Short anterior instrumented fusion and posterior convex non-instrumented fusion of hemivertebra for congenital scoliosis in very young children

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Abstract A variety of treatments has been described in the literature for the treatment of HV. We report the results of early surgical anterior instrumented fusion with partial preservation of the HV and posterior non-instrumented fusion in the treatment of progressive congenital scoliosis in children below the age of six. Between 1996 and 2006, 31 consecutive patients with 33 lateral HV and progressive scoliosis underwent short segment fusions. Mean age at surgery was 2 years and 10 months. Mean follow-up period was 6.1 years. The major scoliotic curve improved from 41° preoperatively to 17° on follow-up. Preoperative segmental Cobb angle averaging 39° was corrected to 15° after surgery, being 15° at the last follow-up (62% of improvement). Compensatory cranial and caudal curves corrected by 47 and 45%, respectively. The angle of segmental kyphosis averaged 16° before surgery, 11° after surgery, and 11° at follow-up. There were two wound infections requiring surgical debridement, one intraoperative fracture of the vertebral body and one case lost correction due to implant failure. All went on to stable bony union. There were no neurological complications. Early diagnosis and early and aggressive surgical treatment are mandatory

for a successful treatment of congenital scoliosis and prevention of the development of secondary compensatory deformities. Anterior instrumentation is a safe and effective technique capable of transmitting a high amount of convex compression allowing short segment fusion, which is of great importance in the growing spine.

Keywords Hemivertebra · Congenital scoliosis · Fully segmented hemivertebra · Congenital spine deformity · Anterior instrumented fusion

Introduction

The natural history of congenital scoliosis has been well documented [6, 7, 11, 14, 21, 36, 39]. Foetal ultrasound scan has made antenatal diagnosis of hemivertebra (HV) possible [32]. Children with HV are, therefore, referred much earlier before severe local and compensatory curves develop. Surgical treatment of patients with congenital scoliosis carries a risk of neurological injury greater than that in patients with idiopathic spinal deformities [12, 19, 38]. Early treatment of progressive deformities helps in minimising the risks of surgery and allows better correction and prevents the development of structural and compensatory curves [7, 21, 38].

The current study describes the result of anterior partial resection and short-segmented instrumented fusion of hemivertebra in a series of 31 consecutive patients below the age of 6 years with progressive congenital scoliosis due to a lateral hemivertebra.

To date, there are no published reports that examine the safety and efficacy of anterior instrumented fusion for the treatment of congenital scoliosis due to HV in such a young patient population.

Study conducted at the Great Ormond Street Hospital for Children and the Royal National Orthopaedic Hospital, Stanmore, London, UK.

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Materials and methods

From 1996 to 2006, 31 consecutive patients with progressive congenital scoliosis due to 33 lateral hemivertebrae underwent short segment anterior instrumented fusion using a single solid rod construct with simultaneous convex non-instrumented posterior fusion corresponding to the levels of the anterior surgery.

Of the 31 patients in the study, 16 were girls and 15 boys. Their mean age at the time of surgery was 2 years and 10 months (range, 8 months to 5 years and 9 months). The mean follow-up period was 6.1 years (range, 2 to 11.3 years).

In total, 19 HV were located in the thoracolumbar junction (T10–L2), 11 in the lumbar spine (L3–L4) and 3 in the thoracic spine.

There were 27 children with single fully segmented HV, 2 semisegmented midlumbar HV, and 2 children had two ipsilateral fully segmented HV.

Only the vertebra above and below the hemivertebra were instrumented in 26 fusions. An additional level was included in seven cases: in three cases with thoracic HV due to the higher rigidity of the spine, in one case were the vertebral body fractured during screw insertion and in three cases with a higher magnitude curve.

Webb–Morley Spine System (Biomet-Merck Ltd, Bridgend, UK) was used on five patients and Downsize Synergy™ Spinal System, (Parsippany, NJ, USA) on 26 patients.

Children were evaluated clinically and by retrospective chart and radiographic review. Spinal cord anomalies were present in four cases (13%). Prior to deformity correction one patient underwent Arnold Chiari decompression and another patient untethering of a lipomyelocele. The remaining two patients had a cervical syrinx and a thoracolumbar syrinx. Other congenital vertebral anomalies were present in 9 patients (29%). Congenital heart disease was found in 6 cases (19%), genitourinary anomalies in 5 (15%) and gastrointestinal abnormalities in 3 (10%) patients. There was one Goldenhar syndrome.

Preoperative radiological imaging included anterior–posterior and lateral radiographs of the whole spine, MRI, echocardiogram and renal ultrasound.

Standing posteroanterior and lateral radiographs of the full spine were analysed and angles measured using the Cobb method. The angles of total main scoliosis, segmental scoliosis, compensatory cranial and caudal curves, segmental and total kyphosis, sagittal alignment of the thoracolumbar junction and lordosis were measured and recorded. The total angle of kyphosis was measured from T4 to T12. Lordosis was measured from L1 to L5. The thoracolumbar junction was measured from T9 to L2. MediCAD software (Hectec GmbH, Niederviehbach, Germany) was used to make all the measurements.

Curve correction was statistically analysed using paired Student's *t* test.

Surgical technique

All the patients are placed in the lateral decubitus position with the hemivertebra facing upward. Spinal cord monitoring is used for all patients. Care must be taken to keep the posterior spine exposed. A transthoracic or thoracoabdominal approach is chosen depending on the level of the HV.

Once the vertebral column is visualised, intraoperative radiographs are taken to confirm levels. Segmental vessels are preserved if possible. After exposure of the HV (Fig. 1), the disc material proximal and distal to the HV including the concave part of the disc is removed. Further resection of the HV is reserved until later stage to minimise bleeding.

Now a posterior midline incision is made. The posterior elements on the convexity are carefully exposed. In the first year to 2 years of life, cartilage forms a significant part of the posterior elements and correction of the deformity can be obtained without a formal posterior release. In older children, facetectomies of the HV are performed. Convex onlay morselised autogenous rib graft augmented with ProOsteon (Interpore Cross International, Irvine, CA) is applied after decortication of the laminae and spinous process corresponding to the levels of the anterior instrumentation.

If there is a failure of segmentation of the posterior elements, an osteotomy of the lamina can be done to further improve mobility at the segment.

The superior and inferior endplates of the HV are now removed to bleeding bone. The posterior body of the HV is partially decancellated. The body of the hemivertebra is

Fig. 1 Anterior–posterior and lateral sequence of the operative technique

resected to a stump corresponding to the pedicle location to achieve union with the adjacent endplates. Partial preservation of the body of the HV prevents nerve root impingement and minimises interference with local blood supply to the spinal cord.

At this point, anterior instrumentation begins. Bicortical transbody monoaxial screws are placed posteriorly and parallel to the endplates above and below the HV. Bicortical purchase of all screws is documented by manual palpation of the contralateral side of the spine. Washers were used in 6 of the 32 patients. A single straight rod is then engaged and the inferior screw is locked. Manual posterior pressure is applied to the spine to open the disc space and rib strut graft is inserted into the anterolateral portion of the concave disc space. Final position of the struts can be adjusted with a small bone punch. The intact posterior longitudinal ligament prevents over distraction during this manoeuvre.

Then careful intersegmental compression is applied. The strut graft prevents inducing kyphosis during anterior compression and serves as a fulcrum to obtain coronal correction. The disc space is further filled with morselized rib autograft.

Intraoperative radiographs are obtained before closure of the wound.

Patients were braced in a TLSO for 6 months postoperatively.

Results

The average operating time in this procedure was 133 min (range 80–210 min). The average blood loss 169 mL (range 50–550 mL), equivalent to a mean external blood volume loss of 10% (range 5–24%).

Segmental scoliosis (Table 1) was corrected from 39° (range 29°–56°) before surgery, to 15° (range 2°–27°) after surgery, and 15° (range 3°–32°) on the average, at the last follow-up. The major curve was 41° (range 29°–60°), 19° (range 4°–40°), and 17° (range 6°–35°), respectively. This represents a 62% of improvement for the segmental curve, and a 59% of improvement for the total main scoliosis curve at last follow-up.

About 47% of correction was obtained in the upper and 45% correction in the lower compensatory curves at the last follow-up.

Table 1 Correction of main and segmental scoliosis curves, compensatory curves, kyphosis and sagittal measurements

	Preoperative	Postoperative	Follow-up	Paired Student's test at follow-up
Total main (mayor) curve (°)	41	19	17	$P < 0.001$
Segmental curve (°)	39	15	15	$P < 0.001$
Cranial curve (°)	15	9	8	$P < 0.001$
Caudal curve (°)	20	12	11	$P < 0.001$
Segmental kyphosis (°)				
Thoracic HV	11	10	9	
Thoracolumbar HV	20	16	13	
Lumbar HV	10	6	8	
Total number HV	16	11	11	
Total kyphosis T4–T12 (°)				
Thoracic HV	17	15	35	
Thoracolumbar HV	18	21	25	
Lumbar HV	11	12	22	
Total number HV	15	17	25	
Thoracolumbar Junction T9–L2 (°) (+ Kyphosis) (– Lordosis)				
Thoracic HV	7	3	2	
Thoracolumbar HV	16	11	10	
Lumbar HV	4	4	–2	
Total number HV	12	9	6	
Total Lordosis L1–L5 (°)				
Thoracic HV	27	28	44	
Thoracolumbar HV	28	30	43	
Lumbar HV	19	21	33	
Total number HV	26	28	40	

In the sagittal plane, the segmental kyphosis averaged 16° (range 3° – 47°) before surgery, 11° (range 2° – 29°) postoperatively and 11° (range 3° – 29°) at last follow-up.

Follow-up focal kyphosis of four patients with a segmental kyphosis of more than 20° preoperatively (average 31° , range 22° – 46°) had a residual average segmental kyphosis of 24.5° (range 21° – 29°) on the latest follow-up. The total thoracic kyphosis T4–T12 averaged 25° (range 4° – 44°) at the latest follow-up. Lumbar lordosis L1–L5 was 40° (range 15° – 58°) at the latest follow-up.

There is a significant range of normal values with respect to the sagittal alignment of the spine [2, 4, 8, 10, 24, 27, 30, 33]. Accepted normal ranges of thoracic kyphosis and lumbar lordosis are 20° – 50° , and 20° – 60° , respectively. In our present study, seven patients were outside this range: five patients with thoracolumbar HV and one patient with a lumbar hemivertebra developed a thoracic hypokyphosis on follow-up, and one patient with a lumbar HV developed lumbar hypolordosis and thoracic hypokyphosis. The average segmental kyphosis for these patients was 17° .

Twenty-nine patients showed a sagittal plumb line with in normal range on follow-up (Figs. 2, 3).

Complications

One patient had an intraoperative fracture of the vertebral body during screw insertion, and required fusion of one additional segment.

Loosening of the screw rod interface (Webb–Morley Spine System, Biomet-Merck Ltd, Bridgend, UK) occurred in one patient. Intraoperative correction was lost in this patient, although went on to stable bony union without further intervention.

Fig. 2 Preoperative anterior–posterior and lateral radiograph in a 19-month-old girl with congenital scoliosis due to fully segmented hemivertebra

Fig. 3 Follow-up radiographs of the spine 7 years later showing satisfactory fusion and deformity correction in both coronal and sagittal planes

There were no neurological complications. Pseudoarthrosis was not observed.

Two patients had a deep posterior postoperative wound infection which required wound debridement, and resolved successfully with a course of antibiotics. No other reoperations were performed.

Discussion

Surgery is often necessary for progressive congenital scoliosis due to HV in the early growth cycle [7, 11, 13, 21, 25]. The severity of the curve, the type of HV, the location and the age of the patient are important factors in choosing the type of surgical intervention needed [3, 6, 22, 39].

Surgery is indicated with severe curvature at presentation, documented progression or predictable course. A corrective procedure to prevent the development of secondary structural curves and keeping the fused segment short to allow spinal growth is preferable [15, 16, 25, 28].

However, surgical treatment for congenital scoliosis carries a higher risk of paralysis [12, 19, 38]. Abnormal vascular supply to the spinal cord in congenital scoliosis has been postulated as one of the reasons for the higher incidence of neurological injury after surgical treatment for congenital deformities [5]. Normotensive anaesthesia and spinal cord monitoring are mandatory [18, 35].

An ideal treatment for congenital scoliosis due to HV is controversial [39] (Table 2).

Posterior spinal fusion alone with or without instrumentation carries a high risk of crank shafting, non-union and curve progression and most surgeons would reserve

Table 2 Summary of previous reports of surgical treatment for progressive congenital scoliosis

Year of publication	No. of patients	Mean follow-up (years)	Age at surgery (years)	Approach + instrument.	Scoliosis main curve		Segmental kyphosis		Complications		
					Preoperative (degree)	Follow-up (degree)	Improvement at FU (%)	Preoperative (degree)		Follow-up (degree)	
Winter et al. [37]	1984	290	6	11	Post. arthrodesis (127 with Harrington rod)	55	44	20	–	–	26% more than 10° loss of correction 20 pseudoarthrosis 2 paraplegias 1 extension of fusion 5 (38%) epiphysiodesis effect with improvement more than 5° 1 epiphysiodesis effect with improvement more than 5° 8 nerve root lesions (1 permanent) 3 pseudoarthrosis 3 Infections 6 extension of fusion Reoperation rate 35% 1 wire removal for pain 1 rod breakage
Winter et al. [36]	1988	13	6.6	3.6	Ant. + post. convex epiphysiodesis and hemiarthrodesis	46	31	32	–	–	
Dubousset et al. [9]	1993	10	2.4	4.1	Ant. + post. convex epiphysiodesis and hemiarthrodesis	46	37	20	–	–	
Holte et al. [13]	1995	37	6	12	Ant. + post. HV resection Post. hooks + rod in 28 patients (18 had previous arthrodesis)	54	35	35	–	–	
Callahan et al. [6]	1997	10	4.5	3.9	Ant. + post. resection 9 spinous process wiring 1 Harrington rod	40	16	60	–	–	
Lazar and Hall [16]	1999	11	2.3	1.5	Ant. + post. HV resection Post. hooks + rod	47	14	70	30	17	1 transient leg weakness 3 metal work removals Short follow-up Suture cutting through lamina
Klemme et al. [15]	2001	6	3.4	1.6	Ant. + post. HV resection Sublaminar suture	38	11	71	–	–	
Shono et al. [28]	2001	12	5.9	14	Post. resection Post. hooks + screws Long instrumentations	49	18	64	40	17	Long instrumentation Concave disc material not completely removed
Nakamura et al. [23]	2002	5	12.8	10	Post. resection	49	26	52	48	17	1 spinal decompensation
Ruf and Harms [25]	2003	25	3.5	3.3	Post. hooks + rod Post. resection Transpedic. screws 20 unisegmental instrumentations	45	13	72	22 ^a	8 ^a	3 implant failures requiring revision 2 pedicle fractures 2 curve progression at the operation site requiring revision 1 Infection and revision 5 pseudoarthrosis 6 curve progression Length of instrumentation unspecified 1 transient paraparesis
Bollini et al. [3]	2006	34	6	3.5	Ant. + post. HV resection Post. hooks + rod	40	27	33	20 ^a	14 ^a	

^a Segmental angle of kyphosis

this procedure for patients presenting late in the growth cycle with no decompensation of the spine [37].

Combined anterior and posterior hemiarthrodesis was designed to benefit of the growth potential on the concave side of the curve [9, 20, 31, 36]. Results are unpredictable but it remains a viable option on young children with low magnitude curves with enough discs above and below to prevent unbalancing of the spine.

An anterior or posterior HV excision lead to instability and it is necessary to apply compression instrumentation for adequate arthrodesis [15, 28]. A number of reports describe the difficulty of closing the wedge created by means of hooks, wiring techniques and sublaminar suture tape requiring frequently longer posterior fusions [3, 15, 16, 23].

HV usually produce acute angular deformity and posterior compression instrumentation can end up in the midline or concavity of the curve, increasing the deformity even further [3]. Spinal cord injury and nerve root compression when the pedicles of the vertebra above and below are approximated has been reported in HV resection [17, 29, 38].

A recent development has been the excision of HV through a posterior only approach [23, 25, 28]. The posterior-only approach for hemivertebra excision is a technically demanding procedure especially above the level of the cauda equina.

Anterior column visualisation and concave disc resection is difficult. Spinal cord and dural manipulation increases the risk for neurological injury [12].

Ruf et al. [25] reported good sagittal and coronal correction on 28 patients with posterior HV resection and pedicle screw instrumentation. He also reported difficulties achieving secure posterior instrumentation.

High intraoperative blood loss has been reported with posterior HV resection [23, 25]; thus the ability of maintaining normotensive anaesthesia in children where the incidence of congenital heart disease averages 26% [1], can be compromised.

Focal kyphosis can improve with further growth after single stage posterior resection of HV with transpedicular fixation [26]. This phenomenon has not been observed in series using a combined approach with posterior instrumentation [3]. In our series, we have not observed an anterior tethering effect with progressive focal kyphosis during further growth.

Our technique described earlier has a low intraoperative blood loss. No dural manipulation is needed and interference with local blood supply to the spinal cord is minimal. The partially preserved HV is the ideal vascularised bone graft to achieve solid fusion.

Short anterior instrumentation is able to transmit higher convex compressive forces to the vertebral body than a

Fig. 4 Preoperative anterior–posterior and lateral radiograph in a 26-month-old boy with congenital scoliosis due to two fully segmented ipsilateral hemivertebrae

Fig. 5 Follow-up radiographs of the spine 8.5 years later showing a balanced spine with satisfactory fusion in the coronal plane and residual thoracolumbar kyphosis

posterior tension band system like pedicle screws, providing good correction and stability [34]. This technique is particularly effective for correction of adjacent ipsilateral hemivertebrae (Figs. 4, 5)

Our technique has many advantages but has only limited ability to correct focal kyphosis. In cases with significant kyphosis, additional posterior instrumentation is required to close the posterior gap.

Conclusions

A range of procedures has been proposed for the treatment of congenital scoliosis. Trend is more towards the radical procedures in young children to correct this condition with significant morbidity. Our series of short anterior instrumented fusions with partial preservation of the HV show that this is a safe and effective technique with minimal blood loss, reliable fusion rate and low morbidity to treat congenital scoliosis in very young children.

It also provides excellent coronal correction and balanced growth in the coronal plane. In the sagittal plane, especially in the thoracolumbar junction persistent kyphosis during correction may require the addition of posterior instrumentation.

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