



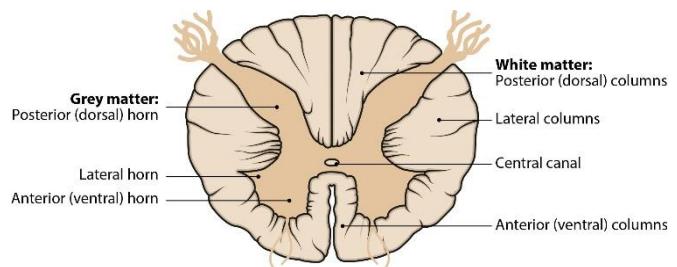
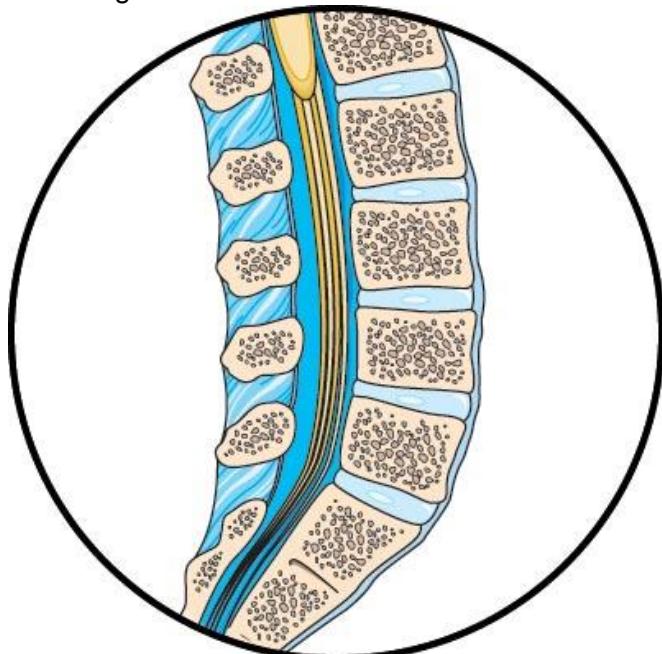
NHS

Great Ormond Street
Hospital for Children
NHS Foundation Trust

Acute flaccid myelitis: information for families

Acute flaccid myelitis (AFM) is a rare condition that tends to occur in ‘outbreaks’ every few years. It affects the spinal cord – particularly the grey matter – and over hours to days muscles weaken and normal limb reflexes disappear. It often follows a mild viral infection affecting the upper airway or stomach. More research is needed to find out why a small proportion of children carry on to develop AFM whereas the vast majority recover from the infection with no lasting effects. This information sheet from Great Ormond Street Hospital (GOSH) explains the causes and symptoms of AFM and how it can be managed. An Easy Read information sheet is included for your child.

The spinal cord is part of the central nervous system and plays an important role in how messages travel to and from the brain. These messages are important for conscious functions such as walking and moving (muscle squeezing and relaxing), but also unconscious things such as breathing.

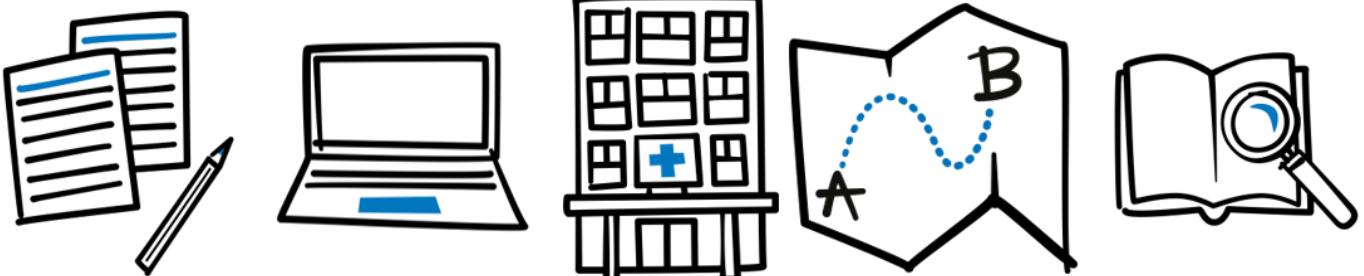


In AFM, the grey matter in the spinal cord is damaged which interferes with message transmission to and from the brain, leading to sudden weakness.

What causes acute flaccid myelitis?

The precise mechanism leading to a child developing acute flaccid myelitis (AFM) has not yet been confirmed but it is clear there is a link with viral infections, often a specific virus caused enterovirus. This means that outbreaks tend to occur in the latter part of the year in the northern hemisphere from August to November when viral infections are more prevalent.

When doctors in the US tested the cerebrospinal fluid (CSF) that surrounds the brain and spinal



cord, they found virus cells, even if a child had recovered from the original viral infection.

It is clear that the grey matter within the spinal cord is damaged in AFM. This particularly affects the area containing motor neurons which are needed for movement.

How common is it?

Acute flaccid myelitis (AFM) is a very rare condition mainly affecting children aged under 18 years. It may be misdiagnosed as there are other similar conditions (such as Guillain Barre Syndrome and Transverse Myelitis) but the Centers for Disease Control in the US have identified over 600 children with the condition to date worldwide. It tends to occur in outbreaks every few years.

What are the symptoms of acute flaccid myelitis?

Initially, the main symptom of AFM is sudden asymmetrical weakness affecting the arms or leg. Asymmetrical means that it can affect each side of the body in a different way or to a different degree. This can develop over hours to days. As well as weakness, the limb becomes floppy (flaccid) and the normal spinal reflexes decrease or disappear.

Muscle weakness can also affect the face, causing the eyelid to droop or one side of the face to weaken or can cause difficulty breathing or swallowing or problems with bladder and bowel. The weakness can be uncomfortable and even painful. Symptoms can occasionally include numbness or tingling.

In around 20 per cent of children, this muscle weakness affects the diaphragm, the curved muscle in the chest that rises and falls when breathing. This can affect breathing and may be serious enough to need support from a ventilator in intensive care.

Other children may have neurological complications that affect automatic body functions

(the autonomic system) such as controlling temperature and blood pressure.

How is acute flaccid myelitis diagnosed?

It is important to take your child to the doctor if they develop any sudden weakness. Identifying AFM quickly and starting treatment can reduce the risk of complications from serious problems such as when breathing is affected.

The doctor will ask lots of questions about which symptoms appeared when and whether your child has had any recent infections, particularly viral ones affecting their airway or stomach. As well as taking a history, the doctor will do a physical examination to see what your child is able to do. They will also organise bloods, urine and stool and a few specific tests.

A **lumbar puncture** is needed to collect a sample of cerebrospinal fluid (CSF) from around the spinal cord – this is then examined in our laboratory. Children with AFM usually have more cells than usual in their CSF. It is also important to analyse the fluid for other viruses.

A **magnetic resonance imaging (MRI)** scan of the spinal cord which may show areas of damage affecting the grey matter of the spine. Early on the MRI can be normal, so it may need to be repeated after a few days.

Electromyography (EMG) and Nerve Conduction studies (NCS) may be carried out to look at how well the message is being sent along the nerve and to try to see where the problem lies.

Further information about these tests is available on our website.

How is acute flaccid myelitis treated?

As acute flaccid myelitis is rare and relatively new, there is not yet an agreed way of treating it. However, the following treatments have been



used in many situations. The doctors will explain which are suitable for your child.

Intravenous immunoglobulin (IVIG) is a blood product and is given often as replacement for people who are unable to make their own antibodies. In neurological conditions, it is often used to dampen an overactive inflammatory response, such as that seen in the spinal cord in AFM. It is usually the first treatment given in AFM. It is given as an infusion into a vein through a cannula over a number of hours, and may be repeated the next day.

Corticosteroid treatment is frequently used to reduce inflammatory symptoms. In AFM it is not clear whether steroid treatment is of benefit or not. If the doctor decides they may be of benefit, corticosteroids are usually given into a vein once a day for three to five days.

Plasma exchange, or plasmapheresis, is a procedure that “washes” the blood, removing plasma, the part of the blood which contains antibodies, from the blood cells and replaces the plasma with a different fluid (usually albumin). Currently, there are no recommendations for using antiviral medications or other medications to damp down the immune system.

Physiotherapy, occupational therapy and if necessary speech and language support are important aspects of treatment to maintain. Detailed assessment should start soon after diagnosis and will be needed on a long term basis.

In cases where outcome is poor, a procedure called '**nerve transfer**' may be suggested. This surgical procedure re-routes nerves so that they connect with areas where the previous nerve supply has been damaged. This is usually considered in the first 9 to 12 months after diagnosis when it is most likely to be successful.

Functional electronic stimulation has been tried in similar conditions with some success. This involves implanting a stimulator to make affected muscles work using electrical impulses.

What is the outlook for children and young people with acute flaccid myelitis?

As acute flaccid myelitis (AFM) is a newly discovered disease, there isn't much data about the long term effects and outlook for those affected. From the cases that have been studied to date, AFM is life-threatening for a small number of children, particularly those whose breathing is affected.

Otherwise, children do recover in time but many continue to have permanent muscle weakness in the worst affected limb. It may also be uncomfortable. Input from a psychologist can also be helpful.

A very small number of children recover completely from AFM with no lasting effects.

Further information and support

Although acute flaccid myelitis (AFM) is different to multiple sclerosis, the effects can be similar so organisations for people with MS may be able to offer support and advice to people affected by AFM. The **MS Society** has a helpline on 0808 800 8000 or visit their website at www.mssociety.org.uk.

The **Transverse Myelitis Society** may also be able to help. Their helpline number is 020 8568 0350 and their website is at www.myelitis.org.uk



The **British Brain and Spine Foundation** helpline is run by neurological nurses. Call them on 0808 808 1000 or visit their website at www.brainandspine.org.uk

The **British Trust for the Myelin Project** supports researchers and other people with an interest in demyelinating disorders. You can call them on 0161 292 3191 or visit their website at www.myelinproject.co.uk

All about acute flaccid myelitis (AFM)



Your spinal cord runs down your back and send messages to and from the brain. Nerves run off the spinal cord to reach every part of your body.



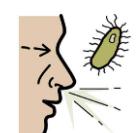
Sometimes, your spinal cord can be damaged, which means that areas of your body like your arms and legs can be weak and floppy.



In AFM, this happens to a small number of children after they have had a viral infection. We're not really sure why this happens.



The first sign of AFM is a sudden weakness in your arm or leg. It may also be numb or tingling.



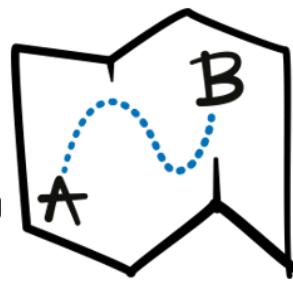
This can happen up to 2 weeks after you have had a viral infection. The types of infection vary but are usually a chest infection or tummy bug.



The doctor will ask lots of questions to find out more. They will want to know when the weakness started and if you have recently had a viral infection.



They will also ask you to do certain things, like walk, stand up from sitting or raise your arms above your head. They will look at how easy or hard this is for you.





You will also need a lumbar puncture. This lets the doctors take a tiny sample of fluid from around your spinal cord. This can be uncomfortable but the doctors will be as quick as they can.



You will also have an MRI scan of your spine. You may need to have an anaesthetic for this so you can keep really still. An MRI scan is noisy so you will have headphones to wear. You can watch a video or your favourite film while it happens



You will usually have EMG and NCS tests – these check how messages travel to and from your brain and spinal cord from your arms and legs.



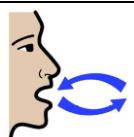
When the doctors have worked out you have AFM, they will start treatment. This will be different for everyone but could include:



Infusions of IVIG medicine or steroid medicine or both. These are given through a cannula in the back of your hand or arm.



Plasma exchange is a bit like kidney dialysis but the machine cleans your blood to replace the straw coloured liquid that holds your blood cells.



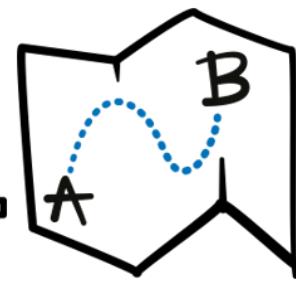
A very small number of children may need help with their breathing. They may have to go to intensive care for a while.



Physiotherapy is important to keep your muscles are strong as possible. You will have sessions with a physio and exercises to do in between.



After you have recovered from AFM, you might still have weak muscles in your arm or leg. You will need to carry on with physio for a while.





An occupational therapist (OT) can help you to find ways of doing things, even if your arm or leg is weak. You may have to learn new ways of doing things or use aids to help you.



Your arm or leg may be sore for a while afterwards. You can ask for help from the Pain team to help you find ways of managing it.



It may help to see a psychologist to work with you to manage your feelings and find ways of coping with everyday life.



Most children carry on having muscle weakness but with help can do most day to day activities, go to school and play sport.



Please ask us if you have any questions.

