**Hydrocephalus Associated with Multiple Tarlov Cysts**

Mieke Hulens MD, PhD[1], Wim Dankaerts MT, PT, PhD[1], Ricky Rasschaert MD[2], Frans Bruyninckx MD[3], Ingeborg StalmansMD, PhD[4], Greet Vansant MSc, PhD[5], Peter De Mulder MD, PhD[6]

**Affiliations**

[1]Musculoskeletal Rehabilitation Research Unit, Department of Rehabilitation Sciences, Faculty of Kinesiology and Rehabilitation Sciences, University of Leuven, Tervuursevest 101, 3001 Heverlee, Belgium

[2]Department of Neurosurgery, AZ Rivierenland, Kasteelstraat 23, 2880 Bornem, Belgium

[3]Clinical Electromyography Laboratory, University Hospitals UZ Leuven, Herestraat 49, 3000 Leuven, Belgium

[4]Department of Neurosciences, Ophthalmology Research Group, University of Leuven KU Leuven), Leuven, Belgium and Department of Ophthalmology, University Hospitals UZ Leuven, Herestraat 49, 3000 Leuven, Belgium

[5]Department of Social and Primary Health Care, Public Health Nutrition, University of Leuven, Herestraat 49, 3000 Leuven, Belgium

[6]Department of Anesthesiology- Pain Therapy, Imelda Hospital, Bonheiden, Belgium

**Corresponding author**

Mieke Hulens, Overwegstraat 14, 3051 Sint-Joris-Weert, Belgium

Tel.: +32 478 338003, Fax: +32 16 470559, Email: miekehulens@skynet.be

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## ABSTRACT

Tarlov cysts (TCs) consist of dilated nerve root sheaths filled with cerebrospinal fluid (CSF) and are most frequently found in the sacrum. It is estimated that 25% of detected TCs cause chronic pain and intestinal and urogenital symptoms due to compression of the sacral nerve root fibers inside the TC. Unfortunately, symptomatic TCs are frequently overlooked.

It is assumed that TCs result from pathologically increased hydrostatic pressure (HP) in the dural sac that forces CSF into the nerve root sheaths. We hypothesize that in patients with TCs, increased spinal hydrostatic pressure is always associated with increased intracranial pressure. This hypothesis of increased cerebrospinal pressure might explain why patients with sacral TCs frequently report distant symptoms, such as headaches and pain in the neck and arms.

In this paper, we describe a case report that provides evidence for this hypothesis.

A 30-year-old man presented for the first time in our clinic complaining of lower back, leg, thoracic, neck, and arm pain; headaches; and bladder, bowel, and sphincter symptoms.

He was born prematurely and suffered cerebral intraventricular bleeding followed by progressive hydrocephalus. Progression was stabilized with acetazolamide and lumbar punctures. At 19 years of age, his head circumference had further increased and he reported back pain and headaches. Fundoscopy showed no papilledema, and lumbar puncture for CSF evacuation improved the headaches and back pain. The former medical team chose not to insert a ventriculo-external shunt.

Brain magnetic resonance imaging (MRI) showed significant dilation of all the ventricles. No CSF flow obstruction between the ventricles was observed.

Surprisingly, MRI of the lumbar and sacral spine showed multiple large TCs.

This case report indicates that hydrocephalus with a patent aqueduct may be associated with TCs because the increased intracranial pressure is transferred to the spinal canal. While increased intracranial pressure causes dilation of the ventricles, the associated increased spinal pressure may cause dilation of multiple spinal nerve root sheaths to form TCs. Furthermore, while the increased volume of the ventricles gradually compresses the neurons and axons of the brain against the bony skull, simultaneously, the increased pressure inside the nerve sheaths may also gradually compress the neurons and axons located inside the dorsal root ganglia and spinal nerves, resulting in neuropathic pain, sensory abnormalities, and neurogenic bladder and bowel symptoms.

Hydrocephalus patients reporting neuropathic pain should be screened for the presence of TCs.

## INTRODUCTION

Tarlov cysts (TCs), or perineural cysts, are the most common type of meningeal cysts and consist of dilated nerve root sheaths containing neurons and nerve fibers (axons) and cerebrospinal fluid (CSF) (1). Dilation is assumed to result from pathologically increased hydrostatic pressure (HP) in the spinal canal, forcing CSF from the dural sac into the nerve root sheaths (2-8). A recent study of 1100 magnetic resonance imaging (MRI) scans detected TCs in the sacral spine in up to 13% of the patients; the affected patients were predominantly women. No TCs were seen in subjects <18 years of age (9). Approximately 25% of TCs are symptomatic at the time of their discovery. Symptoms are often debilitating and include radicular pain; pelvic and perineal pain; and bladder, bowel and sphincter dysfunction (10).

We hypothesize that if TCs are due to increased HP in the dural sac, then the intracranial pressure will also be increased in these patients. As a consequence, at least a proportion of the patients with symptomatic TCs (STCs) should also have intracranial hypertension, which may explain the high prevalence of headaches in patients with TCs (up to 73%) (11).

This case report provides evidence that increased intracranial pressure, e.g., hydrocephalus, is associated with increased HP in the spinal canal and consequently causes dilation of the nerve root sheaths, leading to the formation of TCs.–

## CASE DESCRIPTION

A 30-year-old man presented in our clinic with lower back pain. The medical record revealed that he was born at 28 weeks of gestation. His birth weight was 1.390 kg and his head circumference was 27 cm. At 2 weeks of age, he suffered cerebral intraventricular bleeding followed by progressive posthemorrhagic dilation of the lateral ventricles above the 97$^{th}$

percentile. Acetazolamide was administered and a series of lumbar punctures were performed. During treatment, ventriculomegaly was stabilized and the patient's head circumference remained in the 97$^{th}$ percentile. During ambulatory control, his treatment was continued.

At 6 months of age, treatment with acetazolamide was ceased; however, this caused a further increase in head circumference. Therefore, the acetazolamide treatment was reinstated to avoid insertion of a ventriculo-atrial shunt. Acetazolamide treatment ceased at 2 years of age.

He showed spasticity of the legs, likely due to neonatal cerebral bleeding. His mental development was normal.

At 12 years of age, his head circumference had enlarged to 9 standard deviations (SDs) above the mean. MRI showed pronounced dilation of the lateral ventricles and third ventricle. There was normal CSF flow through the cerebral aqueduct and the fourth ventricle was moderately dilated. Intellectual development and school performance remained normal.

At the age of 19 years, he complained of back pain and headaches. Fundoscopy showed no papilledema, and lumbar puncture and evacuation of CSF improved both the headaches and back pain.

Apparently, during these events the former medical team chose not to insert a ventriculo-external shunt.

When he first presented in our clinic, he was 30 years old and complained of lower back pain, left-sided thoracic pain in dermatomes D5-D6, leg and feet pain and stiffness, neck pain, upper arm pain, and headaches. Additionally, he reported bladder (urinary frequency and urgency), bowel (constipation alternated with diarrhea and abdominal cramps), and sphincter symptoms (painful pressure in the anal sphincter).

Clinical examination revealed hypersensitivity of the right foot sole and lower back. He had bilateral symmetrical hyperreflexia of the Achilles and knee tendons and normal symmetrical strength in the legs.

### MRI of the Brain

Brain MRI showed significant dilation of the lateral and third ventricles (Fig. 1A and B) (obstructive hydrocephalus) and a less pronounced dilation of the fourth ventricle (Fig. 1C). There was a narrowing of the midportion of the aqueduct. Flow voids were seen over the foramen Magendie and foramen magnum, indicating that there was no obstruction of the CSF flow between ventricles (Fig. 1).

### MRI of the Lumbosacral Spine

Multiple large TCs were seen on the lumbar and sacral nerve roots. The TC on the right nerve root S2 was located near the spinal canal, whereas the other TCs were located more distally along the nerve roots (Fig. 2).

## DISCUSSION

This case study describes a patient with hydrocephalus and a patent aqueduct who also developed TCs.

Hydrocephalus without aqueduct stenosis may be due to the overproduction of CSF in the choroid plexus of the ventricles or to the under-resorption of CSF in cerebral and spinal arachnoid granulations and the glymphatic and lymphatic system (12).

Excess CSF in the skull results in increased intracranial pressure. As a consequence, CSF is forced from the lateral ventricles through the foramina Monroi into the third ventricle and through the aqueduct into the fourth ventricle. In our patient, the third and fourth ventricles were dilated. Ultimately, CSF is forced towards the subarachnoid space of the spinal canal and the subarachnoid space of the spinal nerves. Because of the upright position of humans, pathologically increased HP also forces CSF caudally to the lumbar and sacral nerves (6, 7).

Increased pressure in the subarachnoid space of the spinal nerves causes leaking of the CSF between the endoneurium and perineurium near the dorsal root ganglion, where the permeability of the blood-nerve barrier is higher than that of the spinal nerves (13). This increased permeability may explain why TCs are usually located at or near the dorsal root ganglion. Microscopic studies have shown that TCs consist of CSF trapped between the endoneurium and perineurium layers of the nerve root sheaths (7).

In our patient, only one TC was located near the dorsal root ganglion (proximally on the nerve root), whereas the other TCs were located distally on the nerve roots. Some cysts presented multilocular aspects along the nerve root. It is possible that the TCs in this patient were located remotely from the dural sac because in infancy, collagen fibril elasticity is higher than that in adulthood (14). As a consequence, connective tissue of the nerve root sheaths was less resistant to internal pressure and was more likely to expand.

Cases of intracranial hypertension associated with TCs have been described previously (4, 6, 15). A thorough search of the relevant literature yielded no cases of hydrocephalus associated with TCs.

Because 25% of TCs are symptomatic at the time of discovery, this patient's peripheral symptoms, including his neuropathic leg pain, sensory abnormalities in the legs, and bowel and

bladder problems, may have been due to the compression of the lumbar and sacral nerves inside the TCs.

However, because of his intracranial bleeding, it was clinically difficult to differentiate between the central and peripheral origin of his neurological symptoms. Electromyography (EMG) of the lumbar and sacral myotomes and an electrophysiological examination of the anal reflex might differentiate between the origins (16); however, these examinations were not available for this patient.

## CONCLUSION

This case report confirms our hypothesis that increased intracranial pressure is associated with increased intraspinal pressure. While increased intracranial pressure causes dilation of the ventricles, the associated increased spinal pressure may cause dilation of multiple spinal nerve root sheaths to form TCs. Furthermore, while increased intracranial pressure such as in hydrocephalus gradually compresses the neurons and axons in the brain, it probably also gradually compresses the neurons in the dorsal root ganglia and the axons in the spinal nerves, resulting in widespread neuropathic pain, sensory abnormalities and neurogenic bladder and bowel symptoms from the compression of the spinal nerves.

Patients with hydrocephalus reporting unexplained pain should be screened for the presence of TCs and conversely, all patients with TCs should be screened for intracranial hypertension.

## Conflict of interest statement

None

## Data availability

Not applicable

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## FIGURE CAPTIONS

**Fig. 1.** T2-weighted MRI images of the brain. Fig. 1A and B show the dilated lateral ventricles. Fig. 1C shows the dilated third (vertical arrow) and fourth (horizontal arrow) ventricles.

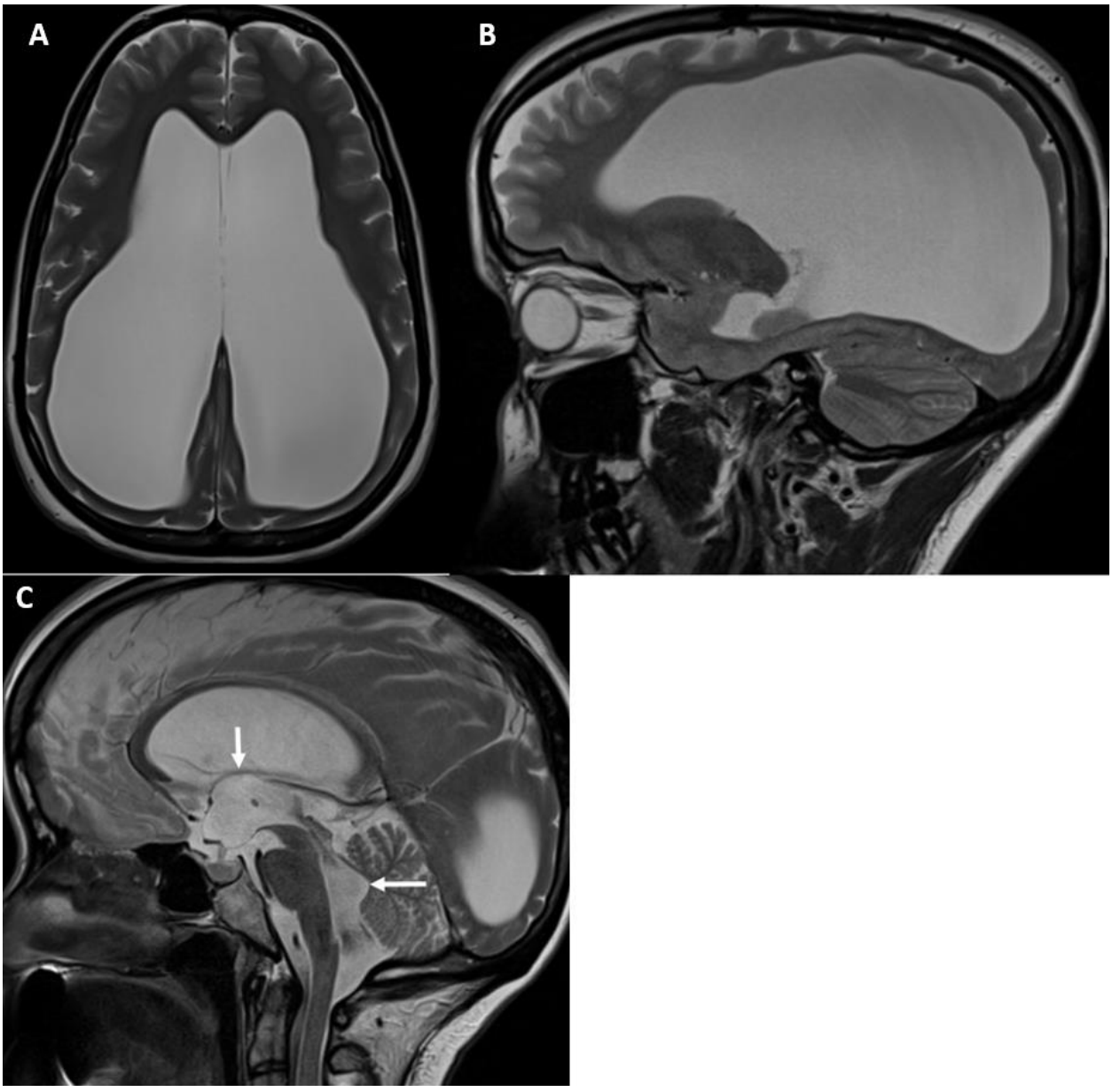

**Fig. 2.** T2-weighted coronal MRI myelography images of the lumbar spine show multiple TCs on the lumbar and sacral nerve roots distant from the spinal canal, except for the right nerve root S2 (large arrow) located at the dorsal root ganglion. The latter TC is located closer to the spinal canal than the former TCs. Some TCs along the nerve root appear to be multilocular (small arrows).