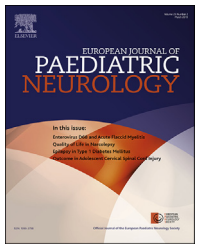
Official Journal of the European Paediatric Neurology Society



Case study

Clinical deterioration despite syringomyelia resolution after successful foramen magnum decompression for Chiari malformation - Case series

J. Shetty [a,b,*], J. Kandasamy [a], D. Sokol [a], P. Gallo [a]

[a] Department of Paediatric Neurosciences, Royal Hospital for Sick Children, Edinburgh, UK
[b] University of Edinburgh, UK

ARTICLE INFO

Article history:
Received 16 September 2018
Received in revised form
27 December 2018
Accepted 8 January 2019

Keywords:
Syringomyelia
Foramen magnum decompression
Chiari malformation

ABSTRACT

Introduction: Neurosurgical treatment is recommended for symptomatic syringomyelia and the post-operative radiological resolution of the syringomyelia is associated with an improvement or at least stability of the patient's pre-operative symptoms.
Methods: We reviewed syringomyelia treatment in our centre over the last five years for clinical outcome, surgical complications, post operative MRI and long term symptom resolution.
Results: 50 cases of symptomatic syringomyelia underwent foramen magnum decompression and expansile watertight duroplasty. While the outcomes for majority are similar to what published in literature, three of them developed typical syringomyelia symptoms after initial good recovery and radiological resolution of syrinx.
Conclusion: Syringomyelia symptoms may appear or worsen following successful surgical treatment and radiological resolution of syrinx and it is important to counsel young people and their family regarding this.

1. Introduction

Syringomyelia is a common clinical problem in paediatric neurology setting, characterised by a cystic cavity (syrinx) within the spinal cord. The pathophysiology of the formation of the syrinx and its propagation is still unclear and a much debated topic in neurosurgery. There are several theories proposed from autopsy studies as well as radiological studies.[1–4] Syringomyelia is most frequently associated with hindbrain herniation also known as Chiari I malformation (CIM). CIM is defined as the descent of the cerebellar tonsils for 5 mm or more beyond the foramen magnum. Majority of the idiopathic syringomyelia seem to be asymptomatic and remain stable not requiring any intervention.[5]

* *Corresponding author*. Department of Paediatric Neurosciences, Royal Hospital for Sick Children, Edinburgh, EH9 1LF, UK.
E-mail address: Jay.Shetty@ed.ac.uk (J. Shetty).
https://doi.org/10.1016/j.ejpn.2019.01.003

Clinical presentation of syringomyelia varies from being asymptomatic to symptoms including scoliosis, sensory, motor and autonomic deficits. Scoliosis is the commonest presentation in paediatric age group. Syrinx extension into the anterior horns of the spinal cord can damage motor neurons (lower motor neuron) and cause diffuse muscle atrophy.

Magnetic resonance imaging (MRI) is the investigation of choice and neurophysiological assessments may be beneficial. Majority of the cases of syringomyelia are associated with hindbrain deformities of which Chiari 1 malformation (CIM) is the commonest. In paediatric population CIM and syringomyelia show much higher incidence of scoliosis compared to other symptoms.[6]

Treatment and monitoring of asymptomatic CIM and syringomyelia is widely debated and there is variation in practice. However, it is standard practice to treat CIM associated with syringomyelia in symptomatic children. Neurosurgical treatment is the widely accepted only treatment option and there are several techniques with varying success.[6–9] The safety, effectiveness and outcome of different surgical modalities of cranio-cervical decompression have been widely reported.[10–14] Although the surgical techniques vary there seems to be a consensus to perform surgery in CIM with syringomyelia in children presenting with scoliosis.

Neurosurgical procedures are not without any complications. Some of the commonest complications including cerebrospinal fluid leak, pseudomeningocele, meningitis, wound infection, hydrocephalus, subdural collections and others are well documented.[15] Aim of the foramen magnum decompression in CIM and syringomyelia is to improve the patient symptoms and at least prevent further increase in size or reduce the extent of the syrinx.

Post-operative radiological resolution of the syringomyelia is associated with an improvement or at least stability of the patient's pre-operative symptomatology. There are no reported cases of post-operative symptomatic deterioration after radiological syringomyelia improvement or resolution.

We report here three cases where typical syringomyelia signs and symptoms appeared after a successful and uneventful cranio-cervical decompression demonstrated by a radiological dramatic improvement or complete resolution of the pre-operative syringomyelia.

2. Methods

We retrospectively reviewed all the 50 paediatric cases who underwent cranio-cervical decompression for CIM and syringomyelia over the last 5 years at our Institution and identified 3 cases. Here we present in detail their clinical presentation, neurosurgical procedure, clinical and radiological outcome.

3. Results

3.1. *Case 1*

A 11-year old girl was seen in Scottish National Scoliosis service and was diagnosed to have adolescent idiopathic triple thoracic and lumbar scoliosis (Lenke type 2a). She was otherwise clinically well. X-rays of the spine showed triple curve pattern with a right upper thoracic curve extending from T1 to T6, a left low thoracic curve extending from T7 to T12 and a right lumbar curve extending from L1 to L4. This was producing a significant deformity due to prominence of the ribs adjacent to the convexity of the left thoracic curve as well as a mild waistline asymmetry. MRI of her spine (Fig. 1) showed a large syringomyelia extending from C4 to T8, measuring up to six millimetres in the antero–posterior plane and nine millimetres in the lateral plane within the cervical cord, no evidence of tethering distally, but a cerebellar tonsillar herniation of nine mm below the foramen magnum.

On clinical examination she had normal cranial nerve findings with normal power and sensation to light touch bilaterally. Her long tract signs were negative and her reflexes were symmetrical. She underwent foramen magnum decompression and expansile watertight duroplasty. The procedure was uneventful and she made a good recovery immediately after surgery but on day three she developed aseptic meningitis which resolved in five days, prolonging her hospital stay over a week. Two months after the surgery she started to complain "*pins and needles*", pain and numbness in her hands and legs. Following this, she had a repeat MRI scan, which showed that the size of her cervicothoracic syringomyelia had reduced significantly (Fig. 1) Patient refused to undergo scoliosis surgery and she is still symptomatic 4 years after the initial surgical procedure.

3.2. *Case 2*

A 12 year old boy with a severe progressive scoliosis was investigated with a whole spine MRI (Fig. 2) which showed a CIM with a 7 mm tonsillar descent and a holocord syringomyelia. He was clinically asymptomatic and no neurological signs were found on examination. He underwent a foramen magnum decompression plus expansile watertight duroplasty. There were no procedure related complications and his post-operative recovery was quick and uneventful. A follow-up MRI performed 4 months after the surgery already showed a remarkable shrinkage of the syringomyelia.

One year later, during his routine follow-up in clinic he was complaining numbness, lack of sensation and weakness in his arms and legs. A second post-operative MRI scan was performed which surprisingly showed a nearly complete resolution of the holocord syringomyelia and a capacious cisterna magna. Despite these imaging findings (Fig. 2), the patient is still experiencing episodes of numbness, lack of sensation and weakness in his arms and legs four years after the operation.

3.3. *Case 3*

A 5-year old girl who was diagnosed to have CIM with a small signal change in the upper cervical cord and apnoea during infancy, underwent a successful foramen magnum osteoligament decompression (without duroplasty) at one year of age and was being followed up in the neurosurgical clinic. In view of her previous scan abnormalities she underwent repeat imaging which revealed a large and extensive syringomyelia extending from C2 down to T10 measuring about eight millimetres in diameter.

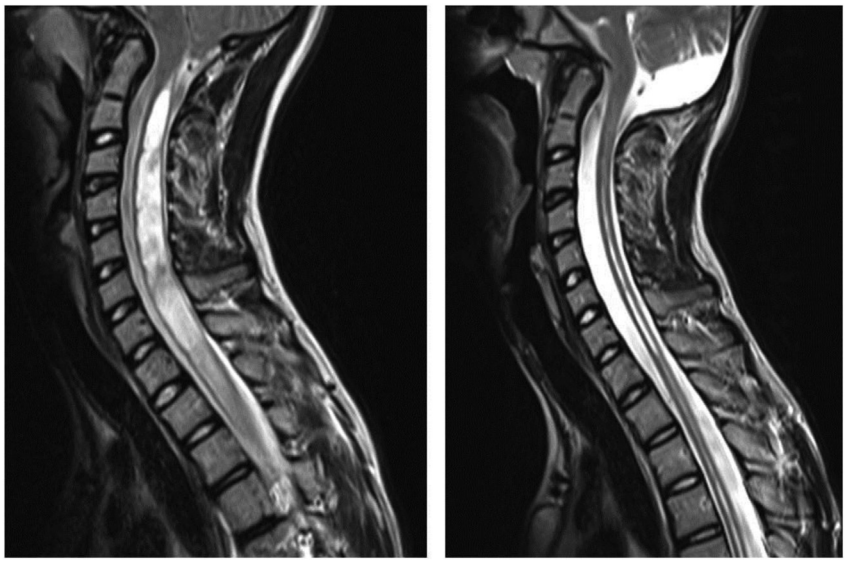


Fig. 1 – MRI spine (left) showing a large syringomyelia extending from C4 to T8 and cerebellar tonsillar herniation of nine millimiters below the foramen magnum. MRI spine (right) 3 months after foramen magnum decompression and expansile watertight duroplasty.

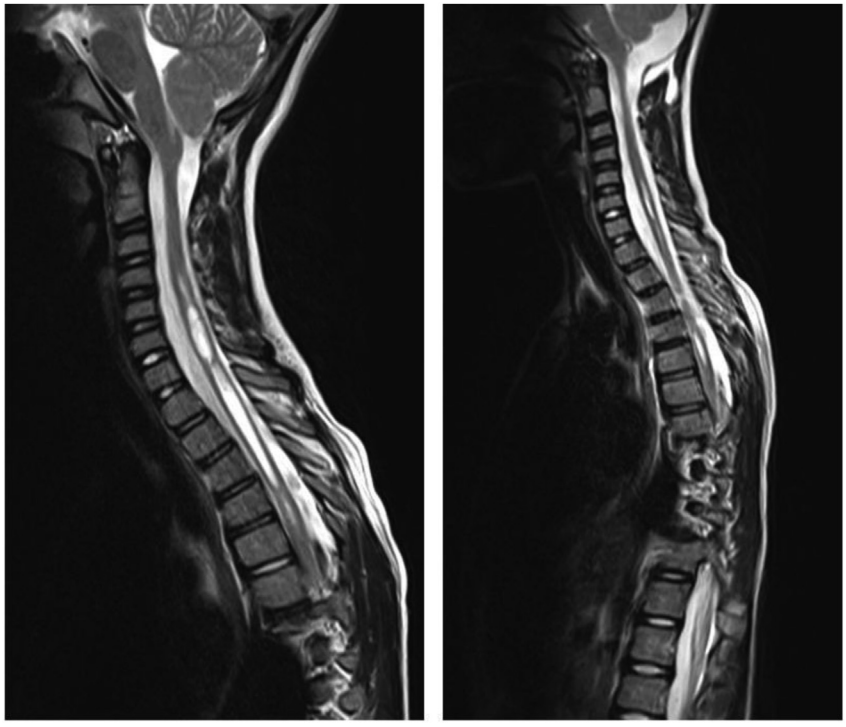


Fig. 2 – MRI (left) showing CIM with a 7 millimiters tonsillar descent and a holocord syringomyelia. Follow up MRI (right) 4 months after a foramen magnum decompression and expansile watertight duroplasty showing shrinkage of the syringomyelia.

On clinical examination she had reduced cold sensation in her upper limbs and reduced pin prick sensation in her left hand; the rest of the neurological examination was normal. Following discussion with parents she underwent repeat foramen magnum decompression plus expansile watertight duroplasty. She recovered well and her symptoms were resolved at the initial follow up visit. Her repeat MRI (Fig. 3) showed a good shrinkage of the syringomyelia. At one-year follow-up, her pre-operative signs were back and she complained of dysesthetic legs pain and urinary issues in terms of urgency and occasional accidents.

This was investigated with a new whole spine MRI which surprisingly showed a complete resolution of the preoperative syringomyelia and a well decompressed cranio-cervical junction. Nearly five years after the surgical procedure she continues to be symptomatic.

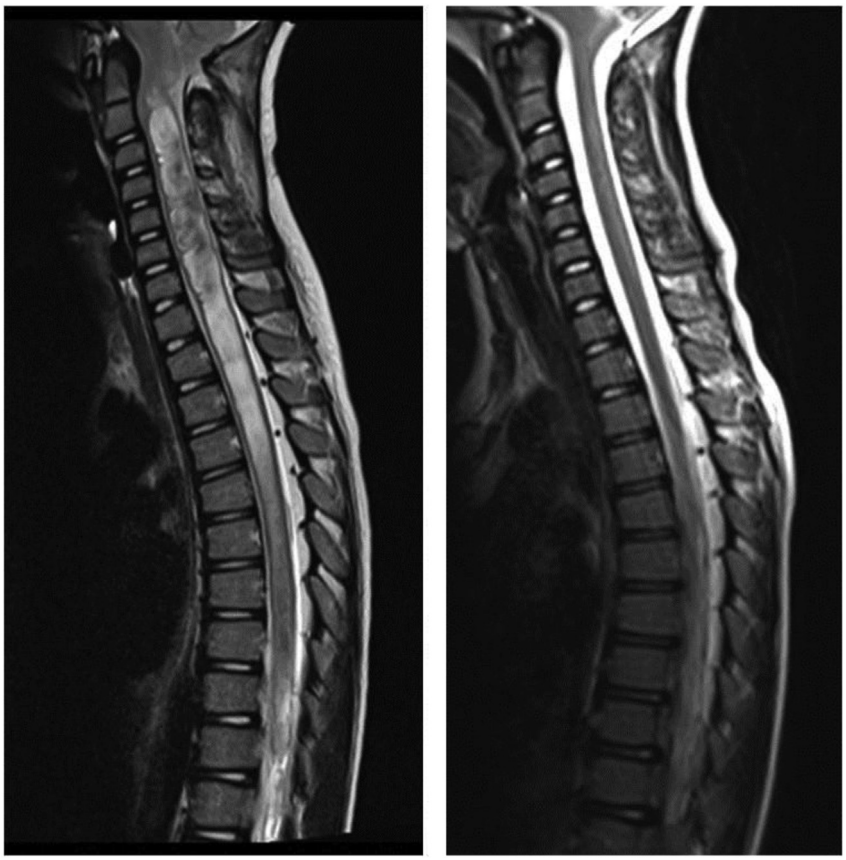


Fig. 3 – MRI spine (left) showing large and extensive syringomyelia extending from C2 down to T10 and the repeat MRI (right) 12 months after foramen magnum decompression plus expansile watertight duroplasty.

4. Discussion

The aim of foramen magnum decompression is to relieve patient's symptoms and prevent further progression of the syringomyelia and the potential damage to the neural tissue. Up to 80% of patients with hindbrain-related syringomyelia experience clinical improvement after cranio-vertebral decompression. Unfortunately, established neurological deficits such as sensory loss, weakness, established spasticity and early myelopathy improve little. The majority of patients (70–80%) shows a radiological improvement of the syringomyelia cavity and another 15% has no change but it does not deteriorate with time either. It is known that a number of patients, despite radiological control of the syringomyelia, continue to deteriorate clinically.[10–14]

It may be possible whilst syringomyelia is developing, the nerve fibres reorganised and the process of decompression may alter this dynamic. However, as we do not know the exact mechanism of syringomyelia development, it is difficult to be sure of these symptoms either. Although the cranio-cervical decompression reverses the pathophysiological process that led to the development of the syringomyelia, it could not reverse the anatomical destruction of the spinal cord caused by the distended syringomyelia before surgery, which explains the persistent signs and symptoms of myelopathy experienced by most patients after surgery despite the consistent collapse of the syringomyelia.[16]

It has been noted in post-mortem examinations that long-standing syringomyelia provokes gliosis formation in the cavity walls and the surrounding cord. Gliosis inside the cord may lead to progressive neurological loss even after successful surgery and collapse of the cavity. It has been described that residual central myelopathy can persist after resolution (also spontaneous) of the syringomyelia, defined these stable symptoms and signs of central myelopathy seen in their patients with absent or spontaneous collapsed syringomyelia in un-operated cases of syringomyelia associated to CMI as "post-syringomyelia" syndrome.[17]

Whatever is the mechanism it is important to counsel the young people and their family, that there is a small possibility of these symptoms appearing or deteriorating after a successful surgery. It is also important to follow these children up for longer time to see if these symptoms persist. It is likely other centres may have seen similar cases and a collaborative approach may help understand the long term outcome. It may also be beneficial to consider neurophysiological testing in these cases.

Acknowledgements

JS and JK are supported by NHS Research Scotland Career Researcher Clinicain award.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.ejpn.2019.01.003.

REFERENCES

1. Ball MJ, Dayan AD. Pathogenesis of syringomyelia. *Lancet* 1972;**2**(7781):799–801.
2. Milhorat TH, Capocelli Jr AL, Anzil AP, Kotzen RM, Milhorat RH. Pathological basis of spinal cord cavitation in syringomyelia: analysis of 105 autopsy cases. *J Neurosurg* 1995;**82**(5):802–12.
3. Koyanagi I, Houkin K. Pathogenesis of syringomyelia associated with chiari type 1 malformation: review of evidences and proposal of a new hypothesis. *Neurosurg Rev* 2010;**33**(3):271–84. discussion 284-5.
4. Jones BV. Cord cystic cavities: syringomyelia and prominent central canal. *Semin Ultrasound CT MR* 2017;**38**(2):98–104.
5. Magge SN, Smyth MD, Governale LS, et al. Idiopathic syrinx in the pediatric population: a combined center experience. *J Neurosurg Pediatr* 2011;**7**(1):30–6.
6. Hida K, Iwasaki Y, Koyanagi I, Abe H. Pediatric syringomyelia with chiari malformation: its clinical characteristics and surgical outcomes. *Surg Neurol* 1999;**51**(4):383–90. discussion 390-1.
7. Arnautovic A, Splavski B, Boop FA, Arnautovic KI. Pediatric and adult chiari malformation type I surgical series 1965-2013: a review of demographics, operative treatment, and outcomes. *J Neurosurg Pediatr* 2015;**15**(2):161–77.
8. Zhao JL, Li MH, Wang CL, Meng W. A systematic review of chiari I malformation: techniques and outcomes. *World Neurosurg* 2016;**88**:7–14.
9. Liu H, Yang C, Yang J, Xu Y. Pediatric chiari malformation type I: long-term outcomes following small-bone-window posterior fossa decompression with autologous-fascia duraplasty. *Exp Ther Med* 2017;**14**(6):5652–8.
10. Di X. Endoscopic suboccipital decompression on pediatric Chiari type I. *Minim Invasive Neurosurg* 2007;**52**:119–25.
11. Tubbs RS, Beckman J, Naftel RP, Chern JJ, Wellons III JC, Rozzelle CJ, et al. Institutional experience with 500 cases of surgically treated pediatric Chiari malformation type I. *J Neurosurg Pediatr* 2011;**7**:248–56.
12. Litvack ZN, Lindsay RA, Selden NR. Dura splitting decompression for Chiari I malformation in pediatric patients: clinical outcomes, healthcare costs and resource utilization. *Neurosurgery* 2013;**72**:922–9.
13. Kennedy BC, Kelly KM, Phan MQ, Bruce SS, McDowell MM, Anderson RCE, et al. Outcome after suboccipital decompression without dural opening in children with Chiari malformation type I. *J Neurosurg Pediatr* 2015;**16**:150–8.
14. Gallo P, Sokol D, Kaliaperumal C, Kandasamy J. Comparison of three different cranio-cervical decompression procedures in children with chiari malformation type I: does the surgical technique matter? *Pediatr Neurosurg* 2017;**52**(5):289–97. https://doi.org/10.1159/000479327. Epub 2017 Aug 25.
15. Greenberg JK, Olsen MA, Yarbrough CK, et al. Chiari malformation type I surgery in pediatric patients. part 2: complications and the influence of comorbid disease in California, Florida, and New York. *J Neurosurg Pediatr* 2016;**17**(5):525–32.
16. Wetjen NM, Heiss JD, Oldfield EH. Time course of syringomyelia resolution following decompression of Chiari malformation Type I. *J Neurosurg Pediatr* 2008 Feb;**1**(2):118–23. https://doi.org/10.3171/PED/2008/1/2/118.
17. Bogdanov EI, Heiss JD, Mendelevich EG. The post-syrinx syndrome: stable central myelopathy and collapsed or absent syrinx. *J Neurol* 2006 Jun;**253**(6):707–13. Epub 2006 Mar 6.