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BMH Med. J. 2021;8(1):30-36. **Case Report**

Hemivertebrae Excision in a One and a Half Year Old

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Abstract

Excision of hemivertebra at an early age offers acute correction of the deformity and prevention of secondary curve development. This limits the amount of fusion required otherwise in the long run and saves fusion levels and permit adequate lung development. Here is a case of fully segmented hemivertebra in a one and a half year old child with eleven years of follow up. The literature reports correction of congenital scoliosis by hemivertebra excision after 2 years of age.

Keywords: Hemivertebra, congenital scoliosis, Early onset scoliosis, Excision, Secondary curves, spinal arthrodesis

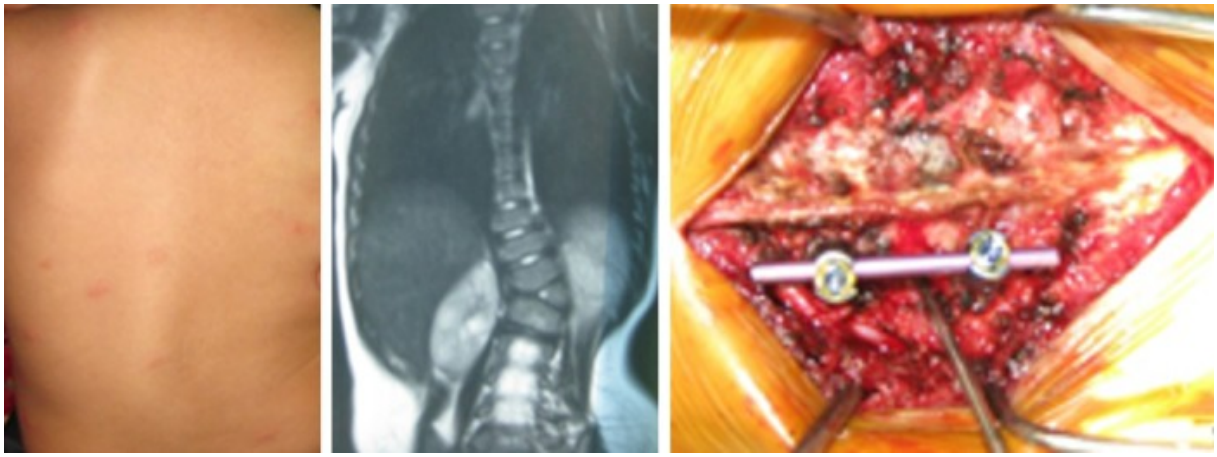
Introduction

Early onset scoliosis is defined by Scoliosis Research Society (SRS) as scoliosis of age of onset less than 10 years irrespective of the etiology [1]. The greatest potential for progression of spinal deformity is during periods of rapid growth. The rapid growth happens in the early years of life and during adolescent growth spurt. The lung develops in the first few years of life, when the number of alveoli and the lung volume increases rapidly. Rapid progression of thoracic curves in this age group can result in grotesque deformity, making severe changes in the thoracic cage, size and shape leading to restrictive lung disease with small lung volumes, decreased chest wall compliance and respiratory muscle dysfunction.

Case Report

A one and a half year old male child was presented by his parents for an apparent deformity of his back. The child was born of full term normal delivery for a non consanguineous parents. As his deformity was progressing he was evaluated with an X-ray and MRI scan of his spine, which showed a fully segmented hemivertebra at D12 on the left side. He was further assessed for any neurological deficit, cardiac abnormalities, urogenital abnormalities, respiratory and gastrointestinal tract abnormalities with ultrasound examination and consultation with concerned specialists.

In view of the progression of the deformity, he was planned for a hemivertebra excision through the posterior approach and fusion of the segment. Under general anesthesia the deformed area was exposed, the abnormal hemilamina and the vertebral body and pedicle was removed from the left side. Pedicle screws were put unilaterally on the left side and connected with a rod. Fusion was achieved with rib graft. The child was further immobilized in a bilateral hip spica cast for two months and the child was further followed up for 11 years and at the final followup the boy is having a balanced straight spine without any secondary curves being developed.



Figures 1: One and half year old boy with fully segmented hemivertebra at D12, Before surgery, Intra operative picture.

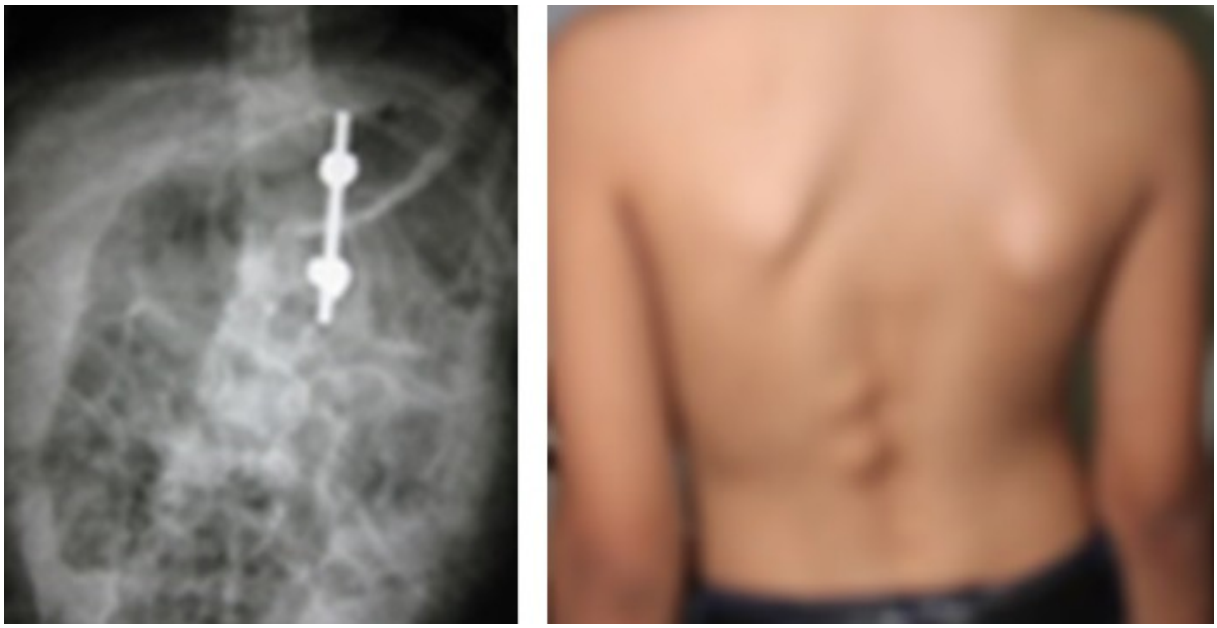


Figure 2: Hemivertebra excision, unilateral fixation, follow up for 11 years with well balanced spine, without secondary curves

Discussion

Based on etiology Early Onset Scoliosis (EOS) may be classified as:

1. Idiopathic scoliosis - exact etiology unknown
2. Congenital - Anomalies of vertebrae (Winter classification)
   - Failure of formation
   - Failure of segmentation
   - Mixed
3. Neuromuscular
   - Neuropathic
   - Myopathic
4. Syndromic
   - Connective tissue disorder
   - Metabolic causes - Rickets

The natural history of EOS is decided by the etiology & age of onset of the deformity. Congenital scoliosis occurs 1:1000 live births and is associated with defect in other organs in 61% of patients. Isolated hemivertebra is the most common form of spinal deformity. It is more common in the thoracic spine (~63%) followed by lumbar spine (~30%), sacral spine (~2.5%) and multiple locations (~3.7%). The reason for this is put forward as interplay of genetic and environmental factors [2]. According to Wynne-Davies [3], isolated hemivertebrae is a sporadic lesion that carried no risk of a similar lesion in siblings or subsequent generations, but multiple level complex anomalies carried a 5-10% risk of such an occurrence. Winter has opined that isolated vertebral anomalies can be expected to occur in first-degree relatives at the rate of 1:100 [4]. Vascular insult, environmental factors like carbon monoxide, hypoxia, alcohol, hypothermia etc. can cause vertebral anomalies. Congenital scoliosis results from an insult to vertebral growth between fifth and eighth week of gestation. The same insult can occur to other organs of the body. Cardiovascular anomalies are seen in 10-26% patients, urogenital anomalies occur in 21-34%, musculoskeletal deformities in 34%, Sprengel's shoulder occur in 40% of patients with cervical or cervico-thoracic scoliosis. Six common abnormalities associated with vertebral malformations are cranial nerve palsy, radial hypoplasia, club feet, dislocated hip, imperforate anus, and hemi facial microsomia. Fascio-auricular vertebral syndrome and VACTERL (vertebral, anorectal, cardiac, Tracheo-Esophageal, renal, limb anomalies) are association with congenital scoliosis. Intraspinal anomalies like syringomyelia, tethered spinal cord, lipoma of filum terminal, and diastomatomyelia, are detected in varying studies with a range of 20-58% [5,6].

According to Winter, hemivertebrae can be sub classified into:

1. Segmented hemivertebra - Both superior and inferior end plates of hemivertebrae have growth potential. Shape of adjacent vertebrae are normal.

2. Semi segmented hemivertebrae - Either upper or lower endplates has growth potential. The other end is fused with adjacent vertebra.

3. Incarcerated hemivertebra - Bother upper and lower endplates have growth potential, but the adjacent vertebrae accommodate for the growth and has abnormal shape of adjacent vertebra.
4. Non-segmented hemivertebra - There is no growth potential. It is fused with vertebra above and below.

50% of congenital scoliosis progress rapidly [7], 25% show mild progression, and 25% do not progress. McMaster and Ohtsuka [8] identified four key risk factors for current progression. They are type of anomaly, site of anomaly, age at diagnosis and number of curves. Maximum progression occur in unilateral unsegmented bar with contralateral hemivertebra [9]. Among the isolated failure of formation a fully segmented hemivertebra pose the greatest risk of progression.

Cervico-thoracic and upper thoracic curves produce greater cosmetic deformity due to neck tilt and shoulder imbalance. In due course secondary curves develop in the lower spine and would increase faster than the primary curve. Hemivertebra produce curves with most rapid rate of progression, ranging from 2-5 degrees per year before puberty, 3.5 degree per year after puberty creating significant trunkal imbalance. The trunkal imbalance can lead to list, pelvic obliquity, leg length discrepancy [8]. Lumbosacral hemivertebrae can cause "oblique take off" and the severe secondary curve with significant rotation in the upper spine. Progression occur between 1-5 years and at pubertal growth spurt. Deterioration of secondary and tertiary curve also occur with growth.

The vertebrae are formed from the condensation of paraxial mesoderm around neural tissue [10] during intrauterine life. Only one-third of the vertebrae are ossified at birth, by five years two-third of the vertebrae are ossified.

By first year the posterior arches begin fusion and complete by three years. By three years the anterior and posterior elements of the vertebrae begin fusion and completes by seven years. The neuro central physis located at he junction of the pedicle and vertebral body is important in the spinal growth. The physis fuses by 9 years of age. The spinal canal achieves 95% of its adult size by 5 years of age. So perivertebral arthrodesis does not significantly affect the dimensions of the spinal canal [11, 12]. Sitting height of a patient directly correlates with trunkal height and may be monitored to assess the spinal growth. A thoracic spine height of 18-22 cm is necessary for normal respiratory functions (normal approximately 27 cm at maturity). A premature arthrodesis of the thoracic spine leads to stunting of the growth of the thoracic spine and can lead to severe respiratory insufficiency [13]. Lumbar spine growth reaches 90% by 10 years of age. So, fusion of lumbar spine after the age of 10 years causes minimal loss of sitting height.

Treatment Options

In 1955 corrective casting was done by Riser using a three point translational force focusing on the apex of the curvature [14]. This would allow longitudinal growth of the spine and an increase in thoracic volume, with a simultaneous attempted 3-D correction of the deformity. Casting should be initiated for EOS (Early Onset Scoliosis) children less than five years of age with more than 10 degree documented progression.

Optimal timing of definitive fusion is not clearly defined [15]. Usually done when adequate chest wall, lung and spinal growth is achieved, or when complications like implant failure or infection are difficult to control. Usual age group is by 10-11 years.

Bracing in congenital scoliosis is mainly to prevent progression of secondary curves. The curves in congenital scoliosis are short and non-flexible. These curves that progress and cause trunkal and spinal imbalances require surgery to achieve safe correction, sagittal balance and to arrest curve progression. The surgical option depends on the type of vertebral anomaly, degree of deformity, age of the patient and skeletal maturity. The procedures for congenital scoliosis can be those that correct the deformity and those that prevent future deformity. In situ fusion prevents further deformity. Convex hemi epiphysiodesis allows gradual correction of the deformity. Similarly hemivertebra excision and osteotomy correct

the deformity acutely. In short rigid curves involving less than 5 vertebrae can be fused early to prevent future severe curves. Curves involving more than 5 vertebrae are fused late to avoid short trunk and thoracic insufficiency syndrome. Instrumented hemivertebrae excision provides the highest rate of correction, particularly if performed before the age of 3 years [16].

In situ fusion is due for cases involving less than 5 vertebrae which is predicted to worsen severely. It can be done for fully segmented hemivertebrae or hemivertebrae associated with contralateral unsegmented bar. Hemivertebrae at the cervico-thoracic or lumbosacral junction are prone for unsightly deformities. These require early fusion to prevent later decompensation. In situ fusion can be done anteriorly, posteriorly, or by combined approaches. Discs above and below the hemivertebra should be excised to decrease the chance of crank shafting and better flexibility of the curve and hence the correction.

Hemiepiphyseodesis is arresting of growth on the convex side of the curve. It may be due for failure of formation where there is growth possible on the concavity of the curve, which would gradually correct the same. It is not indicated for failure of segmentation (unsegmented bar) where there is no growth potential left on the concave side. This procedure should be done for smaller curves at an earlier stage preferably before 5 years, so that there is growth period left for gradual correction by growth in the concave side. Hemiepiphyseodesis is recommended in children less than 5 years of age with single segmented hemivertebra with curves less than 50 degrees. Presence of kyphosis is a relative contraindication as it would prevent anterior growth and gradual correction of kyphosis. Convex hemiepiphyseodesis involves the excision of one-third to half of the intervertebral disc and end plates below and above the hemivertebra on the convexity of the curve. Posteriorly facetectomy and decortication of lamina on convex side should be done. Concave side curve is not exposed to prevent spontaneous fusion. Through the pedicle the hemivertebra and its end plates and adjacent discs on the convex side of the deformity are removed. An autogenous iliac crest graft is used for fusion of the convex side. A brace or cast can be used to maintain the correction for at least 6 months. This procedure is useful at cervico-thoracic junction where exposure anteriorly is not easy.

Hemivertebra Excision

A fully segmented hemivertebra at the thoracolumbar, lumbar, or lumbosacral area can cause significant imbalance and large compensatory curves. So, they are treated with hemivertebra excision before 4-5 years of age, where the deformity is localized and without secondary curve [17,18,19]. This offers immediate correction without significant neurological risks [20,21]. The hemilamina, the transverse process, posterior half of the pedicle are removed.

The annulus on the concave side may be preserved to prevent lateral subluxation of the spine. Spine may be instrumented unilaterally or bilaterally to the vertebra above and below the hemivertebra. Pedicle screws, hooks, wires etc. may be used. The rest of the hemivertebra may be curretted out, discs above and below the hemivertebra is also resected. Iliac crest graft is placed between the adjacent vertebrae and the vertebrae compressed together through the screw rod system. If unable to instrument, postoperative casting (spica cast) and cast wedging may be done. Possible complications with hemivertebra excision are bleeding (epidural/segmental vessel bleeding) [22], neurological injuries, lack of suitable pediatric implants (low profile implants for skinny children. Stable fixation of vertebrae, difficulty in wound closure, wound healing problems and anesthetic risks, newer low profile implants may be developed to prevent many of these complications. In severe fixed and unacceptable deformities, osteotomies are a possibility. In children with failure of segmentation having

severe deformity, osteotomy anteriorly as well as posteriorly may be required. A good bone grafting is critical for a good result.

Conclusion

Isolated congenital fully segmented hemivertebra is the most common congenital spinal anomaly. The aim of management of congenital scoliosis is to achieve maximum spinal, trunkal, chest wall, and lung growth without worsening the curve and correct the spine in safer limits. Many procedures from casting, non-fusion techniques to differential fusion and fusion techniques are described, but judicious and tailor made application of the procedures depending on the age, anomaly, severity of curve, growth potential etc along with good fusion techniques are of paramount importance in the success of treatment in EOS. There are still a lot of scope for development of very low profile implants for very skinny children. The pros and cons of treatment should be made in favor of these young kids when EOS is approached. Intraspinal anomalies possible in these patients should be addressed before correction is embarked. Osteotomies are recommended for complex deformities. Hemivertebra excision for a fully segmented one offers acute correction of the deformity & avoids development of secondary curves most of the time. Thoraco-lumbar and lumbosacral and cervico-thoracic junction hemivertebrae have the potential for gross deformity in the future. These hemivertebrae can be excised at a very young age before the development of secondary curves for excellent results. Here, in this case, the hemivertebra is excised at one and a half years with a follow up of 11 years, even though reported literature suggests hemivertebrae excision after two years of age.

References

1. Dickson RA. Early Onset Idiopathic Scoliosis Vol. 1. New York: Raven; 1994.

2. Erol E. Kusumi K Lou J Dormans JP. Etiology of congenital scoliosis. The University of Pennsylvania Orthopaedic Journal 2002;15:37-42.

3. Wynne-Davies R. Congenital vertebral anomalies: aetiology and relationship to spina bifida cystica. J Med Genet 1975;12(3):280-288.

4. Winter RB. Congenital Deformities of the Spine patients. New York NY: Theme-Stratton; 1983.

5. Loder RT, Hernandez MJ, Lerner AL, et al. The induction of congenital spinal deformities in mice by maternal carbon monoxide exposure. J Pediatr Orthop 2000;20(5):662-666.

6. Belmont PJ Jr. Kukio TR. Taylor KF. Freedman BA Prahinska JR. Kruse RW Intraspinal anomalies associated with isolated congenital hemivertebra. the role of routine magnetic resonance imaging. J Bone Joint Surg Am 2004:86-A/8 1704-1710.

7. Winter RB. Lonstein JE Boachue-Adyes 0. Congenital spinal deformity Instructional Course Lectures. The American Academy of Orthopaedic Surgeons JBJS 1996:78A/2: 300-311.

8. McMaster MJ. Ohtsuka K The natural history of congenital scoliosis. A study of two hundred and fifty-one patients. J Bone Joint Surg Am 1982-64(8): 1128-1147.

9. McMaster MJ Congenital scoliosis caused by a unilateral failure of vertebral segmentation with contralateral hemivertebrae. Spine 1998:23(9) 998-1005.

10. Morcuende JA, Sanders JO. Embryology and development of the neuromuscular apparatus, In: V Weinstein SL, Flynn JM. eds. Lovell and Winter's Pediatric Orthopedics. 7th ed. Netherlands: Lippincott Williams & Wilkins;2013.

11. Dimeglio A. Growth of the spine before age 5 years. J Pediatr Orthop B 1993;1:102-107.

12. Dimeglio A, Canavese F. The growing spine: how spinal deformities influence normal spine and thoracic cage growth. Eur Spine J 2012;21(1): 64-70.

13. Karol LA, Johnston C, Mladenov K, Schochet P. Walters P. Browne RH. Pulmonary function following early thoracic fusion In non-neuromuscular scoliosis. J Bone Joint Surg Am 2008:90(6):1272-1281.

14. Risser JC The application of body casts for the correction of scoliosis Instr Course Lect 1955:12: 255-259.

15. Flynn JM. Tomlinson LA Pawelek J. Thompson GH, McCarthy R Akbarnia BA; Growing Spine Study Croup. Growing-rod graduates: lessons learned from ninety-nine patients who completed lengthening. J Bone Joint Surg Am 2013:95(19): 1745-1750.

16. Repke M. Krtes M. Burda J et al Simple bony fusion or instrumented hemivertebra excision in the surgical treatment of congenital scoliosis. Acta Chir Orthop Traumatol Cech 2008.75(3) 180-184.

17. Callahan BC. Georgopoulos G. Eilert RE. Hemivertebral excision for congenital scoliosis. JPediatr Orthop 1997:17(1):96-99.

18. Klemme WR Polly DW Jr. Orchowski JR. Hemivertebral excision for congenital scoliosis in very young children. J Pediatr Orthop 2001:21(6): 761-764.

19. Ruf M, Harms J. Posterior hemivertebra resection with transpedicular instrumentation: early correction in children aged 1 to 6 years. Spine 2003.28 18):2132-2138.

20. Royle ND. The operative removal of an accessory vertebra. Med J Aust 1928;1:467-468.

21. Leatherman KD, Dickson RA, Two-stage corrective surgery for congenital deformities of the spine. J Bone joint Surg Br 1979:61-B(3):324-328.

22. Bradford DS, Boachie-Adjet O. One stage anterior and posterior hemivertebral resection and athrodesis for congenital scoliosis. J Bone joint Surg Am 1990 72(4)536-540.