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Structural Changes in Conjoined Birth: *A Case Study of Rare Complication of Multiple Pregnancy*

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ABSTRACT

We present a case of a 25 year old multiparons woman in her fifthe pregnancy. She had three previous spontaneous deliveries with one spontaneous miscarriage. Abdominal ultrasound revealed she had conjoined twins at an estimated gestational age of 37.1 weeks. She was delivered through caesarian section of a set of conjoined twins, joined in entire length of thorax and abdomen (thoraco-omphalagus). they died within 10 minutes of birth. Structural changes, obstetrical and gynaecological precautions associated with the case is discussed.

Conjoined twins known to the lay person as Siamese twins are rare and catastrophic obstetric event with the approximate incidence of 1:50,000 - 1:100,000 births. (Nkyeker K, 2002, Noel 2004, Wiilhch 1998). Few obstetricians will have the experience of managing this biologic abnormality. Approximately 40 - 60% of conjoined twins arrive stillborn and about 35% survive only one day. The overall survival rate is between 5 - 25%. Female siblings seem to have better survival rates than their male counterparts. Diagnosis and management of a case of term conjoined twins can be quite challenging. Early diagnosis, close prenatal management and proper route of delivery will assure the best possible outcome for mother and babies.

CASE PRESENTATION

The case involves an unbooked 25 year old multiparous woman in her fifth pregnancy. She had three previous spontaneous deliveries (1 male and 2 females) with one spontaneous miscarriage. Her last confinement was in 2004. She was unsure of her dates but presented to us with an abdominal ultrasound result done on the 27th September, 2006 which suggested that she had conjoined twins at an estimated gestational age of 37.1 weeks.

On examination she was in satisfactory general condition with stable vital signs. The abdomen was grossly enlarged with multiple fetal parts felt on palpation. Presentation of the leading twin was ill defined. Fetal heart sounds were normal. The result of the ultrasound was explained to her and she was counseled for elective lower segment caesarean section. She declined and absconded. She however presented as an emergency on the 2nd of October 2006. She was distressed with pain of uterine contractions, weak and moderately dehydrated. Vaginal examination revealed a cervix that was 8cm dilated with a thick rim. Three fetal legs were prescnting. She was admitted, rehydrated and prepared for an emergency caesarean section. The anaesthetic and paediatric teams on call were

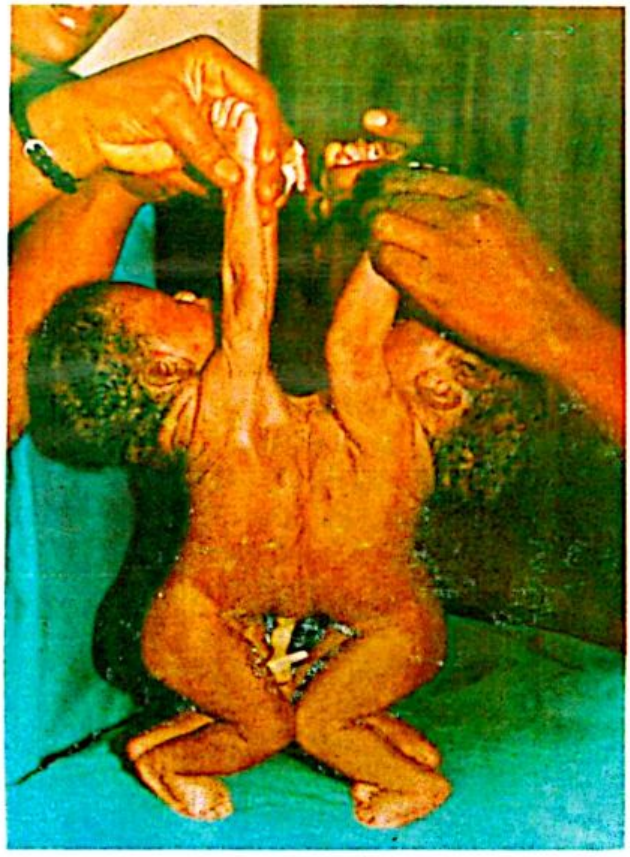

Fig 1. A case of Conjoined Twins joined at the thorax and abdomen.

informed. She was delivered of a set of conjoined twins, joined in the entire length of the thorax and abdomen (thoraco-omphalagus). They were both female and had a combined weight of 6.1kg. Apgar score at birth was 3. The twins failed to cry or improve on their Apgar score despite vigorous neonatal resuscitation. They died within 10minutes of birth. The father refused to give consent for a postmortem examination on the twins.

The patient was transfused with two units of blood postoperatively. She had an uneventful postoperative period and was discharged home on the 8th postoperative day.

DISCUSSION

Conjoined twins are very rare as a result; the diagnosis is often missed antenatally. In the case presented, the diagnosis was made antenatally and the woman was offered elective caesarean section which she refused. The findings at ultrasound which helped clinch the diagnosis in this case were an abnormally large thorax with two cardia closely apposed to each other with two separate heart rates and an abnormally large umbilical cord. Other classical ultrasound signs include, both fetal heads at the same plane, unusual backward flexion of the cervical spine, no change in relative position after maternal movement and manual manipulation and inability to separate both fetal bodies after careful observation. (Kalchbrenner et al 1987). These were not elicited in the case presented.

A plain abdominal X-ray will show) unusual backward flexion of the vertebral column of the fetuses and two heads at the same level. Our patient was not subjected to radiological investigation. Consideration for cost and possible radiation hazards to the fetuses made us prioritise our investigations.

There are two types of twinning Dizygotic (DZ) and Monozygotic (MZ) twinning. DZ twinning represents duplication of the normal process of conception, implantation and further development of the embryo, arising from fertilization of two ova from the same or opposite ovaries. Each fetus will have its own membranes, both chorion and amnion, and its own placenta. When the implantation sites happen to be close together, the placentas may become fused, but separate placental circulation will be maintained (Fisk 2007). The duplication of the membranes is known as dichorionic diamniotic. The fetuses may be like or unlike in sex and will have differences in genetic constitution. Our case was a case of monozygotic twinning both fetuses were of same sex. Both fetuses shared the same placenta and membranes. Unlike DZ twinning which is the duplication of normal development, MZ twinning is a gross departure from the normal process. The several varieties are determined by the time when splitting occurs in the embryo giving rise to different structural arrangements of the membranes.

If splitting occurs not later than the 8 - cell stage (3 days after fertilization) while cells remain unspecialized and thus retain their full potentiality, two separate blastocysts form and if both implant successfully a twin pregnancy results. There are separate implantation sites which mayor may not be close together, and the structural arrangement of the membranes and placentas (dichorionic, diamniotic) are exactly the same as in DZ twinning. However unlike in DZ twins, the fetuses will be identical. This early splitting at the blastomere stage accounts for one third of MZ twins (Fisk 2007). Our case was (monochorionic monoaniotic) with a single placenta.

If splitting is delayed until the inner cell mass is forming (4 - 7 days after fertilization) a single blastomere will implant with a single chorion giving rise to one placenta in which there is anastomoses between the two fetal circulations, but if the amnion has not yet differentiated, each embryo will develop its own amniotic membrane (Monochorionic diamniotic). This intermediate form of splitting, occurs in two thirds of MZ twins. The offspring will be identical. This was not the situation in the case presented. The fetuses though identical, shared a single amnion (Monoamniotic). When later splitting of the inner cell mass occurs after differentiation of the amnion but before the appearance of the primitive streak (8 - 12 days after fertilization), two identical fetuses will develop with a single amniotic cavity as well as sharing a single chorion and placenta with communication

between the two circulations (monochorionic monoamniotic) this accounts for about 1 % of MZ twins and carries a high perinatal mortality rate due to cord entanglement and twin-to-twin transfusion (Nkyeker 2002). The fetuses are not conjoined. This variety though close to the case presented, differs significantly in the sense that the fetuses are not conjoined.

Still later after primitive streak has appeared (from about 13 days after fertilization) incomplete splitting of the germinal disc is an extremely rare aberration that gives rise to conjoined twins (Fisk 2007) as in the case presented.

In view of the fact that vaginal delivery for term conjoined twins could be extremely hazardous (Aiyedun 2002, Omokhodion et aI2001), and that there is the prospect of saving some fetuses, it would appear desirable that an exact diagnosis be reached before the onset of labour and so permit the complication to be treated by elective caesarean section. Also the most experienced surgeon operating under ideal condition will reduce maternal morbidity and/or mortality.

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