Is atlantoaxial instability the cause of Chiari malformation? Outcome analysis of 65 patients treated by atlantoaxial fixation

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OBJECT Understanding that atlantoaxial instability is the cause of Chiari malformation (CM), the author treated 65 patients using atlantoaxial stabilization. The results are analyzed.

METHODS Cases of CM treated using atlantoaxial fixation during the period from January 2010 to November 2013 were reviewed and analyzed. Surgery was aimed at segmental arthrodesis.

RESULTS The author treated 65 patients with CM in the defined study period. Fifty-five patients had associated syringomyelia. Forty-six patients had associated basilar invagination. Thirty-seven patients had both basilar invagination and syringomyelia. Three patients had been treated earlier using foramen magnum decompression and duraplasty. According to the extent of their functional capabilities, patients were divided into 5 clinical grades. On the basis of the type of facet alignment and atlantoaxial instability, the patients were divided into 3 groups. Type I dislocation (17 patients) was anterior atlantoaxial instability wherein the facet of the atlas was dislocated anterior to the facet of the axis. Type II dislocation (31 patients) was posterior atlantoaxial instability wherein the facet of the atlas was dislocated posterior to the facet of the axis. Type III dislocation (17 patients) was the absence of demonstrable facet alignment and was labeled as “central” atlantoaxial dislocation. In 18 patients, dynamic images showed vertical, mobile and at-least partially reducible atlantoaxial dislocation. All patients were treated with atlantoaxial plate and screw fixation using techniques described in 1994 and 2004. Foramen magnum decompression or syrinx manipulation was not performed in any patient. Occipital bone and subaxial spinal elements were not included in the fixation construct. One patient died, and death occurred in the immediate postoperative phase and was related to a vertebral artery injury incurred during the operation. One patient had persistent symptoms. In the rest of the patients there was gratifying clinical improvement. More remarkably, in 7 patients, the symptoms of lower cranial nerve paresis improved. No patient worsened in their neurological function after surgery. Reductions in the size of the syrinx and regression of the CM were observed in 6 of 11 cases in which postoperative MRI was possible. During the follow-up period, there was no delayed worsening of neurological function or symptoms in any patient. Sixty-three patients improved after surgery, and the improvement was sustained during the average follow-up period of 18 months.

CONCLUSIONS On the basis of outcomes in this study, it appears that the pathogenesis of CM with or without associated basilar invagination and/or syringomyelia is primarily related to atlantoaxial instability. The data suggest that the surgical treatment in these cases should be directed toward atlantoaxial stabilization and segmental arthrodesis. Except in cases in which there is assimilation of the atlas, inclusion of the occipital bone is neither indicated nor provides optimum stability. Foramen magnum decompression is not necessary and may be counter-effective in the long run.

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CHIARI malformation (CM) is frequently associated with basilar invagination and syringomyelia.^{1–4,7,11,13–15} Each of these 3 clinical entities can be present in isolation, but they are frequently identified synchronously. Surgical treatment has been evolving for

decades.^{1–4,8,14,15,28,35} A wide range of therapeutic protocols have been described. It can be safely said that the number of forms of treatment described or conducted by various surgeons is probably equal to the number of surgeons performing the surgery.³⁵ Each surgeon has some

ABBREVIATIONS CM = Chiari malformation; CN = cranial nerve; JOA = Japanese Orthopaedic Association; VAS = visual analog scale.

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special trick that he or she finds unique and superior. The very fact that no definite treatment strategy has convincingly found universal acceptance or has produced uniformly gratifying results suggests that the pathogenesis and pathophysiology of the disease process are still unclear and, to say the least, riddled with controversies and opinions. Here, the management of CM associated with or without basilar invagination and with or without syringomyelia is evaluated. All patients were treated primarily with atlantoaxial fixation. The surgical strategy was based on the understanding that atlantoaxial dislocation is the primary initiator of the entire process of structural bone and soft tissue malformation. The nature and probably the subtlety of the dislocation in some cases provided an opportunity for the natural body processes to remodel the entire musculoskeletal system and neural structures of the craniovertebral junction region and the rest of the spine and is Nature's attempt to sustain the instability and avoid or minimize the compromise of critical neural structures. The remodeling may even occur at the cost of self-destructive processes such as syringomyelia and morphological alterations in the form of basilar invagination and CM. Remarkable neurological recovery begins in the immediate postoperative phase following atlantoaxial stabilization without manipulating or handling the region of the tonsils. Observation suggests that CM may be a secondary phenomenon and a natural neural alteration in the face of atlantoaxial instability.

The experience with 65 surgically treated cases of CM associated with or without basilar invagination and with or without syringomyelia is described in this paper.

Methods

Cases of CM surgically treated using atlantoaxial fixation between January 2010 and November 2013 were reviewed and analyzed. The surgical strategy of atlantoaxial fixation was performed in all cases regardless of any demonstrable evidence of craniovertebral instability or physical bone or joint abnormality. Cases of CM and syringomyelia related to any gross structural brain or spinal malformation, hydrocephalus, tumor, infection, or connective tissue disorder were excluded.

All patients underwent atlantoaxial facet stabilization fortified by plate and screw fixation performed using the surgical steps described in previous studies.^{13,15,18–20} The aim of surgery in all cases was atlantoaxial stabilization, and in patients with Group A basilar invagination, an additional attempt was made to reduce basilar invagination and restore craniovertebral alignment. Cervical traction was used only intraoperatively and was applied after the induction of anesthesia. The weights were progressively increased to approximately 5–6 kg, depending on patient age and body weight. The aim of traction was to stabilize the head in the optimum surgical position. The direction of applied traction avoided or reduced direct pressure on the face and eyes during surgery. Distraction of the facets was not the primary aim in applying traction. In all cases with Group B basilar invagination, the C-2 ganglion was sectioned to expose the joint widely. In Group A basilar invagination cases and in cases without basilar invagination,

the ganglion was retracted superiorly to expose the facets and the joint, and sectioning of the ganglion was generally unnecessary. But even in these patients, whenever there was difficulty in exposing the facets widely, the C-2 ganglion was sectioned. The articular cartilage was widely removed using the sharp edge of an osteotome. Varying the sizes of the osteotomes, which were introduced into the joint with the flat edge and then turned 90°, and forcefully repeating these movements in a screwing fashion effect the distraction and denude the cartilage. Whenever necessary, a power-driven drill was used to widely denude the articular surface of cartilage. Corticocancellous bone graft material harvested from the iliac crest was packed into the joint in small pieces. Specially designed titanium spacers were used in 8 cases having basilar invagination and jammed into the joints to provide additional distraction and stability to the joint. Spacers were impacted into the joint whenever it was believed that spacers were mandatory to maintain the distraction and assist in providing stability. Plate and screw fixation of the region subsequently proceeded via the technique described by us.^{18,20} Screws were inserted directly into the substance of the facet of the atlas and via the pars/pedicle into the substance of the facet of the axis. Bone graft pieces were additionally placed in the midline over the lamina of C-2 and arch of the atlas, and in cases in which there was assimilation of the atlas in the suboccipital region. Stainless steel plates, monoaxial nonlocking-type screws, and custom-made titanium material spacers were used in the initial part of the series. After June 2013, the plates and screws used for fixation were made of MRI-compatible titanium material.

Results

Sixty-five patients with CM were treated with atlantoaxial fixation in the defined study period. In 55 cases, there was syringomyelia. According to the standard described craniometric criteria for the diagnosis of basilar invagination (the tip of the odontoid process at least 2.5 mm above Chamberlain's line),⁵ 46 patients had basilar invagination. As per a classification system proposed in 2004, the presence (Group A) or absence (Group B) of manifest instability of the craniovertebral junction, demonstrated by a distancing of the odontoid process from the posterior arch of the atlas, was used to divide all cases with basilar invagination into two groups.^{13,15,29} The clinical and radiological features of the cases are presented in Tables 1–4. Trauma of varying severity was the principle precipitating factor in 7 patients with Group A basilar invagination, 2 patients with Group B basilar invagination, and none of the patients without basilar invagination. Forty-six patients were male and 19 patients were female, with ages ranging from 12 to 50 years (mean 28 years) with Group A invagination (Figs. 1 and 2), 14–58 years (mean 29 years) with Group B invagination (Figs. 3 and 4), and 13–57 years (mean 35 years) in patients without basilar invagination (Fig. 5 and Table 1). All patients had varying degrees of neurological dysfunction at admission. The duration of symptoms ranged from 1 month to 4 years (mean 19 months) in patients with Group A basilar invagination, from 1 month to 15 years (mean 29 months) in those

TABLE 1. Summary of clinical characteristics in 65 patients with CM

Factor	No. of Patients		
	Basilar Invagination		No Basilar Invagination
	Group A	Group B	
Age in yrs			
11–20	2	8	2
21–30	10	9	8
31–40	4	6	3
41–50	2	3	4
51–60	0	2	2
Duration of symptoms in mos			
1–12	9	12	9
13–24	5	7	2
25–36	2	3	0
37–48	2	1	3
49–60	0	2	1
≥61	0	3	4
Presenting symptoms			
Neck pain	18	23	12
Arm pain	2	5	7
Paresthesias	10	16	13
Weakness/stiffness	18	26	15
Hoarseness/nasal regurgitation	5	5	2
Bowel/bladder weakness	0	3	0
Presenting sensory symptoms			
Only posterior columns	1	3	2
Only spinothalamic columns	7	5	2
Both posterior & spinothalamic columns	7	11	7
Normal sensations	7	10	3

with Group B basilar invagination, and from 1 month to 21 years (mean 45 months) in the group without basilar invagination. According to the extent of neurological disability, patients were divided into 5 grades as shown in Table 2: Grade 1, independent and normally functioning; Grade 2, walks on own but needs minimal support/help to perform routine household activities; Grade 3, walks with minimal support and requires help to perform household activities; Grade 4, walks with heavy support and unable to perform household activities; and Grade 5, unable to walk and dependent for all activities. Patients having lower cranial nerve (CN) weakness before surgery were

especially identified in the grading system. Our grading system and the Japanese Orthopaedic Association (JOA)¹⁰ and visual analog scale (VAS) scores²⁵ (Table 4) were used to monitor the clinical state before and after surgery and at the follow-up. The pyramidal symptoms formed a dominant component. Kinesthetic sensations were affected in 38% of cases. The incidence of spinothalamic dysfunction was less frequent (21%). Neck pain was a major presenting symptom observed in 82% of cases. Torticollis was present in 15 patients (23%). Neuropathic pain in the hand and shoulder was present in 14 patients (22%; Tables 1 and 3).

All patients underwent dynamic plain radiography and

TABLE 2. Distribution per clinical grading system

Grade	Description	No. of Patients		No. of Patients w/ Lower CN Deficits
		Preop	Postop*	
1	Independent & normally functioning	11	30	2
2	Walks on own but needs support/help to perform routine household activities	19	25	
3	Walks w/ minimal support & requires help to perform household activities	10	5	2
4	Walks w/ heavy support & unable to perform household activities	13	3	3
5	Unable to walk & dependent for all activities	12	0	5

* One patient died and another continued to have neuropathic pain.

TABLE 3. Type of atlantoaxial dislocation

Type of Dislocation	No. of Patients
I	17
II	31
III	17

CT scanning with the head in flexion and extension, CT angiography, and MRI before surgery.

Depending on the relationship of the facets of the atlas with the facets of the axis in the sagittal plane, 3 types of dislocation were identified (Table 3). Apart from the dynamic CT study, the dislocation was visualized more vividly on MRI and 3D CT in some cases. Type I dislocation was when the facet of the atlas was dislocated anterior to the facet of the axis (Fig. 2D and E). Type II dislocation was when the facet of the atlas was dislocated posterior to the facet of the axis in any head position (Figs. 1D and 3D). Type III dislocation was when no evidence of facet malalignment was demonstrated on plain or dynamic imaging (Fig. 4C). Instability in such cases was termed “central” instability. There was evidence of vertical, mobile, and partially or completely reducible atlantoaxial dislocation (as described by us earlier in 2009) in 18 cases.²³ In patients having basilar invagination, there was occipitalization of the atlas in 33 cases and C2–3 fusion in 4 cases. Radiological images and operative observations revealed that the atlantoaxial joint was “open,” functional, and unstable in all patients. The articular cartilage was intact in all cases. Observation of the structure of the cerebellar hemispheres, vermis, and tonsils revealed in 39 cases that the brain matter was more densely packed in the inferior vermis and in the tonsils when compared with the superior vermis and the superior aspect of the cerebellum.¹⁷ There was atrophy of at least part of the cerebellum in these cases (Fig. 4A).

Because of surgical technical difficulties related to complex anatomy, in 4 patients (3 with Group B basilar invagination and 1 with Group A), the atlantoaxial fixation could not be performed on both sides with our technique, which entails the individual insertion of screws in C-1 and C-2 pars, pedicles, and facets.^{20,22} In 2 patients, transarticular fixation was performed on one side, and our technique was used on the contralateral side. The point of entry and the direction of the transarticular screw were altered to suit the complex local anatomy in these cases. In

1 patient, a spacer was inserted into the facet joint for distraction and fixation on the contralateral side. In 1 patient, fixation was done using our technique on only one side; no instrumented fixation was performed on the contralateral side. Vertebral artery injury during the dissection in the region occurred in 3 patients. Injury occurred during the dissection in the region of the C-1 facet because of an anomalous vertebral artery course in 1 patient and during the insertion of C-2 screws in 2 patients. The artery was sacrificed in all cases. In 1 patient there was a postoperative infarct in the territory of the posterior circulation, and the patient ultimately succumbed. Postoperatively, traction was discontinued in all patients, and each one was placed in a four-post hard cervical collar for 3 months, during which all activities related to neck movements were restricted to allow for bone fusion.

All postoperative assessments were performed using clinical and radiological information obtained at least 3 months after surgical treatment. Dynamic plain radiography and CT scanning were used to confirm postoperative arthrodesis. Fusion of the facets and the posterior elements of atlantoaxial bone were especially observed. Despite the presence of artifacts related to the use of stainless steel implants, the CT image quality was satisfactory to demonstrate alignment and bone fusion. Given that stainless steel metal implants were used for fixation in the majority of cases, postoperative MRI was impossible. As titanium implants were used only after June 2013, it was impossible to evaluate via MRI the long-term status of the syrinx and CM. However, definite reductions in the dimensions of the syrinx and reductions in tonsillar herniation were observed within a period ranging from 2 to 6 months after surgery in at least 6 of 11 cases in which postoperative MRI was available for evaluation (Fig. 6).

Sixty-three patients improved following surgery. Improvement was observed in the immediate postoperative phase, and the progress of improvement was sustained at follow-up. Clinical improvement per our grading system, JOA score, and VAS score is shown in Tables 2 and 4. The follow-up period ranged from 3 to 48 months (mean 18 months). In 17 patients, the follow-up was less than 1 year. One patient continued to have significant neuropathic pain in the limbs; however, the duration of follow-up in this patient was only 3 months. Symptoms improved to varying degrees in 63 patients following surgery. Seven patients who had had preoperative symptoms related to the lower CNs improved following surgery. Two patients who had needed Ryle's tube feeding prior to surgery recovered swallowing function, and the Ryle's tube could be removed in these patients within 3 months of surgery. Hoarseness resolved in 7 patients; the recovery was observed in the immediate postoperative period and was progressive. There was one case in which there was after surgery clear evidence of a posterior circulation infarct related to an intraoperative vertebral artery injury. Apart from this case, there were no postoperative vascular, neurological, or infective complications. No patient suffered a delayed neurological worsening sufficient to warrant a transoral or posterior decompressive surgery or any other type of operative procedure. No patient required reexploration for the failure of implant fixation. Immediate

TABLE 4. Preoperative and postoperative clinical assessments

Factor	No. of Patients	
	Preop	Postop
JOA score		
<7	12	0
8–12	29	7
13–15	20	29
16–17	4	28
Range of VAS neck pain scores in 64 patients	5–8	0–2

FIG. 1. Images obtained in a 21-year-old woman. **A:** A T2-weighted MR image shows Group A basilar invagination, syringomyelia, and CM. **B:** CT scan with the head in flexion shows basilar invagination, assimilation of the atlas, and C2–3 fusion. **C:** CT scan with the head in extension shows mild reduction in the basilar invagination. **D:** CT slice through the facet, showing Type II atlantoaxial dislocation. **E:** Postoperative CT scan showing a reduction in basilar invagination and fixation. **F:** Postoperative CT slice through the facet joint, showing alignment of the facets and fixation. **G:** Postoperative radiograph showing the implant.

postoperative and follow-up radiography at least 3 months after surgery confirmed fixation and fusion. Fusion was considered to be successful when the implant demonstrated the maintenance of fixation on dynamic radiography and bone fusion was observed in the facets and suboccipital region. Successful and sustained distraction and partial or complete reduction of basilar invagination were observed in all patients. Torticollis improved to a varying extent in all cases following surgery. There was at least some degree of C-2 sensory loss in all cases in which the ganglion was sectioned.

Discussion

Chiari malformation and syringomyelia are commonly associated. The twin issues have been addressed jointly in a large number of articles in the literature for over a century. A number of possible pathogenic factors have been speculated. A congenital origin is the most accepted theory. Both CM and syringomyelia are considered to be pathological entities, and accordingly a number of treatment methodologies have been discussed. Further, CM and syringomyelia are frequently associated with basilar invagination. In our series 46 patients (71%) had basilar invagination. It is unclear in the literature whether CM and

syringomyelia in presence of basilar invagination must be treated differently. There is near universal agreement that foramen magnum decompression forms the cornerstone of the treatment strategy for CM. A literature survey of CM treatment via foramen magnum decompression revealed surgical failure rates ranging from 20% to 50%.^{27,34} Although the issue of instability has been occasionally associated with CM, its prime role in the pathogenesis of the disease has not been discussed or evaluated.

In 1998, we divided basilar invagination into two subgroups on the basis of the absence (Group I) or presence (Group II) of CM.¹⁵ According to our observations as well as reports in the literature,³⁰ CM in the presence of basilar invagination was caused by a reduction in the volume of the posterior cranial fossa. On this premise, we discussed the indications for transoral surgery in Group I cases and for posterior fossa or foramen magnum decompression surgery in Group II cases. It was suggested that enlargement of the posterior fossa volume was necessary in these cases and that there was no need to open the dura mater or to perform any kind of duraplasty. In 2004, a new classification of basilar invagination divided cases into Groups A and B on the basis of a radiological indication of the presence or absence, respectively, of instability of the region that was manifested by distancing of the odontoid process

FIG. 2. Images obtained in a 45-year-old woman. **A:** A T2-weighted MR image showing Group A basilar invagination, CM, and syringomyelia. **B:** CT scan with the head in flexion, showing Group A basilar invagination, assimilation of the atlas, and C2–3 fusion. **C:** CT scan with the head in extension, showing a mild reduction in basilar invagination. **D:** CT slice obtained through the facet while the head was in flexion, showing Type I atlantoaxial dislocation. **E:** CT scan with the head in extension, showing a mild reduction in facet dislocation. **F:** Postoperative CT scan showing a reduction of basilar invagination and fixation. **G:** Postoperative CT slice through the facet joint, showing the implant and realignment of the facets.

from the anterior arch of the atlas.¹³ For Group A basilar invagination, in which there was radiological evidence of instability, facet distraction, craniovertebral realignment, and atlantoaxial fixation as a rational form of treatment were recommended.¹³ We proposed the use of a bone graft within the joint with or without the deployment of titanium spacers to retain distraction and reduction of basilar invagination. Bone fusion ultimately assists in arthrodesis and retains the reduction of basilar invagination. Such a surgical strategy avoided the need for transoral surgical decompression, foramen magnum decompression, and inclusion of the occipital bone in the fixation construct. The same surgical strategy was adopted even in cases of Group A basilar invagination associated with CM and with syringomyelia.¹³ Subsequently, some authors have attempted to reduce basilar invagination by a variety of means.^{6,26}

In 2009, we reported our experience treating 170 cases of Group A basilar invagination and identified the reversal of musculoskeletal changes frequently encountered in cases having basilar invagination.²¹ We documented an increase in the length of the neck and the reversal of torticollis in the immediate postoperative phase. There was craniover-

tebral and spinal realignment following facet distraction surgery. On the basis of the analysis, it was speculated that bone fusions like assimilation of the atlas, C2–3 fusions, and other spinal fusions are potentially reversible following atlantoaxial fixation. It was observed that atlantoaxial instability related to basilar invagination was the primary event and that the musculoskeletal changes were secondary natural phenomena designed to limit the extent of cervicomedullary neural compression to the minimum.

The atlantoaxial dislocation in cases with basilar invagination was earlier referred to as “fixed” or “irreducible” dislocation. However, we showed that dislocation in the basilar invagination Group A was not fixed, that the joint in these cases was mobile and in several cases hypermobile, and, more importantly, that the dislocation was reducible by manual distraction and realignment.^{13,21} We introduced the term “vertical” mobile and reducible atlantoaxial dislocation for basilar invagination.²³

Before the present study, until the year 2009, it appeared to us that in Group B basilar invagination, the atlantoaxial dislocation was fixed. Accordingly, posterior foramen magnum decompression was performed in these cases.^{13,15}

FIG. 3. Images obtained in a 19-year-old man. **A:** Sagittal T1-weighted MR image showing Group B basilar invagination, CM, and syringomyelia. **B:** A T2-weighted MR image. **C:** CT scan showing Group B basilar invagination. **D:** CT slice through the facets, showing Type II atlantoaxial dislocation. **E:** Postoperative CT scan showing fixation. **F:** Postoperative CT slice through the facets, partially showing the implant and facet alignment.

As our experience in the matter increased over the years, we observed that basilar invagination is uniformly associated with atlantoaxial dislocation or instability. In cases with Group B basilar invagination, the presence of atlantoaxial instability as the sole and primary pathogenic event was identified even when the instability was not demonstrated by dynamic imaging. Group B basilar invagination cases are more frequently associated with CM and syringomyelia. In the present analysis, 28 cases (43%) with CM had Group B basilar invagination.

More recently, we realized that CM with or without syringomyelia was caused by atlantoaxial dislocation, regardless of the presence or absence of basilar invagination. It appeared that basilar invagination, CM, and syringomyelia were a continuum of the same pathological phenomenon that originates from atlantoaxial instability. The time

of pathogenesis and the nature and intensity of atlantoaxial dislocation seem to define the subsequent processes that partake in natural adjustments. Accordingly, we resorted to atlantoaxial fixation in cases in which there was CM with or without syringomyelia and with or without basilar invagination.

Depending on the relationship of the facets of the atlas with the facets of the axis, 3 types of dislocation were identified in cases of CM with or without basilar invagination. Although some millimeters of facet alignment can be considered within the spectrum of normal physiological limits, the association of this misalignment with other abnormalities in the region indicated the presence of instability. Type I dislocation was when the facet of the atlas was dislocated anterior to the facet of the axis. We earlier identified this type of dislocation of the atlas

FIG. 4. Images obtained in an 18-year-old male. **A:** T2-weighted MR image showing Group B basilar invagination and CM. Mild atrophy of the superior aspect of the vermis is visible. **B:** CT scan showing basilar invagination. **C:** CT slice through the facets, showing central dislocation. **D:** Postoperative CT scan showing fixation. **E:** Postoperative image through the facet, showing the implant.

toaxial articulation with Group A basilar invagination. Anterior atlantoaxial facet dislocation, a phenomenon that we related to as atlantoaxial listhesis, is a more severe form of instability and results in a rather acute form of basilar invagination. This form of instability is less frequently associated with CM or syringomyelia. The average age of clinical presentation in these Group A patients was 28 years. We identified Type II facet dislocation in 31 cases, wherein the facet of the atlas was dislocated posterior to the facet of the axis. Such a form of dislocation has not been reported in the literature. The relationship of the facets on dynamic images was inconsistent. Alignment of the odontoid process and the anterior arch of the atlas was not remarkably abnormal in these cases, and the presence of instability was difficult to detect on plain radiographs and images that focused on the location of the odontoid process. In 17 cases, there was no evidence of facet malalignment on plain or dynamic imaging (Type

III dislocation). In these cases, there was no gross physical abnormality of the facets of the atlas or axis. The operation in such cases was performed on the assumed presence of atlantoaxial instability. Although dynamic imaging did not clearly demonstrate instability in these cases, direct observation of the joint status during surgery clearly revealed its unstable character. We labeled such dislocation as Type III or “central” dislocation.

Types II and III dislocations were frequently associated with the more severe form of basilar invagination (Group B basilar invagination) that resulted in marked bone structural abnormality and a rostral location of the atlantoaxial joint. Such a dislocation was also frequently associated with cases in which there was gross neural structural deformity in the form of CM and syringomyelia with or without any basilar invagination. Analysis of the clinical features indicated that the symptoms and signs were the result of odontoid process–induced brainstem compression in patients

FIG. 5. Images obtained in a 30-year-old man. **A:** T2-weighted MR image shows CM and syringomyelia. There is no basilar invagination. **B:** CT scan showing the craniovertebral junction. **C:** CT slice through the facets, showing Type II dislocation. **D:** An MR image showing the facet joint. **E:** Postoperative scan showing fixation. **F:** Postoperative slice through the facet, showing the implant.

with Type I atlantoaxial instability. Patients with Types II and III instability in general presented with a symptom complex primarily related to CM and syringomyelia and less prominently attributable to brainstem compression.

On evaluating the dynamic images, we identified 18 cases having vertical mobile and reducible atlantoaxial dislocation.²³ In several of these cases, the vertical dislocation was subtle but definite. Identification of such vertical dislocation seems to be an important additional parameter suggesting instability of the region and incompetence of the joints. The presence of facet malalignment and evidence of vertical mobility of the odontoid process were indicators of instability of the atlantoaxial joint.

The symptoms, radiological and physical alterations, and presence of CM with or without syringomyelia seem to depend on the time of pathogenesis and the degree and nature of atlantoaxial instability. In general, the greater the instability, the younger the age at presentation, the more marked the clinical motor and sensory symptoms, and the less prominent the radiological and physical musculoskeletal malformations. On the other hand, when atlantoaxial dislocation is subtle or less marked, the clinical features and neurological deficits are more long-standing and the physical and radiological manifestations are more marked. The presence of CM, syringomyelia, and Group B basilar

invagination was suggestive of the long-standing nature of neural and skeletal alterations and was more often associated with subtle atlantoaxial dislocation. Although our proposed clinical grading system must be validated with additional studies, we found that the system was simple and reproducible and provided a clear impression of the patient's clinical status. Another issue about our proposed grading system is that although the parameter of lower CN weakness is included in the definition, no objective data document the severity of the weakness or the degree of its improvement, and thus the parameter is basically based on subjective information. Considering the potential flaws in our grading scheme, we also used the standard and universally accepted JOA and VAS scoring systems for clinical assessments.

The difficulty in exposing the atlantoaxial joint was related to its location. The more rostral it was, the more difficult the exposure. The surgical procedure was relatively easier in cases in which there was no basilar invagination. In cases with basilar invagination, exposure of the joints was relatively easier when there was no assimilation of the facet of the atlas. Severe basilar invagination and rostral positioning of the joint as identified in Group B basilar invagination cases resulted in a marked difficulty in surgically exposing the joint and manipulating the fac-

FIG. 6. Images obtained in a 22-year-old woman. **A:** Preoperative CT scan showing the craniocervical junction and otherwise normal alignments. **B:** Sagittal CT scan with the cut passing through the facets, showing Type II facet dislocation. **C:** Preoperative T1-weighted MR image with and without contrast showing CM and syringomyelia. **D:** Preoperative T2-weighted MR image showing CM and syringomyelia. **E:** A T1-weighted MR image obtained within 48 hours of surgery, showing persistent syringomyelia and CM. **F:** Immediate postoperative T2-weighted MR image shows persistent syringomyelia and CM. **G:** Postoperative CT scan showing fixation. **H:** CT scan cut through the facets, showing fixation with an implant. **I:** T1-weighted MR image obtained 3 months after surgery, showing a significant reduction in syringomyelia and regression of CM. **J:** Delayed postoperative (after 3 months of surgery) T2-weighted MR image showing reduction of the syrinx and regression of the CM.

ets. Assimilation of the facet of the atlas into the occipital condyle resulted in a thick and strong facet. The course of the vertebral artery posterior to the facet of an assimilated atlas in at least 4 cases posed an additional and sometimes severe difficulty in exposing the joint. However, because of our familiarity with the technique of exposing the atlantoaxial joint for over 25 years, we did not, in any case, resort to inclusion of the occipital bone in the fixation construct. The short length of the plate/rod that was used when direct facet fixation was done provided a mechanical advantage over cases in which long plates or rods were used. Thin occipital bone, the possibility of using short screws, and the need to use long plates/rods result in the inferior biomechanical potential of such a construct as compared with the potential of a short and strong construct, as described and used by us.^{13,20,24} Direct handling of the facets provided an opportunity to assess the status of joint stability. Moreover, the possibility of opening the articular cavity, denuding the articular cartilage, placing bone graft with or without an additional spacer within the articular cavity provided space for bone fusion and additional stability. The primary aim of surgery in all cases was to achieve firm stabilization of the atlantoaxial joint and to obtain segmental arthrodesis. During the follow-up period, all patients had firm arthrodesis of the region, and there was no instance of implant failure or infection. Considering the surgical difficulties associated with direct atlantoaxial fixation and the potential dangers to the vertebral artery, particularly in cases with an assimilated atlas,

including the occipital bone can form a reasonably strong construct and can stabilize the craniocervical junction, although we did not perform such a procedure in the present series. The term “craniocervical stabilization,” rather than “atlantoaxial stabilization,” may be more suitable in such cases.

Direct observation of the joint status was possible during surgery when applying the lateral mass plate and screw fixation technique described by us.^{13,18,20} Instability of the atlantoaxial joint was invariably identified while performing surgery. Although foramen magnum decompression with or without duraplasty has produced a satisfactory clinical outcome, our experience in such cases suggests that atlantoaxial dislocation forms the primary basis of etiopathogenesis. Our successful outcomes, both in general and in particular in cases in which earlier foramen magnum decompression had failed, has convinced us of the paramount significance of fixing the atlantoaxial joint and the futility of resorting to foramen magnum decompression. Prior to 2010, we had resorted to foramen magnum decompression surgery in all cases having Group B basilar invagination.^{13,15} We were satisfied with the clinical outcome in these cases. Although not appropriately quantified and further clinical evaluation will be mandatory to assess this assertion, it appears that the results of treatment with atlantoaxial fixation are significantly superior and long lasting as compared with the clinical outcome observed after foramen magnum decompression surgery. The very fact that a significant majority of patients im-

proved in clinical outcome after atlantoaxial fixation without any foramen magnum decompression suggests that instability is a major factor in the pathogenesis of CM and syringomyelia. Apart from the improvement in motor and sensory symptoms, atlantoaxial fixation led to recovery in lower CN weakness. Such recovery of lower CN function is relatively infrequently observed with conventional forms of treatment.

The degree of heterogeneity in the morphology of cases assessed in our study must be evaluated and compared with that in other similar series available in the literature. Despite the heterogeneous morphology, it appears that the basic pathogenic premise is instability at the atlantoaxial joint.¹¹ The nature and degree of instability, its laterality, the timing of its initiation, and several other known and unknown factors may determine the presence or absence of basilar invagination, syringomyelia, short neck, and other musculoskeletal and neural abnormalities. It has been generally observed that basilar invagination is more frequently encountered in the Indian subcontinent. The exact reason for this epidemiological discrepancy is unclear. In an earlier study, we incriminated nutritional factors and child birth-related trauma due to poor delivery practices related injury to the muscles of the nape of the neck and the consequent instability as a possible cause.²¹ It is difficult to determine if the subgroup of patients with CM treated by us in a relatively poor population of India is different from groups discussed by other authors, the majority of whom are from more affluent countries.^{28,31–33,36} The incidence of basilar invagination (84%) was significantly higher in our series than in the series of Milhorat et al. (12%) and Kleklamp (9.2%).^{16,28,32} There is no or only a passing reference to the presence of basilar invagination or craniovertebral instability in other major series on the subject.^{1,4,9,28,31–33} However, the patients in the above-mentioned were not treated differently from those who did not have basilar invagination.

Our surgical results as described lead us to speculate that CM may be Nature's protective mechanism that assisted in reducing the effect of instability and cord compression by the odontoid process. We speculate that CM may have an "air bag" type of influence that prevented pinching of vital neural structures between the bones. Essentially, we observed that CM is a manifestation of atlantoaxial instability. The prominence and easy identification of CM on MRI could divert the clinician's attention from the relatively poorly delineated atlantoaxial joint and instability. It appears that CM is unrelated to the reduction in posterior cranial fossa volume, and nor is it attributable to the primary or relative increase in the cerebellar mass. In 39 cases in the present study, there was "atrophy" of the superior vermis. Analysis of our cases revealed that in none of the cases without basilar invagination was the posterior cranial fossa volume or foramen magnum volume reduced or compromised. These features suggested that the hypothesis of intracranial or posterior cranial fossa hypertension (as proposed by us and others), cranial constriction, or an increase in cerebellar mass in proportion to the volume of the posterior cranial fossa as a cause of cerebellar herniation may not be correct.³³ It appears that the temporary improvement after foramen magnum decom-

pression is akin to deflating a full air bag, and in the long-term such a form of surgery can be counter-productive. As we speculated earlier, syringomyelia may be a response of the body that balances the pressures within the neural structures in the interest of the patient.¹² The presence of CM and syringomyelia suggests the chronicity of the process and the subtlety of atlantoaxial dislocation. The very fact that there was recovery in the presenting symptoms in all of our surviving patients suggests the effectiveness of stabilization and points to the pathogenesis. Although we could not assess the status of CM and syringomyelia in the majority of our cases given the use of stainless steel material for the fixation, we did observe, in a period ranging from 2 to 6 months after surgery, a definite reduction in the size of the syrinx and reverse superior migration of CM in 6 of 11 cases in which titanium and MRI-compatible implants were used (Fig. 6). Such a reduction in syrinx size and regression of tonsillar herniation without their direct handling does suggest the validity of the proposed hypothesis. Further prospective analysis of cases treated by multiple surgeons and at multiple institutions is mandatory to confirm our observations.

Conclusions

Given our experience, we conclude that CM, with or without basilar invagination, is always associated with instability at the atlantoaxial joint, even if such instability is not clinically manifest or is not demonstrated on radiological imaging. Stabilization of the atlantoaxial joint is the treatment. Foramen magnum bone or dural decompression is unnecessary. We speculate that both syringomyelia and CM are secondary natural events related to long-standing atlantoaxial dislocation and do not need direct surgical manipulation.

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