

CRANIO-CERVICAL DECOMPRESSION FOR CHIARI MALFORMATION: PATIENT & FAMILY INFORMATION SHEET

What is a craniocervical decompression?

A craniocervical decompression is an operation involving the back of the head and top or the neck, i.e., the craniocervical junction, where the neurosurgeon creates extra room for the bottom part of the brainstem and top part of the spinal cord.

In what circumstances is this performed?

A craniocervical decompression operation is performed in a patient where there is not enough space around the in the area where the top of the spinal cord enters through the skull to become the brainstem. This is usually because of the presence of an 'Arnold-Chiari' malformation.

What is an Arnold Chiari malformation?

An Arnold-Chiari malformation, named after the doctors that first described it (also known as a hindbrain hernia), is where a small part of one of the brain structures protrudes through the hole in the base of the skull (known as the foramen magnum) through which the spinal cord enters to become the brainstem. The brain structure concerned, called the cerebellum, usually lies entirely within the skull cavity. It is thought that in some people the skull compartment in which the cerebellum sits has not grown large enough to accommodate the cerebellum and as a result the very bottom part of the cerebellum (known as the cerebellar tonsils) protrudes out through the skull (through the foramen magnum) into the top of the spinal canal. This protrusion or 'herniation' of the cerebellar tonsils is the Arnold-Chiari malformation. As a result, the space around the top of the spinal cord/bottom of brainstem at the level of the craniocervical junction can become very tight. In many people, an Arnold-Chiari malformation may not cause any problems. Some people however can have significant symptoms such as 'Val Salva' headaches (i.e., headaches particularly exacerbated by coughing, sneezing, laughing, or straining on the toilet). In others, an Arnold-Chiari malformation is associated with the development of a spinal cord syrinx.

What is a syrinx?

A syrinx is an abnormal fluid-filled cavity within the spinal cord. The condition is also known as syringomyelia. A spinal cord syrinx is often progressive, that is, over time, it enlarges and affects more of the spinal cord over time causing more and more symptoms. A syrinx can cause irreversible damage to the spinal cord. It can affect the use of your hands, cause disturbing discomfort trunk and limbs, and result in difficulty walking. It is found in some patients with an Arnold-Chiari malformation. An Arnold-Chiari malformation is the commonest cause of a syrinx. We do not understand exactly how an Arnold-Chiari malformation can cause a syrinx at some distant point in the spinal cord. We do know that in the majority of patients with a syrinx caused by an Arnold-Chiari malformation, creating more space around the craniocervical junction, i.e., performing a craniocervical decompression, stops progression of the syrinx and prevents further deterioration in the majority of patients and can result in some symptom improvement.

What is the benefit of a craniocervical decompression?

*If you **do not** have a syrinx:*

The aim of the surgery is to help with any significant Val Salva headaches significantly interfering with lifestyle. Other symptoms may or may not be improved by the surgery. If you do not suffer from significant Val Salva headaches or have not had episodes of loss of consciousness or your imaging does not show a syrinx, a craniocervical decompression is probably not appropriate.

*If you **do** have a syrinx:*

The aim of the surgery is to prevent future neurologic deterioration, i.e., to maintain the status quo/stop things getting worse. Any improvement is a bonus. You are strongly advised to have surgery if your neurosurgeon has identified your spinal cord as having a syrinx and has recommended an operation. Further neurological deterioration over months to years is considered to be the usual course.

How is a cranio-cervical decompression done?

- A cranio-cervical decompression is done under general anaesthetic, that is, you are put asleep for the surgery by an anaesthetist.
- Depending on the circumstances of the operation and individual consultant neurosurgeon practice, you may have a portion of your hair shaved or a small strip hair shave along the course of your scalp cut at the back of your head/neck.
- Your head may be fixed in a head clamp after you have been put to sleep to keep your head very steady during the surgery. If this is the case, you will notice three puncture marks where the clamp was fixed to the skull, including one to two on the forehead after surgery.
- The skin cut is at the top of the neck/back of the head and is usually between 5 to 10 cm in length, depending on an individual patient's anatomy. The cut is deepened through the neck muscles to expose the bottom part of the back of the skull and the back part of the first and/or second vertebral bones in your neck. A small area of bone from the bottom part of the back of the skull is removed and also the back part (also known as arch or lamina) of the first (and sometimes second) vertebral bone in the neck. The canvas-type layer (or dura) over the back of the lower part of brainstem and top of spinal cord is then opened, thus completing the decompression. The wound is then closed.

What can I expect when I wake up and as I progress?

- You will be in the Intensive Care or High Dependency Unit for at least one day, sometimes longer.
- You will be connected to equipment that helps us monitor your condition. Usually, such monitoring is an oxygen measuring probe on your finger and perhaps an electrocardiogram (ECG). These measuring devices will be quickly removed as you progress.
- Some patients find it difficult to sleep in such an environment. We will move you to a quieter less busy ward area as soon as we think it is appropriate.
- You will likely wear a head bandage for a number of days. The main purpose of such a bandage is to stop you itching your head wound when you are asleep.

- You may experience headache, nausea, pain, or discomfort. You will be given medication to relieve these symptoms.
- You will be allowed to eat. You will start by drinking liquids and slowly more foods will be added.
- Your activity will be gradually increased under the supervision of physiotherapy and nursing staff. You will often be got out of bed the first day after you operation.
- In the one to two weeks following surgery, many patients experience what is considered to be a 'chemical meningitis' manifested by headaches and feeling generally unwell. This is presumed to be due to the release of inflammation chemicals from muscle into the CSF through the gap made in the dura at the time of surgery and spontaneously resolves as the surgical wound heals.

What are the risks and possible complications of a craniocervical decompression?

Serious complications are uncommon. There are many steps that we take to try and prevent complications happening and things that we do to reduce the impact of such complications when they do happen. When complications do occur they are often treatable. Problems that can occur include:

1. Bleeding or a blot clot inside the head where the surgery has been performed (<1:100). This would be potentially life-threatening.
2. Stroke, i.e., where an area of brain tissue dies because of blocking off of a blood vessel (for example, occasionally a blood vessel near the site of surgery can become inflamed leading to blood clot sticking to the wall of the blood vessel and blocking the blood vessel leading to a stroke) (<1:100).
3. Wound infection (<1;100).
4. Cerebrospinal fluid leakage (CSF) through wound (1-2:100). This can result in infection in the CSF (bacterial meningitis).
5. Pseudomeningocele (1:100). This is where a large bulge develops underneath the skin in the region of the surgical wound, containing CSF.
6. Hydrocephalus (1:00). This is where the circulation of CSF from cavities inside the brain to the outside surface of the brain gets blocked. These cavities or ventricles swell and can put pressure on the brain from inside. A further operation can be required, i.e., insertion of a 'shunt'.
7. 'Brain sag' (1-2:100). This is a poorly understood condition where a part of the brain surface 'sags' away from its normally close contact with the overlying dura/skull and is a cause of severe headaches following the surgery. It generally resolves with bedrest.
8. Deep venous thrombosis (clot in the leg)/pulmonary embolus (clot in the lung).
9. Medical problems with respect to your general health, e.g., myocardial infarct (heart attack), chest infection etc.

There may in addition be potential problems relevant to your specific operation that your surgeon will tell you about.

What will happen when I go home?

- Take your medicines as directed. Never stop without asking your doctor. Finish all antibiotics, even if you feel better.
- If you have any worries following discharge, you should contact the neurosurgical ward you were discharged from (N1 0114 2712320 or N2 0114 2712896) or alternatively the secretary of the consultant neurosurgeon who operated on you.
- You should also receive a follow-up clinic appointment for about six to eight weeks following your surgery.
- If you had a syrinx, a baseline MRI scan will usually be arranged by about three to four months after your surgery to see to what extent the syrinx has reduced in size.

What will be my long term outlook?

The aim of the surgery is to return you to as normal a life as possible, unless there has been significant neurologic deterioration already from any co-existent syrinx. Some patients may also have symptoms that persist despite treatment of the Arnold-Chiari malformation. For patients with a syrinx undergoing a craniocervical decompression, MR imaging done at around three to four months post-surgery should show collapse or reduction of their syrinx, generally predictive of a good outcome.

Are there any support groups that I can contact for advice?

There are a variety of patient support groups that may be of assistance depending on your circumstances. In particular, you may wish to contact the Ann Conroy Trust, registered charity that supports individuals affected by an Arnold-Chiari malformation or syringomyelia.

T. Carroll/Dept of Neurosurgery/10th December 2012

This information sheet is to be used only in combination with attendance at Arnold Chiari/Syringomyelia Neurosurgical Clinics, Department of Neurosurgery, Royal Hallamshire Hospital, Sheffield Teaching Hospitals NHS Foundation Trust.