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## Syringomyelia

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### Summary

- Syringomyelia is the formation of a fluid-filled cyst (syrinx) within the spinal cord.
  - As the cyst gets larger, it presses on the spinal cord and interferes with the transmission of nerve impulses.
  - Causes include congenital (from birth) brain defects, spinal cord trauma and infection.
  - Treatment options include surgery.
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Syringomyelia is the formation of a fluid-filled cyst (syrinx) within the spinal cord. The most common site is the cervical spine in the neck region. As the syrinx grows, it presses on the spinal cord and interferes with the transmission of nerve impulses.

The condition affects approximately eight out of every 100,000 people, and men are more at risk than women (for reasons unknown). The average age of onset is about 30 years.

There are broadly three types of syringomyelia. The most common type is associated with congenital (from birth) brain abnormalities, while the second type develops as a complication following spinal trauma, infection or tumour. The third type is caused by unknown factors and is called idiopathic syringomyelia.

Without treatment, syringomyelia may result in paraplegia or quadriplegia. Treatment options include surgery.

### Symptoms of syringomyelia

The symptoms vary from one person to the next, depending on the severity and location of the cyst, and the underlying cause. Symptoms can develop slowly and this may delay the diagnosis.

Symptoms include:

- pain and weakness in the back, shoulders, arms or legs
- headaches
- reduced skin sensations, such as not being able to feel extremes of heat and cold
- loss of sensation in the hands, including sensations of pain
- muscle atrophy (wasting), usually beginning in the hands and spreading to include the arms and shoulders
- severe pain in the shoulders and neck
- reduced bowel and bladder control (in the later stages)
- sexual dysfunction.

### Types of syringomyelia

The brain and spinal cord (central nervous system) are surrounded by a clear fluid known as cerebrospinal fluid. Some of the functions of cerebrospinal fluid include nourishing and cushioning the central nervous system.

The three broad categories of syringomyelia include:

- **congenital brain defects** – such as Arnold-Chiari malformation. The bottom part of the brain (cerebellum) lies in the upper part of the neck, instead of within the skull. This obstructs the flow of cerebrospinal fluid. It may be associated with hydrocephalus.
  - **injury to the spinal cord** – including trauma, tumours and infections (such as meningitis, HIV), and where there is severe cord compression.
  - **idiopathic syringomyelia** – the cause is unknown.
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## Formation of the syrinx

Cerebrospinal fluid normally circulates around the brain and spinal cord. If the flow of cerebrospinal fluid is obstructed and turned back on itself, a cyst (syrinx) may form within the delicate tissue of the spinal cord.

The cyst may be small at first. It is thought that the differences in pressure between the brain and spinal cord gradually force cerebrospinal fluid into the cyst. Over time, the cyst grows larger and presses on the surrounding spinal cord nerves. This can eventually damage the nerves.

Syringomyelia usually progresses slowly over years, but symptoms can sometimes advance rapidly after straining or coughing. This is because the temporary increase in cerebrospinal pressure forces fluid into the cyst.

## Complications of syringomyelia

The brain stem is the bridge between the brain and the spinal cord. Messages relay from the brain to the motor and sensory nerves of the body, and vice versa, in a constant 'conversation'. In some cases of syringomyelia, the cyst grows into the brain stem and interferes with vital functions such as breathing and heartbeat. This complication is called syringobulbia.

Some of the symptoms of syringobulbia may include:

- swallowing difficulties
- involuntary flickering of the eyes
- tongue 'wasting', which results in speech problems.

## Diagnosis of syringomyelia

The symptoms of syringomyelia can mimic those of other conditions, so diagnosis can be difficult.

Some of the tests for syringomyelia include:

- medical history
- physical examination
- magnetic resonance imaging (MRI)
- computed tomography (CT) scans
- electromyography – to test muscle strength
- myelogram – an x-ray requiring injection of contrast dye into the cerebrospinal fluid (unusual).

## Treatment for syringomyelia

Treatment depends on the cause and severity of the condition, but may include:

- Wait-and-see approach – if the cyst is small and not causing any symptoms, your doctor may prefer to simply monitor the condition.
- Medications – syringomyelia can't be treated with medication, but pain-relievers and muscle relaxants may help to ease some of the symptoms.
- Shunting – a thin tube is inserted into the cyst to drain the fluid into the abdominal cavity. The shunt contains a one-way valve to prevent backflow. This type of treatment is often used if hydrocephalus (build-up of cerebral fluid inside the brain) is also present.
- Surgery – to treat the underlying cause. If it is a tumour, then this should be removed. With the Arnold-Chiari malformation, surgery aims to give the cerebellum more room in the base of the skull and neck, and improve the flow of cerebrospinal fluid at the same time. If there is hydrocephalus present, then this should be treated first.

## Where to get help

- Your doctor
- Neurologist

## Things to remember

- Syringomyelia is the formation of a fluid-filled cyst (syrinx) within the spinal cord.
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- Causes include congenital (from birth) brain defects, spinal cord trauma and infection.
- Treatment options include surgery.

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