Review Paper:
Congenital Scoliosis: A Current Concepts Review

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ABSTRACT

Context: Congenital scoliosis is a difficult condition for orthopedic surgeons. There are some influencing factors to choose the best treatment option for scoliosis.

Objectives: Patients with congenital scoliosis may encounter different anomalies. There exist various surgical techniques with different indications.

Methods: Electronic databases, such as Google Scholar, PubMed, and Scopus were searched for congenital scoliosis. Articles published from 1928 to 2020 were searched. A narrative review was conducted by focusing on treatment options.

Results: Different methods are presented in the literature that consists of operative and nonoperative approaches. Nonoperative treatment methods are seldom a final choice. They are used to postpone the final surgery. There are different methods of surgeries in the literature; the best treatment strategy concerns the patients' condition and the surgeon's preference.

Conclusions: The critical issue in the management of congenital scoliosis is to diagnose these patients' curves before severe progression, i.e. mandatory to achieve desirable results. Usually, a course of nonoperative treatment can be started, but only to postpone the final surgery. The preferred surgical treatment depends on the type of congenital scoliosis and the age of the patient. The treatment of congenital scoliosis should be a multidisciplinary approach due to concomitant morbidity in these patients.

1. Context

Viewed from the coronal plane, a healthy spine is straight. Scoliosis term derives from the Greek word ''skolios'' (crooked, curved), i.e. first established by Galen (130-201 AD) (1). In orthopedics, scoliosis is a structural deformity of the spine defined by a lateral curvature (2). Scoliosis is accompanied by a rotation, i.e. maximally at the apex of the curve. This deformity is diagnosed with a Cobb angle in patients when it occurs or becomes relevant after skeletal maturity (3, 4). The deviation of <10

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degrees is called spinal asymmetry, whereas the deviation of ≥10 degrees is called scoliosis (5). Scoliosis prevalence has a wide range from 8.3% to 68% of the population (6-8), with a higher prevalence occurring among older patients (9). Patients with scoliosis are classified broadly as congenital, idiopathic, neuromuscular, and syndrome-related (10). Congenital Scoliosis (CS) occurs due to abnormal growth and the development of the vertebral column during embryogenesis (11, 12), i.e. present at birth. However, because of morphological changes during growth, the deformity may not be apparent until later in childhood (13). In children aged <3 years, it is difficult to differentiate between CS and idiopathic infantile scoliosis (14). The worldwide prevalence of CS is equal to 0.5-1 per 1000 live births (15, 16).

Etiology

Understanding the cause of CS is essential for elucidating its pathogenesis. The spine is formed in a process, called somitogenesis. Somitogenesis occurs between the third and fourth weeks of gestation. In this formation, the segments of mesodermal tissue, called somites, are formed in pairs surrounding the eventual spinal cord (16). Somites that are sequentially added to the growing axis establish the characteristic periodic pattern of the spine. The early segmentation stages of the vertebrate of the embryo are displayed by the somitic organization; they also underlie much of the segmental organization of the body, including muscles, nerves, and blood vessels. In the vertebral column, the major component of the paraxial mesoderm is somites that form bilaterally along the nerve cord as a result of primitive/blastopore streak and tailbud regression during body axis formation (17). CS results from a disruption in somitogenesis as evident from animal models. Additionally, various theories have been proposed to explain congenital vertebral anomalies; e.g., failure to ossify, the osseous metaplasia of annulus fibrosus, or persistent notochord (18).

Both environmental and genetic factors influencing spinal development in lower vertebrates might affect the abnormalities associated with human CS (19). Numerous genes are known to play crucial roles in the development of a normal healthy spine. Genes in the "notch" family, such as Pax1 (20) LFNG, MESP2, HES7, and Delta-Like 3 (DLL3) (21-25) have been reported to regulate the development of vertebral precursors. Defects in human notch genes have been associated with normal vertebral development (26). The TBX6-mediated genes that impact somitogenesis are responsible for CS (27). This condition can be related to genitourinary, cardiac, and spinal system abnormalities (28) as well as some syndromes, like Chiari malformation (29). This is suggested as the 1 in 100 risks of a first-degree relative having a single vertebral malformation and a risk of 1 in 10 for multiple vertebral anomalies in either siblings or children of a patient (16).

Classifications

Disease classification systems are created to share a common theme of disease pathologic conditions and provide treatment options for disease states. An ideal disease classification provides a perspective view of a disease state that identifies different severities of the disease state; facilitates communication between health care providers and researchers to assure accuracy and reproducibility in describing the disease state; allows the comparison of different treatment methods, and consequently, allows the creation of accurate treatment recommendation guidelines (30). An ideal CS classification has great significance regarding these aspects. The commonly encountered type is illustrated in Figure 1.

Schwab's classification is focused on the relationship between clinical evaluation and radiological findings; subsequently, it categorizes the apex of the curve, lumbar lordosis, and vertebral body subluxation based on radiological findings (9, 31). The classification of CS based on the 2D images has limitations in clinical usage at present when 3D CT images can be obtained. In 2009, congenital spinal deformity could be mainly classified into 4 types of congenital vertebral abnormalities, as follows: solitary simple, multiple simple, complex, and segmentation failure (32). In 2014, 56 cases of CS were classified in the literature. Besides, the necessity to classify the other cases led to a new classification, consisting of two groups based on the spine dominant deviation in coronal and transverse view; scoliosis due to longitudinal and rotational imbalance (33).

2. Diagnosis

Physical examination

The physical examination of patients with CS should include head to toe, especially the facial features. Neurologic malformations (35% of the patients), congenital heart malformations (25% of the patients), urologic anomalies (20% of the cases), and musculoskeletal anomalies associated with congenital malformations, motor, and sensitive disorders should be checked (34). Tall patients with long fingers and an increased arm span to height ratio should be assessed for other signs

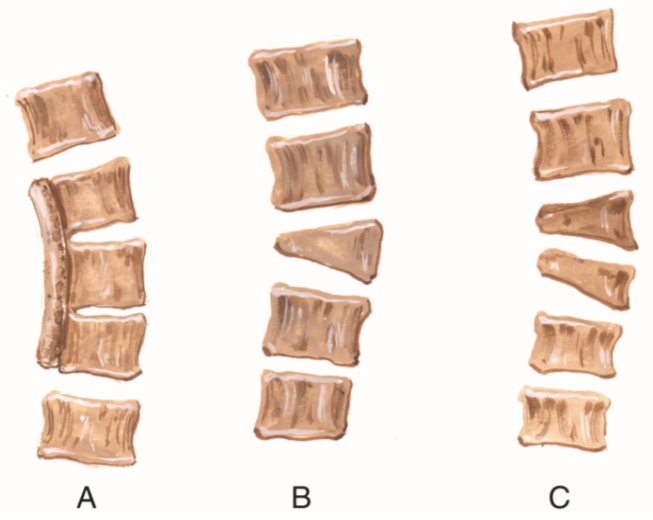

Figure 1. Commonly encountered vertebral malformations associated with CS
A: Unsegmented bar; B: Single hemivertebrae; and C: Multiple hemivertebrae [19]

of Marfan syndrome or homocystinuria. The clinical accompaniment of joint and skin hyperlaxity along with scoliosis may require further workup for a connective tissue condition, like Ehlers-Danlos syndrome. Neurological disorders, like Charcot-Marie-Tooth disease or a spinal cord abnormality, like a tumor, may be associated with high-arched or cavus feet (10). The physical examination of patients with CS should also include obvious deformities with truncal imbalance, the abnormalities of the scapula, like Sprengel deformity and pelvic tilt.

The body tends to list away from the main body axis; thus, it may lead to difficulty in ambulation and balance. Moreover, rib cage abnormalities, the asymmetry of the flank or chest, chest excursion, and anomalies need to be evaluated (16). In general, scoliosis is not a simple deformity in the coronal plane but is also a rotational deformity. Scoliotic curves without rotation may be caused by bony tumors, intraspinal pathology, and nerve root irritation (10).

Imaging

The best time to observe the details of the vertebral anomalies in radiography is before the development of significant deformity (14). Plain radiographs of standing posterior-anterior aspects represent the usual investigation to evaluate the spontaneous evolution during preoperative bracing and after surgery. This evaluation aims to measure the curves' angle by a different method, such as Cobb's or imaging approaches; it also helps to spatially appreciate the deformity and multiple examinations allow the surgeon to identify curve quantification to compare the results of different treatment methods (34). Diagnostic imaging by X-ray is the gold standard of imaging studies in the diagnosis of congenital bony malformation and the measurement of curve magnitude (16).

Three-dimensional reconstruction preparation with sagittal multiplanar reformatting by CT can be very useful in hemivertebrae assessment to detect posterior vertebral anomaly (16). CT is also useful to determine/assess fused segments and vertebral instability; it could also improve the prognosis of the scoliotic curvature progression. CT is often applied for preoperative surgical planning in clinical practice (35). According to the CT radiation doses for the patient (36), CT is rarely employed for the clinical follow-up of CS (37).

Magnetic Resonance Imaging (MRI) is unnecessary for most patients with scoliosis; although this is somewhat controversial. In contrast, with idiopathic scoliosis, all patients with CS should preoperatively undergo MRI scans. The imaging of the brainstem to the sacrum by MRI is necessary to exclude the associated conditions of the spine, the craniovertebral junction, and the viscera. A T2w image through the apex of the curve and a T1w image to determine any cord abnormalities are required (38). Finally, CT scans and MRI methods are usually performed in a supine position, which may underestimate spinal curvature and possible dislocations (39).

Management

The treatment goals are to stop the progression of the spine deformity while ensuring the continued growth

and development of the spine, thoracic cavity, and lungs (13). However, for these patients, no treatment is optimal during spinal growth (40). In general, non-surgical treatment methods are seldom effective but sometimes are used for the selected cases. Corrective instrumentation in combination with arthrodesis is the best method for achieving the best results (Table 1) (41).

Non-surgical treatment

The limitations of non-surgical methods are well-known concerning CS; however, in most cases, conservative treatment aims to avoid spinal surgery. Treating patients with these methods provide a limited effect on the progression of scoliosis; however, it is often needed to help the patient maintain a satisfactory seated position in cases of significant hypotonia and maintain effective chest expansion (42). Casting and bracing are non-surgical approaches for the treatment of patients with CS. Patients with nonprogressive curves should be regularly observed in periods of rapid growth (0-5 & 10-15 years of age).

In this observation, the patient should present high-quality radiographs twice a year. In patients with multiple anomalies due to unpredictable prognosis, observation is recommended. Patients with bracing should undergo radiography every 6 months. Some curves progress very slowly; therefore, it is crucial to compare the current radiography with all previous curves, even with the first radiography to detect any progression (43).

Casting

A crucial nonsurgical method to delay surgery in scoliosis patients is casting. An increase in implementing casting is observed in the management of scoliosis with the realization of the high complication rate associated with surgical techniques. These casts have windows and should be changed every two months until the best possible correction is obtained (42). Casting can delay surgical treatment for up to 2 years (44). Fletcher et al. claimed that casting can postpone the surgery for up to 64 months (45). They concluded that serial casting is an appropriate alternative treatment in moderate to severe early-onset scoliosis (45).

Bracing

Another conservative method of delaying a curve is bracing. This is the oldest treatment for spine scoliosis deformity. This is a complementary method to serial casting in patients who cannot tolerate casting. This approach can also be used as a step down from casting after a satisfactory improvement in the curve with casting (13). Brace application fails to control progression in hemivertebra and unsegmented bar and should not be attempted. There are some different braces (Garches brace, Milwaukee brace, Boston brace, & the Charleston bending brace) available with varying published results (10). Among them, the Garches brace is among the preferred treatment options for very young patients (46). Overall, few patients with CS benefit from bracing.

Surgical treatment

In general, 75% of congenital curves are progressive; therefore, surgery is the fundamental treatment. The goal of scoliosis surgery is to halt progression or to slow the progression of the curve and correct the deformity in the coronal and sagittal planes. The effect of curve surgery on the normal growth potential of the spine is critical (2). The fusion of the spine for young children should be postponed until the age of 8 to 10 years (14). In older children, even with small curves, i.e. <40°, if the progression has been confirmed, surgical intervention is mandatory. In general, surgical methods can be divided into several surgical procedures, as follows:

In situ fusion

In situ fusion is among the safest and reliable surgical treatment approaches for congenital spinal deformities. This operating system does not correct the deformity; however, it is effective in controlling the progression of the curve. Patients with unilateral failure of segmentation, such as a unilateral unsegmented bar with contralateral hemivertebrae (HV) are ideal candidates for in situ fusion (47, 48). Before any significant curve develops, in situ fusion should be accomplished in children. Deformity due to a congenital failure of formation, i.e. <50° in a child with significant growth potential is another ideal indication of this procedure (49).

Growing rods

The operating systems should not restrict the growth of the trunk and thoracic cavity. According to this, numerous techniques have been developed in which the curved spine is either not fused, or is fused only at the level of anchor placements instead of being fused throughout the curve, like the growing rod technique (50). Akbarnia (2005) described a dual growing rod technique for early-onset scoliosis and has used this technique for treating children with CS (51). A new type of these rods can be lengthened in a non-invasive man-

ner through a magnetic mechanism (52). Currently, the Magec system (53) and Phenix system (52) are used. The results of the Magec system are more promising than the Phenix system, i.e. plagued by the lock-up of its internal mechanism. Although multiple complications of rods were indicated in studies, it linearly increases with the number of procedures performed.

Expansion thoracoplasty and vertical expandable prosthetic titanium rib

Rib-based distraction techniques are to some extent similar to usual spine-based distractions; however, the proximal anchors are attached to ribs. The curve is controlled by serial distraction procedures approximately every 6 months, which grows the spine through the unfused segment (13). The concept of Thoracic Insufficiency Syndrome (TIS) was established as an inability in the thorax role in normal respiration or lung growth (54, 55). Patients with a curve >100° present a significant respiratory failure (56). According to these problems, the Vertical Expandable Prosthetic Titanium Rib (VEPTR) was developed to help lung volume, thoracic height, and lung function and correct the deformities of the thorax and spine in patients with TIS (50, 57). VEPTR's ability to improve respiratory function remains questioned (42).

Convex Growth Arrest (CGA)

CGA or the epiphysiodesis of the convexity procedure, a well-accepted technique for surgical treatment of CS, is a simpler procedure with successful results (58). This procedure is ideal for arresting curve growth in young patients with an isolated hemivertebrae and no excessive kyphosis (59-61) with a progressive curve of <70° involving ≤5 and presenting with pure scoliosis not involving the cervical spine (62). CGA is allowing the child either to reach skeletal maturity without needing further treatment or to achieve an adequate torso height to complete the treatment with a classical vertebral arthrodesis (63).

Hemivertebra excision

The first Hemivertebra (HV) excision was described by Royle in 1928 (64). Children younger than 5 years of age with a fully segmented HV at the junctional regions of the spine with spinal imbalance or curve progression are ideal cases for this surgical method (16). Usually, this technique is reserved for a patient with a fixed lateral translation of the thorax which cannot be corrected by other means and pelvic obliquity (43). Anteroposterior (AP) HV resection presents reliable and long-term safety outcomes for CS concerning clinical and radiographic findings. Posterolateral (PL) excision has been performed with rather high implant failure and revision rates. PL technique is technically a more demanding and slightly faster method for HV resection. PL technique has nearly as remarkable a correction rate as the AP technique, but more minor complications. Hemivertebrectomy seems to provide a controllable untethering effect in patients with symptomatic tethered cord (65). The excision of the hemivertebra at the level lumbosacral area can improve the trunk imbalance (43). The safest level for hemivertebra resection is below the conus medullaris in lumbosacral, L3, and L4 levels. In the thoracic spine, due to the narrow canal and the least blood supply in the spine, the risk is high, therefore neuromonitoring

Table 1. Different treatment options for congenital scoliosis

Congenital Scoliosis Treatment Options	
Surgical Approaches	In situ fusion
	Growing rod
	Expansion thoracoplasty and vertical expandable prosthetic titanium rib
	Convex Growth Arrest (CGA)
	Hemivertebra excision
	Reconstructive osteotomy
	Vertebral Column Resection (VCR)
Nonsurgical Approaches	Casting
	Bracing

is mandatory. While HV resection for CS had a higher complication rate than either hemiepiphysiodesis in situ fusion or instrumented fusion without resection, posterior HV resection in younger patients resulted in better percent correction than the other two techniques (66). The other site for hemivertebra resection is cervicodorsal hemivertebra resection.

Reconstructive osteotomy

Common operation systems for CS associated with tethered cord have the complications of untethering; however, the spinal osteotomy technique holds the potential to simultaneously correct scoliosis and decrease spinal cord tension without an extra untethering procedure (67). This technique at the thoracic apical vertebrae level, not only corrects the spine deformity but also simultaneously releases the tension of the tethered cord, resulting in improved neurologic function (68). Osteotomy planning is facilitated by preoperative 3D CT scans and rapid prototyping (69). This osteotomy planning technique is challenging cases and should be conducted by experienced surgeons.

Vertebral Column Resection (VCR)

VCR was performed in two ways staged anterior-posterior procedure and posterior resection (16). Total VCR combined with anterior mesh cage support can effectively correct curves in severe CS and can avoid the injury of the spine cord by spinal crispation. However, intraoperative position and neurologic complications should be considered (70). VCR has a high capability of congenital curves correcting on coronal and sagittal plane relied on the removal of deformity origin. Additionally, selecting appropriate strategies is important to deformity resection and segmental fixation according to different ages and deformity situations of the patient (71).

Results CS involves a broad range of spine deformities with or without a clearly defined etiology for the spine deformity. Advanced classification can help to categories CS for easier scientific communication, prognosticate the disease, and guide the treatment strategy. Early recognition and diagnosis before severe deformity develop is critical for effective treatment. However, there are major advances in the diagnosis of CS. Although the treatment of CS is primarily conservative with bracing and casting, effective operation systems are available. However, successful bracing and casting treatment prevent the radiographic progression of CS and avoids the need for spine surgery. The conventional surgical technique for the patient with a moderate degree of deformity can be successfully managed by fusion with instrumentation. Growing rods may be used in younger children involving long, normally segmented areas of the spine. VEPTR may be the best surgical technique for patients with significant growth remaining and associated chest wall anomalies. Besides, advanced deformities may require traction with reconstructive osteotomy or vertebral column resection. Sometimes, a reconstructive osteotomy is required to obtain a balanced spine.

3. Conclusion

A multidisciplinary treatment strategy is helpful to assure the optimization of medical conditions before and after surgery. Surgeon's preference is an essential factor to choose the best treatment method.

Ethical Considerations

Compliance with ethical guidelines

Patients with congenital scoliosis may encounter different anomalies. There exist various surgical techniques with different indications.

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All authors equally contributed to preparing this article.

Conflicts of interest

The authors declared no conflict of interest.

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