.A.C.A | Syringomyelia Arnold Chiari Association

Registered with the Charity Commission for Northern Ireland NIC 100528

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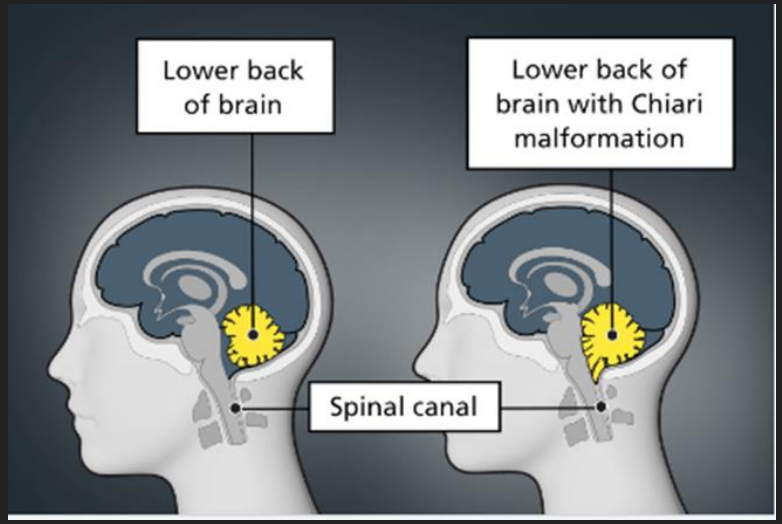
What is Chiari Malformation?

A Chiari Malformation is a complex neurological condition where the bottom part of the brain, the cerebellar tonsils, descend below the opening at the base of the skull, and herniate into the space at the top of the spine, putting pressure on both the brain and spine, and disrupting the normal flow of cerebrospinal fluid (CSF).

(image NHS Uk/conditions)



Types and causes of Chiari malformations

There are two main types of Chiari - Type 1 (CM1) and Type 2 (CM2)

In most cases, **CM1** are congenital, related to **a small posterior fossa** (the space in the skull that holds the cerebellum).

CM2 is associated with the conditions **Spina Bifida and Hydrocephalus,** caused by abnormalities in the structure of the brain and spine before birth.

Chiari can also be **acquired**.

In these cases, Chiari can be caused by a buildup of pressure in the brain eg as a result

of **a tumour, by a problem with the spinal cord**

known as a tethered cord, by leakage of CSF or by an injury.

Chiari Types 3 and 4 are very rare severe embryonic conditions.

Other Chiari types

Chiari Type 0 or 0.5 were only defined recently, in these cases there may be no tonsillar descent and the patient may have a syrinx and chiari-like symptoms.

Chiari Type 1.5 is more advanced than CM1 in addition, the lower part of the brain stem is lower than normal, lying in the upper part of the spinal canal.

Complex Chiari - Some specialists describe a ‘complex Chiari’ when it’s associated with hypermobility syndromes eg Ehlers-Danlos syndrome which is a collagen disorder where the skin and tissue is more lax. Skin does not respond well to surgery, patients have worsening scars, unstable spines, and may need fusions of the neck or spine.

Symptoms

Symptoms can develop at any age, commonly become noticeable in childhood or adults in their early 30's

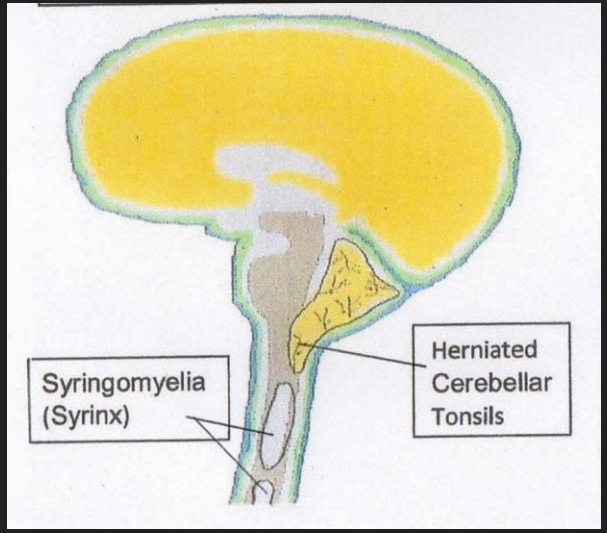
Those who are symptomatic, often experience headaches, and a variety of other symptoms. Valsalva headache is the most common symptom often made worse by straining, coughing, sneezing, laughing, bending over.

Other symptoms include neck pain, trouble sleeping, dizziness, balance problems, tingling/numbness in the arms and hands, hearing and vision issues, cognitive problems, anxiety and depression.

Syringomyelia/syrinx

Some people with Chiari may develop a condition called Syringomyelia (SM).

This is when collection of cerebrospinal fluid (CSF) normally found outside the spinal canal forms a cyst like collection of fluid or cavity in the spinal canal.



This fluid-filled space is known as a syrinx. Over time, a syrinx can expand and elongate, destroying the centre of the spinal cord. As the nerve fibres inside the spinal cord are damaged, a wide variety of symptoms can occur, depending on the size and location of the syrinx.

Symptoms

Symptoms of SM can include:

numbness, muscle weakness, pain, stiffness, unusual sensations (burning or tingling), changes in sensation (loss of pain or temperature sensitivity) and bladder and bowel problems.

In extreme cases, it can cause paralysis.

In some children, scoliosis may be the only symptom, scoliosis meaning curvature of the spine, with a syrinx discovered on MRI scans.

Other causes of a syrinx

SM/syrinx can also occur as a complication **of trauma, meningitis, tumor, arachnoiditis, a tethered spinal cord or spina bifida.** In these cases the syrinx forms in the sections of the spinal cord damaged by these conditions. Some cases of syringomyelia are idiopathic, no known cause.

Syringobulbia is the term used when a syrinx extends into the brainstem. This can affect the nerves in the head, and can cause weakness in the facial muscles, dizziness, involuntary movement of the eyes (nystagmus) and changes in sensation in the face (loss of sensitivity to pain or temperature).

Diagnosis and Treatment

To diagnosis these conditions doctors, usually a **neurologist**, will perform a thorough neurological examination and then carry out **MRI scans.**

Chiari may be diagnosed initially on a **CT scan** at ED or by another specialist.

The patient is then referred to **Neurology** and later to a **Neurosurgeon** to establish if the patient may benefit from surgery. Not every patient with CM1 or SM is a candidate for surgery.

Pain treatment is often the first option.

SACA seeks out resources on pain education and support, and regularly shares this information with our members, we run Mindfulness courses too.
PANI/Versus Arthritis/Healthy Living Centre Alliance - Better Days Pain Support Programme have all been helpful for our members in coping with their chronic pain.

Surgery

The most common type of surgery to treat Chiari malformations is known as a **decompression surgery** carried out at the regional Neurosurgical Unit.

The aim of decompression surgery is to create more room for the bottom of the brain and the brainstem, and to make it easier for the cerebrospinal fluid (CSF) to flow around the brain and the spine.

To quote SACA's medical advisor and patron, Mr Mano Shanmuganathan, Consultant Neurosurgeon, at the last talk he gave to our SACA members – "These are complex conditions, it's doing the right operation at the right time for that patient is important."

What does surgery involve?

The decompression surgery is performed by a neurosurgeon, who cuts out and removes a small piece of bone from the back of the skull at the base.

Parts of the top one or two vertebrae (spinal bones) may also be removed to help create more space.

Sometimes the neurosurgeon will also cut open the thin covering that surrounds the brain and spine (called the dura) and sew in a patch to make it bigger.

The type of patch can vary, but it’s common here for the neurosurgeons to use a synthetic patch.

Possible risks of decompression surgery include:

- stroke or bleeding (haemorrhage)
- nerve or spinal cord damage (causing weakness, numbness, pain or paralysis)
- speech problems
- difficulty swallowing
- memory loss or problems with thinking
- balance problems
- infection / meningitis
- hydrocephalus
- syringomyelia
- seizures
- CSF leak
- pseudomeningocele (a collection of cerebrospinal fluid [CSF] around the surgery site)
- no improvement, recurrence, or worsening of symptoms
- a risk to life

How common are CM and SM?

It is hard to know exactly how common Chiari malformations are.

It has been estimated that **1 child in every 1000 is born with a CM.**

However, because some people don't develop symptoms until adulthood or don't ever develop symptoms, it's likely that the condition is more common than this.

Chiari malformations seem to be more common in women than men.

It was previously thought in North America that the estimated **prevalence of Syringomyelia was 8.4/100 000**

This is why a register of rare diseases is important to our community along with correct coding of our conditions.

Currently both CM and SM are listed on OrphaNet a portal for Rare Diseases, which began in Europe and includes several other countries.

Misdiagnosis and other challenges

Patients with CM and SM may be misdiagnosed with other conditions including: migraine, multiple sclerosis, fibromyalgia and chronic fatigue syndrome.

Currently there are no care pathways or NICE guidelines for the diagnosis and treatment of CM and SM in the UK.

Post-surgery, following several reviews with their neurosurgeon, patients are normally discharged back to their GP.

Patients who are still symptomatic are left fighting for every bit of help and support they can get.

As we straddle Neurosurgery and Neurology, for many patients it is a challenge when seeking care post surgery for new or worsening symptoms, further surgery may not be an option and there are no specialist nurses to offer support here currently.

Chiari patient survey 2018

In a Chiari patient survey* carried out in 2018 by the UK charity Ann Conroy Trust with the help of International charities, there was a total of 741 responses, patients with Syringomyelia or Ehlers Danlos Syndrome were excluded to get clearer results.

- **37% of respondents were diagnosed within 6 months of seeking help**
- **18% diagnosis took 6 years or longer**
- **59% did not feel that the specialist who diagnosed their condition understood it.**
- **48% of the respondents were told their Chiari was an incidental finding,**
- **52% of cases their Valsalva headaches resolved following surgery**
- **38% of respondents reported their other symptoms improved significantly,**
- **44% reported slight improvement.**
- **18% reported no improvement**

*"Presenting symptoms of Chiari Malformation as seen from the patient's perspective" – Burton L, Brereton G, Brereton N, Flint G

About SACA

Syringomyelia Arnold Chiari Association (SACA) is a small, registered charity run by volunteers, based in Northern Ireland.

Our original support group was founded by Frank and Nuala Somers in 1990 and was first known as Ann's Neurological Trust. SACA later gained charitable status in 2014.

We have 40+ full members and a moderated Facebook group with 180+ members.

What we do

SACA provides information and support to those living with Chiari Malformations, Syringomyelia and associated conditions in Northern Ireland; and the families and friends who support them.

We aim to:

- remove the isolation families face when they receive a diagnosis
- support more people who are impacted by these conditions

Our support services

- We operate a **Helpline 078 26 004 008** and provide virtual support via Facebook, messenger, email and zoom
- We produce a members' newsletter
- We arrange face to face meet ups and social events

All of our Trustees have direct experience of these conditions.

Our activities are supported by volunteers donating time and resources.

To date our only financial support has been from annual membership fees, fundraising and generous donations.

Research

Research is ongoing all over the world to learn more about these conditions,

A **UK Chiari 1 Study** supported by the British Syringomyelia-Chiari Group and funded by the Ann Conroy Trust is running in 19/20 neurosurgical centres throughout the UK, it's primary objective: To determine the patient- or parent-reported, health-related quality of life in patients with a new diagnosis of CM1 managed either conservatively or surgically at 12 months follow up.

International Collaboration

2017 Long Island, New York ; 2018 Birmingham; 2019 Milan; July 2025 Liverpool - Syringomyelia 2025

The Milan Chiari / Syringomyelia Consensus Conference brought together the international experts to negotiate consensus views over 60 provisional statements about the diagnosis and treatment of Chiari Malformation.

Paper published "**Diagnosis and treatment of Chiari malformation and syringomyelia in adults: international consensus document**" https://pubmed.ncbi.nlm.nih.gov/34129128/

A European Network of Chiari & Syringomyelia Associations – Syrenet was also established.

- Database project with the Italian, French and Spanish groups
- Chiari gene research project.

Finally

Meet Harvey, a young healthy adorable Cavalier King

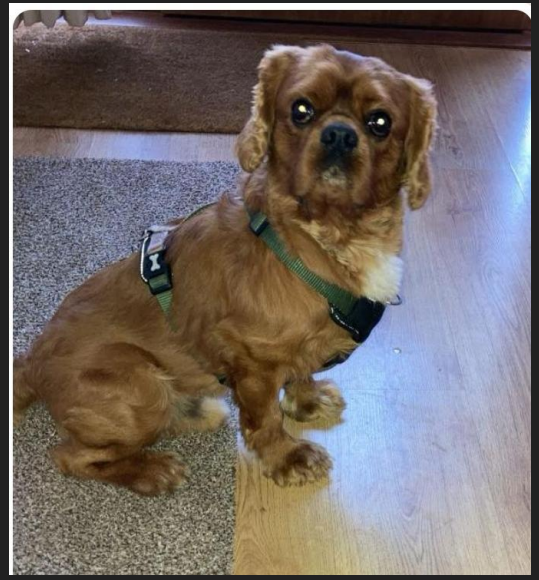
Charles Spaniel, belonging to one of our SACA trustees.

Unfortunately a Chiari-like

Malformation and Syringomyelia are

commonly found in this breed along with several

other toy dog breeds. Their brains are too large for their skulls.



This is not pleasant for the dogs nor their owners. But it has given a naturally occurring animal model to investigate the origins of syringomyelia. We hope the continued research will help increase the understanding of these chronic conditions.

Updated 30/5/2025 MM