London in March

- or Auckland?
Thanks to the organisers
Especially Murali
Choanal Atresia – The GOS approach
History of Great Ormond Street Childrens Hospital, London

Founded 1852
GOS Wards
Great Ormond Street
John Evans – father of UK Paediatric ENT

Harold Hopkins – father of flexible and rigid endoscopes - (and the zoom lens)
Retrograde endoscopic approach to choanal atresia

• Introduction
  • Embryology
  • CHARGE
  • Presentation
  • Differential Diagnosis
  • CT

• GOS Technique

• Results
Embryology

• Failure of breakdown: buccal-nasal / pharyngeal membrane

Dunham, 1998
Choanal atresia in Animals

• Llamas
  – Obligate nasal breathers
  – Incidence “common”
  – Stents
  – Tracheostomy
  – Few successes
  – Euthanasia preferred treatment
Choanal atresia in Humans

• 1 in 8,000 live births
  – Females : males  2 : 1
  – Unilateral : bilateral  2 : 1
  – Unilateral right > left
    – “90% bony, 10% membranous”
    – BUT..
  – All are mixed membranous/bony

Top Tips #1
All are mixed
Associations

• “Choanal Atresia” from Smith’s Recognizable Patterns Of Human Malformation

• CHARGE
• Treacher Collins
• Pfeiffer syndrome
• Arhinia
Original description of association

- Hall, 1979 17 patients

- Choanal atresia 100%
- Mental retardation 100
- Growth delay 100
- Male hypogenitalism 78
- Small ears 76
- Cardiac defects (heart) 71
- Micrognathia 59
- Ocular coloboma 53
- Deafness 40

- 4 of the 6 'C-H-A-R-G-E' features.
  - C oloboma
  - H eart disease
  - A tresia choanae
  - R etarded growth and development
  - G enital hypoplasia
  - E ar anomalies and/or deafness

However several features not included in the acronym (e.g. facial palsy and distinctive ear anomalies) are very specific to CHARGE and rare in other conditions.

AND

Others included in the acronym (H,R,G), are very common in CHARGE, but are also very often seen in other conditions. So they are not very helpful in distinguishing CHARGE from other conditions.
REVISED CLINICAL DIAGNOSTIC CRITERIA
(Blake et al., 1998, Davenport 2003)

3 Major characteristics or 2 Major and 3 Minor characteristics

MAJOR
Coloboma of the eye (80-90%)

Choanal atresia or stenosis (50-60%)

Cranial nerve dysfunction (40-90%)
  – Lack of smell (anosmia)
  – Facial palsy

Characteristic ear shape (frequent)
  – Short, wide ear with small lobe

Middle ear:
  – Ossicular malformations on MRI

Inner ear (Mondini defect) with deformed cochlea and vestibule

MINOR

CHARGE face
  – Square face with broad forehead, arched eyebrows, large eyes, ptosis

CHARGE hand
  – Small or unusual thumb, broad palm with 'hockey-stick' palmar crease

Orofacial
  – Cleft palate, submucous cleft palate

Congenital heart defects

Genital (hypogonadotrophic hypogonadism)

Postnatal growth deficiency

Hypotonia
Less common

- **Renal** - Hydronephrosis, vesicoureteric reflux
- **Larynx** - Laryngomalacia, laryngeal clefts
- **Esophageal** - Atresia, tracheoesophageal fistula
- **Skeletal** - Hemivertebrae, scoliosis, clinodactyly, syndactyly
- **Orofacial clefting** - Found in approximately 30-50% of patients
CHARGE Genetics  
(Vissers, et al.)

- Mutation or deletion Gene chromodomain 7 (CHD7).
- Location 2p14; 7q21; 2q33?

- Mutations in this gene have been found in more than half of all children with CHARGE tested to date.

- This confirms that CHARGE syndrome is a genetic condition caused by a new mutation in a dominant gene.

- Others: 22q11.2 deletions
  - other genes (including SEMA3E)
CHARGE work-up

- ECHO/Cardiology pre-op
- ?
- ?CT scan after
- Ephedrine vasoconstriction
- suction
- Renal ultrasound
- Ophthalmology
- Audiology

Top Tip #2
Suck out nose before CT
? Scope if no CT
Check heart before GA

All GOS patients have CHARGE workup as 39% of our choanal atresia patients have CHARGE
CHARGE: information for parents

- Prevalence: at least 1:10,000.

- Risk of recurrence is at most 1-2%.

- Risk to children of individuals with CHARGE is probably 50%.

- Autosomal dominant
Presentation – Bilateral choanal atresia

• Management:
  - Wait?
  - Airway?
  - Intubation? X
  - Tracheostomy? X
Presentation and Management
Bilateral choanal atresia

• A neonatal respiratory emergency
• “Obligate nasal breather”
• Immediate management with taped-in oral airway
• Urgent CHARGE work-up and CT scan
• Trans-nasal correction in first week of life

Top Tip #3
Do not need intubation
Presentation –
Unilateral choanal atresia

• Non-urgent presentation with unilateral nasal discharge and obstruction

• Management?
Management –
Unilateral choanal atresia

• Non urgent correction

• ? Age 1+

• When there is sufficient septum to support unilateral stent
“CT in the evaluation of choanal atresia”
Laryngoscope, Healy 1987
CT – suck out nose!
Differential diagnosis

• Neonatal rhinitis
• Masses
  — Post nasal space
    • Teratoma
  — Anterior nasal space
    • Glioma
    • *Midline nasal dermoid*
    • Meningocele
• Mid nasal and pyriform aperture stenosis

Top Tips #4

Misting after vasoconstriction
Pass bent cold suction catheter
Pyriform Aperture Stenosis
How to Approach the atresia

• Historical Approaches
  – Blind anterior
  – Palatal approach

• Current Approaches
  – Anterior endoscopy 0°
  – Posterior endoscopy 70-120°

  – Ext rhinoplasty incision
  – Sublabial incision (Koltai)
How to remove the atresia

• Technique
  – Dilatation
  – Drill
  – Microdebrider

• Recurrence
  – Laser
  – Mitomycin
Palatal Approach

• Risk to mid facial growth
Koltai PJ: *The External Rhinoplasty for Unilateral Choanal Atresia*  

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Inject sulcus with 1:100,000 epinephrine
Vasoconstrict nose with oxymetazoline
Inject nasal mucosa with epinephrine
Incision with monopolar cautery at 12 watts
Cuff of labial tissue left on gingival side of incision
Elevate tissues in subperiosteal plane

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Elevate mucosa off of one side of the septum
Leave other side attached to septal cartilage
Dislocate cartilaginous septum laterally
Elevate mucosa off nasal floor on both sides
Provides submucosal access:
  vomer
  atresia plate
Remove bone without mucosa
Mucosal incision is last step

7 cases 1987 – 1991:
1 failure

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## Which way to go?

<table>
<thead>
<tr>
<th>Anterior</th>
<th>Posterior</th>
</tr>
</thead>
<tbody>
<tr>
<td>Familiar</td>
<td>Need to learn orientation</td>
</tr>
<tr>
<td>Equipment available</td>
<td>Need 120° Scope</td>
</tr>
<tr>
<td>But ...</td>
<td>But ....</td>
</tr>
<tr>
<td>Very cramped in neonate</td>
<td>Access even in premature or syndromic infants</td>
</tr>
</tbody>
</table>
GOS - Original approach with otological drill
Original trans nasal approach with 120° telescope and drill
Traditional Dilatation:
Force is mostly longitudinal
Current technique

- 120° Telescope
- Adrenaline
- Dilators to perforate only
- Microdebrider
- Open to same size as anterior nares
- Stent
- Balloon dilatation for early restenosis
- ± Mitomycin
- KTP LASER for late restenosis
120° Telescope to view choanae
Adrenaline: the key to a bloodless field
Dilators: Use with care
Using Dilators to perforate soft centre
120° Telescope and Microdebrider
Microdebrider - what a relief!

A. Pediatric Round Bur, 2.9mm
- Length: 10cm
- Straight shaft
- Application: choanal atresia
- Operating speed: up to 5,000 RPM FWD
- 5/box

G. Choanal Atresia Bur, High-Speed, 4mm
- Length: 13cm
- Curved shaft
- Cannulated suction bur tip
- Application: removal of vomer
- Operating speed: up to 12,000 RPM FWD
- 3/box
- Developed in conjunction with Gary Josephson, MD
Stents

• Dr. Charles Stent, 1807 – 1885
• Dentist

• Improved gutta percha by adding stearine, talc and colour
Original GOS stents: columella necrosis
Redesigned GOS Stents

• Bilateral cases
  – External bridge piece
  – 6 weeks
  – 4.5 Portex for term

• Unilateral
  – Intranasal if at all
Bilateral stents after 6 weeks
Unilateral stent after 6 weeks
Restenosis
KTP laser
Mitomycin

• Sub-cytotoxic dose inhibits fibroblasts
• 2 mg/ml applied topically for 4 minutes

• Topical Mitomycin as an Adjunct to Choanal Atresia Repair -  

• Surgical Management of Choanal Atresia Improved Outcome Using Mitomycin  

• The expression of mRNA for some extracellular matrix proteins (elastase, hyaluronidase, and procollagen) was downregulated in the mitomycin test groups  
Balloon dilatation
1992-2008

new cases and procedures per year

- #/yr
- new cases

10-15/yr

50/yr
GOS Choanal atresia population

  - (108 records from 129 new cases)

  - (241 new cases, 770 procedures)

- Female 64% : Male 36%
- No syndrome : 55%
- Bilateral 51% : Unilateral 49%
- CHARGE : 20%
- Right 66% : Left 34%
- Other syndrome : 25%
Surgical Results

• Unilateral

• 53/108 patients (49%)
• procedures 1 - 8 (mean 3)
• symptomatic at last follow-up 5%

• Bilateral

• 51/108 patients (51%)
• procedures 1 - 37 (mean 5)
• symptomatic at last follow-up 14%

Kubba 2004
Review of persistent restenosis

• 15 year/ 204 patients / 623 procedures.
• Persistent restenosis rate (>six procedures) was 9.8%.

Number of surgical procedures performed per patient in a population of choanal atresia patients

Cochrane 2007
Factors associated with restenosis

- Male gender
- Bilateral disease
- Associated congenital anomalies
- Low birth weight
- Small stent size

Top Tips #5
Make a big hole
Top Tips

• Exclude neonatal rhinitis
• Airway for transfer (not intubation)
• Suction pre CT
• Check heart
• All are mixed: - drill for all
• Make a big hole!
• Preserve vomer
• Balloon if you want to dilate
eSpo 2012 Amsterdam

11th International Congress of the European Society of Pediatric Otorhinolaryngology

20 - 23 May 2012

www.espo2012.com

Pediatric Otorhinolaryngology: From experience-based to evidence-based practice
DATE FOR YOUR DIARY

Saturday 31st May – Tuesday 3rd June 2014
The Convention Centre, Dublin, Ireland

12th INTERNATIONAL CONGRESS OF THE EUROPEAN SOCIETY
OF PEDIATRIC OTORHINOLARYNGOLOGY