

Atlantoaxial fixation for failed foramen magnum decompression in patients with Chiari formation

ABSTRACT

Background: Relationship of atlantoaxial instability with Chiari formation is further analyzed in the report.

Objective: The outcome of 25 patients who had failed conventional treatment for Chiari formation that included foramen magnum decompression surgery and were treated by atlantoaxial fixation is analyzed.

Materials and Methods: During the period January 2010 to November 2019, we treated 25 patients who had undergone conventionally described surgical procedures; all included foramen magnum decompression for Chiari formation. None of the patients had any craniocervical junction anomaly. All patients had syringomyelia. All patients had worsened in their neurological condition following surgery either in the immediate or in the delayed postoperative phase. Atlantoaxial instability was diagnosed on the basis of facet alignment and on the basis of direct observation of joint status by bone manipulation during surgery. The patients were treated by atlantoaxial fixation. Goel clinical grading scale and Japanese Orthopedic Association Score assessed the clinical status both before and after surgery.

Results: Following surgery, all patients improved in the clinical condition. The improvement began in the immediate postoperative period and progressed. During the follow-up period that ranged from 4 to 123 months, "significant" neurological recovery and amelioration of presenting symptoms were observed. During the period of follow-up, reduction in the size of syrinx was observed in 14 out of 18 cases where postoperative magnetic resonance imaging was possible.

Conclusions: Clinical results reinforce the belief that atlantoaxial instability is the nodal point of pathogenesis of Chiari formation. Atlantoaxial fixation is the treatment.

Keywords: Atlantoaxial fixation, atlantoaxial instability, basilar invagination, Chiari formation, syringomyelia

INTRODUCTION

In 2015, we presented our experience with 65 cases with Chiari formation.^[1] Recently, we evaluated the results of surgical treatment of 33 patients in the pediatric age group.^[2] In both of these articles, it was speculated that potential or manifest atlantoaxial instability was the nodal initiating point of pathogenesis of Chiari formation and atlantoaxial fixation was the mode of surgical treatment. We also recommended that foramen magnum decompression of any kind was counter-productive in the long run.^[3,4] Our conclusions had initiated an international debate on the subject.^[5-13] Our increasing clinical experience that we have published recently in the subject reinforces our earlier observation.^[14]

Failure rate of foramen magnum decompression surgery in major surgical series ranges from 15% to 50%.^[1,2,14-16] No pointed treatment has been recommended in the literature for patients with Chiari formation who failed foramen magnum decompression surgery. Despite the "relatively"

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short follow-up periods, considering the possible clinical implications of our observations, we present results of treatment of 25 cases having Chiari formation with atlantoaxial fixation where the conventional and generally accepted form of treatment had failed. Our literature search failed to isolate any clinical study that focused on the treatment of failed foramen magnum decompression surgery.

MATERIALS AND METHODS

During the period January 2010 to March 2019, 25 consecutive cases having Chiari formation were re-operated in our department of neurosurgery (two centers) wherein the previously conducted surgery by conventional forms of treatment had failed and the patients continued to worsen in their symptoms. All patients provided written informed consent, and all clinical tests and surgical procedures were conducted according to the principles of the Declaration of Helsinki. As the study is a retrospective analysis of data, ethics committee permission was not deemed necessary. The patients have been included in our previous study group.^[1,14] In this paper, we present detailed analysis of 25 patients with no craniovertebral junction bone anomaly who had been previously treated with foramen magnum decompression. During the period of study, in one patient who was re-operated for failed foramen magnum decompression, operation could not be completed due to abnormal hemodynamics during surgery. This patient has not been included in the study. There were 18 males and 7 females and their ages ranged from 17 to 48 years (average 29 years). The clinical status was assessed on the basis of Goel clinical grading scale and on the basis of JOA score assessments.^[1] Fifteen patients were unable to walk unaided at the time of surgery, five patients had Ryle's tube feeding tube, and seven patients had difficulty in speaking that was primarily related to breathlessness. Table 1 summarizes the previously done surgical procedures. All patients underwent dynamic (flexion–extension) computed tomography (CT) scan and magnetic resonance imaging (MRI). Tables 2 and 3 summarize the clinical condition, and Table 4 summarizes the radiological findings at the time of first surgery. As discussed previously, atlantoaxial instability was identified on the basis of abnormal alterations in the atlantodental interval (anteroposterior atlantoaxial dislocation) and abnormal vertical movements (vertical atlantoaxial dislocation) of odontoid process on dynamic imaging.^[17] In addition to these conventionally described radiological parameters, we used our recently described parameter to diagnose atlantoaxial instability.^[18] On the basis of lateral profile imaging in the neutral head position and assessment of facet alignments and on the basis of observations on manual manipulations of bones during surgery, atlantoaxial instability was classified into three types. Type 1 atlantoaxial facet

Table 1: The various surgical procedures performed on the patients

Type of initial surgery	Number of patients
Foramen magnum decompression (only bone)	3
Foramen magnum decompression and duroplasty with or without C1 and C2 laminectomy	22
Syringo-subarachnoid shunt	6
Ventriculoperitoneal shunt	3

instability was when the facet of atlas was dislocated anterior to facet of axis. Type 2 atlantoaxial facet instability was when the facet of atlas was positioned posterior to the facet of axis. Type 3 atlantoaxial facet instability was when the facets of atlas and axis were in alignment and instability was diagnosed only on direct manipulation of bone during surgery. In Types 2 and 3, atlantodental interval was not necessarily abnormally altered and such forms of dislocation were labeled as central or axial atlantoaxial instability. Essentially, the presence of Chiari formation by itself was an indicator of atlantoaxial instability and atlantoaxial stabilization was done, irrespective of the observation of instability on radiological imaging and even when there was no evidence of any dural or neural compression by the odontoid process.

Surgical procedure

All patients underwent surgical procedures discussed by us earlier in 1994 and 2004.^[19,20] The patients were placed in prone surgical position. The head was placed in Gardner Wells cervical traction. The head end of the table was elevated by about 30°, to provide counter-traction to the weights. The traction and the table position were aimed to provide a stable head during the surgery. The traction was not primarily aimed at providing distraction of the facets. Bone graft was harvested from the iliac crest. The foramen magnum bone removal during the previous surgery posed difficulties during the joint exposure. Articular cartilage was widely denuded, bone graft was packed into the joint space, and plate–screw fixation was done. The venous bleeding in the lateral gutter and in the extradural space in cases with associated syringomyelia or “external syringomyelia” was significantly higher when compared to patients having no abnormalities of CSF loculation.^[21-23] Packing of the extradural space with gel-foam and/or Surgicel controlled the bleeding. The muscles attached to the spinous process of the axis were widely sectioned. The host bone area of posterior arch of atlas, lamina, and spinous process of axis and the exposed lateral mass bones of both atlas and axis were drilled to make them suitable for accepting bone graft. Traction was removed after the patient was made supine. The patients wore firm cervical collar for 3 months, and all major neck movements were restricted during the time. Postoperative MRI and CT scan were done within 24 h of surgery, at 3 months, and then subsequently at follow-up.

Table 2: The pre- and post-operative neurological status according to Goel's clinical grading scale

Goel's Clinical Grade	Number of patients (preoperative)	Number of patients (postoperative)	L after the number signifies lower cranial nerve affection	V after the number signifies voice abnormalities
Grade 1, independent and normally functioning	-	11	-	-
Grade 2, walks on own but needs minimal support/help to perform routine household activities	1	11	-	1
Grade 3, walks with minimal support and requires and helps to perform house-hold activities	9	3	-	-
Grade 4, walks with heavy support and unable to perform household activities	8	-	2	1
Grade 5, unable to walk and dependent for all activities	7	-	3	5

Table 3: The pre- and post-operative neurological status according to JOA grading scale

JOA score	Number of patients (preoperative)	Number of patients (postoperative)
<7	8	-
8-12	16	3
13-15	1	11
16-17	-	11

JOA - Japanese Orthopedic Association Score

Table 4: The radiological abnormalities

Radiological feature	Number of patients
No craniovertebral junction anomaly	25
Syringomyelia	25
Type of facetal instability	
Type 1	-
Type 2	5
Type 3	20

RESULTS

All patients improved in their symptoms on questioning "immediately" after their recovery from anesthesia. The major symptoms that improved or recovered in the postoperative phase were in voice quality, breathing difficulty, pain in the nape of neck and shoulders, tingling paresthesia, and stiffness of the limbs. The neurological status continued to improve during the follow-up period. The clinical status assessed at the period of follow-up of 3 months is detailed in Tables 2 and 3. Follow-up >1 year was available in 19 patients. None of the patients worsened after an initial improvement. Postoperative MRI examination after 6 months of surgery was possible in 18 cases. In 14 patients, there was clear evidence of reduction in the size of syrinx. There were no postsurgical complications.

Illustrative cases

Case 1

A 26-year-old male patient, a graduate college science student, had undergone a multitude of different surgeries

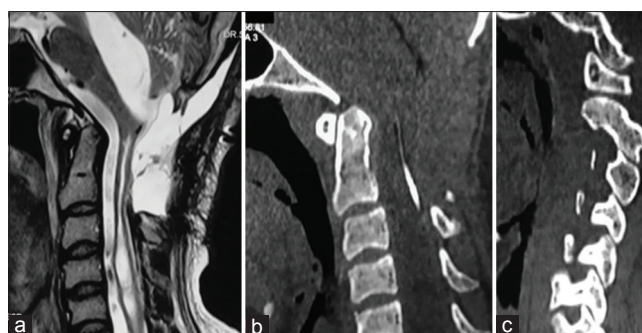


Figure 1: Preoperative images (Case 1). (a) T2-weighted magnetic resonance imaging after the initial surgeries. Syringomyelia can be seen. The fourth ventricle is widely open. Foramen magnum decompression can be seen. (b) Computed tomography scan showing evidence of foramen magnum decompression. No atlantodental interval disturbance is seen. (c) Computed tomography scan with sagittal cut passing through the facets. The facets are in alignment

for Chiari formation, 2 years before presentation to us. At the time of initial presentation, he had complaints of pain in the nape of the neck that radiated to both shoulders and paresthesia of tingling and numbness in both hands. Investigations had shown Chiari formation and syringomyelia. The first operation involved foramen magnum bone and dural decompression and C1 and C2 laminectomy. The patient did not improve following the surgery and had persistent disabling complaints. For these complaints, a right-sided ventriculoperitoneal shunt was performed 6 months following the initial surgery. However, the symptoms of the patients showed no respite and he started worsening. He was then subjected to repeat foramen magnum decompression and widening of the bone decompression and a syngo-subarachnoid shunt placement. Symptoms continued to progress. A syngo-subarachnoid shunt block was suspected, and a left-sided ventriculoperitoneal shunt was performed. However, the patient did not improve after the surgery and his clinical condition continued to deteriorate. He developed weakness and stiffness of all four limbs and was unable to stand or sit even with support. In addition, he developed change in his quality and tone of his

voice. He developed symptoms of difficulty in swallowing with nasal regurgitation of food, more for liquids than for solids. He started experiencing apneic spells, which made him get up at night, and after some deep breaths, he was able to sleep again. When brought to us, the patient had marked spastic quadriparesis. There was wasting of hand muscles with flexor spasms in both lower limbs. He had sensory loss over the chest wall and trunk and both lower limbs. He was unable to turn in bed independently and had poor control of his bowel and bladder. He also had difficulty in breathing and was using his accessory muscles of respiration. He was on Ryle's tube feeds and had a Foley's catheter *in situ*. He was completely dependent on his family members for all his personal activities (Goel Grade 5, JOA 5). Investigations revealed persistent syringomyelia [Figure 1]. The patient was now operated in our department, and atlantoaxial fixation surgery was done. Following surgery, the patient started to improve in all his symptoms in the immediate postoperative phase. At a follow-up of 6 months, he could stand and walk with minimal support and his voice was clearer and he could easily converse. He could eat and drink without Ryle's tube. His urinary catheter was removed. However, he continued to

have urinary incontinence. His breathing was now normal, and he could sleep comfortably. Investigations at this time showed a reduction in the syringomyelia [Figure 2].

Case 2

A 25-year-old female initially presented to another institute with the complaints of neck pain and paresthesia in all four limbs. Investigations revealed Chiari malformation with syringomyelia [Figure 3]. The patient was operated, and a foramen magnum decompression with a C1 and C2 laminectomy was performed. In addition, a syringo-subarachnoid shunt was performed at D3 level. The patient did not improve in her symptoms following surgery and after a period of 1 month started deteriorating neurologically. She started having increased paresthesia and weakness in all her limbs. When she presented to us 1 year following the initial surgery, she had spastic quadriparesis with a power of grade 3–4/5 (Goel Clinical Grade 4, JOA 8). Sensations were decreased in both her lower limbs. She was unable to walk unaided, and she could not perform her routine activities independently. Investigations performed at this time showed persistent Chiari formation and syringomyelia. The patient was operated and atlantoaxial fixation was done [Figure 4]. The patient improved in her symptoms following surgery. At a follow-up of 3 years, the patient is relieved of all her complaints and is back to her routine life. Postoperative imaging showed atlantoaxial fixation and reduction in the size of the syrinx [Figure 4].

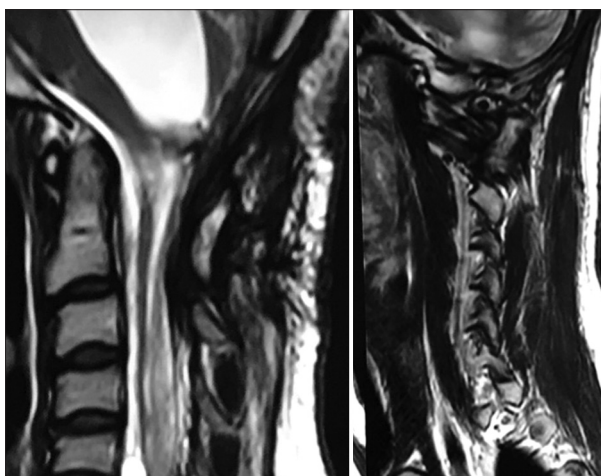


Figure 2: Postoperative images (Case 1). Postoperative T2-weighted magnetic resonance imaging showing reduction in the syringomyelia and the C1-C2 fixation

DISCUSSION

The treatment of Chiari formation has evolved over a century. The very fact that a number of forms of treatment have been recommended as a mode of treatment suggests that the pathogenesis has not been pointed, is unclear, and to say, the least is riddled with controversies and debate. In our earlier article on the subject, we mentioned that the number of forms of treatment of Chiari formation equals the number of surgeons treating the problem.^[1,2,14] Foramen magnum

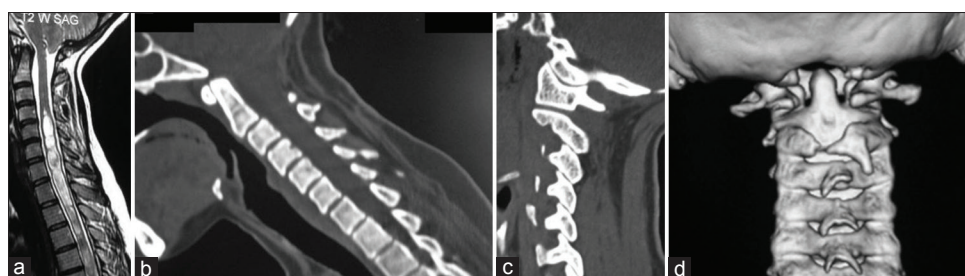


Figure 3: Preoperative images (Case 2). (a) T2-weighted magnetic resonance imaging showing Chiari formation and syringomyelia. (b) Computed tomography scan with the head in flexed position showing foramen magnum decompression. Atlantodental interval is normal. (c) Computed tomography scan with the sagittal cut passing through the facets showing the facets are in alignment. (d) 3-D computed tomography scan showing foramen magnum decompression and resection of arch of atlas



Figure 4: Postoperative images (Case 2). (a) Computed tomography scan showing the atlantoaxial implant. (b) Delayed postoperative magnetic resonance imaging showing reduction in the syringomyelia

decompression has been the gold standard form of surgical treatment. Despite several discussions, a consensus regarding nature and extent of foramen magnum decompression has not been reached. In 1998, we suggested, for the first time in the literature, that opening of the dura may not be necessary in cases with Chiari formation associated with basilar invagination with or without the presence of syringomyelia.^[24] On the basis of our study, we identified that in cases with Chiari formation in association with basilar invagination, there is a small volume posterior cranial fossa, and we had recommended foramen magnum decompression. Small volume posterior cranial fossa that is unable to accommodate the cerebellar volume has been generally accepted to be the cause of Chiari formation and accordingly justified the philosophy of foramen magnum decompression as the form of surgical treatment.

Despite the fact that foramen magnum decompression with or without duroplasty and with or without tonsillectomy has been popularly performed, analysis of a number of major reports suggests a failure rate that ranges from 15% to 50%.^[25-29] In a recently conducted literature survey by Lu *et al.*, the incidence of postoperative complications after posterior fossa bone decompression only was in 11.8% of cases, and in cases where posterior fossa bone and dural decompression was done, the incidence was 15.6%.^[26] Paul *et al.* reported 82% improvement in symptoms after foramen magnum decompression but observed a 21% relapse rate.^[27] Klekamp demonstrated 66% success rate at 5 years after surgery that involved foramen magnum decompression.^[28] A recent review of 116 series dealing with 4206 patients with Chiari formation and treatment by various forms of foramen magnum decompression identified that 3150 (75%) were reported to be improved, 697 (16.6%) had no change in

neurological status, and 359 (8.5%) experienced worsening.^[29] The authors identified that neurological worsening ranged from 5% to 43%. Essentially, the analysis of the multiple large series suggests that foramen magnum decompression surgery has its own share of procedure-related complications. Despite the complications, no definite remedy protocol has been suggested in any reported series.

In our earlier published work, we identified that atlantoaxial instability is the nodal point of pathogenesis of Chiari formation and atlantoaxial stabilization is the aim and atlantoaxial arthrodesis is the final goal of treatment.^[1,2,14] Foramen magnum bone and dural decompression or neural manipulation was considered to be a counter-productive surgical procedure. Atlantoaxial fixation was done in the presence or absence of any bone abnormality of the region and in the presence or absence of syringomyelia. Chiari formation was discussed as a natural protective measure rather than a pathological event. It provided a “soft cushion” and prevented pinching of neural structures between bones in the event of potential or manifest atlantoaxial instability.^[30] Chiari formation appears to be an outcome of longstanding atlantoaxial instability and has a role in stalling or delaying the clinical manifestations. We likened Chiari formation to protective “air bag” of a car. The nature and probably the subtlety of the dislocation provided an opportunity for the natural body processes to remodel the body tissues.

Atlantoaxial instability has been conventionally identified on the basis of dynamic imaging and assessment of the status of odontoid process in relationship to the anterior arch of atlas. In addition, dural or neural compression, by the odontoid process, has been considered to be the evidence of instability. Our recent analysis suggested that there could be atlantoaxial instability even in the absence of abnormality of atlantodental interval or compressive evidence of neural structures at craniovertebral junction. Apart from other known evidence, we evaluated atlantoaxial instability on the basis of facet alignment on the lateral profile imaging with the head in neutral position and on the basis of manual manipulation of bones during surgery. The term central or axial atlantoaxial instability was used for type 2 and 3 atlantoaxial facet instability as per the classification system described by us.^[31] It was recognized that there could be atlantoaxial instability even in the absence of radiological evidence. As our experience in the subject has grown over the years, the presence of Chiari formation by itself was considered to be an evidence of atlantoaxial instability and atlantoaxial stabilization was performed as the treatment. Although inclusion of the occipital bone in the fixation construct is a popular technique, we have identified that instability in such

cases is limited to atlantoaxial instability.^[32] The stabilization techniques must focus on atlantoaxial fixation.

As the results of clinical outcome of our treated cases have become evident, it is clear that atlantoaxial instability forms the basic pathological substrate of Chiari formation both in adults and in pediatric population groups.^[1,2,14] Our further evaluations of craniovertebral junction-related issues conclude that atlantoaxial instability is the basis of a number of musculoskeletal abnormalities, such as basilar invagination, bone fusions or Klippel-Feil abnormality, assimilation of atlas, C2-3 fusion, bifid posterior arch of atlas, platybasia, short neck, and torticollis, as well as neural abnormalities, such as Chiari formation and syringomyelia, when they are present in conjunction or in isolation.^[33-37] The observation has significant clinical relevance and suggests the need for atlantoaxial fixation in all the mentioned clinical entities and futility of foramen magnum decompression surgery as the mode of surgical treatment. As we identified decrease in the size of syrinx and reversal of tonsillar herniation, we identified reversal of musculoskeletal changes of basilar invagination that begins in the immediate postoperative period following atlantoaxial fixation.^[34] A short neck can become normally long, and torticollis can reverse. We speculated that bone fusions have the potential to “un-fuse” following atlantoaxial fixation surgery.

The clinical grading scheme discussed by us in the present report and in our earlier publications was seen to be simple and reproducible and provided a clear image of the clinical profile and status of the patient.^[1] Due to the relatively significant number of patients, we have included the parameter of voice affection in the grading. Other authors have not yet validated the grading scale. To avoid confusion and potential or possible flaws in our grading scale, we have additionally used the universally accepted grading scheme of JOA.

In our earlier published clinical series wherein atlantoaxial fixation was done for Chiari formation, we observed clinical neurological improvement in 100% of cases. Although the duration of follow-up is relatively short in the presented series, a significant and remarkable clinical improvement in 100% of our cases that had failed foramen magnum decompression surgery following atlantoaxial fixation further reinforces the belief that atlantoaxial instability is the cause of Chiari formation. The very fact that improvement started in the early postoperative period is suggestive of the prominent role of atlantoaxial instability in the genesis of symptoms. Voice improvement (7 patients), reduction of hoarseness of voice, improvement in breathing (10 patients), relief from

disabling pain in the neck and hands, ability to walk again without support (22 patients), and possibility of removal of Ryle’s tube as a form of feeding (5 patients) were major successes in the series.

CONCLUSIONS

Atlantoaxial instability is the basic cause of Chiari formation. Atlantoaxial stabilization is a rational and philosophical mode of surgical treatment. Treatment by foramen magnum decompression needs to be reevaluated. Cases where foramen magnum decompression has failed should be treated by atlantoaxial fixation.

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Conflicts of interest

There are no conflicts of interest.

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