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**Letter to Editor**

# Anaesthetic and surgical management of syndromic craniosynostosis in an infant with Apert syndrome amidst geopolitical constraints

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Apert syndrome is a rare autosomal dominant craniofacial disorder caused by mutations in the fibroblast growth factor receptor 2 (FGFR2) gene, essential for cranial and facial bone morphogenesis. It is characterised by polysutural craniosynostosis, midfacial hypoplasia, and complex syndactyly of the hands and feet. Without timely surgical intervention, raised intracranial pressure (ICP) can lead to visual impairment, neurocognitive delays, and structural brain injury.[1] We present the case of an 8-month-old Pakistani infant undergoing the first stage of a multi-stage cranial vault reconstruction, where anaesthetic and surgical challenges were compounded by cross-border geopolitical delays.

The infant, presenting with progressive cranial deformity since birth, was examined and found to be a sociable, alert child with the classic Apert phenotype: acrocephaly, hypertelorism, proptosis, midface retrusion, and syndactyly of all four limbs. Papilloedema and a bulging anterior fontanelle suggested chronically raised ICP (**Figure 1**). The Colorado Pediatric Airway Score (COPUR) was 10, indicating anticipated difficulty in airway management.

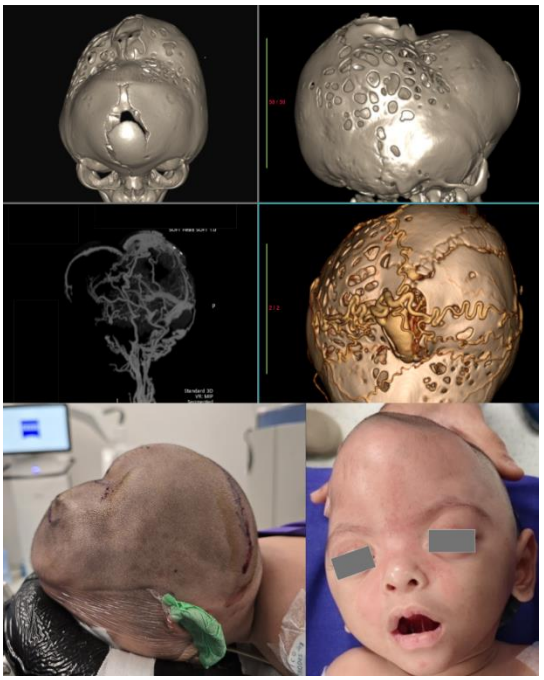

**Figure 1**: Apert phenotype

Systemic examination revealed mild anaemia (Hb 9.8 g/dL), but was otherwise unremarkable. Neuroimaging (MRI and CT) showed multisuture craniosynostosis, with a copper-

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beaten skull, tight brain, and extensive scalp venous collaterals due to superior sagittal sinus compression. Initial assessment was conducted at 5 months, but cross-border visa delays between Pakistan and India postponed surgery by three months, further prolonging the period of elevated ICP.

On the day of surgery, the operating theatre was warmed with underbody blankets to prevent hypothermia. A paediatric breathing circuit with a 500 ml reservoir bag was prepared. Premedication was omitted to maintain baseline neurological status. Anaesthesia was induced with inhaled sevoflurane, followed by intravenous remifentanil (1 µg/kg), propofol (2 mg/kg), and rocuronium (1 mg/kg). A 22G intravenous cannula was inserted in the right hand. Anticipated airway difficulty was managed successfully using a C-MAC videolaryngoscope (Miller size 1 blade) and a size 3.5 mm Micro-Cuff PVC endotracheal tube. Under ultrasound guidance, a 20G, 3Fr catheter was placed into the right femoral artery for invasive blood pressure monitoring, while a 4Fr double-lumen catheter was placed in the right subclavian vein for rapid fluid delivery and potential vasopressor use. The child was positioned prone with meticulous ocular protection using lubricated gauze and Tegaderm. A crystalloid co-load of 20 ml/kg was administered before incision, followed by maintenance fluids at 26 ml/hr. Anaesthesia was maintained with remifentanil infusion (0.05 µg/kg/min), propofol (1 mg/kg/hr), and desflurane (MAC 0.6–0.8).Given the risk of major haemorrhage from vascular scalp collaterals, tranexamic acid was administered (25 mg/kg over 15 minutes, followed by a 0.5 mg/kg/hr infusion).[2] To prevent hypoglycaemia and maintain calcium homeostasis, 10% dextrose with calcium gluconate (30 mg/kg) was infused at a low rate.

Estimated blood loss was 120 ml, replaced with 100 ml packed red blood cells. Neuromuscular blockade was reversed with sugammadex (4 mg/kg), and the patient was extubated smoothly. Analgesia was provided with intravenous paracetamol (15 mg/kg every 8 hours).[3] The postoperative course was uneventful, with ICU observation followed by discharge on day five. The first-stage surgery involved midline sagittal suturectomy to decompress the superior sagittal sinus and reduce venous congestion, combined with spring-assisted posterior cranial vault expansion to create additional intracranial space and reduce ICP.[4,5] The sphinx position, with head elevation above heart level, optimized venous drainage. Scalp incisions were made with sharp-tipped monopolar cautery in coagulation mode.

Intraoperative challenges included dural thinning and prominent bony ridges causing dural impingement. A large venous varix along the midline increased the complexity of suturectomy. Bilateral lambdoid osteotomies were performed, and stainless-steel springs were placed across the bony gaps to facilitate gradual postoperative cranial expansion (**Figure 2**). A second-stage anterior vault reconstruction with orbital advancement was planned to further manage ICP and correct frontofacial deformity. Maxillofacial corrections are reserved for post-pubertal age. However, renewed geopolitical tensions have prevented the timely return for the second stage.

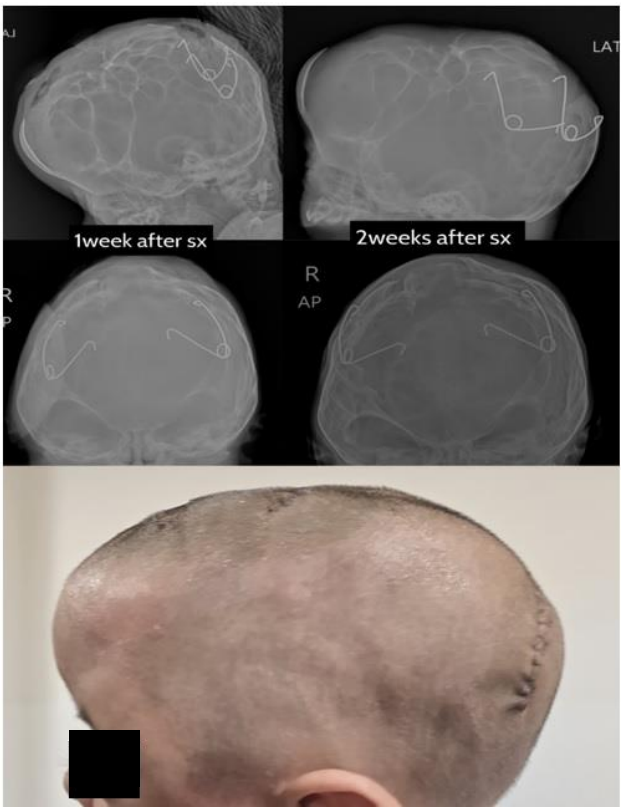


**Figure 2:** Postoperative gradual cranial expansion

This case exemplifies the dual challenges of complex syndromic craniosynostosis—technical demands of surgery and anaesthesia, and the extrinsic barriers imposed by geopolitics. Syndromic patients present unique anaesthetic risks: difficult airway, abnormal venous drainage, increased bleeding potential, and lower physiological reserve. Evidence supports tranexamic acid in reducing perioperative transfusion requirements in paediatric craniofacial surgery. Early invasive monitoring and vigilant fluid management are essential to prevent cerebral hypoperfusion or overload. However, even optimal intraoperative management cannot offset the harm caused by delayed definitive surgery. The initial three-month delay in this case—due to visa restrictions—prolonged the period of raised ICP, potentially affecting neurodevelopment. Now, the indefinite delay in second-stage surgery further diminishes the surgical benefits. This is a real-world example of healthcare's 'butterfly effect,' where diplomatic events far removed from the clinical setting directly impact patient outcomes.

We advocate for the establishment of medical diplomacy protocols—fast-track health visas, cross-border referral agreements, and humanitarian healthcare corridors—to ensure uninterrupted treatment of time-sensitive paediatric conditions, irrespective of the political climate.[6] For craniosynostosis, where early staged intervention is vital,

such measures could be decisive in preventing avoidable disability. This case demonstrates that managing syndromic craniosynostosis in infants demands meticulous, multidisciplinary perioperative planning and technical execution. It also underscores that, without geopolitical solutions to safeguard continuity of care, even the most expertly performed initial surgery may have diminished long-term benefits. Healthcare systems must collaborate across borders to ensure timely access for vulnerable children requiring complex, staged interventions.

## 1. Declaration of Patient Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

## 2. Conflict of Interest

None.

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