Spinal cord and brain stem anomalies in scoliosis

MR screening of 26 cases

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The spinal cord and brain were examined with magnetic resonance (MR) in 26 patients with idiopathic scoliosis to study the prevalence of spinal cord and brainstem anomalies. Two patients had small centrally located spinal cord syrinxes, one at C6–C8 and the other at T6–T8, without association with any brainstem or cerebellar deformity. The lowest position of the cerebellar tonsils was 0.5 cm below the foramen magnum, which was also seen in 2 other patients. Scoliosis associated with syringomyelia may be more common than previously thought, and may be wrongly classified as idiopathic. A neurogenic cause of scoliosis should always be considered, and at least in atypical cases be excluded by MR imaging of the spinal cord. MR should be mandatory before bracing or operative correction of scoliosis.

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Submitted 91-02-24. Accepted 91-05-26

The etiology of idiopathic scoliosis is multifactorial, with several reports indicating that a primary dysfunction of the brainstem is one contributing factor (Nachemson and Sahlstrand 1977, Yamada et al. 1984, Wyatt et al. 1986). Scoliosis is also associated with idiopathic syringomyelia (Woods and Pimenta 1944, McRae and Standen 1964, Huebert and McKinnon 1969, Williams 1979), as well as with congenital malformations of the brainstem (Riley and Swift 1979, Dretakis 1984, Samuelsson and Eklöf 1988). An accurate diagnosis of a nonidiopathic etiology of the curve allows treatment to be directed to the underlying cause, as well as to the spinal deformity. For this reason a neurogenic cause of a scoliosis should always be considered before it is classified as idiopathic. Magnetic resonance (MR) imaging fulfills the needs for a safe and noninvasive evaluation of the spinal cord and brain in children presenting with scoliosis (Hawkes et al. 1983, Norman et al. 1983, Yeates et al. 1983, Samuelsson et al. 1987).

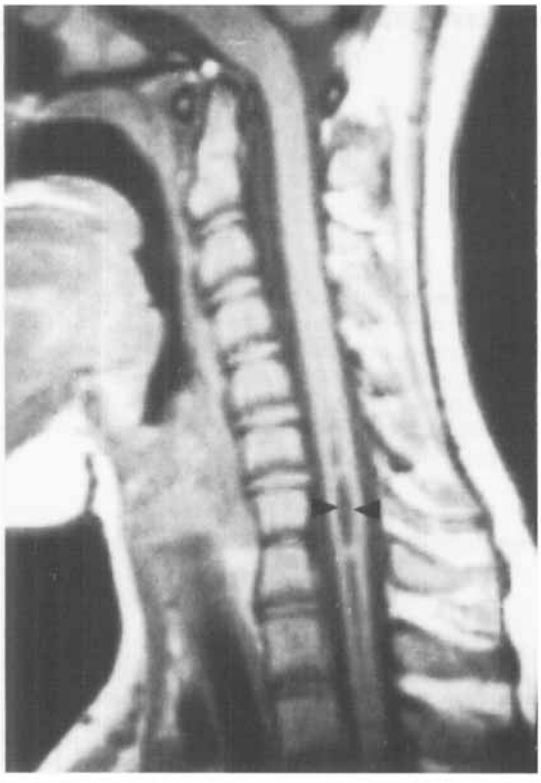
We have studied whether or not patients classified as having idiopathic scoliosis in reality have an associated syringomyelia or Chiari I malformations, or both.

Patients and methods

Totally, 22 females and 4 males classified as having idiopathic scoliosis—mean age 12 (8–16) years and with a mean Cobb angle of 27° (10°–44°) at the first examination—were studied with MR imaging of the brainstem and spinal cord. All the patients were referred for orthopedic assessment after regular school screening for scoliosis. None of them had a neurologic abnormality upon clinical examination. Their curves were thoracic in 16 cases, thoracolumbar in 6, lumbar in 2, and double curves in 2. Twenty-one patients had been braced, whereas 5 had not been braced. The magnitude of the scoliosis was measured with the Cobb method (1948). MR was performed using a 1.0 tesla superconducting magnet (Siemens Magnetom) using a spin-echo technique with T1-weighted images (TR = 600 msec and TE = 15 msec). The brainstem and cervical cord were examined with 15 sagittal scans of 4-mm thickness. The dorsal and lumbar spinal cord were examined with 34 continuous axial scans of 10-mm thickness. Additional axial images of the cervical spinal cord and sagittal images of the thoracic spinal cord were obtained using the same technique when a syrinx was found. No sedation was necessary, and the examination time for each patient averaged 1 hour. We used a historic material of MR images of the normal anatomy of the brain and spinal cord of children of the same age for comparison (Samuelsson et al. 1987).

Results

Two patients had syringomyelia. One syrinx was located at the C6–C8 level, and was associated with a right thoracic scoliosis of 27° (Figure 1). The other



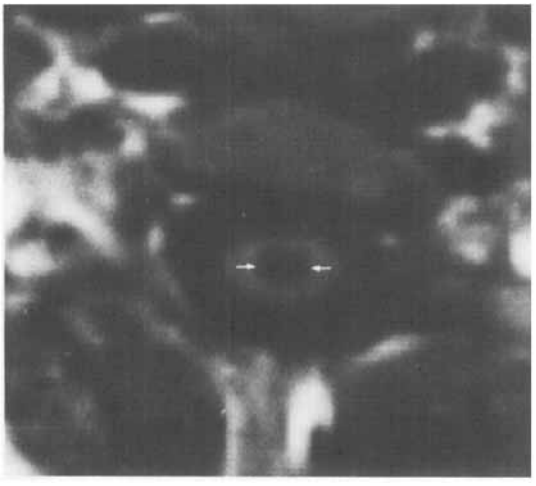
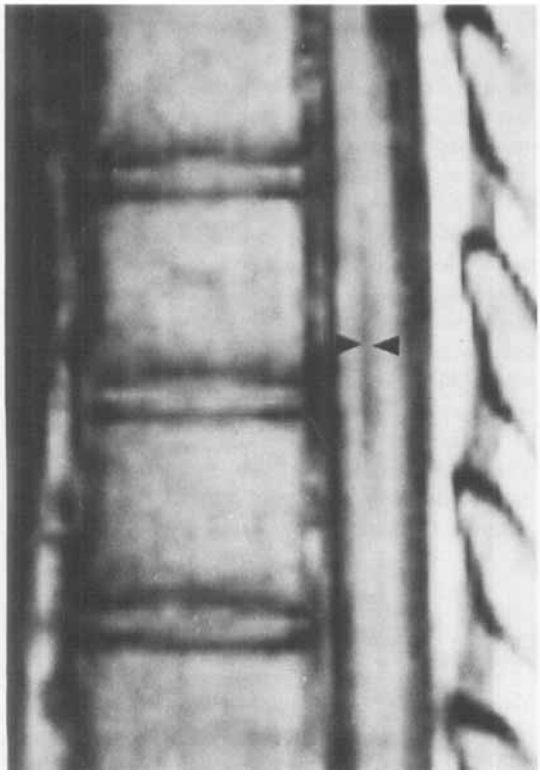


Figure 1. A 16-year-old girl with scoliosis and syringomyelia. Midline sagittal and axial MR images of syrinx located at C6–C8.



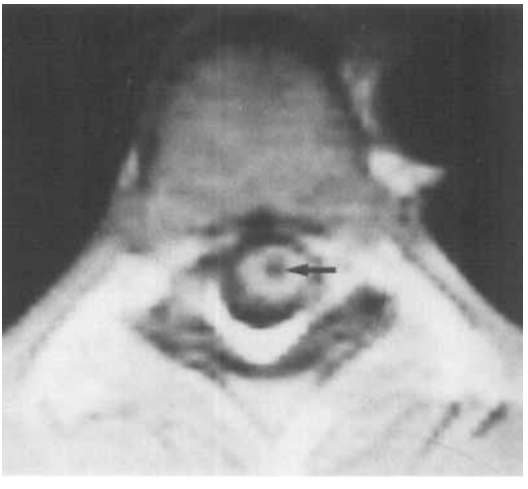


Figure 2. A 14-year-old girl with scoliosis and syringomyelia. Midline sagittal and axial MR images of syrinx located at T6–T8.

syrinx was located at T6–T8, and was associated with a left lumbar curve of 32° (Figure 2). Both were centrally located, and on sagittal images somewhat irregular in shape. The spinal cords were more or less displaced towards the concave side of the spinal canal depending on the magnitude of the scoliosis. The brainstem was normal in all the patients, but there was a wide variation in size and position of the cerebellar tonsils (Figures 3 and 4). The lowest position was 5 mm below the foramen magnum, which was seen in 2 patients. The shape of the tonsils was normal and not peg-like as in Chiari I malformations.

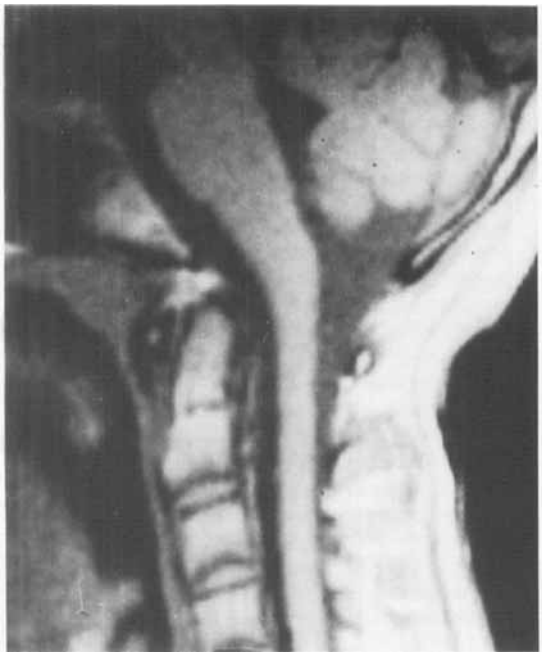


Figure 3. A 14-year-old girl with idiopathic scoliosis. MR image showing a high position of the cerebellar tonsils.

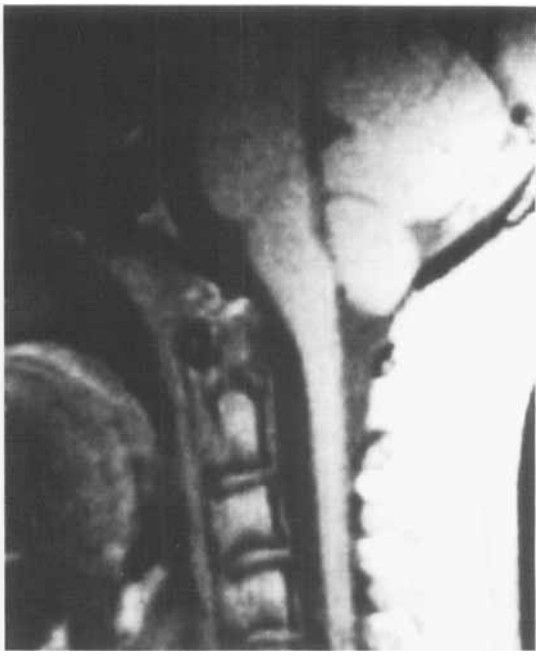


Figure 4. A 14-year-old girl with idiopathic scoliosis. MR image showing a low position of the cerebellar tonsils.

Discussion

MR has become the best method for spinal cord and brainstem imaging (Norman et al. 1983), replacing metrizamide myelography (Pettersson and Harwood-Nash 1982). We preferred sagittal images to assess the form of the brainstem and position of the cerebellar tonsils. These permitted a better view of the gross anatomy of this region and were not as sensitive to variations in positioning of the head and neck during examination as axial scans (Geissle et al. 1989). On the contrary, when examining the spinal cord, axial images show, according to our opinion, the size and extent of a syrinx best, especially when scoliosis is severe (Samuelsson et al. 1987). It is also easier to obtain comparable images on serial examinations of an expanding syrinx with axial scans.

Scoliosis may be the first sign of syringomyelia (Baker and Dove 1983, Dure et al. 1989), and the patient is often first seen by the orthopedic surgeon (Williams 1979, Isu et al. 1990). A cervical or high thoracic double curve (Riseborough and Herndon 1975), left convex thoracic (Coonrad et al. 1985, Phillips et al. 1990), or painful curve (Williams 1979) should incite suspicion. Other presenting features may be of a very insidious nature, and the findings on neurologic examination can vary greatly (Schliep 1978). To our knowledge, there are no reports on the prevalence of syringomyelia without associated neurologic signs or symptoms in patients with scoliosis. One cause may be that the lack of neurologic findings or vague symptoms has not warranted invasive examinations of the spinal canal, like metrizamide myelography, which was used before MR imaging. The high prevalence of CNS lesions, including syringomyelia associated with scoliosis reported by Nokes et al. (1987), is explained by their referral base being selected patients examined before planned surgical intervention. The same holds true for Isobe (1988), who reported 6 patients with syringomyelia out of 33 patients with idiopathic scoliosis who had undergone a preoperative examination with metrizamide myelography.

The fact that we disclosed 2 patients with syringomyelia in a group of 26 adolescents treated for idiopathic scoliosis indicates that scoliosis associated with a syrinx is more common than previously known. This finding is also supported by occasional reports like the one of Nordvall and Wikkelsø (1979) on neurologic complications of scoliosis surgery owing to previously unknown syringomyelia in patients operated on for idiopathic scoliosis.

Both our syrinxes were small, and there were no signs of neurologic deficits. Grant et al. (1987) and Vaquero et al. (1990) reported that symptoms attributed to spinal-cord damage have no relationship to the size of the syrinx on the MR image. It is also well

known that neurologic findings of syringomyelia are rare in young individuals, and the diagnosis is normally not apparent until after 20 years of age (Schliep 1978). There is, however, evidence that the size and extension of a syrinx may be correlated with a neurologic deficit. Jabbari et al. (1990) showed that pathologic somatosensory-evoked potentials correlated with the size of the syrinx. Isu et al. (1990) reported a correlation between neurologic findings and eccentric extension of the syrinx.

Our present knowledge of the natural history of scoliosis associated with syringomyelia is poor. Improved MR technology and increasing number of MR units will facilitate prospective studies with serial neurologic and MR examinations to elucidate the normal history of scoliosis associated with syringomyelia.

Acknowledgement

We gratefully acknowledge the financial support from HKH Kronprinsessan Lovisas förening för barnsjukvård/Axel Thielmans minnesfond, Åke Wibergs stiftelse, Örebro County Council, and Svenska Läkarsällskapet. We would also like to express our thanks to Britt-Louise Eriksson and Ulf Degrell for assistance with the MR examinations.

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