Results at skeletal maturity after double-approach hemivertebral resection

Résultats à maturité osseuse après résection d’hémivertèbres par double abord


* Département de Chirurgie Orthopédique Pédiatrique, Hôpital Timone Enfants, Université de Marseille, 264, rue Saint-Pierre, 13385 Marseille Cedex 5.
** Hôpital Universitaire Saint-Luc, Université Catholique de Louvain, Bruxelles, Belgique.

ABSTRACT

Purpose of the study
The natural history of congenital scoliosis or kyphoscoliosis resulting from a hemivertebra is well documented. The spinal deformation generally worsens in children with a free or semi-segmented hemivertebra situated in the thoracolumbar, lumbar, or lumbosacral region.

Material and methods
From 1982 to 1997, 15 pediatric patients with 15 hemivertebrae causing progressive scoliosis or kyphoscoliosis underwent hemivertebral resection via a double posterior and anterior approach associated with convex fusion. Mean age at surgery was 4.4 years. The 15 patients were reviewed at bone maturity (Risser 4 or 5) to assess outcome.

Results
Genitourinary tract anomalies were associated in five of the 15 patients and intrathecal anomalies in two. Mean follow-up was 12.1 years. Segmental scoliosis was 30.2° preoperatively, 12.3° postoperatively, and 8.9° at last follow-up. The values for total scoliosis were 30.5°, 12.3°, and 12.6°, respectively. This was a 70.5% improvement at last follow-up for segmental scoliosis and 58.7% for total scoliosis. The real trunk imbalance improved from 31% preoperatively to 9% at last follow-up. All these differences were significant.

Discussion
The main objective of surgical treatment for congenital scoliosis due to hemivertebra is to prevent the development of severe deformation that would require a dangerous and difficult procedure to achieve correction. Resection of the hemivertebra via a double approach is the ideal method for early correction. The procedure not only corrects the spinal deformation but also stabilizes the curve.
but also prevents later deterioration. Correction should be performed as early as possible. The result is sustained over time if there is no other associated spinal anomaly. The procedure is safe and the technique well controlled in experienced hands. Patients need to wear a brace for only 6 months after resection of the hemivertebra.

**Key words:** Congenital scoliosis, hemivertebra, hemivertebra resection, double approach, convex fusion.

**INTRODUCTION**

The natural history of congenital scoliosis and kyphoscoliosis has already been documented [Nasca et al. (1), McMaster et al. (2), McMaster and Singh (3)]. The degree of scoliosis produced by a hemivertebra (HV) depends on the type, the site, the number of HVs, and the patient’s age. Progression varies depending on the HV location. The thoracolumbar and lumbosacral junctions are transition zones between the lumbar column and a less mobile or immobile segment (thoracic column or sacrum). HVs in these two transition zones evolve toward trunk imbalance. Moreover, at the thoracolumbar junction, the HV rapidly leads to vertebral rotation. At the thoracolumbar junction and at the level of the lumbar column, the sagittal component is important, with a significant risk of kyphoscoliosis. On the contrary, a lumbosacral HV presents no risk of kyphotic deformation.

A single free HV (completely segmented) can lead to deterioration of the curve at a rate of 2-3.5° per year if it is located at the thoracolumbar junction, 1.7° per year between L2 and L4, and 1.5° per year at the lumbosacral junction [McMaster et al. (2)]. Surgery is indicated in cases of regular deterioration of the spinal curvature or in cases of spinal imbalance that can worsen with no changes in the angle.

This article reports the progression of a series of 15 HV resection cases followed to skeletal maturity. The surgical technique involved a double approach resection with convex fusion.

**MATERIAL AND METHODS**

**Patient series**

From March 1982 to December 1997, 15 hemivertebrae (HVs) were excised in 15 patients (seven girls and eight boys). The mean age at surgery was 4.4 years (range, 3 months to 9.8 years). All spinal deformations were scoliosis or kyphoscoliosis caused by a HV with obvious deformation of the spinal curve. The 15 patients grew to skeletal maturity (Risser 4 or 5) and were assessed retrospectively.

**Patient evaluation**

Preoperative radiographic workup included posteroanterior and lateral standing full x-rays. All but five patients were evaluated by preoperative vertebral spinal MRI to exclude intrathecal anomaly and to study the segmentation of the HV and the growth zones. The five others had preoperative myelography (at the beginning of the series). Preoperative renal ultrasound was also performed in all the children to search for possible associated congenital renal or genitourinary anomalies. A careful cardiac assessment was also done looking for cardiac disorders.

**Anatomic and hemivertebra data (table I)**

Each complete vertebra with a thoracic anatomy (thoracic transverse process and transverse-costal joints) or lumbar anatomy (lumbar transverse processes) was examined. A vertebra with a thoracic transverse process on one side and a lumbar transverse process on the other side was considered a thoracic vertebral column and its side. For example, a left T13-L1 HV corresponded to a HV located between a thirteenth thoracic vertebra and the first lumbar vertebra, located on the left side.

Five HVs were located at the thoracolumbar junction (T10 to L2), five others were lumbar (between L2 and L5), and the last five were lumbosacral. Six HVs were located on the right side and the nine others on the left. Seven HVs were completely segmented and eight were semisegmented. Other spinal bone abnormalities were associated in three cases.

**Surgery (table II)**

All operations were performed by the same senior surgeon (GB).

In the 15 cases, a complete resection of the HV was done by double approach. The anterior approach was performed first in the six first cases of the series, followed by the posterior approach in a single operation (two patients) or after a lapse of 1 week (four patients). For the next nine cases, the posterior approach was done first with the patient in the ventral decubitus position; then the patient was repositioned in lateral decubitus for the anterior approach (in the same operation for eight patients or after 7 months for one patient).

In the posterior approach, only the posterior elements of the convexity were exposed and the posterior part of the HV was resected to the pedicle. The implantation sites of the laminar hooks were prepared at the adjacent proximal and distal laminae. The anterior approach was a thoracophrenolumbotomy on the 10th rib for the five thoracolumbar HVs and a lumbotomy for the ten lumbar or lumbosacral HVs. After complete resection of the HV body, convex posterior compression was applied using the posterior approach.
For two patients, the convex fusion was not instrumented. For the 13 others, posterior instrumentation consisted in a baby-CD (fig. 1) in four cases, a mini-Harrington rod in four cases, and a steel-wire cerclage (fig. 2) in three cases. In one case (case no. 2), a double baby-CD was placed to achieve convex compression and concave distraction. In the last case (case no. 1), the excision of the thoracolumbar HV was associated with anterior and posterior convex fusion of a thoracic HV and double mini-Harrington rods were placed in convex compression at two levels (T9-L1 and T5-T6).

An anterior and posterior autogenous graft was performed using the resected rib or a fibular graft posteriorly and anteriorly. Posterior compression provided a nearly complete closure of the space left by the HV resection from a posterior position, whereas from the anterior aspect, this space was closed by approximately 50%. A blood collector was used during the operation and the blood recycled was returned at the end of the operation. Evoked potentials and intraoperative wakening were not monitored.

A heterologous blood transfusion was necessary for only two patients in the perioperative period. A blood workup was done systematically the day after the operation to check the hemoglobin rate. The child wore a brace molded before the operation for 6 months to protect the convex fusion. The technical details are summarized in table II.

### Table I. – Clinical and anatomical characteristics of the 15 patients.

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Sex</th>
<th>HV location</th>
<th>Side</th>
<th>Segmentation</th>
<th>Fusion at</th>
<th>Other hemivertebra or spinal abnormality</th>
<th>Associated congenital abnormality</th>
<th>Associated intrathecal abnormality</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>T9-L1</td>
<td>L</td>
<td>SS</td>
<td>L1</td>
<td>T1-T2 HV L nonsegmented, T5-T6 HV R free op, T3 and T4 binucleated</td>
<td>One absent kidney</td>
<td>0</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>T13-L1</td>
<td>R</td>
<td>SS</td>
<td>L1</td>
<td>0</td>
<td>One absent kidney</td>
<td>0</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>L1-L2</td>
<td>R</td>
<td>SS</td>
<td>L2</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>4</td>
<td>F</td>
<td>L1-L2</td>
<td>L</td>
<td>SS</td>
<td>L2</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>T11-L1</td>
<td>R</td>
<td>SS</td>
<td>L1</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>6</td>
<td>M</td>
<td>L2-L3</td>
<td>L</td>
<td>Free</td>
<td></td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>7</td>
<td>M</td>
<td>L2-L3</td>
<td>L</td>
<td>SS</td>
<td>L2</td>
<td>0</td>
<td>One absent kidney</td>
<td>0</td>
</tr>
<tr>
<td>8</td>
<td>M</td>
<td>L2-L3</td>
<td>L</td>
<td>Free</td>
<td></td>
<td>Sacralization of L5</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>9</td>
<td>F</td>
<td>L4-L5</td>
<td>L</td>
<td>SS</td>
<td>L4</td>
<td>0</td>
<td>Exstrophy of the bladder, imperforate anus</td>
<td>0</td>
</tr>
<tr>
<td>10</td>
<td>F</td>
<td>L4-L5</td>
<td>L</td>
<td>Free</td>
<td></td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>11</td>
<td>M</td>
<td>L5-S1</td>
<td>L</td>
<td>Free</td>
<td></td>
<td>0</td>
<td>Short tibia and calcaneovalgus foot on the right, club foot left</td>
<td>Lower spinal cord with adhering lipoma</td>
</tr>
<tr>
<td>12</td>
<td>F</td>
<td>L5-S1</td>
<td>R</td>
<td>Free</td>
<td></td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>13</td>
<td>M</td>
<td>L5-S1</td>
<td>L</td>
<td>Free</td>
<td></td>
<td>Vertebral block T3-T4-T5-T6, costal synostosis T4-T5</td>
<td>Operated duodenal stenosis; bilateral femoral anteverision</td>
<td>0</td>
</tr>
<tr>
<td>14</td>
<td>M</td>
<td>L5-S1</td>
<td>R</td>
<td>SS</td>
<td>S1</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>15</td>
<td>F</td>
<td>L4-S1</td>
<td>R</td>
<td>Free</td>
<td></td>
<td>0</td>
<td>Anteposed anus, operated vesicoureteral reflux</td>
<td>Myelocystocele, Chiari malformation</td>
</tr>
</tbody>
</table>

HV, hemivertebra; M, male; F, female; L, left; R, right; SS, semi-segmented.

### X-ray assessment

Radiographic analysis was done on standing posterior and lateral entire spine x-rays. Preoperative x-rays, those taken immediately before hospital discharge (with the brace), and at the most recent follow-up visit were examined.
<table>
<thead>
<tr>
<th>Case no.</th>
<th>HV</th>
<th>Other operated HV</th>
<th>Age (years)</th>
<th>Approach</th>
<th>Instrumentation</th>
<th>Additional procedures, age at time of procedure</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>T9-L1</td>
<td>Convex circumferential arthrodesis T5-T6 HV without resection (10 days later)</td>
<td>3.8</td>
<td>Anterior for anterior resection of T9-L1 HV, then posterior (1 week later) for posterior resection of T9-L1 HV and posterior arthrodesis of T6-T6 HV with simultaneous anterior arthrodesis of T5-T6 HV</td>
<td>Double mini-Harrington rod (convex compression at T9-L1 and T5-T6 levels)</td>
<td>0</td>
</tr>
<tr>
<td>2</td>
<td>T13-L1</td>
<td>0</td>
<td>9.8</td>
<td>Posterior, then anterior</td>
<td>Double baby-CD (convex compression and concave distraction)</td>
<td>Ablation of material (at 11 years of age)</td>
</tr>
<tr>
<td>3</td>
<td>L1-L2</td>
<td>0</td>
<td>4.9</td>
<td>Posterior, then anterior</td>
<td>Baby-CD (convex compression)</td>
<td>0</td>
</tr>
<tr>
<td>4</td>
<td>L1-L2</td>
<td>0</td>
<td>7.0</td>
<td>Posterior, then anterior</td>
<td>Baby-CD (convex compression)</td>
<td>Material removed (at 10 years of age)</td>
</tr>
<tr>
<td>5</td>
<td>T11-L1</td>
<td>0</td>
<td>4.7</td>
<td>Posterior, then anterior (1 week later)</td>
<td>Mini-Harrington rod (convex compression)</td>
<td>Reoperation for abdominal wall hernia and removal of implants (1 month later), vertebral osteotomy and permanent arthrodesis with CD for focal kyphosis (at 14.4 years of age)</td>
</tr>
<tr>
<td>6</td>
<td>L2-L3</td>
<td>0</td>
<td>3.4</td>
<td>Posterior, then anterior</td>
<td>Mini-Harrington rod (convex compression)</td>
<td>0</td>
</tr>
<tr>
<td>7</td>
<td>L2-L3</td>
<td>0</td>
<td>1.1</td>
<td>Posterior, then anterior</td>
<td>Steel-wire cerclage (convex compression)</td>
<td>Posterior arthrodesis with mini-Harrington rod for nonunion with evolutive kyphosis (at 4 years of age)</td>
</tr>
<tr>
<td>8</td>
<td>L2-L3</td>
<td>0</td>
<td>1.0</td>
<td>Anterior and posterior (1 week later)</td>
<td>Steel-wire cerclage (convex compression)</td>
<td>0</td>
</tr>
<tr>
<td>9</td>
<td>L4-L5</td>
<td>0</td>
<td>1.7</td>
<td>Posterior, then anterior</td>
<td>Steel-wire cerclage (convex compression)</td>
<td>0</td>
</tr>
<tr>
<td>10</td>
<td>L5-S1</td>
<td>0</td>
<td>1.6</td>
<td>Anterior, then posterior (1 week later)</td>
<td>Baby-CD (convex compression)</td>
<td>0</td>
</tr>
<tr>
<td>11</td>
<td>L5-S1</td>
<td>0</td>
<td>5.8</td>
<td>Anterior, then posterior</td>
<td>Mini-Harrington rod (convex compression)</td>
<td>0</td>
</tr>
<tr>
<td>12</td>
<td>L5-S1</td>
<td>0</td>
<td>3.6</td>
<td>Anterior, then posterior (1 week later)</td>
<td>No instrumentation</td>
<td>0</td>
</tr>
<tr>
<td>13</td>
<td>L5-S1</td>
<td>0</td>
<td>9.3</td>
<td>Posterior, then anterior</td>
<td>Baby-CD (convex compression)</td>
<td>0</td>
</tr>
<tr>
<td>14</td>
<td>L5-S1</td>
<td>0</td>
<td>9.3</td>
<td>Posterior, then anterior (7 months later)</td>
<td>Mini-Harrington rod (convex compression)</td>
<td>Convex arthrodesis with mini-Harrington rod for nonunion (at 2.2 years of age), material removed (at 9 years)</td>
</tr>
<tr>
<td>15</td>
<td>L4-S1</td>
<td>0</td>
<td>0.3</td>
<td>Posterior, then anterior</td>
<td>Mini-Harrington rod (convex compression)</td>
<td>0</td>
</tr>
</tbody>
</table>
All the x-rays were evaluated by an independent observer (PLD) from another hospital. All angle measurements were taken using the Cobb method.

The different curvatures measured in the coronal plane were: segmental scoliosis, total scoliosis, the compensatory cranial curve, and the compensatory caudal curve (fig. 3). Segmental scoliosis was measured between the two vertebrae immediately adjacent to the HV, whereas total scoliosis was measured between the two most inclined vertebrae. Trunk imbalance was evaluated in two ways on the frontal image: gravity trunk imbalance (GTI) and real trunk imbalance (RTI) (fig. 3). Gravity imbalance is the distance between a vertical line (plumb line) descending from the middle of the T1 body and the middle of the sacrum (to the S2 vertebra). Real trunk imbalance is the distance between the middle of the sacrum and a line descending from the middle of the T1 body and perpendicular to the bi-iliac line (tangential to the two iliac crests). The width of the pelvis was measured as the distance between two points on the iliac crests tangential to the bi-iliac line (fig. 3). The GTI and RTI were related to the pelvis width and expressed as a percentage to prevent errors related to radiological enlargements and the growth of the child’s pelvis. The RTI is more precise than the GTI in cases of pelvic obliquity.

On the lateral incidence, segmental kyphosis (between the two vertebrae adjacent to the HV) were measured. For thoracolumbar HVs, overall kyphosis was measured between T3 and T12). The upper thoracic spine was excluded given that T1 and T2 are often difficult to visualize clearly on the lateral incidence. The kyphosis curvatures were expressed as positive angle values, whereas the lordosis curvatures were expressed by negative values. For lumbar or lumbosacral HVs, overall lordosis was measured between the upper endplate of L1 and the upper endplate of S1.

In three cases that required reoperation (one for progression of the curvature despite HV resection and two for non-union), the x-rays taken just before the reoperation were examined to measure the last follow-up angles.

Statistical analysis

RESULTS

Clinical history of the 15 patients at skeletal maturity

For one adopted child (case no. 5), perinatal data was not available. In the 14 other cases, the mean birth was 2815 g (range, 1730-3500 g). Only one child had a low birth weight (< 2500 g). It was also the only case of a twin pregnancy and cesarian delivery. For the 13 others, delivery was unproblematic and the babies were in cephalic presentation. None was premature (born before 37 weeks of pregnancy). Six children were firstborns, six were second in sibling rank, two others were third and fourth, respectively. Congenital anomalies were associated in seven children out of 15 (table I). Genitourinary defects were found in five patients: three cases of one absent kidney, one case of exstrophy of the bladder and one case of vesicoureteral reflux. None of the patients had cardiac anomalies. One child had duodenal stenosis and femoral anteversion, which required surgical correction. Intrathecal anomalies were found in two children. The first had a lipoma adhered to the spine. This child had a neurological bladder and a neurological deficit of the right lower limb (no active muscle below the knee). He had a short tibia and a calcaneovalgus foot on the right and a club foot on the left. The second had a myelocystocele with imperforate anus and a Chiari malformation. He had a neurological deficit of the left lower limb of

L5-S1 topography and urinary incontinence. The other children had no neurological deficit and normal psychomotor development.

Family antecedents were found in only two patients: Sturge-Weber syndrome in one patient’s brother and sacral agenesis in a maternal uncle of another.

**Surgery**

Surgery lasted a mean of 295 min (range, 240-330 min) from skin incision to the end of skin closure. Blood loss expressed in the difference of the hemoglobin rate between the day before and the day after operation was 2.9 g/dl (range, 1.4-4.2 g/dl), excluding two patients transfused with heterologous blood.

**Postoperative results** *(table III)*

Mean follow-up was 12.1 years (range, 6.9-17.7 years). The 15 patients were at skeletal maturity on the Risser scale: Risser 4 (eight cases) or Risser 5 (seven cases).

On the coronal plane, mean segmental scoliosis was $30.2^\circ$ before surgery, $12.3^\circ$ after surgery, and $8.9^\circ$ at the last follow-up. Mean total scoliosis was $30.5^\circ$, $13.6^\circ$, and $12.6^\circ$ respectively. This reflects an improvement of $70.5\%$ for segmental scoliosis and $58.7\%$ for total scoliosis, and these differences were statistically significant. Gravity trunk imbalance improved from $10\%$ before surgery to $7\%$ at the last follow-up, but this difference was not significant.
RESULTS AT SKELETAL MATURITY AFTER DOUBLE-APPROACH HEMIVERTEBRAL RESECTION

Real trunk imbalance improved significantly, from 21% to 9%.

On the sagittal plane, mean segmental kyphosis was not significantly modified. In the five cases of thoracolumbar HV, total kyphosis at the last follow-up was within the normal values according to Boseker et al. (4) (20°-50°) in four cases. In a single case, this kyphosis was slightly exaggerated at 54°. For the ten cases of lumbar or lumbosacral HV, total lordosis at the last follow-up was within normal values according to the Scoliosis Research Society criteria (-31 to -79°) in eight cases [Knight et al. (5)]. Lordosis was reduced in two cases: -12° and -8°.

The immediate postoperative period showed no complications for all 15 patients. No neurological, infectious, or respiratory complication occurred.

Delayed complications occurred in three patients. An incisional hernia on the thoracophrenolumbotomy scar required reoperation at 1 month after surgery (case no. 5). Two cases of nonunion were found: case no. 7 was treated with posterior arthrodesis with a mini-Harrington rod (3 years later) and case no. 15 was treated with double approach convex arthrodesis (2 years later).

In this series, intrathecal abnormalities were found in two out of 15 patients (13%). McMaster (7) reported intrathecal abnormalities in 18% of a series of 251 patients evaluated using myelography. With MRI, Bradford et al. (8) found intraspinal abnormalities in 38% of their 42 patients, Suh et al. (9) in 31% of their 41 patients, Prahinski et al. (10) in 30% of their 30 patients, and Basu et al. (11) in 37% of their 126 patients. Basu et al. (11) also showed that the HV

### Table III. – Scoliosis, kyphosis, and imbalance in preoperative and postoperative examinations, and at last follow-up.

<table>
<thead>
<tr>
<th></th>
<th>Preoperative</th>
<th>Postoperative</th>
<th>Improvement (paired Student t test)</th>
<th>Last follow-up</th>
<th>Improvement (paired Student t test)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Scoliosis</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Segmental curve (°)</td>
<td>30.2° (range, 13-42)</td>
<td>12.3° (range, 2-28)</td>
<td>59.27% p &lt; 0.001</td>
<td>8.9° (range, 0-22)</td>
<td>70.5% p &lt; 0.001</td>
</tr>
<tr>
<td>Total curve (°)</td>
<td>30.5° (range, 13-42)</td>
<td>13.6° (range, 2-28)</td>
<td>55.41% p &lt; 0.001</td>
<td>12.6° (range, 0-37)</td>
<td>58.7% p &lt; 0.001</td>
</tr>
<tr>
<td>Compensatory cranial curve (°)</td>
<td>15.5° (range, 4-35)</td>
<td>11.4° (range, 2-34)</td>
<td>26.45% p = 0.04</td>
<td>8.9° (range, 0-26)</td>
<td>42.6% p &lt; 0.03</td>
</tr>
<tr>
<td>Compensatory caudal curve (°)</td>
<td>6.9° (range, 0-24)</td>
<td>3.6° (range, 0-15)</td>
<td>47.83% p = 0.02</td>
<td>4.4° (range, 0-43)</td>
<td>36.2% NS</td>
</tr>
<tr>
<td>Gravity imbalance (%)</td>
<td>10% (range, 0-25)</td>
<td>11% (range, 0-35)</td>
<td>-10.00% NS</td>
<td>7% (range, 1-15)</td>
<td>30.0% NS</td>
</tr>
<tr>
<td>Real trunk imbalance (%)</td>
<td>21% (range, 3-49)</td>
<td>17% (range, 2-43)</td>
<td>19.05% NS</td>
<td>9% (range, 1-39)</td>
<td>57.1% p &lt; 0.001</td>
</tr>
<tr>
<td>Segmental kyphosis (°)</td>
<td>+0.6° (range, -18 to +34)</td>
<td>+1.9° (range, -27 to +22)</td>
<td>NS</td>
<td>+1.8° (range, -28 to +40)</td>
<td>NS</td>
</tr>
</tbody>
</table>

DISCUSSION

A hemivertebra is a congenital vertebral abnormality caused by a formation defect. Three major types of spinal deformations can result: congenital scoliosis (the most frequent), congenital kyphosis, or congenital lordosis (the least frequent).

Curvature progression caused by a single HV is extremely difficult to predict: this type of curvature can progress rapidly, slowly, or not at all. The degree of scoliosis produced by an HV depends on four factors: the type, the site, the number of HVs, and the patient’s age [McMaster et David (6)]. To determine whether a curvature is progressing, precise and regular follow-up, both clinical and radiological, is necessary. The main objective of treatment is to prevent the development of a severe deformation requiring substantial and dangerous correction. Curvature progression is fastest during the peak of pubertal growth and stops only at skeletal maturity [McMaster et Singh (3)]. Spontaneous neurological deterioration resulting from spinal cord compression can occur in cases of congenital kyphoscoliosis (in general, during the peak of pubertal growth at a mean age of 13.7 years) [McMaster et Singh (3)].
located higher in the spine (cervical or thoracic) had an increased risk of associated intraspinal anomalies. Our lower rate of intrathecal anomalies could be explained by the distal location of HVs (thoracolumbar, lumbar, and lumbosacral) in our series.

With ultrasound, we found genitourinary defects in five patients out of 15 (33%). This rate is similar to that found in the literature. With intravenous urography (IVU), MacEwen et al. (12) found genitourinary abnormalities in 20% of the patients with congenital scoliosis. With IVU and ultrasound, Guerrero et al. (13) found an incidence of genitourinary abnormalities in 34% of their patients. With ultrasound, Basu et al. (11) found a 21% incidence of genitourinary abnormalities. None of the patients in our series suffered from cardiac abnormalities. Few publications report the incidence of congenital cardiac disease associated with congenital scoliosis. Using echocardiography, Basu et al. (11) found a 26% incidence of congenital cardiac pathology in patients with congenital spinal deformation. Beals et al. (14) found 12%.

Our series had one case of twin pregnancy, but the co-twin had no vertebral abnormality. A positive family history of congenital spinal deformation was found in only one of our patients (sacral agenesis in a maternal uncle). In a series of 1250 patients managed in their center, Winter et al. (15) found a positive family history in only 1% of patients. Wynne-Davies (16), in a review of the families of 337 patients, concluded by saying that an isolated anomaly such as HV was a genetically sporadic lesion that led to no risk of similar lesions in later generations. Twin studies have generally shown that if one twin had an abnormality, the other did not, even in identical twins [Winter et al. (15)]. Only one case of congenital vertebral abnormality in identical twins has been reported in the literature [Sturm et al. (17)].

None of the children in our series was premature (<37 weeks of gestation) and only one (7%) had low birth weight (<2500 g). Rates of prematurity and low birth weight reported by the national perinatal statistics in France in 1995 [Blondel et al. (18)] are 5.9% and 6.2%, respectively.

Since a high percentage of congenital scoliosis curvatures are progressive and do not respond to brace treatment, surgery is generally the rule. The three types of basic operation are in situ arthrodesis, convex epiphysiodesis, and HV resection.

Isolated posterior arthrodesis with or without instrumentation is not recommended for young children because the correction is limited and the crankshaft phenomenon occurs in 15% of patients and in 36% when surgery is done before the age of 4 years [Kesling et al. (19)]. In a series of 31 patients treated with posterior arthrodesis (instrumented with Harrington rods for 18 and noninstrumented in 13), Hall et al. (20) reported a reduction of the mean curvature from 62° to 40° and from 43° to 38°, respectively.

Combined anterior and posterior arthrodesis adds the potential benefit of a greater correction and a correction of the sagittal plane because the discal excision provides greater mobility of the segments. This also reduces the risk of nonunion and prevents the crankshaft phenomenon by removing growth plates. Cheung et al. (21) reported on the long-term follow-up (10.8 years) of a series of six thoracolumbar HVs treated with combined approach convex arthrodesis with subcutaneous instrumented distraction across the concavity. They reported a mean improvement of 41% from 49° preoperatively to 29° at the last follow-up.

Arresting convex growth or convex epiphysiodesis is appropriate only for patients with a potential for growth remaining on the concave side. This type of procedure should therefore be done early (before the age of 5 years) and before the curvature has progressed to 50 or 60° [Winter et al. (22), Uzumcugil et al. (23)]. Taking into account the 123 patients treated with this technique reported in the literature, the technique provides improvement (epiphysiodesis effect) in 48% of cases (20%-77%), no change (arthrodesis effect) in 40% (17%-70%), and progression in 12% (0%-21%) [Winter et al. (22), Uzumcugil et al. (23), Andrew et Piggott (24), Keller et al. (25), Thompson et al. (26), Kieffer et Duboussset (27), Walhout et al. (28)]. Several reasons can be given for the high rate of nonimprovement, even progression: the difficulty of locating the exact summit of the convexity and the solidity of the anterior epiphysiodesis compared to the posterior epiphysiodesis explaining the frequent progression toward kyphoscoliosis. The number of fused vertebrae is also higher with this technique compared to HV resection. Marks et al. (29) concluded that double anterior and posterior convex epiphysiodesis alone does not prevent the progression of the deformation in infantile idiopathic scoliosis and that adding posterior instrumentation can slow or arrest the progression of the deformation but not improve it.

Hemivertebra excision was performed for the first time in 1921 by Royle (30) in Australia. Many authors have reported series of HV resections by successive or simultaneous anterior and posterior approaches or by the posterior approach alone [Leatherman and Dickson (31), Slabaugh et al. (32), Bradford and Boachie-Adjei (33), King and Lowery (34), Leong et al. (35), Holte et al. (36), Callahan et al. (37), Lazar and Hall (38), Klemme et al. (39), Shono et al. (40), Ruf and Harms (41, 42), Nakamura et al. (43)]. The improvement rates in this studies vary from 24.3% to 71.1%.

In our series, HV excision resulted in a high level of immediate correction of total scoliosis (55.4% improvement) and the result was stable over time (58.7% improvement after a mean follow-up of 12.1 years). One case of nonunion can undoubtedly be explained by insufficient instrumentation (steel wire cerclage) used at the beginning of the series. The material used today for instrumentation is the baby-CD designed by our senior author (GB). This posterior instru-
mentation provides convex compression from two sub-and
infralaminar hooks and a rod. This material is very useful
but in the thoracolumbar region, it is only effective for mod-
erate angles. Indeed, when the angle is large with severe
vertebral rotation in the thoracolumbar region, the use of
posterior instrumentation alone is not recommended
because the hooks are close to the spinous process (because
the laminae are not very wide) and are in fact in the concav-
ity because of the severe rotation of the vertebrae. In this
case, it is preferable to use an anterior baby-CD with screws
fixed in the vertebral body and the same rod as for posterior
instrumentation.

In the sagittal plane, the final total kyphosis value was
within normal values (except for one case) for the thoraco-
lumbar HVs and the final lordosis value was within normal
values (except for two cases) for the lumbar and lumbosac-
rnal HVs. The anterior fibular graft is important to prevent
progression of the kyphosis in the postoperative period.

CONCLUSION

Double approach hemivertebra resection is an ideal sur-
gical procedure for early correction of scoliosis. The treat-
ment not only corrects the deformation, but also prevents
later deterioration that the HV would have brought about.
The correction should be done very early, even in the very
small child. The result is maintained over time if no other
associated spinal abnormality is present. The procedure is
safe. Wearing a brace is only necessary for a period of
6 months after the operation.

References

1. NASCA RJ, STILLING FH 3RD, STELL HH : Progression of
congenital scoliosis due to hemivertebrae and hemivertebrae
2. MCMASTER MJ, OHTSUKA K : The natural history of con-
genital scoliosis. A study of two hundred and fifty-one
3. MCMASTER MJ, SINGH H : Natural history of congenital
kyphosis and kyphoscoliosis. A study of one hundred and
twelve patients. J Bone Joint Surg (Am), 1999, 81,
1367-1383.
4. KNIGHT RQ, JACKSON RP, KILLIAN RT, STANLEY EA :
Determination of “normal” thoracic kyphosis; a roentgenographic
study of 121 “normal” children. J Pediatr Orthop, 2000, 20,
796-798.
5. MCMASTER MJ, DAVID CV : Hemivertebra as a cause of
scoliosis. A study of 104 patients. J Bone Joint Surg (Br),
1986, 68, 588-595.
6. MCMASTER MJ, POLLY DW JR, MCHALE KA, ELLENBOGEN RG :
Occult intraspinal anomalies in congenital scoliosis. J Pediatr
7. MCMASTER MJ : Occult intraspinal anomalies and congeni-
8. BRADFORD DS, HEITHOFF KB, COHEN M : Intraspinal
abnormalities and congenital spine deformities: a radiographic
9. SUH SW, SARWARK JF, VORA A, HUANG BK : Evaluating
congenital spine deformities for intraspinal anomalies with
magnetic resonance imaging. J Pediatr Orthop, 2001, 21,
525-531.
10. PRAHNISKI JR, POLLY DW JR, MCHALE KA, ELLENBOGEN RG :
Occult intraspinal anomalies in congenital scoliosis. J Pediatr
11. BASU PS, ELSEBAIE H, NOORDEEN MH : Congenital spinal
deformity: a comprehensive assessment at presentation.
12. MACEWEN GD, WINTER RB, HARDY JH : Evaluation of
(Am), 1972, 54, 1451-1454.
13. GUERRERO G, SAEIH C, DOKCENDORF I, DIAZ V : Geni-
tourinary anomalies in children with congenital scoliosis.
14. BEALS RK, ROBBINS JR, ROLFE B : Anomalies associated
15. WINTER RB, LONSTEIN JE, BOACHE-ADIEI O : Congenital
16. WYNNE-DAVIES R : Congenital vertebral anomalies:
aetiology and relationship to spin bifida cystica. J Med
G ener, 1975, 12, 280-288.
17. STURM PF, CHUNG R, BOMZE SR : Hemivertebra in
18. BLONDEL B, BREART G, DU MAZAUBRUN C, BADEYAN G,
WICSL O M, LOR DIER A, MATE T N : The perinatal situation
19. KESLING KL, LONSTEIN JE, DENIS F, PERRA JH,
SCHWENDER JD, TRANSFELDT EE, WINTER RB : The
crankshaft phenomenon after posterior spinal arthrodesis for
congenital scoliosis: a review of 54 patients. Spine, 2003,
28, 267-271.
20. HALL JE, HERNDON WA, LEVINE CR : Surgical treatment of
congenital scoliosis with or without Harrington instrumenta-
21. CHEUNG KM, ZHANG JG, LU DS, K LUK KD, Y LEONG JC : Ten-
year follow-up study of lower thoracic hemivertebrae
treated by convex fusion and concave distraction. Spine,
2002, 27, 748-753.
22. WINTER RB, LONSTEIN JE, DENIS F, STA-ANA DE LA ROSA H :
Convex growth arrest for progressive congenital scoliosis
23. UZUMCUGIL A, CI L A, YA ZICI M, A CAROGLU E, ALANAY A,
AKSOY C, SURAT A : Convex growth arrest in the treatment of
congenital spinal deformities, revisited. J Pediatr Orthop,
24. ANDREW T, PIGGOTT H : Growth arrest for progressive
scoliosis. Combined anterior and posterior fusion of the


