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Pictorial Review

Abnormalities of the craniovertebral junction in the paediatric population: a novel biomechanical approach

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The craniovertebral junction (CVJ) is the bony transition between the cranium and cervical spine. It is a biomechanically complex articulation comprising the occipital condyles (Oc) the atlas (C1) and axis (C2). Pathologies affecting the CVJ in children are myriad with clinical features resulting from biomechanical instability, deformity, or neuraxial compression. Establishing the natural history and clinical burden of a condition is challenging in infants and young children, often complicated by co-existing neuromuscular and cognitive impairment. This makes investigation and treatment planning difficult. Each disease entity has a predilection for a particular biomechanical abnormality. Investigation using dynamic imaging is most appropriate in instability, computed tomography examination in abnormalities of deformity and magnetic resonance imaging examination in neuraxial compression. Treatment comprises reduction and immobilisation of instability, re-alignment of deformity, or decompression of the neuraxis. We present a review of disease entities affecting the CVJ in children categorised according to a simple mechanistic approach to aid investigation and treatment planning.

Introduction

The craniovertebral junction (CVJ) is the bony transition between the cranium and cervical spine. It is a biomechanically complex articulation comprising the occipital condyles (Oc) the atlas (C1) and axis (C2). Each component contributes distinct mechanical properties. Intervening ligaments provide stability while allowing the Oc-C1–C2 articulation to be the most mobile part of the cervical spine[1] without neural compromise.[2]

Disorders of the CVJ are typically classified by phenotype[3,4] and involve distinct, complex biomechanical and anatomical relationships. Ascertaining the natural history and neurological burden is challenging in infants and young children, often complicated by co-existing neuromuscular and cognitive impairment. Investigation and treatment planning can therefore be arduous.[5]

Clinical features often result from one of three biomechanical processes, namely instability, deformity, and neuraxial compression. Deformity can be subdivided into

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Table 1
Biomechanical abnormalities affecting the craniovertebral junction.

Condition	Instability	Deformity	Neuraxial compression
Down's syndrome	+++	-	+
Mucopolysaccaridoses	+++	-	++
SEDC	+++	-	+
Juvenile idiopathic arthritis	+++	++	+
Tuberculous infection	+++	++	-
Griesel syndrome	-	+++	-
Larsen syndrome	-	+++	-
Klippel–Feil syndrome	+	+++	+
Goldenhar syndrome	-	+++	-
Osteogenesis imperfecta	+	-	+++
Achondroplasia	+	-	+++
Chiari malformation type 1	-	-	+++
Atlas hypoplasia	-	-	+++

+++ Most common, ++ common, + uncommon, - rare/never.
SEDC, spondyloepiphyseal dysplasia congenital.

rotational and sagittal deformities of anatomy, and segmentation anomalies of fusion and form. Each condition has a predilection for one or more of these processes, summarised in Table 1. Treatment comprises reduction and immobilisation of instability, re-alignment of deformity, or decompression of the neuraxis.[5] Here, we present a review of disease entities affecting the CVJ in children, categorised according to a simple mechanistic approach to aid investigation and treatment.

Biomechanics of the CVJ

Movement at Oc-C1 is reliant predominantly on bony elements. Flexion/extension is limited to 23.5–24° by impingement of the dens on foramen magnum. Axial movement of 2.7–7.2° is limited by the Oc-C1 articulation and ligaments.[6,7]

The primary movement at C1–C2 is axial rotation between 23.3–38.9°[6,7] and limited by the C1–C2 articulation, the transverse ligament, contralateral alar ligament, and the capsular ligaments.[8,9] Flexion/extension at C1–C2 ranges from 10.1–22.4° [2,6,7,9]; flexion is limited by the transverse ligament, and extension by the tectorial membrane and C1–C2 articulation[2,10–12] (Fig 1).

Development of the CVJ

The 4th occipital sclerotome (proatlas) is key to the development of the CVJ[13–15] (Fig 2). Several ossification centres are present during C1 development.[16,17] The lateral masses should be present at birth and the axial ring complete at 3 years.[18] The dens is separated from the body by a cartilaginous band; a vestigial intervertebral disc named the neural central synchondrosis.[18,19] This is present up until the age of 8 years, after which the dens and body fuse.[14] Developmental abnormalities of the CVJ are associated

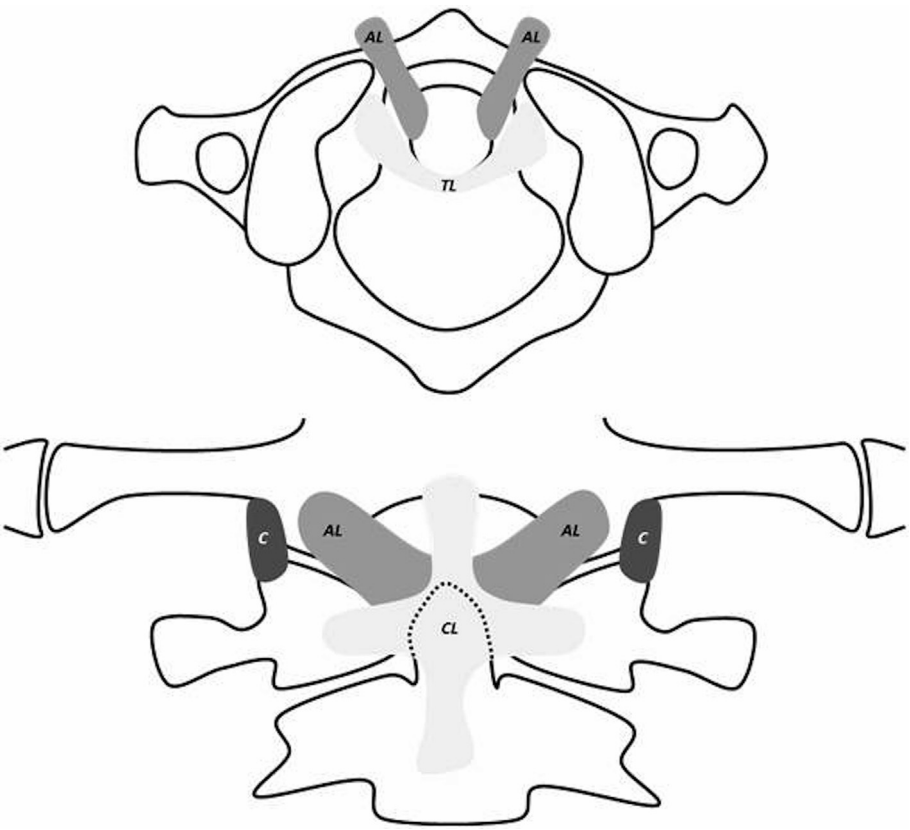

Figure 1 Anatomy of the craniovertebral junction. The tectorial membrane (not shown) is a continuation of the posterior longitudinal ligament uniting the posterior dens with the clivus. AL, alar ligaments, TL, transverse ligament, C, capsule of the atlanto-occipital joint, CL, cruciate ligament, note TL is part of CL.

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