Acute atlantoaxial facet locking in young children following a minor or an otherwise insignificant trauma has been reported to result in torticollis. Sir Charles Bell in 1830 and Corner in 1907 first described rotatory dislocation as a result of the atlantoaxial joint locking in an abnormal position. Wortzman and Dewar introduced the term atlantoaxial rotatory fixation in 1968. A number of possible causes for such an acute pathological event have been speculated. No genetic, chromosomal, or structural abnormality has been identified that could explain the clinical events. The acuity of the episode, young age of the patients, absence of pain and of any neurological symptom makes treatment for this condition a formidable problem. On the basis of our experience, we present a new classification of acute facet locking that is based on the type of abnormality of the facets. We propose an alternative form of surgical treatment and demonstrate with pictures postoperative realignment of dislocation. Although several modalities of surgical treatment have been suggested, our literature survey did not show any case in which there was radiological demonstration of postoperative reduction of dislocation into normal alignment in the clinical setting similar to that discussed in the presented report.

Methods

During the period between 2004 and 2009, 14 children, ranging in age from 7 to 12 years, were shown to have acute facet locking and were treated surgically. There were 6 boys and 8 girls. The symptoms started in all cases with a relatively insignificant trauma, such as tap on the back of the head (2 cases), carrying heavy school bag (2 cases), or a minor fall while playing (10 cases). Apart from neck deformity, there was no other significant symptom in any of these cases. We excluded patients in whom there was evidence of bone abnormality on imaging that could suggest a congenital anomaly or Down syndrome, or in whom there was any clinical evidence of a congenital or metabolic disorder.

Classification

The facet locking was divided into 2 types: rotatory (12 cases) and translatory (2 cases).

Rotatory Locking. Rotatory locking was diagnosed when the head was twisted to one side (torticollis) or was in a rotated position. Rotatory locking was “partial” (4 cases) when the facet of atlas of only one side was dislocated anteriorly over the facet of the axis. The facet on the contralateral side was not abnormally positioned and acted as a pivot for the dislocation (Fig. 1). Rotatory locking was “complete” (8 cases) when the facets of the atlas on both sides were dislocated over the facets of the axis in a rotatory fashion. The process resulted in an atlantal facet being positioned anterior to the axial facet on one side and posterior to it on the other side (Fig. 2).

Translatory Dislocation. Translatory dislocation was
diagnosed when both atlantal facets were dislocated anterior to the facets of the axis. Such a dislocation resulted in a fixed-flexion neck deformity. (Fig. 3) Although a posterior translation is also technically possible, no such cases were encountered.

**Imaging Investigation**

All patients underwent plain radiology, CT scanning with 3D reconstruction, and MR imaging. The degree of neck rotation or tilt in patients with rotatory dislocation was measured as an angle between a line that coursed from the nasion, tip of the nose, and symphysis menti to the horizontal. The angle was then expressed as the degree of tilt from the vertical (Table 1).

**Treatment Protocol**

All 14 patients were referred to our department after initial clinical observation, conservative treatment, and attempts at closed reduction had failed. The surgeries were conducted in our neurosurgical department between 35–140 days (mean 78 days) after the onset of torticollis. Six patients were initially treated elsewhere: attempts at closed reduction in which cervical halter traction (4 cases) and progressively increasing halo traction (2 cases) for a period ranging from 2 days to 2 months. Four patients underwent cervical collar therapy and 2 patients used firm 4-poster neck support. Two patients underwent clinical observation and no direct active treatment was attempted. In none of these patients was there any resolution in the degree of neck deformity. In 2 cases the deformity progressed during the period of conservative observation.

**Surgical Technique of Craniovertebral Realignment**

After induction of anesthesia, the patient is placed under cervical traction, the weights ranging from 3 to 6 kg. The head end of the table is raised approximately 35°.
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to provide countertraction by the body. This position assists in reducing the venous congestion in the operative field, particularly in the vein-rich region of the lateral masses. The exposure of the atlantoaxial joint in cases of facet joint malalignment is significantly more difficult and technically challenging than a normally aligned atlantoaxial joint encountered during the treatment of post-traumatic instability. However, we did observe that, in all cases, intraoperative traction resulted in an improvement in the alignment of the facets compared with their preoperative alignment. The atlantoaxial facet joints are widely exposed on both sides after sectioning of the large C-2 ganglion. Such a wide exposure provides an opportunity to observe the status of the facets and direct manipulation. The joint capsule is excised and the articular cartilage is widely removed using a microdrill. The joints on both sides are distracted using an osteotome. The flat edge of the osteotome is introduced into the joint, and it is then turned vertically to effect distraction and further manipulation. The sizes of the osteotomes can be varied depending on the local situation. The sequence of facet manipulation varies, but in general the more affected facet is treated first. When necessary, the distraction is done simultaneously on both sides. Although visualization of the region is possible with high-definition radiography or intraoperative CT scanning, such investigations were not performed. Direct observation of the position and alignment of the articular surfaces of the atlantal and axial facets is the key to facet realignment. Another indicator of successful realignment is the location of the tip of the C-2 spinous process in relationship to the midline of the arch of atlas. After sectioning of the C-2 ganglion, wide exposure of the region also provides an opportunity for identifying the most appropriate site for placement of C-2 screws, as we have previously discussed.8–12 Wherever possible, C-2 screw placement can even be transarticu-
lar (Fig. 2J). Such a screw placement provides additional stability to the joint. We labeled this method of fixation as “double-insurance” fixation. The use of neuronavigation can assist in screw insertion and avoiding the course of the vertebral artery. Such additional precautions are more relevant in cases with rotatory dislocation because the location of the vertebral artery can be altered due to rotation. Exact preoperative evaluation of the status of vertebral artery is mandatory. Corticocancellous bone graft material harvested from the iliac crest is packed into the joint in small pieces. Subsequent fixation of the joint, with the help of interarticular screws and a metal plate, provides a biomechanically firm fixation as well as sustained distraction and realignment. In cases in which the facet of atlas was dislocated anteriorly and manual realignment was incomplete, the screw (with the plate) was first inserted into the lateral mass of the axis and was tightened. The atlantal screw was then tightened over the fixed plate. This variation resulted in posterior movement of the facet of atlas. A similar technique of reduction has been described in the treatment of spondylolisthesis. Larger pieces of corticocancellous bone graft were placed in the midline over the arch of atlas and lamina of axis after appropriately preparing host bone. Postoperatively, traction is discontinued and the patient is placed in a 4-post hard cervical collar for 3 months, and all physical activities involving the neck are restrained during the period. After a 6-month period, all activities, including sports, were permitted in an unrestrictive manner in our patients.

Course of Surgery

In no case did we observe any ligament tear or any bone fracture that could be identified as a reason for preventing the normal joint movements. In at least 3 cases the ligaments in the region appeared to have an unusually gray hue and be swollen. Histological examination of the tissue was not carried out. There were no operative complications. There were no cases of infection, hardware failure, or need for reexploration. In response to a leading questioning, all children responded they had some degree of numbness in the back of the head that was related to the sectioning of the C-2 nerve root.

Follow-Up

All patients had immediate postoperative and sustained resolution of positional neck abnormality. The degree of neck deformity recovery, as measured by the angle described earlier (angle between a line that coursed from the nasion, tip of the nose, and symphysis menti to the horizontal), is detailed in Table 1 and illustrated in 2 cases (Figs. 1 and 2). The follow-up periods ranged from 3 to 52 months (mean 23 months). All neck movements, particularly rotation, were marginally limited in all cases, but they were significantly improved compared with the preoperative status. Because the implant material was made of stainless steel, postoperative MR imaging was not possible. The presence of metal artifacts on postoperative investigations made it difficult to identify bony arthrodesis. However, arthrodesis was assumed if the neck retained its normal alignment after a period of 6 months.
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Based on this parameter, successful fusion was achieved in all patients.

Discussion

Locking of the atlantoaxial facet joints has been identified to result in rotatory atlantoaxial dislocation. Such a dislocation has been identified more commonly in young children. Although several pathogenetic factors have been considered, the exact cause of the phenomenon is unclear. The dislocation is an acute event that is usually triggered by an episode of relatively minor trauma. Some patients have been identified as having throat and paranasal sinus infections or tonsillitis at the time of event. However, an exact correlation among possible infection, trauma, and atlantoaxial dislocation is only speculative. None of the patients in the present series had any evidence of significant focus of infection. Several other seemingly insignificant events have been associated with initiation of the dislocation. Rheumatoid arthritis, Down syndrome, Morquio disease, and various congenital cervical anomalies have also been associated with rotatory dislocation. However, to avoid overlap, we excluded patients in whom such an etiological factor might possibly be present. The higher incidence of rotatory atlantoaxial dislocation in children may be attributed to the shallower and more horizontally oriented joint surface, the relative elasticity of the ligaments, the not yet fully developed neck muscles, and a relatively large head. The acuity of the neck deformity in an otherwise healthy and neurologically normal child are the hallmarks of the problem. Pain is not a prominent presenting symptom. Patients present instead with the head tilted toward one side and rotated toward the other and in slight flexion. In the case of right unilateral anterior rotatory subluxation, one observes lateral bending of the head and neck to the right and rotation of the head to the left. Rotation of the head contralateral to the direction it faces is difficult; however, rotating it further in the direction toward which it faces is possible. In the acute setting, it is important to distinguish between rotatory locking and muscular torticollis. In rotatory dislocation, the sternomastoid spasm is on the side of the chin whereas in muscular torticollis the spasm is on the contralateral side.

The cause of the facet dislocation and its fixation in an abnormal position remains unknown, because no anatomical or autopsy evidence bearing on this question is available. Trauma or infection of the ligaments leading to laxity or a tear has been implicated as the most possible pathogenetic factor. Obstruction of movements related to impingement of fragments of articular capsule or ligaments on the joint cavity has been speculated. However, with our technique, which involved opening up of the joint and allowing for direct inspection, we found no actual tear or a broken fragment of ligament or bone within the joint cavity. We did identify alteration in texture and an unusually gray hue of the ligaments possibly related to inflammation in at least 3 cases. In the case involving translatory dislocation, extreme laxity or a tear of transverse ligament was possible. However, our investigations did not show this ligament clearly, and operative procedure did not provide an opportunity for direct inspection of this ligament.

A number of possible etiological factors have been posited in the literature. Wittek suggested that there is an effusion in the synovial joints that produces stretching of the ligaments. Coutts proposed the theory that synovial fringes, when inflamed or adherent, may block reduction, whereas Fiorrani-Gallotta and Luzzatti indicated that the deformity is due to rupture of one or both alar ligaments and the transverse ligament. Watson-Jones postulated the presence of a hyperemic decalcification with loosening of the ligaments. Grisel suggested that muscle contracture might follow an upper respiratory tract infection and be a factor. Hess and associates concluded that there is a combination of factors, including muscle spasm, that prevent reduction in the early stages. Wutzman and Dewar postulated there is damage to the atlantoaxial articular processes of an unknown nature. Fielding and Hawkins suggested that reduction of the dislocation is probably obstructed in the early stages by swollen capsular and synovial tissues and by associated muscle spasm. If the abnormal position persists due to a failure to achieve reduction, ligament and capsular contractions develop and cause fixation.

Three-dimensional CT scans are useful for diagnosing facet locking, as the atlas, axis, and their positions can be accurately seen. Although it is possible to determine the degree of neck angulation based on CT scans, the exact parameters necessary have not been clearly described in the literature. Moreover, a uniform CT scan pattern is necessary for validity of such an evaluation. Dynamic scans have been advocated for analyzing the abnormality in relative motion between the atlas and axis, but this investigation was not performed. The basic diagnostic criterion for this condition is a persistent asymmetrical relationship between the dens and the articular masses of the atlas not correctable by rotation.

A number of cases of reducible atlantoaxial facet subluxation can be treated nonoperatively using closed reduction. The patient must undergo a trial of conservative

### TABLE 1: Degree of torticollis in patients with atlantoaxial facet locking before and after surgery

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Neck Tilt (°)</th>
<th>Recovery (°)</th>
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<td>Postop</td>
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treatment. Due to the relative rarity of the dislocation and only limited reports in the literature, it is not possible to provide an approximate incidence of the event or describe which cases with which injury will recover after conservative or nonoperative treatment. On the basis of reducibility of the dislocation, facet locking can be subdivided into reducible or irreducible types. Some authors advise the use of direct cervical traction or a progressive halo brace traction for varying periods. An analysis of the result of traction and modifying the surgical strategy is accordingly carried out. If during the period of conservative therapy, the rotatory dislocation diminishes and remains reduced on dynamic imaging, there is no need for surgery. Immobilization presumably allows the stretched or edematous ligaments to heal and to return to their normal length and function. Full, pain-free motion of the neck should be restored before external immobilization is discontinued. However, we observed that most children tolerated traction poorly, and in none of the cases in the present series did traction result in a permanent repositioning of the facet joints to a normal alignment. Based on our experience, we found that failure of a 2-month period of conservative treatment to restore normal neck stature arguably indicates that surgical realignment should be considered, as proposed. It appears that too much delay can result in local bone fusion that can affect surgery. In all of our patients, closed reduction or a conservative observation had already failed. Although the long-term outcome of untreated rotatory atlantoaxial dislocation has not been elaborately described in the literature, one can expect to find fusion to have occurred in the dislocated position. Delayed lower and midspinal deformities are possible, which will contribute to a patient's head facing straight ahead.

Both rotatory and translatory dislocations result in an alteration in the course of the vertebral artery. Theoretically, acuity of the event can lead to abnormal stretching or even vertebral artery occlusion. However, the suppleness of tissues in children probably allows space for accommodation. Apart from the alteration in the vertebral artery course due to deformity, we did not identify any structural variation in the arterial foramen or any abnormality in the artery's dimensions. The technique distraction-induced of realigning the osseous structures provided an opportunity of return of the vertebral artery into its normal course.

In a recent review Pang and Li studied the normal biomechanics of atlantoaxial rotation, constructed a physiological motion curve for relative motions between these joints, and introduced new diagnostic criteria for rotatory atlantoaxial dislocation. Fielding and Hawkins described treatment guidelines based on their classification of rotary subluxation of the atlantoaxial joint. Depending on the nature of facet dislocation, we classified the deformity into 2 groups: rotatory dislocation and translatory dislocation. This classification varies from those previously described in that it includes translatory dislocation in the subgroup. Considering that facet malalignment was present as an acute phenomenon in a similar clinical setting in both groups of patients, broadening the classification of neck deformity appeared to be reasonable. The rotatory dislocation was further classified into complete rotatory dislocation, where facets on both sides were not properly aligned, and an incomplete rotatory dislocation, where there was only a unilateral facet malalignment, the facets being positioned normally on the contralateral side. The normally aligned facet acts as a pivot on which the contralateral side rotates. Such a rotatory dislocation has been described previously. On physical observation, the degree of torticollis was not remarkably different in the 2 groups.

There are only isolated reports mentioning surgical treatment strategy in cases involving locked atlantoaxial facet joints. Schmidek et al. used a transoral route and Crockard and Rogers employed an extreme lateral approach to remove obstructing ligamentous or bone structures within the joint. Goto et al. released the atlantoaxial lateral facet joint via a transoral route and subsequently performed posterior fixation using interlaminar clamps. Some authors have discussed the usefulness of fixation when the neck is abnormally positioned. They observed that segmental fixation of the dislocation, even in a dislocated position, allows the subaxial spine to move better, and torticollis can be expected to resolve. In a literature survey we found no report of a direct physical manipulation of the facets to effect realignment. Our technique involves sectioning of the C-2 ganglion, creating a wide exposure, opening up of the atlantoaxial joint, and unilateral or bilateral manipulation of the atlantoaxial facet joints. We observed that external traction may not be successful in relocating the facets, as traction had more effect on the lower portion of the neck than the site of problem in the region of the joint. Direct distraction of the facet joints appeared to be the most important component of the operation. Further manipulations of the position of the facets and their realignment were then possible. Opening up of the articular capsule, removal of the articular cartilage, and packing of the bone graft into the articular cavity resulted in dysfunction of the joint and set the stage for its ultimate fusion. Screw fixation of bones that were otherwise normal was strong enough to sustain the vertical, transverse, and rotatory strains of the most mobile region of the spine. We did not locate any broken pieces of bone or cartilage or find any evidence of infection in the region of the joint in any case, although this has been reported.

Our encouraging experience in a relatively small number of patients suggests the validity of our procedure and its positive effects on clinical outcome. Our long-term experience with opening of the atlantoaxial joint and with facet manipulation in the treatment of atlantoaxial dislocation and basilar invagination assisted in the overall operation. Although technically challenging and anatomically precise, the procedure results in excellent restoration of the facet joint alignment and neck shape and satisfactory neck movements. Because the fixation of the craniovertebral junction was segmental at atlantoaxial joint, the rest of the neck and even the occipitoaxial joint provided an avenue for neck movements. Although quantitative assessments were not possible, we did note restriction of neck flexion and rotation compared with healthy children. In at least 3 cases, restriction of neck movements appeared more significant than in patients treated with.

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the same technique for mobile and reducible atlantoaxial dislocations. The exact cause of this variation is unclear, but structural alterations in cases involving facet dislocation appear to be one possibility.

Conclusions

Locking of the facet joints and related neck deformity can be a physically crippling disorder. The difficulties associated with exposure and manipulation of the atlantoaxial joint for reduction of the dislocation, particularly in neurologically intact patients, make surgery a formidable technical challenge. The proposed operation necessitates fixation of the joint. A technique that will reduce dislocation and permit movements of the atlantoaxial joint needs to be identified.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Goel. Acquisition of data: both authors. Analysis and interpretation of data: both authors. Drafting the article: both authors. Critically revising the article: both authors. Reviewed final version of the manuscript and approved it for submission: both authors. Study supervision: Goel.

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Please include this information when citing this paper: published online December 17, 2010; DOI: 10.3171/2010.9.SPINE1010. Address correspondence to: Atul Goel, M.Ch., Department of Neurosurgery, K.E.M. Hospital and Seth G.S. Medical College, Parel, Mumbai 400012, India. email: atulgoel62@hotmail.com.