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Case Report

Neurologist and Neurosurgeon: A 'Sine Qua Non' in Paediatric Intracranial Pathology Management

Ammar Natalwala, Pasquale Gallo, Drahoslav Sokol, Jothy Kandasamy, Krishnaraya Kamath Tallur, Ailsa McLennan, Jayakara Shetty, Richard Chin, Alex Baxter, Paul Eunson, Kenneth McWilliam, Chandrasekaran Kaliaperumal*

Department of Paediatric Neurosurgery, Royal Hospital for Children and Young People, Edinburgh, Scotland, UNITED KINGDOM.

ABSTRACT
Specialist care providers often treat complex, life-threatening intracranial paediatric pathologies and team-based clinical decisions by neurologists and neurosurgeons are vital for good clinical outcomes. We describe six scenarios highlighting multi-disciplinary team-based management of challenging paediatric conditions which provided successful outcomes. This work may aide future management of similar cases.

Keywords: Challenging paediatric cases, Neurology, Neurosurgery, Team-Based, Multi-Disciplinary.

Correspondence:
Mr. Chandrasekaran Kaliaperumal
FRCSEd (Neuro.Surg), Consultant Paediatric and Adult Neurosurgeon, Department of Paediatric Neurosurgery, Royal Hospital for Children and Young People, Edinburgh, Scotland, UNITED KINGDOM.
Email: ckaliaperumal@gmail.com

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INTRODUCTION

Paediatric neurological conditions such as stroke and infection require prompt diagnosis and team-based decisions on management. In some cases, this can be challenging as these conditions are often under recognised. For example, Rafay and colleagues report a median pre-hospital delay for arterial stroke in children to be 1.7 hr from symptoms to presentation, whereas this delay was 12.7 hr in hospital from presentation to diagnosis.[1] Diagnosis can be difficult as children can present with a myriad of symptoms and thus a wide range of differential diagnoses to confirm or refute. Thus, often empirical treatments such as antibiotics are instigated rapidly where appropriate to allow time for investigations and confirmation of diagnosis. Expertise from neurologists, neurosurgeons, intensivists and other healthcare teams is essential to optimise management. This speeds diagnosis and treatment and agreement or consensus can be made on difficult decision where often there is paucity of evidence in the literature, particularly for the paediatric population. Neurological conditions such as these can lead to devastating and life-long poor outcomes if treatment is delayed or misguided and so a sense of urgency and accuracy in treatment is needed per case.

In our series, we describe our experience of six challenging cases where a team-centred approach was applied to enable us to achieve good clinical outcomes. We describe a range of intra-cranial pathologies and challenges highlighted on a case-by-case basis.

Case 1. Meningoencephalitis and cerebral oedema

Presentation

An 11-year-old boy was admitted in July 2016 with signs of infection including pyrexia, headache and vomiting. He was empirically treated for meningitis with Ceftriaxone and Acyclovir but deteriorated neurologically with a fluctuating consciousness on the ward.

Treatment

He was subsequently diagnosed and treated for encephalitis and cerebral oedema (Figure 1A). Following his stay on the paediatric intensive care unit he was extubated and transferred to the neurology ward, however, there were some concerns about deterioration in his vision. Fundoscopy revealed papilloedema (and later optic atrophy) and he was also noted to have a sixth nerve palsy. The raised Intracranial Pressure (ICP) was believed to be secondary to encephalitis and possible venous sinus thrombosis although not confirmed on CT venogram. In August 2016, an attempt was made to insert a right frontal Ventricular Access Device (VAD) to treat the raised ICP which was thought to be contributing to visual loss. Due to dural haemorrhage from raised intracranial pressure, the procedure was abandoned and re-attempted two weeks later where a right frontal VAD was successfully inserted. In September 2016, a left sided Ventriculo-Peritoneal (VP) shunt was inserted and later that month an infected baclofen pump was removed (possibly the

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source of the initial infection). The child did well post-operatively but in December 2016 he re-presented and required burr hole drainage of a left subdural haematoma and revision of the left VP shunt valve. He again re-presented in January 2017 with 3 days of vomiting, abdominal pain and loose stool. A collection was seen around distal shunt catheter and the infected shunt was removed (*Staphylococcus aureus* was the causative agent). He was commenced on intravenous antibiotics and recovered well.

Outcome

Post-op he has ongoing language and visual impairment but did not require re-shunting. His language continues to improve and he is mobilising independently.

Case 2. Ischaemic brain infarct with haemorrhagic transformation in the background of Infective endocarditis and a large intramural vegetation

Presentation

A 9-year-old girl initially presented to the emergency department in February 2017 with altered consciousness (GCS 9; E2 V4 M3). She had been unwell for seven days with pyrexia and a cough with occasional vomiting. She developed a headache and woke during the night distressed and agitated. On arrival to the emergency department, she was very agitated with impaired capillary refill time and pyrexia. She was given a 10 mg/kg fluid bolus and started on Ceftriaxone, Acyclovir and dexamethasone for presumed meningoencephalitis and then intubated for transfer to the Paediatric Intensive Care Unit (PICU). The CT scan prior to transfer was normal (Figure 1B).

Treatment

In PICU she was noted to have a large mobile vegetation on her mitral valve on echocardiogram. Her initial blood cultures grew Streptococcus pneumoniae and here antibiotics were refined accordingly to Cefotaxime and Rifampicin. On advice of the cardiac surgeons, she was started on anticoagulation with enoxaparin. Unfortunately, she deteriorated and an MRI the following day showed a large right frontal haemorrhage and anticoagulation was stopped. This haemorrhage showed increasing mass effect and oedema (Figure 1B) and she underwent emergency craniotomy and evacuation.

Outcome

Post-surgery she had some left sided facial twitching that was treated with phenytoin which has now been stopped. She had 6 weeks of intravenous antibiotics then converted to an oral regimen. She has mild weakness of her left side and mild dysphasia. These have gradually improved with neurorehabilitation which will continue in the community.

Case 3. Hemispheric stroke secondary to carotid dissection

Presentation

A right-handed boy, doing well previously in school, presented to the emergency department in December 2015 with a sudden onset right-sided weakness and neglect; GCS was E4 V1 M5.

Treatment

He was treated empirically with antibiotics and antivirals as suspected to have meningo-encephalitis. An MRI scan showed a left Middle Cerebral Artery (MCA) stroke due to Internal Carotid Artery (ICA) dissection (Figure 1C). He developed dystonic right hemiplegia and aphasia. His echocardiogram showed nothing untoward and the serum thrombophilia and vasculitis screens were also negative. Later it became apparent that the injury was likely due to an incident in a prior rugby match. Due to the fluctuating GCS, the question was asked if he should undergo decompressive craniectomy. It was considered that a conservative approach would be best and since then he has made a partial recovery.

Outcome

He can walk but ability to walk long distances at speed is impaired. He has some speech impairment with difficulty being able to find appropriate words quickly enough in everyday conversation. His sixth nerve palsy at the time of the stroke has fully resolved.

Case 4. Brain abscess and associated infarct

Presentation

A 15-year-old boy presented in March 2016 with six days of severe headache, vomiting, photophobia, rash and a temperature of 39.6°C. His GCS was 15 with no focal neurological deficit. His GCS dropped to 11 and a CT scan confirmed a subdural collection (Figure 1D). Contrast images suggested this was a subdural empyema with associated parasinusitis. The frontal and maxillary sinuses were affected and the likely cause of infection into the subdural space.

Treatment

An emergency craniotomy and evacuation of empyema was performed and drainage of the frontal and maxillary sinuses done endoscopically. He had a right-sided weakness post-operatively and a repeat CT showed raised intracranial pressure and frontal ischaemia. Therefore, an emergency decompressive craniectomy and further evacuation of pus was done. He had a posterior craniotomy and evacuation of a left posterior empyema and later had a cranioplasty to replace bone flap three months afterwards.

Outcome

One-year post-discharge from hospital his seizures are well controlled, he is independently mobile over short distances and has an improving right-sided weakness.

Case 5. Haemorrhagic stroke secondary to ruptured AVM

Presentation

An 8-year-old girl presented to the emergency department in February 2017 with a 2-week history of worsening headache and vomiting. She was drowsy with a fluctuating GCS and was started on intravenous Cefotaxime and Acyclovir empirically for possible encephalitis. The initial CT scan showed a left frontal haemorrhage with 1cm of midline shift (Figure 1E).

Treatment

She developed fixed and dilated pupils and was taken to theatre for an emergency craniotomy for clot evacuation. Intra-operatively she had a seizure and was given a loading dose and regular Levetiracetam. She was extubated 3 days later and was noted to have a residual right-sided weakness and delayed MRI and CT angiogram showed an underlying (micro) Arteriovenous Malformation (AVM) as the cause of this bleed. It was located at the left paramedian frontal pole and the 1cm ill-defined diffuse nidus lay to the left of the midline adjacent to the Anterior Cerebral Artery (ACA) bifurcation into the pericallosal and callosomarginal arteries. There were numerous perforating type nidal feeders from the ACA. Both superficial and deep venous drainage to the AVM was seen to the superior sagittal sinus and internal cerebral veins respectively. The AVM was not suitable for endovascular treatment due to the size of the lesion (Spetzler Martin Grade II; less than 3cm in size, located in a non-eloquent region with superficial and deep venous drainage) and she was not suitable for SRS because of age thus surgical excision was the best option. In June 2017, she had a neuronavigation-assisted redo left frontal craniotomy and complete excision of the frontal AVM.

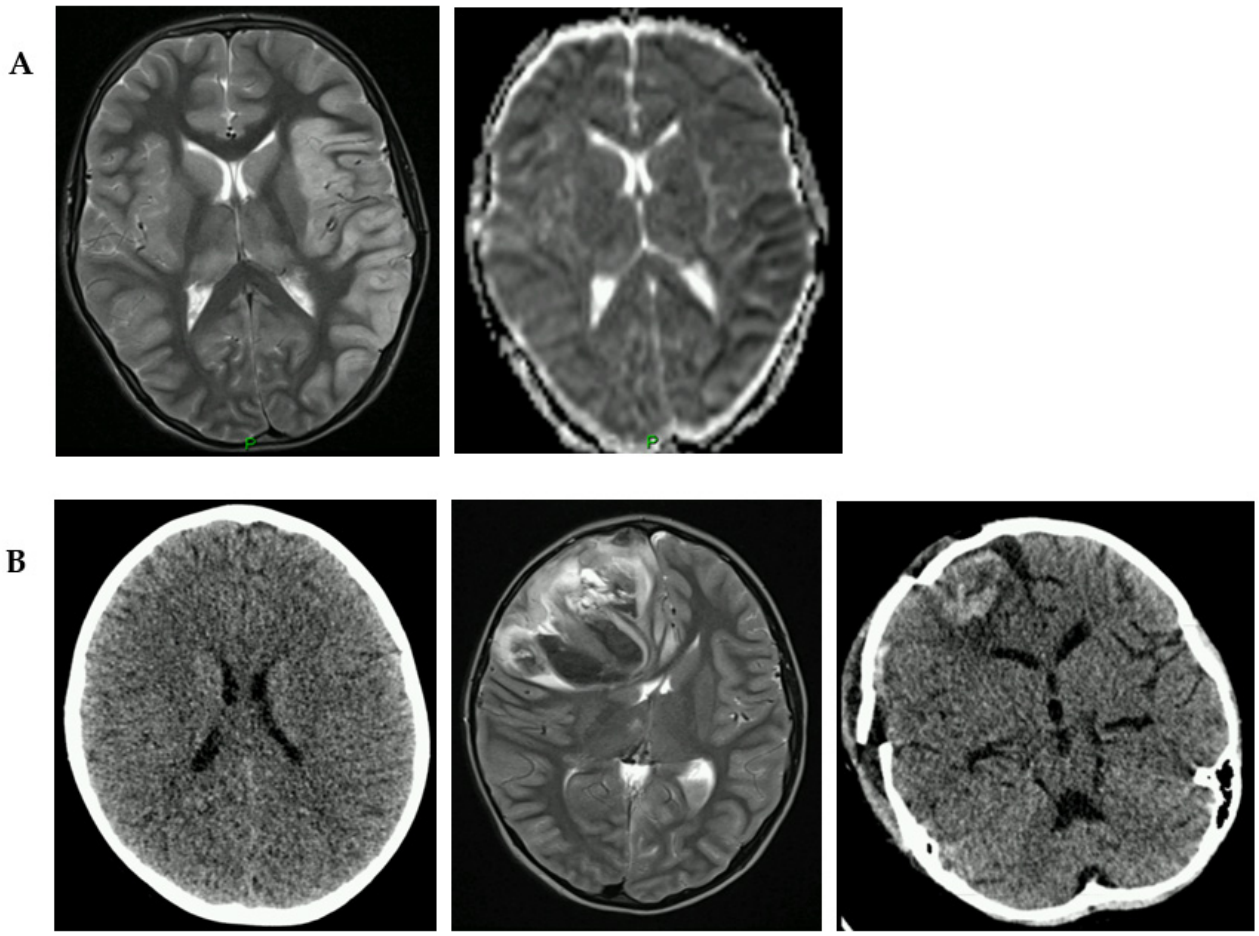
Outcome

Post-operatively she has no neurological deficits and her hemiparesis had completely resolved.

Case 6. Growing skull fracture

Presentation

A 7-month-old child was born at 35+4 weeks in October 2016 by normal (un-instrument assisted) vaginal delivery. He was noted



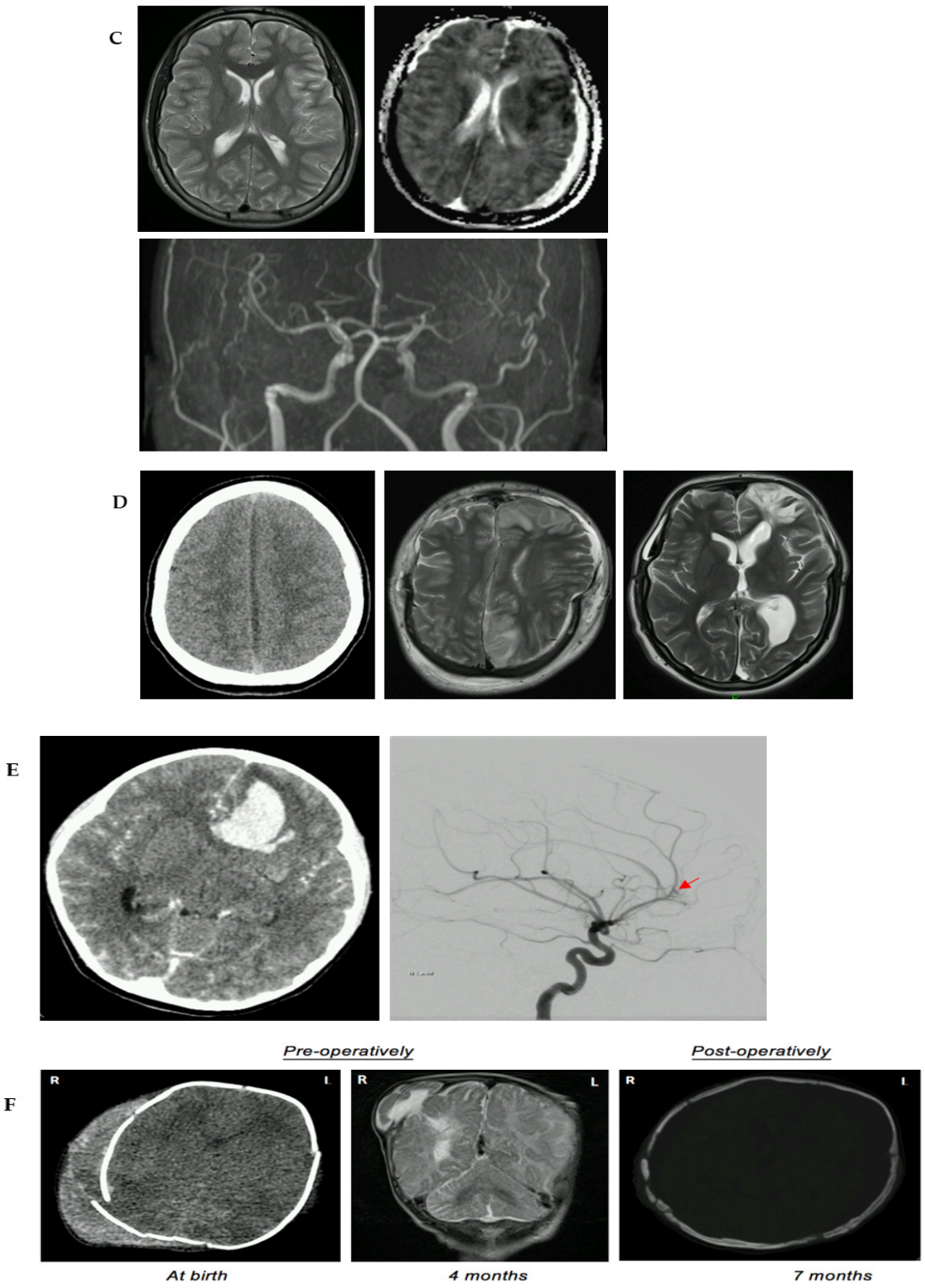


Figure 1: A) T2-weighted (left) and diffusion-weighted (right; DWI) axial MRI images showing areas of cerebral oedema and diffusion restriction mainly in the left hemisphere. B) Initial CT scan on the left which was normal, an MRI in the centre following a decline in GCS showing a right frontal haemorrhage. Given the case this was considered a haemorrhagic transformation from an infarct due to septic emboli. On the right-hand side is the post-operative CT showing good decompression of the clot. C) T2-weighted and DWI axial MRI images (left and right respectively) showing a right-sided MCA territory infarct (restriction on DWI). The MR angiogram below shows evidence of ICA dissection. D) Initial CT (left) showing a parafalcine collection and T2-weighted MRI following craniectomy and washout (centre). The right-hand side image is the most recent MRI following cranioplasty. E) Initial CT axial image for Case 5 showing a left frontal intra-parenchymal haemorrhage and midline shift requiring emergency craniotomy and evacuation. The digital subtraction angiogram on the right shows the diffuse micro AVM (red arrow) where the ACA bifurcates into the pericallosal and callosomarginal arteries. F) Initial CT axial image on the day of birth for Case 6 showing a complex right parieto-occiptal fracture and associated subgaleal collection. The centre image (coronal T2-weighted MRI) shows herniation of the parietal lobe through the defect and associated ischaemia. The post-operative CT shows bony windows following repair of the dura and cranium.

Table 1: Table summarising each case, including the challenges faced, treatment given and outcome achieved working in a team.

Case	Challenge	Treatment	Outcome
1	Decompressive craniectomy vs medical therapy alone.	Antibiotics. No surgical decompression. Hydrocephalus treated with VAD and shunt. Infected baclofen pump removed.	Ongoing language and visual problems (improving). Did not require re-shunting. Mobilising independently.
2	Anticoagulation for endocarditis vs haemorrhagic stroke. Craniotomy for clot evacuation vs medical therapy.	Anticoagulation stopped when ICH diagnosed. Craniotomy and clot evacuation performed. Antibiotics.	Facial twitching stopped with phenytoin. Mild left facial weakness and dysphasia.
3	Decompressive craniectomy vs medical therapy alone.	No surgery. Medical management of carotid dissection and stroke.	Can walk short distances. Residual speech impairment. Sixth nerve palsy fully resolved.
4	Management of subdural empyema including recurrence (craniotomy vs craniectomy).	Initial craniotomy (and sinus washout) then craniectomy upon recurrence and cerebral oedema.	One-year post-discharge from hospital seizures are well controlled. Independently mobile over short distances. Improving right-sided weakness.
5	Initial clot management. When to re-image. Management of residual AVM.	Emergency craniotomy and clot evacuation. Delayed angiogram. Subsequent surgical resection of AVM.	No neurological deficits and hemiparesis completely resolved.
6	When to repair growing fracture. Treatment of CSF leak.	Repair once child gained body weight at 4 months. CSF repaired promptly.	CSF leak has not recurred.

to have a large scalp swelling in the post-natal ward. A CT scan showed extensive occipital and right parietal comminuted and displaced skull fractures and a large subgaleal haemorrhage (Figure 1F). There was no penetration of the dura or skin. Neurologically he was moving all four limbs, reflexes were normal and pupils equal and reactive to light. His MRI scan further delineated partial herniation of the right parietal lobe with ischaemic change and parenchymal haemorrhage. Further infarct was seen in right thalamus and basal ganglia and the mesial temporal lobe. Given the body weight of the neonate, neurosurgical intervention was not deemed appropriate at this stage. The possible diagnosis of osteogenesis imperfecta was raised and a skeletal survey did not reveal other bony anomalies.

Treatment

In February 2017, he had an elective repair of the right parietal growing fracture. Duroplasty and cranial reconstruction was performed (Figure 1F). Post-operatively he did well and continued to achieve the appropriate developmental milestones. However, later that month he was taken back to theatre to evacuate a subgaleal CSF collection and repair the point of CSF leak within the dura.

Outcome

Post-operatively he is doing well, the CSF leak has not recurred and his developmental milestones will be closely monitored.

DISCUSSION

In this report, we describe six complex paediatric cases which highlight the fact that harmonised teamwork amongst neurologists, neurosurgeons and other specialists is key to achieve good clinical outcomes for these children. We allude to some of the difficult management decisions in each of these cases with support from published literature.

The challenge in the first case was deciding upon the initial management of raised ICP secondary to intracranial infection and subsequent cerebral oedema. Although decompressive craniectomy has been described well in the adult setting there is a paucity of its description and outcomes in children in the non-traumatic setting.[2] Aghakhani *et al.* investigated seven children with non-traumatic refractory high ICP undergoing decompressive craniectomy. They found that 6 patients survived and six months after discharge, 5 had a good or moderate recovery with Glasgow Outcome scores of three to five. Of these, four had

infection-related cerebral oedema.[3] In our case, we managed ICP medically in the intensive care setting and once the child's GCS improved, it became apparent that there was visual impairment due to ongoing raised ICP. As per local practice, a ventricular access device (Ommaya reservoir) and a ventriculoperitoneal shunt were inserted. Despite the shunt-related complications of over-drainage and infection, the child made a good recovery without additional morbidity due to craniectomy.

In the paediatric population, ischemic stroke has a reported 30-day mortality of 12.3% and over 50% of survivors acquire neurological deficits.[2] 10% of adults develop malignant infarction following ischaemic stroke with a mortality of up to 80%. Case 2 and 3 were interesting because these children have ischaemic stroke with the former due to septic emboli (transformed into a haemorrhagic stroke) and the latter due to a large left MCA stroke due to an ICA dissection thought to be due to a rugby injury. The clinical challenges included decisions on whether a decompressive craniectomy (limited evidence as described above) would be required given the fluctuating GCS and if anticoagulation is appropriate. In a population-based study by Rahme *et al.*, 12.5% of all paediatric ischemic strokes over two 1-year periods were malignant MCA infarctions.[4] Malignant MCA features often develop within 72 hr of presentation. Shah *et al.* describe three cases of malignant MCA infarct in the paediatric group who underwent surgery and achieve modified Rankin Scores between two and 3 and two to three years follow up.[5] In our case, after a multidisciplinary team discussion the conservative approach was taken. Paediatric stroke guidelines from the Royal College of Physicians guidelines, the American College of Chest Physicians and the American Heart Association (AHA) guidelines were followed.[6] The child did well without surgery and made a good partial recovery.

Case 4 was an intracranial infection like case 1, however, the child had a subdural empyema which was drained twice. Ultimately, a decompressive craniectomy had to be done due to cerebral oedema and refractory intracranial pressure. Cranial subdural empyemas account for 15 to 20% of intracranial infections and a report by Mohindra, highlights the importance of early diagnosis and treatment in these cases.[7] Patel *et al.* identified 27 children with associated extradural, subdural or parenchymal abscesses. 24 required neurosurgical drainage and an initial conservative neurosurgical approach failed in 50% of cases and was associated with longer length of stay. A third of children had significant morbidity at six months, which included cognitive and behavioural problems, residual hemiparesis and expressive dysphasia with no deaths.[8] In our case, we managed co-ordinated with the neurology team to identify and intervene early with surgery to wash out infection and decompress early giving a good clinical outcome.

AVM is the most common congenital vascular pathology causing intra-parenchymal haemorrhage in children. Despite complete surgical excision these AVMs can recur several years later.[9] A study by Kim *et al.*, reviewed 7485 paediatric inpatients with vascular anomalies and found those with AVM had the highest rate of in-hospital length of stay and mortality, compared to those with haemangiomas and other malformations.[10] Trials such as STICH demonstrated in adult patients with spontaneous supratentorial intracerebral haemorrhage, where there was clinical equipoise, that emergent surgical evacuation within 72 hr of bleeding onset did not improve patient outcomes versus best medical management.[6] However, in case 5 given the presenting clinical picture there was no doubt that the mass effect needed to be alleviated surgically and once the haemorrhage had resolved the underlying micro AVM was demonstrated on delayed imaging. Due to the diffuse and small nature of the lesion and its location in a relatively accessible location it was deemed surgical excision versus endovascular was required. SRS would have been another option but this treatment takes months to diminish the AVM and there is a risk of re-bleeding during that time period.

In the case of the child with the growing skull fracture, these fractures often have a traumatic cause and there are few case reports of these fractures occurring in the intra-uterine stage. Examples of this are when the mother has received blunt or sharp trauma to the abdomen prior to delivery.[11] Growing skull fractures in general usually occur during the first three years of the life and almost never after 8 years of life. The incidence reported is only 0.05 to 0.1%.[12] It was unknown as to what caused the growing skull fractures in our case and we hypothesised that head trauma during delivery against the sacral promontory. Risks of cerebral herniation, seizures and CSF leak lead to the decision for surgical repair of the bony defect. The challenge was timing of surgery in the first instance given the body weight was too low when the pathology was first detected. A decision was made to operate when the child was 4 months of age. He did well but required a second operation to repair a CSF leak. A team decision for initial conservative management and timing of surgery helped to achieve a good outcome.

It is suggested that neuroplasticity, compensatory circulation, and lack of systemic comorbidities in children may contribute to a better clinical outcome.[2] These 6 cases (summarised in Table 1) describe the nuances involved in the management of paediatric intracranial conditions emphasizing the need for early communication and teamwork. Communication not only involves the neurologist and neurosurgeons but also keeping the child's family needs and wishes in mind to work as a team. The input from other medical specialties is also important. The same point that we highlight can be extrapolated and can be applicable to adult population as well.

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CONFLICT OF INTEREST

The authors declare no conflict of interest.

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