Big Heads, Lumps and Bumps: Hydrocephalus and Skull Lesions in the Pediatric Population

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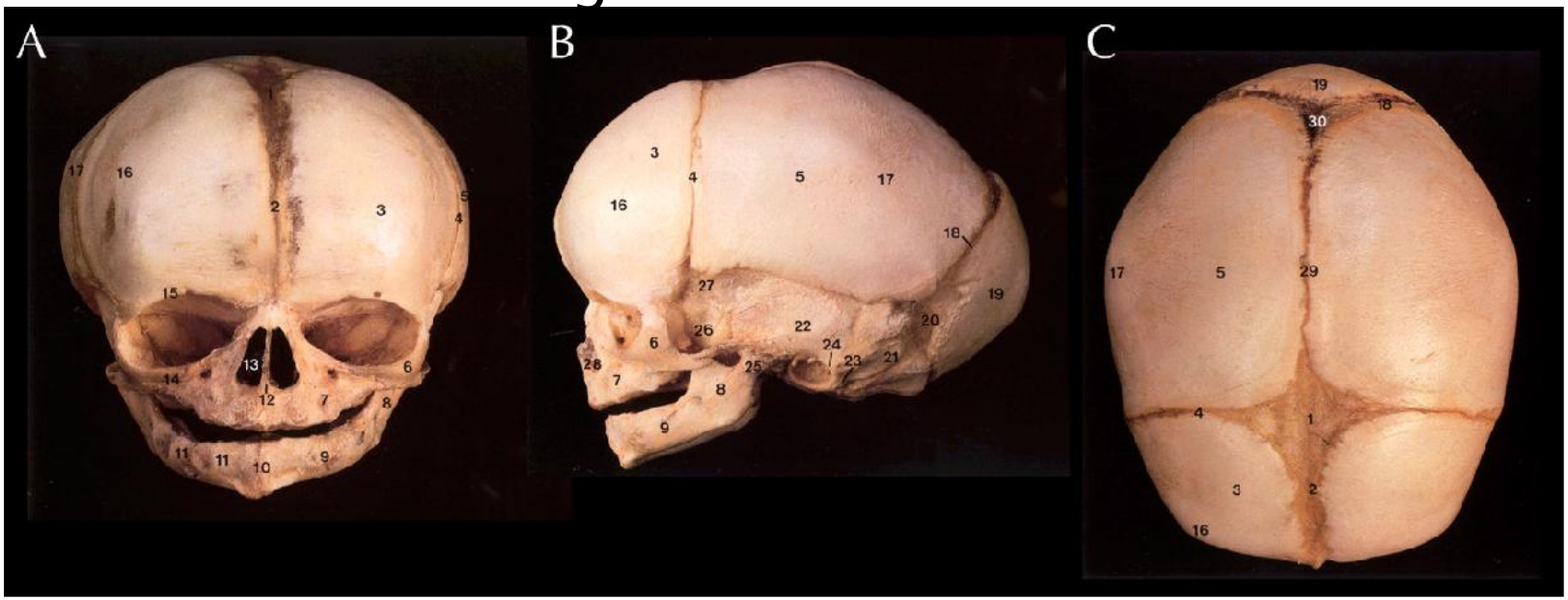
December 8th, 2010

Viscoelastic Properties of the Immature Cranium

- Open fontanels and sutures allow cranial expansion
- High water content allow compensatory ventricular enlargement

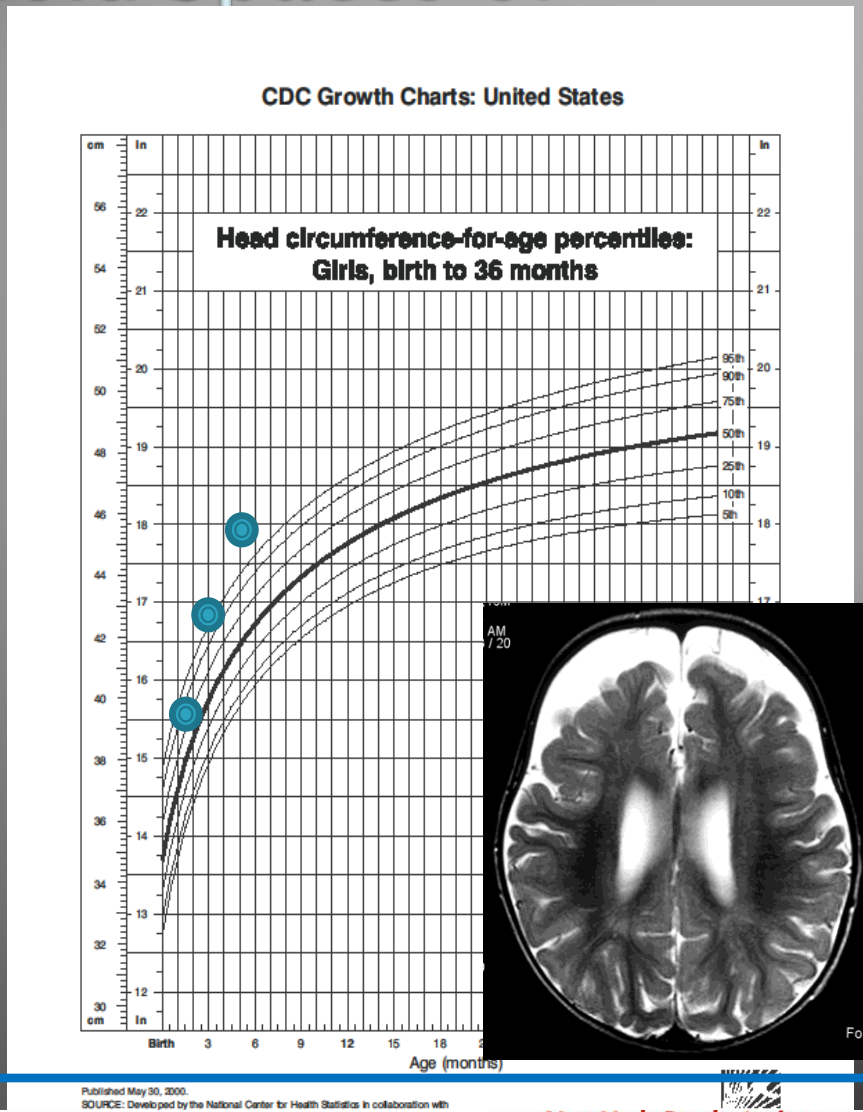


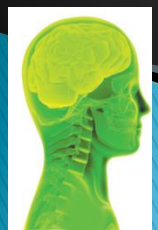
Macrocephaly: Clinically Insignificant

- Constitutional macrocephaly
  - 5% of 'normal' children have an occipitofrontal circumference (OFC) greater than the 95 percentile
  - Nondivergent growth curve
- Increased subarachnoid spaces of infancy
  - External hydrocephalus
  - Divergent growth curve

Increased Subarachnoid Spaces of Infancy

- 6 month old female
- Normal birth and development history
- Growth curve as shown
- Open and full anterior fontanel
- No suture diastasis





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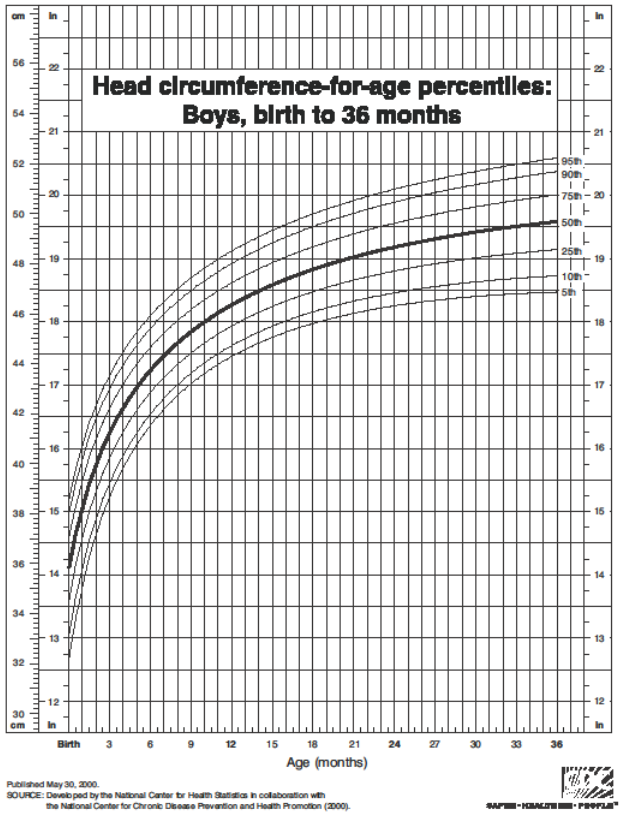
OFC Measurements

CDC Growth Charts: United States



Published May 30, 2000.
SOURCE: Developed by the National Center for Health Statistics in collaboration with the National Center for Chronic Disease Prevention and Health Promotion (2000).

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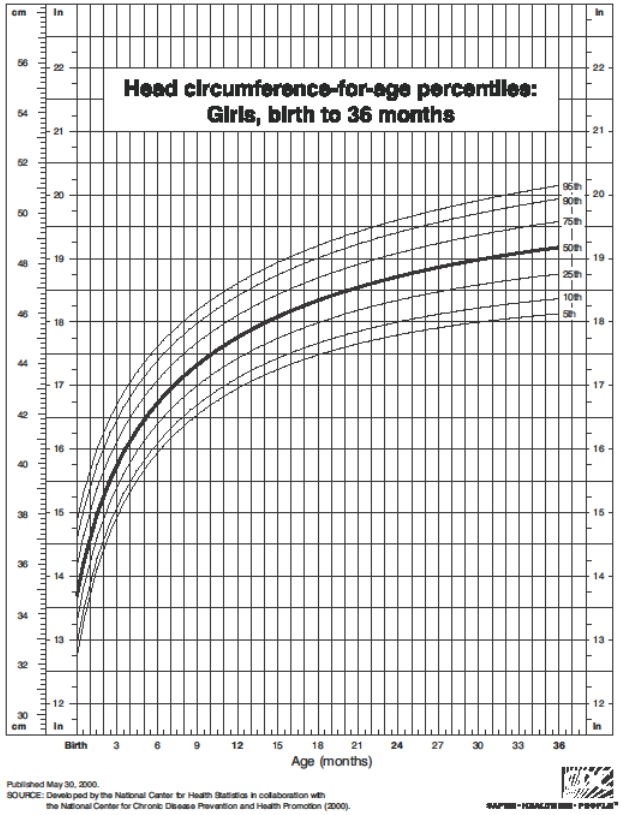

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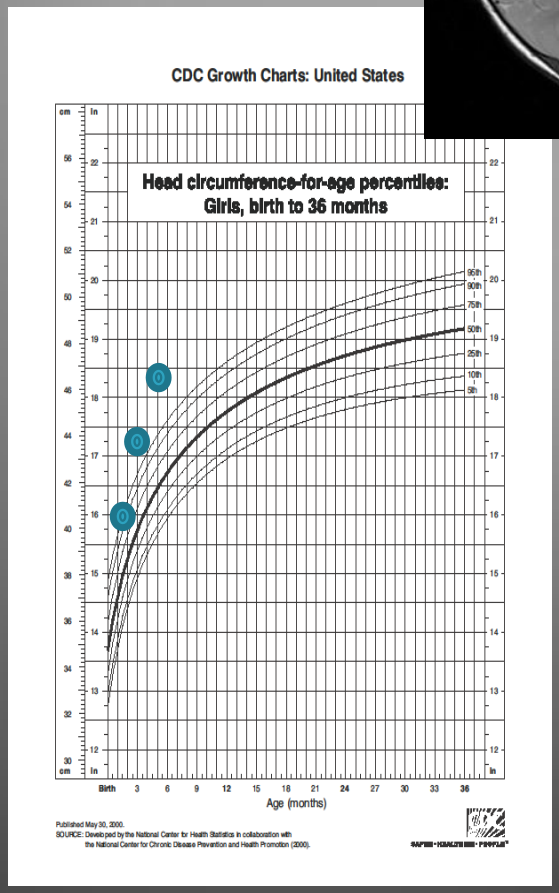
Macrocephaly: Clinically Significant

- Chronic subdural hematomas of infancy
  - Divergent growth curve
  - Usually nonaccidental trauma
- Hydrocephalus and other CSF circulation abnormalities
  - Divergent growth curve
- Neurodegenerative disorders (Canavans and Alexander disease)
  - Divergent growth curve

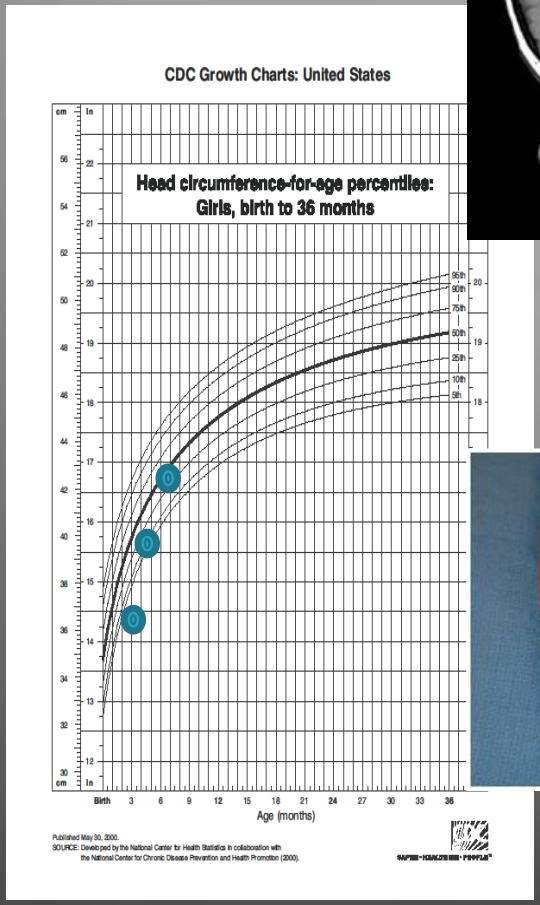
Chronic Subdurals

- 6 month old female
- Normal birth and development history
- Growth curve as shown
- Open and full anterior fontanel
- Suture diastasis
- Irritability, vomiting, seizures, etc.



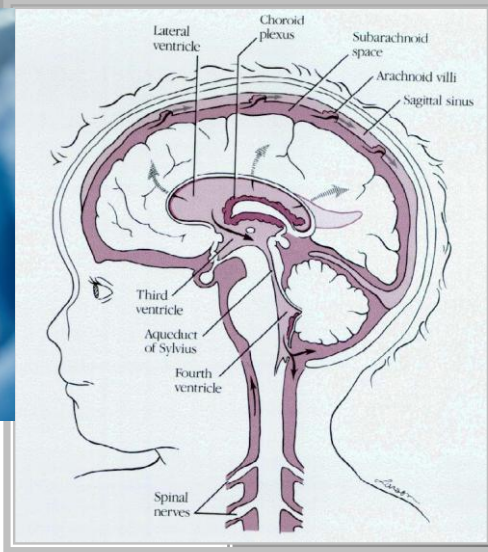
Alterations in CSF Circulation

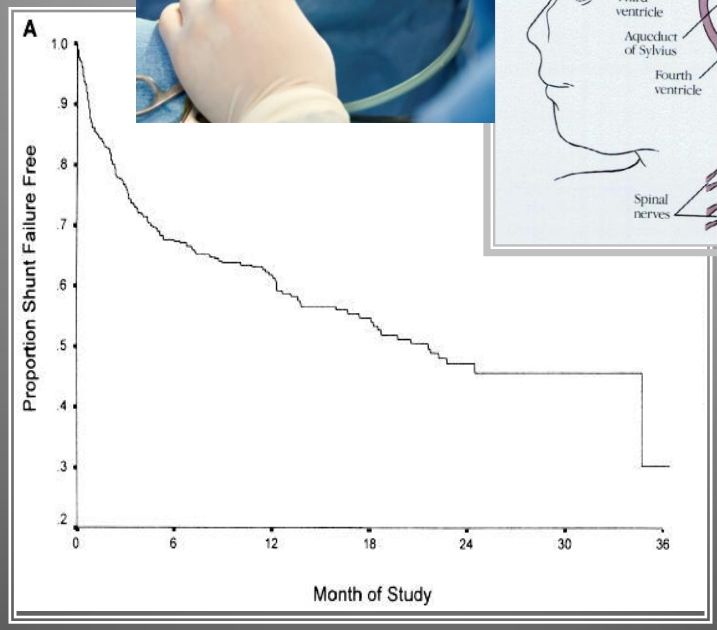
- 6 month old female
- Normal birth and development history
- Growth curve as shown
- Open and full anterior fontanel
- Suture diastasis
- Irritability, vomiting, seizures, etc.
- Clinical manifestations of raised ICP
  - Frontal bossing
  - Forced down gaze
  - Engorgement of scalp veins



Endoscopic Third Ventriculostomy

- Alternative to shunt placement
- Avoids all complications associated with shunts
- Restoration of normal CSF outflow and resorption
- Applicable in all forms of intraventricular obstructive hydrocephalus distal to the posterior third ventricle





Drake JM, Kestle JR, Milner R, et al: Randomized trial of cerebrospinal fluid shunt valve design in pediatric hydrocephalus. Neurosurgery 43:294-305, 1998.

Scalp and Skull Masses

- Posttraumatic
  - Cephalohematoma
  - Leptomeningeal cyst
- Neoplastic
  - neurofibroma
- Inflammatory
  - Lymphadenopathy
  - LCH
  - Cranial fasciitis
  - Infantile myofibormatosis
- Congenital
  - Inclusion cysts
  - Aplasia cutis congenita
- Vascular
  - Capillary hemangioma
  - Sinus pericranii
  - AV malformation
- Dysraphic
  - Cephaloceles
    - Atretic
    - Encephalocele
    - meningocele

Office Evaluation

- History
  - Rate of growth
  - History of trauma including birth history
  - Fevers or associated illnesses
- Physical Examination
  - Anatomical position (midline or eccentric, supratentorial or infratentorial)
  - Pain on palpation
  - Skin integrity
  - Pulsatile
  - Intracranial pressure transmission
  - Associated lymphadenopathy
- Imaging decision

Scalp & Skull Masses

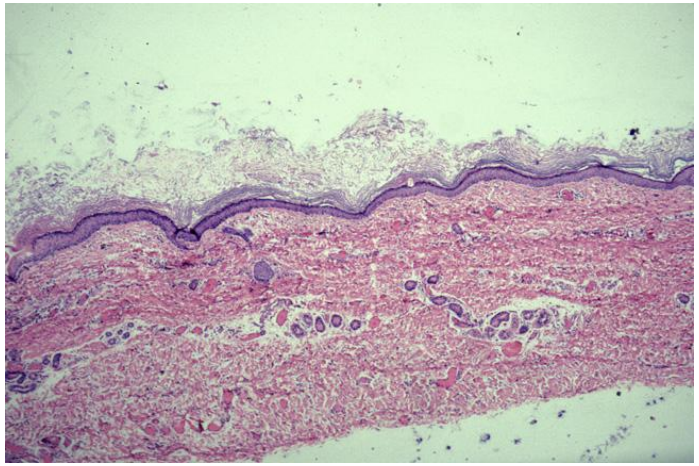
- Generalizations
  - Malignancy is rare
  - Intracrnaial extension is rare
  - Diagnosis is seldom in question
  - Hetereogeneous pathology
    1. Inclusion cysts (dermoid cysts, epidermoid cysts)
    2. Langerhan's cell histiocytosis (eosinophilic granuloma)
    3. Cephaloceles
    4. Subperiosteal hematoma
    5. Lepotomeningeal cyst

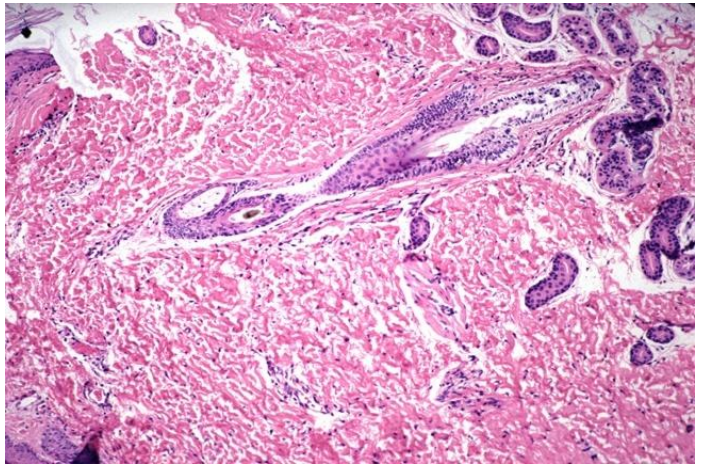
Inclusion Cysts

- Congenital disorder
  - Failed dysjunction
- Epidermoid cyst
  - Intracranial
  - Older children/adults
  - Rare association with dermal sinus
- Dermoid cyst
  - Extracranial
  - Infants
  - Can be associated with dermal sinus (meningitedes)

Dermoid cyst

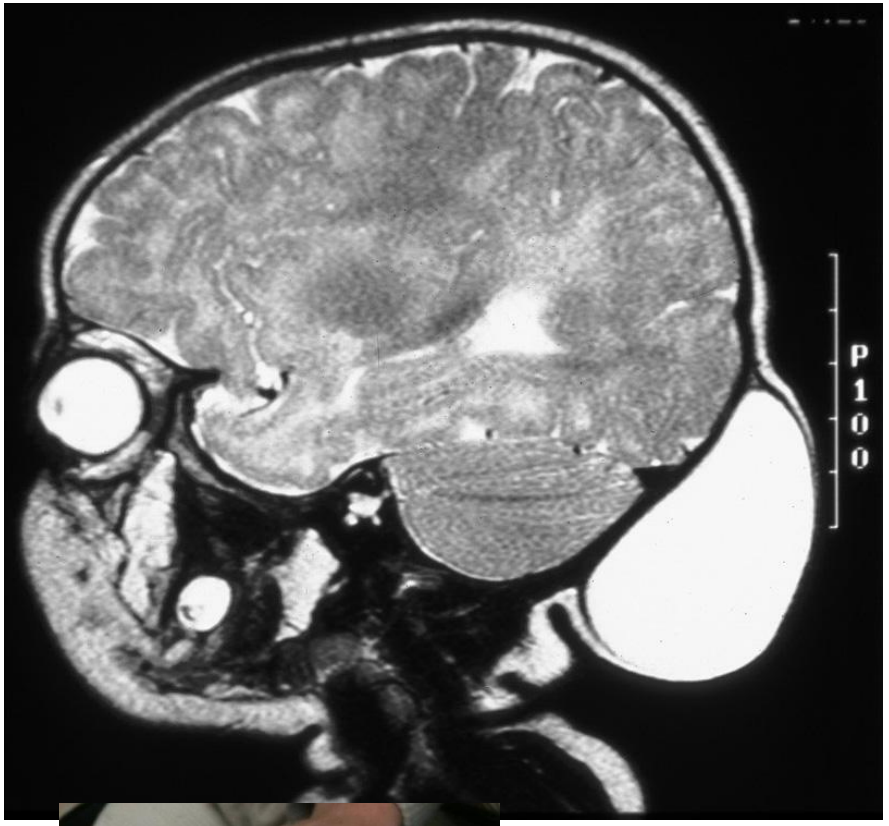
- Most common scalp mass evaluated by neurosurgeon (60%)
- Usually present at birth
- Epidermal and dermal elements
- Slow growth
  - Keratin
  - Cholesteral
  - Sebaceum





Dermoid Cyst

- Anatomical location
  - Anterior fontanel
    - Rare dural penetration
  - Pterion
  - Suboccipital midline
    - Dermal sinus
    - Occipital sinus duplication
    - Dural penetration typical



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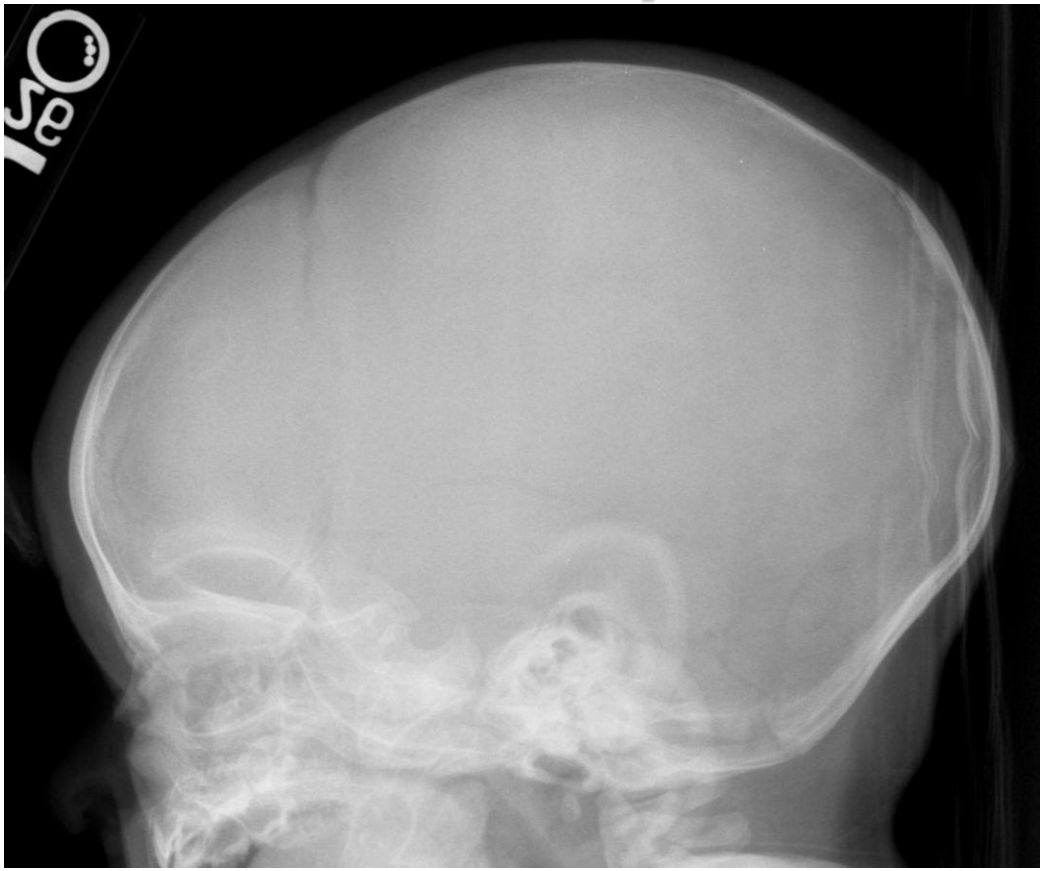
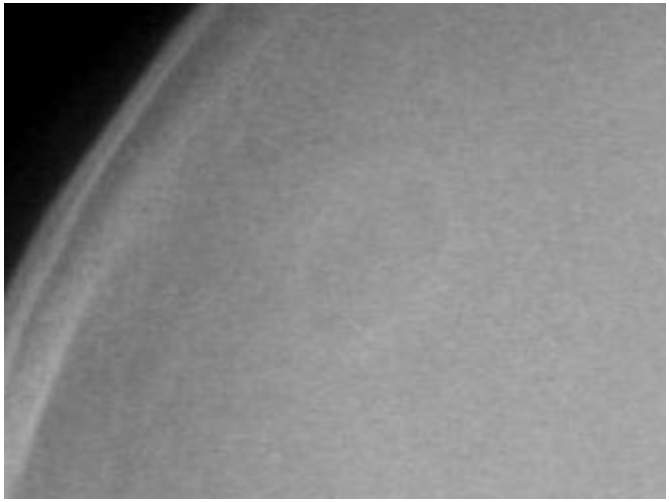
Dermoid cyst

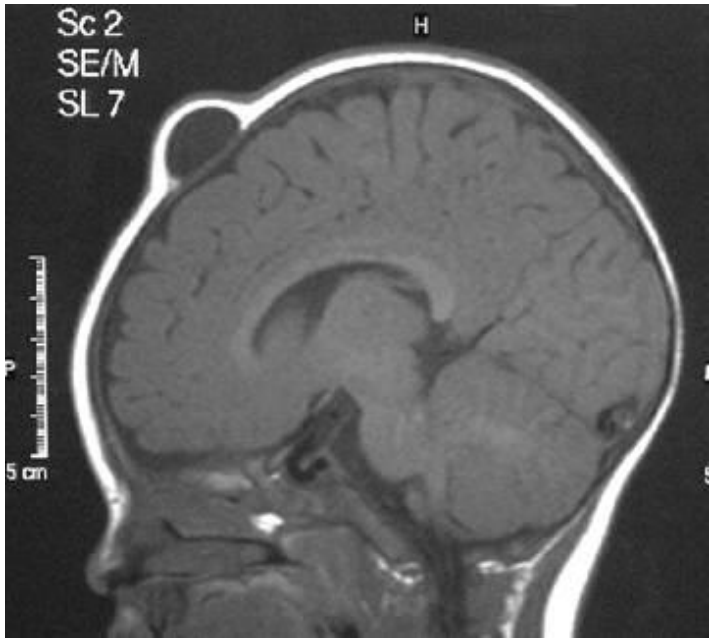


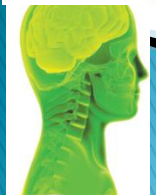
- Plain radiographs
  - Lytic
  - Smooth contours
  - Sclerotic margins



Dermoid cyst: Treatment

- Elective surgical excision
- Split thickness autologous cranioplasty > infancy





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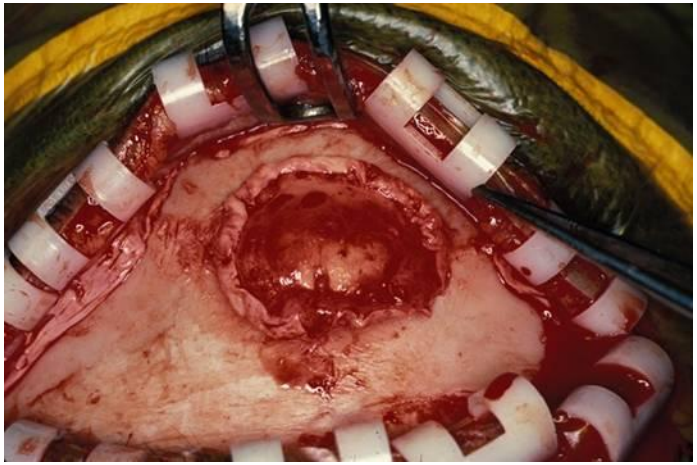
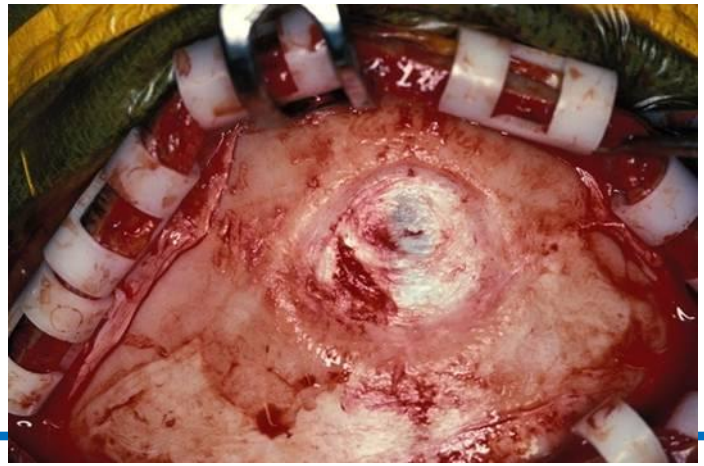
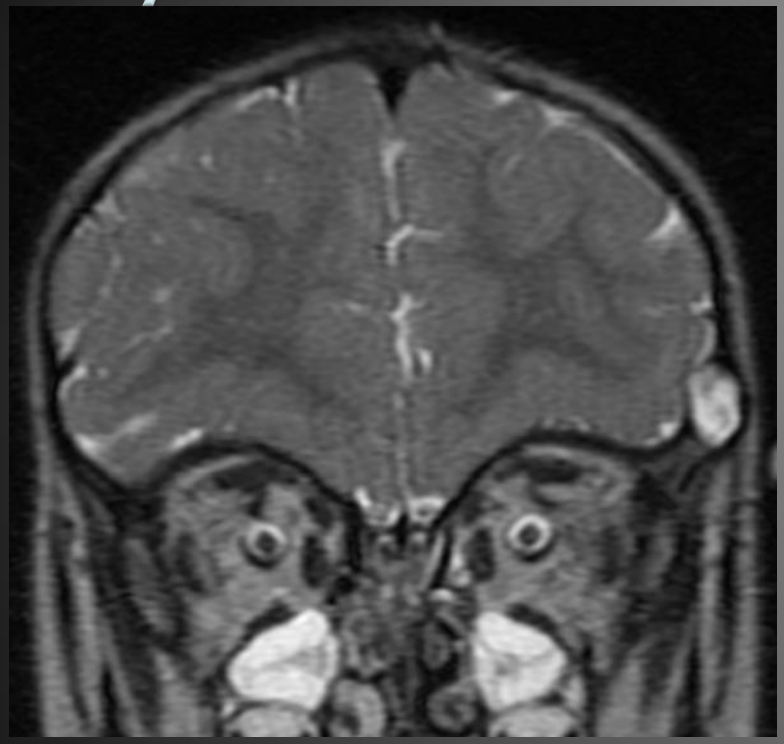
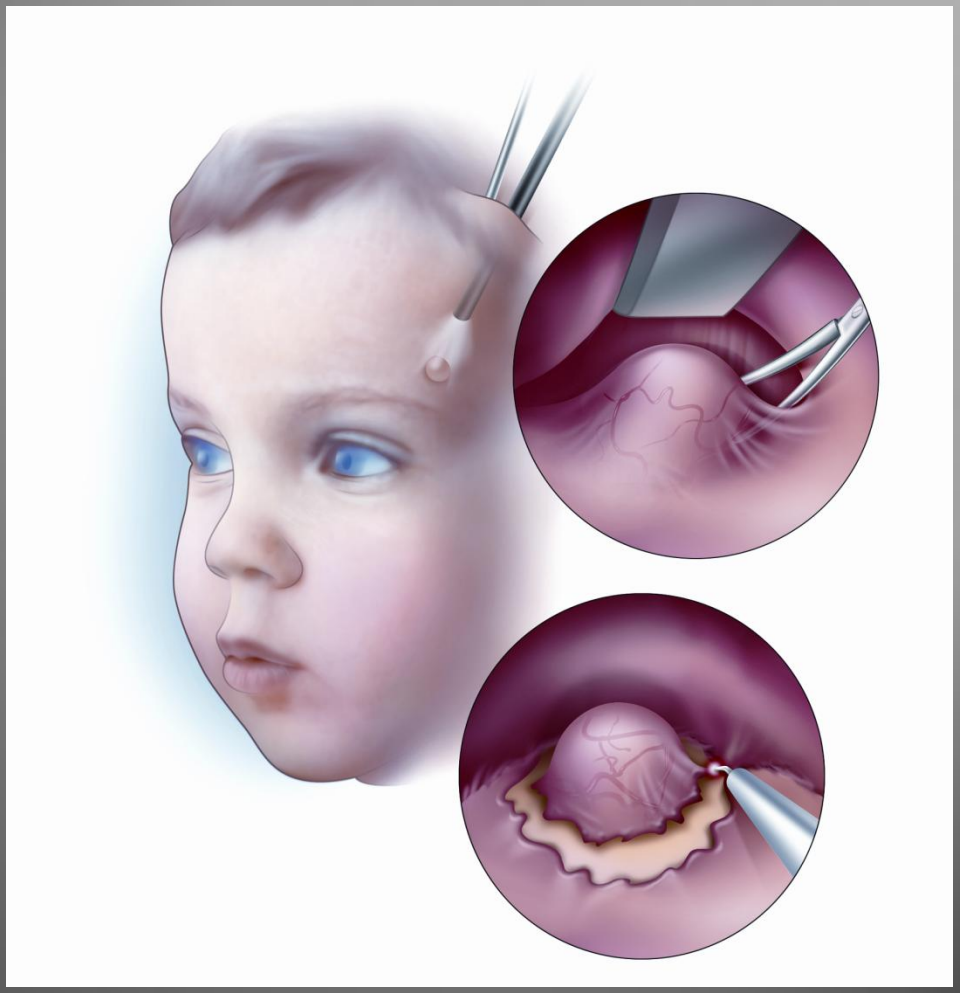
Dermoid Cyst: Surgical Excision

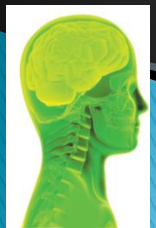




Endoscopic Removal of Dermoid Cysts







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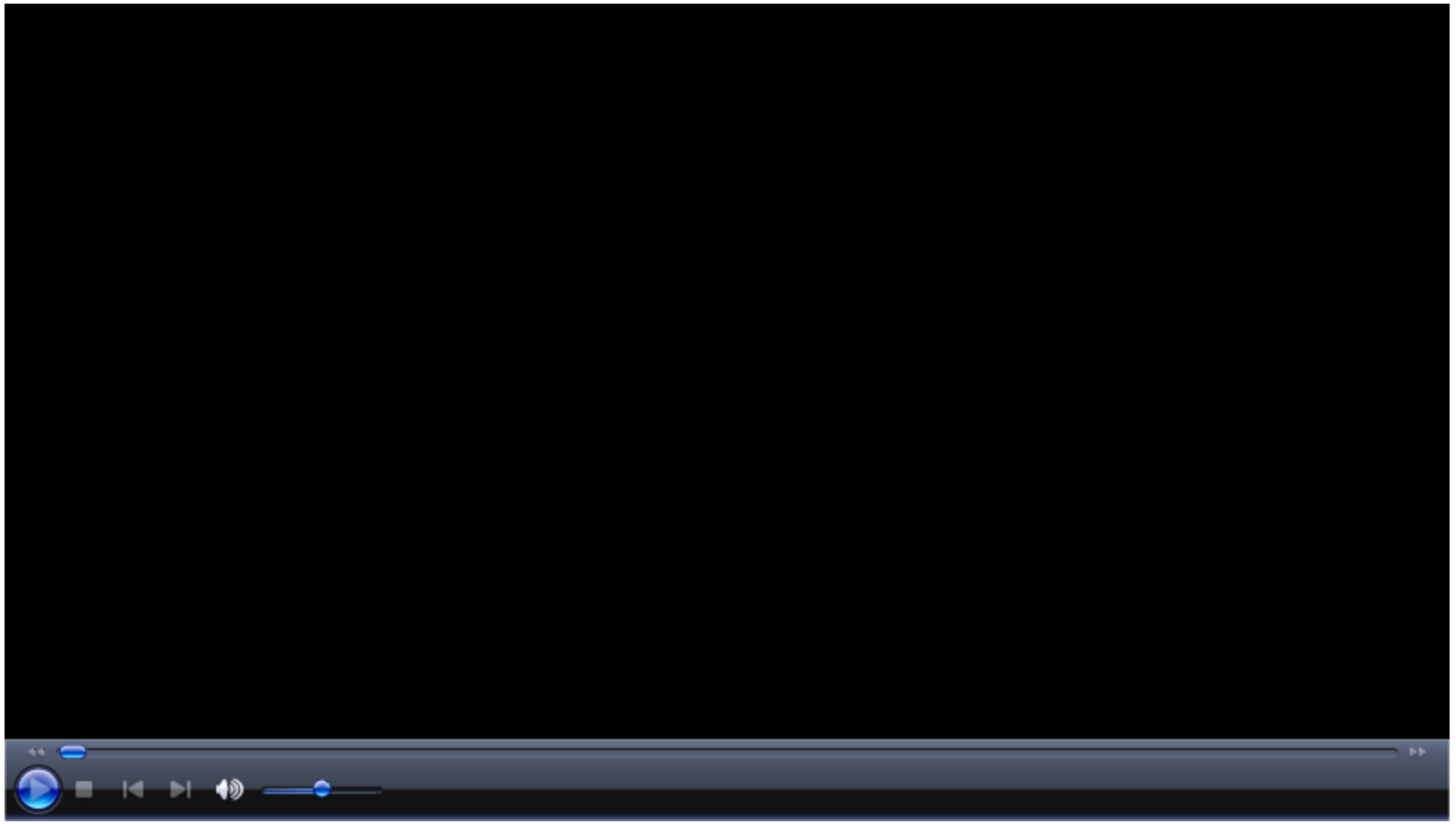
Endoscopic Removal of Dermoid Cyst



Calvarial Dermoid Cysts

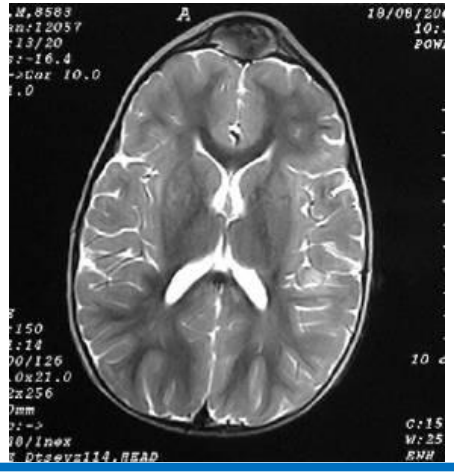
Langerhan's Cell Histiocytosis

- Histiocytosis X
  - Eosinophilic granuloma
  - Hand-Schuller-Christian Disease (diabetes insipidus, hypothalamic lesions)
  - Letterer-Siwe syndrome
- Uncertain etiology
  - Abnormal proliferation of histiocytes
- Skull is most common site
  - 28% polyostotic disease
  - Rarely intracranial
- Painful scalp mass
- Age 4-12 years

Langerhan's Cell Histiocytosis

- Painful scalp mass
- Relatively short history
- Age 4-12 years
- Radiology
  - Plain films ("punched out", no sclerotic margins, scalloped edges)
  - CT
    - External table remodeling
    - Diploic interspace

Langerhan's Cell Histiocytosis



Langerhan's Cell Histiocytosis

- Treatment
  - Surgical
    - Curettage
    - Complete excision
  - Adjunctive Rx
    - Radiation therapy
    - chemotherapy

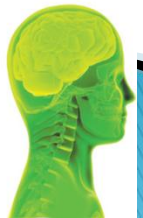
Langerhan's Cell Histiocytosis

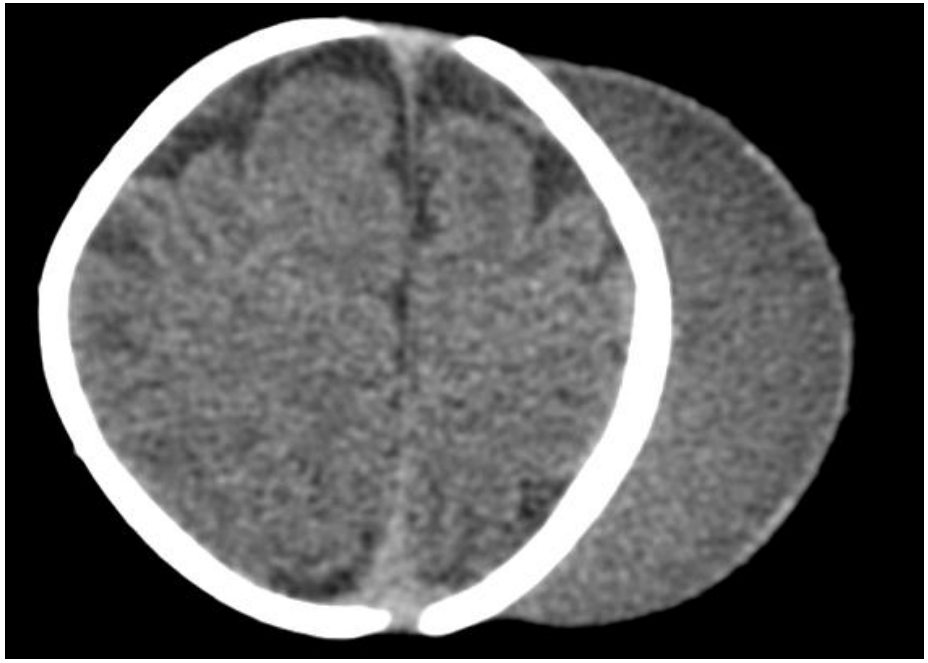
- Outcome
  - Poorly defined
  - Local recurrence 6%
  - New recurrence 22%*
- Negative prognostic features
  - < 2 years age
  - Polyostotic disease
  - Hepatosplenomegaly
  - Thrombocytopenia

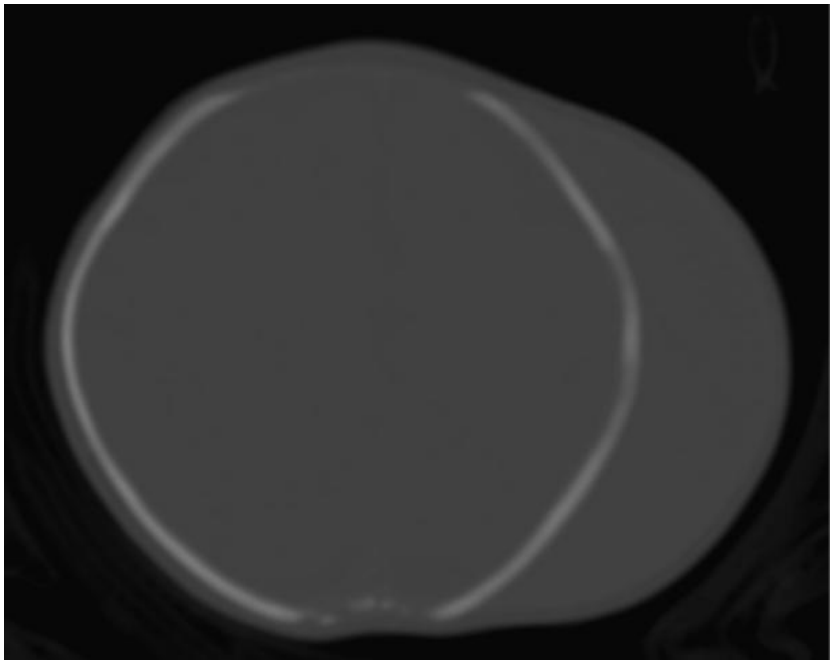
*Kilpatrick et al. Cancer 1995

Cephalohematoma

- Subperiosteal hematoma resulting from birthing (prolonged labor, forceps delivery, etc.)
- Physical examination
  - Absent intracranial extension
  - Respects suture anatomy
- Treatment/Outcome
  - Spontaneous resolution, no need for treatment or aspiration
  - Minority of subperiosteal hematomas calcify
  - cephalohematoma deformans that may require recontouring



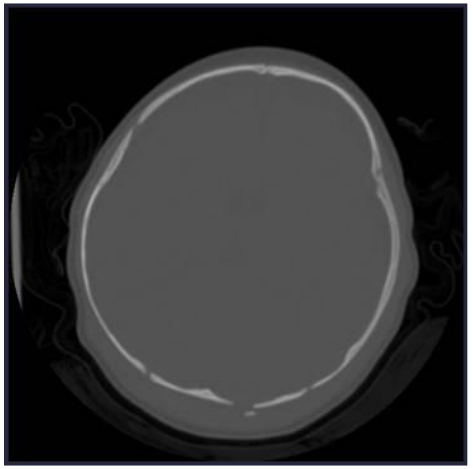




Leptomeningeal Cyst

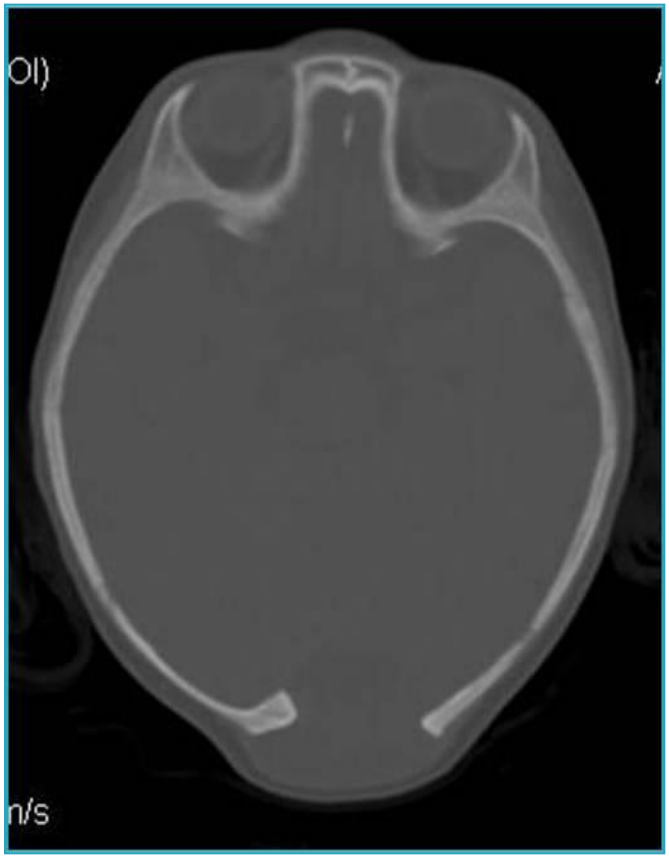
- "Growing skull fracture"
- Rare sequelae of linear skull fractures in infants
- Herniation of leptomeninges and varying cerebral tissue
- Clinical Features
  - Delayed "swelling" following trauma
  - Progressive seizure disorder or neurological deficits
  - Plain X-ray is diagnostic (widely separated linear fracture)
- Repair directed at cranial defect

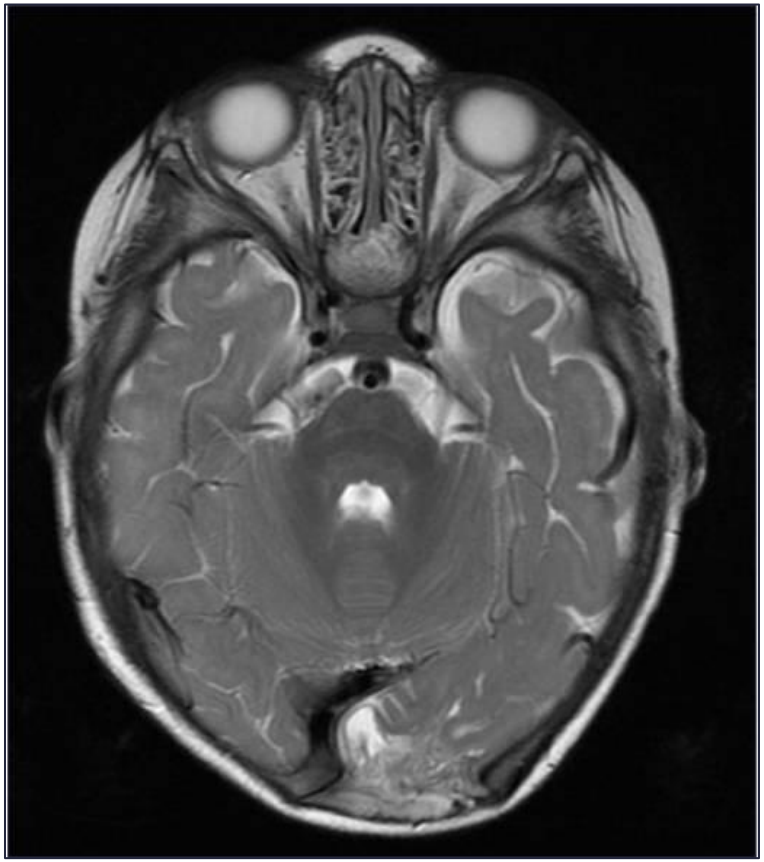
Leptomeningeal cyst



October 2007

Leptomeningeal Cyst





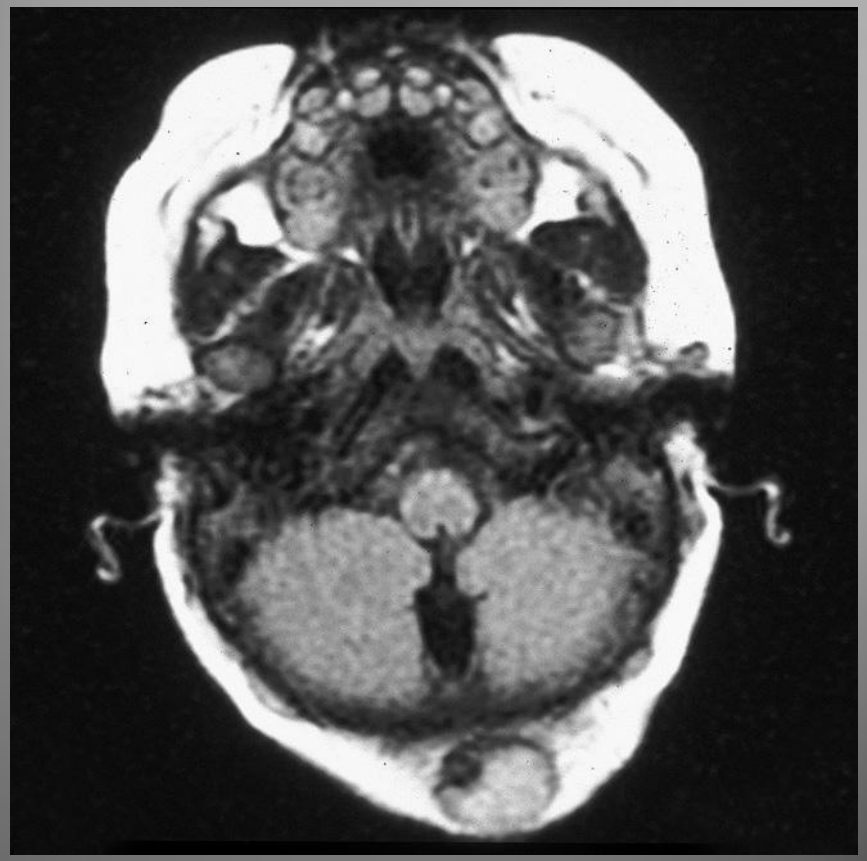
March 2008

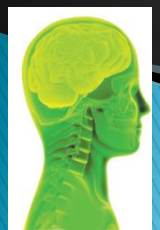
Cepahloceles

- Congenital disorders (encephaloceles, meningoceles) of nondysjunction
  - Skin
  - Calvarium
  - Meninges
  - Cerebrum
- Always midline (cranial base to craniocervical junction), transillumination
- Classification is varied
- Treatment is aimed at repair of defect
- Outcome dependent upon associated intracranial anomalies

Cepahloceles

Cepahloceles





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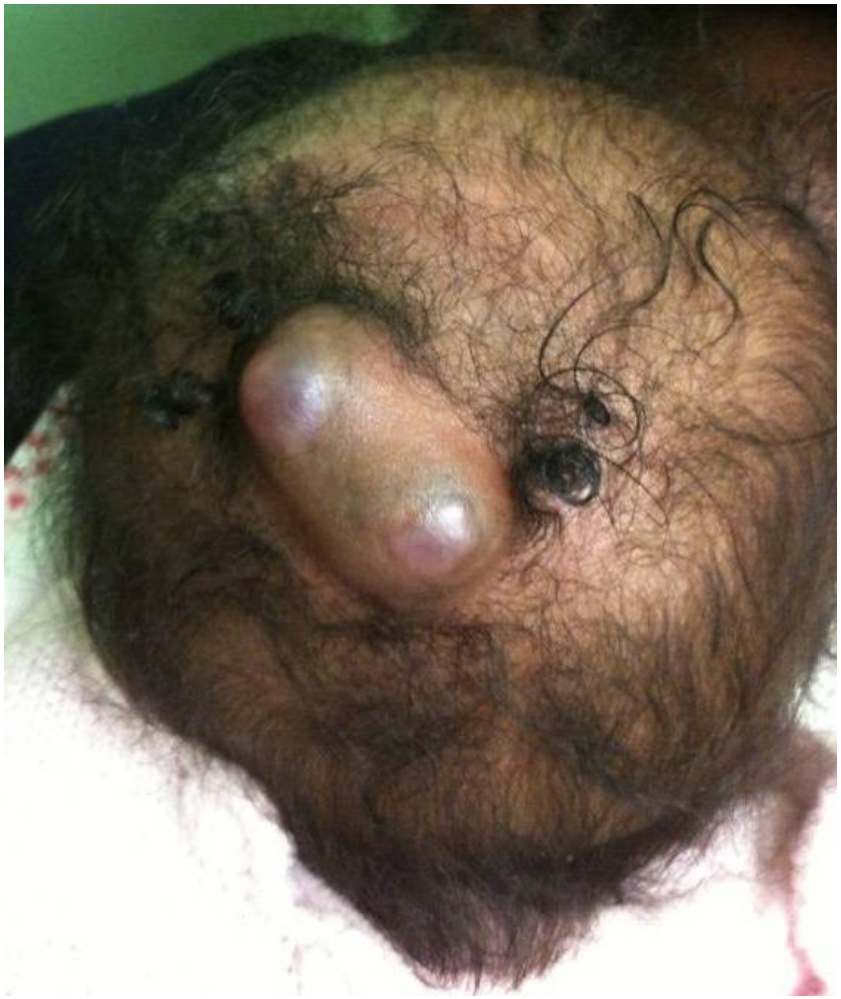
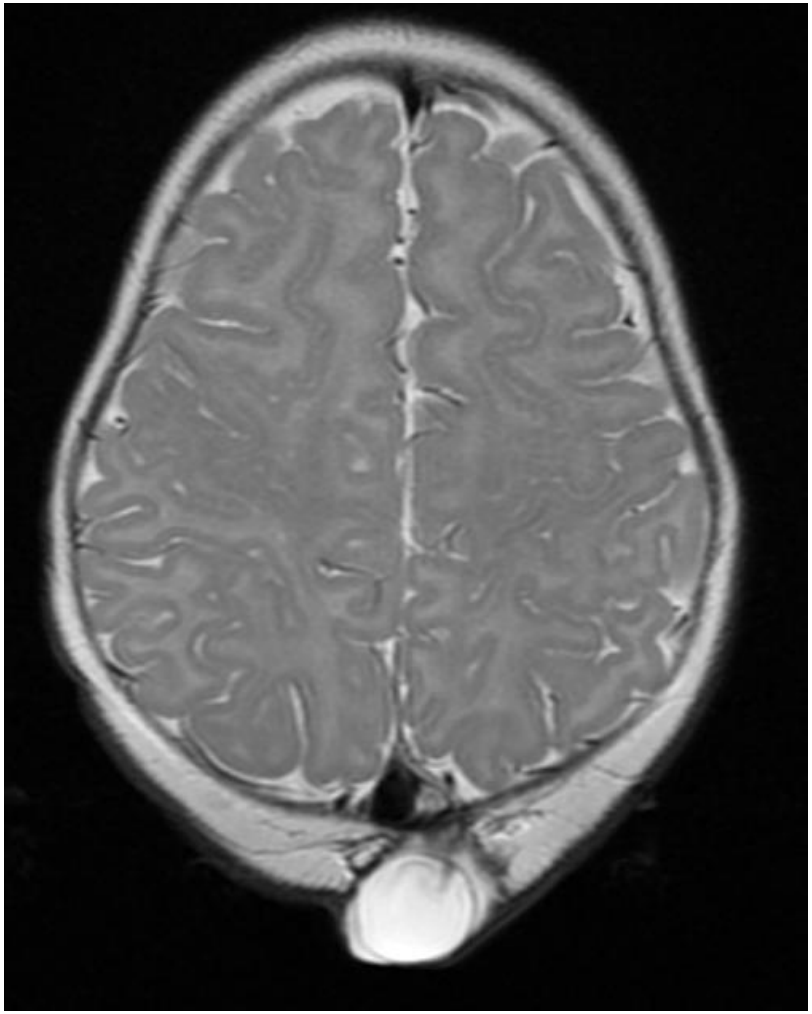


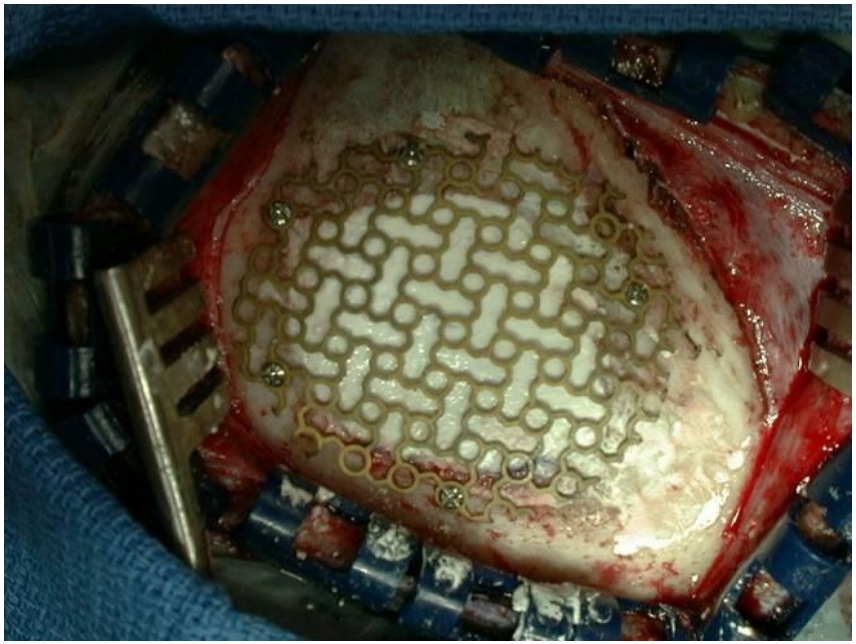


Atretic Encephalocele

- “aborted” cephaloceles
- Congenital disorder, failed dysjunction
- Typically parietal–occipital
- Skin is atypical (discolored, attenuated, hypertrichosis)
- Associated structural cerebral anomalies
- Treatment aimed at normal skin covering
- Outcome dependent upon associated cerebral anomalies

Cranioplasty

- Spontaneous osteogenesis
  - Age dependent
- Autologous cranioplasty
  - Split thickness
- Synthetic cranioplasty
  - Acrylic
  - Titanium



Question:

- 6 month old female
- Normal birth and development history
- Growth curve as shown
- Open anterior fontanel
- Appropriate management includes:
  a. Transfontanel sonogram
  b. MRI of brain
  c. Neurosurgical referral
  d. Neurology referral
  e. None of the above