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ORIGINAL ARTICLE

Are intraspinal anomalies in early onset idiopathic scoliosis as common as once thought? A two centre United Kingdom study

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Abstract
Purpose Rates of neural axis abnormalities in infantile and juvenile idiopathic scoliosis may be as high as 50 %. We aimed to determine the rates of neural axis abnormalities in early onset idiopathic scoliosis patients in a British population.
Methods This retrospective study at two British spinal deformity clinics identified 72 patients satisfying the inclusion criteria of: (1) age at diagnosis of 7 years and younger, (2) idiopathic scoliosis and (3) magnetic resonance imaging of the neuraxis.
Results The mean age at diagnosis was 3.6 years and the mean Cobb angle was 47° with a near equal distribution of left (32) and right (36) sided curves. Eight (11.1 %) neural axis abnormalities consisting of two syrinxes, one Arnold–chiari Type I malformation and five combined (Arnold–chiari malformation Type I and syrinx) anomalies were identified.
Conclusions This multi-centre study on the largest number of British subjects to date helps to establish the rates of neural axis abnormalities.

Keywords Early onset idiopathic scoliosis · Intraspinal anomalies · UK

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Introduction

Previous studies investigating the rates of intraspinal anomalies in early onset idiopathic scoliosis (EOIS)—i.e. inclusive of infantile (IIS) and juvenile (JIS) idiopathic scoliosis—have determined values of up to 50 % [1–7]. This has lead to the recommendation for routine whole spine magnetic resonance imaging (MRI) screening of this condition [1].

Population studies in the United States of America (USA) have found that EOIS patients (defined in the USA as age less than 11 years) compose only 8 % of the idiopathic scoliosis population [8]. Several European studies found IIS to be more common in Europe (12.8 %) compared to North America (0.25 %) [8–12]. In the UK, up to 50 % of patients with idiopathic scoliosis are reported to have the early onset form (defined as less than 8 years of age [8]).

There are no appropriate age group studies conducted in the UK on the rates of intraspinal anomalies in EOIS. We aimed to investigate the rates of intraspinal anomalies in EOIS in a population of British patients. Given that the British and American populations with idiopathic scoliosis appear to vary in terms of age of onset, we hypothesised that the rates of spinal anomalies were also different.

Materials and methods

Our aim was to determine the rates of intraspinal abnormalities in patients diagnosed with presumed idiopathic scoliosis at 7 years of age or less. Following approval from the local research and development committees, we retrospectively reviewed medical records and radiographs for all patients with EOIS who had undergone MRI screening

Table 1 Eight cases (11.1 %) of neural axis abnormalities found in 72 patients

Age of onset (months)	Age of 1st clinic attendance (months)	Cobb angle (°)	Curve sidedness	Neural axis anomaly		Clinical findings	Management
				Chiari	Syrinx		
14	31	60	Right	–	Conus		Growing rod
36	39	32	Left	I	C4–T1		PFD
72	74	38	Right	I	C2–L1		Brace
73	76	34	Left	I	C1–L2	AAR	PFD
78	81	38	Left	I	C6–T6	AAR, FH	PFD
82	84	88	Left	–	Conus		Growing rod
87	89	20	Right	I	–	BP	Fusion
89	93	56	Right	I	C4–T9	AAR	PFD

AAR asymmetrical abdominal reflexes, *FH* family history, *BP* back pain, *PFD* posterior fossa decompression

of the neuraxis. The inclusion criteria were: (1) age of onset before 8 years of age, (2) initial (pre-MRI) diagnosis of idiopathic scoliosis, and (3) underwent screening MRI of the full neural axis. Data were collected from two separate tertiary referral centres in the UK between January 1994 and December 2010. Patients' notes were reviewed to determine the age of onset, age at first presentation, presenting history (family history, back pain, neurology, syndromic abnormalities), Cobb angle, curve sidedness, results of total spine MRI and subsequent management. Radiographs were analysed and Cobb angles measured by consultant spine surgeons and full neuraxis MRIs were further reviewed by consultant radiologists.

Data were statistically analysed to compute the Spearman correlation coefficient to investigate relationships, and the Mann–Whitney test was used to compare means for independent samples. The Fisher exact probability test (φ) was used to compare data from patients with and without neural axis abnormalities. *P* values less than or equal to 0.05 were considered to be significant.

Results

In the 16 years covered, we identified 72 patient records satisfying the inclusion criteria. The average age at diagnosis was 3 years and 7 months (range 1 month to 7 years and 11 months), and the average age at first attendance at a scoliosis clinic was 4 years and 4 months (range 8 months to 9 years and 10 months). The average Cobb angle was 46.6° (range 10°–118°). There were 36 (52.9 %) right-sided and 32 (47.1 %) left-sided curves.

In the study group, eight (11.1 %) patients showed evidence of neural axis abnormality on brain and spine MRI (Table 1). This group had an average age of diagnosis of 5 years and 8 months (range 1 year 2 months to 7 years 5 months) and an average age of first attendance in clinic of 5 years and 11 months (range 2 years and 7 months to 7 years and 9 months). There was an equal distribution of right (4)- and left (4)-sided curves with an average Cobb angle of 45.8° (range 20°–88°). Only one patient had a family history of scoliosis (in his maternal grandfather), and only one had a history of back pain. Asymmetrical abdominal reflexes were observed in three (37.5 %) patients (all of whom had neural axis abnormalities on MRI). Five patients had both Arnold–chiari malformation Type I malformation and a syrinx, two had a syrinx and one had an Arnold–chiari malformation Type I malformation.

In the study group as a whole, there was no significant correlation between Cobb angle and age at diagnosis ($r_s = 0.072$, $p = 0.27$). Comparison of means between the subgroups with normal MRI findings and those with an intraspinal anomaly showed no significant difference in Cobb angles ($p = 0.47$), however the later age of diagnosis in the group with intraspinal anomalies was significant ($p < 0.001$). There was no significant difference between patients with and without neural axis abnormality in terms of presentation before or after 36 months of age ($\varphi = 0.18$, $p = 0.13$), curve sidedness ($\varphi = 0$, $p = 0.64$), back pain ($\varphi = 0.05$, $p = 0.52$), and family history ($\varphi = 0.11$, $p = 0.38$). However, there was a significant difference in the occurrence of asymmetrical abdominal reflexes between the two groups ($\varphi = 0.59$, $p < 0.001$). Of the eight patients with neural axis abnormalities, three had asymmetrical abdominal reflexes but of the 64 patients with normal MRIs, none exhibited this sign. All six patients with Arnold–chiari malformation Type I malformation underwent neurosurgical review. Of these, four were managed with posterior fossa decompression and bracing while another received bracing alone and the final one (with a solitary Arnold–chiari malformation Type I malformation) underwent fusion after the age of seven. The two patients with an isolated conus syrinx were managed with growing rod systems to allow for skeletal and pulmonary development. No patients with intraspinal abnormalities were lost to follow up.

Discussion

Idiopathic scoliosis can be classified according to age of onset. In 1950, Ponseti and Friedman originally classified idiopathic scoliosis as early (less than or equal to 10 years of age) and late (more than 10 years of age) onset [13]. James later classified those <3, 5–8 and 10 or more years of age as infantile, juvenile and adolescent onset, respectively [14]. Dickson returned to the original early/late nomenclature using 3 years of age as the cut-off and subsequently increased this to 5 years of age [15, 16]. A degree of error intrinsically exists in the classification systems as some cases of juvenile scoliosis may be late diagnosed infantile cases and a number of adolescent scoliosis cases may be due to a delay in diagnosing juvenile scoliosis. This error may be compounded by the close proximity of age intervals. In the UK, based on the observed peaks of onset during the first year of life and adolescence, idiopathic scoliosis has been classified as early (before 8 years of age) and late (8 years of age or older) onset [8, 12].

Scoliosis observed during the development of a child with normal neurology is presumed to be idiopathic. At present, screening of the neuraxis for abnormality before surgical intervention is mandatory to prevent permanent neurological injury [1–3, 17, 18]. Previous studies have investigated the rates of intraspinal abnormalities in idiopathic scoliosis and found a wide range.

In the USA, Lewonowski et al. [4] observed the rate of intraspinal pathology in idiopathic scoliosis in patients of 10 years of age or less. Of the 26 patients in their study, five (19.2 %) had an abnormality. These were made up of two Arnold–Chiari malformations, one syringomyelia, one intramedullary tumour and one terminal lipoma. Schwend et al. [18] retrospectively reviewed 95 MRI scans of patients with idiopathic scoliosis between one and 28 years of age. 14 abnormalities were detected including 12 syrinxes, one syrinx and astrocytoma and one dural ectasia. Of note is the fact of the eight patients aged 10 years or less with a left-sided thoracic curve, five had an intraspinal abnormality. The number of anomalies found in the EOIS group is not specifically reported. Gupta et al. [1] prospectively and retrospectively reviewed the MRI scans of 98 patients aged 10 years and younger. They found a rate of 19.4 % (19) of spinal abnormalities. In the subgroup of IIS consisting of six patients, three (50 %) had neural axis abnormalities. Dobbs et al. [5] reported a prevalence of neural axis anomalies of 21.7 % (10) in 46 patients with IIS across three clinics. These consisted of five Arnold–Chiari malformations with syrinxes, four syrinxes and one brainstem tumour. Pahys et al. [6] analysed the MRIs of 54 patients with IIS. They found seven (13 %) anomalies consisting of three tethered cords, two Arnold–chiari malformations and two syrinxes.

In India, Rajasekaran et al. [17] analysed the MRIs of 177 consecutive scoliosis patients. Amongst these patients, there were only four infantile and 11 juvenile onset idiopathic cases of scoliosis. Overall, idiopathic scoliosis showed a 16 % rate of neuro-axial abnormalities with Arnold–chiari malformation being the most common (9/15); however, the rate was higher in the IIS (25 %) and JIS (27 %) subgroups. In Japan, Inoue et al. [2] prospectively preformed MRI scans on 250 patients with idiopathic scoliosis of whom 44 (18 %) had intraspinal anomalies with combined syrinx and Arnold–chiari malformation (22) and isolated Arnold–Chiari malformation (13) being the most common. Only 74 of these patients were less than 11 years of age and this subgroup had 19 (26 %) neural axis abnormalities. Subsequently, Nakahara et al. [7] prospectively studied 472 MRIs of patients with idiopathic scoliosis to find 18 (3.8 %) neural axis abnormalities consisting of six Arnold–chiari malformations, ten combined Arnold–Chiari malformations with syrinx and two isolated syrinxes. They reported age of 10 years or less and asymmetrical abdominal reflexes to be associated with the detection of neural axis abnormalities on MRI. Only 53 of their patients had onset before 11 years of age and, of these, seven (13 %) had intraspinal anomalies on MRI.

In the only British study to date, Evans et al. [3] prospectively performed MRI scans on 31 patients between four and 12 years of age to find a 26 % (8/31) rate of neuroanatomical abnormalities consisting of seven Arnold–chiari malformations (one of which was associated with a syrinx) and one astrocytoma.

In summary, previous studies in Japan, India and the United States investigating the rates of spinal anomalies in EOIS have a range between 13 and 27 % [1, 2, 4, 7, 17]. The subgroup of IIS has been found to have rates of 13 to 50 % and JIS to have rates between 14 and 27 % [1, 2, 4–7]. In the only British study to date, a non-standard age range (four to 12 years of age) representing a combination of juvenile and adolescent onset scoliosis was used and a 26 % rate of spinal abnormalities was found. A summary of the literature is represented in Table 2.

There is a geographical variation in the age of onset of idiopathic scoliosis. Ponseti in North America and Wynne–Davies in the British Isles found that the proportion of cases presenting below 8 years of age was 8 and 50 % respectively [8]. No studies have been performed in the UK in regards to the rates of neural axis abnormalities in this age group. This represents the first study to observe the rate of occurrence of this phenomenon in the EOIS population in Britain.

In our 16 year study on 72 patients with idiopathic scoliosis with an age of onset of less than eight from two British spinal deformity centres, we observed eight (11.1 %) neural axis anomalies consisting of five combined

Table 2 Summary of the literature on rates of intraspinal anomalies in early onset idiopathic scoliosis (EOIS) subdivided into infantile idiopathic scoliosis (IIS) and juvenile idiopathic scoliosis (JIS)

Study	Location	Patients			Intraspinal anomalies		
		IIS	JIS	EOIS	IIS	JIS	EOIS
Nakahara et al. [7]	Japan	N/A		53	N/A		7 (13 %)
Rajasekaran et al. [17]	India	4	11	15	1 (25 %)	3 (27 %)	4 (27 %)
Pahys et al. [6]	USA	54	–		7 (13 %)	–	
Inoue et al. [2]	Japan	2	72	74	N/A		19 (26 %)
Dobbs et al. [5]	USA	46	–		10 (22 %)	–	
Gupta et al. [1]	USA	6	92	98	3 (50 %)	N/A	19 (19 %)
Evans et al. [3]	UK	–	31		–	8 (26 %)	
Lewonowski et al. [4]	USA	4	22	26	2 (50 %)	3 (14 %)	5 (19 %)

Arnold–chiari Type I malformations and syrinxes, one isolated Chiari malformation and two isolated syrinxes. The finding that the majority of abnormalities consisted of an Arnold–chiari malformation with concurrent syrinx is consistent with the majority of previous studies in the literature [2–5, 7]. It is still not clear whether these intraspinal anomalies are a cause of abnormal curvature of the spine or a result of medullary traction secondary to scoliosis [19, 20]. Although age at diagnosis was significantly later (3 years 7 months vs. 5 years 8 months) in those found to have a neuro-axial anomaly, there was no significant difference in Cobb angle between patients with normal (46.6°) and abnormal (45.8°) neural axes. This finding may suggest that a later onset may present an increased risk of developing spinal abnormality independent of the severity of curvature; however, when 36 months is used as a cut-off for subgroup analysis, there was no significant difference in the rates of intraspinal anomalies.

There are a number of limitations to the present study. First, the study is retrospective and as such has the disadvantages of relying on the accuracy and completeness of written records. In addition, due to this shortcoming it is not possible to establish cause and effect. Second, the study was limited to those patients who had undergone MRI. Thus, there may be an unknown number of patients with EOIS who had not undergone MRI scanning over the 16 year study period who were not included in the study. However, both institutions have a policy of performing MRI scanning on all patients with EOIS unless contraindications exist. Thus, given that the EOIS patient population has a low likelihood of these contraindications (e.g. intracranial clips, intra-orbital metal fragments, magnetically activated implants and/or pregnancy), it may be reasonable to assume that the number of patients who were not included in the study due to this limitation is likely to be small if any.

It has previously been claimed that patients with a syrinx show a greater prevalence of left-sided thoracic curves; however, the site and extent of curvature is no different [3, 18, 21–23]. Our patient population had a near equal distribution of right (36)- and left (32)-sided curves overall and an equal distribution of right (4)- and left (4)-sided curves in the subgroup with neural axis abnormalities. The near equal distribution of curve sidedness in our population may support the possibility that, similar to the geographical variation in age of onset of idiopathic scoliosis, there may be a geographical variation in predominance of left- and right-sided curves. Although the subsample of patients with intraspinal anomalies is too small to reach significant conclusions, it may be that, in the British population, curve sidedness does not indicate a more "sinister" curve.

Asymmetrical abdominal reflexes with normal neurological function has been reported to co-exist with the presence of a syrinx [7, 24, 25]. In our study, asymmetrical abdominal reflexes were observed in 37.5 % of patients with neural axis abnormalities, but in none of those with normal MRIs.

Although a total spine MRI is still recommended at presentation in patients with EOIS because of the potential high risk of neural axis abnormalities, this study highlights that the occurrence rates may not be as high as previously expected. This multi-centre study helps to establish the rates of neural axis abnormalities in this group of patients whose growing spines pose continued surgical challenges.

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Conflict of interest None of the authors has any potential conflict of interest.

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