



# Re-imagining early cloverleaf skull deformity management from front to back approach—30 years on

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## Abstract

The cloverleaf skull deformity remains among the most complicated craniofacial conditions to successfully manage. Many cases achieve largely unsatisfactory outcomes due to the requirement for frequent reoperation on the cranial vault and failure to deal with all the elements of the craniofaciostenosis in a timely fashion. Early cranial vault surgery without addressing the cranial base deformity and its attendant cerebrospinal fluid flow changes is invariably challenging and disappointing. A recent focus on the expansion of the posterior cranial vault as a primary procedure with the greater volume change allows a delay in fronto-orbital advancement and reduced need for repeat surgery. We herein describe three cases of complex multisuture craniosynostosis with cloverleaf skull deformity who underwent neonatal posterior cranial vault decompression along with foramen magnum decompression. Our report examines the safety and rationale for this pre-emptive surgical approach to simultaneously deal with the cranial vault and craniocervical junction abnormalities and thus change the early trajectory of these complex cases.

**Keywords** Cloverleaf skull syndrome · Decompressive craniectomy · Foramen magnum · Craniofacial abnormalities

## Introduction

The cloverleaf skull deformity (Kleeblattschadel or triphyllocephaly), with its attendant orbito-facio-stenosis, remains among the most challenging expression of craniosynostosis to manage short and long term [1]. The complex patterns of multisuture craniosynostosis involving both the calvarium and cranial base, with the attendant cerebrospinal fluid flow changes and hydrocephalus, produce a complicated, dynamic, rapidly evolving neurocranial environment,

of which our understanding remains limited. Skull base synostosis, causing the development of short and shallow orbits and gross midfacial retrusion, significantly adds to the potential early ocular and airway morbidity, where treatment is not timely and appropriate.

Thirty years ago, we reported 10 cases of cloverleaf skull deformity with limited follow-up, suggesting that a coordinated multidisciplinary team-based approach was indicated to achieve a useful long-term outcome [2]. Whilst we opined at that time that primary treatment of the calvarial deformity was one of the most uncomplicated aspects of the treatment program, later observation of the clinical progression of our cases and the development of new approaches and timing of cranial vault remodelling procedures suggests otherwise. Frequent reoperation on the cranial vault, reactive management of CSF flow issues, the foreshortened orbits, and the upper airway saw the three of the surviving cases in our series achieve unsatisfactory functional and aesthetic outcomes. These early first-generation craniofacial procedures failed to consistently achieve release and expansion of the constricting cranial vault in a milieu of dynamic and complex CSF flow changes, with a compounding effect over the growing life of the child. This results in frequent revision cranial vault procedures, bony

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resorption, infection, poorly controlled and often multiple ventricular diversionary procedures and delay or failure to perform midface surgery successfully.

Our series sequenced surgery [2], by comprehensive suturectomy and subsequent fronto-orbital advancement (FOA) or lambdoid craniectomy, invariably followed by ventriculoperitoneal (VP) shunt. Whist Jarrahy et al. proposed neonatal VP shunting followed by staged anterior and then posterior vault surgery in the first 12 months of life [3]. Aesthetic and functional outcomes were deemed acceptable, but follow-up was limited to 18 months.

A previous review article on various posterior cranial decompression techniques by Nowinski et al. advocated that they have become the accepted first stage to increase cranial volume in multisuture craniosynostosis, usually around 3–6 months of age [4]. This then permits a delay in FOA until such time as it results in a stable ‘permanent morphological result’—as close to age 2 years as possible.

Improved CT and MRI imaging and quantification in these complex syndromic craniosynostosis cases have identified cranial base changes and CSF flow alterations as early as the neonatal period. Rijken et al. confirmed that the foramen magnum surface area is smaller, and the intra-occipital synchondroses fuse earlier in patients with Crouzon–Pfeiffer syndrome, these changes being present at birth and increasing over time [5]. Whilst the exact cause for ventriculomegaly and raised intracranial pressure (ICP) in these complex syndromic cases is still debated [6], if a reduction in foramen magnum size inhibits normal CSF passage around the posterior cranial base, there exists the possibility of early pre-emptive posterior cranial vault and cranial base decompression to alter the early clinical course of these cases positively.

Herein, we report three recent cases with established complex multisuture craniosynostosis presenting as cloverleaf skull deformity, who underwent neonatal posterior cranial vault decompression in concert with foramen magnum decompression, examining the safety and rationale for such an approach against a background of long-term outcomes.

## Clinical presentation

### Case 1: SM–Crouzon

This male infant born to non-consanguineous Afghan parents at 41 weeks gestation was noted at birth to have a trilobate skull deformity. An antenatal gestational morphology scan at 22 weeks showed a normal craniofacial configuration. He was intubated after meconium-induced pulmonary hypertension produced respiratory distress.

Clinical examination at birth identified a classical cloverleaf skull deformity with frontal turricaphaly, a tense anterior

fontanelle, and bilateral symmetric bulging of the middle cranial fossae with no palpable bone over the temporal lobes. The occiput was flattened with apparent posterior cranial vault constriction. There was a supraorbital bony recession and marked bilateral proptosis. Midface hypoplasia without overt upper airway obstruction, low-set ears, and a high-arched palate without evidence of clefting completed the craniofacial examination. The hands and feet were normal.

Computed tomography (CT) scan with 3D reformatting demonstrated the distinctive cloverleaf deformity, with grossly thinned or absent calvarial bone over the vertex and temporally. The occiput was thickened and flattened in the midline, with thinning laterally and deep bony projections into the dura, with the appearance of craniolacunae. This resulted in grossly constricted posterior cranial fossa, near vertical tentorium cerebelli, and obstructive hydrocephalus (Fig. 1a). Magnetic resonance imaging (MRI) scan confirmed a Chiari I malformation (Fig. 1d) and triventricular hydrocephalus with restricted CSF flow.

Genetic testing confirmed a c.1061C > G mutation with p.Ser354Cys substitution of FGFR2 consistent with Crouzon Syndrome.

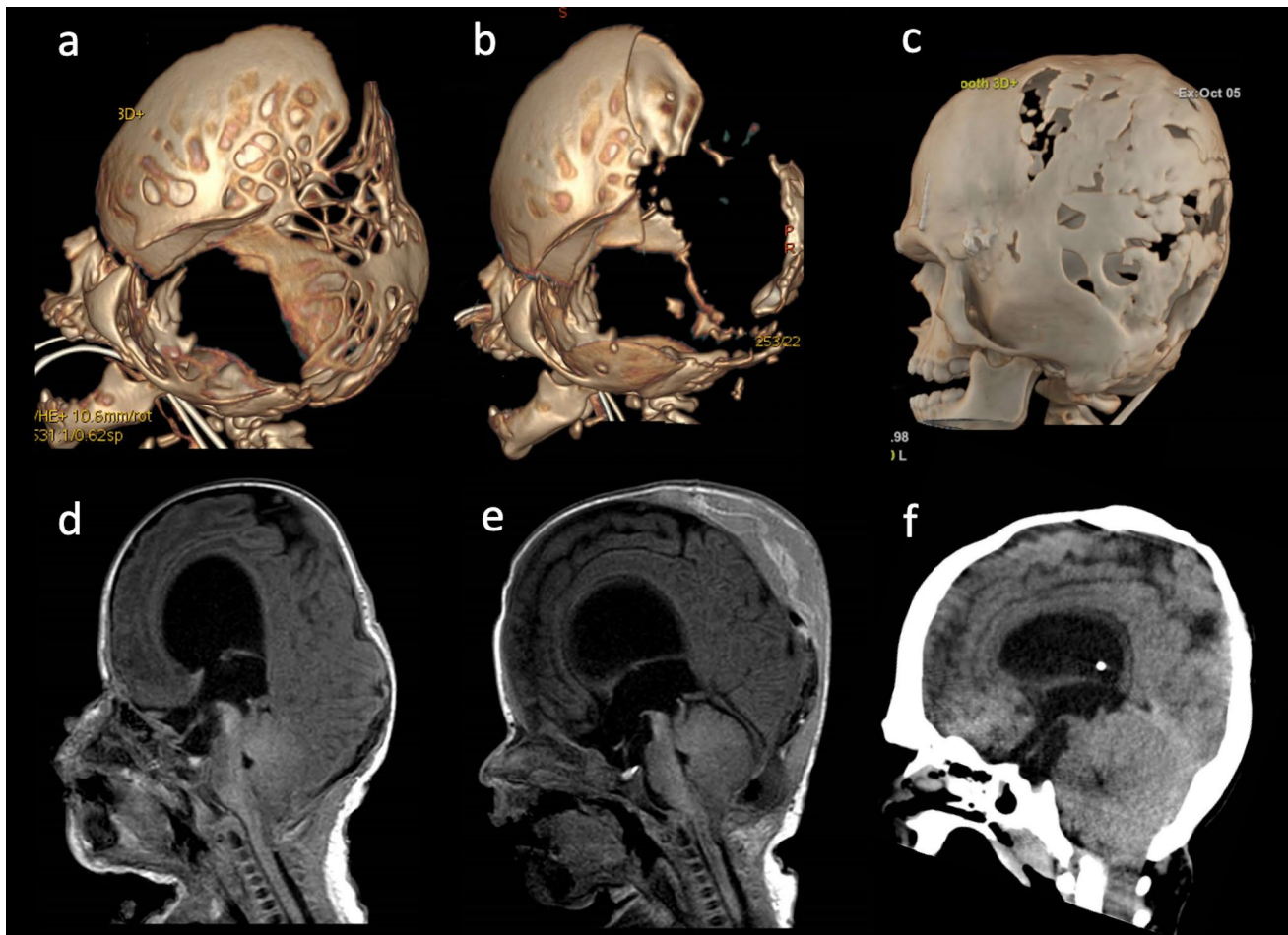
At day 13 of life, he underwent formal lateral tarsorrhaphies for increasing proptosis and resection of the posterior margin of the soft palate posterior to the muscle as to increase the posterior nasal space in alleviating the mechanical airway obstruction.

Two days later, with an obvious increase in anterior fontanelle tension, he underwent a posterior cranial vault craniectomy and foramen magnum decompression [7]. The patient was positioned prone during the operation, and intraoperative blood salvage was used. It is pivotal to provide proper care to prevent corneal exposure and eye globe compression and ensure the endotracheal tube remains in place. The procedure involved a meticulous dissection of the bony spicules projection into the dura, resulting in a wide bony excision from just posterior to the fused coronal ring of sutures back to and including the posterior rim of the foramen magnum. The blood transfusion required was less than 100 ml, and recovery from this surgery was uneventful.

Correction of the significant ventricular enlargement was not achieved, and he initially progressed to a 3rd ventriculostomy at age 4 months, followed by a VP shunt at age 5 months. Careful monitoring of shunt pressures attempted to titrate and promote appropriate skull growth and shape.

Fronto-orbital advancement of 1.5 cm was performed at age 18 months with the VP shunt now turned off—this has remained so till follow-up now aged 3 years.

An adenoidectomy was performed at age 2 years 9 months, and now, at age 3 years, he has an airway not requiring any support, eyes stable, and well protected, and is developing neurologically at an age-appropriate rate (Fig. 2a).



**Fig. 1** Case 1. 3D CT scans show **a** preoperative, **b** immediate postoperative, and **c** at age of 24 months demonstrating reasonable expansion of the posterior skull after the posterior cranial vault and foramen magnum decompression procedure. **d** Preoperative MRI scan which demonstrates significant posterior cranial restriction and Chiari

I malformation. **e** Postoperative MRI at 6 days after posterior cranial decompression revealing improvement at posterior cranial area. **f** CT scan at 24 months of age after definite fronto-orbital advancement procedure

## Case 2: P–multisuture synostosis

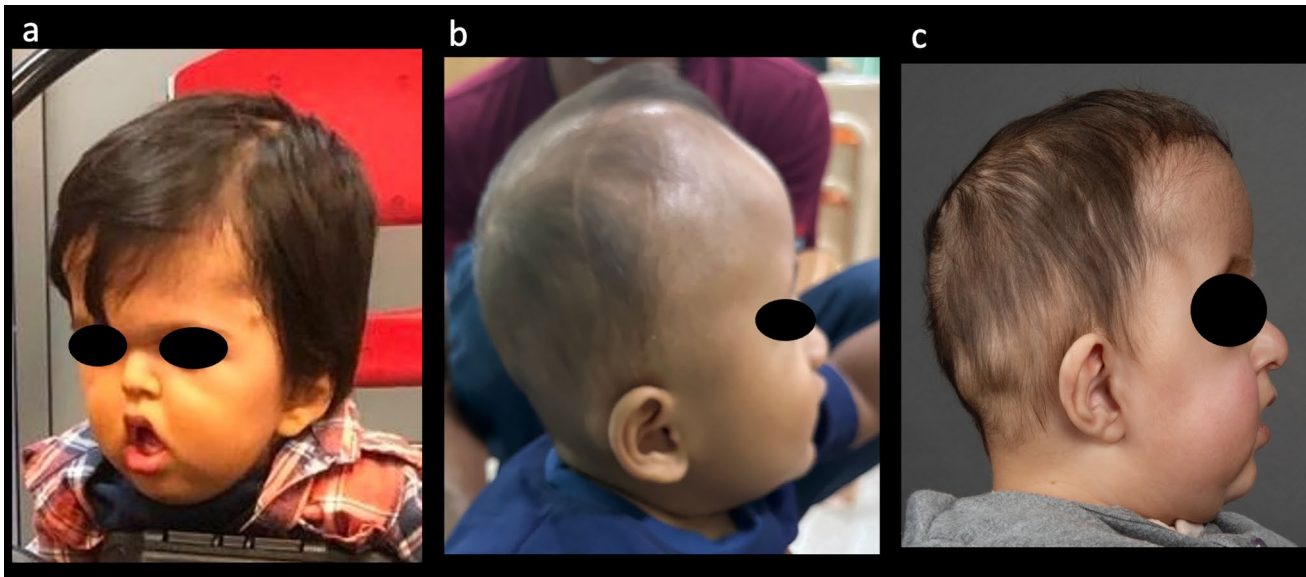
A male infant from non-consanguineous Myanmar parents with no family history of craniofacial disease was born term at 37 weeks of gestational age. He was referred to Siriraj Craniofacial Centre at age of 1 month due to a trilobate head with tense bulging of the anterior fontanelle (Fig. 3). The overnight oxygenation monitoring showed significant desaturations and continuous oxygen treatment was initiated. The physical examination also showed receded supraorbital rims, bilateral eye proptosis, mid-face hypoplasia and bifid uvula. All hands and extremities were unremarkable. The fundoscopy revealed no papilloedema. The laryngotracheobronchoscopy revealed unremarkable and no tracheomalacia.

The 3D CT showed multisuture craniosynostosis involving coronal, sagittal, and lambdoid sutures. There were temporal lobes extruding through the splaying of the squamosal

sutures and bulging of the brain parenchyma through the open anterior fontanelle, consistent with a cloverleaf skull deformity. There was effacement of subarachnoid spaces and mild hydrocephalus, as shown in Fig. 3.

Further genetic testing by whole-exome sequencing revealed heterogenous c.278C > G mutation with p.Ala93Gly of the *SIX3* gene.

The child underwent lateral tarsorrhaphies for proptosis eye management. The posterior cranial decompression by wide craniectomy was performed along with foramen magnum decompression at the age of 2 months (Fig. 3d). The technique is identical to the Case 1 operation. We also used intraoperative blood salvage to reduce the transfusion to 35 ml. Unfortunately, the hydrocephalus has been progressive and managed by undergoing a programmable VP shunt at the age of 4 months. Follow-up CT at postoperative 4 months revealed remarkable cranial expansion a the



**Fig. 2** Recent images of the index cases. **a** Case 1. **b** Case 2. **c** Case 3. All patients demonstrate expanded occiput and rounder head shapes

parieto-occipital area and improved CSF space between the cerebellar and the brain stem (Fig. 3e, f). Moreover, overnight oxygenation at age of 10 months revealed no desaturation. The boy's development is progressively improved and suitable with age (Fig. 2b).

### Case 3: GS–Crouzon

This male infant born to non-consanguineous Indian parents presented at term with a significant cranial vault deformity consistent with sagittal, asymmetrical, coronal, and lambdoid synostosis. Marked frontal prominence, right temporal bulging, and progressively worsening proptosis were evident during the first few days of life. CPAP was required at birth, followed by a continuous oxygen requirement. At 1 week of age, he underwent formal lateral tarsorrhaphies for proptosis and a laryngotracheobronchoscopy which revealed a normal lower airway.

CT scan imaging confirmed multisuture craniosynostosis of sagittal, right coronal, and bilateral lambdoid (Fig. 4a, b). There was no overt hydrocephalus nor Chiari I malformation. MRI scan CSF flow studies showed absent posterior flow through the foramen magnum (Fig. 4c).

With the early progression of the deformity and a tense bulging fontanelle, it was decided to proceed to an early posterior vault craniectomy and decompression of the foramen magnum, occurring on day 17 of life. As in Case 1, bony decompression occurred from the anterior fontanelle backwards—including a 5-cm wide craniectomy of the fused

sagittal suture, removal of the bone over the occiput, down to and including the posterior border of the foramen magnum, leaving exposed the sagittal and transverse sinuses. A small segment of bone over the posterior sagittal suture was preserved, where a sinus pericranii was evident. A blood transfusion of only 45 ml was necessary, and early post-operative recovery was uneventful. We used a head drape for a few days after the operation and recommended the patient sleep on their side to avoid pressure on the back of the head. If they had to lie on their back, we used a doughnut ring pillow to allow for brain expansion towards the back. Further frontal cranial vault remodeling is anticipated (Fig. 2).

A follow-up CT scan at 6 weeks confirmed extensive regrowth of bone over a much more normal posterior head shape, with persistent opening of the posterior arch of the foramen magnum (Fig. 3d, e).

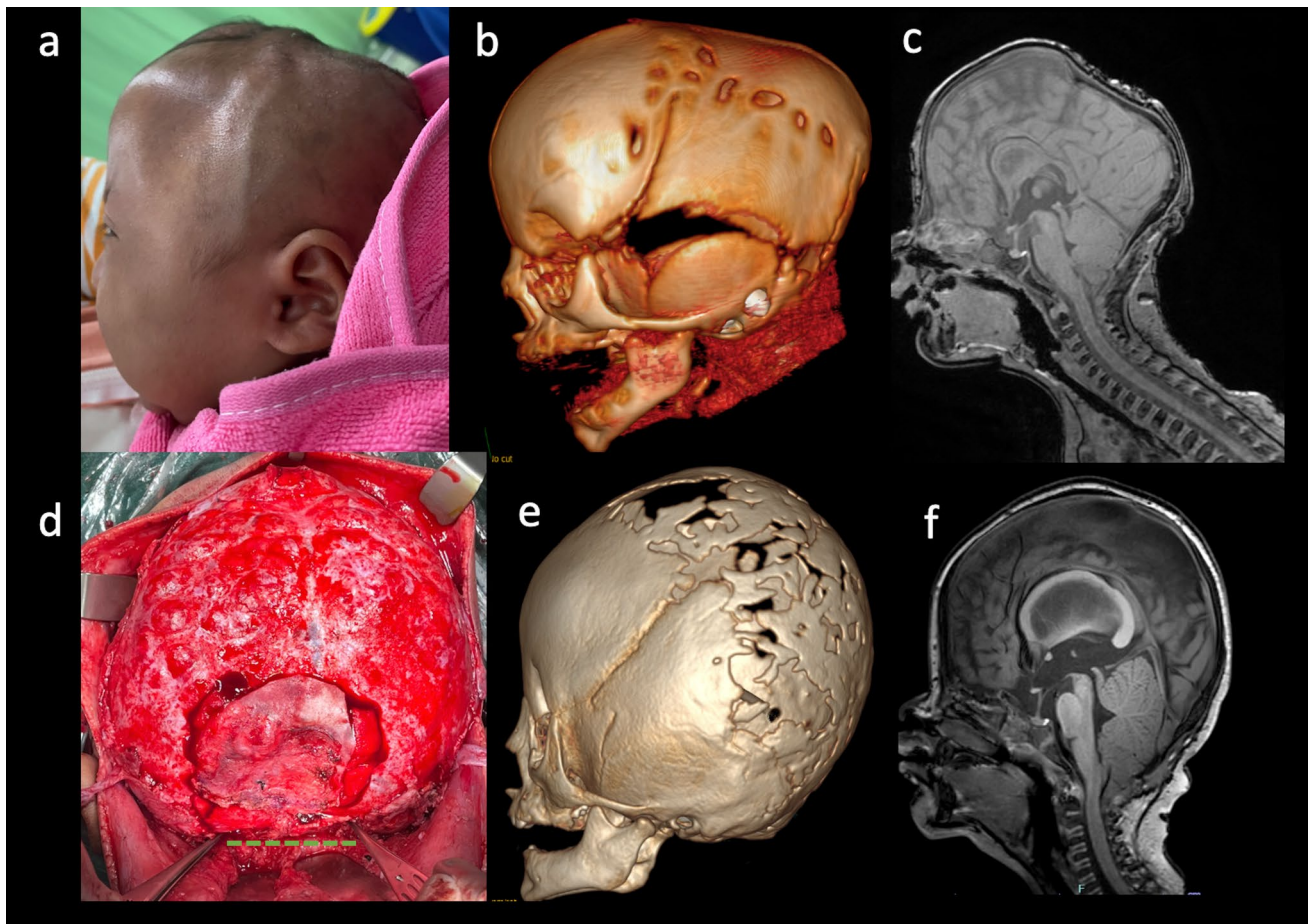
## Discussion

### Managing complex multisuture craniosynostosis

“Insanity is doing the same thing over and over and expecting different results”—Albert Einstein.

In craniofacial surgery, where our primary goals are to remove the stigma of the disease, including the relief of raised ICP, the approaches over the last four decades to manage the expression of complex multisuture craniosynostosis patients have resulted in limited, if any, advance in long-term





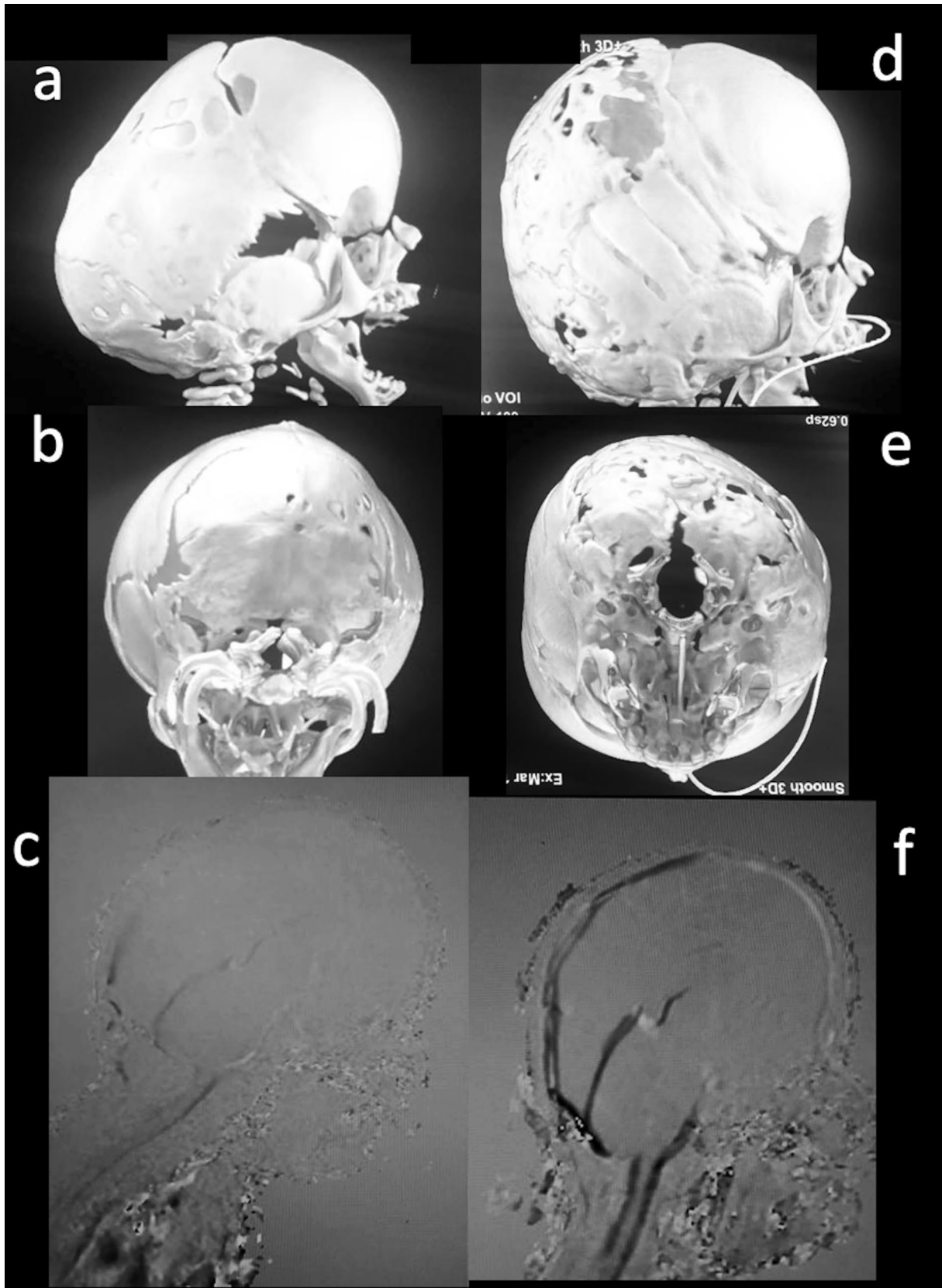
**Fig. 3** Case 2. **a** A preoperative image. **b** Preoperative 3D CT demonstrating multisuture synostosis with constricted posterior cranium. **c** Preoperative MRI showing restriction at posterior cranial fossa. **d** Intraoperative posterior craniectomy with foramen magnum decompression. The green dot line shows the area decompressing occipital

bone as for foramen magnum decompression. **e** Postoperative 3D CT and **f** postoperative MRI showing improved posterior cranial shape with remarkably bone reformation and expanded posterior cranial fossa

aesthetic or functional outcome. The strategies for releasing and expanding a stenotic calvarium, in the presence of related but poorly understood CSF flow changes, to maximise normal brain growth whilst also needing to expand constricted orbits and multi-level airway obstruction has to date been reactionary. Initial cranial vault surgery, either anterior or posterior, at 3–6 months of age or later has often been in response to the threatened complications of a tight cranium, orbit or airway. With the improvement in surgical and anaesthetic techniques, the evolution in imaging the craniofacial region and reflection on long-term outcomes from first-generation craniofacial surgical protocols, we should now attempt to be proactive and interceptive in our multidisciplinary management.

Several studies have suggested early and aggressive decompressive cranial vault procedures in cloverleaf skull deformity. Resnick et al. recommended early and aggressive decompression procedures during the first month of life

to ameliorate the raised ICP. An anterior, followed later by posterior craniectomy, was preferred due to the protrusion of the torcular herophili in the posterior fossa making the dissection difficult [8]. Goh et al. advocated an early cranial vault expansion to delay the shunt procedure, which might reverse the hydrocephalus and restrict the cranial expansion [9]. By performing shunting prior to or simultaneously with a cranial decompression procedure, risks impeding and decelerating cranial expansion during rapid brain growth, especially in the first 6 months of life [10] (Fig. 5). A larger study from a French team of 13 cloverleaf skull patients showed that 5 patients died along the course of treatment. Cardiorespiratory distress associated with airway compromise raised ICP, and Chiari I malformation was the cause of mortality. They found a relation between bilateral lambdoid stenosis and hydrocephalus and, thus, recommended doing posterior fossa decompression and shunting in patients with lambdoid synostosis [11].



**Fig. 4** Case 3. **a–c** Preoperative images show multisuture synostosis (**a**, **b**—coronal, sagittal, and lambdoid suture fusion) and cerebrospinal fluid (CSF) flow obstruction from **c** MRI. **d–f** Postoperative images at 6 weeks after posterior craniectomy with **d**, **e** cranial vault remodelling and foramen magnum decompression reveal a rounder skull shape with remarkable bone reformation posteriorly (**d**, **e**). **f** The postoperative MRI also shows an improvement in CSF flow posterior to spinal cord

### 'Vicious cycle' of raised intracranial pressure in complex multisuture craniosynostosis

The Monro–Kellie Doctrine dictates that the sum of brain volume, CSF, and blood flow is constant in a closed system. Altering any of these factors will change ICP [12]. Several elements, including cranio-cerebral disproportion, hydrocephalus, CSF outflow resistance, venous hypertension, and developing sleep apnea in craniosynostosis, play major roles in developing increased ICP in multisuture craniosynostosis (Fig. 6) [6, 13–16]. The cranio-cerebral disproportion resulting in insufficient intracranial spaces underpinning the growing brain subsequently relates to progressive increased ICP [14]. Given concomitant hydrocephalus, the pathogenesis was believed to be multifactorial and from either obstruction or malabsorption. Besides, in syndromic cases, in which multisutural fusion, both in the cranial vaults and base, results in constriction of the posterior cranial fossa. This subsequently causes the development of the Chiari I malformation and compromising venous return [6, 13, 14]. This, thus, worsens hydrocephalus [16]. The midface hypoplasia causes mechanical restriction of airflow. Moreover, when untreated Chiari I malformation exists, the brain stem herniation may worsen or exacerbate the central apnoea, resulting in hypercapnia and increased cerebral blood flow, aggravating the increased ICP.

Based on these factors, it appears that a restriction in the posterior cranial fossa is the major contributing and causative factor for obstructed CSF outflow, venous hypertension, and Chiari I malformation, all of which can significantly worsen intracranial pressure. The significantly altered posterior cranial fossa anatomy and concomitant ventriculomegaly in syndromic craniosynostosis seem to be the major causes of CSF flow disturbance. Rijken et al. revealed a significantly smaller foramen magnum surface area in Crouzon syndrome [17]. Subsequently, a study from the same group [5] noted that a reduced foramen magnum size might obstruct CSF passage around the posterior skull base, with a potential role in the development of ventriculomegaly and increased ICP. Cerebellar tonsillar herniation present in 44% of Crouzon cases is not directly linked to foramen magnum size, but changes in CSF across this area can set up a vicious cycle of firstly increased ICP leading to cerebellar tonsillar herniation, which further impairs CSF outflow. Doerga et al. demonstrated the relationship between ventriculomegaly and tonsillar herniation

that a 10% increase in ventricle size is related to a 1.6-mm increase in tonsillar herniation in Crouzon syndrome [10]. A study from the US also revealed a significantly smaller foramen magnum surface area in Crouzon and Pfeiffer syndrome [18]. Superimposed on this is the effect of obstructive sleep apnoea causing elevation of ICP, which will likely be accentuated where the foramen magnum is smaller. Releasing of a constrained posterior cranial fossa by decompression of the posterior margin of the foramen magnum seemingly allows for reducing or stopping CSF flow changes in the region. Foramen magnum decompression has been beneficial in addressing all previously stated morbidities associated with increased ICP in the first place before definite cranial vault shaping procedures. Moreover, our 3<sup>rd</sup> case clearly defined improved CSF flow on follow-up MRI.

### Cranial decompression in craniosynostosis: from front-first to back-first

The concept of craniectomy, where cranial suture fusion was thought to be associated with raised ICP, began with Lannelongue in 1890 [19, 20]. Variations on this approach followed with incremental development of anaesthetic and intensive care expertise allowing for more radical surgery performed safely at a young age. The development of distraction osteogenesis and spring-assisted expansion added to the option of conventional vault remodeling procedures. Thatikunta et al., by using the 3D software to investigate the volumetric changes, revealed an average of 13% increase in posterior cranial fossa volume with a mean distraction of 2 cm [21]. Another study from Japan revealed a 20% increased intracranial volume after 3-cm distraction [22].

Changing from the initial operation from front-to-back approach, a focus on volume expansion of the cranial cavity in the last 20 years identified that larger volume changes occur with posterior vault expansion [23–25], in contrast to fronto-orbital advancement, where there is less volume increase, more relapse and need for repeat surgery. Derderian et al. studied multisuture craniosynostosis patients and found the mean advancement of the FOA group was significantly lesser than the PVDO group, 12.5 vs. 24.8 mm. In addition, the volume changing of the PVDO was higher than FOA after adjusting the cranial growth (274 ml vs. 144 ml, respectively). However, there was no difference in terms of the volume changing per advanced distance between the 2 groups, which was around 5 ml per mm [26]. Almost all papers report posterior vault surgery at or about 6 months of age, when the bone is thicker, allowing for better application of fixation devices, and operative descriptions frequently confirm the posterior lower limit of the osteotomy to be at or about the torcula.

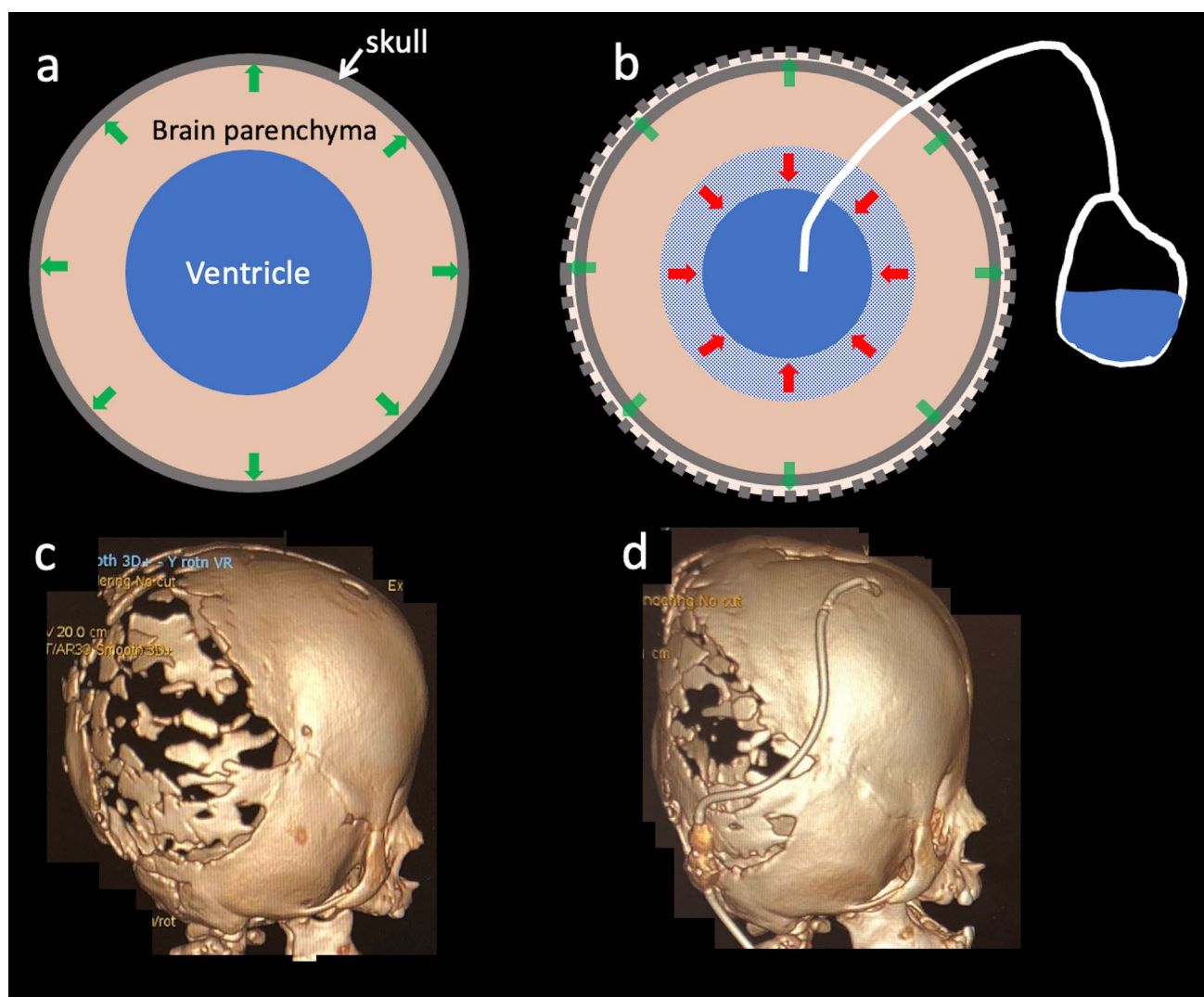
Cranial volume expansion is significant and invariable whether by distraction, springs or formal remodeling, but when performed at 6 months, it is being delayed in these



very cases where CSF flow disturbances across the cranio-cervical junction are present at birth and ongoing thereafter. Posterior vault expansion may change the CSF dynamics in the posterior cranial fossa and reduce Chiari I malformations, but in severe cases with local bony constriction or narrowing of the foramen magnum will have a limited effect. Without attention to the cranial base anomalies and expansion of the cranio-cervical junction, CSF flow disturbances may be attended by the development and progression of hydrocephalus, frequently resulting in early VP shunting. Performing cranial vault expansion and remodelling in the presence of an existing VP shunt is challenging. Inadequate shunting may result in failure to re-ossify cranial defects, whilst over-shunting can produce skull collapse or secondary

synostosis. In Case 1, the flat occiput which developed probably occurred after over-shunting (Fig. 5d). Despite this challenging clinical tightrope, some medical facilities still rely on VP shunts as a primary treatment option to enable delayed timing of cranial vault expansion procedures [27].

The approach reported herein on three infants challenges this conventional approach. Early decompressive posterior vault surgery at age < 1 month was utilised in all cases where there is a rapid progression of the severe clinical expression of multi-suture synostosis—progressive proptosis, tight fontanelle, and impending or actual CSF flow changes. Extensive craniectomy of the posterior cranial vault extending from the anterior fontanelle back to and including the occiput, with extension to the foramen magnum, was safely performed with

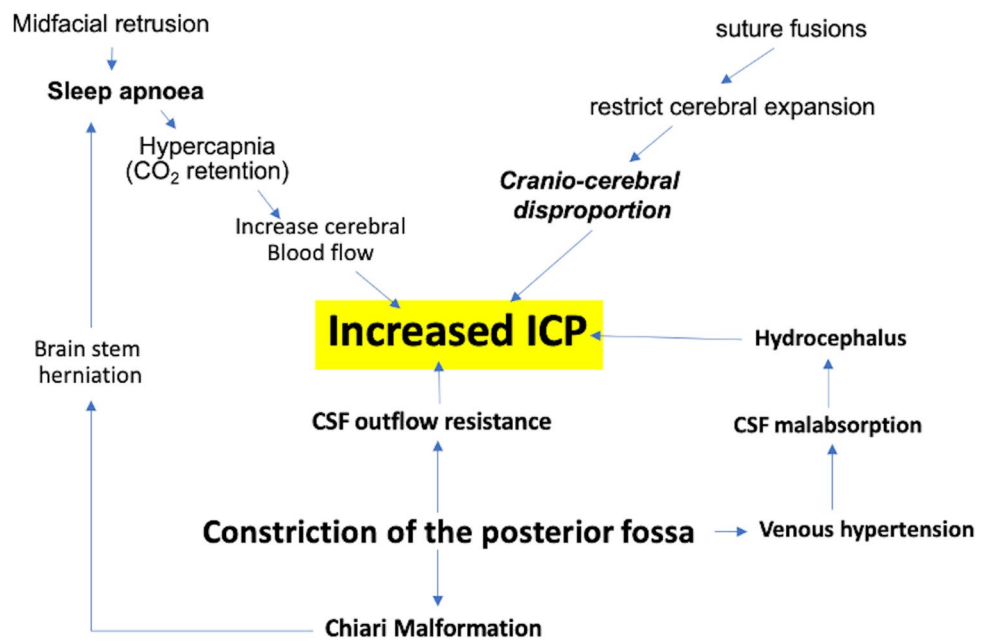


**Fig. 5** Illustrating shunt drainage of CSF (**b**) is able to reduce skull growth during the rapid cranial vault expansion (**a**). The green arrows demonstrate normal brain expansion shaping cranial vaults. The red arrows (ventricle shrinkage) show a deceleration of brain growth after

CSF shunting which will limit the skull shape and size as it should be. Case 1's 3D CT scans (**c** before VP shunt and **d** after VP shunt) show the flat occiput with a suggestive over-shunting



**Fig. 6** Domino effect of increased intracranial pressure in complex multisuture craniosynostosis



a minimal requirement for blood replacement (<50 ml blood in all cases). Lateral tarsorrhaphy was performed before the cranial vault surgery, securing the globes from short- and medium-term exposure. Decompression of the foramen magnum permits alleviation of the Chiari I malformation (Cases 1 and 2) and improves CSF flow across this region (Case 3). Whilst Case 1 went on to require a VP shunt several months after vault surgery, by the time of the FOA at 18 months of age, the shunt was no longer required. As for Case 2, the VP shunt procedure was performed 2 months after the posterior cranial decompression procedure, as the hydrocephalus was remarkable at the temporal horns. This might be due to the associated *SIX3* mutation, which was previously reported in holoprosencephaly 2 (<https://www.omim.org/entry/157170>). However, this has to be confirmed as the pathogenic variant by further trio exome analysis. Early intervention provides the opportunity to prevent the deterioration of CSF flow obstruction across the cranial base, rather than waiting 3–6 months or more in conventional protocols, by which time these changes are established.

Such extensive posterior vault craniectomy has allowed for significant calvarial volume increase and shape change within 4–6 weeks despite the speed with which bone reforms over the regions where the bone is removed. MRI scan imaging in Case 3 shows improved CSF flow through the foramen magnum, coincident with a demonstrable increase in the anteroposterior dimensions of the foramen magnum post-surgically. The post-operative CT scans in these cases confirm extensive bony regrowth, over a wide area, in accordance with our previous studies on sagittal synostosis [28], but with the maintenance of an increase in foramen magnum size.

The immediate management of the severe cloverleaf infant remains one of our greatest challenges. Conventional approaches, waiting 3–6 months or more, have largely resulted in poor functional and aesthetic outcomes. A primary focus not on the cranial vault but on the known changes in the cranial base seems the logical next step. Early tarsorrhaphy to manage the proptosis secondary to the foreshortened orbits protects the globes for the medium term. Management of the upper airway obstruction by uvulopalatoplasty, palatal split, and later adenotonsillectomy [2] similarly buys time and avoids early midface surgery. Lastly, very early decompression of the abnormal foramen magnum and wide craniectomy of the posterior cranial vault allows for improved CSF flow across the cranio-cervical junction and cranial vault volume increase providing the potential to reverse the early downward spiral of multiple cranial vaults and VP shunt procedures that attended the first-generation management of the cloverleaf skull deformity and resulted in such poor long-term outcomes.

## Conclusions

Cloverleaf skull deformity or complex multisuture craniosynostosis, particularly the cranial base suture fusion, plays the main pathology resulting in significant subsequent deformities. Posterior cranial decompression, down to and including the foramen magnum, should be considered the initial decompressive procedure to delay the definite orbitofrontal advancement. Hydrocephalus management may be considerably altered in each patient depending on the

persistent dilatation of the ventricles after cranial expansion procedures. One of the challenges is over-shunting, which results in secondary synostosis, slit-ventricle syndrome, and limiting brain growth. Multidisciplinary team care is mandated in syndromic craniosynostosis management.

**Author contributions** Mark H. Moore: Conceptualisation, Data Curation, Writing - Original Draft, Writing - Review & Editing, Project administration Sarut Chaisrisawadisuk: Conceptualisation, Data Curation, Writing - Original Draft, Writing - Review & Editing, Visualisation Inthira Khampalikit: Conceptualisation, Data Curation, Writing - Review & Editing Xenia Doorenbosch: Conceptualisation, Data Curation, Writing - Review & Editing Alistair Jukes: Conceptualisation, Data Curation, Writing - Review & Editing Cindy J. Molloy: Conceptualisation, Data Curation, Writing - Review & Editing, Project administration

**Availability of data and material** This is not applicable.

**Code availability** This is not applicable.

## Declarations

**Ethics approval** This is not applicable.

**Informed consent and patient details** The patients' parents provided written informed consent for the publication and the use of the images.

**Conflict of interest** The authors report no possible conflicts of interest.

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