

MRI OF 'IDIOPATHIC' JUVENILE SCOLIOSIS

A PROSPECTIVE STUDY

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In a prospective trial we performed MRI of the spine and hind brain in 31 patients with scoliosis of onset between the ages of four and 12 years.

In eight patients (26%) there was a significant neuroanatomical abnormality; there were six cases of Chiari-1 malformation associated with a syrinx, one isolated Chiari-1 malformation and one astrocytoma of the cervical spine. Four of these patients had left-sided curves.

There were no clinical features which could reliably identify those patients with abnormalities on MRI. In particular, the unilateral absence of abdominal reflexes was found to be non-specific (1 of 8 of patients with neuroanatomical abnormalities (12.5%) v 2 of 23 with normal scans (8.7%).

In view of the established risks of surgical correction of scoliosis in the presence of undecompressed syringomyelia and the possible improvement that may follow decompression of the foramen magnum, we feel that MRI of all patients with scoliosis of juvenile onset should be obligatory.

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In 1954 James classified idiopathic scoliosis into three groups according to the age of onset: infantile, juvenile and adolescent. In his study the juvenile group included only 16 of 134 thoracic curves. Although the age criteria for classification vary between different studies, juvenile scoliosis remains relatively uncommon (Tolo and Gillespie 1978; Dickson 1985). Some juvenile cases result from undiagnosed infantile curves and, conversely, a proportion of adolescent scolioses will represent previously undetected juvenile cases. Doubt over the true significance of the juvenile group led Dickson (1985) to use the original classification of Ponseti and Friedman (1950) into early and late onset, with five years as the dividing point.

The association of scoliosis with syringomyelia is well established (Woods and Pimenta 1944; McRae and Standen 1966). In the past the identification of abnormal neurological signs prompted further, usually invasive, investigation. Baker and Dove (1983) were the first to report scoliosis as the presenting sign of otherwise asymptomatic syringomyelia and this has since been confirmed by others (Raininko 1986; Isu et al 1992; Lewonowski, King and Nelson 1992). With the advent of MRI, sporadic accounts of syringomyelia, often associated with the Chiari-1 malformation, have identified a preponderance within the juvenile period (Arai et al 1992).

Zadeh et al (1995) found ten cases of syringomyelia in 12 patients with idiopathic scoliosis who had been selected for MRI after the identification of asymmetrical or absent superficial abdominal reflexes. This review, and that of Schwend et al (1995), emphasised the need for a prospective study of MRI in idiopathic scoliosis and we have performed such a study on the juvenile age group.

PATIENTS AND METHODS

We performed a prospective study of 31 consecutive patients who presented with idiopathic scoliosis first detected between the ages of four and 12 years inclusive. All patients were assessed clinically with a full history and examination including neurological evaluation.

There were 24 girls and seven boys. The mean age at which scoliosis was first noted was 8.6 years (6.6 years for boys and 9.1 years for girls; range 4.0 to 12.5; Fig. 1).

All patients had MRI with a 1.5 Tesla Siemens Magnetom 63SP system (Siemens, Erlangen, Germany). Images of the whole spine were produced by a linearly-polarised

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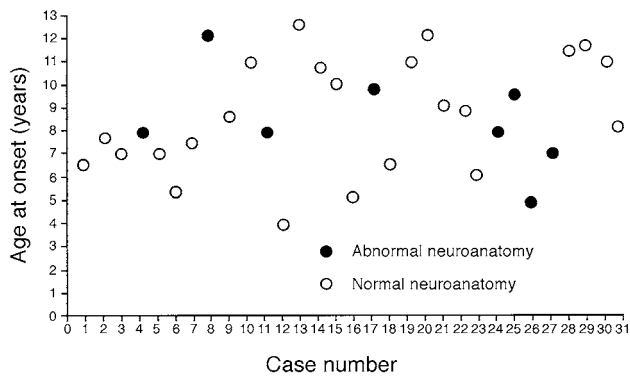


Fig. 1

Scattergram showing the age at onset of scoliosis in 31 patients.

cervical spine coil for the cervical spine and a four-element phased array spine coil for the thoracic and lumbar spine. A combination of sagittal, coronal and axial T1-weighted scans was made in all patients. Those in whom a syrinx was found without an associated Chiari malformation were also examined after intravenous injection of gadolinium-DPTA (Magnevist; Schering, Burgess Hill, UK) to exclude an underlying neoplasm of the cord. These scans were prospectively studied by experienced radiologists. The hind brain and spinal cord were examined for syrinx formation, features of spinal dysraphism and cord neoplasms.

RESULTS

There were 19 single and 12 double curves (Fig. 2). The thoracic curves were right-sided in 24 and left-sided in 7. The Cobb angles of the major curves at presentation ranged from 10° to 100° with a mean of 40°.

In eight patients a considerable neuroanatomical abnormality of the hind brain or spinal cord was identified; there were six cases of Chiari-1 malformation associated with a syrinx, one of an isolated Chiari-1 malformation and one astrocytoma with a syrinx. In four of these patients the thoracic curve was left-sided, whereas this was seen in only three of the 23 patients with normal neuroanatomy ($p = 0.05$).

There was no significant difference in the mean age of onset between the groups with and without neuroanatomical abnormalities (8.3 years and 8.7 years, respectively). The mean curve at presentation was 45° in the group with abnormal neuroanatomy and 40° in the normal group. The proportion of abnormal neuroanatomical findings was similar for both sexes (6 girls and 2 boys, 25% and 29%, respectively).

At presentation only two of the eight patients with neuroanatomical abnormalities had neurological signs. These were marked in one and included nystagmus and ataxia; in the other there was unilateral absence of the abdominal reflex. Both patients had a syrinx with a Chiari-1 malformation. Two patients with normal neuroanatomy had

unilateral absence of the abdominal reflex.

Twenty patients were observed for at least six months before surgery. They had a mean progression of the curve of 7.6°/year (-10 to 44). For the five patients with neuroanatomical abnormalities who were observed for this period the mean progression was 9.8°/year and for the 15 normal cases 6.9°/year.

All six patients with a Chiari malformation and syrinx had decompression of the foramen magnum (Fig. 3). Three with curves greater than 50° at the time of neurosurgery subsequently had two-stage spinal fusion. One of the three remaining patients showed an improvement from 33° to 19° over nine months after decompression. Follow-up of the last two patients, with curves of 28° and 44° respectively, has been too short to make a useful observation.

The patient with an astrocytoma and syrinx had a biopsy and radiotherapy. The patient with an isolated Chiari-1 malformation had a two-stage fusion 47 months after presentation.

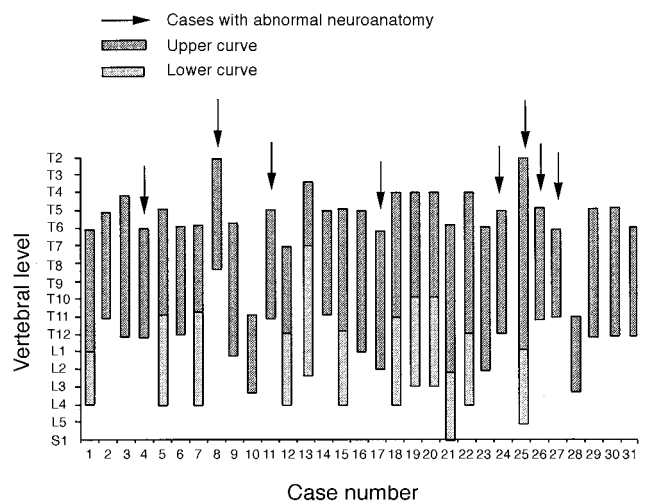


Fig. 2

Chart showing the site of scoliosis in 31 patients.

Of the 23 patients with normal neuroanatomy six were managed by simple observation, four had bracing and 13 had fusion (1 anterior, 1 posterior and the remainder two-stage).

DISCUSSION

We have used MRI prospectively to screen a consecutive group of 31 children with idiopathic scoliosis of juvenile onset. We found abnormalities of the hind brain or cord in eight (26%). Since the clinical presentation did not discriminate between those with and without CNS abnormalities, the underlying abnormalities would have remained undiagnosed without the routine use of MRI.

The unilateral absence of abdominal reflexes did not

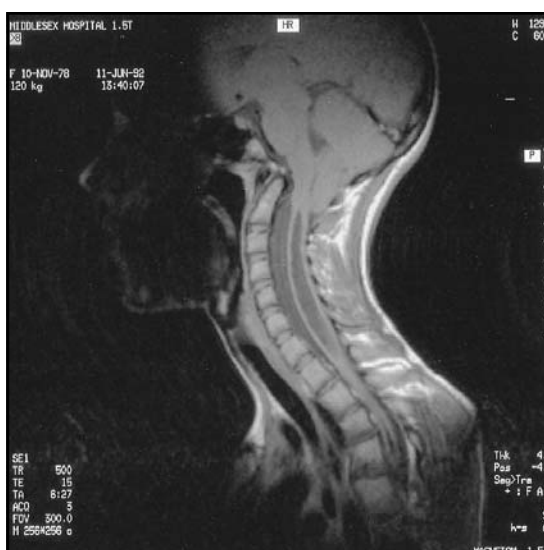


Fig. 3a



Fig. 3b

MRI showing a Chiari malformation associated with a large syrinx in the cervical and thoracic cord before decompression of the foramen magnum (a) and eight months after decompression (b) with reduction in the size of the syrinx.

prove to be a specific finding, being identified in one of eight patients with neuroanatomical abnormalities as against two of 23 with normal neuroanatomy. This appears to conflict with Zadeh et al (1995) who concluded that "an absent superficial abdominal reflex on the same side as the convexity of the curve is an early and sensitive indicator of underlying syringomyelia". The patients whom they report, however, were selected for scanning on the basis of abnormal abdominal reflexes; no comment can be made on the incidence of syringomyelia in patients who had not been scanned. The age distribution of their patients was also younger and we doubt the significance of this reflex when it is absent bilaterally rather than unilaterally.

We found that 50% of the neuroanatomical abnormalities were associated with left-sided curves. In other reviews of syringomyelia Isu et al (1992) and Muhonen et al (1992) had similar findings of approximately 50%, but Schwend et al (1995) found a greater association with left-sided curves (77%). If MRI is done solely on the basis of the direction of the scoliosis then half of all such abnormalities will be missed.

Although only one of our four patients managed by decompression of the foramen magnum has so far shown a reduction of the curve, several reports have indicated either arrest or improvement of the scoliosis after this procedure (Dure et al 1989; Phillips, Hensinger and Kling 1990; Muhonen et al 1992). Early detection of a syrinx by prompt MRI may therefore be of particular value.

The risk of neurological injury during instrumented correction of scoliosis without prior decompression of an associated syrinx has already been documented (Huebert and MacKinnon 1969; Diaz and Lockhart 1987; Noordeen,

Taylor and Edgar 1994). The cause is uncertain, but sensitivity to direct trauma, a tenuous blood supply in the presence of an expanded cord and changes in the pressure of the CSF have all been suggested (Phillips et al 1990).

The identification of a spinal tumour lends weight to the value of MRI. There have been several reports of scoliosis as the presenting feature of an intramedullary cord tumour without neurological signs or symptoms (Allen and Kahn 1933; Dalloz et al 1963; Citron et al 1984). Early detection and treatment of our patient allowed the preservation of complete neurological function.

In a prospective study of routine MRI of 'idiopathic' scoliosis in 26 patients of all ages Samuelsson, Lindell and Kogler (1991) identified two with syrinxes and concluded that "MR should be mandatory before bracing or operative correction of scoliosis". In our study we have failed to identify any clinical features which may reliably distinguish affected cases. For all new patients with scoliosis the cost of routine MRI may be prohibitive in many units, but since we have found significant neuroanatomical abnormalities in 26% of curves of juvenile onset, we feel that in this age group MRI should be obligatory.

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