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Chapter 114

Low Cerebrospinal Fluid Pressure

Steinar T. Vilming and Bahram Mokri

POSTLUMBAR PUNCTURE HEADACHE

Definition of Post-Lumbar Puncture Headache

International Headache Society (IHS) code and diagnosis: 7.2.1 Postdural (postlumbar) puncture headache
World Health Organization (WHO) code and diagnosis: G44.88 Headache associated with other specified disorders
Short description: Bilateral headache that develops within 5 days after lumbar puncture (LP) and disappears within 1 week. Headache occurs or worsens in the upright position and disappears or improves after resuming the recumbent position.
Other terms: postlumbar puncture headache, postdural puncture headache, postmyelogram headache

Essex Wynter in 1889 and H. Quincke in 1891 performed the first dural punctures, and a vast amount of literature followed on the distressing headache that may ensue. Postlumbar puncture headache (PPH) and other headaches due to low cerebrospinal fluid (CSF) pressure are unique because of the postural character, occurring or worsening when assuming the erect position.

Pathophysiology

The exact cause of PPH remains elusive. The favored "leakage theory," which was proposed at the beginning of this century, implies that post-LP leakage of CSF through a dural rent leads to a decreased CSF pressure and volume, whereby the cerebral veins dilate and the brain is deprived of its CSF cushion (72). The consequent gravity-dependent downward sagging of the brain causes traction on the distended and distorted pain-sensitive anchoring veins (the sagittal sinus and its tributary veins) and pressure on equally pain-sensitive large arteries at the skull base and the tentorium (Fig. 114-1). Several facts support this theory.

A *dural rent* is caused by a Quincke-cut needle and it has the form of a nearly opened tin lid hinged on one side (Fig. 114-2). Complete sealing of a rent may take more than 14 days (22). Small rents caused by thin needles heal more rapidly than larger rents, and rents made through the thickest parts of the dura retract more quickly than rents in the thinner parts (22,32). Penetrating the dura with the bevel of a Quincke needle parallel rather than transverse to the longitudinal axis of the dural cylinder reduces the incidence of PPH, possibly because such a technique creates the smaller apertures (29). It has been suggested, although unproven, that a rent created by an oblique needle insertion or a penetration through a richly vascularized area of the dura might be more likely to heal (32,57).

CSF is normally produced at a rate of about 0.3 mL per minute, but despite its great renewal capacity, a low CSF volume and pressure may be maintained by *CSF leakage,* which could be continuously observed by direct inspection isotope studies weeks and even months after a spinal tap. Epidurally lost CSF is absorbed through the intervertebral foramina (32).

CSF pressure closely parallels the venous pressure at all levels of the intracraniospinal system. In the erect human, intracranial CSF pressure is negative at the vertex, approximately 150 mm H_2O. In the horizontal position, intracranial CSF pressure is positive at all levels, normally 50 to 180 mm H_2O. Removal of 30 mL CSF regularly induces headache in the erect position concomitant with a reduction of the vertex CSF pressure down to approximately 220 to 290 mm H_2O. The headache is ameliorated by restoration of the CSF volume or tilting toward the horizontal position and augmented by jugular compression (44,93). In some PPH patients, CSF pressure is zero or low when repunctured (57,82), and intrathecal infusion of saline relieves the headache by restoring the pressure (60). In a controlled clinical study, the average reduction in CSF pressure after lumbar puncture (LP) was significantly greater for patients with headache compared with patients without headache (95). In another study, however,

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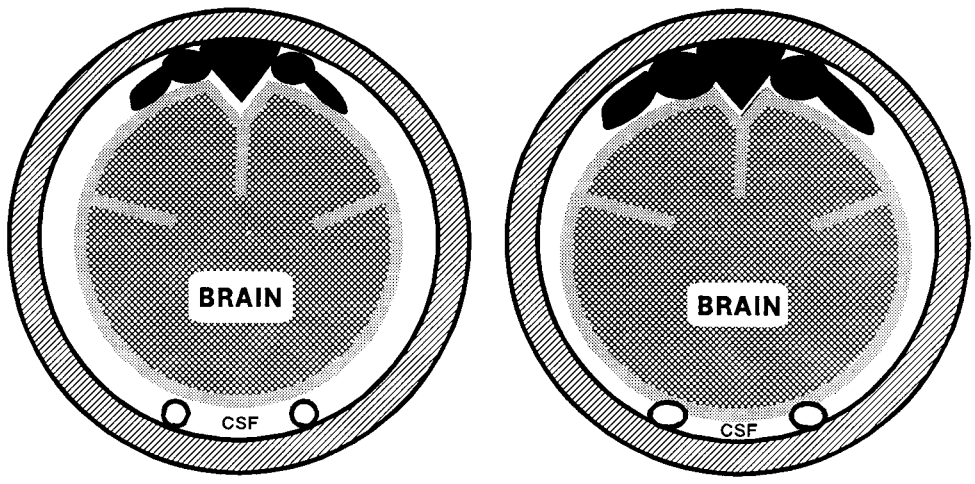

FIGURE 114-1. A schematic presentation of intracranial structures before **(left)** and after **(right)** withdrawal of cerebrospinal fluid explaining the mechanism of postlumbar puncture headache (*PPH*). The loss of volume is compensated for by a significant distention of the pain-sensitive sagittal sinus and its tributary veins. The downward sagging of the brain causes traction on these veins and pressure on the large arteries at the skull base.

only three of five PPH patients had a low CSF pressure with repuncture after 24 hours, and a low CSF pressure was found in 7 of 37 patients who had not developed PPH (47). Thus, the association between PPH and low CSF pressure is not invariable, and other factors may play some role.

CSF serves to buoy about 97% of the weight of the slightly denser brain (61). By magnetic resonance imaging (MRI), it has been shown that the CSF volume is significantly reduced in nearly all patients 24 hours after LP and that there is a trend toward a greater reduction in those who develop PPH than in controls (36). This loss of CSF causes the thin-walled veins to distend, which has been observed directly through a cranial window in cats (31). Also, the brain appeared edematous and cyanotic, and the arteries were constricted slightly. These findings agree with the Monro-Kellie doctrine with Burrow's modification, which postulates that blood volume increases when the CSF volume is reduced and vice versa (37). Elevated blood volume is due exclusively to a venous dilation, since the venoarterial reflex, which occurs within seconds of CSF removal, causes the arteries to constrict and keeps cerebral blood volume unaltered.

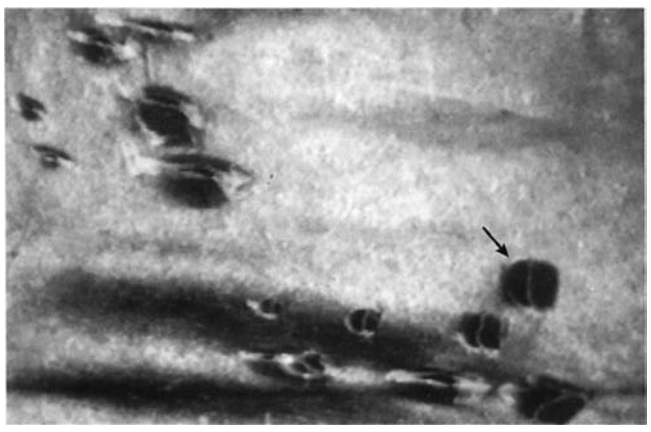

FIGURE 114-2. Effect of dural puncture with Quincke needles of different sizes from 20 to 29 gauge. The typical "tin-lid effect" is visualized, and one example is given (*arrow*). From Dittmann et al. (23).

Clinical Features

IHS diagnostic criteria for postdural (postlumbar) puncture headache (revised International Headache Classification of Headache Disorders [ICHD-II]) are as follows:

A. Headache that worsens within 15 minutes after sitting or standing and improves within 15 minutes after lying, with at least one of the following and fulfilling criteria C and D:
  1. Neck stiffness
  2. Tinnitus
  3. Hypacusia
  4. Photophobia
  5. Nausea
B. Dural puncture has been performed.
C. Headache develops within 5 days after dural puncture.
D. Headache resolves either:
  1. Spontaneously within 1 week
  2. Within 48 hours after effective treatment of the spinal fluid leak (usually by epidural blood patch)

PPH occurs within 2 days (range: minutes to days) after LP in approximately 90% of patients (83). It usually lasts a week, but a duration of several weeks or even months has been reported (44,60,79,82,83), probably because of a persisting fistula. PPH with early onset seems to last longer and tends to be more severe than PPH with a later onset (35,88). PPH is posture dependent, usually starting to increase within 20 seconds when the patient assumes an

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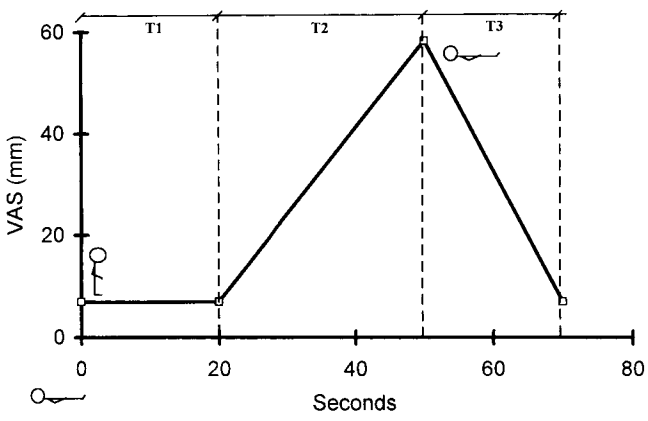

FIGURE 114-3. Headache severity on a 100-mm visual analogue scale (*VAS*) related to time; median values from 79 patients. The body position of the patient is indicated. The pain is minimal when the patient is in the recumbent position, but pain starts to increase after 20 seconds when the patient is in the upright position (*T1*). The time to maximal pain is 30 seconds (*T2*). When the patient lies down, the pain decreases to its lowest level within 20 seconds (*T3*). From Vilming and Kloster (83).

upright position, reaching its maximum within another 30 seconds, and subsiding within 20 seconds of assuming recumbency (Fig. 114-3) (83). The time course of PPH is fairly stable until the last 2 days of its course, when the headache severity usually starts to subside, the patient's mobility increases, and the time required for headache to develop on rising increases (84).

PPH is usually described as aching, dull, deep, constricting, or throbbing, and it is most frequently frontal but often occipital, fronto-occipital, or generalized (1,44,79,82). More than half of PPH patients have associated symptoms, and the more severe the PPH, the more frequently the symptoms occur. Dizziness and nausea, occasionally with vomiting, are most prevalent. Auditory difficulties and tinnitus are not infrequent and may be caused by leakage of perilymphatic fluid from the cochlea to the cerebrospinal space through a cochlear aqueduct, which is functionally open in about 50% of adults (48,90), resulting in a Ménière-like syndrome. Blurred vision and other visual disturbances are quite infrequent. Cranial nerve lesions, usually the abducens nerve, have been reported (1,79,82,88).

Age and *gender* influence the incidence of PPH. Data from old studies that investigated large numbers of patients (79,82) and a more recent report (88) indicate that the incidence of PPH is highest among younger patients, perhaps with the exception of prepubertal children (26). Also, the incidence of PPH may decrease with increasing age, usually above 50 or 60 years, perhaps on the basis of narrowed epidural space (81) or reduced elasticity of pain-sensitive cerebral vessels (82). It should be noted, however, that concerns with study methodology and the potential confounding effect of gender cast doubt on the conclusion that the incidence of PPH is reduced with increasing age (79).

The potential effect of *gender* on the incidence of PPH has been addressed in several studies, but most were not designed specifically for that purpose, and hence the potential for methodologic flaws. In general, the incidence of PPH is higher in women than in men (19,25,79,80, 82,88,95). It is suggested that this observation is on the basis of a higher incidence of PPH in women of childbearing age and does not apply to the prepubertal girls or postmenopausal women (Fig. 114-4). Also, the authors of a large study (n = 300 patients) argued that the apparent reduction in the incidence of PPH with age is gender dependent; that is, the incidence of PPH is reduced in aging women only (88).

The *indication* for LP may impact the development of PPH. The incidence of PPH following spinal anesthesia (SA) is reportedly lower than that following diagnostic LP (79). Perhaps the requirement that patients post-SA remain on bedrest for several days after the procedure hides cases of short-lasting PPH. Also, it is unclear how the introduction of intrathecal chemicals during SA influences the development of PPH.

The influence of *body mass* index on the incidence of PPH remains elusive (45,86).

Several well-designed controlled studies have indicated that the incidence of PPH decreases with a smaller *needle size*, possibly because of reduced leakage through smaller rents (Table 114-1) (30,33,43,58,80). It is recommended that a 20-gauge needle or smaller be used for diagnostic

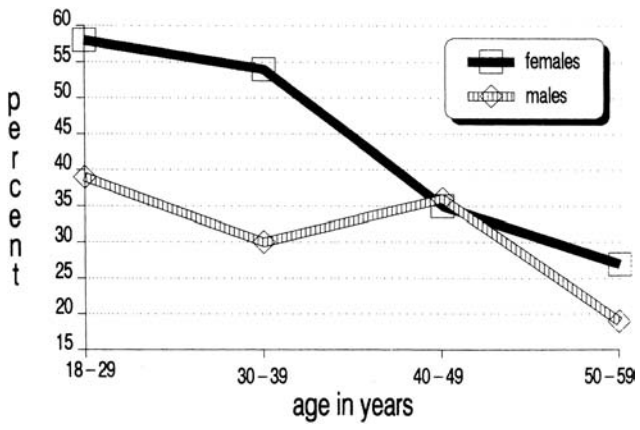

FIGURE 114-4. Incidence of postlumbar puncture headache (PPH) related to age for 150 men and 150 women. There was a significantly higher incidence of PPH in women ($p < 0.025$) and in patients younger than 40 years ($p < 0.005$). A significant difference between the two age groups is only shown for women ($p < 0.005$), and the incidence of PPH is significantly different for the two sexes only below the age of 40 years ($p < 0.01$). From Vilming et al. (88).

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TABLE 114-1 Well-Designed Controlled Studies Regarding the Significance of Needle Size[a]

Design							% PPH					
Author	*Year*	*R*	*B*	*A*	*S*	*n*	*20G*	*22G*	*23G*	*25G*	*26G*	*29G*
Tourtellotte et al. (92)	1972[b]	+	DB	+	−	100		36			12	
Nestvold (60)	1978[b]	+	−	+	+	99	48	27				
Kovanen and Sulkava (44)	1986[b]	−	−	+	−	300	52	37	28			
Flaatten et al. (31)	1989[c]	+	−	+	−	149					7	0
Geurts et al. (34)	1990[c]	+	DB	+	−	80				25		0

PPH, postlumbar puncture headache; +, yes; −, no, or not stated; R, randomized; B, blinded; DB, double blind; A, groups congruent for age; S, groups congruent for sex; n, number of patients; G, gauge.
[a]The table shows details concerning the design of different studies and the incidence of PPH in relation to needle size.
[b]Diagnostic lumbar puncture.
[c]Spinal anesthesia.

LP (14). Also, an oblique penetration of the dura (40), insertion of a Quincke needle with the *bevel parallel* instead of transverse to the longitudinal axis of the dural cylinder reduced the incidence of PPH in one study (29), but this was not proved in another (58). The use of *atraumatic* (blunt-tipped) needles reduce the incidence of PPH (75,78).

The influence of *body position* and *rest* after LP has been the subject of several studies dating back to 1902 (72). Some early reports indicated that bedrest post-LP from 3 to 24 hours reduces the incidence of PPH (11,70) but others (81) and more recent well-controlled studies (3,11,13,15,38,74,87) (Table 114-2) refuted this theory. Accordingly, it is now accepted that bedrest after LP has no prophylactic effect and, in fact, early mobilization may be preferable (3,87). Furthermore, the head-down position in recumbency after LP does not prevent PPH (38,73).

The impact of *personality traits* on the incidence of PPH is unclear. It has been widely believed that apprehensive patients are prone to PPH, but this theory was challenged (79). A recent study indicated that personality profiles of patients with and without PPH were similar, based on the Minnesota Multiple Personality Inventory (85). Furthermore, headache occurs almost as frequently after sham LP as after real LP (42).

The influence of the *underlying diagnosis* (e.g., multiple sclerosis, syphilis) on the incidence of PPH has been investigated but no definite conclusions could be drawn because of lack of prospective, controlled studies.

The following factors are unlikely to influence the incidence of PPH: race, quantity of removed CSF, bloody tap, multiple perforations of the dura, and qualifications of the person performing the LP (79).

Diagnosis

The diagnosis of PPH is made on the basis of its clinical characteristics. MRI may reveal sagging of intracranial structures, pachymeningeal enhancement, dilation of the anterior internal vertebral venous plexus, spinal hygromas, and focal fluid collections in the upper retrospinal region (39,94).

TABLE 114-2 Well-Designed Controlled Studies Regarding the Significance of Body Position after Lumbar Puncture[a]

Design							% PPH					
Author	*Year*	*R*	*B*	*A*	*S*	*n*	*0h*	*2h*	*4h*	*6h*	*24h*	
Carbaat et al. (14)	1981[b]	−	−	+	−	100	38				36	
Andersen et al. (3)	1986[b]	+	−	+	+	112		11			14	
Vilming et al. (89)	1988[b]	+	+	+	+	300	35			39		
Cook et al. (16)	1989[c]	+	+	+	−	102		12			12	
Spriggs et al. (76)	1992[b]	+	+	+	−	110	32		31			

h, hours; +, yes; −, no or not stated; R, randomized; B, blinded; A, groups congruent for age; S, groups congruent for sex; n, number of patients; PPH, postlumbar puncture headache.
[a]The table shows details concerning the design of different studies and the incidence of PPH in relation to bedrest for different lengths of time. No statistically significant differences are observed in any of the studies.
[b]Diagnostic lumbar puncture.
[c]Spinal anesthesia.

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Management

There is no universally accepted standard for the management of PPH except that treatment should not have more side effects than the symptom being treated (41). Often, it is sufficient to advise the patient to lie down as long as PPH lasts and to use mild analgesics when necessary. The clinical practice of advising patients to increase their daily fluid intake after LP is ineffective (20).

Caffeine sodium benzoate 0.5 mg either in 2 mL of saline for bolus injection or in 1 L of Ringer solution for slow infusion and a single oral dose of USP 300 mg anhydrous caffeine powder have shown efficacy in well-conducted studies (12,23,69). Also, theophylline 282 mg three times daily was better than placebo in a controlled study (27). It is of note, however, that the efficacy of these drugs is modest and their long-term effect has not been established.

Case series have indicated that a single lumbar epidural injection of 10 to 60 mL sterile saline provides immediate relief, and recurrence within a few hours occurs in only a few patients (64,81). An alternative to the single injection is the slow infusion of 700 mL of saline at 15 to 30 mL per hour with a success rate of 70 to 100% (7). Epidural injection of as little as 10 mL of saline in the lumbar region raises both epidural and CSF pressures significantly, but the pressure returns to previous values within 3 to 10 minutes (81). It is thought that the beneficial effect of epidural saline is on the basis of sealing a rent (64,81). Side effects of epidural saline infusion include backache, dizziness, nausea, tachypnea, or ocular–frontal pain.

The therapeutic value of an *epidural blood patch* (EBP) has been known for almost 50 years (Gormley) and the procedure gained wide acceptance in the 1970s (21). Several uncontrolled studies indicated that an initial EBP provides immediate relief of PPH in approximately 90% of patients, and almost all patients are relieved after a second EBP (16,21,76). However, data from two small controlled studies (6,70) and a Cochrane Collaboration systematic review (76) indicated lack of sufficient evidence for the routine or prophylactic use of EBP for patients with PPH. Some practitioners recommend EBP for incapacitating pain or PPH lasting longer than 5 days but others perform the procedure as early as day postonset of PPH. EBP may be effective even months after the onset of PPH (91). It is argued that EBP is more effective than an epidural saline injection (6).

The recommended EBP procedure is a slow (0.5 mL/s) injection of 10 to 20 mL of autologous blood near the rent, and a subsequent 1-hour bed rest. Fever, local infection, and coagulopathies are contraindications for EBP (1). Side effects of EBP, which may be troublesome in some people, include nuchal, occipital, or shoulder pain during the procedure, but the symptom could be avoided by a slow injection rate. Also, meningismus may follow an accidental dural puncture. One third of patients who undergo EBP report focal backache for a few days, although rarely patients report pain for weeks to months. Some patients complain of radiating back pain or paresthesias during the injection and for days or weeks thereafter (1).

EBP is effective likely due to initial focal compression of the thecal sac (8) followed by the formation of a seal when blood rapidly coagulates upon contact with the CSF and the bare epithelium and collagen of the rent (65).

Surgical interventions (clipping or suturing of an open dural rent) for the treatment of PPH are sometimes used. Single reports have described the immediate success of surgery, even several months after LP.

CSF FISTULA HEADACHE AND HEADACHE ATTRIBUTED TO SPONTANEOUS (OR IDIOPATHIC) LOW CSF PRESSURE

Definition of CSF Fistula Headache

IHS code and diagnosis: 7.2.2 Cerebrospinal fluid fistula headache

WHO code and diagnosis: G44.88 Headache associated with other specified disorders

Short description: Headache occurs or worsens shortly after assuming the upright position and improves or disappears with recumbency. It is caused by a cerebrospinal fistula, the existence of which may or may not be proven, and disappears within 7 days after successful closure of a fistula.

IHS diagnostic criteria for CSF fistula headache (Headache Classification Committee, 2004) are as follows:

A. Headache that worsens within 15 minutes after sitting or standing, with at least one of the following and fulfilling criteria C and D:
  1. Neck stiffness
  2. Tinnitus
  3. Hyperacusia
  4. Photophobia
  5. Nausea

B. A known procedure or trauma has caused persistent CSF leakage with at least one of the following:
  1. Evidence of low CSF pressure on MRI (e.g., pachymeningeal enhancement)
  2. Evidence of CSF leakage on conventional myelography, computed tomography (CT) myelography, or cisternography
  3. Opening pressure <60 mm H_2O in sitting position

C. Headache develops in close temporal relation to CSF leakage.

D. Headache resolves within 7 days of sealing the CSF leak.

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Definition of Headache Attributed to Spontaneous Low CSF Pressure

IHS code and diagnosis: 7.2.3 Headache attributed to spontaneous (or idiopathic) low CSF pressure

WHO code and diagnosis: G44.88 Headache associated with other specified disorders

Short description and other terms: In 1938, Schaltenbrand used the term "aliquorrhea" to describe a clinical condition presenting chiefly as spontaneous orthostatic headache and very low, unmeasurable, or even negative CSF opening pressure (66,67). To date, these initial cases are diagnosed as spontaneous intracranial hypotension (SIH) (63). The overwhelming majority of SIH cases are caused by spontaneous CSF spinal leaks (50), particularly at the level of the thoracic spine (56). Rarely, CSF leak from the skull base causes SIH.

CSF leak leads to CSF volume depletion. Lately, "CSF hypovolemia" and "CSF volume depletion" have been used interchangeably with SIH as some of the patients with CSF leaks may display consistently normal CSF opening pressures (49,56).

IHS diagnostic criteria for headache attributed to spontaneous (or idiopathic) low CSF pressure (Headache Classification Committee, 2004) are as follows:

A. Diffuse and/or dull headache that worsens within 15 minutes after sitting or standing, with at least one of the following and fulfilling criterion D:
  1. Neck stiffness
  2. Tinnitus
  3. Hypacusia
  4. Photophobia
  5. Nausea

B. At least one of the following:
  1. Evidence of low CSF pressure on MRI (e.g., pachymeningeal enhancement)
  2. Evidence of CSF leakage on conventional myelography, CT myelography, or cisternography
  3. CSF opening pressure <60 mm H_2O in sitting position

C. No history of dural puncture or other cause of CSF fistula.

D. Headache resolves within 72 hours after epidural blood patching.

Etiology

A known procedure or trauma is not required for the diagnosis of headache attributed to low CSF pressure, which is unlike CSF fistula headache. However, a substantial minority of patients do report a history of trivial trauma that preceded the onset of symptoms. Furthermore, factors that predispose to SIH include congenital weakness of the dural sac, meningeal diverticula (similar to Marfan syndrome) (18), disorders of a connective tissue matrix (55), spondylotic spurs (26), or disk herniation (92).

TABLE 114-3 Headache Phenotype in CSF Leak Conditions

- Orthostatic headaches that are preceded by neck pain or interscapular pain or both or by lingering nonorthostatic headaches
- A headache with dampened orthostatic features and with transformation into chronic lingering headaches that can be present even in recumbency. This usually occurs with chronic conditions of CSF leak.
- Acute thunderclaplike headache that mimics subarachnoid hemorrhage before orthostatic features are recognized
- A rare, paradoxically postural headache (i.e., present in recumbency and relieved in the upright position)
- A "second-half-of-the-day headache." Typically, a patient is headache free in the morning but the pain starts and worsens later in the day as the patient remains in the upright position. In some, the headache improves with recumbency but it does not in others.
- Isolated exertional headaches with or without nonheadache manifestations of CSF leaks, and without orthostatic features
- Recurring and remitting headaches that parallel the course of intermittent CSF leaks
- Acephalgic SIH despite documentation of a CSF leak, low CSF pressures, and typical MRI abnormalities

CSF, cerebrospinal fluid; MRI, magnetic resonance imaging; SIH, spontaneous intracranial hypotension

Clinical Manifestations

Spontaneous intracranial hypotension headache is classically orthostatic (i.e., present in the upright position and relieved by recumbency) (63,67), and is often aggravated by Valsalva-type maneuvers. However, not all CSF leak headaches are orthostatic (Table 114-3) (51), and not all orthostatic headaches result from CSF leaks (e.g., with postural tachycardia syndrome [POTS]) (54).

The headache of SIH is often bilateral and nonthrobbing, and the pain location is variable.

Clinical Manifestations Other Than Headaches

One, and more often several, nonheadache symptoms accompany SIH (Table 114-4) (51).

Diagnosis

CSF examination: The CSF opening pressure is typically low, sometimes unmeasurable, and, rarely, even negative. In a minority of patients, however, opening pressures are consistently within normal limits. The fluid color is clear but can occasionally turn xanthochromic. The fluid

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TABLE 114-4 Clinical Features Other Than Headaches

- Interscapular or neck pain (sometimes orthostatic), low back pain
- Nausea, sometimes emesis—often orthostatic
- Horizontal diplopia (unilateral or bilateral sixth cranial nerve palsy)
- Cochleovestibular manifestations (dizziness, change in hearing, tinnitus)
- Photophobia, visual blurring
- Upper limb numbness or pain
- Rare manifestations: facial numbness or weakness, diplopia due to third or fourth cranial nerve palsy, galactorrhea, Ménière disease–like symptoms, upper limb radicular symptoms, encephalopathy, stupor, coma, parkinsonism, ataxia, sphincter trouble, gait unsteadiness, frontotemporal dementia

chemistry profile is that of normal or elevated protein (commonly up to 100 mg/dL and rarely up to 1000 mg/dL) (56) and normal glucose concentrations. The leukocyte count is normal or mildly elevated (up to 50 cells per mm^3) but counts exceeding 222 cells per mm^3 have been reported (56). The erythrocyte count is typically normal and so is cytology and microbiology.

Indium-111 radioisotope cisternography: Normally, substantial radioactivity is detected over the cerebral convexities at 24 hours or earlier. In CSF leaks, however, radioactivity typically does not extend beyond the basal cisterns in the first 24 hours, and its minimal appearance, if any, over the cerebral convexities is delayed from 24 to 48 hours (5,9). Also, radioactivity appears early in the kidneys and urinary bladder (less than 4 hours versus 6 to 24 hours under normal circumstances). Finally, a parathecal (paradural) activity that may point to the approximate site of the leak may be noted (Fig. 114-5). It should be mentioned that meningeal diverticula manifest as foci of parathecal activity that could mimic a site of CSF leakage and may not necessarily be the site of CSF leak.

CT: Head CT is usually normal and of limited diagnostic value. Infrequently, a head CT may reveal subdural fluid collections or increased tentorial enhancement following contrast administration.

MRI (Table 114-5): On head MRI (Fig. 114-6-A and D), a diffuse pachymeningeal and leptomeningeal enhancement pattern is most commonly observed. Enhancement is typically linear, nonnodular, uninterrupted, bilateral, and involving both supra- and infratentorial regions (4,29,52,54,57,61,64). Other MRI signs include cerebellar tonsils descent that may mimic type I Chiari malformations (Fig. 114-7) (54), bilateral subdural fluid collections that may (subdural hematoma) or may not (subdural hygroma) compress the underlying sulci (59), and pituitary gland enlargement that may mimic a pituitary adenoma or hyperplasia (Fig. 114-2-A) (54).

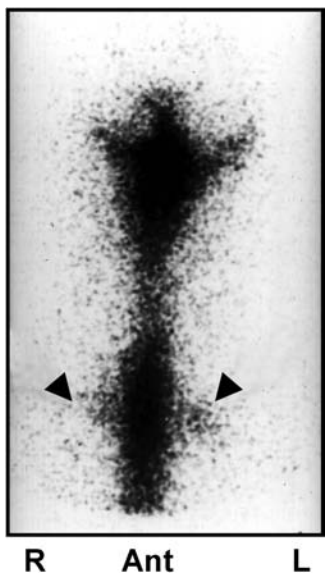

FIGURE 114-5. Indium-111 cisternogram. Note that radioactivity has not extended beyond the basal cisterns. Parathecal activity (arrowheads) points to the extrathecal egress of cerebrospinal fluid (CSF) as the approximate site of the CSF leak.

Spinal fluid collections can be detected on spinal MRI (Table 114-5) and can be either extra-arachnoid (62), which often extend to several levels and rarely reveal the exact leak site, or extradural, which span only a few spinal segments but more likely reveal a leak site.

CT-myelography is a reliable and accurate test that may show extra-arachnoid and extradural extravasation of contrast, meningeal diverticula, dilated nerve root sleeves, and the actual leakage site (Fig. 114-2-B and C) (56). Fast and slow flow CSF leaks may present special challenges, however. Fast flow leaks cause a substantial amount of contrast to distribute across several levels of the spine quite

TABLE 114-5 MRI Abnormalities in CSF Leaks

Head MRI
Diffuse pachymeningeal enhancement
Descent ("sagging" or "sinking") of the brain (descent of cerebellar tonsils, obliteration of prepontine or perichiasmatic cisterns, crowding of the posterior fossa)
Enlargement of the pituitary gland
Flattening of the optic chiasm
Subdural fluid collections
Engorged cerebral venous sinuses
Decrease in size of the ventricles
Spine MRI
Extra-arachnoid or extradural fluid collections
Meningeal diverticula
Spinal pachymeningeal enhancement
Engorgement of spinal epidural venous plexus

MRI, magnetic resonance imaging.

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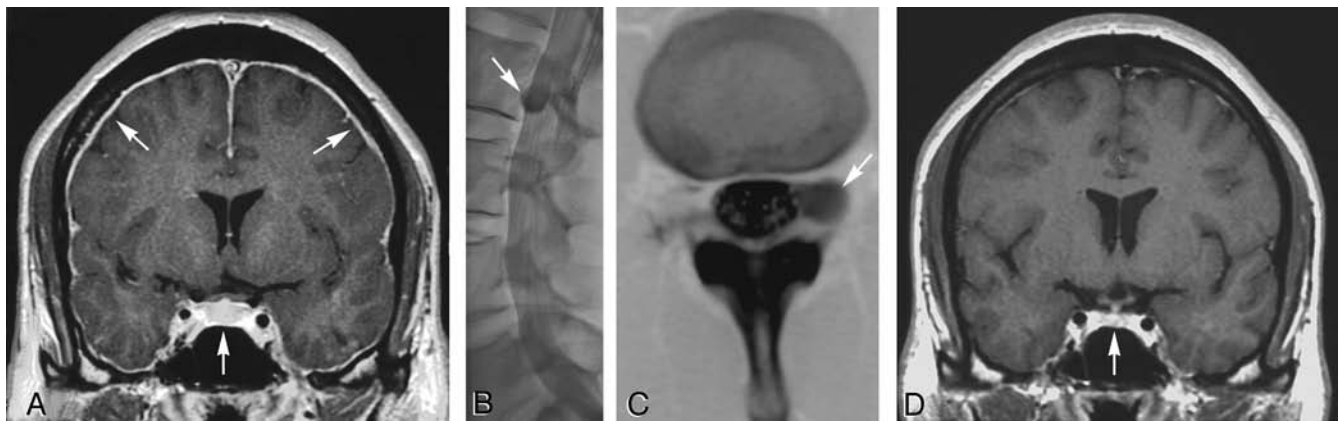

FIGURE 114-6. (*A*) Coronal T1-weighted gadolinium-enhanced head magnetic resonance imaging (MRI) shows diffuse pachymeningeal enhancement (*upper arrows*) as enlargement of pituitary gland (*lower arrow*). Also note the flattened optic chiasm above the pituitary and partially obliterated perichiasmatic cistern (both related to sinking of the brain). (*B*) Myelogram and (*C*) computed tomography myelogram show a meningeal diverticulum at L2 (*arrows*). (*D*) Coronal T1-weighted gadolinium-enhanced head MRI after clinical recovery shows reversal of all of the abnormalities noted in the previous MRI (*A*). Also note that during the symptomatic phase (*A*) when compared with the postrecovery (*D*), the size of the lateral ventricles is somewhat smaller (a manifestation of sinking of the brain and "ventricular collapse"). (From *Mayo Clin Proc* 2002;77:1241–1246, with permission of Mayo Foundation).

rapidly, making it difficult to find the leak site. Dynamic CT-myelography (46) can resolve this problem through the use of a high-speed, multidetector spiral apparatus that allows scanning multiple levels over a short period of time. In contrast, slow CSF leaks can lead to false-negative results if images are completed before enough contrast has oozed out. Delayed imaging techniques (e.g., 3- to 4-hour delayed CT, MR-myelography [77]) may be helpful in such instances.

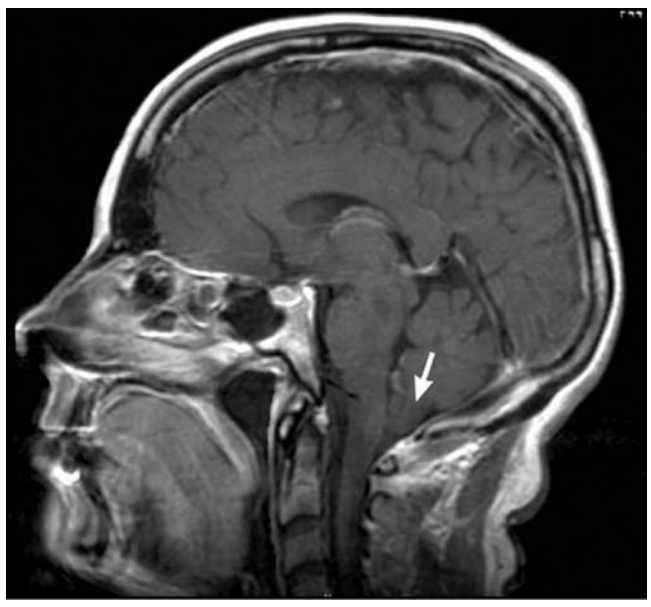

FIGURE 114-7. Median sagittal magnetic resonance imaging demonstrates crowding of the posterior fossa and descent of the cerebellar tonsils (*arrow*).

Treatment

A variety of treatment modalities are advocated for SIH (Table 114-6), when a leak does not stop spontaneously. Bedrest and adequate (over-) hydration are traditional measures that are used but without clear evidence of their benefit.

Pharmacologic interventions using caffeine, theophylline, or steroids may be beneficial for some patients (89), although the effect is reportedly marginal and unsustained.

Epidural infusion of saline (34) or dextran (2), epidural injection of fibrin glue (17), and intradural or even

TABLE 114-6 Treatment of CSF Leaks

- Bedrest
- Hydration/overhydration
- Caffeine, theophylline
- Steroids
- Abdominal binder
- Epidural blood patch
- Continuous epidural saline infusion
- Epidural infusion of dextran
- Epidural injection of fibrin glue
- CSF shunting
- Intrathecal fluid infusion
- Surgery

CSF, cerebrospinal fluid.

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intrathecal fluid infusions are recommended occasionally. An intrathecal infusion may be helpful in rare cases of evolving obtundation or coma (10). These invasive procedures should be used sparingly because of concerns of complications such as infections.

EBPs are in many accounts the treatment of choice in patients who have failed initial conservative treatment (25,73), but the beneficial effect may not be sustained in approximately one third of the patients, requiring several EBPs.

Surgical repair of a CSF leak is considered when conservative and less invasive approaches (e.g., EBP) fail (68). It is essential to determine the site of the CSF leak before surgery is undertaken.

Occasionally, rebound symptomatic intracranial hypertension develops after treatment of spontaneous CSF leaks, whether by EBP or surgery (52). This potential complication is often self limiting and can be managed with acetazolamide. Rarely, CSF shunting may be needed.

REFERENCES

1. Abouleish E, de la Vega S, Blendinger I, et al. Long-term follow-up of epidural blood patch. *Anesth Analg* 1975;54:459–463.
2. Aldrete JA. Persistent post-dural puncture headache treated with epidural infusion of Dextran. *Headache* 1994;34:265–267.
3. Anderson APD, Wanscher MCJ, Hüttel MS. "Postspinaler" Kopfschmerz. Ist die 24-stündige flache Bettruhe eine Prophylaxe? *Reg Anesth* 1986;9:15–17.
4. Atkinson JLD, Weinshenker BG, Miller GM, et al. Acquired Chiari I malformation secondary to spontaneous spinal cerebrospinal fluid leakage and chronic intracranial hypotension syndrome in seven cases. *J Neurosurg* 1988;88:237–242.
5. Bai J, Yokoyama K, Kinuya S, et al. Radionuclide cisternography in intracranial hypotension syndrome. *Ann Nucl Med* 2002;16:75–78.
6. Bart AJ, Wheeler AS. Comparison of epidural saline placement and epidural blood placement in the treatment of post-lumbar-puncture headache. *Anesthesiology* 1978;48:221–223.
7. Baysinger CL, Menk EJ, Harte E, et al. The successful treatment of dural puncture headache after failed epidural blood patch. *Anesth Analg* 1986;65:1242–1244.
8. Beards SC, Jackson A, Griffiths AG, et al. Magnetic resonance imaging of extradural blood patches: appearances from 30 min to 18 h. *Br J Anaesth* 1993;71:182–188.
9. Benamer M, Tainturier C, Graveleau P, et al. Radionuclide cisternography in spontaneous intracranial hypotension. *Clin Nucl Med* 1998;23:150–151.
10. Binder DK, Dillon WP, Fishman RA. Intrathecal saline infusion in the treatment of obtundation associated with spontaneous intracranial hypotension. Technical case report. *Neurosurgery* 2002;51:830–837.
11. Brocker RJ. Technique to avoid spinal-tap headache. *JAMA* 1958;168: 261–263.
12. Camann WR, Murray RS, Mushlin PS, et al. Effects of oral caffeine on postdural puncture headache: a double-blind, placebo-controlled trial. *Anesth Analg* 1990;70:181–184.
13. Carbaat PAT, van Crevel H. Lumbar puncture headache: controlled study on the preventive effect of 24 hours' bed rest. *Lancet* 1981;2: 1133–1135.
14. Carson D, Serpell M. Choosing the best needle for diagnostic lumbar puncture. *Neurology* 1996;47:33–37.
15. Cook PT, Davies MJ, Beavis RE. Bed rest and postlumbar puncture headache. The effectiveness of 24 hours' recumbency in reducing the incidence of postlumbar puncture headache. *Anesthesia* 1989;44: 389–391.
16. Crawford JS. Epidural blood patch. *Anaesthesia* 1985;40:381.
17. Cru JBP, Gerritse BM, Van Dongen RTM, Schoonderwaldt HC. Epidural fibrin glue injection stops persistent postdural puncture headache. *Anesthesiology* 1999;91:576–577.
18. Davenport RJ, Chataway SJ, Warlow CP. Spontaneous intracranial hypotension from a CSF leak in a patient with Marfan's syndrome. *J Neurol Neurosurg Psychiatry* 1995;59:516–519.
19. Dietrich M, Brandt T. Is obligatory bed rest after lumbar puncture obsolete? *Eur Arch Psychiatry Neurosci* 1985;235:71–75.
20. Dietrich M, Brandt T. Incidence of post-lumbar puncture headache is independent of daily fluid intake. *Eur Arch Psychiatry Neurosci* 1988;237:194–196.
21. DiGiovanni AJ, Galbert MW, Wahle WM. Epidural injection of autologous blood for postlumbar-puncture headache. II. Additional clinical experiences and laboratory investigation. *Anesth Analg* 1972;51: 226–232.
22. Dittman M, Schäfer HG, Ulrich J, et al. Anatomical re-evaluation of lumbar dura mater with regard to postspinal headache. *Anesthesia* 1988;43:635–637.
23. Dodd JE, Efird RC, Rauck RL. Cerebral blood flow changes with caffeine therapy for postdural headaches. *Anesthesiology* 1989;71:A679.
24. Duffy PJ, Crosby ET. The epidural blood patch. Resolving the controversies. *Can J Anesth* 1999;46:878–886.
25. Eckstein KL, Rogacev Z, Vicente-Eckstein A, et al. Prospektiv vergleichende Studie postspinaler Kopfschmerzen bei jungen Patienten (<51 Jahre). *Reg Anesth* 1982;5:57–61.
26. Eros EJ, Dodick DW, Nelson KD. Orthostatic headache syndrome with CSF leak secondary to bony pathology of the cervical spine. *Cephalalgia* 2002;22:439–443.
27. Feurerstein TJ, Zeides A. Theophylline relieves headache following lumbar puncture: placebo-controlled, double-blind pilot study. *Klin Wochenschr* 1986;64:216–218.
28. Fishman RA, Dillon WP. Dural enhancement and cerebral displacement secondary to intracranial hypotension. *Neurology* 1993;43: 609–611.
29. Flaatten H, et al. Puncture technique and postural postdural puncture headache. A randomised, double-blind study comparing transverse and parallel puncture. *Acta Anaesthesiol Scand* 1998;42: 1209–1214.
30. Flaatten H, Rodt SA, Vamnes J, et al. Postdural puncture headache: a comparison between 26 and 29-gauge needles in young patients. *Anaesthesia* 1989;44:147–149.
31. Forbes HS, Nason GI. The cerebral circulation: vascular responses to (A) hypertonic solutions and (B) withdrawal of cerebrospinal fluid. *Arch Neurol Psychiatry* 1935;34:533–547.
32. Franksson C, Gordh T. Headache after spinal anesthesia and a technique for lessening its frequency. *Acta Chir Scand* 1946;94:443–454.
33. Geurts JW, Haanschoten MC, van Wijk RM, et al. Post-dural puncture headache in young patients: a comparative study between the use of 0.52 mm (25-gauge) and 0.33 mm (29-gauge) spinal needles. *Acta Anesthesiol Scand* 1990;34:350–353.
34. Gibson BE, Wedel DJ, Faust RJ, et al. Continuous epidural saline infusion for the treatment of low CSF pressure headache. *Anesthesiology* 1988;48:789–791.
35. Gobel H, Schenkl S. Post-lumbar puncture headache: the relation between experimental suprathreshold pain sensitivity and a quasi-experimental clinical pain syndrome. *Pain* 1990;40:267–278.
36. Grant R, Condon B, Hart I, et al. Changes in intracranial CSF volume after lumbar puncture and their relationship to post-LP headache. *J Neurol Neurosurg Psychiatry* 1991;54:440–442.
37. Grant R, Condon B, Patterson J, et al. Changes in cranial CSF volume during hypercapnia and hypocapnia. *J Neurol Neurosurg Psychiatry* 1989;52:218–222.
38. Handler CE, Smith FR, Perkin GD, et al. Posture and lumbar puncture headache: a controlled trial in 50 patients. *J R Soc Med* 1982;75:404–407.
39. Hannerz J, Ericson K, Bro Skejo HP. MR imaging with gadolinium in patients with and without post-lumbar puncture headache. *Acta Radiol* 1999;60:183–188.
40. Hatfalvi BI. The dynamics of post-spinal headache. *Headache* 1977;17:64–66.
41. Hilton-Jones D. What is postlumbar puncture headache and is it avoidable? In: Warlow C, Garfield J, eds. *Dilemmas in the management*

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of the neurological patient. London: Churchill Livingstone, 1984:144–157.
42. Kaplan G. The psychogenic etiology of headache post lumbar puncture. *Psychosom Med* 1967;29:376–379.
43. Kovanen J, Sulkava R. Duration of postural headache after lumbar puncture: effect of needle size. *Headache* 1986;26:224–226.
44. Kunkle EC, Ray BS, Wolff HG. Experimental studies on headache. Analysis of the headache associated with changes in intracranial pressure. *Arch Neurol Psychiatry* 1943;49:323–358.
45. Kuntz KM, Kokmen E, Stevens JC, et al. Post-lumbar puncture headaches: experience in 501 consecutive procedures. *Neurology* 1992;42:1884–1887.
46. Leutmer PH, Mokri B. Dynamic CT myelography: a technique for localizing high-flow spinal cerebrospinal fluid leaks. *AJNR Am J Neuroradiol* 2003;24:1711–1714.
47. Marshall J. Lumbar-puncture headache. *J Neurol Neurosurg Psychiatry* 1950;13:71–74.
48. Michel O, Brusis T. Hearing loss as a sequel of lumbar puncture. *Ann Otol Rhinol Laryngol* 1992;101:390–394.
49. Miyazawa K, Shiga Y, Hasegawa T, et al. CSF hypovolemia vs intracranial hypotension in "spontaneous intracranial hypotension syndrome." *Neurology* 2003;60:941–947.
50. Mokri B. Spontaneous cerebrospinal fluid leaks: from intracranial hypotension to cerebrospinal fluid hypovolemia—evolution of a concept. *Mayo Clin Proc* 1999;74:1113–1123.
51. Mokri B. Low cerebrospinal fluid pressure syndromes. *Neurol Clin North Am* 2004;22:55–74.
52. Mokri B. Intracranial hypertension after treatment of spontaneous cerebrospinal fluid leaks. *Mayo Clin Proc* 2002;77:1241–1246.
53. Mokri B, Atkinson JLD. False pituitary tumor in CSF leaks. *Neurology* 2000;55:573–575.
54. Mokri B, Low PA. Orthostatic headaches without CSF leak in postural tachycardia syndrome. *Neurology* 2003;61:980–982.
55. Mokri B, Maher CO, Sencakova D. Spontaneous CSF leaks: underlying disorder of connective tissue. *Neurology* 2002;58:814–816.
56. Mokri B, Piepgras DG, Miller GM. Syndrome of orthostatic headaches and diffuse pachymeningeal gadolinium enhancement. *Mayo Clin Proc* 1997;72:400–413.
57. Nelson MO. Postpuncture headaches. A clinical and experimental study on the cause and prevention. *Arch Dermatol Syph* 1930;21:615–627.
58. Nestvold K. The influence of lumbar puncture needle size on postpuncture complications particularly with regard to patients under day-care and fully hospitalized patients. *Tidsskr Nor Laegeforen* 1978;98:569–570.
59. Pannullo SC, Reich JB, Krol G, et al. MRI changes in intracranial hypotension. *Neurology* 1993;43:919–926.
60. Pickering GW. Lumbar puncture headache. *Brain* 1948;71:274–280.
61. Plum F, Siesjo BK. Recent advances in CSF physiology. *Anesthesiology* 1975;42:708–730.
62. Rabin BM, Roychowdhury S, Meyer JR, et al. Spontaneous intracranial hypotension: spinal MRI findings. *Am J Neuroradiol* 1998;19:1034–1039.
63. Rando TA, Rishman RA. Spontaneous intracranial hypotension: Report of two cases and review of literature. *Neurology* 1992;42:481–487.
64. Rice GG, Dabbs CH. The use of peridural and subarachnoid injections of saline solution in the treatment of severe postspinal headache. *Anesthesiology* 1950;11:17–23.
65. Rosenberg PH, Heavner JE. In vitro study of the effect of epidural blood patch on leakage through a dural puncture. *Anesth Analg* 1985;64:501–504.
66. Schaltenbrand G. Neure anschauungen zur pathophysiologie der liquorzirkulation. *Zentrablbl Neurochir* 1938;3:290–300.
67. Schaltenbrand G. Normal and pathological physiology of the cerebrospinal fluid circulation. *Lancet* 1953;1:805–808.
68. Scheivink WI, Morreale VM, Atkinson JLD, et al. Surgical treatment of spontaneous spinal cerebrospinal fluid leaks. *J Neurosurg* 1998;88:243–246.
69. Sechzer PH, Abel L. Post-spinal anesthesia headache after lumbar puncture. *Lancet* 1980;1:1245.
70. Seebacher J, Ribeiru V, LeGuillou JL, et al. Epidural blood patch in the treatment of post dural puncture headache: a double-blind study. *Headache* 1989;29:630–632.
71. Sencakova D, Mokri B, McClelland RL. The efficacy of epidural blood patch in spontaneous CSF leaks. *Neurology* 2001;57:1921–1923.
72. Sicard JA. *Le liquide céphalo-rachidien*. Paris: Masson et Gautier-Villars, 1902.
73. Smith FR, Perkin GD, Clifford Rose F. Posture and headache after lumbar puncture. *Lancet* 1980;1:1245.
74. Spriggs DA, Burn DJ, French J, et al. Is bed rest useful after diagnostic lumbar puncture? *Postgrad Med J* 1992;68:581–583.
75. Strupp M, Schueler O, Straube A, et al. "Atraumatic" Sprotte needle reduces the incidence of post-lumbar puncture headaches. *Neurology* 2001;57:2310–2312.
76. Sudlow C, Warlow C. Epidural blood patching for preventing and treating post-dural puncture headache. *The Cochrane Library* 2004;vol 1.
77. Tali ET, Ercan N, Krumina G, et al. Intrathecal gadolinium (gadopentetate dimeglumine). Enhanced magnetic resonance myelography and cisternography. Results of a multicenter study. *Invest Radiol* 2002;37:152–159.
78. Thomas SR, Jamieson DRS, Muir KW. Randomised controlled trial of atraumatic versus standard needles for diagnostic lumbar puncture. *BMJ* 2000;321:986–990.
79. Tourtellotte WW, Haerer AF, Heller GL, et al. *Post-lumbar puncture headaches*. Springfield, IL: Charles C. Thomas, 1964.
80. Tourtellotte WW, Henderson WG, Tucker RP, et al. A randomized, double-blind clinical trial comparing the 22 versus 26 gauge needle in the production of the post-lumbar puncture syndrome in normal individuals. *Headache* 1972;12:73–78.
81. Usubiaga JE, Usubiaga LE, Brea LM, et al. Effect of saline injections on epidural and subarachnoid space pressures and relation to postspinal anesthesia headache. *Anesth Analg* 1967;46:293–296.
82. Vandam LD, Dripps RD. Long-term follow-up of patients who received 10,098 spinal anesthetics. *JAMA* 1956;161:586–591.
83. Vilming ST, Kloster R. Post-lumbar puncture headache: clinical features and suggestions for diagnostic criteria. *Cephalalgia* 1997;17:778–784.
84. Vilming ST, Kloster R. The time course of post-lumbar puncture headache. *Cephalalgia* 1998;18:97–100.
85. Vilming ST, Ellertsen B, Troland K, et al. MMPI profiles in post-lumbar puncture headache. *Acta Neurol Scand* 1997;95:184–188.
86. Vilming ST, Kloster R, Sandvik L. The importance of sex, needle size, height, and body mass index in post-lumbar puncture headache. *Cephalalgia* 2001;21:738–743.
87. Vilming ST, Schrader H, Monstad I. Post-lumbar-puncture headache: the significance of body posture. A controlled study of 300 patients. *Cephalalgia* 1988;8:75–78.
88. Vilming ST, Schrader H, Monstad I. The significance of age, sex, and cerebrospinal fluid pressure in post-lumbar-puncture headache. *Cephalalgia* 1989;9:99–106.
89. Vilming ST, Titus F. Low cerebrospinal fluid pressure. In: Olesen J, Tfelt-Hansen P, Welch KMA, eds. *The headache*. New York: Raven Press, 1993:687–695.
90. Wang LP, Schmidt JF. Central nervous side effects after lumbar puncture: a review of the possible pathogenesis of the syndrome of postdural puncture headache and associated symptoms. *Dan Med Bull* 1997;44:79–81.
91. Wilton NCT, Globerson JH, de Rosayro AM. Epidural blood patch for postdural puncture headache: it's never too late. *Anesth Analg* 1986;65:895–896.
92. Winter SCA, Maartens NF, Amslow P, et al. Spontaneous intracranial hypotension due to thoracic disc herniation. *J Neurosurg (Spine 3)* 2002;96:343–345.
93. Wolff HG. Headache mechanisms. *Int Arch Allergy* 1955;7:210–278.
94. Yousry I, et al. Cervical MR imaging in postural headache: MR signs and pathophysiological implications. *AJNR Am J Neuroradiol* 2001;22:1239–1250.
95. Yu-Lin H, Chia-Erh C. Headache following spinal anesthesia. *Chin Med J* 1951;69:251–257.