SYNDROMES OF CHILDHOOD & ADOLESCENCE IN WHICH EPILEPSY MAY OCCUR

ANGELMAN'S SYNDROME AND EPILEPSY

This is a rare condition which is sometimes called the "happy puppet" syndrome, because children with Angelman's Syndrome may look and move in ways similar to a puppet controlled by a string. The children have epilepsy, learning difficulties, severe speech delay, jerky movements, tongue-thrusting, a characteristic facial appearance and a happy mood with sudden bursts of laughter. The condition is caused by a genetic factor – an abnormality on a chromosome (genetic material), which is inherited from the maternal line in most cases.

SYMPTOMS

Movement development such as sitting or walking can be delayed in children with Angelman's Syndrome. The odd behavioural features (cheerful mood and bursts of laughter) become more obvious after 2 or 3 years of age. The facial appearance also becomes more noticeable in the second year of life, with the development of a pointed chin, thin wide mouth and protruding tongue. Epileptic seizures usually start between 18 months and 2 years of age and may occur with high temperatures (fevers). However, most of the seizures occur without a fever. Eventually seizures will occur in 8 or 9 out of every 10 children who have Angelman's Syndrome. The types of seizures include myoclonic (jerks) seizures, atonic (drop attacks) seizures and tonic seizures in which the child's body stiffens, but generalised convulsive (tonic-clonic) seizures are less common. Children with Angelman's Syndrome often learn to walk, at a later age (between the ages of 2 and 5 years) and their walk is unsteady. There is no development of speech.

DIAGNOSIS

The diagnosis of Angelman's Syndrome is usually made on the basis of the facial appearance, behaviour and epileptic seizures of the children. The EEG (electroencephalogram) test which records the ongoing electrical activity in the brain may also be useful in helping to confirm the condition. Brain scans are not helpful in diagnosing Angelman's Syndrome. More recently a genetic abnormality has been found on one of the chromosomes (genetic material) in almost two thirds of children with Angelman's Syndrome. The abnormality can be looked for in a blood test, but it is not present in every child with the condition.

TREATMENT

In most children with Angelman's Syndrome, the epilepsy can be controlled with anti-epileptic drugs. Drugs that are commonly prescribed include Sodium Valporate (Epilim), Clonazepam (Rivotril) amd Carbamazepine (Tegretol, Tegretol Retard). Lamotrigine (Lamictal) may also be effective. Children also need a lot of help with communication.

PROGNOSIS

General health in Angelman's Syndrome is good and there are no major life-threatening illnesses. The epileptic seizures are frequent and may be difficult to treat with anti-epileptic drugs for the first few years, but the epilepsy usually improves with time. In many children the epileptic seizures disappear completely after the age of 10 or 12 years.