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Simultaneous Surgical Treatment in Congenital Scoliosis and/or Kyphosis Associated With Intraspinal Abnormalities

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Study Design. Retrospective clinical study.

Objective. To show retrospective analysis of 21 consecutive patients who underwent simultaneous surgical treatment for progressive spinal deformity and coexisting intraspinal pathologies (tethered cord and/or diastematomyelia).

Summary of Background Data. The classic advocated approach in patients with congenital spine deformity associated with intraspinal anomalies is first to perform surgery for the intraspinal pathologies and then surgery for correction and stabilization of the deformity 3 to 6 months later. To our knowledge, there is no study on simultaneous surgical treatment for these 2 associated conditions.

Methods. In the surgery; after the exposure of the determined levels, placement of all pedicle screws was performed as the initial part of surgical procedure. Then surgical treatment for intraspinal pathology was performed by the neurosurgical team and then followed by completion of instrumentation and correction of the deformity. Additional anterior surgery was done later to prevent pseudarthrosis and crankshaft phenomenon.

Results. The mean age of the patients at presentation ranged from 3 to 19 years (mean, 13 years). There were 17 female patients and 4 male patients. Four patients had neurologic deficits at the time of presentation, and all 4 had associated kyphosis. The mean operation time was 9.3 hours (range, 7–12 hours) and the mean blood loss was 1980 mL (range, 1500–3000 mL). The average follow-up was 6.8 years (2–12 years). None of the patients experienced deterioration in their neurologic status after surgery. None of the patients had infection, pseudarthrosis, or loss of correction during the follow-up visits.

Conclusion. The simultaneous surgical treatment for congenital deformity and intraspinal abnormality does not involve significant complications and seems to be an alternative and safe treatment option.

Key words: intraspinal abnormality, congenital scoliosis, congenital kyphosis, surgical treatment. **Spine 2007; 32:2880–2884**

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Congenital scoliosis and/or kyphosis occurs as a result of either a failure of formation or a failure of segmentation, or both.[1] This most frequently occurs in the first 8 weeks of prenatal development. During this time, the bony elements of the spine are forming, and the neuraxis is completing its infolding, closing the neural tube.[2] These events are closely related, and any intrauterine event that causes congenital scoliosis and/or kyphosis could also be associated with an intraspinal anomaly. These anomalies include tethering of the cord, diastematomyelia, lipoma and lipomeningocele, teratomas, and syringomyelia.

The intraspinal anomalies can cause progressive neural loss with growth and curve progression. In addition, they greatly increase the risk of neurologic injury during surgical correction of the deformity.[3–6] McMaster[7] reported intraspinal abnormality in 18% of 251 patients with myelography. Magnetic resonance imaging (MRI) is noninvasive and more sensitive in detecting the intraspinal abnormalities. Using MRI, Bradford *et al*[6] reported that 38% of the 42 patients in their study had an intraspinal anomaly.

Congenital scoliosis and/or kyphosis due to the presence of vertebral anomalies cause an imbalance in the longitudinal growth of the spine. Being a developmental anomaly, it is often associated with intraspinal, genitourinary, cardiovascular, and other general abnormalities.[7–9] Congenital deformities of the spine are relatively rigid with a concurrent danger of neurologic complications.[10] All these factors, including the presence of intraspinal abnormalities, are important in decision-making and the management of congenital scoliosis.

The classic advocated approach in such patients is first to perform surgery for the intraspinal pathologies and then surgery for correction and stabilization of the deformity 3 to 6 months later.[7,9] To our knowledge, there is no study on simultaneous surgical treatment for these 2 associated conditions.

In the current study, we have retrospectively analyzed 21 consecutive patients who underwent simultaneous surgical treatment for progressive spinal deformity and coexisting intraspinal pathologies (tethered cord and/or diastematomyelia).

Materials and Methods

We retrospectively reviewed the records of 21 consecutive patients with congenital scoliosis and/or kyphosis associated with intraspinal abnormalities treated by simultaneous surgery between the years 1994 and 2004.

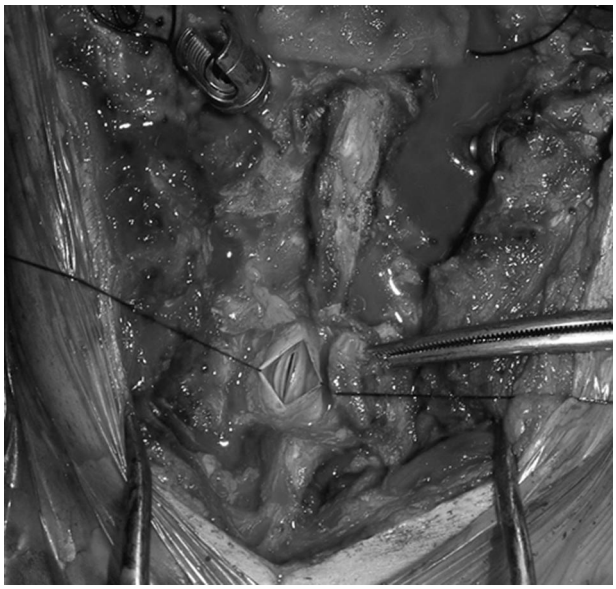

Figure 1. The surgical treatment for intraspinal pathology (release of tethered cord and/or excision of diastematomyelic bone spur) was performed by the neurosurgical team.

The age at presentation, sex, presence of associated kyphosis, and the neurologic status were documented. All the patients had standing anteroposterior and lateral views of the spine from C5 to sacrum. All patients were subjected to neural axis MRI from brainstem to sacrum to detect the associated intraspinal anomalies. Ultrasonography was used as a screening procedure to detect genitourinary anomalies. A cardiovascular evaluation was done of all the patients by both clinical examination and echocardiography to exclude cardiac anomalies. All patients also underwent assessment by a pediatrician.

In the surgery, after the exposure of the determined levels, placement of all pedicle screws was performed as the initial part of surgical procedure. Then surgical treatment for intraspinal pathology (release of tethered cord and/or excision of diastematomyelic bone spur) was performed by the neurosurgical team, then followed completion of instrumentation and correction of the deformity (posterior shortening wedge osteotomy or subtotal vertebrectomy if needed) (Figure 1). Posterior instrumentation for correction of the deformity was short segment in 7 patients and long segment in the remaining 14 patients. Also, vertebrectomy *via* posterior approach was performed in 5 patients and 3 patients had additional anterior surgery later to prevent pseudarthrosis and crankshaft phenomenon. A combination of autograft and allograft was used for achieving fusion in all patients (Figures 2, 3).

The operation time, blood loss, postoperative complications, and the hospitalization time were noted and recorded.

■ Results

The mean age of the patients at presentation ranged from 3 to 19 years (mean, 13 years). There were 17 female patients and 4 male patients.

The congenital vertebral anomalies were classified using standard terminology as either failure of formation, failure of segmentation, or mixed defects.[11] There were 16 cases of congenital scoliosis (including kyphoscoliosis) and 5 cases of congenital kyphosis due to hemivertebra. Of the 16 cases of congenital scoliosis, there were 7 classified as failure of formation, 3 as failure of segmentation, and 6 as mixed defects. The MRI showed the intraspinal pathologies as 7 patients with tethered cord, 1 patient with retethering, and 13 patients with diastematomyelia with tethered cord. Four patients had neurologic deficits at the time of presentation, and all 4 had associated kyphosis. Bilateral weakness of toe extensors with absent ankle jerk was seen in 2 patients. In the remaining 2, there was hypoesthesia at L5, S1 dermatome and weakness of toe extensors as well as flexors.

Genitourinary anomalies were seen in 5 patients (unilateral agenesis of the kidney in 3 patients and ectopic kidney in 2 patients). None of them had clinical features suggestive of genitourinary anomalies but

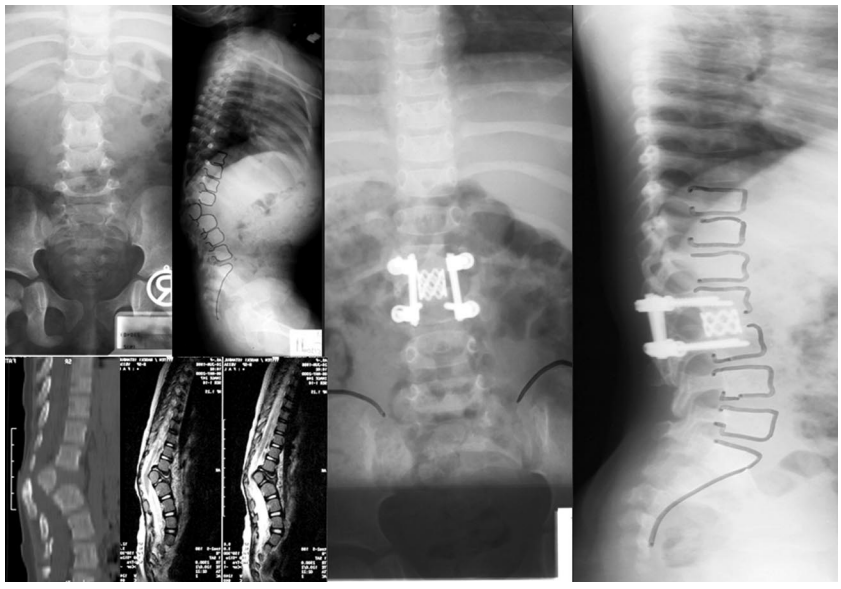

Figure 2. A 3-year-old female patient with congenital kyphosis associated with tethered cord. The patient had neurologic deficit at presentation. The following surgical procedures were performed in order: placement of pedicle screws, release of tethered cord, posterior total hemivertebrectomy, correction and fusion with titanium mesh cage. At postoperative 2 years, she is free from any neurologic symptoms.

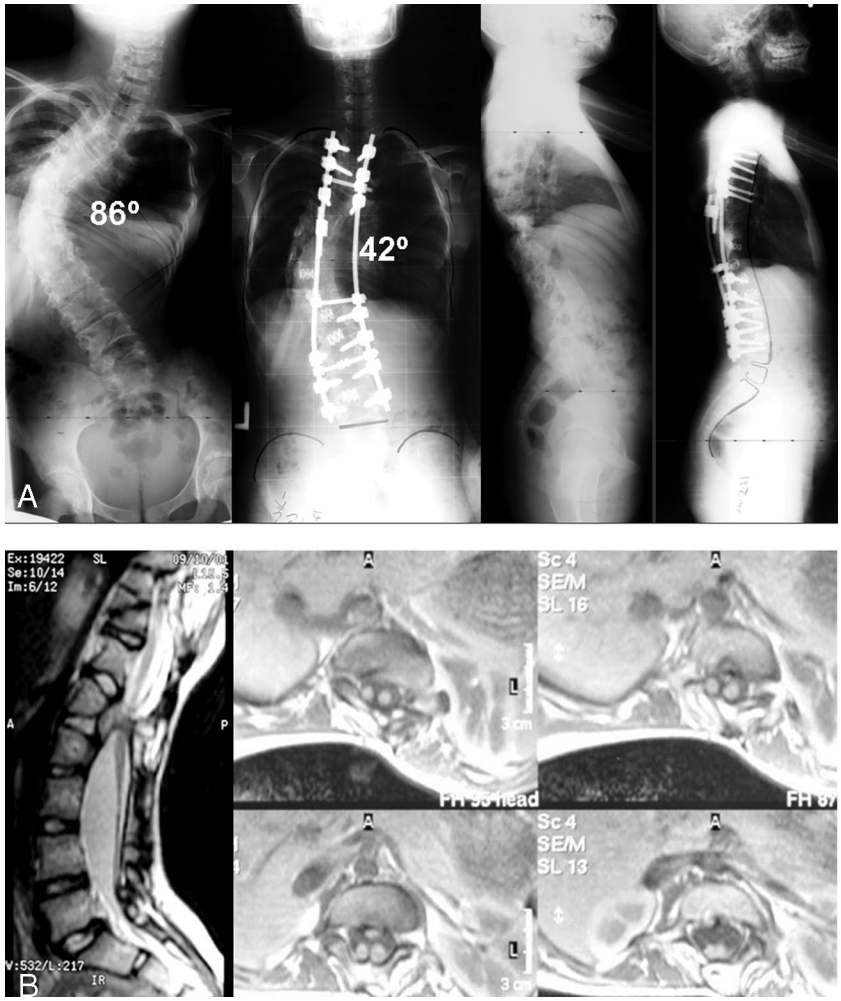

Figure 3. **A**, A 15-year-old female patient with congenital scoliosis associated with diastematomyelia and tethered cord. After exposure and posterior placement of pedicle screws, tethered cord was released and bony spur was excised. Then, we performed posterior shortening wedge osteotomy at T10–T11 to correct severe frontal imbalance. Two weeks later, anterior fusion was done to prevent pseudarthrosis. At fifth postoperative year, she is free of any symptoms. **B**, MRI scans showing the diastematomyelia.

were incidental findings detected on screening. Cardiovascular anomalies were seen in 6 patients with mitral valve prolapse being the commonest anomaly (in 4 of 6 patients).

The mean operation time was 9.3 hours (range, 7–12 hours) and the mean blood loss was 1980 mL (range, 1500–3000 mL). All patients spent at least 1 day in intensive care unit and the mean hospitalization time (excluding 3 patients needing additional anterior surgery) was 7.3 days (range, 5–12 days).

The mean correction in the deformity (either kyphosis or scoliosis or both) was 23%, and the correction loss at the end of follow-up was less than 10%.

The average follow-up was 6.8 years (range, 2–12 years). None of the patients experienced deterioration in their neurologic status after surgery. For the patients with preoperative neurologic deficit, the recovery was complete in 2 patients, neurologic status was better in 1 patient and unchanged in the other. In 1 patient who had associated myelomeningocele, postoperative cerebrospinal fluid leakage was observed, and it was repaired successfully in a second surgical procedure. None of the patients had infection, pseudarthrosis, or loss of correction during the follow-up visits.

■ Discussion

In patients with congenital spinal deformity, the prevalence of intraspinal anomalies that are detected with magnetic resonance imaging has been reported to be 20% to 58%.[3,12,13] MRI is superior to either radiography or myelography for the identification and evaluation of early developmental conditions of the neural axis because it provides more information and is less invasive. In the current study, the most common intraspinal anomalies diagnosed with MRI were tethered spinal cord and diastematomyelia. These findings corroborate those of previous MRI studies of congenital scoliosis that have demonstrated that the most common intraspinal anomalies were tethered spinal cord (prevalence, 13%–29%) and diastematomyelia (prevalence, 3%–12%).[3,4,6,13,14]

The median age at presentation was 13 years. This may support the hypothesis that, although the vertebral anomalies are present at birth, they may manifest as a visible deformity only much later. However, socioeconomic factors may have also contributed to this delay in presentation.

Neurologic involvement at the time of presentation may indicate the underlying occult intraspinal

anomaly.[15] It is therefore essential to exclude underlying spinal cord anomalies if a patient presents with a neurologic deficit, which would then dictate the treatment options. Neurologic deficit at the time of presentation was seen in 4 patients. All of them had associated kyphosis and all were found to have diastematomyelia with tethered cord.

Congenital scoliosis and/or kyphosis is frequently associated with congenital anomalies in other systems, especially those formed from mesenchymal tissue. These anomalies are often asymptomatic and may remain undetected until the patient is fully assessed following the diagnosis of congenital spinal deformity.[7,16] Genitourinary anomalies that can cause obstruction are silent and, if not searched for, may cause serious damage before they are diagnosed. The incidence of genitourinary anomalies was found to be 23.8% in the current study. This is similar to frequencies seen in other studies.[16–18]

Cardiovascular anomalies were the most common of the associated systemic anomalies in our study. They were seen in 28.5% of patients with mitral valve prolapse being the commonest anomaly. According to Winter, the incidence of congenital heart disease in congenital scoliosis was 7%.[8] The study done by Roth *et al*[19] showed that the overall incidence of scoliosis inpatients with congenital heart disease was 12%. The cause of our high rate may be the small size of the patient group.

As decision-making in congenital scoliosis and/or kyphosis depends on the natural history of progression, clinical and radiologic assessment is essential. Detection of inherent problems in the spinal cord and other associated anomalies become relevant in choosing the appropriate surgical management.

According to Winter *et al*,[9] the classic advocated approach in patients having congenital spine deformity associated with intraspinal abnormality is first to perform surgery for the intraspinal pathologies and then surgery for correction and stabilization of the deformity 3 to 6 months later in a different session. However, this different session surgical treatment carries some disadvantages. First, it contains more than 1 surgery and expose the patients to the risks of surgery more than once. Difficult surgical exposure, increased bleeding, adhesion formation, and less clear anatomic landmarks make the later corrective surgery more difficult. Also, after primary neurosurgical procedure, there is a risk of retethering and it often necessitates another neurosurgical procedure before corrective surgery. In addition, more complicated reconstructive procedures like osteotomy, hemivertebrectomy, *etc.*, can be done with difficulty due to preformed adhesion in the surgical site. There is also increased risk of iatrogenic neural element injury during such approaches.

From the year 1994, our surgical approach for the treatment of these patients is the simultaneous surgery for the intraspinal pathology and corrective surgery for the congenital spinal deformity *via* posterior approach. If needed, we perform anterior surgery 2 to 3 weeks later to prevent pseudarthrosis and crankshaft phenomenon. According to our knowledge, there is no study in the English literature involving simultaneous corrective surgical treatment for congenital spinal deformity and neurosurgical procedure for the surgical treatment of the coexisting intraspinal abnormality. It does not increase the surgical time, blood loss, and hospitalization time too much. Although our patient number is not too big, simultaneous surgical treatment for congenital deformity and intraspinal abnormality does not involve significant complications and seems to be an attractive and safe treatment option.

Our decision-making procedure in choosing of short- or long-segment fusion is that in patients having severe, sharp angular kyphosis or kyphosis and/or scoliosis due to a single hemivertebra; the posterior instrumentation and short-segment fusion was done together with hemivertebra resection or with corrective osteotomies. On the other hand, in patients with severe thoracolumbar or lumbar scoliosis even associated with pelvic tilt, posterior instrumentation and long segment fusion was preferred. In patients with severe sagittal and coronal imbalance, transpedicular osteotomy and/or vertebral column resection together with long-segment instrumentation was done. This was especially considered in patients with severely disturbed coronal balance, severe kyphoscoliosis, and presence of trunk shift due to the deformity.

Decision whether to do fusion in situ or to do correction has arisen during the operation according to the last form of the deformity after neurosurgical approach finished and/or corrective osteotomies were done. The flexibility of the deformity is the major concern in those situations.

This study reports on a retrospective study of a small series of patients with congential scoliosis and/or kyphosis associated with intraspinal abnormalities treated surgically for the intraspinal abnormality and the spinal deformity at the same sitting. There were no significant complications, showing that the 1-stage procedure can give comparable results to 2-stage procedures adopted by other workers in the past. This situation is valid only in the presence of the following:

- surgical team experienced in spinal deformity surgery
- well preoperative surgical planning
- neurosurgical team experience in intramedullary abnormalities field
- use of intraoperative neurophysiologic studies (neuromonitorization). We are routinely using neuromonitorization for last 2 years in our all deformity cases
- anesthesiology department experienced in caring both intraoperative and early postoperative phases of these patients
- well working pediatric intensive care unit and also, clear explanation of advantages and disadvantages of

the same sitting surgery to the patients' parents is mandatory before all such kind of procedures.

■ Key Points

- The classic advocated approach in patients with congenital spine deformity associated with intraspinal abnormalities is first to perform surgery for the intraspinal pathologies and then surgery for correction and stabilization of the deformity 3 to 6 months later.
- There is no study in the literature related to simultaneous surgical treatment for these 2 associated conditions.
- The separate session surgical treatment carries some disadvantages related to the patient's general medical condition and surgical site.
- The simultaneous surgical treatment for congenital deformity and intraspinal abnormality does not involve significant complications and seems to be an alternative and safe treatment option.

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