**Science Media Centre Fact Sheet**

**Ohtahara Syndrome**

What is it?
- A very rare neurological disorder affecting very young infants and characterised by seizures and delayed neurological and motor development
- It is one of the most severe and earliest forms of childhood epilepsy
- It was first described by Ohtahara and colleagues in 1976
- Also known as Early Infantile Epileptic Encephalopathy with suppression burst (EIEE)
- It affects newborns, normally within the first three months of life, although seizures can appear as early as ten days
- Figures on the prevalence of Ohtahara syndrome are not well known, although it's listed as a rare disease by the Office of Rare Diseases at the US National Institutes of Health
- It affects a small percentage of children with epilepsy; boys are thought to be slightly more affected than girls
- There have been no recorded incidences of inherited cases

What are the causes?
- Little is known about the causes, although most infants with the disorder have an underlying structural brain abnormality which may be caused either by an inherited genetic abnormality or by brain damage sustained before or at the time of birth

What are the symptoms?
- The main symptoms are tonic seizures (sudden stiffening of the body, particularly the extremities, caused by tightening of the muscles)
- Affected infants may also experience partial tonic seizures affecting only part of the body and, more rarely, myoclonic seizures (jerking movements caused by rapid alternate contracting and relaxing of the muscles)
- Seizures are primarily partial in the early stages, and may become increasingly generalised later on
- Most infants with the disorder also have significant brain underdevelopment, resulting in associated mental and physical disability

How is it diagnosed?
- By encephalogram (EEG) which detects abnormal electrical activity in the brain and reveals a characteristic pattern of activity known as "burst suppression"
Can it be treated?
- There is no cure. Antiepileptic drugs such as phenobarbitone can be used to control the seizures, but are generally not very effective as the seizures show resistance to medication. Corticosteroids can sometimes be helpful.
- In a small number of cases, infants with a clear structural abnormality in a defined area of the brain may undergo surgery to remove the affected area.

What is the prognosis?
- Ohtahara syndrome is a progressive disorder, meaning that seizures become more frequent and severe. Affected children make very little developmental progress and often die within two years; cause of death is often chest infection.
- Those that survive remain severely mentally handicapped and are fully dependent on others for care. They may also develop other epileptic conditions such as West syndrome or Lennox-Gestaut syndrome.

Sources / further information

http://www.epilepsy.org.uk/info/ohtahara.html
http://www.ohtaharasindrome.org/
http://news.bbc.co.uk/1/hi/health/7909564.stm
http://www.wrongdiagnosis.com/o/ohtahara_syndrome/intro.htm