



Symptomatic Tarlov cysts are often overlooked: ten reasons why—a narrative review

Mieke Hulens^{1,8} · Ricky Rasschaert² · Frans Bruyninckx³ · Wim Dankaerts¹ · Ingeborg Stalmans^{4,5} · Peter De Mulder⁶ · Greet Vansant⁷

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Abstract

Purpose Tarlov cysts (TCs) are dilations of nerve roots arising from pathologically increased hydrostatic pressure (HP) in the spinal canal. There is much controversy regarding whether these cysts are a rare source of pain or often produce symptoms. The aim of this review was to identify the reasons that symptomatic TCs (STCs) are easily overlooked.

Methods The literature was searched for data regarding pathogenesis and symptomatology.

Results TCs may be overlooked for the following reasons: (1) STCs are considered clinically irrelevant findings; (2) it is assumed that it is clinically difficult to ascertain that TCs are the cause of pain; (3) MRI or electromyography studies only focus on the L1 to S1 nerves; (4) TCs are usually not reported by radiologists; (5) degenerative alterations of the lumbosacral spine are almost always identified as the cause of a patient's pain; (6) it is not generally known that small TCs can be symptomatic; (7) examinations and treatments usually focus on the cysts as an underlying mechanism; however, essentially, increased HP is the main underlying mechanism for producing symptoms. Consequently, STCs may relapse after surgery; (8) bladder, bowel and sphincter dysfunction are not inquired about during history taking. (9) Unexplained pain is often attributed to depression, whereas depression is more likely the consequence of debilitating neuropathic pain. (10) The recognition of STCs is subject to gender bias, confirmation bias and cognitive dissonance and unconscious bias in publishing.

Conclusion There are several reasons STCs are underdiagnosed, mostly due to persistent misconceptions and biases.

Graphical abstract

These slides can be retrieved under Electronic Supplementary Material.

Key points

1. Symptomatic Tarlov cysts (STCs) can be the cause of debilitating pain, however, are commonly overlooked
2. There are several reasons STCs are underdiagnosed, mostly due to persistent misconceptions and biases.
3. Examinations and treatments usually focus specifically on the cysts as an underlying mechanism for producing symptoms; however, essentially, increased HP may be the main underlying mechanism.

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Reasons Tarlov cysts may be overlooked

1. STCs are traditionally considered clinically irrelevant
2. It is assumed that it is clinically difficult to ascertain that TCs are the cause of a patient's pain
3. Diagnostic studies for low back pain and sciatica only focus on L1 to S1 nerves
4. TCs are usually not reported by radiologists
5. Degenerative alterations are almost always identified as the cause of the pain
6. It is not generally known that small TCs can be symptomatic
7. The focus is usually only on the cysts and not on the increased hydrostatic pressure as an underlying mechanism for producing symptoms
8. Bladder, bowel and sphincter dysfunction are not inquired
9. Unexplained pain is often attributed to depression, whereas depression is more likely the consequence of debilitating neuropathic pain.
10. The recognition of STCs may be subject to gender bias, confirmation bias and cognitive dissonance, and unconscious bias in publishing.

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Take Home Messages

1. In patients with TCs, the mechanism for producing symptoms is probably not only external compression of nerve roots by large valved cysts. Apparently, the main underlying mechanism is increased hydrostatic pressure causing internal pressure which irritates and compresses the axons inside the nerve roots.
2. As a consequence, communicating cysts and small cysts may be symptomatic, and STCs relapse after surgery.
3. From our current understanding of the pathophysiology and symptomatology of STCs, it is clear that STCs should be part of the differential diagnosis of low back pain and sciatica.

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Keywords Tarlov cysts · Meningeal cysts · Unexplained pain · Chronic pain · Intracranial hypertension

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✉ Mieke Hulens
miekehulens@skynet.be

Extended author information available on the last page of the article

Rationale

Tarlov cysts (TCs) or perineural cysts are dilations of nerve roots at the dorsal root ganglion. Cystic structures in the sacrum observed during autopsies were first described in 1902 [1].

In 1938, when Tarlov [2] encountered these cysts, he initially assumed they were clinically irrelevant. Later, however, he understood that cysts can be the source of radicular symptoms, on which he reported starting in 1948 [2].

Since then, controversy has persisted regarding whether TCs are a rare source of pain or often produce symptoms. In 1956, Strully [3] stated that among the causes of low back pain, sacral cysts were “the least suspected, most frequently overlooked, and rarely treated.”

Not much has changed over the past 80 years, as limited knowledge and unwavering misconceptions persist regarding the role of TCs in patients’ pain. The clinical entity “symptomatic TCs (STCs)” remains an underestimated condition that is frequently overlooked [3–13].

Objectives

The first aim of this review was to determine the pathogenesis and pathophysiology of STCs and to list the various symptoms caused by sacral TCs. For this purpose, a literature review was conducted.

The second aim was to challenge the persistent idea that TCs are irrelevant findings, as there are no evidence-based arguments supporting this assumption.

Methods

The search for publications on STCs was performed using Web of Science, PubMed and Google Scholar. The selected studies were retrieved beginning from 1938 (the year Isadore Tarlov described these cysts) and included internationally peer-reviewed publications (published in English and Dutch), case reports, reviews and book chapters on STCs. The MESH terms used were Tarlov cysts, perineural cysts and sacral cysts. The references listed in review papers were searched to find additional relevant studies and reports.

Most of the available publications on TCs are case reports, case series or expert opinions. At present, there are no data available from well-designed research studies. Further, the patients and disease characteristics are usually reported incompletely.

All symptoms described in the case reports of sacral TCs were listed. Next, the pathophysiology of STCs and the reasons why STCs are overlooked were described on the basis of the available literature.

Results

In total, 224 publications were identified. The majority of the publications were concerning sacral meningeal cysts; however, 14 articles described cervical TCs and 21 articles described thoracic TCs. Forty-nine articles described

surgical techniques, and seven articles described fibrin glue injections as a treatment for STCs. Four cases concerned children; however, these were not included. The characteristics and the symptoms of the patients are listed in Table 1.

Discussion

Pathogenesis of TCs

Spinal meningeal cysts are diverticula of the spinal meningeal sac, nerve root sheath or arachnoid. TCs are the most prevalent type of meningeal cysts and are an extradural type of meningeal cysts that contain nerve root fibers [14].

TCs arise from pathologically increased hydrostatic and pulsatile pressure in the spinal canal, forcing cerebrospinal fluid (CSF) into the nerve root sheaths leaving the dural sac [9, 15]. Several authors have observed that radicular pain is a commonly underrecognized symptom of idiopathic intracranial hypertension. Of interest is that dilations of the nerve root sheaths could be seen preoperatively or on fMRI/CT imaging in these cases, and radicular pain resolved immediately following CSF evacuation [16–19].

Thus, under hydrostatic and pulsatile CSF pressures, some nerve root sheaths may dilate significantly. This mechanism is similar to the formation of vascular aneurysms under increased blood pressure. High blood pressure also gradually damages the inner walls of the vessels, whereas increased cerebrospinal pressure damages the axons [20].

It can be argued that most TC patients do not have papilledema, a hallmark of intracranial hypertension [21]. However, papilledema is no longer required to diagnose intracranial hypertension. Additionally, the arbitrary cutoff of 20 cm H₂O to define intracranial hypertension is probably too high. Rather, there may be a continuum between normal and increased ICP [22].

It has been suggested that spinal meningeal cysts can evolve due to the lack of compliance within the subarachnoid space and the inability to compensate for augmentations in CSF volume within a system of pathological CSF dynamics [23].

Several cases of an association between increased intracranial pressure and meningeal cysts have been described. Not all of these patients met the idiopathic intracranial hypertension criteria or showed papilledema [9, 23, 24].

It has been clearly established that lowering hydrostatic pressure (HP) by either external lumbar CSF drainage or the administration of acetazolamide relieves the symptoms of sacral TCs, and in some cases, it may reduce the size of the cysts [23, 25–27].

Changing from supine to standing or sitting upright causes a caudal shift of CSF with an increase in CSF volume in the spinal dural sac. A caudo-rostral pressure gradient

Table 1 Characteristics and the symptoms of the patients described in the case reports

	N	%
<i>Case reports</i>		
Articles describing case reports	94	
Number of cases	565	
<i>Demographics</i>		
Females	423	75
Males	142	25
Age (years)	48.2 ± 13.9 (range 22–74)	
How long (years)	4.2 ± 5.9 (range 0–20)	
<i>Symptoms</i>		
Low back pain	174	31
Buttock pain	49	9
Sacral pain	65	12
Coccygeal pain	54	10
Perineal pain	37	7
Pelvic pain	9	2
Groin pain	13	2
Lateral hip pain	8	1
Leg pain	196	35
Pain in the feet	10	2
Paresthesia of the perineum	12	2
Paresthesia of the leg	25	5
Paresthesia of the foot	14	3
Muscle cramps in the leg	12	2
Involuntary moving toes	1	
Walking difficulties	5	1
Cauda equine syndrome	3	1
Headaches	11	2
<i>Bladder problems</i>		
Urinary tract infections	1	
Bladder dysfunction	47	9
Urinary incontinence	76	13
<i>Bowel problems</i>		
Abdominal pain	7	1
Bowel dysfunction	40	7
Fecal incontinence	23	4
Fecal urgency	1	
Anal pain	6	1
<i>Sexual problems</i>		
Dyspareunia/genital pain	68	12
Persistent genital arousal syndrome	13	2
Impotence	1	
Penile pain	1	
<i>Triggers</i>		
Previous physical trauma	54	10
Heavy lifting straining history	12	2
Standing increases pain	38	7
Sitting increases the pain	20	4
Walking increases the pain/claudeication	47	8
Valsalva increases pain	32	6
<i>Neurological examination</i>		
Sensory abnormalities of the leg/feet/perineum	62	11

Table 1 (continued)

	<i>N</i>	%
Perianal sensory loss	8	1
Weakness of the leg/feet	62	11
Abnormal ankle jerk	8	2

exists within the spinal canal due to the hydrostatic pressure column of length [28]. The consequence of this pressure gradient in the human upright position was demonstrated when nerve roots from 100 autopsies were examined. TCs were detected in nine patients. Cysts were multiple and mostly symmetrical. The largest cysts were found on sacral nerve roots, and smaller cysts were found on dorsal and cervical roots. Several TCs could only be observed in microscopic preparations [29].

The cause of increased HP is not known. There are indications that a genetic factor is involved. Most of patients presenting with STCs are women [9, 10, 30], and familial cases have been reported [31]. TCs are more frequently found in patients with genetic soft tissue disorders such as hypermobility-type Ehlers–Danlos syndrome (EDS) and Marfan syndrome [32–34]. Due to weakness of the soft tissues, the nerve root sheaths in these patients are more susceptible to dilation, and patients with EDS often present increased intracranial pressure [32].

The location of TCs near the dorsal root ganglion [2, 20] may be due to increased permeability of the blood–nerve interface of the dorsal root ganglion compared with nerve roots [35]. As a consequence, when HP increases inside the nerve root sheath, CSF may start to leak between the endoneurium and perineurium [36]. The endoneurium is an endothelial lining that envelops the axons, while the perineurium is connective tissue that envelops the nerve fascicles [37] (Fig. 1).

TCs initially always have a connection with the subarachnoid space of the spinal canal (the cyst neck). The nerve fibers of the dorsal root are centered in the neck and spread over the cyst body or traverse the cavity [38]. Hence, due to this connection, the pressure inside TCs should be equal to the pressure inside the spinal canal [39]. Yet, studies have described cases in which these communicating TCs have caused significant symptoms [9, 20, 40]. This phenomenon can only be explained by increased HP in the nerve root sheaths compressing the axons of the dorsal root in the cyst body.

Larger cysts may act as a buffer system for CSF pressure. TCs are additional expanding CSF containers that may initially prevent a further increase in HP. As a consequence, patients with larger TCs often only become symptomatic later in life (during their fourth or fifth decade) [41].

In some cases, during the development of a TC, narrowing of the TC neck may occur due to proliferation of spinal

arachnoid granulations surrounding the dorsal nerve roots. Such proliferation is a reaction to an increase in CSF pressure [23]. A narrow neck may create a one-way valve system that allows CSF to enter the cyst but significantly limits the outflow. Under these circumstances, pressure inside the cyst rises to a level higher than pressure inside the spinal canal, and axons become even more compressed than in communicating TCs [9, 40].

Therefore, valved cysts are usually larger than non-valved (communicating) cysts.

Figure 2 shows an image of T2-weighted MRI myelography of large TCs on nerve roots S2 bilaterally, similar to those depicted in Fig. 1.

Figure 3 shows MRI myelography and CT myelography with intrathecal contrast injection of a patient with valved and non-valved TCs.

Table 2 shows an overview of the characteristics of valved and non-valved TCs [21].

Onset of symptoms

The dorsal root ganglion contains sensory axons and neurons. Therefore, irritation of these sensory structures initially causes pain and paresthesia [20].

TCs developing a valve system often produce severe pain over a shorter period (weeks or months). Additionally, these enlarging cysts may compress the motor axons in the ventral roots, causing motor dysfunction such as foot drop (L5), weakness of plantar flexion (S1) and sphincter dysfunction (S3S4) [21, 42]. Large TCs may also gradually compress other nearby nerve roots and/or erode the bony neuronal foramen.

Large valved STCs causing motor dysfunction are clinically more obvious. These are typically the examples described in case reports, whereas smaller TCs causing pain and paresthesia and/or bowel and bladder symptoms and anal and urethral sphincter dysfunction remain unnoticed.

Nerve damage

Objective evidence of nerve fiber damage in communicating cysts comes from an autopsy study. Microscopy studies revealed that the walls of TCs consisted of compressed degenerated axons and damaged myelin sheaths [43].

Additional evidence of nerve damage from TCs comes from electrodiagnostic studies. Previous studies revealed

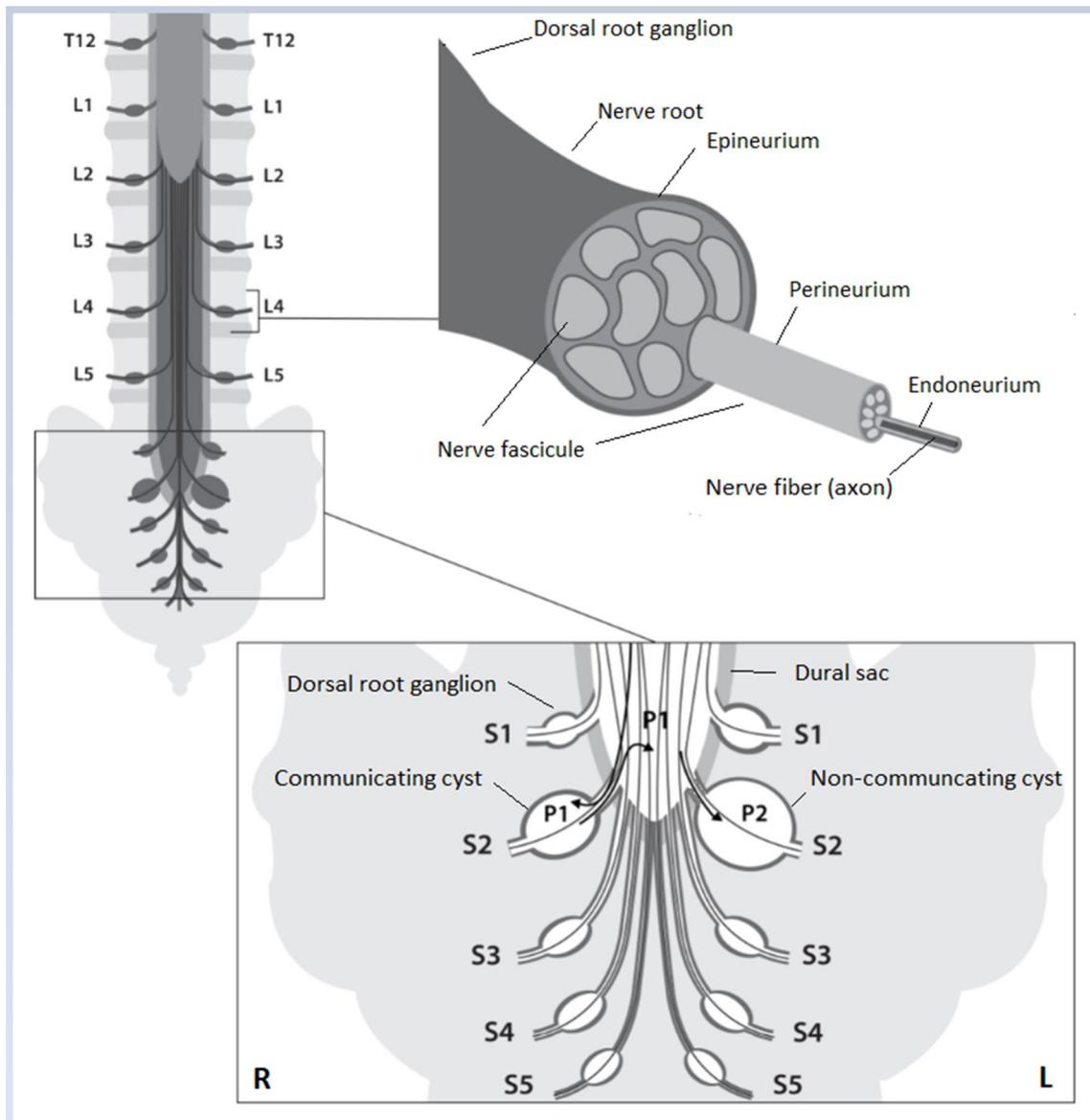


Fig. 1 Schema of the anatomy of a nerve root, a communicating (non-valved) Tarlov cyst on the right nerve root S2 and a non-communicating (valved) Tarlov cyst on the left nerve root S2. P1 is the pressure in the subarachnoid space of the dural sac, which is the same

as the pressure inside the communicating Tarlov cyst. P2 is the pressure inside a non-communicating Tarlov cyst. P2 is always higher than P1

delayed Hoffmann reflexes (H-reflexes), a sign of S1 sensory radiculopathies. An H-reflex is the electrophysiological equivalent of the Achilles tendon reflex [44, 45]. In another study in 30 STC patients, almost all patients with small and/or large TCs showed simultaneous bilateral electromyographic abnormalities in multiple lumbar and sacral myotomes. Additionally, the S3- and S4-innervated ano-anal reflex was delayed [21, 46]. The role of this reflex is to prevent fecal incontinence. Mild-to-moderate fecal incontinence is a common symptom in patients with TCs and is probably due to damage to the sensory branch of the reflex arc

[42]. Clinically, anal muscle tone may be normal (the motor branch), but the sensitivity of the surrounding anal skin may be reduced (the sensory branch) because TCs mostly affect the dorsal root ganglion, where sensory nerves are located.

In patients with idiopathic intracranial hypertension, nerve conduction studies revealed absent or prolonged F-waves, which normalized after ventriculo-peritoneal CSF shunting or repeated lumbar puncture [18]. F-waves are late electrophysiological responses to antidromic activation of motor neurons and are useful for evaluating conduction along proximal root segments (where TCs are located).



Fig. 2 Image of T2-weighted MRI myelography of large Tarlov cysts on nerve roots S2 bilaterally, similar to those depicted in Fig. 1. It is not clear from the MRI if these cysts are valved or non-valved Tarlov cysts. CT myelography using intrathecal contrast injection would be required to show the absence of early filling of valved Tarlov cysts

Symptoms associated with Tarlov cysts

Because HP is highest in the lowest segment of the dural sac, symptoms usually present as a chronic cauda equina syndrome. The intensity and location of the symptoms fluctuate [47]. As the CSF volume is shifted caudally when changing from the lying to the sitting position, pain increases when sitting or standing and is relieved when lying down [9]. In patients with valved TCs, the pain will not be relieved when lying down because the outflow of CFS is limited.

Additionally, because of the overall increased CSF pressure, patients may report distant radicular symptoms such as thoracic pain, pain in the neck and arms and headaches [21].

Table 3 shows an overview of symptoms caused by TCs according to the involved nerve roots.

Female predominance in STCs

MRI studies of patients with back pain have revealed that 70% of patients with TCs are women [30, 40]. We found that 75% of the patients described in the case reports with STCs were women (Table 1).

It has been postulated that gender-related differences in the composition of the dura mater or spinal nerve roots may be the underlying cause of this female predominance [9].

A prospective *in vivo* study measuring the perfusion and permeability of the nerve roots and dorsal ganglia in patients with neuropathic pain showed that women exhibit

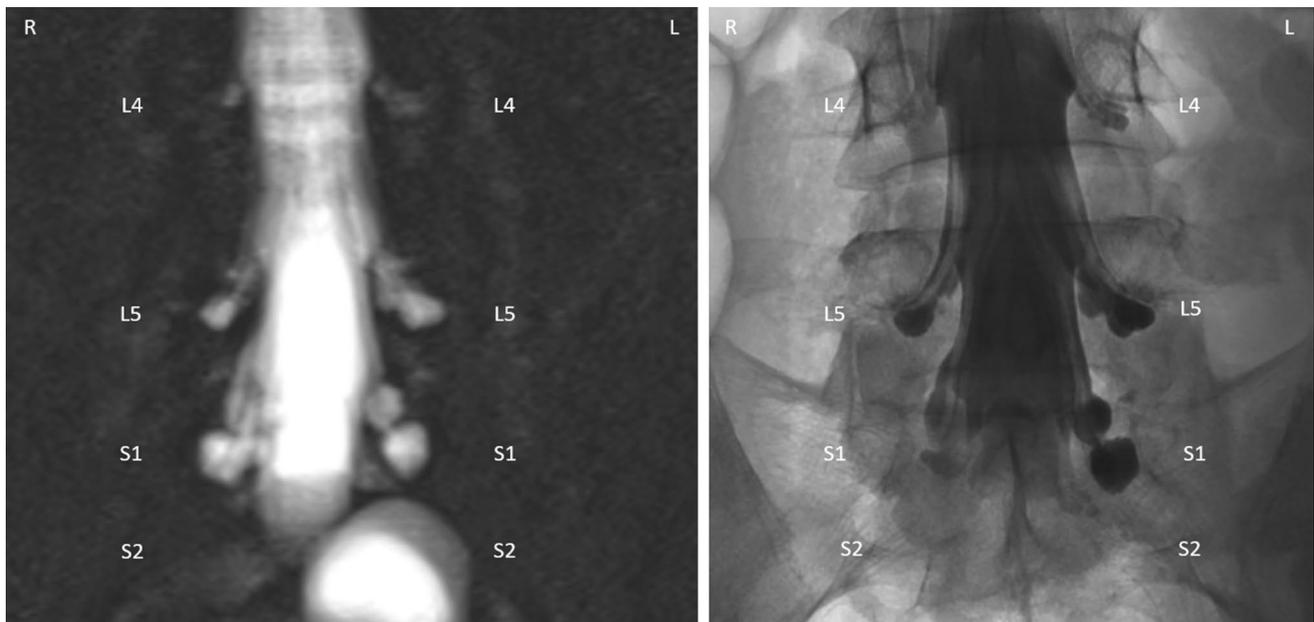


Fig. 3 Image of nerve root dilations and small Tarlov cysts on nerve roots L4 to S1 and a large valved Tarlov cyst on the left nerve root S2. The left image shows the MRI myelography, and the right image shows an early view of the CT myelography with intrathecal contrast

injection. The large cyst does not immediately fill up with contrast medium and can be diagnosed a non-communicating (valved) Tarlov cyst, whereas the smaller cysts can be diagnosed as communicating (non-valved) Tarlov cysts

Table 2 Characteristics of valved versus non-valved TCs

Tarlov cysts	Valved (non-communicating) cysts	Non-valved (communicating) cysts
Size	Commonly larger (> 10 mm)	Commonly smaller than valved cysts
Onset of symptoms	Long pain-free interval	Onset early in life
Mean age at onset of symptoms	± 5th decade	± 2nd–3rd decade
Evolution	Rapid progression	Slow progression
Sensory symptoms (pain and paresthesia/numbness)	Commonly	Commonly
Motor symptoms (muscle weakness)	Commonly	Occasionally
Cauda equine syndrome	Acute	Chronic
Prevalence	Rare	Common
Reported in the literature	Commonly	Rarely
Clinical appearance	Easily diagnosed	Commonly overlooked
Surgical intervention	May be recommended in cases of severe pain or motor dysfunction	Probably inefficient

Table 3 Reported symptoms according to the involved nerve roots

Nerve supply	Symptoms
S2, S3 and S4 <i>Sensory:</i> perineum, clitoris, penis, vagina, scrotum <i>Autonomic:</i> detrusor muscle of the bladder, descending colon, transverse colon, rectum, internal urinary sphincter, internal anal sphincter <i>Motor:</i> external urinary sphincter, external anal sphincter	Pelvic pain Perineal pain and paresthesia Vaginal pain; dyspareunia Testicular or penile pain; prostate pain Pelvic instability symptoms (in pregnancy) Neurogenic bladder symptoms: urine retention, hesitation, Valsalva voiding, urinary frequency, urgency, painful bladder, urge incontinence, stress incontinence Neurogenic bowel symptoms: constipation, alternating with diarrhea; intestinal cramps; defecation frequency; urgency; bloating; false defecation urge, fecal incontinence; stabbing pain; pressure or cramps in the anal sphincter Erectile dysfunction; retrograde ejaculation Female anorgasmia; persistent genital arousal disorder
S2 <i>Sensory:</i> perineum, posteromedial side of the legs, plantar region of the feet <i>Motor:</i> intrinsic foot muscles	Pain, cramps and paresthesia in the feet Atrophy of the intrinsic foot muscles due to compression of nerve root S2
L5 and S1 <i>Sensory:</i> posterior side of the legs (S1), lateral side of the leg, first and fifth toe (L5), dorsal side of the feet <i>Motor:</i> gluteus maximus muscle and calf muscles (S1), gluteus medius muscle and extensor muscles of the feet and toes (L5)	Lumbar and sacral pain Pain in the piriformis muscle and trochanter region Pain and/or paresthesia in the legs and feet Leg cramps Leg weakness Neurogenic claudication: pain while walking, a significantly slower walking pace than before onset, increased pain when walking uphill Weakness of dorsiflexion of the feet; rarely, foot drop (L5) Weakness of plantar flexion of the feet (S1)
L1 to L4	Paresthesia and pain in the legs Weakness of knee or hip extension
Dorsal nerves	Paresthesia and pain in the upper back Intercostal neuralgia
Cervical nerve roots	Cervical pain and pain in the trapezius muscles Pain and paresthesia in the arms and hands
Brain	Headaches

significantly increased permeability and interstitial leaking within the dorsal root ganglia compared with men. As a consequence, in women, the sensory fibers and neurons inside the dorsal ganglion may be more vulnerable to mechanical pressure or toxic agents. These noxious stimuli can gradually produce chronic neuropathic pain. It was speculated that this finding may explain why women are more susceptible to chronic neuropathic pain [36].

Higher permeability compared to men may also explain why the dorsal root ganglia of women are more susceptible to the formation of TCs, which is due to leaking of CSF between the endoneurium and perineurium.

Diagnosis of STCs

Objective findings can confirm that TCs are symptomatic and the cause of pain:

1. TCs are readily visible on MRI. Such TCs should be considered in the differential diagnosis of low back pain or radicular pain, as these structures are not just cysts in the neighborhood of nerve roots, but enlarged nerve root sheaths containing compressed nerves potentially causing pain.
2. During a clinical neurological examination, when comparing the left and right sides, sensory abnormalities can be detected. Some cases may present absent ankle reflexes or loss of strength [42].
3. Urodynamic testing may show specific abnormalities in STC patients, such as early sensation of filling in 70%, involuntary detrusor contractions in 33%, urethral instability in 33% and stress incontinence in 33% [48].
4. Electrophysiological testing of the involved nerves (particularly the L5 and sacral nerve roots) may show delayed F-waves, delayed S1 H-reflexes, delayed ano-anal reflexes and neurogenic abnormalities during needle EMG [21].
5. A lumbar puncture with evacuation of CSF to relieve symptoms may be used as a diagnostic test [21, 27, 49].

Reasons STCs may be overlooked

1. In general, TCs are assumed to be asymptomatic

In the introduction of case reports of STCs or papers on surgical techniques for the treatment of STCs, it is frequently stated that TCs very rarely cause pain or neurological symptoms. However, there is no clear scientific evidence underlying this assertion. On the other hand, a systematic review of 31 case series on the surgical treatment of TCs indicated that there is actually a correlation

between pain and TCs as the majority of patients treated experienced partial or complete relief of pain [50].

It is true that TCs can reside unnoticed in many patients as they progressively become larger, as they may represent a buffer for excess HP within the subarachnoid space. It is likely that when this buffer system decompensates or when a valve system is established, patients become (more) symptomatic [21, 41].

TCs are more prevalent than generally assumed. Studies analyzing the lumbosacral MRIs of patients with lower back pain have reported a 1.5–13% prevalence of TCs [8, 30, 51–53]. A retrospective observational epidemiological study assessed 1100 sacral magnetic resonance (MR) studies for various indications, including 100 studies of children and adolescents. Sacral TCs were found in 13% of adult patients. No sacral TCs were found in children, and the prevalence of TCs increased with age [30].

Microscopic examination of cervical nerve roots from the autopsies of 120 people, predominantly elderly adults, revealed TCs containing degenerated nerves in 30% of cases [29, 43]. Thus, with aging, an increased prevalence of TCs may be associated with progressive neurodegeneration.

It has been estimated that approximately 25% of the TCs seen on MRI are symptomatic at the time of discovery and that a subset of the non-symptomatic cysts may become symptomatic later in life. Consequently, STCs can be found in about 1% of the population [10, 15, 40].

Additionally, TCs were found to be more prevalent in specific patient groups. In a review of lumbosacral MRIs, TCs were found in 13 out of 17 patients (75%) with perineal pain [5] and in 12 out of 18 women (66.7%) with persistent genital arousal disorder [11].

Thus, because STCs are often unrecognized, they may be an important cause of unexplained chronic pain.

2. It is assumed that it is clinically difficult to determine whether TCs seen on MRI are the cause of a patient's pain

One reason for the difficulty of associating TCs with pain is that TCs are located near the dorsal root ganglion and primarily affect sensory axons. Consequently, as a clinical entity, STCs predominantly represent a sensory nerve disorder. Initial symptoms are pain and numbness, which are difficult to objectify [20, 54].

A comprehensive history including very specific questions about bowel, bladder and sphincter symptoms may guide the diagnosis toward STCs [55].

Clinical neurological examinations may reveal sensory abnormalities in involved dermatomes L5 to S4 and sometimes weak or absent Achilles tendon reflexes (S1) and weak dorsiflexion (L5) or plantar flexion (S1) of the feet [42].

3. MRI or electromyography studies focus on the lumbar and S1 nerve roots

In most cases of sciatica, an MRI of the lumbosacral spine does not routinely include axial or coronal sequences of the sacrum, and electromyography studies do not include sacral nerve root myotomes. This is because in the bony sacrum, sacral nerve roots are assumed to be safe from external compression by disk protrusion or degenerative alterations, as opposed to lumbar and S1 nerve roots. However, during the formation of TCs, sacral nerve roots are exposed to internal compression from increased CSF pressure.

4. TCs seen on MRI are usually not reported by radiologists

Because TCs are assumed to be clinically irrelevant, they are usually not reported [7]. Moreover, if TCs are reported, it is often stated that the patient's pain cannot be attributed to TCs. It may be better to state that the clinical relevance of the detected findings should be evaluated clinically and/or electrophysiologically.

5. Degenerative alterations of the lumbosacral spine observed on MRI are almost always identified as the cause of a patient's low back pain and/or sciatica

This conclusion may be overly simplistic. In an MRI study of 96 asymptomatic volunteers (mean age 43.3 years), disk degeneration was observed at least one level in 64% of subjects [56]. In another MRI study of 60 asymptomatic volunteers (20–50 years), disk bulging or disk protrusion was observed on at least one level in 62–67%, and disk extrusion was observed in 18% of subjects [57]. Therefore, in patients with TCs, it should not always be concluded that degenerative spine alterations are responsible for low back pain and that TCs are harmless per se.

6. Small TCs can be symptomatic

The diagnosis of STCs is usually established if a single large cyst on a sacral nerve root causes mono-radicular pain and/or neurological symptoms in the corresponding myotome. If the pain location in patients with multiple TCs does not correspond with the largest TC, it is usually concluded that TCs cannot be responsible. One of the main characteristics of Tarlov cysts is a lack of correlation between radiologic findings and symptomatology. It is presumed that increased CSF pressure forces CSF into the perineural space [47].

This lack of correlation between radiologic findings and symptoms was shown in a study of 30 patients with STCs. In only 59% of the cases, the patients reported worse pain on

the side where the largest TC was located [21]. The reason for this discrepancy is likely that radicular pain is not caused by external pressure on the nerve root exerted by a large cyst, but by internal pressure due to increased HP within the cyst. Indeed, during their development, most TCs do not have a valve system. The pressure within these communicating cysts should be equal to the pressure in the subarachnoid space of the spinal canal [39]. Hence, increased HP affects several nerve roots at the same time and therefore may cause pain and paresthesia in multiple dermatomes bilaterally and simultaneously, regardless of the size of the TCs [21].

Microscopy studies of resected sacral TCs demonstrated that even very small cysts can cause debilitating pain [20].

Figure 4 shows a T2-weighted MRI myelography coronal image of bilateral small sacral cysts in a symptomatic patient.

7. Increased CSF pressure as an underlying mechanism versus TCs seen on MRI

When considering STCs, the focus is mostly on TCs seen on MRI. It is assumed that when the cyst is removed, nerve compression will be relieved, and the patient's problems

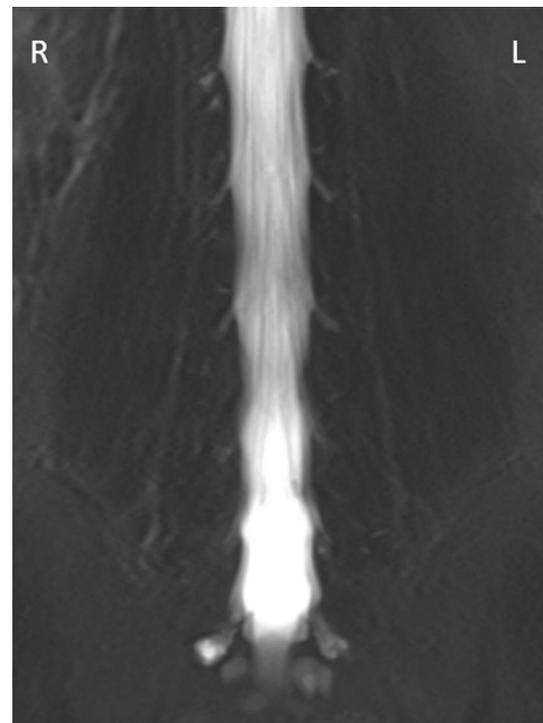


Fig. 4 Image of T2-weighted coronal MRI image of multiple small Tarlov cysts, bilaterally localized (grapes sign) in a 64-year-old male patient who suffered for almost 50 years from unexplained, progressively debilitating sacral and leg pain and bladder and bowel symptoms. This patient showed EMG abnormalities in multiple sacral myotomes

should be solved. However, this is only true in cases of (large) cysts with a valve system, which are rather uncommon. Hence, the focus should be on the primary cause, i.e., the increased HP. If this pressure is not relieved, symptoms will relapse.

This was demonstrated during the follow-up of STC patients who had undergone surgery. Thirty-five TC patients with sacral–perineal pain and a mean cyst size of 3.6 cm were treated. Follow-up data showed improvement in 93% of the patients at some point. However, 50% of patients developed recurrent pain symptoms, despite a reduction in cyst size in 92% of patients [39].

8. You do not know what you do not ask about

Perineal and genital pain, bladder, bowel and sphincter dysfunction or sexual symptoms are assumed to be unrelated to low back pain. When taking the history of patients with low back pain or leg pain, clinicians rarely ask about these symptoms.

Compared to low back pain or leg pain, these symptoms are less frequently described in case series (Table 1), probably partly because these symptoms were not assessed.

Additionally, due to embarrassment, women and men do not talk about these symptoms when they are consulting a clinician [10].

However, asking about these symptoms is a key point in the diagnosis of STCs [55].

9. Unexplained pain is attributed to depression

Patients with undiagnosed STCs usually have a long history of pain and may have undergone several technical investigations and interventions. Moreover, patients are generally unable to perform sitting- or straining-related activities and therefore may be unable to participate in professional and social life. Patients often become housebound and socially isolated. The patient's suffering is often underappreciated by family, friends, peers and physicians. As a consequence, depression is a common comorbidity, and the patient's unexplained pain may be mistakenly attributed to comorbid depression [6, 9, 55].

10. Strong polarization between “believers” and “nonbelievers”

The recognition of STCs is subject to bias. First, there is gender bias, as there is a stronger tendency to attribute atypical physical symptoms to psychological factors in women than in men [58].

Belief that TCs are asymptomatic, irrelevant findings on MRI is also subject to confirmation bias and cognitive dissonance. Confirmation bias is the tendency to interpret

new information in a way that is consistent with preexisting beliefs and hypotheses [59]. Cognitive dissonance is how one responds to conflicting information, that is, the uneasy feeling we get when new evidence calls into question beliefs that we think are correct [60]. As a consequence, those who believe that TCs can be the cause of a patient's symptoms debate with nonbelievers, who assume TCs are clinically irrelevant. Unfortunately, at present, the nonbelievers still outnumber the believers.

Not being diagnosed is harmful for patients because their condition may be extremely debilitating; therefore, patients will continue consulting doctors in search of treatment. Consequently, further unnecessary or ineffective and potentially harmful technical interventions may be performed.

When researchers submit a manuscript on STCs, it may be exposed to unconscious bias. Such manuscripts may be more frequently refused by journals based on assumptions that this condition is very rare and that readers may not be interested. Additionally, the reviewers may be skeptical and more critical of new evidence, indicating that TCs are not harmless, as assumed [61].

Furthermore, readers may not be interested because of the assumption that STCs are so rare that they will encounter this pathology only a few times in their career.

Conclusion

STCs are an underdiagnosed clinical entity associated with poor quality of life. In this paper, the ten most important misconceptions and reasons STCs are overlooked are presented. From our current understanding of the pathophysiology and symptomatology of STCs, it is clear that STCs should be part of the differential diagnosis of low back pain and sciatica.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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Affiliations

Mieke Hulens^{1,8} · Ricky Rasschaert² · Frans Bruyninckx³ · Wim Dankaerts¹ · Ingeborg Stalmans^{4,5} · Peter De Mulder⁶ · Greet Vansant⁷

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 Ophthalmology Research Group, Department of Neurosciences, University of Leuven, Herestraat 49, 3000 Leuven, Belgium

⁵ Department of Ophthalmology, University Hospitals UZ Leuven, Herestraat 49, 3000 Leuven, Belgium

⁶ Department of Anesthesiology and Pain Therapy, Imelda Hospital, 2820 Bonheiden, Belgium

⁷ Department of Social and Primary Health Care, Public Health Nutrition, University of Leuven, Herestraat 49, 3000 Leuven, Belgium

⁸ Sint-Joris-Weert, Belgium