

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

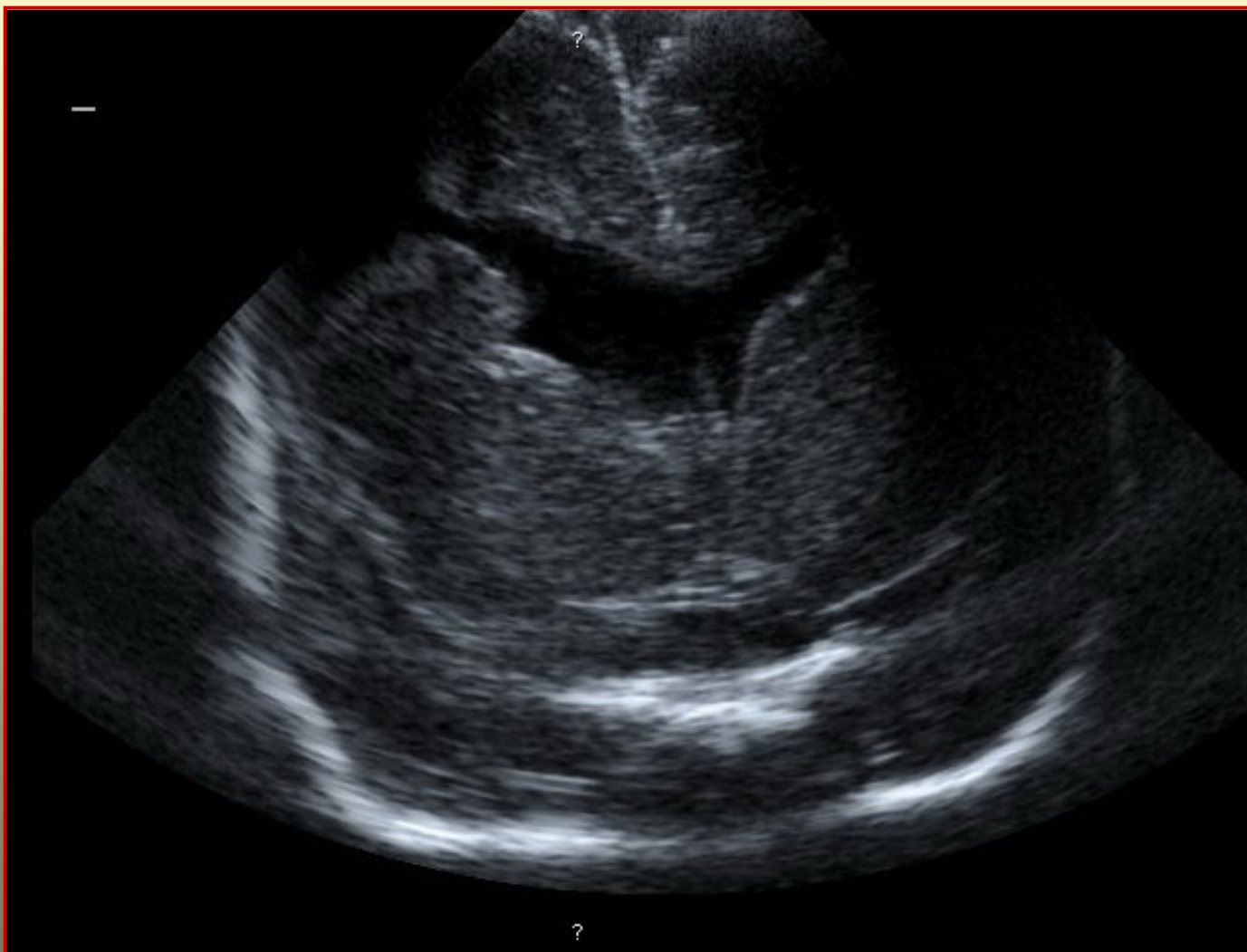
Cranial meningocele mostly has a good prognosis.

Encephalocele is mostly accompanied by hydrocephalus, mikrocephaly, mental retardation, epileptic seizures.



- a) microcephalia
- b) hydranencephalia - a loss of almost all cerebral tissue
- c) holoprosencephalia – hemispherical development disturbance
- d) lissencephalia - severe disturbance of neural tissue migration
 - agyria – completely smooth cerebral surface
 - pachygyria – few flat gyri
 - polymicrogyria – small gyri, shallow sulci (similar to pachygyria)
- e) porencephalia
- f) agenesis of corpus callosum
- g) Dandy-Walker syndrome (cerebellar hypoplasia)
- h) macrocephaly - megalencephaly
- i) schizencephaly

Schizencephalia



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:

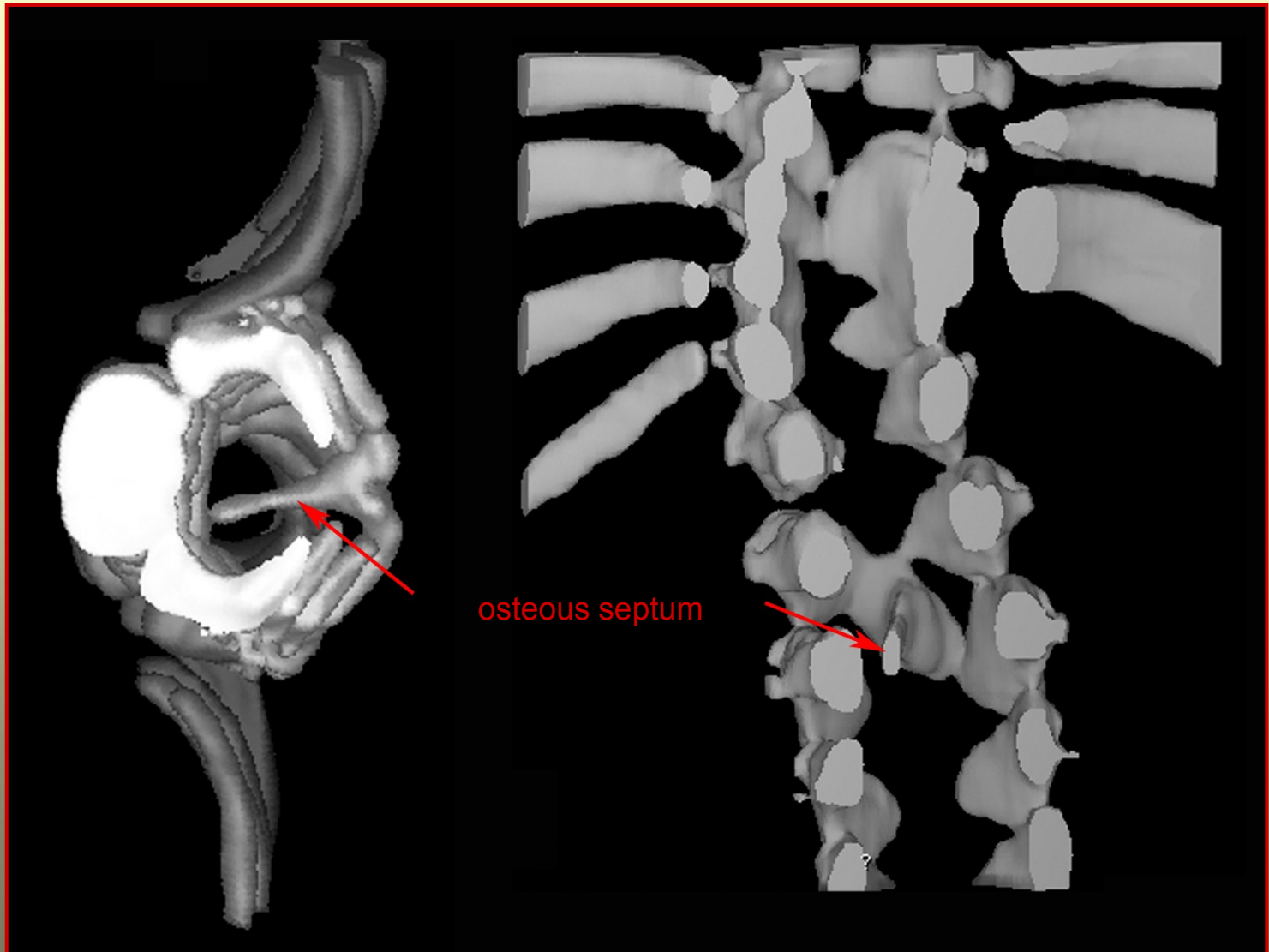
Syringomyelia

Diastematomyelia

Tethered cord syndrom

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

Diastematomyelia in CT 3D



b) Spina bifida aperta seu spina bifida cystica:

meningocele – 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

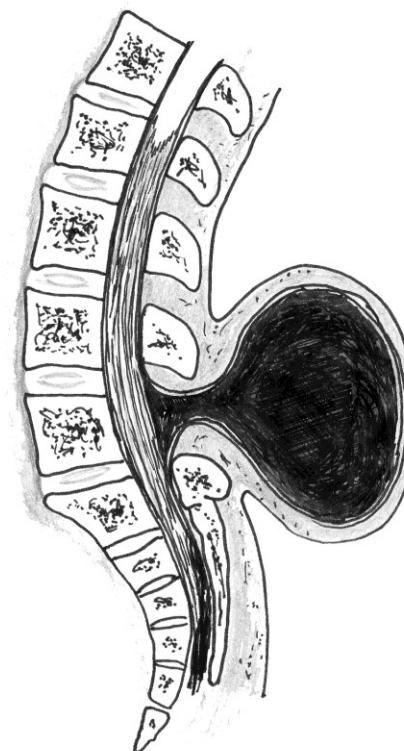
Associated conditions: hydrocephalus (in 65 – 85%)
Chiari malformation (in 80%)

Spinal dysraphism

Myelomeningokéla



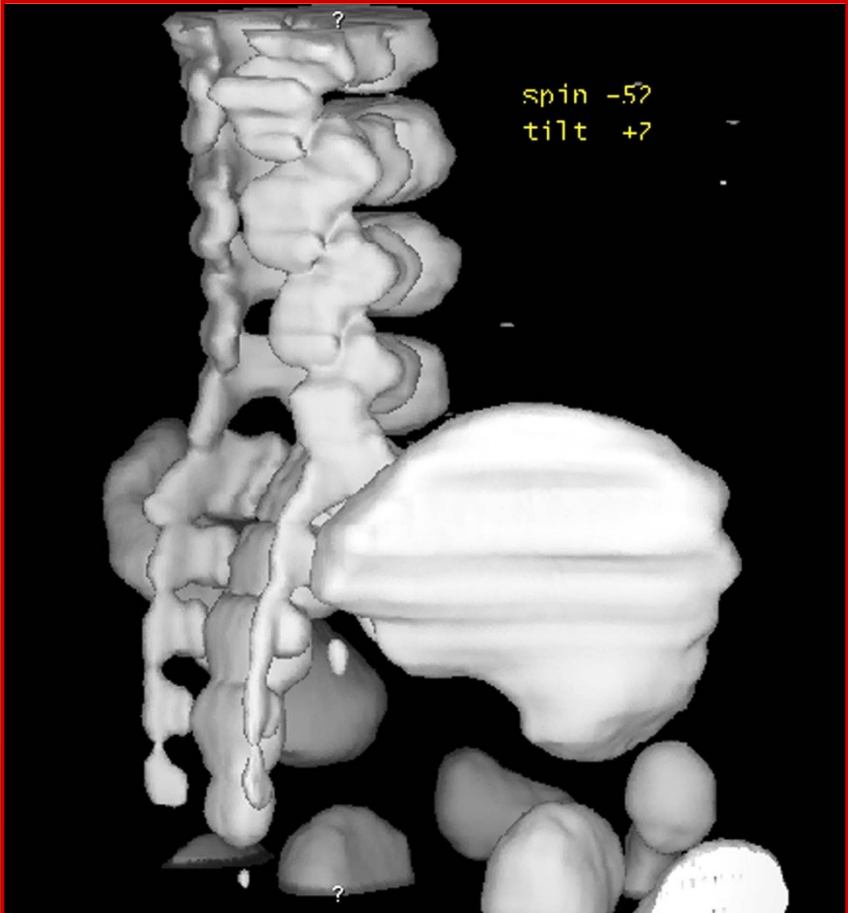
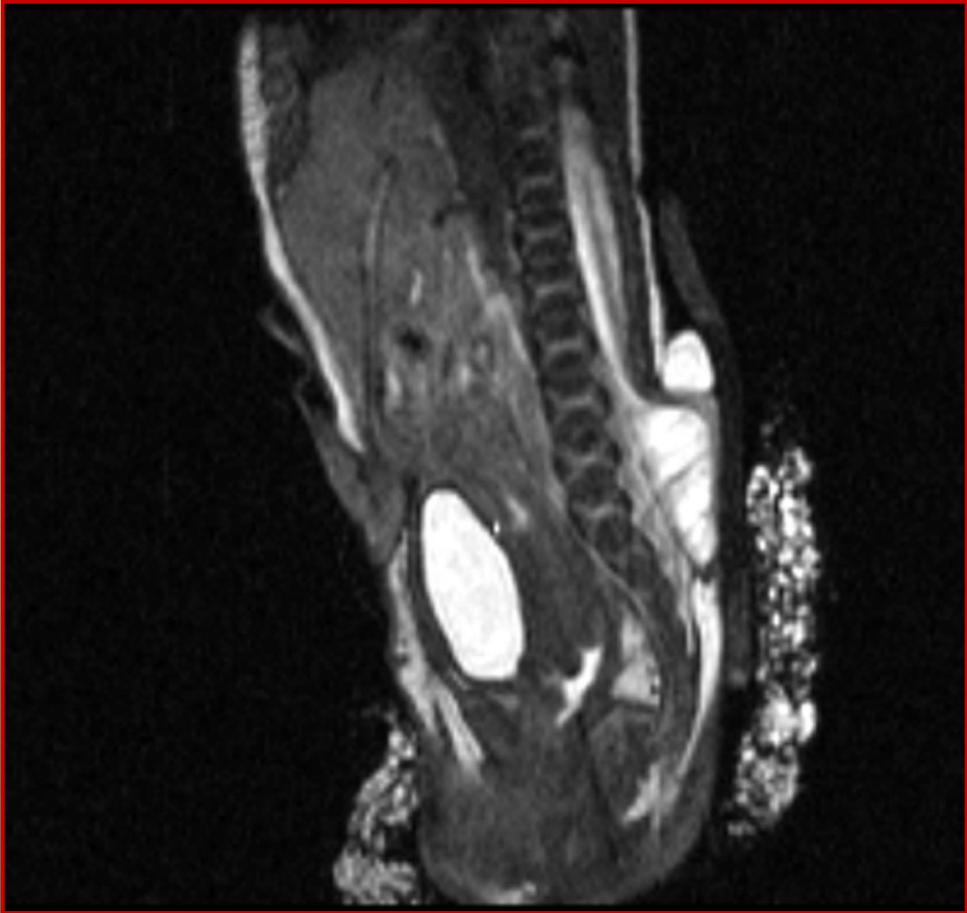
Meningokéla



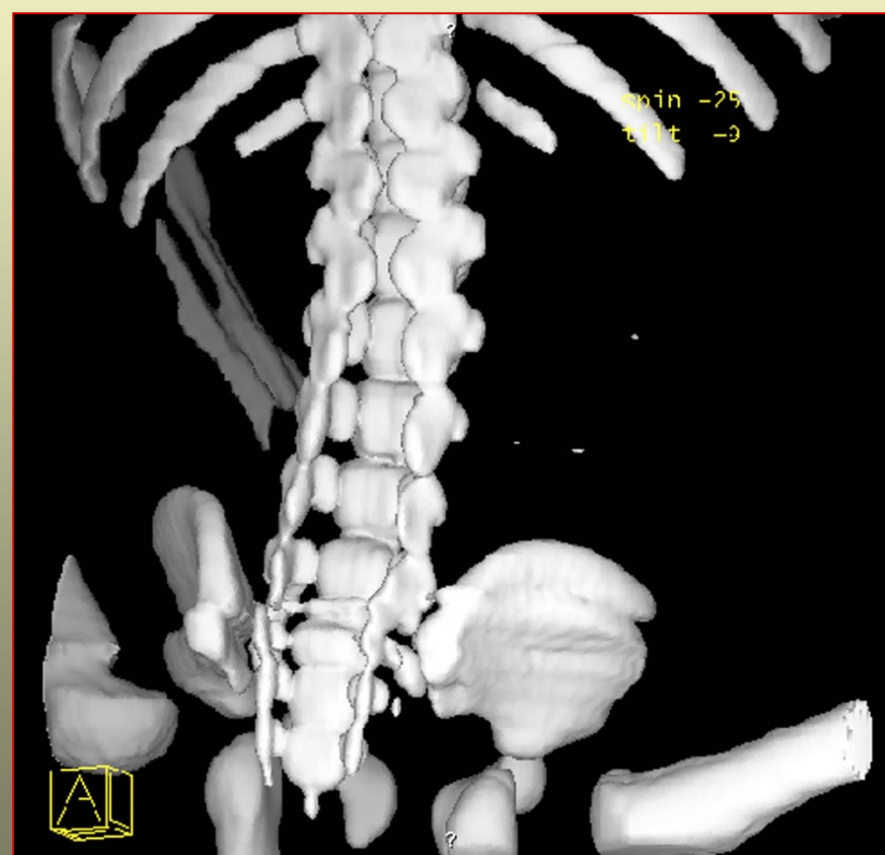
Spinal dysraphism



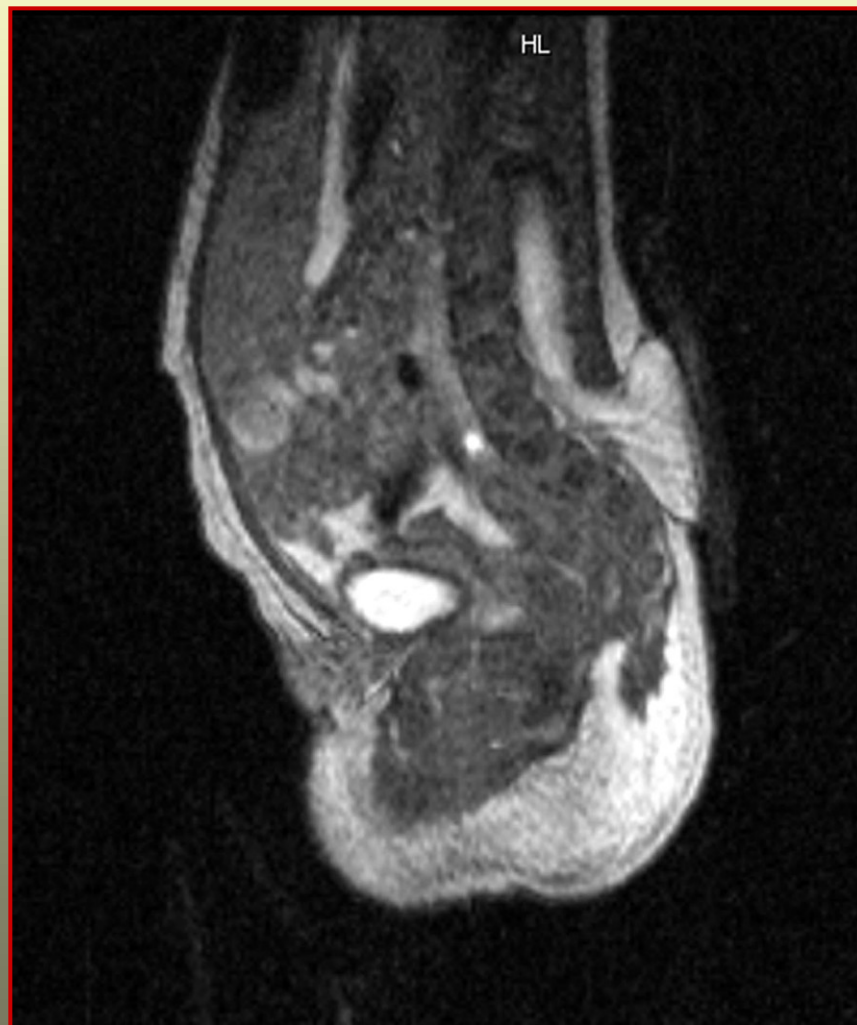
Spinal dysraphism



Spinal dysraphism



Spinal dysraphism



Spinal dysraphism



Spinal dysraphism



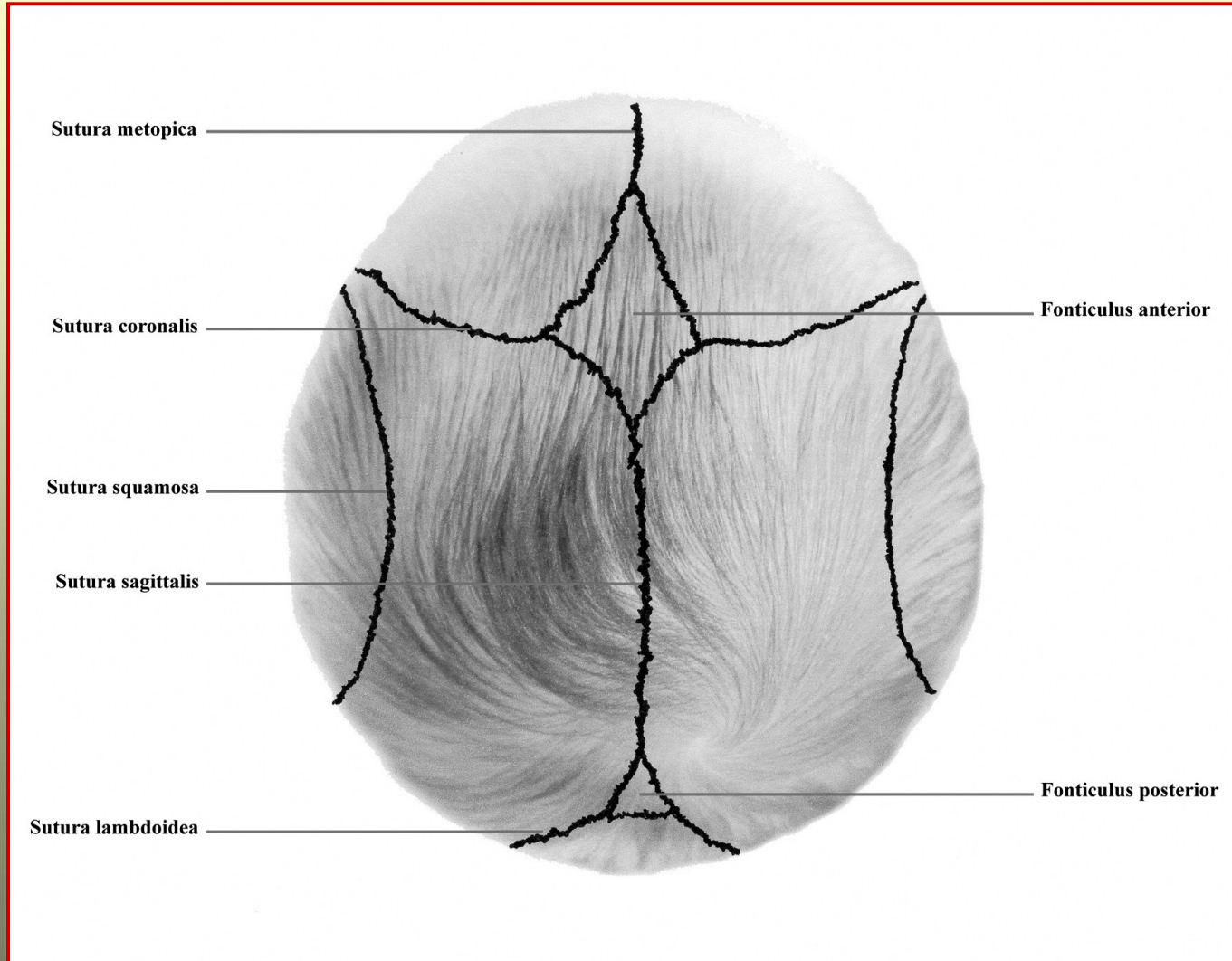
Craniostenosis (caniosynostosis)

Premature skull sutures synostosis.

1852 Rudolf Virchow

1 from 2100 children

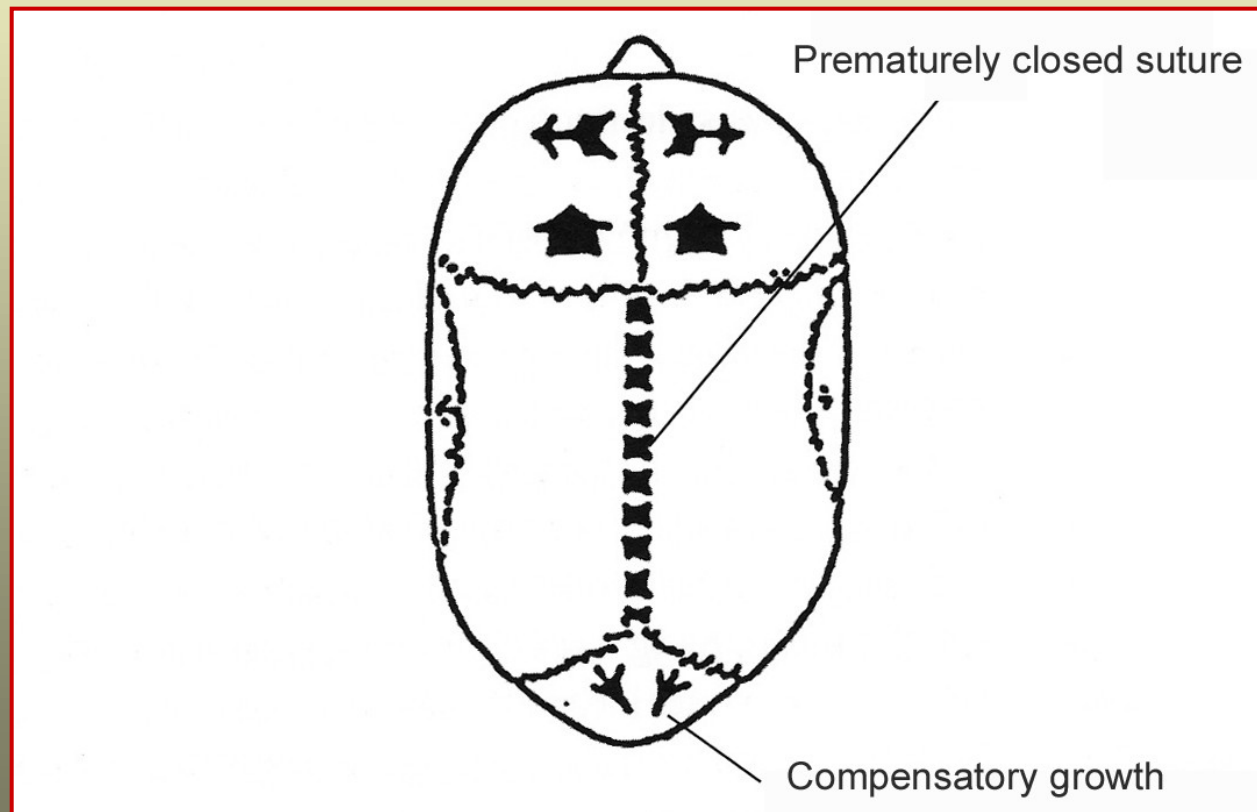
Sutures of the skull



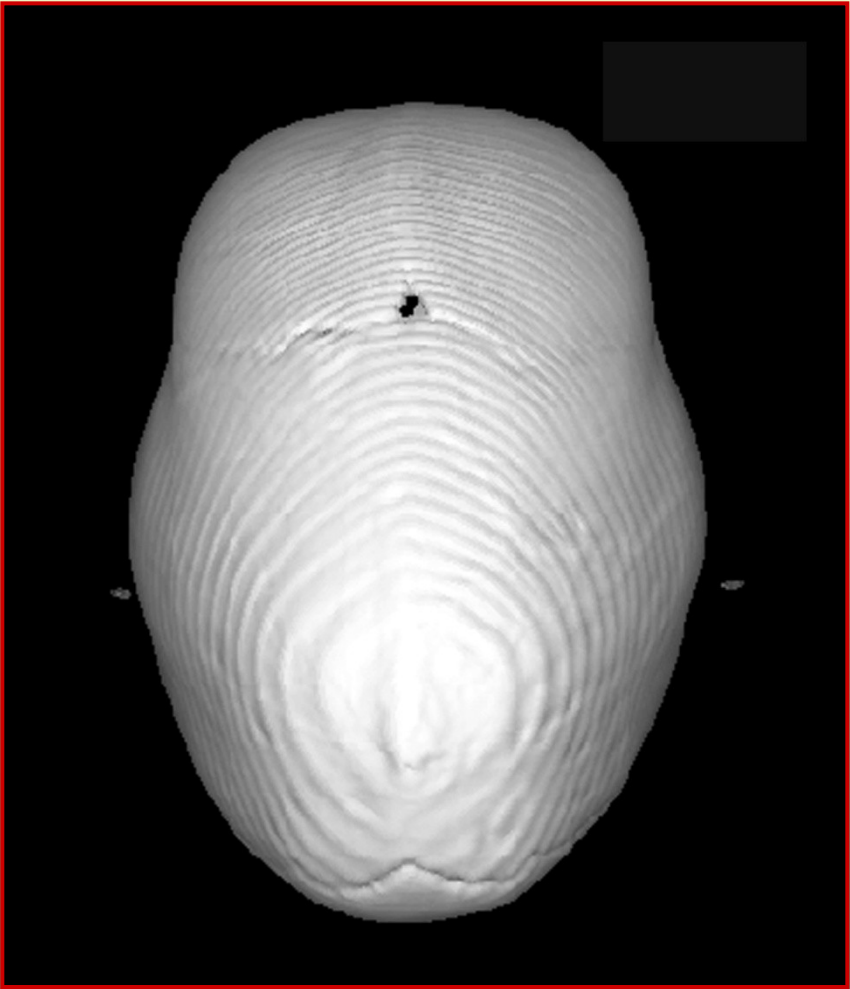
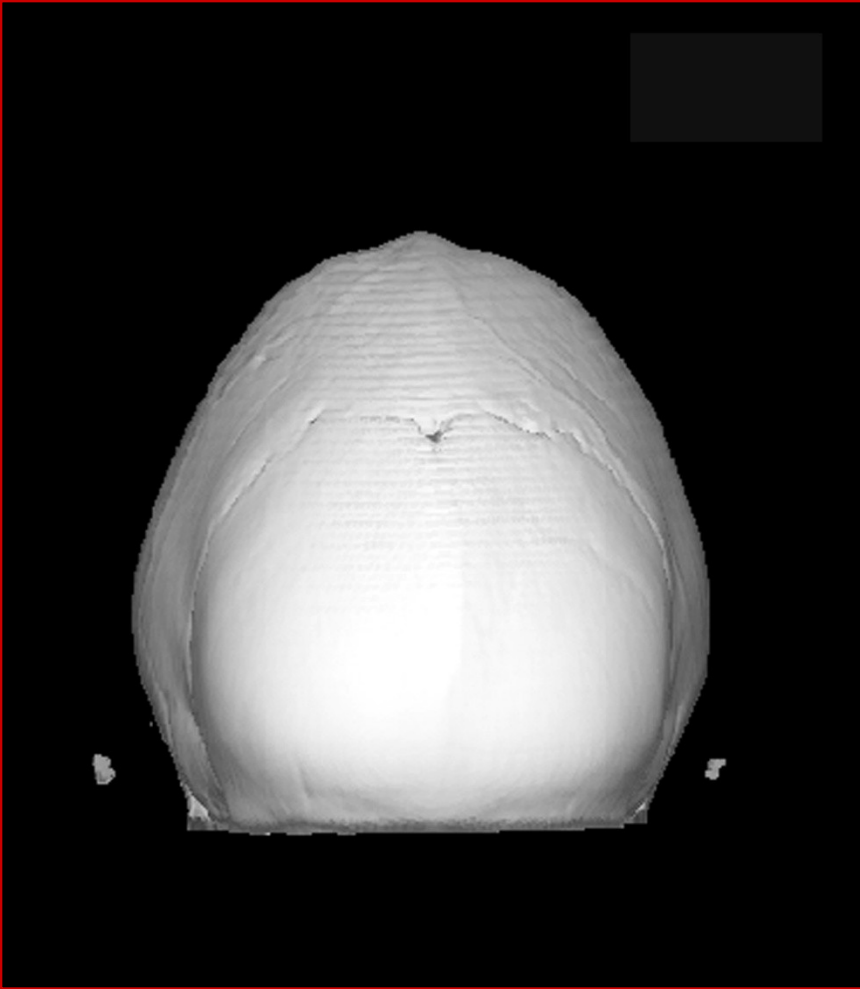
1. Skaphocephalia - dolichocephalia

Premature sagittal suture synostosis

40-60%



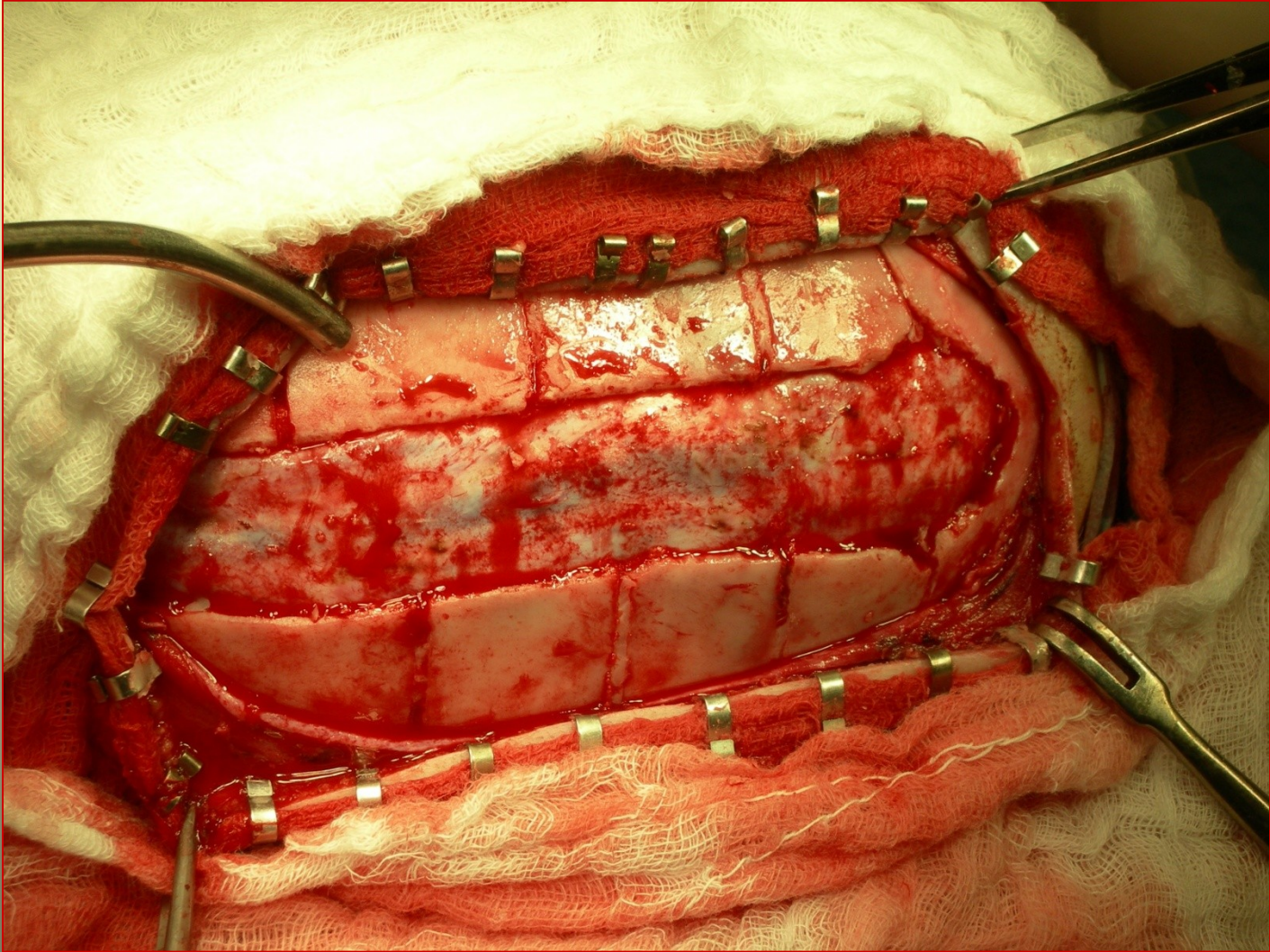
Skaphocephalia - dolichocephalia



Skaphocephalia - dolichocephalia



Skaphocephalia - dolichocephalia



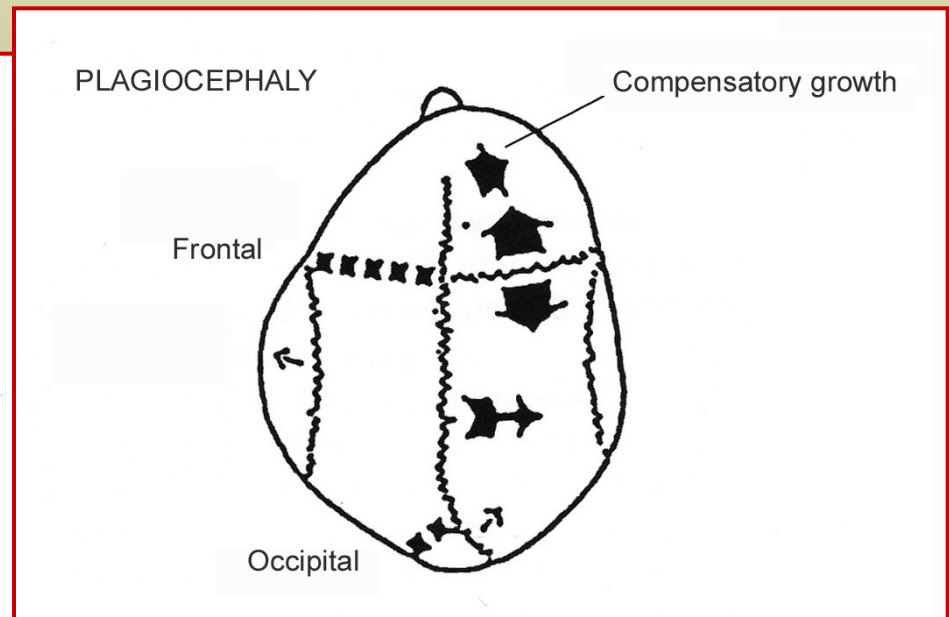
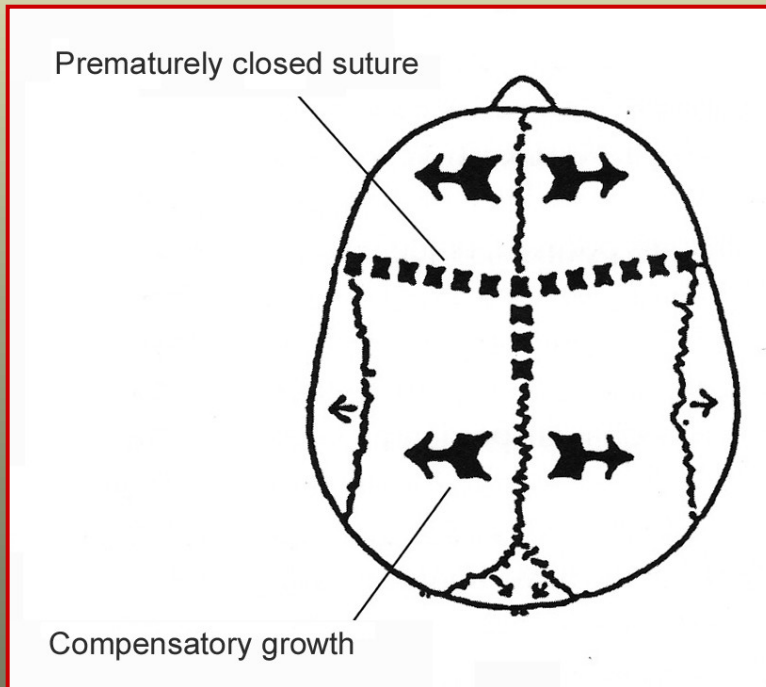
2. Brachycephalia

Premature coronar suture synostosis

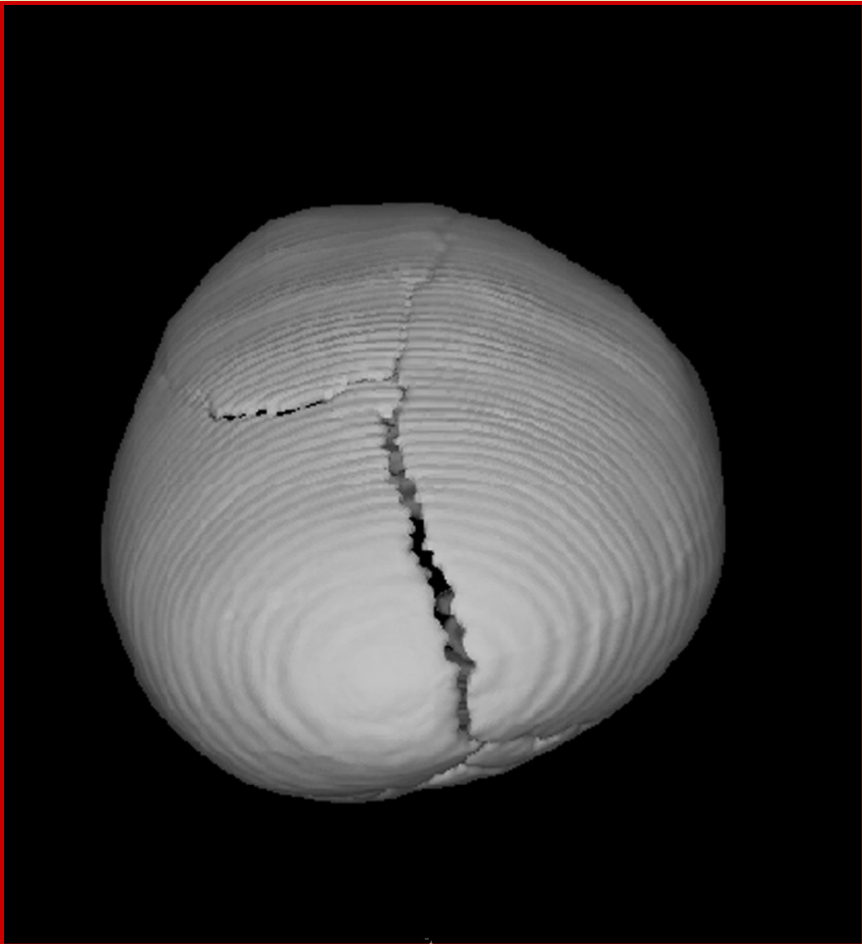
20-30%

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

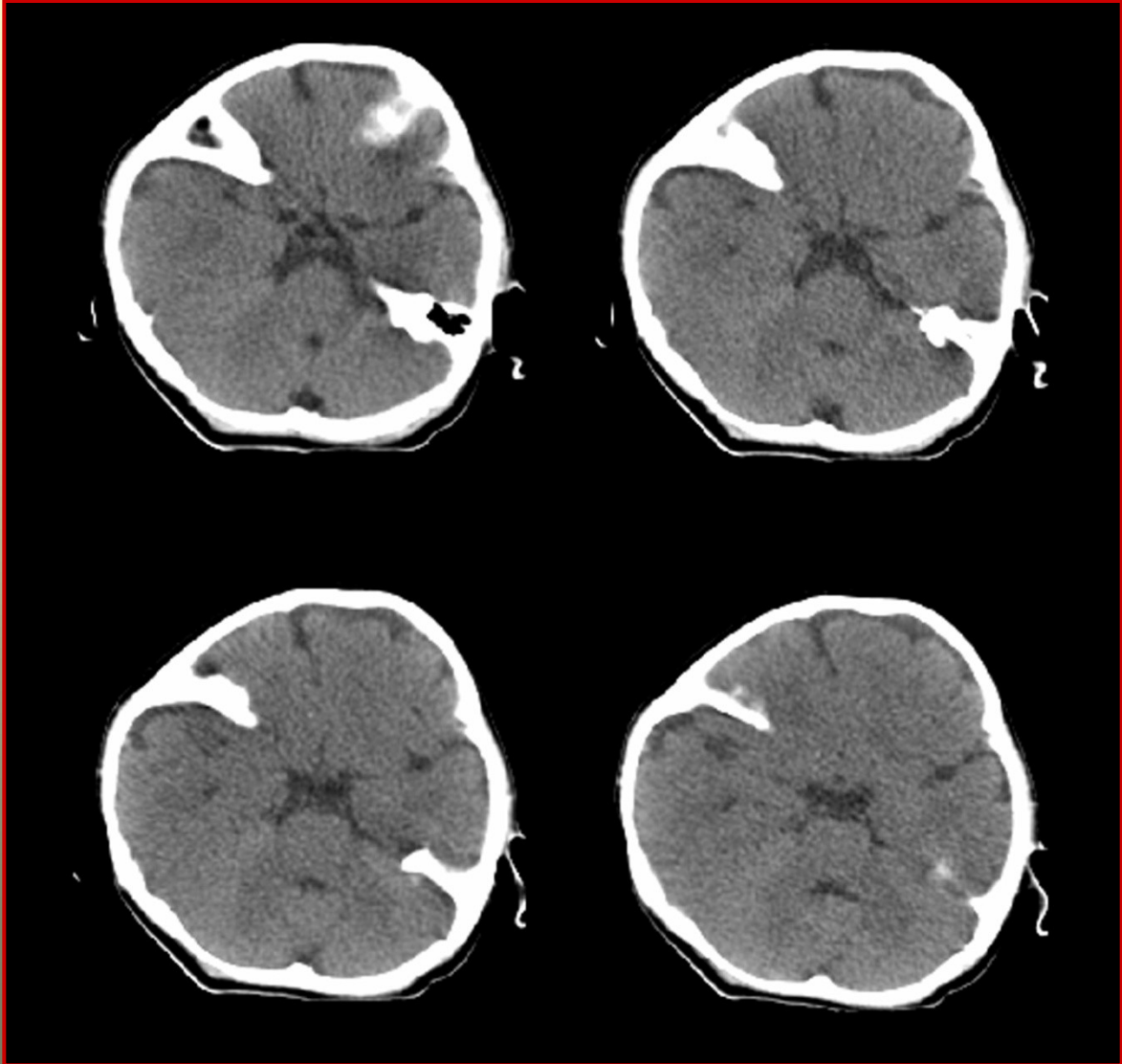
b) **Occipital plagiocephalia** – one side lambdoid suture



Brachycephalia



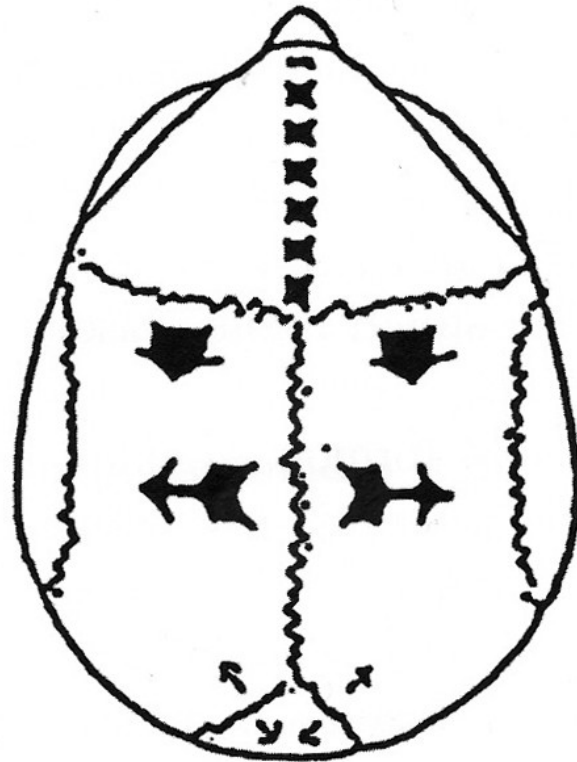
Brachycephalia



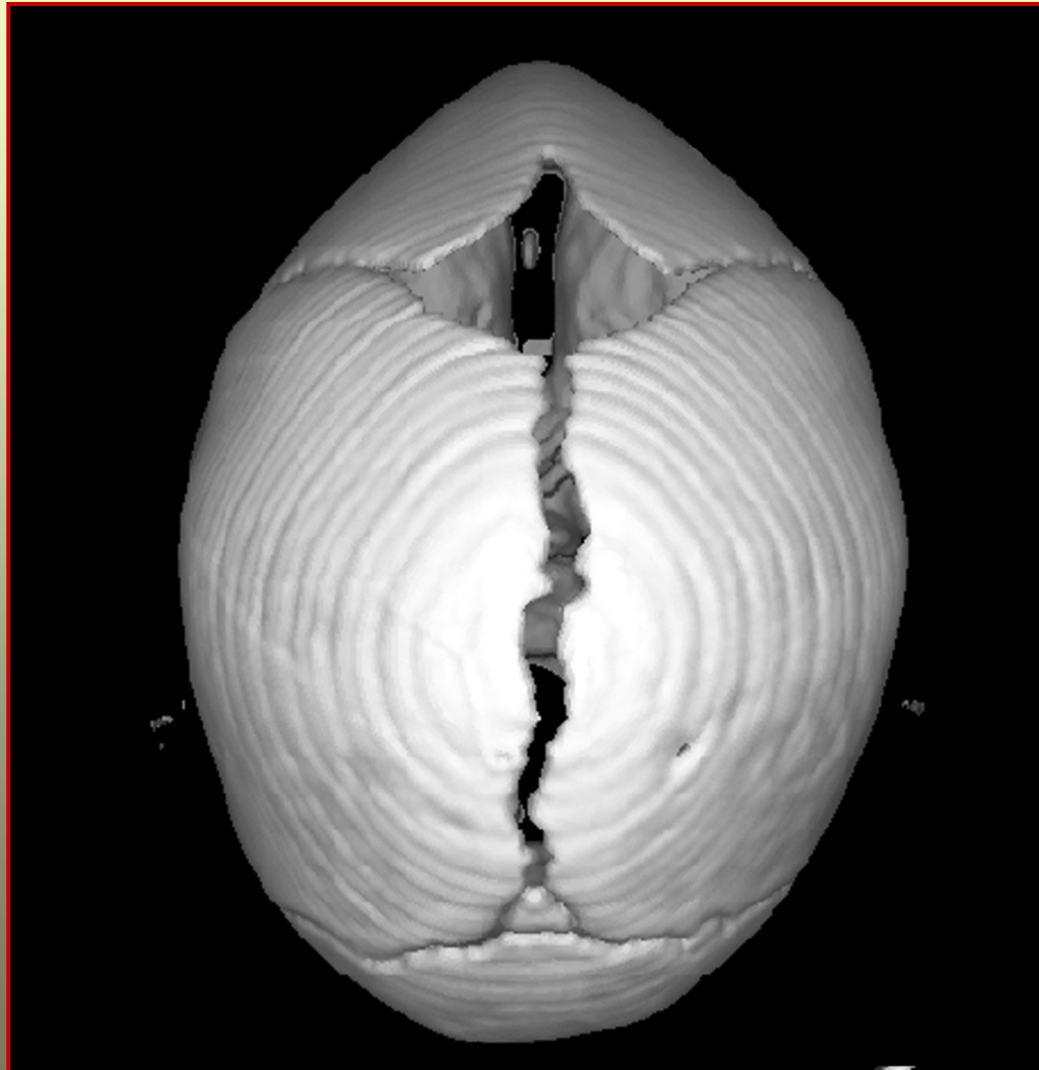
3. Trigonocephalia

Premature sutura metopica synostosis

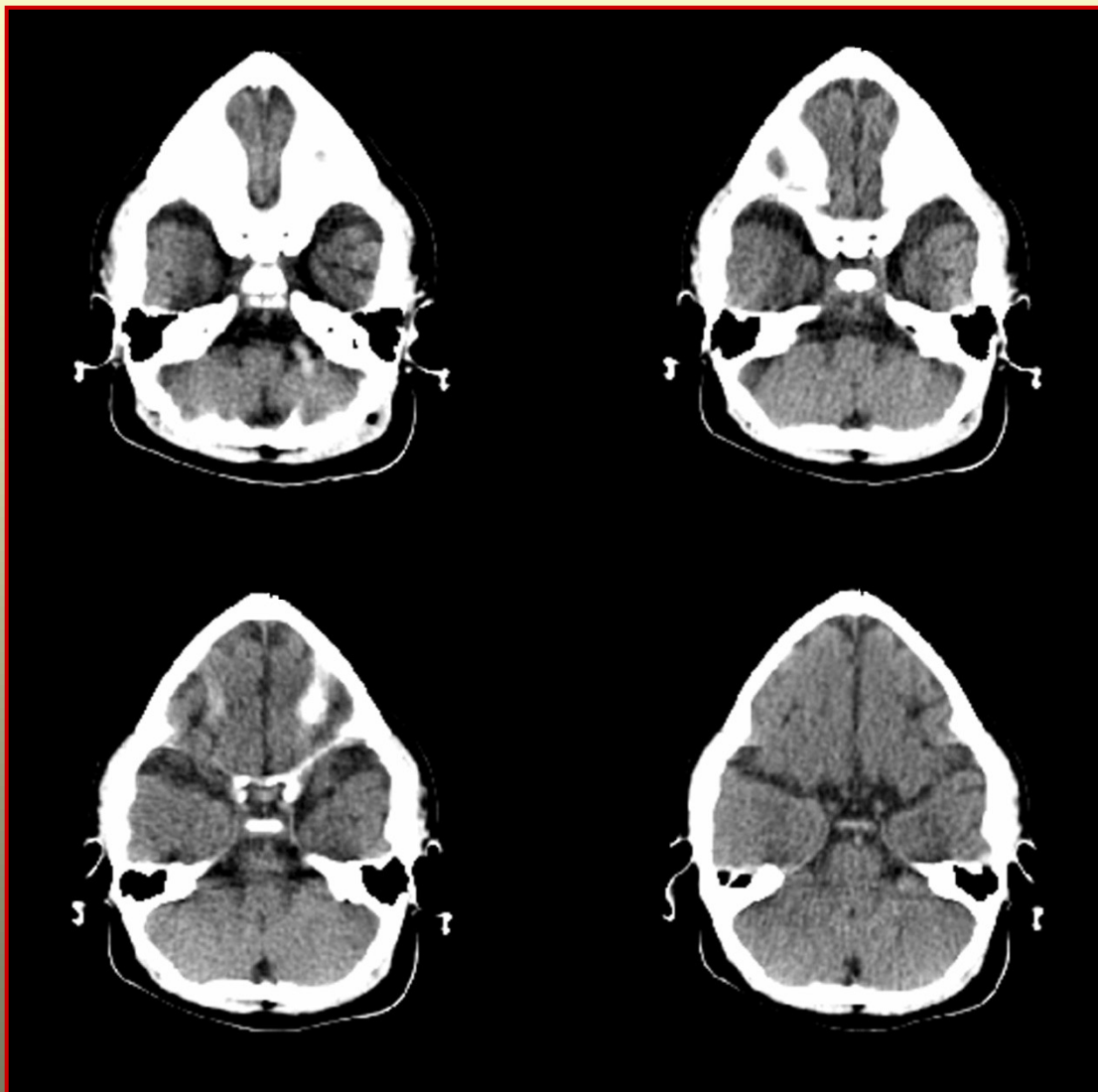
10%



Trigonocephalia



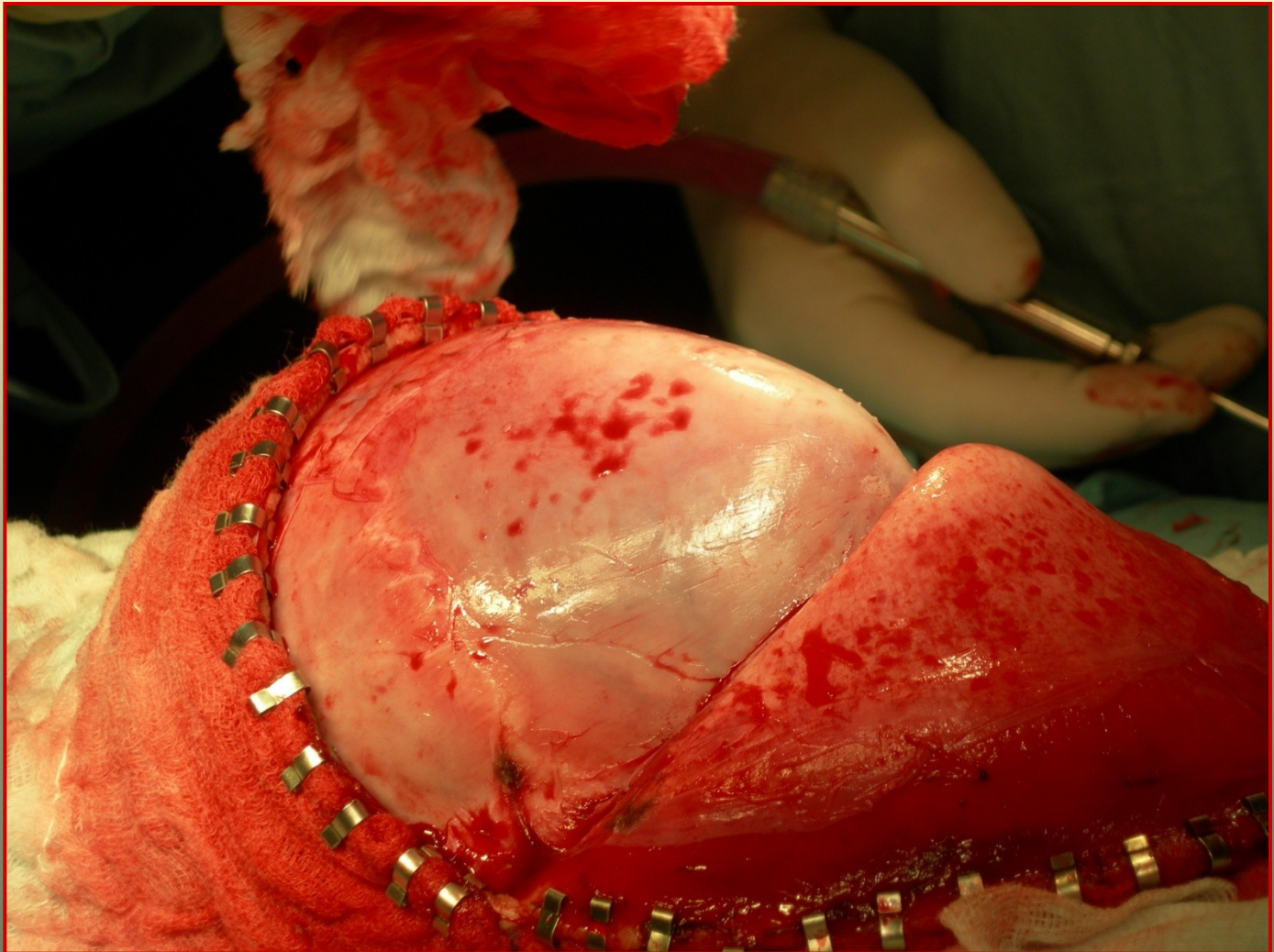
Trigonocephalia



Trigonocephalia



Trigonocephalia



Trigonocephalia



Trigonocephalia



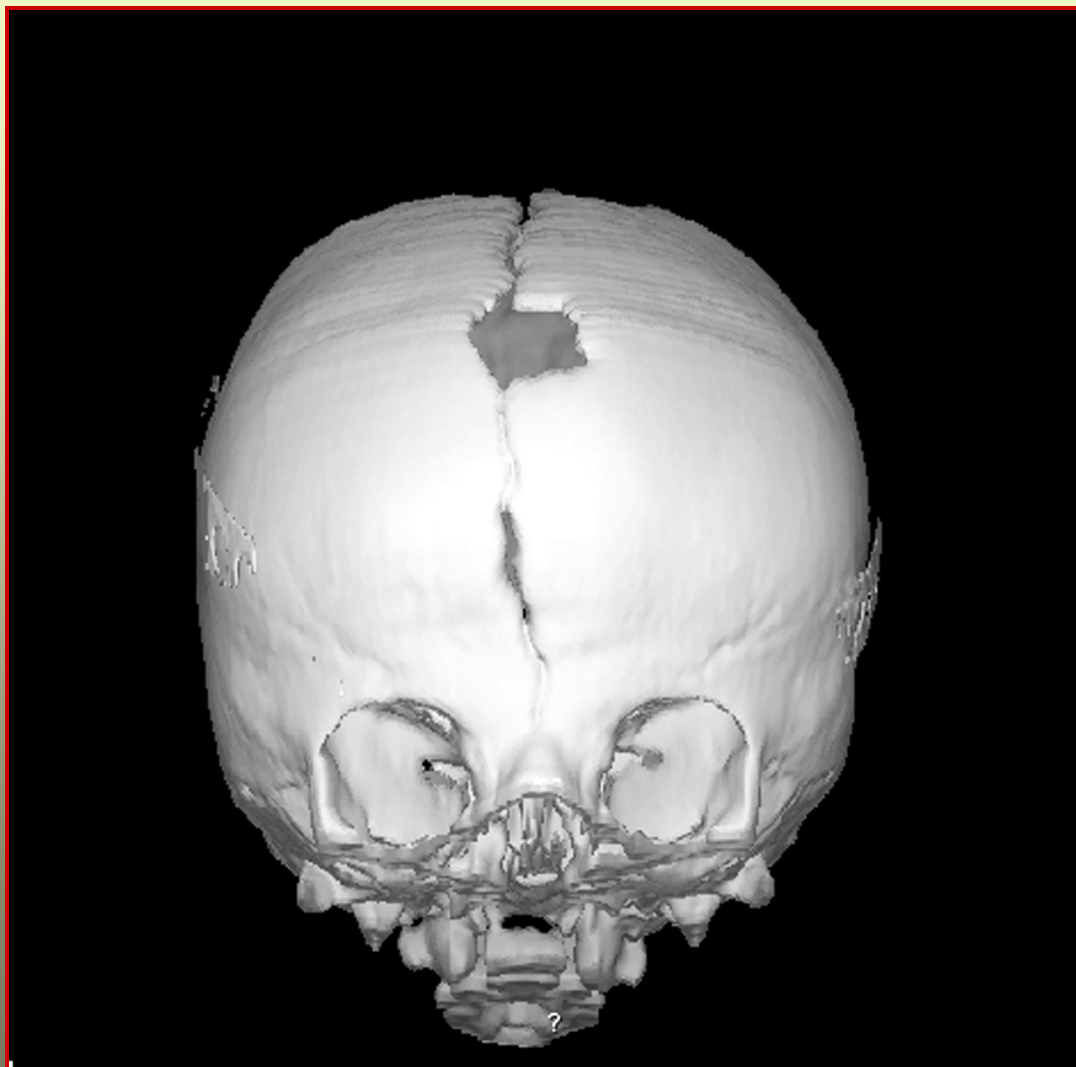
4. Morbus Crouzon – dysostosis craniofacialis

(1912)

- turicephaly
- shallow orbits
- exophthalmos
- hypertelorism
- hypoplasia of middle facial skelet

1 from 25 000 children

Morbus Crouzon



Morbus Crouzon



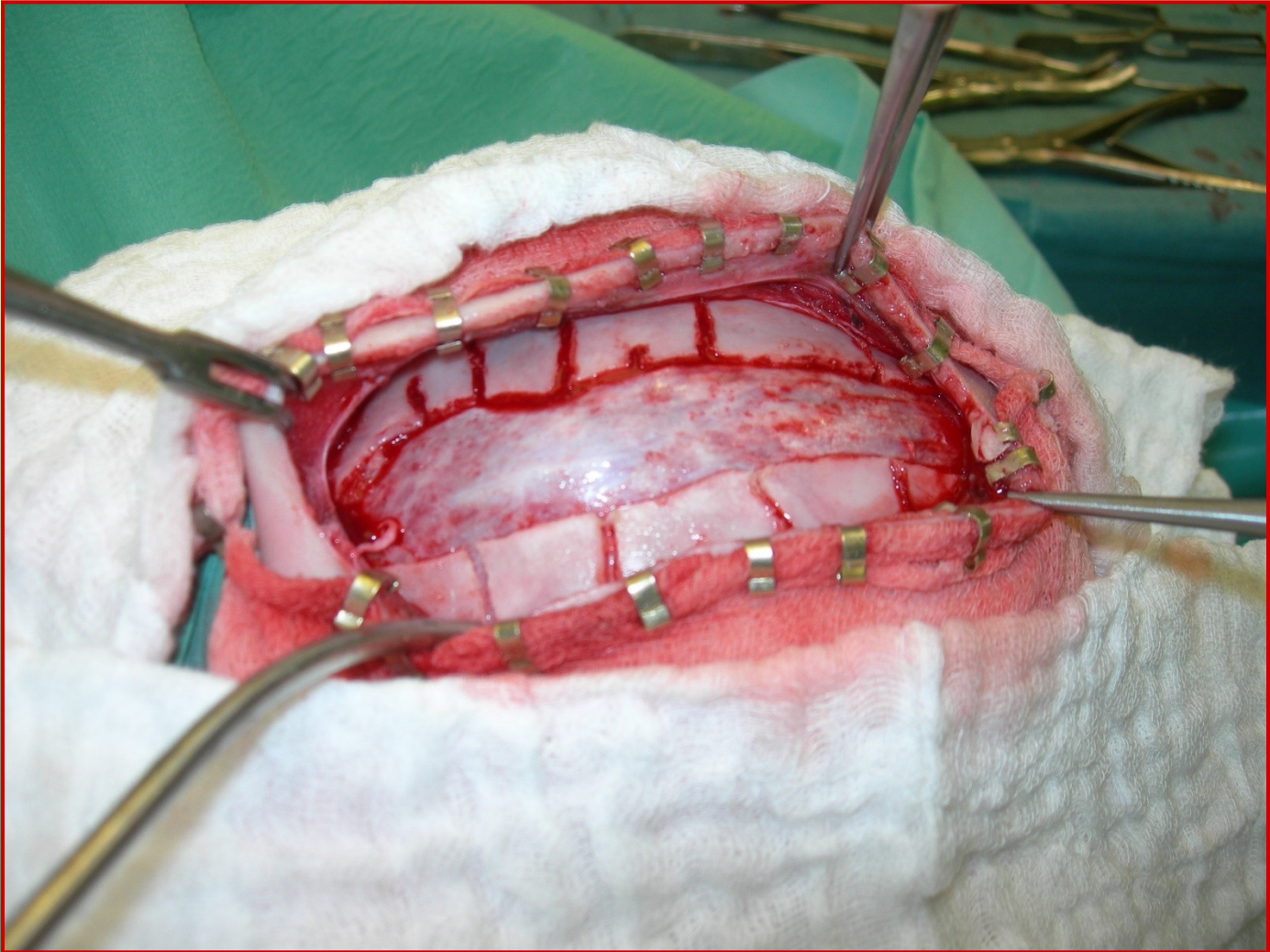
Morbus Crouzon



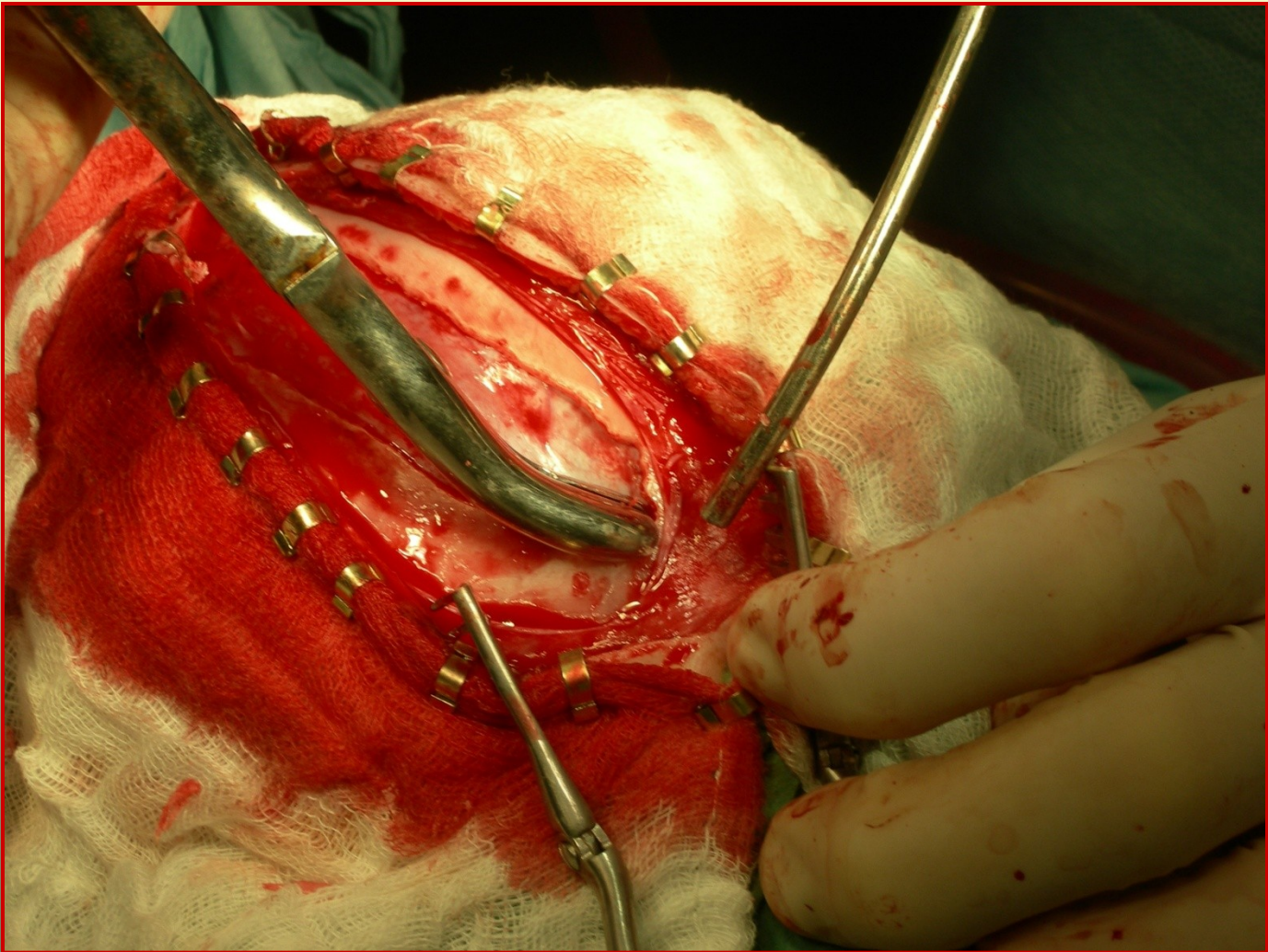
Morbus Crouzon



Morbus Crouzon



Morbus Crouzon



Post surgery care – preventive protective helmets

