Tips and Tricks in Craniosynostosis and Skull Deformity Management

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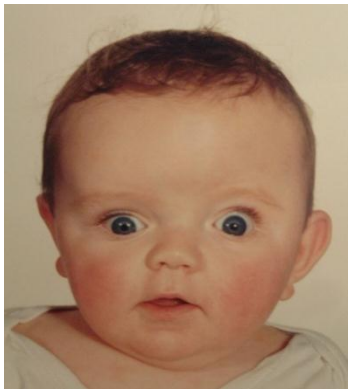
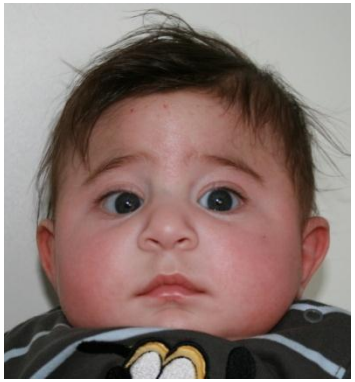
Non syndromic – isolated – single suture

Etiology

- epigenetic /environmental (pressure modulated)
  obstetric related, idiopathic
    - multiple pregnancy
    - oligohydramnion
    - malposition



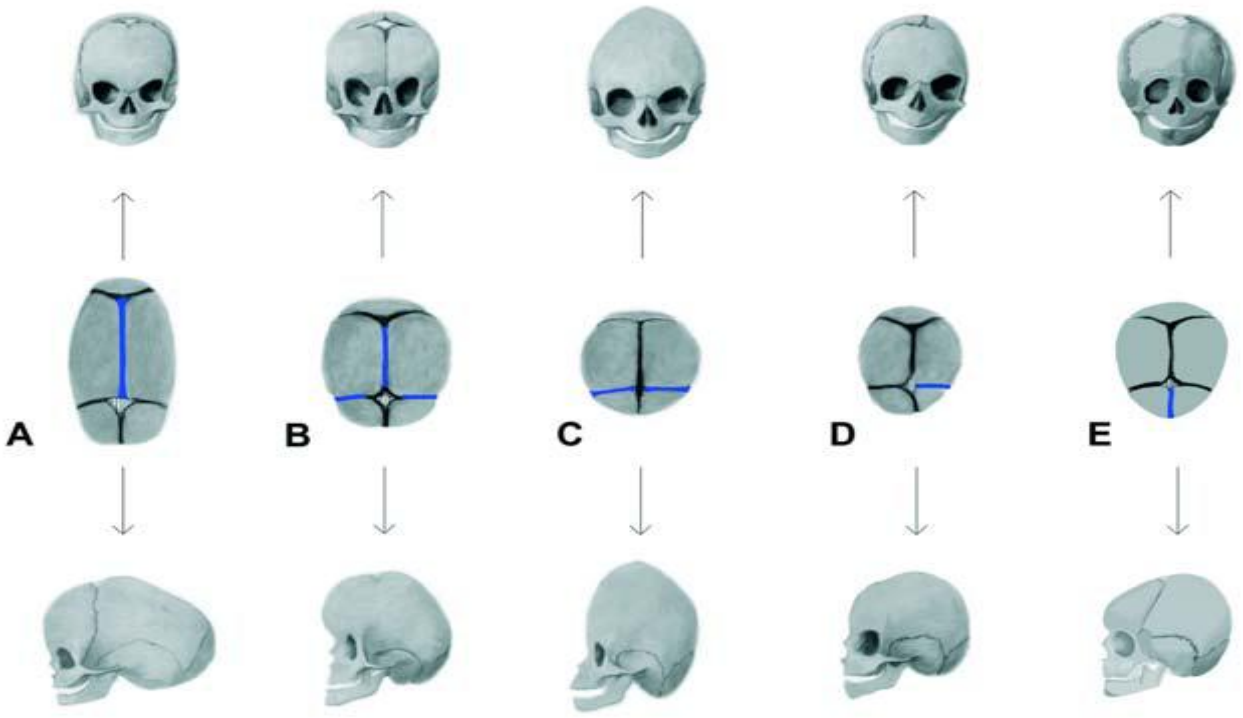
- Genetic (syndromic)
  - chromosomal abnormalities
  - point mutations and microdeletions
    - FGFR (II) and TWIST genes
  - undiscovered



Incidence

overall = 1:2.000-3.000

- syndromic 20-25%
- non-syndromic 75-80%
  - metopic 14% **E** ↑
  - sagittal 40% **A**
  - coronal 20%
    - UCS 12% **D**, BCS 8% **C**
  - lambdoids 1%



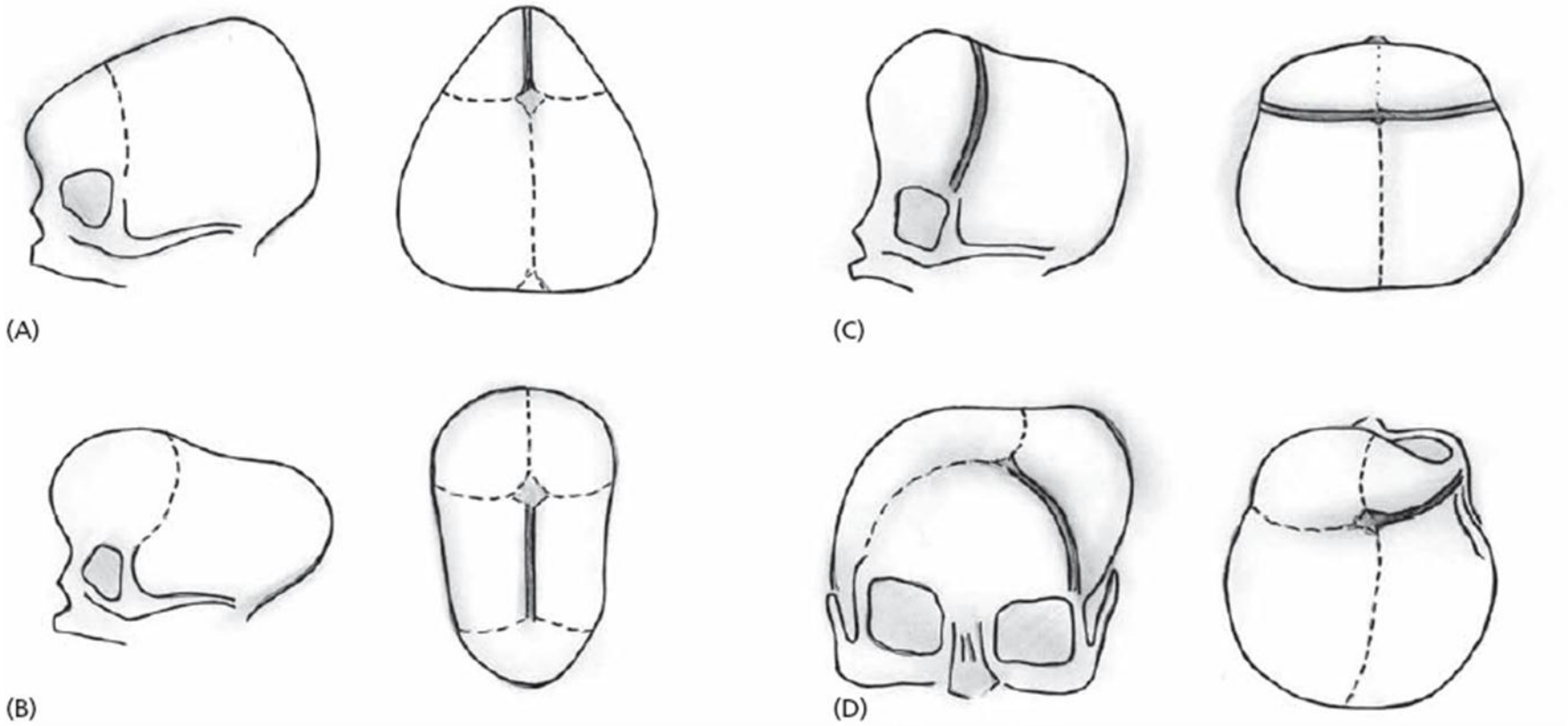


Figure32.2 Single-suture craniosynostosis results in specific cranial deformities. (A) Trigonocephaly is the result of premature closure of the metopic suture and typical features are a frontal keel (↑), anterior displacement of the coronal sutures (↑↑), and compensatory bulging of the parietal squamae (↑↑↑). (B) Scaphocephaly is characterized by bitemporal or biparietal narrowing (↑), frontal (↑↑) and occipital (↑↑↑) compensatory bossing. The shape of the skull resembles the keel of a boat or a saddle. (C) Brachycephaly means short skull and is the result of bicoronal craniosynostosis. The frontal region is flattened (↑) and some cases show early compensatory oxycephalic deformity (↑↑). (D) Anterior plagiocephaly results from unilateral coronal synostosis and causes asymmetrical and rotational deformity of the fronto-orbital region with additional midface and skull deformities.

Anesthesia for Plastic and Craniofacial Surgery, Ehrenfried Schindler, Markus Martini & Martina Messing-Jünger, Gregory's Pediatric Anesthesia, 5. Edition 2012, John Wiley & Sons

Non syndromic – isolated – single suture

Actual questions **?**

Are non-syndromic craniosynostoses really non-syndromic?

→ genetics?

Why does the trigonocephaly incidence increase?

→ environmental / epigenetic factors?

Management

- *Radiation free* diagnostics

- Surgery
  standard
  minimally invasive (sagittal, UCS)
  + additional helmet therapy (?)

- *Radiation free* follow up (8-10 yrs)
  foto scan
  clinical
  ophthalmological

Tips for presurgical management

Diagnostics

in most cases clinical evaluation is sufficient

in difficult / complex cases and for objective follow up

→ non radiating techniques
- photo / video / laser scans (3D)
- transfontanellar sonography
- MRI (hydrocephalus, Chiari and other brain malformations)

Surgical indication

standard → all deformities
(according to deformity: cranioplasty w/wo fronto-orbital advancement)

minimally invasive w/wo endocopic assistance
(sagittal, mild unilateral coronal or metopic, early ICP↑ in syndromic cases)

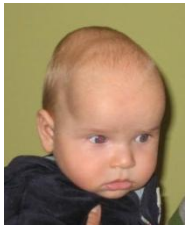
Advantage of early surgery
→ skull base malleable, highest cerebral pulsation forces

diagnostics and follow up in cranial deformities

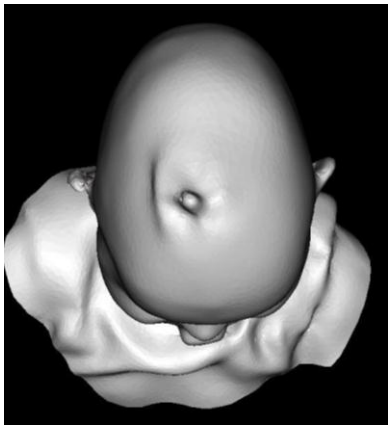
concept of “radiation-free diagnostics” in craniosynostosis

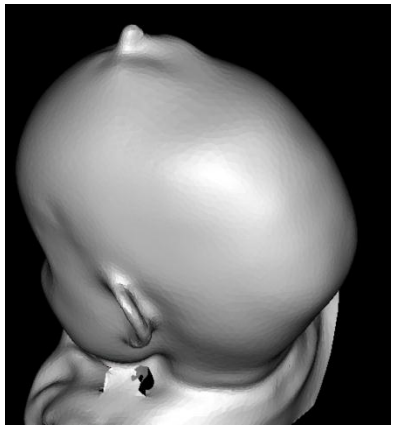
cranial suture sonography

3D photo-laserscan pre / postoperative

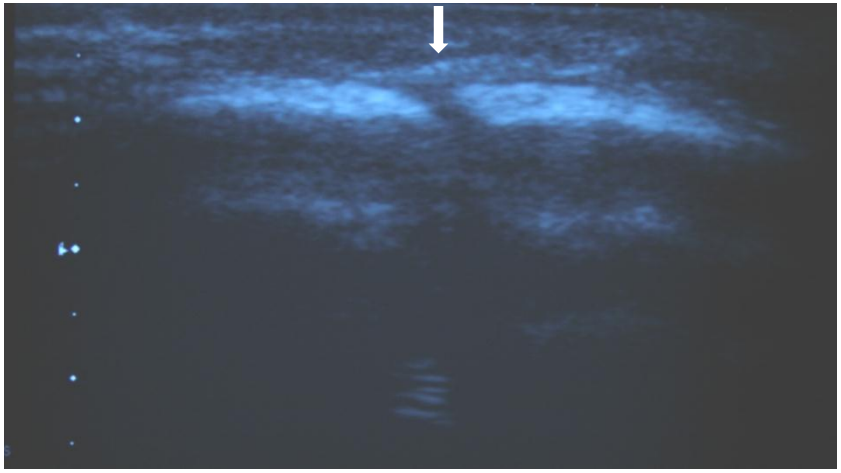


brain sonography / MRI in syndromic patients

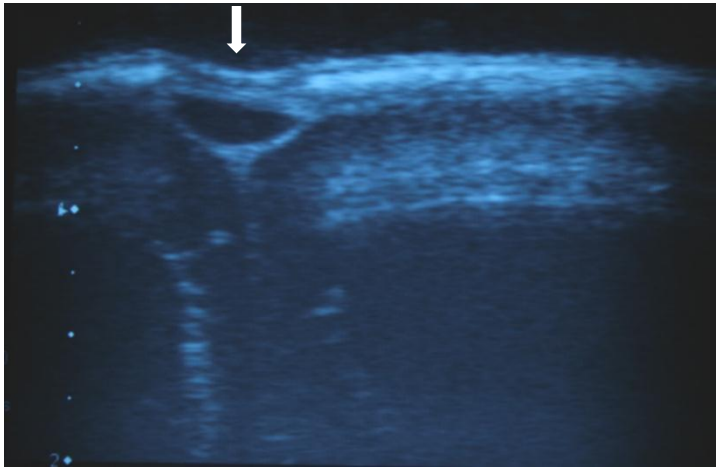




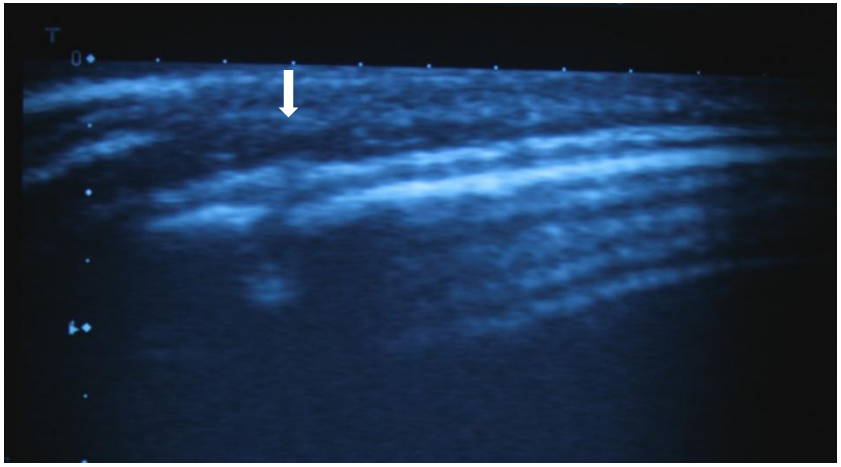
cranial suture sonography



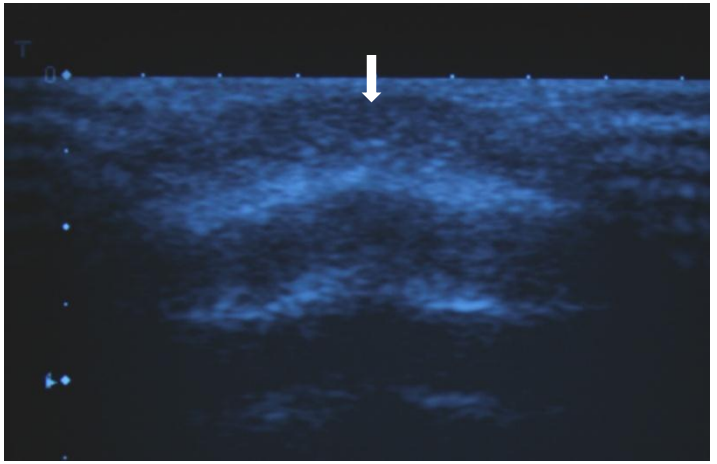
open lambdoid suture in positional plagiocephalus



open sagittal suture with sagittal sinus



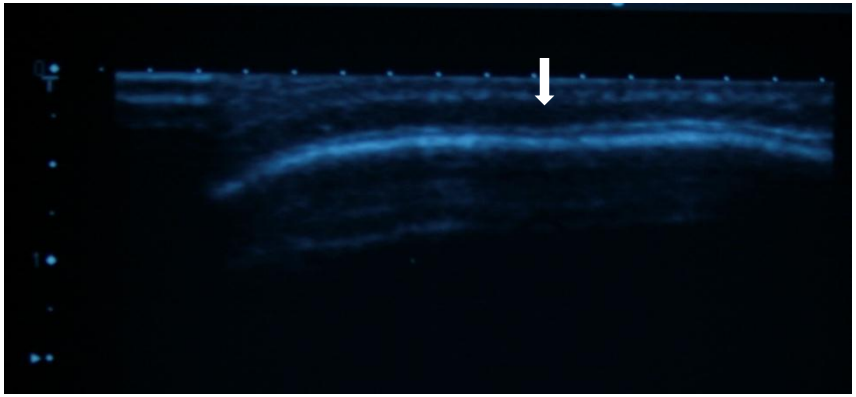
open coronal suture



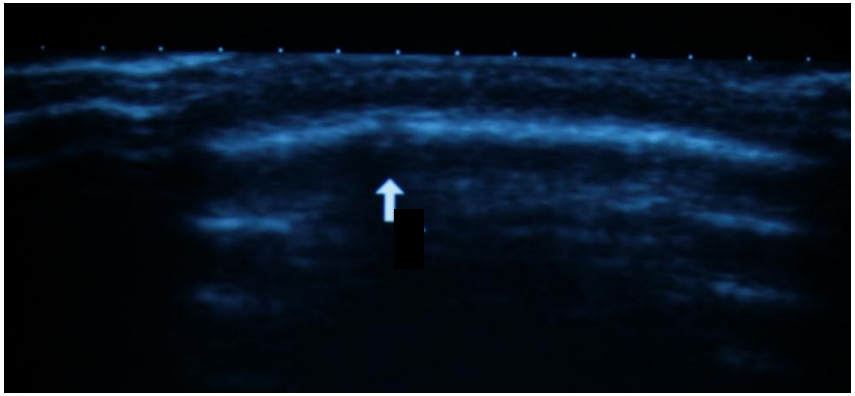
closed sagittal suture in premature sagittal synostosis

cranial suture sonography

patient with unilateral coronal synostosis



closed coronal suture



open contralateral coronal suture

3D photoscan of craniofacial shape

method

3D scan technique basing on combined photographic and video or laser scanning

provides metric information

- head circumference and height
- cephalic index
- cranial volume

direct comparison between follow up scans

no radiation

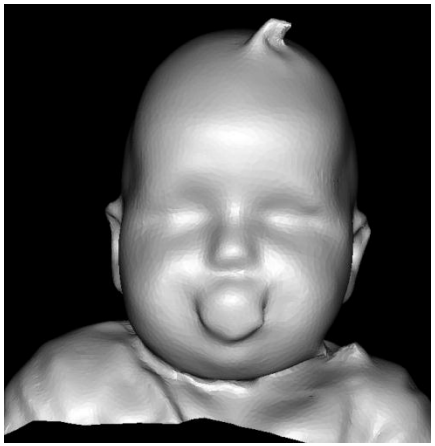
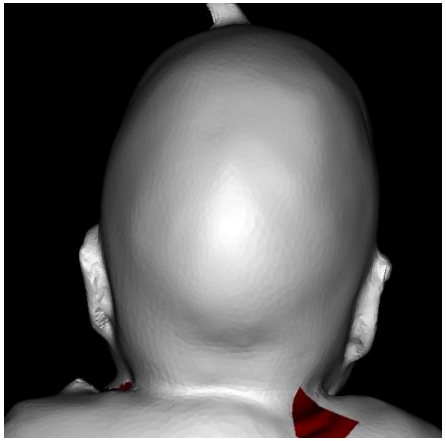
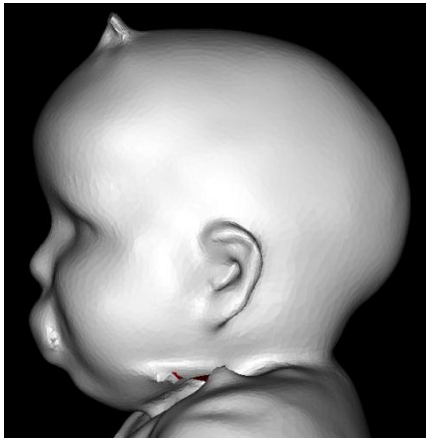
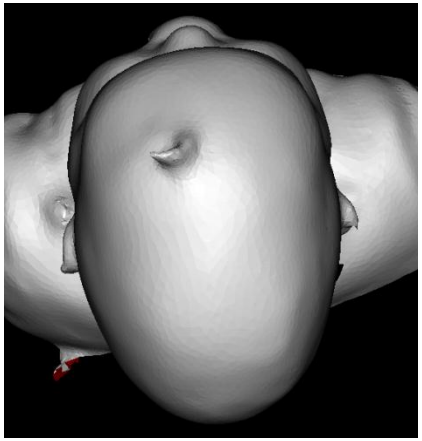
no anesthesia / sedation

indications in skull deformities

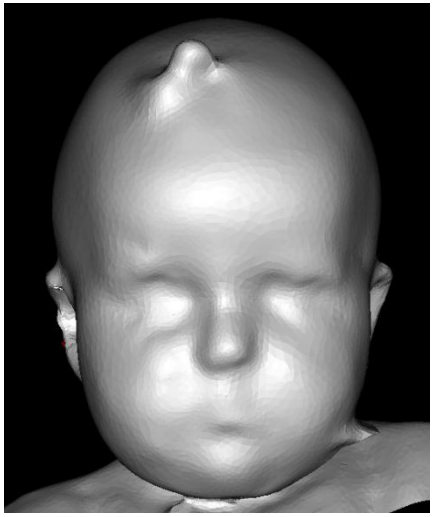
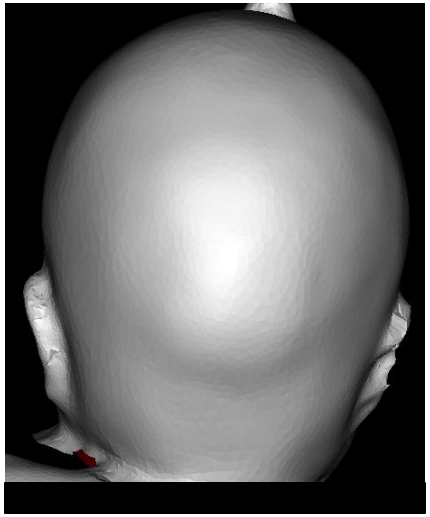
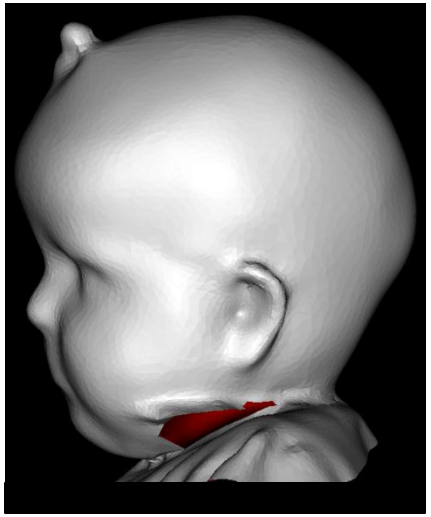
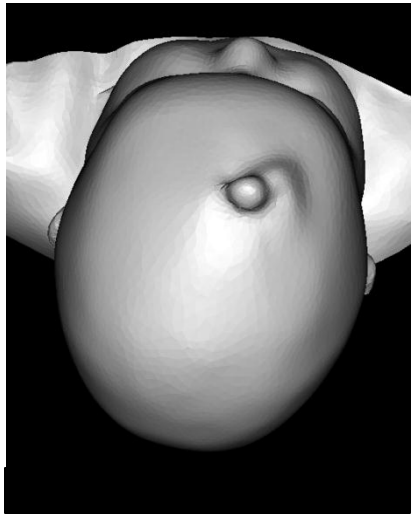
- synostotic
- non-synostotic
- after skull defects

Technical device for 3D photoscan of craniofacial shape

sagittal synostosis

preoperative

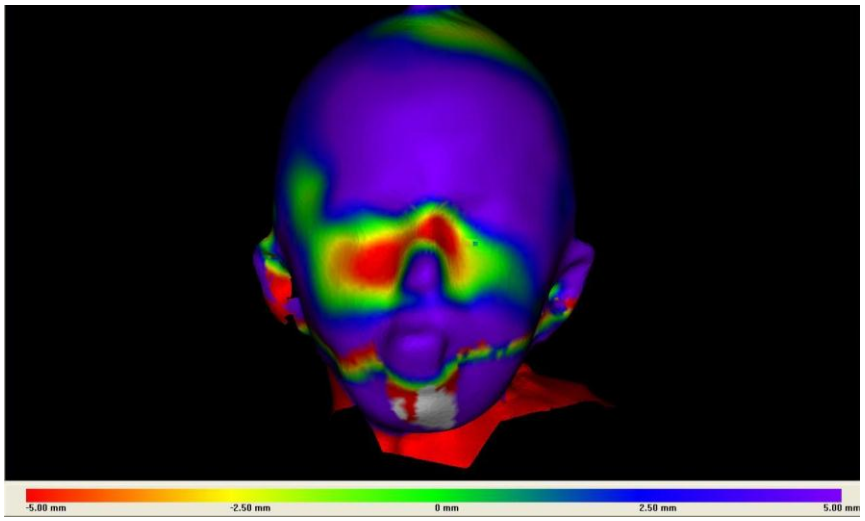





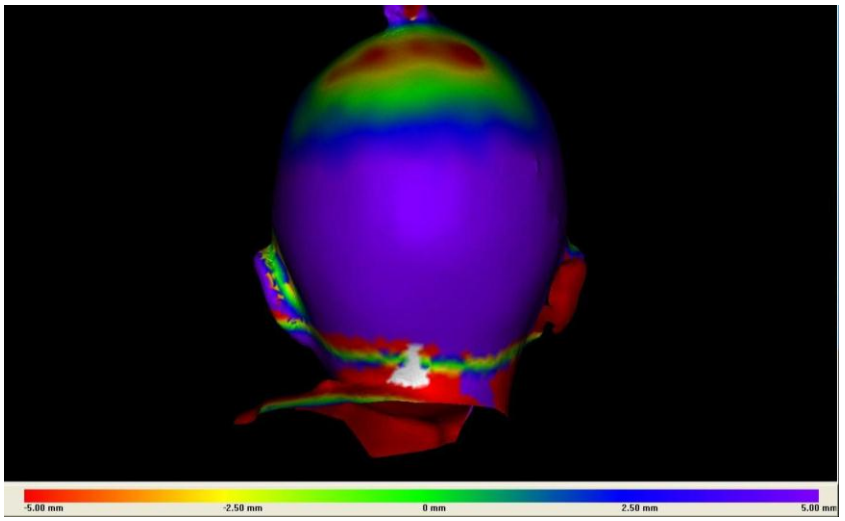
3 months postoperative

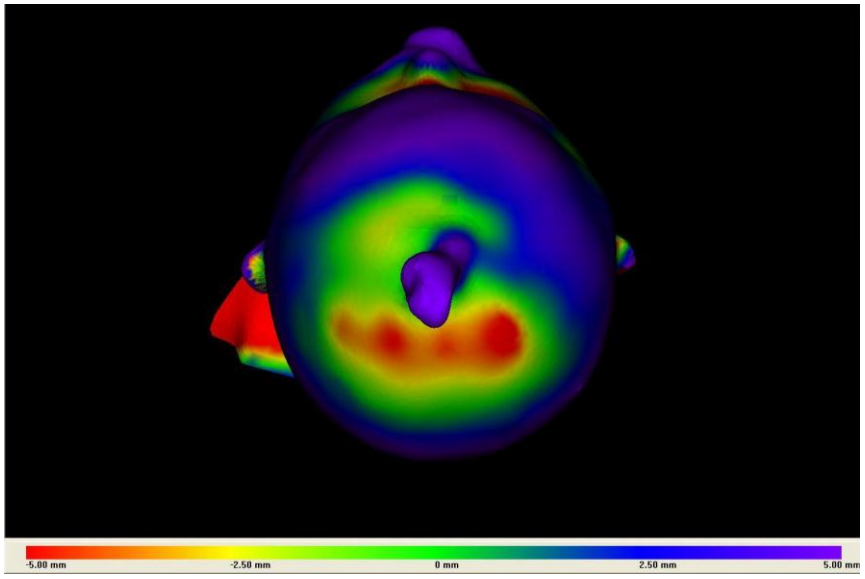
bicoronal synosis

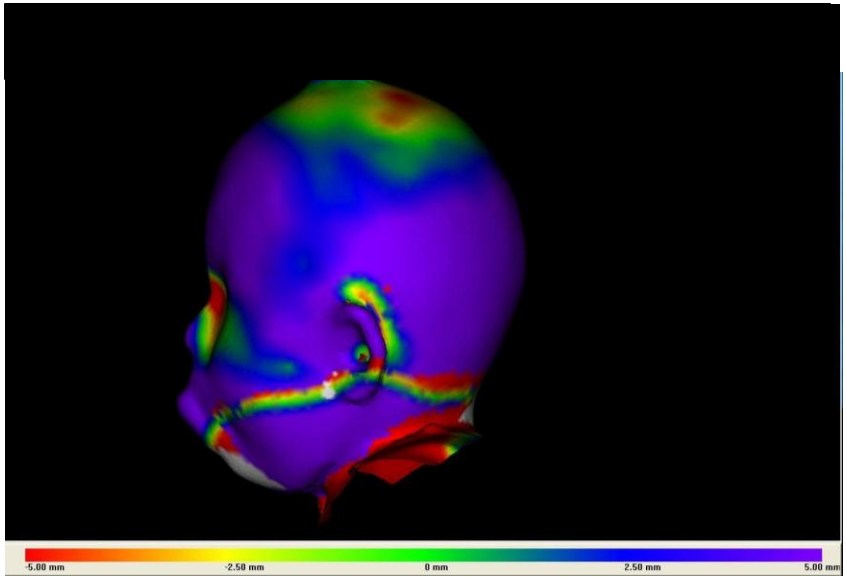
changes after 6 months

green: unchanged
violet/blue: increased





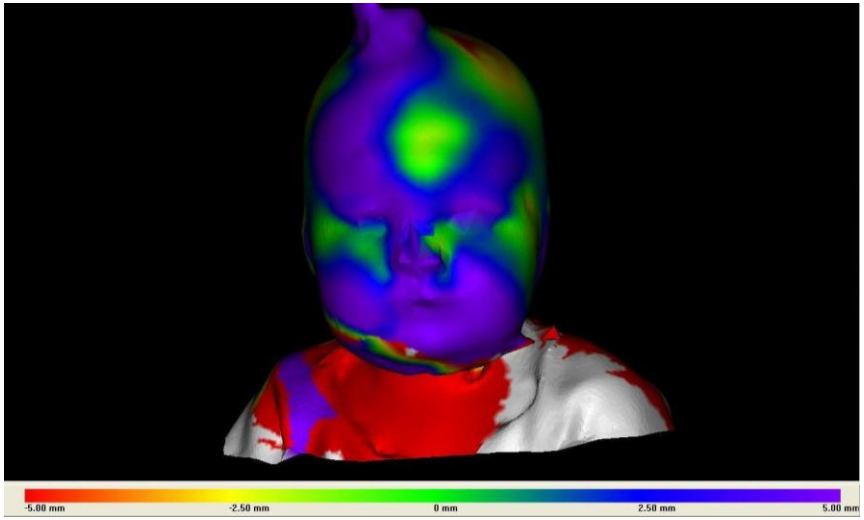


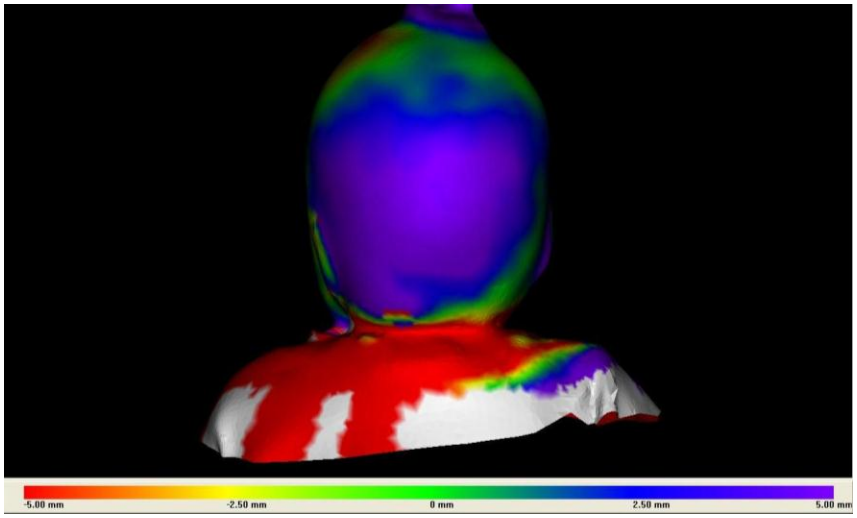


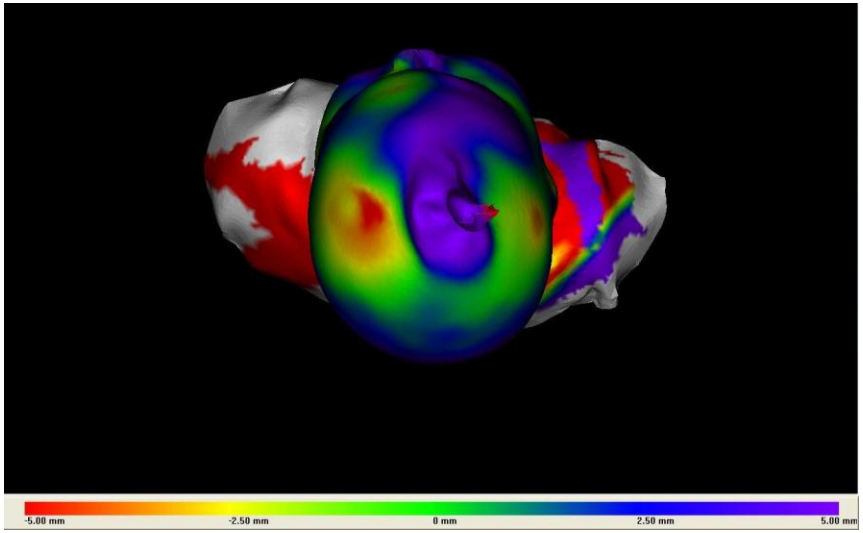
metopic synostosis

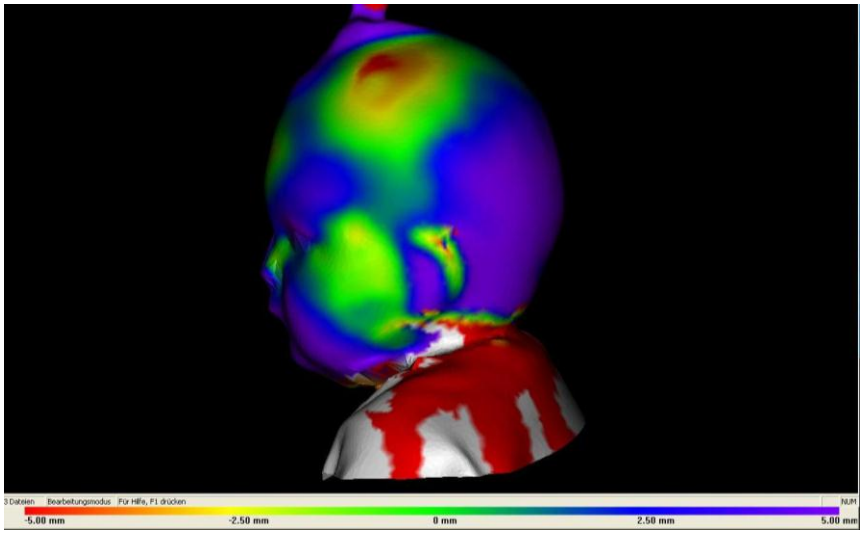
changes after 6 months

green: unchanged
violet/blue: increased









Operative positioning in craniosynostosis surgery

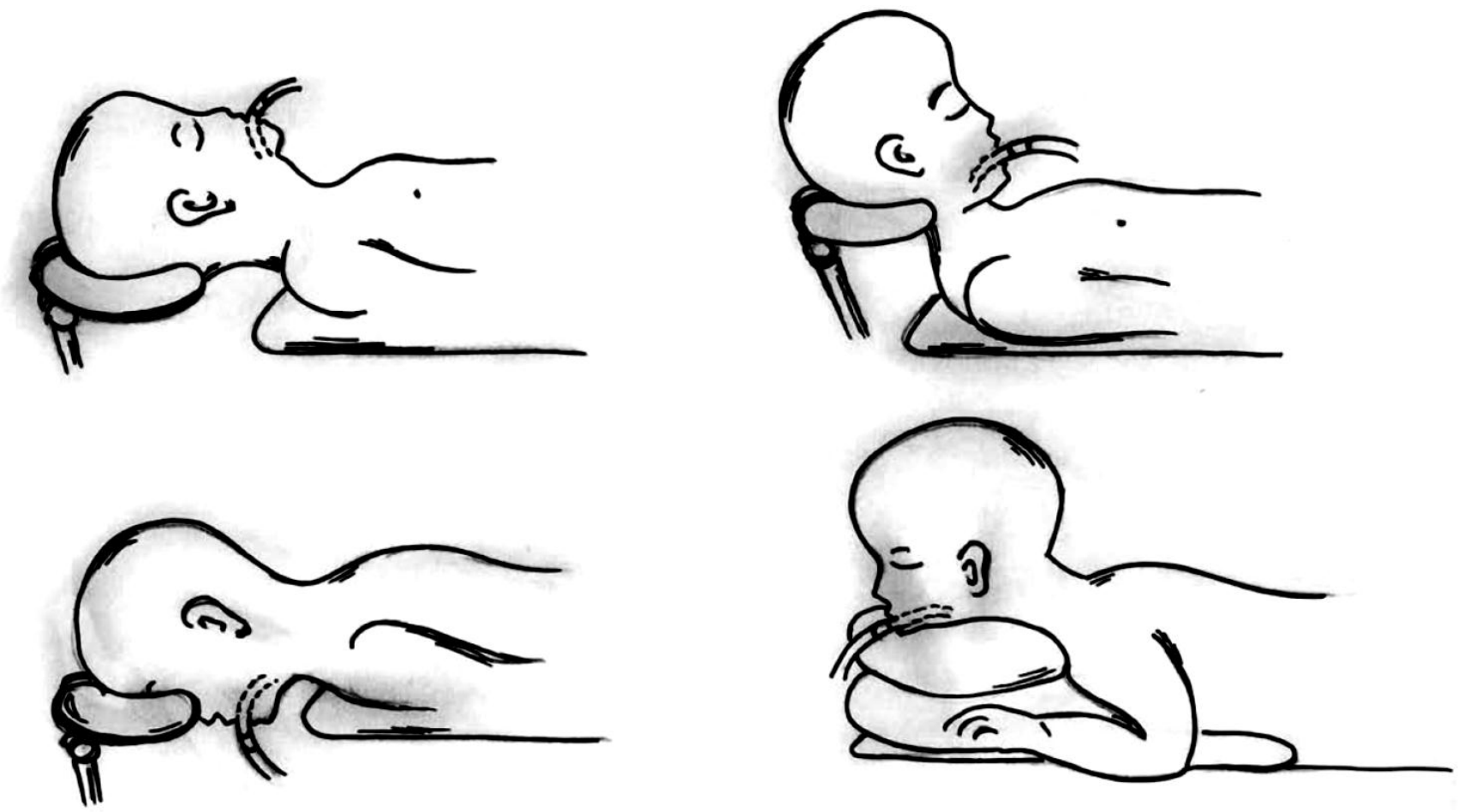


Figure 32.28 Positions in craniosynostosis surgery. (A) Supine position for frontal, frontoparietal and orbitofacial lesions. (B) Supine with head inclination for frontal and parietal and frontobasal lesions. (C) Prone position for parieto-occipital and suboccipital lesions. (D) Prone position with head reclination for total cranial vault exposure.

Anesthesia for Plastic and Craniofacial Surgery, Ehrenfried Schindler, Markus Martini & Martina Messing-Jünger, Gregory's Pediatric Anesthesia, 5. Edition 2012, John Wiley & Sons

Open standard surgery

sagittal synostosis post operative

Technique

total cranial vault reconstruction
no standard bony cutting lines, depending on deformity and bone material
fixation with PDS sutures

timing → around 6th month

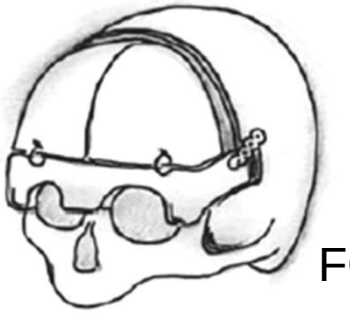
Open standard surgery

Technique

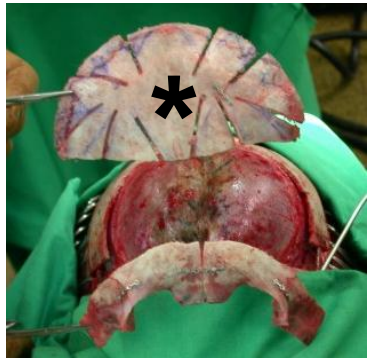
most standardized synostosis surgery
classical fronto-orbital advancement with some hypercorrection (FOA)
bifrontal reconstruction with reshaping* or transposition-osteotomy
rigid fixation (resorbable – non-resorbable)
if non-resorbable
→ explantation necessary after 3 months

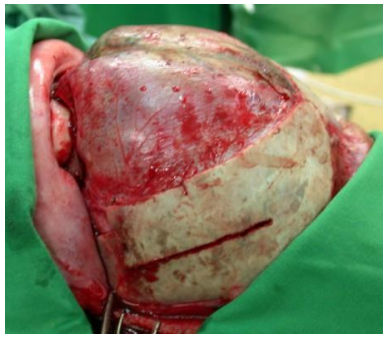
timing → around 6th – 12th month

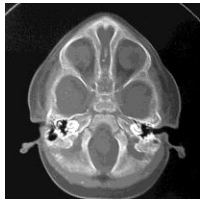
resorbable material sometimes causes local swelling and redness over < 1 year

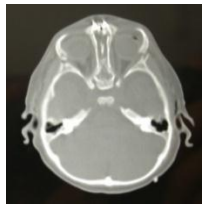


FOA









metopic

Open standard surgery

Technique

most difficult single suture synostosis
to treat
classical fronto-orbital advancement with
some hypercorrection (or one sided)
bifrontal reconstruction with reshaping
rigid fixation (resorbable / non-resorbable)
if non-resorbable
→ explantation necessary after 3 months

timing → around 6th month

resorbable material sometimes causes
local swelling and redness over < 1 year

follow up for ENT- and occlusion problems

unicoronal

Open standard surgery

General remarks

only experienced team on both sides (surgeons and anesthetists)

always crossmatched blood + fresh frozen plasma available

anti-hemorrhagic measures (tranexamic acid)

extubation in theater

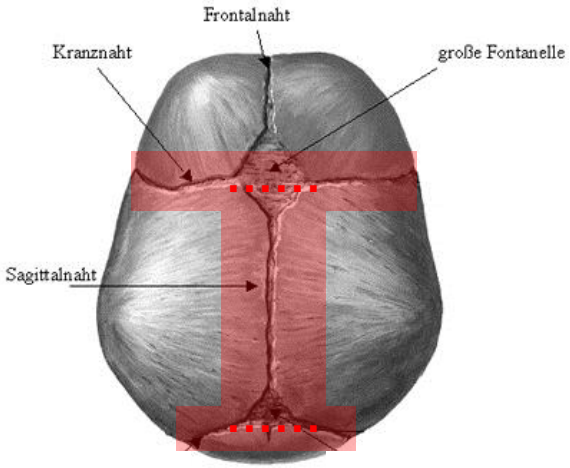
ICU or IMC surveillance over night

avoid postoperative positional plagiocephaly in total cranial vault reconstruction

→ consequent occipital positioning (using pillows or towels) for at least 4-6 weeks

Minimally invasive surgery

(w / wo endoscopic assistance)



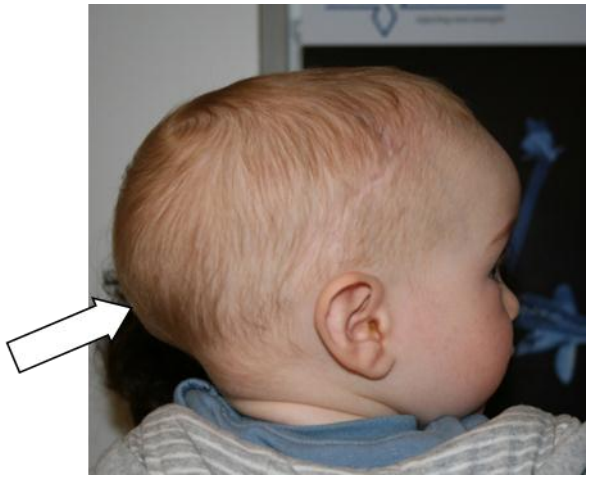
Minimally invasive surgery

(w / wo endoscopic assistance)

best indication: early case of sagittal synostosis

→ early surgery week 8 -12

advantages
→ small skin incisions
→ malleable bone(skull base)
→ most rapid brain growth

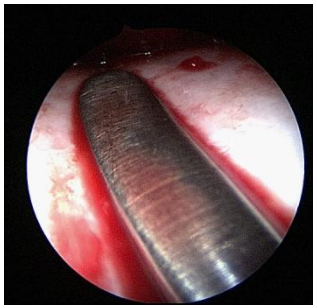


adjuvant helmet therapy

alternatively → active positioning (strictly occipital)
to avoid occipital bossing

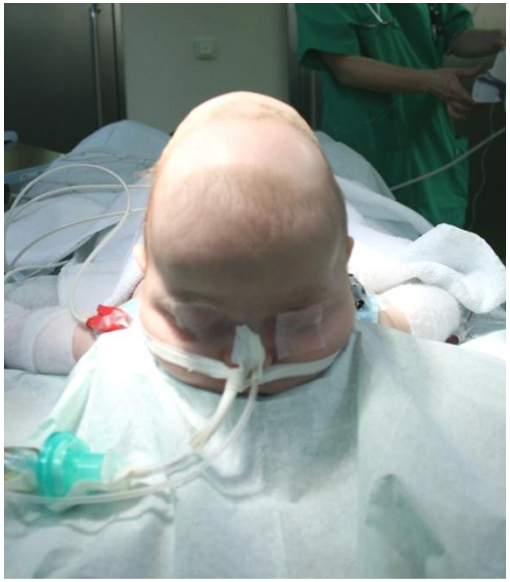
also possible in mild unilateral coronal or metopic synostosis

prone position with head reclined



sagittal synostosis

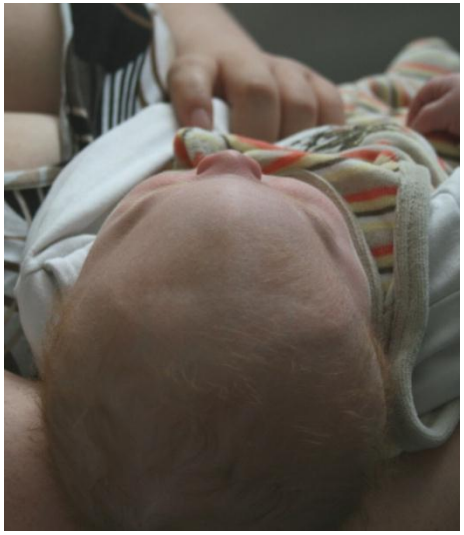
pre OP

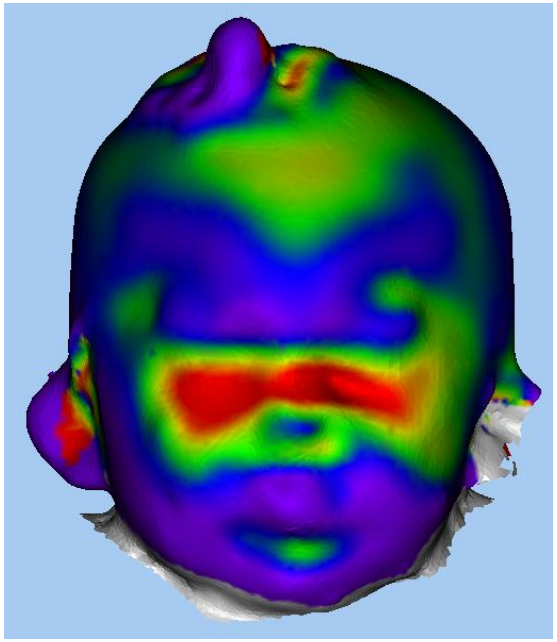


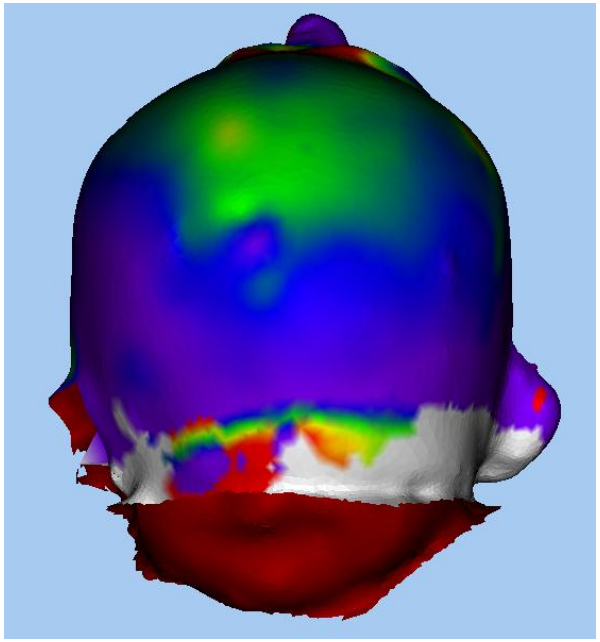
post OP (no helmet)

metopic synostosis







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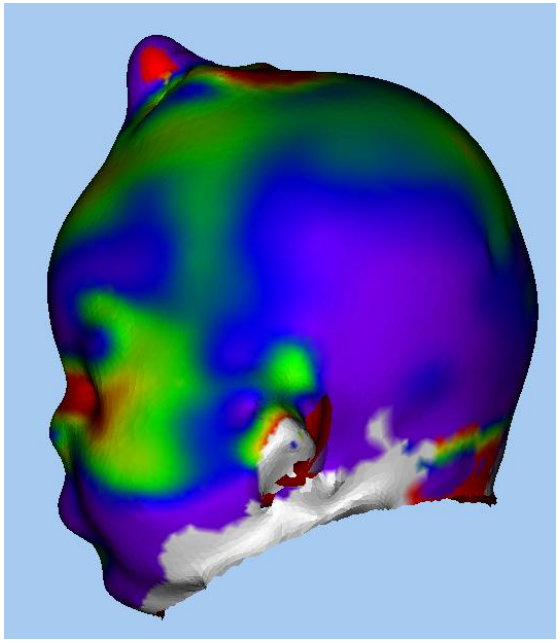
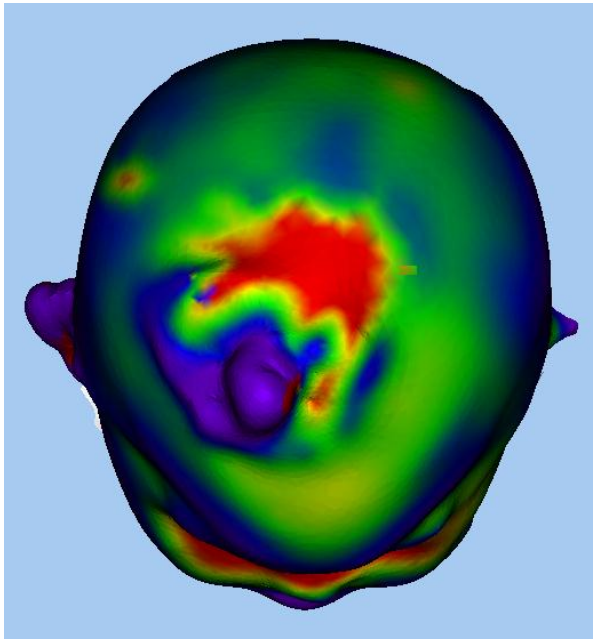
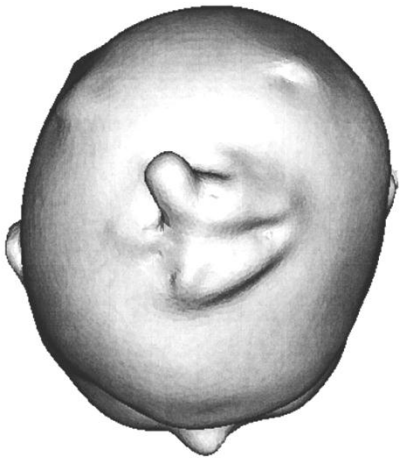
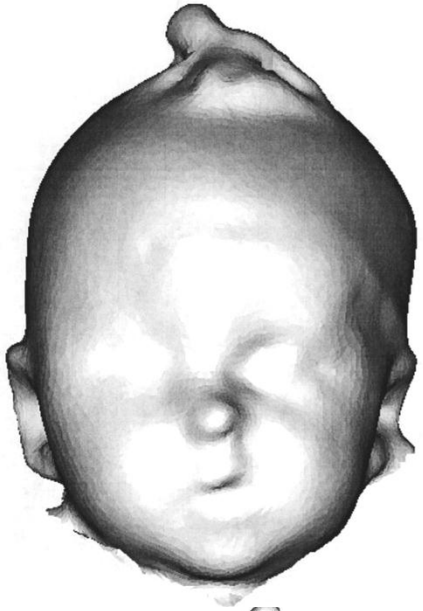
violet/blue: increased

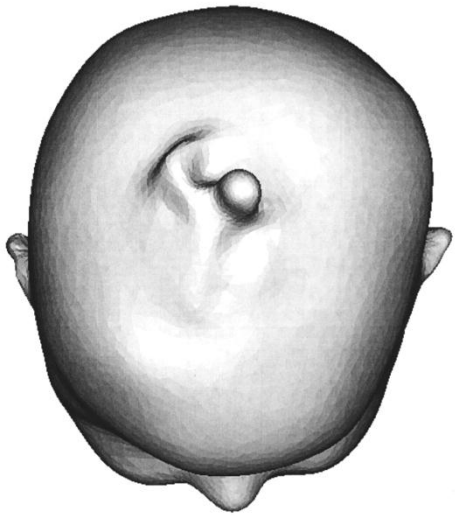


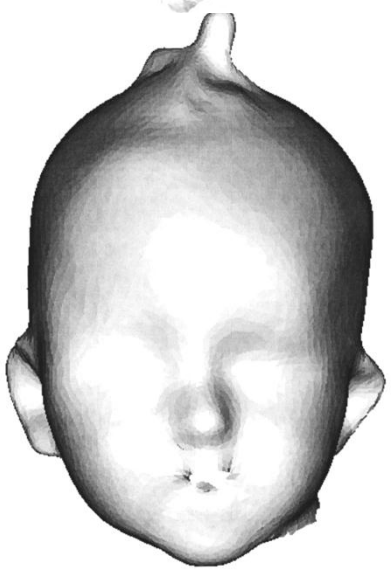


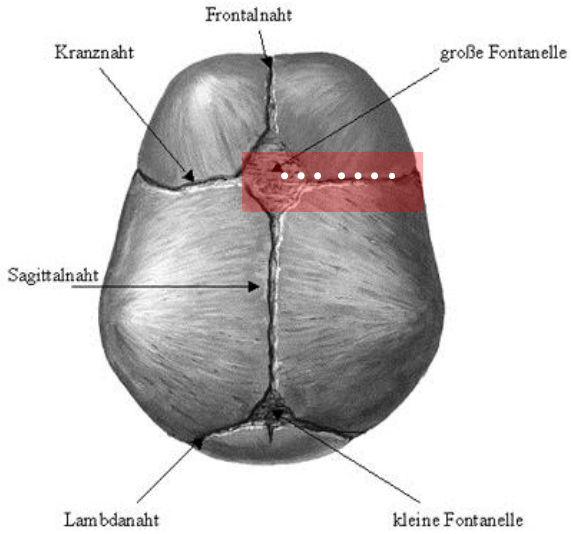
3 months follow up
after minimally invasive surgery
and helmet therapy
in metopic synostosis











3 and 6 months follow up
after minimally invasive surgery
and helmet therapy
in right sided coronal synostosis

Minimally invasive surgery

General remarks

only experienced team on both sides (surgeons and anesthetists)

always crossmatched blood + fresh frozen plasma available

anti-hemorrhagic measures (tranexamic acid)

extubation in theater

ICU or IMC surveillance over night

always Woodbridge tubes to avoid deviation during positioning

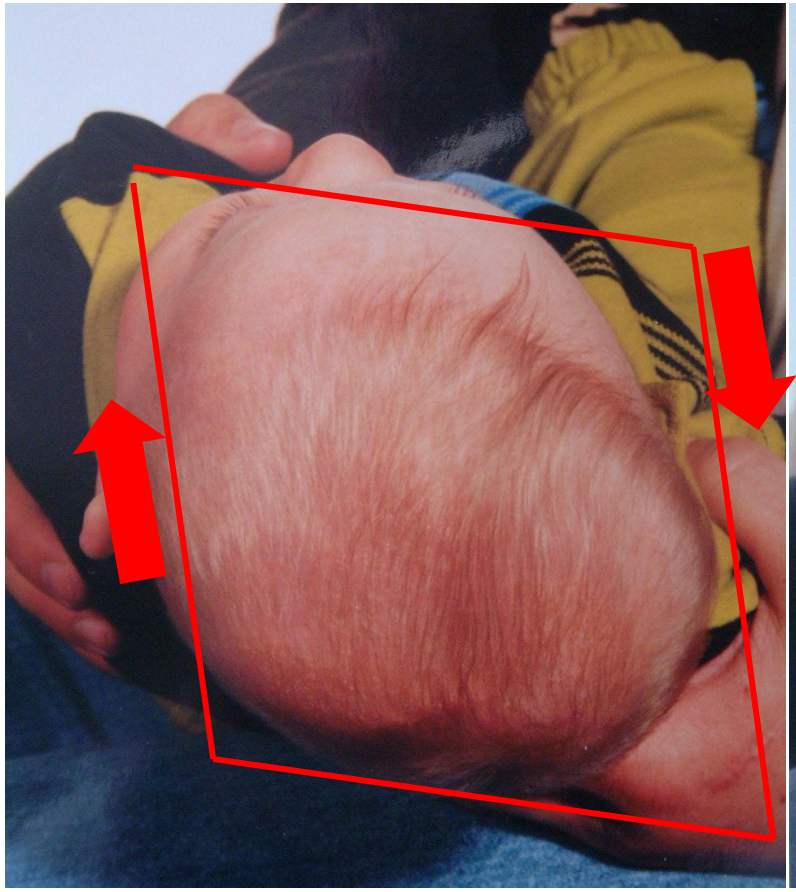
avoid postoperative positional plagiocephaly in minimal invasive suturectomy wo helmet therapy

→ consequent occipital positioning (using pillows or towels) for at least 4-6 weeks

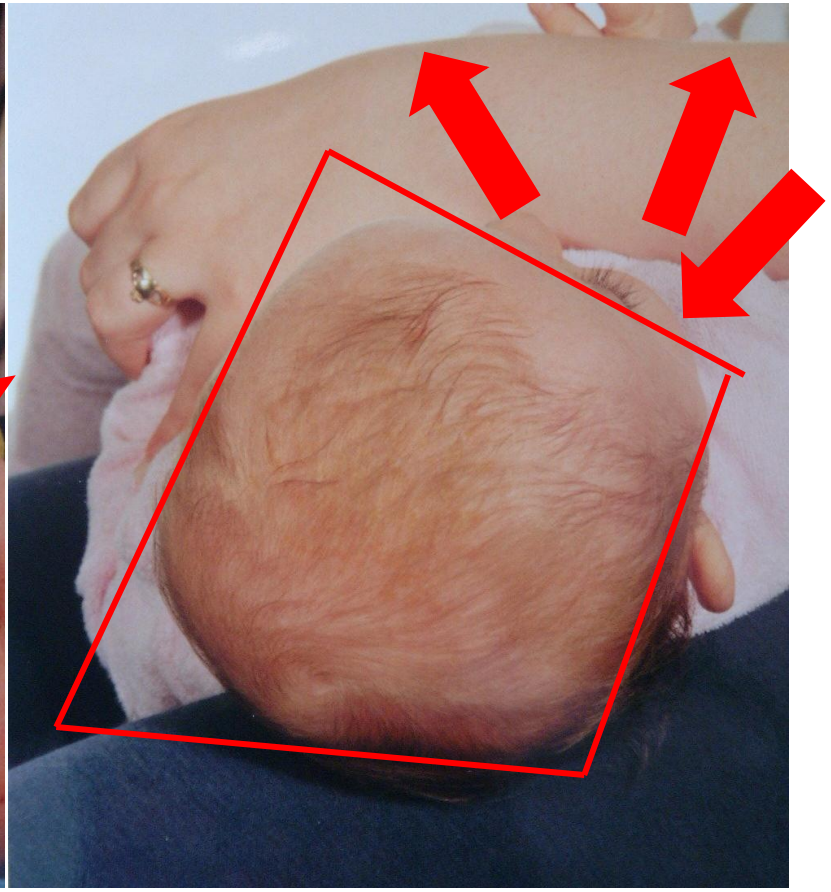
Differentiation between plagio in positional and synostosis cases

positional → head shape = parallelogram
coronal unilateral or lamdoid synostosis → head shape = trapezium

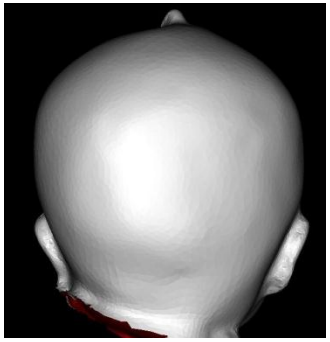
positional plagio

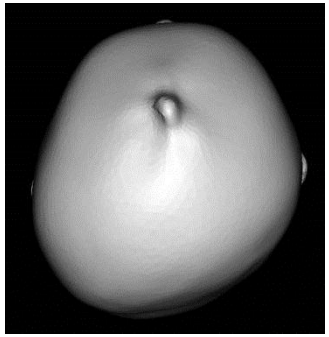


plagio in unicoronal synostosis

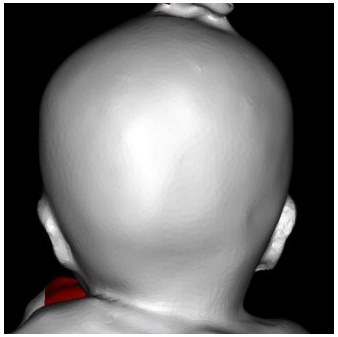


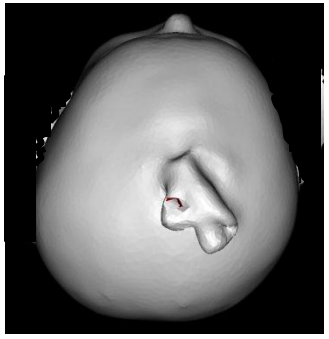
positional plagiocephaly





pre helmet





post helmet

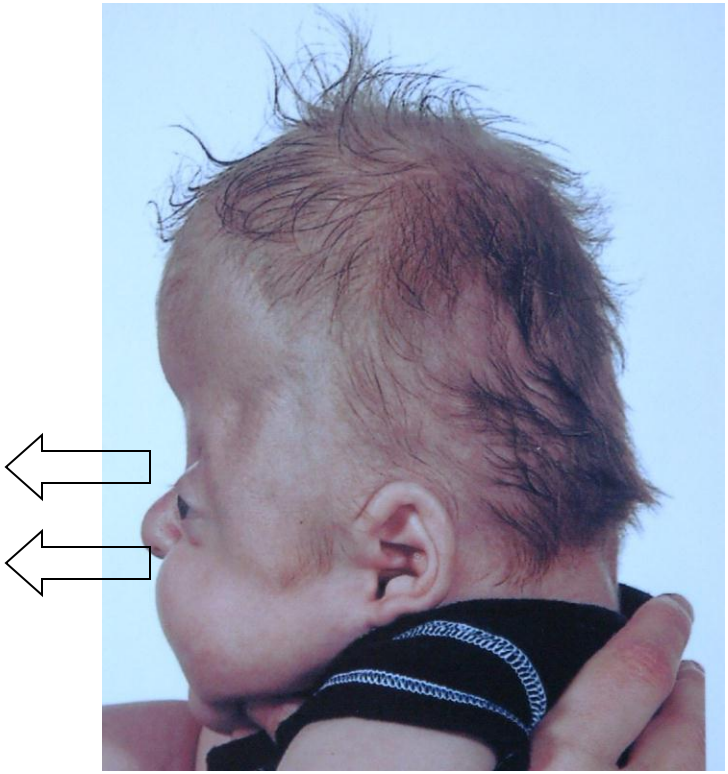
Syndromic cases

cranial vault reconstruction

open total / partial
- morcellation
- transposition
- Rotation
- distraction

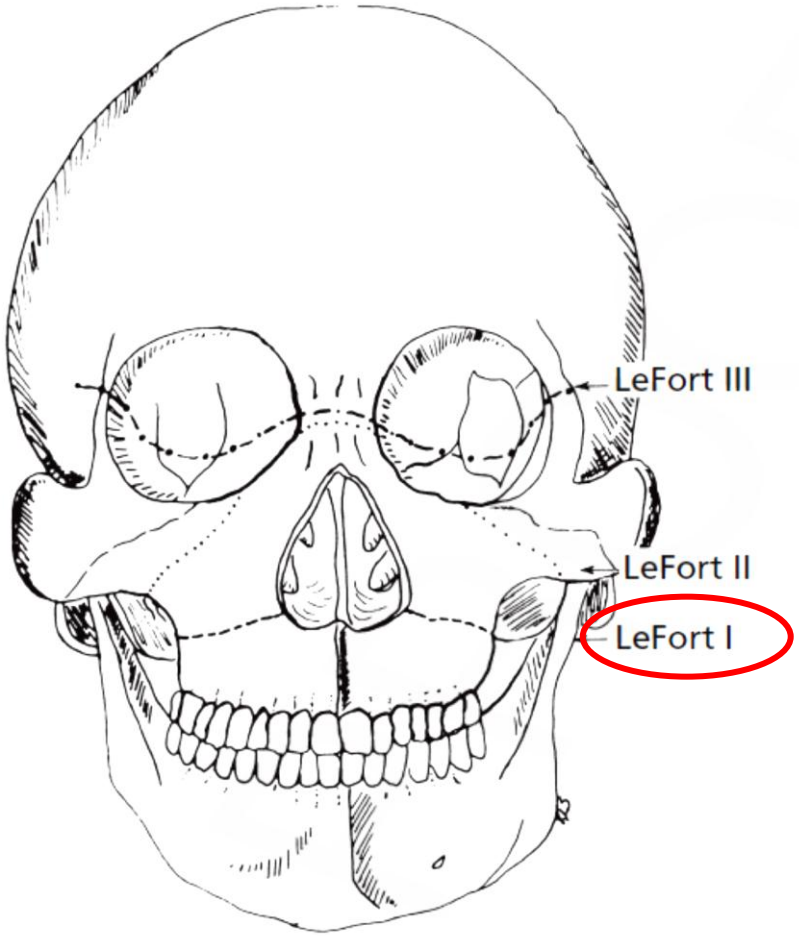
facial reconstruction

fronto-orbital advancement (FOA)
midfacial distraction*
monobloc-distraction
occipital distraction



* after dentation

Midfacial distraction

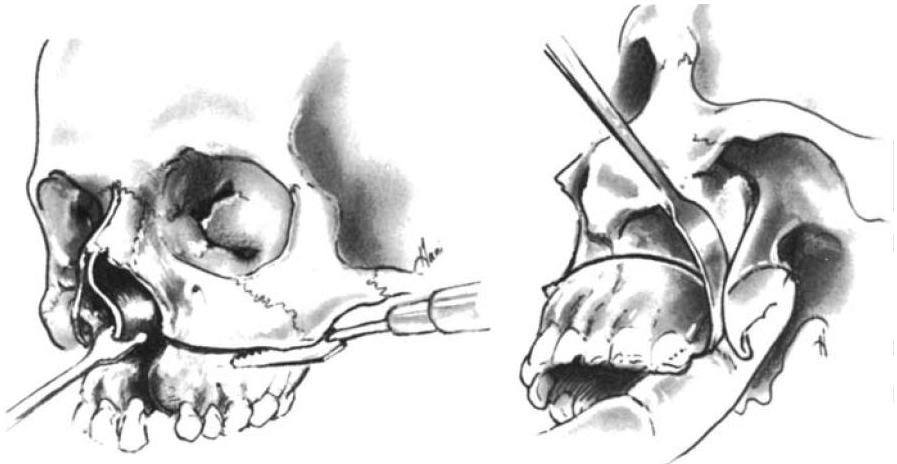


Le Fort osteotomy (I, II, III)

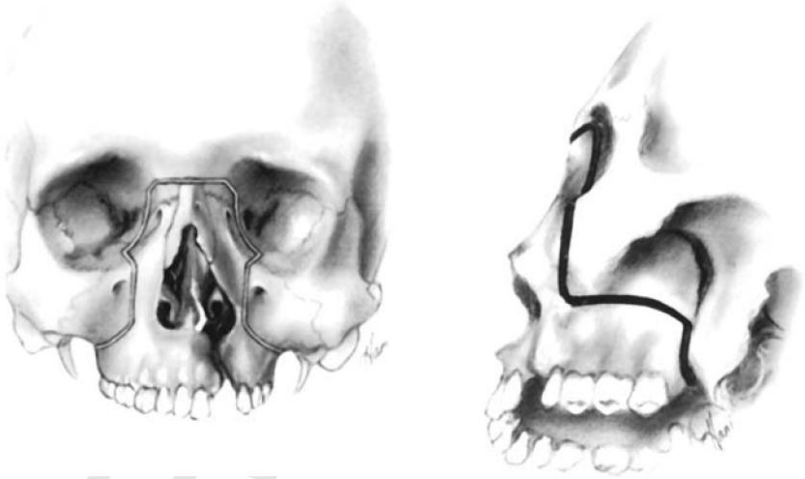
→ preparation for midfacial distraction

Anesthesia for Plastic and Craniofacial Surgery, Ehrenfried Schindler, Markus Martini & Martina Messing-Jünger, Gregory's Pediatric Anesthesia, 5. Edition 2012, John Wiley & Sons

Midfacial distraction



Le Fort I



Le Fort II

Le Fort III

Anesthesia for Plastic and Craniofacial Surgery, Ehrenfried Schindler, Markus Martini & Martina Messing-Jünger, Gregory's Pediatric Anesthesia, 5. Edition 2012, John Wiley & Sons

technical remarks

Ilizarov - principle → distraction osteogenesis

additional neurosurgical treatment options

Gardner – decompression
CSF shunt
ETV

syndromic craniosynostoses

→ often multiple surgeries necessary

Follow up

General remarks

avoid radiation

subjective (surgeon, parents, pediatrician) and objective (measures, scans, fotos)

twice a year ophthalmologic control (papiledema, strabism, astigmatism)

clinical signs of ICP ↑ (following secondary microcephaly or impaired venous drainage)

until 8 to 10 years of age !!!!!

In case of adjuvant helmet therapy → control by surgeon, avoid skull growth restriction and pressure ulcers

Craniosynostoses are interdisciplinary cases – also during follow up !!!!

Many thanks to all actual and recent coworkers, who contributed to the management and documentation of the presented cases:

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Sergey Persits
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Christoph Wiegand
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Roland Albers
Claudia Schmidt
Ehrenfried Schindler
…
and to all parents who gave their consent to use their childrens´ material

Thank you !