



Stichting OverdrukSyndroom NL

Als de druk te hoog wordt

Diagnosis-checker

Goal: recognition that the patient's pain is disabling neuropathic pain due to a physical condition and to prevent further unnecessary ineffective and potentially harmful interventions

This diagnostic checker has been drawn up - supported by scientific research - to achieve a faster diagnosis with regard to **symptomatic Tarlov cysts, which are dilations of nerve roots sheaths in the spinal cord, resulting from **pathologically elevated pressure**. Not only external compression of nerve roots by large Tarlov cysts, but also the internal pressure in cysts irritates the nerve fibers in the nerve. As a result, communicating cysts and small cysts, as well as dilated nerve roots sheaths, can be symptomatic. The hydrostatic increased pressure allows Tarlov cysts to come back after an eventual surgery. There is objective evidence of nerve fiber damage in communicating cysts. Microscopic examination revealed that the walls of the Tarlov cysts consisted of compressed degenerated nerve fibers and damaged myelin sheaths.**

In patients with persistent sacral, perineal, pelvic or leg pain, symptomatic Tarlov cysts should be included in the differential diagnosis. The **Dutch professor G. Padberg, neurologist in Maastricht, already wrote on **October 10th, 1959** in the Dutch. T. Medicine 103. II. 41, about "Perineural Nerveroot Cysts" which he already argued at that time, it was insufficiently known that it can cause low back pain, sciatica and neurological disorders, by physicians in general.**

1. Extensive medical history, also specific questions about:
-> faecal and urinary incontinence
-> bladder retention, bowel symptoms
-> genital and perineal pain
-> cervical, dorsal, thoracic pain
-> pain when sitting and standing
-> pain with exertion
-> headache

Is the patient familiar with a connective tissue disorder?

Note: Tarlov cysts are more often found in patients with genetic connective tissue disorders such as the hypermobility type Ehlers-Danlos syndrome (EDS) and Marfan syndrome. Due to weakness of the connective tissues, the nerve root sheaths in these patients are more sensitive to dilation and in patients with EDS also often elevated intracranial pressure is present.

4. **Electrodiagnostic conductivity test:**
- sensory sural nerves (containing nerve fibers of nerve roots S1 and S2);
- motor peroneal nerves
- S1 Hoffman reflexes (the electrophysiological equivalent of the Achilles tendon reflex)
Needle EMG:
L3 to S3-S4 myotomes
(L3 vastus medialis muscle; L4 vastus lateralis muscle; L5 extensor digitorum muscle; L4-L5 tensor fascia lata muscle and tibialis anterior muscle; S1 gastrocnemius muscle medial head; S2 Tibial nerve innervated intrinsic foot muscle and S3-S4 myotomes (external anal sphincter).

Analysis of S3-S4 ano-anal reflex (the electrophysiological equivalent of the ano cutaneous reflex) (reflex to prevent faecal incontinence)

2. Pinprick test at the top and lower extremities, dorsal at the painful zone.
(this does not always provide a definite answer - important that NCS / EMG, see point 4, is carried out)

Idiopathic Cerebrospinal Pressure Dysregulation Syndrome & Tarlov cysts

3. MRI lumbar / sacral and cervical - T1 and T2 weighted - sagittal, axial and coronal recorded images

Note: if axial and coronal images are not recorded, smaller TC's may be over looked. MRI studies of the lumbo-sacral spine show a prevalence of 9.1% -13% of smaller TCs.

Note: Symptoms in Tarlov cyst and idiopathic cerebrospinal pressure dysregulation syndrome patients, may be similar to Idiopathic Intracranial Hypertension. Tarlov cyst patients do not always have papilloedema. However, papilledema is no longer necessary to make the diagnosis.

Clearly visible on MRI:
- large Tarlov cysts, which can cause bone erosion
Often overlooked:
- smaller Tarlov cysts or dilated nerves root sheaths

Most MRI and EMG investigations regarding to lower backpain and leg complaints focus on the L5/S1 myotomes. These studies are NOT sufficient in case of symptomatic Tarlov cysts and/or idiopathic cerebrospinal pressure dysregulation syndrome

In addition, the limit of 20 cm H2O defining intracranial hypertension, is probably too high. There may be a continuum between normal and increased intracranial pressure (normal values 5-15 cm H2O). American research has shown, that in patients with connective tissue disorders a pressure of 17 or 18 cm H2O, measured at lumbar puncture, can cause debilitating symptoms.

If after extensive anamnesis the pain/symptoms of the patient is similar with the cysts location, the cyst/cysts may be considered symptomatic.
An EMG of the sacral nerve areas is not necessary to make a diagnosis, but can give a definite answer, in case of doubt.

Extra point of attention:
Small Fiber Neuropathy
Research has shown that 50% of fibromyalgia and EDS patients has been diagnosed with small fiber neuropathy.
The diagnosis of fibromyalgia and/or EDS is also often found in Tarlov cyst patients

Note: involvement of the cranial nerves. It has been noted, that the perineural spaces of various cranial nerves, including smell, optic, trigeminal and auditory nerves, show multiple lymphatic pathways of CSF drainage to the lymph nodes. When the cerebrospinal pressure increases, cerebrospinal fluid is forced in the skull nerve root sheaths. This can endanger the blood flow or cause mechanical pressure on the neurons or axons, during their intracranial course.

Note: the worst pain is not always in the same dermatome or ipsilateral to the location of the largest cyst seen on the MRI. Patients may experience worse contralateral pain relative to the side of the largest cyst. Not only larger valved Tarlov cysts, but also smaller (communicating) Tarlov cysts can cause severe pain and nerve damage, so one cannot conclude if symptoms / pain are not corresponding with the largest cyst, that the cause has a different origin. Hence, the extensive EMG / NCS is: measuring is knowing in this case.

This information has been established from the following scientific publications:
https://www.researchgate.net/publication/333025384_Symptomatic_Tarlov_cysts_are_often_overlooked_ten_reasons_why_a_narrative_review
https://www.researchgate.net/publication/318094780_Electromyography_and_A_Review_of_the_Literature_Provide_Insights_into_the_Role_of_Sacral_Perineural_Cysts_in_Unexplained_Chronic_Pelvic_Perineal_and_Leg_Pain_Syndromes
<https://www.dovepress.com/the-link-between-idiopathic-intracranial-hypertension-fibromyalgia-and-peer-reviewed-fulltext-article-JPR>
https://www.researchgate.net/publication/337584451_Idiopathic_intracranial_hypertension_is_not_idiopathic_proposal_for_a_new_nomenclature_and_patient_classification