

Congenital Midline Nasal Masses

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UTMB Grand rounds

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Introduction

- Dermoid sinuses and cysts
- Gliomas
- Encephaloceles

- 1:20,000 to 1:40,000 births
- All three have potential intracranial connections
- May present as a mass on nasal dorsum or as intranasal mass
- Biopsy can lead to meningitis and CSF leak
- Treatment is surgical excision

- Most present in infants and children
- Any unilateral nasal mass in a child should be evaluated for a congenital midline mass

Differential

- Inflammatory lesions
- Traumatic deformity
- Benign neoplasms
- Malignant neoplasms
- Congenital masses

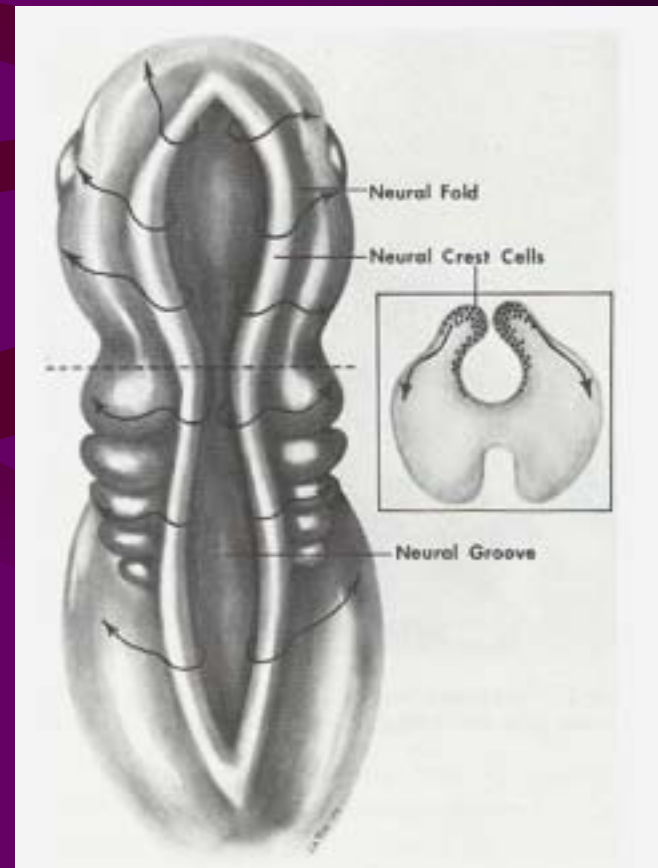
Topics

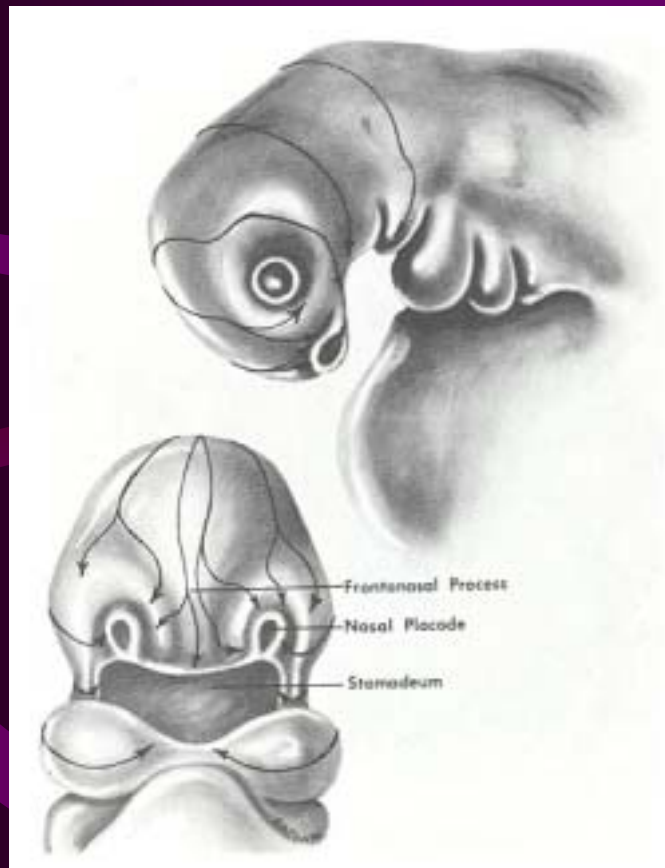
- Embryology
- Dermoid Sinus Cysts
- Gliomas
- Encephaloceles
- Evaluation
- Surgical Treatment

Embryology

- The critical period in nasal development is in first twelve weeks of fetal development
- Abnormalities of development are believed to cause gliomas, dermoids, and encephaloceles

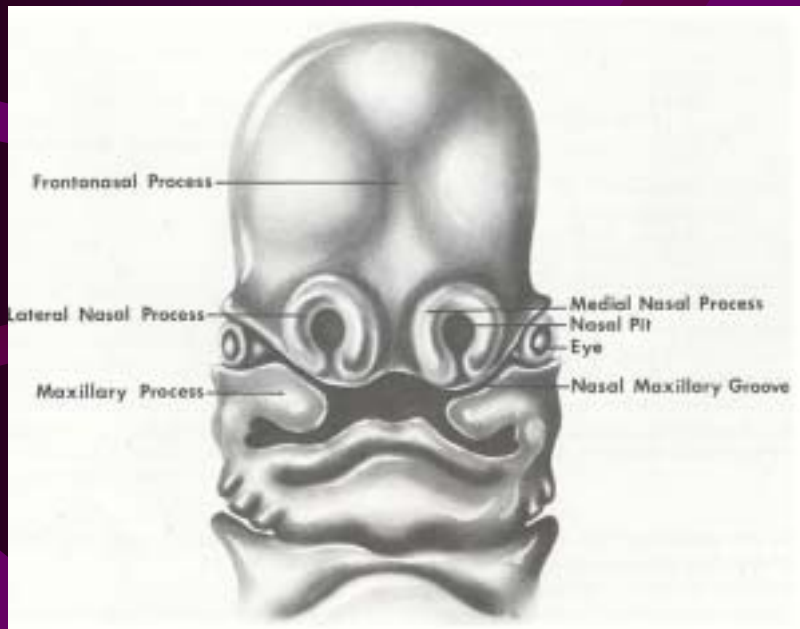
- 3-4 weeks neural fold develops
- Closure occurs from the midline and extends cranially and caudally



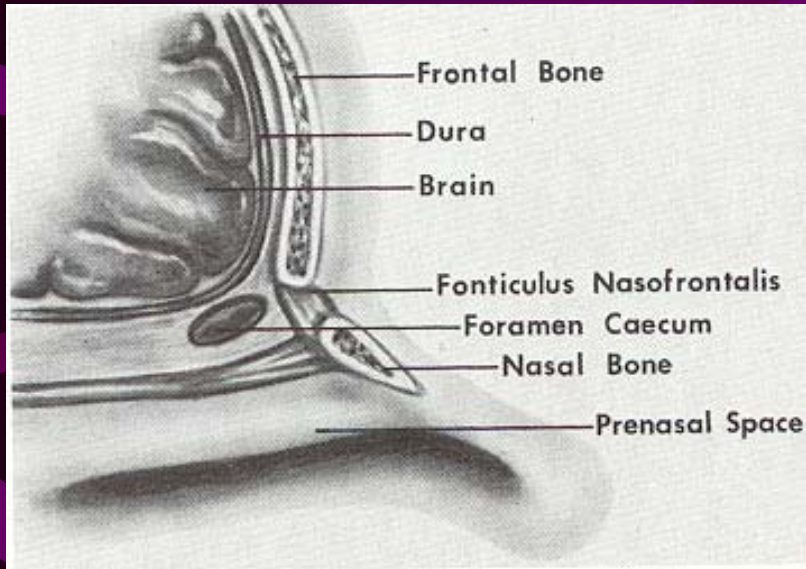


- Neural crest cells play a key role in facial development
- As the neural groove closes neural crest cells migrate around the eyes to the frontonasal process

- In most of the body neural crest cells are involved in ectodermal components
- In the face the primary role is mesenchymal cells
- Bone, cartilage, and muscles of the face are all derivatives



- Nose develops from frontonasal process and two nasal placodes
- Medial processes fuse
- Nasal-maxillary groove becomes the nasolacrimal duct



- Mesenchymal structures form in centers which fuse
- Key spaces are the foramen caecum, fonticulus nasofrontalis, and the prenasal space

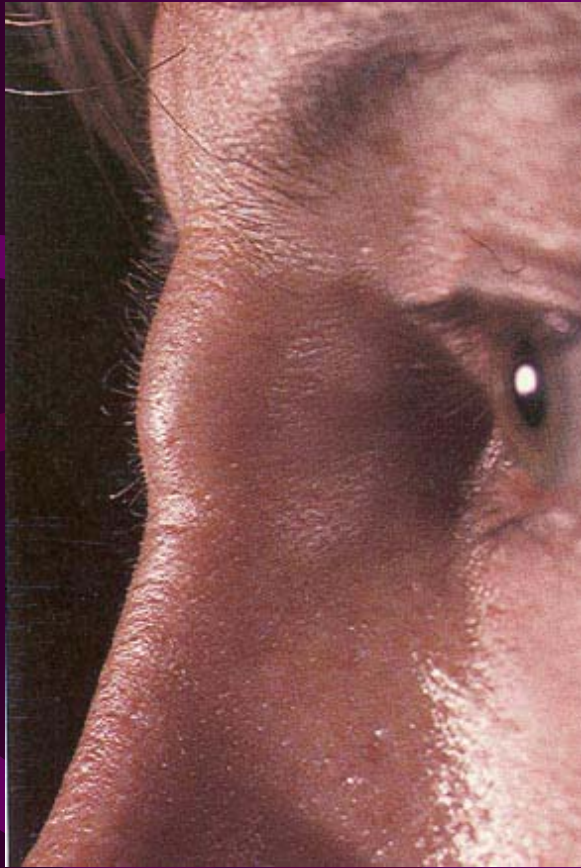
Dermoids

- Cyst or sinus
- Most common congenital midline mass
- 1-3% of all dermoids
- 3-12% dermoids of head and neck

- ectodermal and mesodermal
- Midline nasal pit, fistula, or infected mass from glabella to columella
- Sometimes single cutaneous tract with hair at opening
- May secrete pus or sebaceous material



Complications



- Intermittent inflammation
- Abscess
- Osteomyelitis
- Broaden nasal root
- Meningitis
- Cerebral abscess

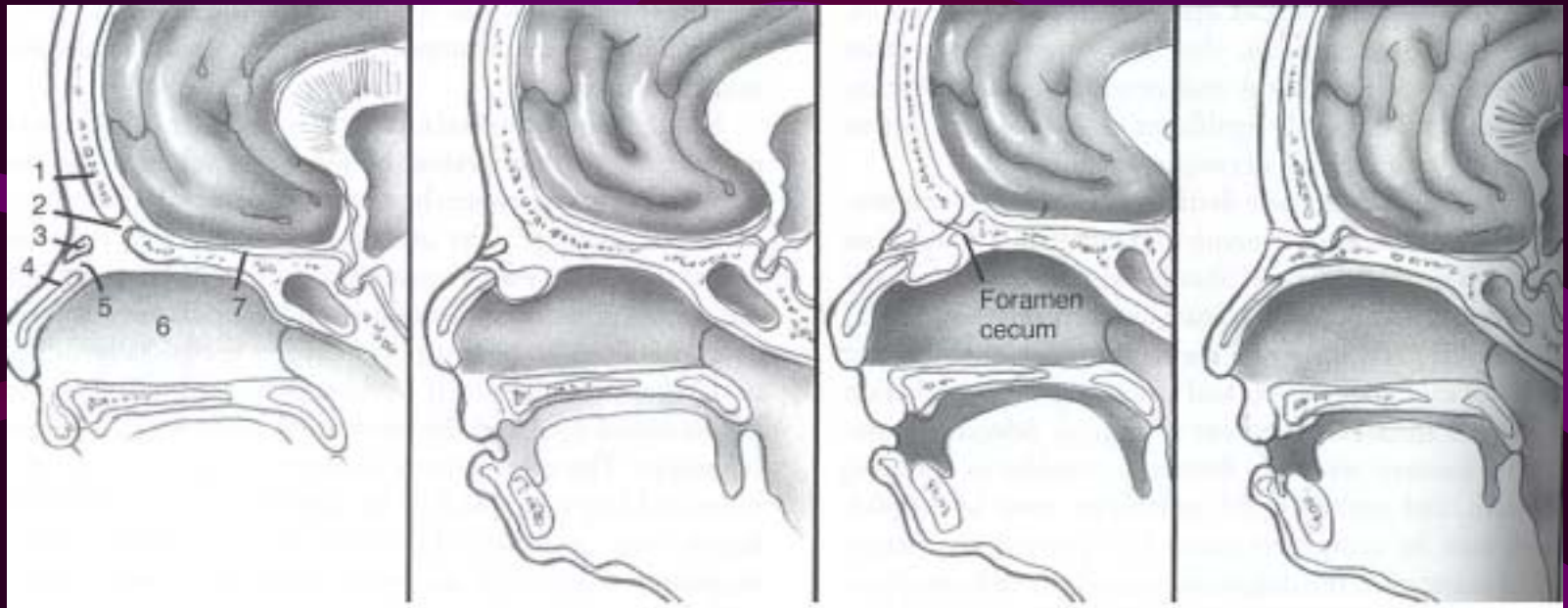
- CNS connection variably reported from 4-45%
- Associated congenital anomalies 5-41%

Associated Abnormalities

- Aural atresia, mental retardation, spinal column abnormalities, hydrocephalus, hypertelorism, hemifacial microsomia, albinism, corpus callosum agenesis, cerebral atrophy, lumbar lipoma, dermal cyst of the frontal lobe, coronary artery anomaly, cleft lip and palate, tracheoesophageal fistula, cardiac, genital and cerebral anomalies

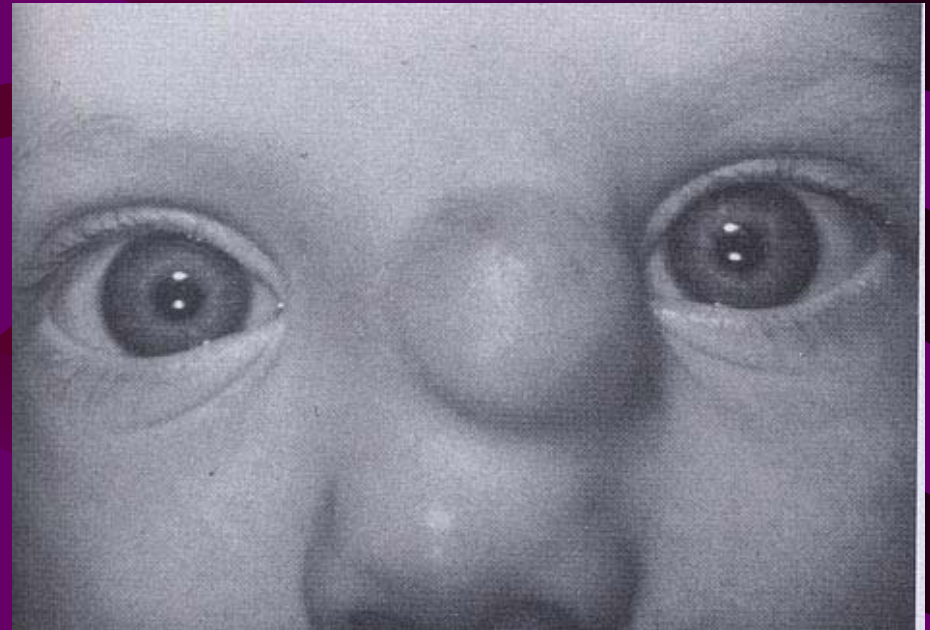
Development

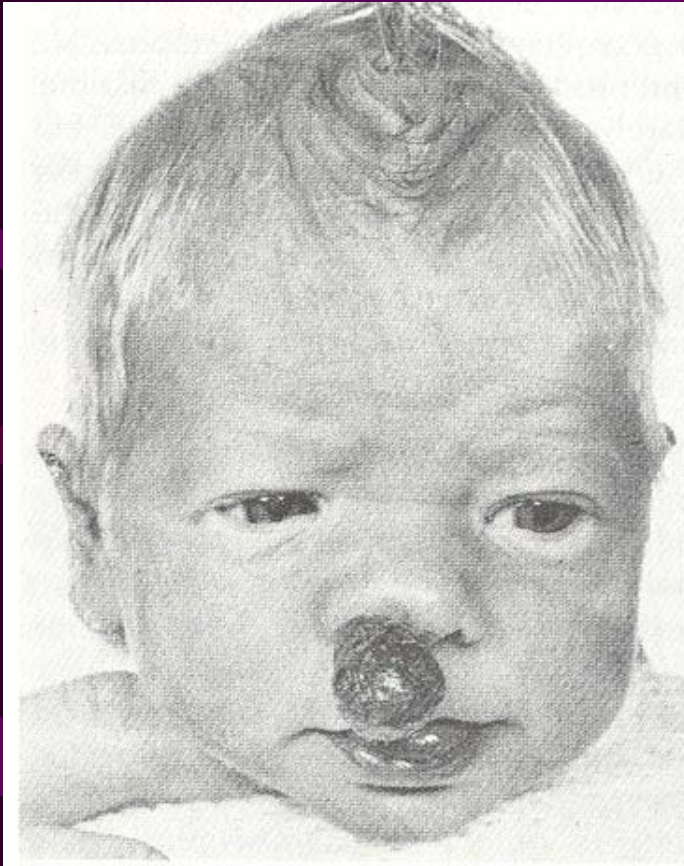
- During development a projection of dura projects through the foramen cecum
- If skin maintains attachment to underlying fibrous tissue, nasal capsule, or dura epithelial elements may be pulled into the prenasal space with or without dural connection



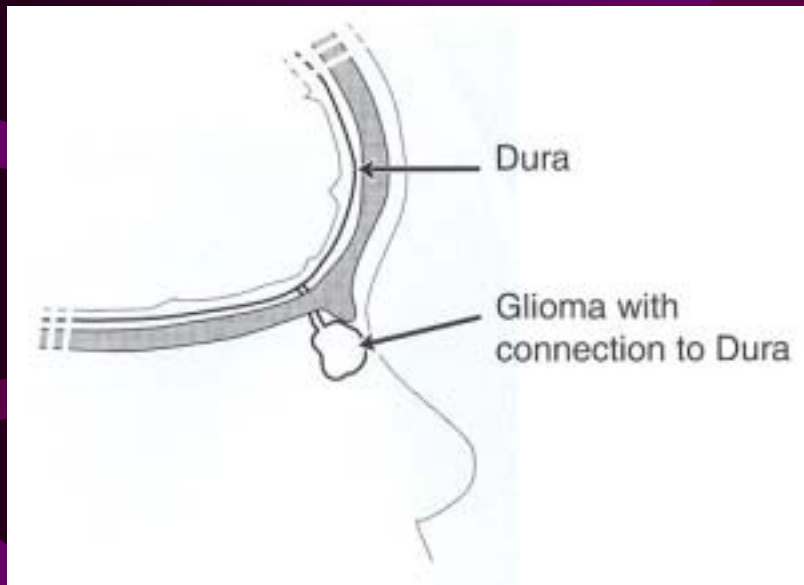
Gliomas

- Glial cells in a connective tissue matrix
- Red or bluish lump
- Glabella, nasomaxillary suture, intranasal
- Firm, noncompressible
- Do not enlarge with crying
- Do not transilluminate
- May have telangiectasias





- Extranasal 60%
- Intranasal 30%
- Both 10%
- Dural connection 35% intranasal, 9% extranasal
- Overall 15% dural connection
- CSF rhinorrhea, meningitis



- Develop from extracranial rests of glial tissue
- Abnormal closure of fonticulus nasofrontalis, possibly encephaloceles which have lost CSF connection

Encephaloceles

- Extracranial herniation of meninges and/or brain
- Subarachnoid connection
- Rare at 1:35,000 births
- 1:6000 live births in Southeast Asia and Russia
- 30-40% associated anomalies: microcephaly, hydrocephalus, microphthalmia, anophthalmia, agenesis of the corpus callosum, porencephaly, cortical atrophy, ventricular dilations



- Bluish, soft, compressible, transilluminate, pulsatile
- Enlarge with crying
- Positive Furstenberg test (bilateral compression of internal jugular veins)
- Originate medially in the nose
- May have associated CSF leak



- Divided into three categories:
- Occipital 75%
- Sincipital 15%
- Basal 10%
- Sincipital-dorsum of nose, orbits, forehead
- Basal- intranasal mass, nasopharynx, posterior orbit

Sincipital encephaloceles



- Nasofrontal
- Nasoethmoidal
- Nasoorbital

Basal Encephaloceles

- Transethmoidal-through cribriform plate into middle meatus
- Sphenoethmoidal-extends through cranial defect between posterior ethmoids and sphenoid to nasopharynx
- Transsphenoidal-presents in nasopharynx
- Sphenomaxillary- through superior and inferior orbital fissures to sphenomaxillary fossa

Development

- Dural projection through fonticulus nasofrontalis
- Abnormal closure results in herniated meninges/brain
- May be closely related to glioma

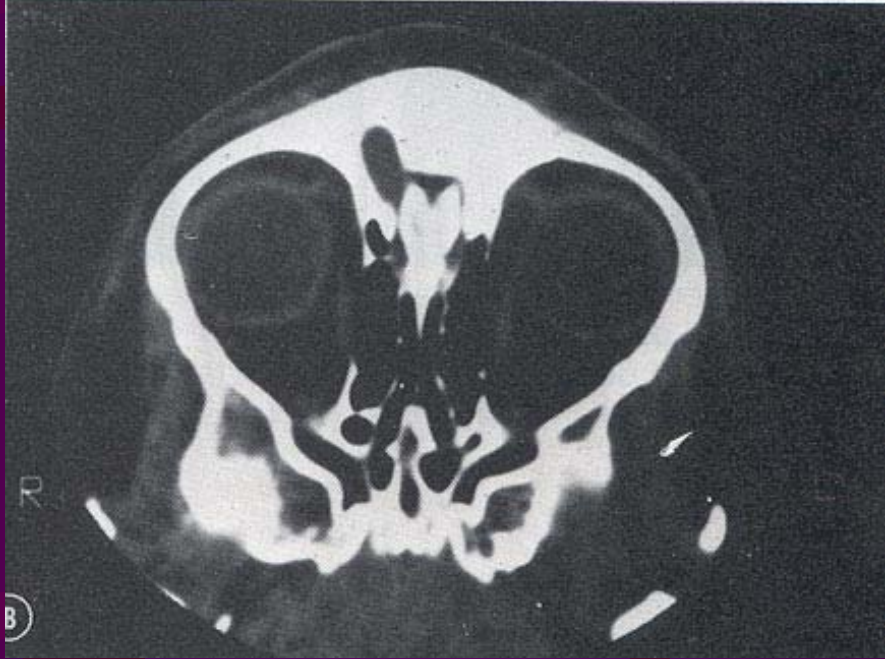
Evaluation

- Most often infants and children
- Dermoids-fistula tract, hair, pus or sebum, midline
- Gliomas- firm, noncompressible, does not transilluminate, telangiectasias
- Encephaloceles- soft, compressible, bluish or red, enlarge with crying, positive Furstenburg test

- Do not biopsy extra or intranasal mass in a child before imaging
- Risk of meningitis or CSF leak
- Get imaging before biopsy

Imaging

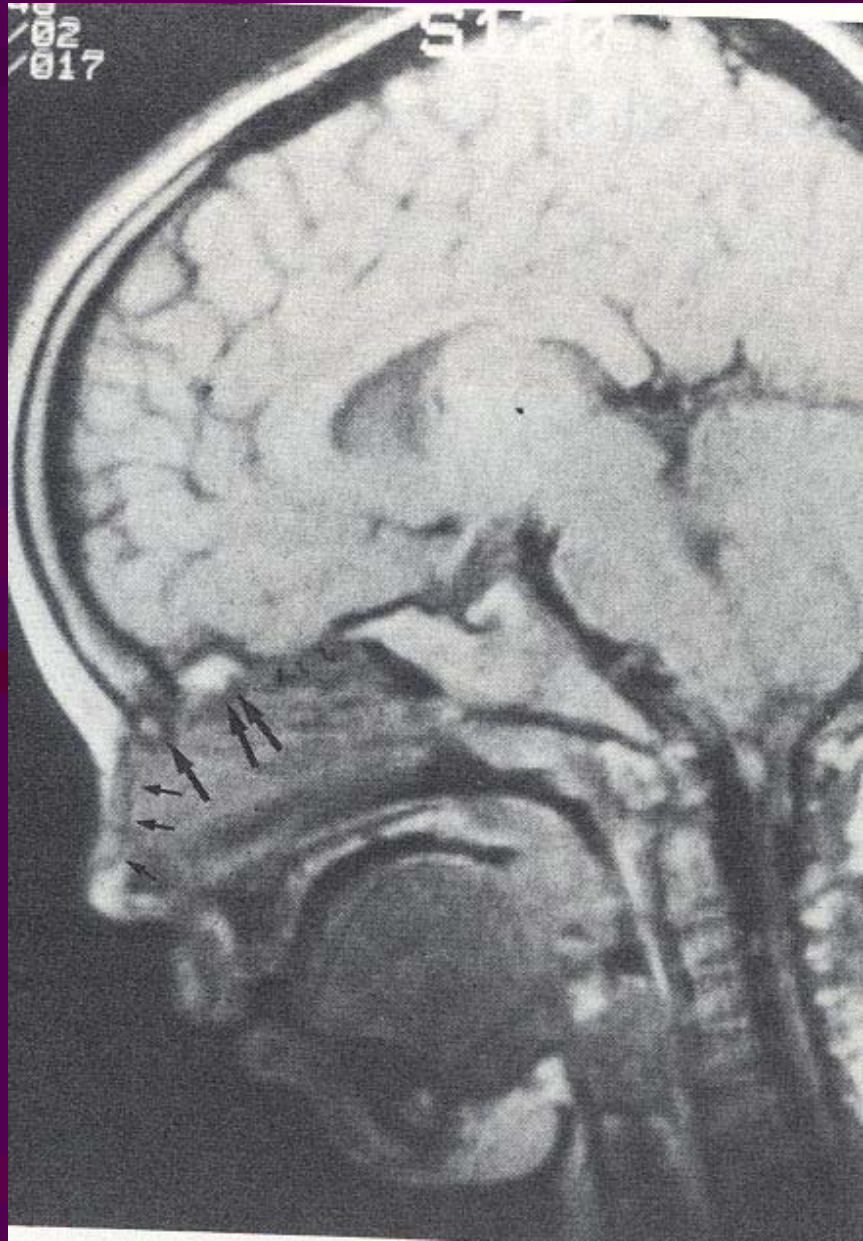
- CT and MRI most used
- CT findings include: fluid filled cyst, soft tissue mass, intracranial mass, enlargement of foramen cecum, distortion of crista galli
- CT findings suggestive of intracranial extension are enlarged foramen cecum and bifidity of crista galli
- Pensler et al reported that these findings were only valuable if absent, (when present may be false positive)



MRI

- Better delineates soft tissue
- Ability to visualize in the sagittal plane
- Denoyelle 36 children with dermoids, 2 patients had CT suggestive of intracranial involvement not found at surgery.
- Recommended CT followed by MRI to confirm intracranial connection

- Bloom et al
- 10 patients with nasal dermoids
- One false positive, one indeterminate CT
- Recommends MRI as first-line test due to increase cost of multiple tests, delay in diagnosis, additional risk if anesthesia required



Surgical Treatment

- Complete excision
- Perform early to avoid nasal distortion, bony atrophy, osteomyelitis, meningitis
- Denoyelle et al reported a recurrence rate of 5.5% of dermoids-must excise entire tract

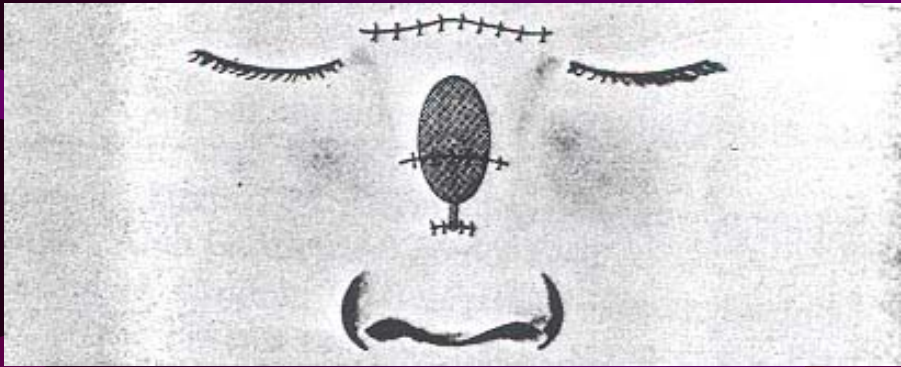
Derموids

- Pollock reviewed surgical treatment:
- Access to all midline cysts and ability to perform medial and lateral osteotomies
- Exposure for repair of cribriform defects, permit control of CSF rhinorrhea
- Allow reconstruction of nasal dorsum
- Offer probability of a favorable scar

Transverse rhinotomy

- Small to moderately sized lesions
- Avoids vertical scar and splaying

Transverse Rhinotomy



- Fistulous opening excised with transverse fusiform incision
- Tract cannulated with lacrimal probe
- Second incision made over dermoid

Tripod-eversion rhinotomy

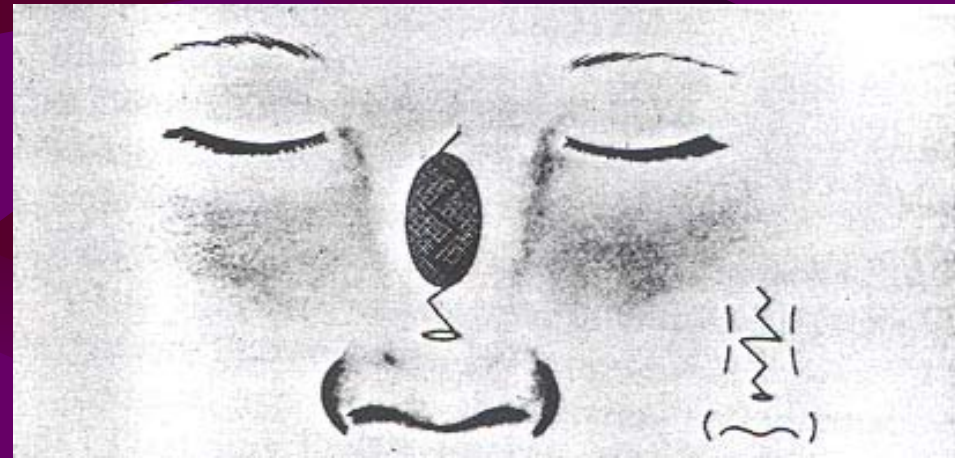


- Larger lesions, especially lower 2/3 of nose
- Transverse columellar incision, transfixion incision carried laterally and between upper and lower lateral cartilages

- Paraalar incisions will permit upward rotation of the nose
- Tract opening-fusiform excision
- Cannulate fistulous tract
- Operating microscope may be used to improve visualization

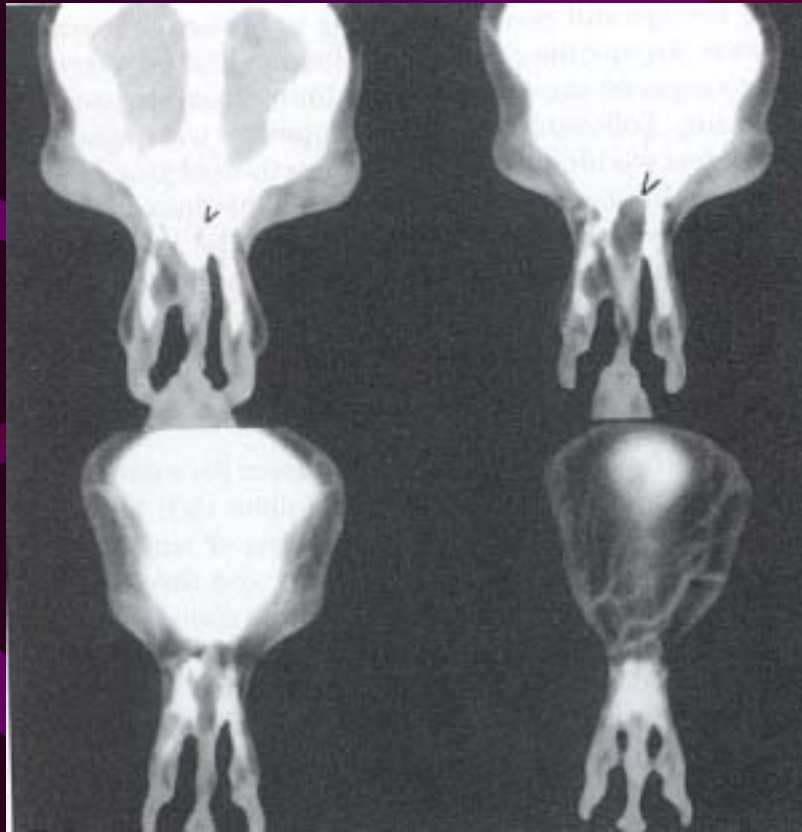
Zig-Zag rhinotomy

- Large lesions
- Underlying bone or cartilage damage
- Known intracranial extension
- Provides wide exposure
- Scar improved over straight vertical rhinotomy



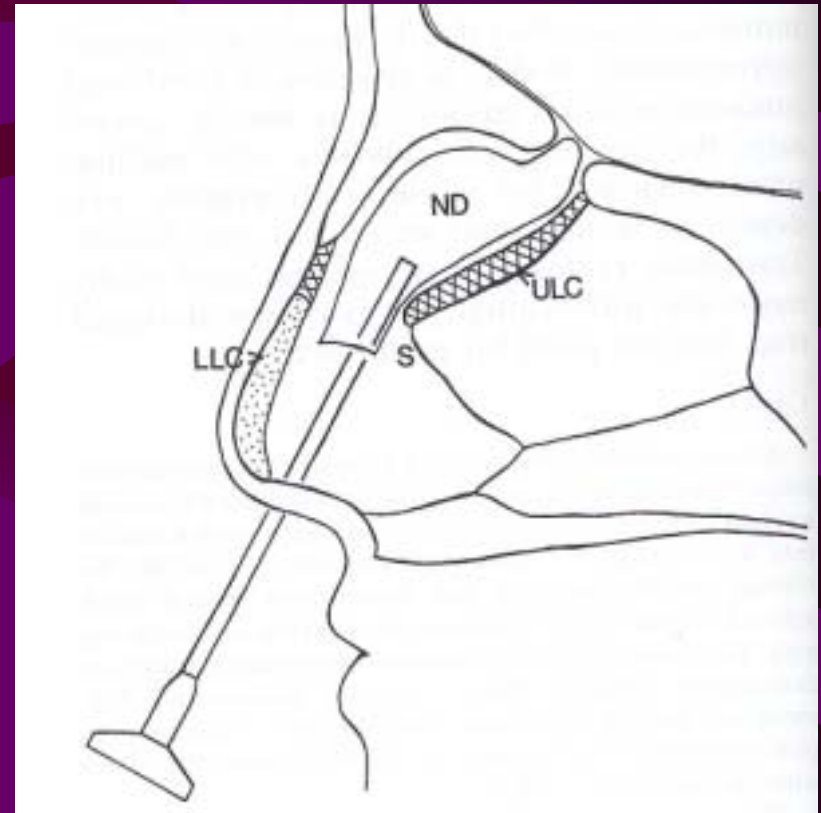
- Incisions designed with limbs which extend vertically >40 degrees <90 degrees
- Fusiform excision of fistulous tract opening
- Scar improved over straight vertical rhinotomy, limbs less than 90 degrees to RSTL running horizontally across the nose

- Rohrich recommends open rhinoplasty for the following reasons:
- Improved aesthetic results
- Ease of exposure
- Wide exposure of entire nasal dorsum
- More control over osteotomy
- Better visualization of the cribriform plate



- Weiss et al described two cases of endoscopic dermoid excision in lesions with little or no skin involvement

- Inferior portion removed via bilateral intercartilaginous and membranous septum incision
- Aufricht retractor in place 0 and 30 telescope sinus tract followed to defect



- Lateral rhinotomy may be used for intranasal gliomas or combine intra-extranasal
- Several authors have reported isolated cases of endoscopic excision of small gliomas without evidence of intracranial extension.

Encephaloceles

- Will require combined approach with neurosurgery
- Frontal craniotomy is performed, intracranial mass excised, bone-dura defect is repaired
- Extracranial mass is then removed
- Turgut et al reported mortality of 46% when there is brain tissue in the encephalocele

Key Points

- Midline nasal masses are rare but must be remembered in the differential
- Don't biopsy without imaging
- Furstenberg's test

