Volume: 24, Issue: 3, July 2013 pp.: 199-212 ORIGINAL ARTICLE / ORJİNAL MAKALE

EVALUATION OF SPINAL AND OTHER SYSTEM PATHOLOGIES IN EARLY ONSET SCOLIOSIS PATIENTS

ERKEN BAŞLANGIÇLI SKOLYOZ HASTALARINDA KARŞILAŞILAN OMURGA VE DİĞER SİSTEM PATOLOJİLERİNİN DEĞERLENDİRİLMESİ

Mehmet Bülent BALİOĞLU[1], Yunus ATICI[1], Akif ALBAYRAK[1], Seçil Sezgin SAKIZLIOĞLU[2], Deniz KARGIN[1], Mehmet Temel TACAL[1], Mehmet Akif KAYGUSUZ[3]

[1]Orthopaedic Surgeon, , Metin Sabancı Baltalimanı Bone Diseases Education and Research Hospital, İstanbul.
[2]Radiologist, , Metin Sabancı Baltalimanı Bone Diseases Education and Research Hospital, İstanbul.
[3]Assoc. Prof. of Orthopaedic Surgery, , Metin Sabancı Baltalimanı Bone Diseases Education and Research Hospital, İstanbul.

SUMMARY

Aim and Purpose: It is very difficult to manage the treatment of early onset scoliosis (EOS). It is necessary to diagnose and start the treatment of patients with EOS as early as possible, and to plan the treatment well. We investigated the pathologies seen in spinal, orthopedic, cardiovascular and other organ systems caused by different etiologies in EOS with the help of magnetic resonance imaging (MRI) and other diagnostic methods, based on etiological reasons.

Materials and Methods: We evaluated 62 EOS patients (26 congenital, 21 neuromuscular, 10 syndromic and 5 idiopathic) who received spinal MRIs, and who were operated on and/or managed due to scoliosis between the years of 2005–2011. The mean age at MRI evaluation was 4.3 years of age (3 months–11 years 10 months), and 38 patients were female and 24 were male. For all patients, we investigated the intraspinal, extraspinal and additional organ anomalies with MRI. We measured the coronal and sagittal plane deformity of the spine with the Cobb method. We also investigated the orthopedic pathologies clinically and radiologically, and the additional organ anomalies with abdominal ultrasound and echocardiogram.

Results: We found intraspinal anomalies in 48 patients (77.4%), extraspinal anomalies in 46 patients (74.1%), and additional organ anomalies in nine patients (14.5%). We documented 98 intraspinal and 101 extraspinal pathologies with MRI. When evaluated according to the spinal regions, 50 (80.6%) of the patients had pathologies in thoracic, 38 (61%) had lumbar, 28 (45%) cervical, and four (6.4%) had pathologies in sacral areas. The most common intraspinal anomaly, observed at a rate of 54.8% (34 patients), was syringomyelia. Intraspinal anomalies were most common in neuromuscular scoliosis patients, at a rate of 55.1% (54 pathologies). The most frequent extraspinal anomaly was hemivertebrae, seen in 38.7% of patients (24 patients). Extraspinal anomalies were most commonly observed in congenital scoliosis patients, at a rate of 62.37% (63 pathologies). The mean Cobb angle in all cases was measured as 46.6° (10–113°). We found increased kyphosis in 18 (29%) cases and lordosis in four (6.4%) cases. In 37 (59.6%) of the patients, an additional 56 orthopedic pathologies were found. The most commonly co-observed problems were motor weakness in the lower extremities in 12 patients (21%), hip dysplasia in nine (16%) patients, and thorax deformities in nine (16%) patients. Orthopedic pathologies were most commonly observed in neuromuscular scoliosis patients, at a rate of 32% (19 patients). We found cardiovascular anomalies in seven patients (11.2%) (most commonly ASD), urogenital anomalies in 11 patients (17.74%) (most frequently bowel and bladder incontinence), and other organic defects in 12 patients (19.35%) (most frequently inguinal hernia). 33 (53.2%) patients had previously received surgery. 18 (29%) of them had had neurosurgical operations, 15 (24.1%) were operated on for orthopedic or other organ anomalies, and only in seven (11.2%) cases were VEPTR (4), telescopic rod (1), epiphysiodesis (1) or hemivertebra (1) excision applied.

Discussion: Even though scoliosis has different etiologies, many spinal and other organ anomalies tend to accompany EOS. Intraspinal anomalies are frequently observed with NS, with syringomyelia as the most common intraspinal anomaly. Extraspinal anomalies of the spine are most frequently seen with CS, and the most common anomaly is hemivertebrae. The thoracic vertebrae are most affected. Orthopedic pathologies are mostly seen with NS. The most commonly encountered cardiovascular anomaly is ASD, which is seen together with SS. In addition to careful clinical and radiological investigations, early MRI investigation can lead to better understanding of accompanying anomalies.

Key words: Early onset scoliosis, Magnetic resonance imaging, Intraspinal anomaly

Level of evidence: Retrospective clinical study, Level III

ÖZET

Giriş-Amaç: İlerleyici erken başlangıçlı skolyoz (EBS) tedavisinin yönetimi oldukça güçtür. EBS hastalarının tanı ve tedavisine mümkün olduğunca erken başlanması ve çok iyi planlanması gereklidir. Çalışmamızda EBS da farklı etiyolojilerin neden olduğu spinal, ortopedik, kardiovasküler ve diğer organ sistemlerinde karşılaşılan patolojiler manyetik rezonans (MRG) ve diğer tanı yöntemleri yardımı ile etiyolojik nedenlere göre incelendi.

Materyal ve Metod: 2005-2011 arası EBS olan, skolyoz nedeniyle opere ve/veya takip edilen tüm omurgası MRG ile incelenmiş 62 hasta (26 konjenital, 21 nöromuskuler, 10 sendrom ve 5 idiopatik nedenlere bağlı) değerlendirildi. MRG ile inceleme yaşı ortalama 4.3 yıl (3 ay-11 yıl 10 ay) ve cinsiyet 38 kız, 24 erkek idi. Tüm hastalarda MRG ile intraspinal, ekstraspinal ve ilave organ anomalileri araştırıldı. Omurganın koronal ve sagittal plan deformitesi Cobb metodu ile ölçüldü. Ortopedik patolojileri klinik ve radyolojik olarak, diğer organ anomalileri ise batın ultrasonografisi ve ekokardiyografi yardımı ile araştırıldı.

Sonuçlar: MRG ile incelemede intraspinal anomali 48 (%77.4), ekstraspinal anomali 46 (%74.1), ek organ anomalisi 9 (%14.5) hastada tespit edildi. MRG ile 62 hastada 98 intraspinal, 101 ekstraspinal patoloji görüldü. Bölgelere göre incelendiğinde torakal vertebralar 50 (%80.6), lomber 38 (%61), servikal 28 (%45), sakral 4 (%6.4) olguda etkilenmişti. İntraspinal anomali olarak en fazla syringomyeli %54.8 (34 hastada) görüldü. İntraspinal anomali en sık nöromuskuler skolyozda %55.1 (54 patoloji) tespit edildi. Ekstraspinal olarak en fazla hemivertebra % 38.7 (24 hastada) ile karşılaşıldı. Ekstraspinal anomali en sık konjenital skolyozla birlikte %62.37 (63 patoloji) görüldü. Koronal planda tüm olguların ortalama Cobb açısı 46.6° (10-113°) ölçüldü. 18 (%29) olguda kifoz artışı, 4 (%6.4) olguda ise lordoz artışı tespit edildi. 37 (%59.6) hastada ilave 56 ortopedik patoloji tespit edildi. En fazla birlikte görülen problem alt ekstremitelerde motor kuvvetsizlik 12 hastada (%21), kalça çıkığı 16%) 9) ve toraks deformitesi 9 (%16) idi. Ortopedik patolojiler en sık nöromuskuler skolyoz ile 19 hastada (%32) görüldü. 7 (%11.2) hastada kardiovasküler anomali (en fazla ASD), 11 (%17.74) ürogenital anomali (en fazla idrar/gayta inkontinansı), 12 olguda (%19.35) diğer organik defektler (en sık inguinal herni) tespit edildi. 33 hasta (%53.2) opere idi. İntraspinal patolojileri nedeniyle 18 (%29) hasta daha önce nöroşirürjikal operasyon geçirmişti, 15 hasta (%24.1) ortopedik ve/veya diğer organ anomalileri nedeniyle opere edilmişti, sadece 7 olguda (%11.2) omurga deformitesine yönelik VEPTR (4), uzatılabilir rodlar (1), epifizyodez (1) ve hemivertebra (1) eksizyonu uygulanmıştı. Tartışma: Skolyoz nedenlerine göre değişiklikler göstermekle birlikte EBS da omurganın intraspinal ve ekstraspinal anomalileri ile birlikte diğer organ anomalileri yüksek oranda eşlik etmektedir. EBS da omurganın intraspinal anomalisi en fazla NS da görülürken en sık intraspinal anomali nedeni siringomiyelidir. Omurganın ekstraspinal anomalisi en fazla KS da görülürken en sık neden hemivertebradır. En fazla torasik vertebralar etkilenmektedir. EBS da yüksek oranda ortopedik patolojiler eşlik ederken en sık NS ile birlikte görülür. Kardiovasküler anomali olarak en fazla ASD, en sık SS a eşlik etmektedir. Ürogenital anomali en fazla inkontinans olarak, en sık SS da görülmektedir. Dikkatli klinik ve radyolojik incelemelerin yanı sıra erken MRG analizi ile eşlik eden diğer anomaliler daha iyi anlaşılabilir.

Anahtar kelimeler: Erken başlangıçlı skolyoz, Manyetik rezonans görüntüleme, İntraspinal anomali.

Kanıt düzeyi: Retrospektif klinik çalışma, Düzey III

Address: Mehmet Bülent Balioğlu, MS Baltalimanı Kemik Hastalıkları Eğitim ve Araştırma Hastanesi, Sarıyer, İstanbul.
Tel.: 0532 2521483
E-mail: mbbalibey@gmail.com
Received: 12th April, 2013
Accepted: 6th June, 2013

INTRODUCTION

James defined idiopathic scoliosis (IS) in 1954 by grouping it into three types: deformity that onset before the age of 3 as infantile, onset between 4–9 years of age as juvenile, and onset from 10 years old to the end of the growth spurt as adolescent[28]. These three periods were predicted to correspond to the different rates of growth during childhood and adolescence. It was indicated that during the juvenile period, spinal growth is slowed down and the onset of scoliosis is rare[13, 15].

Later, Dickinson defined the onset of scoliosis up to 5 years of age as "early onset", and scoliosis seen in children older than 6 years as "late onset". Among early onset scoliosis (EOS), different etiologies were defined, including congenital scoliosis (CS), neuromuscular reasons (cerebral palsy, myelomeningocele, muscular dystrophies etc) as neuromuscular scoliosis (NS), syndromes (neurofibromatosis etc) as syndromic scoliosis (SS), and structural lesions of the central nervous system (diastometamyelia, syrinx, tethered cord etc). Patients with unclear associated anomalies were accepted as early onset idiopathic scoliosis. The term "infantile" was coined for idiopathic etiologies, in the same way as adolescent idiopathic scoliosis (AIS)[12,14,22].

The distinction between early and late onset is very important, due to the increased risk of serious cardiopulmonary consequences with scoliosis found before the age of 5[1,22]. It is believed that the age of onset is very important regardless of the etiology, due to a high probability of other organ anomalies and additional pulmonary complications with major thoracic deformity in children younger than 5 years of age[1,7].

When spinal deformities are evaluated in infantile idiopathic scoliosis (IIS), asymptomatic neural axis anomalies must be considered[9,16,19,24]. In IS patients for whom neural axis anomalies have been detected using magnetic resonance imaging (MRI), 50% may require neurosurgical intervention between the ages of 0–10[20,24,30]. The neural axis and the vertebrae develop simultaneously. Because of this concurrence, neurological anomalies must be investigated in CS patients with or without neurological anomalies[25]. Paraspinal anomalies are mostly silent, but in some children foot deformities, extremity contractures, extremity atrophy, asymmetrical reflexes, and changes in bladder and bowel function may be observed together with neurological symptoms[25]. For progressive curvatures of more than 20°, MRI investigation is required to differentiate central nervous system lesions[1].

Progressive EOS is difficult to treat in children. Treatments available for older children, like orthosis or spinal fusion, are less efficient in smaller children and may have negative effects on growth, immature vertebrae, lungs and the thoracic cage[1]. The rates of surgical intervention in younger patients with spinal deformities are rising, due to techniques like VEPTR or growing rods, which allow growth. In EOS, surgical treatments are required for other commonly encountered abnormalities. Because of this, diagnosis and treatment of EOS patients must be started early and planned meticulously.

In this study, accompanying spinal and extraspinal abnormalities were investigated using assisting diagnostic techniques, particularly MRI, in EOS patients. The etiological causes of spinal, orthopedic, cardiovascular and other organ system pathologies in EOS patients were studied.

MATERIALS AND METHODS

62 consecutive patients with EOS, operated on or under surveillance for scoliosis, who had a full vertebral MRI, and were referred to our clinic during 2005–2011 (38 female, 24 male), were evaluated. The distribution of the patients according to the scoliosis etiology was as follows: 26 patients with CS, 21 with NS, 10 with SS, and 5 with IS. Patients were included in the study with onset before the

age of 5, at least ten coronal plane deformities, or accompanying sagittal plane deformities. The average age of examination with MRI was 4.3 years (3 months–11 years 10 months). The MRIs of all patients were examined by a radiology specialist to detect intraspinal and extraspinal abnormalities of the vertebrae, and additional organ abnormalities.

The whole spinal column was examined in the coronal plane with T2-weighted images, the sagittal plane with T1- and T2-weighted images, and the axial plane with continuous scanned T2- weighted images. Additionally, all the areas included in the section during examination were studied for additional abnormalities.

Coronal and sagittal plane deformities of the vertebrae were calculated using the Cobb method and conventional radiographs. For additional abnormalities, abdominal ultrasonography and echocardiography were performed. Orthopedic pathologies were evaluated in detail, clinically and radiologically. Neurological examination was performed for the upper and lower extremity motor and sensory reflexes. For children with neurological findings and intraspinal abnormalities detected with imaging, neurosurgical consultation was requested. In cases of hiatal hernia, inguinal hernia, endocrine pathologies, urological, genital and cardiovascular abnormalities, the pediatric surgery, urology and cardiology departments were consulted and the patients received surgery for intraspinal and other organ pathologies, if required. For spinal deformities, after treatment for other abnormalities was conducted and an optimal environment was provided, deformity surgery was performed.

The average, standard deviation, median, percentiles and ratios were used for descriptive statistics. For comparison of categorical data, the chi-square test and, if necessary, the Fisher test, were employed.

RESULTS

The vertebrae of 62 consecutive patients with a diagnosis of EOS (26 CS, 21 NS, 10 SS, and 5 IS) were examined with MRI. In the coronal plane, including all patients, the average Cobb angle was 46.6° (10–113°). In the sagittal plane, 29% (n=18) of patients had increased kyphosis, while 6.4% of them (n=4) had increased lordosis.

Intraspinal abnormalities were seen by MRI in 77.41% of patients (n=48; 14 NS, 26 CS, 6 SS). Additional organ abnormalities were seen by MRI in 14.5% of patients (n=9; 6 NS, 1 CS, 2 SS). There were 102 intraspinal and 108 extraspinal abnormalities detected. When the pathologies were reviewed according to the regions of vertebrae, multiple vertebral regions were found to be affected. The thoracic spine was most commonly affected (83.87%, n=52), and the lumbar spine was affected in 66.12% of patients (n=41), the cervical spine in 45.16% (n=28), and the sacral spine in 12.90% (n=8) of patients (Table 1).

INTRASPINAL ABNORMALITIES

When EOS patients were examined with MRI, the ratio of intraspinal pathologies was 77.41% (n=48). These were detected most commonly with NS (95.23%, n=20), followed by CS (73.07%, n=19), SS (70%, n=7) and IS (40%, n=2). The most commonly observed intraspinal abnormality was syringomyelia in 54.83% of patients (n=34). The prevalence of syringomyelia was 66.66% with NS (n=14), 50.00% with CS (n=13), 50.00% with SS (n=5), and 40.00% with IS (n=2). Tethered cord was the second most commonly observed intraspinal abnormality, with a prevalence of 43.54% (n=27). This was observed at a rate of 66.66% with NS (n=14), 46.15% with CS (n=2), and 10% with SS (n=1). Diastematomyelia (split cord) was observed in 19.35% of patients (n=12). This was observed most commonly with NS (28.57%, n=6) and CS (23.07%, n=6). The prevalence of Chiari malformation with EOS was 12.9% (n=8), and this was observed only with NS at a rate of 38.09% (n=8).

Table-1. Distribution of EOS patients according to gender, age, Cobb angle values, and affected spinal region according to MRI results.

EOS	Age	Gender		Cobb Angle	MRG (n=62)							
		M	F	Coronal (n=62)	Intraspinal Anomalies	Extraspinal Anomalies	Additional Organ Anomalies	Number of Intraspinal Pathologies	Number of Extraspinal Pathologies	Cervical Region	Thoracic Region	Lumbar Region	Sacral Region
CS	4.3 years (3 months-11 years 10 months)	8	18	46.6° (10-113)	19	26	1	102	108	45.16% (n=28)	83.87% (n=52)	66.12% (n=41)	12.90% (n=8)
NS		10	11		20	14	6						
SS		6	4		7	6	2						
IS			5		2								
Total		24	38		48(77.41%)	46(74.19%)	9(14.5%)						

Table-2. Distribution of intraspinal anomalies in EOS according to etiology.

EOS	Intraspinal Anomalies	Syringomyelia	Tethered cord	Diastematomyelia (Split cord)	Chiari Malformation	CSF cyst	Neurenteric cyst	Arachnoid cyst
CS	73.07% (n=19)	50.00% (n=13)	46.15% (n=2)	23.07% (n=6)	-	-	1.61% (n=1)	1.61% (n=1)
NS	95.23% (n=20)	66.66% (n=14)	66.66% (n=14)	28.57% (n=6)	38.09% (n=8)	1.61% (n=1)	-	-
SS	70% (n=7)	50.00% (n=5)	10.00% (n=1)	-	-	-	-	-
IS	40% (n=2)	40.00% (n=2)	-	-	-	-	-	-
Total	77.41% (n=48)	54.83% (n=34)	43.54% (n=27)	19.35% (n=12)	12.90% (n=8)	1.61% (n=1)	1.61% (n=1)	1.61% (n=1)

Table-3. Distribution of extraspinal anomalies.

	Extraspinal Anomalies	Hemivertebrae	Butterfly vertebrae	Segmentation Abnormality	Spina Bifida	Myelomeningocele	Hydrocephalus	Sacral agenesis/ hypogenesis	Spondylolisthesis	Atlantoaxial rotatory subluxation	Spino-pelvic instability
CS	100% (n=26)	76.92% (n=20)	61.53% (n=16)	38.46% (n=10)	0.76% (n=8)	-	3.84% (n=1)	3.84% (n=1)	3.84% (n=1)	3.84% (n=1)	-
NS	61.9% (n=13)	19.04% (n=4)	23.80% (n=5)	14.28% (n=3)	14.28% (n=3)	42.85% (n=9)	33.33% (n=7)	23.80% (n=5)	-	-	-
SS	60% (n=6)	20% (n=2)	10% (n=1)	10% (n=1)	-	-	-	10% (n=1)	10% (n=1)	-	10% (n=1)
IS	-	-	-	-	-	-	-	-	-	-	-
Total	74.19% (n=46)	41.93% (n=26)	35.48% (n=22)	22.58% (n=14)	17.74% (n=11)	14.51% (n=9)	12.90% (n=8)	11.29% (n=7)	3.22% (n=2)	1.61% (n=1)	1.61% (n=1)

Additionally, CSF cysts (with NS), neurenteric cysts (with CS), and arachnoid cysts (with CS) were each observed in 1.61% of patients (n=1) (Table-2).

SPINAL ABNORMALITIES

Spinal abnormalities were found in 74.19% (n=46) of patients. They were most commonly observed with CS, with a prevalence of 100% (n=26), followed by NS (61.90%, n=13) and SS (60.00%, n=6). The most common anomaly was hemivertebrae (41.93%, n=26). In order, these were found with a prevalence of 76.92% with CS (n=20), 19.04% with NS (n=4), and 20% with SS (n=1). Butterfly vertebrae were the second most common abnormality, with a prevalence of 35.48% (n=22). In order, these were found with a prevalence of 61.53% with CS (n=16), 14.28% with NS (n=3), and 10.00% with SS (n=1).

Spina bifida was found in 17.74% (n=11) of patients, including in 30.76% of cases with CS (n=8), and 14.28% in cases of NS (n=3). Myelomeningocele was found in 14.51% (n=9) of cases, and only seen with NS (42.85%, n=9). Hydrocephalus was seen in 12.9% (n=8) of cases, with a prevalence of 33.33% with NS (n=7) and 2.84% with CS (n=1). Sacral agenesis/hypogenesis was seen in 11.29% (n=7) of cases, with a prevalence of 23.80% with NS (n=5), 3.84% with CS (n=1), and 10.00% with SS (n=1). Spondylolisthesis was seen in 3.22% of cases (n=2), with a prevalence of 3.84% with CS (n=1) and 10.00% with SS (n=1). Atlantoaxial rotatory subluxation was observed with CS, and spinopelvic instability was observed with SS, each with a prevalence of 1.61% (n=1) (Table-3).

ACCOMPANYING PROBLEMS

71 different accompanying orthopedic pathologies were determined in 64.51% (n=40) of patients. Orthopedic pathologies were most commonly observed with NS (35.21%, n=25), followed by CS (33.80%, n=24), SS (23.94%, n=17), and IS (7.04%, n=5). The most common co-existing problem was motor weakness of the lower extremities (paresis/paralysis) in 25.80% of cases (n=16), which was observed at a rate of 57.14% with NS (n=12), 20% with SS (n=2), and 7.69% with CS (n=2). Other orthopedic problems, in order of abundance, were hip dislocation with a prevalence of 14.15% (n=9), seen with CS in 15.38% of cases (n=4), with NS in 14.28% of cases (n=3), and with SS in 20% of cases (n=2). The incidence of rib cage/rib abnormalities was 14.5% (n=9), which was 15.4% (n=4) with CS, 40% (n=4) with SS, and 20% (n=1) with IS. Pes planovalgus was found with an incidence of 8.06% (n=5), which was 3.84% (n=1) with CS, 14.28% (n=3) with NS and 10% (n=1) with SS. Femur shortness was seen with an incidence of 6.45% (n=4), which was 3.84% (n=1) with CS, 9.52% (n=2) with NS, and 20% (n=1) with IS. PEV was found with an incidence of 4.38% (n=3), which was 7.69% (n=2) with CS and 4.76% (n=1) with NS. The incidence of lumbar hypertrichosis was 3.22% (n=2), which was observed in 7.69% (n=2) of CS cases. Pes cavus showed an incidence of 3.22% (n=2), and this was 4.76% (n=1) with NS and 20% (n=1) with IS. The incidence of spasticity was 3.22% (n=2), observed only in cases of NS, with an incidence of 9.52%. Pes equinus was observed in 3.22% (n=2) of cases, in 4.76% (n=1) of NS cases and 10% (n=1) of CS cases. Metatarsus adductus was seen in 3.22% (n=2) of cases, in 10% (n=1) of CS cases and 20% (n=1) of IS cases. Pectus excavatum, micrognathia, platybasia, lower extremity contracture, pelvic wing hypoplasia, radius head dislocation, joint laxity, short stature, dental structure abnormalities, deformity of femur head, cubitus varus, coxa vara, clavicular aplasia (SS), pes calcaneovarus (IS), and lower extremity hemimelia (CS) were each observed in 1.61% of cases (n=1) (Table-4).

CARDIOVASCULAR ABNORMALITIES

The incidence of cardiovascular abnormalities was 11.29% (n=7), with some patients having more than one abnormality (n=16). The most commonly observed abnormality was atrial septal defect (ASD), seen in 9.76% (n=6) of patients, and seen in 60% of SS cases (n=3), 9.5% of NS cases (n=2), and 3.8% of CS cases (n=1). Ventricular septal defect (VSD) was observed second most commonly, in 4.83% (n=3) of patients (CS 1, NS 1, SS 1). Patent ductus arteriosus was seen in 3.22% (n=2) of patients (CS 1, NS 1). Pulmonary hypertension, coarctation of aorta, right dislocation of arcus aorta, cardiomyopathy and mitral insufficiency were seen in 1.61% (n=1) of patients (Table-5).

UROGENITAL SYSTEM ABNORMALITIES

When urogenital system abnormalities were examined, these were found in 22.58% of patients (n=14), with 22 different pathologies. In some patients, multiple pathologies were observed.

The most commonly observed pathology was urinary/stool incontinence, in 9.67% (n=6) of the patients, in 23.80% of NS cases (n=5) and 10% of CS cases (n=1). Hydronephrosis was seen in 4.83% (n=3) of patients (CS 1, NS 1), vesicoureteral reflux, horseshoe kidney, conjoined kidney, hypoplastic kidney, and neurogenic bladder were each observed in 3.22% (n=2) of patients, and hypospadias, kidney stones and kidney agenesis were each observed in 1.61% of cases (n=1) (Table-6).

OTHER SYSTEM ABNORMALITIES

These were found in 19.35% (n=12) of patients. In order, they were seen in 40% of IS patients (n=2), 20% of SS patients (n=2), 19.23% of CS patients (n=5) and 14.28% of NS cases (n=3). The most commonly observed abnormality was inguinal hernia, with an incidence of 4.83% (n=3), which was 9.52% (n=2) in NS and 10% (n=1) in IS. Hypothyroidism was seen in 3.22% (n=2) of patients, and in 3.84% (n=1) of CS cases and 4.76% (n=1) of NS cases. Reflux was seen in 3.22% (n=2) of patients, in 10% (n=1) of SS cases and 2% (n=1) of IS cases. Pulmonary stenosis was seen in 1.61% (n=1) of cases, and in 10% of SS cases.

Gall bladder stones, round atelectasis, cleft palate and diaphragm herniation were each seen in 1.61% (n=1) of cases, and were all seen with CS, in 3.84% of cases (Table 7).

Statistically, there were no significant differences between the intraspinal pathologies (except for Chiari malformation) seen with CS and NS (p=0.059). However, Chiari malformation (seen in 38.1% of NS and 0% of CS cases, p=0.001) and extraspinal pathologies (seen in 61.9% of NS and 100% of CS cases, p=0.001) were found to be statistically significant. Hemivertebrae and butterfly vertebrae were significant (p=0.000 and p=0.017, respectively). Other organ abnormalities detected by MRI were also found to be significantly different between cases of NS (28.6%) and CS (3.8%) (p=0.035).

Table-4. Distribution of orthopedic problems accompanying EOS.

	Accompanying orthopedic problems	Motor weakness of lower extremity	Hip dislocation	Rib cage/ rib abnormalities	Pes planovalgus	Shortness of femur	PEV	Lumbar hypertrichosis	Pes cavus	Spasticity	Pes Ekinus	Metatarsus Adduktus	Pes Calcaneovarus	Hemimelia	Pectus Excavatum, microgmathia, platisbasis, contracture of lower extremities, pelvic wing hypoplasia, radius head dislocation, joint laxity, short stature, dental structure abnormalities, deformities of the femur head, cubitus varus, coxavara, clavicular aplasia
CS	23.94% (n=17)	7.69% (n=2)	15.38% (n=4)	15.38% (n=4)	3.84% (n=1)	3.84% (n=1)	7.69% (n=2)	7.69% (n=2)	-	-	-	-	-	3.84% (n=1)	-
NS	35.21% (n=25)	57.14% (n=12)	14.28% (n=3)	-	14.28% (n=3)	9.52% (n=2)	4.76% (n=1)	-	4.76% (n=1)	9.52% (n=2)	4.76% (n=1)	-	-	-	-
SS	33.8% (n=24)	20% (n=2)	20% (n=2)	40% (n=4)	10% (n=1)	-	-	-	-	-	10% (n=1)	10% (n=1)	-	-	10% (n=1)
IS	7.04% (n=5)	-	-	20% (n=1)	-	20% (n=1)	-	-	20% (n=1)	-	-	20% (n=1)	20% (n=1)	-	-
Total	64.51% (n=40)	25.80% (n=16)	14.51% (n=9)	14.51% (n=9)	8.06% (n=5)	6.45% (n=4)	4.83% (n=3)	3.22% (n=2)	3.22% (n=2)	3.22% (n=2)	3.22% (n=2)	3.22% (n=2)	1.61% (n=1)	1.61% (n=1)	1.61% (n=1)

Table-5. Cardiovascular abnormalities accompanying EOS.

	Cardiovascular Abnormality	Number of cardiovascular abnormalities	ASD	VSD	PDA	Pulmonary hypertension, coarctation of aorta, right side dislocation of arcus aorta, cardiomyopathy, mitral insufficiency
CS	11.29% (n=7)	n=16	3.8% (n=1)	3.8% (n=1)	3.8% (n=1)	-
NS			9.52% (n=2)	4.76% (n=1)	4.76% (n=1)	-
SS			60% (n=3)	20% (n=1)	-	-
IS			-	-	-	-
Total			9.67% (n=6)	4.83% (n=3)	3.22% (n=2)	1.61% (n=1)

Table-6. Urogenital system abnormalities accompanying EOS.

	Urogenital system abnormality	Number of pathologies observed in urogenital system	Incontinence	Hydronephrosis	Vesico-uretral reflux, horse-shoe kidney, conjoined kidney, hypoplastic kidney, neurogenic bladder	Hypospadiasis, kidney stones, kidney agenesis
CS	22.58% (n=14)	n=22			-	-
NS			23.8% (n=5)	4.76% (n=1)	-	-
SS			10% (n=1)	20% (n=2)	-	-
IS					-	-
Total			9.67% (n=6)	4.83% (n=3)	3.22% (n=2)	1.61% (n=1)

Table-7. Other organ abnormalities.

	Other organ abnormalities	Inguinal hernia	Hypothyroiditis	Reflux	Pulmonary Stenosis	Gall bladder stones, round atelectasia, cleft palate, diaphragm hernia
CS	19.23% (n=5)	-	3.84% (n=1)	-	-	3.84% (n=1)
NS	14.28% (n=3)	9.52% (n=2)	4.76% (n=1)	-	-	-
SS	20% (n=2)	-	-	10% (n=1)	10% (n=1)	-
IS	40% (n=2)	10% (n=1)	-	20% (n=1	-	-
Total	19.35% (n=12)	4.83% (n=3)	3.22% (n=2)	3.22% (n=2)	3.84% (n=1)	3.84% (n=1)

53.2% (n=33) of patients had received previous surgery for various reasons. 29% (n=18) of patients with intraspinal abnormalities detected by MRI were operated on neurosurgically. For orthopedic or other organ abnormality reasons, 24.1% of patients (n=15) received surgery. For EOS, 11.2% (n=7) of the patients were operated on, and the procedures were VEPTR (n=4), growing rod (n=4), epiphysiodesis (n=1) and hemivertebrae excision (n=1). No neurological complications developed postoperatively.

DISCUSSION

EOS typically presents with vertebral deformity in the first five years of life. It may spontaneously regress or may be progressive. Radiological criteria help to distinguish between spontaneously regressive and progressive deformities[22].

Radiological examination of EOS must be performed by evaluating antero-posterior and lateral imaging of whole vertebrae, and calculation of the Cobb angle and rib-vertebrae angle difference (RVAD). The RVAD is useful for showing progression when the rib head is not overlapping with the vertebrae (phase 1). When the RVAD is >20°, this predicts progression, and a RVAD smaller than 20° predicts regression. If the rib head is overlapping with the vertebrae (phase 2), the deformity has already progressed and the RVAD cannot be calculated[33].

After examination for scoliosis and secondary manifestations, patients must be examined for conditions accompanying EOS. For differential diagnosis of the etiological causes of deformation, plagiocephalus, congenital heart diseases, inguinal hernia, hip dysplasia, congenital torticollis and congenital abnormalities of the extremities must be watched for and evaluated in detail in EOS[1,10,17,22,27,45]. When curvature is calculated, congenital vertebral abnormalities must also be examined radiologically. The lumbosacral and hip joints must be included in the radiographic area, to examine additional vertebral abnormalities and developmental hip dysplasia[17]. A careful neurological examination must be conducted of the muscle tone and reflexes, to detect hidden central nervous system abnormalities. Sometimes the only clinical signs of syringomyelia or Chiari type I malformations may be superficial abdominal reflex abnormalities or loss of the gag reflex. The superficial abdominal reflex is lost at the convex side of the deformation and may be the only clue for the coexistence of syringomyelia and Chiari I malformations, and must be examined for a clinical diagnosis of intraspinal abnormalities[8,22,35,41,46,47]. It has been reported thatthoracic hyperkyphosis (>40°) and a juvenile onset (≤10 years of age) are risk factors for neural axis abnormalities[11,37,44]. If there are neural axis abnormalities, the lower extremities may be affected. Foot asymmetry, vertical talus, leg atrophy, cavus feet, and club foot may be correlated with CS. For those patients, MRI examination of the brain stem and spinal cord is necessary[25].

While evaluating EOS patients, the normal developmental physiology of the vertebrae and lung must be known. A significant increase in lung parenchyma occurs in parallel with the growth of the vertebrae and rib cage. Two-thirds of the sitting height is reached before the age of 5. The growth rate of the T1–L5 segments is highest between birth and 5 years of age, growing more than 2 cm, and this rate shows a dramatic deceleration from 5–10 years of age. The growth rate increases after 10 years of age but never reaches as high a rate as before the age of 5. From birth to the maturation of the skeleton, the height of the thoracic vertebrae doubles. The thoracic volume is 6% of the adult volume at birth, before 5 years of age it reaches 30%, and before 10 years it reaches 50%. The final thoracic volume is reached in both males and females at the age of 15[15]. At the beginning and end of treatment, pediatric pulmonary evaluation is very valuable. In patients with chest wall and rib asymmetry, more frequent respiratory function tests may be required, although it is difficult to perform such tests on children younger than 5. Tests such as 3D BT for the determination of lung function and volume may inform the physician about the seriousness of problems and the timing and efficiency of surgery[23]. Extraspinal and intraspinal pathologies have been observed, particularly with EOS. Because of this, the addition of detailed MRI analysis to clinical and radiological evaluation is recommended[22]. Lewonowski found intraspinal abnormalities using MRI in 50% (n=2) of four IIS patients. In both of those patients, Chiari Type I malformations with syringomyelia requiring posterior fossa decompression were detected[30].

Gupta, in a prospective study that included 34 patients younger than 10 years of age, with normal neurological findings, and a Cobb angle smaller than 20°, observed the incidence of neural axis abnormalities as 17.6%. A hidden neural axis abnormality was found in three of the six patients aged younger than 6. In two of those patients,

dislocation of the cerebellar tonsils through the foramen magnum (Chiari type I malformation) was shown, and accompanying cervico-thoracic syrinx

required surgical decompression, while the third patient had diffuse dural ectasia[9]. When patients younger than 10 years were compared with adolescent patients with IS, a high rate of accompanying intraspinal abnormalities were observed (Chiari malformation, syringomyelia, tethered cord or neoplasm). These abnormalities were found together with 20% of idiopathic juvenile scoliosis[5,20,24,30].

Belmont emphasized the inadequacy of patient history and physical examination findings for the determination of intraspinal abnormalities, and stated that for CS patients with isolated hemivertebrae, all of the vertebrae must be scanned with MRI (4). In a retrospective study of 46 patients with IIS and normal neurological examination, with no related syndromes or congenital abnormality, with a Cobb angle smaller than 20° and aged an average of 17 months (2–37), examined with MRI, Cobbs showed that neural axis abnormalities were seen in 21.7% (n=10) of patients, and also observed Arnold-Chiari malformation and accompanying syringomyelia (n=5), syringomyelia alone (n=3), and a lowered position of the conus and brain stem tumor (n=1). A total MRI scan was proposed for EOS patients with deformities of less than 20°[16].

MRI is particularly accepted as a standard for the evaluation of dysraphism. In study on CS and intraspinal abnormality, patient examinations and direct X-rays showed intraspinal abnormalities in three of the nine patients, while MRI detected neural axis abnormalities in 35% of the patients[39]. The incidence of intraspinal abnormalities that were detected by MRI in a congenital patient series was between 32–37%[2,43]. Hedequist indicated that if CS patients show deformity progression and surgical intervention is planned, MRI is required[25]. Even with normal findings in a physical examination, the entire vertebrae must be studied by MRI, due to the high incidence of neural axis abnormalities in IIS with EOS children[1].

In recent years, the meticulous evaluation of EOS children using specific imaging methods has become possible. MRI is indicated for children with deformities of less than 20°, to screen the neural axis for central nervous system lesions[1,22]. Pahys, in a study including 54 IIS patients, found intraspinal abnormalities in 13% of patients (n=7; three tethered cord, two Chiari type I malformations, and two isolated syrinx) using MRI, and 71.4% (n=5) of the patients required neurosurgical intervention[38]. Fernandes and Weinstein emphasized the importance of an MRI scan for EOS patients with major indications for deformity progression[21]. As general anesthesia holds relatively little risk, screening of the progression rate (every 3–4 months) was suggested. If the deformity progression accelerates (>10°/year) and there is no change in neurological examinations or surgical intervention is planned in IIS patients, they must be screened with MRI[38].

McMaster published the rate of abnormality as 18% in a 251 patient series[32]. By MRI, the incidence of neural abnormality was found to be high for mixed or segmentation-defective patients, with spinal abnormalities documented in 37% of patients [(2)]. Diastematomyelia (split cord), syringomyelia, Chiari malformation, tethered cord, intradural lipoma, dural band, cyst, and stretched filum terminale dysraphism were common forms. In some studies in the literature, tethered cord dysraphism was most commonly observed in CS patients. An absence of cutaneous findings of dysraphism and a lack of neurological deficit does not directly indicate a lack of intraspinal dysraphism[2,6,43]. Conversely, in a study which retrospectively evaluated 226 CS patients, the incidence of intraspinal abnormality was found to be 43% (n=99), and as opposed to previous studies, diastematomyelia was found to be the most common intraspinal abnormality. The incidence of

intraspinal abnormality was found to be higher than formation insufficiencies, segmentation defects and mixed defects. More intraspinal abnormalities coexisted with patients with thoracic hemivertebrae than with patients with lumbar hemivertebrae[42].

Sun showed 31.70% (n=13) intraspinal abnormalities in a study including 41 children with congenital spinal deformities (37 CS and four congenital kyphosis), but patients with myelomeningocele were excluded from the study[43].

In our study, the incidence of intraspinal abnormality in EOS patients was 77.41%, higher than seen in other published studies. This might be due to the inclusion of patients with intraspinal abnormalities such as myelomeningocele, with a high intraspinal abnormality incidence. The most commonly observed intraspinal abnormalities in our EOS patients were, in order of frequency, syringomyelia (54.83%), tethered cord (43.54%), and diastematomyelia (19.35%). The number of CS patients was limited compared to the other two studies in the literature, and for those patients, the most common intraspinal abnormality was syringomyelia (50.00%) followed by tethered cord (46.15%), and the rarest was diastometamyelia (23.07%). Extraspinal pathologies of the spine were observed in 74.19% of patients, most commonly hemivertebrae (41.93%).

Treatment of EOS is the most commonly discussed subject in pediatric surgery. Treatment of children younger than 5 years old with progressive scoliosis is difficult. Untreated deformities may lead to serious cardiopulmonary and skeletal deformities[1]. Any suspicion of a murmur or cardiovascular abnormality during examination requires consultation with cardiology. Patients requiring surgery for CS must be evaluated by echocardiogram. Cardiological problems in CS may be very serious, such as atrial and ventricular septal defects, patent ductus arteriosus and Tetralogy of Fallot. In 126 patients with congenital spinal deformities, the most common problem was a ventricular septal defect (26%), followed by an atrial septal defect[2]. Beals et al. reported the incidence of congenital heart disease as 12%[3]. Bollini et al. found 8% congenital heart disease in 75 hemivertebrae patients[6]. In another series with 226 cases of CS, the incidence of cardiac defects was found to be 18%, and the most common organic pathologies were extraspinal pathologies[42]. Our EOS patients were also evaluated for cardiac pathologies, and these were detected in 11.29% (n=7) of patients. Multiple cardiovascular abnormalities were found in some patients. The incidence was 60% in SS, 9.52% in NS and 3.84% in CS.

More than 20% of CS patients had accompanying urological abnormalities that were asymptomatic[31]. Problems of the renal and collecting systems, asymptomatic unilateral kidney agenesis and destructive uropathy can be observed. In every child with CS, during USG or spinal MRI, the abdominal cavity and kidneys must be evaluated[18]. In cases where no spinal cord MRI is obtained, USG is the preferred method. Most urological abnormalities are benign, but the most recent studies have shown that a third of patients require urological treatment[2]. For that reason, appropriate screening must be conducted[25].

The incidence of urogenital abnormality has been reported to be 12–24%[2,31,42]. Suh showed the most common urogenital abnormality co-existing with CS was urorectal abnormality (15%)[43]. When the urorectal and urogenital systems of our patients were investigated, 22.58% (n=14) had abnormalities. Multiple abnormalities were seen in some patients (n=22). Urinary/stool incontinence was most commonly observed, in 9.67% (n=6) of patients, and in 23.80% of NS patients (n=5) and 10% of SS patients (n=1).

The coexistence of IIS with hip dysplasia, congenital heart disease and mental retardationhas been reported[40.] Wynne-Davies found 13% mental retardation and 7% inguinal hernia in male infants

with progressive deformation. A study by Mehta also agrees with these results. Mehta pointed out that infants with hypotonia cannot resist deformation, unlike infants with normal tonus[34].

Conner showed that the risk of scoliosis increased among children with congenital malformation and hiatal hernia[10]. Hearing abnormalities are common, especially with congenital vertebral abnormalities involving the cervical and cervicothoracic regions of the vertebrae[25]. In CS patients, other organic defects were seen in 40% of patients (n=91), cardiac defects in 18% of patients, urogenital abnormalities in 2%, and gastrointestinal abnormalities in 5% of patients[42]. In our study, other system abnormalities were seen in 19.35% of patients (n=12). Inguinal hernia was observed most commonly (4.38%, n=3), at a rate of 9.52% (n=2) with NS, and 10% (n=1) with IS.

Accompanying orthopedic problems were observed in 64.5% (n=40) of our patients. These were most commonly observed with NS (71.42%), followed by SS (70%), IS (60%), and CS (57.69%). The most common co-existing problem was lower extremity motor weakness (paresis/paralysis), seen in 25.80% of patients (n=16), and seen in 57.14% of NS cases, 20% of SS cases, and 7.69% of CS cases. Hip dysplasia was observed in 14.51% (n=9) of patients, and in 20% of SS, 15.38% of CS and 14.28% of NS cases. 24.1% of our patients (n=15) received surgery for orthopedic or other organ abnormalities.

Surgery for intraspinal abnormalities was required before the corrective surgery for the spinal deformation. For congenital curvatures with untreated dysraphism, surgical intervention may cause neurological functional deterioration. If the cord is fixed to a bone spicule (as in diastematomyelia) or distally (as in tethered cord), correction of the deformity may cause spinal cord injury. Deformity progression may also be caused by spinal dysraphism, not by vertebral abnormality. Evaluation with MRI is not indicated for all congenital curvatures, but must be conducted if progression is in a segmented area or neurological findings exist before surgery[25].

30% (n=9) of patients who had MRI scans for congenital spinal deformities had hidden intraspinal abnormalities, and 6.66% (n=2) had surgical intervention[34]. 21.7% (n=10) of the patients had neural axis abnormalities, and 80% (n=8) of those needed neurosurgical intervention (33). In IIS, the intraspinal abnormality incidence by MRI was 13% (n=7), and 71.4% (n=5) of those patients required neurosurgical operations[37].

For 29.03% (n=18) of our patients, neurosurgical operations were performed for intraspinal abnormalities. Subsequently, for vertebral deformities, 11.2% (n=7) of patients received VEPTR (n=4), growing rod (n=1), epiphysiodesis (n=1), or hemivertebra excision (n=1). No neurological deficits developed in our patients after surgery for correction of deformity.

If neural axis abnormalities are not detected, the surgical correction of scoliosis carries risks of neural sequelae[36]. Because of this need for the detection of spinal and extraspinal problems in EOS and the diagnosis of other organ abnormalities, MRI and assisting diagnostic methods have utmost importance, especially at the beginning of treatment. Johnston suggested that MRI should be a necessary reference for preoperative medical and surgical planning for neural axis abnormalities seen in EOS patients[29]. In this age group, the application of MRI is critical, although it has some associated risks, depending on the requirement for intravenous sedation or general anesthesia[16]. We observed no problems related to sedation or anesthesia during MRI scanning for our patients.

Due to the limited number of patients in our study with vertebral and accompanying pathologies (especially intraspinal abnormalities), more comprehensive studies with larger patient numbers are required. However, when EOS is first diagnosed, MRI is useful not only for the detection

of vertebral pathologies, but also for detecting other accompanying abnormalities. A multidisciplinary approach is required for understanding orthopedic, neurosurgical and other organ abnormalities and the accompanying problems, and planning treatment.

In conclusion:

- Scoliosis etiologies differ widely, but intraspinal and extraspinal abnormalities accompany other organ abnormalities frequently in EOS.
- Intraspinal abnormality of the vertebrae is most commonly seen in the NS subgroup of EOS, and the most common intraspinal abnormality modality is syringomyelia.
- Extraspinal abnormalities of the spine are most commonly observed in CS and the most common modality is hemivertebrae. Most frequently, thoracic vertebrae are affected.
- Orthopedic pathologies accompany EOS, and are most commonly seen with NS.
- The most commonly observed cardiovascular abnormality was ASD, and this was most frequently observed with SS.
- The most commonly observed urogenital abnormality was incontinence, and this was most frequently observed with SS.
- The most common pathology in other systems was inguinal hernia.
- Meticulous clinical and radiological examination is needed, together with MRI analysis, of the spine and other abnormalities.

REFERENCES

1. Akbarnia BA. Management Themes in Early Onset Scoliosis. *JBJS* 2007; 89-A(Supplement 1): 42-54.
2. Basu PS, Elsebaie H, Noordeen MH. Congenital spinal deformity. Acomprehensive assessment at presentation. *Spine* 2002; 27(20): 2255-2259.
3. Beals RK, Robbins JR, Rolfe B. Anomalies associated with vertebral malformations. *Spine* 1993; 18(10): 1329-1332.
4. Belmont PJ, Kuklo TR, Taylor KF, Freedman BA, Prahinski JR, Kruse RW. Intraspinal anomalies associated with isolated congenital hemivertebra: The role of routine Magnetic Resonance Imaging. *J Bone Joint Surg* 2004; 86-A(8): 1704-1710.
5. Benli IT, Uzumcugil O, Aydin E, Ateş B, Gürses L, Hekimoğlu B. Magnetic resonance imaging abnormalities of neural axis in Lenke type 1 idiopathic scoliosis. *Spine* 2006; 31(16): 1828–1833.
6. Bollini G, Launay F, Docquier PL, Viehweger E, Jouve JL. Congenital abnormalities associated with hemivertebrae in relation to hemivertebrae location. *J Pediatr Orthop* 2010; 19(1): 90-94.
7. Campbell RM Jr, Smith MD, Mayes TC, Mangos JA, Willey-Courand DB, Kose N, Pinero RF, Alder ME, Duong HL, Surber JL. The characteristics of thoracic insufficiency syndrome associated with fused ribs and congenital scoliosis. *J Bone Joint Surg* 2003; 85-A(3): 399-408.
8. Charry O, Koop S, Winter R, Lonstein J, Denis F, Bailey W. Syringomyelia and scoliosis: a review of twenty-five pediatric patients. *J Pediatr Orthop* 1994; 14(3): 309-17.
9. Citron N, Edgar MA, Sheehy J, Thomas DG. Intramedullary spinal cord tumours presenting as scoliosis. *J Bone Joint Surg* 1984; 66-B(4): 513-517.
10. Conner AN. Developmental anomalies and prognosis in infantile idiopathic scoliosis. *J Bone Joint Surg* 1969; 51-B(4):711-713.
11. Diab M, Landman Z, Lubicky J, Dormans J, Erickson M, Richards BS; members of the Spinal Deformity Study Group. Use and Outcome of MRI in the Surgical Treatment of Adolescent Idiopathic Scoliosis. *Spine* 2011; 36(8): 667–671.
12. Dickson RA. Conservative treatment for idiopathic scoliosis. *JBJS Br* 1985; 67(2): 176-181.
13. Dickson RA. Early-onset idiopathic scoliosis. In: Weinstein S (Ed.), *The Pediatric Spine: Principles and* Practice. Raven Press, New York 1994; pp: 421-429.
14. Dickson RA, Archer IA. Surgical treatment of late-onset idiopathic thoracic scoliosis: The Leeds procedure. *J Bone Joint Surg* 1987; 69-B(5): 709-714.

15. Dimeglio A. Growth of the spine before age 5 years. *J Pediatr Orthop* 1993; 1(2): 102-107.

16. Dobbs MB, Lenke LG, Szymanski DA, Morcuende JA, Weinstein SL, Bridwell KH, Sponseller PD. Prevalence of neural axis abnormalities in patients with infantile idiopathic scoliosis. *J Bone Joint Surg* 2002; 84-A(12): 2230-2234.

17. Dobbs MB, Weinstein SL. Infantile and juvenile scoliosis. *Orthop Clin North Am* 1999; 30(3): 331-341.

18. Drvaric DM, Ruderman RJ, Conrad RW, Grossman H,Webster GD, Schmitt EW. Congenital scoliosis and urinary tract abnormalities: Are intravenous pyelograms necessary? *J Pediatr Orthop* 1987; 7(4): 441-443.

19. Emery E, Redondo A, Rey A. Syringomyelia and Arnold Chiari in scoliosis initially classified as idiopathic: experience with 25 patients. *Eur Spine J* 1997; 6(3): 158-162.

20. Evans SC, Edgar MA, Hall-Craggs MA, Powell MP, Taylor BA, Noordeen HH. MRI of "idiopathic" juvenile scoliosis. A prospective study. *J Bone Joint Surg* 1996; 78-B(2): 314-317.

21. Fernandes P, Weinstein SL. Natural history of early onset scoliosis. *J Bone Joint Surg* 2007; 89-A(suppl 1): 21–33.

22. Gillingham BL, Ham RA, Behrooz BA. Early Onset Idiopathic Scoliosis. *J Am Acad Orthop Surg* 2006; 14(2): 101-112.

23. Gollogly S, Smith JT, Campbell RM. Determining lung volume with three-dimensional reconstructions of CT scan data: A pilot study to evaluate the effects of expansion thoracoplasty on children with severe spinal deformities. *J Pediatr Orthop* 2004; 24(3): 323-328.

24. Gupta P, Lenke LG, Bridwell KH. Incidence of neural axis abnormalities in infantile and juvenile patients with spinal deformity: Is a magnetic resonance image screening necessary? *Spine* 1988; 23(2): 206-210.

25. Hedequist D, Emans J. Congenital Scoliosis. *J Am Acad Orthop Surg* 2004; 12(4): 266-275.

26. Hedequist D, Emans J. Congenital scoliosis: a review and update. *J Pediatr Orthop* 2007; 27(1): 106-116.

27. Hooper G. Congenital dislocation of the hip in infantile idiopathic scoliosis. *J Bone Joint Surg* 1980; 62-B(4): 447- 449.

28. James JI. Idiopathic scoliosis; the prognosis, diagnosis, and operative indications related to curve patterns and the age at onset. *J Bone Joint Surg* 1954; 36-B(1): 36-49.

29. Johnston CE. Preoperative medical and surgical planning for early onset scoliosis. *Spine* 2010; 35 (25): 2239-2244.

30. Lewonowski K, King JD, Nelson MD. Routine use of magnetic resonance imaging in idiopathic scoliosis patients less than eleven years of age. *Spine* 1992; 17(6 Suppl): S109-116.

31. MacEwen GD, Winter RB, Hardy JH. Evaluation of kidney anomalies in congenital scoliosis. *J Bone Joint Surg* 1972; 54-A(7): 1451-1454.

32. McMaster MJ. Occult Intraspinal Anomalies and Congenital Scoliosis. *J Bone Joint Surg* 1984; 66-A(4): 588-601.

33. Mehta MH. The ribvertebra angle in the early diagnosis between resolving and progressive infantile scoliosis. *J Bone Joint Surg* 1972; 54-B(2): 230-243.

34. Mehta MH. Infantile idiopathic scoliosis. In: Dickson RA, Bradford DS (Eds.), *Management of Spinal Deformities*. Butterworths International Medical Reviews, London, 1984; pp: 101-120.

35. Muhonen MG, Menezes AH, Sawin PD, Weinstein SL. Scoliosis in pediatric Chiari malformations without myelodysplasia. *J Neurosurg* 1992; 77(1): 69-77.

36. Noordeen MH, Taylor BA, Edgar MA. Syringomyelia. A potential risk factor in scoliosis surgery. *Spine* 1994; 19(12): 1406-1409.

37. Ouellet JA, LaPlaza J, Erickson MA, Birch JG, Burke S, Browne R. Sagittal plane deformity in the thoracic spine: a clue to the presence of syringomyelia as a cause of scoliosis. *Spine* 2003; 28(18): 2147–2151.

38. Pahys JM, Samdani AF, Betz RR. Intraspinal anomalies in infantile idiopathic scoliosis: prevalence and role of magnetic resonance imaging. *Spine* 2009; 34(12): E434–438.

39. Prahinski JR, Polly DW Jr, McHale KA, Ellen- bogen RG. Occult intraspinal anomalies in congenital scoliosis. *J Pediatr Orthop* 2000; 20(1): 59-63.

40. Riseborough E, Wynne-Davies R. A genetic survey of idiopathic scoliosis in Boston, Massachusetts. *J Bone Joint Surg* 1973; 55-A(5): 74-82.

41. Schwend RM, Hennrikus W, Hall JE, Emans JB. Childhood scoliosis: clinical indications for magnetic resonance imaging. *J Bone Joint Surg* 1995; 77-A(1): 46-53.

42. Shen J, Wang Z, Liu J, Xue X, Qiu G. Abnormalities associated with congenital scoliosis: A retrospective study of 226 Chinese surgical cases. *Spine* 2013; 38 (10): 814-818.

43. Suh SW, Sarwark JF, Vora A, Huang BK. Evaluating congenital spine deformities for intraspinal anomalies with magnetic resonance imaging. *J Pediatr Orthop* 2001; 21(4): 525-531.

44. Whitaker C, Schoenecker PL, Lenke LG. Hyperkyphosis as an indicator of syringomyelia in idiopathic scoliosis: a case report. *Spine* 2003; 28(1): E16–20.

45. Wynne-Davies R. Infantile idiopathic scoliosis: Causative factors, particularly in the first six months of life. *J Bone Joint Surg* 1975; 57-B(2): 138-141.

46. Yngve D. Abdominal reflexes. *J Pediatr Orthop* 1997; 17(1): 105-108.

47. Zadeh HG, Sakka SA, Powell MP, Mehta MH. Absent superficial abdominal reflexes in children with scoliosis: An early indicator of syringomyelia. *J Bone Joint Surg* 1995; 77-B(5): 762-767.