ECTOPIC CORDIS

(A Case Report)

by

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Malformations incompatible with life resulting in stillbirth constitute a fairly discrete group, occurring once in about 200 viable births (Eastman and Hellman, 1966). Ectopic cordis is a condition in which the heart is exposed on chest wall owing to faulty development of sternum. The heart protrudes abnormally ventrally outside the chest. This is a rare congenital malformation and is usually fatal (Patten, 1953; Hollinshead, 1966; and d'Abreu, Collis and Clarke, 1971).

Major and Conn (1953), however, were able to collect 142 cases after reviewing the world literature. They added 5 other cases who underwent surgical correction of this anomaly.

Recently, we came accross a case of ectopic cordis which lead to stillbirth in a primigravida. Because of its rarity this case is reported.

Case Report

Mrs. A.B., 19 years, Hindu, Bengalee housewife, was admitted at 10-45 p.m. on 28-8-1975 with the complaint of loss of foetal movement for 3 days.

Menstrual History

Menarche—13 yrs. Cycles—regular. Flow—average. Last menstrual period—21-12-74. Expected date of delivery—28-9-75.

Obstetric History: She was primigravida. She attended antenetal clinic twice and her last visit was on 19-7-75. The following investigations were done at O.P.D.:—Haemoglobin—10 Gm. per cent, Blood group A, Rh positive, Postprandial blood sugar (after 100 Gm of glucose) —90 mg. per cent, V.D.R.L. test—negative.

General Examination: She was of average built and nutrition. Nothing abnormal was detected.

Abdominal Examination

The uterus was 34 weeks' size. Painful uterine contractions were absent. Head was floating and foetal heart sounds could not be located. There was no foetal movement.

Internal Examination

Cervix was ripe and membranes were intact. Pelvis was adequate. There was no abnormal vaginal discharge. Repeated examinations failed to demonstrate the foetal heart sound. The labour pains commenced at 2 a.m. on 30-8-75 and she gave birth to a female stillborn congenitally malformed baby at 1-25 a.m. on 31-8-75. Prophylactic injection of methergin was given and the third stage was uneventful.

She did not give any history of drug intake (except iron tablets and inj. Tetanus toxoid), or sickness in the present pregnancy. There was no history of previous irradiation. There was no history of any abnormal births on either the maternal or paternal side of the family. Her husband was 23 years old.

She was discharged on 31-8-1975 (discharged on request).

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External appearance of the baby and placenta

The baby weighed 2.1 Kg. and the crown-heel length measured 44 cm. There was a circular defect in the anterior wall of the thorax and upper abdomen of 9.2 cm. diameter (Fig. 2). The heart with the origin of great vessels were bare over the chest wall (Fig. 1). The neck and the upper part of the chest were normal. The liver was covered by a shiny membrane in lower portion of which the umbilical cord was attached. External genitalia were normally developed. No other abnormality was observed.

The placenta was normal in appearance and weighed 800 gm. The umbilical cord measured 36 cm. and contained two umbilical arteries and one vein.

Postmortem Examination (Pertinent findings)

The following observations were made on opening the thorax and abdomen.

1. A radial defect in the lower anterior thoracic wall and upper abdominal wall which permitted the heart to herniate.
2. There was associated ventricular septal defect. The diaphragmatic pericardium was lacking.
3. The lungs were deep in the thoracic cage in their normal position. They did not float in water.
4. There was a gap in the anterior portion of the diaphragm.
5. Liver, Spleen, Stomach, small and large intestines were normal.
6. The supraranal glands, kidneys, ureters and urinary bladder were normal.
7. Internal genital organs were feminine in type and were normally developed.

Discussion

The embryological background of this abnormality has been variously explained. Hamilton, Boyd and Mossman (1964) opined that the ectopic position of the heart is due to failure of the costal cartilages, muscles and sternum to differentiate in the anterolateral chest wall. Patten (1953), however, explained that when the ventral body walls in the cardiac regions are being formed, they 'catch the heart' outside their place of convergence. But in the present case, and also in most of the reported cases, there were associated defects in the body wall which could not be adequately explained by Patten's view. He, however, admitted that this abnormality might also be due to deficient growth pattern of body wall.

According to Hanson (1919) the sternum first arises as 2 laterally situated 'sternal bands'. Subsequently, the two sternal bands fuse in the midline. Non-union of these bands lead to sternal cleft, and thus allow the heart to protrude outside the chest (ectopic cordis). Benson *et al* (1962) classified sternal clefts into upper sternal cleft, distal sternal cleft, and complete cleft of the sternum. In a critical analysis they observed that, because of the associated ventral displacement of the heart, these cases of sternal cleft are usually reported in the literature as cervico-thoracic ectopic cordis (upper sternal cleft) and thoraco-abdominal ectopic cordis (distal sternal cleft). They concluded that the basic defect in all these abnormalities could be due to absence or deficiency of ventral midline mesenchymal tissue into which the migrating mesodermal structures would normally grow. They also suggested that the associated pericardial, diaphragmatic and intracardiac lesions arise as a result of developmental failure of appropriate segments of the mesoderm.

Weese (1818) classified ectopic cordis in three groups: cervical, thoracic and abdominal. Subsequently, Byron (1948) added a fourth category i.e. thoraco-abdominal. Benson *et al* (1962) were able to collect only 20 cases of thoraco-abdominal type of ectopic cordis. The present case belongs to the last type.

In the thoraco-abdominal type of ectopic cordis the following defects are ob-

served (Major and Conn, 1953; Benson *et al*, 1962).

(1) A midline supraumbilical defect of diastasis recti or of omphalocele type.

(2) A defect of the lower sternum (partial absence or cleft of the sternum).

(3) A deficiency of the anterior portion of the diaphragm.

(4) A defect in the diaphragmatic portion of the pericardium.

(5) An intracardiac defect usually ventricular septal defect. The other types of anomalies that may be associated are tetralogy of Fallot, Cortriloculare with a single ventricle, left ventricular diverticulum, tricuspid atresia etc.

Cantrell, *et al* (1958), however, insisted that this association of group of congenital defects should be considered as a separate 'syndrome' and should not be labelled as ectopia cordis.

One must be very careful in diagnosing these cases as Mustard *et al* (1958) has sounded a note of caution. Very rarely a diverticulum of the left ventricle may be misinterpreted as 'ectopic cordis' when a pulsating tissue is protruding through a gap in the chest wall. Similarly, diverticulum of the pericardium only may be confused and labelled as ectopia cordis (Maier and Bortone, 1949; Major and Conn, 1953).

Despite intensive study, the causes of congenital malformations are still little understood (Sultana *et al*, 1975). Screening of maternal obstetric history in the present case failed to reveal evidence of any environmental factor (drugs, maternal infections, high maternal age or irradiation) contributing in the pathogenesis of congenital malformations. In experimental animals cardiovascular malformations have been observed with diets deficient in vitamin A (Wilson and Warkany, 1950) and with induced folic acid deficiency (Baird *et al*, 1954 and Asling *et al*, 1955). Subclinical deficiency of these vitamins, however, could not be excluded in this case.

Summary

A case of ectopic cordis (thoraco-abdominal type) in a stillborn foetus is reported. The embryological background has been discussed and the literature has been reviewed.

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References

1. Asling, C. W. N., Nelson, M. M., Wright, H. V. and Evans, H. M.: Anat. Rec. 121: 775, 1955.
2. Baird, C. D. C., Nelson, H. M., Monie, I. W. and Evans, H. M.: Circulat. Res. 2: 544, 1954.
3. Benson, C. D., Mustard, W. T., Ravitch, M. M., Synder, Jr., W. H. and Welch, K. J.: "Pediatric Surgery" Vol. 1, Ed. 1, Chicago, 1962, Year Book Medical Publishers p. 227-230.
4. Blatt, M. L. and Zeldes, M.: "Ectopic cordis—Report of a case and Review of literature" Amer. J. Dis. Child. 63: 515-529, 1942.
5. Byron, F. J.: Thoracic Surg. 17: 717, 1948.
6. Cantrell, J. R., Haller, J. A. and Ravitch, M. M.: "A syndrome of congenital defects involving the abdominal wall, sternum, diaphragm, pericardium and heart." Surg. Gynec. & Obst. 107: 602-614, 1958.
7. d'Abreu, A. L. Collis, J. L. and Clarke, D. B.: "A Practice of Thoracic Surgery" Ed. 3, London, 1971, Edward Arnold (Publishers) Ltd. p. 64.
8. Eastman, N. J. and Hellman, L. M.: "Williams Obstetrics" 13th Ed., New

Delhi, 1966, Amerind Publishing Co. Ltd. p. 1041.

9. Hamilton, W. J., Boyd, J. D. and Mossman, H. W.: "Human Embryology" ed. 3, (Revised), Cambridge, 1964, W. Heffer & Sons Ltd. p. 200, 258.
10. Hanson, F. N.: "The ontogeny and phylogeny of the sternum". Amer. J. Anat. 26: 41, 1919.
12. Maier, H. C. and Bortone, F.: "Complete failure of sternal fusion with herniation of pericardium". J. Thoracic Surg. 18: 851, 1949.
13. Major, J. W. and Conn, W.: "Thoracoabdominal ectopic cordis". J. Thoracic Surg. 26: 309, 1953.
14. Mustard, W. T., Duckworth, J. W. A., Rowe, R. D. and Dolan, F. G.: Canad. J. Surg. 1: 149, 1958.
15. Patten, B. M.: "Human Embryology", 2nd Ed., New York, 1953, Mc-Grew Hill Book Co. Ltd. p. 509.
16. Sultana, Z., Talib, V. H., Patil, S. D., Magar, D. N. and Sharma, K. D.: "Congenital malformations—an autopsy study". J. Obst. & Gynec. India. 25: 392, 1975.
17. Weese (1818): Quoted by Reference 15.
18. Wilson, J. W. and Warkany, J.: Paediatrics. 5: 708, 1950.

See Figs on Art Paper XIV