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Review Article

Congenital scoliosis: What is new in the present decade?

Athanasios I Tsirikos[1]

[1]Consultant Orthopaedic and Spine Surgeon and Honorary Clinical Senior Lecturer, University of Edinburgh/Royal Hospital for Children and Young People, Edinburgh, United Kingdom

ABSTRACT

Congenital anomalies of the spine occur due to faults in the embryological development of the immature vertebrae as an isolated defect or in association with a systemic condition. The result of the presence of these abnormalities is an asymmetrical growth affecting the longitudinal development of the spine and often producing an aggressive deformity involving the coronal and/or the sagittal planes. Recognition of the anatomical nature of the vertebral anomalies, their location along the spine and relationship to adjacent levels can predict the risk of deformity development and curve deterioration. Treatment is recommended at an early stage for those vertebral defects that are likely to produce a progressive deformity. The key to successful treatment in congenital spinal deformity is to diagnose in a timely manner those patients who are at risk to develop a severe deformity and apply early, prophylactic surgical treatment when the associated risks of major complications are more limited.

Key words: Congenital kyphosis, Congenital scoliosis, Growing rods, Hemivertebra resection, Surgical treatment, Vertical expandable prosthetic titanium rib, Vertebral anomaly

INTRODUCTION

The aim of this review article is to provide an update and present essential guidelines that can be helpful for clinicians treating patients with congenital spinal deformities.

Embryology: Congenital scoliosis can develop due to a range of congenital vertebral abnormalities that produce an imbalance in the longitudinal growth of the immature spine.[1-3] The congenital anomalies impacting on the development of the vertebral column can affect one or multiple levels and carry a varying prognosis. These congenital defects occur during the first 6-8 weeks of embryologic growth and are caused by disruption of the notochord and somites that are responsible for the development of skeletal muscles, dorsal dermis, tendon, ligaments but also play a role in the respiratory, cardiac and genito-urinary system formation. It should, therefore, be appreciated that a multidisciplinary assessment of a patient with congenital vertebral abnormalities is required in order to identify medical co-morbidities beyond the development of the spinal column.

Classification: Having a classification system for congenital spinal deformities can determine the prognosis of the pathological curvature on the basis of the type, number and location of congenital vertebral anomalies, as well as their relation to the adjacent levels of the spine and the presence of contralateral spinal growth that acts as the deforming force that precipitates the deformity. The classification of vertebral abnormalities into failures of formation, failures of segmentation and mixed anomalies provides a useful tool to define the natural history and risk of progression of the curve and communicate treatment outcomes.[4,5]

Assessment: The use of prenatal ultrasound or magnetic tomography imaging (MRI) can allow early diagnosis of congenital vertebral anomalies and timely referral to specialist services. Post-natal assessment involves plain radiographs, computer tomography scans with 3D reconstruction to define the nature of bony defects affecting the vertebral bodies and posterior arches, and MRI to evaluate for intraspinal anomalies.[6,7] The spinal MRI can provide imaging of the kidneys and if any abnormality is identified a renal ultrasound is recommended. A cardiac evaluation including echocardiogram and cardiac ultrasound can be organised to exclude congenital heart disease. In the presence of congenital rib fusions or complex rib abnormalities a respiratory review is advised.[8] A genetic review should also be done to rule out an

***Corresponding author:** Dr. Athanasios I Tsirikos, MD FRCS PhD, Consultant Orthopaedic and Spine Surgeon and Honorary Clinical Senior Lecturer, University of Edinburgh/Royal Hospital for Children and Young People, Edinburgh, United Kingdom. atsirikos@hotmail.com

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underlying syndromic condition such as VATER, VACTERL, Goldenhar, spondylocostal/spondylothoracic dysplasia, Alagille.[9] In the presence of a congenital vertebral anomaly that is likely to produce a progressive deformity these tests should be performed at the time of index review so that there is no delay if surgical treatment is indicated.[10]

Prognosis: A fully **segmented hemivertebra** (most common failure of vertebral formation) is likely to produce a progressive scoliosis, kyphosis or kyphoscoliosis and require surgical treatment. The presence of 2 ipsilateral hemivertebrae produces a more progressive scoliosis and need prophylactic treatment. Two contralateral hemivertabrae (hemimetameric shift), especially those affecting transitional areas such as the thoracolumbar or lumbosacral spine create an unbalanced deformity and necessitate surgery. A semi-segmented hemivertebra produces a slowly progressive scoliosis and treatment may not be required for small curves unless these affect the lumbosacral junction. Lumbosacral hemivertebrae (fully or semi-segmented) create an oblique take-off of the spine and a structural compensatory curve in the lumbar spine that often progresses more than the congenital scoliosis. Early surgical treatment is commonly required to balance the congenitally abnormal spine before a compensatory curve develops.

A **posterior hemivertebra** or a posterolateral quadrant vertebra causing a congenital kyphosis or kyphoscoliosis carries an increased risk of spinal cord compression as the hemivertebra can grow towards the spinal canal producing neurological damage.[4,5] When radiographic imaging is organised during clinical follow-up it is, therefore, critical to include a lateral erect scoliosis view in order to assess for the presence or progression of congenital kyphosis as in this case surgical treatment should be offered before neurological symptoms develop. Congenital kyphosis due to a failure of formation can produce a displaced spinal canal. This occurs as a step-off deformity at the level of a subluxed or dislocated vertebral segment due to the hypoplastic vertebra. This type of deformity is progressive with the spine being highly unstable in the sagittal and axial planes resulting in greater risk of neural damage.

A **unilateral unsegmented bar** (most common failure of vertebral segmentation) produces a rapidly progressive scoliosis due to its tethering effect that impacts on the longitudinal spinal growth in the presence of normal contralateral end plates. The combination of a unilateral unsegmented bar with contralateral hemivertebra (most common mixed anomaly) at the same level carry the worst prognosis in regard to curve deterioration as the tethering impact of the bar is enhanced by accelerated growth in the presence of one or more hemivertebrae on the opposite side. Similarly, rapid curve progression is expected in the presence of an anterolateral unsegmented bar with or without contralateral hemivertebra producing a kyphoscoliosis.

Mixed congenital vertebral anomalies can affect large segments or different areas of the spine combining failures of formation and segmentation. Advances in imaging techniques allow better definition of the anatomy of these vertebral defects which can be very difficult to categorise. There is commonly an association with rib and chest wall abnormalities producing tethering of both the spinal and thoracic growth.

An **unsegmented or incarcerated hemivertebra** or a block vertebra carry very limited or no growth potential and do not create a deformity. An anterior unsegmented bar produces a congenital kyphosis with slow progression that often requires no treatment.

Conservative treatment: Observation is recommended in patients with non-progressive deformities in 6-12 month intervals depending on the stage of skeletal development. Children in their first 3 years of life and teenagers who are in their adolescent growth spurt are at maximum risk of curve deterioration and need to be monitored closely.

Bracing or casting is not effective to control congenital curves or impact on their long-term prognosis as these are usually rigid in the presence of vertebral abnormalities.[11] These treatment modalities are useful to prevent progression of structural compensatory scoliotic curves that develop at the levels proximal or distal to the congenital vertebral defects in an attempt to achieve global coronal spinal balance. Brace treatment can occasionally be used in patients with hemivertebra after resection and segmental fusion or in those with a unilateral bar after a convex fusion when the apical deformity has been controlled if the residual scoliosis extends to longer segments above and below the congenital anomaly.

Surgical treatment: Spinal surgery is indicated in the presence of a progressive deformity. Timing of surgery is of outmost importance and age of the patient is not necessarily a restrictive factor if the deformity is rapidly deteriorating. Prophylactic surgical treatment is recommended when dealing with a type of congenital vertebral abnormality which is expected to produce a highly progressive curve such as when a unilateral or anterolateral unsegmented bar with or without contralateral hemivertebra or a posterior/posterolateral hemivertebra exists. The location of congenital scoliosis in transitional areas of the spine can produce severe spinal imbalance and a deteriorating deformity necessitating early surgery.

Posterior in situ fusion is indicated in the presence of mixed congenital anomalies affecting transitional areas of the spine such as the cervico-thoracic and upper thoracic region and producing a mild scoliosis. This can stabilise the spine across the area of the vertebral defects and prevent progression of the congenital curve, as well as the development of a structural compensatory scoliosis at the levels below. A localised posterior fusion can also be performed when addressing a congenital kyphosis due to a posterior hemivertebra. In this case, the kyphosis should ideally be up to 50o and the child less than 5 years of age.[10] In the presence of residual anterior vertebral growth the posterior bony tether can allow for progressive gradual improvement of the kyphosis occurring with subsequent spinal growth. There should be a low threshold to consider a re-grafting procedure in order to address a non-union or reinforce the posterior fusion mass.

Anterior convex in situ fusion can be performed with the use of an autologous rib strut graft with the aim to stabilise the spine in the presence of a unilateral unsegmented bar. This will convert the congenitally abnormal area in a block segment that will stop deformity progression.

Anterior convex hemiepiphysiodesis (growth arrest procedure) is recommended in young children in the presence of a fully segmented hemivertebra involving the thoracic spine where hemivertebra resection can be associated with increased risks.[12] This should extend to include one level proximal and distal to the congenital anomaly. By preserving anterior concave vertebral growth gradual improvement of the scoliosis is expected to occur over time.

Posterior hemivertebra resection is the treatment of choice when addressing a deteriorating congenital deformity due to the presence of a lateral, posterior or posterolateral hemivertebra.[13,14] This involves complete excision of the hemivertebra (including the posterior elements at the affected level), contralateral disc, as well as the discs and adjacent end plates at the levels cephalad and caudal to the hemivertebra. The benefit of this procedure is that it removes completely the deforming force and can achieve rebalancing of the spine that is usually sustained over skeletal growth with no need for further surgery.

Instrumented posterior spinal correction and fusion can be done in patients who present at an older age with long-standing congenital deformities of any anatomical aetiology and normal neurology. The aim is to balance the spine by using adjacent level spinal flexibility above and below the level of the congenital vertebral abnormality. If the congenital deformity is more severe and rigid the addition of an anterior spinal release can improve the ability to correct the curve; this can also control the risk of crankshaft effect that can occur due to remaining anterior vertebral growth in young patients.

Complex spinal reconstruction *with the use of spinal osteotomies or vertebral column resection* may be required when dealing with severe neglected congenital curves that cause significant spinal decompensation often associated with neurological damage. These techniques will also be needed during revision surgery in the presence of an established fusion mass in order to treat residual or recurrent deformities. The risk of neurological and life-threatening (related to intraoperative blood loss) complications of such procedures is considerable and cannot be underestimated during decision making. The neurological risk is related to segmental mechanical instability produced at the level of vertebral column resection and resulting in spinal cord injury. Neural injury can also occur due to disruption of the vascular supply to the spinal cord at the level of the osteotomy/vertebrectomy.

Growth preserving techniques can be used in isolation in the treatment of congenital scoliosis or alongside the previously described techniques. The aim is to control the deformity at a young age and preserve spinal growth which corresponds to chest development. Good indications for using growing rods in the context of congenital scoliosis is as an adjunct to anterior convex fusion in patients with a unilateral bar (with or without contralateral hemivertebra) or anterior convex epiphysiodesis in patients with a fully segmented hemivertebra. In these cases, the growing rod can control residual adjacent level scoliosis at the levels above and below the congenital abnormality and guide spinal growth during skeletal development which can further improve the remaining deformity. The development of the magnetically controlled growing rods that can limit the number of repeat surgeries has made this technique more attractive but the currently available results have not met the initial high expectations. In addition, growing rod techniques either traditional or magnetically lengthened have a number of inherent complications which can be technical related to implant failure, associated with infection, reduced ability to lengthen the construct over time (so called 'law of diminishing returns'), as well as the development of crankshaft effect or proximal junctional kyphosis.

The vertical expandable prosthetic titamium rib was originally developed as a rib-to-rib construct to address thoracic insufficiency in young children with severe congenital scoliosis and rib fusions.[15] A rib-to-spine construct was subsequently used as an alternative to traditional growing rods as primary treatment of congenital scoliosis with controversial results and limited acceptance.

Hybrid growth preservation techniques have been introduced in the last few years as part of clinical trials. These are based on a combined concept of concave distraction using a growing rod or the spring system and convex growth guidance.[16,17] Even though the initial results are very promising more data is required to allow wider use of these techniques.

CONCLUSION

Successful treatment of congenital spinal deformity requires an early diagnosis and understanding of the nature of congenital abnormalities that carry increased risk to produce a progressive deformity. Surgical treatment depends on factors that include patient's age, remaining spinal growth, type and site of vertebral anomaly, type and size of curvature, as well as the development or impending risk of spinal cord compression. Anticipating deformity progression will allow selecting the correct procedure and applying it at the appropriate time. It is not possible to create vertebral growth where this does not exist. However, early surgery can balance spinal growth and prevent the development of a severe deformity and spinal cord compression which then require much more complex surgery associated with increased risks for major morbidity and potential mortality.

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