

Thoraco Omphalopagus: An Intra Operative Diagnosis of Conjoined Twin Pregnancy

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Abstract

Background: Conjoined twins are a rare type of monoamniotic twins, estimated to occur in 1.5 per 100,000 births worldwide [1]. The incidence appears remarkably similar throughout the world, although the report is higher from Atlanta 1 per 20,000 births [2]. When twins are conjoined, the fusion occurs between the same body parts. Conjoined twins are classified as cephalopagus, thoracopagus, omphalopagus, ischiopagus, parapagus, craniopagus, rachipagus and pygopagus based on the site of fusion.

Case Report: Here we report a case of a 27-year-old Gravida 4 para 2 Abortion 1 mother who presented with labor and rupture of membrane. She had two previous cesarean deliveries. She did not have ultrasound scanning throughout her pregnancy. She was in active labor and an emergency Cesarean section was done. Conjoined twin (Thoraco-omphalopagus) was diagnosed during surgery when the fetuses were extracted from the uterus intra-operatively.

Conclusion: The case of thoraco omphalopagus conjoined twin delivered by cesarean section is presented with a literature review.

Keywords: Monoamniotic-Monochorionic Twins; Thoraco-Omphalopagus; Conjoined Twins

Introduction

Conjoined twins are a rare type of monoamniotic twins. The timing of post-fertilization division of the zygote determines placentation in twins. Monoamniotic, monochorionic placentation occurs with the division on day 8 to 12; Conjoined twins occur when monozygotic twins fail to separate into two fetuses as division occurs at or after day 13 post-fertilization. The ratio of females to males is 3:1, Two theories have been proposed to explain this observation: (1) the process of X-inactivation overlaps with the timing of monozygotic twinning and thus may directly contribute to the development of monozygotic twins and (2) the XX karyotype may confer a survival benefit [7]. The conjunction ranges from the simple joining of ectodermal tissues to the extreme case when one twin is contained within the other [6] the factors responsible for the timing of embryo division are not known. The use of assisted reproductive techniques appears to play a role as *in vitro* fertilization increases the frequency of monozygotic twinning. In some studies, manipulation of the zona pellucida, which is performed with intracytoplasmic sperm injection and assisted hatching, increases the frequency of monoamniotic twins [3]. The diagnosis should be suspected in first-trimester monoamniotic twin pregnancies when the embryo/fetus has the same lie persistently. Other findings, which are not all specific to conjoined twins, include fetal hyperextension, increased nuchal translucency or cystic hygroma, no sign of separate movement of the twins, juxtaposed embryos with a single midline cardiac motion, fewer limbs than expected, a single umbilical cord with more than three vessels, or both heads or breeches consistently at the same level to each other [4]. Color Doppler;

fetal echocardiography, and three-dimensional ultrasound examination can confirm the diagnosis and clarify anatomy, which is critical for assessing prognosis and pre-and postnatal decision-making. Fetal magnetic resonance imaging may be helpful to completely define shared anatomy and for surgical planning before delivery [5]. 40% to 60% of conjoined twins are stillborn, and almost 35% of live births do not survive 24 hours [9].

Conjoined twins are classified by the site of their most prominent union, which is ventral or dorsal in 87 and 13 percent, respectively. The abnormality is named with the suffix pagus, which means fixed. The distribution of ventral unions is as follows [8]: Cephalopagus - 11 percent, Thoracopagus - 19 percent, Omphalopagus - 18 percent, Ischiopagus - 11 percent, Parapagus (pelvis and variable trunk) - 28 percent. The following is the distribution of the less frequent dorsal unions: Craniopagus - 5 percent, Rachiopagus (vertebral column) - 2 percent, Pygopagus (sacrum) - 6 percent.

Thoracopagus fetuses face one another and have the major junction at the level of the chest with a common sternum, thoracic cage, diaphragm, and abdominal walls down to the umbilicus. Thoracopagus constitute 40% of conjoined twins. In these twins, 75% have conjoined hearts, 50% have a fusion of intestinal tracts and virtually all have a shared liver [10].

Case Presentation

This is a 27-year-old Gravida 4 Para 2 (1 alive, 1 infant death) Abortion mother. Her GA is 38+4 weeks calculated from reliable LNMP. She presented to the labor ward with a complaint of labor pain of 6 hours and rupture of the membrane of 5 hours duration. Her entire antenatal care was at a health Centre. Twins were not detected; neither did she have an ultrasound during the care. She had two previous Cesarean sections at Jimma Medical Center for unknown indications the 1st was 6 years back and the 2nd was 3 years back. The first infant died at 2 months of age. There were no antenatal complications. She has no personal or family history of Diabetes Mellitus or Hypertension or any other chronic medical illnesses. The pregnancy is wanted and supported.

On physical examination, she was in labor with normal vital signs. Abdominal Examination confirmed; the uterus was a term-sized longitudinal lie and Breech Presentation. The fetal heart rate was normal at 156, there was a suprapubic transverse old surgical scar and she had 3 contractions in 10 minutes which lasts for 30 - 35 seconds. On digital pelvic examination cervix was 6 cm dilated and both feet of the fetus were felt at the internal os. Obstetric ultrasound was not performed and with the final diagnosis of Early Term Pregnancy, Active First stage of labor, and two previous Cesarean scars she was taken to the operation theatre for emergency cesarean section. Upon the trial of delivery, there was resistance for extraction of the fetus by breech delivery, then by examination of fetal parts, it was noted to be conjoined twins and fetuses extracted by gentle manipulation of the fetal parts. The outcome was conjoined Twins with the weight of 4000 gm together with a good Apgar score. There was no Extension or need for making an inverted T incision. Newborns were given basic essential newborn care and transferred to Neonatal ICU. They were conjoined at the level of the chest to the umbilicus, sharing a single umbilical cord with multiple blood vessels in it and also share the same sternum both having a single nipple each. They have symmetrical 4 extremities and well-formed male external genitalia each. After they stayed in the Neonatal Intensive care unit for about 48 hours, both of them passed away with a possible cause of death being a cardiorespiratory failure due to the underlying condition.



Figure 1: The twins attached together from the mid-sternum down to the umbilicus.

Discussion

The classification of conjoined twins depends upon the site of union and they are classified into Thoracomphalopagus (chest, abdomen, or both), thoracopagus (chest), omphalopagus (abdomen), Pygopagus (buttocks), Ischiopagus (ischium), and Craniopagus (head) depending upon the site where they are united. The analysis of survived and aborted fetuses showed that most of the conjoined twins are females with an M: F ratio of 1:3. The survival depends upon the type of union but overall survival rates reported range between 20 - 25% [11]. The exact cause of this is not known but it is widely accepted that conjoined twins occur due to incomplete splitting of monozygotic twins after 12 days of embryogenesis. The risk factors may include intake of clomiphene citrate, valproate, and griseofulvin during the periconceptual period.

No mutations are yet identified to be associated with conjoint twins. Some researchers proposed the theory of abnormal X-chromosome inactivation while some other researchers have refuted this theory. There is no evidence of increased recurrence risk in case of pregnancy complicated by conjoint twins [12].

The presentation and prognosis depend upon the type of conjoined twins. In the case of craniothoracopagus where there is a union at the level of cranium and thorax prognosis is poor as there is only one brain and surgical separation is not an available option but fortunately this type of conjoint twins are rare. Craniopagus twins may be partial or complete depending upon whether they significantly share the dural venous sinus system. In the bicephalus conjoined twins, there is the presence of 2 heads on 1 trunk. In cases of omphalopagus twins, the fusion occurs at the abdominal wall. Omphalopagus may be isolated or associated with a union at the level of the thorax in which case they are called thoraco-omphalopagus. These types of conjoint twins may undergo complex surgery for separation if they are found not to be sharing vital organs. Other less common forms of conjoined twins include parapagus (union at the trunk), pyopagus (union at sacrum), and rachipagus (fusion at vertebral columns) [13]. In our case it is thoraco omphalopagus, but it was difficult to assess shared visceral organ since imaging study is not done.

Early detection of this condition is of paramount importance to provide the parents with an option of safe termination of pregnancy. Ultrasound is a safe and quick method by which conjoint twins can be diagnosed even in early pregnancy (as early as 8-10 weeks). The twin fetuses maintaining a fixed alignment is a sign of conjoint twins. This may be associated with polyhydramnios [14]. Transvaginal scan may provide a better view and the ability to separately see different parts of the fetuses may help in early diagnosis. Once the diagnosis of conjoint twins is confirmed by ultrasound it is of critical importance to find out the exact site of union, several vital organs including brain, heart, and liver, and the presence of any associated congenital anomaly like congenital heart disease, urogenital anomalies or neural tube defects. Magnetic resonance imaging has the advantage of not being operator-dependent and having better tissue contrast. Moreover, MRI can pick up the presence of associated congenital anomalies at an early stage. The option of medical termination of pregnancy can be offered to the family if diagnosed before 20 weeks of gestational age [15].

In our patient, the diagnosis is made intra-operatively after the patient presents with advanced labor and having previous two cesarean deliveries. An emergency cesarean delivery was performed and the newborns were transferred to Neonatal intensive care units and after staying for about 48 hours both of them passed away with the possible cause of death being a cardiorespiratory failure due to the underlying condition.

Conclusion

Conjoined twins are a rare and complex occurrence. Associated high perinatal mortality makes an early diagnosis by ultrasound an essential requirement to provide parents with an option of safe medical termination of pregnancy. But if the patient presents late, Cesarean delivery is mandatory.

Competing Interest

The authors declare that they have no competing interest.

Consent

Written informed consent was obtained from the mother for publication of this case report and accompanying image.

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