Author's Declaration

The work presented in this thesis was carried out in the Department of Sociology, University of Leicester, and is entirely my own work except where other authors have been referred to and acknowledged in the text. It has not previously been submitted for a degree in this, or any other University. The views expressed in this thesis are my own, and not those of the University.

Signed:

[Kerry Jane Allen]

Date:
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Kerry Allen

Managing the Body? The experience of Prader-Willi syndrome within families

Abstract
This thesis aims to explore the management of Prader-Willi syndrome (PWS) within families. PWS is a rare chromosome disorder, which has among its clinical sequela an intense interest in food, which may develop into an insatiable obsession, likened to addiction. The level of control over diet and the body required within PWS is very hard for people to achieve alone, since the majority of sufferers experience various forms of learning disability, autistic-spectrum disorders, developmental delay and behavioural problems. The thesis is particularly concerned with developing an understanding of the management of diet and food within the household and other everyday practices affecting the body of the child with PWS. The research is based on data from twenty qualitative case studies of English families which incorporate a child with PWS between the ages of eleven and fifteen years. The twenty families are purposively sampled to reflect difference in socio-economic status and family structure. Analysis of management strategies highlights the centrality of embodied agency in directing everyday practices and actions, this being particularly apparent in children with PWS as their experience of embodiment and emotion differs significantly from other family members. Focusing on the multi-dimensional nature of processes surrounding body management, the research identifies management practices and values of family members which guide these. The thesis also addresses the patterning of management strategies by social factors of family structure and socio-economic status. The work relates to contemporary sociological studies of the experience of chronic illness and disability in childhood, food, the body and the family, and contributes to current debates about embodiment, agency and health inequalities. Located in the intersections of the sociology of health and illness, disability studies and social theory, the study represents the first UK empirical sociological study of PWS.
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CHAPTER 1: WHAT IS PRADER-WILLI SYNDROME?

Introduction
The key aim of the research is to explore how Prader-Willi syndrome (PWS) is managed within the family context. However, the incorporation of a medically defined category within a piece of sociological research raises many questions. There is a strong tradition within medical sociology of questioning the objective reality of medical categorisations as biological realities, the formation of disease categories being seen as deriving as much from localised culture and societal modes of surveillance and regulation, as they are from biological bases (Peterson et al 1997). The complexity of the relationship between the epistemological disease category/medical knowledge and the ontological biological state has been widely discussed, as have the workings of power within it. Current debate on this topic is concerned to highlight and synthesise: relativism, in theories of the 'socially constructed' nature of medical categories and realism, so as not to deny the biologically 'embodied' nature of the human condition.

Laying down a preliminary theoretical approach to relativism/realism in relation to the derivation and experience of PWS is important to the thesis overall. As well as introducing literature and theory which can be built upon throughout the analysis, it targets many specific aspects of the everyday experience of PWS within the family. This chapter explores PWS as an epistemologically contested category, providing a sociological insight into what PWS is and how it is experienced. This allows us to build a picture of how PWS is understood in physical and moral terms by respondents in the research. It also allows us to mark the limitations of medical categories for defining individuals with PWS. In particular discussions of realism/relativism lead us to question: whether medical categorisations appreciate diversity amongst people with PWS; the role of biology in day-to-day experiences of PWS, and the extent to which formations of identity and daily interactions of individuals with PWS are affected by social processes and factors external to medical explanations.

Contemporary knowledge of PWS takes various forms. This chapter firstly examines
medical knowledge of PWS describing how it has developed over time, focusing specifically on the development of clinical features and explanations of the aetiology of the disorder. The subjectivity of this development will be examined in terms of how it can be linked to wider political processes of knowledge in medical culture and associated technologies. The chapter then explores how knowledge of PWS is connected with morality, drawing on medical metaphors used in PWS, legal cases, medical ethics and media representations of PWS. The relevance of this subjective and diverse knowledge of PWS to the experience of those with PWS and their carers is illuminated throughout.

1.1 The concept of PWS

Critiques of scientific and medical knowledge can be found in diverse forms across a range of disciplines, such as philosophy, politics, literary theory, sociology, anthropology, critical psychology and history. Further to this there has been much scope for interdisciplinary critique due to specific intellectual movements such as post-modernism, structuralism and post-structuralism, deconstruction and relativism (Woolgar 1988). Critiques of medical knowledge are highly visible in emotive polemics, such as Ivan Illich's (1978) 'Limits to Medicine', and in the work of those seen as figureheads for particular intellectual movements, for example in Michel Foucault's (1973) 'The Birth of the Clinic'. Moreover, the relations between medical knowledge, language, the social and the material/biological are also important and longstanding areas of interest in Feminist, Phenomenologist and Symbolic Interactionist literatures.

These various contributions to the critique of medical knowledge have been linked by the term 'social constructionism'. Vivien Burr (2003) uses the analogy of family as a model for social constructionism in relation to its contributors:

What links them all together is a kind of ‘family resemblance’. Members of the same family differ in the family characteristics that they share. There is no one characteristic borne by all members of a family, but there are enough recurrent features shared amongst different family members to identify the people as basically belonging to the same family group. (Burr 2003: 2)
Burr identifies the characteristics or core tenets of social constructionism as 'a critical stance towards taken-for-granted knowledge; historical and cultural specificity; knowledge is sustained by social processes and knowledge and social action go together', describing social constructionist approaches as being complicit in their 'anti-essentialism' and 'questioning of realism' (Burr 2003: 3-5).

The usefulness of Burr's framing of social constructionism as a distinct approach, albeit through a family analogy, is somewhat limited in relation to literatures concerning the focus of this thesis. On close inspection of the literatures and nature of arguments which fall into the remit of social constructionism, there is a strong tendency to acknowledge that the body has a real material basis, which is subject to material change across time and space. Whilst much feminist literature around the body and disability often focuses on inequalities based around the social construction of knowledge, it simultaneously highlights the existence of material biology through acknowledgement of embodiment, as Birke (1999) explains:

In emphasising social constructionism, in opposing it to biological determinism, we have perpetuated the dualism; and have played down the importance of the biological body itself. Like many women, I have trouble thinking about theories of social construction that ignore my bodily pain and bleeding, that ignore the ways that desire (however constructed) finds expression through my material body. My experiencing of both pain and desire is both materially and culturally constructed. (Birke 1999: 25)

Similarly, constructionist accounts which focus on the influence of professional, organisational and corporate powers in pathologising the body (Foucault 1973, Armstrong 1983) and the workings of social class in relation the patterning of health (Bartley 1998, Scambler 2002, Higgs et al in Carter et al 2004) are also keen to draw attention to the material existence of the body as a real entity, built upon by structural powers and processes.
Thus, although various strands of thought within the social sciences have highlighted the culturally constructed nature of medial knowledge and categories such as PWS, this attempt to highlight social factors in the construction of knowledge should be viewed as a reaction to 'biological determinism' and rather than a statement of existential denial of biological differences and biology's impact on individuals. However, in light of the focus of this thesis, important tensions between the epistemological bases of scientific/medical knowledge and social science knowledge remain. That is, concepts and ideas from general medicine, genetics and clinical psychology relating to PWS cannot be drawn upon unquestioningly throughout the thesis. These epistemological tensions are sometimes framed as questions of relativism verses realism.

Relativism is the view that human knowledge, beliefs and behaviour have no absolute reference. In its strongest form, relativists claim that humans understand and evaluate beliefs and behaviours only in relation to their historical and cultural context. Realism, in contrast holds that universal truths exist, independently and somehow prior to the social world. To claim that relativist thought underlies social science approaches, and realism acts as the equivalent in medical science, would be to overlook the complexity of research and ideas in both disciplines. However, much medical research into PWS, even that which attempts to link the disorder to social, behavioural outcomes such as educational achievement, reflects basic realist principles in its reliance on universalised abstract measures. For instance, within experimental research, individuals are grouped and generalised based on genetic subtypes within PWS. Further to this, these groups are often measured against pre-designed, discipline specific scales, such as Intelligence Quota (IQ) or psychometric scales. These abstract scales, categories and measures clash with social science understandings of behaviour, failing to acknowledge the complex processes which contribute towards human behaviour and social action. The linear correlations made between these medical/psychological concepts and scales can lead easily into biologically deterministic understandings of behaviour and action. As Birke (1999) noted, it is as an attempt to redress this biological determinism that much thought within social science has tended to concentrate on relativism based theories of knowledge.
construction.

The 'dualism' referred to by Birke (1999) places the biological firmly as the subject matter of medical and traditional sciences, removing it from the research agendas of the social sciences. One important step in resolving this dualism can be found in The Body and Society: Explorations in Social Theory, where Bryan Turner (1984) made a call for the human body, explicitly, to become central to the analysis of social processes, noting that differing interpretations of the body and its role in the social world are present in classical sociology, social anthropology and philosophy, reappearing significantly in medical sociology and feminist theory in the 1970s. From the mid 1980s onwards sociology has witnessed a significant rise in analyses of the body/society relationship often linked with consumer culture, feminist theory, embodiment, social action and the self in late modernity (Shilling 1993, 2005, Synott 1993, Brook 1999, Price et al 1999, Crossley 2001, Evans et al 2002, Howson 2004), with biology becoming the explicit focus of many texts (Birke 1999, Williams et al 2002). Williams et al (1998) highlight the significance of ‘emotions and embodiment’ as an emerging key theme on research agenda of the sociology of health and illness. They detail the reciprocal benefits between sociology of the body and sociology of health and illness, derived from the inclusion of ‘embodiment’ in health based research. Firstly the phenomena of illness, pain and disability provide interesting cases to exemplify embodiment and relations of body, society and self. Studies of disease and disability, therefore, allow greater insight into broad ontological debates of nature/culture, the role of biology and nature of emotions in social action. Further to this, the value of the interjection of ‘the body’ in studies of health and illness can be seen as,

Enabling sociologists of health and illness to re-read existing themes and traditional concerns in a new corporeal light, from the problems of the medicalisation of everyday life to the dilemmas of chronic illness and disability. (Williams et al 1998, online: http://www.socresonline.org.uk/socresonline/3/4/1.html)
Adding to the literature in the area of embodiment, chronic illness and disability, highlighted by Williams et al., the study of PWS within the family provides terrain upon which to explore biological-social interplay in a primarily grounded and empirical fashion. As much of the major work on embodiment tends to be highly theoretical in nature (Turner 1984, Shilling 1993, Burkitt 1999, Crossley 2001), this provides the opportunity to add to applied research literatures drawing on this theory.

Ascribing agency to biology is a complex and politically sensitive task. As Carolan (2005) notes, 'Sociology has been cautious of looking too far into the realm of the biophysical for causal potentials out of fear that such analyses might mark the beginning of a slippery slope toward biological reductionism' (Calolan, 2005: 1). The project of exploring the agency of the biological within social and political science has therefore not concentrated on the conceptualisation of any straight forward causal relationships of biology affecting society. Rather, through embodiment theory and critical realism the object of reintroducing biology is more subtle; to acknowledge it as one of the conditions from which social action results and as one of the mediums through which social action is conducted. The relationship between the biological and social is viewed as one of codependency or synergy. This makes the isolation of biology, as a site for the ascription of agency impossible, but at the same time highlights the inappropriateness of ignoring biology in the study of phenomena.

Attempts to negotiate synergy between relativist and realist conceptions of the body and its biology provide a useful starting point in formulating an approach to the existence and nature of PWS. William’s (1999) critique of social model disability theory (Oliver 1990), uses ‘critical realism’ to address relativist/realist dualism and explore the role of biology. He draws attention to two dualism related problems in the social model of disability’s focus on ‘disability as social oppression’, as have Hughes and Paterson (1997). Firstly, that it is ‘uni-directional’ (towards a relativist position) in its approach to the biology/society equation, not accounting for the part biology can play in affecting the individual, specifically through the biological nature of emotion. Secondly, viewing disability as a form of social oppression based on ‘impairment’, accepts unquestioningly
impairment as defined by biomedicine. This adds to the problem of dualistic segregation of disciplinary understandings and subject matter, discussed earlier in relation to Birke (1999). As Williams notes, ‘endorsing this view of the impaired body is complicit, wittingly or otherwise, in a medicalised approach to disability’ (Williams 1999: 803).

Avoidance of this ‘medicalised approach to disability’ can be sought through a thorough examination of the concept of PWS. William’s observation of the significance of the direct role of biology on the individual’s emotions and sense of identity is particularly relevant to experiences of PWS. Autistic spectrum disorders and the dysfunction of regulating processes i.e. appetite, temperature, both commonly described within PWS, have directly affect on the individual’s comprehension and everyday embodied emotional experience. To deny the role of biology as a disabling factor in its own right would certainly mean understating the severity of everyday experiences of disability for those with PWS.

Towards harmonising biomedical and social science approaches, Williams advocates critical realism as a way forwards. Although, Roy Bhaskar did not initially assign the term ‘critical realism’ to his work, it is his contribution to this philosophy that has the most useful application in this context, as he considered specifically the nature of philosophies of science and social science (Bhaskar 1978, 1989, 1994, 1997, 1998). In Reclaiming Reality: A Critical Introduction to Contemporary Philosophy (1989), Bhaskar sets out a theoretical framework which works equally for the social and natural world and as such can be used as the foundation for research in science and social science. He identifies three dimensions: ‘actual’ which are events; the ‘empirical’ where events are experienced by humans; and the ‘real’, the often unrealised steering mechanisms behind events. The ‘real’ are mechanisms which exist separately from human knowledge and that remain, relative to the other dimensions, constant through time and space.

Through critical realism, the workings of biology can be viewed as ‘real’; as mind-independent mechanisms, affecting events and experience, upon which biomedical knowledge has been built, yet in its entirety exists, to the most part, undetected. Williams
(1999) observes the capacity of critical realism in 'bridging the divide', giving natural and social sciences a common philosophical base. In doing so it avoids the conflation of epistemology and ontology that Williams believes is inherent to social model disability theory circa 1990, where the biological body is reduced to what is known about it. This type of theorization is described by Bhaskar as an 'epistemic fallacy'. In his words, the 'epistemic fallacy' is the conviction that '...statements about being can always be analysed in terms of statements about our knowledge' (Bhaskar 1989: 13).

Scambler and Scambler (2003) usefully build on Williams' ideas, demonstrating the benefits of critical realism for understanding medical conditions, specifically the rare genetic disorder, 'Juvenile Batten Disease'. They outline the need for a viewpoint which acknowledges both the impact of the biologically impaired body on the individual and the social implications of this for the individual and their family. To this end, they utilize critical realism's concept of 'ontological differentiation'; that the objects of biomedical and social science are both real and different, as are the generative mechanism governing these objects. They note:

Such a position (critical realism) permits acknowledgement of both the unyielding physicality of those multiple disabilities which are its issue and which invade all aspects of the lives of affected children and their families...research into gene therapy and stem cell transplants are suggestive of generative mechanisms which have little in common with generative mechanisms like class, gender and ethnicity which feature in sociological research. (Scambler and Scambler in Williams et al. 2003: 61-62)

In a similar way, critical realism provides a useful base for conceptualising PWS. The biologically distinct properties of people with PWS such as altered body composition involving poor fat: muscle ratio, learning and speech difficulties and excessive appetite, have real and different generative mechanisms from these individuals' socio-economic

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1 Degenerative childhood disease, death occurring ten to fifteen years after onset. Symptoms include: visual impairment, seizures, jerky speech, loss of speech and intellectual deterioration.
characteristics.

It is important to note that the social model disability theory critiqued by Williams (1999) has developed considerably over recent years. Notably, it has expanded the view of disability to incorporate an appreciation of the embodied nature of human life and the simultaneous impact of biological and social factors on identity. Moving away from a conceptualisation of disability as solely social oppression, in 2000 Shakespeare et al, called for a four dimensional approach to disability involving physical experience, social dimensions, psychological processes and cultural patterns and representations. He notes, 'these four dimensions of analysis, inextricably entwined, produce the disability phenomenon which millions of people experience everyday' (Shakespeare and Erickson in Rose et al 2000: 195).

This opening up of the concept of disability to incorporate embodiment is also visible in disability theorists' use of the phenomenological ideas of Maurice Merleau-Ponty. Phenomenological philosophy is commonly seen as providing the foundations for contemporary conceptualisations of embodiment, in particular Merleau-Ponty’s The Phenomenology of Perception (1962). In The Phenomenology of Perception (1962) his account of the lived and existential body explicitly rearticulates the relationship between the subject and object and tackles a series of related binaries, all connected with Cartesian dualism. He purports the idea that all scientific knowledge and theory is derived from practical human experiences of bodily exposure to the world. Iwakuma (1999: 78) observes Merleau-Ponty’s exploration of the relationship between the subject and object and displays the applicability of his notion of ‘embodiment’ to disabled people’s use of tools and technologies as extensions of the body.

By placing the body as the subject of perception, Merleau-Ponty initiates a framework where individuals’ social worlds can be shaped by their bodies and bodily differences. What is unique about the way Merleau-Ponty views this process is that the differences in the way that people perceive society is not solely the result of society’s response to the
individual’s body and the discourses and representations commonly held in that society. Rather, Merleau-Ponty gives embodied beings agency, arguing that their own sensory make-up may perceive the world in its own way. That is, that their biology may directly affect how they perceive the world. Considering the stubbornness, rigidity, lack of social awareness and obsessive traits described in the personalities of those with PWS, their perception of the world is arguably as altered as their biology, making Merleau-Ponty’s phenomenology of perception a useful way of theorising social action and perception.

Returning to theories that create synergy between realist and relativist conceptions in order to outline my own approach to PWS, Ian Burkitt’s (1999) work on embodiment and identity in modernity Bodies of Thought, draws heavily on the theory of Merleau-Ponty, whose applicability to this thesis has been noted. In addition, his work on the body in social relations unifies three traditionally distinct ways to theorise the body; as a thinking entity, a symbol and a physiological being. Burkitt’s theorisation of social action is involved in a constant project of breaking down Cartesian dualisms, as such creating a synergy between realist and relativist approaches. Burkitt notes that social constuctionism is useful in historical analyses of knowledge, but not an adequate framework for examining relations between humans, how humans interact with and use materials and how they transform the ‘real’. He instead advocates a multi-dimensional approach, arguing that knowledge is located primarily in the experiences of the real and active body. Burkitt avoids placing social construction and materiality in opposition by arguing that social constructionism is a material, as well as a linguistic process and that reality is a constantly changing state created by people, both materially and discursively. The concept of PWS, through Burkitt’s analysis, can thus be a biological reality, whilst the culturally subjective nature of its construction is also appreciated.

My theoretical approach to PWS draws on ideas from critical realism and Burkitt’s (1999) theory of embodiment and social structure/action. In order not to understate the severity of everyday experience for people with PWS, I take an embodied approach, acknowledging the real biological differences which exist in the bodies of those with PWS, directly affecting their emotion, identity and action. However, I do not conflate this
biological reality to the biomedical classification of PWS. Instead, in line with Williams, I view this label as 'merely descriptive, not constitutive of disease itself' (Williams 1999: 806). Following critical realism I recognise the equal importance and validity of scientific and social scientific research and understandings of PWS. However at this point I move away from a critical realist theorisation. Rather than advocating the separation of medical and social investigations of disability as outlined by Scambler e.g. ‘the most effective way of enhancing knowledge of processes of disease/disability may well be to forsake (premature) inter-disciplinary integration’ (Scambler 2003: 61), I take from Burkitt a view that humans are physical, thinking, symbolic and productive, continually involved processes of transforming the real e.g. altering biology through pharmacological tools and lifestyle. Thus the real biological bodies of those with PWS are as much the result of social processes as they are of nature. More specifically, contemporary bodies are the result of social processes which incorporate and build upon nature ‘social relations are lodged in material contexts’ (Burkitt 1999: 73).

As well as acknowledging the real existence of biology and the natural and social factors affecting it, my approach to PWS simultaneously appreciates the culturally constructed nature of medical knowledge and understandings of PWS. With this approach in mind the concept of PWS will be assessed in terms of its biological reality and its socially constructed nature, posing the questions:

1. Is PWS a distinct biological reality and biological sub-group of individuals? and
2. Are variations within the disorder accounted for in medical understandings?

That is, to what extent do individuals with PWS display and share the specific and real biological characteristics which make up its clinical diagnostic features? Did people displaying these characteristics exist before PWS was named, and if so, what factors can be seen to have led to its medical interception? As we have seen, social constructionist approaches to medicine do not generally seek to deny the existence of a biological or material reality. Rather they are concerned with how and why cultures highlight and organise knowledge using these phenomena (Blaxter 2004). Thus, even where biological characteristics of PWS may be visible in individuals over time, recognition of the cultural influences which led to their specific form of medicalisation remains a vital component to
the redefinition of the disorder, leading to the further questions:

3. How are developments in medical knowledge of PWS linked to broader medical trends and technologies? and

4. What is the current dominating medical framework through which PWS is understood and explained and what impact is this having on the setting of future research agendas?

That is, how is the concept of PWS culturally specific and how might medical culture, paradigms and their associated technologies sustain or shape future knowledge within the concept?

These four questions first point to a need to trace the existence of the biological and behavioural characteristics associated with PWS in history. And secondly to the need for a close examination of how and why these biological characteristics were intercepted by the medical profession and have developed from this point. The formation of medical knowledge on PWS can be broken down to focus specifically on changes in clinical features over time, then on aetiology, in line with rapidly changing genetic trends and technologies. The current (2006) diagnostic criteria can be found in appendix 1.

1.2 The biological existence of PWS

Following a critical realist conception, the biological reality of what is now classified as PWS has existed throughout time and space, independently of medical knowledge about it. The earliest evidence explicitly illustrating the characteristics in an individual now commonly associated with PWS can be traced to 184 years before its medical interception.
In the late 17th Century, King Charles II commissioned the painting of these two portraits. The subject, Eugenia Martinez Vallejo, was a six-year-old, weighing 120lbs. She displays excessive central obesity, small hands and feet and the characteristic facial features.

Less explicit evidence of the disorder can be traced to the Palaeolithic era, in the form Taurodont teeth from a Maltese skeletal archaeological record. These are associated with PWS but are also a common feature in genetic abnormalities such as the presence of an
extra X or chromosome 21. To add to this, Taurodontism\(^2\) is thought to be reasonably common in homo neanderthalis, so this cannot be taken as definitive evidence of congenital abnormality (Savona-Ventura 1996, online: http://geocites.com).

Holland et al (2004) along with many other PWS specialists argue that because of neonatal hypotonia\(^3\) and feeding problems it is unlikely that many infants would have survived prior to the advent of 20\(^{th}\) Century neonatal care. Thus, the second, over-eating or hyperphagic phase, which begins after weaning, would have been rarely observed and then only in isolated individuals. Specifically, developments in feeding techniques such as the introduction of tube feeding technologies in the early twentieth century, made it possible for significant numbers of babies to survive and hence, groups of people with apparently similar characteristics to be noticed and eventually reported in scientific literature.

In summary, it is apparent that the biological reality now categorised as PWS has been present throughout history. Medical specialists have drawn attention to the difficult neonatal phase in the disorder and the likelihood of infant deaths as a reason why nothing was recognisably recorded on the condition until 1864. This chapter will now move on to outline the construction of the disorder, highlighting additional reasons for the timing of developments in medical knowledge on PWS.

\(^2\) A malformed multi-rooted tooth characterized by an altered crown-to-root ratio, the crown being of normal length, the roots being abnormally short, and the pulp chamber being abnormally large. (2001, Marquette School of Dentistry, online: http://www.dental.mu.edu/)

\(^3\) A state of reduced tension in muscle. (Martin 1998: 322)
1.3 The Development of Medical Knowledge

The first known scientific recording of the syndrome came in 1864. John L.H Down, who first described Down’s syndrome, also described a single case, which was named ‘polysarcia’. The patient was 4’4” tall and weighed 210lbs and had small hands and feet. She was described as having little body hair, being 'mentally retarded' and not menstruating. Her history also reflected the common medical criteria for PWS; she had been thin and delicate up to the age of seven, but then had become hyperphagic (Down, 1864).

In 1956 the Swiss paediatricians Prader, Labhart and Willi published a brief article describing a new syndrome. And this is the point where it was named PWS. The main features noted at this time were neonatal hypotonia, impaired sexual development, short stature, a propensity towards severe obesity and mental retardation. A paper was given at the 8th International Paediatric Congress in Copenhagen; this emphasised the most distinctive feature to be hyperphagia leading to excessive obesity. In 1963 Prader and Willi followed this up with a study of 14 cases in addition to the original features they

In 1968, Gabilan and Royer analysed 11 cases and were the first to describe strabismus⁴ and orthopaedic problems such as scoliosis⁵, lordosis, coxa valga⁶ and hip dislocation. As more people were identified with the syndrome, advances were made in delineating common characteristics and in distinguishing PWS from other conditions with some similar features. Again in 1968 Zellweger and Schneider described 14 people with the syndrome and reviewed literature on 79 others. It was at this point that the distinction was made between the early hypotonic phase and the later obese hyperphagic phase. They also reported on the non-consistency of diabetes mellitus as a connected characteristic.

Hall and Smith added to the clinical features in 1972. Tabulating the abnormalities in 32 cases, they found delayed developmental milestones, male hypogenitalism⁷ in 100% of the cases, and personality problems, delayed bone age and male cryptorchidism⁸ in over half of the cases. They also drew attention to the temper tantrums and stubbornness that occur from childhood. After this it was not until 1993 that definitive Diagnostic Criteria for PWS were drawn up.

Whittington et al (2004) have suggested that early twentieth century developments in neonatal feeding technologies allowed significant numbers of children with PWS to

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⁴ Squint (heterotropia): any abnormal alignment of the two eyes. The strabismus is most commonly horizontal, but it may be vertical. Or in rare cases both eyes look towards the same point but one is twisted clockwise or anti-clockwise in relation to the other (Martin 1998: 627)

⁵ Laternal deviation of the backbone caused by congenital or acquired abnormalities of the vertebrae, muscles and nerves. (Martin 1998: 589)

⁶ Coxa - Hip bone or joint (Martin 1998: 153) Valgus – describing any deformity which places the hand or foot away from the midline (Martin 1998: 691)

⁷ Hyp/hypo – denoting deficiency, lack or small size.

⁸ The condition in which the testes fail to descend into the scrotum and are retained within the abdomen or inguinal canal.
survive through infancy, this rise in the PWS population being one factor leading to its 1956 definition. Utilizing the social constructionist arguments of Foucault (1973) and Armstrong (1983) relating to developments in medical knowledge, we can outline a second reason for the timing of medical recordings of PWS. In *The Birth of the Clinic* (1973) Foucault introduced the concept of the ‘anatomical atlas’ as both the tool and the ongoing project of medical knowledge in the discovery and explanation of disease. Measurement and interrogation of the biological body, Foucault argues, became central to medicine as disease was understood through mapping characteristics of the physical body. Foucault observes that it was not until the late Eighteenth century that the biological body became central to interpretations of health and illness in this way. Foucault also observes this biological emphasis on mapping of the human body, through medical science as a limited process, in particular subject to the influence of social, economic and political structures.

The widespread surveys and population surveillance, common to the post World War II era in European medicine, form a second reason for the 1956 definition of PWS. Down's 1864 recording of 'polysarcia' was not purposely built upon by Prader, Labhart and Willi. It can be seen as an exercise typical of it's time, where emphasis was on classification and collation of physical bodies. Developments in knowledge of the characteristics of PWS were visibly most rapid in the 1950s and 60s. Although Prader, Labhart and Willi were Swiss, many of David Armstrong's (1983) insights into 20th Century medical knowledge in England have relevance to developments in PWS, particularly in consideration of Lawrence, an English paediatrician who conducted a study of 6 children in the late 1950s and who was said to be marginally beaten in the naming of the disorder by his Swiss counterparts.

As Armstrong (1983) suggests, the two decades following WWII saw an increased interest in the patient within the community setting, resulting in large scale surveying to ascertain a 'norm' and identify outliers. The definitive discovery of PWS being made by paediatricians also has some resonance with the work of Armstrong, as he highlights that
the post war years witnessed a specific interest in the discovery of illnesses particular to children and again to population surveys of children's characteristics.

Within the development of knowledge of clinical features of PWS, what is seen as the main feature was subject to change and remained unresolved. Where Prader and Willi highlighted increased appetite leading to obesity, Lawrence highlighted the main feature as neuromuscular. Holm's (1993) and Gunay-Aygun's (2001) diagnostic criteria, however, highlight the multi-system nature of the disorder and the many various combinations of characteristics that can occur within it. This perhaps highlights another cultural shift in medicine, towards acceptance of the decentralised and fragmented nature of disorders. Medical fields such as genetics have arguably moved away from straight forward 'specific cause and effect' models towards an appreciation of complex variations within and between individuals' biology and how it interacts with its environment (Rose et al 2000, Davey in Williams et al 2004).

From Down's initial recording of polysarcia in 1864 through to the late 1960s, no
an explanation for the possible cause of PWS was put forward. Zellweger and Schneider's 1968 analysis of 14 cases, which had distinguished the two phases within PWS, also speculated that the disorder had a hypothalamic cause. The hypothalamus is a region of the brain which secretes hormones, controls the release of hormones in the pituitary gland, and controls water balance, sleep, temperature, appetite, sugar balance, fat metabolism and blood pressure. Hypothalamic abnormalities are still thought to be central to the causation of at least some of the disorder's clinical features. However, the exact processes at work in the hypothalamus and techniques to correct them remain unknown.

Rapid developments in aetiology started in 1981. Genetics have since become and remain very much the central explanatory factor. Ledbetter et al (1981) discovered that most individuals with the syndrome have a small deletion at a certain area (q11-13) on the long arm of chromosome 15. In 1983 Butler et al reported that the chromosome 15 deletion was on the paternal chromosome. Then in 1989 Nicholls isolated the cause of the disorder in the majority of non-deletion cases. Instead of a deletion from the paternal chromosome, they had two chromosome 15s from the mother and none from the father—a situation called Uni-parental Disomy (UPD). The chromosomes appeared to be a normal size and structure under a microscope; only through imprinting or investigating the inheritance patterns of the chromosomes was this addition to aetiology made possible.

In 1993 Nicholls produced a further explanation for the imprinting abnormality he had earlier outlined, i.e. UPD. He asserted that genes are expressed differently depending on whether they are maternally or paternally inherited. Thus having two maternal chromosome 15s will have a similar effect as a paternal deletion, as the paternal information remains missing. Nicholls findings are seen as revolutionary in the field of molecular genetics, as PWS is the first human disorder to show effects of genomic imprinting.
Buiting et al in 1995 as Saitoh et al in 1997 located the cause of 1-5% of individuals with the disorder resulting from neither deletion nor disomy. They found that these remaining cases were caused by a micro deletion in the centre controlling the imprinting process within 15q11-13. They found that in these cases there is a likelihood of recurrence within a family, unlike in any other genetic route to the disorder. The most recent contribution to genetic aetiology came in 1999 when Khan and Wood claim to have isolated the exact genes within the critical region which are significant to the PWS phenotype⁹, these being SNRPN and NDN.

From 1989 onwards links have been made from these genetic routes back to the diagnostic criteria i.e. the creation of specific genetic phenotypes. As early as 1989, Butler observed that individuals with deletions seem more likely to show the syndrome's typical facial features and hypopigmentation. And in 1995 Robinson found that people with UPD have increased birth weight, a shorter course of tube feeding in infancy and a later onset of hyperphagia. Apart from recurrence in families, little has been found linking those with micro deletions in the imprinting centre other than none of them have hypopigmentation.

Returning to the identification of cultural influences in the construction of knowledge on PWS, the impact of the dominance of molecular biology and the paradigms of new genetics within science (Davey in Williams et al 2003), have clearly had a massive impact on understandings of PWS. Further to this, the specific impact upon knowledge of shifts between different phases of genetics and their related technologies are also apparent.

In her examination of the history of human cytogenetics, Thurman (1993) distinguishes between five distinct phases, the last three of which are of particular relevance to the

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⁹ The observable characteristics of an individual, which result from interaction he possesses (genotype) and the environment. Also, the expression in a person or on a cell of characteristics determined by the genes that are not fully defined. (Martin 1998: 503)
development of knowledge around PWS. The 'Trisomy' period, starting in the late 1950s made the connection between chromosome analysis and the study of congenital abnormalities and 'syndromes', with forerunners such as Down's and Turner's syndromes. These were both connected to specific chromosome abnormalities as early as 1959, leading the way for other syndromes to test similar hypotheses.

The 'banding' era was brought about by technological advances, chiefly Fluorescence Microscopy. This technology made visible horizontal bands of differential staining intensity. This allowed the identification of each chromosome and importantly the recognition of structural abnormalities associated with specific genetic syndromes. It was shortly after the introduction of high resolution chromosome banding techniques, in 1981 that the paternal deletion was discovered and linked to the vast majority of people with the syndrome.

The last cytogenetic phase described by Thurman is the molecular era, typified by advances in the use of DNA probes. The imprinting discoveries made by Nicholls in the late 80s and early 90s, highlighting uni-parental disomy, were reliant on the signature technologies of this phase.

These developments in the genetic aetiology of the disorder can be seen to have a great influence on how knowledge around PWS will develop in the future. Research into aspects of PWS carried out in other fields such as psychology, endocrinology, and neurology now tend to include a breakdown of results into the different genetic subgroups.

Returning to the questions initially posed:

1. Is PWS a distinct biological reality and biological sub-group of individuals?

Following the critical realist principle that the 'real' and its generative mechanisms exist
independently of human knowledge, individuals displaying the characteristics now commonly associated with PWS existed prior to its naming and formal classification in medical science. This is exemplified in the paintings of Eugenia Martinez Vallejo, painted 184 years before Down's initial recording of polysarcoma. These real biological differences have a direct impact on the individual, affecting their modes of processing information, perceiving social situations, experiencing emotion and hunger and communicating.

Equally, understandings of the distinction of people with PWS are an integral part of the concepts which are specific to our social environment, expressed in language and sustained by the social processes which have formed around them. That is, although the biological differences found in people with PWS are real, what makes them meaningful are the social structures within which these people interact. Drawing on Burkitt's (1999) analysis, multidimensional networks develop between embodied beings, who share common understandings and symbols, they actively transform the real, utilising the materials around them. The existence of such a network in relation to PWS could have the capacity to change the experience of that illness. For example, in contrast to the paintings of Eugenia Martinez Vallejo, where the subject is described as 'the monster', the fact that PWS has been medically defined may alter everyday experiences for individuals with the condition and their families. Although the stigma attached to obesity, learning difficulties and behavioural problems in contemporary western societies should not be underestimated, various types of emotional, financial, legal and educational support, as well as, biology-altering pharmacological treatments are now available to people with PWS and their families. Early diagnosis through genetic testing allows families to be informed of the nature of the disorder and prepare for managing their child. Thus, although the biology remains constant over time, the social structures, networks and materials which have developed since PWS's medical definition affect how the disorder is experienced day-to-day.

2. Are variations within the disorder accounted for in medical understandings?

Any discussion of variation within the disorder needs firstly to concede that these
variations themselves, no matter how biologically real, also have some degree of cultural specificity. Whether these variations are behavioural manifestations, intelligence quota, physical disabilities or genetic route information, all of these concepts have been previously constructed in relation to various medical fields and reflect wider trends within the medical disciplines.

Developments in defining the clinical features of PWS have displayed a shift away from looking at one key feature to typify or define the essence of the syndrome and towards an acceptance of the many systems involved and the many forms the syndrome can take. Similarly ideas on the causes of the syndrome have diversified. As Foucault (1973) explained, the physical body became central to medical thought and medical explanation from the late eighteenth century, with bodies that were deemed abnormal becoming subject to medical interrogation. As the dominance of the physical body in medicine is replaced by the genetic paradigm, physical features become secondary to more abstract notions of chromosomes. A large amount of research continues into hypothalamic causes to account for particular physical aspects of the disorder. So, the development of medical knowledge on PWS displays a move towards acknowledgement of variation within the disorder.

3. How are developments in medical knowledge of PWS linked to broader medical trends and technologies?

The medical interception of individuals with characteristics now commonly associated with PWS did not occur in a vacuum. Trends in medicine described by Foucault (1973), physical examination and the medicalisation of 'abnormal' physical properties, and Armstrong (1983), child population surveys, clearly played a part in shaping the development of the concept of PWS. Its discovery and the description of its features were typical of medical enterprises at that time. Down's recording of Polysarcia and the rapid development of clinical features throughout the 1950s and 60s reflecting the trend in nineteenth and 20th century medicine of recording, measuring and classifying deviation
from the norm. Blaxter (2004) notes the centrality of deviations from the norm ‘especially the normal range of measurable biological variables’ as a core principle of the biomedical model, the prominent model at this time.

Developments in the aetiology of the disorder were extremely closely linked to technological advances and prevailing ideas in human cytogenetics. These aetiological advances were made during the 1980s and 90s, as Western medicine entered a new ‘genetic’ paradigm. Becoming less concerned with simply measuring and classifying, medical conceptions of the body and its workings came to be redefined via the application of new genetic technologies (Charlton 1993).

4. What is the current dominating medical framework through which PWS is envisaged and explained and what impact can this be seen to be having on the setting of future research agendas?

PWS is described as a ‘genetic’ or ‘chromosomal’ disorder. Although viewed as a multi-system disorder, affecting neurological, endocrine, craniofacial and musculoskeletal aspects of individuals, genetic information has become the primary feature in the disorder’s diagnosis. Where diagnosis was once derived from the presence of clinical criteria (Holm et al 1993), since the late 1990s the definitive diagnostic test for the syndrome is chromosome analysis of blood tests, searching for the absence of the specific gene SNRPN RNA (Wevrick et al 1996). Thus, genetic abnormalities are present in all cases and research continues into how these relate to the body’s systems, in particular the hypothalamus.

The shift to genetic diagnosis has had an important influence on who is now included as having PWS, pointing towards a major medical redefinition of PWS as a concept. Paediatricians Gunay-Augun et al (2001) note that 16.7% of patients with a molecular diagnosis did not meet the 1993 diagnostic criteria. The extent to which PWS is now viewed through a genetic medical paradigm is illustrated in their comments, as they
interpret this discordance to suggest 'the published criteria may be too exclusive...we suggest revised clinical criteria to help identify the appropriate patients for DNA testing for PWS' (Gunay-Augun et al. 2001: 92)

Where clinical features now serve only to raise suspicion of PWS, it is suggested that these criteria are broadened to maximise the potential of finding individuals missing SNRPN RNA. This demonstrates the secondary status that clinical features now have in understandings of PWS, as genetics come to be seen as the definitive factor in the disorder. As a result of this redefinition an ever increasing number of individuals are being diagnosed with PWS.

The dominance of the genetic paradigm within medicine has been observed widely within sociological fields of science, technology and medicine. Basiro Davey (2003) notes the tendency towards determinism within the genetic framework:

> The gene remains the dominant biological motif at the start of the new millennium. Dawkins' gene-dominated vision chimes with the sentiment expressed by Watson that everything beyond the molecular is subsidiary to the action of genes and that human culture is a consequence of genetic evolution. (Davey 2003)

Opposition to the viewpoint of the gene as the sole agent of evolutionary change has come from various disciplines, not least molecular biology itself (Rose et al. 2000). Within sociology the response has taken the form of close investigation of new genetics; its underlying assumptions, possible motivating factors, the work carried out in its name and the overall effects of the industry on individuals' and society's perception of self, kinship, behaviour etc. (Finkler 2000; Pilnick 2002).

In terms of shaping the research agenda and hence the concept of PWS in the future,
advances in knowledge of the genetic routes have meant that where research is conducted in other medical fields some comparison of these genetic concepts is built into the research design as standard. For example findings from recent research into mental health in the disorder are organised into UPD cases compared to paternal deletions (Boer et al. 2002), as were findings from neurological research into the physiology of the corticospinal tract\textsuperscript{10} (Civardi 2004).

Answering these questions provides an applied demonstration of my theoretical contention, that PWS can be taken both as a biological reality and a social construct. In line with conceptions from critical realism, medical research is seen as valid and of great value in alleviating the experience of disability for people with PWS; biology has a direct impact on individuals' emotional experience and identity and medical sciences offer legitimate ways of understanding and managing this biology. However, utilization of Burkitt's (1999) theory of embodiment illustrates that biology is only part of social process which determines these individuals' everyday experience of disease. Networks of relations between embodied beings, incorporating the biological, material, ideological, linguistic and symbolic, continuously transform the real and in this case govern the experience of people with PWS and their families. Tracing the history of the concept of PWS allows us to recognise the culturally subjective nature of medical knowledge, thus bringing it into a social science framework. If we can integrate medical knowledge, to some degree, into social science knowledge, this will help to develop a theoretical framework from which we can fully understand the experience of PWS and in this thesis, draw on scientific sources where appropriate.

1.4 Morality and Knowledge

We have seen that people with PWS share real, biological characteristics that determine the manner in which they perceive and interact with their social environment. Also, that this is a two-way process, where the ideas which underpin that social environment also

\textsuperscript{10} A type of neurone found in the cerebral cortex, with a pyramid-shaped cell body, a branched dendrite extending from the apex towards the brain surface, several dendrites extending horizontally from the base, and an axon running in the white matter of the hemisphere (Martin 1998: 552)
give meaning to and define these biological characteristics. So far we have examined ideas from medical science in relation to what PWS is and commented on the subjectivity of this knowledge in terms of its close connection to prevailing paradigms and practices in medicine. We now turn to look at another facet of the subjective nature of this knowledge; that is, the various ways in which medical discourses on PWS are related to social moralities.

Like Armstrong, Turner's (1987) investigation of *Medical Power and Social Knowledge* concentrates on the importance of the ideas of Michel Foucault. As he justifies in the second edition, Foucault's work offers 'a systematic approach to medical institutions, governmentality and the human body' (Turner 1987: 1). Thus, Turner observed a particular aptness of Foucault's philosophy to the sociology of medicine, as his analysis of power and knowledge was highly concerned with the body and regulation of bodies in society (Foucault 1973, 1977). Foucault's contention that power and knowledge imply one another is a useful starting point for assessing how morality and values have become ingrained in medical knowledge and practice around PWS.

Commenting generally on medicine's position in society, Turner argues that wider processes of secularisation and rationalisation have afforded the medical profession and its knowledge the position of moral guardian over individuals. He observes that the power of the medical profession to define and classify deviance, the development of institutions around the hospital and the organisation of medical surveillance of society have resulted in a situation where 'the doctor has replaced the priest as the custodian of social values' (Turner 1997: 35). Lupton (2003) expands on Turner's ideas through an exploration of 'illness as metaphor'. In her work on *Medicine as Culture* she describes how religious metaphors have been intertwined with ideas about medicine, the body, health and illness since the seventeenth century in Europe.

Examples of the use of moral metaphors in PWS are visible in the way medical research groups present themselves and their work. The homepage for 'The Foundation for Prader-Willi Research' (FPWR) [http://www.pws.pwsresearch.org/](http://www.pws.pwsresearch.org/) illustrates clearly how
medicalisation is imbued with political and moral discourse. The first animated sequence on the website displays medical components such as hormones, pharmacological agents and gene sequences as pieces of a jigsaw puzzle. When the puzzle is complete it reads ‘Hope’ and the site asks, ‘Help us find the missing pieces of the puzzle, help us cure PWS’. The research foundation’s slogan, ‘Harnessing the power of research to render PWS powerless in the lives of those we love’ sets out a distinct moral battle. Medical knowledge being the protagonist, PWS the antagonist and the patient as the passive battlefield through which the conflict occurs. This analogy displays extreme negativity towards the disorder. It effectively deems the subject with PWS, or through their image the ‘PWS part’ of the person, to be deviant and undesirable, whilst depicting that ‘hope’ and ‘a cure’ will be derived from medical components.

Foucault’s analysis of the power/knowledge relationship and its societal effects are useful in reflecting on how medical knowledge informs other structural knowledge and action, such as law, social welfare and benefit structures and media reporting. However, observations of the personification or attribution of morality to PWS is more resonant with bodies of literature concerning medical metaphors. Susan Sontag’s (1979) essay ‘Illness as Metaphor’ describes the dualistic nature of the way western thought conceptualises health and illness into good and bad. She uses the cases of tuberculosis and cancer to illustrate how our apprehensions of illnesses are saturated with emotive images, which she argues negatively skew our understanding and experience of them.

Everyone who is born holds dual citizenship, in the kingdom of the well and in the kingdom of the sick. Although we all prefer to use only the good passport, sooner or later each of us is obliged to identify ourselves as citizens of that place...My point is that illness is not a metaphor, and that the most truthful way of regarding illness- and the healthiest way of being ill- is one most purified of metaphoric thinking. Yet it is hardly possible to take up one’s residence in the kingdom of the ill unprejudiced by the lurid metaphors with which it has been landscaped. (Sontag 1979: 3-4)
Focus on medical metaphors in specific illnesses and biological processes has become commonplace in critiques of scientific knowledge. Emily Martin (1991) draws attention to the naturalization of socially constructed gender roles via medical descriptions of reproductive processes. Literature on the representation of HIV/AIDS is also prominent in this field, with Sontag returning to this subject (1988) to expand her original ideas on illness as metaphor. The simplistic dualism described by Sontag is detectable in the FPWR website (Foundation for Prader-Willi Research 2002-2005, source: http://www.pws.pwsresearch.org/). The medicine verses illness battle, which serves to abstract the subject from their condition, contrasts sharply with ideas from disability theory and even with current UK government agendas and ideas of disability.

Social constructionist ideas form the basis of much disability theory and are clearly evident in the ‘social model’ of disability. The social model highlights the agency of social knowledge, organisation and action in ‘disabling’ the individual, rather than, as traditionally seen, the agency of that individual’s biology. Although this model has been open to the same ‘social determinist’ criticisms as social constructionism, as seen in William's (1999) critique, it remains the most important model of disability within the field. The social model of disability stands against both the biomedical conceptualisation of illness and the moral values placed on medicine therein, both illustrated in the FPWR website and assuming the key biomedical aim of normalisation. As Barnes describes:

Biomedicine has as its focus on deviations of body and mind from socially recognised norms. Much biomedical hope and expectation are currently invested in a fast moving ‘genetic’ science which appears to its practitioners to offer a brave new world of disease eradication and radical new treatments for the restoration of normal functioning. In this ‘medical model’ perspective, disability continues to be equated with the impairment itself- ‘the disability’ is the impairment. The emergence of the social model of disability has had little or no impact in the heartlands of Western scientific medicine’. (Barnes et al 2001: 40-41)
At its strongest, the social model, being borne out of a political agenda towards the resistance of social oppression of the disabled, offers a reorganisation of morality. In line with Sontag’s (1979) objectives it sees biological difference, through illness, disease and disability as value-free, as opposed to negative phenomena. Instead of viewing medicine as holding all the answers to quality of life for the disabled, it advocates wider social change- in structures, organisations and attitudes, thereby stripping the medical profession of the moral status and power it holds in the medical model.

The moral battle of medicine against PWS, illustrated in the Research Foundation website, not only conflicts with the ideas which underpin disability theory, but also with government agendas on disability and thus public conceptions. Government initiatives such as ‘Images of Disability’, clearly display principles of the social model.

The Government is committed to promoting the rights of and improving the opportunities for disabled people. To affect real change, we need to bring disabled people into the mainstream of society. The implementation of the Disability Discrimination Act and the setting up of the Disability Rights Commission are examples of steps being taken to achieve this. However, the government realise that more work needs to be done to change stereotypical attitudes entrenched in society. Our vision for IOD is that: Government advertising and publicity will accurately promote the diversity of our society through the fair, honest and creative representation of disability. (Dept. for Work and Pensions 2002: 13)

The government's use of social model ideas, i.e. inclusion of difference by society, instead of normalisation of individuals by medicine, can be seen as part of a wider social movement of loss of faith in medicine (Le Fanu 2000) and the normalization of illness (Furedi 2003).

So with tensions in knowledge of conceptualisations of disability and the perceived role of medicine, between the public, government and medical science, what are the implications for PWS? Literature produced by the British Prader-Willi Syndrome Association (PWSA (UK)) can be seen to take the line of current government concepts of
disability. In the ‘Handbook for Carers’ (1996) there is an emphasis away from straightforward acceptance of medical interpretations of the syndrome, and information on PWS is not presented as belonging to medicine. For instance, the chapter from the Handbook on psychological issues and behavioural management starts, ‘In recent years a growing body of knowledge from both parents and professionals has identified several behavioural problems which occur frequently in people with PWS’ (Waters 1996: 30). The 2001 edition also stresses the importance of individuality within the disorder and suggests different approaches or solutions may work in different cases. This is seen in Waters’ advice on educational choices; ‘because children with PWS vary so much with regard to their abilities, it is important to remember that the school that is right for one child with PWS is not so right for another’ (Waters 2001).

The change in attitude can be seen when comparing this to the 1984 edition:

Some communities offer educational opportunities for handicapped children at a very early age, some even from birth. Contact the special education director of your school district. An early start can make a big difference. Some children with Prader-Willi syndrome have started pre-school at a time when they were functioning a year or more below their age level and have made enough progress to be able to enter kindergarten with normal children. If the child is involved in group activities with other handicapped children it is wise to also provide opportunities for him to be with normal children. It is your right and responsibility as a parent to obtain an education for your child that is appropriate to his needs. (PWSA 1984: 8)

This earlier advice is noticeably more instructive, generalises the needs of people with PWS and concurs with the biomedical principle of normalisation.

So far our analysis of the concept of PWS has shown that people with PWS and their significant others are portrayed as passive agents in terms of contributing to concepts of PWS. PWSA (UK) was founded in 1981 by parents of people with PWS, its main objective being ‘supporting people with Prader-Willi syndrome (PWS), their families,
The tradition for parental involvement in the charity is still apparent. Jackie Waters, a director of PWSA (UK) and the author of many of their publications, is the mother of a 28-year-old with PWS. As the only UK charity dedicated to providing information and support regarding PWS, the charity's work is widely read by people with PWS and their carers as well as health, education, and welfare professionals. Through their work, we see not just the agency the families of people with PWS to inform and add to understandings of PWS, but also a contribution from people with PWS themselves. The PWSA (UK) website and handbooks include descriptions, life stories, case studies, photographs, and the drawings of people of all ages, many of these accounts written by the people with PWS themselves.

There has been a recognisable shift in concepts of disability towards an acceptance of aspects of the social model, however it is important not to overstate the tension between medical and public knowledge. Although current literature from PWSA (UK) displays a movement away from biomedical principles and objectives of normalisation, it has been argued that a similar movement is occurring within medicine, in particular in the area of new genetics, where PWS research concentrates. Scully observes the post-modern nature of genetics, arguing that accusations of biological determinism are misplaced.

In its purest form, the genetic model claims only that a variation in the DNA sequence is the prime cause of a pathology, but makes no statements about exactly how the path from gene to physical manifestation - the phenotype of the organism - is followed. The way is left open for a variety of ideas about how genes function in development and disease, including approaches that hold no single component - whether genes, cellular morphology, biochemistry, environment - to be 'the bearer of ultimate control. (Scully in Corker et al 2002: 50-51)

Issues regarding how people with PWS should be governed by state service providers and within families are complex, due to the equally complex nature of the disorder. In particular, there are tensions between domination and restraint, and individuals'
autonomy and freedom. As a governmental model, the social model of disability stresses the importance of self-management and autonomy as the rights of people with disabilities. However, due to the lack of capacity for decision-making around food within the disorder, it can be argued that placing some external constraint (around food) actually allows these individuals greater freedom and quality of life, protecting them from the potential dangers of their preoccupation with food. It follows that adhering to social model principles, the carers of people with PWS have an obligation to intervene with regard to limiting access to food, even where this conduct conflicts with the immediate wishes of the person with PWS.

Clinical psychologist Anthony Holland and Human Rights law specialist Josephine Wong (1999) outline the ethical and legal dilemmas that can arise in cases of PWS and set out suggestions for future decision-making. Disagreements occur most commonly in relation to the level of autonomy the PWS adult should be granted in making decisions about what he/she eats. European Human Rights law sees this autonomy as central to any democratic society, as long as the person is ‘appropriately informed’, has the ‘capacity’ to make the decision and ‘is free from undue influence’ (Holland et al 1999: 232).

Psychologists have argued against the idea that people with PWS have the capacity to make decisions around eating behaviour. Dykens (1997) describes the relationship of people with PWS to food as ‘physiological not motivational’, arguing that eating behaviour is biologically determined and outside of the control of individuals. Holland et al (1999) suggest that PWS should be made a special case or exception to human rights law and that, in these cases, the common law principles of ‘acting in a person's best interests’ using ‘the least restrictive alternative’ should be applied.

The details of legal discussions on this matter show that medical knowledge is still central to determining legal decisions. That is, even though a public movement towards lack of faith in medicine and science has been suggested (Le Fanu 2000), these disciplines and their knowledge are still powerful authorities in the structuring of social life for those with PWS. In most European countries, if someone is classified under the
Mental Health Act, treatment is compulsory. Thus if individuals with PWS come to be certified as having a 'mental impairment', legal issues around autonomy, individual choice and civil liberties can be avoided. Even though this labelling may not be the most appropriate description of the complex factors involved, it forms a common solution, especially where obesity becomes life-threatening and restriction of food intake the major priority for the individual and their support network.

The recent, highly publicised, case of Chris Leppard, a twenty-three year old with PWS, highlights many of these issues (See figure 1.5). Shortly after featuring in a BBC television documentary on the disorder, Chris Leppard was sectioned by police and social services against his wishes and those of his family. The documentary had portrayed Chris Leppard struggling with his eating behaviour, putting on weight and making an argument towards infringement of civil liberties, as he compared his addiction to smoking behaviour. After his sectioning he was detained in a mental health clinic for a week. He has subsequently lost weight. Although sectioning under the mental health act is usually avoided, Chris Leppard's example displays this medical/legal categorisation being used to control eating behaviour. It also displays an attitude within social services and health professionals that people with PWS lack capacity in decisions around food and that the state has an obligation to people with PWS, where eating behaviour becomes life threatening.

The issues raised in these cases are problematised by the widespread reporting of obesity in the Western general population. Holland et al (1999) outline the conceptualisation of PWS as seen by those who argue for individual choice and highlight how the rise in obesity in the general population undermines arguments for the forced restriction of eating behaviour in PWS.

One way to conceptualise this dilemma is to consider people with PWS no differently from those in the general population, and more specifically, no different from those people who are overweight. Obesity, with all its associated health problems, is endemic in Western society. In the UK a reduction in obesity
has been one of the targets for the “Health of the Nation” campaign. As humans we appear constitutionally ill-prepared for an environment where food and the money to buy food are readily available. Governments and health care professionals can inform and encourage but cannot insist. As a society we would not sanction regulation or restriction of food intake for people whose health is at risk, without their consent. This would be considered an unacceptable infringement of civil liberties. Is overeating behaviour in PWS an extension of the same behaviour so common in the general population? (Holland et al 1999: 232)

Thus, the sharp rise in levels of obesity across Western society calls into question the boundaries of PWS as a medical concept. It questions the medical concept of PWS by asking; to what extent should we view eating behaviours in PWS as different from the general population? therefore also questioning the differential treatment through medical, legal and welfare structures, in relation to enforced food restriction.

A second problem in the reported rise of obesity, and its prominence on the public health agenda, is the associated media coverage. Particularly where childhood obesity is concerned media coverage can be likened to a moral panic. Although an increase in the body mass index (BMI) of children has been widely documented, many commentators, from sociological, as well as political and journalistic, backgrounds are of the opinion that the current policy agenda and media coverage of childhood obesity is an overreaction.

These critiques often draw attention to the rather moralistic nature of reporting of childhood obesity issues in the media. The Social Issues Research Centre conducted an analysis of the statistics on obesity within the government’s Health survey for England 2003. Their conclusions illustrate this concern over the misrepresentation of childhood obesity:

The framing of appropriate strategies to tackle obesity needs to be firmly based on accurate assessments of both the scale of the problem and the populations most at
risk, rather than on population-wide approaches coupled with sensationalist claims and the quite unjustified use of terms such as ‘epidemic’ or ‘exponential rise’. (Social Issues Research Centre 2005: 3)

A sociological approach to childhood obesity makes clear that it is a far more complex issue than is suggested by an apparently ‘rational’ government reaction to the medical facts. Rich and Evans (2005) raise a number of ethical issues about the ways in which the ‘obesity crisis’ has been socially created and publicly represented. Lee Monaghan (2005) similarly questions the obesity epidemic. He draws attention to the subjective nature of ‘excess’ weight and calls for ‘the science legitimating the war against fat to be questioned as well as the arguments offered by those playing a key role in constructing this as a massive public health problem’ (Monaghan 2005: 1).

This moral crisis and its media representations have been seen to demonise ‘excess’ weight to an extent that may, ironically, damage health; ‘The proponents of the ‘obesity epidemic’ are pushing...towards dangerous surgeries, crash diets and harmful diet drugs’ (Oliver 2006: 3).

(Figure 1.5 Hastings Today 2005,
The media attention and public interest which constitute the moral crisis on obesity have visibly skewed public knowledge and perceptions of PWS. Until recently media coverage of PWS was scarce. It most commonly took the form of occasional ‘life-story’ profiles in women’s magazines, where food, eating behaviour, weight-loss and body size have long been part of the agenda. Following the BBC documentary that portrayed Chris Leppard and other cases, there has been a marked interest in the disorder, especially from the tabloid press. The pictures and headlines from Leppard’s case clearly display the bias in media conceptions of PWS towards the eating behaviour aspect of the disorder. The front page of the British tabloid newspaper ‘The Sun’ read, ‘Fat Police Lock up Chris’ (The Sun 2005: 1). Headlines in local press had similar levels of sensationalism; ‘31-stone man eating himself to death’ and ‘31 stone man locked up for his own good’ were headlines in ‘Hastings Today’. The photographic material i.e. camera angles used, can also be seen to exaggerate or sensationalise the real (see figure 1.5).

As with the PWSA publications, media coverage such as Chris Leppard’s story and television documentaries on PWS (BBC1 2005, Channel 4 2006), present conceptualisations of PWS which allow the voices of people with PWS to be heard. However, the individuals selected by British tabloid newspapers such as The Sun, are extreme cases which focus specifically on food related behaviours and obesity within the disorder. There is evidence of the same ‘selective’ and misrepresentative practices in the content of television documentaries, which would claim to give a fuller picture of the disorder. Whilst conducting this research I have been contacted by two television documentary makers, both specifically seeking participants who were obese and had marked problems in their behaviour around food. Similarly media requests for participants for various television documentaries, circulated via PWSA (UK) to their members, specified the importance of obesity and problematic behaviour around food. Thus media conceptualisations of PWS tend to present extreme and misrepresentative images of PWS, accounting for diversity within the disorder to a lesser extent than other knowledge bases discussed.
In addition to sensationalising and misrepresenting PWS, the moral panic around childhood obesity may have a considerable impact on those with PWS and their carers. Within media coverage of childhood obesity, the emphasis is on parents and guardians to take precautions against the ‘epidemic’. For instance, advice and information on childhood obesity detailed on the BBC official internet pages is clearly aimed at parents rather than the children themselves, the main subheading include: ‘Should kids ever be made to finish what’s on their plate? Can giving kids food as a reward cause obesity? Should I tell my child off for being fat? Is it worth forbidding fattening foods? Should I offer my child a choice of food? Are ‘weight management camps’ a good way to treat obesity? (Wardle, 2004, source: http://www.bbc.co.uk/science/hottopics/obesity/advice). Parents of people with PWS may find themselves under increased pressure to control the weight of their children, as avoidance of childhood obesity is increasingly viewed as the moral responsibility of the parent, rather than merely a medical issue.

1.5 Conclusion

PWS is simultaneously a biological reality and a social construction within medical knowledge. Although medical knowledge is seen as valid and of great use in alleviating the experience of disability for people with PWS, i.e. Through various medications and treatments, the medical label is not constitutive of the biological reality it describes. The defining of PWS and medical knowledge concerning its clinical features and aetiology has derived as much from societal modes of surveillance and regulation and shifting scientific paradigms and their associated technologies, as it has from the biological reality which forms its subject. This recognition of the subjectivity of medical knowledge allows it to be brought into a social scientific framework, reducing the polemics of realism and relativism which can be seen to underlie science and social science in more determinist approaches and creating a theoretical framework through which the disability experience can be fully appreciated.
The concept of PWS is diverse appearing in medical, legal/ethical, charity/welfare support and media contexts. Representations of PWS vary between these contexts, particularly in relation to moral issues around the disorder. Some medical representations of PWS, as with other disorders, tend to portray their work through the metaphor of a moral battle of good against evil; medical research against disabling condition. Legal and ethical knowledge concentrates on the moral conduct of society in supporting those with PWS. In both of these conceptualisations the person with PWS is treated more or less as a passive agent, the definitions produced and decisions around them occurring at an institutional level. Charity/welfare support literatures and media representations include the voices of people with PWS and their significant others, allowing them agency in the process of knowledge construction of their condition. However, where charity literatures are keen to portray variation within the disorder, much media coverage focuses specifically on obesity and behaviours around food, often ignoring physical and behavioural aspects of PWS which are not related weight issues.

One other context in which the concept of PWS is understood, but is yet to be explored fully, is of individuals with PWS and their families. How people with PWS and their significant others understand PWS, how they draw on various existing knowledge as and its linking to morality, as well as their own lived experience and how this affects identity and everyday management strategies can be explored further within this research.
CHAPTER 2: LITERATURE REVIEW

Introduction
Chapter one drew on a number of literatures in order to explore the diverse knowledge of PWS and develop a theoretical conceptualisation of PWS that can be used throughout the thesis. We have so far examined social science literatures on the construction of medical knowledge, how relativist and realist philosophies appear in medical and social sciences and attempts to synergise these positions. This examination focused specifically on how this has been achieved in the fields of medical sociology and disability studies. Literatures on morality and medical knowledge have also been related to our subject.

A study of the management of children with PWS within the family, however, requires a number of further sociological literatures to be drawn upon. Specifically this calls for a brief overview of research in areas of embodiment, the experience of chronic illness and disability within the family, the impact of social factors on the experience and management of illness, research on obesity and the sociology of food. This chapter will also examine literatures on the management of PWS within the family from medical and social work perspectives. The aim of this review is to illustrate the nature and findings of sociological research connected to the research topic, with a view to the formulation of research questions and research design.

2.1 Embodiment
Chapter one introduced the concept of embodiment through the ideas of Merleau-Ponty’s Phenomenology of Perception (1962) and Burkitt’s Bodies of Thought (1999). However there is a need to explore wider literature on this topic in order to identify how we intend to define and use the concept of embodiment throughout this research.

The past decade has seen the utilisation of the concept of human embodiment to explain social action in various fields of sociological enquiry. Phenomenological philosophy is credited for providing the basis for recent conceptualisations of embodiment in sociology, particularly the contribution of Maurice Merleau-Ponty’s Phenomenology of Perception (1962). In Phenomenology of Perception, his account of the body unites the objectified
and symbolic body with the everyday subjective body through which individuals perceive the world. Through his recognition that human knowledge is gained through humans' corporeal experience of the world, he breaks down the Cartesian dualism of mind and body. According to his analysis it is this practical experience that is key to affecting human perception of the world and which influences human action within it.

This conceptualisation of perception, not only emancipates the material body to the status of the inner mind, but also demonstrates an essential connectedness 're-establish the roots of the mind in its body and in its world' (Merleau-Ponty 1962: 3). Although this is a work of philosophy, its implication as a theory of social action is strong as it paints a distinct picture of the social world and the relationships and interactions that occur within it.

We are in the world through our body...we perceive that world within our body...by thus remaking contact with the body and with the world, ...we discover ourself, perceiving as we do with our body, the body is a natural self and, as it were, the subject of perception. (Merleau-Ponty 1962: 206)

Merleau-Ponty’s recognition of the essential connectedness of the corporeal body and the development and nature of human knowledge allows the acknowledgement that bodily differences may lead to differing perceptions of the social world. His theory of perception affords individuals agency by acceding that their experience is dictated by more than the external discourses and knowledge held within their society. Although this process can be built upon by exploring some of the meanings and attributes attached to bodies in any society, the process itself is not reliant on these external factors. Iwakuma (1999: 78) observes Merleau-Ponty’s exploration of the relationship between the subject and object and displays the applicability of his notion of ‘embodiment’ to the sociology of the body, health, illness and disability.

Merleau-Ponty (1962) calls the body a ‘grouping of lived-through meanings which move towards equilibrium’ (p.153). He introduces the innovative idea that the body ‘extends’ an object, for example a cane for the blind, so that it literally
becomes part of the body. This ‘extension of the bodily synthesis’ is a process of embodiment (p.152), and this search for equilibrium also seems to be the core of adjustment by a person to his/her impairment. (Iwakuma 1999: 78)

The incorporation of human embodiment into the sociology of health and illness research agenda has occurred against the backdrop of ‘the body’ being explicitly introduced back into sociology more broadly. It was not until the publication of Bryan Turner’s The Body and Society that the sociology of the body was delineated and pushed forward as a key area of social enquiry with a specific research agenda. With hindsight, it has been noted that differing interpretations of the body and its role in the social world are present in classical sociology, social anthropology and philosophy, reappearing significantly in medical sociology and feminist theory in the 1970s (Turner 1984). From the mid 1980s onwards sociology has witnessed a significant rise in analyses of the body/society relationship often linked with consumer culture, feminist theory, embodiment, social action and the self in late modernity.

Unlike studies about the body, where the body is the central subject of investigations, the term ‘embodiment’ or an ‘embodied sociology’ (Bendelow et al 1998) invoke a more subtle approach. That is the idea of embodiment as used by Burkitt (1999), Bendelow and Williams (1998) and Crossley (2001) suggests that what is social is simultaneously corporal or as Crossley (2001: 3) puts it, questions of embodiment seek to explore, ‘both the corporeal nature of social life and the social nature of the human body. To take an embodied view is to acknowledge that social action occurs through the medium of the body.

An embodied sociology addresses the problem of Cartesian dualisms within social science approaches to action. Bendelow and Williams (1998) highlight that the notion of mind being separate from the body, originating in the philosophies of Descartes and Kant, has led to many binary assumptions which underpin Western thought. These assumptions, from an embodied approach are false and have been the base of various social inequalities throughout modernity. For example the dualism between men/women
became mapped on to public/private divisions, similarly throughout colonialisation the ideas of ration/emotion were used to characterise occident/orient (Hall 1992). Crossley (2001: 23) puts forward the notion that the mind itself is in fact a fallacy, constructed to represent the spiritual essence of the person. He conceptualises humans as whole bodies which incorporate brains and sees the brain as by no means reducible to the mind.

It is from the work of Merleau-Ponty (1962), Bendelow and Williams (1998), Crossley (2001) and Burkitt (1999) that I conceptualise embodiment in relation to the study of the management of PWS within the family. Acknowledging that the body is central to social action and order, on both micro and macro scales, has an interesting implication to our subject. As the PWS body is significantly differentiated from other family members this may affect the social perception and behaviour of the person with PWS, meaning extra work must be undertaken by all family members to maintain order within the family, with differences in biology simultaneously equating with differences in the social.

2.2 Experience of Chronic illness and Disability
Akin to embodiment literatures, literatures on the experience of illness and disability acknowledge that the changes in the biophysical have significant social consequences, impacting upon sense of self, relation to others and to our material environment. However, many of the classic texts in this field, such as Corbin and Strauss’s (1985) study of the work chronically ill people undertake to maintain their social position and Bury’s (1982, 1991) explorations of individuals’ adaptation to chronic illness, approach the experience of illness/disability from the point of view of adults who have at some point been well and value that status. As PWS is a condition present from birth within which people experience learning disability and autistic-spectrum disorders, thus experiencing a significantly altered social perception, it is hard to apply these theories to the ways in which people with PWS cope with everyday life.

The work of Connors et al (2002) provides an alternative, child-centred account of the experience of illness, from both disabled children and their siblings. Drawing on child-friendly, qualitative sessions with twenty-six disabled children aged seven to fifteen years
and twenty-four of their siblings aged five to nineteen, the identities and daily relationships of the children are explored, as well as their dealings with professionals. The findings reveal a positive and pragmatic approach amongst the young people, who describe being generally happy both at home and at school. The children identified some level of bullying which tended to occur outside of the home and a sense of being restricted by the attitudes of certain teaching staff and over-protective parents. Siblings describe the disrupting effects and limitations within the family setting. Although antagonistic relations between siblings were described, the general picture of family life is harmonious. Brothers and sisters display concerns for their disabled sibling’s physical and emotional health which often remain unexpressed within the home for fear of burdening parents.

Although Connor’s (2002) work importantly addresses the child’s approach to everyday experience, it takes a strong social model approach, primarily focusing on how findings should affect health and welfare provision. Rather than exploring the everyday practicalities of dealing with the actual, physical illnesses, it examines social experiences of prejudice and oppression, highlighting the need for an extension of the disability rights movement amongst children with disabilities and a wider support structure for their siblings. These issues have similarly been raised by other authors such as Burke (2004), Davis et al (2003) and Bevan (2004). These studies draw attention to the need to explore the roles of all family members in negotiating daily life within the family, particularly the impact on siblings. As this thesis is concerned to display the practical actions and strategies that constitute the management of a complex physical disorder, it is necessary to move beyond the conceptualisation of the experience of disability as oppression, adding to this an acknowledgement of the physically disabled body within the family.

Research findings drawn from work on the intersection of the sociology of childhood and the sociology of the body, provide more in-depth considerations of children’s relation to their bodies. Berry Mayall’s book ‘Children, Health and the Social Order’ (1996), is the result of two in-depth school studies; parents, staff and children aged 6-10 years. Taking
on the theoretical view of the child’s body as a ‘lived entity’, she discusses in detail the political aspects and everyday interactions of home and school life. Her findings have some interesting implications for children living with PWS. When commenting on the politics of health in society in general, she observes that parents find their experience-based inputs and thoughts referred to as ‘lay knowledge’, whilst children’s’ accounts of their own bodies are often treated with suspicion at school:

Children in particular find themselves the objects of health care rather than the subjects. They are objects of large-scale interests: those of market-led purchasers and providers in the NHS, of health professionals, the epidemiologists of the normal, of the psy-complex across the home, pre school and school’ (Mayall, 1996: 41)

As PWS affects the child in multifaceted ways and is seen to incorporate a variety of threats to health, it is inevitable that health will be a much greater focus for these children and their families. This may result in Mayall’s ideas of processes of objectification in childhood being magnified for these children. This objectification may pass on to the child a reduced perception of their own capacity for agency in decision-making and responsibility in health, more so than with unaffected children. This opens up a debate about whether the high levels of dependency occurring throughout the PWS life are to some extent learnt in childhood. There are obvious limitations to the PWS body and it cannot be denied that living with PWS in contemporary society demands more support than otherwise. However, if as children these individuals are taught to respond to a range of institutions in a passive and submissive manner, it is possible that these interactions will shape PWS individual’s self-image and persist later in life.

Societal responses to child illness are important to Mayall, but as a researcher who adheres to notions of the lived experience of the body, she also looks beyond this, narrowing her study down to individual perceptions and experiences. From mothers’ accounts of their children’s health, Mayall asserts, ‘women’s day-to-day people work forces them to recognise the bodily, and interlinkages between bodily and emotional
well-being’ (Mayall, 1996: 88). What Mayall is advocating is that people experience life as embodied beings; the physical, social, psychological and emotional all working on each other. Further to this, she recognises that parents acknowledge this aspect in their children and to some extent children themselves are aware of it, ‘Children recognise that physical input to learning promotes both enjoyment and achievement’ (Mayall, 1996: 109). As PWS bodies are medically considered as physically limited, through Mayall’s conceptualisation, this may have a profound effect on the children’s overall well-being.

The work of Allison James (1995, 2000) also takes an embodied approach to the child body. She expands on the ideas we have seen presented by Mayall, using a methodology that resembles Mayall’s; an ethnography of school children between 4 and 9 years of age. Unlike Mayall, James’s work addresses the impact of social environment in relation to other embodied peers. In extension to this, James notes that her ideas about bodily change being used as a marker of child identity, are based on the premise that children experience their bodies in terms of others. James (1995) describes a process where comparing bodily difference becomes crucial to children’s meaningful definitions of themselves and of others. According to James, this intense focus on the body in childhood is partly due to the rapid physiological changes that occur at this time, thus in adulthood meaning is derived from the body less so. James (2000) outlines the specifics of how children attach meaning to aspects of the body such as, height, size, shape and gender. She finds that although children’s beliefs generally reflect those of adult society e.g. fat bodies holding negative connotations, the children actively produce reasoning and explanations for these convictions, based within the lived context of their environment. Christensen (2001) confirms James’s findings on the significance of the body in day-to-day interactions within childhood, drawing attention to the way children display cuts and bruises, asking for reassurance that their bodily reaction is normal (Christensen 2001: 40).

In connection with PWS, children create meaning around their bodies in relation to the bodies of their cohorts. Due to the rarity of PWS in the population, this indicates that children with the disorder may develop a more negative self-identity, their physical
development displaying marked differences from those around them. James summarises this point about the importance of physical difference in childhood with reference to height. Her comments are particularly relevant to children with PWS who often experience short stature:

Achieving and fulfilling the status of a child is...understood by children to be accomplished partly through abandoning the status of infant and one very eloquent statement of being a normal child lies in possessing a body which is child sized. Thus children whose bodies are smaller than those of their contemporaries may gradually become self-conscious of their difference through a dawning recognition of their own body's failure to conform'. (James, 2000: 70)

As has been previously described, the PWS body is extremely prone to weight gain. First, its composition has a high fat to muscle ratio, making it hard to burn off calories, i.e. the average PWS intake maintaining weight is 900 – 1200 calories per day, much less than for an unaffected person. In addition, these people are characteristically in a constant state of hunger and obsessive behaviour around food is common. Considering these limitations, the PWS body is under the constant threat of obesity, although in many cases this is avoided through regulating strict regimes of diet, exercise and lifestyle. With the risk of obesity being constant within the syndrome, the social meaning of obesity is relevant to the experience of all young people with PWS and their carers. James (2000) examines the social meaning of obesity within her ethnography of 4-9 year-old school children. Here we return to the process of understanding bodies in relational terms, 'children do not passively accept the stereotypes of body shape and size which they encounter, but, instead, seek plausible explanations for negative stereotyping from their own bodily encounters' (James, 2000: 31). The children expressed notions of greed in relation to overweight people, always presuming that the weight resulted from eating too much. The children's reasoning for not wanting to be fat were based on problems that would occur within the context of their own environment e.g. not being able to do up shoelaces and not being able to run fast.
In her ethnography, James observed that the whole system of primary schooling was biased towards slim, agile people, ‘teachers exhort children to ‘walk properly’... ‘sit up straight’. Fat bodies cannot walk tall or sit straight. The body must be orderly: tying shoelaces and ties...neatens the body’s appearance...fat bodies are untidy bodies’ (James, 2000: 32). She also notes that there is an emphasis on physical play at school, where children are expected to go outside at every break, as well as partake in physical education lessons. According to James’s process of meaning creation it is exactly this kind of mainstream cultural valuing that forces children to work out, in their own context, why being fat is negative. Even more strikingly than this, the children associated fatness with being a bully or a generally nasty person. When hearing a poem about a bully a child said the character was a ‘fat bully’ and a ‘fat pig’. It seems, at least in James’s ethnography, that assumptions, meanings and values around obesity are deeply negative in Western society, forcing children to discover their own explanations based in their own environment. In addition to health concerns, these attitudes may put extra pressure on children with PWS and their families to make the PWS body conform to mainstream standards.

Bluebond-Langner’s (1978) ethnography of child leukaemia camps outlines the positive effects association of bodily experience can have amongst physically disabled people. Within these camps the terminally ill children used the characteristics which isolated and stigmatised them in normal society to identify with each other. As active, creative performers, the children chose to reverse the values of mainstream culture, using the bodily aspects it values least, such as gauntness and hair loss, as signs of shared identity. Unconditional acceptance of peers was a theme central to the camp, the children and workers created a social environment opposing James’s classroom culture, the concept of unconditional acceptance contrasts sharply with a world where every bodily difference invites scrutiny and moral judgement. Bluebond-Langner’s research is relevant to the experience of young people with PWS. Again due to the low incidence of the disorder within the population, it is unlikely that these young people will be able to access positive opportunities to share experience of their bodies with others with PWS.
It is important to note that although the empirical studies discussed have interesting implications towards the experiences of the young people in this research, the authors concentrate only on physical difference and disability. As people with PWS also experience learning disability and autistic spectrum disorders, their social understandings and emotional response to their environment is differentiated from those without the disorder. Lack of awareness of their physical bodies and underdeveloped social awareness are commonly reported within the syndrome. These facets of PWS mediate their experience of the body in childhood and may, to a certain extent, exclude them from the processes described in these studies.

Literatures on disability within the family and disability draw attention to the process of 'stigmatization' (see Cahill 1995, Link et al 2001, Gray 1993, 2002, Scambler et al 1986, 2004, Voysey 1975, and MacRae 1999). These studies are applications of Goffman's (1963) analysis of stigma, which describes the relational interactions, assumptions and symbols involved in the social stigmatization of individuals and the possible effects of this on the 'self'. Central to Goffman's theory are the concepts of 'virtual social identity' i.e. the assumptions commonly drawn about the nature of individuals, based on first appearances and 'actual social identity' i.e. the attributes an individual can be proved to possess (Goffman 1963: 12). Goffman describes stigmatization as a social process whereby a person is 'reduced in our minds from a whole and usual person to a tainted, discounted one' (Goffman 1963: 12) and proposes that this occurs when attributes are revealed (actual social identity), which differ from the attributes we expect a person to possess (virtual social identity). Goffman categorizes three different types of stigma: abnormalities of the body; blemishes of individual character; and tribal stigmas of race, nation and religion. These experiences of stigma occur for the person with the disability as well as their significant others; Goffman (1963: 30) refers to this as 'courtesy stigma'. Whether processes relating to stigmatisation are described by children with PWS and their families forms an interesting topic of enquiry in itself. Relating to the main aim of exploring management of the body, it is possible that avoidance of stigma, in addition to health concerns, may shape the management of the condition within the family.
As well as stigmatizing effects, the impact of disability on other family members has been discussed widely in relation to responsibility for caring within the family. Earlier in this section the caring work of siblings was discussed. Gender is also thought to play a key role in the patterning of care work within families of disabled children (Graham 1984, Beresford 1995, Thomas 1995, 1997, Ungerson 2000). Arber et al (1999) draw attention to the heavily gendered patterning of caring in UK families with disabled children, also noting how gender is intertwined with factors of class, ethnicity and changing social care structures. Questions of who undertakes childcare work within the families of children with PWS or who effectively manages PWS will be addressed within the research.

The high proportion of disabled children being looked after in lone-parent families has been commented upon within social trends literatures (Beresford 1995, Lewis 2002). Read (2000) highlights the decrease in material resources and emotional support which can be found within this group. The frequency of lone-parenthood in families with disabled children points to a need to include the accounts of such families within this research. In addition, questions of whether this impacts the everyday management of the condition will be addressed.

2.3 Health beliefs, behaviours and the sociology of food
Further to experience of illness and health status being determined by micro structures such as the family, sociological and epidemiological literatures identify the effects of wider social structures such as socio-economic status (SES) on human morbidity and mortality. Materialist explanations for health inequalities have drawn attention to contributory factors such as poverty, income, unemployment and housing conditions (Payne 1991). More commonly material factors are drawn into cultural explanations for differences in health outcomes. These explanations concentrate on the role of lifestyle, beliefs and behaviours in determining health. Although the chromosome disorder PWS is not caused by material or social factors, differing lifestyles, health beliefs and access to material resources could have important impacts on everyday management practices.
Behavioural and cultural explanations for health inequalities were initially highlighted in the Black Report (1982) as a more complex social explanation for social differences in health behaviours and attitudes, not reduced to blaming individuals with perceived risk-prone lifestyles:

[Non-cultural] explanations focus on the individual...emphasizing unthinking, reckless or irresponsible behaviour...as the moving determinant of poor health status. What is implied is that people harm themselves or their children by excessive consumption of harmful commodities, refined foods, tobacco and alcohol, or by lack of exercise, or by their under utilisation of preventative health care, vaccination, ante-natal surveillance or contraception. Some would argue that such systematic behaviour within social groups is a consequence only of lack of education, or individual thoughtlessness...Others [cultural explanation] see behaviour which is conducive to good or bad health as embedded more within social structures—as illustrative of socially distinguishable styles of life, associated with, and reinforced by, class. (Townsend et al, 1982: 110)

Research and surveys following the Black report outline these persistent differences in leisure and consumption habits. The most notable being smoking, exercise and amount of fat, sugar and salt in diet (Bartley, 2004: 64). Evidence from national surveys of health display that obesity outcomes are strongly correlated to social class position. For instance, obesity was reported as occurring in just 11.1% of women in social class I, yet 23.6% of women in social class IV (Colhoun et al, 1996: 24).

Understandings of what constitutes health (lay beliefs) vary between social groups such as age, SES and ethnicity, as do the behaviours that impact health status (Blaxter 2004). These cultural differences have most frequently been viewed in relation to social class. For instance D'Houtard et al's (1986) survey of health definitions in the French population found that definitions varied significantly in line with the respondent's occupation. Similarly participation in physical leisure activities is found to be more
common in higher SE groups (Wilson 2002, Jarvie 1994: 193). One theory which has been utilized to account for the higher social classes’ apparent ‘healthier’ lifestyle is Lefcourt’s (1976) ‘locus of control’. Locus of control refers to an individual's expectations concerning where control over subsequent events resides. In relation to health, this can be viewed in terms of social position, with members of the middle classes tending to see their behaviours as responsible for their future health outcomes and members of lower class social groups taking a more fatalistic approach, seeing themselves as out of control of their health (Elstad 1998).

It has also been noted however, that relationships between health beliefs and behaviours are not straightforward, particularly where it comes to social class (Blaxter 1990). That is, health behaviours and lifestyles do not suddenly transform upon the acquisition of new health information. Williams (1995) draws on Bourdieu’s notions of ‘practical knowledge and knowledge of practice’ (1990) to explain the complex relationship between class, health and lifestyles. William’s explains, as Bourdieu (1990), that individuals are not always aware of why they behave in certain ways. When asked, they may draw on ‘official accounts’ to justify their actions, but in truth their behaviour is based on their own practical logic; their own experience of what works, grounded within their own material and social environment or class culture.

Within the families of children with PWS, cultural and lifestyle aspects of SES may impact upon management strategies or goals. Differences between management approaches and health beliefs of families could be usefully explored in light of SES within this research. One area which may be of particular interest here is family conceptions of ‘healthy’ foods. Coveney (2005) found that beliefs about food and health were expressed differently dependent on the SES of families, commenting that this was particularly the case where children’s eating habits were concerned. Higher Socio-economic (SE) groups tended to use the terminology and concepts of nutritional science, where lower SE groups spoke about food in terms of its effect on the child’s appearance.

Differences between the foods consumed across classes are most notably illustrated in
Bourdieu's (1984) discussion of class distinction and taste, which incorporates detailed empirical data about class based food preferences drawn from a survey of over one thousand French people in the 1960s. Bourdieu observed differences not only in the types of foods eaten, but also in the manner food is prepared, presented and consumed. Similarly to Bourdieu's discussion of practice, food and other tastes are drawn from individual's immediate social and material environment. Caplan et al's (1998) study of food choice in Lewisham, London, noted similar class based distinctions between SE groups. Middle class families were seen to spend a higher proportion of their budget on food, emphasising the greater value placed on expensive, quality food products within this group. Within working class families, Caplan et al (1998: 177) identified that food often became the expendable part of the household budget when families faced economic hardship. Working class respondents described cutting out what were seen as healthier food choices in order to meet other financial needs.

Lupton (1998: 37-69) illuminates the particular significance of food within the family, its place in the maternal-child relationship, the nostalgic memories of food in childhood and the role of food in special occasions. She (1998: 38) observes that the emotion of love is constantly linked with food, describing how both motherly love and romantic love have come to be demonstrated in social rituals around food in Western family life. These observations of the symbolic importance of food within the family, understood and made real through embodied experience i.e. consumption, bring to light the emotional as well as practical complexities of managing PWS within families. How parents renegotiate their role to involve the denial of food to their children, whether food takes on the same emotional connotations within the families and how families deal with special occasions, are questions which can be carried through to the research.

Warde and Hetherington’s (1994) survey of food in families in greater Manchester, found that women still undertook the vast amount of cooking related duties within the home. The social symbolic importance of the feeding relationship between mother and child has been widely commented upon in social studies of food and the family (Homans 1983, Murcott 1988, Lupton 1995, 1998, Oakley 1979 and Shuttleworth 1993). Food has been
seen to take on immense importance for mothers in Western society from the point of conception, with the ‘good’ mother paying attention to what she consumes and its effect on the child (Lupton, 1998: 43). This responsibility is maintained throughout motherhood, with the infant’s body signifying the mother’s virtue. As Oakley (1979) describes:

A baby that is feeding and growing “well”, is a prize for the mother’s efforts, a tangible token of her love and work. Conversely, a baby who gains weight more slowly than it “should”, and perhaps who cries a lot, and seems unsatisfied, is a thorn in the mother’s flesh, a sign of maternal failure. (Oakley, 1979: 165)

With regard to these comments we can begin to see how these traditional relations are problematised for the parents of children with PWS. The infancy stage of PWS is marked by an inability to feed and an extremely weak and floppy child. The insatiable appetite that develops after infancy may also harbour a symbolic threat to the virtue of the mother, as teaching children to nourish and discipline themselves is traditionally seen as her undertaking. As Lupton (1998) explains:

The woman’s role as wife and mother is to keep the household harmonious, provide emotional stability for the family and acculturate children into appropriate norms of behaviour, including conventions of emotional management and eating habits. Mothers domesticate children, propelling them from the creature of pure instinct and uncontrolled wildness of infancy into the civility and self-regulation of adulthood. (Lupton, 1998: 39)

The centrality of nutrition and feeding to conceptions of what constitute femininity and ‘good’ motherhood, should be taken into consideration by myself in terms of the kind of questions asked and how to ask them, when dealing with issues around food in the data collection process.

Charles and Kerr (1988) measured the frequency of consumption of various food types in families, as broken down between fathers, mothers and children. The results suggest that
particular types of food were seen as more appropriate to children. These included: breakfast cereal, biscuits, puddings, sweets, soft drinks, baked beans and milk (Charles et al, 1988: 142). In light of this, food preferences will be explored not only in terms of SES, but also in terms of perceived appropriateness for age and whether or not the child has PWS.

2.4 Managing PWS

To date there have only been two social science studies of the management of PWS within the family. Van Hooren et al (2005) approached the subject from a social work perspective, exploring the values of 23 parents and 14 non-parent caregivers of children with PWS. Their findings highlighted the caregivers’ beliefs that family life had to be enjoyable and meaningful for everyone involved (although siblings did not appear in the study). Four values were identified which motivated the everyday interventions of caregivers, these were ‘physical health; well being; freedom and responsibility; and a liveable life’ (van Hooren et al, 2005: 311). Thirteen interventions were identified, these were the ways care was given to the child with PWS, these included ‘not to make high demands; to be moderate in imposing rules; to be alert; to warn in advance’ (van Hooren et al 2005: 311). Although the values were seen to guide the interventions, many of the interventions were also described as creating conditions which limited other values. For instance, the intervention of ‘correcting the person with PWS constantly’ was guided by concerns for physical health, however it created a condition of ‘constant supervision and control’ which jarred with the value of ‘freedom and responsibility’. Van Hooren et al’s findings successfully highlight the complex practical and moral demands placed upon the caregiver in the management of PWS. Acknowledgement of the role of siblings and of the person with PWS in everyday management practices could be drawn upon to give a fuller picture of the phenomena of managing PWS.

Educational psychologists Brown and James (1992) produce an overview of PWS in fifty families in Western Canada. This includes a break down of medical characteristics, a psychological summary of social development and much discussion of the families’ access to professional services. Of most relevance to this research, it includes some data
about food management strategies within the families. Findings suggest that the majority of families had used some sort of ‘calorie counting’ method or special diet. Typical everyday adaptations to food practices included: ensuring food was eaten only in the dining area; colour coding food containers; using low calorie substitutes; and enforcing table etiquette and rituals’ (Brown et al 1992: 120-121). Brown and James offer little further explanation of these practices or how they occurred throughout the families. They do however draw attention to family tensions occurring as a result of siblings’ greater access to food (Brown et al 1992: 121). Again family management is described in terms of parental intervention with little insight into the formulation and experience of these practices within families. There is an apparent need to develop an in-depth understanding of food and body management practices within the families of children with PWS, accounting for the actions, understandings and intentions of all family members.

2.5 Conclusion

This review raises many important questions and areas of enquiry for the research. Firstly a review of literatures on embodiment and their application to the subject posed the question:

As human action is mediated through the body, is the child’s behaviour different from other family members, and does this affect the order in the micro structure of the family?

And,

What work is done by family members to restore this order?

These questions highlight a need for an embodied investigation of action within the family. In particular, attention must be given to how the individual with PWS perceives their environment and actions within it.

Research on the experience of illness in childhood displays the significant contribution of siblings in families with disabled children. This points towards a need to incorporate all family members within the exploration of PWS management. Research also suggests the heightened importance of the body in childhood and adolescence, this can be examined in relation to the children with PWS and their siblings accounts of everyday life. Literature relating to the emotional impact of disability within the family identifies the need to
explore if and how family members feel stigmatised by the condition and whether this influences behaviour and management strategies.

The impact of social and structural factors, namely family structure and SES, are pronounced in sociological literatures. The high proportion of lone-parent families with disabled children implies the value of incorporation of this family type within the research, in order to present a rounded picture of management within families. Similarly SES has been seen to affect many aspects of the health experience that relate to our subject. Higher class groups are documented as taking more purposeful and confident approaches towards illness, recognising their own agency in determining health outcomes. SES differences in terms of nutritional knowledge and foods consumed are also particularly marked. Variables of family structure and SES will be purposively sampled within the research in order to build on these findings.

The emotional significance of food in family relations may affect the management of diet within the families. This will be explored with particular reference to: How parents renegotiate their role to involve the denial of food to their children, whether food takes on the same emotional connotations within the families and how families deal with special occasions. Further to this the sparse nature of systematic studies of body management within the families of those with PWS, points towards the need to develop an in-depth understanding of management practices drawing on experiences and contributions of all family members. Further literature and research from the areas identified in this review will be examined throughout the analysis, where relevant.
CHAPTER 3: METHODOLOGY

Introduction

This thesis is concerned with the experience of Prader-Willi Syndrome and the body in childhood; specifically it aims to discover how the child's body is managed within the family. The literature raised a number of questions for the research, pointing towards possible areas of interest within the main question, 'How is PWS managed within the family?' This chapter examines the research methods and analysis employed within the research and the rationale behind this.

Firstly, it provides an outline of the main aims and objectives of the research. It then outlines the research questions and areas of interest for data collection. Issues of sampling and all data collection methods are discussed in terms of issues affecting their design and reflections on carrying out the case studies. The theoretical and ethical dimensions of the methodology are explored before discussing the approach to analysis and outlining findings from the preliminary analysis. Relevant methods literatures are drawn upon throughout.

3.1 Aims and Objectives

Prader-Willi syndrome (PWS) is a chromosome disorder, currently thought to affect approximately 1:16,000 live births (Waters, 2001). Within the UK population overall it is estimated to have a prevalence of 1:45,000, based on findings in one UK health region (Whittington et al. 2001). The disorder has, among its clinical sequela an intense interest in food (hyperphagia), which may develop into an insatiable obsession, likened to addiction. Body composition is also affected by the disorder, with the muscle: fat ratio being very poor in comparison with the general population, resulting in the requirement of a lower daily calorie intake. Consequentially, unless diet and physical exercise are carefully monitored, weight gain can be very rapid, leading to obesity, disease and early death. The level of control over diet and the body required within this syndrome is very hard for the PWS person to achieve alone, since the majority of sufferers experience various forms of learning disability, developmental delay, autistic spectrum disorders and behavioural problems.
The main aim of the research was to develop a rich and in-depth understanding of body management among children with PWS and their families, exploring how and why approaches to body management are employed within the family. Particular attention was given to examining dietary control, in relation to the lifestyle and everyday routines of the child and family. Significant variations were possible, for example, in terms of the socio-economic resources of the household and the family structure (i.e. number of parents and siblings). Due to the dynamic nature of family life, an insight was also needed into how relationships with food and approach to body management vary over time, as the child grows up. The research therefore focuses on different family formations and looks at changes over time, both retrospectively and prospectively.

3.2 Initiation: learning the realities of PWS

Before any questions could be posed or research designed, it was necessary for the researcher, myself, to gain some practical experience of working with people with PWS. There are two main reasons for this concerning ‘what’ to research and ‘how’ to research it. Firstly, it was important to learn first hand how the different physical and behavioural aspects of the disorder presented themselves in real life, in addition to knowledge of the disorder gained from medical and social care literatures. This also gave an opportunity to examine the diversity within the group. Secondly, it was advantageous to get practice in communicating with people with PWS, exploring how to phrase questions to maximise comprehensibility to the group, and what behaviour and approach, in a responsible adult, the group seemed to be comforted or put at ease by.

In the summer of 2003 I worked as a volunteer on two outdoor activities weekends. The weekends were organised by PWSA (UK) to facilitate research workshops for 16-24 years olds with PWS for a separate research project on transition in young people with PWS. The groups were single sex, one male and one female. The male weekend consisted of 6 young people and 4 volunteers. The female weekend was made up of 8 young people and 3 volunteers. Over the course of the weekends we took part in activities such as, abseiling, caving, canoeing, assault courses and indoor craft activities such as painting, jigsaws etc.
Again, in the summer of 2003, I worked as an in-house (staying in residence) volunteer in a permanent respite care facility. Over a period of seven days I worked with a group of people with PWS aged between 17-38 years, approximately half of the group came from family homes and the other half residential homes. Daytime activities included shopping, walks along the beach and around the area, a visit to a children’s farm, indoor fairground and museum. Activities within the Centre were swimming, television (Wimbledon tennis tournament), games, jigsaws and a disco.

These work placements allowed me to develop my personal skills and knowledge, viewing the range and variety of medical conditions and behavioural aspects within PWS in young people and adults. The separate sex groups allowed me some insight to gender and PWS. The mixed group permitted me a chance to see how romantic relationships are negotiated within the group. All of the placements provided me with the opportunity to watch how the group interacted with each other. It became clear very quickly that despite any cognitive or language difficulty these people may experience they do not need a non-PWS person present in order to communicate with each other effectively. Recognising what they prioritised in conversations with each other gave an intriguing insight to both their experience of and relationship with the everyday social world.

Helpful as the placements were, bringing previously undetected issues to my attention and instilling myself as a researcher with the confidence and skills to interact with PWS people, there are many inconsistencies with this work and the research itself. The age ranges were different, as were locations and the ‘family’ were absent. This was not a piloting exercise, but a personal opportunity to develop skills and knowledge and become initiated to PWS.

This initial phase of the research addressed Bendelow et al’s (1999) call for an ‘embodied sociology’, where issues of embodiment are considered in every aspect of the research process and the embodiment of the practitioners as well as the subjects is taken into account (Bendelow et al 1999: 3). The call for the researcher to reflect on their own physical and social character within processes of theorisation and fieldwork
can be noted through methods literatures from the early 1990s (Scott et al 1993, Coffey 1999). In these initial interactions with people with PWS I was able gain a practical understanding of successful rapport.

Observations carried forward to research design included:

- A tendency for people with PWS to comprehend and respond in a ‘literal’ and factual manner. In line with the diagnostic criteria of ‘concrete and rigid’ social approaches, people with PWS tended to prefer talking about real events and the factual content of what happened, as opposed to why they happened or how it felt. Further to this, any use of metaphor within my own speech caused confusion. This excerpt from my first residential break with people with PWS, demonstrates this point. It also gives an insight into the difficulties I faced in adapting my behaviour and language in order to communicate better with the group:

  ‘Day 2: Really struggling to get the hang of not using metaphor/idioms. So use to using them, don’t realise I have until met with blank faces. Can remember at least five I’ve done already: face the music; back seat driver; fly on the wall; hold your horses; and pull out all the stops. This morning Penny [with PWS] was quite intrigued about ‘holding her horses’, thought it would be better to try to explain why I had said it, what I meant by it. She didn’t seem to understand even after I’d explained though, so must just stop doing it.

- Concepts of time also caused confusion for many of the people with PWS. There was some difficulty around explaining how often events occurred or conceptualising terms such as ‘recent’. This was observed frequently in work placements and noted in my field diary:

  ‘Am sure one of the reasons I keep getting asked [by people with PWS] about when activities etc are going to happen is that they find it hard to visualise concepts of time. Jacob [with PWS] was telling me about his book but couldn’t tell me how long he’d been reading it for, or even how long it took
him to read a chapter. This came up before when Chloe [with PWS] was talking about her holiday and couldn't remember how long ago it had been.’

•Pleasure and comfort experienced by people with PWS by knowing about future events and their order emerged very strongly. Individuals appeared more relaxed and able to converse once they were familiar with the order of the day’s events and knew what they could expect. Contradicting information regarding future events or changes to practices and schedules caused anxiety within the group. On all the respite trips I worked on it was very important to plan ahead, with all staff knowing the schedule for the day’s activities. People with PWS tended to ask for confirmation of events extremely frequently often searching for more accuracy e.g. exactly what time, instead of just this afternoon. This excerpt from my field diary on an activities weekend with fifteen young people with PWS displays this:

‘From now on I’m just going to say [to the people with PWS] the sequence of what we’re doing rather than estimate times, unless I def [definitely] know for sure that something will be at a certain time. Also try to be more vague with times- ‘between 4 and 5’ instead of ‘at 4.30’. Today we were late back from abseiling, most didn’t notice because asleep in the minibus but Leanne [person with PWS] pointed it out to Bella [social worker] on the bus. B diffused situation before anyone else picked up on it’.

One change in events that caused a huge amount of anxiety and several temper tantrums occurred on the fifth day of a seven-day respite break. The room that we had been using as the dining room was booked as the venue for a meeting by a local company. The staff of the respite facility were not aware of the meeting until a few hours before it started. As the meeting was only meant to last three hours, whilst we were all on an outing it was presumed that this wouldn’t cause any inconvenience. However, because the meeting overran by an hour, it was still in session when we returned. The people with PWS were denied entry to make a hot drink [which had become a routine associated with
returning to the centre after trips] and couldn’t be given an exact time when they would be allowed in.

‘[On returning to the centre and hearing the news about the meeting] Callum [social worker] decided the best thing to do was to ask if one of the staff could go in and make the drinks for the group. As he made the drinks, we all [other staff] tried to keep as cheerful as possible; talking about the fairground rides [that afternoon’s activity] and trying to involve everyone in T.V. or jigsaws. This was quite a pointless effort really, as they were already noticeably extremely wound-up and wanted to know what these ‘other’ people were doing in ‘their’ dining room, with ‘their’ tea, coffee, milk and kettle [Many of the young people on the trip preferred to do activities in the dining room rather than the lounge and recreation rooms, in particular making friends with the chef]. Demands for information came thick and fast, voices were raised—seemed to spread like mass hysteria. Lily [with PWS, having a hard time anyway as she was adamant that the only activity she had any interest in was shopping. Was refusing to join in with other activities] screamed and ran out and had to be followed by Paulette [social worker]. This shocked people into calming down a bit. We all agreed it would probably only be half an hour and that we could be patient. Lily’s tantrum involved screaming, crying and abusive language, lasting for about 10 mins [minutes] then 20 mins of crying. Paulette and Zeena [social workers and volunteer] gave her a bath and calmed her down. Meanwhile, the calm in the rest of the group didn’t last very long and despite our requests the majority of the young people refused to stay in the recreation rooms. Instead they congregated around the door of the dining/meeting room. As we tried to keep the noise level down, voices from the group asked ‘What if they don’t come out before dinner?’ , ‘Who are they?’ and ‘Can’t you tell them to use another room?’ . Consensus amongst the group was that this was not fair and that these people had no right to be there. Harry [person with PWS] tried to reassure his friends saying that it would be okay and they were bound to come out soon. This didn’t work and just raised all the same angry points again. I’d stopped trying to explain the predicament at this point for the same reason. I kept silent and just tried to appear as calm as
possible, remaining close to all of them so that they couldn't storm in! We then heard a loud banging noise on the lounge window, Danny [with PWS] being extremely stressed and upset by the situation had gone outside and was banging the back of his head on the window.'

Despite witnessing temper tantrums on several occasions during my voluntary work placements, I always found, self-harming behaviour in particular, extremely disturbing. By the end of the episode mentioned above, I too thought the situation was unfair. Although I did not express my feelings to any of the young people with PWS, along with other volunteers and workers, I was angry and felt that the situation could have been easily avoided.

• Positive reassurance was often sought by people with PWS from staff and volunteers. Gaining this reassurance seemed to allow individuals to carry out activities at ease and take greater pleasure in them. As this excerpt from my field diary, about abseiling with young people with PWS displays:

'Abseiling was amazing. Kath [volunteer] went down first to put everyone at ease a bit. You could tell no one thought they could actually do it and they looked pretty scared. In the end even the ones who had been convinced there was no way they could go over the top managed to. It took forever! They seemed to just need persistent and absolute assurance and reassurance that they could do and they were doing it right. By the end we'd attracted quite a crowd of passers-by who were joining in, shouting encouragement, clapping etc. Everyone's still on a high, seem totally proud of themselves and keep asking me if I saw them!'

• When closed questions were asked there was a tendency for people with PWS to confirm, although on many occasions it transpired that the information they were confirming had not been understood. This tendency came up repeatedly in work placements and I discussed it with one of the PWS specialist social workers:
'Spoke to Laura [social worker] about people saying 'yes' when they haven’t understood. She was saying it’s very common and goes even further than that, sometime people have been known to actually rephrase things as if they understand, but when it comes to it they haven’t. She mentioned giving scenario cases to people in her work and getting the correct answer before assuming that people had understood. People don’t tend to feel patronised by this; in her experience they quite enjoy it. Idea for informed consent and other questions.'

These aspects of communication had initial implications for my conduct within the research, in terms of content of questions and use of language. Firstly, to get a full response there was a necessity to include many factual questions before seeking to probe explanatory or emotional dimensions of everyday actions and events. Use of metaphor was avoided throughout the data collection interactions. Questions avoided the use of abstract concepts of time e.g. ‘how often? How many times a week/month/year?’ Instead questions were phrased in terms of the sequence of events e.g. ‘Did this event occur before/after that event?’ Before all data collection sessions with people with PWS, the format of the session and what could be expected was explained fully. Positive reassurance was given throughout sessions and closed questions were avoided where possible. Informed consent with children with PWS involved not just telling them their rights but asking them what they would do if they got angry or felt upset during the interviews. As I didn’t want to introduce a negative image of the data collection, I also asked what they’d do in other circumstances e.g. ‘if you can’t stop laughing’, ‘if the cat jumps on your lap’, ‘if your friend knocks on the door’.

As the work placements involved people with PWS ranging from sixteen to thirty-eight, outside of the family home, the data was not used to address the main research questions in this thesis. Data, in the form of the field diary was collected with the specific function of preparing me for working with people with PWS in an effective and ethical fashion. Analysis of the diary for this purpose consisted of open coding, I highlighted observations I thought may be relevant as I went along and wrote memos concerning the links between these observations and the research process.
3.3 Research Methods

Research Questions
The two key questions posed by the research are initially descriptive then explanatory in nature:

How are the bodies of children with PWS managed within the family? And Why are the apparent management strategies employed?

The research addresses these questions using a predominantly qualitative methodology. Thus, it is the data collected that generates the theory and discussion within the thesis. At the stage of drawing up the research questions, it was crucial not to place limits on the findings by being over specific, or making assumptions about the processes involved. However, in order to devise a comprehensive research design and anticipate problems both practical and ethical in nature, some extension of the key questions was required.

These key questions and the literature review opened up a number of subsequent questions and areas for further inquiry. When looking at how the PWS body is managed within the family, a number of areas need to be addressed. Firstly, how is the body ‘physically’ managed? With reference to diet, the research aims to uncover objective factual information on, what types of food are consumed by the child with PWS; what amount of food is consumed; at what times meals are eaten and whether this is a strict routine; and to what extent eating practices of the PWS child vary from the rest of the family. The research will also aim to disclose any non-food strategies that affect the physical body. For example, how much calorie burning activity is built into the everyday lifestyles and routines of the children and whether they have home activities which act to distract attention away from food. Secondly, what beliefs and attitudes are held by children, and their families, towards health, food, exercise and the body? As chapter two displayed, these understandings and beliefs have been seen to differ in light of age and SES. Questions of how these understandings impact management strategies and practices are assessed.
In order to measure the input of the PWS child within these processes, it was necessary to pose research questions and data collection techniques that isolated the child’s perceptions and experiences analytically. To ascertain the impact of somatic factors on the child and in turn the family, there was some exploration of embodied experience within PWS. That is, how sensory aspects such as hunger, pain and emotional dispositions were negotiated within the family environment. In order to get a clear idea of how the children relate to and understand their bodies, perceptions of health were explored.

Unit of analysis
Due to the pronounced need for the child with PWS to be externally monitored and controlled in terms of food intake, the unit of study is the child within the family context. Any study into how the PWS child body is managed and how the child relates to the social world must acknowledge the crucial role the family play, in terms of diet control and their influence over the social environment that the child inhabits.

Each family unit is uniquely linked to its own biological variation of PWS (i.e. level of language skill, cognitive ability, medical treatment choices, related medical problems), structure, resources, location, hierarchy and beliefs. Placing the child within the family context allows these factors and their effects to be fully recognised. Comparison of these units has the potential both to correlate body management approaches with isolated factors and to draw out trends that have a high prevalence amongst the families of children with PWS regardless of these factors.

Sample
The sample was drawn from members of the British Prader-Willi syndrome Association (PWSA (UK)). The sample was comprised of a total of twenty families. Children were aged between eleven and fifteen years at the beginning of the study. Due to the effects of developmental delay on cognition and communication skills within PWS, eleven was thought to be the youngest age appropriate for children to participate in research. The upper limit of 15 years would imply that the sample spanned childhood and adolescence within the normal population. However, within PWS puberty is normally delayed in both males and females, although some children
in the sample may have experienced some early traits of pubescence it is much less pronounced than in the general population (Waters 2001).

Given that the management of PWS was likely to vary in different family formations and household structures, a diversity of families were recruited into the sample. The three main discriminating factors in drawing the sample were:

1. Child living with one or both parents
2. Only child or child with siblings

(See figure 3.1)

There are many more factors that could potentially affect management approaches e.g. geographic location, religion, gender of child with PWS, ethnicity etc. The three factors selected were anticipated to have the most direct effects. As the unit of analysis was the child within the family, and the research involved exploring familial interaction, factors 1 and 2 presented as primary variables for the research sample. Further to this the literature review raised the importance of lone-parent families the care of disabled children in UK society, as well as the care work performed by siblings within the household. Factor 3 was privileged as it allows for patterns in management to be revealed along gradients of material and cultural circumstance. The significance and overriding agency of which has been proven consistently within the sociology of health and illness, medical sociology, public funded health research and epidemiology (Acheson 1998; Scambler 2001; Wilkinson 1986).

The most meaningful method of operationalising SES within health research is an ongoing discussion within sociological literatures (Nettleton 1998: 167). Inequalities of mortality and morbidity in relation to SES have most commonly been measured against class as defined by Registrar-General’s occupational social class (Blane 1991, White et al 1993). One problem of defining class by occupation within this research is the inclusion of lone-parent full-time carers within the sample. Although UK work practices have gone through a process of dramatic restructuring over the last hundred
years (Scamber 2002: 89), the percentage of lone parents engaged in employment is below 40% (ONS 2000a: 68). Thus the likelihood of occupation reflecting class culture of lone-parents with care responsibility for children with PWS is low. Nettleton (1998: 169) notes that issues of defining and measuring class within research are strongly influenced by methods of analysis and theoretical conceptualisations of class within findings. As the literature review highlighted the importance of class culture and lifestyle in determining health beliefs and behaviours, occupation of parent would seem an adequate measure. However due to the problem of inclusion of lone-parents, ‘highest educational qualification within the household’ and ‘previous occupation’ were added to defining categories. In the case of lone-parents these variables allow some insight into social status previous to becoming a carer.

Using the sampling frame of members of the PWSA (UK), information on SES was not apparent prior to drawing the sample. The only indicator towards SES was the geographic area in which the families were based. Using the index of multiple deprivation (Office of National Statistics 2000b, source: http://www.statistics.gov.uk/StatBase/Product); a ranking of English geographical wards in relation to home ownership, crime rate, income, health and employment, families were selected on the basis that their geographic location scored particularly high or low on this index. This was seen as the best indicator before information about occupation and education could be ascertained. The geographic locations of half of the sample (ten families) scored below the English national average on all variables within the index and the other half scored above the average. Once the sample had been drawn factors of education and employment/previous employment were checked in order to confirm the individual families’ place within the group allocated.

As a funder of this research, the national charity for PWS (PWSA (UK)) allowed me access to many of their information resources throughout the research. The PWSA membership database was used as the sampling frame, this included the names of parents, addresses and ages of children within families of people with PWS in the UK. A recruitment pamphlet (appendix 2) and information request form (appendix 3) were sent out to all families with a child with PWS in the specified age range of eleven to
fifteen years, fifty-three families in total. Thirty-two families responded and after selecting families on the basis of SES and family structure, letters were sent out to initiate the first data collection session (appendix 4). All twenty families responded and participated fully in the research.

Working as an associate of PWSA (UK) was advantageous in terms of both accessing opportunities for the development of personal skills and knowledge, accessing families and developing trust and the credibility the research. All correspondence between to the families included the corporate logos of the University of Leicester, the Economic and Social Research Council (ESRC) and PWSA (UK) (See appendices 2, 3 and 4). Although associations with the University of Leicester and the ESRC undoubtedly went some way to affording the research credibility, the association with PWSA (UK) could be seen as more meaningful to families of children with PWS. As all families were members of PWSA (UK), they had prior experience of the work of the organisation, their decision to become members indicating that this experience was positive.

In total there were eighty respondents in the research: thirty-four parents (fourteen male and twenty female), twenty children with PWS (twelve male and eight female) and twenty-six siblings, (eleven male and fifteen female).
Figure 3.1 - Sample structure

Characteristics

Total sample

Socio-economic status of area

1. Above national average
2. Below national average

Family structure - parental

1. Lone parent household
2. Two-parent household

Family structure - children

1. Only child
2. Child with siblings

Number of Respondents

20

1. 10
2. 10

1. 3
2. 7
1. 3
2. 7

1. 1
2. 2
1. 2
2. 5
1. 1
2. 2
1. 2
2. 5
**Data collection**

The primary method of data collection was qualitative family case studies. Since the intention was to focus on the child in relation to the family, each member of the immediate household was included in the research. The application of the case study method to this research was advantageous in light of the unit of analysis, theoretical approach, and the research questions posed. As Yin (2003: 2) notes, case studies have been traditionally best placed to answer ‘how’ and ‘why’ questions, such as the key research questions outlined in this thesis. The need for case studies arises out of a desire to understand complex social phenomenon, involving relations between agents. As such, case studies are frequently used in social work research (Gilgun 1994). The flexibility of the method allows a range of data collection methods to be employed, appropriated to the particular respondent and research question or theme the data are being gathered on. The individuals within the family units are extremely diverse in terms of age and abilities, making the case study method invaluable to this research on a practical level.

The case studies combine the following methods: initial questionnaires for parents; semi-structured interviews with parents and older siblings; themed write and draw/discussion sessions with children with PWS; semi-structured interview and write and draw sessions with younger siblings and observation. All interviews and discussion take place in the family home with observation work sometimes occurring outside of the home. The data were collected through two to three contacts with each family, over a year period.

**Initial Questionnaires**

Although the nature of most of the data collected in the research are subjective and the methods predominantly qualitative, some areas of interest were straightforwardly factual. For instance, if the child was undertaking Growth Hormone treatment (hGH) this would affect the composition of the body and may be of significance to the research, yet in-depth interviewing techniques are not necessary to bring out this kind of information. The surveys covered details of family members including SES (age, occupation, previous occupation, education), medical treatments and aspects of
medical history, type of school attended by PWS child i.e. mainstream or special needs and age of child at point of diagnosis (see figure 3.2). The questionnaire was administered verbally, responses were recorded as well as written to cover where respondents elaborated on the questions.

**Figure 3.2 Initial Questionnaire**

1. Age and gender of all family members currently living in household.

2. Occupations of parents or occupation previous to full-time caring.

3. Highest level of education within household.

4. Age of child when first diagnosed with PWS.

5. Any medical problems or major surgery.

6. Medications currently taken by child.

7. Type of school attended.

**Semi-structured interviews**

*With parents*

These interviews were designed to draw out data relating to questions on how PWS was managed within the families. The first interview focused specifically on managing food within the family and the second on family life more generally (See figure 3.3). The interview questions were designed to cover areas brought out of the literature review e.g. the significance of food within the family, dealing with special occasions, the work of siblings etc. In many cases these questions were not asked directly with the intention of not generating bias results. For instance, asking what sorts of activities and pastimes the family enjoy doing as a group, rather than asking directly how much they value physical exercise or whether food is significant within the family.
As the main objective was to gain an in-depth understanding of everyday food management, how parents prioritised the issues they spoke about was of importance. For this reason the initial questions in both interview schedules were very open, allowing parents to speak at length about aspects of food management and family life that they found important and introduce their own areas of interest. In most cases this was successful and further questions were used only when needed.

All interviewees were steered towards describing events which had actually happened rather than talking in abstraction about approaches to family life or food management e.g. 'Where did you go on holiday last and what was it like?', 'Can you tell me about one particular occasion of conflict within the household that you remember?' The use of specifics as opposed to generalities within qualitative interviewing is a useful way of ensuring validity in responses (Mason 2002: 227). For instance, instead of asking parents about their views or values in abstraction, these can be drawn out through analysis of how they perceived and dealt with real events. The same technique was used in the interviews with children, picking up on issues they discussed and asking them to describe past events, how they acted and how they felt.

**Figure 3.3 Parent interview schedules**

A. Food

1. How do you approach everyday diet and food issues in the home?

2. On an average school day what is eaten by child, when, where and with who?

3. Does this differ at weekend or in school holidays?

4. Are there any exceptions regarding food such as special occasions?

5. Has child ever displayed food seeking/stealing behaviour?

6. What have been situations of conflict around food? How were they resolved or how do you avoid?
7. What do you find the hardest part of managing the dietary issues?

B. Family Life
1. What are the family’s main interests or what do you enjoy doing as a group?

2. What was your last family holiday? How was it?

3. How does child get on with brothers and sisters (where applicable)?

4. How do you feel they are affected either negatively or positively by having brother/sister with PWS?

5. How do you and partner (where applicable) share childcare/domestic responsibilities?

6. What are your main concerns for child with PWS, anything particular you are working towards at the moment?

With older siblings (13 years and above, N=10)

Inteviews were conducted with siblings of the child with PWS, they addressed the topic of family life generally, aiming to generate data on how this was viewed and experienced by siblings of those with PWS. The interviews also sought to identify any input that the siblings have had on everyday management strategies. These interviews were unstructured apart from their general topic and aim. Interview dialogue built upon information given by parents in semi-structured interviews. Figure 3.4 displays basic questions used as prompts where necessary.

**Figure 3.4 Sibling interview schedule**

Family life
1. Tell me about the last thing you did as a family that you really enjoyed/didn't enjoy.
2. Do you have friends visiting very often, what do you do?

3. Tell me about your mum, dad, brother/sister.

Similarly to the interviews with parents, where possible only the first question was asked directly with conversation being steered around the other topics. This unstructured style of interviewing allowed the respondent to offer new areas of relevance to the research questions. Food was not spoken about directly with this group, enabling them to raise the topic in relation to their sibling with PWS or family life in general if they felt it was of relevance. The length of all interviews varied (between 15 minutes and 2 and a half hours) with parental sessions tending to be much longer than child sessions.

**Themed sessions with child with PWS**

A large part of the data collection process involved the child with PWS. Different themed sessions involving drawing and discussion were carried out during the research. Sessions included, ‘Getting to know you’ exploring self-perception, identity, personality, hobbies and family; ‘Food and eating’ examining their perception of how they deal with food and bodily sensations such as hunger; and ‘Health’ drawing out their perceptions of what constitutes good and bad health, thus gaining a greater understanding of how the children conceptualise the body and its functions.

The children in the study had differing communication skills in terms of language, comprehension and modes of expression e.g. writing, drawing, and speech. The sessions were tailored towards the strengths and needs of the particular child. Children were asked to express their thoughts using ‘write and draw’ and ‘draw and label’ techniques which have been found to be a very useful way of encouraging children to describe and explore their experiences (Pridmore *et al* 1995). Write and draw methods were used in conjunction with unstructured, themed interviews. That is, ‘adult’ research methods were not replaced by child-centred ones, rather children were given the opportunity to express their ideas through drawing in addition to speech. Backett-Milburn *et al* (1999) and Punch (2002) warn of the dangers of stereotyping and homogenising childhood, by assuming child-centred data collection
At the stage of research design it was thought that drawing may be particularly useful in children with expressive language problems; that pictures may aid communication by revealing a level of emotion not apparent in verbal communication alone, which tends to be more factually driven. Pictures did provide useful talking points, however they did not overcome the factually driven emphasis of children with PWS. In many cases they acted to highlight this linear, fact-based perception of the children e.g. where children chose to depict events in text rather than illustration (see figure 4.5 chapter four) and where pictures of favourite activities were depicted in an abstract fashion. For example one child with PWS (Gemma) described their favourite activity as playing with their dog, the illustration of this (figure 3.5) did not portray either the child or the dog but a set of different coloured balls used, showing how many were a particular colour.
Keeping the sessions on one theme was a deliberate and central aspect of design for these sessions. This strategy aimed to reduce anxiety and behavioural challenges in the child with PWS. Amongst other characteristics, Waters (2001) lists, ‘temper outbursts, similar to those of a two-year-old child, stubbornness, inability or reluctance to adapt change in routine, obsessiveness and perseveration (repetition of the same question, or reverting to the same subject over and over again)’ as the main behavioural challenges of PWS (Waters 2001). Keeping to a relatively narrow topic acts to avoid overloading the respondent with demands and information and allows for the narrow, focused, concrete conceptualisations that many people with PWS find it comfortable to work within.

These themed workshops, conducted with children with PWS, were used to explore the child’s understanding of day-to-day management practices. The ‘Getting to know you’ session was conducted on the first visit. As well as gaining data on lifestyle, interests and personality, this session allowed the child’s communication style to be shown and future sessions to be tailored accordingly. For instance, many children were extremely vocal and seemed to enjoy giving narratives of events in their life. In these cases keeping the child’s focus on the theme of the session was the main priority within the sessions. Other children gave minimal answers and appeared to be shyer within the workshops. With these children the priority became finding a subject that interested them and that they were happy talking about in order to get the session moving. Sessions on ‘Food and eating’ and ‘Health’ were conducted on subsequent visits to the families. Children’s beliefs and ideas on these topics were linked to their everyday experiences. That is, when expressing an idea about food or health, children were asked how and when they put this into practice.

As explained in the ‘ethics’ section later in this chapter, six families decided a ‘food and eating’ workshop would be problematic for their child with PWS and these sessions were not carried out. Due to the nature of the data collected from the ‘food and eating’ workshops, it was decided that a ‘food and eating’ workshop would be problematic for these families. Therefore, these sessions were not carried out.

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1This information after data excerpts indicates whether the family is in the higher or lower SES sample 'SESH'=higher 'SES L'=lower, how many parents within the household 'P2'=two parents and 'P1'=one-parent and whether there are other children within the household 'NOSIBS'=only child 'SIBS'=child with siblings.
and eating’ workshops that were carried out, this restriction is not judged to have affected the overall analysis in a major way. As described in more detail in chapter four, although children could list many foods and talk about when they ate them, they had problems describing sensations of hunger and describing why they liked particular foods.

The generally limited nature of discussions with children with PWS, however, did have implications for the analysis overall. It is noticeable in the following analysis chapters that parental quotes are longer, more detailed and used more often. This reflects the embodied nature of the research process; people without PWS communicated in a way that was comprehensive and meaningful to myself, the researcher, and thus are dominant in the analysis. Despite attempts to redress this balance, I found no way of either getting children with PWS to offer more detailed accounts or finding meaning within the accounts that they gave.

Sessions with younger siblings (under 13 years, N=16)

Siblings under ten years took part in the same themed sessions as their brothers/sisters with PWS. As well as aiming to understand these children’s perspective and contribution to family life and management strategies, data from these sessions acted as a control group throughout analysis. That is, a way of gauging whether aspects of data were specific to PWS or common amongst children more generally.

Observation

At least one participant observation session was carried out within each family; where possible this took the form of a family meal. This technique allowed family relations and food practices to be viewed from an objective position. The family meal was of particular interest as information about the type of food consumed and its presentation could be gained first hand. Observation of kitchen spaces, their layout in terms of how and where food was stored and use of locks was also carried out on the initial visit to each family home. A great deal of observation work carried out was informal, as notes were taken on behaviours and interactions of family members in between interview sessions.
Observation of family meals posed particular methodological considerations regarding how my presence might disrupt the behaviour and interactions between respondents. The disruptive presence of participant observers is well documented within methodological literatures (Wilmot 1979, Mack et al 2005), as such I was able to take precautions to limit this disturbance. The rationale behind the technique was explained to the families before the observation was conducted. Families were encouraged to interact with one another as they normally would instead to conversing with me, as their guest, and informed that I would be taking some notes. After the meal parents were asked whether they perceived my presence to have influenced anyone’s behaviour and in what ways.

In these observations and in general during the research, parents and siblings commented that their children with PWS had been on ‘best behaviour’ while I was with them. Many of them also pointed out how much the children enjoyed the idea of having their own researcher and receiving special attention. In light of these comments the importance of complementing the family case studies with work placements became very apparent. Within the work placements I was viewed as a ‘helper’ by people with PWS rather than a ‘special researcher’. Temper tantrums, disputes and other challenging behaviour was experienced on a daily basis, allowing me to get a full picture of people with PWS. This allowed my analysis to form from a more informed base. Although the data from respite breaks were not used as data for this research, my experience allowed me a greater understanding of problem behaviour as explained by family members.

### 3.4 Ethnographic and Ethical Approach

All the data drawn upon in the analysis were collected within the family homes. An ethnographic approach was taken, as each case study aimed to discover the specific practices, behaviours and routines unique to that family and the meanings behind that everyday action. As such the structure of the data collection sessions in terms of their timing, location of interviews within the home and whether parents and siblings were interviewed in generational groups or individually, was not imposed strictly by the researcher. This relaxed approach allowed a more detailed insight into the structure, hierarchy and responsibility for care work within the families.
For instance, in some cases mothers arranged the research interviews to be carried out at times when the father was at work. Although the fathers' participation in research was expected, in three cases this took the form of a brief interview due to the time constraints of employed work. In three cases an older (fifteen years or above), sibling joined in the parental interview about food management strategies, displaying their significant role and felt responsibility in the everyday management of their sibling. Multiple siblings were asked whether they wanted to conduct their sessions as a group or individually, with most opting for group work. Sessions with the child with PWS were conducted individually, however they often called a parent into the room in order to help them explain a particular event or to show a picture to them. All such instances formed valuable data themselves for the analysis.

The importance of researcher reflexivity is frequently highlighted in relation to research with children (Punch 2002, Backett-Milburn et al 1999), children with disabilities (Davis et al 2002, Alderson et al 1996) and ethnographic approaches more generally (Edwards et al 2002, Hertz 1997). The researcher’s ability to analyse their own thoughts, questions and interpretations throughout the research process has been seen to increase the validity of research findings and greatly enhance ethical practice. Allowing the respondents freedom to structure aspects of the data collection process enabled me the detachment necessary for this reflection. For instance, problems I had anticipated around parental intrusion on interview sessions with children were not played out, rather the children actively asked for their parent’s contribution, demonstrating the intensity of this relationship within the family.

As part of this reflexive approach I kept a personal logbook. This detailed how I had felt within various situations in the fieldwork setting; decisions I had made about content of interviews and why I had made them, as well as recording initial theoretical and analytical ideas. The journal revealed it was not only with the children with PWS and their siblings which I exercised caution and anticipated the response to my actions and requests. Topics of parenthood, specifically motherhood in relation to food, as suggested by Lupton (1998), are imbued with emotive and moral connotations. In many interview situations with parents I noted strong feelings of personal pride,
accomplishment and a sense that the parent's identity was closely linked to how they dealt with these everyday issues of food within the home. I noted that in many cases I often felt compelled to state an agreement with parents’ views and approaches to health and lifestyle and commenting positively about the effectiveness of their strategies, as not to do so would have felt impolite.

Ethics

Ethical considerations for the research formed a part of the research design prior to fieldwork as well as being actively considered and implemented throughout. Edwards and Mauthner (2002) outline an 'ethics of care' approach to research practice in contrast with a more traditional 'ethics of justice' approach. Justice-based models of ethics depict the researcher as an autonomous, rational being. This approach is concerned with drawing up and adhering to a set of guidelines concerning what practices are right or wrong, these guidelines, having the capacity to be applied universally to research contexts. Ethics of care, in contrast, acknowledges the researcher as a 'self-in-relation' to other, such as research respondents. Ethics of care is concerned with the subjectivity of morality within different research contexts, maintaining that rather than simply following set guidelines ethics must be carefully considered in light of the complex relations within each research setting (Edwards et al 2002: 22-23).

Although many ethical interventions were initially drawn out of consideration of the British Sociological Association (BSA) Statement of Ethical Practice (1994), ethics of care had an overriding application to the nature of this research and informed the research process throughout. Thus prior to the research ethical interventions were planned in light of the BSA guidelines. For example, BSA guideline 1.6 notes,

In some situations access to a research setting is gained via a 'gatekeeper'. In these situations members should adhere to the principle of obtaining informed consent directly from the research participants to whom access is required, while at the same time taking account of the gatekeepers' interest. Since the relationship between the research participant and the gatekeeper may continue long after the sociologist has left the research setting, care should be taken not to disturb that relationship unduly (BSA 1994).
Access to families had been negotiated via the PWSA (UK) membership database, parents of the families were sent publicity materials on the basis of which they volunteered their families for the research (see appendices 2, 3 and 4). However, before data were collected from family members, each was asked individually for their consent, reminded that they could withdraw at any time and encouraged to mention any aspect of the research that they found uncomfortable or had questions about, for informed consent form see appendix 5. In addition to the assurances of confidentiality and right to opt out of the research, included in the recruitment correspondences (appended 3), these issues were the first topics covered in the initial face-to-face meetings with families. Families were informed of their names appearing as pseudonyms in the write up of the research and any associated publications. Issues of data protection in terms of how their personal details would be dealt with was also covered i.e. would not be accessible to any other party and shredded when no longer needed. As the data collection progressed many families would ask me directly for information about other families management practices and the character and behaviour of other children with PWS in comparison to their own. In light of this, all families were asked on subsequent visits whether they minded this sort of information being communicated to other families. All families agreed to this, perhaps reflecting that their appreciation of the benefits of pooling information and sharing experience was a key motivation for being involved in the research.

Informed consent forms were explained and filled out by all family members as part of the initial visit. A key finding of the sensitising work placements was that young people with PWS have a tendency to agree to statements, often without fully comprehending the meaning of the statement. As noted on page 65, the relevance of this to the process of successfully gaining informed consent was immediately apparent. In light of this, the process of gaining the informed consent of children with PWS involved giving the children scenarios, asking them what they would do if they got angry or felt upset during the sessions. Bearing in mind another finding of the sensitising work, that people with PWS had a tendency to take information literally, I was concerned that the respondents would not appreciate the hypothetical nature of this scenario and that they would be expecting to be upset at some point in the sessions. As I didn’t want to introduce a negative image of the data collection, I also
asked what they’d do in other circumstances e.g. ‘if you can’t stop laughing’, ‘if the
cat/dog jumps on your lap’, ‘if your friend knocks on the door’.

The complexities of gaining informed consent have been widely raised in connection
to research with people with learning disability (Stalker 1998, Moore et al 1998,
contradiction between the emphasis on conducting research with instead of on people
with learning disabilities, seen in the participatory action research approach, and the
legal requirements of gaining consent from parents, guardians and carers. As the
respondents with PWS in this research were also aged below 16, there was an even
greater emphasis on the correct involvement of parents and guardians. Similarly to
Thomas (2003), I felt particular aspects of the research involved an ethical tension
between devising an approach which took on board the concerns of parents, whilst
allowing the person with PWS freedom to express themselves fully.

As I explain in more detail in this section, a large part of the initial session with
families consisted of detailed discussions with parents about what subjects may be
problematic to cover with their children. Effectively this meant parents perceptions an
experiences with their children shaped the data collected in sessions with children
with PWS. This problem highlights the embodied nature of research in practice.
Parents were able to express and articulate what topics may cause distress to their
children in a manner that I understood, without becoming angry themselves. Although
I would have preferred to ask the people with PWS directly, thus treat them equally in
the research process, I felt that this may simply invite the opportunity for them to get
stuck on a negative topic, that may cause them to become upset or agitated. This
methodological and ethical dilemma indicates the difficulties involved in negotiating
multiple perspectives of respondents within families (Greenstein, 2006).

It has been noted that interventions and innovations around informing respondents of
their rights are of particular significance in research with children (Alderson 2004,
Mayall 2000). Although differences in researching children and adults should not be
overstated to the point of portraying children as a separate and homogeneous group,
issues of societal power relations between adults and children and how this affects the
child must be taken into account. That is, children may be more used to following
adult instructions and thus less aware of their rights and choices where the research
was concerned. For this reason children were reminded of these rights before every
data collection session. Interventions based on BSA guidelines proved to be
successful measures, as children utilised these rights, opting out of answering certain
questions and enquiring about aspects of the research process such as what would
happen to their tape recordings and drawings.

However, these interventions formed a very small part of the ethical work within the
research overall. Before any data collection sessions were carried out with the
children with PWS, the nature of the sessions were discussed with one or both parents
at length. Due to the sudden emotional outbursts and behavioural problems
experienced within the disorder, I sought parental opinions of whether the children
would enjoy the sessions, what problems they perceived and any possible negative
effects on the child or family I had overlooked. In many cases parents saw no problem
with the unstructured discussions and drawing work around identity, food and health.
However, six of the twenty families expressed a wish that I didn’t cover the topic of
food. They described that the child gets stuck on this topic, causing prolonged anxiety
and possibly problematic questioning and behaviour once I had left. Other families
listed a range of topics that should be avoided, subjects to which the child was
particularly sensitive at the moment. These topics included toys, transport to school,
school friends and upcoming events such as holidays, trips or medical procedures in
the children’s lives. Three families asked me not to mention Prader-Willi syndrome
explicitly, as the child was not aware of the medical condition. Many other parents
commented that the child knows about PWS, but does not fully understand what it
means.

So between case studies there was a huge variation in what constituted an ethical
approach to practice in this research. As seen in ethics of care literatures there was no
standard for ethic practice, rather it was negotiated through parental interpretations of
their children and my interpretation of the families and individuals. This resulted in a
highly reflexive research practice with the children with PWS, as I constantly
monitored the impact of my choice of topics and actions both with the children themselves and other family members.

3.5 Analysis

Interview scripts, observational notes, journal entries and children's illustrations formed the data for this research. Qualitative coding-based analysis was carried out aided by QSR NUD*IST software. The insights and techniques of Glaser and Strauss's (1967), Glaser (1978), Strauss (1987) and Strauss and Corbin (1990) were drawn upon to guide analysis practices. As empirical research, with a central research question ('How is PWS managed with the family?') firmly based in everyday action and interaction, it was important to adopt an analysis where theory was generated in close relation to empirical data. A principle concern of Glaser and Strauss's (1967) The Discovery of Grounded Theory was that analysis should involve the act of moving between data and theory until they reach a point of synthesis. It is their ideas that were used to guide this analysis.

The 'Constant comparative method' (Glaser et al 1967) was utilised throughout all levels of coding. That is, as data were coded they were compared to other data coded in that group, where differences were found new categories and codes were created. In the initial 'open' coding, following guidelines of Glaser (1978) and Strauss (1987), simple questions were asked of the data i.e. is it displaying what, why, how, where etc. in relation to the main research question. This allowed the first division between the data into 'how' PWS is managed within the family and 'why' it is managed in this way. From this point 'axial' coding was employed within each of these branches, interrogating which category was related to another and how these categories could be ordered and combined to produce key theoretical concepts.

It is interesting to note that the majority of open coding, and notes on possible directions for axial coding were conducted long before QSR-NUD*IST software was employed. Even before interviews had been transcribed, the long train journeys back from family visits, when impressions of the family were still fresh in my mind, provided some of the most fruitful times to reflect upon the direction of analysis. Similarly, once case studies were transcribed, much of the open coding was marked.
on the paper copies before being transferred to QSR-NUD*IST. Although this may have been more time consuming as it involved repeating the same coding on paper and then transferring it to QSR NUD*IST, it seemed necessary as the analysis process started as soon as I left the first family visit and my thoughts needed to be recorded.

Within the 'how' branch, four features of the practices relating to management of the PWS child body were identified. These features form the subject of chapter four. The central category of the 'why' branch was 'embodied interactions' within this there were sub categories of biological/sensory, emotionality, family values, existing knowledge and materials. That is, all actions directing the management of PWS within the family were seen to derive from embodied interactions between family members, influencing factors came from various sources but were bought together in processes governed by these individuals. Chapter four lays out this analysis in more detail, examining these embodied interactions, the role of the biological and the use of materials in addition to describing the four features of the management of PWS. Chapter five concentrates on how epistemological features such as family values and knowledge existing outside of the family are drawn into this process. It outlines the identification of four family values and their sub categories.

The social factors structuring the sample i.e. family structure and SES were included within the analysis as 'base information'. Using QSR NUD*IST software all data were attached to codes which indicated whether the family was part of the higher or lower SE group, a lone or two-parent family and a child with siblings or an only child. As with other codes attached to data, it was possible to search for correlations between this 'base information' and the concepts identified. Strauss (1987: 32) was keen to draw attention to the point that the significance of categories should not be assumed or hypothethsised and the inclusion of variables, such as our 'base information', should only be included within the theory if they demonstrate a right to be there. Following this, only where significant correlations between codes within data were found were these social factors drawn into the theory overall. Chapter six discusses how the values and features of the management of PWS are patterned (or not) by these social factors, continuing with the analysis where relevant.
A note on the analysis of drawings: the somewhat paradoxical method for analysing write and draw materials, as noted by Backett-Milburn (1999: 392) has been quantitative in nature, tallying the amount of times certain object or ideas are portrayed (Pridmore et al 1995). Within this research, the nature of the pictures drawn by children with PWS very generally was compared to those of their non-PWS siblings. Four trends emerged from this which are discussed in relation to the overall analysis in subsequent chapters. These were a tendency within PWS drawings towards: illustrating in text rather than pictures, the inclusion of food; focus on objects rather than people, and often copying of the same object several times.

The following chapters outline the selective coding process, giving a theoretical narrative of the analysis.
CHAPTER 4 – PRACTICES IN THE FAMILY MANAGEMENT OF PWS

Introduction

This chapter addresses the main research question of how Prader-Willi syndrome (PWS) is managed within the family, focusing specifically on dietary practices. The term management may be seen to presume a top-down process, where those with power judge aspects of the child’s eating behaviour as in need of correction and act accordingly. However, a sociological exploration of the ‘management’ of PWS takes the form of identifying and describing the everyday strategies, routines, behaviours and practices which are played out within the families. Highlighting the diverse locations of agency and network of relations involved in these practices forms an ongoing theme throughout the analysis. Although explicit rules and decisions about managing diet were made at a parental level, analysis demonstrates that these decisions were the result of emotional, embodied interactions within the family, where children with PWS play a significant part in dictating everyday practices.

The analysis highlights the importance of human embodiment in the everyday experience of PWS and its management within the family. This analysis of embodied action first of all seeks to draw out the role that biological and emotional factors play in the formulation of family practices, before turning to examine how these factors are connected to the values and beliefs of family members, socio-cultural and economic factors. An embodied analysis of family interaction illuminates the many ways that the child with PWS shapes and influences everyday actions, thereby permitting his or her agency to be fully recognised.

Four key features in food management practices were identified from the data:
‘Access-to-food’ practices
‘keeping occupied’ practices
‘Use of routine’ practices and
‘Biological alteration’

This chapter will describe the nature of these practices within the families.
It is useful at this point to return to some of the issues raised in chapter one, on diversity within the disorder. When posing the question of how PWS is managed, it is crucial to bear in mind that the individuals display very different physical and behavioural traits. This means that the nature of what is being managed varies greatly and this is clearly evident within the study sample. Before exploring the key features in food management it will be beneficial to briefly demonstrate some of the differences between the respondents with PWS. As the thesis focuses on dietary control and management of hunger, food seeking behaviours, the nature of hunger and incidence of overweight/obesity will be addressed.

4.1 Food seeking and hunger
The vast majority of families (nineteen of twenty), reported the existence of excessive hunger or a preoccupation with food. In one family the child had never developed this trait, however because his/her fat: muscle ratio was poor, diet still had to be monitored carefully. Of the remaining nineteen, three families had noticed some elevated interest in food, but found that this did not result in food seeking behaviour or pose any immediate problems day-to-day. The rest of the families (sixteen in total) described food seeking and/or stealing behaviours, excessive appetite or obsession with food. However, only five claimed that food management and weight loss was their main concern.

Food seeking and stealing behaviours also varied greatly within the sample in terms of their nature and frequency. In four families, the incidents were occasional and isolated (less than five times in a year), in five there are phases of food stealing typically lasting three to six months, and in seven this behaviour occurred daily, with children described as constantly in the process of devising new ways of obtaining food. The following excerpts from interviews with two of these families illustrate this:
Mother10: ...in a way it moved on from Joe just trying to take food, which he will do given the chance, to working out other ways to get food, and one of those is stealing money. So my handbag is kept under there [cupboard under stairs] which is locked. I used to check his pockets and bag for money. I have a system at school were I 'phone the school if he's bringing in money for a legitimate purpose. We then had a problem because he hid it in his shoes, it's very difficult. The girls [his sisters] have a locked box under their bed. And we have to tell people that come into the house if they leave their bag. Because once he's got money... at school should be reasonably safe, they know no one should ever accept money from him for food, without having the authority from me. He then moved on to asking others for money, which makes him sound terrible, but he'd stop strangers and say "excuse me can I have some money?". Which is completely inappropriate and also very dangerous, you know you ask the wrong person and you could have a real problem. So that's very difficult, him working out that there are other ways he can get food. Having to check the junk mail that comes in. It all gets put in a pile with the newspapers. We found that Joe had been taking out the leaflets for pizza! To ring up! [laughs] And I interrupted him once, so then he realised and actually now the phone's off the hook. There are two handsets and that's why I had that system set up because he might do that and, but actually, also he phones the place. I don't know why! The place know him and Joe knows he shouldn't do it, I don't understand. I can't take the risk, he did it a couple of times and he didn't put the phone on the hook and because they couldn't get through they knocked on the door with a pizza. But it also means if he picks up a number and he could pick up a number for food from anywhere, from so many places. But with all those measures in place we are reasonably sure. But it does mean being several steps ahead of him. (SESH, P1, SIBS)

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Father11: Well what I do is, she goes to school in a taxi, there and back and the way it works it's always the same driver. He's a very nice man really. And everyone at school knows that Kelis eats what she goes to school with and that's it, no one at school will

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1 The identifiers used to refer to families are coded as follows: Mother/Father/Sister or Brother identifies which family member is speaking. Where the child with PWS is speaking a pseudonym is used. The last two characters indicate which family the data belongs to, from 01 to 20. K indicates the interviewer.
give or sell her any food. That’s how it is, all set up. And I never thought there was any need to tell the taxi driver the ins and outs of the disorder, he just takes her there and back you know? Anyway last Christmas he gave her a present which was sweets. And when I spoke to him after that it turned out that she’d really befriended this man and they’d quite often stop for sweets, and he’d buy them. It sounds really dodgy when I say it but, he was very apologetic when he realised, nothing untoward, just he felt sorry for her I think. She’s good with people like that, she’s a bit too good at getting what she wants sometimes. Yes, so Christ knows how long that had been going on and she never said a word. (SESL, P2, SIBS)

Lay conceptions, especially where derived from sensationalised media coverage of the disorder, as discussed in chapter one, tend to characterise people with PWS as biologically distinct due to their experience of constant hunger and possession of ‘insatiable appetite’. This hunger, in turn, is seen as the basis of behavioural, psychological and physical problems within the disorder. However, medically little is known about how people with PWS experience hunger and neurological research continues in this area (Hinton 2004, 2006). There is a suggestion that the satiety response is impaired (Holland et al 1993) and that this may result from hypothalamic dysfunction (Swaab et al 1995). Many health professionals, parents and carers are keen to draw attention to a link between fixations on food and associated behaviours and the trend towards ‘obsessive traits’ prevalent in PWS. There seems to be some consensus amongst these groups that these ‘obsessive traits’ play a major role in exacerbating desire for food.

This is not to say that people with PWS do not experience some high level of physical hunger. They develop various obsessions, but food and food practices remain the most prominent, as seen in the fact that nineteen out of twenty families reported increased interest in food. Moreover, it is very important to draw attention to the rigid and focused understandings and obsessive traits experienced by people with PWS and the extent to which these traits exacerbate the relationship with and desire for food. That is, it is important to appreciate that hunger is the result of an interaction between the individual’s
biological drive and their social understandings. This forms the widely documented highly emotional relationship that people with PWS have with food.

Themed sessions (see chapter three) on food and hunger were carried out with fifteen of the young people with PWS. Although they could all list many of their favourite foods and talk about where and when they had consumed them, they could not describe why they liked them. Neither could any describe or draw the feeling of hunger. This inability to describe emotion and sensation is peculiar to PWS and autistic spectrum disorders. In typical lesson plans for primary school classes about world hunger issues it is standard for children to draw and describe what hunger feels like (Learning to give world hunger lesson plan Source: www.learningtogive.org/lessons/unit140/lesson3.html). However, despite this inability to articulate their hunger, aspects of the children’s food fixations were visible across the data (interviews and drawings) drawn from the themed sessions on ‘hobbies and interests’ and on ‘health’.

(Figure 4.1 drawing by Ed) (SESH, P2, SIBS)
Fourteen of the twenty children with PWS listed cooking or baking as one of their hobbies. When asked to draw one of their favourite activities one child, aged twelve, drew and labelled twelve jam doughnuts.

K: What I want you to do now is think about all the hobbies that you’ve mentioned so far and draw me a picture of one of them. You might want to draw your favourite hobby. Could you do that do you think?
Ed: [nods, starts to draw]
K: Do you mind talking and drawing at the same time?
Ed: No
K: So what is it you’ve decided on drawing?
ED: Baking
K: So you actually made the doughnuts yourself?
Ed: Yes
K: how did you make them?
Ed: Don’t know [pause] with Mummy.
K: Why did you enjoy it, can you remember?
Ed: [pause] We used special\(^1\) jam and we used the bread machine too. I don’t normally have doughnuts. (SEH, P2, SIBS)

The children’s preoccupations with food were also displayed as they frequently brought food into conversations and themed sessions on other subjects. For instance, one 14 year old drew a picture of their favourite hobby, horse riding (figure 4.2), the picture portrayed a horse eating.

(Figure 4.2, drawing by Victor) (SESH, P1, SIBS)

\(^1\) Reduced sugar
Figure 4.3, a child's depiction of their twelfth birthday party, also demonstrates the significance of food in everyday life for people with PWS. Sonia described her party as follows:

K: That's [picture] brilliant, can you talk me through what you've drawn. Tell me about the party and what you enjoyed.
Sonia: [nods] I got three dolls. This one's my favourite [shows K the doll].
K: Oh she's lovely, did you get her at the party?
Sonia: No
K: I'm very interested in your picture, can you tell me about it?
Sonia: yes, this is the food we had, it's a buffet. There were sausage rolls, but vegetarian, these are twiglets, crisps. These are samosas, fruit salad and fruit juice. Then we had jelly and we watched my DVD. It was my idea to have samosas. I made the food with mummy.
K: And who are all these people?
Sonia: My friends.
K: What was you're favourite part of the day, can you remember?
Sonia: The buffet and getting my presents. (SESH, P2, SIBS)

It is interesting to note in figure 4.3 and the child's description, the people at the party seem secondary to the objects, such as the food, TV stack and presents. As much as indicating an increased interest in food, the data reflects a general perception of the world often related to autistic-spectrum disorders. That is, with social relationships featuring less significantly than more tangible objects such as food, TVs, computers. This draws attention back to the interplay between obsessive traits and somatic experience that constitutes hunger within PWS.

The complexity of the hunger in PWS and the idea that it is experienced differently was widely commented upon by parents. For example:
Father15: No I don't think it's hunger like you or I would be hungry. It's not just the eating, it's everything around food that he enjoys, reading the labels and comparing. He'll often find something lower calorie than me if we go to the supermarket, he's very helpful in that way.

K: Does he help with the cooking?

Father15: Oh yes, always supervised though. He loves the cooking programmes [on television], he has said he'd like to be a chef, but we try to steer him away from that a little. Yes, he just got a passion for food and everything that goes with it. He's happier in the kitchen than anywhere else in the house and I think to a point he should be allowed to enjoy it. (SESH, P2, SIBS)

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Mother12: [on hunger] I see it as much more of an obsession than actually feeling hungry, although he does feel hungrier than a child without PWS I think. He isn't that aware of his body at all really, he just knows what time he should be eating and he can get very upset if that doesn't happen! (SESH, P2, SIBS)

This conceptualisation of hunger, as part physiological drive, exacerbated by the tendency towards obsessive traits and the rigid, literal perception of the social world described in autistic disorders, provides an embodied concept of hunger in PWS. Hunger can be viewed as an emotion. The interplay between these two aspects breaks down the idea of separation between mind and body, as the physiological drive for food and the social perceptions common to autistic-spectrum disorders, combine inseparably to produce the embodied emotion of hunger. Synergy between biological and social factors experienced emotionally through the body, as a model for hunger, is not distinct to PWS. As Lupton notes:

Hunger is not often regarded as an emotion, as it is viewed more as a drive or instinct unmediated by social states. Yet it would be difficult to argue that hunger is purely a biological phenomenon, given the web of cultural significations that surround and govern the ways and amounts and times that we eat. The
physiological relationship between the body's recognition of the need for food and the emotional state is clearly complex. (Lupton 1996: 33)

Further to Lupton's comments, it is not simply the symbolic values of food and the body in Western society and micro level cultures such as the family, that are relevant, but how biological differences, traditionally theorised in genetics, neurology and psychology are involved in wider social processes. Within medical sociology there is a broad literature of similar arguments as applied to pain. Thus Bendelow remarks that:

...the dominant biomedical model, which prioritises sensation over emotion, pain, we argue, needs to be seen as a fundamentally embodied experience: one which combines both physical and emotional dimensions of human suffering. (Bendelow et al 1998: 7)

It is important to note here that although it was initially an objective of this research to explore embodied accounts of hunger from the children with PWS themselves, their inability to articulate their experience of hunger, explained earlier in this chapter and chapter three, made this impossible. This research makes no claim to have achieved such accounts. However, through the frequency of food in the children's conversation and drawings and from parental accounts and my own observations, a basic theoretical approach to hunger has been sketched and placed upon the backdrop of current research theorisations of hunger from the sociology of health and illness.

To summarise the theoretical approach to hunger taken in this analysis, physiological factors and social perceptions of individuals are inseparably woven to form the embodied emotion of hunger. Within PWS hunger is experienced uniquely due to particular biology/physiology i.e. hypothalamic dysfunction, and particular modes of relating to the social world i.e. rigidity and obsessive characteristics of autism. The following analysis of food management strategies within families of children with PWS points to the multidimensional nature of hunger by highlighting the various strategies used to control hunger.
Two children in the sample were classified as obese, nine were overweight and nine were a healthy weight, as classified by Body Mass Index (BMI). This thesis acknowledges the contested and culturally specific nature of constructions of fat bodies and medical risk, the objectivity or meaningfulness of these conceptions within the contemporary political climate of ‘obesity epidemic’ is highly questionable (Monaghan 2005). However, these figures give some idea of the weight range within the sample, the real picture contrasting sharply with media representations of the disorder, which exaggerates weight issues within PWS (see chapter one). Having outlined a theoretical approach to hunger and displayed the diversity of food and weight related issues within the PWS, we will now move on to examine the four features of food management practices identified in this analysis; access to food, keeping occupied, use of routine and biological alteration.

4.2 Access to food

One widely used and effective strategy of controlling and monitoring diet within the families was denying the child physical access to food both within and outside of the household. Although a total denial of access to all foods without supervision was uncommon (four out of twenty families), the vast majority (seventeen out of twenty) employed some physical form of denying access, such as alarm systems, locking kitchen or particular cupboards/refrigerators, food placing\(^3\), waste disposal systems and food storage\(^4\).

Locking part or all of the kitchen area was a strategy employed by the majority of families in the study:

- One family used a timed alarm system
- Three families keep all food/kitchen locked
- Eight families keep certain cupboards or the refrigerator locked
- Eight families used no kitchen locks

\(^3\) Placing food on high shelves, out of child’s reach or hiding it in other rooms in the house.

\(^4\) Storing food in an inedible form e.g. refrigeration.
Five of the families that used no locks, placed certain foods out of the child’s sight and reach. Most often this involved placing foods on the top of cupboards or on the highest shelves. In two cases, parents looked after gifts such as Easter eggs and sweets in a separate room, unknown to the child, to ensure they could ration them appropriately. This food placing typically occurred with high calorie foods and foods that needed no preparation, such as sweets and baked products. The way food was disposed of and stored (frozen) was also given attention within the families. In practical terms these physical measures were credited by parents as an extremely important part of day-to-day food management. For example,

K: Have you ever made any large or sudden changes in the way food is dealt with at home?

Mother14: No, not really, we just take it day-by-day. Apart from the single most useful thing we bought for the kitchen was this [points to sink] waste disposal, it’s really good. You put your leftovers in it and it’s just gone, like that. We all use it.

K: Did you have problems before that with the left-over food then?

Mother14: Yes, you couldn’t just leave it on the side to be washed; even in the bin I’m not sure it was always safe. So we all got used to it, just every time all food goes in there.

K: And could you actually notice a difference in his weight because of that?

Mother14: Oh yes. We did see it, half a stone, three quarters maybe...I hadn’t realised before that how much he must’ve been able to get his hands on. I only caught him once or twice you see...he was very good at it! (SESL, P2, SIBS)

K: So would you say that starchy foods are her favourite type of food?

Father03: Hmm, I’m not sure she has a favourite type of food. I know it is anything that’s filling though. If I buy bread I usually put it straight in the freezer, because otherwise it tends to disappear. But vegetables and fruit I can leave out and have fresh no problem. So, yes her preference is towards that way, potatoes, bread, she loves pasta. (SESL, P2, NOSIBS)
A variety of justifications were given by parents for the use/non-use of locks. Responses included not just practical reasons but, in most cases, discussion of moral/ethical implications of these practices for the child with PWS and other family members. These discussions reveal an interesting connection with the widely documented trend toward the privileging of self-discipline and regulation in health (Pollock 1993), diet (Lupton 1996: 131, Bordo 2003: 99) and patterns of individualisation in broader social life (Beck et al 1994). That is, the ability to show self discipline and take individual responsibility for your health, including diet, have been encouraged in contemporary Western societies e.g. UK government policy to educate citizens so that they can self-regulate such as the ‘Five-A-Day’ strategy to increase fruit and vegetable consumption (Sharp, Department of Health 1997). In addition, this ability to self regulate is seen as indicative individual’s moral standing; to look after your own health is to act as a moral citizen. In a society where the value of self regulation in health is so strong, many parents initially made a negative association with the necessity of placing external locks and described a sense of failure where a child could not be taught to self-regulate.

From both users and non-users of locks there was an expression of negativity at the initial idea of food locking. Families’ reactions to external management e.g. locks, can be seen to reflect these wider societal trends, privileging individual self-management and self-discipline. Families made reference to external locking feeling ‘harsh’ and compared it to incarceration, a lifestyle not fitting for a family home. As this non-user recalls:

K: What was it that horrified you, can you remember?
Mother18: [pause] well...I think...I just remember imagining this obese child, this little monster you know. Just trying to get food all the time and into everything. And the thought of living in this prison were everything's locked away. But eventually we got put on to a very good doctor from the city hospital and we worked out it doesn't have to be like that at all. (SESL, P2, SIBS)

This lock user expresses a similar initial reaction:
Mother09: We were quite reluctant at first [to lock food], but it's just part of the set-up now. I don't really think about it. Yes, I'd say the food side of things pretty much runs itself these days.

K: What was it that made you reluctant at first?

Mother09: I don't know really, just seems a bit harsh doesn't it? Maybe something you'd do as a last resort.

K: Do you ever think like that now?

Mother09: I have my moments, but basically we have to go on 'if it works, it works' cos there's too much else to worry about with Jason. (SESL, P2, SIBS)

This quote from family 09 also highlights a theme that was recurrent within the analysis. That is, as locks prevent self-management and external control is dim-viewed, at least initially, in many families, use of locks presents families with a conflict of values and practicalities. Several families also gave descriptions of their attitude towards external control and management becoming more positive over time, in light of their practical experience within the home.

Four families, however, firmly upheld values of self-discipline around food despite their practical experiences. They saw their non-use of locks as a central aspect of how diet/lifestyle was approached within their household. One parent spoke emotively of the importance of allowing the child to overcome obsession with food internally:

Mother01: We don't calorie count, we never have. We don't use diets or any special diet foods. We just stick to healthy food and plenty of it. We don't use any locks on the fridge or in the kitchen. Food seeking is not a problem for us. We have a deal that if Alex wants something extra he comes and asks me and most of the time I say yes...if you want to go on a diet the last thing you should be thinking about is food and calories. They should go and get a hobby and that's a darn sight more important than counting any calorie. It's the same with him. Sometimes if he's busy he'll miss lunch, other things completely override it. Yes there is an obsession there I won't deny it. But if you keep the mind busy with other things the battle's half won. (SESH, P2, SIBS)
Three of the families who used partial locking also strongly privileged internal control over external control where access to food was concerned. These families had moved in and out of using locks over time, but expressed that they were working towards an 'ideal' of a lock-free environment. As expressed in family 02:

Mother02: She does steal food at home. I thought we were on top of that, I’ve noticed things are missing again over the past few months. I think actually the lock is going to have to go on the pantry again.
K: You don’t use the lock at the moment then?
Mother02: No, we always had the lock from when she was about seven or eight, then when she was ten we took it off and that was fine for a while. Then I think we had it on again for a year, and now it’s been about 6 months without.
K: So you prefer not to have locks if possible?
Mother02: Of course. I don’t enjoy doing it. And there’s the rest of the family [children aged ten and twelve and I don’t like to deny them. We want her eventually to be able to live independently and fit in, so she’s got to learn because I won’t always be there.
(SESH, P2, NOSIBS)

To recap, four out of the twenty parents articulated a belief in the value of internal self-regulation on the part of the child with PWS over external control of food. From this viewpoint the objective to be worked towards was for the child with PWS to achieve a level total of self control. Living in a house with no locks was viewed as a more civilized lifestyle, which was fairer to and more pleasant for other family members. Encouraging self-discipline was also seen as an investment for the future, allowing greater social inclusion and thus a greater number of living choices in adult life. Nine out of twenty families voiced an initial feeling of negativity towards the idea of locks in the household.

In contrast, six families who deny access to food exhibit a very different approach to food locking, morality and the emotional effects on both the child with PWS and on the
family. Rather than seeking to cure food obsessions and behaviour around food, these families see their moral duty as resolving the extreme psychological stress caused to their child when he/she is in a situation where there is access to food that they know they are not allowed to consume. They view the behavioural and practical problems resulting from not having locks as more disruptive within the family than other family members not being able to access food. From their own unique experiences of the disorder these families have come to view their children’s obsessive relationship with food as an inevitable part of who they are and are resigned to the prospect that their child will never have a capacity for rational decision making around food. This is made clear in the following interview excerpt:

Mother 10: I know when Joe was young we didn't want to put a lock on the kitchen door thinking that would be awful, and it would be a real sort of sign that something had failed. Although now I think I've gone through the process of realising that I'm at a point where I simply can't control his compulsion to take food...And if I give him a situation where the opportunity is there he will either take it or he will get quite stressed knowing that he shouldn't do it. He becomes agitated and we get problems, so now there are locks on the cupboards where there is food. [Family are about to install alarm system to dispense of locks, family have piloted the system]. If he comes downstairs on to the landing area, that will trigger it. It's not very convenient because if the girls or I come down it triggers it too. But at least I know his movements and without that it's very difficult to get a night's sleep. Once Joe knows the alarm is set he doesn't try. He will sometimes come and say 'can you lock the door' because I think it takes the pressure off. He can carry on with the Playstation. So when he was little I wouldn't have wanted to lock or have alarms, I would have seen it as a failure. But actually it works reasonably well and it means that I can relax when I'm out in the garden [door through kitchen], without looking over my shoulder all the time. (SESH, P1, SIBS)

As seen in the above quote, families who view locking practices positively have reconceptualised notions of ‘freedom’, based on their ‘lived’ experience of managing
PWS within the home. In contrast to viewing external constraints as a means of incarceration, these families outline how extra freedom is gained for both the parent and the child through denying access to food. These two conflicting approaches to the use of external control can be seen to map on to wider discourses. As we have already seen, non-locking families uphold notions reflecting a shift towards individual self-discipline in health and the management of illness. The sentiments of families who use locks can be seen to reflect contemporary medical discourse, drawing attention to the responsibility of the carer to intervene in light of people with PWS' reduced capacity for decision-making around food.

The conceptualization of PWS within the six families who see locks as a valuable and ethic approach to food management, and the shift of responsibility away from the child, strongly reflect the conceptualizations within medical ethics towards the responsibility of others. It mirrors on a micro level the suggested modes for legality of treatment. As seen in chapter one, Holland et al. (1999), Holland being a psychiatrist who gives evidence in PWS related human rights cases, support the notion that, in the vast majority of cases people with PWS lack the capacity to make rational decisions around food. Although they call for the assessment of each child based on their own unique circumstances, based on certain criteria, they strongly highlight the responsibility of the state and its agencies for curbing weight gain where this capacity is lacking:

We examine the ethical and legal dilemmas that can arise in the care of people with PWS. A tension exists between a genetic deterministic perspective and that of individual choice. We conclude that the determination of the capacity of a person with PWS to make decisions about his/her eating behaviour and to control that behaviour is of particular importance in resolving this dilemma. If the person is found to lack capacity, the common law principles of acting in a person's "best interests" using the "least restrictive alternative" may be helpful. Allowing serious weight gain in the absence of careful consideration of these issues is an abdication of responsibility. (Holland et al 1999: 230)
So far we have seen how access to food is dealt with in the families and the various values, beliefs and justifications parents espouse in connection with these practices. From the responses there is a clearly identifiable group who value self-control of the child around food, and who reflect the wider social trend towards individual responsibility, evident in social theory and visible in recent health policy initiatives. There is a second group who recognise a responsibility of care, involving the external control of access to food. This viewpoint reflects macro level concepts of PWS from medical ethics and the legal structuring of responsibility that these conceptualisations command. These discourses of internal/external control are strong and in some cases were passionately argued in the interviews by parents in defence of their lifestyle. However, this should not be taken to mean that the discourses alone determine or cause the everyday actions and practices of the families in the study. The picture is more complex than this, as I will now go on to discuss.

Analysis revealed overwhelmingly that access-to-food practices were connected to the behaviours presented by the child to the parents, who mediate, judge and respond. These practices change gradually in relation to changes in the child’s behaviour, again as perceived by the parent. Ten out of the twenty families did not venture an ethical/moral justification for their use/non-use of locks and described the practice as a straightforward response to the extent of problematic food seeking behaviours presented. For instance, the quote from the mother in family 02 on page 90, displays how locks are placed and removed over time as parents judge the situation in relation to the child’s behaviour. Six other families describe themselves as alternating between periods of locking and non-locking over time. These two quotations taken from interviews with ‘non-locking families’ also illustrate the importance of child behaviour and parental reaction to it:

K: So what were your first impressions? Was there anything you imagined that then hasn’t been a problem or vice versa [diagnosis at 6 weeks]?
Father20: Erm...well it was so overwhelming it’s hard to say for sure…I do remember a few years after that and we’d been reading up – every article and pamphlet we could get
our hands on! We were so prepared for the locks on the fridge and the kitchen you know and it just never happened.

K: You never felt you needed locks?

Father20: No, not at all. It’s her back [scoliosis] and getting her through the operation that’s important now. (SESH, P2, NOSIBS)

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K: So those instances [food seeking/stealing] never made you consider fitting locks or anything like that?

Mother06: Well yes I considered but, when he climbed up on the counter that time and had all those bowls of cereal it scared me so much. He’s so tiny, you know so short and so unsteady, I have no idea to this day how he got up there. So I thought to myself, if I put things up high or put locks on things it’s just going to make the whole thing worse and he’s going to end up really hurting himself or getting angry and trashing a cupboard! And then it’ll be a war and that’s what I try to avoid.

K: So what do you do now to stop him? Or has that sort of behaviour corrected itself?

Mother06: Well I think what’s happened really is I just don’t buy sweets or anything like that anymore. Or if I do I’ll keep them in my bag. I shop a lot throughout the week and so we never have masses in, just as we need it. I stock up on the things you can freeze.

K: Do you find that you freeze things like bread, erm like things you wouldn’t necessarily have to?

Mother06: Oh bread yes I do. All bread like crumpets, pitta breads, pizza bases. Also summer fruits, strawberries what else? All meat is frozen then defrosted in the microwave before I cook it. Yeah basically anything I can I freeze. (SESH, P1, NOSIBS)

The process of negotiating decisions around access to food in the home can be seen as complex and dynamic over time, involving a number of factors. Decisions are made at a parental level. However, the behaviour of the child and its emotional impact on other family members, and in turn their values and ways of perceiving situations, play an important role in the process. Even where parent’s actions and comments can be connected to specific beliefs and trends, existing externally from the family such as the value of self-regulation and internal control, their decisions have a strong relation to the
everyday embodied experiences of family members. That is, parents draw on ideas from wider society to understand and explain their everyday life, however it is the everyday embodied interactions of family members that directs action within the families not these ideas.

Thus the starting point of the analysis of the phenomena involved in managing PWS within families, is that the human embodiment of family members and the relationships between members are the central processes creating the social actions (everyday behaviours and practices involved in body management) that shape the PWS body. The idea of a network of relations, in cases of PWS body management in families, therefore is very useful. Within this first theme of ‘access-to-food’ strategies it also becomes apparent that factors external to humans are incorporated into this network.

The significance of ‘the material’ in influencing everyday actions in food management is highly visible in ‘access-to-food’ practices. Alarm systems, locks and various kitchen technologies; refrigerators, freezers, microwave ovens, waste disposal units and shelving are of great value to families in inhibiting access to food. Existing knowledge about PWS (outlined in detail in chapter one), general societal perspectives on health and the social institutions they are borne out of are also part of this network. Discourses of individual responsibility, self-discipline and control around food, as well as contrasting medical discourse on the nature of PWS i.e. individuals with PWS cannot possess capacity around food and moral responsibility for food control shifts to state and carers, were visible in parental explanations of access-to-food strategies.

Material resources and ideas from medical and other conceptions of PWS must therefore be granted some agency in the processes of the formation of food management strategies. Through the analysis it emerged that these two aspects (material resources and institutional discourse) were secondary to embodied interaction in terms of their agency in determining access-to-food practices. Tools/materials were used to aid food management, yet their use and non-use over time alternated depending on parents’ perceptions of the child’s behaviour. Various discourses were drawn upon to make sense.
of, justify and narrate access-to-food practices, but again the practicalities of child behaviour had a more direct link with practices. That is, access to food was denied within families where food seeking and stealing was a problem.

Where access to food did not need to be denied because food seeking is not a problem, it is unclear whether it is the everyday social practices and beliefs of particular families that, as well as resulting from child behaviour, also cause the child not to exhibit these behaviours. That is, the acts of individual agents within families reproduce and modify the family’s social structure. This includes the family’s belief/action system; the very system that they draw upon to create this action. As we have seen, via embodied interaction, the biological body plays a part in influencing family strategies, however it also acts out these strategies and is the final product of them (See figure 4.4).
(Figure 4.4) THE CHILD WITH PWS WITHIN THE FAMILY MANAGEMENT PROCESS

FACTORS DETERMINING MANAGEMENT STRATEGIES

RESOURCES OUTSIDE OF THE FAMILY

EMBODIED INTERACTION
(Family members including CHILD WITH PWS)

(Everyday actions (family members including CHILD WITH PWS)

OUTPUT: MANAGED CHILD

(Materials, technologies, knowledge and services drawn upon by family to devise and act out management strategies)
4.3 Keeping occupied

Keeping the child engaged in activities as a way of controlling hunger was the main food management strategy in families who did not use external restraints e.g. food locking and placing. However, this practice was described or observed across all of the families in the sample. Even in families where access to food is physically denied, this practice was used to curb hunger and prevent the child instigating a discussion about food. In cases where the child showed no increased interest in food, 'keeping active' was described as a lifestyle and it was implied by the parents that adherence to this lifestyle had prevented food from becoming an issue for the young person.

Eleven of the families described keeping the child occupied as a planned strategy around food. Eight of these families also commented on the usefulness of this strategy in controlling behaviour and emotional outbursts. In the remaining nine families, distracting the child from food through activities was mentioned or observed, but families did not state directly that this was an intentional food or behaviour management strategy.

Keeping occupied took a number of forms. Amongst families where this was a planned strategy for food/hunger management, there was an emphasis on physical activity and active lifestyle. For example:

Mother20: We've always encouraged exercise as a way of life, a healthy life, you know? We tried her at all the special needs sports clubs. Team sports we had a lot of problems with didn't we?
Father20: Yes, when you're playing a game like cricket there was just too much going on for her really to comprehend.
Mother20: I think we had a couple of tantrums and decided that team sports were out [laughs]. But no, she got there in the end, she’s completely obsessed with her gymnastics. They take her for 5 hours a week.
K: Is that after school?
Mother20: Yes, 2 hours at the weekend and 3 days after school in the week. And we worked that out for us too because between school and dinner is the hard time food-wise. So yes that works for everyone. (SESH, P2, NOSIBS)

Many families described the process of subtly working exercise into lifestyle:

Mother09: On Saturday afternoons we quite often have a treat. We'll go to the baker's and he can pick a cake or biscuit. But the deal is we walk there and back, it's about a mile each way and because the whole thing is about food he gets excited and he doesn't even notice the exercise. Yes that works well, it's nice to treat them, I think you can as long as you're sensible about it. (SESL, P2, SIBS)

Within eighteen of the twenty families, a way of 'keeping occupied' aside from physical exercise was described or observed. Parents described their children becoming absorbed in a number of solo activities within the home. These included: jigsaws, puzzles (particularly word searches), knitting, sewing, collecting objects, computer PC, games consoles, snooker, reading, jewellery making (particularly beads), reading and copying text (handwriting or typing out text from the Internet or Ceefax/Teletext, particularly sports scores and lottery numbers). The tendency towards obsessive focus and linear information and activity, often characterised as autistic spectrum (Bass 2000: 219) seems to result in people with PWS being able to spend long periods of time involved in these activities. This behaviour has been documented in psychological and psychiatric studies, for example, the work of Dykens (2002) and of Clarke et al (2002).

The psychologist and PWS specialist Elizabeth Dykens (2002) found that children with PWS have unusual jigsaw puzzle and word search skills. She compared three groups: children with PWS, children with learning disabilities and a similar IQ level to those with PWS and a control group of ‘normally functioning’ children, all three groups were the same age. She found that the PWS group far outperformed the learning disability group on both word searches and jigsaws. She also found that the word search skills of children with PWS matched those of the ‘normal’ peers and that in PWS jigsaw skills were
significantly more developed than the ‘normal’ peers, children with PWS being able to place more than twice as many pieces (Dykens 2002: 343).

Clarke et al (2002) associated PWS with ‘high rates of ritualistic behaviours, such as the need to ask or to tell something, insistence on routines, hoarding and ordering objects and repetitive actions and speech’ (Clarke et al 2002: 358). He also noted a difference between these characteristics and the behaviour of people with obsessive compulsive disorder. Checking, cleaning and counting compulsions were not found in people with PWS. Thus Clarke (2002) suggests that the way in which people with PWS experience obsessive traits and tendencies is unique to the disorder.

The language of Cartesian dualism is often used by parents when describing the success of this strategy, elevating the position of the mind by displaying its power over the body. For example,

Mother01: If you can keep the mind occupied then the battle is half won. (SESH, P2, SIBS)

Father04: When she's got her mind engaged with her reading or sewing then we don't get any of the problems, because the body doesn't matter at those times, she doesn't feel hungry. (SESL, P1, SIBS)

Mother14: He forgets about his body [hunger] when he's using his brain. (SESL, P2, SIBS)

However, this behaviour equally could be viewed as a display of the essential connectedness of the whole body. As the person with PWS becomes engrossed in one activity, they do so as a physical as well as thinking being. The process is therefore essentially embodied, seamlessly incorporating emotion and physiology. These non-food focused activities act to prohibit both food seeking and emotional outbursts and are thus an important part of everyday behaviour and food management in the home. Some
families (nine of twenty) are aware of their significance and encourage these activities, in others (nine of twenty) the phenomena occur naturally and are not thought of as food management techniques. This is clear from the following interview excerpts:

Mother03: When she gets in from school she has a Cup-a-Soup and Weight Watcher bread, one slice with no margarine. Then she'll go and play on the computer and she normally stays up there until supper.
K: How long is that?
Mother03: Well she gets in at 3.30 and I make supper for 6 sometimes 6.30. I'm trying to make supper later gradually. Because it's between supper and bed time she will start asking for more food and we have the behaviour problems. I think it's because I've got a rule about no computer in the evenings. But yes, that's a bit of a problem time. (SESL, P2, NOSIBS)

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Mother08: She's a good reader. She can read quite well for her age...she gets quite involved with it, so that's useful if we [mother and siblings] need a bit of peace and quiet. (SESL, P1, SIBS)

Some of the activities described and observed could be construed as obsessions themselves. Parents in some cases voice concern and speak of the problems of finding the balance between managing food and managing other obsessions. Thus:

Mother10: He does have this obsession with magazines, we try to take them away and he says no, and he'll be scribbling. He does, I think he has quite an obsession; sometimes he'll seek activities that are very limited. Like switch Ceefax on and write the cricket scores down and he'll write pages and pages and pages of it. He likes [laughing] he goes through phases of using playing cards. He has cards and he likes to play pairs and he'll take several packs and lay them all out beautifully in rows and if you disturb them he gets very upset. And he plays quite a bit, he's very good at it, very good memory for doing that. He does it again and again and again. But at the moment he's not doing that. But it's cyclical, it will come round again and he will, that will be his thing. When he was little it
was the Yellow Pages, if we went to someone's house he'd ask for a copy, and go through it. A lot of them seem to relate to paper... so he carried the bag...I'll show you. When he's stressed he just takes magazines and takes cuttings, all sorts of things. He's cutting out pictures and he never actually does anything with them. But he'll go through religiously and cut. If you try to throw it away he gets upset. He often takes it when we go out, he doesn't do anything with it, just takes it with him.

K: Does that sort of behaviour concern you?

Mother10: Well only in so much as I know he's happiest when he feels like he fits in and he's part of things. That sort of thing makes him stand out and in the end adds to his unhappiness and frustration. I try to train him to fit in socially but it's a big problem for us. (SESH, P1, SIBS)

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[Jason sitting at computer copying tennis sets and games tables]

K: What's that?

Jason: It's tennis

K: Is it Wimbledon?

Jason: Yes

K: How often do you write out the scores?

Jason: Just when they're playing.

K: Who do you want to win?

Jason: I want Tim, Tim Henman.

K: There's lots of scores aren't there, lots of matches? How long does it take you, do you know?

Jason: Not sure. I do it sometimes before bed.

K: Does it make you sleepy?

Jason: No. (SESL, P2, SIBS)

As well as evidencing some of the non-food obsessive behaviours displayed by the children with PWS, the quote from family 10 also raises parental objectives of the child's 'happiness' in the management process. This emerging theme will be discussed further in
chapter five, in terms of how the child's long and short term happiness is negotiated by parents.

The phenomenon in 'keeping occupied' as a food/behaviour strategy, again illustrates that everyday practices central to managing diet are the outcomes of family members' human embodiment. In the case of 'keeping occupied' practices, the capacity of people with PWS to focus rigidly on what others may perceive as simplistic or two-dimensional activities for long periods of time, has come to play a key role in how hunger is controlled within the families. Everyday routines form around these activities regardless of whether or not they are recognised or encouraged at a parental level.

Although the agency of the child with PWS is clearly evident here, key decisions about these practices are still taken at a parental level. Parents encourage sporting activities and active lifestyles, or sometimes curb food-distracting activities such as computer games or hoarding. This draws attention back to the importance of embodied interaction and relations between family members, particularly parents' emotional responses to their child's behaviour.

Again there is space within the processes described to acknowledge the influence of knowledge external to the family on everyday action, specifically the media attention given to the danger of allowing children to over use computers and other sedentary behaviour, related to the childhood 'obesity epidemic'. The agency of material objects and technologies, however, emerges as a strong theme in 'keeping occupied' practices. The way children with PWS interface with computers, texts, jigsaws, playing cards etc. suggests these objects have elevated agency and social significance in their perception of the social world, as depicted in figure 4.3.

Actor Network Theory (ANT) is a useful interpretive framework for 'keeping occupied' activities, which to a point also correspond with other themes in the analysis so far. The basic principle behind ANT is that society is made up from networks. The networks that make up grand organisations are, in essence, the same in nature as those that make up the
minutiae of society. This relational approach fits with the importance of interaction as discussed earlier. The congruency between micro and macro levels, acknowledged by ANT theorists, is also well suited to the analysis of management, at a micro level, within family units. ANT draws attention to the idea that these networks are not simply relations between individuals and groups of individuals; they are made of heterogeneous materials: ‘machines, animals, texts, money, architectures – any material that you care to mention [...] we wouldn’t have a society at all if it weren’t for the heterogeneity of the networks of the social’ (Law et al 1999: 7). Although this framework is extremely fitting to the process of the child with PWS engaging with texts and machines and the agency bestowed by these material agents, it fails to explicitly acknowledge the pivotal role of embodiment that is so important to the current analysis. Thus, although ANT has an interesting and relevant connection to the emerging analysis and is central to trying to overcome the subject/object divide, ANT alone is an inadequate backdrop upon which to set many of the other behaviours described.

As a relational theory, stressing the multidimensional nature of these dimensions and highlighting the role of real materials, Burkitt’s (1999) theory of embodiment has many similarities with ANT. In addition, the theory takes embodiment as its central theme and provides an appropriate theoretical foundation upon which the analysis of family PWS management can be set. His work on the body in social relations unifies three traditionally distinct ways to theorise the body; as a thinking entity, a symbol and a physiological being. Through this synthesis and the discussions it raises, Burkitt’s theory incorporates the emerging theoretical themes within this analysis. Firstly his theory, where it pertains to social action, makes relations between embodied subjects a focal point:

We are located in relations that transform the natural and social worlds in which we live. It is within networks of interdependence that we can affect the actions of other people and also change the face of reality [...] we are locked in relations of communication with one another and through the discourses or speech genres of our culture, give expression to the conditions of our lives. As with the power of
production, communication can bring about change and can allow people to challenge the prevailing ideas of their day. In both these ways all individuals become powerful bodies, with abilities and capacities which can radically alter the conditions of life. (Burkitt 1999: 2)

Secondly, Burkitt works both discourse or existing knowledge and materiality into the network of relations, demonstrating the connected parts they play in processes resulting in social action, yet primarily, marking how these dimensions are used by embodied beings:

There is no absolute separation between materiality and knowledge, for these can be understood as dimensions, interconnected through relations and practices mediated by artefacts, involving the thinking bodies of persons and selves. (Ibid: 89)

The ideas articulated by Burkitt on the reciprocal nature of the social relations of the body, also incorporate an embodied interpretation of 'structuration' that could occur within the micro environment of the family. This occurs first by his continued assurance that embodied beings are central to the creation of the social world (structure) and second, through his observations of the significance of artefacts and knowledge, created by persons over time, returning to form part of individuals. Having examined 'access to food' and 'keeping occupied practices and begun to theorize the actions occurring within the families, we will now turn to the third feature in family management 'use of routine'.

4.4 Use of routine
The examination of 'keeping occupied' practices illustrated the way in which embodied qualities of PWS shape food management strategies. 'Use of routine' practices within households, in terms of types of food consumed, location of consumption and time of consumption, display a second way in which the social perception of the child with PWS fashions the nature of everyday food management. Within the sample, three types of family were identified. Firstly, (seven of twenty) those that stick to a very rigid structure of food provision in response to the child’s preference and to the behavioural problems that can occur if a rigid structure is not followed. Secondly, (nine of twenty) families that
have a loose routine which changes to accommodate family life and activities. These families however acknowledge their child’s desire for structure and repetition and make gestures towards it. In the third family type (four of twenty) children with PWS do not display a preference for rigid routines and thus integrate into the families’ natural eating patterns.

The majority (sixteen of twenty) of children with PWS present a preference towards rigid routine, in terms of both food provision and general social life. In medical terms this behaviour is often linked with the common incidence of autistic spectrum traits within chromosome 15 abnormalities (Cook 1997). This manifests in the current data through a need for certainty about future events and a preference for food types tried before. Where children are used to a rigid structure, diversions from it can cause high levels of anxiety.

Within seven of the twenty families, food provided for the child with PWS was described as adhering to a very rigid structure. Meals within the household and food taken to school were eaten at the same time everyday and, in three families, always eaten in the same room. In four families, meals were eaten in the living room and dining area/kitchen. The types of food eaten for breakfast, lunch and snack remained constant on weekdays for the child with PWS, but often varied amongst other family members. The content of evening meals varied, but the child was told in advance what the meal would be. In all of these accounts this structure was not something enforced on the child by the parents for ease of monitoring calories, but had developed because it was what the child felt most comfortable with. Diversion from the routine was described as causing anxiety for the child and often leading to angry outbursts. In two of these families the child with PWS ate different food from the rest of the family. In five, breakfast, lunch and snacks were different from the rest of the family, but the evening meal was shared with siblings and/or parents. Eating out was problematic for these seven families unless planned well in advance:

Mother09: When he gets home – has to have his cup of tea and a Go Ahead [low calorie] biscuit.
K: Is that everyday?
Mother09: Yes everyday at the same. The others [children] like different things, but he never wants what they’ve got, like normal children do you know? It’s strange. For him it’s all about knowing in advance, just being certain about what’s going to happen.
K: That’s interesting.
Mother09: And that’s not just food that could be, just visiting friends or whatever: “Where are we going? What’s going to happen when we get there? Who’s going to be there? Why are we going?”[laughs] “When will we be coming home? What’s going to happen when we get home?” It can get very tiring! But he just always needs to know. (SESL, P2, SIBS)

Nine families described themselves as having a rough schedule, but one that varied with the fluctuations of events and demands of family life. Meals could be an hour either side of the demarcated time. Similar, but not exactly the same, foods were eaten as breakfast, lunch and snacks, the child with PWS having a more constant diet in terms of food types than other family members. Evening meals varied, and in 3 of the nine families, were eaten with siblings and/or parents. In six families evening meals were eaten with siblings and/or parents where possible, but after-school clubs and parents’ work and other commitments prevented this from happening on a daily basis. Meals were eaten in the dining area/kitchen and living room. Within these families eating out was sometimes problematic, but was not avoided.

Although the routine within these nine families was looser than the seven identified earlier, these families acknowledged that their child was most comfortable when they knew exactly where, when, what and with whom they would be eating. Within five of the families, the idea of children preferring the same food everyday was raised. Seven of the families spoke of resisting their child’s desire for a stricter routine, either for the sake of quality of life for other family members or training the child with PWS to cope with flexibility and change in preparation for the future, or so they can be included in family activities:
Mother06: In terms of the routine, breakfast is pretty much the same. At lunch time we have sandwiches. Frank tends to like to have the same sort of thing, the same sort of bread, as you saw it today. So in that sense it's quite easy to restrict his calories because he likes everything and he likes a pattern to what he eats. So you can feed him the same quite limited foods. And meals aren't necessarily at the same time because it's not practical and also it wouldn't be terribly helpful if Frank expected dinner at the same time, we couldn't do it. So we try to keep it reasonably flexible so that he doesn't get fixed on certain times.

K: That's interesting because I think there are families that feel like they don't have a choice.

Mother06: I know, it's one of the ways they recommend for managing. Saying you eat at these specific times and not outside it. But the way that we live that's just not realistic. And if he were too, with Frank, used to specific times and no food outside of that he would find it very hard to cope. And there are so many things that we do socially that in some way or another involve food – if he goes to watch cricket he likes a cup of tea, if we visit friends there might be food, and it won't be the same as here. No it's just I think, recognising that Frank likes routine is fine, but it's important he learns to cope with variables and people living differently and that would seem quite hard if he went to stay with somebody else and they did things differently, and he wasn't prepared for that he wouldn't be able to cope with that. It would bring other problems because, with change it's difficult. I think the main thing with Frank is knowing in advance, if you tell him in advance that lunch is at one, two or three that's when he will be eating, then that's when he will be ready to eat. (SESH, P1, NOSIBS)

Five of the families who did not adhere to the child's preference for a strict eating routine at home expressed that they are more likely to give in to the child's wishes when out of the home. This occurred in order to limit behaviour problems.

K: And do you get a chance to go on holidays as a family?
Mother05: Yeah we go on holidays every year. We stay [Northern England] on a farm, self catering, it's not a problem. We eat more or less there as we do here. We have lollies, ice creams perhaps a bit more often.

Mother05: We have fish and chips sometimes don't we? It's ok because it's a treat, we don't do it very often. It's more trouble than it's worth, saying no.

Father05: He doesn't have chips at home. But if we're out, we learnt very early on, there is absolutely no point in saying 'you can only have this and this' because it just causes so many problems.

Mother05: But that's not just food, I think that's generally the case with Ed. It's not that we give in just on the food side, perhaps that's too strong a word. But we're more relaxed with whether it's what's on the menu or other things that he wants to do when we're out. Because we know that he'll just erupt, from experience.

Father05: And for five years we couldn't even go out as a family and enjoy it, because every single time, you know, we'd have an 'event' [emotional outburst]. We took him to a wildlife park and he loves trains and there was a train. So his focus was train and picnic and that was it. So we did the train and then it was just, "when's lunch?" And so, because he's focused towards that, it creates tension. Because he just wants to do certain things and he wants to do them straight away and he won't do the other things...(SESH, P2, SIBS)

Within the families with more flexible routines, shared rules about balancing out treats were common, as demonstrated by these parents' comments:

Mother19: If there's an occasion and there's a buffet we still go, you can't deny them that sort of thing. We do our best to monitor, but usually we are a bit more lax. Sonia knows if she overeats we make up for it later, that's always been the case and she accepts that. (SESH, P2, SIBS)

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Mother14: We have a rule that if he has a treat if we're out or something then he will go without later. He knows it's going to happen, it's automatic, no debate. (SESL, P2, SIBS)
These families included many ‘gestures towards routine’ within eating habits:

K: Does he choose what spread he has [at breakfast]?
Mother 18: Yes, but he loves his marmite on toast. He did ask for sandwich spread once, but now it’s back to marmite every morning.
K: How long has that been?
Mother 18: Oh, probably three, four years now! (SESL, P2, SIBS)

Where approaches to food are more relaxed on special occasions or in public places, rules to negotiate ‘treats’ are drawn up within the family and gestures towards routine are made, the importance amongst families of occasionally letting the child dictate what is eaten is clear. In some cases this can be seen as a strategy to avoid problematic behaviour, elsewhere it results from parents’ belief that their children should be allowed ‘a break’ in order to enjoy themselves. This trend highlights the two-way (child/parent) nature of food management strategies within the families of those with PWS.

Within only four of the families, a desire for rigidity in eating practices on the part of the child did not come across in the data. These families did not keep to rigid time structures; instead meals were eaten when there was a chance to do so. The families described eating together once or twice a week, with siblings eating together more often. The content of all meals varied for all family members. Food consumption generally occurred in the dining/kitchen area or living room. Three of these families made reference to the child wanting to know when and what they would be eating leading to occasional behaviour problems. Although desire for routine was not clear in these families, two families mentioned that the child had and enjoyed one permanent daily food habit, which might be seen as a ‘gesture towards routine’ e.g. ‘hot chocolate before bed’ (family 06) and ‘sausages with ketchup on a Friday’ (family 10).

As with ‘keeping occupied’ practices, the majority of the respondents shape everyday food provision towards the social perceptions of the child with PWS and how they relate emotionally to everyday events. Where this is resisted is in consideration of other family...
members, their needs, activities and quality of life. These findings highlight the creation of social action through the embodied relations between family members.

Both ‘keeping occupied’ and ‘use of routine’ practices are simultaneously behaviour and food management strategies. This highlights the emotional nature of hunger and draws attention to the connectedness of embodied processes which traditionally have been set apart in popular understandings and scientific discourse. Concepts of behaviour and hunger have been subject to characterisations at the heart of Cartesian dualism; behaviour in the cognitive realm of the mind and hunger in the sensory realm of the body. Examples from the management of hunger in PWS demonstrate that managing the body is not merely a question of nutritional science and biology, but of managing the whole emotional, thinking and biological being. We will now move on to examine the fourth key feature identifies within management practices, biological alteration.

4.5 Biological alteration

A number of pharmacological products were used by families to modify the biology and behaviour of their children with PWS. However, use of Human Growth Hormone (hGH) in particular was described by parents as being central the management of the child’s body, both that it direct affects the body and changes the child’s behaviour around food. HGH is a potent hormone with a wide variety of biological effects including increasing rate of growth, final height, bone density and muscle-to-fat ratio. Having been the subject to high levels of controversy for its use to improve athletic performance and in defying the aging process, the prescribing regulations differ widely from country to country. Throughout Europe and the US it is most commonly used in children with poor growth caused by growth hormone insufficiency, renal failure, Turner syndrome, PWS or idiopathic short stature (Hintz 2004: 907).

Of the respondents, ten out of twenty children with PWS have been receiving hGH for over a year, one for six months. Eight of the twenty families described the effects of hGH as playing a powerful and central role in the maintenance of weight. Several families explained how hGH had allowed them to be less strict about food. Thus:
Mother05: The effects of growth hormone have been brilliant. It took a while to notice, but he has so much more energy and that has a knock on effect. The more he’s out expending that energy obviously that’s time when he’s not hanging round the kitchen, but mainly, the more I can give him at meal times. Most days I give him exactly the same as everyone else, which makes a big difference to him.

K: That makes him happy?

Mother05: Yes, there are slight differences in the dessert, Muller Lite. But the main meals are almost always just the same and that’s nice for me too really.

K: It’s easier practically or you feel better with the idea of their meals being equal?

Mother05: Both, definitely easier. But not having to go through, ‘but why this, why that’ and sometimes it carried on way after the meal [shakes head]. (SESH, P2, SIBS)

A raised level of energy was commonly associated with hGH in the families. As well as the basic energy output/input influence on diet, the positive psychological effects of being able to be active and participate in family activities were attributed to making food management easier:

Mother06: It’s [hGH] allowed him to do much more.

K: Because he’s physically stronger?

Mother06: Yes, like we can play out in the garden for longer without getting tired or irritable...he’s more happy generally.

K: Are there less food related behaviour problems?

Mother06: He is more relaxed about food, maybe because he’s busier, more interested in other things. (SESH, P1, NOSIBS)

As well as the enhanced physical performance and it’s positive psychological effect on self image and general behaviour, families associate other physical regulating qualities with hGH i.e. sleep and temperature.
Mother12: In the morning we used to have to be very careful because he’d get up early, come down and have breakfast and creep back up. Then we’d all get up and we didn’t realise sometimes and we’d give him another breakfast.

K: That doesn’t happen anymore? Was it a phase do you think?

Mother12: I think what’s happened is since he’s been on the growth hormone he just sleeps through. He used to nap a lot in the day but that stopped with the growth hormone treatment as well. (SESH, P2, SIBS)

Claims that hGH regulate behaviour and have positive psychological effects on the individual are interesting here as the opposite was also often claimed. In two families with young males with PWS a phase of violent behaviour, as family 13 describe:

Mother13: It took eighteen months before we really started to see it but it’s all very positive. To start off with they said that because he also can be very violent, and he had a lot of it round about that time [start of hGH treatment] they were concerned about a connection. But he had shown that behaviour a lot earlier as well, and I think it just happened that he had a particularly bad episode at that time. He’s very good with times [remembering when to take medication] actually he will come up to you and go [gestures cupping hands]. (SES, P2, SIBS)

Both the positive psychosocial effects of hGH on the individual and the connection to violent behaviour have been identified on in paediatric research in the area. For example, Whitman et al. (2002) conducted a two-year case control study on adolescents with PWS. Although their hypothesis of positive psychosocial effects on behaviour was proven, showing a major reduction in depressive symptoms, the authors also documented some negative behavioural effects, increase attention deficit/hyperactivity disorder symptoms. However, they conclude that the benefits were much more frequent and thus outweighed the relatively rare occurrence of these negative effects (Whitman et al 2002).

This conclusion was reflected by the families using hGH in the sample. Positive psychological effects and detailed, specific social reasons for these were given by the
families. Where violence was reported it was short lived, so the general improvements in quality of life for all family members won out.

Of the nine families not using hGH, one family had no knowledge of the medication; one family had decided against because of possible worsening of their son’s scoliosis caused by sudden growth; and three families were in the process of considering use but had concerns about scoliosis and the unknown long-term effects of the drug within the disorder. The remaining four families expressed negativity towards the idea of using hGH on a variety of grounds:

K: And does Diane [child] use Growth Hormone at all?
Mother04: No, the thing is I’m short, Marks’s [ex-husband] short and Diane is not out of proportion for that. We’re just short people and I’ve never seen it as a problem and neither has Diane. It’s all part of who she is, I think it’s possible to go too far trying to change things. I concentrate on helping Diane accept herself for who she is and being happy, that’s what’s important. (SESL, PI, SIBS)

Father15: No we avoid all that [hGH]. Good for them if they’re happy with the results [other families]. No we try all the ways that don’t involve drugs first and we cope fine. I think it would have to be really unliveable before we looked to drugs.
Mother15: Basically yes she is what she is and yes, she’s different but at some point people have to accept that. (SESH, P2, SIBS)

These comments chime with the sentiment of the social-model disability rights movement (UPIAS 1976, Oliver 1990), refusing to alter their child’s biology in order to adhere to social pressures to be a standard height or display behaviour to conform to a socially considered norm. The quotation from family 04 above also indicates that families make sense of their child’s body and how it should be treated with reference to their own bodies and to their family history. This practice of understanding bodily normality with reference to other members of the family, rather than medical science definitions, has been highlighted in health research into families and interpretations of weight in their
adolescent children (Backett-Milburn et al 2006). Backett-Milburn et al found that in low-income Scottish families, understandings of body size were contextualised within notions of family inheritance of characteristics. Further to this these understandings were put forward by parents and grandparents as justifications for non-interventionist strategies. The quote from family 04 displays this process occurring within the families of children with PWS, where short stature is described as a family characteristic rather than a symptom of PWS and this is used as a justification for the non-use of hGH.

The remaining two families who chose not to use hGH had concerns about the general character of the product. Thus:

Mother07: No we’re going to steer clear of that for as long as we can.
K: What is it that worries you about Growth Hormone?
Mother07: Well it’s just so strong isn’t it? I’m sure it must be quite dangerous, when you see the athletes on it and everything. That’s not what we want for Gemma. (SESL, P2, NOSIBS)

Using hGH as part of food management strategies, effectively alters the way individuals with PWS relate to and respond to their social environment by altering their biology. As well as being a stark indicator of the agency and role of the biological in processes determining social action, this practice exemplifies that human embodiment involves the biological meeting and combining with the social, mediated by emotion and resulting in behaviour/action. This is particularly evident in this study in terms of the positive emotional effects attributed to greater achievement at sports activities or being included in family life or peer activities to a greater extent, and the way these feed through to identity and behaviour. Or what medical research refers to as psychosocial effects; effects involving both psychological and social aspects, relating social conditions to mental health (Hansen 2003).

So what can this examination of ‘biological alteration’ practices bring to our theorisation of family practices so far? All four themes of food management and the use the hGH in this chapter have highlighted embodiment as a pivotal factor. ‘Access to food’ explored
the use of external constraints and 'keeping occupied' and 'use of routines' focused on
how the child with PWS apprehends their world. Use of hGH in families combines
external (a prescribed hormone) with internal factors (its embodied effects).
The use of hGH in family management of the PWS body draws us back to Burkitt's
theory of embodiment and social action (1999). Firstly, as Bhaskar he appreciates the
ontological realm and draws the materiality of world into the processes he discusses.
However, unlike Bhaskar he does not support a division between the ontological and
epistemological. Rather he maintains that they are brought together in relations and
actions of embodied beings. These actions also have the power to transform the real;
transform those embodied beings themselves. The use of hGH is a good example of this
process, hGH being a tool created by humans which transforms the human body. People's
decisions to produce or use hGH are based on (epistemic) existing medical knowledge
and symbolic, cultural ideas of how the body should be.

4.6 Embodied interaction

As this theoretical treatment of the action involved in managing PWS in the family
begins by acknowledging the centrality of embodied relations between family members,
it is useful at this point to look explicitly at the interaction between the child with PWS
and the parent(s). Of particular interest here are negotiation structures and the use of
language and emotion in relation to food management.

The children with PWS in this research displayed ways of expressing and understanding
which were significantly differentiated from those of other family members. In medical
literatures such as the diagnostic criteria for PWS, people with PWS are described as
having impaired social cognition and perform poorly compared to control groups in
psychological tests such as 'facial emotion recognition'. Speech impairments are also

The differences in ways that children with PWS and their parents relate to language and
apprehend their environment has a substantial impact on communication. One noticeable
aspect of comprehension in PWS is that words and phrases are taken literally, as seen in
the following conversation with a mother:

Mother10: I was so busy one day and I was going out. He always has to know where, so I
said to him 'I've got to see a man about a dog' and Joe got very agitated because he really
thought it meant we were getting a dog “who was this dog and when could he go and see
the dog”. I said “It’s just something people say”, but it was too late because I’d already
said about the dog so he was just fixed on it and [laughing] I think that's an autistic thing.
K: That's something I had to change. Not being sarcastic or ironic in any way. I didn’t
realise I was until I started working with people with PWS.
Mother10: No, it doesn't work, they don't understand. Joe has a sort of naivety, there's
something about the sort of innocence. He's very spontaneous there are lots of lovely
things about him. And I just think that if we could manage all the other stuff then you
could see that lovely side all the time instead of the other side, which can be very
difficult. (SESH, P1, SIBS)

Parents’ accounts of conversation around food management showed that interesting use
could be made of this literal comprehension. If the words used to describe a food
reflected a certain image, children tended to accept that, to a degree, even if it did not
match up with reality:
Mother18: it used to be Weight Watchers’ soup but it's now Campbells’, condensed you
see, double the amount [laughs], or so he assumes. But we're talking very little calorie
difference, so that's absolutely fine by me [laughs]. (SESL, P2, SIBS)
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Mother04: ...well we call it milkshake; it’s just low fat flavoured soy milk. But if you
say, you know, “ohh let’s have a milkshake as a special treat” for him it’s like chocolate
or cake or something. (SESL, P1, SIBS)
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Mother13: He loves his mini pizzas and chips. It’s not pizza at all really it’s low calorie
bread with lots of veg and low fat cheese, just a sprinkle. Then a very small amount of
McCaine’s low fat oven chips, just a handful. But because the bread is only 50 calories a
slice that means he can have 3 mini pizzas and chips [laughs]. Which to him, yeah, it's just like “wow thank you Mam”. He thinks it's Christmas, and job done [laughs]. (SESL, P2, SIBS)

One interesting and common form of negotiation involves letting the child make choices around food. Parents describe how it is geared towards their child’s way of seeing and useful in controlling emotional outbursts:

Mother05: We never, we don't say you can't have...we say well yes you can have a little bit if...when you get home you can't have a snack and let him decide. So if Ed wants a treat he can have one if he goes without a little later.
Father05: Yes and that's really worked for us because well firstly your not saying “No” because he'd just go off if you did! And also you're telling him about what's going to happen in the future, which he likes too. And if he makes the choice if he tries to go back on it you say “this was your decision” and he remembers and he takes it on board, he’s very good about that. It's not, when you say it like that it's like we're tricking him.
K: No, I can see what you're saying. You found a way that gets through, that's effective.
Father05: Yes, just finding a way to communicate that actually works, it can be very hard. Because a lot of the time you say things to him and he says 'yes I agree, I understand' but he doesn't really. (SESH, P2, SIBS)

The behaviours and strategies exhibited how everyday practices can be tailored to suit the emotional needs of the child with PWS. The nature and workings of emotion, and it's synthesis of the social and biological has become a field of sociological enquiry in it's own right, addressing areas such as gender, labour, childhood, education and health. In this sense the ‘sociology of emotion’ has appreciated the embodiment of human life since it's formulation in the 1970s. As a founder of the sociology of emotions, Arlie Hochschild notes:

Emotion is a biologically given sense and our most important one. Like other senses, hearing, touch, smell, emotion is a means by which we continually learn
and relearn about a just-now-changed, back-and-forth relation between self and world, the world as it means something just now to the self. (Hochschild in Bendelow et al. 1998: 6)

Although the sociology of emotion recognises the agency of the biological body in processes connecting the self to society, the emphasis has broadly been upon measuring how social values, factors and structures affect individuals’ emotional experience. Embodied studies of childhood touch on how biology over the lifecourse can alter emotional experience (Prout, 2000), yet still takes as its central focus the role of the social and what is external to the body, as that which differentiates emotional experience in individuals. Exploring emotional relations within the management of a complex, multi-system chromosome disorder like PWS, displays the immense influence biology can have on individuals’ experience of emotion and thus, the self, this experience still being based in relations with others and mediated through language and social environment.

Burkitt’s (1999) concept of emotion in his theory of embodiment leaves space for biology and biological difference to play a major role in individuals’ relationships to the social world and the self. He argues that social structures, discourses and the body all play a part in the formation of emotional experience, but does not give preference to detailed explanations of how social factors effect emotion with biology/the body forming a backdrop or context for these processes. He leaves the triadic relationship open, allowing his theory to incorporate the diversity of human forms:

Emotions exist only in the context of relations and are to be conceptualised as complexes; that is, as irreducible to social structures, discourses or the body. All these elements are constitutive of emotions, which are felt only by active, embodied beings who are locked into networks of interdependence. It is not discourses alone then that construct human subjectivity, but the relations in which humans are engaged. (Burkitt 1999: 127)
This idea of networks, primarily between embodied beings, but including the non-human aspects in materials and discourse, has emerged as central to the analysis of the family management of PWS. In respect of relationships between family members, the analysis so far has concentrated on the agency and embodied form of the child with PWS in creating social actions. However, of equal importance are the embodied forms and understandings of other family members, such as siblings.

Incorporating a family member with a significantly different way of relating to the social world poses many problems within the families. Not least of these were personal relationships between various family members. Each case family was unique in this respect and, since the nature of family life being dynamic and multi-dimensional, it is hard to pinpoint the precise effects on family members. However the data convey a strong sense of the emotional impact on all family members. This is demonstrated in the following interview excerpts:

Mother08: Her [child with PWS] and Chris [brother] don't get on...full stop. And I think partly he was not quite 2½ when she was born and he got all this, he'd always been a clingy baby and the whole atmosphere of the house was very angry [Mother08 perceives this as husband's influence]. My oldest son was quite introvert and glad to be out of the house and Abby [sister] at 16 ran away from home, tried to commit suicide and was self-harming. And I said that's it, it has to stop and you have to go [to husband, divorce]. I was trying to hold things together for the sake of family unity, but it was just destructive. And Chris [brother] was growing up to be a very angry little boy [due to Father also being of an angry disposition], and at 2½ Hannah's [child with PWS] back in hospital for a week and Mummy's disappeared [from viewpoint of brother]. My mum was here. He just went into a depression and since then ...They had to share a bedroom until he was eight and she was five, and he can't stand any of the noises she makes, just eating or chewing on her hair. He gets into such a rage. He'll be on his best behaviour if someone's here, but he can really lay into her. I've had to physically drag him off before. So I don't push it. If he's got something to distract him, like when Abby [sister] was here, that was fine because he would joke around with her. And now she's moved out and it's just us. But he's kind of
coming through that now. And I make sure she's in bed by eight, that's it, and Chris [brother] and I can spend the evening together and I try to do things for him as well. (SESL, P1, SIBS)

Mother 10: It's difficult for other people to be tolerant of the behaviour that he demonstrates than it is of the fact that he needs to watch what he eats. Then he feels excluded, then that simply exacerbates the problem because he desperately doesn't want to be excluded. *He is very hard to live with, for the girls in particular. He's to the point now that they don't want to play games with him. Because he doesn't behave properly. Sometimes it's nothing more than just speaking too loud or asking the same question over and over again.* So yes behaviour is definitely...it led to his exclusion from school and in the long term that will handicap him much more. The fact that he's reasonably intelligent is great, but it doesn't really matter if he can read and write and do maths very well, but he can't integrate into society and the behaviours around him. So if I could wave a wand and deal with any of it would be to help the behaviour. (SESH, P1, SIBS)

Figure 4.5 displays the emotionally turbulent relations between children with PWS and their siblings and its impact on the family. Matt (aged 11 years) depicts an argument with his sister (aged 8 years) and how they called on their mother to mediate the situation. It is interesting to note that he chooses to represent the scene as text rather than pictures, this was a common in write and draw sessions, reflecting children with PWS's interest in text and numbers as opposed to images. The text reads:

Sister 18 [aged 10]: Mum he's calling me stupid.
Matt: No, she's calling its me her. No she calling me pea-brain and she called me a pea-brain.
Mother 18: Stop it right now or you are grounded, ha ha!
Sister 18: But I don’t want to be grounded.
Mother 18: Well you are grounded for a month and a week. (SESL, P2, SIBS)
he's calling

me stupid.

no she calling. it's me he

no she calling

me for bran

and she call

me peabran

stop it right

now
The relationship between parent and child is often described as very intense:

Mother13: This is the third time she's been away from home with the school and she was away once with your Mum and Dad [to other parent] and that is the sum total really, apart from the odd overnight with Heather [family friend] as well. It was awful actually cos when she first went away, it was a shock to my system. In a sense it was awful cos I realised just how much I give her. I felt guilty.

K: Really?
Mother13: Yeah...in a sense because I was glad, cos it was lovely that she was away and I felt guilty about that. Suddenly my life was a lot, I was really quite lazy. So I'm looking forward to next week and I'm not going to feel guilty. I haven't said that to her. (SESL, P2, SIBS)

Parents often describe loneliness or feeling dehumanised as a result of their relationship with their child, particularly in response to the child not understanding the parent’s emotions. This is vividly illustrated in the following comments from Mother04:

Mother04: He just has no concept of other people’s needs or feelings. Which isn’t his fault; it’s all part of the syndrome isn’t it? But just on occasions I wish he’d realise that I’m not solely here for him and sometimes I need my own space too. He can be extremely affectionate, it’s not all bad. It’s just that he’s the centre of his universe and I can’t seem to teach him any different. It’s hard to live with that day in day out you know? (SESL, P1, SIBS)

These feelings of loneliness and isolation are exacerbated by lack of public understanding around PWS and the invisibility of the work that parents do. Thus:

Mother14: And it's really hard to get people to understand that. I used to get really narked about it. Perhaps I'm mellowing with age. But especially professionals I really couldn't understand, that they know that, but it doesn't matter. Even when we went to Wyvern for his first special needs assessment, with the head of special needs. They said does anyone want a drink and Nick says 'yes I want coffee' and they said 'here you are have some biscuits' and this is 9.30 in the morning, he's already had breakfast! They didn't need to offer biscuits. And he was very good because it was a pack of three. And I said “no” because I got all narky. And he had one and I said 'I'll keep those for you for when you get home’. But it's really; people don't understand that aspect of it at all. It's quite hard to find substitutes. Like you can get an Easter egg for a pound and if people have suddenly got to buy a present instead you're talking fifteen quid. Then Alice [sister] goes “I want fifteen pounds. (SESL, P2, SIBS)
We will return in more detail to this topic of isolation in chapter six, addressing to how it is experienced in relation to family structure.

4.7 Conclusion

Four key features emerged as central to managing the PWS body in the within the families. Firstly, ‘Access to Food’ strategies. Most households employed some physical form of denying access to food, either locks, food placing or food storage. Secondly, all families were seen to use ‘keeping occupied’ practices. Families where no physical restraints were used to block access to food described the lifestyle of their child as active and busy, with physical activity being the central to their lifestyle. This emphasis was also visible in many families that did deny access to food. ‘Keeping occupied’ practices within the home often took the form of simple, solo activities, such as jigsaws, cards, copying text, where the children were described as becoming absorbed for long periods of time. The third key practice was families’ ‘use of routine’. The majority of families found their child was most comfortable when they knew in advance what they would be eating and when. In some families, a very rigid eating structure had evolved; others made ‘gestures to routine’ which the child was seen to enjoy. The use of Human Growth Hormone formed the forth key feature in practical management. Families described the many advantageous physical effects of the drug and ways in which it had improved the quality of life for their child. They also spoke of the knock on effect this had in taking the pressure of food provision within the home. But others were more negative and had decided not to use hGH. The four themes in management displayed that managing the ‘body’ could not be abstracted from managing the person; food management was always linked to behaviour management beyond food seeking and stealing. Parental discussion of food management techniques throughout the data was embedded with moral as well as practical justifications for everyday action in the households.

The idea of ‘embodied action’ forms the theoretical starting point for understanding these behaviours/social actions. Within all of these management practices, the impact of emotion and biology is clear and everyday action is seen to stem from the relations
between embodied agents. The phenomenological idea of embodiment (Merleau-Ponty 1962) displays the role of the physical, emotional body in perceiving and relating to the social world. It also provides a theoretical basis which accommodates those individuals who have significantly differentiated physiological forms and apprehensions of their environment. As well as highlighting the role of biology and emotion, the data display the connectedness of the traditionally divided realms of the cognitive and the corporeal. Traits commonly associated to cognitive functioning are found to play a key role in controlling hunger, which is commonly seen as sensory. This connectedness points towards a need to redefine the nature of autistic spectrum cognition and hunger, by highlighting the social context and processes that surround them, thus removing them from biologically reductive abstraction.

In order to make these processes transparent, the idea of a network becomes useful. Within the families, we have seen that interactions between embodied agents are of primary importance to the creation of everyday actions. These relations are impacted by the physicality of the individuals and how they emotionally relate to their environment and each other. Closer inspection of relations between family members highlights the importance of the use of language. Although family members' relations with each other are central, the networks involved in the formulation of management strategies also stretch beyond the family members themselves. Families draw upon existing knowledge, materials and technologies to understand, justify and implement these strategies.

Sources of agency in the everyday management of the PWS body are therefore diverse, but embodied relations are central with a secondary network of the individuals' relation to knowledge and materiality. Within families, a micro process of embodied structuration can be isolated focusing on the PWS body. The networks described as creating behaviours and strategies, contain the PWS body. The PWS body acts out these behaviours and is the product of them (See figure 4.4).
CHAPTER 5: THE ROLE OF VALUES IN FAMILY MANAGEMENT OF PWS

Introduction

Having identified four key everyday practices in the family management of PWS we will now examine the core values within families which guide these practices. We have seen so far the importance of embodied emotion and perception, linked to the biology of the child with PWS, in directing practices around body management, as well as the use of materials in these practices. Following Burkitt’s (1999) theoretical position on the formation of social processes directing action, knowledge and cultural meanings, as well as, material factors play a part in these processes. As such we will examine in detail the beliefs and values of family members and how they are seen to impact the practices identified.

Four key values emerged in family accounts of the day to day management of PWS:

1. Physical health
2. Emotional health
3. Concerns for the future
4. Quality of life for all family members

After addressing the nature of values in these families generally, the chapter will examine each of these values in turn, specifically focusing on their relationship with management practices. It will then examine what these values indicate about family conceptions of the nature of disability and how it should be addressed.

5.1 Values guiding food management strategies

Within the sociology of childhood, families and the embodied interactions within them have long been conceived as powerful mediators between children and wider societal structures of knowledge and materiality, shaping children’s understandings and actions. As Dunn (1988) notes, children’s understandings of social relationships develop as such because of a pragmatic need to ‘get things done in their family relationships’ (Dunn in Christensen et al 2000: 25). Thus in modern Western societies families provide important environments, within which children develop social understandings.
Studies of childhood have moved towards separating the child from the family and allowing study of children in their own right. The placing of the child within the family context is often viewed as misrepresenting children's agency and limiting their voice within sociological research (James et al 1996: 41). However, in the case of children with PWS, the role of the family as a mediating level between child and society, is arguably heightened and magnified due to the intensity of the relationships between parent and child with PWS. A high level of child dependency on parents was observed and described throughout the research. The child's need for close monitoring and differentiated social understandings led to the family playing an extremely central role in regulating the child's actions and shaping relations to their social environment. This kind of dependency has been commented upon within various studies of disability and the family (see Hillyer 1993, Pillemer et al 1991).

This example from family 02 portrays the intensity apparent in the majority of parent/child relationships within the families:

[Finished session with child with PWS: Becky. Becky shouts through to kitchen where her mother is]
Becky: Mummy I've finished!
Mother02: That's good. Are you going to go and read now so that K and I can talk some more?
Becky: Yes, I, no, mummy? [Goes through to kitchen] I told her about school and Alison [friend at school] and then about riding. Also what books and...
Mother02: That's ok. I could hear you mostly, you did really well. Now I'm going to talk to K.
K: Yes, that was really useful, thank you.
Becky: [getting agitated, raised voice] But, this important, one thing. I told her about Sheppie [dog] when he went in the field with the cows and I got angry with him because he knows about that he shouldn't because I've told him. Is that OK, she said it would be alright to tell her that?
Mother02: [serious tone] Yes, that's fine, you don't need to tell me that, you did really well. Now can you go up and read your book for me?
Becky: How long?
Mother02: Half an hour.
Becky02: Ok [leaves]
Mother02: [lowered voice] That's typical, like what I was saying earlier. There's a constant need for reassurance about absolutely everything, you never know what she'll get stuck on next.

K: And there always has to be time for the story?
Mother02: Even when there isn't, you have to make it. She can be extremely demanding. It's just easier and actually less time consuming to go along with it. What she wants is right and right because she wants it [laughs]. (SESH, P2, NOSIBS)

Because of this intense and dependent relationship, where children with PWS frequently seek approval and instruction from their parents, the design of management strategies tends to come from a parental level. The child with PWS affects these strategies as we have seen in chapter four, with parents basing strategies around what works in practice. However the values concerned in guiding these practices were identified above all as the parental values. For this reason, the most relevant data for this chapter came from discussion with parents rather than with children.

Accounts of management strategies and practices were littered with justifications for everyday family actions, both practical and moral. These justifications were often based on everyday interactions within the family, however they also made reference to wider cultural health beliefs, either from media and government health promotion, medical commentary on PWS and advice and literature from PWSA (UK). Parental biography i.e. experience in their own family was also seen to affect conceptions of health and thus guide values, as described in other research on the family and health (see Lawton 2003). This will be examined in more detail in chapter six in relation to social class.

Bearing in mind the powerful mediating effect of the family on the understandings and behaviours of children (Rosser 2003, David 2003, Dunn 1988), it is useful to identify the core values within families that guide their management of children with PWS. Management practices can be conceived as the result of embodied family interactions, with factors external to the family such as technologies, medical
knowledge on PWS and wider social beliefs about health playing a part in this process. The analysis revealed a set of core values or priorities at the family level which played a key role in guiding everyday practices in households. As the beliefs of the family members, these values are part of embodied interactions and can be seen to draw on the wider discourse identified above.

Van Hooren et al (2005) conducted qualitative interviews with the parents and caregivers of 18 children with PWS in the Netherlands. They concluded that the families' main concern was to create a life that was 'meaningful' for all members of the household. This was facilitated by creating an atmosphere of trust and allowing the young person with PWS to take responsibility and make their own choices where possible. They characterised four values which directed behaviours managing the child. These were: 'Physical health, well-being, freedom and responsibility, and a liveable life' (van Hooren et al 2005: 309).

Van Hooren et al's analysis is compatible in certain ways with the image of PWS management emerging from my own data. Chiefly, their emphasis on 'a meaningful life' indicates that the subjective and emotive relations between family members are central to creating everyday actions and strategies around the person with PWS. Although expressed in terms of gender and emotion work rather than 'embodied relations', Hochschild also notes the importance of emotional balance amongst family members, particularly women:

Hochschild argues that there is a third shift of emotional work that women disproportionately perform. This refers to attempting to maintain harmony and orderliness in the home, comfort children and spouse, and generally ensure that the home provides a haven from the difficulties of the outside world. (Adams et al 2001: 34)

Analysis identified four family values which were central to decision making around and reflection upon, food management practices. These values were the underlying priorities across all of the families. In many cases these were highlighted in justifications and explanations of effectiveness. They were also sometimes highlighted through concerns. Everyday life was presented as allowing these values to
be upheld or viewed as problematic when practicalities prevent behaviours that support these values. In the analysis that follows, values will be traced through to the behaviours which they have been seen to affect (see figure 5.1). The discussion begins with a consideration of the value of physical health and the care of the body.

The values identified: physical health, emotional health, considerations for future and quality of life for all family members, display similarities to those of van Hooren et al (2005). However, as well as identifying key values, analysis uncovered sub values within these, such as social inclusion, independence and flexibility. As in van Hooren et al's (2005) findings, values were not always fully achieved due to their contradictory nature. For instance, strategies used to control 'physical and emotional health', such as use of rigid routine, were judged by many parents to impair the achievement of values such as 'considerations for the future'. Adding to the work of van Hooren et al (2005) this chapter not only identifies values and sub values, but links them to the practices described in chapter four, using empirical evidence to demonstrate where practices are seen to aid or impede the achievement of values.
Figure 5.1 - Family Values and Practices

**Value and sub-values identified**

**Physical Health (child with PWS)**
- Weight loss/maintenance
- Other physical/medical conditions

**Emotional Health (child with PWS)**
- Inclusion
- Identity/self image
- Various conceptualisations of emotional health

**Considerations for the future**
- Inclusion
- Flexibility
- Independence

**Quality of life for other family members**
- Emotional balance/harmony
- Freedom and choice for other siblings
- Inclusion

**Practices**
- Access to food
- Keeping occupied
- Use of Routine
- Biological alteration

**Key**
- Value guides management practice
- Management practice impairs achievement of value

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5.2 Physical health and care of the body

The themes of managing weight and the body were the initial focus of the data analysis. As such, physical health and the body emerged as the first key priority in food management practices within the families. Many parents expressed the idea that, despite the effects of PWS on their child, the child was generally in good physical health. Even in the cases of obesity or scoliosis parents made statements to this effect, as if health could be measured in separation from these physical conditions.

Weight control was the central theme in issues relating to physical health, although a number of other physical conditions such as skin picking, related infections and scoliosis were described as everyday concerns guiding management behaviours. In cases where the child with PWS was overweight, parents tended not to see this as a major health concern, expressing a view that excess weight was the result of the medical condition and to some extent, to be expected:

Mother20: She [child with PWS] is a bit chunky, especially around the middle, that’s where she carries it. But that’s normal for the condition, she’s always been that shape. (SESH, P2, NOSIBS)

As reported in a Scottish study of perceptions of body size amongst young teenagers (Wills et al 2002: 399), most families used non-medical, descriptive terms talking about weight. For example:

Mother04: She does have a bulky look, it’s the lack of muscle tone.

In most cases day-to-day management strategies were seen as preventing the risk of extreme obesity, which was seen as the potential risk of PWS, maintaining body weight even where children were slightly over-weight rather than correcting an excess weight problem.

In only two cases did the child themselves talk about wanting to lose weight. In these cases, the act of losing weight was not connected to health or altered body size by the
children and, as seen in this quote from a 13-year old with PWS, weight is spoken about in numbers rather than bodily descriptions:

Simon: [showing K weight management diary, recording weight every week over past two years] In April it [weight] was nine and seven [9 stone 7 pounds], then ten [stone] and now it's nine- eleven [9 stone 11 pounds].

K: So you lost some weight recently, well done. Were you happy about that?

Simon: Yes

K: What do you enjoy about having lost weight?

Simon: [blank]

K: I mean, what was it that made you want to lose weight?

Simon: I keep the weight book [diary] and Mum weighs me and we write down every week. (SESH, P2, SIBS)

A general lack of self-awareness and concern around the body was evident in the data from children with PWS. As well as not connecting issues of weight loss to physical changes in their bodies, an absence of general care of the body was commented on. As this sister remarks:

Sister13 [aged 7]: I like her because she makes me laugh.

K: When did she last make you laugh?

Sister13: We go to school, and she comes down [stairs] and her hair is all messy, cos she forgots to brush it. (SESL, P2, SIBS)

Although weight tends not to be conceptualised by the children with PWS in relation to their own bodies, there is evidence that children with PWS connect food to health, as displayed in figure 5.2. The child was asked to draw or write down some things (food not specified) that are healthy and unhealthy; figure 5.2 also suggests the preoccupation with food described in chapter four.
Having discussed approaches to physical health in terms of body management within the families, we will now examine how these are connected to the management practices. Where access to food was physically denied, straightforward connections to weight control were made. By physically restricting food intake, diet could be monitored more accurately by parents and weight more easily controlled. Thus:

Mother09: We have two [locks]. That one on the pantry and one on the kitchen door that we use at night.

K: Is that something you’ve always done?

Mother09: It was just that cupboard until about three years ago. When we realised we had a problem with food at night, because he has times when he doesn’t sleep that well. You know you’d just notice things had moved and he was looking a bit bigger too. It gives us peace of mind and we can watch his weight that bit better with the lock on the door. (SESL, P2, SIIBS)

Within two families, locks had always been in place in preparation for food seeking behaviour rather than as a reaction to weight gain. These families linked this to the portrayal of PWS in the initial information they were given by medical professionals or the PWSA charity. Interestingly, both of these families expressed a view that food was not a problem and that weight was not a major concern for them. The failure to acknowledge the value of physical health perhaps indicates how longstanding and ingrained into the lives of families with PWS this value is. This is displayed in family 17, who use locks but contrastingly report that their child doesn’t have much of an issue with food:

Mother17: No I don’t worry about it really, food isn’t really an issue for Vick, it’s much more behaviour that poses the real problem. I think it’s because from day one we knew about the eating disorder side of things and we prepared ourselves. Because that’s the main thing that comes across in the advice you’re given and everything points towards that. Yes what didn’t come across was all the behaviours and how
challenging they can be. Unfortunately Victor's got a lot of those behaviour aspects and not so much the food.

K: Have you ever thought about taking the locks away?
Mother17: Yes, we tried it once and we did have problems, but it's hard to say how much of that is because he's used become used to them, so when they came off obviously it was a big deal. (SESH, P1, SIBS)

'Keeping occupied' practices were also clearly linked to the objective of physical health. This was seen most strongly in the three families who used no physical restraints to food but based their management strategy around having active lifestyles. This emphasis on the physical body is clearly visible in the detailed account of a parent from one of these families:

Mother01: As a biologist obviously the first thing I did was read up on all the genetics, because I was quite interested in it. So I looked at it and I thought 'right, ok so I've got this sleepy, dopey little blob of jelly...that's going to eat like a horse'. So how do we go about working against all the things that are going to happen? So one of the things I did was decided that he shouldn't be allowed to sleep all of the time, he was only allowed to sleep at certain times for so long then that's it, he's woken up. Even when we're in the car we like to keep him alert, so he doesn't sleep in the car. The other thing is exercise, when he was young, I don't really go in for massage, but just generally moving him around, that then developed the muscle, that's one of the things that I think is really important. Because of the 'we will sue' policy, a lot of the physiotherapists, when you tell them the kinds of things you'd like them to do, if there's a lot of movement, they say no. So we were very lucky in having a very old physio. I said to her I want him to go in one of these baby bouncers, and she said 'wait 'til he can hold his head up, because you have to be really careful with scoliosis. Now he was floppy but he could just about hold his head up and as he used the bouncer, he'd move around and when he came out he was rigid... And although we spoke about scoliosis, my attitude was that if you're a blob and you have no muscle, you're going to get scoliosis anyway. The only way you're going to stop scoliosis is by building muscle around the spinal chord. So it depends what way you look at really, as long as you've got a child who is capable of developing muscle and you're not doing a
ridiculous amount of exercise at a young age, then you should be able to produce the muscle without scoliosis occurring. We've found this to be really successful, but the professionals won't to it, they won't give that advice...it's so ridiculous. And when you think of how metabolic rate increases with exercise, so that someone who is fit will naturally be able to eat more without putting on weight. Again if you keep a Prader-Willi child on a starvation diet, ok so it might keep the weight down, but it's not the way to promote health or anything else. If you increase the exercise, increase the metabolic rate, increase the amount of calories being burnt, they can eat more food, this has got to be beneficial to health and also they can lead a normal life. (SESH, P2, SIBS)

This account displays that the value of physical health stretches beyond simple weight control. Scoliosis (pronounced curvature of the spine) entered into several families' concerns about physical health throughout the research, since all children with PWS are at risk of this condition. Twelve of the twenty children in the sample suffered from scoliosis, in three cases this was severe and the children wore body brace supports.

'Keeping occupied' practices (discussed in chapter four) were described as having positive effects on physical health by either exercising the body, distracting the child from food or both simultaneously. However, concerns about physical health were also raised in connection to 'keeping occupied' behaviours. These concerns were mostly based around the repetitive activities in the home. However, one parent voiced concern for physical health related to swimming. The account illustrates again the many non-weight related conditions within PWS and their significance in everyday life:

Mother 17: He does [swim], yes it’s one of the only forms of exercise he doesn’t really moan about. But I do limit how many times a week he goes because of his eyes [sufferers from eye irritations] and also he’s got a scab at the moment that he won’t leave and the last thing we need is for that to get infected too. So it’s just once a week at the moment. (SESH, P1, SIBS)
Skin picking was also a concern for some parents during ‘keeping occupied’ practices within the home:

Mother09: Another thing that can be a worry is his skin picking. We’re seeing a psychologist next month, so I’m going to ask her the best ways to deal with it I think. It changes places but at the moment he’s scratching the back of his leg, it’s really quite bad. I try to keep an eye on him, but you can’t all the time. I’m pretty sure he does it when he’s up there with the X-box after school, I’m not even sure he’s aware he’s doing it half the time. (SESL, P2, SIBS)

Scoliosis was another aspect of physical health, thought to be worsened by activities such as prolonged computer use within ‘keeping occupied’ practices:

Mother15: Yes, I am trying to discourage it a bit at the moment [computer use]. Well it’s not something I’m happy about anyway but also his GP has pointed out a mild scoliosis over the last year. So yes, I think the computer’s going to have to be limited to certain times, which he won’t be pleased about. (SESH, P2, SIBS)

Within the ‘use of routine’ practices, physical health is achieved through weight control, as described below:

Mother06: Yes, it is good in a way that he sticks to a fairly set pattern [diet]. It’s very easy for me to monitor his calorie intake that way
(SESH, P1, NOSIBS)

Many families view the effects of hGH as having a great positive impact on physical health. In chapter four, we saw that parents’ accounts associated extra energy levels, improved physical abilities and muscle tone with hGH, and described the easing effect this had on everyday food management. For example:

Mother03: I mean she has grown so much since she’s been on it [hGH]. We looked at a picture of her the other day didn’t we?
Father03: Oh as a young girl, yeah [child’s current age is 14].
Mother03: You're nearly as tall as me now aren't you? And her muscle tone is much better.

Father03: And you could see as soon as she went on it, in the space of just a couple of weeks, she was doing so much more. She wasn't feeling tired no more, she was up and energetic. And even the way she used to run, it used to be such a funny run, but in the space of a couple of weeks she was lifting her legs up to run properly. So you did notice a big difference straight away.

Mother03: They do, they run so funny, we used to say she run like an elephant, so heavy footed. But she's so much better, and she finds it a lot easier now. (SESL, P2, NOSIBS).

Scoliosis featured again in concerns over physical health, in relation to the use of hGH:

Mother05: He's got scoliosis quite bad and he has to have an operation on his back, lined up in 6 weeks, to have a rod put in his spine. And that's been monitored for the last 7 years. He had dislocated hips when he was born and he was in a harness. I guess he was 2 or 3 when his back was monitored and it's got worse and worse. So yes there are other things. Hopefully in 6 weeks time that will sort it, every year it causes him more and more discomfort.

Father05: And he's very hunched.

Mother05: Yeah...it used to be just when he didn't have a top on you could see it, but now with any clothes on.

Father05: They spoke about a brace, but it just wasn't going to be practical. One, I think long-term they knew he would have to have an operation anyway, and the discomfort and restriction that a brace would put on just wasn't going to affect it. And doesn't even like wearing his glasses, so there's absolutely no chance of him wearing a brace 2 or 3 hours a day. And for something that would ultimately be of little benefit because he'd have to have the operation anyway.

Mother05: At the moment his back is really stopping him doing thing that he possibly could and making progress. He can't lie on his back very well.

Father05: He's obviously been going to clinics for a number of years. But we only got the letter this week anyway. And because of the way he is if we tell him now,
everyday it would be “when's my operation”. And he'll get himself really worked up or get us worked up. It's like anything with him, you need to narrow the timescale between telling him and the event.

Mother05: It even tells you in the letter. It advises you not to tell the child until the day before hand.

Father05: Yeah so that’s our main thing. He had growth hormone for 1 year.

K: Did you notice a difference?

Mother05: I noticed he has sprouted quite a lot.

Father05: It wasn't instant, but over the year he has grown more than he would have done. And he's obviously still of quite short stature. The down side is, effectively it's made his scoliosis worse because he's growing faster than normal and it's affecting that. (SES H, P2, SIBS)

Issues around physical health and hGH are seen to be complex by parents. The above except from an interview with family 05 illustrates a family trying to balance two physical health concerns. Where the family are using hGH it is noticeably worsening scoliosis and causing the child pain. However, in this case, as the child is due to have major surgery for the scoliosis anyway, the benefits are seen to outweigh the downsides. Physical health was also described as the key motivating factor for non-use of hGH. Three families who did not use hGH saw it as too risky since in their opinion not enough is known about the long term effect on the body. All of this points towards physical health being a major issue in decisions around use of hGH.

The value of physical health and care of the body was evident throughout the four trends in food management discussed in chapter four i.e. access to food, keeping occupied, use of routine and biological alteration. Weight was an obvious consideration within families, but the data reveal the importance of other physical conditions associated with PWS. These include scoliosis, eye infection, skin picking and wound care, and the risk of unknown physical conditions in relation to hGH.

5.3 Emotional health

A second key value in families was the emotional health of the child with PWS. This was a major concern within the families, with thirteen of twenty stating that
'behavioural problems' such as uncontrollable outburst of emotion were the thing that worried them most about their child. Accounts of food management made reference to emotional health as a factor that guided family’s everyday practices, but also as a concern. Emotional health was conceptualised in a variety of ways, however behaviour such as violent/angry outbursts or obsessive traits were commonly seen as the key indicator of emotional health problems.

Around ‘access to food’ strategies parental concern for their child’s emotional well being on a day-to-day level was seen as central to the formulation of locking practices. Parents described children getting ‘anxious’ and ‘preoccupied’ where there is opportunity to take food. Locks were placed not only to stop physical access to food, but also to relieve these stressful situations for the child:

Mother16: Food is always going to be a problem; he just gets really wound up if he knows there’s food there [accessible].
K: So do you tend to use the locks all the time then?
Mother16: Yeah, it’s the best way and it’s easier for him as well as me.
K: Is it kinder to him?
Mother16: I think so, otherwise the whole time he’s wound up [about food]. (SESL, P1, NOSIBS)

Many conceptualisations of emotional health can be seen within parental accounts of ‘keeping occupied’ practices. In families where ‘active lifestyle’ is seen as the central management strategy, a holistic view to health is taken; where physical health impacts emotional health and vice versa. Within this research all families who used a general ‘active lifestyle’ as a body management strategy also displayed a holistic view of health. These families express the view that keeping the body in good condition will have knock on positive emotional effects. They also display the belief that keeping mentally engaged in activity is an emotionally healthy way of life:

Father20: She’s always enjoyed riding and so have I. We had to stop for a year when the stable shut down…yes we really missed it, being outdoors and focusing on the
activity, on achieving that relationship with the horse, it’s a very beneficial to her I think. Well, it’s just a lovely thing to do for anyone really. (SESH, P2, NOSIBS)

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Mother01: We taught him about keeping active from a very early age, we never left him alone to get bored and we did whatever it took to get him involved in what was going on around him. Puzzles were great we’ve got literally bookcases full of puzzles. So even if he can't always be doing physical activity, he can keep his brain active. I think even with normal children, if you just stick them in front of the TV and expect them to entertain themselves, there’s no wonder children are becoming obese and square-eyed and all the rest of it. (SESH, P2, SIBS)

Further to this, families where children are focused on one particular sport interpret this as contributing to the well being of the child by giving them a sense of identity, achievement and pride:

Mother01: He's got this gala coming up and it's hard, he knows there's a lot of competition, but he's absolutely determined that he will enter every competition and try to win it. It really is his thing, his focus, he gets an awful lot out of it.

K: Do you think that’s when he’s happiest?

Mother01: I would say so, when he’s competing he’s really in his element, it’s good to see. (SESH, P2, SIBS)

In chapter four, ‘keeping occupied’ practices involving focused, simple, repetitive tasks were seen to occur to some extent naturally; that is, whether the parent(s) were aware of their use as a food management measure or not. This indicates that they are activities that are pleasing for the child. The children’s enjoyment of these activities was also highlighted in parent accounts:

Mother09: He's quite content. I'd rather see him happy than stressing about what will be for dinner or constantly asking me questions. It's nice for everyone to have some peace sometimes. [pauses] No it's just one of his quirks [writing and copying lists], but it doesn't hurt anyone and he's happy. (SESL, P2, SIBS)
However, in cases where these activities take extreme forms and begin to be seen as obsessions, parents express concerns over negative impacts on emotional health, as seen in the following excerpt:

Mother10: Yes it’s very strange and I don’t know why he does it [hoarding paper and articles, carrying them round with him]. We saw a psychologist but that was of limited use I thought. It seems with Joe he will just have these focuses, go in and out of phases and unfortunately there’s very little anyone can do to stop it. (SESH, P1, SIBS)

Where a strict routine of eating was followed by families the main justifications were the problematic behaviour which occurred if the routine was broken and the widely documented pleasure the child took in having certainty about what and when they would be eating. This highlights that emotional health of the child, was the central value involved in decisions around ‘use of routine’ as a strategy. Family 13 describe the emotional security that they believe routine gives their child:

Mother13: No, food isn’t a concern anymore. John’s quite rigid in that he expects to have breakfast, lunch and dinner.
Father13: Yeah...and if we’re out and about and things don’t go to plan. And we might be saying we’ll have breakfast then a late dinner, then he’ll want to have at least something, as a token, for lunch.
Mother13: Absolutely and he would normally expect to have a sweet.
Father13: yeah...but he doesn't always. Like I didn't make one today did I?
Mother13: He definitely likes the security of knowing what and when the next meal is going to be.
Father13: Which is fair enough though isn't it, we’re glad to meet him half way. (SESL, P2, SIBS)

The ‘gestures to routine’ in families with less rigid eating patterns are also done as a special treat for the child in acknowledgement of the function of these gestures to improve the quality of life for the child. As family 10 tellingly observe: ‘we have sausages on Friday...which he loves to look forward to’.
The fourth management practice, biological alteration was also justified by parents in relation to the value of emotional health. The use of hGH was seen as beneficial to the child not only through its physical effects but also through the improvements to quality of life and self-image achieved as a result. Inclusion was central to this process as families noted the importance to the child of being able to be included and treated equally in family meals and being able to match peers sporting ability. The positive significance of sporting achievements to identity and self-image were again highlighted as improving emotional health, as recorded in chapter four.

Within families who did not use hGH, emotional health also emerged as a central deciding principle (see comments by families 4 and 15 in biological alteration section, chapter four). These families described their decision not to use hGH as part of an effort to allow their children to accept themselves as they are.

The analysis therefore suggests that emotional health is a core value within the families, guiding practices around food management. Improving self-image, building personal identity and inclusion with family/peers were identified as conditions that aid positive emotional health. There is a significant body of literature within the sociology of health and illness which traces lay conceptions of health (e.g Blaxter 2004, 48-53, Nettleton 1998: 36-68, Conway et al 1998). Within accounts surrounding family practices and justifications for them, a number of conceptualisations of ‘emotional health’ were displayed. These included emotional health as: a holistic approach- physical health was equated to emotional health inseparably; constant engagement in mental activity as well as physical activity; the absence of problem behaviour (obsessive behaviours and angry outbursts); and self-acceptance. As observed by D’Houtard (1986), who conducted a large-scale social survey of the health beliefs of the French population, these beliefs were clearly influenced by the life, biography and everyday experiences of those who held them. Chapter six will examine in more detail how these core values and the associated conceptualisations of health are distributed amongst the families and related to their socio-economic and cultural positions.
As embodiment and the essential connectedness of the physical body and emotional body have been found to be central in the analysis of the behaviour of children with PWS in this study, it is interesting to see that physical and emotional health are apparently categorised separately in family accounts, particularly when being asked about priorities directly. When asked, ‘is there an aspect of life with your child which concerns you most?’ families tended to separate perceived physical and emotional factors: thirteen of twenty indicated that the aspect that concerned them most was behaviour; five weight loss; and one scoliosis. One family did however articulate a belief that all aspects of their child’s health were related:

K: Is there any particular aspect of life with Becky which concerns you most?
Mother02: It's the whole package, you can't separate, you have to take her as an individual, as a person and every facet is important. I mean socially she acts like a toddler and at that time that becomes really important to focus on, and other times would be she's telling lies to everybody, you know. All the things change all the time and I think it's part of her ability to manipulate so well. She can almost change tack from one problem to another. You think you've just got on top of something and you shift the attention to something else. But she was dreadful at self-harming, and that has improved, but it hasn't gone.

(SESH, P2, NOSIBS)

The above excerpt from an interview with family 02, evidences the view that physical, emotional, and behavioural aspects of the disorder are connected. It points to the dynamic, changeable nature of concerns and symptoms within the disorder and the constant interplay between social and biological factors, charting events at school and the child’s emotive response and perception of them. Even where families initially distinguish between physical and emotional issues, many of the processes they go on to describe, such as the effects of hGH, clearly indicate the synergy of physical and emotional factors.

The families’ tendency to compartmentalise physical and emotional management goals may reflect how ingrained into western thought and language the dualism between mind/body remains. However some data suggests that this separation may be
symptomatic of the potentially conflicting requirements which managing PWS involves. For instance, where food treats were allowed in order to avoid emotional outbursts in public places, parents were involved in a direct play-off between physical and emotional risks. However, not all families acknowledged the physical and emotional aspects of management being at odds with each other. In families where physical activities formed a core part of the management of PWS, they acted as a distraction from food and the active lifestyle was viewed as being simultaneously beneficial to physical and emotional management. However the everyday relationship between physical and emotional aspects of the disorder were conceptualised by parents, the data clearly displays the breadth of considerations taken on a day-to-day level and the importance of negotiating emotion in order to manage the body.

Within the data the ‘happiness’ of the child with PWS emerged as a key priority, guiding decisions around the everyday management of the disorder. However, these notions of happiness can be seen to shift and change, particularly in terms of short and long term happiness. Many measures that act to enhance the child’s immediate mental health, e.g. allowing them to indulge in obsessive behaviours, cause parents concern in light of the child’s long-term happiness. The following section considers how children with PWS are prepared for life as adults, identifying the values that parents feel are important to this transition.

5.4 Considerations for future
Considerations for the future form the third key value in managing PWS. Transitions from childhood and adolescence to adulthood can be problematic for those with PWS and their carers (Whitman in Butler et al 2006: 122). Traits associated with childhood are displayed throughout the life course i.e. intellectual ability, child-like speech, and adults with PWS are legally and socially viewed as a vulnerable group. The majority of adults with PWS remain within the family home (Waters 1996: 34), with others moving into various forms of residential living. As such, considerations for the future happiness of their children with PWS are a key area of concern for many parents. This was clearly displayed within the families through parental explanations of decisions around various food management practices.
Within ‘access to food’ strategies, whether access was denied or not, many families were seen to privilege internal control. One way parents accounted for this was by stressing the importance of teaching independence/autonomy so that the child can have more choices as an adult. Independence forms a key value within concerns for the future more broadly, see figure 5.1. This is demonstrated in the following interview excerpt:

Mother16: The problems come when we go out mostly. Because here [home] we’ve [mother and son] everything set up [locks]. It was, I did worry that because I run things so tightly with the food it would be hard for Paul to ever move away from home or to have a job, it is a worry. I haven’t given that much thought to it yet [Paul aged 12 at time of interview].

K: Would you ideally like Paul to live independently, move away from home at least?
Mother16: Yes, just like with any child. He is intelligent enough to have some sort of job, but you can’t help thinking everything else is going to get in the way, the behaviour side. That would be a shame. Although I did speak to someone at PWSA and there are homes which run the food as tightly as me, it’s just getting the funding [funding to get Paul into one of the homes]. (SESL, P1, NOSIBS)

This theme of training the child for future independence also arose in justifications of ‘keeping occupied’ practices. The following parent in family 01 spoke about socialising or conditioning her child not to be preoccupied with food, with the objective of making them an inclusive member of society:

Mother01: I know when he gets home it’s not food that’s on his mind. Because he usually has a club or something to be getting on with. That’s always been very important to me, I’ve always taught Alex to get involved. There is an obsession [food] there I won’t deny it. But we’ve taught him a way of life where it’s not always about food and hopefully that will always be with him. When you read some of the medical articles they paint it as worse than it actually has to be in my opinion. The only way you can help someone to live a normal life is to treat them normally, at the end what you’re doing is trying to teach them to fit in. (SESH, P2, SIBS)
Where the routine desired by the child was resisted by the family, a reason commonly given was the value of teaching ‘flexibility’ in order for the child to be able to deal with situations better in the future. Thus:

Mother11: She needs to get used to get used to things changing. Everything isn’t always just how you like it…that’s life isn’t it? (SESL, P2, SIBS)

Similarly, where a rigid routine is kept, parents express concern for the child’s ability to eventually move away from the family, integrate and achieve independence:

Mother16: everything’s [food provision] as so and it’s the way he’s used to. So no, it’s [food] not a major worry. I do wonder how he’ll cope when he’s older or will there always be someone to sort everything out for him? It’s hard because he’s almost 14 and he still has the behaviour of a toddler sometimes. I don’t think we encourage that, it’s just hard to imagine him getting on in a different home. I don’t think he’ll ever be independent and that’s something we’ve accepted really. (SESL, P1, NOSIBS)

Choices around the use of hGH were heavily influenced by perceived positive and negative impacts on the child’s future quality of life. Resistance to hGH treatment due to unknown long-term effects reveals parental concern for the future. Where the effects of hGH are viewed in a positive light, they are also seen as an investment for the future. Again parents made particular reference to social inclusion:

Mother13: He will be taller than he would have been [without hGH] and the energy and strength that has a knock on effect. It means he’s much more able. The stronger he is the more exercise he can do and that makes he even stronger, it’s a cycle. (SESL, P2, SIBS)

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Mother03: …she can do so much more [on hGH]. We thought that’s so much better for her, she can live that bit fuller and she’ll have more choices at the end of the day. (SESL, P2, NOSIBS)
Considerations for the future impacted upon the four key trends in practical food management. Perceptions of the qualities the young people with PWS would need in their adult lives guided management strategies. Where everyday practices such as locking and strict use of routine were seen to impair future quality of life, this was raised as a concern. Several concepts emerged as central to parental perceptions of successful future living: inclusion, flexibility and independence. Importance was given to the child being able to fit into various environments and situations, thus increasing their ability to cope with the choices available and to be made in the wider context of adult life.

5.5 Quality of life for other family members

The quality of life of other family members forms the fourth key value in family management of PWS. As our theoretical analysis so far places relations between embodied family members as central to actions within the family, it follows that parents also explicitly highlighted this value and its effect on management practices. Although this was visible in many forms throughout parental accounts, the relations between siblings, including frequent tensions and arguments, as described by children and documented in chapter four, also play key role in directing everyday practices. Here again it is useful to draw on Hochschild’s (1997) idea of the parent working to maintain an emotional balance within the household.

Incidents between siblings and the perceived needs of non-PWS children play a major role in guiding management strategies. Aspects of food management practices were often viewed as having some negative impact on other children in the families. This was particularly the case where everyday structures and practices were heavily based around the young person with PWS, i.e. rigid eating routines or heavy usage of external restraints to food. The practicalities of denying access to food were often seen as unfair to brothers and sisters. For example, the parents in family 02 expressed the difficulty of finding a balance with access to food:

Father19: Before we moved down here, when I was working, it was very much the case that Hillary [mother] was the full-time carer for Sonia. I was only home at usual father times, seven to ten at night you know? Here, now, we tend to do it all. Share the
supervision, check food intake, it takes the pressure off of both of us. Food, we are
getting more and more concerned about. We are finding more and more theft and
finding something on the floor in the kitchen that, funnily enough, is nowhere near
where it should be. We're looking at various ways in which we can cope with it. Also
having two other kids who deserve to be able to come in and have a chocolate biscuit
or whatever as they please. (SESH, P2, SIBS)

This extract from a family that uses locks on kitchen cupboards, displays the moral
difficulties encountered by parents in providing for children with different needs. It
also points towards problems encountered with parents occupying traditional gender
roles. That is, with two other children, being a ‘full-time carer’ for the child with PWS
and running the household day-to-day was seen as too much for the mother, so the
father became more active within the household. The father of family 19 had changed
career and at the time of interview both the mother and father ran a business from
home and took equal responsibility for childcare. Family structure and division of
labour within the home will be discussed in more detail in chapter six.

As well as the practical and ethical aspects of locking and other access to food
restrictions, parents also displayed a concern for the impact on other children of
growing up in a household with such a strong emphasis on food. One mother with two
non PWS daughters aged seven and nine articulated this concern:

Mother10: It does bother me sometimes that the girls are growing up around all these
strict diets and locked kitchens and weight loss stuff. I think sometimes they get the
wrong idea. Emma [daughter seven years old] has said to me “I shouldn’t eat that
should I Mummy or I don’t want too many calories” and I am worried for them
because it’s hard enough for young women these days anyway and I would hate for
this to add to that. Yes, that is something I’m keeping a close eye on.

The same mother also went on to discuss how she counters this:

Mother10: Like when he goes out this afternoon [taken swimming by respite worker],
all the locks will come off and we’ll open up all the doors and we’ll probably do some
baking. If he's not here for the evening we sometime have crisps and treats. Just to show it's OK for us to do sometimes really. So as not to scare them about food and calories. (SESH, P1, SIBS)

Five other families described similar practices. For example:

Mother18: Well on Thursdays it's his drumming evening and we actually, we have fish and chips quite regularly and the others look forward to that. I think it's those things that help the others [three children without PWS] put up with the sort of, everyday demands of having a PW brother.

K: Do they find it hard do you think?

Mother18: Oh yes. At school it's very hard, they do stick up for him and look out for him though. No, he is extremely difficult to be with sometimes. They all have to keep their money and valuables locked in boxes under their beds, they can't eat in front of him apart from meal times and despite all that he still picks a fight with them when things aren't going his way. I do as much as I can for them when I can because he takes up so much of my time. They are very good. (SESL, P2, SIBS)

'Keeping occupied practices' were seen as giving other family members 'peace' from behaviours which can aggravate relations between the child with PWS and other family members. This is portrayed in an earlier quote from Mother09, page 149. In relation to 'keeping occupied practices she states 'It's nice for everyone to have some peace sometimes'.

The main reason given for resisting rigid routine within the families was that it was not achievable or practical within the demands of family life. Children and parents had work or leisure commitments which were high priority and everyday eating routines had to be shaped in consideration of these commitments. As the following excerpt from an interview with family 12 shows, families also valued being able to attend social or extended family events even where they sometimes posed problems around food monitoring:
Mother12: Well it varies. It's probably weekends and one day in the week we all eat together. Then the others it's either Gary's [husband] out or one of the kids has got a club or swimming. But yeah, it just depends on how things are going. We do make exceptions on special occasions, family do's. We let him have a bit more than he normally would and then we make up for it later. (SESH, P2, SIBS)

The extent to which other children partake in the everyday monitoring of food intake came across strongly through family observation, interviews with siblings and parental accounts. This sibling gives an account of his role:

K: Can you think of any ways that you help Hannah. Either at school or at home?
Brother08 [aged 10]: I do watch if I see her taking something from the kitchen. I have to go and tell mum, and ask her whether she’s allowed to or not.
K: Is she normally allowed?
Brother08: Sometimes. But I have to do it carefully and not accuse because she gets really angry sometimes.
K: Do you let Mum deal with that part?
Brother08: Yeah. I just keep look-out and make sure she doesn’t see me. (SESL, PI, SIBS)

Families who used hGH often described consequential benefits to the whole family. Improvements in behaviour and families being able to all eat the same things or partake in activities together were all seen as positive effects stemming from use of hGH. As well as again highlighting the importance of inclusion in everyday family life, families also described a general relaxing of food management and monitoring allowed by the drug and the gain to all family members. For example:

Mother19: There’s less tension there. They [other children] know if she eats something it’s not the end of the world anymore, it all affects them, sometimes you forget how much. They’re ever so good, they will play with her sometimes even if it’s not what they want and they’ve always looked out for her with the food. I tried to teach them: it’s not telling tales, it’s part of being a good brother. (SESH, P2, SIBS)
The idea of emotional balance within the family was a major concern, guiding actions within these families. This is evident in terms of a balance of responsibility and work within the marital relationship and a balance between siblings through avoidance of arguments. In the majority of families, emphasis was placed on all family members living full lives in terms of having choices and interests, this sentiment being particularly strong in families with other children. The child with PWS being able to partake in activities alongside the rest of the family was seen as beneficial to the family as a whole. Where strict management strategies were often seen to inhibit choices and freedom for other family members, concern was also expressed about an overemphasis on food and the effects of this on siblings, particularly where younger and female. Consideration of all family members guided practices, enabling family members to partake in social and sporting activities. Families with an only child with PWS found this balance easier to strike, this will be discussed in more detail in chapter six.

5.6 Family values and the nature of disability

Having examined the four family values we will now address what these values suggest about parents’ conceptions of disability more generally. The complexity of hunger; as involving social/biological synergy was discussed in relation to sociological literatures and parental perceptions. In a similar way, the family values guiding everyday practices and actions within the households cast light on the nature of disability. Of the four values identified in connection with managing the physical body, only one was explicitly concerned with the physical body, the other three values addressed social aspects of living with PWS. That is, how children’s ‘impairments’ or biologically determined attributes have simultaneous social impacts upon the child and the family. This allows disability to be viewed in line with the social model perspective, where the biological impairment (PWS) is the basis upon which disability is based, and disability refers to the individual’s experience of social exclusion (Oliver in Barnes et al 1996:139-54).

‘Emotional health’ was conceptualised in various ways, but was centrally concerned with the child’s relation to social structures such as the family, educational and recreational settings. This relation was highlighted in the sub categories of ‘inclusion’
and 'identity' assigned to this value. Within 'considerations for the future', there was also an emphasis on social inclusion, with attention being given to the nature of residential provision. 'Quality of life for other family members' addresses the more immediate social structure of the family and stresses the benefits of the inclusion of the child with PWS within family life, but also frequently made reference to wider structures of employment, education and leisure provision.

Although expressions of family values related to management strategies in the data point to the social nature of disability, the political ends of the social model i.e. society's acknowledgement that it should develop to accommodate the impaired, remain only half explicitly realised within the families. Accounts and observations of behaviours and values demonstrate that managing the body cannot be achieved without addressing non-physical aspects of the child such as their preference for rigid routine, and the families' location within social structures over time. In many cases these themes underpinning the social model were acknowledged directly by parents.

Limiting the isolating effects of being defined medically is a key focus of the social model. That is, the social model stresses that individuals should not be defined by their medical condition but recognised as humans with equal rights and needs as their non-impaired counterparts. Shakespeare and Watson (1998) outline this point with particular reference to disabled children and education:

Rather than considering disabled children as having special needs, it is more appropriate to accept that all children and adults have the same basic needs. Disabled people, regardless of impairment, are first and foremost human beings, with the same entitlements and citizenship rights as anybody else. It is up to society to ensure that the basic needs of disabled people are met within the systems and structures of education. (Shakespeare and Watson in Robinson et al 1998: 24)

The father from family 14 echoes these sentiments within the micro structure of the family:
Father14: Nicky's just a lovely child. Every child is different and you take them as they come... We try not to make distinctions between the other children and Nick just because he's, I don't know, apparently medically distinct. We try to say everyone gets the same type of treatment and not single Nicky out as a special child. Try not to give him more attention than the others. He realises he's got a problem, but not fully the physical/medical side of it. I doubt if he ever will and that's not of importance really. They're all special; they all have their own activities and personalities. (SESL, P2, SIBS)

Many families spoke at length of the problems of raising public awareness of the disorder amongst their families, friends and local communities, as well as within their local education and health authorities. This reflects the core principle within the social model that social inclusion should be achieved through social change as opposed to individual adjustment (Oliver 1996: 34). The following interview excerpt make this point:

Mother03: When she got it, cos I used to live in Peterborough and I went to my doctor with it, and she didn't have a clue. She was asking me for the information on it, but then she obviously got a lot better and read up on it and stuff.
Father03: We go down the surgery now and we always look but there's nothing on it, not one leaflet.
Mother03: My mum belongs to the society [PWSA] as well, and when the magazines come she take them down the doctor's surgery.
K: Oh that's a good idea.
Mother03: Yeah 'cos that's a way of other people getting to know. Cos it's a hard one to understand for other people. (SESL, P2, NOSIBS)

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Mother12: When we first moved in we did give leaflets out to the neighbours and the parents. But since then no one has ever mentioned it. I don't think anyone really says anything about it now. Maybe because he is more the normal side of things, he can have slip ups, as we all do...
Mother12: [on media coverage of obesity] It always amazes me that it's [PWS] never mentioned. You know 'cos they're always saying 'it's their own fault' and I always feel...
there should be a little thing saying 'by the way there are diseases which can cause obesity'. (SESH, P2, SIBS)

Despite these accounts and the social nature of disability displayed in the interviews, many of the key practices and values of parents point strongly towards an individual or biomedical understanding of disability within families. Both family 03 and 12 above can be seen actively contributing to public awareness of the disorder. And both have children on hGH. Thus individual treatment and adjustment are valued alongside changing public and service provider attitudes and awareness around PWS. Use of hGH is perhaps the most radical form of ‘normalisation’ (Wolfensberger in Flynn et al 1980) displayed within the data on management practices. In keeping with an individualistic model of disability, this practice aims to transform the individual to meet social and biological standards of ‘normality’ instead of questioning social definitions of ‘normality’.

Normalising practices are common throughout families’ management behaviours, with inclusion into everyday family life and wider social structures forming a central aspect of family values. These normalising practices take the form of pharmaceutical treatments altering physiology (hGH) and behaviour (fluoxitine, risperdal), but are also evident in the general aims of upbringing as seen in these accounts from families 01 and 07. Family 01 take a normalising approach to aiding their child’s inclusion within the family, whereas family 07 alter the structures around the child within the home:

Mother01: At the moment the main thing that’s an obstacle to us is behaviour. All the time you get very little warning, shouting, crying. What I really want is for us to be able to take him anywhere and for him to be able to get through it without an episode and not stick out like a sore thumb, for his sake as well as ours really. Whether it's going on holiday, for an outing or just a walk around town, to be able to be reasonably certain that we won't have a big outburst… It's the behaviour side that drives me nuts, the violence and all that, that's what the others [in the family] can find quite frightening. But the rest of it like this [gestures to the kitchen] it's part and parcel of Alex being normal really. He fits in relatively well. We, or I certainly, have always
taken the line that we treat Alex as normal...after all that's what you're doing isn't it, teaching them to fit in.

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Mother07: We try to keep things [everyday life for Gemma with PWS] as simple as possible. We do keep the pantry locked, don't spend a lot of time in the kitchen...we clear away the dishes as soon as we've finished. Really try not to make a big thing out of eating at all. (SESL, P2, NOSIBS)

Oliver's (1996) description of qualities within the individual and social model of disability draws attention to the emphasis in the individual model on 'care and control' in comparison to the stress on 'rights and choice' in the social model. The above quotation on locking practices from family 10 highlights the prominence of 'care and control' within families where access to food is denied. As seen in chapter four, the shift of responsibility for care from the person with PWS to the family in childhood and adulthood is an ongoing theme in medical ethics around PWS. Holland et al (1999), for example, draw attention to food as an area where rights and choice of the PWS individual should be overridden by law, in light of the individual's lack of capacity to make rational decisions in this area.

In accordance with Holland et al's (1999) observations on lack of capacity for decision-making around food, much of the data collected from families who deny access to food suggests that under a social model governmental framework, these forms of external control promote social inclusion, quality of life and freedom for the disabled person. As displayed in the quote from family 10 (access to food section, chapter four), the freedom from the stress and anxiety of his overwhelming preoccupation with food is one of the most important freedoms this mother feels she can give her son, this being achieved through alarm systems and locks. However, in light of the families within this research who have developed successful dietary routines without denying access to food, it is clear that the need for interventions must be judged on an individual basis, appreciating the diversity within PWS.

Whether practices can be seen to reflect biomedical or social model principles, the underlying aim within families is for the child to be able to integrate within their
community. Examination of family values has illustrated the importance of 'social inclusion' and data reveal families taking any route to achieve this i.e. social change via public awareness, often combined with individual treatments altering physiology and behaviour. Returning to figure 5.1 we can see that the reasons for valuing 'social inclusion' were primarily associated with embodied interaction between family members. Firstly, children with PWS were seen as being happy when they could be included in the same activities as others, as the following extract illustrates:

Mother10: Emma's [non- PWS child, 9 years] very good aren't you [to Emma]? And is a very staunch defender of Alex [PWS child] and will stand up for him, even when some people are less than patient with him. And Emma's very good at sort of saying 'he can't help it, it's not his fault'. It's hard for all of us because he doesn't appear so different. And when we meet someone new we have to remember that they won't know. It can be awkward. And you're very patient, if you're watching [TV] programmes you sometimes say 'haven't you already asked that question?' But Alex is desperate to be part of the family in a sense that, on a Saturday night if the girls are playing he likes to be part of that, it's a treat. And they paint his nails, the girls paint his nails!
Sister10 [aged 9]: He wants to be like us. (SESH, P1, SIBS)

Secondly, behaviour perceived as 'abnormal' was judged to have a negative impact on other family members:

Mother01: It's the behaviour side that drives me nuts, the violence and all that, that's what the others can find quite frightening. (SESH, P2, SIBS)

Thus, although practices and values can be linked to wider political stances on disability, these are not discourses which directly guide everyday actions. Behaviours moreover can be seen through this analysis as responses to embodied interactions which ease family life on an emotional and practical level. However, the synthesis of biological and social factors in the experience and management of disability comes across clearly through the data, perhaps suggesting that the concept of disability goes beyond biomedical versus social model definitions, as (Shakespeare et al 2000, 2001).
5.7 Conclusion

The micro structure of the family provides a location for the regulation of children with PWS, as well being a site for mediating between child and society. Although these processes connecting the family to wider social structures have been widely commented upon within the sociology of childhood and the family (Rosser 2003, David 2003, Dunn 1988), the importance of family environment for grounding the child’s relation to wider society is arguably heightened for children with PWS, due to the increased dependency on the family resulting from the child’s differentiated apprehension of social environment. As such, the analysis identified family values as being of great importance to management strategies and practices.

Physical health, emotional health, considerations for the future, and quality of life for all family members, were identified as the core values amongst the families. Analysis also identified various qualities commonly associated with these values. The sub-values highlighted the diversity of physical conditions presented within children with PWS and the social aspects that play a part in the experience of disability and its management. ‘Social inclusion’ emerged as a particularly significant practical aim within the families, appearing as a sub-category in three of the four core values: emotional health, considerations for the future and quality of life for other family members.

The significance of ‘social inclusion’ in management values and practices, points towards social model understandings of disability as social exclusion by parents. However, the data suggest that this value results from family members’ everyday experience of interaction and perceptions of children’s’ quality of life. Practices highlighting aspects of biomedical and social model conceptualisations of disability were used intermittently within families toward the key objective of social inclusion.

Within the data there is a general ‘messiness’ concerning how management practices interact with the values identified. That is, in many cases there is conflict between values within families. This chapter demonstrated these conflicts; firstly as conflicts between physical and emotional health; secondly as promoting long term happiness.
whilst allowing the child enjoyment on a day-to-day, short term basis; and lastly negotiating conflict between quality of life for the child with PWS and quality of life for other family members. This finding demonstrates the complex contradictions faced by those caring for people with PWS on an everyday basis.

So how do 'family values' fit into the theoretical analysis of everyday actions within the family? Values are seen to be primarily influenced by embodied interactions between family members, thus embodied interactions remain central to the process. Family values are also seen to have a more direct impact on everyday action than wider structures of social knowledge and discourse. However, some connection between wider discourse and family values is evident, as medical definitions of illness are upheld in everyday management and central values in terms of 'inclusion' parallel government agendas in social care.

Burkitt's (1999) notion of the 'thinking body' allows family values to be conceptualised as part of embodied interactions, as the beliefs held by individuals are not viewed as a separate realm of knowledge, but as an essential part of their emotional being. Burkitt also acknowledges the connection between values and wider societal structures, also visible within this analysis:

Thinking bodies are always connected to a transformed materiality and sociality. There is no absolute separation between nature and culture, body and mind, materiality and knowledge, for these can be understood as dimensions interconnected through relations and practices, involving the thinking bodies of persons and selves. (Burkitt 1999: 89)

Family values then are an essential part of embodied interaction between family members, they draw upon wider social structures and knowledge and act to mediate children’s relation to these structures. We now turn to examine the patterning of the practices and values identified in light of family structure and socio-economic status.
CHAPTER 6: THE IMPACT OF FAMILY STRUCTURE AND SOCIO-ECONOMIC STATUS ON FAMILY MANAGEMENT OF PWS

Introduction

The sample was designed to ensure variation in socio-economic status (SES) of household area and family structure. This chapter examines how these variations impact the family management of PWS, by addressing the patterning of practices and values identified so far.

Family structure variations were built into the sampling design (see chapter three): of the twenty families: six were lone-parent families. Two of the lone-parent families had only one child as did four of the two-parent families. This chapter examines how family structure impacts family management of PWS. It examines whether family structure affects the patterning of practices and values identified in previous chapters and identifies the various emotional effects on family members which underlie this process. This involves interpretation of data, drawing on sociological literatures around gendered caring responsibilities, the experience of lone-parenthood with a disabled child, as well as experiences of stigma and courtesy stigma within the family and children’s care work.

As seen in chapter three SES was operationalised in the sample using census data on the area in which the family lived, with half the sample (ten families) living in areas with SES above the average for England and the other half (ten families) below the average. This method of sampling was used to ensure difference of SES, which could be taken into consideration as contextual influences of family management of PWS. However the conceptualisation of SES and social class goes beyond these two groups, using the ideas of Bourdieu (1977, 1984) to theorise the embodied processes, related to class and lifestyle, which impact upon the family management and health outcomes within the families. That is, the two groups identified at the sampling stage were not intended to reflect, in any simple or stereotypical way, the existence of two distinct social class groups within the total population of families with PWS.
After addressing family structure, the chapter examines the main ways in which SES was linked to practices and values. Using Bourdieu’s (1984) theoretical conception of ‘habitus’ it will outline processes specifically relating to the acquisition of habitus for the PWS child, assessing to what extent this process is impeded by the embodied dispositions of the child associated with PWS.

6.1 Family structure: gendered caring, children as carers and the intensity of the caring relationship

The question of 'who' manages food in the families is complex. The analysis so far has identified the influence of embodied action of all family members, incorporating both differences in social perception based in the biology i.e. social effects of autistic spectrum-disorders in people with PWS and family beliefs and understandings in terms of 'values'. We have also seen that formal day-to-day decision-making tends to occur at the parental level. As these decisions can be seen to arise from interactions, agency cannot be straightforwardly apportioned to any individual or family level of individuals. In practical terms, siblings' role in helping to monitor food intake has been highlighted, as well as children with PWS directing their own choices around types of food and, to a greater extent, the timing of meals. However, within the families, particular individuals tend to take responsibility for the monitoring, planning and preparation of food for the child. This domestic division of labour within the households will be examined with reference to family structure.

Traustadottir (1991: 212) and others have highlighted a tendency for the gendered nature of caregiving to be overlooked in research on disability in the family. More recent research in this area tends to draw attention to differences in both the quality and quantity of mothers' and fathers' involvement with their disabled children (e.g. Read 2000: 52). A national survey conducted in 1994, of 1100 two-parent households in the UK with a disabled child, found that in 96% of cases the mother was identified as the main care giver (Beresford 1995). As Ungerson (2000) observes, literatures on care have moved away from being heavily gendered to incorporate factors such as class, race and wider processes such as privatization in the health services. However her article on the
production and consumption of long-term care in Britain highlights that caring is still strongly gendered, particularly within the family, contributing socially and economically to gender inequalities. This trend is also visible in census data (2001, www.statistics.gov.uk/census2001).

Observations of the centrality of gender for caring in families with disability are, however, apparent in literatures from the early to mid 1970s onwards, as Voysey’s (1975) US study of the experience of disability within the family illustrates. Voysey acknowledges that where she writes about ‘parents’ she is referring to ‘mothers’. She justifies this disparity on the grounds that: mothers were more frequently respondents in studies of disability in the family; they have more contact with outside agencies who in turn direct their services towards mothers rather than fathers; and that mothers tend to take overall responsibility for the care of disabled children in the family thus feeling more responsible for public presentations of the family (Voysey 1975: 215).

These findings were reflected in the data from families of children with PWS. Of the fourteen two-parent families, eleven mothers reported that they took responsibility as main caregiver in the household, attending to day-to-day food management and other intimate care of the child with PWS. In five of these cases the mother stated explicitly that caregiving was her domain and that the father had little input into everyday decisions around the child. Thus:

Mother19: He's [father] not back until half six, seven o'clock most days. So it is me that looks after Sonia? That's just the way we work it...I cook for them [children] at six and we [parents] normally eat later...I make the decisions about diet and treatments because I'm here all the time and I know slightly better what the issues are. If we book to see the dietician or psychologist it's always at times when he's [father] at work, so I deal with that side of things myself. (SESH, P2, SIBS)
Other families, where the mother reported that she was the main caregiver gave accounts of the father having some input. Most commonly this took the form of leisure activities and major medical decisions,

Mother12: We try to keep up to date with the research. Darren’s [father] is quite good at that actually. When Louie [child with PWS] went on Growth Hormones it was him [father] that spoke to the doctor and he [father] seemed to have read more of the articles than he [doctor] had. (SESH, P2, SIBS)

Father07: On Saturday afternoon I normally do something with her [Gemma with PWS], to give Rebecca [mother] some time to herself and because I enjoy it most of the time. K: What sort of thing do you do? Father07: We avoid shops, they tend to cause problems (behaviour). We’ve done a round of pitch and put, cinema quite often, feeding the ducks that kind of stuff. And of course she gets to stretch her legs a bit that way as well. (SESL, P2, NOSIBS)

Research on childhood illness such as Angst’s (1996) study of decision making in families of school aged children with cystic fibrosis, has highlighted the role of the child, alongside the parents in making major medical decisions. In this research however, children were not seen to be involved in this process. Possibly due to their age and to learning difficulties they experience, the kind of reasoning these decisions require was judged as beyond their capacity by their parents.

Where justifications were given by mothers for their position of main caregiver, they were usually practical and centred around the time constraints of the father's employment commitments. However, additional factors were given as justifications including professional medical expertise of the mother and preferences of the child with PWS. One family, whose situation was unique in the study, justified the mother's care giving role for the child with PWS as the father was a full time carer of another child in the family with multiple sclerosis. The couple had married after having five children in previous
relationships. Neither parent was in formal employment and care of other children in the family was shared equally.

Three mothers expressed that their previous educational and professional experience qualified them as the more able parent to be the main caregiver:

Mother03: Before I had Chetna [child with PWS] I was a nurse, so I can understand a lot of the terminology and the medical side. (SESL, P2, NOSIBS)

Mother20: [mother ex-nurse] My medical background is very handy, I do a lot of my own research and I tend to make decisions about diet and medical decisions, although we [husband/partner and self] always discuss it too. (SESH, P2, NOSIBS)

Four parents stated that having one main caregiver was more practical in light of the child's understandings and temperament. That is, as the child preferred to know what would be happening in detail on a day-to-day basis, having one parent in charge left less room for conflicting information and thus less likelihood of upsets and emotional outbursts. Thus:

Mother11: As far as she's concerned, Mum is boss and she's happy with that most of the time. He'll come and check the same things over with me again and again. I think he really needs that structure, he asks for it. We've had some real problems when one person says one thing and one person says something else, but that's mainly at school really. At home we have more of an 'ask Mum' policy don't we [to father]?

Father11: [nods]

Mother11: And Mum's word is final, usually. (SESL, P2, SIBS)

The child's preference for this straightforward authoritative governance by one parent was demonstrated frequently throughout the research by the children themselves. As this except from an observation of family meal displays:

Victor: Am having two, two, two potatoes now aren't I mummy?
Mother17: Yes that's right. We [child with PWS and mother] talked about this because we're [rest of family, Mother, sister aged 15] going to have a few more and so does Victor normally. [to Victor] But yesterday we had ice creams and we decided not to eat so many potatoes today didn't we?

Victor: Yes, me not mind.

Mother17: No, you don't mind that rule do you, because you get ice cream.

Victor: Like that rule a lot.

Mother17: Good boy.

[Victor is smiling and seems happy that he remembered and followed the rule] (SESH, P1, SIBS)

This preference for rigid structure and rules means that if parents contradict each other over food or more general issues, children with PWS can become extremely agitated. Whitman's (1994) findings from an international survey of parents with a child with PWS supports the notion that conflict between parents can contribute to extreme stress and behavioural problems in the child. And, as Waters reports:

> Any disagreement and lack of common goal will confuse the person with PWS, especially where one parent or family member is saying the person can do one thing, whilst another relation is saying he cannot: the person needs to know where he or she stands. (Waters 1996: 23)

Within three of the two-parent families, mother and father took equal responsibility for the day-to-day care of the child. Two families did not put forward any justification for this, it seemed to be part of how they had always approached family life. One family described the father's temporary unemployment as the reason for this dual responsibility for care. None of the fathers in this study took sole responsibility for care.

The division of labour within the two-parent families clearly demonstrates that responsibility for the care of the child with PWS was predominantly the mother's. This finding is in line with other research into caring, chronic illness and disability in the
family (Graham 1984, Beresford 1995, Thomas 1995, 1997, Ungerson 2000). Apart from the three mothers who cited professional medical or biological knowledge as an asset in everyday care of the child, little explanation was given by either parent in the families for the gendered division of labour in caring. This suggests the naturalised nature of family work carried out by women in the home perceived as being borne out of ‘motherly’ love (Sevenhuijsen 1998, Van Every 1997, Morgan 1996)

The intensity of the relationship between carer and child was observed in light of embodied interactions in chapter four. Accounts of the emotionally draining nature of this relationship in everyday life were expressed particularly strongly in accounts from lone-parents. Read (2000: 53) has drawn attention to the elevated number of lone-mothers with disabled children as compared to the general U.K population. More recent government statistics highlight this point, as Lewis et al explain ‘more than one third of non-working lone-parents have children with a long standing disability’ (Lewis 2002 source: http://www.statistics.gov.uk/events/NSOD/default.asp). Whilst acknowledging that loneparenthood by no means automatically indicates any kind of ‘impoverished personal experience’ for parent or child, Read (2000) highlights some significant practical and emotional factors, such as limits to household income and lack of day-to-day emotional support.

The six lone-parent families in the sample were all lone-mothers (rather than fathers). This was due to the absence of lone fathers with children in the age range within the sampling frame and again highlights the gendered nature of caring in families with disabled children. As this small sample of lone-parent families was selectively stratified by SES, little can be shown about how the economic restraints of loneparenthood in general affect everyday life and management of PWS. However, accounts of the emotional impact on parent and family of living with someone with PWS were both more frequent and detailed within this group, compared to the two-parent families in the sample. This excerpt from a lone-parent displays the feeling of isolation:
Mother08: We've (parent and child with PWS) got into a routine now. It's if we have to go somewhere else and I have to warn everybody, nobody ever believes me, it's really very frustrating. I think that's, (pauses) if there's one thing about dealing with life with Hannah that gets to me personally, it's that.

K: Why do you think that is?

Mother08: Because it's like everyone thinks I'm this neurotic woman, being over protective and fussing. They can't believe that you've got to be that strict. We stayed with my mum and she was putting on a pound every other day. And I think then she suddenly realised, this is serious. She's gone away on church weekends and come back having put on 2 lbs. They think she just needs to eat a little bit less than everyone else, they find it very hard too. (SESL, P1, SIBS)

The discussion of embodied interactions in chapter four illuminated the intense nature of the relationship between parent and child with PWS. Parents expressed feelings of isolation, exacerbated by a general lack of understanding of PWS in day-to-day contacts with professionals, their extended support networks and the public. Loneliness and isolation were described in part as the effect of caring for a child with an altered conception of the social world, who has a limited appreciation of the parent’s and other family member’s emotional needs. A Swedish psychological study by Olsson (2001) displays the high level of emotional impact associated with mothers of autistic children. Olsson found that depression is common in mothers of children with autistic-disorders, more so than in mothers of children with other learning disabilities. Comparing this to fathers Olsson found the same pattern, but with the number of fathers experiencing depressive symptoms being much less. In fact, the number of depressed mothers of non-autistic, learning disabled children was higher than the number of depressed fathers of autistic children (Olsson 2001: 535). Within this research, the isolation felt was often described in terms of the stigmatization of being seen as a bad parent in public places, where the underlying reasons for behaviour problems in the child are not directly apparent to the onlooker. All six (female) lone-parents alluded to the emotionally testing nature of managing PWS in everyday life, as did four (three female, one male) of the fourteen parents from two-parent families.
Although there has been no sociological research on the emotional impact of parenting children with PWS, Gray’s (1993, 2002) study of courtesy stigma in Australian parents of autistic children forms a useful starting point to explore these issues. Gray’s research utilised Goffman’s framework of social interactions (Goffman 1963) to explain how parents of children with autism experienced stigma in public places. His findings acknowledged the role of gender of parent in perception of stigma, ‘[T]he sex of the parent was strongly linked to the perception of stigma...Although the number of fathers was small, there was a marked tendency for them to feel less stigma than the mothers (Gray 1993: 114). Gray attributes this gender difference to mothers feeling more responsibility for the presentation of their family in public and feeling more personally responsible for their child’s autism than the fathers (Gray 1993: 114-115).

Gray highlights the role of normal appearance of the child as a key factor heightening the experience of stigma for parents. He explains how courtesy stigma is increased in the parent as their child’s appearance is that of a normal child, whilst the nature of behavioural problems displayed in the condition are severe. The fact that the presence of a medical disorder is effectively ‘hidden’ in public encounters makes the presentation of anti-social behaviour in the child more stigmatizing for the parents (Gray 1993: 119).

The problematic nature of ‘normal appearance’ was also drawn upon by parents of children with PWS to explain awkward situations in public. In many cases this was linked to autism-related behaviour. For example,

K: So it doesn't stop you doing things you might otherwise?
Mother1: It cuts short things at times. It's not worth the bother sometimes. *When she makes an awful racket and people start looking at you as if you're abusing her!* Cos they haven't got a clue, cos they don't know.
K: She presents quite normally?
Mother1: Yeah, so in a sense. The drawback with Kelis, unlike children who are um...like Downs [syndrome], you can see something's wrong with them, cos they do look
that bit different. But with Kelis, although they [medical literature] say they've all got similar characteristics, similar things on their faces or whatever, you can't see that when you're looking at Kelis on her own. (SESL, P2, SIBS)

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Mother16: That’s my main priority at the moment. Being able to go out wherever, whenever and to be reasonably sure we weren’t going to have any dramatics!

K: Is it quite frequent that he’ll have some sort of episode when you go out then?

Mother16: Most times I would say, and if he doesn’t it’s because you’re constantly working towards stopping it happening, so you can’t really relax and enjoy whatever it was you’re supposed to be enjoying in the first place. It can be quite embarrassing because the way he looks you can’t really see there’s anything too different about him. I sometimes think, not that I’d ever wish for him to be more handicapped, but if he looked handicapped it would help other people to understand and not be judgemental. I think it would help them take the diet stuff more seriously too.

K: Do people actually say things to you?

Mother16: Not really, just I get the odd disapproving look, ‘what’s she doing to him?’ or ‘can’t she control her child?’ type look. And you can’t stop and explain it to them can you, but you want to every time. Yes I’m getting used to it now though, it affects me less and less. (SESL, P1, NOSIBS)

As the above excerpt from family 16 hints, the problems of normal appearance in social interaction are not only associated with autism-related behaviour problems but also issues around weight. There is a common theme, especially among lone-parents, of unseen and hidden body work and a parental desire to have this work acknowledged by others. Not enough data was collected on the subject to be able to make causal statements about the reasons for this feeling amongst single parents. However, following Read (2000) and Beresford’s (1995) observations on lone-motherhood in families of the disabled, it could be suggested that this need for acknowledgement reflects the absence of emotional support provided by the partner in two-parent families, as displayed in the following interview excerpts:
Mother08: [on obesity in other children] It does annoy me, cos you see more and more, really big children don’t you?...I manage to get Hannah to just about what she’s meant to be (weight), and that’s with all the extra PW problems and you think ‘why can’t you? (SESL, P1, SIBS)

Mother16: It’s really very annoying, I do have to check myself regularly with people. It’s because they think I’m exaggerating and ‘it can’t possibly be that bad’. It’s because he’s not very overweight so people don’t see the risk, they can’t imagine that one biscuit, one ice cream will hurt and ‘children should have treats’ and all that. But they don’t appreciate the very reason he’s normal is because of everything we do day-in-day-out and have done for years. (SESL, P1, NOSIBS)

Having examined the impact of whether the family had two parents or a lone-parent we will now examine how number of children within families affected family management of PWS. The sample selection addressed not only number of parents, but also number of children within the families. In six of the families, the child with PWS was the only child. ‘Use of routine’ practices such as eating at set and rigid times were more common amongst this group compared to families with other children. Seven families reported keeping to a ‘strict’ structure of food provision in the household, five of these seven were families with only children. In the absence of siblings it seemed the needs and preferences i.e. for routine, of the child with PWS could determine food management to a greater extent. Unsurprisingly the values of ‘consideration for other family members’ was expressed less in this group, as there were less family members to consider.

Drawing on a study of twelve mothers of children with disabilities in the West Midlands, Read (2000: 28) identified a trend for siblings to contribute towards the care of the child with disability. Burke’s (2004) Brothers and Sisters of Disabled Children focuses specifically on children’s care work and services provided to support the siblings of disabled children. In this study there was a tendency for non-disabled children to aid their parents emotionally and practically in care work, as described by Burke (2004: 54). As we saw in chapter five, this most frequently took the form of monitoring food intake and
informing parents. It was also common for children to demonstrate consideration for their sibling with PWS by clearing away food after they had eaten and not eating excessively in their presence. As well as practical help in food management, siblings children also displayed aspects of emotional care work. These most commonly took the form of defending their sibling in the school environment and explaining his/her behaviours to peers. This behaviour was encouraged and rewarded by parents:

Brother17 [aged 13]: Most people [at school] are OK, I’ve told them what he’s like and that’s just him and they get used to it.

K: How often do you have friends round?
Brother17: About one time a week or more in the holiday.

K: And does that normally go ok?
Brother17: Yeah, they know not to eat sweets in front of him because I told them and what’s going to annoy him and stuff.

K: Do you normally do stuff all together, your friends and F(child with PWS) when they come over?
Brother17: Not really, sometimes. He’s really annoying sometimes. I do stuff with him when it’s just us.

K: Do you prefer doing stuff with your friends?
Brother17: [nods] Yep. He’s alright, at school I sometimes see him at break times and I do go and talk to him. If he’s in trouble or something the teachers sometimes ask me and I help. (SESH, P1, SIBS)

Although the age and gender of siblings were not considered as a factor when the sample was drawn, they emerged as important in determining the way siblings cared for the child with PWS. Where children were the same age or younger, relationships often were marked by conflict and tension, regardless of gender. These children contributed to the care of the child with PWS by aiding in the monitoring of food intake and acting as educators and mediators amongst peers. In three families including a female sibling who was five or more years older than the child with PWS, the sister’s relationship to the child
was much more in line with the mother’s. This was not evident where siblings were significantly older and male.

In each of the three households with older sisters, the sisters aligned themselves with the mothers during the data collection process. They contributed to interviews on the child’s medical history and personality, practical and personal problems of day-to-day management and commented on how and why management strategies had developed. All three described having an input into everyday decisions around the child and taking on caring duties to allow parents time off. This tendency illustrates that Voysey’s (1975: 215) observations on the highly gendered nature of caring still carry much resonance for families with disabled children.

Young carers are defined by the Carers National Association as: ‘Anyone under 18, whose life is in some way restricted because of the need to take responsibility for the care of someone who is ill, has a disability, is experiencing mental distress, is affected by substance misuse of HIV/AIDS. (Rollins 2002, source: http://wwwcarersinformation.org.uk/showdoc). The siblings in this study can therefore be described as such. Burke (2004: 65) suggests that young carers suffer significant emotional effects related to their role and that this can have a considerable impact on their developing identities. The emotionally turbulent nature of relations between PWS and non-PWS siblings has been discussed in chapter four (see figure 4.5 and related discussion).

The impact of family structure on the management of PWS shows a continuation of trends already identified in literature on disability and the family e.g. gendered caring within the family, the emotional impact of disabled children within families, and siblings as young carers. The gendered nature of care work is visible, not just between parents but also in siblings, particularly where older than the child with PWS. In addition, the only lone-parent families available in the sampling frame were those with lone-mothers. Problems of stigma, related to autism and felt moreover by mothers than fathers (Gray 1993, 1999) were also apparent in the data. The emotional strain of care work and desire
for recognition of hidden work was expressed most strongly in the accounts of lone-mothers. Children contributed in various ways to the care of the child with PWS. In younger children this tended to take the form of food monitoring and mediation work amongst peers. In older children, especially females, a more supervisory role was displayed. In families with only children, stricter 'use of routine' practices were apparent. Having examined the impact of family structure on family management we will now turn to look at SES.

6.2 Impact of socio-economic factors on ‘practices’ and ‘values’

Within the sample, SES was operationalised using government-defined measures from local area level census data (see chapter three). These measures of home ownership, education, crime and self-reported health are far from a fully realized conceptualisation of social class and its complex relation with health. Literatures on class and health point beyond quantifiable material factors towards more complex issues of lifestyle and everyday interaction (Blane in Scambler 2004, Macintyre 1997). However, indicators towards class are apparent in the data and a more detailed analysis of the impact of SES on family management and health outcomes was possible drawing on interview and observation data. The sample was divided such that half of the families lived in geographic locations scoring above the English average on the multiple deprivation index (Office of National Statistics 2000, source: http://www.statistics.gov.uk/StatBase/Product), and half below this average. However, there was no indication of a corresponding distinction in the families’ management practices, health beliefs and values. Many practices and values occurred throughout the families, regardless of SES. Nonetheless, certain patterns did emerge in relation to the two SES ‘groups’, as certain approaches to management occurred more frequently in one group or the other.

In terms of practices, ‘keeping occupied’, ‘use of routine’ and ‘biological alteration’ occurred evenly throughout the two SE groups. ‘Keeping occupied’ practices were observed in all families, with 11 explicitly stating this as a food management strategy. Although 'use of routine' was strongly affected by family structure, with strict use of
routine occurring mainly in families with no other siblings, this had no connection to SE group. The utilisation and objection to hGH also occurred equally in both groups. This lack of association can be seen in part as the result of embodied dispositions of PWS overriding SES. That is, children with PWS have predisposed preferences towards rigid routine, and participation in focused activities regardless of their family’s economic and social position. This draws attention back to the centrality of embodied agency in determining management practices.

Although in some ways the human embodiment of people with PWS overrides SES, there are also many associations between health and class in this research which tie into the broad sociological literature on this subject. For instance, the tendency of higher social classes to place increased value upon participation in sporting activities than lower class groups is well documented in sociological literatures (Wilson 2002, Jarvie 1994: 193). More active, sports based variations within practices of ‘keeping occupied’ were more common in the higher SE group. Families in this group were also more likely to state this explicitly as a management strategy. Six out of ten children with PWS in the high SE group took some form of physical sports activity outside of school hours everyday, compared to two of ten in the lower SE group. This could partly be explained through access to resources. For instance, two of the families in the higher SE group had an indoor swimming pool within their home and a further three had various gym equipment and a room in the family home dedicated to physical exercise.

Drawing on Lefcourt’s (1976) theory of ‘locus of control’ the emphasis on physical activity within the higher SE group could be explained in cultural terms. Locus of control refers to an individual’s expectations concerning where control over subsequent events resides. In relation to health, this can be viewed in terms of social position, with members of the middle classes tending to see their behaviours as responsible for their future health outcomes and members of lower class social groups taking a more fatalistic approach, seeing themselves as out of control of their health (Elstad 1998). In line with this, families in the higher SE group stressed the importance of ‘healthy, active lifestyles’ much more frequently. The patterning of ‘access to food’ practices also highlighted this
trend in relation to the promotion of internal versus external control around food. The three families who used no restraints to food in their households, neither locks nor food placing, were all in the higher SE group and placed a high emphasis on active lifestyle as a key management strategy.

Values of 'concerns for the future' and 'quality of life for all family members' were not visibly patterned by SES. The values of physical and emotional health were also a high priority across all the families. However, the various conceptualisations of these two concepts differed between the two SE groups. Across all of the families, physical health was spoken about in terms of medical conditions related to PWS, such as weight gain, scoliosis and skin picking. Physical health was also expressed as an abstract form, as general health and fitness. The latter was much more frequent in the higher SE group, where physical activity was promoted as away of life for the child with PWS and other siblings. As discussed in chapter five, within these families there was an holistic approach to health where physical and emotional health were often expressed as being essentially connected, if not the same thing. In contrast, three of the families in the lower SE group and one from the higher, alluded to managing PWS as maintaining a balance between physical and emotional health, in that too much attention on weight control could have a negative emotional effect on the child. That is, in the lower SE group conceptualization of health, physical and emotional health seemed to be conflicting concerns, compared to the higher group where increases in physical health were seen as improving the quality of emotional health. The three families from the lower SE group stressed the importance of forgetting about weight management every so often, to allow their children to enjoy themselves. As this quote from family 03 describes:

K: Do you ever get a chance to have a holiday or anything like that?
Chetna: [shouts] Yarmouth.
Father03: Yeah, every year it's a jaunt to the self same place cos it's a nice small place, stuff for the kids to do and it's only a mile or so away from Great Yarmouth itself. But because it's a nice small place you know where the kids are, guaranteed. We know where
they are so we always go there. *It's only a week that they go for, but we do tend to let the reigns go a bit just slack, 'cos we need a holiday too and that.*

Mother03: Plus we're pretty active on holiday, do a lot of walking. So she's still exercising, we try to do some swimming.

K: So what sort of dinners do you have on holiday compared to what you might have at home?

Chetna: [shouts] Fish and chip!

Mother03: Oh yeah, we do have a treat of fish and chips. But if we go to a restaurant she normally has a child's meal. She's now got to the stage where she's a bit bigger and she'll try and say ‘I want a bigger meal’. But she still has a small one with a little portion of ice cream, as well. *It's really hard cos they all ask don't they, and you want to give them a good holiday and so they do sometimes get extras.* (SESL, P2, NOSIBS)

In summary, a few of the family practices and values already identified in the analysis were patterned by the SES of the family. This patterning reflects a distinct approach used by three families in the higher SE group. This approach involved a lifestyle based around physical and sporting activities, with open access to food. Individual behaviours were seen as crucial to controlling health outcomes. Values and practices amongst other families were mixed, many having no visible relation to SE position.

6.3 Socio-economic status, food choice and medical knowledge

Further analysis of the data, revealed that types of food consumed and relationship with medical discourse had a more direct association with SE group. Food choice has been widely examined in relation to social class (see Bourdieu 1984, Mennel 1985, Calnan 1990, Murcott 1992, 1998, Lupton 1998). Fitting with Lupton’s (1998) observations of the ‘body as aesthetic’ amongst the middle classes and functional amongst the working classes, Calnan’s (1990) study of women and food choice showed that middle class women tend to focus on the ‘balanced diet’ and are cautious not to overeat, whereas working class women tend to stress the importance of substantial and filling meals.
In the interviews, notions of what constitutes ‘healthy’ meals were so taken-for-granted that extracting from the research participants what it was they actually ate was often a lengthy process. Problems in ascertaining what foods were eaten from interviews meant that observation sessions were based around meals wherever possible.

The following examples demonstrate this problem. The first is taken from a family in the lower and the second from the higher, SE group.

K: Can you describe some of the typical meals that you prepare?
Mother16: At 6ish they eat and it’s just a normal meal, I use low fat and his meal is just a bit smaller portions.
K: So do you cook from scratch everyday or do you use some frozen foods?
Mother16: Sometimes I cook from scratch, but that’s normally at the weekends, if we have a roast or something.
K: Can you give me some examples of meals that they would usually eat?
Mother16: They’re just standard meals. Usually potatoes, vegetables, and lots of them and then lean meat or fish.
K: So when you do a roast that’s usually lean meat like poultry?
Mother16: Yes, it’s normally chicken. That’s the favourite at the moment.
K: Do you ever have vegetarian meals?
Mother16: Not that often, once a week maybe I’ll do pasta and veg. (SESL, P1, NOSIBS)

K: So can you give me a couple of examples of typical evening meals?
Mother01: We always eat as much fresh as possible, he stocks up on fruit when he’s back from school and then the main meals are often more like snacks because I give him a big packed lunch.
K: What sort of thing? Is it not always a hot meal then?
Mother01: No, well especially not in the summer. We do lots of salads.
K: Like pasta salad?
Mother01: I tend to avoid too much pasta because it’s so high in carbohydrates. It’s usually a leafy salad with some chicken or tuna. Sometimes I use a bit of couscous or
homemade croutons, you know toast cut up basically. Always lots of seeds and herbs for flavour.

K: In the winter you have more hot meals?
Mother Ol: Yes.

K: Do the meals get bigger then or you still manage to keep them quite moderate.
Mother Ol: Yes I think they probably are bigger. We have jacket potatoes with lots of salad or tuna and vegetables. We normally have a starter like soup or a slice of melon.

K: Do you usually buy a ready made soup?
Mother Ol: Yes, but I get the fresh rather than the packet ones.

K: Oh, like the Covent Garden ones?
Mother Ol: we have had those. I tend to get Sainsbury’s own.

K: Do you go for reduced fat at all or just the standard?
Mother Ol: Never no, just one’s with fresh ingredients. I’ve always thought it’s important to have nutritious whole foods and not be scared of calories, as long as it’s good food.

(SESH, P2, SIBS)

Thus it was from observational notes rather than interviews that data on food types were derived for this analysis. Food choices were not completely distinct in the two SE groups. However, there was a tendency in the lower group to consume higher levels of carbohydrates and more processed foods. These processed foods often took the form of calorie controlled products such as Slim-a-soup, ‘Go-ahead’ or ‘Be-good-to-yourself’ bars and reduced calorie ready meals, ‘Muller-lite’ yoghurts and processed cheeses which come in small portions e.g. ‘mini Baby Bell’. Two of the families in the lower SE group had experimented with the ‘Weight Watchers’ diet plan for their children and themselves and were pleased with the results.

Families in the higher SE group, by contrast, often explicitly stated that they did not count calories and avoided diet foods due to their lack of nutritious value. The emphasis for these families tended to be on what was viewed as nutritious, unprocessed foods which were naturally low in fat. Meals tended to be smaller. Foods were more likely to be cooked from scratch. Patterns in food choices connected to SES were apparent but not all
encompassing within the families. Five families in the higher group followed this social
class based mode of healthy eating rigidly, as did six of the lower group. The remaining
nine families ate a mixture of processed, fresh, frozen, homemade and diet foods, and so
could not be bracketed into one style of food choice.

Families in the higher SE group spoke much more frequently about medical aspects of
PWS, often utilising the language of medical science in doing so. Emily Martin’s classic
book The Woman in the Body (1987) described similar relations between social class and
access to medical language and knowledge. Choices of medication, the nature of the
condition and its symptoms were often discussed at length. Use of medical terminology
was apparent in this mother’s description of her child’s weight:

Mother05: He is slightly overweight but the percentile shows he’s below average for
people his age with PWS so I don’t let it bother me too much, for now. (SESH, P2, SIBS)

Families in the higher SE group tended both to utilize and question medical discourse on
PWS and medical professionals. hGH was used equally in both SE groups and
individuals from both groups voiced concerns and objections to it. However use of
dieticians and psychiatrists was more frequent in the higher SE group, with seven of the
ten having regular contact with one of these. In the lower SE group, four of the ten had
used one of these in the past but only one family maintained regular contact. This finding
is not specific to families of people with PWS; it is typical of class related practices in the
literature connecting individuals with lower SES to poorer utilization of health services
than those of higher SES.

This can again be linked to ideas of locus of control over health with middle class
families becoming more involved in medical discussion and displaying a greater
confidence in terms of understanding medical knowledge and dealing with professionals.
The tendency of families in the lower SE group to withdraw from contact with medical
professionals is indicative of the assumptions of fatalism and lack of control over medical

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phenomena suggested in the locus of control model. The confidence of middle class families to both access and critique health services and knowledge in this research is portrayed in the following example taken from the higher SE group:

K: Have you ever accessed or been referred to professionals, like Psychologist, Dieticians?
Mother02: Yes we did. We went to a psychologist for quite a while. It tended to be just me that got there. Sometimes Becky came along sometimes not. I thought it became quite awkward because they'd ask a lot of searching questions, but Becky couldn't really cope with it. It worried her and would play on her mind. Why are we talking about that? What do you mean it was this that and the other? Although they were specialists in their own fields I don't think they had a good appreciation of Prader-Willi issues, and it was doing more harm than good. So I found that difficult at times.
K: Do you still access those services at all?
Mother02: I'm looking out for one at the moment really, but someone like I saw who has a proper appreciation of what you should and shouldn't say to Prader-Willi people.
(SESH, P2, NOSIBS)
The confidence of the higher SE group with medical knowledge is also demonstrated in the quote from family 01 in the 'physical health and care of the body' section, chapter five. This mother went against professional advice and carried out a large amount of successful muscle work with her child as an infant.

It is important not to overstate the presence of distinct class cultures corresponding to the two SE groups. The three families in the higher SE group, with no 'access to food' restraints and highly physical lifestyles also displayed typically middle class food tastes and a confidence with medical discourse. These families had a distinctive 'lifestyle' approach to managing PWS and the health of the family in general, which they stated explicitly, took responsibility for and saw as having a direct impact on the child's emotional and physical health. In other families, although trends could be isolated in food choices, contact with medical profession and locking behaviours, these aspects were mixed within families, across the SE groups. Blaxter (1990) highlights the complexity of
health related behaviours, finding similarly that totally ‘healthy’ or ‘unhealthy’ practices in individuals are rare and that mixed patterns of behaviour and beliefs are more typical.

6.4 Habitus, taste and logic of practice

The theoretical analysis so far and the nature of findings relating to SE group lend themselves to a theorisation of class based around the ideas of Pierre Bourdieu (1977). Williams (1995) draws attention to the useful nature of Bourdieu’s ideas for theorising class, health and lifestyle, in particular the relationship between health-related knowledge and behaviour. He emphasises the value of the centrally embodied nature of Bourdieu’s work, combined with its link to material and structural factors.

As a theory which is based around the everyday behaviours and dispositions of individuals or ‘practice’, Bourdieu’s work has a relational foundation, as has the theorisation of the management of PWS in the family so far. His explanations place significant importance upon the interaction between embodied individuals and the way individuals relate emotionally to aspects of the social world i.e. via embodied ‘dispositions’ in the construction of ‘taste’. Embodiment and emotionality are crucial to the workings of the concept of ‘habitus’. Habitus links individual behaviour to material factors (i.e. agency to structure) on the basis that our tastes reflect the everyday environments that we are familiar with and with which we have built an embodied and experiential security. The embodied nature of Bourdieu’s concept of habitus is clearly displayed in this quote: ‘As an acquired system of generative schemes objectively adjusted to the particular conditions in which it is constituted, the habitus engenders all the thoughts, all the perceptions and all the actions consistent with those conditions and no others’ (Bourdieu 1977: 95).

Bourdieu’s ideas around ‘habitus’, ‘taste’ and ‘practice’ can be related to the research findings on the socioeconomic patterning of management beliefs and practices and the dispositions displayed by family members. ‘Taste’ in Bourdieu’s (1984) analysis refers to a wider remit than food choices, including bodily size and shape, deportment, style of dress and décor. However, food preferences and arguably conceptions of ‘healthy’ foods
are explained in terms of social class and material structure. Findings on food choices
displayed a preference for light, non starchy, natural foods in the higher group and more
substantial meals and produced diet foods in the lower group. William’s (1995)
interpretation of Bourdieu’s ‘taste’ describes the role of structure and the material in the
construction of distinct, class based, tastes. Describing how people develop tastes for
‘what is available to them’, Bourdieu writes ‘…virtue made out of necessity which
continuously transforms necessity into virtue by inducing ‘choices’ which correspond to
the condition of which it is a product’ (Bourdieu in Williams 1995: 590).

These class dispositions which make up the ‘habitus’ include vocabulary; this offers
some explanation for the differences in confidence when interacting with medical issues
and dealing with medical professionals. It also feeds into the locus of control attributes
displayed in the SE groups; the higher group is emotionally disposed from their
experience of their social environment to feel in control of their own and their family’s
health, whilst the lower group inherit a resigned and disassociated attitude towards
medical knowledge and practitioners and health outcomes.

The notion of underlying materialism reflects Bourdieu’s ideas on the ‘logic of practice’,
which also forms an interesting backdrop against which to set our SE findings. In ‘Logic
of practice’, behaviours in everyday life are guided by, often unknown to the individual,
practical or implicit knowledge from the experience of that individual. As these guiding
logics are unknown to the individual, they justify behaviour with ‘official accounts’
which often take the form of collectively held discourses that the individual can apply to
their situation. In application to the SE patterning of behaviours and values, the differing
conceptualisations of physical and emotional health demonstrate this process well.
Chapter five depicts that, across all families, physical and emotional health are core
guiding values to management practices. However the various conceptualisations of
health within these families can be seen as a reflection of their learnt experiences and
cultures. These conceptualisations have been seen to have a relation to SE position,
further highlighting the value of this theoretical base.
6.5 Class habitus, health outcomes and the individual with PWS

To this point our theorizing of class has concentrated on the family as a unit rather than on the child with PWS. In Bourdieu’s analysis, class structures and behaviour, including health behaviours, are reproduced through habitus. As the development of tastes and behaviours which form the individual’s habitus are grounded in the materials and structures which surround them, the family can be seen as an extremely influential cite for the development of habitus in children. This section will address whether this works in the same way for children with PWS or whether this social process is inhibited by their predisposition towards certain ways of thinking, feeling and acting. We have seen the agency that these biologically based, embodied dispositions of the child with PWS have in directing action, in chapter four and in our discussion of the lack of association between SES and certain family management practices i.e. ‘keeping occupied’ and ‘use of routine’.

There are certain identifiable ways, demonstrated in this research, in which individuals with PWS do not take on the same dispositions as their peers or develop at the same pace. For instance, where the child with PWS had younger siblings it was common for parents to describe the process of younger siblings overtaking the child with PWS socially and intellectually:

Mother15: Simon doesn’t get as much money as her sister [sister is one year younger than Simon]. And there are difficulties because his sister, being very close in age but actually younger is becoming the older sister and we work very hard to make Olivia feel like she is the older sister. Like her sister’s allowed a mobile phone and of course Olivia wanted one too. And I said no because you don’t need one. And we had to work on that quite hard. To try to justify why in a way she can understand. It’s very hard realising how she can keep the role of big sister without obviously being big sister. (SESH, P2, SIBS)

Mother10: And it used to be...Joe [15 years old] could hold his own. He could read before they [younger sisters 9 and 7] could, now they can do everything better than him. (SESH, P1, SIBS)
Interviews with children with PWS demonstrated that use of language and comprehension of events was significantly different that of their siblings, as this session on ‘food and hunger’ with a child with PWS and his two younger brothers displays:

K: [to Victor 13, years old, child with PWS] So when you’re hungry, can you tell me what that feels like?
Victor: [pause] not really.
K: That’s ok, but you do feel hungry a lot you said?
Victor: yes
K: Could you say where about in or on your body you get that feeling?
Victor: don’t know.
Brother17 [aged 10]: He doesn’t know.
K: [to brother] Do you know?
Brother17: You’re tummy rumbles and when you see a nice burger your mouth waters because you want to eat it so much.
K: [to other brother] What do you think?
Brother17 [aged 12]: yeah, like that. [pause] If I’m really hungry I feel a bit sick, but that’s only happened once. (SESH, P1, SIBS)

Parents and siblings also give accounts of differences in the development of social skills of their children/sibling with PWS. The 15 year-old sister of Jason gives the following account:

Sister09 [aged 15]: I used to play with him more when I was little. But to be honest he was a bit of bully, even though I was older. It’s not his fault and everything like that, but he’s got a really short temper.
K: Did you enjoy playing with him sometimes?
Sister09: yeah, I must have. But I wouldn’t choose to now. Well, I don’t really ‘play’ anymore like he does anyway. I do help him with home work sometimes, that’s okay but
it's still mostly him having everything his way. That's just what he's like I guess. (SESL, P2, SIBS)

However, other data suggest that these embodied dispositions of the children with PWS that set them apart from their siblings, do not exclude them from achieving a habitus in line with their family members. Firstly, the two incidences of obesity in the sample occurred in families where at least one parent described struggling with weight issues themselves, one family from the higher SE group and one from the lower. Of the three families in the higher SE group who lead 'active lifestyles' and had open access to food, all of the children with PWS were a 'healthy weight', as were all family members. This patterning of weight outcomes in the children with PWS suggests that weight management of the child with PWS is more easily achieved in families where other members do not experience overweight or obesity.

Overweight and obesity of children with PWS were distributed evenly over the sample, irrespective of SES. The higher SE group contained one case of obesity, five overweight and one obese, the lower group contained five healthy weight, four overweight and one obese. A recent epidemiological study of childhood obesity demonstrates an extremely close link between social class (defined by parental occupation) and obesity. Ness et al (2006) reported total fat in the highest social class (I) to be 6.5 kilograms, whilst in social class IV it was 7.6. Between these results a clear gradient displaying an inverse correlation between class and fat was evident. The discrepancy between class/weight outcomes in the people with PWS in this research, and findings from the general population, can perhaps be seen as a reflection of the vast amount of body management work carried out in families of those with PWS from all backgrounds.

A second way in which the children with PWS successfully achieve a habitus in line with their significant others is through their capacity to mimic (see diagnostic criteria Appendix one). When describing an event many of the children with PWS in this research had a tendency to act the event out, very convincingly and emotively playing the
parts of the characters involved, remembering and repeating what was said and by whom.
As displayed in these conversations:

Matt: Mum, Mum and after we got him [Rabbit] our house got burgled didn't it.
Mother18: Yes
Matt: and our next door neighbour innit? There's a lady called Lou innit? and she looks after our rabbits doesn't she? And she was doing the washing wasn't she? And she see the computer cupboard open. And she said "why is that open? I'd better go and check".
Mother18: [to K] [sighs] Do they all go off on a tangent?
K: Yes it's fine
Matt: And she came in [voice louder to keep the focus on him], listen, and this was the first day [of holiday] this was the day we first got there wasn't it, the just day we first, the day we first, first just got there. And the next day Jackie and stuff came [family friend], my big brother and sister came. Didn't they ...didn't they Mum? And then you had to ring home and then she say “I’m sorry, the tower’s not there” [TV, Video etc] [pauses] didn't she? Everything broke up...didn't she?
Mother18: Yes
Matt: And all the rings, who give you the rings
Mother18: Well it was all my jewellery wasn't it?
Matt: And it was from Nanny May and Nanny Breen and one from Uncle Marve.
Mother18: Yes
Matt: And they took your jewellery box as well
Mother18: Mmm
Matt: And they took Dad's stuff. Then someone came who works, and he said “knock, knock, knock... is Mr Gunn here...Mrs Gunn? Mr Gunn? Your house has been burgled” and dad said “oh shit!” [Voice raised
K & Mother18: [laughing]
Mother18: You're not supposed to say things like that are you?
Matt: I mean oh sugar...Mum I meant oh sugar. [Squeals, seems happy]
Mother18: I know you did. (SESL, P2, SIBS)

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K: Do you go to the football?
John: Yeah.
K: Do you enjoy that?
John: Yeah.
K: How do you feel when you're there?
John: Amazing. When they score I go 'YEASSS, GET IN, GET IN, GO ON, GET IN' 'GO ON SON, GO ON, GET IN' [voice raised, punching the air- reliving the moment]. (SESL, P2, SIBS)

This capacity to remember and imitate, aids the formation of habitus in line with those around the individual with PWS.

6.6 Conclusion
Family structure and SES impacted strategies for the management of PWS within families. In terms of family structure, data displayed the gendered nature of caring within the family. Although in many two-parent families both parents took equal responsibility, all the lone-parents in the research were female due to lack of male lone-parents in the sampling frame (PWSA (UK) members database). Further to this, across the twenty families no fathers took sole responsibility as carer. Lone-parents were found to be more aware of experiencing courtesy stigma, and described the emotional impact of parenting a child with PWS in more detail than parents in two-parent families. A key feature in the emotional impact described by lone-parents was frustration related to public, professional and familial misconceptions of the disorder and lack of recognition of the 'hidden work' involved in family management. Siblings were found to contribute to the care of the child with PWS both emotionally and practically, with older female siblings aligning themselves more with parents in terms of responsibility, knowledge and supervisory duties within the household.

Many practices and values were common in the two groups. This lack of association indicating the importance of the embodied dispositions specific to PWS and their impact on management practices, overrode SES factors. However, 'active lifestyle' approaches
to management and open access to food within the home were distinct to families in the higher SE group. Food choices and conceptions of what constitutes ‘healthy food’ were visibly differentiated between the two groups. There was a tendency within the lower SE group to use calorie controlled foods, whilst higher SES families emphasised the importance of nutrition, in many cases explicitly stating an avoidance of calorie counting approaches. A greater accessing of health care services and a more confident and critical approach to medical knowledge and services was also apparent in the higher SE group.

The two groups identified in the sampling process were not intended as a conceptualisation of class in the population of families of children with PWS more generally. Processes relating to class distinctions and reproduction were conceptualised through Bourdieu’s (1984) notion of ‘habitus’, this conceptualisation appreciating the corporeal dimensions of class reproduction. Further to this Bourdieu’s notion of habitus is drawn upon by Burkitt (1999) in his theory of embodiment and social action, thus this conceptualisation of class fits the theoretical backdrop of analysis in previous chapters. The case of PWS demonstrates how biology must be taken into account as one of the objective conditions out of which habitus arises, especially as much Bourdieu-inspired work has reduced those objective conditions to only a matter of class position or gendered identity (Colley et al., 2003). Intellectual functioning and differing modes of social perception and interaction are seen to mediate the formation of habitus in individuals with PWS. However, the process of habitus acquisition remained valid for children with PWS, as ‘healthy weight’ and ‘obesity’ outcomes for children with PWS were found to be in line with weight as experienced by other family members.
CHAPTER 7: CONCLUSION

7.1 Thesis Summary

The everyday actions which constitute management of PWS within the family result primarily from embodied relations between family members. The processes creating these actions are multi-dimensional, in that family members draw on their own experiential knowledge, existing knowledge e.g. medical science, and material environment. Management practices demonstrate the role of biology in this process, as children with PWS are seen to possess common predetermined, emotional and embodied dispositions e.g. distress triggered by unexpected changes in events, preference for rigid structure in day to day activities, obsessive focus on food and enjoyment of focused simplistic tasks such as puzzles, copying text, computer games. These emotional dispositions guide the actions of children with PWS and are accounted for in the actions of other family members, thus becoming central to everyday management practices.

This research identified four key practices in the management of the PWS body: denial of access-to-food; keeping the child occupied; using rigid routine to structure everyday activities of the child; and biological alteration of the child using human Growth Hormone (hGH). These practices present a holistic image of the PWS child, breaking down the Cartesian mind/body dualism. They demonstrate that managing the body is achieved by addressing every aspect of the person with PWS, including their emotional relation to their environment and social perception. Use of hGH displays human capacity for the design and use of tools to alter biological reality i.e. the child with PWS, in turn altering their emotional, embodied dispositions.

As actions have their base in relations between embodied, emotional individuals, the values and beliefs of these individuals play an important role in the formulation of management strategies. The main guiding values identified were: physical health; emotional health; concerns for the future; and quality of life for all family members. Examination of these values demonstrated the pivotal role of the family as mediator between the child with PWS and wider societal structures. A connection between family values and knowledge existing outside of the family was evident, as medical
definitions of illness are upheld in everyday management and recurrent themes within key values such as ‘inclusion’ parallel government agendas in social care.

The immediate context of the family was seen to impact management practices, most notably, stricter ‘use of routine’ practices were found in families with only children. The impact of managing PWS on other family members emerged strongly through this analysis, particularly parents’ frustration at the ‘hidden’ nature of their management work. Most management practices occurred in families irrespective of socio-economic factors, again displaying the agency of the child with PWS in terms of the demanding nature of their embodied dispositions. There was a similar lack of association between incidence of overweight and obesity and socio-economic status of the families. However weight outcomes tended to reflect body size of other family members, suggesting that despite the embodied dispositions related to PWS they were not excluded from social structuring processes such as the acquisition of habitus.

7.2 Key findings and Discussion

The first key finding and point for discussion is the role of biology in embodied agents, highlighted in this thesis. Individuals with PWS experience the social world in a significantly differentiated way to their peers and siblings without PWS, as was evident throughout the research. This altered perception has its base in the embodiment of these individuals. That is, regardless of their location and experience within micro and macro social structures such as family and class structures, these individuals share common emotional and embodied dispositions, such as differentiated bodily awareness, differentiated social awareness and obsessive focus, most commonly on food. The individuals also share many common biological characteristics such as poor fat: muscle ratio in body composition, decreased bone density leading to scoliosis in extreme cases, altered functioning of the hypothalamus and abnormalities on chromosome 15q11-q13 region, which is currently thought to be the cause of the biological and behavioural differences presented by this group.

Following Merleau-Ponty’s (1962) theory, the embodied nature of the human condition is central to how the social world is perceived and how it is acted upon. Although this process brings together the biological and the social, demonstrating
their connectedness, it highlights the need for recognition of the biological body and its impact on identity and action. Calls for role of the physical/biological body to be examined within social analyses of health and illness have been widespread, culminating in the mid 1990s (see Kelly et al 1996, Williams 1996, Bury 1995). The direct and profound effects of biology on the social identity of the individual are arguably hidden in everyday interactions. The case of children with PWS and the physiological and behavioural qualities unique to their condition, allows the role of biology in impacting social action through the embodied agent to be illuminated.

Bhaskar’s (1989) contribution to critical realist philosophy provides a useful theoretical base for an initial conceptualisation of PWS and its biological aspects. The biology specific to PWS can be viewed as a real ontological state, existing throughout time and space and being governed by ‘mind-independent mechanisms’. However when addressing the management of PWS and the embodied experience of PWS, it is necessary to break with critical realist approaches. As this study of the management of PWS makes apparent the ontological and epistemological converge through the embodied agents (family members), their physical bodies, emotional dispositions, linguistic capacities, beliefs and values, relation to existing cultural knowledge and symbols (medical knowledge, current trend in thinking about disability and obesity) and their use of tools (hGH) to transform the real. These phenomena in management strategies theorized through Burkitt’s (1999) work on embodiment, identity and social action, draw attention to the multi-dimensional network of relations between embodied beings which underpins social action.

As highlighted by Benton (1991), the history of relations and collaborations between the social and biological sciences has been marked by politically contentious movements such as ‘eugenics’ and ‘socio-biology’. So it is with caution to avoid genetic or biological determinism that biology is brought back in to social science research. However, in order to fully capture and understand the experience of rare, multi-system conditions such as PWS, its impact on carers and the social processes involved in its management, the role and agency of biology cannot be ignored.
The second key finding regards the role of family values within the overall management process. Dutch research into the values of caregivers in families of children with PWS identified the four key values of ‘physical health; well being; freedom and responsibility; and a liveable life’ (van Hooren 2005: 311). Values identified from this research displayed similarities to these: physical health, emotional health, considerations for future and quality of life for all family members. As well as identifying key values, analysis uncovered sub values within these such as social inclusion and independence. As in van Hooren’s (2005) findings, values were not always fully achieved due to their contradictory nature. For instance, strategies used to control ‘physical and emotional health’, such as use of rigid routine, were judged by many parents to impair the achievement of values such as ‘considerations for future’ where flexibility was desirable. This finding highlights the complex and demanding nature of care-giving within families. The families in this research were seen to be negotiating several conflicting interests: firstly, between the physical and emotional health of their child; secondly, as promoting long term happiness whilst allowing the child enjoyment on a day-to-day, short term basis; and lastly, negotiating conflict between quality of life for the child with PWS and quality of life for other family members.

The analysis of values added to van Hooren’s model, not just identifying values, but offering a greater explanation of their derivation and role within management overall. Through parental accounts, values were traced to the practices which they were seen to guide. In this way, the influence of everyday embodied interactions on the formation of values was visible. For instance, in parents’ experience of emotional distress in their children. The analysis also drew on the accounts of siblings where relevant, what they considered improved life within the family for their sibling with PWS and how this affected their behaviour.

Building on the analysis of management practices within the families, values- of family members were seen to partially guide management within the households. These values were seen as one aspect of the embodied relations of family members, deriving primarily from everyday practical experience, as well as drawing on wider moral discourse from medical knowledge and political agendas. These findings again
highlight the relevance of Burkitt’s (1999) theoretical framework of embodied social action, displaying the role of knowledge in creating action whilst simultaneously displaying the centrality of lived embodied relations to the overall management process.

The third key finding relates to the nature of the emotional impact of managing PWS on parents. Gray (1993, 2002) explored Goffman’s (1963) notion of ‘courtesy stigma’, as uniquely felt by parents of autistic children. Many of his findings were replicated in this research. The presentation of the child as apparently ‘normal’, in both Gray’s analysis and this thesis, had an extremely stigmatising effect on parents when children displayed anti-social behaviour in public. This study also confirmed Gray’s observation that this stigma was felt and described by mothers more commonly than fathers. Although descriptions of this stigma were described across many families, it was found to particularly affect lone-parents (female) of children with PWS.

As well as confirming the problem of normal appearance of the child in parental experiences of courtesy stigma, for parents of children with PWS a second dimension to this problem was apparent. Normal appearance was not only perceived as a problem where anti-social behaviour was displayed by the child, causing parents to feel that their morality and child-rearing skills were being questioned by onlookers. Normal appearance was also perceived as a problem in terms of the body size of the child. In the majority of cases in this research, children were a healthy weight or overweight, rather than obese. Parents expressed that, in interactions with professionals, family and friends alike the severity the disorder in terms of weight management was doubted. Parents described a stigmatising effect of this as being made to feel as if they were lying, exaggerating or being needlessly overprotective.

In addition to these stigmatising effects, parents drew attention to the lack of recognition they felt they received for the weight management work they undertook which was effectively ‘hidden’ by the relatively normal body size of their children. Hidden work within the family forms an ongoing theme in feminist, economic, political and legal literatures (Coltrane 2000), particularly in relation to caring duties.
(Lawton et al. 2002). In the case of care work in families of children with PWS, this work is hidden not just by the private nature of family life, but by lack of public awareness of the condition combined with the normal appearance of the child.

Although conceptions of stigma have been drawn upon frequently in the field of disability studies (Cahill 1995, Link et al. 2001), they are less commonly seen in the study of disability within the family (Gray 1993, 2002, Scambler et al. 1986, 2004, Voysey 1975). These three studies address specifically courtesy stigma as felt and enacted within families with disabled children and the coping strategies developed. Experiences of stigma in family members of those with PWS display a unique case, adding to findings from this small body of literature.

7.3 Policy and practice
The 2001 white paper, Valuing People: A New Strategy for Learning Disability for the 21st Century, sets out a number of key areas, priorities and changes to services, aimed at improving quality of life for all people with learning disability and their carers. As this section will examine, reflecting on the findings of this research many of the ideas within the white paper are extremely relevant to those with PWS and their families. Implementing structural changes and altering the work of frontline services, as set out in the paper, would appear to be an extremely worthwhile direction to take. As the data that indicate this to be the case were collected two to three years after the publication of the paper, we can acknowledge that the objectives of the strategy have yet to be fully achieved.

The multifaceted nature of PWS and of its management within the family is clearly demonstrated within this thesis. These young people encounter a wide range of physical and behavioural problems, and management of the physical body is mediated via the unique social perceptions and emotional needs of individuals in this group. Calls for social and health services to work in partnership through ‘Learning Disabilities Partnership Boards’, the appointment of specialist learning disability community nurses and more general measures (Department of Health, 2001: 59-69), acknowledge the close relation between learning disability and physical health displayed in this research. In particular, that experiencing learning difficulties and, in
the case of people with PWS autistic spectrum disorders, impacts individuals’ abilities to manage their own health.

‘Person-Centred planning’ is a concept that places the recipient of social care at the centre of the planning process. It derives from the ‘person-centred approach’; a non-directive humanistic approach, which played a part in early developments in social work in the 1950s and 1960s (Rogers 1951, 1961). The (2001: 42, 49) white paper made this the primary technique employed for care planning in local government across the UK. As person centred planning focuses on the needs, beliefs and aspirations of individuals, it has the capacity to acknowledge diversity within the disorder and differences in ways of managing PWS displayed in this research. The white paper (2001: 42) draws particular attention to this planning method for young people with learning disability, during the difficult stage of transition from childhood to adulthood. It also calls for greater integration of child and adult services in health and social care. These measures can also be seen to have a potential benefit for the young people and families in this research. Families saw issues of transition as being particularly problematic, sometimes viewing the immediate emotional needs of their children as being in conflict with their long-term quality of life as adults. An early intervention from care services, involving the person with PWS may help to alleviate the conflict of values felt by families managing PWS.

However, the high levels of emotional intensity and dependency in child/parent relationships highlighted in this research and the key role families play in structuring everyday dietary and health practices, must be taken into account during these person-centred planning sessions. ‘Person-centred planning’ has some obvious benefits in line with the social model disability, maximising autonomy and recognising the rights and choices of individuals. But as these plans are drawn up it is crucial that they are workable within the everyday context of the family and fully appreciate the nature of PWS. As seen in the discussion of autonomy and the social model of disability (chapter five), in many cases the freedom and inclusion of the child with PWS were achieved through particular kinds of intervention and forms of external management, undertaken by the family.
The white paper (2002: 53-58) strategy for supporting carers goes some way towards acknowledging the desirability of including family carers in care planning. A key objective being, ‘They [carers] need to be treated as valued partners by local agencies, not as barriers to their son’s or daughter’s greater independence. (Department of Health, 2001: 53). The introduction of ‘information centres’ and a telephone helpline for carers of those with learning disabilities (Department of Health, 2001: 56), are much needed forms of support. However, the extreme emotional impact of caring for a young person with PWS, illustrated in this research, demonstrates the need to extend this service to include specialist information on rare disorders that include learning disability.

The multi-faceted nature of the disorder and the problems encountered by families trying to manage weight, behaviour and other medical conditions connected to PWS such as scoliosis, point towards the benefits of multi-disciplinary clinics for PWS. The Glasgow multidisciplinary clinic was founded in 1991 and includes PWS specialists in fields of endocrinology (growth), neurology, psychiatry, general medicine, education and dieticians. There is also a multidisciplinary clinic in Bristol which deals with a variety of rare disorders. Awareness of and access to these clinics should be improved amongst the families of those with PWS.

Over recent years the UK government has implemented a strategy to tackle childhood obesity including monitoring children’s weight at school (Department of Health, 2007). The stigma of obesity can pose particular problems for overweight children with PWS and their families. When ‘childhood obesity’ is discussed in the media and government strategies formulated there is a notable failure to acknowledge diverse medical conditions, such as PWS, which can also cause obesity in childhood. Although reports sometimes make vague references to ‘genetic factors’ as a partial cause of obesity, these are seldom discussed in any detail and they usually refer to inherited traits. This means that there is little or no appreciation amongst policy makers and general public alike that people with PWS are far less able than the general population to take actions to reduce their weight. A greater awareness of the PWS and other conditions causing obesity is needed amongst policy makers and front line health and social care professionals alike.
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Appendix 1.

Published Diagnostic Criteria for PWS

Major Criteria
1. Neonatal and infantile central hypotonia with poor suck, gradually improving with age
2. Feeding problems in infancy with need for special feeding techniques and poor weight gain/failure to thrive
3. Excessive or rapid weight gain on weight-for-length chart (excessive is defined as crossing two centile channels) after 12 months but before 6 years of age; central obesity in the absence of intervention
4. Characteristic facial features with dolichocephaly in infancy, narrow face or bifrontal diameter, almond-shaped eyes, small appearing mouth with thin upper lip, down-turned corners of the mouth (3 or more are required).
5. Hypogonadism— with any of the following, depending on age:
   a. Genital hypoplasia, (male: scrotal hypoplasia, cryptorchidism, small penis and/or testes for age (5th percentile); female: absence or severe hypoplasia or labia minora and/or clitoris
   b. Delayed or incomplete gonadal maturation with delayed pubertal signs in the absence of intervention after 16 years of age (male: small gonads, decreased facial and body hair, lack of voice change; female: amenorrhea/oligomenorrhea after age 16)
6. Global developmental delay in a child 6 years of age; mild to moderate mental retardation or learning problems in older children
7. Hyperphagia/food foraging/obsession with food
8. Deletion 15q11-13 on high resolution (650 bands) or other cytogenetic molecular abnormality of the Prader-Willi chromosome region, including maternal disomy

Minor Criteria
1. Decreased fetal movement or infantile lethargy or weak cry in infancy, improving with age
2. Characteristic behavior problems—temper tantrums, violent outbursts, and obsessive-compulsive behavior; tendency to be
argumentative, oppositional, rigid, manipulative possessive, and stubborn;
perseverating, stealing, and lying (5 or more of these
symptoms required)
3. Sleep disturbance and sleep apnea
4. Short stature for genetic background by age 15 (in the absence of growth hormone
intervention)
5. Hypopigmentation—fair skin and hair compared with family
6. Small hands (25th percentile) and/or feet (10th percentile) for height age.
7. Narrow hands with straight ulnar borders
8. Eye abnormalities (esotropia, myopia)
9. Thick viscous saliva with crusting at corners of the mouth
10. Speech articulation defects
11. Skin-picking
Supportive Findings
1. High pain threshold
2. Decreased vomiting
3. Temperature instability in infancy or altered temperature sensitivity in older
children and adults
4. Scoliosis and/or kyphosis
5. Early adrenarche
6. Osteoporosis
7. Unusual skill with jigsaw puzzles
8. Normal neuromuscular studies

To score, major criteria are weighted at 1 point each, and minor criteria are weighted
at 1/2 point each. Supportive findings increase the
certainty of diagnosis but are not scored. For children 3 years of age or younger, 5
points are required, 4 of which should come from the
major group. For children 3 years of age and for adults, a total score of 8 is required
and major criteria must comprise 5 or more points
of the total score.

(Gunay-Aygun et al 2001: 94)
Managing the Body: Health and the Experience of Children with Prader-Willi Syndrome
Aims and objectives of research

- The research aims to fill existing knowledge gaps regarding everyday problems of weight by those with Prader-Willi Syndrome (PWS) and their families during childhood and adolescence, with a particular focus on attention to how they cope with diet-related problems. 

- Even though Guillemin et al. (1998) developed for hyperphagia and the Prader-Willi Association developed for knowledge of PWS and their diet, the actual ways to cope with diet-related problems of weight and body image is not as systematic.

- The research aims to fill many ways families cope with different aspects of weight during childhood.
Aims and objectives of the research

- The research aims to extend existing knowledge of the everyday problems that are faced by those with PWS and their families during childhood and adolescence, paying particular attention to how food is dealt with.
- Even though Guidelines have been developed for the management of hyperphagia/appetite (Prader-Willi Association 2001), knowledge of how people with PWS and their families actually cope with diet and deal with problems of weight gain, and body image is anecdotal rather than systematic.
- The research aims to explore the many ways families cope with different aspects of PWS in childhood.

Who can take part?

Households with PWS children aged 10-15. All kinds of families are welcome to take part in the research, from lone-parents with only children, to households with many children and members spanning more than two generations. However, it is essential that all participants speak basic English language.

The research is funded by the Economic and Social Research Council (ESRC) and the Prader-Willi Association UK, and is based at the University of Leicester.

For further details or an informal discussion please contact the researcher (myself):

Kerry Allen
Tel: 0116 2522829
E-mail: kia3@le.ac.uk
12th February 2004

Dear Miss Jones,

I am writing to you in connection with a forthcoming research project 'Managing the Body: Health and the Experience of Children with Prader-Willi Syndrome'. The project is based at The University of Leicester and funded by the Economic and Social Research Council and the Prader-Willi Syndrome Association (UK).

Twenty families will be recruited for the research and will be visited 3 to 4 times over a year period. This will allow profiles of the families to be developed, in relation to how they manage PWS throughout childhood and adolescence.

Families with a child aged 10-15 years who has PWS are invited to become part of the research. If you and your family are interested in taking part in the research please complete the attached reply slip and return it in the pre-paid envelope provided. Having completed and sent this section you will be under no obligation to proceed with the research if you later decide not to participate.

Yours faithfully,
Kerry Allen  
(Researcher, University of Leicester)  
(UK))

Narinder Sharma  
(Chief Executive, PWSA)

Parent name(s):

Full postal address:

Telephone number:

e-mail (if applicable):

PWS child’s age:

Other children’s age(s) (if applicable):
Dear Mrs Jones,

Many thanks for the interest you have shown in the research. A preliminary sample for the project has now been drawn and if you are still willing I would be very interested in including your household in this sample. At the moment I am in the process of setting up some initial visits. The visit will last roughly two hours and will involve:

1. Collection of some basic background information about Helen e.g. medical history, type of school she attends etc
2. An informal chat about the daily routines of your household (especially food related routines)
3. A session with Helen- 'Getting to know you' activities such as drawing and discussion based around hobbies and interests.

The only people that will need to be involved this initial session are yourself and Averil. I am happy to visit at evenings and weekends. I will be available between April 22nd to May 17th, then from May 24th to May 31st.

Do let me know a date and time that would suit you (home phone with 24 hour answer service -0116 244 8562). If you have any further questions don't hesitate to get
in touch. I will be away between 4\textsuperscript{th} April and 22\textsuperscript{nd} April but feel free to leave a message on my answer phone.

Best wishes and look forward to meeting you all
Kerry Allen

(Researcher, Leicester University)
Appendix 5. Informed consent form

RESEARCH SUBJECT INFORMED CONSENT FORM

Prospective Research Subject: Read this consent form carefully and ask as many questions as you like before you decide whether you want to participate in this research study. You are free to ask questions at any time before, during, or after your participation in this research.

Project Information

| Project Title: Managing PWS within families |
| Investigator: Kerry Allen                  |
| Location: University of Leicester         |
| Phone: 0116 252 2829                      |

PURPOSE OF THIS RESEARCH STUDY

- You are being asked to participate in a research study designed to explore the experiences of families with a child with PWS aged 11-15. The research focuses on how families manage all aspects of the disorder, with a particular focus on everyday dietary management.

PROCEDURES

- You will be asked to participate in three or four family case studies. These involve interviews and observations within the family home.
- The expected duration of subject's participation is three years.

CONFIDENTIALITY

Your identity in this study will be treated as confidential. The results of the study, may be published but will not give your name or include any identifiable references to you.

TERMINATION OF RESEARCH STUDY

You are free to choose whether or not to participate in this study. There will be no penalty or loss of benefits to which you are otherwise entitled if you choose not to participate. You will be provided with any significant new findings developed during the course of this study that may relate to or influence your willingness to continue participation. In the event you decide to discontinue your participation in the study,

AVAILABLE SOURCES OF INFORMATION

- Any further questions you have about this study will be answered by the Investigator:
Name: Kerry Allen
Phone Number: 0116 252 2829

AUTHORIZATION

I have read and understand this consent form, and I volunteer to participate in this research study. I understand that I will receive a copy of this form. I voluntarily choose to participate.

Participant Name (Printed or Typed):
Date:
Participant Signature:
Date:
Investigator Signature:
Date: