Early prenatal diagnosis of parapagus conjoined twins

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Abstract

Conjoined twinning occurs in 1/100 of monozygotic twins, 1/50,000 gestations and 1/250,000 live births. It is the consequence of a division event at the primitive streak stage of the human embryonic development, about 13-14 days after fertilisation, in monochorionic monoamniotic gestations. A healthy pregnant woman, Gravida 2 Para 1, was admitted into our Fetal Medicine Unit to perform the first trimester ultrasound. A diagnosis of conjoined parapagus twinning based on ultrasound features was made at 11 weeks of gestation, and the couple decided to terminate the pregnancy. The ultrasound showed two independent skulls and hearts, a shared spine below the thoracic level, and a shared stomach. The pathological findings were slightly different, showing two independent stomachs draining into a common duodenum. The karyotype was 46 XY. Early prenatal ultrasound may provide a window to counsel the family and to offer an early termination of pregnancy.

Introduction

Conjoined twinning consists on a rare obstetrical phenomenon that represents the most extreme complication of monozygotic twinning. The estimated incidence is 1/50,000 to 1/200,000 livebirths, and is more common in Africa and Southwest Asia, for unknown reasons.[1] There seems to be no association with maternal age, race, parity, or heredity and the risk of recurrence is negligible.[2] Two theories have emerged to explain conjoined twinning: It could be a consequence of either the embryo division after 14 days of the initial formation, with incomplete splitting of the embryonic axis (*incomplete fission theory*) or it could occur after the cellular fusion stimulated by stem-cells with similar characteristics in both twins (*fusion theory*).[3] Currently, the fusion of monozygotic twins is no longer believed to be the basis of conjoined twinning.[4]

The risk factors for conjoined twinning are not yet fully understood. An increase in the incidence of monozygotic twinning occurs in pregnancies after induced ovulation with exogenous gonadotrophins. It also has been reported in pregnancies that occurred within 6 months of stopping oral contraceptives. It has been hypothesised that in these situations, there is an abnormal uterine environment that leads to abnormalities of zygote division, but the mechanism remains unknown.[4] As the prevention of monozygotic pregnancies in general, and conjoined twinning in particular, remains impossible, the early ultrasound diagnosis in the first trimester provides an opportunity to counsel the couple and to offer early termination of pregnancy.

Case Report

A healthy 36-year-old pregnant woman, Gravida 2 Para 1, with a previous normal pregnancy that culminated in a spontaneous delivery, with light smoking habits of two cigarettes/day, with no history of congenital or hereditary disease in her family, was admitted into the Fetal Medicine Unit of the authors to perform the first trimester ultrasound regarding the combined approach for aneuploidy screening. The body mass index was 22.5 kg/m^2 and the blood type was A+. The pregnancy was spontaneous, with no use of ovulation inducing agents. There was no previous exposure to any medication.

The ultrasound, performed at 11 weeks and 2 days, revealed parapagus-dicephalus conjoined twins (Figure 1) with two separated skulls and hearts, a single abdominal cavity with an apparent common stomach (Figure 2), four superior limbs and two inferior limbs. Additional findings were the fusion of the spine at the thoracic level, with dorsum-lumbar and sacrum union. The nuchal translucency was above the 90th percentile for both twins.

The couple decided to terminate the pregnancy after being given detailed information about the neonatal prognosis. The procedure was done with 200 mg of oral mifepristone, followed by a dose of 800-µg misoprostol applied in the vaginal *cul-de-sac* 24 h later. Expulsion of the conjoined twins happened 4 h later, with intact umbilical cord and placenta (Figure 3) The karyotype analysis, performed in the postnatal period, showed a normal male karyotype, 46 XY. The embryopathological study showed two independent heads and necks, two chests with a common diaphragm, four lungs with normal morphology, two independent hearts each with an independent pericardium, fusion of the dorsum-lumbar spine and a single dysplasic sacrum. No cardiac or central nervous system malformation was described. Regarding the abdominal cavity, the pathological analysis was different from the ultrasound findings, showing two stomachs draining to a common duodenum, a single small intestine, a single microcolon and a shared liver, pancreas and gallbladder. The urinary system encompassed two kidneys with normal morphology for each fetus and two independent bladders.

Discussion

Conjoined twinning represents the extreme form of monozygotic twinning. Risk factors continue to be misunderstood, although women who have undergone induced ovulation and IVF *(in vitro fertilization)* seem to be at greater risk, as do women from populations with high levels of endogamous relationships.[5]

Monochorionicity seems to be a teratogenic phenomenon on its own, resulting in more congenital anomalies in these fetuses, with 25% of congenital anomalies in mono-

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chorionic-monoamniotic gestations. Even in conjoined twins without shared organs, the incidence of congenital anomalies is about 80%.[6] This complication affects female fetuses at a ratio of 3:1, for unknown reasons. Below is a rarer case with a male karyotype. Spencer suggested eight types of conjoined twins, based on the level of the fusion: omphalopagus (umbilicus), thoracopagus (chest), cephalopagus (head), ischiopagus (hip), craniopagus (skull), rachipagus (spine), pygopagus (buttocks) and parapagus (side). Spencer also suggested that conjoined twins should be classified by the proposed site of union in ventral (joined over a single yolk sac with a shared abdomen and umbilicus) and dorsal (joined in the neural tube with a separate abdomen and an umbilical cord for each twin).[7] According to the Spencer classification, our case refers to dicephalic parapagus conjoined twins with ventral fusion. Chih-Ping Chen *et al.* reported an incidence of parapagus conjoined twins of 13.5% in a series of 74 cases, establishing the rarity of this condition.[7] In this group, the fusion happens in the lower part of the body, usually in the lumbo-sacral region, while the region facing this this is often duplicated, as we can see in our case by independent organs for each fetus above the duodenum.[4] This type is associated with two trunks that gradually merge in their caudal regions. In a review of parapagus twin cases, the major aspects of these models of twinning is the doubling of all cranial structures, two vertebral columns as far as the lumbosacrum level, two hearts, with one supplying one half of the body and two sets of lungs. Outside the pyloric sphincters, the alimentary tract fused and both gut and the urogenital systems are common.[8] This type of conjoined twinning is different from the more common thoracopagus or thoraco-omphalopagus fusion, in which the chest and the abdomen are fused ventrally, with two independent heads facing each other.

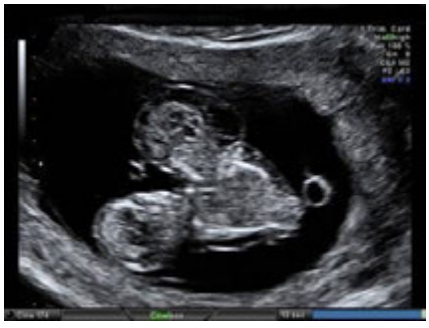

Figure 1. Parapagus - dicephalus conjoined twinning.

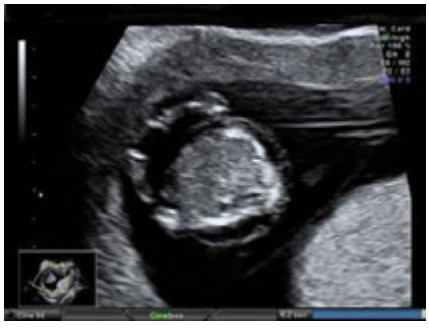

Figure 2. Parapagus conjoined twinning - abdominal wall.

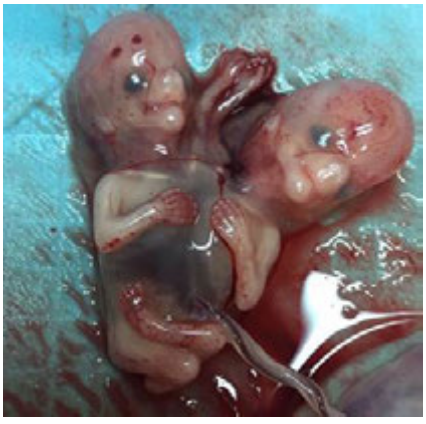

Figure 3. Parapagus conjoined twinning - postnatal anterior view.

Congenital anomalies are often found in the set of conjoined twins, observed in more than 60% of cases.[9] The malformations more frequently reported are those affecting the genitourinary tract, the neural tube defects, hydrocephalus, microphtalmia and those from the musculoskeletal system, such as polydactyly. Gastrointestinal atresia and facial clefts are also common.[9] In the first trimester ultrasound these malformations may be difficult to discern. In fact, in the first trimester, only 25% of conjoined twins had an abnormality detected in the ultrasound in the work of Chih-Ping Chen *et al*., including increased nuchal translucency, hydrops fetalis, abdominal wall defects, neural tube defects, congenital heart defects or renal agenesis.[7] The only abnormality that we found in the ultrasound apart from the fusion of the fetus, was an increased nuchal translucency.

There are some ultrasound signs that could help the diagnosis of conjoined twins, such as the presence of both fetal heads in the same plane, unusual backward flexion of the cervical spine and no change in the relative position of the fetuses after maternal movement. [1] Diagnosis can be made as early as eight weeks of pregnancy for some authors, but accurate evaluation of common structures is not possible. Some signs, such as a single extra-amniotic vitelline vesicle or a single embryo with bifid appearance before 10 weeks, can indicate the diagnosis of conjoined twins.[2]

In our experience, body fusion is easy to identify in the first trimester ultrasound, between 11-13w+6 days, and allows a diagnosis to be made early and the couple to be counselled early on. We have presented a rare case of dicephalic parapagus on twins, with male karyotype, where the first trimester ultrasound achieved an important overlap with embryopathological findings.

Conclusions

The first trimester (11-13w+6days) ultrasound is the best method for diagnosing conjoined twins early in pregnancy. The diagnosis remains easy, even if some of the congenital abnormalities cannot be seen at these gestational stages. Increased nuchal translucency is common, even in fetuses with two independent hearts and no cardiac congenital abnormalities in the embryopathological study. The early diagnosis of this condition is mandatory to allow an early termination of pregnancy.

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