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The Characteristics of Temper Outbursts in Prader-Willi Syndrome

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ABSTRACT

The purpose of this study was to develop a comprehensive understanding of temper outbursts in Prader-Willi syndrome (PWS). A survey was developed from interviews conducted with individuals with PWS and their caregivers. The survey was completed by 101 primary caregivers. The findings suggest that outburst frequency decreases with age while duration increases. Adolescents exhibited more severe behaviors than children or adults. No differences were found across gender or genetic subtype. Provocations fit into three themes: goal blockage, social injustice and difficulty dealing with change. Distracting the person or giving them space to calm down were the only management strategies judged effective. Risperidone, Sertraline and fluoxetine were the most common medications prescribed for outbursts, though parents reported only minor effects.

Keywords: Prader-Willi syndrome, PWS, outbursts, aggression, rages.

Prader-Willi syndrome (PWS) is a neurodevelopmental disorder that arises from the absence of expression of paternally-inherited imprinted genes in the chromosome 15q11-q13 region This can be due to one of three mechanisms: in 70% of cases it is due to a paternal deletion; in 20-30% of cases it is due to maternal uniparental disomy (UPD) of chromosome 15; and in fewer than 5% of cases it is due to gene translocation or mutation of the imprinting center (Cassidy & Driscoll, 2008).

Parents of children with PWS report higher rates of parental control, worry, anger, marital conflict and inconsistent parenting than do parents of children with fragile X or Williams syndrome (van Lieshout et al., 1998). The increased stress and reduced well-being experienced by parents correlate with the behavior problems displayed by the individual with PWS (Hodapp et al., 1997; Lanfranchi & Vianello, 2012). Temper outbursts are one of, if not *the* most common, maladaptive behavior reported by parents of children, adolescents and adults with PWS (Dykens & Cassidy, 1995; Einfeld et al., 1999a; Holland et al., 2003). Compared to typically developing (TD) children and children with Down syndrome, children with PWS display a more rapid and later onset of outbursts, more severe outbursts, and continue exhibiting outbursts to a much later age (Dimitropoulos et al., 2001; Dykens et al., 1992; Einfeld et al., 1999b; Rice et al., 2015). Despite the severity and negative impact of outbursts, little is currently known about the characteristics of these behaviors in PWS.

Stein and colleagues (1994) conducted the first empirical investigation into the observable characteristics of PWS outbursts. This study involved the completion of a survey by 369 caregivers. The authors found that 90% of caregivers reported that their child displayed outbursts. Outbursts were more common in children and adolescents than in adults and this finding is supported by a more recent longitudinal study (Rice et al., 2015).

However, these studies only compared children and adolescents with adults. Thus, it is not yet known whether there are any changes in outburst frequency in the earlier years, such as childhood to adolescence. Furthermore, these studies examined outbursts based on measures that asked caregivers to rate how likely behaviors were to occur. The results were therefore largely based on outburst frequency or frequency and intensity combined. Understanding whether the frequency, intensity or duration of outbursts change over time within the PWS population would allow clinicians to predict prognoses and monitor treatment outcomes more effectively.

The only other known study that investigated the observable characteristics of PWS outbursts did so through semi-structured interviews with 14 caregivers of people with PWS (Tunnicliffe et al., 2014). The authors found variability in both the frequency and duration of outbursts. Due to the small sample size, it was not possible to investigate individual factors that would likely influence this variability; such as age, gender or genetic subtype. There is some evidence that externalizing behaviors are lower in individuals with a deletion compared to those with UPD (Dykens, Cassidy, & King, 1999). However, measures of externalizing behaviors are not a direct measure of outbursts. In regard to gender, we have previously reported similar rates of verbal aggression, physical aggression and outbursts in males and females with PWS (Rice et al., 2015). However, this study was conducted with adolescents and adults and there is some evidence to suggest that young boys with PWS may have greater difficulty with anger and self-control then young females (Dykens & Cassidy, 1995). Further research is therefore needed to determine whether the frequency, severity or duration of outbursts differ across genders and genotypes in PWS.

In the study by Tunnicliffe and colleagues (2014), caregivers were read a list of 21 antecedents and asked whether each had resulted in an outburst in the previous 12 months. Caregivers reported an average of ten antecedents, suggesting individuals with PWS could

become excessively distressed over a range of situations. The most common provocation was a change in routine or expectation. However, participants in this study were recruited as part of a larger study, which required all participants to display outbursts that were due to a change in routine. Consequently, conclusions about provocations must be interpreted with caution due to a potentially biased sample. Understanding the provocations to outbursts is important for two reasons. First, it is the essential piece of information needed for the development of a behavior management plan. Second, examination of the situations that lead to outbursts may provide clues to underlying psychological or physiological mechanisms driving such behaviors. Caregivers reported using a range of strategies to manage outbursts. However, the sample size for each intervention was too small to draw conclusions (Tunnicliffe et al., 2014). Understanding the effectiveness of interventions currently implemented by caregivers can potentially help direct future behavioral interventions.

Stein et al. (1994) used the clinical global impression (CGI) to assess response to medications in treating outbursts. The types of psychotropic medication participants were prescribed to manage behavior problems included serotonin reuptake inhibitors (SSRIs), antipsychotics, stimulants and anticonvulsants. Caregivers reported SSRIs to be helpful in treating outbursts; however, analysis of the CGI responses found no significant improvements. As this study was conducted in 1994, it did not include new-generation or atypical antipsychotics, such as risperidone.

Almost all individuals with PWS experience hyperphagia, making it the most common feature of PWS (Holland et al., 2003). Research conducted within the TD population has shown that people become more irritable when they are hungry (Bushman et al., 2014; Macht, 1999). Thus, it is logical to consider whether outbursts displayed by individuals with PWS could be an effect of hyperphagia. Dimitropoulos et al. (2001) examined the age of onset for hyperphagic and outburst behaviors in children with PWS.

These authors found that the age of outburst onset moderately correlated with the age of hyperphagia onset. However, the correlation analysis excluded participants who had not yet developed hyperphagia, making it difficult for conclusions to be drawn. A replication of the study by Dimitropoulos et al. (2001) that does not exclude participants who have not yet developed hyperphagia may help establish whether there is an association between these behaviors.

In sum, there are currently only a handful of studies that have directly investigated the characteristics of outbursts in the PWS population. These studies show that the onset of PWS outbursts is later for children with PWS than it is for TD children and children with Down syndrome. Children with PWS also display more severe outbursts and continue exhibiting outbursts to a much later age. Two studies suggest PWS outbursts decrease in frequency and intensity combined from around 18 years. However, it is not yet known whether there are any changes in outburst frequency in the earlier years, such as childhood to adolescence, or whether the intensity and duration of outbursts individually change over the lifespan. Understanding the situations that are likely to lead to a PWS outburst may provide clues to the causal mechanisms of these behaviors.

The aim of this study was to develop a comprehensive understanding of the characteristics of PWS outbursts that could then be used to direct behavioral interventions and investigations into the causal mechanism of these behaviors. Specifically, this study examined whether:

1. the onset of PWS outbursts and hyperphagia occurred around the same time
2. age, gender or genetic subtype influenced the frequency, intensity and duration of PWS outbursts
3. people with PWS shared common patterns in the provocations, behaviors and setting events of outbursts
4. caregivers were using any behavioral and/or pharmaceutical interventions that they

found effective in managing PWS outbursts.

METHOD

Participants

The study was reviewed and approved by the University of Sydney Human Research Ethics Committee. Participants were invited to complete an online questionnaire circulated by Australian and international Prader-Willi syndrome associations. The eligibility criteria included (1) being a primary carer of a person with PWS, (2) being someone who had spent at least 20 hours a week with the individual with PWS over a minimum of 6 months and (3) the person with PWS displayed at least one outburst a month. An outburst was defined as becoming very angry or upset in a way that seemed excessive for the situation and beyond the person's control. All participants provided informed consent prior to completing the survey. Data was collected between May 2013 and May 2014. Genetic diagnosis of PWS was confirmed by either genetic report or confirmation from a medical professional.

Development of the survey

A PWS outburst survey was developed for this study based on information collected through 22 semi-structured interviews. The purpose of the interview was to gather a list of provocations, behaviors, setting events, and management strategies that could be used to develop a PWS outburst survey. The interviews were conducted until there were no new responses. In total, 13 mothers, 3 fathers, 5 individuals with PWS and 1 day-center coordinator participated in the interviews. The interview focused on individuals with PWS aged between 7 and 42 years and with a mean age of 20.91 years. The interview questions were open-ended and asked participants about outburst duration and frequency, situations that were likely to lead to an outburst, setting events, behaviors commonly displayed during

an outburst, and strategies individuals had tried to manage outbursts.

The interviews were recorded and transcribed verbatim and hand-coded by LR as either a provocation (situations that would likely lead to an outburst), setting event, behavior displayed during an outburst or a management strategy. Clips of each item were then extracted along with any additional information to ensure the context was not lost. These were then reviewed and discussed with SE. Together, LR and SE developed terms for descriptions that had the same meaning (e.g. yell, shout, and raise voice became shout/raise voice). The final list of each category (provocation, behavior, setting event, management strategy) was then used to form the items for the close-ended questions in the survey. The survey was reviewed by KW to ensure that the questions and the responses were correctly understood. See supplementary material for a summary of the survey questions.

For outburst frequency, intensity and duration, parents were asked to base their responses on outbursts that had occurred in the previous month. Parents chose their response from a seven-point Likert scale with geometric steps. The scale was based on a model that was used to examine outburst frequency and duration in TD toddlers (Potegal & Davidson, 2003). A list of provocations, behaviors and setting events was provided. Caregivers were asked to indicate how often, based on the following options, each provocation, behavior or setting event occurred within the previous 12 months: ‘1. Never’, ‘2. Sometimes’, ‘3. Often’, ‘4. Always’. This scale has been used in several well-validated measures, such as the Developmental Behavior Checklist (Einfeld & Tonge, 2002), as well as in outburst research conducted with TD children (Potegal & Davidson, 2003). Management strategies were also listed, and caregivers were asked to choose from the following options: ‘Not tried’, ‘Not effective at all’, ‘Somewhat effective’, or ‘Very effective’. The provocations, behaviors, setting events and management strategies listed were based on answers from the interviews as well as from information collected through a review of the literature. There was also always

an ‘Other’ option for parents to provide answers that were not listed. The survey was conducted via an online survey program called Lime Survey and was made available to participants in English and Spanish. The survey was translated into Spanish following an expression of interest for more research to be made available to Spanish speaking PWS families. The survey was translated into Spanish by Dr Ana Fernández, a Postdoctoral Research Associate at the University of Sydney, fluent in both Spanish and English. Following the first translation, the author met with Dr Fernández to discuss any issues. There were several words that could not be translated directly into Spanish. The intended meaning of each word was discussed, and an alternative Spanish word or phrase were agreed upon.

Statistical analysis

Descriptive statistics were used to analyze outburst behaviors, provocations, setting events and interventions. Test-retest reliability of three questions, pertaining to outburst frequency, duration and intensity, was measured by inviting 30 participants to complete a second questionnaire two weeks later. A weighted version of Cohen’s kappa was used to calculate absolute agreement for frequency and duration. For intensity, test-retest reliability was assessed using intra-class correlation (ICC). An attempt was made to collect data from a sub-sample of second informants to assess inter-informant reliability but only 3 of 15 caregivers consenting to this provided data. Participants were classified into three age groups: children (1-10 years), adolescents (11 to 18 years) and adults (19 years and up). Data for frequency and length of outbursts were nominal so non-parametric analyses were conducted. Two Kruskal-Wallis tests were conducted to explore whether outburst frequency and duration differed across the three age groups, where age was a between-subject variable.

RESULTS

Participants

A total of 101 primary caregivers completed the survey. Individuals with PWS ranged in age from 1 to 64 years, with a mean age of 16.14 (SD 10.91). The mean age for the three age groups were 6.85 (SD 2.12) years for children, 14.65 (SD 1.81) years for adolescents and 27.97 (SD 10.85) years for adults. There were 62 males and 39 females with PWS; 57 people with a deletion, 36 with uniparental disomy, two with an imprinting defect and six participants for which genetic subtype data was not available. The number of participants in each age group is provided in Table 1. Genetic confirmation of diagnosis was available for 80% of participants. The 20% for whom genetic confirmation was not available were compared on all of the below questions to the 80% with confirmation. No difference was found, so all participants were included in the analysis. Of the people with PWS, 87 were living at home with their parents or guardians, five lived in a PWS-specific group home, two in a non-PWS-specific group home, four in their own home with supported living, one person was living independently, and one was in a residential school. Participants were from Australia (n = 15), Argentina (n = 1), Canada (n = 8), Chile (n = 2), England (n = 8), Mexico (n = 1), New Zealand (n = 5), Peru (n = 2), Philippines (n = 1), Puerto Rico (n = 1), South Africa (n = 1), Spain (n = 1) and the United States (n = 55).

Place Table 1 about here

Onset of outbursts

The participants ranged in age from 1 to 64 years. To avoid memory inaccuracy, the online survey was designed to ask about the order of onset of outbursts and hyperphagia only of caregivers to a person with PWS who was under the age of 12. Of the 41 caregivers who met this criterion 41% reported that outbursts began between 1 and 3 years; 49% reported outbursts occurred between 4 and 6 years; 7% reported outbursts occurred between 7 and 9 years; and 3% reported outbursts occurred between 10 and 12 years. Regarding the onset of

outbursts and hyperphagia, 68% of caregivers reported that outbursts started before excessive eating, 18% reported excessive eating occurred before outbursts, and 15% reported the two behaviors started around the same time.

Outburst characteristics across age

Frequency

Test-retest reliability was assessed using a weighted Cohen's kappa. A significant moderate level of agreement was found: Kappa = 0.56 (p = <.0.001), 95% CI (0.45, 0.68) (Landis and Koch, 1977). A Kruskal-Wallis non-parametric test was used to determine whether outburst frequency differed across the three age groups. With alpha set at 0.05, the test was significant, $x2$ (2, n = 101) = 8.94, p =.011. Three post hoc comparisons using Mann-Whitney U tests were conducted to compare children with adolescents, adolescents with adults and children with adults. A Bonferroni adjustment was made for three tests (p = 0.016). The only Mann-Whitney U test that was significant was between children and adults, z (n = 63) = 2.4, p = <0.015, with children (2 to 3 times a week) displaying significantly more outbursts than adults (once a week).

Intensity

Test-retest reliability was assessed using ICC. A high level of agreement was found, ICC = 0.76, with a 95% confidence interval from 0.6 to 0.86. A one-way analysis of variance was conducted to determine whether there was a difference in outburst intensity across the three age groups. There were no significant differences.

Duration

Test-retest reliability was assessed using a weighted Cohen's kappa. A significant moderate level of agreement was found, Kappa = 0.52 (p = <.0.001), 95% CI (0.35, 0.69) (Landis and Koch, 1977). A Kruskal-Wallis non-parametric test was used to determine whether outburst

duration differed across the three age groups. With alpha set at 0.05, the test was significant, $x2$ (2, n = 100) = 17.8, p = <.001. Three post hoc comparisons using Mann-Whitney U tests were conducted to compare children with adolescents, adolescents with adults and children with adults. After Bonferroni adjustments for the three tests (p = 0.016), a significant difference was found between children and adolescents (z (n = 68) = 3.9, p = <0.001) and between children and adults (z (n = 62) = 2.8, p = <0.005). For children outburst duration was typically between 0 and 15 minutes and for adolescents and adults outburst duration was typically between 10 and 30 minutes. See Table 2 for descriptive statistics of outburst frequency, intensity and duration.

Place Table 2 about here

Outburst characteristics across gender and genetic subtype

Two Mann-Whitney U tests were conducted to compare males and females on the frequency, length and intensity of outbursts. There was no difference between genders for any measure. Nine Mann-Whitney U tests were conducted to compare the frequency, length and intensity of outbursts for two genetic subtypes (deletion and UPD) for each age group (children, adolescents and adults). There were no differences across genetic subgroups for any measure.

Provocations

Table 3 outlines the percentage of caregivers who reported whether a situation would likely cause an outburst for each age group. The average number of provocations for one individual was 14, with a range of 6 to 19. Caregivers were also asked to list any additional situations that were likely to result in an outburst. No new situations were described. As can be seen in Table 3, being told no, being asked to do something non-preferred, and changes to routines/expectations were the most commonly reported provocations, reported by upwards of 90% of the sample.

Place Table 3 about here

Behaviors

Table 4 outlines the percentage of caregivers who reported whether each behavior listed would, at least sometimes, occur during an outburst.

Place Table 4 about here

Setting events

Table 5 outlines the percentage of caregivers who reported that outbursts occurred at least sometimes with each person, in each location, during each event and associated with each biological factor.

Place Table 5 about here

Behavior management strategies

Table 6 outlines the percentage of caregivers who reported each behavior management strategy to be somewhat or very effective. Caregivers reported the most effective strategies to be distracting or divert the person's attention and giving the person space to calm down.

Place Table 6 about here

Clinical behavior interventions

Table 7 outlines the percentage of caregivers who reported each clinical behavior intervention to be somewhat or very effective. Caregivers reported the most effective strategies to be behavior management plans and parenting programs.

Place Table 7 about here

Pharmaceutical interventions

Four individuals with PWS were prescribed a psychotropic drug to be administered at the

time of an outburst (pro re nata, PRN). These participants were males aged 14 to 21 years. The medications included valium (rated as not effective), topiramate (rated as somewhat effective), quetiapine (rated as very effective) and lorazepam (rated as very effective). Thirty-two participants were prescribed ongoing medication to manage outbursts. These participants ranged in age from 6 to 33 years, with a mean of 18 years; there were 19 males and 13 females; 17 people had PWS due to a deletion and 14 due to UPD. Nineteen people were taking one medication, eight were taking two medications, three were taking three medications and two were taking four medications. Types of medication included anticonvulsants (n =4), stimulants (n = 2), mood stabilizers (n = 1), benzodiazepine (n = 2) and anxiolytic (n = 1). The most commonly prescribed medications were risperidone (n = 10), sertraline (n = 8) and fluoxetine (n = 7). Parents reported these medications to be somewhat effective.

DISCUSSION

The aims of this study were to determine whether (1) the onset of PWS outbursts and hyperphagia occurred around the same time, (2) PWS outbursts changed in frequency, intensity or duration over the course of development or across genetic subtypes or gender, (3) there were shared qualities or patterns in the provocations and behaviors displayed during PWS outbursts and (4) there were any behavior management strategies or psychotropic interventions that parents found useful in managing PWS outbursts. Results are discussed in detail below.

Does the onset of PWS outbursts occur at the same time as hyperphagia onset?

In the TD population, 91% of children display outbursts by the time they turn 3 (Potegal & Davidson, 2003). In contrast, only 41% of children with PWS were reportedly displayed

outbursts by 3 years. For the remaining 59%, outbursts onset occurred between 4 and 12 years. These findings are in line with previous research, which suggests outburst onset occurs later in children with PWS than the TD population and children with Down syndrome (Dimitropoulos et al., 2001).

If the onset of outbursts and hyperphagia occurs at the same time, then there would be reason to suggest that they may share a causal link. Dimitropoulos and colleagues (2001) compared the age of onset for hyperphagic and outburst behaviors in children with PWS. These authors found that the age of outburst onset moderately correlated with the age of hyperphagia onset. However, the correlation analysis excluded participants who had not yet developed hyperphagia, making it difficult for conclusions to be drawn. In the present study, 68% of caregivers reported that outbursts started before excessive eating, 18% reported excessive eating occurred before outbursts and 15% reported the two behaviors started around the same time. These findings suggest that outbursts most often occur before hyperphagia.

However, Butler et al. (2010) found the age at which body mass index starts to increase in children with PWS beyond that of the TD population predates the age at which parents start noticing changes in eating behaviors. This suggests that the physiological mechanisms of hyperphagia may be present before parents begin noticing signs of hyperphagia, and therefore may be present at the onset of outbursts. Thus, while outbursts may not be directly caused by hyperphagia, the mechanisms driving these two symptoms may still arise around the same time and therefore be related.

Are PWS outbursts influenced by age, gender or genetic subtype?

Previous research has shown that PWS outbursts decline, in both intensity and frequency, from around 19 years (Rice et al., 2015; Stein et al., 1994). The present study aimed to extend on this research by looking at changes in outburst frequency, duration and intensity over the

lifespan. Our results suggest that while people with PWS have fewer outbursts as they get older, the duration of these outbursts increases from childhood to adolescence. We did not find a change in outburst intensity, suggesting that parents' perception of the level of upset or anger experienced by the individual during the outburst remains the same across the lifespan. Frequency, intensity and duration of outbursts did not differ between males and females. This was in line with previous research, which found the combined frequency and intensity of outbursts to be similar across genders in individuals with PWS (Rice et al., 2015). This was also consistent with the duration and frequency rates of outbursts displayed by TD children (Österman & Björkqvist, 2010). It is, however, dissimilar to rates of outburst in clinical populations, where outburst rates are much higher in boys than girls (Carlson et al., 2016; Potegal et al., 2009). Examining the differences in symptoms across genetic subtypes is important for elucidating causal mechanisms. Consistent with previous research, the present study found that the frequency, duration and intensity of outbursts for those with PWS due to a deletion and UPD was similar (Didden et al., 2007). This finding suggests that if outbursts are due to a genetic abnormality, then this abnormality is likely to be present in both genetic subtypes.

Are there shared qualities or patterns in the provocations to PWS outbursts?

One aim of this study was to determine whether individuals with PWS shared similarities in the provocations to outbursts. When the provocations were compared with previous research, three overarching themes emerged. Previous research has shown that individuals with PWS display outbursts following a change in routine or expectation (Tunnicliffe et al., 2014; Woodcock et al., 2011) and that this is due to difficulties in switching attention (Woodcock et al., 2009). In line with this, the present study found difficulties with 'change in routine and expectation' and 'inability to take their mind off a thought or idea' were common

provocations to PWS outbursts likely to be associated with a deficit in task-switching.

The most common provocation to outbursts for TD children is conflict with parents. Family conflict studies suggest conflict between children and parents and children and children are most often due to the blocking of a desired goal or a violation of social expectations, such as not following rules or treating others unfairly (Carpenter & Halberstadt, 2000; Fisher & Johnson, 1990). The provocations to PWS outbursts fit these categories. Provocations associated with the blocking of a desired goal include being told no to something they asked for other than food, being asked to do something they do not want to do, being denied food, and misplacing a possession. Examples of provocations associated a violation of social expectations include being accused or confronted, feels he/she is being treated unfairly, feels someone is getting more/better food than they are, feels he/she is not receiving the independence they deserve, being teased and being ignored. These three themes (a change in routines, blocking of a desired goal and a violation of social obligations) could be used to help guide the development of new behavior management strategies.

Are there shared qualities or patterns in the behaviors displayed during PWS outbursts?

Crying, raising voice, screaming, and arguing were the most common behaviors displayed during an outburst for all three age groups. Severe outburst behaviors were reported to occur more commonly in adolescents than in children or adults. These behaviors included biting self and others, punching, head banging, throwing objects at others and damaging property. Research conducted with the TD population suggests that the onset of puberty is associated with intensified emotions (Hare et al., 2008; Larson et al., 2002). A meta-analysis further found that while rates of parent-child conflict decrease, the emotional intensity of disagreement increases from early adolescence (Laursen, Coy, & Collins, 1998). Thus, the

increase in severe behaviors during adolescence may reflect heightened emotional experience associated with this period.

Are there setting events that are likely to lead to an increase in PWS outbursts?

Outbursts displayed by TD young children most commonly occur at home (Belden et al., 2008). Carlson and Dyson (2012) found that the context in which an outburst occurred differed depending on the diagnosis. Children whose parents and teachers reported outbursts were most likely to have a diagnosis of a severe mood disorder; children whose parent only reported outbursts were more liable to be diagnosed with an anxiety disorder and children whose teacher only reported outbursts were typically diagnosed as having language and learning disorders. We found that for individuals with PWS of all ages, outbursts most commonly occurred at home with parents. However, PWS outbursts also frequently took place at school, in work environments and public settings, as well as with teachers or support workers. These findings suggest that the setting of PWS outbursts is most similar to that of children with severe mood disorders.

Embregts et al. (2009) found that an overly exciting environment was a common precipitating factor to aggression in people with mild ID. Similarly, we found that 82% of parents reported outbursts to at least sometimes occur during a celebration, such as Christmas or a birthday. We also found that parents of adolescents and adults reported outbursts to be common during the holidays and parents of children reported outbursts to be common when visitors came over. These occasions are typically associated with a change in routine, which may be the reason for the increased outbursts. Daily schedules that allow people to know what is happening each day as well as signals for when changes are about to occur may help people with PWS deal with these events (Bull, Woodcock, & Oliver, 2017).

Are there effective behavior management strategies for PWS outbursts?

The most effective behavior management strategy was to distract or divert the persons attention. The remaining strategies that were reported to be at least somewhat effective by the majority of parents all related to giving the person time and space to calm down. These included, taking the person away from the situation, removing anyone at risk and walking away from the person. Reminding the person of the consequences was endorsed as being somewhat effective by 47% of parents. Verbal insistence and trying to reason with the person was only reported to be effective by 34% and 28%, respectively.

Geleerd (1945) suggests that whether an intervention is effective in reducing an outburst is dependent on the person's emotional maturity. A child with the maturity of an infant is unable to respond appropriately to reasoning due to an ability to effectively regulate emotions. A recent study provided support for this hypothesis by showing that self-regulation skills strongly predicted outburst characteristics of TD children (Roben et al., 2013). Children begin to show vast improvement in these regulatory skills between the ages of 24 and 30 months, the same age when outbursts start to decline (Potegal & Davidson, 2003). Similarly, a study conducted with adolescents with ASD found that poor emotion regulation, specifically a reduction in cognitive reappraisal, was associated with increased negative emotions and maladaptive behavior, such as tantrums and aggression (Samson, 2015).

To date, there has been little empirical work examining emotion regulation abilities of people with PWS. We know that despite their level of cognitive abilities, individuals with PWS have limited coping skills (Dykens et al., 1992), difficulty perceiving the view of others (Lo Siemensma, Collin, & Hokken-Koelega, 2013) and poor social judgement (Benarroch et al., 2007). However, more work is needed to evaluate the level of emotion regulation abilities in people with PWS.

Are there clinical behavior interventions for PWS outbursts?

The most effective and commonly tried clinical behavior intervention was behavior management plans, which were reported to be somewhat effective for all three age groups. Parenting programs were also reported to be somewhat effective for all three age groups, however, only 17% of participants had tried parenting programs. Family therapy and psychological interventions, such as anger management plans, were reported to be slightly less effective than behavior management plans and parenting programs.

Are there effective pharmaceutical interventions for PWS outbursts?

Only four participants had been prescribed a psychotropic medication to take at the time of an outburst. This is not surprising given outbursts typically last less than 30 minutes, and most medications take up to 20 minutes to be absorbed and have an effect. Thirty-two participants were prescribed ongoing medication to manage outbursts. The most common ongoing medications prescribed were risperidone, sertraline and fluoxetine. However, most parents reported these medications to be only be 'somewhat effective' at reducing outbursts. Thus, it seems that there is currently no known pharmaceutical intervention that effectively reduces PWS outbursts.

Potential limitations

Several limitations should be noted. First, this study employed a cross-sectional design. The primary limitation of using a cross-sectional design is that it is difficult to distinguish between differences that are due to changes in age and cohort effects. Second, attempts to test inter-rater reliability of the survey produced insufficient data. Nevertheless, test re-test reliability was moderate to high. Third, the study relied on participant recall rather than direct observation of outbursts. There is some evidence that greater weight can be placed on

negative behaviors when measured through recall. Finally, we did not collect information about cognitive or adaptive functioning, which may relate to some of the variables examined.

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Tables

Table 1. Number of participants in each age group by genetic subtype

	Children (1-10 years)		Adolescents (11-18 years)		Adults (19 years +)	
	Male	Female	Male	Female	Male	Female
Deletion	11	8	12	3	7	15
UPD	9	6	9	5	4	3
Imprinting/translocation or unknown	2	1	1	0	1	0
Totals	22	15	22	8	9	18

Table 2. Descriptive statistics for frequency, intensity and duration of outbursts

	Children	Adolescents	Adults
Frequency			
n	37	31	33
Mean	4.1	3.5	2.9
SD	1.6	1.6	1.7
Median	4	4	2
Intensity			
n	37	31	33
Mean	5.6	5.8	5.7
SD	1.3	1.4	1.3
Median	6	6	6
Duration			
n	37	31	33
Mean	1.7	2.8	2.7
SD	0.7	1.2	1.3
Median	2	3	2

Note: Frequency: 1 = one outburst a month, 2 = 2 – 3 outbursts this month, 3 = once a week, 4 = 2 – 3 times a week, 5 = one a day, 6 = 2 – 3 times a day and 7 = 4 a day or more. Intensity: 1 = a little upset or angry and 7 = very upset or angry. Duration: 1 = less than 5 minutes, 2 = 10 to 15 minutes, 3 = 15 to 30 minutes, 4 = 30 minutes to an hour, 5 = 1 hour to 2 hours, 6 = more than 2 hours and 7 = a day or more.

Table 3. Percentage of caregivers who reported provocation would lead to an outburst

Provocation	Children	Adolescents	Adults	Total
Being told no to something they have asked for (other than food)	97	93	91	94
Being asked to do something he/she does not want to do, e.g., take a shower	100	90	88	93
Change in expectations, plans or routine	91	93	94	93
Being accused of or confronted about something they may have done wrong	85	90	94	90
Being denied food	86	93	90	90
Being rushed/hurried along	86	77	94	86
Having difficulty understanding/ considering another person's point of view	81	87	90	86
Feeling he/she is being treated unfairly	72	90	91	84
Someone touching or moving their possession/s	82	69	91	81
Having difficulty taking mind off a thought or idea	76	83	85	81
Misplacing a possession	84	67	85	79
Someone did not understand what they were saying	77	53	94	75
Feeling someone is getting more/better food than they are	56	80	81	72
Feeling he/she is not receiving the independence they deserve	69	69	76	71
Being teased	48	63	79	63
Being ignored	62	54	64	60

Table 4. Behaviors displayed during an outburst

Behavior	Children	Adolescents	Adults	Total
Crying	92	97	94	97
Raising voice/shouting	94	93	97	95
Screaming	92	97	97	94
Arguing	88	93	94	93
Stomping off	69	75	67	70
Slamming doors	56	82	75	70
Damaging property	63	83	58	67
Pushing	56	71	70	66
Throwing objects - not at anyone	68	72	56	66
Talking to self	53	52	73	60
Kicking	62	64	39	55
Running away	58	68	43	53
Name calling	31	67	64	52
Swearing	13	51	76	49
Dropping to the ground	77	54	15	48
Punching	38	61	33	44
Throwing objects at people	39	59	36	44
Self-injury	30	43	48	40
Pulling hair	38	35	29	31
Biting others	29	46	19	30
Head banging	24	41	21	28
Biting self	18	46	3	16

Table 5. Accompanying person and location of outburst

	Children	Adolescents	Adults	Total
Accompanying person				
Mother	97	100	85	94
Father	100	96	80	92
Teacher/support worker	89	87	79	86
Sibling (within 3 year age difference)	82	80	62	74
Peers	81	73	66	73
Sibling (older by more than 3 year)	72	61	59	63
Grandparent	75	74	34	54
Sibling (younger by more than 3 years)	43	30	8	27
Location of outburst				
Family home	89	100	87	96
Public places	92	97	91	93
School/college/work/day program	94	93	79	91
Place of residence if not family home	0	42	81	73
Social setting/friend's house/social support groups	84	78	53	71
Events				
Celebrations (e.g. Christmas, Birthday)	80	83	82	82
Visitors over	74	64	60	66
Home for the holidays	39	82	75	65
Holiday away from home	20	86	70	59

Table 6. Behavior management strategies

Strategy	Child	Adolescent	Adult	Total
Distract or divert the person	82 (2.2)	90 (2.1)	81 (2.1)	84 (2.1)
Take the person away from the situation or the situation away from the person	81 (2.1)	66 (1.8)	71(2.0)	74 (2.0)
Remove anyone who may be at risk of harm and wait out the outburst	80 (2.1)	61 (1.8)	70 (1.8)	70 (1.9)
Ignore the individual/walk away	60 (1.6)	62 (1.6)	74 (1.9)	64 (1.7)
Remind the person of consequences	43 (1.4)	45 (1.6)	37 (1.2)	47 (1.4)
Verbal insistence, e.g. ‘Stop it.’ or ‘That's enough.’	37 (1.4)	31(1.4)	34 (1.3)	34 (1.4)
Try to reason with the person	43 (1.5)	18 (1.2)	22 (1.2)	28 (1.3)

The percentage of parents who reported that they have tried the strategy is listed first and then how effective parents reported the strategy to be is listed in brackets. Note: 1 = not effective at all, 2 = somewhat effective, 3 = very effective.

Table 7. Clinical behavior interventions

Strategy	Child	Adolescent	Adult	Total
Behavior management plan	42 (2.0)	76 (1.7)	83 (2.1)	67 (2.0)
Parenting program	22 (2.0)	14 (1.75)	14 (2.0)	17 (1.9)
Family therapy	15 (1.8)	43 (1.6)	32 (1.8)	30 (1.7)
Psychological intervention such as anger management program	15 (2.0)	57 (1.5)	33 (1.6)	35 (1.6)

The percentage of parents who reported that they have tried the intervention is listed first and then how effective parents reported the strategy to be is listed in brackets. Note: 1 = not effective at all, 2 = somewhat effective, 3 = very effective.