Expert's comment concerning Grand Rounds case entitled “Syringomyelia with irreducible atlantoaxial dislocation, basilar invagination and Chiari I malformation” (by Shenglin Wang, Chao Wang, Ming Yan, Haitao Zhou, Liang Jiang)

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The authors describe a case of a 27-year-old woman with a 5-year history of progressive quadriparesis caused by gradual decompensation of cranio-vertebral junction (CVJ) anomaly. The bone anomalies—assimilation of atlas, synostosis of C2–3 and most probably also platybasia were complicated by anterior atlanto-axial dislocation (AAD) and basilar invagination (BI) concomitantly with Chiari I malformation and cervical syringomyelia. Due to lack of movement on dynamic flexion-extension films and no evidence of reduction after 10 min of traction under general anesthesia, the AAD was considered irreducible (IAAD) as described by Wang et al. [9]. The authors

therefore proceeded with elective tracheostomy and anterior transoral, muscular and ligamentous release without an actual odontoidectomy. The anterior release allowed for appropriate reduction of AAD that was subsequently stabilized through a posterior occipito-cervical fusion (O-C2) during the same operation. The final re-alignment was excellent and persisted during the 2-year follow-up.

I would like to commend the authors on an excellent clinical and morphological result. This and similar cases of decompensated congenital anomalies of CVJ represent one of the most challenging tasks for spine surgeons. The etiology has been the subject of discussions regarding question of which pathology is primary and which is secondary. Similarly, the selection of appropriate surgical approach is not clear [1–3, 6–8]. In general, orthopedic spine surgeons prefer posterior fixation first and neurosurgeons anterior decompression followed by posterior fixation and fusion. Overall, the experience with congenital CVJ anomalies is limited. This is especially the case in countries where systematic antenatal screening is applied, or those where congenital anomalies are detected early and thus fixed before becoming decompensated and irreducible. Such a lack of continuous experience can result in a large variability in treatment approach among different countries and surgeons.

Undoubtedly, the presented patient requires a surgical treatment to achieve an adequate medullary decompression and good cervical spine alignment with respect to long term follow up. Personally, I differ slightly in the approach to such a problem, in particular with respect to safety of the procedure.

A dynamic MRI is a better tool than plain dynamic films to demonstrate whether an adequate foramen magnum decompression is achievable in extension [2]. Even a dynamic CT scan can add further useful information [5].

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Rationale for treatment and evidence based literature-debate

Re: 2.1 Posterior fossa decompression with/without fusion

Regarding the presented case, I am not sure why a posterior fossa decompression and C1 laminectomy were not performed. This should be of no biomechanical consequence and certainly would be of benefit given a significant Chiari malformation, especially not being able to predict the postoperative effect of pure reduction.

Re: 2.2 Anterior decompression by odontoidectomy

1. This is not really true for the documented case. The target area can be reached transorally as visible on Fig. 1 (extended neck). The intraoperative photograph (Fig. 2b) also demonstrates a clearly visible odontoid tip, even with a curette above to allow for manipulation.
2. Yes, it is sometimes challenging to separate the odontoid process from surrounding structures but most of the described complications can usually be avoided by the use of a surgical microscope and micro-technique. Currently, the complication rate for transoral surgery is acceptably low. The essential point is not to open the subarachnoid space.
3. I have to disagree with the third argument against transoral decompression. Cases of recurrent BI after transoral odontoidectomy described by Goel were not all total odontoidectomies and further—not a single one underwent a posterior fixation after the anterior decompression [4].
4. This argument is also debatable in the presented case given the congenital C2–3 fusion where the C3 lateral mass screws could and, in my opinion, should be used to fortify the construct.

Re: 3.2 Anterior reduction

Transoral odontoidectomy has the same effect on stability as the extended release described by authors. Even if the transverse ligament is left in place behind the odontoid process, this structure could not be of any value because of AAD.

Technically, it is not clear to me how a resection of the transverse ligament was performed with the odontoid process still in place. Possibly the ligament was cut at its attachments but not resected. Such maneuver is difficult and certainly more dangerous than odontoidectomy itself.

Other surgeons supporting authors' opinion are mentioned but unfortunately not cited.

Procedure

After the induction of general anesthesia, the authors applied 8 kilograms of traction (no muscle relaxants are mentioned) and waited 10 min without success. Neither the traction attempt nor the prone positioning was monitored electrophysiologically (SSEP, MEP). Without experience, this represents an unnecessary risk taking. The need for a tracheostomy for a simple anterior release is also questionable.

A partial C1 anterior arch resection is described followed by apical ligament removal—this is difficult if the odontoid tip (according to Fig. 1e) is not distracted from its initial position in foramen magnum that could be achieved after an anterior ligamentous and muscular release. Nevertheless, the anterior arch of C1 was transected or removed (as seen on Fig. 3d).

The retro-odontoid tissue disappeared after 6 months but this unpredictable appearance of soft mass represents another additional risk of a “closed reduction maneuver”.

The follow up imaging shows an excellent realignment of cranio-cervical junction and slight decrease of the medullary syrinx size. I cannot comment on the resolution of Chiari malformation as midline sagittal images were provided for follow up (Fig. 4c—different plane to 3c and 1c).

Personal preference

I prefer to attempt a preoperative closed reduction for a longer period of time, usually for at least 3 days of gradually increasing halo ring traction. If unsuccessful, I proceed with microsurgical transoral resection of assimilated anterior atlas arch and transoral odontoidectomy under continuous electrophysiological monitoring. I perform anterior atlantoaxial joint release if necessary. This is always followed by a posterior fixation with or without decompression during the same operative session. When turning the patient prone, a halo vest could be of certain advantage, however, even with a free hand manipulation, evoked potentials are always monitored. The turn is the most dangerous part of the procedure and, without monitoring, a significant morbidity will go unnoticed. In prone position, a bony posterior fossa decompression and occipito-cervical fusion is performed with a midline occipital bone (thicker and stronger) screw anchorage. I would extend the construct to the lateral masses of C3 because of

the congenital fusion. Angular reduction, if necessary, for cervical spine alignment and sagittal balance, is performed manually with the head fixed in a halo ring (again under monitoring). Alternatively, posterior distraction or angular correction of the atlanto-axial joint can be achieved by means of C2 pedicle screw levers or distraction forceps.

Conclusion

In my opinion, a pure anterior release alone is a controversial technique in cases where the surgeon has made a decision to perform an anterior transoral surgery for the treatment of IAAD. If the deformity can be reduced by a pure anterior release procedure, the remaining odontoid process does not aid in any further stability and only represents an unnecessary risk for the rest of the procedure, especially positioning and posterior closed reduction. Should an anterior release not result in adequate reduction, then a complete odontoidectomy and anterior neural decompression is required anyway.

References

1. Abumi K, Takada T, Shono Y, Kaneda K, Fujiya M (1999) Posterior occipitocervical reconstruction using cervical pedicle screws and plate-rod systems. *Spine* 24:1425–1434
2. Fenoy AJ, Menezes AH, Fenoy KA (2008) Craniocervical junction fusions in patients with hindbrain herniation and syringohydromyelia. *Spine* 9:1–9
3. Goel A (2004) Treatment of basilar invagination by atlantoaxial joint distraction and direct lateral mass fixation. *J Neurosurg Spine* 1:281–286
4. Goel A (2005) Progressive basilar invagination after transoral odontoidectomy: treatment by atlantoaxial facet distraction and craniocervical realignment. *Spine* 30:E551–E555
5. Goel A, Shah A, Rajan S (2009) Vertical mobile and reducible atlantoaxial dislocation. *J Neurosurg Spine* 11:9–14
6. Kerschbaumer F, Kandziora F, Klein C, Mittlmeier T, Starker M (2000) Transoral decompression, anterior plate fixation, and posterior wire fusion for irreducible atlantoaxial kyphosis in rheumatoid arthritis. *Spine* 25:2708–2715
7. Menezes AH (2008) Craniocervical junction database analysis: incidence, classification, presentation, and treatment algorithms. *Childs Nerv Syst* 24:1101–1108
8. Mouchaty H, Perrini P, Conti R, Di Lorenzo N (2009) Craniocervical junction lesions: our experience with the transoral surgical approach. *Eur Spine J* 18:S13–S19
9. Wang Ch, Yan M, Zhou HT, Wang SL, Dang GT (2006) Open reduction of irreducible atlantoaxial dislocation by transoral anterior atlantoaxial release and posterior internal fixation. *Spine* 31:E306–E313