

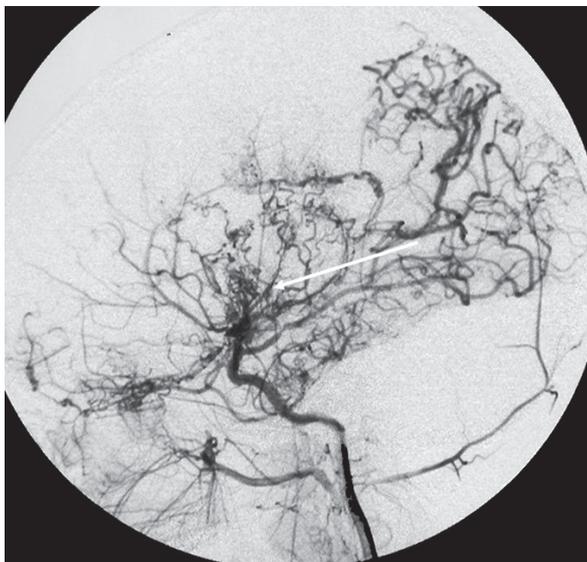


Moyamoya disease

This information sheet explains about moyamoya disease, some possible causes and how it can be treated. It also explains what to expect when your child comes to Great Ormond Street Hospital (GOSH) for assessment and treatment.

What is moyamoya disease?

It is a rare disease caused by narrowing (stenosis) or blockage (occlusion) of the four main blood vessels in the brain known as internal carotid arteries. The name 'moyamoya' means 'puff of smoke' in Japanese. This name describes the appearance of the network of small blood vessels that develop in the brain to compensate for the narrowing of the major blood vessels.



Brain angiogram showing typical 'puff of smoke' blood vessels seen in Moyamoya disease

What are the symptoms of moyamoya disease?

The symptoms of moyamoya disease in children can include headaches, problems with movement or difficulties with learning. Some of these symptoms are permanent, while others are temporary. Some children are not affected at all. A number of children with moyamoya disease have one or more strokes or 'mini-strokes' (transient ischaemic attacks or TIA). These strokes and mini-strokes can cause movement difficulties, such as muscle weakness, numbness or paralysis affecting one side of the body. The strokes and mini-strokes can lead to loss of function, such as speech, thinking or learning difficulties. Some children may also have seizures or involuntary movements in their arms or legs.



How is it diagnosed?

The diagnosis of moyamoya disease is usually based on findings from scans of the brain and blood vessels. Before being diagnosed, children may have a clinical assessment as well. Other scans that might help arrive at a diagnosis include CT or MRI scans to tell if areas of the brain and blood vessels have been affected, SPECT scans to examine blood flow to the brain and a cerebral angiogram to give detailed information about the blood vessels in the brain. For more information about these scans, please see our information sheets, available from your healthcare team or on our website.

What causes moyamoya disease?

The cause of moyamoya disease is not known. Some associations include injuries to the brain, infections or genetic conditions such as Down syndrome, neurofibromatosis or sickle cell disease. However, the majority of children with moyamoya disease are otherwise healthy.

How common is it and whom can it affect?

Moyamoya disease is very rare, affecting about 1 in every 1,000,000 (million) people in the UK. It affects more females than males. It is more common in Asia, especially Japan, but it can affect people from any racial background. In children, moyamoya disease is often diagnosed before the age of 10 years. The effects of moyamoya disease, when diagnosed in childhood, can be less in adulthood.

What treatments are available?

There are several treatment options or combinations of treatments available.

- **Watchful waiting** – Doctors may recommend that parents ‘watch and observe’ their child and inform the team if they are worried. Children will still require regular check ups, which may include repeat scans if there are any concerns.
- **Medicines** – Aspirin, for example, can be given at a low dose to stop blood becoming ‘sticky’ and forming clots. Although aspirin is generally not prescribed to children under the age of 16 years, in this case, the benefit of reducing blood clots is greater than the risk of side effects. The dose of aspirin required will be worked out according to the child’s weight.
- **Surgery** – There are many different types of ‘revascularisation surgery’, which can help return blood flow to the brain by opening narrowed blood vessels or bypassing blood vessels that are blocked. Extracranial-intracranial bypass (ECIC bypass) surgery uses healthy blood vessels to bypass the blockage and to provide additional blood supply to the areas of the brain that have been deprived of blood. For more information about ECIC bypass surgery, please see our information sheet.



What about other therapies?

Children with moyamoya disease may have learning difficulties or physical difficulties. These may impact on their general development and/or progress at school. For many children, an assessment by an occupational therapist (OT) or an evaluation by a psychologist may offer strategies to help support their day to day activities. If you have any concerns about your child's progress, please discuss it with your child's medical team.

What is the outlook for children with moyamoya disease?

The outlook can be variable. People diagnosed in childhood can often experience improved symptoms in adulthood.

Is there a support group?

There is currently no support group in the UK specifically for moyamoya disease. The website www.moyamoya.com is based in the United States but may be helpful in finding out more information and support. In the UK, the following groups may be able to help, although they are focused mainly on stroke.

Different Strokes

Helpline: 0845 130 7172

Email: webcontact@differentstrokes.co.uk

Website: www.differentstrokes.org.uk

Stroke Association

Helpline: 0845 30 33 100

Email: info@stroke.org.uk

Website: www.stroke.org.uk

**If you have any questions,
please contact the Clinical Nurse
Specialist for Neurology on
020 7405 9200 ext. 0569**

Notes

Compiled by the Neurology department in collaboration with the Child and Family Information Group with help from families of children with moyamoya disease.

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