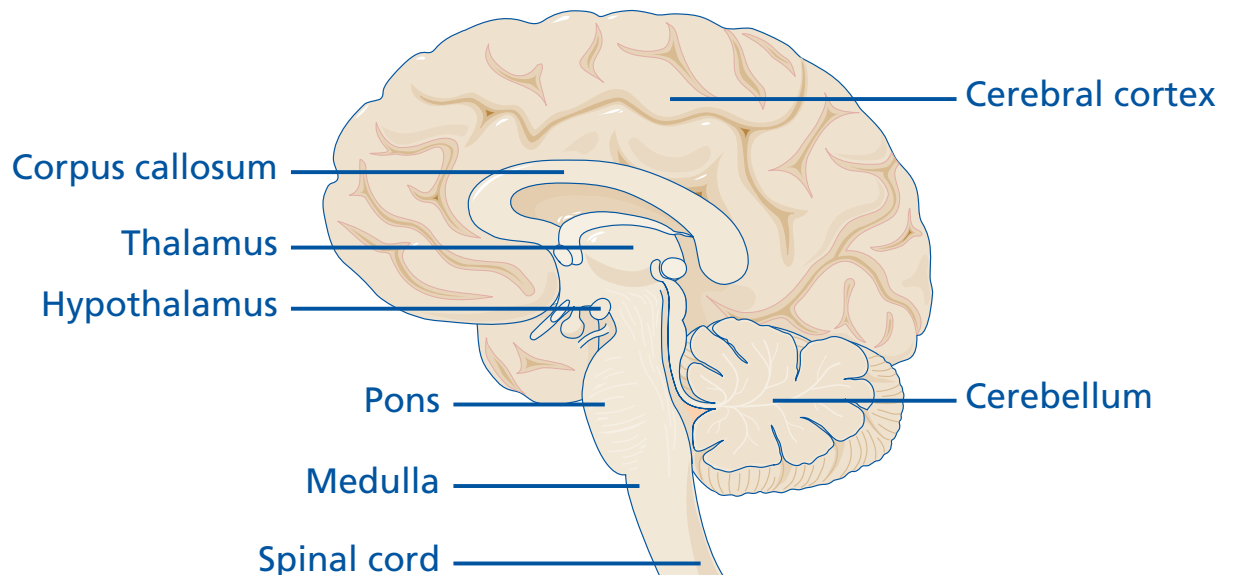




Great Ormond Street Hospital for Children NHS Foundation Trust: Information for Families

Opsoclonus Myoclonus Syndrome/ Dancing Eye syndrome (OMS/DES)

This information sheet from Great Ormond Street Hospital (GOSH) explains the causes, symptoms and treatment of Opsoclonus Myoclonus Syndrome/ Dancing Eye syndrome (OMS/DES) – also known as Kinsbourne syndrome – and where to get help.



What is opsoclonus myoclonus syndrome?

Opsoclonus-myoclonus syndrome (OMS), also known as dancing eye syndrome (DES), is a rare neurological condition which develops over days or weeks in early childhood. The key features are unsteadiness (ataxia), jerky movements of the trunk and limbs (myoclonus), rapid involuntary eye movements in all directions (opsoclonus) and usually marked irritability with sleep disturbance. The occurrence of marked irritability with behavioural change and sleep disturbance in an infant with new onset ataxia is one of the strongest clues that this may be OMS/DES. Most children develop the condition in the second or third year of life but rarely it may occur earlier or later. In children the condition

may occur with no clear trigger or it may follow a viral illness or it may be associated with a neuroblastoma.

What causes opsoclonus myoclonus syndrome?

Opsoclonus myoclonus is thought to be an autoimmune disorder although further research is needed to prove the actual mechanism of how the condition develops. Autoimmune disorders occur when the body mistakenly attacks itself rather than a foreign invader such as a bacterium or virus. It appears that in opsoclonus myoclonus syndrome an area of the brain called the cerebellum is attacked. The cerebellum is responsible for coordinating eye movements, muscle coordination and speech.



Around half of children diagnosed with opsoclonus myoclonus syndrome are found to have a neuroblastoma tumour. This is usually, but not always, a benign form of the tumour. Most children identified with a neuroblastoma will need to undergo staging and surgical resection, followed by monitoring by the neuro-oncology team. Treatment of the neuroblastoma, while important in its own right, does not appear to alter the outcome of opsoclonus-myoclonus syndrome. In children who do not have neuroblastoma, the condition may follow an infectious illness or no clear trigger may be found.

What are the signs and symptoms of opsoclonus myoclonus syndrome?

The symptoms of opsoclonus myoclonus syndrome are very variable and may not all be present at the same time. Typically opsoclonus myoclonus appears around the age of one to two years but can occur at any time in childhood.

Unsteadiness (ataxia) and muscle jerks affecting all or some areas of the body are usually the first symptom to appear. Muscle tone seems to decrease causing floppiness and lethargy (tiredness). Behavioural problems, irritability and sleep disturbance are also present. These symptoms tend to appear quite quickly, often leading to skills such as sitting and walking being lost in a matter of days. Speech difficulties also occur, for instance losing previously-fluent speech or not speaking at all. Rapid eye movements are one of the later symptoms to develop.

How is opsoclonus myoclonus syndrome diagnosed?

There is much discussion around how best to diagnose opsoclonus myoclonus syndrome and recently diagnostic criteria have been agreed. To be diagnosed with opsoclonus myoclonus syndrome, a child must have three of the following four conditions

1. rapid eye movement (opsoclonus or ocular flutter)
2. ataxia (unsteadiness) or jerky muscle spasms (myoclonus)
3. behavioural or sleep problems and
4. neuroblastoma.

Opsoclonus myoclonus can be mistaken for other similar appearing conditions, so diagnosis at a specialist centre is advised.

Affected children undergo extensive investigations including MRI brain scan and infection screening (blood tests and a lumbar puncture) to exclude the presence of alternative CNS disorders. Children are also examined for the possibility of an underlying neuroblastoma, including an MRI scan of the body, perhaps with a specific nuclear medicine scan such as an MIBG scan.

How is opsoclonus myoclonus syndrome treated?

The aims of treatment are firstly to treat the neuroblastoma tumour if present and secondly to damp down the immune system to reduce the attacks to the cerebellum and prevent any lasting damage.

- Neuroblastoma tumours in children with opsoclonus myoclonus syndrome are usually very small so are best removed using surgery. Occasionally chemotherapy or radiotherapy is needed before and afterwards, especially if the entire tumour cannot be removed during the operation.
- The immune system is modulated by using a combination of medicines. Steroids are given either by mouth or in 'pulses' intravenously (into a vein), sometimes alongside another



medicine such as rituximab. Rituximab removes some of the white blood cells in the body called B cells. Sometimes another medicine called cyclophosphamide is used instead of rituximab but this affects all types of blood cell. Intravenous immunoglobulin (IVIG), which damps down the immune system, may also be used. Treatment continues until the symptoms of opsoclonus myoclonus syndrome improve.

Who will be involved in your child's care?

There may be many different people involved at GOSH and these may change over the years. This may seem confusing, but different members of the team can help children and young people with opsoclonus myoclonus syndrome in different ways.

Consultant Paediatric Neurologist

These are doctors who have specialist knowledge of MS in children. They play an important role in diagnosing, treating and prescribing appropriate treatment. This is the person you will see each time you come to your appointment in the clinic.

Consultant Oncologist

This highly specialist doctor will treat your child's neuroblastoma before and after surgical removal with chemotherapy and/or radiotherapy.

Consultant Surgeon

These are the highly specialist doctors who will remove your child's neuroblastoma in an operation.

Clinical Nurse Specialist

Your clinical nurse specialist works closely with the neurologists. They have an in-depth knowledge of the condition and are usually your main contact person in between appointments.

Clinical Psychologists

Clinical Psychologists can support children, young people and their families in many aspects of care. Feelings of anger, anxiety, fear and sadness, for example, are common and understanding and managing these may be easier with the support of the Clinical Psychologist.

Clinical Neuropsychologists

We know that opsoclonus myoclonus affects a person's brain, and as a result there can sometimes be effects on a person's ability to concentrate, on their memory and on their learning. Neuropsychological testing will help identify specific strengths and weaknesses that your child may be experiencing. The Clinical Neuropsychologist can use the results of the testing to make recommendations to help at school. Even if there are no problems, it is very helpful for us to establish a baseline and re-evaluate progress every few years.

What happens next?

The outlook for children with opsoclonus myoclonus syndrome is variable depending on the severity of the symptoms when the condition is diagnosed. Occasionally children recover fully without treatment. Others respond to treatment partially or completely. The majority of children however, will have a chronic relapsing disease course and will require long-term immunosuppressive treatment. Relapses may be triggered either as the treatment is withdrawn or following a viral infection.

Many children will experience ongoing problems with learning, coordination, behaviour and sleep. These difficulties require targeted intervention from the wider multidisciplinary team including speech and language therapy, occupational therapy and physiotherapy. Children with severe initial symptoms and those who are very young when symptoms start are at particular risk of developing long-term effects. It is important for those affected to



be identified early as they might benefit from new advances in immunomodulating therapy in specialist centres.

Once the condition is stable and symptoms are under control, support in school can be helpful. The Dancing Eye Syndrome Trust (details below) produces guidance for teachers which may be helpful in recognising support needed.

Further information and support

In the UK, the main support organisation offering information and support to anyone affected by opsoclonus myoclonus syndrome is the Dancing Eye Syndrome Support Trust. Call them on 01475 794 577 or visit their website at www.dancingeyes.org.uk