POSNA

The Core Curriculum

Atlanto-axial subluxation

Objectives

1. Discuss the spectrum of severity of atlanto-axial subluxation in children
2. Define Grisel's syndrome, and discuss the pathophysiology of this condition
3. Describe a treatment approach to non-traumatic atlantoaxial subluxation in children
4. Discuss imaging of atlantoaxial subluxation in children

Discussion points

1. Why does upper respiratory infection or surgery often precede atlantoaxial subluxation?
2. What is the relationship of duration of symptoms to effectiveness of treatment?
3. Does CT or MRI imaging add any useful information to that obtained by clinical exam?

Discussion

Atlanto-axial subluxation in children remains incompletely understood despite its relatively frequent appearance. It is sometimes called rotary subluxation or atlanto-axial rotary subluxation. Fielding and Hawkins described four types in 1977, almost all cases seen clinically are types I or II, in which the displacement is not severe. It often follows an upper respiratory infection, or surgery, in which case it is known as Grisel's syndrome. It has been postulated that pharyngovertebral veins can provide an avenue for the spread of inflammation from the pharynx to the ligaments of the upper cervical spine. Meniscal like synovial folds have also been demonstrated in the upper cervical spine of children, but not in adults; and these could also contribute to fixed subluxation. Similar changes have been reported in the midcervical spine.

Clinically, the child presents with a head tilt much like a child with congenital torticollis. However, the spasm of the sternocleidomastoid is on the long side in the presence of subluxation, in an attempt to right the neck; instead of the short side in congenital torticollis.

Plain radiography is generally of little help; dynamic CT scanning is often recommended. However, Villas noted that normal children could produce identical CT findings with neck rotation. The relationship to the odontoid to the atlas does not change when there is rotary subluxation, but the same findings can also be noted clinically.

Treatment is incremental, based on the degree of displacement and duration of symptoms. For a subluxation of a few days duration, a cervical collar may suffice. In hospital head halter traction, with muscle relaxants, is the next step; if this is unsuccessful, halo traction or surgical stabilization may be necessary to prevent re-subluxation. Results of treatment are generally quite good.

Several cases of traumatic subluxation have been reported accompanying a clavicle fracture, which presumably allows rotation past physiologic limits.

Not surprisingly, generalized conditions such as Downs syndrome and Marfan's syndrome which are accompanied by ligamentous laxity have been associated with rotary subluxation.

References

1. Fielding JW, Hawkins RJ. Atlanto-axial rotary fixation. (Fixed rotary subluxation of teh atlanto-axial joint. J Bone Joint Surg(Am) 1977;59:37.

2. Herzka A, Sponseller PD, Pyeritz RE. Atlantoaxial rotatory subluxation in patients with Marfan syndrome. A report of three cases. Spine 2000;25(4):524-6.

3. Hoffman B, Kaar G. Traction and television for reduction of unilateral childhood rotatory atlanto-axial subluxation. British Journal of Neurosurgery 1999;13(1):71-2.

4. Kawabe N, Hirotani N, Tanaka O. Pathomechanism of atlantoaxial rotary fixation in children. J Pediatr Orthop 1989;9:569.

5. Lopes DK, Li V. Midcervical postinfectious ligamentous instability: a variant of Grisel's syndrome. Pediatric Neurosurgery 1998;29(3):133-7.

6. Muniz AE, Belfer RA. Atlantoaxial rotary subluxation in children. Pediatric Emergency Care 1999;15(1):25-9.

7. Niibayashi H. Atlantoaxial rotatory dislocation. A case report. Spine 1998;23(13):1494-6.

8. Parke WW, Rothma RH, Brown MD. Thae pharyngovertebral veins: an anatomic rationale for Grisel's syndrome. J Bone Joint Surg(Am) 1984;66:568.

9. Subach BR, McLaughlin MR, Albright AL, Pollack IF. Current management of pediatric atlantoaxial rotatory subluxation. Spine 1998;23(20):2174-9.

10. Tseng SH, Cheng Y. Occiput-cervical fusion for symptomatic atlantoaxial subluxation in a 32-month-old child with Down syndrome: a case report. Spinal Cord 1998;36(7):520-2.

11. Villas C, Arriagada C, Zubieta JL. Preliminary CT study of C1-C2 rotational mobility in normal subjects. European Spine Journal 1999;8(3):223-8.