

Information for Local Authorities when considering whether to assess a child/young person with Prader-Willi Syndrome for an Education Health and Care Plan

Legal test for an EHC Plan

The tests that an LA must apply when considering assessment are contained in Section 36(8) of the Children and Families Acts 2014; there are two parts:

- Part one of the test is that the child/young person has or may have special educational needs
- Part two of the test is that it may be necessary for special educational provision to be made for the child/young person through the issuing of an EHC Plan

The Prader-Willi Syndrome Association UK's position is that every child/young person with PWS will need an assessment for an EHC plan to establish what provisions must be in place to enable them equitable access to an education.

Part One: The child/young person has or may have special educational needs

Prader-Willi Syndrome (PWS) is a rare, complex, genetic condition caused by an abnormality of chromosome 15, which causes:

- Hyperphagia - Excessive uncontrollable appetite
- Hypotonia - Low muscle tone leading to mobility delays
- Hypogonadism - Immature sexual development
- Learning disabilities and some autistic tendencies
- Immature emotional and social development
- Challenging Behaviours
- Speech and language difficulties

Some children/young people with PWS will have a dual diagnosis of disorders such as:

- Autism spectrum disorder
- Attention deficit disorder
- Dyspraxia

amongst others, less common. Children/young people with PWS may also exhibit characteristics of these disorders without a formal diagnosis.

Children with PWS will require input from

- Dietician
- Endocrinologist
- Geneticist
- Occupational Therapist
- Ophthalmologist
- Orthopaedic nurse or surgeon
- Orthoptist
- Orthotist
- Paediatrician
- Physiotherapist
- Speech therapist

PWS has a particular group of behaviours associated with it; this is called a phenotype, and includes:

- Food seeking/stealing
- Outbursts of rage or prolonged crying and other forms of challenging behaviour
- Obsessive and/or compulsive behaviour – repetitive questioning about the same topic (perseveration) often with regard to food, but can also be other topics or people
- Stubbornness/Difficulty in adapting to change in routine
- Autistic tendencies
- Argumentative or oppositional behaviour
- Manipulative behaviour
- Hoarding, collecting and non-food theft
- “Lying” and blame-shifting and story-telling
- Skin picking

Challenging and obsessive behaviours are a feature of PWS, caused both by the Hyperphagia and by the immature emotional and social development. Children/young people with PWS have reduced cognitive abilities and will usually present as abler than they are, and will need support throughout their life.

In addition to these behaviours, there are additional medical issues that will impact on a child's care within an educational setting;

- Sleep apnoea resulting in abnormal daytime sleepiness
- Body temperature abnormalities potentially resulting in both overheating and being too cold
- Lack of vomit reflex masking gastric issues and the stomach not rejecting inappropriately ingested items
- High pain threshold resulting in reduced awareness of injury
- A tendency to skin picking and bruising

Part two: It may be necessary for special educational provision to be made for the child/young person through the issuing of an EHC Plan

It is important that a child/young person with PWS receives adequate and appropriate support within the school setting in order to access the curriculum, develop key skills and make social gains.

Children with PWS have an overwhelming interest in food and have an insatiable, compulsive hunger for most of the time and this is not usually lessened by eating. This Hyperphagia, coupled with Hypotonia (low muscle tone) and unusual metabolism, makes it very difficult for a child/young with PWS to maintain a healthy weight. They will put on weight very easily and very quickly, and need, on average, 60% of the calories of other people. This means that controlled and restricted access to food is extremely important.

Hyperphagia, if not rigidly managed, results in anxiety, distraction, frustration and irritability, leading to challenging behaviour. Environmental changes will need to be made within the school setting to accommodate a child/young person with PWS. Food security is essential; kitchens and cupboards containing food must be locked, and snacks and food always kept out of sight. A child/young person with PWS will find a way to access extra food, sometimes through stealing or from their peers, and this makes them very vulnerable as overeating or bingeing can lead to stomach rupture with potentially fatal consequences.

A child/young person with PWS will almost always need one to one support for most parts of the day, and in particular during meal or snack times. Staff within the chosen school will need:

- An understanding of PWS and the challenging behaviours that can occur
- To learn strategies that will enable them to manage behaviours related to food
- To learn strategies that will enable them to manage behaviours not related to food
- To provide a safe and structured environment
- To differentiate teaching methods to allow for reduced cognitive ability and allow extra time to process information
- To give clear, simple instructions, reinforced where appropriate with pictorial diagrams
- To be aware of and manage sensory issues that may overwhelm the child/young person
- Provide additional support when moving around school and participating in physical activities
- To provide a balance between physical and quiet activities and allow for additional rest breaks
- To assist with toileting needs if and when necessary

Life with a child with PWS is very difficult and challenging and getting the right support to maximise their life chances is vital to both the person with PWS and the family.

For further information visit www.pwsa.co.uk

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