Central Nervous System Congenital Abnormalities

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Neural tube defects

- **Dysraphism**
  - uncomplete neural tube closure with possible herniation

1. **Cranial dysraphism**
2. **Spinal dysraphism**

- **Total dysraphism – craniorachischisis**
  - non developed calvarium with complete spinal canal splitting (mostly abortus)

1. Cranial dysraphism

- uncomplete neural tube closure with „cranial bifidum“ (middleline calvaria defect) with possible cephalocele

- **Cephalocele**
  - **a) cranial meningocele:** dura mater and CSF herniation
  - **b) encephalocele:** cerebral tissue herniation
  - **c) anencephaly:** open dysraphism, without calvaria bones

- **Localisation**
  - middle line, frontal, parietal or occipital

- **Diagnosis**
  - X-rays skull and spine – skeleton defects
  - Ultrasound – hernia content
  - CT or MRI – detailed information

Cranial dysraphism

- Cranial meningocele mostly has a good prognosis
- Encephalocele is mostly accompanied by hydrocephalus, mikrocephaly, mental retardation, epileptic seizures

Cranial dysraphism

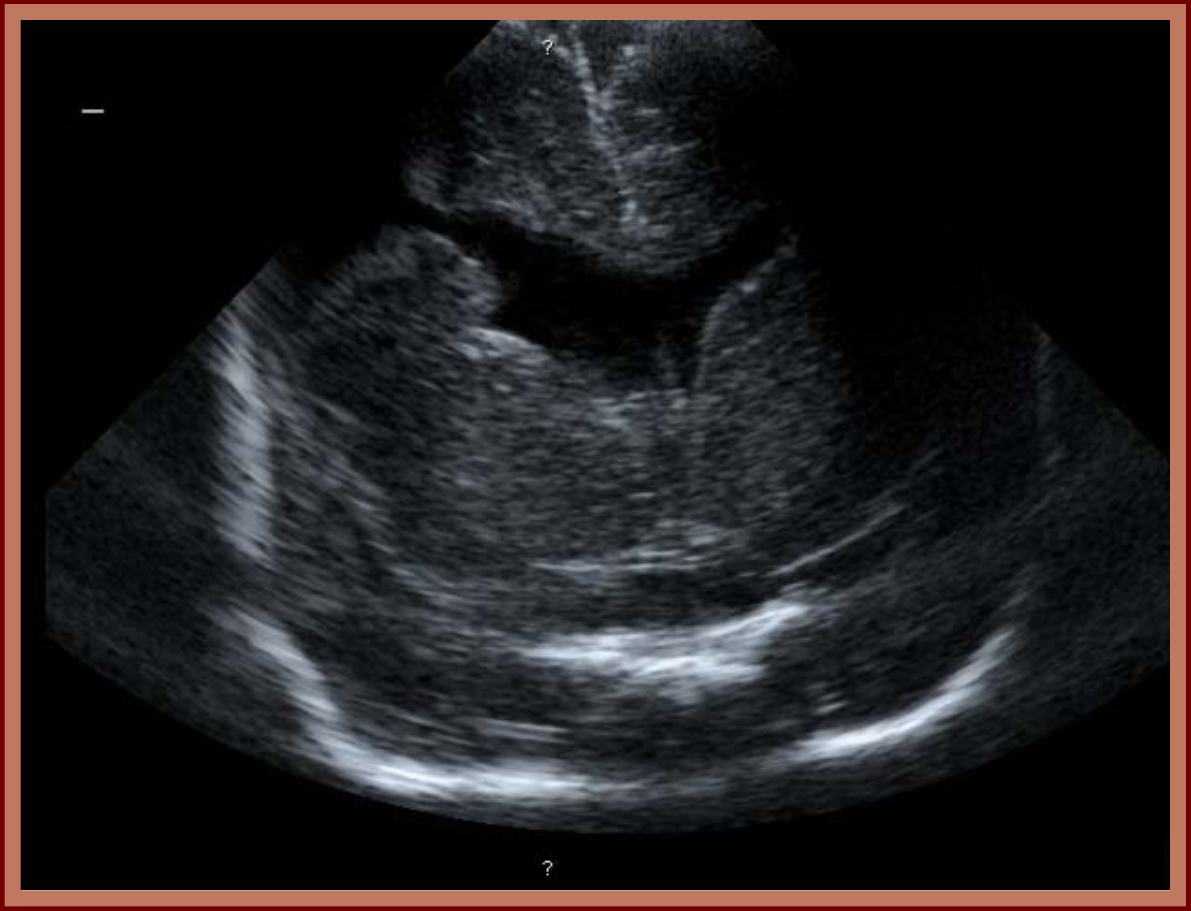
a) **Microcephalia**
b) **Hydranencephalia** – a loss of almost all cerebral tissue
c) **Holoprosencephalia** – hemispheral development disturbance
d) **Lissencephalia** – severe disturbance of neural tissue migration

- **agyria:** completely smooth cerebral surface
- **pachygyria:** few flat gyruses
- **polymicrogyria:** small gyruses, shallow sulci (similar to pachygyria)

e) Porencephalia
f) Agenesis of corpus callosum
g) Dandy-Walker syndrom (cerebellar hypoplasia)
h) Macroencephaly – megalencephaly
i) Schizencefaly

Cranial dysraphism

- Schizencefalia



2. Spinal dysraphism - spina bifida

a) **Spina bifida occulta**
   - congenital absence of processus spinosi and vertebral arches changes

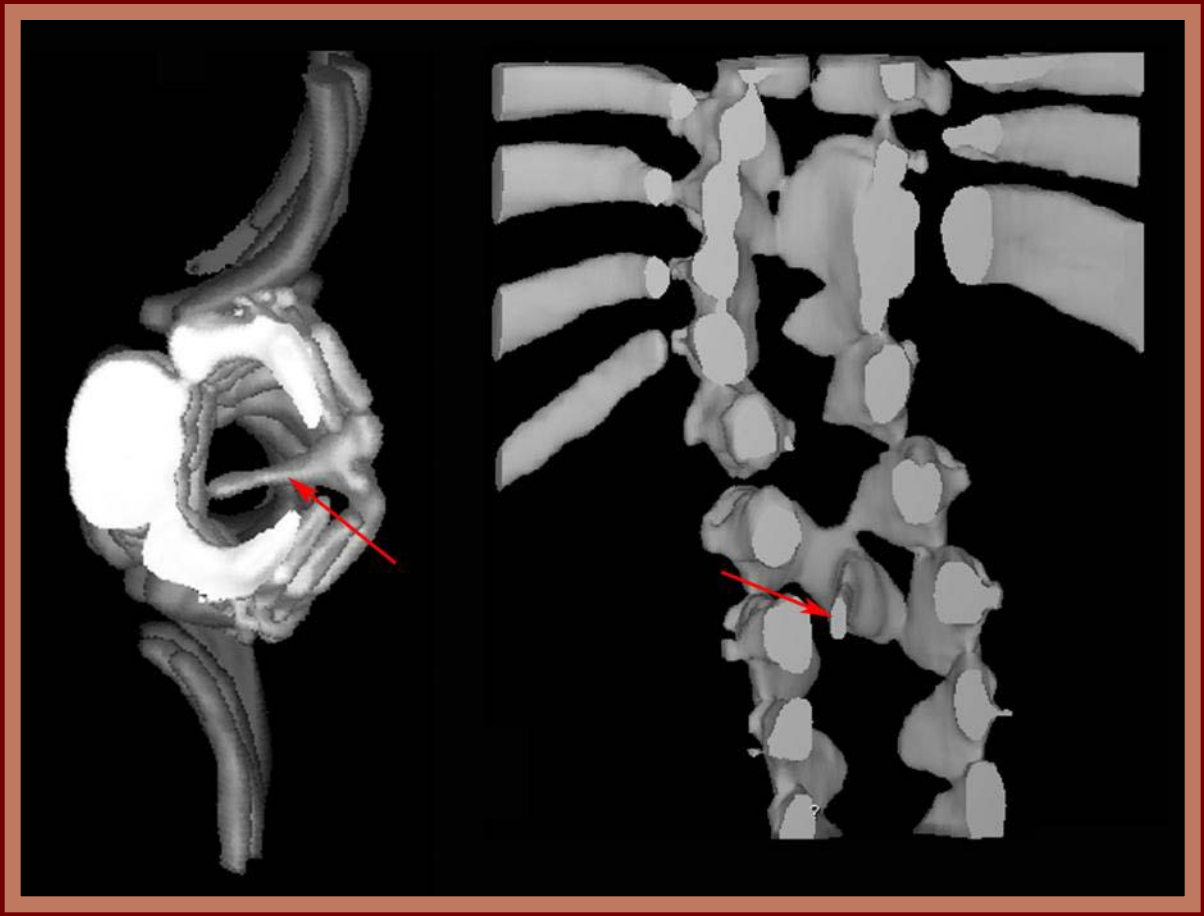
- cutaneous changes in middle lumbosacral region:
  - hypertrichosis
  - lipoma
  - dyscoloration
  - dermal sinus (cave infectious complications)

- **Serious conditions associated**
  - Syringomyelia
  - Diastematomyelia
  - Tethred cord syndrom

- **X-rays diagnosis**
  - L5-S1 level – dorsal part of spinal canal - closure defect

Spinal dysraphism

- Diastematomyelia in CT 3D



Spinal dysraphism

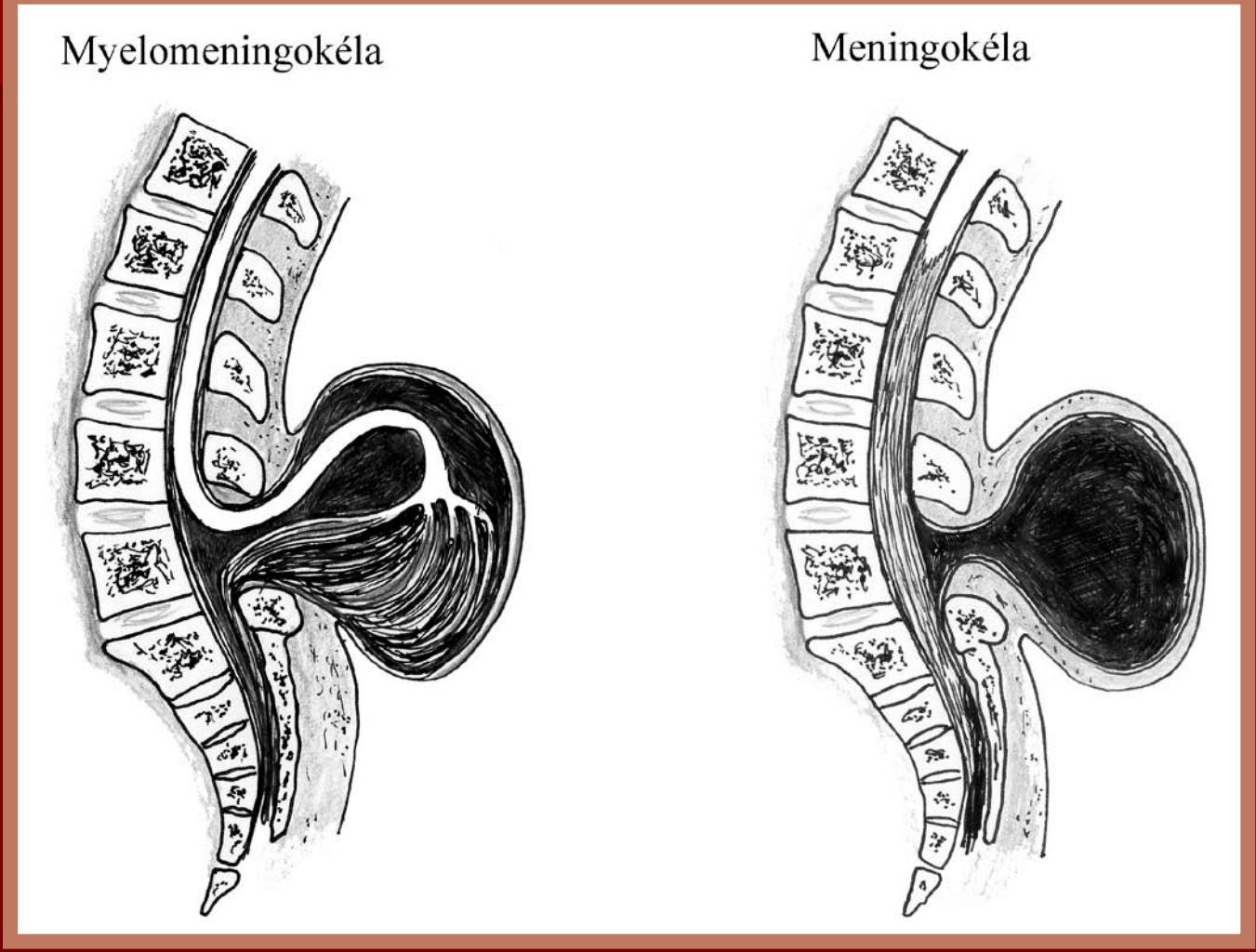
a) **Spina bifida aperta seu spina bifida cystica**
   - **eningocele** – vertebral arches defect, meningeal cyst, in 1/3 neurological deficit
   - **myelomeningocele** - vertebral arches defect, meningeal cyst, structural and functional nervous tissue abnormalities

- **Myelomeningocele epidemiology**: 1 from 1000 newborns

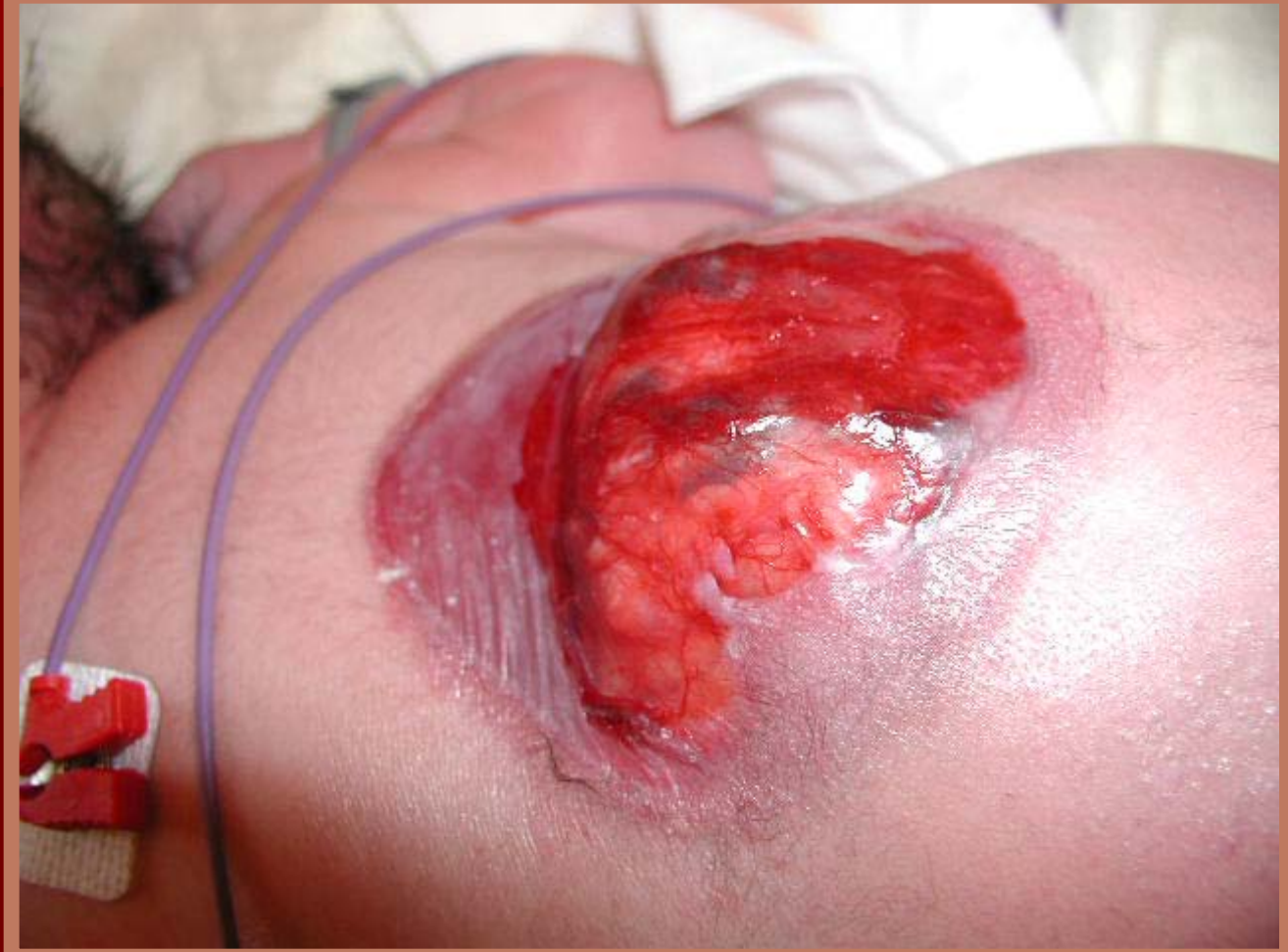
- **Clinical features**
  - lower extremities paresis proprioceptive reflexes disturbance incontinence

- **Associated conditions**
  - hydrocephalus (in 65–85 %)
  - Chiari malformation (in 80 %)

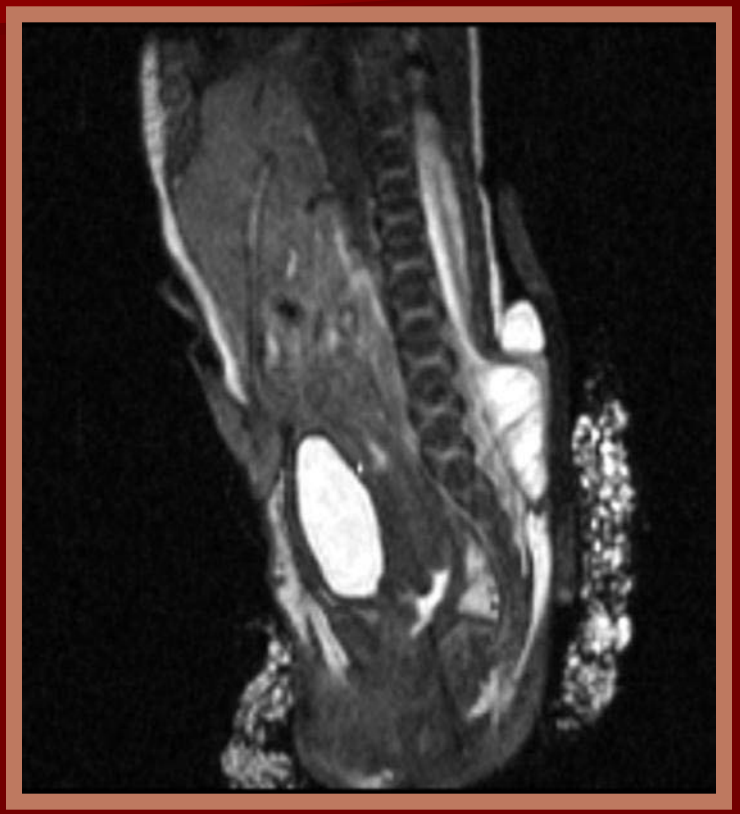
Spinal dysraphism

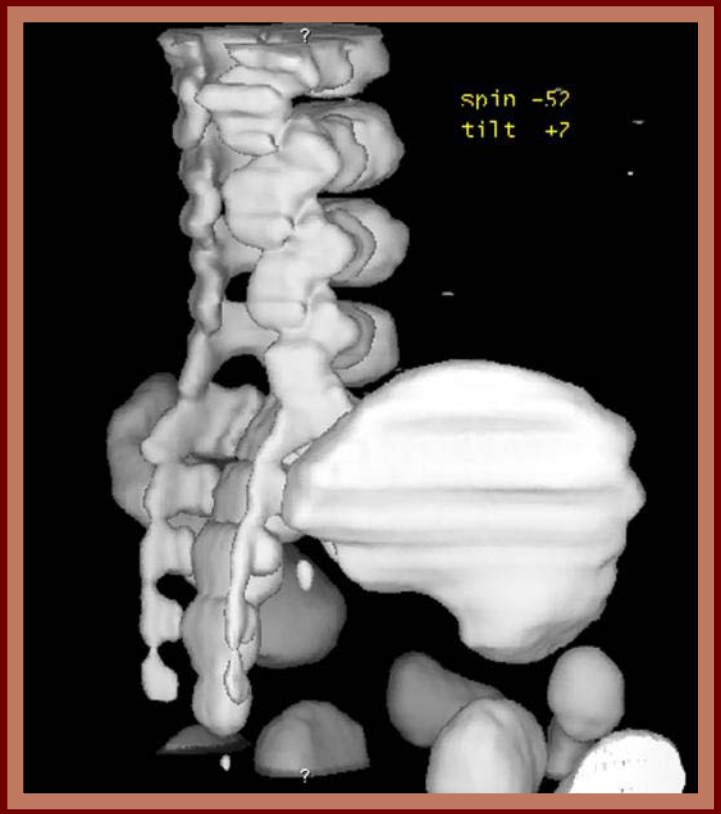


Spinal dysraphism

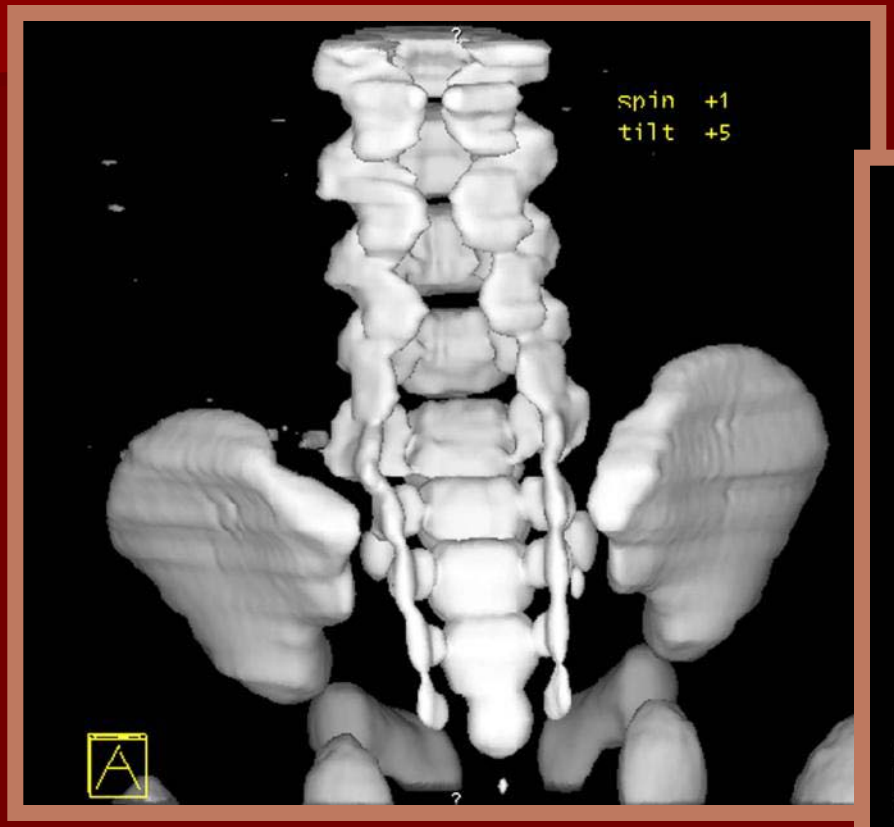


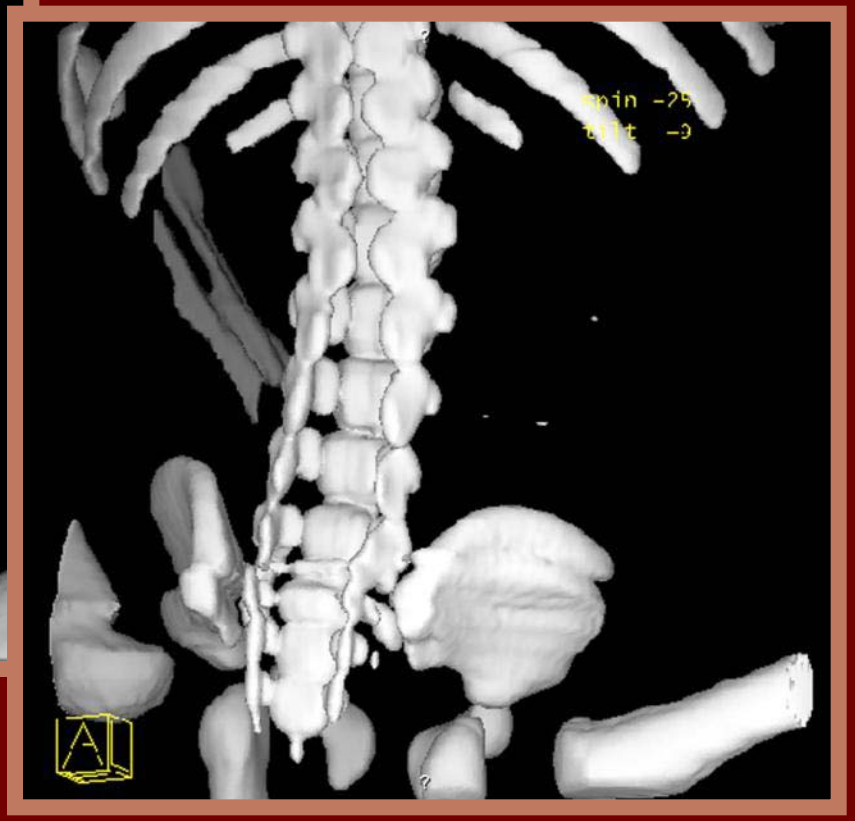
Spinal dysraphism





Spinal dysraphism





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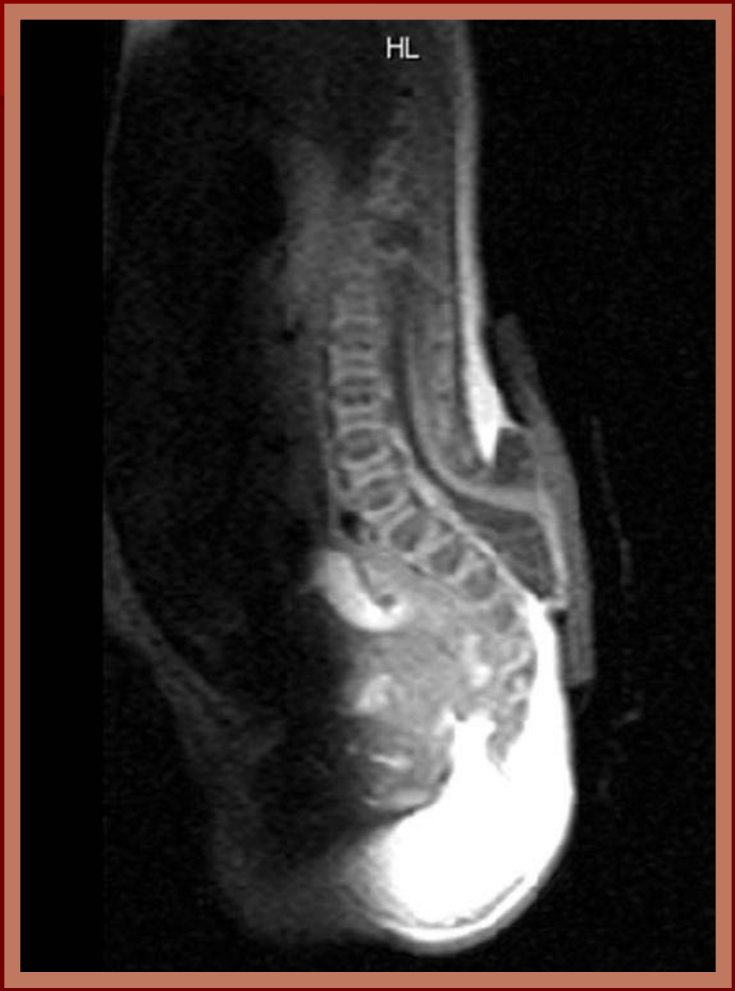
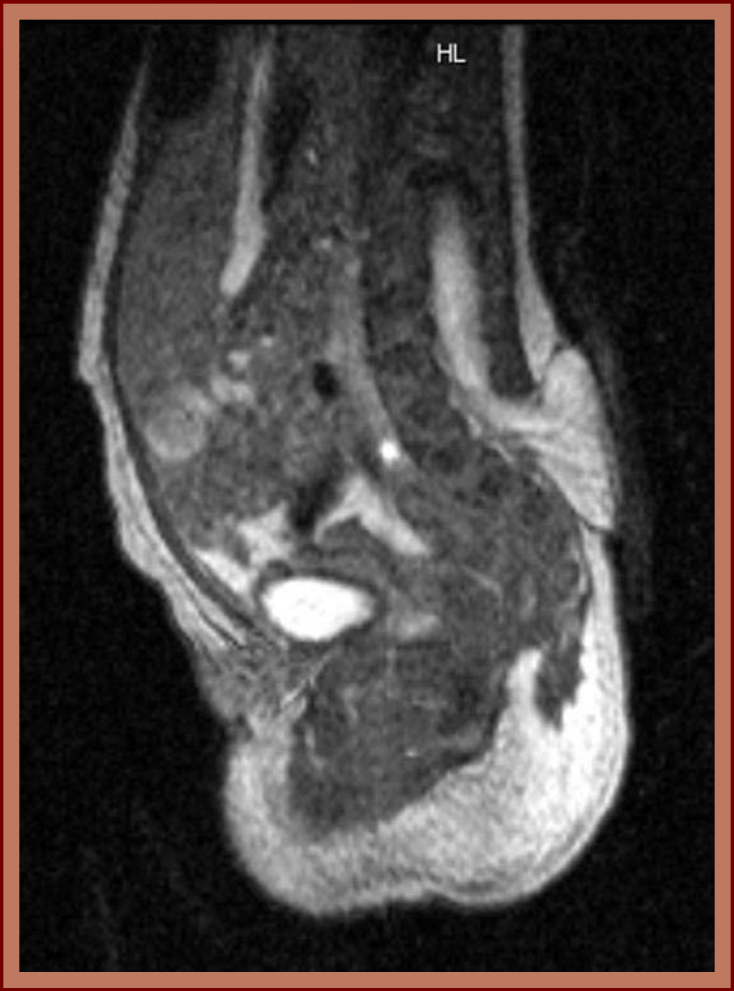
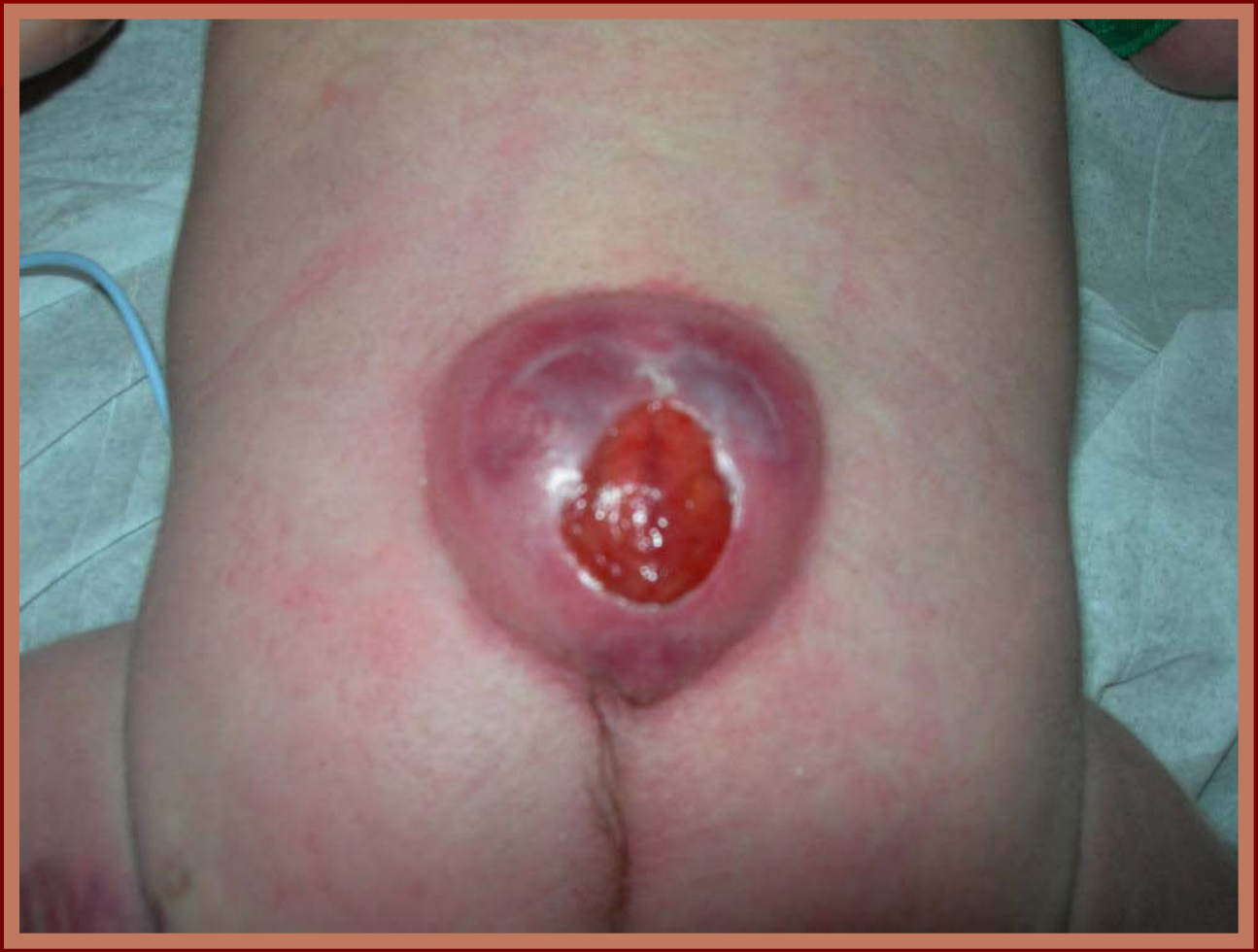




Spinal dysraphism



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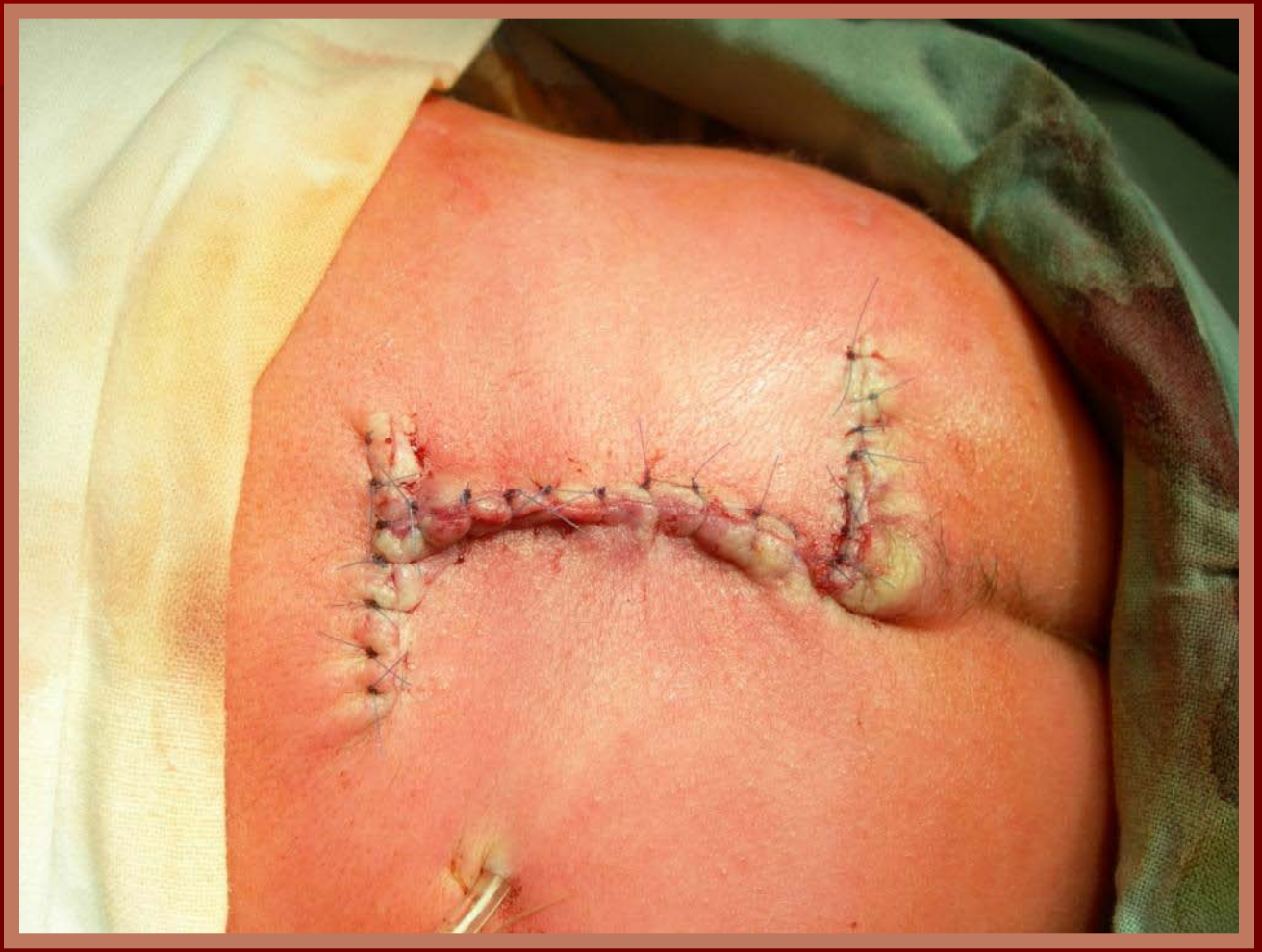
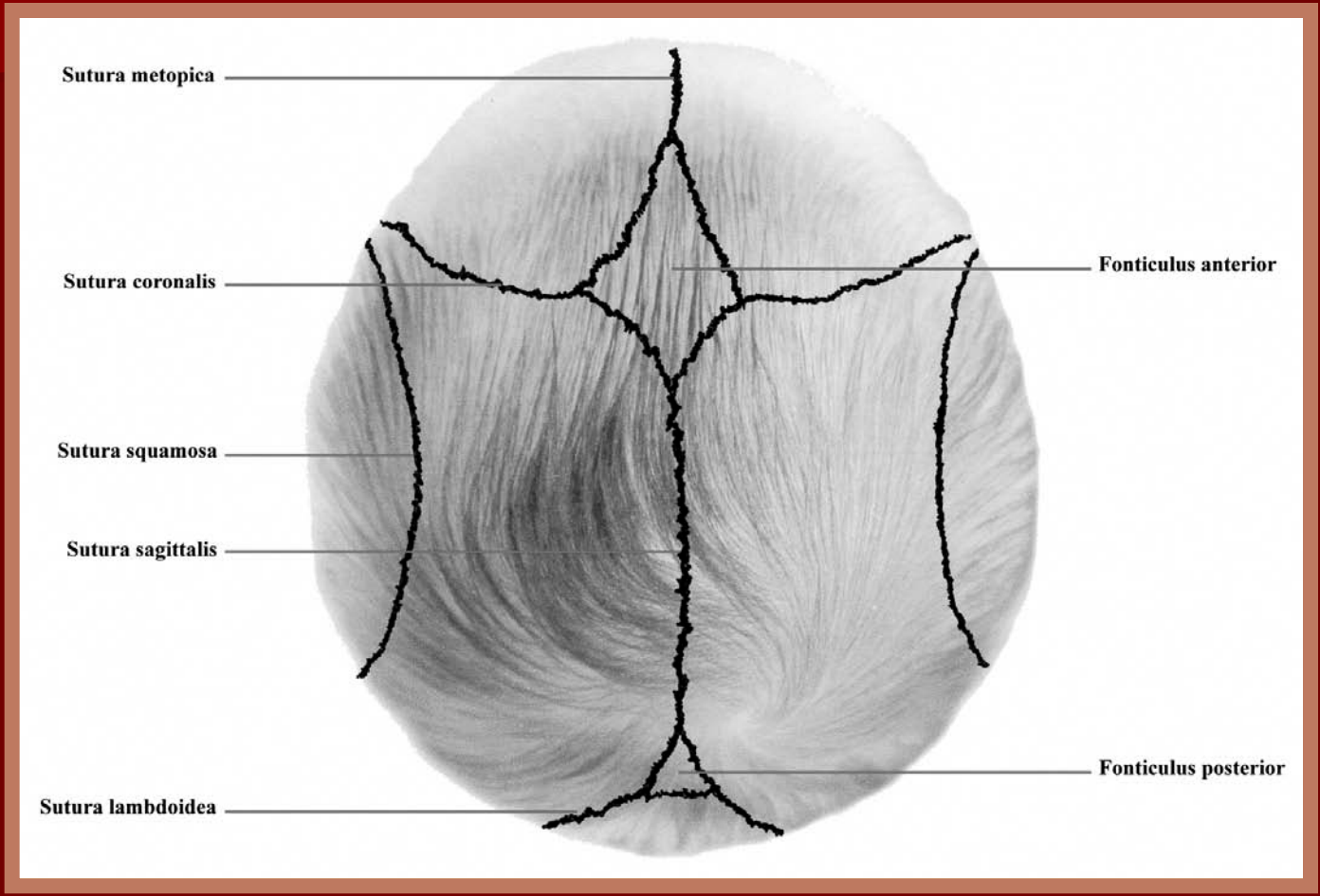


Craniostenosis (caniosynostosis)

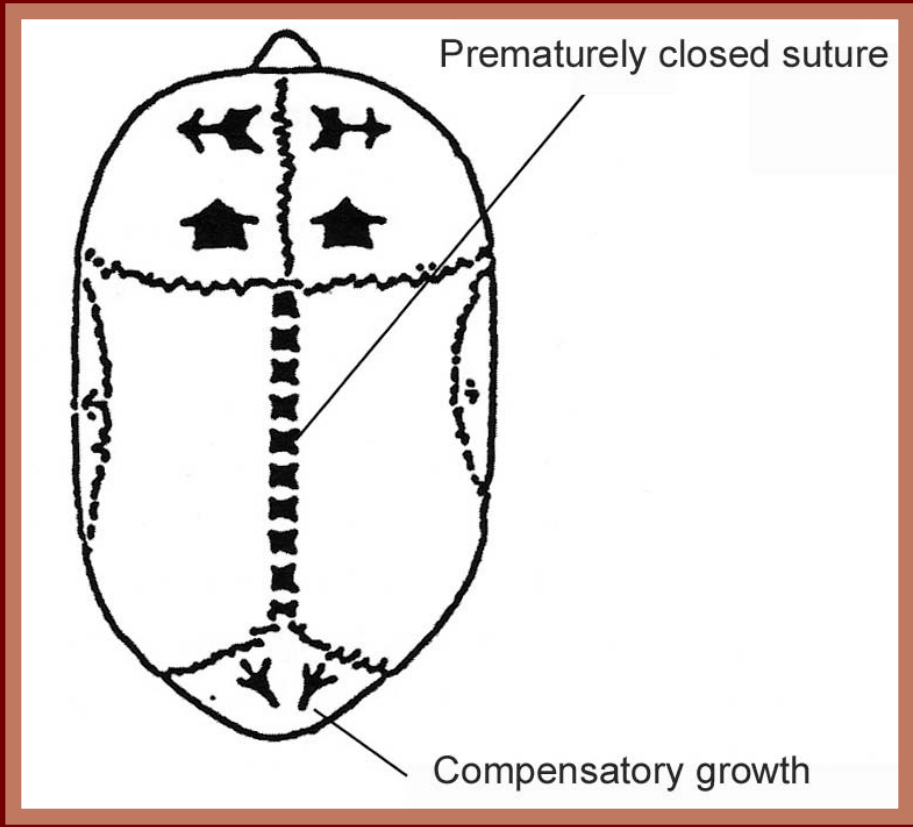
- Premature skull sutures synostosis
- 1852 Rudolf Virchow
- 1 from 2100 children

Sutures of the skull



1. Skaphocephalia - dolichocephalia

- Premature sagital suture synostosis
- Výskyt 40–60 %



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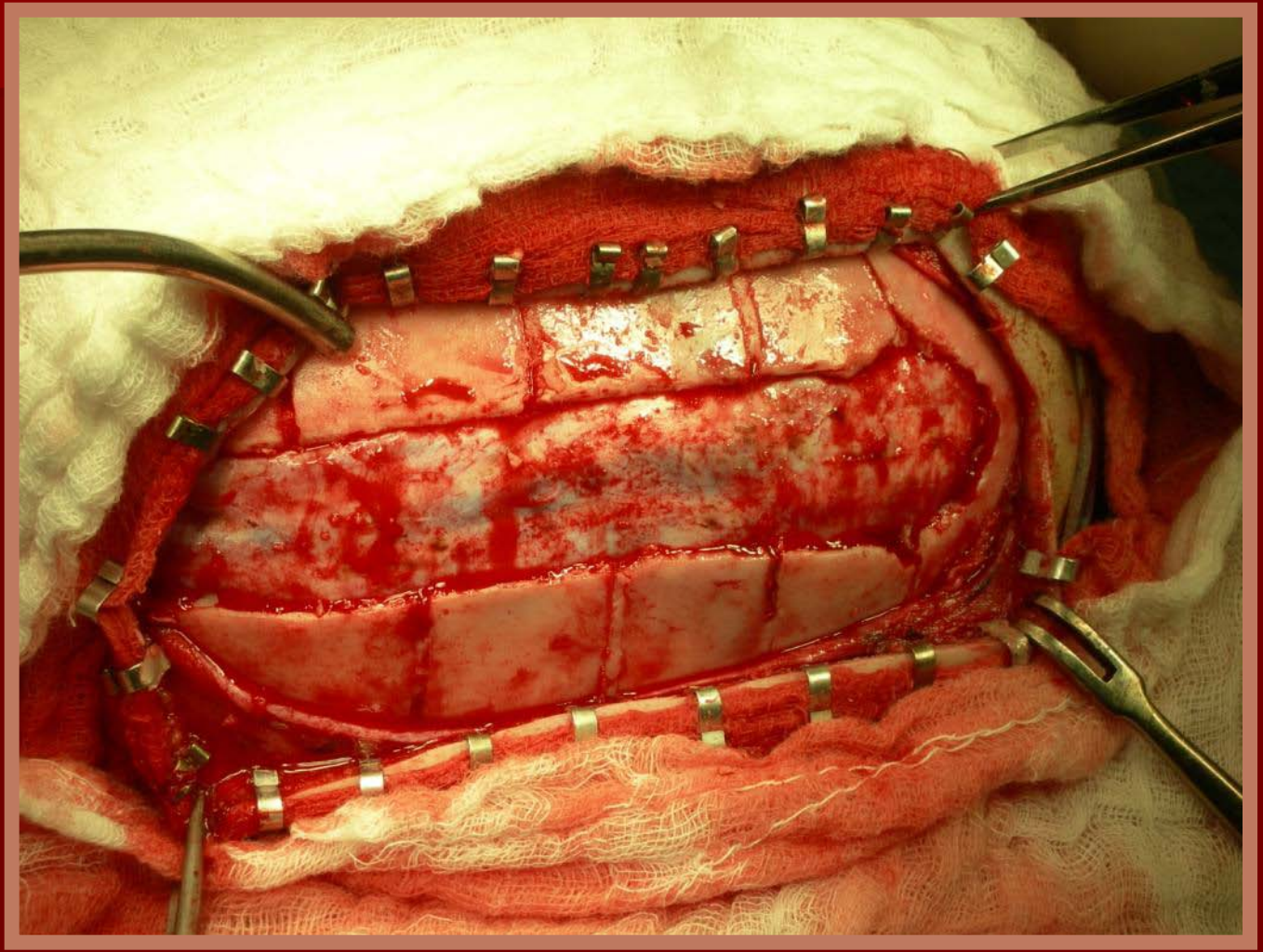
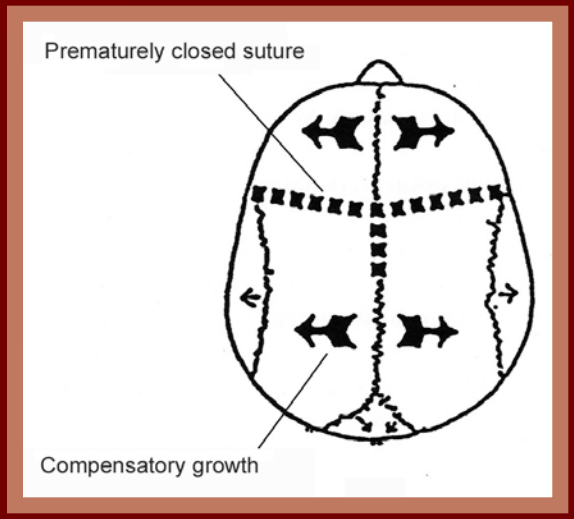


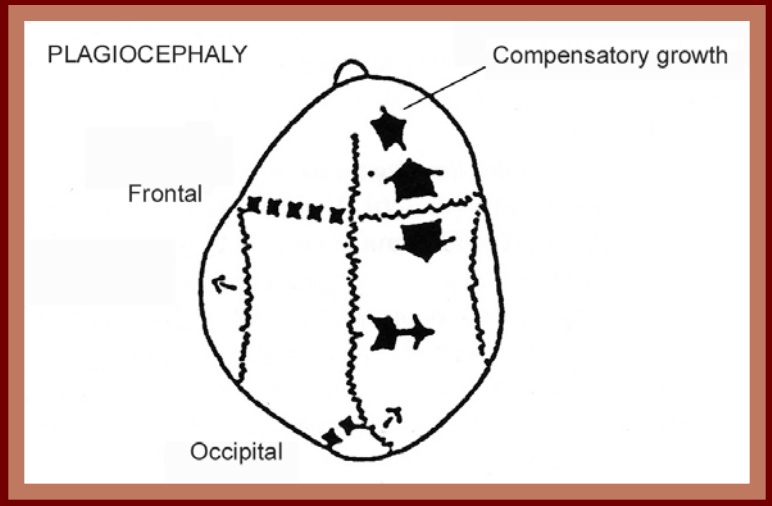
2. Brachycephalia

- Premature coronar suture synostosis
- 20–30 %

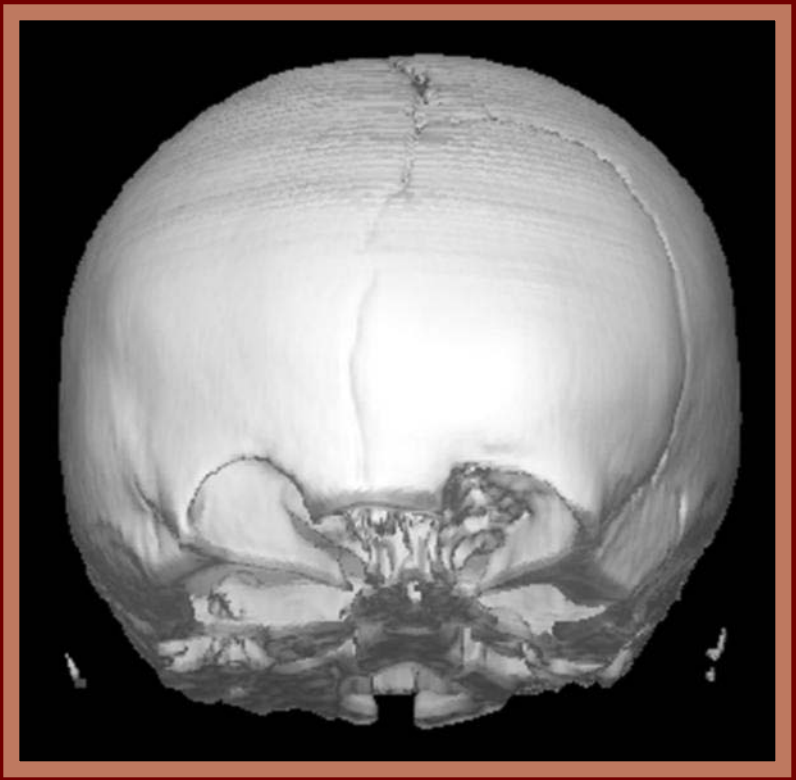
a) **Frontal plagiocephalia**
   - one side coronar suture

b) **Occipital plagiocephalia**
   - one side lambdoid suture

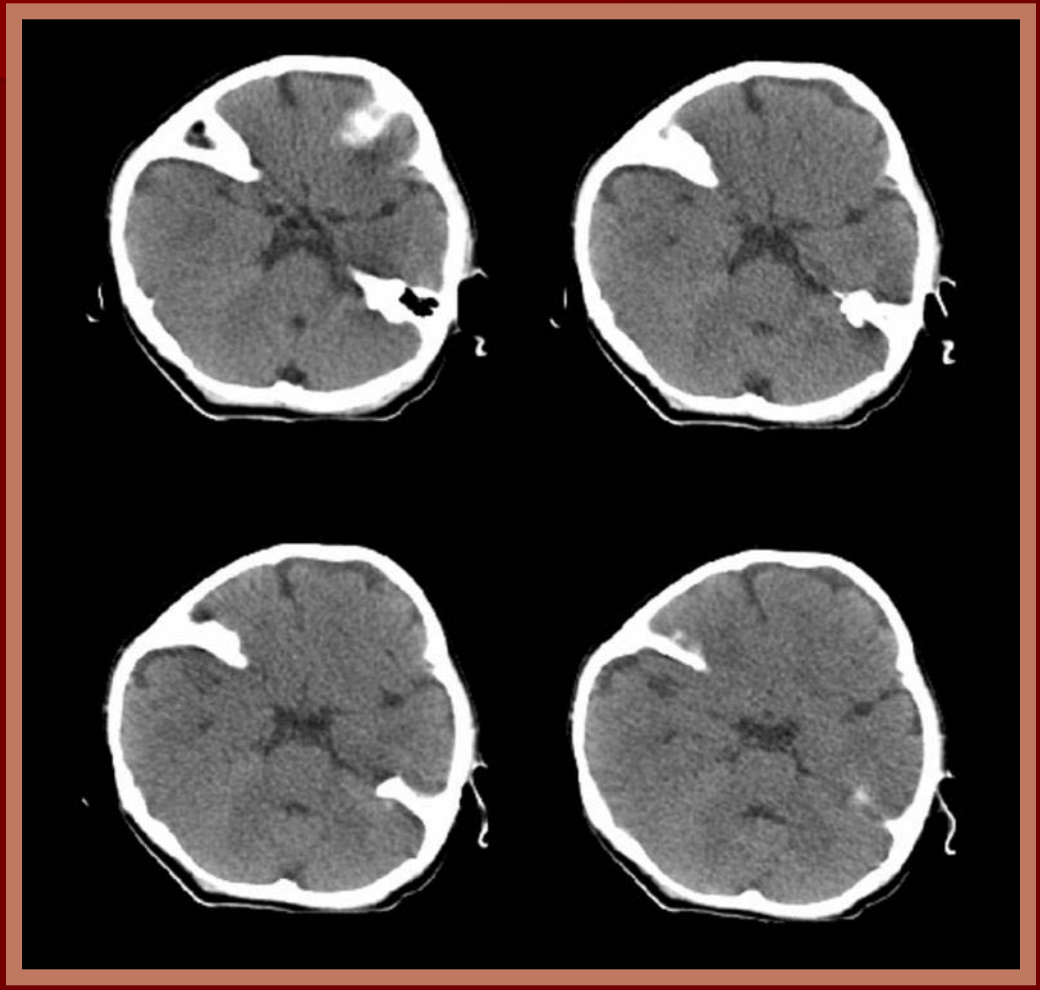




Brachycephalia

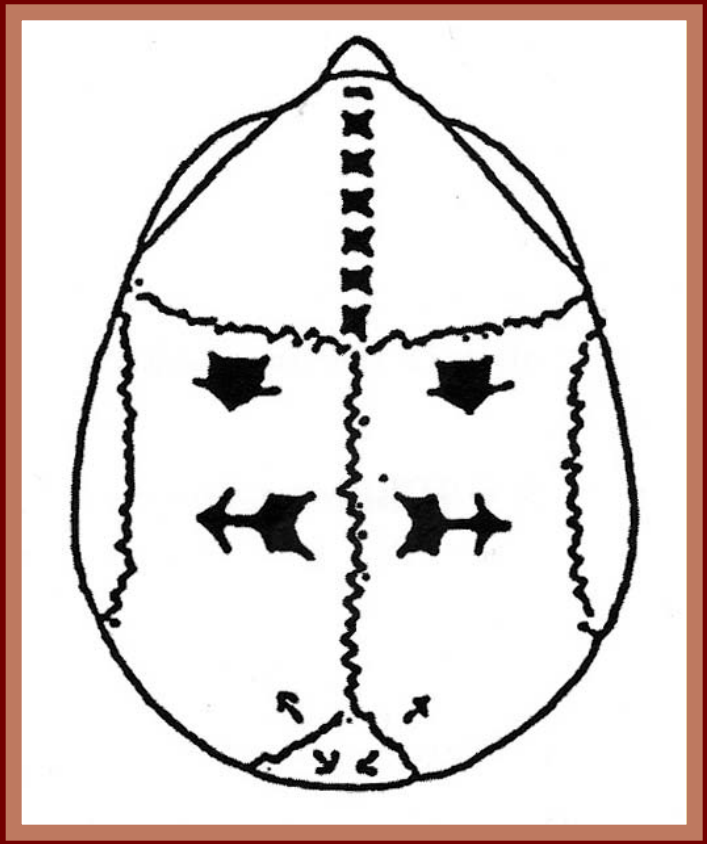


Brachycephalia

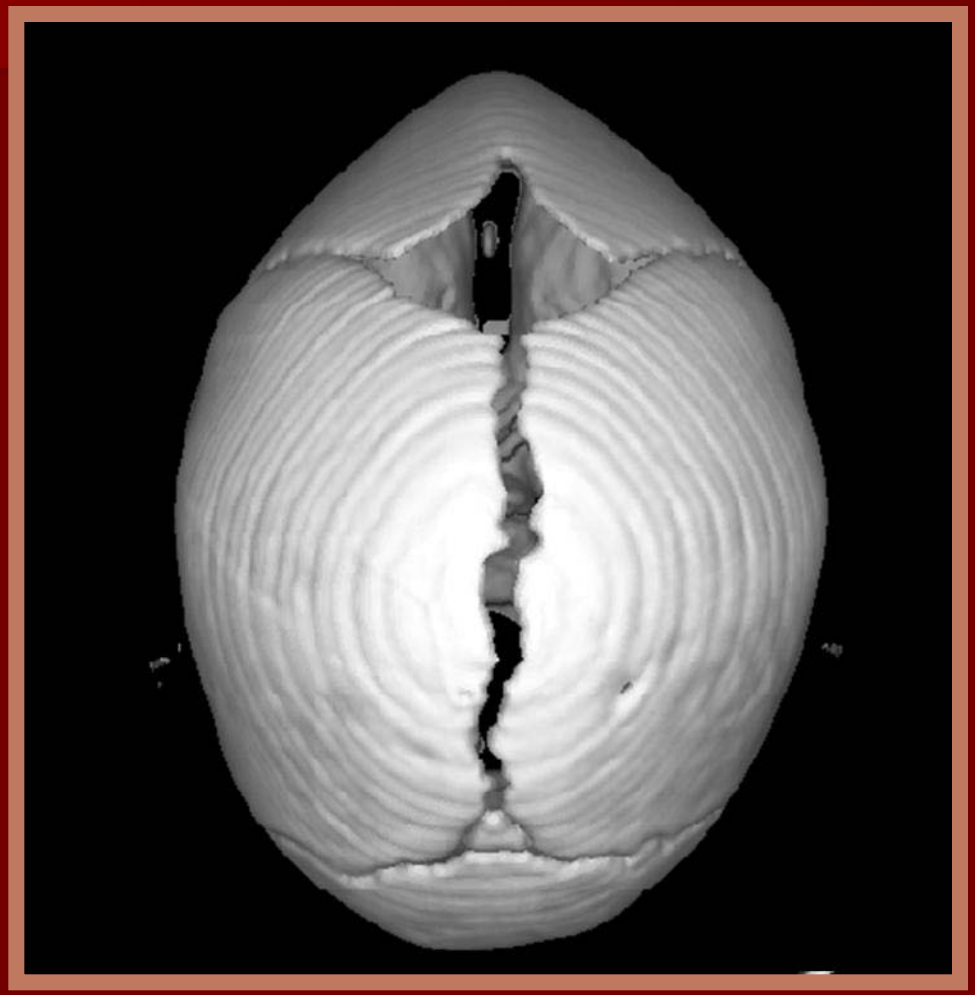


3. Trigonocephalia

- Premature sutura metopica synostosis
- 10%



Trigonocephalia



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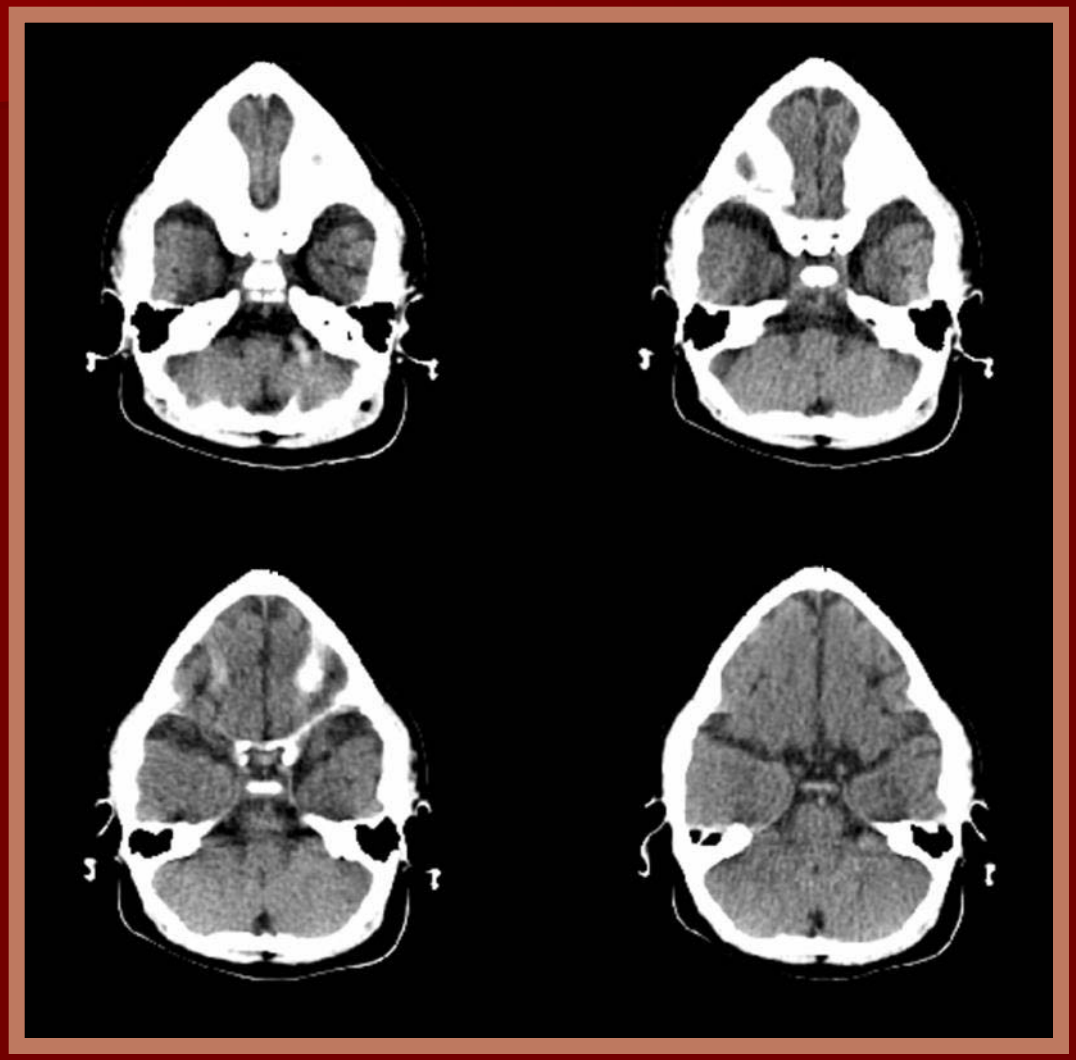

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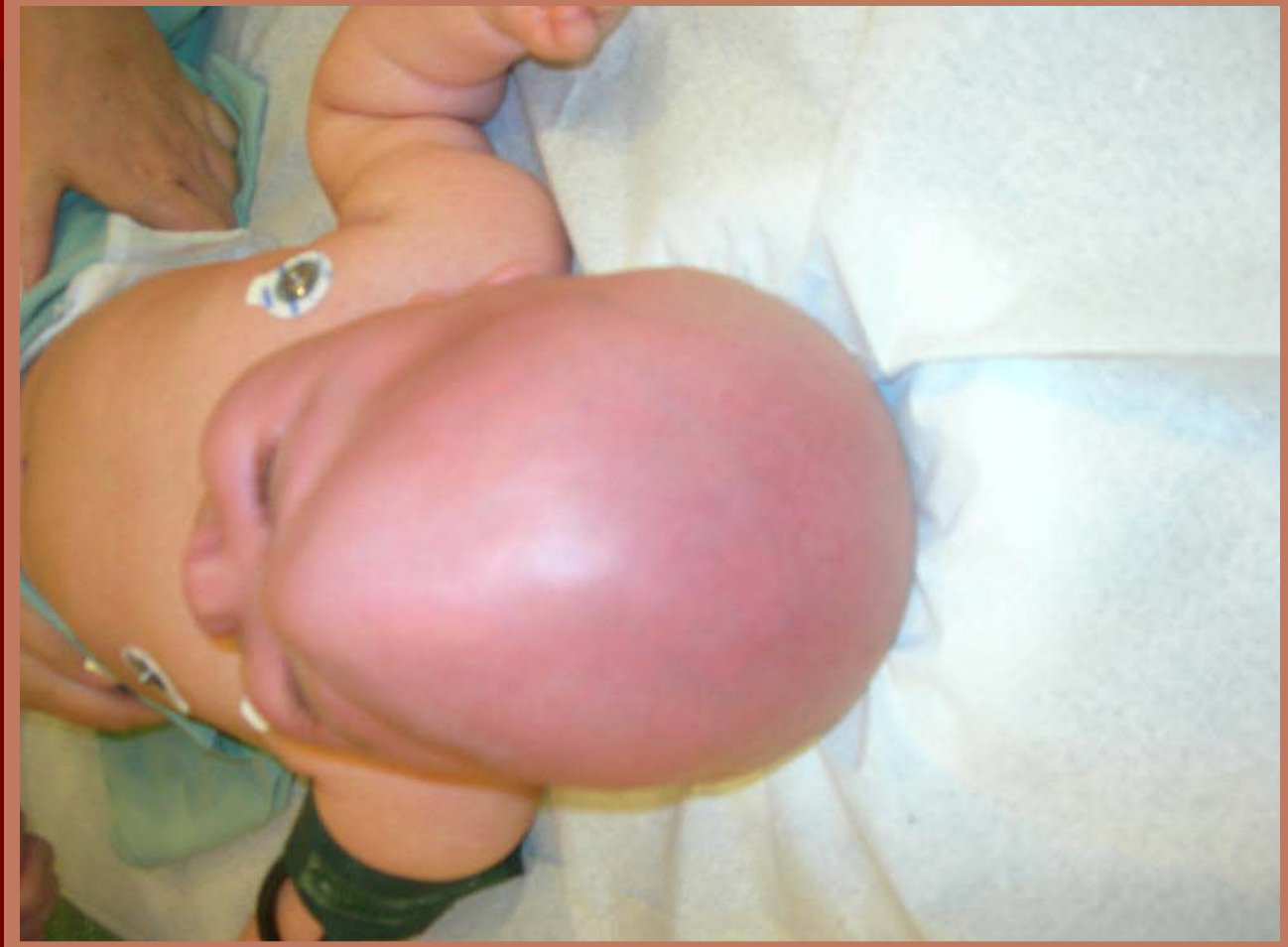

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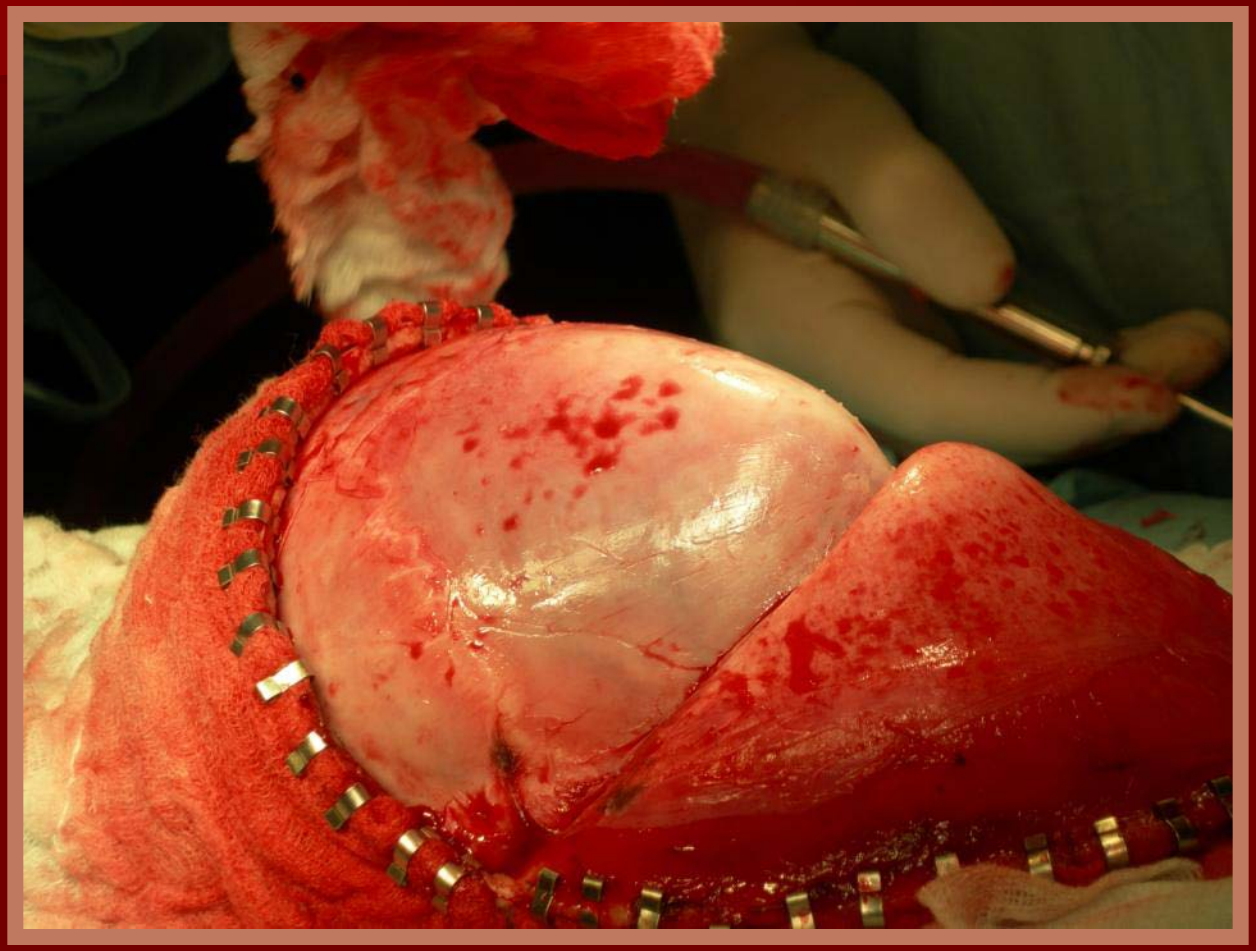

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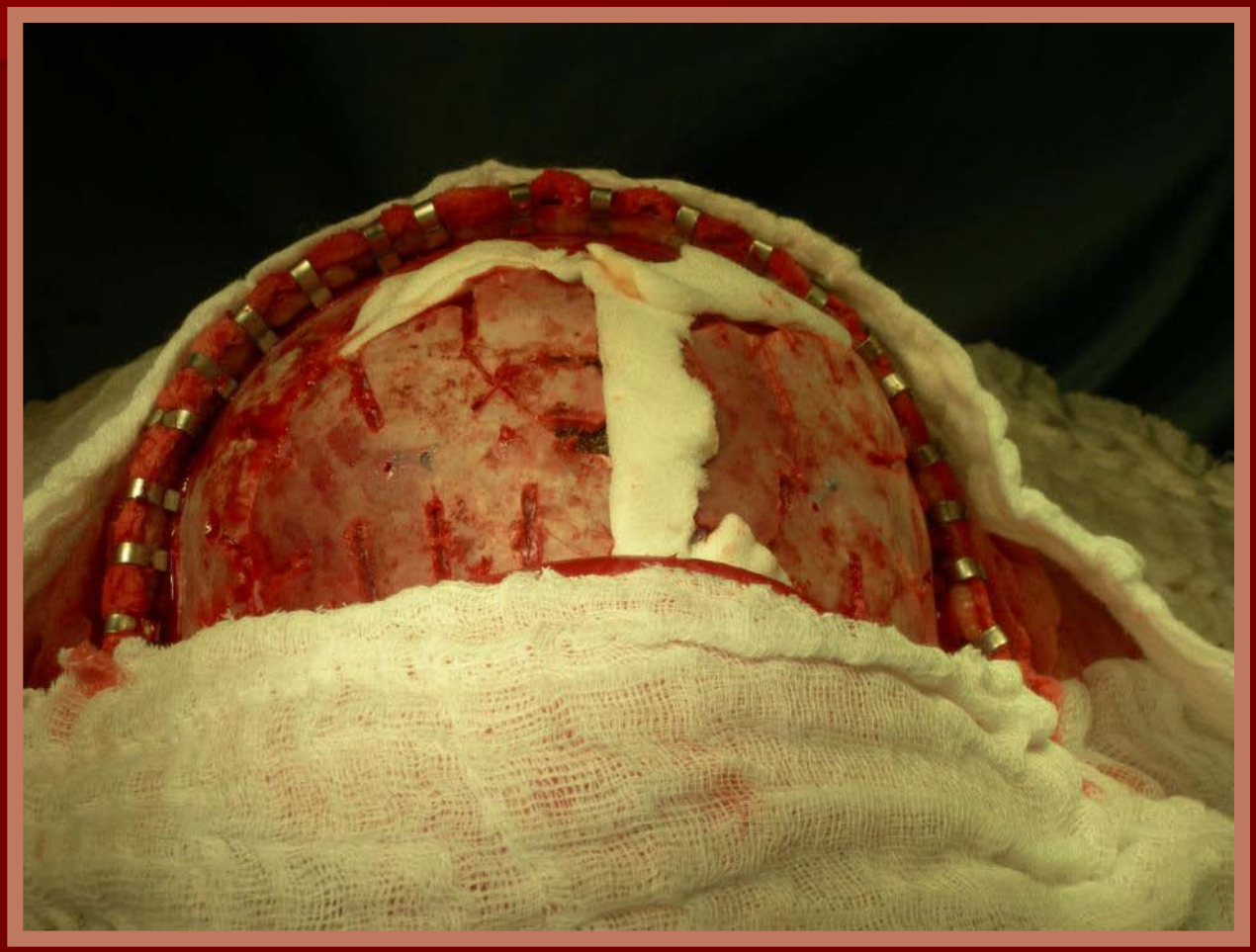

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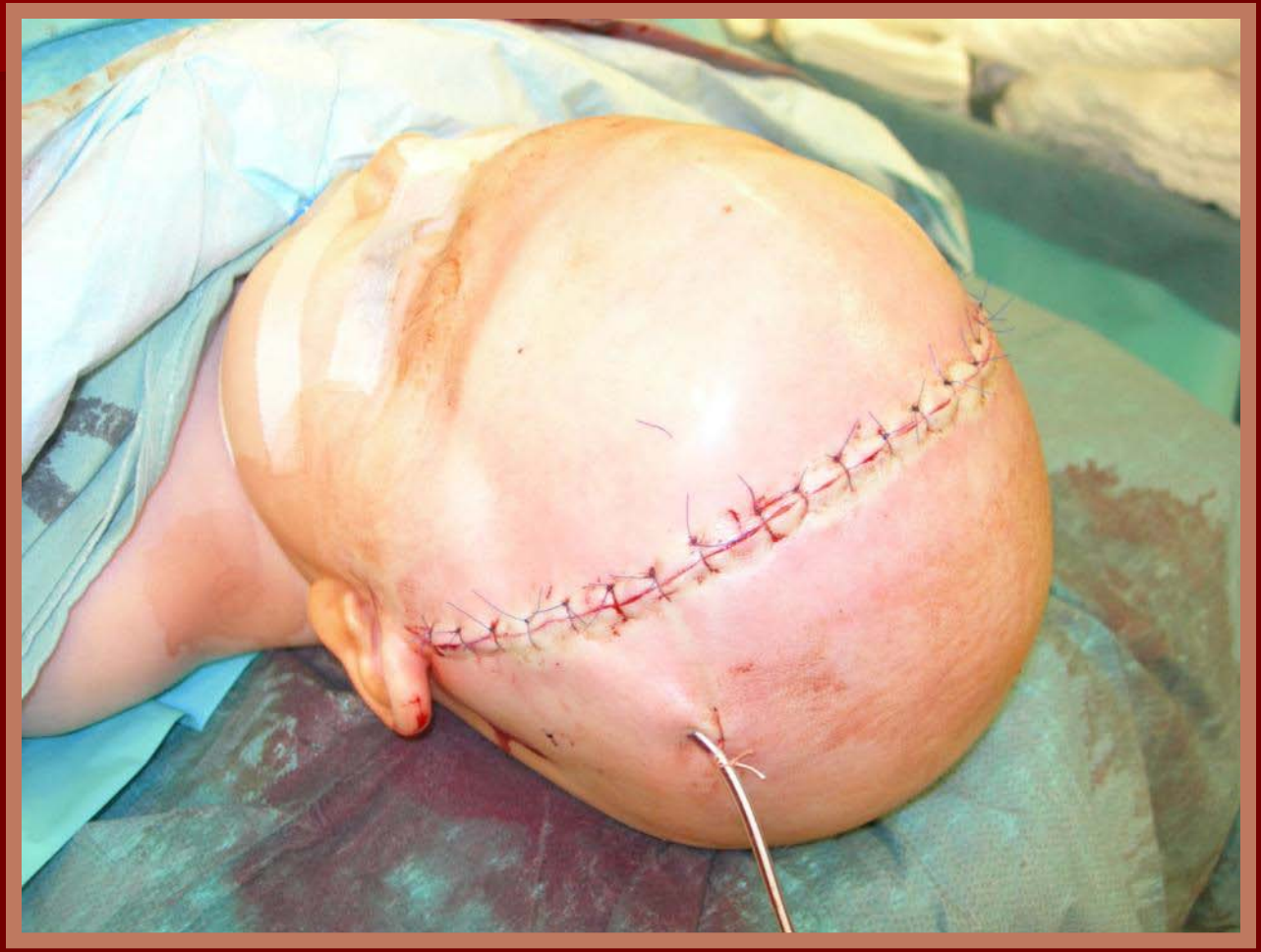
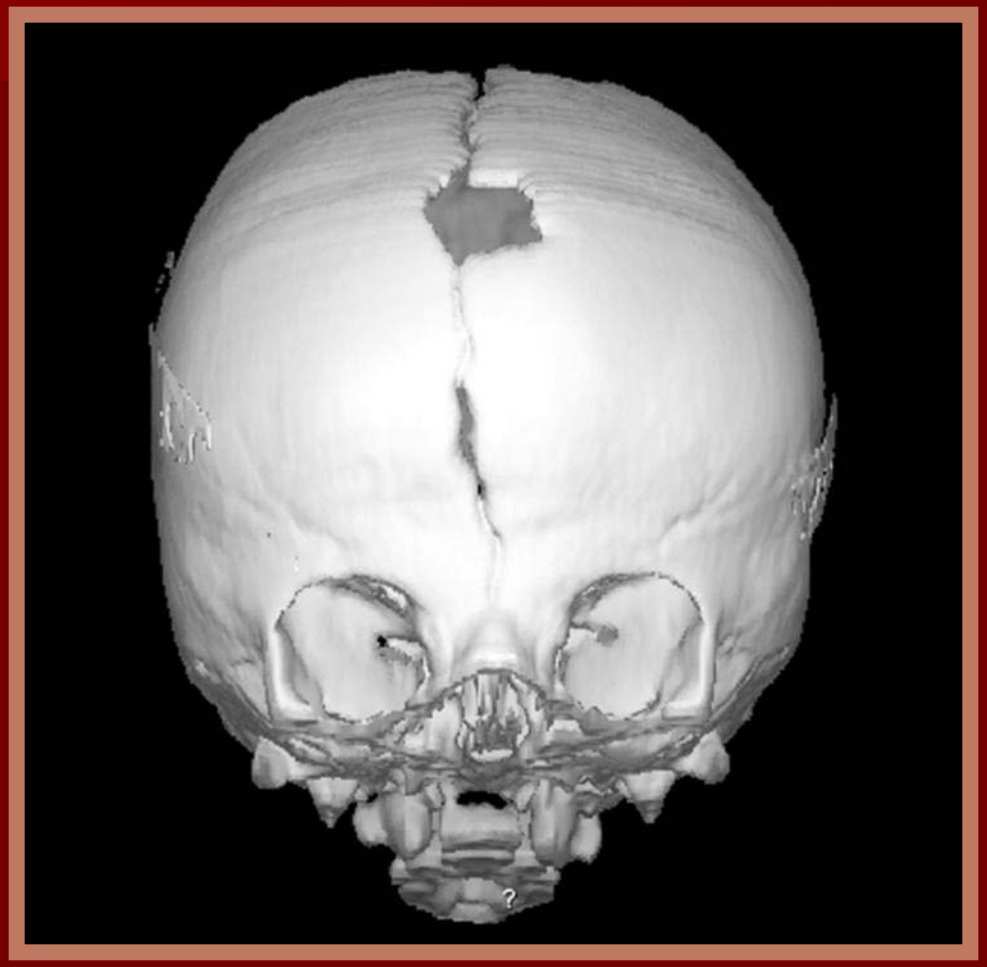


4. Morbus Crouzon – dysostosis craniofacialis (1912)

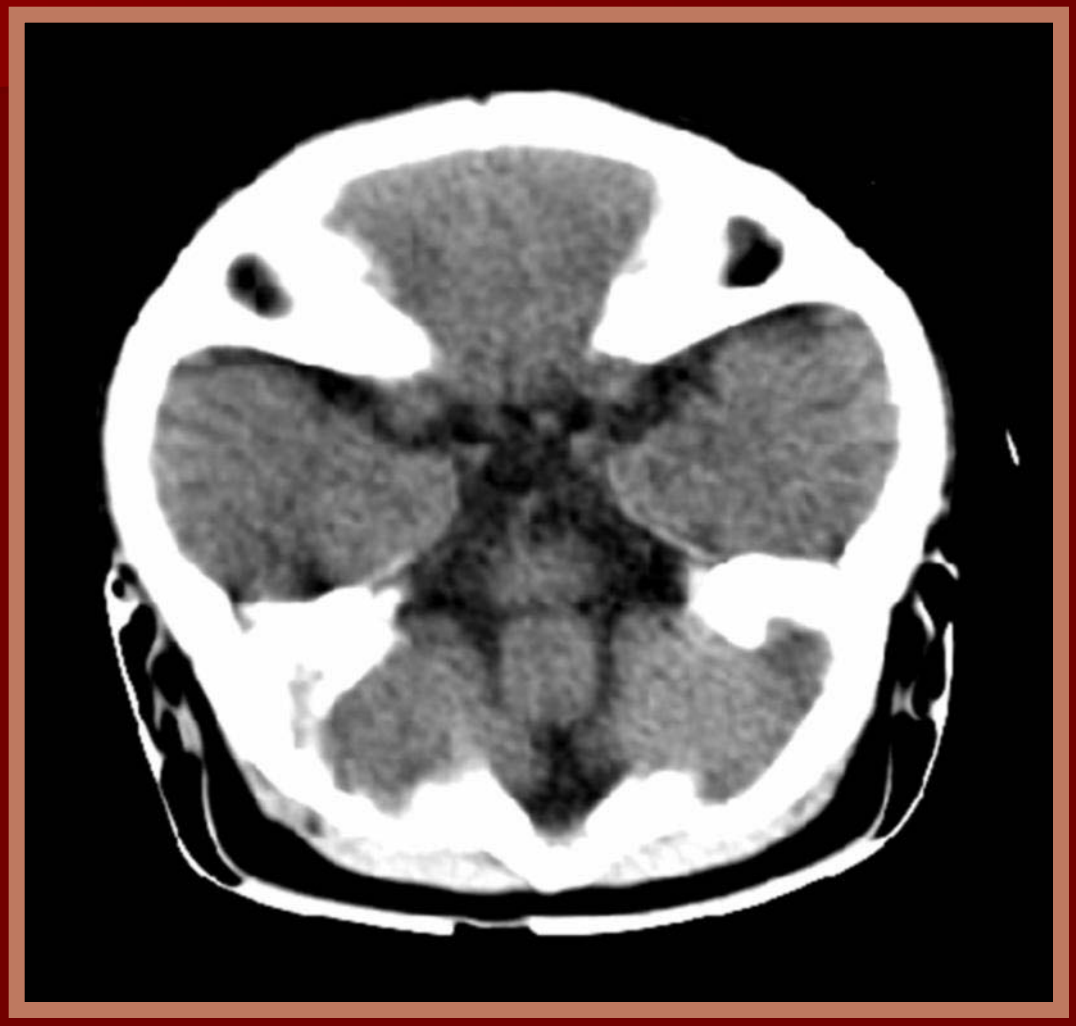
- Turicephaly
- Shallow orbits
- Exophalmos
- Hypertelorism
- Hypoplasia of middle facial skelet

- **1 from 25 000 children**

Morbus Crouzon



Morbus Crouzon



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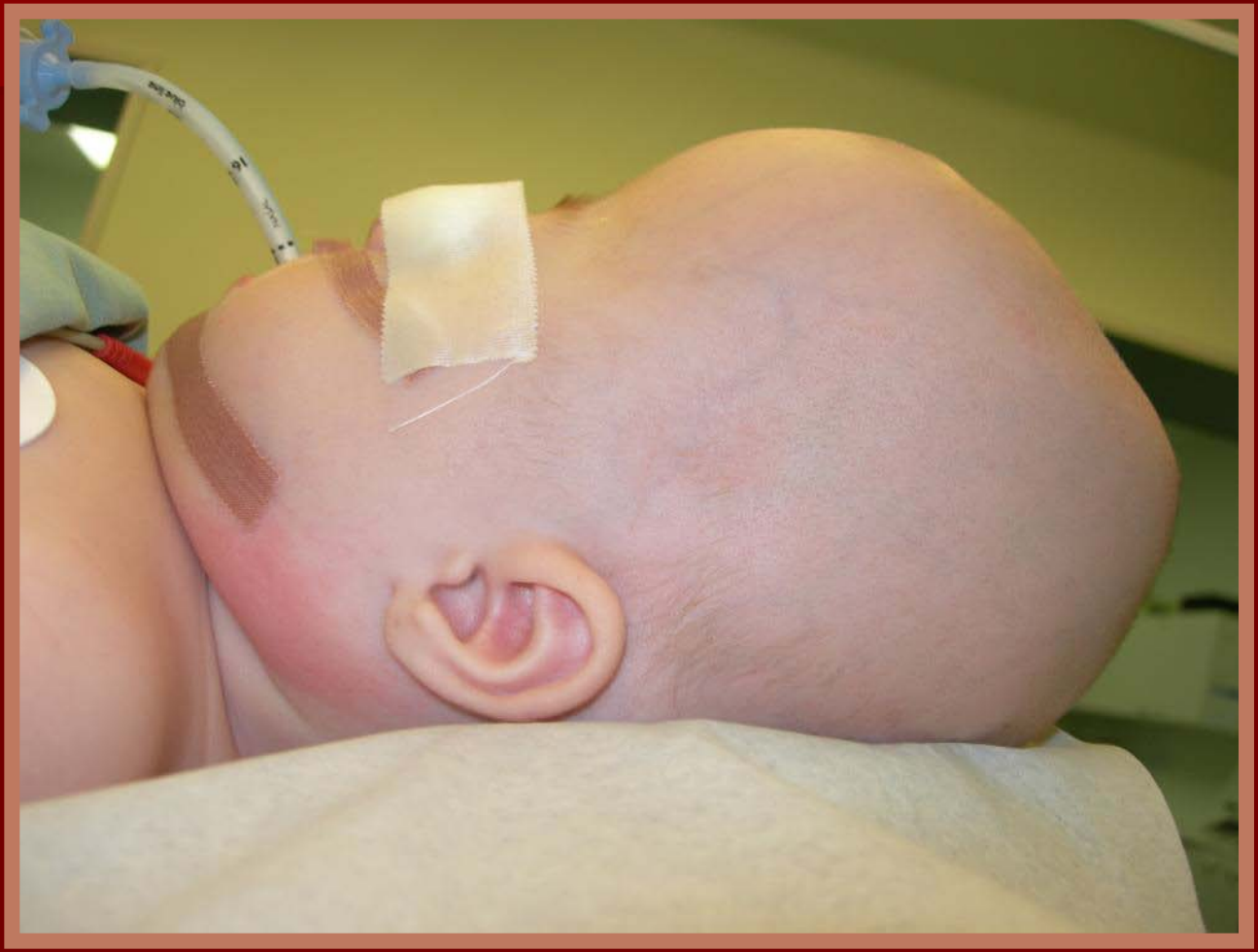

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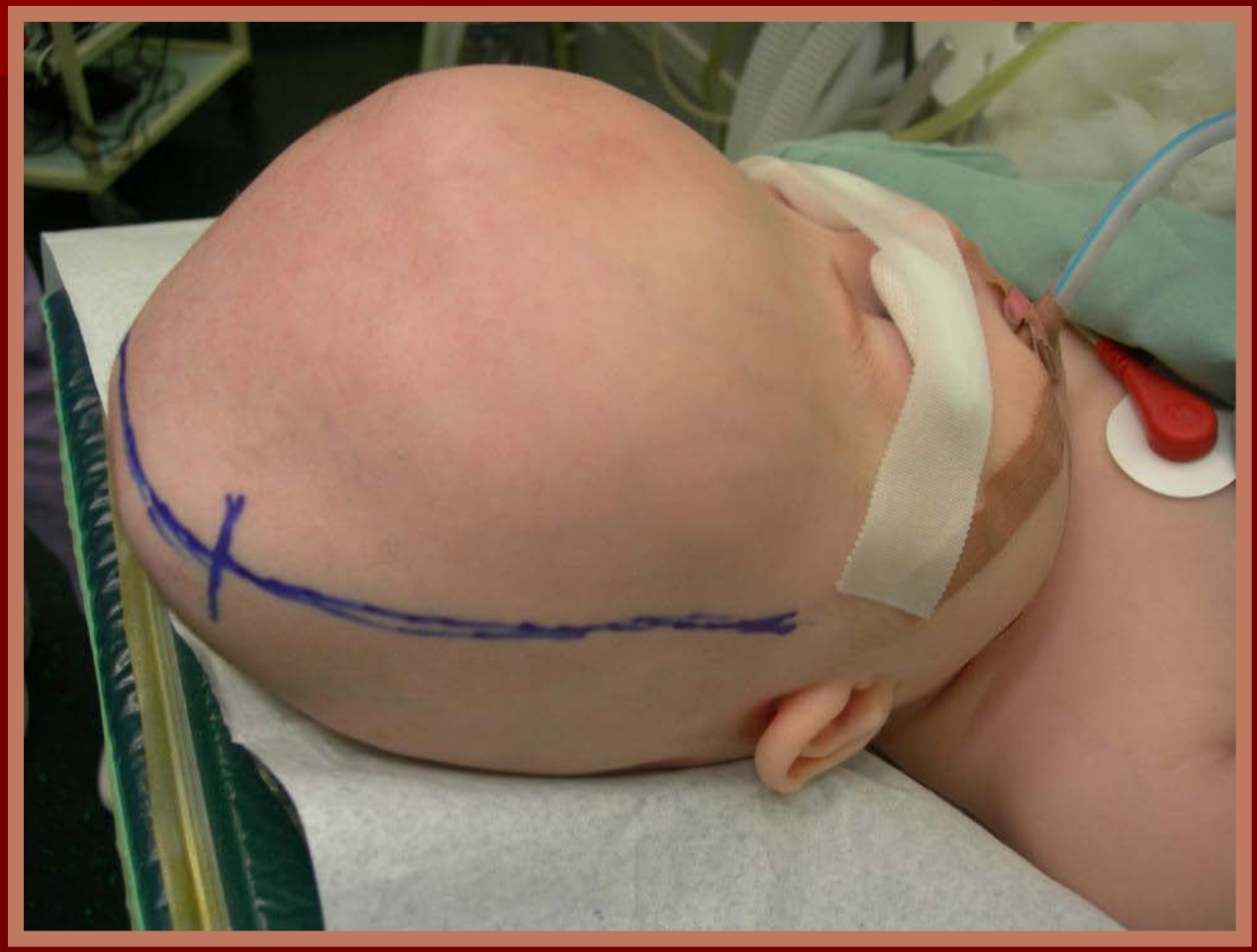

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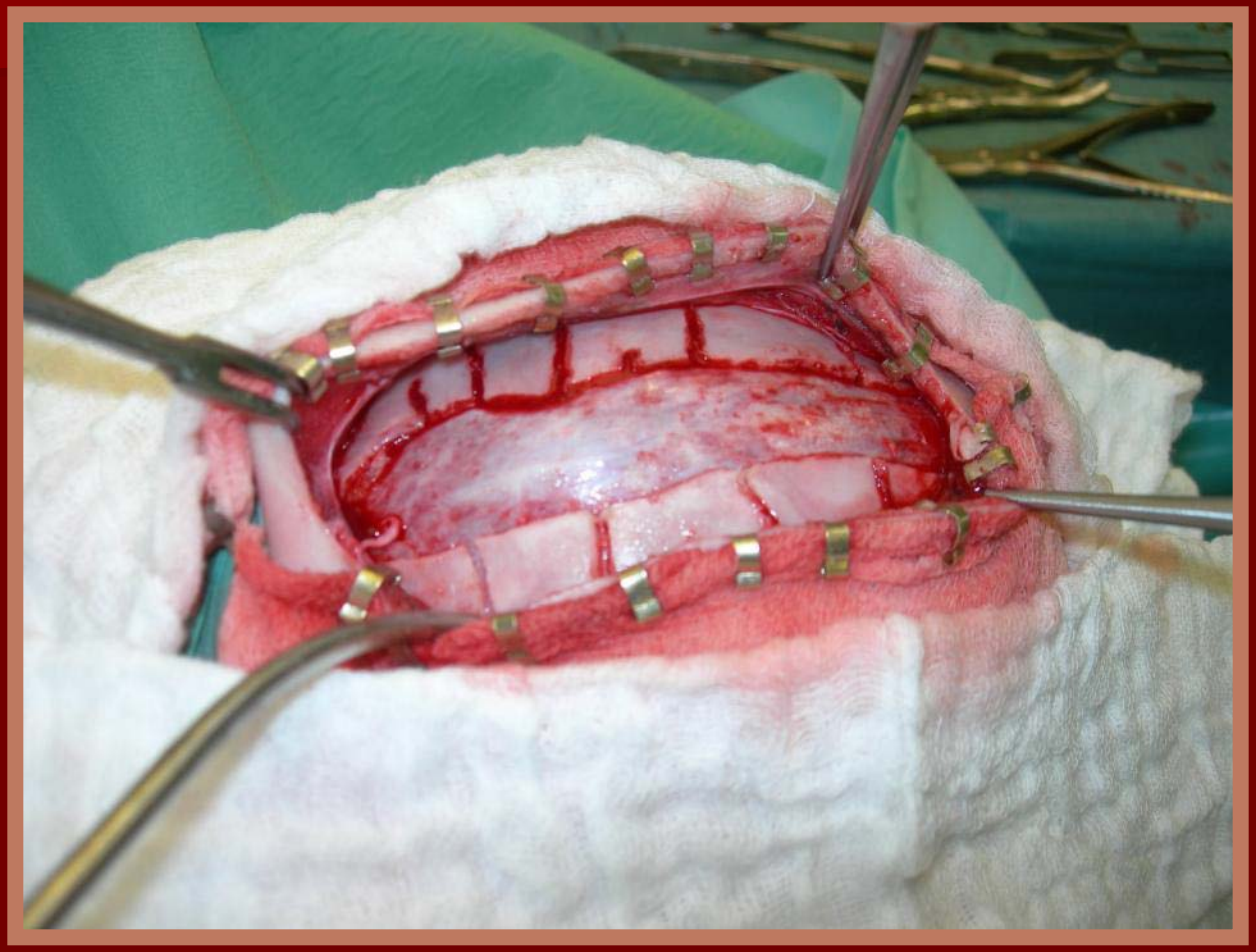

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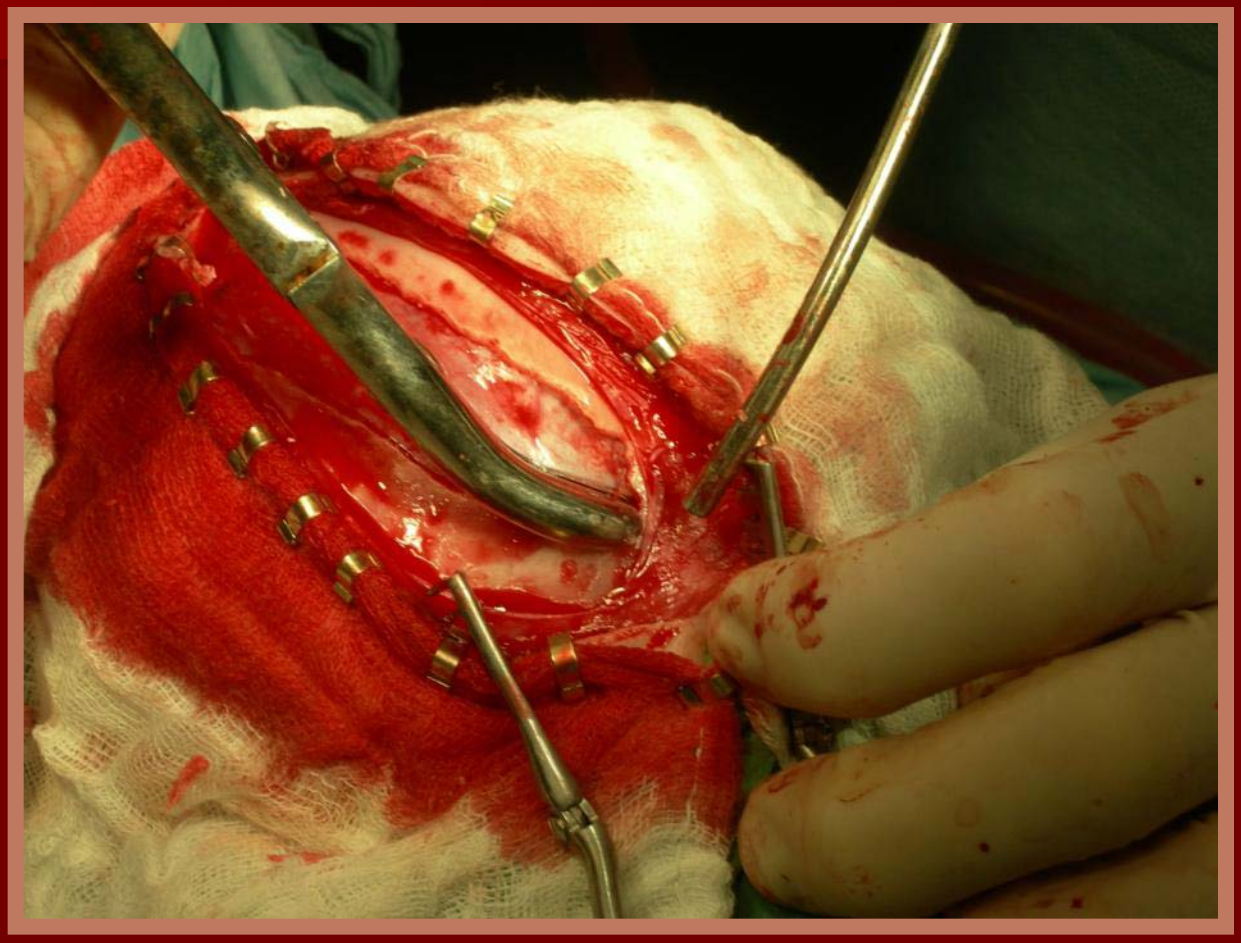

Post surgery care

Preventive protective helmets