

Scoliosis and Syringomyelia With Chiari Malformation After Lumbar Shunting

Hsin-Hung Chen^{1,3*}, Feng-Chi Chang^{2,3}, Tai-Tong Wong^{1,3}

¹Department of Neurosurgery, The Neurological Institute, ²Department of Radiology, Taipei Veterans General Hospital, and ³National Yang-Ming University School of Medicine, Taipei, Taiwan, R.O.C.

Unsteady gait was noted in a 2-year-old boy with a lumboperitoneal (LP) shunt that had been inserted 1 year earlier for increased head circumference caused by communicating hydrocephalus. Scoliosis was also noted during postoperative follow-up. Magnetic resonance imaging revealed new hindbrain tonsillar herniation and an extensive syrinx from C3 to L1. The malfunctioning LP shunt was removed and posterior fossa decompression with ventriculoperitoneal shunt insertion was performed. The unsteady gait recovered completely and scoliosis improved. Magnetic resonance imaging demonstrated resolution of the syrinx and tonsillar herniation. Acquired Chiari I malformation after LP shunt is well documented; usually, patients have no symptoms. This is the first report to have all the cause and effect mechanisms among syringomyelia, scoliosis and Chiari I malformation in 1 patient. We review the literature and discuss the possible mechanisms. [*J Chin Med Assoc* 2010;73(7):384–388]

Key Words: acquired Chiari malformation, hydrocephalus, lumboperitoneal shunt, scoliosis, syringomyelia

Introduction

Chiari first described 2 specific types of hindbrain deformities in 1891. Type I Chiari malformations are characterized by the presence of cerebellar tonsils in the upper cervical canal (at least 3–5 mm below the foramen magnum) while the 4th ventricle remains above the foramen magnum. Over the past 3 decades, there have been a number of reports of Chiari malformation and syringomyelia occurring either singly or together following cerebrospinal fluid (CSF) shunting from the lumbar subarachnoid space.^{1–7} Whether this “acquired” configuration is synonymous with a Chiari I malformation may be a matter of semantics, since the acquired descent of the cerebellar tonsils is radiographically indistinguishable from that of Chiari I malformations. So the term “acquired Chiari malformation” is used to describe this situation. The relationship between lumbar shunting procedures for hydrocephalus and Chiari I malformation has been established. Although no theory proves the mechanism by which this acquired Chiari malformation evolves, published theories invoke

a craniospinal pressure gradient^{8,9} or cephalocranial disproportion.^{1,10}

Syringomyelia is frequently associated with abnormalities of the craniocervical junction, particularly Chiari I malformation. A group of hydrodynamic mechanisms were introduced: it has been proposed that obstruction of CSF outflow from the 4th ventricle leads to dissection of CSF into the central canal and syrinx formation.^{11,12} It has also been pointed out that the abnormal pressure gradient from the cranial to the spinal subarachnoid space could suck CSF downward along the central canal and produce the syrinx.⁹ Ball and Dayan emphasized that with craniospinal dissociation, pressure waves in the spinal subarachnoid space remain confined to its space rather than dissipating throughout the entire CSF compartment. With a Valsalva maneuver, CSF could dissect into Virchow-Robin spaces and eventually produce gross cavities.¹³

Chiari malformations are often associated with spinal deformities, including scoliosis. Studies have suggested a causal relationship between syringomyelia and scoliosis. The incidence of scoliosis in patients with



*Correspondence to: Dr Hsin-Hung Chen, Department of Neurosurgery, The Neurological Institute, Taipei Veterans General Hospital, 201, Section 2, Shih-Pai Road, Taipei 112, Taiwan, R.O.C.
E-mail: Chen0403@tpts5.seed.net.tw • Received: October 2, 2009 • Accepted: May 14, 2010

Chiari malformation and syringomyelia is higher in the pediatric population than in adults. Early decompression of Chiari I malformation with syringomyelia and scoliosis resulted in improvement of stabilization of the spinal deformity.¹⁴⁻²⁰

We report the case of a boy who developed both Chiari I malformation and syringomyelia after lumboperitoneal (LP) shunt placement for the treatment of communicating hydrocephalus. Posterior fossa decompression and replacement of the LP shunt with a ventriculoperitoneal (VP) shunt resulted in radiographic resolution of both lesions as well as clinical improvement.

Case Report

The patient received a head injury when he was 2 months old, and brain computed tomography revealed hydrocephalus with marked ventriculomegaly. Magnetic resonance imaging (MRI) showed compatible findings (Figure 1), and I-131 RISA cisternography demonstrated progressive communicating hydrocephalus a few months later. He received LP shunt insertion in May 1996 when he was 10 months old. The head circumference grew within normal limits thereafter, with improvement of headache. Scoliosis of Cobb angle 28 degrees (Figure 2A), trunk tilting to the left side, and unsteady gait were noted 1 year later in May 1997 when he was 2 years old. MRI on May 27, 1997 (Figure 3) revealed obvious hydrocephalus, downward herniation of the cerebellar tonsils, and syrinx from C3 to L1. This was interpreted as a Chiari I malformation.



Figure 1. Preoperative sagittal T1-weighted magnetic resonance imaging of the brain.

On June 23, 1997, posterior fossa decompression surgery (suboccipital craniectomy with C1 laminectomy and duroplasty) and replacement of the LP shunt with a VP shunt were performed. MRI follow-up in September 1997, March 1998, and July 1999 revealed progressive resolution of the cerebellar tonsillar herniation and syrinx (Figure 4).

The unsteady gait improved straight after the operation, but clumsiness of fine motor skills was sustained. The scoliosis was also almost completely resolved (Figure 2B). At present, the patient's performance in elementary school is excellent.

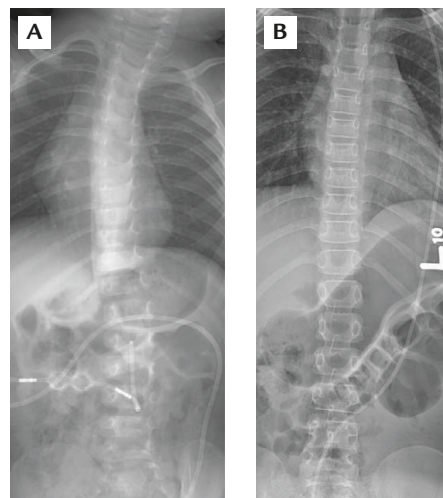


Figure 2. (A) After lumboperitoneal shunt insertion, progressive scoliosis change is noted on X-ray. (B) Regression of scoliosis after revision of lumboperitoneal shunt to ventriculoperitoneal shunt.

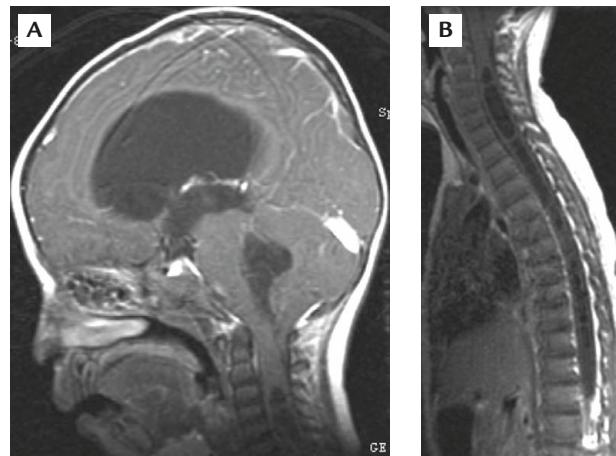


Figure 3. Twelve months after lumboperitoneal shunt insertion, contrast-enhanced magnetic resonance imaging of the (A) brain and (B) spine show hindbrain tonsil herniation below the foramen magnum and syrinx formation from C3 to L1.

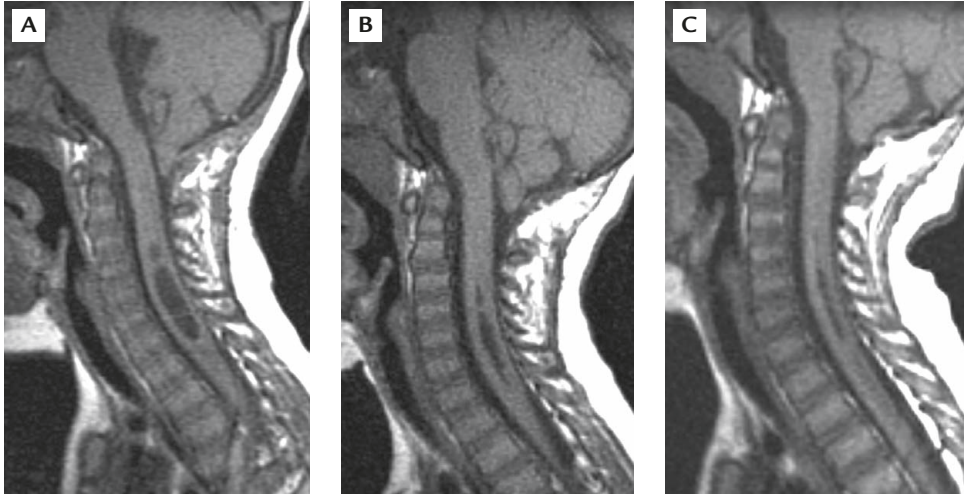


Figure 4. Non-contrast sagittal T1-weighted magnetic resonance imaging of the cervical spine shows progressive resolution of hindbrain downward herniation and cervical syrinx after surgical intervention: (A) 2 months; (B) 9 months; and (C) 2 years postoperatively.

Discussion

Chumas et al²¹ reported an incidence of 70% for asymptomatic tonsillar descent subsequent to lumbar shunting. We perform LP shunting instead of VP shunting in young patients with hydrocephalus to avoid the complication of overdrainage shunt malfunction. This case is the first experience of so-called “acquired Chiari malformation” with symptoms of syringomyelia and scoliosis in Taipei Veterans General Hospital. Two theories have been proposed to explain the development of low-lying tonsils in previously normal patients. In the first theory, cephalocranial disproportion attributable to underdevelopment of the posterior fossa, the cranial contents grow faster than the surrounding cranium and ultimately cause cerebellar tonsillar herniation into the foramen magnum.^{1,10} Previous studies indicate that shunting procedures usually arrest the growth of the skull. As the brain volume enlarges to fill the space previously occupied by the ventricular system, skull growth is assumed to be stimulated after occurrence of downward migration of cerebellar tonsils. Nevertheless, no acquired Chiari malformation has been found in association with ventricular shunting in any patient, and the skull growth is only transiently arrested subsequent to shunting (not every patient demonstrates evidence of gross posterior fossa maldevelopment in the end). Could this account for symptomatic brain stem compression many years later? This seems unlikely and leads to the postulation of another theory.

The second theory (craniospinal pressure gradient theory) invokes a pressure gradient across the cranial and spinal shunting procedure. This artificially created

pressure gradient is the suspected driving force for tonsillar descent. In addition, the compromise of normal CSF flow dynamics at the level of the foramen magnum results in acute obstructive hydrocephalus, further worsening the craniospinal pressure gradient.^{8,9}

The mechanism of syringomyelia formation with Chiari malformation is not clearly understood. Three hydrodynamic theories of origin exist: the “water-hammer” theory of Gardner,^{11,12} Williams’ “1-way-valve” or “craniospinal dissociation” theory,⁹ and revision of the Heiss-Oldfield theory.^{22,23} In the water-hammer theory, arterial pulses from the choroid plexus are thought to be transmitted down through an abnormal 4th ventricle to the cord, “hammering” out a dilatation in the cord; this mechanism has been essentially disproven using MRI. In the 1-way-valve theory, the abnormal communication between the 4th ventricle and the spinal cord causes an unequal pressure gradient with Valsalva maneuver, resulting in high pressure in the spinal column that causes cord tissue dilatation by “hydrodissection”. However, this process could not be replicated by Heiss et al and Oldfield et al using cine MRI and intraoperative ultrasound study, respectively. The Heiss-Oldfield theory: occlusion at the foramen magnum causes CSF pulsations during cardiac systole to be transmitted through the Virchow-Robins spaces, which increases the extracellular fluid, which coalesces to form a syrinx. Finally, Levine²⁴ concluded that in the presence of subarachnoid obstruction at the foramen magnum, erect posture, coughing or straining, and pulsatile fluctuations of CSF pressure during the cardiac cycle produce transiently higher CSF pressure above the block than below it. There are corresponding changes in transmural

venous and capillary pressure favoring dilation of vessels below the block and collapse of vessels above the block. The mechanical stress on the spinal cord, coupled with venous and capillary dilation, partially disrupt the blood-spinal cord barrier, allowing ultrafiltration of crystalloids and accumulation of a protein-poor fluid. The hypothetical assumption is consistent with the neuropathological findings in syringomyelia (which include: the pressure in syringomyelia is higher than CSF pressure, extensive gliosis, edema, and vascular wall thickening regularly occur around the syrinx, and the fluid of syringomyelia is not identical with that of CSF).

The relationship between Chiari malformation and scoliosis has been recognized but not completely understood. Multiple theories have been proposed to explain the association. Huebert and MacKinnon theorized that syringomyelia compromises cells in the cord that are responsible for muscle balance of the trunk, leading to scoliosis.²⁵ Decompression of the Chiari formation with syringomyelia has been shown to halt or improve neurological abnormalities associated with this condition. The incidence of scoliosis in patients with Chiari malformation and syringomyelia is higher in the pediatric population: 82% of patients younger than 20 years with syringomyelia have scoliosis, compared with 18% of patients older than 20 years.¹⁸ Scoliosis in patients with Chiari malformation and syringomyelia can occur at very young ages and can rapidly progress, necessitating early diagnosis and treatment to prevent the need for spinal fusion (before permanent structural spinal changes).²⁰

Our case supports Welch et al's conclusion once again that Chiari I malformation can be an acquired condition related to disproportionate spinal CSF absorption and resultant hydrocephalus pushing the tonsils further downward.⁸ The mechanism for syrinx formation in our case is best explained by the Levine theory.²⁴ Then, it is the syringomyelia-compromised cells in the spinal cord that are responsible for scoliosis. After performing posterior fossa decompression and VP shunting revision simultaneously (since subarachnoid obstruction at the foramen magnum and subsequent hydrocephalus were the 2 main causes), both the Chiari I malformation and syrinx were shown radiographically to be resolved due to normalized CSF dynamics. According to the Levine theory, we neither plugged the obex nor opened the arachnoid membrane during decompression surgery to prevent the risk of CSF leakage and arachnoiditis. The syrinx completely resolved within several months, implying an active movement of CSF between the cavity and spinal subarachnoid space. The Cobb angle of scoliosis was

reduced in turn. Consequently, no syringomyelia shunting or orthopedic surgery for scoliosis was needed for the patient. After reviewing the literature, we believe that this is the first case to demonstrate such stereotypical presentation and resolution of disease according to the proposed pathophysiological mechanisms in 1 patient.

References

- Hoffman HJ, Tucker WS. Cephalocranial disproportion: a complication of the treatment of hydrocephalus in children. *Childs Brain* 1976;2:167-76.
- Johnston I, Jacobson E, Besser M. The acquired Chiari malformation and syringomyelia following spinal CSF drainage: a study of incidence and management. *Acta Neurochirurgica* 1998; 140:417-28.
- Kushner J, Alexander E Jr, Davis CH Jr, Kelly DL Jr. Kyphoscoliosis following lumbar subarachnoid shunts. *J Neurosurg* 1971;34:783-91.
- Padmanabhan R, Crompton D, Burn D, Birchall D. Acquired Chiari I malformation and syringomyelia following lumboperitoneal shunting for pseudotumour cerebri. *J Neurol Neurosurg Psychiatry* 2005;76:298.
- Riffaud L, Moughty C, Henaux PL, Haegelen C, Morandi X, Riffaud L, Moughty C, et al. Acquired Chiari I malformation and syringomyelia after valveless lumboperitoneal shunt in infancy. *Pediatr Neurosurg* 2008;44:229-33.
- Fischer EG, Welch K, Shillito J Jr. Syringomyelia following lumboureteral shunting for communicating hydrocephalus: report of three cases. *J Neurosurg* 1977;47:96-100.
- Sathi S, Stieg PE. "Acquired" Chiari I malformation after multiple lumbar punctures: case report. *Neurosurgery* 1993;32: 306-9.
- Welch K, Shillito J, Strand R, Fischer EG, Winston KR. Chiari I "malformations": an acquired disorder? *J Neurosurg* 1981; 55:604-9.
- Williams B. On the pathogenesis of syringomyelia: a review. *J R Soc Med* 1980;73:798-806.
- Venes JL. Arnold-Chiari malformation in an infant with Kleblattschadel: an acquired malformation? *Neurosurgery* 1988; 23:360-2.
- Gardner WJ. Hydrodynamic mechanism of syringomyelia: its relationship to myelocoele. *J Neurol Neurosurg Psychiatry* 1965; 28:247-59.
- Gardner WJ, Angel J. The mechanism of syringomyelia and its surgical correction. *Clin Neurosurg* 1958;6:131-40.
- Ball MJ, Dayan AD. Pathogenesis of syringomyelia. *Lancet* 1972;2:799-801.
- Bertrand SL, Drvaric DM, Roberts JM. Scoliosis in syringomyelia. *Orthopedics* 1989;12:335-7.
- Dyste GN, Menezes AH. Presentation and management of pediatric Chiari malformations without myelodysplasia. *Neurosurgery* 1988;23:589-97.
- Farley FA, Song KM, Birch JG, Browne R. Syringomyelia and scoliosis in children. *J Pediatr Orthop* 1995;15:187-92.
- Ghanem IB, Londono C, Delalande O, Dubousset JF. Chiari I malformation associated with syringomyelia and scoliosis. *Spine* 1997;22:1313-8.
- Isu T, Iwasaki Y, Akino M, Abe H. Hydrosyringomyelia associated with a Chiari I malformation in children and adolescents. *Neurosurgery* 1990;26:591-7.

19. Phillips WA, Hensinger RN, Kling TF Jr. Management of scoliosis due to syringomyelia in childhood and adolescence. *J Pediatr Orthop* 1990;10:351-4.
20. Eule JM, Erickson MA, O'Brien MF, Handler M, Eule JM, Erickson MA, O'Brien MF, et al. Chiari I malformation associated with syringomyelia and scoliosis: a twenty-year review of surgical and nonsurgical treatment in a pediatric population. *Spine* 2002;27:1451-5.
21. Chumas PD, Armstrong DC, Drake JM, Kulkarni AV, Hoffman HJ, Humphreys RP, Rutka JT, et al. Tonsillar herniation: the rule rather than the exception after lumboperitoneal shunting in the pediatric population. *J Neurosurg* 1993;78:568-73.
22. Heiss JD, Patronas N, DeVroom HL, Shawker T, Ennis R, Kammerer W, Eidsath A, et al. Elucidating the pathophysiology of syringomyelia. *J Neurosurg* 1999;91:553-62.
23. Oldfield EH, Muraszko K, Shawker TH, Patronas NJ. Pathophysiology of syringomyelia associated with Chiari I malformation of the cerebellar tonsils. Implications for diagnosis and treatment. *J Neurosurg* 1994;80:3-15.
24. Levine DN. 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* 2004;220:3-21.
25. Huebert HT, MacKinnon WB. Syringomyelia and scoliosis. *J Bone Joint Surg Br* 1969;51:338-43.