

# **AFRICAN JOURNAL OF MEDICINE**

**and medical sciences**

**VOLUME 38 NUMBER 3**

**SEPTEMBER 2009**



**Editor-in-Chief  
O. BAIYEWU**

**Assistant Editor-in-Chief  
O. O. OLORUNSOGO  
J. O. LAWOYIN**

**ISSN 1116—4077**

## Primary closure of an isolated superior sternal cleft in a Nigerian neonate

RO Ugwu<sup>1</sup>, AU Eneh<sup>1</sup>, I Gbobo<sup>2</sup> and BE Otaigbe<sup>1</sup>.

Departments of Paediatrics<sup>1</sup> and Surgery<sup>2</sup>, University of Port Harcourt Teaching Hospital, Port Harcourt, Nigeria

### Summary

Sternal cleft is a rare congenital anomaly which results from failure of the midline fusion of the lateral plates of the sternum during embryonic life. Only few cases have been reported in the literature. This is a case report of a 2-day old female who presented with a defect in the anterior chest wall from birth. Primary closure during the neonatal period is the preferred approach because of the compliant chest wall at that period with little danger of causing cardiac or respiratory compromise.

**Keywords:** Superior sternal cleft, paradoxical respiration, primary closure

### Résumé

La fente sternale est une anomalie congénitale rare qui résulte d'une absence de fusion entre les plaques latérales du sternum pendant la vie embryonnaire. Seulement quelques cas ont été reportés dans la littérature. Ceci est le cas reporté d'une femelle de deux jours d'âge admise ayant une déformation de la cage thoracique depuis la naissance. La fermeture primaire de la fente sternale durant la période néonatale est l'approche préférée due aux plaintes du patient de sa cage thoracique diminuant le risque de causer des problèmes cardio-respiratoires.

### Introduction

Sternal cleft is a rare congenital anomaly that results from complete or partial failure of the two lateral sternal bars to fuse during embryonic development [1]. The spectrum of the defect varies from a small superior or inferior sternal cleft to a total cleft of the sternum. It may be isolated or occur as one component of defined syndromes like Cantrell's pentalogy (a midline supraumbilical abdominal wall defect, a deficiency of the anterior diaphragm, a defect in the diaphragmatic pericardium, congenital heart malformation and a defect of the lower part of

the sternum) [2] or PHACES (posterior fossa brain anomalies, facial hemangiomas, arterial anomalies, cardiac anomalies and aortic coarctation, eye anomalies and sternal cleft and supraumbilical raphe syndrome [3]. We report a case of isolated superior sternal cleft that was managed successfully with primary repair.

### Case report

Baby GD was a 2 day old female who was admitted into the special care baby unit (SCBU) with the complaint of a defect in the anterior chest wall from birth. She was delivered to a 22 year old mother at term after an uneventful pregnancy. She cried immediately after birth, and weighed 2800g. Examination revealed a wide defect measuring 5cm x 6cm at the upper part of the sternum, covered by a thin circular transparent membrane which bulges during crying and expiration and forms a depression during inspiration (Fig 1 A and B). Cardiac pulsations were visible through the thin membrane overlying the defect. No other abnormalities were detected on physical examination. The chest radiograph and, computed tomography scan confirmed the absence of the manubrium sternum. Echocardiogram showed no cardiac defect.



**Fig. 1:** (a) Baby FG with superior sternal cleft with depression at rest and during deep inspiration, (b) with bulging of the membrane over the sternal cleft during crying and expiration.

Primary repair of the sternal cleft was done and consisted of mobilization and direct apposition of the sternal bars with non-absorbable 2-0 nylon sutures inserted through the intercostal spaces and tied after ensuring that there was no haemodynamic compromise. The overlying soft tissues and muscles were closed over the repair with 1-0 chromic catgut. Post operatively she developed wound sepsis which was managed with intravenous antibiotics and wound dressing with Eusol and honey. She was discharged home in stable condition on the 14th post-operative day. At 8 months of age no complications have been reported and she had a good aesthetic sternal appearance with no deformities (Fig 2).



**Fig. 2:** Baby FG at 8 months of age showing good aesthetic sternal appearance.

### Discussion

The sternum originates from the lateral plate mesoderm that fuse craniocaudally in the midline by the tenth week. Rarely, the two sternal bars do not join, leaving a defect. The failure of fusion may involve the upper sternum (superior sternal cleft) [4-7], the lower sternum (inferior sternal cleft) [8], or the entire sternum (complete sternal cleft) [9]. The aetiology is unknown, however, Fokin [1] had suggested riboflavin deficiency as a possible cause. The superior sternal cleft with fusion of the distal part is however the most common form of sternal clefts and is almost always an isolated abnormality. They can be U- shaped if the cleft extends to the last sternal body segment [5,6] or V- shaped if it extends to the xiphoid process [4-7]. Our patient had an isolated superior sternal cleft as echocardiography, plain chest radiograph and computed tomography scan confirmed no other abnormality. Inferior and complete sternal clefts on

the other hand are almost always associated with severe congenital abnormalities.

Common congenital abnormalities usually associated with sternal cleft include protrusion of the pericardium and heart (ectopia cordis) [8], congenital cardiac defects [6,8,10] facial haemangiomas [1,7,9,10] divarification of the rectus muscles [5,8,9] omphalocele [1], and syndromes like Cantrells Pentalogy [2] and PHACES syndrome [4]. None of these were found in our patient. Luis *et al* [6] reported a case of superior sternal cleft associated with enterogenous cyst, while Schimdt *et al* [11] reported one with gastroschisis.

Most reported series of sternal cleft as in the reported case were in females. This female preponderance has led to the speculation that the syndrome might represent an X-linked dominant condition [12]. Sternal clefts can be easily diagnosed at birth by inspection and palpation, however, there is also the need for investigations and imaging studies to exclude the infrequent associated anomalies.

The clinical implication of the sternal cleft is that the heart and other mediastinal structures are left unprotected and thus prone to trauma apart from the unsightly appearance of the defect. Again, the instability of the sternum may cause paradoxical movement of the chest wall during respiration which may result in ventilatory insufficiency and changes in intrathoracic pressure that may affect venous return to the heart [1]. Recurrent respiratory tract infection can also occur as a result of reduced aeration of the lungs. For these reasons, early surgical reconstruction is advised even in isolated and asymptomatic cases.

Management includes primary closure of the sternal cleft preferably during the neonatal period when the cartilaginous thorax is still very flexible with little danger of cardiac compression. Delayed reconstruction increases the risk of cardiac compromise due to increased rigidity of the chest wall and the physiologic accommodation of the thoracic organs to the circumference of the chest.

**Conclusion:** Sternal cleft is a rare congenital abnormality and primary repair during the neonatal period is the best management option because of the compliant chest wall at that period with little danger of causing cardiac or respiratory compromise.

### References

1. Fokin AA. Cleft sternum and sternal foramen. *Chest Surg Clin North Am* 2000; 10:261-276.
2. Cantrell JR, Haller JA and Rivitch MM. A syndrome of congenital defects involving the

- abdominal wall, sternum, diaphragm, pericardium, and heart. *Surg Gynecol Obstet* 1958;107:602-614
3. Metry DW, Dowd CF, Barkovich AJ and Frieden IJ. The many faces of PHACE syndrome. *J Pediatr* 2001;139:117-123.
  4. Mazzie JP, Lepore J, Price AP, Driscoll W, Bohrer S, Perlmutter S and Katz DS. Superior Sternal Cleft associated with PHACES Syndrome, Postnatal Sonographic Findings. *J Ultrasound Med* 2003;22:315-319.
  5. Yavuzer S and Kara M. Primary repair of a sternal cleft in an infant with autogenous tissues. *Interact CardioVasc Thorac Surg* 2003;2:541-543.
  6. Luis AL, López Gutierrez JC, Fernández A, Avila LF, Encinas JL, Andrés AM, Hernández F *et al*. Sternal Cleft Associated with enterogenous cyst treated during the newborn period. *Eur J Pediatr Surg* 2007; 17: 275-277.
  7. Chakkarapani E, Barnard I and Couriel J. Superior sternal cleft, cutaneous, and airway haemangiomas. *Arch Dis Child Fetal Neonatal Ed* 2007;92:F3-F4.
  8. Singal AK, Balamourougane P and Bhatnagar V. Primary closure of thoraco-abdominal ectopia cordis. *J Indian Assoc Pediatr Surg* 2006;11:250-252.
  9. Snyder BJ, Robbins RC and Ramos D. Primary Repair of Complete Sternal Cleft With Pectoralis Major Muscle Flaps. *Ann Thorac Surg* 1996;61:983-984.
  10. Acastello E, Majluf R, Garrido P, Barbosa LM and Peredo A. Sternal cleft: a surgical opportunity. *J Pediatr Surg* 2003;38:178-183.
  11. Schmidt AI, Jesch NK, Gluer S and Ure BM. Surgical repair of combined gastroschisis and sternal cleft. *J of Pediatr Surg* 2005;40: E21-E23.
  12. Forzano F and Daubeney PEF. Midline raphe, sternal cleft and other midline abnormalities: a new dominant syndrome? *Am J Med Genet* 2005;135A:9-12.

Received: 14/03/07

Accepted: 15/09/09