PWSA UK
PRADER-WILLI SYNDROME ASSOCIATION UK

Speech and language in PWS 5—13 years

Introduction

Children with PWS may have the same range of disorders of speech and language development that can occur in any child.

There is also the possibility of additional problems associated with the syndrome.

These may be caused by:

- Learning disabilities e.g. retrieval of information and memory deficits.
- **Physiological** characteristics e.g. high arched palate causing inaccurate articulation.
- Low muscle tone making it difficult to make the fast accurate movements required for clear speech.

Most people with the syndrome do have some difficulty with speech and/or language at some time during their lives.

NOTE:
Words in orange are explained in the glossary at the end of this leaflet.

Difficulties associated with PWS

Articulation

Inaccuracy of movements and inability to change quickly from one tongue or lip position to another (sometimes described as **dysarthria**) are often features of PWS. This is caused by the combination of the characteristic high arched palate and the small lower jaw. **Hypotonia** in the oral muscles can also contribute.

More rarely **dyspraxia** occurs, although it is now thought that dyspraxia is more common in children with PWS than in the general population. This condition can lead to extremely poor intelligibility and may mean that an augmentative method of communication is needed e.g. a communication book or Voice output device.

The role of the speech and language therapist

The Speech and Language Therapist's (SLT) role is to assess and diagnose which, if any, speech and language, communication or feeding difficulty the child is presenting with.

This may not be because of PWS. If any disorder is found the SLT will advise on the best way to enable the child to reach their maximum communication potential. S/he will liaise with parents, teachers, carers, nursery staff and anyone else who is a regular part of the child's environment.

Language delay

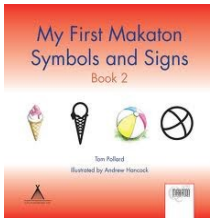
Language delay will often occur, with **verbal comprehension** being better than **expressive language.** This can lead to extreme frustration and exacerbate temper episodes if the child knows what they want but cannot put it into the right words.

Early stimulation at the appropriate level for the child's understanding is essential for him to maximise skills in this area, and the use of signing such as Makaton (see **Useful Agencies and Websites** https://www.pwsa.co.uk/assets/files/useful-agencies-children.pdf) for details) to aid development of comprehension is often a valuable technique suggested by SLTs. The discrepancy between receptive and expressive language can cause frustration at school if the child has learnt something but cannot demonstrate their knowledge by answering questions.

Fluency

Most non-PWS children experience a stage of non-fluency, usually between the ages of 3 and 5 but for the child with PWS it can be later occurring and last longer. Occasionally it can develop into a true stammer, but this would probably have been present if the child had not had PWS as there is a strong genetic component in the incidence of stammering within families. Some children with PWS are unusually **fluent.**

Language disorder



More rarely a specific language learning difficulty exists. The child may not be able to understand language adequately, leading to confusion, frustration and difficulty learning. This may be mistaken for a learning disability, but only collaboration between an SLT and an educational psychologist can determine the origin of the problem.

Some children may experience difficulty with expressive language, with their verbal comprehension more intact, this is similar to a language delay, but more pervasive and persistent in nature, so that the ability to express ideas and comment on events does not develop along expected lines.

Pragmatic Disorder

Pragmatic skills concern the child's ability to use language appropriately in social situations; this can cause a significant barrier to learning if not dealt with. Problems in development of these skills are less easy to spot and to diagnose, and may be thought to be behavioural.

The children will talk a lot, but the language is inappropriate and social interaction, conversation and turn-taking skills are affected, making it harder to form relationships and friend-ships. These children are often not referred to SLT service because they can speak, although SLTs are the appropriate professionals to give advice and support in this situation.

Autistic Spectrum Disorder

A small number of children with PWS may also have an Autistic Spectrum Disorder (ASD) and it is easy miss this because of the difficulty or delay in social skills development. We have also heard of a few children with an ASD diagnosis where it was not realised that their difficulties with social skills were because of a delay in this area of development due to PWS, not ASD.

Repetitive use of language (Perseveration)

This is the term used for the habitual discussion of one topic, often associated with an obsession with food. It may take the form of the constant repetition of a question, even after an answer has been given several times. This is extremely difficult for people who are not familiar with the child to understand, and has lead to situations where the child is punished or ignored because of it.

Difficulties of direct (hands on) therapy

The SLT supporting your child may advise that direct therapy is not the way forward. This will be decided on your child's clinical needs. Children who are resistant to therapy, as is the case with many children with PWS, will become distressed and not achieve as much as they could. There may be a need for constant and prolonged repetition of work for learning to take place; this cannot be achieved in weekly therapy sessions.

Children may not generalise what they learn in a specific situation, so that the trend now is for development activities to be devised which can be built into the child's daily routine, or carried out within his school curriculum, rather than specific 'therapy sessions' taking place.

Rewards for carrying out therapy activities must be carefully prepared according to the child's interest, and must not be food-based. Suggestions could be playing with a favourite toy or a colouring sheet.

How to access SLT services

Every child with difficulties in the following skills should be assessed by an SLT:

- Eating and drinking (if it is suspected there is a physical cause)
- Saliva control (excessive salivation for the child's age)
- Speech (if not intelligible to the adults in the child's environment)
- Language development (if not in line with the child's other development, or to establish whether this is the case)
- Pragmatic skills or any other problem that is causing difficulty with communication

Most Health Trusts operate an open referral system, so that parents can request that an SLT give advice about their child by telephoning the local SLT department. If the child has a Statement of Need or co-ordinated support plan and SLT is specified, it is a legal requirement for the Education Authority to ensure that this is provided.

Speech and Language Therapy (SLT)

Service Delivery may include:

- **Consultation and advice about how people can support the child's communication needs.**
- **Teaching parents/ carers to work with their child and giving specific work for them to do.**
- **Advice to others involved with the child to maximise communication opportunities in different environments e.g. home, nursery, school.**
- **Assessment and explanation of the result, which includes the impact of any difficulty the child may have.**
- **Hands on therapy with the child, probably with backup practice for home (the therapy programme may be carried out by an SLT support worker under the guidance of a qualified SLT).**
- **Re-assessment of the impact for the child.**
- **Speech and language therapists work in episodes of care so the child may move in and out of the service as their needs change.**

Remember!

Not all children with PWS will have all or any of these problems, and some of them might have been there if the individuals had not had PWS.

Glossary

Articulation Production of meaningful speech sounds by co -ordinated movement of tongue, lips, teeth palate and jaw.

Clinical Relating to treatment of a medical condition.

Dysarthria Inability to make accurate rapid speech movements. Generally such speech is slow, with indistinct consonants and long intervals between words. Overall effect is of slurred or indistinct speech.

Dyspraxia Disability of motor programming of articulatory movements or lack of voluntary control over the muscles needed for speech, neurological in origin. Characterised by difficulty in imitating words or repeating them accurately and/or poor sequencing of sounds in words, and/or sentences.

Expression The ability to use language in a way meaningful to the listener.

Fluency The flow of speech.

Hypotonia Lack of tension and strength in the muscles.

Oral Relating to the mouth.

Physiological Physical characteristics (e.g. shape of lips, tongue, palate).

Signing The use of a sign language system e.g. Makaton.

Verbal comprehension The ability to understand the spoken word.

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