

# Basilar impression in osteogenesis imperfecta treated with staged halo traction and posterior decompression with short-segment fusion

### ABSTRACT

Basilar impression is a cranial base abnormality associated with osteogenesis imperfecta (OI) with serious neurologic implications but controversial treatment options. Combined anterior and posterior decompression with long-segment posterior fusion is often recommended. We report a patient with OI (Sillence type III) with basilar impression treated with halo traction followed by posterior surgery. The patient was a 12-year-old female with a presentation of hiccups and change in upper extremity function. Diagnostic imaging revealed syringomyelia, compensated hydrocephalus, basilar impression, and Chiari type I malformation. The patient was treated with halo traction followed by posterior decompression fusion from the occipital bone to C2. Bone fusion and improved syrinx were evident on images during the 5 years of follow-up. Five years after surgery, syrinx recurred and the fourth ventricular catheter was revised. The treatment with halo traction followed by posterior-only surgery of basilar impression associated with OI resulted in a good postoperative outcome.

**Keywords:** Basilar impression, osteogenesis imperfecta, syrinx

### INTRODUCTION

Osteogenesis imperfecta (OI) is characterized by bone fragility causing several orthopedic complications including fractures, kyphoscoliosis, and lumbar spondylolysis<sup>[1-3]</sup> and anomalies of the craniovertebral junction, which can lead to basilar invagination, basilar impression, and platybasia.<sup>[4]</sup>

In this case report, we present a patient with OI type III diagnosed with the basilar impression, Chiari type I malformation, hydrocephalus, and cervical syrinx treated with indirect decompression by halo traction, followed by posterior Chiari I decompression, placement of the fourth ventricular catheter shunt, and short-segment occipitocervical fusion.

### CASE REPORT

The patient is a 12-year-old female with OI type III with a history of severe scoliosis later treated with posterior spinal fusion and extremity fractures treated surgically. At the age of 11, she underwent a posterior fossa Chiari decompression for a Chiari malformation with syringomyelia. On routine imaging

the next year, the syrinx recurred concurrent with progressive basilar impression without marked signs of myelopathy. The patient's primary complaint was hiccups, though her mother noticed occasional difficulty with manual dexterity. While they noted no ataxia, the patient had not ambulated in a year.

During the period of diagnosis, magnetic resonance images of the cervical spine taken 6 months apart showed an increase in the size of the cervical syringohydromyelia but unchanged


**MUTLU COBANOGU<sup>1,2</sup>, JENNIFER M BAUER<sup>3</sup>,  
JEFFREY W CAMPBELL<sup>4</sup>, SUKEN A SHAH<sup>1</sup>**

Departments of <sup>1</sup>Orthopaedics and <sup>4</sup>Neurosurgery, Nemours/Alfred I. Dupont Hospital for Children, Wilmington, Delaware, USA,

<sup>3</sup>Department of Orthopaedics, University of Washington, Seattle Children's Hospital, Seattle WA, <sup>2</sup>Department of Orthopedics and Traumatology, Faculty of Medicine, Adnan Menderes University, Aydin, Turkey

**Address for correspondence:** Dr. Suken A Shah, Department of Orthopaedics, Nemours/Alfred I. Dupont Hospital for Children, 1600, Rockland Rd, Wilmington, DE 19803, USA. E-mail: suken.shah@nemours.org

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basilar impression and hydrocephalus [Figure 1]. The basilar impression was documented by magnetic resonance imaging and computed tomography. McRae's measure was 9.5 mm, Chamberlain's was 22 mm, and McGregor's was 24 mm.

The surgical plan was for indirect decompression with cervical traction in preparation for occipital cervical fusion and definitive treatment of the syrinx. A full ring carbon fiber halo was applied with 12 fixation pins at 2-inch pounds each. Traction was applied at approximately 2 pounds and gradually increased to 19 pounds over the course of 1 week, with careful monitoring of neurological symptoms and pain. She had some evidence of hypoglossal nerve traction. The weight was decreased and her symptoms resolved. The halo was applied for 2 weeks; the patient returned to the operating room at this point for posterior decompression and occipitocervical fusion. At the same time, placement of a fourth ventricular catheter to alleviate the recurrent syringomyelia was performed.

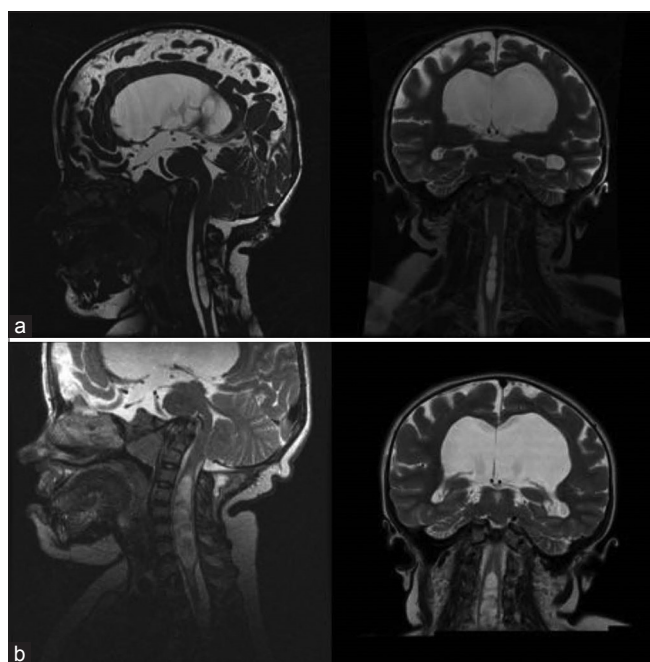
At the start of the operation, while prone, gentle traction was applied to simulate traction and indirectly decompress her basilar impression with maintained baseline neuromonitoring. The previous posterior midline exposure was used to expose

her occipital bone, foramen magnum, C1, and C2. A revision suboccipital craniectomy and decompression with the placement of a fourth ventricular-to-cervical subarachnoid catheter was performed. Preoperatively-studied vertebral anatomy could accommodate a C1–C2 transarticular screw only on the right, and a C2 isthmic pedicle screw was placed on the left with both connecting to an occipital plate with four-screw fixation [Figure 2a-g]. Bone morphogenetic protein-2 sponges and local autograft were applied to the area for fusion due to the very little calvarial periosteum. A custom halo vest was applied for 2 months after surgery and replaced with a cervical collar for 3 more months. Postoperatively, McRae's measure was 4 mm, Chamberlain's measure was 20 mm, and McGregor's measure was 22.5 mm and symptoms fully resolved. At 5-year follow-up, imaging shows stable occipital bone-C2 fusion without a change in craniocervical bony relationships [Figures 2g and 3a]. However, the cervical syrinx recurred without clear symptoms due to an occluded fourth ventricular catheter revealed at the yearly examination; the patient underwent re-exploration of Chiari decompression with the revision of the fourth ventricular subarachnoid catheter and duraplasty, resolving her symptoms [Figure 3b]. At the last outpatient visit (6 months after the second surgery), the patient was communicative. She complained about a tractable migraine without aura and status migrainosus.

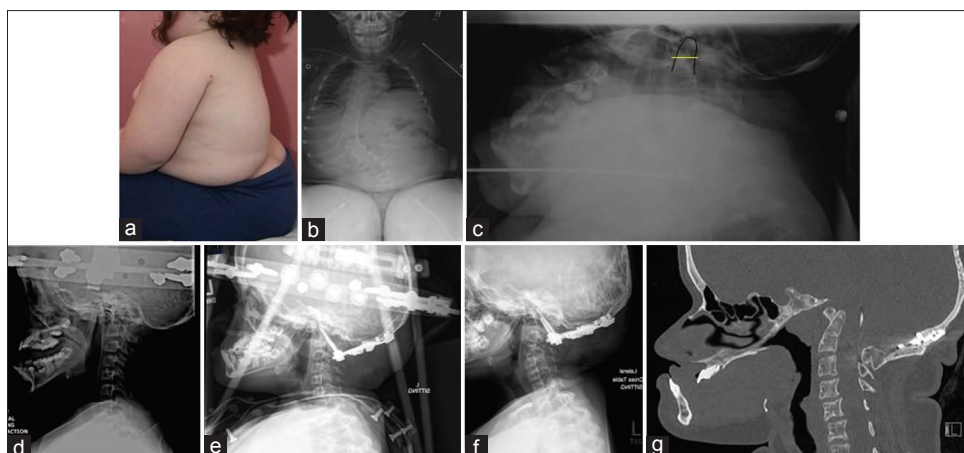
## DISCUSSION

The incidence of cranial base abnormality is observed more frequently in the more severe Type III OI.<sup>[4]</sup> Cheung *et al.* reported a prevalence of 22% for cranial base abnormality in pediatric and adult patients with OI (187 patients with type I–VII OI aged 3–47 years), some of whom had undergone bisphosphonate treatment.<sup>[5]</sup> The cranial base abnormality is present in more than one-third of patients with OI regardless of bisphosphonate treatment status.<sup>[6]</sup> Our patient presented with progressive basilar impression and underwent bisphosphonate treatment before and after surgery.

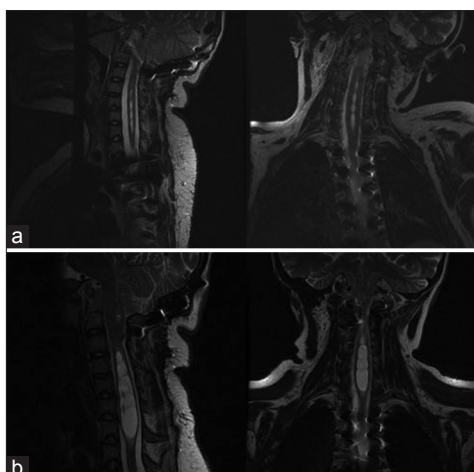
Treatment recommendations have evolved. While some originally recommended conservative Minerva brace treatment for asymptomatic cases,<sup>[3]</sup> modern recommendations are to await symptoms, then treat surgically. Surgical decompression of the brain stem is required for symptomatic cases. Initially, posterior decompression alone without fusion led to an acceleration in the progression of brain stem compression,<sup>[7]</sup> which led to a shift to anterior decompression. However, anterior approaches were technically difficult even in the absence of anatomical irregularities and had serious



**Figure 1:** (a) Initial magnetic resonance image. Sagittal and coronal images demonstrate odontoid impression on the medulla and cervicomedullary junction. No signal abnormality was seen within this portion of the brain stem or upper cervical cord. Maximal diameter of the syrinx at the cervical level measures 0.7 mm × 4.9 mm × 0.7 mm at the C6–C7 level. (b) Repeat magnetic resonance image 6 months later. Sagittal and coronal images show unchanged basilar impression but worsened syringohydromyelia. The syrinx now measures 1.3 cm × 6.5 cm × 1.2 cm in the transverse craniocaudal and anteroposterior planes and extends from C3 to T1



**Figure 2:** (a) Preoperative lateral view; clinical appearance of shortening neck. (b) Posteroanterior spine radiograph showing  $>50^\circ$  scoliosis. (c) Lateral cervical radiograph with basilar impression by McRae's line with dens outlined. (d) Preoperative halo application with distraction between cervical vertebrae. (e) Initial postoperative lateral radiograph showing occipitocervical fixation and halo vest. (f) Four months postoperative lateral imaging after halo vest removal. (g) Five-year postoperative sagittal computed tomography image showing bony fusion between the occipital bone and C2 without basilar impression progression



**Figure 3:** (a) Sagittal and coronal magnetic resonance images reveal decrease in the size of syringohydromyelia (C6–C7 measures 4 mm  $\times$  3 mm) at the 1<sup>st</sup> year of follow-up after the operation. (b) Five years after operation, basilar impression remained stable but significant increase in syrinx with maximal dimension at C6 recurred

approach-related complications such as speech disturbances and nasopharyngeal reflux, fracture of the facial skeleton and cosmetic deformity, and temporomandibular joint pain and limitation in opening the mouth.<sup>[8]</sup> Anterior decompression alone may lead to postoperative instability requiring fixation, therefore, supporting the need for fusion.<sup>[9]</sup>

Recognizing the need for decompression and fusion, treatment shifted to anterior and posterior surgery. There have been several cases of successful posterior-only surgery, as with our patient. Noske *et al.* presented a patient (type IV, 22 years old) treated with staged halo traction followed by posterior decompression and fusion, similar to our patient. The patient was symptom-free for 2 years after surgery

until worsened hydrocephalus despite fusion required a ventriculoperitoneal shunt.<sup>[10]</sup> The same posterior treatment was used successfully without recurrence by Imagama *et al.* in their patient (type IA, 29 years old).<sup>[9]</sup> Our patient was type III OI (12 years old). Despite treating the most severe survivable OI type, with current modern implants, we were able to obtain stable fusion with the shortest documented fusion length that we are aware of in the literature, stopping at C2. Prior studies extended to C6<sup>[9]</sup> and T1.<sup>[10]</sup>

## CONCLUSION

The treatment of basilar impression associated with OI can be treated by distraction with axial halo application followed by posterior-only surgery. The patients need to be monitored for the risk of recurrence in neurologic symptoms and hydrocephalus and syrinx enlargement even if there is no evidence of basilar impression progression.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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## Conflicts of interest

There are no conflicts of interest.

## REFERENCES

1. Silience D. Osteogenesis imperfecta: An expanding panorama of variants. *Clin Orthop Relat Res* 1981; 159:11-25.
2. Yilmaz G, Hwang S, Oto M, Kruse R, Rogers KJ, Bober MB, *et al.* Surgical treatment of scoliosis in osteogenesis imperfecta with cement-augmented pedicle screw instrumentation. *J Spinal Disord Tech* 2014;27:174-80.
3. Sawin PD, Menezes AH. Basilar invagination in osteogenesis imperfecta and related osteochondrodysplasias: Medical and surgical management. *J Neurosurg* 1997;86:950-60.
4. Kovero O, Pynnönen S, Kuurila-Svahn K, Kaitila I, Waltimo-Sirén J. Skull base abnormalities in osteogenesis imperfecta: A cephalometric evaluation of 54 patients and 108 control volunteers. *J Neurosurg* 2006;105:361-70.
5. Cheung MS, Arponen H, Roughley P, Azouz ME, Glorieux FH, Waltimo-Sirén J, *et al.* Cranial base abnormalities in osteogenesis imperfecta: Phenotypic and genotypic determinants. *J Bone Miner Res* 2011;26:405-13.
6. Arponen H, Evälahti M, Waltimo-Sirén J. Dimensions of the craniocervical junction in longitudinal analysis of normal growth. *Childs Nerv Syst* 2010;26:763-9.
7. Harkey HL, Crockard HA, Stevens JM, Smith R, Ransford AO. The operative management of basilar impression in osteogenesis imperfecta. *Neurosurgery* 1990;27:782-6.
8. Bertrand J, Luc B, Philippe M, Philippe P. Anterior mandibular osteotomy for tumor extirpation: A critical evaluation. *Head Neck* 2000;22:323-7.
9. Imagama S, Wakao N, Kitoh H, Matsuyama Y, Ishiguro N. Factors related to surgical outcome after posterior decompression and fusion for craniocervical junction lesions associated with osteogenesis imperfecta. *Eur Spine J* 2011;20 Suppl 2:S320-5.
10. Noske DP, van Royen BJ, Bron JL, Vandertop WP. Basilar impression in osteogenesis imperfecta: Can it be treated with halo traction and posterior fusion? *Acta Neurochir (Wien)* 2006;148:1301-5.