General considerations about atlantoaxial dislocation management

Atlantoaxial dislocation (AAD) is an uncommon entity due to a loss of stability in the upper cervical spine, between atlas (C1) and axis (C2). In some cases, it can lead to sagittal deformity and severe neurological deficits when not treated properly.

Atlanto-occipital (AO) and atlantoaxial (AA) joints allow complex movements of the craniocervical junction and stabilize the head. Almost 50% of cervical spine flexion and extension movements occur at AO joint, while rotation is provided by the articulation of the dens (C2) with C1 and transverse ligament. The anatomical relationship among these structures is based in osseous elements (occipital condyles and lateral masses of C1 and C2), C1-C2 facet joints (oriented in the axial plane), and ligaments (the most important are the transverse ligament and the alar ligaments), with no intervertebral discs participating in stabilization and load sharing.

There are several etiologies related to AAD, but commonly, this pathology is multifactorial. Among the congenital causes of AAD, Down syndrome has been widely studied. In this population, ligamentous laxity and osseous abnormalities result in an increased incidence of AAD, up to 20%. Different skeletal dysplasias have been also related to increased incidence of AAD, including mucopolysaccharidosis Type IV (Morquio syndrome), Goldenhar syndrome, and spondyloepiphyseal dysplasia. The occipitalization of the atlas, C2–C3 fusion, and asymmetrical occiput-C3 facet joints are congenital osseous abnormalities also frequently associated with AAD. Traumatic AAD, with no other risk factors predisposing to it, is extremely rare and is normally due to a disruption of the transverse ligament secondary to forced displacement of the neck. In these cases, the C1 anterior arch may move completely superior and posterior after losing its articulation with the odontoid process (C2). Some Type II odontoid fractures may also be associated with AAD. Chronic rheumatoid arthritis is commonly associated with AAD, particularly in adult population, but in the last years, the incidence has decreased because of the advances in medical treatment, including immunoregulatory drugs. In these patients, chronic synovitis results in bony erosion and ligamentous laxity, with secondary instability and AAD, normally anterior.

Clinically, AAD may present with a high variety of signs and symptoms. Some possible clinical presentations include neck movement restriction and pain, weakness (even quadriplegia), numbness, sphincter disturbances, lower cranial nerve palsy, respiratory distress and failure, pyramidal signs, vertebral artery dissection, and even death. It is also important to consider the effects of upper cervical spine instability in the subaxial levels. Some patients with AAD can present severe subaxial hyperlordosis to compensate the diminished lordosis in the occipital-C1–C2 segment (swan-neck deformity). Moreover, for some authors, the pathogenesis of Chiari malformation is primarily related to atlantoaxial instability. In the differential diagnosis, we must consider torticollis, atlantoaxial rotatory fixation, and odontoid and odontoid fractures without AAD.

Several methods are applied to establish the diagnosis of AAD, without a clear and accepted consensus. The atlantodental interval (ADI) is the distance between the posterior aspect of the C1 arch and the anterior dens (C2) and can be measured in lateral cervical X-rays in neutral, flexion, and extension. The ADI is normally constant during head movements, and AAD is defined generally when ADI is exceeding 3 mm in adults and 5 mm in children. Most patients with clinical repercussion present anterior dislocations of C1, which increase ADI and decrease the spinal canal diameter, compromising the spinal cord. The patients at higher risk of paralysis where those with a space available for the spinal cord (SASC, the distance between the posterior odontoid process and the anterior aspect of the posterior arch of C1) <14 mm in dynamic X-rays. Of course, magnetic resonance imaging (MRI) and computed tomography scan are mandatory to complete the diagnosis and determine the need or not for treatment.

Our aim when we consider treatment of AAD must be stabilization of the craniocervical junction, correcting at the same time the sagittal alignment to prevent future subaxial events, and the decompression of the affected neural structures. Different strategies have been proposed, from nonoperative treatment (traction and orthotic immobilization) to different surgical approaches. Surgical treatment for patients with symptomatic AAD is accepted for the vast majority of spine surgeons, but there is still a debate on which is the best management

in asymptomatic cases. For adults with asymptomatic AAD, surgery can be considered if ADI is >5 mm. In children, we will indicate surgical fusion if one or more of the following conditions exist: neurological compromise, ADI persistently >4 mm with deformity present for >3 months, or recurrence after external reduction and 6 weeks of immobilization. In cases of AAD associated with rheumatoid arthritis, we must indicate surgery, even in asymptomatic patients, if one of the following is encountered: neck pain in a patient with radiographic instability not responding to nonnarcotic pain drugs, spinal canal stenosis with compromise of the spinal cord seen radiographically (SASC <14 mm), cervicomedullary angle <135°, or odontoid migration ≤5 mm rostral to McGregor line. Is recommended to follow-up patients with Down syndrome and those with os odontoideum performing annual lateral and dynamic X-rays, to identify changes in their deformity which will lead us to suspect, considering also clinical aspects, a worsening in spinal cord compression. We must confirm those findings performing an MRI exam.

The first measure that we have to apply when treating an AAD is the correction or reduction of the dislocation using traction. A reducible AAD should be operated through a posterior approach alone (C1 lateral mass screw – C2 pedicle screw fixation with or without extension to the occiput or to subaxial levels, C1–C2 transarticular screw fixation, C1 lateral mass screw – C2 laminar screw fixation), while in a nonreducible AAD causing spinal cord compression, we must perform an anterior decompression of the odontoid process (open transoral approach, endoscopic transoral, endoscopic transnasal, or endoscopic retropharyngeal approaches) combined, most of times, with a posterior fusion procedure. Nowadays, these procedures can be performed using the support of intraoperative neurophysiological monitoring and neuronavigation and/or robotic techniques, when available. Different classification systems have been described in the literature (Greenberg, Fielding, Wang, among others), but there is still no consensus on determining the best method to decide the best treatment options and the preferred surgical technique.

In conclusion, AAD is a challenging, rare, and sometimes fatal condition that needs an especial consideration among the spine pathologies. Getting the maximal agreement about the best treatment modality and surgical technique for each patient population must be the target in the years to come.

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