REVISIÓN DE LITERATURA

CIFOESCOLIOSIS CONGÉNITA CON HEMIVÉRTEBRA: SERIE DE 14 CASOS Y REVISIÓN DE LA LITERATURA

CONGENITAL KYPHOSCOLIOSIS WITH HEMIVERTEBRA: A 14-CASE SERIES AND LITERATURE REVIEW

CIFOESCOLIOSE CONGÊNITA COM HEMIVÉRTEBRA: SÉRIE DE 14 CASOS E REVISÃO DA LITERATURA

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Resúmen

Antecedentes: La hemivértebra resulta de una anomalía en el desarrollo del cuerpo vertebral y es causa de escoliosis congénita.

Objetivo: Describir la experiencia con la corrección quirúrgica de la cifoescoliosis congénita con hemivértebra y revisar la literatura para orientar su diagnóstico y manejo oportuno.

Material y métodos: Se realizó un estudio retrospectivo de serie de casos, desde enero de 2006 a julio de 2017, mediante la revisión de las historias clínicas de los pacientes menores de 14 años incluídos.

Resultados: Se incluyeron 14 pacientes con edades comprendidas entre 3 meses y 13 años, con una edad media de 6,9 años, el 78,6% eran varones. El seguimiento osciló entre 18 y 144 meses. Dos pacientes se perdieron durante el seguimiento. Se documentaron veintidós hemivértebras, 10 en la columna torácica (45,4%), seis en la columna cervical (27,3%) y seis en la columna lumbar (27,3%). El procedimiento quirúrgico más común fue la resección de hemivértebras junto con fusión instrumentada, retirándose el material de osteosíntesis en cuatro pacientes con ≥2 años de seguimiento. Tres pacientes tuvieron complicaciones no neurológicas.

Conclusión: La resección de hemivértebras junto con la fusión instrumentada es un procedimiento quirúrgico seguro y eficaz para prevenir y corregir la deformidad de la columna. Además de un diagnóstico y tratamiento tempranos, es necesario un seguimiento adecuado por parte de un equipo multidisciplinario para obtener mejores resultados.

Palabras clave: columna vertebral, escoliosis congénita, diagnóstico, tratamiento.

Abstract

Background: Hemivertebra results from an anomaly in the development of the vertebral body and is a cause of congenital scoliosis.

Objective: To describe our experience with the surgical correction of congenital kyphoscoliosis with hemivertebra and review the literature to guide its diagnosis and timely management.

Material and methods: A retrospective case series study, from January 2006 to July 2017, was conducted by reviewing the medical records of patients under 14 years of age included.

Results: The study included 14 patients aged 3 months to 13 years, with a mean age of 6.9 years, 78.6% were male. Follow-up ranged from 18 to 144 months. Two patients were lost to follow-up. Twenty-two hemivertebrae were documented, 10 in the thoracic spine (45.4%), six in the cervical spine (27.3%), and six in the lumbar spine (27.3%). The most common surgical procedure was hemivertebra resection along with instrumented fusion, the osteosynthesis material being removed from four patients at ≥2 years of follow-up. Three patients had none-neurological complications.
Conclusion: Hemivertebra resection along with instrumented fusion is a safe and effective surgical procedure to prevent and correct spinal deformity. In addition to early diagnosis and treatment, adequate follow-up by a multidisciplinary team is needed for better results.

Keywords: spine, congenital scoliosis, diagnosis, treatment.

Resumo

Introdução: A hemivértebra resulta de uma anomalia no desenvolvimento do corpo vertebral e é causa de escoliose congênita.

Objetivo: Descrever a experiência com a correção cirúrgica da cifoescoliose congênita com hemivértebra e revisar a literatura para orientar seu diagnóstico e manejo oportuno.

Materiais e Métodos: Foi realizado um estudo retrospectivo de série de casos, de janeiro de 2006 a julho de 2017, por meio da revisão dos prontuários dos pacientes menores de 14 anos incluídos.

Resultados: foram incluídos 14 pacientes com idades entre 3 meses e 13 anos, com média de idade de 6,9 anos, 78,6% eram homens. O acompanhamento variou de 18 a 144 meses. Dois pacientes foram perdidos no acompanhamento. Foram documentadas 22 hemivértebras, sendo 10 na coluna torácica (45,4%), seis na coluna cervical (27,3%) e seis na coluna lombar (27,3%). O procedimento cirúrgico mais realizado foi a ressecção de hemivértebras juntamente com fusão instrumentada, com remoção de material de osteossíntese em quatro pacientes com ≥2 anos de seguimento. Três pacientes apresentaram complicações não neurológicas.
Conclusões: A ressecção de hemivértebras juntamente com a fusão instrumentada é um procedimento cirúrgico seguro e eficaz para prevenir e corrigir deformidades da coluna vertebral. Além do diagnóstico e tratamento precoces, é necessário um

acompanhamento adequado por equipe multidisciplinar para obtenção de melhores resultados.

Palavras chave: coluna vertebral, escoliose congênita, diagnóstico, tratamento.

Introduction

Hemivertebra results from abnormal development of the vertebral body in the early stages of embryonic development (first 6 weeks), with subsequent asymmetric growth of the spine.1,2 Congenital scoliosis is defined as a lateral spinal curvature >10° measured by the Cobb method, though it is rarely clinically evident at birth. The deformity increases in two periods of rapid growth, childhood, and adolescence,1,2 and represents 10% of scoliotic deformities, being present in 0.5-1 out of every 1000 live births in North America.3-6 Thus, it is unusual but not rare.

It is unclear the mechanism conducing to congenital developmental anomalies of the spine. Animal studies suggest maternal exposure to toxins such as carbon monoxide,7 maternal diabetes,8 antiepileptic drugs, and genetic susceptibility factors.6 Multiple isolated genetic alterations have been implicated, among which the PAX1 gene has been deemed important for signaling in the ventromedial differentiation of the sclerotome. Alterations in other genes involved in mitogenesis have been described.9-12 The spinal anomaly types can be divided into formation failures (wedge vertebra and hemivertebra), segmentation failures (block vertebra and vertebral bar), and mixed types. The indications for surgical treatment of hemivertebrae are not clearly defined; however, spinal curve progression caused by a single hemivertebra, in addition to cardiopulmonary and neurological involvement, are all indicators. When surgery is necessary, options include in situ fusion, convex hemiepiphysiodesis, and hemivertebra resection.2,13

The approach to these patients is not simple, as the natural history of curve progression along with the risk of thoracic insufficiency and neurological deficits poses a challenge for neurosurgical treatment. Here, we describe our experience with the surgical correction of congenital kyphoscoliosis with hemivertebra at the [Blinded] and review the literature to guide its diagnosis and timely management.

Materials and Methods

A retrospective case series study, from January 2006 to July 2017, was conducted by reviewing the medical records of 14 patients under 14 years of age recruited from the outpatient clinic and operated on by the principal investigator at the [Blinded]. The

variables recorded were age, sex, deformity type, hemivertebra location, surgical procedure, Cobb angles, complications, and follow-up time.

Most patients were referred from the pediatric outpatient clinic. Diagnostic imaging for all patients included full-spine anteroposterior and lateral radiographs with dynamic views, full-spine computed tomography (CT) scan with 3D reconstruction, and full-spine magnetic resonance imaging (MRI). Spinal angiography was performed in two cases and myelo-CT scan in one case.

Results

The study included 14 patients with congenital kyphoscoliosis due to hemivertebra, aged from 3 months to 13 years, with mean age of 6.9 years, 78.6% of whom were male and 21.4% female. Follow-up ranged from 18 to 144 months. Two patients were lost to follow-up.

A total of 22 hemivertebrae were documented, 10 in the thoracic spine (45.4%), six in the cervical spine (27.3%), and six in the lumbar spine (27.3%). The most common surgical procedure was hemivertebra resection along with instrumented fusion (nine cases), after which we removed the osteosynthesis material in four patients at ≥2 years of follow-up. Three patients had complications: one intraoperatively (case 13, cerebrospinal fluid fistula) and two postoperatively (case 1, surgical site infection, treated with surgical lavage and enterostomal therapy; and case 11, pressure-related ulcers that resolved with medical treatment). There were no neurological complications associated with the procedure (Table 1).

Table 1. Description of cases

# Case	Age years	Sex*	Deformity**	H+	Instrumentation	Intervention++	Cobb angle-i	Complication	Follow-up (months)
1	6	M	Scoli	Left T11 & right L3	T10-T12-L2-L4	HR+ discectomy + IF by pa	28	Surgical site infection	48
2	12	F	Kyphos	Posterior T5 & T6	T4-T7 / C7-T1 -T10-T11	HR+ discectomy by ap + bone graft + Instrumentation by pa	30	No	144
3	6	M	Scoli	Right C4 & C5	-	HR+ discectomy by ap	20	No	NS
4	9	F	Scoli	Left L1	T8-T9 & L2-L3	IF by pa	25	No	84
5	13	M	Kyphos	Right T11	Cylinder + T10-T12 by anterior approach/T4-T5 & L1-L2	HR+ discectomy by ap + IF by pa	40	No	120

					by posterior approach				
6	11	M	RotoScoli	Right L2	T11-T12-L1 & L3-L4-L5	HR+ discectomy by pa + IF by pa	12°	No	NS
7	2	M	Scoli	Left C7	-	HR+ discectomy by ap + anterior cervical plate	15	No	84
8	1	M	Scoli	Left T6 & right T11	-	HR+ discectomy	22	No	24
9	0.5	M	Scoli	Left C7 - T1	-	HR+ discectomy + bone graft by ap cervicotomy		No	96
10	0.3	M	Scoli	Left T11 - right T5	-	HR+ discectomy by pa		No	21
11	0.7	M	Kyphos	Left L2	T11-L1, L3-L5, titanium wire T12 & L4	HR+ discectomy + IF by pa	30	Pressure-related ulcers	24
12	10	M	Scoli	Left T7	T7-T8-T9	HR+ discectomy + IF by pa		No	27
13	11	F	Kyphos	Right L1 - L2	T9-T10-T11-L3-L4-L5	HR+ discectomy IF by pa	40-50	Intraoperative CSF fistula	18
14	4	M	Scoli	Left C6 - C7 + Bone malformation at C1-C2	C6-C7 – anterior plate C0-C3-C4 by posterior approach	HR+ discectomy by ap + resection of bone malformation at C1 - C2 by pa + occipitocervical fixation		No	90

*M = male and, F = female. ** Kyphos = Kyphoscoliosis and, Scoli = Scoliosis. +H = Hemivertebra. ++ HR = Hemivertebra resection, IF =Instrumented Fusion, posterior approach = pa, anterior approach = ap. **Cobb angle-i = Initial Cobb angle**

Illustrative cases

Case 1

Six-year-old male patient with diagnosed with congenital scoliosis due to left T11 and right L3 hemivertebrae without neurological deficit at 5 months of age. Orthotic treatment was initiated, with poor adherence. In September 2011, progression of the scoliotic curve was evident, reaching a thoracic Cobb angle of 28° and lumbar Cobb angle of 20°, which increased the following year to 33° and 22°, respectively. Instrumented hemiepiphysiodesis was used for surgical correction of the left T10-T12 and right L2-L4 segments, which decreased the thoracic and lumbar Cobb angles to 12° and 10°, respectively. Twelve months after surgery, the patient developed a surgical site infection. We treated it with surgical lavage, enterostomal therapy, plastic surgery, and wound closure for the second time. Two years after surgery, there was significant improvement of the curve and the T11 and L3 unsegmented hemivertebrae, leading to the removal of surgical material from both segments without complications (Fig. 1).

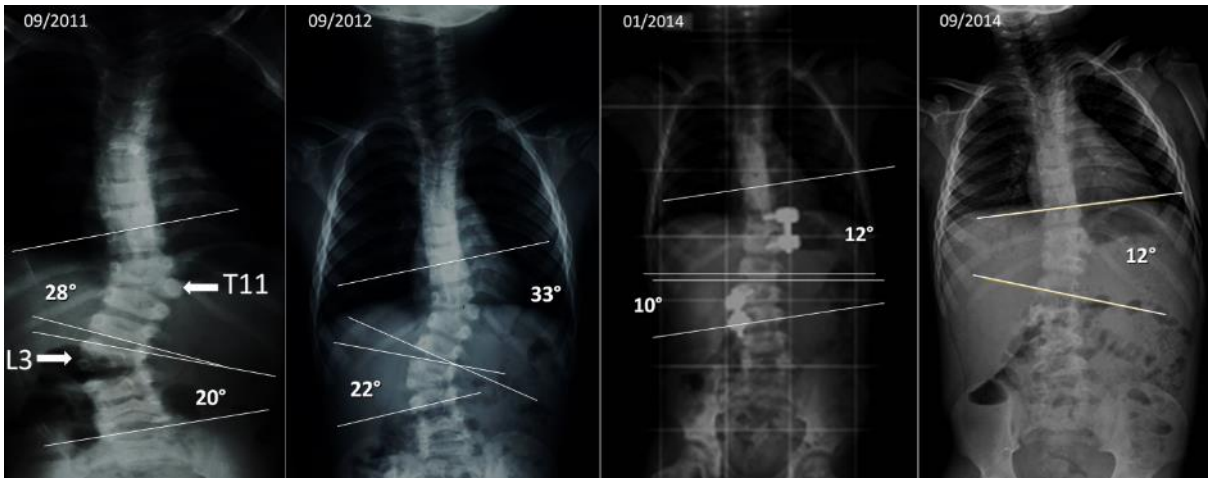

Fig. 1. Case 1. A 6-year-old boy with left T11 and right L3 hemivertebra, treated with hemivertebra resection and instrumentation, with subsequent removal of osteosynthesis material.

Case 2

Twelve-year-old female patient was admitted for consultation in 2006 due to a 6-month clinical picture of paraparesis with severe left lower limb involvement (T6 sensory level). Spinal MRI showed compressive myelopathy by posterior hemivertebrae at T5-T6 with rotoscoliosis and marked kyphosis of 80°. A two-stage surgical procedure was undertaken. The first operation was performed in 2006, accessing the convexity (left side) and removing the T5-T6 hemivertebrae and adjacent discs. A fibular allograft was placed and fixed with transpedicular screws at the T4-T7 levels along with a left lateral rod. The second operation was performed by placing hooks at C7, T1 and T10, T11 plus bars and grafts. The radiographic following in 2014 and 2016 showed a left scoliosis T1-T8 of °28 and kyphosis T1-T12 of °75. Finally, an acceptable coronal and sagittal balance was observed, the latter slightly posterior to the sacrum with compensatory hyperlordosis and a horizontal sacrum. Currently, the patient has no sensory symptoms or lower limb weakness and shows hyperreflexia and the Babinski sign.

Case 5

Thirteen-year-old male patient with a history of progressive axial pain for 2 years hampering physical activity. The patient had no neurological deficit but showed a progressive thoracolumbar kyphosis deformity with a Cobb angle of 40°, right T11 hemivertebra, and anterior T12-L2 spinal cord compression, with signs of myelopathy and syringomyelia. The patient underwent two-stage surgery through anterior and posterior approaches. The anterior approach involved thoracic surgery, retroperitoneal thoracolumbotomy, and repair of the diaphragm with unusual access on the side of the

concavity, contralateral origin of the Adamkiewicz artery arising from the right side, T11 corpectomy, T10-T11 and T11-T12 discectomy, a bone-filled titanium cylinder, and anterior fusion of T10-T12 with titanium screws and rod. The second surgical stage consisted of posterior multilevel fusion with transpedicular screws at the T4-T5 and L1-L2 levels and a left T8 sublaminar hook. There were no systemic or neurological complications (Fig. 2).

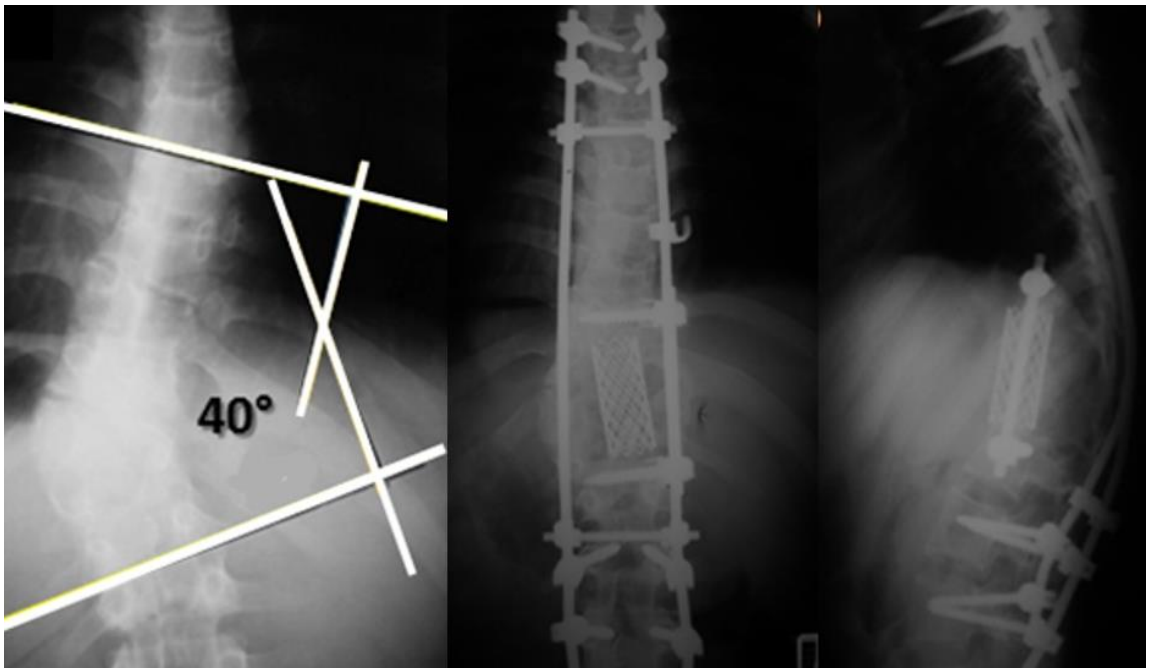

Fig. 2. Case 5. A 13-year-old boy with T11 hemivertebra, treated with hemivertebra resection with placement of a cylinder by the anterior approach, then instrumented fusion by the posterior approach.

Case 11

Seven-month-old male patient with progressive deformity due to left L2 hemivertebra with kyphoscoliosis. The patient underwent fixation with T11-L1 and L3-L5 sublaminar hooks and T12 and L4 titanium wire attachments. We removed the instrumentation 12 months after surgery (Fig. 3).

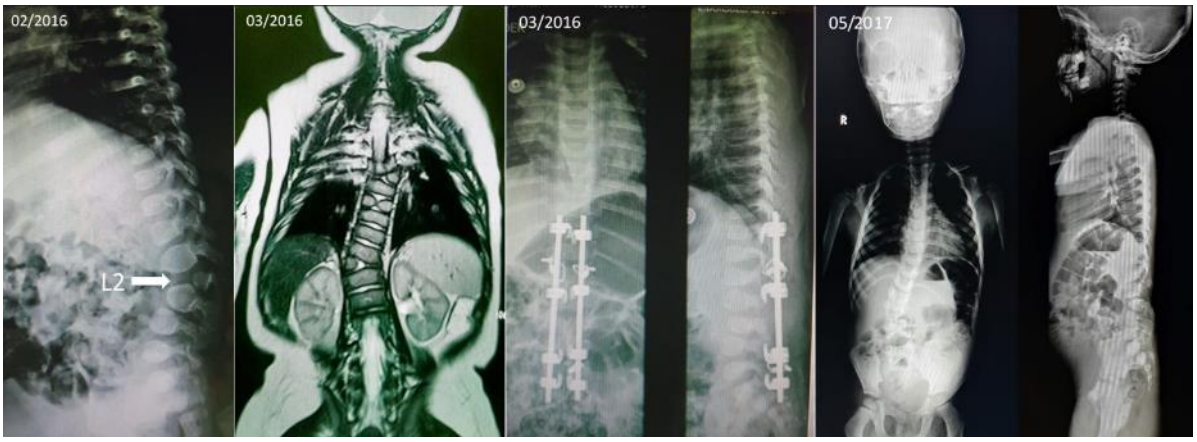

Fig. 3. Case 11. A 7-month-old boy with L2 hemivertebra, treated with hemivertebra resection and instrumentation, with subsequent removal of osteosynthesis material.

Case 14

Four-year-old male patient with cervicalgia, right lateral neck tilt, and left C6-C7 hemivertebra with ipsilateral C1-C2 bone malformation displacing the spinal cord. The patient underwent two-stage surgery. The first consisted of hemivertebra resection by left C6-C7 cervicotomy, the second, resection of bone malformation and C1-C2 laminectomy with occipital fixation using screws and C3-C4 sublaminar hooks.

Based on our experience with the observed cases, we drew a flowchart to guide the surgical treatment of patients with congenital scoliosis (Fig. 4).

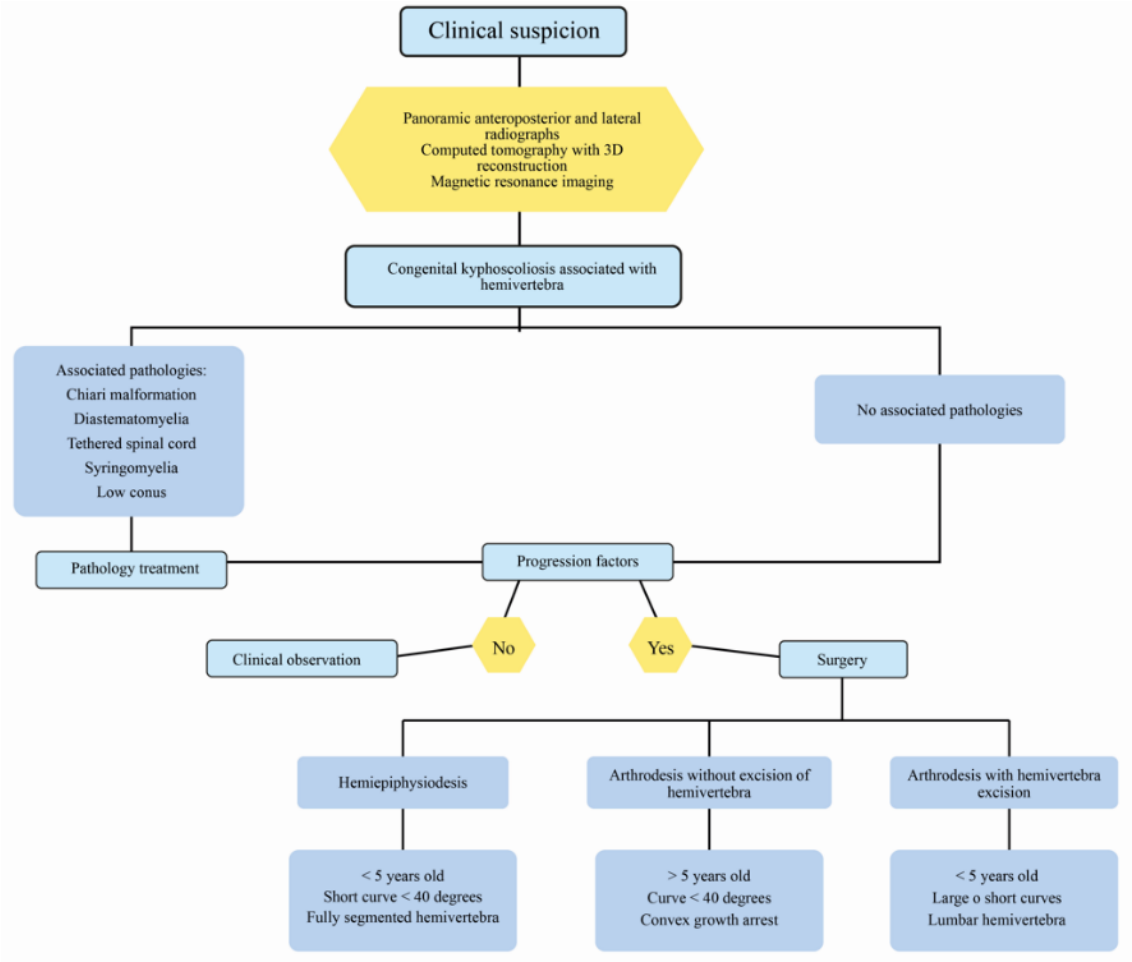

Fig. 4. Algorithm for managing congenital Kyphos with hemivertebra.

Discussion

Congenital spinal malformations are classified into three main groups, neural tube closure defects (spina bifida, meningocele, myelomeningocele), segmentation disorders (block vertebra, unsegmented bar), and formation disorders (wedge vertebra, hemivertebra). Congenital scoliosis is classically secondary to segmentation or formation failure, or a mixed form. The results of the deformity depend on the growth asymmetry on the opposite side of the spine and its location in the specific spinal segment(s).13-16

The most common anomaly is hemivertebra, classified according to its relationship to the vertebrae above and below: fully segmented when separated from each adjacent vertebra by a disc, partially segmented when separated from one of the adjacent vertebrae by a disc, and unsegmented when there is no disc material separating the

hemivertebra from the adjacent vertebrae. The presence of disc material is important for growth potential.15

Kyphoscoliosis and congenital kyphosis are less common than congenital scoliosis, but the latter can have serious consequences if left untreated due to the risk of spinal cord compression and paraplegia. It was classified into three types by McMaster & Singh: type I for anterior formation failure (posterior hemivertebra), type II for an anterior unsegmented bar, and type III for mixed defects. The worst prognosis is associated with type I in the upper thoracic segment.15-17

Hemivertebra can be diagnosed early in the prenatal period by ultrasound, although one-third of cases are usually observed after delivery. In the newborn, skin anomalies on the lumbosacral midline may suggest spinal dysraphism, and trunk asymmetries detected during the follow-up of the child's growth and development are also warning signs of this condition. In the physical examination, the back of the patient must be examined while standing and during forward trunk flexion. The shoulder level, waist folds, and lower limb length must be assessed. Last, a complete neurological examination must be done.

It is recommended to study panoramic radiographs in the anteroposterior and lateral views to evaluate the presence of kyphosis in the deformity, the coronal and sagittal balance, lumbopelvic angle, and pelvic incidence. Interobserver variability of 12° has been documented in severe curves when not using the same reference points for measurement. Axial, coronal, sagittal, and 3D-reconstruction CT scans can be very helpful in understanding the complex malformation patterns. Neuroaxis MRI, angiography, and myelo-CT scan are reserved for cases of abnormal neurological examination, findings suggestive of spinal dysraphism, and progressive curves that are candidates for surgery.18,19

According to McMaster & Singh,17 curve progression in congenital scoliosis depends on four aspects: defect type, defect location, age at presentation, and number of curves. Spinal growth is greatest in the first 3 years of life and puberty, so deformities diagnosed at an early age have a severe prognosis due to the curve progression until skeletal maturity. Thoracic curves have a worse prognosis, particularly in the thoracolumbar junction, and cervicothoracic curves progress slowly but have a greater aesthetic impact. The most severe malformation is a unilateral unsegmented bar with contralateral hemivertebra, followed by a unilateral unsegmented bar, multiple hemivertebrae, single hemivertebra, wedge vertebra, and last, block vertebra.

The curve pattern in a patient is difficult to predict; curves of 10-25° are deemed to show little progression, and those >25° have a high risk of progression. Most curves in

congenital scoliosis are progressive, and at most 25% of cases are nonprogressive.20 Skeletal maturity should be assessed by radiological signs such as Risser's sign, triradiate cartilage closure, and carpal ossification. Patients with Risser stage 0-1 (bone immaturity) and curves >20° are at high risk of progression.21

The main goal of treatment is to prevent severe deformity. Orthotic treatment has not been effective at correcting the congenital scoliosis curve and may hinder thoracic cavity development. However, some studies consider that the main goal of bracing treatment is to prevent curve progression above and below the congenital curve, while others reserve its use to ensure postoperative stability in infants.22

Surgery is justified for scoliotic curves exceeding 40-50°, curves showing a steady progression, or curves with unilateral unsegmented bars (with or without contralateral hemivertebra), particularly in children under 5 years of age. Early surgery is advised and widely accepted. Fixation approaches include in situ fusion, convex hemiepiphysiodesis, hemivertebra resection, and instrumented fusion.23 Nonfusion surgery such as expansion thoracoplasty or vertical expandable prosthetic titanium ribs may be useful in young patients in whom the deformity may develop, although this is disputed.

To reduce the risk of neural injury associated with correction, it is necessary to conduct intraoperative monitoring of neural function with motor- and sensory-evoked potentials. In case of any abnormality, some studies recommend routine evaluation with the intraoperative wake-up test.23-26

Hemiepiphysiodesis

This technique requires growth to achieve correction over time and is reserved for patients with small preoperative curves. It is the procedure of choice for young patients (under 5 years) with a high risk of progression and segmented hemivertebra with no other associated anomalies. It is performed on the convex side of the curve while the concave side retains its growth potential, theoretically allowing some correction of the deformity, but this is negligible in practice (mean 10°).27

Fusion without hemivertebra resection

In early series of patients with congenital scoliosis, pseudarthrosis developed without much correction of the deformity. Currently, osteotomy and facetectomy allow the creation of a stable and balanced spine. The procedure is performed in older patients. It has a lower risk of complications than fusion with hemivertebra resection.28

Fusion with hemivertebra resection

This technique provides the greatest correction with little instrumentation. It is recommended in very young patients, under 4 years of age. The procedure is ideally used in lumbosacral hemivertebrae and is performed via anterior and posterior approaches or only a posterior approach. When resection is performed by anterior access to the thoracolumbar junction, it is recommended to use, according to the extent, the lumbotomy approach of Hodgson (intrapleural retroperitoneal from T9 to L5) or Mirbaha (extrapleural retroperitoneal from T11 to L5). Most cases require posterior transpedicular fixation to ensure adequate stability and alignment. The posterior-only approach has been taken with transpedicular subtraction. This approach to hemivertebra resection has slightly more neurological complications than the other surgical techniques.29,30

In postoperative management, it is recommended to follow-up with panoramic radiographs of the spine in the anteroposterior and lateral views, along with axial, coronal, and sagittal CT scans of the spine every 6 months for 2 years and then continue with annual imaging follow-up. It is advisable to use a polypropylene orthosis for 3 months and administer postoperative physical therapy.

Congenital scoliosis due to hemivertebra is an uncommon condition that should be diagnosed at an early age to avoid neurological and cardiopulmonary complications and the development of major, difficult-to-correct curves. However, in the cases we report, the mean age at the time of surgery was 9.2 years. Some studies show that late treatment of this deformity requires more fusion and has a high risk of neurological deficit.15,31 Fusion with hemivertebra resection is the surgery that ensures the greatest correction (29°), with no neurological deficit documented in the present series. Although Holte et al. reported a high rate of neurological complications in their series,32 these complications occurred mostly when taking a single posterior approach for resection by transpedicular subtraction in thoracic or thoracolumbar segments. In our experience described here, this procedure was not performed because patients older than 5 years had deforming curves in need of combined anterior and posterior approaches, which brings less risk of neurological deficit. Many series report no or minimal neurological complications.33,34 Hemivertebra resection continues to be more widely accepted, providing excellent correction. Kyphoscoliosis due to hemivertebra produces greater neurological deficit; in case 2, the patient was admitted preoperatively with compressive myelopathy, and after T5-T6 hemivertebra resection, there was significant symptomatic improvement with minimal sequelae.

Limitations

Some data may be missing since we reviewed the medical records and could not contact some of the patients again.

Conclusion

Congenital kyphoscoliosis with hemivertebra is a challenging condition. Its early diagnosis can prevent severe deformity and reduce its impact on the neurological and cardiopulmonary prognosis. In the patients described here, hemivertebra resection was accompanied in most cases by instrumented fusion, which is a safe and effective surgical procedure to prevent and correct spinal deformity, as described by other studies. In addition to early diagnosis and treatment, adequate follow-up must be ensured to obtain the best results, for which a multidisciplinary team is needed (spine surgery, pediatric neurosurgery, pediatrics, pediatric surgery, physiatry, anesthesiology, nursing).

Statements

Statement of Ethics

Study approval statement: Ethical approval for this study was not required in view of the retrospective nature of the study and all the procedures being performed were part of the routine care. Written informed consent was obtained from the participants' parent/legal guardian/ next of kin to participate in the study. All data collected and used in the study were fully anonymized prior to analysis and submission for publication.

Consent to participate statement: Patient consent were not required in accordance with national guidelines.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Carlos Antonio Llanos-Lucero: Conceptualization; Data curation; Formal analysis; Investigation; Methodology; Project administration; Software; Supervision; Validation; Visualization; Roles/Writing - original draft; Writing - review & editing.

William Ricardo Vargas-Escamilla: Conceptualization; Data curation; Formal analysis; Investigation; Methodology; Project administration; Software; Supervision; Validation; Visualization; Roles/Writing - original draft; Writing - review & editing.

Lina María Villegas-Trujillo: Conceptualization; Data curation; Formal analysis; Investigation; Methodology; Project administration; Software; Supervision; Validation; Visualization; Roles/Writing - original draft; Writing - review & editing.

Data Availability Statement

The data that support the findings of this study are available from the corresponding author, upon reasonable request.

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