

## Congenital Anteroposterior Spinal Dissociation in Larsen's Syndrome: Report on Two Operated Cases With Long-Term Follow-up

[Case Reports]

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### Abstract

**Study Design.** The outcome of two patients with Larsen's syndrome after spinal surgery was evaluated after follow-up for 9 and 16 years.

**Objective.** To report on a new phenomenon of anteroposterior dissociation of the vertebrae in Larsen's syndrome. To demonstrate that it can be visualized before surgery with imaging and also seen intraoperatively. To show that the neurologic recovery after surgery is sustained and to review the technical reasons for the difficulties in achieving a surgical fusion. The critical role of CT scanning will be illustrated.

**Summary of Background Data.** The results of preoperative and postoperative radiologic investigations and intraoperative findings are presented to support this new phenomenon.

**Methods.** Two patients underwent multiple spinal surgeries because of a deteriorating myelopathic clinical status. Intraoperatively, anteroposterior dissociation was documented in both patients. There was great difficulty in obtaining a surgical fusion, and an unusually long circumferential fusion was eventually necessary to obtain stability. Prolonged halo-vest immobilization was essential.

**Results.** Neurologic recovery was sustained over time, and the spinal deformity did not deteriorate.

**Conclusion.** Awareness of this phenomenon is essential to the management of spinal deformities in Larsen's syndrome when presenting with myelopathy. Special features in the radiologic workup should be sought after so as to plan staged surgical procedures. Conventional principles of planning of fusion levels are inadequate. Early treatment is advocated, as the neurologic compromise is reversible.

Larsen's syndrome was actually first described by Sorrel in 1932, although it was Larsen in 1950 who later described the first series consisting of six patients and whose name was subsequently accredited to this condition. This disorder is characterized by distinct facial features and multifocal musculoskeletal features. The facial features consist of flat faces, prominent forehead, depressed nasal bridge, and hypertelorism. There are also multiple major joint dislocations and mixed vertebral anomalies throughout the spine. Siblings with the same disorder have been reported. The spine is also uniquely involved with severe dysraphism and spondylolysis often resulting in substantial spinal deformities. However, the cervical spine is less commonly involved and Tachjian reported an incidence of 20%. Cervical spine involvement when present can be severe with difficult to manage deformities such as midcervical kyphosis. The typical young patient is particularly difficult to evaluate, as there is concurrent hypoplasia, dysraphism,

and immature cartilaginous elements. Micheli was the first to emphasize that beside the functional and biomechanical aspects, these deformities are potentially lethal. One such consequence of sudden respiratory arrest was reported in Larsen's original series. The frequency of such incidents has prompted the anesthetic literature to highlight extreme precaution with positioning and intubation of these patients. Of all the operated cases of Larsen's syndrome reported in the literature, the indications have all been due to the presentation of abnormal neurology. They have all occurred below the age of 3 years. The indications for our patients were also for neurologic complications. The neurologic picture of our two patients improved significantly after surgery unlike the other reported cases.

Although the principle of spinal fusion has been discussed in Larsen's syndrome for those with significant spinal instability or deformity, we are unaware of any reports as to detailed intraoperative findings, nor have there been technical suggestions as to the need to modify conventional principles to ensure a stable fusion can be achieved.

### Case Reports

#### Case 1.

An 8-year-old girl was diagnosed to have Larsen's syndrome, presenting with flexion deformities of both elbows and knees, bilaterally dislocated hips, clubfeet, and severe spinal deformities. Two spinal problems were identified. Plain radiographs showed (1) a thoracic kyphosis between T11 to L2 and (2) a significant cervical kyphoscoliosis due to multiple vertebral body hypoplasia with associated posterior spinal dysraphism from C4 to C7 (Figure 1A).

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The surgery was performed at the Duchess of Kent Children's Hospital, Hong Kong.

Acknowledgment date: July 6, 2001.

Revision date: November 21, 2001.

Acceptance date: February 4, 2002.

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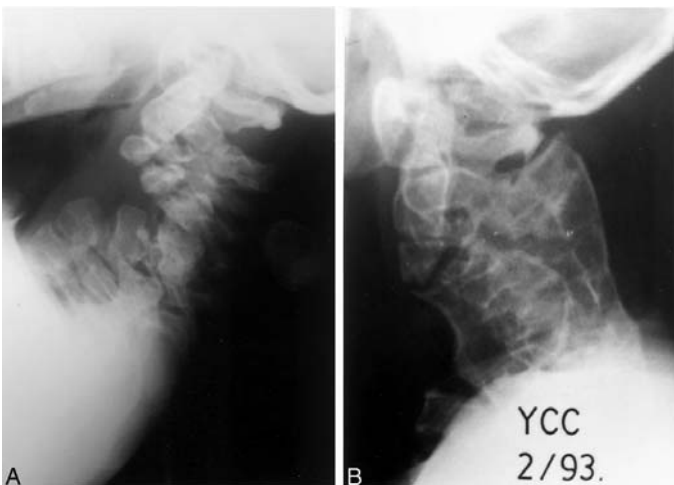
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The manuscript submitted does not contain information about medical device(s)/drug(s).

No funds were received in support of this work. No benefits in any form have been or will be received from a commercial party related directly or indirectly to the subject of this article.

[Key words: Larsen's syndrome; spinal dysraphism; anteroposterior dissociation] *Spine* 2002;27:E296-E300

Figure 1. Case 1. **A**, Lateral preoperative radiograph of cervical kyphoscoliosis at C4–C7. **B**, Lateral postoperative radiograph showing that the cervical fusion was achieved.



Over time the thoracic kyphosis was determined to be progressive. A discectomy and anterior bone grafting between T12–L3 was therefore performed. Rib graft was used because of the poor quality of her iliac bone stock. Intraoperatively, the anterior spinal bodies were noted to be dissociated from the posterior elements such that one could almost pull the vertebral body out from the spinal column. A second-stage elective posterior fusion was performed 1 month later. The spinous processes were confirmed to be fibrocartilaginous and on this second occasion, the posterior elements were separable from anterior structures with a gentle distraction force. No instrumentation was implanted but a plaster jacket was worn for 8 months after surgery. Serial radiographs and tomograms over 3 years showed no evidence of fusion. Because the thoracic deformity was static, further surgery to deal with the pseudarthrosis was declined by the patient and her family.

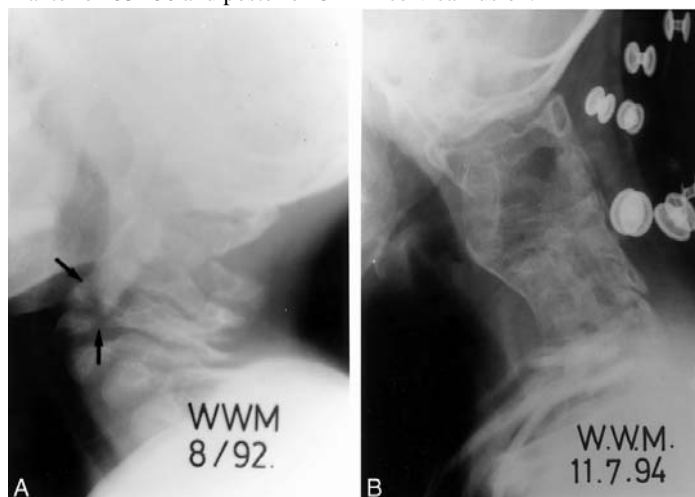
While being treated for the thoracic kyphosis, she complained of an increasing sensory deficit in the upper extremities. A myelogram was performed as shown showing severe lack of contrast at the apex of the kyphosis. MRI confirmed maximum stretching of the spinal cord over the C5–C6 internal kyphus. The cervical spine required an anterior decompression and stabilization from C3 to C7. Postoperative immobilization in a halo-thoracic vest was used in view of the inherent instability and the long fusion. During the immediate postoperative period she developed transient weakness in both the upper and lower extremities, which subsequently recovered completely.

Of particular interest was that intraoperatively several levels of cervical vertebral bodies were also noted to be independently mobile as experienced in the lumbar spine. This suggested that there was also gross dissociation between the anterior and posterior spinal elements at several cervical levels. Therefore, a longer fusion than had been anticipated before surgery was performed.

Unfortunately, she developed a nonunion despite wearing a well-maintained halo-thoracic vest for a period of 3 months followed by 3 months of cervical collar protection. Nine months later a posterior fusion (C2–C7) was done for the progressing cervical kyphotic deformity. The posterior approach allowed us to visualize the posterior dysraphism. We were able to reexamine the anteroposterior disassociation and confirm the discontinuity to be at the level the body–pedicle junction. Eventually, a solid anterior and posterior fusion mass of the cervical spine was achieved only after a total of 18 months (Figure 1B).

Case 2. A 6-year-old girl was referred to our center for clubfeet, flexion deformity of both hips and knees, and cervical spine anomalies. Plain radiographs showed she had multiple levels vertebral body hypoplasia with posterior dysraphism leading to a collapsing lordoscoliotic cervical spine. The plain radiographs also showed multiple levels of spondylolysis from C4 to C6 (Figure 2A). In addition, a MRI demonstrated occipital condylar invagination, congenital C1–C2 fusion, and mild spinal cord atrophy over the curvature of the lordosis. This was important in being able to exclude the possibility of contributing compression from C1–C2 instability and/or condylar invagination. Clinically, she developed progressive symptoms of myelopathy because of the dynamic cervical instability from C4 to C6. Initially, a halo-thoracic vest alone was applied to stabilize and reverse the neurologic deterioration and in doing so demonstrated the dynamic nature of the compression. This led to marked clinical improvement and thus a long posterior spinal fusion from C1 to T1 was performed.

Figure 2. Case 2. **A**, Preoperative lateral radiograph showing cervical kyphoscoliosis at C1–C6. An anteroposterior dissociation defect is illustrated (black arrows). **B**, Postoperative radiograph showing anterior C3–C6 and posterior C1–T1 cervical fusion.



Intraoperatively, the C5 and T1 laminae were found to be completely dissociated from the vertebral bodies at the pedicle region. The other areas of spondylolysis seen on the preoperative radiograph were actually fibrocartilage. Having seen this pathoanatomy and because of our previous experience we elected a second-stage anterior fusion from C3 to C6 4 weeks later. At the second surgery we confirmed the vertebral bodies to be hypoplastic and also dissociated from the posterior structures at the pedicle region. During this operative session the instability between consecutive vertebrae was even more pronounced after completing an adjacent discectomy. The vertebral bodies could be literally plucked out from the spinal column with one's fingers. This would not normally occur and confirmed without doubt the independence of the anterior and posterior spinal elements.

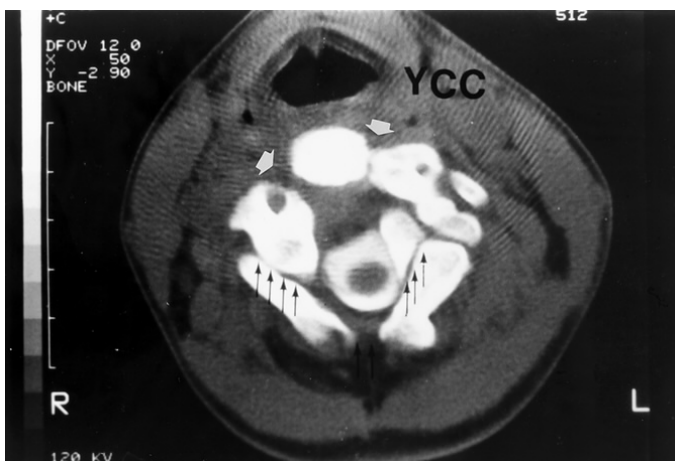
Expectedly, the symptoms of myelopathy resolved after surgery. However, follow-up radiographs demonstrated a nonunion between C3–C4 and C4–C5 anteriorly. In anticipation that the nonunion may cause further myelopathy in the future and also because the instability was deemed dangerous, the anterior fusion from C3 to C5 was redone. Four months later radiographic evaluation of the cervical spine in flexion and extension views showed persistent nonunion at C4–C5 both anteriorly and posteriorly. The patient further underwent a repair of the pseudarthrosis anteriorly and augmentation of the fusion posteriorly from C2 to C6. There was eventually solid anterior and posterior

fusion of the cervical spine determined by radiographic evaluation after a total period of 19 months of postoperative halo-vest immobilization ([Figure 2B](#)).

## Results

Case 1 has been followed up for 16 years and has reached skeletal maturity. Case 2 has been followed up for 9 years. No further deterioration of either cervical spine has arisen. Their neurologic statuses are stable. A supplementary MRI ([Figure 3](#)) was taken of case 1, 6 years after the fusion. Case 2 is now 15 years old and has needed further surgery also for thoracolumbar scoliosis. Both patients enjoy full usage of their upper limbs with normal sensory and motor function, but independent ambulation has been limited because of anatomic problems arising in their hips.

Figure 4. Case 1. CT myelogram scan at C3 showing spinal dysraphism at the embryonic junction between centrum and pedicle (white arrows) and bilateral pedicle defects causing spondylolisthesis and anteroposterior dissociation (black arrows).

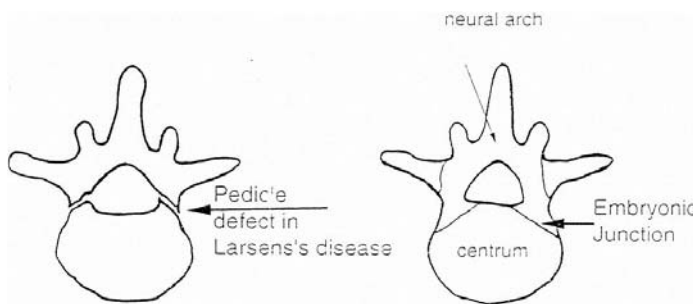


The MRI shows the bony defects not as well ([Figure 3](#)) because it cannot outline the bone cortex clearly. The minimal amount of marrow in the dysplastic vertebral bone does not show up well. Its main role is in identifying nonbony pathology, especially of intraspinal lesions, such as the severity of cord atrophy as in our patients, which is important for estimating prognosis.

The vertebra develops from three morphologic components the centrum and the right and left neural arches ([Figure 5](#)). Costal elements add further components to the basic vertebrae. For example, the foramen in the transverse process of a typical cervical vertebra is formed from both the true transverse process and its costal elements. In the thoracic spine the costal elements develop separately as the ribs, which articulate with the neural arches. In all the other parts of the vertebral column the costal elements become fused to the neural arches and become incorporated as morphologic parts of the vertebrae. It should be noted that the morphologic centrum is not the same as the anatomic body of the vertebra as part of the neural arch is incorporated into the anatomic body. This neurocentral junction lies anterior to the costal facets on the body.

In Larsen's syndrome this normal development is disturbed. We have consistently found the location of the abnormality at the pedicle at all levels of the spine. The abnormality is not at the normal embryonic junctional area ([Figure 5](#)) and therefore

Figure 5. Diagrammatic representation of the different morphologic components of the vertebra. A defect in the region of the pedicle in Larsen's syndrome is responsible for the anteroposterior dissociation.



cannot be explained by a simple failure of the ossification centers but instead represents a specific feature of this condition. Sometimes the defect is completely discontinuous and sometimes there is some cartilaginous component maintaining a tenuous continuity. The consequence of this fact is that the cartilage, if present, may eventually ossify and ensure rigidity. This cartilaginous area can thus be managed for the moment, as if it were of normal morphology. The morphologic appearance of the pedicular dysplasia is variable. It is this discontinuity at the pedicle that gives rise to this phenomenon of anteroposterior disassociation of the vertebrae. With the increasing availability of the CT and MRI, these methods should be liberally used to distinguish areas of complete pedicular discontinuity and pedicular bridging by fibrocartilage. We found that the CT images were clearly superior in providing this preoperative information ([Figure 4](#)). This condition should be distinguished from spondylolysis, which usually affects one level only and more commonly at the pars interarticularis. It is also different from spina bifida, which affects the lamina region instead. Historically, the term "spinal dysraphism" was a general term used for any part of the vertebrae, which was morphologically abnormal. We think that anteroposterior dissociation is the result of a specific defect at the pedicle region.

## Discussion

The natural history of this condition and in particular of the spinal deformity is unknown. The largest series of this syndrome is that reported by Laville et al [3](#) on 38 patients living on Reunion Island. This high incidence is probably related to the high degree of consanguinity practiced on the island. The mode of inheritance remains controversial, as both autosomal dominant and recessive patterns have been shown.

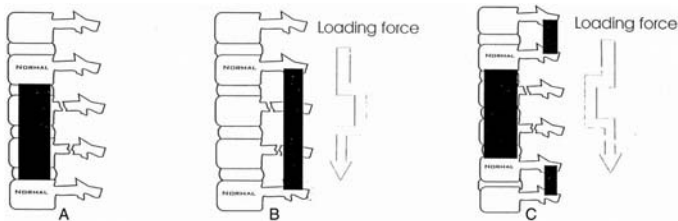
Bowen et al [1](#) tried to analyze the pattern of presentation of the spinal deformities by systematically analyzing the whole spine in a series of eight patients (ages 1–28 years). They [1](#) concluded that the cervical spine was the most severely affected by the abnormalities. Scoliosis was the most common presentation in the thoracic spine and spondylolysis in the lumbar spine. They noticed that the posterior spinal elements were radiologically more dysplastic than the anterior elements but did not give any details of the abnormality.

A review of the literature has highlighted that almost all the reported cases that needed a spinal operation were of a much younger age, usually <3 years of age. [2–5](#) The reported results were less successful when compared with our experience. Our two patients were slightly older and both had

good results. We would postulate that neurologic success is probably more related to the timing of the intervention, the earlier the better, rather than age per se.

The literature reports to date have tended to document only epidemiologic, physical, and simple plain radiographic features. Of the reported operated cases, detailed discussions of the possible reasons for their success or failures are not available. Little information was available to us as to the potential technical difficulties before we embarked on surgical stabilization. Although the erratic and asymmetric spinal dysraphism is well documented in the literature, its technical relevance has not been appreciated. Our experience suggests that in treating this condition, even obtaining a good anterior fusion mass and a good posterior fusion mass still does not guarantee a stable construct, as the two columns may not be interconnected. For this condition one must pay particular attention to the spinal column as a whole. There are three solutions to obtaining a stable construct. If either an anterior fusion ([Figure 6A](#)) or a posterior fusion alone ([Figure 6B](#)) is planned, then it must extend to vertebral levels above and below, which have continuity between the anterior and posterior elements of the vertebra. An alternative fusion configuration would be to perform a fusion, which bypasses the area with anteroposterior dissociation by a combination of anterior and posterior fusion ([Figure 6C](#)). Furthermore, whenever anteroposterior dissociation is suspected, single- or two-level interbody fusion should be avoided because of the dangerous instability resulting from the discectomy in these situations. On the issue of nonunion, we think that the higher nonunion rate is a result of several factors including the presence of the dysplastic elements and that younger patients have a relatively larger cartilaginous component. There is therefore a relative intrinsic reduction of bone surface for fusion to take place.

Figure 6. **A**, Anterior interbody fusion alone to levels with normal vertebrae. **B**, Posterior spinal fusion alone to levels with normal vertebrae. **C**, A long C-shaped bypass fusion.



## Conclusion

Our current strategy on the management of Larsen's syndrome, based on our experiences is that, first, any peripheral surgery or peripheral orthopedic problem cannot be dealt with properly until the whole spine has been thoroughly investigated. The spine and the spinal cord may be the cause of any distal pathology, such as a late presenting major joint dislocation. It can certainly affect the outcome of any treatment. Second, the neurologic compromise is progressive, especially in the presence of spinal instability. Early intervention will give a better outcome, as the neurologic deficit is reversible. Third, the spinal correction can be permanent if the stabilization is done according to our principles described. Patients with Larsen's syndrome should be followed up closely as the spine can decompress dramatically

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