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COMPLETE THORACIC ECTOPIA CORDIS. REPORT OF A CASE

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SUMMARY

Complete thoracic ectopia cordis is a rare anomaly. 17-hour of age a newborn baby had been attempted to the operative correction is reported and the pertinent literature is briefly reviwed. Unfortunately surgery is not satisfactory on this anomaly.

Key words: Ectopia cordis, Complete thoracic ectopia cordis.

The term ectopia cordis applies to a condition in which the heart is not placed within the thoracic cavity. Four types of ectopia cordis have been described: thoracic, thoracoabdominal, cervical, and abdominal [1]. Partial and complete forms are recognized. Complete ectopia cordis a rare anomaly.

CASE REPORT

K.B. (no. 196583). A 15-hour of age, full-term, newborn baby with a pulsating mass prodruding out of the midline of the torax was transfered to our hospital. There was no skin and pericardium over the mass (Fig. 1).

It was explained to the cihld's parents that the operation offered little chance of correcting the defect.

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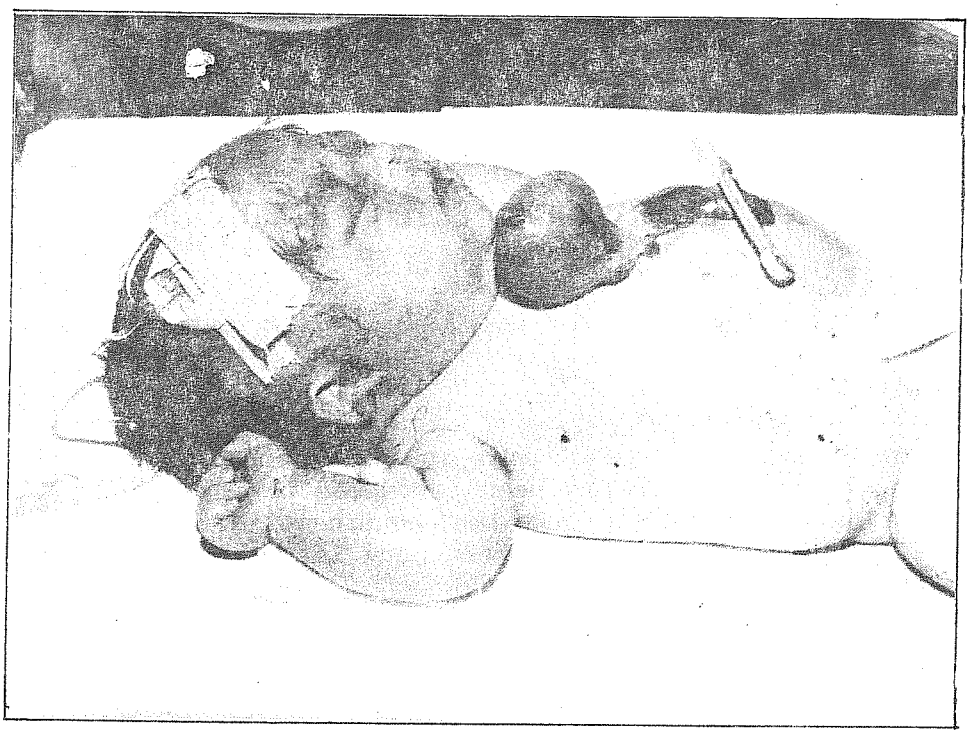

Fig. 1. The heart lies neked outside the thorax.

Under general anesthesia a midline vertical skin incision was made abowe and below the part from the heart. A total cleft sternum was confirmed. Fibrous tissues were dissected around the heart and great vessels. The heart entered the thorax through an opening in the left hemidiaphragm. When the heart was manuplated into this new position, its action had embarrassed and it was necessary to allow it to return to the original ectopic situation.

After the heart entered into the thorax, only the skin was approximated over the heart. When the operation was completed the heart beats gradually weakened and the patient died one and a half hours after the operation.

The post-mortem examination revealed a large ventricular septal defect, hypoplastic pulmonary artery associated with infundibulary stenosis and large patent ductus arteriosus, persistent left superior vena cava, and a dextroposed aorta.

DISCUSSION

According to Kaplan et al[2], the occurence of thoracic ectopia cordis with a cephalic-pointing cardiac apex suggest an arrest of cardiac descent at 3 weeks of developments consistent with their finding of ectopia cordis in a 28-day human embryo. Mechanical compression secondary to rupture of the chorion and/or yolk sac at 3 weeks of gestation would interfere with normal cardiac descent and compress, yielding thoracic and pulmonary hypoplasia.

Although complete ectopia cordis is a rare anomaly, in 1948 Bayron[1] 142 cases, and in 1985 Kaplan et al[2], had collected 200 cases from literature. But many of them probably have had only a part of the heart exposed, and others have been simply cases of bifid sternum, the heart remaining within the thorax, albeit just beneath the skin, namely these were may be partial ectopia cordis. In the case of complete ectopia cordis, that is to say, genuine ectopia of the heart as described by Ravitch[3], sternum is cleft and the heart protrudes anteriorly far beyond the chest wall, covered only by pericardium, or lies altogether naked outside the body, as in some reports[2,4-6], an our case.

Some cardiac and other malformations often are associated with ectopia cordis, but the main problem of correction of the anomaly is the heart's not being able to adopt the reposition because of kinking the great vessels. There are neither surviving cases of complete thoracic ectopia cordis without operation nor after attempted operations.

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