# Modern Management of Craniosynostosis

# 79

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## 79.1 Introduction

The different presentations of craniosynostosis form the majority of the workload of craniofacial services, and it is important to note that the surgical interventions only form a small part of the overall management of this vulnerable group of patients and their families.

### 79.1.1 Definition

Craniosynostosis is defined as the premature fusion of one or more of the cranial sutures. This premature fusion can either be an isolated disorder (e.g. sagittal synostosis) or form part of a syndrome (e.g. Apert syndrome). In many cases the resultant head shape is typical of the involved suture such that the Greek/Latin descriptive terms are used synonymously with the description of the affected suture. For example scaphocephaly (boat-shaped head) is used to describe sagittal synostosis; however this approach can lead to confusion as can be seen in the use of the term plagiocephaly (flat head) which may refer to a unicoronal synostosis but also can be used in unilateral lambdoid synostosis, skull base torsion, the deformity resulting from torticollis and positional skull deformities. For this reason it is probably best to avoid the Latin/Greek descriptive terms and identify the affected suture(s) by name.

The majority of craniosynostoses are primary in nature and congenital; however a small proportion are termed secondary—caused by another pathology, usually resulting in reduced brain growth as seen in microcephaly or post-shunting.

### 79.1.2 Aetiology

#### 79.1.2.1 Primary Craniosynostosis

In the majority of cases, no cause for the synostosis is identified, but in an increasing proportion (currently about 25%), a mutation is identified. A significant proportion of these mutations are related to six genes FGFR2, FGFR3, TWIST1, EFNB1, TCF12 and ERF, however there is an increasing frequency of additional mutations being identified by more complex genetic analysis.

The incidence of identifying a mutation is much higher in syndromic craniosynostosis cases (69%), but the rate of mutation identification in the non-syndromic cases is increasing (currently 5%) particularly in the bicoronal, multisuture and unicoronal (in decreasing frequency) [1].

These genetic advances have assisted in the diagnosis of these conditions and influence the genetic counselling but have yet to impact on the management of the resultant problems related to the premature suture closure. As the mechanisms of sutural control and homeostasis are elucidated, novel therapies may be introduced.

#### 79.1.2.2 Secondary Craniosynostosis

Microcephaly and babies who have been shunted as well as other systemic conditions such as sickle cell disease, thalassaemia and rickets can lead to synostosis usually affecting all the sutures. In cases of microcephaly, it is important to differentiate between a primary pansynostosis resulting in a small head often with raised intracranial pressure from a secondary synostosis with normal intracranial pressure [2].

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### 79.1.3 Epidemiology

The incidence of craniosynostosis ranges from 1 in 1400 to 1 in 2100 live births worldwide with a slight male preponderance.

The different types of craniosynostosis occur in the following proportions:
- Sagittal 45%.
- Metopic 20%.
- Bicoronal and unicoronal 15%.
- Multiple suture 5%.
- Lambdoid <2% [3].

### 79.1.4 Classification

The classification of craniosynostosis has, in recent years, changed; the previously utilised terminology of simple and complex, referring to single-suture and multiple suture abnormalities, respectively, has been abandoned. It has little utility and is in fact misleading—the deformity as a result of a unicoronal synostosis is extremely complex and as more is understood about the genetic basis the more complex the condition becomes.

In terms of utility, the craniosynostosis is best classified by the suture(s) involved and whether this is associated with a syndrome (i.e. other abnormalities). See Table 79.1 and Illustrations 79.1, 79.2, 79.3, 79.4, 79.5, 79.6 and 79.7. The condition can be further defined by the genetic abnormalities. In general the syndromic craniosynostosis cases tend to be more severe and have a greater incidence of complicating features.

## 79.2 Management

The care of babies and children with craniosynostosis is best delivered in a centre with access to a full multidisciplinary team. This team involves the core specialities of paediatric neurosurgery and craniofacial surgery (either OMFS or plastics based) as well as support from paediatricians, neonatologists, geneticists, respiratory specialists, ophthalmologists and ENT surgeons. The input from specialist nurses and psychologists is vital [4].

**Table 79.1** Overview of craniosynostoses

| Suture affected | Description of shape | Diagram of shape | Description |
|---|---|---|---|
| Metopic | Trigonocephaly | Illustration 79.1a, b | Pointed, protuberant, prow-shaped forehead<br>Reduced intercanthal distance<br>Bitemporal narrowing |
| Unicoronal | Anterior plagiocephaly | Illustration 79.2a, b | Flattened forehead on affected side with raised eyebrow and shallower orbit<br>Failure of forward growth of the hemiforehead with the ipsilateral ear also posterior |
| Bicoronal | Brachycephaly | Illustration 79.3a, b | Shortened AP—Coronal dimension<br>Flattening of whole forehead<br>Flattened supraorbital rims |
| Sagittal | Scaphocephaly | Illustration 79.4a, b | Forehead bossing<br>Occipital bulleting<br>Bitemporal narrowing<br>Increased AP—Coronal dimension |
| Lambdoid | Posterior plagiocephaly | Illustration 79.5a, b | Bossing of mastoid on affected side<br>Contralateral parietal bossing<br>Ipsilateral ear posteriorised |
| Pansynostosis | Turricephaly or Oxycephaly | Illustration 79.6a, b | Restricted growth at all or multiple sutures—Upward growth in the region of the Fontanelle<br>Small circumference<br>'Cone'- or 'turret'-shaped head |
| Positional | Plagiocephaly | Illustration 79.7a, b | Parallelogramming of the whole head<br>Ears move with the segments<br>No ridging of any sutures |

**Illustration 79.1** (**a**, **b**)
Metopic suture affected in trigonocephaly

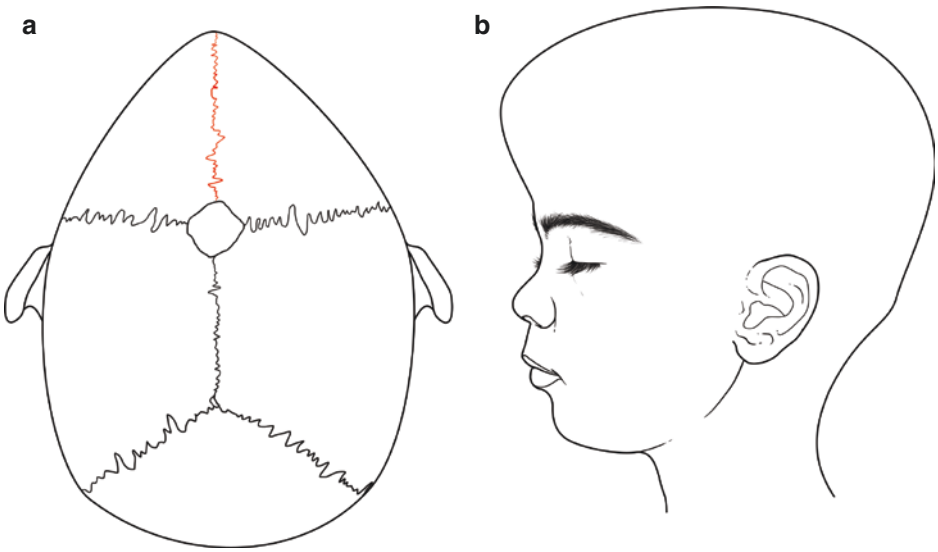





**Illustration 79.2** (**a**, **b**)
Unicoronal suture affected in anterior plagiocephaly

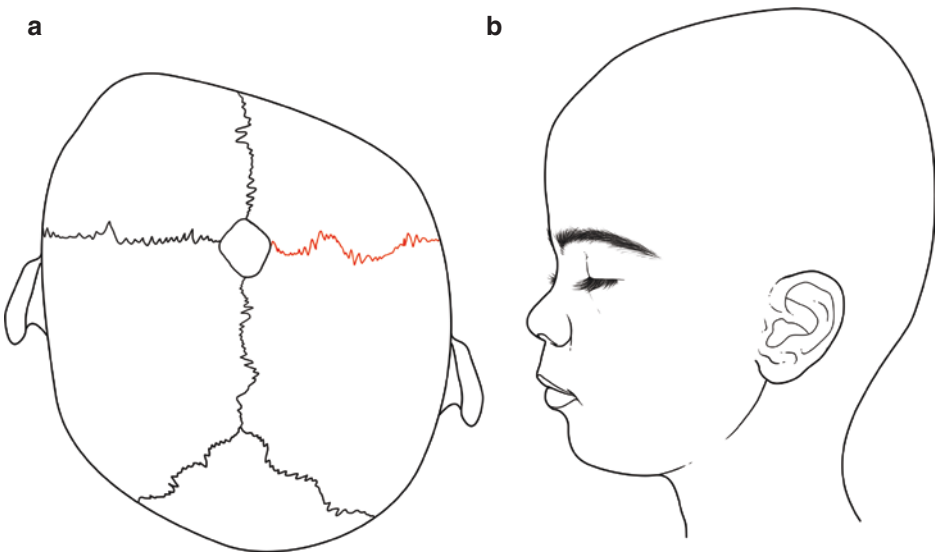





**Illustration 79.3** (**a**, **b**)
Bicoronal suture affected in brachycephaly

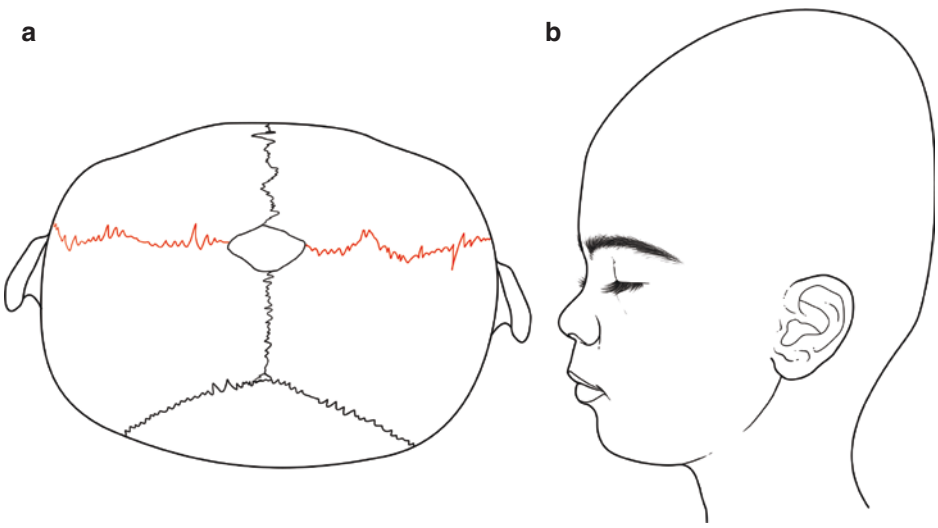


**Illustration 79.4** (**a**, **b**)
Sagittal suture affected in scaphocephaly

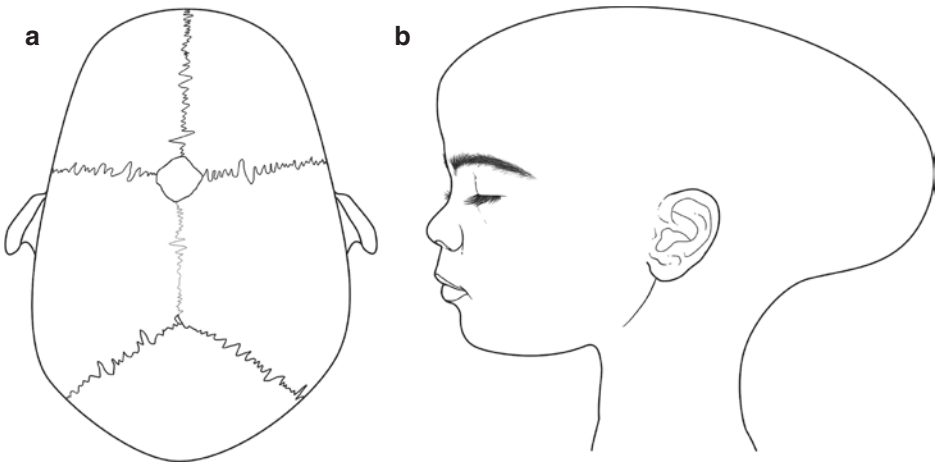





**Illustration 79.5** (**a**, **b**)
Lambdoid suture affected in posterior plagiocephaly

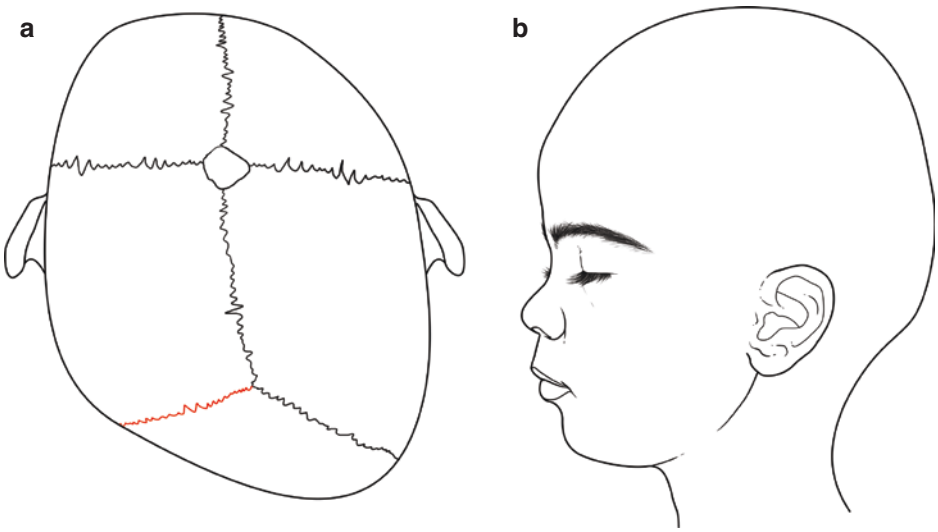





**Illustration 79.6** (**a**, **b**)
Pansynostosis causing turricephaly or oxycephaly

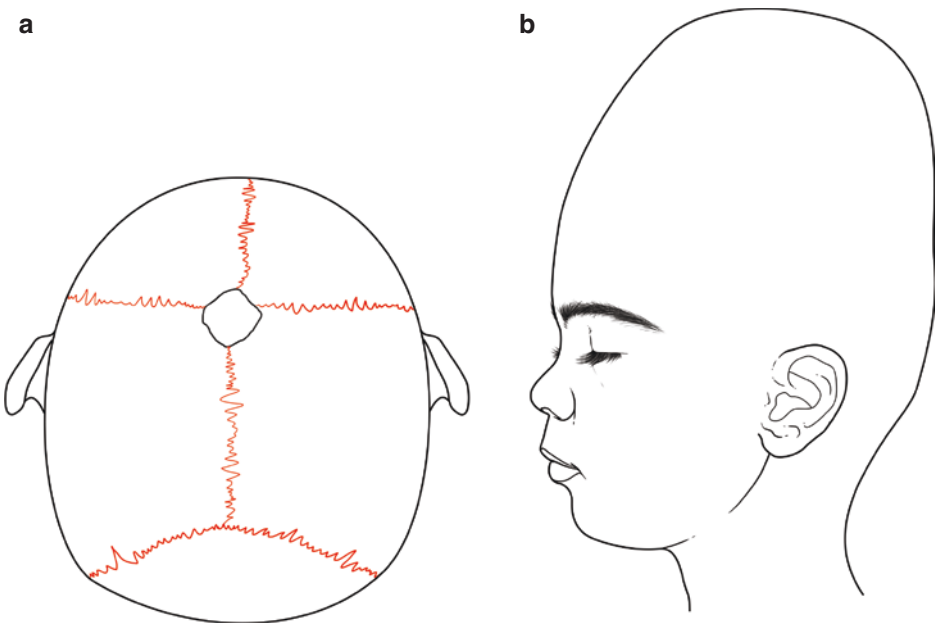


**Illustration 79.7** (**a**, **b**) Positional plagiocephaly

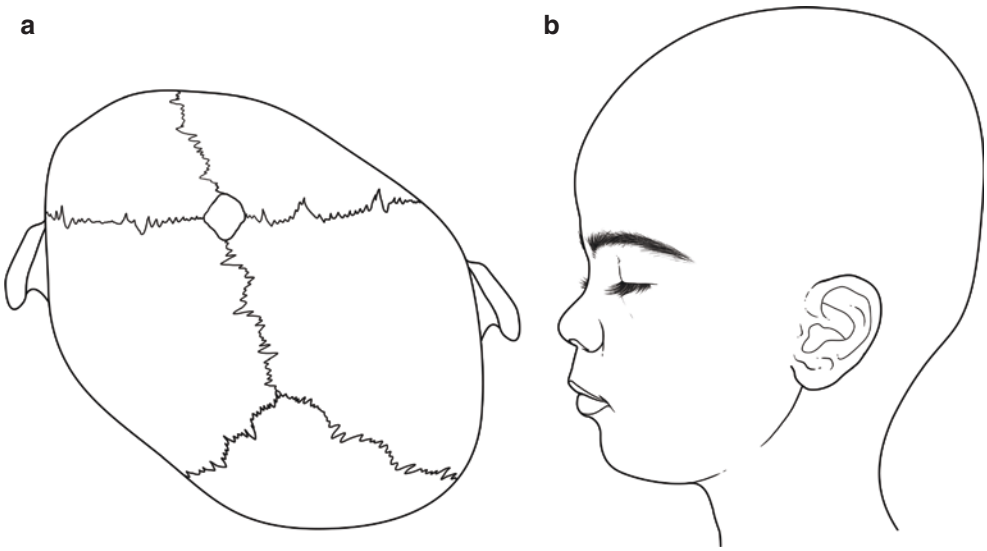





**Table 79.2** Treatment options in infancy for complex synostosis problems

| Clinical problem | Emergency/urgent treatment options | Definitive treatment options |
|---|---|---|
| Upper airway obstruction 2° to midface retrusion | Nasopharyngeal airway<br>Intubation, $O_2$ support, CPAP, tracheostomy | Transcranial monobloc advancement (± distraction), CPAP, tracheostomy,<br>Subcranial Le fort III advancement |
| Corneal exposure | Intensive eye care, tarsorrhaphy | Fronto-orbital advancement, Monobloc advancement |
| Hydrocephalus | External ventricular drain, ventriculo-peritoneal shunt, third ventriculostomy (in selected cases) | Ventriculo-peritoneal shunt |
| Elevated ICP | Medical treatment with acetazolamide, steroids | Skull vault expansion, fronto-orbital advancement and remodelling (FOAR), monobloc advancement ± implanted telemetric device |

### 79.2.1 Prenatal Diagnosis

The use of high-definition prenatal ultrasound has increased the early diagnosis of both hydrocephalus and craniosynostosis [5]. This facilitates the preparation of parents for the birth of a child who is likely to have additional medical, surgical and emotional needs. In these circumstances parents getting to know the craniofacial team and being better informed of the potential problems often makes supporting them much easier.

### 79.2.2 Perinatal Care (Table 7.2)

In the perinatal period, care is directed at optimising the airway, ensuring the child is well oxygenated, preventing corneal damage (ocular protection) and confirming the diagnosis. In the majority of single-suture abnormalities, there are often no issues that prompt early intervention; however in the complex syndromic cases, this is not the case. Airway problems due to severe midface retrusion; breathing disturbance due to central problems, often due to posterior fossa crowding and Chiari malformation; corneal exposure secondary to midface and brow retrusion; and associated non-craniofacial anomalies prompt more urgent interventions. The necessity for such interventions is greatest in the syndromic conditions such as Apert, Pfeiffer, Crouzon and Carpenter syndromes. In cases where there is significant midface retrusion, there may also be feeding difficulties [6].

In the severe cases, initial care is supportive. The airway should be assessed and intervention initiated if necessary. It is important to assess the oxygen saturation and the work of breathing and correlate these findings with formal blood gas measurements. This assessment can be augmented with a formal sleep study, and on occasion simultaneous measurement of intracranial pressure may be helpful. Airway support can range from positional nursing, the use of a nasopharyngeal airway, intubation and in some cases tracheostomy. As part of this assessment, when the airway is an issue of concern, consideration should be given to performing a CT scan, and this will be useful to look at cranial anatomy but also the nasal anatomy, particularly to exclude choanal atresia. In assessing the airway, babies often manage satisfactorily but may suddenly decompensate with a minor upper respiratory tract infection or whilst feeding [6].

Surgical intervention in this period is only undertaken when the airway is such that no simple measures are able to overcome the symptoms; often a period of intubation is helpful to allow for a more thorough assessment rather than rushing to either tracheostomy or a monobloc fronto-facial advance.
During this period parents and family members have to come to terms with the often unexpected consequences of having a child with additional needs and medical interventions. Early and ongoing support needs to be delivered; this can and should be through multiple avenues. Many such children are born in institutions with little or no experience of the condition(s), and this professional unfamiliarity often adds to parental anxiety. For these reasons early contact with a dedicated craniofacial team is invaluable. Specialist nurses, psychologists and support groups can often ease the stress of this difficult time [7].

### 79.2.3 Care in Infancy

The management of craniosynostosis in infancy is primarily aimed at addressing the primary consequences of the premature suture closure as well as dealing with the associated problems.

The management of the synostosis has, over the years, evolved considerably and in some respect completed a complete circle. The treatment is aimed at ensuring cranial volume is satisfactory to ensure that there is no elevation of ICP (in the absence of hydrocephalus) as well as attempting to normalise the head shape. The normalisation of head shape is done to maximise the chances of a normal head shape at the completion of growth. Initial treatment introduced in the second half of the last century involved a procedure known as suturectomy [8]. The principle of this was to excise the pathological, prematurely fused suture with the idea that removal of the pathology would allow the skull to continue to grow into a normal shape possibly aided by the continued growth of the brain. This was found to be partially effective especially when done before the age of 18 months [9]. It was however noted that in many cases re-fusion of the suture occurred and a number of surgeons began wrapping the bone edges on either side of the resected suture with silicone edging in an attempt to minimise the chances of re-fusion. Unfortunately this approach did not prevent re-fusion, and this often occurred on the dural side of the silicone edging. In pansynostosis cases the sutures were excised and the remainder of the skull was morselised [9]. These simple techniques have been superseded with more complex reconstructive procedures involving suturectomy and remodelling. (See case series examples (Figs. 79.2, 79.3, 79.4 and 79.5). When the coronal sutures are involved, either unilaterally or bilaterally, and/or the metopic sutures, the mainstay of treatment is a fronto-orbital advancement, with remodelling of the affected bones. When the sagittal suture is involved, the affected suture is excised and the remaining skull remodelled. The extent of remodelling varies from case to case but can involve addressing the frontal bossing, bitemporal narrowing and the occipital bullet [4]. These procedures involve extensive exposure and blood loss with their attendant risks, and for these reasons there has been a move to attempt to correct these abnormalities with less invasive procedures. These less invasive approaches utilise endoscopic suturectomy coupled with active postoperative helmet therapy to mould and harness the ongoing brain and hence skull growth [10]; another approach has used a minimal access suturectomy and specially designed springs to actively separate the affected suture edges [11]. A further technique, particularly in bicoronal or pansynostosis cases, is to utilise distraction osteogenesis; a craniotomy of the occipital bone is performed, and two or three distraction devices are applied; these are then activated over a period of 2–3 weeks, expanding the skull volume and in many cases improving the posterior fossa crowding seen in such cases. For reasons not fully understood, this technique often leads to a significant improvement in the contour of the frontal bones [12].

In terms of timing of surgery, there is considerable variation between different centres. In general elective (not motivated by functional concerns), suturectomy and skull remodelling procedures are performed between the ages of 5 months and 2½ years. The younger the patient, the greater the contribution of normal brain growth in normalising head shape; however the drawback of earlier surgery is the fragility of the skull bones and most importantly the risks of complications—particularly excessive blood loss [13].

During this period patients are generally followed up to monitor for signs of raised intracranial pressure, obstructive sleep apnoea (OSA) and evidence of a significant change in the rate of head growth as indicated by head circumference. Discrepancies between the plots on the growth charts (Fig. 79.1a, b) for length, weight and head circumference as well as plots crossing the centile lines should prompt further investigation.

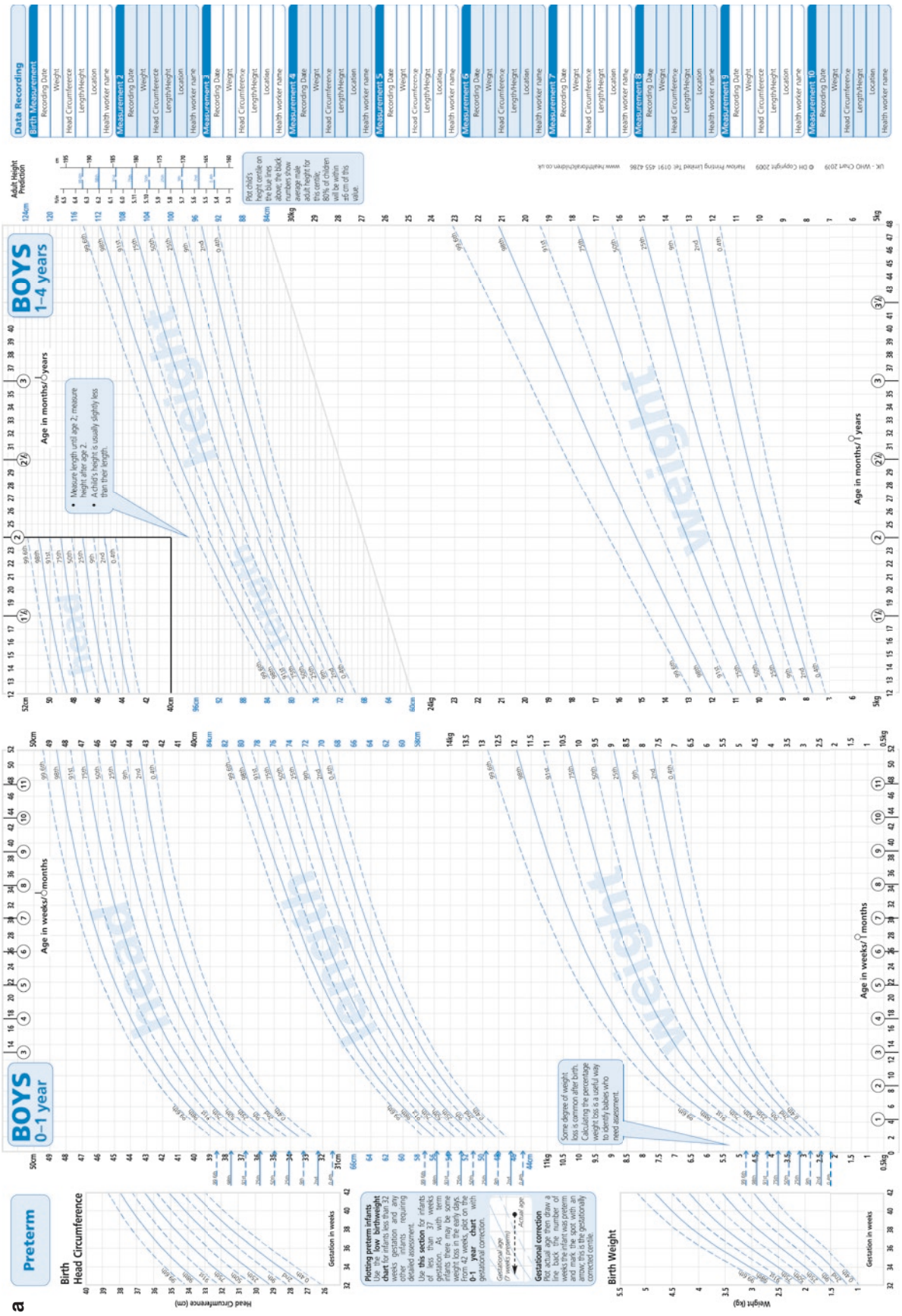



**Fig. 79.1** (**a**) (boys), (**b**) (girls) WHO UK growth the chart (**a**) and (**b**) is available Online at https://www.rcpch.ac.uk/resources/uk-who-growth-charts-0-4-years

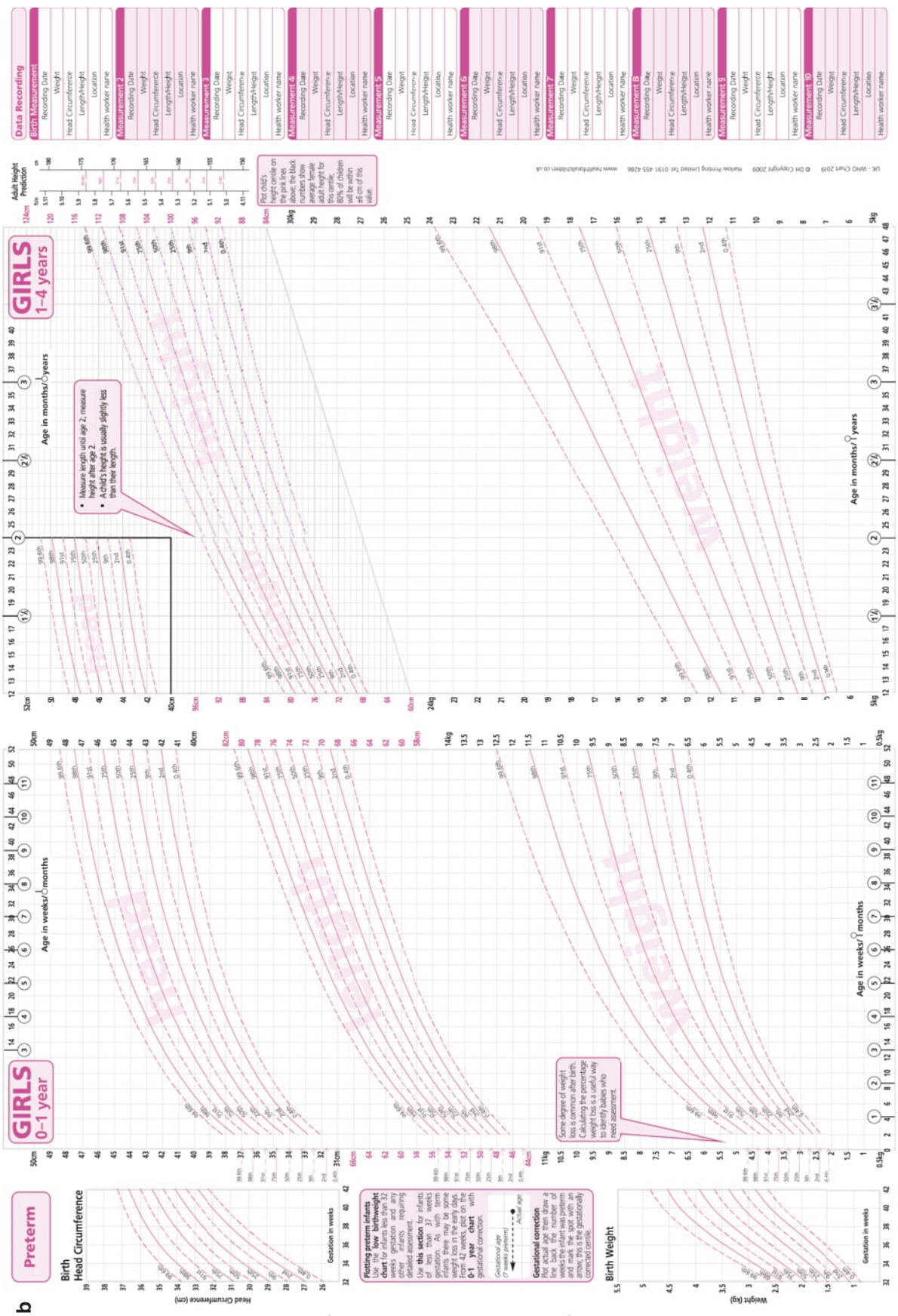



**Fig. 79.1** (continued)

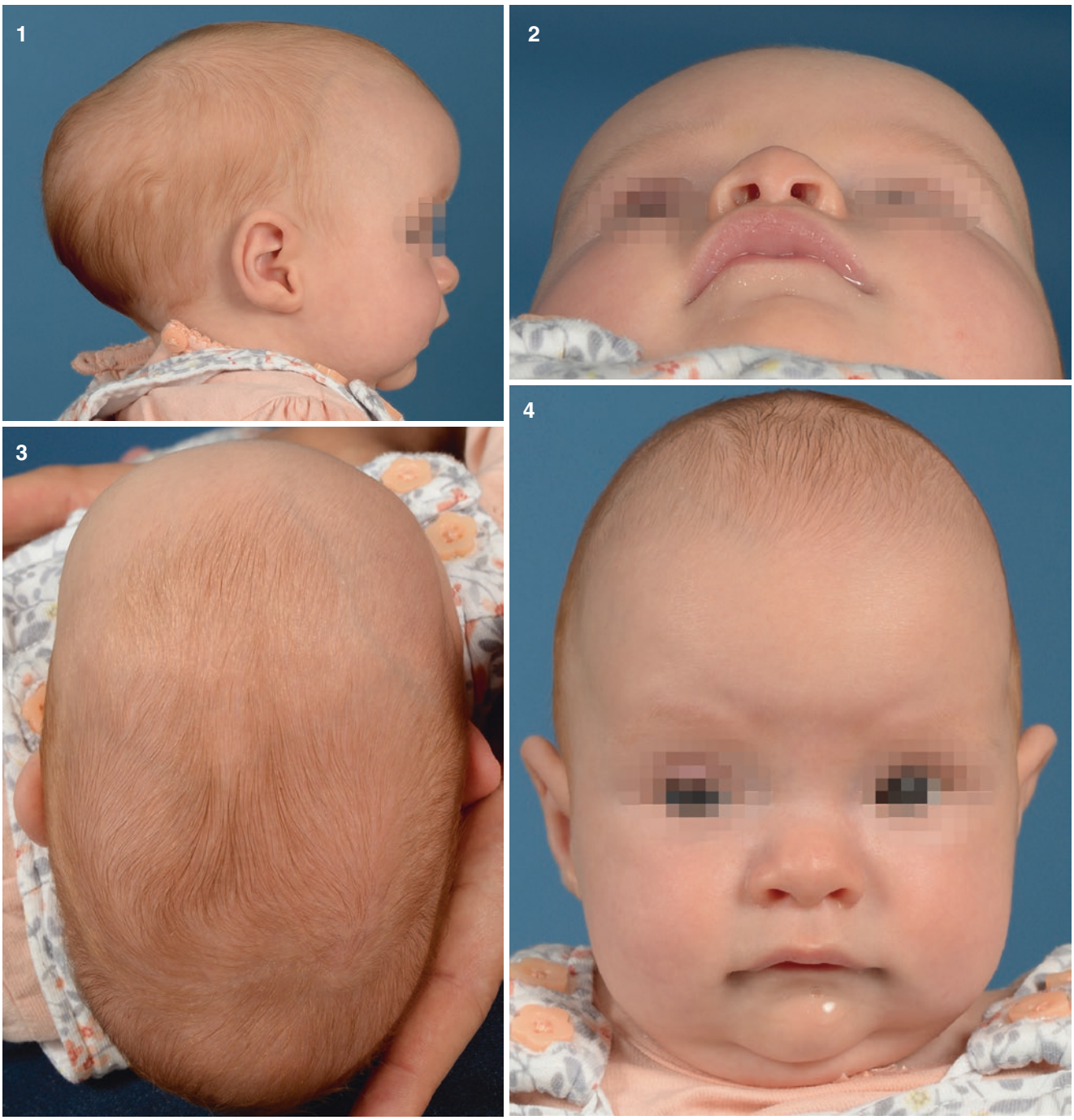
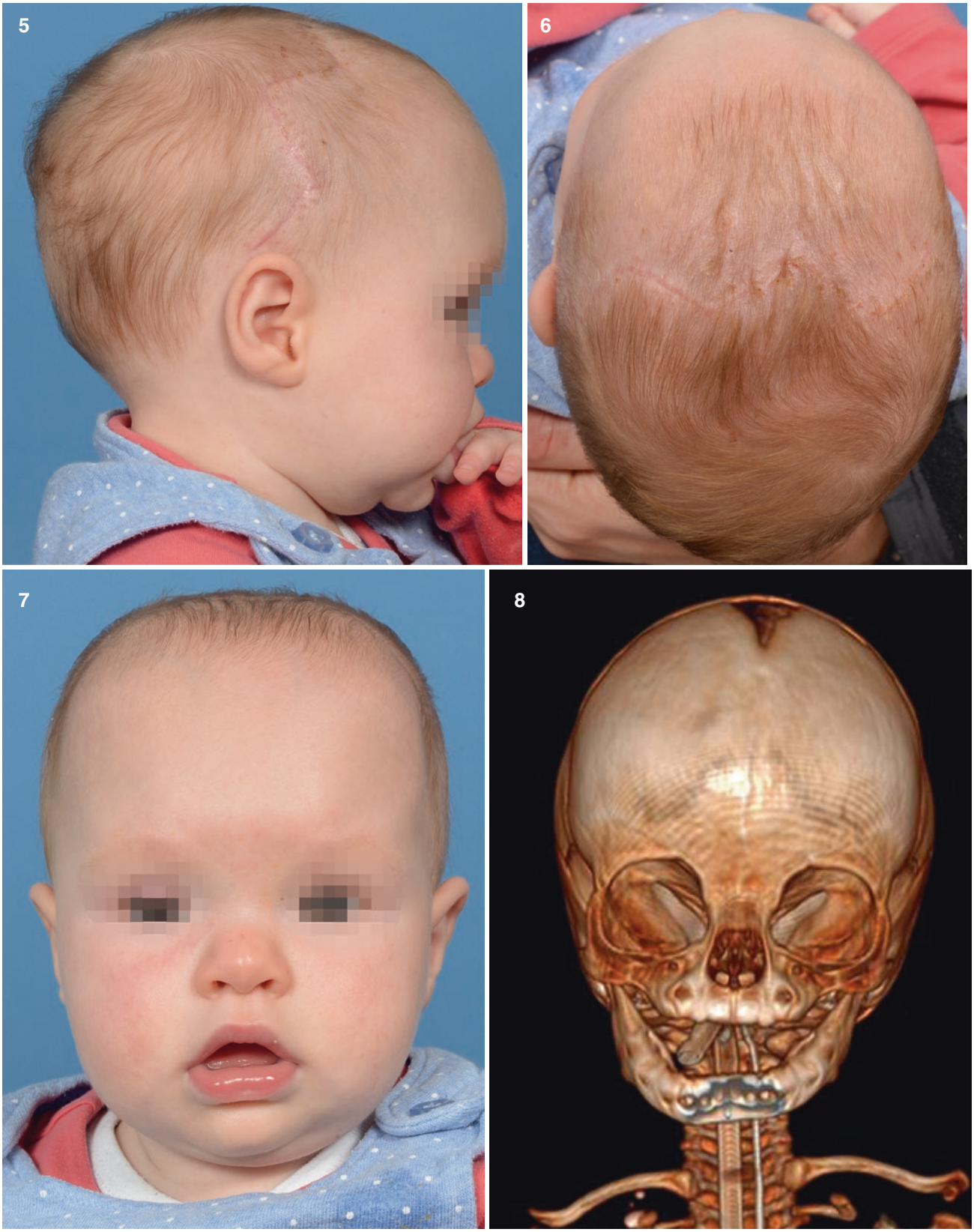



**Fig. 79.2** (2.1–2.9) Case series of sagittal synostosis. (2.1–2.4) Preoperative facial views of patient with sagittal synostosis aged 13 weeks. Note elongated A-P dimension and bitemporal narrowing with prominent forehead and occiput. (2.5–2.7) Postoperative facial views of patient with sagittal synostosis after full calvarial remodelling. (2.8 and 2.9) Preoperative CT scan of the same patient with sagittal synostosis

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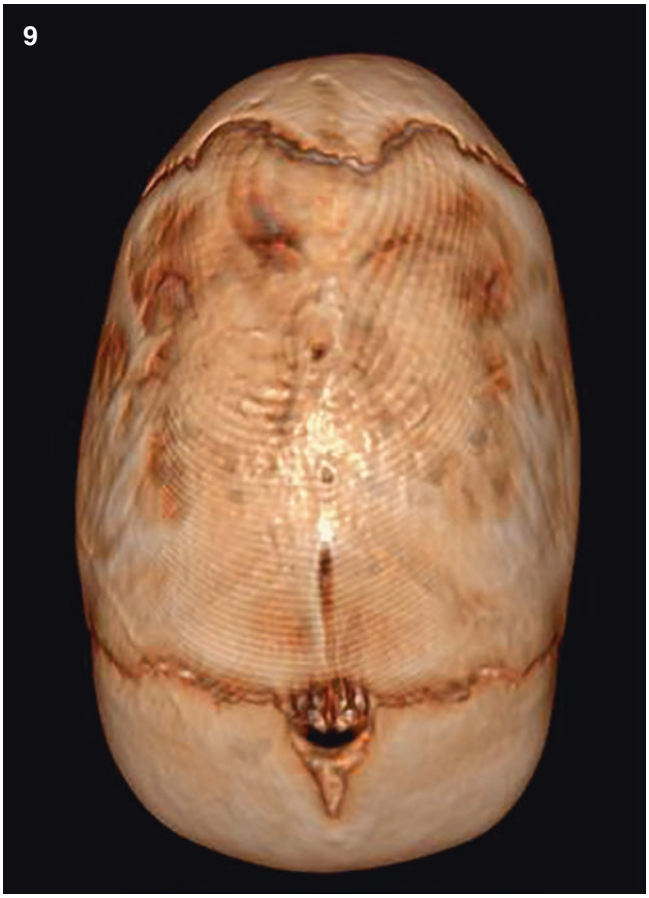





**Fig. 79.2** (continued)

Elevated ICP manifests as distressed behaviour, inconsolable crying and headbanging with symptoms being worse at night. When symptoms are combined with papilloedema, urgent intervention is indicated. The absence of papilloedema does not exclude raised ICP, but its presence is of significance. In cases of doubt, formal ICP monitoring can be undertaken. In established cases the use of an indwelling telemetric ICP measurement device may be helpful [14].

Formal ophthalmological assessment should be undertaken and appropriate intervention initiated. Regular assessments are necessary to identify deterioration, particularly to identify corneal exposure and avoid the development of amblyopia.

Routine computerised tomography (CT) cross-sectional imaging is not indicated, and the diagnosis can, in the vast majority of cases, be made on clinical findings alone. In cases where the diagnosis is in doubt or there is the suspicion that there may be posterior fossa crowding, Chiari malformation or craniocervical abnormalities, a CT is indicated. Magnetic resonance imaging (MRI) is indicated to investigate the brain for structural abnormalities. The objective with this very conservative approach to imaging is to minimise the ionising radiation exposure, which has been demonstrated to adversely affect brain development and damage the developing eye [15].

Obstructive sleep apnoea can manifest as noisy snoring, characteristically crescendoing to a maximum followed by a period of silence, representing the apnoeic period with a cyclical restart, or a failure to thrive coupled with daytime tiredness. If OSA is suspected, an initial overnight pulse oximetry study can be undertaken, and if this proves to be suggestive of OSA, a full polysomnogram sleep study should be performed. This can be combined with formal ICP monitoring should there be any concerns about raised ICP. The active management of established OSA requires intervention from the multidisciplinary team, and the respiratory/ENT specialist input is paramount. It is important to establish the causes of sleep disorder and the level(s) of airway obstruction so that the most appropriate intervention(s) can be employed. Further investigations might involve microlaryngoscopy to exclude tracheal abnormalities. Imaging with CT is useful, and interventions should be stepwise with consideration being given to tonsillectomy and adenoidectomy [16].

In the majority of cases of non-syndromic single-suture abnormalities, a single intervention for the surgical correction of the head shape is the norm [4].Whilst there is considerable variation between different units regarding

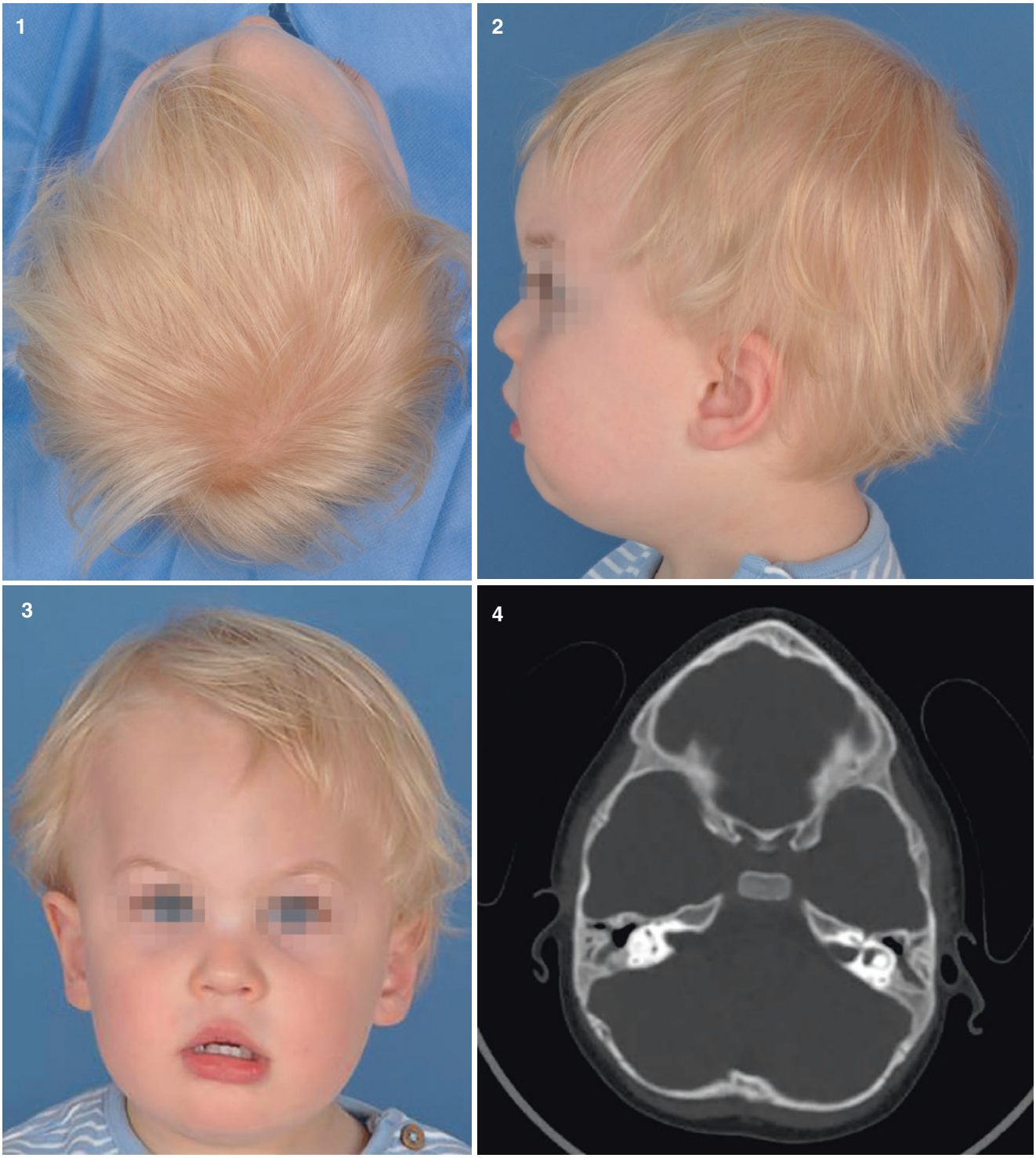
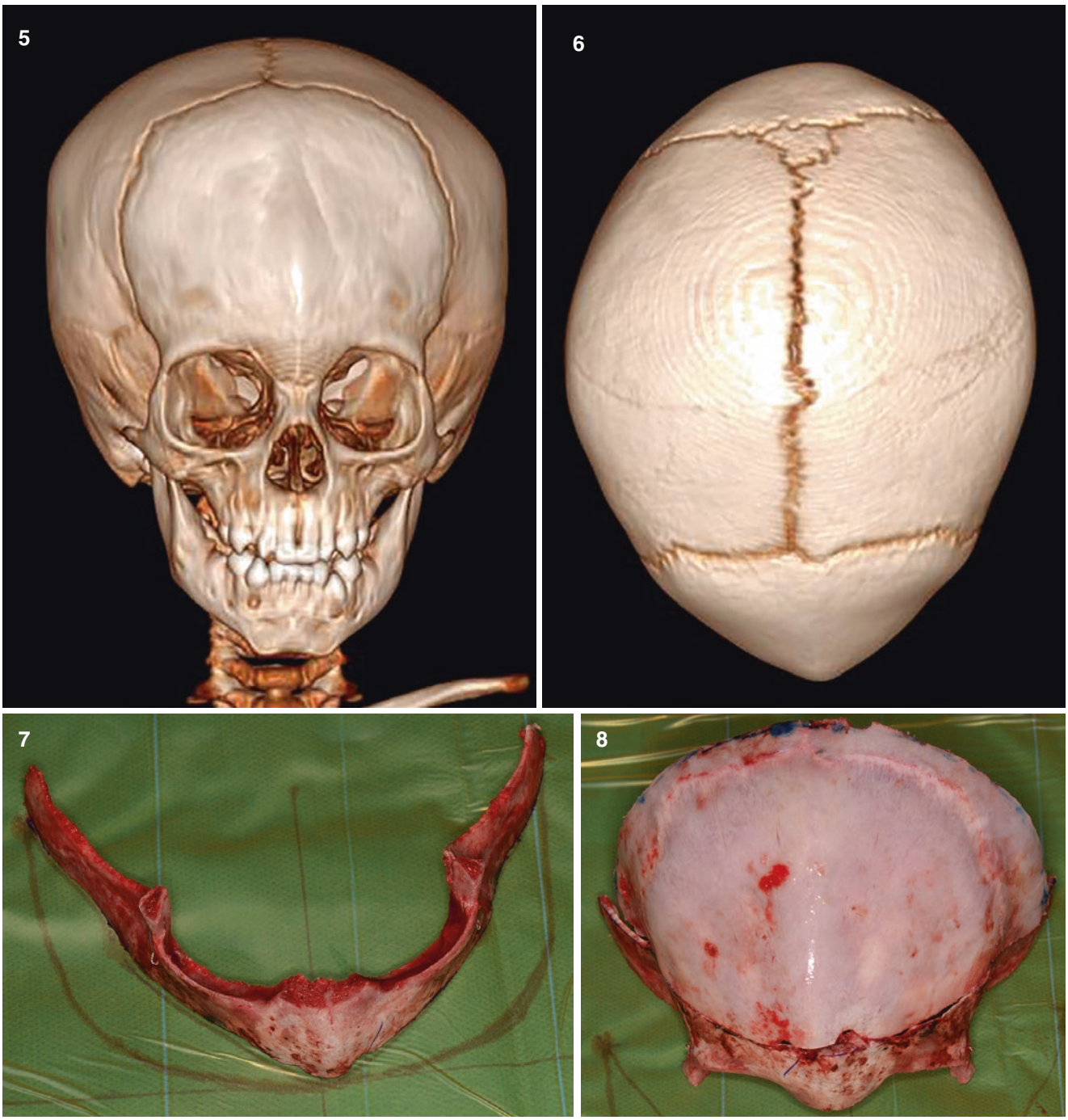



**Fig. 79.3** (3.1–3.17) Case series of metopic synostosis. (3.1–3.3) Preoperative facial views of patient with metopic synostosis, age 23 months (late presentation), note pointed brow, triangular-shaped skull and hypertelorism. (3.4–3.6) Preoperative CT scan of the same patient with metopic synostosis—note it is not our usual practice to obtain preoperative CT in metopic synostosis; however this was taken due to the subtle (and late) presentation. (3.7–3.14) Intraoperative views of fronto-orbital remodelling. (3.7) Frontal bar after removal prior to remodelling. (3.8) Frontal bar and fontal bones after removal prior to remodelling. (3.9, 3.10) Frontal bar after remodelling with midline graft to correct hypotelorism. (3.11) Frontal bone osteotomised ready for remodelling. (3.12–3.14) Final views after fixation—fronto-orbital remodelling or anterior two-thirds remodelling. Note use of absorbable plates in load−/tension-bearing areas and absorbable sutures or stainless steel wires in less critical regions. Bone dust and bone dust putty (fibrin sealant mixed with bone dust) are used in resultant bone defects to encourage ossification. (3.15–3.17) Postoperative facial views of patient with metopic synostosis

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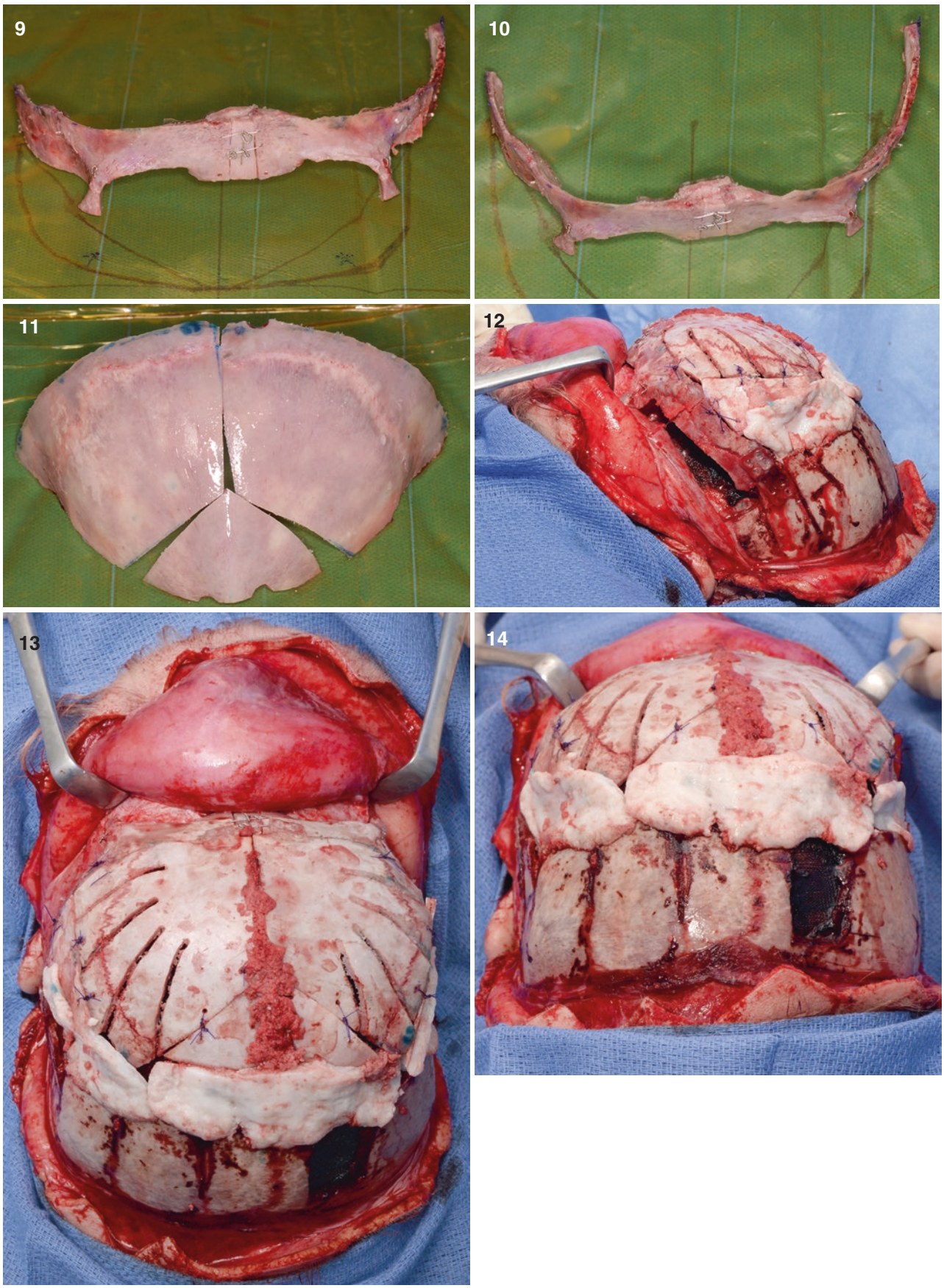


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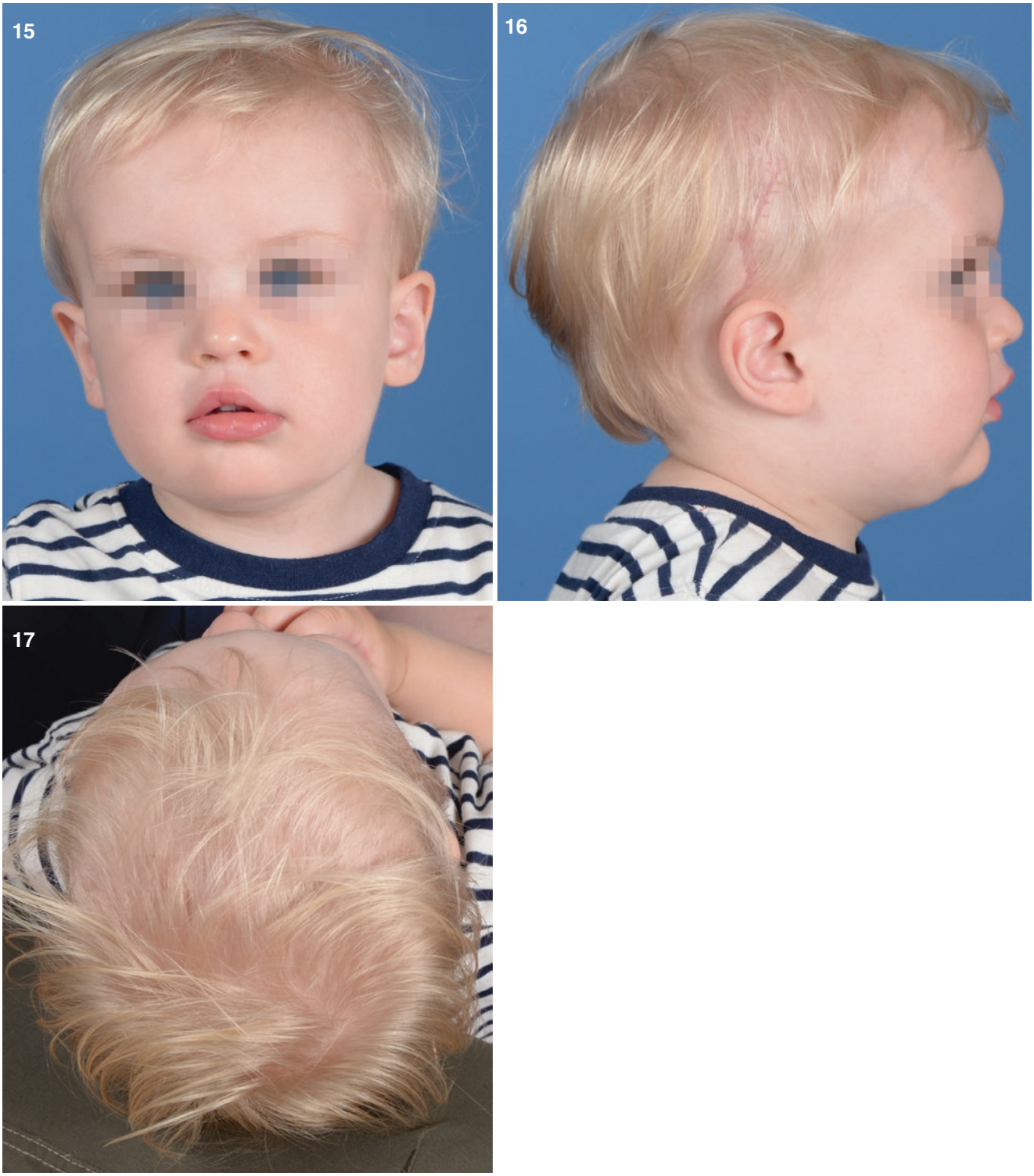




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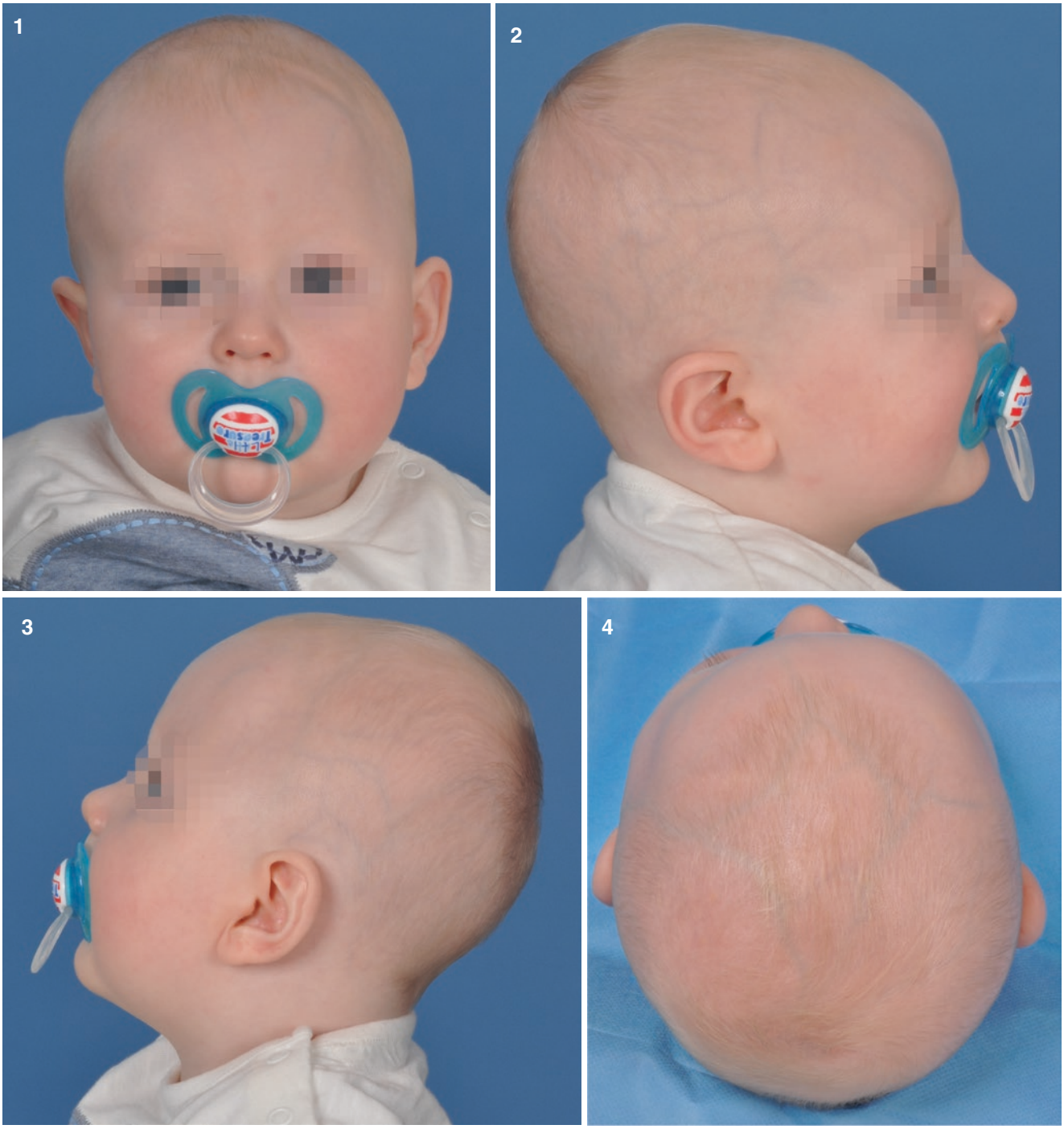
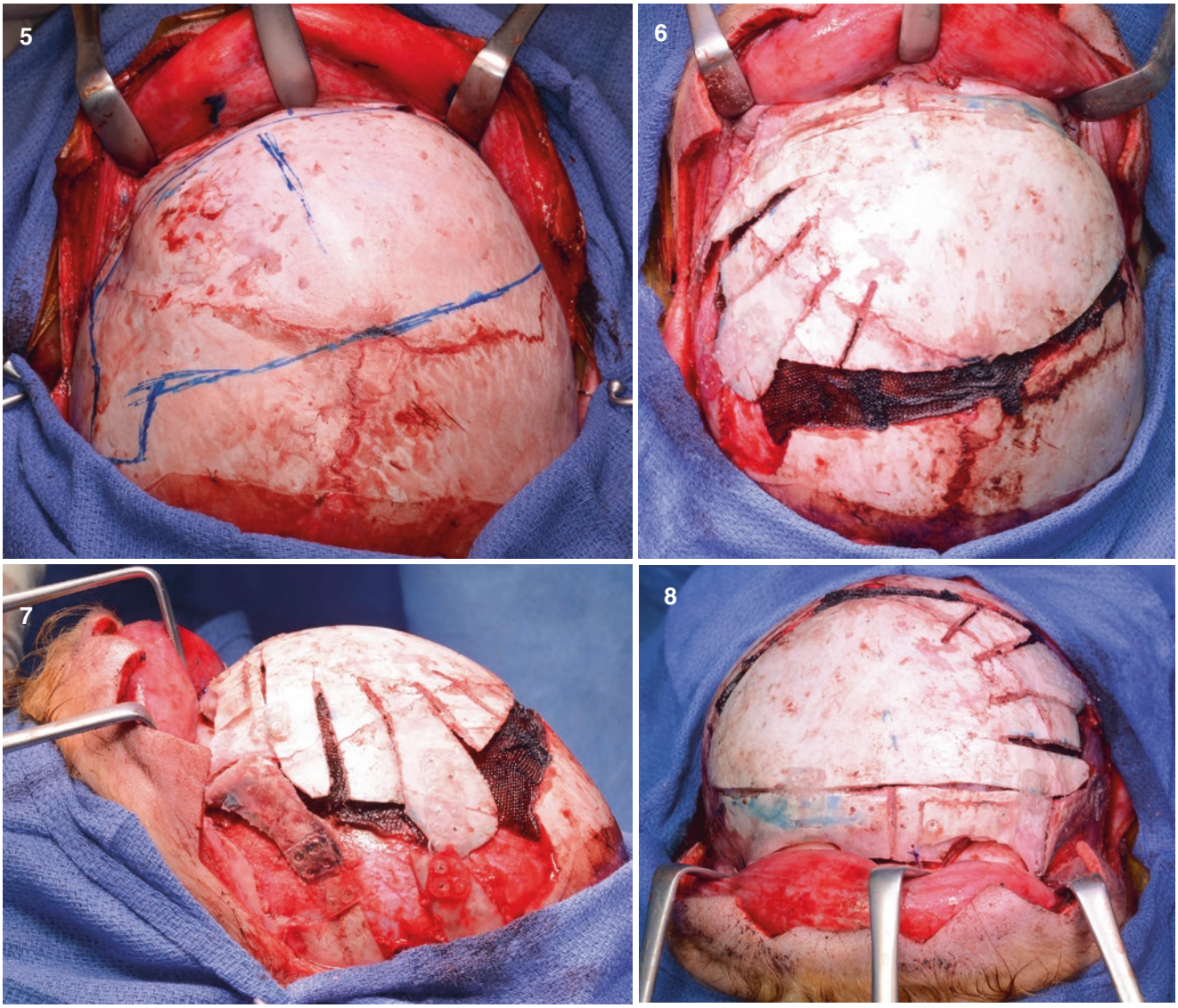



**Fig. 79.4** (4.1–4.12) Case series of unicoronal synostosis. (4.1–4.4) Preoperative facial views of patient with unicoronal synostosis, patient aged 31 weeks—left unicoronal synostosis—flattened left brow anterior plagiocephaly when viewed from above and ridging of the involved left coronal suture. (4.5–4.9) Intraoperative views of asymmetric fronto-orbital advance. (4.5) Precraniotomy planning—note the fused left coronal suture. (4.6–4.9) After fixation of the osteotomised segments—asymmetric fronto-orbital advancement; note onlay bone graft to left supraorbital region, fixation with absorbable plates and pins as well as absorbable sutures. (4.10–4.12) Postoperative facial views of patient with unicoronal synostosis

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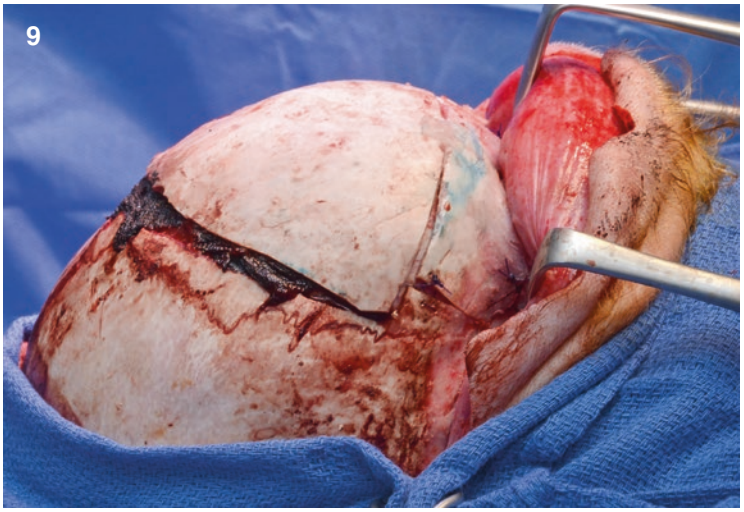


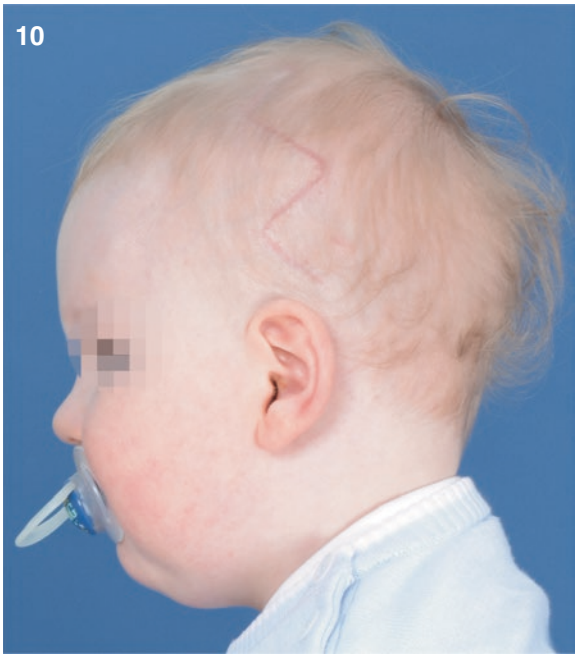



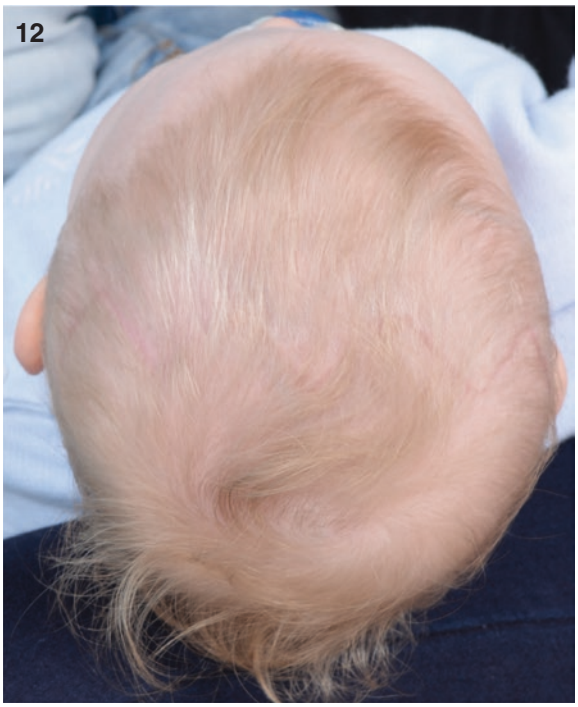




**Fig. 79.4** (continued)

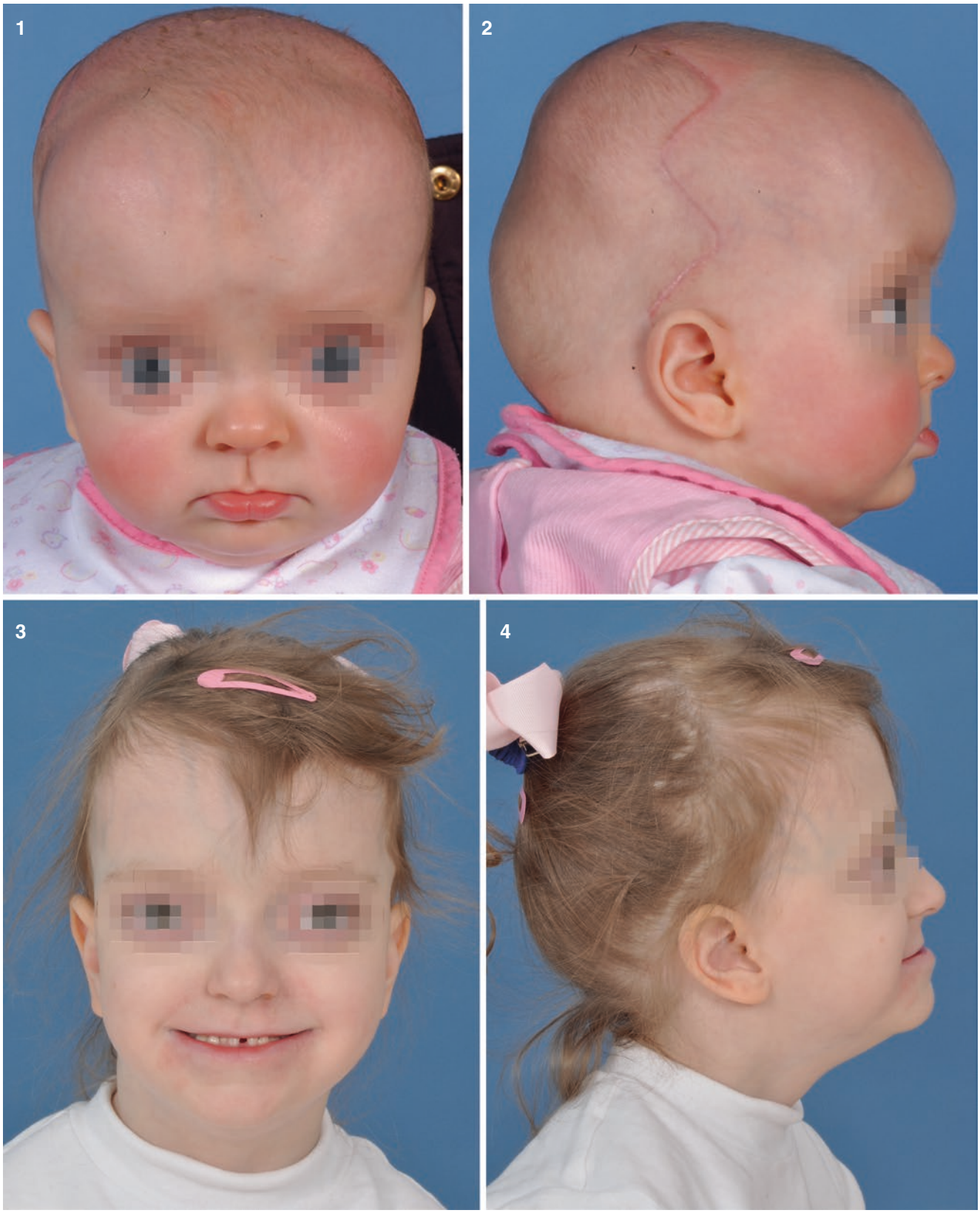


**Fig. 79.5** (5.1–5.13) Case series of Crouzon syndrome. (5.1 and 5.2) Patient with Crouzon syndrome after initial calvarial remodelling aged 34 weeks. (5.3 and 5.4) Preoperative facial views of same patient aged 5 with Crouzon's type facies. (5.5–5.7) Postoperative facial views after Le Fort III distraction osteotomies and static fronto-orbital advance with the rigid external distractor (RED frame) and internal distractors in place (day 1 post-op). The use of the internal distractors allows for the early removal of the external device after the active distraction period, acting as fixation devices for the retention period. (5.8–5.9) Final post-operative result immediately after RED frame distractor removal. (5.10–5.11) Late postoperative result following internal distractor removal. (5.12 and 5.13) Pre- and mid-distraction CT images of Le Fort III distraction and static fronto-orbital advance. Note internal distractors engaging zygoma (pushing Le fort III and external RED frame pulling midface). It is not our usual practice to obtain a CT mid treatment, but this was carried out due to concerns around a possible CSF leak. The static fronto-orbital advance is secured with absorbable, radiolucent, plates, but the multiple pin holes are visible

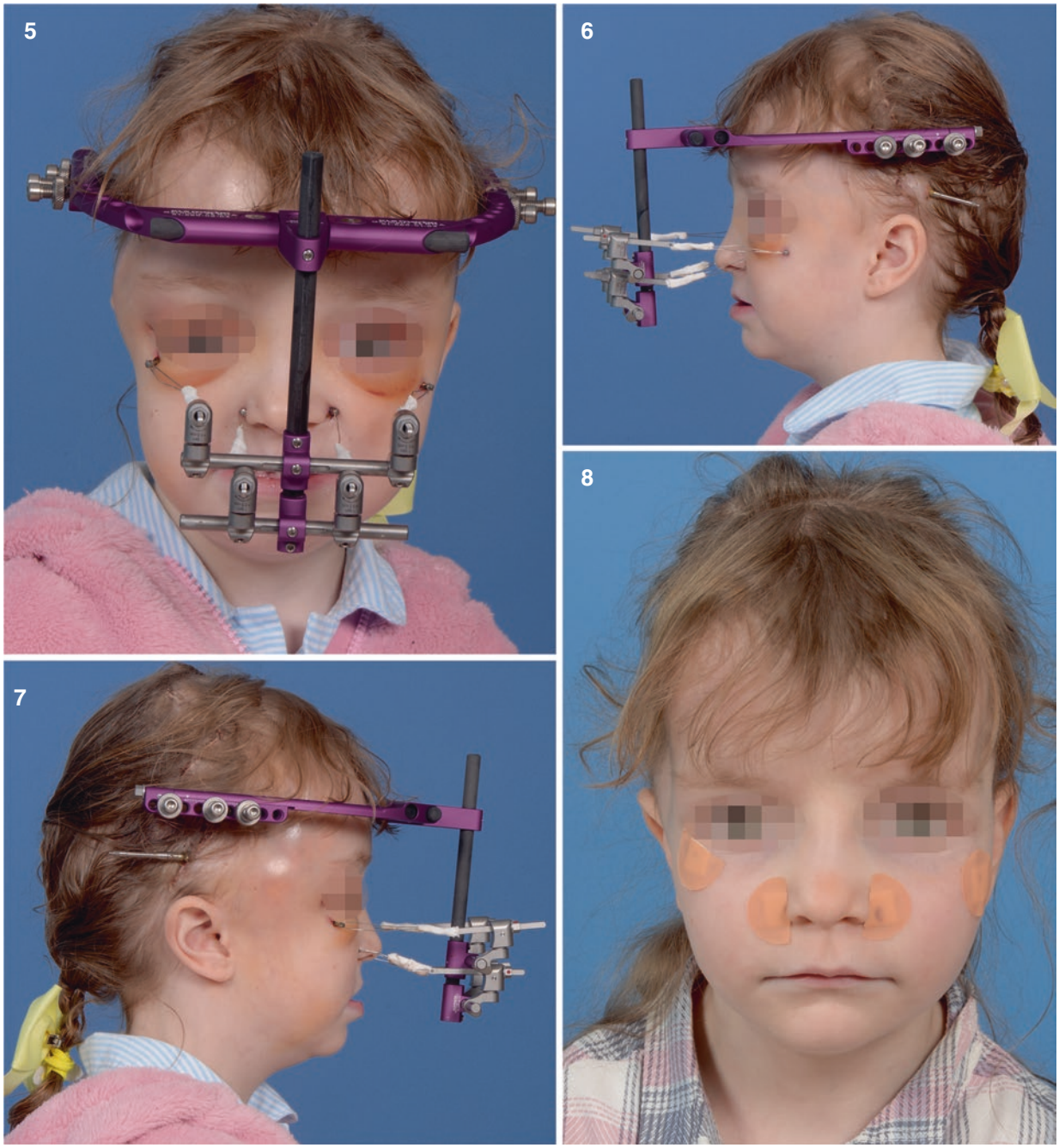


**Fig. 79.5** (continued)

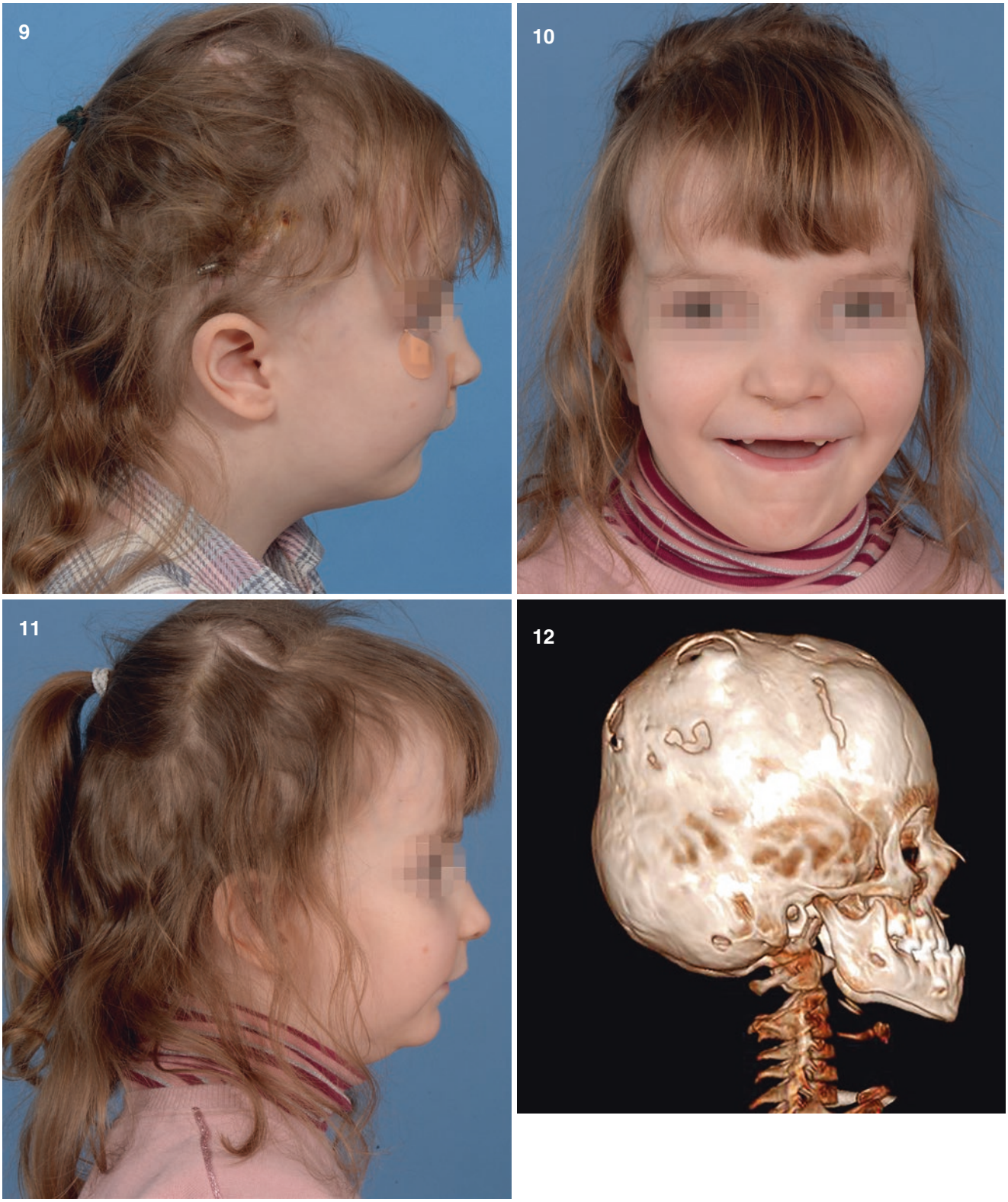


**Fig. 79.5** (continued)

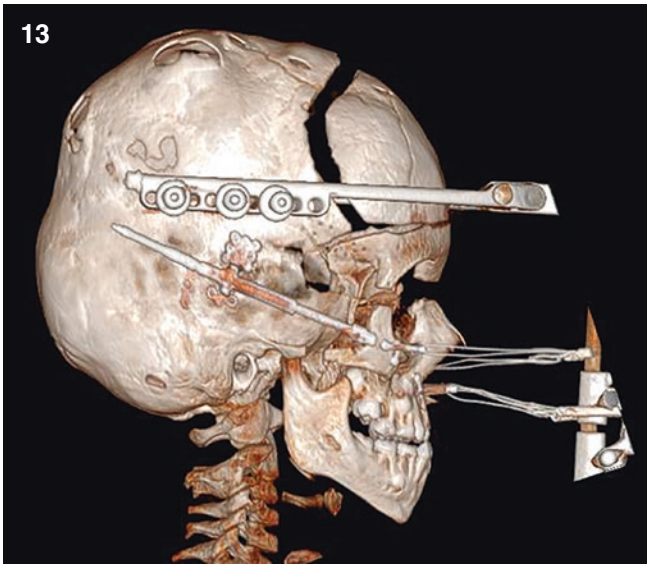




**Fig. 79.5** (continued)

timing and actual intervention, the Glasgow craniofacial service protocol is outlined in Table 79.3. See also Table 79.4 for an outline of the surgical procedures commonly carried out and the case series (Figs. 79.2, 79.3, 79.4 and 79.5) for an overview of treatment outcomes.

In the management of the syndromic cases, the treatment has to be more flexible and problem driven. There is considerable variation in the severity and comorbidities in this group of patients. Overall the aim is to minimise the number of surgical interventions; however re-synostosis, systemic problems, corneal exposure, airway obstruction and raised ICP are much more common in the group [14]. In the absence of additional pressing clinical issues, the syndromic synostosis cases can be managed as the non-syndromic cases in terms of the timing and extent of skull surgery.

### 79.2.4 Management in Early Childhood (3–8 Years)

In this period the management is primarily of observation. The non-syndromic cases are followed up 6 weeks, 6 months and 1 year post-skull vault surgery with growth, particularly head circumference, development and vision being monitored. A final review is usually undertaken immediately prior to the child starting school. Some units have a much longer follow-up period; however we reserve longer follow-up for the small proportion of patients who encounter ongoing development problems or symptoms. This protocol should be modified if there is a less well-developed community-based health surveillance programme.

**Table 79.3** Glasgow protocol for the management of non-syndromic synostosis

| Condition | Ideal age for intervention | Usual procedure |
|---|---|---|
| Sagittal synostosis | 5–7 months | Total vault remodelling |
| Metopic synostosis | 10–14 months | Fronto-orbital advancement, anterior remodelling ± hypotelorism correction |
| Unicoronal synostosis | 10–14 months | Fronto-orbital advancement, anterior remodelling—Bilateral |
| Bicoronal synostosis | 5–18 months | Posterior vault distraction |
| | 10–14 months | Bilateral fronto-orbital advancement, anterior remodelling |
| Lambdoid synostosis[a] | 10–14 months | Posterior vault expansion static or with distraction |

[a]In the presence of elevated ICP

The syndromic cases are followed up more closely with a minimum of annual reviews. In addition to the parameters noted above, the development of OSA may be insidious, and therefore at every review this should be asked about specifically. Towards the end of this period, consideration can be given to performing either a midface advancement or hypertelorism correction. This decision should be primarily driven by clinical problems rather than purely aesthetics, though teasing, bullying and psychological issues can, and should, play a role in decision-making. The input of child psychologists is often very helpful in developing a treatment plan and choosing the optimal timing [17].

During this period it is also important to monitor dental development and try to optimise routine dental care. In addition, at some time the patient should be referred for a full genetic consultation, though the timing of this can be determined by the needs and desires of the family. Throughout the care of the patient development, psychological, educational, physical and speech and language development should be monitored and intervention initiated when necessary.

**Table 79.4** Surgical procedures in craniofacial surgery

| Procedure | Access | Technical notes | Illustrations |
|---|---|---|---|
| Fronto-orbital advance and remodelling | – Bicoronal stealth incision in the Alice band region<br>– Anterior dissection to superior orbit for access to frontal bandeau inferiorly (may require osteotomies to free supraorbital nerves/vessels) | 1. Design mapped out and bur holes made for access with matchstick craniotome.<br>2. Frontal bone removed (pre−/post-suture).<br>3. Frontal bandeau removed—Cuts from anterior cranial fossa with direct vision inferiorly via the superior orbit.<br>4. Frontal bandeau and frontal bone remodelled as necessary and secured in advanced position. | Illustration 79.8a, b |
| Sagittal remodelling<br>Total vault remodelling | – Mid-cranial stealth incision for access to anterior and posterior calvarium<br>– Start in prone position with careful eye positioning and intermittent relief and then turn supine<br>– Dissection posteriorly to base of occipit and anteriorly to brow | 1. Map out dural sinuses.<br>2. Draw plan for osteotomies and create bur holes for access with matchstick craniotome.<br>3. Remove occipital bullet and barrel stave (sun) (bone harvesting as required from inner table).<br>4. Sagittal suturectomy and lateral barrel staving of mid calvarium/moulding laterally as far anteriorly as permitted whilst prone.<br>5. Secure occipital portion with PDS.<br>6. Turn the patient.<br>7. Frontal craniotomy/remodelling and inner table bone harvesting as required.<br>8. Complete suturectomy and barrel staving anteriorly.<br>9. Secure frontal bone (resorbable plates).<br>10. Fill in areas of bony discontinuity with bone putty (bone dust and tisseal). | Illustration 79.9 |
| Metopic remodelling | – Bicoronal stealth incision in the Alice band region<br>– Anterior dissection to superior orbit for access to frontal bandeau inferiorly (may require osteotomies to free supraorbital nerves/vessels) | 1. Design mapped out and bur holes made for access with matchstick craniotome.<br>2. Frontal bone removed (pre−/post-suture).<br>3. Frontal bandeau removed—Cuts from anterior cranial fossa with direct vision inferiorly via the superior orbit.<br>4. Frontal bandeau remodelled with interposing bone graft to increase width and flatten midline prominence.<br>• Internal resorbable strut plate for strength.<br>• Resorbable plates bitemporally ± bone graft in gap from advancement.<br>• Brow resecured in midline (resorbable plate/PDS).<br>5. Frontal bone remodelling (sectioning and repositioning ± barrel staving).<br>6. ± barrel staving and moulding of mid calvarium<br>7. Frontal bone placed and secured (resorbable plates/PDS). | Same as Illustration 79.8b |
| Unicoronal | – Bicoronal stealth incision in the Alice band region<br>– Anterior dissection to superior orbit for access to frontal bandeau inferiorly (may require osteotomies to free supraorbital nerves/vessels) | 1. Design mapped out and bur holes made for access with matchstick craniotome.<br>2. Frontal bone removed (post-coronal suture).<br>3. Frontal bandeau removed—Cuts from anterior cranial fossa with direct vision inferiorly via the superior orbit.<br>4. Frontal bandeau remodelled and secured:<br>• Internal resorbable strut plate for strength.<br>• Resorbable plates bitemporally ± bone graft in gap from advancement.<br>• Brow resecured in midline (resorbable plate/PDS).<br>5. Frontal bone remodelling (sectioning and repositioning ± barrel staving).<br>6. Frontal bone placed and secured (resorbable plates). | Asymmetric fronto-orbital advancement for unicoronal synostosis Illustration 79.10 |

**Table 79.4** (continued)

| Procedure | Access | Technical notes | Illustrations |
|---|---|---|---|
| Monobloc | – Bicoronal stealth incision in the Alice band region<br>– Anterior dissection (separate pericranial flap) to superior orbit for access to frontal bandeau inferiorly (may require osteotomies to free supraorbital nerves/vessels)<br>– Dissection to zygomatic arch bilaterally<br>– Intraoral access posterior to hamulus for pterygomaxillary disjunction<br>– Transconjunctival access to orbital floor | 1. Design mapped out and bur holes made for access with matchstick craniotome.<br>2. Frontal bone removed (pre−/post-suture).<br>3. Access to anterior cranial fossa and superior orbit.<br>4. Cuts as per frontal bandeau cuts in anterior cranial fossa with direct vision from superior orbit.<br>5. Cuts at zygomatic arches bilaterally.<br>6. Inferior orbital cuts from inferior orbital fissure laterally and then superiorly to join superior orbit cuts and medially from inferior orbital fissure running superiorly to join superior cuts.<br>7. Osteotomes used to achieve pterygomandibular disjunction.<br>8. Smiths/osteotomes to propagate splits.<br>9. Fixation may be for:<br>• Static advancement with plates at frontal bandeau butt joint posterior frontal bone bilateral zygomatic arches.<br>• Distraction—Internal distractor at posterior part of frontal process of zygomatic bone and pins for the RED frame in the body of the zygoma and paranasally.<br>• A combination—Static fronto-orbital advancement/remodelling (as for FOAR) with distraction of the Le fort III segment. | Illustration 79.11a, b |
| Subcranial Le fort III | – Bicoronal stealth incision in the Alice band region<br>– Anterior dissection to superior orbit for access to frontonasal junction (may require osteotomies to free supraorbital nerves/vessels)<br>– Dissection to zygomatic arch bilaterally<br>– Intraoral access posterior to hamulus for pterygomaxillary disjunction ± anteriorly for placement of distractors<br>– Transconjunctival access to orbital floor | 1. Cuts in orbital floors avoiding lacrimal apparatus leaving medial canthal tendon attached to bone.<br>2. Zygomatic cuts may be made from above or below depending on access.<br>3. Osteotome used to achieve frontonasal disjunction and pterygomaxillary disjunction.<br>4. Smiths to propagate splits.<br>5. Fixation may be for distraction as shown (and as per monobloc) or static advancement with fixation at the zygomatic arch, frontozygomatic suture, frontonasal junction and intermaxillary fixation. | Illustration 79.12—subcranial le fort III |
| Posterior vault distraction | – Mid-cranial stealth incision for access to the posterior calvarium<br>– Posterior dissection to base of occiput<br>– Stealth mapping of dural venous sinuses | 1. Posterior bullet osteotomy with matchstick bur and craniotome.<br>2. Dural mobilisation.<br>3. Positioning of 2/3 distractors and tunnelling anteriorly to exit either through the wound or anterior to it.<br>4. May incorporate foramen magnum decompression or insertion of telemetry can ICP monitor if required. | Illustration 79.13—Posterior vault distraction |
| Facial bipartition/orbital osteotomies | – Bicoronal stealth incision in the Alice band region<br>– Anterior dissection (separate pericranial flap) to superior orbit for access to frontal bandeau inferiorly (may require osteotomies to free supraorbital nerves/vessels)<br>– Dissection to zygomatic arch bilaterally<br>– Intraoral access posterior to hamulus for pterygomaxillary disjunction and anteriorly to separate nasal septum and split palate<br>– Transconjunctival access to orbital floor | As per monobloc with additional:<br>1. Separation of the nasal septum from the palate via an intraoral incision.<br>2. Midline osteotomy of the palate.<br>3. Midline removal of bone as required from glabella/nasal region.<br>4. Secure as per monobloc with additional midline fixation and a palatal splint.<br>Box osteotomies of the orbits to achieve orbital movement separate from the maxillary movement involve the same cuts without disjunction at the Le fort I level | Illustration 79.14a, b |

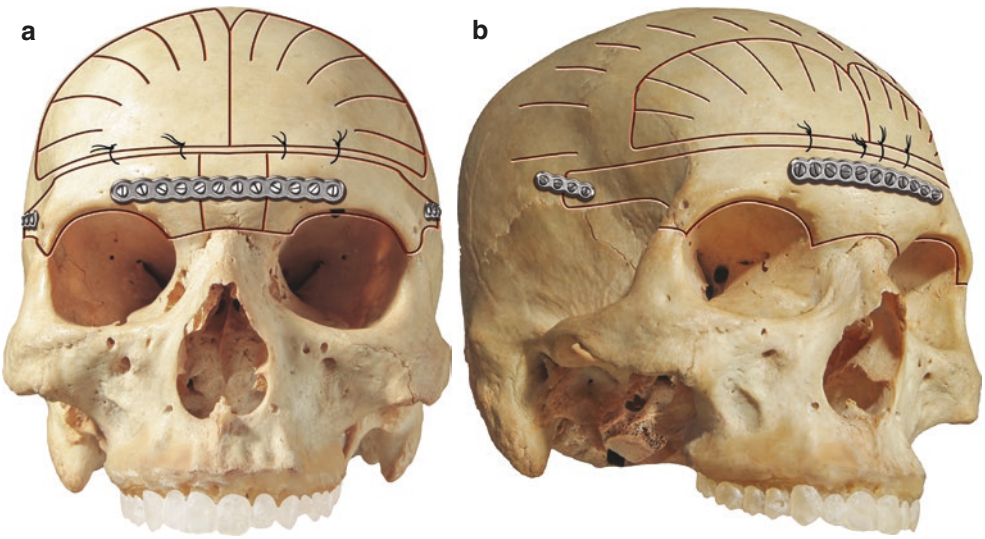


**Illustration 79.8** (**a**, **b**) Fronto-orbital advance and remodelling





**Illustration 79.9** Sagittal remodelling/total vault remodelling



**Illustration 79.10** Asymmetric fronto-orbital advancement for unicoronal synostosis

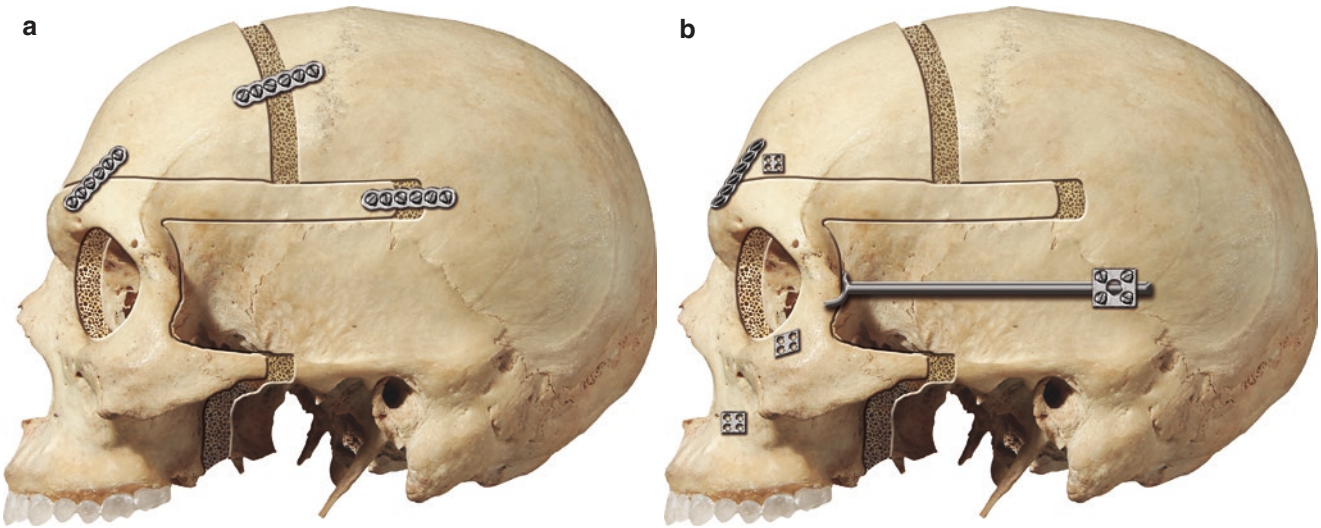




**Illustration 79.11** (**a**, **b**) Monobloc advancement

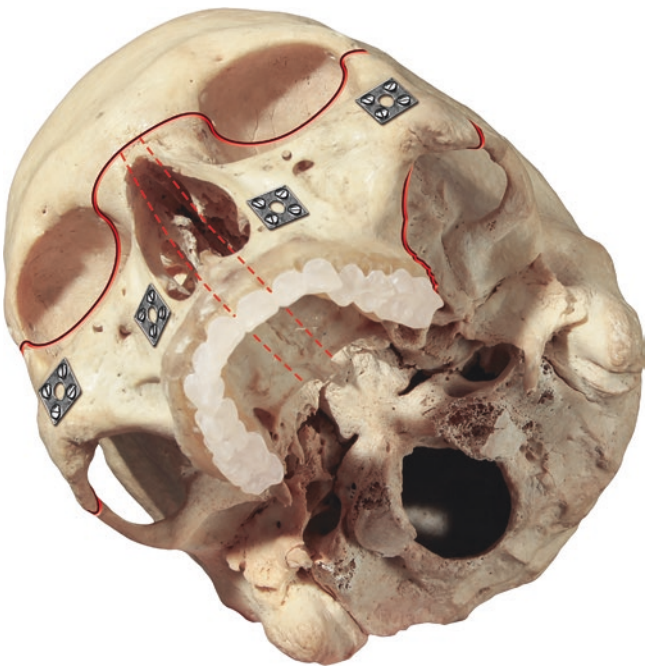



**Illustration 79.12** Subcranial Le Fort III

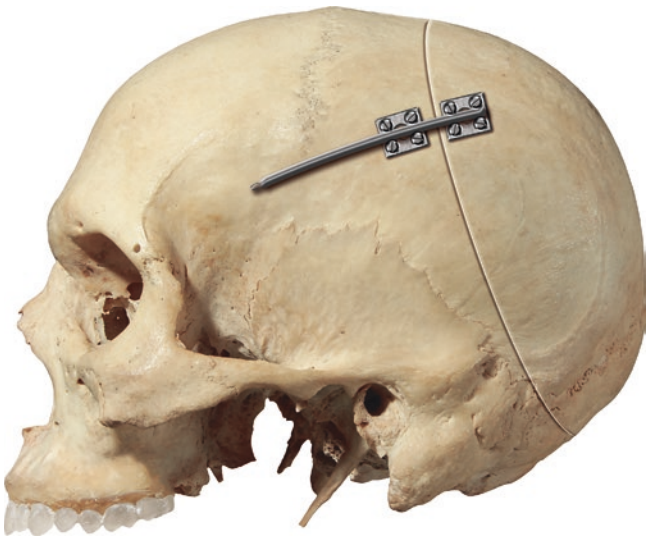



**Illustration 79.13** Posterior vault distraction

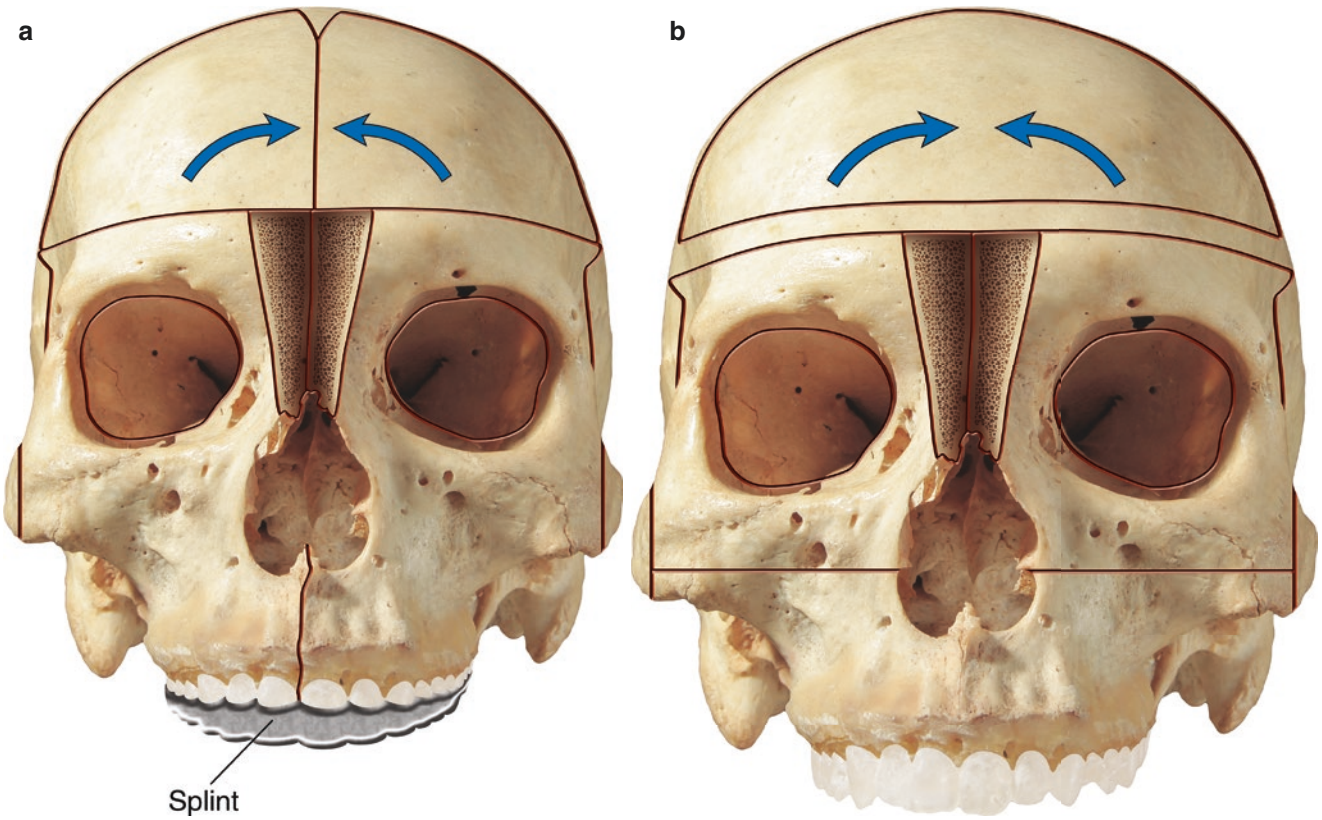




**Illustration 79.14** (**a**) Facial bipartition. (**b**) Box osteotomy to correct hypertelorism

### 79.2.5 Management in Middle Childhood (8–12 Years)

During this period routine monitoring is necessary, and as the face and dentition develop, attention can be turned to managing the midface and occlusion. During the transition from the deciduous to permanent dentition, it is important to have an orthodontic assessment—so that a definitive orthodontic/orthognathic plan can be developed. One of the major factors that may prompt early midface surgery would be the development of OSA; thus regular questioning and targeted investigations at review appointments are necessary. In considering orthodontics it is important to be mindful of the fact that repeated courses of orthodontics will lead to root shortening and also potentially exhaust the tolerance and cooperation of the patient and his/her family [18].

In general, the routine correction of hypertelorism can be safely performed from about the age of 8 years; however this is determined by the chosen technique (facial bipartition vs box osteotomy) (illustration 79.14a, b) and whether an early midface advancement is necessary for the management of OSA [19]. As with all surgery aimed primarily at improving facial appearance, the indications, objectives and expectations of surgery must be discussed at length with the patient and his/her family and a consensus reached. The input from specialist child psychologists is often very helpful. The transition from primary to middle or secondary school is often traumatic, and interventions before this time can be very helpful from a psychological perspective, but it needs to be acknowledged and accepted that further surgery at the completion of growth may well be necessary [18].

### 79.2.6 Management in Adolescence and after the Completion of Growth

At this stage in development, definitive corrective surgery (when necessary) should be planned and an integrated orthodontic/surgical plan developed. In the syndromic cases, ongoing monitoring for functional and developmental issues is required, and as the child matures, the drive and the responsibility for decision-making will move towards the patient and away from their parents. Many adolescents at this stage are tired of multiple hospital appointments and are not keen on further interventions; in these circumstances it is

important to work with the patient and their family to provide care but also respect the patient's wishes and desires. It is better to postpone treatment rather than have a half completed course of orthodontics with poor cooperation and the consequent complications and finally lose the confidence of the patient.

The planning of comprehensive surgery will need assessment in the following areas:

1. Skull shape—irregularities, defects.
2. Orbital position and symmetry.
3. Upper midface morphology.
4. Lower midface morphology.
5. Mandibular morphology.
6. Nasal form.
7. Soft tissue issues.

It is always best to offer and plan for a total correction in the first instance. The discussion around further transcranial surgery is often key to the decision-making. Many patients and families do not want to undertake any further transcranial surgery, in spite of the significant benefits, particularly for hypertelorism or dystopia corrections. None the less these procedures can offer significant improvements in the final result [19].

For residual skull defects of approximately 2 cm diameter or larger, it is usual to offer some form of reconstruction; this could be autologous (usually cranial bone), or alloplastic. Alloplastic reconstructions may be of a variety of materials and can be CAD/CAM designed or shaped intraoperatively. One of the most common residual defects following a fronto-orbital advancement is a degree of temporal hollowing; this can be effectively addressed with the use of mouldable hydroxyapatite synthetic bone material. This technique can be used to satisfactorily address minor skull irregularities and defects, and it is also possible to combine this technique with the use of alloplastic custom-made cranioplasties (e.g. titanium or PEEK) [20].

The assessment of orbital position is critical to deciding on what operative procedure to offer. The combination of hypertelorism and marked maxillary narrowing (particularly if the upper incisors are crossed), as is often seen in Apert syndrome, lends itself to correction with a facial bipartition; this technique can also be used early as the bone cuts do not result in damage to the dentition. The major drawback of this technique is that orbital asymmetries cannot be addressed. The more conventional box osteotomy is used to effect asymmetric movements, hypertelorism and dystopia corrections. The overall aesthetic outcome is often enhanced with nasal augmentation and soft tissue surgery, but the final improvement (particularly in severe hypertelorism cases) unfortunately often does not match the skeletal changes achieved.

The upper midface can be advanced, either as part of the bipartition procedure, transcranial Le Fort III (monobloc), subcranial Le Fort III or Le Fort II. These procedures can be performed conventionally with bone grafting and fixation with either absorbable or titanium plates and screws or with the use of distraction techniques. If distraction is being utilised, the devices can be a halo-based external device or buried devices. It is sometimes helpful to combine external distraction devices with buried devices which facilitate the early removal of the somewhat cumbersome halo device for the retention period [21]. In addition to advancement movements, it is possible to incorporate height changes, but rotatory movements (e.g. to correct the upper dental midline) are very difficult unless the symmetry is equal at all facial levels. Furthermore if the maxillary advancement required has a significant differential between the upper component and dental component, it is possible to combine a Le Fort III (transcranial or subcranial) or a monobloc advance with a simultaneous Le Fort I osteotomy. This additional level of movement allows for differential advancements and/or height changes as well as rotations at the lower level. See case series in Fig. 79.5 of Crouzon syndrome.

The mandible including the chin should be considered as one would for conventional orthognathic surgery, but caution should be exercised in setback procedures as correction of the occlusion to a retrusive maxilla may worsen or precipitate OSA.

Nasal deformity is best dealt with as close to the end of treatment as possible; the range of deformity is considerable; each patient should be treated on their own merits. In cases with severe deficiency, dorsal augmentation with alloplastic or autologous material may be necessary, and this can be refined with nasal tip surgery.

The final phase of surgery is the management of the soft tissues, and again a variety of techniques can be employed to improve the overall aesthetic result. As well as rhinoplasty techniques, facelifting, brow suspension, eyelid surgery, soft tissue augmentation with fillers or fat transfer and soft tissue reduction with direct excision or liposuction all have their place.

See Table 79.2 for an outline of the management of common clinical problems and Table 79.4 for an outline of surgical procedures.

## References

1. Wilkie AO, Byren JC, Hurst JA, Jayamohan J, Johnson D, Knight SJ, et al. Prevalence and complications of single-gene and chromosomal disorders in craniosynostosis. Pediatrics. 2010;126(2):e391–400.
2. Bhadkamkar MA, Albright SB, Wolfswinkel EM, Bollo R, Buchanan EP. Posterior cranial vault distraction in the treatment

of shunt-induced craniosynostosis. J Craniofac Surg. 2015 Jan 1;26(1):e70–2.
3. Di Rocco F, Arnaud E, Renier D. Evolution in the frequency of non-syndromic craniosynostosis. J Neurosurg Pediatr. 2009;4(1):21–5.
4. Mathijssen IM. Guideline for Care of Patients with the diagnoses of Craniosynostosis: working group on Craniosynostosis. J Craniofac Surg. 2015;26(6):1735–807.
5. Cornelissen MJ, Apon I, van der Meulen JJ, Groenenberg IA, Kraan-van der Est MN, Mathijssen IM, Bonsel GJ, Cohen-Overbeek TE. Prenatal ultrasound parameters in single-suture craniosynostosis. J Matern Fetal Neonatal Med. 2018 Aug 3;31(15):2050–7.
6. Edmond JC, Nischal K, Forbes B, Katowitz W, Costakos D. Update on the management of patients with craniostenosis. J AAPOS. 2011;15(1):e35.
7. Burokas L. Craniosynostosis: caring for infants and their families. Crit Care Nurse. 2013;33(4):39–50.
8. Mehta VA, Bettegowda C, Jallo GI, Ahn ES. The evolution of surgical management for craniosynostosis. Neurosurg Focus. 2010;29(6):E5.
9. Ingraham FD, Alexander E, Matson DD. Clinical studies in craniosynostosis: analysis of fifty cases and description of a method of surgical treatment. Surgery. 1948 Sep 1;24(3):518–41.
10. Jimenez DF, Barone CM, Cartwright CC, Baker L. Early management of craniosynostosis using endoscopic-assisted strip craniectomies and cranial orthotic molding therapy. Pediatrics. 2002;110(1 Pt 1):97–104.
11. Lauritzen CG, Davis C, Ivarsson A, Sanger C, Hewitt TD. The evolving role of springs in craniofacial surgery: the first 100 clinical cases. Plast Reconstr Surg. 2008;121(2):545–54.
12. Khechoyan D, Schook C, Birgfeld CB, Khosla RK, Saltzman B, Teng CC, Ettinger R, Gruss JS, Ellenbogen R, Hopper RA. Changes in frontal morphology after single-stage open posterior–middle vault expansion for sagittal craniosynostosis. Plast Reconstr Surg. 2012 Feb 1;129(2):504–16.
13. Garza RM, Khosla RK. Nonsyndromic craniosynostosis. Semin Plast Surg. 2012;26(2):53–63.
14. Beez T, O'kane R, Piper I, Koppel D, Sangra M. Telemetric intracranial pressure monitoring in syndromic craniosynostosis. J Craniofac Surg. 2016 Jun 1;27(4):1032–4.
15. Benson ML, Oliverio PJ, Yue NC, Zinreich SJ. Primary craniosynostosis: imaging features. AJR Am J Roentgenol. 1996;166(3):697–703.
16. Gonsalez S, Hayward R, Jones B, Lane R. Upper airway obstruction and raised intracranial pressure in children with craniosynostosis. Eur Respir J. 1997 Feb 1;10(2):367–75.
17. Broder HL. Using psychological assessment and therapeutic strategies to enhance Well-being. Cleft Palate Craniofac J. 2001;38(3):248–54.
18. Evans RD, Bradford R. The dental and orthodontic management of craniosynostosis. In: Bux M, editor. The clinical Management of Craniosynostosis ch 11. London: Mac Keith Press; 2004. p. 211–7.
19. Gonzalez AM, Elahi M, Barakat K, Yavuzer R, Brinkmann B, Jackson IT. Hypertelorism: the importance of three-dimensional imaging and trends in the surgical correction by facial bipartition. Plast Reconstr Surg. 2005 May 1;115(6):1537–46.
20. Feroze AH, Walmsley GG, Choudhri O, Lorenz HP, Grant GA, Edwards MS. Evolution of cranioplasty techniques in neurosurgery: historical review, pediatric considerations, and current trends. J Neurosurg. 2015 Oct;123(4):1098–107.
21. Schulten A, Lim AA, Bruun RA, Hayes C, Mulliken JB, Padwa BL. Combined push-pull distraction for correction of syndromic midfacial hypoplasia. J Oral Maxillofac Surg. 2006 Jan 1;64(1):23–30.