Worster-Drought Syndrome

This information sheet explains about Worster-Drought Syndrome and what it could mean for your child. It also gives suggestions for how you can ensure the best treatment and support for your child.

What is Worster-Drought syndrome?

Worster-Drought syndrome (WDS) is a type of cerebral palsy (movement disorder) that affects the muscles around the mouth and throat. This causes problems with swallowing, feeding, talking, dribbling and other actions controlled by these muscles such as coughing, nose-blowing, kissing and crying.

Children with WDS often have additional difficulties with co-ordination, learning, behaviour and epilepsy.

What causes WDS?

WDS is thought to be due to abnormality in the part of the brain which normally controls the mouth and throat muscles (perisylvian area). This abnormality appears to occur in the 12 to 16 week period of pregnancy. In around 15 in 100 children with WDS, this abnormality is visible on a brain scan as bilateral perisylvian polymicrogyria. However there is not always a close relationship between the scan appearances and the difficulties the children experience.

There appear to be several causes of WDS, such as problems with blood supply to the perisylvian area or genetic vulnerability, all of which act in early pregnancy. In around 15 out of 100 cases, several family members are affected suggesting a genetic cause and this is true for cases with or without scan abnormalities. WDS is not due to injury around the time of birth, although babies with WDS often have so much difficulty with swallowing and feeding that they need to go to a special care baby unit after birth.

How common is WDS?

There have been no large studies of WDS. One cerebral palsy study suggested it occurs in around 3 in 100,000 live births. In our experience, this is probably an underestimate as many children are diagnosed late (often older than five years) or looked after without ever receiving a diagnosis.

How is WDS diagnosed?

WDS is diagnosed on the medical interview and examination. It may be suspected in very young children, but diagnosis is not always certain until the child is older than four years, as some younger children have co-ordination problems that look similar to WDS, but which improve as they mature.

What are the main features?

WDS is very varied in how it affects people, both in extent and severity. Even two people from the same family will not necessarily be similar to each other. The main features are listed below, and then discussed in more detail.

Essential Features:
Children have obvious difficulty in moving the tongue, lips, jaw and palate that is present from birth, so they have problems with swallowing, feeding, speech, dribbling, coughing and so on. The muscles involved may seem stiff and slow, but are not wasted and there will be no obvious structural reasons such as tongue tie. Most children will have a brisk jaw jerk when examined by a doctor.

Common additional features:

- Mild difficulty with movements (for example, late in walking, clumsy)
- Moderate learning difficulties
- Behavioural difficulties (such as short attention, problems with social interaction)
- Epilepsy
- Club feet and restricted movement at joints
- Abnormality on brain scan (congenital bilateral perisylvian polymicrogyria)
- Consequences of the mouth and throat problems (for instance glue ear, chest infections, gastro-oesophageal reflux)

Swallowing, dribbling and feeding:
Swallowing may be disorganised, with the risk that food, drink, or even saliva spills into the lungs (aspiration). Symptoms to look out for include frequent chestiness, coughing/choking or colour changes during feeding and/or constant wheeziness. There may also be gastro-oesophageal reflux, where the stomach contents move back up the food pipe and into the mouth. This can be painful and is another way that food can spill over into the lungs.

Breast or bottle feeding may be slow, and there may be a delay in progressing to more solid food as there are often problems with chewing and moving food through the mouth. There is a distinct improvement in feeding abilities for most children with time, although many will always need a modified diet (for example, avoiding particular textures such as apple skins or crisps). Older children usually still have feeding difficulties, but appear to learn to put up with them or find unusual ways to overcome them, such as using their fingers rather than their tongue to move food around the mouth.

Around a quarter of children will need tube feeds at some stage as their swallowing is unsafe (risk of aspiration), most commonly as infants. However around 1 in 10 will need long term tube feeding, usually by gastrostomy (tube directly through the skin into the stomach). It is important for all children to have their height and weight checked regularly to monitor their growth. Many children will have problems with constipation, poor fluid intake and food aversion.

Most children have problems with drooling because of their difficulties with swallowing and with keeping their lips closed. This may improve with age, but equally may cause increasing distress as it is socially isolating, interferes with school work and using computers, and

leaves chronically irritated skin. Some children benefit from simple positioning and behavioural techniques, others use medication and some have surgery. Specialist assessment may be needed, as interventions need to be planned to minimise side-effects on dental hygiene or swallowing.

Some children with WDS have rather impassive faces, however many have normal emotional facial expressions but cannot voluntarily or on command make the same facial movements. This is because the brain controls voluntary and involuntary (that is, in response to emotions) movements separately and in WDS it is particularly the voluntary control that is affected.

Mouth:
Because the muscles around the mouth do not work properly, the jaw and palate may have an unusual shape and the teeth are often crooked. Sometimes children have a jaw contracture, which means they cannot open their mouth fully, making eating and teeth cleaning very difficult. Tooth decay is particularly common in children who are tube fed or who have reduced saliva due to medication or surgery.

Speech, language and learning:
All children with WDS understand more language than they are able to speak. They will often be unusually quiet babies who do not babble, and develop speech late, often using gesture and pointing to communicate instead. When they do start to speak, they have enormous difficulty pronouncing words so that even family members may not be able to understand them.

In our experience, children with WDS continue to have severe difficulty with talking as they grow older. They often do not benefit from therapy directed specifically at pronunciation. It is important to provide them with additional and alternative ways of expressing themselves from an early age, such as gestures, sign language, symbols and communication aids. Often this will need specialist assessment and provision.

Speech difficulty may be made worse by hearing problems caused by the Eustachian tubes (drainage tubes) in the ear not draining properly because of the muscle problems around the throat. This causes glue ear or middle ear infections and may need treatment.

Many children with WDS also have some general learning difficulties, including difficulty understanding language. The earlier these difficulties are noticed the better, as early intervention can help reduce their impact and allow you to organise any special help needed at nursery or school. About one third of children with WDS have mild learning difficulties needing some help in mainstream school. Another one third may need more intensive support but the remaining one third have no significant learning difficulties at all.

Behaviour and social aspects:
At least 4 in 10 children with WDS have behavioural difficulties. Poor concentration skills and hyperactivity are particularly common and need to be addressed as they can make it difficult for the child to settle and learn in school. It is important to undertake a careful analysis of any difficulties as they may require

specific treatment, behavioural and educational approaches.

Some children with WDS struggle in their interactions with other children and adults. Sometimes it is because of the problems they experience with expressing themselves and finding a group of friends similar to themselves with whom they can feel comfortable. These are important reasons why providing early communication support is so vital.

Other children have such difficulty with social interaction that they fall within the autistic spectrum and require specialist education.

Epilepsy:
Around one-third of children with WDS have seizures, and this is more common if children have a visible abnormality on brain scan. Usually the seizures can be controlled with medication, however sometimes they can be difficult to treat, interfere with learning and need referral for specialist advice.

Motor abilities:
Children with WDS are often late in reaching motor milestones. They commonly have co-ordination problems and find it difficult to perform tasks that require dexterity and careful control of movement. This will affect their ability to use sign language and also communication aids. Some children are born with club foot (talipes) or other joint contractures, which will need specialist management.

How to ensure the best for your child

Most children with WDS are first known to speech and language therapists because of the early feeding and speech problems. These therapists are well placed to identify the unusual features in WDS and to refer the children for further evaluation by either neurodisability or neurology services. Once the diagnosis is made, the local team should take responsibility for arranging comprehensive assessment and management of the child, including monitoring for complications. Because of the multi-faceted nature of difficulties, a multidisciplinary approach is essential. It will commonly include:

Feeding:
- Observation of feeding and safety of swallow
- Height and weight, indicators of nutrition
- Where indicated: videofluoroscopy, pH study, chest X-ray, dietary modification, tube feeds

Speech and language:
- Early provision of alternative and augmentative communication channels to promote communication
- Hearing
- Epilepsy
- Development and learning, including educational support
- Behavioural screening
- Motor skills
- Genetic advice
- Support for the family

Children will often have a brain scan to look for a visible abnormality to explain their difficulties, but this is not essential for diagnosis and can be performed at a later date.

Children with WDS are all very different. Some show a wide range of problems, while others appear to have isolated feeding and speech difficulties. Careful assessment is therefore extremely important to identify each child's personal strengths and weaknesses and to ensure that their individual needs are met. Co-ordination between schools and health services is essential to deliver the right support.

Medical terms associated with WDS

Pseudobulbar and suprabulbar palsy:
These terms are interchangeable and simply describe the neurological level of the problem (in the cerebral cortex or cortical tracts). In WDS the term 'congenital' is normally added, as the problem is present from birth. Thus WDS may also be referred to as congenital pseudobulbar palsy or congenital suprabulbar palsy.

Perisylvian:
This is the area of the brain's surface that controls the muscles of the mouth and throat. It is present on both sides of the brain.

Opercular syndrome:
Operculum is another name for the perisylvian region, and was the name originally used for what is now known as congenital bilateral perisylvian polymicrogyria.

Congenital bilateral perisylvian polymicrogyria (CBPP):
In CBPP there is an abnormality in brain development that appears to occur between 12 and 16 weeks of pregnancy. This causes the brain surface in the perisylvian area to have an excessive number of small folds and means that the region, which normally controls the muscles of the mouth and throat, may not work properly. Typically children with CBPP have difficulty with feeding and speaking, and often also have epilepsy. However there is not always a good relationship between the visible imaging abnormalities and the difficulties the children experience. Indeed some children with CBPP do not have many difficulties. If children with CBPP have difficulties with eating and speaking, they fit the WDS group and benefit from the same approaches and management as other children with WDS.

Further information

GOSH switchboard – 020 7405 9200

Neurodisability department - ext 5293

Family Support Group
The Worster Drought Syndrome Support group is a UK based charity that supports children and adults with WDS. As well as supporting those with WDS, the group also supports anyone else affected by WDS and offers information and support to parents, carers and professionals working with children that have WDS

The Worster Drought Syndrome Support group offers regular newsletters and family days allowing people to meet up and talk about their experiences of WDS.

For more information about the support group please go to. www.wdssg.org.uk

Newly diagnosed families - please email national.contact@wdssg.org.uk

Useful contacts:

ACE Centre Advisory Trust
The ACE (Aiding Communication in Education) Centre Advisory Trust carries out assessments and provides information in the use of technology as an aid to communication.
Website: www.ace-centre.org.uk

AFASIC
UK charity representing children and young adults with communication impairments working for their inclusion in society and supporting parents and carers
Helpline: 0845 355 5577
Website: www.afasic.org.uk
Email: info@afasic.org.uk

Communication Matters
National voluntary organisation of members concerned with augmentative and alternative communication (AAC) needs of people with complex communication needs.
Website: www.communicationmatters.org.uk

Contact a Family
Contact a Family provides support, advice and information for families with disabled children, no matter what their condition or disability.
Helpline 0808 808 3555 or
Textphone 0808 808 3556
Website: www.cafamily.org.uk

Department of Education and Employment (DfEE) Publications Centre
Tel: 0870 000 2288

Half PINNT (Patients on Parenteral or Enteral Feeding)
This is the children's section of PINNT, providing help and information on alternative feeding methods
Website: www.pinnt.com

Hyperactive Children's Support Group
Tel: 01243 539966
Website: www.hacsg.org.uk
Email : hacsg@hacsg.org.uk

I CAN
A charity working for children with speech and language difficulties, including special schools for such children.
Website: www.ican.org.uk

Independent Panel for Special Education Advice (IPSEA)
Provides advice and information to parents whose children have special educational needs. Professional advice for parents appealing to SEN tribunal
Helpline: 0800 0184 016
Website: www.ipsea.org.uk
Email: ipsea.info@intamail.com

KIDS
KIDS is a charity that works to create an inclusive world for disabled children, young people and their families. Range of services provided for children with disabilities including home based learning, respite care, holiday play schemes and independent educational advisory service
Website: www.kids.org.uk

Makaton
Makaton is a system that uses signs and symbols to teach communication, language and literacy skills to people with communication and learning difficulties
Website: www.makaton.org

MENCAP
Mencap is the leading UK charity for people with a learning disability and their families.
Tel: 020 7454 0454
Website: www.mencap.org.uk
Email: information@mencap.org.uk

Network 81
National network of parents working towards properly resourced inclusive education for children with special needs.
Website: www.network81.org

PECS
Picture exchange communication system

Website: www.pecs.org.uk

Rare Diseases UK
Rare Disease UK aims to develop strategic planning for rare diseases in the UK

Website: www.raredisease.org.uk

Rathbone
Rathbone is a UK-wide voluntary youth sector organisation providing opportunities for young people to transform their life-circumstances by re-engaging with learning, discovering their ability to succeed and achieving progression to further

Website: www.rathboneuk.org

Skill (National Bureau for Students with Disability)
Provides information, advice and publications regarding post 16 education, training and employment for people with disability
Information service Tel: 0800 328 5050
Text : 0800 068 2422
Website: www.skill.org.uk
Email : info@skill.org.uk

Widgit
Widgit has pioneered the use of symbols in learning and communication, including software to create symbol-supported materials in print, onscreen and online
Website: www.widgit.com

Notes

Compiled by the Neurodisability Service in collaboration with the Child and Family Information Group

Great Ormond Street Hospital for Children NHS Trust
Great Ormond Street,
London WC1N 3JH

www.goshfamilies.nhs.uk www.childrenfirst.nhs.uk

