INTRODUCTION

Conjoined twinning (CT) is a rare phenomenon caused by the abnormal embryological development of monozygotic twins. The exact mechanism by which CT is formed is not known. Two theories exist: The fission theory (incomplete separation of the fertilized ovum) versus the fusion theory (early reattachment of two separate ovums).[2]

The incidence of CT is about 1 in 50,000 to 1 in 100,000 births,[1] or about 1 in 400 monozygotic twin births.[3] There is a female:male predominance of 3:1.[3,4] CT was broadly classified by Spencer[5] into 8 types: Cephalopagus (11%), thoracopagus (20–40%), omphalopagus (18–33%), ischiopagus (6–11%), parapagus, (28%), craniopagus, (2%) rachipagus (<1%), and pyopagus (18–28%).[6]

However, each case of CT has to be studied individually to recognize the site, type, and extent of the union, and the presence of vascular connections or malformations.[7] The prognosis, especially vitality and separability of each case of CT depends on the characteristics previously mentioned.[2]

CASE REPORT

A 17-year-old primiparous woman with a 26-week gestation, was referred to the hospital in active labor. Her antepartum course has been significant for diagnosis of current gestation at 12 weeks with only one prenatal consult at a primary care clinic, irregular ingestion of folic acid, and one obstetrical ultrasound by a radiologist that reported a 24.3 weeks monochorionic, monoamniotic twin pregnancy with both products of conception alive and active, heartbeat of the first fetus 154 beats/min, heartbeat of second fetus not measurable, with normal amniotic fluid.

Her gynecological history
Menarche age 14, 28 × 5, no history of the sexually transmitted disease or cervical cancer screening. After proper
ultrasonographic scan the diagnosis was a 26 week old pregnancy with thoracopagus twins in an adolescent (17 year old female) in active labor, twins were sharing one heart. The rest of her history is insignificant for the case.

The patient begins with irregular, painful uterine contractions 5 h before her referral.

On examination, the patient was alert and oriented, gravid, with palpable uterine activity, uterine fundus of 29 centimeters, vaginal exam with 7 centimeters dilatation, 90% effacement, fetal foot palpable, and intact membranes.

At her arrival we did an ultrasound scan finding a twin pregnancy, identifying two thorax sharing one heart, approximately 900 grams each, with a gestational age of 26.1 weeks [Figure 1].

**Final diagnosis**
Thoracopagus twins of 26 weeks gestational age in a primiparous 17-year-old female in active labor.

**Procedure plan**
After proper identification of the thoracopagus twins, and diagnosis of the single heart being shared, considering the present active labor and risks involved on proceeding with a vaginal delivery, a multidisciplinary team, including geneticists, neonatologists and obstetricians was assembled to perform a cesarean section delivery [Figure 2].

**Pregnancy outcome**
A cesarean section with a Kerr incision was used to obtain female thoracopagus twins in breech presentation [Figure 3], gestational age by Ballard score of 26 weeks, weight of 1450 g, first fetus with cleft palate, [Figure 4] the second fetus with cleft lip and cleft palate [Figure 5], both with brachydactyly, resulting in spontaneous death 30 min post-delivery. The family did not accept the further assessment of the twins.

**DISCUSSION**
CT, an infrequent and spontaneous event, poses a challenge to obstetricians in making a proper diagnosis. Two theories to explain this phenomenon exist: The fission theory versus the fusion theory.

The fission theory is based on the timing of division of the fertilized ovum, used classically to describe the chorionicity of all monozygotic pregnancies: Division from the time of fertilization to day 3 post-fertilization produces a dichorionic diamniotic pregnancy, division from day 4 to day 7 results in a monochorionic diamniotic pregnancy (in normal embryogenesis, the chorion begins to form on day 3), division between day 8 and 13 gives a monochorionic monoamniotic pregnancy (the amnion usually forms between day 6 and 8), and finally, division beyond 13 days results in CT (after the formation of the primitive streak); however, the exact mechanism is not known.
The fusion theory opposes this theory because it lacks definitive evidence and does not explain cases of atypical twinning (chimeric twins, phenotypically discordant monozygotic twins, mirror-image twins, polar body twins, vanishing twins, hydatiform mole with coexistent twin, fetus papyraceus, fetus-in-fetu, superfetation, and superfecundation), and proposes that CT is a result of the fusion of two originally distinct mono-ovular embryos. However, neither theory has concrete evidence.

Several authors mention that no significant associated genetic and environmental risk factors have been identified, however, Mutchinick et al. stratified ethnicity in four categories (Anglo-Saxon/Caucasian, Chinese, Latin American, and Latin European) with statistical differences observed with higher prevalence of CT in Latin Americans than in the other 3 categories.

Overall, 40-60% of CT are stillborn and 35% die within 24 h of birth. Results of several studies suggest a 2-3-fold increase in congenital anomalies in monozygotic twins, with 50% of CT presenting structural anomalies of major organs. The most frequently reported anomalies are of the genitourinary tract (19.8%), central nervous system (18.9%), neural tube defects (9.9%), hydrocephalus (3.6%), and microphthalmia (0.9%).

The diagnosis of CT is frequently made by ultrasound during the first trimester, and can be made as early as 12 weeks gestation with some authors describing the diagnosis at 7 weeks gestation, but with higher false positives cases. At around 8 weeks of gestation, fetal activity increases and it is easier to differentiate between a monochorionic, monoamniotic twin, and CT. Signs that suggest CT are the presence of 3 or more vessels in the umbilical cord, fewer limbs than would be expected, no change in relative positions of twins, hyperflexion of the spine, a single yolk sac, and bifid appearance of the fetal pole.

After the initial sonographic imaging in early pregnancy, there must be more advanced imaging of the CT to improve anatomic detail and physiologic information. Magnetic resonance imaging has superseded imaging by computed tomography due to its high radiation burden.

Specifically to thoracopagus CT, a detailed cardiac assessment is necessary through fetal ECHO for two reasons: The overall increase in congenital heart disease in CT and to evaluate the degree of cardiac fusion typically seen in thoracopagus CT. There are four possible cardiac anatomy variations: (1) Separate hearts and pericardium, (2) separate hearts but common pericardium, (3) fused atria and separate ventricles, and (4) fused atria and ventricles. Approximately 80-90% of thoracopagus CT are not candidates for surgical separation, as 90% share a common pericardium and 75% share a heart. Ventricular union has never been successfully separated, with both twins surviving and only one case has been reported of separation of twins with the atrial union.

Besides a conjoined heart, thoracopagus twins share a liver in 100% of cases, biliary tree in 25% of cases and small intestine in 40% of cases, therefore the hepatic venous draining must be evaluated and must be separate for the CT to be eligible for separation.

As previously mentioned, obstetrical management must be individualized depending on the exact determination of the affected structures and estimated prognosis.

In general, CT have a poor prognosis and many patients consider elective termination of pregnancy in the first or second trimester. The mode of delivery in second-trimester deliveries has been cesarean section because of the risk of maternal soft tissue injury, labor dystocia, and uterine rupture with a vaginal delivery. However, several case reports exist describing successful vaginal deliveries of the second trimester.
trimester and term CT in multi and primiparous women, without complications.\textsuperscript{[10,11]}

If a vaginal delivery is chosen, the medical team should be comfortable with performing destructive fetal procedures, and there should be an operating room available for emergencies.\textsuperscript{[11]} It is important to mention that Mitchell et al. administered fetal intracardiac potassium chloride to avoid live births of nonviable fetuses, with the added advantage of the softening of fetal parts to ease manipulation.\textsuperscript{[11]} The authors of these cases conclude that if fetal survival is not an issue, and maternal trauma less likely, vaginal delivery is an appropriate and safe option,\textsuperscript{[10]} thereby reducing the risks and morbidity associated with an operative delivery.\textsuperscript{[11]}

**CONCLUSION**

A case of CT was presented, diagnosed in a primiparous adolescent female with low socioeconomic status without adequate prenatal care, at the time of active labor. The diagnosis was made with a routine ultrasound that is performed on all patients arriving at the hospital in labor, demonstrating the importance of ultrasound screening at admission in this population. An obstetrical decision was made to perform a cesarean delivery to avoid soft tissue damage, dystocia, or a destructive fetal procedure. The neonatal outcome was poor due to the presence of a shared heart. It is important to have adequate prenatal care and early diagnosis to improve obstetrical outcomes.

**REFERENCES**