

ORIGINAL PAPERS

Surgical treatment of congenital scoliosis resulting from an isolated hemivertebra: 27 cases

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KEYWORDS

Congenital scoliosis;
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Abstract

Purpose: Congenital scoliosis resulting from hemivertebrae can be treated surgically through resection or convex epiphysiodesis. Our purpose was to assess the results obtained in a series of children operated using both methods.

Materials and methods: A clinical and radiological review was carried out of a non-comparative series of 27 isolated hemivertebrae of which 16 had been treated by means of resection and 11 by means of epiphysiodesis. Mean age at surgery was 50 months (24-132) and mean Cobb angle was 33° (20°-75°). In the case of lumbar hemivertebrae, resection was performed through an anterior and posterior approach; compressive CD instruments were used. Thoracic vertebral epiphysiodesis required prior excision at two levels, followed by a subsequent 4-level decortication phase: in 6 cases the vertebrae were instrumented and in 5 a plaster cast was applied in the reduced position.

Results: No neurological complication was found. With respect to the 16 resections, mean reduction was 75% postoperatively and 73% at the end of the study. For the 11 epiphysiodeses mean correction obtained was 25% postoperatively and 39% at 4 years' mean follow-up (1-8 years). There were 2 cases of long-term failure in the absence of instrumentation.

Conclusions: Resection of hemivertebrae provides immediate and stable satisfactory results. However, in the thoracic area, and given the neurological risk involved, instrumented convex epiphysiodesis guarantees a good long-term result.

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PALABRAS CLAVE

Escoliosis congénita;
Hemivértebra;
Resección;
Epifisiodesis convexa

Tratamiento quirúrgico de la escoliosis congénita por hemivértebra aislada. A propósito de una serie de 27 casos

Resumen

Objetivo: Las escoliosis congénitas evolutivas por hemivértabras pueden ser tratadas quirúrgicamente mediante resección o epifisiodesis convexa. Nuestro objetivo fue evaluar los resultados de una serie de niños operados según una u otra modalidad.

Material y método: se realizó una revisión clínica y radiológica de una serie no comparativa de 27 hemivértabras aisladas de las cuales 16 habían sido tratadas por resección y 11 por epifisiodesis. La media de edad en el momento de la intervención fue 50 (24-132) meses y el ángulo de Cobb fue de 33° (20°-75°). La resección para las hemivértabras lumbares se hizo por una vía de abordaje anterior y posterior y como instrumentación se utilizó el material Baby CD a compresión. La epifisiodesis para las hemivértabras torácicas necesitó de un tiempo anterior de escisión a dos niveles, seguido de un tiempo posterior de descorticación a cuatro niveles: en 6 casos fueron instrumentadas y en 5 casos, escayoladas en posición de reducción.

Resultados: no se constató ninguna complicación neurológica. Respecto a las 16 resecciones, la reducción media fue del 75% en postoperatorio y del 73% al final del estudio. Para las 11 epifisiodesis la corrección media obtenida fue del 25% en el postoperatorio y del 39% a los 4 años de seguimiento medio (1,8 años). Hubo 2 casos de fracaso a largo plazo en ausencia de instrumentación.

Conclusiones: la resección de las hemivértabras proporciona resultados satisfactorios inmediatos y estables en el tiempo. Sin embargo, en las localizaciones torácicas y a causa del riesgo neurológico, las epifisiodesis convexas con instrumentación garantizan el resultado a largo plazo.

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Introduction

Congenital scoliosis is the most frequent deformity of all congenital anomalies. These can be classified into three groups: growth deformities, segmentation deformities and mixed deformities, which combine growth and segmentation defects. Hemivertebrae are a kind of growth deformity. Mac Master et al¹⁰ classify hemivertebrae according to their relation with adjacent vertebrae and distinguish between fully segmented, partially segmented, unsegmented and incarcerated hemivertebrae. This is a prognostic classification; indeed, an intervertebral disc can be interpreted as a potential site of hemivertebral growth and of progression of spinal malformation.

The progression of angular scoliosis is an indication for surgical treatment. Two methods are possible: convex hemiepiphysiodesis^{3,13,15,18} and hemivertebra resection^{1,2,4,6,8,17}.

Convex epiphysiodesis arrests scoliosis progression. Gradual correction is also possible, but it is dependant on the prospective contralateral growth. Hemivertebra resection results in complete correction or, more frequently, in partial correction, but it exposes the patient to a greater neurological risk.

The aim of this study is to assess the results obtained through surgical treatment in a series of 27 isolated hemivertebrae treated by means of resection or convex epiphysiodesis.

Materials and methods

We reviewed a series of 27 consecutive isolated hemivertebrae in 24 infants (12 males and 12 females) operated between 1996 and 2004. Three patients presented 2 isolated non-contiguous hemivertebrae. Mean age at time of diagnosis was 27 (0-108) months.

The type of hemivertebrae was diagnosed through radiologic study and MRI. Following Mac Master's classification we found 19 hemivertebrae of the totally segmented type, 7 partially segmented hemivertebrae, and 1 of the incarcerated type. Systematic studies for other deformities found no medullary abnormalities or heart defects, except for one patient with a renal anomaly. One other patient presented with cerebral palsy.

Mean Cobb angle was 24° (5°-46°) at diagnosis, and 33° (20°-75°) at the moment of treatment. Mean age for surgical treatment was 50 (24-132) months. 16 patients were treated by means of hemivertebra resection and 11 by means of epiphysiodesis.

Surgical technique (resection)

Resection is performed through 2 successive approaches and on only one application of anesthesia.

The first phase is performed with the patient in the prone position. Through a posterior approach, half of the posterior arch and the whole pedicle up to the vertebral body are

resected. The Baby CD instrumentation is prepared, consisting of supralaminar and infralaminar hooks, which are situated in the vertebrae adjoining the hemivertebrae. Decortication of the posterior arches is carried out, in preparation for arthrodesis. The posterior wound is closed with a continuous suture.

Next, the child is placed in the lateral position so as to enable an anterior approach that can be, depending on localization, a thoracotomy, a thoracophrenolumbotomy or a simple lumbotomy. The body of the hemivertebra is exposed up to the pedicle base. Superior and inferior discs are removed and the hemivertebral body is resected. Epiphysiodesis is performed by removing the vertebral endplates of the adjacent vertebrae. Subsequently, the posterior incision is made and the instrumentation introduced. Compression is applied, checking its effect on both anterior and posterior regions. Following this, the bone graft is introduced in the anterior and posterior zones. Post-operatively, a thermoformed corset is applied for 6 months. The patient can stand on the third day after treatment.

Surgical technique (convex epiphysiodesis)

This technique can be performed through two different approaches: first, by means of an anterior approach, with the patient in the lateral position. A convex excision of vertebral discs and plates is carried out above and below the hemivertebra. By performing a bone excision a space is created both in the hemivertebra and in adjacent vertebrae. This cavity is filled with a graft from the iliac crest or from the ribs. Next, the patient is placed in the prone position. A posterior approach is performed, only on the convex side of the spine. Decortication of the posterior arch of the hemivertebra and of the adjacent upper and lower vertebrae is performed. An iliac crest graft is placed. Compression with Baby CD type material may be needed.

In cases with osteosynthesis, post-op support is assured by means of the use of a thermoformed-fiber corset for a period of 6 months. In cases without osteosynthesis, a plaster cast is prepared, and applied also for 6 months. In all cases, standing is recommended on the third day.

Results

Table 1 shows the results of our study. With respect to the 16 resection cases, mean follow-up was 4 (1-9) years. There were no neurological complications. In one case, a superficial post-op infection required surgical treatment without hardware extraction and with prolonged intravenous antibiotic therapy. Mean Cobb angle was 32° (20°–75°) at the pre-operative stage, 8° (0°–35°) in the immediate post-operative stage, and 7.5° (0°–31°) at the end of follow-up. Mean reduction was 75% postoperatively and 73% at the end of the study.

With respect to the 11 cases of convex epiphysiodesis, mean follow-up was 4 (1-8) years. There were no complications. In one case (T1 hemivertebra) only a posterior epiphysiodesis was performed. In 6 cases epiphysiodesis was stabilized by means of compression with

Baby CD type instrumentation and immobilization was achieved, on reduction, by means of a plaster corset. Mean Cobb angle was 31° (20°–45°) in the pre-op stage, 23° (10°–35°) postoperatively, and 19° (10°–32°) at the end of follow-up (4 years). Mean correction was 26% postoperatively and 39% at the end of follow-up. In other words, mean angular correction was 2.6° (0°–15°) yearly. In 4 cases the effectiveness of epiphysiodesis was obtained with gradual correction of the defect. In 5 cases, there was only a fusion effect and a stabilization of the defect. In 2 cases, however, there was a failure and the scoliosis progressed. There were no differences in the immediate postoperative results between epiphysiodeses instrumented with Baby CD and epiphysiodeses performed without instrumentation. The 2 long-term failures occurred in cases of epiphysiodeses without instrumentation.

Discussion

Surgical indications are not well defined in cases of hemivertebrae. In theory, they are based on the progression of the scoliosis. But in many cases progression may be difficult to determine. Loder et al⁹ studied the variability of Cobb's angle in children with congenital scoliosis. They found an intra-observer variability of around 9.6°, and an inter-observer variability of around 11.8°. This would imply that to prove the evolution of scoliosis an angular progression of 23° is needed. In many cases this aggravation would not become evident till after the patient is 5 years old, and this reduces the possibilities of successful surgical treatment. The classification contributed by Mac Master et al^{10,11} is helpful, since it enables a better definition of surgical indications. In general, we favor early surgical treatment in cases of free and partially segmented hemivertebrae, as well as in cases of lumbosacral hemivertebrae causing spine obliqueness. However, our indications for unsegmented and incarcerated hemivertebrae are exceptional (only one case in our series).

Our series does not allow us to carry out a comparison between epiphysiodesis and resection. We based our indications on the topography of the defect: epiphysiodesis in the thoracic area and resection in the lumbar area, with the intention of avoiding neurological risks that we deem to be greater in resections in the thoracic area.

Whenever possible, we perform both an anterior and posterior epiphysiodesis. In our series, we performed an isolated posterior epiphysiodesis in only one case located at the level of T1.

Winter et al¹⁸ showed that isolated posterior epiphysiodesis could bring about the so-called crankshaft defect, which causes a loss of efficiency. However, Winter et al¹⁹ have recently presented the case of a patient treated by means of posterior epiphysiodesis for a segmentation defect at the age of 12 months; this patient still showed excellent results at the age of 44.

In order for epiphysiodesis to be effective, the posterior fusion must extend over 4 levels. Ominus et al¹³ showed that a short fusion, at 2 levels, causes angular progression; a fusion at 4 levels, on the other hand, corrects the angulation.

Table 1 Cases

Level	Type	Age at diagnosis (months)	Age at operation (months)	Operation type	Instrumentation	Cobb angle (initial)	Cobb angle (pre-op)	Cobb angle (post-op)	Cobb angle (final)	Follow-up (years)
L4	1/2 F	6	36	Resection	Yes	22°	22°	0°	5°	8
L5	F	10	24	Resection	Yes	10°	35°	20°	15°	4
L5	1/2 F	34	106	Resection	Yes	25°	25°	0°	0°	1
L2	F	2	24	Resection	Yes	46°	75°	35°	20°	8
L5	F	84	96	Resection	Yes	18°	30°	15°	15°	5
L5	F	3	56	Resection	Yes	20°	30°	5°	5°	3
L3	1/2 F	3	42	Resection	Yes	20°	27°	0°	0°	2
L4	F	7	24	Resection	Yes	30°	35°	0°	0°	3
L4	F	12	48	Resection	Yes	20°	25°	0°	0°	5
L4	F	0	26	Resection	Yes	20°	30°	5°	5°	1
L2	1/2 L	58	72	Resection	Yes	30°	30°	0°	5°	9
L4	1/2 F	18	48	Resection	Yes	5°	27°	5°	5°	1
L4	F	12	24	Resection	Yes	35°	35°	0°	0°	2
L3	F	102	102	Resection	Yes	15°	20°	0°	0°	4
L4	F	5	24	Resection	Yes	20°	35°	28°	31°	7
L4	F	3	18	Resection	Yes	25°	25°	10°	15°	6
D10	INC	3	33	EP; D9D11	Yes	30°	40°	30°	5°	4
D1	F	108	132	EP; C7D2	No	27°	30°	30°	30°	6
L2/L3	1/2 F	36	48	EP; D12L3	No	20°	30°	24°	23°	6
L3	1/2 F	29	70	EP; L1L4	No	26°	28°	20°	17°	2
L1/L2	F	13	36	EP; L1L3	Yes	35°	35°	35°	10°	2
D12	F	11	24	EP; D11L1	Yes	20°	20°	20°	10°	2
D6/D7	F	60	87	EP; D6D8	Yes	20°	25°	20°	10°	1
D8	F	3	24	EP; D6D10	Yes	25°	30°	18°	15°	2
D8	F	9	32	EP; D6D9	No	25°	30°	15°	35°	5
D9	F	10	60	EP; D8D10	Yes	29°	45°	20°	20°	5
D9	F	5	24	EP; D7D11	No	20°	30°	25°	32°	8

EP: epiphysiodesis; INC: incarcerated; F: free; 1/2 F: partially segmented.

Equally significant is age at the moment of surgical treatment. The earlier the treatment the greater the chances of achieving an effective epiphysiodesis (fig. 1). Dubouset et al³ recommend operating when the patient is less than 5 years of age and suggest that the graft area must be inspected after 4 months, which we have not done and which could explain our 2 failed results.

On the other hand, we obtained satisfactory results when we used compressive osteosynthesis. This technique, which is generally used in resection surgery, can be adapted to epiphysiodesis. It contributes a small perioperative correction and seems to enhance the effect of the bone graft used in epiphysiodesis (fig. 2).

Results of epiphysiodesis are difficult to compare in the series found in the literature. This is due to the fact that the results depend on the patient's age at the moment of treatment, on the extension of the epiphysiodesis and, most importantly, on the contralateral growth potential.

In their series, Dubouset et al³ found 20 corrections, 19 fusions and 4 failures, which they attributed to the crankshaft defect. Thompson et al¹⁵ contributed 15 cases with 23 corrections, 5 stabilizations and 2 failures. They observed a 1.9° angular progression prior to surgical treatment and a 1.2° yearly reduction following treatment; this yielded a 3.1° differential.

Keller et al⁷ recommend anterior transpedicular epiphysiodesis. In a series of 19 cases that were operated in patients with a mean age of 4 years and 8 months, they obtained 7 corrections, 8 stabilizations and 4 failures. These results do not seem to be as good as those obtained with the two levels. This may be due to the difficulty found in determining the growth zone in a vertebral body that is for the greater made of cartilage.

As regards resections, the angular correction in the perioperative stage is significant. Results are stable over time (figs. 3 and 4). There is very little or no complementary epiphysiodesis effect. Our results are equivalent to other results in the literature. The correction percentage obtained in T12 and L4 resections is 71% in the series presented by Hedequist et al⁵ and 67% in the Callahan et al⁴ series. Klemme et al⁸ have extended the indication of resection to thoracic vertebrae obtaining a 67% reduction and no neurological complications. Holte et al⁶, instead, have reported 7 neurological complications in a series of 37 operated cases. Nevertheless, their series is different, since their mean age is 12 years and their angulations reach 132°. This series confirms the need for early surgical treatment. Some authors have performed resections at only one posterior level^{12,14,16}. Resection of the vertebral body is performed by either an extrapleural or a retroperitoneal approach depending on the level being treated. The published results showing a correction percentage between 54 and 72% are satisfactory.

In conclusion, lumbar hemivertebra resection yields satisfactory immediate results and stability over time. We recommend this technique for hemivertebrae located at the lumbar level. For the thoracic area, however, and given that the neurological risk is greater, we prefer convex epiphysiodesis with a combined approach. Posterior osteosynthesis with compression yields better long-term results and enhances the effectiveness of epiphysiodesis.

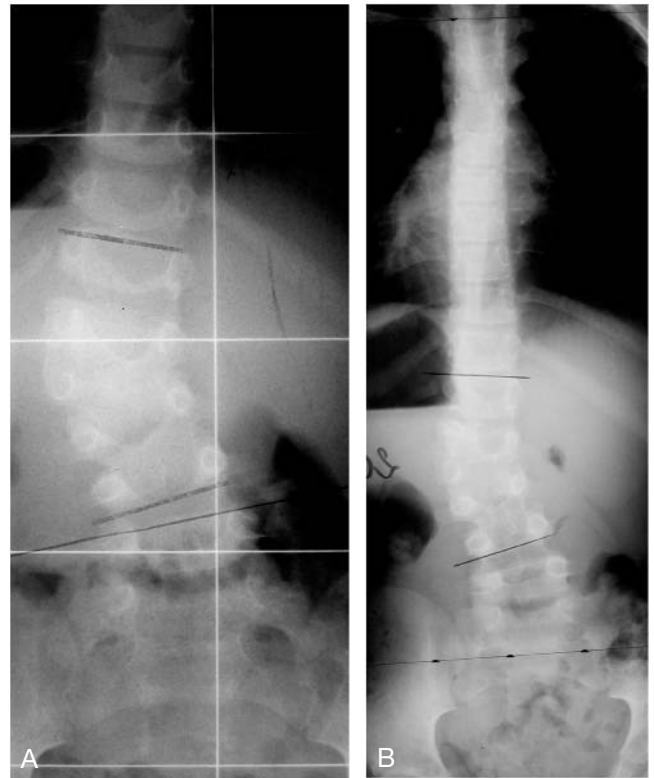


Figure 1 A: lumbar hemivertebra causing progressive scoliosis. Treatment through anterior and posterior convex epiphysiodesis. Surgical treatment at 3 years of age. B: result at age 12. Residual 20° angulation.

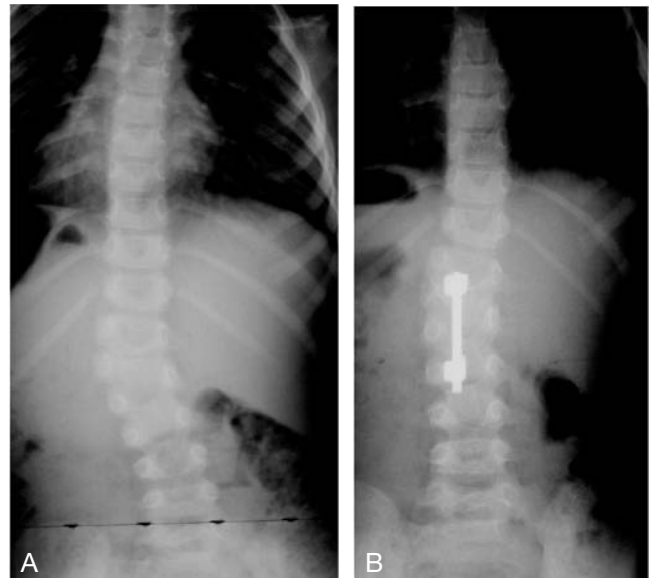


Figure 2 A: lumbar hemivertebra causing developmental scoliosis. Treatment by anterior and posterior convex epiphysiodesis with posterior instrumentation with compression at 3 years of age. B: result after 2 years follow-up.

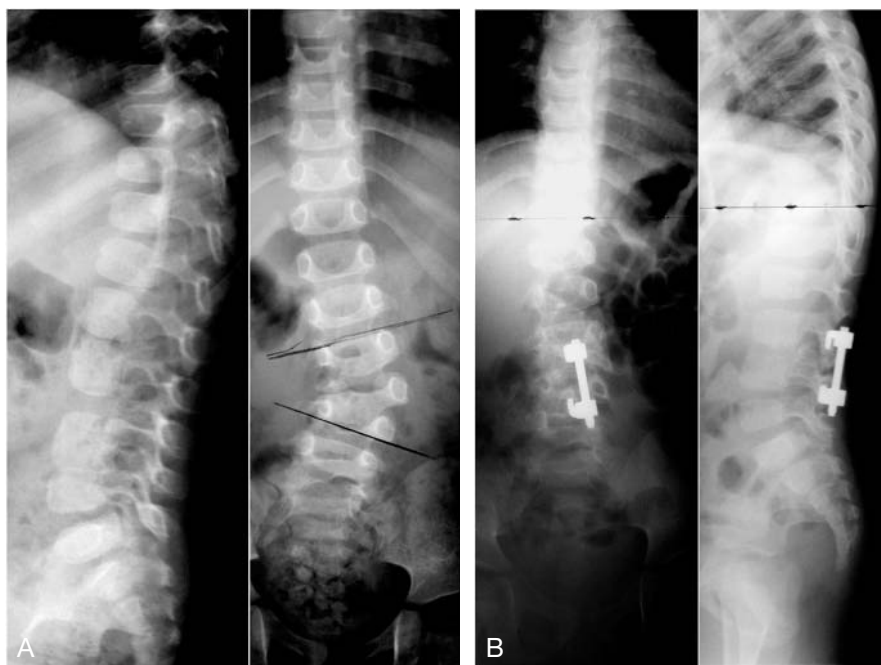


Figure 3 A: Lumbar hemivertebra treated by means of resection and instrumentation at 2 years of age (AP and Lat). B: result at age 5 (AP and Lat).

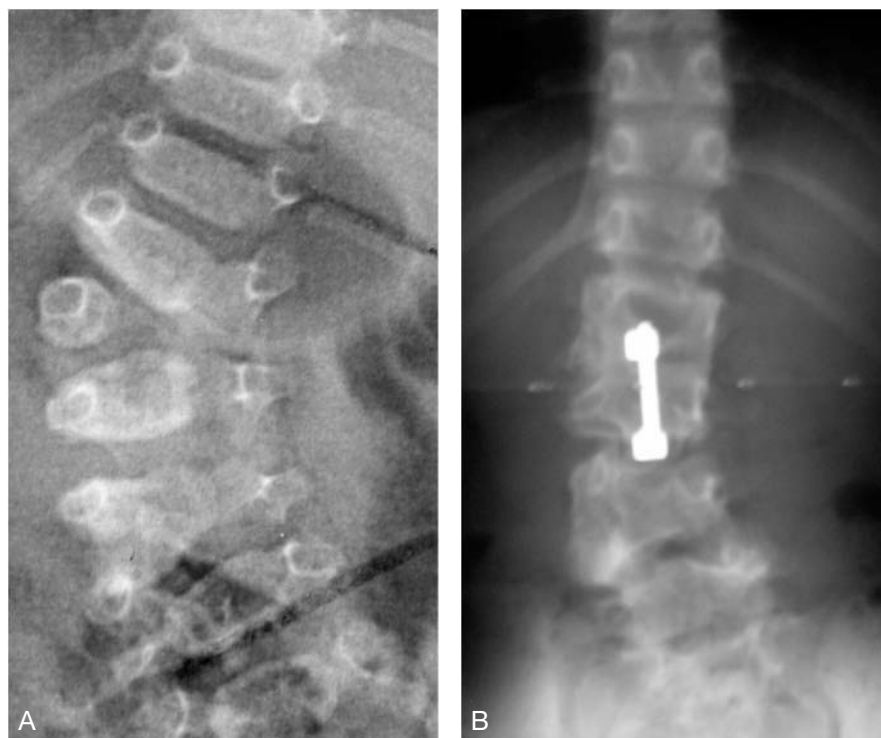


Figure 4 A: Lumbar hemivertebra treated with resection and instrumentation at 3 years of age. Note that there is an untreated segmented hemivertebra. B: result at age 7.

Conflict of interests

The authors have declared that they have no conflict of interests.

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