Natural history of syringomyelia associated with Chiari I abnormality: Appearance of de novo cervical spinal cord syrinx in a child: Case report. Sequential imaging in an asymptomatic

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A 34 month old boy was referred to neurosurgery after an MRI of the brain obtained during workup for intractable vomiting disclosed a Chiari I abnormality. By the time he appeared for his initial visit to the Neurosurgery Clinic, dietary adjustment had effected cessation of his vomiting. Survey spine MRI was normal. Since he was asymptomatic, his physical examination was normal, and no pre-syrinx changes were present, we recommended observation.

One year later, age 4, though he remained asymptomatic with a normal physical examination and BMI, surveillance MRI revealed a dilated multi-loculated syrinx extending from C3-T1. We recommended FMD and duraplasty.
Surgery proceeded uneventfully, followed by a > 50% decrease in syrinx volume on follow-up imaging one year postop. He remains asymptomatic at age 6.
Nishizawa et al (2001)\(^1\) described 9 adult patients with incidentally discovered, asymptomatic syringomyelia. Discussing the natural history of syringomyelia associated with Chiari I abnormality, they noted that the MRI parameters of their patients did not provide predictable values to recommend interventional surgery. They considered foramen magnum decompression (FMD) unnecessary in the absence of symptomatic or imaging changes. Amin-Hanjani et al (1995)\(^2\) reported 2 toddlers with simultaneous \textit{de novo} development of a Chiari I and syrinx, however, one patient had previously suffered IVH, placement of VP shunt, and shunt revision. The other patient’s \textit{de novo} Chiari I and syrinx appeared on an MRI obtained 1 year after a normal brain study obtained for developmental delay. It is difficult to determine if the original studies of either patient included adequate spinal cord imaging documenting a normal spinal cord.


Of 22 children aged 1 to 16 with Chiari I followed 3 to 19 years by Novegno et al (2008), three developed a de novo syrinx $\leq 4$mm maximum diameter; only one had an “uncomplicated” Chiari I, the other 2 also had hydrocephalus associated with the Chiari I abnormality.

Strahle et al (2011), reporting the natural history in a series of un-operated Chiari I patients followed for at least 1 year with imaging and physical examination, encountered 2 children who developed a syrinx in a previously normal-appearing spinal cord. Apparently both eventually underwent Chiari decompression, although time elapsed from initial MRI study to discovery of the new syrinx was not listed specifically for these two children, the interval for the group of 7 patients developing a “new” syrinx ranged from 2-74 months.


CONCLUSION

Reports of the development of a *de novo* syrinx in a clinically normal child being followed for an “incidental Chiari I” are rare. Our case, together with the few others referred to above, may challenge skepticism regarding the need for imaging follow-up of these frequently referred, newly diagnosed “incidental” Chiari I patients.