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AN ISOLATED SUPERIOR STERNAL CLEFT IN A NEONATE IN WESTERN KENYA: CASE REPORT

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SUMMARY

Sternal cleft is a rare congenital anomaly and very few cases have been described especially in Africa. This is a case of an isolated superior sternal cleft in a term neonate in Western Kenya.

INTRODUCTION

Although the first case of sternal cleft was reported more than three centuries ago(1), and several successful repairs have been done in the developed world, very little reporting has been done in the developing world. Almost no surgeries have been reported in East Africa. Although this condition is rare, attention needs to be given to those affected by it to improve their survival and quality of life. This is a case report of an isolated superior sternal cleft in East Africa, in a context where there is limited access to surgical repair.

CASE REPORT

A one day old neonate was referred from a peripheral health centre after having been noted to have a midline chest wall abnormality at birth. The neonate was born to a 28 year old para 3+0 mother via spontaneous vertex delivery at 39 weeks gestation by dates ,cried immediately and had a birth weight of 2.5kgs. The baby had no respiratory distress or difficulties in breastfeeding after birth. There was no family history of similar or other congenital anomalies. She passed meconium within one hour of birth and was passing urine well. Antenatal period was uneventful and the mother had received the routine antenatal care in the same health centre.

On examination we found a stable female baby with an anterior sternal defect measuring 2.5 cm covered with thin skin and visible pulsations of the great vessels. There was an anterior abdominal wall median raphe measuring seven centimeters. There was no tachypnea or respiratory distress. The breath sounds were normal with no added sounds. There was no murmur. The head circumference was 32.5cm, the anterior fontanel measured 6x4 cm with normal suture lines. There were no dysmorphic features or other clinically evident congenital anomalies. A chest X-ray showed absence of sternal ossification centres. A 2D echocardiogram done on the third day of life showed no structural heart defects but had a moderate pulmonary hypertension (63mmHg) and mild Right Atrial and Right Ventricular dilatation. A repeat echocardiogram at day eight of life showed a decline in the pulmonary pressures to 54 mmHg. Abdominal Ultrasound was normal. Renal function tests and the complete blood count were normal. The baby was dischargedwith advice on conservative management from the cardiothoracic team. The baby was reviewed at two weeks of age and was weighing 3.3kg (average weight gain of 20g/kg/day), stable and was reported to be breastfeeding well without exhaustion. Further follow up was to be done at the age of three months.

Figure 1
A Photograph of the child showing the sternal cleft and *abdominal raphe*

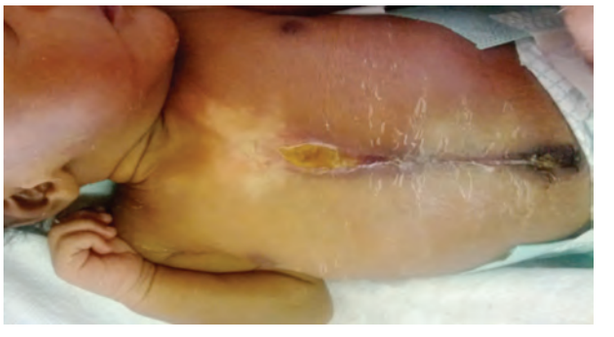
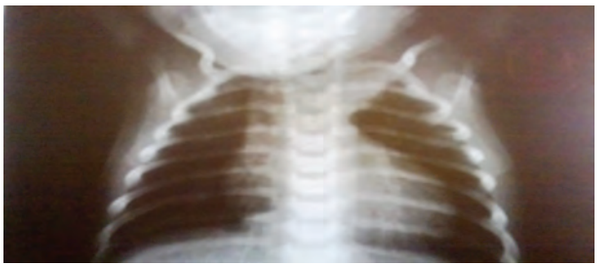


Figure 2
Chest X-ray with absence of sternal ossification centres



DISCUSSION

The sternum has its origin form the lateral plate mesoderm. These two lateral bands move anteriorly and superiorly and fuse in the ventral midline in the third month of life to form the sternum(2). The midline fusion is craniocaudal, and a concurrent lateral fusion occurs with the costal cartilages. On rare occasions, the midline fusion fails to occur (complete sternal cleft) or occurs partially (incomplete sternal cleft)(2,3). Complete sternal clefts are very rare and may be associated with other midline fusion defects. Complete sternal cleft also occurs in association with pentalogy of Cantrell, a syndrome consisting of a midline supraumbilical abdominal wall defect, a defect of the lower sternum, a deficiency of the diaphragmatic pericardium, a deficiency of the anterior diaphragm, and intracardiac anomalies4. Sarper *et.al* also described a case of a 13 year old girl who had complete sternal cleft with pectum excavatum(5).

Incomplete sternal fusion occurs either inferiorly or superiorly. Inferior sternal clefts are often associated with other anterior chest wall defects such as ectopiacordis. It has also been described as part of pentalogy of Cantrell and in association with other midline defects such as disorders of the oesophagus and anterior abdominal wall(6).

Superior sternal cleft, such as is in our case, may be isolated or occur with other congenital malformations. Singh *et.al* (India) reported a caste of a six month old male infant who presented with recurrent pneumonia and a midline thoracic bulge. He was found to have a 4.5 cm isolated u shaped sternal cleft and underwent a successful primary repair. This team also described a successful repair of an isolated u shaped sternal cleft in an 18 day old male neonate (7). Superior sternal cleft has been described in association with PHACES syndrome(8, 9). PHACES is a neurocutaneous syndrome consisting of posterior fossa malformations, haemangioma, arterial anomalies, coarctation of the aorta, cardiac defects, eye abnormalities, and sternal cleft with or without abdominal raphe. Mazzie *et.al*. described a term female neonate with PHACES syndrome who was successfully managed at Winthrop University Hospital-USA(10).In this case, the neonate was diagnosed with multiple congenital heart defects on echocardiogram at birth. The sternal cleft was successfully repaired. An aberrant subclavian artery was diagnosed at two weeks of age. At one month of age she was diagnosed with a proximal tracheal haemangioma causing subglottic stenosis together with haemangioma on the lips and gums. She underwent a successful tracheal laser surgery. Our patient appeared to have isolated type. We could not find any other anomalies from the clinical examination and the investigations done.

Prenatal ultrasonography diagnosis has been described in a case of an upper sternal cleft (11). At birth the diagnosis of sternal cleft is easily done by inspection and palpation. Diagnostic investigations are thus directed to exclude the infrequent associated anomalies. Prognosis of isolated sternal cleft is good although early surgical repair is advised even in asymptomatic cases. The repair improves respiratory dynamics, protects the heart form external trauma and is aesthetic. Early repair in the neonatal period is advised due to flexibility of the cartilaginous thorax and ability of the chest viscera to accommodate closure of the chest wall (12,13). Delayed closure increases the risk of serious compromise of the heart and lungs especially so with complete sternal cleft.

Although surgical repair performed in the neonatal period in patients with sternal cleft is the most advocated, a safe and favourable operation may also be performed with the use of autogenous tissues even in late infancy. Yavuzer and Kara reported a case of a four year old girl with sternal cleft who showed a favourable clinical outcome following successful primary surgical repair with the use of autogenous tissues (14). The case presented here was to be scheduled for surgery at a later date. This plan was agreed upon bearing in mind the limitations in neonatal critical care in our set up and the potential for a later repair though with likely increased surgical risks. This approach, however is not unusual as Chakkarapani *et.al* in United Kingdom, have reported previously of a case of a term baby diagnosed at birth but was on follow up as at three months awaiting repair later (15). It has been a common practice in developing countries to delay surgery if possible to avoid complications which may relate to critical care in the neonatal period, a service which is normally not available in many facilities.

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