Chapter

25

Anaesthesia for Craniofacial Surgery in Children

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Introduction

Craniofacial surgery encompasses the management of congenital and acquired anomalies of the cranium and face. This ranges from single suture craniosynostosis to complex multi-sutural craniofacial and orbital dysostoses, encephalocoeles and craniofacial clefts (Figure 25.1). The surgery may be highly complex and is often protracted, but when conducted within the multidisciplinary setting of a specialist craniofacial unit, the incidence of serious complications is fortunately low.

Optimal perioperative management of these patients is dependent on a multidisciplinary team approach that includes thorough preoperative assessment and preparation, meticulous intraoperative planning, anticipation and proactive management of intraoperative complications and a high level of postoperative care within an appropriate setting.

Craniosynostosis

Craniosynostosis is a congenital condition in which premature fusion of one or more of the cranial sutures occurs, leading to cranial deformity and possible compromised intracranial volume. Craniosynostosis occurs in approximately one in 2,500 live births and can be an isolated finding (non-syndromic synostosis) or part of a multisystem syndrome (syndromic synostosis).

Non-syndromic Synostosis

Non-syndromic (simple) synostosis is the most common form of craniosynostosis, accounting for approximately two thirds of cases. It results from the premature fusion of a single cranial suture, most commonly the sagittal, coronal and metopic sutures. The resulting skull deformity depends on the suture involved (Table 25.1).

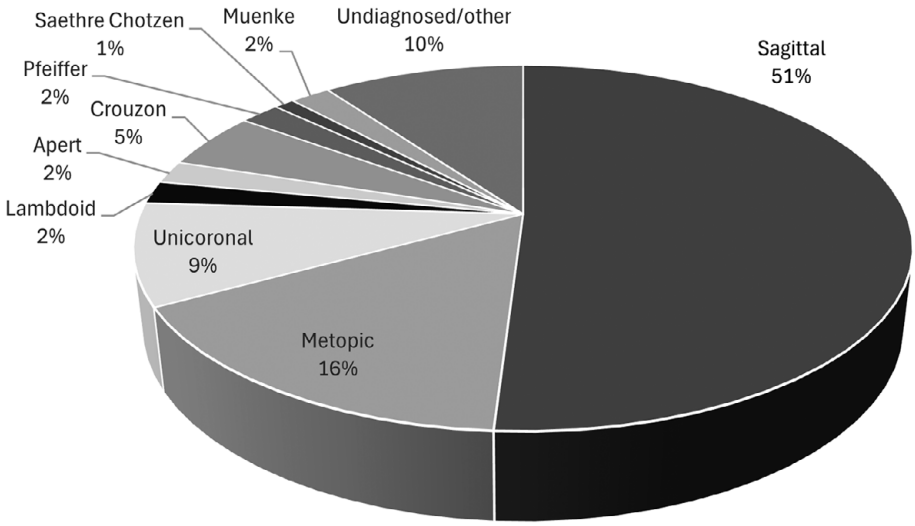

Figure 25.1 Distribution by diagnosis amongst children undergoing major/ complex major cranial vault surgery.
Source: Great Ormond Street Hospital (GOSH) for Children data for 2020–2021.

Table 25.1 Classification of craniosynostosis

Suture	Problem	Solution
Sagittal *1:5,000* *M:F = 2.5–4:1*	Elongated and narrow head *Scaphocephaly*	<6 m: spring-assisted cranioplasty >18 m: total calvarial remodelling (± Melbourne modification)
Metopic *1:7,000–15,000* *M:F = 1.8–2.8:1*	Broad triangular forehead *Trigonocephaly*	Fronto-orbital remodelling/ advancement
Unicoronal *1:10,000* *F:M = 1.6–3.6:1*	Unilateral forehead flattening Contralateral bossing *Anterior plagiocephaly*	Fronto orbital remodelling/ advancement
Bicoronal *1:150,000* *M:F = 1:1.5*	Short, broad head *Brachycephaly*	Fronto orbital remodelling/ advancement (if *not* associated with maxillary hypoplasia)
Lambdoid *1:200,000* *F > M*	Unilateral occipital flattening Contralateral bossing *Posterior plagiocephaly*	Posterior vault expansion/remodelling

Non-syndromic synostosis is usually an isolated anomaly, and there are no associated skeletal defects or other anomalies. Indications for surgery are primarily aesthetic, although raised intracranial pressure (ICP), which occurs in 14% of patients with single suture synostosis, is a functional indication for surgery.

Syndromic Craniosynostosis

Syndromic (multi-sutural) craniosynostosis affects the skull vault and facial skeleton and constitutes up to a third of all cases. The coronal and/or lambdoid sutures are usually affected, often in combination with other sutures. It is a genetically heterogeneous disorder, with most cases showing autosomal dominance and involving mutations in several genes, predominantly the fibroblast growth factor receptor (FGFR1–3) and TWIST 1 genes. Associated malformations are common, and involvement of the facial skeleton often leads to functional problems, including raised ICP, which occurs in approximately half of patients with multi-sutural synostosis, airway obstruction, exorbitism, feeding difficulties and behavioural and psychological problems. There are several craniofacial associated syndromes (Table 25.2), the most common ones being:

- Apert syndrome
- Crouzon syndrome
- Pfeiffer syndrome
- Muenke syndrome
- Saethre–Chotzen syndrome

Timing of Surgery

Craniofacial surgery may be classified according to the region of the craniofacial skeleton involved, the degree of complexity and whether it is performed using an open or minimally invasive technique (Table 25.3).

The ideal timing of surgery varies between institutions, individual surgeons and whether the procedure is to be performed using an open or minimally invasive technique. To achieve lasting results, definitive surgery should ideally be delayed until the period of maximum growth of the affected area is complete. Earlier surgical intervention is performed for minimally invasive procedures, where success is dependent on the presence of pliable bones, or to treat an urgent functional problem such as raised ICP, airway obstruction and exorbitism. Consideration should also be given to the potential psychological impact the deformity has on the child, and waiting for the ideal time to perform reconstructive surgery may not be acceptable to the patient or their parents.

Preoperative Assessment

Two groups of patients present for craniofacial surgery: non-syndromic patients with simple craniosynostosis and syndromic patients with

Table 25.2 Craniofacial associated syndromes

Syndrome	Gene mutation	Synostosis	Features
Apert *1:65,000–1:100,000*	FGFR-2 AD or sporadic	Multisutural (bicoronal)	• Maxillary hypoplasia/hypertelorism ± cleft palate (dental crowding) • Proptosis • Hearing impairment • Choanal stenosis/atresia • Cervical vertebral fusion (C5/C6) • Complex syndactyly, broad thumb • Cardiac defects (~10%) • Developmental delay
Crouzon *1:30,000–60,000*	FGFR-2 (FGFR-3) AD or sporadic	Multisutural (bicoronal ± lambdoid)	• Maxillary hypoplasia/hypertelorism • Proptosis, strabismus • Cervical spine abnormalities • Arnold–Chiari malformation • No hand/feet anomalies • Mostly normal intellect
Pfeiffer *1:100,000*	FGFR-1 FGFR-2 AD or sporadic	Multisutural Type 1: bicoronal Type 2: cloverleaf skull Type 3: no cloverleaf deformity	• Maxillary hypoplasia/ hypertelorism (± cleft palate) • Proptosis, ptosis • Syndactyly, broad thumbs /toes • Multiple other abnormalities (tracheal, cardiovascular, intestinal) • Developmental delay (Type II, III)
Muenke *1:30,000*	FGFR-3 AD	Unicoronal Bicoronal	• Maxillary hypoplasia (mild)/ hypertelorism • Macrocephaly • Proptosis, ptosis • Epilepsy (~10%) • Syndactyly (mild) • Sensorineural hearing loss • ± Developmental delay
Saethre–Chotzen *1:25–50,000*	TWIST-1 AD	Coronal ± metopic	• Facial asymmetry, high forehead • Maxillary hypoplasia/ hypertelorism ± cleft palate • Deviation of nasal septum, convex 'beaked' nasal ridge • Ptosis, strabismus • Syndactyly, clinodactyly of 5th finger, broad big toe • Mostly normal intellect

Notes:
AD = autosomal dominant.
FGFR = fibroblast growth factor receptor.

multi-sutural synostosis and craniofacial dysostoses. In the latter group, a thorough preoperative assessment is essential, with a particular focus on assessment of the airway. Preoperative investigations should be tailored to the patient's clinical status and the proposed surgical procedure. As a minimum, baseline haematological tests should be performed. Other investigations may include sleep studies in patients with obstructive sleep apnoea (OSA) and visual evoked potentials in those at risk of raised ICP (pattern reversal may precede papilloedema or other signs of raised ICP). The

Table 25.3 Classification of craniofacial procedures

Complex major surgery
Craniofacial/midface
• Monobloc frontofacial advancement for anterior-posterior and vertical anomalies
• Frontofacial bipartition for three-dimensional anomalies of the skull, face and orbits
• Orbital box osteotomy for lateral or vertical orbital anomalies
Major surgery
Cranial vault/ transcranial
• Fronto-orbital remodelling/advancement for metopic and coronal synostosis
• Total calvarial remodelling (± Melbourne modification) for sagittal synostosis
• Posterior vault expansion for lambdoid synostosis
Subcranial
• Le Fort osteotomies (commonly Le Fort III)
Cranial vault
• Spring-assisted cranioplasty for sagittal, lambdoid synostosis
• Endoscopic-assisted suturectomy for metopic, unicoronal, sagittal synostosis

appropriate volume of blood should be cross-matched and ready for use prior to surgery. This should be guided by the hospital's Maximum Surgical Blood Order Schedule (MSBOS), but one unit of packed red cells are usually adequate for major procedures and two to four units for complex major procedures.

Airway Assessment

Craniofacial abnormalities are often associated with airway problems, and up to half of patients with syndromic craniosynostosis will present with some degree of supraglottic airway compromise secondary to maxillary hypoplasia. This ranges in severity from mild nasal obstruction to severe OSA. Patients with moderate to severe symptoms require a preoperative sleep study. Symptomatic airway obstruction is managed preoperatively with nasal prong airways or nasal continuous positive airway pressure (CPAP). Surgical options include adenotonsillectomy, uvulopharyngo-palatoplasty or, in the most severe cases, tracheostomy.

Physical appearance may be an unreliable predicter of a potentially difficult airway in some craniofacial patients. Whilst the airway of some syndromic patients with maxillary hypoplasia may look challenging, they rarely present any difficulty at laryngoscopy. On the contrary, syndromic patients whose physical appearance has been improved through corrective surgery may be difficult to intubate due to maxillary advancement and reduced temporomandibular joint movement. The presence of a rigid external distraction (RED) frame may also make laryngoscopy awkward if not impossible.

Indications for Tracheostomy

A minority of syndromic patients with severe airway obstruction and OSA may present for surgery with a tracheostomy in situ. The decision to perform a covering tracheostomy is made on an individual patient basis following a multidisciplinary team discussion with members of the craniofacial, ENT and anaesthetic teams and the patient (if appropriate) and their parents. Indications for a covering tracheostomy include:

- Severe and deteriorating upper airway obstruction, including life-threatening OSA.
- Patients less than two years of age scheduled for complex major surgery, such as frontofacial advancement
- Patients undergoing extensive facial osteotomies where reintubation may be difficult

When a decision is made to perform a covering tracheostomy, it is best done as a separate procedure at least two weeks prior to the scheduled craniofacial surgery. This ensures the presence of a well-formed tracheostome and avoids the potential risk of iatrogenic tension pneumothorax developing due to positive pressure ventilation during prolonged complex major surgery. A tracheostomy may also be associated with an increased risk of surgical infection, and subsequent decannulation may be challenging.

Premedication

If intravenous induction of anaesthesia is planned, a topical anaesthetic cream should be applied preoperatively to potential cannulation sites to facilitate cannulation. Patients who are anxious or those with significant behavioural problems may benefit from preoperative psychological preparation with or without the use of sedative premedication.

Caution should be exercised when using sedative premedication in children with severe airway obstruction or raised ICP, although this may be necessary in some cases and is generally well tolerated. Anti-embolism compression stockings (TED™) should be applied in older patients and in those undergoing prolonged surgery.

Conduct of Anaesthesia

Induction

Induction of anaesthesia may be achieved using an inhalational or intravenous technique, the technique chosen depending on the preference of the patient, parent and anaesthetist. Inhalational induction may be preferable where a difficult airway is anticipated. Prior to induction of anaesthesia, standard monitoring should be applied, with a pulse oximeter being an absolute minimum.

Airway Management

Following induction of anaesthesia, maintenance of the airway may be challenging in patients with syndromic craniosynostosis. Relaxation of the upper airway compounds pre-existing supraglottic airway obstruction, and it may be difficult to achieve an adequate face mask seal due to facial asymmetry, maxillary hypoplasia and exorbitism. Use of a soft-seal face mask, rotated through 180° if necessary, combined with simple airway manoeuvres and adjuncts, may be necessary to achieve adequate mask ventilation. Application of CPAP via a close-fitting face mask will also help in alleviating upper airway obstruction.

Difficult laryngoscopy is unusual in patients with midface hypoplasia, although children who have previously had corrective surgery may be more difficult to intubate. For these cases, a full range of intubation equipment, including a laryngeal mask airway, should be readily available in theatre.

The choice of tracheal tube is dependent on the type of surgery. A pre-formed south-facing Ring-Adair-Elwin (RAE) orotracheal tube is suitable for patients undergoing surgery in the supine position. A reinforced orotracheal tube with secure facial strapping is required for patients undergoing surgery in the prone position. Where intraoral surgical access is required, a reinforced orotracheal tube secured with circum-mandibular wiring is preferred at our institution. The tracheal tube should be optimally positioned to avoid accidental extubation or inadvertent bronchial intubation. This may best be achieved by deliberate endobronchial intubation followed by withdrawal of the tube until breath sounds can be auscultated bilaterally. The tube should be taped securely and the position rechecked with the head in flexion and extension. Patients presenting for surgery with a tracheostomy in situ should have the tracheostome intubated with a reinforced tube secured to the anterior chest wall.

Intravascular Access

At least two appropriately large bore cannulae should be sited for complex surgery and complex major surgery. This may be challenging in syndromic patients with syndactyly or other limb abnormalities, but ultrasound has made this task much easier. The long saphenous vein is a particularly useful vessel, as it allows cannulation with a relatively wide bore cannula that is easily accessible intraoperatively. All children undergoing major and complex major craniofacial surgery require an arterial line to facilitate continuous arterial blood pressure monitoring and serial measurement of haemoglobin and blood gases. A femoral central venous line and monitoring of central venous pressure (CVP) is useful in patients undergoing prolonged complex major surgery.

Antibiotic cover is administered after induction of anaesthesia and prior to the commencement of surgery. Local policies should dictate the choice of antibiotics.

Maintenance

Maintenance of anaesthesia is achieved using a balanced technique involving a volatile inhalational agent such as isoflurane or sevoflurane delivered in a mixture of oxygen in air with fentanyl or a remifentanil infusion for analgesia. A total intravenous technique using propofol may be considered in older children. In addition to intravenous opioids, multimodal analgesia includes the preoperative infiltration of a tumescent solution containing triamcinolone acetate, lignocaine, levobupivacaine, hyaluronidase and adrenaline (up to 7 ml kg^{-1}) and administration of intravenous paracetamol. Intravenous morphine (up to 0.1 mg kg^{-1}) is titrated towards the end of surgery, especially if remifentanil has been used intraoperatively.

Intraoperative Monitoring and Patient Positioning

Standard monitoring includes electrocardiogram (ECG), oxygen saturation (SpO_2), end-tidal carbon dioxide ($ETCO_2$), invasive blood pressure, central and peripheral temperature and peripheral nerve function. Measurement of central venous pressure may be useful in patients undergoing prolonged major or complex major surgery. Maintenance of normothermia is essential and is best achieved using a forced-air warming blanket and fluid warming device with careful monitoring to avoid the risks of inadvertent hyperthermia. As the duration of surgery may exceed six hours, meticulous attention should be paid to patient positioning:

- For most craniofacial procedures, the supine, head-up (semi-Fowler's) position is used to reduce cerebral venous congestion.
- Prone positioning is used for sagittal spring cranioplasty, posterior vault expansion and total calvarial remodelling. In this position, it is important to ensure that the abdomen is free to allow unrestricted diaphragmatic movement with ventilation and avoid venous congestion.
- In all cases, pressure points should be adequately padded to avoid cutaneous or peripheral nerve injury.
- The eyes should be well lubricated and adequately protected. This may be difficult in patients with marked exorbitism in whom suturing of the palpebral fissures may be required (tarsorrhaphy).

Management of Blood Loss

Extensive blood loss and massive transfusion may be unavoidable and remains the greatest risk to infants and children undergoing complex major craniofacial surgery. Haemorrhage may arise from the extensive scalp incision, bone edges or less commonly from extradural venous or sinus tears. Intraoperative blood loss may be rapid and can easily exceed a patient's estimated blood volume, especially in infants and young children. Accurate assessment of blood loss may be difficult due to loss onto surgical drapes and gowns as well as dilution by irrigation fluid used for the surgical power tools.

Intravascular volume replacement and transfusion requirements must therefore be guided by careful monitoring of the following parameters:

- Vital signs: ECG/heart rate, arterial blood pressure and waveform, capillary refill time, core–peripheral temperature gradient, central venous pressure, urine output
- Acid-base status (pH, BE, HCO_3, lactate)
- Haemoglobin concentration and haematocrit
- Coagulation parameters and platelet count

The goals of intraoperative fluid management are to maintain a normal circulating volume, an appropriate haemoglobin concentration, normal electrolyte balance and normoglycaemia. Of paramount importance is the avoidance of hypovolaemia and maintenance of adequate organ perfusion. This is best achieved through meticulous matching of transfusion volumes and blood loss using a combination of balanced salt solution (e.g. Hartmann's solution) and packed red cells to maintain an acceptable haemoglobin concentration. As fluid dynamics can change very rapidly, a proactive approach should be adopted, with all fluids being warmed prior to infusion. Transfusion practice also depends on the experience of the anaesthetist, and whilst blood loss is often underestimated, the risks of over-transfusion remain, especially in infants and young children. In addition to intravenous fluids, vasopressors (e.g. phenylephrine) may be required in the management of intraoperative hypotension.

Reducing Allogeneic Blood Transfusion

Despite donor blood being extensively tested and monitored, allogeneic blood transfusion is not without risk from viruses, reactions or the incorrect blood being transfused. Complications associated with the massive transfusion of allogeneic blood, including metabolic acidosis, hyperkalaemia, hypocalcaemia and coagulopathy, may also be associated with significant morbidity. Strategies should therefore be employed to avoid or at least reduce the use of allogeneic blood.

Preoperative Measures

- Iron deficiency anaemia should be treated with iron supplements.
- Recombinant erythropoietin therapy should be applied. Preoperative elevation of haemoglobin with erythropoietin has been shown to reduce

transfusion requirements in children undergoing craniofacial surgery, but an unfavourable cost-benefit ratio limits its usefulness, and it is not used in patients undergoing craniofacial surgery at the author's institution.
- Preoperative autologous blood donation is not used routinely in children in the United Kingdom.

Intraoperative Measures

- Patient positioning (supine, head-up semi-Fowler's position).
- Scalp infiltration with an adrenaline-containing tumescent solution.
- Meticulous surgical technique should be used with particular attention to incremental or marginal losses at each stage of surgery and use of fibrin sealants or fibrin glue.
- Antifibrinolytics (e.g. tranexamic acid) are commonly used for major and complex major surgery and have been shown to reduce blood loss and the requirement for blood transfusion in children undergoing craniosynostosis surgery. The optimal dose is still a matter of debate, but at the author's institution a loading dose of 10–15 mg kg^{-1} is administered, followed in some cases by an infusion of 5 mg kg^{-1} h^{-1}.
- Intraoperative cell salvage can be a safe and effective means of reducing intraoperative allogeneic blood use. The newer blood salvage machines have relatively fast processing times and low processing volumes and provide a continuous supply of washed red cells in proportion to the rate at which the patient is bleeding. Despite this, cell salvage alone is often inadequate for infants undergoing craniofacial surgery. The overall efficacy of red cell recovery by cell salvage is dependent on the ability to recover the blood lost in a useable form. This comprises suction losses as well as blood loss to surgical swabs. Washing the blood-soaked swabs in a normal saline/heparin solution can increase the amount of recoverable blood by 30–50%.
- Induced hypotension may be beneficial in the older child undergoing midfacial advancement but has not been widely adopted owing to the increased risk of intraoperative venous air embolism (VAE) and rapid blood loss. There are several agents that may be used to achieve moderate intraoperative hypotension, a commonly used combination being remifentanil and clonidine. The use of induced hypotension in infants and young children undergoing craniosynostosis repair is controversial and is not practised at the author's institution.
- Acute normovolaemic haemodilution is limited by the child's relatively small circulating blood volume and so has limited value.

At the author's institution, preoperative optimisation of iron levels where possible, meticulous attention to patient positioning (particularly when in the prone position with avoidance of venous congestion) and surgical technique and the use of tranexamic acid and intraoperative cell salvage for complex major cases are routinely used.

Postoperative Measures

- Lower the transfusion threshold in otherwise healthy children (currently 70 g l^{-1} at the author's institution in haemodynamically stable children with no pre-existing cardiac disease).
- Iron supplements may be used. Oral iron therapy may be given to children with a low haemoglobin. Intravenous iron therapy is not used at the author's institution.
- Postoperative cell salvage involves the collection of blood from surgical drains followed by reinfusion, with or without a wash cycle. Although there have been concerns about the safety of transfusing unwashed red cells, postoperative cell salvage is commonly used in orthopaedic surgery. Despite the potential benefits, postoperative cell salvage is not currently used for craniofacial surgery in the United Kingdom.

Management of Dilutional Coagulopathy

Dilutional coagulopathy may occur in patients undergoing complex major craniofacial surgery. The intraoperative use of Octaplas, a pooled plasma solution (10–15 ml kg^{-1}), should be determined by the extent of blood loss, clinical evaluation and serial measurement of coagulation parameters. If blood loss persists in the presence of a low fibrinogen level (<0.8 g l^{-1}), use of a highly concentrated source of fibrinogen such as

cryoprecipitate (5–10 ml kg^{-1}) may be required. In the author's experience, platelets are rarely required, although they may be necessary in some complex major cases in the presence of demonstrable thrombocytopenia ($<50 \times 10^9$ l^{-1}).

Intraoperative Complications

Intraoperative complications are fortunately uncommon but may include:

- Unintentional extubation, endobronchial intubation or severing of the tracheal tube pilot balloon.
- Acute bradycardia secondary to the oculocardiac reflex. This may occur in response to orbital manipulation and responds in most cases to immediate cessation of the stimulus. Occasionally a vagolytic such as glycopyrrolate or atropine is required.
- VAE. Use of the head-up position places patients at increased risk for VAE. The incidence of VAE during surgery for craniosynostosis has been reported as high as 83%, although only 1–2% were clinically significant with haemodynamic compromise. At the author's institution, capnography is used to monitor for clinically significant VAE, and the incidence of haemodynamically significant VAE is fortunately very low.
- Sudden major blood loss resulting in hypotension and impaired organ perfusion. This responds to fluid resuscitation in most cases, although a vasopressor such as phenylephrine is often necessary.
- Significant metabolic acidosis. Children undergoing craniofacial surgery may develop a varying degree of intraoperative metabolic acidosis as evidenced by an increased base deficit. This occurs despite the patient being haemodynamically stable and clinically well perfused and is most likely the result of hyperchloremic acidosis. In the author's experience, the base deficit usually reverts to baseline over a period of approximately 12 hours.

Postoperative Care

Most children undergoing major and complex major craniofacial surgery can be extubated at the end of surgery. Following a period of stabilisation on the recovery ward, further postoperative care should be provided on a neurosurgical high-dependency unit where invasive monitoring can be continued. Admission of a patient to the intensive care unit for mechanical ventilation is uncommon at the author's institution but may occasionally be required in the event of an unexpected or significant intraoperative complication.

Bilateral nasopharyngeal airways are routinely used in children at risk of postoperative upper airway obstruction following complex major surgery, such as frontofacial advancement. These remain in situ for up to five days or until such time as the facial oedema has resolved. To reduce the potential for further facial oedema within the initial 24–48 hours postoperatively, patients are nursed in a head-up position. Frost sutures (between the lower eyelid and eyebrow) may also be used in this group of patients to protect the cornea.

Fluid Management

Fluid losses may continue into the initial postoperative period, and meticulous attention should be paid to postoperative fluid management with ongoing monitoring of haemodynamic parameters, drain losses and coagulation profile. A falling haematocrit in association with minimal drain losses should alert one to the possibility of an intracranial haemorrhage. This constitutes an emergency for which an immediate neurosurgical opinion should be sought.

Pain Management

It is important that adequate pain control is achieved prior to transfer of a patient to the neurosurgical high-dependency unit. This may involve the titration of further doses of intravenous morphine in the recovery ward. Thereafter, satisfactory analgesia can be achieved in most patients with a combination of regular intravenous paracetamol (15 mg kg^{-1} every six hours), oral morphine (0.1–0.2 mg kg^{-1} every four hours) as required and a non-steroidal anti-inflammatory drug (e.g. ibuprofen, diclofenac) once the clotting profile has normalised. Intravenous morphine delivered via a patient- (PCA) or nurse-controlled analgesia (NCA) device is used for patients following complex major surgery. It may also be used following some major procedures (e.g. fronto-orbital

remodelling) but may be associated with an increased incidence of nausea and vomiting and delayed mobilisation.

Postoperative nausea and vomiting are common following craniofacial surgery, and dual antiemetic therapy should be used.

Specific Craniofacial Procedures

Minimally Invasive Surgery

Minimally invasive approaches to the surgical management of craniosynostosis is gaining wider acceptance, and early suturectomy with some form of distraction to passively reshape the head has become an accepted treatment for single-suture craniosynostosis. Surgical techniques include spring-assisted cranioplasty, commonly performed for sagittal synostosis and for posterior vault expansion. More recently, endoscopic suturectomy with helmet therapy (ESCH) has emerged as a successful treatment for metopic, unicoronal and sagittal synostosis. Minimally invasive surgery offers many advantages over traditional open techniques, including shorter operative times, less blood loss, fewer transfusions and a reduced length of hospital stay.

Spring-Assisted Cranioplasty

Dynamic distraction with steel springs is increasingly being used in the management of sagittal and lambdoid craniosynostosis. Spring-assisted cranioplasty is the treatment of choice for sagittal synostosis at the author's institution and is increasingly being used for posterior vault expansion.

Spring-Assisted Sagittal Cranioplasty

Spring-assisted cranioplasty involves performing a small craniectomy straddling the sagittal sinus, approximately halfway along the fused suture, followed by two parasagittal osteotomies. Two titanium springs are then placed across the defect to gradually separate the bones (Figure 25.2). Surgery should be performed between three and six months of age as the development of frontal bossing in late infancy precludes the use of this technique. Spring removal is typically undertaken three months later through the same incision as a day-case procedure.

This minimally invasive procedure has had an enormous impact on the surgical management of infants with sagittal synostosis, and it has several advantages over the traditional open technique. A quantitative analysis of 100 consecutive cases at the author's institution showed a reduced operating time (~1 h vs 2–3 h), smaller scar with minimal dural dissection, minimal blood loss, reduced blood transfusion (9% vs ~ 95%), rapid recovery and shorter hospital stay (one night vs three nights) compared with alternative treatment strategies.

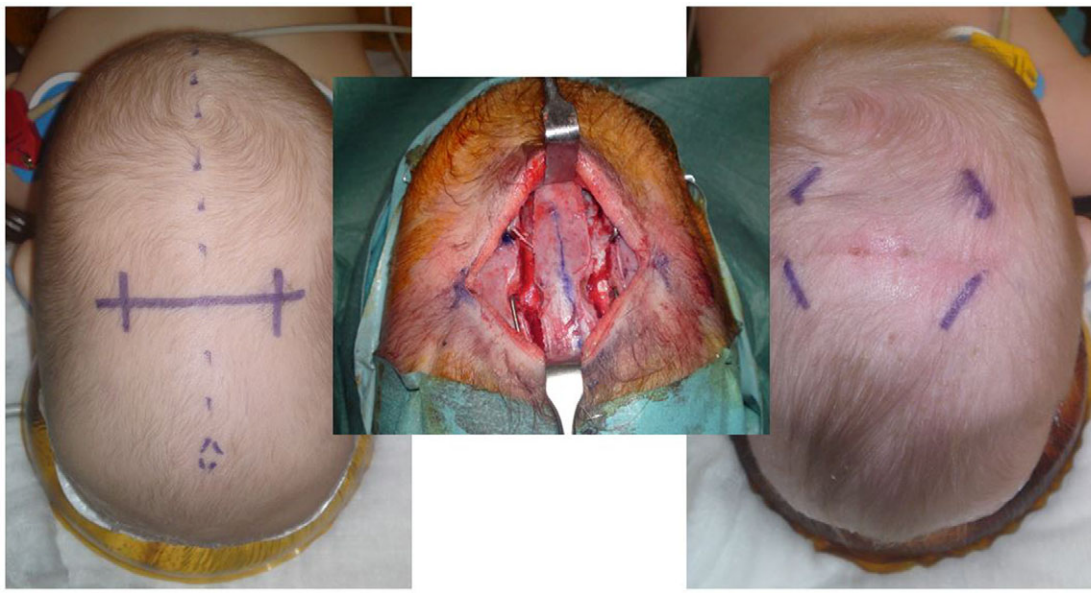

Figure 25.2 Operative technique for sagittal spring cranioplasty demonstrating 'sphinx' positioning and location of scalp incision (left), location of parasagittal osteotomies and springs (centre) and residual postoperative scar post spring removal (right).

Anaesthetic Considerations

- Patient position: modified prone with neck extension ('sphinx position')
- Surgical approach: small transverse incision
- Surgical duration: one hour
- Reinforced orotracheal tube with full neurosurgical strapping
- Peripheral venous cannula × 1
- Arterial line not essential

Spring-Assisted Posterior Vault Expansion

Spring-assisted posterior vault expansion is also commonly performed at the author's institution. Although it has some advantages over the traditional surgical technique, spring cranioplasty has not had quite the same impact as it has for sagittal synostosis. The procedure remains of similar duration to the traditional open technique, and although blood loss may be less, the risk of significant and rapid blood loss remains.

Anaesthetic Considerations

- Patient position: modified prone position with neck extension ('sphinx position')
- Surgical approach: bicoronal incision
- Surgical duration: three to four hours
- Reinforced orotracheal tube with full neurosurgical strapping
- Peripheral venous cannulae × 2
- Arterial line essential

Endoscopic Suturectomy for Craniosynostosis with Postoperative Helmeting (ESCH)

ESCH is increasingly being used in the treatment of single-suture synostosis in the United Kingdom. The procedure involves the early surgical removal of the affected cranial suture with postoperative guidance of remaining brain growth using helmet orthoses.

At the author's institution, a recent retrospective cohort study of 18 patients (mean age 4.6 months [range 2.5–7.8 months]) showed this to be a safe and well-tolerated technique for the treatment of metopic, unicoronal and sagittal synostosis with low morbidity (one surgical complication, suture abscess), transfusion requirement (2 of 18) and short hospital stay (median length one night).

Anaesthetic Considerations

- Patient position: supine/modified prone with neck extension ('sphinx position')
- Surgical approach: small transverse incision
- Surgical duration: one hour
- Reinforced orotracheal tube with full neurosurgical strapping
- Peripheral venous cannula × 1
- Arterial line not essential

Open Surgery

Fronto-orbital Remodelling/ Advancement

Fronto-orbital remodelling/advancement is currently the surgical procedure of choice for unicoronal, bicoronal and metopic synostosis. These forms of craniosynostosis affect the forehead and upper part of the eye sockets, producing a characteristic triangular deformity of the frontal bone with lateral forehead deficiency (trigonocephaly) in metopic synostosis, flattening of the ipsilateral forehead with bossing of the contralateral side (frontal plagiocephaly) in unilateral craniosynostosis and a skull that is short in the anterior–posterior dimension (brachycephaly), tall vertically (turricephaly) and wide in bicoronal synostosis. Fronto-orbital remodelling/advancement involves a bifrontal craniotomy with release of the affected suture(s) and simultaneous supraorbital and frontal reconstruction. The ideal timing of surgery varies between institutions and surgeon preference; advocates for early reconstruction (typically 9–12 months of age) claim that greater bony pliability results in a more favourable result, whilst those who prefer to delay surgery until 12–18 months of age do so to reduce the chance of recurrence.

Anaesthetic Considerations

- Patient position: supine, head up
- Surgical approach: bicoronal incision
- Surgical duration: three to four hours
- Pre-formed south-facing RAE orotracheal tube
- No eye covering (eye ointment only)
- Peripheral venous cannulae × 2
- Arterial line essential
- Intraoperative cell salvage may be useful for redo surgery

Total Calvarial Remodelling

Sagittal craniosynostosis is the most common form of synostosis and results in a skull shape that is typically long and narrow (scaphocephaly). The timing of reconstructive surgery is paramount to both the nature and extent of the surgical procedure performed. If surgery is performed within six to eight months of birth when the cranial bones are still very pliable, a spring-assisted sagittal cranioplasty is performed. This is the preferred technique at the author's institution, but a more extensive strip craniectomy involving a combination of wide sagittal craniectomy with plication of the vertically divided parietal bones is still performed in some centres.

Correction of sagittal synostosis in older children involves total calvarial remodelling. This is a more extensive procedure involving reconstruction of at least the anterior two thirds if not all the calvarium. The modified 'Melbourne' technique allows remodelling of both the anterior and posterior cranium. This involves an intraoperative change in patient position from the prone to supine position, during which meticulous care is required to avoid unintended dislodgement of the airway, arterial line or cannulae.

Anaesthetic Considerations

- Patient position
  Anterior two-thirds procedure: modified prone with neck extension ('sphinx position')
  Modified Melbourne procedure: combined modified prone and supine
- Surgical approach: bicoronal incision
- Surgical duration: total calvarial remodelling three to four hours; modified 'Melbourne' technique five to six hours
- Reinforced orotracheal tube with full neurosurgical strapping
- Peripheral venous cannulae × 2
- Arterial line essential
- Intraoperative cell salvage may be useful for redo total calvarial remodelling

Posterior Calvarial Vault Expansion/ Remodelling

Posterior vault expansion/remodelling is performed to treat raised ICP, which can be related to craniosynostosis (multi-sutural or unilateral lambdoid synostosis), abnormal venous drainage, OSA or hydrocephalus. Posterior vault expansion can be performed as an open procedure involving the removal and reshaping of the posterior skull with bone grafting, or more commonly by spring cranioplasty. As posterior vault expansion is used to treat raised ICP, it may be performed at any age, although this typically occurs at 6 months of age and usually before 10 years of age. There is a risk of significant bleeding with this procedure whether it is performed using an open or minimally invasive technique.

Anaesthetic Considerations

- Patient position: modified prone with neck extension ('sphinx position')
- Surgical approach: bicoronal incision
- Surgical duration: three to four hours
- Reinforced orotracheal tube with full neurosurgical strapping
- Peripheral venous cannulae × 2
- Arterial line essential
- Intraoperative cell salvage may be useful for redo surgery

Monobloc Frontofacial Advancement and Facial Bipartition with or without Distraction Osteogenesis

Monobloc frontofacial advancement and bipartition have an important role to play in the management of syndromic synostosis. They can be used to reverse the associated facial deformity seen in children with Apert, Crouzon and Pfeiffer syndromes and to treat functional problems such as raised ICP, severe exorbitism with or without globe subluxation and upper airway obstruction. Monobloc frontofacial advancement addresses all these functional issues in a single procedure. The operation combines transcranial frontal advance and extracranial Le Fort III midfacial advance with the frontal bones and maxilla being brought forward in one piece either immediately at surgery or more slowly by distraction using a RED frame (Figure 25.3). The RED frame comprises a halo device which is attached to the mobilised facial skeleton through a combination of cranial pins, rods and wires. Postoperative distraction (~1.5 mm/day) is achieved by progressively shortening the wires. The distractor is removed under general anaesthesia once the midface has advanced the required distance and following a consolidation period of six weeks.

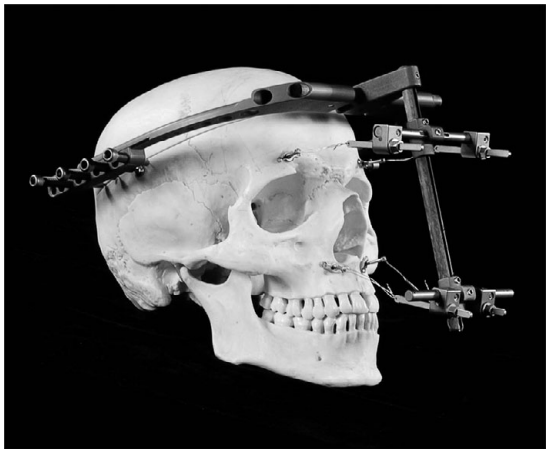

Figure 25.3 A rigid external distractor (RED) in situ.

A facial bipartition occurs naturally as part of a midline facial cleft. A midline split can also be created surgically as an extension of a monobloc frontofacial advancement to correct frontonasal dysplasia, facial dysostoses and orbital hypertelorism (horizontal dystopia). Following mobilisation of the maxilla and orbits, removal of a midline segment of bone and extension of the central osteotomy down to the incisors allows rotation of the two hemifacial segments together, thereby narrowing the interorbital distance and expanding the maxilla. It also corrects any down-going slant to the eyes, which is a feature of Apert syndrome.

Both procedures may be associated with high morbidity, especially in young infants and children. Timing of surgery is crucial, and the procedure should ideally be delayed until skeletal maturity has been achieved at approximately 12 years of age. Indications for early frontofacial advancement include severe upper airway obstruction, ocular exposure or subluxation and raised ICP.

Orbital Box Osteotomy

Orbital box osteotomy is used in the treatment of hypertelorism and vertical orbital dystopia, particularly when lateral movement of a single orbit is required. It involves making a 360° osteotomy around one or both orbits, thereby allowing them to be moved medially either upwards or downwards. As the osteotomy extends between the orbital floor and the roots of the permanent dentition, this procedure should only be performed once puberty has been reached.

Anaesthetic Considerations

- Patient position: supine head up.
- Surgical approach: combined intraoral and transcranial (bicoronal) incisions.
- Surgical duration: six to eight hours.
- Reinforced orotracheal tube with circummandibular wiring. In patients with a tracheostomy in situ, a reinforced tracheal tube is placed via the tracheostome and secured to the anterior chest wall.
- A throat pack is essential to protect the airway from blood and bony fragments and assist with securing the tracheal tube.
- Eye ointment and temporary tarsorrhaphies are used to protect the eyes intraoperatively (replaced with Frost sutures for 48 hours postoperatively).
- At least two large bore peripheral venous cannulae.
- Femoral central venous line is recommended, especially in the younger child.
- Arterial line is essential.
- Urinary catheter.
- Intraoperative cell salvage.
- Care should be taken to ensure that all pressure points are padded, and these should be rechecked during the procedure.
- Anti-embolism compression stockings (TED™) for older teenagers with mechanical deep venous thrombosis (DVT) prophylaxis system (e.g. Flowtron).
- Bilateral nasopharyngeal airways and a nasogastric tube are inserted at the end of surgery.
- It is usual practice at the author's institution to extubate all patients at the end of surgery if surgery has been uneventful, the patient is haemodynamically stable and there is no respiratory compromise.
- As the RED frame is bulky, it may impede access to the airway, making reintubation very difficult. If there are any postoperative airway concerns, the halo may be placed with delayed fitting of the suspension frame.

Removal of the RED Frame

In the older child, removal of a rigid external distractor is a relatively simple procedure involving disconnection of the halo device followed by

removal of the cranial pins. The situation is very different in infants, owing to the insertion of titanium mesh sheets to prevent penetration of the skull by the cranial pins. This necessitates removal of the distractor and sheets via a bicoronal incision.

The presence of a RED frame may make airway management difficult, so it is important to ensure that the necessary tools and expertise to remove the frame are readily available prior to induction of anaesthesia. In cooperative children, the suspension frame may be removed prior to induction of anaesthesia. If this is not feasible, the face mask may have to be inverted and then changed for a laryngeal mask airway once the patient is anaesthetised, as the frame's wires and bars make conventional placement of a face mask impossible. Laryngoscopy and intubation may also be difficult if not impossible, and for procedures other than removal of the frame where intubation is deemed necessary, it is advisable that the surgeon remove the wires and bars.

Key Points

- Craniosynostosis results from the premature closure of one or more cranial sutures and can occur in isolation or as part of an associated syndrome.
- Preoperative evaluation should focus on any comorbidities and the possibility of difficult airway management. Children who have undergone previous corrective surgery may be difficult to intubate, despite the lack of any history of difficult laryngoscopy.
- Successful perioperative management is dependent on understanding the proposed surgical procedure and the anticipation and prompt treatment of the many potential complications that may arise.
- Blood loss and major transfusion remain the greatest risk to infants and children undergoing complex major craniofacial surgery. Blood loss, which may be dramatic or insidious, and dilutional coagulopathy should be managed proactively.
- Intraoperative cell salvage is useful in reducing the need for allogeneic transfusion, particularly in prolonged major or complex major surgical cases.
- Most patients can be extubated at the end of surgery, with postoperative care being continued on the neurosurgical high-dependency unit (HDU) with continued invasive monitoring.

Further Reading

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