Atlanto-axial subluxation in Down syndrome

General information

Not all cases of Atlanto-axial subluxation are unstable (an unstable spine, by definition, needs treatment).

The incidence of AAS in Down syndrome (DS) is 20%, [1] but only 1–2% of DS patients have symptomatic AAS. [2] AAS in DS appears to be due to laxity of the transverse atlantal ligament (TAL). This laxity may decrease with age as the TAL stiffens.

Management

Controversial. There have been position statements [3] and rebuttals. [4], [5].

Recommendations (modified [6]); ADI = atlantodental interval, PADI = posterior atlantodental interval:

1. children who have been screened and do not have AAS: no further screening after age 10 years (since AAS does not develop later; the cutoff age is controversial)

2. os odontoideum: surgical fusion

3. symptomatic AAS

a) symptoms may include:gait difficulties,neck pain,limited neck motion,torticollis,clumsiness, sensory deficits, and other symptoms of myelopathy

b) forADI>4.5mm or PADI <14 mm or spinal cord damage on cervical MRI:surgical fusion

4. asymptomatic AAS saw on lateral C-spine X-ray:

a) forADI ≤4.5 mm andPADI ≥14 mm:no need for further testing

b) forADI>4.5mmorPADI<14mm:cervical MRI

● if the MRI shows spinal cord damage: surgical fusion

● if MRI shows no spinal cord damage: surgical fusion is optional. If fusion is not done, prohibit high-risk activities and restudy in 1 year

Case reports

A case of Down syndrome complicated by congenital atlanto-occipital dislocation. The patient presented with severe cervical myelopathy at 13 years of age after a 10-year follow-up. Radiography and computed tomography revealed os odontoideum protruding into the foramen magnum and congenital anterior atlanto-occipital dislocation. Additionally, a bifurcated internal occipital crest with a thinned central portion of the occipital bone was noted. Magnetic resonance imaging revealed

kyphotic alignment of the spinal cord with severe compression at the foramen magnum level. As the neurological impairment was partially improved by halo vest immobilization, we performed in situ O-C2 fusion with an iliac autograft and decompression of the foramen magnum and posterior arch of C1. An improvement was observed immediately after surgery. Two years after surgery, radiography and computed tomography showed solid O-C2 segment fusion. The accumulation of similar cases is essential for determining the prognosis or optimal treatment for this rare congenital condition [7].

1)
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3)
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5)
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6)
Brockmeyer D. Down syndrome and craniovertebral instability. Topic review and treatment recommendations. Pediatr Neurosurg. 1999; 31:71–77
7)
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