Abstract:
1 week old baby, isolated sternal cleft, one in one lakh live births, normal APGAR score, normal investigations except CT chest which had defect in sternal region, primary closure done, findings isolated sternal cleft, intact xyphoid cartilage, V shaped defect, intact linea alba, intact endo thoracic fascia and pericardium, absent sternum, bilateral clavicular ends normal, primary closure with approximation of sternal plates post operative period uneventful, discharged

Keyword : isolated sternal cleft, primary closure

A rare case of isolated sternal cleft Ab-stract: We report the surgical manage-ment of a 1 week old baby with isolated sternal cleft, this is a rare condition of one in one lakh live births and it was done with primary repair

CASE REPORT:
The baby was born by Caesarian section in our hospital, the indication was moderate fetal distress, mother was a primigravida and the prenatal USG failed to diagnose the defect. It was a first born male baby, term, appropriate for gestation. APGAR scoring was 6/10 at 1 mt and 8/10 at 5mts. Neonatologist referred the patient to us due to the deformed chest wall and visible heart beat, the provisional diagnosis was congenital absence of sternum. On examination, it was a one day old baby with wt 2.8kg. there was a 'V' shaped defect in the upper anterior chest wall with concave defect in the sternum covered by skin which was slightly hyperpigmented. there was paradoxical skin movement with changes in intrathoracic pressures with bulging of skin with crying. all other systems were clinically normal and all basic investigations including ECG and neurosonogram were normal. CT chest revealed defective
sternum with intact pericardium and normal lung fields. Echo showed a PFO immediately after birth and closure of it after one week with no residual shunt. Difference of opinions given by our colleagues - Radiologist - congenital absence of sternum Paediatric surgeon - sternal cleft. Finally after concluding the diagnosis as CONGENITAL ISOLATED CLEFT STERNUM, primary sternal closure was planned.

Patient was hydrated well and with antibiotic coverage, vit. K and complete anaesthetic work up, taken for surgery.

Peroperative findings were - 1. isolated sternal cleft 2. intact xiphoid cartilage, 'V' shaped defect. 3. intact linea alba 4. endothoracic fascia and pericardium intact 5. sternum absent 6. bilateral clavicular ends were normal

Procedure:
Under GA with ET tube upper midline incision made over the defect. Both sides flaps were raised over over pectoral muscles. Bilateral cartilages were undermined, keeping both pleurae intact. Possibility of primary approximation was confirmed. There was no CVS and RS distress on trial primary approximation. Hence sliding chondrotomy was deferred. Sternum was closed with 3-0 ethibond horizontal intracartilage mattress sutures. Reconstruction was done with pectoral major muscle flap raised on both sides. Subcutaneous layer was closed with 3-0 vicryl. Skin was closed with 3-0 monocryl subcuticular stitches without any drain. Patient was kept in NICU postoperatively. There was no postoperative cardiac or respiratory compromise. Patient was fed orally on the 2nd POD. Post operative CXR confirmed adequate repair. He was discharged on 10th POD.

DISCUSSION:
After the first reported case in 1740, Burten did the first successful surgical repair in an 11 weeks old child in 1943. Marcello et al did the first neonatal repair in a 14 days old baby.
Sternal defects with ectopia cordis are of severe type and are usually associated with intracardiac defects and pentology of Cantrell. They are difficult to correct and have high mortality. The sternal clefts with incomplete defects are usually isolated; 'V' or 'U' shaped; asymptomatic; with orthotopic, normal heart and with normal skin coverage. They have a very good prognosis and are associated with facial hemangiomas, midline supraumbilical raphe and PHACES syndrome (Posterior fossa malformations, Hemangiomas of face and airway, Arterial anomalies, Cardiac anomalies, Eye abnormalities, Sternal cleft and or Supraumbilical raphe).

The patient with sterna defects are to be investigated with CXR, echo, CT brain and chest, bronchoscopy, USG-abdomen etc. The aim of repair is to protect the heart and great vessels, to improve respiratory mechanics and aesthetics.

The surgical techniques:
Primary closure in the neonatal period is possible because the flexibility of chest wall is maximum and compression of the structures minimum in neonates.
After 3 months we can do sliding or rotating chondrotomies using autologous or prosthetic materials.
Costal cartilages, parietal skull or tibial bones, periostium and ribs can be used as autologous materials. Marlex, acrylic, silicone elastomer, Teflon and stainless steel mesh are used as prosthetic materials.