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Case Report

Conjoined Twin: Late Presentation and Challenges in Management

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Abstract
Conjoined twin is a rare occurrence and its management presents with unique challenges. We report a case of a 23 year primigravida with a thoracophagus conjoined twin with complex cardiac fusion who presented to us at 34weeks of gestation. Condition was diagnosed by ultrasound; degree of fusion was assessed by computed tomography. Inspite of poor prognosis of the babies, patient was delivered by lower segment cesarean section to avoid maternal genital tract trauma.

Keywords: *Cesarean section, conjoined twins, prenatal diagnosis, ultrasound*

Introduction
Conjoined twins are one of the rarest congenital anomalies occurring in approximately 0.2:10,000 pregnancies but only 0.5:10,000 live births and its management is one of the greatest challenges to the modern era pediatric specialist including pediatric surgeons, anesthetists, neonatologists, urologists, neurosurgeons, and orthopedic surgeons [1-4]. This anomaly can be classified according to the type of twins′ fusion. Various organs can be fused making the separation difficult. Conjoined twins are usually diagnosed antenatally by ultrasound which can be detected as early as 12 weeks and can be confirmed by 20 weeks. Detailed fetal echocardiography is necessary to counsel the parents during pregnancy. When a diagnosis is made around 50-70% of parents will opt for termination [5]. Definite indications for terminations include extensive cardiac or cerebral fusions or when the expected deformity following separation is unacceptable but nevertheless termination should be discussed as an option for all conjoined twins as overall outcome is poor. Early termination is minimizing the morbidity to the mother associated with later termination and is preferable [6].

Case
A 23 years primigravida at 34 wks. of POG presented to antenatal outpatient department for decreased fetal movements. She had no previous antenatal visits. On clinical examination, the patient was normotensive with overdistended abdomen and presentation was difficult to appreciate. Ultrasonography showed conjoined twin which was thoracophagus variety with BPD corresponding to 34 weeks' period of gestation and only one cardiac activity with fetal bradycardia. Ultrasound of the abdomen revealed sharing of the extra hepatic biliary system, common bile duct and part of the intestine as well. There was partial cardiac fusion as well. Computed tomography showed similar findings. Since she was in

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advanced gestation confinement was planned by caesarean section. The patient and spouse were counseled regarding the prognosis of the babies which was poor and she was taken up for emergency cesarean section under spinal anesthesia. Midline vertical skin incision was given while a transverse lower segment uterine incision was given. She delivered conjoined twin female babies the combined weight being 2.8kg. Babies were having bradycardia and were gasping and as parents had consented against resuscitation no such attempt were made. The babies died within half an hour of life. The post operative period of the mother was uneventful.

Discussion

The diagnosis of conjoined twins should be made antenatally and can be done by ultrasound as early as 12 weeks of gestation and confirmation by a repeat second trimester anomaly scan at 16-22 weeks which also helps to define the site and extent of joining.

The ultrasound features that suggest the diagnosis are constant relative position of fetus over time with the head and other body parts at the same level persistently; inseparable body contours; fetus facing each other with hyperflexion of the cervical spines; fewer limbs than expected; single umbilical cord with more than three vessels; shared organs and sometimes it can be straight forward when fusion of fetuses is detected [4,7].

In our patient, unfortunately she had no previous antenatal visit so conjoined twins remained undiagnosed till late third trimester and was picked up by ultrasound at 34 weeks by features described previously. The poor prognostic factors for survival of conjoined twin in our patient were: late presentation when one twin was dying due to which proper evaluation antenatally was not possible, nor could the patient be transferred to centre of advanced pediatric surgical care for complete evaluation. The most important factor was the type of twining. It was a thoracophagus twin with partial cardiac f\usion though complete evaluation was not possible. Data from previous studies show that 90% of thoracophagus twins have common pericardial sac and there is almost always a degree of cardiac fusion. In 50% a common small intestine and in 25% a shared biliary system may be present [8].

The most important evaluation prenatally is the fetal echocardiography as the degree of cardiac fusion and severity of associated anomalies determine the post natal viability and also because it is easier to visualize the anomalies in utero as amniotic fluid acts as a buffer [7]. Studies done have shown that separation in such cases of thoracophagus twins with complex cardiac fusion is usually unsuccessful and the general consensus is to counsel the parents and give option for termination of pregnancy at the earliest [9]. If diagnosed early patient can undergo medical termination of pregnancy in second trimester. But in our case as the patient presented late we had to take her up for cesarean section to avoid trauma to maternal genital tract.

Vaginal delivery for conjoined twins has been reported but this is mainly for small preterm twins if they are considered non viable or because prenatal assessment of unfavorable anatomy has been made. But this mode is often fatal for twins and carries risk for the mother due to trauma, obstructed labor, dystocia and uterine rupture [10] hence even if the prognosis of twins is dismal the choice of mode of delivery is cesarean section in third trimester.

The cesarean section for conjoined twins also poses with unique challenges. It can be performed under regional block. If a regional block fails conversion to general

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anesthesia may be opted for. Consideration should be given to the skin and uterine incision. Like any other difficult cesarean vertical skin incision is preferable and in general classical cesarean section is preferred as incision can easily be extended and will hence avoid trauma to the delicate vascular connections of the twins [6].

In our case as prognosis of the twins was unfavorable and operation was undertaken for the mother a vertical skin incision was made but babies were delivered by a transverse incision in the uterus by breech extraction. We were prepared for management of post partum hemorrhage which is expected in such cases but luckily no such complication occurred in our patient.

Regular antenatal care is required and early diagnosis of conjoined twins by ultrasound prevents unnecessary intervention in mother due to diagnosis in late gestation. Surgeries for separation of conjoined twins is still a distant reality for most of our patients but an accurate prenatal diagnosis is crucial in counseling the parents and timely referral to centers with advanced pediatric care for a few affording couples. Cesarean section appears to be the safer route for termination in late pregnancies even for non salvageable babies keeping in view the safety to the mother.

Figure 1: Thoracophagus conjoined twins

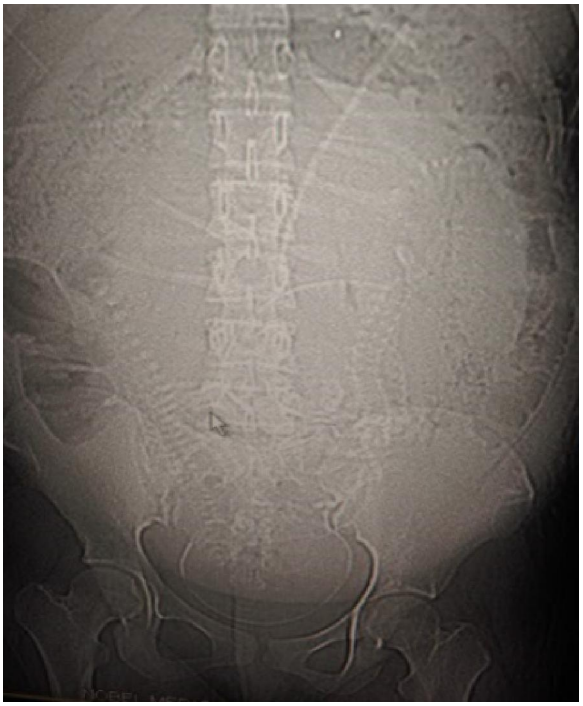

Figure 2: X-Ray of the mother with conjoined twins in utero showing

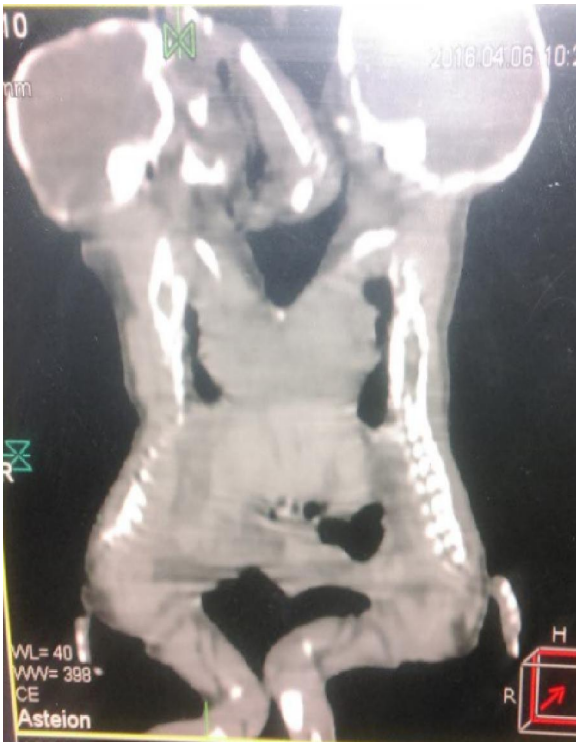

Figure 3: CT scan of the conjoined twins post delivery showing level of fusion

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