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ANENCEPHALY IN UNIOVULAR TWINS

REPORT OF A CASE

BY

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Anencephaly is a relatively common congenital malformation occurring with a frequency of up to 0·3 per cent. of births. Malpas (1937) reported an incidence of 0·308 per cent. in 13,964 births, and Record and McKeown (1949) from 158,307 births found an incidence of 0·23 per cent. Hurwitz (1955) reviewed the whole literature and included his own series of 5,264 births, among which he found fifteen cases of anencephaly, representing an incidence of 0·28 per cent.

Of greater interest than the occurrence merely of a particular type of malformation is its occurrence in one of a pair of uniovular twins. So far as the causes of malformations are concerned, they may be either hereditary or environmental, or a combination of both. When a malformation has occurred in one of a pair of uniovular twins it has been suggested that the causative factors must have been associated with the environment of the affected foetus, because both foetuses would have been malformed if an hereditary factor alone had been operating. This is not to imply that the genetic constitution of the twins is in no way responsible, but that without an additional environmental factor any genetic factor will not operate. That anencephaly can be caused by environmental factors is also suggested by experimental work (*e.g.* Ingalls, Curley, and Prindle, 1952; Hicks 1952). From the practical standpoint this means that certain malformations may be classed as preventable diseases, although it is difficult to see how satisfactory prophylactic measures can be taken at a time when the state of pregnancy cannot definitely be ascertained.

A case in which one of a pair of twins was anencephalic was reported by Josephson and Waller (1933). Here, however, both of the foetuses were abnormal; twin A was an iniencephalic and twin B was anencephalic. Moreover, there was no specific report on the placental membranes; "there was only one placenta, the babies were both females, and probably identical twins". These authors quoted a report by Thompson (1925) of a case of anencephaly in one of a pair of twins. In fact, in Thompson's case, the normal living child was a male and the one malformed foetus was not a true anencephalic but, rather, an acardiac acephalic. There was no comment on the state of the placental membranes.

Potter (1952) reported having seen two instances of twins in a single amniotic sac and attached to a single placenta in which one twin was normal while the other grossly abnormal. In one of these pairs the malformed foetus was anencephalic. The presence in this case of one chorion and one amnion seems definite evidence of an origin from a single ovum. When a single chorionic membrane is associated with two amniotic sacs it is usual to consider the twins as being uniovular; but this is not invariably true, it seems, for Potter and Crunden (1941) reported an instance of a monochorionic placenta associated with twins of the opposite sex. However, where there is a single chorion and the twins are of the same sex, it is extremely probable that the twins are uniovular.

The present report describes such an example of uniovular twins, both of them male; one was anencephalic and the other without malformation of any kind.

Case Report

The mother was aged 37 years and had had only one previous pregnancy; this had resulted in the birth of an healthy female infant. During her present

pregnancy there was no history of trauma or disease and she had received no inoculation. Exposure of the abdomen to *x* rays had been limited to a simple diagnostic procedure one week before delivery at which time the presence of twins including an anencephalic was confirmed. Her blood group was A rhesus negative, but during the course of pregnancy no rhesus antibodies had been demonstrated. The father was also aged 37 years and was in good health. There was no history of deformity occurring in the sibs, parents, or grandparents of either mother or father.

During the present pregnancy, vomiting had occurred early, and by the 30th week pronounced hydramnios and oedema of the ankles had developed. The pregnancy was terminated at the 35th week by artificial rupture of the membranes, and resulted in the spontaneous delivery of male twins (Figs 1 and 2) neither of which survived. The placenta, weighing 675 g., was expelled complete and consisted of one chorion and two amnions; from the form of the placenta (Fig. 3, overleaf) and the fact that the twins were of the same sex it was concluded that the twins were uniovular.

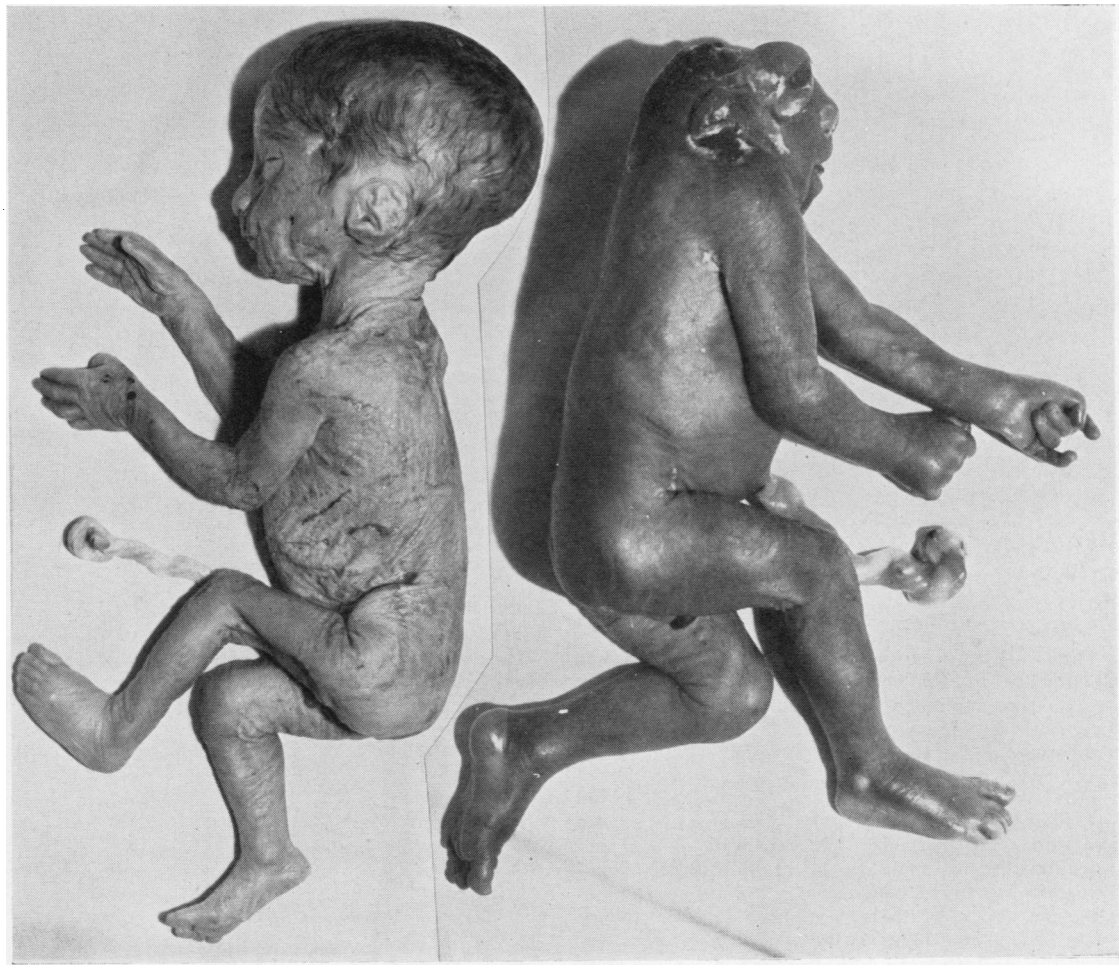

FIG. 1.—Twin 1, normal.

FIG. 2.—Twin 2, anencephalic.

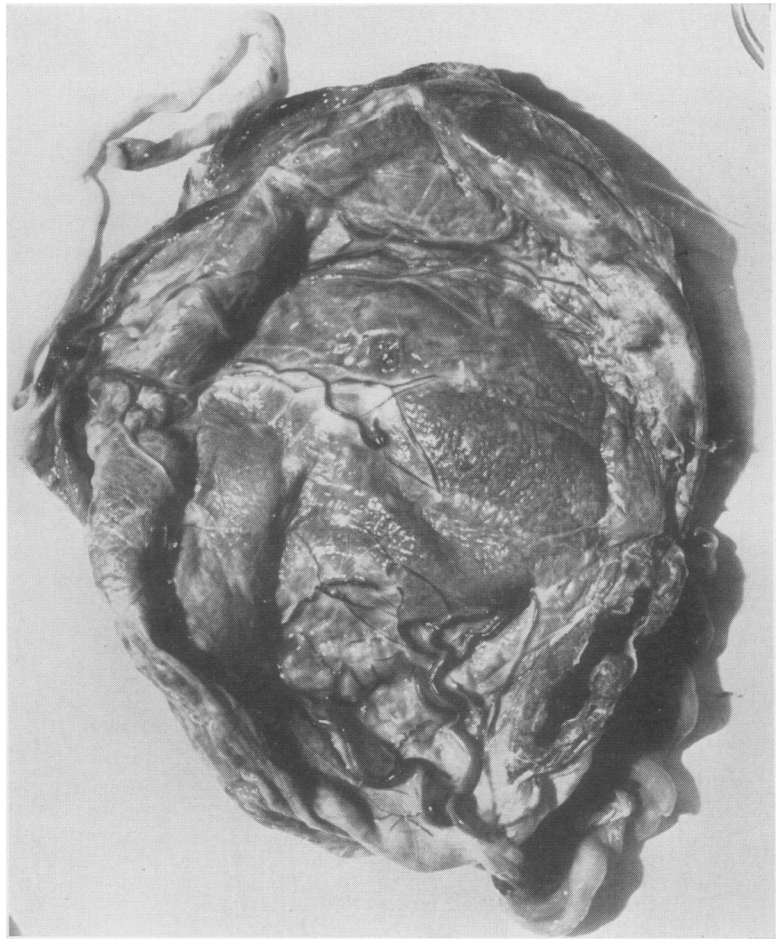

FIG. 3.—Placenta.

Post-mortem Findings:

Twin 1.—This male foetus weighed 526 g. and showed no obvious external or internal malformation. The weights of certain organs were, heart 4·1 g., liver 23·5 g., thymus 0·7 g., and paired adrenals 1·2 g. The lungs were unexpanded and airless.

Twin 2.—This foetus, also male, weighed 621 g. and had anencephaly with the facies characteristic of this condition. Over the lumbar spine was a tuft of hair and dissection revealed spina bifida occulta. No other malformation was found on external and internal examination. The heart weighed 8·6 g., liver 46·0 g., thymus 3·6 g., and paired adrenals 0·3 g. The lungs were unexpanded and airless, and histological examination failed to demonstrate in them any recognizable brain tissue.

SUMMARY

Anencephaly is described in one only of a pair of twins with a single chorion.

I am indebted to Mr. G. Davey of Shoreham for the photographs.

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