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CURRENT TREATMENT APPROACHES OF SPINE DEFORMITY ASSOCIATED WITH INTRASPINAL PATHOLOGIES

İNTRASPİNAL PATOLOJİLERİN EŞLİK ETTİĞİ OMURGA DEFORMİTELERİNDE GÜNCEL TEDAVİ YAKLAŞIMLARI

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SUMMARY

Pathologies of the spinal cord may be associated with scoliosis. When the spine deformity is corrected, neurological deficits due to spinal cord tension may develop. For this reason, surgical procedures may be required for intraspinal pathology. In this study, we evaluate the treatment approaches for spinal deformities with a high incidence of spinal abnormalities.

Key Words: Scoliosis; Intraspinal anomalies; Chiari malformation; Tethered cord; Magnetic resonance imaging

Level of Evidence: Review Article, Level V

ÖZET

Skolyozla birlikte spinal kord patolojileri birlikte görülebilir. Omurga deformitesi düzeltilirken spinal kordda gerilmeye bağlı nörolojik defisit gelişebilir. Bunu için öncelikle intraspinal patolojiye yönelik cerrahi işlemlere gerek duyulabilir. Bu çalışmada, intraspinal anomali insidansının yüksek bulunduğu omurga deformitelerinde tedavi yaklaşımları incelenmiştir.

Anahtar Kelimeler: Skolyoz; intraspinal anomali; Chiari malformasyonu; tethered kord; manyetik rezonans görüntüleme

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INTRODUCTION:

Spine deformities, such as scoliosis and kyphosis, can be observed together with Chiari malformation (CM), tethered cord and other intraspinal pathologies. Deformities should be evaluated in terms of spine curvature and spinal canal pathologies. Canal pathologies are rarely presented with clinical signs. When planning the treatment of patients, it is useful to detect these pathologies as soon as possible. Magnetic resonance imaging (MRI) is used for scoliosis patients with intraspinal pathologies who don't have any clinical signs.

Intraspinal anomalies are often observed in patients, especially those with congenital, infantile and juvenile onset scoliosis (20%)[13,17,22]. In patients with adolescent idiopathic scoliosis (AIS), the risk of the incidence of intraspinal anomalies increases with abnormal neurological examination signs, left curvature of the spine, and kyphotic deformity at the apical site[42,51,60.] The most common intraspinal anomalies associated with scoliosis are CM and/or syringomyelia[65-66]. Other common anomalies are tethered cord, lipoma and diastematomyelia. Vermis herniation is observed due to the herniation of the cerebellar tonsils down from the foramen magnum in Chiari malformation type I (CM I), and due to prolapsus of spina bifida and rhombencephalon in Chiari malformation type II (CM II). In MRI, CM is defined as >4–5 mm herniation of the cerebellar tonsils from the foramen magnum to the caudal[44]. In patients with CM, 50–90% scoliosis and 15–65% scoliosis in CM I patients has been observed[6]. The mechanism by which scoliosis occurs with CM or syringomyelia has not yet been determined. In some patients with scoliosis and syringomyelia, denervation of the paraspinal muscles can occur. However, no relationship between the amount of prolapse of the paraspinal muscles and the size of the syrinx and cerebellar tonsil herniation has been observed[60]. It should be remembered that scoliosis can develop without CM and syringomyelia[48].

Surgical correction of scoliosis increases the risk of neurological damage caused by cord tension[10,42]. Detailed neurosurgical evaluation is recommended before the treatment of deformity. If there is any intraspinal pathology, neural axis malformations should be primarily treated, due to the risk of neurological disruption during the correction of deformity[21,47,63]. In tethered cord syndrome (TCS), progressive sensorimotor deficit, fecal and/or urinary incontinence, and musculoskeletal deformities in the lower extremities are observed, due to adhesion of the distal spinal cord or filum to adjacent structures. Distal cord tension can be idiopathic. It can be observed with intraspinal anomalies such as lipoma, and can be related with the adhesion of distal spinal cord residues during the repair of dysraphism in patients with myelopathy. Relaxation of the cord surgically is recommended the patients' pain symptoms get worse, progressive leg deformities or acute correction of spasticity or associated spinal deformity. Neurophysiological intraoperative monitorization decreases the risk of undesired nerve root or spinal cord injuries during relaxation procedures, and indicates any change in cord function during the correction of spinal deformity[57,69]. In the presence of a tethered cord, loosening the affected cord is one of the best methods for preventing or correcting scoliosis[44]. In the literature, the rate of which tethered cord occurs together with CM I has been reported as 14%[1,26,30,52]. Therefore, it is recommended that intervention is necessary before the surgical decompression of intraspinal anomalies such as CM[23,52,56,61].

While spine deformity is being corrected, surgical processes in the craniocervical or lumbar regions can be required to prevent neurological deficit due to tension of the spinal cord. For the correction of spinal deformity, it is necessary to improve treatment techniques and to prefer safe methods in patients with a high neurological risk. Therefore, treatment approaches applied to spine deformities associated with intraspinal pathologies were investigated.

DISCUSSION:

Intraspinal pathologies:

About 20% of patients with congenital scoliosis can show intraspinal anomaly (Chiari malformation, syringomyelia, diastematomyelia and tethered cord)[6,18]. Similarly, spinal cord anomalies were observed in MRI of 20% of patients with infantile and juvenile scoliosis[(18,23)]. However, this incidence is quite low for idiopathic scoliosis[15,17,28]. Most of the intraspinal anomalies detected with scoliosis do not require surgery, such as small syrinx or minimal cerebellar ectopy[58].

In a prospective study including 327 AIS patients, were detected abnormal MRIs in seven patients, none of which required a neurosurgical approach[17]. The incidence

of intraspinal anomalies requiring surgical correction is greater in infantile scoliosis. While Gupta stated that three out of six patients required surgery[24].

Dobbs reported that eight of ten anomalies received surgery[18].

Conservative treatment of adolescent spine curves is still controversial. It is known that bracing is not effective for the treatment of scoliosis associated with CM I or syringomyelia[65]. In the treatment of spine deformities such as scoliosis and/or kyphosis associated with spinal pathologies (CM I, tethered cord, other intraspinal pathologies), the aim is primarily to perform surgical treatment of intraspinal pathology and then to correct the spine deformity with a second surgery. There have been studies investigating the applicability of direct posterior or other methods for spinal deformity correction without a neurosurgical approach.[61,66]. In the same session, surgical treatment for both pathologies was applied sequentially. According to the degree of scoliosis and CM I, Xie showed that these adolescent patients can be treated with vertebral column resection from the posterior alone, or posterior instrumentation without suboccipital decompression[68]. Although associated scoliosis is corrected by the interventions performed for intraspinal anomalies, this is not possible for people who are elderly or admitted late[42,46,47]. In recent years, it has become possible for both of these procedures to be applied under a single anesthesia, due to the increased opportunities for neuromonitorization. Similar applications have also been defined for congenital scoliosis[51].

Tethered Cord:

When scoliosis is corrected, the risk of neurological injury increases due to tension of the cord. Therefore, distal relaxation of the tethered cord is required. Generally, this is applied 4–6 weeks before scoliosis correction. During this healing period, the possibility of re-adhesion decreases[58]. The other approach is applied for deformity correction from a minimum of six weeks until six months after the relaxation procedure. The aim is to allow the curves to be stabilized and corrected due to relaxation, or to decrease the risk of spinal cord injury during correction. However, there are no absolute proofs supporting this in the literature. Samdani and Ayvaz investigated the necessity of gradual surgery in their studies[4,58]. In order to decrease the risk of neurological complication before the treatment of spine deformity associated with intraspinal anomalies requiring intervention, it has been recommended that these pathologies should be treated. For a male patient aged 15, with low lying conus at the L4 level in MRI, progressive kyphoscoliosis, lower extremity paresthesia and urinary incontinence, Samdani applied simultaneous surgery, including relaxation of the stretched spine and posterior fusion from the T2 level to the pelvis. When simultaneous relaxation of the spinal cord and correction of scoliosis with intraoperative neuromonitorization is compared to gradual procedures in selected patients, this has been shown to be an applicable option[58]. For tethered cord, treatment of the affected cord by relaxation is suggested to prevent or correct scoliosis[12].

The effectiveness of the relaxation of a tethered cord for kyphosis has been less well defined, but it has been shown that small kyphotic curves can recover without progression of deformity. Tethered cord relaxation and simultaneous scoliosis or kyphosis deformity correction has only been described in two patients. Samdani performed surgical deformity correction with tethered cord relaxation and fusion from the T2 level to the pelvis in a single stage in a patient aged 15 with progressive scoliosis with fatty filum and tethered spinal cord[58]. Hamzaoğlu et al. performed surgery in a single session for 21 patients with an average age of 13[26]. For all patients, it was observed that correction of deformity was maintained after 6.8 years, and the rates of complications were similar.

Chiari Malformation:

Charry stated that iatrogenic factors need to be considered for patients with scoliosis associated with CM I or syringomyelia, due to the high risk of neural tissue damage during corrective procedures. He demonstrated that prolapse of the cerebellar tonsils caused hypertension of the cord, an expanded syrinx reduced blood circulation by compressing the spinal cord, and the pressure of the cerebrospinal fluid significantly changed intraoperatively. Therefore, he reported that decompression of craniocervical bone and fossa elements and syrinx shunt should be applied, before spinal fusion and correction procedures can be applied with confidence regarding the neurological condition after 3–6 months[13].

In progressive scoliosis with CM I, decompression applied at an early age and when the curvature angle is small has a positive effect on the correction of scoliotic curvature. In children with mild scoliosis and CM I malformation, either stabilization or correction of spine deformities with neurosurgery alone has been proven in the literature[21]. On the other hand, results are not very good for adolescents with large curves. Brockmeyer showed that suboccipital decompression and duraplasty provided correction or stabilization of the curve for many patients with scoliosis together with CM I and syringomyelia (62%). In children aged over 12 and whose curve is more than 50°, he stated that the chance of correcting the curve is low, and this increases with suboccipital decompression and duraplasty for patients aged less than 10 with small curves[11]. When following up patients after neurosurgery, Flynn found that an age greater than 11, the presence of neurological symptoms, rotation of the vertebral body, a double scoliosis curve and the observation of large curves (>50° kyphosis or >40° curve) caused the progression of scoliosis[23]. Bhangoo reported the mean age of patients requiring scoliosis correction surgery as 158 months and the mean Cobb angle as 76°, for 13 patients diagnosed with symptomatic CM I and clinical scoliosis who received craniovertebral decompression[7]. Sengupta demonstrated that 37,5% of 16 patients' curves were corrected, and 71.4% of them were under the age of 10 during hindbrain decompression[65]. Muhonen published that scoliosis regressed in patients with a preoperative curve of greater than 40° for patients aged under 10 years old[45]. Eule reported correction in five out of five patients who received decompression when they were under the age of 8 years old[21].

Myelodysplasia:

It is known that Chiari and tethered cord pathologies are often encountered in patients with myelomeningocele. In order to prevent further progression of current problems, a surgical approach with diagnosis at an early age is important in terms of solving deformity and extra neurological problems.

Myelomeningocele (myelodysplasia) is associated with scoliosis, kyphosis and lordosis at high rates in children[37]. The incidence of scoliosis associated with myelomeningocele has been reported to be between 62% to 90% veya 62-90% [50,59]. Raycroft and Curtis reported the incidence of spine deformities as 62% in patients with myelomeningocele[45]. Associated spinal deformities are generally progressive and affect the prognosis of the patient. Vertebral anomalies cause skeletal deformities and make the clinical landscape complex. The most obvious and common congenital anomaly is an inadequate posterior arch of the lumbosacral vertebra. These anomalies affect the treatment of scoliosis and kyphosis from many aspects[2,32]. Other congenital malformations include hemivertebrae, butterfly vertebrae, diastematomyelia and unsegmented bar[67]. When spine curves are compared with other developmental anomalies in patients with myelomeningocele, they are frequently observed in teens[45]. They can occur between the age of 2 and 3, and can be very serious by the age of 7[2,50,64].

In pediatric patients with myelodysplasia, the most common reason for neurological disruption is shunt dysfunction, with the second most common reason being symptomatic tethering[5,8,12,14]. For 25 cases with or without congenital syndrome who received tethered cord relaxation, 64% correction in motor function, 14% recovery in bladder and intestinal function, and 29% reduction in pain were detected for the patients without congenital syndrome. In 18% of the patients with congenital syndrome, recovery of bladder and intestinal function was detected. In both groups, no deterioration of motor functions, bladder and intestinal functions or pain were observed. Postoperatively, improvement of symptoms was clearly revealed in 6–8 weeks[32]. Similar results were also published by Quinones-Hinojosa[56]. He evaluated 15 adult patients retrospectively, and observed recovery of bladder and intestinal functions in 46% of the patients, recovery of back pain in 39% of patients, and recovery of motor functions in 31% of the patients, while loss of bladder control and postoperative worsening of back pain was detected in 8% of the patients.

In a retrospective study investigating 50 children with TCS, Daszkiewicz found that a significant clinical recovery was obvious in the lower extremities instead of the syrinx in 32.2% of the patients, and the degree of relaxation was the most obvious factor affecting the results[16]. In a large single-center clinical series, it was published that 23% of patients with myelomeningocele and spinal cord tension received surgery, and scoliosis correction was performed after relaxation procedures for the patients with myelomeningocele[8]. For patients with scoliosis and myelomeningocele, it has been suggested that the tension is relaxed before the correction of deformity, due to large and progressive curves[12, 25].

Syringomyelia:

In patients with scoliosis and syringomyelia, Qiu demonstrated that it was safe to correct spine deformity without neurosurgical decompression[55]. He obtained successful results with a posterior or combined anterior-posterior approach by detecting the last safe correction amount with Halo traction or radiographs taken from the fulcrum. Osteotomy alone with pedicle instrumentation, or deformity correction without vertebral column resection (VCR), was applied to cases with mild or moderate spine curve and no neurological deficit. In adolescent scoliosis with large curves and obvious progression, Xie suggested correction of the spine deformity and fusion. In 13 patients with CM I and adolescent scoliosis, he compared seven patients treated posteriorly with complete single-stage without suboccipital decompression, and six patients whose deformity was corrected and posterior pedicle screw fixation alone was applied. He found the mean Cobb angle as <90° (a mean scoliosis of 77.3° and kyphosis of 44°). Also, he found the postoperative correction rates to be 60.8% and 53.4% in the coronal and sagittal planes, respectively, for patients with CM who had no neurological deficit and were treated with only posterior instrumentation and correction of deformity. In the group with VCR, the mean Cobb angle was found to be greater than 90° for scoliosis and kyphosis (a mean scoliosis of 100.1° and kyphosis of 97.1°), and the correction rate was 63.3% for scoliosis and 71.1% for kyphosis. In this study, no neurosurgical processes were performed for CM. As a result, he concluded that suboccipital decompression is not always necessary before the correction of spine deformity in adolescent patients with CM I together with scoliosis[68].

Bradley performed initial neurosurgical decompression for 13 adolescent patients. While thoracic scoliosis was 46° (29–69°) and kyphosis was 71° (31–119°), the mean correction rate was found to be 48% (6–83%) after deformity surgery[9].

For asymptomatic syringomyelia, many authors support careful observation instead of prophylactic neurological operations[27]. In scoliosis, the effects of applying a direct shunt for isolated syringomyelia (without CM) are not clear. Some studies find it useful[59], while other studies do not[46].

Neuromonitorization:

During spinal surgery, monitorization with motor-excited potentials (MEP) provides evaluation of the intraoperative integrity of the corticospinal way[17]. Bridwell reported that spinal cord potentials cannot always show neurological deficits, and although they supported and applied the routine wake-up test after spinal correction procedures, they stated that it is a waste of time and reduces the maximum efficiency of deformity correction, and monitorization should be maintained[10]. TCS develops due to adhesion of the spinal cord or residues of the spinal cord to adjacent structures, is often related to spinal dysraphism, and generally involves the distal portion of the spinal cord. Therefore, as the cord naturally rises during growth, tension is applied to the cord due to this movement. Theoretically, these repetitive tensions can cause potentially irreversible dysfunction and structural damage to the spinal cord, the development of progressive motor and sensorial clinical signs in the lower extremities, pain, dysfunction of the bladder and/or intestine, and muscle-skeletal deformities such as scoliosis. When progressive symptoms can be seen or correction of an obvious spinal deformity is considered, surgery is required. As the normal anatomy changes due to the tethering, surgical repair can cause undesired injuries to the nerve roots. Because of this, neurophysiological intraoperative monitorization (NIOM), electromyelography, somatosensorial evoked potentials (SEP) and transcranial evoked potentials (TCMEP) are used to minimize the risk of iatrogenic neurological injury and postoperative deficit[54]. The recovery of neurological functions has been reported after relaxation, and the result of the treatment was related to the degree of relaxation[34,38,54]. NIOM is an integral part of TCS surgery due to the risk of nerve root injuries caused while the filum is isolated, dissected and cut. Typically, this is suitable for patients with progressive surgical signs and symptoms. Indications for surgery are pain, scoliosis with progressive orthopedic deformities, neurological deficit and spasticity.

Simultaneous surgery:

The improvements to neuromonitorization techniques make simultaneous surgery for these procedures possible[35-36,41,48]. For 15 sequential pediatric patients, retrospectively, Mehta published the results of simultaneous spinal cord untethering and deformity correction with fusion for scoliosis and/or kyphosis.

Clinically and radiologically, the initial values, operation details, morbidity and postoperative results were evaluated. He compared the results of this group with 21 patients who received scoliosis correction following a two-stage untethering surgery[43]. The mean corrections were 27% and 50% for scoliosis and kyphosis, respectively, and this was found within the correction range obtained for those procedures[4,40,58]. Mehta observed that simultaneous tethered cord relaxation and spinal fusion in a single-stage approach for the correction of scoliosis and kyphosis is clearly safe, with no operative or postoperative morbidity[43]. When compared with a two-stage approach performed by the same surgical team, the simultaneous approach resulted in less blood loss, a shorter operation time, a shorter hospitalization period, better deformity and tethered cord correction, and fewer perioperative complications[41]. While inadequate fusion for patients with intraspinal anomalies and spine deformities was observed in 12–50% of cases, pseudoarthrosis was observed in only one patient (6.7%) in this series[19,24,31-33]. Barley published a case showing sudden intraoperative improvement in weak neurophysiological responses in the upper extremities after relaxation and correction of scoliosis was simultaneously applied, and tethered cord surgery was applied for myelodysplasia, Chiari II malformation, severe scoliosis and tethered cord at the lumbar level. A harmony between the monitorization changes in the patient and the clinical improvement was noticed postoperatively[5]. There have been various studies defining progression of curvature after tethered cord relaxation[48-49, 55-60] and these patients can benefit from simultaneous tethered cord relaxation and spine fusion for correction of the deformity. According to Pierz and McLone, spine fusion after untethering is necessary for progressive deformity for more than 80% of patients with large curves (>40° and 50°)[41,51]. Mehta observed the incidence of progression requiring fusion as 83% in patients with an immature skeleton and curves >40° (Risser 0–2)[43].

For the correction of scoliosis and/or kyphosis, spine fusion and simultaneous tethered cord relaxation can be an effective and safe method for patients with possible progression of deformity[48]. It can minimize the risks that could be caused by a second operation, especially for patients with an immature skeletal system, rapid progression of curvature and a large curve, and also in cases with increased scarring and neurological injury. Earlier simultaneous interventions aim to improve the results of skeletal deformity correction during tethered cord relaxation. For small curves, stable curves, or for children with a skeletal system that is possibly stable and mature, a simultaneous approach is contraindicated. For this approach, it is important to choose the patients properly and to avoid any unnecessary approaches. While making the decision, the main factors can be curves of more than 40°, an immature skeletal system, and thoracic curve, lower lumbar and sacral motordysfunction[60]. However, patients that will benefit from a simultaneous approach should be chosen carefully[48].

Complications:

Neurological complications related to scoliosis surgery have been reported[13,62,70]. In a case with transient neurological deficit, Tomlinson observed headache, dystaxia and neck pain two weeks after spinal fusion, and detected syringomyelia with MRI[67]. Despite the application of laminectomy, Schlesinger observed no correction after scoliosis surgery in a patient with paraplegia[64].

Suggestions:

Xie suggested that primary correction of deformity should be performed gradually, by compression from the convex during correction, and stated that the remaining curve angle in preoperative convex bending radiographies shows the last point of correction and plays a positive role in the protection of the spinal cord[70]. Shrinkage and bending of the dural sac of more than 2–3 cm should be considered, and it has been stated that more compression can cause neurological pressure. In animal experiments, Kawahara demonstrated that reduction of up to 1/3 of the height of the vertebra caused no negative changes to the spinal cord[31].

CONCLUSION

Neural canal abnormalities can be encountered more frequently with congenital or neuromuscular scoliosis than other spine deformities. It is important to diagnose intraspinal pathologies early and to follow up scoliosis closely. Sometimes, intraspinal pathologies reveal no neurological signs. MRI, in particular, plays an important role in the diagnosis of these pathologies before scoliosis surgery. Intraspinal pathologies should be diagnosed and treated before surgical correction of curves. One of the most important aims of the surgical technique is to

apply correction without stretching the spinal cord. The application of neuromonitorization increases the safety of surgery during instrumentation and correction.

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