Irreducible basilar invagination and atlanto-axial dislocation: Are they really "irreducible"?

Basilar invagination (BI) is a condition characterized by telescoping of the upper cervical spine (more specifically, the odontoid process) into the foramen magnum.[1-8] BI may or may not be associated with atlanto-axial dislocation (AAD).

The symptoms are usually very severe and consist of progressive neurological deficits. Conventional strategies over the past three decades included a transoral excision of odontoid process, followed by a posterior instrumented fixation.[9-12] More recently, it has been shown that distraction at the C1/C2 joints leads to a possibility of reduction of BI even in "irreducible" cases.[4,6,9-14] This has led to a possibility of considering BI with AAD pathology similar to "spondylolisthesis," which may be reduced through a posterior approach only using certain "specific" intraoperative manipulations. There have been a few studies where, by using intra-operative manipulations such as distraction and other movements, satisfactory reduction has been obtained. One of the oldest techniques was that described by Sonntag *et al*.[15] way back in 2004.

Following this, there have been some more technique descriptions of reducing BI and AAD from a posterior approach only.[16-29]

EVOLUTION OF MANAGEMENT

AAD and BI is a highly complex pathology that can cause progressive cervico-medullary compression and disabling deficits. The underlying principle of treatment is not just to relieve the compression but also to provide optimal stability and correction of deformity.[2,4,5,30-37] In contrast to dislocation caused by trauma, inflammation, and other factors, AAD associated with BI is mainly due to congenital etiology. It is considered irreducible in the majority of patients, as it does not reduce on dynamic X-rays or with cervical traction. Perhaps this definition is not so accurate as traction itself does not provide optimal forces to reduce the deformity. The understanding of this disease is closely linked to the development of various surgical strategies.

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The development of surgical strategies may be roughly divided into three phases of development as follows.

Era of Palliative Fusions

Before the evolution of transoral decompression and odontoidectomy, surgeons understood the grave nature of this pathology.[38-41] Due to unavailability of optimal micro-neurosurgical techniques, they suggested open surgery, posterior decompression, and autologous bone fusion in an attempt to arrest of the progress of the pathology. However, this was not enough in most cases, and the disease relentlessly progressed leading to death or permanent disability.

Era of Decompression and Instrumented Fusions

With the development of better micro-neurosurgical techniques, transoral procedure slowly started becoming a standard for relieving the compression ventrally. This was followed by posterior instrumented fusions. This procedure was followed for almost three decades. For a long time, the posterior procedure was performed at a second stage usually within a week after transoral procedure during which the patient was kept on skeletal traction to maintain stability.[9-11,42-44] Over a period of time, with increase of expertise, both procedures were being performed during the same sitting. Wang *et al*. have also suggested a transoral release of the ligaments around the odontoid process[12,32] as the first stage of the treatment, followed by a posterior instrumented fixation in a second surgery. Our institute has followed a standard policy of transoral excision of odontoid process, followed by posterior instrumented fixation for nearly three decades.[45] The same has been true for other centers worldwide.[9-11,13,42,46] Dickman *et al*.[42] reported that the complication rate of transoral surgery, even in experienced hands, was 9.4% (14 of 148 patients). Complications included cerebrospinal fluid leakage, wound dehiscence, wound infection, pneumonia, etc. Death also occurred in 2% of patients (3 of 148 patients).

Era of Intraoperative Manipulations and Realignment

The conventional understanding of management of BI and AAD depended on its "reducibility," i.e. whether it would reduce on traction or not.[35,44,46] While this may occur for some AADs, BIs usually cannot be reduced on traction[4,5] and it was well accepted that these cases must be treated with decompression and stabilization only without correction of deformity.[9-12,44] With the introduction of Goel's technique, the differentiation between reducible and irreducible AAD and BI became more blurred, as it was shown that BIs could be realigned by distraction (by placement of spacer within the joint) along with a C1/C2 fixation.[4,5,13,47]

Intraoperative manipulation to reduce BI and AAD is relatively a new concept. Jian *et al*.[17] introduced a concept of intraoperative distraction cases of BI with assimilated C1 arch, where a rod was connected to a C2 pedicular screw and occipital screw, following which distraction was performed reducing both BI and AAD. They achieved satisfactory results. However, the shortcoming of this procedure was that it could provide distraction only as a method of reduction for both AAD and BI. AAD for its optimal reduction also requires a forward movement of dens as compared to BI, which requires only a vertical distraction. This is reflected in their results, where BI could be reduced in almost all patients, but the AAD could be reduced completely in only 85% of their cases. In addition, distraction only without a spacer placement carries a risk of re-settling, which also was reflected in some of their cases.

Hsu *et al*.[16] overcame this shortcoming by describing a novel technique in two cases of acquired (one infection and other in metastasis) occipito-cervical instability. Here, apart from intraoperative occipito-cervical distraction, they also provided an extension of neck by applying compression between the upper occipital screw and another screw tightened more superiorly on the rod, which resulted in correction of AAD. This technique clearly demonstrated that while distraction corrects BI, extension while maintaining distraction results in correction of AAD.

We have been using a technique of first placing spacers to distract the joint to correct the BI; the spacers were then used as a fulcrum over which compression and extension was provided, which then corrected the AAD. Since the procedure involved movements of distraction, compression, and extension, we named it as Distraction, Compression, and Extensive Reduction (DCER). We used it satisfactorily for all irreducible cases of BI and AAD and now have an experience of over 50 cases. This technique varied depending on whether the C1 arch was assimilated or not. In cases where the C1 arch was not assimilated, we preferred C2 translaminar screw connected to rod to C1 lateral mass screw. We preferred this over a C2 pedicular screw as it was easier to place and has been shown to have equal pullout strength as pedicular screws.[48-50] In addition, C2 translaminar screw provides a longer lever arm. Thus, the amount of force transmitted into the C2 bone may be less, providing a better chance of long-term screw retention as compared to C2 pedicular screw. C2 translaminar screws also form a torque with C1 lateral mass screws, thus resulting in a better horizontal compressive force along with a vertical compressive force as well. C2 laminar screw does not have any risk of vertebral artery injury also, which is a possibility especially in patients with congenital BI and AAD. Thus, because of all these reasons, we believe that C2 translaminar/C1 lateral mass screws formed a better and a stable construct and serve an ideal model for DCER in cases where C1 arch was not assimilated.

In cases where the C1 arch was not assimilated, we used an occipital and C2 translaminar screws as they provided long lever arms, thus once again reducing the amount of force being transmitted into the bone.

Using the technique of DCER, we were able to reduce the AAD completely in 94% cases and BI satisfactorily in all cases. We feel that this technique may be used satisfactorily in all cases of BI and AAD. In cases where the C1 arch is thin or broken, an occipito-C2 DCER may be performed.

LIMITATIONS AND CONCERNS OF INTRAOPERATIVE MANIPULATIONS

The primary concern with our technique involves manipulation of the cervical spine, and the concern of overdistraction and its consequent effects on the cervico-medullary junction cannot be ignored. To minimize this, it is important to provide distraction of not more than 5–6 mm, just enough to place in the spacers. Hsu *et al*.[16] have described the use of intraoperative ultrasound to ensure adequate spinal cord decompression during the reduction maneuvers. We have only used fluoroscopic guidance during DCER.

It is also important to note that patient selection is critical to ensure successful application of this technique. The reduction maneuvers place a high degree of stress across the bone–screw interface, though much less than those described earlier,[16,17] due to the spacers also acting as a channel for significant weight transmission. However, it is imperative that the screws have adequate cortical purchase to prevent hardware failure. Medical conditions that would reduce bone strength (e.g. osteoporosis, acute infection) are relative contraindications to this procedure. It is important to note that in few of the cases, it may not be possible to completely reduce the AAD and BI. In these cases, one should not still shy away from performing a transoral procedure.

CONCLUSIONS

BI with AAD is a complex pathology to manage. With the development of better imaging techniques, micro-neurosurgical techniques, along with remarkable development of instrumentation, it is now possible to develop surgical methodologies which can provide intraoperative manipulation to reduce the BI and AAD during surgery. These procedures have demonstrated that the definition between reducible and irreducible BI and AAD is blurred. In almost all cases, optimal alignment may be provided for this pathology, hitherto considered earlier not to be possible. These techniques provide an immense scope for development of specialized instrumentations in future, which would allow complete reduction, and realignment of BI and AAD to be performed during surgery itself.

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REFERENCES

1. Ding X, Abumi K, Ito M, Sudo H, Takahata M, Nagahama K, *et al*. A retrospective study of congenital osseous anomalies at the craniocervical junction treated by occipitocervical plate-rod systems. Eur Spine J2012;21:1580-9.
2. Fenoy AJ, Menezes AH, Fenoy KA. Craniocervical junction fusions in patients with hindbrain herniation and syringohydromyelia. JNeurosurg Spine 2008;9:1-9.
3. Furlan JC, Kalsi-Ryan S, Kailaya-Vasan A, Massicotte EM, Fehlings MG. Functional and clinical outcomes following surgical treatment in patients with cervical spondylotic myelopathy: A prospective study of 81 cases. J Neurosurg Spine2011;14:348-55.
4. Goel A. Treatment of basilar invagination by atlantoaxial joint distraction and direct lateral mass fixation. J Neurosurg Spine 2004;1:281-6.
5. Goel A. Progressive basilar invagination after transoral odontoidectomy: Treatment by atlantoaxial facet distraction and craniovertebral realignment. Spine (Phila Pa 1976) 2005;30:E551-5.
6. Goel A. Double insurance atlantoaxial fixation. Surg Neurol 2007;67:135-9.
7. Goel A. Basilar invagination, Chiari malformation, syringomyelia: A review. Neurol India 2009;57:235-46.
8. Menendez JA, Wright NM. Techniques of posterior C1-C2 stabilization. Neurosurgery 2007;60(1 Supp1 1):S103-111.
9. Landeiro JA, Boechat S, Christoph Dde H, Gonçalves MB, Castro I, Lapenta MA, *et al*. Transoral approach to the craniovertebral junction. Arq Neuropsiquiatr 2007;65:1166-71.
10. Lee ST, Fairholm DJ. Transoral anterior decompression for treatment of unreducible atlantoaxial dislocations. Surg Neurol 1985;23:244-8.
11. Mummaneni PV, Haid RW. Transoral odontoidectomy. Neurosurgery 2005;56:1045-50; discussion 1045-50.
12. Wang T, Zeng B, Xu J. Transoral reduction of irreducible posteriorly displaced odontoid fracture. Eur Spine J2011;20 (Suppl 2):S227-30.
13. Goel A, Pareikh S, Sharma P. Atlantoaxial joint distraction for treatment of basilar invagination secondary to rheumatoid arthritis. Neurol India 2005;53:238-40.
14. Goel A, Shah A. Atlantoaxial joint distraction as a treatment for basilar invagination: A report of an experience with 11 cases. Neurol India 2008;56:144-50.
15. Kim LJ, Rekate HL, Klopfenstein JD, Sonntag VK. Treatment of basilar invagination associated with Chiari I malformations in the pediatric population: Cervical reduction and posterior occipitocervical fusion. J Neurosurg 2004;101(2 Suppl):189-95.
16. Hsu W, Zaidi HA, Suk I, Gokaslan ZL, Wolinsky JP. A new technique for intraoperative reduction of occipitocervical instability. Neurosurgery 2010;66(6 Suppl Operative):319-23; discussion 323-14.
17. Jian FZ, Chen Z, Wrede KH, Samii M, Ling F. Direct posterior reduction and fixation for the treatment of basilar invagination with atlantoaxial dislocation. Neurosurgery 2010;66:678-87; discussion 687.
18. Cronin CG, Lohan DG, Mhuircheartigh JN, Meehan CP, Murphy J, Roche C. CT evaluation of Chamberlain's, McGregor's, and McRae's skull-base lines. Clin Radiol 2009;64:64-9.
19. Kwong Y, Rao N, Latief K. Craniometric measurements in the assessment of craniovertebral settling: Are they still relevant in the age of cross-sectional imaging? AJR Am J Roentgenol 2011;196: W421-5.
20. A W. Roentgen diagnosis of the cranio- vertebral region. New York, NY: Springer-Verlag; 1974.
21. McRae DL, Barnum AS. Occipitalization of the atlas. Am J Roentgenol Radium Ther Nucl Med 1953;70:23-46.
22. Goel A, Shah A. Vertical atlantoaxial dislocation as a cause of failure of midline fixation. J Clin Neurosci 2010;17:1345-6.
23. Goel A, Shah A, Gupta SR. Craniovertebral instability due to degenerative osteoarthritis of the atlantoaxial joints: Analysis of the management of 108 cases. J Neurosurg Spine 2010;12:592-601.
24. Goel A, Shah A, Rajan S. Vertical mobile and reducible atlantoaxial dislocation. Clinical article. J Neurosurg Spine2009;11:9-14.
25. Goel A, Sharma P. Craniovertebral realignment for basilar invagination and atlantoaxial dislocation secondary to rheumatoid arthritis. Neurol India 2004;52:338-41.
26. Mummaneni PV, Lu DC, Dhall SS, Mummaneni VP, Chou D. C1 lateral mass fixation: A comparison of constructs. Neurosurgery 2010;66(3 Suppl):153-60.
27. D'Agostino R, Tarantino V, Calevo MG. Blunt dissection versus electronic molecular resonance bipolar dissection for tonsillectomy: Operative time and intraoperative and postoperative bleeding and pain. Int J Pediatr Otorhinolaryngol 2008;72:1077-84.
28. Cherekaev VA, Bekiashev A, Filippov Iu A, Belov AI, Gol'bin DA. Experience in using a molecular resonance coagulator in neurooncology. Zh Vopr Neirokhir Im N N Burdenko 2005:33-6.
29. Ma XY, Yin QS, Wu ZH, Xia H, Riew KD, Liu JF. C2 Anatomy and Dimensions relative to translaminar screw placement in an Asian population. Spine (Phila Pa 1976) 2010 Feb 26 [Epub ahead of print].
30. Goel A. High cervical C3-4 'disc' compression associated with basilar invagination. Neurol India 2008;56:68-70.
31. Nishikawa M, Ohata K, Baba M, Terakawa Y, Hara M. Chiari I malformation associated with ventral compression and instability: One-stage posterior decompression and fusion with a new instrumentation technique. Neurosurgery 2004;54:1430-4; discussion 1434-5.
32. Wang C, Yan M, Zhou HT, Wang SL, Dang GT. Open reduction of irreducible atlantoaxial dislocation by transoral anterior atlantoaxial release and posterior internal fixation. Spine (Phila Pa 1976) 2006;31:E306-13.
33. Goel A, Laheri V. Plate and screw fixation for atlanto-axial subluxation. Acta Neurochir (Wien) 1994;129:47-53.
34. Aryan HE, Newman CB, Nottmeier EW, Acosta FL Jr., Wang VY, Ames CP. Stabilization of the atlantoaxial complex via C-1 lateral mass and C-2 pedicle screw fixation in a multicenter clinical experience in 102 patients: Modification of the Harms and Goel techniques. J Neurosurg Spine 2008;8:222-9.
35. Madawi AA, Casey AT, Solanki GA, Tuite G, Veres R, Crockard HA. Radiological and anatomical evaluation of the atlantoaxial

transarticular screw fixation technique. J Neurosurg 1997;86:961-8.
36. Abumi K, Takada T, Shono Y, Kaneda K, Fujiya M. Posterior occipitocervical reconstruction using cervical pedicle screws and plate-rod systems. Spine (Phila Pa 1976) 1999;24:1425-34.
37. Jun BY. Complete reduction of retro-odontoid soft tissue mass in os odontoideum following the posterior C1-C2 tranarticular screw fixation. Spine (Phila Pa 1976) 1999;24:1961-4.
38. Goodbody RA, Roberts LV. Basilar invagination in Paget's disease. Lancet 1950;1:809-11.
39. Allison FG, Penner DW. Paget's disease of skull with basilar invagination (platybasia) and internal hydrocephalus. Can Med Assoc J 1951;65:476-8.
40. Cossa P, Duplay M, Martin J, Aubert, Ducos, Hemmi M. Arnold-Chiari malformation, basilar impression, basilar invagination, platybasia, convexobasia, aplasia of the basilar lamina: Report of two cases. Rev Neurol (Paris) 1951;85:392-8.
41. Mateos Gomez JH, Dorfsman Figueroa J. Basilar invagination. Rev Med Hosp Gen (Max) 1961;24:181-5.
42. Dickman CA, Locantro J, Fessler RG. The influence of transoral odontoid resection on stability of the craniovertebral junction. J Neurosurg 1992;77:525-30.
43. Hwang SW, Heilman CB, Riesenburger RI, Kryzanski J. C1-C2 arthrodesis after transoral odontoidectomy and suboccipital craniectomy for ventral brain stem compression in Chiari I patients. Eur Spine J 2008;17:1211-7.
44. Zileli M, Cagli S. Combined anterior and posterior approach for managing basilar invagination associated with type I Chiari malformation. J Spinal Disord Tech 2002;15:284-9.
45. Yerramneni VK, Chandra PS, Kale SS, Lythalling RK, Mahapatra AK. A 6-year experience of 100 cases of pediatric bony craniovertebral junction abnormalities: Treatment and outcomes. Pediatr Neurosurg 2011;47:45-50.
46. Mummaneni PV, Haid RW. Atlantoaxial fixation: Overview of all techniques. Neurol India 2005;53:408-15.
47. Goel A, Sharma P. Craniovertebral junction realignment for the treatment of basilar invagination with syringomyelia: Preliminary report of 12 cases. Neurol Med Chir (Tokyo) 2005;45:512-7; discussion 518.
48. Heuer GG, Hardesty DA, Bhowmick DA, Bailey R, Magge SN, Storm PB. Treatment of pediatric atlantoaxial instability with traditional and modified Goel-Harms fusion constructs. Eur Spine J 2009;18:884-92.
49. Hu Y, He XF, Ma WH, Xu RM, Ruan YP, Feng JX, *et al*. Comparison study of biomechanical test among fixation techniques of three types screw of posterior approach for C2. Zhongguo Gu Shang 2009;22:17-20.
50. Ren ZW, Ni B, Song HT, Zhang MC, Guo X, Wang MF, *et al*. A finite element investigation of bilateral atlantoaxial trans-articular screws and atlas laminar hooks instrumentation. Zhonghua Wai ke Za Zhi 2008;46:657-60.

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