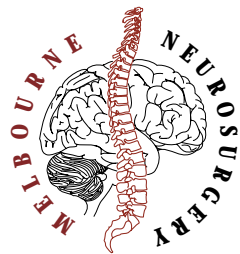
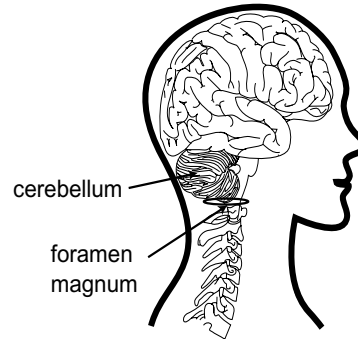


# CHIARI MALFORMATION



## WHAT IS A CHIARI MALFORMATION ?

This is a developmental abnormality where parts of the brain inside the head prolapse out through the hole in the back of the skull called the FORAMEN MAGNUM. This hole normally only allows the spinal cord to pass through.



There may also be an abnormality where a cyst forms in the spinal cord (called a syrinx). Depending on the degree of the chiari abnormality it can be divided into many types.

## WHAT ARE THE TYPES OF CHIARI MALFORMATION ?

There are four main types

Type I. This is when the cerebellar tonsils alone are displaced through the foramen magnum.

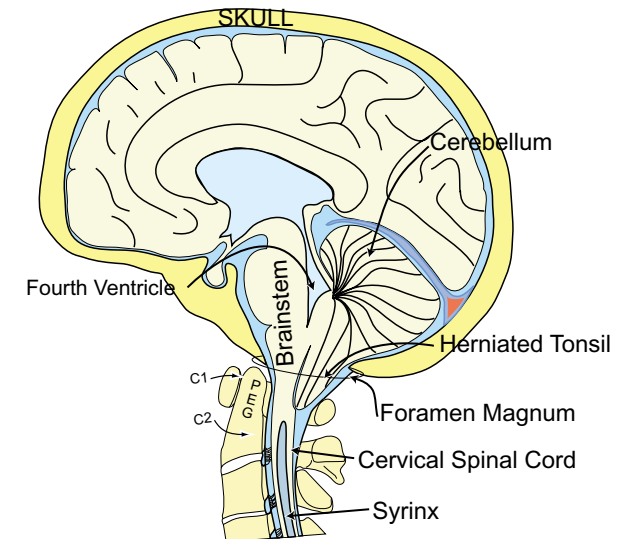
Type II. Not only are the tonsils displaced, but the brainstem and fourth ventricle may also be in the spinal canal. This usually associated with spina bifida (myelodysplasia) The tonsils are displaced further down the spinal canal than is type I.

Type III Here there is a defect in the bone at the back of the spine with brain herniated into a fluid filled cyst in the defect.

Type IV (v.rare) in this type the cerebellum does not develop properly and parts are missing.

These abnormalities can be associated with changes in

1. The brain  
extra folds in the brain
2. The bones around the base of the skull  
Sinking of the skull onto the top of the spine  
(basilar Impression)
3. The bone of the neck  
fusion of segments  
(e.g. Klippel-Feil syndrome)



## Midline picture through the brain and upper cervical spinal cord

Shows the tonsils protruding down through the Foramen Magnum and the syrinx that has developed in the spinal cord

## What symptoms could you have ?

The symptoms can either occur as a child or adult.  
The commonest ones are due to:

General:	headache
Cerebellum Problems:	balance coordinatiuon
Brainstem/Spinal Cord:	visual disturbance jerking of eyes swallowing/talking problems drop attacks cough headache muscle stiffness sensation abnormalities

## What is a Syrxinx

This is a general term for the dilation of the centre of the spinal cord with fluid. This may be due to obstruction to the top of the spinal cord from pressure or inflammation. There are two types

Syringomyelia: Dilation of the canal and rupture into the substance of the spinal cord.

Hydromyelia: Dilation of the canal

(see leaflet on syringomyelia)

## HOW IS THIS DIAGNOSED ?

Your doctor will usually make the diagnosis from the symptoms that you have. There are lots of diagnoses that are possible from your symptoms.

When the possibility is raised that you have Chiari malformation then you will need to have some tests.

### PLAIN X-RAYS

These are often the first test. This may show that there is an abnormality of the bones of the neck.

### CAT SCAN

This is usually next. This is a computerised X ray of your brain and spine that will show how much the diameter of the foramen magnum is reduced and may show the tonsils herniated. It is unlikely to show a syrxinx.

### M.R.I. (Magnetic Resonance Imaging)

This is the next test. It produces pictures similar to the CAT scan but they are generated using a magnetic field and not using radiation. This is the best test. It shows the brain and the spinal cord substance.

You are referred to a specialist after the CAT scan in most cases and they organise any further investigations. The common specialists to be referred to are:

Neurosurgeons  
Neurologists

## HOW IS THIS TREATED ?

This depends on the type and severity of any symptoms.

If you have no symptoms and this has been found by accident then nothing will usually need to be done.

If your symptoms have been transient and do not recur then we may just observe for a while.

If your symptoms are from the pressure on the brainstem and there is no syrxinx then the posterior fossa is decompressed.

If you have no posterior fossa symptoms and there is a syrxinx then the decompression of the posterior fossa will usually treat this.

## IF YOU NEED SURGERY WHAT WILL IT BE ?

### THERE ARE TWO TYPES OF OPERATION

The first is the most usual and the second is to treat any possible Syrxinx if the first does not work.

### POSTERIOR FOSSA DECOMPRESSION

This is where the bone of the foramen magnum is removed from the back and a small part of the upper two cervical vertebra may also be removed. The lining of the brain is opened and a patch is sewn into the defect to allow the whole to expand

### DRAINAGE OF SYRXINX

The syrxinx may be opened directly or a small tube may be placed into it to allow the fluid to drain out. This could be drained into the fluid around the spinal cord or to another site.

## FOR ABOVE PROCEDURES SEE THE RELEVANT PROCEDURE LEAFLET

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