CONGENITAL SCOLIOSIS DUE TO HEMIVERTEBRA
TREATMENT OPTIONS

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Congenital scoliosis due to hemivertebra: Treatment options

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Abstract

Study design. A review of English literature about treatment of congenital scoliosis due to hemivertebra.

Objective. Make a review of the treatment of hemivertebra describing the types of surgical procedures available, as well as, the indications and contraindications of each procedure and results obtained.

Summary of background data. Congenital scoliosis due to hemivertebra is most often progressive and requires surgical treatment. The recommended surgical options includes: “in situ” fusion; convex hemiepiphysiodesis and hemivertebra excision.

Methods. Comprehensive review of articles published on “congenital scoliosis - treatment”. Medline, pubmed, b-on, science citation index yielded about 1000 titles each of which was surveyed for content related to treatment of hemivertebra.

Results. “In situ” fusion does not address the anomaly directly, allows a moderate correction rate but is associated with long spinal fusion which has repercussion in spinal mobility and pulmonary function. Convex hemiepiphysiodesis is a good option for young patients, less than 5 years old, with single fully segmented hemivertebra and with Cobb angle < 60º. However, the degree of correction is unpredictable and dependent on the concave growth. Finally, hemivertebra resection addresses directly the spinal anomaly, produces the best correction results and it is a safe procedure that can be performed during all childhood and even in adults. The evolution of the posterior-only approach allowed it to be applied in very young children and adults with less perioperative neurological risk and with promising results.

Conclusions. The treatment of congenital scoliosis focuses on early diagnosis and appropriate surgical management before the development of large curves. All the procedures described can be effective and safely performed if correctly selected and preformed by an experienced team of surgeons. The main goal of surgery is to achieve a straight spine and a physiological sagittal profile while preserving as much as possible, normal spinal growth.

[Key words: Congenital scoliosis; Hemivertebra; Treatment]

Introduction:

Congenital scoliosis is defined as lateral curvature of the spine due to a developmental abnormality. An incidence of approximately of 1 /1000 births has been observed for congenital scoliosis. In most cases congenital scoliosis are non-hereditary. Congenital scoliosis is believed to be related to an insult to the fetus during spine embryology development (between the 5th and 8th week of gestation). This is why other malformations such as congenital heart disease, spinal cord dysraphism, or kidney malformations are frequently associated. Only cases with syndromic associations (Jarcho-Levin, spondylocostal dysplasia) or multiple defects of segmentation can have hereditary factor. Vertebral anomalies causing congenital scoliosis may be caused by failure of formation (wedge vertebra and hemivertebra), by failure of segmentation (“en bloc” vertebra and unilateral bars) or by a combination of these 2 factors, resulting in a mixed deformity. Hemivertebra is the most common cause of congenital scoliosis and may be further classified according to the fusion of the vertebral bodies above and/or below in fully segmented, partially segmented or unsegmented. The natural history is highly unpredictable and the degree of scoliosis produced by hemivertebra depends on the type of anomaly, site and number of hemivertebra and the patient’s age. Curve progression occurs more rapidly when hemivertebra is fully segmented especially when associated with an unilateral bar, during the first 5 years of life and during the adolescent growth spurt and when the vertebral anomaly is located at cervicothoracic and lumbosacral junctions. Statistically 25% of curves are non-progressive, 25% mildly progressive and 50% highly progressive and will require treatment.
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Treatment of congenital scoliosis will consist in either observation of the curve or surgery or, rarely bracing. Observation should be applied only for non-progressive curves with balanced spine. Bracing in most instances is totally inefficient in congenital scoliosis. It may be indicated for long flexible curves or to control the compensatory lumbar curve or help rebalancing the spine, or it may be used after an operation, for instance, until the fusion is solid. In cases of anticipated curve progression (hemivertebra associated with unilateral bar), severe trunk imbalance, or large curve, surgical intervention is indicated. The goal of surgery is to achieve a straight spine, a physiological sagittal profile while maintaining flexibility, to arrest progression of the curve and as short a segment fusion as possible, preserving as much normal spinal growth as possible. Surgery for congenital scoliosis carries a greater neurological injury risk than in idiopathic spinal deformities.

Early treatment of progressive deformities helps in minimizing the risks of surgery and allows better correction and prevents the development of structural and compensatory curves. Preoperative evaluations should include: a comprehensive history and physical examination paying particular attention to the prenatal history, neurological examination, signs of spinal dysraphism, spinal balance, rib cage deformities, cardiac and renal abnormalities. It is also essential to evaluate spinal flexibility and skeletal maturity. Plain radiographs remain standard for diagnosis of congenital anomalies and measuring curve magnitude, progression and growth potential of the vertebral anomaly. CT scans (2D and 3D) may help define the anatomy, avoid any unexpected intraoperative posterior element deficiencies and help planning the surgical approach. As congenital scoliosis is frequently associated with spinal dysraphism, MRI assumed an important role in preoperative evaluation. Surgical procedures may be broadly divided according to their goal in: those preventing further deformity and those that correct the present deformity.

Prevention of further deformity:

In situ fusion – can be done with a single posterior fusion with or without instrumentation or with an anterior fusion or anterior and posterior fusion. The aim is to achieve halt in the progression of the deformity. However, the compensatory curve above the fusion area may still progress on its own after such operation. Some correction of the so-called fusion may be achieved if one uses a corrective cast postoperatively. In young children if the fusion is only posterior there is risk of crankshaft phenomenon to occur.

Convex hemiepiphysiodesis – is done by removing the convex lateral half of disks and growth plates adjacent to the hemivertebra, with no exposure of the spine on the concave side. It is indicated for a unilateral formation failure i.e. hemivertebra. Because there must be adequate growth potential on the concave side, this procedure is contraindicated in segmentation defects, such as bars.

Hemivertebra excision – a hemivertebra causing marked truncal imbalance and progression of the curve can be managed with wedge resection in patients whom an isolated “in situ” fusion or convex hemiepiphysiodesis would not result in balanced spine. It is best performed at approximately 2 years of age when the child can still tolerate a cast and fusion is more likely. It offers nearly complete correction over a short fusion segment and may be performed via anterior-posterior surgery or posterior only surgery. Initially it was only performed for thoraco-lumbar or lumbar curves because of the neurological risk associated to the surgery at thoracic level, but recently thoracic hemivertebra resection has been performed in safety and with good results.

Correction of the present deformity

Correction and Fusion with instrumentation – Posterior spine fusion without instrumentation and correction with a cast can be indicated in young children, but the lack of anterior fusion exposes the spine to the crankshaft phenomenon if the anterior growth plates overcome the posterior fusion. Posterior spine fusion with instrumentation is indicated in older teenagers, where no risk of crankshafting exist but there is a risk of neurological complications if too much distraction is applied. Anterior and posterior spine
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Fusion with discectomies and instrumentation can achieve a significant correction in the mobile segments of the spine. However, the danger is over-correction with distraction of the curve and neurologic complications. Distraction should never be done first. The use of spinal cord monitoring and immediate wake-up test after correction is here, more than anywhere else, mandatory.

Hemiepiphyseodesis and Hemiarthrodesis – these techniques rely on the future growth of the spine on the concave side, as so, they can only be performed for failure of formation anomalies. The goal is to arrest the growth on the convex side of the curve; the concave side is not exposed surgically, as doing so, could lead to spontaneous fusion. Much of the correction is achieved acutely at the time of the initial procedure, thus postoperative a corrective cast is used to encourage fusion of the spine in a somewhat corrected position. The total correction will be dependent on the child’s age, and the younger the child is, the more potential correction exist. According to Winter et al. this procedure should be reserved for patients younger than 5 years of age, with a progressive curve of < 70º involving 5 segments or less, and presenting with a pure scoliosis, not involving the cervical spine and with no major kyphosis or lordosis. However the results of this procedure are somewhat unpredictable.

Hemivertebra resection – hemivertebra resection is done either through a posterior approach only or through a sequential or simultaneous front and back approach. These procedures usually provide an average of 25º-30º of correction, with some correction of the associated kyphosis. The correction is usually obtained with a pedicle screw rod system. The best indications of hemivertebra resection are the lumbosacral hemivertebra or the hemivertebra situated below the spinal cord and responsible for the take-off with pelvic obliquity. In the thoracic spine, these resections are definitely more dangerous and should only be performed by experienced spine surgeons. Recent publications tend to show that hemivertebra resection is safe even in the thoracic spine. Hemivertebreal excision has a potential advantage over alternative techniques for the surgical management of congenital scoliosis by addressing the deformity directly and allowing immediate, better controlled and more predictable correction, particularly for coronally decompensated patients.

Single and Dual Growing Rods: The growth of the spine is greatest during the first 5 years of life while thoracic volume reaches 30% of the adult size by five years of age. Long fusions performed on younger children with scoliosis may have a further deleterious effect on the trunk height and the thoracic volume, leading to thoracic insufficiency. If the child is still very young, the primary congenital curve can be treated with “in situ” fusion, hemiepiphyseodesis and/or hemiarthrodesis, excision, or osteotomy, and the corrected curve can then be treated with a growing rod until the child is older. This avoids fusing the entire curve, which will lead to growth retardation and potentially harmful pulmonary effects. This technique was pioneered by Paul Harrington with single growing rod in the 1960’s and evoluted by Akbarnia and McCarthy to dual growing rod. However, they expose the patient to multiple lengthening operations and carry a significant risk of complications, mostly infections or instrumentation complications.

Expansion thoracoplasty and VEPTR – congenital spine deformities with rib fusions may be associated with thoracic insufficiency. The surgical concept of the expansion thoracoplasty and stabilization with the VEPTR implant (vertical expander prosthetic titanium rib) is based on the expansion of the thorax by rib distraction on the concave side of the curve achieving indirect correction of the curve. The best indications are in cases of congenital scoliosis associated with fused ribs and/or patients with thoracic insufficiency syndrome and/or chest hypoplasia before the age of 8.

The aim of this study was to make a review of the published literature about the treatment of congenital scoliosis due to hemivertebra describing the types of surgical procedures available, as well as, the indications and contraindications of each procedure and results obtained.
Materials and methods:
Comprehensive review of articles (English language only) published on “congenital scoliosis - treatment”, whose content yielded data on the treatment of hemivertebra. Medline, pubmed, b-on, science citation index and other searches yielded about 1000 titles each of which was surveyed for content related to treatment of congenital scoliosis due to hemivertebra.

Results:
The three main surgical procedures recommended are: “in situ” fusion, convex-side growth arrest (hemi-epiphysiodesis) and hemivertebra resection.

Posterior spinal fusion has considerable limitations. The goal of posterior surgery is stabilization in order to prevent further progression rather than correction of the curve. Winter et al\(^3\) reported 290 patients with congenital scoliosis who had posterior fusion with or without Harrington instrumentation. Correction was limited to 28% in those fused without instrumentation and to 36% in those whom Harrington implants were used. Instrumented distraction across the concavity was associated with the risk of paraplegia. Deformation of the fusion mass because of continued anterior growth was observed in 40 patients (14%) – Crankshaft phenomenon. Hall et al\(^18\),\(^19\) reported a mean correction of the curve of 12% in posterior fusion without instrumentation improving to 35% with Harrington instrumentation. Slabaugh et al\(^2\) compared hemivertebra excision with posterior “in situ” fusion for lumbosacral hemivertebra and found better correction of the curve in the group who had excision. Combined anterior and posterior fusion adds the potential benefit of greater correction and of sagittal plane correction because the excision of discs allows greater mobility of the segments. It also decreases the likelihood of pseudoarthrosis and prevents the crankshaft phenomenon by removing the growth plates anteriorly. Since this technique does not address the wedge deformity directly the entire measured curve must be encompassed in the fusion, including normal segments.

Convex epiphysiodesis of the spine was designed to arrest growth while allowing concave growth to correct the deformity. It is necessary to perform convex hemiepiphysiodesis across the entire measured curve, often including a normal segment above and below, in order to achieve a satisfactory improvement. The results of this procedure have been variable and unpredictable. Roaf\(^18\) described convex hemiepiphysiodesis and proposed that further growth on the concave side would correct the deformity. He achieved correction of more than 20º in 23% of patients, but less than 10º in 40%. Winter and Moe\(^7\) reported early results in 13 children treated by convex hemiepiphysiodesis showing curve progression arrest in seven patients (54%) and improvement of more than 5º in five (38%). An overview of the english-language articles published till the year of 2006\(^20\) revealed that 123 patients have been treated with convex-side epiphysiodesis. The results obtained were the improvement of curve in 48% of patients (range 20% to 77%), stopped progression only in 40% (range 17% to 70%), and continued curve progression in 12% (range 0% to 21%). Ginsburg et al\(^21\) reported the technique of transpedicular hemiepiphysiodesis for treatment of hemivertebra. They concluded that this procedure is a safe and effective treatment method to halt the progression of congenital scoliosis due to hemivertebra in patients who are skeletally immature.

Excision of a hemivertebra was first reported in 1928 by Royle\(^22\) in Australia. Bradford and Boachie-Adjei\(^23\) reported a mean correction of the scoliosis of 68.1% (from 47º to 15º). Lazar and Hall\(^14\) also used a single compression rod in 11 patients through the combined approach. The mean correction of the scoliosis was 70.2% (from 74º to 14º). Leatherman and Dickson\(^25\) and Slabaugh et al\(^2\) performed dual Harrington rod instrumentation through the combined approach. The correction average was 44.2% (from 77º to 43º) and 35.9% (from 39º to 25º), respectively. Callahan et al\(^17\) reported a mean correction of 60% (from 40º to 16º) in 9 patients after placement of spinous process wires. Klemme et al\(^26\) reported a mean correction of 71.1% (from 38º to 11º) in 6 patients in whom large sublaminar suture tapes were used after hemivertebra
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resection through a combined approach. Deriven et al.\textsuperscript{27} reported a 59\% correction with thoracic and thoracolumbar hemivertebra excision in 10 patients, and concluded that these procedures are safe for thoracic curves in experienced hands. No neurological injuries were seen in these series. More recent studies have reported on hemivertebra excision from the posterior approach only. Ruf and Harms\textsuperscript{28} reported on 28 cases of posterior-only hemivertebra resection with a mean correction of 71.1\% (from 45\degree to 13\degree). Complications included 2 pedicle fractures, 3 failures of instrumentation, 2 additional operations for curve progression and 1 infection. Posterior instrumentation allows an early intervention in very young children. Excellent correction in the frontal and sagital planes and short segment fusion allows for normal growth in the unaffected parts of the spine. Shono et al.\textsuperscript{29} and Nakamura et al.\textsuperscript{30} reported similar results with the same technique.

The ideal age for hemivertebrectomy is controversial. Potential advantages of early surgery are several\textsuperscript{26}: first, and perhaps most importantly, surgery for lower magnitude curves requires less correction and less surgical aggression and carries less neurological risk; second, despite a congenital origin, immature congenital curves may retain some local flexibility and often lack structural compensatory curves. Such curves, therefore, may allow shorter arthrodesis. Finally, more than one study states that hemivertebra excision has superior results in young children.

Klemme et al.\textsuperscript{26} and Callagahn\textsuperscript{17} concluded that it is safely preformed in very young children. Ruf and Harms\textsuperscript{31} in their study of posterior hemivertebra resection with transpedicular instrumentation recommended that correction should be preformed early before the development of severe local deformities and secondary structural changes, especially in patients with expected deterioration. However, surgery in very young children is associated with several potential problems, namely, neurological injuries with pedicle screws, suitable pediatric instruments, stable fixation of vertebral segments, difficulty in wound closure, wound healing problems and infections and anesthetic risks.\textsuperscript{32} These problems were partially overcome with the evolution and more experience of the technique and anesthetic procedures, as well as, the evolution of new instruments namely the new low-profile implants.

To date there has been very little written on congenital spinal deformity presenting in adulthood. The literature published till date,\textsuperscript{14, 33, 34} about surgical treatment of congenital scoliosis in adulthood reports that curve correction is limited but effective and safe and with promising results in pain relief. Vitale et al.\textsuperscript{35} in a retrospective study of pulmonary function and quality of life in children with congenital scoliosis revealed that these children treated by spinal fusion have significantly worse quality of life scores and pulmonary function. Comparing patient who had thoracic fusion vs nonthoracic fusion they realize those who had thoracic fusion had shorter spinal height, lower pulmonary function and reported more pain at follow-up visit after an average of 7 years of follow-up. The results of this study will add to a growing literature, which supports alternatives to early spinal fusion such as growing rods, epiphysiodesis and thoracoplasty.

Conclusion:

The treatment of congenital scoliosis focuses on early diagnosis and appropriate surgical management before the development of large curves. Vertebral anomalies that have a natural history of progression need to be managed aggressively. Knowing that surgery in congenital scoliosis is more risky than in idiopathic scoliosis and it is frequently associated with other systemic malformations or anomalies, a thorough preoperative assessment is mandatory and includes a standard history and physical examination, as well as, obtaining appropriate imaging tests. The best choice for surgical procedure depends on the anomaly itself, the degree of deformity and experience of the surgical team. Three main procedures are recommended: posterior or combined “in situ” fusion; convex hemiepiphysiodesis with or without instrumentation; and hemivertebra excision through combined approach or posterior-only approach. “In situ” fusion does not address the anomaly directly, allows a moderate correction rate but is associated with long spinal fusion which has repercussion in spinal
Congenital scoliosis due to hemivertebra mobility and pulmonary function. Convex hemiepiphysiodesis is a good option for young patients, less than 5 years old, with single fully segmented hemivertebra and with Cobb angle <60º. However, the degree of correction is unpredictable and dependent on the concave growth. Finally, hemivertebra resection is a technique that addresses directly the spinal anomaly, produces the best correction results and after overcoming the initial problems of neurological injury and instrumentation it is a safe procedure that can be performed during all childhood and even in adults. The evolution of the posterior-only approach allowed it to be applied in very young children and adults with less perioperative neurological risk and with promising results.

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