Teaching *NeuroImages*: Platybasia and basilar invagination in osteogenesis imperfecta

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Figure 1 Face photograph

(A) Blue sclera in osteogenesis imperfecta and (B) short neck.

A 15-year-old girl with osteogenesis imperfecta (OI) and multiple bone fractures since early childhood presented with occipital headache. She had blue sclera and short neck (figure 1), right hypoglossal palsy, bilateral pyramidal tract signs, and gaze-evoked nystagmus. Cervical MRI showed platybasia, basilar invagination, brainstem/high cord compression, and narrowing of upper cervical canal (figure 2). Cervical traction relieved headache. OI is characterized by diffuse osteopenia and bone fragility.¹

Platybasia (flattening of the skull base) with secondary basilar invagination (developmental anomaly of the craniovertebral junction in which the odontoid process abnormally prolapses upward through the foramen magnum)² in OI results from abnormally soft bone at the cranio-cervical junction and the odontoid process protruding through the foramen magnum.³ Appropriate screening may prevent severe neurologic consequences.

AUTHOR CONTRIBUTIONS

Dr. P.S. Ghosh: drafting/revising the manuscript, analysis or interpretation of data, acquisition of data. Dr. Taute: study concept or design, obtaining pictures to accompany manuscript. Dr. D. Ghosh: drafting/revising the manuscript, study concept or design, analysis or interpretation of data, study supervision.

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Figure 2 MRI of the cranio-cervical junction

(A) Narrowing of upper cervical canal (white arrow) and myelomalacia at C2 (black arrow). (B) Tip of the odontoid and anterior arch of atlas are well above Chamberlain line (white line) and the clivus canal angle is <150° (yellow line).

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Disclosure: The authors report no disclosures.

Neurology®

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Neurology 2011;77:e108

DOI 10.1212/WNL.0b013e31823648de

This information is current as of October 31, 2011

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