Complete Sternal Cleft in a Filipino Newborn

Barbra Charina V. Cavan¹, Jonna A. Masongsong², Peter Y. Mancao², Arnold A. Tan²

¹Department of Pediatrics, Cebu Doctors’ University Hospital, Osmena Blvd., Cebu City, Philippines; ²Department of Surgery, Cebu Doctors’ University Hospital, Osmena Blvd., Cebu City, Philippines

ABSTRACT
We report the case of a female newborn with complete sternal cleft and patent ductus arteriosus. No other dysmorphic features were noted. Repair was accomplished at 18 days of age and the respiratory and cardiac conditions remained stable.

Key Words: sternal cleft, cleft sternum, chest wall deformity, congenital malformation or anomaly, surgery

Introduction
Sternal clefts are rare occurrences. Embryologically, the sternum is formed by the fusion of the paired parallel sternal bars between the 8th and 10th week of pregnancy.¹,² Incomplete fusion or failure of the fusion of the sternal bars leads to the development of a congenital sternal cleft.³ If fusion is partial, this is referred to as an incomplete sternal cleft, designated as superior or inferior depending on the location of the cleft. The superior cleft is the most common.¹ If the whole sternum failed to fuse, this condition is called a complete sternal cleft.⁴ Hazari et al conducted a review of the of sternal clefts from 1800 to 1998 and noted 73 cases, 40% of which had associated facial hemangiomas.¹ Sarper et al in 2002 cited a 1990 study of Shamberger and Welch which documented only 23 cases of complete sternal clefts specifically.⁵ Shalak et al, also in 2002, reported one patient with complete sternal cleft with a congenital heart disease and reviewed 35 previously reported cases, including those mentioned by Hazari.⁴

Common malformations reported to be associated with sternal clefts are ectopia cordis, vascular malformations/dysplasia such as hemangiomas, supraumbilical raphe, and even abdominal abnormalities.¹,³ Cantrell reported a syndrome also known as Cantrell’s pentalogy where there is a combination of an omphalocele, diaphragmatic hernia, sternal cleft, pericardial defect, and cardiac abnormalities.⁶ Surgical techniques used to repair sternal clefts range from primary closure to grafts to the use of metal plates.¹,⁷

Case Report
The patient was a female born term at 38 weeks age of gestation, by primary caesarian section (secondary to variable deceleration) to non-consanguineous Filipino parents. The pregnancy course was unremarkable except for thin meconium staining during labor. Her birth weight was 2.693 kg and she had feeble cry and marked chest indrawing.

On closer examination, there was a marked abnormality on the midline chest characterized by thin, transparent skin and an absent sternum. The apex beat was evident through the skin and noted to be at the 5th intercostal space. The patient’s umbilicus was at the level of the 12th rib. A thin ligament connected the base of the umbilicus to the thin skin (Figure 1). There were no cutaneous hemangiomas, no dysmorphic features, and no median raphe on the sternal area. The thin skin became less transparent and developed central atrophic scar later on. Chest radiographs showed no other abnormalities of the rib cage. Two-dimensional echocardiography revealed a small patent ductus arteriosus (PDA), a patent foramen ovale, and an intact pericardium. Abdominal ultrasound showed a laterally placed esophagus.

At 7 hours old, the baby had an episode of generalized tonic-clonic seizures. Metabolic investigations (blood sugar, arterial blood gases, and electrolytes) were normal. A cranial ultrasound revealed mild cerebral edema. Recurrence of the convulsions prompted Phenobarbital treatment which provided good control in the succeeding days. Hypoxemic ischemic encephalopathy (HIE) stage 1 due to intrapartum asphyxia was considered as the cause of the seizures.

Primary repair of the complete sternal cleft was done on the 18th day of life. The surgical technique involved the trimming of the parallel unfused sternal bars and closure of the sternum with wires. The vertical wound was closed subcutaneously. Chest tube thoracostomy was also performed. The patient was discharged on the 27th hospital day improved.

On subsequent clinic visits, the patient was noted to have normal milestones, normal repeat 2-D echo at the age of 3
years, but her growth curve has been following the 5th centile 
curve (FNRI-PPS, 1992). Phenobarbital was discontinued 
2 weeks after discharge since the patient remained seizure 
free.

The family was counseled regarding the sporadic 
ocurrence of these clefts making the recurrence risk in 
subsequent pregnancies negligible.

The family was counseled regarding the sporadic 
ocurrence of these clefts making the recurrence risk in 
subsequent pregnancies negligible.

The index case did not fully meet the classical criteria of these 
enumerated syndromes. She only had a small PDA. The 
PDA is not reported as a common major finding associated 
with sternal clefts, having been mentioned in only a few 
reports. The PDA is not reported as a common major finding associated 
with sternal clefts, having been mentioned in only a few 
reports. Unfortunately, angiography that would exclude 
other vascular malformations was not done on the patient.

There are many hypotheses on the formation of sternal 
clefts. They include the absence of the pre-sternum or 
secondary splitting and intrauterine rupture of the chorion 
or yolk sac. Early yolk sac rupture might affect the descent 
of the heart, heart development and failure of fusion of the 
sternum, while late rupture may just affect the sternum 
alone. Sternal clefts associated with vascular dysplasia may 
be due to persistent midline angioblastic tissue.

The absence of a complete sternum evidently leads to an 
unstable chest wall. Most surgeons prefer a surgical repair 
in the neonatal period, with primary direct closure. Repair during this period is favorable because of the 
compliant chest wall. Other advantages are protection of 
the vital structures within the thoracic cavity and correction 
of the visual deformity of the chest. The timing of the repair 
has to be considered in accordance with the stability of the 
patient’s condition. Fortunately for this patient, there was 
no major cardio-respiratory compromise that would have 
complicated the contemplated procedure. It was likewise 
important to make sure that after the surgical approximation, 
the cardio-respiratory status remained stable.

The lateral displacement of the esophagus noted on the 
patient’s ultrasound may be a normal variant. Such variants 
were studied by Smith et al using computed tomography 
with the cricoid as the reference point. There is no mention 
of a similar displacement of the esophagus in other case 
reports of complete sternal clefts. Her seizures were 
considered to be due to hypoxic ischemic encephalopathy 
(HIE) stage 1 secondary to antepartum and intrapartum 
asphyxia, as evidenced by the thinly meconium stained 
amniotic fluid. The neurologic sequelae of perinatal 
asphyxia include seizures, hypotonia and coma although 
this stage of HIE usually has a good prognosis.

Complete sternal cleft is reported as a sporadic event, 
whether associated with other anomalies or isolated. Counseling on recurrence risks being low in such cases was 
done with the couple, especially as this patient was their 
first child.

Conclusion

Complete sternal clefts are rare types of birth defects. 
Repair of this malformation during the neonatal period, 
especially in the absence of other major congenital 
abnormalities leads to a good outcome as seen in this case.
Acknowledgment

We would like to thank the family for giving their consent for this report, and Dr. R. Diaz for the pictures.

References