TWO-STAGE CORRECTIVE SURGERY FOR CONGENITAL
DEFORMITIES OF THE SPINE

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Sixty patients with congenital deformities of the spine were operated upon in the past fifteen years using a two-stage procedure. In the fifty patients with scoliosis half of the deformities were due to hemivertebrae and half to unilateral bars. The average correction of the deformity was 47 per cent. Early neurological signs observed in two patients with a diastematomyelia resolved. Of the ten patients with kyphosis nine had neurological signs of impending paraplegia and one was completely paraplegic before operation; all improved markedly.

Posterior spinal fusion alone in the rapidly progressing congenital deformity may not prevent further progression, particularly in those cases with unilateral bars. Anterior resection of the vertebral body with later posterior fusion with Harrington instrumentation is safe and effective.

The Scoliosis Research Society has accepted the classification of spinal deformities proposed by Goldstein and Waugh (1973) which includes a classification of congenital deformities devised by MacEwan, Conway and Miller (1968) with subdivision into scoliosis, kyphosis or lordosis. The embryological aetiology may be a failure of bone formation, a failure of bone segmentation, or both. Failures of formation may be complete and unilateral (hemivertebrae) or partial and unilateral (wedge vertebrae). Failures of segmentation may be unilateral, producing “bars”, or bilateral, producing “blocks”. Long-term studies of the behaviour of these deformities emphasise the need for a clear understanding of their aetiology (Winter, Moe and Eilers 1968; Winter, Moe and Wang 1973; James 1975). Solitary hemivertebrae, solitary wedge vertebrae, or balanced hemivertebrae on each side of the spine are not usually associated with rapid progression. This is the case in approximately 50 per cent of congenital deformities of the spine. Multiple hemivertebrae on the same side, and particularly unilateral unsegmented bars, are, however, notorious for their progression, there being normal or near-normal growth on one side of the spine and no growth or virtually no growth on the other (Bradford, Moe and Winter 1975).

In the management of congenital deformities of the spine this asymmetry of growth potential must not be allowed to cause progression of the deformity: if progression is noted, treatment is indicated. Because external support by casts or braces is successful in only a small percentage of cases, an operation is the definitive treatment (Winter 1973). Harrington instrumentation is contraindicated due to the high incidence of paraplegia as a result of tethered spinal cords (Winter 1973). As a consequence posterior “fusion-in-situ” has become the recommended procedure, even in very young individuals (Winter 1973), but it is at best only a holding operation. Furthermore, in rapidly progressive deformities, not only does progression continue, but it may do so at a greater rate due to the tethering effects of the fusion (Roaf 1966). This is hardly surprising as the fusion is posterior, some distance from the area of asymmetric growth which is anterior. In addition, these deformities are rigid.

In order to correct a rigid spine without incurring the danger of traction paraplegia, the spine must be shortened as well as straightened. A two-stage corrective procedure has been introduced for this purpose (Leatherman 1969, 1973). This paper describes the operative considerations and the results of sixty patients treated thus in this centre over the past fifteen years.

DEVELOPMENT OF THE TWO-STAGE PROCEDURE

The first report of the removal of a portion of the spine for a fixed deformity was by Royle (1928). A two-stage procedure was reported by von Lackum and Smith (1933): in the first stage the vertebral body and posterior elements were removed and in the second stage the spine was fused. Serious complications arose from such operations where the vertebral body and posterior elements were removed at the same time (Wiles 1951). A two-stage procedure was, therefore, conceived with the vertebral body being removed through an anterior approach in the first stage and the posterior elements removed and fusion performed in the second stage. With his interest in the anterior approach

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to the spine for tuberculosis, Hodgson (1965) reported two cases of fixed kyphosis corrected by anterior opening-wedge osteotomy and anterior strut grafts. He suggested that alternatively a closing-wedge osteotomy could be done on the convex side of the deformity. This forms the basis of the first stage of the two-stage procedure and enables the spine to be shortened as well as straightened to preserve the neurological function. As a result the anterior asymmetric growth is halted. However, the curve remaining above and below the osteotomy will continue to deform throughout growth. A posterior fusion of the entire curve is therefore necessary; this is the basis of the second stage, usually performed three weeks later when the anterior wound has healed and the patient has recovered from the first operation. Between the two stages the patient is nursed free in bed without external support to the spine. In the second stage the posterior elements are removed at the same level to complete the wedge resection which is then closed with a compression system. Additional stability and correction are gained by the insertion of a distraction rod on the concave side.

**OPERATIVE TECHNIQUE**

Stage 1: anterior resection of the vertebral body. The apical vertebral body is excised and, depending on the location of the curve, thoracic, thoracolumbar, or lumbar exposures are necessary. These approaches have been well described (Riseborough 1973) but certain points of technique are most important when resecting a vertebral body. The spine is approached from the convex side of the curve which greatly facilitates exposure of the scoliotic spine which, in severe cases, may lie in contact with the rib cage or abdominal wall. Exposure of the kyphotic spine from the front is more exacting.

When approaching the thoracic spine exposure is facilitated by entering the chest through the bed of the rib above the vertebral body that is to be excised. This eliminates the overhang provided by the rib above. The spine is best located by dissecting the rib back and disarticulating it from its transverse process (Fig. 1). When there is adherence or rib fusion, disarticulation may be eased by incising the pleura along the line of the rib to its attachment to the spine, thus allowing the entire costovertebral articulation to be seen. The pleura is then gently lifted upwards and downwards to expose the apical vertebral body and the bodies above and below. The segmental vessels at these three levels are then doubly ligated and divided. This should be performed on the anterior surface of the vertebral bodies so as not to impair the vertically orientated anastomosis which is important in the nutrition of the spinal cord. The periosteum is then incised in the same line as the pleura and these two layers afford a satisfactory closure at the completion of the resection. The rib attached to the apical vertebral body is now disarticulated at the costovertebral joint and its posterior 2 inches excised. The vertebra is now exposed ready for excision.

The thoracolumbar region of the spine is exposed by means of a thoraco-abdominal approach through the bed of the tenth or eleventh ribs. After rib resection the deep periosteum together with the pleura is incised throughout the length of the wound which is enlarged by means of a rib spreader. The diaphragm is detached peripherally leaving a fringe for reattachment at the end of the operation. The abdominal part of the exposure is entirely retroperitoneal, the peritoneum and its contents being mobilised by blunt dissection towards the concavity. An excellent view of the thoracolumbar region of the spine is thereby obtained and the periosteum and segmental vessels are dealt with as in the thoracic approach.

Exposure of the lumbar spine is achieved by the extraperitoneal approach used for lumbar sympathectomy. The peritoneum is displaced medially forwards off the posterior abdominal wall, the ureter and genital vessels being raised with it. The periosteum and segmental vessels are dealt with as before. The spine thus approached must be exposed laterally and anteriorly.

The apical vertebral body is resected by a closing-wedge osteotomy, more of the convexity being removed than of the concavity (Fig. 2). Thin layers of bone are removed with a sharp chisel or gouge. Bleeding from the richly vascular cancellous bone of the vertebral body is controlled by means of bone wax. The greater part of the vertebral body can thus be removed before the spinal canal is breached. Entry into the spinal canal is best gained by enlarging the intertransverse foramen after removal of the transverse process by rongeur. The posterior longitudinal ligament with a thin shell of cancellous bone attached is then removed by pituitary punches (Fig. 3). Bleeding is controlled by thrombin-soaked Gelfoam and cottonoid pledgets. The pedicle, transverse process, and facets on the concave side are left intact and provide stability to the spine between the two stages. The resection area is smoothed by rongeur, filled with Gelfoam soaked in thrombin and closed by suturing the pleura and periosteum.
Stage 2: posterior resection, fusion and instrumentation. The spinal column is approached posteriorly in the midline and the resection area is identified. The posterior elements and the pedicle and transverse process on the concave side are removed to complete the closing wedge. This area is inspected to ensure that there are no sharp bony spicules or loose fragments. The wedge deficit is then closed by means of a compression system applied to the convex side and additional stability is obtained by inserting a distraction rod on the concave side. There must, however, be no strong distraction such as might embarrass the spinal cord or nerve roots. Posterior fusion of the entire structural curve is then performed. The patient is kept supine in a cast or a crutch-type brace for six months; walking is then allowed but the spine is supported by this same well-fitting brace for a further six months.

CASE REPORT

Case 1. This girl, aged one year two months, had a congenital thoracic scoliosis of 56 degrees due to a unilateral bar (Fig. 4) and underwent a posterior “fusion-in-situ” of the entire curve. She was referred to this centre at the age of five years four months with a deformity of 110 degrees (Fig. 5). Posterior fusion had not halted progression and may have accelerated it. Studies of pulmonary function showed a severe restrictive defect but there were no neurological deficits and myelography revealed no abnormality. Two-stage correction was performed but in the second stage the previous fusion mass was osteotomised in five places to facilitate adequate correction. The deformity was thereby corrected to 35 degrees with restoration of chest symmetry and trunk balance (Fig. 6). The patient made an uneventful recovery.

CLINICAL MATERIAL

Sixty patients with congenital deformities of the spine have been treated in this manner with a minimum follow-up of eighteen months. There were fifty with scoliosis and ten with kyphosis.

Congenital scoliosis

Patients and methods. There were thirty girls and twenty boys, the mean age at operation being eleven years (range two years three months to sixteen years eight months) and the mean follow-up five years five months (range one year six months to twelve years seven months). Half of the deformities were due to hemivertebrae and half to unilateral bars. Pulmonary function studies were performed on all patients before operation and, although diminution in pulmonary function did not correlate with the magnitude of the curve, it was observed that in thoracic curves the pulmonary function was never normal while in lumbar curves it was always normal. Myelography was performed routinely and a diastematomyelia was found in four patients, two of whom had clinical evidence of early neurological deficit.

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<th>Table I. Results of the two-stage procedure</th>
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These bony spurs were removed in a separate procedure. The deformity was measured on a standing radiograph before operation, on a supine radiograph after the second stage, and on a standing radiograph at the follow-up examination (Cobb 1948).

Results. The measurements of the curves are shown in Table I. The mean correction obtained after the second stage was 47 per cent. At follow-up the mean curve was 43 degrees, but the large standard deviations indicate the wide range of curves treated. The early neurological signs observed in the two patients with a diastematomyelia resolved, and the following case illustrates such a patient.
Case 2. This girl, aged eight years one month, had a congenital scoliosis measuring 95 degrees due to a unilateral bar (Fig. 7) and early neurological signs of impending paraplegia. A myelogram revealed a diastematomyelia at the apex of the curve (Fig. 8). After a two-stage corrective operation including removal of the bony spur, the deformity was reduced to 26 degrees (Fig. 9) and her neurological signs disappeared.

Congenital kyphosis

Patients and methods. There were four girls and six boys, the mean age at operation being twelve years three months (range two years seven months to twenty-nine years three months) and the mean follow-up three years eight months (range fourteen months to ten years). Eight of the deformities were due to hemivertebrae, one to a wedge vertebra, and one to an anterior bar, and all were at the thoracolumbar junction. Nine patients had neurological signs of impending paraplegia and one was completely paraplegic before operation. Myelography performed in all cases showed evidence of compression of the cord from the front. The curve was measured as for the scoliosis patients.

Results. The size of the curves and the corrections (Table I) were similar to those for the scoliosis group but were of less importance as these individuals were operated upon because of their neurological signs. The nine patients with early neurological deficit recovered completely, while the patient with paraplegia recovered sensation and control of the bladder but only minimal motor function. At follow-up he was able to walk with long leg braces and crutches. This case has already been described in detail (Leatherman 1973).

COMPLICATIONS

Early complications comprised three cases of delayed healing, two of atelectasis, and one pleural effusion, making a total of six, or 10 per cent of all cases. These less severe problems resolved uneventfully. There was only one late complication, a pseudarthrosis, but there were no neurological problems, no wound infections, and no deaths in this series.

DISCUSSION

Posterior spinal fusion is successful in preventing progression of adolescent idiopathic curves because the potential for asymmetric growth is not great. This, however, is not so in rapidly progressive congenital deformities, particularly those due to unilateral unsegmented bars where the potential is at a maximum and a posterior “fusion-in-situ” may not prevent progression (Figs 4 and 5). Furthermore, as these deformities develop they also become rigid. Neurological problems are easily produced by attempting to correct them with Harrington instrumentation alone. The only way of correcting a rigid deformity without incurring the danger of neurological complications is to shorten the spine as well as to straighten it. The development of anterior resection as the initial part of a two-stage technique is particularly relevant when there are coexistent neurological problems, for these can only be solved by an anterior approach. Hall (1973) stated that a spinal cord compressed from the front by a kyphosis or any other cause must be decompressed from the front as a
laminectomy will not only be futile but actually harmful.

Patients with congenital deformities of the spine need a very thorough evaluation before operation. This should include a careful neurological examination and myelography is mandatory. When dealing with the thoracolumbar junction or the lumbar spine, intravenous pyelography is essential so that any associated congenital renal abnormality can be appreciated before operation. Pulmonary function studies are performed routinely and varying degrees of restrictive lung defects noted in thoracic curves. These spirometric tests are only a part of the evaluation and it is most important that these patients be examined by an expert in cardiopulmonary function. In cases where there is considerable embarrassment of heart and lung function the assistance of such an expert during the postoperative phase is most helpful. Similarly, these patients should be carefully assessed by the anaesthetist. When the anterior surgical approach can be expected to the particularly exacting, as in a case of marked kyphosis at the thoracolumbar junction, we have no hesitation in obtaining the help of an experienced general surgeon when exposing the spine. Similarly, when a diastematomyelia is resected the assistance of a neurosurgeon should be sought by those unfamiliar with this procedure.

By such scrupulous attention to detail in the planning of the operative procedure and its performance, complications are kept to a minimum.

REFERENCES


