Nasal obstruction and chronic rhinosinusitis in children – Sommer School 2018

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Nasal obstruction in children

- congenital
  - Inflammatory/infectious
  - trauma/iatrogenic
- acquired
  - neoplasm
  - systemic
Pediatric airway – anatomy

- high larynx
  - cricoid cartilage = C4
- long epiglottis
  ⇒ lies against the soft palate
- long uvula
  ⇒ neonate obligate nasal breather

Bilateral congenital nasal obstruction

- choanal atresia/stenosis
- piriform apertura stenosis
- craniofacial malformation (Treacher–Collins, Crouzon…)
- cleft lip and palate
- nose agenesis (arhinia)
- proboscis lateralis
- congenital cyst, nasolacrimal duct cyst
- meningoencephalocele
- encephalocele
- glioma
- tumor: chordoma, hamartoma, teratoma

⇒ developmental errors of the anterior neuropore
Bilateral congenital nasal obstruction

- acute respiratory distress ⇒ emergency
- intermittent cyanosis relieved by crying
- clinical evaluation:
  - nose
    - external aspect of the nose
    - 6Fr/8Fr suction catheter via nostrils
    - laryngeal mirror test
    - stethoscope
  - oral cavity
  - other malformations
  - feeding difficulties

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Embryology – nose and palate

6th week

7th week

Choanal atresia – pathogenesis

4 theories:
- persistence of the buccopharyngeal membrane from the foregut
- abnormal persistence or location of mesoderm forming adhesions in the nasochoanal region
- abnormal persistence of the nasobuccal membrane of Hochstetter
- misdirection of neural crest cell migration

Choanal atresia

- anatomical closure of the posterior choanae
- 1:5000 – 1:8000 births
- "2:1 rule":
  - 2w:1m
  - 2unilateral:1bilateral
  - 2rights;1left
- unilateral: mostly isolated, no symptoms at birth, diagnosis later in life
- bilateral: often associated malformations (CHARGE syndrome)
Bilateral choanal atresia

- respiratory distress at birth
- paradoxical cyanosis: cyanosis relieved by crying
- diagnosis:
  - 5 or Fr catheter via nostrils: obstruction 3–3.5 cm
  - ev. nasal endoscopy
  - CT-scan = gold standard

Bilateral choanal atresia and other congenital anomalies

- other associated syndromes:
  - Treacher–Collins
  - Apert–Crouzon
  - Pfeiffer
  - mandibulofacial dysostosis
- upper airway anomalies: laryngomalacia, tracheomalacia, subglottic stenosis
Bilateral choanal atresia and other congenital anomalies

Coloboma
Heart defects
Atresia of the choanae
Retardation of growth and/or development
Genital abnormalities
Ear abnormalities

- incidence: 0.1–1.2/10’000 live births
- CHD7 gene mutation
- mostly de novo autosomal dominant mutation
- tracheotomy 10–60% of cases by multilevel upper airway obstruction

Transnasal endoscopic opening of choanae

* after Ibrahim et al., Int J Pediatr Otorhinolaryngol, 2010
Transnasal endoscopic opening of choanae

- primary success rate: 67–85%
- re-stenosis most common complication
- risk factors for re-stenosis:
  - reflux
  - age < 10 days
  - syndromal patients
- prevention of complications:
  - frequent nasal saline irrigation
  - second look procedure

Transnasal endoscopic opening of choanae

- to stent or not to stent: controversial
- most likely no difference in re-stenosis rates
- postoperative phase easier without stents
- Mitomycin C: not supported by the available evidence (! potentially carcinogenic!)
- laser: to-date no evidence of better results (! complications, fire!)
Bilateral congenital nasal obstruction

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→ developmental errors of central midface

Embryology early face – nasal cavities

FIG 5. Frontal drawings of 4- to 5-week (A) and 5- to 6-week (B) embryos illustrate the progressive displacement of the nasal sacs toward the midline as a result of medial growth of the maxillary processes. Frontal view of a 6- to 7-week embryo (C) shows the nasolacrimal groove. Closure of this groove establishes continuity between the side of the nose formed by the lateral nasal process and the cheek formed by the maxillary process. (Modified with permission from Lavinie I., Clainveille M.P., eds. Chapter 1, Surgical Anatomy of the Paranasal Sinus, China: 2005. Sinus Surgery Endoscopic and Microscopic Approaches, Figures 1-2, Thieme Medical Publishers Inc., Georg Thieme Verlag Stuttgart.)

Som PM, AJNR, 2013
Embryology early face – nasal cavities

- Pyriform apertura stenosis
  - anterior nasal stenosis
  - 1:50’000 births
  - bony overgrowth of the nasal process of the maxilla
  - pyriform apertura: narrowest portion of the nasal cavity
  - symptoms similar to bilateral choanal atresia
  - diagnosis: CT
  - milder forms: conservative therapy, otherwise surgery
  - association with megamaxillary incisor, holoprosencephaly

Som PM, AJNR, 2013
Pyriform apertura stenosis

- translabial approach
- drilling of the hypertrophic bone
- stenting 2 to 6 weeks
Cleft lift and palate

- 1:800 births
- 85% isolated
- >200 syndromes associated with cleft lip and palate
- significant upper airway obstruction at birth mostly with associated malformations /syndrome
- Pierre Robin sequence:
  - retrognathia
  - glossoptosis
  - cleft palate
  - + other malformations = syndrome

Craniofacial malformations

- Crouzon
- Treacher–Collins

complex malformations oft associated with cleft palate/choanal atresia ! multilevel upper airway obstruction oft tracheotomie
Congenital nasal tumors

- tumors:
  - teratomas
  - hamartomas
  - lymphatic malformations
  - Rhabdomyosarcoma
  - ...

- teratomas/hamartomas may arise from the nasopharynx, same symptoms as congenital nasal masses, oft associated cleft palate

Bilateral congenital nasal obstruction – summary

- diagnosis:
  - endoscopy
  - examination of the oral cavity
  - radiological evaluation before any surgery!
    - CT
    - MRI

- acute management depending on the etiology and severity of obstruction:
  - special pacifier (dummy): McGovern nipple, with an opening
  - oroendotracheal Intubation
  - tracheotomy in complex syndromal malformations
Nasal obstruction in children

- congenital
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  - neoplasm
  - systemic
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Nasal obstruction – tumors

- juvenile nasopharyngeal angiofibroma
- malignant tumors:
  - rhabdomyosarcoma
  - lymphoma
  - nasopharyngeal carcinoma
  - ...

22.08.2018
Tumors – rhabdomyosarcoma

- 3–4% all pediatric malignancies
- most common solid tumor in the pediatric population
- affects head & neck in 35–40% of cases
- bimodal age distribution: 2–4 yrs and 12–16 yrs
- 60% cases < 6 yrs
- 2 histology subtypes:
  - embryonal: more common, better prognosis
  - alveolar
- oft late diagnosis
- diagnosis: biopsies and imaging (CT/MRI), staging
- treatment modalities depending on the staging

Tumors – juvenile nasopharyngeal angiofibroma

- rare highly vascular benign neoplasm with local aggressive and invasive behavior
- almost exclusive nasopharynx
- originates at the sphenopalatine foramen in the posterolateral aspect of the nasal wall
- adolescent males
- etiology unclear
- triad: unilateral nasal obstruction, epistaxis, nasopharyngeal mass
- diagnosis: CT / MRI /angiography
- treatment: embolization before surgery
Nasal obstruction – infection / inflammation

- recurrent upper airway infections
- acute rhinosinusitis
- adenoiditis
- adenoid hypertrophy
- chronic rhinosinusitis
- allergic rhinitis

Recurrent upper respiratory tract infections (URTI)

- 8–12 viral upper airway infections per year in small children
- 5–13% of these viral infections end up with a bacterial infection (otitis media or rhinosinusitis)
- in most cases, spontaneous resolution with symptomatic therapy
- no symptom in-between episodes
Rhinitis ↔ Rhinosinusitis

Acute rhinosinusitis – viral

- sudden onset of ≥ 2 symptoms:
  - discoloured nasal discharge
  - nasal blockage / obstruction / congestion
  - cough at daytime and night-time
- selbstlimitierend
- symptomatic therapy
- symptoms free intervals if the problem is recurrent
- duration < 12 weeks

European position paper on rhinosinusitis and nasal polyps 2012, Rhinology
Acute rhinosinusitis – bacterial

- RARE!
- illness extends beyond 7–10 days without improvement or acute increase of symptoms after initial improvement of a URTI or symptoms more severe
- fever
- purulent nasal discharge (with unilateral predominance)
- purulent secretions in cavum nasi
- severe local pain (with unilateral predominance)

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European position paper on rhinosinusitis and nasal polyps 2012. Rhinology

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Acute rhinosinusitis – bacterial

- presence of at least 3 symptoms:
  - discoloured nasal discharge (with unilateral predominance)
  - purulent secretions in cavum nasi
  - severe local pain (with unilateral predominance)
  - fever
  - elevated WBC/CRP

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European position paper on rhinosinusitis and nasal polyps 2012. Rhinology
Acute bacterial rhinosinusitis – therapy

The diagnosis is mostly based on history of symptoms and their duration as well as physical findings. In most cases this is a self-limited process but, treatment with antibiotics seems to accelerate resolution. **Whether this benefit outweighs the risks associated with frequent antibiotic prescriptions remains to be clarified.** Intransal steroids might be useful adjuncts to antibiotics in the treatment of ARS and very limited evidence in older children suggests that they may be useful as a single agent in the treatment. **Ancillary therapy in the form of nasal irrigations, antihistamines, decongestants, or mucolytics have not been shown to be helpful.**
Adenoid hypertrophy

- very common pediatric ENT problem
- most frequent cause of nasal obstruction in children
- peak age: 2–7 yrs
- boys = girls
- oft family history
- more common among Afro-Americans

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Adenoid hypertrophy

- main symptoms:
  - mouth breathing
  - snoring
  - OSAS
  - runny nose
  - serous otitis media
  - "dynamic" problem, as children grow...
  - duration of symptoms
  - evolution over time

When to treat?
How to treat?
What to treat?
Adenoid hypertrophy – when to treat

- permanent nasal obstruction
- nasal discharge
- mouth breathing
- night snoring
- apneas
- serous otitis media

- duration and importance of symptoms, as well as parents/caregivers, play an important role in the treatment making process

Adenoid hypertrophy – how to treat

- wait and see
- nasal saline
- topical corticosteroids
- adenoidectomy
Adenoid hypertrophy – topical steroids

How does it work? 3 hypothesis:

- direct reduction of adenoidal size by lympholytic action of steroids on adenoids
- reduction in adenoidal and nasopharyngeal inflammation by anti-inflammatory effects of steroids
- reduction in the significance of the adenoids as a reservoir for infection

Does it really work?

Zhang L et al., Cochrane, 2010

Intranasal corticosteroids for nasal airway obstruction in children with moderate to severe adenoidal hypertrophy

Zhang L et al., Cochrane, 2010

- Cochrane review 2008, update 2010
- 6 randomized studies, 394 patients
- topical steroids vs. placebo (saline) 8–24 weeks
- outcomes measures: Nasal Obstruction Index (NOI), questionnaires, endoscopy
- 5 studies with improvements (nasal obstruction, adenoid size)
- 1 no significant improvement of symptoms after 8 weeks beclomethasone although there was a 5-fold decrease of the adenoids size
Intranasal corticosteroids for nasal airway obstruction in children with moderate to severe adenoidal hypertrophy
Zhang L et al., Cochrane, 2010

• BUT: several methodological flaws (allocation concealment unclear, no standardized reporting of results, sample size... )
• how does it work still unclear
• very few secondary effects
• remaining questions:
  • dosage?
  • duration of therapy?
  • long-term secondary effects?
  • follow-up and long-term outcomes?

Systematic review and meta-analysis of randomized controlled trials on the role of mometasone in adenoid hypertrophy in children

• 8 randomized controlled trials
• Mometasone nasal vs nasal saline
• dose and duration of therapy: 100, 200 or 400 µg p. day for 4-9 weeks
• significant improvement: nasal obstruction, snoring, total nasal symptoms, QOL

• Conclusions: some beneficial effect of mometasone spray but some limitations::
  • few randomized controlled trials
  • minimal dose and duration to obtain some beneficial effects not known
  • assessment of adenoid size not standardized (size, adenoid/choana ratio)
  • poor methodological quality of all randomized controlled trials
Adenoid hypertrophy – topical nasal steroids

- YES
- utilisation of mometason and fluticason seems to be safe in children
- duration of therapy min 2 weeks, mean 4–6 weeks
- but administration in small children with a runny nose most likely not optimal
- in case of important obstruction an operation will probably be necessary anyway
- it does not help in case of enlarged tonsils

Adenoid hypertrophy – adenoidectomy indications

- in cases of mild symptoms is a trial of steroids a good first option
- obstruction of the upper airways (permanent blocked nose, nasal discharge, mouth breathing, snoring at night)
- OSAS, BUT watch the tonsils!
- by insertion of ventilation tubes: only if adenoid symptoms
- children > 18 months old, BUT rare exceptions
- not a clear indication by “only” recurrent viral upper airway infections
Adenoidectomy – with tonsillektomy?

Table 3. Odds of Future Tonsillectomy by Tonsil Size at Initial Adenoidectomy

<table>
<thead>
<tr>
<th>Tonsil Size</th>
<th>Cases (n = 132)</th>
<th>Controls (n = 24)</th>
<th>Odds Ratio (95% Confidence Interval)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>13</td>
<td>18</td>
<td>0.8 (0.4-1.7)</td>
</tr>
<tr>
<td>1+</td>
<td>11</td>
<td>7</td>
<td>1.0 (0.6-1.5)</td>
</tr>
<tr>
<td>1+ to 2+</td>
<td>7</td>
<td>7</td>
<td>1.0 (0.4-2.0)</td>
</tr>
<tr>
<td>2x to 5+</td>
<td>19</td>
<td>34</td>
<td>0.5 (0.3-1.0)</td>
</tr>
<tr>
<td>5x to 10+</td>
<td>14</td>
<td>7</td>
<td>2.0 (0.6-6.0)</td>
</tr>
<tr>
<td>10x</td>
<td>3</td>
<td>1</td>
<td>3.0 (0.5-18.3)</td>
</tr>
</tbody>
</table>

*From the nested case-control study. P = .001 for test of homogeneity equal odds; P<.001 for test to trend of odds.

Figure 1. Kaplan-Meier estimates demonstrating odds of tonsillectomy by age at the time of initial adenoidectomy.

2462 patients
5 mths – 18 yrs

Kay DJ et al., Arch Otolaryngol Head Neck Surg, 2005
Pediatric chronic rhinosinusitis – definition

- similar definition to adults
- ≥2 symptoms > 3 months:
  - nasal blockage/obstruction/congestion
  - nasal discharge (anterior/posterior nasal drip)
  - ± facial pain/pressure
  - ± cough
  - ± relevant changes on the CT scan
- not a well studied condition

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Pediatric chronic rhinosinusitis

- different management if patients < or > than 12 years old
- management of patients between 13–18 years similar to adults
- allergy is not a significant factor in pediatric disease
- adenoid hypertrophy plays a major role in children < 12 years, and therefore is the first line treatment in case of nasal obstruction/nasal discharge, etc.
- no diagnosis of chronic rhinosinusitis in healthy children before removal of adenoids, as adenoidectomy resolves the problem in most cases

- cave: nasal polyposis!
Pediatric chronic rhinosinusitis – diagnosis

- medical history: tobacco, allergies, asthma
- clinical examination:
  - anterior rhinoscopy
  - nasal endoscopy (> 4 years) and epipharyngoscopy
- NO radiological examination in healthy children, except before surgery
- allergy tests in case of positive medical history

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Pediatric chronic rhinosinusitis – treatment

- children > 12 years: similar to adults
- children < 12 years: saline irrigation and topical steroids, if no improvement and/or moderate to severe symptoms, consider adenoidectomy
- FESS not as first line treatment in children < 12 yrs

**BUT BE ALERT**
in case of unusual evolution, persistence of symptoms without any change after adequate treatment, nasal polyposis
3 systemic diseases with unspecific ENT symptoms:

- Primary ciliary dyskinesia
- Immunodeficiency
- Cystic fibrosis

Cystic fibrosis

- most common genetic disease in Switzerland
- autosomal recessive. Mutation in the cystic fibrosis transmembrane conductance regulator (CFTR) gene on chromosome 7, which leads to production of a defective chloride channel, and thick secretions

- chronic rhinosinusitis in ~ 100% of patients over time, most with polyps
- oft no development of frontal and sphenoid sinuses
- diagnosis:
  - endoscopy
  - CT only if surgery planned
Cystic fibrosis – treatment

- conservative:
  - saline irrigation (hypertonic)
  - topical antibiotics
  - oral antibiotics
  - topical steroids
- surgery
  - endoscopic opening of the sinuses the first time
  - very high rate of recurrence, re-polypectomies

Primary ciliary dyskinesia and immunodeficiency

- often first manifestation = ENT field: not disease-specific symptoms: runny nose, serous otitis media...
- often diagnosis after adenoidectomy for recurrent URTI and otitis with same symptoms after surgery...
- a nasal polyposis infrequent, but possible, mainly by primary ciliary dyskinesia
Nasal obstruction and chronic rhinosinusitis in children – summary

- A blocked nose or a runny nose is a very common complaint during childhood, and mostly responds to conservative treatment.
- Recurrent upper airway infections as well as adenoid hypertrophy are the more common etiologies.
- Adenoidectomy is the first line treatment.
- In rare cases with no improvement over time although an adequate treatment has been conducted, one should look for a systemic disease such as cystic fibrosis, primary ciliary dyskinesia or immunodeficiency.
- Polyposis nasi in children = cystic fibrosis almost all the time!!!