Rett Syndrome: Therapies and Parents' Views

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Most of all, though, my thanks go to those families who freely offered their personal thoughts and intimate experiences. It is my hope that schools everywhere will come to understand the unique needs of the children who suffer from Rett syndrome, and offer them the best possible support. It is my hope, too, that the causes of Rett syndrome will be understood sufficiently for the girls to be given the opportunity to enjoy healthy and contented lives.

Patricia Moore, May 2000.
Abstract

Rett syndrome is a profoundly handicapping neurological disorder with an incidence of approximately 1 in 10,000 live female births. The recently acknowledged syndrome has neither known cause nor cure. The nature of the syndrome is particularly distressing for parents, since regression occurs in infants after apparently normal early development. Rapid therapeutic intervention after diagnosis is considered vital in order to ameliorate detrimental stereotypical behaviours and to limit painful physical deterioration. However, there is little published research dealing with the effects of therapies or the manner in which they are delivered in schools.

The National Curriculum in Britain aims to ensure that pupils have access to a broad and balanced education, regardless of ability. The rights of children are recognised in legislation, and it is accepted that some pupils will need specialist support. Recent trends are towards greater involvement of parents in educational issues and towards encouraging the development of partnerships in education.

This research examines therapy provision for girls with Rett syndrome aged between 7 and 12 years, investigates parental opinions regarding effectiveness of therapies in key skill areas of communication, hand function, motor ability and learning ability, and touches upon the broader issues of how essential needs can be considered alongside the rights of children in the classroom environment. The study shows that the roles of teachers and therapists are complementary, and suggests that enhanced professional interaction may be a future objective.

Music therapy, physiotherapy, hydrotherapy and speech therapy are the most common forms of intervention, and the perceived values of each are reported. Inconsistency in provision is apparent, and whilst all therapies are considered beneficial, no single therapy is identified as significantly more beneficial than others.

To consolidate this study, further areas for research are suggested.
Contents

Chapter 1 Rett syndrome – a review of the literature.................................................................1

1.1 Introduction .........................................................................................................................1
1.2 Normal development ............................................................................................................1
1.2.1 The central nervous system .........................................................................................2
1.2.2 Neurological Disorders ...............................................................................................3
1.3 Possible origins ....................................................................................................................5
1.3.1 Genetic clues ...............................................................................................................5
1.3.2 Clinical clues ..............................................................................................................7
1.3.3 Biochemical clues .....................................................................................................7
1.3.4 Infectious and immunological clues .........................................................................8
1.3.5 Summary of the nature of Rett syndrome .................................................................8
1.4 Time course of Rett syndrome .........................................................................................9
1.4.1 Stage 1 / Pre-regression ...........................................................................................10
1.4.2 Stage 2 / Regression ...............................................................................................11
1.4.3 Stage 3 / Early post-regression ...............................................................................12
1.4.4 Stage 4 / Late post-regression ................................................................................13
1.5 Diagnostic criteria .............................................................................................................14
1.5.1 Necessary criteria ......................................................................................................15
1.5.1.1 Motor disorder ..................................................................................................15
1.5.1.2 Stereotypic hand movements .......................................................................17
1.5.1.3 Mental disability .........................................................................................18
1.5.1.4 Communication difficulties .........................................................................20
1.5.2 Supportive criteria ....................................................................................................20
1.5.2.1 Breathing difficulties ..................................................................................21
1.5.2.2 Epilepsy ......................................................................................................21
1.5.2.3 Scoliosis .......................................................................................................22
1.5.3 Exclusion criteria .......................................................................................................22
1.5.4 Common symptoms ..................................................................................................22
1.5.4.1 Emotional difficulties ..................................................................................22
1.5.4.2 Problems with nutrition ..............................................................................23
1.5.5 Atypical Rett syndrome ..........................................................................................23
1.5.6 Differential diagnosis ...............................................................................................24
1.5.7 Summary of symptoms ............................................................................................26
1.5.8 Response to diagnosis ............................................................................................26
1.6 Parental involvement ......................................................................................................27
1.6.1 Parent / professional relationships .........................................................................28
1.7 The education of children with Rett syndrome ..............................................................30
1.7.1 Therapeutic intervention .......................................................................................33
1.7.2 Education and therapy ............................................................................................34
1.8 Summary .........................................................................................................................35

Chapter 2 Therapies – a review of the literature.................................................................38

2.1 Therapies ..........................................................................................................................38
2.2 Physiotherapy ..................................................................................................................38
2.2.1 Developmental issues ..............................................................................................40
2.2.2 Theories of physiotherapy .......................................................................................40
2.2.2.1 Neurodevelopmental treatment ....................................................................40
2.2.2.2 Muscle education and braces ......................................................................41
2.2.2.3 Proprioceptive neuromuscular facilitation (PNF) .......................................42
2.2.2.4 Sensory stimulation ....................................................................................42
2.2.2.5 Progressive pattern movements .................................................................42
2.2.2.6 Patterning ....................................................................................................43
2.2.2.7 Conductive education .................................................................................44
2.2.2.8 Movement education ..................................................................................46
2.2.3 Treatment principles ...............................................................................................46
2.2.3.1 Movement ....................................................................................................47
2.2.3.2 Hand stereotypes ......................................................................................48
2.2.3.3 Scoliosis .....................................................................................................49
2.2.4 Organisation of therapy .........................................................................................49
2.2.5 The role of the physiotherapist .............................................................................50
2.2.5.1 Parental involvement .................................................................................51
2.2.6 Summary ................................................................................................................52
2.3 Speech therapy ...............................................................................................................53
2.3.1 Theoretical approaches to language and language development .........................54
2.3.1.1 The psycholinguistic approach .................................................................54
<table>
<thead>
<tr>
<th>Section</th>
<th>Page</th>
</tr>
</thead>
<tbody>
<tr>
<td>3.5.5 Population</td>
<td>104</td>
</tr>
<tr>
<td>3.5.6 Interview data</td>
<td>105</td>
</tr>
<tr>
<td>3.6 Summary</td>
<td>106</td>
</tr>
<tr>
<td><strong>Chapter 4 Results of the survey</strong></td>
<td>107</td>
</tr>
<tr>
<td>4.1 The survey</td>
<td>107</td>
</tr>
<tr>
<td>4.2 Numbers, ages, and diagnosis</td>
<td>107</td>
</tr>
<tr>
<td>4.3 Abilities</td>
<td>108</td>
</tr>
<tr>
<td>4.3.1 Severity of disability</td>
<td>108</td>
</tr>
<tr>
<td>4.3.2 Importance of improvement</td>
<td>108</td>
</tr>
<tr>
<td>4.4 Therapies</td>
<td>108</td>
</tr>
<tr>
<td>4.4.1 Length of time children had received therapies</td>
<td>110</td>
</tr>
<tr>
<td>4.4.2 Organisation of therapy sessions</td>
<td>111</td>
</tr>
<tr>
<td>4.4.3 Number of hours received</td>
<td>112</td>
</tr>
<tr>
<td>4.4.4 Inclusion of therapy on statement of Special Educational Needs</td>
<td>113</td>
</tr>
<tr>
<td>4.5 Effects of therapies on skill areas</td>
<td>116</td>
</tr>
<tr>
<td>4.5.1 Perceived effect of music therapy</td>
<td>116</td>
</tr>
<tr>
<td>4.5.2 Perceived effect of speech therapy</td>
<td>117</td>
</tr>
<tr>
<td>4.5.3 Perceived effect of hydrotherapy</td>
<td>119</td>
</tr>
<tr>
<td>4.5.4 Perceived effect of occupational therapy</td>
<td>120</td>
</tr>
<tr>
<td>4.5.5 Perceived effect of physiotherapy</td>
<td>120</td>
</tr>
<tr>
<td>4.5.6 Perceived effect of aromatherapy</td>
<td>121</td>
</tr>
<tr>
<td>4.6 Summary</td>
<td>122</td>
</tr>
<tr>
<td><strong>Chapter 5 Analysis and discussion</strong></td>
<td>124</td>
</tr>
<tr>
<td>5.1 Introduction</td>
<td>124</td>
</tr>
<tr>
<td>5.2 Parents’ perceptions of severity of disability</td>
<td>124</td>
</tr>
<tr>
<td>5.2.1 Parents perceptions of importance of improvement</td>
<td>125</td>
</tr>
<tr>
<td>5.2.2 Severity of disability and the importance of improving</td>
<td>126</td>
</tr>
<tr>
<td>5.3 Diagnosis</td>
<td>127</td>
</tr>
<tr>
<td>5.4 Statement of Special Educational Needs</td>
<td>130</td>
</tr>
<tr>
<td>5.4.1 Inclusion of therapy on statement of Special Educational Needs</td>
<td>132</td>
</tr>
<tr>
<td>5.4.2 Therapies given but not specified on statement</td>
<td>135</td>
</tr>
<tr>
<td>5.4.3 The relationship between the statement of Special Educational Needs and whether a therapy is given</td>
<td>135</td>
</tr>
<tr>
<td>5.5 Organisation of therapies</td>
<td>136</td>
</tr>
<tr>
<td>5.5.1 The relationship between the organisation of the therapy and the statement of Special Educational Needs</td>
<td>136</td>
</tr>
<tr>
<td>5.5.2 Perceived benefits of therapies, relating to organisation</td>
<td>137</td>
</tr>
<tr>
<td>5.6 Perceived effects of therapies</td>
<td>142</td>
</tr>
<tr>
<td>5.6.1 Music therapy</td>
<td>142</td>
</tr>
<tr>
<td>5.6.2 Speech therapy</td>
<td>143</td>
</tr>
<tr>
<td>5.6.3 Hydrotherapy</td>
<td>144</td>
</tr>
<tr>
<td>5.6.4 Physiotherapy</td>
<td>146</td>
</tr>
<tr>
<td>5.6.5 Other therapies</td>
<td>146</td>
</tr>
<tr>
<td>5.7 Correlation between variables</td>
<td>147</td>
</tr>
<tr>
<td>5.7.1 Severity of disabilities and the perceived value of therapies</td>
<td>147</td>
</tr>
<tr>
<td>5.7.2 Importance of improving and the perceived value of therapies</td>
<td>148</td>
</tr>
<tr>
<td>5.7.3 The length of time a therapy has been given</td>
<td>149</td>
</tr>
<tr>
<td>5.7.4 Hours per month for therapies</td>
<td>149</td>
</tr>
<tr>
<td>5.8 Perceived differences in values of therapies within a single skill area</td>
<td>150</td>
</tr>
<tr>
<td>5.8.1 Communication</td>
<td>150</td>
</tr>
<tr>
<td>5.8.2 Hand function</td>
<td>152</td>
</tr>
<tr>
<td>5.8.3 Motor ability</td>
<td>153</td>
</tr>
<tr>
<td>5.8.4 Learning ability</td>
<td>154</td>
</tr>
<tr>
<td>5.9 Summary</td>
<td>156</td>
</tr>
<tr>
<td><strong>Chapter 6 Interview Data</strong></td>
<td>158</td>
</tr>
<tr>
<td>6.1 Introduction</td>
<td>158</td>
</tr>
<tr>
<td>6.2 Developmental issues</td>
<td>158</td>
</tr>
<tr>
<td>6.3 Extent of disability</td>
<td>160</td>
</tr>
<tr>
<td>6.4 Physical aspects</td>
<td>161</td>
</tr>
<tr>
<td>6.4.1 Physical aspects – priorities</td>
<td>162</td>
</tr>
<tr>
<td>6.4.2 Physical aspects – perceived value of therapies</td>
<td>163</td>
</tr>
<tr>
<td>6.5 Communication</td>
<td>164</td>
</tr>
<tr>
<td>6.5.1 Communication - with others</td>
<td>166</td>
</tr>
<tr>
<td>Section</td>
<td>Page</td>
</tr>
<tr>
<td>------------------------------------------------------------------------</td>
<td>------</td>
</tr>
<tr>
<td>6.6 Hand function</td>
<td>169</td>
</tr>
<tr>
<td>6.6.1 Hand function - stereotypical behaviour</td>
<td>170</td>
</tr>
<tr>
<td>6.6.2 Hand function - modification of hand movements</td>
<td>171</td>
</tr>
<tr>
<td>6.6.3 Hand function - perceived value of therapies</td>
<td>172</td>
</tr>
<tr>
<td>6.7 Learning ability</td>
<td>173</td>
</tr>
<tr>
<td>6.7.1 Learning ability - priority areas</td>
<td>174</td>
</tr>
<tr>
<td>6.7.2 Learning ability - perceived value of therapies</td>
<td>175</td>
</tr>
<tr>
<td>6.8 Summary of the value of therapies</td>
<td>177</td>
</tr>
<tr>
<td>6.8.1 Speech therapy</td>
<td>177</td>
</tr>
<tr>
<td>6.8.2 Music therapy</td>
<td>177</td>
</tr>
<tr>
<td>6.8.3 Physiotherapy</td>
<td>178</td>
</tr>
<tr>
<td>6.8.4 Hydrotherapy</td>
<td>178</td>
</tr>
<tr>
<td>7.1 Areas for further research</td>
<td>181</td>
</tr>
<tr>
<td>7.1.1 Therapies and parents' knowledge</td>
<td>181</td>
</tr>
<tr>
<td>7.1.2 Interactions between therapies</td>
<td>181</td>
</tr>
<tr>
<td>7.1.3 Funding</td>
<td>181</td>
</tr>
<tr>
<td>7.1.4 National Curriculum</td>
<td>182</td>
</tr>
<tr>
<td>7.1.5 Teachers' perceptions</td>
<td>182</td>
</tr>
<tr>
<td>7.1.6 Placement</td>
<td>182</td>
</tr>
<tr>
<td>7.1.7 Needs of families</td>
<td>183</td>
</tr>
<tr>
<td>7.1.8 Alternative therapies</td>
<td>183</td>
</tr>
<tr>
<td>7.2 Educational implications of Rett syndrome</td>
<td>184</td>
</tr>
<tr>
<td>7.2.1 Policies for Excellence</td>
<td>184</td>
</tr>
<tr>
<td>7.2.1.1 Early identification</td>
<td>185</td>
</tr>
<tr>
<td>7.2.1.2 Early intervention</td>
<td>186</td>
</tr>
<tr>
<td>7.2.2 Working with parents</td>
<td>186</td>
</tr>
<tr>
<td>7.2.3 Improving the SEN framework</td>
<td>187</td>
</tr>
<tr>
<td>7.2.3.1 Statements</td>
<td>189</td>
</tr>
<tr>
<td>7.2.3.2 Individual Education Plans</td>
<td>190</td>
</tr>
<tr>
<td>7.2.4 Inclusion</td>
<td>191</td>
</tr>
<tr>
<td>7.2.4.1 Value of therapies</td>
<td>193</td>
</tr>
<tr>
<td>7.2.4.2 Financial issues</td>
<td>195</td>
</tr>
<tr>
<td>7.2.5 Developing skills</td>
<td>196</td>
</tr>
<tr>
<td>7.2.5.1 Role of teachers</td>
<td>197</td>
</tr>
<tr>
<td>7.2.5.2 Role of learning support assistants</td>
<td>198</td>
</tr>
<tr>
<td>7.2.6 Multi-professional approach</td>
<td>198</td>
</tr>
<tr>
<td>7.3 Summary</td>
<td>200</td>
</tr>
</tbody>
</table>
Figures

Figure 1  Number of girls receiving each therapy ................................................................. 107
Figure 2  Parents' perceptions of the severity of disability in four skill areas ......................... 108
Figure 3  Parents' perception of the importance of improvement in each skill area ................. 109
Figure 4  Length of time children had received therapies ................................................... 110
Figure 5  The organisation of therapy sessions .................................................................... 111
Figure 6  Number of hours per month .................................................................................. 112
Figure 7  The inclusion of therapies on statements of Special Educational Needs ................. 113
Figure 8  Perceived effect of music therapy on skill areas .................................................... 117
Figure 9  Perceived effect of speech therapy on skill areas .................................................. 118
Figure 10  Perceived effect of hydrotherapy on skill areas .................................................... 119
Figure 11  Perceived effect of occupational therapy on skill areas ....................................... 120
Figure 12  Perceived effect of physiotherapy on skill areas .................................................. 121
Figure 13  Perceived effect of aromatherapy on skill areas .................................................. 122
Tables

Table 1  Comparison of characteristics of Rett syndrome and childhood autism ................................. 25
Table 2  Parents’ perceptions of severity of disability............................................................................ 109
Table 3  Parents’ perceptions of the importance of improvement.......................................................... 110
Table 4  The number of children receiving each therapy, related to its inclusion on statement of Special Educational Needs................................................................. 114
Table 5  The organisation of music therapy in 30 Rett cases ................................................................. 115
Table 6  The organisation of speech therapy in 30 Rett cases ................................................................. 115
Table 7  The organisation of hydrotherapy in 30 Rett cases ................................................................. 116
Table 8  The organisation of physiotherapy in 30 Rett cases ................................................................. 116
Table 9  Total and average scores of value of music therapy on each skill area .................................. 117
Table 10  Total and average scores of value of speech therapy on each skill area ............................. 118
Table 11  Total and average scores of value of hydrotherapy on each skill area................................ 119
Table 12  Total and average scores of value of physiotherapy on each skill area ............................... 121
Table 13  Perceived differences in severity of disability against each skill area .................................... 125
Table 14  Perceived differences in importance of improvement against each skill area ..................... 126
Table 15  Correlation between severity of disability and the importance of improving in each of the four skill areas ........................................................................................................ 127
Table 16  Relationships between statements and whether therapy is given ........................................ 136
Table 17  Relationships between how therapy is organised and statements of Special Educational Needs, all therapies included ........................................................................................................... 137
Table 18  Perceived benefits of music therapy, relating to organisation - average scores .................... 138
Table 19  Perceived benefits of speech therapy, relating to organisation - average scores .................... 139
Table 20  Perceived benefits of hydrotherapy, relating to organisation - average scores .................... 140
Table 21  Perceived benefits of physiotherapy, relating to organisation - average scores .................... 141
Table 22  Differences in perceived value of music therapy on skill areas .......................................... 142
Table 23  Differences in perceived value of speech therapy on skill areas ......................................... 144
Table 24  Differences in perceived value of hydrotherapy on skill areas .......................................... 145
Table 25  Differences in perceived value of physiotherapy on skill areas .......................................... 146
Table 26  Correlation between the perceived severity of disability and value of therapy .................... 147
Table 27  Correlation between the perceived importance of improving and value of therapy ............. 148
Table 28  Correlation between length of time therapy has been given and perceived value of therapy ........................................................................................................................................ 149
Table 29  Correlation between monthly quantity of therapy and perceived value of therapy .......... 150
Table 30  Differences in effect of therapies on communication ............................................................ 151
Table 31  Differences in effect of therapies on hand function ............................................................... 153
Table 32  Differences in effect of therapies on motor ability ................................................................. 154
Table 33  Differences in effect of therapies on learning ability ............................................................. 155
Table 34  Developmental issues of girls with Rett syndrome ............................................................... 159
Table 35  Greatest overall difficulties .................................................................................................... 160
Table 36  Current physical abilities ....................................................................................................... 161
Table 37  Gross motor priorities ........................................................................................................... 162
Table 38  Value of therapies in assisting gross motor skills .................................................................. 164
Table 39  Means of communication ....................................................................................................... 165
Table 40  Ability to communicate with outsiders .................................................................................. 166
Table 41  Priority areas for communication and how they are addressed ............................................ 167
Table 42  Value of therapies in assisting communication skills ............................................................ 168
Table 43  Current hand function ........................................................................................................... 170
Table 44  Stereotypical hand behaviours ............................................................................................... 170
Table 45  The importance of modifying hand movements ...................................................................... 171
Table 46  Value of therapies in assisting hand function ......................................................................... 172
Table 47  Parents’ perceptions of current level of learning ability ......................................................... 174
Table 48  Learning ability priority areas ................................................................................................. 175
Table 49  The value of therapies in assisting learning ability .................................................................. 176
Table 50  The four clinical stages of Rett syndrome ............................................................................. 203
Table 51  Necessary criteria .................................................................................................................. 204
Table 52  Supportive criteria ................................................................................................................ 205
Table 53  Exclusion criteria .................................................................................................................. 205
Table 54  Criteria for formes frustes ..................................................................................................... 206
Chapter 1 Rett syndrome – a review of the literature

1.1 Introduction

This study looks at the views of thirty parents of children with Rett syndrome, a low incident, profoundly disabling neurological disorder which to date has only been found in girls. Medical research has progressed significantly in recent years, but there has been little published work relating to aspects of the education and the therapies girls with Rett syndrome receive in school (Lindberg, 1994, Lewis and Wilson, 1998). Parents have the most intimate contact with their daughters, yet their collective opinions are not widely reported. This study examines therapies in an educational context, provides some theoretical rationale for the different interventions, and in particular, asks the question 'Do parents demonstrate preferences in relation to therapeutic management?' Detailed research questions, resulting in part from this generic question, and in part from a review of the literature, can be found in Section 3.3. Although specifically dealing with Rett syndrome, the study has relevance to other low incidence conditions and conditions within the autistic spectrum.

1.2 Normal development

It is inappropriate to consider the pathology and consequences of any disabling condition without reference to normal patterns of development, and the individual's condition should be understood in context with anticipated child development before remedial action is taken. As well as offering a sound foundation, knowledge of normal developmental patterns can give therapeutic intervention greater sensitivity (Davies, 1997). If a therapist only deals with, or has knowledge of, children who display abnormalities, there could arise a normalising effect or a desensitising regarding a child's particular circumstances. Awareness of other patterns of development, including other conditions and including normal development, enhances the therapist's skill in planning and conducting therapeutic intervention appropriate to specific disorders.

This section looks at normal development in general, and begins to investigate abnormalities engendered by neurological disorders. Development is frequently classified in terms of physical, mental (including language), emotional and social categories, but it is important to remember that these areas overlap and interact to complete a whole person. Acquisition of new skills, increased understanding, social
interaction, and physical growth are among the processes of development, which Holt (1991) considers to be a complex unfolding and expanding, while moving toward a state of completion. Satisfactory development requires not only an appropriate environment and adequate nutrition plus opportunities to learn reinforced by challenges and rewards, but also a well formed and normally functioning body and brain (Atkinson, 1974). A disturbance to any of these factors affects the whole child.

1.2.1 The central nervous system

One of the first organised body structures to develop after conception is the central nervous system. The formation of the brain and spinal cord starts as a strip of ectodermal cells running the length of the embryo. As the embryo grows, other tissues close over the strip, eventually to form the skull and the vertebrae. The neurones, or nerve cells, of the central nervous system differ from other cells in their ability to receive and transmit stimuli to other cells that are adapted to generate movement. Each neurone consists of a cell body, with several dendritic processes and a single long process, the axon, which may extend to a significant proportion of the body length. The parts of the neural tube in the head expand to form the brain while the remainder forms the spinal cord. The development of the nervous system continues and new nerve cells are added until the end of the first year after birth (Wingate, 1979, Atkinson, 1974, Donnai, Kerzin-Storrar and Wigmore, 1993). Aggregates of conducting processes develop into the nerve fibres of the body, their ability to conduct enhanced by a sheath of fatty myelin that serves to accelerate conduction, and to protect and heal.

The central nervous system facilitates reaction of the body to the environment. The system, though largely dealing with reflex actions is concerned with physical (motor, sensory and autonomic), intellectual and emotional activities and, in consequence, any dysfunction may involve any one or all three of these major functions (Atkinson, 1974). The greatest number of neurones is contained within the brain, in particular within the cerebrum. The early reflex activities and movement responses develop long before birth, although at birth the central nervous system is not completely myelinated and is therefore incapable of functioning in a mature manner. With regard to human development, maturation refers to the elaboration of structure and function of the nervous system (Cash, 1974). As the nervous system matures,
Some reflex pathways become more dominant for a period before becoming modified and integrated into more mature movement patterns. If there is disturbance of the synaptic mechanism, or damage or dysfunction within the central nervous system, the development of the more mature neurological reactions will be delayed, and the pattern of development both in sequence and in the pattern of each reaction may be disordered (Levitt, 1984).

1.2.2 Neurological Disorders

When, for whatever reason, development processes are interrupted or do not follow normal pathways, the effects can vary between dramatic and almost unnoticeable. To avoid the potential complications associated with terms such as 'disease' and 'illness', the term 'disorder' is frequently used. Whereas diseases affect the nervous system through secondary interference with structure (Wingate, 1979) the term disorder is used to imply the existence of a clinically recognisable set of symptoms or behaviour associated with interference with personal functions in the absence of a causative disease (World Health Organization, 1992).

The World Health Organization (WHO) International Classification of Diseases identifies a category of pervasive developmental disorders, containing childhood autism, atypical autism, Rett syndrome, other childhood disintegrative disorder, overactive disorder associated with mental retardation and stereotyped movements, and Asperger's syndrome. In these neurological developmental disorders, the all-encompassing features of social abnormality, restricted and stereotyped interests and activities, and communication patterns are a 'pervasive feature of the individual's functioning in all situations, although they may vary in degree' (WHO, 1992, p.252).

The Diagnostic and Statistical Manual of Mental Disorders IV (American Psychiatric Association, 1994) describes Autistic Spectrum Disorder (ASD) as a pervasive developmental disorder. The spectrum is defined by presence of impairments in social interaction, in communication, and in behaviour, (lack of flexibility in thinking and behaviour manifest in repetitive and stereotyped patterns of behaviour, interests and activities).

These two diagnostic systems have agreement in their use of this triad of impairments for diagnosis of autism and other medical conditions that share
common developmental difficulties (Jordan, Jones and Murray, 1998). A comparison of the characteristics of Rett syndrome and the characteristics of childhood autism, based on these impairments can be found in Section 1.5.6. Jordan (1999a) suggests that Rett syndrome is appropriately placed within the autistic spectrum, and that similar teaching approaches are beneficial for both groups, particularly during childhood. However, it will be seen that there are differences between childhood autism and Rett syndrome that may have implications in the classroom.

Rett syndrome is, then, a pervasive developmental disorder within the autistic spectrum, that follows a very specific progression, and that has implications for other disorders. ‘... the Rett syndrome is an excellent model for many such neurological diseases of this kind, and for what should be done to increase the knowledge of all aspects from the development of scoliosis to the relief of symptoms’ (Gordon, 1993, p. x).

Unusual repetitive patterns of behaviour in the form of hand movement were a key factor in Rett's appreciation of the condition. Haas describes how Rett syndrome was first discovered in Vienna, in 1965. In Professor Andreas Rett's own words:

One day in the Spring of 1965, two mothers holding their children on their laps were sitting in the waiting room. Both children were swaying and the mothers held their arms. Both children, who were treated for epileptic seizures, were well known to me. That morning I passed them repeatedly and, incidentally, the mothers let go of their children's arms. At once, the children put their hands together and started nearly identically-looking washing movements. I asked the mothers not to stop these movements again, and was startled at the similarities. It was the same gaze, same facial expression, same weak muscles and the same stereotyped movements of their hands. Moreover, they were both females. At first, I was amazed by this enormous coincidence and thought I must have come across several other children with these symptoms. I asked my head nurse to look at these children and she immediately recalled some more names of children who behaved similarly. We invited them, placed them next to each other and found that all of them showed the same kinds of movements. Further, all patients were female.

(Haas, undated)

The original publication of Rett's findings, which characterised the stereotyped neurological disease that now bears his name, appeared in 1966 in the Austrian
medical newsletter, Wiener Medizinische Woehenschrift. This clinical paper, like most of Rett's publications, was not widely circulated, and it remained unnoticed in the English-speaking world. The first English description appeared in 1977 (Rett, 1977), but it was an article published in 1983 by a Swedish doctor and his colleagues in a widely-read English language neurology journal that brought Rett syndrome to the attention of the English-speaking world (Hagberg, Aicardi, Dias and Ramos, 1983). The historical significance of Rett's earlier descriptions was acknowledged by these authors in this paper.

From this time, awareness of the syndrome grew and previously unidentified cases were subsequently diagnosed (Hagberg, 1993). Today, Rett syndrome is known to exist all over the world, in all ethnic groups, and with an incidence of approximately 1 in 10,000 live female births, (Rett Syndrome Association United Kingdom, 1999b) and Andreas Rett is now recognised as a pioneer in the care of mentally and physically handicapped children in Austria (Hunter, 1999). Support organisations are seeking to publicise the condition, and in Britain the Rett Syndrome Association United Kingdom (RSAUK, formerly the UK Rett Syndrome Association, UKRSA) has published many informative booklets and leaflets, and has promoted research into the causes and treatments of the syndrome.

1.3 Possible origins

Rett syndrome is, in the female sex, one of the major clinically defined conditions within the concept of a severe mental disability, yet it still constitutes a major clinical, biological and genetic mystery (Hagberg, 1995b). In his original paper, Rett reported raised ammonia levels in the blood, and concluded that the illness was due to an inborn biochemical disturbance of nitrogen metabolism (Wilson, 1986). He later withdrew this claim because he doubted the accuracy of older methods of ammonia measurement. Recent work has concentrated on the search for a genetic origin (Armstrong, 1999).

1.3.1 Genetic clues

The absence of boys with classic Rett syndrome suggests a genetic fault on the X chromosome (Kerr, 1994a) and an extensive search for molecular genetic X chromosome abnormalities has been performed in recent years (Anvret, Wahlström and Äkesson, 1993). This suggestion is supported by work of Amir, Veyver, Wan, Tran, Franke and Zoghbi (cited by Armstrong, 1999) who have recently reported
mutations on the X chromosome of 8 out of 23 Rett patients, on a gene which codes for a regulator protein. Without the regulator, other proteins are over-expressed, although the form and effect of over-expression are not yet known.

The effects of a genetic fault on one chromosome may be compensated for by the existence of a functioning equivalent gene on the other member of the pair. In the case of the sex chromosomes, a female child inherits an X chromosome from each parent while the male inherits a maternal X and a paternal Y. Possession of only one X renders the male more vulnerable than the female to a faulty gene on the X chromosome. If a fault develops in a gene with a major role in organising brain development, as is suspected in Rett syndrome, (Kerr, 1994a) the male foetus cannot survive whereas the female survives, but is impaired. If this were the case, one would expect fewer brothers than sisters of girls with Rett syndrome (Äkesson, Hagberg, Wahlström and Engerström, 1992). These authors suggest that the size of population needed for such a study makes it unfeasible, although a study of 30 families (Fyfe, Leonard, Dye and Leonard, 1999) found no disturbance in the siblings sex ratio when case and control families were compared. It should be possible to access families of girls with Rett syndrome through bodies such as the International Rett Syndrome Association (IRSA) and the RSAUK, and this is an area suited to long term research.

The genetic basis of Rett syndrome is supported by the existence of familial cases and by concordance among monozygotic twins, which are genetically identical, and discordance among dizygotic twins, which are genetically different from one another (Zoghbi, 1988). In 1992, the IRSA register included 7 concordant monozygotic twin pairs and 11 discordant dizygotic twin pairs. Of the dizygotic twin pairs, four are female / female and seven are female / male (Anvret, Wahlström and Äkesson, 1993).

Collected data relating to families with more than one Rett syndrome female supports a genetic transmission, working through both maternal and paternal lines (Hagberg, 1993). Genealogical data from Sweden, going back over seven to ten generations, also conform to a genetic inheritance, albeit of quite complex and so far unexplained type (Äkesson, Hagberg, Wahlström and Witt-Engerström, 1992).

The same research suggests that a significantly increased consanguinity rate, in both maternal and paternal lineages, contributes further support to the genetic basis
of Rett syndrome. Witt-Engerström (in Kerr and Corbett, 1994) cited the case of a woman with classic Rett syndrome giving birth to a girl with classic Rett syndrome, the child being more disabled than her mother, suggesting dominant inheritance.

The search for the genetic origin of Rett syndrome is a specialised field involving microbiological and demographic research and is an area that has generated much interest to researchers in recent years. Personal experience does not extend to hereditary or genetic matters, and such work is beyond the scope of this study.

1.3.2 Clinical clues

As understanding of Rett syndrome grows, so the condition becomes more distinct from other progressive neurological diseases. Increasing motor impairment is typical of this type of condition, but there is a characteristic remission in Rett syndrome that is not typical (Hagberg, Anvret, Percy and Wahlström, 1993). Diseases such as Parkinson and Alzheimer show progressive and permanent degeneration due to ageing changes in already mature brains of males and females, but Rett syndrome affects only infant girls. Kerr (1992a) believes Rett syndrome is better viewed as 'a developmental disorder in which the evolving clinical signs are due to the effects of maturation and ageing on an abnormal brain rather than to primary degenerative disease' (p. S43).

1.3.3 Biochemical clues

Another dissimilarity between Rett syndrome and progressive degenerative diseases can be found in biochemical research. ‘Only exceptionally in a progressive condition does one see such a relative chemical normality associated with such extensive neurological impairments’ (Percy, 1988, quoted in Hagberg, Anvret, Percy and Wahlström, 1993, p.112).

Efforts to trace deviating biological parameters of primary importance (metabolic derangements, hormonal deficiencies, and chromosomal aberrations) have all been unsuccessful. However, the deceleration of skull growth in early infancy, along with an abnormal deficiency of brain cells, support either a defect in the arrest of programmed normal neuronal cell death or lack of a specific brain growth factor (Hagberg, 1995b). This suggestion adds strength to Kerr’s view of the syndrome as a developmental disorder relating to the maturing of an infant brain.
1.3.4 Infectious and immunological clues

Parents of girls with Rett syndrome may initially associate the manifestation of the symptoms with various common infections, particularly of the upper respiratory or gastrointestinal tracts. Some girls start the period of regression abruptly, causing encephalitis to be suspected. This suggests the possibility that Rett syndrome may be due to a slow virus state. However, if a virus does have a role in Rett syndrome, it is difficult to explain why it only affects females, and at present there is no supporting evidence for immunological dysfunction in this disorder (Hagberg, Anvret, Percy and Wahlström, 1993). There is a lack of detailed research into this area, but personal experience does not support activation of Rett syndrome by common infections. Prior to this study, out of four cases encountered personally, there was no association with suggested triggers.

1.3.5 Summary of the nature of Rett syndrome

On current knowledge it is still unclear whether Rett syndrome is an inherited disorder and if so which specific chromosomal region is involved. The hypothesis mostly favoured is an X linked dominant inheritance that is lethal in males. Neither clinical nor epidemiological data contradict this, but equally no positive molecular evidence has so far been found despite intensive investigation (Anvret, Wahlström and Åkesson, 1993).

Rett syndrome appears to be neither a neurodegenerative nor a traditional neurometabolic condition. An age-limited neurodevelopmental, as yet unknown, brain growth deficiency is at present indicated (Kerr, 1992a). There is still much work to be done in the field of research, since at present there is no biological marker, an incomplete understanding of the cause and development of the disease, and few clues regarding the type of genetic transmission. Hagberg (1995b) believed that ‘Combined efforts from brain research, genetics and molecular biology may hopefully soon lead to identification of diagnostic markers and causative factors for RS’ (p.976).

The real effects of Rett syndrome are severe loss of communication skill, loss of mobility, loss of fine motor skill, and in the broadest sense, significant reduction of the ability to learn. The cause and development are still uncertain, but this does not mean that attempts to incorporate the children within the normal educational framework should not be made. Part of the difficulty of knowing what is right arises
because the cause is not understood; therefore a programme of education cannot be built upon medical foundations. Neither can the child be exempt from the legislation which directs education on a national scale. The degree to which medical necessity and legislation interact may vary from county to county, and from one medical condition to another, and it is not improbable that there is compromise which may not be in the best interest of the child. It will be seen that mainstream education is likely to be inappropriate for girls with Rett syndrome, and that special measures are essential for their education and wellbeing.

1.4 **Time course of Rett syndrome**

The classic Rett syndrome begins very early. Girls are born apparently healthy and their psychomotor development is apparently normal up to 6 - 18 months. This is followed by a period when learning achievement stops and acquired skills are impaired or lost. Regression brings new behaviours ranging from screaming and inconsolable crying, to total withdrawal and isolation. In time some improvement is noted, particularly in social aspects, and a number of specific symptoms appear which allow a tentative diagnosis (Lindberg, 1990).

Autistic spectrum disorders are not detectable at birth, although Kerr (1995a) insists that a close scrutiny of classic Rett syndrome cases indicates that the cognitive and motor problems of this particular disorder are detectable from birth even though the child looks normal and makes early progress within the accepted range. Video recordings support families' accounts of early communication and manipulative skills, but also show pre-regression children to be poorly mobile with an excess of repetitive limb and trunk movements. These abnormalities, while quite subtle and usually missed by routine observations or tests under nine months of age, nevertheless signify the onset of the disease well before the regression period (Witt-Engerström, 1987).

It was over twenty years after the first recognition of the syndrome that attempts were made to describe the progress of the condition. Hagberg and Witt-Engerström (1986) constructed a staging system to facilitate the characterisation of the patterns and profiles of Rett syndrome from infancy to adolescence. This was based on four clinical stages, derived from a synthesis of clinical observations over a number of years in 50 Swedish cases. Whilst acknowledging that this was to some extent a simplistic framework, the authors believed this staging system to be
a useful instrument for an approach to the complex clinical symptoms of Rett syndrome. This was adapted by Hagberg (1989) and a substaging of certain parts was introduced by Witt-Engerström (1990) (Appendix I). An alternative approach is taken in Britain, where doctors and researchers refer to the level of regression (Kerr and Corbett, 1994), rather than to stages. The sub-type is reported according to the predominant abnormality of tone i.e. hypotonic, variably increased tone but retained ambulation, and rigidity and inability to walk (Kerr and Stephenson, 1985).

The two approaches describe a single syndrome, and both have aptness. Kerr (in Kerr and Corbett, 1994) relate the regressional basis to levels of cortical incompetence, and the referral to the level of regression does give recognition to what is an important characteristic of Rett syndrome. It also gives a clear indication of general status, whereas the staging notation implies a stepped progression. However, by definition, pre-regression as a stage cannot be recognised until regression occurs, but recognition of the 'stagnation' of stage 1 is possible.

The British Education system is accustomed to dealing with stages, but from the point of view of a parent just coming to terms with Rett syndrome, the language of the original model could be rather daunting. Although the staging system was described as 'sometimes crude and a somewhat simplistic frame' by the very people who introduced it (Hagberg and Witt-Engerström, 1986, p.58), it was considered a necessary step for outlining the crucial features of a lifelong disorder (Witt-Engerström, 1990). Many interpretations have been put upon certain characteristics, and the literature describing the stages of Rett syndrome is often vague or based on small samples. What may be required now is revision of the terminology, perhaps based on the International Classification of Mental and Behavioural Disorders (ICD-10) (WHO, 1992, WHO, 1993), but with concise descriptions, relating to specific stages which would encompass the requirements of professionals and be easily understood by parents. To some extent, each existing model is appropriate, and the following summary draws on both, and on literature from established authorities.

1.4.1 Stage 1 / Pre-regression

Stage 1 referred to the early onset stagnation period when the developmental profile was still characterised by an unspecific, general slowing, rather than an established deterioration (Witt-Engerström, 1990). This stage can begin when the
child is between 6 and 18 months of age, and the duration is generally a few months. Until this time the child has progressed and acquired new skills, although possibly at a slower rate than would have been expected. Most girls with Rett syndrome are able to speak with single words or short phrases. Successful feeding is usual and self-feeding with fingers or even a spoon is not uncommon, nor is the ability to reach for objects, build with bricks, choose appropriate holes for shapes or even operate switches. This stage is seen as a consequence of increasing cortical incompetence (Kerr, 1994d).

In a study of 28 class teachers, relating to 42 girls with Rett syndrome it was reported that great difficulty was experienced in focussing the child's attention (Lewis and Wilson, 1996). Non-specific, episodic hand waving may occur alongside functional use of hands. The rate of skull growth decreases at approximately 8 months, and the final head circumference rarely exceeds 52 centimetres. This decline in head circumference growth is followed by a decline in total body weight (Armstrong, 1992).

1.4.2 Stage 2 / Regression

Stage 2 was defined as the stage of deterioration, when obvious loss of acquired abilities and skills occurred (Witt-Engerström, 1990). Kerr (1995a) described this period as 'silent but dramatic'. Between 1 and 3 years of age a period of regression begins and lasts for some weeks or months. This is regarded as the cortical crisis (Kerr 94d) and is characterised by a general developmental deterioration with loss of acquired skills, including the ability to use hands functionally. Typical hand stereotypies manifest themselves. Gross motor skills are often affected, with unsteady, unco-ordinated and jerky movements becoming common (Lindberg, 1994). Speech is severely disrupted and most girls rarely use more than a single word utterance. This regression may be sudden, gradual, or in steps, apparently precipitated by seemingly trivial events or illnesses. The child may show signs of distress, including episodes of screaming; relationships are interrupted to such a degree that autism may be suspected, and sleep is interrupted. Some girls simply stop using their skills whilst most show clearly disturbed muscle tone and involuntary movements which affect the whole body, particularly the hands and mouth (Kerr, 1994d). Such developments obviously have educational implications and these are discussed in Chapter 5.
Onset of the typical alternating hyperventilation and apnoea of Rett syndrome usually follows the other regression signs and may do so after an interval of months or even years, suggesting that the trigger is a maturational step parallel to but distinguishable from that which precipitates the rest of the regression event (Kerr, 1995a).

There has been no evidence of upper or lower motor neurone abnormality at this early stage, and the primary motor defect appears to be cortical (Eyre, Kerr, Miller, O'Sullivan, Ramesh, 1990). Kerr (1994d) reports that the time of arrival at this 'developmental ceiling' suggests an abnormality of higher centre processing and evidence of the essentially developmental and cortical character of the disease, since late infancy is the time when cortical networks undergo rapid maturation and their influence becomes increasingly evident. The normal infant also progresses in 'dips and drops' (von Euler, Forssbarg and Lagercrantz, 1989) and this is thought to be due to reorganisation of existing mechanisms of the nervous system for more adaptable modes of operation. It is now thought that it is in the course of such reorganisation that the child with Rett syndrome begins to regress. By the end of this period it is obvious that the child is profoundly disabled with a level of skill under that expected at one year (Kerr, 1994d).

1.4.3 Stage 3 / Early post-regression

Stage 3 was defined as the stage without further mental deterioration, when the bottom level for hand use and speech ability had been reached, while acquired gross motor abilities were relatively well spared (Witt-Engerström, 1990). This early post-regression stage can last for many years, from pre-school into school years. The girl no longer regresses in her development but stabilises at the level reached. Kerr (1994d) believes that the infant skills achieved before regression remain substantially intact, but in some unknown way they become inaccessible. The post-regression child may regain some use of her hands and she may experience improvement in communication. The capacity for words never completely disappears, and one perplexing feature of the syndrome is the rare, sudden, complex and complete sentence uttered by the child in an appropriate context (Zappella, 1992).
Emotional contact with surroundings improves, and gross motor abilities are still largely maintained, with only slow deterioration. Truncal ataxia with poor muscle co-ordination is prominent, and epileptic seizures are common (Kerr 1994d).

Whether or not seizures follow regression, the electroencephalogram becomes abnormal but although characteristic of Rett syndrome (Lugaresi, Cirignotta and Montagna, 1985), the abnormality is not diagnostic. The irregularity is most marked in sleep and is diminished when the child is alerted (Southall, Kerr, Tirosh, Amos, Lang and Stephenson, 1988).

The presence of considerable infant skill remains an important feature of Rett syndrome and is often discounted when viewed from the perspective of later disabilities (Kerr, 1994d). Unfortunately, many professionals, including educationalists and therapists, are unaware of this aspect of Rett syndrome, and until all features are recognised and understood, they may not be able to take full advantage of this important characteristic.

1.4.4 Stage 4 / Late post-regression

In stage 4, gross motor deterioration accelerates and physical deformities are inclined to worsen. Most girls develop scoliosis, a condition apt to progress, especially in the most hypotonic and sedentary girls. Weakness, wasting, spasticity and scoliosis make most girls non-ambulatory, with much time having to be spent in a wheelchair (Witt-Engerström, 1990). Limbs tend to be held rigid during voluntary effort and a variety of fixed deformities of the feet may develop. Children who do not walk may suffer dislocated hips. It is important that measures, generally therapeutic, be introduced to reduce the rate of deterioration, and it is in a school environment where they are usually carried out. There is an obvious trade-off between 'subject-based' education and 'fitness for life' education. While one of a school's specific targets might be attainment related, in the case of conditions such as Rett syndrome, there really should be no compromise with health and welfare, even if the more formal aspect of education is damaged as a result. The health and welfare of all children are important, but severe low incidence disability cases are exceptionally vulnerable, and children may have uncertain life expectancy. Mainstream children do not have the same imperative in terms of medical support, so it is unlikely there will be a need for compromise. There are conditions more damaging than Rett syndrome that fit less comfortably within the education
framework. Progeria, for example, with its certainty of death in or soon after the teen years, raises difficult questions regarding the purposes of preparing for adulthood. No child should be denied essential care, but for some there may be a case for withdrawing somewhat from the formal learning routine.

Kerr (1988) found no evidence of inevitable continuing mental deterioration in this stage but she noted some improvement in emotional contact. Epileptic seizures become less problematic and easier to control with medication. It is not unusual for girls to survive into the fourth decade. Although the physical disabilities tend to increase, this seems to be in a comparable fashion to those that follow severe birth injury, where non-progressive damage to the brain manifests itself in new ways at different ages.

Therefore, the course of the disease is predetermined, with specific symptoms and disabilities. However, there are variations in the visible onset of the disorder as well as in the speed and degree of the deterioration. With increasing age and advanced stage of disease, the final differences in severity of motor disability, pattern of neurology and degree of impairment are striking (Hagberg, 1995b). There may be ambulant Rett syndrome adults, not dissimilar to other uncomplicated severely mentally disabled adults, while others may never have learned to walk independently, some remaining completely helpless, with secondary body deformities. These may conceal the original clinical features (Garber and Veydt, 1992) and may cause great difficulties in obtaining correct diagnosis in adults when detailed data of early history is not available.

1.5 Diagnostic criteria

Rett syndrome has only become widely recognised during the 1980s and it is likely that many girls and women remain undiagnosed. The onset of the disorder and rapidity of its progress varies with each case, as does the intensity of the symptoms. In its early stages, many of the individual characteristics of Rett syndrome are common to other disorders and possible misdiagnoses can include autism, Angelman syndrome, tuberous sclerosis, cerebral palsy or epilepsy, while in its later stages, diagnoses of cerebral palsy, spinocerebellar degeneration or unknown degenerative disorder may have been made (Perry, 1991, Hagberg, 1993) and these need to be ruled out before Rett syndrome can be confirmed.
As yet, there are no laboratory tests that can confirm the clinical diagnosis. Because of this absence of accurate biological markers, diagnostic criteria for Rett syndrome have been set up. These were first constructed in 1984 at the 2nd International Rett Syndrome Conference in Vienna (Hagberg, Goutières, Hanefeld, Rett and Wilson, 1985) and served as a model for the necessary, supportive and exclusion criteria presented in 1988 by the professional advisory board of the IRSA and the Centers for Disease Control in the USA (Trevathan and Moser, 1988). An updated version was presented by Hagberg (1993) (Appendix II). The criteria were intentionally kept strict so that other developmentally deviating disorders were not falsely included (Hagberg, 1995b). The differential diagnosis of Rett syndrome is discussed in Section 1.5.6.

Anatomical structure and physiological function in conditions of neurological developmental disorders are inextricably linked; not only does structure determine function, but also function modifies structure (Baddeley, 1984). Impairments are described as problems in body function or structure as a significant deviation or loss (WHO, 1999). In the following section, structure and function are considered in each of the main areas of disability.

1.5.1 Necessary criteria

There are problems and disabilities which are so common amongst girls with Rett syndrome, that they must be considered characteristic of the disorder (Lindberg, 1994, Witt-Engerström, 1993). These symptoms can be categorised broadly into four areas, relating to motor disorder, stereotypic behaviours, mental disability, and communication difficulties. It is only when these symptoms and behaviours are fully understood that progress can be made towards treating this disorder and addressing educational issues, including implementing appropriate therapeutic and educational programmes.

1.5.1.1 Motor disorder

The development of motor function is focused on balancing and independent movement. Witt-Engerström (1993) interprets the early signs of Rett syndrome and their sequential occurrence as a reflection of dysfunction or damage to certain neural circuits at cortical and sub-cortical levels at an early stage of development. This deprives the girls of necessary control mechanisms, resulting in 'very immature and deranged behavioural and motor patterns' (p.37).
Girls with Rett syndrome have limited mobility in all respects. Both gross and fine motor skills are impaired and after the regression period, girls are left with paucity of voluntary movements, poor co-ordination, and jerky, writhing involuntary movements which are most marked in the hands and arms but also affect the lower limbs, trunk and face (Hagberg, Aicardi, Dias, and Ramos, 1983). Lindberg (1994) considers the inability to carry out purposeful movements and actions to be ‘apraxia’ and suggests that although it has an overall effect on the child, its consequences are most evident in speech, fine motor skills and complex gross motor skills. She describes apraxia as being one of the most fundamental handicaps in Rett syndrome, affecting almost all active behaviours. Not only does it make it difficult to get a true picture of the intellectual capacity of an individual, but it also affects her emotional development, identity and self-image. Witt-Engerström (1993) suggests that an affected area of the brain is unable to play any direct role in causing positive symptoms, and that negative symptoms are likely to be due directly to loss of function of the damaged part.

Apraxia is used here in relation to loss of controlled movement, described by the Gray Laboratory (2000) as the ‘inability to execute a skilled or learned motor act, not related to paralysis or lack of comprehension, caused by a cortical lesion’. However, the term apraxia is also understood to mean no movement, and is not considered clinically correct. Dyspraxia, meaning ‘difficulty with movement’ is the term preferred in neurology. Unfortunately dyspraxia has been described rather vaguely as ‘an impairment or immaturity of the organisation of movement, with associated problems of language perception and thought’ (The Dyspraxia Foundation, 2000) and as ‘impaired or painful function of any organ of the body’ (Gray Laboratory, 2000). The Foundation believes dyspraxia to be an immaturity of neurone development in the brain, particularly the myelination process, with no clinical neurological abnormality, although there are many soft neurological signs present. There are references in the Rett syndrome literature relating to both ‘apraxia’ (WHO, 1992, Lindberg, 1994) and ‘dyspraxia’ (Kerr, 1995a, Hunter, 1999) and in defining the necessary criteria, Hagberg (1993) used both terms (Appendix II). Divided professional opinion and lack of clarity of definition can only lead to more uncertainty for parents, who are likely to be confused by inconsistency both in the meaning of terminology and in the explanation of symptoms.
When the cells and their pathways in the cerebellum are damaged, preventing normal synapsis, movement becomes unco-ordinated and control is lost (Shepherd, 1980). This lack of co-ordination and poor balance is referred to as ataxia. Truncal ataxia is common in Rett syndrome and ambulant girls often walk with a stiff, broad-based gait (Hagberg, Aicardi, Dias and Ramos, 1983). Ataxia is characterised by great irregularity of functions or symptoms, or by a want of co-ordinating power in movements (Gray Laboratory, 2000). Bedford and McKinlay (1993) describe unsteadiness of movement, imperfect balance and intention tremor when discussing ataxia.

Development of tone characteristically occurs before the first 18 months, in a craniocaudal direction, to allow the infant to stand and walk (Witt-Engerström, 1993). She suggests that hypotonia is an expression of the suppression of 'multivariable control' of the regulation of posture, both at the level of the motor centres of the cerebral cortex and at the spinal level. Early low muscle tone can turn to rigidity and spasticity, with cold and swollen feet being an additional handicap (Lindberg, 1994). Consequently, even those girls who are able to walk frequently lose this ability as the condition progresses, and many become wheelchair bound.

As well as having difficulty in integrating their sensory impressions, girls with Rett syndrome may also have difficulty in co-ordinating their actions (Lindberg, 1994). Delayed responses are common, and deep concentration may be interpreted as lack of interest or comprehension (UKRSA, undated). There is a suggestion that some girls with Rett syndrome may actively want to carry out basic movements (such as feeding) but know and understand that they are not capable of doing so (Personal communication, parent of a girl with Rett syndrome, 1996). The implications for education and therapy are great, and if efforts are not to be seriously misdirected, the teacher and therapist must understand and address this feature.

1.5.1.2 Stereotypic hand movements

Some authors believe that the disappearance of purposeful, voluntary hand use constitutes the most important manifestations of Rett syndrome (Witt-Engerström, 1990). Kerr, Southall and Samuels (1992) suggest that stereotypic movements probably arise because the brain centres for control of fine intentional movement
fail to develop normally and some of the underlying rhythms which should be smoothly co-ordinated escape from control and become simple repeated patterns. The higher co-ordination that would, in time, develop into complex patterns of movement or of speech, is deficient in girls with Rett syndrome. Movement difficulties are exacerbated by agitation, which may be an unsuccessful effort by the brain's own feedback system to alert the defective control centres.

The repetitive movements over which a girl with Rett syndrome has little, if any control become increasingly troublesome during the regression period. Repeated, involuntary, movements of the tongue, hand, trunk and legs make it difficult for useful movement to be achieved. The hands seem to lose any voluntary control and begin to make repetitive clapping, wringing or squeezing movements, often with hands together at the level of chest or mouth. The movements are rhythmic, simple and slightly different for each hand and seem to prevent constructive use of the hands. Prolonged monitoring studies suggest that these are not under the control of the child and exacerbate automatically with evident agitation when she is alerted (Kerr, Southall, Amos, Cooper, Samuels, Mitchell and Stephenson, 1990).

Referring to the literature relating to the autistic spectrum disorder, Jordan (1999a) believes that 'It is not clear ... whether repetitive stereotyped behaviour reflects a truly cognitive difficulty in expressing spontaneous creative behaviour or whether it represents a form of 'displacement' behaviour in the face of overwhelming social, emotional or cognitive overload' (p.119). It is clear, that whatever the reason for stereotypic hand movements in Rett syndrome, they severely interfere with a girl's ability to participate in normal activities which would increase life's experiences, and as such cannot be ignored when educational needs are considered.

1.5.1.3 Mental disability

Rett syndrome is due to a flaw in development that interferes with the synaptic process. Although probably present from birth, problems only become evident within the first year of life, when development of the brain gradually brings the faulty but hitherto unused brain networks into operation. During the regression period mental confusion appears to increase, possibly due to the faulty internal communication systems of the brain increasingly dominating performance as the brain matures (Kerr, 1992b). It is axiomatic that many of the features of normal development are dependent upon and occur as a result of the development of the
brain (Holt, 1991). Uvebrant, Bjure, Sixt, Witt-Engerström, and Hagberg, 1993) found that in comparison to controls, children with Rett syndrome have a significant and early appearing hypoperfusion in the area of the midbrain and upper brainstem as well as the frontal lobes. The growth of the brain is reflected in the increase of head size with age and the clinician's most useful guide to this growth is measurement of the head circumference. One significant clinical feature of Rett syndrome already described is a deceleration of head and brain growth (Armstrong, 1992).

Reporting on 18 girls with Rett syndrome, Fontanesi and Haas (1988) found relative preservation of gross motor and daily living skills, at the developmental level corresponding to the age of onset of the condition, while other adaptive functions were more depressed, indicating that islands of motor and intellectual functions persist.

Girls with Rett syndrome have certain deficiencies in perception and sensory integration, leading to difficulties in interpreting and integrating sensory impressions from the outer world as well as from their own bodies. This becomes particularly noticeable during regression, but gradually the ability to interpret different signals improves, although their ability to perceive and integrate sensory stimuli will never be normal (Lindberg, 1994). In order to understand something they must experience it with their own bodies and through their own senses, and in order to remember, they must experience it many times. This is not a phenomenon unique to Rett syndrome. The same is true of very young children and children with severe learning difficulties and has implications for their treatment and education. It has never been suggested that infants should be educated in the same way as, for example, young adults. Similarly, children with profound disabilities may need different educational management to those of 'average' intelligence.

Whereas girls with Rett syndrome visibly lose their previous fine motor, verbal and social skills, they seem rather to come to a standstill in mental development. As they grow older, this aspect of the disability becomes more and more evident, they generally behave in a way normal for children of about 18 months of age (Lindberg, 1994) and remain totally dependent upon others for every aspect of their care.
1.5.1.4 Communication difficulties

Studies using magnetic resonance imaging have confirmed that the volume of the frontal and motor cortex is selectively reduced in Rett syndrome. These regions of the brain can be related to some of the major symptoms of Rett syndrome - difficulties with motor control, speech and emotional lability. The same damage to neural circuits at cortical and subcortical levels during early development which lead to immature motor patterns, also results in dysfunction of communication and breathing (Witt-Engerström, 1993).

The importance of oral motor skills also needs to be recognised, as they may be linked to feeding and communication abilities. Many girls display physiological problems relating to excessive tongue thrusting, chewing and swallowing (Van Acker, 1991). A study of 20 girls led Morton (in Kerr and Corbett, 1994) to report that the oral phase of swallowing was disturbed, with poor lip and palate seal, and jaw movement. Tongue behaviour was immature and dystonic, becoming rigid in older girls. Although early successful feeding is usual (Kerr, 1994b), at regression this skill commonly deteriorates and is seldom recovered, with failure to manage food in the mouth characteristic of older girls (Morton, 1995). Exacerbation of involuntary movements seems to contribute to this deterioration, and also to loss of speech.

Most girls have lost skills in speech before the age of three years (Hagberg, 1993) and never regain their previous range of speech after regression, although communication improves right into adult life (Kerr, 1995a). Occasionally a few useful words are retained or learned and it is a feature of the condition that these may be suddenly and aptly produced (Kerr, 1992b, Lindberg, 1994). Most girls do not have a useful vocabulary but do understand some language, at about the 12 month level (Kerr, 1988). Although girls with Rett syndrome do not have the ability to express their wish for contact in a conventional manner, pleasure and displeasure can be shown by emotional expressions, body language and facial expression (Lindberg, 1994).

1.5.2 Supportive criteria

All children diagnosed with Rett syndrome will suffer from the above disabilities. Many, however, will have some or all of the following symptoms. These are
described as supportive criteria (Hagberg, 1993) and may be used to strengthen the diagnosis of Rett syndrome if doubts remain.

1.5.2.1 Breathing difficulties

Research by Lugaresi, Cirignotta and Montagna (1985) indicates that breathing impairment in Rett syndrome represents a functional disturbance of the behavioural control of breathing, which occurs during active wakefulness.

The breathing irregularity that is seen in many girls with Rett syndrome usually begins at the end of the regression period, from 3 years onwards. It generally occurs when the girl is awake and, like other movements, is most noticeable when she is alert or agitated (Lewis and Wilson, 1996). Hyperventilation is common, with deep rapid breathing lasting for several seconds, invariably followed by apnoea, after which the breath may be ‘forcibly and noisily released’, accompanied by ‘short grunts or cries’ (Kerr, Southall and Samuels, 1992, p.2).

A direct consequence of hyperventilation is a fall in the level of carbon dioxide in the blood, as deep breathing expels an excess amount of carbon dioxide from the body. Carbon dioxide is a normal waste product of the body and while it is carried in the blood, it serves the purpose of maintaining the pH balance at an appropriate level for normal cell function. A fall in the blood level of carbon dioxide leads to a less than ideal environment for cellular activity (Kerr, Southall and Samuels, 1992). These authors have found no evidence that the brain is damaged by the events that may be associated with these breathing abnormalities. They regard this breathing irregularity as a disordered movement pattern, a direct consequence of the abnormal brain development responsible for Rett syndrome.

1.5.2.2 Epilepsy

Videos of girls later found to suffer from Rett syndrome taken at 2 - 3 months of age show repeated small twitching movements of the eyes and mouth, giving rise to the suggestion that the cortical abnormality is manifesting itself in barely perceptible seizure activity at this early stage, before Rett syndrome has been considered (Kerr, 1995a). However, routine electroencephalograms have been reported as normal before regression.

Seizures are sometimes difficult to differentiate from spells of breath holding, associated with abnormal breathing (Hagberg, 1993). Rarely, hyperventilation may
actually trigger an epileptic fit but most seizures seen in Rett syndrome do not appear to be directly associated with breathing abnormality. Many girls with Rett syndrome have brief interruptions of awareness which are not epileptic fits and some of these 'vacant spells' seem to be related to the breathing abnormality (Kerr, Southall, Samuels, 1992, Amos, Kerr and Cooper, 1992). The treatment for epilepsy is principally with anti-convulsant drugs, but the use of medication was not explored in this study.

1.5.2.3 Scoliosis

When the syndrome was initially described by Rett, there was no mention of spinal deformity as part of the abnormalities associated with the condition itself, but over the course of time it has become clear that spinal deformity is not uncommon (Webb, 1996). This may simply be because Rett's initial observations were made on young girls who had not yet developed scoliosis. Scoliosis in Rett syndrome is now well documented (Hagberg, Aicardi, Dias and Ramos, 1983, Hanks 1986, Van Acker, 1991) and at a recent World Congress, this condition was reported in 74% of females, the figure rising with age (Kerr, 1996b). Information from 282 Rett families on the RSAUK database indicates that scoliosis was often noted before Rett syndrome itself was diagnosed (Webb, 1996).

1.5.3 Exclusion criteria

The exclusion criteria refer mainly to medical disorders, and are used to eliminate misdiagnosis relating to other disabilities or diseases. These include microcephaly at birth, the existence of identifiable metabolic or other heredodegenerative disorder, acquired neurological disorder resulting from severe infections or head trauma and evidence of perinatally acquired brain damage (Hagberg, 1993).

1.5.4 Common symptoms

There are other difficulties common to many girls with Rett syndrome, but not included in either necessary or supportive criteria. These are discussed in the limited amount of educational research into Rett syndrome (Lindberg, 1994, Lewis and Wilson, 1998) and are considered of sufficient importance to be included here.

1.5.4.1 Emotional difficulties

During the regression stage, skills which have been mastered are lost, physical development deteriorates rapidly, and speech is no longer possible. Not
surprisingly, girls with Rett syndrome commonly seem withdrawn and frightened (Lindberg, 1994, Hagberg, 1993). The child is likely to react in various ways - screaming and crying, inappropriate laughing, self-biting or fighting are not unusual. Following regression, these symptoms stabilise and life becomes less confused. However, girls continue to have an incomplete and confused image of themselves as they grow, and they experience rapid mood changes. They also suffer fluctuations in behaviour, in capacity for contact and interaction with surroundings, and in performance as a whole (Lindberg, 1994).

1.5.4.2 Problems with nutrition

Kerr (1991) indicated that nutrition was a major area of difficulty for a minority of girls. Rice (undated) describes the typical child with Rett syndrome as extremely thin, with a good appetite but difficulty in gaining weight, to such an extent that it may become life threatening.

Motil, Schultz, Brown, Glaze and Percy (1994) and Schultz, Motil and Glaze (1994) have concluded that the difference in energy balance between girls with Rett syndrome and a healthy control group may not be due to excessive involuntary muscular movement, but to an inefficient energy intake. While they accept that the suggestion is unproven, they still consider it noteworthy that the difference in energy balance would account almost exactly for the difference in growth between the control and sample groups over a two year period.

1.5.5 Atypical Rett syndrome

Presence of any of the exclusion criteria negates a diagnosis of 'classic' or 'typical' Rett syndrome. It is, however, possible for a person to have 'Rett syndrome like' characteristics which do not fulfil the defined diagnostic criteria. In Britain, these girls are described as having 'atypical' Rett syndrome (Kerr and Corbett, 1994), while Hagberg and Witt-Engerström (1986) describe 'formes frustes' (Appendix III). Atypical children are not categorised in the necessary, supportive and exclusion diagnostic system. This could be likened to autism and autism-like conditions, moving away from types and categories towards continua. A major factor in the diagnosis of Rett syndrome depends more on understanding the sequence of events than on the appearance of any one sign or symptom at a precise age (Witt-Engerström, 1993).
1.5.6 Differential diagnosis

The presentation of Rett syndrome differs considerably depending upon the stage and age of observation of the child (Van Acker, 1991). Whereas a child of 4 - 5 years of age with classical Rett syndrome can be correctly diagnosed with relative ease, very young children may exhibit vague symptoms, making diagnosis less assured. Van Acker describes the late stage of adolescence as displaying a 'common, complex, multihandicapped picture of extreme severity with secondary contractures that resemble any number of disorders' (p.358) leading to frequent misdiagnosis.

ICD-10 (WHO, 1992, p.256) reports that: ‘Initially, Rett’s syndrome is differentiated primarily on the basis of lack of purposive hand movements, deceleration of head growth, ataxia, stereotypic “hand-washing” movements, and lack of proper chewing. The course of the disorder, in terms of progressive motor deterioration, confirms the diagnosis’. The entire disease process must be recognised and considered for the condition to be fully understood (Trevathan and Naidu, 1988) and subsequently addressed.

Because of difficulties in diagnosis, a range of descriptions is likely be used by professionals to describe a child within the autistic spectrum, including ‘autistic tendencies’ and ‘autistic features’ (Jordan, Jones and Murray, 1998) and these authors make it clear that - ‘There are no behaviours per se that by their presence or absence indicate autism; it is the overall pattern and underlying difficulties that define autism’ (p.14).

It has been seen that many of the individual characteristics of Rett syndrome are common to other disorders, and infantile autism is often suspected, particularly during the regression period when skills are lost in communication, sociability and hand use, but mobility remains (Witt-Engerström, 1993, Olsson and Rett, 1987). Initially the two conditions may overlap, with both having an early onset, absent or disordered speech and language, lack of social responsiveness and repetitive stereotypical mannerisms, but it is now generally accepted that the two disorders are distinguishable in a number of ways (Olsson and Rett, 1990, World Health Organisation, 1993, Naidu, Hyman, Harris, Negrin, Hosain, Yablonski and Moser, 1994, American Psychiatric Association, 1994).
Table 1 shows a comparison of the characteristics of Rett syndrome and childhood autism, taken from ICD-10 (World Health Organisation, 1993).

### Table 1  Comparison of characteristics of Rett syndrome and childhood autism

<table>
<thead>
<tr>
<th>Classification</th>
<th>Childhood autism F84.0</th>
<th>Rett syndrome F84.2</th>
</tr>
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<tbody>
<tr>
<td>A</td>
<td>Abnormal or impaired development is evident before the age of three years in at least one of the following areas:</td>
<td>A There is an apparently normal prenatal and perinatal period and apparently normal psychomotor development through the first 5 months and normal head circumference at birth.</td>
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<tr>
<td></td>
<td>1 receptive or expressive language as used in social communication;</td>
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<td></td>
<td>2 the development of selective social attachments or of reciprocal social interaction;</td>
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<td></td>
<td>3 functional or symbolic play.</td>
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<tr>
<td>B</td>
<td>A total of at least six symptoms from 1, 2 and 3 must be present, with at least two from 1 and at least one from each of 2 and 3:</td>
<td></td>
</tr>
<tr>
<td>1 Qualitative abnormalities in reciprocal social interaction</td>
<td>Manifest in at least two of the following areas:</td>
<td>B There is deceleration of head growth between 5 months and 4 years and loss of purposeful hand skills between 5 and 30 months of age that is associated with concurrent communication dysfunction and impaired social interactions and the appearance of poorly coordinated/unstable gait and/or trunk movements.</td>
</tr>
<tr>
<td></td>
<td>a. failure adequately to use eye-to-eye gaze, facial expression, body posture and gesture to regulate social interaction;</td>
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<td></td>
<td>b. failure to develop (in a manner appropriate to mental age and despite ample opportunities) peer relationships that involve a mutual sharing of interests, activities and emotions;</td>
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<td></td>
<td>c. lack of socio-emotional reciprocity as shown by an impaired or deviant response to other people's emotions; or lack of modulation of behaviour according to social context; or a weak integration of social, emotional and communicative behaviours;</td>
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<tr>
<td></td>
<td>d. lack of spontaneous seeking to share enjoyment, interests or achievements with other people (e.g. a lack of showing, bringing or pointing out to other people objects of interest to the individual).</td>
<td></td>
</tr>
<tr>
<td>2 Qualitative abnormalities in communication</td>
<td>Manifest in at least one of the following areas:</td>
<td>C There is severe impairment of expressive and receptive language, together with severe psychomotor retardation.</td>
</tr>
<tr>
<td></td>
<td>a. a delay in, or a total lack of, development of spoken language that is not accompanied by an attempt to compensate through the use of gesture or mime as an alternative mode of communication (often preceded by a lack of communicative babbling);</td>
<td></td>
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<td></td>
<td>b. relative failure to initiate or sustain conversational interchange (at whatever level of language skills is present), in which there is reciprocal responsiveness to the communications of the other person;</td>
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<td></td>
<td>c. stereotyped and repetitive use of language or idiosyncratic use of words or phrases;</td>
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<td></td>
<td>d. lack of varied spontaneous make-believe or (when young) social imitative play.</td>
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### Classification

<table>
<thead>
<tr>
<th><strong>Classification</strong></th>
<th><strong>Childhood autism F84.0</strong></th>
<th><strong>Rett syndrome F84.2</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>3 Restricted, repetitive and stereotyped patterns of behaviour, interests and activities</td>
<td>Manifest in at least one of the following areas: an encompassing preoccupation with one or more stereotyped and restricted patterns of interest that are abnormal in content or focus; or one or more interests that are abnormal in their intensity and circumscribed nature though not in their content or focus;</td>
<td>D There are stereotyped midline hand movements (such as handwringing or &quot;hand-washing&quot;) with an onset at or after the time when purposeful hand movements are lost.</td>
</tr>
<tr>
<td>a.</td>
<td>apparently compulsive adherence to specific, non-functional routine or rituals;</td>
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<tr>
<td>b.</td>
<td>stereotyped and repetitive motor mannerisms that involve either hand or finger flapping or twisting, or complex whole body movements;</td>
<td></td>
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<tr>
<td>c.</td>
<td>preoccupations with part-objects or non-functional elements of play materials (such as their odour, the feel of their surface, or the noise or vibration that they generate).</td>
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<td>d.</td>
<td>The clinical picture is not attributable to the other varieties of pervasive developmental disorder: specific developmental disorder of receptive language (F80.2) with secondary socioemotional problems; reactive attachment disorder (F94.1) or disinhibited attachment disorder (F94.2); mental retardation (F70-F72) with some associated emotional or behavioural disorder; schizophrenia (F20.-) of unusually early onset; and Rett syndrome (F82.4).</td>
<td></td>
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</tbody>
</table>

#### 1.5.7 Summary of symptoms

In summary, a young girl with Rett syndrome will have a motor disability, stereotypical hand movements, poor communication skills and severe learning difficulties. In addition, there may be evidence of breathing abnormalities, epilepsy and emotional difficulties. Add to this the lack of tangible evidence and it is not surprising that Rett syndrome is not easily recognised or diagnosed by medical experts. It is unlikely, then, that schools will be aware of the complexity of Rett syndrome, and consequently may not be able to plan appropriately for their children with Rett syndrome. There are many low incident complex disorders, and there are many implications for teachers and therapists. It would be difficult to disseminate knowledge of every condition, although the Green Paper (DfEE, 1997) outlines a commitment to training in Special Educational Needs and this is addressed in more detail in Chapter 7.

#### 1.5.8 Response to diagnosis

After diagnosis of Rett syndrome, parents show common reactions in a series of stages (Goodship, 1987). Anxiety, shock and disbelief are followed by numbness, sadness and grief. 'Why' questions follow and feelings of depression and
helplessness may appear at any time. The need for a tentative Rett syndrome diagnosis for use in daily practical life, for understanding and management has been emphasised by parents and others involved (Hagberg, 1995b). The need for a diagnosis in all autistic spectrum disorders is discussed further in Section 5.3.

Frequently, the confirmation of a diagnosis is the step which enables parents to look beyond the present (Hunter, 1987). How to cope in the family environment, how to plan for the future, how to arrange a school education, are all issues which are hard to address when the nature and cause of the symptoms are not understood. 'For parents, a diagnostic label can give then access to a voluntary organisation which can provide not only further information, but also a source of emotional, psychological and practical support both from helplines and from meeting other parents' (Gascoigne, 1995, p. 17).

As well as support from parental sources, it is likely that parents will need the support of a number of professionals to help them, both through this initial period, and as their child matures and grows. As well as providing support, this can cause a number of problems. Parents may not be aware of what help is available, how to access it, or how to deal with the situation, and there may be resentment at the intrusion of so many people into an essentially private situation. It will be seen that the form of the early professional / parent interaction may determine the later character of the relationship and affect the role of parents and their perceptions of any management of their child.

1.6 Parental involvement

There has been an increasing emphasis by the Government on the importance of parental involvement in the education of their children, no more so than in the area of special needs (DES, 1978, DFE, 1994). The Green Paper (DfEE, 1997) highlights the importance of parents' roles in helping children learn, whilst accepting that 'for many parents, learning of their child's problems will be a devastating blow' (p.25) and that varying degrees of support will be needed. The paper accepts that therapies will form an important part of the education of children with special needs. Given the wide range of difficulties encountered by children with special needs, both within one syndrome and in the wider sphere of disability, is it unlikely that there will be one 'best' therapy. Individual symptoms and circumstances will need to be considered and taken into account, and it seems obvious that some of the
information can only be acquired from parents. In an age when individual rights are at the forefront of national processes (DES, 1978, DFE, 1994, DfEE, 1997), the views of parents should be sought frequently in relation to children’s education.

In a single generation, relationships between parents and teachers have changed significantly (Wolfendale, 1984, Topping, 1986), and studies such as this are necessary from a rights perspective and valuable from the child’s perspective. The parent of one child with Rett syndrome summed up the feelings of many: ‘I feel I know what is good for my child and what will help her’ (Personal communication, parent of girl with Rett syndrome, 1997).

Given the likely potential variation in experiences and feelings, it is necessary to listen to individuals, and to distil social opinions and trends from larger groups. Whilst individuals are important, it simply might not be possible to accommodate the extremes of individual viewpoints, whereas response to the aggregate view might be more achievable. Once the listening is complete (and it maybe never will be) some kind of action or response should be initiated. To do nothing would be to let down the parent community, and discourage further consultation. ‘The essence of good consultation is to ask people for information or to express their views, to pay close attention to what they say, and to act on it where possible’ (Ball, 1997, p.5).

In appraising research literature relating to the benefits of parental participation in children’s development and education, Wolfendale (1984) found there to have been a radical departure from the traditional approaches whereby professionals and parents kept their distance. Topping (1986) found that parental involvement in schools only began to be documented with some degree of certainty this century. He cites Wall (1947) who found some benefits in such co-operation, and also highlighted the fears of some headteachers that it might lead to criticism of the teacher in the home. It may be that the ‘aloofness’ so prevalent in the past was thought to preserve respect, with approachability bringing a perceived loss of respect. In reality, approachability followed by positive response might enhance respect felt by parents for teachers.

1.6.1 Parent / professional relationships

Cunningham and Davis (1985) demonstrate the use of Personal Construct Theory as a framework to enable professionals to best use their knowledge and skills in
partnership with parents. These authors talk strongly about the rights of parents, and the need for a trusting and co-operative relationship between parents and professionals. They examine, discuss and contrast three models of relationships that they believe to be evident in current practices, the expert, transplant, and consumer models. Very much simplified, in the expert model, professionals take control, maintain a ‘distance’, and instruct parents within their protected position. In the transplant model, the parent becomes a resource, and receives new skills from the professional in an environment of co-operation. In the consumer model, the parent is the consumer, and the professional the supplier. The parent is the decision-maker, and the professional acts as consultant.

The expert view could not be described as a partnership (Cunningham and Davis, 1985), and despite its long tradition, even though expert knowledge is indeed used for the benefit of the client, in this model the benefits of a holistic approach can be overlooked in favour of the treatment of specific symptoms. Most parents are not and never will be ‘experts’ in the same way as professionals, and in the trauma of disability will need some form of partnership. Parents might be vulnerable, and experience guilt or a sense of failure, but on the other hand, they may be convinced of their own potential for dealing with their child’s disability to the extent that they challenge the professionals in their own sphere. Parents of children with special needs will meet many professionals during their child’s education, and probably encounter views from each model. This may, in turn, colour their outlook on the value of the service being offered. Professionals should never lose sight of their ultimate purpose, and effective parental involvement should be part of the plan of action. ‘Ideally, the process of negotiation should lead to mutually agreed solutions in which the parents have executive control over major decisions. This should reduce the likelihood of failure due to incompatibility between family resources and professional advice’ (Cunningham and Davis, 1985).

What is certain is that during the life of a child with special needs, parents will have contact with many professionals, some more competent than others, but all with a degree of influence on the shaping of their children’s futures.

There is a natural variation among parents, but whatever the particular age, race, creed, experience, demeanour and circumstance might be, each parent is likely to be aware of some fact or feature pertinent to the growth and development of their
child, and this information will need to be conveyed to others. The skilled professional will need competence in their own professional discipline, and they must have the ability to communicate this competence (Cunningham and Davis, 1985). An understanding of the likely reactions of parents to having a child with special needs will help professionals judge how best to formulate the parent / professional relationship.

Parents will already have seen their children develop since birth, while the specialist will see them periodically, probably only after a critical event has flagged a problem. Parents see them daily, so developmental flaws might be difficult to notice. In any case, perceptions of development are likely to differ between parents and between professionals, although there are patterns or sequences of development that are accepted as being characteristic for all children (Kramer and Hinojosa, 1993).

1.7 The education of children with Rett syndrome

The Education (Handicapped Children) Act (DES, 1970) established the right of children with severe learning difficulties to enter the education system. The publication of the Warnock Report followed mounting pressure to recognise the right for the educational needs of all pupils to be met (DES, 1978) and signalled the beginning of a new recognition of the entitlement of minority groups of children.

Appreciation and meaning of the word 'education' have changed over time. Formerly given a simple interpretation, to educate, meaning to bring up or to instruct (Reid 1861), or to cultivate or train (Skeat 1888), there are wider implications in the meaning today and Summer (Ed.) (1992) defines education as the process by which a person's mind and character are developed through teaching or through formal instruction at a school or college. Education has also been described as the acquisition of knowledge and skill (Hanks, 1971). Just as there are different definitions of education, we all have different experiences of education and the education needed by one child may not be appropriate for another.

The individual needs of a girl with Rett syndrome will be assessed by the Local Education Authority, and a statement of Special Educational Needs issued. The process should, in theory, reflect the views of all who have been involved in the process of assessment; pre-school teacher, paediatrician, educational psychologist
and other specialists. The specific objectives contained in the statement, combined with each girl's right to a broad and balanced curriculum will form the basis of the work undertaken by the class teacher.

Jordan (1999a) suggests that it is necessary to look beyond behaviour to psychological functioning in order to teach a child with autism effectively and this is equally applicable to Rett syndrome. On a practical level, not only does the teacher have to consider how to address the essential criteria of motor disability, poor communication skills, stereotypical hand movements and learning difficulties which are certain to be present, but there is also the possibility of added complications such as hyperventilation and epilepsy. It is likely that the parents are more concerned with the physical condition and general good health of the child than the broad and balanced curriculum demanded by unnamed persons far removed from the practical difficulties caused by this distressing syndrome. For many parents, an appropriate curriculum is of far more significance that breadth and balance. It has to be hoped that the statement of Special Educational Needs will reflect health concerns, and that the annual review of the statement will ensure that both individual educational needs and health needs are reflected as appropriate. Whilst not suggesting that girls with Rett syndrome do not have a right to schooling, the value of 'education' as recognised in the National Curriculum has to be queried. There is much written in education regarding 'meeting a child's needs' and how that can be achieved, but possibly less regarding the difficult task of discovering what those needs really are, although legislation and official documents in recent years have emphasised the need for assessment (DES, 1978, DFE, 1994, DfEE, 1997).

Fagg, Aherne, Skelton and Thornber (1990) believe that the curriculum consists of all deliberate experiences provided by a school in which an individual participates, and will consist of the National Curriculum subjects, and all other aspects considered vital by the school. A broad curriculum will ensure opportunities for areas of work pertinent to the development of the individual that may not be included in the National Curriculum. So, whilst accepting the value of the National Curriculum, it is important not to forget the many therapies which offer girls with Rett syndrome the only hope of even approaching a degree of functionality in the essential and fundamental matter of day to day living. Special programmes and 'hidden curriculum' are needed if individual special needs are to be met, and in a sense programmed forms of therapy could be the only real education the girls need.
For example, in a broader, less specific sense, Longhorn (1988) believes the sensory curriculum can enhance the National Curriculum in order to provide the necessary learning experiences for children with special needs.

Jordan and Powell (1997) do not advocate any single teaching approach, but suggest that for some children there must be structure in order to minimise stress and to promote learning. This is of particular relevance for any disorder within the autistic spectrum. It has been seen that girls with Rett syndrome have difficulty in integrating their sensory impressions and that during the regression period mental confusion increases (Lindberg, 1994, Kerr, 1992b) and so any structure which can be given will be of benefit. It has been suggested that familiar routines based on everyday activities should be given consistency of time, person, place and physical and verbal cues, leading to an understanding of daily activities and routine, including an increased understanding of the immediate environment (McInnes and Treffry, 1993, Van Dijk, 1991).

Programmes such as TEACCH (Treatment and Education of Autistic and related Communication handicapped CHildren) provide necessary structure, and ‘clarify the dimensions that need to be considered when teaching communication skills and stress the need to teach one dimension only at a time’ (Jordan and Powell, 1995, p.66). By focusing on the strengths, interests and emerging skills of the individual child (Jordan, Jones and Murray, 1998), this approach could help to reduce the confusion frequently felt by children within the autistic spectrum. There may need to be some consideration of stereotypic hand movements when planning the programme for children with Rett syndrome, and this would be best undertaken collaboratively by teacher, therapist and parent.

The TEACCH approach increases the potential of the parent to play a role in the educational programming of their child (Gallagher, 1988) and so is a good model of parent / professional collaboration. Other approaches to educational intervention that have been used with children within the autistic spectrum include interactive and communication approaches, as well as those based on the principles of behaviour management (Jordan et al., 1998). In a study into educational provision for children with autism, Jordan and Jones (1997) found it encouraging that in most school and units, single approaches were not being adopted en masse, but that aspects of different approaches were incorporated into a broad curriculum. For this
to be successful, there are implications for staff expertise and knowledge, and subsequent implications for training.

The question of what must be included in a broad and balanced curriculum for all pupils with Special Educational Needs was an issue raised at National Curriculum Council seminars. Access to the National Curriculum is part of a total curriculum for these pupils, but other elements across the curriculum remain essential. In the seminars teachers stressed the importance of meeting individual needs through such means as programmes of therapy, including speech therapy, physiotherapy and the art therapies, particularly for those pupils with profound and multiple learning difficulties (National Curriculum Council, 1993).

1.7.1 Therapeutic intervention

There has been a variety of attempts at therapeutic intervention for Rett syndrome (Kerr, 1988). Therapy is the treatment of a disorder and in as much as medical knowledge allows, intervention programmes either target physical problems such as scoliosis and mobility caused by loss of cortico-spinal influence or focus on the behaviours and skills of children with disabilities. Intervention for physical symptoms include physiotherapy, speech therapy, hydrotherapy, hippotherapy and occupational therapy, with a variety of approaches used to adapt behavioural characteristics and enhance daily living skills. The majority of intervention programmes targeting behaviours and skills of girls with Rett syndrome have concentrated on increasing functional hand use by decreasing stereotypical hand movements (Mount, 1998).

If, as has been suggested, infant skills are not truly lost in girls with Rett syndrome, then surely it requires very specific methods of searching out and re-vitalising those abilities. ‘The existence of these skills provides an opportunity for the therapist and a challenge for the neurologist’ (Kerr, 1994d, p.1). Therapy can be directed right to the heart of specific areas of the disability: targets of therapy might be physical and emotional as well as addressing areas of communication and learning difficulties. The intention of a therapy may be to ensure that a girl has access to the National Curriculum, and as such must be considered a necessary and valid precursor to formal learning.
The standard therapies available in this country, the theories behind these therapies, and the implications for the treatment and education of children with Rett syndrome, are described in Chapter 2.

1.7.2 Education and therapy

Within the school setting, the concepts of education and therapy taken in the commonly accepted sense of the words initially appear uncomplicated, but their interactions are not entirely free from disharmony, although the two co-exist side by side in schools for children with profound and multiple learning difficulties. The two words have evolved separately; education has roots in the Latin ‘educatus’ - to bring out, while therapy derives from the Greek ‘therapeia’ - service done to the sick. Whilst education is concerned with the acquisition of skills and knowledge, therapy has an emphasis on curative processes. By implication, therapy is directed towards putting right that which is wrong, whereas education is directed more to developing already present but immature abilities and characteristics. Differences in brain function, manifest in physical symptoms and behaviours, lead traditional therapy towards specific methods, often with a very individual approach developed around an understanding of individual needs, an understanding which is less developed in the group approach of education where principles of equality and rights tend to be emphasised.

Within this tension, the common ground of education and therapy is progress from a less desirable to a more desirable state, even though in extreme cases progress might equate to a kind of stasis. Ware and Healy (1994) offer a philosophical summary of what is desirable and what progress might mean in terms of how society sees fitness for life, and they point out that the views of individuals and society about what represents quality of life, and thus progress towards it, may change radically with changing circumstances. They believe that the definition of objectives and the measurement of movement towards them are ‘issues central to the whole educational enterprise’ (p.1) and accept that apparent lack of progress is a major concern for teachers working with children with profound and multiple learning difficulties.

The National Curriculum states that a belief in education as a route to the ‘spiritual, moral, social, cultural, physical and mental development, and thus the wellbeing of the individual’ (DfEE and QCA, 1999, p.10) is essential. So for a child with
profound and multiple learning difficulties, it is no easy task to separate education from therapy. The two should go hand in hand, within a mutually supportive relationship, but there are occasions when the relationship is modified to some extent by separate agendas. The teachers' role historically, has been to teach, whereas the role of the therapist has been to give or organise therapy. From a position of distance, the roles have converged to some extent, but the professionals still bear different names, are trained differently, and have different qualifications. They are different people, with different priorities, but they work with the same children and to some extent share common aims.

1.8 Summary

Rett syndrome is far more common that was at first thought, and is now believed to be the commonest cause of profound mental and physical disability in girls and women, with sufferers becoming dependant upon others for all aspects of their daily care (UKRSA, 1996a). Rett believed that the condition has existed for many years and that if the girls were born years ago, they would have died at a very young age (Rett, 1985b).

As the condition has become recognised world wide, with increased knowledge and more accurate diagnosis, it has become clear that it is not the steadily deteriorating disorder that was assumed at first. There are, however, a number of sudden, unexplained deaths, approximately 1% of the girls recorded on the UKRSA database (Kerr, 1996a). This is fewer than might be expected among profoundly disabled people, but more than for normal healthy girls. Many women enjoy sound health and it is not unusual for communication and movement skills to improve in later years (Kerr, 1992b).

In Rett Syndrome we must plan for prolonged survival with continuing positive learning and enjoyment of skills in spite of profound disabilities and physical therapies are of primary importance both in order to develop full potential and because lack of stimulation and use of the disabled mind and body plays a major role in producing secondary disability. (Kerr, 1988, p.2)

The preservation of acquired skills and the continued improvement of re-learned skills are paramount. It is unlikely that even the most dedicated teacher will be able to achieve these objectives by following the National Curriculum alone. If girls with Rett syndrome are to genuinely have equal opportunities with their able bodied
peers, then every effort must be made, by all concerned, to ensure that their education is personalised to meet their individual needs. If this does not meet the accepted model of education, then another, less conventional route must be found.

The effects of Rett syndrome have been described, but how they may best be addressed in schools is not clear. In an environment of increasing parental involvement, the role of the school in areas such as therapy provision and support is growing. Parents are now being seen as important partners in education (DFE, 1994, DfEE, 1997, Wolfendale, 1992, Wolfendale, 1997) and it is to parents that schools must look for feedback regarding the policies and practices which are intended to meet the needs of the children. Researching parents' perceptions is just one way of gathering opinions, not only of the effect of their daughters' conditions, but of how valuable the many therapies are perceived to be in preserving and nurturing essential life skills.

Hunter (1987) claims that all parents of daughters with Rett syndrome want the opportunity to talk about their child and to be heard and this study has offered a means, albeit limited, for them to do so. Of course, relationships may not be that straightforward and there are still barriers to effective communication between researcher and the researched. Not least among them is the sensitive and very painful nature of the problem; there may well also be considerable inertia to overcome, with less motivated parents feeling reluctant to engage in dialogue because they have not learned its value. For parents of girls with Rett syndrome, it is hoped that this study will move the wider process onwards, and at the same time gather evidence of how parents perceive the various therapies to work. It should also prompt debate within the education system and generate some suggestions for how the special needs of children with Rett syndrome and other low incidence disabilities might be determined and addressed.

For the professional, it is vital to recognise what is best for the child in terms of learning, experience, and therapies. Just as Rett syndrome may be difficult to diagnose, the complexities and variations of the condition may make it difficult to educate and treat the girl with Rett syndrome even in the context of the special school, even though therapists and teachers have parallel or common aims.

The increasing emphasis on parent/school partnerships may provide a vehicle for channelling parental input into school policy development. Ideally, parents of girls
Chapter 2 Therapies – a review of the literature

2.1 Therapies

In spite of the extensive medical research that has been carried out in recent years, there is currently no cure for Rett syndrome. There is neither standard provision nor uniform delivery of therapies to children in British schools, and it is accepted that for some conditions the prognosis is not hopeful.

...there are conditions within the autistic spectrum, such as Rett's syndrome, where there is physiological and neurological deterioration over time which, sadly, often results in loss of function. In such cases, teaching has to be directed at maintaining current levels of functioning wherever possible, and finding new ways to enhance the quality of life with a deteriorating range of skills and abilities.

(Jordan and Powell, 1995, p. 156)

There is a vast array of therapeutic intervention methods used to treat the wider variety of medical conditions existing throughout the world, from the ambitious but experimental dolphin therapy under development in North America to the technical application of hyperbaric oxygen therapy (James, 1995). All share common aims in seeking to improve the wellbeing of the patient although the routes are not always specified and the results sometimes unclear.

Therapies chosen in this study for examination include music therapy, speech therapy, hydrotherapy, physiotherapy, and occupational therapy. Hippotherapy, aromatherapy and cranial osteopathy are mentioned, but not discussed in depth. The basis for this choice derives from an examination of the effects of Rett syndrome, discussion with professionals and parents, and from the results of the survey, which will be discussed later. All of these therapies are manageable to a greater or lesser degree in a school setting, and while they do not themselves constitute 'education' in the traditional meaning of the word, they are all ways of producing reactions (perhaps only short term) in girls with Rett syndrome that will allow, through the conception and nurturing of dormant or absent skills, an indirect access to forms of education which would otherwise be inaccessible.

2.2 Physiotherapy

The most common visible manifestation of disability other than aesthetic appearance is physical dysfunction. Intellectual impairment may not be immediately apparent to the casual observer, but few can fail to notice the outward...
signs of conditions such as cerebral palsy, spina bifida, thalidomide deformity, and, indeed, established Rett syndrome. The paediatric physiotherapist is concerned with the assessment, treatment and management of children who have a general developmental delay, disorder of movement, disability or illness which may be improved, controlled or alleviated by physiotherapeutic skills and / or the use of specialised equipment. They are likely to treat children with physical problems caused by neuromuscular, musculo-skeletal and cardiovascular / respiratory conditions (Association of Paediatric Chartered Physiotherapists, undated).

Levitt (1982) suggests that when the neurological mechanisms of posture, balance and movement are disorganised, the muscles that are activated for maintaining these mechanisms become unco-ordinated and weak. The physiotherapist should therefore aim treatment primarily at the neurological mechanisms in the central nervous system which activate and control these muscles. It is true that physiotherapy programmes still aim to correct, as far as possible, any deformities or dysfunction resulting from the physical condition of children (Ouvry, 1987), but in addition, they seek to prevent the development of any additional deformities or loss of function which may seriously impede the ability to participate in functional or educational activities.

In referring specifically to girls with Rett syndrome, Hanks (1990) suggested that any physiotherapy programme must be tailored to the individual child, based on a careful analysis of the factors that are interfering with functional movements or causing deformities. She considers that essential areas for treatment goals will include the development or maintenance of ambulation and of transitional skills, the prevention or reduction of deformities, the alleviation of discomfort and irritability and an improvement in independence. It is the opinion of Kerr, Southall and Samuels (1992) that the physiotherapist should strive to minimise joint deformities during childhood, as the fragile skills of a child with Rett syndrome can be used in later life, when control over movement tends to improve. This view is reiterated by Hanks (1990) who believes that the development of a therapy programme for a girl with Rett syndrome must evaluate her present state, and also anticipate future problems from what is already known about the usual course of the disease.
2.2.1 Developmental issues

Because abnormalities in a child's physical condition are not necessarily apparent at birth, a child's health and development are checked from a very young age against accepted criteria in order to obtain warning of developmental delays or more serious motor and neurological incapacity. Routine developmental assessment takes place at regular intervals between six weeks and five years. Children vary greatly in their development, therefore the 'milestones' which should occur at or near to these stages can only be timed approximately, and it is usually a cluster of achievement delays or abnormalities that indicate problems, rather than the absence or delayed occurrence of a single milestone (Levitt, 1984).

Witt-Engerström (1987) in fact attempted to systemise early development milestones and key age data for girls with Rett syndrome, and to identify existing but unrecognised deviations from birth. At 6 months, no predictive developmental features could have been revealed by routine screening, although by 9 months suspicion of developmental pathology may have arisen as a result of gross motor delay. Between 9 and 14 months there are clear abnormal neurodevelopmental signs and clinical symptoms, and the signs leading to the earliest referrals of girls who were subsequently confirmed to have Rett syndrome were failures to attain gross motor milestones.

2.2.2 Theories of physiotherapy

Many philosophies can be adopted for use in the treatment of neurological disorders of childhood (Eckersley and King, 1993). The more common approaches, and particularly those that may be used successfully in the treatment of children with neurodevelopmental disorders, are discussed in the following sections.

2.2.2.1 Neurodevelopmental treatment

The basic theory behind the Bobath neurodevelopmental treatment is that inhibition of abnormal reflex activity leads to facilitation of normal automatic movement (Bobath, 1980). The Bobath concept outlined a number of basic principles which should be incorporated into any treatment approach - patterns of movement, use of handling and prerequisites for movement (Eckersley and King, 1993).

Bobath programmes are systematically tailored to meet the personal requirements of individual children and much traditional physiotherapy in schools is based on this
neurodevelopmental treatment. Children are withdrawn from the class lesson and work in isolation with the physiotherapist. Classwork missed can be experience lost forever, or more pertinently, never gained. However, if without the Bobath programme the child can never attain a state in which educational achievement can actually be reached, there may be little point in attempting to teach that particular skill. There is also a risk that if the treatment is administered away from the classroom, the teacher is unable to continue the work of the physiotherapist, and opportunities for consolidating physical improvement may be lost.

Today, paediatric physiotherapists are very aware of the educational needs of children, and accept that access to education is important for all children and that there are times in a child's life when education is of paramount importance (Association of Paediatric Chartered Physiotherapists, undated). There may be occasions when physiotherapy treatment in education time is not appropriate and a decision involving parents will need to be made about the most suitable time and place for treatment. Equally, there may be times when physical needs are paramount, and educational needs must be subordinate.

2.2.2.2 Muscle education and braces

Specific treatment techniques, including muscle education and the use of braces and callipers, were developed by Phelps, an American orthopaedic surgeon. He advocated the use of massage for hypotonia, passive movements for maintaining soft tissue length and for demonstrating the movement required, active assisted movements and conditioned motion and synergistic motion involving resistance to one muscle group in order to facilitate contraction of an inactive muscle group in the same synergy (Levitt, 1982). This is a familiar concept in proprioceptive neuromuscular facilitation.

Braces or callipers are still in use in many schools today and in some cases they are considered a necessary adjunct to physiotherapy treatment, although there are views advocating and condemning their use (Naganuma and Billingsley, 1988, Tuten and Miedaner, 1989, Aron, 1990). It will be seen that some parents have strong views concerning the use of splints and braces for children with Rett syndrome, and this is discussed in more detail in Chapter 6.
2.2.2.3 **Proprioceptive neuromuscular facilitation (PNF)**

This technique of facilitation was devised in America 50 years ago, and the majority of British trained physiotherapists will be familiar with at least the basic principles of the technique (Eckersley and King, 1993). Proprioception is the understanding of the body’s position in space (consciously and unconsciously) based on feedback from joint, muscle and skin receptors. Proprioception contributes to balance and equilibrium, muscle tone and cocontraction. Proprioceptors such as muscle spindles work in conjunction with the vestibular system to give a sense of balance and position in space. All muscles and joints are involved in this process; however, the neck joints and proximal limb joints, such as shoulders and hips, are of primary importance and give the most feedback to the central nervous system (Kramer and Hinojosa, 1993). PNF is a treatment that is particularly useful for those who are unable to co-operate as techniques to stimulate deep muscles can be done actively by the therapist with only passive co-operation, making it appropriate for children with profound and multiple learning difficulties.

2.2.2.4 **Sensory stimulation**

The approach advocated by Rood (Cash, 1974, Burr, 1984, Stanton, 1992) suggested activating muscles through sensations of heat, cold and brushing, and is based on the theory that tactile and vestibular mechanisms play an important part in normal sensori-motor development. Ayres' sensory integrative therapy was also based on this theory. The treatment method is mainly based on constant motion stimulation such as swinging in a hammock or on a swing or whirling in a swivel chair (Stanton, 1992).

2.2.2.5 **Progressive pattern movements**

A great deal of physiotherapy is based on Temple Fay's developmental pattern of movement, when the child is taught to move according to evolutionary development (Eckersley and King, 1993). The theory behind this work is that the child with a motor disability has the same reflex movements that can be provoked in the non-disabled new-born child. The treatment programme elicits patterns of reflex motion by manual pressure on 'trigger zones' which are then 'imprinted' in the central nervous system and stored within the brain, thus allowing normal motor patterns to occur (Stanton, 1992).
2.2.2.6 Patterning

In the late 1940s, there was a growing body of evidence that sensory enrichment as well as sensory deprivation can alter the structure of the brain. This inspired Glenn Doman (a physiotherapist) and Carl Delacato (an educator) to develop a technique which aimed to treat the brain by altering the neurological system within the brain (Stanton, 1992). The Doman-Delacato principle is that it is possible to treat the brain itself to enable function to be restored naturally through the restoration of the brain's capacity to dictate appropriate function. The theory is that the function of the child's damaged central neurological organisation can be improved by means of an intensive programme of repetitive movements, enabling undamaged parts of the brain to take over the damaged area. The basic principle behind patterning is that mobility is only attained through frequent, intense and repetitive movement. These movements are patterned in accordance with the sequences of movements that 'normal' children go through (Bowley and Gardner, 1980).

The Doman-Delacato system is not a treatment that sits well with the educational system in this country. It is recommended by Doman-Delacato supporters that at least two years are spent in patterning before decisions are made about its effectiveness (Stanton, 1992). However, this 'trial' period is a long time, and if no result is detectable, there must be a suggestion that the time could have been used more productively in other areas. There has to be a certain risk commensuracy, and when chances are taken with children's development, unsuccessful cases are bound to be emotionally debilitating, and doubts may remain as to whether proven improvement can be attributed with certainty to the treatment.

One mother of a child with Rett syndrome experienced the treatment from the Institute for the Achievement of Human Potential in Philadelphia for two years.

The worst part of the treatment is that all the burden, both credit and blame, is placed on the parents in regards to the implementation of what they tell you to do. If you are honest and tell whatever did or didn't work, and if they think it should have worked and it didn't, it is 'all your fault' if your child doesn't see dramatic results, and you end up with a heavy burden of guilt.

(RettNet Internet Newsgroup, 1997)

At the University of Western Australia, Leonard reports that she has encountered a number of children who have been on the programme; some have made gains, but
they might equally well have done so using more conventional programmes (RettNet Internet Newsgroup, 1997). The disadvantage of the programme is the financial, physical, and emotional stress put on parents when such a programme permeates their whole existence to the detriment of a family life.

2.2.2.7 Conductive education

The multi-disciplinary approach began to be widely accepted with the work of Professor Pető, neurologist and educationalist, who was amongst the first to understand the need for a unity of approach to the treatment, education and management of the disabled child. His work began in 1951 and was continued by his students and colleagues after his death in 1967 (Cotton, 1998).

Conductive education differs from traditional methods of physiotherapy, being essentially an educational approach for motor disorders based on learning, rather than a therapy or a treatment (Grant, 1995). This method breaks away from the traditional treatment carried out by several separate therapists and teachers, and advocates instead a unified approach, which aims to deal with all aspects of the disabled person's development. This includes not only motor development, but perception, attention, language, and early educational skills, in accordance with the carefully devised strict programme of systematic sequential training (Hári and Ákos, 1988). An important principle of Conductive Education is the intensive work towards orthofunction, with the ultimate aim of enabling the child to attend a mainstream school.

Hári and Ákos (1988) believe that it is impossible to isolate principle features of conductive education, as the details can never be separated, and that if one area is neglected, the entirety suffers. On the other hand, Lewis and Wilson (1998) believe that aspects of conductive education have been found to be helpful for girls with Rett Syndrome to overcome motor problems, with rhythmical intention in particular being a powerful motivator. ‘The principles of rhythmic intention, in which actions are practised within a repetitive routine with clear cues and signals, would appear to offer a way forward for some girls with Rett Syndrome in relation to their profound learning difficulties, apraxia and delayed response …’ (p. 13).

The use of rhythmic intention encourages children to verbalise, helps to regulate the timing of motor actions and can assist tempo and the general cohesion of the group. It is thought that linking speech with motor actions helps to organise the
brain to signal motor performance with an increased level of control (Cottam and Sutton, 1986). ‘At a time when the cerebral organisation of an activity leading to a particular goal progresses relatively slowly, counting has a co-ordinating role in filling out the rather lengthy period between verbally intending a task and accomplishing it’ (Hári and Ákos, 1988, p.210).

A major problem with using conductive education within the current educational system of this country is the use of time. Schools for pupils with a physical disability have always had to balance the demands of an academic programme and the essential need for a range of treatment and therapy. In recent years it has been found that the time essential for conductive / movement education leaves insufficient time to provide a broad and balanced curriculum, although Bakehouse (1995) believes it is possible to meet the needs of the physically disabled without compromising the essential element of quality. However, it has already been seen (Section 2.2.2.1) that there may be times when educationalists and therapists need to prioritise the needs of children.

A report commissioned by the Government calls into question the value of conductive education, suggesting that ‘conventional’ treatments were equally successful (Bairstow, Cochrane and Hur, 1993, Rogers, 1993). Conductive education is seen by some as a holistic approach to the disabled child in which a motor disorder is a learning difficulty to overcome rather than a condition to be treated (Hári and Ákos, 1988) although Bairstow, Cochrane and Hur, (1993) are very critical of some assertions made by the Pető Institute, such as dysfunction being a personality problem. These authors also identify unchallenged, untested and conflicting statements, poorly explained key features, and incorrect use of terminology.

It is understandable that parents want to do whatever they consider best for their child, and conductive education is still used in some special schools today. However, there is an inter-disciplinary approach being increasingly employed in this country to teach children with a motor disorder, which is derived from conductive education and influenced by British Primary practice, and this is considered in the following section.
2.2.2.8 Movement education

Movement education aims to stimulate rich and varied movement experiences that enhance the development of psychomotor, cognitive and affective abilities. Central to this work is a learning model that educates children towards orthofunction, thus enhancing their integration into society. The approach is holistic, and child-centred activities and medical therapies are skilfully integrated into a broad and balanced curriculum designed to meet the needs of children with motor disabilities (Fairfields School, 1996).

The content of movement education sessions includes opportunities to develop physical, musical, language and play skills, as well as mathematical or scientific concepts, social interaction and creativity. A thematic approach to the National Curriculum, and movement education sessions incorporate the same themes, reinforcing and developing work undertaken in classrooms. Teams of teachers, nursery nurses, physiotherapists, speech therapists, occupational therapists and their assistants work together, sharing expertise. Regular multi-disciplinary planning meetings ensure high standards in both movement education and National Curriculum are met by evaluating current work and by preparing new programmes for the children. This approach is less rigid and more easily implemented than conductive education, and seems to offer a combination of treatment and education that can be adapted to meet the needs of the individual child.

2.2.3 Treatment principles

All children who are referred with a physical difficulty are assessed to identify problems that may indicate that physiotherapy is appropriate. An initial assessment followed by continuous holistic assessment of the child is fundamental to a successful treatment outcome. Assessment may be carried out by a mixture of detached observation in a free play situation, parent questioning, physical examination and using a series of specific tests (Association of Paediatric Chartered Physiotherapists, undated).

Eckersley and King (1993) have found no empirical evidence to suggest whether an integrative and eclectic approach or a specific concept approach is the most effective when working with children. It is, however, the common ground between the different systems which forms the principles of treatment (Levitt, 1982). The following general principles of treatment have been commonly accepted by various
schools of thought: team work, early treatment, repetition of a motor activity, whether it is neuromuscular techniques in treatment sessions or in the motor activity during all day management, sensori-motor experience and motivation of the child. Common factors in the specific principles of treatment are: development training, treatment of abnormal tone, training of movement patterns, use of afferent stimuli, use of active movement, facilitation, abnormal and normal overflow, and prevention of deformity.

There are some symptoms specific to all children with Rett syndrome and these are considered now, and although some proposals are made for treatment methods, it has been suggested that: 'The therapist should always be prepared to examine and use other ideas as and when appropriate so that the therapeutic approach becomes broad based and capable of adaptation to the needs and circumstances of individual children and their families' (Eckersley and King, 1993, p.324).

2.2.3.1 Movement

It has been seen that ataxia of general movements and of gait in girls with Rett syndrome may arise as a result of damage to the cerebellum or its tracts. This damage interferes with the normal sequence and organisation of movement patterns, which become unco-ordinated and clumsy (Cash, 1974). Whole body movement planning will be affected, with movements becoming difficult to achieve (Armstrong, 1992).

Physiotherapy aims to give the child experiences that will, through sensations and movement, themselves improve motor response and skill (Eckersley and Steel, 1993). A programme of treatment will be set in motion by the physiotherapist, involving the training of normal postural mechanisms and locomotor reactions and the correction any asymmetries of posture, movement or balance. It is not until coordination of gross motor skills have been assessed and evaluated with respect to a child’s age and experience, that her particular level of dysfunction and its effects on classroom function becomes clear (Watter, 1984). This information provides the therapist with a basis for beginning treatment, as well as revealing other areas in which further specialist assessment may be necessary.
Access to appropriate seating and standing equipment is an essential part of the treatment of a child with a motor disability. This will need to be reviewed regularly by the therapist and is another opportunity for meaningful interaction with parents.

2.2.3.2 Hand stereotyphies

Stereotyped hand movements represent one of the most distinguishing characteristics of Rett syndrome, with ritualistic hand wringing, hand tapping and hand to mouth movements resulting in the loss of purposeful hand function (Hagberg, 1992a, Kerr, 1995a, Rett, 1992). Although it has been suggested that these repetitive hand movements should be ignored except when actual injury occurs, or if it is found that the performance of one hand improves when the other is restrained (Kerr, Southall and Samuels, 1992), educational opportunities could be increased if some degree of functional hand movement was established. This is an area of potential conflict between teachers, therapists and parents, and some agreement will need to be reached concerning the restriction of hand movements. Physiotherapy is not solely concerned with gross motor movements and the slow reversal PNF technique could be helpful in preparing a child for the activities of grasping and releasing, or for constructional activities.

There is controversy over the value of the practice of splinting and confining the arms and hands (Rett, 1985b). Naganuma and Billingsley (1988) examined the effect of bilateral hand splints on the persistent stereotypic hand movements on three girls with Rett syndrome. Although they reported significant reductions in stereotypic hand behaviours, a replication study by Tuten and Miedaner (1989) did not validate this. Aron (1990) used elbow splints to inhibit incessant hand to mouth movements, with positive results. As well as decreased hand to mouth movements and decreased hand wringing behaviour, increased socialisation and interaction with the environment were observed. Many girls with Rett syndrome are unable to tolerate the application of hand or arm splints for even short periods, and the effect may be worse than the condition (Van Acker, 1991). This may be one reason why studies in this area are limited.

Budden (1995) considered that reducing repetitive purposeless movements is effective in decreasing agitation, self-injurious behaviour, improving focus on activities, maintaining sustained attention and prompting hand use and socialisation. He recommended the use of soft elbow splints to achieve this and the
use of hand splints to prevent deformities of the thumbs. Research is continuing in this area, and the effect of splinting techniques on hand stereotypies and hand function is currently being examined (Freeman, undated).

Parents' views on the restraint of hands stereotypies and the use of splints are discussed in Chapter 6, but it seems that this is an area where the sensitivity of the physiotherapist should be a priority and where consultation with parents should be mandatory.

2.2.3.3 Scoliosis

Scoliosis has been a repeatedly common finding in more severely affected Rett girls (Harrison and Webb, 1990). The development of scoliosis in girls with Rett syndrome is dependent more on complex neurological factors and stage of disease than age (Stokland, Lidström and Hagberg, 1993). As the disorder progresses, scoliosis gradually manifests itself, and an earlier low muscle tone can change into rigidity and spasticity. The spinal deformity itself is diagnosed by the appearance of asymmetry of the ribs noticed from behind, sometimes obliquity of the pelvis, and a loss of balance in a wheelchair. Symmetrical postures should be the starting positions for symmetrical movements. There will also be movements that involve each limb carrying out a different pattern at the same time. Maintenance of head trunk and pelvis in the starting position of symmetry can be continued if both arms and hands are used together in bilateral, alternate or reciprocal actions (Levitt, 1984).

Harrison and Webb (1990) argue that spinal orthosis can be used successfully to improve sitting balance and limit the tendency to spinal curve progression, although they recognise the limitations of such treatment, particularly the need for frequent adjustment and advice. Brace treatment is not always successful and is sometimes badly tolerated by Rett syndrome girls, due to their particular mood deviations (Stokland, Lidström and Hagberg, 1993). As with hand splinting, there is a danger that the effect of bracing could actually be worse than the condition itself.

2.2.4 Organisation of therapy

It has been seen that much physiotherapy work is carried out individually (Stanton, 1992). Individual treatment is only a very small part of a child's life and in order to succeed in improving a child's performance, practice of the necessary skills must
be incorporated into normal daily activities (Association of Paediatric Chartered Physiotherapists, undated). A realistic management plan should be devised and agreed with everyone regularly involved with the child and necessary training should be given. This should ensure that even individual treatment is not given in total isolation, and that improvements made can more easily be consolidated in other areas.

In group situations, a variety of behavioural and social problems, including short attention span, distractibility and hyperactivity may be evident in children with disabilities (Magill-Evans and Restall, 1991) which may cause disruption, particularly if the tasks are repetitive, with little consideration given to individual interests. It is difficult to see how continual group work can either maintain interest or cater for each end of the spectrum of competence, even allowing that children are grouped roughly according to ability. Just as some children will respond well to an individual approach, so some treatments will determine the organisation of the therapy. Even group work such as Conductive Education relies largely on individual help within the group and this is an area where parental input can be beneficial.

2.2.5 The role of the physiotherapist

An increased understanding of the complexities of treating children with profound and multiple difficulties, of which a physical disability is only a part, has led to a change in role of the physiotherapist over the last few decades. Although the more established systems and methods of neurodevelopmental treatment, proprioceptive neuromuscular facilitation, muscle education and braces and patterning vary somewhat, both in theoretical background and in the use of aids and techniques, they are united in their separation between treatment and education (Cotton, 1984). Cotton refers to this as the linear approach, where movement, function and language are taught as separate items and according to the preferred methods of the therapist or educator.

More recently, the linear approach has been referred to as the reductionist approach (Kramer and Hinojosa, 1993). These authors suggest that in linear progression, several components of a process must occur before the skill as a whole is acquired or learned. The therapy techniques of 'drilling away' at medical signs and symptoms, as frequently as possible, are no longer the whole story
(Levitt, 1984) and the more holistic approaches of conductive education and movement education seem to have gained in popularity.

One of the physiotherapist's roles is the management of the therapy, and it has been claimed that '... the effectiveness of a therapist is measured not by what happens in her presence but by that which occurs in her absence' (Rushforth, 1984, p.106). Parents may not be aware of this unseen role, which in a school is likely to involve, at minimum, some consultation with staff involved with pupils on a day to day basis.

Another important role for the physiotherapist is the assessment of need and provision of aids, both in the school and at home. Access to all areas will need to be considered, and this is likely to be particularly problematic in a mainstream school. This role is likely to be shared by a number of professionals, including occupational therapists and speech therapists, and may 'create difficulties for the family if co-operation between professionals is fragmented' (Kennedy, 1984, p.227). Teachers, as well as parents, will be involved and Kennedy suggests that one therapist should maintain liaison between all the different parties.

2.2.5.1 Parental involvement

As well as a clinical role, the paediatric physiotherapist has an important role as an adviser to children, their parents and families, carers and professional colleagues (Association of Paediatric Chartered Physiotherapists, undated). Every effort should be made to ensure that both the child and the parents understand the objectives of physiotherapy and how these are going to be achieved.

Time spent explaining in simple terms 'the why, when and how' is well rewarded in co-operation and trust. The physiotherapist must remember that listening is often more important than talking and to do so will often reveal problems or anxieties that the parents were too shy to mention in case they sounded too trivial or complaining.

(Hyde, 1984, p.188)

Parents have a wealth of information about their child and have a vital role to play in their development. They should be encouraged to assist their children and participate in opportunities to increase their understanding of their special needs, and this is acknowledged in recent Government publications (DFE, 1994, DfEE, 1997).
It is essential for the optimal improvement in the child's function that treatment, like assessment, be multidisciplinary, and dictated by the child's specific needs. At the same time the utmost care must be given to maintaining the integrity of the family and providing supportive measures where needed. Overloading the child and the family by the involvement of too many professionals can be avoided by establishing the priority of the family's need. Therapy should enable the family to cope better, and should never increase the stresses under which it operates.

(Watter, 1984, p.159)

2.2.6 Summary

A girl with Rett syndrome has many disadvantages in life, not least of which is the inability to control gross and fine motor actions and physiotherapy is strongly placed to assist in this area. Although the term physiotherapy implies some form of correction to restore normal function, within the concept of many disabilities including Rett syndrome, it has much wider connotations. It has been found that therapy applies not only to the deficit needs created by the children's disabling conditions, but also to their growth needs (Ouvry, 1987), and so it overlaps, contributing to both care and education.

Difficulties with movement are a serious disability for the child with Rett syndrome, but there are methods of treatment. A knowledge and understanding of her difficulties in gaining control of movement, along with her inability to interrupt the repeated sequences of involuntary movements, is essential. Situations in which she is able to achieve pleasurable and useful movement - usually in which her interest in gained and no pressure is imposed - need to be discovered, and clearly these are important tasks for the physiotherapist. Hanks (1990) believes that physiotherapy treatment should be more frequent during periods of loss of transitional skills, and aimed at maintaining function, slowing deformities, reducing irritability and providing comfort in order to improve the quality of life for the girls and their caregivers.

While many physiotherapy treatments can be comfortably incorporated in a school environment, it appears that the special nature of the process requires a professional person in a specific role. The therapist needs to be teacher and physician, combining the skills of both. To some extent it would be possible to see these roles combined, but there seems to be an absence of literature regarding this point. It is not so easy to see teachers administering therapy, though the notion of
the teacher participating in and directing the process may not be that unrealistic given adequate resources and funding.

2.3 **Speech therapy**

The ability to communicate is a basic human need. Speech and language therapy is the process of enabling people to communicate to the best of their ability (Royal College of Speech and Language Therapists, 1997). Communication is a keystone of society, facilitating interaction between individuals, and between groups. It is a keystone too of education. The spoken word is our primary communication tool, supported by body language, gestures and eye contact, which all act as communication modifiers. Progressive speech acquisition through infancy into adulthood is considered the norm, but 2.5 million people in the UK are considered to have a speech or language difficulty (Royal College of Speech and Language Therapists, 1997). This figure should be viewed with caution, since the term 'difficulty' is open to interpretation, but the number serves to illustrate the possible scale of the problem. The difficulties associated with Rett syndrome and other profound disabilities are very complex, and there are many symptoms, other than loss of speech, which can be addressed through the medium of speech and language therapy, referred to here simply as speech therapy.

Careful documentation of progress is critical in determining whether intervention is needed, and the nature and extent of intervention (Garber and Veydt, 1992). These authors believe that as social and communicative skills emerge, therapy should work towards increasing the variety of early intentions, increasing the forms that they take, and increasing the variety of contexts under which they occur. Professional guidelines for good practice by speech therapists emphasise skill sharing, flexible working practices and the integration of therapy and educational programmes (Royal College of Speech and Language Therapists, 1996).

Meyerson (1985) emphasised the need for full knowledge of a syndrome when a person is first seen by a speech therapist: ‘... it is of critical importance that we understand the physical characteristics, the neurologic implications, the possibility of progression of symptoms, the prognosis, and the multiplicity of problems that can arise’ (p.47).
Meyerson also suggests that intervention is often not planned until a diagnosis is made, with a subsequent diagnostic delay resulting in a critical loss of remedial opportunity. Although early intervention is desirable (Kerr, 1988, Van Acker, 1991) it is crucial that the therapist has full information before implementing an intervention programme. In some conditions, such as pseudo-bulbar palsy, it is essential that exercises that might exacerbate a neuromuscular degenerative condition are not used; similarly with voice disorders, a speech therapist would never intervene until there was a clear picture regarding pathology.

2.3.1 Theoretical approaches to language and language development

Until the 1970's there were two main theories of language and language development. The behaviourist approaches of Skinner, who regarded language as part of the process by which human beings interact with their environment, and the grammarian approach of Chomsky who focused attention on the acquisition of syntactic rules as a basis for language (Kiernan, 1988) led to a large number of intervention studies with children with a wide range of types of language delay or disorder (Harris, 1990).

More recent research on normally developing infants and young children can be unified into two theoretical approaches, (Coupe O'Kane and Goldbart, 1998) and a knowledge of these is necessary if effective intervention strategies are to be devised for children with language and communication difficulties.

2.3.1.1 The psycholinguistic approach

This approach offers an explanation of observations in the early utterances of young children and has four underlying principles (Coupe O'Kane and Goldbart, 1998, p.2).

1. Some level of cognitive intentionality is required for the development of communication.

2. The first words or protowords that children produce express things to do with meanings.

3. Expressions of meaning and content are an encoding of the child's existing knowledge about the world.
Language development is built, at least in part, on cognitive development.

A summary of the stages in development according to the psycholinguistic approach is given below. This is drawn from Piaget's findings on the sensori-motor period. The stages defined by Piaget are still a major source of reference today. His work is important when examining the cognitive foundations for language development, as it provided one of the first developmental accounts of the emergence of logical thought (Harris, 1990). The sensori-motor stage from 0 - 2 years is typified by what Piaget referred to as 'egocentric thinking'. Children centre on their own actions, their body and their knowledge, and are unable to take anyone else's needs or interests into account. Gradually infants learn about objects, themselves, their world, and the people in their world. They begin to understand how one thing can cause another, and acquire simple ideas of time and space, as well as acquiring prelinguistic skills. The ages below are approximate and it is not assumed that the cognitive behaviour necessarily implies that the semantic or symbolic behaviour will be established (Coupe O’Kane and Goldbart, 1998).

In stages 1 and 2 (1 - 4 months) the object presents sensory impressions which may be 'sensed' by the child. Continued looking (or touching, smelling or listening to) is encouraged by the pleasure evoked by the sensory impressions, which may persist after the object is moved. During stage 3 (4 - 8 months) the child begins to extrapolate in time and space by a series of manoeuvres: (a) he shows visual anticipation of movements of the object; (b) he searches for the object; (c) he plays with a 'there - not there' situation - he looks at the object, then turns away, then looks back at the object again, and does this repeatedly; (d) he anticipates the whole object when seeing only a part; (e) he will remove an obstruction to see the object. Stage 4 (8 - 12 months) is when the child searches for the object with increasing degrees of sophistication and in Stage 5 (12 - 18 months) he focuses his search upon the place where the object was last seen. Stage 6 (18 - 24 months) is when the child searches for objects by imagining a series of possible positions that they might be occupying. In this way, he demonstrates his awareness that objects have an existence apart from himself.
Although the psycholinguistic approach gives some explanation of the early speech of young children (Coupe and Goldbart, 1988), it is the sociolinguistic approach that explains why language and communication skills develop.

2.3.1.2 The sociolinguistic approach

In the sociolinguistic approach, the emphasis is on the child in a social setting, emphasising the importance of the child as an active participant in learning language and communication. Many children who experience difficulties with language and communication also experience limited opportunities for engaging in social routines (Harris, 1990). This author suggests that any naturally occurring activity, such as feeding, shopping or playing games, is a potential context for language learning, and intervention strategies are planned which make use of this.

Coupe O’Kane and Goldbart (1998, p.2) discuss five fundamental principles of the sociolinguistic approach:

1. Language is acquired only if the child has reason to communicate.

2. Interactions between the language learner and more mature language learners are very important.

3. Language is learned in ‘dynamic social interactions’ and so positive contacts with adults in essential.

4. Language is a progression from simpler forms of communication.

5. Children are active participants in the process of learning communication.

The importance of the social context in which the child learns is now widely accepted, as are the affective issues that influence performance. With respect to acquired language, the implications for measurement of communication ability are significant, because performance is situation-specific.

2.3.2 Development of language

Goldbart (1994) argues that by looking at the way a baby learns cognitive and communication skills, it is possible to recognise and value small changes in the development of children with profound and multiple learning difficulties, so helping to plan further small steps for teaching or therapy. Within the autistic spectrum, the
general level of intellectual functioning covers almost the full range of intellectual ability (Jordan and Powell, 1990) and the range of language difficulties associated with autism is also wide (Jordan and Powell, 1995). The cognitive and linguistic abilities of girls with Rett syndrome are without exception very limited (Perry, 1991) and most of them have not exceeded Piaget’s 3rd sensori-motor stage (Fontanesi and Haas, 1988, Olsson and Rett, 1985).

The Rett Syndrome Diagnostic Criteria Work Group (1988) reports that expressive skills do not develop beyond the earliest level even in pre-regression, and that regression in verbal skills is accompanied by a general regression in psychomotor skills. There are similar findings in the work of Coleman, Brubaker, Hunter and Smith (1988) and Kerr and Stephenson (1985). Reactions to external stimuli such as hunger or thirst were noted, but in a study of 63 girls, 81% were considered by their parents to be insensitive to pain. Kerr (1987b) stated that language was never completely normal in the child with Rett syndrome and that imaginative and imitative activities were never demonstrated, regardless of the child’s age at regression.

2.3.3 Intervention strategies

Coupe, Barber and Murphy (1988) confirm that teachers, speech therapists and psychologists have struggled for years to understand and apply appropriate intervention strategies for pupils who function at a very early stage of communication. They suggest that throughout the stage of pre-intentional communication a developmental sequence can be taken into account that progresses through three levels, reflexive, reactive and proactive. At the reflexive level, the child has a limited repertoire of behaviours that can be interpreted by familiar people, although these behaviours are non-intentional. At the reactive level the child has a response of voluntary behaviours and is beginning to react to both objects and people. When a child reaches the proactive level, purposeful acts on objects, events and people in the environment can be observed.

There are a number of approaches to ensure appropriate intervention strategies, and the following examples are particularly appropriate to children with severe cognitive delay. Coupe O’Kane and Goldbart (1998) describe affective communication as being concerned with ‘interpreting or placing communicative meaning on individual’s responses to a variety of internal and external stimuli’
Coupe, Barber and Murphy (1988) discuss the Affective Communication Assessment (ACA) and believe this can open up channels for teaching and monitoring development. The ACA is concerned with interpreting or placing communicative meaning on individuals' responses to a variety of internal and external stimuli (Coupe O'Kane and Goldbart, 1998) and was developed as a tool for enabling adults to make predictions regarding the child's range of affective communication, so allowing appropriate programmes of intervention to be planned. This assessment does rely on an adult's interpretation of a child's response and Goldbart (1994) cites Yoder (1987) who demonstrated that signals from children with severe disabilities are more difficult to interpret, suggesting that some form of training is advisable before programmes are planned.

The Pre-verbal Communication Schedule (PVCS) allows teachers and other people working with children with minimal verbal or signing skills to identify the child's current abilities and to develop teaching programmes for the development of communication skill (Kiernan, 1988). The PVCS aims to provide an assessment for all pupils with severe learning difficulties, and relies on a thorough knowledge of the child by therapist, teacher or parent.

2.3.3.1 Lack of speech

The term 'non-verbal' has been used to describe children who seem unable to use spoken language but are able to communicate using signs and gestures, whereas 'non-communicating' is used to describe those children who seem uninterested in communication of any kind (Harris, 1990). Even girls with Rett syndrome who appear to try to communicate have restricted means from which to choose, as speech is limited and the interpretation of non-verbal signals is likely to be difficult.

One explanation for the failure to regain speech is related to the severe cognitive disability of girls with Rett syndrome, resulting from disorders in the brain and the nervous system (Woodyatt and Ozanne, 1994). Another is the presence of dysarthria, which arises from difficulties with the control of speech musculature (Harris, 1990) and is found in neurological disorders such as Rett syndrome. It affects the muscles of speech, such as jaws, cheeks, tongue, soft palate, larynx and lips and may lead to unintelligible speech or even lack of speech. Therapy programmes will be aimed at motor exercises, such as in the Nuffield programme, for lips, tongue and breathing, with the emphasis on motor planning and
sequencing, rather than strengthening muscles, although it should be remembered that it may not be sufficient to concentrate solely on the acquisition of skills.

Whatever the cause, the therapist has to make the decision whether language intervention should focus on spoken language, or whether opportunities should be given to the child to increase skills which use a visual rather than an auditory process (Harris, 1990). This decision is likely to be taken in conjunction with parents and teachers, but the prognosis for regaining speech with girls with Rett syndrome is not good, and this feature will need to be taken into consideration. It will be seen that many girls have the ability to communicate by means other than speech, and this could well influence the route taken.

Although girls with Rett syndrome are considered to be 'non-verbal', as they rarely use more than single word speech or carry out more than the simplest spoken instructions in the pre-regression phase, some girls have meaningful words and a few can manage phrases (Zappella, 1992).

**Single words in context**

Occasionally a few useful words are retained or learned and it is a feature of the condition that these may be suddenly and aptly produced (Kerr, 1992b, Lindberg, 1994). Babbling and occasional sudden bursts of words are not uncommon, although it has remained a constant mystery why single words may be uttered, apparently in the right context, even after several years without speech.

They have also lost babble, and learned single words if the regression period starts later. So the loss of words during the regression period is absolute but then I have some examples of where 5 or 10 years later they learn some words. Suddenly they say that word or around half a sentence then they are silent again for 10 years. It comes suddenly without any connection to anything. Some girls when they are severely stressed can suddenly say something which has relevance to the situation.

(Hagberg, 1992a, p.16)

Reports on expressive language skills focus on lists of retained words or general statements on the presence or absence of words (Woodyatt and Ozanne, 1992b), with little detailed descriptions of the communicative functions of the words, nor their context or situational appropriateness.
Preserved speech

Zappella (1992) reports instances of girls with preserved speech who were able to say numerous words and even fully formed phrases and sentences. These girls showed all the features originally described by Rett (Rett Syndrome Diagnostic Criteria Work Group, 1988) with this one noticeable difference. The existence of variants of Rett syndrome has been considered for a number of years (Goutières and Aicardi, 1986, Suzuki, Matsuzaka, Hirayama, Sakuragawa, Arima, Tateno, Tojo and Suzuki, 1986) and Zappella (1992) suggests that these cases could be considered as a definite, 'speaking' variant of the syndrome.

2.3.3.2 Comprehension deficit

Language comprehension deficit is rarely mentioned in reports of Rett syndrome girls even though it is considered by the Rett Syndrome Diagnostic Criteria Work Group (1988) to be a necessary criterion for diagnosis (Garber and Veydt, 1992). Studies have shown that girls typically perform at a profoundly impaired level of disability (Olsson and Rett, 1985, 1987, Perry, Sarlo-McGarvey and Haddad, 1991, Woodyatt and Ozanne, 1992a, 1992b, 1993). This does imply a serious receptive language disorder, although post regression expressive language skills show a wider range of abilities than observed in receptive language (Woodyatt and Ozanne, 1992b). Hagberg and Witt-Engerström (1986) support the hypothesis of cognitive arrest at the developmental level achieved at the age of onset of the condition, in association with an expressive language disorder.

It is often believed, particularly amongst parents and carers, that girls with Rett syndrome do understand more than is apparent (Lindberg, 1994) and numerous anecdotal reports exist that suggest these girls may understand more than they can express (Weisz, 1987), although the limited amount of research in this area does not substantiate these claims. It is likely that parents desperately want their girls to communicate, and therefore miss no opportunity to interpret non-verbal signals and consequently assume a comprehension that may not be present.

Nomura, Segawa and Hasegawa (1984) reported that comprehension of verbal and non-verbal commands was non-existent, although Trevathan and Naidu (1988) stated that minimal comprehension might continue beyond four years. The ability to understand words is difficult to assess, due to fluctuating attention and their limited ability to perform voluntary movements (Witt-Engerström, 1993) although pictures
of familiar situations appear to be understood (Lindberg, 1994). Certainly, there is
an indication that girls with Rett syndrome have a lack of, or an extremely limited,
ability to decode verbal stimuli.

Speech therapy is one method of searching for comprehension deficit, and an
improvement in communication skill, however achieved, would almost certainly
influence perceptions of how much or little a child could understand. An early
communication curriculum framework as outlined by Coupe and Jolliffe (1988)
would highlight the main areas of development and experience and provide 'the
direction and justification for the education which pupils subsequently receive'
(p.93).

2.3.3.3 Other symptoms

Stereotyped oral and perioral movements including bruxism, tongue protrusion,
licking and slow involuntary movements are not uncommon in all stages of Rett
syndrome (Kerr and Stephenson, 1985, Hagberg, Aicardi, Dias and Ramos, 1983).
Rett (1977) reported smiling in girls of 3 years or above, but this was sporadic, short
lived and seemingly unrelated to environmental events. Nomura, Segawa and
Hasegawa (1984) noted a lack of facial expression, while Hagberg et al. (1983)
found facial grimacing to be prominent in many Rett children aged 2 - 5 years.

Kerr and Corbett (1994) cite a study by Morton of 20 children, which found that the
oral phase of swallowing was disturbed, with poor lip and palatal elevation and jaw
movement. This may result in nasal escape of food, and may lead to impairment of
chewing ability. The subsequent failure to manage food in the mouth is
characteristic of Rett syndrome girls (Morton, Bonas, Minford, Kerr and Ellis, 1997).
As well as having a profound impact on communication skills, these symptoms
inevitably detract from normal feeding development and nutrition can be a major
problem for girls. The typical girl with Rett syndrome is extremely thin and in spite
of having a good appetite, has difficulty gaining weight. Kerr (1991) suggests that
possible problems include the shape or posture of head and neck; inability to keep
the lips closed on food; defective chewing; defective swallowing; choking;
involuntary or obstructing movements; vomiting or regurgitation; excessive
secretions; defective appetite and dependence in feeding. It is likely that feeding
techniques will need to be adjusted by parents, and speech therapists are strongly
placed to advise in this area.
Bedford and McKinlay (1993) suggest that speech therapists and physiotherapists need to work together to address the problems associated with control of jaw, lips and tongue. Motor exercises to stimulate and facilitate movement in the mouth may be appropriate, and PNF techniques are often used to facilitate feeding. To gain maximum benefit for the child, by ensuring continuity and generalisation into other contexts, the involvement of parents and teachers is essential.

Visual assessment has shown that the functional use of vision and hearing is unimpaired in children with Rett syndrome, although it seems that the functionality is not utilised (von Tetzchner, Jacobsen, Smith, Skjeldal, Heiberg and Fagan, 1996, Kerr 1988). Lenn, Olsho and Turk (1986) felt that a study of auditory function may contribute to an understanding of Rett syndrome and provide an additional basis for planning speech and language therapy. Consistency in approach is vital for children with profound disabilities, even more so for those who are not fully utilising vision and hearing (Mclnnes and Treffry, 1993). The child may require specific sessions to stimulate or train residual hearing and the speech therapist will enable individual children to heighten their awareness of their ability to make sounds and to communicate. Again, consultation with other professionals and parents will ensure the generalisation of new skills into other areas.

2.3.4 Communication

The most common type of communicative act for girls with Rett syndrome appears to be non-verbal, such as looking, touching and gesturing for such purposes as requesting or protesting (Coleman, Brubaker, Hunter and Smith, 1988). Woodyatt and Ozanne (1993) indicate that one factor in determining the ability of a girl to communicate seems to be the awareness of caregivers of the range of behaviours that may be interpreted as communication. They emphasise the need for the assessment and treatment of communication skills to focus on the intentionality and functionality of the child's communication.

During the regression stage of Rett syndrome, girls may withdraw from the outer world into a state of isolation, or even exhibit behaviours which may initially lead to a diagnosis of infantile autism (Lindberg, 1994).
However, childhood autism does not typically present the apparently intense desire to communicate through eyes, gestures, body language and pointing, found in Rett syndrome.

One parent has stated:

Our daughter is 3½ years old. Her name is Sally. She was diagnosed with Rett Syndrome at 20 months. Sally can no longer feed herself, but she wants to very badly. Sally, by touching my outstretched hand, answers "yes" to questions. Today at lunch she grabbed her hamburger over and over. So I asked her, "Sally, do you want to be able to feed yourself?". She touched my hand to say yes. It broke my heart.

(Personal communication, 1996)

When speech and language are lost, parents may well turn to communication modifiers, such as natural gestures (pointing, eye gaze), vocalisations, facial expression and body motions (Garber and Veydt, 1992) in an attempt to hold onto that which they have lost.

As a child with Rett syndrome leaves the regression stage, so her ability to communicate improves and the establishment of an effective means by which the girl can make her wishes known is of primary importance to her and her family (Kerr 1988). Normal speech, however, is impossible, and so other channels of communication need to be found. The speech therapist will need to evaluate the girls' responses to cause and effect, making choices, following simple directions and showing recognition of objects and people, in order to establish appropriate forms of communication.

2.3.4.1 Use of eyes

Eye contact is one of the earliest communications between mother and baby (Latham, 1984). Direct eye contact continues to be important throughout adult life, and use of the eyes for referring to external objects during ordinary conversation becomes automatic for the majority of people.

"One of the fundamental mechanisms which allow social behaviours to develop is that of eye contact" (Jordan and Powell, 1995, p.19). These authors report that if children with autism do use eye gaze they are not able to use it appropriately for communication. They suggest that gaze behaviour has to be interpreted within its
interactive setting, and that the behaviour of the caregiver develops according to feedback gained in social interchanges.

Vision and gaze are considered paramount in enabling girls with Rett syndrome to learn about their environment and to interact with their surroundings (Olsson and Rett, 1990, Rett, 1992). A recent study (von Tetzchner, Jacobsen, Smith, Skjeldal, Heiberg and Fagan, 1996) found that persistent 'looking' was associated with low cognitive functioning, a counter-intuitive for parents and professionals who regard this persistent looking as an indication of interest and an ability to concentrate. Active gaze, as opposed to persistent looking, was considered a more useful tool in assessing language comprehension in Rett syndrome. Research by von Tetzchner et al. (1996) indicated that girls with Rett syndrome need more visual-processing time than is normally required. Confusing information is received from the sense organs and reaction time is relatively slow (Kerr, 1988).

The critical element in the development of a meaningful communication system depends upon the successful interaction of the person and her environment (Van Acker, 1991). A study of six girls with Rett syndrome by Woodyatt and Ozanne (1993) supported reports of improvement in social interaction skills, but found that girls were still operating at a pre-intentional level, regardless of age or stage of the syndrome. These authors suggested that this apparent improvement is actually an increase in the number of behaviours which caregivers interpret as communicative, rather than a change in the communicative behaviour of the child herself.

There are, without doubt, interrelationships between cognition, physical skills and communication (Woodyatt and Ozanne, 1994), but just as the age of onset and duration of each stage is individual to each girl, so may these interrelationships differ, and it is the responsibility of the carer to ensure effective means of communication for each individual. Given time and opportunity to choose, most girls will make a choice. Many girls eye-point spontaneously and for the non-ambulant and more immobile girls, eye-pointing for choice will be the most practical tool (Lindberg, 1994). Lindberg suggests the use of pictures to make selection and communication easier, as many girls are able to make quick and stable associations between the pictures and the things they represent.
2.3.4.2 Social interaction

Shortly after recognising the syndrome, Rett (1977) found that the social interactions of the girls he was studying were severely limited by lack of active and passive vocabularies, language comprehension, learning (defined as memory and imitation) and play activity. Swedish studies showed that play repertoire never advanced beyond a functional level i.e. the girls never took the decisive step from the sensori-motor to the imaginative stage of maturation (Witt-Engerström, 1993). In similar studies Nomura, Segawa and Hasegawa (1984) and Rolando (1985) reported lack of eye contact, absence of response to persons and objects and no recognition of familiar people and objects, although Lindberg (1994) found that pleasure and displeasure could be shown by emotional expressions, body language and facial expression.

A survey of twelve girls by Lewis and Wilson (1998) showed that a range of communication skills was apparent. All girls were able to indicate likes and dislikes, comfort and discomfort and feelings of sadness or pleasure. Communication and learning seem to be facilitated by close cuddling contact with a known carer, but became impossible in a noisy or insecure environment (Kerr, 1988). Noise and sounds, though, when used in a planned manner, may enhance the girls' weakened ability to communicate. Wesecky (1986) and Zappella (1986) discuss the value of musical instruments in the promotion of a girl's desire to interact with her environment.

Personal communication through touch and sounds is a particularly rewarding area for the girl with Rett syndrome. Kerr (1991) believes that everyone involved in her care can help this communication, by protecting her long term relationships with the people who matter most to her, by making time for undemanding face to face interaction, by learning to use and respond to non-speech vocalisation.

2.3.4.3 Alternative forms of communication

In considering all elements of communication, careful evaluation must take into consideration parent and caregiver observations, the girl's responses to cause and effect, making choices, following simple directions and showing recognition of objects and people. This information can provide a basis on which to establish alternative forms of communication, including communication boards and switch-activated systems which facilitate environmental access (Budden, 1995, Van Acker,
Lindberg (1990) felt that direct selection was a good beginning as it enabled girls to make known their own opinions and preferences. Simple non-speech communication using eye pointing, sounds and gestures may be taught. Basic mechanical or electrical devices have an important place, and it may be possible to teach ways of indicating 'yes' or 'no'. Preliminary trials with a 'yes / no' panel and touch pads were carried out by Kerr (1992b). The results suggest that some girls may be able to use simple communication boards for choice and may learn to associate action, colour and word with the wish to accept, refuse or change.

Computer learning programmes also provide opportunities for communication, although the stereotypical involuntary hand movements may make the success of this difficult to judge. Kerr (1997) warned that although written symbols can be useful in offering choices, written words and computer responses are of much less importance to girls with Rett syndrome than other kinds of communication. This is of some significance in education, and while teachers may be aware of the limitations and benefits of computer aided learning, they should also be aware of the personal nature of Rett girls' communication difficulties and therefore use computer options cautiously. Jordan, Jones and Murray (1998) have suggested that augmentative systems of communication have an additional role in autism, and can successfully be used to assist learning and communication regardless of the level of speech.

2.3.5 Organisation of speech therapy

Lewis and Wilson (1998) suggest that the individual approach to speech therapy is most common for girls with Rett syndrome, and case studies by Garber and Veydt (1992) support this view. However, Golding (1997) highlighted the importance of a peer group for children within the autistic spectrum, in providing opportunities for tolerance, turn taking and communication. It has been pointed out that although most children with autism will need some access to individual teaching, some children will find this difficult to endure, and respond better in a small group situation (Jordan and Powell, 1997). Reid, Millar, Tait, Donaldson, Dean, Thomson and Grieve (1996) believe that although there is a place for individual therapy for children with special needs, withdrawal-based intervention should not be considered the only option. Again, speech therapy is well placed in a school
environment and is frequently used, but more information is needed to allow schools to structure delivery in the most beneficial way.

2.3.6 The role of the speech therapist

In has been seen that speech therapy is not involved just with the production of the spoken word, but rather with all aspects of communication, as well as aspects of feeding. For example, proper seating is essential to reduce the risk of choking during feeding (Van Acker, 1991) and input from physiotherapist and speech therapist together can determine appropriate positioning.

Harris (1990) discusses the varying roles of the speech therapist, from 'hands-on' to enabler, and suggests that people without specialist qualifications can play an important part in the delivery of language intervention programmes. The joint approach of therapists, parents and teachers should ensure that the individual needs of the child remain a priority, and this is discussed further in Chapter 5.

2.3.7 Summary

With such a complex syndrome as Rett, it is difficult to isolate the effect of any single influence. The conclusion drawn from a variety of sources in the literature is that speech therapy should be able to influence the child in several ways, from the ability to chew and so manage food in the mouth to better emotional expression, although there is little published research specifically directed at the application of speech therapy to girls with Rett syndrome.

In summary, there is possibility for inhibiting regression, for improving post regression communication, for learning words, and for improving oral skills including food management. An experienced and qualified speech therapist should be able to address all these issues, whether directly or indirectly. Appropriate intervention strategies for pupils who function at a very early stage of communication need to be considered. Eye contact, looking, listening, attention, turn-taking and developing meaningful interaction are all areas addressed by the speech therapist. These aspects are likely to be addressed in an educational context, and should be programmed into a structure appropriate to the individual needs of a child with Rett syndrome.
2.4 **Music therapy**

Music therapy has developed internationally as an authentic paramedical profession alongside clinical psychology and speech therapy. The therapy extends to music psychotherapy in which therapeutic issues are addressed through various levels of verbal discourse or experience occurring within a musical environment (Bruscia, 1998), but this study will not explore the issues of psycho-analysis and the difficult to define process of transference, since in many disabilities, including those within the autistic spectrum, the root cause is neurological, and not a psychological response to past events and experiences. In education, music therapy is most widely used in the assessment and treatment of children with emotional, behavioural, social or psychological impairment (Association of Professional Music Therapists, 1992). Music therapy is particularly appropriate in cases where verbal communication is impaired or impossible, and it has been said that the therapy can provide an alternative to speech as a means of communicating (Wigram, 1995). This viewpoint needs to be considered with some caution as music therapy quite clearly will never replace speech as a detailed information carrier although it will aid expression of emotions and feelings that precedes language function.

Jordan, Jones and Murray (1998) cite Christie, Newson, Newson and Prevezer (1992) when discussing musical interaction therapy as an autism-specific approach that has been developed over a number of years at a school for autistic children. The therapy forms part of the school’s curriculum that prioritises the development of communication skills. Jordan et al. suggest that this approach is ‘compatible with other approaches which prioritise the building of relationships and communication skills and which acknowledge the particular needs of children with an asd’ (p.40). It has been seen that Rett syndrome is a pervasive developmental disorder within the autistic spectrum, and much research has been aimed specifically at this particular syndrome (Postacchini, 1993, Wigram, 1995). Professor Andreas Rett, the International Rett Syndrome Association and the UK Rett Syndrome Association have all strongly advocated the use of music therapy for minimising the disabling effects of Rett syndrome (Montague, 1988). However, it is not enough for individuals and organisations with a specific interest in the amelioration of the symptoms to promote the therapy, unless they can endorse their views by valid research findings. Schools are unlikely to include music therapy in the curriculum for one syndrome only, without unbiased confirmation of its benefits.
All children have access to music, as specified in the National Curriculum, where the aims are to teach composition and specific musical concepts. It will be well beyond the reach of some children to benefit from the structure imposed by this curriculum, and achievement of musical competency is not the purpose of music therapy, although it could aid proficiency. There has recently been evidence that music can be used beneficially for sufferers from profound disabilities, who seem to respond to musical sounds in remarkable ways. It has been found that girls with Rett syndrome, in spite of their apparent inability to make sense of much of their surroundings and experiences, seem to be very receptive to and discriminating of musical sounds (Etkin and Eisler, in UKRSA, undated). Why this should be is not clear, but the point is that simple musical activities can transcend the limited function of speech. This is likely to be through arousal of inhibited neural pathways.

2.4.1 Music and the brain

There is evidence that functions of intellect are localised in discrete areas of the brain, and that learned cognitive skills are associated with areas of the cerebral cortex. In life, each hemisphere of the brain becomes specialised to some extent, and for the right handed majority, language control appears to be located in the left hemisphere, while non-verbal and spatial orientation are concentrated in the right. This is an oversimplistic representation, and a succinct review of biological aspects of music function is offered by Sloboda (1990), who goes on to examine patterns of disorder in music. He concludes that ‘Music, if not a totally distinctive neural function, almost certainly employs a distinctive configuration of neural resources’ (p265).

Wesecky (1986) believes the receptivity for rhythm and melodies must be located within a primitive region of the hierarchical structure of the brain, as even severely disabled children respond to music:

We do not know which particular part of the brain is responsible for that reaction yet it remains a fact that in severely mentally retarded patients the capacity to perceive and accept rhythm and melodies remains at times the only access to social communication. Therefore it is obvious to use music as a therapeutic means with children with Rett syndrome in order to preserve or enhance motoric functioning. (p.254)
Significant work is being done in the fields of neurology. Modern neurological techniques are beginning to reveal the effects of music upon the brain. Electroencephalographic studies suggest that music creates a unique level of coherence of electrical activity in separate parts of the brain (Storr, 1993), to some extent complying with the suggestion of Sloboda (1990) that it is interaction between separate areas which make up musical function, rather than activity in a single locality. If this were the case, then it would not be surprising to find that music therapy held influence over other functions. It is likely that most music therapists do not think of specific areas of the brain as an integral part of the music therapy experience, but rather determine the aims of a session according to the emotional needs of the child, and it could be argued that the effects and results are more important than the theory. However, Erdonmez (1993) believes that:

... we, as music therapists, can add to the knowledge of brain functions in that we work with a highly creative art form, which activates many areas of the brain. We need to work alongside psychologists and neuropsychologists in order that we are better informed about the effect of music on brain functions, and so we can better inform other professionals about our clients’ non-verbal functioning level.

(p. 123)

Medical research has found that music is able to activate neurones for purposes of relaxing muscle tension, changing pulse and producing long-range memories that are directly related to the number of neurones activated in the experience. This can now be measured by injecting the brain with radioactive chemicals that are detected when the brain cells are active (Campbell, 1984).

Warwick (1995) believes the lack of knowledge about the efficacy of music therapy to be a major factor in the lack of popularity of music therapy in education. Personal experience has shown that music therapists working with children with special needs have little awareness of neurological effects of the therapy, and are primarily concerned with the overall manifested outcome. More credibility would be afforded to the profession if this imbalance were to be addressed, and if critical research were able to identify and explain real benefits.

2.4.2 Music therapy - theories and models

There are a number of practical models of music therapy, and those that relate particularly to children with physical and learning difficulties are discussed briefly in
the following section. Although most models of practice encourage expression, there are instances where there is emphasis on the feelings created during the sessions, rather than free expression, for example, when working with terminally ill children or children with degenerative conditions.

Whatever the theory behind the method of therapy, the approach is holistic, aiming to promote the balance between the emotional, physical, intellectual and social development of the child (Warwick, 1995). The balance between each area will depend on the individual needs of the child and will vary, possibly even from session to session.

Of course, in the world of education, music therapy, if it is provided at all, will form only part of a much wider sphere of activity. It may be extremely difficult, if not impossible to separate out the various effects of school activities when judging both the value of therapy and education, and what is important to the child and parent is the overall result. 'Music therapy is a holistic approach working with the skills, knowledge, experience and attitudes of each child. By its very nature, music therapy enables the child to benefit from, and increase attainment across, the full curriculum' (Fairfields School, 1995).

There are many approaches to music therapy, but all therapists will use some form of improvisation. This is child-centred, with the therapist responding musically to the output and the perceived emotional state of the child (Lawes and Woodcock, 1995). The child’s responses are the starting point for improvisation and remain central to the direction that the therapy takes (Howat, 1995).

These early interactions can lead to the formulation of specific goals, and these will be considered within the context of the improvised musical setting. In practice, what is fundamental to all approaches is the development of a relationship between the child and the therapist. Shared experiences using music can help the exploration and expression of an individual’s conscious and unconscious feelings.

2.4.2.1 Improvisational model

A number of improvisational models of therapy are based on an approach to music education known as Orff-Schulwerk (Bruscia, 1987). Although originally designed for children without special needs it has been widely used in special education and music therapy settings. Elemental music is defined as ‘the universal and primordial
tendency of human beings to make music spontaneously, using the natural rhythms of movement and speech’ (p.219) and this concept is at the root of all Orff-based models. The approach is based on a philosophy of music education that develops music concepts and skills while actively involving children in singing, movement, speech, rhythm, playing instruments and improvisation (University of Massachusetts, 1999). In teaching, the music is the main objective, in therapy the music is the tool for relating and communicating, not an end in itself (Warwick, 1995). The therapy should always take place in a non-competitive atmosphere where one of the rewards is pleasure (American Orff-Schulwerk Association, 1999).

Some children, including those with profound and multiple learning difficulties, may not have the ability to develop high level musical concepts and skills. Musical proficiency is not the aim of therapy and lack of musical skill should not preclude the use of this approach.

2.4.2.2 Creative model

The improvisation techniques of Nordoff and Robbins have become widely used and are known as creative music therapy. The techniques have been found particularly effective for children with a variety of disorders, including autism, emotional disturbance, learning disorders, physical impairments and neurological disorders (Bruscia, 1987). The overall goals of creative music therapy are to develop ‘expressive freedom, communication, inter-responsiveness while dispelling pathological behaviour patterns’ (p.67). The creative music therapy approach of Nordoff and Robbins has traditionally been understood and interpreted as focusing on the musical event and bypassing words (Pavlicevic, 1997).

There appear to be no intellectual prerequisites for participating in creative music therapy and it has been said that the child may be ‘non-verbal and low functioning or verbal and high functioning’ (Bruscia, 1987). This makes it especially appropriate for girls with Rett syndrome and other disorders within the autistic spectrum, where there may be variance of speech and learning ability.

2.4.2.3 Developmental model

The pioneering work of Juliette Alvin (Heal and Wigram (Eds.), 1993) with children with learning disabilities, set in motion the first realisations that music could be used as a way of reaching children with significant difficulties. She discovered that
allowing people with a developmental disability to watch and listen to her playing evoked an enormous reaction, and children with special needs were stimulated while they participated in an enjoyable activity. The main goals of this approach are to develop interpersonal relatedness through non-verbal and verbal modalities (Bruscia, 1987). It is suggested that the therapy room is equipped with musical instruments and projective play materials such as dolls, puppets, and that the child may either gravitate towards an instrument or materials, or that the therapist may attempt to engage the child.

Alvin's ideas progressed to a more interactive approach. The interactive feature of a music therapy assessment gives a clear indication of the child's receptive skills, expressive skills, non-verbal communication as well as their ability to engage in social relationships (Wigram, 1995).

As relationships develop, so opportunities are provided for developmental growth (Bruscia, 1987). Alvin used the relationships inherent in music to promote intellectual, physical and social-emotional developments. It has been seen that children with Rett syndrome may become withdrawn and frightened (Lindberg, 1994, Hagberg, 1993) as well as having learning and physical difficulties, and an interactive approach can be of benefit.

2.4.3 Music therapy and the treatment of symptoms

Music therapy is used with individuals of all ages and with a variety of conditions, including Rett syndrome. It is not the role of the music therapist to teach specific musical concepts, but to look specifically at and address the needs of the individual child (Warwick, 1995). Nordoff and Robbins (1977) believe that for the therapist to find the 'individualised musicality inborn in each child' there must be an initial element of direction. In debating the issue regarding directive and non-directive intervention, Bean (1995) considers that each has a value when it meets the needs of the child. On the one hand, the freedom for musical expression that non-directive therapy offers is a vital component in the therapeutic process, on the other hand, freedom without clear boundaries, musically or behaviourally, is not always desirable or appropriate. Bean believes that it is not simply a question of whether a programme should be directive or non-directive, but rather a question of incorporating different aspects of each approach with a bias towards one or other during the course of a balanced therapy programme. ‘These timely decisions of the
therapist are developed as part of the therapeutic and musical artistry in his or her work' (p.208). What is essential is that the music therapist knows the child, and the disability, well enough to select the most appropriate approach at any given time.

Over the last five years Wigram has been actively working with girls who have Rett syndrome and states that almost all of the people he has seen have responded very positively to music therapy (Wigram, 1995). The aspects of disability that may benefit from music therapy are outlined in the following sections, and it will be seen that all are in some way appropriate to the school setting.

2.4.3.1 Communication

Proficient communication is a human achievement that is unique in the natural world. No other species is able to transmit meaning, intention, and knowledge as effectively as we do. Communication is a skill acquired early in life. As we grow, our ability to receive information from our senses is extended, and we learn to appreciate variation in tone, timbre and pitch in spoken words. It could be that variation in tone and pitch can be detected long before the meaning of words is learned, and discriminatory skill is a much more basic function than interpretation of language. Since music is pitch, tone and timbre, very young infants are likely to be receptive to music before they learn to speak, and the fundamental emotions of comfort and discomfort could be triggered by sounds. It is logical then that responses to music are instinctive, a point made by Storr (1993) who considered that music can be the carrier for emotion just as can spoken words, sometimes even more so, transcending the barriers of language.

While casual giving of sounds may achieve some stimulus, a degree of structure is clearly needed in the ‘treatment’ in order to achieve the best results, just as structure turns infant sounds into language. Before structure must come an appreciation of what music therapy actually is. It is not straightforward, and has been described as ‘an interpersonal process in which the therapist uses music and all of its facets - physical, emotional, mental, social, aesthetic, and spiritual - to help children to improve, restore or maintain health’ (Bruscia, 1991, p.5).

Making music can provide a powerful medium for communication in the hands of the skilled music therapist. Non-verbal interaction, such as gesture and eye contact, as well as the dynamic use of musical elements such as volume, timbre
and frequency, can provide a child with Rett syndrome with a means of expression (Wigram, 1995). Actual vocalisation has also improved, from vague unrelated sounds to deliberate noises, including some approximations, such as 'Hello'. An increasing awareness of surroundings and an improved ability to relate to peers and adults have become apparent in girls who receive music therapy.

Whilst acknowledging that all therapists working with children with communication difficulties need to be aware of the exact nature of the difficulties, and consciously adapt methods of communication in order to meet the specific needs of the child, Oldfield (1995) believes that for music therapists the challenge is even greater.

For music therapists using the medium of music, the non-verbal aspects of communication become particularly important. There are no words to interpret and no complex sentences to 'cover up' basic difficulties in concentration and listening. With no verbal content to focus on, the manner in which the music therapist chooses to communicate is going to be of particular importance.

(p.226)

Etkin and Eisler (UKRSA, undated) believe that the effect of losing speech and the means of purposeful communication is one of the most serious disabilities of Rett syndrome and that girls should have access to music therapy by right. Purposeful communication is fundamental in the context of education, and therefore music therapy should be a prime candidate for school based therapy, although personal experience shows that access is far from automatic.

2.4.3.2 Stereotypic movements

The stereotypic movements of hands and fingers peculiar to Rett syndrome sufferers may preclude or seriously interfere with other movements, and with regular tasks and activities in school. Wing (1976) explains the movements as the only action available to a child who could not obtain pleasure from communicating with others or from activities involving understanding of complex abstractions. Schulz (1987) sees stereotypic movements as activities that can bring immediate satisfaction and provide a sense of achievement. The child is familiar with the actions, and is not exposed to anything new, threatening or beyond her understanding.
The therapeutic approach is likely to have to confront or work round some very deeply rooted neurologically driven behaviours such as hand plucking, while trying to motivate the child to make any necessary changes (Wigram, 1991). Jordan and Powell (1995) describe how stereotypic behaviour in children with autism can lead to a kind of sensory deprivation. Music therapy can encourage creativity that can, in turn, open up opportunities for interaction with others. Schulz (1987) believes that by incorporating stereotypic movements into acceptable activities, music therapy can offer ways to encourage and help children out of the small, isolated world of their stereotypies, enable new experiences and learning processes to take place, improve social contacts, and provide the child with a medium for emotional expression. Children with autism often have motor stereotypies (World Health Organisation, 1993), and although these are not restricted to the hand and finger movements seen in Rett syndrome, music therapy could offer benefits to children with these stereotypic movements.

2.4.3.3 Movement

A study by Hill (1997) indicates that music therapy can be particularly valuable for girls with Rett syndrome in maintaining skills already acquired before regression, particularly physical skills including fine motor movements. A case study by Postacchini (1993) described a child whose movements were initially characterised by considerable rigidity. Movement games that incorporated sound were introduced in a play situation. By relaxing in a rewarding and satisfying situation of multi-sensory stimuli, there was a perceptible improvement in balance and walking, with a subsequent improvement in spatial awareness.

2.4.3.4 Learning difficulties

Wigram (1995) considers that music therapy provides a strong and motivating stimulus for girls with Rett syndrome and believes this is a way of developing attention and concentration. A personal case study reported a child with Rett syndrome who had limited attention and total unconcern about herself or her surroundings. It was found that during a sustained period of music therapy, the child increasingly focused upon and responded to musical activities, and developed attention and concentration. This was particularly evident from the information gained from the use of records and documents, and reinforced by the class teacher's observations (Moore, 1996). The case study described by Postacchini
reported that music therapy sessions led to enhanced interest in objects, people and activity. However, it is difficult to measure the improvement in these cases and equally difficult to identify the relationship between cause and effect.

2.4.3.5 Other symptoms

Research has shown that breathing irregularity, including hyperventilation, is common in many girls with Rett syndrome, and can be particularly problematic when they are agitated (Lugaresi, Cirignotta and Montagna, 1985, Lewis and Wilson, 1996, Kerr, Southall and Samuels, 1992). The study by Postacchini (1993) described a girl whose breathing was laboured, and whose vocal expressions, when heard, were weak, subdued and hesitant. During music therapy sessions, improvement in the relationship with her therapist, coupled with her increased control of space, resulted in improved breathing and clearer voice quality.

Girls with Rett syndrome can often be miserable, unhappy, anxious, frustrated and tense (Lewis and Wilson, 1998). Music making and the ability to vocalise or create simple sounds can provide a medium for emotional expression. A skilled music therapist will organise a framework through which a child can communicate her anxiety and fear, as well as her happiness and joy (Wigram, 1995). Relaxation induced by music can facilitate a child's ability to respond in a variety of situations. 'Music therapy is very valuable to help a girl maintain contact with people and her environment, especially during irritable times' (Hanks, 1990, p.160). The benefits in a school environment and for children with similar disorders are not difficult to see.

2.4.4 Organisation of music therapy

Music therapy has been successfully carried out in group and individual sessions. By adopting a supportive role and providing a 'safe' environment, individual therapy aims to build trust and confidence, develop skills of listening and attention and enrich non-verbal communication (Association of Professional Music Therapists, 1992).

Group music therapy allows the therapist to create musical situations that aim to meet as far as possible the therapeutic needs of the individuals in the group and of the group as a whole. Many children are isolated by their individual disabilities (Fairfields School, 1995). Group therapy can instil personal confidence in a group situation, as well as provide opportunities for observing the functioning of an
individual child within a group (Association of Professional Music Therapists, 1992). In group sessions, children should be grouped according to their therapeutic needs and compatibility, not necessarily with their class group, with subsequent timetabling implications.

There is a place for both the individual and group approach within the school system, although the reality may be that financial constraints will prevent individual therapy in favour of the more economic group sessions, regardless of the individual needs of children.

2.4.5 The role of the music therapist

Bunt (1994) suggests that ‘Music is an all-embracing form, providing a rich source of information that can be viewed from a range of different standpoints’ (p.20) and believes that different professionals, with different viewpoints, can work as a team as part of the assessment procedure for individuals. Video recordings could be viewed and evaluated by members of the team, and used to allow parents the opportunity to observe sessions.

Wigram and Weekes (1985) believe that appropriate use of music can encourage mobility and thus reduce the risk of deformity. They advocate a joint treatment approach, neither specifically music therapy nor physiotherapy, but using the expertise unique to each profession. They suggest that physiotherapists should learn and understand the rhythmic influences in movement and that music therapists should learn and understand the physical and anatomical problems of children, in order that they may design the musical input appropriately. Rhythmic intention can be particularly effective for girls with Rett syndrome (Lewis and Wilson, 1998) and there appears to be an obvious potential progression from music to movement education. The overlapping of the two therapies could enhance the effect of both, and lead on to a combined approach with benefit to the child and favourable cost implications for schools.

2.4.6 Summary

A number of case studies have been cited here to illustrate the benefits of music therapy to a child suffering from Rett syndrome although they have not shown whether these effects are lasting. The individual goals of a student will determine which musical experiences will be most effective from a therapeutic standpoint.
The student's problems and special needs will be related to the basic characteristics of the musical experience and its potential effects on her. Music therapy sessions are designed to take advantage of the innate tendencies of all human beings to make music at their own developmental levels (Bruscia, 1991). The therapist actively encourages the release of emotions enabling the child to share and work through her feelings with the therapist.

Wigram (1991) emphasises the need for flexibility and variety in the therapist's approach.

For any parent or therapist to come to terms with a child who has Rett's syndrome, the biggest challenge must be to overcome purposeless and manneristic activity, and to rechannel natural energy and need for human contact into an engagement at a therapeutic level that is productive, dynamic and satisfying.

(p.51)

This study looks at parents' perceptions of the value of therapies, and it will be seen that this varies, and it is not always possible to ascertain by what criteria judgement is made. Jordan, Jones and Murray (1998) cite Radhakishan (1991) who concluded that the literature on music therapy research showed consistent methodological weaknesses, principally the absence of a neutral control and the presence of observer bias, leading to reservations about reported benefits.

Music therapist themselves acknowledge the difficulty. ‘...due to the importance of the therapeutic relationship, and the subtle aspects of emotional expression and creativity developed during the course of treatment that scientific methods of research do not lend themselves well to measuring the efficacy of music therapy’ (Diamond, 1999).

If assessment and methods of record keeping are not comprehensive and reliable then music therapy runs the risk of seeming uncertain in its aims and subjective in its assessment. Because music therapy is not prescriptive, with direction and content substantially affected by the individual child, there can be no universal model for music therapy record keeping or standardised assessment procedures (Association of Professional Music Therapists, 1992) and benefits can be difficult to measure.
However, the literature does seem to suggest benefits to recipients that are similar to the aims of teachers in schools and there do seem to be good reasons for incorporating music therapy into a school curriculum. If the academic objectives of the National Curriculum are not addressed directly, the therapy may open up ways of providing pathways to objectives. If the therapy provides access to forms of education, then the child is ethically as much entitled to receive the therapy as the education.

2.5 Hydrotherapy

It is not known precisely when hydrotherapy was first used therapeutically, but Reid Campion (1997) suggests that it could date back to 2400 BC. Today it is recognised as ‘the performance of physiotherapy exercises in warm water, combined with therapeutic and recreational aspects’ (Lewis and Wilson, 1998). Kerr (1995b) believes hydrotherapy to be one of the most essential therapies for girls with Rett syndrome and that daily hydrotherapy with good physiotherapy support should be one of the core facilities.

2.5.1 Hydrodynamic principles

Reid Campion (1997) describes hydrodynamic principles in a therapeutic perspective. Hydrotherapy makes use of hydrodynamic principles to modify movement processes of the body. Buoyancy, first described in physical terms by Archimedes, is the best known effect. In water, gravity is opposed by buoyancy to the extent that a young child can float and appear weightless. When standing in shallow water, the forces needed to support the body in an upright posture are reduced in proportion to the depth. Muscular forces needed for body support are less in water, but the energy required to move is normally greater due to fluid displacement and turbulence. Turbulence and water flow can also be used to assist movement.

The human body is neither uniform in density nor symmetric, whereas the effect of buoyancy acts equally on the body surface. The effect is to create an imbalance that turns or rotates the immersed and unrestrained body towards a position of equilibrium. Resistance to displacement normally has the effect of slowing voluntary and involuntary movement, and a combination of these factors can be used to develop balance and co-ordination.
Anticipation or anxiety is common among children preparing to enter the pool, and there is opportunity for the therapist to build relationships before and during therapy sessions (Reid Campion, 1997). If parents assist in a school situation, there is also an ideal opportunity for parent/therapist relationships to develop.

The Halliwick method of teaching swimming was devised in 1949 (Martin, 1981). Hydrodynamic principles and the scientific understanding of the human body in water form the basis of the method, which stresses ability instead of the disability which is apparent out of the water (Bell, 1993). Although initially devised as a method of teaching swimming, rather than as a therapeutic intervention, it has been used successfully with children with neurological disorders.

2.5.2 Treatment of symptoms

Just as physiotherapy can help with symptoms other than movement, so has hydrotherapy been found to be beneficial in several areas.

2.5.2.1 Movement

Girls with Rett syndrome are particularly liable to lose muscle bulk, strength and flexibility because the brain is not capable of using the existing nerve pathways in a normal fashion. This leads to poor internal nutrition of the tissues (Kerr, 1995b). Health of muscle and nerve requires active movement, but the movement repertoire available to the Rett child in the air is very restricted and becomes increasingly so in later years. Consequently, the problem cannot be overcome by daily physiotherapy alone, since only passive movement can be achieved in this way. Kerr believes that it is only in water that the fullest range of movements can be achieved and that as other forms of mobility deteriorate, it may only be movement in water that remains possible.

Because Rett syndrome produces a movement disorder that deprives the individual of the freedom to plan and carry out voluntary movement, walking is never secure, and is sometimes impossible. Girls with Rett syndrome may experience discomfort due to increased tone, voluntary tightness or actual muscle spasm. The difficulties which lead to the movement problem include fear which seems to be induced by perceptual difficulties when a girl find herself balanced some distance above the ground (Kerr, 1995b). Proximity to the surface of the water and its buoyant support of her limbs appear to diminish this fear, allowing her to find confidence in walking
on the base of the pool, and controlling her movements, even if she is not actually swimming. By assisting in supporting the child, the water can help with standing and gait re-education (Reid Campion, 1997).

2.5.3 Other symptoms

Kerr (1992b) suggests that girls with Rett syndrome should be offered opportunities for personal communication through touch. Kerr (1997) considered that the richest communication is likely to be through means other than speech, and suggests that a girl’s enjoyment of personal contact could be one way of encouraging interaction. By providing one-to-one support for the child, in a secure situation, hydrotherapy may provide the catalyst for the beginnings of communication.

The warmth and security of the hydrotherapy pool can help reduce the supportive criterion of breathing irregularity (Kerr, 1995b), which may in turn allow communication skills to develop. Cold and noise may cause fear and withdrawal in girls with Rett syndrome and exacerbate the involuntary disorder of movement, including hand stereotypies and breathing irregularities. The warm, quiet and well supported position provided by the hydrotherapy pool can reduce these symptoms and allow active learning to take place (Kerr, 1995b).

Distressing behavioural and emotional symptoms of girls with Rett syndrome, although not essential criteria, are common. Hanks (1986) and Lewis and Wilson (1998) found that hydrotherapy alleviates the stress, fear, anxiety and muscle spasm that may create tension. On the other hand, Reid Campion (1997) has found that fear or anxiety about water can create tension and suggests that musical sounds can be used to diminish these fears. Personal experience has shown that hydrotherapy is one of the most valued therapies for children with severe disabilities. Parents regard it as a means of providing therapeutic and learning experiences, and they also see it as a valuable recreational experience for children who experience few pleasures.

2.5.4 Organisation of hydrotherapy

Reid Campion (1997) believes that group hydrotherapy sessions, with children made up of differing levels of ability, can be beneficial, by improving attention and concentration span, as well as allowing group members to be educationally stimulated by each other. She accepts that if only children with severe difficulties
are grouped together, interaction will be poor and may inhibit progress. Some considerable skill, then, will be necessary to group children in such a way that the most severely disabled do not benefit at the expense of others.

The Halliwick method of swimming relies totally on one-to-one support, and aims to enhance breath-control, relaxation and balance. It does, however, use group dynamics and games for motivation, and would appear to offer the joint benefits of individual combined with group therapy. 'The opportunity to work on a one-to-one basis with a physiotherapist within a group provides the optimal situation for children' (Reid Campion, 1997 p.34). This is unlikely to be a realistic option because of costs, but by encouraging and using parental involvement during hydrotherapy sessions, schools can make the very most of the opportunity to help individual children.

2.5.5 Summary

Reid Campion (1997) believes that it is the combined properties of buoyancy and turbulence offered by water that enable the disabled child to move with greater freedom, and to execute movement which would be impossible on land. Hydrotherapy supplies the support of water deep enough to stand and broad enough to swim, in a warm quiet environment and under the care and guidance of the physiotherapist. The water assists the movements of girls with Rett syndrome as no other process can and Kerr (1995b) suggests that daily hydrotherapy with good physiotherapy support should be one of the core facilities for girls of all ages, and that it is one of the most essential of the physical therapies for this group of people.

Hydrotherapy provides benefits for the whole child, and is able to focus jointly on physical and emotional wellbeing of the child. In order to offer daily sessions, there must be access to a warm water pool. This places limitations on many schools, and could cause a special school placement to be the most appropriate for a girl with Rett syndrome.

2.6 Occupational therapy

'Occupational therapy is always based on purposeful activity, which is relevant to the client's lifestyle, physical and mental capacities and ... disability' (Penso, 1987, p.9). This therapy has been described as the treatment of physical and psychiatric
conditions through specific activities in order to help people reach their maximum level of function and independence in all aspects of family life (British Association of Occupational Therapists, 1982). Evaluation of the child's abilities and disabilities enables the therapist to determine what intervention is needed (Kramer and Hinojosa, 1993).

2.6.1 Benefits

Occupational therapy can improve all aspects of childhood by facilitating self-care at a level appropriate to the child's physical abilities and mental development, by devising and adapting play material to promote symbolic, imaginative, cognitive, constructional and manual skills and by suggesting suitable equipment to facilitate formal education (Penso, 1987).

2.6.1.1 Mobility

Mobility is likely to be impaired in girls with Rett syndrome, and some form of aid is likely to become essential for the child and the family. Both the nature of the specific disability and the process of development and growth must be constantly assessed and re-assessed (Kennedy, 1984). This need may be addressed by the physiotherapist, especially if she is based in a school environment, but it is likely that the expertise of the occupational therapist will be sought at some point, and in schools, the two disciplines should work together.

2.6.1.2 Hand function

Lewis and Wilson (1998) believe that 'In the planning and carrying out of programmes to increase the hand use of a person with Rett Syndrome, the occupational therapist has a vital part to play' (p.45). Current research issues on the advantages and disadvantages of the practice of splinting and confining the arms and hands have been discussed in Section 2.2.3.2. Cass (1994) suggests that a planned hand use programme would appear to present a possible way forward.

2.6.2 Equipment

Assessment of the need and provision of equipment may be shared by a number of professionals, with physiotherapist and occupational therapists most likely to be involved in the provision of equipment (Kennedy, 1984). Furniture, toilet facilities,
eating and drinking utensils, wheelchairs and even play apparatus will come under
the jurisdiction of the occupational therapist.

The knowledge and expertise of the occupational therapist may be needed to find
individual solutions and aids in relation to Rett girls’ specific difficulties, which affect
more than one aspect of life. Hand function and levels of perception need to be
examined, as well as equipment necessary to daily living skills. Penso (1987)
believes that ‘For the severely impaired child a proportion of occupational therapy
will be directed towards the parents or carers to advise on handling techniques and
the best methods of carrying out the child’s daily care’ (p.13).

It would seem logical that the occupational therapist, as well as other therapists,
should be involved in such issues as planning the physical attributes of the
classroom, even the school, where profoundly disabled children are placed. The
direction and form of a child's day is very much influenced by her condition, and by
her status within that condition on a given day, so the importance of planning right
down to the day to day activities and the daily routine of attendance at school
should not be underestimated.

2.7 Other therapies

There are other therapies which may be considered beneficial, and it has been
found that some parents are prepared to try alternative forms of therapy in an
attempt to improve the quality of life for their daughters (Gorman, 1998). A study by
Lewis and Wilson (1998) found that hippotherapy, aromatherapy, reflexology,
rebound therapy and cranial osteopathy were received by some girls with Rett
syndrome, although the sample size was too small to allow statistical support.

Hippotherapy, aromatherapy and cranial osteopathy were the only peripheral
therapies to be mentioned by parents in this study, and these are considered briefly
in the following sections.

2.7.1 Hippotherapy

By the 1950s, British physiotherapists were exploring the possibilities of horseback
riding as therapy, and the British Riding for the Disabled Association was founded
in 1969 (Riding for the Disabled Association, undated). Hippotherapy is the use of
the horse as a treatment modality with a patient not a rider (Association of
Chartered Physiotherapists in Riding for the Disabled, undated) and is carried out
on a one-to-one basis by a physiotherapist trained in this field. The aim of hippotherapy is the unimpeded transfer of movement from the horse’s back to the patient and the patient’s adaptation to that movement. Bream and Spangler (1997) offer a definition which, while directed primarily at the benefits of horseback riding, applies equally well to almost any form of therapeutic intervention. ‘The aim of any treatment program is to improve circulation, respiration, balance, co-ordination, proprioception, agility, self confidence and mental relaxation. In addition, one would hope for significant carry-over of improvements from the therapy sessions to activities of daily living’ (p. 1).

Hippotherapy has now become well established and is often sought as a therapy by parents of girls with Rett syndrome (Lewis and Wilson, 1998). Research has found that stereotypic hand movements have diminished after sessions of horseback riding, because purposeful hand movements are encouraged (Lindberg, 1994).

In addition to physical benefits, horseback riding can improve inter-personal relationships between the child and her helpers, as well as promoting a positive self-image and enhancing self-confidence (Bream and Spangler, 1997). Clearly, very few schools can offer hippotherapy on site, and the expense of transport added to the expense of providing suitable horses and adequate one-to-one (or even two to one) supervision makes this one of the less common therapies. There is also a paucity of serious scientific analysis of the outcomes, and while there is some evidence of value, the therapy cannot rank alongside those already discussed in terms of school based therapeutic intervention.

2.7.2 Aromatherapy

Aromatherapy is an ancient art using the essential oils of plants to enhance mental and physical wellbeing (Plucknett, 1995). It has been suggested that aromatherapy will ‘foster a sense of wellbeing and self-esteem, an important part of her Personal and Social Education Curriculum’ (Lewis and Wilson, 1996, p. 101) as well as providing a leisure activity for older girls.

Although a complementary therapy in the education system, Annis and Sillitoe (1995) believe that it is important to treat aromatherapy and massage as a serious part of the curriculum for some pupils and feel that knowledge and training could help dispel the mistrust which exists around this therapy. They consider that
massage is beneficial for both the physically and mentally disabled child, and can create an excellent bond between carer and child. Some components of PNF techniques can be successfully integrated into massage, so combining physiotherapy benefits with those of aromatherapy. In an educational context, the bond may be of limited value if the carer is not active in the classroom. It is difficult to envisage the class teacher taking on a therapist's role in this sense, especially considering that the teacher is responsible for other children, and has other educational expectations.

Constipation can be a very real problem for many girls with Rett syndrome (Rice, undated) and massage with essential oils, given by a qualified aromatherapist, can relieve the symptoms and help with restlessness and nervous tension (Annis, undated). There may be peripheral benefits for schooling, but again, it is harder to see them in an educational context.

2.7.3 Cranial osteopathy

Osteopathy is a system of health care based on the theory that the body is capable of making its own remedies against disease (Oldham, 1999). Cranial osteopathic treatment involves hands-on manipulation to gently free areas of the body that have restricted motion. It involves cradling the head and using gentle pressures to free movements of restricted bones or other tissues (Remington and Moore, 1999).

There is no real research published regarding the effect of cranial osteopathy on a child's ability to advance socially and educationally in school, and there appears to be a somewhat lesser credibility regarding the self-healing concept. More research is required before realistic assessments of this therapy can be made.

2.8 Conclusions

It has been seen that it is not enough to look at the behaviours of children within the autistic spectrum in order to teach a child with autism (Jordan, 1999a). In referring specifically to Rett syndrome, it has been noted that: 'Therapists must have an understanding of the complex problems associated with the four stages of the condition in order to develop appropriate treatment plans' (Lieb-Lundell, 1988, p.S34). There are many rare medical conditions, each with their own idiosyncrasies and it is unlikely that the full implications of the complex problems associated with
Rett syndrome are fully appreciated by the average therapist, whatever their discipline.

The literature relating to the therapies involved in the treatment of Rett syndrome indicates that positive benefits can be attained by the application of these therapies although little reference is made to the lasting effects of treatment. It is not always easy to categorise a condition or symptom into an area of therapy. Although many of the benefits of a specific therapy are logical - few people would dispute the benefits of physiotherapy in the area of motor disability - others, such as the use of music therapy to reduce stereotypical hand movements (Schulz, 1987) are less obvious. Nutrition has been described as ‘the cornerstone by which all other forms of therapies are made possible’ (Rice, undated). Difficulties with feeding and other oral reflexes may come under the jurisdiction of physiotherapy, but since the problems have considerable implications for speech, liaison with the speech therapist is also very important (Eckersley, 1993). There are other examples of the benefits of professionals working in unison, such as the joint physiotherapy and music therapy approach advocated by Wigram and Weekes (1985). Close liaison by the occupational therapist, with the physiotherapist and speech therapist in particular, will ensure that technical aids are adapted to the personal needs of each child.

Shepherd (1980) recommends a complete link-up between the various people responsible for the child’s treatment and suggests that the best approach is one in which all therapists are trained to have a similar approach to treatment, and also to have an understanding of each other’s work so there can be continuity in a practical sense. It may even be possible for two professionals to share their expertise, whilst working together with a child. ‘The speech therapist, for example, may encourage vocalization and stimulate more normal oral function during those times when the physiotherapist has facilitated more stable head control’ (Shepherd, 1980, p.113).

The school would seem an obvious place in which to share therapies, therapists, and therapy rooms, and for those special schools serving the most physically disabled children, therapy and withdrawal from the classroom are a major part of school life. There is a need to co-ordinate therapies to ensure maximum benefit for the children, and success will rely to a large extent upon day-to-day management processes within a particular school. Personal experience suggests that this will be
influenced by the individual personalities of headteachers and senior management staff, as well as the competence of individual therapists.

For the Rett child in the UK, therapies are administered mostly in school time, and are normally funded by the Local Education Authority or Health Authority, within the framework of the National Curriculum (DFE, 1994). Local issues may affect the choice and quantity of therapies; indeed, opinions of individual officers within the Authority may influence choices. What is important is the notion that these therapies are at the beginning of a life that will be dependent upon different forms of intervention. Their effects may not be fully understood, in the long term at least, and there is a need to assess and monitor the epidemiology of Rett syndrome in relation to applied therapies so that the best options are available for newly diagnosed children and for young Rett adults as they grow into later life. 'In syndromes of this kind the statement 'There is no treatment' is unacceptable: no specific treatment maybe, but there is much that can be done to help the patient and the family' (Hagberg, 1993, p.x).

There is only one place where all the common features of life skills are brought together on a regular basis. The school is where children learn to cope, where they assimilate knowledge, where they learn new skills and become competent in new activities. For the girl with Rett syndrome, it is where she should find the calm which allows her awareness, where the personal bonds which touch her everyday life outside the family are formed, where she learns to cope with stereotypies, and where some of the most beneficial therapies take place.
Chapter 3 Methodology

3.1 Rationale

It has been seen that Rett syndrome is an uncommon condition occupying a narrow position within the broader spectrum of autistic disorders. The neurology is complex and not fully described, and theoretical support for the various therapies is complicated. Personal experience suggests that parents are equally, if not more concerned with the outcome of intervention than with the complexity that underlies the theory, and the practical element of this research concentrates on parents' views of the values of individual therapies.

My first contact with Rett syndrome was in 1986, when I first met and taught a girl with this condition. I have since taught other girls, both in the United Kingdom and Australia. The research in this study stems from the realisation that there are many inconsistencies in the way in which the needs of girls with Rett syndrome are met in education. Personal contact with parents reiterated my intuitive thoughts that traditional methods of education may not be wholly appropriate for children displaying such debilitating and complex symptoms. Rett syndrome is unique and puzzling in its presentation, and while collectively the intellectual and physical manifestations comprise a mix not found in any other condition, each feature taken alone may not be exclusively specific to the syndrome. Differential diagnosis regarding Rett syndrome and childhood autism, both pervasive developmental disorders, can be found in Section 1.5.6. Other disabling conditions present similar difficulties for education, and evoke similar emotions in parents and carers. This study, while concentrating upon the Rett enigma, will have obvious implications for other syndromes and conditions within the British education system.

Teachers have stressed the importance of meeting individual needs through such means as programmes of therapy, the development of basic skills and the promotion of 'life skills' at all levels (National Curriculum Council, 1993) and the differences and similarities between education and therapy are discussed in Chapter 1. It is parents who have the best opportunity for consistent and close observation of their children, and consequently they may be most suitably placed to appreciate the effects of the various treatments in terms of living skills. The National Curriculum has little to say regarding the specific needs of children with complex prognoses, although it does accept that 'for pupils with profound and
multiple learning difficulties therapies can be an essential element of the total curriculum' (National Curriculum Council, 1993, p.3).

Since the acquisition of essential life skills normally precedes academic learning, it is the special treatments administered to girls with Rett syndrome that provide the platform for abilities in later life. Whilst research on the value of therapies with specific reference to Rett syndrome is sparse, recent work appears to indicate that therapeutic intervention can have a critical effect on minimising the effects of this disability (Hanks, 1990, Glaze, 1995, Budden, 1995, Kerr, 1995b and Lewis and Wilson, 1998).

It is known that therapies of various kinds are delivered in schools, but there is no central control, and no co-ordinated mechanism for assessing results. Therapies sometimes appear to be given on an ad-hoc basis, depending on geographic location and financial circumstances of individual local education authorities, or even individual schools, and there is no guarantee that they will be offered at all, regardless of the individual need of the child. Personal experience has shown that if therapies are provided, it is likely to be at the expense of 'time-out' from regular classroom activities, and so it is important to know the comparative value in terms of early educational achievement.

3.2 Research methods

Whilst quantitative researchers use scientific techniques to produce measurable conclusions, researchers adopting a qualitative perspective are more concerned with an understanding of individuals’ perceptions of the world (Bell, 1996).

The data collected in this study represent the views of parents with regard to their children with Rett syndrome, and as such are subjective rather than objective. The measurements were given numerical labels to turn them into quantity-like form. The questionnaire data were subjected to quantitative analytical techniques using either descriptive or complex statistics, and the results are presented in Chapter 4 and discussed in Chapter 5.

In Chapter 6 the subjective views of the parents were in part analysed with quantitative and in part with qualitative methodology. Quantitative methods were used to analyse the interview data, which were counted, collated and tabulated. Open-ended questions were used to collect qualitative data and to allow greater
flexibility of response. This aspect of the study identified themes within the data, including differences and similarities between views of parents. Parents’ comments were used when appropriate to illustrate commonly held views. It is acknowledged that the decision to categorise the parents’ views in particular ways were subjective on the part of the researcher.

3.3 Research questions

This research is in the nature of a market survey, and so strength of opinion is the major outcome measure. In order to supplement classroom experience, and to test professional intuition, it was decided to gather parents’ perceptions of the difficulties encountered by their daughters with Rett syndrome, and to ask the question ‘Do parents demonstrate preferences in relation to therapeutic management?’ A survey instrument in the form of a questionnaire was designed to ask the following questions:

1. How severely do parents perceive their children to be affected by Rett syndrome compared to children without a disability?

2. How important do parents think it is for their children to improve in specific areas?

3. Which therapies are commonly used in schools, within the age group 7 to 12 years?

4. Is provision for therapies included on the statement of Special Educational Needs, and does this make a difference to provision?

5. How are therapies organised in schools?

6. How long have children been receiving therapies?

7. How much therapy do children receive?

8. How valuable do parents perceive each therapy to be?

The questions were grouped into three sections. Section One related to the present ability of the child compared to a child without Rett syndrome, and the importance of addressing main areas of disability. Section Two referred to the
therapies themselves, and Section Three to the parents' perception of the value of each therapy.

3.3.1 Search of the literature

A search of existing literature was carried out to identify current research and it confirmed that little work had been undertaken on the value of therapies with specific reference to Rett syndrome. This search was conducted using traditional methods, leading to an examination of medical journals, educational publications, internet publications, and resources provided by several agencies dedicated to the support of Rett syndrome girls and their families.

The aim of the literature review was to establish the knowledge base for Rett syndrome and to view this disability in the context of other general issues that influence management, such as parent attitudes and medical and educational models of philosophy and practice.

Personal contact was made with leading researchers in England, Australia and Sweden and the Rett Syndrome Association United Kingdom provided access to the database of girls who were suffering from the syndrome.

3.3.2 The survey

It was realised from the outset that the only practicable method of gathering information would be by using a postal questionnaire to target parents on the RSAUK database, and so a survey instrument was designed to collect primary information.

3.3.3 Survey population

During 1997, questionnaires (Appendix IV) were supplied to parents of 100 girls with Rett syndrome. In order to ensure anonymity of Rett families, the RSAUK agreed to administer the postal distribution of the questionnaires, and so the geographical location of the children is not known, other than that they are located in the United Kingdom. The issue of anonymity is important, and will be discussed later in this chapter. It was anticipated that approximately one half of the 100 questionnaires would be completed, and the low incidence of Rett syndrome meant that the families would probably be spread thinly throughout the whole country. Girls in the group chosen for study were aged between 7 years and 12 years, when
diagnosis was likely to have been confirmed and educational programmes, including therapeutic intervention, established.

3.3.3.1 Pilot study

The purpose of the pilot study was to identify which therapies were commonly used to support the education of children with Rett syndrome, to test a prototype survey instrument, and to enable the method of analysis to be tested before the main survey was carried out.

It was important to know that the questionnaire was socially acceptable to parents, and politically acceptable to RSAUK, since this element of the study relied upon the co-operation of the Association. Prior to the main survey, a pilot study was carried out with members of the RSAUK committee, who have daughters with Rett syndrome, and with other known families. Ten families agreed to complete a draft questionnaire and then offer critical appraisal of content and style. This led, through a discussion of the process through parents' eyes, to the final version of the questionnaire, with minimal changes to style only being considered necessary (see Appendix V).

3.3.4 Survey instrument

The advantage of using a printed questionnaire is that the maximum number of subjects can be invited to participate in a pressure-free way, allowing structured data to be gathered and at the same time allowing opportunity for additional personal opinions to be expressed. The greatest disadvantage is that a significant proportion of the target group may not respond, and those who do respond may not be truly representative if the sample is small (Bell, 1996). A more subtle disadvantage is that the questioning process may modify expressed opinions, and questionnaire design is therefore very important (Borg and Gall, 1993).

Johnson (1994) suggests that the use of a questionnaire can direct the thinking of others in relation to their own experiences, and consequently many low level surveys are seriously flawed because on one hand the very act of questioning subconsciously alters opinions, and on the other, it frequently does not give an opportunity for the full range of responses to be expressed.

Given the limitations of the printed questionnaire, it was decided that this research would seek a minimum of demographic data, but would ask for opinions regarding
the severity of disability and the value of the most common therapies. It was suspected that there would be cases where unconventional therapies were delivered, so the opportunity to assess unlisted treatments was provided.

An important element of the study embraces the opinions of parents regarding values of specific therapies. It was expected that two parents would have different perceptions of the value of the same intervention, because it is unlikely that the parents will have identical experience of their daughters' lives. There is some difficulty with objectively evaluating perceptions, feelings and opinions, particularly where the enquiry relates to the effects of processes that are not necessarily understood by the research subjects, and applied to others who are unable themselves to report how they feel. Important factors regarding perception may be hard to quantify, and because techniques for structuring such qualitative information are not well known, the information may not even be collected (Cameron, 1994). In this research, the source data (opinions and feelings) are subjective but have been quantified in order to facilitate comparison. Analysis in this study was kept simple, and was restricted to examination of differences and basic interactions between variables.

There are ways other than point scales for assessing subjective opinion (for example, the fuzzy logic style of Hesketh, 1989) but the trade-off is added complexity for the respondent, and consequent loss of responses.

It was decided that the value of graphic rating scales might be compromised by the investment in time and concentration demanded of the respondent, and as such it was decided to use a method similar to the more familiar Likert point scales for indicating the strength of parents' opinions. A conscious decision to exclude 'don't know' categories was made early in the design, because it was felt that everyone would have an opinion of some kind in each area, and that to allow uncertainty would in a sense encourage it. In the event, a very small number of blanks suggested that a few respondents found some difficulty in allocating scores, or did not actually know the circumstances of their daughter's therapy.

3.3.5 Survey Procedure

Although it has been said that the postal questionnaire is frequently the best form of survey in educational enquiry (Cohen and Manion, 1994) research has shown that
a high percentage of postal questionnaires are not completed or returned (Bell, 1996). A short covering letter from the RSAUK was sent out with the questionnaire (Appendix IV) which outlined the aim of the survey, assured the respondents of confidentiality, and encouraged parents to complete and return questionnaires. In spite of this support, there was only a 30% response. The RSAUK did not support the distribution of follow-up letters to remind parents to return their questionnaires, as they felt it would be an additional pressure for parents.

3.3.5.1 Questionnaire

The questionnaire was designed to acquire the following:

a: simple demographics:

Date of birth, age at diagnosis

b: severity of disability related to specific areas of learning

i) Perceived effect of Rett syndrome, measured in four areas, communication, fine motor ability (functional hand use), gross motor ability and learning ability

These four areas were derived from an examination of the necessary, supportive and exclusion criteria (Hagberg, 1993). Only those disabilities relating to the current educational needs of the child were included in the study. Whilst accepting that supportive criteria contribute to the overall picture of individual children, it was considered inappropriate to include those features not common to all girls. Only the following conditions are known to be significant to all sufferers of Rett syndrome.

* communication difficulties

* loss of functional hand use (fine motor ability)

* poor motor skills (gross motor)

* learning difficulties

Personal experience has shown that parents are not always comfortable with the difference between fine and gross motor skills. Loss of functional hand use is an important feature of Rett syndrome, and although hand use is an aspect of motor ability, it was decided to look at the two areas separately.
ii) Importance of improving in each of the four areas

In the case of severely disabled children it may be possible to discern improvement or deterioration in ability, but to quantify those changes requires regular objective assessment designed for the particular situation. On the one hand, periodic objective testing by detached professionals may identify interval trends, whereas everyday intimate contact of parents and carers may allow a greater intuitive understanding. The child’s responses may differ day to day, so periodic testing can be influenced by the particular manifestation of the condition on a given day, whereas changes may not be observed by the carer if they occur gradually over a period of time.

In this kind of study, it is very difficult to measure any kind of improvement, or to gain access to stored information that could offer some objective indications of change. In the case of Rett syndrome, there is a possible interaction between different therapies used, adding to the complexity of analysis and so for the purpose of this study, perceptions of the effects of therapies were sought. The views of teachers are often coloured by external factors – including the need to ‘prove’ that the children are making academic progress and so it was decided to seek the views of parents or primary carers. It is possible that the views of parents, too, will be equally coloured, whether by a desperate wish for improvement or by denial of the severity of disability.

The difficulty for this study is that on one hand parents and carers are well placed to judge effects of treatments and events because of their intimate closeness to the condition, but on the other, their emotions, their love of the child and desire for a ‘cure’ are not likely to be objective, and neither are their responses. The questionnaire was designed to force an objectivity in some areas, while leaving free expression in others, even though it was felt that some participants would be dissuaded from responding because of the difficulty in giving values to perceptions. It may never be possible to reconcile both aspects in this type of study, but the problem is at least recognised.

c: effect of therapies, related to the four areas of disability

In the questionnaire the main therapies suggested by leading researchers in the field of Rett syndrome (Percy and Hagberg, 1993, Van Acker, 1991) were listed in
relation to each of the four ability areas. Hunter (1987) pointed out the need for all parents of Rett children to be allowed to talk about their child, and so an opportunity was provided for additional therapies to be considered and discussed.

The effect of a therapy and the relationship between therapies can be influenced by:

a. age of child

b. severity / complexity of disability

c. length of time therapy has been used

d. intensity of therapy

e. the skill of the therapist

This last area was not addressed. Lay carers may not have the opportunity or the ability to judge the skill of professional therapists and it was felt that this issue was beyond the scope of this study.

3.3.5.2 Measurement

The aim of the study was to measure the perceived values of therapy as viewed by the parents since it was not possible to acquire objective measures. There is then the immediate problem of understanding what actually is being measured, and the different ways in which individuals may see things. To address this issue, the scales used on the questionnaire were kept relatively simple and consistent.

It is the views of parents that are being examined in this study. These comprise parents’ perception of how well a child responds to the real effect of the therapy, an adjustment of perception due to the actual presence of the therapy (Hawthorne type effect) and the influence of ‘other non-treatment variables such as the enthusiasm and dedication of those carrying out the programme’, as well as the support given by professionals (Jordan, Jones and Murray, 1998, p.129). It is suspected that the therapists’ influence may even moderate that of the therapy - in effect a ‘poor’ therapy delivered by a ‘good’ therapist would have a greater perceived benefit than a ‘good’ therapy delivered by a ‘poor’ therapist.
We can give a child to three therapists; one with the best resources; one with so-so resources and one who says nothing can be done. In my opinion (after 35 years of working with the handicapped), the child feels better than any other that "yes, this therapist likes me and accepts me" and, if she feels this, then she will work much better and doesn't cry.

(Rett, 1985b, p.3)

Rather than an evaluation of an applied therapy in general, this study examines a series of individual cases, each influenced by a variety of factors which are acknowledged but which cannot at this point be separated. Opinions may be 'contaminated', but they might be all a parent has to offer and because they do represent the total expression of experience, they must be considered worthwhile.

3.3.6 Anonymity

While it may be preferable to know the identity of the respondent, it is established practice in educational surveys to allow anonymity. It was felt to be more important to receive an honest response than an identified response, and it was expected that some parents might be critical of some features of their daughter's treatment. This kind of measurement cannot be verified in the same manner as can objective recording, and educational / sociological research tends not to be subject to the same techniques. It was consequently decided to offer elective anonymity. Although this meant that only those people who offered their names could be chosen for further study, it was hoped that the offer of anonymity would encourage a greater response.

3.3.7 Interviews

Initial analysis of the questionnaires clearly showed which therapies were available to Rett girls in school, and it was decided to interview only parents of children who were receiving the four most common therapies – physiotherapy, music therapy, hydrotherapy and speech therapy. The instances of other therapies being used were so low as to be insignificant. Consequently, the interview method was used with eight parents to gather additional data and extend the scope of the study.

An interview differs from a questionnaire, by allowing data to be gathered through direct verbal interaction between individuals. It was felt that this direct verbal interaction would allow far greater depth than a questionnaire, but it was recognised that it may be prone to subjectivity and bias on the part of the interviewer. With this
in mind, it was decided to control the situation by using a focused approach (Cohen and Manion, 1994). Previously completed questionnaires were used as a starting point for planning the interview schedule, and provided guidance during prior analysis in the most appropriate direction.

All eight interviews were conducted over the telephone, and these formed the basis of Chapter 6 (Appendix VI). Although these eight interviews may not represent a typical sample, they do have some common ground: all the children involved received a minimum of the four main therapies and were aged between 7 and 12 years. 30 parents responded to the initial questionnaire. Of the eleven families who met the selective criteria for the follow-up interview, three declined to give their names, so could not be approached for interview. It was felt that eight was an adequate number to provide additional data.

3.4 Statistical testing

The data returned in the completed questionnaires were of categorical or ordinal class, with the exception of those responses that related to periods of time. In effect, these data too were reduced to ordinal class during analysis. The Wilcoxon test was considered the most appropriate for use when examining differences between variables, and Spearman's correlation coefficient was used when examining variables which were likely to be interactive. The Mann Whitney and Chi-square tests were used to examine differences occurring in single variables across the sample in relation to other influencing factors.

Throughout this analysis, it was decided where possible to look for statistically significant differences in perceived values, and it was necessary to conduct tests against a statistical hypothesis in each case. The hypotheses used were not derived from published literary opinions, but were of a simplistic nature, generally of the type that assumed no differences between variables. For all the tests, the critical value for $p$ was taken to be 0.05 and values less than this (shown in bold typeface) permitted rejection of the null hypothesis. In each of the analyses examined, the null hypothesis is that there is no difference between each of the two sets of responses or that there is no association between variables.

The Wilcoxon test is a non-parametric test that makes no assumptions regarding distributions within the data, and is capable of dealing with ordinal data. The test
compares two sets of ordinal data obtained from the same (or matched) subjects and determines whether there is a significant difference between the medians (Hicks, 1990). This test was considered the most appropriate for use when examining differences between variables, and the null hypotheses that there are no differences between each of the two sets of responses were used to give some strength to the ranking procedure.

The Spearman test is another non-parametric test that indicates whether scores are positively or negatively correlated, and is suited to ordinal data from two sets of scores. It might be expected that associations exist between related variables such as those which represent parents’ perceptions of the nature of disability, how important it is to improve, and the benefit values offered by therapies. For example, it would be reasonable to anticipate that the perceived benefit value of a particular therapy might be influenced by the hours per month that the therapy was given. Spearman’s rank correlation ($r_s$) was used to test the strength of association between variables in these relevant areas.

Mann Whitney and Chi-square are also non-parametric tests that can be used to formally test for differences between observed proportions of independent samples, and to test for associations between two categorical or ordinal variables. It might, for example, realistically be expected that presence of a therapy on a statement of Special Educational needs would influence whether or not the therapy was actually delivered, and that the effectiveness of a particular therapy might be influenced by the organisation into groups or individual sessions.

3.4.1 Analysis

The questionnaire data were stored in a spreadsheet, and descriptive statistics were produced in tabular and graphic forms. An advanced statistical package was used to identify possible interaction between the perceived effects of therapies. A value $p<0.05$ was taken to be significant throughout the analysis. Low numbers limited the analysis. Less powerful non-parametric tests were used on data that were only of ordinal category. It was not possible to examine interactions in any depth because of the small sample size.

Cohen and Manion (1994) discuss two possibilities for coding and scoring interview data. One suggestion is that interview schedules are precoded, to enable the
interviewer to classify responses during the interviewing process. An alternative method is to postcode the interview data once they have been collected, and then subject it to content analysis. This was the method used in this research; the response data were subsequently counted and the results were collated and tabulated. The common threads appearing in the interview responses were identified and discussed. Where appropriate, comments from parents made on questionnaires and during interviews were used to illustrate clusters of opinion.

3.4.2 **Interpretation**

In this type of study, there is no manipulation of variables to establish causal relationships, and there is no requirement to query large statistically representative samples. The nature of the syndrome selected for this work is such that it is rare on a local basis, but there are large numbers of individual sufferers spread around the world. It may or may not be a reasonable assumption that the effects of therapies applied to one sufferer may be similar to the effects upon another, but the fact that the very nature of any therapy is that it is designed to benefit, and the nature of a medical syndrome is such that it places its sufferers into a similarity class, means that there is a level of probability that effects may be replicated. This is not an assumed conclusion, but a logical hypothesis that needs eventually to be tested in a wider study.

3.5 **Critical analysis of methodology**

The main issues relating to the methodology used in this study are appraised in this section, and a number of shortcomings have been highlighted. Research methods are always open to criticism, and whilst some of the difficulties were anticipated during the planning stages, others were not so evident, and were only identified as the research progressed.

3.5.1 **Questionnaire**

Although ideally a pilot study should be undertaken with respondents who would be eligible to take part in the main study (Johnson, 1994) the small incidence of Rett syndrome made this difficult, and not all children in the pilot study were within the age group selected for the main study. All the families in the pilot study were connected with the RSAUK and were approached in person, either by myself or by a member of the RSAUK committee. Consequently, they were all committed to the
project, and all questionnaires in the pilot study were fully answered and comment sections were completed. The pilot study did not, therefore pick up the fact that some parents would not know the answers to some of the questions, or may not be able to express a view about values of therapies.

Some questions assumed that parents would be fully aware of what was happening in their daughters’ schools. This was not always the case.

   My daughter has speech therapy but I don’t know if it is in a group or not and I do not know how long she has speech therapy, as I don’t see her speech therapist unless I need to. She makes a report about once a year on how she is doing.
   (Questionnaire 34)

This difficulty could have been addressed if contact had been made with individual schools, and this would have increased the scope of the research, although anonymity would have been compromised. Similarly, the theoretical premise of the therapy is not known, and contact with therapists would have been needed to include this aspect.

3.5.2 Statistical testing

The casual construction of the null hypotheses was only possible because of the small number of variables under consideration. In large-scale studies, such ad-hoc selection of hypothesis could lead to data overload, and in these cases, a proper construction of research hypotheses is critical. In retrospect, a more rigorous relationship should have been developed between the research questions asked and the hypotheses tested in this study.

3.5.3 Non-response

A survey conducted in a more personal manner may have encouraged a greater proportion of parents to respond, and although a personal appeal for participation at the RSAUK conference had some success, only a relatively small proportion of parents attended the conference; not all had children within the selected age group, and those that did had mostly responded already. It was not possible within the terms of reference of this study to make personal contact, but if this work is repeated, it is felt that a direct approach may be better. Although it may be difficult to maintain anonymity in a direct approach, the findings here suggest that most parents would not object to less rigorous anonymity.
The questionnaire was not thought by the parents in the pilot study to be difficult to complete, but with the knowledge of hindsight, a simpler version may have gained greater support. The layout was not unduly complicated, but when faced with two sides of tables and questions there may have been a dissuasive effect due to first impressions, or the intensely personal nature of Rett syndrome may have had a deterrent effect. Of the many disabilities encountered personally during a career in special education, Rett syndrome is characterised in parents by a strong group bonding which tends to set them apart from others.

Bell (1996) cites Moser and Kalton (1971, p.267) as reporting that 'non-response is a problem because of the likelihood - repeatedly confirmed in practice - that people who do not return questionnaires differ from those who do' (p.86). The parents who did respond may be those who were simply more outgoing and more prepared to share their experiences. The danger then is that the sample is virtually self selected on a personality basis, and may not represent a random selection.

3.5.4 Use of benchmarks

The inclusion of benchmarks by which to judge seriousness of disability and effect of therapies would have helped stabilise the overall results, but was difficult to achieve without making the questionnaire longer, and was not thought necessary by participants in the pilot study. There would have been more precision if parents had been asked separately about particular communicative functions, as contained in the Pre-verbal Communication Schedule, for example, but this would have been inappropriate without eliciting information about other areas of development, such as fine and gross motor functions. This would add considerably to the length of the questionnaire and could discourage parents from participating in the research. It would, however, have increased the rigour of the research and if this work were to be repeated, then the use of benchmarks should be seriously considered.

3.5.5 Population

The nature of Rett syndrome means that sufferers are widely dispersed throughout the country, therefore the effects due to geographical factors cannot be isolated within the data. For example, effects due to the skill factor of the individual therapist are unknown. A sample considerably larger than obtained here is needed to overcome these issues.
A wider age range would have captured more subjects and given a chance of more responses, but then additional problems associated with puberty and the progress of the syndrome become more influential.

It was not possible to include all families with Rett syndrome daughters in the survey. The RSAUK is the only national body with records of incidence of Rett syndrome, but it is probable that not all cases are included. However, it is difficult to see any other starting point for research.

Johnson (1994) suggests that the survey approach may not be suitable for research into sensitive issues because it does not have sufficient flexibility to enable the supportive atmosphere needed between the respondent and the researcher. It may be that only the more articulate parents felt able to complete the questionnaire; some parents may have resented a perceived intrusion into their personal life, others may have been unable to spare the time involved or simply be unwilling to help. Parents of children with disabilities have many pressures (Hunter, 1987) and even the most caring or willing parent may have been unable to cope with this additional demand on their time.

The RSAUK did not support the inclusion of personal questions as it was felt that these would be intrusive, and so information requesting socio-economic information was not sought. It has been seen that if the sample is not in fact representative of the 'identified population' then generalising from the survey findings can produce seriously biased statements (Johnson, 1994). Consequently, it is acknowledged that this sample may contain bias, and the results should accordingly be viewed with appropriate caution.

3.5.6 Interview data

There was a number of scoring procedures that could have been used to aid the content analysis of the interview data other than response counting. Because of the low number, response counting was considered the most appropriate at the time. In retrospect, more rigour could have been applied to the interview process and analysis by using a recognised coding technique, and a pilot interview would have been beneficial in identifying areas of interviewer bias.
3.6 Summary

The study seeks to answer a number of research questions relating to the ability of children with Rett syndrome, the importance of addressing main areas of disability, organisational aspects of therapies and parents' views of the value of therapy. Data were obtained using firstly a postal questionnaire distributed to 100 families and then telephone interviews with 8 families where the four most common therapies were given. The questionnaire data were stored in a spreadsheet, descriptive statistics were produced in tabular and graphic forms and the data were analysed using quantitative methodology. Interview data were response counted and common threads were identified and discussed.

The study aims to broaden knowledge relating to the status of Rett syndrome in British schools by reporting hitherto unpublished analysis of parental views regarding therapeutic management.
Chapter 4 Results of the survey

4.1 The survey

During 1997, questionnaires (Appendix IV) were supplied to parents of 100 girls with Rett syndrome and 30 completed responses were received. The following figures and tables show raw results only; interpretation and analysis are given in Chapter 5.

4.2 Numbers, ages, and diagnosis

At the time of the survey the average age of the girls was 9 years and 5 months, ranging from 7 years 5 months to 11 years 7 months, and the average age at diagnosis was 4 years and 2 months, ranging from 2 years to 10 years. Figure 1 shows the number of girls receiving each therapy.

Figure 1. Number of girls receiving each therapy
From the 30 responses, 11 cases were identified where girls received the 4 most common therapies, and from these the parents of 8 girls agreed to a telephone interview. Structured interviews were carried out later with these in order to acquire more information (Chapter 6).

Instances of hippotherapy (2), homeopathy (1) and osteopathy (1) were reported as isolated cases. The basic results of the other six therapies are shown here in graphical or tabular form.

4.3 Abilities

Parents were asked to give their views on how severely their child was affected by Rett syndrome and the importance of increasing proficiency in the areas of communication, functional hand use, motor skills and learning skills.

4.3.1 Severity of disability

Parents were asked to indicate how seriously they felt their child was affected in the four fundamental areas of disability of Rett syndrome. Figure 2 shows that the majority of parents chose the values of 4 and 5, indicating that the level of disability in all four skill areas was considered severe or moderately severe. Table 2 shows the average and distribution of scores.

![Graph showing parents' perceptions of the severity of disability in four skill areas](image)

**Figure 2. Parents' perceptions of the severity of disability in four skill areas.**
Most parents perceived their daughter to be severely affected in all four areas, with learning difficulties scoring an average 4.17. Communication was thought to be the least severely affected, scoring an average 3.60.

**Table 2. Parents' perceptions of severity of disability**

<table>
<thead>
<tr>
<th>Area</th>
<th>Score</th>
<th>Total</th>
<th>Average</th>
</tr>
</thead>
<tbody>
<tr>
<td>Communication</td>
<td>1 4 10 6 9</td>
<td>108</td>
<td>3.60</td>
</tr>
<tr>
<td>Hand function</td>
<td>2 3 0 11 14</td>
<td>122</td>
<td>4.07</td>
</tr>
<tr>
<td>Motor ability</td>
<td>3 4 5 4 14</td>
<td>112</td>
<td>3.73</td>
</tr>
<tr>
<td>Learning ability</td>
<td>2 1 3 8 16</td>
<td>125</td>
<td>4.17</td>
</tr>
</tbody>
</table>

4.3.2 Importance of improvement

Figure 3 shows that the consideration of most parents was that it was very important to achieve an improvement in all four areas. Table 3 summarises the result, and it can be seen that while communication was considered the least severe disability, it was considered most important to improve in this area. Although motor skills scored the lowest, the average score was a high 4.0.

![Figure 3. Parents' perception of the importance of improvement in each skill area](image-url)
Table 3. Parents' perceptions of the importance of improvement

<table>
<thead>
<tr>
<th>Area</th>
<th>Score</th>
<th>Total</th>
<th>Average</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>Communication</td>
<td>0</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Hand function</td>
<td>1</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>Motor ability</td>
<td>2</td>
<td>3</td>
<td>5</td>
</tr>
<tr>
<td>Learning ability</td>
<td>0</td>
<td>3</td>
<td>2</td>
</tr>
</tbody>
</table>

4.4 Therapies

Parents were asked to comment on the length of time each therapy had been given, the organisation of the therapy, the numbers of hours given each month, and whether it was included on their child's statement of Special Educational Needs.

4.4.1 Length of time children had received therapies

Parents were asked to indicate the length of time their child had been receiving each therapy, and Figure 4 shows a summary of the results.

Figure 4. Length of time children had received therapies
15 girls had never received music therapy, and 12 of the remaining 15 that had received the therapy had only received it in the last 4 years. A total of 7 girls had never received speech therapy; those that did receive it had done so for a range of time from 1 to 9 years, with most, 20, being given the therapy for between 1 and 7 years. Hydrotherapy was given to all but 8 girls, with a fairly even spread from 1 to 7 years. The results also showed an even time spread for occupational therapy but only 9 girls received it, whereas 21 did not. Only 3 girls did not receive physiotherapy, and those who did had done so through the range 1 to 9 years, with the majority falling into the range from 2 to 7 years. Aromatherapy was given to 21 girls, most in recent years, but 3 cases were reported where this therapy had been given for between 6 and 7 years.

4.4.2 Organisation of therapy sessions

Parents were asked how the therapy sessions were structured. Figure 5 shows how therapies were administered, either by group sessions only, individually, or a mixture of both.

For music therapy and hydrotherapy, the majority of girls receiving the therapies did so in groups. Speech therapy appeared to be spread more evenly across the
methods, whereas physiotherapy and aromatherapy were delivered to most girls in individual sessions. Only 9 girls received occupational therapy, 3 in groups and 6 individually.

4.4.3 Number of hours received

Parents were asked to specify the number of hours each child received, for each therapy, every month, and Figure 6 shows the results for this question.

![Figure 6 Number of hours per month](image)

Music therapy was given for between 1 and 5 hours per month, with an average of 3.3 hours per month, and 3 parents reported that although their daughters received the therapy, they did not know for how long. Speech therapy showed a similar pattern, with a range between 0.1 to 10 hours per month, and an average of 3.2 hours per month. 6 parents could not say how long this therapy was given for.

All parents were able to report the amount of time given to hydrotherapy probably because it requires some involvement on the parents' behalf, even if only to pack the swimming costume. The range of times was between 1 and 6 hours per month,
with an average of 3 hours per month. Occupational therapy was not widely available, and of the 9 cases where it was used 6 parents did not know how long their daughters were spending with the therapy. The other 3 cases reported different times ranging between 5 minutes and 4.5 hours per month.

Physiotherapy was one of the most widely available therapies, and of the 30 cases, 27 reported that it was given, but 7 respondents did not know how much time was involved. The majority of girls received between 2 and 4 hours per month, but 2 received more than 15 hours per month. Aromatherapy was given in 13 cases, for a maximum of 4 hours per month.

4.4.4 Inclusion of therapy on statement of Special Educational Needs

Parents were asked whether therapies were specified on their daughter's statement of Special Educational Needs. The results are given in Figure 7.

Every respondent was aware of the inclusion or otherwise of therapies on their child's statement. The only therapies to appear on statements more often than not were speech therapy and physiotherapy. These were also the two most common
therapies to appear on statements, with 16 and 17 cases respectively. There was a clear majority for 'not included' on the remaining therapies, with very large differences apparent for aromatherapy and occupational therapy. 9 children had no therapy specified on their statement of Special Educational needs.

The responses from the questionnaires showed that 90% of children whose statement of Special Educational Needs specified a particular therapy did in fact receive that therapy. Table 4 shows a summary of the therapies in relation to statements.

Table 4. The number of children receiving each therapy, related to its inclusion on statement of Special Educational Needs

<table>
<thead>
<tr>
<th>Therapy</th>
<th>Included on statement</th>
<th>Not included on statement</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Receiving</td>
<td>Not receiving</td>
</tr>
<tr>
<td>Music therapy</td>
<td>8</td>
<td>1</td>
</tr>
<tr>
<td>Speech therapy</td>
<td>15</td>
<td>1</td>
</tr>
<tr>
<td>Hydrotherapy</td>
<td>9</td>
<td>1</td>
</tr>
<tr>
<td>Occupational therapy</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>Physiotherapy</td>
<td>16</td>
<td>1</td>
</tr>
<tr>
<td>Aromatherapy</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Homeopathy</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Hippotherapy</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Osteopathy</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>53</td>
<td>5</td>
</tr>
</tbody>
</table>

Table 5 shows that 70% of children did not have music therapy listed on the statement. 80% of those who did receive music therapy did so in groups. In 3 out of the 4 cases when therapy was given individually, the therapy had been included on the statement.
Table 5. The organisation of music therapy in 30 Rett cases

<table>
<thead>
<tr>
<th>Music therapy</th>
<th>Included on statement</th>
<th>Not included on statement</th>
</tr>
</thead>
<tbody>
<tr>
<td>Therapy given individually</td>
<td>3</td>
<td>0</td>
</tr>
<tr>
<td>Therapy given as a group</td>
<td>5</td>
<td>6</td>
</tr>
<tr>
<td>Therapy given both individually and as a group</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Therapy not given</td>
<td>1</td>
<td>14</td>
</tr>
<tr>
<td>Total</td>
<td>9</td>
<td>21</td>
</tr>
</tbody>
</table>

47% of children did not have speech therapy listed on the statement (Table 6). The figures show that for children receiving speech therapy, when the therapy is included on the statement, 56% receive some individual therapy, but this number is reduced to 21% if speech therapy is not on the statement.

Table 6. The organisation of speech therapy in 30 Rett cases

<table>
<thead>
<tr>
<th>Speech therapy</th>
<th>Included on statement</th>
<th>Not included on statement</th>
</tr>
</thead>
<tbody>
<tr>
<td>Individual therapy</td>
<td>6</td>
<td>2</td>
</tr>
<tr>
<td>Group therapy</td>
<td>5</td>
<td>5</td>
</tr>
<tr>
<td>Individual and group therapy</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>Not known</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Therapy not given</td>
<td>1</td>
<td>6</td>
</tr>
<tr>
<td>Total</td>
<td>16</td>
<td>14</td>
</tr>
</tbody>
</table>

Table 7 showed that 67% of children did not have hydrotherapy listed on the statement. When the therapy is included on the statement, 50% receive some individual therapy, but this number is reduced to 10% if hydrotherapy is not on the statement. When the therapy was not detailed on the statement, 35% did not receive the therapy, and 55% received group therapy.
Table 7. The organisation of hydrotherapy in 30 Rett cases

<table>
<thead>
<tr>
<th>Hydrotherapy</th>
<th>Included on statement</th>
<th>Not included on statement</th>
</tr>
</thead>
<tbody>
<tr>
<td>Individual therapy</td>
<td>5</td>
<td>2</td>
</tr>
<tr>
<td>Group therapy</td>
<td>4</td>
<td>11</td>
</tr>
<tr>
<td>Individual and group therapy</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Therapy not given</td>
<td>1</td>
<td>7</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>10</strong></td>
<td><strong>20</strong></td>
</tr>
</tbody>
</table>

Table 8 showed that 43% of children did not have physiotherapy listed on the statement. Individual therapy was more prevalent than in all other therapies, and 94% of children with statemented physiotherapy received some individual treatment. 69% of children who did not have physiotherapy on their statements still received some individual therapy.

Table 8. The organisation of physiotherapy in 30 Rett cases

<table>
<thead>
<tr>
<th>Physiotherapy</th>
<th>Included on statement</th>
<th>Not included on statement</th>
</tr>
</thead>
<tbody>
<tr>
<td>Individual therapy</td>
<td>13</td>
<td>7</td>
</tr>
<tr>
<td>Group therapy</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Individual and group therapy</td>
<td>3</td>
<td>2</td>
</tr>
<tr>
<td>Therapy not given</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>17</strong></td>
<td><strong>13</strong></td>
</tr>
</tbody>
</table>

4.5 Effects of therapies on skill areas

Parents were asked to indicate how effective they thought each therapy was for their child in each of the four skill areas specified.

4.5.1 Perceived effect of music therapy

The responses show that 15 girls received music therapy, and their parents reported varied opinions of the therapeutic value. All respondents expressed an opinion, and the majority of scores were for 3 and above, with communication and functional hand use appearing to be the areas where most benefit was noticed.
Generally, motor skills were thought not to be improved by music therapy.

Table 9 showed that the area considered to benefit most from music therapy was communication, with 35% of the total score.

<table>
<thead>
<tr>
<th>Rank</th>
<th>Skill area</th>
<th>Total score</th>
<th>Average</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>communication</td>
<td>69</td>
<td>4.6</td>
</tr>
<tr>
<td>2</td>
<td>hand function</td>
<td>52</td>
<td>3.5</td>
</tr>
<tr>
<td>3</td>
<td>learning ability</td>
<td>44</td>
<td>2.9</td>
</tr>
<tr>
<td>4</td>
<td>motor ability</td>
<td>30</td>
<td>2.0</td>
</tr>
</tbody>
</table>

*n = 15*

4.5.2 Perceived effect of speech therapy

There were 23 girls who received speech therapy, but the parents of 2 girls were not able to express an opinion as to the value for motor ability and hand function. Figure 9 shows that the majority of scores were for values between 1 and 3. The only skill area to be rated 4 or 5 by more than half the parents was communication,
and the effect of speech therapy on the other three skill areas was somewhat lower. Motor skill was the area that scored the greatest number of low scores, with over half the respondents awarding a score of only 1. Only one respondent gave a score of 4 for the effect of speech therapy on motor ability, and there were no scores of 5 in this category.

![Figure 9: Perceived effect of speech therapy on skill areas](image)

Similarly, Table 10 showed that communication was also thought to benefit most from speech therapy, with 34% of the total score.

![Table 10: Total and average scores of value of speech therapy on each skill area](table)

<table>
<thead>
<tr>
<th>Rank</th>
<th>Skill area</th>
<th>Total score</th>
<th>Average</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>communication</td>
<td>73</td>
<td>3.3</td>
</tr>
<tr>
<td>2</td>
<td>learning ability</td>
<td>61</td>
<td>2.8</td>
</tr>
<tr>
<td>3</td>
<td>hand function</td>
<td>49</td>
<td>2.3</td>
</tr>
<tr>
<td>4</td>
<td>motor ability</td>
<td>34</td>
<td>1.6</td>
</tr>
</tbody>
</table>

*n = 22*
4.5.3 Perceived effect of hydrotherapy

Hydrotherapy was given to 22 girls, and every parent was able to express an opinion regarding the therapy's value. Figure 10 shows, not unexpectedly, that views were fairly evenly distributed across the range of scores, with a cluster around the value 3. Scores were evenly divided for motor ability. Scores for communication skill were concentrated around 3, 4 and 5, and scores for hand function and learning ability were generally 3 or below.

![Perceived effect of hydrotherapy](image)

Figure 10 Perceived effect of hydrotherapy on skill areas

Table 11 shows an equal score for communication and motor skill for areas thought to benefit most from hydrotherapy, with each having 29% of the total score.

<table>
<thead>
<tr>
<th>Rank</th>
<th>Skill area</th>
<th>Total score</th>
<th>Average</th>
</tr>
</thead>
<tbody>
<tr>
<td>1a</td>
<td>communication</td>
<td>74</td>
<td>3.4</td>
</tr>
<tr>
<td>1b</td>
<td>motor ability</td>
<td>74</td>
<td>3.4</td>
</tr>
<tr>
<td>3</td>
<td>hand function</td>
<td>54</td>
<td>2.5</td>
</tr>
<tr>
<td>4</td>
<td>learning ability</td>
<td>53</td>
<td>2.4</td>
</tr>
</tbody>
</table>

n = 22
4.5.4 Perceived effect of occupational therapy

Occupational therapy was delivered to 9 girls, and of these 9 cases, 2 parents were unsure of the value of the therapy in some skill areas (Figure 11).

![Perceived effect of occupational therapy](image)

**Figure 11. Perceived effect of occupational therapy on skill areas**

The results suggest that parents generally thought the therapy offered only a low value in the four skill areas considered, with only one score of 5 recorded, for functional hand use, and three scores of 4, one each for communication, hand function and learning ability.

There was a strong clustering of scores of 1 for all skill areas, with motor ability attracting the greatest number of lowest scores.

4.5.5 Perceived effect of physiotherapy

Of the 27 cases where physiotherapy was given, only one respondent failed to give an opinion on the therapy's value.

Figure 12 shows that whilst opinion was divided, motor skills were thought to benefit quite widely with 13 respondents awarding a score of 5, whereas learning skills and communication skills were not thought to be greatly improved by physiotherapy.
The greatest response for hand function was ten awards of 3, the greatest for learning ability was nine awards of 1, and the greatest for communication was nine awards of 3.

The area which most benefited by physiotherapy is motor skill with 31% of the total score, followed by hand function, communication and learning skill, as shown in Table 12.

<table>
<thead>
<tr>
<th>Rank</th>
<th>Skill area</th>
<th>Total score</th>
<th>Average</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>motor ability</td>
<td>99</td>
<td>3.8</td>
</tr>
<tr>
<td>2</td>
<td>hand function</td>
<td>80</td>
<td>3.1</td>
</tr>
<tr>
<td>3</td>
<td>communication</td>
<td>72</td>
<td>2.8</td>
</tr>
<tr>
<td>4</td>
<td>learning ability</td>
<td>68</td>
<td>2.6</td>
</tr>
</tbody>
</table>

n = 26

4.5.6 Perceived effect of aromatherapy

Aromatherapy was given to 13 girls, but was not thought to be very beneficial in any of the skill areas. The only area to attract a score of 5 (from 3 cases) was
communication. Figure 13 shows the response for aromatherapy, and shows an obvious clustering of scores of 1 for each of the four skill areas.

![Perceived effect of aromatherapy](image)

Figure 13 Perceived effect of aromatherapy on skill areas

### 4.6 Summary

At 30%, the response to the survey was disappointing, but the sample size of 30 allowed basic statistical analysis on the data to be carried out (Cohen and Manion, 1994). The features of diagnosis of Rett syndrome were within the expected ranges, and there were no unusual results in this area. The most common therapies were physiotherapy, speech therapy and hydrotherapy, with each of these being given to more than two thirds of the girls studied. Half the sample received music therapy, and approximately one third received aromatherapy and approximately one third received occupational therapy. There were a few reports of 'minor' therapies, with hippotherapy, homeopathy and osteopathy all present in the survey.

Parents generally considered their daughters to be severely disabled, although communication and motor ability were in the moderate range. Parents saw communication as the area in which improvement was most needed. The length of time therapies had been given was varied and there was no obvious pattern between therapies. Physiotherapy and aromatherapy were often delivered to girls.
Chapter 5 Analysis and discussion

5.1 Introduction

The results given in the previous chapter are analysed and discussed in this chapter. Whilst focusing on parents’ views of their children with Rett syndrome, the chapter looks at relevant research and practice in other areas of special needs in order to consider issues more clearly and to set Rett syndrome in a wider educational context.

5.2 Parents' perceptions of severity of disability

Parents’ perceptions of the severity of disability are likely to vary according to their own experiences - parents of a first born child may not have personal benchmarks for comparisons, whereas parents with other children may be more aware of developmental stages. Parental anxiety may also be an issue, and may be more pronounced with the first child. This study does not indicate whether a child has siblings, so it is not possible to tell if this was a factor in determining parents’ views of severity of disability.

68% of the scores were for the values of 4 and 5, indicating that the overall level of disability was considered severe. Overall, parents considered communication to be the area causing the fewest problems, with 50% scoring 4 and 5, although the loss of previously acquired speech, so characteristic of Rett syndrome, may be considered to be one of the most significant disabilities of all (Jones and Creegan, 1986). The apparent belief of parents in their daughters’ underlying ability to communicate (Lindberg, 1994) may be due to a subconscious desire to maintain a close bond, or it might be simply a result of sustained practice in which previously acquired interaction continues in an altered form. There may also be a foundation in reality of the frequent claims of parents that their daughters understand more than they can express. It is probably impossible to quantify how much more important it may be to parents of girls with Rett syndrome to communicate in a social environment where personal rewards in any area are likely to be minimal.

As with communication ability, many parents believe that girls with Rett syndrome understand more than is readily apparent (Lindberg, 1994, Weisz, 1987), although there is little evidence to substantiate these claims. The results of the questionnaires, however, showed that learning ability was the area perceived by
parents to be most severely affected, see Table 13. The loss of physical functionality is not that uncommon in the wider population, and is a disability that can be overcome in the sense that our society is paying more attention to issues of equality for the disabled person. The publicity given to Stephen Hawkins, for example, demonstrates that it is possible for a remarkable intellect to succeed in the face of overwhelming physical adversity. Parents may find it easier to cope with a disability in which arms and legs fail to work than with a dysfunctional intellect.

<table>
<thead>
<tr>
<th>rank</th>
<th>skill area (a)</th>
<th>skill area (b)</th>
<th>n</th>
<th>W</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>1&gt;2</td>
<td>learning ability</td>
<td>hand function</td>
<td>30</td>
<td>31</td>
<td>0.3580</td>
</tr>
<tr>
<td>1&gt;3</td>
<td>learning ability</td>
<td>motor ability</td>
<td>30</td>
<td>91</td>
<td>0.0360</td>
</tr>
<tr>
<td>1&gt;4</td>
<td>learning ability</td>
<td>communication</td>
<td>30</td>
<td>142</td>
<td>0.0270</td>
</tr>
<tr>
<td>2&gt;3</td>
<td>hand function</td>
<td>motor ability</td>
<td>30</td>
<td>90</td>
<td>0.0370</td>
</tr>
<tr>
<td>2&gt;4</td>
<td>hand function</td>
<td>communication</td>
<td>30</td>
<td>126</td>
<td>0.0361</td>
</tr>
<tr>
<td>3&gt;4</td>
<td>motor ability</td>
<td>communication</td>
<td>30</td>
<td>125</td>
<td>0.3681</td>
</tr>
</tbody>
</table>

Sample size (n), Wilcoxon statistic (W) and probability (p)

It is realised that the term 'severity of disability' may be interpreted in a variety of ways, and that parents will not have a uniform base against which to make comparison. Learning ability and hand function are the two areas considered to be most severely affected, but the difference between the two areas is not significant. The severity in each area is, though, significantly greater than in the remaining two areas, and there is no significant difference between the lowest two, motor ability and communication skills.

5.2.1 Parents perceptions of importance of improvement

Communication, thought to be the area of least disability, was perceived to be the skill most important to improve, with 83% of parents scoring 5. Perhaps this again is related to the belief that physical dysfunction is a disability which can be carried, whereas communication is considered a fundamental requirement in achieving any real quality of life. The score for importance of improving communication is significantly greater than hand function and motor ability, but there are no other significant differences in Table 14.
There are three main issues in the assessment of progress, what is being measured, how it is being measured and the relevance of the assessment in guiding further teaching (Ware and Healey, 1994). Even using checklists with small steps it is sometimes difficult to measure the improvement of children with profound and multiple learning difficulties and such methods are not necessarily effective guides for future planning. Criterion-referenced tests, such as the Standard Assessment Tasks are not effective progress measures for children who are unable to reach Level 1, and do not help the teacher determine teaching strategies. Performance criteria have been developed to support schools by providing a common basis for measuring the progress of pupils for whom the early levels of the National Curriculum are not appropriate (DfEE, 1998c). These are designed for use by teachers when making summative assessments, rather than for use by teachers when making day-to-day assessments of pupils' progress, and again have little relevance for determining teaching strategies.

All parents in this study were able to express an opinion about the importance of improving ability in each particular skill area. One concern regarding the inclusion of this issue on the questionnaire was that respondents would consider the importance of improving to be maximum in each skill area. Parents have different expectations and degrees of tolerance of behaviour (Jordan, 1999a) and this may be why there were different perceptions of the importance of improving.

5.2.2 Severity of disability and the importance of improving

It might be expected that a positive correlation would exist between severity of disability and the importance of improving, that the more severe the disability was
thought to be, the more important it would be to gain some improvement. This did in fact prove to be so, but the relationships were not strong, and Table 15 shows that there were no significant correlations.

Table 15 Correlation between severity of disability and the importance of improving in each of the four skill areas

<table>
<thead>
<tr>
<th>Skill area</th>
<th>Severity</th>
<th>Importance</th>
<th>rs</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Mean</td>
<td>Mode</td>
<td>Mean</td>
<td>Mode</td>
</tr>
<tr>
<td>Communication</td>
<td>3.60</td>
<td>3</td>
<td>4.77</td>
<td>5</td>
</tr>
<tr>
<td>Hand function</td>
<td>4.07</td>
<td>5</td>
<td>4.37</td>
<td>5</td>
</tr>
<tr>
<td>Motor skill</td>
<td>3.73</td>
<td>5</td>
<td>4.03</td>
<td>5</td>
</tr>
<tr>
<td>Learning ability</td>
<td>4.17</td>
<td>5</td>
<td>4.50</td>
<td>5</td>
</tr>
</tbody>
</table>

The Spearman statistic is positive in each case, but the values indicate only very weak associations. It appears then, that parents consider that it is important to improve in the four ability areas no matter how serious they perceive their child's level of disability to be. Perhaps the wish for improvement is consistently too high to allow variation to be detected. Parents naturally want 'the best' for their children, and their desire to see improvement is a natural feeling reflected in these results. The mode score of 5 occurs in all cases except the severity of communication disability, another pointer to the widely held parental view that girls with Rett syndrome are able to communicate more extensively than tests would suppose.

5.3 Diagnosis

Diagnosis of autistic spectrum disorders is based on the triad of impairments that form the basis of diagnostic criteria (World Health Organisation, 1993, American Psychiatric Association, 1994). There may be a number of different disorders, with different diagnostic criteria, within this spectrum, but they all share common developmental difficulties (Jordan, Jones and Murray, 1998) and there are no laboratory tests that can confirm the conditions. Rett syndrome can be found within this spectrum and its clinical diagnosis depends on observation of a child's early growth and development and on an ongoing assessment of medical history and physical and neurological status (Rett Syndrome Diagnostic Criteria Work Group, 1988, WHO, 1993). From the 30 responses to the questionnaires in this study, the
average age at diagnosis was 4 years, with a span of 2 - 10 years. It is likely that children are now being diagnosed with Rett syndrome at an earlier age than even ten years ago, although in this study, the 2 oldest children, both born in 1986 were diagnosed at 2 years and 10 years respectively. It is doubtful that this large differential is due to a differing level of disability, it is more likely due to a geographical variation in levels of awareness. The condition is still not widely recognised by the medical profession and in an interview with a British Rett parent in 1994, Professor Andreas Rett observed that 'The number of cases in any country still depends on the doctors in those countries making the right diagnoses' (in Freeman, undated). Even today professionals can, because of a lack of knowledge, be sceptical when parents first begin to explain the problems that they have with their children (Hill, 1997).

Klauber (1999) considers the diagnosis of autism to be particularly traumatic and states that 'There are many professionals who deliver the diagnosis without offering follow-up appointments to help parents take in what it means' (p.37). Klauber adds that some professionals underestimate the impact of throwaway remarks relating to the nature of the condition and its likely impact upon the family. This lack of sensitivity, described by Cunningham and Davis (1985) and considered to be an 'expert' stance, is likely to exacerbate feelings of confusion in the parents, and may lead to other feelings, such as bitterness or even helplessness.

Disabilities that are obvious from birth allow therapeutic involvement to be set in motion immediately, thus optimising the opportunity for benefit. As girls with Rett syndrome appear to develop normally, deviations from the physical or intellectual norm may not be noticed until an age when these opportunities for early intervention are lost. Referring to children with autism, Jordan and Powell (1995) point out that although early intervention is advantageous, it is never too late to improve quality of life. These authors argue that 'an understanding of the fundamental difficulties faced by a pupil at a psychological level is crucial to developing a curriculum and a teaching approach that addresses the pupil's needs' (p.5). A diagnosis is needed before this can take place. It has been seen that there may be a gap between diagnosis and provision of educational advice and therefore close communication between those making the diagnosis and educational providers is necessary (Jordan and Jones, 1997).
Parents find it particularly distressing when their child shows symptoms of autism after appearing to develop normally for the first year or two of life (Jordan, 1999a). The shock and deep disbelief felt by parents of Rett syndrome children appears to be more severe than that of parents of children diagnosed with a disability at birth, since they have believed their child to be normal and have shed the early anxiety felt by all new parents (Goodship, 1987). Lewis and Wilson (1998, p.65) quote Field (1990) in describing the feelings of parents before diagnosis - 'Every Rett girl and her family before this date went through grotesque and seemingly inexplicable events which neither they nor the professionals could understand or predict'.

A parent of the first child in the United States to be diagnosed with Rett syndrome, and the founder of the International Rett Syndrome Association summarises the aims of the association and the thoughts of other parents:

We share information about the medical aspects of the syndrome and try to help parents sort out their feelings of guilt, anger, frustration, resentment and sorrow. We also find that many of us who have waited many years to find out just what went wrong are relieved to at last have a diagnosis.

(Hunter, 1987, p.534)

Whereas early diagnosis brings with it the chance for early intervention, hope may turn to despair as parents begin to realise the full implications of the identification of Rett syndrome. 'A diagnosis of Rett syndrome implies a severe, combined dysfunction of motor, cognitive and communication abilities, and a growth disturbance, appearing in infancy or early childhood' (Witt-Engerström, 1993, p.26).

Houghton (undated) found that it was not until the diagnosis of Rett syndrome had been made that the family was able to plan for the future. These plans must inevitably include thoughts about the child's educational needs as well as day to day coping strategies for the family. Families will become involved with a number of professionals once a disability has been confirmed, whether or not there has been a definite diagnosis. Cunningham and Davis (1985) discuss the phases of shock, reaction, adaptation and orientation which parents experience when faced with the reality of a child with special needs. These phases are accompanied by a number of emotions, including anger, resentment and guilt, and parents' distress can be vented on professionals (Klauber, 1999). Professionals need to be aware of these feelings, and avoid reactions to parents' emotions that can be construed as negative. This is important when giving information to parents and Harris (1990)
suggests that '... it is necessary to consider not only what information to provide for parents, but how this information can be most usefully presented' (p.103).

Once a disability has been confirmed, the individual needs of the child can be assessed by the Local Education Authority and a statement of Special Educational Needs should be issued.

5.4 Statement of Special Educational Needs

A statement of Special Educational Needs outlines how, where and with what resources a child's needs will be met by the Local Education Authority (DFE, 1994). Material for the statement is sought and acquired from educational and medical sources, and includes a contribution by parents. For most parents, writing a report about their child is likely to be both difficult and painful (Gascoigne, 1995). Lewis and Wilson (1995) agreed that this may be a challenging task and they quote one mother 'I apologise for the long delay in submitting this personal view of my daughter's needs. It has been a very painful experience for me' (p.4).

An annual review of the statement should ensure that Special Educational Needs are re-examined, progress is evaluated and the provision reappraised to ensure its continued suitability. Although it is the responsibility of the LEA to ensure this annual review takes place, in practice the duty is delegated to schools.

As well as identifying educational needs and resources, additional non-educational provision will be specified on the statement. This section includes recommendations for the use of therapies, the cost of which is usually borne by the Health Authority. At each annual review of the statement, the LEA should invite representatives from the Health Authority to contribute to the review, either by submitting a report or attending the annual review meeting (DFE, 1994).

The Government Green Paper 'Excellence for all children' (DfEE, 1997) is the first step in a reappraisal of the way in which the Special Educational Needs of children are met. The paper recognises that the distinction between educational and non-educational provision is unclear, accepting that therapies may benefit a child's educational progress, even if provided for health reasons. In the recent response to the paper (DfEE, 1998a) there was almost universal support for promoting better co-operation between education and health services. Suggestions for improving
relationships included clarifying or changing the statutory responsibilities of different agencies and introducing more flexible funding arrangements between agencies.

If the proposal to end the distinction between educational and non-educational needs and provision in statements is accepted, then the present lack of conformity for the provision of therapies may be redressed. How the effect of therapies can be assessed is more problematic, but there will be a need to determine the value of therapies for an individual child.

Currently there is no uniform objective method in widespread use in the UK for dealing with the issue, and a system is required for monitoring therapy effects. However, there is already a mechanism for regular inspection of schools, and there is also an accreditation process through the National Autistic Society that attempts to evaluate the quality of provision for children with autism, based on achieving and maintaining specific criteria (Jordan, Jones and Murray, 1998). School inspections tend to focus on aspects of teaching and learning, but it is not impossible that they could provide a foundation upon which to build a more comprehensive evaluation system.

Confirmation is needed that a treatment is actually better than doing nothing, and that an appropriate quality of provision is being implemented. Weiss (1998) makes a distinction between efficacy studies, effectiveness studies, and routine monitoring as methods of evaluating child psychotherapy in the USA, and suggests reasons why routine monitoring might be needed. All three research methods offer ways of gathering information, but there are ethical difficulties with the notion of control groups and because the therapy has been implemented for the purpose of treating a child and not for research, efficacy studies do not always lend themselves easily to special school environments. Effectiveness studies seek to evaluate treatments under the conditions where they are most likely to be applied, in naturalistic settings, but give snapshots in time. Routine monitoring, although described by Weiss in the context of American clinics delivering psychotherapy, could provide a framework for ongoing therapy assessment in British schools, provided that the purpose of the monitoring was clearly defined. There are issues which would need to be addressed, such as the need for control groups in order to determine whether observed change is attributable to treatment, and the fact that improvement is not sufficient in itself to demonstrate treatment effectiveness, but the multiple
information sources necessary are already present in school, i.e. teachers, parents, therapists, and the children themselves.

The objective of routine monitoring is, in Weiss' view, not to establish the general effectiveness of a treatment but rather to determine, for a particular clinic or practice, the parameters of the treatment's effectiveness. As the need for accountability grows, establishments should offer effective treatments and be able to document their effectiveness on an ongoing basis.

The reasons for conducting routine monitoring offered by Weiss are adapted and developed by Jordan (1999b) and given greater focus on educational evaluation. Whilst raising questions dealing with the success of intervention in a particular situation, absolute effectiveness, comparative effectiveness, treatment integrity and subgroup effectiveness, Jordan touches on the problems of involving participants in the research, the difficulties associated with control groups, and matters of ethics.

In discussing educational interventions for children with autism, Jordan, Jones and Murray (1998) propose reasons why evaluative research in schools is potentially problematic. Areas of difficulty may include observer skills, small population effects, focus distortion, the use of control groups, assessment of parental input, observer bias and long-term versus immediate reaction conflict.

Statements of Special Educational Needs reflect and are controlled by national legislation and assessment at local level; therefore evaluations of interventions need to be influential at both levels. It might be the case that small scale research will sway opinions locally, while collective reporting and larger scale projects are needed to influence national policy.

5.4.1 Inclusion of therapy on statement of Special Educational Needs

The DfEE (1997) claims that difficulties in securing therapy services for children with Special Educational Needs result partly from the different statutory responsibilities and priorities of Health Authorities and Local Education Authorities, and from lack of clarity over funding. In this study, it was anticipated that some differences in therapy provision would be identified, although it was decided not to ask for geographic information in the questionnaire as substantial research is needed to explore this issue fully. Differences in provision were indeed identified, and it was found that although all children had been diagnosed with the same
critical disabling condition, nine girls had no therapies included on their statements, while five girls had four or more.

Only two girls had statements that included therapies that were not given. One case related to statemented speech therapy, physiotherapy and occupational therapy, where only occupational therapy was not given, but in the other case the girl’s statement specified music therapy, speech therapy, hydrotherapy and physiotherapy, but no therapies were given. The parent of this child was disappointed with her daughter’s situation, and commented: ‘These girls need the maximum input to get the maximum output. Without these therapies they are left to waste away without any help in the condition. With no help they make no progress’ (Questionnaire 16). Clearly the respondent in this case felt some resentment towards the authorities. The reasons for non-provision of statemented therapies are not known, and although few, the omissions may be indicative of wider issues such as lack of funding, shortages of trained therapists, and disagreement between parents and assessors regarding actual needs.

A Local Education Authority is legally obliged to ensure that funding is made available to support the statement, but it would seem that this legal requirement is not always met. Parents may need help to challenge the authority, and if disputes cannot be satisfactorily resolved, parents have the right to appeal to the Tribunal, the 'final arbiter in disputes between parents and LEAs' (DfEE, 1997). This is seen as a last resort when all other negotiations have failed. LEAs have employed professional advisers to work alongside parents, but unless it is clear that there is no conflict of interests, this partnership will not succeed and only those ‘articulate and energetic minority’ of parents (Simmons, 1997, p.123) will challenge LEA decisions.

Simmons also points out that a two-tier system of statements is emerging in many LEAs:

In one group are those parents who challenge LEA decisions and whose child’s Statement is ultimately re-written by the Tribunal in a way that is consistent with the law. In the other, much larger, group are those who do not challenge and as a result end up with inadequate statements which fail to guarantee provision for their children.

(p. 123)
One parent in the study group felt so strongly about the inclusion of a therapy on her daughter's statement that she was prepared to fight for what she considered to be the rights of her daughter. 'I had to campaign very hard to get music therapy and was one of the first to have it statemented at this school. Subsequently all Rett girls now have it on their statement at this school' (Questionnaire 65). There is no clear indication how other parents felt, and it is not known if they were satisfied with the services provided.

The Green Paper (DfEE, 1997) states that 'Improved arrangements for encouraging dialogue between parents, schools and LEAs should be reflected in a reduction in the number of appeals to the SEN Tribunal' (p.30) and it is to be hoped that this will eliminate the need for parents to endure the anxiety of such appeal. However, it remains to be seen where the true motivation behind the Green Paper lies, and whether political and financial mechanisms eventually prove to be dominant.

20% of parents showed an awareness of financial constraints, with comments such as 'Music therapy is purchased as an extra, it depends on funds' (Questionnaire 32), 'I think the value of a wide range of therapies for Rett girls is very important but unfortunately where we live there is not a great deal of funding available to give the children all the different therapies' (Questionnaire 23), 'It is obviously all down to money!' (Questionnaire 207) and 'There does not seem to be the money made available in parts of the country for effective therapies' (Questionnaire 199).

Parents of children with special needs feel many emotions, and dissatisfaction with the educational system is likely to compound feelings of frustration, helplessness and even anger. Hunter (1987) felt that the incessant strain and responsibility of caring for a child with Rett syndrome could originate from a combination of sources:

> The emotional stress of having lost their child is compounded by the physical stress of having to care for her, at the same time trying to provide for the needs of other family members. For the parent of such a child, every day is filled with physical, emotional and financial tolls.

(p.537)

The issue of funding should not concern parents; the delivery of statemented therapies is not a privilege, but a considered and agreed measure designed to alleviate some of the physical burden suffered by the child. It is the right of children

134
and their parents to have necessary therapies delivered in a professional way, yet there is evidence here that lack of resources interferes with the process.

5.4.2 Therapies given but not specified on statement

80% of children actually received at least one therapy when there was no obligation on the part of the school to ensure that therapy was given. In every therapy mentioned, there were instances of the therapy being given although not specified on the statement. It is unclear who provided the funding for this therapy. This may be for a variety of reasons. If a school values the results of a therapy to such an extent that the benefit for the child is considered to be paramount, then it may be prepared to bear the cost of that therapy from its own funding, possibly taking advantage of activities formalised through other pupils. Parental pressure can be a powerful instrument in implementing change, particularly if a united front is presented, with one parent claiming: 'Physiotherapy has improved recently due to parental complaints, but still not sufficient 1 to 1 with qualified physiotherapist' (Questionnaire 99).

Even schools may not be able to support the parents and children in the way they would wish. In a personal study (Moore, 1996) it was found that whilst claiming to recognise the value of music therapy, one LEA would not permit it to be included on the statement of Special Educational Needs of a girl with Rett syndrome, in spite of appeals from the school on behalf of the child. The LEA indicated that music therapy did not make such a 'significant contribution to provision as to merit specific inclusion in a statement, although the school may wish to provide it' (unpublished correspondence, 1996). The school remains convinced of the value of music therapy for children with Rett syndrome and continues to bear the cost of this facility.

5.4.3 The relationship between the statement of Special Educational Needs and whether a therapy is given

The legal status of a statement has been discussed, and it is reasonable to suppose that the appearance of a therapy on a statement will influence provision, and Table 16 shows that this appears to be the case. When the results are pooled, the Chi-square statistic of 74.46 relates to a p value of less than 0.001, indicating that there is a strongly significant effect. This is as expected, and shows that therapies are much more likely to be given when they appear on the statement. To
eliminate any effect from minor therapies, if only the four commonest therapies are considered, Chi-square reduces to 18.06, but $p$ remains less than 0.001. Chi-square cannot be accurately computed for individual therapies because of the low numbers in some cells.

Table 16: Relationships between statements and whether therapy is given

<table>
<thead>
<tr>
<th></th>
<th>Included on statement</th>
<th>Not included on statement</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total therapies given</td>
<td>53</td>
<td>60</td>
</tr>
<tr>
<td>No therapy given</td>
<td>5</td>
<td>152</td>
</tr>
<tr>
<td>Chi-square</td>
<td>74.46</td>
<td></td>
</tr>
<tr>
<td>$p$</td>
<td>&lt;0.001</td>
<td></td>
</tr>
</tbody>
</table>

pooled for $n = 30$ cases and $N = 9$ reported therapies

5.5 Organisation of therapies

Although a child's statement of Special Educational Needs should specify if a therapy is to be provided, it is unlikely to stipulate if this therapy is to be given individually or in a group session. Although girls with Rett syndrome are diagnosed by their symptoms, and therefore display numerous similarities, their specific therapeutic problems and responses to treatment vary considerably (Van Acker, 1991) and so the needs of the individual child should be paramount when considering how a therapy is administered. Who makes this decision, and on what grounds, is likely to vary from authority to authority or even from school to school. The decision is likely to be dependent on issues such as availability of staff, timetabling constraints and financial matters, as well as the needs of the child.

There is a need for a peer group for children with autism and Golding (1997) believes that in normal development this can facilitate individual autonomy, although it may be, of course, that a therapy session is not the most appropriate time for group interaction.

5.5.1 The relationship between the organisation of the therapy and the statement of Special Educational Needs

When the numbers are pooled for group and individual therapies, (Table 17) it can be seen that the results are permissive of a relationship in which presence of a
therapy on a statement of Special Educational Needs influences the way the therapy is given.

Table 17. Relationships between how therapy is organised and statements of Special Educational Needs, all therapies included

<table>
<thead>
<tr>
<th></th>
<th>Included on statement</th>
<th>Not included on statement</th>
</tr>
</thead>
<tbody>
<tr>
<td>Individual therapy</td>
<td>21</td>
<td>34</td>
</tr>
<tr>
<td>Group therapy</td>
<td>14</td>
<td>31</td>
</tr>
<tr>
<td>Chi-square</td>
<td>6.38</td>
<td>&lt;0.02</td>
</tr>
</tbody>
</table>

The Chi-square statistic of 6.38 for all therapies is significant, with p < 0.02. When the four main therapies only are pooled, Chi-square increases to 8.11, and p falls to under 0.01. However, cause cannot be attributed to effect in this type of study, and there are other explanations which could be equally realistic. The two likely explanations are that statementing may indeed influence therapy organisation, or that those therapies which tend to be given individually, such as physiotherapy, are the ones most likely to appear on the statement anyway, whereas fringe therapies tend to be given in groups, and omitted from statements. In general though, this study found that if a therapy is specified on a statement of Special Educational Needs, and is subsequently given to the child, it is more likely to take the form of individual therapy than group therapy. If the therapy does not appear on the statement, then there is less likelihood of the child receiving the therapy.

Undoubtedly group therapy is a cheaper option, and it may be that if groups are already running, it is possible to 'top up' with additional pupils, with minimal expense. Some therapists may prefer the group approach, feeling that it gives additional opportunities for interaction between pupils.

5.5.2 Perceived benefits of therapies, relating to organisation

Whether a child receives individual or group therapy should depend on individual circumstances. Individual therapy is not necessarily the more appropriate therapy, and it has been seen that group therapy can offer benefits of, for example, cooperation and can allow children to learn from each other (Association of Professional Music Therapists, 1992, Reid Campion, 1997, Reid, Millar, Tait,
Donaldson, Dean, Thomson and Grieve, 1996). This study found that group music therapy was more common than individual music therapy. The analysis shows that there are no significant differences in perceived values of group music therapy and individual music therapy (Table 18).

Table 18. Perceived benefits of music therapy, relating to organisation - average scores

<table>
<thead>
<tr>
<th>Grouping</th>
<th>n</th>
<th>Communication</th>
<th>Hand function</th>
<th>Motor ability</th>
<th>Learning ability</th>
</tr>
</thead>
<tbody>
<tr>
<td>Individual</td>
<td>3</td>
<td>4.67</td>
<td>3.33</td>
<td>1.33</td>
<td>2.67</td>
</tr>
<tr>
<td>Group</td>
<td>11</td>
<td>4.64</td>
<td>3.73</td>
<td>2.27</td>
<td>3.00</td>
</tr>
<tr>
<td>Both</td>
<td>1</td>
<td>4.00</td>
<td>1.00</td>
<td>1.00</td>
<td>3.00</td>
</tr>
<tr>
<td>u</td>
<td>17</td>
<td>19.5</td>
<td>20.5</td>
<td>18.5</td>
<td></td>
</tr>
<tr>
<td>p</td>
<td>1.0000</td>
<td>0.7692</td>
<td>0.6593</td>
<td>0.8846</td>
<td></td>
</tr>
<tr>
<td>N</td>
<td>14</td>
<td>14</td>
<td>14</td>
<td>14</td>
<td></td>
</tr>
</tbody>
</table>

Mann Whitney (u), probability (p), excluding 'both' category, and sample sizes (N,n)

In groups, the emphasis is on providing not only an opportunity for children to gain a greater self awareness through musical interaction with the therapist, but also to develop awareness of others through spontaneous and appropriate pupil / pupil interaction, as well as providing diagnostic information concerning the function of an individual pupil within a group (Association of Professional Music Therapists, 1992). It is, without doubt, cheaper to give music therapy to a group of children than to individuals. Before a decision is made about the organisation of sessions, it is hoped that the therapist has considered the needs of the individual child, rather than financial issues.

No significant differences were found between perceived values for speech therapy delivered in group sessions and speech therapy given in individual sessions (Table 19).
Table 19 Perceived benefits of speech therapy, relating to organisation - average scores

<table>
<thead>
<tr>
<th>Grouping</th>
<th>n</th>
<th>Communication</th>
<th>Hand function</th>
<th>Motor ability</th>
<th>Learning ability</th>
</tr>
</thead>
<tbody>
<tr>
<td>Individual</td>
<td>8</td>
<td>3.00</td>
<td>2.13</td>
<td>1.63</td>
<td>2.50</td>
</tr>
<tr>
<td>Group</td>
<td>10</td>
<td>3.67</td>
<td>2.63</td>
<td>1.38</td>
<td>3.11</td>
</tr>
<tr>
<td>Both</td>
<td>4</td>
<td>3.25</td>
<td>2.25</td>
<td>1.75</td>
<td>2.75</td>
</tr>
<tr>
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<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>u</td>
<td>43.5</td>
<td>44</td>
<td>33</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>p</td>
<td>0.5414</td>
<td>0.2345</td>
<td>0.9591</td>
<td>0.4234</td>
<td></td>
</tr>
<tr>
<td>N</td>
<td>17</td>
<td>16</td>
<td>16</td>
<td>17</td>
<td></td>
</tr>
</tbody>
</table>

Mann Whitney (u), probability (p), excluding 'both' category, and sample sizes (N,n)

Reid et al. (1996) found reported benefits from parents and professionals when programmes were integrated into the curriculum rather than kept separate from it. In a study of 116 parents, Jordan and Jones (1997) found that professionals who were able to work directly with a child, such as speech therapists, were generally rated higher by parents than professionals who just gave advice, particularly if they were thought to know little about the condition of their child. It may be that actual provision of the therapy and close contact with the therapist is sufficient to influence opinion.

Just as speech and language is an integral part of everyday life, so speech and language therapy should be incorporated into all aspects of daily life, both at home and at school. Not only does this entail skills of collaboration on the part of the therapist, it also pre-supposes willingness and competence on the part of parents to reinforce the therapy process in the home.

Perceived values for group hydrotherapy and individual hydrotherapy likewise do not reveal any significant differences (Table 20).
Reid Campion (1997) believes the ideal solution in hydrotherapy is for a child to receive individual therapy, within a group situation. From a safety perspective, it would be unwise not to use one-to-one support irrespective of other factors, and group therapy without individual support is untenable. Lewis and Wilson (1998) highlight the dangers of leaving a girl with Rett syndrome unattended in the water irrespective of any buoyancy aids, and personal experience has shown that without parental support, it is not always possible for pupils to take part in hydrotherapy sessions. The use of parents to support children in hydrotherapy sessions is not uncommon and Reid Campion (1997) believes that parents may become very effective handlers of their children in the water. As well as providing an opportunity for parents to forge a bond by the closeness essential to hydrotherapy, it is without doubt a very cost effective means of providing a facility that may otherwise be financially prohibitive. A weakness of this analysis is the lack of clarity about what is meant by individual and group hydrotherapy, since individual could be a one-to-one process in a group environment. The use of a hydrotherapy pool for swimming does not necessarily constitute therapy, although there are common elements between the two (Lewis and Wilson, 1998) and it is unclear whether the provision received by the girls in this study was, in fact, hydrotherapy or warm water swimming.

The results so far do not come close to significance for any of the four ability areas for music therapy, speech therapy and hydrotherapy, and do not identify any perceived differences for group or individual therapy; however, the results for physiotherapy do reveal differences. Table 21 shows that for communication skill and motor ability, physiotherapy is perceived to be significantly more valuable when
given individually. The values for learning ability approach, but just fail to reach, significance with a p value of 0.0838. In every case, the scores for individual and for 'both' are higher than the scores for group therapy alone.

Table 21 Perceived benefits of physiotherapy, relating to organisation - average scores

<table>
<thead>
<tr>
<th>Grouping</th>
<th>n</th>
<th>Communication</th>
<th>Hand function</th>
<th>Motor ability</th>
<th>Learning ability</th>
</tr>
</thead>
<tbody>
<tr>
<td>Individual</td>
<td>20</td>
<td>2.95</td>
<td>3.11</td>
<td>4.05</td>
<td>2.84</td>
</tr>
<tr>
<td>Group</td>
<td>2</td>
<td>1.00</td>
<td>2.50</td>
<td>1.00</td>
<td>1.00</td>
</tr>
<tr>
<td>Both</td>
<td>5</td>
<td>2.80</td>
<td>3.20</td>
<td>4.00</td>
<td>2.40</td>
</tr>
<tr>
<td>u</td>
<td>35</td>
<td>22.5</td>
<td>37</td>
<td>33</td>
<td></td>
</tr>
<tr>
<td>p</td>
<td></td>
<td>0.0464</td>
<td>0.6636</td>
<td><strong>0.0216</strong></td>
<td>0.0838</td>
</tr>
<tr>
<td>N</td>
<td>21</td>
<td>21</td>
<td>21</td>
<td>21</td>
<td></td>
</tr>
</tbody>
</table>

Mann Whitney (u), probability (p), excluding 'both' category and sample sizes (N,n)

Although physiotherapy has conventionally been administered to individual children, on a one-to-one basis, since the introduction of conductive education into the British education system, group physiotherapy has become increasingly popular (Cottam and Sutton, 1986). Girls with Rett syndrome have very individual physical disabilities and it may be this that causes the parents to believe that individual therapy is most appropriate.

It is difficult to draw firm conclusions from these analyses. Better results might be anticipated from therapies delivered individually, but the results do not indicate that this is so. There is no obvious reason to suppose physiotherapy should depend upon individual or group delivery for its level of success any more so than other therapies, yet for communication and motor ability the perception of respondents is that individual delivery is better. Respondents may have had preconceived notions of physiotherapy and its benefits, to the extent that their answers were modified not only by what they observed in their children, but by contact with doctors and other agencies during the many discussions which must have occurred as Rett syndrome was diagnosed. Possibly, the significant differences found here are no more than artefacts of an over simple methodology that does not isolate influences. Further research is required in this area to eliminate the possible preconceptions that may be influential, and an objective assessment of the various effects is required.
The number of children receiving other therapies was small, as shown in the previous chapter. Only one girl received osteopathy, one received homeopathy, and two received hippotherapy. The numbers are insufficient to enable any form of analysis to be completed.

5.6 Perceived effects of therapies

If specific therapies are designed to facilitate improvement or progress in specific areas, it would be unlikely that a particular therapy would be perceived to be equally effective across all areas, and it would be unlikely that different therapies would have an equal effect in all skill areas. The underlying theory of each therapy relates to neuropathology, but it is not possible here to relate therapies and effects to brain or central nervous system function, since it is the outward manifestations which parents observe and report. The following sections deal with differences in perceived benefit, and attempt to identify significant differences between perceived effects in skill areas.

5.6.1 Music therapy

The differences in perceived effect of music therapy are summarised in Table 22. The value for communication was significantly greater than for all other skill areas. Second was the effect on hand function, itself significantly better than motor skill (fourth), but not better than learning skill (third). The effect of music therapy upon learning skill was seen as greater, but not significantly greater, than its effect on motor skill.

<table>
<thead>
<tr>
<th>Rank</th>
<th>Skill area (a)</th>
<th>Skill area (b)</th>
<th>n</th>
<th>W</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>1&gt;2</td>
<td>communication</td>
<td>hand function</td>
<td>15</td>
<td>36</td>
<td>0.0039</td>
</tr>
<tr>
<td>1&gt;3</td>
<td>communication</td>
<td>learning ability</td>
<td>15</td>
<td>66</td>
<td>0.0010</td>
</tr>
<tr>
<td>1&gt;4</td>
<td>communication</td>
<td>motor ability</td>
<td>15</td>
<td>91</td>
<td>0.0001</td>
</tr>
<tr>
<td>2&gt;3</td>
<td>hand function</td>
<td>learning ability</td>
<td>15</td>
<td>22</td>
<td>0.1094</td>
</tr>
<tr>
<td>2&gt;4</td>
<td>hand function</td>
<td>motor ability</td>
<td>15</td>
<td>45</td>
<td>0.0020</td>
</tr>
<tr>
<td>3&gt;4</td>
<td>learning ability</td>
<td>motor ability</td>
<td>15</td>
<td>36.5</td>
<td>0.0645</td>
</tr>
</tbody>
</table>

Sample size (n), Wilcoxon statistic (W) and probability (p)
The findings of this study are consistent with work by leading experts in the field of music therapy (Wigram, 1995, Bruscia, Oldfield, 1995), who all believe that the therapeutic value of music therapy can enhance the ability to communicate. However, these are all experts in their own field, and likely to want to believe in their area of specialism, and it has been seen that unbiased research in the field of music therapy is difficult to achieve (Diamond, 1999). On the other hand, it has been pointed out that uncertainty about approaches should not preclude their use: 'It is important that their potential benefits are researched fully so that there is no premature dismissal of a possibly beneficial approach simply because the evidence is missing' (Jordan, Jones and Murray, 1998, p.120).

Wigram (1995) has stated that music therapy should be on the statement of Special Educational Needs of every child with Rett syndrome and it should be continued as a therapy treatment for this client group for as long as possible, a view echoed by one parent: 'It is a great pity that music therapy is so thin on the ground, and I do feel that research shows that Rett girls should be given greater priority' (Questionnaire 49). It is not known in this particular case why music therapy is thought to be 'thin on the ground'. It may be a reference to lack of availability of qualified music therapists, or there may be an implicit assumption that music therapy is not sufficiently valued to be considered mandatory. Whilst not significant in itself, this comment does highlight the view that children with Rett syndrome need to be given special consideration.

The views of music therapists were not sought, and so the rationale for giving the therapy was not known. It has been seen that one of the rewards of music therapy is pleasure (American Orff-Schulwerk Association, 1999), and there is a possibility that appreciation of a child's pleasure by a parent might influence their perceptions of its value.

5.6.2 Speech therapy

The effect of speech therapy on communication, Table 23, was significantly greater than in all other areas. The effect on learning skill was second, better than motor skill in fourth place but not significantly better than hand function in third place. The effect of speech therapy upon hand function was significantly better than its effect on motor skill.
Table 23 Differences in perceived value of speech therapy on skill areas

<table>
<thead>
<tr>
<th>Rank</th>
<th>Skill area (a)</th>
<th>Skill area (b)</th>
<th>n</th>
<th>W</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>1&gt;2</td>
<td>communication</td>
<td>learning ability</td>
<td>22</td>
<td>51</td>
<td>0.0068</td>
</tr>
<tr>
<td>1&gt;3</td>
<td>communication</td>
<td>hand function</td>
<td>21</td>
<td>75.5</td>
<td>0.0012</td>
</tr>
<tr>
<td>1&gt;4</td>
<td>communication</td>
<td>motor ability</td>
<td>21</td>
<td>103</td>
<td>0.0002</td>
</tr>
<tr>
<td>2&gt;3</td>
<td>learning ability</td>
<td>hand function</td>
<td>21</td>
<td>44.5</td>
<td>0.5270</td>
</tr>
<tr>
<td>2&gt;4</td>
<td>learning ability</td>
<td>motor ability</td>
<td>21</td>
<td>109</td>
<td>0.0017</td>
</tr>
<tr>
<td>3&gt;4</td>
<td>hand function</td>
<td>motor ability</td>
<td>21</td>
<td>52</td>
<td>0.0049</td>
</tr>
</tbody>
</table>

Sample size (n), Wilcoxon statistic (W) and probability (p)

That parents perceive speech therapy to influence communication skills more than any other skill area is not unexpected, although it will be seen later (Table 18) that music therapy and hydrotherapy are thought to have an even greater effect than speech therapy on communication.

Wright and Graham (1997) consider successful communication between speech therapists and teachers to be essential because although it is therapists who assess, plan, and carry out interventions, it is likely to be the teachers who work with the children on a daily basis across the curriculum.

The extent of interaction between speech therapists and teachers is not clear in this study, although the varying roles of the speech therapist has been discussed in Chapter 2 from ‘hands-on’ to enabler (Harris, 1990). Not only may therapists have responsibility for more children than they can cope with, but teachers may prefer to work with children themselves, but under the guidance of the speech therapist. Communication forms an important part of any curriculum and no more so than for children within the autistic spectrum. 'Communication is at the core of the autistic difficulty with learning and it must therefore be at the heart of any effective educational approach with pupils with autism' (Jordan and Powell, 1995, p.70).

5.6.3 Hydrotherapy

The results for hydrotherapy were not so clear, and the scores for communication and motor skill were equal. The perceived effects on hand function in third place were significantly lower than on communication and motor skill, with slightly different values for p, (Table 24). Similarly, the perceived effect of hydrotherapy
was significantly greater on communication and motor skill than it was on learning skill, in fourth place. Finally, there was no observed significant difference between hand function in third and learning skill in fourth order of benefit.

Table 24 Differences in perceived value of hydrotherapy on skill areas

<table>
<thead>
<tr>
<th>Rank</th>
<th>Skill area (a)</th>
<th>Skill area (b)</th>
<th>n</th>
<th>W</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>1a&gt;1b</td>
<td>communication</td>
<td>motor ability</td>
<td>22</td>
<td>75.5</td>
<td>1.0000</td>
</tr>
<tr>
<td>1a&gt;3</td>
<td>communication</td>
<td>hand function</td>
<td>22</td>
<td>110</td>
<td>0.0145</td>
</tr>
<tr>
<td>1b&gt;3</td>
<td>motor ability</td>
<td>hand function</td>
<td>22</td>
<td>69.5</td>
<td>0.0081</td>
</tr>
<tr>
<td>1a&gt;4</td>
<td>communication</td>
<td>learning ability</td>
<td>22</td>
<td>130</td>
<td>0.0047</td>
</tr>
<tr>
<td>1b&gt;4</td>
<td>motor ability</td>
<td>learning ability</td>
<td>22</td>
<td>105</td>
<td>0.0042</td>
</tr>
<tr>
<td>3&gt;4</td>
<td>hand function</td>
<td>learning ability</td>
<td>22</td>
<td>42</td>
<td>0.4250</td>
</tr>
</tbody>
</table>

Sample size (n), Wilcoxon statistic (W) and probability (p)

It might be expected that hydrotherapy would have the same effect on the gross motor ability of the children as physiotherapy, as it has been described: 'Hydrotherapy is simply physiotherapy in water' (Lewis and Wilson, 1998, p.104) although it has been suggested that hydrotherapy is different from but complementary to physiotherapy (Kerr, 1995b). The literature indicates that although they may address the same or similar underlying neurological deficits, they do so using different physical principles. Neither of the therapies can take over the role of the other, and each operates in its own framework. Hydrotherapy could never replace physiotherapy in the treatment of children with neurodevelopmental disorders, but there is evidence that it provides benefits for the whole child through its holistic approach. Kerr (1995b) suggests that the environment of the hydrotherapy pool can be conducive to active learning although in this study, learning ability appeared to be the area considered by parents to benefit least from hydrotherapy.

Although Reid Campion (1997) warns that the effect of the extra noise in a pool, water in the ears and the position of the physiotherapist may compromise communication there was no evidence of this in this study. Reid Campion (1997) suggests that the same physiotherapist should treat the child both in and out of the water. This study does not indicate if this is the case, although there are instances of parents being involved as helpers, providing one-to-one support. The physical
support needed for children with a profound disability in a hydrotherapy pool may help the communication process, although other children within the autistic spectrum may not tolerate this intimate support easily.

5.6.4 Physiotherapy

The therapy's perceived effect on motor skill is significantly greater than all the other skill areas, and the effect on hand function, ranked second, is significantly greater than on learning ability, ranked fourth, Table 25.

<table>
<thead>
<tr>
<th>Rank</th>
<th>Skill area (a)</th>
<th>Skill area (b)</th>
<th>n</th>
<th>W</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>1&gt;2</td>
<td>motor ability</td>
<td>hand function</td>
<td>26</td>
<td>116.5</td>
<td>0.0053</td>
</tr>
<tr>
<td>1&gt;3</td>
<td>motor ability</td>
<td>communication</td>
<td>26</td>
<td>124.5</td>
<td>0.0016</td>
</tr>
<tr>
<td>1&gt;4</td>
<td>motor ability</td>
<td>learning ability</td>
<td>26</td>
<td>149</td>
<td>0.0003</td>
</tr>
<tr>
<td>2&gt;3</td>
<td>hand function</td>
<td>communication</td>
<td>26</td>
<td>69.5</td>
<td>0.1387</td>
</tr>
<tr>
<td>2&gt;4</td>
<td>hand function</td>
<td>learning ability</td>
<td>26</td>
<td>30.5</td>
<td>0.0389</td>
</tr>
<tr>
<td>3&gt;4</td>
<td>communication</td>
<td>learning ability</td>
<td>26</td>
<td>34</td>
<td>0.2496</td>
</tr>
</tbody>
</table>

Sample size (n), Wilcoxon statistic (W) and probability (p)

Physiotherapy was clearly perceived to be more beneficial in helping motor ability than with any other skill areas, and it could be argued, that if physiotherapy is able to offer such benefits that the child's life expectancy is increased then other benefits almost pale in significance. It has been found that the high prevalence of scoliosis and associated joint deformities in Rett syndrome have important implications for the continued physical management of affected girls (Harrison and Webb, 1990), with subsequent interference with aspects of learning. Concentration suffers when comfort levels are low and if a child receives regular physiotherapy which controls the onset of deformity, then other benefits must accrue in areas of mobility, motor control, and general wellbeing.

5.6.5 Other therapies

The perceived benefits of occupational therapy were generally lower than for other therapies. The Wilcoxon test does not detect any differences between benefits applying to each skill area, and since occupational therapy is not commonly in use, other details are not reported here.
Communication is the area which parents considered to be improved by aromatherapy, but there were no significant differences between the therapy's perceived effect on skill areas. Details are not reported here.

The sample sizes for homeopathy, hippotherapy and osteopathy were too small for analysis.

5.7 Correlation between variables

Factors that might affect the value benefit of a therapy cannot be readily quantified, but clearly there are features that could be influential. The skill of the therapist, for example, could be a significant factor, but it is not possible here to consider this as a variable. The actual amount of therapy, in terms of monthly hours and overall length of time it has been given may be influential, and a therapy may be of differing benefit to children with more or less severe disability. The perceived value might be influenced by the parent's desire to see improvement. These relationships are tentatively explored in the following sections. The statistic for osteopathy and hippotherapy could not be computed due to the small sample for these specific therapies, but the remaining therapies are shown in Table 26 to Table 29.

5.7.1 Severity of disabilities and the perceived value of therapies

There are 18 negative and 6 positive correlations in Table 26, the predominance of negatives suggesting that the value of therapy is generally considered to be less with more severely disabled girls.

Table 26 Correlation between the perceived severity of disability and value of therapy

<table>
<thead>
<tr>
<th></th>
<th>Communication</th>
<th>Hand function</th>
<th>Motor skill</th>
<th>Learning ability</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>rs</td>
<td>p</td>
<td>rs</td>
<td>p</td>
</tr>
<tr>
<td>Music therapy</td>
<td>-0.16</td>
<td>0.5578</td>
<td>-0.76</td>
<td>0.0009</td>
</tr>
<tr>
<td>Speech therapy</td>
<td>0.04</td>
<td>0.8538</td>
<td>-0.37</td>
<td>0.1004</td>
</tr>
<tr>
<td>Hydrotherapy</td>
<td>0.12</td>
<td>0.5966</td>
<td>-0.36</td>
<td>0.1019</td>
</tr>
<tr>
<td>Occupational</td>
<td>0.24</td>
<td>0.6082</td>
<td>0.05</td>
<td>0.8982</td>
</tr>
<tr>
<td>Physiotherapy</td>
<td>-0.19</td>
<td>0.3446</td>
<td>-0.20</td>
<td>0.3351</td>
</tr>
<tr>
<td>Aromatherapy</td>
<td>-0.43</td>
<td>0.1460</td>
<td>-0.60</td>
<td>0.0318</td>
</tr>
</tbody>
</table>

Spearman correlation (rs) and probability (p)
These associations are mostly very weak, with the exception of the patterns that relate to specific therapies rather than to skill areas. For example, there are no real strengths of association between communication skills and any of the therapies, but there are evident patterns for music therapy and aromatherapy. The results here suggest that both these therapies are more effective with girls considered to be less severely disabled. There may be an element of tautology here in the sense that if a parent considers, for example, a child to be only mildly disabled in communication, then the possible effect of therapy would be necessarily limited in that particular ability area.

5.7.2 Importance of improving and the perceived value of therapies

There was no variation in score for the importance of improving communication skills and the perceived value of music therapy, therefore analysis was not possible. There are no evident patterns in Table 27 and none of the correlations come close to being significant. There is therefore a wide variation in the ways parents have scored their responses in this area.

It was of possible concern that because of the difficulty of assigning values to characteristics that are hard to measure, parents would see benefits where they wanted to see improvements. When a parent is desperate for a sign, even little incidents can be interpreted as significant effect and therefore attributed to some cause. This did not appear to happen in this study.

Table 27 Correlation between the perceived importance of improving and value of therapy

<table>
<thead>
<tr>
<th></th>
<th>Communication</th>
<th>Hand function</th>
<th>Motor skill</th>
<th>Learning ability</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>rs</td>
<td>p</td>
<td>rs</td>
<td>p</td>
</tr>
<tr>
<td>Music therapy</td>
<td>-</td>
<td>-</td>
<td>0.03</td>
<td>0.9127</td>
</tr>
<tr>
<td>Speech therapy</td>
<td>0.16</td>
<td>0.4721</td>
<td>-0.24</td>
<td>0.3047</td>
</tr>
<tr>
<td>Hydrotherapy</td>
<td>0.07</td>
<td>0.7521</td>
<td>0.05</td>
<td>0.8269</td>
</tr>
<tr>
<td>Occupational</td>
<td>0.34</td>
<td>0.4586</td>
<td>0.12</td>
<td>0.7689</td>
</tr>
<tr>
<td>Physiotherapy</td>
<td>-0.06</td>
<td>0.7726</td>
<td>0.20</td>
<td>0.3286</td>
</tr>
<tr>
<td>Aromatherapy</td>
<td>0.22</td>
<td>0.4671</td>
<td>0.33</td>
<td>0.2649</td>
</tr>
</tbody>
</table>

|                     | rs             | p             |
|---------------------|----------------|
| Motor skill         | -0.28          | 0.3070        |
| Learning ability    | 0.04           | 0.8804        |
| Spearman correlation (rs) and probability (p) | 148 |
5.7.3 **The length of time a therapy has been given**

It was suspected that the longer a therapy had been given, the more appreciated it would be by parents, but Table 28 shows that this did not appear to be the case.

**Table 28 Correlation between length of time therapy has been given and perceived value of therapy**

<table>
<thead>
<tr>
<th></th>
<th>Communication rs</th>
<th>p</th>
<th>Hand function rs</th>
<th>p</th>
<th>Motor skill rs</th>
<th>p</th>
<th>Learning ability rs</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Music therapy</td>
<td>-0.12</td>
<td>0.6804</td>
<td>-0.29</td>
<td>0.2904</td>
<td>-0.11</td>
<td>0.6915</td>
<td>-0.07</td>
<td>0.8135</td>
</tr>
<tr>
<td>Speech therapy</td>
<td>-0.24</td>
<td>0.2844</td>
<td>0.02</td>
<td>0.9399</td>
<td>0.13</td>
<td>0.5619</td>
<td>-0.18</td>
<td>0.4114</td>
</tr>
<tr>
<td>Hydrotherapy</td>
<td>0.08</td>
<td>0.7180</td>
<td>0.08</td>
<td>0.7338</td>
<td>0.02</td>
<td>0.9159</td>
<td>-0.04</td>
<td>0.8461</td>
</tr>
<tr>
<td>Occupational</td>
<td>-0.18</td>
<td>0.7259</td>
<td>-0.65</td>
<td>0.1107</td>
<td>0.53</td>
<td>0.2780</td>
<td>-0.39</td>
<td>0.3902</td>
</tr>
<tr>
<td>Physiotherapy</td>
<td>-0.14</td>
<td>0.5085</td>
<td>0.01</td>
<td>0.9581</td>
<td>0.07</td>
<td>0.7350</td>
<td>0.02</td>
<td>0.9353</td>
</tr>
<tr>
<td>Aromatherapy</td>
<td>-0.02</td>
<td>0.9455</td>
<td>-0.04</td>
<td>0.9085</td>
<td>0.12</td>
<td>0.7053</td>
<td>0.07</td>
<td>0.8291</td>
</tr>
</tbody>
</table>

Spearman correlation (rs) and probability (p)

There were no significant cases, and there were in fact 13 negative correlations fairly evenly distributed among the total of 24. Music therapy generated negative values for all four skill areas, the only therapy to do so, suggesting perhaps that parents may grow to devalue the therapy with time, although the associations were all very weak. Again, the results in Table 28 need to be viewed with a degree of caution.

5.7.4 **Hours per month for therapies**

The only significant correlation is between the level of speech therapy and perceived value for motor ability. This seems an unlikely association, and in view of the great majority of weak associations, this result should be treated with caution. The only other strong relationship was between occupational therapy hours and communication, but there were only three discrete values in this cell, so this result should also be viewed with caution (Table 29).

A study by Reid, Millar, Tait, Donaldson, Dean, Thomson and Grieve (1996) found that although some parents wanted changes such as more individual work, more group work, or better continuity of provision, most parents simply wanted more speech therapy time. Although decisions about the quantity of therapy given to individual children may depend on external factors, the parents may hold the
therapist responsible for what they consider to be shortcomings and undervalue the therapy as a consequence. However in this study, it seems that the number of hours given for any therapy does not influence parents' opinions with respect to the value of the therapy.

Table 29 Correlation between monthly quantity of therapy and perceived value of therapy

<table>
<thead>
<tr>
<th></th>
<th>Communication</th>
<th>Hand function</th>
<th>Motor skill</th>
<th>Learning ability</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>rs</td>
<td>p</td>
<td>rs</td>
<td>p</td>
</tr>
<tr>
<td>Music therapy</td>
<td>0.00</td>
<td>0.9895</td>
<td>0.09</td>
<td>0.7925</td>
</tr>
<tr>
<td>Speech therapy</td>
<td>0.28</td>
<td>0.2861</td>
<td>0.28</td>
<td>0.3204</td>
</tr>
<tr>
<td>Hydrotherapy</td>
<td>0.08</td>
<td>0.7071</td>
<td>-0.14</td>
<td>0.5316</td>
</tr>
<tr>
<td>Occupational</td>
<td>1.00</td>
<td>&lt;0.0001</td>
<td>0.50</td>
<td>0.6667</td>
</tr>
<tr>
<td>Physiotherapy</td>
<td>0.30</td>
<td>0.2031</td>
<td>0.24</td>
<td>0.3137</td>
</tr>
<tr>
<td>Aromatherapy</td>
<td>0.31</td>
<td>0.3787</td>
<td>0.40</td>
<td>0.2552</td>
</tr>
</tbody>
</table>

Spearman correlation (rs) and probability (p)

5.8 Perceived differences in values of therapies within a single skill area

It was expected that there would be some perceived differences in effects of therapies within a single skill area. Physiotherapy, for example, by its name implies a physical benefit, whereas parents possible perceive speech therapy is being primarily directed at improving speech and language. Music therapy, on the other hand, does not imply one specific area of influence and parents may be less likely to be directed in their thinking.

5.8.1 Communication

When the questionnaire results are examined using skill areas as the base, the relative values of therapies can be compared directly. As in the previous analysis, the Wilcoxon test was used with a significance level of 0.05. Table 30 shows that the perceived value of music therapy was significantly greater than the second, third and fourth placed therapies, (hydrotherapy, speech therapy and physiotherapy) but not significantly greater than the fifth and sixth placed.
Table 30 Differences in effect of therapies on communication

<table>
<thead>
<tr>
<th>Rank</th>
<th>Therapy</th>
<th>Therapy</th>
<th>n</th>
<th>W</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>1&gt;2</td>
<td>music therapy</td>
<td>hydrotherapy</td>
<td>13</td>
<td>45</td>
<td>0.0020</td>
</tr>
<tr>
<td>1&gt;3</td>
<td>music therapy</td>
<td>speech therapy</td>
<td>13</td>
<td>21</td>
<td>0.0156</td>
</tr>
<tr>
<td>1&gt;4</td>
<td>music therapy</td>
<td>physiotherapy</td>
<td>14</td>
<td>78</td>
<td>0.0002</td>
</tr>
<tr>
<td>1&gt;5</td>
<td>music therapy</td>
<td>aromatherapy</td>
<td>7</td>
<td>10</td>
<td>0.0625</td>
</tr>
<tr>
<td>1&gt;6</td>
<td>music therapy</td>
<td>occupational therapy</td>
<td>3</td>
<td>6</td>
<td>0.1250</td>
</tr>
<tr>
<td>2&gt;3</td>
<td>hydrotherapy</td>
<td>speech therapy</td>
<td>17</td>
<td>46</td>
<td>0.5000</td>
</tr>
<tr>
<td>2&gt;4</td>
<td>hydrotherapy</td>
<td>physiotherapy</td>
<td>22</td>
<td>127</td>
<td>0.0087</td>
</tr>
<tr>
<td>2&gt;5</td>
<td>hydrotherapy</td>
<td>aromatherapy</td>
<td>13</td>
<td>52</td>
<td>0.0508</td>
</tr>
<tr>
<td>2&gt;6</td>
<td>hydrotherapy</td>
<td>occupational therapy</td>
<td>6</td>
<td>16</td>
<td>0.1563</td>
</tr>
<tr>
<td>3&gt;4</td>
<td>speech therapy</td>
<td>physiotherapy</td>
<td>21</td>
<td>48</td>
<td>0.1201</td>
</tr>
<tr>
<td>3&gt;5</td>
<td>speech therapy</td>
<td>aromatherapy</td>
<td>10</td>
<td>15</td>
<td>0.0313</td>
</tr>
<tr>
<td>3&gt;6</td>
<td>speech therapy</td>
<td>occupational therapy</td>
<td>5</td>
<td>6</td>
<td>0.1250</td>
</tr>
<tr>
<td>4&gt;5</td>
<td>physiotherapy</td>
<td>aromatherapy</td>
<td>13</td>
<td>18</td>
<td>0.5273</td>
</tr>
<tr>
<td>4&gt;6</td>
<td>physiotherapy</td>
<td>occupational therapy</td>
<td>7</td>
<td>13</td>
<td>0.4219</td>
</tr>
<tr>
<td>5&gt;6</td>
<td>aromatherapy</td>
<td>occupational therapy</td>
<td>4</td>
<td>0</td>
<td>1.000</td>
</tr>
</tbody>
</table>

Sample size (n), Wilcoxon statistic (W) and probability (p)

It would be reasonable to expect that these latter differences would also be significant, since they are arithmetically greater, but the sensitivity of the test has been reduced by small sample sizes. The small samples occurred because there were few girls who received the two specific therapies in each case. This effect occurs repeatedly through the skill-area based analysis, and the differences in sample sizes mean that comparisons of p values should be made with caution.

It appears that music and hydrotherapy are seen as the therapies having the greatest effect in addressing communication difficulties, with speech therapy, traditionally used for assisting with problems of communication (Lees and Unwin, 1991) appearing less favourable.

There are suggestions in the literature that the richest communication for a girl with Rett syndrome is likely to be through means other than speech, and that therapies other than speech therapy may also be beneficial for communication (Kerr, 1997, Wigram, 1995 and Etkin and Eisler, undated). The findings in this study support the
view that music and hydrotherapy are seen by parents as helpful in addressing communication difficulties.

Storr elaborates on the importance of music as an alternative to speech:

It isn’t really surprising that music provides an alternative to speech as a means of communication. When mothers talk to their infants, the meaning of the words which they employ is unimportant; but the changes in pitch, stress, rhythm, speed, and volume which characterise a mother’s utterance convey a message of concern and love to which the infant responds with gurgling pleasure.

(Storr, 1993, p.xi)

It is unlikely, however, that some form of language intervention programme has not been delivered to the children in this study, and it has been seen that, given support, people without specialist qualifications can play an important part in this delivery. Lack of communication between therapist, teachers and parents can lead to misunderstandings and the complementary role of the speech therapist and that of the teacher need to be clearly discussed with parents.

5.8.2 Hand function

Table 31 shows that the perceived beneficial effect on hand function of music therapy, while close to being significant, is not significantly greater than the second placed therapy (physiotherapy), but the differences between first and third / fourth places are strongly significant. Physiotherapy, ranked second, has an effect which is significantly better than hydrotherapy, speech therapy and aromatherapy. It should be remembered that within Rett syndrome, improvement in hand function may relate more to a reduction in stereotypies than to an increase in controlled movement.

Music therapy and physiotherapy both impact upon hand movements, and it is reasonable to suppose that the difference between them is not significant because the scores reflect an actual benefit perceived by respondents for both therapies.
Physiotherapy is concerned with both gross and fine motor movements, and so it is not surprising that there is a positive input in the management of stereotypical hand movements. The skilled physiotherapist will provide the parents with information and a range of options, but will understand that the decision making lies ultimately with the parents (Cunningham and Davis, 1985). Continual involuntary hand movement may have implications in the classroom, so the issue is not straightforward and the views of the teacher should also be considered.

5.8.3 Motor ability

Highly significant differences are shown in Table 32 between physiotherapy, the highest ranked therapy for motor ability, and the therapies ranked 2 to 5. The difference between physiotherapy and occupational therapy, the lowest ranked, is almost significant, but the result is desensitised by the small sample size. This overall result is rather as expected, since the primary aim of physiotherapy is to address problems with motor ability (Hanks, 1990). The therapy ranked second, hydrotherapy, is also designed to improve muscle control and motor ability, and is
perceived to be more beneficial than the remaining therapies, with some very high levels of significance. When considering third rank and below, there are virtually no significant differences in perceived effect, probably because of the smaller sample sizes, but possibly because therapies such as aromatherapy and speech therapy really are perceived to be of equally little value in assisting deficits in motor ability.

Table 32 Differences in effect of therapies on motor ability

<table>
<thead>
<tr>
<th>Rank</th>
<th>Therapy</th>
<th>Therapy</th>
<th>n</th>
<th>W</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>1&gt;2</td>
<td>physiotherapy</td>
<td>hydrotherapy</td>
<td>22</td>
<td>38</td>
<td>0.0371</td>
</tr>
<tr>
<td>1&gt;3</td>
<td>physiotherapy</td>
<td>music therapy</td>
<td>14</td>
<td>45</td>
<td>0.0020</td>
</tr>
<tr>
<td>1&gt;4</td>
<td>physiotherapy</td>
<td>aromatherapy</td>
<td>13</td>
<td>45</td>
<td>0.0020</td>
</tr>
<tr>
<td>1&gt;5</td>
<td>physiotherapy</td>
<td>speech therapy</td>
<td>20</td>
<td>136</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>1&gt;6</td>
<td>physiotherapy</td>
<td>occupational therapy</td>
<td>7</td>
<td>10</td>
<td>0.0625</td>
</tr>
<tr>
<td>2&gt;3</td>
<td>hydrotherapy</td>
<td>music therapy</td>
<td>13</td>
<td>55</td>
<td>0.0010</td>
</tr>
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<td>aromatherapy</td>
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<td>speech therapy</td>
<td>17</td>
<td>91</td>
<td>0.0001</td>
</tr>
<tr>
<td>2&gt;6</td>
<td>hydrotherapy</td>
<td>occupational therapy</td>
<td>6</td>
<td>1</td>
<td>0.5000</td>
</tr>
<tr>
<td>3&gt;4</td>
<td>music therapy</td>
<td>aromatherapy</td>
<td>7</td>
<td>2</td>
<td>0.8750</td>
</tr>
<tr>
<td>3&gt;5</td>
<td>music therapy</td>
<td>speech therapy</td>
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<td>15</td>
<td>0.0313</td>
</tr>
<tr>
<td>3&gt;6</td>
<td>music therapy</td>
<td>occupational therapy</td>
<td>3</td>
<td>0</td>
<td>1.0000</td>
</tr>
<tr>
<td>4&gt;5</td>
<td>aromatherapy</td>
<td>speech therapy</td>
<td>10</td>
<td>5</td>
<td>0.3750</td>
</tr>
<tr>
<td>4&gt;6</td>
<td>aromatherapy</td>
<td>occupational therapy</td>
<td>4</td>
<td>0</td>
<td>1.0000</td>
</tr>
<tr>
<td>5&gt;6</td>
<td>speech therapy</td>
<td>occupational therapy</td>
<td>5</td>
<td>1</td>
<td>0.5000</td>
</tr>
</tbody>
</table>

Sample size (n), Wilcoxon statistic (W) and probability (p)

The findings in this study are compatible with the literature which suggests that both physiotherapy and hydrotherapy are therapies primarily concerned with physical ability. Kerr (1996a) believes that they are '... of primary importance both in order to develop full potential and because lack of stimulation and use of the disabled mind and body plays a major role in producing secondary disability' (p.2).

5.8.4 Learning ability

Table 33 shows the perceived effects of therapies on learning ability, and it is evident that there are fewer significant differences in effects in this area than in the other three skill areas. This may be because therapies are generally devised to
address a specific feature of physical deficit, whereas learning skill is more an intellectual characteristic, and therapies examined here may in reality have little effect. An alternative consideration might be that parents and carers find the term 'learning ability' difficult to define, and so assessments are likely to vary between parents who themselves may have different intellectual capabilities.

Table 33 Differences in effect of therapies on learning ability

<table>
<thead>
<tr>
<th>Rank</th>
<th>Therapy</th>
<th>Therapy</th>
<th>n</th>
<th>W</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>1&gt;2</td>
<td>music therapy</td>
<td>speech therapy</td>
<td>13</td>
<td>13</td>
<td>0.4219</td>
</tr>
<tr>
<td>1&gt;3</td>
<td>music therapy</td>
<td>physiotherapy</td>
<td>14</td>
<td>19</td>
<td>0.2344</td>
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<td>hydrotherapy</td>
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<td>1&gt;5</td>
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<td>aromatherapy</td>
<td>7</td>
<td>10</td>
<td>0.0625</td>
</tr>
<tr>
<td>1&gt;6</td>
<td>music therapy</td>
<td>occupational therapy</td>
<td>3</td>
<td>1</td>
<td>0.5000</td>
</tr>
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<td>physiotherapy</td>
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<td>42</td>
<td>0.4250</td>
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<td>0.0156</td>
</tr>
<tr>
<td>2&gt;6</td>
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<td>6</td>
<td>0.1250</td>
</tr>
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<td>3&gt;4</td>
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<td>hydrotherapy</td>
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<td>0.4197</td>
</tr>
<tr>
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<td>aromatherapy</td>
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<td>19</td>
<td>0.0781</td>
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<td>occupational therapy</td>
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<td>15</td>
<td>0.2813</td>
</tr>
<tr>
<td>4&gt;5</td>
<td>hydrotherapy</td>
<td>aromatherapy</td>
<td>13</td>
<td>6</td>
<td>0.1250</td>
</tr>
<tr>
<td>4&gt;6</td>
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<td>occupational therapy</td>
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<td>8</td>
<td>0.3125</td>
</tr>
<tr>
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<td>aromatherapy</td>
<td>occupational therapy</td>
<td>4</td>
<td>0</td>
<td>1.0000</td>
</tr>
</tbody>
</table>

Sample size (n), Wilcoxon statistic (W) and probability (p)

In a sense, judgmental values and development objectives are redefined as the manifestation of the Rett disorder appears and comes to be understood by parents, and one person's concept of learning skill may be different to that of another. The difference between music therapy and hydrotherapy, ranked 1 and 4 for their effect on learning ability, is only just significant at the 5% level.

A search of the literature showed little evidence that learning ability can be directly enhanced by the use of therapies, although the DfEE (1997) accepts that therapies may benefit a child's educational progress, even if provided for health reasons. So if, for example, physiotherapy ensures the prolonged physical wellbeing of a child,
enabling her to attend school on a regular basis, then it could be argued that this is contributing to her ability to learn. One parent commented 'Music is a key to a locked door as far as Maria is concerned' (Questionnaire 21). In this sense the therapy is a facilitator, a vehicle for learning which should be as much an essential part of daily routine as the learning process itself.

5.9 **Summary**

The role of therapists was not defined to parents in this study, neither is the degree of interaction between teachers and therapists known. Parents may not be aware of the commonality of purpose in this respect, and just as there might be a difficult distinction between the roles, parents might not appreciate that in some cases teachers are working within the framework set out by the therapist. It is a weakness in this study that parents are assumed to accept a clear distinction between the two.

However, in all of these analyses, an important point to remember is that the results reflect parents’ opinions of the effects of therapy, they do not represent an objective investigation of actual effects. Nor does a fixed relationship between score values exist for all respondents, who may differ in their assignment of actual numbers to a concept of benefit which is at best difficult to assess. There is also the issue of preconception. For example, when assessing skill areas, there is possibly a bias in reporting, a bias that emphasises a therapy known to be designed to assist specific problems (and perhaps even named accordingly) in respect of skill areas. Similarly, aims, objectives and delivery techniques of therapies such as occupational therapy may not be well understood by parents, who subconsciously adjust their scores depending upon their actual understanding.

Parents want what is best for their children, and if no hard evidence is available, their opinions will be based on their interpretations of what they see and may be easily influenced by opinions of others. ‘They may lack good information on the value of these approaches and so be vulnerable to claims for success’ (Jordan, Jones and Murray, 1998, p.9).

In real life situations there is probably no way to overcome these limitations, because therapies cannot be given anonymously, and severely disabled children cannot ethically be subject to experimental methods which for some may jeopardise the opportunity for effective help. These finding must also be viewed in an
Chapter 6 Interview Data

6.1 Introduction

The following interview data is based on telephone interviews with eight families of girls with Rett syndrome (Appendix VI). All parents had previously completed questionnaires, and had indicated that they were prepared to take part in further research. All the girls received a minimum of four therapies, including music therapy, speech therapy, physiotherapy and hydrotherapy, and it is the perceived effect of these four therapies, related to the main areas of disability associated with Rett syndrome (communication, hand function, mobility and learning ability) which are considered in this section.

It has been seen (Chapter 3) that interview response data were counted and the results collated and tabulated. Open-ended questions were favoured in order to allow greater flexibility of response, although in retrospect more structure could have been given to the interview schedules. This would have allowed more precision in the responses, enabled more accurate conclusions to be drawn and added validity to the research.

6.2 Developmental issues

Parents who agreed to take part in the interviews were asked what first caused them to think that their daughter was not progressing in the expected manner, and at what age they first noticed this lack of development (Table 34). All of these cases conform to the classic Rett syndrome diagnosis, in which girls are born apparently healthy and begin to regress within the first eighteen months.

The diagnosis of Rett syndrome relies to a large extent on the observations of the parents. In every case parents were aware that their child was not developing normally by the age of 18 months, with the average ‘realisation’ at approximately 12 months. The average age of actual diagnosis was 3 years 6 months. So, typically a family might have been in a state of uncertainty for over two and a half years, living either with no diagnosis, or an incorrect diagnosis. This was particularly difficult for five families, evident from comments such as - ‘I couldn’t convince the doctors that this was not normal’ (Interview 4) and ‘The doctors didn’t know what was wrong’ (Interview 7). Then, the diagnosis itself was equally distressing. Two parents admitted to being ‘devastated’ and ‘crying for days’. Studies by Hunter
(1987) and Goodship (1987) reveal that these initial reactions are not unusual in families with a daughter with Rett syndrome, and Klauber (1999) found a similar impact on families of children with autism. Personal experience has shown that some parents, whilst able to discuss practical elements of their child’s disability, still find it difficult to talk about the emotional impact it has had on their lives.

Table 34. Developmental issues of girls with Rett syndrome

<table>
<thead>
<tr>
<th>No.</th>
<th>Failure to reach milestones</th>
<th>Other traits e.g. too floppy</th>
<th>Apparently precipitated by illness</th>
<th>Wrong or late diagnosis</th>
<th>Age when delay was realised</th>
<th>Age at diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>★</td>
<td></td>
<td>★</td>
<td></td>
<td>12 months</td>
<td>5 years</td>
</tr>
<tr>
<td>2</td>
<td>★</td>
<td></td>
<td>★</td>
<td></td>
<td>10 months</td>
<td>5 years</td>
</tr>
<tr>
<td>3</td>
<td>★</td>
<td></td>
<td></td>
<td></td>
<td>6 months</td>
<td>4 years</td>
</tr>
<tr>
<td>4</td>
<td>★</td>
<td>★</td>
<td>★</td>
<td></td>
<td>12 months</td>
<td>2 years</td>
</tr>
<tr>
<td>5</td>
<td>★</td>
<td></td>
<td></td>
<td></td>
<td>11 months</td>
<td>6 years</td>
</tr>
<tr>
<td>6</td>
<td>★</td>
<td>★</td>
<td>★</td>
<td></td>
<td>8 months</td>
<td>2 years</td>
</tr>
<tr>
<td>7</td>
<td>★</td>
<td></td>
<td></td>
<td></td>
<td>18 months</td>
<td>3 years</td>
</tr>
<tr>
<td>8</td>
<td>★</td>
<td></td>
<td></td>
<td></td>
<td>18 months</td>
<td>2 years</td>
</tr>
</tbody>
</table>

It has been seen (Section 5.3) that some professionals do not demonstrate consideration for parents’ feelings, nor do they consider themselves to be responsible for issues beyond the diagnoses and treatment. Cunningham and Davis (1985) refer to counselling as a skill which should be fundamental in all professionals, and suggest that it is never more important than when a paediatrician gives notification of a child's medical condition.

Three of the eight parents interviewed thought that an illness had triggered the problems that ultimately led to the diagnosis of Rett syndrome, with an observation from one parent: ‘Debbie was progressing normally. She could stack toys and was learning some words. Then she had measles at 11 months. This seemed to trigger Rett syndrome. Everything she had learnt just went’ (Interview 4). This belief is not uncommon, and Kerr (1994d) found that regression takes many forms including apparent precipitation by illness. It has been suggested that Rett syndrome may be due to a slow virus state (Hagberg, Anvret, Percy and Wahlström, 1993) and one parent seemed to give identity to the syndrome with the comment ‘It’s as if it’s lying
in the body waiting for something to happen' (Interview 7), although this opinion is not representative of the sample.

6.3 Extent of disability

Parents were asked what they considered to be their daughter’s greatest overall difficulties. At this stage they were not asked to refer specifically to the four main areas of disability specified in the essential criteria of Rett syndrome, and Table 35 shows the wide range of problems mentioned. Hunter (1987) found that parents’ views about specific symptoms differ with the age of the child and the severity of the symptoms, and so the range of perceived difficulties was not unexpected.

Neither mental deficiency, nor hand stereotypies, both necessary criteria for Rett syndrome were mentioned by parents. Four parents considered conditions relating to physical disability as being the most difficult for their daughter, with the thought of deformity caused by scoliosis, being a cause for concern in one case. Two parents mentioned communication difficulties.

<table>
<thead>
<tr>
<th>No.</th>
<th>Epilepsy</th>
<th>Poor mobility</th>
<th>Inability to communicate</th>
<th>Scoliosis</th>
<th>Poor sleep patterns</th>
<th>Incontinence</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>★</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2</td>
<td></td>
<td>★</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td></td>
<td></td>
<td>★</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>4</td>
<td>★</td>
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<td>★</td>
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<td></td>
<td></td>
<td>★</td>
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<tr>
<td>7</td>
<td>★</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>★ ★</td>
</tr>
<tr>
<td>8</td>
<td></td>
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<td></td>
<td></td>
<td>★</td>
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</tr>
</tbody>
</table>

Epilepsy is a supportive rather than necessary criterion, yet three parents considered it to be the greatest problem for their daughter, with one parent saying ‘This interferes with all her activities, although it now seems to be controlled by the use of drugs’ (Interview 1). The extent of this perceived problem may be because of the stigma attached to epilepsy, taken in a relative context, for epilepsy is frequently controllable under drug therapy. It is likely that what is considered a
priority at one time, may be less important later, depending on the physical health of the child at the time. If a child is so ill that her life may be at risk, then her inability to walk, for example, may be thought less important than during periods of good health. Certainly, this aspect of the condition should be considered when proposing therapy, as the effects of epilepsy undoubtedly impinge upon a child’s overall wellbeing and ability to respond to treatment. There is a need for education and therapy staff to understand medical implications of conditions. Staff need to be aware that some parents become very knowledgeable about aspects of their child’s disability, both generally, and individually, and should be given the opportunity to discuss their opinions.

6.4 Physical aspects

Table 36 shows parents’ views on the current physical needs and abilities of their daughters. Degrees of mobility and spinal deformation were the issues of concern. Only two of the eight girls were able to walk completely independently, with one deteriorating and already using a wheelchair. Three girls were able to walk with support, all for short distances only. Three girls were unable to walk even with help.

<table>
<thead>
<tr>
<th>No.</th>
<th>Mobile</th>
<th>Mobile with help</th>
<th>Not mobile</th>
<th>Scoliosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>★</td>
<td></td>
<td></td>
<td>★</td>
</tr>
<tr>
<td>2</td>
<td></td>
<td>★</td>
<td></td>
<td></td>
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<tr>
<td>3</td>
<td>★</td>
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<td>4</td>
<td></td>
<td>★</td>
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<td>5</td>
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<td>★</td>
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<tr>
<td>6</td>
<td></td>
<td>★</td>
<td>★</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>deteriorating</td>
<td></td>
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<td></td>
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<tr>
<td>8</td>
<td></td>
<td>★</td>
<td>★</td>
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</tbody>
</table>

It has become clear over the course of time that spinal deformity in girls with Rett syndrome is not uncommon (Webb, 1996). None of the girls studied were over twelve years of age, and scoliosis was already apparent in three cases. Whereas the profound mental disabilities evident at regression remain essentially unchanged, physical disabilities, scoliosis and lower limb deformities tend to
increase with age (Harrison and Webb, 1990). A further study to monitor the incidence of scoliosis would reveal if the problem had indeed become more prevalent with age.

6.4.1 Physical aspects – priorities

Most children within the autistic spectrum do not have a physical difficulty, whereas for parents of girls with Rett syndrome, this may be an area of considerable concern, bearing in mind the potential level of degeneration attached to the syndrome. Parents were asked to suggest priorities for the future and Table 37 shows their greatest concerns.

<table>
<thead>
<tr>
<th>No.</th>
<th>Improve mobility</th>
<th>Aid general wellbeing</th>
<th>Maintain present skills</th>
<th>Control scoliosis</th>
<th>Aid future management</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>★</td>
<td>★</td>
<td>★</td>
<td>★</td>
<td>★</td>
</tr>
<tr>
<td>2</td>
<td>★</td>
<td>★</td>
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<td>★</td>
<td>★</td>
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<td>3</td>
<td>★</td>
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<td>★</td>
</tr>
</tbody>
</table>

In six cases, maintaining or improving current levels of mobility were seen as a priority. It is interesting to note that in two interviews, the need to keep the effects of scoliosis to a minimum was recognised, although at the time the girls were not suffering from this aspect of the condition. This indicates that some parents are aware of the progression of the disability and highlights the concerns of parents for the future.

It could be argued that all parents should be alerted to the expected progression of the condition, in order to be in a position to make informed decisions about their daughter’s treatment. However, it is uncertain how parents will react to this news, and it is unclear whose responsibility it is to pass on such information. Just as consideration to feelings needs to be given during initial confirmation of diagnosis,
so must further information be given 'sympathetically, but with a balanced honest appraisal of implications which do not just list negative aspects' (Cunningham and Davis, 1985, p.55) and with an assurance of continuing support.

There is little hope of a 'cure' in the immediate future for children within the autistic spectrum, and informed parents who have chosen to learn about Rett syndrome will be aware of its regressive nature, and will find it difficult not to be concerned for the years to come. Five parents mentioned future physical management of the girls as a cause for concern. They felt that the ability of the girls to walk or take weight would help carers with management in later life.

To keep supple and to keep the mobility she already has - that is really important. We don't want her to have a wheelchair in the house. This is really important for parents as well as for the children. It takes up a lot of space and makes her look really handicapped. It would be heavy to move and so would Samantha be and that would make it difficult for everything.

(Interview 3)

In this kind of situation the physiotherapist has two issues to deal with, the disability of the child and the needs of the parent. How these related needs are managed may influence the parent / therapist relationship and subsequently have future bearing on the perceived value of the therapy itself. There must be a genuine understanding of the parents’ needs; for example, it may be possible for school staff to manage the physical needs of child in a situation that it is not able to replicate in the home, and the additional pressures of parents must be recognised.

6.4.2 Physical aspects – perceived value of therapies

It was already known that in all eight cases the children were receiving a minimum of four therapies. Not all of these, however, were thought to help with all aspects of their child’s development. Parents were invited to comment on the value of all therapies used to aid physical improvement, although only the four most common therapies (physiotherapy, hydrotherapy, music therapy and speech therapy) were used comparatively.

Table 38 summarises the key points of the perceived value of therapies in assisting gross motor skills.
Six parents believed that more than one therapy contributed towards assisting physical difficulties. In all eight cases, physiotherapy was thought to help with physical needs. Neither music therapy nor speech therapy was mentioned in the therapies thought to be helpful in improving the gross motor skills of the child. Discussions with parents at annual meetings to review a child’s statement have supported the view that for children with a physical disability, physiotherapy is a priority.

6.5 Communication

Preliminary research in this study has shown that communication was a key issue for many parents, and they were asked to indicate in greater depth the ways in which their daughters were able to communicate (Table 39). Parents were not asked about particular communicative functions, as it was thought this might
influence the response, although in retrospect there would have been more precision if this aspect had been included in the interviews.

Table 39 Means of communication

<table>
<thead>
<tr>
<th>No.</th>
<th>Hovers nearby</th>
<th>Uses her eyes</th>
<th>Facial expression</th>
<th>Words, noises</th>
<th>Body language</th>
<th>Grabs or touches</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>★</td>
<td>★</td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>2</td>
<td>★</td>
<td>★</td>
<td></td>
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<tr>
<td>3</td>
<td>★</td>
<td>★</td>
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<td>4</td>
<td>★</td>
<td>★</td>
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<td>7</td>
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<td>8</td>
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</tbody>
</table>

The loss of previously acquired communication skills is a well-documented prominent feature of Rett syndrome and is considered by some to be one of the most significant disabilities of all (Jones and Creegan, 1986). As a child with Rett syndrome leaves the regression stage, so her ability to communicate improves. In all eight cases, the parent or carer reported that the girl was able to communicate through eye contact. Comments such as 'Harriet has amazing eye contact. I can tell from her eyes if she is hurting or is unhappy' (Interview 5) and 'The mother can tell if she is happy, unhappy, upset, agitated or excited by looking into her eyes' (Interview 6) demonstrate the widely held view among parents that the child understands more than she can demonstrate (Lindberg, 1994). Food seemed to be a powerful motivator, with four parents mentioning the attraction of food encouraging eye pointing or intense staring.

Whereas lack of eye contact, or inappropriate use of eye contact is a feature of the behaviour of children with autism which causes parents to become discouraged in their attempts to communicate (Davis, 1997), girls with Rett syndrome often display an intense eye contact that has the opposite effect, sustaining a belief in some kind of hidden understanding which encourages and gives hope to parents. The difference in condition contrasts with the similarity in parental emotion, both sets of
parents valuing the ability to communicate, to some extent at least, through eye contact whether it is actually present or not.

6.5.1 Communication - with others

All children were able to communicate to some degree with those close to them. Table 40 shows how well parents thought their daughters were able to communicate with people other than family or close friends.

Table 40 Ability to communicate with outsiders

<table>
<thead>
<tr>
<th>No.</th>
<th>Limited</th>
<th>Able</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>★</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>★</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>★</td>
<td></td>
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<tr>
<td>4</td>
<td>★</td>
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<tr>
<td>7</td>
<td>★</td>
<td></td>
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<tr>
<td>8</td>
<td>★</td>
<td></td>
</tr>
</tbody>
</table>

Considering the recognised ability of girls with Rett syndrome to communicate, it is interesting to note that none of the girls were thought to be able to communicate easily with people other than family or close friends, but also, none were thought to be entirely without communication. It has been seen that eye contact is one of the most important early communications between mother and baby (Latham, 1984) and it may be that parents wish to hold onto that, as so much has been lost in other areas. Eye contact relies to a large extent upon its interpretation by others, and it is possible that parents apply their own interpretation of eye use of their children.

Those involved in the care of a girl with Rett syndrome can help this communication, by protecting her long term relationships with the people who matter most to her, by making time for undemanding face to face interaction and by learning to use and respond to non-speech vocalisation. It may be that girls are attempting to communicate with others, and that casual contacts need to learn how to interpret their signals, with one parent believing: 'It is easy to miss things she is looking at if you are not expecting it' (Interview 7). This is a good reason to aim for
consistency in daily routine, and the value of consistency should be acknowledged and addressed in an educational environment in order that the greatest possible opportunity for learning can take place (Ouvry, 1987).

It can be seen that communication ability of girls with Rett syndrome is something of an enigma, and it therefore becomes vital that everyone concerned works together in order to learn how best to communicate.

6.5.2 Communication - priorities

Parents were clear about the needs of their daughters, but Table 41 shows they were less clear about how to address these issues, with four parents unable to offer suggestions about how they should best be addressed.

<table>
<thead>
<tr>
<th>No.</th>
<th>Priorities</th>
<th>How addressed</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>yes / no choice</td>
<td>keep existing skills</td>
</tr>
<tr>
<td>1</td>
<td>★</td>
<td>★</td>
</tr>
<tr>
<td>2</td>
<td>★</td>
<td>★</td>
</tr>
<tr>
<td>3</td>
<td>★</td>
<td>★</td>
</tr>
<tr>
<td>4</td>
<td>★</td>
<td></td>
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<tr>
<td>5</td>
<td>★</td>
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<tr>
<td>6</td>
<td>★</td>
<td></td>
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<tr>
<td>7</td>
<td>★</td>
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<tr>
<td>8</td>
<td>★</td>
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</tbody>
</table>

Seven of the eight cases studied mentioned the ability to choose as a priority, with the development of eye pointing as a means of achieving this in 3 instances. Research has shown that this is within the expectation of girls with Rett syndrome. Lindberg (1994) found that given time and opportunity to choose, most girls would be able to make a choice. The means of expression might vary, but for many, particularly those girls unable to use their hands, eye pointing will be preferred. Children with autism also find it difficult to make meaningful choices (Jordan and Powell, 1995) and this skill will need to be taught as part of a communication programme.
6.5.3 Communication - perceived value of therapies

Parents were asked to name specific therapies that have been used to assist with communication difficulties and to specify in which ways they had been beneficial. Their views are summarised in Table 42.

Speech therapy has traditionally been linked to communication difficulties, and it might be expected that all parents would consider this particular therapy to be beneficial and five of the eight parents thought speech therapy helped their daughters. Only one parent mentioned the therapists' role in giving advice. It is not known to what extent therapists have liaised with class teachers in this study, who in turn may have advised parents about communication. Personal experience has shown that if parents have confidence in the general ability of the class teacher, they are more likely to recognise the enabling role of the speech therapist, and accept the class teacher having overall responsibility for the child's communication skills.

<table>
<thead>
<tr>
<th>No.</th>
<th>Speech therapy</th>
<th>Music therapy</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>advice from therapist</td>
<td>helps with eye contact, choice and co-operation</td>
</tr>
<tr>
<td>2</td>
<td>helps with choosing</td>
<td>helps with choosing</td>
</tr>
<tr>
<td>3</td>
<td>encourages choice</td>
<td>gives opportunity to choose</td>
</tr>
<tr>
<td>4</td>
<td></td>
<td>helps with choice and turn taking</td>
</tr>
<tr>
<td>5</td>
<td>helps with choosing</td>
<td>fun, gives choices, relaxes</td>
</tr>
<tr>
<td>6</td>
<td></td>
<td>encourages vocalising</td>
</tr>
<tr>
<td>7</td>
<td>helps with eye contact</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td></td>
<td>helps focus on sounds</td>
</tr>
</tbody>
</table>

Seven parents found music therapy to be beneficial to their children. A point was raised about the validity of the term 'therapist'. Whilst accepting that her child responded well to music, one mother queried the validity of calling the treatment a therapy, when the session was not conducted by a qualified therapist. It would have added validity to the study if the professional status of the person delivering
the therapy was known. The role of the physiotherapist and the speech therapist as enablers has been discussed (Harris, 1990, Levitt, 1984), but there is little in the literature to suggest that music therapists pass on their skills to allow others to implement programmes. There is clearly an issue regarding what constitutes music therapy as distinct from simply experience of music, and the role of the teacher in this situation is discussed in Chapter 7.

It is interesting to note the diversity of values, with eight different benefits being mentioned for music therapy, but only three for speech therapy, and this may be an example of the name of the therapy suggesting the purpose to the parent, rather than a real effect. References to benefits other than the four main areas of communication, hand function, mobility and learning disability mentioned improved co-operation, fun and relaxation as benefits of music therapy. Much of the literature regarding Rett syndrome and aimed specifically at parents mentions the diverse value of music therapy (Montague, 1988, Wigram, 1995) and this may have affected opinions.

6.6 Hand function

As the disappearance of purposeful, voluntary hand use constitutes an important manifestation of Rett syndrome (Witt-Engerström, 1990), parents were asked to talk about the way their daughters used their hands (Table 43).

Although four children were thought to reach out purposefully with their hands, and seven were able to pick up, only two girls were able to finger feed. At regression this skill commonly deteriorates and is seldom recovered (Kerr, 1994b). This regression in oral skills leads to impairment of chewing ability in girls with Rett syndrome, such that the subsequent failure to manage food in the mouth is a recognised characteristic (Morton, Bonas, Minford, Kerr and Ellis, 1997).

There appears to have been little research on what constitutes cause - whether this is linked to the girls’ inability to put food into their mouths, or whether their inability to do so leads to regression in oral skills. The roles of the therapists in helping with feeding programmes were not known.
Table 43 Current hand function

<table>
<thead>
<tr>
<th>No.</th>
<th>Picks up</th>
<th>Mouths</th>
<th>Holds / grasps</th>
<th>Finger feeds</th>
<th>Throws</th>
<th>Hits out or swipes</th>
<th>Reaches out</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>★</td>
<td>★</td>
<td>★</td>
<td></td>
<td>★</td>
<td>★</td>
<td>★</td>
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<td>2</td>
<td>★</td>
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<tr>
<td>3</td>
<td>★</td>
<td>★</td>
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<td>★</td>
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</tbody>
</table>

6.6.1 Hand function - stereotypical behaviour

Children with autism show signs of stereotyped behaviour, and these may involve either hand or finger flapping or twisting, or complex whole body movements (WHO, 1993). However, it is the stereotyped midline hand movements (such as handwringing or “hand-washing”) that are so characteristic of Rett syndrome. It was expected, therefore, that all children would have unconventional hand behaviours and Table 44 shows parents’ observations of these.

Table 44 Stereotypical hand behaviours

<table>
<thead>
<tr>
<th>No.</th>
<th>Wringing</th>
<th>Mouthing</th>
<th>Opening, closing</th>
<th>Tapping, clapping</th>
<th>Plucking clothes</th>
<th>Flapping, waving</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td></td>
<td></td>
<td>★</td>
<td></td>
<td></td>
<td>★</td>
</tr>
<tr>
<td>2</td>
<td></td>
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<td>★</td>
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<td>3</td>
<td>★</td>
<td>★</td>
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<tr>
<td>4</td>
<td></td>
<td></td>
<td>★</td>
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<td>★</td>
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</tbody>
</table>

It is during the period of regression, when acquired hand skills and the ability to use hands functionally are lost, that typical hand stereotypies manifest themselves.
They may include handwringing, squeezing, washing, patting, rubbing, mouthing or tongue pulling. It was not surprising, given the variations of hand movements possible, that the movements reported took many varied forms, with a clear description from one parent of an intricate hand movement pattern. ‘She has a different, detailed movement for each hand. This is very complex and involves opening and closing each hand, with thumbs in different but specific places and at different speeds’ (Interview 4). Discussions with parents have indicated that they show what is almost a pride in their daughters’ abilities to use their hands in such a way.

6.6.2 Hand function - modification of hand movements

It has been seen that opinion over the use of restraint for modifying hand movements is divided (Kerr, 1991, Budden, 1995, Rett, 1985b). Parents were asked if they felt that it was important to restrain or modify the stereotypical hand movements of their children (Table 45).

Table 45. The importance of modifying hand movements

<table>
<thead>
<tr>
<th>No.</th>
<th>Important</th>
<th>Not important</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td></td>
<td>★</td>
</tr>
<tr>
<td>2</td>
<td></td>
<td>★</td>
</tr>
<tr>
<td>3</td>
<td></td>
<td>★</td>
</tr>
<tr>
<td>4</td>
<td></td>
<td>★</td>
</tr>
<tr>
<td>5</td>
<td></td>
<td>★</td>
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<tr>
<td>6</td>
<td></td>
<td>★</td>
</tr>
<tr>
<td>7</td>
<td></td>
<td>★</td>
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<tr>
<td>8</td>
<td></td>
<td>★</td>
</tr>
</tbody>
</table>

All parents were in full agreement that modifying hand movements was unnecessary, and seven parents actually felt that it was important that hands were not restrained in any way, with a typical comment: ‘No, I would never agree to this. It is part of them, it is what they need to do’ (Interview 6). Five parents agreed with Rett, that this stereotypic use of hands was part of the child, and should not be restricted in any way. ‘It’s as if she needs to use her hands’ (Interview 2) and ‘Her hand movements are part of her and she can’t stop them. It wouldn’t be right to try’
(Interview 5) were amongst the comments made. The use of splints may also have emotional overtones. They are usually visible and may appear to the uninformed as restraints, and as such may exacerbate the distress of parents. In general, the use of splints or restraints is rather controversial, and it seems logical that parents should be consulted when decisions are made regarding how stereotypical hand movements should be viewed. Parents can be very protective of their children, both with regard to physical discomfort and how they are perceived by friends and acquaintances, and they may resent professionals taking an expert stance in this instance.

6.6.3 Hand function - perceived value of therapies

Table 46 shows which therapies had been used beneficially to assist with the difficulties related to hand function. As hand function comes under the ‘umbrella’ of physiotherapy, it is surprising that there was only one reference to the value of physiotherapy. This referred to the physical aspect of the hand itself, rather than purposeful hand movements.

<table>
<thead>
<tr>
<th>No.</th>
<th>Music therapy</th>
<th>Speech therapy</th>
<th>Physiotherapy</th>
<th>Hippotherapy</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>encourages her to hold onto things</td>
<td></td>
<td></td>
<td>encourages her to hold onto things</td>
</tr>
<tr>
<td>2</td>
<td>encourages her to hold onto things</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td></td>
<td></td>
<td></td>
<td>encourages her to hold onto things</td>
</tr>
<tr>
<td>4</td>
<td>encourages purposeful hand use</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>encourages purposeful hand use</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>encourages purposeful hand use</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>she will hold on to things</td>
<td>helps with feeding</td>
<td></td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>encourages her to hold onto things</td>
<td></td>
<td>slows down hand deformation</td>
<td></td>
</tr>
</tbody>
</table>
Music therapy emerged clearly as the therapy most able to help with hand function, with motivation being seen as a strong incentive to hold onto objects. By incorporating stereotypic movements into acceptable activities, music therapy can ‘... offer ways to encourage and help children out of the small, isolated world of their stereotypies: to make possible new experiences, learning processes, social contacts: to provide the child with a medium for emotional expression and exchange to give her an opportunity to play’ (Schulz, 1987, p.16). The benefits of therapies interacting for the good of the child (Wigram and Weekes, 1985) has been discussed in Section 2.8, although this study does not look specifically at instances of therapies working together. This would seem to be an area where physiotherapists could impact upon the work being done by music therapists.

6.7 Learning ability

Learning ability is a key area of development, and in British education, it is the foundation upon which achievement is measured. Testing at the key stages reflects the pupil’s ability to learn, reason, and interpret meaning. For the majority of parents, their child’s learning ability is a fundamental issue, but for parents of children with an autistic spectrum disorder there may be a different perspective. There may well be a desire to use school time for giving of life skills or even relaxation, more so than for academic purposes, although Jordan, Jones and Murray (1998) remind us that ‘... children are the ultimate clients of the education provided by LEAs, and that provision must meet children’s special educational needs’ (p.128). These authors accept that on occasions this provision may not meet the wishes of parents. The eight parents in this study were asked to discuss in their own words their child’s current level of learning ability (Table 47).

This area caused considerable distress to two respondents, with one parent explaining ‘This is really difficult’ (Interview 6) and another becoming so distraught she was unable to continue talking. When a child is very young, a lack of academic ability can be easily disregarded or even go unnoticed, but a profound learning disability in an older child cannot easily be disguised. Again, this issue might be one of how school time should be used, and as such parents’ considerations should not be ignored.
Table 47: Parents' perceptions of current level of learning ability

<table>
<thead>
<tr>
<th>No.</th>
<th>Deteriorating, no progress</th>
<th>Knows more than is apparent</th>
<th>Will never achieve</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>★</td>
<td></td>
<td>★</td>
</tr>
<tr>
<td>2</td>
<td></td>
<td>★</td>
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<td>3</td>
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<td>4</td>
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<td>7</td>
<td></td>
<td></td>
<td>★</td>
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<tr>
<td>8</td>
<td></td>
<td></td>
<td>★</td>
</tr>
</tbody>
</table>

Five parents had an expectation that their child would never achieve academically. ‘We know she will never achieve. We expect nothing from her, just to be there for us’ (Interview 5) and ‘I know she will stay how she is for the rest of her life’ (Interview 7). However, parents of three children believed their daughters knew more than was apparent, with one comment: ‘It’s just that she can’t show what she knows without us helping her’ (Interview 4). This reflects the view held by Lindberg (1994) who believes that girls with Rett syndrome do have a capacity for learning, and that capacity, albeit limited, will be the greatest in areas where there is opportunity to obtain the most experience.

6.7.1 Learning ability - priority areas

Parents were asked to indicate their priorities for their daughter in the area of learning ability. These are summarised in Table 48 and the paucity of comments may be due in part to subconscious feelings of hopelessness, which some parents appeared to experience, as well as to an absence of ideas in a ‘conceptual’ area they find to be less tangible than the other three skill areas.

This was a difficult aspect of Rett syndrome for parents to discuss, with one parent commenting: ‘I don’t think about it. Then I’m just grateful for what comes along’ (Interview 1). Seven parents felt that happiness was of more concern to them than academic learning, with comments ‘It is important she has fun’ (Interview 3) and ‘Being healthy and happy is the only really important thing for Hannah’ (Interview 7). Personal experience has shown that many parents, once they have accepted that
their child has a profound learning disability, have little regard for academic achievements, valuing freedom from pain and happiness above all else.

Table 48 Learning ability priority areas

<table>
<thead>
<tr>
<th>No.</th>
<th>Make choices</th>
<th>Maintain current skills</th>
<th>Develop hand control</th>
<th>Improve communication</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>★</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2</td>
<td></td>
<td></td>
<td></td>
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</tr>
<tr>
<td>3</td>
<td></td>
<td>★</td>
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<td>4</td>
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<tr>
<td>5</td>
<td>★</td>
<td></td>
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<tr>
<td>6</td>
<td></td>
<td>★</td>
<td>★</td>
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<td>8</td>
<td></td>
<td></td>
<td></td>
<td>★</td>
</tr>
</tbody>
</table>

Girls with Rett syndrome, who have profound and multiple learning difficulties are, in the eyes of the National Curriculum, like all other children, entitled to a broad and balanced curriculum. This may be an evocative area, and significantly, three parents were unable to indicate any learning priority for their child.

The meeting for the annual review of the statement is an opportunity for parents to contribute towards their child's Individual Education Plan (DFE, 1994), and to explore suggestions for future planning from teaching staff. Structured approaches, such as TEACCH, have been successfully used with children with autistic spectrum disorders and those parents who have been involved in this approach may be more perceptive about their own child's learning ability.

6.7.2 Learning ability - perceived value of therapies

Conventionally, therapies relate to more esoteric areas than academic learning, which tends to be placed firmly in the hands of educationalists. Parents were asked for their views on the value of therapies in aiding learning.

Table 49 shows that only speech and music therapies were mentioned by parents in this section.
Learning ability was the aspect of Rett syndrome least influenced by the effect of therapies, with four parents believing them to have no educational value at all in this area. Speech therapy and music therapy were the only two therapies thought to be beneficial, with improvements in communication being most noticeable. One parent commented that everything her child does helps learning, not just one therapy, suggesting that interaction between therapies, and indeed education, may need to be considered.

There is a particular relevance to therapies in support of learning, but no clear opinions on how interactions develop. On one hand, therapies may be a vehicle for learning as well as a necessary medical treatment, on the other, they may be expensive to administer, of uncertain effect, and unconventional in education terms. It may be that there is a difference between what parents want and what others think is right. It is 'a fundamental principle' of the Code of Practice (DFE, 1994) that the 'knowledge, views and experience of parents are vital' (p.2). It may never be accepted that learning ability is not paramount, whatever the views of parents, but recent publications (DfEE, 1997, QCA, 1999) may help sway opinion away from targets of academic excellent towards more realistic life-skill objectives for children with profound and multiple learning difficulties.

It is possible that it is in the area of learning ability that real interaction between therapist, educator and parent should take place, to ensure that the familiar routines necessary for an understanding of the immediate environment are put into practice (McInnes and Treffry, 1993, Van Dijk, 1991). An interdisciplinary approach, where

<table>
<thead>
<tr>
<th>No.</th>
<th>Speech therapy</th>
<th>Music therapy</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>improves communication</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>improves communication</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5</td>
<td></td>
<td>helps with choosing</td>
</tr>
<tr>
<td>6</td>
<td></td>
<td>improves communication, controls hand movements</td>
</tr>
<tr>
<td>7</td>
<td></td>
<td></td>
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<tr>
<td>8</td>
<td></td>
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</tbody>
</table>
all adults involved with the child work as a team, accepting the expertise of parents and liaising closely with them, should provide the features of good practice advocated by Van Dijk (1991). Once this has been established, then therapeutic intervention can be integrated into this routine with a minimum of disruption.

6.8 **Summary of the value of therapies**

For each therapy parents were asked to summarise ways in which they thought this particular therapy was beneficial, or not, for their child and to indicate which therapy they would choose to retain if all but one had to be removed.

6.8.1 **Speech therapy**

Although six parents agreed that communication skills were helped by speech therapy, two added the proviso 'probably'. Only one parent thought the value was minimal, with one parent supportive of the therapy but openly critical of the provision. Parents were asked for their opinions of the therapy itself, but it is perhaps inevitable that their perception is coloured both by the standard of provision and by the skill of the therapist. None of the respondents chose speech therapy as the one to be retained in preference to the others.

6.8.2 **Music therapy**

All parents agreed that music therapy had many benefits. Enhanced hand function was thought to be a benefit by seven parents and improved communication was mentioned in six instances. Pleasure and relaxation were seen as important issues, although not all comments were totally positive, with two parents indicating that they would appreciate more involvement: 'This gives a lot of pleasure to Debbie. There is not enough feedback, so we don't know what she is doing' (Interview 4) and 'I would really like to see Elizabeth do her music therapy, but no one else is allowed into the room' (Interview 6). Jordan and Jones (1999) suggest that a video may be used if direct observation affects the pupil, and that 'Parents should be asked if they would like to see how their child works and plays at school and consulted on how this might be arranged' (p.46).

In one case only would music therapy be retained if all others had to be removed, with the comment: 'All the other therapies I could do myself at home' (Interview 5). It has been seen that, with guidance, it is possible for non-specialists to help with the delivery of intervention programmes (Harris, 1990, Levitt, 1984) and it would be
interesting to find out which therapeutic interventions are more conducive to a shared or facilitating role.

6.8.3 Physiotherapy

All parents thought physiotherapy helped their child with mobility, with the general health of the child also being addressed. Many girls with Rett syndrome suffer from constipation and in two cases, the exercise associated with physiotherapy was thought to help ameliorate this condition. Three parents valued physiotherapy above the other therapies. Apart from the physical benefits, and the related management issues, the role of the physiotherapist as a mediator was appreciated by two parents. ‘They are very good at passing on information to other people’ (Interview 6).

6.8.4 Hydrotherapy

Hydrotherapy was the favoured therapy, named by four parents as the therapy they would most wish to retain. Three parents thought that hydrotherapy helped in all areas of a child’s development, with pleasure and relaxation also featuring in its benefits. ‘She is also getting all the other therapies at the same time, plus she is having fun’ (Interview 3).

One parent felt hydrotherapy was the one therapy that could not be managed at home, whereas the others could be carried out at home if guidance was given. The essential one-to-one support needed in the water was one reason given for retaining hydrotherapy, a valid reason, although a consequential benefit. An experienced physiotherapist agreed with the views of the parents on the value of hydrotherapy.

When you are working with Rett syndrome girls, physiotherapy is essentially passive. The child often doesn't like the treatment and will resist. She becomes very hard to motivate. If in water, which is a medium they like, they are happy, then water induces relaxation and more can be done in the way of stretching and active movement. I would certainly say it is the most important therapy.

(Personal communication, 1999)

6.9 Summary

All parents wanted a good quality of life for their children, with happiness being considered more important than academic learning. The routes for achieving
happiness are many and varied for the majority of children, but less so for girls with Rett syndrome, and parents are obviously reluctant to cause any further suffering for their children. Even potential benefits were treated with misgivings if the means of achieving them could cause distress. For example, stereotypical hand movements were thought to be an essential part of the girls’ behaviours, and the possible benefits of restraining movements were not sufficient reason to cause discomfort. The use of restraints is an area of concern for parents, as instinct seems to be going against the views of professionals.

Communication, and predominantly the ability to choose, was considered to be very important. Music was thought to be a powerful motivator in encouraging choice, and thus indirectly communication, with pleasure being a consideration. All parents felt that they were able to understand their daughters’ basic needs and wishes, with the use of eyes underpinning all means of communication. Much communication is based on the parents’ ability to interpret signals, with people other than family and close friends less able to understand these signs.

Maintaining standards of ability already achieved was considered important, particularly physical achievements, and physiotherapy and hydrotherapy were considered most beneficial in this area. Many parents were knowledgeable about the condition of Rett syndrome, and were anxious to avoid the onset of scoliosis, which is more prevalent in non-ambulant children. Physical programmes were thought to not only help children to retain physical skills and maintain good health, but help prevent future deformities, with the future management of the girls also a cause for concern.

Whilst talking to the parents of children with Rett syndrome, it became clear that the stresses faced by most families are extreme. Although the issue was not explicitly addressed in the study, some parents seemed compelled to talk about the emotional difficulties involved, although this itself was not easy for them. Parents of all children who suffer from a disability are likely to experience a whole range of responses, including guilt, sadness and anger. This may be especially so for families of children with an autistic spectrum disorder. Whilst not experiencing the immediate stress of giving birth to an obviously disabled child, families may initially enjoy the pleasures of a ‘normal’ child and may subconsciously blame themselves for regression. ‘After diagnosis, there is shock and deep disbelief. This appears to
be more severe with parents of Rett syndrome children, as they have believed their child to be normal up to this point‘ (Goodship, 1987, p.539).

Alvarez and Reid (1999) described how living with a child with autism could lead to a state of permanent shock, hurt and grief. ‘... lives may become severely limited, with consequent stress on the parents' marriage and effects on the mental health of everyone’ (p.3). Hunter (1987) believes that most parents acknowledge the enormous stress a child with Rett syndrome brings to the marriage and to the family, and one parent in this study openly blamed the condition for problems in his own life. He described his daughter as hyperactive with an irregular sleeping pattern. ‘We spent all our time sleeping in shifts. My wife and I never spent a night in the same bed. Now we are divorced’ (Interview 7).

Apart from physical and emotional issues, thoughts of the future were very much in the minds of parents. Parents were realistic about their expectations for their child, and although some claimed to have come to terms with this, it was still an emotional subject for many. Most parents can look forward to watching their children mature, gain their independence and possibly have children of their own. Parents of girls with Rett syndrome know they will not have this satisfaction, with one parent saying: ‘It's really hard not knowing what to expect in the future, how long she will live’ (Interview 7).

The majority of parents accepted that their daughters would never regain that which was lost, and many held little hope for the future. There are suggestions that some therapies give pleasure, and assist a little in addressing the limiting effects of the condition. It may be that therapies can be used for the greatest benefit for the greatest numbers only within a school environment, and it is there that the balance between traditional teaching methods and less traditional treatments needs to be made.
Chapter 7 Conclusions

7.1 Areas for further research

The majority of published work concerned with Rett syndrome is of a medical nature, and this study has shown the need to bridge the gap between neurology and classroom. Although specifically centring on Rett syndrome, the conclusions have relevance for other autistic spectrum disorders. Therapies have been examined, and parents’ views surveyed, and from the results come suggestions for further research and implications for education.

7.1.1 Therapies and parents’ knowledge

This study looks at parents’ views concerning the values of therapies in relation to therapeutic management. One of the weaknesses inherent in surveying parental views is the variable basis against which those views are formed. While parents are not expected to gain expert knowledge in all the matters relating to Rett syndrome, some will be more informed than others. Parental assessments of therapy values are influenced by many factors that may be unequally weighted from person to person. Parental participation in intervention, knowledge of the syndrome, knowledge of the theory base for the different therapies, and perception of the actual effect will all make a contribution to the overall view, and further research is needed to determine how parents perceive intervention with respect to their own knowledge.

7.1.2 Interactions between therapies

It is possible that forms of interaction and cross-effect occur within the delivery of therapies. One therapy may be enhanced by another, or the length of time treatment is given may influence progress. It is not known by what criteria session lengths are determined, but the variation in the results suggests that the planning of each therapy should be examined in more detail, perhaps with regard to other disabilities, for it is entirely likely that group therapy sessions are not limited to a single syndrome or disability. In an alternative methodology, continuous data could be acquired and subjected to appropriate analysis to isolate those interactions.

7.1.3 Funding

It is suspected that available funding plays a more important role than admitted in determining therapy delivery. It would be useful to look at this issue across the
country to compare how different counties operate, and to determine the regional cost effectiveness of interventions.

7.1.4 National Curriculum

There seems to be a distinction between the aims of the National Curriculum and the wishes of parents of girls with Rett syndrome. Many parents expressed the view that enjoyment of life is paramount for their children, and the idea of relinquishing National Curriculum targets for the sake of enhancing the girls’ quality of life would be an interesting area for further research.

There is much recent literature examining the needs of children with disabilities (Ware, 1994, DfEE, 1997, Powell and Jordan, 1997, DfEE, 1998c, QCA, 1999,) and it would be interesting to look at how parents perceive the education of their children in the light of their own situations. School time takes a very significant portion of each weekday, and opinions regarding how that time should be used might vary greatly between parents of disabled and parents of able-bodied children.

7.1.5 Teachers’ perceptions

A parallel study to this, with the same questions, could be carried out using teachers’ perceptions about the value of different therapies. By using teachers’ knowledge of their pupils with Rett syndrome and having a working knowledge of the organisation of the school day, it may be possible to ascertain not only the benefits of therapies, but also the implications of withdrawal from the classroom environment.

7.1.6 Placement

The severity of Rett syndrome is such that all cases encountered personally have been in special school environments. It was assumed at the outset of this study that the girls would all be placed in special schools, and consequently no enquiry was made regarding placement. Nothing in any of the responses suggested otherwise, but in retrospect, the question should have been asked. In the light of the Green Paper (DfEE, 1997) it would be interesting to consider parents’ and teachers’ views regarding the most appropriate placement, and the problems associated with delivering therapies in a mainstream setting. It is possible that some girls with Rett syndrome are placed in mainstream schools, and if this is so, it
is important to know whether therapies were delivered, how they were delivered, and if their values were perceived differently by parents.

7.1.7 Needs of families

An Australian study to establish the needs of families with children with Rett syndrome found that not only were therapy services a frequently offered and highly valued source of assistance (Fyfe, Leonard and Callaghan, 1996), but that the diagnosis of Rett syndrome influenced the provision of therapy for the girls, with access to therapies increasing after diagnosis. There seems to be an absence of similar studies in the UK, and the effects of Rett syndrome upon the families of affected girls would be a significant area for research. Although this may not be a predominantly educational issue, the needs of families and children are inexorably linked.

7.1.8 Alternative therapies

There are several alternative, or ‘fringe’ therapies which are available in North America, and the International Rett Syndrome Association Internet mail server has carried reports of treatments as common as drug therapy and as unorthodox as dolphin therapy. While the latter is clearly not a practical option in the United Kingdom, drugs here tend to be administered as part of a medical regime, and not considered to be a therapy in the strict sense of the word. ‘Current therapeutic approaches to girls with Rett Syndrome are directed along the lines of providing habilitation with regard to physical and occupational therapy and communication skills and of providing suitable medication for the management of seizures’ (Percy and Hagberg, 1993).

There are, though, other less common therapies that may have a bearing on treatments for disability. One parent expressed an interest in the research of alternative therapies such as cranial osteopathy, reflexology homeopathy and herbal treatment. ‘I think that a collection of findings would be very useful for lots of children with special needs’ (Questionnaire 62). A broad based investigation into world-wide application of lesser known therapies would be interesting, but perhaps not of direct relevance to the British education system.
7.2 Educational implications of Rett syndrome

This study has been concerned with parents' perceptions of their Rett daughters' disability, and in particular with how affected skill areas can best be influenced by the use of therapies. It is to be expected that parents' views about specific symptoms will vary according to family circumstances, and also with age and medical condition of their child.

The onset of the disorder and rapidity of its progress varies with each case, as does the intensity of the symptoms. Just as it is difficult to quantify what is meant by 'severity', so the perceived importance of improvement is likely to vary. It was clear, however, that the importance of enhancing quality of life was acknowledged by some parents.

The DfEE (1997) highlights the need for suitable provision for all children with Special Educational Needs, ranging from those with profound and multiple learning disabilities to those whose difficulties may be addressed in a mainstream setting, with minimum disruption. Rett syndrome is a severe, low-incidence condition that is common enough for related national and international associations to flourish. Its effects are so profound that normal academic achievement targets are no longer appropriate, and many professionals and parents consider that exceptional treatment is needed to bring out the best in their daughters (Kerr, 1988, Lindberg, 1994, Lewis and Wilson, 1998). The role of therapies is vital in addressing the complex needs of girls with Rett syndrome, and needs to be considered in the light of recent Government policies.

7.2.1 Policies for Excellence

The policies of the Green Paper (DfEE, 1997) are intended to be applied to all children, and their aim is to improve the achievements of children with Special Educational Needs. The Green Paper states that it is expected that these policies for excellence should 'enable schools to reduce over time the proportion of children they identify as having SEN' (p.12). The causes of Rett syndrome are not fully understood, but are known to be due to complex neurological deficiencies within the brain. No policy or target setting will reduce the effects of this condition, neither will they reduce the statementing requirement for these girls, who are likely to be amongst those children who will always require significant assistance.
The Code of Practice (DFE, 1994) made it clear that both non-educational needs and non-educational provision must be specified on the statement of Special Educational Needs, although the responsibility for ensuring their implementation is less clear. Only clear directives supported by adequate funding will allow therapies to be administered appropriately, thus enabling a child to get maximum access to education, as well as enhancing quality of life.

On the one hand the Green Paper takes a holistic view and addresses the epidemiology of disability, but what is common to parents consulted in this study is the individual need for measures which to all intents and purpose seek to prevent further regression, and preserve whatever life quality remains. Academic objectives deal with the progress of learning, but they do not always address issues of health and happiness. It would be difficult to legislate for quality of life, because the varied aspects of life are valued so differently when illness or disability occurs, but it is an area which must not be ignored by those who are responsible for decisions which affect the lives of our most vulnerable children. This study began with an intent to identify educational benefits within therapeutic intervention, but has also demonstrated how the concept of life quality overlaps the traditional issues of education, infiltrates the rationale of special education and might easily dominate parents' wishes for their children.

7.2.1.1 Early identification

The 30 responses in this study showed a span of 2 - 10 years before diagnosis of Rett syndrome was made, showing clearly that early diagnosis cannot be assured. In all of the eight interviews, parents knew there was a problem with their child's development before diagnosis. This is not uncommon due to the nature of the syndrome (Kerr, 1995a), but what is unacceptable is the seeming lack of sensitivity of General Practitioners to the concerns of parents, with comments such as: 'I took her to my GP at about 10 months. He didn’t believe me at first' (Interview 2), 'At 11 months we started to get worried and took her to our GP. He said everything was OK' (Interview 5) and 'She reassured me by saying my daughter was just slow - “she won’t be a brain surgeon”. Tracey was then referred to a paediatrician, who again said she was just slow but didn’t diagnose her. She was then referred to an orthopaedic surgeon because she wasn’t walking. He reassured me' (Interview 8).
Hope for earlier and more rapid diagnosis lies with the medical profession, but it is education and other agencies that must mobilise treatments once there is a diagnosis. ‘… there is still no predictive test available to indicate the likelihood of having a Rett baby. This means that the development of practical resources and best remedial therapies and management techniques is of paramount importance’ (Freeman, undated, p.3).

There is clearly room for improvement in the relationships between medical staff and parents of girls with Rett syndrome, and other children within the autistic spectrum. Referring to parents of children with Special Educational Needs, Gascoigne (1995) agreed that ‘Some may have felt let down by medical personnel, perhaps through delays with referrals or mishandling of disclosure or diagnosis in the early stages’ (p.97). General Practitioners are under great pressure just from routine problems of the day, and probably do not view an ill-defined parental anxiety with the same urgency as a recognisable condition, illness, injury or disease. Yet the suffering Rett syndrome causes to families cannot be taken too lightly, and parental concerns must never be dismissed lightly. The education of doctors and other professionals may be an area that needs to be developed in order to reduce the likelihood of parents suffering in the way some of the study parents did.

7.2.1.2 Early intervention

The significance of prompt intervention in the early stages of difficulties is widely accepted, and is known to contribute towards acquiring the best educational and therapeutic opportunities (Kerr, 1988, Van Acker, 1991). The difficulty with Rett syndrome is that regression occurs in a manner which is unexpected and insidious; it occurs in infancy when common childhood maladies and subsequent recovery are expected, and what few early symptoms there are can be hard to detect and hard to define (Witt-Engerström, 1990, Kerr, 1995a). It is this characteristic factor of regression which makes Rett syndrome both difficult to recognise and difficult to deal with. Regression may also occur at dissimilar stages of development, making it impossible to specify a narrow time frame in which clues are concentrated.

7.2.2 Working with parents

The issue of parental involvement has already been discussed, and there is a widespread acceptance of the importance of consultation. The actual process of involvement may not always be easy (70% of parents questioned here did not
respond) but the data gathered may not be available elsewhere. In this study, the survey data represents a snapshot of some parent’s views at a particular time in their daughters’ lives. Respondents had opportunities to comment on their involvement with school, but less than 1 in 5 did so, and those that did expressed negative feelings. This is only one aspect of the survey, but with a condition as serious as Rett syndrome, it would be hoped that all parents would be satisfied with their level of interaction. There is no evidence here to imply that this result is typical of the national situation, but it does suggest that there are at least some dissatisfied parents, and that some work is needed to address the issue.

Research has shown that ‘Whilst parents are grateful for the identification of appropriate therapies for their child, they remain confused about how this will be put into practice’ (Northamptonshire County Council, 1998, p.4). Not all parents in this study were aware whether therapies were being delivered individually or in groups, and some were unaware for how long each month. It is clearly the responsibility of the school to inform parents of what is happening during the day, and personal experience has shown that it is not uncommon for pupil timetables to be sent home to parents. Lewis and Wilson (1998) point out that timetables alone do not represent a true picture of what is offered to pupils, as they often indicate the ‘how’ or ‘where’ of teaching rather than the actual curriculum content.

There remains a need to develop child support that overlaps and interacts with the wider needs of the family and community. ‘For the therapist involved with the child and family, that responsibility carries with it a commitment to develop practical solutions for day-to-day problems in caring for the child with Rett syndrome’ (Lieb-Lundell, 1988, p.S34).

7.2.3 Improving the SEN framework

A ‘robust framework’ for assessing and monitoring Special Educational Needs is essential if a child is to benefit fully from the most appropriate education (DfEE, 1997). Personal experience suggests that many practising teachers believe that the National Curriculum relates to rights more than to needs. For the child who cannot walk, talk or dress herself, and for her parents, there are far more important issues than the normal targets of the National Curriculum. Freedom from pain in later life, or indeed, a life expectancy extended by even a few years into adulthood, is more critical than academic achievements. There is a priority within special
needs which conforms to Maslow's hierarchical theory of needs (Maslow, 1968). Some children with profound and multiple learning difficulties have deficiency needs that outweigh growth needs, and even the first level of physiological needs may be difficult to satisfy. The role of the school is to fulfil these needs as far as is possible, and build upon them in successive layers, but the manifestations of Rett syndrome make it unlikely that cognitive and aesthetic growth or self-actualisation would ever be realistic objectives.

How the dilemma of attempting to satisfy deficiency needs within a growth based National Curriculum can be addressed is not clear, indeed, it may be true to say that such issues form a whole new consideration outside the National Curriculum. Equality of opportunity is a fundamental aim of our education system, but the reality is that girls with Rett syndrome will never achieve the independence and academic successes of their mainstream peers, or even that achieved by some other children within the autistic spectrum. The revised National Curriculum should ensure that the needs of all pupils are taken into account in any appropriate way by identifying the full range and type of non-statutory provision which is a recognised part of the school curriculum (Wade, 1999). It has been suggested that schools should receive guidance on the range and types of provision, outside the National Curriculum, such as therapy, which could be included in the school curriculum to meet the particular requirements of individuals or groups of pupils (QCA, 1999).

Emphasising equality and standardising levels of achievement only serve to isolate and mark as ‘different’ those children who will have special needs for life. Performance criteria have been developed to support schools by providing a common basis for measuring the progress of pupils for whom the early levels of the National Curriculum are not appropriate (DfEE, 1998c). They are, however, designed to support schools in setting targets to raise standards for all pupils, rather than assist in classroom planning, and meeting needs of individuals. There is a powerful argument for promoting ‘quality of life’ training to a position above regular curriculum activities, since with the current state of medical knowledge, girls with Rett syndrome will never take a competitive place in the real world. In essence, therapeutic intervention may be more beneficial than the National Curriculum in addressing some of the needs of children with Rett syndrome, and as such must be afforded the same, if not more, regard.
7.2.3.1 Statements

It is essential that the proposed reduction in the number of children with statements (DfEE, 1997) will have no repercussions for the child with a high risk, low incidence disability, such as Rett syndrome. Without the protective umbrella of a statement of Special Educational Needs, it is difficult to perceive how well expensive extra-curricular therapies could be delivered. If Local Education Authorities or Health Authorities were not compelled to provide therapies, then it may be that academic aims would take priority, regardless of individual needs. At the moment, the LEA must ensure that the needs of individual children are 'clearly and accurately described and that there is full agreement on the nature and quantity of the provision necessary to meet those needs, consulting the relevant responsible professionals as necessary' (DFE, 1994, 4.33).

The DfEE (1997) suggests that the distinction between educational and non-educational needs and educational and non-educational provision is often unclear, and recommend that the distinction be dropped. They accept that speech therapy, for example, can benefit a child's progress even if provided for health reasons, and propose to improve co-operation and co-ordination between Local Education Authorities and the Department of Health. It has to be hoped that both authorities share the aim of meeting children's Special Educational Needs more effectively, and it is suspected that the main barrier to effective collaboration is lack of clarity over funding. One suggested approach to resolving difficulties of funding is to give education authorities control over financial resources, and to enable therapy services to be purchased from health authorities. Another approach is for a system of joint responsibility for funding and managing resources to be established. Either solution, or some compromise between them, should keep the child's interest as the priority.

All parents in this study were aware of the inclusion or otherwise of therapies on their child's statement of Special Educational Needs. In this study, the most common therapies to appear on statements were speech therapy and physiotherapy, each one a 'traditional' therapy found in many special school environments. Some parents requested therapies that were not included on the statement, and there were examples of therapies being provided when not specified on the statement.
One parent in the study, when commenting on the value and availability of therapies, raises a simplistic view of the issue: ‘The criteria seems to be what is available, and not what is useful. The same is true for each statement’ (Questionnaire 49). It is not known how representative of the parents’ views this is, but it does highlight the issue of responsibility for ensuring the precision of statements.

Unless senior officers in a local authority are aware of each and every issue relevant to the syndrome, and indeed all conditions likely to be encountered in a school situation, and are aware also of the value of each therapy, there is every chance that decisions will be made for the wrong reasons. To deliver the best and right options, the right and best options have first to be identified and acknowledged. This also means that those who make the decisions need also to be well informed and knowledgeable of the processes by which the therapy benefits the child.

There appears to be little specific published evidence of the effect of therapies on girls with Rett syndrome, and this lack of evidence is itself instrumental in preventing full application of the complete range of additional facilities available in special education. 'Perhaps therapy as a concept, particularly when related to the creative arts, is still viewed with suspicion by some in the education field' (Warwick, 1995, p.209).

7.2.3.2 Individual Education Plans

It has been said that Individual Education Plans (IEPs) are generally most helpful when they are ‘crisply written, focusing on three or four short-term targets relating to key skills, such as communication skills, literacy, numeracy, behaviour and social skills’ (DfEE, 1998a, p.16). By acknowledging and legitimising behaviour and social skills, the DfEE seem to be endorsing aspects of the curriculum other than academic achievement, indicating that therapies may be seen as broadly ‘educational’. This perspective is already accepted in Australia, and the outcome of the partnership approach is encouraging. In Queensland, therapy services are delivered as part of a student’s educational programme and are directed towards educational outcomes. The purpose of the therapy is to ‘enhance and support the student’s educational goals, and to maximise the student’s access to and
participation in the curriculum’ (Low Incidence Unit, 1998). Personal experience in Queensland is of therapists fully integrated into the teaching team.

If a child receives a therapy, then the targets of therapists should be reviewed at the meeting for the annual review of the statement. If the statement has been accurately written, with full information about the child’s disability, then the targets of therapists should relate to the needs created by the disability itself. In the case of Rett syndrome, it is hoped that reference will be made to the essential criteria for diagnosis of the condition thus enabling appropriate therapy targets to be set.

By incorporating the targets of therapists into the IEPs, teachers could perhaps help to ensure that parents are aware of these targets, and reassure parents that the value of therapeutic intervention in the classroom is recognised. Such a process would add to the already considerable workload of teachers, but the benefits would be significant. It would increase the incidence of programmed therapy, and raise the profile of therapeutic intervention. It may also help to dispel parents’ concerns about the delivery of therapies.

7.2.4 Inclusion

Jordan and Jones (1999) discuss the benefits and disadvantages of mainstream placement of children with autistic spectrum disorders, but accept that not only will there be variation within the mainstream sector, but that the needs of pupils may change over time. The Green Paper (DfEE, 1997) makes it clear that, where possible, children should be educated in mainstream schools, with appropriate support, although it does recognise ‘the continuing need for special schools to provide - in some cases temporarily - for a very small proportion of pupils whose needs cannot be fully met within the mainstream sector’ (p.49). In the case of some low incidence disorders, including Rett syndrome, the disability is not temporary, and academic progress may be negligible.

This study did not seek to determine whether the children were being educated in special schools, special units or mainstream schools, nor did it elicit views from parents on the most appropriate placement, although the severe and complex nature of Rett syndrome makes it likely that most girls would attend special schools (Houghton, undated). It did show that communication between schools and parents is not perfect, and the question arises of how mainstream schools would need to
adapt in terms of parent relationships, if children with profound and multiple disabilities were to be included in a regular school environment.

Personal experience indicates that special schools are probably better at communicating with parents than mainstream schools, due to lower pupil numbers, higher staff / pupil ratios, and a greater interdependency between school and home. Some parents indicated that their child did attend a special school, and there was an expectation from one parent that the special school should offer provision which was over and above that of mainstream school. 'Claire attends a special needs school, and so speech and music therapy are part of her education' (Questionnaire 70). Even within special schools, there is a danger that therapies become routine, and are delivered as a matter of course.

What is clear is that the role of therapies alongside appropriate education is invaluable, and whatever the means of developing inclusion, the provision of therapies should not become a casualty of progress. In reality, examples can be found of real disadvantages that can be influential when a child with special needs attends a local mainstream school.

- Therapists may not be 'on site'
- Opportunities for group therapy may be limited
- Staff in mainstream school are unlikely to be used to the disruption to routine which can be caused by specialist support in the classroom
- Teachers from special schools are likely to have chosen that particular career route, and welcome the challenge of minority group children, whereas mainstream teachers may have had inclusion thrust upon them

If a girl with Rett syndrome is placed in a mainstream environment, it is essential that the support structure is in place for her. When a transfer is contemplated, the structure needs to be in place or evolving before the transfer takes place.

The DfEE (1997) advocates standardising the inclusion of children with similar needs across the country. The attitude of staff is likely to play an important part in the success or failure of inclusion schemes, and personal experience in the world of
education confirms that individual attitudes hold just as much influence over the relative success of integration as does legislation.

Inclusion should mean more than the education of children with Special Educational Needs in mainstream schools, alongside their able-bodied peers. Wade (1999) describes the Qualifications and Curriculum Authority's definition of inclusion as 'securing appropriate opportunities for learning, assessment and qualifications to enable the full and effective participation of all pupils in the process of learning' (p.81).

At a recent consultation conference on curriculum guidelines for pupils working below level 1 of the National Curriculum, the QCA spoke passionately about inclusion. They have a vision of a curriculum that is truly inclusive and embraces the needs of all learners (personal communication, 1999). The QCA recognised the importance of therapy in a child's timetable, both in terms of time spent and actual content. Unfortunately they were unable to provide a solution to the issue of funding, stating that this was a DfEE matter.

Whether in mainstream or special schools, 'children with the most severe and complex difficulties will continue to need specialist support' (DfEE, 1997, p.53). The Green Paper highlights the differences in quantity and quality of provision across the county, with wide variations in funding levels. It accepts that whilst variations in the ways in which educational services are provided are acceptable, quality must not be compromised. In a sense, the Green Paper is pre-empting the issue, as the best ways of dealing with Rett syndrome are not yet known. This research is in line with the DfEE requirements; it examines and notes the views of parents, and the conclusions, with those of similar studies, should be a foundation upon which to build. Further research is urgently needed at Government level to drive legislation in the right direction.

7.2.4.1 Value of therapies

Although this study has suggested that all therapies should be viewed in an educational context, it has become clear that parents value interventions that contribute to the health of their child, and at the same time give enjoyment.

Music therapy and hydrotherapy were perceived as being the therapies most beneficial in addressing the communication difficulties of girls with Rett syndrome.
Parents were particularly anxious that their child should be given the opportunity to choose, and music therapy was seen as a powerful motivator that encouraged expressive choice. There is much literature to support the therapeutic value of music therapy in communication skills, with particular reference to girls with Rett syndrome (Storr, 1993, Wigram, 1991 and Oldfield, 1995) and the value of hydrotherapy is also recognised by a leading researcher in the field of Rett syndrome (Kerr, 1995b).

Functional hand use was also helped predominantly by music therapy and again the research of others supports these findings (Schulz, 1987, Bruscia (Ed.), 1991, Heal and Wigram (Eds.), 1993, Hanks, 1986 and Wesecky, 1986). The interview data showed that 7 out of 8 parents were opposed to restricting their child’s stereotypical hand movements, and the same parents thought that music therapy could provide a route to purposeful hand movement.

Physiotherapy is specifically aimed at enhancing physical ability (Hanks, 1990), and was widely thought by parents to benefit motor skills, with a strong emphasis on preserving existing skills and ensuring the wellbeing of the child. The assistance offered by the physiotherapist was also seen as an area of support for parents.

Learning was an area which parents found difficult to address, both emotionally and rationally, with no one therapy seeming to be predominantly beneficial. A search of the literature confirmed that although there is scant evidence to show that learning ability can be directly enhanced by the use of therapies, they may indirectly benefit a child’s educational progress, even if provided for health reasons (DfEE, 1997).

As such, they need to be given the same regard as are other routes to education - a rationale accepted by one parent. ‘Although in a school inspection therapies are not considered under the education label, I think they should be seen as essential. The basis of learning is communication - speech therapy and music therapy, and this should be recognised by the LEA’ (Questionnaire 65). If, as appears to be the case, therapies are beneficial for certain features of Rett syndrome, then there is a strong case for their inclusion on a statement of Special Educational Needs.

There are clear concomitant effects between some therapies. For example, elements of physiotherapy can be difficult and sometimes uncomfortable for children, but may be a necessary part of the therapy programme, and can be made less uncomfortable if incorporated into a music therapy session (Bean, 1995).
Music therapy can offer variety in having to perform similar activities and a music routine can lighten the task, a view shared by Wigram and Weekes (1985) who believe that music therapy and physiotherapy can work together to provide a new dimension within which the child can work.

It is not fully known how therapies interact in the case of Rett syndrome, indeed, individual cases may respond in widely different ways. There is clearly much to be discovered, and while progress is steady in the medical field, this does not appear to be the case in education. No medical cure for Rett syndrome has yet been found, and until there is a greater understanding of the educational implications of the condition, it is unlikely that the complex needs of girls with Rett syndrome will be adequately met.

7.2.4.2 Financial issues

It has been argued that everyone concerned with the education of children with Special Educational Needs needed to work effectively together on a regional basis to 'ensure both the general availability of services, particularly for low-incidence disabilities, and the efficient provision of such services' (DfEE, 1998a, p.33). The cost of therapeutic provision should not be of concern to parents, but the survey revealed there was a degree of awareness of financial issues.

The Green Paper (DfEE, 1997) recognises the inconsistencies of quality and quantity of specialist provision across the country, as well as wide variations in funding levels, and states that 'We will work to secure a continuum of provision across the country so that, no matter where pupils live and whatever their needs, an appropriate level of support is available' (p.54). There is also the issue of minimum effect - and the influence of the length of therapy sessions is not known, although this research found no correlations between length of sessions and perceived benefit.

It is important that disputes over funding should not delay the development of processes by which therapies are introduced and sustained. It could be argued that while a child is taking part in a session of therapy, she is not being taught by a teacher, and so somehow it ought to be possible to offset the educational savings against the cost of therapy.
7.2.5 Developing skills

The DfEE (1997) acknowledges the need to raise the standards of all new and established members of the teaching profession, and accepts that much training and support will be required. Education is never complete; teachers constantly need to update their knowledge and skills if they are to work effectively in today's education system. There has never been, nor should there be, any suggestion that teachers should be expert in all areas. It is unlikely that even experienced practitioners in special education will have encountered every low incidence syndrome, nor will they have gained the understanding of the fundamental difficulties of individuals at the psychological level needed in order to develop programmes and appropriate teaching approaches (Jordan and Powell, 1995). Rett syndrome is a particularly complex disability, which until the mid 1980s had never been heard of in this country. Yet girls with Rett syndrome did exist before this, and presumably passed though the education system. How well this system treated them will probably never be known. The DfEE (1997) believes it to be the responsibility of 'all teachers and support staff in a school to be aware of the school's responsibilities for children with special needs' (p.61). Ideally, this should be addressed in the early stages of teacher training, when ideas are more easily assimilated. There may, of course, be a need for re-adjustment of course content. 'Inclusive education demands that initial teacher training (ITT) provides teachers with the necessary competence to identify and address pupils' SENs as set out in the Code. The current predominance of concern for NC content in ITT prevents an adequate allocation of training to achieve the necessary competencies' (Wedell, 1998).

Practising teachers also need time to rejuvenate and to increase and update their knowledge. This cannot happen spontaneously, and some choices will need to be made concerning how teachers' time is spent. Recently the Secretary of State for Education recognised the need for teachers to concentrate their efforts on improving the achievements of their pupils. 'Cutting unnecessary burdens on teachers helps us to raise standards in schools, and that is our top priority' (DfEE, 1998b). He suggests that school documents should be kept short, reports to parents should be crisp and concise and undue length and complexity should be avoided in lesson preparation. The time recovered by freeing teachers from burdensome paperwork might be used for improving classroom techniques and
extending knowledge. Whilst it is not realistic to expect teachers to be knowledgeable about every low incidence disability, non-contact time would offer some opportunity to research the disabilities encountered within a single school.

7.2.5.1 Role of teachers

The perceived role of teachers in delivering therapies in this study was not generally known, but the literature suggests that they have a valuable part to play in ensuring that therapies are carried out (Harris, 1990, Levitt, 1984). There was some confusion from parents about the different roles of the therapist and the teacher. This is not surprising as teachers and therapists themselves are often unclear about each other's roles and responsibilities. Schools have a responsibility to ensure that parents understand these complementary roles, and some considerable work may be needed before a proper understanding can be passed on.

The responsibility of teachers in delivering therapy is open to debate. There is a case for suggesting the class teacher take some responsibility in administering, even delivering therapies, but teachers' workloads are already high, and it is difficult to see how a teacher can become therapist without yielding some other responsibility. The responses in this study suggest that teachers and support staff do go beyond the duties generally expected of them, but how widespread this is, and how effective they are is not known.

For children with profound and multiple disabilities, school may not be a place for achieving key stages, but a place for learning how to live. The children do not necessarily need a teacher skilled in academic excellence, but they do need someone who understands and is able to address their real needs. ‘Teaching is more of an art than a technology (at least, within most educational approaches in the UK) and an inspired and dedicated teacher (whether this is a professional or a parent) can often succeed against the odds, with whatever approach he or she believes in’ (Jordan, Jones and Murray, 1998, p.119).

It could be argued that regular input from a competent and informed teacher might be better than a therapy programme that is poorly administered or suffers from regular disruption. It also raises the issue of the teacher's pivotal position in the web of learning, and of how much responsibility teachers should take in these
matters that are not strictly education centred. Teachers in special schools already
fulfil roles beyond those of mainstream teachers, and if therapies really are of great
benefit, perhaps there is a case for reviewing how they are delivered.

7.2.5.2 Role of learning support assistants

Routine tasks may be delegated by the class teacher to learning support assistants
(LSAs), and it is possible that they give some therapy. It is interesting to note that
the Green Paper, in acknowledging the role of LSAs refers specifically to supporting
speech therapy programmes, whilst other therapies are not specified. It could be
that 'helping pupils to access the curriculum' (DfEE, 1997, p.64) is an implicit
reference to other therapies, but in practice it is likely that either the school or the
class teacher will decide upon the role of support staff. It is suggested that 'Health
authorities may have an important role in training LSAs to support children with
substantial and complex difficulties, including medical needs' (DfEE, 1997, p.65).

It was not clear from this study, whether support staff were being used to support
therapies, although one parent suggested: ‘Physiotherapy is most important and a
daily routine should be followed by LSAs if there is no physiotherapist’
(Questionnaire 32). Personal experience has shown that the role of support staff
varies not just between authorities, or even schools, but between individual classes.
If the practice of working individually with children is widespread, then there are
implications, both for therapists, teachers, support staff and the children
themselves. The question then becomes one of responsibility and culpability. If a
child is injured or harmed by an LSA or a even a teacher attempting to deliver an
essential therapy when no trained therapist is present, there could be serious
repercussions.

7.2.6 Multi-professional approach

Few therapies can be considered to operate successfully in isolation, and the most
success is likely to be achieved within a multi-disciplinary approach. Professional
jealousies and insecurities may need to be overcome (Cotton, 1984) to ensure that
all practices can be best integrated for the benefit of the child. Lewis and Wilson
(1988) acknowledge the problems that may be encountered. 'Liaison between
teacher and therapist can be difficult, since each is working to different managers,
with different agendas and budgets. Time needs to be allocated to build a working
A professional in any field is likely to be supportive for their own role, and the enthusiasm of the therapist for the work undertaken would tend to enhance rather than dilute the effect of the work. It could also be argued that such enthusiasm would be passed on to the receivers of the therapy, and their carers. Rett (1985b) believed that the personalities of therapists involved with the child are vital to the success or otherwise of any treatment. Pragmatically, it matters not to a parent whether a child's improvements were fed by the therapy, by the environment, or by the therapist. The fact that all three combined to give a vehicle for development may be sufficient for them. However, it is not adequate to leave such an important issue to chance, and it is the responsibility of the educator to ensure that the diverse needs of these girls are met.

It is difficult for even the most dedicated professional to deliver an appropriate curriculum when adequate provision is not made. The DfEE (1997) emphasises the need for liaison between the Education and Health Authorities and indicates that the Government will 'consider funding joint research by the DfEE and the Department of Health into the factors which lead to the most effective provision of ... therapy for children' (p.72).

By inference, there must also be liaison between teacher and therapist. What appears to be needed is an environment where education and therapy are not treated in isolation. This concept has already been considered in the field of motor disability, where a highly trained conductor works on all aspects of a child's development, ensuring that there is transfer of learning from one activity to another. Hári and Ákos (1988) point out that it is not unusual for the various problems facing children with special needs to be 'shared out' by as many as ten or twelve professionals, including educationalists and therapists. They believe this to be the greatest obstacle to rehabilitation. If one person could take on this role, the benefits to the child would be great, although the responsibilities of the person involved would be even greater.

This approach would involve a whole new concept in training, but if successful could be the ultimate in a multi-professional approach.
7.3 **Summary**

This study has reviewed current medical and educational literature and discussed the benefits of therapeutic management with reference to Rett syndrome, and has touched on the broader conditions within the autistic spectrum. It has been seen that educational literature, in particular, is sparse and knowledge about the complex nature of the syndrome is limited. Yet Rett syndrome may even prove to be the commonest cause of profound mental and physical disability in girls and women (UKRSA, 1996a).

The position of Rett syndrome on the autistic spectrum, within the wider category of pervasive developmental disorders described by the World Health Organisation (1993) and the American Psychiatric Association (1994), is not always apparent from the literature, and frequently only brief acknowledgement to its position on the spectrum is made in Rett syndrome research. Similarly, literature dealing with autistic disorders often does not make reference to all specific conditions within this broader spectrum. Like all autistic spectrum disorders, Rett syndrome is characterised by abnormalities in reciprocal social interactions, in patterns of communication and by restricted, stereotyped, repetitive repertoire of interests and activities (WHO, 1992), but there are features of particular significance which sets the syndrome apart from other conditions.

Whilst there are good reasons for considering Rett in the wider context, those involved in education will see clear differences. Unlike childhood autism, for example, Rett syndrome carries with it:

- The certainty of severe mental disability;
- Repetitive and enervating hand movements unlike the stereotypies of autism;
- The appearance of poorly co-ordinated / unstable gait and / or trunk movements;
- The development of social interaction (including eye contact).

Although the profile of Rett syndrome has been raised in recent years, its place in the autistic spectrum, and differences as well as similarities are not always acknowledged, and need a wider appreciation.
The growing recognition of parents’ rights has been discussed in this study. Parents are now actively encouraged to express concerns about their child’s education, from discussion within individual schools right through to complaints to the Secretary of State (Northamptonshire County Council, 1997). A genuine desire for partnership requires there to be alternative pathways for dialogue, and this research opens up one such pathway. Rett syndrome was chosen as the unifying condition because of the growing recognition of the condition and its debilitating effects. The views of parents of girls who are in the early stages of their education were sought. These parents were invited to share their views on the value of therapies in the four essential areas of disability shared by all Rett sufferers. It was found that certain therapies were perceived to be more effective than others in addressing these areas of disability. The significance of the statement was highlighted, with the importance of non-educational needs recognised.

The study highlighted the shortcomings in the present system of identifying and educating children with low incidence disabilities. Many parents felt a certain dissatisfaction with their involvement in the educational process, particularly their views on which therapies should be specified on a statement of Special Educational Needs. Whilst it is accepted that it may not always be possible to agree to the demands of parents, their opinions are actually a valuable aid in educational planning.

In the academic sense, the ability of their child to communicate, with an emphasis on choice, was considered by parents to be the most important area, although health and happiness were considered paramount, with formal educational requirements of less concern.

The interviews (Chapter 6) offered the opportunity to speak personally with parents, and revealed that, whilst acknowledging the unconditional love they feel for their child, many parents still feel the hurt and desolation this syndrome has caused. ‘The fact that a child has been diagnosed as having a certain condition or syndrome many years earlier does not mean that the emotions experienced by parents have faded away by the time their child begins school’ (Gascoigne, 1995, p.11). The study also revealed the anguish of parents who had given birth to seemingly healthy daughters, but then seen a relentless deterioration to the point of severe
disability. Some parents felt a sense of despair that was compounded by the disbelief of the professionals to whom they turned for help.

There is, however, always some hope for the future. Medical research is continuing to scrutinise Rett syndrome (Kerr, 1997) and there have been significant recent developments although there is no guarantee that understanding the cause is synonymous with finding a cure. On an educational front, recent Government publications (DfEE, 1997, DfEE, 1998a, DfEE, 1998b, DfEE, 1998c, QCA, 1999) indicate that the special needs of minority groups are finally being taken seriously, and it is to be hoped that this study will generate support and encouragement for deeper research into the difficult issues facing the girls, their parents, and their teachers.
Appendix I The clinical stages of Rett syndrome

Table 50 shows the original staging system of Rett syndrome, including the points added in 1990 (Witt-Engerström, 1990).

Table 50 The four clinical stages of Rett syndrome

<table>
<thead>
<tr>
<th>Stage I: Early onset stagnation</th>
<th>Added points</th>
</tr>
</thead>
<tbody>
<tr>
<td>Onset age: 6 months to 1.5 years</td>
<td>Onset from 5 months of age</td>
</tr>
<tr>
<td>Developmental progress delayed</td>
<td>Early postural delay</td>
</tr>
<tr>
<td>Developmental pattern still not significantly abnormal</td>
<td>Dissociated development</td>
</tr>
<tr>
<td>Duration: weeks to months</td>
<td>'Bottom-shufflers'</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Stage II: rapid developmental regression</th>
<th>Added points</th>
</tr>
</thead>
<tbody>
<tr>
<td>Onset age: 1 – 4 years</td>
<td>Loss of acquired skills</td>
</tr>
<tr>
<td>Loss of acquired skills / communication</td>
<td>fine finger, babble / words, active playing</td>
</tr>
<tr>
<td>Mental deficiency appears</td>
<td>Occasionally 'in another world'</td>
</tr>
<tr>
<td>Duration: weeks to months, possibly 1 year</td>
<td>Eye contact preserved</td>
</tr>
<tr>
<td></td>
<td>Breathing problems yet modest</td>
</tr>
<tr>
<td></td>
<td>Seizures only in 15%</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Stage III: Pseudostationary period</th>
<th>Added points</th>
</tr>
</thead>
<tbody>
<tr>
<td>Onset: after passing stage II</td>
<td>'Wake up' period</td>
</tr>
<tr>
<td>Some communicative restitution</td>
<td>Prominent hand apraxia / dyspraxia</td>
</tr>
<tr>
<td>Apparently preserved ambulant ability</td>
<td>Stage III / IV</td>
</tr>
<tr>
<td>Inapparent, slow neuromotor regression</td>
<td>Non-ambulant patients</td>
</tr>
<tr>
<td>Duration: years to decades</td>
<td>Stage IV-A</td>
</tr>
<tr>
<td></td>
<td>previous walkers, now non-ambulant</td>
</tr>
<tr>
<td></td>
<td>Stage IV-B</td>
</tr>
<tr>
<td></td>
<td>never ambulant</td>
</tr>
</tbody>
</table>

### Appendix II Diagnostic criteria for classical Rett syndrome

Table 51, Table 52 and Table 53 show the diagnostic criteria for classical Rett syndrome.

**Table 51 Necessary criteria**

<table>
<thead>
<tr>
<th>Necessary criteria of Rett syndrome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Apparently normal pre- and perinatal period</td>
</tr>
<tr>
<td>Apparently normal development through at least first 5 - 6 months of age</td>
</tr>
<tr>
<td>Normal head circumference at birth</td>
</tr>
<tr>
<td>Deceleration of head growth (age 3 months to 3 years)</td>
</tr>
<tr>
<td>Loss of acquired skills (age 3 months to 3 years):</td>
</tr>
<tr>
<td>1. Learned purposeful hand skills</td>
</tr>
<tr>
<td>2. Acquired babble / learned words</td>
</tr>
<tr>
<td>3. Communicative abilities</td>
</tr>
<tr>
<td>Appearance of obvious mental deficiency</td>
</tr>
<tr>
<td>Appearance successively of intense hand stereotypies:</td>
</tr>
<tr>
<td>Hand wringing / squeezing</td>
</tr>
<tr>
<td>Hand washing / patting / rubbing</td>
</tr>
<tr>
<td>Hand mouthing / tongue pulling</td>
</tr>
<tr>
<td>Gait abnormalities among ambulant girls:</td>
</tr>
<tr>
<td>Gait apraxia / dyspraxia</td>
</tr>
<tr>
<td>Jerky truncal ataxia / body dyspraxia</td>
</tr>
<tr>
<td>Diagnosis tentative until 2 - 5 years of age</td>
</tr>
</tbody>
</table>
### Table 52 Supportive criteria

<table>
<thead>
<tr>
<th>Supportive criteria of Rett syndrome</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Breathing dysfunction:</strong></td>
</tr>
<tr>
<td>Periodic apnoea during wakefulness</td>
</tr>
<tr>
<td>Intermittent hyperventilation</td>
</tr>
<tr>
<td>Breath-holding spells</td>
</tr>
<tr>
<td>Forced expulsion of air or saliva</td>
</tr>
<tr>
<td><strong>Bloating / marked air swallowing</strong></td>
</tr>
<tr>
<td><strong>EEG abnormalities:</strong></td>
</tr>
<tr>
<td>Slow waking background and intermittent rhythmical slowing (3-5Hz)</td>
</tr>
<tr>
<td>Epileptiform discharges, with or without clinical seizures</td>
</tr>
<tr>
<td><strong>Epilepsy - various seizure forms</strong></td>
</tr>
<tr>
<td>Spastic signs, later muscle wasting and / or dystonic traits</td>
</tr>
<tr>
<td>Peripheral vasomotor disturbances</td>
</tr>
<tr>
<td>Scoliosis of neurogenic type</td>
</tr>
<tr>
<td>Hypotrophic small and cold feet</td>
</tr>
<tr>
<td>Growth retardation</td>
</tr>
</tbody>
</table>

### Table 53 Exclusion criteria

<table>
<thead>
<tr>
<th>Exclusion criteria of Rett syndrome</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Organomegaly or other signs of storage disease</strong></td>
</tr>
<tr>
<td>Retinopathy or optic atrophy</td>
</tr>
<tr>
<td>Microcephaly at birth</td>
</tr>
<tr>
<td>Existence of identifiable metabolic or other heredodegenerative disorder</td>
</tr>
<tr>
<td>Acquired neurological disorder resulting from severe infections or head trauma</td>
</tr>
<tr>
<td>Evidence of intrauterine growth retardation</td>
</tr>
<tr>
<td>Evidence of perinatally acquired brain damage</td>
</tr>
</tbody>
</table>

From Hagberg (1993, p.8)
Appendix IV Questionnaire and explanatory letter to parents

A Research Project into the Value of Therapies for Girls with Rett Syndrome

Dear Parent,

Thank you for taking part in this project. The information gathered as a result of your participation will be used to further the understanding of the effects of different therapies on children with Rett syndrome.

My first contact with Rett syndrome was in 1986, when I taught a girl at Fairfields Special School, Northampton, where I am now deputy headteacher. I have since taught other girls, both in this country and in Australia. My current research is part of a three year project at the University of Leicester and stems from the realisation that there are many inconsistencies in the way in which the needs of Rett syndrome girls are met in education.

This questionnaire attempts to identify which therapies are commonly used in schools, whether they are included on the statement of Special Educational Needs, and how valuable parents and carers feel them to be. In particular it aims to help determine which therapies parents believe are most beneficial for specific symptoms of Rett syndrome.

The questions are grouped into three sections. Section One relates to the present ability of your child compared to a child without Rett syndrome, and the importance of addressing main areas of disability. Section Two refers to the therapies themselves and Section Three to your own perception of the value of each therapy.

The research is endorsed by the University of Leicester and has the full support of the Rett Syndrome Association (UK). A summary of the findings will be made available to the Association when the research is complete.

The questionnaire may be completed anonymously, but if you are willing to take part in follow-up research, please write your name, address and telephone number in the space provided at the bottom of page 2. The distribution of the questionnaires has been organised by the RSAUK, and any information acquired by them in the process of distribution is confidential.

If you have any questions or comments about the study, please contact me at the address below.

Thank you for your support.

Patricia Moore
38 Homestead Way
Northampton
NN2 6JG

01604-719587
awm@compuserve.com
The value of therapies for girls with Rett Syndrome
University of Leicester in conjunction with the Rett Syndrome Association (UK)

Date of birth of child
Year of RS diagnosis

Part 1 - Abilities

<table>
<thead>
<tr>
<th>Area</th>
<th>Communication (eye pointing, sounds etc.)</th>
<th>Functional hand use</th>
<th>Motor skills (walking, crawling etc.)</th>
<th>Learning skills</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>□</td>
<td>□</td>
<td>□</td>
<td></td>
</tr>
</tbody>
</table>

For each of the 4 areas, say how seriously you think your child is affected. In each box enter a number between 1 and 5 where 1 is not affected and 5 is seriously affected.

How important do you feel it is for your child to become more proficient in these areas? In each box, enter a number between 1 and 5, where 1 is not important and 5 is very important.

Please use this space to add your own comments about your child's abilities.

Part 2 - Therapies

For how long has your child been receiving each therapy?
Are the sessions group (G) or individual (I)?
For each therapy how many hours does she receive each month?
Enter a tick if the therapy is specified on statement.

If your child does not receive a therapy, please mark the box with an X.

Music Therapy
Speech / Language Therapy
Hydrotherapy
Occupational Therapy
Physiotherapy
Aromatherapy

Please add any other therapies below

Please use this space to add your own comments about the use of therapies.
Part 3 - Effects of Therapies

<table>
<thead>
<tr>
<th>Therapy</th>
<th>Communication (eye pointing, sounds etc.)</th>
<th>Functional hand use</th>
<th>Motor skills (walking, crawling etc.)</th>
<th>Learning skills</th>
</tr>
</thead>
<tbody>
<tr>
<td>Music Therapy</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Speech Therapy</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hydrotherapy</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Occupational Therapy</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Physiotherapy</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Aromatherapy</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Please add any other therapies below

On a scale of 1 to 5, how effective do you think each therapy is for your child in each of the 4 areas shown. Enter a number from 1 to 5, where 1 is not effective at all and 5 is very effective, or X if the therapy is not received. Please use your own judgement of the effect of each therapy.

Please use this space to add your own comments about the effects of therapies on specific symptoms of Rett Syndrome.

Please use this space to add any general comments about the value of therapies for girls with Rett Syndrome.

Thank you for completing this questionnaire.
If you are willing to take part in further research into the value of therapies for girls with Rett Syndrome, please write your name, address, and telephone number in the space provided.

Please return this questionnaire in the envelope provided to Patricia Moore, 38 Homestead Way, Northampton NN2 6JG as soon as possible.
Appendix V Pilot Questionnaire.

The value of therapies for girls with Rett Syndrome
University of Leicester in conjunction with UKRSA

<table>
<thead>
<tr>
<th>Date of birth</th>
<th>Date of diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Part 1**

**Abilities**

<table>
<thead>
<tr>
<th>Communication</th>
<th>Functional hand use</th>
<th>Motor skills</th>
<th>Learning skills</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

For each of the 4 areas, say how seriously your child is affected. In each box, enter a number between 1 and 5, where 1 is not affected, and 5 is seriously affected.

How important do you feel it is for your child to become more proficient in these areas? In each box, enter a number between 1 and 5, where 1 is not important and 5 is very important.

Please use this space to add your own comments about your child's abilities.

**Part 2**

**Therapies**

<table>
<thead>
<tr>
<th>Music Therapy</th>
<th>Speech Therapy</th>
<th>Hydrotherapy</th>
<th>Occupational Therapy</th>
<th>Physiotherapy</th>
<th>Aromatherapy</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

For how many months has your child been receiving each therapy? Are the sessions group or individual? For each therapy, how many hours does she receive each month? Enter a tick if the therapy is specified on statement.

Please add any other therapies below

Please use this space to add your own comments about the use of therapies.

Pilot Study January 1997
Part 3
Effects of Therapies

On a scale of 1 to 5, how effective do you think each therapy is for your child in each of the 4 areas shown. Enter a number from 1 to 5, where 1 is not effective at all and 5 is very effective, or X if the therapy is not received. Please use your own judgement of the effect of each therapy.

Communication  Functional hand use  Motor skills  Learning skills

Music Therapy
Speech Therapy
Hydrotherapy
Occupational Therapy
Physiotherapy
Aromatherapy

Please add any other therapies below

Please use this space to add your own comments about the effects of therapies.

Please use this space to add any general comments about the value of therapies for girls with Rett Syndrome.

Thank you for completing this questionnaire. If you are willing to take part in further research into the value of therapies for girls with Rett Syndrome, please write your name, address, and telephone number in the space provided.

Please return this questionnaire to:
Patricia Moore, 38 Homestead Way, Northampton, NN2 8JG

Pilot Study January 1997
Appendix VI Structured interview

Name of child: Date of birth: Age at diagnosis:

What caused you to first think that your daughter was not developing in the way you would expect? How old was she at that time?

What do you consider to be your daughter’s greatest difficulties?

Gross motor skills

Tell me about your child’s current physical needs and abilities?

What do you think the priorities are for her in this area and how are they being addressed?

Have any therapies been used to assist with these difficulties? How have they been beneficial?

Communication

How does your daughter communicate?

Is she able to communicate to people other than family or close friends?

What do you think the priorities are for her in this area and how are they being addressed?

Have any therapies been used to assist with these difficulties? How have they been beneficial?

Hand function

Tell me about the way your daughter uses her hands?

Does she exhibit stereotypical behaviour?

Do you feel it is important to restrain or modify her hand movements? If so, how do you think this can best be achieved?

Have any therapies been used to assist with these difficulties? How have they been beneficial?
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