Neurosurg Rev (2010) 33:271–285
DOI 10.1007/s10143-010-0266-5

REVIEW

Pathogenesis of syringomyelia associated with Chiari type 1 malformation: review of evidences and proposal of a new hypothesis

Izumi Koyanagi · Kiyohiro Houkin

Received: 30 November 2009 / Revised: 23 March 2010 / Accepted: 2 May 2010 / Published online: 8 June 2010

Abstract The exact pathogenesis of syringomyelia associated with Chiari type 1 malformation is unknown, although a number of authors have reported their theories of syrinx formation. The purpose of this review is to understand evidences based on the known theories and to create a new hypothesis of the pathogenesis. We critically review the literatures on clinicopathological, radiological, and clinical features of this disorder. The previously proposed theories mainly focused on the driven mechanisms of the cerebrospinal fluid (CSF) into the spinal cord. They did not fully explain radiological features or effects of surgical treatment such as shunting procedures. Common findings of the syrinx in clinicopathological studies were the communication with the central canal and extracanalicular extension to the posterior gray matter. Most of the magnetic resonance imaging studies demonstrated blockade and alternated CSF dynamics at the foramen magnum, but failed to show direct communication of the syrinx with the CSF spaces. Pressure studies revealed almost identical intrasyrinx pressure to the subarachnoid space and decreased compliance of the spinal CSF space. Recent imaging studies suggest that the extracellular fluid accumulation may play an important role. The review of evidences promotes a new hypothesis of syrinx formation. Decreased absorption mechanisms of the extracellular fluid may underlie the pathogenesis of syringomyelia. Reduced compliance of the posterior spinal veins associated with the decreased compliance of the spinal subarachnoid space will result in disturbed absorption of the extracellular fluid through the intramedullary venous channels and formation of syringomyelia.

Keywords Syringomyelia · Chiari type 1 malformation · Extracellular fluid · Compliance · Cerebrospinal fluid · Pathogenesis

I. Koyanagi (✉) · K. Houkin
Department of Neurosurgery,
Sapporo Medical University School of Medicine,
South 1, West 16, Chuo-ku,
Sapporo 060-8543, Japan
e-mail: koyai@sapmed.ac.jp

Introduction

The exact pathogenesis of syringomyelia associated with Chiari type 1 malformation has not been clarified. This disorder is characterized by ectopia of the cerebellar tonsils with or without displacement of the brainstem through the foramen magnum. Disturbed pathway of the cerebrospinal fluid (CSF) around the foramen magnum is assumed to be the primary cause of syringomyelia. However, hydrocephalus is usually absent, and the degrees of subarachnoid blockade at the foramen magnum and descent of the cerebellar tonsils are not associated with presence or absence of syringomyelia. Although recent advances of neuroradiological imaging provided static and dynamic information on the anatomical structures around the foramen magnum, none of the previously reported theories fully explained the clinical or radiological features. Until now, no animal models successfully reproduced this disorder. In the known experimental models, syringomyelia was produced by induction of adhesive arachnoiditis, spinal cord injury or hydrocephalus.

In this article, we critically review the previously proposed theories and clinical studies of syringomyelia associated with Chiari type 1 malformation. The anatomical and pathophysiological evidences are analyzed to infer the mechanisms of syrinx formation. The purpose of this review is to create a new hypothesis for the pathogenesis of syringomyelia associated with Chiari type 1 malformation.

Previous theories for the pathogenesis

Table 1 summarizes the previously reported theories. Most of theories in 1900s focused on how CSF entered into the spinal cord as the pathogenesis of syringomyelia [16, 57–59, 130, 145, 200, 201]. The main source of CSF entrance was considered to be the fourth ventricle via the central canal [57–59, 200, 201] or the spinal subarachnoid space via the perivascular spaces [16, 130, 145]. The latter theory that the syrinx fluid originates from the subarachnoid CSF has been supported by many clinical or experimental studies. However, the subarachnoid CSF origin theory was not based on direct evidences. Recent articles in 2000s proposed that the syrinx fluid derived from the extracellular fluid from the spinal cord microcirculation, not from the CSF in the subarachnoid space or the fourth ventricles [69, 70, 104, 115]. These studies did not show new clinical evidences but provided novel insights into the pathogenesis of syringomyelia. The idea that the syrinx fluid originates from the extracellular fluid may explain the

Table 1 The authors and study descriptions of the known theories for the pathogenesis of syrngomyelia associated with Chiari type 1 malformation

Authors	Theory	Study description
CSF entrance from the fourth ventricle		
[57–59]	Ventricular CSF fluid enters into the central canal by the arterial pulsation. Narrowed portion of the central canal acts as a one-way valve	Speculation from clinical studies on patients with Chiari malformation who underwent posterior surgeries (17 patients in 1950; 45 patients in 1958; 68 patients in 1965). The theory assumed presence of mild or compensated hydrocephalus
[200, 201]	The pressure dissociation between the intracranial and spinal subarachnoid spaces secondary to venous pressure changes sucks the fourth ventricle CSF into the central canal	Speculation based on common clinical observations and the literature review and the clinical study in 37 patients with syringomyelia and Chiari malformation. The lumbar CSF pressure became higher firstly by cough or Valsalva maneuver and fell faster than the ventricle pressure in 24 out of 37 patients
CSF entrance from the subarachnoid space		
[16]	CSF enters into the spinal cord via perivascular spaces. The elevated thoraco-abdominal pressures are transmitted via the epidural venous plexus to the spinal subarachnoid space	Speculation from the pathological study using human spinal cord specimen with syringomyelia. Intra-syrinx injection of Indian ink resulted in spread and pool in the dilated perivascular spaces
[131]	Tonsillar herniation blocks the upward flow of the central canal fluid. CSF may enter into the spinal cord via the perivascular spaces	Speculation from the clinical and pathological study on 20 autopsy specimens (6 fetuses and 14 adults) and 45 patients with hindbrain lesions including 25 patients of Chiari type 1 malformation
[145]	Piston action of the cerebellar tonsil forces the subarachnoid CSF into the spinal cord through the perivascular or interstitial spaces	Speculation from the clinical study using phase-contrast MR imaging and intraoperative ultrasonography findings in 7 patients with syringomyelia and Chiari type 1 malformation
Extracellular fluid origin		
[69, 70]	Syringomyelia is produced by mechanical distension of the spinal cord and filling with extracellular fluid from the spinal cord microcirculation	Speculation based on phase-contrast MR imaging study on 16 patients with spinal cord cysts including 7 patients with Chiari type 1 malformation, and literature review
[104]	Syringomyelia is originated from accumulation of the extracellular fluid in the spinal cord	Speculation from the literature review
[115]	Dilatation of intramedullary vessels below the subarachnoid blockade partially disrupts the blood-cord barrier and produces the syrinx with accumulation of the fluid from the intramedullary microcirculation	Speculation from the literature review

pathophysiology of syrinx formation in adhesive spinal arachnoiditis but is still difficult to explain effectively the mechanism in Chiari type 1 malformation.

Clinicopathological studies

There have been only several studies reporting human spinal cord specimens of syringomyelia with Chiari type 1 malformation. In 1953, Netsky reported autopsy findings of 8 patients with syringomyelia and found abnormal vessels around the syringes [141]. He suggested that the intramedullary abnormal vessels were the cause of syringomyelia. However, Chiari malformation was present in only one patient in the series. From 1987 to 1996, autopsy findings of 18 cases of syringomyelia with Chiari type 1 malformation were reported in four papers [20, 80, 91, 132]. These studies demonstrated that there was no direct communication between the fourth ventricle and the syrinx, but the central canal to the fourth ventricle was patent in eight of these 18 cases. Ependymal lining of the syrinx or communication of the syrinx with the central canal was observed in all cases. The syrinx usually extended into the posterior gray matter and sometimes communicated with the spinal subarachnoid space.

Radiological evidences of CSF dynamics

CT-scan with intrathecal water-soluble contrast materials

Computed tomographic (CT) scan after intrathecal administration of water-soluble contrast materials (CT myelography (CTM)) was introduced for radiological examination of syringomyelia in the end of 1970s [51, 159]. The delayed CTM several hours after intrathecal injection of metrizamide (MW 789) displayed enhancement of syringomyeliac cavities [13, 26, 27, 29, 35, 100, 101, 111, 117, 168, 198, 206]. Such CTM findings supported the theory of parenchymal CSF entrance because the contrast medium injected into the spinal subarachnoid space was accumulated in the syrinx without entrance into the fourth ventricle. Similar intramedullary contrast accumulation was also present in other intramedullary cystic lesions in cervical spondylosis, intramedullary tumors, and syringomyelia due to other etiologies [95, 99].

Several studies demonstrated dynamics of the intrathecally injected water-soluble contrast materials in the normal spinal cord. These studies indicated that a significant part of the intrathecally injected metrizamide was eliminated to the blood via the spinal routes in rabbits [66] and humans [45, 146]. It is also known that the intrathecally injected water-soluble contrast materials penetrate into the normal brain and spinal cord parenchyma in dogs [40, 161], rabbits [44, 85] and humans [86, 88, 203]. The mechanism of metrizamide penetration from the subarachnoid space into the spinal cord was thought to be a simple diffusion because of lack of a barrier between subarachnoid CSF and the extracellular fluid of the spinal cord. Tracer studies using HRP (MW 43,000) demonstrated rapid entrance of the subarachnoid HRP into the spinal cord [176, 177] or the brain [179] via the perivascular spaces in normal rats, cats, dogs and sheep. These studies suggested the role of arterial pulsation as a driving force.

Considering the results of CTM and tracer studies, intramedullary penetration of the water-soluble contrast materials from the subarachnoid space will not be specific to syringomyelia. Delayed clearance of the contrast from the syrinx cavities may explain delayed visualization of the syrinx in CTM.

CSF dynamics by cine-mode MR imaging

Cine-mode magnetic resonance (MR) imaging enables analysis of CSF dynamics in a cardiac cycle in the patients with Chiari type 1 malformation. Most of the published studies utilized phase-contrast techniques [3, 6, 21, 28, 38, 69, 74, 76, 83, 90, 105, 120, 126–128, 150, 154–156, 174, 196, 204]. Some studies demonstrated CSF movement as the displacement of the bands [185] or stripes [164]. According to these MR studies, there was a significant variety in the degree of subarachnoid blockade and physiological parameters of the CSF flow in Chiari type 1 malformation. The CSF movement in the posterior subarachnoid space at the foramen magnum was disturbed or completely blocked by the displaced cerebellar tonsils. However, some studies on pediatric population reported normal CSF flow in 19–33% of the patients with Chiari type 1 malformation [126, 127, 196]. The reported data on the CSF velocities in the spinal subarachnoid space were more confusing. Some studies [3, 6, 21, 164] reported that the systolic CSF velocities in Chiari patients were lower than those in healthy controls. Other studies [76, 89, 120] reported significantly higher systolic velocities. Simultaneous bidirectional CSF flow at the foramen magnum was also reported [190]. None of cine-mode MR imaging studies showed CSF entrance from the fourth ventricle or the spinal subarachnoid space into the syrinx. Also, most of them did not explain why some Chiari patients developed syringomyelia and others did not. Only one study compared cine MR findings of 32 patients with syringomyelia and 15 patients without syringomyelia in Chiari type 1 malformation [154] and reported that the duration of the caudal CSF movement in the ventral subarachnoid space was significantly longer in syringomyelia.

Thus, the cine-mode MR imaging studies demonstrated abnormal CSF dynamics in Chiari type 1 malformation.

However, they failed to display definite evidences that CSF enters into the syrinx.

Pressure studies of syringomyelia

Direct recordings of the pressure in the syrinx were performed in four studies [34, 46, 76, 133]. In 1970, Ellertsson and Greitz first recorded pressures of the subarachnoid space and the syrinx using electromanometric equipment after percutaneous puncture in ten patients [46]. They described that the pressures in the syrinx were above those in the subarachnoid space in most cases, but the difference was not significant. Unfortunately, they did not specify the type of syringomyelia. Davis and Symon recorded the intrasyrinx pressure with a simple manometric technique during surgery in 17 syringomyelic patients including 5 Chiari malformations [34]. The recorded pressures were relatively low (4.0 to 7.0 cmH_2O in 15 patients and 0 to 1.0 cmH_2O in the other two patients) probably because their measurement was performed after draining of the subarachnoid CSF and syringomyelic fluid. Milhorat et al. performed manometric recordings of the intrasyrinx pressure in 32 patients including 21 Chiari type 1 patients during syrinx surgery [133]. They recorded the pressure through an 18-gage needle inserted into the syrinx after opening the dura and arachnoid. The recorded pressures ranged from 0.5 to 22.0 cmH_2O (mean, 7.7). They described that the patients with syrinx pressures greater than 7.7 cmH_2O tended to have more rapid progression of symptoms. Heiss et al. recorded the pressures of the cervical subarachnoid space and the syrinx through 22-gage spinal needles during surgery in 20 patients of syringomyelia with Chiari type 1 malformation [76]. They reported that the syrinx pressure (15±5.8 mmHg) was identical to the cervical subarachnoid pressure (15.1± 4.7 mmHg). Relatively larger values of the syrinx pressure in this study compared with the other two studies may be explained by preservation of the spinal subarachnoid space during recordings. They also reported that the CSF compliance (milliliters of CSF per milliliters of mercury) of the spinal subarachnoid space was significantly low in Chiari-syringomyelia patients than normal controls.

Several studies reported the relationship between the intracranial and spinal subarachnoid pressures in Chiari type 1 malformation. Williams reported the pressure dissociation between the intracranial and spinal subarachnoid spaces during Valsalva maneuver [201]. Häckel et al. reported that eight of nine patients with syrinx had a CSF block, while only three of 13 patients without syrinx showed a block by Valsalva maneuver of Queckenstedt test [73]. Using a manometric Queckenstedt test technique, Tachibana et al. demonstrated severe or complete CSF block with neck flexion and no CSF block with neck extension in the patients of syringomyelia with Chiari type 1 malformation [180]. According to the study by Heiss et al., the Valsalva maneuver during surgery failed to produce significant pressure differences between the intracranial and lumbar subarachnoid space in 20 Chiari patients with syringomyelia [76].

From these pressure studies, there is a variety of the degree of the CSF blockade in patients with Chiari type 1 malformation. The intrasyrinx pressure is almost identical to that of the surrounding subarachnoid space. It is unlikely that a simple pressure gradient is the main mechanism of syrinx formation.

Morphometric studies

Posterior fossa size

Morphometric studies on the posterior fossa and neural structures provided quantitative evidences on etiology of Chiari type 1 malformation. The posterior fossa volume was significantly reduced in the patients with Chiari type 1 malformation compared to normal controls [15, 134, 179, 187, 195]. There were some small differences in the results among the morphometric studies. Nishikawa et al. reported that there was no significant difference in the mean posterior crania fossa volume between Chiari type 1 patients and normal controls in adults [142]. However, the volume ratio of the neural structure (the brainstem and cerebellum) and the posterior cranial fossa was significantly larger in the Chiari patients. From the analysis of MRI in 42 pediatric patients with Chiari type 1 malformation, Sgouros et al. reported that there was no significant difference of the posterior fossa volume between the patients with Chiari malformation only and normal controls, but Chiari patients with syringomyelia had a significant smaller posterior fossa volume [172]. Studies measuring the parameters of the posterior fossa such as length of the supraocciput and clivus also showed small posterior fossa in Chiari type 1 malformation [14, 102, 144, 167]. These studies indicate that Chiari type 1 malformation is a disorder of paraxial mesoderm that induces underdevelopment of the occipital bone and overcrowding in the posterior fossa [134, 142]. However, the relationship between the presence of syringomyelia and size of the posterior fossa has not been clarified.

Tonsillar herniation

Chiari malformation has been defined as the descent of the cerebellar tonsil of 3 or 5 mm below the foramen magnum [1, 18]. Degree of tonsillar herniation was reported to be

associated with the severity of the brainstem or cerebellar compression symptoms [47, 207]. However, the literature indicated that tonsillar herniation of less than 3 or 5 mm can cause symptoms consistent with syringomyelia with Chiari type 1 malformation [53, 134, 170]. Even the patients without tonsillar herniation showed clinical presentation of syringomyelia with Chiari type 1 malformation [89, 109, 110, 189, 210] and were successfully treated by posterior fossa decompression.

It was also reported that the degree of tonsillar herniation did not correlate with presence of syringomyelia and size of the syrinx [125, 134, 175, 178, 207, 208]. Some studies demonstrated that intermediate level of tonsillar herniation was most frequently associated with syringomyelia. Stevens, et al. reported that syringomyelia was present in 57% of the patients showing the tonsillar descent at occiput-C1, 70% at C1–C2, and 20% at lower than C2 [175]. Stovner, et al. also reported that syringomyelia was significantly more associated with a herniation of 9 to 14 mm (56%) than smaller (13%) or larger (13%) herniations [178]. In a clinical study on surgical series of Chiari type 1 malformation by Yamazaki et al., the length of the ectopic tonsil was significantly larger in the patients without syringomyelia than those with syringomyelia [208].

According to these morphological studies, the role of mechanical effects of the displaced tonsil on the upper cervical cord may be limited.

Effects of surgical treatment

Posterior decompression

Gardner initially reported suboccipital craniectomy with opening of the fourth ventricle and plugging of the obex as a surgical treatment of syringomyelia associated with Chiari type 1 malformation [59]. The rationale of obex plugging was based on the idea that CSF entered into the central canal from the fourth ventricle. The Gardner's operation had been performed by many neurosurgeons [24, 25, 29, 43, 82, 116, 124, 153, 186]. However, simple decompressive procedures at the craniovertebral junction proved to have similar effects on reduction of syringomyelia with lower incidence of complications [56, 121]. Suboccipital craniectomy with laminectomy of the upper cervical spine and expansive duraplasty has been a standard surgery in this disorder [5, 7, 11, 12, 23, 36, 67, 81, 138, 165, 168, 188–192]. Several variations in procedures were reported. The arachnoid membrane was opened to explore the foramen magendie and excise adhesions [10, 31, 32, 39, 48, 63, 64, 71, 103, 107] or was left intact [37, 173, 199]. Some authors left the dura mater open with arachnoid dissection [22, 107] or intact [151]. Displaced tonsils were sometimes manipulated, coagulated or resected [4, 8, 9, 33, 54, 68, 72, 108, 112, 140, 205]. To prevent CSF-related complications, some authors did not open the dura, but removed the dural band (occipitoatlantal membrane) or outer layer of the dura [30, 55, 61, 75, 94, 98, 118, 136, 147, 148, 209, 211]. Meta-analysis of 582 pediatric patients in the literature revealed that foramen magnum decompression without duraplasty was associated with higher risk of reoperation but showed lower risk of complication compared to that with duraplasty [42]. There was no significant difference between these two methods in clinical improvement and reduction of syringomyelia after surgery. Several authors recommended suboccipital expansive craniotomy using autologous bone or synthetic materials to obtain dural expansion [84, 162, 163, 181, 193]. Too wide suboccipital craniectomy was also reported to produce downward displacement of the hindbrain [41, 84]. Thus, enlargement of the subarachnoid space around the hindbrain will be important to provide therapeutic effects. Recent variations in surgical procedures aimed to reduce complications or to achieve sufficient decompression.

Shunting procedures

Shunting procedures such as syringo-subarachnoid (S-S), syringo-peritoneal or syringo-pleural shunting are another option of surgical treatment. Syrinx shunting was developed as an additional procedure to foramen magnum decompression [4, 48, 52, 130, 160, 183, 197] or as a surgical treatment of syringomyelia without hindbrain abnormalities [17, 114, 122, 152, 182, 194]. Several authors reported that S-S shunting was effective in reduction of syrinx and improvement of syringomyeliac symptoms as the primary surgical treatment in syringomyelia with Chiari type 1 malformation [77–79, 92, 93, 96, 97, 149]. Although the syrinx shunting has shown higher incidence of reoperation [19, 171, 202], shunting procedures are the important option for syringomyelia of various etiologies including Chiari type 1 malformation. S-S shunting, which drains the syrinx fluid into the surrounding subarachnoid space, theoretically does not alter the CSF flow around the foramen magnum. The previous theories proposing CSF entrance from the subarachnoid space does not explain why S-S shunting works well as far as the shunt tube is patent.

Pre-syrinx state

In 1999, Fischbein et al. reported 5 patients showing enlarged spinal cord with parenchymal T1 and T2 prolongation but no cavitations on MR imaging, and called this condition as the presyrinx state [50]. Their series included one case of Chiari type 1 malformation. They proposed that the increased CSF

pressure by the pulsatile tonsillar descent drives CSF into the spinal cord parenchyma via perivascular spaces. The driven CSF will enlarge the central canal in syringomyelia. If the central canal is not patent, the driven CSF will distribute more diffusely in the spinal cord parenchyma and result in the presyrinx state. Several authors reported similar MR imaging features as the presyrinx state in Chiari malformations [65, 119], trauma [157, 169], arachnoiditis [87], hydrocephalus [137], or posterior fossa arachnoid cyst [143]. The MR appearance may be identical to that in posttraumatic microcystic degeneration [49, 113, 123] or adhesive spinal arachnoiditis [106]. Although the driven mechanism of CSF from the subarachnoid space into the central canal or the spinal cord parenchyma via perivascular spaces should be further verified, explanation for the extracellular fluid accumulation is plausible.

Recently, we investigated MR imaging findings of the spinal cord parenchyma in syringomyelia with Chiari type 1 malformation [2]. Parenchymal hyperintensity areas were present around the central canal and base of the posterior column adjacent to the syringomyelic cavity on T2-weighted images. This study indicates that the elevated extracellular fluid state is commonly present in the spinal cord in syringomyelia with Chiari type 1 malformation (Fig. 1). Such centrifugal pattern of the extracellular fluid accumulation is most likely produced by the disturbed absorption mechanisms of the extracellular fluid, not by the driven force of CSF from the spinal cord surface [2].

A new hypothesis for syrinx formation

The evidences of CSF dynamics, pressure studies, morphology of the hindbrain structures, effects of surgical intervention and recent MR imaging findings of "pre-syrinx state" promote new insights into the pathogenesis of syrinx in Chiari type 1 malformation.

Anatomical consideration

Human spinal cord has a characteristic vascular distribution over the cord surface. The outer layer of the pia mater covers the anterior spinal artery and vein at the anterior surface. There are no arachnoid trabeculae in the anterior subarachnoid space. In contrasts, the posterior subarachnoid space contains a longitudinal midline dorsal septum, which becomes only a few strands immediately below the foramen magnum [139]. The posterior spinal veins and arteries are situated in the true subarachnoid space with arachnoid trabeculations [184]. The posterior spinal veins receive

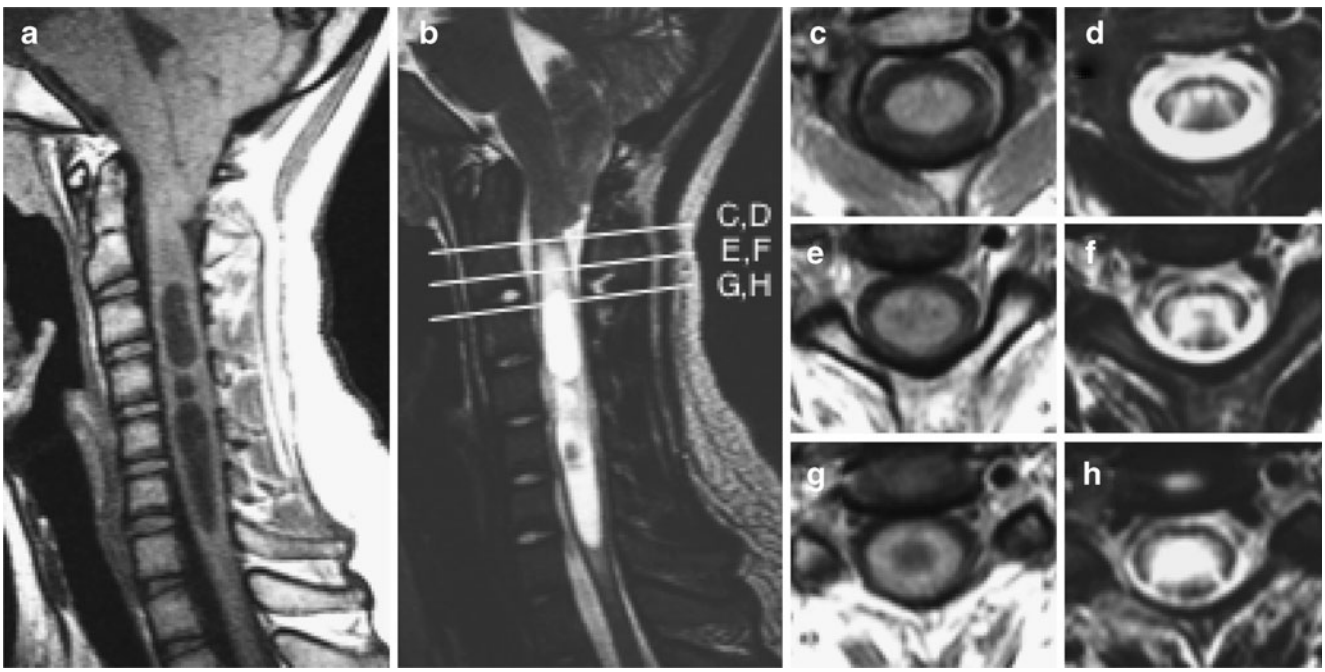

Fig. 1 MR imaging of the cervical spine in a 16-year-old girl showing syringomyelia with Chiari type 1 malformation. **a** T1-weighted sagittal image showing displaced tonsil and syringomyelia from the C2/3 to C6/7 levels. **b** T2-weighted sagittal image demonstrates intramedullary hyperintense areas at the C2 and C7 levels. The *three lines* indicate the levels of axial slices. **c**–**h** T1- (**c**, **e**, **g**) and T2-weighted (**d**, **f**, **h**) axial images at the upper (**c**, **d**), middle (**e**, **f**), and lower C2 (**g**, **h**) levels. T2-weighted images clearly demonstrate hyperintense areas at the central canal and the posterior gray matter (**d**, **f**), while T1-weighted images show only slightly hypointense signal (**c**, **e**). At the C2/3 level, T2-weighted image (**h**) demonstrates more extensive abnormal signal area in the spinal cord than T1-weighted image (**g**)

venous tributaries from the base of the posterior columns [62, 135] and constitute an important venous drainage of the spinal cord.

In the spinal cord, extracellular fluid is intimately associated with blood circulation. At the capillary level, fluid moves from the blood flow into the interstitial space at the arteriolar end of the capillary, where the filtration pressure exceeds the oncotic pressure, and from the interstitial space into the capillary at the venular end, where the oncotic pressure exceeds the filtration pressure [60]. It has been known that CSF is produced not only at the choroid plexus but also at the brain and spinal cord [129, 166]. Clinical and experimental studies using CTM and tracer techniques indicate that there is a significant fluid communication between the subarachnoid CSF and the extracellular space in the spinal cord. Considering these evidences, the extracellular fluid of the spinal cord contains both the filtrate from the spinal cord microvasculature and the CSF, and at least some part of the extracellular fluid will be absorbed into the intramedullary venous channels (Fig. 2-a).

Venous compliance and syrinx formation

The spinal CSF shows pulsatile movement with arterial pulsation. At the foramen magnum level, CSF enters into the spinal CSF space during systole and goes back to the intracranial space during diastole. The spinal CSF space will respond such CSF volume changes by altering the venous blood volume of the spinal cord and/or the epidural venous plexus. These venous blood volume changes during cardiac cycle may help to absorb blood from the capillary bed and the extracellular fluid from the spinal cord parenchyma.

There is evidence that compliance (the volume change per the pressure change) of the spinal CSF space is reduced in syringomyelia with Chiari type 1 malformation [76]. Reduced intracranial compliance determined from cine-mode MR imaging was also reported in Chiari type 1 malformation [3]. The low compliance of the CSF space is most likely produced by the tonsillar blockade of the posterior subarachnoid space at the foramen magnum. Because the posterior spinal veins exist in the true subaracnoid space, the spinal CSF pressure directly influences the posterior spinal veins and will reduce compliance of the posterior spinal veins. That is, the posterior spinal veins reduce the ability to expand during diastole of cardiac cycle and the absorption mechanism of the extracellular fluid from the spinal cord parenchyma will be most likely disturbed. The spinal cord blood flow may be preserved because of the preserved arteriovenous perfusion pressure. Thus, the reduced venous compliance results in decreased absorption of the extracellular fluid through the intramedullary venous channels. Because the central canal acts as the active transport of the fluid, the decreased venous absorption will produce enlargement of

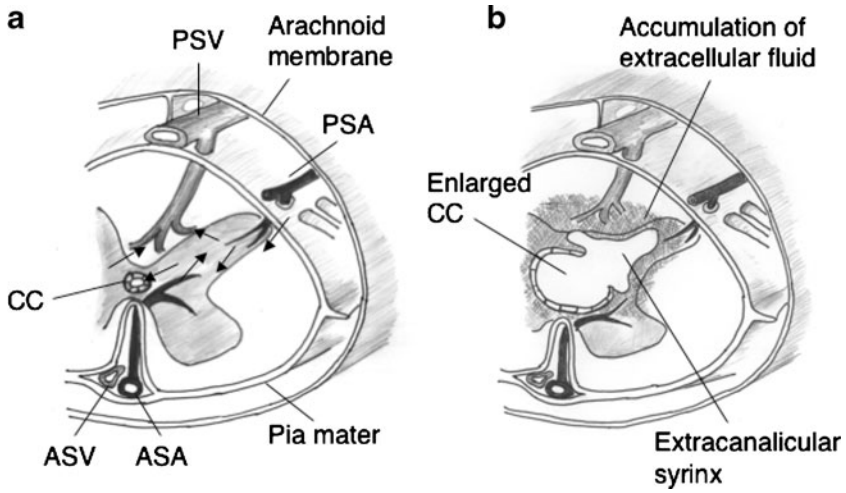

Fig. 2 Schematic presentations of the extracellular fluid circulation in the cervical spinal cord. *ASA* anterior spinal artery, *ASV* anterior spinal vein, *PSA* posterior spinal artery, *PSV* posterior spinal vein, *PCC* central canal. **a** Normal spinal cord. *Small arrows* indicate the flow of the extracellular fluid. The fluid in the extracellular space is derived from the filtrate of the arteriole end of the capillaries and the subarachnoid space via the perivascular spaces. The central canal acts as the active transport of the fluid. The extracellular fluid is absorbed through the intramedullary venous channels. The posterior spinal veins are situated in the posterior subarachnoid space and are directly influenced by the CSF pressure of the posterior subarachnoid space. **b** Syringomyelia associated with Chiari type 1 malformation. Reduced compliance of the posterior spinal veins due to the decreased compliance of the spinal subarachnoid space produces disturbed absorption of the extracellular fluid through the intramedullary venous channels. The accumulated extracellular fluid results in the enlarged central canal and the interstitial edema. Cleft formation of the expanded central canal and the accumulated extracellular fluid produce the extracanalicular syrinx

the central canal and increased extracellular fluid (interstitial edema) around the central canal (Fig. 2b). The extracellular fluid will be accumulated also in the relatively coarse areas such as the central gray matter and the posterior gray matter. Cleft formation initiated by rupture of the distended central canal may contribute to formation of the extracanalicular syrinx (Fig. 3).

Spinal dural arteriovenous fistula (AVF) also shows venous congestion and the spinal cord edema, but syringomyelia is uncommon. This should be noted. Spinal dural AVF produces significant decrease in spinal cord perfusion pressure. The abnormal perfusion state will result in both the extracellular fluid accumulation and intracellular edema caused by ischemia. Such ischemic edematous state will not result in syringomyelic cavity. Accumulation of the extracellular fluid with the preserved perfusion pressure may be important in expansion of the fluid pathways in the spinal cord.

Although most of our supposed mechanisms lack the experimental or clinical evidences and consist of speculations, this decreased absorption hypothesis can explain several radiological and clinical features. For example, delayed visualization of syringomyelia by CTM is the result of delayed clearance of the contrast via the intramedullary veins after influx of the subarachnoid contrast into the syrinx via the perivascular spaces. S-S shunting drains the syrinx fluid (the accumulated extracellular fluid) into the subarachnoid space where the usual CSF circulation and absorption mechanisms exist. It is still unclear why some patients with Chiari type 1 malformation develop syringomyelia and some do not. Differences in capacity of the venous absorption of the extracellular fluid or the fluid transport mechanism of the central canal may underlie such variation in clinical presentation of Chiari type 1 malformation.

Conclusions

This study critically reviews the evidences of the clinicopathological, radiological and clinical presentations of syringomyelia associated with Chiari type 1 malformation. The previous theories for the pathogenesis do not fully explain the radiological features and effects of surgical treatment such as shunting procedures. The MR appearance of syringomyelia demonstrates the extracellular fluid accumulation in the spinal cord parenchyma and suggests decreased absorption mechanisms of the extracellular fluid.

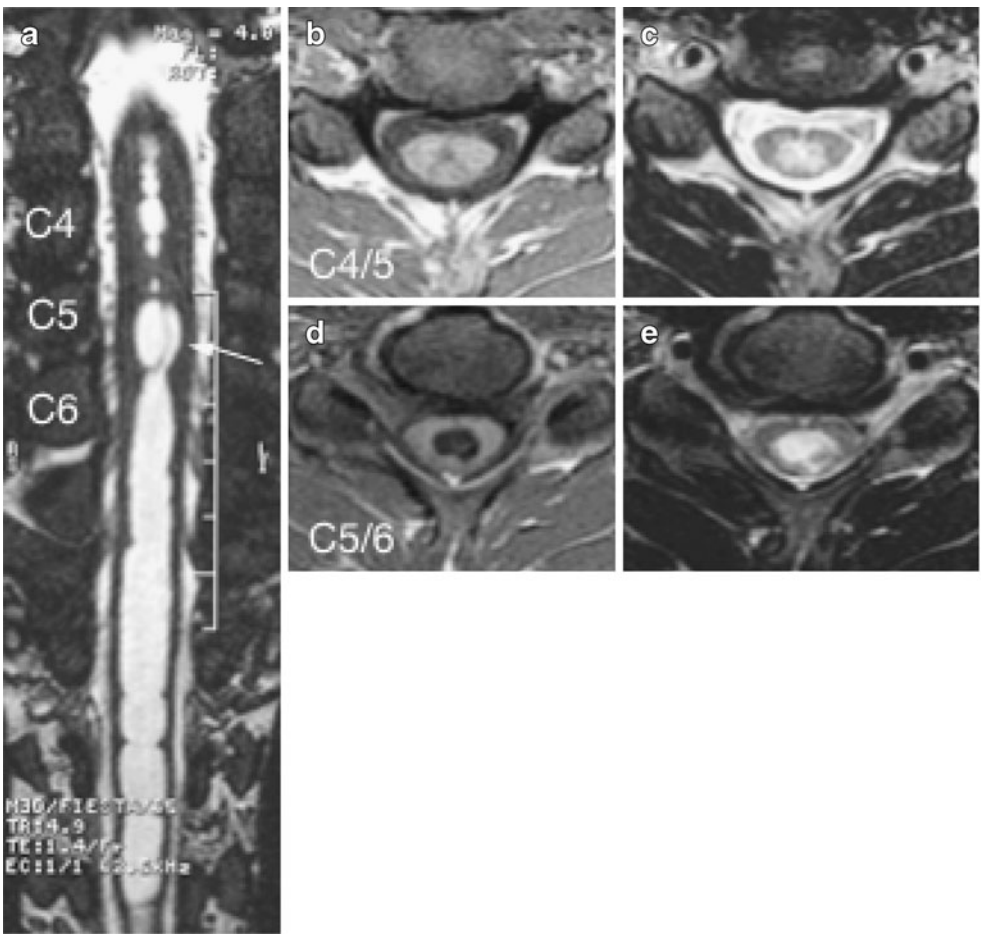

Fig. 3 MR imaging of the cervical spine in a 28-year-old woman showing syringomyelia with Chiari type 1 malformation. **a** Coronal image with fast imaging employing steady-state acquisition (FIESTA). This heavily T2-weighted image clearly demonstrates the enlarged central canal and the extracanalicular extension of the syrinx (*arrow*). **b–e** T1- (**b**, **d**) and T2-weighted (**c**, **e**) axial images at the C4/5 (**b**, **c**) and C5/6 (**d**, **e**) levels. Abnormal hyperintense areas around the central canal (**c**) or the syrinx (**e**) indicate accumulation of the extracellular fluid or interstitial edema

The review of the evidences promotes a new hypothesis of syrinx formation: Reduced compliance of the posterior spinal cord veins, that is associated with the decreased spinal CSF compliance due to the foramen magnum blockade, will produce disturbed absorption of the extracellular fluid through the intramedullary venous channels and result in syringomyelia in Chiari type 1 malformation.

References

1. Aboulezz AO, Sartor K, Geyer CA, Gado MH (1985) Position of cerebellar tonsils in the normal population and in patients with Chiari malformation: a quantitative approach with MR imaging. J Comput Assist Tomogr 9:1033–1036
2. Akiyama Y, Koyanagi I, Yoshifuji K, Murakami T, Baba T, Minamida Y, Nonaka T, Houkin K (2008) Interstitial spinal-cord oedema in syringomyelia associated with Chiari type 1 malformations. J Neurol Neurosurg Psychiatry 79:1153–1158
3. Alperin N, Sivaramakrishnan A, Lichtor T (2005) Magnetic resonance imaging-based measurements of cerebrospinal fluid and blood flow as indicators of intracranial compliance in patients with Chiari malformation. J Neurosurg 103:46–52
4. Alzate JC, Kothbauer KF, Jallo GI, Epstein FJ (2001) Treatment of Chiari I malformation in patients with and without syringomyelia: a consecutive series of 66 cases. Neurosurg Focus 11:E3
5. Anderson RC, Dowling KC, Feldstein NA, Emerson RG (2003) Chiari I malformation: potential role for intraoperative electrophysiologic monitoring. J Clin Neurophysiol 20:65–72
6. Armonda RA, Citrin CM, Foley KT, Ellenbogen RG (1994) Quantitative cine-mode magnetic resonance imaging of Chiari I malformations: an analysis of cerebrospinal fluid dynamics. Neurosurgery 35:214–223
7. Arora P, Behari S, Banerji D, Chhabra DK, Jain VK (2004) Factors influencing the outcome in symptomatic Chiari I malformation. Neurol India 52:470–474
8. Arruda JA, Costa CM, Tella OI Jr (2004) Results of the treatment of syringomyelia associated with Chiari malformation: analysis of 60 cases. Arq Neuropsiquiatr 62:237–244
9. Asgari S, Engelhorn T, Bschor M, Sandalcioglu IE, Stolke D (2003) Surgical prognosis in hindbrain related syringomyelia. Acta Neurol Scand 107:12–21
10. Attal N, Parker F, Tadi_ M, Aghakani N, Bouhassira D (2004) Effects of surgery on the sensory deficits of syringomyelia and predictors of outcome: a long term prospective study. J Neurol Neurosurg Psychiatry 75:1025–1030
11. Attenello FJ, McGirt MJ, Gathinji M, Datoo G, Atiba A, Weingart J, Carson B, Jallo GI (2008) Outcome of Chiari-associated syringomyelia after hindbrain decompression in children: analysis of 49 consecutive cases. Neurosurgery 62:1307–1313
12. Attenello FJ, McGirt MJ, Garcés-Ambrossi GL, Chaichana KL, Carson B, Jallo GI (2009) Suboccipital decompression for Chiari I malformation: outcome comparison of duraplasty with expanded polytetrafluoroethylene dural substitute versus pericranial autograft. Childs Nerv Syst 25:183–190
13. Aubin ML, Vignaud J, Jardin C, Bar D (1981) Computed tomography in 75 clinical cases of syringomyelia. AJNR 2:199–204
14. Aydin S, Hanimoglu H, Tanriverdi T, Yentur E, Kaynar MY (2005) Chiari type I malformations in adults: a morphometric analysis of the posterior cranial fossa. Surg Neurol 64:237–241
15. Badie B, Mendoza D, Batzdorf U (1995) Posterior fossa volume and response to suboccipital decompression in patients with Chiari I malformation. Neurosurgery 37:214–218
16. Ball MJ, Dayan AD (1972) Pathogenesis of syringomyelia. Lancet 2:799–801
17. Barbaro NM, Wilson CB, Gutin PH, Edwards MS (1984) Surgical treatment of syringomyelia. Favorable results with syringoperitoneal shunting. J Neurosurg 61:531–538
18. Barkovich AJ, Wippold FJ, Sherman JL, Citrin CM (1986) Significance of cerebellar tonsillar position on MR. AJNR 7:795–799
19. Batzdorf U, Klekamp J, Johnson JP (1998) A critical appraisal of syrinx cavity shunting procedures. J Neurosurg 89:382–388
20. Beuls EA, Vandersteen MA, Vanormelingen LM, Adriaensens PJ, Freling G, Herpers MJ, Gelan JM (1996) Deformation of the cervicomedullary junction and spinal cord in a surgically treated adult Chiari I hindbrain hernia associated with syringomyelia: a magnetic resonance microscopic and neuropathological study. Case report. J Neurosurg 85:701–708
21. Bhadelia RA, Bogdan AR, Wolpert SM, Lev S, Appignani BA, Heilman CB (1995) Cerebrospinal fluid flow waveforms: analysis in patients with Chiari I malformation by means of gated phase-contrast MR imaging velocity measurements. Radiology 196:195–202
22. Bhangoo R, Sgouros S (2006) Scoliosis in children with Chiari I-related syringomyelia. Childs Nerv Syst 22:1154–1157
23. Bidziński J (1988) Pathological findings in suboccipital decompression in 63 patients with syringomyelia. Acta Neurochir Suppl (Wien) 43:26–28
24. Blagodatsky MD, Larionov SN, Manohin PA, Shanturov VA, Gladyshev YuV (1993) Surgical treatment of "hindbrain related" syringomyelia: new data for pathogenesis. Acta Neurochir (Wien) 124:82–85
25. Blagodatsky MD, Larionov SN, Alexandrov YA, Velm AI (1999) Surgical treatment of Chiari I malformation with or without syringomyelia. Acta Neurochir (Wien) 141:963–968
26. Bonafé A, Manelfe C, Espagno J, Guiraud B, Rascol A (1980) Evaluation of syringomyelia with metrizamide computed tomographic myelography. J Comput Assist Tomogr 4:797–802
27. Bosley TM, Cohen DA, Schatz NJ, Zimmerman RA, Bilaniuk LT, Savino PJ, Sergott RS (1985) Comparison of metrizamide computed tomography and magnetic resonance imaging in the evaluation of lesions at the cervicomedullary junction. Neurology 35:485–492
28. Brugieres P, Idy-Peretti I, Iffenecker C, Parker F, Jolivet O, Hurth M, Gaston A, Bittoun J (2000) CSF flow measurement in syringomyelia. AJNR 21:1785–1792
29. Cahan LD, Bentson JR (1982) Considerations in the diagnosis and treatment of syringomyelia and the Chiari malformation. J Neurosurg 57:24–31
30. Caldarelli M, Novegno F, Vassimi L, Romani R, Tamburrini G, Di Rocco C (2007) The role of limited posterior fossa craniectomy in the surgical treatment of Chiari malformation Type I: experience with a pediatric series. J Neurosurg 106(3 Suppl):187–195
31. Calliauw L, Dehaene I (1977) The surgical risk in the treatment of Arnold Chiari malformations. Acta Neurochir (Wien) 39:173–179
32. Danish SF, Samdani A, Hanna A, Storm P, Sutton L (2006) Experience with acellular human dura and bovine collagen matrix for duraplasty after posterior fossa decompression for Chiari malformations. J Neurosurg 104(1 Suppl):16–20
33. da Silva JA, Holanda MM (2003) Basilar impression, Chiari malformation and syringomyelia: a retrospective study of 53 surgically treated patients. Arq Neuropsiquiatr 61:368–375
34. Davis CH, Symon L (1989) Mechanisms and treatment in post-traumatic syringomyelia. Br J Neurosurg 3:669–674
35. DeLaPaz RL, Brady TJ, Buonanno FS, New PF, Kistler JP, McGinnis BD, Pykett IL, Taveras JM (1983) Nuclear magnetic

resonance (NMR) imaging of Arnold-Chiari type I malformation with hydromyelia. J Comput Assist Tomogr 7:126–129
36. Depreitere B, Van Calenbergh F, van Loon J, Goffin J, Plets C (2000) Posterior fossa decompression in syringomyelia associated with a Chiari malformation: a retrospective analysis of 22 patients. Clin Neurol Neurosurg 102:91–96
37. Di Lorenzo N, Palma L, Palatinsky E, Fortuna A (1995) "Conservative" cranio-cervical decompression in the treatment of syringomyelia-Chiari I complex. A prospective study of 20 adult cases. Spine 20:2479–2483
38. Dolar MT, Haughton VM, Iskandar BJ, Quigley M (2004) Effect of craniocervical decompression on peak CSF velocities in symptomatic patients with Chiari I malformation. AJNR 25:142–145
39. Dones J, De Jesus O, Colen CB, Toledo MM, Delgado M (2003) Clinical outcomes in patients with Chiari I malformation: a review of 27 cases. Surg Neurol 60:142–148
40. Dubois PJ, Drayer BP, Sage M, Osborne D, Heinz ER (1981) Intramedullary penetrance of metrizamide in the dog spinal cord. AJNR 2:313–317
41. Duddy MJ, Williams B (1991) Hindbrain migration after decompression for hindbrain hernia: a quantitative assessment using MRI. Br J Neurosurg 5:141–152
42. Durham SR, Fjeld-Olenec K (2008) Comparison of posterior fossa decompression with and without duraplasty for the surgical treatment of Chiari malformation Type I in pediatric patients: a meta-analysis. J Neurosurg Pediatr 2:42–49
43. Dyste GN, Menezes AH, VanGilder JC (1989) Symptomatic Chiari malformations. An analysis of presentation, management, and long-term outcome. J Neurosurg 71:159–168
44. Ekholm SE, Foley M, Kido DK, Morris TW (1984) Lumbar myelography with metrizamide in rabbits. An investigation of contrast media penetration and resorption. Acta Radiol Diagn (Stockh) 25:517–522
45. Eldevik OP (1983) Elimination of metrizamide from the spinal subarachnoid space: a study of patients with abolished intracranial circulation. AJNR 4:585–587
46. Ellertsson AB, Greitz T (1970) The distending force in the production of communicating syringomyelia. Lancet 1 (7658):1234
47. Elster AD, Chen MY (1992) Chiari I malformations: clinical and radiologic reappraisal. Radiology 183:347–353
48. Ergün R, Akdemir G, Gezici AR, Tezel K, Beskonakli E, Erg_ng_r F, Taskin Y (2000) Surgical management of syringomyelia-Chiari complex. Eur Spine J 9:553–557
49. Falcone S, Quencer RM, Green BA, Patchen SJ, Post MJ (1994) Progressive posttraumatic myelomalacic myelopathy: imaging and clinical features. AJNR Am J Neuroradiol 15:747–754
50. Fischbein NJ, Dillon WP, Cobbs C, Weinstein PR (1999) The "presyrinx" state: a reversible myelopathic condition that may precede syringomyelia. AJNR Am J Neuroradiol 20:7–20
51. Forbes WS, Isherwood I (1978) Computed tomography in syringomyelia and the associated Arnold-Chiari type I malformation. Neuroradiology 27:73–78
52. Fujii K, Natori Y, Nakagaki H, Fukui M (1991) Management of syringomyelia associated with Chiari malformation: comparative study of syrinx size and symptoms by magnetic resonance imaging. Surg Neurol 36:281–285
53. Furuya K, Sano K, Segawa H, Ide K, Yoneyama H (1998) Symptomatic tonsillar ectopia. J Neurol Neurosurg Psychiatry 64:221–226
54. Galarza M, Sood S, Ham S (2007) Relevance of surgical strategies for the management of pediatric Chiari type I malformation. Childs Nerv Syst 23:691–696
55. Gambardella G, Caruso G, Caffo M, Germanò A, La Rosa G, Tomasello F (1998) Transverse microincisions of the outer layer of the dura mater combined with foramen magnum decompression as treatment for syringomyelia with Chiari I malformation. Acta Neurochir (Wien) 140:134–139
56. Garcìa-Uria J, Leunda G, Carrillo R, Bravo G (1981) Syringomyelia: long-term results after posterior fossa decompression. J Neurosurg 54:380–383
57. Gardner WJ, Goodall RJ (1950) The surgical treatment of Arnold-Chiari malformation in adults. An explanation of its mechanism and importance of encephalography in diagnosis. J Neurosurg 7:199–206
58. Gardner WJ, Ange J (1958) The mechanism of syringomyelia and its surgical correction. Clin Neurosurg 6:131–140
59. Gardner WJ (1965) Hydrodynamic mechanism of syringomyelia: its relationship to myelocele. J Neurol Neurosurg Psychiatry 28:247–259
60. Ganong WF (1985) Dynamics of blood flow and lymph flow. In: Review of Medical Physiology, 12th edn. Lange Medical Publications, Maruzen Co, Tokyo, pp 470–484
61. Genitori L, Peretta P, Nurisso C, Macinante L, Mussa F (2000) Chiari type I anomalies in children and adolescents: minimally invasive management in a series of 53 cases. Childs Nerv Syst 16:707–718
62. Gillilan LA (1970) Veins of the spinal cord. Anatomic details; suggested clinical applications. Neurology 20:860–868
63. Goel A, Bhatjiwale M, Desai K (1998) Basilar invagination: a study based on 190 surgically treated patients. J Neurosurg 88:962–968
64. Goel A, Desai K (2000) Surgery for syringomyelia: an analysis based on 163 surgical cases. Acta Neurochir (Wien) 142:293–302
65. Goh S, Bottrell CL, Aiken AH, Dillon WP, Wu YW (2008) Presyrinx in children with Chiari malformations. Neurology 71:351–356
66. Golman K, Wiik I, Salvesen S (1979) Absorption of a non-ionic contrast agent from cerebrospinal fluid to blood. Neuroradiology 18:227–233
67. Grabb PA, Mapstone TB, Oakes WJ (1999) Ventral brain stem compression in pediatric and young adult patients with Chiari I malformations. Neurosurgery 44:520–528
68. Greenlee JD, Donovan KA, Hasan DM, Menezes AH (2002) Chiari I malformation in the very young child: the spectrum of presentations and experience in 31 children under age 6 years. Pediatrics 110:1212–1219
69. Greitz D, Ericson K, Flodmark O (1999) Pathogenesis and mechanics of spinal cysts. A new hypothesis based on magnetic resonance stdies of cerebrospinal fluid dynamics. Int J Neuroradiol 5:61–78
70. Greitz D (2006) Unraveling the riddle of syringomyelia. Neurosurg Rev 29:251–264
71. Guo F, Wang M, Long J, Wang H, Sun H, Yang B, Song L (2007) Surgical management of Chiari malformation: analysis of 128 cases. Pediatr Neurosurg 43:375–381
72. Guyotat J, Bret P, Jouanneau E, Ricci AC, Lapras C (1998) Syringomyelia associated with type I Chiari malformation. A 21-year retrospective study on 75 cases treated by foramen magnum decompression with a special emphasis on the value of tonsils resection. Acta Neurochir (Wien) 140:745–754
73. Häckel M, Benes V, Mohapl M (2001) Simultaneous cerebral and spinal fluid pressure recordings in surgical indications of the Chiari malformation without myelodysplasia. Acta Neurochir (Wien) 143:909–918
74. Haughton VM, Korosec FR, Medow JE, Dolar MT, Iskandar BJ (2003) Peak systolic and diastolic CSF velocity in the foramen magnum in adult patients with Chiari I malformations and in normal control participants. AJNR 24:169–176
75. Hayhurst C, Richards O, Zaki H, Findlay G, Pigott TJ (2008) Hindbrain decompression for Chiari-syringomyelia complex: an outcome analysis comparing surgical techniques. Br J Neurosurg 22:86–91

76. Heiss JD, Patronas N, DeVroom HL, Shawker T, Ennis R, Kammerer W, Eidsath A, Talbot T, Morris J, Eskioglu E, Oldfield EH (1999) Elucidating the pathophysiology of syringomyelia. J Neurosurg 91:553–562
77. Hida K, Iwasaki Y, Koyanagi I, Sawamura Y, Abe H (1995) Surgical indication and results of foramen magnum decompression versus syringosubarachnoid shunting for syringomyelia associated with Chiari I malformation. Neurosurgery 37:673–679
78. Hida K, Iwasaki Y, Koyanagi I, Abe H (1999) Pediatric syringomyelia with chiari malformation: its clinical characteristics and surgical outcomes. Surg Neurol 51:383–391
79. Hida K, Iwasaki Y (2001) Syringosubarachnoid shunt for syringomyelia associated with Chiari I malformation. Neurosurg Focus 11:E7
80. Hinokuma K, Ohama E, Oyanagi K, Kakita A, Kawai K, Ikuta F (1992) Syringomyelia. A neuropathological study of 18 autopsy cases. Acta Pathol Jpn 42:25–34
81. Hoffman CE, Souweidane MM (2008) Cerebrospinal fluid-related complications with autologous duraplasty and arachnoid sparing in type I Chiari malformation. Neurosurgery 62(3 Suppl 1):156–161
82. Hoffman HJ, Neill J, Crone KR, Hendrick EB, Humphreys RP (1987) Hydrosyringomyelia and its management in childhood. Neurosurgery 21:347–351
83. Hofmann E, Warmuth-Metz M, Bendszus M, Solymosi L (2000) Phase-contrast MR imaging of the cervical CSF and spinal cord: volumetric motion analysis in patients with Chiari I malformation. AJNR 21:151–158
84. Holly LT, Batzdorf U (2001) Management of cerebellar ptosis following craniovertebral decompression for Chiari I malformation. J Neurosurg 94:21–26
85. Holtas S, Morris TW, Ekholm SE, Isaac L, Fonte D (1986) Penetration of subarachnoid contrast medium into rabbit spinal cord. Comparison between metrizamide and iohexol. Invest Radiol 21:151–155
86. Ikata T, Masaki K, Kashiwaguchi S (1988) Clinical and experimental studies on permeability of tracers in normal spinal cord and syringomyelia. Spine 13:737–741
87. Ikushima I, Korogi Y, Hirai T, Yamashita Y (2007) High-resolution constructive interference in a steady state imaging of cervicothoracic adhesive arachnoiditis. J Comput Assist Tomogr 31:143–147
88. Isherwood I, Fawcitt RA, St Clair Forbes W, Nettle JR, Pullan BR (1977) Computer tomography of the spinal canal using metrizamide. Acta Radiol Suppl 355:299–305
89. Iskandar BJ, Hedlund GL, Grabb PA, Oakes WJ (1998) The resolution of syringohydromyelia without hindbrain herniation after posterior fossa decompression. J Neurosurg 89:212–216
90. Iskandar BJ, Quigley M, Haughton VM (2004) Foramen magnum cerebrospinal fluid flow characteristics in children with Chiari I malformation before and after craniocervical decompression. J Neurosurg 101(2 Suppl):169–178
91. Isu T, Iwasaki Y, Sasaki H, Abe H, Tashiro K, Nakamura N (1987) An autopsy case of syringomyelia associated with Chiari malformation and basilar impression [in Japanese]. No Shinkei Geka 15:671–675
92. Isu T, Iwasaki Y, Akino M, Abe H (1990) Syringo-subarachnoid shunt for syringomyelia associated with Chiari malformation (type 1). Acta Neurochir (Wien) 107:152–160
93. Isu T, Iwasaki Y, Akino M, Abe H (1990) Hydrosyringomyelia associated with a Chiari I malformation in children and adolescents. Neurosurgery 26:591–597
94. Isu T, Sasaki H, Takamura H, Kobayashi N (1993) Foramen magnum decompression with removal of the outer layer of the dura as treatment for syringomyelia occurring with Chiari I malformation. Neurosurgery 33:844–850
95. Iwasaki Y, Abe H, Isu T, Miyasaka K (1985) CT myelography with intramedullary enhancement in cervical spondylosis. J Neurosurg 63:363–366
96. Iwasaki Y, Koyanagi I, Hida K, Abe H (1999) Syringo-subarachnoid shunt for syringomyelia using partial hemilaminectomy. Br J Neurosurg 13:41–45
97. Iwasaki Y, Hida K, Koyanagi I, Abe H (2000) Reevaluation of syringosubarachnoid shunt for syringomyelia with Chiari malformation. Neurosurgery 46:407–413
98. James HE, Brant A (2002) Treatment of the Chiari malformation with bone decompression without durotomy in children and young adults. Childs Nerv Syst 18:202–206
99. Jinkins JR, Bashir R, Al-Mefty O, Al-Kawi MZ, Fox JL (1986) Cystic necrosis of the spinal cord in compressive cervical myelopathy: demonstration by iopamidol CT-myelography. AJR 147:767–775
100. Kan S, Fox AJ, Vinuela F, Debrun G (1983) Spinal cord size in syringomyelia: change with position on metrizamide myelography. Radiology 146:409–414
101. Kan S, Fox AJ, Vinuela F (1985) Delayed metrizamide CT enhancement of syringomyelia: postoperative observations. AJNR 6:613–616
102. Karagoz F, Izgi N, Kapijcijoglu Sencer S (2002) Morphometric measurements of the cranium in patients with Chiari type I malformation and comparison with the normal population. Acta Neurochir (Wien) 144:165–171
103. Klekamp J, Batzdorf U, Samii M, Bothe HW (1996) The surgical treatment of Chiari I malformation. Acta Neurochir (Wien) 138:788–801
104. Klekamp J (2002) The pathophysiology of syringomyelia—historical overview and current concept. Acta Neurochir (Wien) 144:649–664
105. Koc K, Anik Y, Anik I, Cabuk B, Ceylan S (2007) Chiari 1 malformation with syringomyelia: correlation of phase-contrast cine MR imaging and outcome. Turk Neurosurg 17:183–192
106. Koyanagi I, Iwasaki Y, Hida K, Houkin K (2005) Clinical features and pathomechanisms of syringomyelia associated with spinal arachnoiditis. Surg Neurol 63:350–355
107. Krieger MD, McComb JG, Levy ML (1999) Toward a simpler surgical management of Chiari I malformation in a pediatric population. Pediatr Neurosurg 30:113–121
108. Kumar R, Kalra SK, Vaid VK, Mahapatra AK (2008) Chiari I malformation: surgical experience over a decade of management. Br J Neurosurg 22:409–414
109. Kyoshima K, Kuroyanagi T, Oya F, Kamijo Y, El-Noamany H, Kobayashi S (2002) Syringomyelia without hindbrain herniation: tight cisterna magna. Report of four cases and a review of the literature. J Neurosurg 96(2 Suppl):239–249
110. Kyoshima K, Kuroyanagi T, Toriyama T, Takizawa T, Hirooka Y, Miyama H, Tanabe A, Oikawa S (2004) Surgical experience of syringomyelia with reference to the findings of magnetic resonance imaging. J Clin Neurosci 11:273–279
111. LaMasters DL, Watanabe TJ, Chambers EF, Norman D, Newton TH (1982) Multiplanar metrizamide-enhanced CT imaging of the foramen magnum. AJNR 3:485–494
112. Lazareff JA, Galarza M, Gravori T, Spinks TJ (2002) Tonsillectomy without craniectomy for the management of infantile Chiari I malformation. J Neurosurg 97:1018–1022
113. Lee TT, Arias JM, Andrus HL, Quencer RM, Falcone SF, Green BA (1997) Progressive posttraumatic myelomalacic myelopathy: treatment with untethering and expansive duraplasty. J Neurosurg 86:624–628
114. Lesoin F, Petit H, Thomas CE 3rd, Viaud C, Baleriaux D, Jomin M (1986) Use of the syringoperitoneal shunt in the treatment of syringomyelia. Surg Neurol 25:131–136

115. Levine DN (2004) The pathogenesis of syringomyelia associated with lesions at the foramen magnum: a critical review of existing theories and proposal of a new hypothesis. J Neurol Sci 220:3–21
116. Levy WJ, Mason L, Hahn JF (1983) Chiari malformation presenting in adults: a surgical experience in 127 cases. Neurosurgery 12:377–390
117. Li KC, Chui MC (1987) Conventional and CT metrizamide myelography in Arnold-Chiari I malformation and syringomyelia. AJNR 8:11–17
118. Limonadi FM, Selden NR (2004) Dura-splitting decompression of the craniocervical junction: reduced operative time, hospital stay, and cost with equivalent early outcome. J Neurosurg 101(2 Suppl):184–188
119. Lipson AC, Ellenbogen RG, Avellino AM (2008) Radiographic formation and progression of cervical syringomyelia in a child with untreated Chiari I malformation. Pediatr Neurosurg 44:221–223
120. Liu B, Wang ZY, Xie JC, Han HB, Pei XL (2007) Cerebrospinal fluid dynamics in Chiari malformation associated with syringomyelia. Chin Med J (Engl) 120:219–223
121. Logue V, Edwards MR (1981) Syringomyelia and its surgical treatment—an analysis of 75 patients. J Neurol Neurosurg Psychiatry 44:273–284
122. Lund-Johansen M, Wester K (1997) Syringomyelia treated with a nonvalved syringoperitoneal shunt: a follow-up study. Neurosurgery 41:858–865
123. MacDonald RL, Findlay JM, Tator CH (1988) Microcystic spinal cord degeneration causing posttraumatic myelopathy. Report of two cases. J Neurosurg 68:466–471
124. Mariani C, Cislaghi MG, Barbieri S, Filizzolo F, Di Palma F, Farina E, D'Aliberti G, Scarlato G (1991) The natural history and results of surgery in 50 cases of syringomyelia. J Neurol 238:433–438
125. Masur H, Oberwittler C, Reuther G, Heyen P (1995) Cerebellar herniation in syringomyelia: relation between tonsillar herniation and the dimensions of the syrinx and the remaining spinal cord. A quantitative MRI study. Eur Neurol 35:162–167
126. McGirt MJ, Nimjee SM, Floyd J, Bulsara KR, George TM (2005) Correlation of cerebrospinal fluid flow dynamics and headache in Chiari I malformation. Neurosurgery 56:716–721
127. McGirt MJ, Nimjee SM, Fuchs HE, George TM (2006) Relationship of cine phase-contrast magnetic resonance imaging with outcome after decompression for Chiari I malformations. Neurosurgery 59:140–146
128. McGirt MJ, Atiba A, Attenello FJ, Wasserman BA, Datoo G, Gathinji M, Carson B, Weingart JD, Jallo GI (2008) Correlation of hindbrain CSF flow and outcome after surgical decompression for Chiari I malformation. Childs Nerv Syst 24:833–840
129. Milhorat TH, Hammock MK, Fenstermacher JD, Rall DP, Levin VA (1971) Cerebrospinal fluid production by the choroids plexus and brain. Science 173:330–332
130. Milhorat TH, Johnson WD, Miller JI, Bergland RM, Hollenberg-Sher J (1992) Surgical treatment of syringomyelia based on magnetic resonance imaging criteria. Neurosurgery 31:231–245
131. Milhorat TH, Miller JI, Johnson WD, Adler DE, Heger IM (1993) Anatomical basis of syringomyelia occurring with hindbrain lesions. Neurosurgery 32:748–754
132. Milhorat TH, Capocelli AL Jr, Anzil AP, Kotzen RM, Milhorat RH (1995) Pathological basis of spinal cord cavitation in syringomyelia: analysis of 105 autopsy cases. J Neurosurg 82:802–812
133. Milhorat TH, Capocelli AL Jr, Kotzen RM, Bolognese P, Heger IM, Cottrell JE (1997) Intramedullary pressure in syringomyelia: clinical and pathophysiological correlates of syrinx distension. Neurosurgery 41:1102–1110
134. Milhorat TH, Chou MW, Trinidad EM, Kula RW, Mandell M, Wolpert C, Speer MC (1999) Chiari I malformation redefined: clinical and radiographic findings for 364 symptomatic patients. Neurosurgery 44:1005–1017
135. Miyasaka K, Asano T, Ushikoshi S, Hida K, Koyanagi I (2000) Vascular anatomy of the spinal cord and classification of spinal arteriovenous malformations. Interv Neuroradiol 6(Suppl 1):195–198
136. Munshi I, Frim D, Stine-Reyes R, Weir BK, Hekmatpanah J, Brown F (2000) Effects of posterior fossa decompression with and without duraplasty on Chiari malformation-associated hydromyelia. Neurosurgery 46:1384–1390
137. Muthukumar N, Venkatesh G, Thiruppathy S (2005) Arrested hydrocephalus and the presyrinx state. Case report. J Neurosurg 103(5 Suppl):466–470
138. Nagib MG (1994) An approach to symptomatic children (ages 4–14 years) with Chiari type I malformation. Pediatr Neurosurg 21:31–35
139. Nauta HJ, Dolan E, Yasargil MG (1983) Microsurgical anatomy of spinal subarachnoid space. Surg Neurol 19:431–437
140. Navarro R, Olavarria G, Seshadri R, Gonzales-Portillo G, McLone DG, Tomita T (2004) Surgical results of posterior fossa decompression for patients with Chiari I malformation. Childs Nerv Syst 20:349–356
141. Netsky MG (1953) Syringomyelia; a clinicopathologic study. AMA Arch Neurol Psychiatry 70:741–777
142. Nishikawa M, Sakamoto H, Hakuba A, Nakanishi N, Inoue Y (1997) Pathogenesis of Chiari malformation: a morphometric study of the posterior cranial fossa. J Neurosurg 86:40–47
143. Nomura S, Akimura T, Imoto H, Nishizaki T, Suzuki M (2002) Endoscopic fenestration of posterior fossa arachnoid cyst for the treatment of presyrinx myelopathy–case report. Neurol Med Chir (Tokyo) 42:452–454
144. Nyland H, Krogness KG (1978) Size of posterior fossa in Chiari type 1 malformation in adults. Acta Neurochir (Wien) 40:233–242
145. Oldfield EH, Muraszko K, Shawker TH, Patronas NJ (1994) Pathophysiology of syringomyelia associated with Chiari I malformation of the cerebellar tonsils. Implications for diagnosis and treatment. J Neurosurg 80:3–15
146. Olsson B, Eldevik OP, Gronnerod TA (1985) Absorption of iohexol from cerebrospinal fluid to blood: pharmacokinetics in humans. Neuroradiology 27:172–175
147. Ono A, Suetsuna F, Ueyama K, Yokoyama T, Aburakawa S, Numasawa T, Wada K, Toh S (2007) Surgical outcomes in adult patients with syringomyelia associated with Chiari malformation type I: the relationship between scoliosis and neurological findings. J Neurosurg Spine 6:216–221
148. Ono A, Suetsuna F, Ueyama K, Yokoyama T, Aburakawa S, Takeuchi K, Numasawa T, Wada K, Toh S (2007) Cervical spinal motion before and after surgery in patients with Chiari malformation type I associated with syringomyelia. J Neurosurg Spine 7:473–477
149. Padovani R, Cavallo M, Gaist G (1989) Surgical treatment of syringomyelia: favorable results with syringosubarachnoid shunting. Surg Neurol 32:173–180
150. Panigrahi M, Reddy BP, Reddy AK, Reddy JJ (2004) CSF flow study in Chiari I malformation. Childs Nerv Syst 20:336–340
151. Perrini P, Benedetto N, Tenenbaum R, Di Lorenzo N (2007) Extra-arachnoidal cranio-cervical decompression for syringomyelia associated with Chiari I malformation in adults: technique assessment. Acta Neurochir (Wien) 149:1015–1023
152. Phillips TW, Kindt GW (1981) Syringoperitoneal shunt for syringomyelia: a preliminary report. Surg Neurol 16:462–466
153. Pillay PK, Awad IA, Little JR, Hahn JF (1991) Symptomatic Chiari malformation in adults: a new classification based on

magnetic resonance imaging with clinical and prognostic significance. Neurosurgery 28:639–645
154. Pinna G, Alessandrini F, Alfieri A, Rossi M, Bricolo A (2000) Cerebrospinal fluid flow dynamics study in Chiari I malformation: implications for syrinx formation. Neurosurg Focus 8:E3
155. Pujol J, Roig C, Capdevila A, Pou A, Marti-Vilalta JL, Kulisevsky J, Escartin A, Zannoli G (1995) Motion of the cerebellar tonsils in Chiari type I malformation studied by cine phase-contrast MRI. Neurology 45:1746–1753
156. Quigley MF, Iskandar B, Quigley ME, Nicosia M, Haughton V (2004) Cerebrospinal fluid flow in foramen magnum: temporal and spatial patterns at MR imaging in volunteers and in patients with Chiari I malformation. Radiology 232:229–236
157. Reed CM, Campbell SE, Beall DP, Bui JS, Stefko RM (2005) Atlanto-occipital dislocation with traumatic pseudomeningocele formation and post-traumatic syringomyelia. Spine 30:E128–E133
158. Rennels ML, Gregory TF, Blaumanis OR, Fujimoto K, Grady PA (1985) Evidence for a 'paravascular' fluid circulation in the mammalian central nervous system, provided by the rapid distribution of tracer protein throughout the brain from the subarachnoid space. Brain Res 326:47–63
159. Resjö IM, Harwood-Nash DC, Fitz CR, Chuang S (1979) Computed tomographic metrizamide myelography in syringohydromyelia. Radiology 131:405–407
160. Rhoton AL Jr (1976) Microsurgery of Arnold-Chiari malformation in adults with and without hydromyelia. J Neurosurg 45:473–483
161. Sage MR, Wilcox J, Evill CA, Benness GT (1982) Brain parenchyma penetration by intrathecal ionic and nonionic contrast media. AJNR 3:481–483
162. Sahuquillo J, Rubio E, Poca MA, Rovira A, Rodriguez-Baeza A, Cervera C (1994) Posterior fossa reconstruction: a surgical technique for the treatment of Chiari I malformation and Chiari I/syringomyelia complex—preliminary results and magnetic resonance imaging quantitative assessment of hindbrain migration. Neurosurgery 35:874–885
163. Sakamoto H, Nishikawa M, Hakuba A, Yasui T, Kitano S, Nakanishi N, Inoue Y (1999) Expansive suboccipital cranioplasty for the treatment of syringomyelia associated with Chiari malformation. Acta Neurochir (Wien) 141:949–961
164. Sakas DE, Korfias SI, Wayte SC, Beale DJ, Papapetrou KP, Stranjalis GS, Whittaker KW, Whitwell HL (2005) Chiari malformation: CSF flow dynamics in the craniocervical junction and syrinx. Acta Neurochir (Wien) 147:1223–1233
165. Sansur CA, Heiss JD, DeVroom HL, Eskioglu E, Ennis R, Oldfield EH (2003) Pathophysiology of headache associated with cough in patients with Chiari I malformation. J Neurosurg 98:453–458
166. Sato O, Asai T, Amano Y, Hara M, Tsugane R, Yagi M (1971) Formation of cerebrospinal fluid in spinal subarachnoid space. Nature 233:129–130
167. Schady W, Metcalfe RA, Butler P (1987) The incidence of craniocervical bony anomalies in the adult Chiari malformation. J Neurol Sci 82:193–203
168. Schlesinger EB, Antunes JL, Michelsen WJ, Louis KM (1981) Hydromyelia: clinical presentation and comparison of modalities of treatment. Neurosurgery 9:356–365
169. Scholsem M, Scholtes F, Belachew S, Martin D (2008) Acquired tonsillar herniation and syringomyelia after pleural effusion aspiration: case report. Neurosurgery 62:E1172–E1173
170. Sekula RF Jr, Jannetta PJ, Casey KF, Marchan EM, Sekula LK, McCrady CS (2005) Dimensions of the posterior fossa in patients symptomatic for Chiari I malformation but without cerebellar tonsillar descent. Cerebrospinal Fluid Res 2:11
171. Sgouros S, Williams B (1995) A critical appraisal of drainage in syringomyelia. J Neurosurg 82:1–10
172. Sgouros S, Kountouri M, Natarajan K (2007) Skull base growth in children with Chiari malformation Type I. J Neurosurg 107(3 Suppl):188–192
173. Sindou M, Chavez-Machuca J, Hashish H (2002) Craniocervical decompression for Chiari type I-malformation, adding extreme lateral foramen magnum opening and expansile duroplasty with arachnoid preservation. Technique and long-term functional results in 44 consecutive adult cases—comparison with literature dat. Acta Neurochir (Wien) 144:1005–1019
174. Sivaramakrishnan A, Alperin N, Surapaneni S, Lichtor T (2004) Evaluating the effect of decompression surgery on cerebrospinal fluid flow and intracranial compliance in patients with chiari malformation with magnetic resonance imaging flow studies. Neurosurgery 55:1344–1351
175. Stevens JM, Serva WA, Kendall BE, Valentine AR, Ponsford JR (1993) Chiari malformation in adults: relation of morphological aspects to clinical features and operative outcome. J Neurol Neurosurg Psychiatry 56:1072–1077
176. Stoodley MA, Jones NR, Brown CJ (1996) Evidence for rapid fluid flow from the subarachnoid space into the spinal cord central canal in the rat. Brain Res 707:155–164
177. Stoodley MA, Brown SA, Brown CJ, Jones NR (1997) Arterial pulsation-dependent perivascular cerebrospinal fluid flow into the central canal in the sheep spinal cord. J Neurosurg 86:686–693
178. Stovner LJ, Rinck P (1992) Syringomyelia in Chiari malformation: relation to extent of cerebellar tissue herniation. Neurosurgery 31:913–917
179. Stovner LJ, Bergan U, Nilsen G, Sjaastad O (1993) Posterior cranial fossa dimensions in the Chiari I malformation: relation to pathogenesis and clinical presentation. Neuroradiology 35:113–118
180. Tachibana S, Iida H, Yada K (1992) Significance of positive Queckenstedt test in patients with syringomyelia associated with Arnold-Chiari malformations. J Neurosurg 76:67–71
181. Takayasu M, Takagi T, Hara M, Anzai M (2004) A simple technique for expansive suboccipital cranioplasty following foramen magnum decompression for the treatment of syringomyelia associated with Chiari I malformation. Neurosurg Rev 27:173–177
182. Tator CH, Meguro K, Rowed DW (1982) Favorable results with syringosubarachnoid shunts for treatment of syringomyelia. J Neurosurg 56:517–523
183. Tator CH, Briceno C (1988) Treatment of syringomyelia with a syringosubarachnoid shunt. Can J Neurol Sci 15:48–57
184. Tator CH, Koyanagi I (1997) Vascular mechanisms in the pathophysiology of human spinal cord injury. J Neurosurg 86:483–492
185. Terae S, Miyasaka K, Abe S, Abe H, Tashiro K (1994) Increased pulsatile movement of the hindbrain in syringomyelia associated with the Chiari malformation: cine-MRI with presaturation bolus tracking. Neuroradiology 36:125–129
186. Tokuno H, Hakuba A, Suzuki T, Nishimura S (1988) Operative treatment of Chiari malformation with syringomyelia. Acta Neurochir Suppl (Wien) 43:22–25
187. Trigylidas T, Baronia B, Vassilyadi M, Ventureyra EC (2008) Posterior fossa dimension and volume estimates in pediatri.c patients with Chiari I malformations. Childs Nerv Syst 24:329–336
188. Tubbs RS, Elton S, Grabb P, Dockery SE, Bartolucci AA, Oakes WJ (2001) Analysis of the posterior fossa in children with the Chiari 0 malformation. Neurosurgery 48:1050–1055
189. Tubbs RS, McGirt MJ, Oakes WJ (2003) Surgical experience in 130 pediatric patients with Chiari I malformations. J Neurosurg 99:291–296

190. Tubbs RS, Webb DB, Oakes WJ (2004) Persistent syringomyelia following pediatric Chiari I decompression: radiological and surgical findings. J Neurosurg 100(5 Suppl Pediatrics):460–464
191. Tubbs RS, Iskandar BJ, Bartolucci AA, Oakes WJ (2004) A critical analysis of the Chiari 1.5 malformation. J Neurosurg 101 (2 Suppl):179–183
192. Ur-Rahman N, Jamjoom ZA (1991) Surgical management of Chiari malformation and syringomyelia: experience in 14 cases. Ann Saudi Med 11:402–410
193. Vanaclocha V, Saiz-Sapena N, Garcia-Casasola MC (1997) Surgical technique for cranio-cervical decompression in syringomyelia associated with Chiari type I malformation. Acta Neurochir (Wien) 139:529–540
194. Vaquero J, Mart_nez R, Salazar J, Santos H (1987) Syringosubarachnoid shunt for treatment of syringomyelia. Acta Neurochir (Wien) 84:105–109
195. Vega A, Quintana F, Berciano J (1990) Basichondrocranium anomalies in adult Chiari type I malformation: a morphometric study. J Neurol Sci 99:137–145
196. Ventureyra EC, Aziz HA, Vassilyadi M (2003) The role of cine flow MRI in children with Chiari I malformation. Childs Nerv Syst 19:109–113
197. Vernet O, Farmer JP, Montes JL (1996) Comparison of syringopleural and syringosubarachnoid shunting in the treatment of syringomyelia in children. J Neurosurg 84:624–628
198. Wang AM, Jolesz F, Rumbaugh CL, Zamani A (1983) CT assessment of thoracic extension and of concomitant lesions in syringohydromyelia. J Comput Assist Tomogr 7:18–24
199. Wetjen NM, Heiss JD, Oldfield EH (2008) Time course of syringomyelia resolution following decompression of Chiari malformation Type I. J Neurosurg Pediatr 1:118–123
200. Williams B (1969) The distending force in the producing of "communicating syringomyelia". Lancet 26:189–193
201. Williams B (1980) On the pathogenesis of syringomyelia: a review. J R Soc Med 73:798–806
202. Williams B, Sgouros S, Nenji E (1995) Cerebrospinal fluid drainage for syringomyelia. Eur J Pediatr Surg 5(Suppl 1):27–30
203. Winkler SS, Sackett JF (1980) Explanation of metrizamide brain penetration: a review. J Comput Assist Tomogr 4:191–193
204. Wolpert SM, Bhadelia RA, Bogdan AR, Cohen AR (1994) Chiari I malformations: assessment with phase-contrast velocity MR. AJNR 15:1299–1308
205. Won DJ, Nambiar U, Muszynski CA, Epstein FJ (1997) Coagulation of herniated cerebellar tonsils for cerebrospinal fluid pathway restoration. Pediatr Neurosurg 27:272–275
206. Woosley RE, Whaley RA (1982) Use of metrizamide in computerized tomography to diagnose the Chiari I malformation. J Neurosurg 56:373–376
207. Wu YW, Chin CT, Chan KM, Barkovich AJ, Ferriero DM (1999) Pediatric Chiari I malformations: do clinical and radiologic features correlate? Neurology 53:1271–1276
208. Yamazaki Y, Tachibana S, Takano M, Fujii K (1998) Clinical and neuroimaging features of Chiari type I malformations with and without associated syringomyelia. Neurol Med Chir (Tokyo) 38:541–546
209. Yeh DD, Koch B, Crone KR (2006) Intraoperative ultrasonography used to determine the extent of surgery necessary during posterior fossa decompression in children with Chiari malformation type I. J Neurosurg 105(1 Suppl):26–32
210. Yilmaz N, Kiymaz N, Mumcu C (2005) Surgical treatment of craniocervical decompression without Chiari malformation in syringomyelia. Hiroshima J Med Sci 54:109–111
211. Yundt KD, Park TS, Tantuwaya VS, Kaufman BA (1996) Posterior fossa decompression without duraplasty in infants and young children for treatment of Chiari malformation and achondroplasia. Pediatr Neurosurg 25:221–226

Comments

Fumio Suzuki and Kazuhiko Nozaki, Shiga, Japan

The authors are to be congratulated for this comprehensive overview on the pathophysiology of syringomyelia. They made a modification of the theory proposed by Greitz D. in that a decreased compliance of the large veins in the subarachnoid space, which results from the reduced compliance of CSF below the obstruction, decreases the absorption of the extracellular fluid from intramedullary venous channels, resulting in the accumulation of extracellular fluid in spinal cord. This phenomenon might contribute partly to the development of syringomyelia but does not seem to be a main cause of syrinx formation. Although Greitz D. reported in his review that venous congestion might contribute to syrinx formation, venous congestion is not so obvious in Chari malformation type 1 as in spinal dural AVFs, in which large veins of the spinal cord are congested severely and compromised veins should reduce their compliance. These abnormal venous conditions may induce necrotizing myelitis but do not necessarily accompany syrinx formation. The authors referred to the report by Heiss J. et al. as an evidence of reduced CSF compliance, but the data was not statistically significant. More data about the changes in compliance of CSF space in Chiari Type 1 should be needed before establishing their modified theory.

Ricardo V. Botelho, São Paulo, Brazil

The authors performed a comprehensive review of mechanisms and concepts related to the pathogenesis of syringomyelia in Chiari malformation and have designed a hypothetical model of pathogenesis for syringomyelia.

Some of the factors reviewed are well established and others are hypothetical:

1. Patients with CM and sirirngomielia have smaller posterior fossa than those who did not have syringomyelia.

2. In patients with Chiari malformation, smaller tonsillar herniations are associated more frequently with syringomyelia than larger herniations.

The combination of these two features, small and shallow posterior fossa and small herniation of the tonsils might suggest a lower compliance of the foramen magnum, at the same time, prevents the descent of the tonsils and produces an early and intense blockage of free flow of craniocervial CSF in patients with syrinx.

3. Patients with syringomyelia have a blockage of subarachnoid CSF flow and less complacency of the subarachnoid space and posterior spinal veins.

4. The reduced absortion mechanism from the extracellular fluid from the spinal Cord parenchyma would result in syringomyelia in Chiari type 1 malformation, as speculated by the authors.

One real and observed effect in patients with syringomyelia and MC is that decompression of the posterior fossa often decreases syringomyelia cavity, probably by restoring the caniocervical flow of CSF.

The importance of reducing capacity venos absorption of extracellular fluid is an interesting suggestion posed by the authors that future works will confirm or not these suggestions.

Jörg Klekamp, Quakenbrück, Germany

In this paper, Koyanagi and Houkin present a hypothesis that was supposed to explain the development of syringomyelia in patients with a Chiari type I malformation. The authors correctly summarize in their paper that previous theories trying to explain syringomyelia by cerebrospinal fluid (CSF) entering the spinal cord via the 4th ventricle or other avenues have failed to demonstrate such a communication and are not able to explain several observations in these patients. Even though several thoughts and conclusions by the authors are well founded, I do have some reservations against this paper.

In table 1, the authors provide a list of previous theories and disqualify each of these as speculative. This statement is grossly negligent. Gardner's and Williams' theories, for instance, may no longer be tenable but were based on careful clinical tests, pressure recordings in patients and several animal studies. Given the technical conditions at the time, these works were state of the art and well founded on the observations made. Likewise, the theories of extracellular origin relating syringomyelia to edema formation are based on animal experiments and clinical observations and by no means just the result of a literature review.

The concept of syringomyelia as a spinal cord edema is by no means new. Tannenberg in 1924 and Liber and Lisa in 1937 were the first to propose this view. Taylor and Byrnes in 1974, Aboulker in 1979, and Yamada et al. in 1996 further elaborated on this theory and already emphasized the importance of venous obstruction, which they thought to cause syrinx formation in combination with CSF flow obstruction.

I do not agree with the authors' initial statement, that theories concerning the pathophysiology of syringomyelia on this basis do not apply to patients with a Chiari malformation. Several experimental studies have provided new insights into the physiological exchange between extracellular fluid (ECF) of the spinal cord and CSF under normal conditions as well as with CSF-flow obstructions. It appears that any pathology causing a CSF-flow obstruction and/or spinal cord tethering as well as certain intramedullary tumors are able to disturb the balance between ECF und CSF in the spinal canal, which may then lead to syrinx formation. This concept applies to patients with a Chiari malformation just as well as to those with posttraumatic syringomyelia, for instance. After all, syrinx formation in Chiari patients is the result of CSF-flow obstruction at the foramen magnum as it is in posttraumatic syringomyelia with CSF-flow obstruction at the level of the posttraumatic arachnopathy. With their hypothesis, Koyanagi and Houkin simply add a reduced compliance of posterior spinal cord veins to this concept of ECF/CSF imbalance. Spinal cord veins may turn out to contribute to syrinx formation in this setting but this assumption does not imply a completely novel hypothesis.

References

1. Aboulker J. (1979) La syringomyelie et les liquides intrarachidiens. Neurochirurgie 25 (Suppl):1–144.

2. Liber AF, Lisa JR. (1937) Rosenthal fibres in non-neoplastic syringomyelia: a note on the pathogenesis of syringomyelia. J. Nerv. Ment. Dis. 86:549–558.

3. Tannenberg J. (1924) Über die Pathogenese der Syringomyelie, zugleich ein Beitrag zum Vorkommen von Capillarhämangiomen im Rückenmark. Z. Neurol. 92:119–174.

4. Taylor AR, Byrnes DP. (1974) Foramen magnum and high cervical cord compression. Brain 97:473–480.

5. Yamada H, Yokota A, Haratake J, Horie A. (1996) Morphological study of experimental syringomyelia with kaolin-induced hydrocephalus in a canine model. J. Neurosurg. 84:999–1005.