Management of Pediatric Craniovertebral Junction Disorders
Napa, August 2007

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Craniovertebral junction is inherently more unstable in children:

- Delicate musculature, supple ligaments, smaller condyles, horizontal joint orientation, presence of odontoid synchondrosis
- Larger head places fulcrum of maximal flexion/extension in infants and young children at C1

Kinematics

<table>
<thead>
<tr>
<th></th>
<th>rotation</th>
<th>flex/ext</th>
<th>lateral bending</th>
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<tbody>
<tr>
<td>O-C1</td>
<td>0-4</td>
<td>20</td>
<td>6</td>
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<tr>
<td>C1-2</td>
<td>32-47</td>
<td>20</td>
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Classification of Craniocervical Abnormalities

Acquired:
1. Trauma
2. Os Odontoideum
3. Infection: Grisel’s Syndrome

Genetic / Metabolic:
1. Skeletal Dysplasias
2. Down Syndrome

Congenital
1. Occipital bone: basilar invagination
2. Atlas: aplasia, occipital assimilation
3. Axis: dens dysplasia
4. Chiari

Indications for Treatment:

- Neurological Compression
  - manifest by signs and symptoms
  - imaging

- Clinical Instability determined by
  - MRI
  - Plain film criteria
  - Anterior pathology
Neurological Compression:

**Signs and Symptoms**

- head tilt, stiff neck
- scoliosis
- myelopathy
- ataxia
- nystagmus
- sleep apnea
- hoarseness, dysphagia
- neck pain
- suboccipital headache
- dizziness, syncopy
- bowel bladder dysfunction
- behavior change

**Imaging (MRI)**

- Compression
- Cord signal change
- Syrinx

Imperceptible changes in compression on imaging may alter the CSF dynamics enough to produce a new syrinx.

Imaging of clinical Instability at the Craniovertebral Junction

1. MRI: Cord signal change
   - Torn tectorial membrane or transverse ligament
2. Plain films remain the major practical consideration to determine instability at the craniovertebral junction

Clinical Instability at the Craniovertebral Junction

**Atlantodental interval (ADI)**

- Upper limit in flexion:
  - 4mm
  - 5mm (Punjabi and White)
  - 6mm (Cadaver studies <10yo)
  - >10mm: Alar and tectorial membrane secondarily damaged with O-C1 instability as well

**Dens Basion Interval (DBI):**

- 10-12.5 mm
  - Variable ossification of proatlas from 2-6yrs
- Range of motion in flex/ext:
  - <1mm in adults (Wiesel)
  - <2mm (Punjabi and White)

**Basion-axial Interval (BAI):**

- Does not extend posterior to the posterior axial line
- No flex/ext data

Clinical Instability at the Craniovertebral Junction
Clinical Instability at the Craniovertebral Junction

O-C1 Joint Interval:
5mm (Kauffman)
3mm (CT data, Pang)

Unique Consideration of Clinical Instability at the Craniovertebral Junction

3. **Anterior Pathology**: basilar invagination, platybasia

Thought to represent potential instability which is initiated or compounded by posterior decompression

Grisel’s Syndrome

- Subluxation of atlantoaxial joint following parapharyngeal inflammation: infection, post tonsillectomy.
- Torticollis

Treatment:
- Antibiotics, muscle relaxant, pain medication
- Reduction: turn to midline
  - traction (tongs)
- Immobilization - stiff collar
- Follow-up for instability (rare)

Treatment Paradigm

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<th>Stability</th>
<th>Unstable</th>
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<td>Fusion (what levels)</td>
<td>Decompression</td>
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<tr>
<td>Irreducible compression</td>
<td>Open Reduction</td>
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<table>
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<tr>
<th>Fusion</th>
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+/- open reduction
Skeletal Dysplasia:
Morquio’s, Spondyloepiphyseal Dysplasia
- O-C2 instability: ligamentous laxity
- Bone abnormalities: hypoplastic C1, dens.
- Foramen magnum, C1 stenosis.

- Normalization of soft tissue thickening, continued osseous development after surgery

Treatment: reduction and fusion; decompression (posterior). Preop complete spine MRI.

SED 9yo, 14 kg, 78cm (3yo), delayed ossification

Treatment: Halo
reduction, turn, post-op
C-C6 fusion
C1-2 Laminectomy

Morquio’s 11yo 20 kg 83 cm (5yo)

Treatment: O-C2 fusion
C1 sublaminar cables
Collar
Morquio’s 8 yo 14 kg (3yo) with mild myelopathy

Treatment: C1 laminectomy
Close f/u, likely will need fusion

Down Syndrome:
- Ligamentous laxity leads to AAD, AOD, AARS
- Hypoplastic C1 and C2, os odontoideum, basilar invagination
- 1% with symptomatic instability
- Screening radiographs: x1 3-5yr AAP, Special Olympics (do not change after 6)
- Different imaging criteria of instability for treatment

Down Syndrome:
- Recommend Fusion:
  - Preop brain image.
  - May need concurrent C1 laminectomy for posterior compression.
  - Basilar invagination may be reduced by traction, otherwise anterior decompression with fusion.
  - Fusion rate 40-95%; Graft resorption..

11yo Down Syndrome, Os Odontoideum, failed fusion at 6yo
Surgical Techniques
C2 translaminar screws

Down Syndrome 3.5 month; Craniocervical dislocation
Halo
O-C2 fusion, bilateral iliac crest

7 months (1st procedure C1 laminectomy, fusion 3.5 mo
Redo fusion 7 months rib graft, 2.0mm plate and screw fixation)
Congenital Segmentation Failure: Assimilation of the Atlas

- Isolated or in conjunction with Klippel-Feil, Chiari I.
- Excessive load on C1-2 motion segment or abnormal development of transverse ligament: AAD

- Reducible AAD: Fusion
- Irreducible AAD with basilar invagination requires anterior decompressive procedure (traction successful 1/57 pt P. Salunke et al); or open instrumented reduction (A Goel)

A Goel, J Neurosurg (Spine) 3, 2004
"Basilar Invagination" with Chiari:

- Likely not reduce with traction
  (A. Goel et al: 0/102 basilar invagination; Menezes: Clival canal angle < 127)

A Menezes, Youman's 2003

- Literature suggests addressing anterior compression via odontoidectomy along with posterior decompression and fusion.
  Boogard’s Angle > 130 (J Greenlee et al); pB-C2 > 9mm (Grabb et al)

J Greenlee et al, Neurosurg Focus 6, 1999
Grabb et al, Neurosurg 44, 1999

5yo with Klippel Fiel, basilar invagination, syrinx

Top loading: reduction
Selected basilar invagination and ventral brain stem compression with Chiari I and/or syrinx may be treated with posterior decompression, open reduction with instrumentation and fusion (L Kim et al).

Second stage odontoidectomy may be performed for persistent or recurrent symptomatic ventral compression.

Needs follow-up.

Basilar Impression (Twining’s Line >30mm)
Bone softening disorders: Osteogenesis Imperfecta

Sawin & Menezes: 25 pts (!)
- 40% reducible with traction
- Irreducible compression treated by ventral decompression and posterior fusion
- No problems with fusion
- It’s the progressive bone infolding: 80% basilar impression progressed
- Goal of treatment is palliative

L Kim, J Neurosurg (peds) 2004
**Trauma**

Injuries of the O-C2 region are the predominate form of cervical injury in children<10

- Soft tissue
- Ligamentous
  - O-C2 subluxation (AOD- longitudinal AAD)
  - transverse AAD (rare)
- Synchondrosis fracture
- AARF
- C2, occipital condyle fractures rare

**Cervical MRI Protocol for Trauma Patients (CHOP)**

Cervical MRI performed in all high risk trauma patients:

- Altered mental status or nonverbal patient with a likely mechanism of injury
- signs or symptoms referable to the cervical spine
- abnormal or equivocal radiographs
- 75 pts 95-98
Tectorial Membrane Injury

Along the Spectrum of O-C2 Injury

- Isolated muscular abnormality
- Ligamentous abnormality:
  - Apical ligament
  - Unilateral joint disruption
  - Tectorial membrane: bilateral joint disruption
  - SCI
  - Complete AO dislocation

Involvement of the Tectorial Membrane (TM) is a critical threshold for unstable ligamentous injury of the O-C2 region

- All SCI occurred with TM injury
- All TM injuries also had AO joint disruptions
- Those treated without fusions had progressive MRI changes
- Abnormal O-C2 measurements were found only with TM injury

Radiographic criteria of O-C2 instability in MRI tectorial membrane abnormalities

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(CT data)
Radiographic criteria of O-C2 instability in MRI tectorial abnormalities

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Synchondrosis Fracture:
- Synchondrosis ossify @ 7
- Most neurologically intact
- Treatment: CTO
Cervical MRI Protocol for Trauma Patients (CHOP)

Cervical MRI performed in all high risk trauma patients:

• Altered mental status or nonverbal patient with a likely mechanism of injury
• Signs or symptoms referable to the cervical spine
• Abnormal or equivocal radiographs
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Cervical MRI Protocol for Trauma Patients

Utility of MRI to “clear” the pediatric neck (<10)

• No occult compressive lesions
• No occult instability (unstable MRI ligamentous findings) in the absence of plain film abnormality with C1-2:2-3 ratio included
• Spinal cord injury (including SCIWORA) noted clinically (unpublished)