Expert Review

Journal of Trauma & Orthopaedic Surgery 2016; July - Sep; 11(3):5-11

Congenital Scoliosis and Kyphosis - A Brief Clinical Overview

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Abstract

Background: Spinal deformity which is secondary to malformed or absent vertebrae along with bony bar is called as congenital kyphoscoliosis. Congenital scoliosis is more common that the congenital kyphosis. The deformed curves are secondary to malformed vertebrae, absent vertebrae, accessory vertebrae and fused vertebrae which pose as bony bars. Because of nature of the deformity they are more severe and rigid as compared to idiopathic scoliosis. They form pure scoliosis, kyphosis, kyphoscoliosis and lordoscoliosis. Treatments of these deformities are challenging and require osteotomy and correction if the curves are progressing rapidly. It is paramount to diagnose and evaluate non progressive and progressive curves at early age.

Introduction

Etiology [1-6]:

Environmental factors like hypoxia, cigarette smoking, exposure to certain chemicals, fetal alcoholic syndrome, genetics, deficiency of folic acid, have been recognized as causative agents either singly or in combination. Formation and segmentation defect take place as early as between 3 to 8 weeks. Damage to somites during development leads to hemivertebrae, accessory vertebral formation and fused vertebrae [7-14].

1. An untreated scoliosis curve over 30 degrees in an immature child will likely progress to needing surgery at some time during the child's life. Curves increasing > 10 deg/year = 100% risk of needing surgery. (Alain DiMeglio)
2. Congenital scoliosis is an inherited disease and has familial association
3. Homobox genes are as well shown to be responsible for segmentation defect. Mutation of these genes lead to congenital spinal deformities

Red Flags for Identification of congenital scoliosis/ spinal deformity [15-19]

1. Child with a abnormally stiff Back
2. Abnormal foot/ associated foot deformity
3. Chest wall deformity
4. Skin Pigmentation e.g. Café au lait spots
5. Imperforate Anus or Anal sinus
6. Painfull scoliosis in young patient (suggestive of dural malformations, like dural ectasia)
7. Short stature, abnormal size head with facial asymmetry

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Classification:

Congenital scoliosis is classified into further types depending upon structural integrity

1. Hemivertebra (Defect of formation)
   - Unilateral defect of formation- complete Hemivertebra
   - Semisegmented
   - Incarcerated
   - Non-segmented
2. Unsegmented Bony Bar (Unilateral failure of Segmentation)
3. Block vertebra (Failure of segmentation)

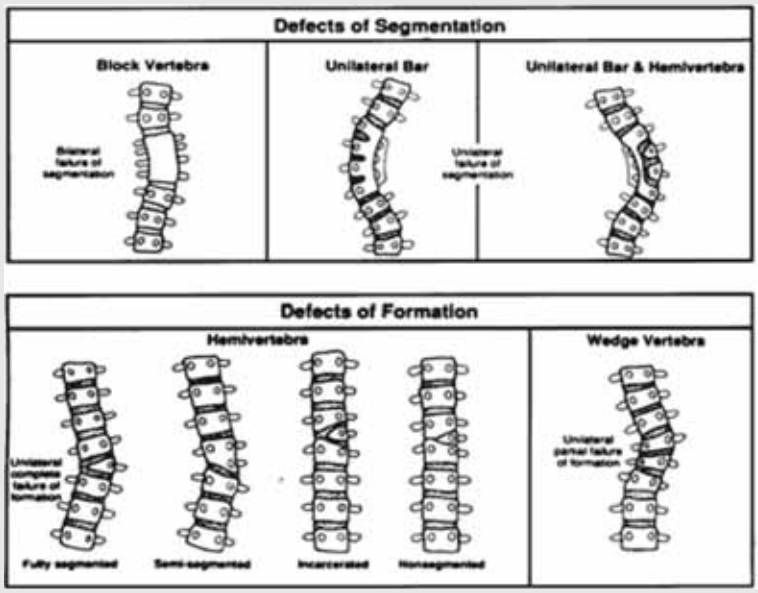
Natural History of Congenital Scoliosis [18-19]

Progression of congenital scoliosis is approximately seen in 70 % individuals. The rapid nature depends upon the type of defect. Hemivertebra has progression potential because of wedged body and occurrence of unequal growth. The cases in which there is unilateral segmented bony bar with contralateral hemivertebra has the worse prognosis and definitive risk of rapid progression. This type of anomalies, surgery is the sole answer. As against that, cases in which there is incarcerated vertebra or Block vertebra, have lower risk of progression and mostly get managed by conservative measures. Location of anomaly has impact on progression of the curve, upper thoracic anomalous vertebra has lower risk of progression as compared to hemivertebra present at thoracolumbar or Cervicothoracic junctional level.

Following factors decide the natural progress of the scoliotic deformity:

1. Type of anomaly
2. Location
3. Number of anomalies and involved vertebral segments]

 | pISSN 2278 – 6651



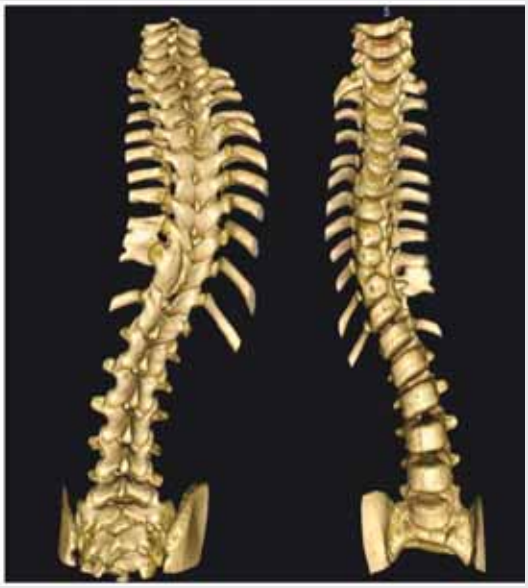


Figure 1: Kawakami also gave 3 D classification which gave 4 types which should help in surgical planning. 1-Solitary Simple, 2- Multiple Simple, Multiple complex, 3- Mismatched Complex, Mixed complex, 4-Segmentation Failure.

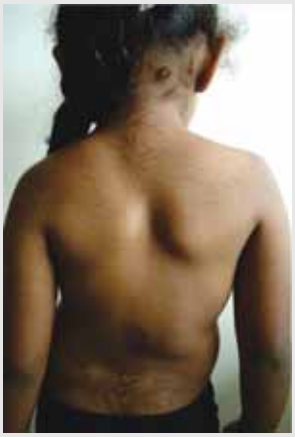


Figure 2: During clinical evaluation, it is paramount to see gait of the patient. Sagittal balance, coronal balance assessment need to be done. Shoulder mal-alignment, truncal shift, head over pelvis need to be assessed. Motor and sensory examination and look for skin pigmentation
A- Both shoulder balance must be assessed from front as well back of the patient. In this case right shoulder appears to be at higher level
B- Right side Rib Hump with prominent higher level scapula. Scoliotic curve seen on right side with Kyphotic sagittal imbalance seen at thoracolumbar junction
C- History of previous surgery, surgical mark of de-tethering seen
D- Pelvic Obliquity- Right side lower than left side

Figure 3: Patient has standing difficulty, because of hip problem. Patient examined in sitting position.
A- Shoulder Balance is assessed
B- Left side rib Hump with congenital Kyphoscoliotic curve more on left side
Scar of previous surgery, operated case of myelomeningocoele

4. Initial severity of the curve and remaining growth potential
5. Associated Chest wall deformities

Clinical Evaluation of Patient

Cross sectional studies [20-27]

They involve Magnetic Resonance Imaging (MRI) and Computed Tomography (CT scan). Both the investigations are required for specific evaluation. For Spinal cord anomaly, malformation, and visualisation of soft structures, MRI is the gold standard. CT scan is performed to delineate the bony bars, diastomatomyelia, fibrous and bony septums in the canal.

MRI scan is carried out in cases of congenital Scoliosis to rule out following things:

A- Tethered Cord and spinal cord anomaly
B- Fibrous, complete or incomplete diastomatomyelia
C- Low lying tonsils
D- Syrinx present in the thoracic cord
E- Y shaped disc in defect of formation, hemivertebrae

Management of Congenital Scoliosis [28-33]

Conservative Options-

Bracing and casting is advisable to prevent further progression of compensatory curves. In case of wedged hemivertebra with congenital bony bar on contralateral

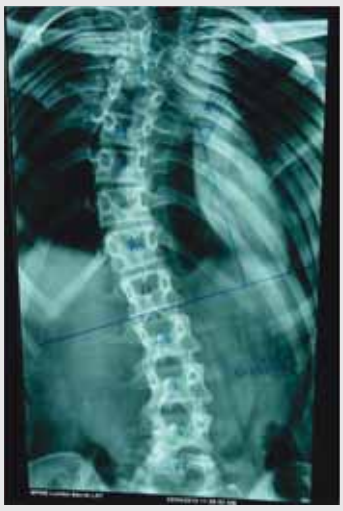


Figure 4:
- Whole spine standing radiographs are essential.
- Right thoracic scoliotic curve extending from D2 to L1 with apex of the curve at D10-D11 disc space
- A- At the Upper thoracic vertebra there is failure of segmentation and thus block vertebra is present. It is as well associated with crowding of the ribs on both the side of curve
- Radiological assessment should calculate Cobb s angle, shoulder asymmetry, RVAD, pedicle rotation and pelvic obliquity

Case 1

Figure 5: Kawakami also gave 3 D classification which gave 4 types which should help in surgical planning. 1-Solitary Simple, 2- Multiple Simple, Multiple complex, 3- Mismatched Complex, Mixed complex, 4-Segmentation Failure.

Case 2

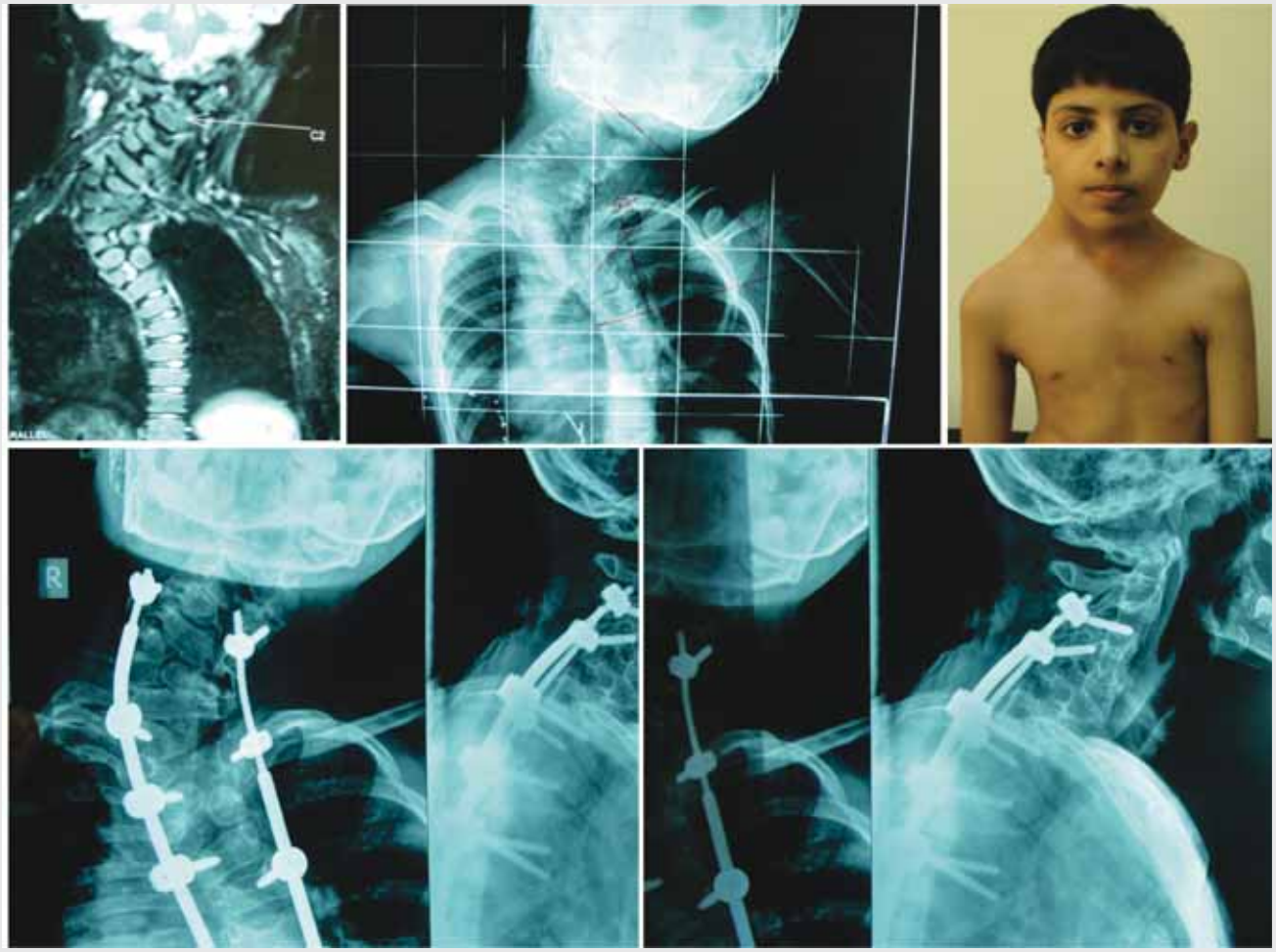


Figure 6: Multiple Complex hemivertebrae leading to congenital scoliotic deformity in cervicothoracic area. Apex of the deformity in cervicothoracic junction leading to obnoxious deformity. This X ray shows segmentation defect, hemivertebrae, unilateral unsegmented bony bar. Surgical correction was done by doing unilateral pedicle subtraction osteotomy and deformity correction, spinal Fusion. Surgical correction was aimed at arresting further deformity progression and solid fusion. Spinal instrumentation and fusion has been extended into cervical area. Cervicothoracic curves are very challenging to correct. Getting the Head over shoulders and correcting torticollis is the objective.

side , non-operative treatment has no role. In treatment of congenital scoliosis, only block vertebrae and incarcerated hemivertebrae to some extent needs conservative treatment. Non-operative treatment essentially involves clinical monitoring of static vertebral anomaly.

Surgical Treatment

Treatment depends on mainly 2 principles-

a. Removal/excision of deforming force(vertebra/ Y shaped disc)
b. Fusion at the apex of deformity and allow further growth of spine

Till date surgical approach was dictated by the presence of anomolous vertebra and bony bar. If hemivertebra is located posteo-laterally then, posterior approach was preferred. Anterior apporoach has advantage of prevention of crank shaft and limiting surgery to shortest possible levels. If Posterior surgery is performed under the age of 9 years in more than 4 levels of thoracic fusion, then earlier it was said that it leads to restrictive Lung disease. Long fusion surgeries limits the pulmonary function. Now a days most of the surgeries are performed through posterior approach because of its modification and vast usage of pedicle subtraction and Posterior column osteotomies. Objective of the surgery in congenital scoliosis is to achieve apical fusion involving shortest segments and levels. In growing spine, in addition to apical fusion, guided distraction and growth of the spine has been achiveed with Non fusion techniques like- Dual growth rods, SHILLA, and VEPTR. Different Surgical options are discussed with following cases. Every case of verebral deformity is different and requires taiolred made surgical option keeping principles of surgery in mind [34-35].

These vertebral anomalies are associated with other congenital anomalies like VACTERL, vertebral defects, anal atresia, cardiac anomalies, tracheo-oesophageal fistula, radial club hand, renal problems and limb deformities. It is known to have genitourinary problems associated with vertebral anomalies, the reason being the development of Spine coincides with development

Case 3

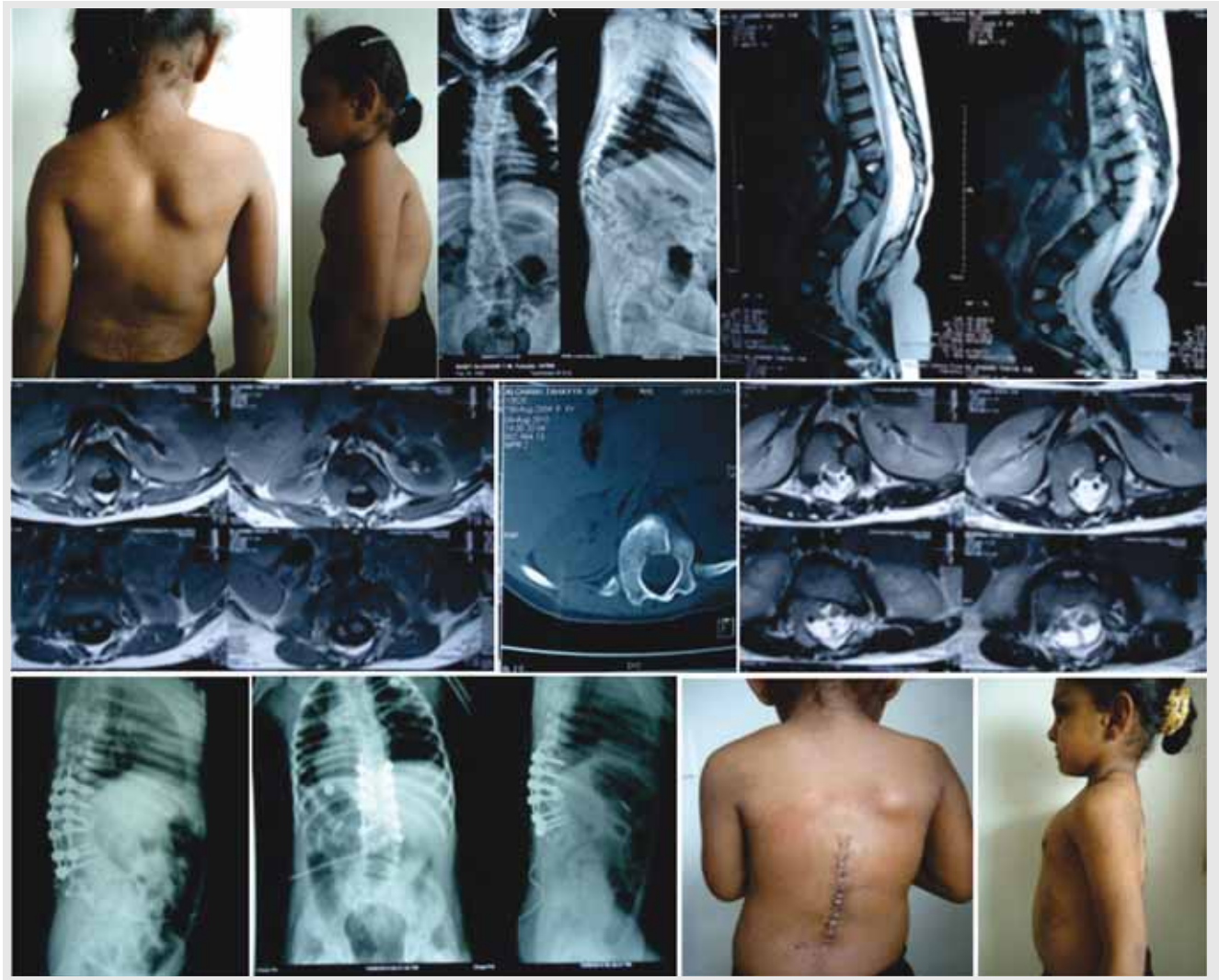


Figure 7: 5 year old child with congenital kyphosis and tethered cord. Failure of segmentation anteriorly, leading to anterior bony tether with angular kyphosis. Cord was detethered at first stage and then with Smith Peterson osteotomy, deformity correction and spinal fusion achieved by posterior approach. Preoperatively D11, D12 and L1 vertebrae were fused anteriorly by bony bar leading to angular kyphosis. After Osteotomy anterior Column was lengthened by reconstructing Anterior harms Mesh cage and Posteriorly vertebral column was shortened with Pontes osteotomy and posterior pedicle screw tension band.

Case 4

Figure 8: 5 year old girl with an incarcerated hemivertebra along with progressive congenital scoliosis was operated with posterior hemivertebra excision and then short posterior fusion.

Case 5

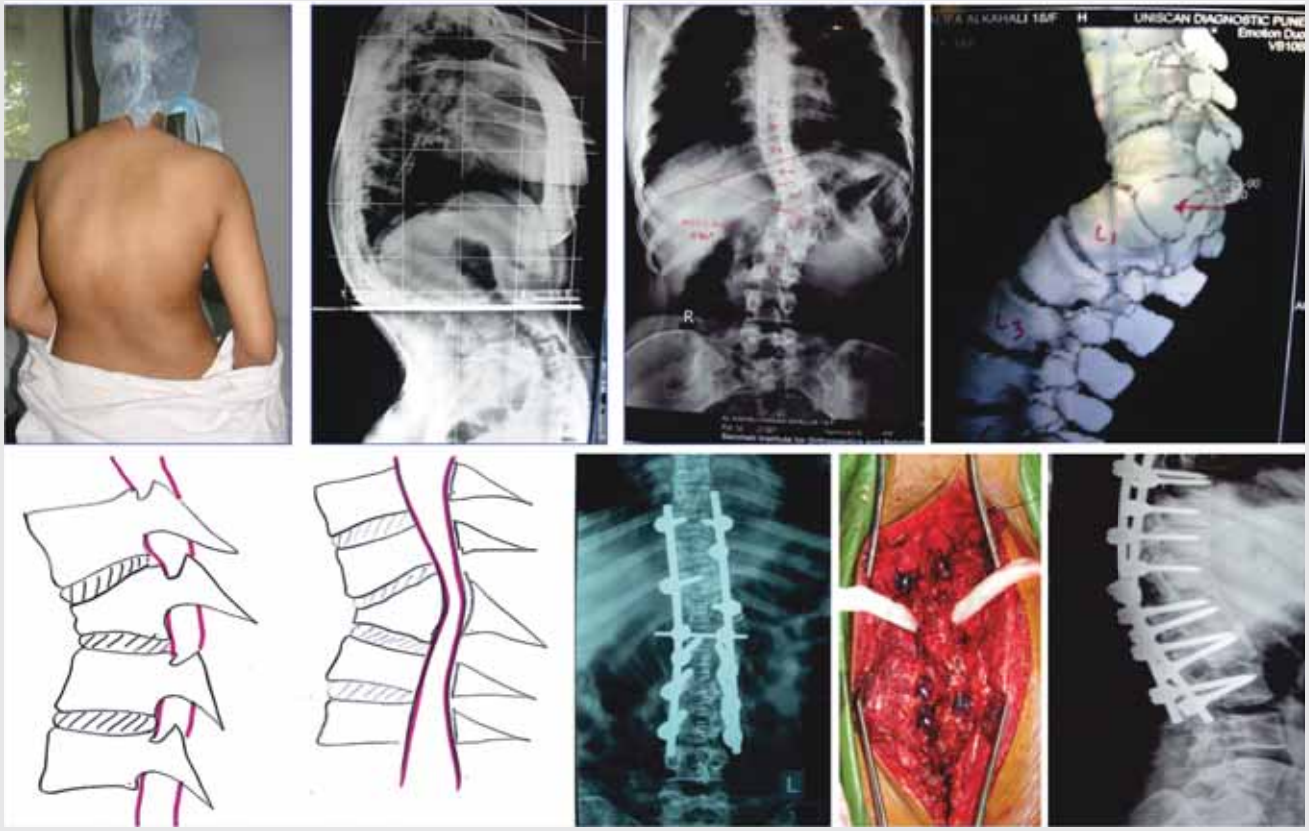


Figure 9: 19 year old girl with posterolateral based L1 hemivertebra leading congenital Kyphoscoliotic deformity. Patient was operated with posterior based Hemivertebra excision, Pedicle subtraction Osteotomy on contralateral side with posterior column shortening with anterior bone to bone contact. This deformity correction procedure corrected kyphoscoliotic deformity in one stage.

Case 6

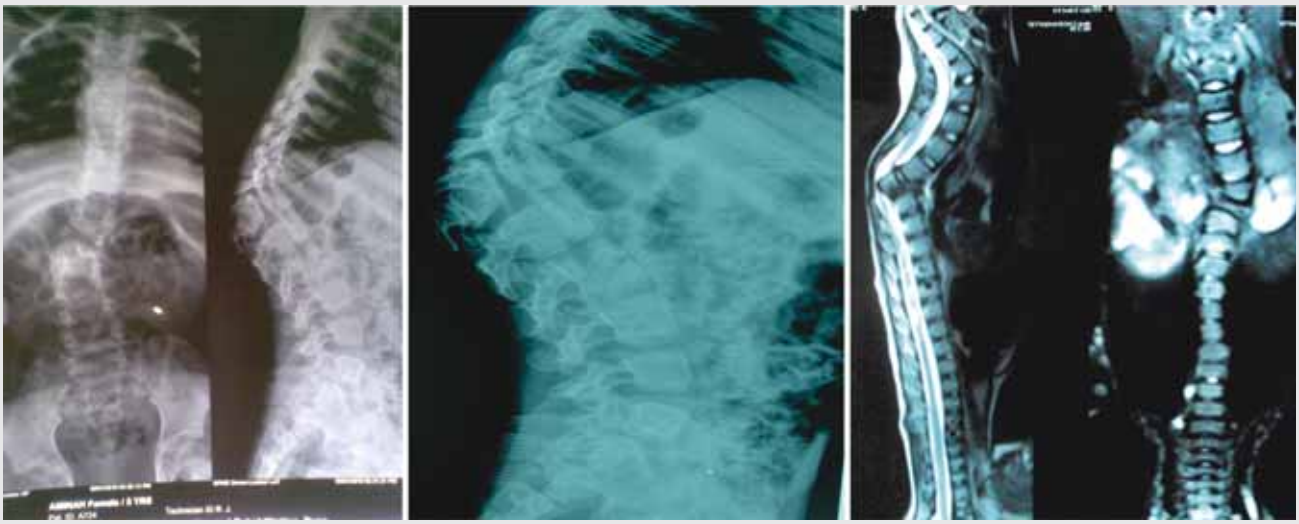


Figure 10: Congenital Angular Kyphosis in a 4 year old child. There is defect of formation anteriorly. It has lead to only formation of posterior elements and classical Y shaped disc is seen anteriorly on xray and MRI image. At the age of 4 years there is 100 degree of angular kyphosis. These patients should not wait till maturity and should be operated at the earliest.

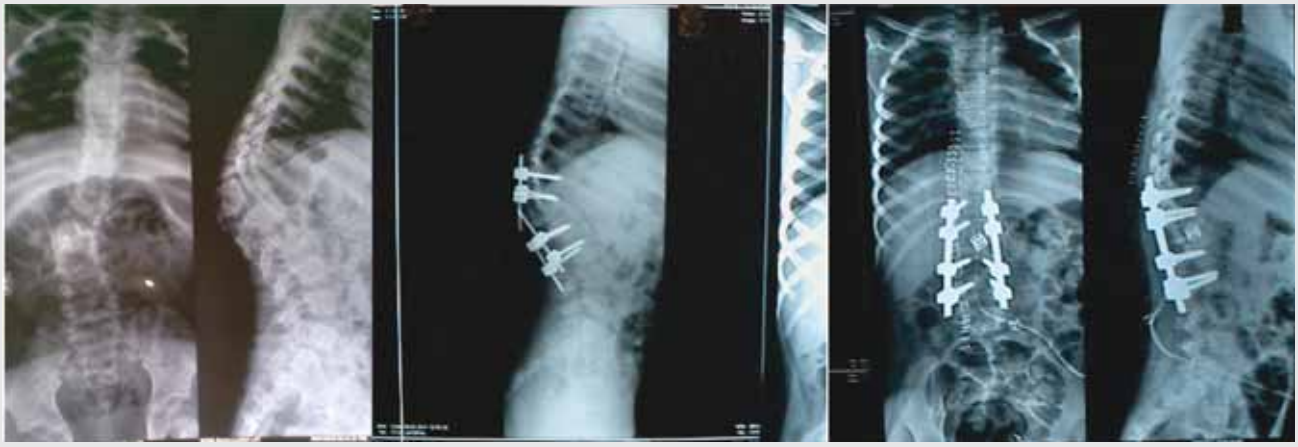


Figure 11: In the index surgery, Y shaped disc was excised, Angular Kyphosis was corrected. Anteriorly bone to bone contact was achieved with Tricortical anterior strut graft and posteriorly Pedicle screw stabilisation and deformity correction was performed. Patient had a fall at the end of 3 months and implants broke.
Patient was revised with stouter pedicle screws, anterior column was lengthened and reconstructed with cage to prevent cord buckling and reduce the stress over posterior implants.

of multiple Organ System.

Pearls:

1. Congenital scoliosis and kyphosis is as a result of failure of formation and failure of segmentation
2. Assessment of vertebral anomalies should be done in childhood and associated anomalies should be looked for, especially genitor-urinary system.
3. Bracing has no role in congenital scoliosis and Kyphosis. Bracing is done to monitor static structural anomalies
4. Surgery should be done at the earliest to prevent future structural changes. Unlike idiopathic scoliosis, deformity correction should not wait till maturity
5. First radiograph should be compared to assess rapidity of progression of the curve
6. MRI is must in cases of congenital scoliosis if rapid progression is observed or before any surgical intervention
7. Congenital anomalies of vertebral body, especially posterior elements, demand 3 D CT scan for evaluation of structural abnormality
8. Neurologic risk is higher with congenital curves as compared to idiopathic ones
9. Objective of the surgery is to arrest rapid progression of the curve, apical fusion and sagitally and coronally balanced spine

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Conflict of Interest: NIL
Source of Support: NIL

How to Cite this Article

Khurjekar K, Khurjekar D, Hadgaonkar S, Kardile M. Congenital Scoliosis and Kyphosis - A Brief Clinical Overview. Journal of Trauma and Orthopaedic Surgery. July - Aug 2016;11(3):5-11