Worster-Drought Syndrome

This condition is a form of cerebral palsy. The main problems occur with the mouth, tongue and swallowing muscles as a result of the bulbar muscles being affected. There are usually no obvious causes in the pregnancy or birth but some varieties are genetically determined. Because of the range of problems the diagnosis is often made quite late.

The signs and symptoms of Worster-Drought Syndrome include difficulties with voluntary lip, tongue and palate movements with a brisk jaw jerk. Sometimes there is an abnormal shape to the jaw and tooth alignment and, therefore, dental care is important. The arms and legs show signs of mild spasticity and incoordination common.

The first indication of the condition is when the baby has difficulties with feeding. Attempts at feeding may cause choking and inhalation which may lead to chest infections. In the severest cases sucking difficulties may necessitate tube feeding for several months.

In less severe cases the major problems 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 or hard palate. Chewing and swallowing are often impaired which may lead to inhalation of food into the respiratory tract. These problems may gradually improve over the first 2-3 years or persist for many years. The second major clinical problem is usually a severe speech delay which may even prevent all early speech.

Persistent dribbling is very common but may show steady improvement during childhood. In some cases speech therapy, medication and occasionally surgical intervention are sometimes required. As a result of the swallowing problems there is an increased incidence of middle ear infection and conductive hearing loss.

A mild delay in walking and running with clumsiness of the hands is common. Many children have mild learning and behaviour difficulties which may include hyperactivity.

In a minority of cases Epilepsy can occur which is treated with medication in the usual way.

The management of WDS is on the degree of feeding and speech problems. The condition is non-progressive.

by Brian Neville