

A CASE OF CANTRELL'S PENTALOGY

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SUMMARY : "Cantrell's Pentalogy is very rare congenital anomaly which is related to lower sternal cleft. A 2 days old newborn with Cantrell's Pentalogy diagnosed and treated successfully at Erbil Cardiothoracic surgical unit is presented and the literature is reviewed.

Key Words : Ectopia Cordis, Cantrell's Pentalogy, Omphalocele, Sternal Cleft.

Failure of midline fusion of the sternum may be associated with combined congenital defects of the thoracic and abdominal wall, diaphragm, pericardium and heart (Cantrell et al. 1958; Symbas and Ware, 1973; Toyoma, 1972).

A lower sternal cleft is almost always a part of a pentalogy of congenital defects first described as a syndrome in 1958 by Cantrell et al.

A patient with lower sternal cleft, omphalocele, ectopia cordis, diaphragmatic defect, pericardial defect and cyanotic congenital heart disease is described.

CASE REPORT

A 2 days newborn presented to the Cardiothoracic surgical unit of Erbil Teaching Hospital with an abdominothoracic ventral defect containing the heart and intra - abdominal organs which was extending from the midpoint of the sternum down to the umbilicus and covered only by the amniotic membrane with no skin covering. The heart protruded through the defect and was beating freely outside the chest cavity (Fig 1). The defect measured 12 cm.

vertically and 7 cm. transversely. The newborn otherwise looked healthy but mildly cyanosed. PR was 140 / min, RR was 35 / min. There was no cardiac murmur. Physical examination of the other systems was not significant. Hb was 23 gr/dl. and blood group O Rh⁺.

The chest and abdominal X-rays showed clear lung fields but an abdominothoracic opacity was seen which was related to the defect. The apex was pointing to the right side. The electrocardiogram showed right axis deviation and right atrial enlargement.

At operation an elliptical thoracoabdominal incision was carried out excising the whole amniotic cover and the umbilical stump. The operative findings were as follows (Fig 2).

1. Omphalocele containing the left lobe of the liver, stomach and transverse colon.
2. Diaphragmatic anterior defect and diaphragmatic pericardial defect.
3. Lower sternal cleft.

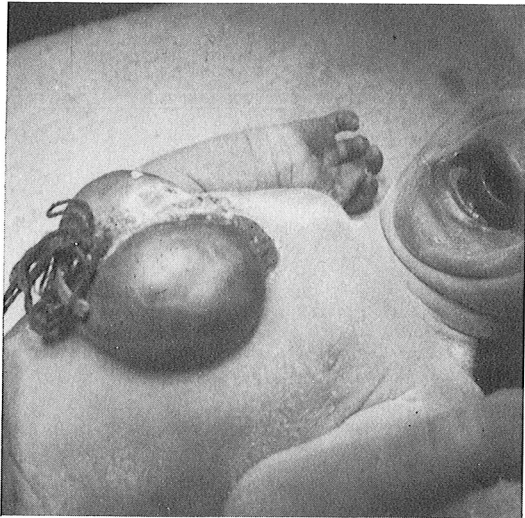


Fig. 1

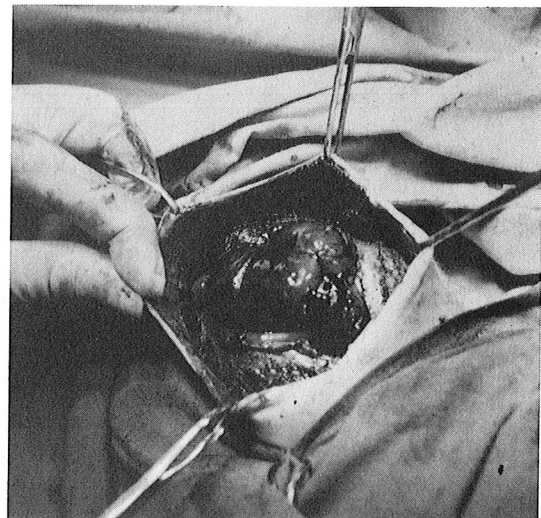


Fig. 3

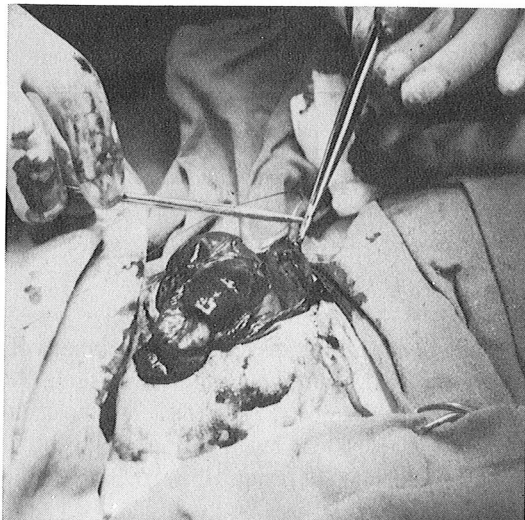


Fig. 2

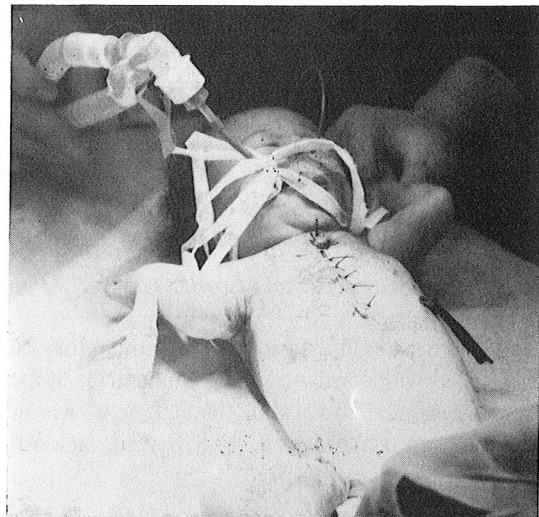


Fig. 4

4. Ectopia cordis and a direct communication between the heart and intra-abdominal organs.

5. Dextrocardia and suspected T.O.F.

The diaphragmatic defect was closed by using teflon sheat which was also sutured to the defective sternal edges. The heart was replaced inside the mediastinum thereby separating the chest cavity from the abdominal one (Fig 3, 4). The abdominothoracic defect was repaired at this stage by primary skin suturing only after carrying out extensive bilateral skin undermining and a release incision. The newborn was sent home on the subsequent reconstructive step.

DISCUSSION

The failure of sternal fusion causes three types

of sternal cleft deformities, which include : 1- superior sternal cleft 2- total sternal cleft 3- lower sternal cleft (Cantrell's pentalogy). In the three types of split sternum, the heart is generally unusually prominent and may appear to be ectopic, in cases at superior sternal cleft, the heart may appear to be in the neck. Much the same is true of thoracoabdominal ectopia cordis, in which the heart is uncovered by the distal sternal cleft and not be in any sense ectopic. There are of course instances of true ectopia cordis. Those occurring with the heart in the neck or the heart in the abdomen are extremely rare. The pentalogy, as first described by Cantrell and coworkers has subsequently been reported by others (Haller and Cantrell, 1966; Murphey et al. 1968; Symbas and Ware, 1973; Toyoma, 1972). Confir-

ming as well as in our case the invariable association of these congenital anomalies with a lower sternal cleft rather than a superior or total cleft.

The pentalogy syndrome consists of; 1- A ventral abdominal omphalocele - like defect or an actual omphalocele. 2- A cleft distal sternum, 3- A crescentic deficiency of the diaphragm anteriorly, 4- A deficiency of the diaphragmatic portion of the pericardium so that the pericardium and peritoneum communicate freely through the diaphragmatic defect, 5- A congenital cardiac defect usually one involving ventricular septal defect (commonly tetralogy of Fallot) and occasionally a left ventricular diverticulum.

There are number of reports of total sternal cleft with successful primary closure in the neonatal period (Aytaç and Saylam, 1976; Castro et al. 1977; Firmin et al. 1980; Heraldot et al. 1980; Verska, 1975), but in none of them was there a ventral diaphragmatic or pericardial defect.

Although the symptoms arising from this syndrome are related more to the type of cardiac defect, the clinical situation may occasionally demands a staged repair, with the priority given first to the extracardiac defect, namely repair of the huge omphalocele (Cantrell et al. 1958; Edgett et al. 1969; Eraklis et al. 1967; Heraldot et al. 1980).

The surgical techniques to be used must be chosen individually for each patient (Aytaç and Saylam, 1976).

Two operative methods are described :

1- Two-staged repair : The sternal gap is closed and the diaphragm is attached to the repaired sternum, the ventral abdominal defect is closed.

2- A complete one stage repair : Excision of the left ventricular diverticulum, repair of a left ventricular septal defect, and repair of the parietal defect.

In our case we did prefer the two-staged method because of the age of the patient and the size of the omphalocele.

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