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# PREVALENCE OF NEURAL AXIS ABNORMALITIES IN PATIENTS WITH INFANTILE IDIOPATHIC SCOLIOSIS

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**Background:** Although reports in the literature have demonstrated an approximately 20% prevalence of neural axis abnormalities in patients with juvenile idiopathic scoliosis who have a curve of  $>20^\circ$ , the prevalence of neural axis abnormalities in patients with infantile idiopathic scoliosis is not well documented. In two previous studies involving a total of only ten patients with infantile idiopathic scoliosis, five patients were noted to have a neural axis abnormality on magnetic resonance images.

**Methods:** The records of forty-six consecutive patients who were seen between 1992 and 2000 at three spinal deformity clinics were retrospectively reviewed. The inclusion criteria included presumed idiopathic scoliosis at the time of presentation, an age of three years or less, a curve magnitude of  $\geq 20^\circ$ , normal neurological findings, no associated syndromes, and no congenital abnormalities. All patients were evaluated with a total spine magnetic resonance imaging protocol for examination of neural axis abnormalities from the skull to the coccyx.

**Results:** Ten (21.7%) of the forty-six patients were found to have a neural axis abnormality on magnetic resonance imaging. This group included five patients with an Arnold-Chiari malformation and an associated cervicothoracic syrinx, three with syringomyelia, one with a low-lying conus, and one with a brainstem tumor. Eight of these ten patients needed neurosurgical intervention for treatment of the abnormality.

**Conclusions:** The 21.7% prevalence of neural axis abnormalities in this group of patients with infantile idiopathic scoliosis was found to be almost identical to that reported in the literature on patients with juvenile idiopathic scoliosis. Because of the high prevalence of abnormalities and the fact that eight of the ten patients with abnormal findings on magnetic resonance images required neurosurgical intervention, a total spine magnetic resonance imaging evaluation at the time of presentation is recommended for all patients with infantile idiopathic scoliosis who have a curve measuring  $\geq 20^\circ$ .

The evaluation and treatment of patients with infantile-onset idiopathic scoliosis is a challenging problem. When evaluating the spinal deformity, the treating physician must be aware that scoliosis in children can be the presenting sign of an asymptomatic neural axis abnormality<sup>1,4</sup>. If the neural axis abnormality remains undetected, there is a risk of neurological sequelae resulting from the use of instrumentation for correction of the scoliosis<sup>5-7</sup>. Of additional concern is the reported need for neurosurgical intervention between the time of birth and the age of ten years in  $>50\%$  of patients with idiopathic scoliosis who have neural axis abnormalities on magnetic resonance imaging<sup>1,8,9</sup>. However, those studies included only a very small number of patients in the infantile age-group (birth to the age of three years).

Although reports in the literature have demonstrated an approximately 20% prevalence of neural axis abnormali-

ties in patients with juvenile idiopathic scoliosis who have a curve of  $>20^\circ$ <sup>1,8,9</sup>, the prevalence of neural axis abnormalities in patients with infantile idiopathic scoliosis is not well documented. In the study by Gupta et al.<sup>1</sup>, three of six infants with scoliosis and normal neurological findings were found to have neural axis abnormalities on magnetic resonance images. In a similar study by Lewonowski et al.<sup>8</sup>, two of four infants with scoliosis and normal neurological findings were found to have abnormalities on magnetic resonance images. However, the sample sizes in those studies were small, and without more data it is difficult to make recommendations regarding the need for routine use of magnetic resonance imaging for patients in the infantile age-group. Informed decision-making in this age-group is even more critical because the performance of a screening magnetic resonance imaging examination in these young children often involves the risks

associated with intravenous sedation or general anesthesia.

The purposes of the present study were to determine the prevalence of neural axis abnormalities in a large multicenter cohort of patients with idiopathic infantile scoliosis who had normal neurologic findings on history and physical examination and to report on the clinical importance of the identified neural axis abnormalities as indicated by the need for further neurosurgical intervention.

### Materials and Methods

The databases at three spinal deformity clinics were retrospectively reviewed to identify all patients with infantile-onset scoliosis who had been seen between 1992 and 2000. The inclusion criteria were presumed idiopathic scoliosis at the time of presentation, an age of three years or less at the time of diagnosis, a curve magnitude of  $\geq 20^\circ$  as measured on a radiograph made with the patient supine (for infants) or standing (for patients who were old enough to stand without support), normal findings on a neurological examination, no associated syndromes, and the absence of congenital spinal abnormalities. Forty-six consecutive patients met the criteria for inclusion in the study.

The neurological examination, which was performed by a pediatric and/or spinal orthopaedic surgeon, consisted of an evaluation of the motor, sensory, and reflex function of the upper and lower extremities as well as an evaluation of the abdominal reflex. Anteroposterior and lateral radiographs of the entire spine were made at the time of presentation to rule out congenital anomalies and to allow for measurement of the Cobb angle. All patients with a curve of  $\geq 20^\circ$  underwent a total spine magnetic resonance imaging protocol for examination of neural axis abnormalities from the skull to the coccyx. If a neural axis abnormality was detected, the patient was referred for neurosurgical evaluation, treatment, and follow-up. All patients were followed continually by the pediatric deformity service at regular intervals.

### Total Spine Magnetic Resonance Imaging Protocol

The magnetic resonance imaging examinations were done in the same manner at all three institutions. T1 and T2-weighted sagittal screening images of the cervical, thoracic, and lumbosacral spine were made for all patients. Additional sagittal and axial screening images of the craniocervical junction, cervicothoracic junction, thoracolumbar junction, lumbosacral junction, and the area of major deformity were also made. The studies were reviewed to check for all neural axis abnormalities, including Arnold-Chiari malformation, syringomyelia, hydromyelia, low-lying conus, tethered cord, and neoplasm. All studies were reviewed by an attending pediatric neuroradiologist as well as by the attending spine or pediatric orthopaedic surgeon.

### Statistical Analysis

Descriptive statistics were calculated with use of the combined data from all three centers. Odds ratios were used to describe the association between the gender of the patient, the direc-

tion of the curve, the apex of the curve (T10/11 and above compared with T11/12 and below) and the prevalence of abnormal magnetic resonance imaging findings. The chi-square test for trend was used to evaluate the relationship between a more caudad apex and abnormal magnetic resonance imaging findings. T tests were used to test for differences with regard to average age and average Cobb angle between patients with normal and abnormal findings. The level of alpha was set at 0.05.

### Results

Forty-six consecutive patients were reviewed. The female-to-male ratio was 1.3:1, with twenty-six girls and twenty boys. The mean age at the time of presentation was seventeen months (range, two to thirty-seven months). The mean Cobb angle at the time of presentation was  $40^\circ$  (range,  $20^\circ$  to  $90^\circ$ ). In thirty-three patients the apex of the curve was in the thoracic spine (cephalad to the level between the eleventh and twelfth thoracic vertebrae), whereas in thirteen the apex was in the thoracolumbar-lumbar spine (at or caudad to the twelfth thoracic level). Of the main thoracic curves, 52% were right-sided (that is, the apex was to the right) and 48% were left-sided.

Ten (21.7%) of the forty-six patients had abnormal findings on magnetic resonance imaging. According to the Student t test, there was no difference between the patients who had abnormal findings on magnetic resonance imaging and those who had normal findings with regard to age ( $p = 0.95$ ) or the Cobb angle ( $p = 0.57$ ). The average age for patients with normal findings on magnetic resonance imaging was  $17.23 \pm 10.45$  months, compared with  $17.56 \pm 12.17$  months for those with abnormal findings ( $p < 0.95$ ). With regard to the direction of the curve, 26% of the right-sided curves were associated with abnormal findings on magnetic resonance imaging compared with 18% of the left-sided curves (odds ratio, 1.88; 95% confidence interval, 0.40 to 8.82). Of the patients with abnormal findings on magnetic resonance imaging, 17% were female and 28% were male (odds ratio, 0.52; 95% confidence interval, 0.12 to 2.30).

Of the ten patients with abnormal findings on magnetic resonance imaging, five had an Arnold-Chiari Type-I malformation and an associated cervicothoracic syrinx, three had syringomyelia alone, one had a low-lying conus, and one had a brainstem tumor. Eight of these ten patients needed neurosurgical intervention for treatment of the abnormality.

In the group of five patients with an Arnold-Chiari Type-I malformation, the mean age at the time of presentation was twenty months (range, five to thirty-four months). The mean Cobb angle at the time of presentation was  $37^\circ$  (range,  $20^\circ$  to  $54^\circ$ ). All five patients had a main thoracic curve; three of the curves were right-sided and two were left-sided (Figs. 1-A and 1-B). All five patients underwent an initial decompression of the posterior fossa. In four of the five patients, the associated syrinx had to be surgically addressed either at the time of decompression of the posterior fossa or at a later time because of progression of the syrinx or the development of neurological findings. Three of the five patients eventually

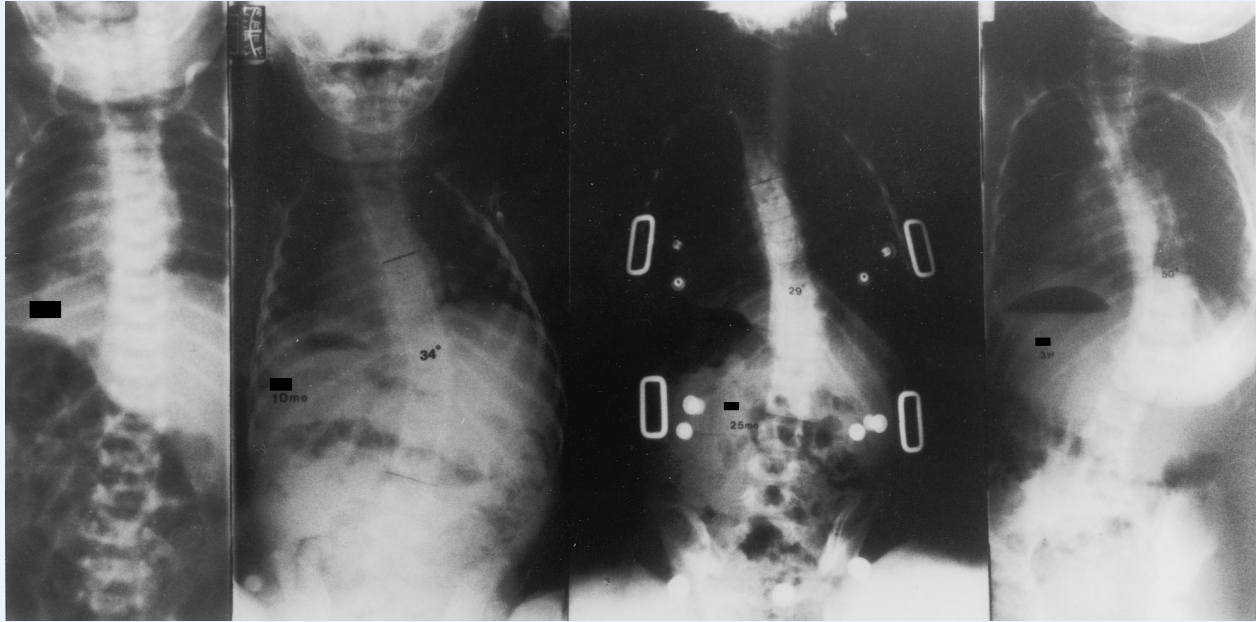
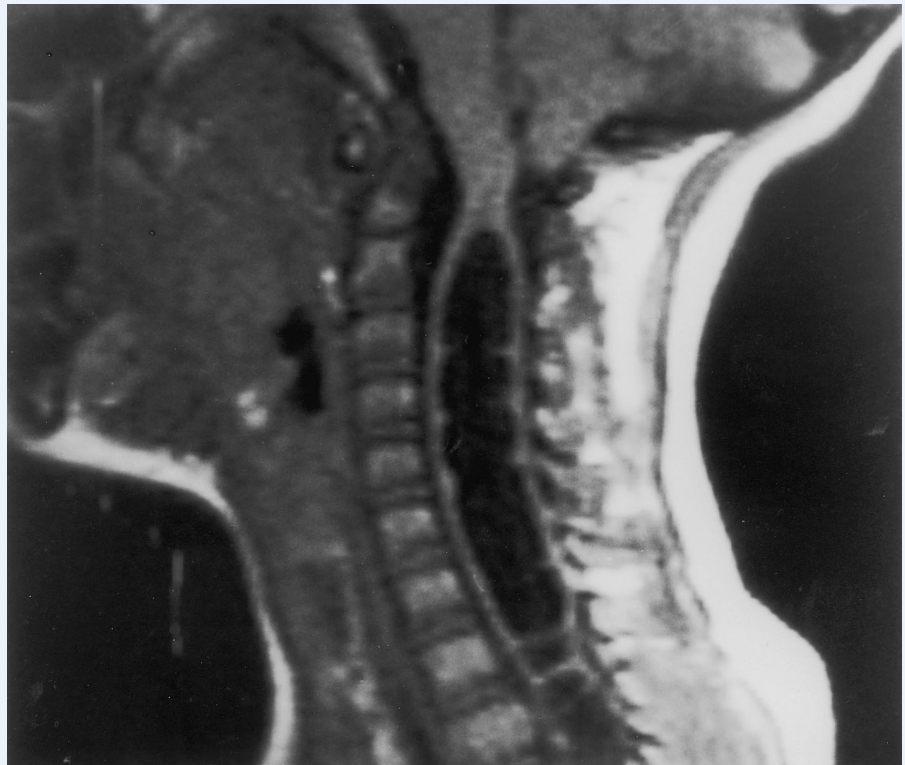


Fig. 1-A

Serial radiographs, made between the ages of five months and three years, of a girl who presented with scoliosis and an otherwise normal history and examination, showing rapid progression of a right thoracic curve despite treatment with a brace.

Fig. 1-B

Magnetic resonance image, made at the age of three years, demonstrating an Arnold-Chiari Type-I malformation and syringomyelia.



underwent spinal arthrodesis because of continued progression of the curve.

Of the three patients who had syringomyelia without other abnormalities, one had a cervicothoracic syrinx and was managed with observation only whereas the other two had a large thoracic syrinx (involving the entire diameter of the cord) and required surgical decompression and/or shunting. The mean age at the time of presentation was ten months (four, eleven, and fourteen months). The mean Cobb angle at the time of presentation was 41° (28°, 34°, and 60°). In two of the three patients, the scoliosis was successfully managed with a brace. The third patient underwent spinal arthrodesis because of progression of the curve.

The patient with a low-lying conus presented at the age of eighteen months with a right lumbar curve of 32°. The conus, which ended at the third lumbar level, was detected with use of magnetic resonance imaging. Normally, the conus lies opposite or cephalad to the disc space between the first and second lumbar vertebrae by one to two months after birth<sup>10-12</sup>. The patient was successfully treated with a brace without curve progression and did not require neurosurgical intervention.

The patient with a brainstem tumor presented at the age of thirty-four months with a left lumbar curve of 45°. A mass in the medulla was noted on magnetic resonance imaging. A diagnostic biopsy demonstrated a low-grade astrocytoma, and treatment consisted of partial surgical resection of the tumor followed by radiation therapy. The patient subsequently underwent anterior-posterior spinal arthrodesis from the ninth thoracic to the third lumbar level at the age of six years.

## Discussion

The results of the present large multicentered study demonstrated a 21.7% prevalence of neural axis abnormalities in otherwise asymptomatic patients with infantile idiopathic scoliosis. The high percentage of neural axis abnormalities in our patients is noteworthy because eight of the ten patients who had such abnormalities required neurosurgical intervention, compared with 50% of juvenile patients with neural axis abnormalities as reported in the literature<sup>1,8</sup>. Similarly, in the study by Gupta et al.<sup>1</sup>, two of the three infantile patients with neural axis abnormalities required neurosurgical intervention. Two of those patients had a Chiari Type-I malformation associated with a cervicothoracic syrinx, and the third patient had diffuse dural ectasia. All three of those patients were being treated with a brace for correction of the curve at the time of the study. In the study by Lewonowski et al.<sup>8</sup>, both patients with neural axis abnormalities required neurosurgical intervention to treat a Chiari Type-I malformation and an associated syrinx.

An infant with a suspected spinal deformity should be carefully examined for the presence of scoliosis as well as associated abnormalities of the head, neck, pelvis, and hips. Once the diagnosis of scoliosis is made, the differential diagnosis in this age-group includes congenital scoliosis, scoliosis of neuromuscular origin, scoliosis secondary to intraspinal pathology, scoliosis in association with a genetic syndrome (e.g.,

neurofibromatosis), and idiopathic scoliosis. The diagnosis of idiopathic scoliosis is one of exclusion. A history should be obtained with regard to birth weight and developmental milestones. A complete physical examination, including a careful neurologic examination, is imperative. For instance, the loss of abdominal reflexes or an absent gag reflex is sometimes the only clinical finding in a patient with scoliosis associated with a syrinx or a Chiari malformation<sup>13-18</sup>. The presence of plagiocephaly or congenital abnormalities of the extremities should be noted. Any associated condition warrants appropriate evaluation and treatment<sup>19</sup>.

Anteroposterior and lateral radiographs of the entire spine should be made at the initial visit to evaluate the Cobb angle and to rule out congenital vertebral anomalies. In addition, the lumbosacral junction and the hip joints should be visualized on the radiographs to rule out congenital anomalies and developmental hip dysplasia<sup>19</sup>.

It is likely that a substantial number of infants, like juvenile patients, who are diagnosed with idiopathic scoliosis have subtle neurological findings that should lead the treating physician to perform a further diagnostic workup<sup>1,8,19</sup>. Interestingly, the 21.7% prevalence of neural axis abnormalities in the present study of patients with infantile idiopathic scoliosis mirrors the reported prevalence in studies of patients with juvenile idiopathic scoliosis (range, 17.6% to 26%)<sup>1,8,9</sup>. In addition, the percentage of infantile patients in the present study who had neural axis abnormalities that required neurosurgical intervention is similar to that reported for the juvenile population<sup>1,3,9</sup>.

In the present study, which we believe to be the largest multicenter study of patients with infantile idiopathic scoliosis to date, there was no association between abnormal findings on magnetic resonance images and gender, curve magnitude, curve location, or curve direction. Unfortunately, these findings do not help to identify a particular subset of patients who are at risk. However, because of the 21.7% prevalence of abnormal findings on magnetic resonance images and the fact that eight of the ten patients with abnormal findings required neurosurgical intervention, we recommend that magnetic resonance imaging of the neural axis be performed at the time of presentation for all patients with infantile scoliosis who have a curve measuring  $\geq 20^\circ$ , even if the findings of a neurological examination are normal. ■

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