HOLOACARDIUS

(A Case Report)

by

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Ever since Benedetii, in 1533, described the first case, acardiac anomalies or monster are reported in literature from time to time. Malformations are considered to be rare because so far even less than two hundred cases have been reported in the world literature. It has been estimated that acardiac monster occur once in 34,600 deliveries and in 1% of monozygotic twins.

An acardius is a monster that is characterized by the absence of heart, and only develop in single ovum twin pregnancies owing to inequalities in the communicating placental, circulation. One twin is well developed and normal while the other is imperfectly formed. The affected twin is thought to depend upon the functioning heart of the normal twin for its circulation and survival.

CASE REPORT

L. B. a 26 year old Hindu, gravida 5, para 4 who had prenatal complications such as mild hydramnios and toxaemia of pregnancy, was admitted in active labour at 38 weeks gestation. Only one foetal heart tone was detected on admission. The patient gave no history of diabetes, hypertension or previous twin in the family Labour was uneventful, and after artificial rupture of membrane, she delivered a 2800 gm. male infant who appeared active and normal. Placenta expelled spontaneously followed by second twin which was a gross monstrosity presenting no heart beat or other evidence of life. The still-born foetus, however, was not macerated.

Subsequently the viable twin has developed normally and has presented no problem in the neonatal period. The patient has normal puerperium.

Pathology

The malformed infant was an amorphous mass weighing 1 pound and measuring 5" x 5" x 4" with uniform shape of cephalic and caudal poles. The caudal pole of the body was identified by rudimentary appendage which did not resemble the normal structure in any way (Fig. 1). The cephalic pole was identified by diffuse hair growth. There were no structures recognizable as eyes, ears, nose or mouth. The only indication of upper extremities was a rudimentary appendage. Below the umbilical cord the external genitals appeared to be normal but the testes were undescended.

Radiographic films confirmed the polarity and indicated a recognizable but highly abnormal anatomic structure which was masked by a heavy investment of subcutaneous tissue. There was a complement of ribs which could not be counted but no indication of a sternum. The thoracic verteberae could be identified, but these were somewhat disorganized and could not be counted. There were no cervical vertebrae. The cranium was very small and rudimentary. The identity of rest of the bones was questionable.

The viscera consisted of bowel (a coil of intestine) undescended gonads in the form of two small nodules. There were no signs of heart, kidneys, pancreas, liver, suprarenals, diaphragm, lungs thymus or thyroid.

The most striking feature of this foetus was the vasculature. There was one umbilical artery and one umbilical vein in the umbilical cord. Above the diaphragm the umbilical vein divided into several smaller branches to intercostal vessels and the base of the skull. The single

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umbilical artery was derived from aorta which gave of branches to base of the skull.

The placenta was monochorionic, diamniotic structure weighing 450 gm. Large anastomotic vessels between cord insertions were visible on the amniotic surface of the chorionic plate. The placenta was otherwise grossly and microscopically normal.

Discussion

The author can still recall a twin delivery during his training period. The first twin was normal while the second twin presenting as breech could not be delivered inspite of all skillful manouvres and subsequently a caesarean section was required. A monster of omphalus variety was born. To date this case is unreported and thus unrecognized. Although this abnormality is very rare, its incidence is not exactly estimated due to unreported cases.

Defective germ plasm theory, chromosomal aberrations and severe mosaicism are discarded. Two most popular theories are in field. Potter (1952) stated that the primary defect is failure of the heart to develop and that the acardiacus foetus survives in utero only where there is an anastomosis in the placenta with the vessels of the normal twin. Others Cladius (1859), Ahfeld (1879), Hunziker (1907) Loescheke (1948) and Benirschke (1970) believe that the primary defect is an abnormal placental vascular communication between the two twins where one twin receives the arterial blood from the other twin and as a result the heart of the one twin degenerates after having been established. Although the circulatory support provided by the normal twin allows the acardiacus twin to survive, it is probably inadequate and therefore responsible for other characteristics of acardiacus such as the peculiar massive overgrowth of mucoid subcutaneous tissue. Severn and Holyoke (1973) are of the view that the overgrowth of the subcutaneous tissue is responsible for the lack of form characterstic of these monsters and since the recognizable form is hidden under this tissue rather than absent, they suggest that the term acardiacus cryptomorphous (hidden) form is more appropriate than the term "amorphous" (noform).

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See Fig. on Art Paper XIII