Postoperative Imaging Findings After Cranial and Facial Reconstructive Surgeries in Syndromic Craniosynostosis

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Objective

Review imaging appearances & potential complications following surgical procedures in patients with syndromic craniosynostosis.

Background

Craniosynostosis is a congenital disorder marked by premature fusion of one or more cranial sutures, resulting in abnormal cranial growth. Children with craniosynostosis typically have cranial expansion in the direction parallel to the fused suture, leading to abnormal head shape.

Syndromic craniosynostoses are conditions with additional manifestations (craniofacial and/or extracraniofacial deformities)

Majority of syndromic craniosynostoses are associated with FGFR mutations, demonstrating autosomal dominant inheritance.

Craniofacial treatment procedures in patients with syndromic craniosynostosis can be divided into five phases:

1st Phase (< 6 months of age)

Procedures to treat respiratory distress, sleep apnea, poor nutrition, corneal exposure, hydrocephalus & raised intracranial pressure & Strip craniectomies.

2nd Phase (6-9 months of age)

Fronto-orbital advancement

3rd Phase (9 months to 3 years)

Additional cranial vault remodeling

Monobloc or Craniofacial advancement

4th Phase (3 to 5 years)

Lefort III advancement osteotomy through a subcranial (extracranial) route

5th Phase Adolescence to Adulthood

Lefort III advancement osteotomy combined with Lefort 1 Advancement osteotomy

Lefort II advancement osteotomy

CRANIAL VAULT PROCEDURES can be divided into two broad categories:

Fronto-orbital advancement & remodeling

- Total or subtotal cranial vault remodeling
- Barrel stave osteotomy with cranial remodeling
- Endoscopic suturectomy
- Monobloc advancement &cranioplasty
- Revision cranioplasty

FACIAL/ORTHOGNATHIC SURGERIES FOR CORRECTION OF MIDFACIAL HYPOPLASIA

- Le Fort II and III osteotomies
- Orthognathic surgery/orthodontic preparation (Lefort I, split mandibular osteotomies, and genioplasty)
- Hypertelorism correction (box osteotomies, facial bipartition, subcranial medial orbital translocation).

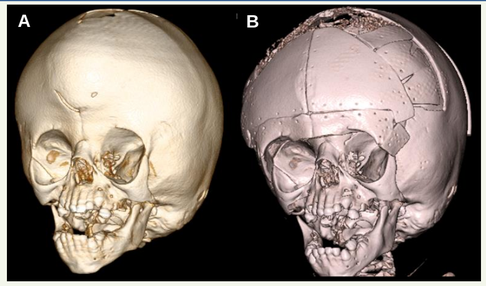


Fig. 1: Fronto-orbital advancement & cranial vault expansion in a 17-mth old F with Apert syndrome: Preop oblique frontal view (A) shows craniosynostosis of the coronal & lambdoid sutures and partial fusion of the sagittal suture & moderate to severe midface hypoplasia. Postop image (B) shows anterior cranial vault expansion and fronto-orbital advancement, coronal suturectomy & barrel stave osteotomies with decreased brachycephaly and better covering of the orbits.

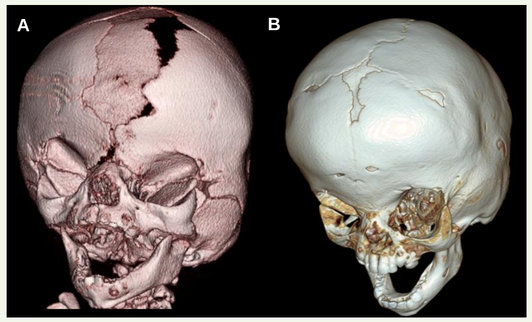
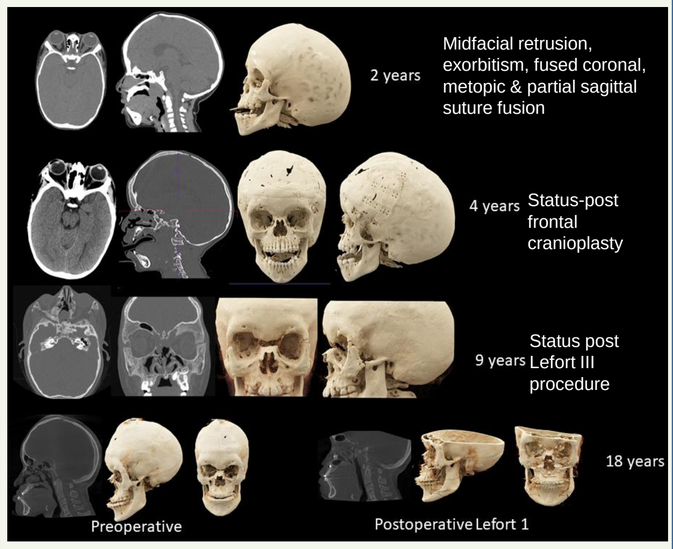


Fig. 2: Endoscopic Suturectomy in a child with Apert syndrome: Preop image (A) at 3 months of age shows bilateral coronal sutural synostosis, widened anterior fontanelle, metopic & sagittal sutures. Left lambdoid suture was also partially fused. Postop image 7 months post endoscopic suturectomy (B) shows bony defects along expected locations of the previously fused right, and to a lesser extent, the left coronal suture.

Fig. 3: Serial surgical procedures in a patient with Pfeiffer syndrome



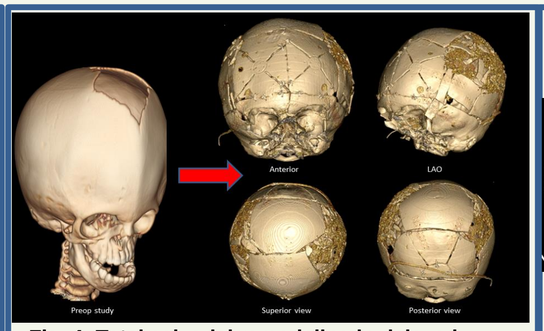


Fig. 4: Total calvarial remodeling in delayed pansynostosis in 3Y with Crouzon syndrome: Frontoorbital advancement (1.3 cm), posterior advancement (1 cm), lateral barrel staving, and reconstruction of apical bone to decrease the severe turribrachycephaly seen on the preoperative CT. Autogenous bone grafting of open defects.

Imaging appearances following facial or orthognathic surgeries for correction of midfacial hypoplasia

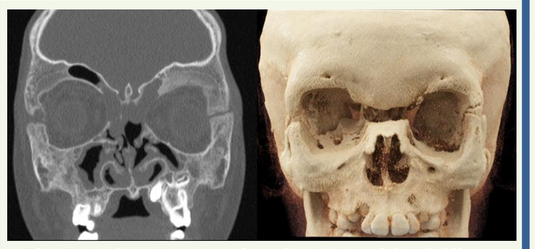


Fig. 5:.Lefort III procedure in 9Y M with Pfeiffer syndrome

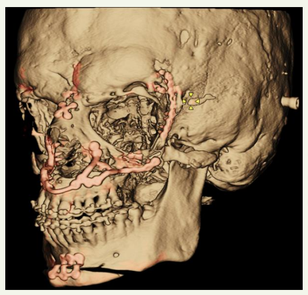


Fig. 6:.Lefort I procedure in a teenager with Pfeiffer syndrome: Lefort 1 osteotomy for maxillary advancement with grafting & genioplasty to correct maxillary hypoplasia, microgenia & malocclusion

Post-surgical complications following craniofacial surgery in patients with syndromic craniosynostosis

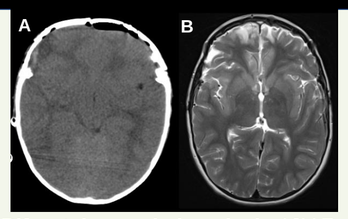
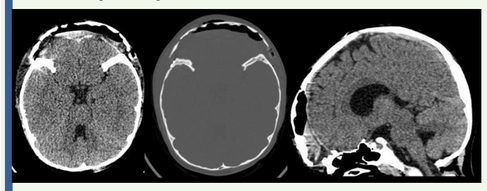


Fig. 7: Bifrontal encephalomalacia:12Y F s/p fronto-orbital advancement shows small right frontal hemorrhage (A). MRI obtained for headaches 1 year post surgery shows bifrontal encephalomalacia

Fig. 8: CSF leak following fronto-orbital advancement & repair of cranial defects in 5Y F with Apert syndrome



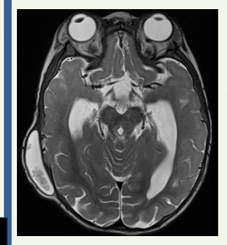


Fig. 9: Focal subgaleal collection following cranioplasty in patient with Crouzon syndrome

Take home points

Treatment protocol in syndromic craniosynostosis can involve several surgical procedures starting from the immediate postnatal period to teenage years. These procedures are preceded & followed in many cases by imaging studies (mainly CT with 3D reconstructions or limited MRIs) to:

- Plan surgical procedures including virtual surgical planning & simulated surgeries on 3D-printed models
- Compare pre-and postoperative landmarks.
- Serially track calvarial growth & effectiveness of craniofacial correction procedures over time.
- Identify complications & undesirable results so that these can be treated in time & minimize long-term functional & cosmetic problems.

Thus it is important to be familiar with expected postoperative imaging appearances and potential complications in this population.

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