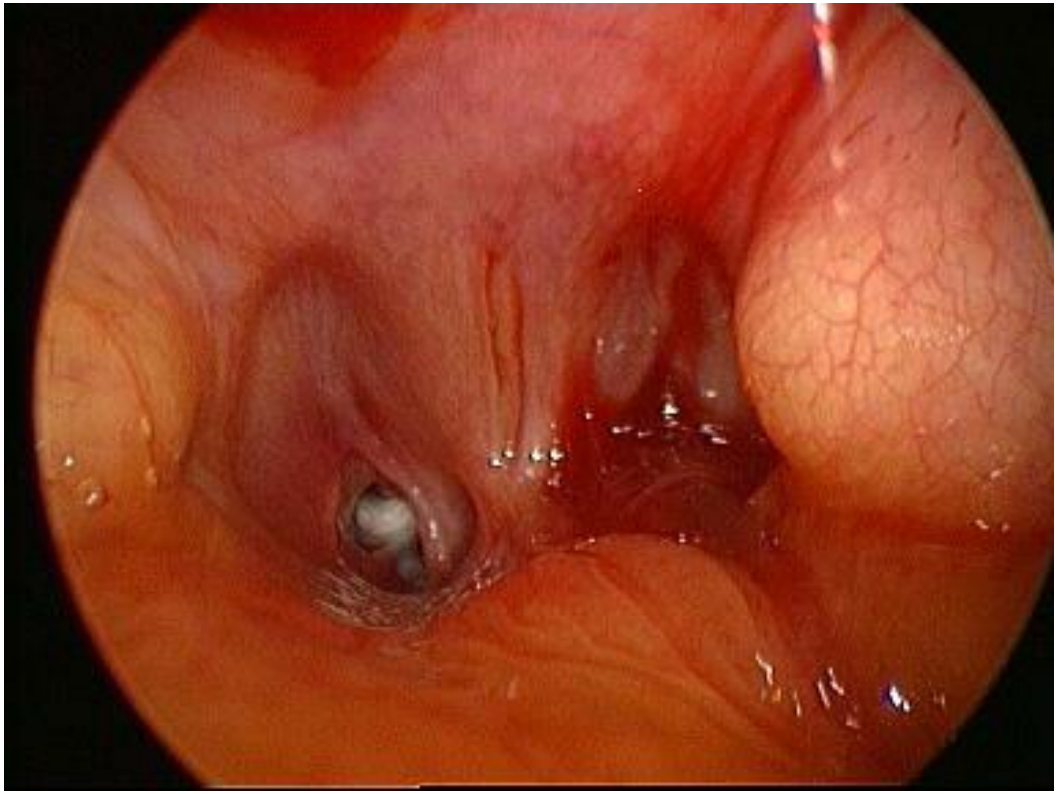
London in March - or Auckland?

12th Asia-Oceania Otolaryngology Congress Auckland 2011

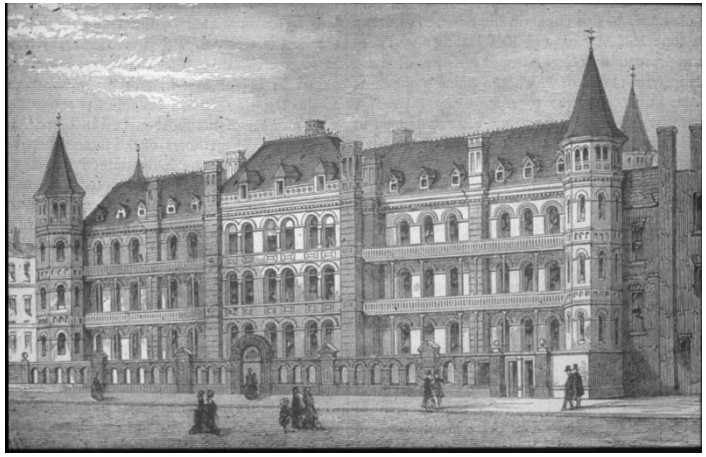

Fisheries
POACHING IS THEFT - REPORT ILLEGAL FISHING

Choanal Atresia – The GOS approach



History of Great Ormond Street Childrens Hospital, London

Founded 1852

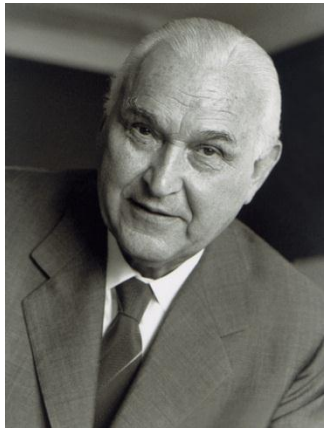


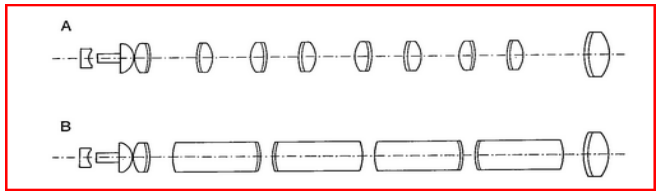
GOS Wards

Great Ormond Street

GOS ENT Department

John Evans – father of UK Paediatric ENT

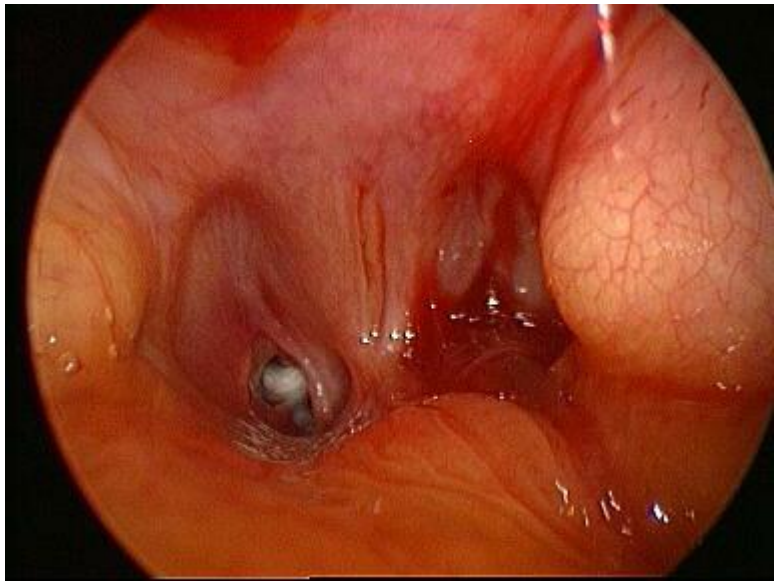




Harold Hopkins – father of flexible and rigid endoscopes - (and the zoom lens)

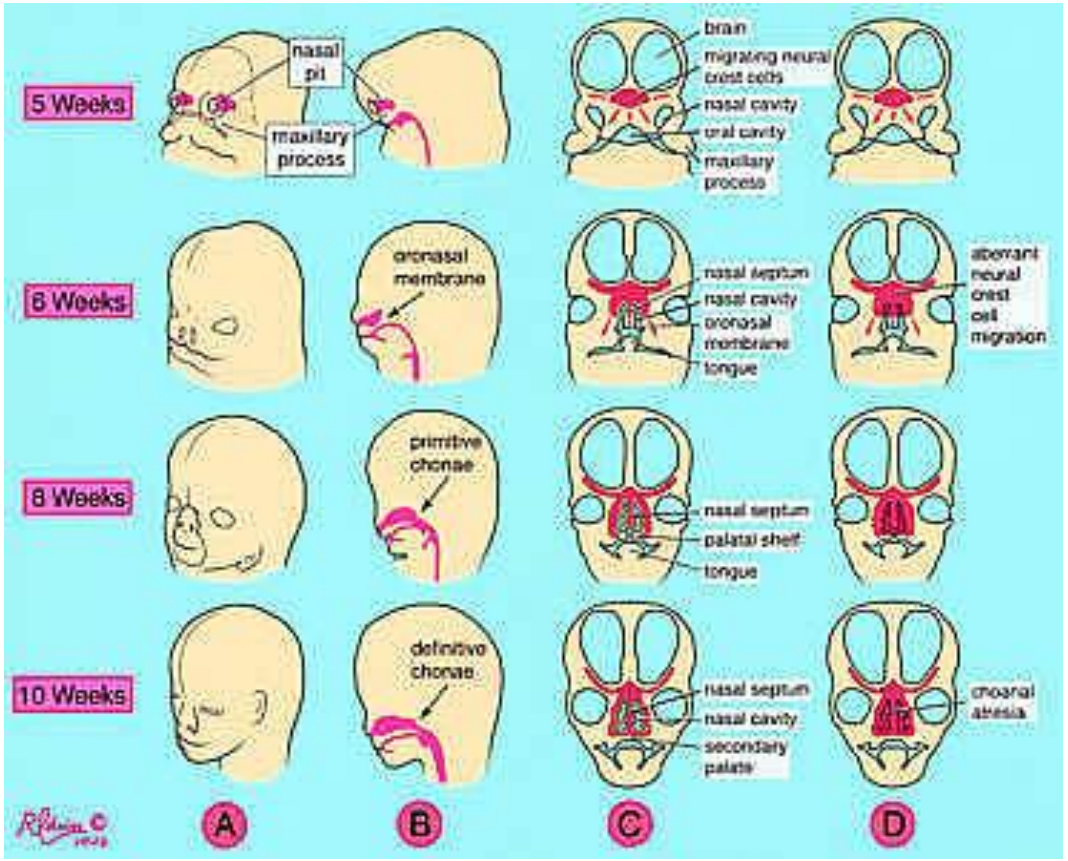
Retrograde endoscopic approach to choanal atresia

- Introduction
  - Embryology
  - CHARGE
  - Presentation
  - Differential Diagnosis
  - CT
- GOS Technique
- Results



Embryology

- Failure of breakdown: bucco-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

Original DIAGNOSTIC CRITERIA of C.H.A.R.G.E. (Pagon et al,1981)

- 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 anomolies 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 (hypogonadotropic 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?
- Tracheostomy?

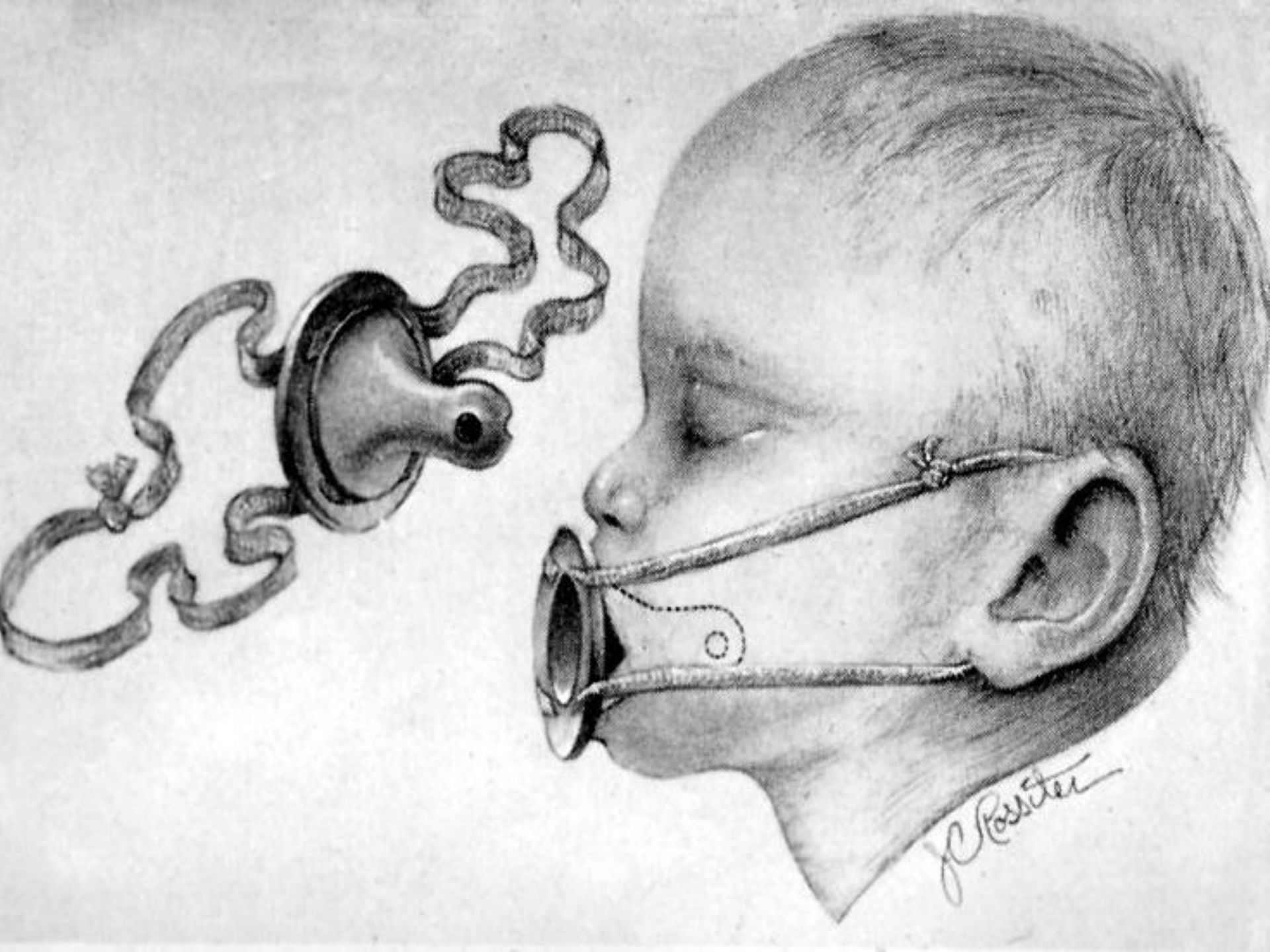
Presentation and Management
Bilateral choanal atresia

- 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

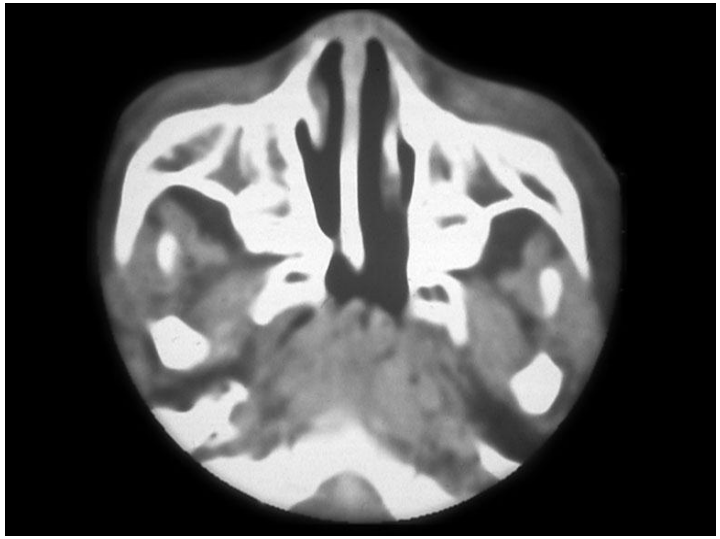
- Non-urgent presentation with unilateral nasal discharge and obstruction

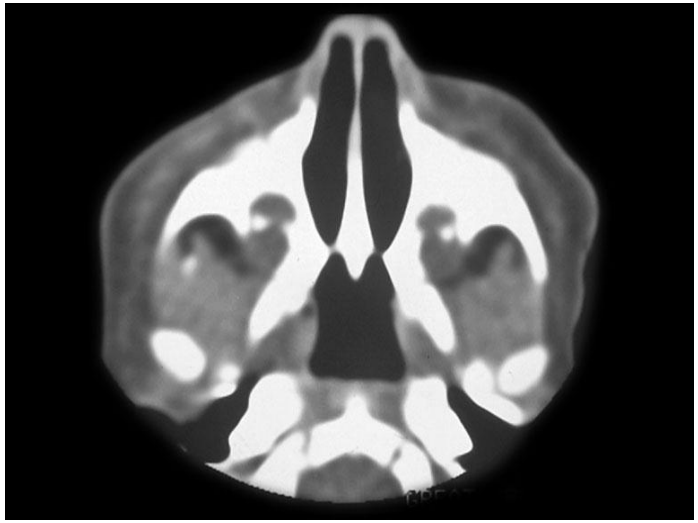
- Management?

Management – Unilateral choanal atresia

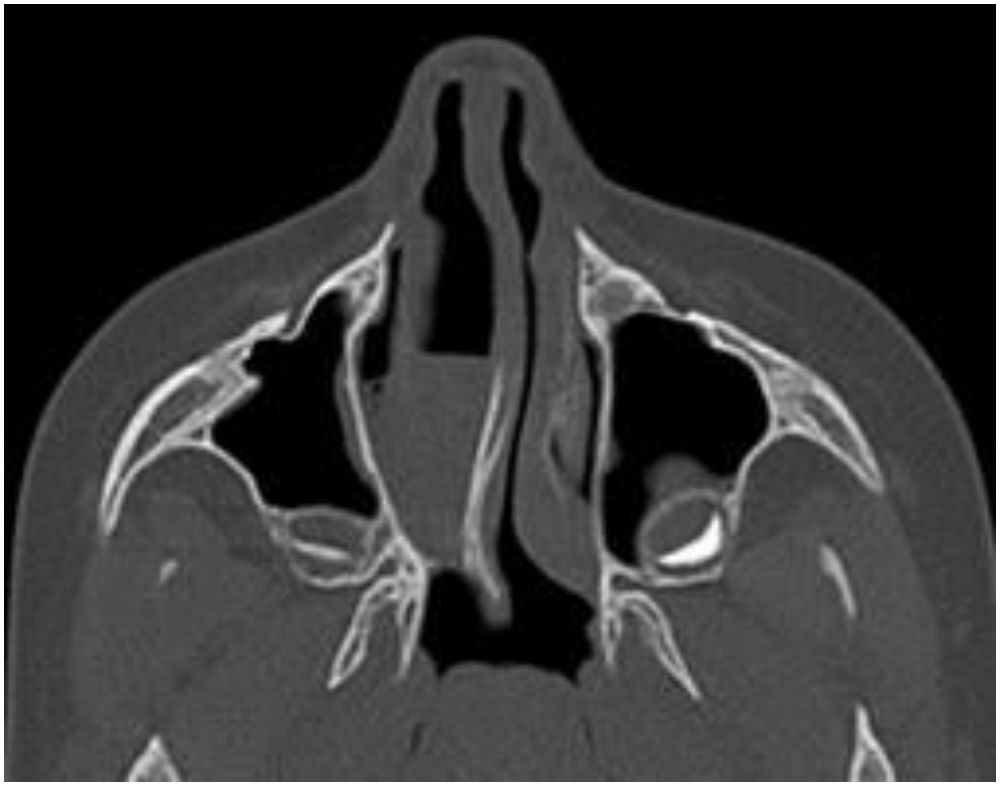
- Non urgent correction
- ? Age 1+
- When there is sufficient septum to support unilateral stent

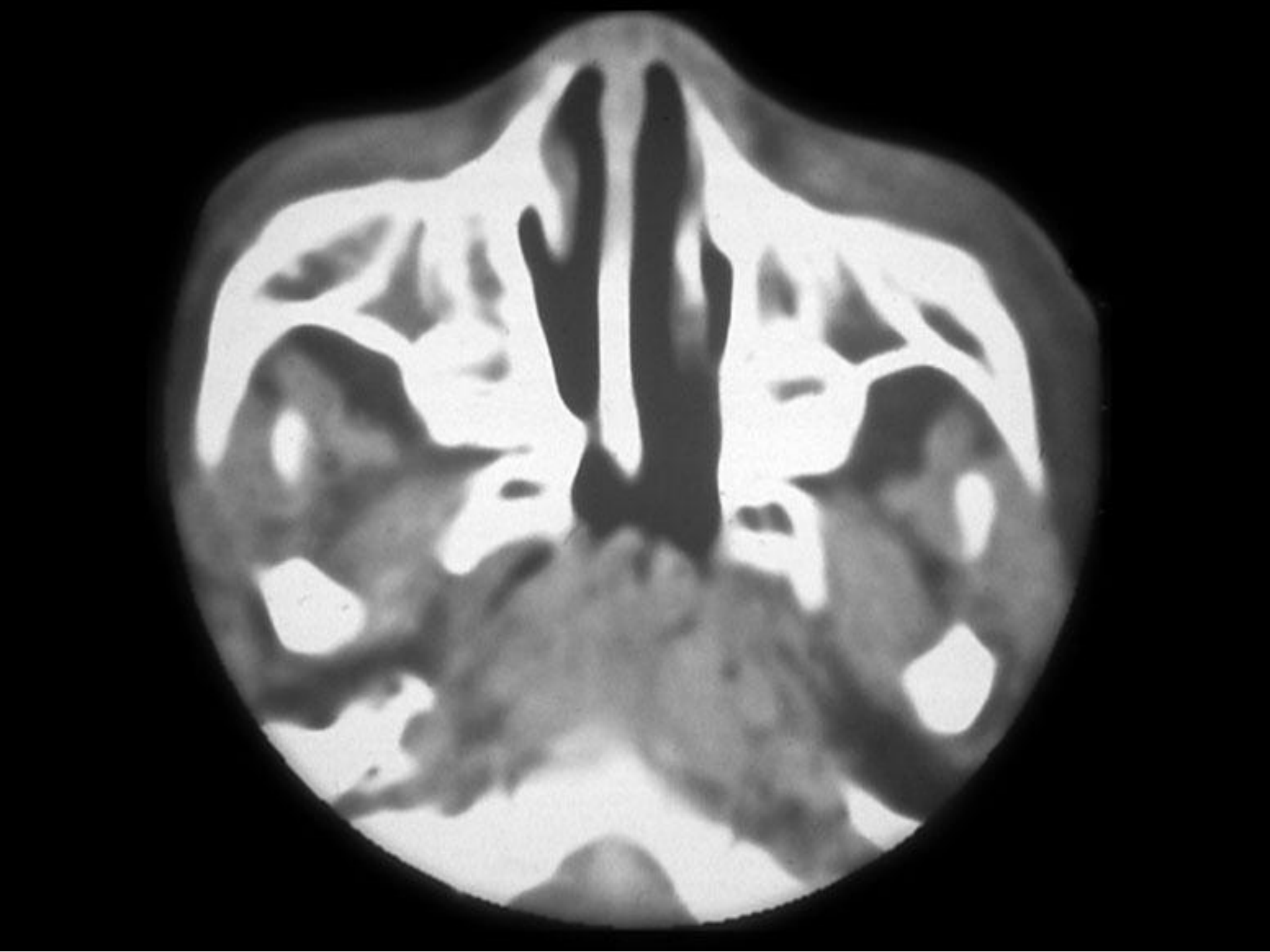
“CT in the evaluation of choanal atresia”
Laryngoscope, Healy 1987





CT – suck out nose!





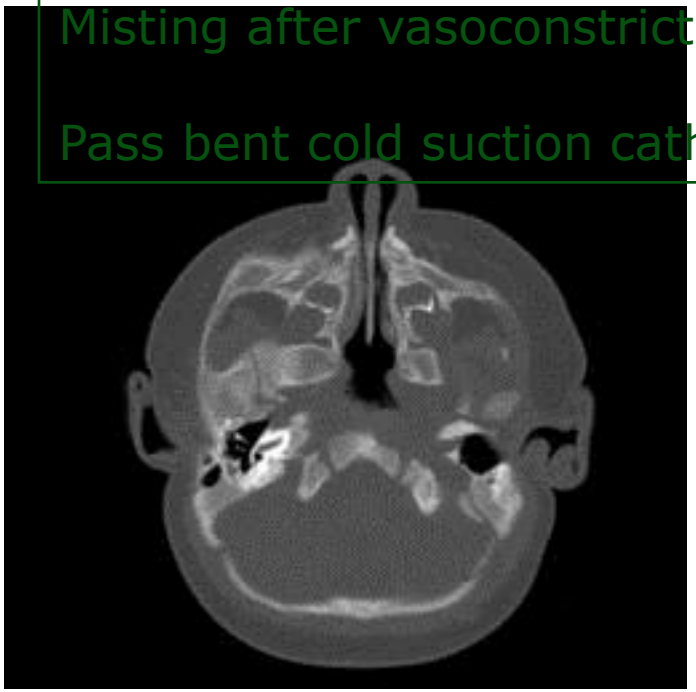
Differential diagnosis

- Neonatal rhinitis
- Masses
  - –Post nasal space
    - Teratoma
  - –Anterior nasal space
    - Glioma
    - *Midline nasal dermoid*
    - Meningocoele
- Mid nasal and pyriform aperture stenosis

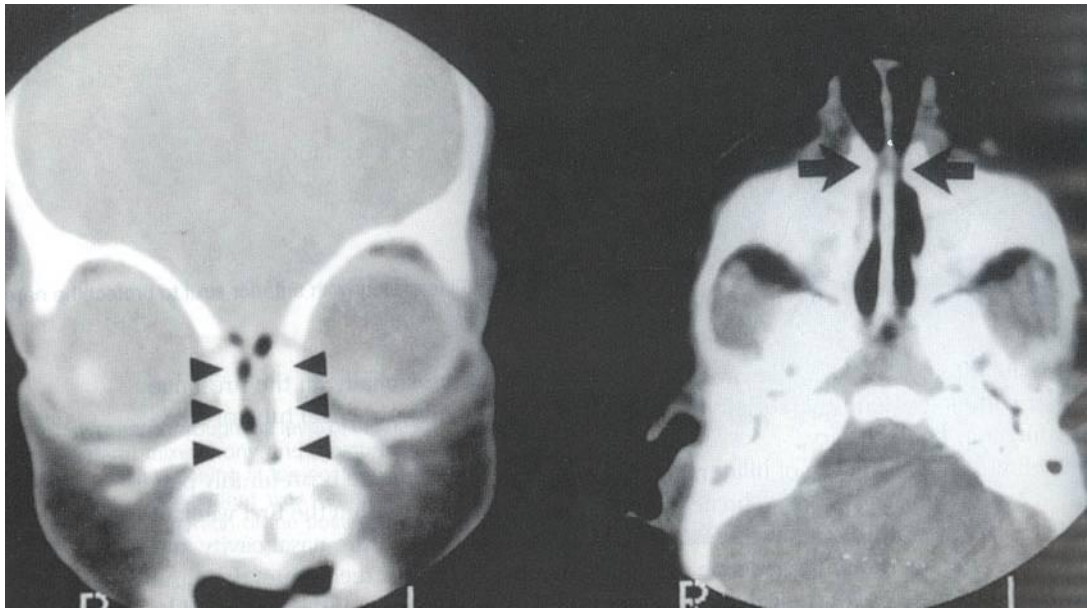
Top Tips #4

Misting after vasoconstriction

Pass bent cold suction catheter

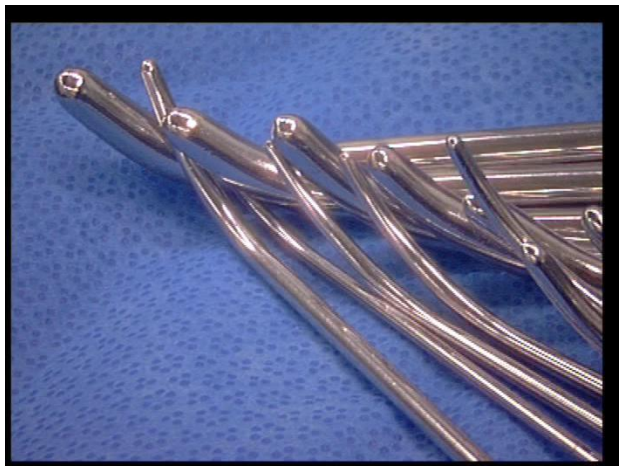


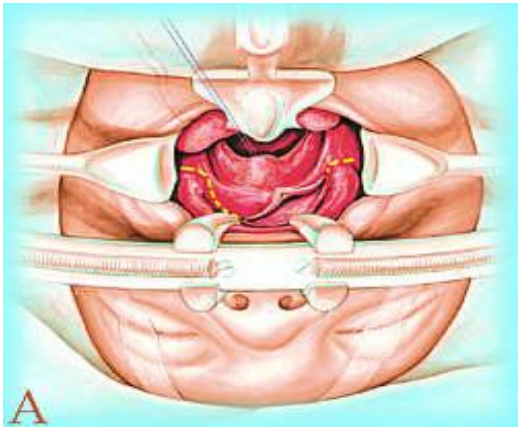
Pyriform Aperture Stenosis



How to Approach the atresia

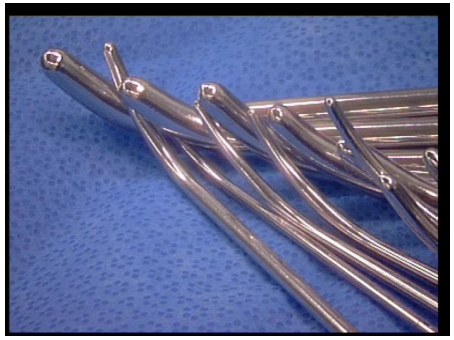
- Historical Approaches
  - –Blind anterior
  - –Palatal approach
- Current Approaches
  - –Anterior endoscopy 0°
  - –Posterior endoscopy 70-120°
  - –Ext rhinoplasty incision
  - –Sublabial incision (Koltai)



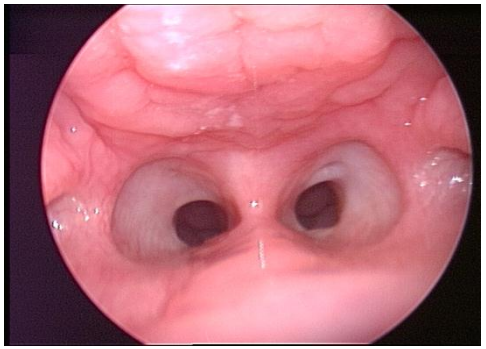


How to remove the atresia

- Technique
  - –Dilatation
  - –Drill
  - –Microdebrider

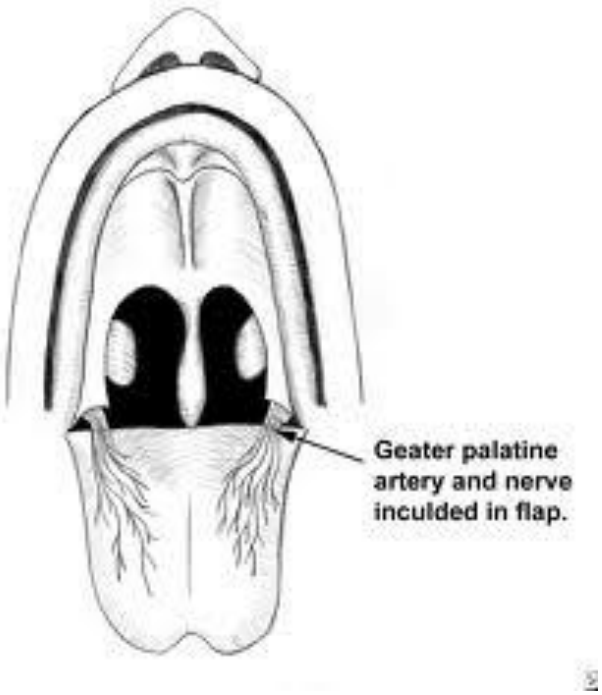


- Recurrence
  - –Laser
  - –Mitomycin

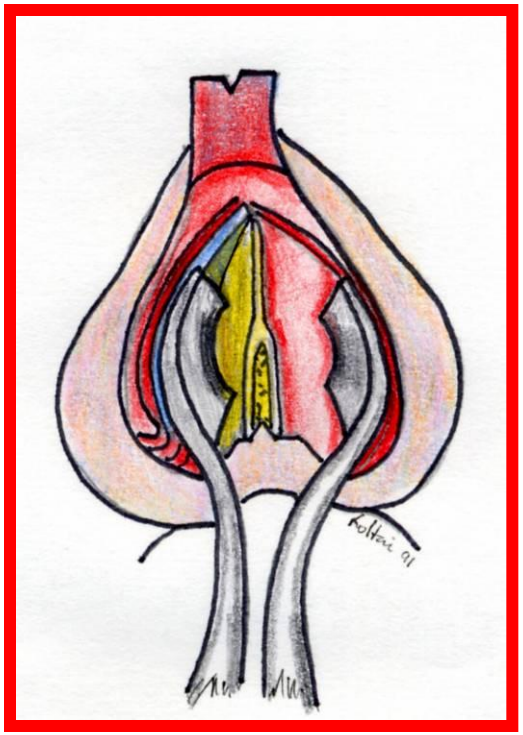


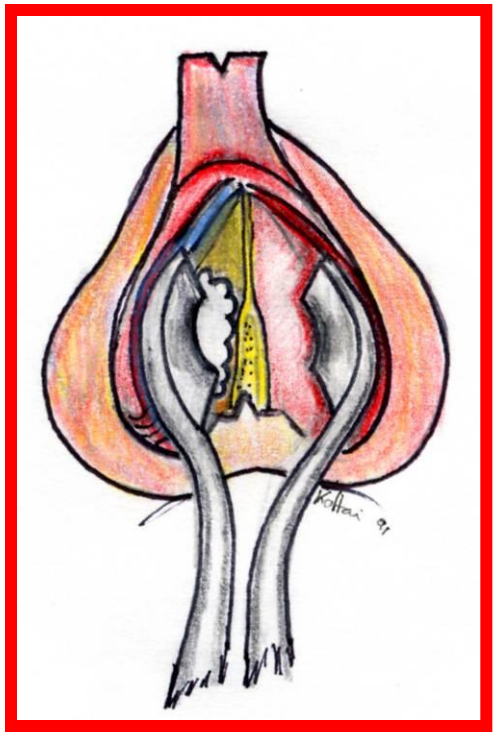
Palatal Approach

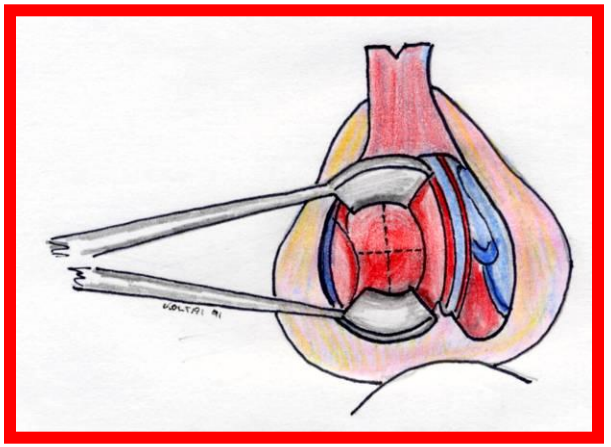
- ? Risk to mid facial growth

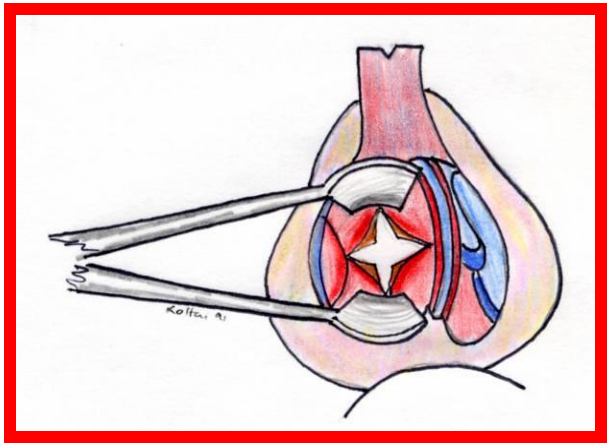


External rhinoplasty



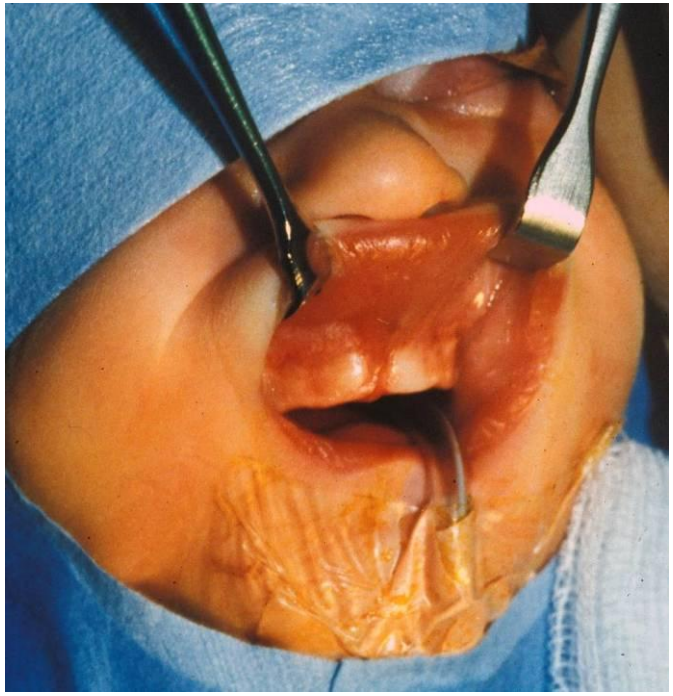


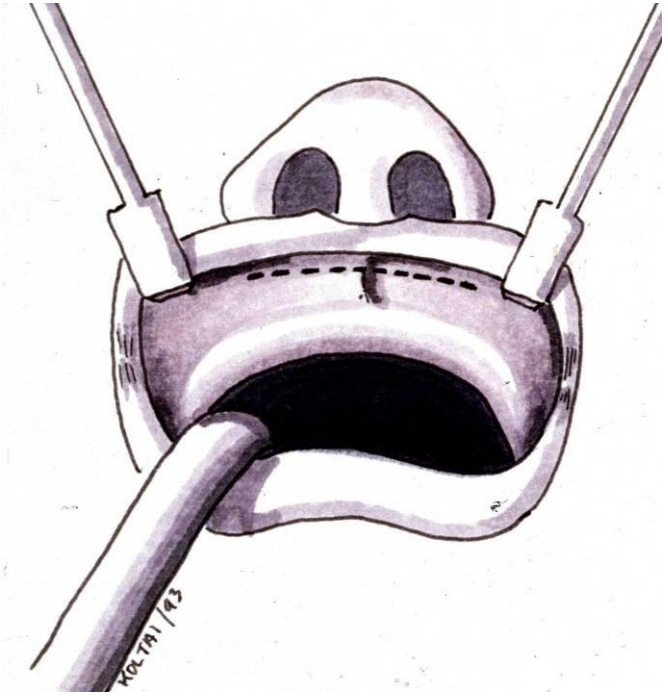




Koltai PJ: *The External Rhinoplasty for Unilateral Choanal Atresia* E.N.T. Journal. 1991.

Peter Koltai - sublabial approach



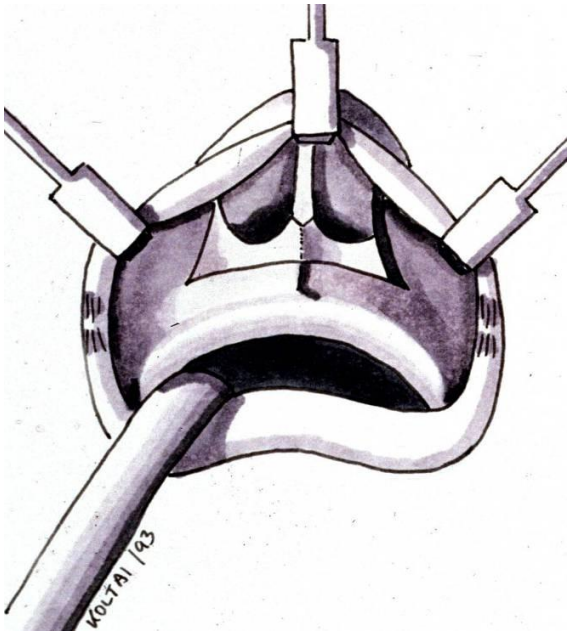


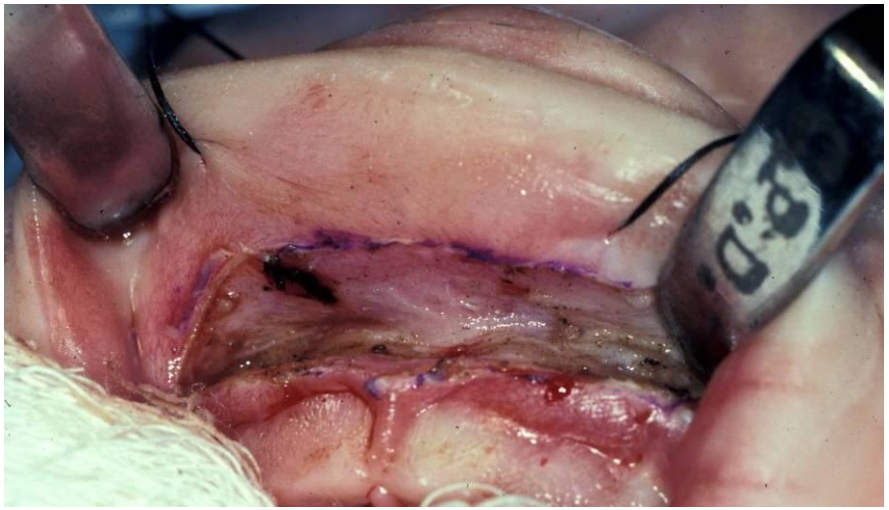
Inject sulcus with 1:100,000 epinephrine

Vasoconstrict nose with oxymetazoline

Inject nasal mucosa with epinephrine

Peter Koltai - sublabial approach



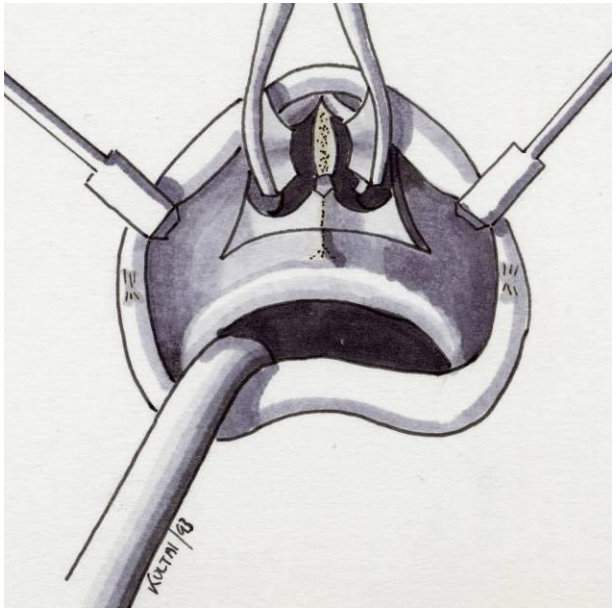


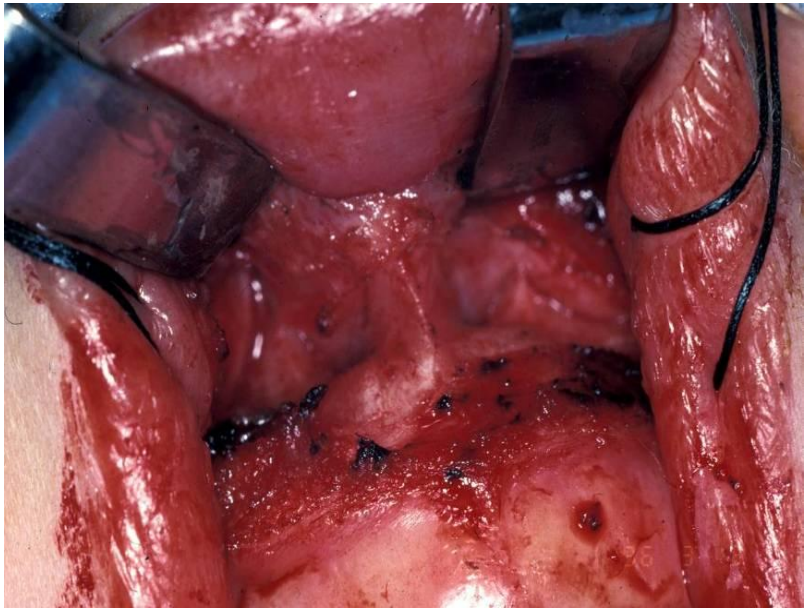
Incision with monopolar cautery at 12 watts

Cuff of labial tissue left on gingival side of incision

Elevate tissues in subperiosteal plane

Peter Koltai - sublabial approach





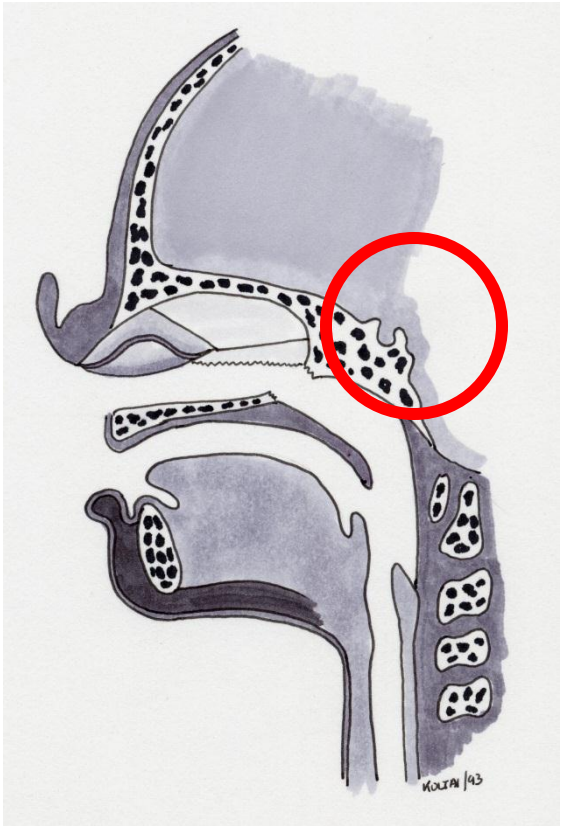
Elevate mucosa off of one side of the septum

Leave other side attached to septal cartilage

Dislocate cartilaginous septum laterally

Elevate mucosa off nasal floor on both sides

Peter Koltai - sublabial approach



Provides submucosal access:

- vomer
- atresia plate

Remove bone without mucosa

Mucosal incision is last step

7 cases 1987 – 1991:

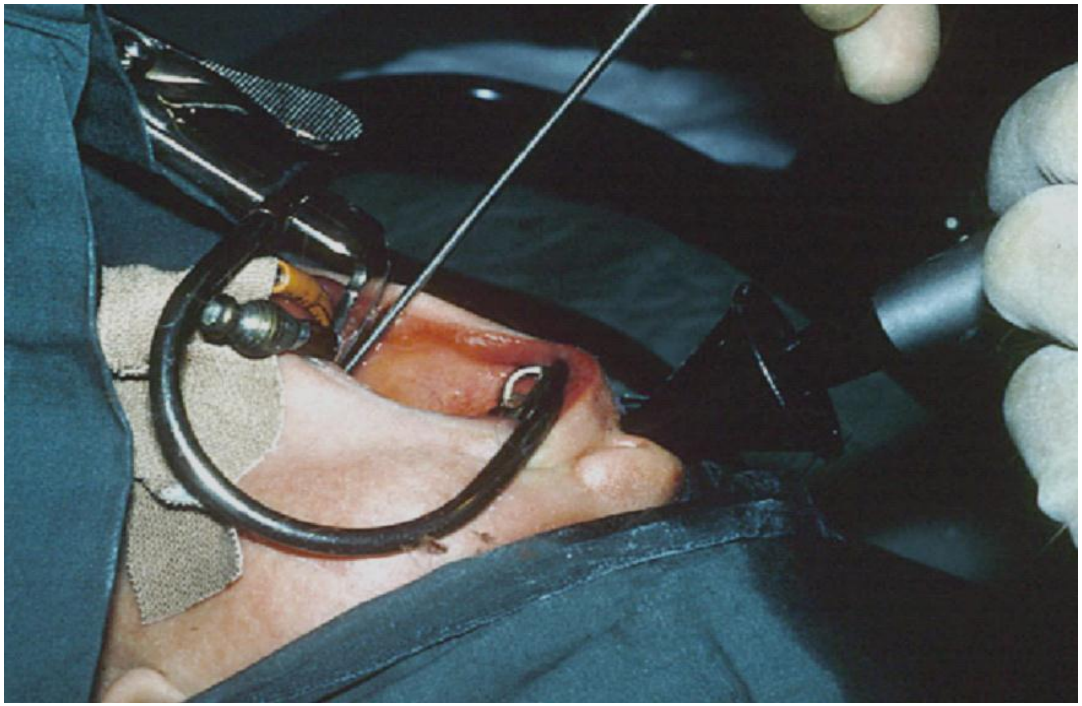
1 failure

albert.uk.com

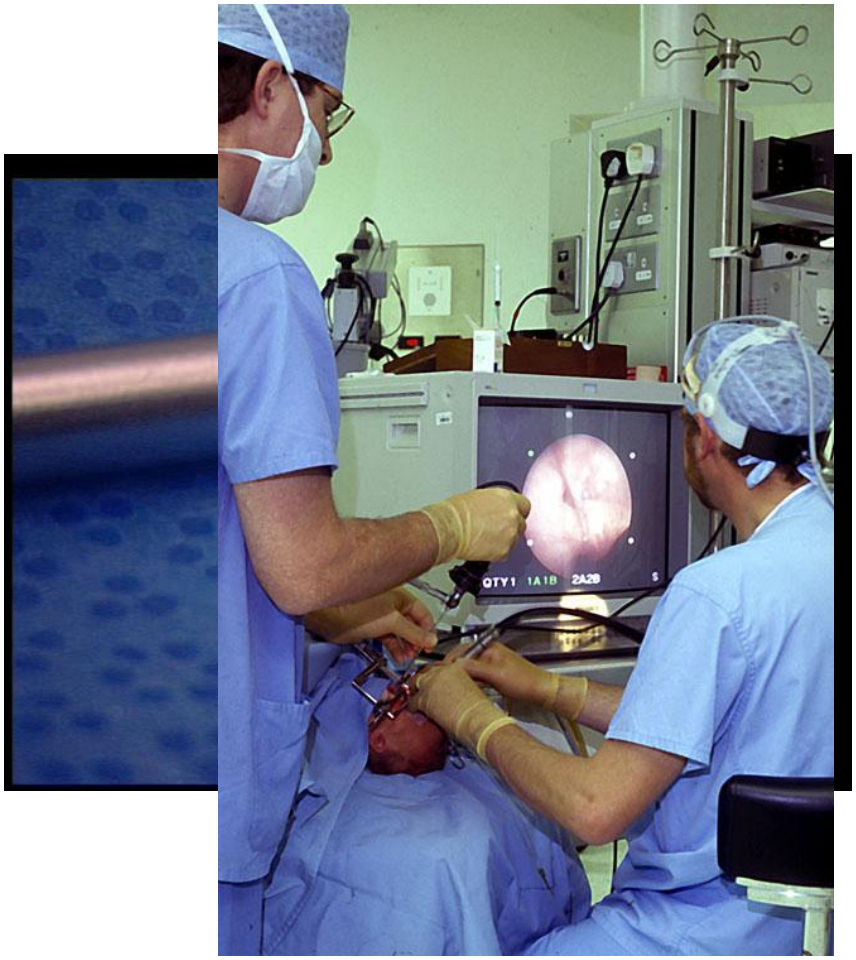
Which way to go?

Anterior	Posterior
Familiar Equipment available But … Very cramped in neonate	Need to learn orientation Need 120° Scope But …. Access even in premature or syndromic infants

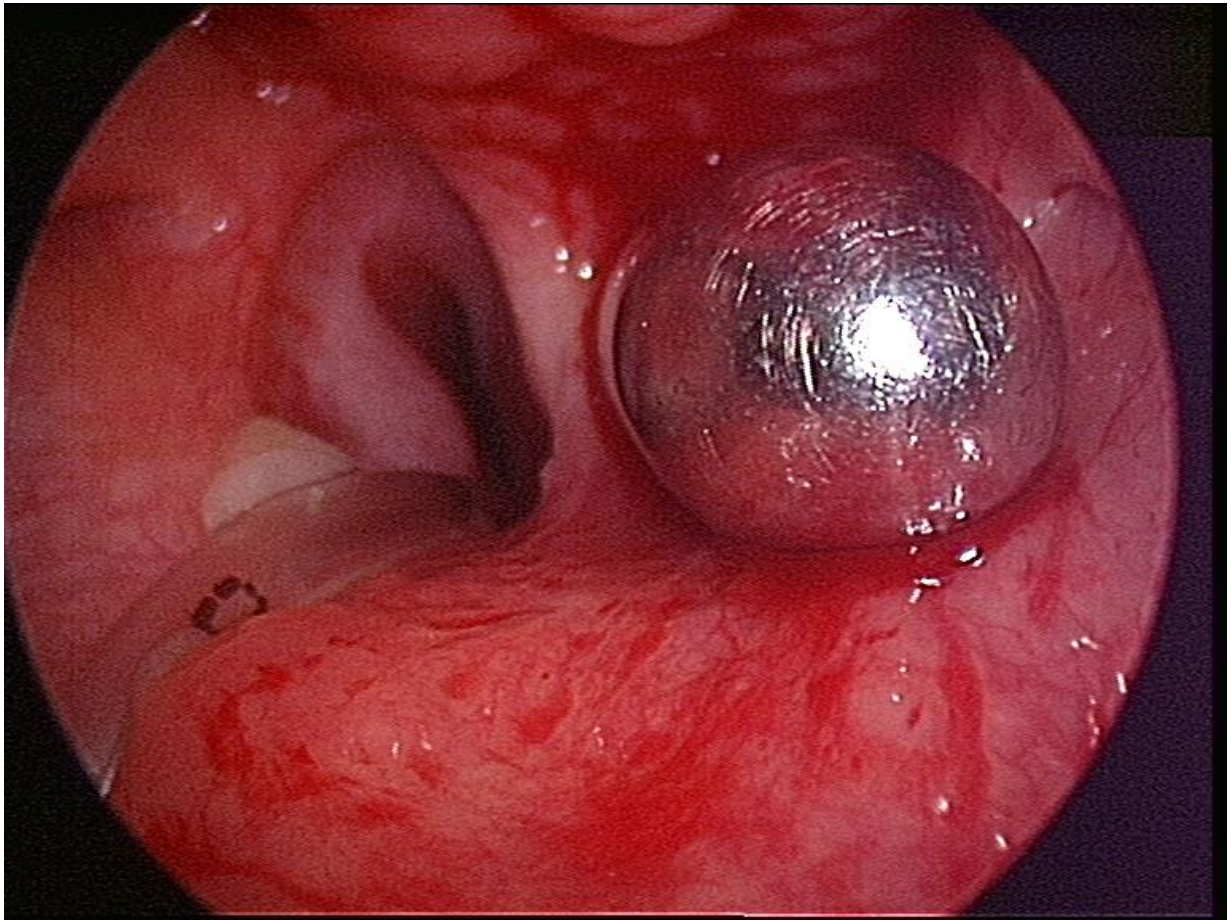
GOS - Original approach with otological drill



Original trans nasal approach with 120° telescope and drill



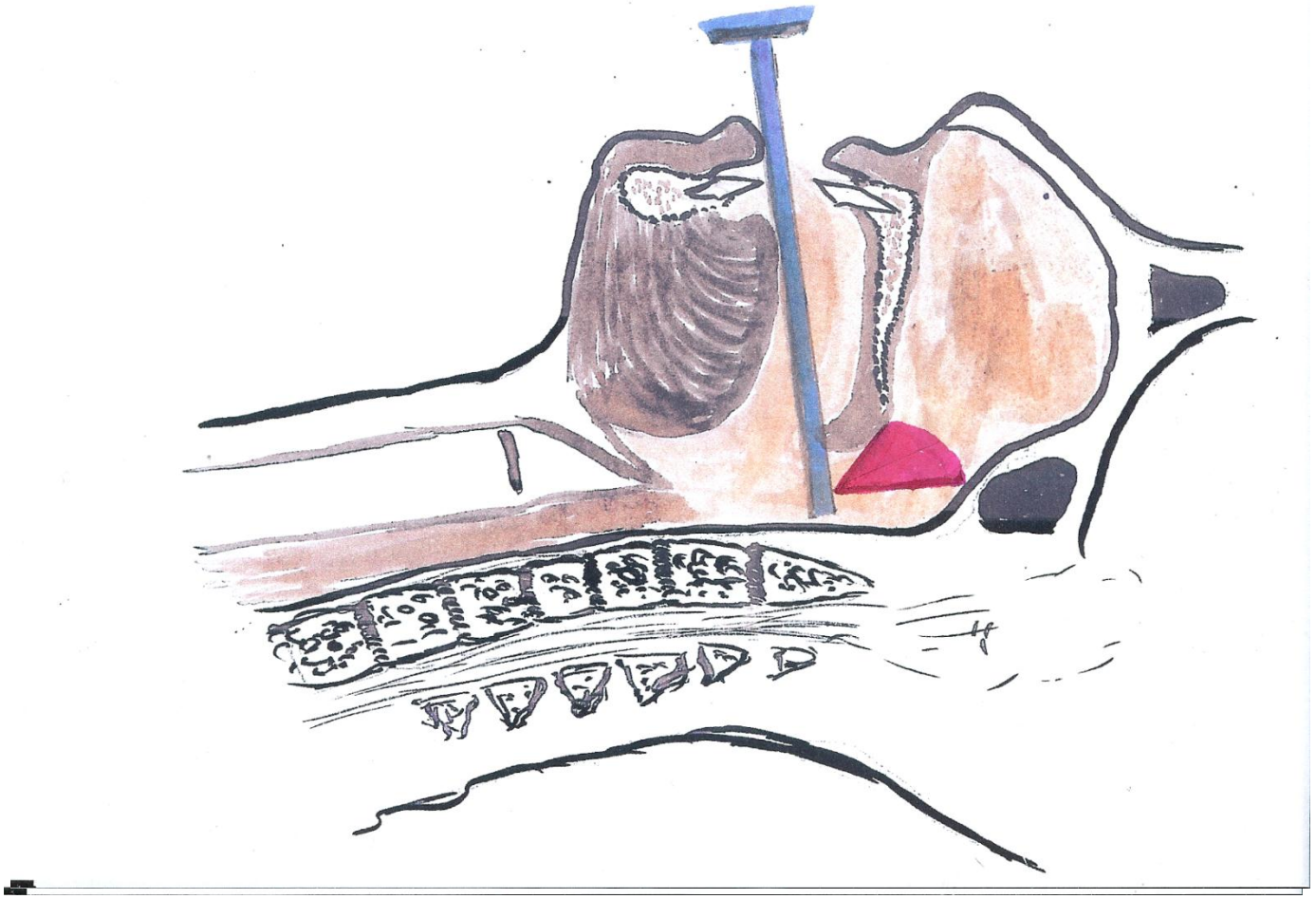
Traditional Dilatation:
Force is mostly longitudinal

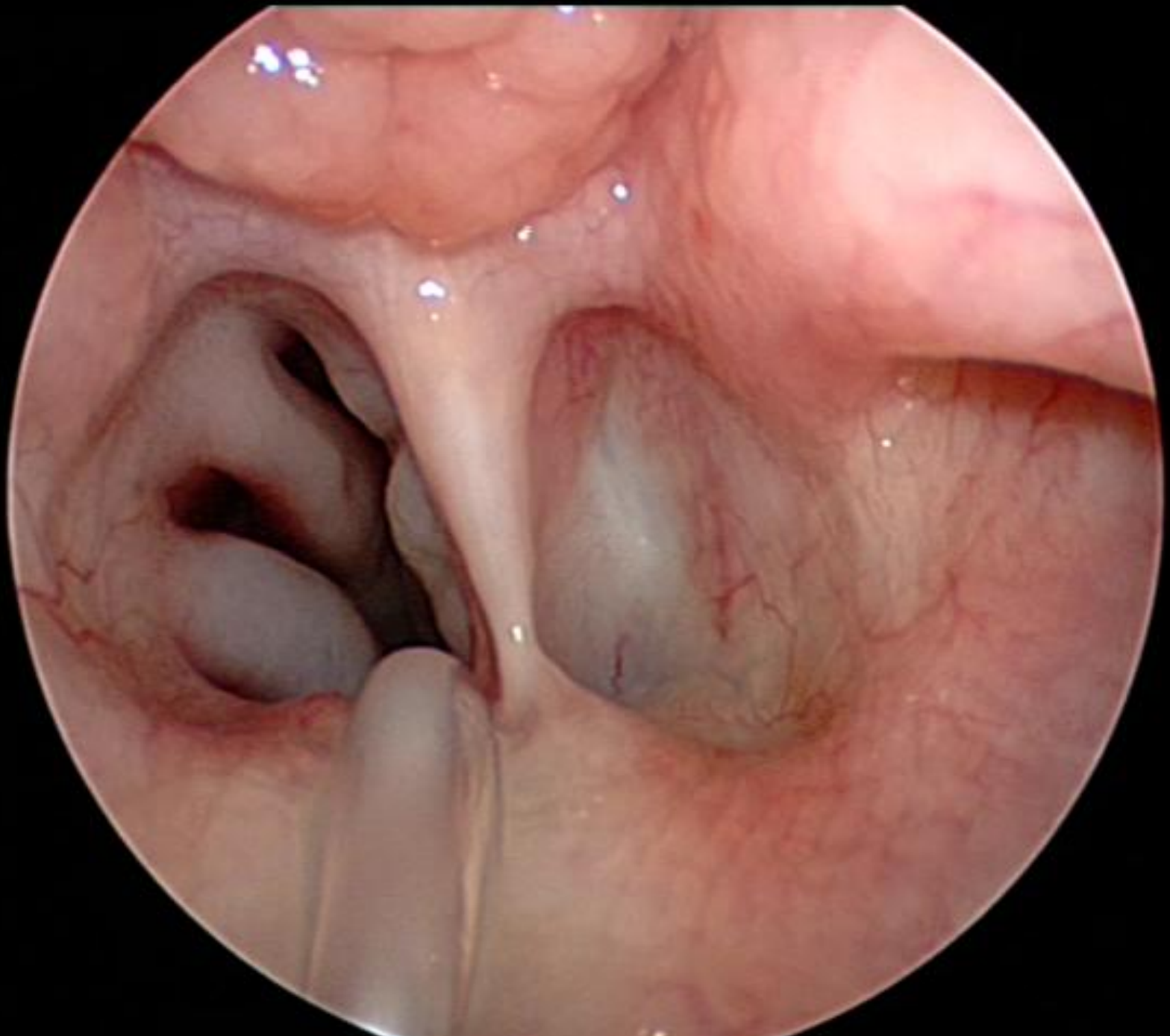


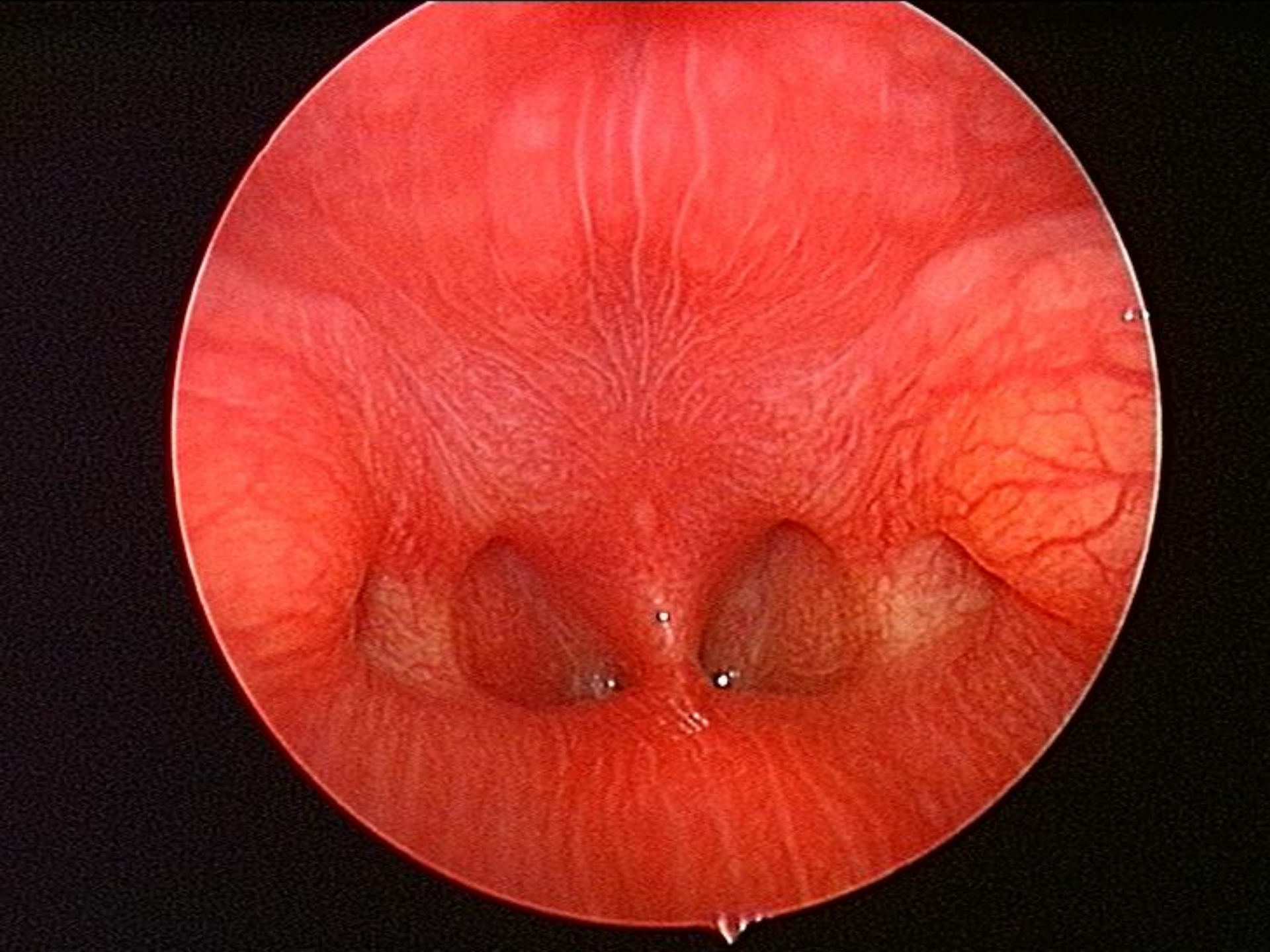
Current technique

- 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

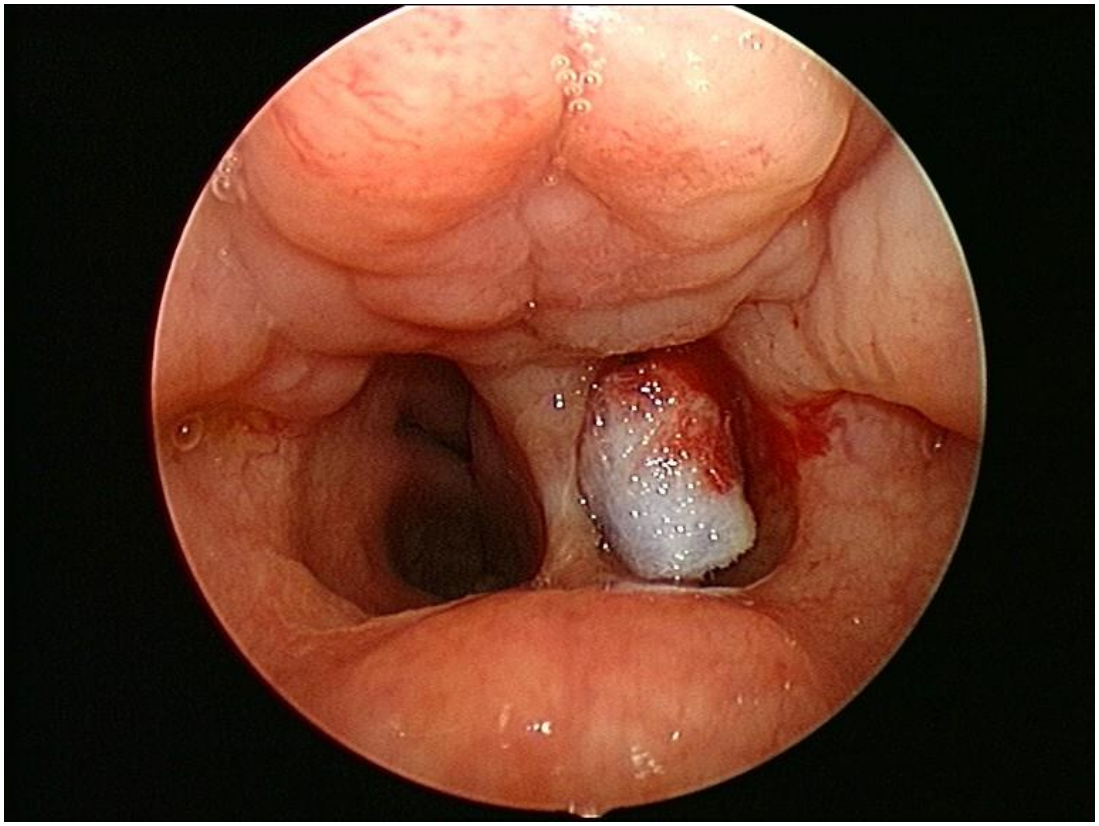
120 ^{0} Telescope to view choanae



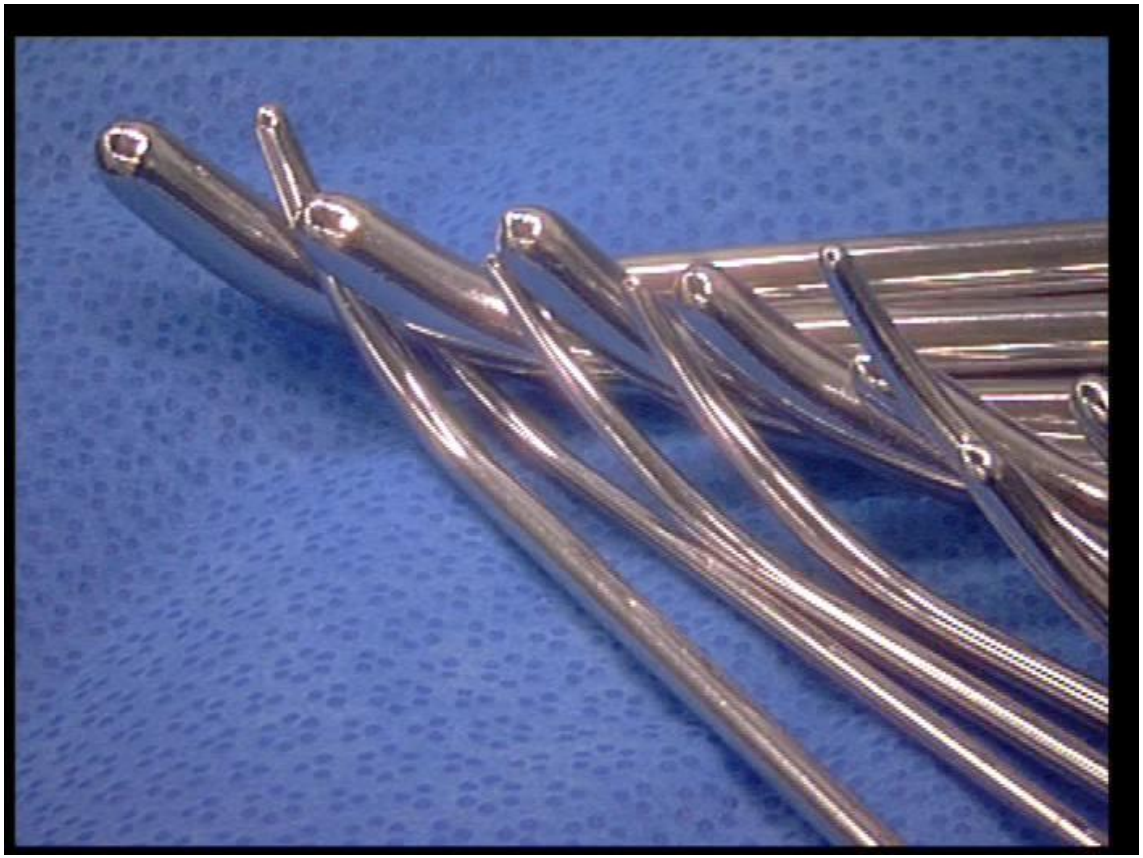




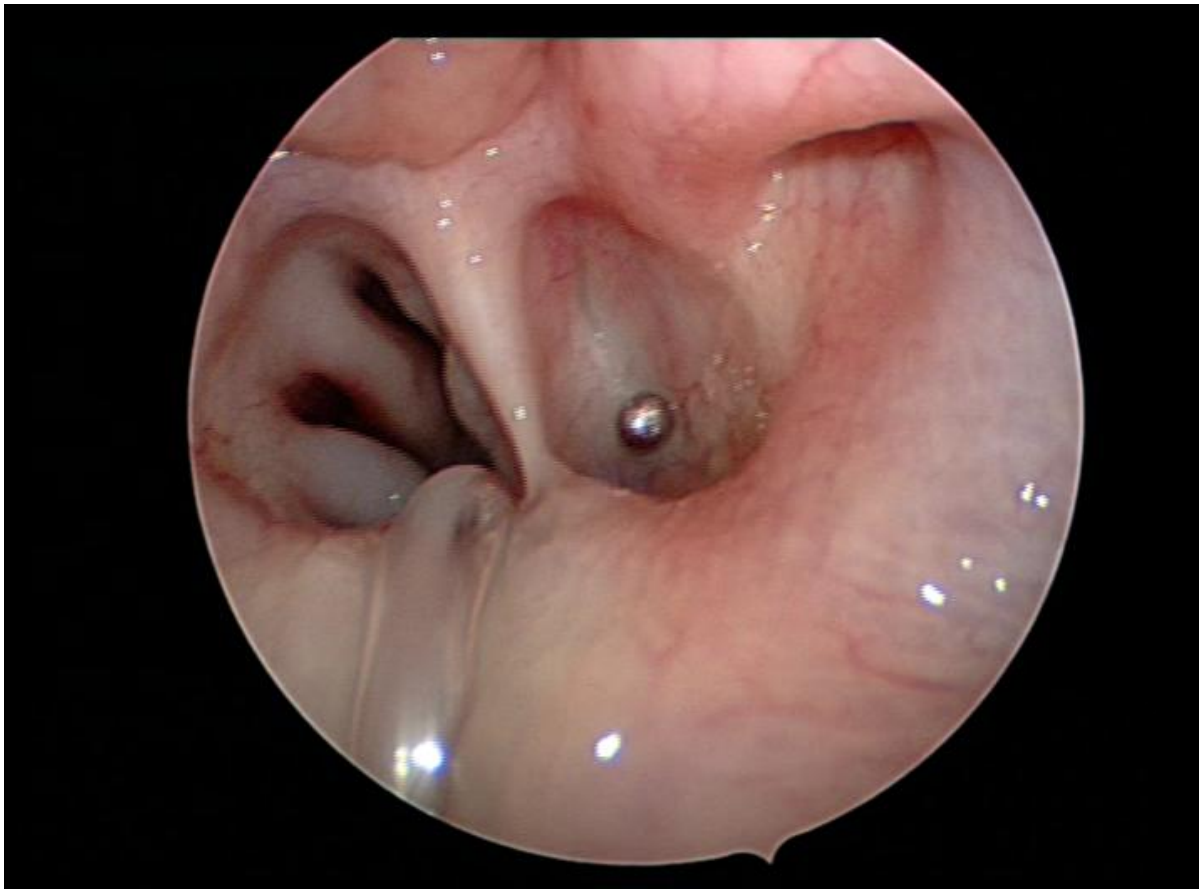
Adrenaline:
the key to a bloodless field



Dilators: Use with care



Using Dilators to perforate soft centre



120 ^{0} Telescope and Microdebrider

Microdebrider - what a relief!

A. Pediatric Round Bur, 2.9mm

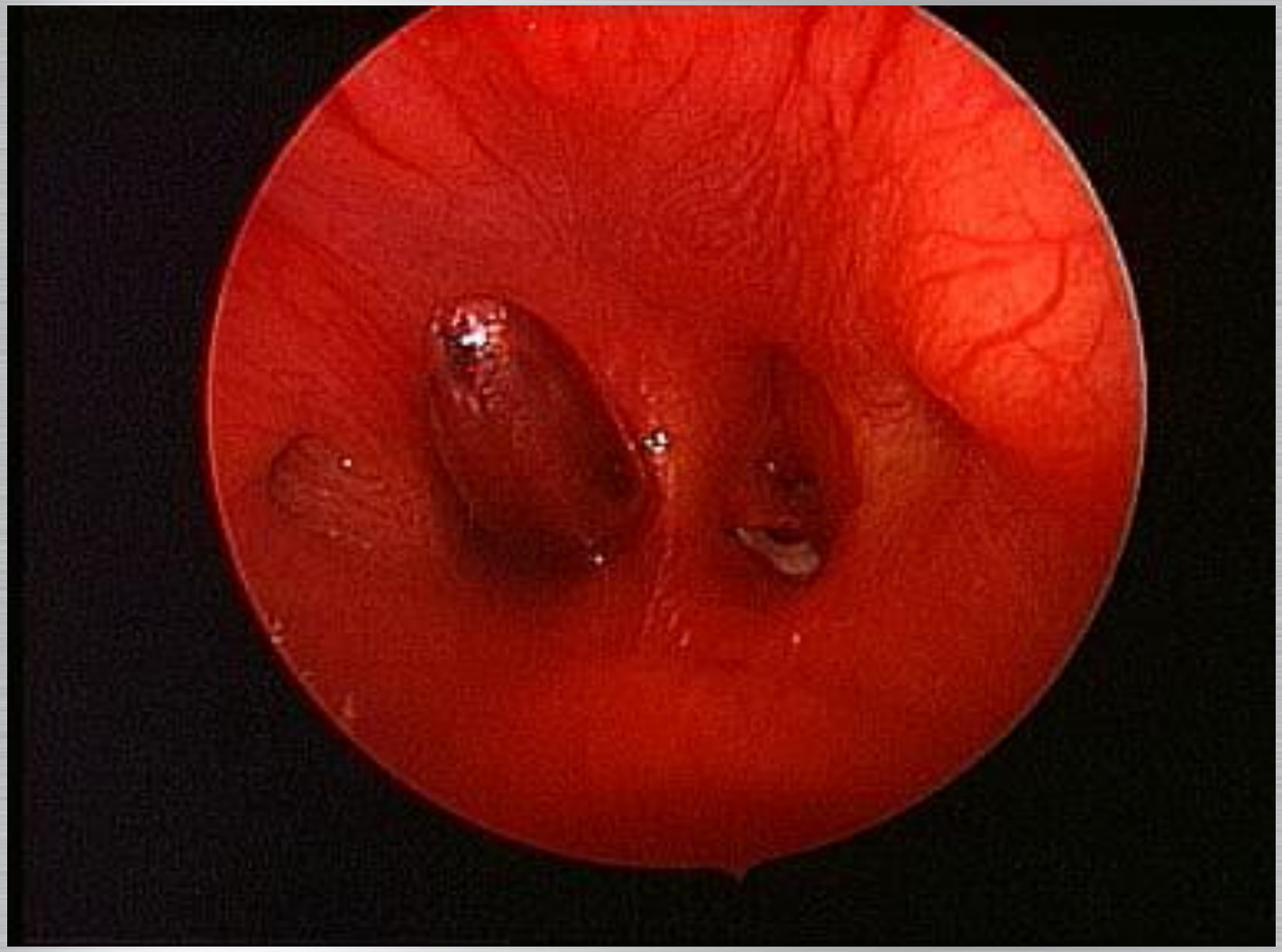
- Length: 10cm
- Straight shaft
- Application: choanal atresia
- Operating speed: up to 5,000 RPM FWD
- 5/box

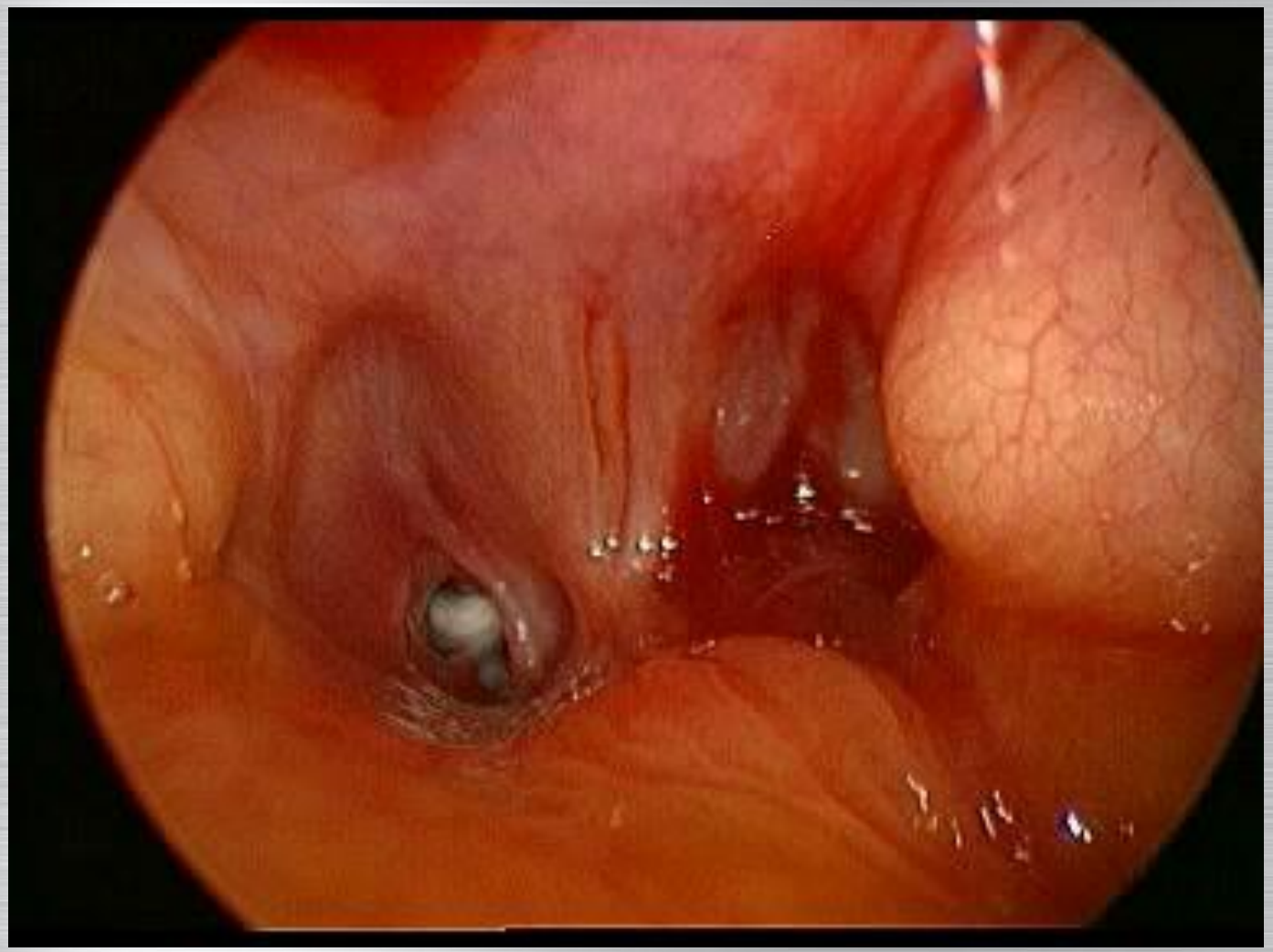
18-82960

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

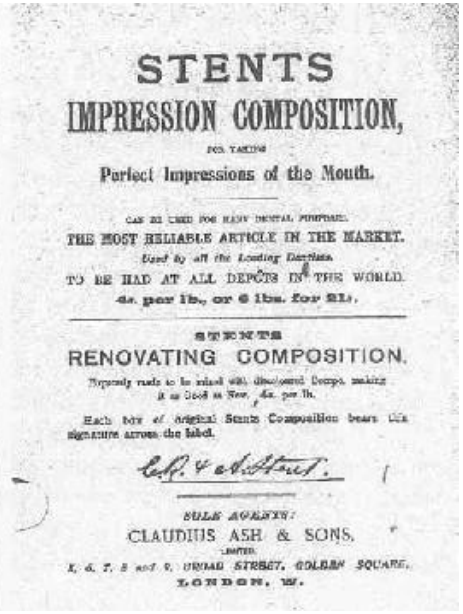
18-83673HS



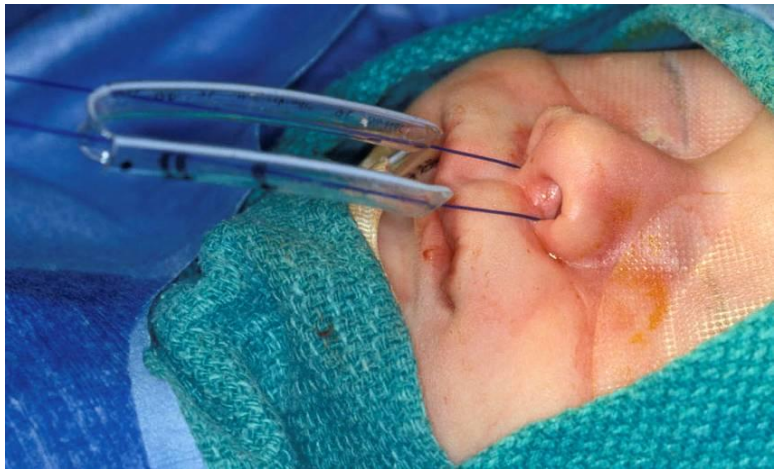


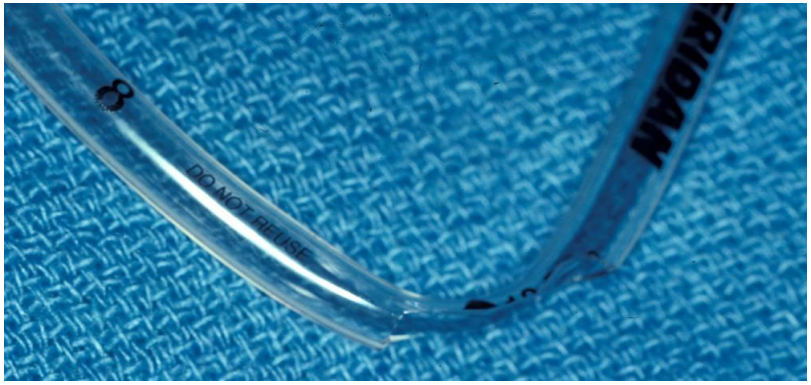
Stents

- Dr. Charles Stent, 1807 – 1885 Dentist
- Improved gutta percha by adding stearine, talc and colour



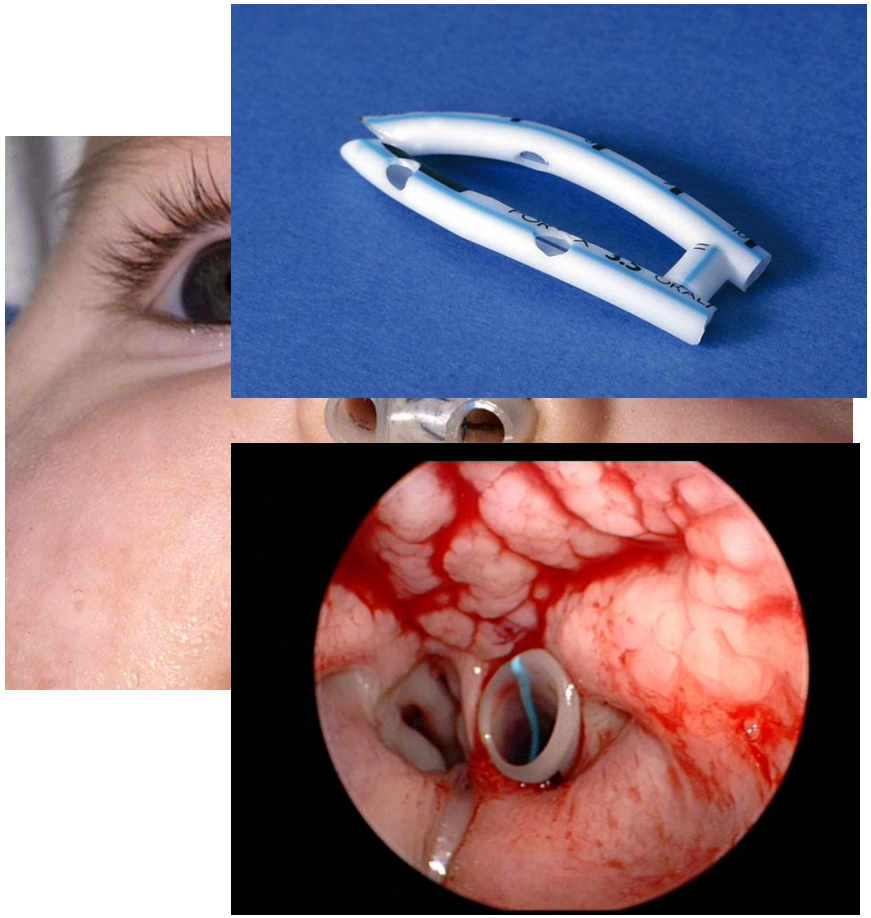
Original GOS stents: columella necrosis



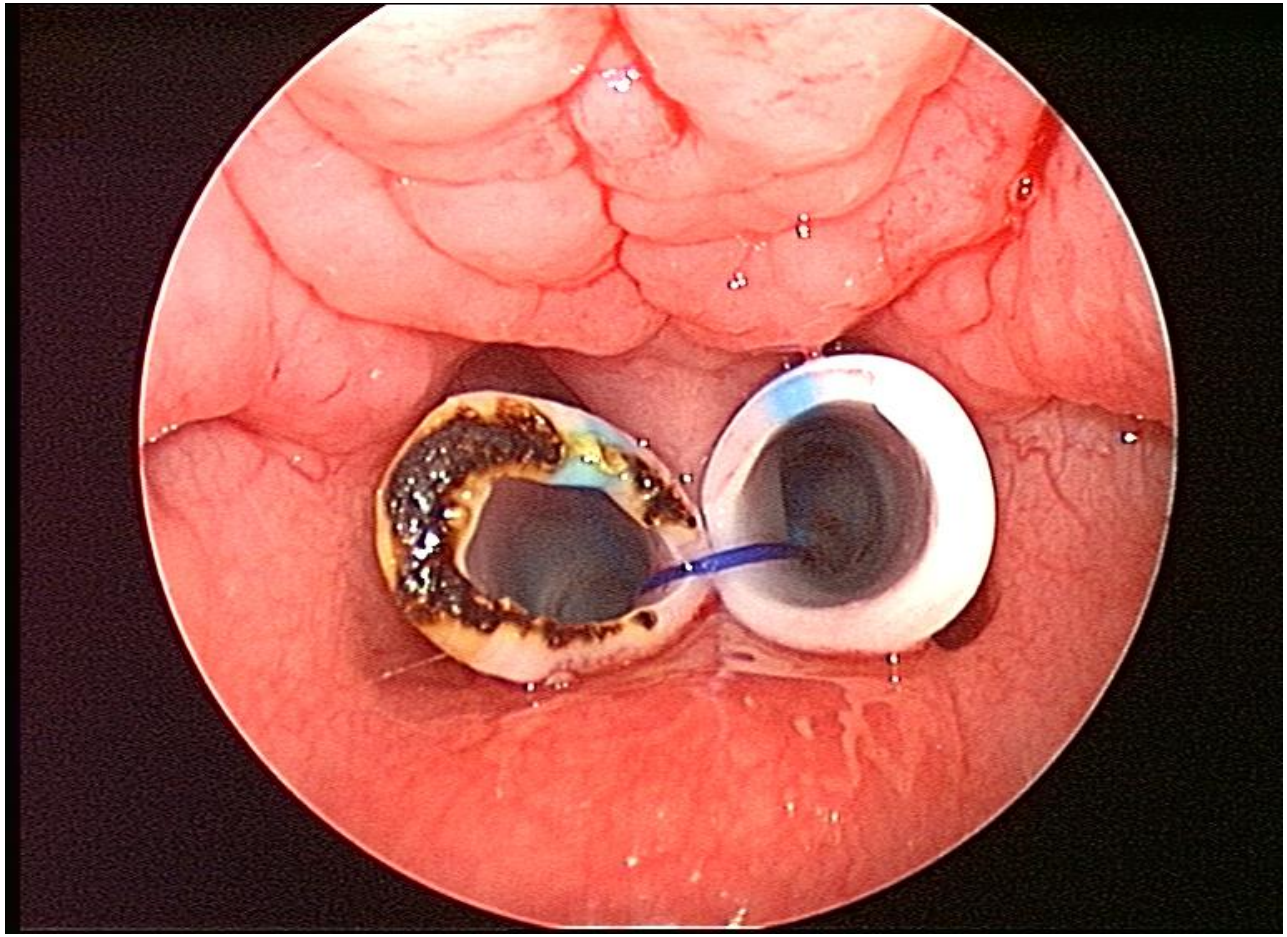


Redesigned GOS Stents

- Bilateral cases
  - External bridge piece
  - 6 weeks
  - 4.5 Portex for term
- Unilateral
  - Intranasal if at all

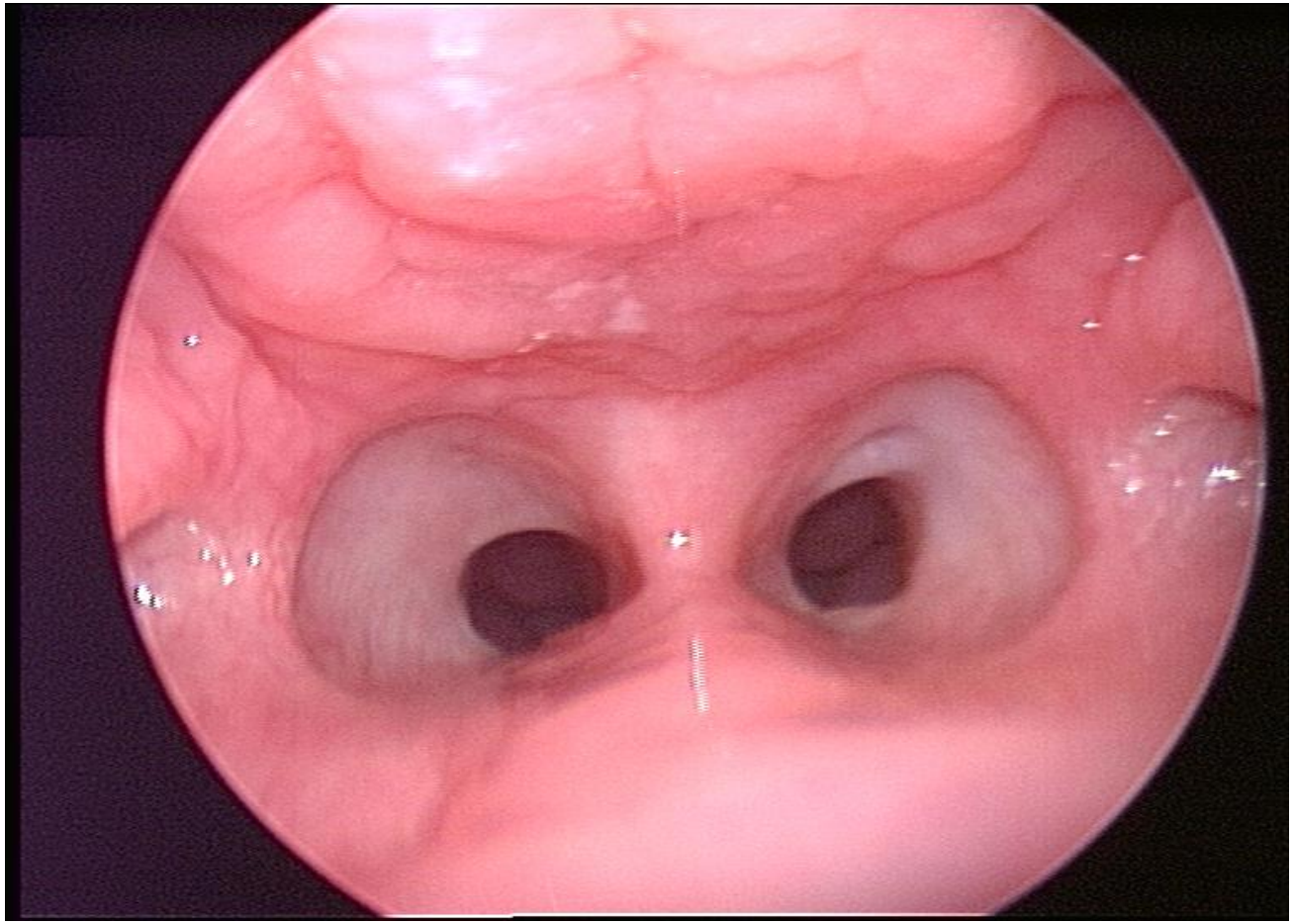


Bilateral stents after 6 weeks

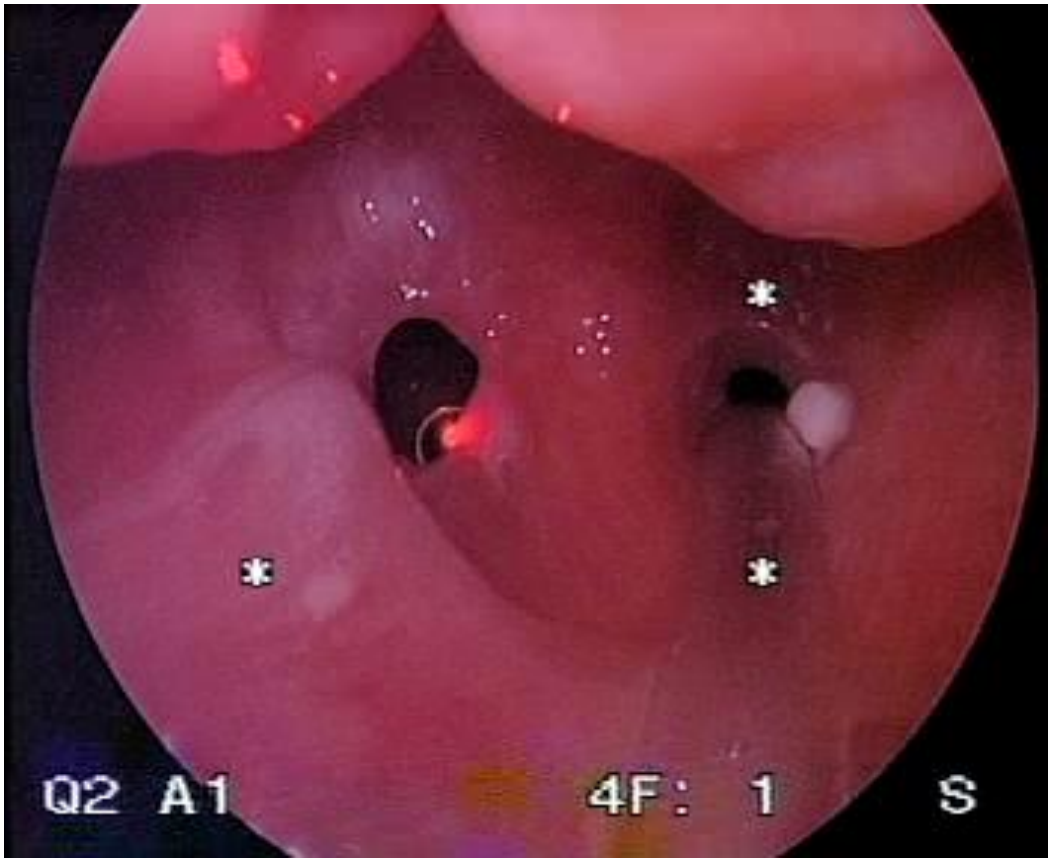


Unilateral stent after 6 weeks

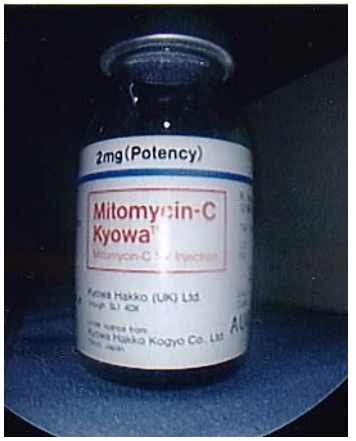
Restenosis



KTP laser



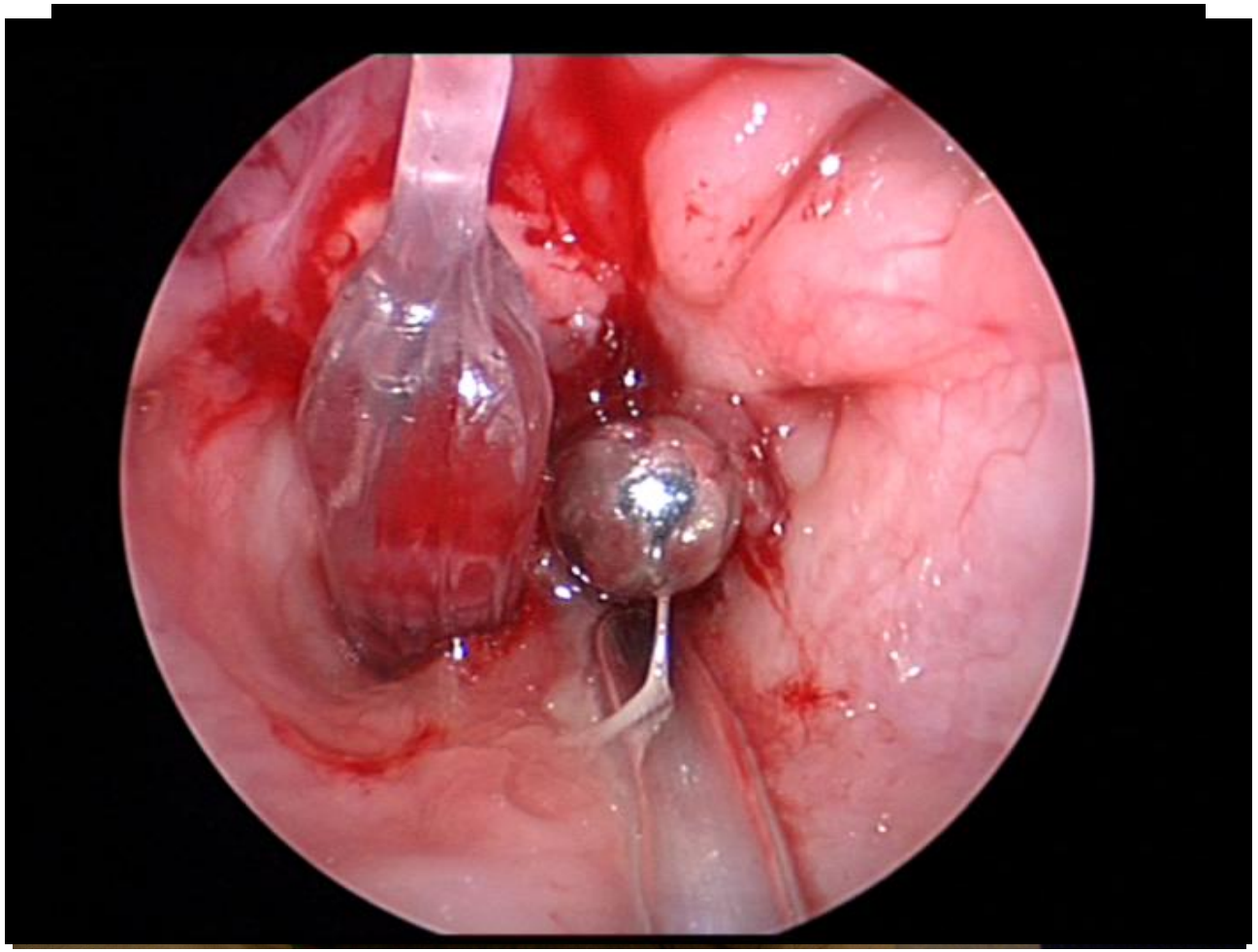
Mitomycin



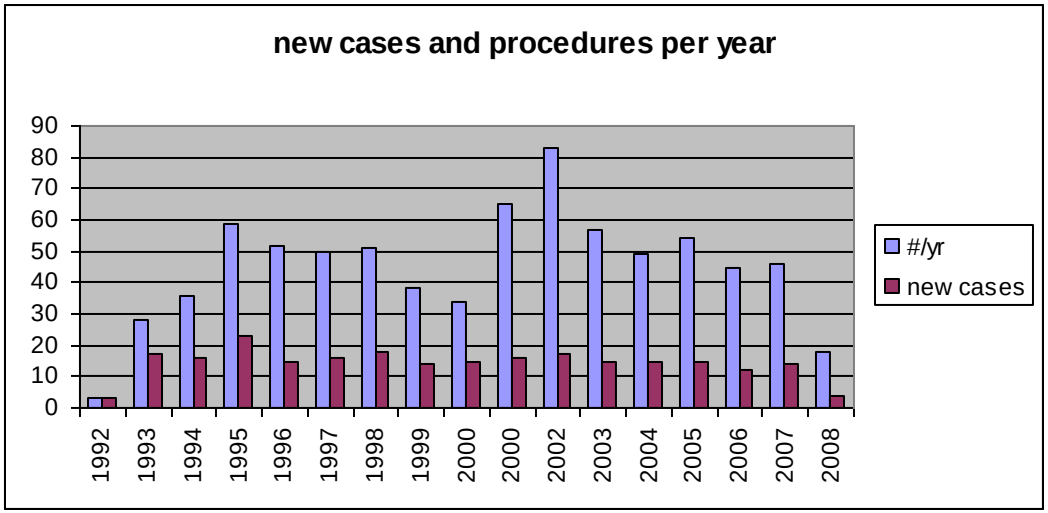
- Sub-cytotoxic dose inhibits fibroblasts
- 2 mg/ml applied topically for 4 minutes

- Topical Mitomycin as an Adjunct to Choanal Atresia Repair -
  - *Arch Otolaryngol. 2002;128:398-400.*
- Surgical Management of Choanal Atresia Improved Outcome Using Mitomycin (0.4 mg/mL)
  - *Arch Otolaryngol. 2001;127:1375-1380.*
- The expression of mRNA for some extracellular matrix proteins (elastase, hyaluronidase, and procollagen) was downregulated in the mitomycin test groups
  - *Laryngoscope. 113(2):237-242, February 2003*
  -

Balloon dilatation



1992-2008



50/yr

10-15/yr

Albert 2008

GOS Choanal atresia population

- 10 year: 1992 – 2002 (Kubba 2004)
  - (108 records from 129 new cases)
- 15 year: 1992 – 2007 (Cochrane 2007, updated Albert 2008)
- (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)

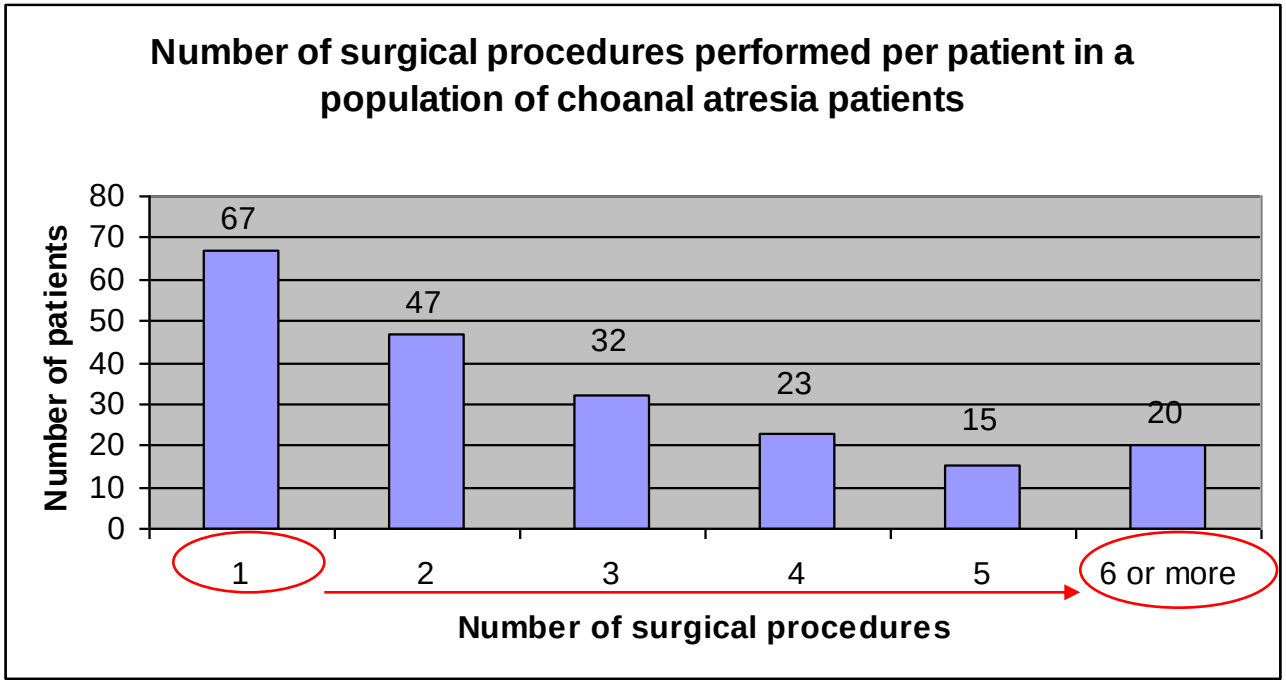
•symptomatic at last follow-up

14%

Kubba 2004

Review of persistent restenosis

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



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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