

CONGENITAL MALFORMATIONS IN A MONOAMNIOTIC TWIN

(Report of a case)

by

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Monoamniotic twinning has not been frequently reported in the Indian literature. There are hardly a few scattered reports. Thus, the estimates of frequency of this condition are not exactly known in the Indian literature. There have been frequent reports of monoamniotic twin pregnancies in the American literature since Quigley's original review of world literature in 1935. The condition is not an extreme rarity. It requires recognition; many more cases remain undetected or unreported.

Foetal mortality rate is relatively high in monoamniotic twinning, mainly due to cord complications and congenital malformations which are more common in multiple than in single gestation. The present case which has been studied by the author was a set of monoamniotic twins; one had congenital malformations, while the other was a normal stillborn macerated foetus.

CASE REPORTS

H. B., aged 20 years, 2nd gravida, 1 para expected to confine after 8 weeks, was admitted in the labour room for painless bleeding. Her general examination did not reveal any contributory finding.

Per abdomen examination revealed 28 weeks' size pregnancy, a small floating head

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presenting as vertex. Per vaginal examination revealed 5 cm. dilated, effaced cervix. Membranes were intact, and ruptured artificially. After a eight hour first stage of labour and a 30 minutes second stage, one male macerated foetus was delivered. The foetus had normal umbilical cord. Five minutes after, another foetus was delivered along with the placenta. This foetus had a very short cord and some marked congenital malformations. The weight of the normal macerated foetus was 500 Gms. and the weight of the abnormal foetus was 350 Gms. Congenital defects in the abnormal foetus were (1) the absence of external genitalia, (2) the absence of anal opening (3) non development of right hip, (4) left foot 6 toes, the right foot only a great toe and (5) umbilical hernia.

There was a single pale placenta with areas of congestion at various places. The diameter, circumference and thickness of the placenta were 14 Cm, 44 Cm. and 2 Cm respectively. The gestation sac was monochorionic and monoamniotic. The umbilical cords which were inserted close to each other, were showing almost a common origin. One of the umbilical cords was 47 cm long and 1 cm wide. That cord connected the macerated foetus to the placenta. The umbilical cord connecting the abnormal foetus to the placenta was only 3 cm long. Distal to that umbilical cord, there was a pyramid-shaped swelling (Umbilical hernia) with the liver and a few coils of intestine of the foetus in the sac. Each umbilical cord had three vessels but there was one common vessel connecting both the cords.

Comments

Logically, all monoamniotic twins take origin from single ovum and this has been

explained by Coulton (1947) and Corner (1955). The division of germ disc between day 8-13 after fertilization would result in twin foetuses in a single amniotic sac because the sac begins to form on day 8 after fertilization (Primitive unity theory). In some monoamniotic twinning it is supposed that originally there are two amniotic sacs and that the partition between these two sacs is broken in early foetal life due to pulsation of two cords lying together on either side of the partition. (Theory of primitive duality).

The incidence of monoamniotic twins as reported by various workers, is variable. It is estimated to be in the range of 1 in 1,000 to 1 in 90,000 deliveries. Muller and Rosenberg (as quoted by Boyle and Richter) estimated the incidence of 1 in 16,000 and 1 in 60,000 deliveries respectively. Ahlfield and Alfieri (quoted by Salerno 1959) reported an incidence of 1:168 and 1:256 twin pregnancies respectively, King (1952), reported 5 cases of monoamniotic twinning in the city of New Orleans in a period of 18 months while Potter and Crunden (1944) did not report even a single case in a review of 332 twin pregnancies at-Chicago-lying-in-hospital between a period of 10 years. The incidence of the pathology is not known in Indian literature.

Prior to the use of amniography, the antenatal diagnosis of monoamniotic twins was impossible. The condition had been suspected only after the delivery of the first twin, if there was prolapse of a twisted or knotted umbilical cord or if there was absence of the second amniotic sac. The routine use of amniography in twin pregnancies to look for an entity as rare as monoamniotic twinning has not obtained much importance.

According to Quigley (1935) the incidence of monstrosities is about 10 per

cent in monoamniotic twins. Pedlow (1961) found that 9.2 per cent of reported monoamniotic twins had congenital abnormalities. King (1952) reported 5 cases of congenital abnormalities out of 34 cases in the world literature while Salerno (1959) reported 4 cases of foetal abnormalities in 35 cases in the American literature. The author, in a recent review, has not been able to find a single report which is concerned with the malformation of genito-urinary system and gastrointestinal tract in a monoamniotic twin.

Foetal mortality has been estimated to be 68 per cent by Quigley who analysed 109 pregnancies for foetal outcome in monoamniotic twins. He estimated the chances of double survival to be 16 per cent. Raphael (1961) stated that the survival rate was 46 per cent 1961, and Wensinger and Daly (1962) found it to be 72.9 per cent. King estimated the foetal salvage of 56 per cent in world literature while Salerno observed 70 per cent survival rate in 35 cases which he reviewed in the American literature. A high foetal mortality rate in Quigley's report and a high survival rate in the subsequent reports shows that the efficiency in the management has been certainly improved since then. The clinical significance of identification of monoamniotic twins does not require further elucidation. The condition is not an extreme rarity and is recognized only at delivery. The danger of death to one or both of the foetuses from twisting or knotting of the umbilical cords which is very common, is great.

Summary

The occurrence of congenital malformation such as absence of anal opening and external genitalia along with some defects in the bones in a monoamniotic twin is reported.

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See Figs. on Art Paper V