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Complete Extrathoracic Ectopia Cordis: Proposal of a New Surgical Approach

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Ectopia cordis is a quite rare congenital deformity, first described by Niels Stensen in 1971 in a case of Tetralogy of Fallot (cited by Shao tsu. L., and Cremer, W.)[1,2] Abbott reported 8 cases of ectopia cordis in 1000 cardiac malformations (0.8 %) in 1936. The ratio of this anomaly on total population is about 0.004-0.05 % (all quoted by Cremer, W.)[2] More than 200 cases of ectopia cordis have been presented to date.[2]

Case Report

A 2300 gm premature male infant (file number: 264886) was transferred to our clinic one hour after birth. This was the first pregnancy of a 16 year-old mother who had a spontaneous delivery with head presentation. Physical examination revealed a premature baby in fairly poor condition with mild cyanosis. The heart was totally placed outside of the thorax and not covered either with pericardium or skin (Figure 1). The rest of the body looked normal and liver and spleen were nonpalpable. The heart was beating regularly and effectively, located extrathoracicly through a defect of the sternum. ECG showed no abnormality.

The infant was taken to the operating room urgently on the 23rd of June, 1971, two hours after his admission; and tracheal intubation and general anesthesia were applied throughout the procedure. Figure 1 was taken in the operating room after the incisions were made.

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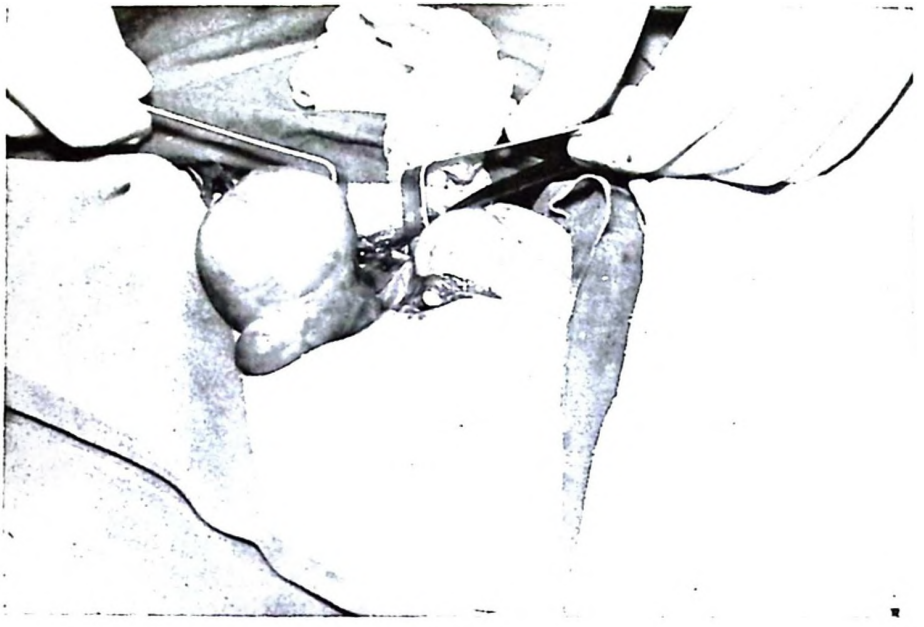

Figure 1

Picture of the patient taken in the operating room at the beginning of the operation.

Figure 2

Incisions made to open the left thoracic cavity.

Two incisions were made, one as a left anterior thoracotomy between the 4th and 5th intercostal cartilages, and the second one vertical to it extending down towards the abdomen from the hole where the heart was located extrathoracicly (Figure 2). Left lower lobectomy was performed first to create a suitable space for the heart in the chest cavity and the heart was placed into this new place. The heart slowed down after 5-6 minutes of beating due to impaired venous return because of inadequate diastole. The heart was moved back to its original location and massaged and placed again into the place of the removed lobe. After several attempts it was decided that the space was not large enough for the heart to beat effectively. Following these attempts, a left pneumonectomy was performed and the heart was placed into the left thoracic cavity. This time there was no impairment in the cardiac function and the beating of the heart was observed for 30 minutes. The chest was closed after it was decided that the space was adequate for the heart to be left in (Figure 3).

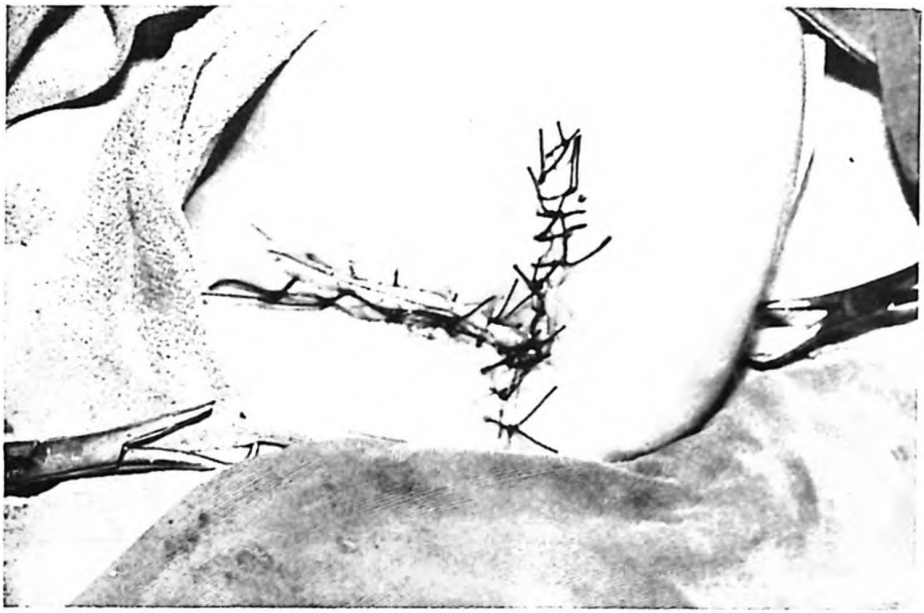

Figure 3
Incisions closed at the end of the operation.

The patient lived 20 hours after surgery while the heart kept beating satisfactorily and peripheral pulses were strong meanwhile. After 20 hours the heart slowed down and respiration became poor, and the patient died when he was 26 hours old. Urinary output was never satisfactory.

Autopsy of the heart showed patent foramen ovale, hypoplastic left ventricle, transposition of the great arteries and anomaly of the coronary arteries (orifices of both coronary arteries originated from the same location on the aortic wall). See Figure 4.

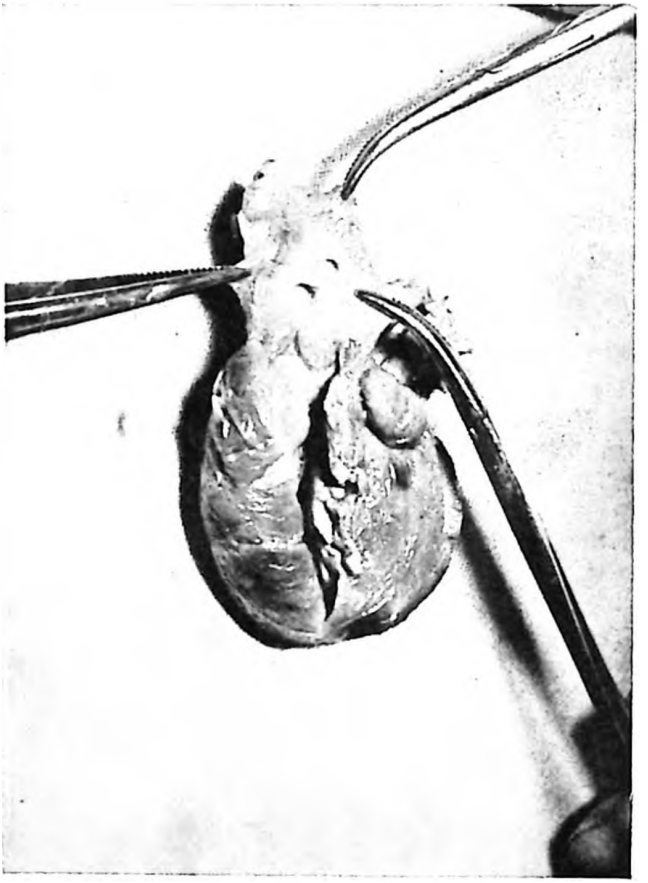

Figure 4
Abnormal coronary arterial orifices on the aortic wall.

Discussion

Exact meaning of the ectopia cordis is still not very clear as it was mentioned by Barlow who wrote the first well-studied case in 1938.[3] In earlier days some authors accepted even dextrocardia as a form of ectopia cordis. Abbott[4] described it as "a displacement so that the heart poses out of the thorax and comes lie upon the outer surface of the body or in the abdominal cavity". In 1962 Kanagasuntheram and Verzin[5] classified the types of ectopia cordis as (1)cervical, (2) thoracocervical, (3) thoracic,

(4) thoracoabdominal and (5) abdominal. Cantrell et al[6] reported a syndrome complex consisting of defects of the thoracoabdominal wall, thoracoabdominal ectopia cordis, defects of diaphragm, pericardium and heart in 1958 (so called Cantrell's syndrome). Similar cases were reported later.[7-9, 10]

Ectopia cordis can be classified as complete and incomplete. In the incomplete form the heart lies in a subcutaneous position, where in the complete form there is no pericardium or skin covering the heart as it was for our case. Extrathoracic ectopia cordis above the diaphragm can be located either in the cervical or thoracic portion of the upper body. Cervical and thoracic ectopia cordis are usually fatal. Abdominal ectopia cordis is generally compatible with life, several cases in the adults having been reported to date including some women who had healthy pregnancies.[11]

Patten[12] believes that the defect is established during the 3rd week of foetal life and he states that this is the time the demarcation between intra-and extra-embryonic coelom is produced by the folding of the body. Millhouse and Joos[13] studied the details of 52 cases and showed that the most common associated cardiac anomaly is the ventricular septal defect, and the extracardiac anomaly, the cleft palate or cleft lip. Incidence of ectopia cordis was slightly higher among premature infants and males. Pregnancy was uneventful among all of these cases except one. Surgery was attempted in only 10 cases including theirs with no effect on survival. Average life expectancy was 2 or 3 days.[13]

Surgical intervention in ectopia cordis excluding pure abdominal forms is the only hope despite poor results up to date. The most difficult problem is to find a suitable space for the heart to be replaced in the thorax. In Millhouse and Joos' paper it was mentioned as a suggestion of Dr. B. Meyer that the left lower lobectomy might be performed if there is not enough space to put the heart back into the chest cavity.[13] However, this has never been attempted so far Left pneumonectomy has never been suggested or attempted until our case. We believe this procedure should be done as a last surgical maneuver if left lower lobectomy does not provide large enough room for the heart to beat effectively. Our patient might have lived after left pneumonectomy if he had had no other cardiac abnormalities.

Surgery in Cantrell's syndrome is successful in some cases using the techniques of open-heart surgery in the current practice.[10]

Summary

A case of complete extrathoracic ectopia cordis was reported and the literature reviewed. A new surgical approach was applied by removing the entire left lung to find a suitable space to replace the heart into the chest cavity. Although the result ended in failure, it is hoped that this new approach might give the last and only hope to those children if there is not enough space following left lower lobectomy to put the heart back into the thorax. Our case showed that patients may survive this procedure and be a candidate for a longer life if they do not have other serious defects within the heart.

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