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Research Project Report
(C85RES)

University of Nottingham
Institute of Work, Health and Organisations

Doctorate in Clinical Psychology

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Assessing the Impact of Hyperphagia on the Behaviour of Children with Prader-Willi Syndrome

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Submitted in part fulfilment of the requirements for the

Doctorate in Clinical Psychology
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Abstract

Background: Prader-Willi Syndrome (PWS) is a genetic disorder, characterized by: hypotonia, short-stature, craniofacial abnormalities, hypogonadism, hypersomnolence and developmental delay. However, it is best known for hyperphagia (excessive appetite) and behavioural problems. In the past, there has been limited research examining the relationship between hyperphagia and behavioural problems in PWS, as the difficulties were viewed as distinct phenotypes and the measurement of Hyperphagia was challenging. However, Dykens et al. (2007) recently devised the Hyperphagia Questionnaire (HQ), to measure hyperphagia severity, drive and behaviour in PWS. The primary aim of the current study was to use the HQ to explore how well levels of hyperphagia could account for a variety of behavioural and emotional problems in children with PWS, whilst controlling for potentially mediating factors such as: age, gender, IQ and weight. It was hypothesised that there would be an association between hyperphagic drive and severity (level of food-related distraction and distress) and disruptive behaviour, anxiety, self-absorbed behaviour and social relating problems.

Method: Following ethical approval, 350 questionnaire packs were sent by post to parents and carers of children with PWS (aged 4-18 years) via the PWS-Association. Data was collected on: age, gender, weight, hyperphagia and behavioural and emotional problems of children with PWS. A total of 105 responses were received (30%). Following this 19 were excluded from the analysis. Of the remaining 86 children included in the study, 60% were male, with a mean age of 9.63 years (SD: 4.19).

Results: Initial analyses were conducted to confirm that data met the criteria for parametric tests. Bivariate correlational analyses were then performed to determine which variables were suitable for entry into regression. Following this a number of multiple regressions were conducted to examine how well hyperphagic drive and behaviour predicted emotional and behavioural problems (whilst controlling for potentially mediating variables such as age, gender, and weight). Hyperphagic drive significantly predicted levels of antisocial/disruptive
behaviour, anxiety, social relating problems, communication disturbances and self-absorbed behaviours. Whilst hyperphagic behaviour did not significantly predict any behavioural/emotional problems.

Conclusions: The results suggested an association between hyperphagia and a variety of behavioural and emotional problems in children with PWS, with those with higher levels of hyperphagic drive experiencing more problems. This finding reinforces previous research, which has suggested an association between hyperphagia and non-food related behavioural problems in PWS (Dykens et al, 2007). However, the factor structure of the HQ was not supported in this study. This suggests that it requires further validation and exploration. Furthermore, the HQ was only able to assess the behavioural and emotional expression of hyperphagia, not the internal experience of it. Therefore more research into hyperphagia in PWS is required. Despite this though, these findings have important implications for the understanding of behavioural problems in people with PWS. In particular, finding a link between hyperphagia and behavioural problems could lead to the development of bio-psycho-social interventions, which consider both problems together, rather than the current practice of treating the hyperphagia and behavioural problems separately.
Statement of Contribution

The author of this study was responsible for the project design, applying for ethical approval, reviewing the literature, data collection, scoring questionnaires, entering data and data analysis. However, support and guidance was gratefully received on all areas from my clinical and academic supervisors; Dr Shirley Thomas, Dr Roshan das Nair and Dr Martha-Laxton Kane. I would like to thank them for sharing their expertise and for providing support and guidance throughout the completion of this research.

I would also like to thank everyone at the Prader-Willi Syndrome Association, in Derby who helped in the identification and recruitment of all research participants. Furthermore, I would like to acknowledge the many parents and carers who agreed to take part in this study, without whom this study would not have been possible.

Finally, a very big thank-you goes to all of my family and friends, who have been an invaluable source of support for me over the two years. In particular: my partner Danny, my Mum, and my great friends Lisa and Rosie. A special mention also goes to my grandparents, whose boundless generosity and enthusiasm made my whole education possible.
Research Paper

Abstract

Background

Prader-Willi Syndrome (PWS) is a complex genetic syndrome associated with hyperphagia and behavioural problems. Recent research suggested a link between hyperphagia and behavioural and emotional problems in PWS such as anger and anxiety. The current study aimed to explore this relationship further.

Method

Through parental report postal questionnaires, data was collected on the age, gender, weight, hyperphagia and behavioural and emotional problems of 105 children with PWS aged 4-18 years (M: 9.63 years).

Results

Following preliminary analysis, a series of multiple regressions were performed. Hyperphagic drive significantly predicted antisocial/disruptive behaviour, anxiety, social relating problems, communication disturbances and self-absorbed behaviours. Whilst hyperphagic behaviour did not significantly predict any behavioural/emotional problems.

Conclusions

This study reinforces research which has suggested an association between hyperphagia and non-food related behaviour in PWS. This has implications for the understanding of PWS and the development of psychological interventions for behavioural and emotional problems.

Keywords: Prader-Willi Syndrome; Challenging Behaviour; Behaviour; Hyperphagia
Prader-Willi Syndrome (PWS) is a complex genetic disorder caused by abnormalities on chromosome 15q11-q13 (Cassidy et al., 1984; Curfs et al., 1995). It affects males and females equally and occurs in one in 15,000 to 30,000 births (Cassidy & Driscoll, 2009; Couper & Couper, 2000). The primary physical characteristics are; hypotonia, short-stature, craniofacial abnormalities, hypogonadism, hypersomnolence, and intellectual disabilities (ID). These are caused by widespread central nervous system, endocrine gland and hypothalamic dysfunction (Curfs et al. 1995; Goldberg et al. 2002). However, the syndrome is perhaps best known for the food-related symptoms related to hypothalamic dysfunction. These include: hyperphagia (excessive appetite), food-preoccupations and foraging (Dykens & Kasari, 1997).

(See Section 1.1 of the Extended Paper for further description of the causes, diagnosis and symptoms of PWS)

Hyperphagia generally presents between the ages of 18months and six-years (Cassidy, 1984; Clarke, et al. 1996; Dimitropoulos et al. 2000) and is attributed to the fact that individuals with PWS possess unusually high levels of the appetite stimulating hormone Grehlin (Haqq et al. 2003). Individuals also experience reduced satiation responses from the impaired functioning of the hypothalamus (Swaab, 1997). Hyperphagia is widely considered to be the most debilitating aspect of PWS (Kundert, 2008). Consequently, without dietary management; individuals often become obese (Dykens, et al. 1996).
Lindgren et al. (2000) explored the hyperphagia in PWS by comparing the eating of children with PWS (n=9) with healthy-weight (n=20) and obese controls (n=20), and then measuring the time taken to eat a meal. A Kruskal-Wallis 1-way analysis of variance was conducted to examine the differences in eating rates between groups, and ‘typical eating curves’ were calculated. Both control groups showed ‘decelerating’ eating curves (high initial rate, gradually slowing until satiation occurred) whilst the PWS group showed ‘non-decelerating’ curves (consistently high rate) indicating difficulty reaching satiation. However, as participants were given a set meal, it could not be calculated whether satiation could be reached.

In addition to this, there is evidence for a specific behavioural phenotype (Clarke et al. 1996; Clarke et al. 2002; Dykens & Kasari, 1997; Hiraiwi, et al. 2007). Key problems identified are: temper-tantrums, stubbornness, argumentativeness, and inflexibility (Clarke et al.1998; Dykens et al. 1996; Steinhausen et al. 2004). Such difficulties become evident in childhood and are more prevalent among people with PWS than participants matched for age, gender and ID (Clarke et al. 1998; Reddy et al. 2007). Dykens and Kasari (1997) measured the behaviour of children with PWS (n= 43) and compared them with age-and-gender matched children with Down Syndrome (DS, n= 43) and Non-Specific ID (NS, n= 43). The PWS group displayed significantly more frequent and severe behavioural problems, with 72% scoring over the clinical cut-off (compared to 23% of the DS group and 39% of the NS group).
Such behavioural problems have been found to be associated with age, with older children and adults displaying higher levels (Dimitropolous et al. 2001; Dykens, 2004). Steinhausen et al. (2004) examined behaviour in people with PWS aged two to 29 years. They found the prevalence of disruptive behaviours was significantly higher for the older group (over 13 years) than the younger group (under 13 years). This is consonant with research on behaviour in children with other ID (Einfeld & Tonge, 2002). Level of ID has also been found to be associated with behaviour problems in PWS, with those with higher levels of ID displaying more emotional and behavioural problems (Einfeld et al. 1999).

However, gender has not been consistently found to be associated with behavioural problems in PWS. Some have found no relationship between gender and behaviour (Dykens, et al. 1991; Steinhausen, et al. 2004). However, others found males display significantly more externalising and disruptive problems, and that females display more anxiety (Dykens & Cassidy, 1995; Dykens, 2004). Finally, one further factor which may be associated with behaviour in PWS is weight. Ackefeldt and Gillberg (1999) found no association between Body Mass Index (BMI) and behavioural problems in PWS. However, Steinhausen et al. (2004) found a significant relationship between challenging behaviour and BMI in PWS, with those with higher BMIs displaying more behaviour problems.

(See Section 1.2 of the Extended Paper for further description of Behavioural and Emotional Problems in PWS)

There has been little research examining the impact of hyperphagia on individuals with PWS or its association with behavioural and emotional
problems. However, Holland et al. (2003) stated that: “Although people with PWS are not starved, their behaviour and much of the associated physiology is as if they are in a state of starvation” (p.990). Research into the reactions of healthy individuals to food restriction demonstrates that experiencing hunger over long periods can cause a variety of psychological, behavioural and cognitive changes (Keys et al. 1950; Polivy, 1996; Weinreb et al. 2002). For example, Keys et al. (1950) took a group of 50 healthy weight participants and restricted food intake by 25% over six-weeks. The men were observed becoming increasingly ‘obsessed’ with food. They also reported increased anxiety, irritability and depression, leading to increased conflict with people around them. Furthermore, the symptoms persisted for sometime after the study ended. Similar psychological effects have been found in studies of long-term dieters and children who are subjected to hunger through social deprivation (Polivy, 1996; Weinreb et al. 2002). These effects were reflected in a qualitative study involving eight adults with PWS (Haselip, 2006), which revealed that all participants considered coping with incessant hunger the most difficult aspect of having the syndrome. Participants attributed feelings such as sadness, anger and anxiety to feeling continually hungry. (See Section 1.3 of the Extended Paper for a discussion of the implications of examining the link between hunger and behaviour)

One reason for the limited research into the impact of hyperphagia on people with PWS has been that the measurement of hyperphagia is complex (Dykens et al. 2007). Self-report measures are generally not utilised, as studies have indicated that people with PWS cannot reliably report on their eating (Young et al. 2006). Furthermore, it has been suggested that individuals with ID are more
likely to be affected by response biases such as social desirability (Jones et al. 1997).

Therefore, at this time the only method of measuring hyperphagia is to assess how people with PWS behave around food. In the past, researchers have utilised eating disorder inventories or observational methods for this (Sarimski, 1996; Young, et al. 2006). However, recently attempts have been made to devise more reliable and valid methods with a view to examining links with non-food related behaviour (Dykens, et al. 2007; Russell & Oliver, 2003). Russell and Oliver (2003) devised the ‘Food-Related Problems Questionnaire’ (FRPQ), a 16-item informant response questionnaire designed through focus groups with parents of individuals with PWS, in which the common food-related problems observed in PWS were grouped together. These were then divided into three subscales: ‘Preoccupation with food’ (e.g. excessive talking about food), ‘Impairment of satiety’ (e.g. complaining of feeling hungry) and ‘food-related negative behaviours’ (e.g. stealing, pica).

However, on the FRPQ items required verbal descriptions from the individual about their hunger and/or satiety (e.g. “how often will the person say they still feel hungry?”). It would be questionable whether some individuals would be able to do this. As a result of this limitation, Dykens et al. (2007) devised the Hyperphagia Questionnaire (HQ), a parental report measure which, like the FRPQ also assessed levels of hyperphagia as observed through expressions and/or behaviour.
Individual items for the HQ were devised from clinical work with parents and carers of people with PWS and from the definitions in the fourth edition of the Diagnostic and Statistical Manual of Mental Disorders (American Psychiatric Association, 2000). Following this, the measure’s psychometric properties were assessed using a sample of parents and carers of children and adults with the syndrome (n=153) aged from four to 51 years. Exploratory factor analysis revealed a three factor structure, leading to the creation of three subscales, labelled: ‘Hyperphagic Behaviour’ (frequency of stealing, foraging and bargaining for food), ‘Hyperphagic Drive’ (severity of distress when denied food and the ease of re-directing from food-related activities), and ‘Hyperphagic Severity’ (overall time engaged in food-related behaviour and the impact of food-related distraction on daily functioning).

Dykens et al. (2007) used the HQ to explore the relationship between hyperphagia and non-food related behaviour. Analysis revealed moderate positive correlations between hyperphagic drive and severity and internal difficulties (e.g. anxiety/depression and withdrawal). Whilst hyperphagic behaviour, drive and severity were all correlated with non-compliant behaviour and aggression. These results suggested a relationship between hyperphagia and behavioural and emotional problems in people with PWS.

However, there were some limitations. Firstly, Dykens et al. (2007) recruited a sample aged four to 51 years and used the same measures throughout. The challenging behaviour measure used (Checklist of Challenging Behavior: Achenbach, 1991) was developed for typically developing children and is not
validated with adults. The Developmental Behaviour Checklist (DBC-P - Einfeld & Tonge, 1992, 1995, 2002) may have been more suitable, as it is a similar, well validated measure of emotional and behavioural problems, however it was developed specifically for people with ID (Einfeld et al. 2002). Therefore, all items relate to behaviour commonly observed in children with ID and exclude questions which may be attributed to disabilities (e.g. does not speak). The subscales of the DBC-P are: Antisocial/Disruptive behaviour (e.g. Manipulative; Abusive; Irritable), Self-Absorbed behaviour (e.g. Preoccupied), Communication Disturbance (e.g. Echolalia; Perseveration), Anxiety (e.g. Fears, phobias; Cries) and finally Social Relating (e.g. Aloof; Unhappy).

A second limitation is that it has been suggested that the hyperphagic expression of PWS changes throughout the lifespan, with increasing hyperphagia in childhood and decreasing hyperphagia in middle-age (Descheemaeker et al. 2002). This indicates that the correlations between may have been better examined by age, to provide more detailed information about the relationship between hyperphagia and behaviour at different ages.

Although Dykens et al. (2007) have made progress towards examining the relationship between behavioural difficulties and hyperphagia in PWS, further research is necessary to address sampling and measurement problems and explore the relationship with more in-depth analysis. Furthermore, the HQ requires further validation. Therefore, the primary aim of this study was to build on the work of Dykens et al. (2007) to use the HQ and the DBC-P to examine how well hyperphagia accounts for levels of emotional and behavioural
problems in children with PWS, whilst also controlling for potentially mediating factors such as: age, weight, gender and IQ. This was to establish whether there was a relationship between hyperphagia and non-food related behaviour. Furthermore, the secondary aim was to explore the validity and reliability of the HQ through exploratory factor analysis.

It was hypothesised that there would be an association between levels of hyperphagia and non-food related behavioural and emotional problems displayed. More specifically, it was hypothesised that:

1. Increased levels of hyperphagic drive and severity (food related distraction and distress) would be associated with higher levels of non-food related antisocial/disruptive behaviour, resulting from hyperirritability associated with chronic hunger.

2. Increased levels of hyperphagic behaviours (e.g. stealing food, foraging) would be associated with increased antisocial/disruptive behaviours.

3. Increased hyperphagic drive and severity would be associated with high levels of Self-Absorbed behaviour, as hunger is associated with increased withdrawal.

4. Increased hyperphagic drive and severity would be associated with higher scores on the anxiety subscale, as hunger is associated with increased anxiety.

5. Increased hyperphagic drive and severity would be associated with higher levels of social relating problems (aloof, unhappy), as hunger has been found to relate to low-mood and depression.
6. Increased hyperphagic drive, severity and behaviour would not be associated with communication impairments, as these difficulties are more commonly observed in young people with severe ID (Einfeld et al. 2002).

7. Hyperphagic behaviour would not be associated with scores on the anxiety, self-absorbed behaviour and social relating subscales.

Examining the link between hyperphagia and behaviour in people with PWS could aid clinical practice as many psychological interventions currently address behavioural problems only and not hyperphagia (Luiselli, 1988). As such, they may not provide individuals with a sense of control over the syndrome (Singh et al. 2008). If a link between hyperphagia and behaviour was identified, then interventions could be modified to incorporate support with managing the impact of the hyperphagia on behaviour, in a similar manner to cognitive-behavioural interventions for chronic-health problems (White, 2001). Such interventions could provide individuals and their families with skills in managing the physical, emotional and behavioural impact of PWS.

(See Section 1.3 of the Extended Paper for an extended Clarification of the aims and rational for the study)

**Method**

**Design**

This study employed a cross-sectional design in which parents of children with PWS were asked to complete a battery of postal questionnaires assessing hyperphagia and behavioural and emotional problems.
Participants

Parents and carers of children with PWS were recruited for this study, rather than children with PWS themselves, as no self-report measures for hyperphagia have been developed. Participants were recruited via the Prader-Willi Syndrome Association for the UK (PWSA-UK) who hold a database of parents and carers of children with PWS, who had previously consented to being contacted by the association regarding research.

To be included, participants were required to have at least one child diagnosed with PWS aged 18 years or under at the time of testing.

Following this, participants were excluded if:

- The child did not reside with them; as they may not be able to reliably comment on their recent food intake or challenging behaviour.
- The child was under four-years old; as they would be less likely to experience hyperphagia (Cassidy, 1984; Russell & Oliver, 2003).

A total of 350 potential participants from the database were contacted for the current study.

Measures

Demographic Questionnaire

Demographic child and parent information was obtained using a locally developed nine-item Demographic Questionnaire (Extended paper appendix 1.4), which assessed: the participant’s relationship to the child, where the child
resides, age, gender, age at diagnosis, type of school attended, weight, height, and IQ.

Hyperphagia Questionnaire (HQ - Dykens et al. 2007)

The HQ is a brief 11-item informant measure that focuses on food related problems in individuals with PWS. Items are rated on a 5-point Likert scale by either the severity or the frequency of the problem. For example: responses range from 1=not a problem, up to 5=severe and/or frequent problem; or from; 1=never, up to 5=4-7 times a week. The HQ was developed through focus groups with parents of people with PWS. The psychometric properties of the measure were assessed using a sample of parents and carers of individuals with PWS (n=153) aged four to 51 years (Dykens et al. 2007). Factor analysis of the measure revealed a three factor structure assessing three key components of hyperphagia labelled: hyperphagic behaviour, hyperphagic drive and hyperphagic severity. These factors accounted for 58.93% of variance and also possessed acceptable levels of internal consistency (Cronbach’s alpha ranging from.60-.80). However, as this was a new measure, it was planned that the factor analysis would be re-run and further reliability analysis would be conducted with this sample.


The second edition of the DBC-P is a 96-item informant response questionnaire designed to assess behavioural and emotional disturbances in children with intellectual disabilities (aged four to 18 years). Parents and carers indicate the
extent to which items apply to their child using a 3-point Likert scale (responses may be 0=not true; 1=somewhat true or sometimes true; 2=very true or often true). The measure then provides a ‘Total Behaviour Problem Score’, which provides an indication of the severity of any behavioural/emotional disturbances. The measure also provides scores on the five behavioural subscales: Disruptive/Antisocial, Self-Absorbed, Communication Disturbance, Anxiety, Social Relating.

This scale was originally developed in Australia as an instrument for assessing the psychopathology of children with intellectual disabilities. However it has been used extensively within Europe and with PWS samples (Clarke et al. 2002; Einfeld, et al. 1999). Extensive reliability and validity analyses have been conducted on the DBC-P. The measure has been found to have good internal consistency and inter-rater and test-retest reliability (Einfeld & Tonge, 1992). Furthermore, it was shown to have high criterion group validity in differentiating clinical from non-clinical cases and strong criterion and concurrent validity (Dekker, et al. 2002; Einfeld & Tonge, 1992, 1995, 2002). In a recent study using Australian and European samples, the factor structure of the five subscales was found to be strong, accounting for 44% of the total variance (Dekker, et al. 2002). The internal consistency of the measure has been found to be good, with Cronbach’s alpha of .94 reported on the total problem behaviour score (Einfeld & Tonge, 1992). In this sample, Cronbach’s alpha on the total problem behaviour score was .94, and on the five subscales it ranged between .69-.90.

(See Section 2.1 of the Extended Paper for further discussion of the Measures)
Procedure

Following favourable ethical approval from the Institute of Work, Health and Organisations Ethics Committee at the University of Nottingham (See Extended paper Appendix 3.0), 350 questionnaire packs were dispatched to the PWSA-UK. These packs were then addressed to parents and carers on the database by the PWSA-UK and sent by post. Each questionnaire pack included: a letter of invitation from the PWSA-UK introducing the researchers, an information sheet providing comprehensive details about the study aims and objectives, a consent form, and the three measures. (See Extended paper Appendix 2.1 to 2.5)

(See section 2.2 of the Extended Paper for further discussion of the key ethical points considered)

Those parents and carers who wished to take part in the study were requested to sign the consent form and complete the demographic sheet and two self-report questionnaires (the HQ and the DBC-P). This was estimated to take between 25-30 minutes. Participants were requested to return the completed forms using a prepaid reply envelope, within two months of receiving the questionnaire pack.

Analysis

Data was analysed using SPSS™ for Windows version 16.0 (SPSS Inc. 2008). All statistical tests were two-tailed and the significance level was defined at $p \leq .05$. Initial exploration of the data was conducted to confirm that the data met the assumptions of parametric tests.

(See Sections 3.2 and 3.4 of the Extended Paper for a description of the data screening process)
To address the main aims, first Exploratory Factor Analysis was completed on the HQ to explore the factor structure and assess reliability. Following that, bivariate correlations were calculated between the subscales of the HQ and the subscales of the DBC-P to assess whether investigation of the relationship between hyperphagia and various forms of behavioural and emotional problems would be relevant using Regression Analysis. Bivariate correlations were also calculated to examine the relationship between the subscales of the DBC-P and other potentially moderating demographic factors (such as age, weight category, gender, or IQ). Following this, any significantly correlated predictors of behavioural problems were entered into a series of multiple regression analyses.

**Results**

**Sample**

Responses were received from 105 parents of children with PWS (30% response rate). Following this, 15 (14%) were excluded from the study as their child was under four years, and three (3.8%) were excluded because their child did not reside with them. One further case was removed from the dataset, as over 50% of data points were missing.

*(See section 3.1 of the Extended paper for a participant recruitment flow chart)*

This left 86 parents and carers of children with PWS. Of these, 86% were the child’s mother, whilst 13% were the child’s father and 1% was a step-parent.
The children included in the study were aged between four and 18 years (M: 9.63 years, SD: 4.19 years) and 60% were male (See Table 1 for a summary).

Based on the age, gender, height and weight information provided by parents, children were categorised into either a ‘normal-weight’ or ‘overweight/obese’ category (Cole et al. 2000). Of the children included in the study, 62.2% were categorised as being overweight or obese, with Body Mass Indices ranging from 14 to 50 (M: 24.18, SD 8.14).

The mean Total Behaviour Problem Score from the DBC-P was 50.36 (SD: 25.87), which is above the cut-off point for clinically significant emotional and behavioural difficulties (Einfeld & Tonge, 2002).

Initial exploration of the data revealed that, only seven respondents provided information on their child’s IQ. Therefore, this variable could not be included in further statistical analysis.

(See section 3.2 of the Extended Paper for further description of the study population)

Table 1

<table>
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<th>Characteristics of the sample meeting inclusion criteria</th>
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<td>Mean Age in years (SD)</td>
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<td>9.63 (4.19)</td>
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Factor Analysis of the HQ

Factor analysis using Principle Components Analysis was used to examine the scaling properties of the HQ in this sample. Preliminary analysis was conducted, revealing the presence of many inter-item correlations above the recommended level of .3 (Pallant, 2007). Furthermore, no correlations were above .9 indicating no problems with multi-collinearity. The Kaiser-Meyer-Oklin Measure of Sampling Adequacy was .87, over the recommended value of .6 (Kaiser, 1974). This supported the suitability of the sample for factor analysis. Finally Bartlett’s test of sphericity was statistically significant (p<.001) supporting the factorability of the correlation matrix.

(See section 3.3 of the Extended Paper for the correlation Matrix, Eigen values and Scree Plot)

Initial results revealed two factors with eigenvalues over one, explaining 59.99% and 11.45% of variance respectively. Furthermore, inspection of the scree plot using Cattell’s scree test revealed a clear plateau after the second component (Field, 2005). Therefore, it was decided to retain two components for further analysis.

As the two components both explored aspects of hyperphagic behaviour, they were considered likely to be interrelated. Therefore, oblique ‘Direct Oblimin’ rotation was performed. This revealed a simple two factor structure, with both components demonstrating strong loadings (above .4) and items generally loading onto only one component (Table 2). There was a moderate positive correlation between the two factors (r=.534).
### Table 2

**Pattern and Structure Matrix for Principle Components Analysis of the Hyperphagia Questionnaire Items**

<table>
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<th>Item</th>
<th>Pattern Matrix</th>
<th>Structure Matrix</th>
<th>Communalities</th>
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<tr>
<td>9. When others try to stop your child from talking about food or engaging in food-related behaviours, it generally leads to:</td>
<td>.936</td>
<td>.098</td>
<td>.883</td>
</tr>
<tr>
<td>1. How upset does your child become when denied a desired food?</td>
<td>.930</td>
<td>-.127</td>
<td>.862</td>
</tr>
<tr>
<td>3. Once your child has food on their mind, how easy is it for you or others to re-direct your child away from food to other things?</td>
<td>.920</td>
<td>-.058</td>
<td>.889</td>
</tr>
<tr>
<td>6. How persistent is your child in asking or looking for food after being told &quot;no&quot; or &quot;no more&quot;?</td>
<td>.779</td>
<td>.138</td>
<td>.853</td>
</tr>
<tr>
<td>11. To what extent do food related thoughts, talk or behaviour interfere with your child's normal daily routines, self-care or schoolwork?</td>
<td>.771</td>
<td>.053</td>
<td>.799</td>
</tr>
<tr>
<td>7. Outside of normal mealtimes, how much time does your child spend talking about food or engaged in food related behaviours?</td>
<td>.639</td>
<td>.211</td>
<td>.752</td>
</tr>
<tr>
<td>2. How often does your child try to bargain or manipulate to get more food at meals?</td>
<td>.617</td>
<td>.296</td>
<td>.775</td>
</tr>
<tr>
<td>How often does your child get up at night to food seek?</td>
<td>-.131</td>
<td>.913</td>
<td>.357</td>
</tr>
<tr>
<td>4. How often does your child forage through the trash for food?</td>
<td>.045</td>
<td>.762</td>
<td>.452</td>
</tr>
<tr>
<td>10. How clever or fast is your child in obtaining food?</td>
<td>.424</td>
<td>.556</td>
<td>.357</td>
</tr>
</tbody>
</table>

*Note: Major loadings for each item are shown in bold*

These results raised questions about the three factor structure found by Dykens et al. (2007) and as a result the new components were retained. Component one contained items, which related to individual’s expression of hyperphagia and their general drive for and/or preoccupation with food day-to-day, e.g. how often they talk about food and how upset they become when denied food. This component retained all items formerly in Dykens et al’s (2007) ‘Hyperphagic Drive’ subscale, with the addition of item two (from the ‘Hyperphagic Behaviour’
subscale) and both items (eleven and seven) from the ‘Hyperphagic Severity’ subscale. As all items still appeared to describe ‘Hyperphagic Drive’, the label was retained for component one.

Component two, on the other hand contained items which appeared to describe the child’s tendency to act on their hyperphagia, e.g. how often they steal/forage for food. This component retained all items formerly in the ‘Hyperphagic Behaviour’ Subscale except for item two which was moved into the Hyperphagic Drive subscale as it loaded more strongly onto component one. Furthermore, on examination it appeared to fit better as a measure of ‘drive’, as it was not about actions, it was about expressing a desire for food (more in keeping with items three, six and eleven in component one). Component two retained the label ‘Hyperphagic Behaviour’ as the four items reflected food-related behaviour (e.g. stealing, foraging).

On completion of the factor analysis, the internal consistency of the two new subscales was assessed. The Cronbach’s alpha for the new seven item ‘Hyperphagic Drive’ subscale was .923 and for the four item ‘Hyperphagic Behaviour Subscale’ it was .854. These figures suggested good levels of internal consistency (Field, 2005).

*Initial exploration*

Initial exploration of the data with Pearson’s correlations revealed that Hyperphagia total score was significantly positively related to the TBPS ($r (81) = .585, p<.01$). Therefore, further investigation of the relationship between hyperphagia and behaviour was supported.
Following this, a number of bivariate correlations were conducted to assess whether or not proposed predictor variables (Hyperphagic Drive, Hyperphagic Behaviour, Age, Gender and Weight Category) were associated with the five behavioural subscales of the DBC-P. The results are shown in Table 3.

### Table 3

**Results of the Bivariate Correlation Analyses**

<table>
<thead>
<tr>
<th>Variable</th>
<th>Mean</th>
<th>SD</th>
<th>N</th>
<th>2.</th>
<th>3.</th>
<th>4.</th>
<th>5.</th>
<th>6.</th>
<th>7.</th>
<th>8.</th>
<th>9.</th>
<th>10.</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Age</td>
<td>9.63</td>
<td>4.19</td>
<td>86</td>
<td>.071</td>
<td>.097</td>
<td><strong>.225</strong></td>
<td><strong>.398</strong></td>
<td><strong>.360</strong></td>
<td>.056</td>
<td>.031</td>
<td>-.009</td>
<td>.192</td>
</tr>
<tr>
<td>2. Gender ($r_{pb}$)</td>
<td>-</td>
<td>-</td>
<td>75</td>
<td>.249*</td>
<td>.085</td>
<td>.141</td>
<td>.045</td>
<td>.098</td>
<td>.075</td>
<td>.048</td>
<td>.012</td>
<td></td>
</tr>
<tr>
<td>3. Weight Category ($r_b$)</td>
<td>-</td>
<td>-</td>
<td>66</td>
<td><strong>.269</strong></td>
<td><strong>.290</strong></td>
<td><strong>.295</strong></td>
<td>.191</td>
<td>.038</td>
<td>.030</td>
<td><strong>.290</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4. Hyperphagic Drive</td>
<td>18.13</td>
<td>7.29</td>
<td>85</td>
<td>.689**</td>
<td>.609**</td>
<td>.547**</td>
<td><strong>.367</strong></td>
<td><strong>.392</strong></td>
<td><strong>.414</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5. Hyperphagic Behaviour</td>
<td>7.42</td>
<td>4.40</td>
<td>78</td>
<td>.476**</td>
<td>.422**</td>
<td>.173</td>
<td>.191</td>
<td>.299**</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>6. Antisocial/Disruptive</td>
<td>15.55</td>
<td>9.10</td>
<td>83</td>
<td>.563**</td>
<td>.607**</td>
<td>.537**</td>
<td><strong>.580</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>7. Self-Absorbed</td>
<td>13.36</td>
<td>8.62</td>
<td>83</td>
<td>.645**</td>
<td>.586**</td>
<td>.668**</td>
<td><strong>.684</strong></td>
<td><strong>.554</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>8. Communication Disturbance</td>
<td>8.83</td>
<td>4.66</td>
<td>83</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>9. Anxiety</td>
<td>4.41</td>
<td>2.85</td>
<td>83</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td><strong>.461</strong></td>
</tr>
<tr>
<td>10. Social Relating</td>
<td>5.25</td>
<td>3.30</td>
<td>83</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*p<.05; **p<.01; ***p<.001

*Note.* $r_{pb}$ indicates Point-biserial correlation coefficient and $r_b$ indicates Biserial correlation coefficient.

(See Section 3.5 of the Extended Paper for extended description of these correlations)

### Regression Analysis

Following correlational analysis, a number of linear regressions were then conducted using the five behavioural subscales of the DBC-P as outcome variables and any factors, which were significantly related to them as predictor variables. The results are summarised in Table 4 below.
<table>
<thead>
<tr>
<th>Outcome Variable</th>
<th>Predictor Variable(s)</th>
<th>Unstandardised Beta</th>
<th>Std. Error</th>
<th>Standardised Beta</th>
<th>t</th>
<th>Sig.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Disruptive/Antisocial Behaviour</td>
<td>Age</td>
<td>.518</td>
<td>.242</td>
<td>.238</td>
<td>2.14</td>
<td>.037</td>
</tr>
<tr>
<td></td>
<td>Weight Category</td>
<td>2.63</td>
<td>2.02</td>
<td>1.40</td>
<td>1.30</td>
<td>.199</td>
</tr>
<tr>
<td></td>
<td>Hyperphagic Drive</td>
<td>.672</td>
<td>.176</td>
<td>.539</td>
<td>3.81</td>
<td>.001</td>
</tr>
<tr>
<td></td>
<td>Hyperphagic Behaviour</td>
<td>-.067</td>
<td>.313</td>
<td>-.033</td>
<td>-.215</td>
<td>.831</td>
</tr>
<tr>
<td>Self-Absorbed</td>
<td>Hyperphagic Drive</td>
<td>.577</td>
<td>.159</td>
<td>.488</td>
<td>3.63</td>
<td>.001</td>
</tr>
<tr>
<td></td>
<td>Hyperphagic Behaviour</td>
<td>.168</td>
<td>.264</td>
<td>.086</td>
<td>.636</td>
<td>.527</td>
</tr>
<tr>
<td>Communication Disturbance</td>
<td>Hyperphagic Drive</td>
<td>.236</td>
<td>.067</td>
<td>.367</td>
<td>3.53</td>
<td>.001</td>
</tr>
<tr>
<td>Anxiety</td>
<td>Hyperphagic Drive</td>
<td>.155</td>
<td>.041</td>
<td>.392</td>
<td>3.81</td>
<td>.001</td>
</tr>
<tr>
<td>Social Relating</td>
<td>Hyperphagic Drive</td>
<td>.168</td>
<td>.075</td>
<td>.370</td>
<td>2.23</td>
<td>.030</td>
</tr>
<tr>
<td></td>
<td>Hyperphagic Behaviour</td>
<td>-.014</td>
<td>.126</td>
<td>-.018</td>
<td>-.109</td>
<td>.914</td>
</tr>
<tr>
<td></td>
<td>Weight Category</td>
<td>1.371</td>
<td>.863</td>
<td>.201</td>
<td>1.59</td>
<td>.118</td>
</tr>
</tbody>
</table>

**Disruptive/Antisocial Behaviour**

Multiple linear regression using the forced entry method was used to explore how well hyperphagic drive and hyperphagic behaviour could account for levels of Antisocial/disruptive Behaviour, whilst controlling for age and weight category. The four predictor model accounted for 44% of variance in disruptive/antisocial behaviour ($R^2=.44$, Adjusted $R^2=.40$) and a significant model emerged ($F(4,58)=10.63$, $p<.001$). Regression coefficients are shown in Table 4. Hyperphagic Drive significantly predicted levels of Disruptive/Antisocial behaviour and demonstrated the highest beta value (.539) indicating that it was performing well as a predictor. Age also significantly predicted Disruptive/Antisocial behaviour, however it had a lower beta value (.238), indicating that it was not performing so well as a predictor.
Finally, Hyperphagic Behaviour and weight category did not emerge as significant predictors of disruptive/antisocial behaviour.

Self-Absorbed Behaviour
Multiple linear regression using the forced entry method was used analyse how well levels of hyperphagic drive and hyperphagic behaviour could account for levels of Self-Absorbed Behaviour. The two predictor model was accounted for 30% of variance in self-absorbed behaviour ($R^2=.30$, Adjusted $R^2=.29$) and a significant model emerged ($F(2,75) = 15.912, p<.001$). Regression coefficients are shown in Table 4. Hyperphagic Drive significantly predicted Self-Absorbed Behaviour, with greater levels of hyperphagic drive predicting more self-absorbed behaviour. It also had the highest beta value (.488) indicating that it was performing well as a predictor. However, Hyperphagic Behaviour did not emerge as a significant predictor and had a much lower beta value (.086), indicating that it was not performing well as a predictor of self-absorbed behaviour.

Communication Disturbance Subscale
Linear regression was performed to explore the association between Hyperphagic Drive and Communication Disturbance Scores. Hyperphagic Drive accounted for a significant proportion of variance in Communication Disturbance scores ($R^2=.14$, Adjusted $R^2=.12$) and a significant model emerged ($F(1,81) = 12.45, p=.001$).
**Anxiety Subscale**

Linear regression was performed to explore the association between Hyperphagic Drive and Anxiety Scores. Hyperphagic Drive accounted for a significant proportion of variance in anxiety scores ($R^2=0.15$, Adjusted $R^2=0.14$) and a significant model emerged ($F(1,81) = 14.513, p<0.001$).

**Social Relating Behaviour**

Multiple linear regression using the forced entry method was used to analyse how well hyperphagic drive and hyperphagic behaviour could account for levels of Social Relating, whilst controlling for weight. The three predictor model accounted for 21% of variance in social relating ($R^2=0.21$, Adjusted $R^2=0.17$) and a significant model emerged ($F(3,58) = 4.817, p=0.005$). Regression coefficients are shown in Table 4. Hyperphagic Drive was a significant predictor of social relating behaviour, with higher levels of hyperphagic drive predicting higher levels of social relating difficulties. Hyperphagic Drive also had the highest beta value (.370) suggesting it was performing well as a predictor of this subscale. Finally, Hyperphagic Behaviour and weight category did not emerge as significant predictors of social relating behaviour problems.

*(See Sections 3.5 to 3.6 of the Extended Paper for an extended description of the assessment regression assumptions and additional analyses)*
Discussion

This study aimed to explore the relationship between hyperphagia and emotional and behavioural problems in children with PWS. High rates of emotional and behavioural problems were identified, with 57% scoring above the clinical cut-off on the DBC-P (Einfeld & Tonge, 2002). Furthermore, the significant positive correlation between total hyperphagia score and total problem behaviour score suggested that the higher the level of hyperphagia, the more emotional and behavioural problems experienced. This is consistent with Dykens et al. (2007) who also found a significant correlation between hyperphagia and behaviour problems. However, the aim was to examine the relationship in more detail by looking at the impact of hyperphagia on the emotional and behavioural subscales of the DBC-P. It was hypothesised that: higher levels of hyperphagic drive and severity (food-related distraction and distress) would be associated with increased antisocial/disruptive behaviour, anxiety, social relating problems and self-absorbed behaviour. Whilst, hyperphagic behaviour (stealing/foraging for food) would only be associated with antisocial/disruptive behaviour.

The results of multiple regression analyses indicated that hyperphagic drive and age significantly predicted disruptive/antisocial behaviour, whilst hyperphagic behaviour and weight category did not. This reflects research, which suggests that individuals experiencing persistent hunger experience anger and frustration, leading to increased conflict (Keys et al. 1950). It also fits with reports from individuals with PWS, who attributed frustration to constantly feeling hungry (Haselip, 2006). Age is also commonly associated with increased
antisocial/disruptive behaviour in children with ID, with older children and adolescents displaying higher levels of disruptive/Antisocial behaviour (Dykens, 2004; Steinhausen et al. 2004). However, hyperphagic behaviour was not a significant predictor. This result contradicts Dykens et al. (2007) who found that hyperphagic behaviour was significantly related to disruptive behaviour.

Multiple regression revealed that hyperphagic drive significantly predicted levels of self-absorbed behaviour, whereas hyperphagic behaviour did not. This supports previous research, which has found that individuals experiencing hunger may become distracted from alternative activities as they become fixated on food and eating. For example, Keys et al. (1950) described individuals withdrawing from non-food related interests and activities (1950).

Results indicated that hyperphagic drive significantly predicted levels of anxiety, with higher drive associated with higher anxiety. This reflects research, which has found that people experiencing hunger experience increased anxiety. For example, Weinreb et al. (2002) found that persistent hunger was associated with high anxiety in school-aged children. Furthermore, individuals with PWS reported that the constant sense of hunger made them feel distracted and worried (Haselip, 2006).

In this sample, hyperphagic drive predicted levels of Communication Disturbance. This result was not anticipated, as previous research has not indicated an association between hunger and communication impairments (such as echolalia, whispering, and talking to self). However, this subscale also
includes items that relate to repetitive speech, preoccupations and obsessions. The literature indicates that individuals with PWS are more likely to display both food and non-food related obsessions and compulsions including: repetitive actions; insistence on routines; and repetitive speech (Clarke et al. 2002). In this study it is difficult to ascertain whether increased hyperphagia leads to increased food-related obsessions and compulsions (as measured by the Hyperphagic drive subscale), or also to increased obsessions in non-food areas. The measures employed in this study were not able to control for this adequately. Therefore, more research is needed to explore this further.

Increased hyperphagic drive significantly predicted Social relating problems, whereas hyperphagic behaviour and weight category did not. This fits with other studies which have found that hunger predicts withdrawal from others, low mood and depression in non-PWS groups (Hill et al. 1991; Keys et al, 1950; Weinreb et al, 2002). Furthermore, individuals with PWS also reported feeling ‘sad’ and ‘depressed’ due to continual hunger (Haselip, 2006).

When the results are considered overall, hyperphagic drive emerged as a significant predictor of all subscales whilst hyperphagic behaviour did not significantly predict any, even though it was significantly correlated with disruptive behaviour, social relating and self-absorbed behaviour. This finding is consistent with Dykens et al (2007) who found that hyperphagic behaviour was only significantly associated with non-compliant behaviour. One explanation could be that multi-collinearity existed between the HQ subscales and this may negatively impacted on the regression. However, the levels of multi-collinearity
were well within the acceptable range (Field, 2005), therefore this did not seem to be an issue. One hypothetical explanation is that hyperphagic behaviour is a consequence of hyperphagic drive, like any of the other behavioural and emotional problems measured on the DBC-P. Therefore, Hyperphagic drive leads to emotional changes (e.g. anger or frustration), which then leads to antisocial/disruptive or hyperphagic behaviours (e.g. stealing food) depending on the situation and availability of food. If this were the case, then hyperphagic behaviour would not be expected to predict emotional and behavioural problems. However, this theory requires exploration through further empirical analysis.

There were some limitations to this study. Firstly, the hyperphagia measure (HQ) is still in early stages of development and Dykens et al’s (2007) three factor structure was not supported in the current study. This may be attributed to the small sample, as Field (2005) suggests that 10-15 cases are required per item for factor analysis, and for this study there were only 81 cases for 11 items. However, the two factor structure appeared robust and displayed good levels of internal consistency. Furthermore, the original measure contained one subscale with only two items in it (Hyperphagic Severity). Costello and Osborne (2005) stated that a subscale with less than three items should be regarded as ‘weak and unstable’; therefore the two component structure is likely to be stronger. Further research would be required to examine factor structure and to explore and confirm the reliability and validity of the scale.
Additionally, whilst the HQ takes researchers a step closer to being able to explore hyperphagia in PWS, one salient limitation is that it is only able to measure the external expression of hyperphagia, not internal experiences or severity of hunger. Therefore, the results may have been confounded by the fact that both the HQ and DBC-P measured emotional and behavioural problems. To address this in future research and gain a more complete understanding of the impact of hyperphagia on emotions and behaviour, attention needs to be paid to developing other means of assessing hyperphagia. This may be achieved through qualitative research with people with PWS exploring the construct of hyperphagia, which could then lead to the development of standardised self-report questionnaires. Radimer et al. (1990) utilised a similar methodology to develop a measure of hunger in adults without PWS. Self-report measures and qualitative research have previously been ruled out due to the varying levels of ID in PWS and possible response biases (Jones, et al. 1997; Young, et al. 2000). However, it is important to involve people with ID and children in research, as it is only by supporting people to comment on their own lives that we can gain a full picture of experiences (Chappell, 2000; Lewis & Porter, 2004; Walmsley, 2001). Furthermore, it could also be useful to look at the impact of appetite stimulating hormones such as Grehlin (Haaq et al. 2003) on eating behaviour and non-food related behaviour in PWS to examine whether changes in such hormones predict levels of non-food related behavioural and emotional problems.

Another limitation is that, whilst the recruitment methods were intended to be representative, postal questionnaires are often subject to response and non-
response bias (Vink et al. 2004). This study may have been affected, as it obtained a lower than expected response rate of 30%, furthermore 86% of respondents were mothers. Previous studies have shown that parent's responses to questions about their child's emotions and behaviour are associated with factors such as their gender and their own mental-health (Jensen et al. 1988; Kroes et al. 2003; Vink et al. 2004). Najman et al. (2000) found that depressed or anxious parents were more likely to over-report behavioural problems. This was supported by Kroes et al. (2003) who found that parents with high stress or mental-health problems are more likely to project their own symptoms onto their children when reporting on behaviour. In addition to this, Jensen et al. (1988) found that mothers rated significantly more behavioural problems than fathers. None of these factors were controlled for in this study. However, as the levels of behavioural and emotional problems reported in this study were in line with previous studies of behaviour in PWS (Einfeld et al. 1999), it seems unlikely that excessive over-or-under-reporting was a major problem in this study. However for future research, information could be collected on parent stress/ mental-health to control for such bias.

(See section 4.2 of the Extended Paper for further discussion of the limitations of this study)

It is important to consider that the factors explored in this study are unlikely to be the only factors associated with behavioural and emotional problems in PWS. Therefore, the consideration of other possible risk factors in further analyses may improve the predictive power of these models and the understanding of behaviour in PWS. For example, previous research suggests that level of intellectual ability is associated with behavioural and emotional
problems. For example Einfeld et al. (1999) found that those with more severe ID displayed higher levels of disruptive and antisocial behaviour and self-absorbed behaviour. In the current study, only seven parents provided information on their child’s intellectual ability. Therefore, level of intellectual ID may also account for some variance in behaviour in this sample. As a result, further research would be required which incorporated an assessment of intellectual abilities.

Further analyses may also include systemic factors such as socio-economic status, family functioning, parent stress and mental illness, as studies with children with ID of other aetiologies have found a link between these factors and behaviour (Feldman, et al. 2007; Wallander, et al. 2006). These factors have not yet not been investigated in PWS, however parents of children with PWS have been found to display higher levels of stress, compared with parents of children with ID of other aetiologies (Hodapp et al. 1997).

Despite the limitations, the results of the present study have important implications for the understanding and treatment of behavioural and emotional problems in PWS. The possibility that the two symptoms frequently observed in people with the syndrome may be related has not been well researched to date, as they have been regarded as distinct phenotypes. This has led to distinct treatments, which focus on the problems separately. For example, the recommended treatment for hyperphagia is strict environmental control (Whitman & Jackson, 2006). Whilst for behavioural and emotional problems,
interventions vary from pharmacological treatments to behavioural interventions that target specific problem behaviours like self-harm (Luiselli, 1988).

Finding a link between hyperphagia and non-food related behaviour provides support for the development of interventions that take into account the bio-psycho-social components of the syndrome, and therefore, the possible impact of hyperphagia on the emotions and the behaviour of people with PWS. For example, Singh et al. (2008) devised a mindfulness-based intervention for an adolescent with PWS in which they taught mindful meditation to manage hunger. This significantly aided weight loss. They found that this intervention also had a positive impact on the participants’ challenging behaviour, as they also developed skills in managing their emotions. Similar interventions could provide individuals and their families with skills in managing the physical, emotional and behavioural impact of PWS.

(See Section 4.3 of the Extended Paper For further discussion of the clinical implications)

Conclusion

In summary, this study has added to the understanding of a syndrome, which has been neglected in psychological research. The results suggest that children who experience high levels of hyperphagic drive also experience high levels of emotional and behavioural problems, including: disruptive/antisocial behaviour, anxiety, social-relating problems, communication disturbances and self-absorbed behaviour. This supports research, which indicates that long-term hunger may cause a variety of psychological, behavioural and cognitive changes in individuals, including: anger, anxiety, withdrawal, obsessions and
depression (Weinreb et al, 2002). However, the method of measuring hyperphagia has been called into question. This indicates that further research is required to address limitations. It is hoped that this will lead better understanding of hyperphagia in PWS and also to the development of more effective and comprehensive psychological interventions for behavioural problems.

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"Refer to": - 310
Total Research Paper Word Count: 6999
References


children with or at risk of developmental delay. *Journal of Intellectual Disability Research* 51 (8), 606-619.


Section 1: Extended Background

1.1 Extended description of the causes, diagnosis and symptoms of PWS

Prader-Willi syndrome was first discovered in 1956 by Swiss endocrinologists: Prader, Labhart and Willi. Who described the cases of nine individuals who all displayed evidence of: intellectual disabilities, excess body fat, short stature, hypogonadism and infantile hypotonia (Butler, Hanchett & Thompson, 2006). However, it was not until the early 1980s with the advent of genetic testing that it was discovered that the syndrome is caused by abnormalities on chromosome 15q11-q13 (Cassidy, Thuline, & Holm, 1984; Ledbetter et al., 1981). Since then, it has been discovered that in around 60-70 percent of cases these abnormalities are due to a new deletion on chromosome 15q11-q13 inherited from the father; a further 25-30 percent are caused by inheriting two chromosome 15q11-q13’s from the mother and none from the father (maternal uniparental disomy- ‘UPD’); and in the remaining 2-5 percent of cases it is caused by either a mutation or imprinting defect in the same region (Gunay-Aygun, Schwartz, Heeger, O’Riordan & Cassidy, 2001; Kundert, 2008). The syndrome is not generally thought to be directly passed down through families, although Cassidy, Dykens and Williams (2000) estimated the chance of recurrence within families as being around one percent. Furthermore, cases of UPD have been associated with increased maternal age (Kundert, 2008).

A definitive diagnosis of PWS is currently only obtainable though genetic testing, however clinical diagnostic criteria for the disorder have also been developed (see Table 3). These criteria provide an overview of the range and complexity of the symptoms, which an individual with PWS may experience. The diagnostic criteria are often employed by clinicians to confirm the need for further genetic testing (Gunay-Aygun et al. 2001). Early methods of genetic testing were often vulnerable to false diagnoses, as other genetic syndromes such as Angleman Syndrome can present in a similar way to PWS in the early stages (with severe infant hypotonia and poor suck reflex) and are also caused by a deletion on chromosome 15 (Cassidy & Driscoll, 2009). In fact, specific guidelines for genetic testing of PWS were not published until 1996, when the
American College of Medical Genetics (ACMG) and the American Society of Medical Genetics (ASHG) identified the most appropriate methods for diagnosis. Therefore, as definitive genetic testing has only become possible relatively recently, the average age at diagnosis is not currently known.

Gunay-Aygun et al. (2001) studied 90 participants who had recently undergone genetic testing for PWS in the USA and had received a positive result. The age range in this sample was five months to 60 years (with a median of 14 years). Eiholzer, L'Allemand and Zipf (2003) stated with the ease of testing, it is now becoming increasingly popular for clinicians to genetically test all newborns presenting with poor muscle tone (hypotonia) at birth. Eiholzer et al. (2003) reported that in Switzerland up to 50% of newborn babies with hypotonia who were given genetic testing received a diagnosis of PWS. In a recent study in the UK, Russell and Oliver (2003) found that in their sample of 58 individual with PWS, the age at diagnosis ranged from birth to 17 years (mean = 4.1 years; standard deviation = 4.7 years). This indicates that people may be beginning to be diagnosed with the syndrome much earlier.

As mentioned in the paper, the cause of many of the clinical symptoms (see Table 3) of the syndrome is thought to be widespread hypothalamic dysfunction. This is caused by a marked decrease in the size of the paraventricular nucleus, an area responsible for sexual development, eating behaviour, growth and body temperature (Crino et al. 2003; Swaab, 1997). This is coupled with the production of fewer oxytocin-expressing neurons (Kundert, 2008). In addition to this, Swaab (1997) investigated the hypothalamus in PWS and found a significant reduction in growth hormone releasing neurons in the arcuate nucleus, an area responsible for the regulation of eating behaviour. This is combined with the fact that the hormone Ghrelin (an appetite stimulant) was found by Haqq et al. (2003) to be significantly elevated in people with PWS.

Other structural abnormalities in the brain have also been noted. For example, Miller et al. (2007) conducted three-dimensional magnetic resonance imaging (3D-MRI) scans on 20 people with PWS aged three months to 39 years old.
The results were compared with a normal weight and an obese control group. They found multiple abnormalities such as: decreased brain volume in the parietal-occipital lobe (although only in the over five's); enlarged ventricles (venticulomegaly); Sylvian fissure polymicrogyriasome (which is related to language disorders); Lack of complete insula closure (which is linked with lower pain perception and autonomic control); cortical atrophy; and small brain stem. However Miller et al. (2007) themselves recognise that this was a cross-sectional study with a relatively small sample size. Further longitudinal studies would be required to study whether these abnormalities are present from birth or whether they occur later as a result of other difficulties and therefore may be treatable.

As demonstrated in the clinical diagnostic criteria (Table 3), the prominent food related problems associated with PWS usually actually begin at birth with a poor suck reflex and severe hypotonia, which can often lead to a failure to thrive (Dykens & Kasari, 1997). As such, infants with PWS can often require assisted feeding during the first year of life. However, once the hyperphagia has begun. Dimitripoulos et al. (2000) stated that individuals with PWS generally “report being hungry, seem never satiated, and rarely vomit.” (p.126). Therefore individuals with PWS are required to permanently remain on a low calorie diet to prevent obesity (Pipes & Holm, 1973). In fact, the combination of low muscle tone, low energy expenditure and slow metabolism affecting people with the syndrome means that to maintain a normal weight, an average adult with PWS only needs 1,000-1,200 calories per day (Kundert, 2008) compared to the 2,000-2,500 calories per day for the average adult without PWS (NHS Choices, 2009). A further consequence of this is that individuals with PWS also often develop food related behavioural problems. For example, common food related difficulties reported are: excessive consumption of food, intense preoccupation or obsession with food or talking about food, incessant food seeking (including foraging for food and stealing food or money to buy food), and pica (consumption of non-food items) (Dimitropoulos et al. 2000; Dykens & Kasari, 1997).
Table 3

**Clinical Diagnostic Criteria for Prader-Willi Syndrome (Adapted from Holm et al. 1993)**

| Major criteria (1 point each) | 1. Infantine central hypotonia  
2. Infantine feeding problems/failure to thrive  
3. Rapid weight gain between 1 and 6 years  
4. Characteristic facial features  
5. Hypogonadism, with any of the following:  
   a. Genital hypoplasia,  
   b. Pubertal deficiency  
6. Developmental delay/mental retardation  
7. Hyperphagia/food foraging/obsession with food  
8. Deletion on 15q11-13 or other cytogenic molecular abnormality of that region |
|---------------------------|----------------------------------------------------------------------------------|
| Minor criteria (½ point each) | 1. Decreased foetal movement and infantile lethargy  
2. Typical behavioural problems  
3. Sleep disturbance/sleep apnoea  
4. Short stature for the family by age 15 years  
5. Hypopigmentation (lighter hair, eye and skin colours)  
6. Small hands and feet for height age  
7. Narrow hands with straight ulnar border  
8. Esotropia, myopia  
9. Thick, viscous saliva  
10. Speech articulation defects  
11. Skin picking |
| Supportive criteria (no points) | 1. High pain threshold  
2. Decreased vomiting  
3. Temperature control problems  
4. Scoliosis/kyphosis  
5. Early adrenarche  
6. Osteoporosis  
7. Unusual skill with jigsaw puzzles  
8. Normal neuromuscular studies |

**For diagnosis:**  
- 5 points required for children under 3 years of age  
  (with three from major criteria)  
- 8 points required in those above 3 years of age  
  (with four from major criteria)  

In addition to the physical characteristics described above and the eating difficulties, people with PWS often also experience a number of cognitive symptoms from birth onwards. For example, individuals usually display a definitive intellectual impairment coupled with borderline or mild to moderate intellectual disabilities (Steinhausen, Eiholzer, Hauffa, & Malin, 2004). The average IQ of a person with PWS is around 60-70 points; although average IQs
have been found to occur in approximately five percent of cases (Whittington et al. 2004). Whittington et al. (2004) found that individuals with PWS have particular cognitive strengths in visual processing tasks, but they commonly perform more poorly in tasks requiring auditory processing, arithmetic and/or short-term memory. People with the syndrome have also been found to have particular skills in completing jigsaw puzzles and word searches. For example, Dykens (2002) found that a group of children with PWS outperformed a matched group of children with intellectual disabilities of other aetiologies on a word search task. They also out-performed a group of children with no intellectual disabilities on a jigsaw puzzle task. However, it has been proposed that individuals with the syndrome may often fail to perform at their optimum IQ due to diminished social skills and emotional capabilities (Rosner, Hodapp, Fidler, Sagun, & Dykens, 2004). This can often lead to academic underachievement in young people with PWS (Whittington et al. 2004).

The apparent diminished social skills and understanding of emotions in PWS has been linked to an increased susceptibility towards Autistic Spectrum Disorders (ASD) in people with the syndrome. Veltman, Craig and Bolton (2005) proposed that there are high incidences of ASD-type symptoms and behaviour in individuals with PWS, for example; poor skills in social interactions and a tendency towards stereotyped and ritualistic behaviour. Greaves, Prince, Evans and Charman (2006) compared the repetitive and ritualistic behaviour of children with PWS and those with autism. They found that both groups of children displayed similar levels of repetitive and ritualistic behaviour overall, but that parents of the children with PWS more frequently endorsed items relating to the collecting and storing of objects, whilst the parents of the children with Autism children more frequently endorsed lining up objects and awareness of detail. Veltman et al. (2005) related this to the fact that maternally derived duplications of chromosome 15 have been linked to ASD, therefore individuals with PWS with a diagnosis of Uni-parental Disomy may be more vulnerable ASD than those with a paternal deletion. This was supported by Dimitropoulos and Schultz (2007) who concluded that poor social skills and repetitive behaviours seen in PWS are likely to be related to genetic factors. Although they do note that so far the genes have only been linked to autism and there is
not yet a definitive genetic marker of Autism or ASD. Furthermore, others have argued that the behaviours seen in PWS are better labelled as a form of obsessive- compulsive disorder (Dykins, Leckman & Cassidy, 1996). See the section below for an extended discussion of this.
1.2: Further Exploration of Behavioural and Emotional problems in PWS

As mentioned in the paper, behavioural and emotional difficulties frequently occur in addition to the food-related problem behaviours in individuals with PWS. Such difficulties have been found to commonly develop in children with the syndrome at approximately two years of age (Dimitropolous, Feurer, Butler, & Thompson, 2001). This is around the same time as the onset of the hyperphagia and like the hyperphagia, the behavioural problems are also believed to persist throughout life (Clarke, Boer, Chung, Sturmey & Webb, 1996). However, behavioural problems are thought to be most severe in adolescence and early adulthood (Steinhausen, et al. 2004; Whitman & Jackson, 2006).

No unitary definition or label for the emotional and behavioural problems has emerged and as such, in past research they have been labelled: maladaptive behaviours (Clarke et al. 1996; Dykens & Kasari, 1997), problem behaviours (Dykens et al. 2007; Steinhaussen et al. 2004), behavioural disorders (Hirawi, Maegaki, Oka, & Ohno, 2007), Behavioural disturbances (Einfeld, Smith, Durvasula, Florio, & Tonge, 1999). Many attempts to define behavioural problems, involve the use of the umbrella term ‘challenging behaviour’. This label is commonly utilised to describe problem behaviours within wider research, education and health and social care with children with intellectual disabilities. Challenging behaviour in people with intellectual disabilities has been defined as:

“behaviour of such an intensity, frequency or duration that the physical safety of the person or others is likely to be placed in serious jeopardy, or behaviour which is likely to seriously limit or deny access to and use of ordinary community facilities”

(Emerson 2001, p.1).

However, Emerson’s description may automatically include all people with PWS, as their hyperphagic behaviour may place them in jeopardy and/or limit their access to community facilities. Therefore, this definition does not adequately encompass some of the emotional difficulties thought to be
experienced by people with the syndrome, such as anxiety or depression. Einfeld and Tonge (2002) proposed a broader definition of ‘emotional and behavioural problems’ in children with intellectual disabilities. They defined this as being:

“Where behaviours and emotions are abnormal by virtue of their qualitative or quantitative deviancy and cannot be explained on the basis of the intellectual disabilities alone, and cause significant distress to the young person, carers or the community, as well as significant added impairment, then they are defined as disordered.” (Einfeld & Tonge, 2002, p.xiv)

This definition appears to more adequately encompass individuals with PWS, as it includes the statement that the behaviour cannot be explained by the intellectual disabilities alone.

The different definitions adopted in research have led to a variety of different measures being used to assess behaviour, which in turn makes the estimation of the prevalence of behavioural and emotional problems in PWS quite difficult. A review of studies was conducted as part of this investigation. The review examined the prevalence and nature of behavioural and emotional problems in PWS and the results are summarised in Table 4. This review demonstrates the wide variation in the types of behavioural and emotional problems experienced by people with PWS, as well as the prevalence of such problems and the measures used to assess them.

However, Table 4 does appear to demonstrate overall empirical support for the increased prevalence of behavioural and emotional problems in PWS when compared with other groups. This extends beyond the Dykens and Kasari, (1997) study, (cited in the paper). For example, Einfeld, et al. (1999) compared the behaviour of 46 children with PWS (as measured through parental report questionnaires) with that of children with intellectual disabilities of other aetiologies from a community sample (n=454). They found the parents of the PWS group reported significantly higher levels of psychopathology than the
control group. In particular, the PWS group had high levels of antisocial behaviour (for example, 83% of the PWS had severe temper tantrums compared to 26% of the control group). However, as the groups were not matched for age, gender or level of intellectual disabilities, it is difficult to ascertain whether or not demographic factors may have accounted for some of these differences (for example, the mean age of the control group was 12 years, whilst the mean age of the PWS group was 17.7 years). Therefore these results should be viewed with caution.

In addition to the behaviour problems, it has also been identified that people with PWS often engage in other more specific forms of challenging or problematic behaviour like self-harm. In particular, picking at or damaging skin tissue is common, as is using objects (such as badges and scissors) to scratch the skin, pulling out hair, fingernails or toenails and swallowing inedible objects or poking them in the ears or the nose (Dorman, 2001). In fact, in a study with the families of 62 people with PWS, self-injury was reported in 81% of the sample (Symons, Butler, Sanders, Feurer & Thompson, 1999). However this is thought to be due to the fact that people with PWS have a high pain threshold and therefore do not experience warning signs related to that behaviour. It has also been proposed that the feelings of pleasure associated with the release of endorphins make this activity attractive, especially to provide individuals with a sense of relief from emotions such as anxiety or frustration (Dorman, 2001).

Researchers have also found that individuals with PWS are at an increased risk of developing obsessive-compulsive disorder (OCD), extending beyond the food-related obsessions to other areas. In fact, obsessions and repetitive or compulsive behaviours are said to be another key behavioural feature of the syndrome (Beardsmore et al.1998; Clarke et al. 2002; Curfs, Hoondert, van Lieshout, & Fryns, 1995; Dykens et al.1996; Reddy, Steven & Pfeiffer, 2007). In a survey of compulsive and ritualistic behaviour in PWS, Clarke et al. (2002) found that the syndrome is commonly associated with the following ritualistic behaviours:

- the need to ask or tell something repeatedly
- insistence on routines and rituals
- hoarding and ordering of objects
- and repetitive actions and speech

Interestingly, though Clarke et al. (2002) found little evidence of more ‘typical’ compulsive behaviours, which may be related to a clinical diagnosis OCD such as checking, cleaning and counting. Dimitropolous et al. (2000) stated that as this compulsive behaviour begins in early childhood and often persists throughout life, that biological mechanisms rather than anxiety are likely to play a significant role. This is thought to be related to the vulnerability to autistic spectrum disorders (ASD) mentioned previously. There has long been a connection made in the literature between the ritualistic and stereotyped behaviours of ASD and the compulsive behaviours in OCD. The prevalence of OCD in people on the autistic spectrum is said to range from 1.5% to 81%, depending on the measure and diagnostic criteria used (Leyfer et al. 2006). However this has not yet been investigated empirically, therefore it remains uncertain whether biological mechanisms or the psychological and social experiences of PWS trigger the ritualistic and inflexible behaviours noted, or whether it may be a mixture of the two.

Other mental health problems have also been identified in some individuals with PWS. In fact, it has been proposed that around 10% of adults and adolescents with PWS go on to develop “major psychiatric problems”, such as severe and agitated depression, bipolar disorder and even psychotic episodes (Descheemaeker et al., 2002, p 42). Beardsmore, Dorman, Cooper and Webb (1998) studied 23 adults with PWS and compared them with a group of adults with intellectual disabilities of other aetiologies (n=73). They found that the PWS group had higher rates of affective disorders, with 17.4% of individuals with PWS suffering from an affective disorder, compared to 4.1% for people with intellectual disabilities of other aetiologies. They also found higher rates of schizophrenic/ delusional disorders in the PWS group. However, this is based on a small sample (n=23) of adults aged between 16 and 51 years old. Therefore, it is not necessarily clear how prevalent such difficulties are in the wider population of people with the syndrome (especially children and older adults). This indicates that individuals with PWS may have a vulnerability to
mental health problems, although the factors contributing to this have not yet been investigated.

However, it seems important to note that the results of the studies showing increased behavioural problems and increased vulnerability to mental health problems in individuals with PWS should be viewed with caution. Firstly, many of these studies are based on adults with PWS rather than young people. Furthermore, more recent studies have suggested that behavioural difficulties and mental health problems may be more common in many other groups of individuals with intellectual disabilities than some have suggested. Research indicates that the number of people with intellectual disabilities regularly displaying severe challenging behaviour in the population may actually be between 10-20% (McClintock, Hall & Oliver, 2003).

Furthermore, two major surveys conducted in 1999 and 2004 by The Office for National Statistics, indicated a high prevalence of mental health problems in children and adolescents with intellectual disabilities in the UK (Emerson & Hatton, 2007). In a recent report by Emerson and Hatton (2007), the data collected from the two reports was combined, giving a total sample of over 18,000 children and adolescents (aged five to fifteen years). It was discovered that over 36% (one in three) children with intellectual disabilities possess a diagnosable mental health problem and far more than that show significant signs of challenging behaviour or distress (Emerson & Hatton, 2007). The authors conclude that this is likely to be related to the “increased exposure to poverty and social exclusion than being something inherent in having learning disabilities” (page iii). It does not appear that these factors have been considered in the research looking at PWS and challenging behaviour and/or mental health problems.
Table 4

Showing sample size, measures used and results found in a sample of studies looking at behaviour in PWS

<table>
<thead>
<tr>
<th>Authors</th>
<th>n (PWS)</th>
<th>Cohort</th>
<th>Assessments</th>
<th>Behaviours Assessed</th>
<th>Findings</th>
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</table>
| Beardsmore, Dorman, Cooper & Webb (1998)     | 96 (23) | Adults with PWS living in residential care (Mean age: 29.3 years, SD: 8.2 years); compared with adults with intellectual disability of other aetiologies (n: 73, Mean age: 39.2 years, SD: 12.2 years) | Present Psychiatric Scale-Learning Disabilities (PPS-LD) (Cooper, 1997) Adaptive Behaviour Scale (Nihira et al. 1993) Mini PAS-ADD (Prosser et al. 1997) | - All areas of psychopathology - Maladaptive Behaviour - Activities of daily living | - Affective disorders in 17.4% of PWS sample compared with 4.1% in the control.  
- Behaviour disorders in 65.2% of the sample compared with 15.1% of the control. |
| Clarke, Boer, Whittington, Holland, Butler & Webb (2002) | 140 (97) | Parents and carers of people with PWS (Mean age 20.8 years, SD 12.5 years) were compared to 43 people with learning disabilities of other aetiologies (mean age: 20.2, SD 14.6 years) | PWS Structured Interview Questionnaire (PWS-SIQ: Clarke et al. 2002) | - Compulsive and ritualistic behaviour - Mood - Behavioural problems - Eating behaviour | - Compulsive symptoms were not associated with age or obesity in the PWS sample.  
- Ritualistic and compulsive behaviours occurred significantly more in the PWS group than the control group, except for repetitive behaviour. |
| Clarke, Boer, Chung, Sturme & Webb (1996)    | 60 (30) | Adults with PWS (N: 30, aged 16-44 years) compared with 30 adults with non-specific intellectual disability (matched for age, gender and intellectual ability) | Aberrant Behaviour Checklist (ABC– Aman et al. 1985a, b) completed by staff and researchers | - Irritability - Lethargy/Withdrawal - Stereotypy - Hyperactivity/Non-compliance - Inappropriate Speech | - The PWS group scored significantly higher on Irritability and inappropriate speech, but not on lethargy/withdrawal; stereotypy or hyperactivity/non-compliance.  
- No relationship was found between ABC scores and age, gender or BMI. |
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<th>Authors</th>
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<tr>
<td>Deschemaeker et al. (2002)</td>
<td>(53)</td>
<td>Parents of 31 Children &amp; adolescents (aged; 1–18 years) and 22 adults (aged; 18 - 47 years) with PWS in the community.</td>
<td>Unstructured clinical interviews by psychiatrists and clinical psychologists, conducted yearly over a 15 year period.</td>
<td>- Interviews focussed on the medical, emotional, behavioural and cognitive symptoms of the syndrome.</td>
<td>- Four participants were diagnosed with acute cycloid psychosis. Four were diagnosed with unspecified bipolar disorder.</td>
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<td>Dimitropoulos, Feurer, Butler &amp; Thompson (2001)</td>
<td>237 (105)</td>
<td>Parents of children with PWS (2-6 years). Compared with the parents of two groups: Down Syndrome (N: 56, 2-5 years); Typically Developing (N: 76, 2-5 years).</td>
<td>Compulsive Behaviour Checklist for Clients with Mental Retardation (Gedye, 1992) Tantrum Behaviour Survey (Dimitropoulos et al. 2001) Early Child Development Inventory/ Pre-school Development Inventory (Ireton, 1992)</td>
<td>- Compulsive behaviour - Tantrums</td>
<td>- Significantly more compulsive behaviours were reported overall in the PWS group than DS and typically developing groups. - 89.5% of the PWS group had rapid tantrum onset compared to 68% in the DS group and 63% in the typically developing.</td>
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<td>Dykens &amp; Kasari, (1997)</td>
<td>129 (43)</td>
<td>Parents of children with PWS (n. 43). Compared with the control groups: Down Syndrome (DS; n.43); Non-specific learning disabilities (N: 43) (Aged between 4 to 19 years)</td>
<td>Child Behavior Checklist (CBCL- Achenbach, 1991)</td>
<td>- Internalising problems (e.g. withdrawn, somatic complaints, anxious/depressed) - Externalising problems (e.g. delinquent and aggressive behaviour)</td>
<td>- The PWS group demonstrated significantly more frequent and severe internalising, externalising and total problem behaviours. - Seven behaviours predicted PWS group membership with 91% accuracy (skin-picking, overtired, obsessions, impulsivity, speech problems, talks too much and hyperactive)</td>
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<td>Authors</td>
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<td>Dykens, Leckman &amp; Cassidy (1996)</td>
<td>134 (91)</td>
<td>Parents and carers of people with PWS (aged 5-47 years) were selected.</td>
<td><em>Leyton Inventory</em> (measuring OCD) (Murray, Cooper &amp; Smith, 1979)</td>
<td>Obsessions and compulsions</td>
<td>- 64% of PWS group showed OCD symptom related distress, 80% showed symptom related adaptive impairment. - The PWS and OCD groups were not significantly different in terms of severity of and numbers of compulsions.</td>
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<td>A group of 43 individuals with diagnosis of OCD (aged 18-47 years) were also selected and matched with 43 people from the PWS group for age and sex.</td>
<td><em>Questionnaire on Resources &amp; Stress – Freidrich edition</em> (QRS-F; Freidrich, Greenberg &amp; Crnic, 1983)</td>
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<td><em>Yale-Brown Obsessive Compulsive Scale</em> (Y-BOCS; Goodman et al. 1989a,b)</td>
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<td>Einfeld, Smith, Durvasula, Florio &amp; Tongue (1999)</td>
<td>500 (46)</td>
<td>Parents of people with PWS (n= 46, mean age: 17.7 years). Compared to a control group of 454 people with intellectual disabilities of other aetiologies (mean age 12 years).</td>
<td><em>Developmental Behaviour Checklist</em> (DBC: Einfled &amp; Tongue, 1992, 1995)</td>
<td>Disruptive behaviour</td>
<td>Higher levels of overall psychopathology than the controls (Mean Total problem behaviour score = 51.7 vs. 42.3 for the control group), especially with regards to antisocial behaviour.</td>
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<td>Self-absorbed behaviour</td>
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<td>Communication disturbance</td>
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<td>Anxiety</td>
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<td>Autistic relating</td>
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<td>Antisocial behaviour</td>
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<td>Greaves, Prince, Evans &amp; Charman (2006)</td>
<td>169 (80)</td>
<td>Parents of children &amp; adolescents with PWS (aged 3-18 years) compared to a control group of children with autism (N:89, aged 3-17 years).</td>
<td><em>Childhood Routines Inventory</em> (CRI: Evans et al. 1997)</td>
<td>Repetitive and rigid behaviours</td>
<td>Levels of repetitive and ritualistic behaviours in autism and PWS were not significantly different.</td>
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<td>Hiraiwi, Maegaki, Oka &amp; Ohno (2007)</td>
<td>207 (165)</td>
<td>Parents of people with PWS aged 2-31 years. Compared with a matched control group of people with mixed intellectual disabilities of other aetiologies (N: 42, aged 18-31 years).</td>
<td>Unstandardized semi-structured questionnaire designed to assess; health, ability, behavioural and psychiatric problems (Hirairwi et al. 2007)</td>
<td>- Behavioural problems (e.g. stubbornness, tantrums, self-injury, aggression, lying, repetitive speech, hyperactivity, wandering, compulsions, hyperphagia, laziness).&lt;br&gt;- Psychiatric symptoms (e.g. depression, mania, inactivity, delusion).</td>
<td>- 37% of PWS group showed evidence of psychiatric symptoms compared to 11.9% of control group.&lt;br&gt;- Young adults with PWS had significantly higher levels of: Stubbornness, Hyperphagia, Temper Tantrums, Self-injury, Laziness, Hypersomnia and stealing than the control group.</td>
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<td>Reddy &amp; Pfeiffer, (2007)</td>
<td>73 (13)</td>
<td>13 young people with PWS (aged 11-20 years) were compared with:30 with intellectual disabilities of other aetiologies (MR group) and 30 with intellectual disabilities and coexisting psychiatric disorders (DD group). All were in full time residential care.</td>
<td>Devereux Scale of Mental Disorders (DSMD; Nagleiri et al. 1993)</td>
<td>- Conduct&lt;br&gt;- Delinquency&lt;br&gt;- Anxiety&lt;br&gt;- Depression&lt;br&gt;- Autism&lt;br&gt;- Acute problems&lt;br&gt;- Internalising behaviours&lt;br&gt;- Externalising behaviours</td>
<td>- Statistically significant higher levels than the MR group on the Total, Externalising, Internalising, Conduct, Delinquency, Anxiety and acute problems scales.&lt;br&gt;- Comparable levels of psychopathology with the DD group, but lower depression scores.</td>
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<td>Steinhaussen, Etholzer, Hauffa &amp; Malin (2004)</td>
<td>58 (58)</td>
<td>Parents of people with PWS aged 2-29 years, living in the community.</td>
<td>Developmental Behaviour Checklist (DBC; Einfield &amp; Tongue, 1992, 1995)</td>
<td>- Disruptive behaviour&lt;br&gt;- Self-absorbed behaviour&lt;br&gt;- Communication disturbance&lt;br&gt;- Anxiety&lt;br&gt;- Autistic relating&lt;br&gt;- Antisocial behaviour</td>
<td>- Behaviour problems were significantly higher in those over 13 years old than under.&lt;br&gt;- Particularly high scores on disruptive and antisocial behaviour were found.</td>
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| Walz & Benson   | 187     | Parents of children aged 5-19 with: PWS (n. 28); compared with Down Syndrome (DS; n.91); and Angleman Syndrome (AS; n.68). | Parent form of the *Nisonger Child Behaviour Rating Form* (CBRF; Aman et al. 1996) | - Compliant/calm behaviour  
- Adaptive social skills  
- Conduct problems  
- Insecurity /Anxiety  
- Hyperactivity  
- Self-Injury/ Stereotypy  
- Self-Isolation/ Ritualistic  
- Overly Sensitive | - Significantly higher levels of under activity, tantrums, argumentativeness, obsessive-compulsive behaviours, anxiety and over-sensitivity in the PWS group when compared with both the DS and AS groups. |

*Note.* The ‘n’ column reflects the total number of participants recruited for the study whilst the figure in the brackets reflects the total number of PWS participants recruited.
1.3: Extended Clarification of the rationale for the study

1.3.1. Implications for the understanding of the disorder

As the previous section suggests, no clear model of PWS currently exists which is able to consider the behaviour, emotions and physiology of the syndrome together. O'Brien (2000) stated that those investigating behaviour disorders in people with learning disabilities have a duty to attempt to understand it at a variety of different levels, from the macro-social, through to individual cognitions to biological and genetic factors. He went on to state that the over-emphasis of any one of these factors will inevitably “miss the point” (p.620), Woodcock, Oliver and Humphreys (2009) have recently developed a model which explains the links between genotypes and behaviour profiles in complex genetic syndromes. In this model, they hypothesised that the cognitive, behavioural, social and physiological mechanisms of complex genetic syndromes are in fact more closely related to one another than previously believed.

Woodcock et al. (2009) used PWS as an example to demonstrate these links and Figure 1 demonstrates their hypothetical model of PWS. In this model, the authors propose that the biological differences in people with PWS may affect their ability to process new information (according to their level of cognitive impairment). When high demands are then placed on individuals (either by social situations, the environment, or limited cognitive processing abilities) they may enter a state of high physiological arousal. This may then trigger ‘typical’ PWS behaviours such as repetitive questioning (to attempt to clarify the situation or what is expected of them) and ultimately may lead to temper outbursts, as the person feels more and more out of control of the situation. The outcome of this is then affected by the environment and the response of people around them. Woodcock et al. (2009) recommend more research examining the complex interplay between these factors in all genetic conditions.

Woodcock et al’s (2009) model is in the very early stages of development and it has not yet been tested empirically. Nonetheless, it appears to be useful for beginning to unpick the complex factors feeding into the presentation of people with genetic
syndromes. However, the model has a number of limitations when applied to PWS. Firstly, the focus of the paper was not necessarily on advancing the understanding of PWS, therefore much of what the authors suggest is purely speculative and not based on clear research evidence, and as such the model needs to be viewed cautiously. For example, the authors state that repetitive questioning is a symptom of arousal, when in fact it may also be a symptom of cognitive impairment experienced by many people with intellectual disabilities rather than a ‘typical’ PWS behaviour. Furthermore, the authors do not present a clear reason why they attribute some difficulties to ‘CNS abnormalities’ (e.g. temper outbursts) whilst other problems they relate simply to ‘brain abnormalities’ (e.g. cognitive impairment and arousal). In fact, as the brain is part of the CNS, all of the difficulties could fall under the category of CNS abnormality. The authors themselves acknowledge that not enough is known about the exact biological factors which underpin PWS related behaviour yet to be able to accurately speculate on these areas at all.

In addition to this, the one final salient limitation of the model at this time appears to be that the authors appear to have entirely neglected to consider the impact of the most common physiological symptom of PWS: hyperphagia. The absence of hyperphagia in Woodcock et al’s (2009) model is extremely concerning, as it is thought to be the most debilitating aspect of this complex syndrome (Calliandro et al., 2007).

Research into the reactions of healthy individuals to prolonged food restriction and long-term hunger demonstrates that experiencing hunger over long periods has the potential to cause a number of psychological, behavioural and cognitive changes. For example, in one of the most notable studies of food deprivation by Keys, Brozek, Henschel, Mickelsen and Taylor (1950) they took a group of 50 healthy weight male participants and restricted their food intake by 25% over six weeks. Within this study, the men were observed becoming increasingly focussed on or ‘obsessed’ with food (including stealing food and changing careers to food-related careers), and also reported feeling increasingly anxious, irritable and depressed, often leading to increased arguments with their partners and family members. Furthermore, these
symptoms persisted for a number of months after the study ended and normal food intake was restored. Similar psychological effects to these have been found in more recent studies of long-term dieters and children who are periodically subjected to hunger through social deprivation (Polivy, 1996; Weinreb et al. 2002). Polivy (1996) proposed that this indicates that prolonged hunger can in itself cause “increased emotional responsiveness and dysphoria, and distractibility” (page 589). This provides some support for the suggestion that individuals with PWS experiencing reduced satiety or excessive hunger throughout their lives may experience negative mood (such as anger, depression and anxiety) as a result.
**Figure 1:** A hypothetical model of how the genetics of PWS may be associated with cognitive and physiological changes which, via environmental influence, may result in behavioural phenotypic behaviours by Woodcock, Oliver and Humphreys (2009)
1.3.2. Implications for Treatment

It is hoped that this study could contribute to the understanding of behavioural and emotional problems in children with PWS. This could lead to the development of PWS specific treatment packages. Such packages could provide individuals with support in coping with the insatiable hunger as well as for management of the behavioural and emotional difficulties.

At present, there are many different treatments for the various individual symptoms of PWS however, as yet no ‘cure’ for the hyperphagia has been found (Calliandro et al. 2007). Attempts at utilising common pharmacological treatments such as appetite suppressants for limiting hunger in PWS have failed (Whitman & Jackson, 2006). This is thought to be due to the fact that the hyperphagia in PWS results from the central nervous system dysfunction interrupting the messages between the stomach and the brain, rather than a problem with the usual hunger mechanisms (Holland, Treasure, Coskeran & Dallow, 1995; Whitman & Jackson, 2006). In the past, attempts have been made to utilise medications which work on the central nervous system, such as ‘Naloxone’ (an opioid antagonist) to control hunger in PWS, however these have also proved ineffective (Zipf & Berntson, 1987). In fact, some medications (such as ‘Fluoxamine’ and ‘Fluoxetine’) have even been found to exacerbate the hyperphagia and food-related behaviour problems in adolescents with the syndrome (Kohn, Weizman & Apter, 2001). Finally, many children and young people with PWS are now currently treated with Growth Hormone Therapy. This has been found to improve their growth and body composition (as it reduces body fat and increases muscle mass); however this treatment also has no significant effect on the hyperphagia (Eiholzer et al. 1998; l’Allemand, Bachman, Greiser & Eiholzer, 2003).

Therefore, it is currently accepted that the hyperphagia in children and adults with PWS can only be managed through external control from caregivers until an effective and safe medication becomes available (l’Allemand et al. 2003; Whitman & Jackson, 2006). Whitman and Jackson (2006, page 324) state that effective management of individuals with PWS involves the consistent delivery of the following four elements:
1. A physical environment structured so that food access is completely eliminated
2. An appropriate dietary and exercise plan
3. A procedure for ensuring that the effective person is always informed regarding the time and menu for the next meal or snack
4. Elimination of all other avenues for obtaining food

Treatments therefore rely on parents and carers to restrict access to food and provide consistent parenting, in order to prevent an individual from over-eating. This involves the locking of cabinets, fridges, bins or kitchen doors at home, and more recently families have been using electronic methods such as tagging devices and alarm systems to police this. However, Whitman and Jackson (2006) themselves acknowledge that this kind of plan commonly initially leads to an increase in behavioural problems, as individuals challenge the restrictive boundaries. Furthermore, such plans are very difficult to put in place from a practical perspective, as the complete restriction of access to food in the ‘real world’ (e.g. schools, shops and social clubs) is almost impossible. Goldberg, Garret, van Riper and Warzak (2002) stated that parents of children with PWS often find such plans very difficult to cope with, as they can often cause increased conflict within families. As a result, PWS has been linked with significantly higher levels of family and parent stress, when compared with children with intellectual disabilities of other aetiologies (Hodapp, Dykens & Masino, 1997). In a comparative study of PWS and Angleman syndrome, parental stress in PWS was associated with: the need for information and professional support about the syndrome; lack of control over their lives caused by the child’s disabilities; and anxiety about negative consequences for their child (van den Borne et al. 1999). However, Hodapp et al. (1997) found that family stress was more related to the amount of behavioural and emotional problems experienced by children with PWS.

Furthermore the fact that people with PWS currently need so much support and monitoring to control their food intake, makes it unlikely that they will ever achieve full independence as adults. This is counter to adults with intellectual disabilities of other aetiologies, who are increasingly being encouraged to live more independently in the community in supported living environments (‘Valuing People’, 2001).
Caliandro et al. (2007) investigated the quality of life of a sample of 40 children and adolescents with PWS. They found that quality of life was intensely impaired both in terms of mental (emotional) and physical aspects, especially in the group aged fourteen and over. Furthermore, in a qualitative study exploring adult’s experiences of living with PWS (Haselip, 2006), a number of participants reported feeling frustrated by needing to live in residential care and being closely monitored continually, and that they felt as if they were in prison.

Other techniques reported for treating food-related problem behaviours (such as stealing food or sneaking prohibited foods) have been to implement a behavioural treatment programmes. For example, in one study by Maglieri, DeLeon, Rodriguez-Catter and Sevin (2000), an adolescent with PWS was left in a therapy room with access to prohibited food items. She was then observed though a one-way mirror and verbally reprimanded each time she took food. This intervention was then transferred to her home environment over 90 ten-minute sessions. This technique resulted in a decrease in stealing of prohibited food items. However, the authors did not report whether or not the results were maintained over time. Also, the results are difficult to generalise, as they are based on intensive work with one individual with PWS only. In addition to the methodological limitations of the study, there are ethical concerns about using this form of treatment for individuals who may not be able to give informed consent, where they are not in any immediate risk from the behaviour (Emerson, 1998). Furthermore, the impact of the treatment on the individual’s emotional wellbeing or other aspects of behaviour were not considered.

Other behavioural techniques commonly employed have been to use food as reinforcement for positive behaviour. Whitman and Jackson (2006) state that preferred foods could be made contingent on the completion of a desired behaviour (such as getting ready for school on time), alternatively restaurant trips or low calorie confectionary could be used as the reward on a regular schedule based on meeting certain behavioural goals. For example, Ellis, Cress and Spellman (1992) examined the effects of food related reinforcers on the exercise behaviour of an overweight child with PWS. The child studied was required to walk laps of a 47 metre course and on the beginning of each lap; she collected a baton, which she then placed in a rack on completion of the lap. When she had collected all of the batons, she could
then exchange them for a food related token reinforcer. However, although the participant continued with this exercise plan, the intervention was not effective in her becoming quicker over time, nor did she increase the number of laps she was completing. In fact, her pace slowed over sessions. Therefore, it is uncertain whether the use of such behavioural reinforcers was able to promote independent exercise in this individual. Furthermore, other similar studies have actually reported increases in problem behaviour arising when individuals with PWS fail to meet the criteria required to gain the reward (Whitman & Jackson, 2006).

It is clear that much of the emphasis in the treatment of PWS is on managing diet and weight rather than directly approaching the many behavioural and emotional problems associated with the syndrome. This is perhaps because it is considered to be the simplest problem to manage (Holland et al. 2003). Like the hyperphagia, there is no consistently used treatment for the mental health and behavioural difficulties. Treatments for non-food related behavioural and emotional difficulties or are often either pharmacological (e.g. Individuals with PWS are commonly prescribed pharmacological treatments such as Fluvoxamine and Fluoxetine for behavioural difficulties and symptoms of low mood) or from a purely behavioural perspective (Luiselli, 1988).

The risk with relying solely on such interventions is that they may not increase an individual’s sense of control over the syndrome (Singh, et al. 2008). Furthermore, such behavioural methods are unlikely to offer any benefit to people with learning disabilities in respect to their emotional distress, a problem which commonly goes unrecognised by behaviour therapists (Senfert-Kroese, 1997; Wilner, 2005). It is possible that other psychological interventions may be more suitable for this purpose. For example, cognitive-behavioural interventions for people with chronic health problems (such as pain) have been found to be efficacious in reducing psychological distress by providing individuals with techniques for managing the physiological and emotional aspects of their condition with a variety of cognitive and behavioural strategies. White (2001) stated that a large number of chronic and persistent medical conditions require self-management from the individual to control the impact of the symptoms. It is believed that using psychological interventions with individuals can help them to feel more in control and minimise the negative
psychological impact of chronic health problems (White, 2001). Furthermore, in a review of the use of cognitive-behavioural techniques with people with intellectual disabilities, it was demonstrated that self-management procedures can be efficacious in reducing distress in this group, provided that the approach is collaborative and that individuals are fully involved in the planning and design of the intervention (Harchik, Sherman & Sheldon, 1992).

Cognitive-behavioural approaches (CBT) have also been utilised for children and adolescents with primary obesity. For example, Braet, Van Winckel and Van Leeuwen (1997) designed a cognitive-behavioural treatment designed to help children and their families to change their lifestyles, enhance self-regulation skills and to enhance their problem solving skills. The results indicated that CBT can be effective in treating childhood obesity. The authors also conducted a long-term follow up study with the same cases after 4.6 years (Braet & Van Winckel, 2000) in which they found that significantly less of the children who received CBT were obese when compared to the control groups who simply received dietary advice or self-help material. However, Braet et al. (1997; 2000) did not study the impact of the CBT intervention on the non-food related behaviour or emotions of the children in their study. Therefore, it is difficult to speculate how the intervention would impact on the emotions of children who are obese.

In a recent study by Singh et al. (2008) the use of a mindfulness-based health wellness programme for an adolescent with PWS was reported. This programme employed an ABCD design in which:

- **Phase A** was the baseline (retrospectively over 10 months)
- **Phase B** was an intervention involving daily walks (over 12 months)
- **Phase C** was an intervention which combined daily exercise with food awareness (over 12 months)
- **Phase D** was an intervention which combined exercise, food awareness and mindfulness training (which involved mindful eating, visualising and labelling hunger and meditation on the soles of the feet)

This intervention led to significant weight loss over the period of three years, which was maintained at follow-up. Also, interestingly, the participant in this study showed evidence of being able to apply the mindfulness techniques to non-food related
behaviour problems. Reportedly this had a positive impact on his behaviour overall. Singh et al. (2000) recognised that as these techniques have only been studied with this one individual so far, therefore they are not generalisable to the PWS population as a whole. However, the results do suggest that interventions which enable individuals to develop skills in self-management may have beneficial effects on individuals with PWS, which warrants further exploration at least.

In the present study, it is hoped that by learning more about the possible relationship between the physical effects of the syndrome (e.g. the constant hunger) and the behavioural and emotional symptoms, a better understanding may be reached about the factors contributing to and maintaining behavioural and emotional problems in PWS. With this information, more can then be done to create specific holistic interventions which can address behaviour and eating together.
1.4: Extended Clarification of the aims/ Additional Aims and Hypotheses

1.4.1. Extended Clarification of the Aims and Hypothesis

The primary aims and hypotheses of this study are detailed in the research paper. The aim was to examine how well hyperphagia could account for levels of behavioural and emotional problems (measured using the various subscales of the Developmental Behaviour Checklist) in children with PWS. The researchers also aimed to control for age, gender, level of learning disability (IQ) and weight in their analysis of the relationship between hyperphagia and behavioural and emotional problems. Multiple linear regression was planned for the analysis of this study. Listed below is a justification for why each variable would be included in this analysis:

Age was controlled for, as a variety of behavioural problems in PWS have been found to increase with age (Dykens & Kasari, 1997; Dimitropolous et al. 2001; Dykens, 2004; Steinhausen et al. 2004). For example; Steinhausen et al. (2004) examined challenging behaviour in people with PWS aged from two to 29 years old. They found that the prevalence of all forms of problem behaviours was significantly higher for the older age group (over 13 years old) than the younger age groups (under 6, and 7-13 years).

Weight was also controlled for, as some previous studies with people with PWS have found that the higher the body mass index (BMI), the more behavioural problems displayed. For example; Steinhausen, et al. (2004) found a relationship between challenging behaviour and body mass index (BMI) in children and young adults with PWS. However, Dykens and Cassidy (1995) found that the opposite was true; that participants with lower BMI’s had higher problem behaviour scores. Furthermore Ackefeldt and Gillberg (1999) and Dykens and Kasari (1997) both found no association between weight and behaviour in PWS.

Gender was included, as previous studies with children with intellectual disabilities have suggested that boys display significantly more behavioural and emotional problems than girls, when all other factors are matched (Einfeld, Piccinin, MacKinnon et al. 2006; Einfeld & Tonge, 2002; Emerson & Hatton, 2007). However, studies with
people with PWS have found mixed results. Some studies have reported no effect of
gender on the behaviour of people with PWS (Dykens & Kasari, 1997; Einfeld et al.
1999; Steinhausen et al. 2004; Symons et al. 1999). On the other hand, some
researchers have reported that boys display significantly more externalising
problems, aggressive behaviour and higher levels of depression, whilst girls display
more self-harm behaviour and anxiety (Dykens & Cassidy, 1995; Dykens, 2004).

Furthermore, the level of intellectual disability has been found to be associated with
level of emotional and behavioural problems in children with learning disabilities, with
people with severe intellectual disabilities experiencing higher levels of behavioural
problems than those with mild, moderate or profound learning disabilities (Einfeld et
al. 2006; Einfeld & Tonge, 2002). In PWS, Einfeld et al. (1999) found a relationship
between level of intellectual disabilities and behaviour, with those with more severe
intellectual disabilities displaying higher levels of disruptive and antisocial behaviour.

It was hypothesised that there would be a relationship between hyperphagia and
level of emotional and behavioural problems, even when the child’s age, gender,
level of LD and weight was taken into account.

In addition to this, as the HQ has only recently been developed and has not yet been
extensively used in research, the aim was to use the data collected from this study to
assess the reliability of the Hyperphagia Questionnaire as a tool for measuring
hyperphagia in PWS. Part of this would involve repeating a factor analysis on the
data to assess the reliability of the three factor structure found by Dykens et al
(2007).

1.4.2. Additional Aims
Some additional aims were also identified, based on the literature discussed. These
were as follows:

- Dykens, Maxwell, Patino, Kossler and Roof (2007) found that hyperphagia in
  PWS could conceptually and statistically be divided into the following three
domains, as measured by the three subscales of the HQ:
  - Hyperphagic Behaviour: indicates attempts to obtain, steal or forage for
    food.

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- Hyperphagic Drive: indicates distress when denied food and/or the ease of re-directing them from food related activities or discussions.
- Hyperphagic Severity: indicates the amount of time engaged in food behaviour or how much food interferes with daily routines.

Furthermore, they reported that the level of hyperphagic behaviour problems was not related to behavioural and emotional problems in their sample. In fact, only Hyperphagic Drive and Hyperphagic Severity was associated with non-food related behavioural problems. Therefore, as part of the additional analysis, the aim was to examine how well the three original subscales of the HQ predict overall behavioural and emotional problems.

- As mentioned above, the research into the relationship between gender and behaviour in PWS has yielded mixed results. Some studies found no relationship between gender and behaviour in PWS when examining overall levels of behaviour problems (Dykens & Kasari, 1997; Einfeld et al. 1999; Steinhausen et al. 2004; Symons et al. 1999). However Dykens (2004) and Dykens and Cassidy (1995) both found a relationship between gender and more specific types of behavioural problems in PWS (e.g. self-harm, anxiety, aggression and depression). Therefore the aim was to examine participant’s scores on the five behavioural subscales of the DBC-P (e.g. Disruptive/Antisocial, Self-Absorbed, Communication Disturbances, Anxiety and Social Relating problems) and to examine whether or not males and females differ on any of these aspects of behaviour.

- It is believed that individuals with PWS display higher levels of behavioural and emotional problems than individuals with intellectual disabilities of other aetiologies, when matched for age, gender and level of LD (Einfeld et al. 1999). Therefore, to examine this further, the aim was to compare the total level of behavioural and emotional problems in children with PWS with the normative sample in the DBC-P (Einfeld & Tongue, 2002) to see whether there is a significant difference in the level of emotional and behavioural problems between the groups.
Section Two: Extended Method

2.1 Exploration of Measures

2.1.1: Behavioural and Emotional Problems

Many different methods of assessing the behaviour of children with intellectual disabilities exist, such as clinical interviews, behavioural questionnaires, direct observational methods, and physiological methods (Einfeld & Tongue, 2002). However, for this study, a parental-report measure of behavioural and emotional problems in childhood was required. The measure selected for use in this study was required to be valid for children aged from four to 18 years old and for children with intellectual disabilities. What follows is not an exhaustive list of possible measures for behaviour in children with intellectual disabilities; rather it is a summary of the key measures considered for this study.

One of the most common measures of challenging behaviour and emotional and behavioural problems used in research with children and adults with PWS is the Child Behavior Checklist (CBCL: Achenbach, 1991, used in: Dykens, Cassidy & King, 1991; Dykens et al. 2007; Wallander, Dekker & Koot, 2005). This measure comprises of 118 items which describe problem behaviours in children and adolescents. Parents and carers are asked to indicate the extent to which items apply to their child using a 3-point Likert-style scale (0 = not true; 1 = somewhat true; 2 = very true). The measure then provides a total score of the overall level of problem behaviours and also provides scores on:

- Social Competence
- Behaviour Problems
- Internalising Problems
- Externalising Problems
- Sex Problems

In addition to this, scores can be obtained on eight ‘syndrome scales’ encompassing: Social withdrawal; somatic complaints; anxiety/depression; social problems; thought problems; attention problems; delinquent behaviour; and aggressive behaviour (Achenbach, 1991). From extensive research, clinical cut-off scores have been devised for the syndrome scales, the total problem score and the internalising and
externalising subscales, to provide researchers with an idea of whether a score falls into the ‘normal’, ‘borderline’ or ‘clinical’ range (Achenbach, 1991). Extensive reliability and validity analyses have shown that the CBCL has good retest reliability over seven days (Correlation coefficients ranging from .74 to .89) and internal consistency was also good, with Cronbach’s alphas ranging from .62 to .92 for boys aged from four to 11 years and .66 to .92 for girls aged from four to 11 years (Achenbach, 1991). It has also been shown to have strong construct validity (Achenbach, 1991). However, the CBCL was actually developed in the USA as an instrument for assessing the psychopathology of children within the general population, and not for children with intellectual disabilities. A number of the items within the measure have been criticised for addressing thoughts and behaviours that may be too complex or sophisticated to apply to children with moderate or severe intellectual disabilities (Einfeld & Tonge, 2002). Furthermore, Aman et al. (1996) stated that the expression of challenging behaviour or psychopathology in children with intellectual disabilities is often very different to children without; therefore specific instruments are required for this group.

The CAMHS outcome research consortium for the UK (CORC, 2006) has recently compiled a list of recommended measures to use with children with and without intellectual disabilities. It recommends the use of the Strengths and Difficulties Questionnaire (SDQ- Goodman, 1997, 1999, 2001) for measuring behaviour in all children. The SDQ is a 25-item Likert-style measure which explores both the positive and negative aspects of a young person’s behaviour. It has shown good levels of both reliability and validity (Emerson, 2005; Goodman, 2001), and has been utilised in wide-scale national research studies investigating child behaviour (Green, McGinnity, Meltzer, Ford & Goodman, 2005). Emerson (2005) stated that the SDQ is a straightforward and robust measure of behaviour and mental health problems in young people with intellectual disabilities. However, the main limitation is that there are different versions of the measure for 3-4 year olds, 4-16 year olds and 11-17 year olds. There is not a version which has been validated across the required 4-18 age range.

In addition to the SDQ, the Nisonger Child Behaviour Rating Form (Nisonger CBRF-Aman, Tasse, Rojahn & Hammer, 1996) was developed to assess the severity of
behavioural and emotional problems, specifically in children with intellectual disabilities. The Nisonger CBRF consists of a total of 76-items, with 66 items measuring challenging behaviour and 10 items measuring pro-social behaviours. It then provides a score on eight behavioural subscales (Aman et al. 1996):

- Compliant/Calm
- Adaptive social
- Conduct problem
- Insecure/Anxious
- Hyperactive
- Self-injury/Stereotypic
- Self-isolated/ritualistic
- Overly sensitive

The measure has demonstrated good levels of internal consistency: on the problem behaviour subscales the median Cronbach’s alpha score was .84, whilst on the prosocial behaviour subscales the median Cronbach’s alpha score was .78 (Aman et al. 1996). Furthermore, this measure has good levels of content validity when correlated with other measures of behaviour (Waltz & Benson, 2002). The measure has been widely used in research with children with intellectual disabilities (Lecavalier, 2006; Sarimski, 2004) and has also been utilised with PWS samples (Waltz & Benson, 2002). However, like the SDQ this measure is only recommended for use with children aged between four and 16 years old, as it has not been validated on an older teenage population (Aman et al. 1996).

One other popular measure for use with children with PWS is the Developmental Behaviour Checklist (DBC; Einfeld & Tonge, 1992, 1995, 2002). This measure is structurally very similar to the CBCL (Einfeld et al. 2000) but it has been specifically developed for use with children with intellectual disabilities. It is a 96-item informant response questionnaire designed to assess behavioural and emotional disturbances (in children and adolescents aged from four to 19 years). Parents and carers are asked to indicate the extent to which items apply to their child using a 3-point Likert-style scale (in which 0 = not true; 1 = somewhat true or sometimes true; 2 = very true or often true). The measure then provides a ‘Total Problem Behaviour Score’ which provides an indication of the severity of any behavioural/emotional disturbances. The
measure can also provide a total score on one of the five behavioural subscales, including:

- Disruptive/Antisocial Behaviour: Manipulative, irritable, lies, kicks, hits, abusive
- Self-Absorbed Behaviour: Preoccupation with certain items, eating non-food items, gorging food.
- Communication Disturbance: Perseveration, Talks to self, echolalia
- Anxiety: Appears distressed, shows fears, cries easily
- Social Relating: Not showing affection, aloof, not regarding other’s feelings.

These subscales are based on behavioural problems frequently observed in young people with learning disabilities and they exclude behaviours which may be related to the disability itself (e.g. does not speak). However, they are not based on specific diagnostic categories or constructs, therefore should not be used for diagnosis (Dekker et al, 2002).

To assist in the interpretation of the results from the DBC, normative data is available from a general population of young people with intellectual disabilities (n=454). This data has been used to develop percentiles based on total score, which have also been broken down by age, gender and IQ level (Einfeld & Tongue, 2002). Furthermore, a clinical cut-off score has been provided for the total problem behaviour score. This scale was originally developed in Australia as an instrument for assessing the psychopathology of children with intellectual disabilities. However it has been used extensively within Europe and more specifically with PWS samples (Clarke et al. 2002; Einfeld, Smith, Durvasula, Florio & Tonge, 1999). Extensive reliability and validity analyses have been conducted on the DBC and the measure has been found to have good levels of internal consistency (Cronbach’s alpha= .94) and test-retest reliability with parents (intra-class correlation= .80) (Einfeld & Tonge, 1992). Furthermore, it was shown to have high criterion group validity in differentiating clinical from non-clinical cases and strong criterion and concurrent validity (Dekker, Nunn & Koot, 2002; Einfeld & Tonge, 2002). In a recent study using Australian and European samples, the factor structure of the five subscales was
found to be strong, accounting for 44% of the total variance (Dekker, Nunn, Einfeld, Tonge & Koot, 2002).

In summary, following the review of the available measures for this study, it was decided that the DBC was the most appropriate and robust tool available for the measurement of emotional and behavioural problems in this study. This was because it was specifically developed for children with intellectual disabilities and has been well validated within PWS populations. Furthermore, it covered all aspects of emotional and behavioural problems which may be experienced by children with PWS.

Reliability analysis was conducted on the DBC to ascertain the internal consistency of the overall Total Problem Behaviour Score and also of the five subscales in the current study. The scores are shown in Table 5 below. All scores fell on or above the cut-off point of .7 recommended by Pallant (2007). This indicates good levels of internal consistency for all DBC subscales.

Table 5

<table>
<thead>
<tr>
<th>Scale</th>
<th>No. Items</th>
<th>Cronbach’s Alpha</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total Problem Behaviour Score</td>
<td>95</td>
<td>.94</td>
</tr>
<tr>
<td>Disruptive/Antisocial Behaviour Subscale</td>
<td>26</td>
<td>.85</td>
</tr>
<tr>
<td>Self-Absorbed Behaviour Subscale</td>
<td>30</td>
<td>.82</td>
</tr>
<tr>
<td>Communication Disturbance Subscale</td>
<td>12</td>
<td>.71</td>
</tr>
<tr>
<td>Anxiety Subscale</td>
<td>9</td>
<td>.69</td>
</tr>
<tr>
<td>Social Relating Subscale</td>
<td>10</td>
<td>.74</td>
</tr>
</tbody>
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2.1.2: Hyperphagia
A parental-report measure of hyperphagia was also required. Prior to selecting a measure for this study, the measurement of hyperphagia in PWS was reviewed. It emerged that a wide variety of approaches to the measurement of hyperphagia in PWS have been employed in the past, such as; the direct observation of eating behaviour; the use self-report methods (interviews and questionnaires with
individuals with PWS); and informant based questionnaire methods (often completed by parents or residential staff). The strengths and weaknesses of these methods will be explored below, concluding with an overview of the measure selected for this study.

Firstly, researchers have used observational methods, whereby direct observation of eating behaviour is undertaken. These studies either consist of leaving individuals with PWS (and healthy weight or obese controls) in a room with unlimited access to food and measuring how much they consume in a given time period (Holland, Treasure, Coskeran, Dallow, Milton & Hillhouse, 1993; Holland, Treasure, Coskeran & Dallow, 1995; Young et al. 2006). Alternatively, researchers have given a set quantity of food to individuals and used scales placed under their plates to measure the rate of eating compared with control groups (Lindgren et al. 2000). These studies have been useful for confirming the idea that individuals with PWS often display appetitive behaviour that is distinct from individuals with healthy weights and those who are obese. In particular they found that in general individuals with PWS will eat larger quantities of food over longer periods and appear to possess an impaired satiation response (Lindgren, 2003). However, the main shortcoming of these approaches is that they often used adults with PWS and very small sample sizes, therefore the results cannot necessarily be extended to the general population of people with PWS, particularly children or adolescents. Furthermore, it is important to note that information extracted from such artificial environments may not accurately reflect the behaviour observed by parents and carers in the home environment.

There have also been studies, which have conducted interviews with individuals with PWS in order to explore their food seeking behaviour. Young et al. (2006) conducted a survey with 19 individuals with PWS, seven with intellectual disabilities of other aetiologies and 25 typically developing individuals. The exact age range of participants was not provided however, the PWS group was aged between aged one and 50 years, the intellectual disabilities group was aged one to 17 years and the typically developing group was aged from one to 50+ years (mean ages were not provided). The acceptability of food in different situations was explored through interviews in which participants were shown digital photos of food matched with different environments and asked to rate which of the items they would be willing to
eat. They found that, in the response to the survey, the PWS group differed from the typically developing group, but not from the control group with intellectual disabilities and that both groups provided inconsistent responses. It was concluded that inconsistent responses in these two groups, may be related to the intellectual disabilities rather than their actual food preferences and that they may have simply been related to confusion with the survey instructions. This indicates that the intellectual disabilities in individuals with PWS may have affected their ability to provide accurate self-report information on their eating behaviour. It has been argued that people with intellectual disabilities will often struggle with reliably reporting on their behaviour and are more likely to be affected by such variables as social desirability (Jones, Miller, Williams & Goldthorp, 1997). Therefore, any results from this study must be viewed with caution, as the self-report information may be unreliable.

In addition to the direct methods with individuals with PWS, a number of informant-based studies have also been designed using carer/parental report questionnaires. For example, the Children’s Eating Behaviour Inventory (Archer, Rosenbaum & Streiner, 1991) was utilised by Sarimski (1996) to assess the eating behaviour of people with PWS. This is a 40-item measure which asks parents or carers to rate the frequency of their child’s problematic eating behaviour on a five point Likert-style scale where 1= Never and 5 = Always, they are then asked to indicate whether this behaviour represents a problem for them with a yes/no response. The measure has demonstrated good levels of test-retest reliability, with the correlation co-efficient of the total problem eating score at .87 and showed acceptable levels of internal consistency (Cronbach’s alpha ranging from .58 to .76). However, only one person with PWS was recruited for the study which examined the reliability and validity of the scale (Archer et al. 1991). Furthermore, this measure was designed for use with other clinical groups such as children with eating disorders, picky eating and autism, and therefore it is unlikely to be able to adequately encompass the experience and the breadth of behaviour shown by individuals with this rare syndrome (Dykens et al. 2007). Therefore, this measure may well lack validity in its use for people with PWS. This has not been assessed to date.
Gilmour and Skuse (2003) also utilised a 23 item parental report questionnaire relating to eating behaviour and appetite problems in children and young people. This measure was developed directly from a semi-structured interview schedule utilised in research into hyperphagia in other clinical groups (Skuse, Albanese, Stanhope, Gilmour & Voss, 1996). However, this measure has not been subjected to reliability or validity analysis; therefore, it cannot be considered to be a robust measure.

In addition to these measures, Russell and Oliver (2003) went on to devise the Food Related Problems Questionnaire (FRPQ) to specifically measure food-related problems in individuals with PWS. This measure was developed through focus groups and semi-structured interviews with parents of individuals with PWS. These interviews focussed on all aspects of food-related behaviour and led to the creation of a 16-item informant response measure, with three main subscales:

- Preoccupation with food
- Impairment of satiety
- Other food-related challenging behaviour, including
  - Eating inedible items (‗Pica’)
  - Storing foods inappropriately or hoarding foods
  - Responding inappropriately when food is not available

Parents and carers were asked to rate their child’s behaviours on a seven-point Likert-style scale, ranging from 0=never to 6=always. This measure was piloted with the parents, carers or key workers of: adults with PWS living in residential care (n=23, mean age: 27.7 years); adults with intellectual disabilities of other aetiologies also living in residential care (n=12, mean age: 43.1 years); and a community sample of children and adults with PWS (n=, mean age: 18.3). In reliability and validity analyses, the measure showed good levels of internal consistency (Cronbach’s alpha for the total FRPQ score was good at .87). Furthermore, test-retest and inter-rater reliability coefficients for the total FRPQ score were both .86.

However, this questionnaire’s main limitation was that it required verbal responses from individuals with PWS on six of the sixteen items. For example: Question 8 - ‘after a normal sized meal, how often will the person say they still feel hungry?’ and Question 15 - ‘Does the person ever describe “feeling full”?’ (Russell & Oliver, 2003,
These questions are problematic; firstly due to the fact that people with PWS have varying levels of intellectual abilities and in moderate to severe intellectual disabilities, this could lead to impaired verbal communication skills. Secondly, these questions require the individual with PWS to discuss their feelings about food or eating behaviour with their carers, these conversations are often very sensitive to people with the syndrome and they may be less likely to engage in such discussions (Dykens, et al. 2007). Finally, and perhaps most saliently, the measure could not provide information on the severity of hyperphagia, which makes its use in research and clinical practice quite limited in terms of exploring the impact of hyperphagia on other aspects of behaviour (Dykens, et al. 2007).

As mentioned in the literature review, to address the shortcomings of the FRPQ Dykens et al. (2007) devised the Hyperphagia Questionnaire (HQ). The individual items for the HQ were extracted from reports from carers of people with PWS obtained during lengthy research and clinical work with individuals with PWS and their parents and carers. The severity items were also derived from the definitions of symptoms and impairments from the fourth edition of the Diagnostic and Statistical Manual of Mental Disorders (American Psychiatric Association, 2000). Following the development of the measure, its psychometric properties were assessed using a sample of parents and carers of children and adults with the syndrome (n= 153) aged from four to 51 years of age. Exploratory factor analysis on this measure revealed a clear three factor structure assessing three key components of hyperphagia:

- **Hyperphagic Behaviour Subscale**
  - Contains five items around the person’s attempts to obtain, steal or forage for food

- **Hyperphagic Drive Subscale**
  - Contains four items around the person’s distress when denied food and the ease of re-directing them from food related activities or discussions.

- **Hyperphagic Severity Subscale**
  - Contains two items around the amount of time engaged in food behaviour or how much food interferes with daily routines or functioning.
These factors were able to account for 58.93% of the total variance. The Hyperphagic Behaviour subscale accounted for 34.47% of variance and showed good levels of internal consistency (Cronbach’s alpha = .76). The Hyperphagic Drive Subscale accounted for 15.28% of the variance and also showed good levels of internal consistency (Cronbach’s alpha = .80). Finally, the third subscale, Hyperphagic Severity accounted for 9.17% of the variance and showed acceptable levels of internal consistency (Cronbach’s alpha = .60). Items on this measure are rated on a 5-point Likert-style scale by either the severity or the frequency of the problem, for example: responses may range from 1 = not a problem, up to 5 = severe and/or frequent problem; or from; 1 = never, up to 5 = 4-7 times a week. The measure then provides a total hyperphagia score, followed by individual scores for hyperphagic severity, hyperphagic drive and hyperphagic behaviour.

In summary, although the Hyperphagia Questionnaire has only had limited use so far in Prader-Willi research, it appears to be the most robust and valid measure of hyperphagia in PWS at this time. Therefore, this measure was selected for us in this study, with the acknowledgement that further validation of the measure is required and further research is need to explore whether assessment of the behavioural expression of hyperphagia is the best way to assess its severity.
2.2: Ethical Approval

Prior to commencement of this study, full ethical approval was gained through the Institute of Work Health and Organisations ethics committee at the University of Nottingham (See Appendix 4.0 for a copy of the ethical clearance).

The key ethical point considered for this research was that having a child with a complex genetic syndrome and/or challenging behaviour may be a sensitive or distressing subject for parents and carers to discuss. To address this, it was emphasised to potential participants that participation in the study was entirely voluntary and that parents or carers could choose to not take part with no penalty to themselves or their family. It was also emphasised on the information sheet that participants would be able to withdraw from the study at any point during the data collection process if they decided that they would no longer like to take part (this was facilitated through the provision of a randomly assigned reference number on each questionnaire pack). Finally, the full contact details of the lead researcher were also provided to address any concerns or queries participants may have had about participating in the study. In addition to this the contact details for the PWSA-UK were provided for support and advice in case of any concerns or distress arising from participating in the study.

The second key ethical consideration in this study was that researchers planned to only seek informed consent from the parents and carers of children with PWS, rather than the children themselves. Informed consent from children with PWS was not sought separately, as individuals with PWS vary widely in terms of their intellectual abilities, ranging from average ability to severe or profound learning disabilities (Whittington, Holland, Webb, Butler, Clarke & Boer, 2004). Furthermore, the children and young people were aged between four and eighteen years, so it is likely that this group will have had extremely varied levels of literacy and understanding. The British Psychological Society code of conduct and ethical guidelines (BPS, 2006) states that when obtaining informed consent from children is not possible in research, consent can be obtained from parents or those in loco parentis instead. This is further supported by the fact that other studies with parents and carers of people with PWS have not sought informed consent from individuals themselves (Dykens et al. 2007; Russell & Oliver, 2003).
In addition to the above, participants’ anonymity was preserved by not asking them for any personal or identifying information on any of the response sheets (such as names, date of births or addresses).
2.3: Calculation of Body Mass Index

Body mass index (BMI) is a measure of body fat based on weight and height and was calculated for all ages and genders, using the following calculation:

$$\text{BMI} = \frac{\text{Bodyweight (kilograms)}}{(\text{Height [metres]}^2}$$

(Taken from Cole, Bellizzi, Flegal & Dietz, 2000, page 1)

This calculation was used for calculating the BMI of the children and adolescents in this study.
2.4 Pre-Test Sample Size Calculations

Linear regression was planned for the analysis of this study. The aim was to examine how well a maximum of six predictor variables (hyperphagic behaviour, Hyperphagic drive; age; gender; IQ and weight) could account for levels of behavioural and emotional problems (measured using the subscales of the Developmental Behaviour Checklist – below) in children with PWS.

For multiple linear regression, Field (2005) states that a minimum of 15 cases per predictor variable is required to achieve a suitable level of power. Therefore for six predictor variables, a minimum of 90 cases would be required.
Section Three: Extended Results

3.1: Participants

- $n=350$ Parents and carers invited to participate
- $n=105$ Responses Received
- $n=245$ Did not respond
- $n=15$ Excluded, as the child was under 4 years old
- $n=3$ Excluded, as the child did not reside with them
- $n=1$ Excluded, as significant amount of missing data*
- $n=86$ Parents and carers met inclusion criteria

* = See section 3.3.1 for an explanation of how missing data was assessed

Figure 2. Participant Inclusion Flow Chart
3.2: Demographic Information

3.2.1. Age at Diagnosis

Participants were asked the age at which their child was diagnosed with PWS. The mean age at diagnosis was 7.92 months old (SD: 16.03 months), with a range of between birth and 7½ years. The mode was 1 month old.

3.2.2. Level of Learning Disabilities

Only seven respondents provided information on their child’s IQ. Of those, the mean value was 68 (SD: 20), with a range of 37-98. This variable was excluded from further analysis for not having enough data points to achieve a desirable level of power.

In addition to being asked about IQ, on the demographic questionnaire, parents were asked: ‘What level of learning disability does your child have?’ On this question 58.8% of parents responded that their child had moderate learning disabilities and 10.6% responded that their child has severe learning disabilities (see Figure 3). This question may have a number of limitations, as the term ‘learning disability’ may have many different definitions and interpretations. Furthermore, the question required parents to estimate their child’s level of intelligence. Previous research has found that parent’s estimates of their child’s level of intelligence can be extremely unreliable, as estimates are significantly influenced by the age and gender of the parent as well as the age and gender of the child (Furnham & Gasson, 1998; Furnham, 2000). For example, in a study of British parents (n=184), Furnham and Gasson (1998) found that parents rated the intelligence of sons higher than daughters and that older parents gave higher estimates of intelligence than younger. Furthermore, it has been shown that mothers tend to rate intelligence lower overall than fathers.

In addition to this, the figures obtained in this study do not appear to accurately reflect the PWS population, as the average IQ of a person with PWS is around 60-70 points (mild learning disability) (Whittington et al 2004). This indicates that parents may have underreported their child’s intelligence. Therefore, based on all of the factors, the LD category was not utilised in further analysis.
Figure 3. Bar chart of the frequency of parent’s responses to ‘What level of learning disability does your child have?’

On the demographic questionnaire, parents were also asked to provide information on the type of school attended by their child: 55.8% of the PWS children attended mainstream schools, whilst 32.6% attended a ‘special’ school (for those with special educational needs). The remainder attended mainstream Nurseries or Colleges (See Figure 4). The high rate of people in mainstream education provides a further indication that the high rate of endorsement of the moderate to severe learning disabilities category by the parents may be inaccurate.

Figure 4. Bar chart of the frequency of parent’s responses to ‘What type of school does your child attend?’
3.2.3. Age of onset of Hyperphagia

Sixty-three parents and carers provided information on their child’s age at onset of the hyperphagia. Of these, the mean age of onset was 4.05 years (SD: 2.4 years), with a range of 6-months to 16 years old. This fits with previous research which suggests that hyperphagia does not begin until between the ages 18 months and six-years (Cassidy, 1984; Clarke, Boer, Chung, Sturmey, & Webb, 1996; Dimitropoulos et al. 2000).

3.2.4 DBC-P Overall Problem Rating

As part of the DBC-P, each parent was asked to rate whether or not they felt that their child had emotional or behavioural problems. Of the 75 parents who answered this question, 20% responded ‘no problems with feelings or behaviour’, 42.7% responded that their child had ‘minor problems with feelings or behaviour’ and 37.3% responded that their child had ‘major problems with their feelings or behaviour’. Figure 5 represents these results.

![Figure 5. Bar chart representing parent’s responses to ‘overall, do you feel that your child has problems with feelings or behaviour’ in addition to problems with development?](image-url)
3.3 Factor Analysis

3.3.1 Prior Analysis

Prior to conducting factor analysis on the Hyperphagia Questionnaire, the data was screened to ascertain the factorability of the data. Initially, a correlation matrix was created (See Table 6). This revealed that all items on the scale correlated at least .3 with another item on the scale, supporting the factorability of the measure. Furthermore, no items correlated above .9 with other items. This signifies that there were no problems with multicollinearity in the data (Field, 2005).

Table 6

*Showing inter-item correlation matrix for the Hyperphagia Questionnaire*

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Following this, the Kaiser-Meyer-Olkin measure of sampling adequacy was examined. This was .868, which is above the recommended level of .6 (Pallant ). Furthermore, Bartlett’s test of sphericity was significant. The diagonals of the anti-image correlation matrix were also examined and all items had figures over .7, this suggests that all items shared some common variance with other items on the measure Proportion of common variance (average = 7.857/11 = .714) above .7, with less than 30 variables and a small sample size, this is good using Kaiser’s criterion. Given these overall results, it was decided that factor analysis could be conducted on all 11 items of the measure.
3.3.1 Scree Test

**Figure 5. Scree Plot for Factor Analysis on the Hyperphagia Questionnaire**

Using Cattell’s scree test, it was decided to retain two components for further analysis, as the graph began to plateau after the second factor (Field, 2005).
3.4 Initial exploration of the data: Normality, Distribution, Outliers

Initial exploration of the data was conducted to check for outliers, missing data and finally to assess whether the data met the criteria for parametric tests. Previous research on this population, examining hyperphagia and behaviour in PWS suggested that data would meet the criteria for parametric tests (Dykens et al. 2007; Russell & Oliver, 2003).

3.4.1 Missing Data

The data set was scanned for missing information. Where this was found, the response sheets were examined to check for data entry errors or omissions. Where a significant amount of data (more than 50%) was found to be missing, from a particular measure (e.g. the HQ or the DBC-P) then cases were excluded listwise (this happened with one case). Where there was not a significant amount of data missing, from a particular measure, cases were excluded pairwise.

3.4.2 Outliers and Distribution of data

Histograms and boxplots were then created for each of the variables to provide a visual indication of any outliers and the distribution of the data. However, further assessment of the distribution was undertaken. Firstly, the values of skewness and kurtosis were examined, and from these, z-scores were calculated so that the scores were standardised. These were calculated using the following equation, taken from Field, 2005 (p.72)

\[ z_{\text{skewness}} = \frac{S - 0}{SE_{\text{skewness}}} \]

Field (2005) recommends that these z-score values should fall between 1.96 and -1.96, any scores above or below this value should be considered as skewed and may not be appropriate for parametric methods.

3.4.2.1 BMI/Weight Category

Initial boxplot exploration of the BMI data revealed one significant outlier (see Figure 6). This is due to the fact that for one respondent, BMI was recorded as 82. Investigation into this data revealed that it was likely to be a response error; therefore this data point was removed from the database.
A Histogram (with normal curve) was then completed to assess whether or not the BMI data was normally distributed. Figure 7, shows that this variable was significantly positively skewed. This was supported by the $z$ skewness figure calculated at 4.46 (significantly over the recommended cut-off of 1.96) and a Kolmogorov-Smirnov test (K-S 0.132, $p= .008$).

Further exploration of this revealed that although BMI is calculated in the same way for children as for adults, the figures are not on a continuous scale
for children, as the meaning of the BMI value is heavily dependent on the age and the gender of the child (Dietz & Robinson, 1998). Therefore, Cole et al. (2000) published internationally validated cut-off points for overweight and obesity in childhood, which are age and gender specific. These cut-off points were utilised to categorise children’s weight into two categories: healthy weight or overweight/obese (see Appendix 4 for the cut-off points used).

3.4.2.2. Age
A box plot (Figure 8) revealed no outliers in the age data.

![Boxplot of Age](image)

*Figure 8. Boxplot of Age*

A Histogram (with normal curve) of the age data (Figure 9) appeared slightly skewed; although the $z$Skewness and $z$kurtosis values both fell within the acceptable range (-1.96 to 1.96). Although, a Kolmogorov-Smirnov (K-S) test indicated that data were not normally distributed (K-S 0.129, $p=.01$). However, some researchers have argued that the K-S test can often find distributions to be significantly non-normal, when in fact they are relatively symmetric and smooth and suitable for use in research (Micceri, 1989). Therefore this result was viewed with caution, given that the $z$Skewness and $z$kurtosis values were acceptable, as was the histogram.
3.4.2.3. Hyperphagia Total Score (from the Hyperphagia Questionnaire)

A box plot (Figure 10) revealed no outliers in the total score data.

A Histogram (with normal curve) of the data (Figure 11) appeared slightly positively skewed; the $z$Skewness value was calculated at 2.57 (which is slightly over the recommended cut-off of 1.96). $z$Kurtosis values fell within the acceptable range (-1.96 to 1.96). However, a Kolmogorov-Smirnov test confirmed that data were normally distributed ($K-S 0.105, p=0.079$).
Figure 11. Histogram of Hyperphagia Total Scores

3.4.2.4. Hyperphagic Drive Score (from the Hyperphagia Questionnaire)
A box plot (Figure 12) revealed no outliers in the Hyperphagic expression scores.

Figure 12. Boxplot of Hyperphagic Expression Scores

A Histogram (with normal curve) of the data (Figure 13) appeared slightly kurtotic; however both the $z$Skewness and $z$Kurtosis values fell within the acceptable range (-1.96 to 1.96). A Kolmogorov-Smirnov test confirmed that data were normally distributed (K-S 0.97, p=0.055).
3.4.2.5. Hyperphagic Behaviour Score (from the Hyperphagia Questionnaire)

A box plot (Figure 14) revealed one outlier in the total score data. This was assessed for data entry error, but was confirmed to be a correct score.

Field (2005) recommends that where significant outliers are found, they may be replaced with a score representing the mean plus two standard deviations. Therefore the problematic case was corrected as such. Following this, further
analysis of normality was completed. A Histogram (with normal curve) of the data was created (Figure 15). This appeared positively skewed; the zSkewness value was calculated at 4.25 (significantly over the recommended cut-off of 1.96). zKurtosis values fell within the acceptable range (-1.96 to 1.96). However, a Kolmogorov-Smirnov test confirmed that data were not normally distributed (K-S 0.228, p<.001).

![Histogram](image)

*Figure 15. Histogram of Hyperphagic Action Scores*

3.4.2.6. Total Problem Behaviour Score (from the DBC)
A box plot (Figure 16) revealed one outlier in the total score data. This was assessed for data entry error, but was confirmed to be a correct score.
A Histogram (with normal curve) of the data (Figure 17) appeared slightly skewed towards the right; although the $\zeta$Skewness and $\zeta$Kurtosis values both fell within the acceptable range. A Kolmogorov-Smirnov test confirmed that data were normally distributed (K-S 0.073, $p=0.20$). 

![Boxplot](image16.png)

*Figure 16. Boxplot of Total Behaviour Problem Scores*

![Histogram](image17.png)

*Figure 17. Histogram of Total Problem Behaviour Scores*

3.4.2.7. Disruptive/antisocial Behaviour Score (from the DBC)
A box plot (Figure 18) revealed some outliers in the Disruptive/Antisocial score data.
The dataset was examined to check for data entry errors, however these were found to be true scores. Field (2005) recommends that where significant outliers are found, they may be replaced with a score representing the mean plus two standard deviations. Therefore the scores of the two problematic cases were replaced with this value. Following this, further analysis of normality was completed. A Histogram (with normal curve) of the data (Figure 19) appeared slightly skewed towards the right; although the $z$Skewness and $z$kurtosis values both fell within the acceptable range. A Kolmogorov-Smirnov test confirmed that data were normally distributed (K-S 0.086, $p=0.19$).
3.4.2.8. Self-Absorbed Behaviour Score (from the DBC)

A box plot (Figure 20) revealed one outlier in the total score data. This was assessed for data entry error, and confirmed to be a correct score. Field (2005) recommends that where outliers are found, they may be replaced with a score representing the mean plus two standard deviations. Therefore the problematic case was corrected as such. Following this, further analysis of
normality was completed. A Histogram (with normal curve) appeared slightly skewed (Figure 21); the $z$Skewness value was 2.28 (which is slightly over the recommended cut-off of 1.96) although the $z$Kurtosis value fell within the acceptable range. A Kolmogorov-Smirnov test indicated that data was not normally distributed ($K$-S 0.115, $p<.05$). However, some researchers have argued that the K-S test can often find distributions to be significantly non-normal, when in fact they are relatively symmetric and smooth and suitable for use in research (Micceri, 1989). Therefore this result was viewed with caution, given that the $z$Skewness and $z$Kurtosis values appeared acceptable, as did the histogram.

![Histogram of Self-Absorbed Behaviour Scores](image)

*Figure 21. Histogram of Self-Absorbed Behaviour Scores*

3.4.2.9. Communication Disturbance Score (from the DBC)
A box plot (Figure 22) revealed no outliers in the communication disturbance scores.
Figure 22. Boxplot of Communication Disturbance Scores

A Histogram (with normal curve) of the data (Figure 23) appeared acceptable and the skewness and kurtosis values both fell within the acceptable range. A Kolmogorov-Smirnov test confirmed that data were normally distributed (K-S 0.076, p=0.20).

Figure 23. Histogram of Communication Disturbance Scores

3.4.2.10 Anxiety Score (from the DBC)
A box plot (Figure 24) revealed one outlier in the Anxiety data. This was assessed for data entry error, but was confirmed to be a correct score.

![Boxplot of Anxiety Scores](image)

*Figure 24. Boxplot of Anxiety Scores*

Field (2005) recommends that where significant outliers are found, they may be replaced with a score representing the mean plus two standard deviations. Therefore the problematic case was corrected as such. Following this, further analysis of normality was completed. A Histogram (with normal curve) of the data (Figure 25) appeared acceptable and the $z$Skewness and $z$kurtosis values both fell within the acceptable range. However, a Kolmorgorov-Smirnov test suggested that data was not normally distributed (K-S 0.123, $p=0.003$). However, some researchers have argued that the K-S test can often find distributions to be significantly non-normal, when in fact they are relatively symmetric and smooth and suitable for use in research (Micceri, 1989). Therefore this result was viewed with caution, given that the $z$Skewness and $z$kurtosis values were acceptable, as was the histogram.
3.4.2.11. Social Relating Score (from the DBC)

A box plot (Figure 26) revealed one outlier in the total score data. This was assessed for data entry error, but was confirmed to be a correct score.

Figure 26. Boxplot of Social Relating Scores

A Histogram (with normal curve) of the data (Figure 27) appeared acceptable and the $z$-Skewness and $z$-kurtosis values both fell within the acceptable range. However, a Kolmorgorov-Smirnov test suggested that data were not normally distributed ($K-S 0.118, p=0.006$). However, some researchers have argued...
that the K-S test can often find distributions to be significantly non-normal, when in fact they are relatively symmetric and smooth and suitable for use in research (Micceri, 1989). Therefore this result was viewed with caution, given that the $z$-skewness and $z$-kurtosis values were acceptable, as was the histogram.

![Histogram of Social Relating Scores](image)

*Figure 27. Histogram of Social Relating Scores*
3.5 Preliminary Analysis

Initial bivariate correlation analyses were then conducted to assess whether or not the proposed predictor variables (Hyperphagic Drive, Hyperphagic Behaviour; Age; Gender; Weight Category) were associated with the five behavioural subscales of the DBC. The variable IQ was excluded from this analysis, as not enough data was gathered for it to be used in a correlational analysis.

3.5.1 Gender

A number of point-biserial correlations (Coolican, 2004 p.470) were conducted to see if there was a relationship between gender and the five behavioural subscales of the DBC.

This revealed that gender was not significantly related to any of the five subscales. Therefore, this variable was not included in any of the multiple regression analyses.

3.5.2 Weight Category

A number of biserial correlations were conducted to examine the relationship between weight category and the five behavioural subscales of the DBC. The following formula from Field (2005, p.132) was used to calculate these correlation coefficients:

\[ r_b = r_{pb} \sqrt{\frac{P_1P_2}{y}} \]

In this context, \( P_1 \) was the percentage in the healthy weight group and \( P_2 \) was the percentage of people in the overweight/obese group.

This analysis revealed that weight was not significantly related to the Self-Absorbed behaviour, Anxiety or Communication disturbance subscales. Therefore, this variable was excluded from these regression analyses.

However, weight category was significantly related to the Disruptive/Antisocial Behaviour Subscale, \( r_b \text{ (df; 63) } = .304, p=.016 \). Therefore, this variable was included into the analysis as a predictor variable. Furthermore, weight category was significantly related to the Disruptive/Antisocial Behaviour Subscale, \( r_b \text{ (df; 63) } = \)
.299, p=.017. Therefore, this variable was included into the analysis as a predictor variable.

3.5.3. Age
A number of Pearson’s correlations were conducted to examine the relationship between age and the five behavioural subscales of the DBC. This analysis revealed that weight was not significantly related to the Self-Absorbed behaviour, Anxiety, Social Relating or Communication disturbance subscales. Therefore, this variable was excluded from these regression analyses.

However, age was significantly related to the Disruptive/Antisocial Behaviour Subscale, \( r(\text{df}; 83) = .360, p=.001 \). Therefore, this variable was included into the analysis as a predictor variable.

3.5.4. Hyperphagic Drive
A number of Pearson’s correlations were conducted to examine the relationship between total Hyperphagic Drive scores (from the HQ) and the five behavioural subscales of the DBC.

This revealed significant positive correlations between hyperphagic drive and all five of the subscales of the DBC. Therefore, hyperphagic drive was included as a predictor variable in all regression analyses.

3.5.5. Hyperphagic Behaviour
A number of Pearson’s correlations were conducted to examine the relationship between total Hyperphagic Behaviour scores (from the HQ) and the five behavioural subscales of the DBC.

This analysis revealed that Hyperphagic Behaviour was not significantly related to the Anxiety or Communication disturbance subscales. Therefore, this variable was excluded from these particular regression analyses.

However, Hyperphagic Behaviour was significantly related to Antisocial/Disruptive behaviour \( r(76) = .476, p<.001 \). Therefore, this variable was selected for entry into
the regression analysis as a predictor variable. Hyperphagic Behaviour was significantly related to Self-Absorbed behaviour $r (76) = .422, p < .001$. Therefore, this variable was selected for entry into the regression analysis as a predictor variable. Finally, Hyperphagic Behaviour was significantly related to the Social Relating subscale $r (76) = .299, p = .009$. Therefore, this variable was selected for entry into the regression analysis as a predictor variable.
3.6 Assessing the assumptions of the regression models:
Following the regression analyses, the assumptions of the model(s) were confirmed through the following steps:

3.6.1 Multiple regression of Antisocial/Disruptive Behaviour Subscale

3.6.1.1 Normally distributed errors
The assumption of normally distributed errors was confirmed by examining the standardised residuals. Field (2005, p.170) states that 95% of these should be between -2 and 2, and none should be over 3. This regression revealed no cases over 2. Furthermore a Histogram (Figure 28) and Normality Plot of the Residuals (Figure 29) both appeared normally distributed. Therefore, one can assume that the assumption of normally distributed errors in the model is acceptable.

![Histogram](image)

*Figure 28. Histogram of the standardised residuals*
3.6.1.2. Linearity & Homoscedasticity

In order to assess the assumption of linearity and homoscedasticity, a scatterplot of standardised predicted values with standardised residuals was plotted (see Figure 30). The data points on this graph were evenly dispersed around zero and there was no identifiable curve, this indicated that both assumptions were met.

Figure 29. Normal plot of the standardised residuals

Figure 30. Scatterplot of the regression standardised residuals with the Standardised Predicted Values
3.6.1.3. **Co-linearity**

The assumption of no multi-collinearity was initially assessed by examining the correlations between the predictor variables. There were no correlations above .8 between predictors, therefore multi-collinearity was not a problem. The VIF and Tolerance statistics were also consulted to assess for multi-collinearity (see Table 7). According to Bowerman and O'Connell (1990), the largest VIF should not be over 10 for the results to be acceptable. In this case, VIF values were between 1.195 and 2.217, which do not indicate any great cause for concern. Furthermore, according to Menard (1995) tolerance values below .2 indicate a problem with multi-collinearity. All values were above this, therefore these results were also acceptable.

| Co-linearity statistics for the multiple regression of the subscales of the HQ |
|-------------------------------|-------------------|
|                                | Tolerance | VIF    |
| Age of child                  | .837      | 1.195  |
| Weight Category               | .897      | 1.115  |
| Hyperphagic Drive             | .518      | 1.931  |
| Hyperphagic Behaviour         | .451      | 2.217  |

3.6.1.4. **Casewise Diagnostics**

Cases that may be affecting the regression were assessed using Mahalanobis distance, Cooks distance and the Leverage values. No problematic cases were identified and none of the Mahalanobis distances were over 15, which supports that they are acceptable (Field, 2005). The Cook’s distances also were all under 1, which is also within the acceptable level. Finally, the average leverage was calculated at 0.085, this was multiplied by two to give a value of 0.254; none of the leverage values were above this point. Therefore, it appeared that no cases were impacting on the regression analysis.
3.6.1.6. Summary
On the whole, the assumptions of multiple linear regression seem to have been met in this data; therefore the model appears to be accurate for this sample.

3.6.2 Multiple regression of Self-Absorbed Subscale
3.6.2.1. Normally distributed errors
The assumption of normally distributed errors was confirmed by examining the standardised residuals. Field (2005, p.170) states that 95% of these should be between -2 and 2, and none should be over 3. This regression revealed only three cases over 2 and none over 3. Furthermore a Histogram (Figure 31) and Normality Plot of the Residuals (Figure 32) both appeared normally distributed. Therefore, one can assume that the assumption of normally distributed errors in the model is acceptable.

Figure 31. Histogram of the standardised residuals
3.6.2.2. Linearity & Homoscedasticity

In order to assess the assumption of linearity and homoscedasticity, a scatterplot of standardised predicted values with standardised residuals was plotted (see Figure 33). The data points on this graph were evenly dispersed around zero and there was no identifiable curve, this indicated that both assumptions were met.

Figure 32. Normal plot of the standardised residuals

Figure 33. Scatterplot of the regression standardised residuals with the Standardised Predicted Values
3.6.2.3. Co-linearity

The assumption of no multi-collinearity was initially assessed by examining the correlations between the predictor variables. There were no correlations above .8 between predictors; therefore multi-collinearity was not a problem. The VIF and Tolerance statistics were also consulted to assess for multi-collinearity (see Table 8). According to Bowerman and O’Connell (1990), the largest VIF should not be over 10 for the results to be acceptable. In this case, VIF values were 1.903, which do not indicate any great cause for concern. Furthermore, according to Menard (1995) tolerance values below .2 indicate a problem with multi-collinearity. All values were above this, therefore these results were also acceptable.

Table 8

<table>
<thead>
<tr>
<th>Co-linearity statistics for the multiple regression of the subscales of the HQ</th>
<th>Tolerance</th>
<th>VIF</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hyperphagic Drive</td>
<td>.525</td>
<td>1.903</td>
</tr>
<tr>
<td>Hyperphagic Behaviour</td>
<td>.525</td>
<td>1.903</td>
</tr>
</tbody>
</table>

3.6.1.4.4. Casewise Diagnostics

Cases that may be affecting the regression were assessed using Mahalanobis distance, Cooks distance and the Leverage values. Three potentially problematic cases were identified; however none of the Mahalanobis distances were over 15, which indicates that they are acceptable (Field, 2005). The Cook’s distances were all under 1, which is also within the acceptable level. Finally, the average leverage was calculated at 0.051, this was multiplied by two to give a value of 0.154; none of the leverage values were above this point. Therefore, it appeared that no cases were impacting on the regression analysis.

3.6.2.5. Summary

On the whole, the assumptions of multiple linear regression seem to have been met in this data; therefore the model appears to be accurate for this sample.
3.6.3. Multiple regression of Communication Disturbance Subscale

3.6.3.1. Normally distributed errors

The assumption of normally distributed errors was confirmed by examining the standardised residuals. Field (2005, p.170) states that 95% of these should be between -2 and 2, and none should be over 3. This regression revealed no cases over 2. Furthermore a Histogram (Figure 34) and Normality Plot of the Residuals (Figure 35) both appeared normally distributed. Therefore, one can assume that the assumption of normally distributed errors in the model is acceptable.

![Histogram](image)

*Figure 34. Histogram of the standardised residuals*
3.6.3.2. Linearity & Homoscedasticity

In order to assess the assumption of linearity and homoscedasticity, a scatterplot of standardised predicted values with standardised residuals was plotted (see Figure 36). The data points on this graph were evenly dispersed around zero and there was no identifiable curve, this indicated that both assumptions were met.
3.6.3.4. Casewise Diagnostics
Cases that may be affecting the regression were assessed using Mahalanobis distance, Cooks distance and the Leverage values. No potentially problematic cases were identified; furthermore none of the Mahalanobis distances were over 15, which indicates that they are acceptable (Field, 2005). The Cook’s distances were all under 1, which is also within the acceptable level. Finally, the average leverage was calculated at 0.02, this was multiplied by two to give a value of 0.71; none of the leverage values were above this point. Therefore, it appeared that no cases were impacting on the regression analysis.

3.6.3.6. Summary
On the whole, the assumptions of multiple linear regression seem to have been met in this data; therefore the model appears to be accurate for this sample.

3.6.4. Multiple regression of Anxiety Subscale
3.6.4.1. Normally distributed errors
The assumption of normally distributed errors was confirmed by examining the standardised residuals. Field (2005, p.170) states that 95% of these should be between -2 and 2, and none should be over 3. This regression revealed two cases over 2 and only one over 2.5, however a Histogram (Figure 37) and Normality Plot of the Residuals (Figure 38) both appeared normally distributed. Therefore, one can assume that the assumption of normally distributed errors in the model is acceptable.
3.6.4.2. Linearity & Homoscedasticity

In order to assess the assumption of linearity and homoscedasticity, a scatterplot of standardised predicted values with standardised residuals was plotted (see Figure 39). The data points on this graph were evenly dispersed.
around zero and there was no identifiable curve, this indicated that both assumptions were met.

### Figure 39. Scatterplot of the regression standardised residuals with the Standardised Predicted Values

![Scatterplot]

3.6.4.3. **Casewise Diagnostics**

Cases that may be affecting the regression were assessed using Mahalanobis distance, Cooks distance and the Leverage values. Three potentially problematic cases were identified; however none of the Mahalanobis distances were over 15, which indicates that they are acceptable (Field, 2005). The Cook’s distances were all under 1, which is also within the acceptable level. Finally, the average leverage was calculated at 0.02, this was multiplied by two to give a value of 0.71; none of the leverage values were above this point. Therefore, it appeared that no cases were impacting on the regression analysis.

3.6.4.5. **Summary**

On the whole, the assumptions of linear regression seem to have been met in this data; therefore the model appears to be accurate for this sample.
3.6.5. Multiple regression of Social Relating Subscale

3.6.5.1. Normally distributed errors

The assumption of normally distributed errors was confirmed by examining the standardised residuals. Field (2005, p.170) states that 95% of these should be between -2 and 2, and none should be over 3. This regression revealed only two cases over 2, furthermore a Histogram (Figure 40) and Normality Plot of the Residuals (Figure 41) both appeared normally distributed. Therefore, one can assume that the assumption of normally distributed errors in the model is acceptable.

Figure 40. Histogram of the standardised residuals
Figure 41. Normal plot of the standardised residuals

3.6.5.2. Linearity & Homoscedasticity
In order to assess the assumption of linearity and homoscedasticity, a scatterplot of standardised predicted values with standardised residuals was plotted (see Figure 42). The data points on this graph were evenly dispersed around zero and there was no identifiable curve, this indicated that both assumptions were met.

Figure 42. Scatterplot of the regression standardised residuals with the Standardised Predicted Values
3.6.5.3. Co-linearity

The assumption of no multi-collinearity was initially assessed by examining the correlations between the predictor variables. There were no correlations above .8 between predictors; therefore multi-collinearity was not a problem. The VIF and Tolerance statistics were also consulted to assess for multi-collinearity (see Table 8). According to Bowerman and O’Connell (1990), the largest VIF should not be over 10 for the results to be acceptable. In this case, VIF values were between 1.115 and 1.957, which do not indicate any great cause for concern. Furthermore, according to Menard (1995) tolerance values below .2 indicate a problem with multi-collinearity. All values were above this, therefore these results were also acceptable.

Table 9

<table>
<thead>
<tr>
<th>Co-linearity statistics for the multiple regression of the subscales of the HQ</th>
<th>Tolerance</th>
<th>VIF</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hyperphagic Drive</td>
<td>.520</td>
<td>1.921</td>
</tr>
<tr>
<td>Hyperphagic Behaviour</td>
<td>.511</td>
<td>1.957</td>
</tr>
<tr>
<td>Weight Category</td>
<td>.897</td>
<td>1.115</td>
</tr>
</tbody>
</table>

3.6.5.4. Casewise Diagnostics

Cases that may be affecting the regression were assessed using Mahalanobis distance, Cooks distance and the Leverage values. Two potentially problematic cases were identified; however none of the Mahalanobis distances were over 15, which indicates that they are acceptable (Field, 2005). The Cook’s distances were all under 1, which is also within the acceptable level. Finally, the average leverage was calculated at 0.069, this was multiplied by three to give a value of 0.206; none of the leverage values were above this point. Therefore, it appeared that no cases were impacting on the regression analysis.

3.6.5.5. Summary

On the whole, the assumptions of multiple linear regression seem to have been met in this data; therefore the model appears to be accurate for this sample.

3.7 Additional Analyses
3.7.1. Multiple Regression of the HQ
Multiple regression analysis was conducted to explore the association between the three original subscales of the HQ (Hyperphagic Drive, Hyperphagic Severity and Hyperphagic Behaviour) and total problem behaviour score in children with PWS.

3.7.1.1. Screening
Boxplots plotted for the three subscales of the HQ (Figures 43-45) revealed no significant outliers in the data.

![Boxplot of Hyperphagic Behaviour Subscale Scores](image1)

*Figure 43 Boxplot of Hyperphagic Behaviour Subscale Scores*

![Boxplot of Hyperphagic Drive Subscale Scores](image2)

*Figure 44. Boxplot of Hyperphagic Drive Subscale Scores*
However, initial screening revealed that the three subscales of the HQ were not normally distributed (See Table 10).

| Normality Test Results for All Three Hyperphagia Subscales |
|---------------------------------|---------|-------|-------|------|
|                                 | zSkewness | zKurtosis | K-S   | p    |
| Hyperphagic Behaviour Subscale  | 4.035     | 0.086    | .218  | <.001|
| Hyperphagic Drive Subscale      | 0.85      | 1.212    | .100  | .034 |
| Hyperphagic Severity Subscale   | 2.542     | -0.895   | .160  | <.001|

For Hyperphagic Behaviour, the zSkewness figure was calculated at 4.34 (which is significantly over the recommended cut-off of 1.96) and a Kolmogorov-Smirnov test was significant (K-S 0.218, p< .001).

For Hyperphagic Drive, the zSkewness and zKurtosis values both fell within the acceptable range (-1.96 to 1.96). Although, a Kolmogorov-Smirnov test indicated that data were not normally distributed (K-S 0.100, p=.034).
For Hyperphagic Severity, the $z$-Skewness figure was calculated at 2.54 (which is over the recommended cut-off of 1.96) and a Kolmorgorov-Smirnov test was significant (K-S 0.160, $p< .001$).

However the outcome variable; total problem behaviour score was normally distributed. Non-normal distributions in clinical data such as this are actually very common. In fact, Micceri (1989) investigated the normality of 440 distributions from previous research and found that none of the distributions examined passed all tests of normality. Many researchers endorse the use of statistical transformations which may be applied to the dataset to normalise the distribution (Bland & Altman, 1996; Field, 2005; Tabachnick & Fiddell, 2007).

However, Osborne (2002) states that although transformations may normalise data, they are not always advisable on clinical data as you may alter the fundamental nature of the variable (e.g. by creating a curvilinear relationship or changing the measurement scale). Therefore, transformations were not applied to these variables.

However, Micceri (1989) pointed out that most parametric statistics should be fairly robust to mild to moderate skew in the data and that some skew in the data should not prevent researchers from utilising parametric tests, as long as they are aware of the potential limitations. Furthermore, for multiple regression, Field (2005, p. 170) states that multiple regression requires only the outcome variable to have a normal distribution, not the predictor variables. Therefore, multiple regression was still utilised for this analysis.

3.7.1.2. Preliminary analysis
As part of the preliminary analysis for the multiple regression, bivariate Pearson’s correlational analyses were conducted to assess the relationship between the three predictor variables and Total problem behaviour score (TBPS). This was to assess whether an investigation of the relationship between the variables would be relevant using multiple regression analysis and to check for multi-collinearity. The results are displayed in Table 11.
Table 11

*Pearson’s Product correlations between Hyperphagic behaviour, drive and severity with Total Behaviour Problem Score*

<table>
<thead>
<tr>
<th></th>
<th>Hyperphagic Drive Subscale</th>
<th>Hyperphagic Severity Subscale</th>
<th>TBPS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hyperphagic Behaviour Subscale</td>
<td>.693”</td>
<td>.672”</td>
<td>.473”</td>
</tr>
<tr>
<td>Hyperphagic Drive Subscale</td>
<td>.757”</td>
<td></td>
<td>.596”</td>
</tr>
<tr>
<td>Hyperphagic Severity Subscale</td>
<td></td>
<td></td>
<td>.519”</td>
</tr>
</tbody>
</table>

= Significant at p<.001 level (two-tailed)

These initial correlational analyses revealed significant positive correlations between Total Problem Behaviour Score and Hyperphagic Drive, Hyperphagic Severity and Hyperphagic Behaviour. Furthermore, the correlations between the subscales themselves revealed correlation coefficients between the subscales of between r = .672 and r = .757. As none of these coefficients are above r = .8 (Field, 2005), these figures indicate that there was not a significant problem with multi-collinearity.

3.7.1.3. Multiple Regression

Multiple regression analysis using the forced entry method revealed a good fit (see Table 12) with the three hyperphagic subscales explaining \( R^2 = .37 \) 36.9% of the variance in total problem behaviour score. Furthermore, the Analysis of Variance (ANOVA) revealed that the overall model was significant (\( R^2 = .607 \) \( F(3,79) = 15.38, p < 0.01 \)).

Table 12

*Multiple regression of Hyperphagic behaviour, drive and severity with Total Behaviour Problem Score.*

<table>
<thead>
<tr>
<th></th>
<th>B</th>
<th>SE B</th>
<th>( \beta )</th>
<th>t</th>
<th>( p ) (two-tailed)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Constant</td>
<td>10.38</td>
<td>6.39</td>
<td></td>
<td>1.62</td>
<td>.11</td>
</tr>
<tr>
<td>Hyperphagic Behaviour Subscale</td>
<td>.37</td>
<td>.61</td>
<td>.08</td>
<td>.60</td>
<td>.55</td>
</tr>
<tr>
<td>Hyperphagic Drive Subscale</td>
<td>2.68</td>
<td>.90</td>
<td>.44</td>
<td>2.98</td>
<td>.004</td>
</tr>
<tr>
<td>Hyperphagic Severity Subscale</td>
<td>1.59</td>
<td>1.72</td>
<td>.13</td>
<td>.92</td>
<td>.36</td>
</tr>
</tbody>
</table>

Note, \( R^2 = .37 \), Adjusted \( R^2 = .35 \)
However, the Pearson’s correlations revealed that only Hyperphagic Drive significantly predicted Total Problem Behaviour in children with PWS.

3.6.1.4 Assessing the Assumptions of the model

Following the linear regression, the assumptions of the model were confirmed through the following steps:

3.6.1.4.1. Normally distributed errors

The assumption of normally distributed errors was confirmed by examining the standardised residuals. Field (2005, p.170) states that 95% of these should be between -2 and 2, and none should be over 3. This regression revealed only two cases over 2 and one over 2.5. Furthermore a Histogram (Figure 46) and Normality Plot of the Residuals (Figure 47) both appeared normally distributed. Therefore, one can assume that the assumption of normally distributed errors in the model is acceptable.

Figure 46. Histogram of the standardised residuals
3.6.1.4.2. Linearity & Homoscedasticity

In order to assess the assumption of linearity and homoscedasticity, a scatterplot of standardised predicted values with standardised residuals was plotted (see Figure 48). The data points on this graph were evenly dispersed around zero and there was no identifiable curve, this indicated that both assumptions were met.
3.7.1.4.3. Co-linearity

The assumption of no multi-collinearity was initially assessed by examining the correlations between the predictor variables. The VIF and Tolerance statistics were also consulted to assess for multi-collinearity (see Table 13). According to Bowerman and O’Connell (1990), the largest VIF should not be over 10 for the results to be acceptable. In this case, VIF values were between 2.132 and 2.741, which do not indicate any great cause for concern. Furthermore, according to Menard (1995) tolerance values below .2 indicate a problem with multi-collinearity. All values were above this; therefore these results were also acceptable.

Table 13

Co-linearity statistics for the multiple regression of the subscales of the HQ

<table>
<thead>
<tr>
<th>Subscale</th>
<th>Tolerance</th>
<th>VIF</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hyperphagic Behaviour Subscale</td>
<td>.469</td>
<td>2.132</td>
</tr>
<tr>
<td>Hyperphagic Drive Subscale</td>
<td>.365</td>
<td>2.741</td>
</tr>
<tr>
<td>Hyperphagic Severity Subscale</td>
<td>.385</td>
<td>2.596</td>
</tr>
</tbody>
</table>

3.7.1.4.4. Casewise Diagnostics

Cases that may be affecting the regression were assessed using Mahalanobis distance, Cook’s distance and the Leverage values. Three potentially problematic cases were identified; however none of the Mahalanobis distances were over 15, which indicates that they are acceptable (Field, 2005). The Cook’s distances were all under 1, which is also within the acceptable level. Finally, the average leverage was calculated at 0.036, this was multiplied by three to give a value of 0.108; none of the leverage values were above this point. Therefore, it appeared that no cases were impacting on the regression analysis.

3.7.1.4.5. Summary

On the whole, the assumptions of multiple linear regression seem to have been met in this data; therefore the model appears to be accurate for this sample and generalisable to the population.
3.7.2. Comparison of DBC Behaviour Scores with group Norms

The data collected from this study was analysed using Graphpad™ Prism Software for Windows (version 5.0, 2009).

An independent samples t-test with was conducted to compare the Total Problem Behaviour Scores (TPBS) obtained by the PWS sample, with the Norm Group from the DBC (Einfeld & Tonge, 2002). The normative data consisted of only summary measures (e.g. Standard Deviation and sample size) therefore equality of variances could not be assumed between the two groups. In order to control for any differences in terms of variances, a Welch’s correction was therefore utilised for the analysis, as this would apply a more stringent $p$ value. The results of the comparison are demonstrated in Table 14.

<table>
<thead>
<tr>
<th>TBPS</th>
<th>Norm Group (N: 511)</th>
<th>PWS Group (N: 83)</th>
<th>Welch's $t$</th>
<th>$p$ (2-tailed)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean</td>
<td>42.8</td>
<td>50.36</td>
<td>2.494</td>
<td>0.0142</td>
</tr>
<tr>
<td>Std. Dev</td>
<td>24.0</td>
<td>25.87</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Std. Error of Mean</td>
<td>1.06</td>
<td>2.84</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

There was a significant difference in Total Problem Behaviour Scores for the PWS group ($M = 50.36, SD = 25.87$) and the DBC Norm group ($M = 42.8, SD = 24.0$); $t(106) = 2.49, p = 0.0142$ (two-tailed). However, the magnitude of the differences in the means (mean difference = 7.560, 95% CI: 1.550 to 13.570) was small according to Cohen (1988, pp. 284-7) at: $\eta^2$ squared = .01.
3.7.3. Examining Behaviour by Gender

Analysis was conducted to explore whether or not males and females obtained different scores on any of the behavioural or emotional subscales of the DBC. As the data was not normally distributed, non-parametric Mann-Whitney U Tests were conducted to compare the difference in DBC-P subtest scores between males and females. Furthermore, as multiple comparisons were completed, a Bonferroni Correction was applied to reduce the chance of a Type-1 error. Therefore, significance was set at:

\[
\text{Alpha} = \frac{0.05}{5} = 0.01
\]

The median scores on the DBC-P subscales by gender are presented in Table 10, as are the results of the Mann-Whitney U Tests. No statistically significant differences were found between males and females on disruptive/antisocial behaviour, self-absorbed behaviour, communication disturbances, anxiety or social-relating.

Table 15

<table>
<thead>
<tr>
<th>The median scores on the DBC-P subscales by gender</th>
<th>Male (N: 43)</th>
<th>Female (N: 30)</th>
<th>Mann-Whitney U</th>
<th>( p ) (2-tailed)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Disruptive/Antisocial Subscale</td>
<td>15.00</td>
<td>15.00</td>
<td>6.37.50</td>
<td>.933</td>
</tr>
<tr>
<td>Self-Absorbed Subscale</td>
<td>14.00</td>
<td>9.50</td>
<td>510.50</td>
<td>.131</td>
</tr>
<tr>
<td>Communication Disturbance Subscale</td>
<td>9.00</td>
<td>8.50</td>
<td>567.50</td>
<td>.384</td>
</tr>
<tr>
<td>Anxiety Subscale</td>
<td>5.00</td>
<td>4.50</td>
<td>602.00</td>
<td>.627</td>
</tr>
<tr>
<td>Social Relating Subscale</td>
<td>5.00</td>
<td>4.50</td>
<td>643.50</td>
<td>.987</td>
</tr>
</tbody>
</table>
3.7.4. Assessing the Reliability of the Hyperphagia Questionnaire

Reliability analysis was conducted on each of the subscales of the HQ, the results were as follows:

3.7.4.1. Hyperphagic Behaviour

Corrected item-total correlation scores were calculated for this scale to assess how well each item correlated with the total subscale score. For each of the five items in this scale, correlation coefficients varied from .59 to .84. These values are above the .3 cut-off point recommended by Pallant (2007), which indicates that each item correlates well with the total Hyperphagic behaviour score.

This subscale demonstrated good levels of internal consistency; the Cronbach’s alpha coefficient was calculated at .87, which is above the recommended cut off of .7 (Devillis, 2003). Furthermore the mean inter-item correlation coefficient was .58, with values ranging from .41 to .82. This suggests a strong relationship among items within the subscale.

3.7.4.2. Hyperphagic Drive

Corrected item-total correlation scores were calculated for this scale to assess how well each item correlated with the total subscale score. For each of the four items in this scale, correlation coefficients varied from .62 to .84. These values are above the .3 cut-off point recommended by Pallant (2007), which indicates that each item correlates well with the total Hyperphagic Drive score.

This subscale demonstrated good levels of internal consistency; the Cronbach’s alpha coefficient was calculated at .91, which is above the recommended cut off of .7 (Devillis, 2003). Furthermore the mean inter-item correlation coefficient was .72, with values ranging from .62 to .84. This suggests a strong relationship between the items within the subscale.

3.7.4.3. Hyperphagic Severity

Corrected item-total correlation scores were calculated for this scale to assess how well each item correlated with the total subscale score. For the two items in this scale, the correlation coefficient was .63. This value is above the .3 cut-off point
recommended by Pallant (2007), which indicates that both items correlate well with the total Hyperphagic Drive score, and suggests a strong relationship between the two items in the subscale.

This subscale demonstrated good levels of internal consistency; the Cronbach’s alpha coefficient was calculated at .75, which is well above the recommended cut off of .7 (Devillis, 2003).
Section Four: Extended Discussion

4.1 Discussion of additional analyses

4.1.1. Multiple Regression of the original HQ subscales

Preliminary analyses revealed that all original subscales of the HQ had significant positive correlations with the Total Behaviour Problem Score. However, once entered into the multiple regression analysis, it emerged that Hyperphagic behaviour and Hyperphagic severity were not statistically significant predictors of behavioural and emotional problems in the sample. In fact, the only statistically significant predictor was Hyperphagic Drive. These results indicate that the higher the level of hyperphagic drive, the higher the level of emotional and behavioural problems. Therefore, the children who become most distressed when denied food and/or those who are not easily re-directed from food-related activities or discussions are also those who display the higher levels of emotional and behavioural problems. These findings are in contrast to Dykens et al (2007) who found that both hyperphagic drive and hyperphagic behaviour were related to non-food related behavioural problems.

No causal direction should be inferred from the current findings, as this study did not demonstrate that a change in hyperphagic drive directly causes significant changes in behavioural and emotional problems. However, one explanation for this finding could be that individuals with higher levels of hyperphagic drive are more easily distressed generally, therefore they also display more emotional and behavioural problems. There is no evidence to support this assumption, as this possibility has not been researched to date.

However, there could be an alternative explanation for this finding. Parents of children with PWS are increasingly being encouraged to manage their child’s hyperphagia through strict management of the environment in which access to food is completely eliminated (Whitman & Jackson, 2006). As technology advances, the techniques for doing this have progressed considerably (for example; electronic tagging, kitchen alarms and locks are all now frequently utilised in family homes to prevent individuals from over-eating). Hyperphagic behaviour scores could be influenced by the level of this external control. For example; attempts to steal or
forage for food could be prevented by the use of door alarms and sensors. Therefore, scores on this subscale may be artificially deflated by the level of external control by caregivers.

Similarly, the hyperphagic severity subscale may have also been confounded by the fact that highly structured daily routines are also recommended to manage hyperphagia (Whitman & Jackson, 2006). Depending on the how rigorous these routines are, they should be structured to allow less time for a child to engage in food related behaviours and provide less opportunities for hyperphagia to interfere with their daily routines (Whitman & Jackson, 2006). This could also reduce the score they then obtained on the Hyperphagic severity subscale.

Therefore, both the hyperphagic behaviour and hyperphagic severity items could be controlled by external sources, such as parents and teachers. On the other hand, Hyperphagic drive is a construct which cannot so easily be influenced by such controls, as it involves the child’s emotional reaction to being denied food and also the amount of time that child talks about and/or thinks about food. Therefore, this subscale may reflect the child’s level of hyperphagia when external controls are removed. However there is no research evidence to support this hypothesis to date. One way to explore this in future research would be to ask parents to complete the hyperphagia questionnaire as if no external controls were in place (e.g. how would your child respond if left alone?) and to then ask parents to complete the same questionnaire with the current controls in place to see how much scores on hyperphagic severity and behaviour subscales are influenced by this. Alternatively, parents could simply be asked to provide information on the way that they are managing their child’s hyperphagia when completing the measure and this could then be controlled for in any further analysis.

4.1.2. Comparison of DBC Behaviour Scores with group norms

The scores from this study were compared with the scores from the DBC-P normative group (Einfeld & Tonge, 1996a, 1996b, 2002). This group was made up of 511 children aged from 4-18 years with intellectual disabilities of mixed aetiologies residing in both rural and urban areas of Australia. The results of this analysis showed that the PWS group had statistically significantly higher levels of behavioural
and emotional problems than the normative group. This confirms previous research into behaviour and PWS, which has found higher levels of behavioural and emotional problems in children with PWS when compared to children with intellectual disabilities of other aetiologies (Clarke et al., 1998; Einfeld et al. 1999; Reddy, Steven, & Pfeiffer, 2007). This provides support for the suggestion that PWS has a specific pattern of behaviour (or behavioural phenotype) which is different to the expected patterns of behaviour in the wider learning disability population (Einfeld et al. 1999).

However one limitation of this analysis is that the normative group from the DBC-P was made up of a mixture of children with mild, moderate and severe intellectual disabilities. As no data was collected on the child’s level of intellectual disabilities in the current study, we were not able to accurately compare these results by level of intellectual disability. Previous research has suggested that individuals with PWS generally experience only mild to moderate intellectual disabilities (Whittington et al. 2004). Furthermore, past research has found a relationship between behaviour and intellectual abilities in PWS (Einfeld et al. 1999) with those with more severe intellectual disabilities displaying more severe behavioural problems. Therefore, these results should be viewed with caution.

In addition to this, one further limitation of this study was that the children for the normative sample were all recruited within Australia. No normative data for the DBC-P from a UK population are available.

4.1.3. Examining Behaviour by Gender

The scores on the behavioural subscales of the DBC-P were compared between male and female participants, to see if there was any significant difference in the types of behavioural and emotional problems between girls and boys. No significant differences were found on any of the behavioural subscales between males and females. This finding contradicts previous research, which has shown that boys with PWS are more likely to display externalising and aggressive behaviour than girls, and that girls are more likely to display anxiety and self-harm behaviour (Dykens & Cassidy, 1995; Dykens, 2004). This finding also contradicts research which has looked at gender differences of behaviour with children with intellectual disabilities of
other aetiologies and has shown that boys display significantly more behavioural and emotional problems overall, with particularly high scores on disruptive and self-absorbed behaviour (Einfeld & Tongue, 2002). On the other hand, this finding is consistent with other studies with individuals with PWS, which have found no significant differences in the types of behavioural, and emotional problems exhibited males and females (Dykens, et al. 1991; Einfeld et al. 1999; Steinhausen et al. 2004). Therefore, the evidence appears inconclusive at this time and further research would be warranted in this area.
4.2 Strengths and Limitations

Overall, the current study was able to add to the knowledge and understanding of part of a syndrome, which has been neglected in research. However, there are a number of methodological limitations of this study. Firstly, the response rate to the questionnaires was significantly poorer than anticipated. Other research into PWS, using the same PWSA-UK data base received a 60% response rate from parents and carers of children with PWS (Russell & Oliver, 2003). This study only achieved 105 responses from 350 questionnaire packs (30% response rate). It is difficult to speculate on the reasons for the low response rate for this study when compared with Russell and Oliver (2003). It may be that, as it is more than six years since their research was conducted, that the individuals on the database have changed. Conversely, as PWS is such a rare syndrome and the PWSA-UK is the only resource in the UK, which provides access for research to such a large number of people with PWS, it may be that the families on the database have been saturated with requests for research over the years. However, in a review of response rates to postal questionnaires (Edwards et al. 2009) it was found that many factors may significantly influence response rates, such as:

- Pre-notification of the study
- Length of questionnaires
- Providing financial and non-financial incentives
- Providing a reminder letter with another set of questionnaires
- University vs. corporate identity of the researchers
- Use of stamped rather than franked reply envelopes
- First class outward mailing
- Being offered opportunity to opt out of the study
- Sensitive questions

Many of these factors were already considered in the planning of this study, for example, participants were given prior notification via the PWSA-UK website and newsletter; sensitive questions were excluded where possible; and confidentiality was maintained by not asking for any identifying personal data. Furthermore, the questionnaires sent out were of a similar length to those used by Russell and Oliver (2003). In fact, the DBC-P contains only 96 items, whilst the measure of challenging behaviour utilised by Russell and Oliver (2003) - the Child Behavior Checklist
(CBCL: Achenbach, 1991) contains 118 items. However, some of the other factors were more difficult to control for, due to the time and financial constraints of this particular study. For example, financial constraints on the project prevented the provision of financial incentives to participants, the use of stamped (rather than franked) reply envelopes, or the use of first class mail. The possibility of sending reminder letters to those who had not responded was explored to increase participant numbers. However, this was not possible, due to the fact that researchers did not have access to the names and addresses on the database or any identifying information about the participants. Therefore, it would have meant sending out another 350 letters and questionnaire packs, at a significant cost. The PWSA-UK were approached to explore the possibility of either placing a reminder notice on their website, or sending out an e-mail reminder to parents on the database. However, the association do not have access to e-mail addresses for the majority of parents and carers and they were not able to place a notice on the website within the required time scale. In addition to these factors, the rarity of PWS means that recruiting participants from other sources (such as schools or the National Health Service) would have been very difficult indeed, as many regions may only have a very small number of children with PWS within them, and those children may not be known to services. Therefore, no further participants were recruited for this study.

However, the limited response rate and subsequent response bias were taken into consideration when approaching the analysis, the interpretation of data and in the discussion of the results. Furthermore, the number of responses received was compared to the estimated population of children with PWS in the UK, to ascertain whether or not the population recruited represented a significant proportion of the PWS population. The Office of National Statistics (ONS - online, 2008) was consulted for population estimates. The ONS estimated that the population of children aged 4-18 years in the UK in 2008 was 9,689,424 people. Prader-Willi Syndrome is thought to affect between 1 in 15,000 and 1 in 30,000 population (Cassidy & Driscoll, 2009; Couper & Couper, 2000). As such, it could be estimated that between 322 and 645 children aged 4-18 years old have PWS at this time in the UK. Therefore, the recruitment of 105 participants actually represents a significant proportion of the whole population 16.28 – 32.61% of children aged 4-18 years with PWS. This potentially increases the generalisability of the findings.
4.3 Implications

Despite the limitations of the current study, the outcome has some important clinical implications. As mentioned in the research paper, the most significant clinical implication of this study is that it has added to the understanding of the complex behavioural and emotional problems in PWS. In particular it has indicated a link between the physical symptoms, emotions and behaviour in PWS. In future research, this finding could inform the development of a bio-psycho-social model of the behaviour in PWS, similar to that of Dodge and Petit (2003). As mentioned in the literature review, Woodcock et al. (2009) created a hypothetical model of PWS (See Figure 1), which include some of these elements. However, this model has many limitations, most saliently that the proposed links between cognitive, physical, and behavioural elements of the syndrome have not all been empirically tested as yet, therefore much of the model is speculative. Furthermore, Woodcock et al (2009) failed to consider the potential impact of hyperphagia on the behaviour of people with PWS.

However, despite this, the model has been useful for attempting to begin to map out a working model of PWS. Following the results of the current study, the model has been adapted to include a link between hyperphagia and behavioural problems (such as temper outbursts and repetitive questioning). As shown on Figure 49., high levels of hyperphagia may be associated with behavioural problems. However, as this study could not determine linear causality at this time, the lines are bidirectional.

A further limitation of Woodcock et al’s (2009) model is that it does not allow for the consideration of multiple explanations for the behavioural outcomes. For example; Dykens (2000) proposed that there may be multiple reasons why children with learning disabilities behave in a certain way other than physiological factors. So, when an individual with PWS asks repetitive questions, it may reflect a deficit in task-set reconfiguration arising from brain defects, however it may also indicate that they did not understand the answer, they may not be satisfied with the response or they may be frightened, and so on. Furthermore, the model also only included temper tantrums and repetitive questioning as the non-food related behaviour problems. Whilst, it is known that individuals with PWS may display a whole range of behavioural and emotional problems such as: aggressive behaviour, stubbornness,
ritualistic behaviour; self-harm, depression, anxiety, sleep abnormalities, and inflexibility (Clarke et al. 1998; Dorman, 2001; Dykens et al., 1996; Steinhausen, et al. 2004). Therefore, for future development of the understanding of PWS, this model would require expanding. Furthermore, the various elements of the model require empirical validation.

The creation of a model of behavioural and emotional problems in PWS would be useful, to guide the development of psychological interventions for this complex group of individuals. As mentioned in the paper, currently no holistic treatments exist designed specifically for people with PWS and current interventions are limited and expensive and do not provide individuals or their families with a sense of control over the syndrome (Singh et al. 2008; Whitman & Jackson, 2006). In people with learning disabilities of other aetiologies, holistic interventions, which incorporate biological, psychological and social factors, are recommended for behavioural and emotional problems, as the most effective treatment (Xeniditis, Russell & Murphy, 2001). The observation that hyperphagia is related to the behavioural and emotional problems indicates that the two are interconnected. This provides support for the development of similar treatments, which consider the hyperphagia as well as the behavioural problems.
Figure 49: Revised version of Woodcock et al.'s (2009) model of PWS (Additions shown in dotted lines)
4.4 Future Directions
In terms of research, a number of different areas possible areas of exploration have been indicated by this study.

Firstly, it would be interesting to extend this investigation to examine hyperphagia and non-food related behaviour with different groups of people with PWS. For example, adolescents and young adults with PWS are thought to have the highest levels of behavioural problems for people with the syndrome (Steinhausen et al. 2004). Therefore, it would be interesting to investigate whether or not they also have higher levels of hyperphagia, or whether other social or personal factors related to adolescence and adulthood may account for this increase. Furthermore, a comparison with different groups who are known to also experience hyperphagia and behavioural problems would be very interesting, such as those with ‘Hyperphagia Short Stature Syndrome’ (Gilmour, Skuse & Pembrey, 2001). This would give an idea of whether or not hyperphagia is associated with behavioural and emotional problems for all groups of people, or whether the difficulties observed in this study are related to PWS specific factors.

In addition to this, during the data collection process for this study, a number of telephone calls were received from parents of children with PWS who reported that their child was not displaying any signs of hyperphagia. The age of the children discussed ranged from six to fourteen years old, which is above the usual age of onset of between 18 months and six-years (Cassidy, 1984; Clarke et al. 1996; Dimitropoulos et al. 2000). Interestingly, it emerged that all of the children discussed had diagnoses of PWS with a maternal disomy, rather than the more common paternal deletion (Dimitropoulos et al. 2000). Based on anecdotal evidence shared between other parents within the PWSA-UK, the parents and carers believed that individuals with a maternal disomy may experience significantly lower levels of hyperphagia than other individuals. However, Dykens et al (2007) found no association between hyperphagia levels and genetic status (e.g. deletion, uniparental disomy or imprinting error) in their study. Therefore, in this study, parents and carers were not asked to record their child’s exact PWS diagnosis, as it was believed that this information may not be readily available to parents and there was no evidence in the literature to suggest that it would affect behaviour or hyperphagia.
However, it would be interesting if future research into PWS could further investigate the possible differences within the genetic subgroups in terms of behaviour and hyperphagia.

Another point of interest for future research may be to explore the relationship between behavioural and emotional problems in PWS and the possible risk factors (e.g. hyperphagia) with more in depth statistical techniques. So far, only traditional ‘one-step’ methods have been used to explore the relationship between various factors and behaviour in PWS. These have been sufficient to suggest common patterns and relationships. However, techniques such as structural equation modelling (SEM) could be used to explore the direction and specific impact of these variables. Anderson and Gerbing (1998) stated that SEM can be a more comprehensive method of assessing and modifying theoretical models within psychology, leading to the development of more robust theories.

Finally, for future research into the relationship between behaviour and hyperphagia in PWS, it would be interesting to look at the impact of specific psychological interventions on both the levels of behavioural problems and levels of hyperphagia in individuals with the syndrome. To date, very few outcome studies have been published which look at the impact of psychological interventions on PWS at all. When they have, they are often based on single case examples or very small sample sizes and the outcomes are often weight or BMI (Singh et al. 2008). This is perhaps because in the past, no reliable method of assessing the impact of an intervention on hyperphagia existed (Dykens et al. 2007).
4.5 Reflective report

This study may appear to fit well within a traditional positivist (or empiricist) framework. Positivism purports that observation and measurement should form the core of scientific work and that only observable and measurable traits should be investigated through experimental (quantitative) research (Guba, 1992). However, this position has long been criticised in the field of social sciences, which maintain that human behaviour, thoughts and feelings cannot easily be reduced to numerical constructs in the same way as in other sciences (such as chemistry and biology). This is because human behaviour and emotions are not fixed, but are determined by individual, social and situational factors (Littlejohn, 2003). Therefore, in recent years, a wide range of ‘post-positivist’ epistemological approaches to social science research have emerged.

Two dominant post-positivist approaches within psychology are ‘critical realism’ and ‘constructivism’ (Ponterotto, 2005). Constructivism purports that realities are constructed in individual’s minds through language and social interaction and that the goal of research is understanding individual’s meaning through exploring lived experience (so often through qualitative research methods) (Hoshmand & Martin, 1994). On the other hand, critical realism maintains that there exists a ‘reality’ independent of our thinking about it that science can study and explore, however it acknowledges that all methods of observation are fallible and have the potential of error (Littlejohn, 2003; Okasha, 2002). Therefore critical realists believe that all theory is revisable, and that you can never truly ‘know’ psychological experiences, but we may be able to use research to understand common patterns of behaviour (Bhaskar, 1989). As a result, for critical realists both qualitative and quantitative methods are valid methods for the exploration and development of theory (Hoshmand & Martin, 1994).

In terms of epistemology, traditional positivist approaches emphasise the need for academics to maintain complete objectivity throughout research, so as not to bias results (Guba, 1992). On the other hand, the constructivist view is that the interaction between researcher and participant is fundamental for comprehending experiences and that the research process is subjective (Smith, 2004). Critical realists once again take the epistemological ‘middle ground’ (Hoshmand & Martin, 1994) in which they...
are aware that researchers may influence research or be influenced by it, but objectivity and researcher-subject independence are still important goals to aim for during the research process (Ponterotto, 2005).

The current study arose from a critical realist stance. The development of the central research question was influenced by a qualitative study exploring the lived experiences of PWS, conducted by the author (Haselip, 2006). From this study, a number of important issues arose which indicated that further research looking at the relationship between hyperphagia and behavioural and emotional problems may be useful. However, the author was mindful of the fact that qualitative studies can only reflect the experiences of a very small number of people and also that the analysis may be influenced by the author’s own interpretations of the data. Therefore, it was felt that at this point, a quantitative approach would more effectively aid the development of understanding of PWS, by providing information on a wider proportion of the population. Furthermore, by using standardised measures it was hoped that the level of subjectivity in the analysis of the data would be lessened.

As this research is directed from a critical realist perspective, the following section has been used to reflect on some of the central ethical and scientific issues pertinent to this study. In particular, the methodology used in this study has been critically considered from this epistemological stand-point, in relation to the limitations of Null Hypothesis Significance Testing (NHST) and more specifically, regression analysis. Furthermore, how this study may fit within the wider academic discourse around PWS and behavioural phenotypes was explored. Finally, some of the key ethical concerns related to this study were addressed.
4.5.1 Use of NHSTs

NHST’s have been used throughout the current study (and for the additional analyses), as they are perhaps the most commonly used and accessible form of data analysis in published psychological research literature (Balluerka, Gómez, & Hidalgo, 2005; Loftus, 1996; Nickerson, 2000). However, for many years now, the use of NHSTs has been heavily criticised (Balluerka, et al. 2005; Cohen, 1962, 1994; Cronbach, 1975; Howard, Maxwell & Fleming, 2000; Loftus, 1996; Nickerson, 2000). Time was taken to reflect on the central limitations of NHST methods and to consider the strengths and limitations of some of the main alternatives to these methods, before reaching a conclusion based on this study.

The central and predominant criticism of NHST’s is that they are widely misunderstood and misinterpreted throughout the literature (Abelson, 1997). The major reason for this is that researchers are commonly confused by what the p value actually represents. Many researchers using NHSTs believe that the p value is the probability of the null hypothesis (Ho) being true (Balluerka et al. 2005). The danger of this is that it can lead to false conclusions as in fact the p value represents the probability of observing results as extreme (or more extreme) as observed if the Null Hypothesis is false (Balluerka et al. 2005).

A further criticism of NHST’s is that the Null Hypothesis can never be true within the social sciences (Howard et al. 2000). This argument fits with the arguments of the post-positivists who emphasised that, human beings are shaped by their own unique situation or contexts. As such they will vary so widely, that there will always be differences across participants, no matter how small those differences are. Therefore, it is possible that with a large enough sample size and a two-tailed hypothesis; you could always obtain a significant result in psychological research, even when differences between groups are actually very small (Howard et al. 2000). Some researchers have stated that this therefore makes the Null Hypothesis meaningless, as it is impossible to achieve within the social sciences (Loftus, 1996).

A further limitation of NHSTs is that the p<.05 cut off point for significance is used stringently within the social sciences; however it is essentially an arbitrary point (Field, 2005). As a result of this, ‘statistically significant’ (p<.05) results have become
favoured in the literature, and there is a danger that this can lead to the publication of relatively meaningless papers with significant results, or conversely that interesting papers with p values of .051 may not make it to publication, (Loftus, 1996). A consequence of this over-reliance on the p value is that researchers may also pay little attention to the need to ensure that the underlying assumptions of the statistical method selected have been met (Loftus 1996). Abelson (1997) states that to overcome these limitations, assumptions of tests should be rigorously adhered to and effect sizes and confidence intervals should also be reported in addition to p values.

The final disadvantage of NHST’s is that they are not designed for amassing evidence over multiple studies as they only provide a researcher with a ‘yes or no’ (significant or non-significant) outcome. Howard et al (2000) stated that this is a problem as you cannot ‘scientifically’ make comments about human behaviour based on the hypothesis and statistical results of a single study. Therefore, NHSTs are not well designed to assess theories and so they cannot advance scientific knowledge (Balluerka et al. 2005)

Alternatives to traditional NHST’s have been proposed which are designed to overcome some of the weaknesses of NHSTs. For example, Bayesian analysis, meta-analysis and Planned comparisons (contrasts) are all examples of possible alternatives (Howard et al. 2000). Bayesian approaches to analysis are philosophically very different to the traditional NHST approach, as they allow the researcher to test prior beliefs about a phenomenon, which may either come from their own personal beliefs or previous empirical research (Howard et al. 2000). However, they are not always an alternative to NHST, as they are not necessarily appropriate to use when studying entirely new phenomena when a researcher cannot make prior assumptions about the results. Instead it is proposed that the philosophy of Bayesian approaches can be used in conjunction with NHST’s to ensure that researchers create strong hypotheses prior to conducting studies (Trafimow, 2003).

Another approach, Meta-analysis has been heralded as a superior alternative to NHST’s (Howard et al. 2000). This is because Meta-analysis is specifically designed
to focus on accumulating evidence from research, which can provide stronger support for a hypothesis or theory (Levin, 1998). This can also allow us to make more accurate predictions about a hypothesis in future research, something which NHST’s are unlikely to be able to ever do accurately (Howard et al. 2000).

However, like Bayesian methods, Meta-analysis is also limited by this as is not appropriate to use when a phenomena has not been explored empirically in research before. Therefore, one advantage of the NHST over meta-analysis is that it can inspire and provoke new research more than meta-analysis ever could hope to (Abelson, 1997). This is necessary for the further development of psychological theory and understanding.

So in summary, from the literature explored, it seems that NHSTs may have many pitfalls and be open to abuse by overzealous academics wishing to assert their theories. However, it also appears that if they are used properly with the aid of other complementary statistical procedures to support their evidence (such as confidence Intervals and effect sizes) they can be meaningful and can provide a good starting point for further research (Abelson, 1997; Levin, 1998). For this study in particular, the research into the link between hyperphagia and behaviour in PWS is still in its very early stages. Therefore NHST’s represented an appropriate means of opening up the dialogue about this relationship to pave way for future research. However, they have been used throughout the current study with a critical awareness of their limitations. As such, the assumptions of each of the tests were adhered to as best as possible and complimentary methods of supporting the results (e.g. effect sizes) were also adopted where possible. It is hoped that in doing this, the chance of error in the analysis has been limited.

4.5.2 Issues around the use of Regression Analysis

In addition to thinking about the limitations of NHSTs generally, Linear Regression Analysis was also considered. Regression analyses are extremely popular in psychological research however, like all other NHST’s they are not without their theoretical limitations (Fox, 1997).
The salient limitation of Linear Regression is that it can only provide information on the extent of a relationship between variables, not direction or causality (Cohen, 2003). Therefore, an apparent significant relationship may actually originate from a wide range of different sources, including variables not included in the research question (Tabachnick & Fiddell, 2007). Cohen (2003, page 7) gives the example that a researcher may find a significant relationship between an individual’s attitude to abortion and favoured political party. However, from this it would be wrong to infer that political views predict attitude to abortion or vice versa. In this example Cohen (2003) provides examples of other factors such as socio-demographic factors and religious views (to name but a few), which may also play a part in attitude to abortion.

In fact, the only way to be able to speculate on causality, is to conduct experimental research in which variables are directly manipulated by the researcher and the affect another – in this case this would be by performing an Analysis of Variance (Brace, Kemp & Snelgar, 2006). However, ‘human behaviour can be variable and difficult to manipulate, therefore it is actually a strength of regression analysis is that can measure ‘natural’ human behaviour (Brace et al. 2006).

4.5.3: How does this study fit with the wider understanding of phenotypes in PWS?

The current study has added to the knowledge of the behavioural and emotional problems in PWS. Throughout the literature on PWS, it is suggested that the presence of a plethora of specific and complex behavioural and emotional problems in PWS provides support for the existence of a distinct behavioural phenotype for the syndrome (Clarke et al. 1996; Clarke et al. 2002; Dykens & Kasari, 1997; Hiraiwi et al. 2007). These factors have been linked to the imprinting defects on chromosome 15 (Veltman et al. 2005).

However, in recent years it has been proposed that the concept of behavioural phenotypes in genetic disorders may be intrinsically and/or ethically flawed. This is due to the fact that huge variations in behaviour exist in even the simplest genetically diagnosable trait like gender, for example. In fact, gendered behaviour is said to be more on a ‘continuous pathway’ than a dichotomy (Flint, 1995). Goodey (2006) states that when typecasting individuals with learning disabilities, it is important to
bear in mind that “Most of today’s social stereotypes started out as yesterday’s known scientific facts, subsequently discarded,” (page 402). Therefore, we should not overlook other potential individual emotional, environmental and social factors contributing to challenging behaviour and mental health problems in PWS (O’Brien, 2000).

This debate actually reflects the debate between positivism (which views the body as a scientific entity) and post-positivism (that individuals are a product of their social world (Littlejohn, 2003). The critical realist perspective adopted in this study is able to incorporate the impact of both underlying physical structures and personal agency in behavioural and physical outcomes (Williams, 1999). Therefore, using this approach to consider the results, it may be proposed that the physical and social impact of PWS could account for the development of emotional and behavioural problems. However, more research is required to explore this further and to highlight the salient physical and social factors responsible for this.

4.5.4 Conducting research about children with learning disabilities.

One ethical point, which caused some concern in the current study, was the level of involvement from people with PWS themselves and the issue of informed consent. As mentioned in the paper, parents were selected to participate in the study rather than children with PWS themselves. This extended to informed consent, which was only obtained from parents, rather than the children themselves. This decision to include only parents in this study was made for a number of reasons. Firstly, the questionnaires were designed for completion by parents only, as no self-report measure of hyperphagia exists to date (Dykens et al. 2007). Secondly, previous research has indicated that individuals with PWS may find the subject of hyperphagia sensitive to discuss, and this may affect the accuracy of their reports (Dykens et al. 2007). Finally, it is known that individuals with PWS vary widely in terms of their intellectual abilities, ranging from moderate learning disabilities to average intelligence (Whittington et al. 2004), this was coupled with the fact that the sample were aged 4-18 years. Both of these factors would have meant that participant’s levels of comprehension, reading, literacy and ability to give informed consent could be extremely varied and consequently very difficult to provide for in a
postal survey (the only method of recruitment open to the researchers for the current study).

This raised some ethical concerns for the author, as increasingly, those conducting research with children and adults with learning disabilities are being expected to make provision for the inclusion and participation of the individuals themselves (Chappell, 2000; Lewis & Porter, 2004; Walmsley, 2001). Lewis and Porter (2004, page 192) state that participation should be at all stages of the research process and they provide four key ethical areas for researchers to consider before conducting any research about children with learning disabilities:

1. How will the research be useful and/or contribute to the life of people with learning disabilities?
2. Will the research bring about any change?
3. Have (or could) people with learning disabilities contribute to establishing the aims and purposes of the research?
4. Could participants be harmed in any way by participating in the study?

Very little research has been conducted in the past directly with adults or children with PWS, as most PWS research takes the form of parental report surveys (Chertkoff et al. 2002; Clarke et al. 2003; Deschemaeker et al. 2002; Dykens et al. 2007; Russell & Oliver, 2003). As a consequence, no self-report tools have been developed and this makes conducting direct quantitative research very difficult. However, as mentioned previously, the aims of the current study were actually devised from the outcome of a previous qualitative study with adults with PWS, in which the participants all emphasised the need for support with their hunger and also the negative emotional impact of feeling continually hungry (Haselip, 2006). Therefore, people with PWS did directly contribute to establishing the aims of the study. Furthermore, it was hoped that the study could contribute to the life of people with PWS, by helping people to understand their behaviour in more depth and possibly leading to more holistic treatments packages.

Despite this, it is regrettable that on this occasion, individuals with PWS were not able to contribute more fully to the research process. For further research it would be important to bear in mind the need to include people with learning disabilities in the
development, design and participation of research. For this to happen, more needs to be done to devise reliable ways of measuring hyperphagia in children with PWS, as it will be difficult to ever fully understand the experience of hyperphagia in PWS until we do that.
References

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age, and insulin concentrations in normal children and are markedly increased in Prader-Willi Syndrome. *Journal of Clinical Endocrinology and Metabolism, 88*, 174-178.


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Appendix 1: Author Guidelines for the Journal of Applied Research in Intellectual Disabilities


Journal of Applied Research in Intellectual Disabilities

Edited by:
Chris Hatton and Glynis Murphy

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ISI Journal Citation Reports® Ranking: 2009: Rehabilitation (Social Science): 27 / 52; Psychology, Educational: 20 / 44
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Top Author Guidelines

1. GENERAL

The Journal of Applied Research in Intellectual Disabilities is an international, peer-reviewed journal which draws together findings derived from original applied research in intellectual disabilities. The journal is an important forum for the dissemination of ideas to promote valued lifestyles for people with intellectual disabilities. It reports on research from the UK and overseas by authors from all relevant professional disciplines. It is aimed at an international, multi-disciplinary readership.

The topics it covers include community living, quality of life, challenging behaviour, communication, sexuality, medication, ageing, supported employment, family issues, mental health, physical health, autism, economic issues, social networks, staff stress, staff training, epidemiology and service provision. Theoretical papers are also considered provided the implications for therapeutic action or enhancing quality of life are clear. Both quantitative and qualitative methodologies are welcomed. All original and review articles continue to undergo a rigorous, peer-refereeing process.

Please read the instructions below carefully for details on submission of manuscripts, the journal's requirements and standards as well as information concerning the procedure after a manuscript has been accepted for publication. Authors are encouraged to visit http://authorservices.wiley.com/bauthor/ for further information on the preparation and submission of articles.

2. ETHICAL GUIDELINES

The Journal of Applied Research in Intellectual Disabilities adheres to the below ethical guidelines for publication and research.
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**Authorship:** Authors submitting a paper do so on the understanding that the manuscript has been read and approved by all authors and that all authors agree to the submission of the manuscript to the journal. ALL named authors must have made an active contribution to the conception and design and/or analysis and interpretation of the data and/or the drafting of the paper and ALL authors must have critically reviewed its content and have approved the final version submitted for publication. Participation solely in the acquisition of funding or the collection of data does not justify authorship.

It is a requirement that all authors have been accredited as appropriate under submission of the manuscript. Contributors who do not qualify as authors should be mentioned under Acknowledgements.

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**Conflict of Interest:** Authors are required to disclose any possible conflict of interest. These include financial (for example patent ownership, stock ownership, consultancies, speaker's fee). Author's conflict of interest (or information specifying the absence of conflict of interest) will be published under a separate heading.

The *Journal of Applied Research in Intellectual Disabilities* requires that sources of institutional, private and corporate financial support for the work within the manuscript must be fully acknowledged, and any potential conflict of interest noted. As of 1st March 2007, this information is a requirement for all manuscripts submitted to the journal and will be published in a highlighted box on the title page of the article. Please include this information under the separate headings of 'Source of Funding' and 'Conflict of Interest' at the end of the manuscript.

If the author does not include a conflict of interest statement in the manuscript, then the following statement will be included by default: 'No conflict of interest has been declared'.

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Singapore 188778
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Manuscripts should be submitted via email to patchelland@wightcabelnorth.net and copy it to both chris.hatton@lancaster.ac.uk and g.h.murphy@kent.ac.uk

3.1 Manuscript Files Accepted

Manuscripts should be uploaded as Word (.doc) or Rich Text Format (.rtf) files (not write-protected) plus separate figure files. GIF, JPEG, PICT or Bitmap files are acceptable for submission, but only high-resolution TIF or EPS files are suitable for
printing. The files will be automatically converted to HTML and PDF on upload and will be used for the review process. The text file must contain the entire manuscript including title page, abstract, text, references, tables, and figure legends, but no embedded figures. Figure tags should be included in the file. Manuscripts should be formatted as described in the Author Guidelines below.

Please note that any manuscripts uploaded as Word 2007 (.docx) will be automatically rejected. Please save any .docx files as .doc before uploading.

3.2 Blinded Review

All articles submitted to the journal are assessed by at least two anonymous reviewers with expertise in that field. The Editors reserve the right to edit any contribution to ensure that it conforms with the requirements of the journal.

4. MANUSCRIPT TYPES ACCEPTED

Original Articles, Review Articles, Brief Reports, Book Reviews and Letters to the Editor are accepted. Theoretical Papers are also considered provided the implications for therapeutic action or enhancing quality of life are clear. Both quantitative and qualitative methodologies are welcomed. Articles are accepted for publication only at the discretion of the Editor. Articles should not exceed 7000 words. Brief Reports should not normally exceed 2000 words. Submissions for the Letters to the Editor section should be no more than 750 words in length.

5. MANUSCRIPT FORMAT AND STRUCTURE

5.1 Format

Language: The language of publication is English. Authors for whom English is a second language must have their manuscript professionally edited by an English speaking person before submission to make sure the English is of high quality. It is preferred that manuscripts are professionally edited. A list of independent suppliers of editing services can be found at http://authorservices.wiley.com/bauthor/english_language.asp. All services are paid for and arranged by the author, and use of one of these services does not guarantee acceptance or preference for publication.

5.2 Structure

All manuscripts submitted to the Journal of Applied Research in Intellectual Disabilities should include:

Cover Page: A cover page should contain only the title, thereby facilitating anonymous reviewing. The authors' details should be supplied on a separate page and the author for correspondence should be identified clearly, along with full contact details, including e-mail address.

Running Title: A short title of not more than fifty characters, including spaces, should be provided.

Keywords: Up to six key words to aid indexing should also be provided.
Main Text: All papers should be divided into a structured summary (150 words) and the main text with appropriate sub headings. A structured summary should be given at the beginning of each article, incorporating the following headings: Background, Materials and Methods, Results, Conclusions. These should outline the questions investigated, the design, essential findings and main conclusions of the study. The text should proceed through sections of Abstract, Introduction, Materials and Methods, Results and Discussion, and finally Tables. Figures should be submitted as a separate file.

Style: Manuscripts should be formatted with a wide margin and double spaced. Include all parts of the text of the paper in a single file, but do not embed figures. Please note the following points which will help us to process your manuscript successfully:

- Include all figure legends, and tables with their legends if available.
- Do not use the carriage return (enter) at the end of lines within a paragraph.
- Turn the hyphenation option off.
- In the cover email, specify any special characters used to represent non-keyboard characters.
- Take care not to use l (ell) for 1 (one), O (capital o) for 0 (zero) or ß (German esszett) for (beta).
- Use a tab, not spaces, to separate data points in tables.
- If you use a table editor function, ensure that each data point is contained within a unique cell, i.e. do not use carriage returns within cells.

Spelling should conform to The Concise Oxford Dictionary of Current English and units of measurements, symbols and abbreviations with those in Units, Symbols and Abbreviations (1977) published and supplied by the Royal Society of Medicine, 1 Wimpole Street, London W1M 8AE. This specifies the use of S.I. units.

5.3 References

The reference list should be in alphabetic order thus:

Journal titles should be in full. References in text with more than two authors should be abbreviated to (Brown et al. 1977). Authors are responsible for the accuracy of their references.

We recommend the use of a tool such as EndNote or Reference Manager for reference management and formatting.

EndNote reference styles can be searched for here:
http://www.endnote.com/support/enstyles.asp
Reference Manager reference styles can be searched for here:
http://www.refman.com/support/rmstyles.asp
The Editor and Publisher recommend that citation of online published papers and other material should be done via a DOI (digital object identifier), which all reputable online published material should have - see www.doi.org/ for more information. If an author cites anything which does not have a DOI they run the risk of the cited material not being traceable.

5.4 Tables, Figures and Figure Legends

Tables should include only essential data. Each table must be typewritten on a separate sheet and should be numbered consecutively with Arabic numerals, e.g. Table 1, and given a short caption.

Figures should be referred to in the text as Figures using Arabic numbers, e.g. Fig.1, Fig.2 etc, in order of appearance. Figures should be clearly labelled with the name of the first author, and the appropriate number. Each figure should have a separate legend; these should be grouped on a separate page at the end of the manuscript. All symbols and abbreviations should be clearly explained. In the full-text online edition of the journal, figure legends may be truncated in abbreviated links to the full screen version. Therefore, the first 100 characters of any legend should inform the reader of key aspects of the figure.

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Although low quality images are adequate for review purposes, print publication requires high quality images to prevent the final product being blurred or fuzzy. Submit EPS (line art) or TIFF (halftone/photographs) files only. MS PowerPoint and Word Graphics are unsuitable for printed pictures. Do not use pixel-oriented programmes. Scans (TIFF only) should have a resolution of at least 300 dpi (halftone) or 600 to 1200 dpi (line drawings) in relation to the reproduction size. Please submit the data for figures in black and white or submit a Colour Work Agreement Form. EPS files should be saved with fonts embedded (and with a TIFF preview if possible).

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Acrobat Reader will be required in order to read this file. This software can be downloaded (free of charge) from the following website: www.adobe.com/products/acrobat/readstep2.html

This will enable the file to be opened, read on screen, and printed out in order for any corrections to be added. Further instructions will be sent with the proof. Proofs will be posted if no e-mail address is available; in your absence, please arrange for a colleague to access your e-mail to retrieve the proofs.

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REFERENCE: 001

16th January 2009

Dear Parent or Carer,

This is an invitation to participate in a piece of research being conducted by Ms Leanne Haselip (Trainee Clinical Psychologist) under the supervision of Dr Shirley Thomas at the University of Nottingham.

This study aims to explore levels of hyperphagia (excessive appetite) in children with Prader-Willi Syndrome and how this may relate to their non-food related behaviour. Participation in the study involves the completion of questionnaires as well as the provision of some background information. The information sheet enclosed provides details on why the study is being conducted, what the aims of the study are and also what the information provided in the questionnaires will be used for.

If you would like to know more, please read the enclosed information sheet for details on how to participate in this study. If you have any concerns or queries, please do not hesitate to contact the PWSA-UK on 01322 365 676, or Leanne Haselip on 0115 846 6646.

Many Thanks,

Yours Sincerely,

Jackie Waters
Dear Parent/ Carer,

My name is Leanne Haselip and I am a trainee clinical psychologist from the University of Nottingham. I would like to invite you to take part in a research study I am completing as part of my Doctorate in Clinical Psychology training course.

Before you decide whether to take part, you need to understand why the research is being carried out and what it would involve for you. Please take the time to read the following information carefully.

**What is the study about?**
It is well known that the primary symptom of Prader-Willi Syndrome (PWS) is hyperphagia (an excessive appetite). In the past, how this impacts on other areas of people’s lives has not been looked at very closely. This is because measuring hyperphagia can be really difficult.

However, a questionnaire for measuring hyperphagia has been developed. The aim of my study is to use this questionnaire to assess hyperphagia levels in children with PWS and to also look at how this might relate to their behaviour.

**Why have I been invited?**
These forms have been circulated on my behalf by the Prader-Willi Syndrome Association UK (PWSA-UK) to all families of children with PWS on their database. I have invited you to take part in this research study, in order to find out about the relationship between hyperphagia and the behaviour displayed by your child with PWS. I hope you can use this opportunity to pass on your experiences. It is hoped that if we can have a better understanding of how hyperphagia impacts on children’s’ behaviour, then this will inform how we may be able to help parents and carers to manage this.

**Do I have to take part?**
It is up to you to decide. Taking part in the research is entirely voluntary. If you are happy to provide information about your own experiences, then please complete the questionnaires attached. Deciding not to take part in the study will not affect your child’s care in anyway.

**What will I have to do?**
You will be asked to fill out all of the pink sheets of paper in the pack provided to you and return these using the pre-paid envelope supplied.

This pack comprises of two questionnaires, which should take about 20 -30 minutes to complete in total. The “Developmental Behaviour Checklist” includes questions about your child’s behaviour The “Hyperphagia Questionnaire” includes questions about your child’s eating habits. In addition to this,
you will also be asked to provide some basic information about you and your child on the sheet provided and to sign that you consent to participating in the study on the consent form in the pack.

**What will happen to the information?**
This study is completely **anonymous** – you will not be asked to provide any identifying information about yourself (such as names, addresses etc.). Instead, I ask that you make note of the **reference number** on the top of your invitation letter. This number has been randomly assigned by the researchers before the letters were sent out and we have no way of knowing which family received each number. However, if you wish to withdraw from the study at any time, you can quote that number to us and your results can be removed from the database.

Once we have received the completed questionnaires, the information will be analysed to look for common responses. The results will then be written up as my doctoral thesis for university and may also be published in a peer-reviewed journal. All study data will be stored in a secure location at the University of Nottingham and will only be accessible by myself and my research supervisor from the university.

**Who has reviewed the research study?**
All research for the university is looked at by a Research Ethics Committee to protect your safety, rights, wellbeing and dignity. This study has been reviewed and given favourable opinion by the ethics committee at the Institute of Work, Health and Organisations at the University of Nottingham.

**Will there be any follow up of the research study?**
If you would like any feedback about the research, please contact me on the details below, and I would be happy to send you a summary of the results once the study is complete. I also hope to provide a summary of the results to the PWSA-UK for their PWS News publication.

**Further information and contact details**
If you have any questions or concerns about this study, then please do not hesitate to contact me on the details below:

**Email:** lwxljh@nottingham.ac.uk  
**Tel:** 0115 846 6646

Alternatively, if you require any advice or support regarding any issues raised in this study, then please contact the Prader-Willi Syndrome Association on:

**Email:** admin@pwsa.co.uk  
**Tel:** 01332 365676

If you would like to participate in this study, please return the completed pink forms in the enclosed pre-paid envelope by **Friday 20th March 2009**, to:

Leanne Haselip (Doctorate in Clinical Psychology)  
Institute of Work, Health & Organisations  
Floor B, International House  
Jubilee Campus  
The University of Nottingham  
Nottingham. NG8 1BB

-------------------------------------------------------------------------------------------------------------------

**Thank you for reading this information**
Appendix 2.3: Participant Consent Form

REFERENCE: 001

Trent Doctorate in Clinical Psychology
Institute of Work, Health & Organisations
Floor B
International House
Jubilee Campus
The University of Nottingham
Nottingham. NG8 1BB
Tel: 0115 846 6646

If you have read the information sheet provided and would like to continue with this study. Please tick the boxes that apply to you and sign below:

- I have read the project information sheet
- I have asked any questions that I want to
- I understand that taking part in the study is my choice
- I understand that I can stop taking part in the study, if I wish
- I agree to take part in the study

____________________________  __________________________
Date  Signature

Leanne Haselip
Tel: 0115 846 6646
INFORMATION ABOUT YOU AND YOUR CHILD

On this questionnaire, we would like some basic information about you and your child with PWS. Please answer all questions to the best of your knowledge. Any questions you do not know the answer to, please leave blank and move to the next question.

REFERENCE NUMBER: ______________

1. What is your relationship to the child? (Please tick)

Mother ☐  Father ☐  Grandparent ☐
Sibling ☐  Other carer ☐

2. Does the child live with you full time? (Please tick)

Yes ☐  No ☐

If no, please give brief details below of where your child lives:
____________________________________________________________________
____________________________________________________________________

3. How old is the child?

_____ years _______ months

4. What gender is the child?

Male ☐  Female ☐

5. Age when first diagnosed with PWS

_____ years _______ months
6. What type of school does the child currently attend? (please tick)

Local Mainstream School  ☐  Local Special School  ☐
Residential School  ☐  Other (please state below)
_______________________________

7. Has the child ever received intelligence testing (IQ tests)?

Yes  ☐  No  ☐  Don’t know  ☐
If yes, what was their score?
_______________________________

8. What level of learning disability would you say the child has?

No learning disabilities  ☐
Mild learning disabilities  ☐
Moderate learning disabilities  ☐
Severe/profound learning disabilities  ☐
Don’t know  ☐

9. What is the child’s current weight and height?

Weight _________________  Height _________________
## Appendix 2.5: Hyperphagia Questionnaire

**HYPERPHAGIA QUESTIONNAIRE**  
(Dykens, Maxwell, Pantino, Kossler & Roof 2007)

This questionnaire has been designed to explore your child’s level of hyperphagia (appetite) and other food-related related behaviours in more depth. Please read each question and circle the response that best applies to your child.

<table>
<thead>
<tr>
<th></th>
<th>Question</th>
<th>Not particularly upset at all</th>
<th>A little upset</th>
<th>Somewhat upset</th>
<th>Very upset</th>
<th>Extremely upset</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>How upset does your child generally become when denied a desired food?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>How often does your child try to bargain or manipulate to get more food at meals?</td>
<td>A few times a year</td>
<td>A few times a month</td>
<td>A few times a week</td>
<td>Several times a week</td>
<td>Several times a day</td>
</tr>
<tr>
<td>3</td>
<td>Once your child has food on their mind, how easy is it for you or others to re-direct your child away from food to other things?</td>
<td>Extremely easy (takes minimal effort to do so)</td>
<td>Very easy (takes just a little effort to do so)</td>
<td>Somewhat hard (takes some effort to do so)</td>
<td>Very hard, (takes some effort to do so)</td>
<td>Extremely hard (takes sustained and hard work to do so)</td>
</tr>
<tr>
<td>4</td>
<td>How often does your child forage through the trash for food?</td>
<td>Never</td>
<td>A few times a year</td>
<td>1-2 times a month</td>
<td>1-3 times a week</td>
<td>4-7 times a week</td>
</tr>
<tr>
<td>5</td>
<td>How often does your child get up at night to food seek?</td>
<td>Never</td>
<td>A few times a year</td>
<td>1-2 times a month</td>
<td>1-3 times a week</td>
<td>4-7 times a week</td>
</tr>
<tr>
<td>6</td>
<td>How persistent is your child in asking or looking for food after being told “no” or “no more”?</td>
<td>Lets go of food ideas quickly and easily</td>
<td>Lets go of food ideas pretty quickly and easily</td>
<td>Somewhat persistent with food ideas</td>
<td>Very persistent with food ideas</td>
<td>Extremely persistent with food ideas</td>
</tr>
</tbody>
</table>
### Additional items:

<table>
<thead>
<tr>
<th>12. How old was your child when they first showed an increased interest in food?</th>
<th>13. How variable is your child’s preoccupation or interest in food?</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Less than 15 minutes a day</strong></td>
<td><strong>Hardly ever varies</strong></td>
</tr>
<tr>
<td><strong>15-30 minutes a day</strong></td>
<td><strong>Usually stays about the same</strong></td>
</tr>
<tr>
<td><strong>30 minutes to an hour a day</strong></td>
<td><strong>Goes up and down occasionally</strong></td>
</tr>
<tr>
<td><strong>1 to 3 hours a day</strong></td>
<td><strong>Goes up and down quite a lot</strong></td>
</tr>
<tr>
<td><strong>More than 3 hours a day</strong></td>
<td><strong>Goes up and down all the time</strong></td>
</tr>
</tbody>
</table>
Appendix 3.0: Research Advertisement

Leanne Haselip, Dr Martha Laxton-Kane and Dr Shirley Thomas from the University of Nottingham are about to undertake a research project looking at the relationship between levels of hyperphagia and amount of challenging behaviour in children with PWS. Their aim is to explore whether or not children with increased levels of hunger and hyperphagia also display increased challenging behaviour.

They are going to be sending out questionnaire packs to parents and carers of children (aged 18 and under) with PWS, who are also members of the PWSA-UK over the next few months. These packs will take about 30 minutes to complete and will provide the researchers with important information about any link between hyperphagia and challenging behaviour. If you would like any further information about this study, or are interested in taking part, please contact Jackie Waters at the PWSA (UK) office address.
Appendix 4.0: Ethical Approval Letter

Institute of Work, Health & Organisations
http://www.i-who.org

07 October 2008

Leanne Haselip
Doctorate in Clinical Psychology
University of Nottingham

Dear Lu

I-WHO Ethics Committee Review

Thank you for submitting your proposal on “Assessing the impact of hyperphagia on challenging behaviour in children with Prader-Willi syndrome”. This proposal has now been reviewed by I-WHO’s Ethics Committee to the extent that it is described in your submission.

I am happy to tell you that the Committee has found no problems with your proposal and is able to give approval.

If there are any significant changes or developments in the methods, treatment of data or debriefing of participants, then you are obliged to seek further ethical approval for these changes.

We would remind all researchers of their ethical responsibilities to research participants. The Codes of Practice setting out these responsibilities have been published by the British Psychological Society. If you have any concerns whatsoever during the conduct of your research then you should consult those Codes of Practice and contact the Ethics Committee.

You should also take note of issues relating to safety. Some information can be found in the Safety Office pages of the University web site. Particularly relevant may be: Sections 6.9, 6.10, 6.11, 6.14 of the Safety Handbook, which deals with working away from the University.
http://www.nottingham.ac.uk/safety/

Safety circulars:

Responsibility for compliance with the University Data Protection Policy and Guidance lies with all researchers.

Ethics Committee approval does not alter, replace or remove those responsibilities, nor does it certify that they have been met.

We would remind all researchers of their responsibilities:

- to provide feedback to participants and participant organisations whenever appropriate, and
- to publish research for which ethical approval is given in appropriate academic and professional journals.

Sincerely

[Signature]

Professor Nadina Lincoln
Chair, I-WHO Ethics Committee
Appendix 5.0: International cut off points for body mass index for overweight and obesity by gender, taken from Cole, Bellizzi, Flegal, & Dietz (2000) p. 4

<table>
<thead>
<tr>
<th>Age (years)</th>
<th>Body mass index 25 kg/m²</th>
<th>Body mass index 30 kg/m²</th>
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<tr>
<td></td>
<td>Males</td>
<td>Females</td>
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<tr>
<td>2</td>
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<td>18.0</td>
</tr>
<tr>
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