Pre-Operative Services Teaching Rounds 14 April 2011

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<table>
<thead>
<tr>
<th>Down’s Syndrome</th>
<th>Dental procedures</th>
</tr>
</thead>
<tbody>
<tr>
<td>History</td>
<td>Positioning</td>
</tr>
<tr>
<td>Genetics</td>
<td>Anesthesia</td>
</tr>
<tr>
<td>Incidence</td>
<td>Airway management</td>
</tr>
<tr>
<td>Clinical features</td>
<td></td>
</tr>
<tr>
<td>Anesthesia implications</td>
<td></td>
</tr>
</tbody>
</table>
CASE

- 29 yr old Down’s syndrome for dental rehab under GA
- Typical Down’s features
- VSD
- No previous anesthesia
DOWN’S SYNDROME - HISTORY

- John Langdon Down, a British physician, first described the syndrome in an article in 1866.
- Mongol
  - synonym used by Dr Down, referring to work of Johann Friedrich Blumenbach on ethnic classes because of facial similarity to Mongoloid class
- A letter to the Lancet in 1961 by 18 geneticists saying that *Mongolian idiocy* had "misleading connotations," and had become "an embarrassing term," and should be changed. *The Lancet* supported *Down's Syndrome*.
- WHO accepted this in 1965, as requested by Mongolian delegate
- Down’s syndrome is the accepted medical term today
The cause was not identified till 1959 when Jerome Lejeune in France discovered trisomy 21.

Found in all ethnicities

*Down’s equivalent found in animals — well described in mice and chimps*
GENETICS

Usual karyotype:
- 23 pairs of chromosomes
  - 22 paired
  - XX or XY sex chromosomes
- Down’s: additional chromosome
GENETICS

○ Trisomy 21.
  - > 90%
  - three copies of chromosome 21 in all cells
  - abnormal cell division during the development of the sperm cell or the egg cell.

○ Mosaic Down syndrome.
  - Rare
  - some cells with an extra copy of chromosome 21.
  - abnormal cell division after fertilization.

○ Translocation Down syndrome.
  - uncommon
  - part of chromosome 21 becomes attached (translocated) onto another chromosome, before or at conception.
  - two copies of chromosome 21, with additional material from chromosome 21 stuck to the translocated chromosome.

No known environmental trigger for abnormality
INCIDENCE

- Most common chromosomal disorder
- 1 of every 600–800 births
- 5500 born /yr in the US
- Advanced maternal age is implicated
  - 1-4% after age 40
  - although 80% born to younger women as majority of babies are born to younger women
- Paternal age recently implicated
- Life expectancy – 49 years (used to be 10 years)
CLINICAL FEATURES

- Mental retardation
  - IQ is below 65 (average 50)
  - Delayed fine motor skills
  - Delayed gross motor skills
  - Language delay

- Hypotonia

- Excessive flexibility
  - upper cervical spine instability produced by ligamentous laxity, skeletal anomalies, or both
  - can result in neurologic impairment, including quadriplegia
CLINICAL FEATURES

- Craniofacial abnormalities
  - Brachycephaly
  - Flattened facial features / Flat nose
  - Large Protruding tongue
  - Small chin
  - Upslanting palpebral fissures
  - Prominent epicanthic folds
  - Unusually shaped ears
CLINICAL FEATURES

- Upper airway
  - Macroglossia
  - Tonsillar hypertrophy
  - High arched palate
  - Narrow trachea for age
  - Small teeth
- Broad, short hands with a single palmar crease
- Clinodactyly of 5th finger, hypoplasia middle phalanx
- Relatively short fingers
- Short limbs
- Infants born with Down syndrome may be of average size, but typically they grow slowly and remain shorter
Trisomy 21 - Hand Features

clinodactyly
single flexion crease
short broad hands
CLINICAL FEATURES

Cardiovascular

- congenital heart disease (50%),
  - Endocardial cushion defects (AV canal) (32-40%)
  - Ventricular septal defect (30-40%)
  - Tetralogy of Fallot /ASD / PDA
  - Cyanosis/Eisenmenger’s syndrome
  - Pulmonary hypertension

- Lower risk of atherosclerosis
COMMON CARDIAC LESIONS

- Endocardial cushion defects
- Ventricular septal defect
CLINICAL FEATURES

Gastrointestinal findings:
Congenital:
  • Imperforate anus
  • Trache-oesophageal fistula
  • Duodenal atresia
  • Hirschprung’s
Acquired:
  • GERD
  • Celiac disease
  • Constipation
CLINICAL FEATURES

- Seizure disorder (2-9%)
- Psychiatric
  - Early dementia (age 40)
  - Depression
  - OCD
  - ADD/autism
- Eye lesions
  - Strabismus
  - Cataracts
  - Glaucoma
  - Refractive errors
  - Brushfield spots (small white/grey/brown spots found on iris periphery)
CLINICAL FEATURES

- Sleep apnea
- Obesity
- Thyroid
- Infertility males > females
- Tumors:
  - Leukemia
  - Testicular
  - Solid tumours are less common – increased tumor suppressor cells
- Immunocompromise
- Respiratory and ear infections
ANESTHESIA IMPLICATIONS

- Airway:
  - Difficult – anatomy:
    - Tongue
    - Small chin
    - C-spine
C-spine

Atlanto-axial instability (15%)

Laxity of transverse ligament

AADI greater than 4–5 mm in any lateral view is abnormal.

Natural progression
- can become unstable (3-24%)
- can stabilize (less common)

Atlantoaxial instability. Note the increased anterior atlantodental interval (AADI) with flexion and the corresponding decrease in the space between the posterior aspect of odontoid and the posterior arch of C1 (through which the spinal cord passes).
Atlanto-axial instability:
Greater subluxation when the neck is flexed, with normalization during extension.

But extension, particularly with “lifting” of the skull and C1 that occurs during laryngoscopy, may result in movement.
Rotation of the head may also result in C1–C2 subluxation.
Atlanto-occipital instability (9-20%)  

Powers ratio used to diagnosis occipitocervical dislocation

\[
Powers \ ratio = \frac{C-D}{A-B}
\]

**C-D:**
distance from basion to posterior arch  
**A-B:**
distance from anterior arch to opisthion

Ratio ~ 1 is normal  
if > 1.0 concern for anterior dislocation ratio  
< 1.0 raises concern for posterior atlanto-occipital dislocation  
(odontoid fractures  
ring of atlas fractures)
Fig. 1

A: Lateral radiograph in extension shows normal position of occiput on C-1 with basion directly over odontoid.

B: Flexion results in 1.2 cm forward movement of occiput along relatively flat articular surfaces of C-1.
C-SPINE

- cervical spinal stenosis
- only 1-2% will become of these 3 neck c-spine abnormalities are symptomatic.

  symptoms include
  
  - neck pain
  - torticollis
  - weakness / gait abnormality
  - quadriplegia
  - clonus
HINTS AS TO NEW SPINAL CORD ABNORMALITY

History:
Has the patient’s behavior changed (refusal to participate in usual activities)?
Ability to ambulate worsened?
Fine motor function decreased?
Any change in bladder or bowel function?
Any pain in the head or neck area?
Does the patient refuse or seem unable to turn his or her head?
Have there been any episodes of dizziness or syncope?
Physical examination:
Abnormal range of motion
Neck tenderness
Gait disturbances
Weakness
Spasticity
Abnormal reflexes or clonus
American Academy of Pediatrics recommends:

- screening once between ages 3-5 especially if they are involved in contact sports.
- lateral flexion/extension c-spine Xray
- Controversial - not endorsed by Special Olympics
- Flexion recommended first while patient is cooperative (more important view)
- Specifically review:
  - C1/C2
  - Atlanto-occipital
  - Size of neural canal
- CT/MRI more sensitive but cost is high, may need anesthesia
  - Used for abnormal Xray or symptoms

Literature review:
- no evidence-based practical guidelines
- risk of spinal cord injury during anesthesia –unknown
- very few anesthesia complications in Down’s
Intraoperatively – stabilize head for procedures e.g. For BMTs – strap head and patient to table and tilt whole table laterally.

Ethical discussion regarding c-spine fixation for minor procedure under GA
Risks/benefits and burdens
ANESTHESIA IMPLICATIONS (CONT)

Easy desaturations intra- and post-

- Airway
  - macroglossia,
  - hypotonia of pharyngeal muscles
  - enlarged T’s and A’s
- Respiratory infections
- Cardiac
- OSA
- Obesity
- ETT that is 1-2 sizes smaller (subglottic stenosis and for nasal ETT – smaller nares with midface hypoplasia)

- Postoperative stridor more common
Cardiac

- Uncorrected congenital defects beyond the scope of this talk
- History of effort tolerance
  - Cyanotic spells
  - Squatting
  - Ability to drink bottle without cyanosis/stopping/sweating
- Exam
  - Clubbing
  - Cyanosis
  - Murmurs
  - Loud P2
  - Edema – sacral/calves
- Measure oxygen saturation

- Air embolus
- SBE prophylaxis
### SBE PROPHYLAXIS (AHA 4/2007)

*NEED TO HAVE BOTH LESION AND SURGERY*

<table>
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<tr>
<th>Heart lesion</th>
<th>Surgery type</th>
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<td>- Prosthetic heart valves,</td>
<td>- all dental procedures that involve manipulation of either gingival tissue or the periapical region of teeth</td>
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<td>- A prior history of IE.</td>
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<td>- Unrepaired cyanotic congenital heart disease, including palliative shunts and conduits.</td>
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<td>- Completely repaired congenital heart defects with prosthetic material or device, during the first six months after the procedure.</td>
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<td>- Repaired congenital heart disease with residual defects</td>
<td>- perforation of the oral mucosa. (T’s A’s etc)</td>
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<td>- Cardiac valvulopathy in a transplanted heart.</td>
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• **IV access – obesity and xerodermia**
• ? exaggerated response to atropine, with tachycardia and cycloplegia.
• ? altered concentrations of neurotransmitters, such as serotonin, dopamine, norepinephrine and acetylcholine. Altered sensitivity to sedatives and narcotics.

**Labs:**
- C-spine lat flex/extend Xray
- Recent ECG and oxygen saturation if heart disease
- CBC if pale
- as per our guidelines
DENTAL PROCEDURES

- Positioning – supine
- Shared airway
  - C- spine
  - Aspiration risk
ANESTHESIA

- Endotracheal tube
  - Oral
  - Nasal
- Throat pack – wet (saline)
  - Sore throat
- Arrhythmias common
- ASC candidate – cooperation/airway
**RECOMMENDATIONS**

<table>
<thead>
<tr>
<th>C-spine Xray</th>
<th>Repeat Xray</th>
</tr>
</thead>
<tbody>
<tr>
<td>- Any age</td>
<td>- New signs and symptoms</td>
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<td>- Flexion first</td>
<td>- If first done before age 3</td>
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<tr>
<td>- Especially if surgeon involved in head and neck</td>
<td>- Prior abnormality</td>
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<td></td>
<td>- Extreme positioning in surgery</td>
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