Commentary on: "To the Occiput or Not? C1–C2 Ligamentous Laxity in Children with Down Syndrome"

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Siemionow and Chou review the surgical management of atlantoaxial instability in the context of Down syndrome, using two interesting illustrative cases with different craniocervical abnormalities (atlantoaxial rotatory subluxation and os odontoideum). Both patients present with significant craniocervical spinal cord compression, and in this context, they highlight the specific case nuances that prompt occiptocervical fusion in one case and isolated atlantoaxial fixation in the other. However, it is important to note that while the authors refer to the presence of basilar invagination (and cranial settling) in these cases, a review of the accompanying radiographic images suggests that while significant craniocervical deformity and atlantoaxial instability is present, neither case truly has a diagnosis of basilar invagination nor cranial settling.

Basilar invagination is strictly defined as a developmental anomaly of the craniocervical junction in which the odontoid process protrudes rostrally into the foramen magnum. Basilar invagination is, in essence, a radiographic diagnosis caused by several congenital etiologies and is classically defined based on plain radiographic imaging. It is traditionally diagnosed using morphometric lines such as the Chamberlain, McGregor, or McRae line (see the comprehensive review by Smith et al[1] for further details) but current high-resolution cross-sectional imaging has greatly facilitated diagnosis. Basilar invagination is typically caused by congenital hypoplasia of one or more elements of the craniovertebral junction such as the clivus, occipital condyles, or atlas with or without occipitoatlantal assimilation.[2] Further, it is important to understand the difference between basilar invagination and conditions such as basilar impression, cranial settling and platybasia. While basilar invagination is a congenital condition, basilar impression refers to the acquired variant that results from softening of the bones of the skull base, typically secondary to conditions such as Paget disease, hyperparathyroidism, skull base osteomyelitis, or osteogenesis imperfecta. Cranial settling refers to the odontoid migration of rheumatoid arthritis that prototypically follows atlantoaxial erosive synovitis and subsequent C1 lateral mass erosion. Platybasia simply refers to a flattening of the skull base and may occur with basilar invagination or in isolation.

Correct use of this terminology is paramount, as a diagnosis of basilar invagination has management implications that are typically and substantially different than for patients with atlantoaxial instability in the absence of basilar invagination. As highlighted in the extensive case series of Goel et al,[3] patients with basilar invagination typically undergo a trial of axial cervical traction to determine the degree to which odontoid reduction (and thus brainstem decompression) can be achieved. Cases with nonreducible basilar invagination require anterior decompression either via standard transoral surgical techniques, or more common in recent years, via endoscopic surgical techniques. Further, given the pervasive craniocervical congenital abnormalities that promote basilar invagination, posterior foramen magnum decompression and fusion extending from the occiput to the cervical spine is the mainstay of treatment,[1] either as a standalone treatment in reducible forms of basilar invagination or following anterior decompression for nonreducible forms.

Despite the confusion in terminology present in the article, this should not detract from an otherwise excellent review of specific aspects of the surgical management of atlantoaxial instability in the setting of Down syndrome. As Siemionow and Chou nicely delineate, the decision to extend fusion to the occiput should be made on a case-by-case basis. Factors such as the degree of craniocervical deformity reduction, the presence of multiplanar deformity, the existence of abnor-

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malities of occipitoatlantal joint architecture (which are particularly pertinent in the Down syndrome population), and anatomic considerations that preclude placement of C1 instrumentation are all reasons to extend fusion to the occiput. As Siemionow and Chou state, extension of fusion to the occiput is well recognized to be associated with increased operative complications and higher rates of pseudarthrosis. In addition, occipitoatlantal fusion results in substantial restriction in flexion and extension and thus has significant long-term functional range of motion consequences. The individualized case-by-case decision making advocated by Siemionow and Chou is critical for optimal patient outcome in this subset of patients with complex craniovertebral abnormalities.

References

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