Indian Journal of Orthopaedics Surgery 2019;5(4):282–286

Content available at: iponlinejournal.com

Indian Journal of Orthopaedics Surgery

Journal homepage: www.innovativepublication.com

Case Report

Occipito -cervical fusion in Cranio-Vertebral anomalies with basilar invagination

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A R T I C L E I N F O

Article history:
Received 20-11-2019
Accepted 27-12-2019
Available online 06-01-2020

Keywords:
Basilar invagination
AtlantoAxial dislocation
Platybasia
OC Fusion
Craniovertebral junction anomaly
Brainstem compression

A B S T R A C T

Basilar invagination are uncommon syndrome that occurs when the superior part of the odontoid (part of the C2 vertebrae) migrates upward through foramen magnum and that causes bulbomedullary compression. Basilar invagination are divided on bases of that causation in trau matic and other one is developmental. Mostly seen basilar invagination are developmental. Here with describe a case of 13yr old female with developmental craniovertebral junction abnormality in form of congenital atlanto axial dislocation with Platiebasia with basilar invagination which is treated with decompression and occipital plate and screw based posterior fixation with its clinical as well as radiological outcome.

1. Introduction

Basilar invagination has several etiologies it should be considered a radiographic finding and the underlying etiology should be identified. Basilar invaginations are classified as developmental and traumatic . Among the reported etiologies of developmental basilar invagination are clival hypoplasia, condylar hypoplasia, hypoplasia of the atlas, incomplete ring of atlas with spreading of the lateral masses, achondroplasia, and atlanto -occipital assimilation. “Basilar impression” is the term used to describe the acquired form of basilar invagination, which results from softening of the bone at the base of the skull. Common conditions leading to basilar impression are Paget’s disease, rehumatoid arthritis, osteomalacia, hyperparathyroidism, osteogenesis imperfecta, Hurler syndrome, rickets, and skull base infection.

The Chamberlain line- from the dorsal lip of foramen magnum (opisthion) to the posterior portion of the hard palate. The superior tip of odontoid usually does not exceed beyond this line, extension beyond 5 mm is typically considered abnormal.

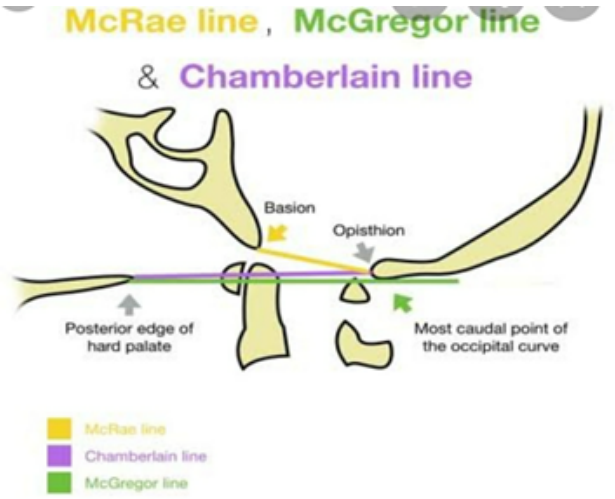

Fig. 1: Normal anatomical lines

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https://doi.org/10.18231/j.ijos.2019.057

McRae line- delineates the foramen magnum from anterior margin to the posterior border, for which the odontoid tip should not cross.

McGregor described a modification of the Chamberlain line from the posterior portion of the hard palate to the lowest edge of the midline occipital surface.

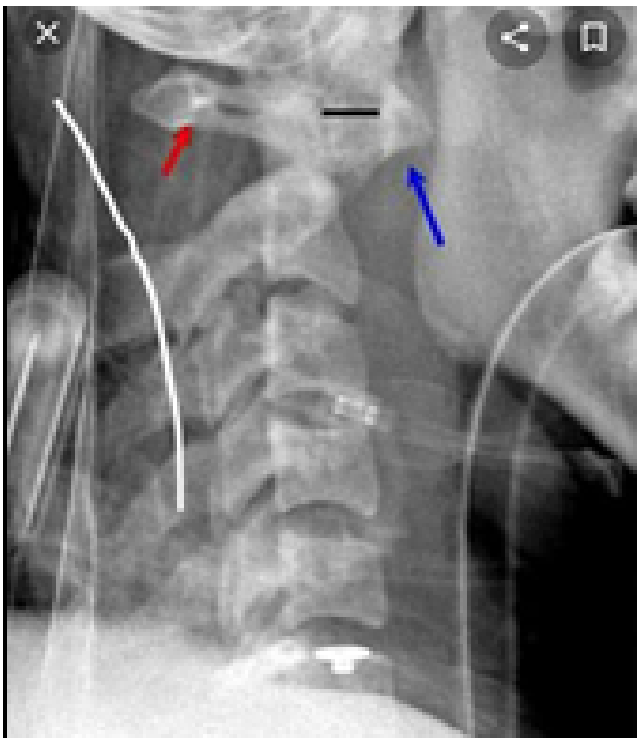

Fig. 2: Atlanto-Axial Dislocation

1.1. Predental space

Space between anterior arch of atlas and axis of dens.

Indications of AAD are >3mm in adults & >5mm in children.

Goel's classification of atlanto axial instability

Type 1- Anterior atlanto axial facetal dislocation

Most common as well as most symptomatic type

Type 2- Posterior atlanto axial facetal dislocation

Type 3- Central atlanto axial facetal dislocation

Generally type 2 and type-3 predental space is not affected and they are generally less symptomatic. Generally they become symptomatic after trauma.

1.2. Platybasia

The association of platybasia and basilar invagination is known.earlier in studies both entites are described as synonymus by Chamberlain. Subsequently, various authors have questioned the clinical significance of platybasia and have considered it to be of anthropological significance only.in platybasia there is a flattening of skull bone so it was associated with a more horizontal angulation and shortening of the clivus which resembles as superior positioning of odontoid process. Klaus also identified two groups of basilar invagination on the basis of the clival line of Wackenheim. He noted that in basilar invagination associated with platybasia, the tip of the odontoid process almost never reaches the Wackenheim clival line, while in a steeply shelving or normal clivus, the line from the dens often reaches or even over-shadows it. Platybasia was seen in both the groups but was relatively less in number and severity in Group A. From the study of Group B patients, it appears that platybasia was as important as invagination of the odontoid process in causing the anterior concavity of the brainstem and in reducing the volume of the posterior fossa. Marin-Padilla concluded from their study that the Chiari-like deformities reflect the effects of clival and occipital molding, which act mainly anteriorly. Platybasia did not directly result in any neurological symptoms, but it participated with basilar invagination in critically reducing the posterior cranial fossa volume. In our recent study, we demonstrated reversal of platybasia in Group A patients following the craniovertebral realignment surgery that involved distraction of facets of atlas and axis.

1.3. Case description

13yr old female with complain of neck pain and b/l upper limb and lower limb weakness since 15 days which is progressive and patient became unable to walk or even stand and since last 4 days with normal bowel and bladder function without any history of trauma or any clinical complain like fever, cough etc. So patient brought to Rajkot civil for treatment where all radiological investigation done and in MRI and CT scan patient found to have predental space of 6.5mm which is suggestive of atlanto - axial dislocation and tip of dens is 11mm above level of chamberlain line which suggestive of basilar invagination with Patiebasia which cause compression over brainstem due which pateint has weakness in b/l upper and b/l lower limb. When patient came to CHA tong insertion done and traction given then traction CT scan done for checking stability of Atlanto axial joint and patient was operated oc fusion with plate and screw fixation with posterior approach and after 2 days of surgery patient's neurology got improved and post operative ct scan done.

2. Discussion

Cervical instability due to trauma or congenital pathologies are the most common indications

leading to instrumented spinal fusions in children. I Zehan et al shows fusion gives significantly better clinical outcome with low morbidty if performed with skilled surgeon who has impwerative knowledge about anatomy of

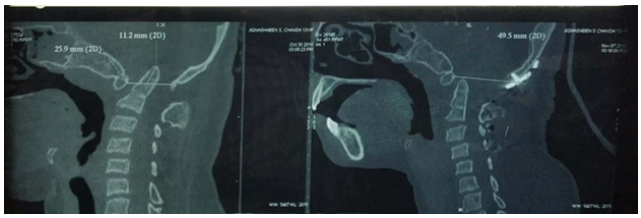

Fig. 3: Pre op CT scan (Left side), Post op CT scan(Right side)

Fig. 4: Pre op MRI

Fig. 5: Post op MRI

cranio -vertebral junction. M. lastikka shows better outcome of it in children with skeletal dysplasia.

The main indications for instrumented cervical spinal fusions in the current case was instability due to developmental atlanto -axial dislocation with basilar invaginations with platybasia. Fusion is sometimes necessary to treat and/or prevent neurologic deficits in developmental anomalies of craniovertebral junction.occipito-cervical fusion can be done with two different techniques. 1) Rigid internal fixation with screws and 2) semi rigid wiring techniques and in which first has been documented to be superior in terms of lower complication rates and higher fusion rates. The biomechanical stability of C1 to C2 fusion has also been found to be in favour of a screw system over wiring techniques in studies. It is also thought that rigid internal fixation will diminish the need and duration of long post operative external stabilisation devices, Screw/rod implants offer rigid skeletal fixation for occipito -cervical stabilization. Rigid cervical fixation is advantageous in that it minimizes axial rotation. Such fixation is attained through C1-2 transarticular screw fixation or through C1 lateral mass fixation coupled with C2 pars interarticularis, pedicle, or laminar fixation. Occipital plate is connected with rods to C1 lateral mass screws and C2 translaminar screws. Note the large surface area covered by the plate. The first step is to achieve fixation in the upper cervical spine by using techniques described previously. The next step is to bend the rods to match the anatomical configuration of the Cranio vertebral junction. Low-profile connectors allow the rods to be secured to the occiput with cortical bone screws. This avoids the use of polyaxial screws that have large heads and are therefore prone to break through skin because of inadequate soft tissue in the occiput. Alternatively, rods can also be secured to an occipital keel plate. Occipital keel plates are available in various configurations. They cover more surface of the occiput than low-profile connectors. This may hinder occipital graft placement. Bone thickness should be assessed through preoperative axial imaging studies. Ideal occipital fixation is achieved in the midline because the thin, squamous portion of the occiput does not allow sufficient screw purchase. The midline bone usually can accept a 10 mm screw. Often 12 or 14 mm screws can be placed into this structure. Rigid fixation should be achieved through at least four bicortical occipital screws. Six screws are optimal, but the patient‘s anatomy may not allow for this many to be implanted. Drilling is performed with a high-speed drill, and the drill holes should begin lateral to the midline and below the superior nuchal line. A midline screw may be utilized in certain occipital plate designs once the skull is sculpted with a burr to allow the plate to sit

flat below the superior nuchal line. The drill should be advanced very slowly. A probe is helpful for determining when the hole is bicortical. After the drill holes have been made, tapping is necessary before placing the fixation screws. Bleeding is often encountered from the bone but is usually self-limited following screw placement. Occipito -cervical fusion with lateral mass screws in C1 and pars interarticularis screws in C2. Here patient is operated for occipitocervical fixation through posterior approach without odontoidectom. Immediate post operatively forepost collor givrn to patient for stabilization Post operatively patient's improved drastically from 4/5 power in b/l lower limb and 3/5 power in b/l lower limb to 5/5 power in b/l lower limb and 4+ power in b/l upper limb so that patient impro ved to neurick grade-1 from Neurick grade-3 and in post op CT scan tip of dens is at the level of chamber lands line before surgery which is 11 mm away from the line which is significant for basilar invagination in predental space there is no significant change due partial assimilation of atlas to oociput. Many complications of occipito cervical fusion are neurological deficit, non fusion, perforation in skull, infection, abnormal motions of fused segment. For better understanding of clinical course of patient advised for regular follow up.

3. Conclusion

Here by, this is concluded that OC fusion with posterior screw and plate fixation is a treatment option in patient with developmental atlanto-axial dislocation with basilar invagination. Patient shows significant improvement clinically as well as radiologically without any significant increasing chances of immediate and early post operative complications.

4. Source of Funding

None.

5. Conflict of Interest

None.

6. References

1. Ackermann JF. Über die Kretinen, einebesondere Menschenabart in den Alpen. Gotha: Ettingersche Buchhandlung; 1790.
2. Spillane JD, Pallis CH, Jones AM. Developmental abnormalities in the region of the foramen magnum. *Brain* 1957;80:11–48.
3. Virchow R. Untersuchungenuber die Entwicklung des Schädelgrundes. *Berlin: Reimer* 1856.
4. Virchow R. Beiträgezurphysischen Anthropologie der Deutschen, mitbesonderer Berucksichtigung der Friesen. Berlin: G. Vogt; 1876. [
5. Grawitz P. BeitragzurLehre von der basilaren Impression des Schadels. *Arch Path Anat Physiol* 1880;80:449.
6. Schmidt H, Sartor K, Heckl RW. Bone malformations of the craniocervical region. In: Vinken PJ, Bruyn GW, editors. Handbook of Clinical Neurology. Vol. 32. Amsterdam: North Holland Publishing; 1978:1–98.
7. Chamberlain WE. Basilar impression (Platybasia). A bizarre developmental anomaly of the occipital bone and upper cervical spine with striking and misleading neurologic manifestations. *Yale J Biol Med* 1939;11:487–96.
8. Schuller A. Zur Roentgendiagnose der basilaren impression des schädels. *Wien Med Wochenschr* 1911;61:2594. [
9. Vet AD. Basilar impression of the skull. *J Neurol Psychiatry* 1940;3:241–250.
10. Bharucha EP, Dastur HM. Craniovertebral anomalies.(A report on 40 cases) *Brain* 1964;87:469–80.
11. Dastur DK, Wadia NH, Desai AD, Sinh G. Medullospinal compression due to atlanto-axial dislocation and sudden haematomyelia during decompression. Pathology, pathogenesis and clinical correlations. *Brain* 1965;88:897–924.
12. Menezes AH, VanGilder JC, Graf CJ, McDonnell DE. Craniocervical abnormalities. A comprehensive surgical approach. *J Neurosurg* 1980;53:444–455.
13. Menezes AH. Primary craniovertebral anomalies and hindbrain herniation syndrome (Chiari I): Database analysis. Pediatr Neurosurg 1995;23:260–269.
14. Goel A, Sharma P, Dange N, Kulkarni AG. Techniques in the treatment of craniovertebral instability. *Neurol India* 2005;53:525–533.
15. Goel A, Laheri VK. Plate and screw fixation for atlanto-axial dislocation.(Technical report) *Acta Neurochir* (Wien) 1994;129:47–53.
16. Goel A, Desai K, Muzumdar DP. Atlantoaxial fixation using plate and screw method: A report of 160 treated patients. *Neurosurg* 2002;51:1351–1357.
17. Jeanneret B, Magerl F. Primary posterior fusion C1/2 in odontoid fractures: Indications, technique, and results of transarticular screw fixation. *J Spinal Disord* 1992;5:464–75.
18. Goel A, Bhatjiwale M, Desai K. Basilar invagination: A study based on 190 surgically treated cases. *J Neurosurg* 1998;88:962–968.
19. McRae DL. Bony abnormalities in the region of foramen magnum: Correlation of anatomic and neurologic findings. *Acta Radiol* 1953;40:335–354.
20. Thiebaut F, Wackenheim A, Vrousos C. New median sagittal pneumostratigraphical findings concering the posterior fossa. *J Radiol Electrol Med Nucl* 1961;42:1–7.

21. Von Torklus D, Gehle W. The upper cervical spine. In: Von Torklus D, Gehle W, editors. Regional Anatomy, Pathology, and Traumatology: A Systematic Radiological Atlas and Textbook. New York: Grune & Stratton; 1972:1–98.
22. Goel A. Treatment of basilar invagination by atlantoaxial joint distraction and direct lateral mass fixation. *J Neurosurg Spine* 2004;1:281–286.
23. Kothari M, Goel A. Transatlantic odonto-occipital listhesis: The so-called basilar invagination. *Neurol India* 2007;55:6–7.
24. Chandra PS, Kumar A, Chauhan A, Ansari A, Mishra NK, Sharma BS. Distraction, compression, and extension reduction of basilar invagination and atlantoaxial dislocation: A novel pilot technique. *Neurosurg* 2013;72:1040–1053.
25. Ihab Zidan, Wael Fouad, occipitocervical fixation in the management of craniocervical instabilities. *Alexandria J Med* 2011:47:3:185-192
26. M. Lastikka, J. Aarnio, I. Helenius, Instrumented cervical spine fusion in children: indications and outcomes. *J Children's Orthopaed* 2017:11:6

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Cite this article: Modi JV, Mungalpara N, Desai S, Vala P. Occipito -cervical fusion in Cranio-Vertebral anomalies with basilar invagination. *Indian J Orthop Surg* 2019;5(4):282-286.