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%
Figure 32.2 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.
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 endoscopic 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 reclamation for total cranial vault exposure.
Open standard surgery

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 6\textsuperscript{th} – 12\textsuperscript{th} month

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

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
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
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

→ consequent occipital positioning (using pillows or towels) for at least 4-6 weeks
Differentiation between plagio in positional and synostosis cases

- Positional plagio
  - Head shape = parallelogram

- Coronal unilateral or lamdoid synostosis
  - Head shape = trapezium
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
Midfacial distraction

Le Fort I

Le Fort II

Le Fort III
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 !!!!
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... better to visit a neurosurgeon ...

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