Atypical congenital kyphosis

REPORT OF TWO CASES WITH LONG-TERM FOLLOW-UP

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We describe two patients with an atypical congenital kyphosis in which a hypoplastic lumbar vertebral body lay in the spinal canal because of short pedicles. There were no defects in the posterior elements, or any apparent instability of the facet joints.

Both patients were treated successfully by anterior fusion to the levels immediately above and below the affected vertebra, and posterior fusion which extended one level more both proximally and distally. This gave progressive correction of the kyphotic deformity by allowing some continued anterior growth at the levels which had been fused posteriorly.

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Congenital kyphosis is a serious condition in which progressive deformity may result in paraplegia.¹⁻⁶ Three types have been defined: failure of formation (type I), failure of segmentation (type II) and a combination of defects of both formation and segmentation of the vertebral body (type III).^{1,2,7,8} Failure of formation has the worst prognosis and the greatest risk of neurological involvement of any spinal deformity.^{1-3,5-12} For these cases, the accepted principle of treatment is early detection and posterior fusion. For children younger than five years of age with a kyphosis of less than 50 to 55°, posterior fusion alone is adequate,^{2,4,6,11} but for older patients or those with greater deformity, combined anterior and posterior fusion is recommended. The posterior elements are usually preserved, but pure anterior and symmetrical failure is rare, especially when a hypoplastic vertebral body is located in the line of the spinal canal and there are no abnormalities of the posterior arch, which determines the stability of the curve.¹

We report the long-term follow-up of two children with hypoplastic lumbar vertebral bodies displaced posteriorly with no associated defects of the posterior element or posterior instability. In these patients the anomaly was not typical of posterior hemivertebrae in terms of location of the lesion and its treatment.

Case Reports

Case 1. An 18-day-old white boy presented with a thoracolumbar kyphotic deformity. A patent ductus arteriosus had been repaired at the time of birth. There was a palpable bony prominence over the upper lumbar region, but the spine was otherwise straight with no lateral deformity or scoliosis and there were no neurological deficits. Radiographs showed a hypoplastic second lumbar vertebral body which was displaced posteriorly into the spinal canal (Figs 1a and 1b), and a kyphotic deformity of 10°.

At 15 months of age, tomography showed segmental narrowing of the spinal canal by a vertebral body which was displaced posteriorly by about 70% of its width (Fig. 1c). There were no signs to suggest posterior instability in dynamic radiographs. CT confirmed the short pedicles of the second lumbar vertebra (Fig. 1d). At 18 months, the kyphosis had increased to 25° (Fig. 1e) and operation was advised.

At 22 months of age we performed a combined anterior (L1 to L3) and posterior (T12 to L4) spinal arthrodesis without correction of deformity using autogenous rib grafts anteriorly and iliac bone posteriorly.

In the lateral decubitus position and through a retroperitoneal approach, an anterior defect was palpable at the apex of the kyphosis. Fibrous tissue connecting the first and third lumbar vertebral bodies was removed to a depth of 1.5 cm to expose the anterior aspect of the displaced body. Both proximally and distally, the intervertebral disc, growth plate and fibrocartilaginous tissue were removed. A rib graft was impacted between the bodies of L1 and L3 and additional iliac-crest bone was packed around this. Bilateral posterior fusion, using autogenous iliac bone, was performed from T12 to L4. No laminar defects or facet joint instability were seen at operation. Postoperatively, the patient remained in a body-jacket cast for six months.

Five years and six months after operation, both anterior

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Fig. 1a

Fig. 1b

Fig. 1c

Fig. 1d

Fig. 1e

Fig. 1f

Fig. 1g

Figures 1a and 1b – Radiographs of the spine of an 18-day-old boy showing a hypoplastic second lumbar vertebral body located in the spinal canal. Figure 1c – Local view and metrizamide tomogram showing a segmental narrowing of the spinal canal, but no dislocation of the facet joint. Figure 1d – CT showing a hypoplastic vertebral body and short pedicles of the second lumbar vertebra. Figures 1e to 1g – Serial lateral radiographs of the thoracolumbar spine at 18 months of age, at 5 years 6 months and at 14 years 6 months postoperatively. The clinical results were excellent and there was no kyphotic deformity.

Fig. 2a

Fig. 2b

Fig. 2c

Fig. 2d

Fig. 2e

Figures 2a and 2b – Plain radiograph and tomogram showing a hypoplastic first lumbar vertebral body that was posteriorly displaced into the spinal canal. Figures 2c to 2e – Serial lateral radiographs of the thoracolumbar spine at the age of 2 years 4 months, at 5 years 4 months and at 13 years and 5 months. There was no kyphosis at the thoracolumbar junction and the cosmetic result was excellent.

and posterior fusions appeared to be solid and the kyphotic deformity had decreased from 25° to 15° (Fig. 1f). At the latest follow-up, at the age of 16 years and 4 months, there was no kyphotic deformity of the thoracolumbar junction (Fig. 1g); the patient had no pain or neurological deficit and an excellent cosmetic result. He participates in all sports.

Case 2. A four-month-old black boy was transferred to our hospital with congenital laryngomalacia and respiratory distress syndrome. There was a gibbus deformity in the thoracolumbar area with no sign of scoliosis. Neurological examination was normal except for a slight increase in muscle tone in the limbs.

Radiographs and tomograms showed a hypoplastic first lumbar vertebral body, displaced posteriorly into the spinal canal by 30% of its width (Figs 2a and 2b) and a kyphotic angle of 32°.

At 19 months of age, a combined anterior (T12 to L2) and posterior (T11 to L3) spinal arthrodesis was performed without correction as described for case 1, and again no defects of the posterior element or instability of the facet joints were seen. At the age of five years four months, radiographs showed a solid fusion mass and the kyphotic deformity had improved from 44° to 24° at 2 years 2 months (Figs 2c and 2d). At our latest follow-up at the age of 13 years 5 months, there was no definite kyphosis (Fig. 2e), no neurological deficit and an excellent cosmetic result.

Discussion

Treatment of known congenital anomalies of the spine in children is often delayed because of failure to recognise the potential for deterioration and the need for early treatment. The aetiology of these deformities is unclear, but the vertebral column is known to be well formed at the end of the embryological period.^{7,8} Congenital kyphosis develops prenatally as a result of specific growth defects in the centrum and osseous metaplasia of the anterior annulus fibrosus. Hypoplasia and aplasia of the vertebral body are part of a spectrum of growth deficits of the centrum during the late stages of chondrification and ossification, when there is a rapid increase in vertebral size.^{7,8}

Tsou et al⁸ suggested that there were six common forms of deficiency of the centrum, and that the development of pure kyphosis or kyphoscoliosis depended on the location of the defect. A posterior hemicentrum or aplasia of the centrum produces a pure kyphosis and our cases may fit into this category. It usually exerts direct nerve pressure, causing a progressive traction myelopathy due to displacement of the osseous ring.^{7,8} Our patients differed in that the vertebral body was in the spinal canal, occupying space but with no definite posterior displacement of the osseous ring.

Dubousset¹ classified failure of formation into two types: type I is partial failure with a well-aligned spinal canal and an intact posterior arch and type II is partial failure with a dislocated canal that can almost always be seen in the anteroposterior plane as a bayonet type of appearance. He also noted that the bodies, pedicles and posterior arches may each be absent. In our cases, the cause of the posterior displacement of the hypoplastic vertebral bodies with no displacement of the osseous ring or associated defects of the posterior element seemed to be an embryological mal-development resulting in a hypoplastic body with short pedicles, as shown clearly on CT.

Shapiro and Herring¹³ and Zeller, Ghanem and Dubousset¹⁴ have suggested another type of congenital kyphosis, using the terms 'congenital vertebral displacement' and 'congenital dislocated spine', respectively. They both stressed that abnormal posterior elements were always present, and

all their cases had posterior displacement of anomalous vertebrae showing both anterior failure of formation and defects of the posterior element. Zeller et al¹⁴ described the loss of continuity of the posterior cortex of adjacent vertebral bodies as the 'step-off sign', a diagnostic hallmark of a congenital dislocated spine. Our patients both showed this radiological sign but the vertebral column below the hypoplastic vertebra was not subluxed or dislocated and there were no defects of the posterior arch. Scott et al¹⁵ and Faciszewski et al¹⁶ introduced the term 'segmental spinal dysgenesis', characterised by anterior subluxation of the normal segment of the spine above the dysgenesis, with absence of the pedicles and transverse and spinous processes, as well as failure of formation of the vertebral body.

Treatment of congenital kyphosis should aim to prevent progression of the deformity. Failure of formation produces an average progression of 7° a year,^{1,5,6,11,12} but surgical treatment gives good results. In younger children without severe deformity (under 50 to 55°) anterior fusion is not usually advised. Posterior fusion alone extending to one vertebra above and one below the deformity allows any anterior growth to continue, with the possibility of slow but steady correction of the kyphosis. Winter et al,⁶ among others,^{2,4} have found little spontaneous correction after both anterior and posterior fusion in younger children, but with a compromised spinal canal in 'congenital dislocated spine' or 'segmental spinal dysgenesis', posterior fusion alone gave poor results. For these cases combined anterior and posterior fusion is regarded as the only reliable procedure.^{13,16,17}

The two patients whom we describe had excellent results after a short anterior fusion with a longer posterior fusion extending one level above and below the anterior fusion. In similar cases with a hypoplastic vertebral body with short pedicles deeply located in the spinal canal, posterior fusion alone may not provide spontaneous correction or prevent neurological damage secondary to bending of the fusion mass. Because of the high incidence of pseudarthrosis in a posterior fusion, re-exploration at about six months after the first operation is often recommended, with the addition of more autogenous bone. We believe that apex pseudarthrosis may be prevented by our anterior and posterior fusion. This may achieve a more reliable result than posterior fusion alone, particularly in patients with the unusual findings which we have discussed.

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