Educating Students with Prader-Willi Syndrome

Prader-Willi Syndrome (PWS) is a complex genetic condition characterised by a propensity to abnormal and excessive eating behaviour and a range of medical and behavioural features. There is also evidence of a distinctive cognitive profile which has implications for how the affected children learn. Having an understanding of the cognitive and behavioural profile of PWS is essential in helping affected children reach their potential in educational settings.

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INTRODUCTION

Prader-Willi Syndrome (PWS) is a genetically determined neurodevelopmental disorder resulting from the absence of expression of one or more genes on chromosome 15 (Whittington and Holland, 2004). In the majority of cases, the critical genes are lost from the chromosome normally contributed from the father - known as 'Deletion Type PWS'. In most of the remaining cases the entire chromosome of the father is missing and there are instead two chromosome 15s from the mother - known as 'Uniparental Disomy (UPD) PWS'. Estimates for the prevalence of the disorder range from 1 in 10,000 to 1 in 22,000 (Reddy and Pfeiffer, 2007), and it occurs in both genders and all races. Although it is a genetic condition, it is not usually hereditary and the vast majority of affected individuals are the only affected individuals in their family.

In PWS three significant areas of concern interact (Whittington and Holland). First, there is the developmental delay and the cognitive and functional impairments characteristic of those with an intellectual disability. Children with PWS typically function in the borderline to moderate intellectual disability range (Curfs, 1992). Second, there is the marked tendency to over-eat (hyperphagia). This most striking behaviour involves the tendency to severe and persistent over-eating which if unrestricted, leads to life-threatening obesity (Holland, Treasure, Coskeran, Dallow, Milton and Hillhouse, 1993). Third, there is the propensity to particular behaviour patterns that can have a negative impact on the child's quality of life and that of his or her family. PWS is associated with an increased risk for a range of behavioural difficulties including temper tantrums, impulsivity, skin-picking, and repetitive and compulsive behaviours including hoarding, ordering items, and repetitive questioning (Dykens, Hodapp and Finucane, 2000).

Physical Characteristics and Associated Medical Conditions

The physical characteristics of children with PWS include short stature, small hands and feet, hypopigmentation (reduced skin pigmentation), hypogonadism (incomplete pubertal development), hypotonia (weak muscle tone) at birth, and characteristic facial features (Holm, Cassidy, Butler, Hanchett, Greenswag, Whitman and Greenberg, 1993). The short stature of people with PWS has been attributed primarily to decreased levels of growth hormone and appears to be present from birth

(Gillessen-Kaesbach, Robinson, Lohmann, Kaya-Westerloh, Passarge and Horstthemke, 1995) and is compounded by a lack of growth spurt prior to the time puberty would normally occur (Butler and Meaney, 1991). Treatment with growth hormone has been shown to improve short stature and improve physical development of children with PWS (Cassidy and McCandless, 2005). Hypopigmentation for the family, manifested as fairer skin, hair, and eye colour, occurs in about one-third of affected individuals (Cassidy and McCandless). Children with PWS have underdeveloped sexual organs (hypogonadism) and puberty may be delayed and incomplete. Hypotonia (weak muscle tone) at all ages is reflected in decreased muscle bulk and tone as well poor coordination and often decreased strength. Characteristic facial features including almond-shaped eyes, narrow nasal bridge, and a down-turned mouth with a thin upper lip, are either present from birth or evolve over time in most affected individuals (Cassidy and McCandless). Children with PWS are at higher risk than the general population for scoliosis and osteoporosis (Whittington and Holland, 2004). Sleep difficulties are also a common feature in PWS. Children with PWS may also have raised pain thresholds and self-harm, usually in the form of skin-picking, is common. Poor temperature perception is also common and this may include mild symptoms such as wearing clothing unsuited to the season of the year (Whittington and Holland). Other characteristics include thick viscous saliva and a high threshold for vomiting (Cassidy and McCandless).

Hyperphagia

One of the most prominent features of PWS is the abnormal interest in food, the excessive eating behaviour (hyperphagia), and the resultant obesity (Whittington and Holland, 2004). At birth babies with PWS are initially hypotonic and have poor suck, usually necessitating tube feeding. At some point, usually in the first two years of life, appetite appears to improve and the child begins to gain weight excessively (Greenswag, 1987). It appears that the basis of the excessive eating behaviour in PWS is that the satiation response is impaired so that the child never feels full. As a result, the feelings of hunger only briefly diminish following significant calorie intake and affected children have a continuous urge to eat that they cannot learn to control (Whittington and Holland). To compound this problem children with PWS need less food than their peers without the syndrome because their bodies have less muscle and tend to burn fewer calories. Hyperphagia in individuals with PWS is not explained by weak character or due to a lack of willpower around food. Rather, hyperphagia in individuals with PWS results from altered function of the hypothalamus (Swaab, Purba and Hofman, 1995). Appetite suppressant and behaviour modification techniques aimed at reducing overeating have been found to have limited effects (Dykens, Leckman and Cassidy, 1996).

Behavioural Profile

In addition to the eating and food related behaviours, temper tantrums, skin-picking, and extreme mood fluctuations are among the most prevalent of maladaptive behaviours in children with PWS (Whittington and Holland, 2004). Reddy and Pfeiffer (2007) found that children with PWS exhibited more emotional and behavioural problems than children with similar levels of intellectual disability without the syndrome. Obsessional and compulsive behaviours in PWS are common, and are not just food related and can include ordering and arranging of items and

engaging in ritualistic and routine behaviours (Dimitropoulous, Feurer, Butler and Thompson, 2001). Repetitive questioning, needing to tell, and hoarding are also common (Dykens, Hodapp, Walsh and Nash, 1992), while cleaning, counting, and checking behaviours may also be present (Whittington and Holland).

Learning Profile

Some of the characteristic learning strengths and weaknesses of children with PWS are displayed in Table 1.

Table 1: Learning Strengths and Weaknesses associated with Prader-Willi Syndrome

Characteristic Learning Strengths	Characteristic Learning Weaknesses
Long-term memory for information	Short-term auditory memory leading to difficulties in acquisition of arithmetic skills and verbally presented information
Receptive language	
Visually based learning through pictures, illustrations and videos	Expressive language (although verbal abilities improve with age)
Hands-on experiences	Fine motor skills, related to strength, tone, and motor planning
	Interpreting subtle social cues, learning subtle social norms and applying knowledge in new situations

Adapted from Chedd, Levine and Wharton (2006).

Four main cognitive or learning difficulties have been identified in children with PWS: intellectual disability, language processing problems, specific learning difficulties associated with short-term auditory memory and sequencing deficits, and failure to develop the ability to apply knowledge in new situations (metacognitive ability) (Eihozler, Gellatly, Lee, Ritzen and Whitman, 2001). It is generally found that children with PWS fall within the range of mild intellectual impairment although there is a wide range of abilities ranging from average functioning to moderate intellectual disability. As well as a global lowering of ability, there may also be more specific cognitive strengths and weaknesses (Whittington and Holland, 2004). Language development may also be delayed although verbal skills are an ultimate strength in most individuals (Cassidy and McCandless, 2005). Language difficulties for children with PWS may include poor speech-sound development, reduced oral motor skills, hypernasality, flat intonation patterns, an abnormal pitch, and a harsh voice quality (Lewis, 2006).

Difficulties with auditory short-term memory mean that children with PWS typically have difficulties with mathematics and may be unable to cope with complex verbal instructions. Affected children frequently have difficulty with sequential processing (Dykens et al., 1992), resulting in difficulties in telling the time and comprehending temporal order (days of week and months of year). However, children with PWS

typically display a relative strength in simultaneous processing in comparison with sequential processing. Information presented using a simultaneous approach resembles a “visual whole” or "gestalt". In terms of academic subjects, children with PWS have a relative strength in word reading and less so in spelling, but not in mathematics compared to other children with intellectual disability (Whittington and Holland). Children with ‘UPD Type PWS’ may perform better than children with ‘Deletion Type PWS’ in reading, spelling, and mathematics, with this advantage being greatest in reading (Whittington and Holland). In contrast, children with ‘Deletion Type PWS’ may show greater ability to complete jigsaw puzzles and relatively better fine motor skills (Whittington and Holland). Children with PWS often have difficulties applying learned information in new situations and generalising learning across environments. However, they respond well to ‘hands-on’ and experiential teaching strategies and such strategies are likely to increase the generalisation of their learning.

EDUCATIONAL PROVISION AND TEACHING STRATEGIES

Children with PWS frequently present with an apparently advanced level of vocabulary and conversational skills, and thereby appear cognitively very able. This can have a significant disadvantage of raising expectations to a level that then results in high degrees of stress and failure (Whittington and Holland, 2004), and in general academic performance is poor for cognitive ability (Milner, Craig, Thompson, Veltmann, Thomas, Roberts, Bellamy, Curran, Sporikou and Bolton, 2005). Children with PWS sometimes have significant behavioural problems such as stealing food or money, immature social behaviour, temper tantrums, and obsessive-compulsive behaviours. Because of these behaviours, they may find mainstream school very demanding and such children may be placed in special schools despite the fact they may have the ability to cope with the academic demands of mainstream schooling (Whittington and Holland).

As children with PWS tire easily, it is often better to introduce new ideas early in the day when the child is more receptive (Prader-Willi Syndrome Association, United Kingdom (PWSA, UK), 2006). Formal social skills training is invaluable because most individuals with PWS have difficulty reading social cues and maintaining friendships with peers. Social skill instruction should include teaching the child not to interrupt conversations, standing at an appropriate distance from the person they are talking to, and sharing and turn taking, as these skills all tend to be poorly developed in people with PWS (PWSA, UK). Social skills resources suitable for children with high functioning autism spectrum disorders may be helpful for children with PWS. It is important that the child should be prepared in advance for changes in routine or planned activities (Cassidy and McCandless, 2005). The use of visual schedules and timers may be particularly useful to give the children plenty of warning that activities will soon finish or to indicate that a transition is coming up. Anticipatory planning is essential especially for non-routine events or situations in the school day.

It may be best to avoid using open-ended questions as they can lead to difficulties in decision making for children with PWS (PWSA, UK). The use of closed questions with only 2-3 alternatives may be preferable. Rules, regulations, and procedures must

be spelled out, written down, and prominently displayed in the classroom. The use of question cards to limit the number of questions asked may also be helpful with regard to repetitive questioning. Limiting or rationing the time a child can talk about a particular topic may be necessary if the child is inclined to fixate or obsess about certain topics. Visual cues and aids to learning should be used as much as possible to capitalise on relatively strong simultaneous and visual spatial skills. Teachers should be aware that visual materials such as photos, illustrations, computers, and videos are highly motivating and useful teaching aids for most children with PWS (Chedd, Levine and Wharton, 2006). The children also display better performance on long-term memory tasks on tests of cognitive ability, so new learning tasks should be introduced slowly and linked to concepts with which the children are already familiar.

In the area of mathematics, concepts should be taught in a concrete and visual manner and always linked to real-life activities. Given that the deficit in mathematics appears to be related to short-term memory deficits, specific training in improving short-term auditory memory may improve mathematical abilities and support the ability to do tables and other arithmetical tasks (Bertella, Girelli, Grugni, Marchi, Molinari and Semenza, 2005). In terms of reading, the visual and simultaneous processing strengths displayed by children with PWS mean that sight word approaches are more likely to be productive than phonics based approaches.

Behaviour Management

Provision of clear behavioural expectations and limits, beginning at an early age, is an extremely important part of behaviour management for children with PWS. Applying consistent limits at school and home is critical. Preparing parents very early in the life of the affected individual to be able to set and enforce limits consistently, appears to be a good predictor of behaviour problems in the future (Cassidy and McCandless, 2005). Successful behaviour management primarily requires a consistent supportive environment. Clearly stated rules regarding acceptable and unacceptable behaviour along with specified consequences that are consistently enforced at home and at school, and schedules that are as much as possible invariant in time and task routines, are prerequisites for reducing the occurrence of challenging behaviours (Whitman and Jackson, 2006). It is also important to have strategies for promoting positive behaviour, including attending to appropriate behaviour (with verbal praise or concrete rewards) on a regular and consistent basis while ignoring inappropriate behaviour. Table 2 outlines common behavioural difficulties in children with PWS and possible interventions for dealing with them:

Table 2: Common Behavioural Difficulties Displayed by Children with PWS and Possible Interventions to Limit Display of Difficulties

Behavioural Difficulties	Possible Interventions
Difficulty with flexibility and change and a strong need for routine	● Organise weekly and daily schedules of activities (visual schedules of the child performing the activity) ● Provide praise for flexible behaviour and responses ● Negotiate solutions to resolve stubborn issues

Obsessive thinking and repetitive questioning	● Provide limits (I will tell you one more time and after that we have to move on, or use of question cards) ● Refer child to visual schedule or timetable ● Distract child to another activity
Frustration leading to tantrums – yelling swearing, aggression, destruction or self-injury	● Avoid environments that might be over stimulating (assembly hall) or allow child to come to class earlier or later than other children ● Set behavioural expectations for the child telling them what you expect during the day ● Include student in behavioural contract ● Create a safe area in the classroom or outside the classroom where student can go or be brought to, if behaviour becomes very challenging. ● Have a plan in place whereby all staff know what to do if behaviour becomes very challenging ● Limit verbal interaction with child and establish a routine of not arguing stating "this discussion is now over"
Poor stamina	● Get child up and moving (movement breaks, doing school messages) ● Schedule exercise activity after lunch
Scratching and skin picking	● Provide distracting activities – keep hands busy with colouring, computers ● Keep nails short and apply skin lotion liberally to keep skin slippery and moist ● Praise for not picking/scratching ● Cover area with bandage or other covering
Poor social skills	● Provide opportunities to learn in small groups as child may learn better and display more appropriate social skills in small groups as opposed to whole class situations ● Provide social skill classes that emphasise sharing and taking turns

Adapted from Whitman and Jackson (2006).

Dietary Management

A low calorie well-balanced diet combined with a regular exercise programme and close supervision to minimise access to food should be instituted early in the child's life (Cassidy and McCandless, 2005). According to Whitman and Jackson (2006) effective dietary management of PWS requires the following elements:

- A physical environment structured so that access to food and money is completely eliminated
- An appropriate dietary and exercise plan: Non-competitive activities, such as swimming, walking, jumping on a trampoline, can be good alternatives to team sports
- A procedure for ensuring that the affected person is always informed about the time and menu for the next meal or snack
- Elimination of all other avenues for obtaining food
- Education for all family members, neighbours, friends and school staff regarding the restricted dietary needs
- Supervision in all food related areas

- Home-school communication around dietary issues is vital
- Avoidance of the use of food as a reward or incentive.

Nutritional needs and diet management goals can be incorporated into an individual education plan.

THERAPEUTIC INTERVENTIONS

The complexity of needs of children with PWS has led to the recognition that the skills of a range of different professionals are required in supporting affected children. Professionals on a multidisciplinary team supporting these children are likely to include dieticians, paediatricians, psychiatrists, and psychologists to help advise on the prevention of obesity and on the other maladaptive behaviours. Occupational therapists and physiotherapists can provide advice on physical development and exercise, and speech and language therapists can support communication development. Educational psychologists and educators can provide support in assessing and ameliorating the consequences of specific and general learning difficulties.

CONCLUSION

Educators of children with PWS need an awareness of the complexity of characteristics associated with the condition. As well as the prominent eating and food related behaviours there is an increasing understanding of the learning strengths and needs of affected children. Many of the children will also present with behavioural difficulties which if not appropriately addressed can result in significant difficulties in the classroom. In order to reach their potential in educational settings children with the syndrome need the support and understanding of a range of health and education professionals. Without such support the participation and inclusion of the children in educational settings will be put at grave risk.

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