De novo syrinx formation in a patient with type I Chiari malformation: Case report and review of the literature

ABSTRACT
This report highlights the case of a 56-year-old woman with an incidental type I Chiari malformation who on initial presentation had no associated cervical syrinx on magnetic resonance imaging (MRI), but who on subsequent MRI 8 years later was found to have developed a de novo upper cervical cord syrinx.

Keywords: Development, magnetic resonance imaging scan, syrinx, type I Chiari malformation

INTRODUCTION
Type I Chiari malformations occur when there is herniation of the cerebellar tonsils thought the foramen magnum by a minimum of 5 mm. They can be asymptomatic or present with a range of symptoms including Valsalva induced headache and syncope. Type I Chiari malformations may also be complicated by a spinal cord syrinx. In the absence of specific indications for surgery, such as a spinal cord syrinx, patients are often managed conservatively and do not have routine follow-up imaging.

Here we present the case of a 56-year-old lady who presented with an asymptomatic type I Chiari malformation and was discharged for routine follow-up. She went on to develop a spinal cord syrinx years later. Along with the case we review previously published literature around this topic and any potential implications for practice.

CASE REPORT
A 56-year-old, right-hand-dominant female was referred to our Chiari malformation outpatient clinic following cranial magnetic resonance imaging (MRI) for new visual symptoms. She had a history of minor occipital headaches worsened by Valsalva maneuver dating back to childhood not significantly interfering with lifestyle. She had no history of Valsalva syncope. Her past medical history included diabetes, hypertension, stroke, and gout. She was a known Chiari on the basis of an historic MRI 8 years back, but no syrinx was present [Figure 1]. On examination, she had residual left-sided altered sensation and weakness from the previous stroke. She had a positive Hoffman’s sign on the left, but no other upper motor neuron signs and no nystagmus.

Her current MRI of head and spine further demonstrated her known Chiari malformation, but now with the development of an upper cervical cord syrinx [Figures 2 and 3]. She subsequently underwent an uncomplicated cranio cervical decompression procedure with postoperative MRI imaging at 2 months postsurgery, showing decompression of the Chiari malformation and reduction in syrinx appearances [Figure 4].

This patient was managed within a single surgeon specialist clinic, in which 295 new patients with Chiari 1 were seen...
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over a 9-year period. Forty-nine of these patients had had a syrinx (including this patient), but this patient was the only individual who had historic imaging showing a previous normal cervical cord.

DISCUSSION

Chiari malformations are a group of related hindbrain abnormalities, with type I malformation being the most prevalent. In type I Chiari malformations, the cerebellar tonsillar herniate through the foramen magnum by >5 mm. The prevalence of asymptomatic/incidental type I Chiari malformations has been calculated to be 0.24% (95% confidence interval: 0.04–0.58) of the population.\(^1\) When symptoms are present, it can include occipital headaches and syncope induced by the Valsalva maneuver (where intrathoracic pressure is increased by action such as coughing and straining). They may also be associated with a wide range of neurological symptoms and occasionally signs including vertigo, paresthesia, and nystagmus.\(^2\) In addition, Chiari malformations can be associated with syrinx formation in the spinal cord in an undetermined percentage of cases and if present can cause irreversibly progressive neurologic deficit. Strahle et al. described syringomyelia in 13.5% of 147 children with Chiari undergoing MRI.\(^3\) The indications for surgery in patients with Chiari malformation include severe Valsalva headaches affecting the patient’s quality of life and the presence of a syrinx in the spinal cord.

Incidental (asymptomatic) or minimally symptomatic Chiari malformations not meeting indications for surgery will be managed conservatively with no long-term arrangements for clinic or imaging follow-up. Such individuals, however, may enquire about the risk if any of developing a syrinx over time.
This case report highlights a patient with a minimally symptomatic Chiari malformation without syrinx initially who went on to subsequently develop a syrinx in her cervical spinal cord 8 years after initial presentation. There is minimal information in the published literature about the risk of patients with Chiari malformation who are initially identified as not having a syrinx going on subsequently to develop a \textit{de novo} syrinx. In the adult literature, there is only one case series which describes two patients who subsequently go on to develop a syrinx. These patients developed their syrinx over 3 and 4 years between initial and subsequent scan. Both these patients experienced an increase in body mass index and their “headaches worsened and neurological function deteriorated” – a feature not seen in our patient.\cite{arnautovic2013association}

In the pediatric literature, there is just one paper describing subsequent \textit{de novo} syrinx development in individuals with Chiari type I malformation. In this case series of 147 children managed conservatively, three children developed a syrinx in previously intact spinal cord, suggesting that this may be more common in the pediatric population.\cite{strahle2011natural}

This report demonstrates that subsequent syrinx formation can occur in an individual with Chiari and no syrinx on presentation MRI. However, this occurrence is likely to be infrequent. It would seem reasonable therefore to feedback to individuals with Chiari being seen in the clinic that there is a possibility of syrinx, but not so much of a concern to warrant ongoing surveillance MRI. There should be a low threshold to reimage in the event of significant change to neurologic status, and this low threshold to reimage should be recorded in documentation provided to the patient and primary care physician/referrer.

\textbf{Declaration of patient consent}

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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\textbf{Conflicts of interest}

There are no conflicts of interest.

\textbf{REFERENCES}

1. Morris Z, Whiteley WN, Longstreth WT Jr., Weber F, Lee YC, Tsushima Y, \textit{et al.} Incidental findings on brain magnetic resonance imaging: Systematic review and meta-analysis. BMJ 2009;339:b3016.
2. Chavez A, Roguski M, Killeen A, Heilman C, Hwang S. Comparison of operative and non-operative outcomes based on surgical selection criteria for patients with Chiari I malformations. J Clin Neurosci 2014;21:2201‑6.
3. Strahle J, Muraszko KM, Kapurch J, Bapuraj JR, Garton HJ, Maher CO, \textit{et al.} Natural history of Chiari malformation type I following decision for conservative treatment. J Neurosurg Pediatr 2011;8:214‑21.
4. Arnautovic KI, Muzevic D, Splavski B, Boop FA. Association of increased body mass index with Chiari malformation type I and syrinx formation in adults. J Neurosurg 2013;119:1058‑67.