

Pediatric Pathologies in Rhinology

Working Group Pediatric ENT
Swiss Society of ORL
Bettlach 29. August 2014
(Dres D. Leuba und P. Dubach)

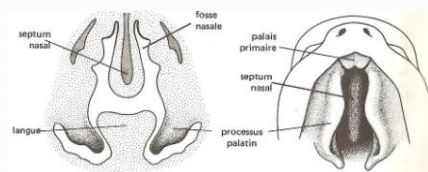


Physiology



Dr D. Leuba
(29.8.14)

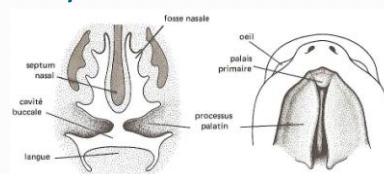
Embryo 6 ½ weeks



Frontal cut

Ventral view

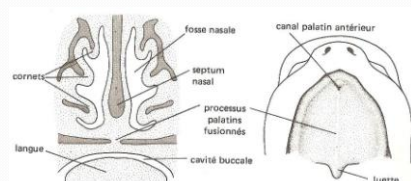
Embryo 7 ½ weeks



Coupe frontale

Vue ventrale

Fetus 10 weeks



Frontal Cut

Ventral View

The new born

- New borns are compulsory nasal breathers during the first 3 to 6 months of life
 - Nasal obstruction causes respiratory distress and feeding difficulties
- During deglutition the larynx is elevated: Epiglottis contacts and seals the velum (separation between airway and way for nutrition)

Klinik für HNO-, Kopf- und Halschirurgie

Case Presentation

- Spontaneous birth delivery at 24.12.11
- Oligohydramnion in prenatal ultrasound
- Immediate zyanosis and respiratory distress after transection of umbilical chord
- The ENT resident gets a call to evaluate the intubated child the next morning



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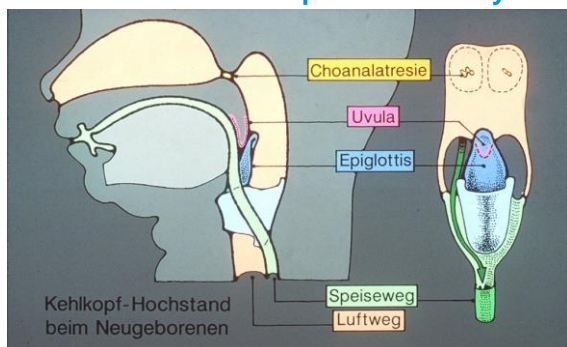
Choanal Atresia

- 1:8000 (females are affected twice often)
- 10% purely membranous
- **10% bilateral RED FLAG: Emergency situation for obstetrician, misfortunes never come singly (look for syndromal association)**
- Intubation/Mc Gown Nippel (Guedel)
- Feeding tube
- Establish the diagnosis (endoscopy)
- Evaluate for syndromes
- Surgery (but neochoanae does not grow)



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Problem with the separated air way



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CHARGE Syndrom

- Coloboma
 - Heart anomaly
 - Atresia of the choanae
 - Retardation
 - Genital Hypoplasia
 - Ear deformities
- Amotio Retinae
VSD, ASD, Fallot
Growth, Development
Caudal position or malformation of ears
- In our case: Coloboma, Patent ductus Botalli, ear deformity,..... esophageal atresia type 3b

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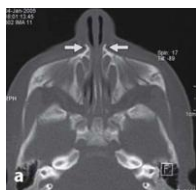
Kasuistiken

HNO 2007 55:121-127
DOI 10.1007/s00381-006-1385-8
Online publiziert: 14. März 2006
© Springer Medizin Verlag 2006

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A. Borgeas, München

Kongenitale Stenose der Apertura piriformis

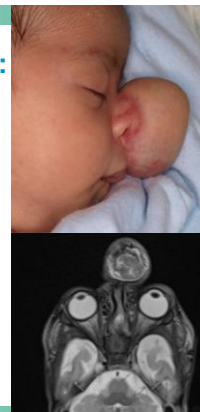
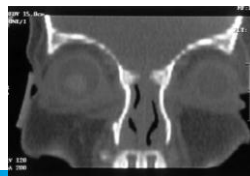
Seltene, potenziell lebensbedrohliche Ursache einer Nasenatmungsbehinderung bei Neugeborenen



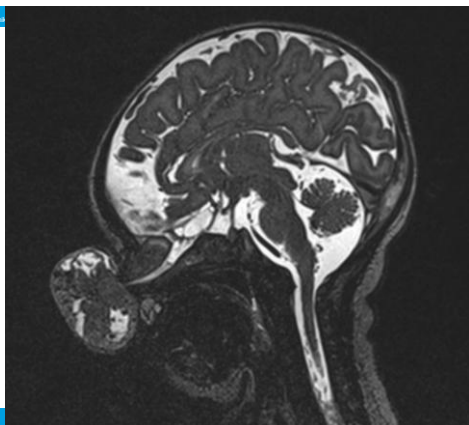
The child's paranasal sinus

- Most salient features:
 - **Maxillary sinus:** present at birth, rapid growth with second dentition, final dimensions at age 14-16, approx. 15ml
 - **Sphenoid sinus:** pneumatization at age 5-7 and slow growth until age 14-16, approx. 1-3 ml
 - **Ethmoid:** present at birth, 10-15 cells, final configuration at age 12-14, approx 2-4 ml in total
 - **Frontal sinus:** absent at birth, reaches level of orbit at age 6, pneumatization at about age 10 with large variations, completely formed at age 20, approx. 5-7 ml volume

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Problem: Wet dimpel on the nose**Midline fusion problems: (Dysraphias):****from dermal sinus to encephalocele**

Klinik

**Rhino-Sinusitis****Diagnosis on clinical grounds**

2 major criteria or 1 maj.+ 2 minor

Major criteria:

- Rhinorrhea (ant or posterior, purulent)
- Nasal obstruction
- Anosmia, Hyposmia
- Facial Dysesthesia

Minor criteria:

- Fever
- Otagia
- Halitosis
- Headache
- Cough
- Others....

Acute rhinosinusitis**Definition:**

- Infection lasting less than 30 days fulfilling the major or minor criteria
- Full recovery without persistence of complaints

Subacute chronic rhinosinusitis**Definition:**

- Diagnostic combination of symptoms during 30 to 90 days
- Asymptomatic patient after the episode

Chronic rhinosinusitis

Definition

- Inflammation of the nose and paranasal sinus longer than 90 days with respiratory signs: cough, rhinorrhoe, nasal obstruction.
- Patient remains symptomatic despite adequate treatment.

Acute Rhinosinusitis

- Clinical Workup
 - GP exam (almost indistinguishable signs of viral or bacterial infections during the **first days**)
 - ENT exam (concomitant otitis, tonsillitis)
 - Anterior rhinoscopy and 25° endoscopy
 - Painful percussion or trigger points over the sinus
 - Radiological exam only in case of expected complication or if surgery is scheduled (CT !!!)
 - Objective Signs: either pathologic CT or endoscopy of middle meatus with pus

Acute Rhinosinusitis

- E.g. 4 year old child



Microbiology

Virus: rhinovirus, parainfluenza and influenza virus, Corona virus

Bacterial: *S. pneumoniae* 30%, *Haemophilus influenzae* 20%, *Moraxella catarrhalis* 20%, others (Group B streptococci, *Staph aureus*,...)

NB: *Pseudomonas* in case of cystic fibrosis

Fungal: *Aspergillus fumigatus*, mucormycosis during bone marrow depression

Medical Treatment: Acute Rhinosinusitis

First 5 to 7 days

- Nasal decongestants
- Saline douche
- Topical steroids
- Analgetics
- Mucolytics

Longer than 5 to 7 days:
(or rapid progression of complaints)

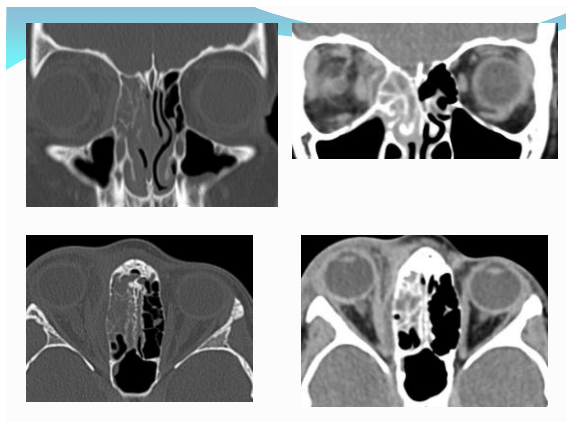
Antibiotics (10d)
NSAR

Complication of acute rhinosinusitis

- E.g. 5-year old child



Rhinosinusal endoscopy. Diagnosis and surgery
Prof. G. Terrier, 1991



Acute Rhinosinusitis

• Complications

- Orbital: More frequent in children because delicate and often dehiscent septa, open sutures and well developed vascular channels for spread of infection.
- Intracranial: 4% of hospitalized children (meningitis, epidural empyema, cerebral abscess, sinus thrombosis)

Microbiology and antimicrobial treatment of orbital and intracranial complications of sinusitis in children and their management. I Brook. Int.J. of Pediatric Otorhinolaryngology (2009)

Orbital complications

- I edema and PRE-septal cellulitis
- II orbital cellulitis
- III subperiosteal abscess
- IV intraorbital abscess
- V cavernous sinus thrombosis

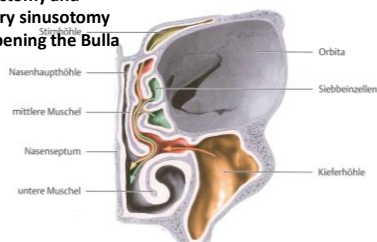
Emergency Surgery: How?

• **Endonasal:** Minimal invasive, reaches medial orbit, but has limited overview especially if bleeding occurs

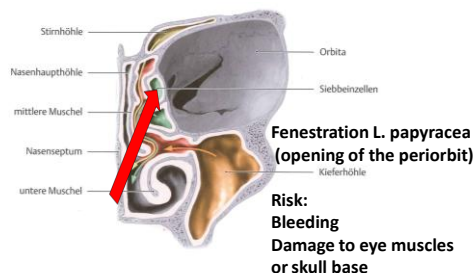
• **Transfacial:** Better overview, technical easier and less risky, reaches upper and lateral orbital pathologies (frontal sinus drainage)

Surgery: Endonasal

Uncinectomy and maxillary sinusotomy with opening the Bulla

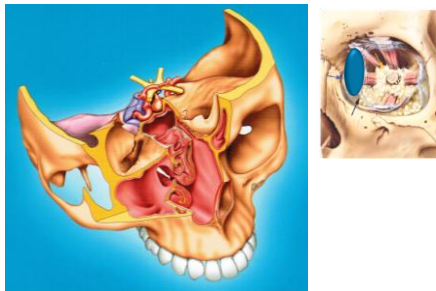


Surgery: Endonasal



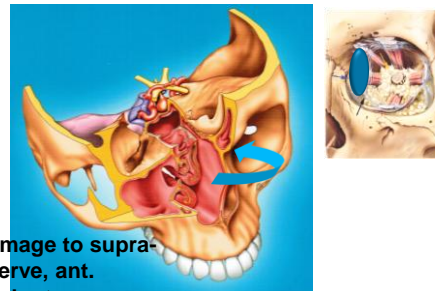
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Surgery: External Approach



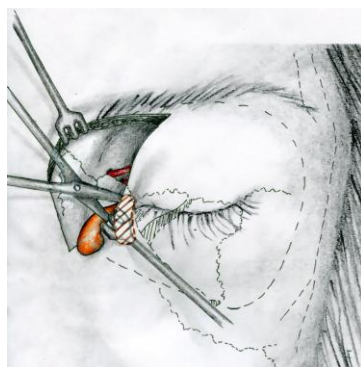
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Surgery: External Approach



Risk: Damage to supra-orbital nerve, ant. Ethmoidal artery

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Additional location of complication after age of 6

15 year old male after 5 days of acute rhinosinusitis
Rapid development of painful frontal swelling

Progressive lid swelling

Normal acuity, no double vision, GCS 14, no meningeal signs

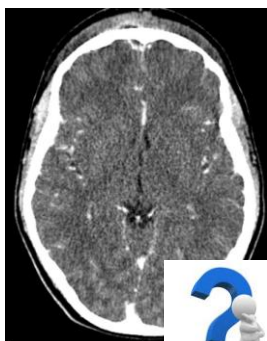


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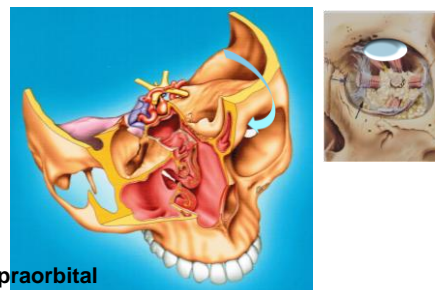


(with permission)



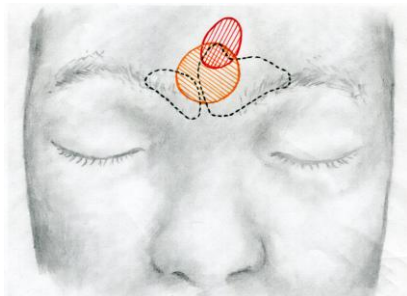
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Surgery: External Approach



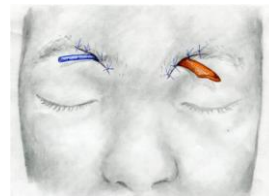
Risk: supraorbital nerve injury

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■ subperiostaler Abszess
■ epiduraler Abszess

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Drawings by the courtesy of Dr. Hergen Friedrich
AA Rhinology, Inselspital



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10 days following surgery

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Pott's Puffy Tumor:

Frontal swelling by subperiosteal abscess by definition with osteomyelitis of the frontal bone

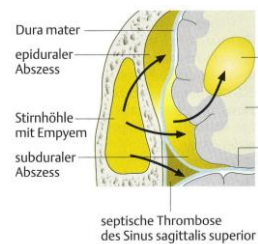
Percivall Pott (1714-1788)
- Surgeons of St. Bartholomew's Hospital London wrote in 1760
„a puffy circumscribed, indolent tumor of the scalp and a spontaneous separation of the pericranium from the skull under such tumor“

„... observed after head trauma.“

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Therapy of the Pott Puffy Tumor: **Surgical**
(up to 50% repeated)

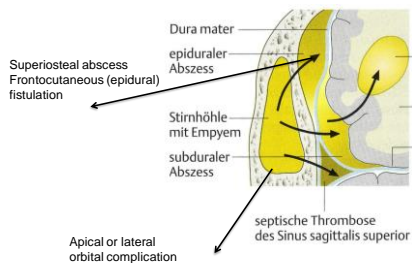
Pott Puffy tumor is prone to severe complications: **insgesamt ca. 75%**



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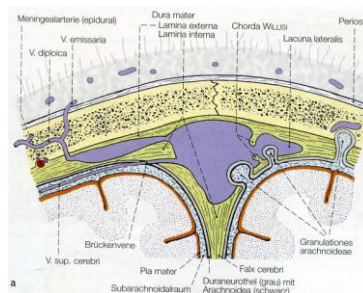
Therapy of the Pott Puffy Tumor: **Surgical**
(up to 50% repeated)

Pott Puffy tumor is prone to severe complications: **up to 75%**



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Spread of disease intracranially: Valveless diploic veins through Breschet Foramina



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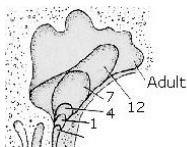
Potential risk factors:

- frontal (ethmoidal) sinusitis
- Frontale Trauma / iatrogenic after operation

Epidemiology: Adolescent males (M:F 5:1)

Etiology:

- Pneumatization frontal sinus exceeds orbita level at age 6 or later
- Maximum development of the diploic veins
- Teens are prone to „Trauma“



Main microbiological germs:

- Anaerobic mixed
- Streptococci spec.
- coagulase negative Staphylococci
- Pseudomonas aeruginosa
- etc.

In our case:
Streptococcus milleri...

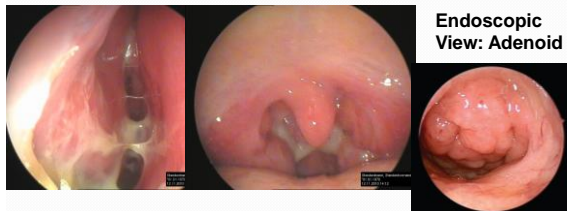
Chronic Rhinosinusitis

• Work up:

- ENT exam (adenoids)
- Anterior rhinoscopy and 25° endoscopy
- Radiological work up (CT-scan as a basis for surgery)
- Systematic work up for other pathologies (allergies, immuno-deficiency, mucoviscidosis, ciliary dyskinesia)

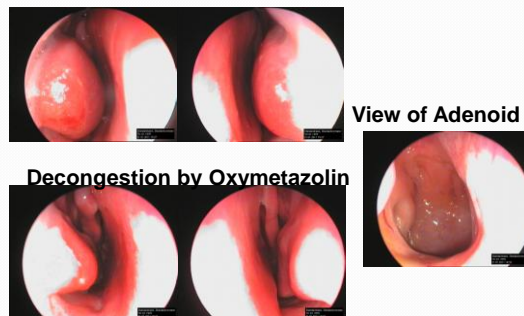


Blocked nose: Chronic adenoiditis?



5.13 Endoscopic view of adenoidal hypertrophy causing choanal obstruction. Aus Isaiing und Leunig ENT Infections, Paleri and Hill 2010

Blocked nose: Turbinate hypertrophy



Chronic Rhinosinusitis with Polyps

- E.g. Child ten year old



Rhinosinusal endoscopy. Diagnosis and surgery
Prof. G. Terrier, 1991

Chronic Rhinosinusitis

Associated factors

- Allergies (80% of children with CRS have a positive familial history of allergies)
- Otitis media and asthma
- Exposition to tobacco smoke
- Nursery
- Gastro-esophageal reflux

Chronic Rhinosinusitis

- Medical Treatment:
 - Nasal lavage with saline douche
 - Topical steroids (short time systemic steroids)
 - Systemic antibiotics (sometimes long time, only in special cases)

Chronic Rhinosinusitis

- Surgical treatment
 - Hardly ever necessary for children younger than 6 years
 - Only after failure of full medical treatment
- AIMS:
 - Better control of symptoms
 - Better quality of life
 - Prevention of complications

Cystic fibrosis / Mucoviscidosis

Symptoms

- Baby's skin tastes salty, intestinal obstruction (no meconium)
- Cough, thick sputum
- Lung infections
- Sinus infections and polyposis

Cystic fibrosis Mucoviscidosis

Diagnosis

- Genetical screening
- Sweat test
- Chest x-ray
- Lung function

Mucoviscidosis

- Infections of the respiratory tract take their origine from the nose and paranasal reservoir
- Sinu-nasal surgery prior to lung transplantation cannot prevent pulmonary contamination/colonization by sinunasal pathogens

Leung M.-K., Am. J. Rhinol. 22, 192-196, 2008

Mucoviscidosis

- Following sinunasal surgery patients show better quality of life and better access for topical medication.

FESS. A retrospective analysis of 115 children and adolescents with chronic rhinosinusitis. V.Siedek. Int.J.of Pediatric Otorhinolaryngology 73: 741-45, 2009

Primary ciliary dyskinesia Kartagener

- Symptoms
 - Respiratory infections
 - Inverted situs
 - Male infertility
 - Chronic otitis media with effusion (contraindication to grommets)

Primary ciliary dyskinesia Kartagener

- Diagnosis
 - Often delayed (50% before the age of 5 years)
 - Defects in the ciliary ultrastructures in TEM
 - Ciliary function analysis

- Work up:
 - FENO highly elevated
 - Brush cytology/biopsy of nose and bronchi for TEM and LM ciliary beat function

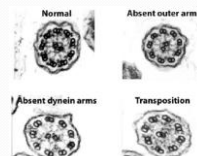


Fig. 11.5 Scanning electron micrographs of respiratory cilia (courtesy of Dr. Ann Dewar)

14 year old female with epipharyngeal mass

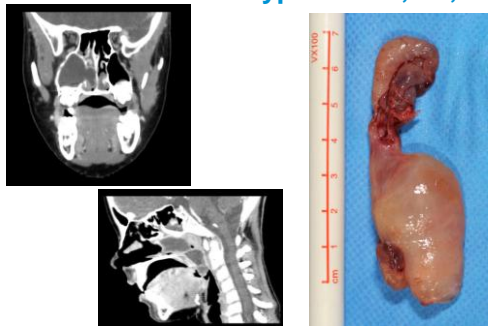


Antrochoanale Polyp COT 32, L2, D



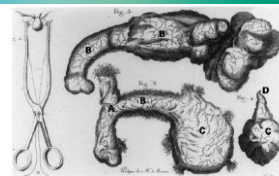
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Antrochoanale Polyp COT 32, L2, D



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Antrochoanal Polyps



- Frequency:
 - Adult 0.6%
 - Pädiatric population 7.8% bis 33%
- Antro-choanal
 - But spheno-, septoturbinar, ethmoido- und conchal origins have amply been described
- Leading symptoms: Nasal obstruction, rhinorrhea, dysphagia, dyspnea... OSAS up to cachexia

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Ad fontes...

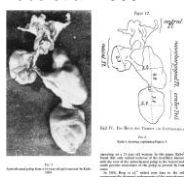
THE ORIGIN OF CHOANAL POLYPI.

BY GUSTAV KILLIAN, M.D.,

PROFESSOR OF LARYNGOLOGY AND RHINOLOGY, UNIVERSITY OF
FREIBURG IN BREISGAU.

AMONG nasal polypi the so-called choanal or benign nasopharyngeal polypi occupy a special place. This has already been emphasised by Moldenhauer and lately Gerber has confirmed it. My own experience, which extends over 22 cases, also coincides. Choanal polypi are usually unilateral.

Kubo et al. 1909



Min YG. 1995

- Origin of stalk in the maxillary sinus:
- Dorsal wall: 92.3%
- Lateral wall: 61.5%
- Floor: 38.5%
- Rarely anterior wall
- Stalk must be completely removed to prevent relapse

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Unilateral nasal mass/polyp:



- 12 year old boy
- Unilateral obstructed nasal breathing, suspected foreign body by the general practitioner
- Careful history taking: episodes of unilateral nose bleeding and ipsilateral ear pressure
- Endoscopy: Unilateral big polyp/mass
- Otoscopy: „Seromucotympanon“, tuning fork lateralized in affected ear.

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Problem for male gender Juvenile Angiofibroma

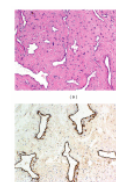
Male gender

Hallmarks: Obstruction, epistaxis...
proptosis, double vision, swellingTumor arising behind the middle turbinate
at the sphenopalatine foramenClinical and MR presentation is
characteristical, biopsy prone to massive
bleeding is not necessary!!

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Juvenile Angiofibroma

- Expansile growth into nasal- and paranasal cavities
- Into clivus and middle cranial fossa
- Into pterygopalatine and – infratemporal fossa

Histologic picture: Endothelium without
muscular layer in fibrous stroma surrounded
by pseudocapsule

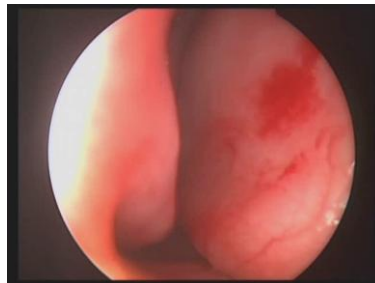
- Therapy: Primarily surgical
- Experimental: Pre-op Flutamid antiandrogen (Steroids, antiestrogens Tamoxifen not tried)
- Relapse or persistence 14-30% (radiation, hormon therapy)

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Case: The top of the iceberg

- 4 4/12 year old female
- Normal birth and uneventful development until..
- Acute right sided otalgia
- Intermittent right sided nasal obstruction
- Rapid progressive facial nerve paralysis H&B 3 bis 5
- Emergency mastoidectomy during holidays in Serbia...

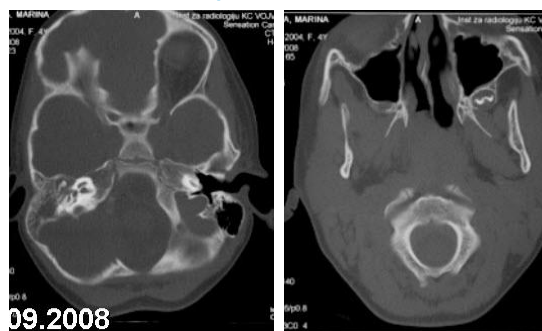
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Unilaterale Polyp....

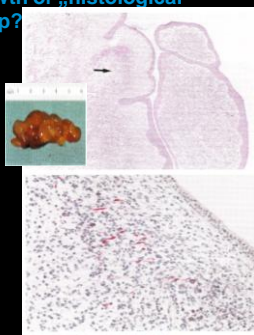
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CT of the nose and paranasal cavities...

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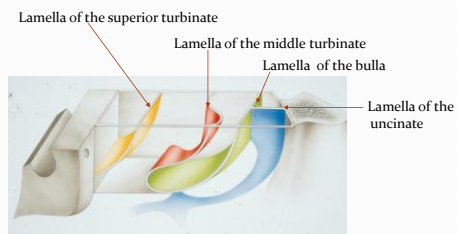
Misdiagnosis rapid regrowth of „histological proven“ antrochoanal polyp?**Rhabdomyosarcoma**

- Grapelike (botryoid) Mass
- Myxoid odemateous Stroma
- Subepithelial Layer: Cambium with:
- Spindel shaped POLYMORPHIC cells IMMUNHISTO: Vimentin, Mb, Desmin positivity

**Rhino-Sinusitis****Surgical Measurements and Technique:**

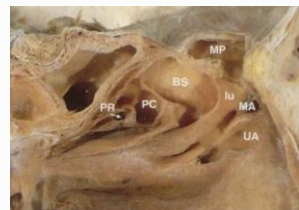
- Sinusoscopy (Maxillary sinus trepanation)
- Adenoidectomy
- Functional Endoscopic Sinus Surgery

The Anatomy of the Ethmoid



Cadaver Dissection Training

- The ethmoid in formaline fixed head



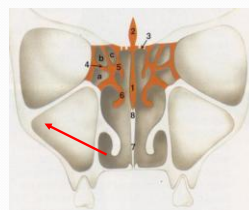
Sinuscopy: sinus trephination

Indications:

(failure of medical treatment)

- Drainage and rinsing of the sinus
- Microbiological sampling (Gram, Culture, Resistance, Fungal stains and culture)
- Evaluation of the sinus ostium and observation for the directed mucociliary transport.

Sinuscopy of the maxillary sinus



Rhinosinusal endoscopy. Diagnosis and surgery
Prof. G. Terrier, 1991

Sinusoscopy of the maxillary sinus

- E.g Child with sinus ventilation tube in place



Rhinosinusal endoscopy. Diagnosis and surgery
Prof. G. Terrier, 1991

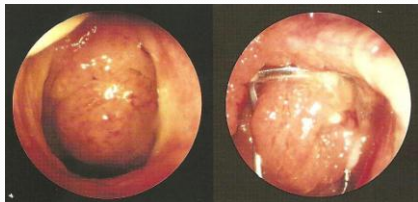
Sinuscopy

Tooth bud in the maxillary sinus



Rhinosinusal endoscopy. Diagnosis and surgery
Prof. G. Terrier, 1991

Endoscopic controlled Adenoidectomy



Rhinointral endoscopy, Diagnosis and surgery
Prof. G. Terrier, 1991

Adenoidectomy

- **Aim:** Reducing bacterial load/bacterial reservoir
- Facilitating nasal clearance from secretion
- Reducing blockage of rhinopharynx and osteomeatal complex
- Microbiological sampling by sinuscopy

Effect of adenoidectomy in children with complex problems of rhinosinusitis and associated diseases. K. Ungkanont, Int.J. of Pediatric Otorhinolaryngology (2004) 68, 447-451

FESS

- - In case of complication of rhinosinusitis
- - Persistent symptoms of chronic rhinosinusitis despite optimized medical treatment for 6 weeks.
- Persistent symptoms despite of adenoidectomy
- Following:
 - - Immuno-allergic work up
 - - Sweat test in case of polypoid chronic rhinosinusitis
 - - CT low Dose

FESS

- **Aim:**
- Optimization of natural drainage pathway
- Diminishing frequency of superinfections
- Facilitating the use of nasal lavage and application of topical medication

FESS

- Surgical technique:
 - Similar to adults but
 - Paranasal cavities underdeveloped, sutures and bone dehiscent
- Experienced surgeons
- Navigation system

Sphenoidotomy prior to CT based CAS

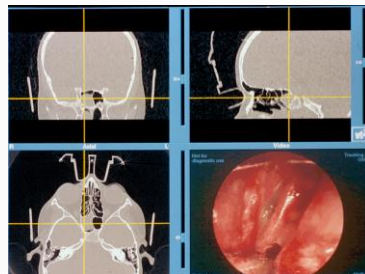


Conventional radiological localization of the sphenoid sinus

CAS systems



CT based CAS for sphenoidotomy



FESS surgical technique

- Landmarks: Middle turbinate and ethmoidal bulla
- Infiltration of middle turbinate, uncinate process and bulla.

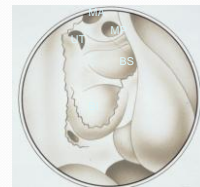


FESS surgical principles

Vertical uncinectomy

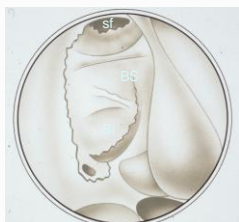


Bullectomy



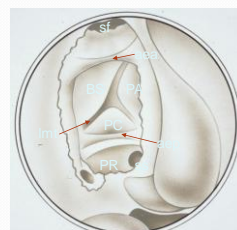
FESS technique chirurgicale

- Anterior ethmoidectomy and frontal sinusotomy



FESS surgical principles

- Posterior ethmoidectomy



Bibliography

- Clinical practice guideline: Management of sinusitis. American academy of pediatrics. Pediatrics Vol. 108 N°3, sept 2001
- Pediatric sinusitis: when do we operate? J.D.Lieser. Current opinion in Otol. & Head and Neck Surgery, 13:60-66, 2005
- FESS for chronic rhinosinusitis. (Review) The Cocherane Collaboration. 2009
- FESS. A retrospective analysis of 115 children and adolescents with chronic rhinosinusitis. V.Siedek. Int.J.of Pediatric Otorhinolaryngology 73: 741-45, 2009

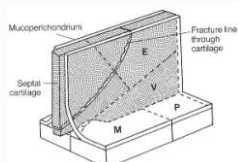
Pediatric nasal fractures

- Nasal fractures represent 30% of the pediatric fractures.
- 32% are mandibular fractures and 28 % maxillary or zygomatic fractures
- Causes:
 - 50% car accidents
 - 12% fights
 - 9% falls

Patterns and outcomes of pediatric facial fractures in the US: A survey of the National data Bank. Imahara, J.Am Coll Surg. Vol.207, n°5, nov 08, p.710-16

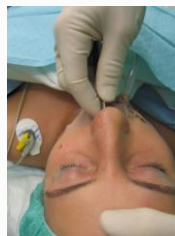
Nasal fracture

- Anatomy



Nasal fracture

- Closed Repositioning



Nasal fracture



Work up: Nasal trauma

Soft tissue coverage of denuded bone/cartilage

Red flag: Septal hematoma and superinfection (abscess): Early diagnosis and drainage

If deviation cannot be evaluated: Await reduction of swelling, **time limit for closed reduction: 7-10 days after trauma**

documentation for scheduled control and organization for repositioning if necessary

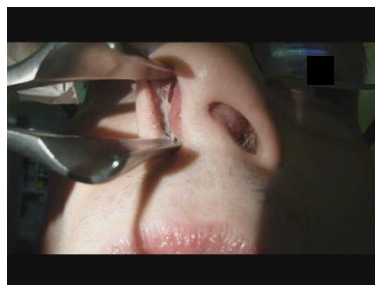
Klinik für HNO-, Kopf- und Halschirurgie

Complication of nasal trauma



Klinik für HNO-, Kopf- und Halschirurgie

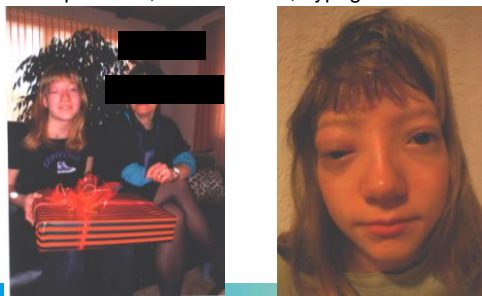
Inspection, Palpation



Klinik für HNO-, Kopf- und Halschirurgie

Mucocele, Mucopyocele

- Minimal pressure, double vision, hypoglobus

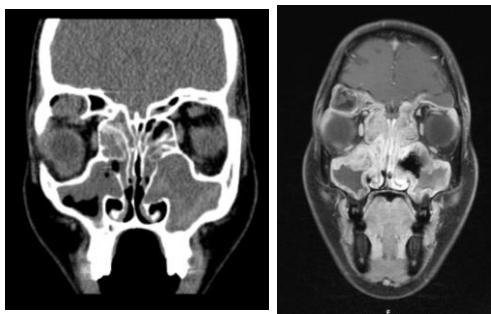


Klinik für HNO-, Kopf- und Halschirurgie

Mucocele, Mucopyocele

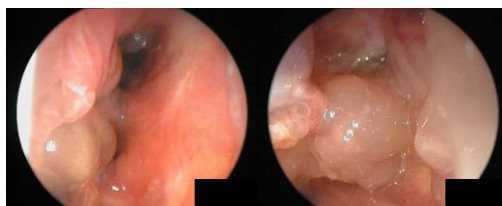
- Retention and dilatation of preformed paranasal sinus
- Etiology: post-traumatic, iatrogenic, inflammatory
- Location: ethmoidal, frontal, sphenoidal, maxillary

Klinik für HNO-, Kopf- und Halschirurgie



Klinik für HNO-, Kopf- und Halschirurgie

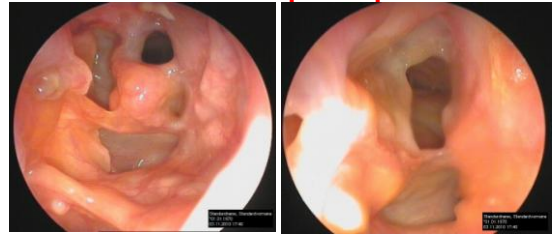
Pre-operative frontal recess



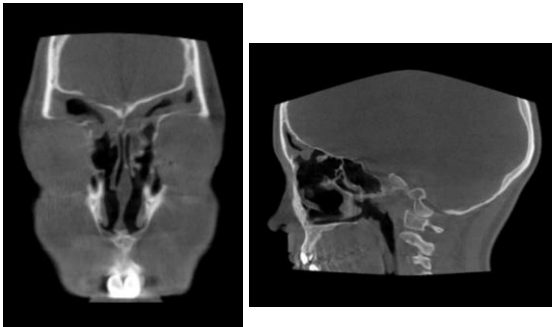
Klinik für HNO-, Kopf- und Halschirurgie

4 weeks post-op

Klinik für HNO-, Kopf- und Halschirurgie

8 weeks post-op

Klinik für HNO-, Kopf- und Halschirurgie

1 year post operative: Digital volume tomography

Klinik für HNO-, Kopf- und Halschirurgie



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your children