Case Reports

FOETUS ACARDIUS ACEPHALUS

K. R. S. POOL, M.B., B.S., F.R.C.S., M.R.C.O.G.

Senior Registrar, Obstetric and Gynaecological Department, Kings College Hospital, London, S.E.5

Fœtus Acardius Acephalus is a monster which can occur only in a monozygotic multiple pregnancy. The majority occur in twin pregnancies but can occur in triplet pregnancy as described by Ross (1951). In order that the acardius or heartless monster may live, it must receive its blood supply from the twin with a functioning heart. Schatz's (1898) classification recognizes two main groups of acardii, the hemicardius with an imperfectly formed heart and the holoacardius with complete absence of the heart. Das (1902) subdivided the holoacardius group into acephalus, amorphus, anceps and acormus. In a review of the world literature Napolitani and Schreiber (1960) noted that holoacardius monsters had been reported 149 times. It has been calculated that they occur once in 34,600 deliveries. This figure is based on their occurrence in about one per cent of monozygotic twins. The specimen to be described is an acardius acephalus. (Fig. 1).

Case Report

Mrs. A., an unbooked para 4, aged 29, was admitted to hospital as an emergency in established labour with the diagnosis of a shoulder presentation. The pregnancy had been uneventful with no abnormal findings. Labour began at 36 weeks after spontaneous rupture of the membranes. On admission, abdominal examination revealed a uterus larger than expected from the dates, with a fetus lying in the transverse position. The foetal heart was audible. On pelvic examination the presenting part was in mid-cavity and with the cervix fully dilated. Two digits were felt but the exact identity of the presenting part remained obscure. The patient was anaesthetized in order to carry out a more thorough pelvic examination. A few minutes later a fetus acardius was delivered 'cephalic' end first. A second twin was diagnosed and delivered as a breech following internal version. The third stage of labour was normal.

X-ray Report. 'This maldeveloped fetus has a vertebral column of only 24 segments, as opposed to the normal 29, excluding the coccyx. The upper 6 cervical segments are missing. There are 6 lumbar segments, the third of which shows a lateral hemivertebra deformity. The shoulder girdle and upper limbs are very imperfectly developed. On the left side there is an unrecognizable mass of bone representing an attempt at an upper limb. On the right, several ill-formed long bones are present but they cannot be identified individually. The ribs are normally developed with the exception of bilateral vestigial first ribs. The pelvis is well ossified, as are the femora, tibiae and fibulae but there is a congenital dislocation of the left hip. Bilateral talipes are present with only a few maldeveloped bones in each foot'. A photograph and X-ray of the partially opened fetus is shown in Figs. 2 and 3.

FIG. 1.

Pathology. The specimen was that of an acephalic fetus, 30 cm. in length weighing 790 g. after fixation. There was absence of recognizable skull. The pelvis, spine and lower limbs were present but only one rudimentary upper limb. There was bilateral talipes equinovarus. The heart and thoracic cavity were absent. The blood supply came direct from the cord and there appeared to be a posterior 'aorta'. No stomach, liver, pancreas or kidneys were recognized. A urinary bladder, ovaries and female genitalia were present. There was a spinal cord which shows thickening of its upper end. A flattened structure 1 cm. x 1 cm. x 0.2 cm. in size

FIG. 2.

was shown on histological examination to be adrenal tissue.

The placenta was healthy and weighed 630 g. after fixation. The umbilical cord of the monster was atrophic and contained two umbilical arteries and one vein. There was one chorion and two amnions.

Discussion

The existence of the monster is completely dependent on the normal twin. Some of the blood reaching the placenta via the umbilical artery of the normal foetus passes directly into the umbilical artery of the monster. The direction of the flow of blood in the umbilical arteries of the foetus acardius is therefore reversed so supplying the monster with blood deficient in oxygen and nutrients. The ætiology remains obscure. It was originally thought

FIG. 3

that the vascular connection between the foetus would cause hypertrophy of the heart of one twin and atrophy of the other but it seems more probable that this condition is due to an original failure of parts to form rather than to secondary atrophy (Potter, 1952).

My grateful thanks to Mr. S. G. Clayton for his permission to report this case; to Dr. C. Dulake for his help with the dissection of the specimen and to Dr. M. J. Turner for the X-ray report.

REFERENCES

- DAS, M. (1902): Acardiacus Anceps, *J. Obstet. Gynec. Brit. Emp.*, **2**, 341.
 NAPOLITANI, F. D., and SCHRIEBER, I. (1960): The Acardiac Monster, *Amer. J. Obstet. Gynec.*, **80**, 582.
 POTTER, E. J. (1952): 'Pathology of the Foetus and the Newborn', p. 184. Chicago: Year Book Publishers Inc.
 ROSS, J. R. W. (1951): An Acardius Amorphus in a Triplet Pregnancy, *J. Obstet. Gynec. Brit. Emp.*, **58**, 835.
 SCHATZ, C. F. (1898): 'Die Acardii und ihre Verwandten'. Berlin: A. Hirschwald.