Introduction
Although the largest group of scoliosis is idiopathic, a subset has some neurological abnormality. This can be extremely subtle clinically despite marked neurological pathology. Such a case is presented which demonstrates the value of magnetic resonance imaging (MRI) in this group of patients.

Case report
A boy aged 7 years and 10 months is referred for a progressive idiopathic scoliosis from a peripheral hospital, where he had been followed up for 18 months. His mother first noticed the deformity 2 years ago, which had since then progressed. Obstetric history was unremarkable with a normal vaginal delivery, without any antenatal or perinatal problems. The family history was negative for congenital or developmental diseases. His three brothers have no scoliosis. According to the mother, he is a functionally normal child with normal developmental milestones and average to good progress at school. The only presenting complaint was that of a spinal deformity and the patient denied any headaches or back pain and did not report any weakness or neurological deficits.

On examination he was fit and healthy, of average weight and size, walking and running around. The systemic examination was normal, with no evidence of a syndromic cause for the scoliosis. On examination of the spine, he presented with a left-sided thoracic curve and a right-sided lumbar curve, still relatively mobile with correction on right lateral flexion and traction. A left-sided rib hump was evident with the Adams forward bending test. His shoulders were level and his head was centred over his sacrum.

Neurological examination revealed intact sensation, symmetrical reflexes (including superficial abdominal reflexes) and 5/5 strength in both upper and lower extremities. The only positive finding was that of some difficulty in undoing his shirt’s buttons when asked to undress, which was initially ascribed to his age.
The standing anteroposterior radiograph revealed a left thoracic curve with the apex at T9, and a Cobb angle measuring 38°. The right lumbar curve had a Cobb angle measuring 36° (Figures 1 and 2).

MRI was arranged shortly after admission to exclude neural axis pathology because of the atypical left thoracic curve. MRI confirmed our suspicions showing a syrinx formation extending the whole length of his cord and a Chiari 1 malformation of the brainstem (Figures 3a and 3b).

In retrospect the lateral cervical spine radiograph indicated increased anteroposterior canal dimensions to accommodate the dilated cord (Figure 4).

The patient was referred to the neurosurgeons, where a posterior fossa decompression was performed. The surgery was successful, with an uneventful postoperative course.

Discussion
James initially classified idiopathic scoliosis on the basis of the patient’s age at when the scoliosis was first identified: it was classified as infantile at an age of less than three years, as juvenile at an age of three to ten years, and as adolescent at an age of ten years to skeletal maturity.\(^1\) Juvenile idiopathic scoliosis (JIS) makes up approximately 8 to 16% of childhood idiopathic scoliosis. Unlike adolescent idiopathic scoliosis relatively little is known about juvenile idiopathic scoliosis. In a series of 109 patients with juvenile idiopathic scoliosis evaluated by Robinson and McMaster, the boys presented earlier at a mean age of 5 years 8 months, compared to an age of 7 years 2 months for the girls. The ratio of girls to boys was 1:1.6 for those younger than 6 years and 2.7:1 for those older than 6 years at presentation. Although left-sided curves are regarded as atypical in adolescent onset scoliosis, their study showed that in juvenile onset scoliosis there were equal numbers of right- and left-sided curves in the younger group (less than 6 years), with a predominance of right-sided curves (3.9:1) only in the patients older than 6 years.\(^2\) Curves with onset in this age group are often progressive with potential for trunk deformity and eventual cardiac and pulmonary compromise. The rate of progression is 1 to 3 degrees per year before age 10 years, and sharply increases to 4.5 to 11 degrees per year after that age. Juvenile onset scoliosis is more likely to have an underlying spinal cord abnormality as the cause of the deformity, with the incidence of abnormality approximately 20% in this group.\(^3\) In this younger group of patients (less than 10 years) with a neurological cause, actual neurological findings on physical examination are often absent, and often the spinal curvature itself must then be considered the initial sign of a neural axis abnormality.\(^4\)
Lewenowski et al warns us that any unusual spinal deformity in a child is highly suggestive of an underlying neurogenic problem. Therefore utmost vigilance is required when evaluating such patients, and this is in agreement with what Tachdjian and Matson said, "Since musculoskeletal abnormalities constitute the initial manifestation of intraspinal lesions in a large percentage of cases, the orthopaedist should keep his index of suspicion high with respect to the possibility of intraspinal lesions in evaluating all children he is asked to see" 6.

Syringomyelia was first described in 1837 but the association with scoliosis was only described in 1944. Scoliosis associated with syringomyelia has been characterised by the following factors: left thoracic and other atypical curve patterns, rapid curve progression (more than 1° per month), neurological deficits, and cervical curve components. 7,8 These atypical curve patterns associated with syringomyelia were further studied by Spiegel et al 9 and they identified curves for which a high index of suspicion is warranted. These include: ‘an atypical group’, including left thoracic curves, double and triple curves, and long right thoracic curves with end vertebrae caudal to T12 (King 4 and variations thereof), and a ‘typical group’ (right thoracic, right thoracic/left lumbar) with superior or inferior shift of one or both end vertebrae and/or the apex of the curve. Most patients with syringomyelia present with some symptoms including back pain, neck pain, headaches, weakness, abnormal reflexes, ataxia, urologic complaints, and cranial nerve deficits. However sometimes they can be remarkably normal on examination with only the deformity being present. 1,5 This is evident from our case report and confirms that idiopathic scoliosis is still a diagnosis of exclusion and specifically in this juvenile group the responsibility to exclude underlying pathology remains vital. Magnetic resonance imaging (MRI) has become more accessible in most parts of our country, and should be used more frequently in the diagnosis of such conditions. That having been said, we agree with Lewenowski et al who concluded in their study that routine MRI is necessary in all patients presenting before 11 years of age to exclude intraspinal pathology, irrespective of a normal neurological examination and scoliotic curve pattern. 5

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

References