



## Congenital scoliosis : Management and future directions

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**Congenital scoliosis remains an interesting and challenging diagnostic entity. Vertebral absence, partial formation or lack of segmentation may cause asymmetrical growth and resultant deformity. Because of the high frequency of associated anomalies within and outside the spine, a detailed history and physical examination are mandatory. Maternal, perinatal history, family history, and developmental milestones must be fully explored.**

**Plain radiographs remain standard for diagnosis of congenital anomalies and measuring curve magnitude, progression and perhaps growth potential of the vertebral anomaly. Preoperative CT scan defines the anatomy and avoids any unexpected intraoperative posterior element deficiencies. MRI can exclude associated conditions of the spine, cranio-cervical junction and viscera. The recognition of curves with a bad prognosis at an early stage is pertinent to prevent curve progression and possible neurological complications.**

**The goal of surgery is to achieve a straight spine and a physiological sagittal profile while maintaining flexibility, to arrest progression of the curve with a short fusion segment preserving as much normal spinal growth as possible.**

**Developments in gene research continue to be promising and may potentially lead to early detection of congenital vertebral malformations.**

**Keyword :** congenital scoliosis.

### BACKGROUND

Congenital scoliosis (CS) is defined as a lateral curvature of the spine due to a developmental

abnormality. Scoliosis present at birth that is not associated with an underlying developmental anomaly is referred to as infantile scoliosis. The term CS implies that there is always spinal curvature at birth, but only the vertebral malformations are present at birth in all patients, while the scoliosis for some may not develop until later.

An incidence of approximately 0.5 to 1/1,000 births has been observed for CS (37). With an isolated single vertebral malformation, the chance of a first degree relative having a similar anomaly was estimated to be approximately one out of a hundred in one study (45), but no pattern of inheritance was found in another report (45). If multiple vertebral anomalies are present, then the risk of similar anomalies in either siblings or children of the patient is between 10 to 15% (45).

Genetic inheritance has been shown responsible for some congenital vertebral anomalies ; however, there is no clear-cut genetic aetiology of CS to

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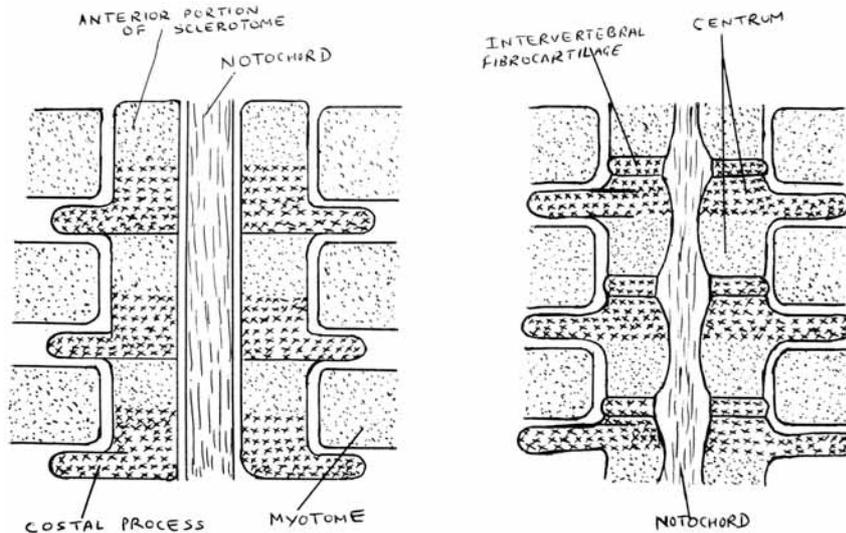


Fig. 1. — Vertebral centrum development from two adjacent segments.

date (15). Basic science research evidence in mice suggests that maternal exposure to toxins, such as carbon monoxide exposure, may cause congenital scoliosis (16). Associations with maternal diabetes and ingestion of antiepileptic drugs during pregnancy have also been postulated as possible causes (16).

The vertebral column develops from pairs of somites, which begin to appear at 3 weeks gestation. The somites are mesenchymal segments, which are on both sides of the neural tube, and the antero-medial wall of the somite is termed a sclerotome. Cells from the sclerotomes spread out centrally to form an unsegmented, cellular perichordal sheath, which eventually forms the centrum of the vertebral column. Next a zone of loose cells in the sclerotome forms superiorly where the intersegmental and spinal nerve pass through the plate and a dense zone forms inferiorly which goes on to become the posterior neural arch of the vertebra and the rib (fig 1). One sclerotome pair forms one level of ribs and posterior elements of the spine. In the cellular sheath of the notochord, alternating zones of loose and dense zones also appear, but the superior zone of loose cells goes on to form the centrum of the vertebra, while the inferior dense zone goes on to form the intervertebral disk.

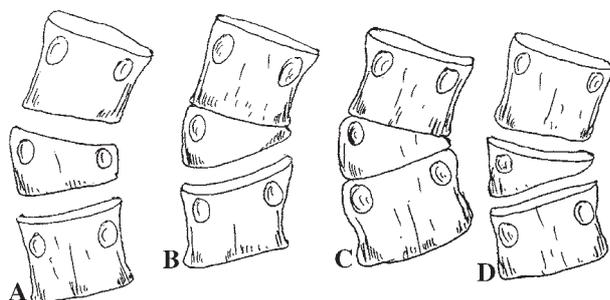
Various theories to explain congenital vertebral malformations have included failure of ossification as the cause of defects of vertebra formation, osseous metaplasia of the annulus fibrosus as the cause of defects of vertebral segmentation, and vertebral development hindered by persistent notochord (5).

## CLASSIFICATION

Moe *et al* (35) classified congenital scoliosis according to morphologic characteristics on plain AP frontal and lateral images as an embryological defect into formation failure, segmentation failure, and a mixed type. This classification also includes several other factors, such as the level of formation failure and the presence or absence of intervertebral disc space. Based on this information, the natural history of congenital scoliosis and strategic approaches to its treatment have been evaluated and reported (35).

The failures of formation (fig 2) are characterised by the deficiency of a portion of a vertebral element, causing 1) hemivertebra (complete) or 2) wedge vertebra (partial).

Wedge vertebrae present with a height asymmetry, with one side being hypoplastic, there are

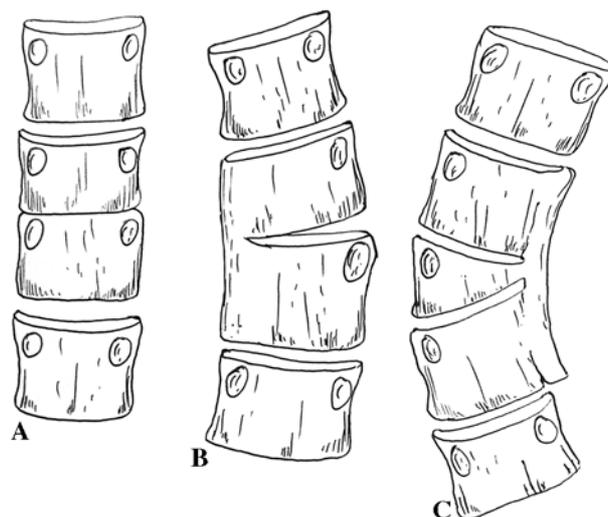


**Fig. 2.** — Representation of formation failures : (A) wedge vertebra (B) Fully segmented hemivertebra (C) Partially segmented hemivertebra (D) Unsegmented hemivertebra.

two pedicles. Next in severity is a hemivertebra, with the absence of one pedicle and a region of the vertebral body. Hemivertebrae may be further classified on the basis of the presence or the absence of fusion to the vertebral bodies above and/or below (29). An unsegmented hemivertebra is fused to the vertebral body above and below ; a partially segmented hemivertebra is fused to the vertebral body either above or below ; and a fully segmented hemivertebra is separated from the body above and below by disk space. Hemivertebrae may occur at ipsilateral adjacent levels of the spine, which produces significantly asymmetrical spine growth, or one hemivertebra may be counterbalanced by another hemivertebra on the contralateral side of the spine in the same region, separated by one or several healthy vertebrae (termed as hemimetameric shift) (38,41). This anomaly occurs most commonly in the thoracic spine.

The defects of segmentation are characterised by abnormal bony connections between vertebrae (fig 3). These bony connections may be bilateral and symmetrical, resulting in a block vertebra. Segmentation defects caused by unilateral bony fusions are termed bars and may act as a unilateral growth tether. Occasionally, a segmentation defect may span an ipsilateral formation defect, resulting in a unilateral bar and a contralateral hemivertebra (24).

The third category or mixed type comprises the complex anomalies that include both segmentation and formation errors and at first may be difficult to define, as the spine is only 30% ossified at birth.



**Fig. 3.** — Representation of segmentation failures : (A) block vertebrae (B) Partially unsegmented bar. (C) combination of hemivertebra and unsegmented bar.

Hemivertebrae usually occur as extra spinal segments and are often accompanied by an extra rib. They may result from an abnormal cleavage of the primary chondrification center. Wedge vertebrae do not present as extra segments or ribs and thus may involve a unilateral failure of development of the chondrification center (31).

Anomalies tend to occur at the apex of a curve. A curve can be upper thoracic (33% of curves), lower thoracic (31%), thoracolumbar (20%), lumbar (11%), or lumbosacral (5%) (40). In general, thoracolumbar curves tend to have the worst prognosis and the greatest progression, followed by lower thoracic curves and then upper thoracic curves.

The ribs are formed in close association with the vertebrae, and it is, therefore, not surprising to have a combination of developmental abnormalities affecting both the ribs and the vertebrae. An extensive thoracic congenital scoliosis due to mixed vertebral anomalies associated with fused ribs may affect thoracic function and the growth of the lungs in young children and lead to a thoracic insufficiency syndrome. An imbalance in the mechanical thrust of the ribs may also adversely affect spinal growth as well as the function of trunk muscles and the pressure within the thorax.

However, since only the vertebral body and the pedicle of the vertebral arch in the malformed vertebrae can be evaluated by this classification, an evaluation of the morphology of the posterior components has not been performed. Furthermore, it is difficult to evaluate the morphology of vertebral bodies in severe kyphoscoliosis, or in cases of severe complex malformed vertebrae, using plain AP images alone.

### NATURAL HISTORY

Reports in the 1950s by Kuhns and Hormell (12) estimated that congenital scoliosis had a generally benign course: only 37% of the children in their series had curves greater than 30° at maturity. This conclusion was refuted by the independent natural history studies by Winter *et al* (44) and McMaster and Ohtsuka (26) who reviewed a combined total of 485 patients, most until skeletal maturity. They reported that the rate of deterioration and the severity of final deformity were predictable according to the type of anomaly and curve location. Upper thoracic curves tend to be less severe than thoracolumbar curves, which tend to be most severe. Curve progression occurs more rapidly during the first 5 years of life and, again, during the adolescent growth period of puberty; these two periods represent the most rapid stages of spine growth (12).

These authors found that nearly 75% of patients studied required treatment, and that 84% of patients who were untreated developed curves greater than 40° at maturity. For most anomalies, the deterioration of the curve was steady and unremitting, starting at birth, accelerating during the preadolescent growth spurt, and slowing or stopping at skeletal maturity. Children with clinical deformities in the first year of life had the worst prognosis and had severe early progression.

Curve progression is caused by unbalanced growth of one side of the spine relative to the other. Spine growth comes from the superior and inferior end plates of each vertebral body. Thus by assessing the quality of the disks surrounding the anomalous segment of the spine, growth potential may be inferred. Radiographically definable disks signify the presence of vertebral growth plates and,

when asymmetrical or more present on one side of the spine than on the other, they have the potential for asymmetrical growth in that area of the spine. Thus, fully segmented hemivertebrae with healthy, definable disks above and below have much more potential to cause a curvature compared with unsegmented hemivertebrae, which are fused to the vertebrae above and below (33). The presence of either a bar or fused ribs is a good predictor of curve progression. Either can act as a tether, and the combination tends to produce even more rapid curve progression.

With recognition of the 'at-risk' types of vertebral malformations, patients with congenital scoliosis can now be classified according to their risk of curve progression and the frequency of follow-up adjusted accordingly. Some deformities, such as a unilateral unsegmented bar, were recommended for treatment at time of recognition because of their invariable tendency for severe progression. The thoracic curves have the poorest prognosis, with the worst anomaly being a unilateral unsegmented bar combined with single or multiple convex hemivertebrae, followed by a unilateral unsegmented bar, double convex hemivertebrae, and a single convex hemivertebra, with a block vertebra having the best prognosis (20).

A unilateral unsegmented bar adjacent to a contralateral hemivertebra with an average curve progression in excess of 10 degrees per year in the thoracolumbar region is the most aggressive anomaly. It may not be radiographically visible until it ossifies when the patient is 3 to 4 years of age. A hemimetameric shift does not always present with a balanced spine, and produces occasionally a progressive deformity especially in thoracolumbar and lumbosacral junctions (20).

Block vertebrae are uncommon and tend to be the most benign anomaly producing curves of less than 20°. Complex curves consisting of multiple anomalies occur in 10% of cases (20). Both conditions should be observed until progression is documented.

Mixed deformities are unpredictable, and their severity depends on the amount of unbalanced growth potential. The combination of mixed deformities and rib anomalies can lead to a global loss of trunk and thoracic height and width, resulting in

severe volume and motion restriction leading to respiratory failure.

### ASSOCIATED SURFACE DEFORMITY

A cosmetic deformity occurs with most curves. In upper thoracic curves, elevation of the shoulder on the convexity of the curve, with tilting of the head into the concavity, may be seen. Structural congenital curves tend to have only a mild rotational component; thus, they produce only a mild rib hump. Unbalanced curves in the lower thoracic region and the lumbar region produce a pelvic obliquity with apparent shortening of the leg on the concave side of the curve. The trunk tends to list away from the apex of the curve, and this can cause difficulty in ambulation and balance.

### ASSOCIATED ANOMALIES

Another important aspect of congenital scoliosis is the recognition of the presence of concurrent abnormalities of the spinal cord, the kidneys, and the heart, which may be present in 61% of patients and in an even higher percentage in patients with mixed defects (2). There is some evidence in the literature to support the development of organ malformations at the body segment at which vertebral malformations occur. The developmental field concept described by Opitz (32) is useful in providing a framework for understanding multisystem involvement and a defect in one system should prompt evaluation of others.

Neural axis abnormalities are present in up to 35% of patients, as detected with MRI, patients with mixed or segmentation defects being at highest risk (5). These abnormalities include (but are not limited to) diastematomyelia (split cord), cord tethering, Chiari malformations, and intradural lipomas. The absence of cutaneous signs of dysraphism and the absence of neurological deficit do not rule out an intraspinal dysraphism. Diastematomyelia is found in up to 20% of patients with congenital scoliosis and should be treated by resection before proceeding with corrective surgery of the spine because of the risk of stretching the spinal cord tethered at the site of the anomaly (46). Other lesions of the cord seen in congenital scoliosis

include dermoid cysts, epidermoid cysts, teratomas, and lipomas as well as tethered spinal cord and they may all need treatment prior to spinal surgery to minimise risk of subsequent neural injury. With such a high incidence of associated intraspinal anomalies, routine MRI screening of patients with congenital scoliosis has been suggested (4). The specific indications for MR imaging for possible intraspinal anomalies in congenital scoliosis include: the presence of neurological defects such as weakness, sensory loss, bowel or bladder dysfunction; an associated skin abnormality over the spine such as a dimple, hairy patches, or nevi; complaints of back or leg pain; patients with lumbosacral kyphosis; radiographic evidence of interpedicular widening, diastematomyelia or presence of a unilateral congenital bar with a contralateral hemivertebra; and any patient who is to undergo spinal stabilisation surgery (4).

Unrecognised abnormalities of the renal system may be present in 25 to 33% of patients and include unilateral horseshoe kidney, renal agenesis, duplicated kidney/ureters and hypospadias (13). All patients with congenital scoliosis warrant a renal evaluation by either intravenous pyelogram or renal ultrasound/MRI.

Congenital heart disease is present in 10% of patients, ranging from atrial and ventricular septal defects, which are the most common abnormalities, to complex congenital heart defects, such as tetralogy of Fallot and transposition of the great vessels. A screening echocardiogram and cardiological evaluation is needed for patients undergoing surgical correction for a congenital spine deformity.

The development of restricted pulmonary function is a concern. Recent studies imply that curve magnitude directly affects pulmonary function, but the development of restrictive lung function occurs only as the curve approaches 90° (30). Severe curves in young children tend to produce the most severe pulmonary compromise. This restriction may be due more to hypoplastic lung development than to a physical constriction from the curve; alveolae continue to develop in the growing child up to 8 years of age. Vital capacity screening is recommended for patients with severe curves.

Musculoskeletal anomalies occur frequently in association with congenital spine anomalies. Disorders, such as clubfeet, Sprengel deformity, Klippel-Feil deformity, developmental dysplasia of the hip, and upper and lower limb deformities warrant a detailed evaluation and management prior to spinal surgery.

Vertebral malformations have been shown to be associated with hemifacial microsomia, Alagille, Jarcho-Levin, Klippel-Feil, Goldenhar, Joubert, and VACTERL syndromes as well as basal cell naevus, trisomy 18, and diabetic embryopathy (3).

### CLINICAL AND RADIOGRAPHIC ASSESSMENT

#### Clinical Evaluation

Clinical evaluation begins with a comprehensive history and physical examination. A prenatal history of the mother, including all health problems, previous pregnancies, and medications, is recorded. Birth history of the child should include details such as length of gestation, type of delivery (vaginal or caesarean), birth weight, and complications. Given that the presence of cognitive delay has been shown to correlate with curve progression in some patients, particular attention should be paid to whether the child has appropriately reached developmental milestones (10).

The physical examination should start with the height and the weight of the patient, given that growth plays a significant role in curve progression. The skin must be examined for abnormalities such as "café au lait" spots or axillary freckles as seen in neurofibromatosis and midline patches of hair for any evidence of spinal dysraphism. Spinal dysraphism may also manifest itself in the lower extremities, and signs would include asymmetrical calves, cavus feet, clubfeet, vertical tali, and abnormal neurological findings. The spinal examination itself focuses on any evidence of truncal or pelvic imbalance. Rib cage deformities, chest or flank asymmetry, chest excursion and anomalies need to be evaluated, as does the inspiratory and expiratory capacity of the chest wall. Limitation in chest excursion may indicate a syndromic scoliosis and

thoracic insufficiency syndrome (10). In young children, the Adams forward bending test (looking for prominence of the ribs in the thoracic spine or transverse processes in the lumbar spine) is not possible, but the test can be simulated by laying the child in a prone position over the knee of the examiner. Curve flexibility can be assessed by placing the child in a lateral position over the knee of the examiner or by suspending the infant over the arm of the examiner.

Spinal balance in both the coronal and the sagittal planes must be evaluated. Truncal imbalance, head tilt, shoulder inequality, and pelvic balance should be addressed. Photographs serve as an important aid in sequential evaluation of progress. Given the association of neural axis abnormalities and the possibility of neurological compromise in congenital spine deformities, a detailed motor, sensory and reflex examination, (including abdominal reflexes) is mandatory. Muhonen *et al* (29) described the absence of an abdominal reflex as the only objective finding seen in some patients with a Chiari malformation. When the results of reflex testing are abnormal, the reflex is usually absent on the convex side of the curve.

The vital capacity of patients with congenital scoliosis has been compared with those with idiopathic scoliosis. For any given Cobb angle the loss in vital capacity was approximately 15% greater in congenital compared with idiopathic scoliosis. This greater impairment in lung function in congenital scoliosis has been proposed to be due to the associated rib deformity or to an underlying lung anomaly (30). Vital capacity screening has been recommended for patients with severe curves. A full spirometry work-up is recommended if surgery is planned for those with a vital capacity < 60% of normal (30).

#### RADIOGRAPHIC ASSESSMENT

Plain radiographs remain standard for diagnosis of congenital anomalies and measuring curve magnitude, progression and perhaps growth potential of the vertebral anomaly. Conventional radiography often is difficult to interpret because of the patient's small size, the complex nature of the deformity,



*Fig. 4.* — Preoperative three-dimensional reconstructed computed tomography scan defines the anatomy of the curve and may aid in the positioning of the implants.

superimposed structures obscuring visualisation of the anomaly, and spine deformity in a plane other than that of the radiograph.

Preoperative CT scan helps define the anatomy and avoid any unexpected intraoperative posterior element deficiencies (fig 4). The combination of 3-D images and 2-D coronal and sagittal multiplanar reformatted CT images curved to match the contour of the spine can be extremely beneficial in the assessment of hemivertebrae for which it is difficult to determine the degree of segmentation or the presence of a contralateral unsegmented bar by plain radiography or 2-D CT imaging. We obtain CT with 3-dimensional reconstructions for preoperative assessment and evaluation of complex deformities, but not for routine observation or serial documentation.

Computed tomography scan helps in evaluation of chest wall deformity and lung volume in congenital deformities with chest wall anomalies, chest deformity, or thoracic insufficiency. The use of 3-D CT data to define lung volumes in patients who were too young for pulmonary function tests has been described and subsequently used to measure improvement in lung function following expansion thoracoplasty (18).

Magnetic resonance imaging (MRI) has had a revolutionary impact on imaging the neural axis and its role has been widely studied and documented in patients with congenital scoliosis and spinal dysraphism (4). The prevalence rate of spinal dysraphism detected using MRI approaches 30% in patients with congenital spine deformities. Imaging of the brainstem to the bottom of the sacrum is required to exclude associated conditions of the spine, the cranio-cervical junction and the viscera. In our institution, we perform coronal T1, sagittal T1 and T2 weighted scans through the whole spine with axial gradient echo images through the cervical spine and T2 weighted images through the apex of the curve and through the lumbar spine. The coronal T1 weighted images are useful for demonstrating if the cord is single and for assessing the shape of the scoliosis.

## MANAGEMENT

The skill in managing a patient with a congenital spine deformity lies not only in the ability to perform major complex salvage surgery in patients presenting at a late stage with a severe rigid deformity, but in recognising those curves with a bad prognosis at an early stage to prevent curve progression and possible neurological complications. Meticulous management planning requires an astute knowledge of the natural history of all types of congenital spine deformity and the methods of treatment that are available. Once high-risk anomalies such as unilateral unsegmented bars with contralateral hemivertebrae are recognised, treatment is initiated regardless of age to prevent deformity.

## NON-SURGICAL TREATMENT

Congenital vertebral anomalies can be recognised fortuitously on radiographs of infants before clinical deformity develops. Both these and patients diagnosed later in life should be followed carefully at 4 to 6 month intervals until the end of growth. The goals of observation are to recognise the presence of vertebral anomalies, which require immediate treatment, and to detect the progression of curves.

## Bracing

Bracing plays a very limited role in the management of congenital scoliosis, especially as a primary form of treatment because these curves are typically inflexible. Less than 10% of congenital curves can be treated by bracing. Bracing is sometimes used to prevent progression of secondary curves that develop above or below the congenital curve and are causing balance problems. The Milwaukee brace is the brace of choice for upper curves. A TLSO (thoracic/lumbar/sacral orthosis) brace is adequate for lumbar curves. The optimum duration of daily brace wear is still under investigation. As long as the curve remains controlled, treatment should continue until skeletal maturity. The brace may not arrest or correct development of the curve, but it may slow progression and maintain flexibility, allowing surgery at a later age. Bracing serves no function in short, sharp, rigid curves.

## SURGICAL TREATMENT

### Principles of surgery

As a guiding principle, it has been widely recognised in studies that it is easier to prevent a deformity than allow it to progress and attempt correction at a later stage (17). 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 fusion segment as possible, preserving as much normal spinal growth as possible. The risk of neurological injury with surgical treatment of patients with congenital spinal deformities is greater than in patients with idiopathic spinal deformity (34). The occurrence of a perioperative neurological deficit may be minimised in multiple ways by :

- (1) A routine use of MRI evaluation of the spinal cord.
- (2) Earlier treatment of a spinal cord anomaly.
- (3) Preventing lengthening the spinal cord intraoperatively by avoiding distraction and using shortening procedures.
- (4) Carefully monitored use of controlled hypotension to minimise blood loss to prevent the occurrence of cord ischaemia, especially during any corrective manoeuvres.
- (5) Motor- and sensory-evoked potential monitoring, supplemented by a wake-up test in cases with intraoperative changes in neurological monitoring that do not return to baseline.
- (6) The postoperative monitoring of a patient's neurological status, given that paraplegia after deformity surgery may present in a delayed fashion, especially in the first 72 hours (28).
- (7) Early and aggressive treatment of deformities before they become severe.

The procedures are broadly divided into (a) those preventing further deformity and (b) those that correct the present deformity ; with the latter type further subdivided into techniques that correct the curve gradually and those that correct the curve acutely.

### Prevention of Future Deformity

#### In Situ Fusion

*In situ* fusion is a safe technique and a good choice for many progressive curves with minimal deformity involving a relatively short section of the spine.

It is ideally done in patients with unilateral failure of segmentation such as a unilateral unsegmented bar and should be accomplished very early, before a significant curve develops, which would then make a more extensive correction necessary.

A patient with no trunk imbalance with a curve less than 40 degrees not requiring substantial correction is best fused *in situ* without instrumentation with some modest correction by postoperative casting (17). One of the major controversies surrounding *in situ* fusion is whether a combined anterior and posterior fusion is required (24). There is less potential for anterior growth in a spine with congenital scoliosis because the growth plates may not be properly formed. In an *in situ* fusion for congenital scoliosis, a shorter segment is fused and there is less rotation in the segment, which makes a

true crankshaft phenomenon (continued anterior growth in presence of posterior fusion) unlikely.

McMaster (25) and Winter and Moe (43) recommended an early prophylactic *in situ* fusion to prevent the unbalanced growth of the spine in a unilateral unsegmented bar, with or without contralateral hemivertebrae with best results reported before the age of two years. A convex growth arrest procedure or simply dividing the unsegmented bar will not correct this type of deformity because there is no growth potential in the unsegmented bar on the concavity of the curve. This usually necessitates a combined anterior and posterior fusion to control the severe spinal growth imbalance in these patients. The argument that an early spine fusion will stunt the growth of the spine in these young patients is of no relevance because the unsegmented bar is not contributing to vertical height and only making the spine more crooked.

The decision regarding which curves may be fused *in situ* can be complex. The location of the curve and likelihood of progression and the impact of the curve on the balance and cosmetic appearance of the child must be taken into account. Deformities at the lumbosacral and cervicothoracic junction are more likely to lead to cosmetically disfiguring deformities and therefore should be fused *in situ* very early or alternate treatments considered.

#### Convex hemiepiphysiodesis

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 that have no potential for concave growth. The epiphysiodesis is done by removing the convex lateral half of disks adjacent to the hemivertebra, with no exposure of the spine on the concave side. Winter *et al* (42) suggest that this procedure should be reserved for patients younger than 5 years of age, with a progressive curve of < 70 degrees involving 5 segments or less, and presenting with a pure scoliosis not involving the cervical spine with no major kyphosis or lordosis. In their series, 38% of patients had an average correction of 10 degrees, 54% had a stabilisation effect without significant correction, and the only compli-

cation was curve progression in one patient. More predictable alternatives to convex hemiepiphysiodesis for a short segment deformity include hemivertebra excision or wedge resection, and growth-oriented procedures, such as VEPTR (Vertical expandable prosthetic titanium rib) or growing rods, when a longer segment of spine is involved.

#### 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 epiphysiodesis would not result in a 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 (19). To close the gap after resection of the hemivertebra, compression instrumentation is necessary and allows for strong fixation into adjacent vertebrae. Ruf and Harms (35) previously argued that hemivertebra resection via a posterior-only approach with transpedicular instrumentation was the ideal procedure for correction in young children. They opined that this methodology was not only less invasive but also prevented the development of secondary curves and severe local deformities.

Suk *et al* (39) also support the use of a posterior-only approach, arguing that the combined approach requires prolonged operative time, and that when used in the treatment of lumbosacral deformity, it poses a risk to the anterior vascular and visceral structures. Consequently, based on their retrospective review of treatment of fixed lumbosacral deformity with posterior vertebral-column resection, they maintained that this technique offers the advantages of single-stage surgery, the ability to address the deformity at the apex, and of controlled shortening across the resection gap.

Bollini *et al* (5) on the other hand advocate a combined anterior and posterior fusion because of greater correction capabilities, including correction in the sagittal plane secondary to disc excision. This approach gives better visualisation of the

anatomy including the spinal column and nerve roots, as well as affording the surgeon the opportunity to apply a corrective force to the spine from an anterior position while simultaneously applying compressive instrumentation. It also decreases the likelihood of pseudoarthrosis and prevents the development of crankshaft phenomenon by the removal of the growth plates (6). The procedure is best performed in the thoracolumbar junction or lower because the spinal cord in the thoracic spine is not as amenable to manipulation, but this view has been recently refuted (11).

### **Correction of the present deformity**

*Gradual correction* is obtained through the use of a hemiepiphysiodesis and/or hemiarthrodesis of the spine. Immediate or acute correction may be accomplished through several different techniques : excision or decancellation of the hemivertebra and definitive or staged instrumentation using a growing rod. For patients with thoracic insufficiency syndrome, expansion thoracoplasty and stabilisation via the VEPTR addresses this issue by rib distraction on the concave side of the curve. For fixed deformities, it may be necessary to perform an osteotomy to balance the spine and correct the cosmetic deformity.

#### *Gradual correction*

### **Hemiepiphysiodesis and Hemiarthrodesis**

These techniques are used for failure of formation, relying on the future growth of the spine on the concave side. In segmentation failure, there is no potential for growth to occur. The procedure is unlikely to correct curves with low concave growth potential such as those due to a convex hemivertebra opposed by a concave unsegmented bar (41). Dubousset (14) has contributed much to growth arrest procedures of the spine by pointing out the relationship between sagittal and coronal growth disturbance of the spine with congenital deformity and how mechanical instability must be considered in the treatment. The goal is to arrest the growth on the convex side of the curve ; the concave side of the spine is not exposed surgically, as doing so could lead to spontaneous fusion. Much of the correction associated with hemiepiphysiodesis is

achieved acutely at the time of the initial procedure, thus postoperatively, a corrective cast is used to encourage fusion of the spine in a somewhat corrected position, which will therefore improve the starting point and permit the ultimate correction to rely less on the growth of the spine. The total correction obtained by performing a convex hemiepiphysiodesis varies because the younger the child at the time of the operation, the more potential that exists for correction over time.

#### *Growing Rods*

### **Single and Dual Growing Rods**

The growth of the spine is greatest during the first 5 years of life as sitting height reaches two thirds of the adult level by age 5, while thoracic volume reaches 30% of the adult size by five years of age. Congenital scoliosis is associated with short stature and diminished trunk height. Long fusions performed on younger children may have a further deleterious effect on trunk height and thoracic volume, leading to thoracic insufficiency. In the absence of congenital rib fusions, early progressive deformities may best be managed using a growing rod technique, pioneered by Paul Harrington (43) in the 1960's. If the child is still very young, the primary congenital curve can be treated with fusion *in situ*, hemiepiphysiodesis 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.

The goals of treatment have been to achieve correction of the spinal deformity without extensive fusion, maintain correction during the subsequent growth period, allow spinal growth and lung development, and avoid or eliminate the need for definitive fusion of the spine at an early age.

Akbarnia and McCarthy (1) reported their series of a dual growing rod construct due to problems associated with a single-rod implant, including hook dislodgement and rod breakage. This is performed by subperiosteal dissection of the anchor sites proximally and distally and by placement of claw constructs. Rods are then placed subcutaneously on each side and joined with tandem con-

nectors placed at the thoracolumbar junction, where the lengthening may occur. An apical fusion involving the congenital anomaly may be needed; this will obtain control of the apex of the curve and, if a hemiepiphysiodesis and/or hemiarthrodesis has been performed, will possibly permit improvement of the apical curvature with growth.

However, apical fusion is a must if the apical curvature is due to failure of segmentation. Their patients showed an average Cobb angle improvement from 82 to 38 degrees at follow-up and an average growth of the T1-S1 segment of 1.2 cm per year.

#### *Acute Correction Techniques*

##### Correction and Fusion with Instrumentation

The partial or complete correction of a deformity depends on the congenital anomaly itself and the degree of deformity. The choice of fusion levels is difficult when there are multiple noncontiguous anomalies but the aim should be to achieve a balanced spine with the safest possible correction. The whole curve needs to be fused, but there may be abnormal segments cephalad or caudad to the curve that will not be included or two curves may need to be included in the instrumentation. High thoracic and cervical curves need special precaution as overcorrection of a lower thoracic or thoracolumbar curve may lead to shoulder imbalance or neck obliquity. Congenital anomalies associated with relatively normal segmentation, flexibility (as revealed by radiographs), and less severe truncal deformity may be managed by means of a standard posterior arthrodesis and instrumentation. Those cases with well-defined disk spaces seen on imaging imply significant remaining growth; these patients may be at risk of a crankshaft phenomenon and should have in addition anterior surgery in the form of either an open or a thoracoscopic anterior release and fusion. The amount of correction obtained by instrumentation in congenital scoliosis is usually much less than that obtained in idiopathic scoliosis and one should not attempt to gain dramatic correction of large, stiff congenital curves. Complications of treatment have declined as experience has increased with the most common problem being progression of the curve. Occasionally,

intraspinal abnormalities must be dealt with prior to the instrumentation procedure. Computed tomography with three-dimensional reconstruction is very helpful for the assessment of local anatomy and as an aid in pre-operative planning.

The safe use of traction in congenital spine deformities, particularly in cases involving preexisting neurological deficits, was questioned in past reports by MacEwen *et al* (22) due to the risk of traction-induced paraplegia in congenital deformities. Recently, the use of traction has been popularised for severe deformities, including congenital deformities (34). Rinella *et al* (34) reported the use of Halo gravity traction which can be applied in bed or while in a wheelchair or walker device allowing for gradual curve correction before or following an associated anterior release.

##### Hemivertebra excision

Hemivertebra excision may be an ideal procedure for long standing curves with significant deformity or truncal imbalance. The ideal indication is a hemivertebra at the lumbosacral junction, as this particular deformity often leads to major imbalance or very serious compensatory curves.

It is possible to combine this procedure with a convex hemiepiphysiodesis and/or hemiarthrodesis, with use of a nonfusion rod or a definitive instrumentation of a longer curve. The excision of a hemivertebra may be performed by using combined anterior and posterior procedures. Posterior-only hemivertebra excision in growing children has recently been reported with successful results. Ruf and Harms (35) reported their results on hemivertebra excision using a posterior-only approach and segmental transpedicular instrumentation. They reported excellent results in patients younger than 6 years, with an average Cobb measurement of 45 degrees. At 3.5 years follow-up, the Cobb measurement had been maintained at 14 degrees, with no patient having a neurological complication.

##### Reconstructive osteotomy

Pelvic obliquity, severe truncal decompensation, progressive deformity, and developing neurological deficit are indications for reconstructive osteotomies. They are considered for fixed curves with



**Fig. 5.** — Postoperative radiograph of a four-year-old boy with a volume-depletion deformity resulting from fused ribs and congenital scoliosis. The VEPTR stabilisation expansion thoracoplasty lengthened and widened the constricted hemithorax, corrected the scoliosis indirectly without growth inhibition, restored truncal balance, and corrected pelvic obliquity.

unacceptable cosmesis and cannot be successfully managed with procedures described so far. An osteotomy may be part of a combined approach that involves resection of the hemivertebra and instrumentation and fusion of a more extended curve. A cosmetically displeasing but well balanced curve is probably best treated with stabilisation alone.

#### EXPANSION THORACOPLASTY AND VEPTR

Congenital spine deformities with rib fusions may be associated with thoracic insufficiency, a term coined by Robert Campbell *et al* (8) to explain

poor thoracic and lung parenchymal development with inability to support normal lung function and growth. The tethering effect of congenital rib fusions add to the scoliotic effect of the spine to produce a concave, constricted hemithorax, which is diminished in height and function.

The growth of the lung, the respiratory tree, and the alveoli cell multiplication are greatest in the first 8 years of life. Fifty percent of the thoracic volume is attained at 10 years of age ; thus, early fusions may have an even more profound effect on thoracic development than on spine height. Having borderline pulmonary capacity at the end of growth can be a serious risk factor for developing respiratory insufficiency in late adulthood because approximately 400 cc's of vital capacity is lost in the normal aging process and even further losses may occur from obstructive respiratory disease or infection. The possible effect of early spinal fusion on thoracic development may compound the preexisting thoracic insufficiency associated with congenital scoliosis and fused ribs (8).

In children with congenital scoliosis and severe curve progression early surgical curve stabilisation was based on the belief that 'a short straight spine is always preferable to a long crooked one' (21). Therefore, early posterior or combined anterior and posterior spinal fusion, convex anterior epiphyseodesis or arthrodesis has been performed in the past (23). The total growth inhibition effect on the thoracic spine due to these procedures was thought to be negligible because of the assumption of diminished growth potential of the concave side (21). A recent study (7) challenged this assumption by showing an increase in length of the unilateral unsegmented bars and equal increases in length of the concave and convex sides of the thoracic spine in children with congenital scoliosis, unilateral fused ribs and unsegmented bars treated by VEPTR. The surgical concept of the expansion thoracoplasty and stabilisation with the VEPTR implant is based on the expansion of the thorax by rib distraction on the concave side of the curve achieving indirect correction of the curve (fig 5). Spinal fusion may be necessary once skeletal maturity is reached and thoracic spinal growth is no longer an issue.

## DIRECTIONS FOR FUTURE WORK

Congenital scoliosis remains an interesting and challenging diagnostic entity. Developments in gene research continue to be promising and may potentially lead to early detection of congenital vertebral malformations. It is likely that different approaches will enable an elucidation of the genetic and environmental basis of CS. Since CS is due to localised alterations in vertebral body development as opposed to a more widespread distribution of vertebral malformations, a model needs to be developed that incorporates position identity and failure of segmentation. The association of renal, cardiac, skeletal and spinal cord malformations with CS may reflect the involvement of different genes that are associated with developmental pathways in several organs. Epidemiologic studies can provide assistance in determining whether various environmental factors contribute to CS.

John Cobb (9) wrote in 1958 that "In the future study of scoliosis it will be necessary to keep our eye on the patient and not on the curve." Important issues to be addressed in the future about thoracic insufficiency syndrome and VEPTR include characterisation of the natural history of the thoracic insufficiency syndrome by anatomic classification, correlating progressive three-dimensional thoracic deformity with the onset of respiratory insufficiency and whether the procedure can halt or reverse progressive three-dimensional thoracic deformity after the onset of respiratory insufficiency. Furthermore, continued advances in surgical technique and instrumentation will provide surgeons with an array of safe and efficacious modalities for these patients with complex conditions.

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