SUCCESSFUL OCCIPITOCERVICAL FUSION FOR BASILAR INVAGINATION IN A RURAL AFRICAN MISSION HOSPITAL: CASE REPORT

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ABSTRACT

Basilar invagination is a developmental anomaly in which the odontoid abnormally prolapses into the foramen magnum. Although congenital, it may remain asymptomatic until adulthood. It is a progressively worsening condition and if left untreated, it can lead to death. We aim to describe successful occipito-cervical arthrodesis of a case of basilar invagination in a resource- challenged setting.

INTRODUCTION

Basilar invagination is a primary developmental anomaly of the cranio-vertebral junction in which the odontoid prolapses into the foramen magnum (1-3).

It results in compression of the posterior fossa contents (cerebellum and midbrain) and the spinal cord (2). It has several aetiologies and associations and some of the reported are basi-occiput (clivus) hypoplasia, occipital condyle hypoplasia, atlas hypoplasia, incomplete C1 ring with spreading of the lateral masses, atlanto-occipital assimilation, achondroplasia, osteogenesis imperfecta, Klippel-Feil syndrome, Morquio syndrome and cleidocranial dysplasia (1,2). Therefore, due to its multiple possible aetiologies, basilar invagination should be considered a radiographic finding and one should endeavour to identify the underlying pathology (1).

Based on the degree of compression, patients can be asymptomatic or can exhibit symptoms consistent with compression in this region (2).

CASE REPORT

A fifteen year old male patient presented with history of progressive weakness of all the 4 limbs over a 3 month period. This was after he was stepped on his neck while playing soccer with his colleagues in school 2 weeks prior to onset. In addition, the weakness onset was preceded by tonsillitis (5 days prior). He was able micturate but had bowel incontinence. Also, he was experiencing difficulty with expectoration and coughing.

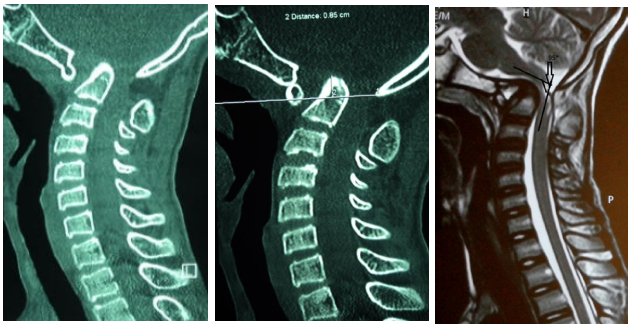
On examination, he was found to be weaker in the lower limbs (Medical Research Council grade 3) than upper limbs (MRC grade 4) and hyper-reflexic and with lower limb clonus. Sensation had been preserved. He was graded using the Ranawat neurologic grading system as stage IIIB.

His X-rays, CT scans and MRI revealed basilar invagination (8.5mm above Chamberlain line and a cervico-medullary angle of 95^0) associated with atlanto-occipital assimilation and a hypoplastic atlas (Figure 1).

Figure 1

Images of pre operative CT and MRI scans

Figure 1 *Figure 1a* *Figure 1b*



He was then put on skull traction using Gardner-Wells tongs and within 3 days he had regained his lower limb strength (MRC grade 5) and by the 4th day all his upper limb strength (MRC grade 5). Six days later he had surgery- posterior foramen magnum decompression and instrumented occipito-cervical fusion using 3.5 mm reconstruction plates and screws (as occipital and lateral mass [C3 to C5] screws) and posterior iliac crest autograft. Postoperatively he was admitted to the High Dependency Unit for 6 days then to the general ward for 3 more days before discharge home. A Philadelphia cervical collar was applied from the immediate post-operative period for 2 months then a soft cervical collar for 2 months.

He was ambulated aided with a walker then crutches for 1 week. Since then he has ambulated unaided and he is currently in Ranawat stage I. His postoperative X-rays showed good fusion by 3 months (Figure 2). He was able to return to school.

Figure 2
Post operative radiographs showing the position of the implants

Figure 2a *Figure 2b*

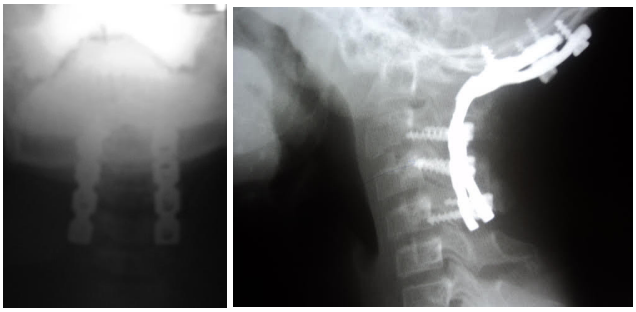
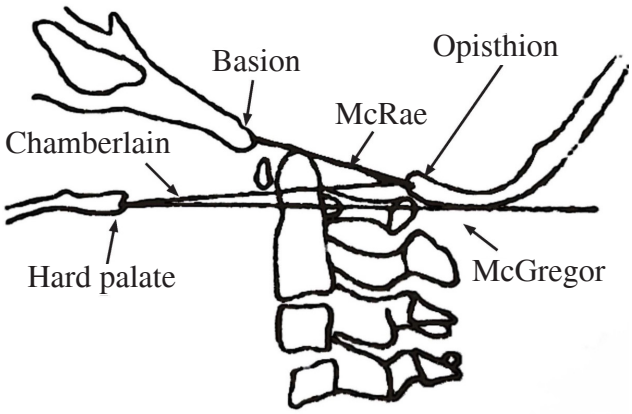


Figure 3
Image showing various landmarks and lines used in diagnosis



DISCUSSION

Although congenital, basilar invagination can remain asymptomatic and unrecognized until adulthood (1). This delay in presentation was described by Goel *et al* (3) who reported on 190 surgically treated patients with basilar invagination and stratified them into 2 groups with and without Chiari malformation. They noted that trauma was the principal precipitating factor of the symptoms in 48% of those with basilar invagination without Chiari malformation. In contrast, the other subset with basilar invagination associated with Chiari malformation, had no antecedent trauma. In addition, among those without Chiari malformation, 58% presented in the second decade of life and 86% presented within the first 3 decades- our patient's age bracket. In addition, our patient had antecedent tonsillitis and the possibility of ligamentous laxity seen in atlanto-axial rotatory instability associated with Grisel syndrome was considered too.

Basilar invagination is diagnosed by radiographic criteria (1,2). Usually using plain radiographs but CT scan and MRI have greatly expanded the ability to define cranio-vertebral anomalies (1). Definitive characterization entails use of the Chamberlain line, McGregor line or McRae line (Figure 3). When using the Chamberlain line, basilar invagination is considered present if the dens extension beyond the foramen magnum is greater than 5mm (4). As for the McGregor line the cut-off is 6mm in women and 7 mm in men (4,5); and for the McRae line, the odontoid should be below this line (1). If the sagittal diameter of the foramen magnum, is reduced to less than 19mm, neurologic deficit usually ensues (6).

Due to the compression of the posterior cranial fossa contents, there can be a compromise of the posterior inferior cerebellar artery circulation and blockage of normal cerebrospinal fluid flow. Thus, the diagnosis can be confused with syringomyelia, amyotrophic lateral sclerosis and multiple sclerosis (2).

The patient should be fully evaluated for renal, respiratory and cardiac problems if symptoms related to them are present. In addition, due to brainstem or cranial nerve compromise these patients may have problems swallowing hence impaired nutritional status (1).

Basilar invagination is a progressively worsening condition and if left untreated, it can lead to death (7). The treatment usually follows the order of traction then decompression and occipito-cervical fusion. The patients who improve on traction (reducible) usually undergo posterior decompression and fusion and those that do not improve on traction (irreducible), are first decompressed anteriorly then fused posteriorly (7-9). The patient described in this case report had the reducible type; this is similar to Goel *et al's* (3) patients, 82% of those without associated Chiari malformation improved 'almost instantaneous' after application of traction.

Since Pilcher (1910) first described occipito-cervical fusion, there has been an evolution in various surgical procedures in an attempt to improve alignment maintenance, increase fusion rates and decrease the incidence of complications (10). For adequate correction of basilar invagination, a rigid fixation anchor for the occiput and cervical spine is typically required (11).

Wiring of rib grafts from the occiput to the cervical spine is frequently used and has been shown to result in up to 99% fusion rates (12); but it is associated with prolonged immobilization due to lack of immediate stability.

Rod-screw and rod-plate systems offer rigid fixation of the occipito-cervical region (13) and hence preclude need for prolonged immobilization, especially with 'patient-unfriendly' appliances like Halo-vests. Autogenous bone graft is favoured for arthrodesis.

With this background knowledge, we elected to do occipito-cervical fusion with two 3.5mm reconstruction plates (Synthes, USA) that are easy to contour, and fixed them with 3.5mm and 4.0mm screws. Sub-axially, we placed lateral mass screws

from C3 to C5 and layered autogenous bonegraft. Fusion was noted radiographically by 3 months postoperatively (Figure 2).

CONCLUSION

Cervical spine anomalies in children and adolescents are rare, however, knowledge on how to recognize and treat these anomalies is vital to avert possible neurologic complications (2). Even in resource-challenged settings, such cases can be successfully managed with the use of available surgical armamentarium.

Disclosures: The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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