Abstract:
A cleft sternum is a rare congenital anomaly, which is often asymptomatic at birth. Its incidence is about 1 in 50,000 with a slight male preponderance. Clinical outcome may be unfavourable when an associated anomaly is present, particularly an intra cardiac anomaly if it coexists with the defect. Primary repair should be employed in the neonatal period because the flexibility of the chest wall is maximal and compression of underlying structures is minimal.

Keyword: Sternal cleft, Surgery, Congenital anomaly.

Fig 1
Fig 2
Introduction:
Sternal cleft is the separation of the sternum with Orthotopic normal heart and normal skin coverage [1]. Sternal clefts are extremely rare and very few cases have been reported [2]. They may be classified as complete or incomplete (Fig 3). The complete cleft is the rarer form. Isolated Sternal clefts, however, have a favorable prognosis because they present without intra cardiac anomalies, which allows a potential for primary repair of the defect. Repair of a Sternal cleft should preferably be employed early in the neonatal period when the thorax is relatively more compliant, and the primary closure is generally safe and easy [2,3]. We report herein a case of newborn baby presenting with a Sternal cleft who underwent a successful surgical repair.

Case report
Our patient was a 1 day-old, full-term male baby weighing 3 kg, who was delivered by LSCS and was diagnosed to have bifid sternum by attending neonatologist. Baby had normal APGAR scores at birth. He had a midline 'U'-shaped Sternal cleft (Fig 1). Patient's mother had complained of bulging of skin during respiration (Fig 2). There was no other obvious congenital anomaly present. Laboratory examinations were within the normal limits. A chest roentgenogram and computed tomography scans (Fig. 3,4) showed widening between the medial ends of the clavicles with the absence of the Manubrium. The Doppler echocardiography on 2nd day of life showed small ASD. After one week repeat Echo showed spontaneous closure.

Fig 3 Fig 4
Fig 5
The patient underwent Sternal repair on the 18th day of life. Midline Skin incision was made (Fig 6). Endo thoracic fascia was exposed. Skin flap raised, taking care not to injure pericardium, endo thoracic fascia and pleura (Fig 7). The Sternal bars were exposed and the pericardium and pleura separated from the undersurface (Fig 8). Sutures were taken on both Sternal bars with Ethibond sutures (Fig 9). The patient's Haemodynamic status was monitored for 5 min before tying the knots (Fig 10). Skin was sutured. Post operatively patient was nursed in NICU. There was no postoperative respiratory or cardiac compromise. Oral feeds was started on 2nd POD. Sutures were removed on 10th POD. Post operative X-ray confirmed adequate repair. Patient had a haemodynamically uneventful postoperative recovery. At a follow-up visit, the child had no complaints (Fig 12).

**Discussion:**
The first case of Sternal cleft was reported in 1740. [4] The first surgical correction was reported by Lannelongue in 1888, [3] but the first successful surgical repair of the anomaly was done by Burton in an 11-week-old child in 1943. [5]
The embryology of the Sternal cleft remains obscure. The sternum is derived from paired concentrations of thoracic lateral bands that fuse in a cranio caudal direction by the ninth week of gestation. The superior part of the Manubrium arises from three small mesenchymal primordiums, while the Sternal bands fuse laterally with the costal cartilage. Disturbances of normal ventral midline thoracic fusion can present as a spectrum of abnormalities, including a prominent supra Sternal notch, irregularities in shape of the Xiphoid, Ectopia Cordis, superior Sternal cleft or complete Sternal cleft.\[1],[2],[5]\n
The resultant anatomical defect produces a concave defect in the sternum covered by skin, with an Orthotopic heart and an intact pericardium. The skin moves paradoxically with intra thoracic pressure changes. The shape of the defect can vary from a narrow 'V' to a wider 'U'-shaped cleft. The Sternal defect, if partial, involves the upper sternum and Manubrium, in contrast to the Sternal defect in thoracic and thoraco-abdominal Ectopia Cordis, in which the defects are mostly in the lower sternum. The clinical presentation due to the defect is bulging of the skin with crying as seen in this patient. Mostly these infants with Ectopia Cordis are asymptomatic. Recurrent respiratory tract infection can occur due to reduced aeration of lungs due to associated decrease in the thoracic cavity space. If the anomaly occurs as a part of upper midline syndrome, e.g., Cantrell's Pentalogy, then associated diaphragmatic and pericardial defects are also present. Fischer\[7] reported an association with Cervico-facial Haemangiomas in 1879 and Ingelrans and Debeugny later reported occurrence of fatal post-operative haemorrhage from presumed Haemangiomas of the trachea.\[8]\n
The timing of appearance of Haemangiomas is variable and parents should be informed before surgery. Other associated conditions are supra umbilical raphe, band-like scars extending from umbilicus to the inferior aspect of the Sternal defect, and rarely a split mandible (gnathoschisis). Intra cardiac anomalies are rare in cases of Sternal cleft, a distinction from the other Sternal defects. There is a general agreement that the earliest possible primary direct closure is the best treatment.\[9]\n
There is a general agreement that the best treatment is primary closure as a neonate. As the infant grows, the cartilage becomes less malleable and the thoracic contents move anteriorly into the defect.\[10]\n
A V-shaped defect in the sternum can often be approximated by suture closure with relaxing Osteotomy of the edges if needed. The U-shaped defects often require transection of the limbs of the U at the juncture with the normal sternum, followed by suture approximation.\[10]\n
The flexibility of the thoracic cage is inversely proportional to the age of the patient. Repairs done after the age of 3 months have always required more supportive postoperative care with higher incidence of cardiac complication. Beyond 1 year of age, autologous rib/coastal cartilage grafts/prosthetic materials are required. Good mobilization of the two Sternal edges is the key to a secure approximation.

Wedge excision of the xiphoid process is required to approximate the lower edges. All the peri Sternal sutures are placed before tying the knots. Hemodynamic stability is reassessed by the anaesthetist and after approximation the knots are tied after waiting for a brief period. The patient is to be watched in the early postoperative period. The risk of complication decreases with the early age of repair.
References:


4 De Torres JI. Extract of a letter from Jos. Ignat. De Torres, MD to the Royal Society, containing the extraordinary case of the heart of a child turned upside down. London: Philosophical Transactions; 1739;41;776-8


