

THE
Worster-Drought
SYNDROME

A review for Parents of the
research carried out by The
Institute of Child Health and Great
Ormond Street Hospital
2000

Produced with the help and collaboration
of the members of the
Worster-Drought Support Group

Worster-Drought Syndrome

Support Group

Affiliated to Contact a Family and the Rare Disorders Alliance UK.

WORSTER-DROUGHT SYNDROME

By Dr. Maria Clark

WHAT IS WORSTER-DROUGHT SYNDROME (WDS)?

WDS is named after Dr. Worster-Drought who first described it in 1956. It is a form of cerebral palsy in which the main effect on movement, is on the control of muscles which normally move the lips, jaw, tongue, palate, back of the throat (pharynx) and upper gullet (oesophagus or food pipe). Any number of these areas can be affected to variable degrees, and the children may have problems with eating, drinking, swallowing, dribbling and/or speech.

Like many forms of cerebral palsy, the condition is complex and can be associated with difficulties in many areas (e.g. learning, behaviour, epilepsy). This means that the children can appear very different from each other, and often their main difficulties can be in the associated areas, rather than predominantly focused on the oral problems. Thus children with WDS can come to the attention of a variety of different specialists, (speech therapist, teacher, educational psychologist, G.P., paediatrician...). As the condition is not well known, it can take some time before the whole picture is recognised and WDS diagnosed, enabling the child to receive the multifaceted support they need.

HOW IS WDS DIAGNOSED?

WDS is a clinical diagnosis, which means that it is made on the basis of the history and examination. The core features are the difficulty with movements around the mouth and throat from an early age (usually below 2 years) and a brisk jaw jerk. For example, many children have difficulty in wagging the tongue from side to side, or even in and out.

Most children have difficulty in additional areas, as previously mentioned. There is no specific test, although there are a number of investigations that can be helpful, and may be arranged by the child's doctor.

WHAT CAUSES WDS?

WDS is thought to be due to an abnormality in the area of brain that is responsible for the sophisticated control of movement of the muscles around the mouth and throat. In some cases an abnormality can be seen in this area on a brain scan. This abnormality is thought to occur very early in the development of brain of the baby during the first third of pregnancy. There is usually no obvious cause in the pregnancy or birth. In a minority of children, there is a family history of WDS, although which gene is involved, or how it has an effect, is not understood.

From the range of difficulties encountered in children with WDS, it seems likely that there may be subtle changes in other areas of the brain would explain why these children are vulnerable to epilepsy, learning difficulties, slow motor development...etc. Given that the abnormality appears to occur very early in the formation of the brain, and that this process is very complex and vulnerable, this is not too surprising.

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Common areas of difficulty

Feeding and associated areas

Difficulties may start as a baby, with difficulty sucking that may lead to choking and inhalation (*aspiration*). In severe cases, tube feeding (either through the nose or directly into the stomach using a gastrostomy) may be needed. In milder cases, problems may occur when solid foods are introduced. These include difficulties with lip closure and tongue mobility, so that food is not moved to the back of the mouth or cleared efficiently from the mouth cavity. Chewing and swallowing are often impaired which may lead to inhalation of food (*aspiration*). These problems may gradually improve over the first few years, or persist for many years.

Aspiration:

This is where food is inhaled into the lungs and this can lead to chest infections. Each chest infection can be very severe, and repeated aspiration can cause serious long term damage to the lungs. *Videofluoroscopy* is the test used to check that feeding is safe for a particular child. In severe cases, it may be suggested that a child is fed through a tube (either through the nose or directly into the stomach - *gastrostomy*).

Gastro-oesophageal reflux:

Food and acid from the stomach can pass back up the gullet (food pipe/oesophagus) and into the mouth. This can cause pain (especially around meal times and when lying down) and even vomiting. *Videofluoroscopy* or *Barium meal*, are both X-ray tests to look for this. A *pH study* is used to look for acid that has passed from the stomach, back into the gullet.

Reflux can be treated with drugs to reduce acid production, or by drugs that make the stomach empty more quickly. In severe cases, an operation (fundoplication which 'tightens' the entrance into the stomach) may be needed.

Nutrition:

Because of all the difficulties with feeding, children can be underweight which in itself can lead to ill-health by reducing energy and reserve to cope with infections. Advice from a dietician can be very important, especially for children who cannot eat solid food and need a special diet.

Dribbling:

Persistent dribbling is very common but may show steady improvement during childhood. For some children, speech therapy, drugs and occasionally operations are needed.

Ear and hearing problems:

Because these children have swallowing problems, the Eustacean tube (which usually ventilates the middle ear) often does not work properly. This means many children have problems with middle ear infections, glue ear and (conductive) deafness. It is particularly a problem for young children, and may need to be treated with grommets to help the ventilation of the ear.

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Dental care:

Sometimes there is an abnormal shape to the jaw and tooth alignment, so dental care is important. Some children have unusual teeth (e.g. fused teeth), and build-up of plaque can be a particular problem for those who cannot take food by mouth.

Speech and Language:

Difficulty with moving muscles of the mouth and throat, interferes with multiple aspects of voice production. In severe cases a baby may be unusually quiet and not produce normal babble. For other children, parents may notice that speech and early words are much delayed compared with the understanding of language, and that the children may spontaneously use gesture and pointing to express themselves. Even when the children do start to speak, pronunciation and articulation of words is very difficult for them, so it can be hard, even for familiar people to understand them. These difficulties mean that many children are referred to Speech and Language Therapists. However, at present it is not certain that traditional speech and language therapy is able to compensate for the weakness of the relevant muscles, and many children need alternative ways of expressing themselves (e.g. through sign language, symbol use or even voice out-put communication aids).

Motor development:

The arms and legs often show mild signs of spasticity and in-coordination. This means that children may be slightly late with sitting (e.g. around 1 year) and walking (e.g. around 2 years), and have clumsiness of the hands. Occupational therapy and physiotherapy may be helpful.

Learning and behaviour:

Many children have learning difficulties, which can be moderate or severe. Behavioural problems are also common, particularly hyperactivity. Carefully planned educational and behavioural programmes can be very helpful, and a minority of children may benefit from drugs to treat their hyperactivity.

Epilepsy:

A minority of children can develop epilepsy, which is treated with medicine in the usual way. Good treatment is very important as some children have a tendency to frequent fits, which can interrupt their awareness and attention span, so that they are less able to learn from new experiences and enjoy life.

Temperature (or febrile) fits are also more common in the younger children.

Minor birth defects:

A minority of children are born with unusual features such as small or tight jaws, club feet, limb contractures, or even excessive amounts of hair. These effects must have happened early in the baby's development in the womb, but are not fully understood.

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WHAT TESTS MAY BE DONE?

MRI: (Magnetic resonance imaging)-Brain scan:

This produces a very sophisticated image of the brain. The child has to lie inside a machine, which is like a big tunnel, and can be noisy. The machine uses a big magnet and radio waves to take a picture of the brain, a bit at a time. Then a computer creates the picture. It does not involve X-rays.

The scan takes quite a long time (up to 40 minutes) and so many children will need either sedation or a general anaesthetic to help them to lie still.

In some cases, children with WDS have changes on the scan in the areas which control the mouth and throat, and these changes usually consist of 'bilateral perisylvian polymicrogyria'. However, for most children, the scan appears normal.

CT (Computer Tomography) - Brain scan: This also produces a picture of the brain, but it is less detailed than an MRI. It uses X-rays, and is much quicker to perform.

EEG: (Electroencephalogram) - Brain wave record: The EEG is a special test, which records the electrical activity from the brain. It is particularly used to look for signs of fits. The child has wires stuck onto his/her head with special glue. These wires record electricity coming from the brain (simply recording the brain's normal activity). During the recording, the child will be asked to open and shut their eyes, and at one point to breathe deeply (or blow a windmill). They will also be asked to look at a flashing light. There may also be an attempt to record whilst the child is asleep. All of these activities, in some children may increase or reveal abnormalities, which can then help to guide their care.

EMG (Electromyogram) Muscle recording: The EMG uses a very fine needle to record activity from a muscle to help clarify whether a problem with movement of that muscle arises from the muscle itself or the messages, which should reach the muscle, from the nerves.

Videofluoroscopy and Barium swallow: In this test, the child swallows a mouthful of food or liquid containing a form of label or dye that shows up on X-rays. This allows the swallowing mechanisms to be examined in great detail, highlighting any difficulties the child has with eating and swallowing, and revealing if the child is at risk inhaling food onto the lungs (aspiration), or if there is a tendency for food to come back up into the gullet (gastro-oesophageal reflux or heart-burn).

pH study:

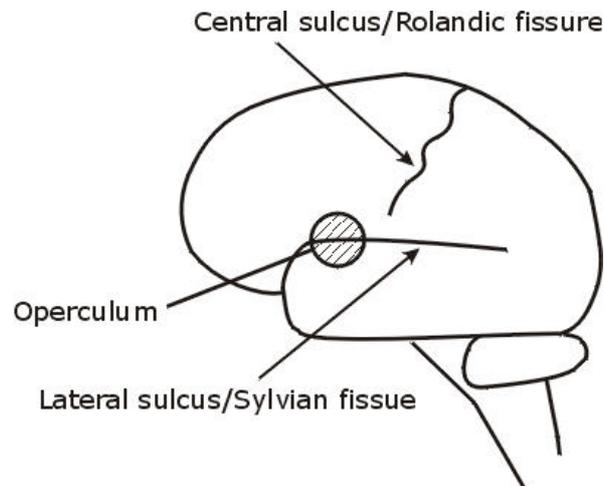
This test is used to check whether acid from the stomach 'refluxes' (i.e. goes back up into) the gullet. It is done by passing a very narrow wire through the nose, down the throat into the gullet. An X-ray is taken to check that the end of the wire is just above the opening to the stomach (the X-ray is also useful to look for any signs that food has been inhaled onto the lungs). The wire stays in position for 24 hours, and changes in acid level are monitored by a little box (about the size of a walk-man) that the child can wear. During this time, the child can eat, drink and move normally.

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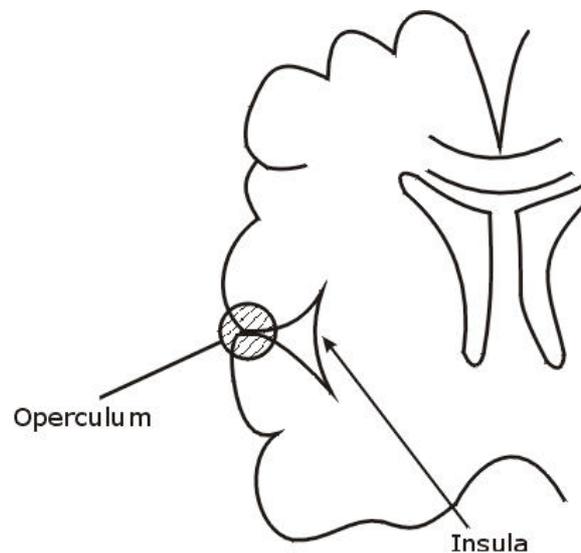
Blood tests:

There are no specific blood tests for WDS. However a doctor may ask for a blood test to check on conditions that superficially may look similar to WDS, or to look at related issues such as nutrition. In certain centres, blood may be taken to help with genetic research into WDS.

Anatomy



Cross Section



The sylvan fissure is deep and contains a “buried” area referred to as the insula. The anterior part of the insula is the operculum.

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ASSOCIATED MEDICAL TERMS

Pseudo-bulbar palsy:

This means that messages from high level, sophisticated brain cells that control the muscles of the mouth and throat (found in the perisylvian region of both cerebral hemispheres) are not reaching the muscles correctly so that the muscles do not move properly and appear weak. This can arise if either the relevant brain cells themselves do not work, or if the fibres (which carry messages) from them are interrupted as they descend through the brain.

Congenital bilateral perisylvian syndrome:

In this condition the child is born with an abnormality in the area of the brain that controls the mouth and throat, and this abnormality is visible on a brain scan. The abnormality affects both sides of the brain (bilateral) in the perisylvian region (this is the large fold that is easily seen on the outside of the brain). The abnormality usually consists of polymicrogyria. It seems likely that this condition is a subgroup of WDS, as there is considerable overlap between the 2 conditions. (NB only a minority of children with WDS have this visible abnormality on their scan)

Polymicrogyria:

‘poly’ – many
‘micro’ – small
‘gyria’ – folds

Here an abnormality has occurred very early in brain development of the unborn child. The abnormality is visible as an area in the cerebral hemispheres where the surface of the brain has very many small folds.

Opercular syndrome:

Operculum means ‘mouth’ and the term refers to the fold that is clearly visible on each side of the cerebral hemispheres. It is the same as the perisylvian region. The term was originally used for people who developed severe epilepsy and were found to have a visible abnormality in this area on their brain scan.

Foix-Chavany-Marie syndrome:

This term is usually used in relation to adults as it refers to damage that is acquired later in life, after the brain has been formed. There are visible bilateral perisylvian abnormalities on the brain scan, and it causes an unusual situation in which reflex or automatic movements of the muscles of the mouth and throat remain intact (such as swallowing or yawning). But movements that are voluntary or under conscious control (such as speech or following commands) are lost.

What does the future hold?

The condition is non-progressive, which means that the children are born with particular features about their brain which means that they have difficulty with movement of the mouth and throat, and possibly some of the additional features mentioned previously (although they may not show signs of them until they are older,

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such as speech delay). There is no tendency for them to acquire new problems with the brain.

Many of the features may have a tendency to improve with maturity (e.g. feeding, dribbling, behaviour).

Some areas are more problematic, and many children are likely to need life-long support with their speech.

Because WDS has only been recognised for a relatively short time, and is not well known, it is not easy to be precise about the future.

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Aims of the WDS Support Group:

- ✍✍ To offer support to other families affected by Worster-Drought syndrome
- ✍✍ To raise awareness of WDS with professional workers and other interested individuals
- ✍✍ To raise awareness of WDS Support Group
- ✍✍ To support and promote research into WDS.

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