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Hemivertebra Excision for Congenital Scoliosis

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Hemivertebrae result from a failure of complete formation of the vertebral body and present as a challenging clinical entity. Progression and ultimate curve magnitude is associated with the degree of segmentation. An accurate assessment and a close follow-up are necessary to make appropriate treatment decisions. The goal of any intervention is to stop progression and provide the smallest degree of deformity at skeletal maturity. If a high degree of deformity is anticipated, intervention should be attempted sooner rather than later. In these cases, an in situ fusion may be appropriate. Single-stage hemivertebra excision is an effective method of both correction and stabilization; however, the procedure is technically demanding.
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Hemivertebrae result from a failure of complete formation of the vertebral body and are one of the most common causes of congenital scoliosis. Various degrees of incomplete formation occur, which range from subtle to severe, and these differences correlate with relative deformity progression. In addition to the altered bone morphology, the adjacent disks may also be deficient. Most hemivertebrae possess a normal disk above and below the malformed bone, which is termed a fully segmented hemivertebra. When no disk exists either above or below, it is termed incarcerated or nonsegmented; this scenario is the least common.[1] Any part of the spine may be affected, and hemivertebrae may occur at 1 or more locations. The degree of altered formation, location, number, age of the patient, and segmentation dictate the behavior of the spine as the patient grows.

Scoliosis associated with fully segmented hemivertebrae progresses at a rate of 1-2 degrees per year.[1,2] When the defect occurs at the lumbosacral or thoracolumbar region, a severe trunk shift may occur. A secondary compensatory curve may develop above and become structural over time. Kyphosis is also a concern, which may result in neurologic complications in patients with thoracic hemivertebrae. Semisegmented hemivertebrae cause a slowly progressive scoliotic curve that usually does not exceed 40 degrees at maturity. A nonsegmented hemivertebra is synostosed above and below its adjacent vertebrae, and, typically, a progressive deformity does not occur because of the lack of growth potential. Defects in the thoracic spine tend to progress at a higher rate than those of the lumbar spine.[3]

Assessment

An accurate assessment to determine whether the curve is progressing is paramount in choosing the appropriate intervention. Careful clinical and radiographic assessments should be made every 3 or 6 months. It is often difficult to obtain accurate radiographic measurements in patients with hemivertebrae; care should be taken when comparing radiographs. Serial follow up imaging allows an estimation of the degree of deformity at skeletal maturity. A common error in treatment of congenital scoliosis is the failure to measure radiographs accurately. Skeletal immaturity and aberrations in the vertebra contribute to difficulties in selecting measurement points. To confirm that progression has occurred, measurements have to overcome interobserver variability. Inter- and intraobserver reliability studies have been performed to assess the accuracy of measurements in congenital scoliosis. In 1 study, interobserver variability was ±11.8 degrees, requiring that there should be at least a 23-degree increase to determine curve progression, whereas another article showed interobserver variances to be ±7.86 degrees, which corresponds to a 15.7-degree increase to document progression.[4-5] When documented progression occurs, it is important to act on the newfound data.

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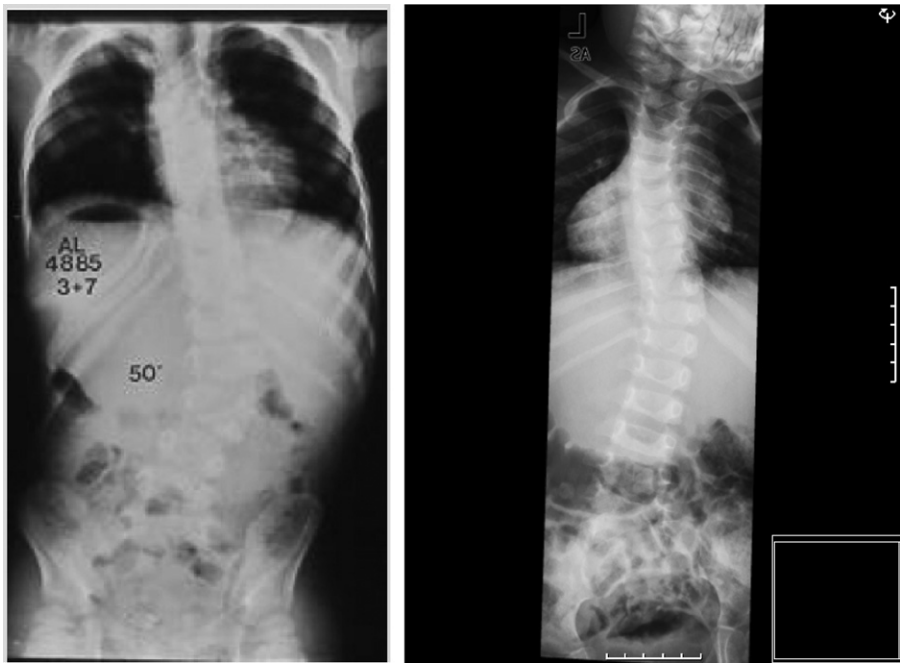

Figure 1 Illustrative case.

In patients with a question of intraspinal pathology, magnetic resonance imaging, computed tomography, or myelography may be warranted.

Treatment

The overall goal of treatment is to prevent the development of a severe deformity. Bracing is far less effective in patients with hemivertebrae than in patients with idiopathic scoliosis.[6] In cases where the degree of curvature at skeletal maturity is projected to be severe, surgery is the mainstay of care. Types of surgical intervention include posterior arthrodesis, convex-sided growth arrest, and hemivertebra excision.

Posterior Arthrodesis/Growth Arrest

Isolated posterior procedures have been used in the past in the treatment of hemivertebra. Posterior fusion, the most common surgical technique in the treatment of congenital scoliosis, may not alone provide the desired balance correction in patients with hemivertebrae. By only creating a fusion mass posteriorly, there is always the risk of bending of the fusion mass over time in young patients because of the crankshaft effect. Loss of correction may occur, warranting further intervention.

Convex growth arrest is appropriate in patients with remaining growth potential; however, this technique must be performed early.[7-9] The patient is generally <5 years of age with curves <50 degrees. An anterior/posterior fusion is performed on only the convex side of the deformity, and the patient is placed in a corrective cast. Close monitoring of the patient is critical because the procedure is done before the

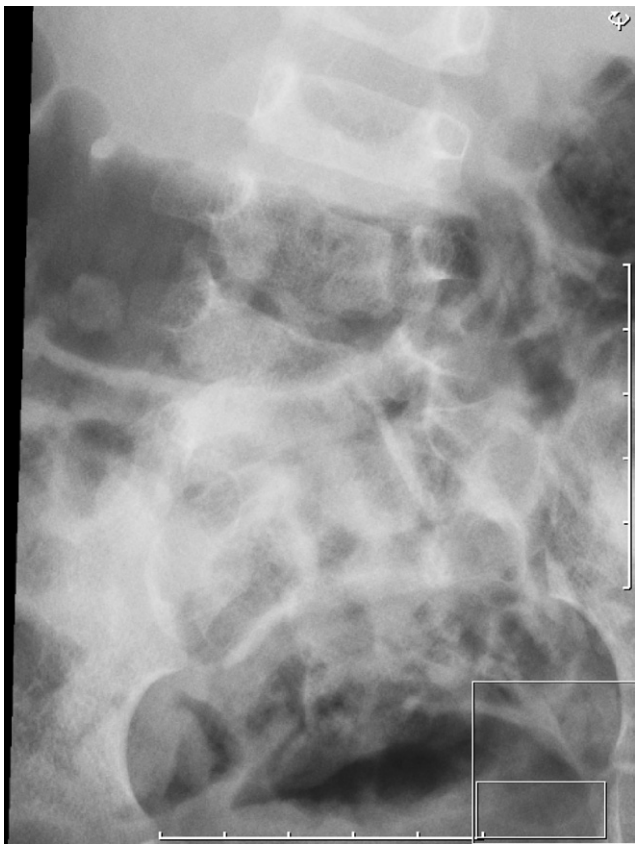

Figure 2 A 3-year-old patient with low lumbar hemivertebra treated with single-stage excision and fixation.