Central Nervous System Congenital Abnormalities

Eva Brichtova

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:

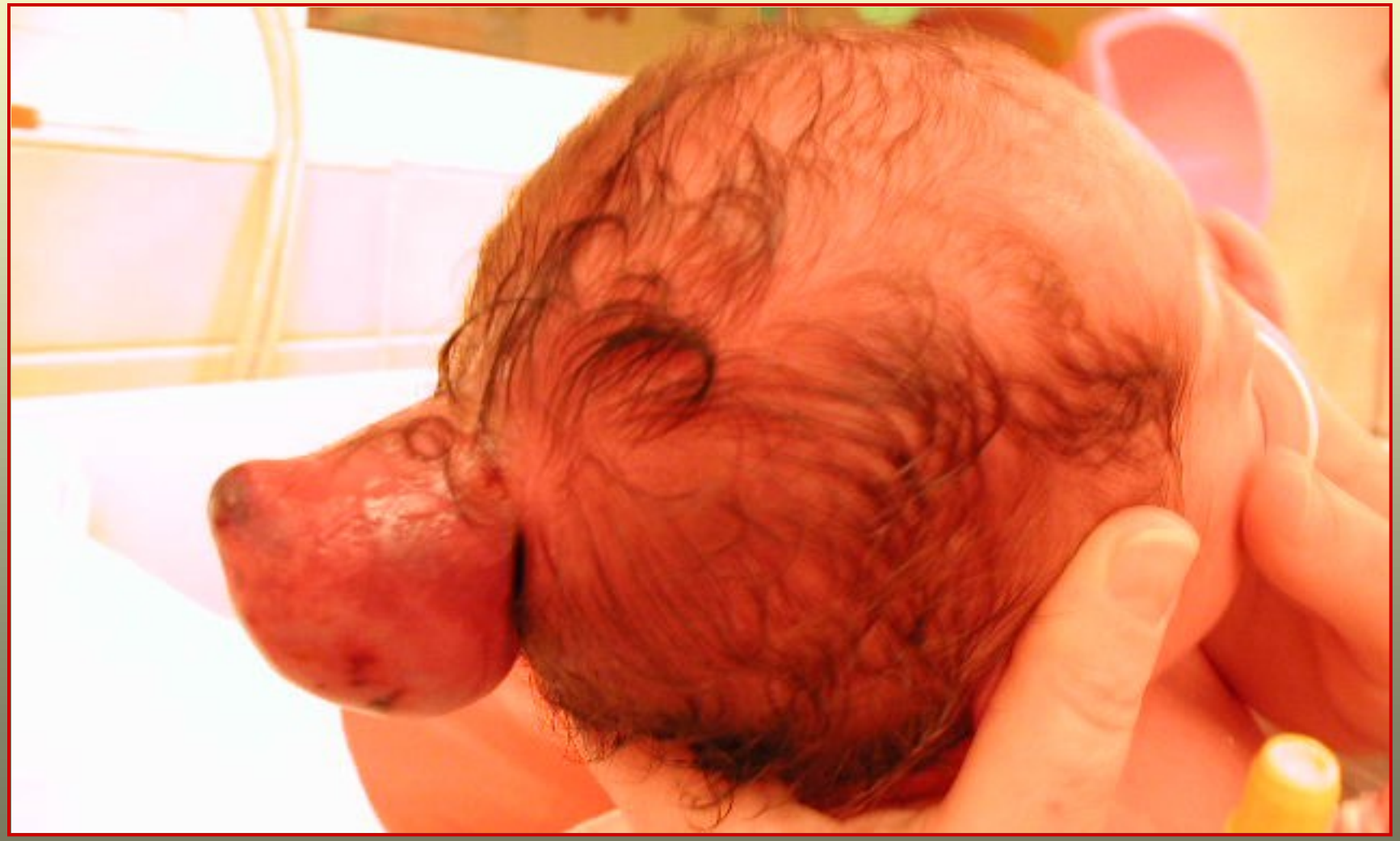
a) cranial meningocele - dura mater and CSF herniation
b) encephalocele – cerebral tissue herniation
c) anencephaly - open dysraphism, without calvaria bones

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 – 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

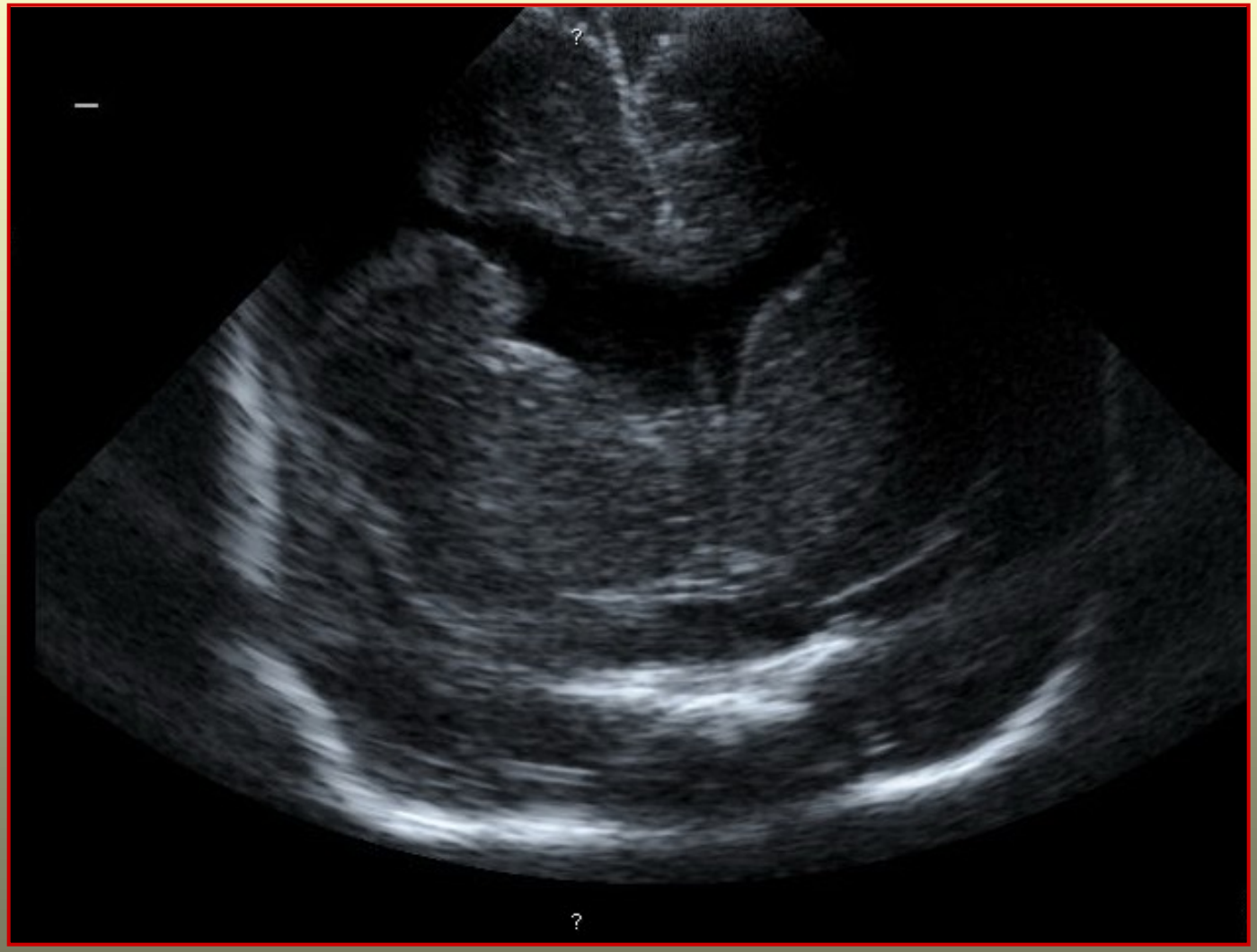
f) agenesis of corpus callosum

g) Dandy-Walker syndrom (cerebellar hypoplasia)

h) macroencephaly - megalencephaly

i) schizencefaly

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:

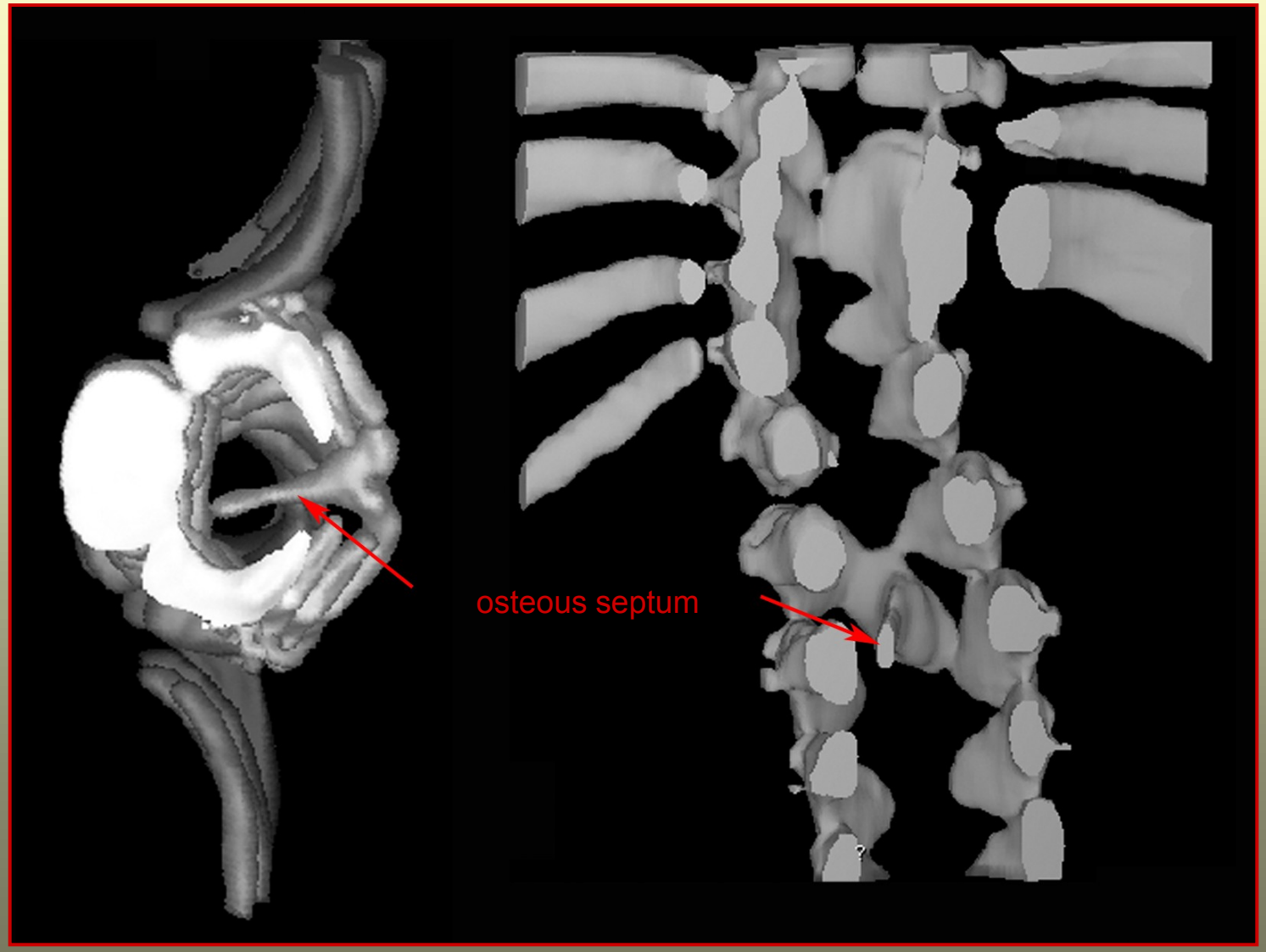
Syringomyelia

Diastematomyelia

Tethred cord syndrom

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

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

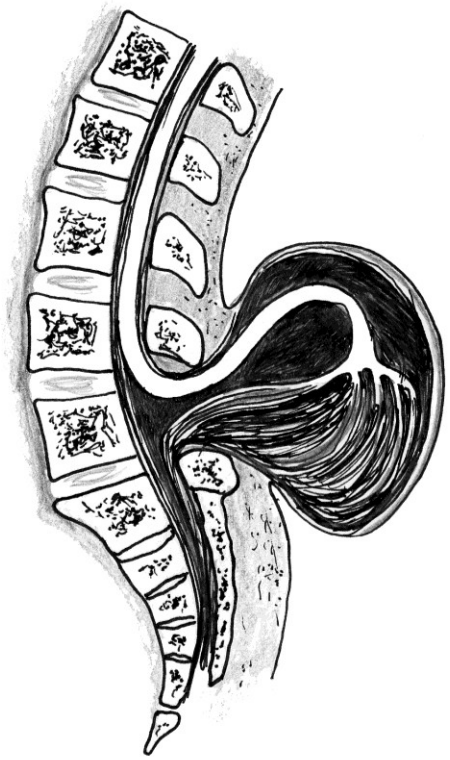
Myelomeningocele epidemiology: 1 from 1000 newborns

Clinical features: lower extremities paresis
proprioceptive reflexes disturbance
incontinence

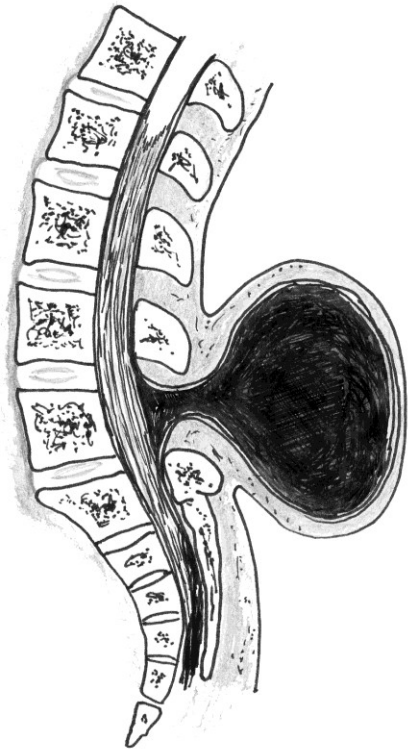
Associated conditions: hydrocephalus (in 65 – 85%)
Chiari malformation (in 80%)

Spinal dysraphism

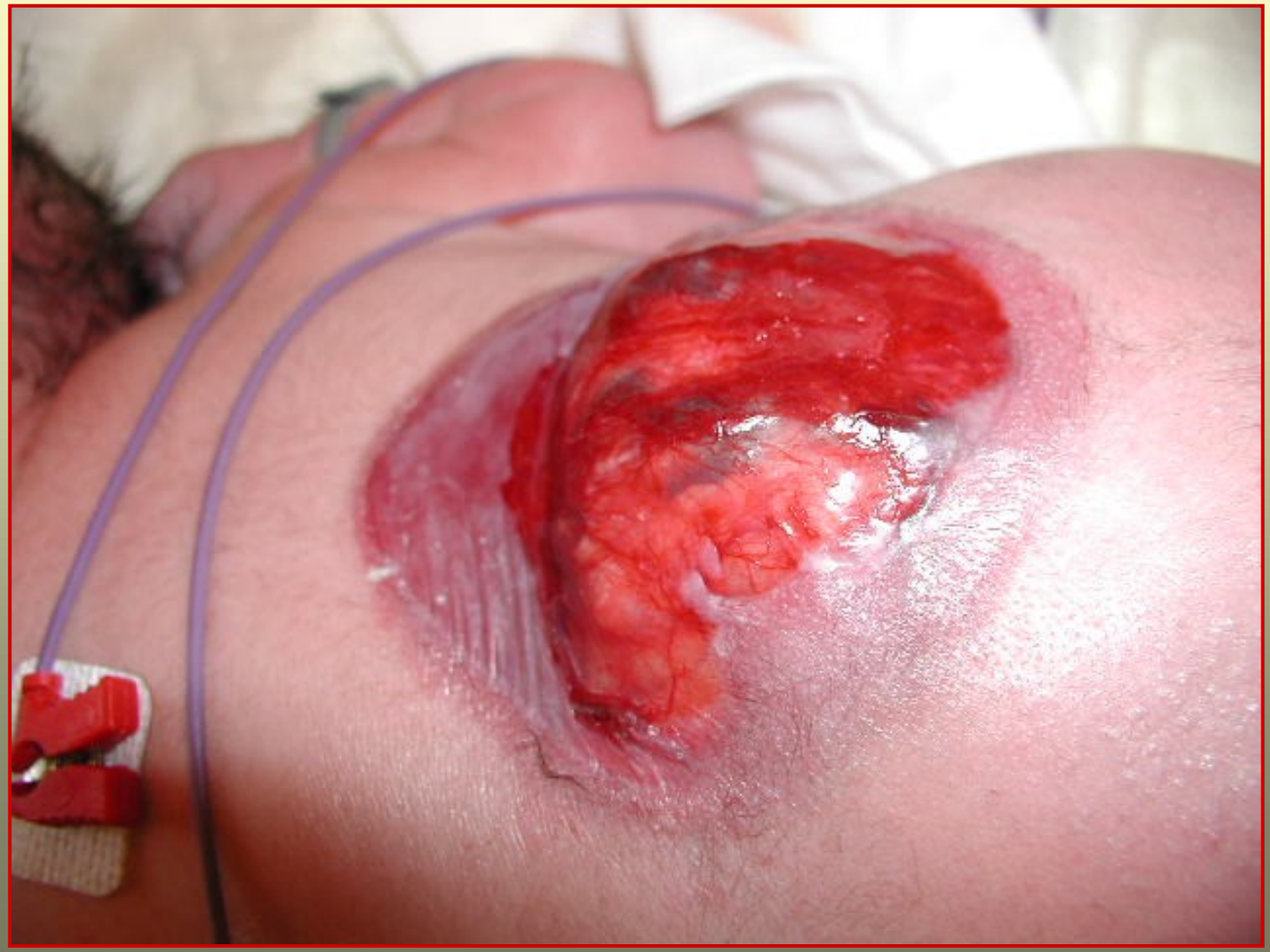
Myelomeningokéla



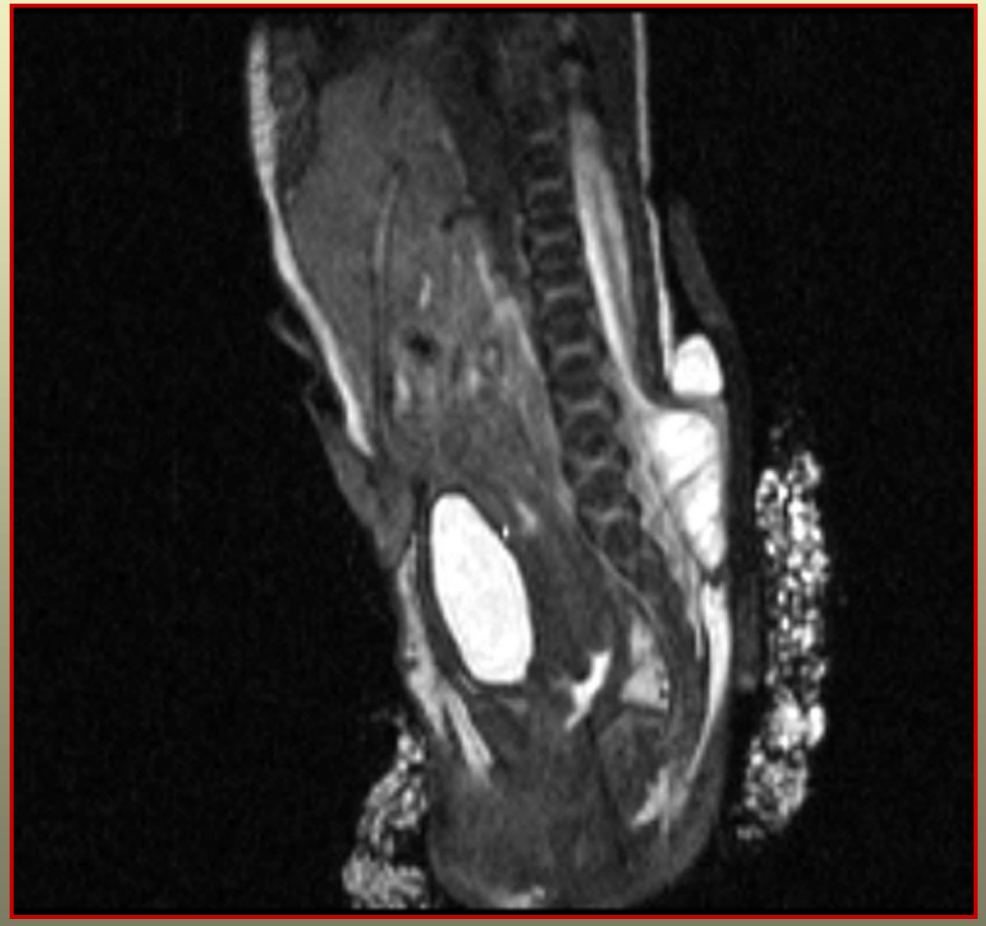
Meningokéla

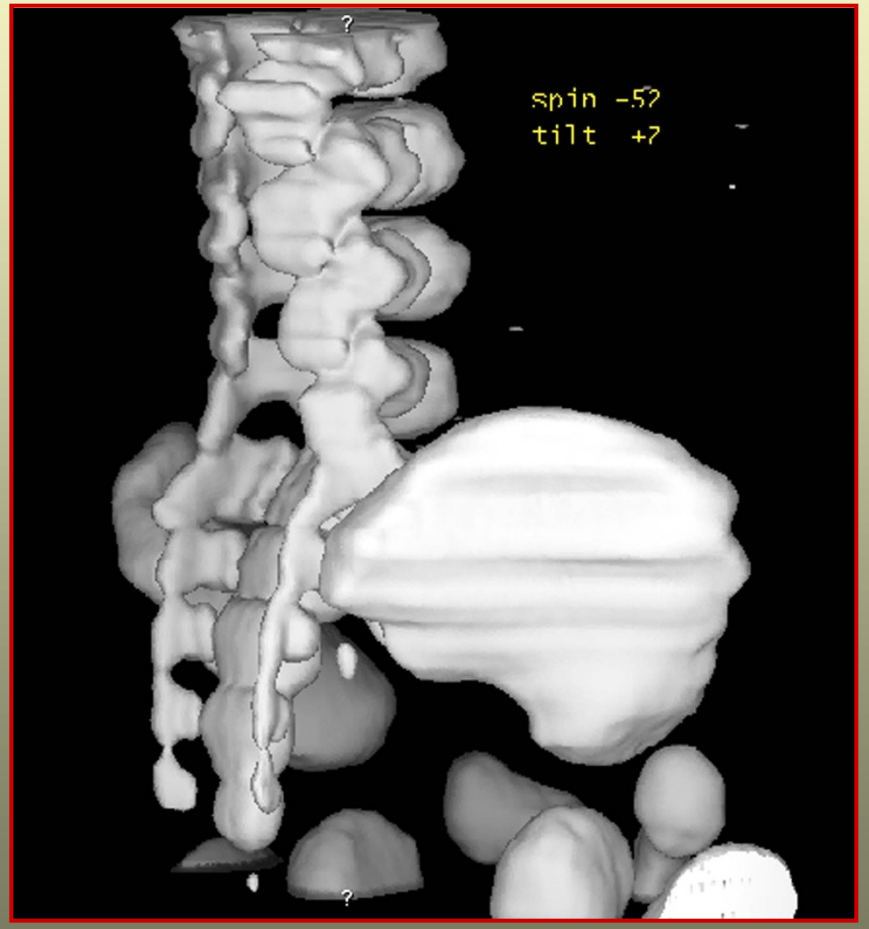


Spinal dysraphism

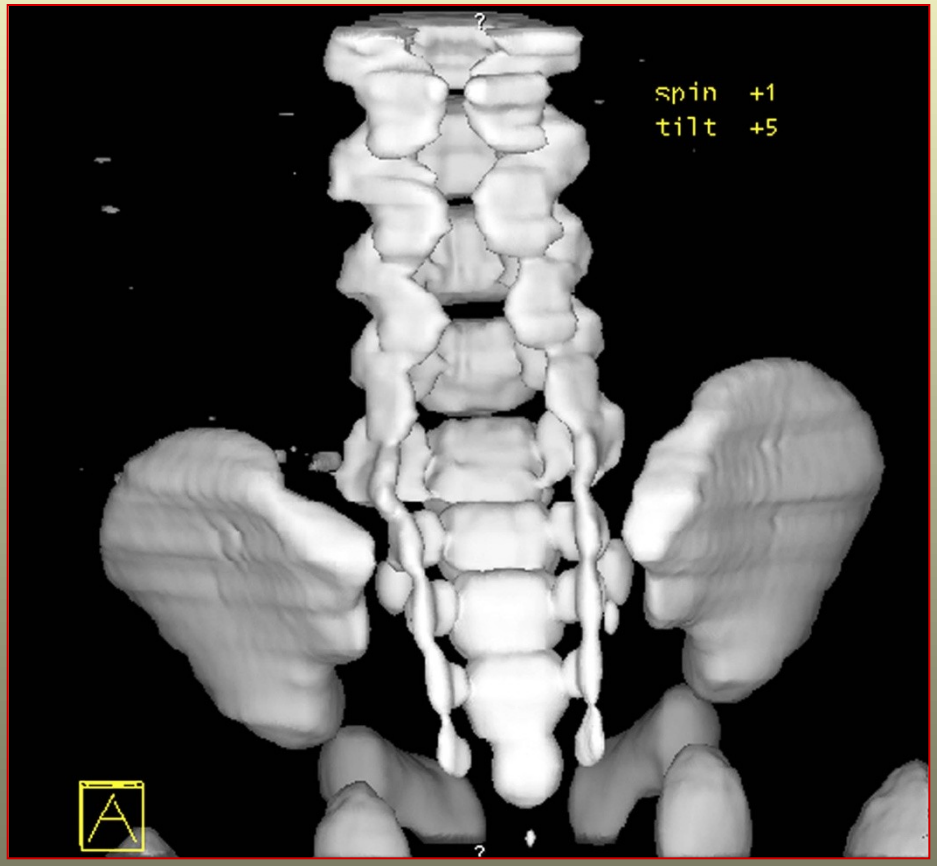


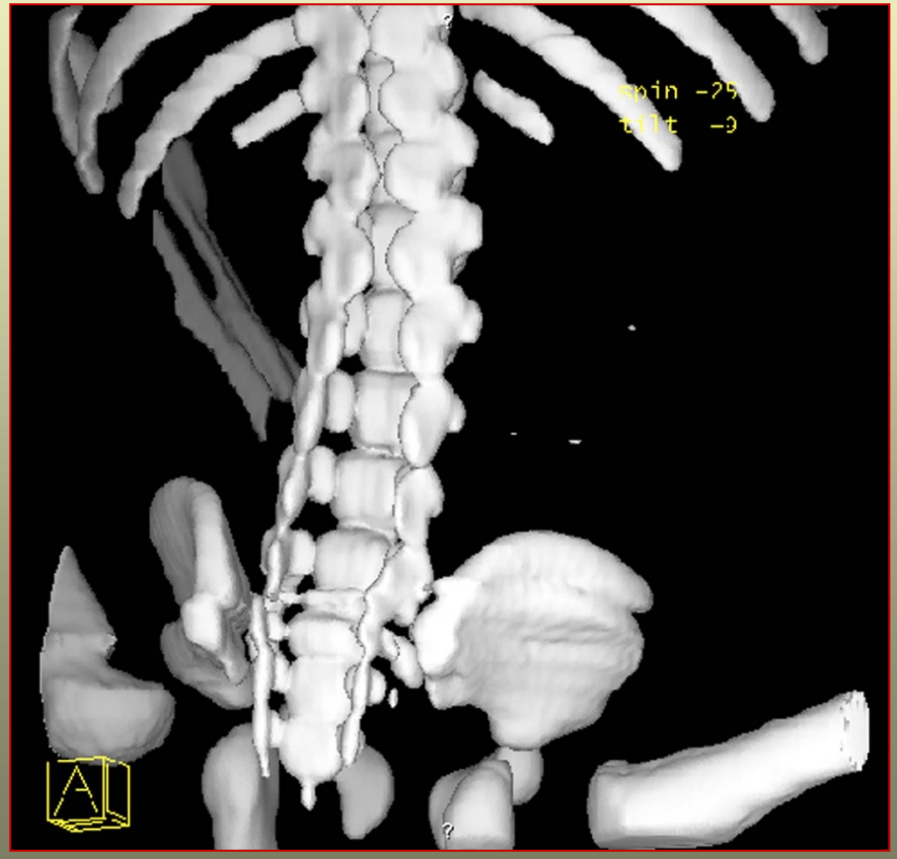
Spinal dysraphism



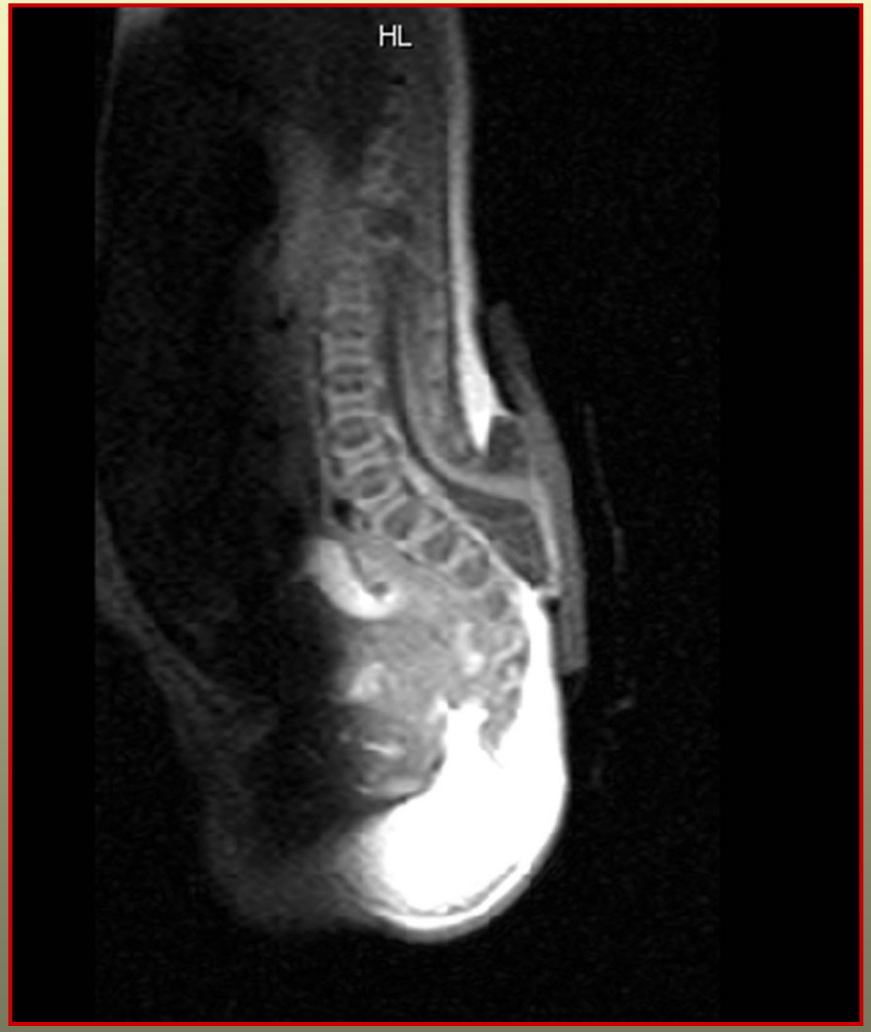


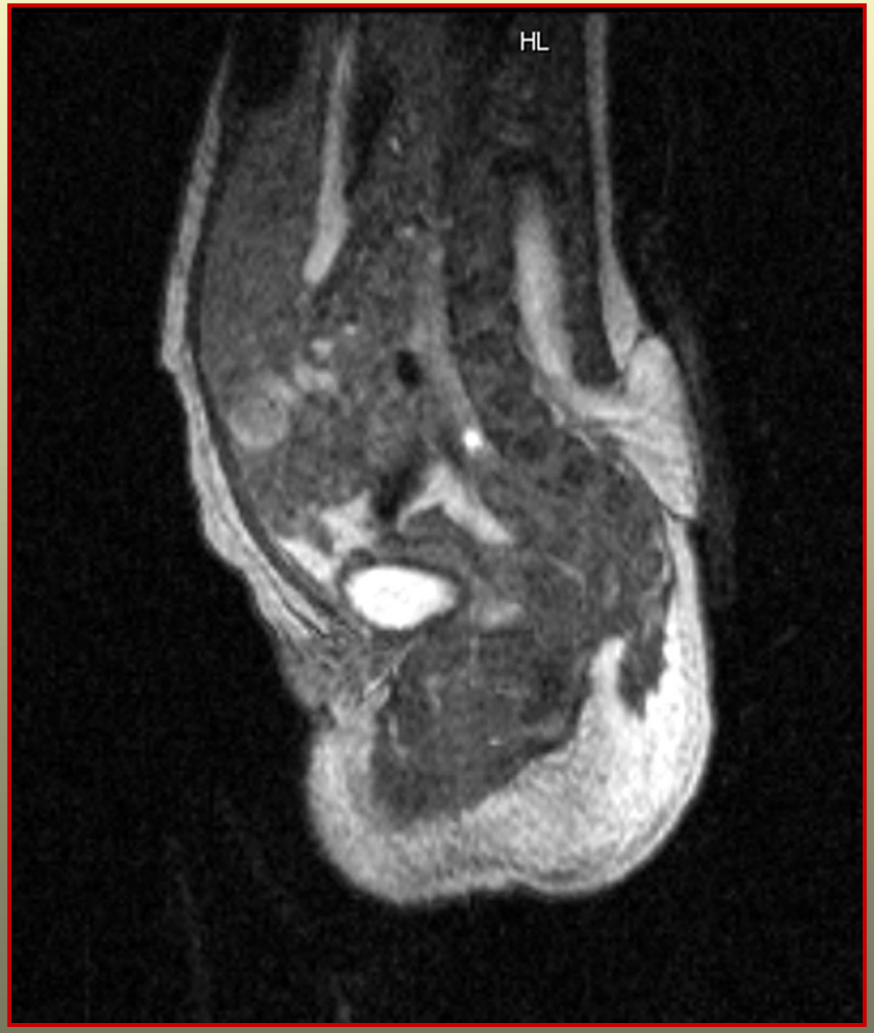
Spinal dysraphism



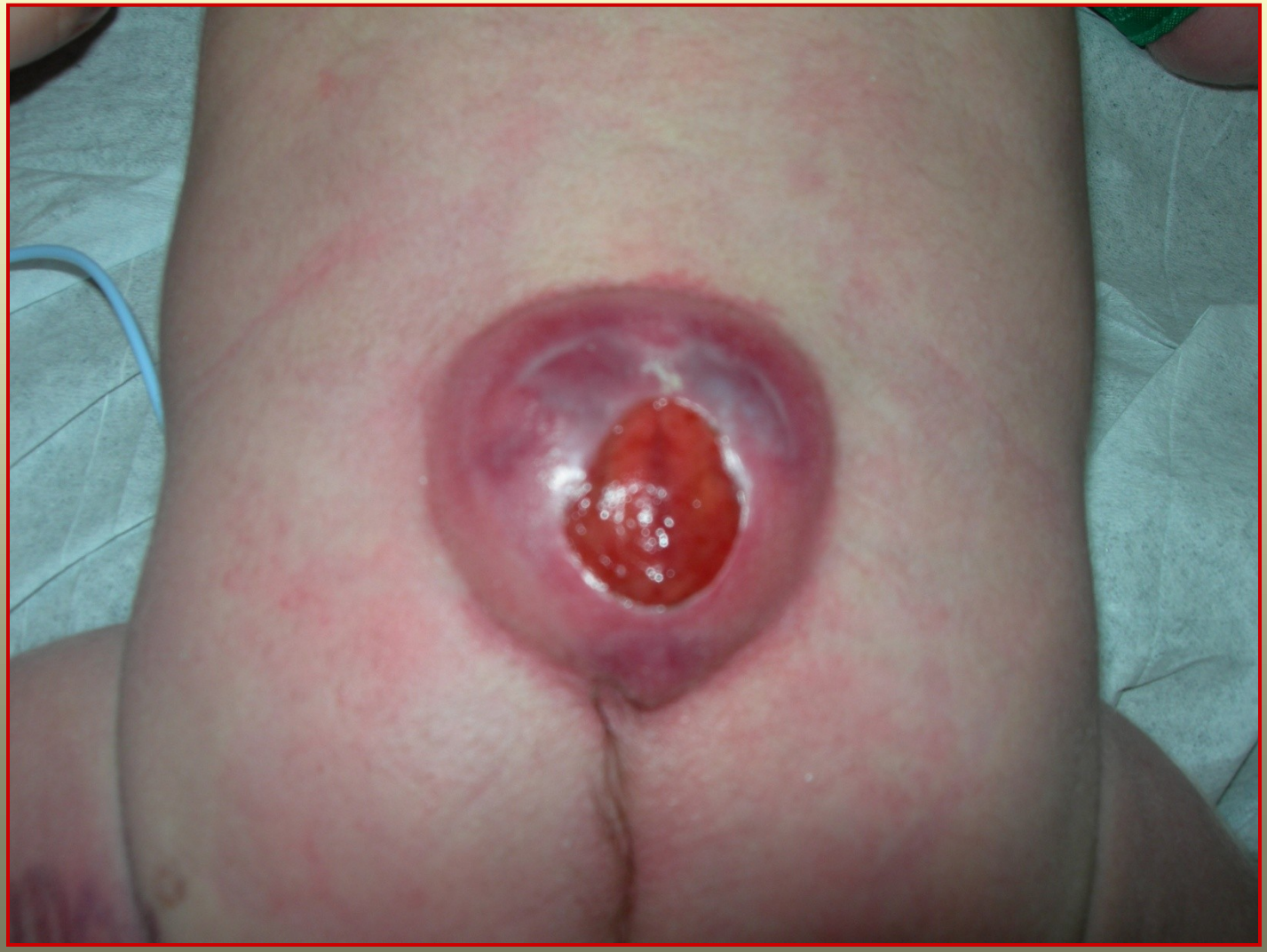


Spinal dysraphism

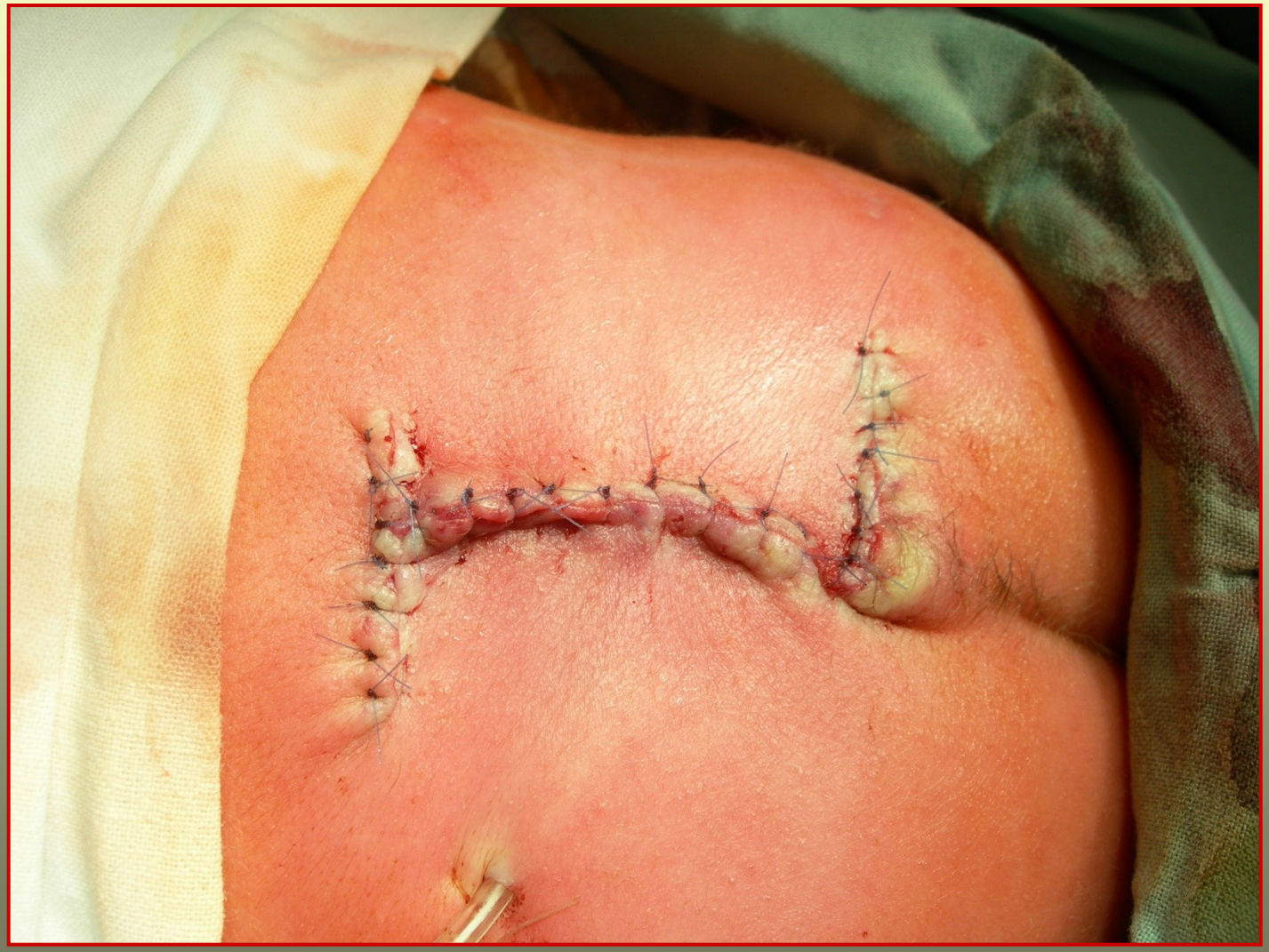




Spinal dysraphism



Spinal dysraphism



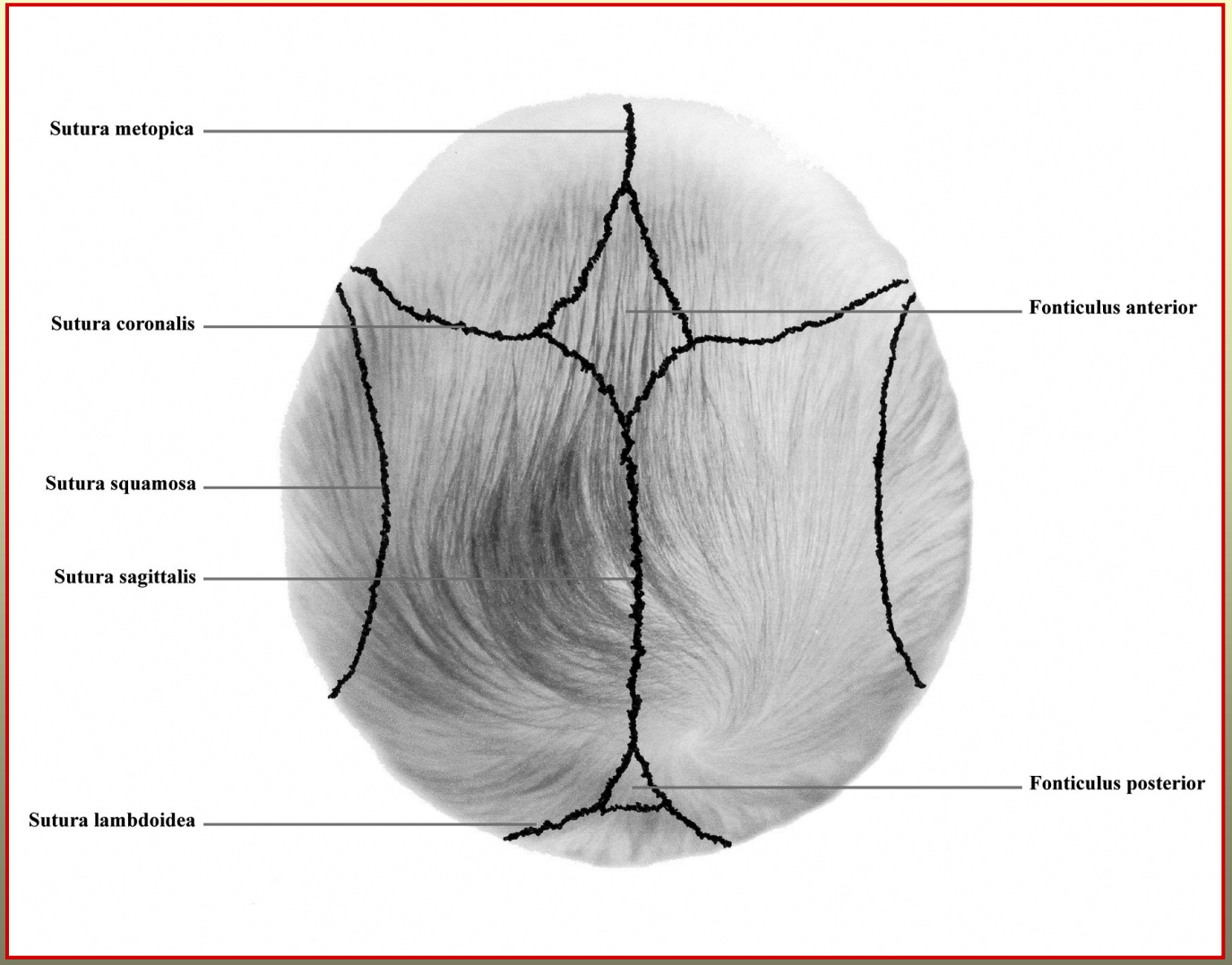
Craniostenosis
(caniosynostosis)

Premature skull sutures synostosis.

1852 Rudolf Virchow

1 from 2100 children

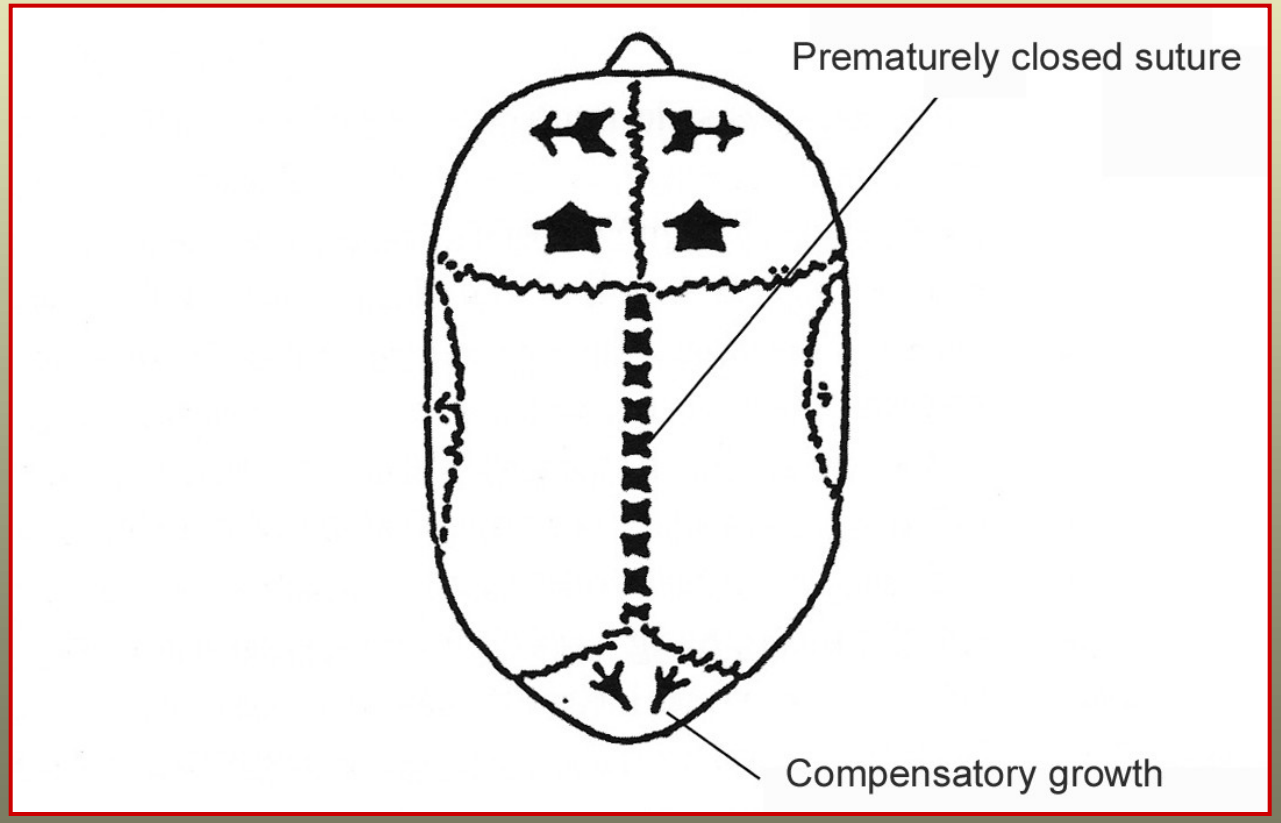
Sutures of the skull



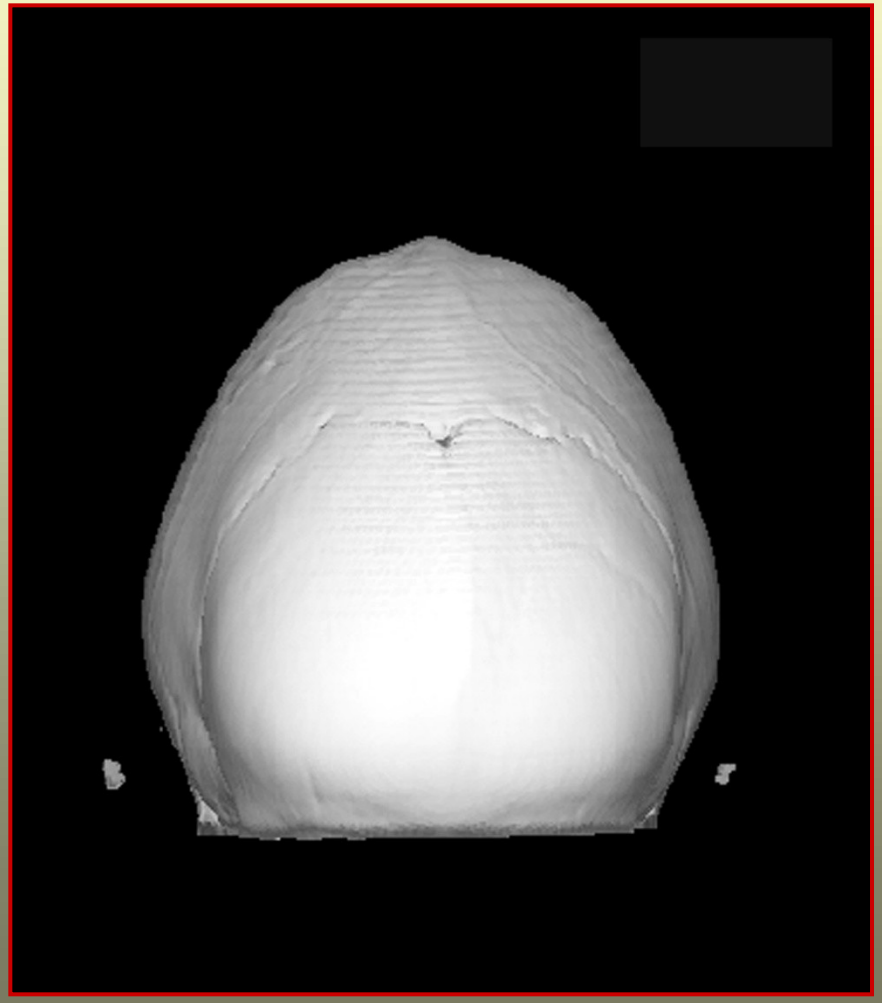
1. Skaphocephalia - dolichocephalia

Premature sagital suture synostosis

40-60%

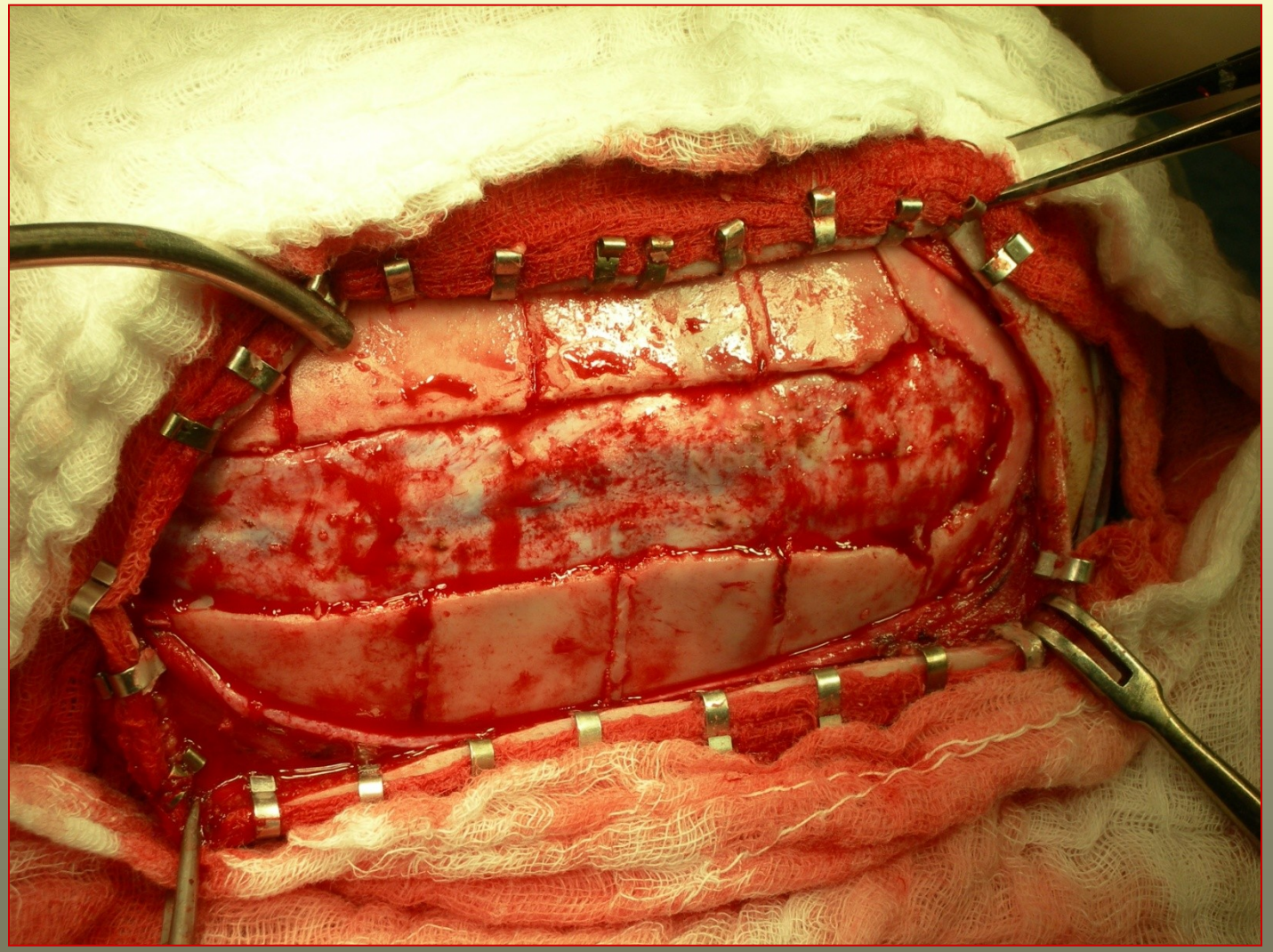


Skaphocephalia - dolichocephalia



Skaphocephalia - dolichocephalia

Skaphocephalia - dolichocephalia

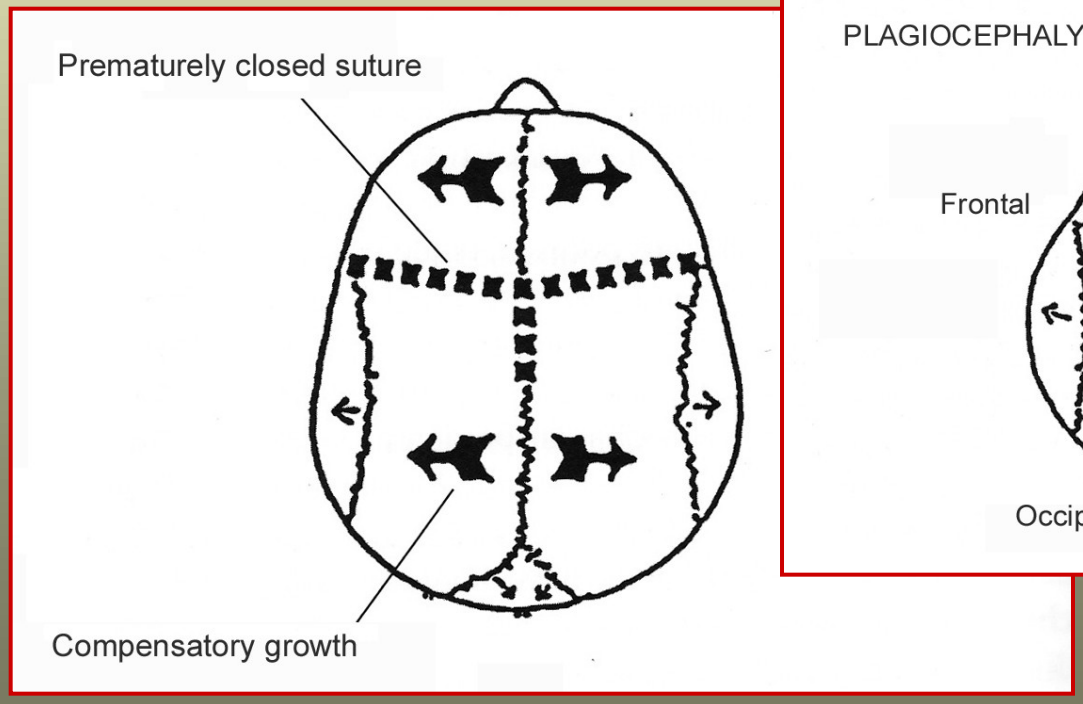


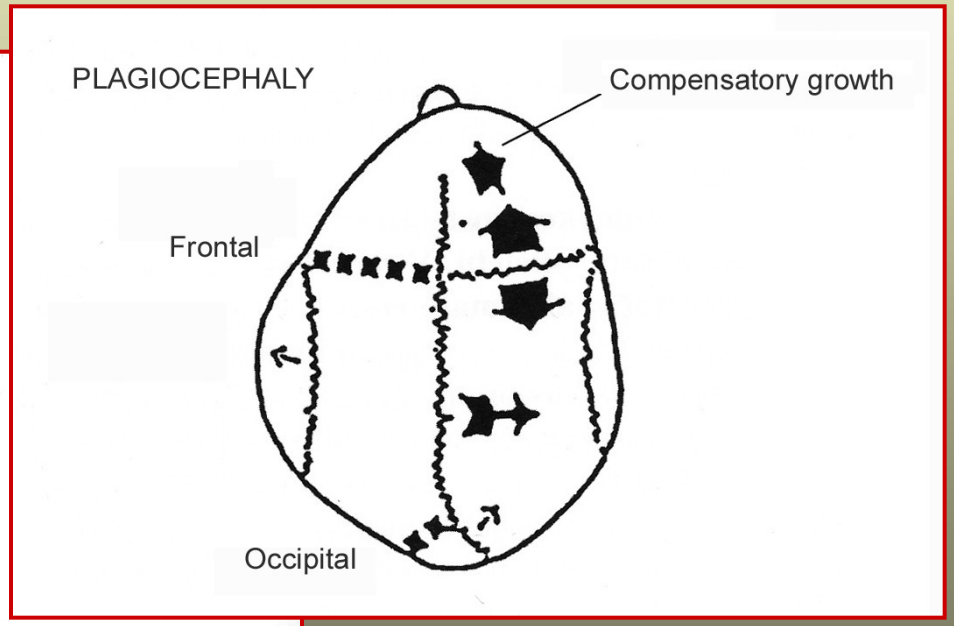
2. Brachycephalia

Premature coronar suture synostosis

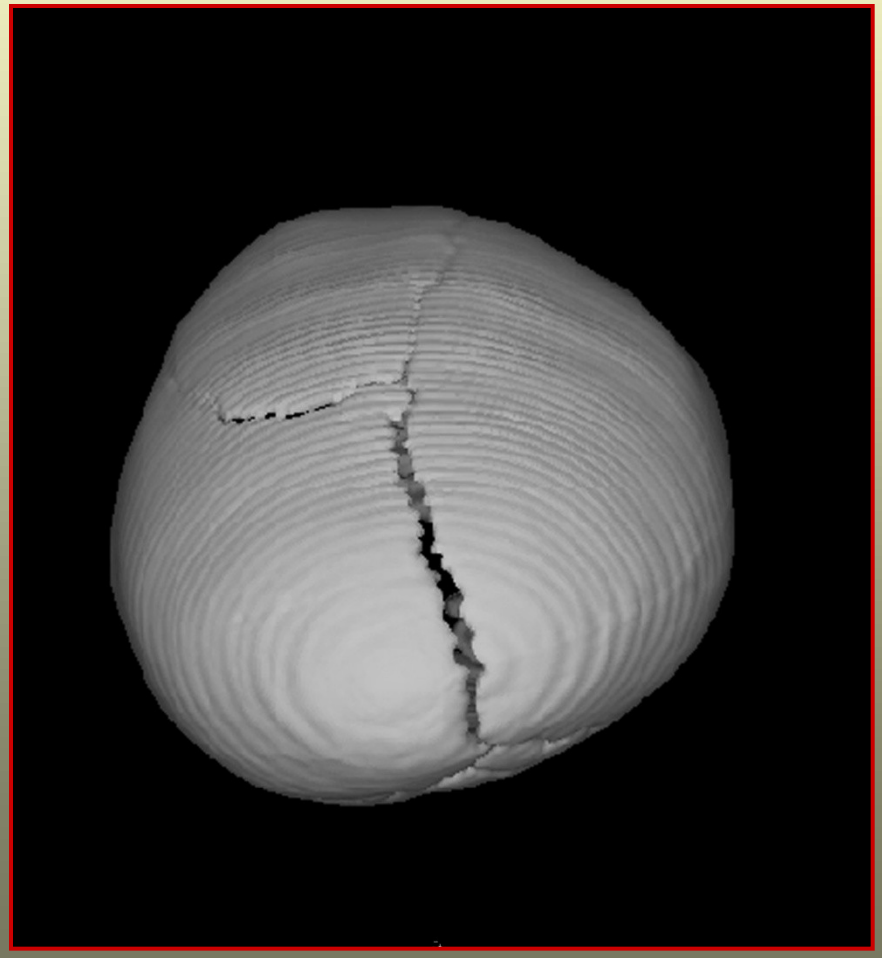
20-30%

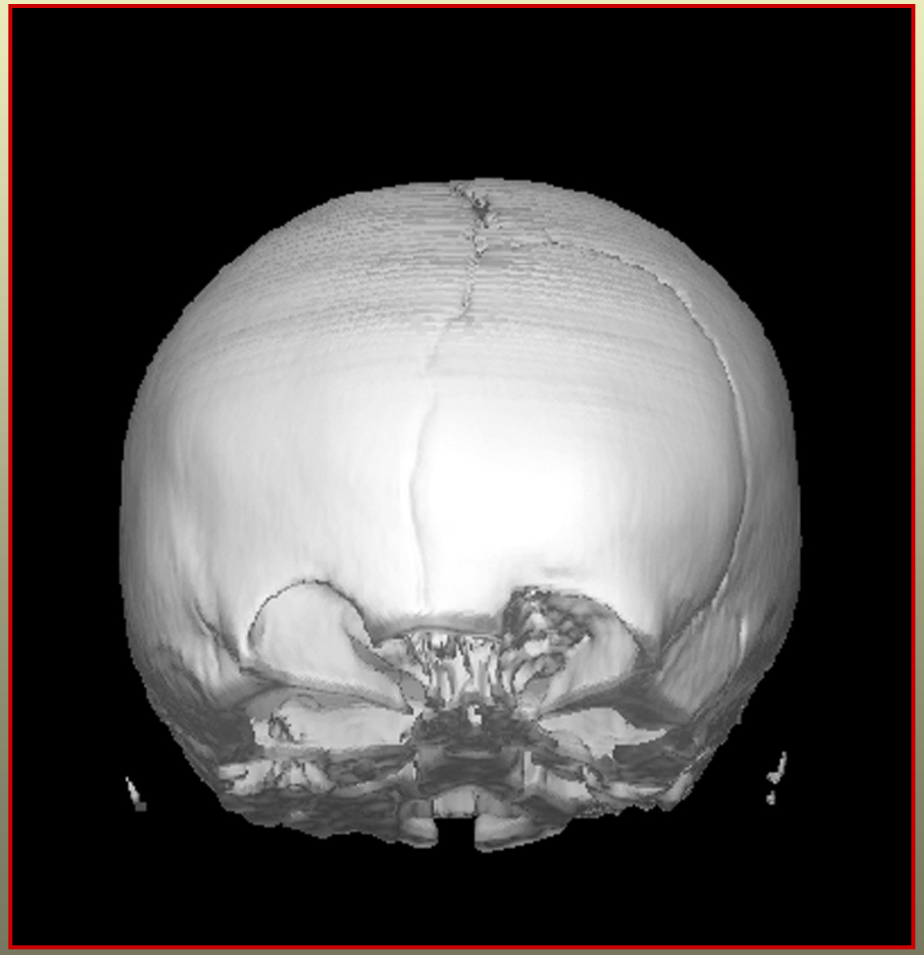
a) **Frontal plagiocephalia** – one side coronar suture
b) **Occipital plagiocephalia** – one side lambdoid suture



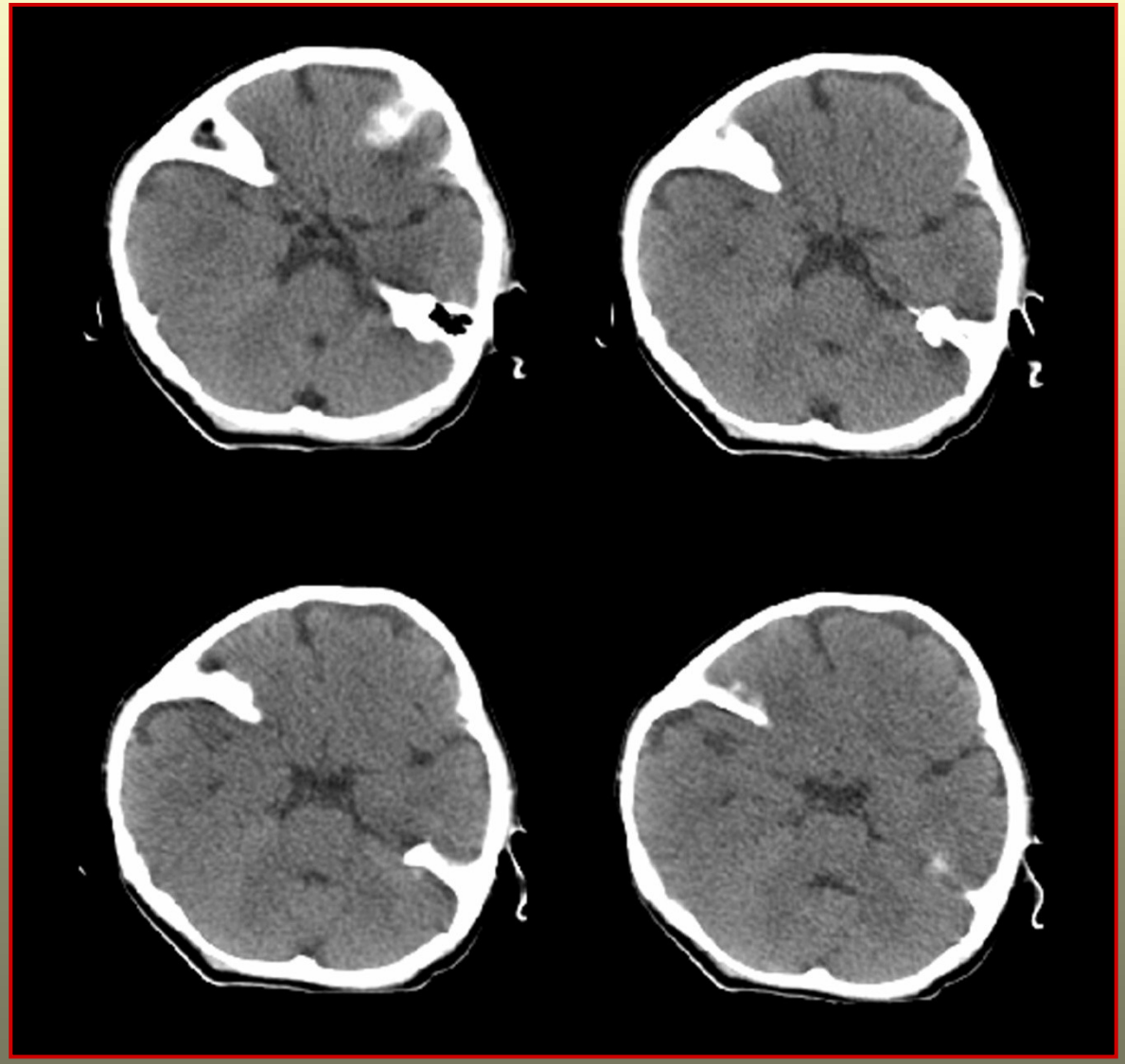


Brachycephalia





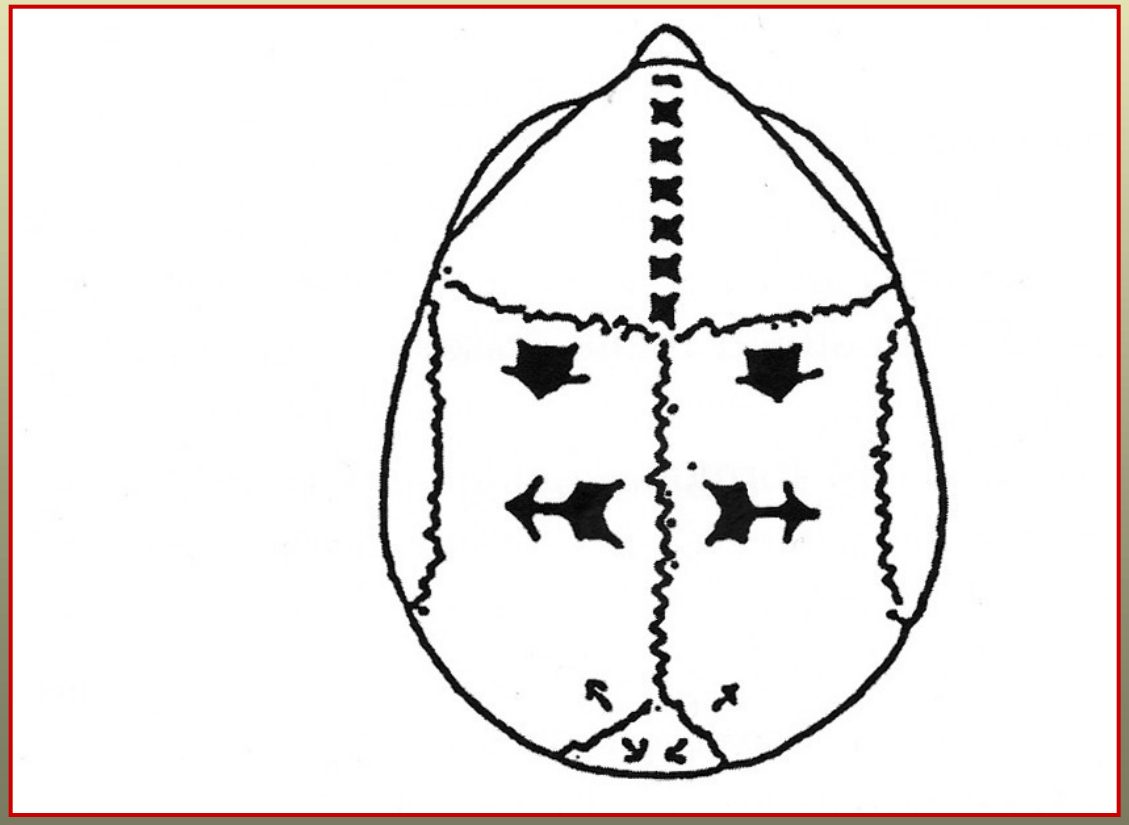
Brachycephalia



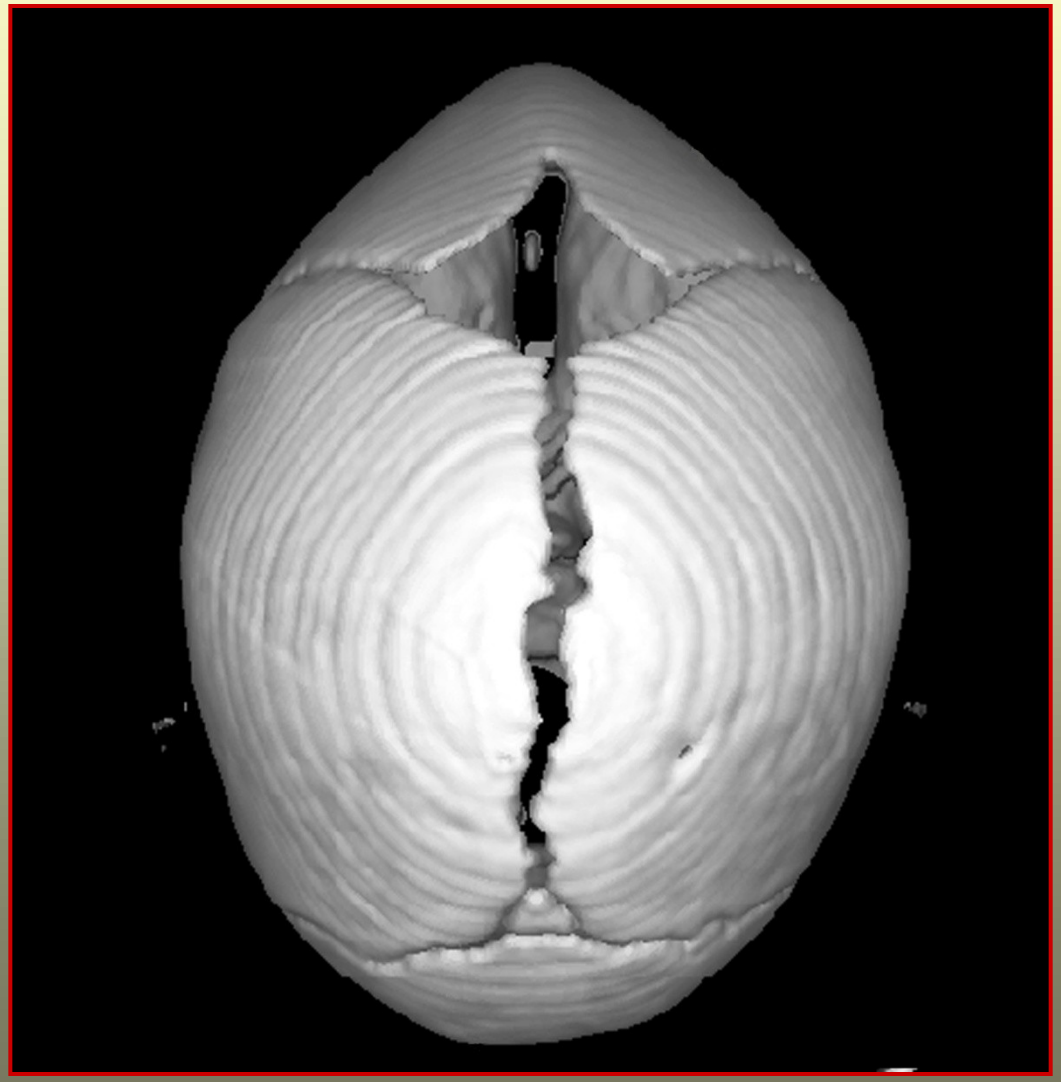
3. Trigonocephalia

Premature sutura metopica synostosis

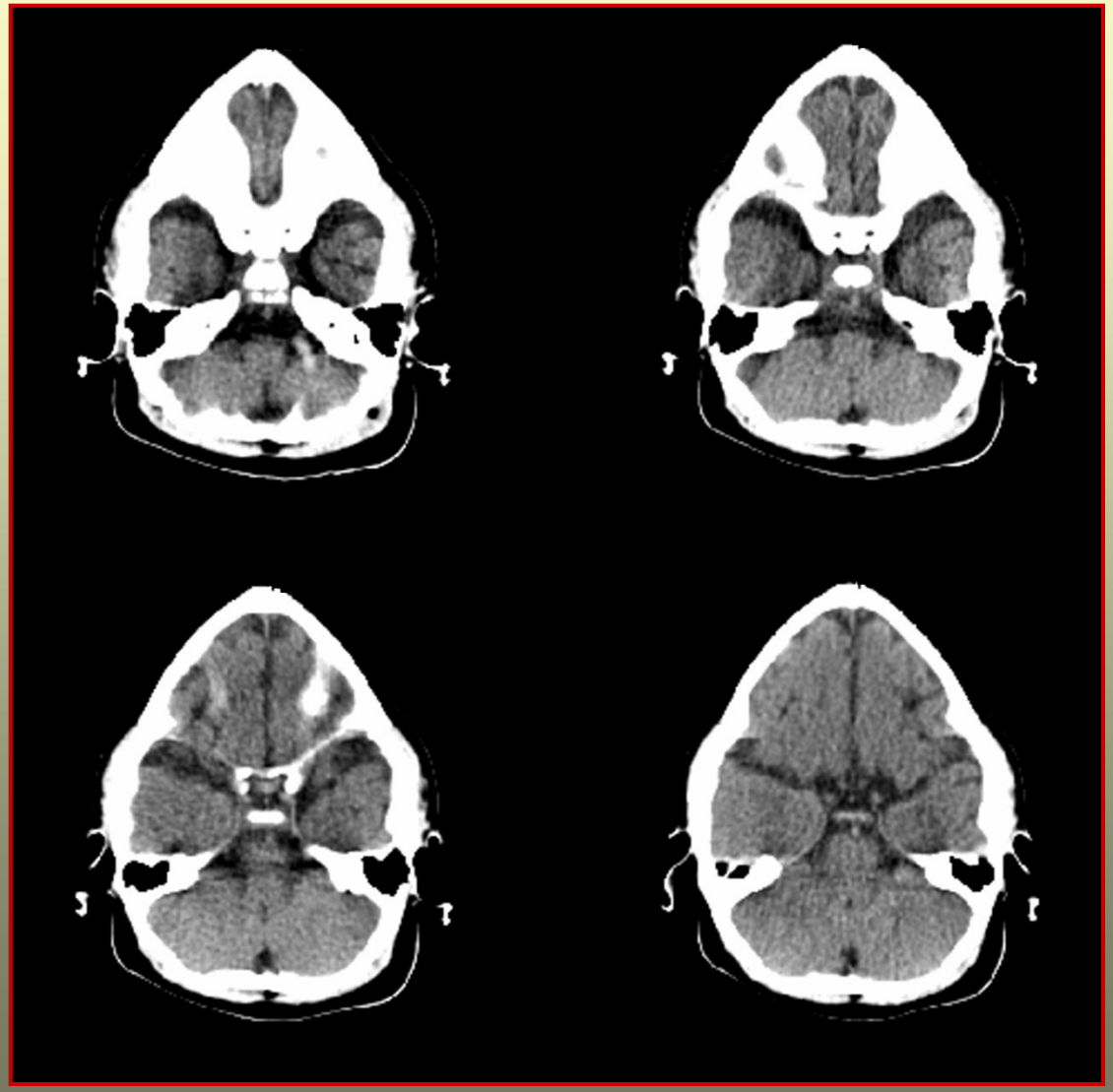
10%



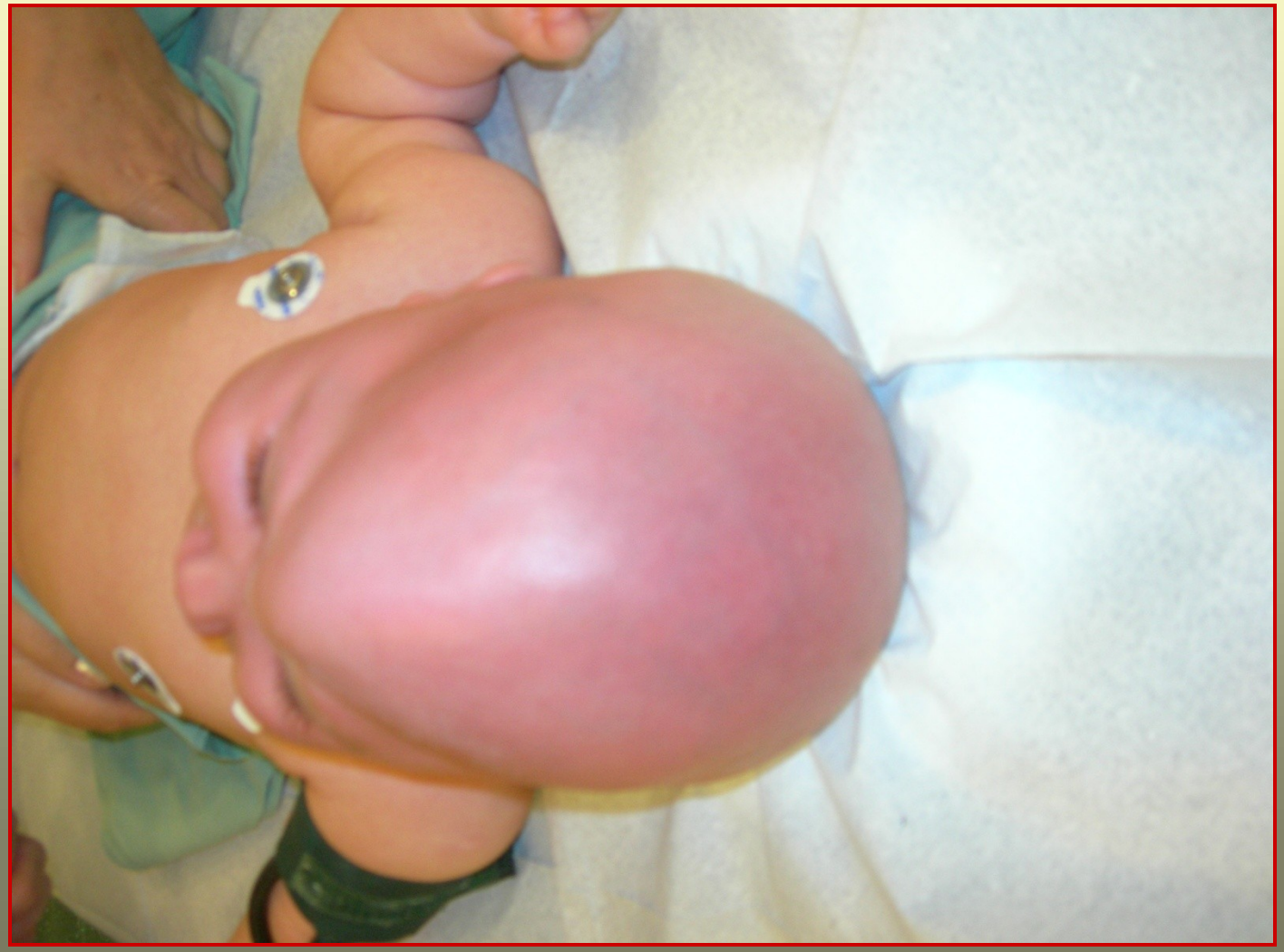
Trigonocephalia



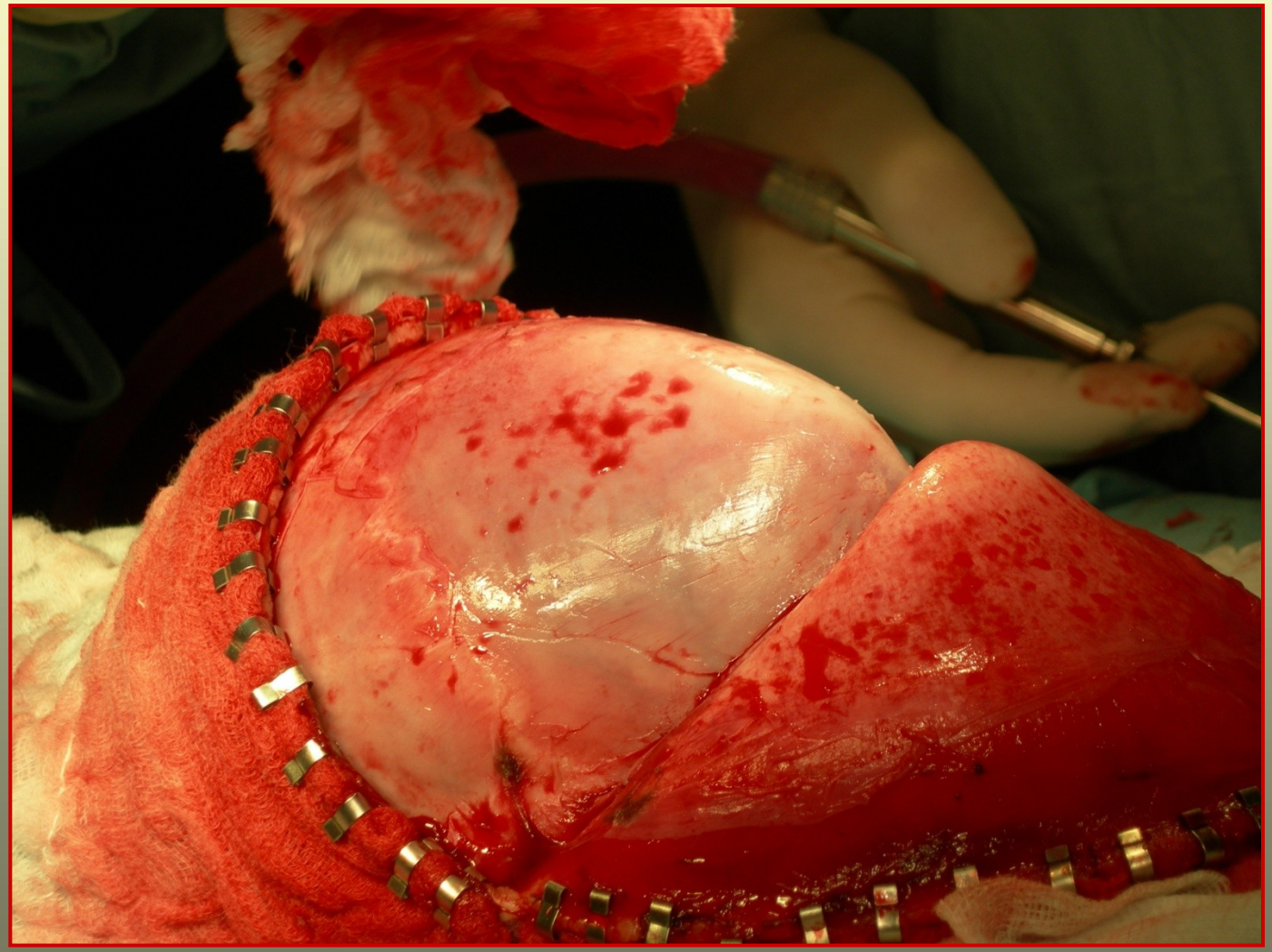
Trigonocephalia



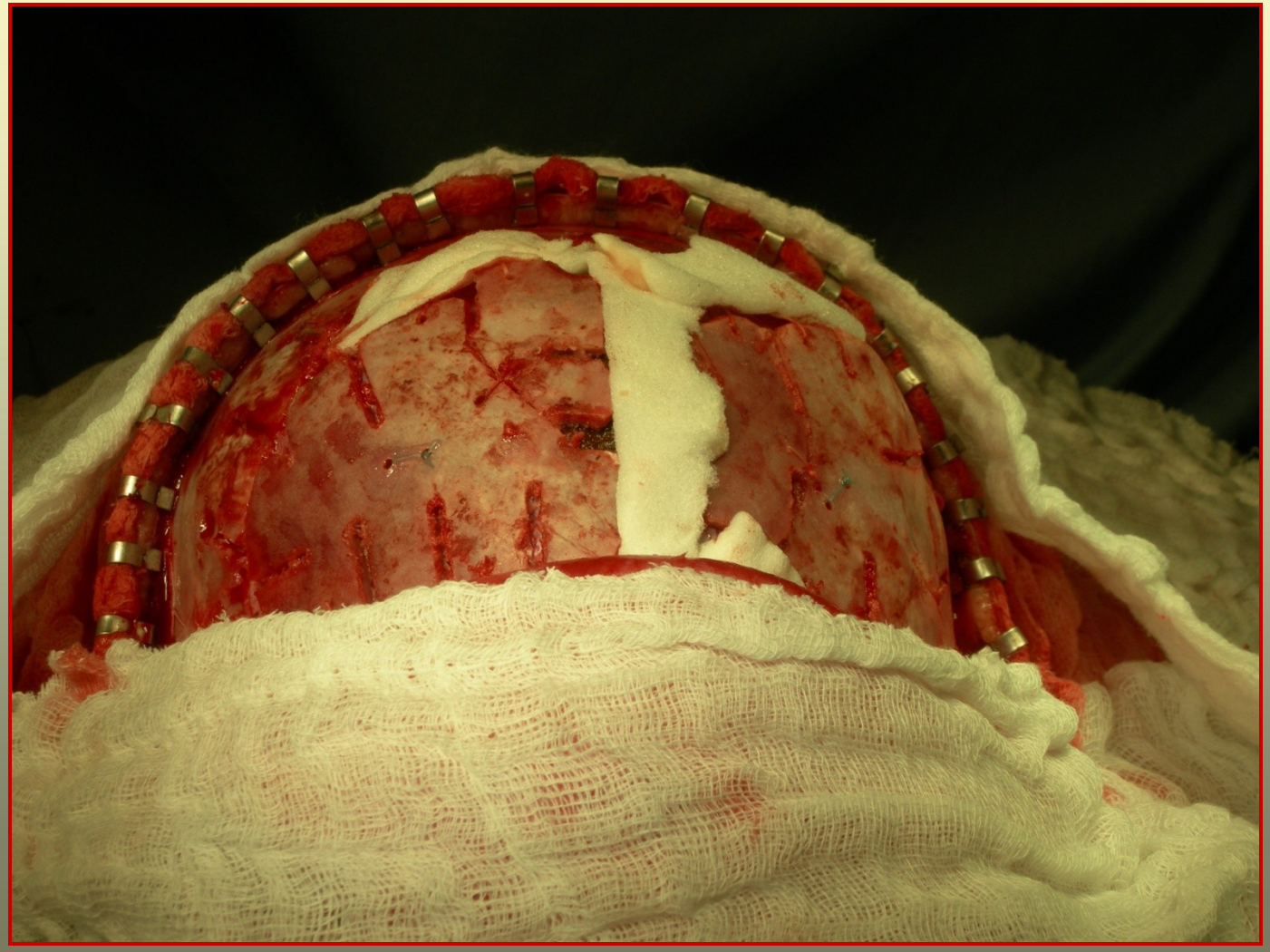
Trigonocephalia



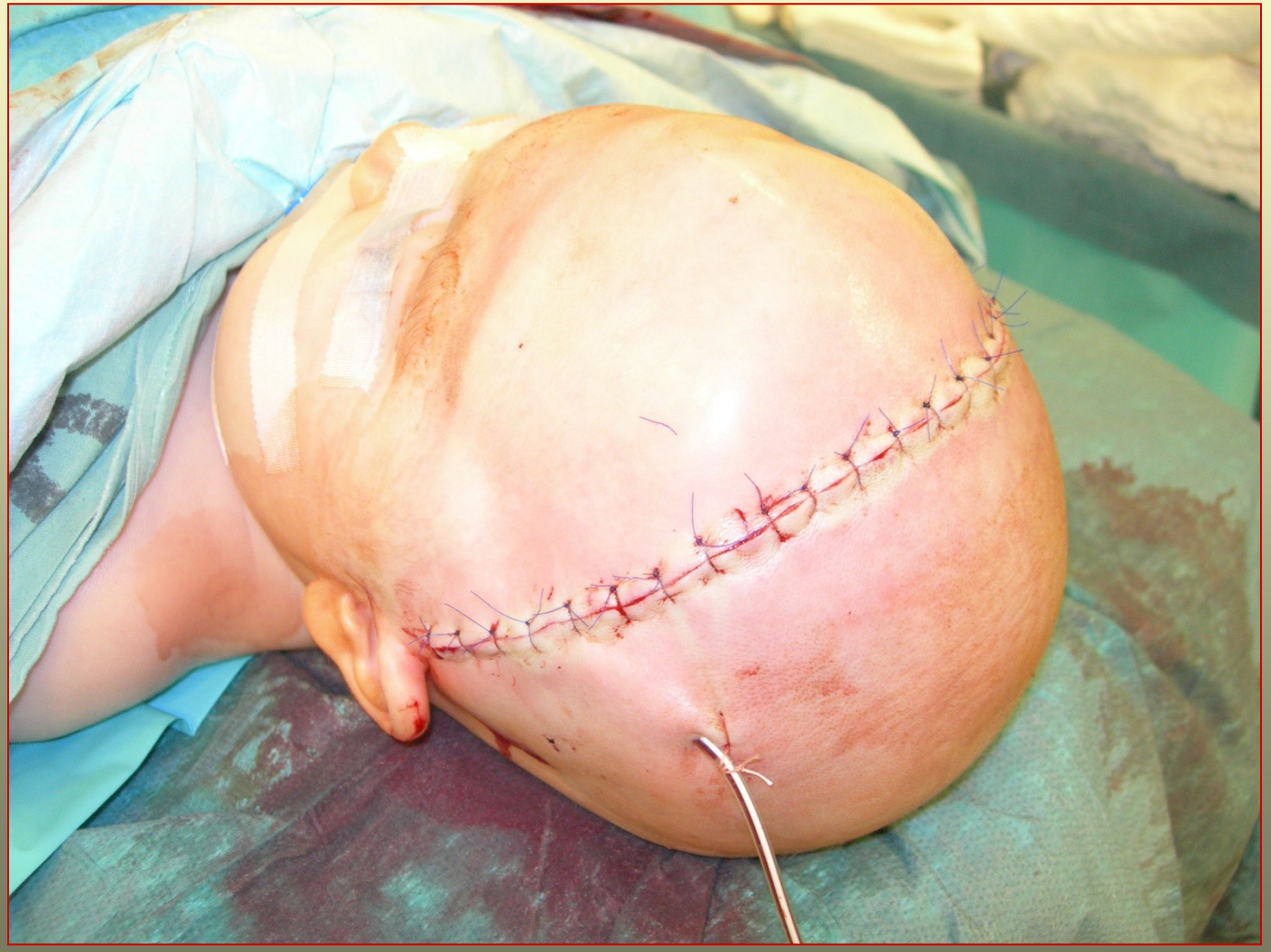
Trigonocephalia



Trigonocephalia



Trigonocephalia



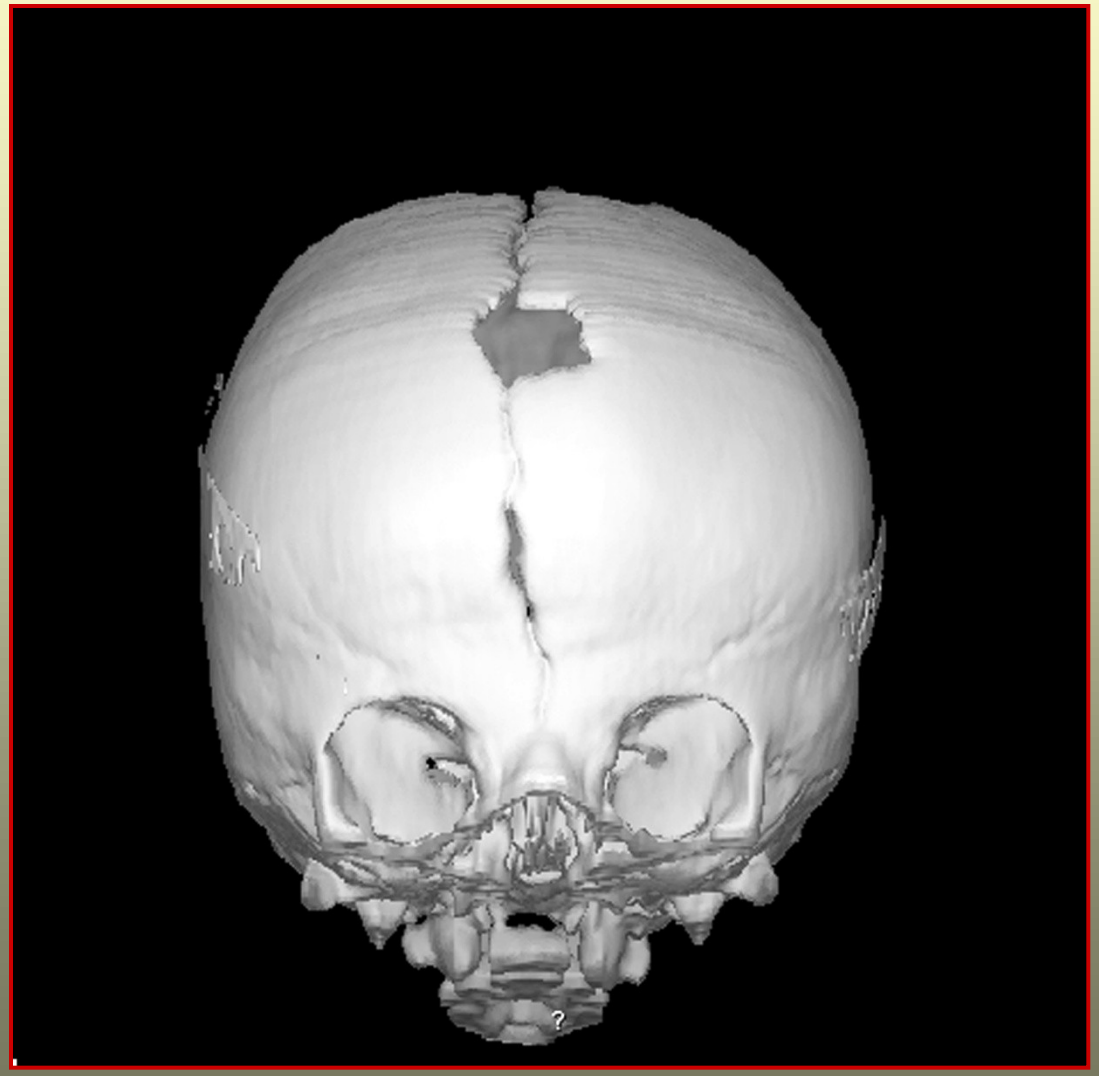
4. Morbus Crouzon – dysostosis craniofacialis

(1912)

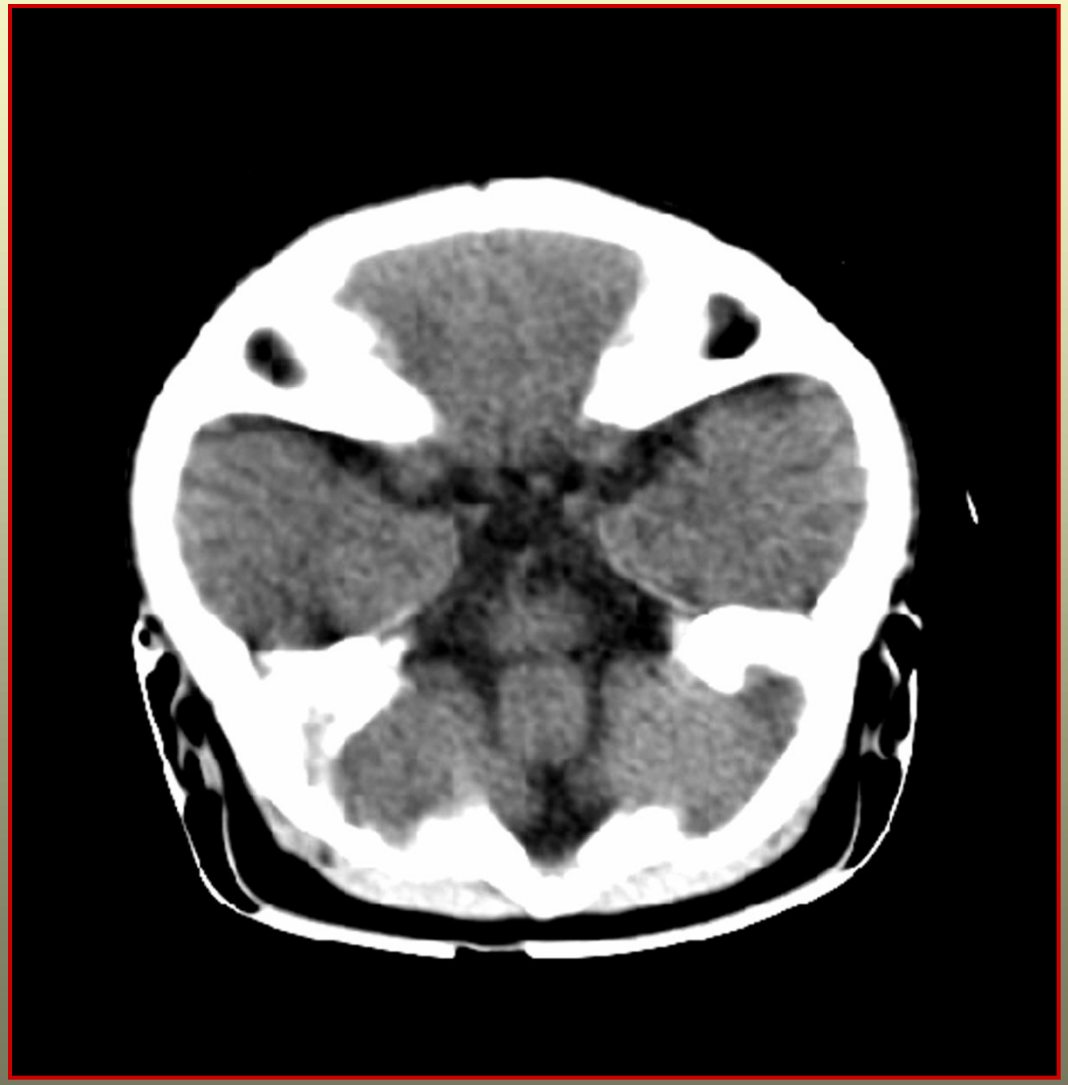
- turicephaly
- shallow orbits
- exophalmos
- hypertelorism
- hypoplasia of middle facial skelet

1 from 25 000 children

Morbus Crouzon



Morbus Crouzon



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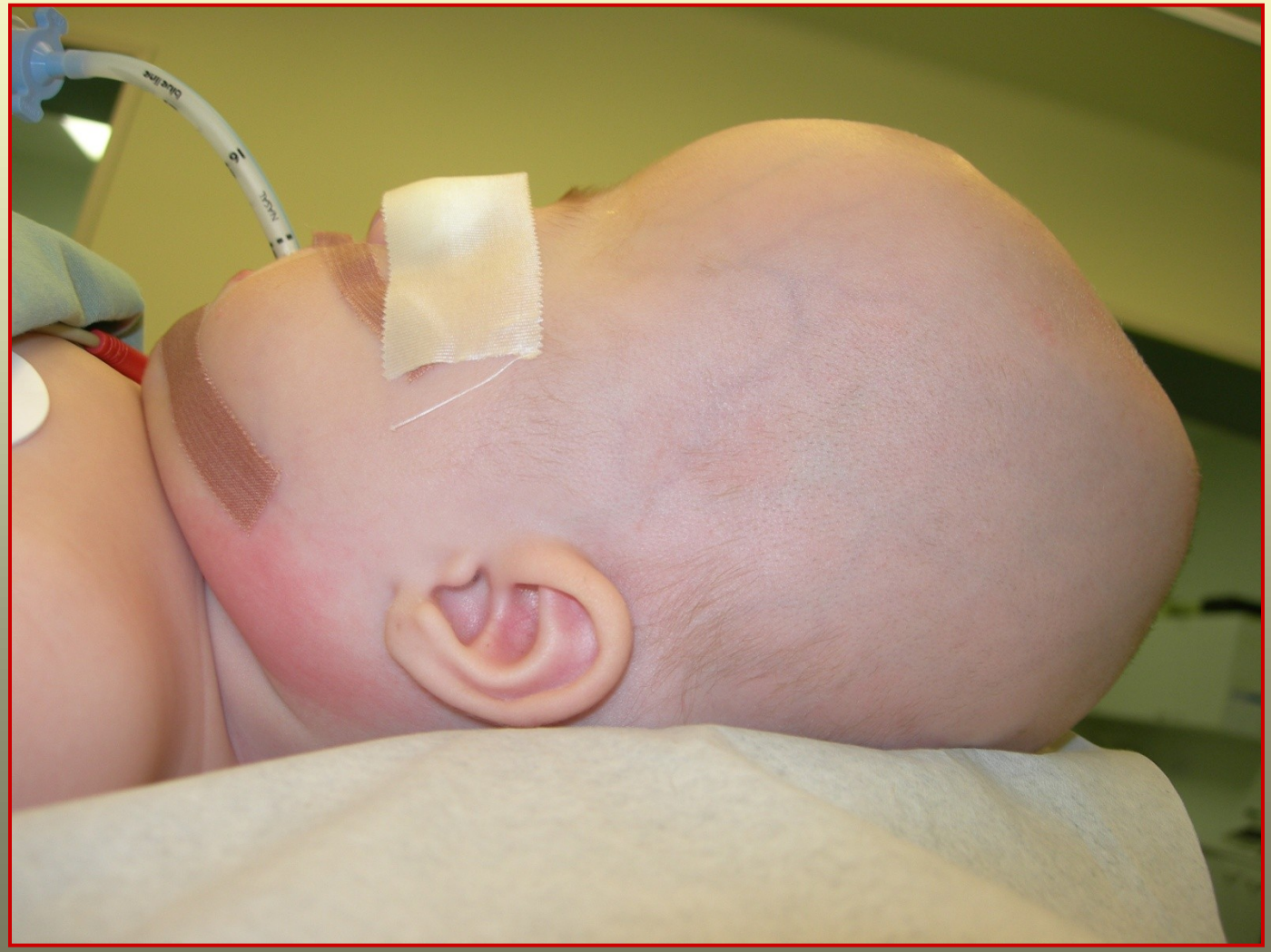

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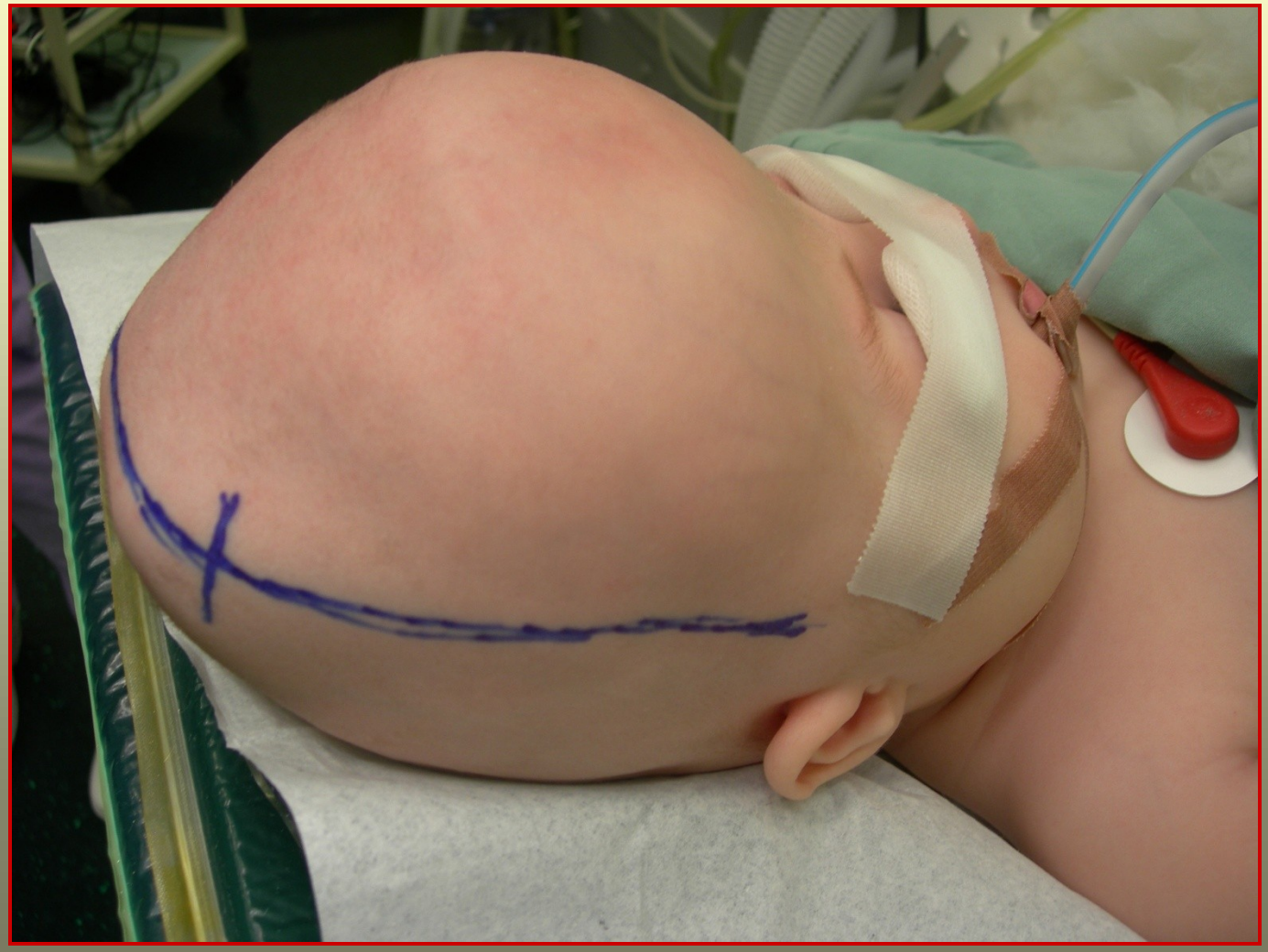

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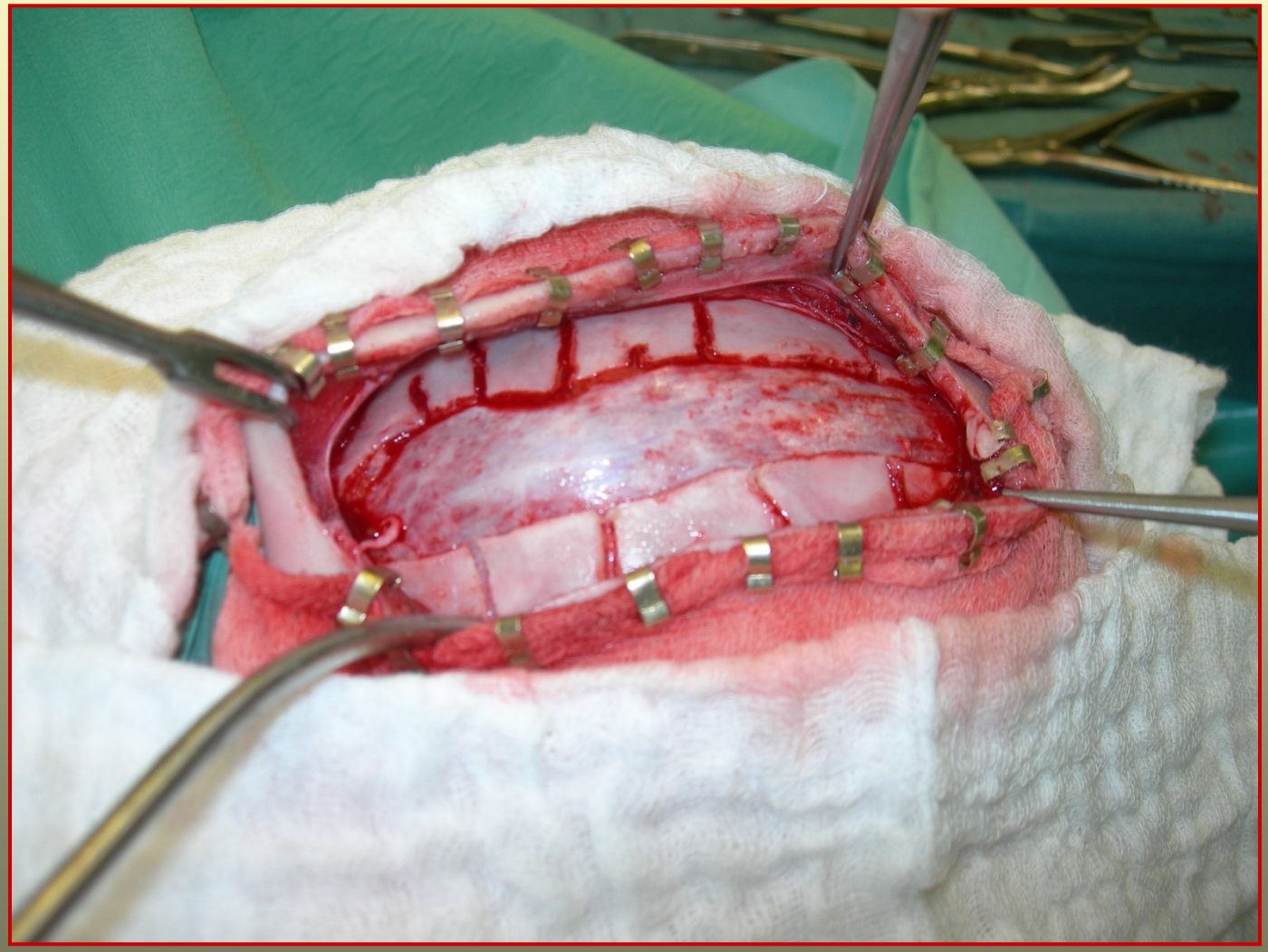
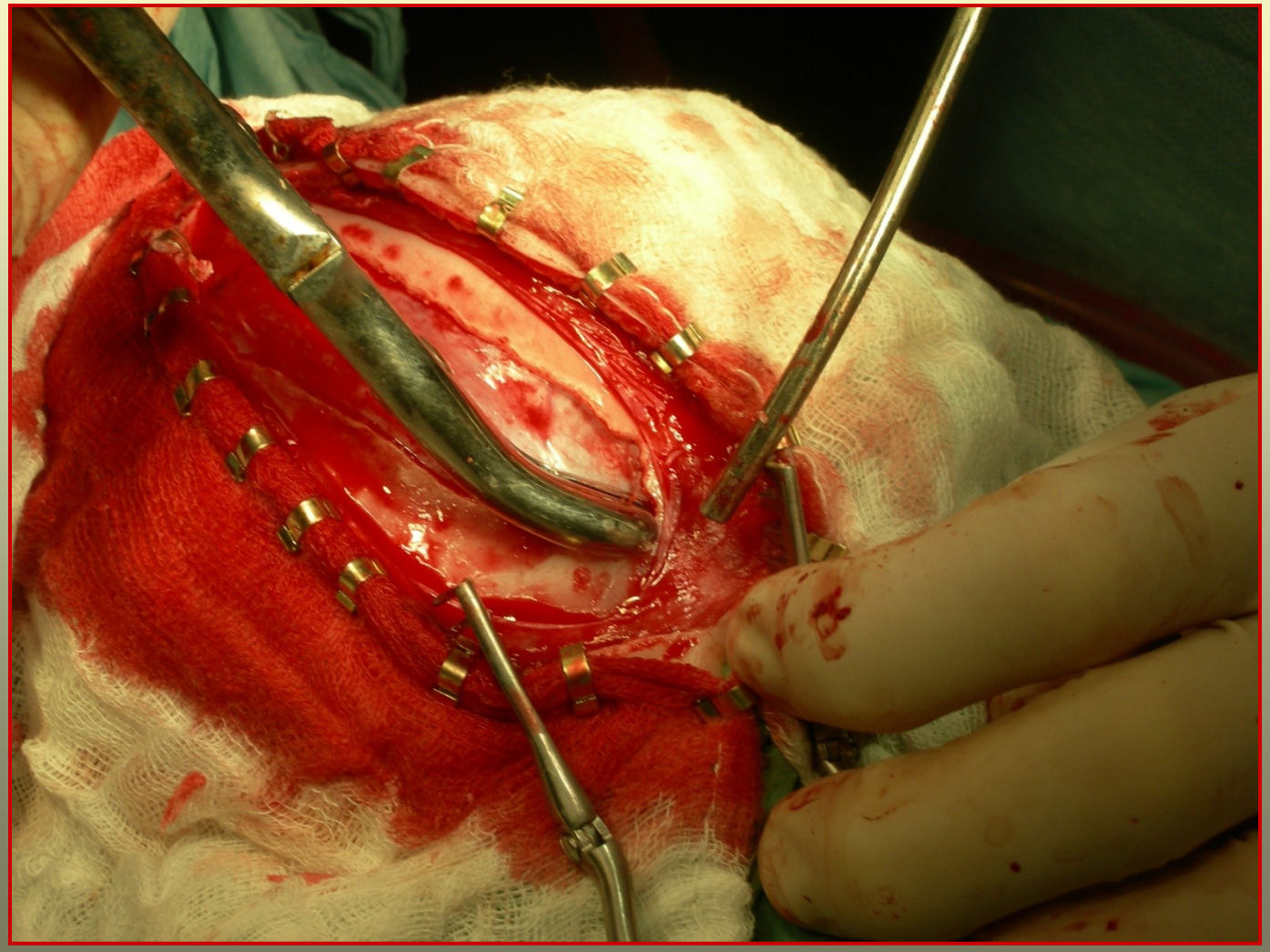


Morbus Crouzon



Post surgery care – preventive protective helmets