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Case Report

Staged Repair of Complete Sternal Cleft and Interrupted Aortic Arch Associated With PHACES Syndrome in a Very Low Birth Weight Infant

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Posterior fossa malformations, hemangiomas, arterial anomalies, cardiac defects, eye abnormalities, and sternal malformations' (PHACES) syndrome comprises various defects that require a concrete therapeutic plan. Herein, we report a case of a very low birth weight infant with PHACES syndrome presenting complete sternal cleft, interrupted aortic arch with ventricular septal defect, and absence of communicating cerebral arteries. Due to the infant's very low birth weight, we planned staged surgery for this complex disease. First, bilateral pulmonary artery banding was performed to improve unstable hemodynamics. Then, after sufficient body weight was obtained, aortic arch and intracardiac repair was done. However, when the tracheal tube was removed 2 weeks after surgery, the patient could not breathe well due to paradoxical chest movement related to complete sternal cleft. Consequently, to improve chest wall compliance, the upper sternum was constructed with dislocated ribs. After these

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treatments, the patient gained adequate weight and was in stable condition. Staged surgical treatment of sternal cleft and congenital heart defects is a potential therapeutic option for PHACES syndrome, particularly in very low birth weight infants with complex cardiovascular disease.

Key words: Neonatal surgery – Sternal cleft – Congenital heart disease

Posterior fossa malformations, hemangiomas, arterial anomalies, cardiac defects, eye abnormalities, and sternal malformations' (PHACES) syndrome was first proposed by Frieden *et al*[1] in 1996. They defined a syndrome comprised of: (1) posterior fossa malformations, (2) hemangiomas, (3) arterial anomalies, (4) coarctation of the aorta and cardiac defects, (5) eye abnormalities, and (6) sternal malformations or malformations of the abdominal midline.[1] The patient had some hemangiomas; arterial anomalies (hypoplasia of the right anterior cerebral artery, the right anterior communicating artery and the left posterior communicating artery); cardiac defects (interrupted aortic arch with a ventricular septal defect); and a complete sternal cleft. Although the etiology of PHACES remains unknown, some authors have reported that complications associated with the syndrome, such as complete sternal cleft and hypoplasia of the circle of Willis, can be problematic when total correction is required.[2,3] Herein, we report the first case of a very low birth weight infant with PHACES syndrome presenting complete sternal cleft and complex congenital heart defects who was treated by staged surgery. Hence, we introduce a potential strategy to manage this syndrome, particularly in very low birth weight infants with complex cardiovascular disease.

Case

The patient was born at 36 weeks of gestation weighing 1370 g. She presented extracardiac malformations such as hemangioma and complete sternal cleft, which are identical to the diagnostic criteria of PHACES syndrome (Fig. 1). In detail, there were hemangiomas in 4 places: the nose, the upper lip, the lower lip, and the posterior head. These hemangiomas were diagnosed with their appearance by skilled plastic surgeons since they had grown rapidly and regressed spontaneously. Additionally, cardiovascular anomalies including type B interrupted aortic arch (IAA), ventricular septal defect (VSD), patent ductus arteriosus (PDA), and aberrant right subclavian artery were confirmed on echocardiography and cardiac computed tomography (Fig. 2). Endotracheal intubation was required to control heart failure due to the cardiac malformations, while a gastric tube also was inserted through the nose.

Considering the patient's very low body weight, we decided to perform bilateral pulmonary artery banding though a median sternotomy prior to total repair of the cardiovascular and extracardiac deformities. She was observed in the neonatal intensive care unit until she gained sufficient body weight to undergo the correction. Meanwhile, considering that PHACES syndrome can present intracranial artery malformations, as in our case (Fig. 3), we considered that she also should undergo cannulation of both carotid arteries. After sufficient body weight was obtained, the patient underwent intracardiac surgery comprising IAA repair, VSD closure, and PDA closure though a median sternotomy. After dissection of the heart and vessels, the right carotid artery was partially clamped and a 3.5-mm tube (Gore-Tex, W. L. Gore & Associates, Newark, DE) was anastomosed as a blood source to the brain from the cardiopulmonary bypass. After bicaval cannulation to drain the venous blood, the descending aorta was cannulated to deliver oxygenated blood to the inferior body. After establishment of cardiopulmonary bypass, both the descending aorta and the aortic arch between the right and left carotid arteries were clamped. After the aortic arch was incised, a cannula was inserted in the left common carotid artery to send oxygenated blood from the cardiopulmonary bypass. Then, the aortic arch was reconstructed with extended direct end-to-end anastomosis, followed by aortic cross-clamping to repair the intracardiac anomaly. The VSD was at the perimembranous portion, and was closed with 5 pairs of 5-0 mattress Ethibond sutures. After declamping of the aorta, heart motion recovered satisfactorily. After the heart was weaned from the cardiopulmonary bypass, the chest skin only was closed by a skilled plastic surgeon due to the lacking sternum associated with PHACES syndrome. The

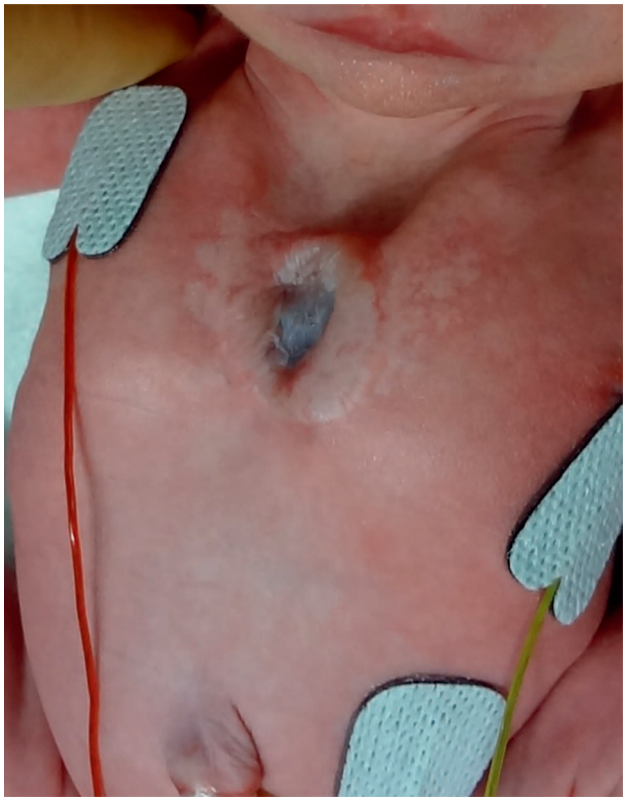


Fig. 1 Photograph, showing complete sternal cleft in a very low birth weight infant.

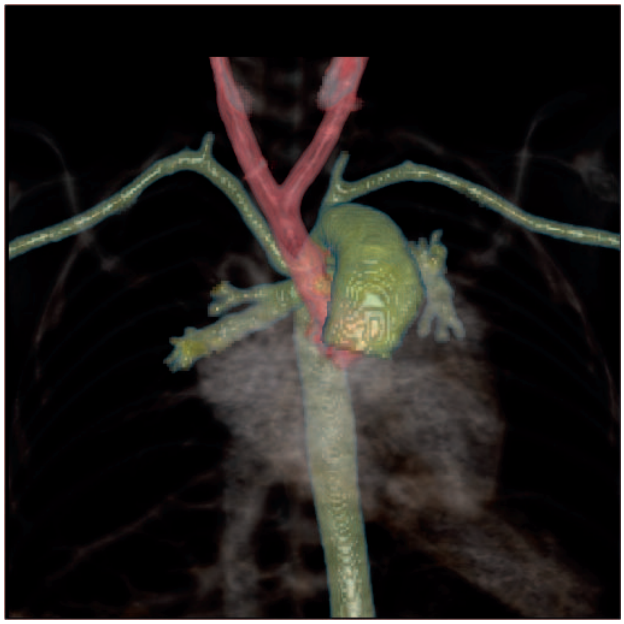


Fig. 2 Three-dimensional cardiac computed tomographic image, showing interruption of the aorta (type B) and an aberrant right subclavian artery.

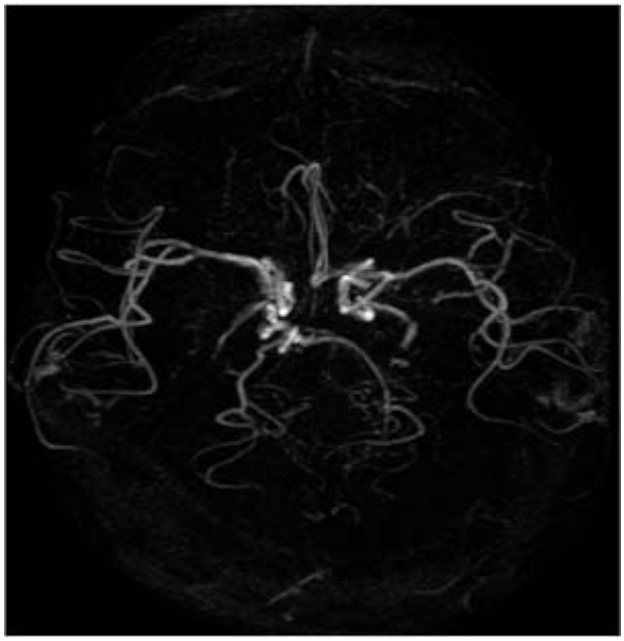
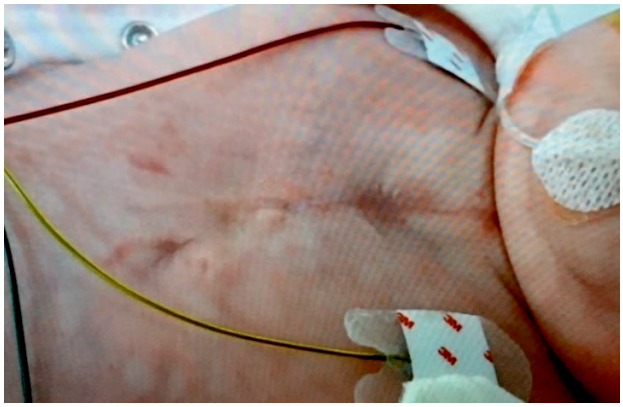


Fig. 3 Magnetic resonance angiogram, showing the circle of Willis with absence of communicating arteries.

patient was transferred to the intensive care unit in stable condition.

At 2 weeks after surgery, removal of the tracheal tube was planned; however, effective breathing could not be maintained due to paradoxical chest movement related to complete sternal cleft. This paradoxical movement not only generated respiratory failure but also had a considerable effect on circulatory instability due to compression of the mediastinum including the aorta (Fig. 4). Consequently, reintubation was required immediately. As a next step, complete sternal cleft repair was planned, after which, the patient was to be weaned from the ventilator. At 5 months of age, she underwent sternal construction with dislocation of the right sixth and seventh rims via a median sternotomy (Fig. 5). At the same time, nasal plasty was performed to remove obstruction of the left nasal cavity caused by inflammation associated with the hemangioma. After surgery, the paradoxical chest motion disappeared, adequate chest wall compliance and protection from aortic compression were achieved, and the patient was in stable condition. As a result of surgery, she was extubated without difficulty. Although she was intubated during the 5-month period, any iatrogenic hemangiomas and granulomas did not present in the airway.

(a)



(b)

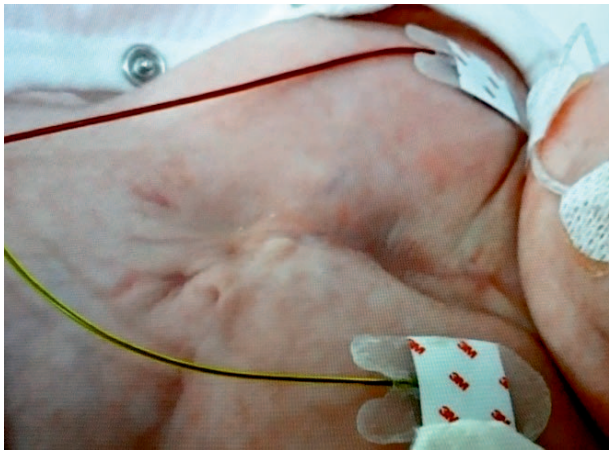


Fig. 4 Photographs, showing paradoxical chest movement causing compression of the aorta. (a) Exhalation phase. (b) Inhalation phase.

Discussion

PHACES syndrome comprises various defects that require a concrete therapeutic plan. Here, we reported a case of a very low birth weight infant with PHACES syndrome presenting complete sternal cleft, IAA with VSD, and the hypoplasia of the right anterior cerebral artery, the right anterior communicating artery and the left posterior communicating artery, and the left posterior communicating artery, those were diagnosed with the cerebral magnetic resonance imaging. It has been reported that patients with PHACES syndrome can have cerebrovascular anomalies that cause fatal complications.[2–4] Our case showed the hypoplasia of the anterior and the posterior communicating arteries. Therefore, during intracardiac surgery,

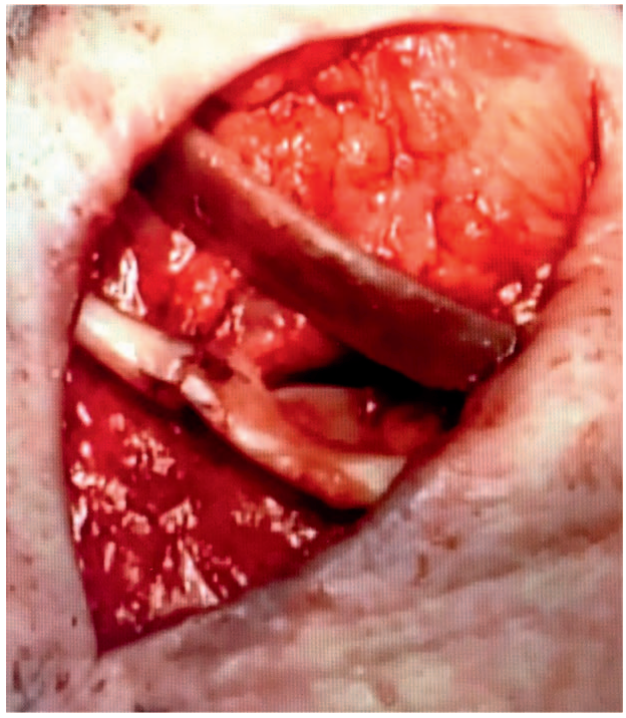


Fig. 5 Intraoperative photograph, showing complete sternal cleft repair with dislocated right ribs.

oxygenated blood was sent from the cardiopulmonary bypass to both the right and left common carotid arteries to supply the appropriate cerebral blood. Therefore, we recommend that children with PHACES syndrome should undergo cerebral magnetic resonance imaging before intracardiac surgical repair.

Torre *et al*[5] reported that primary closure of sternal cleft is preferred, possibly in the neonatal period when the chest wall is most compliant, thereby avoiding complex procedures, such as clavicular dislocation.[6] However, as in our case, it may be feasible to perform secondary repair of sternal cleft. Infants with complicated heart defects, especially those with very low birth weight, require staged surgery for cardiac anomalies, such as bilateral pulmonary artery banding and total cardiac correction, to control heart failure and hemodynamic instability prior to sternal cleft repair. Additionally, combined single-stage repair of sternal cleft and IAA with VSD should be avoided in very low birth weight infants because intracardiac repair in these cases has been reported to have higher risks. Moreover, some neonatal cases may require the chest to be left open for a few days after surgery to prevent deterioration of hemodynamic stability due

to an enlarged heart associated with long-running cardiopulmonary bypass.[7] Therefore, sternal cleft repair after total heart correction may be preferable.

PHACES syndrome is known to present hemangiomas as one of the symptoms.[8] Our case also had a somewhat iatrogenic complication, which was left nasal cavity obstruction associated with hemangioma of the nose. The obstruction also might have been generated by insertion of the nasal gastric tube after endotracheal intubation, which might have caused inflammation around the hemangioma. Although a gastric tube was required to manage feeding, we should have opted for an oral gastric tube rather than a nasal gastric tube, even after immediate endotracheal intubation for respiratory failure. Eventually, nasal plasty to remove the obstruction was performed concomitantly with sternal cleft repair because it would have been difficult for the patient to breathe, as infants generally breathe nasally. Consequently, we recommend using oral gastric tubes, especially in patients who require long-term ventilation to control severe heart and respiratory failure.

Conclusion

We presented a case of a very low birth weight infant with PHACES syndrome presenting complete sternal cleft and IAA with VSD. Staged surgical treatment of sternal cleft and congenital heart defects is a potential therapeutic option for PHACES syndrome, particularly in very low birth weight infants with complex cardiovascular disease.

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