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Congenital Scoliosis (Mini-review)

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Abstract: Congenital scoliosis is a lateral deformity of the spine with a disturbance of the sagittal profile caused by malformations of vertebra and ribs. Typically, early surgical intervention is the suggested treatment (before three-years-old) for young patients with congenital scoliosis. While a previous study was conducted in 2011 to investigate long-term studies supporting the necessity for this recommendation and no evidence was found, this current review, is an updated search for evidence published from 2011 through March 2015. This also failed to find any prospective or randomized controlled studies to support the hypothesis that spinal fusion surgery in patients with congenital scoliosis should be considered as evidence-based treatment. Contradictory results exist on the safety of hemivertebra resection and segmental fusion using pedicle screw fixation. When using the VEPTR (vertical expandable prosthetic titanium rib) device, studies show a high rate of complications exist.

It is difficult to predict the final outcome for patients with congenital scoliosis. However, it is possible that many patients with congenital scoliosis may be able to avoid spinal surgery with the application of advanced bracing technology. Therefore, it is only prudent to advocate for conservative management first before spinal surgery is considered.

Keywords: Bracing, congenital scoliosis, conservative treatment, surgery.

INTRODUCTION

Congenital scoliosis is a lateral deformity of the spine with a disturbance of the sagittal profile caused by malformations of vertebra and ribs. The sagittal disturbance is mainly kyphotic due to malformations of the vertebral bodies, however, lordotic deformities are also possible.

In principle, there are two possible forms of malformation. Failures of formation or failures of segmentation are possible, or a combination of both, leading to a mixed deformity [1-4]. Winter [4] has noted that congenital deformities can be mostly benign to incredibly severe. These deformities can be associated with multiple other problems, may cause possible paraplegia and result in early death from cor pulmonale. These malformations present challenges for pediatricians, physicians, and surgeons because of the high frequency of severe curves, secondary medical conditions and curve stiffness compared with those of idiopathic and neuromuscular patients [4].

With congenital scoliosis there is a high frequency of associated anomalies within and outside the spine. The physical examination of the skin in the spinal midline to investigate the existence of nevi, hemangiomas or hairy patches is deemed necessary as they are signs of underlying spinal dysraphism [5]. In addition, spinal examination should focus on the cervical region due to the link between congenital scoliosis and Klippel-Feil syndrome. Moreover, a neurological examination is necessary to examine the existence of latent ataxia or myelopathy [1, 5]. These often coexist with other syndromes such as Alagille, Jarco Levin, Joubert, basal cell nevus and diabetic embryopathy. Congenital scoliosis may be associated with musculoskeletal disorders such as Sprengel's deformity, clubfeet, or DDH (developmental dysplasia of the hip) [6].

McMaster and Ohtsuka [7] were the first to focus on the natural progression of congenital scoliosis and define the risk of further deterioration (progression) in detail, in relation to four key factors: the type of congenital anomaly, spinal location, patient's age at the onset of the disorder, and solitary or multiple curves. These disorders are divided primarily into four main categories. Concerns include failure of formation, failure of segmentation, mixed defects and complex unclassifiable defects [5, 7, 8].

As stated by Kaspiris *et al*. [5], failure of formation could lead to wedge vertebra in the lower thoracic and thoracolumbar regions, however these vertebrae have a relatively low rate of progression of 1° to 2° per year. Failure of formation can be categorized into unsegmented, semi-segmented, fully segmented and multi-hemivertebrae. Unsegmented hemivertebra have minimal risk for progression since there is no potential for growth. Typically, curves do not exceed 30º at bone maturity with unsegmented hemivertebra. However, in the latter three categories the risk of progression correlates with the location, number and degree of segmentation. Progressions of curvature due to hemivertebra in relationship to location are as follows: a) upper thoracic 1º to 2º per year prior to age ten followed by 2.0º to 2.5º after this age; b) lower thoracic spine 2º per year prior to puberty followed by 2.5º to 3º afterwards; c) thoracolumbar area progresses more

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aggressively at a rate of 2.0º to 2.5º prior to puberty and 3.5º per year afterwards which results in a significant trunk imbalance. Lumbar hemivertebra progress less aggressively than thoracic vertebra [5, 7, 8].

The failure of segmentation is another category. This category includes block vertebrae which exhibit slow progression (less than 1º per year) and are typically multiple. The extent of the unilateral unsegmented bar and its position determine its natural development. In the upper thoracic spine, the rate of progression is 2° per year before puberty and 4° after. In the lower thoracic area, it is 5° and 6.5° respectively. In the thoracolumbar area, the rate of progression is to 6° before puberty and after puberty increases to 9°. In the lumbar area, it is about 5° per year [5, 7, 8].

Other categories include mixed or complex-mixed-pattern anomalies that are unrelated to the first two categories and whose development is particularly difficult to predict [5,7,8].

Cases with formation defects such as non-incarcerated, semi-carcerated or incarcerated hemivertebrae receive a variety of treatments ranging from observation to brace treatment [9, 10] or surgical intervention [11, 12]. In general, most congenital scoliotic curves are not flexible and are resistant to brace treatment. For this reason, the primary goal of brace treatment is to prevent the progression of secondary curves that develop above and below the congenital curve, which can potentially result in further imbalance [5]. Since the secondary curves are flexible bracing can have a positive influence on these curves and even though the main curve may be resistant to treatment there is potential for stabilization. As a result, it is likely that patients can have a good outcome regarding cosmetic and physical complaints.

Early surgery is suggested even in mild cases with formation failures in the first three years of life [11, 12], although that it has been cited that in this group of patients, a conservative approach may be beneficial [5].

As shown in the review from 2011 [5], despite the large number of studies concerning both the pathogenesis and the surgical and conservative treatment of congenital scoliosis, few focused on the long-term results of therapy [5]. Only two studies concerned the mid-term results and the remaining eight, the long-term results [13-20]. In most of the cases presented, skeletal maturity was not yet attained [13, 17-20].

As early surgical intervention is the suggested treatment within the first three years for patients with congenital scoliosis, a new literature search has been undertaken to find current long-term studies supporting this recommendation.

MATERIAL AND METHODS

A comprehensive review in 2011 was performed to support the hypothesis that surgical intervention is indicated in patients with congenital scoliosis [5]. No evidence was found to support early surgery in patients with congenital scoliosis [5]. This Pub Med review has been performed from 2011 to March 2015 as an update of the search made in 2011 seeking controlled studies that would support surgery for congenital scoliosis.

RESULTS

No prospective controlled or randomized controlled long-term outcome studies have been found which support the hypothesis that spinal fusion surgery in patients with congenital scoliosis can be considered as evidence-based treatment.

The safety of hemivertebra resection and segmental fusion using pedicle screw fixation [21,22] yield contradictory results. It should also be noted that utilization of a VEPTR device results in a high rate of complications [23].

DISCUSSION

No evidence (with respect to prospective controlled or randomized controlled outcome studies) has been found to support the hypothesis that spinal fusion surgery in patients with congenital scoliosis may be superior to conservative management or no treatment at all.

Indications for surgery are not in question for patients with rib synostosis and unilateral bar (failures of segmentation). However, failures of formation very often do not progress [5] and should not always be treated surgically first.

There are patients with curvatures exceeding 25° during the pubertal growth spurt whose curves did not progress even without treatment [5] (Fig. **1**).

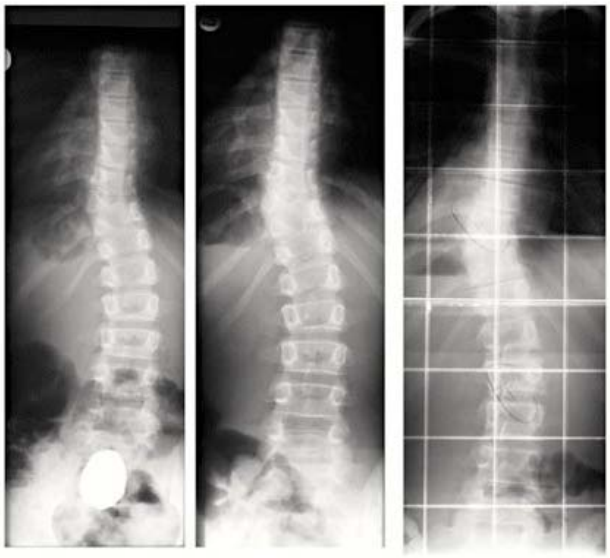

Fig. (1). No progression in a patient with a 26° thoracic and 21° lumbar curve and failure of formation from the age of 10 (premenarchial as seen on the left) to the age of 14 (2 years postmenarchial as seen on the right). This case was described in more detail in [5].

There are no clear guidelines with respect to surgical indications in children and adolescents with congenital scoliosis. Clinically, there have been instances when skeletally mature patients present with no obvious deformity or complaints; yet surgery is recommended (Fig. **2**).

There are patients with failure of segmentation whose progression may be halted with conservative measures [5] (Fig. **3**). For this reason, prior to considering surgery, consulting a specialist in the conservative management of congenital spinal deformities should be the first course of treat-

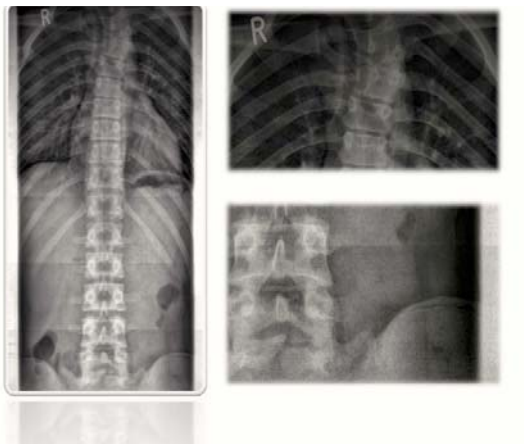

Fig. (2). Asymmetric high thoracic butterfly vertebra. Clinically, the patient was straight and without complaints. Although at Risser 4, without significant residual growth expected, a German University center recommended surgery.

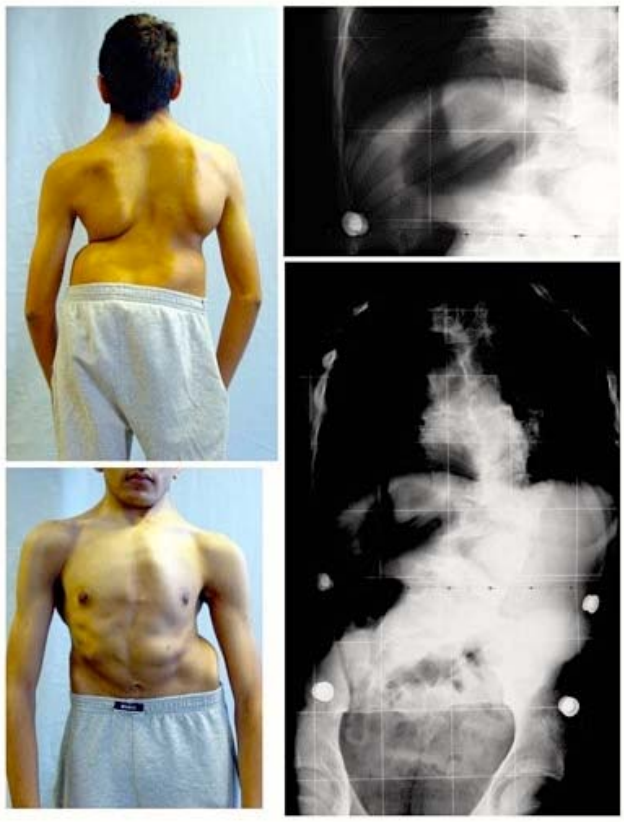

Fig. (3). Patient with failure of segmentation (rib synostosis) with clinical and radiological improvement due to conservative treatment. This fifteen-year-old boy presented at age nine with congenital scoliosis with rib synostosis due to failure of segmentation, prior to the pubertal growth spurt. At the time of his last follow-up, his clinical appearance remained severe but with an improved cosmetic appearance. The radiographs demonstrate a scoliotic curve of 59 degrees with Risser sign 4. VC was 1.640 ml, 33% of the predicted value. This case was described in more detail in [5].

ment explored. Another systematic review [23], demonstrated a high rate of complications as well. This may be due to the fact that congenital scoliosis is not a uniform condition, but varies to a great extent from one patient to another. In one study, hemivertebra excision in combination with pedicle screw fixation (Fig. **4**) rarely demonstrated complications [21] while in another study [22], complications were reported. Those complications included: fourteen of 140 subjects with pedicle screw fractures, rod fractures, pedicle elongations, removed implants, delayed incision healing, additional surgeries required due to curve progression, and one patient required prolonged respiratory support. There were no neurological complications [22].

Fig. (4). Hemivertebra excision and pedicle screw fixation on two levels in a patient with vertebral malformation.

When surgery occurs at an early age and complications occur, it could result in impairment of the young patient, potentially affecting residual growth. Therefore, the protocol to operate on all patients with malformations at an early stage is questionable.

With the use of advanced bracing technology, many patients with congenital scoliosis may be able to avoid spinal surgery [5]. However, it should be noted that the final course of the deformity is hardly predictable. It is prudent that specialists in conservative management administer the appropriate bracing protocols before spinal surgery is considered (Fig. **5**). When the curve exceeds 90º and the cardiopulmonary system is compromised surgery may be performed, however, surgery is a personal option. There are many adult scolitics with curves greater than 90º who are functioning with minimal problems. Scoliosis surgery is often performed for cosmetic and psychological reasons [24].

CONCLUSIONS

No evidence (with respect to prospective controlled or randomized controlled outcome studies) has been found to support the hypothesis that spinal fusion surgery in patients with congenital scoliosis is superior to no treatment or conservative management.

For many patients with congenital scoliosis, the advancements in bracing technology may mean that spinal sur-

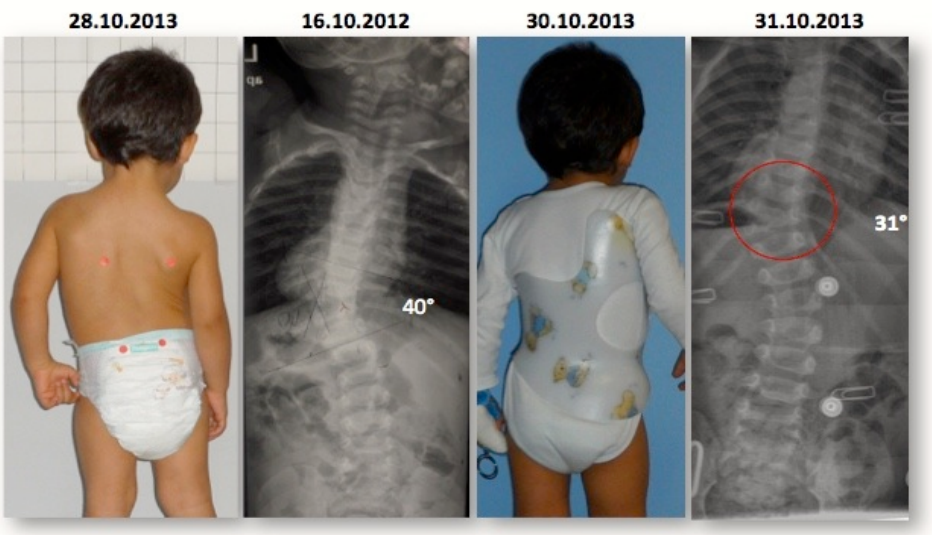

Fig. (5). Brace treatment in a two- year-old boy. Although the in-brace correction in the brace is not significant (right x-ray), clinically the deformity of the trunk is mirrored (in-brace picture compared to the picture without a brace on).

gery may potentially be avoided. However, the course of congenital scoliosis is highly unpredictable and parents may want to pursue management via conservative options prior to surgery since there are cases where successful management has occurred [5].

COMPETING INTERESTS

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CONFLICT OF INTEREST

The authors confirm that this article content has no conflict of interest.

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HRW prepared the initial draft. Both authors worked on the paper and both have read and approved the final manuscript. Written informed consent has been achieved from all patients visible in the pictures.

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REFERENCES

[1] Hedequist D, Emans J. Congenital Scoliosis: A review and update. J Pediatr Orthop 2007; 27(1): 106-16.

[2] Arlet V, Odent T, Aebi M. Congenital scoliosis. Eur Spine J 2003; 12: 456-63.

[3] Hensinger RN. Congenital scoliosis Aetiology and Associations. Spine 2009; 34(17): 1745-50.

[4] Winter RB. Congenital Spinal Deformity. In: Lonstein JE, Bradford DS, Winter RB, Ogilvie JW: Moe´s Textbook of Scoliosis and Other Spinal Deformities. Saunders, Philadelphia 3rd edition, 1995.

[5] Kaspiris A, Grivas TB, Weiss HR, Turnbull D. Surgical and conservative treatment of patients with congenital scoliosis:a search for long-term results. Scoliosis 2011; 6: 1.

[6] Debnath UK, Goel V, Harshavardhana N, Webb JK. Congenital scoliosis - Quo vadisż. Indian J Orthop 2010; 44(2): 137-47.

[7] McMaster M, Ohtsuka K. The natural history of congenital scoliosis: a study of 251 patients. J Bone Joint Surg 1982; 64A: 1128-47.

[8] Marks DS, Qaimkhani SA. The natural history of congenital scoliosis and kyphosis. Spine 2009; 34(17): 1751-5.

[9] Weiss HR. Congenital scoliosis - presentation of three severe cases treated conservatively. Stud Health Technol Inform 2008; 140: 310-3.

[10] Repko M, Krbec M, Burda J, *et al*. Simple bony fusion or instrumented hemivertebra excision in the surgical treatment of congenital scoliosis. Acta Chir Orthop Traumatol Cech 2008; 75(3): 180-4.

[11] Ruf M, Harms J. Posterior hemivertebra resection with transpedicular instrumentation: early correction in children aged 1 to 6 years. Spine 2003; 28(18): 2132-8.

[12] Winter RB, Moe JH, Eilers VE. Congenital scoliosis: A study of 234 patients treated and untreated. J Bone Joint Surg Am 1968; 50: 1-15.

[13] Winter RB, Smith MD, Lonstein JE. Congenital scoliosis due to unilateral unsegmented bar: posterior spine fusion at age 12 months with 44-year follow-up. Spine 2004; 29(3): E52-5.

[14] Winter RB, Lonstein JE. Congenital scoliosis with posterior spinal arthrodesis T2-L3 at age 3 years with 41-year follow-up: A case report. Spine 1999; 24(2): 194-7.

[15] Winter RB, Lonstein JE. Congenital thoracic scoliosis with unilateral unsegmented bar and concave fused ribs: rib osteotomy and posterior fusion at 1 year old, anterior and posterior fusion at 5 years old with a 36-year follow-up. Spine 2007; 32(26): E841-4.

[16] Thompson AG, Marks DS, Sayampanathan SR, Piggott H. Long-term results of combined anterior and posterior convex epiphysiodesis forcongenital scoliosis due to hemivertebrae. Spine 1995; 20(12): 1380-5.

[17] Marks DS, Sayampanathan SR, Thompson AG, Piggott H. Long-term results of convex epiphysiodesis for congenital scoliosis. Eur Spine J 1995; 4(5): 296-301.

[18] Bollini G, Docquier PL, Viehweger E, Launay F, Jouve JL. Thoracolumbar hemivertebrae resection by double approach in a single procedure: long-term follow-up. Spine 2006; 31(15): 1745-57.

[19] Lazar RD, Hall JE. Simultaneous anterior and posterior hemivertebra excision. Clin Orthop Relat Res 1999; (364): 76-84.

[20] Chang DG, Kim JH, Ha KY, Lee JS, Jang JS, Suk SI. Posterior Hemivertebra Resection and Short Segment Fusion with Pedicle Screw Fixation for Congenital Scoliosis in Children under Age 10 Years: Greater than 7-Year Follow-Up. Spine (Phila Pa 1976) 2015.

[21] Guo J, Zhang J, Wang S, *et al*. Complications and prophylaxis strategies of posterior hemivertebra resection for congenital scoliosis. Zhonghua Wai Ke Za Zhi 2014; 52(8): 566-70.

[22] Lucas G, Bollini G, Jouve JL, *et al*. Complications in pediatric spine surgery using the vertical expandable prosthetic titanium rib: the French experience. Spine (Phila Pa 1976) 2013; 38(25): E1589-99.

[23] Weiss H, Goodall D. Rate of complications in scoliosis surgery - a systematic review of the Pub Med literature. Scoliosis 2008; 3: 9.

[24] Westrick E, Ward W. Adolescent idiopathic scoliosis: 5-year to 20-year evidence-based operative results. J Pediatr Ortho 2011; 31(1 suppl): S61-68.

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