

ADDENDUM TO THE ADHESIVE ARACHNOIDITIS SYNDROME

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The syndrome of low-pressure CSF headaches was first recognized following diagnostic lumbar puncture.

In the 1940s and 50s, Woltman in the United States ([\[1\]](#)) and Schaltenbrand in Germany ([\[2\]](#)) described "spontaneous" posture-related low-pressure headaches that did not develop after lumbar puncture.

In spontaneous CSF leaks, there is sometimes a history of strenuous activity or trivial traumatic injury.

Mokri et al. ([\[3\]](#)) have recently described cases associated with postural tachycardia syndrome (POTS).

Schievink et al. ([\[4\]](#)) have also linked intracranial hypotension with connective tissue disorders. Ferrante et al. ([\[5\]](#)) reported a case of Marfan's syndrome and postural headache in whom MRI myelography revealed bilateral multiple large meningeal diverticula at sacral nerve roots level; the authors postulated that

"The intracranial hypotension syndrome likely resulted from a CSF leak from one of the meningeal diverticula. In conclusion patients with spinal meningeal diverticula (frequently seen in Marfan's syndrome) might be at increased risk of developing CSF leaks, possibly secondary to Valsalva maneuver or minor unrecognized trauma."

Bachmann-Mennenga et al. ([\[6\]](#)) suggest that when CSF leak is involved, the leaks are mainly located cervically or at the cervicothoracic junction.
Implantation of the pump may also cause CSF leak.

I have come across several individual diagnosed with arachnoiditis who have known recurrent CSF leaks.

Schievink and Jacques ([\[7\]](#)) reported on a recurrent CSF leak in a patient found at surgery to have an absence of the entire nerve root sleeve at multiple thoracic levels.

They dubbed this the "nude nerve root" syndrome and suggested that these patients were at higher than normal risk of recurrent CSF leak.

Schievink ([\[8\]](#)) remarked,

"Spontaneous intracranial hypotension is an important cause of "new daily persistent headaches" but is not a well-recognized entity. The misdiagnosis of spontaneous intracranial hypotension can have serious consequences."

He suggested,

"Migraine, meningitis, and psychogenic disorder were the most commonly entertained diagnoses".

These headaches tend to be *postural*, i.e. they come on when standing/sitting and are relieved by lying down. Essentially it is the same headache as experienced post-lumbar puncture or if there is an accidental dural puncture during epidural anaesthesia/epidural steroid injection/spinal surgery.

However, Mokri ([\[9\]](#)) noted

“In a substantial minority of patients, headaches are not orthostatic and may mimic other types of headache. Additional diverse neurological manifestations may dominate the clinical picture and patients may occasionally have no headache at all.”

The headaches may be localized or diffuse, often frontal or occipital (at the back of the head) and can be of sudden onset when changing posture, shaking the head, coughing, sneezing or straining on the toilet.

They tend to be severe and may be dull or throbbing. Mokri ([\[10\]](#)) suggests that

“with chronicity the orthostatic features may blur into a chronic, lingering headache.”

There may be associated nausea, vomiting, blurred vision, dizziness or tinnitus. More unusual features include diplopia (double vision), upper limb pain/tingling/weakness and interscapular pain (between the shoulder blades).

Typically, the headache is relieved by lying down. Occasionally, patients may complain of back or neck pain that may or may not correspond to the level of the CSF leak.

Loss of CSF volume causes downward displacement of the brain when sitting/standing (cf. Chiari Malformation) which stretches arteries and veins and therefore affects the nerve fibres supplying them.

Low CSF volume may be associated with apparently normal CSF pressure due to compensatory intracranial vascular dilation.

However, these 'normal' values, which may be >60mmHg, may in fact be lower than the patient's normal values.

In arachnoiditis, there may be fluctuating CSF pressure due to the impairment of normal CSF flow by the scar tissue.

There could be areas of reduced pressure but also, as I have already mentioned, other areas of locally increased pressure, which may be associated with development of arachnoid cysts or syringomyelia.

MRI scan of the brain with gadolinium shows diffuse pachymeningeal enhancement and possibly sinking of the brain, subdural fluid collections, enlargement of the pituitary, engorgement of venous sinuses, and engorgement of epidural venous plexus. Lumbar puncture (not advised in arachnoiditis patients) may demonstrate low opening CSF pressure.

As regards spontaneous low CSF volume/pressure headaches, Mokri concludes that

“The disorder has a broad clinical and imaging spectrum with substantial variability in clinical and imaging features, in CSF findings, and in response to treatment.”

Treatment is difficult once the problem is chronic, although caffeine may be beneficial. Epidural blood patch, used in the acute situation, is obviously not a viable option in arachnoiditis patients, particularly because blood is, as we have seen, highly irritant to nerve roots and thus likely to exacerbate arachnoiditis.

If the source of the leak can be identified, surgical closure may be an option.

Probably in arachnoiditis with chronic intracranial hypotensive headaches, rest and analgesia are the most viable options and may resolve the symptoms at least partially within a few days.

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[1] Woltman HW: *Med Clin North Am* 1940; 24:1159-1170, Headache: a consideration of some of the more common types.

[2] Schaltenbrand G: *Lancet* 1953;1:805-808, Normal and pathological physiology of the cerebrospinal fluid circulation.

[3] Mokri B, Low PA. *Neurology*. 2003 Oct 14;61(7):980-2. Orthostatic headaches without CSF leak in postural tachycardia syndrome.

[4] Schievink WI, Gordon OK, Tourje J. *Neurosurgery*. 2004 Jan;54(1):65-70

Connective tissue disorders with spontaneous spinal cerebrospinal fluid leaks and intracranial hypotension: a prospective study.

[5] Ferrante E, Citterio A, Savino A, Santalucia P. *Cephalalgia*. 2003 Sep;23(7):552-5. Postural headache in a patient with Marfan's syndrome.

[6] Bachmann-Mennenga B, Philipps J, Haukamp F, Reinbold WD. *Anaesthesist*. 2004 Jan;53(1):45-52. [Spontaneous intracranial hypotension. A rare syndrome with good treatment options]

[7] Schievink WI, Jacques L. *Neurosurgery*. 2003 Nov;53(5):1216-8; discussion 1218-9. Recurrent spontaneous spinal cerebrospinal fluid leak associated with "nude nerve root" syndrome: case report.

[8] Schievink WI. *Arch Neurol*. 2003 Dec;60(12):1713-8. Misdiagnosis of spontaneous intracranial hypotension.

[9] Mokri B. *Curr Opin Neurol*. 2003 Jun;16(3):319-26. Headaches caused by decreased intracranial pressure: diagnosis and management.

[10] Mokri B. *Curr Neurol Neurosci Rep*. 2004 Mar;4(2):117-24 Spontaneous low cerebrospinal pressure/volume headaches.