Key Points

1. Over the years there have been many theories on why syrinxes form, many of which have focused on the flow of CSF being blocked by the cerebellar tonsils
2. One recent theory holds that the tonsils act like a piston and with every heartbeat drive down into the spinal area and create large pressure waves of CSF; this drives CSF into the spine and creates a syrinx
3. This paper disputes this claim by pointing out that the pressure inside a syrinx is higher than the outside CSF and that the syrinx fluid and CSF are not identical
4. New theory puts forth that activities such as coughing, straining, standing up, etc., create a pressure imbalance between the skull and spine
5. This leads to abnormal expansion and contraction of blood vessels in the spine and cause stress and damage to the spine
6. This damage leads to a breakdown of the blood-spine barrier and allows fluid to leak from blood vessels and create a syrinx
7. While this theory fits with current observations, it needs to be proven experimentally

Definitions

capillary - the smallest of the body's blood vessels, through which oxygen, nutrients, and waste products can pass into cells

caudal - towards the tail; when referring to the spine, down

cerebellar tonsils - portion of the cerebellum located at the bottom, so named because of their shape

cerebrospinal fluid (CSF) - clear liquid in the brain and spinal cord, acts as a shock absorber

cervical - relating to the upper

New Theory On How Syrinxes Form

Many theories on syrinx formation have come and gone over the years. In the very early days, syrinxes were thought to be like tumors. More recently, researchers have focused on the flow of cerebrospinal fluid and how a Chiari malformation disrupts this flow.

In a normal person, when the heart beats, blood rushes into the brain. Since the skull is rigid and has a fixed volume, the inrushing blood pushes CSF out of the skull area and into the spinal area. During the second phase of the cardiac cycle, this is reversed; blood flows out of the skull area and CSF flows back in from the spinal area. In people with Chiari, the cerebellar tonsils block this natural flow of CSF between the brain and spinal areas.

One of the more recent theories on syrinx formation is the so-called piston theory. The piston theory states that with every heartbeat, the cerebellar tonsils - which are jammed into the top of the spine acting like a plug - are pushed down into the spinal area. The result is like a piston repeatedly driving into the CSF of the spine. This in turn creates pressure waves of CSF which drives CSF into the spinal cord itself and forms a syrinx. While many doctors and researchers have accepted the piston theory - at least in part - the theory is not universally accepted and has not been proven definitively.

Now, Dr. David Levine, a Professor of Neurology at NYU, has published a critique of the piston theory (and other existing theories) in the May issue of the Journal of the Neurological Sciences, and proposes his own theory on syrinx formation.

In his paper, Dr. Levine points out that the current theories, including the piston theory, are difficult to reconcile with many physical findings regarding syrinxes. Specifically, he points out that studies have shown that the pressure of the fluid inside a syrinx is higher than the surrounding CSF. If CSF were being forced into the spine from the outside, the opposite would be true.

Additionally, Dr. Levine points out that autopsies have revealed that blood vessels around syrinxes are enlarged with thickened walls. Again, if CSF were being pushed in from the outside, the vessels should be smaller, or even crushed, from the pressure. Autopsies have also revealed the presence of gliosis - a thickening of the connective tissue in the nervous system - which the current theories fail to account for. Finally, Levine points out that while both CSF and syrinx fluid are clear and colorless, a study in 1969 showed that the protein content of the two fluids in 7 cases was significantly different.

In contrast to the piston theory, Dr. Levine believes that syrinxes form from fluid that leaks out of blood vessels due to damage to the spine. First, Levine describes how in people with Chiari actions such as coughing, sneezing, straining, and even the heart beating, create a pressure difference between the brain area - intracranial pressure - and the spinal area - intraspinal pressure. Since the cerebellar tonsils are blocking the flow of CSF out of the skull, the body is essentially divided into two compartments, above the block and below the block. Levine shows how actions such as coughing lead to abrupt changes in pressure just above and below the block, with the pressure in the brain being abnormally high and the pressure in the spinal area being abnormally low.

This sudden change in pressures that occurs with events such as coughing have a direct impact on the blood system of the spine. Since the pressure of the blood inside the veins and capillaries stays the same, the increased pressure outside of the blood vessels in the skull area puts pressure on them and shrinks them. Below the tonsils, in the spinal area, the pressure inside the blood vessels is higher than the pressure of the CSF outside of them, so the vessels expand. Eventually, some time after an event like a cough, the pressures even out. But this dramatic expansion and contraction happens every time a Chiari person coughs, sneezes, or even stands up - activities that patients all know aggravate symptoms.

According to Levine's theory, this repeated, sudden, and uneven expansion and contraction puts real stress on the structure of the spine. He likens it to what happens to roads in response to repeated temperature changes - the stress creates potholes. This stress damages the tissues of the spine and leads to the gliosis that is found associated with a syrinx.

His theory goes on further to state that the repeated expansion of the blood vessels in the spine, and the resulting stress, cause the capillaries to rupture and leak plasma - the fluid of the blood - into the spine. This fluid collects and forms a syrinx. Levine believes that plasma that is filtered through a partially ruptured blood vessel would match the characteristics of the fluid found in syrinxes.

Although Levine does a nice job of citing existing evidence in support of his theory, only time, experiments, and the critical eye of fellow scientists will determine whether this will finally be a universally accepted theory on syrinx formation. If it is proven to be true, it will be interesting to see how it effects the treatment of Chiari and syringomyelia.

part of the spine, neck area

glia - connective tissue of the brain and spine; cells which provide structure and serve other functions

gliosis - abnormal thickening and hardening of the glia

intracranial pressure - the pressure of the CSF inside the skull

intraspinal pressure - the pressure of the CSF inside the spinal area

plasma - the liquid part of blood in which blood cells float

rostral - towards the mouth; when referring to the spine, up

syringomyelia (SM) - neurological condition where a fluid filled cyst forms in the spinal cord

syrinx - fluid filled cyst in the spinal cord

thoracic - relating to the middle part of the spine, or chest area

Valsalva maneuver - straining

Meet The Researcher: Dr. David Levine

Qualifications:

- Professor of Neurology at NYU
- Associate Director, Neurology Service, Tisch Hospital
- Director, Transcranial Doppler Laboratory
- Board Certified Neurologist

Education:

- Med School - Harvard Medical School
- Residency - Massachusetts General Hospital (Neurology)
- Postgrad - Research Associate, National Institutes of Health
- Clinical Fellowships -
  Massachusetts General Hospital (Neurology)

Selected Publications:

- Levine DN. The pathogenesis of syringomyelia associated with lesions at the foramen magnum: a critical review of existing theories and proposal of a new hypothesis.*J Neurol Sci.* 2004 May 15;220(1-2):3-21.
- Levine DN; Rapalino O. "The pathophysiology of lumbar puncture headache". *Journal of the neurological sciences* 2001 Nov 15;192(1-2):1-8
- Levine DN. "The pathogenesis of normal pressure hydrocephalus: a theoretical study". *Bulletin of mathematical biology* 1999;6:875-916

Disclaimer: This publication is intended for informational purposes only and may or may not apply to you. The editor and publisher are not doctors and are not engaged in providing medical advice. Always consult a qualified professional for medical care. This publication does not endorse any doctors, procedures, or products.

© 2003-2020 C&S Patient Education Foundation