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NEURAL AXIS ABNORMALITIES IN EARLY ONSET SCOLIOSIS

ERKEN BAŞLANGIÇLI SKOLYOZDA NÖRAL AKS DEFORMİTELERİ

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SUMMARY

Early onset scoliosis includes infantile idiopathic scoliosis, juvenile idiopathic scoliosis, and congenital scoliosis. The types of early onset scoliosis are neuromuscular scoliosis, syndromic scoliosis, and thoracic insufficiency syndrome. Spinal deformities in the growing spine may be observed alongside abnormalities of other organ systems, with neural axis abnormalities being one of the most common. The gold standard in the diagnosis of intraspinal abnormalities is MRI evaluation, and doctors dealing with scoliosis treatment should be cautious about the signs of asymptomatic neural axis abnormalities, in order to avoid the development of neurological deficit.

Key words: Neural axis deformities, syringomyelia, early onset scoliosis, MRI

Level of evidence: Review article, Level V

ÖZET

Erken başlangıçlı skolyoz infantil idiopatik skolyoz, juvenil idiopatik skolyoz ve konjenital skolyozu kapsar. Nöromusküler skolyoz, sendromik skolyoz ve torasik yetersizlik sendromu da erken başlangıçlı skolyoz tipleridir. Büyüyen bir omurgada spinal deformiteler sıklıkla nöral aks anomalileri olmakla beraber diğer organ anomalileri ile birliktelik gösterebilirler. MR görüntülemenin intraspinal anomali değerlendirmesinde altın standart olmasına rağmen, skolyoz tedavisi ile ilgilenen bir hekimin asemptomatik nöral aks anomalileri belirtileri hakkında bilgi sahibi olması gerekir.

Anahtar Kelimeler: Nöral aks deformiteleri, siringomiyeli, erken başlangıçlı skolyoz, MRI

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INTRODUCTION:

Early onset scoliosis is defined by the SRS as curvature of the spine in the lateral plane diagnosed before the age of 10. Infantile idiopathic scoliosis, juvenile idiopathic scoliosis, and congenital scoliosis are included within this definition. Neuromuscular scoliosis, syndromic scoliosis, and thoracic insufficiency syndrome are types of early onset scoliosis[9].

Spinal deformities in the growing spine may be observed alongside abnormalities of other organ systems, with neural axis abnormalities one of the most common abnormalities seen[11,18,24]. Neural axis abnormalities that are seen together with spinal deformities include Arnold-Chiari malformation, syringomyelia, hydromyelia, low-lying conus, tethered cord, tonsillary ectopia, and tumors. These abnormalities can be both symptomatic and asymptomatic. Doctors dealing with scoliosis treatment should be cautious about the signs of asymptomatic neural axis abnormalities[18], otherwise they may be faced with the risk of neurological deficit development,especially during scoliosis correction. In scoliosis patients, a neurological examination that involves motor, sensory, and reflex evaluations of the upper and lower extremities should be performed in detail. Hypertrophic reflexes, a unilateral superficial abdominal reflex, muscle atrophies, motor weakness, and sensory loss may be signs of neural axis abnormality. The risk of intraspinal abnormalities were found to be higher in patients with curves that were atypical and over 20°, that had rapid progression, and in the presence of lower back pain[21,30].

The gold standard in the diagnosis of intraspinal abnormalities is MRI evaluation. All the rates reported in the literature are the results of diagnostic studies performed using MRI[2,18,24].

INFANTILE AND JUVENILE IDIOPATHIC SCOLIOSIS:

The rate of encountering neural axis abnormalities in patients with infantile or juvenile idiopathic scoliosis with curves greater than 20° has been reported to be approximately 20%.

Although MRI evaluation of all idiopathic scoliosis patients is controversial, it is recommended that an MRI is taken in the presence of early onset (infantile and juvenile) scoliosis, male gender, an abnormal sagittal spine profile, atypical curve shapes (curves to the left), associated abnormalities or lower extremity deformities, rapid progression, and pain[6,11,18,24,30].

Wu et al. studied 64 patients diagnosed with idiopathic left-sided thoracic scoliosis and found neural axis abnormalities in 54% [33]. The neural axis abnormalities detected by MRI in patients previously diagnosed with idiopathic scoliosis were syringomyelia, Arnold-Chiari malformation, tonsillary ectopia, hydromyelia, spinal cord tumors, tethered spinal cord, diastematomyelia, and intraspinal lipoma. A study in England showed that among 72 patients under the age of 7 with idiopathic scoliosis, the rate of encountering neural axis abnormalities (syrinx and Arnold-Chiari type 1) was 11%[19]. Pahys et al. took MRI scans of 54 patients with infantile idiopathic scoliosis and normal neurological examinations and detected neurological axis abnormalities in seven patients (13%). Neurosurgical intervention was necessary in five patients (three cases of tethered cords and two Chiari malformations)[24].

In a study by Gupta et al., the incidence of neurological axis abnormalities in cases with infantile and juvenile spinal deformities was 17.6% [15]. Nakahara reported that the incidence of neurological axis abnormalities in idiopathic scoliosis for patients diagnosed before or after the age of 11 was 13.2% and 2.6%, respectively, and they were reported to be more common in King type 4 and 5 curves[22]. Inoue et al. performed MRI evaluation of 250 idiopathic scoliosis cases and found neural axis abnormalities in 44 patients. Neurological examinations were abnormal for 26 of these patients. The abnormalities detected were Chiari malformation together with syringomyelia in 22 patients, syringomyelia and tonsillar ectopia in two patients, Chiari malformation in 13 patients, tonsillar ectopia in six patients, and low-lying conus medullaris in one patient [18].

There have been theories stating that Chiari malformation together with syringomyelia may result in scoliosis by causing muscle imbalance. However, studies by Eule and Flynn have stated that decompression of Chiari malformations may lead to an increase in spinal deformity[12,13].

Intraspinal abnormalities detected in patientswith idiopathic scoliosis should be evaluated with a multidisciplinary approach. Our approach to abnormalities that require surgical treatment is to simultaneously perform treatment of neural axis abnormalities and scoliosis correction. This eliminates the need for a second operation. Various studies have supported the suggestion that this method can be used safely and effectively[23,28]. In their experience with concurrent tethered cord release and correction of scoliosis and kyphosis with spinal fusion, Mehta et al. observed that a single-stage approach was safe, with no significant operative or postoperative morbidity. When compared with a two-stage approach carried out by the same operating team, the concurrent approach was associated with less blood loss, shorter operative time, a shorter hospital stay, better deformity and tethered cord correction, and fewer perioperative complications[20].

The use of intraoperative neuromonitoring is necessary to decrease surgical risks. Spinal cord monitoring is able to accurately detect significant deviations in patients with neural axis abnormalities[10].

CONGENITAL SCOLIOSIS:

The rate of intraspinal abnormalities in congenital scoliosis cases is 30–40%. This high rate results from the close proximity of the vertebral column to the spinal cord and organs developing from the mesoderm during embryonic development. Growth and increase of the curvature may result in progressive neurological deficits in these patients. The risk of neurological injury during surgical correction of congenital scoliosis is also much higher than for idiopathic cases.

Positive physical examination findings may be seen in approximately half of the cases with abnormalities. Therefore, MRI evaluation of the whole vertebral column must be performed routinely for all congenital scoliosis patients[11,17,29].

The most common intraspinal abnormalities seen in congenital spinal deformities are tethered cord lesions and diastematomyelia. The next most common lesions are syringomyelia, thickened filum terminale, low-lying conus, intra- and extradural masses, Chiari malformation, arachnoid cyst, and Dandy Walker malformation[11,25,29].

Physical examination findings, such as hairy patches or pigmentation over the spine, bladder symptoms, foot deformities, an asymmetrical abdominal reflex, or abnormal posterior column sensitivity are warning signs for an intraspinal abnormality[11]. In a study by Shen et al., 43% of 226 patients with scoliosis had intraspinal abnormalities. The most commonly seen abnormality was diastematomyelia. The rates of encountering intraspinal abnormalities were found to be higher in segmentation defects and mixed defects compared to formation defects, and in thoracic hemivertebra cases compared to lumbar hemivertebra cases. A clinical sign was present in 51% of cases with intraspinal abnormalities[29]. In a study by Suh et al., 13 of 41 congenital scoliosis patients were detected to have intraspinal abnormalities, with tethered cord as the most common pathology. There were no clinical signs in 31% of patients[30].

Belmont et al. evaluated 160 congenital scoliosis patients with MRI and found similar rates of intraspinal abnormalities in isolated hemivertebra and complex deformities (28% and 21%). The authors reported that patient history and clinical examination were not predictive, and all patients, even those with an isolated hemivertebra, should be evaluated by MRI[4].

The approach classically advocated for such patients is first to perform surgery for intraspinal pathologies, and then to perform surgery for correction and stabilization of the deformity 3–6 months later. In our opinion, the second session of surgical treatment has some associated disadvantages, related to the patient's general medical condition and the site of surgery.

Simultaneous surgical treatment for the congenital deformity and the intraspinal abnormality does not involve significant complications and seems to be an alternative and a safe treatment option[16]. Oda et al. lists the advantages of simultaneous surgery as including a single anesthetic exposure and single hospitalization, no surgical dissection through an area of the spine traumatized by a previous laminectomy, improved resource utilization and lower cost, and patient comfort and convenience[23].

NEUROMUSCULAR SCOLIOSIS:

Loss of muscle power or voluntary control, or sensory deficits such as deep sensation, may result from the development of a deformity in the growing spine. The causes of neuromuscular scoliosis may be classified into two groups: neuropathic and myopathic. Numerous disorders in which the neural axis is primarily involved may lead to early onset scoliosis. Traumatic spinal cord injuries, tethered cord syndrome, syringomyelia, and spinal cord tumors are among the intraspinal anomalies that result in scoliosis.

The incidence of scoliosis rises to 100% for spinal cord injuries that have occurred before maturation of the skeletal system[3,11,32].

Scoliosis that occurs in myelomeningocele patients is also assessed in this group. In studies by Trivedi et al., structural curves over 20˚ were found at a rate of 52%[31]. It is crucial to determine the underlying pathology as a priority, and then structure the management plan together with scoliosis treatment[5]. Although the use of intraoperative neuromonitoring is possible in these cases, it should be remembered that reliable results may not always be obtained. According to Dicindio et al., both transcranial electric motor and posterior tibial nerve somatosensory evoked potentials can be monitored reliably in most patients with neuromuscular scoliosis[8].

Accadbled et al. advocated that the use of a single epidural electrode allowing alternate recording of somatosensory evoked potentials and spinal cord stimulation was a safe and valid method for intraoperative monitoring[1].

SYNDROMIC SCOLIOSIS:

Children with neurofibromatosis type 1 who develop scoliosis may be diagnosed with either dystrophic (degenerative) or non-dystrophic forms of the condition. Non-dystrophic scoliosis resembles idiopathic adolescent scoliosis, with similar types of curvature and curve patterns. Dystrophic neurofibromatosis may result in short and sharp spinal curves with early onset and rapid progression, eventually culminating in severe deformity. Dystrophic curves may occur together with kyphosis and high rates of neurological injury.

Due to the poor quality of the bone, the characteristics of dystrophic scoliosis also include a number of bone abnormalities, including thinning of the ribs, significant rotation of the vertebrae, vertebral wedging, and erosion of the vertebrae by the spinal fluid. Meningocele, pseudomeningocele, dural ectasia, dumbbell lesions and intraspinal tumors may be seen in patients with neurofibromatosis[7,11,14,27,32].

In Marfan's syndrome, intraspinal abnormalities may be observed together with scoliosis. In a study by Robins, 35 of 64 patients with Marfan's syndrome had scoliosis, and scoliosis had begun in the infantile and juvenile age periods for 44% of these patients[27]. Pyerits et al. found widening of the lumbosacral spinal canal in 63% of 57 patients with Marfan's syndrome[26]. Dural ectasia develops as a result of fibrillin deficiency in Marfan's syndrome. It is known that the laminae are thinned in areas of dural ectasia, and so the use of hooks must be avoided. This is seen frequently at the most caudal part of the lumbosacral spinal column. A widened interpedicular distance, increased vertebral scalloping, and an increased sagittal diameter can indicate dural ectasia in patients with Marfan's syndrome. The clinical findings of this condition may manifest as lumbar pain or headache. In symptomatic dural ectasia, posterior laminectomy has sometimes been implemented as a means of relieving the back pain[11].

REFERENCES

1. Accadbled F, Henry P, de Gauzy JS, Cahuzac JP. Spinal cord monitoring in scoliosis surgery using an epidural electrode. Results of a prospective, consecutive series of 191 cases. *Spine* 2006; 31(22): 2614-2623.
2. Akhtar OH, Rowe DE. Syringomyelia-associated scoliosis with and without the Chiari I malformation. *J Am Acad Orthop Surg* 2008; 16(7): 407-417.
3. Allam AM, Schwabe AL. Neuromuscular scoliosis. *PM R* 2013; 5(11): 957-963.
4. Belmont PJ Jr, 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. Cassar-Pullicino VN, Eisenstein SM. Imaging in scoliosis: what, why and how? *Clin Radiol* 2002; 57(7): 543-562.
6. Crawford AH, Herrera-Soto J. Scoliosis associated with neurofibromatosis. *Orthop Clin North Am* 2007; 38(4): 553-562.

7. Cardoso M, Keating RF. Neurosurgical management of spinal dysraphism and neurogenic scoliosis. *Spine* 2009; 34(17): 1775-1782.
8. DiCindio S, Theroux M, Shah S, Miller F, Dabney K, Brislin RP, Schwartz D. Multimodality monitoring of transcranial electric motor and somatosensory evoked potentials during surgical correction of spinal deformity in patients with cerebral palsy and other neuromuscular disorders. *Spine* 2003; 28(16): 1851-1855.
9. Early onset scoliosis. <http:// www.srs.org/ patient_and_ family/ scoliosis/ early_ onset_scoliosis>
10. Elsebaie HB. Co-morbidities associated with early onset scoliosis. In: Akbarnia A, Yazici M, Thompson GH (eds.) *The Growing Spine*. Springer, Berlin, 2011; pp: 69- 78.
11. El-Hawary R, Sucato DJ, Sparagana S, McClung A, Van Allen E, Rampy P. Spinal cord monitoring in patients with spinal deformity and neural axis abnormalities: a comparison with adolescent idiopathic scoliosis patients. *Spine* 2006; 31(19): E698-E706.
12. Eule JM, Erickson MA, O'Brien MF, Handler M. Chiari I malformation associated with syringomyelia and scoliosis: a twenty-year review of surgical and nonsurgical treatment in a pediatric population. *Spine* 2002; 27(13): 1451-1455.
13. Flynn JM, Sodha S, Lou JE, Adams SB Jr, Whit- field B, Ecker ML, Sutton L, Dormans JP, Drummond
14. DS. Predictors of progression of scoliosis after decompression of an Arnold Chiari I malformation. *Spine* 2004; 29(3): 286-292.
15. Funasaki H, Winter RB, Lonstein JB, Denis F. Pathophysiology of spinal deformities in neurofibromatosis. An analysis of seventy-one patients who had curves associated with dystrophic changes. *J Bone Joint Surg* 1994; 76-A(5): 692-700.
16. 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* 1998; 23(2): 206-210.
17. Hamzaoglu A, Ozturk C, Tezer M, Aydogan M, Sarier M, Talu U. Simultaneous surgical treatment in congenital scoliosis and/or kyphosis associated with intraspinal abnormalities. *Spine* 2007; 32(25): 2880-2884.
18. Hensinger RN. Congenital scoliosis: etiology and associations. S*pine* 2009; 34(17): 1745-1750.
19. Inoue M, Minami S, Nakata Y, Otsuka Y, Takaso M, Kitahara H, Tokunaga M, Isobe K, Moriya H. Preoperative MRI analysis of patients with idiopathic scoliosis: a prospective study. *Spine* 2005; 30(1): 108-114.
20. Koç T, Lam KS, Webb JK. Are intraspinal anomalies in early onset idiopathic scoliosis as common as once thought? A two centre United Kingdom study. *Eur Spine J* 2013; 22(6): 1250-1254.
21. Mehta VA, Gottfried ON, McGirt MJ, Gokaslan ZL, Ahn ES, Jallo GI. Safety and efficacy of concurrent pediatric spinal cord untethering and deformity correction. *J Spinal Disord Tech* 2011; 24(6): 401-405.
22. Morcuende JA, Dolan LA, Vazquez JD, Jirasirakul A, Weinstein SL. A prognostic model for the presence of neurogenic lesions in atypical idiopathic scoliosis. *Spine* 2004; 29(1): 51-58.
23. Nakahara D, Yonezawa I, Kobanawa K, Sakoda J, Nojiri H, Kamano S, Okuda T, Kurosawa H. Magnetic resonance imaging evaluation of patients with idiopathic scoliosis: a prospective study of four hundred seventy-two outpatients. *Spine* 2011; 36(7): E482-E485.
24. Oda JE, Shah SA, Mackenzie WG, Akbarnia BA, Yazici M. Concurrent tethered cord release and growing- rod implantation-is it safe? *Global Spine J* 2012; 2(4): 207-212.
25. Pahys JM, Samdani AF, Betz RR. Intraspinal anomalies in infantile idiopathic scoliosis: prevalence and role of magnetic resonance imaging. *Spine* 2009; 20; 34(12): E434-E438.
26. Prahinski JR, Polly DW Jr, McHale KA, Ellenbogen RG. Occult intraspinal anomalies in congenital scoliosis. *J Pediatr Orthop* 2000; 20(1): 59-63.
27. Pyeritz RE, Fishman EK, Bernhardt BA, Siegelman SS. Dural ectasia is a common feature of the Marfan syndrome. *Am J Hum Genet* 1988; 43(5): 726-732.
28. Robins PR, Moe JH, Winter RB. Scoliosis in Marfan's syndrome. Its characteristics and results of treatment in thirty-five patients. *J Bone Joint Surg* 1975; 57-A (3): 358-368.
29. Samdani AF, Asghar J, Pahys J, D'Andrea L, Betz RR. Concurrent spinal cord untethering and scoliosis correction: case report. *Spine* 2007; 32(26): E832-E836.
30. 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.
31. Suh SW, Sarwark JF, Vora A, Huang BK. Evaluat- ing congenital spine deformities for intraspinal anomalies with magnetic resonance imaging. *J Pediatr Orthop* 2001; 21(4): 525-531.
32. Trivedi J, Thomson JD, Slakey JB, Banta JV, Jones PW. Clinical and radiographic predictors of scoliosis in patients with myelomeningocele. *J Bone Joint Surg* 2002; 84-A (8): 1389-1394.
33. Vialle R, Thévenin-Lemoine C, Mary P. Neu- romuscular scoliosis. *Orthop Traumatol Surg Res* 2013; 99(1-Suppl): S124-S139.
34. Wu L, Qiu Y, Wang B, Zhu ZZ, Ma WW. The left thoracic curve pattern: a strong predictor for neural axis abnormalities in patients with "idiopathic" scoliosis. *Spine* 2010; 35(2): 182-185.