Multilevel Spinal Segmental Fixation for Kyphotic Cervical Spinal Deformity in Pediatric Age Group—Report of Management in 2 Cases

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OBJECTIVE: **We discuss the role of instability of multiple spinal segments including the atlantoaxial joint in the pathogenesis of cervical kyphotic deformity.**

MATERIAL AND METHODS: **Two male patients (5 and 17 years old) had severe cervical kyphosis and presented with symptoms related to myelopathy. The patients underwent multisegmental spinal distraction and fixation that included atlantoaxial joint. No bone decompression was done.**

RESULTS: **At a follow-up of >30 months, both patients had significant neurologic recovery. Investigations at follow-up showed successful arthrodesis of treated spinal segments. Although incomplete, there was recovery in kyphosis.**

CONCLUSIONS: **Multisegmental spinal distraction and fixation can lead to reduction in kyphosis and relief from symptoms related to myelopathy. Role of spinal instability in general and atlantoaxial joint instability in particular in pathogenesis of cervical kyphosis need to be assessed on the basis of studies with a larger number of patients.**

INTRODUCTION

Kyphotic cervical spinal deformity is relatively rare in nonsyndromic pediatric age group patients. Treatment is usually indicated when kyphosis is associated with neurologic symptoms or deficits.[1-6] We report our experience of treating 2 patients with severe kyphotic cervical spinal deformity with multisegmental spinal distraction and fixation. Both patients had no known syndromal affection. Atlantoaxial joint was included in the fixation construct in both patients. No decompression by bone or soft tissue removal was done. The strategy and outcome of treatment are evaluated. Our literature survey did not reveal any report where such a method of multilevel spinal distraction and fixation was done and where atlantoaxial joint was included in the fixation construct for kyphotic cervical spinal deformity.

CASE ILLUSTRATION 1

A 5-year-old male child was born of a normal delivery and had normal milestones for his age. He did not have any syndromic affliction. For the last 1 year the parents noticed that the boy was having frequent falls and was unable to walk or run as before. He progressively worsened in his functional activities. When admitted for treatment, the child had inability to stand or walk and was carried by his father. He was unable to eat on his own, which he was able to do earlier. The child was bed-ridden and needed help for all activities. He was unable to hold his head straight. Neurologic examination showed severe spastic quadriparesis. The power was grade 2—3/5 in all 4 limbs. On clinical evaluation he had a Japanese Orthopaedic Association score of 6, and on Goel functional scale he was grade 5.[7] Radiologic investigations showed severe cervical kyphosis from C2—T1 levels (**Figure 1**). The C1—C2 joint formed the rostral point (point A) of the kyphotic arc. The tip of the dome of the arc was at the C4—C5 level (point B), and the T1 level was the inferior limit of the arc (point C).[6] There was narrowing of the cord at the C4—C5 level. The kyphotic angle was 89 degrees.

Operative Technique

The patient was placed in a prone position under Gardner-Wells traction. The traction aided in stabilization of the head. The cervical spine from the C1 to C7—T1 level was exposed. Severe instability was noted at the facetal articulation at all cervical levels. Manual manipulations of bones revealed atlantoaxial instability (type C atlantoaxial instability). The articular cavities were widely exposed, articular cartilage was denuded, and bone graft chips harvested from the iliac crest were stuffed in the joint spaces. C1—C2 lateral mass plate and screw fixation (as described by us in 1994),[8,9] C3—C4 and C4—C5 facetal distraction using intra-articular spacers and bone graft,[10] and C5—T1 transarticular screw fixation (as described by Camille in 1972)[11] were

Key words
- Atlantoaxial instability
- Cervical kyphosis
- Spinal instability

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Citation: World Neurosurg. (2017) 106:661-665.
http://dx.doi.org/10.1016/j.wneu.2017.07.055

*Journal homepage: **www.WORLDNEUROSURGERY.org***

*Available online: **www.sciencedirect.com***

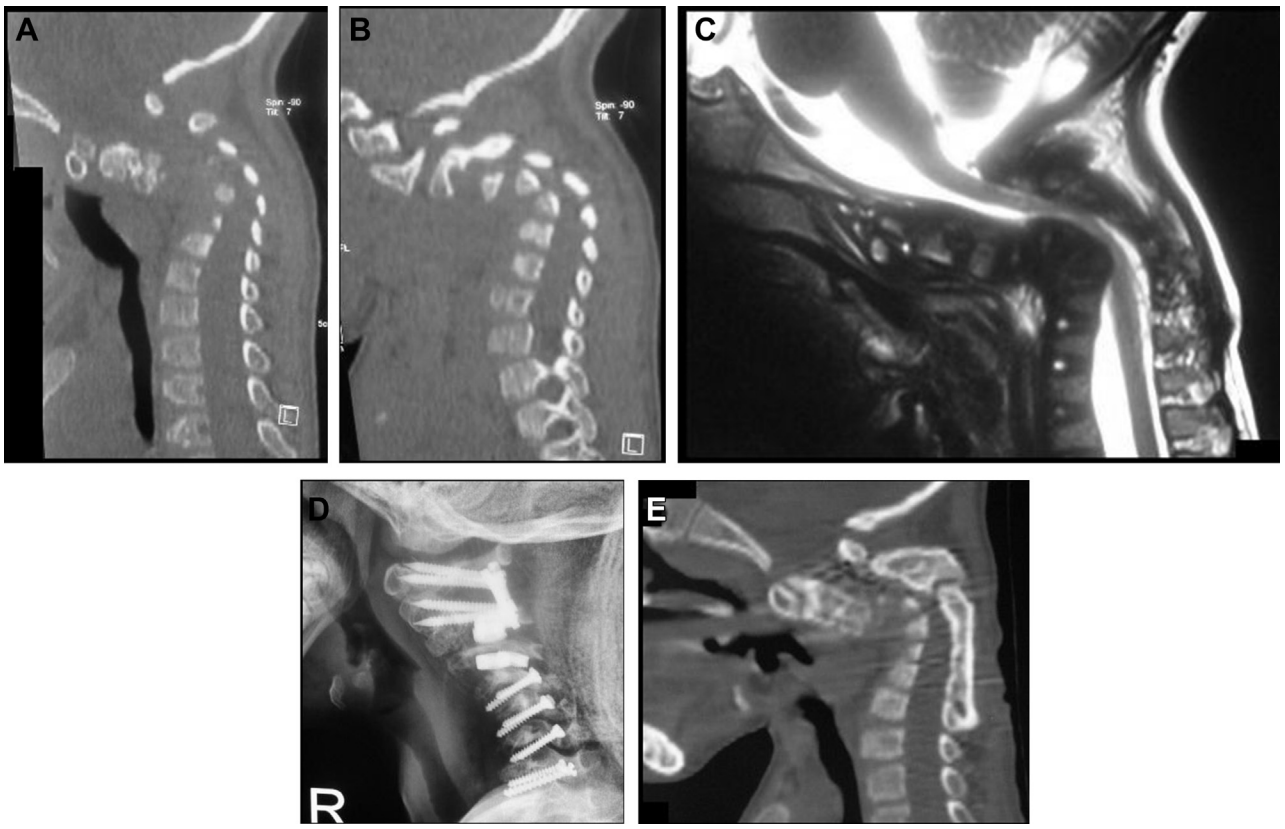

Figure 1. Images of a 5-year-old male. (**A**) Sagittal cut of computed tomography (CT) scan shows severe cervical kyphotic deformity. (**B**) Sagittal cut of CT scan showing the facets and the kyphotic deformity. (**C**) T2-weighted magnetic resonance imaging shows the cervical deformity. (**D**) Postoperative radiograph shows the fixation implants. (**E**) Delayed postoperative image of CT scan shows multilevel spinal fusion.

performed. Bone graft pieces were additionally placed over the midline after elaborately preparing the surface of the host bone by removing all soft tissues and drilling of the outer cortex.

Postoperative Course

The patient showed immediate postoperative neurologic improvement. The improvement was sustained and progressive. At a follow-up of 36 months the patient was able to walk unaided and could run at moderate speed. He was able to eat and dress himself and had started attending school. Computed tomography showed reduction in the kyphosis (kyphosis angle 60 degrees) with fusion from the C1–T1 levels.

Case Illustration 2

A 17-year-old male presented with progressively increasing weakness of all 4 limbs for the past 3 years. He had no syndromic affliction. He was unable to carry out all his routine activities independently and needed help. He needed support to walk. On neurologic examination he had spastic quadriparesis of grade 4/5. His Japanese Orthopaedic Association score was 9, and Goel clinical functional score was grade 5.[7] Radiologic investigations showed cervical kyphosis extending from the C2–C7 level (Figure 2). The rostral point of the kyphotic arc (point A) was at C1, the tip of dome (point B) was at C4, and the inferior point of the curve (point C) was at C7. A C1–C7 posterior fixation was performed, with surgical steps similar to that discussed for case 1. The patient's symptoms improved postoperatively, and he was well at a follow-up of 30 months. He was able to perform all routine work himself but could not be involved in any professional job. Delayed computed tomography scan showed a reduction in the cervical kyphosis with satisfactory posterior fusion.

DISCUSSION

Cervical kyphosis may be attributed to degenerative spondylosis, iatrogenic causes (e.g., postlaminectomy), trauma, infection, inflammatory arthritis (e.g., ankylosing spondylitis), and neuromuscular and metabolic disease.[4,12-16] Cervical kyphosis in pediatric patients is relatively rare. Such deformities are generally identified in children with syndromic disorder that leads to affection of multiple joints. Myelopathy as a result of such kyphotic deformity has only rarely been identified. The exact cause of development of kyphosis in both our patients can only be speculated. There was no definite evidence of injury to the head or spine or any known metabolic or genetic factor. Nutritional, infective, or environmental factors also seemed unlikely.

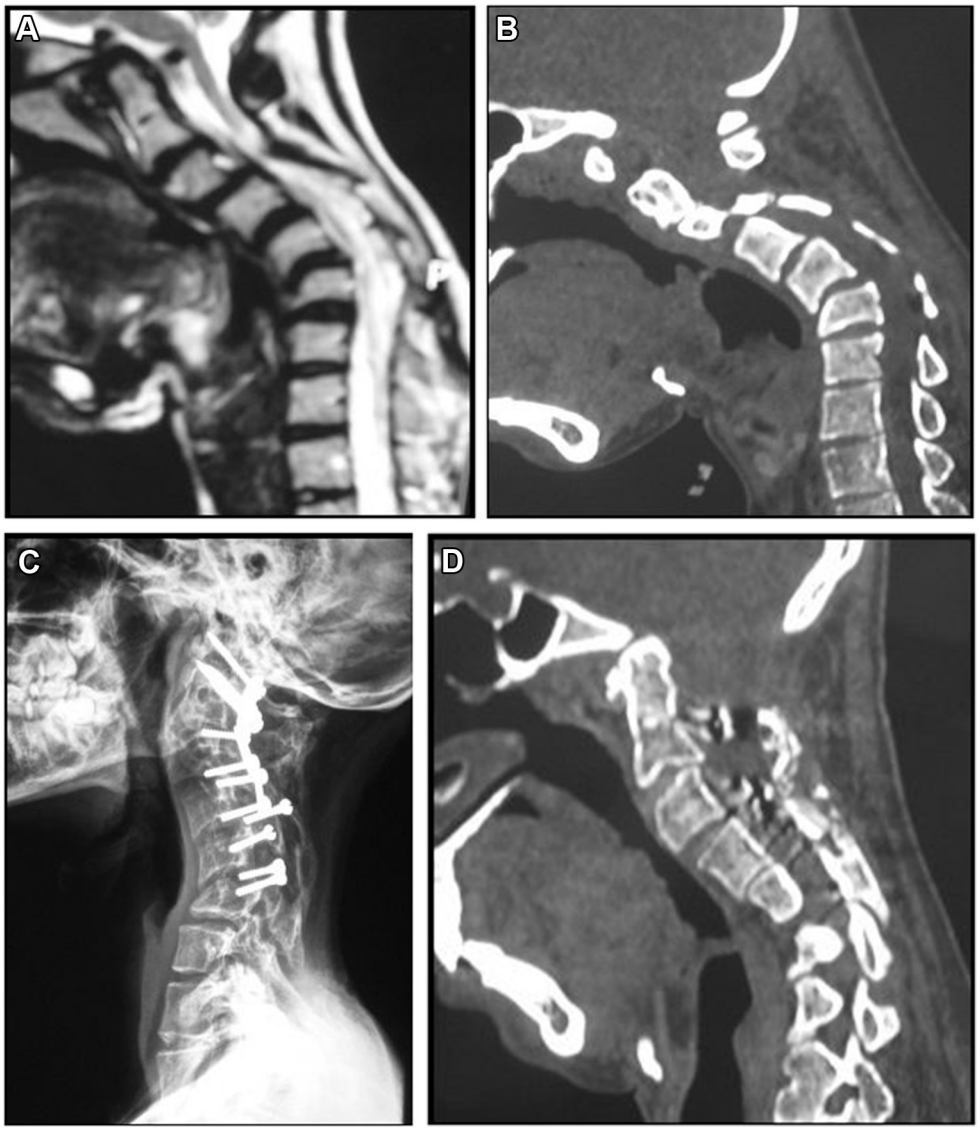

Figure 2. Images of a 17-year-old male. (**A**) T2-weighted magnetic resonance imaging shows cervical kyphosis. (**B**) Sagittal image of computed tomography (CT) scan showing kyphosis. (**C**) Plain radiograph showing multilevel fixation implants. (**D**) Delayed postoperative image of sagittal CT scan showing arthrodesis of treated segments.

Cervical kyphosis—related compression of the spinal cord by the posterior surface of vertebral bodies and consequent reduction of spinal canal dimensions has been identified as the major cause of myelopathy symptoms.[16-18] The existing surgical opinion is that symptomatic myelopathy in the presence of kyphotic deformity warrants a 2-pronged operation, the first involving wide decompression of the cord of compressing bone elements and the second involving stabilization. Numerous techniques of decompression of indenting bone and increasing the space for free traverse of the spinal cord have been discussed and range from multilevel discoidectomy, corpectomy, and facetectomy to laminectomy/laminoplasty.[1,3,5,19-22] Benzel opined that deformity correction is a viable treatment option in such cases and identified that correction of focal kyphosis has direct correlation with neurologic outcomes.[23] Instability of the spinal segments is considered to be an issue in the pathogenesis of kyphosis by some authors.[9] However, its role as a defining factor in development of kyphosis and generation of symptoms related to myelopathy has not been appropriately addressed or therapeutically exploited. More often stabilization is considered, as extensive bone removal for decompression has been generally associated with development of instability of the spine in the longer run. Multiple-stage operations have been advocated for

decompression, reduction, and subsequent stabilization in the treatment of cervical kyphosis.

Although the articular anatomic and morphologic peculiarities of the atlantoaxial joint permit wide-range circumferential movements, they also subject the joint to an exaggerated risk of instability. On the other hand, subaxial facet joints that permit restricted movements in anteroposterior and vertical perspectives are obliquely articulated. Consequently, the instability in the subaxial spine is essentially "vertical" in nature.[24] Atlantoaxial dislocation has been diagnosed essentially by a single parameter of abnormal alteration of atlantodental interval on dynamic flexion-extension images of the craniovertebral junction. We recently identified an alternative parameter to diagnose atlantoaxial instability by observing facetal malalignment on sagittal imaging with the head in neutral position.[25] Type A atlantoaxial instability occurred when the facet of atlas was dislocated anterior to the facet of axis. The atlantodental interval is increased in this type of dislocation, neural compression by the odontoid process is obvious, and the symptoms are relatively acute in nature. In type B atlantoaxial facetal dislocation, the facet of atlas is dislocated posterior to the facet of axis. In this situation, alteration of the atlantodental interval is frequently present but is not the hallmark, neural compression may not be evident, and the symptoms are generally chronic and longstanding. Type C atlantoaxial facetal instability happened when the facetal alignment was normal on imaging, but instability was identified by manual manipulation of bones during the surgical procedure. A high degree of clinical and radiologic suspicion in addition to subjective judgment based on surgical experience is mandatory to identify the presence of instability. Because the dural and neural compression of the craniocervical cord may not be evident on imaging in both types B and C, such dislocation has been labeled as central or axial atlantoaxial dislocation.[26] Central or axial atlantoaxial dislocation has been identified in cases associated with longstanding atlantoaxial instability like Group B basilar invagination, Chiari 1 malformation, syringomyelia, short neck, Klippel-Feil abnormality, and Hirayama disease.[7,27-31] We also identified such dislocation in cases with cervical kyphosis related to spondylotic degeneration.[6] We identified type C atlantoaxial instability to be associated with subaxial cervical joint/s instability in both presented cases. Accordingly, atlantoaxial and multisegmental subaxial spinal fixation was done. We also identified that it is not the cord compression or deformation that is the cause of symptoms in cases with kyphotic cervical spine but rather repeated microtrauma related to instability that causes symptoms.[32]

In our earlier study on cervical kyphosis that developed secondary to degenerative cervical spondylosis, we identified a "kyphotic arc."[6] Point A was the rostral end of the arc, point B was the tip of the dome of the arc, and point C was the inferior limit of the arc. All spinal segments between points A and C were unstable and needed fixation. Like in the presented cases, in cases where central or axial atlantoaxial instability was identified, the rostral limit of fixation was the atlantoaxial joint. The rostral limit of the arc was formed by the atlantoaxial joint in both presented cases. Additionally, type C instability of the atlantoaxial joint was identified during surgery. Despite the tremendous progress of computer-based imaging, instability of the spine as is evident during surgery by abnormal movements at the level of facets has not yet been clearly pictured. The diagnosis of instability is therefore based more on subjective and direct visual assessment of facetal stability during surgery than by radiologic imaging.

The technique of posterior atlantoaxial facetal fixation by the technique described by us in 1994[9] and transarticular screw fixation of the subaxial spine by technique described by Camille and Saillant[11] are relatively safe, strong, and remarkably quick when compared with techniques that involve anterior cervical multilevel bone decompression and subsequent fixation. In both discussed cases, intraoperative neural monitoring was not used but can certainly be of help and add to the safety of the procedure. Manual distraction of each spinal segment by opening of the joint, wide removal of articular cartilage, and introduction of bone chips with or without the facet distraction spacers[10] resulted in solid fixation, indirect spinal decompression, and reduction in the degree of spinal kyphosis.[8,9,33-35] The aim of surgery in our cases was focused on spinal stabilization and achieving solid bony arthrodesis and not primarily on reduction of kyphosis. The other advantage of lateral fixation was that in case the technique failed to produce the desired clinical outcome, both anterior and posterior midline bone structures could be decompressed at a subsequent second-stage surgery. As the follow-up in both our cases is relatively short, it is unclear if any anterior fixation should have been additionally performed or will be necessary or if new kyphotic deformity will develop below the fixated levels. Our successful clinical outcome in cases with kyphosis related to spinal degeneration and in the presented cases suggests that multisegmental spinal instability has a defining role in pathogenesis of cervical kyphosis and multisegmental spinal stabilization is the mode of treatment. However, more clinical experience is mandatory to confirm the proposed hypothesis.

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Received 14 June 2017; accepted 11 July 2017

Citation: World Neurosurg. (2017) 106:661-665.
http://dx.doi.org/10.1016/j.wneu.2017.07.055

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