J Neurosurg Spine 11:9–14, 2009

Vertical mobile and reducible atlantoaxial dislocation

Clinical article

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Object. The authors' experience with treatment of 8 patients with "vertical mobile and reducible" atlantoaxial dislocation is reviewed. The probable pathogenesis, radiological and clinical features, and management issues in such cases are discussed.

Methods. Between January 2006 and March 2008, 8 patients who presented with vertical mobile and reducible atlantoaxial dislocations were treated at the Department of Neurosurgery at King Edward Memorial Hospital in Mumbai, India. The vertical atlantoaxial dislocation/basilar invagination reduced completely on extension of the neck, with no need of any cervical traction. According to the extent of superior migration of the odontoid process, and measurements based on the vertical atlantoaxial instability index, the dislocation was graded as mild, moderate, or severe. All patients were treated using the C-1 lateral mass and C-2 pars plate and screw method of fixation.

Results. The study group was composed of 5 male and 3 female patients (mean age 24 years, age range 8–54 years). All patients presented with the physical features of short neck, torticollis, pain in the nape of the neck, and varying degrees of quadriparesis. In 6 patients there was a history of trauma prior to the onset of major neurological symptoms. The dislocation was mild in 3 cases, moderate in 1, and severe in 4. All patients had clinical neurological improvement following surgery. The follow-up duration ranged from 4 to 30 months (mean 18 months).

Conclusions. Vertical mobile and reducible atlantoaxial dislocation is a discrete clinical entity. Abnormal inclination and incompetence of the facet joint appears to be the primary causative factor that resulted in vertical dislocation or basilar invagination. Posterior fixation in the reduced dislocation position forms the basis of treatment.

(DOI: 10.3171/2009.3.SPINE08927)

Key Words • **atlantoaxial dislocation** • **basilar invagination** • **craniovertebral anomaly**

In an earlier report we classified basilar invagination into 2 groups based on the presence (Group A) or absence (Group B) of craniovertebral instability.[4,5] In the present report, we discuss 8 patients who had Group A basilar invagination in which there was complete reduction of the invagination on extension of the head, with no need of any cervical traction. We have labeled this group of patients as having "vertical mobile and reducible" atlantoaxial dislocation. Considering that the imaging characteristics with the head in the flexed position are of Group A basilar invagination, the term "mobile and reducible basilar invagination" can also be used to classify the clinical condition. We could not locate any report from the literature describing such a clinical condition.

Methods

During the period between January 2006 and March 2008, 8 consecutive patients with vertical mobile and reducible atlantoaxial dislocation underwent surgery at our hospital. There were 5 male and 3 female patients who ranged in age from 8 to 54 years (mean 24 years). A short neck and a range of torticollis were constant clinical findings in all patients and had been present since early childhood. In 6 patients, there was a history of a relatively moderate degree of trauma at the time of onset of major neurological symptoms. The torticollis worsened after the injury in all cases. The history of trauma ranged from 15 days to 4 years (mean 6 months) prior to diagnosis and treatment. All patients had varying degrees of neck pain, neck muscle spasm, and spastic quadriparesis. Sensory symptoms were relatively mild and predominantly included bilateral upper- and lower-extremity paresthesias and kinesthetic sensation deficits (Table 1).

All patients underwent preoperative MR imaging, dynamic CT scanning, and plain radiography, and we also obtained dynamic CT scans and plain radiographs postoperatively (Figs. 1–3). Because stainless-steel plates and screws were used during surgery, postoperative MR imaging was not possible. Three patients experienced assimilation of atlas. The atlantoaxial articulation was inclined in all patients, as is seen in Figs. 2D and 3C.

We chose the more commonly used parameters of

Abbreviation used in this paper: VA = vertebral artery.

the clival line described by Wackenheim, and the line of foramen magnum described by McRae to evaluate basilar invagination in these cases. The tip of the odontoid process was above the Wackenheim clival line and the McRae foramen magnum line in all patients when they flexed the neck.[1,12,14] On extension of the neck, the tip of the odontoid process was below these lines. To measure the vertical instability in these cases, we previously proposed a vertical atlantoaxial instability index.[11] The index basically measures the vertical relationship of the atlas and axis. A horizontal line is drawn through the lower endplate of the axis. A second line is drawn parallel to this and tangential to the lower border of the anterior arch of the atlas. Furthermore, a third line is drawn parallel to these lines and tangential to the superior margin of the dens. The shortest distance between the first 2 lines (x) is divided by the shortest distance between the first and third lines (y). Depending on the severity, the vertical instability was graded from 1 to 3, or mild, moderate, and severe. We used the same parameter with the neck of the patient in the flexed position to evaluate the extent of vertical instability. Accordingly, 3 patients had mild, 1 had moderate, and 4 had severe vertical dislocation.

TABLE 1: Principal presenting clinical features in 8 patients with unstable atlantoaxial dislocation

	No. of Patients (%)	
Clinical Features	Preop	Postop
neck pain	8 (100)	0
torticollis & restricted neck movements	7 (88)	2 (25)
weakness		
able to walk unaided	5 (63)	8 (100)
needed support to walk	3 (38)	0
unable to walk even w/ support	0 (00)	0
sensations		
normal sensations	4 (50)	7 (88)
kinesthetic sensations affected	4 (50)	6 (75)
spinothalamic sensations affected	3 (38)	4 (50)
lower cranial nerves affected	1 (13)	0
respiratory distress	1 (13)	0

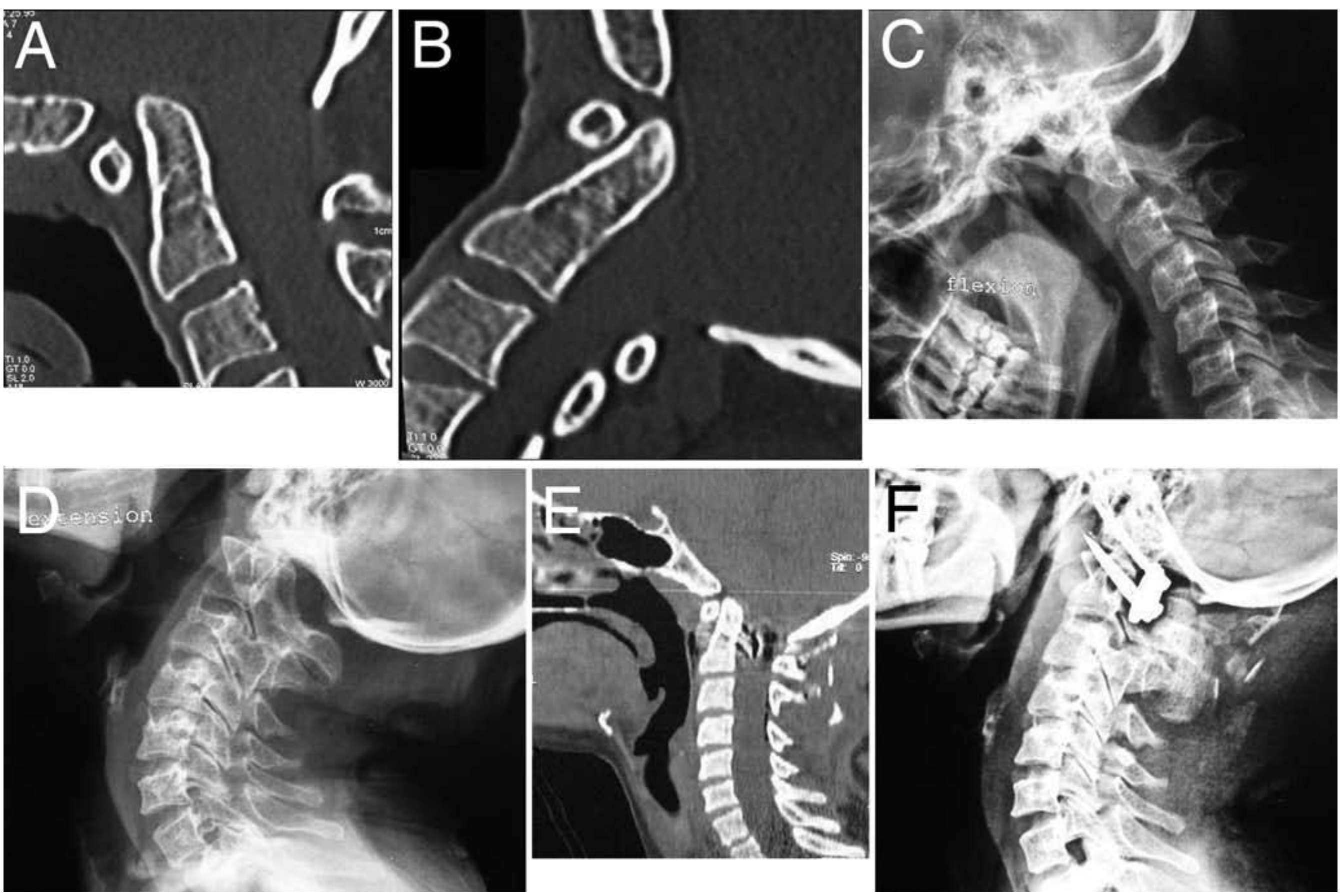

Fig. 1. Neuroimages obtained in a 54-year-old woman with unstable basilar invagination. A: Sagittal CT scan reconstruction with head in flexion, showing vertical dislocation (or basilar invagination). B: Sagittal CT scan obtained with the head in extension, showing reduction of the dislocation. C: Radiograph of the craniovertebral junction with the head in flexion, showing evidence of basilar invagination and atlantoaxial dislocation. D: Radiograph obtained with the head in extension, showing reduction of the dislocation. E: Postoperative CT scan showing fixation of the atlantoaxial dislocation in reduced position. F: Postoperative radiograph showing fixation with plate and screws.

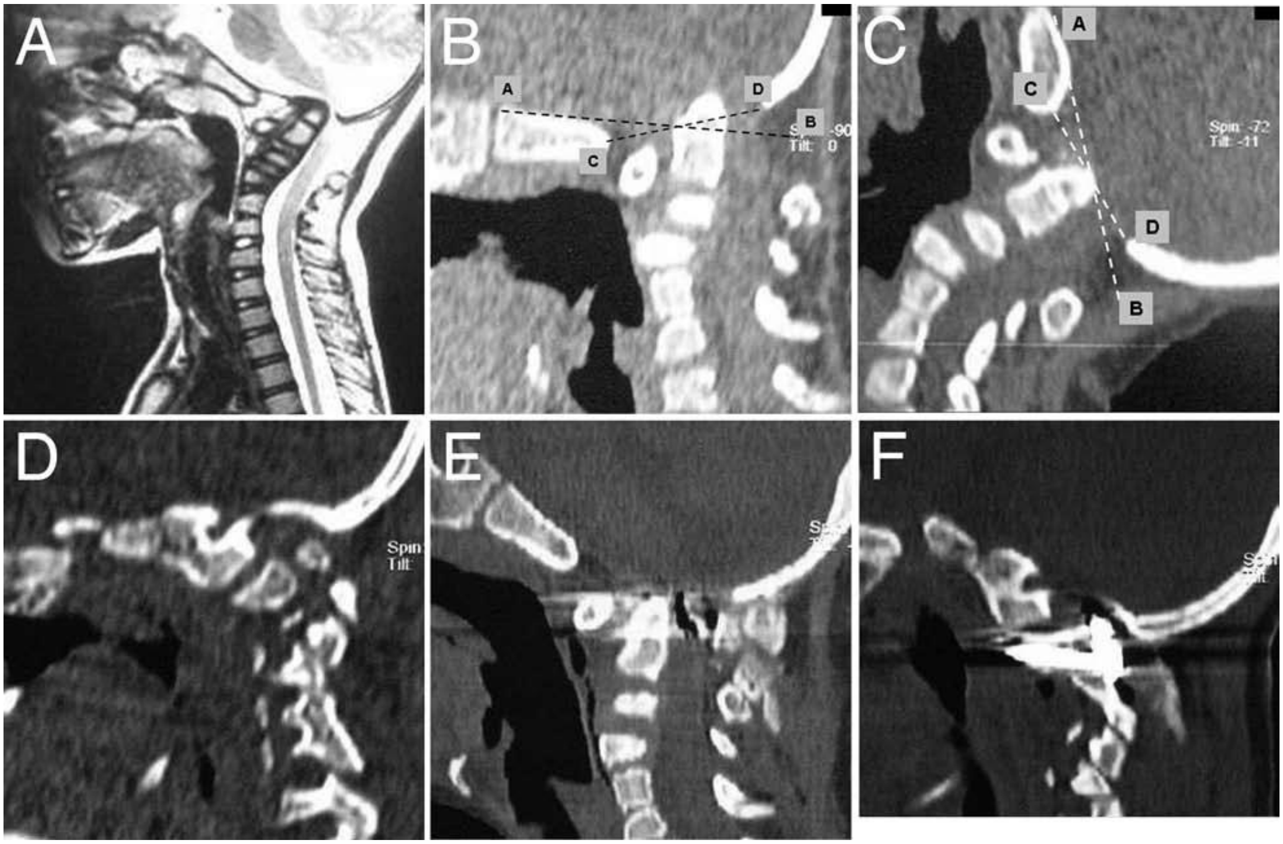

Fig. 2. Neuroimages obtained in an 8-year-old girl with unstable basilar invagination. A: Sagittal T2-weighted MR image showing extensive basilar invagination and cervicomedullary spinal cord compression. B: A CT scan reconstruction with the head in flexed position showing severe basilar invagination and vertical atlantoaxial dislocation. The Wackenheim clival line *(A – – – B)* and the McRae foramen magnum line *(C – – – D)* show the relationship with the odontoid process. C: A CT scan with head in extension showing reduction of dislocation. Note the altered relationship of the odontoid process with the McRae line and the Wackenheim line. D: A CT scan with the sagittal cut passing through the atlantoaxial joint. An abnormal oblique orientation of the joint can be appreciated. E: Postoperative CT scan showing fixation of the dislocation by using the lateral mass plate and screw fixation method. Reduction of the vertical dislocation can be appreciated. F: Postoperative sagittal CT scan showing plate and screw fixation and realignment of the joint.

Surgical Technique

The basic steps of the surgery are the same as those discussed in our previous papers, and are briefly summarized here.[4,6,7] Short neck and torticollis, presence of assimilation of the atlas, and abnormal inclination of the lateral masses of both atlas and axis made exposure of and insertion of screws into the lateral mass for fixation relatively tedious and complex. Cervical traction is given prior to induction of anesthesia, and the weights are progressively increased to ~ 5–8 kg or one-sixth of the total body weight. With the traction in place, the patient is placed prone with the head end of the table elevated to ~ 35°. Cervical traction stabilizes the head in an optimally reduced extension position and prevents any rotation. The traction also ensures that the weight of the head is directed superiorly toward the direction of the traction and that the head is essentially floating in air so that pressure over the face or eyeball by the headrest is avoided. Elevation of the head end of the table, which acts as a countertraction, helps in reducing venous engorgement in the operative field.

The suboccipital region and the upper cervical spine are exposed through an ~ 8-cm longitudinal midline skin incision centered on the spinous process of the axis. The spinous process of the axis is identified, and the attachment of paraspinal muscles to it is sharply sectioned. The large second cervical ganglion is closely related to the VA on its lateral aspect. It is first exposed widely and then sectioned sharply. Although it is possible to displace the ganglion superiorly or inferiorly, it was seen that in the relatively more complex anatomy in this group of patients, ganglion sectioning provided a panoramic view to the joint and the lateral masses, and the entire procedure could be completed under direct vision. Control of venous bleeding, which is sometimes considerably troublesome, is also superior after such an exposure. Packing of the region with Surgicel and Gelfoam can assist in the control of venous bleeding. The joint capsule is cut sharply, and the articular surfaces of the joint are exposed. The articular surfaces were drilled and denuded of their cartilage with a microdrill, and pieces of bone harvested from the iliac crest were stuffed into the joint space. Actual VA

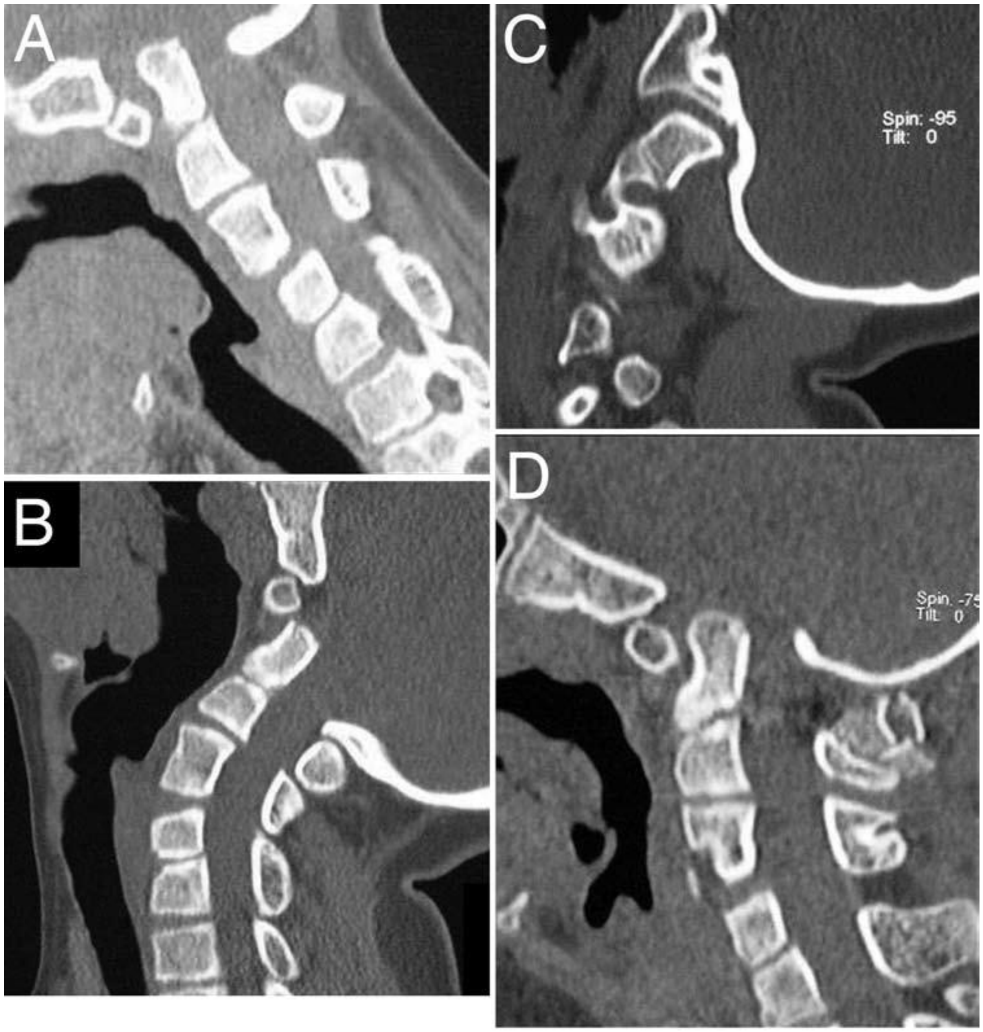

Fig. 3. Neuroimages obtained in a 12-year-old boy with unstable basilar invagination. A: Sagittal CT scan with the head in flexion, showing the presence of basilar invagination. B: A CT scan with the head in extension position showing reduction of the basilar invagination. C: A CT scan with the cut traversing the atlantoaxial joint. An abnormal configuration of the joint can be appreciated. D: Postoperative CT scan showing fixation in a partially reduced position.

exposure is unnecessary either lateral to the pars of the axis or superior to the arch of the atlas.

Metal screws are implanted into the previously created guide holes in the lateral mass of the atlas and axis through a 2-hole (~ 2-cm-long), adequately selected and shaped, double-compression stainless-steel (or titanium) plate. First, a screw is placed into the atlas. It is directed at an angle of ~ 15° medial to the sagittal plane and ~ 15° superior to the axial plane. The preferred site of screw insertion is at the center of the posterior surface of the lateral mass, 1–2 mm above the articular surface. Whenever necessary, careful drilling of the inferior surface of the lateral aspect of the posterior arch of the atlas in relation to its lateral mass can provide additional space for the placement of the plate and screw implantation. Screw implantation in the axis needs to be precisely directed, because of the intimacy of VA relationships. Presence of torticollis makes the screw insertion more tedious. The quality of cancellous bone in the lateral masses of the atlas and axis in the proposed trajectory of screw implantation was good, providing an excellent purchase for the screw, and avoided the VA.

Large pieces of corticocancellous bone graft from the iliac bone are then placed in the adequately prepared receptor area of the posterior arch of the atlas and the lamina of the axis. After the wound is closed, cervical traction is removed. The patients are mobilized within a few days of surgery. They wear a hard cervical collar for 3 months, and all physical activity related to neck movements is restricted during that period.

Results

The follow-up ranged from 4 to 30 months (mean 18 months). All patients improved radiologically and clinically following surgery (Table 1). The fixation of the atlantoaxial region was in the reduced position. On direct questioning, all patients reported a patch of numbness in

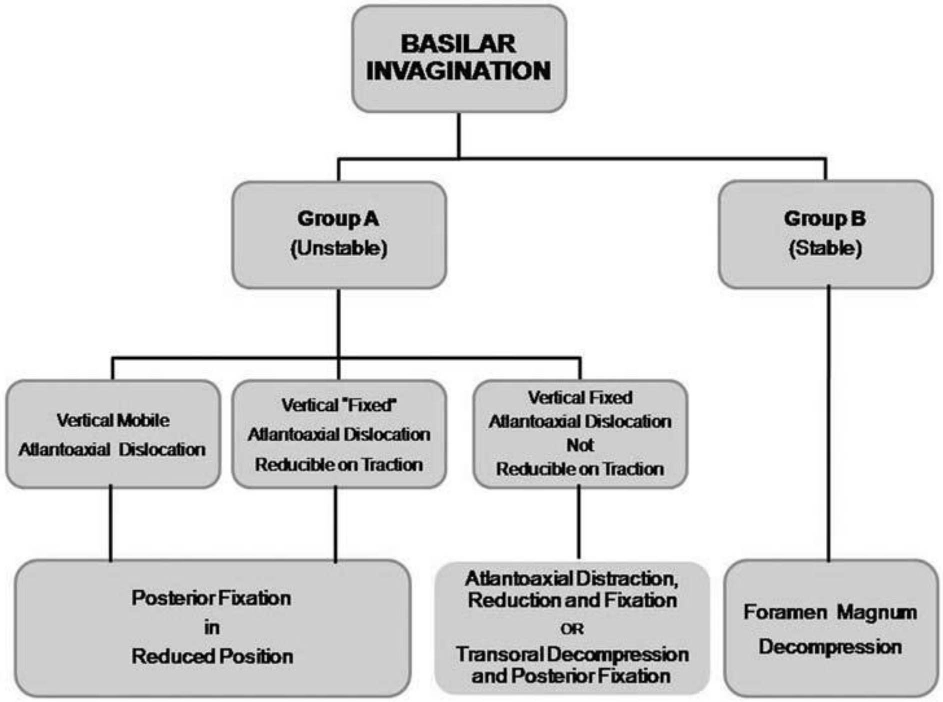

Fig. 4. Algorithm for management of basilar invagination cases.

the suboccipital region related to sectioning of the C-2 nerve, but none was unduly concerned about this symptom.

Discussion

In 2005, we classified basilar invagination into 2 groups.[4,5] Group A basilar invagination is found in cases where the tip of the odontoid process invaginated into the foramen magnum. Essentially, in this group of patients the odontoid process migrated away from the anterior arch of the atlas, and there was clinical and radiological evidence of craniovertebral instability. The definition of basilar invagination as prolapse of the cervical spine into the base of the skull, as suggested by Von Torklus and Gehle[16] and Grawitz,[8] is suitable for this group of patients. Group B basilar invagination exists when the alignment of the odontoid process and clivus is maintained despite the presence of basilar invagination and other associated anomalies. There is no instability in these patients.

After our recognition of the presence of mobile and reducible vertical dislocation, and after conducting dynamic CT scanning, we identified the 8 cases presented in this study from among the 64 patients with Group A basilar invagination who were seen to be experiencing vertical mobile and reducible dislocation during the period. It is critical to differentiate this group of patients from the other cases of Group A basilar invagination in which "fixed" atlantoaxial dislocation is diagnosed, because the treatment of the 2 clinical entities is discrete.

Although several authors consider congenital basilar invagination as a fixed anomaly, others have identified it to be a result of vertical craniocervical instability.[2,4,5,9,13,14] Essentially, it appears that patients with Group A basilar invagination can be subclassified in either a fixed atlantoaxial dislocation category (where the basilar invagination does not disappear with neck movement) or vertical mobile and reducible atlantoaxial dislocation (where the basilar invagination disappears on extension of neck). The more accepted method of treatment of the former subgroup (Group A basilar invagination with fixed atlantoaxial dislocation) is transoral decompression of the odontoid process and subsequent craniovertebral stabilization[2–5,13] or the recently described atlantoaxial joint distraction (with or without the assistance of metal spacers), reduction of the dislocation, and subsequent fixation.[4] However, in the latter subgroup (Group A basilar invagination with vertical mobile and reducible atlantoaxial dislocation), fixation of atlas and axis in the reduced position suffices, and distraction of the facets is not necessary. Based on the identification of the presented subgroup of basilar invagination, a treatment algorithm for patients with basilar invagination is presented in Fig. 4. This algorithm has philosophical similarities with that described by Menezes[13] and colleagues.[15]

Vertical instability of the atlantoaxial joint has been identified and more often linked with cranial settling associated with rheumatoid arthritis. Vertical instability has also been linked by some authors as a cause of basilar invagination secondary to congenital anomaly in the region. Reduction of basilar invagination when cervical traction is instituted has been observed by us and by others.[2,4,5] Such a reduction of basilar invagination suggests the presence of vertical instability. We had earlier identified a relatively inclined profile of the facets of atlas and axis to be the primary cause of basilar invagination. Progressive slip of the facet of C-2 over C-1 could be the cause of listhesis of C-1 over C-2, a phenomenon that eventually results in basilar invagination.[10] In all the presented cases, the atlantoaxial joint was markedly inclined, an anomaly that appeared to be primarily responsible for incompetence of the lateral masses and an abnormal vertical mobility.

The Wackenheim clival line and the McRae line of foramen magnum were used to evaluate the basilar invagination or the superior migration of the tip of the odontoid process in relationship to the clivus or to the anterior arch of the atlas.[12,14] To grade the degree of vertical dislocation, we used our previously described vertical atlantoaxial instability index.[11] The majority of patients were either adolescent or middle aged. The dislocation was generally more severe in younger patients. Because the compression of the cervicomedullary spinal cord is not as acute as is seen in horizontal atlantoaxial dislocation, the symptoms are relatively mild and long-standing. Pain in the nape of the neck, spasm of neck muscles, and short neck formed the principal symptom complex.

We used our technique of plate and screw fixation of the lateral mass of atlas and axis for stabilization of vertical atlantoaxial dislocation.[6,7] Bicortical purchase of screws in both atlas and axis was achieved in all cases. Although identified by some, in our present series and also in our earlier experience, damage to the carotid artery by a too deeply penetrating screw has not occurred. On direct intraoperative observation, it was clear that the joint was active and mobile and that the articular surfaces were functional. The articular surfaces were drilled and denuded of their cartilage, and iliac crest bone graft was introduced into the joint cavity to effect bone fusion. Immediate postoperative and long-term results following successful stabilization by surgery were extremely gratifying.

Conclusions

Considering the number of cases seen by us in a relatively short period of time, it appears that vertical mobile and reducible atlantoaxial dislocation is a discrete clinical entity. The movements of the odontoid process in a vertical plane result in indentation of the cervicomedullary cord and related symptoms. From our observations, it appears that all patients with basilar invagination should undergo dynamic flexion-extension CT scanning to determine the presence or absence of vertical instability and reducibility. Stabilization and fixation in reduced and anatomical position resulted in cure.

Disclaimer

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

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Manuscript submitted December 11, 2008.
Accepted March 5, 2009.
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