Original Research Paper

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CONGENITAL STERNAL CLEFT-OUTCOME & MANAGEMENT AT ICH-SINGLE CENTRE EXPERIENCE.

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ABSTRACT **Background:** Sternal cleft is a rare congenital malformation with a reported incidence of 1:1,00,000 case per live birth and with less than 1% of overall incidence among chestwall deformities. Surgical intervention plays a important line of management in the prognosis/outcome in these children. These cases generally are asymptomatic or will present with recurrent respiratory infections or for disfigurement .Herein we report our institutional experience in surgical management of this rare congenital anomaly. **Method:** We present a series of 5 cases diagnosed in neonatal period. It is a retrospective observational study , conducted between January 2016 to June 2024 at Department of Pediatric Surgery, Institute of Child Health and Hospital for Children, Madras Medical College. **Result:** Out of 5 cases, three had incomplete sternal cleft and two had complete sternal cleft , 2 cases survived and on regular follow up. This article enhances various syndromic associations with sternal cleft and the risk , prognosis associated with these conditions.

KEYWORDS : Congenital Malformation, Chestwall Deformities, Sternal Cleft , Incomplete/complete Sternal Cleft

INTRODUCTION

Sternal cleft (SC) is a rare congenital malformation that results from the defective embryologic fusion of mesenchymal cells in the ventral midline.

The first case was reported in 1739 by De Torres. Based on the clinical manifestation, the SC is classified into complete or incomplete, and the incomplete form being further subdivided in a superior and inferior type. In 67% of cases the SC is incomplete and two-thirds are female. Therefore, incomplete sternal clefting in a female is the more common anomaly. Associated malformations such as cardiovascular anomalies, midline fusion defects, hemangioma, maxillofacial defects, and pectus excavatum are common. Syndromic malformations coinciding with sternal clefting include PHACES (posterior fossa malformations–hemangiomas–arterial anomalies–cardiac defects–eye abnormalities–sternal cleft and supraumbilical raphe syndrome) or Cantrell pentalogy syndrome.

The fusion defect leaves the mediastinal viscera unprotected, resulting in an increased risk of harmful events to the underlying mediastinal organs. To restore the protective function of the skeleton by establishing bony integrity as soon as possible and to benefit from the high flexibility of the chest wall in newborns, surgical repair should be performed in the first weeks of life.

Aim & Objective

1) To emphasize the importance of sternal cleft repair in neonatal period.
2) To study various associated syndromes with congenital sternal cleft which determines the prognosis.

MATERIAL & METHODS :

It is a Retrospective Observational Study , Duration- January 2016 to June 2024 at Department of Pediatric Surgery, Institute of Child Health, Madras Medical College ,Chennai.

Sample Size- 5

Out of 5 cases, three had incomplete sternal cleft and two had complete sternal cleft , 2 cases survived and on regular follow up.

2 cases expired even before surgical intervention due to associated cardiac anomalies.

Case Series :

Case 1 – Admitted on 7th day of life, male child, diagnosed as incomplete superior sternal cleft(fig 1). Chest x-ray showed-widely placed sternal end of clavicle (fig 2)

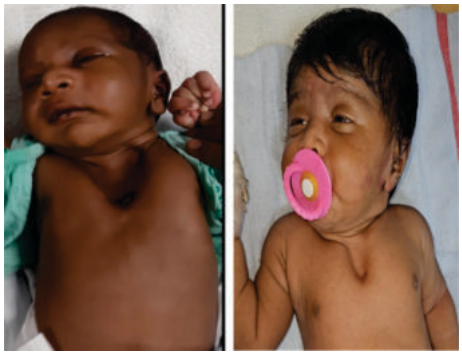


Fig 1 – Incomplete Superior Sternal Cleft

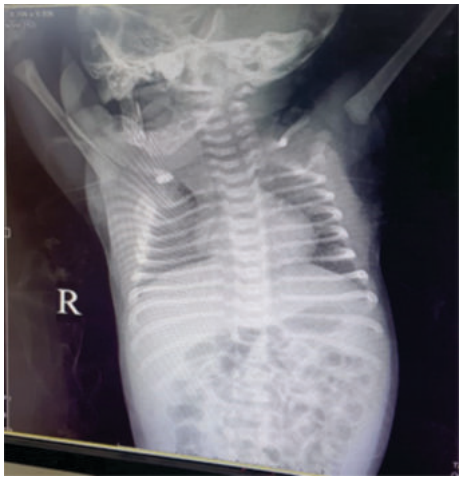


Fig 2 - Chest X-ray Showing Both Clavicle Head Facing Upward Vertical Direction, Widely Placed Sternal End Of Clavicle

Associated Abnormality – Nil .

Child was taken for surgery at day 10 of life under ETGA, sternal cleft defect measured-upper midline incision made, by extra pleural-extra pericardial approach(Fig 3) ,'U' shaped sternal cleft defect converted to 'V' shaped defect by freshening sternal bar edges (fig 4a). We used 1-0 VICRYL simple sutures for primary closures(fig 4b), ICD Placed due to inadvertent injury to lung, wound closed in layers. Post operative period uneventful, child completed successful follow up for 5 years without recurrence.

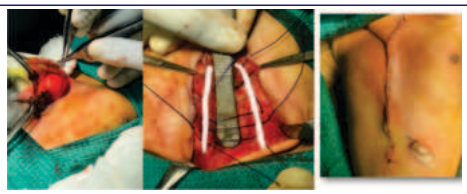
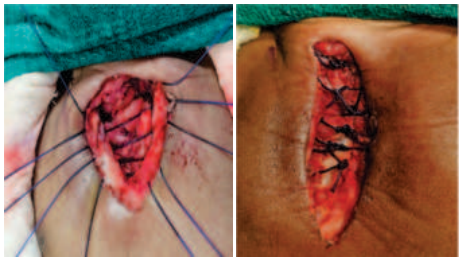


Fig 3 Extra Peritoneal & Extra Pericardial Approach



a. **b.**

Fig 4 A. intra Op Image- V Shaped Converted Defect, B. After Primary Closure Of Sternal Cleft Using 1-0 Vicryl

Case 2 – Presented at 17th day of life, female child, incomplete superior sternal cleft, child has associated facial hemangioma, to rule out PHACES SYNDROME- chest x-ray, USG abdomen, USG cranium, ECHO done.

Associated abnormality -posterior cranial fossa cyst, ventricular septal defect, fascial haemangioma more than 5 cm and sternal cleft, which satisfies definition of PHACES syndrome criteria.

This child operated at 20 days of life. Child was extubated on postoperative day 2 and discharged with propanolol therapy. Child completed 2 years of regular follow up and is thriving well.

Case 3- 24 Hours of life, female child-brought with respiratory distress, on examination child had complete sternal cleft with exomphalos(fig 5) of more than 5cm with liver as content.

Fig 5- Complete Sternal Cleft with Exomphalos Major

Associated abnormality-exomphalos major with complete sternal cleft and ventricular septal defect. This case comes under CLASS3-TOYAMA CLASSIFICATION of PENTALOGY OF CANTRELL.

Child was taken up for surgery on 7th day of life, through extra pericardial, extra peritoneal approach, abdominal wall defect and sternal cleft defect repaired. Post op period child developed wound infection-left upper lobe lung atelectasis. Child could not be weaned off from mechanical ventilation and child expired due to respiratory failure.

Case 4- 24 hours old, male child, preterm with very low birth weight with complete sterna cleft. Child expired before surgical intervention.

Case 5- 7 days old, female child ,incomplete superior sterna cleft, preterm/IUGR with major cardiac anomaly. Child expired before surgical intervention. Comparative description of each cases given in TABLE 1

Table 1- Comparative Description of Each Cases

	CASE 1	CASE 2	CASE 3	CASE 4	CASE 5
Age At Admission	7th Day	17th Day	1st Day	1st Day	7th Day
Sex	Male	Female	Female	Male	Female
Type Of Cleft	Incomplete -Superior Sternal Cleft	Incomplete -Superior Sternal Cleft	Complete Sternal Cleft	Complete Sternal Cleft	Incomplete - Superior Sternal Cleft
Associated Anomaly/ Medical Condition	Nil	Facial Heman-gioma VSD Posterior Cranial Fossa Cyst	Exomph alos ASD	Preterm/ Sepsis/ VLBW	Preterm/ IUGR/ Cardiac Anomaly
Age At Repair	10th Day	20th Day	7th Day	Repair Not Done	Repair Not Done
Survived	Yes 5 Years Follow Up	Yes 2 Years Follow Up	Repair Done. Expired Due To Res-piratory Failure	Expired Before Inter-vention	Expired Before Inter-vention

DISCUSSION

Sternal clefts result from defective embryologic fusion of paired mesodermal bands in the ventral midline. In regular development, two mesenchymal bars fuse between the seventh and tenth week of gestation, starting cranially at the manubrium and finishing distally at the xiphoid process (2). Disturbances of normal ventral midline thoracic fusion can present as a spectrum of abnormalities, including a prominent suprasternal notch, irregularities in the shape of the xiphoid, ectopia cordis, superior, inferior or complete sternal clefts (10).

Many etiological factors had been linked to this malformation (2), including alcohol consumption early in pregnancy (10) and nutritional deficiencies, especially lack of methylcobalamin (vitamin B12) or riboflavin (vitamin B2) (1, 2). Disruption of the HOX b gene might be involved in the development of sternal clefts (1)(5)(10). The malformation occurs sporadically, although autosomal recessive forms have also been described. Notably, many cases have been reported from the Middle East (2).

Sternal clefts are often asymptomatic (74 %), and diagnosis can be easily made clinically at birth by palpation and paradoxical respiratory movements. Therefore, diagnosis is usually possible in the neonatal period (64 %), however, several authors have reported patients diagnosed at older ages, (i.e., in adolescence or even adulthood). Imaging studies, including chest X-ray, USG abdomen, spine and cranium , computerized tomography and careful cardiologic evaluation (electrocardiogram and echocardiography) can help to identify any associated anomalies. Prenatal diagnosis is difficult and sternal clefts seem to be more easily identified when associated with cardiac anomalies. The second trimester is the best period for prenatal sternal study (2)(5)(10). Early recognition of the malformation is important since it is associated with an increased risk of life-threatening events involving mediastinal viscera and vessels (10). Prior to surgery, associated defects and syndromes should be excluded (5).

Surgical treatment is generally recommended, if possible in the neonatal period to take advantage of the elasticity of the thoracic cage and to achieve primary closure. At older age surgical approaches demand primary closure with autogenous tissue (73 %), bone graft interposition (10 %), prosthetic closure (7 %) or muscle flap interposition (3 %). During the first month of life, the pliability of the chest wall is maximum, which aids in easy primary closure without compressing the underlying structures. Patients undergoing surgical

repair after the age of three months have required more postoperative supportive care and shown a higher incidence of cardiac complications due to the reduction of the thoracic volume (10). Sometimes, partial or total removal of the thymus is necessary (2, 5). Intraoperative complications have been reported due to pericardial or pleural tears. Postoperative complications include retrosternal seromas or pneumothorax(5). Generally child with low birth weight, exomphalos major, major cardiac defect with complete sternal cleft reported to have poor prognosis.

CONCLUSION

STERNAL CLEFT is the rare congenital malformation, mostly diagnosed at new born period. Indications for surgery, not only cosmetics but also to improve respiratory dynamics and to protect mediastinal structures from direct injuries. We also emphasize the need for primary closure in neonatal period, ruling out associated syndromes before surgery helps in prognostication. Overall sternal cleft repair has satisfying functional and cosmetics results with low complication rates.

Declarations

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Conflict of Interest: The authors declare no conflict of interest about this manuscript

Ethical Approval : consent obtained before this publication

REFERENCES

1. Engum, S. A. (2008). Embryology, sternal clefts, ectopia cordis, and Cantrell's pentalogy. Seminars in Pediatric Surgery, 17(3), 154–160. https://doi.org/10.1053/j.sempedsurg.2008.03.004
2. Acastello, E., Majluf, R., Garrido, P., Barbosa, L. M., & Peredo, A. (2003). Sternal cleft: A surgical opportunity. Journal of Pediatric Surgery, 38(2), 178–183. https://doi.org/10.1053/jpsu.2003.50038
3. Forzano, F., Daubeney, P. E., & White, S. M. (2005). Midline raphé, sternal cleft, and other midline abnormalities: A new dominant syndrome? American Journal of Medical Genetics Part A, 135A(1), 9–12. https://doi.org/10.1002/ajmg.a.30682
4. Daum, R., & Zachariou, Z. (1999). Total and superior sternal clefts in newborns: A simple technique for surgical correction. Journal of Pediatric Surgery, 34(3), 408–411. https://doi.org/10.1016/s0022-3468(99)90487-6
5. Dòmini, M., Cupaioli, M., Rossi, F., Fakhro, A., Aquino, A., & Chiesa, P. L. (2000). Bifid sternum: neonatal surgical treatment. The Annals of Thoracic Surgery, 69(1), 267–269. https://doi.org/10.1016/s0003-4975(99)01206-0
6. Hazari, A., Mercer, N. S. G., Pawade, A., & Hayes, A. M. (1998). Superior Sternal Cleft: Construction with a Titanium Plate. Plastic & Reconstructive Surgery, 101(1), 167–170. https://doi.org/10.1097/00006534-199801000-00028
7. Hoffman, E. (1965). Surgical correction of bifid sternum. Archives of Surgery, 90(1), 76. https://doi.org/10.1001/archsurg.1965.01320070078017
8. Torre, M., Rapuzzi, G., Carlucci, M., Pio, L., & Jasonni, V. (2012). Phenotypic spectrum and management of sternal cleft: literature review and presentation of a new series. European journal of cardio-thoracic surgery : official journal of the European Association for Cardio-thoracic Surgery, 41(1), 4–9. https://doi.org/10.1016/j.ejcts.2011.05.049
9. Torre, M., Rapuzzi, G., Guida, E., Costanzo, S., & Jasonni, V. (2008). Thymectomy to achieve primary closure of total sternal cleft. Journal of Pediatric Surgery, 43(12), e17–e20. https://doi.org/10.1016/j.jpedsurg.2008.09.011
10. Fokin, A. A. (2010). Thoracic defects: Cleft Sternum and Poland Syndrome. Deleted Journal, 20(4), 575–582. https://doi.org/10.1016/j.thorsurg.2010.06.001