A rare case report of conjoined twins: Thoraco-omphalopagus with anterior abdominal wall defect

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INTRODUCTION

Conjoined twin is an exceptionally rare congenital anomaly of two babies physically joined to each other, caused by partial separation of an early embryo after the 13th day of fertilization. Conjoined twinning occurs in 1 in 100 sets of monozygotic twins, 1 in 50,000 gestations or 1 in 250,000 live births.¹ Around 40% of the conjoined twins are still born and approximately 35% die within 24 hours of delivery.² Prognosis to some extent can be related to the vitality of the organs shared and precariousness of the accompanying congenital anomalies.

The cause of conjoined twins is unknown, but two theories have been postulated to explain the origin: fission theory is the traditional one. The fertilized egg is incompletely split, resulting in delayed separation of the embryonic mass after twelfth day of fertilization.

Fusion theory, in which the fertilized egg is completely separated, but the ‘stem cells’ of one twin fuse with the ‘like-stem cells’ of other twin, leading to fusion of both twins together.³,⁴

Termination should be offered in early pregnancy if conjoined twins are diagnosed antenatally to prevent the mother from further misery of having a malformed baby after the whole duration of pregnancy.

Overall, approximately 500 cases of conjoined twins are reported in the literature. Here we report a rare but unique case of conjoined twin (thoraco-omphalopagus) with anterior abdominal wall defect in one and unrevealed sex in other. Current report emphasizes on making an early diagnosis of conjoined twin pregnancy, so that it can be managed at the earliest according to the gestational age.

ABSTRACT

Conjoined twins are a rare deviation of monozygotic monoaomniotic twins which results from fusion of the two at any part of their body. The prognosis is not good and associated with high mortality. Here we report a case of conjoined twin which was referred to our tertiary care hospital as twin pregnancy with impacted breech in obstructed labour. Taking mothers condition into account, she was taken for emergency caesarean section, performing which we realised that it was a conjoined twin. The baby were still born and conjoined at thorax and abdomen (thoraco-omphalopagus) with anterior abdominal wall defect in one and unrevealed sex in other. Current report emphasizes on making an early diagnosis of conjoined twin pregnancy, so that it can be managed at the earliest according to the gestational age.

Keywords: Anterior abdominal wall defect, Caesarean section, Case report, Conjoined twin, Obstructed labour, Thoraco-omphalopagus

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CASE REPORT

A 20 year old G3P2 at 36 week gestational age with 2 full term vaginal delivery was referred to our hospital, from a CHC as an emergency case on account of impacted twins with breech presentation (Figure 1). Her last pregnancy was 2 years back and her current pregnancy was by spontaneous conception. Her index pregnancy was unsupervised and uninvestigated since the beginning such that the patient wasn’t aware that it was a twin pregnancy.

On admission, patient was pale, dehydrated and exhausted. Her BP was normal but pulse rate was 110 per minute. On per abdominal examination, uterus was full of fetus with multiple fetal parts felt. Fetal heart could not be localized with stethoscope or Doppler. On local examination, perineum and thigh of first baby and legs with feet of second baby along with placental tissue were lying outside introitus. On per vaginal examination, buttocks of first baby and thighs of second baby were felt in the birth canal. No active bleeding was present.

Patient’s investigations and cross match were send. After explaining poor prognosis for babies and taking high risk consent, patient was shifted immediately for emergency cesarean section. Lower segment cesarean section was performed under general anesthesia. Extraction of the babies was complicated, and it was at that point when we found out that it was a conjoined twin. An assistant was asked to push the baby upwards at the same time as attempting extraction. Feet of second baby were delivered followed by feet and body of first baby, eventually head of first baby and lastly head of second baby. The transverse incision of the uterus has converted into a T-shaped incision with 5 cm vertical arm extending down into the lower segment (due to the massively stretched out lower segment from obstructed labour). There was no injury to the bladder and the tear was stitched with restoration of complete hemostasis. Patient’s intraoperative and postoperative periods were uneventful.

It was a still born conjoined twin with a combined weight of 3.0 kg. First baby was male and the sex of other baby could not be revealed. Placenta was single with single umbilical cord ascertained. On clinical examination, the babies were fused from the level of xiphisternum downwards (thoraco-omphalopagus) with the fold reaching upto the perineum in second baby. First baby was male with anterior abdominal wall defect but the sex of second baby was not identifiable due to the skin fold covering the perineum (Figure 2). The babies were handed over to the relatives as they did not consent for further workup.

DISCUSSION

The first case of conjoined twin, born in 1811 who survived to adulthood was reported from Thailand, formerly called Siam, and hence the name Siamese twins. Conjoined twinning is a random event, which is not related to hereditary factors. No effect of maternal age has been found either. Conjoined twins share a single common chorion, placenta and amniotic sac, thus is exclusively monoamniotic monochorionic.

The outcome of monozygotic twinning and the number of layers of amnion and chorion depends on the time of division of fertilized ovum. As a rule, once any tissue is
differentiated, it is no longer capable of splitting. The chori
on differentiates on day 4 and the amnion on day 8
after fertilization. Hence, if zygotes divide within the first
72 hours after fertilization (which occurs in 25% of
monozygotic twins); two embryos, two amnions and two
chorions develop and a diamnionic, dichorionic twin
pregnancy evolves. Two distinct placentas or a single
fused placenta may develop. If division occurs between
4th and 8th day after fertilization when the chorion has
already differentiated (as in 75% cases), a diamnionic,
dichorionic twin pregnancy results. In less than 1% of
monozygotic conceptions, by approximately 8 days after
fertilization, the chorion and amnion have already
differentiated, and division results in two embryos within
a common amnionic sac that is monoamnionic, 
dichorionic twin pregnancy. But after 13 days of
fertilization, division of embryo is usually incomplete and
it leads to the development of conjoined twins of various
varieties.2

The medical term used to describe conjoined twins ends
with the suffix “pagus” from the Greek word “fixed”. Conjoined twins are classified according to the most prominent site of conjunction: thorax (thoracopagus), abdomen (omphalopagus), sacrum (pygopagus), pelvis (ischiopagus), skull (cephalopagus), and back (rachiopagus). Thoracopagus is the most frequent type. 75% of conjoined twins are seen as thoracopagus.3 The overall survival rate for conjoined twins is approximately 25%.9

Conjoined twins are reported as early as the 10th week of
gestation.80 When the diagnosis of the conjoined twins is
made, the type and severity of the abnormality should be
assessed with two- and three-dimensional ultrasound, CT
scan or MRI.5 When severe forms are diagnosed prior to
24 weeks gestation, termination via vaginal delivery
should be considered.11

Moreover, this case report emphasizes the limitation of
health facilities and lack of awareness among patients in
developing countries, where the patient didn’t bother for
even a preliminary ultrasound or antenatal checkup; and
hence focuses on the necessity of health facilities to be
catered to as many as possible.

CONCLUSION

Conjoined twins are an extremely rare developmental
anomaly with high perinatal mortality and morbidity.
Available modalities should be used to diagnose such a
pregnancy before viability and termination be offered to
the parents at the earliest. This can be made possible by
strengthening health care set up and improving patient
awareness towards early antenatal assessment as per
guidelines. This way we can reduce an unnecessary
burden of taking the pregnancy to term which carries
grave prognosis and adds to maternal morbidity as well;
as happened in our case.

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