This fact sheet provides information on Chiari malformations. It focuses on Chiari malformations in adults. Our fact sheets are designed as general introductions to each subject and are intended to be concise. Sources of further support and more detailed information are listed in the Useful Contacts section. Each person is affected differently by Chiari malformations and you should speak with your doctor or specialist for individual advice.

What is a Chiari malformation?
A Chiari malformation is when part of the cerebellum, or part of the cerebellum and part of the brainstem, has descended below the foramen magnum (an opening at the base of the skull).

The cerebellum is the lowermost part of the brain, responsible for controlling balance and co-ordinating movement.

The brainstem is the part of the brain that extends into the spinal cord.

Usually the cerebellum and parts of the brainstem are located in a space within the skull above the foramen magnum. If this space is abnormally small, the cerebellum and brainstem can be pushed down towards the top of the spine. This can cause pressure at the base of the brain and block the flow of cerebrospinal fluid (CSF) to and from the brain.

Chiari malformations are named after Hans Chiari, the pathologist who first described them. Chiari malformations are sometimes referred to as Arnold-Chiari malformations (this name usually refers specifically to Type II Chiari malformations; see below). Another term sometimes used is hindbrain hernia.

Cerebrospinal fluid (CSF) is a clear, colourless fluid that surrounds the brain and spine. Its main functions are to protect the brain (it acts as a shock absorber), to carry nutrients to the brain and to remove waste.

Are there different types of Chiari malformations?
Yes. Chiari malformations are divided into different groups. Your specialist will tell you which type you have.
Type I Chiari malformations are the most common type. Type I malformations involve part of the cerebellum (the lowest part called the cerebellar tonsils) extending down into the spinal canal.

The other types of Chiari malformation are different from the Chiari I and are associated with other conditions such as spina bifida, hydrocephalus and some conditions existing at or before birth. Type II Chiari malformations are less common than Type I malformations and are more serious. They usually present in childhood and are associated with hydrocephalus in adults. Type II malformations involve parts of the cerebellum and brainstem (the lowest part called the medulla or medulla oblongata) extending downwards towards the spine.

Types III and IV Chiari malformations are very rare and more serious. They affect children and symptoms appear at birth and during infancy.

What causes Chiari malformations?
The Chiari I malformation is thought to be in most cases related to a small posterior fossa (the cranial bit of the skull that holds the cerebellum). The Chiari II and III malformations are caused by abnormalities in the structure of the brain and spine which develop in the womb before birth. This might be due to genetic factors or other reasons, such as a lack of vitamins and minerals during pregnancy.

Although rare, some Chiari malformations can be caused by an injury or an infection. Chiari I malformations can also be caused by a build up of pressure in the brain, for example, hydrocephalus or tumour, or by the spinal cord being held down (for example, a tethered cord).

Is it hereditary?
It is possible that children born with this disorder may have inherited the gene. Research is currently being carried out to identify this gene. However, the risk of passing a Chiari malformation on from parent to child is very small. Also, if the child does inherit the gene, most will not develop symptoms.

What are the symptoms of Chiari malformations?

Type I Chiari malformations
Often people with Type I malformations will not experience any symptoms. For some people, symptoms might only develop later in life. People can be born with a Type I malformation and not experience any symptoms until they are adults. If people with Type I malformations do experience symptoms they might be similar to those experienced by people with Type II malformations (see below). Headaches and neck pain are especially common.

Type II Chiari malformations
The main symptoms people with Type II malformations might experience are headaches (usually at the back of the head and often made worse by coughing, sneezing or straining), back pain, dizziness and balance problems, unusual feelings in the arms or legs (numbness or tingling), muscle weakness and paralysis, visual problems and involuntary movement of the eyes (nystagmus), swallowing problems, hearing loss and tinnitus.
People might also experience nausea (feeling sick), vomiting (being sick), insomnia (difficulty sleeping) and depression. For help with depression, your GP can advise you of mental health services.

Children with Types III and IV Chiari malformations experience severe neurological symptoms, which are not covered in this factsheet.

**Syrinx and Syringomyelia**

Those with type I Chiari malformations often develop a condition called syringomyelia. This is when a hollow space forms within the spinal cord and fills with fluid - which is known as a syrinx. Over time, this syrinx can enlarge and press on the spinal cord, which can damage it. The symptoms of this include numbness, weakness, pain, stiffness, problems with bladder and bowel function and in some cases, paralysis.

Patients with a Chiari II or III are much more likely to have a syrinx, but their treatment is often different that those with a Chiari I.

This can be treated through surgery or through releasing the fluid (see 'treatments' section below).

**Tests and investigations**

For people who do not experience any symptoms, Type I Chiari malformations might be discovered by chance after a scan to investigate a different illness or condition.

For people with symptoms, doctors will perform a thorough neurological examination. The key tests to diagnose Chiari malformations are CT scans or MRI scans to check for abnormalities in the structure of your brain and spine.

(You might like to read our fact sheet on brain and spine scans for further information.)

**What are the treatments?**

Pain relief medication can be given to relieve headaches and neck pain.

The key treatment for Chiari malformations is surgery. Surgery will be considered and discussed on an individual basis and will not be suitable for everyone. The particular type of surgery will differ for individuals and you might need more than one operation as part of your treatment. Your neurosurgeon will discuss your options with you.

The main type of surgery to treat Chiari malformations is to increase the space at the top of your spinal cord and back of your brain to allow more free movement of fluid between the brain and spinal canal.

Other treatment options include:

- **Endoscopic third ventriculostomy (ETV):** In patients with hydrocephalus, fluid in the brain ventricles (fluid filled cavities within the brain) is released by a small hole made in the wall of one of the ventricles.
- **Ventriculoperitoneal shunting:** In patients with hydrocephalus, fluid in the brain can also be drained by the drilling of a small hole into the skull and a catheter (thin tube) being passed from the ventricle usually into the peritoneal cavity (the abdomen).
- **Untethering:** This is a process to divide a band of tissue that can hold the bottom end of the spinal cord down to the end of the spinal canal.
- **Syringo subarachnoid shunt:** In cases where the syrinx worsens or continues to be symptomatic once the Chiari malformation has been treated, a catheter can be placed into the syrinx cavity to drain it into the cerebrospinal fluid (CSF) pathway.
Hydrocephalus: a build-up of CSF in the brain. Hydrocephalus might be treated with a shunt (a thin tube implanted in the brain to drain away the excess CSF). (You might like to read our fact sheet on hydrocephalus and shunts for further information.)

Syringomyelia: a condition in which a cavity called a syrinx grows in the spinal cord and fills with CSF. Syringomyelia might be treated with a shunt.

Spina bifida: a group of birth defects involving problems with the development of the spine. Spina bifida is associated with Type II Chiari malformations in children.

Tethered cord syndrome: a condition in which the spinal cord attaches itself to vertebrae (spinal bones).

Curvature of the spine: the spine might curve abnormally to the left or right (scoliosis) or curve abnormally outwards (kyphosis).

As with any form of surgery, there are risks associated with surgery to treat Chiari malformations. Sometimes, surgery leads to no improvement or even worsening of symptoms. Your neurosurgeon will discuss these with you before your operation.

Possible risks of decompression surgery for Chiari malformation include:

- risk to life
- stroke or haemorrhage (bleeding)
- paralysis of the arms and legs
- meningitis or other infection
- impaired speech
- memory loss or problems with thinking
- difficulty swallowing
- balance problems
- hydrocephalus
- seizures (although these are rare)
- reoccurrence of symptoms

However, most people who have surgery find that their symptoms improve afterwards. Even if symptoms do not improve significantly, surgical treatment of Chiari malformations might prevent existing symptoms from worsening.

**Other conditions associated with Chiari malformations**

People with Chiari malformations might also experience:

- **Hydrocephalus**: a build-up of CSF in the brain. Hydrocephalus might be treated with a shunt (a thin tube implanted in the brain to drain away the excess CSF). (You might like to read our fact sheet on hydrocephalus and shunts for further information.)
- **Syringomyelia**: a condition in which a cavity called a syrinx grows in the spinal cord and fills with CSF. Syringomyelia might be treated with a shunt.
- **Spina bifida**: a group of birth defects involving problems with the development of the spine. Spina bifida is associated with Type II Chiari malformations in children.
- **Tethered cord syndrome**: a condition in which the spinal cord attaches itself to vertebrae (spinal bones).
- **Curvature of the spine**: the spine might curve abnormally to the left or right (scoliosis) or curve abnormally outwards (kyphosis).
Physiotherapy
Depending on your individual symptoms, you might benefit from having physiotherapy. A physiotherapist can help with physical problems like muscle weakness and difficulties with balance, movement and co-ordination. Many people find that physiotherapy is an important part of their longer-term rehabilitation.

Useful contacts
Brain & Spine Helpline
0808 808 1000
www.brainandspine.org.uk

Run by neuroscience nurses, providing support and information on all aspects of neurological conditions for patients, their families and carers, and health professionals.

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