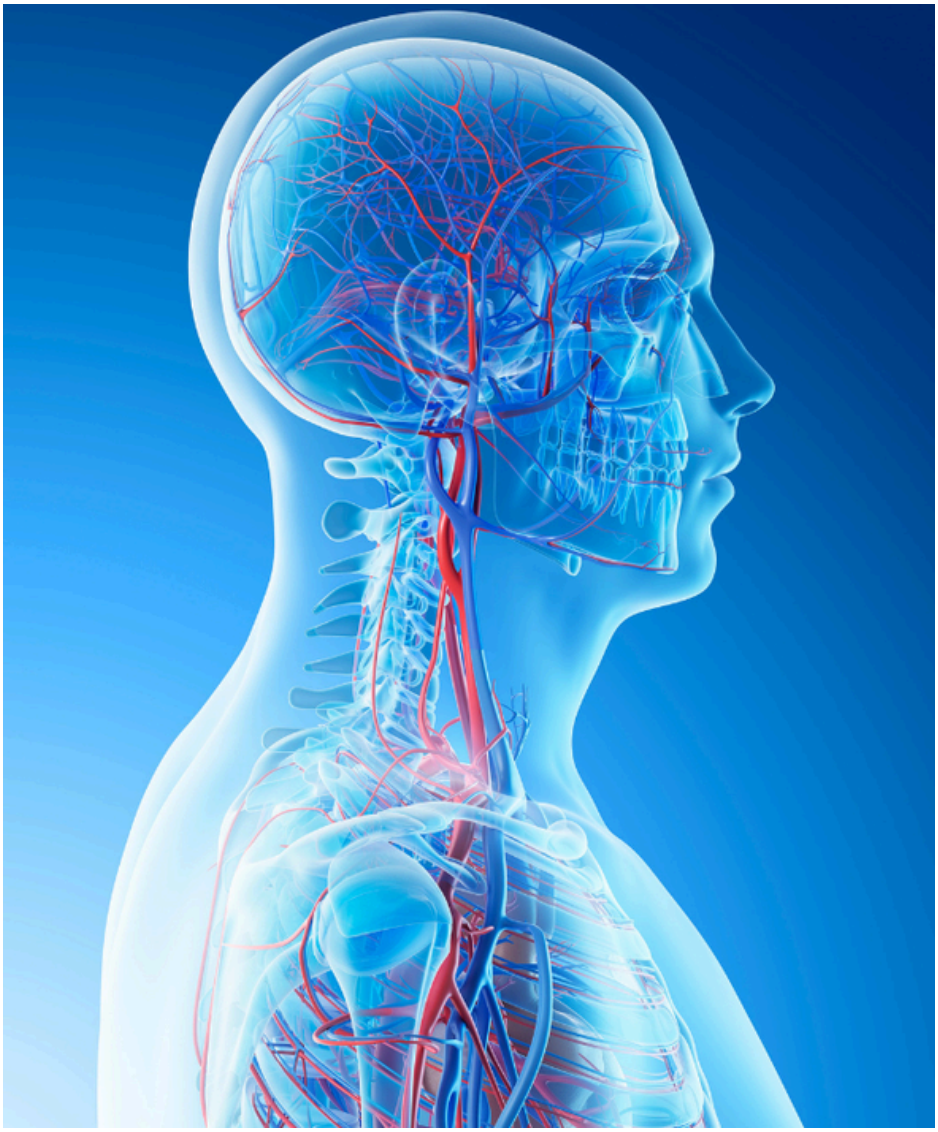
DOWN'S SYNDROME ASSOCIATION

The Down Syndrome Medical Interest Group

Moyamoya

July 2026



A Down's Syndrome Association publication

Moyamoya – Some Different Terms

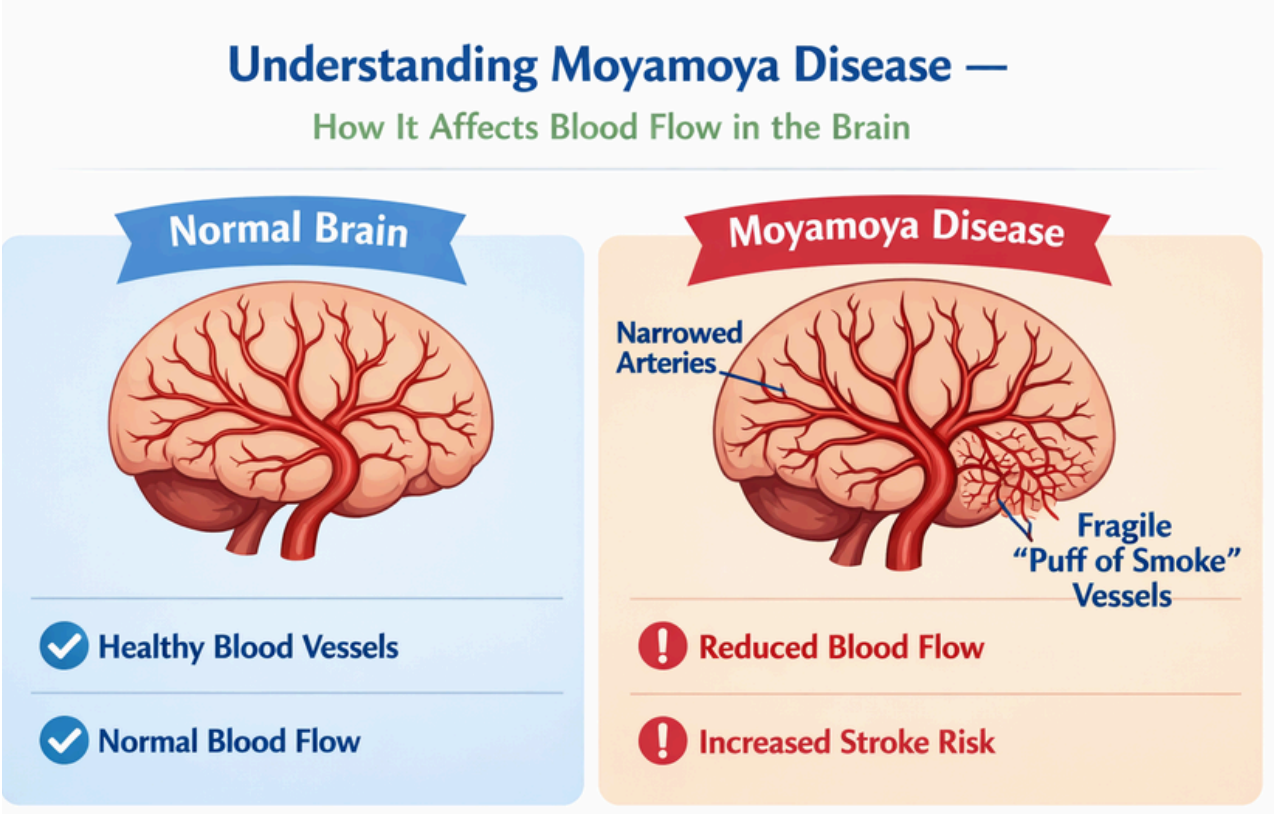
Moyamoya disease or syndrome is a rare condition that affects the blood vessels of the head and neck and can cause strokes. Moyamoya Disease is the name given to the condition when it affects someone without any other related health problems. When it is related to another condition, such as Down syndrome, it is called Moyamoya Syndrome. This can all be very confusing, so for this resource, we'll use the term moyamoya on its own, for ease.

What is Moyamoya?

In moyamoya, the arteries which take blood from the heart through the neck and into the brain become more and more narrowed. This leads to less blood reaching the brain. The role of these arteries is to deliver oxygen and nutrients to the brain, which are essential for the brain to work properly.

When the brain gradually gets less blood than it should, the body grows extra blood vessels to try to bypass the blocked or narrowed arteries. These extra blood vessels are tiny and fragile and not as good as the original blood vessels, so the body grows lots of them to try to make up the difference.

These are called 'collateral' blood vessels. This new network of tiny collateral blood vessels can look hazy like a 'puff of smoke' on CT or MRI scans, which is where the name moyamoya comes from – it means 'puff of smoke' in Japanese. In most cases it is not clear what causes moyamoya to develop.



How does Moyamoya Cause Strokes?

It's similar to what might happen if a motorway is closed or blocked. Cars divert off through a network of other roads, some of which might just be narrow tracks, which can also become blocked if too many cars try to drive down them. In the same way, the collateral blood vessels can become blocked too. Sometimes, they can burst, as they are so fragile, and cause bleeding into the brain.

When a part of the brain temporarily receives less blood than it should, it can temporarily stop working properly. This is a mini-stroke, or TIA (transient ischaemic attack).
When a part of the brain doesn't receive blood for a longer period of time, the brain tissue becomes damaged. This is an ischaemic stroke.

When there is bleeding into the brain, this also damages the brain and can put pressure on the brain. This is a haemorrhagic stroke.

Who gets Moyamoya?

Moyamoya is very rare but is more common in people who have Down syndrome than in the rest of the population. It is also more common in people of East Asian (particularly Japanese) descent, as well as people with conditions such as neurofibromatosis type 1, sickle cell disease and some autoimmune conditions.



In Japan, between 1 in 10,000 and 1 in 30,000 people has moyamoya. In Europe, it is estimated that about 1 in 100,000 people has moyamoya. For comparison, about 1 in 1,000 people have Down syndrome in the UK. It is not completely clear how much more common moyamoya is in people who have Down syndrome, with studies quoting between 4 and 26 times more likely compared to people without Down syndrome.

It can develop at any age, although about two-thirds of people with moyamoya will develop it as a child (most often around 4 to 10 years of age) and about one-third will develop it as an adult (most often around 30 to 40 years of age).

Males and females can both develop moyamoya, but girls and women develop it slightly more than boys and men.

What Symptoms or Problems does Moyamoya Cause?

The symptoms or problems someone with moyamoya has depends on the area of the brain that is affected. Mini-strokes, or TIAs, cause temporary problems, lasting from a few minutes up to 24 hours. Ischaemic strokes cause problems which last longer. It can be difficult for doctors to know whether a stroke is a mini-stroke or a 'proper' stroke, especially if it is only a short time since the symptoms started.

Both TIAs and strokes can cause problems such as:

- Weakness of an arm, leg or both – this may look like new clumsiness.
- Drooping face on one side – this might be more obvious when a person smiles.
- Changes in sense of touch, taste, hearing or vision.
- Abnormal movements.
- Confusion – this may cause changes in learning and development in a child or being more muddled or upset in an adult.
- Altered speech – speech may be more slurred or difficult to understand compared to normal, or the person might get more words muddled up or struggle to find the right word.
- Seizures.
- Changes in behaviour.

Haemorrhagic strokes can also cause loss of consciousness or collapse.
Symptoms might be triggered by coughing, straining, exercise, crying or fever, particularly in children.

In moyamoya, children are more likely to have TIAs or ischaemic strokes and adults are more likely to have haemorrhagic strokes.

What Should I do if I Think Someone is Having a Stroke or Mini-stroke?

If you think someone is having a stroke or a mini-stroke, call 999. It is very important that the person is assessed by a doctor or nurse as soon as possible. Don't wait to see if the symptoms get better.

It can be difficult to tell, sometimes, if someone has had a stroke, especially if the only changes are changes to the way someone is behaving. If you are unsure, seek medical help. Don't be afraid to ask 'could this be a stroke?' and remind the doctor or nurse that people who have Down syndrome are more likely to have moyamoya which can cause strokes.

How do I Know Someone has Moyamoya?

Unless someone with moyamoya has symptoms, as in the list above, it can be impossible to tell they have the condition. Sometimes it might be picked up during a scan for another reason, but most often it is found after someone has a stroke or mini-stroke.

What Tests Might be Done?



If someone has new, persistent or severe headaches, or has collapsed, a doctor or nurse should examine them and decide if a scan is needed. There are lots of other causes of headaches and collapse and not all of them require a scan.

If someone has symptoms of a stroke or mini-stroke then it is very likely they will have a CT scan. This type of scan can usually be done quite quickly. The person lays on a special bed which moves in and out of the scanner. The scanner is shaped like a large donut or polo-mint. It does not touch the person. It can sound a bit like a washing machine.

A CT scan of the head will usually only take a few minutes. It is often possible for someone to remain in the scanning room with the person, so long as they are not pregnant and they wear a lead apron to protect them from the x-rays used by the scanner. If the person is very scared, doctors will decide whether it is safe to use sedation. There are always risks with sedation, so these must be balanced against the benefits of getting the scan result.

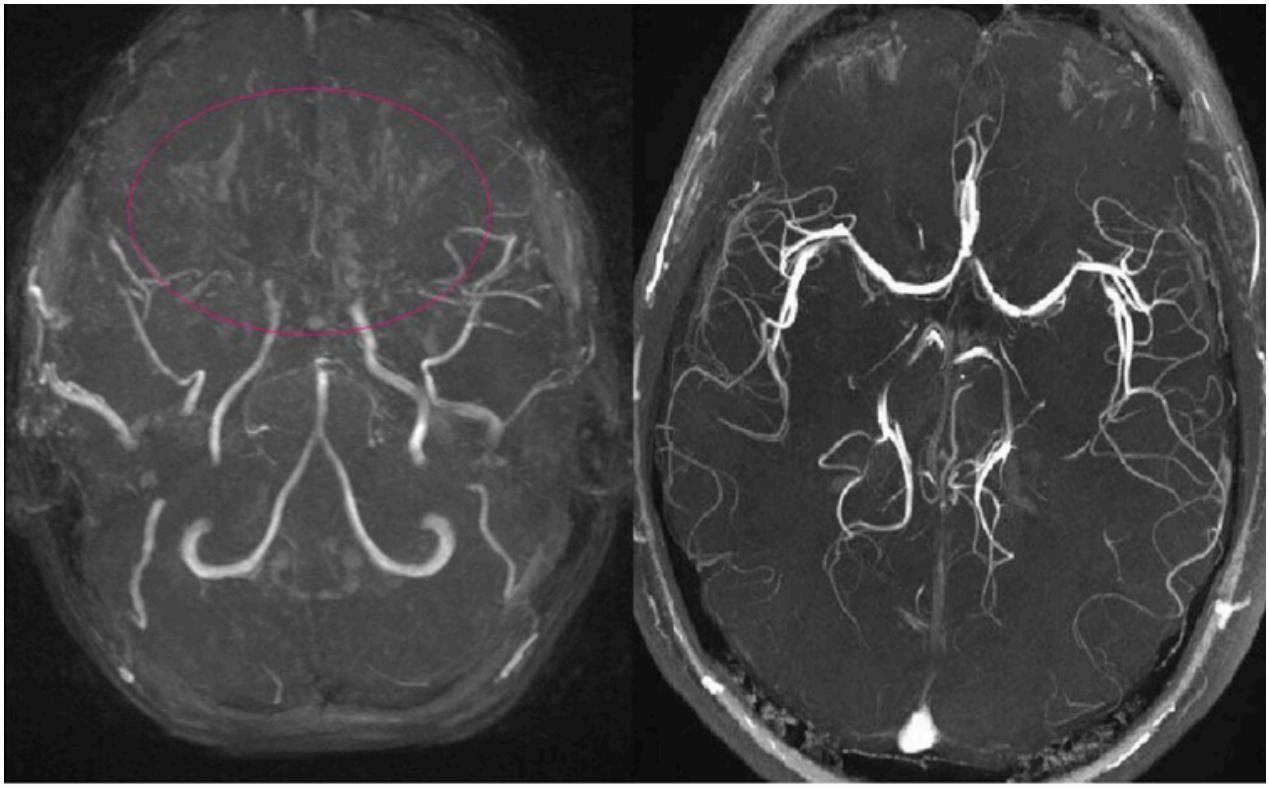




It is likely that contrast, or dye, will need to be given to get better pictures of the blood vessels. This is given through a drip or cannula, usually in the person's arm. Cold sprays or numbing cream may be used to help insert the cannula, although in some cases it might be decided that waiting for the cream to work is too dangerous (creams often take 30-45 minutes at least to work properly). If so, cold spray might still be possible. Some people might find it easier if someone explains things clearly first, especially with pictures or demonstrating the equipment, whilst others prefer to be distracted with a phone, tablet or book for example.

Sometimes an MRI scan is also needed. This shows much more detail compared to a CT scan, but is more difficult to arrange and to carry out.

It takes much longer than a CT scan (30 minutes to an hour) and involves laying on a bed inside the scanner, which is a narrow tunnel. It can be quite noisy. Some people manage well with music to listen to or a virtual reality screen to look at. Others may need sedation or a general anaesthetic.



Smaller collateral blood vessels seen on left image.

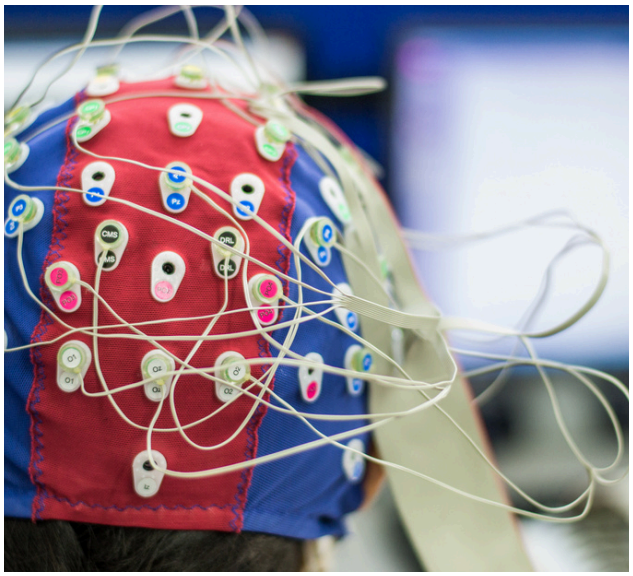
CREDIT commons.wikimedia.orgwikiFileMRA_Moya-moya-disease

An EEG (electroencephalogram) may also be carried out.

This involves electrodes attached to the scalp to record the electrical activity in the brain.

This can be easier if a person is allowed to get used to the electrodes beforehand and if the procedure (which is painless) is explained clearly.

Sedation may also be considered.



How is Moyamoya Treated?

Moyamoya cannot be cured and is a lifelong condition. Patients with moyamoya are often looked after by neurologists (brain specialists) and neurosurgeons (brain surgeons).

A referral might need to be made to a specialist centre in a different part of the country. Surgery might be carried out to help improve blood flow to the brain. Blood thinning medication might be used to try to prevent ischaemic strokes. Medications may also be used to control the person's blood pressure, as high blood pressure can make strokes more likely.

A healthy lifestyle, including a healthy diet, exercise and avoiding smoking, is also important.





A healthy lifestyle, including a healthy diet, exercise and avoiding smoking, is also important.

More information can be found here:

mencap.org.uk/easyread/moyamoya

nhs.uk/conditions/stroke/

CONTACT US

Down's Syndrome Association

Langdon Down Centre,
2a Langdon Park, Teddington,
Middlesex, TW11 9PS

0333 1212 300

info@downs-syndrome.org.uk
training@downs-syndrome.org.uk
downs-syndrome.org.uk

Wales
wales@downs-syndrome.org.uk

Northern Ireland
enquiriesni@downs-syndrome.org.uk



A Down's Syndrome Association publication
© Down's Syndrome Association 2026

Photosymbols®
Professional Licence 6132156530743