

Cephalothoracopagus Janiceps Dissymmetricus with Gastroschisis, a Very Rare Form of Conjoined Twins: Case Report

Gastroşisizli Sefalotorakopagus Janiseps Disimetrikus, Çok Az Görülen Bir Yapışık İkiz Formu

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ABSTRACT In 1:50 000 to 1:100 000 births, conjoined twins occur, caused by incomplete division of the embryonic disc more than 13 days after fertilisation. We present a case of cephalothoracopagus janiceps dissymmetricus with gastroschisis, a very rare form of conjoined twins, which was diagnosed at 13th weeks of gestation. Early prenatal diagnosis of conjoined twins allows better counselling of the parents regarding the management options, including continuation of pregnancy with post-natal surgery, termination of pregnancy or selective fetocide in case of a triplet pregnancy. The cephalothoracopagus twin's prognosis is extremely poor because surgical separation is not an option. Thus early prenatal diagnosis of cephalothoracopagus twins is important to provide an opportunity for pregnancy termination if desired. Autopsy confirms the prenatal diagnosis.

Key Words: Ultrasonography, prenatal; prenatal diagnosis, twins, conjoined, gastroschisis

ÖZET Fertilizasyon sonrası embriyonik diskin inkomplet ayrılmasına bağlı 1/50 000 ile 1/100 000 doğumda bir yapışık ikizler görülür. Bu raporda çok az görülen ve 13. gestasyon haftasında tanı konulan gastroşisizli yapışık ikiz sefalotorakopagus janiseps disimetrikus olgusunu sunduk. Yapışık ikizlerin erken tanısı sayesinde, postnatal cerrahisi ile gebeliğin devamı, gebeliğin terminasyonu veya yalnızca fetosit uygulamasını içeren yönetim opsiyonları aileye daha iyi danışmanlık verilmesini sağlar. Cerrahi ayrımı olmadığı için sefalotorakopagus ikizlerinin prognozu çok kötüdür. Bundan dolayı sefalotorakopagus ikizlerinin erken prenatal tanısı gebelik sonlandırma seçeneği sunduğu için önemlidir. Otopsi prenatal tanıyı konfirme eder.

Anahtar Kelimeler: Prenatal ultrasonografi; prenatal tanı, yapışık ikiz, gastroşisiz

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Division of the embryonic disc more than 13 days after fertilisation is usually incomplete and results in conjoined twins.^{1,2} This occurs in 1:50 000 to 1:100 000 births. Seventy percent of the embryos are female. No maternal age effect has been noted, which is consistent with the fact that monozygotic twinning is not affected by maternal age.³ An increased risk was reported in black African population.^{4,5} An early and exact diagnosis of conjoined twins is a precondition for taking early and adequate action.² The first prenatal diagnosis of conjoined twins by two-dimensional ultrasound was published in 1976.⁶

Due to improvement in ultrasound imaging it is now possible to diagnose conjoined twins as early as the first trimester.⁷

CASE REPORT

A 20-year-old nulliparous woman was referred to our department at 13th weeks of gestation. The 2D transabdominal ultrasound examination was performed with a TOSBEE® SSA-240A® Toshiba® (abdominal convex probe, 7.5 MHz) and revealed a twin pregnancy. Detailed examination showed that these twins were conjoined with fusion of the head and chest (Figure 1).

After induction of labor with prostaglandin, a vaginal delivery of conjoined twins was achieved. Placental examination confirmed mono-chorionicity, with single insertion of umbilical cord which contained two arteries and one vein. The fetuses, 8.5 cm in length, had a large conjoined head with two faces, one located on each of the opposite sides of the head; fused from head dissymmetrically and also fused from chest. Four arms and four legs were present (Figure 2). One fetus had gastroschisis that was a herniation of abdominal contents through the body wall directly into the amniotic cavity (Figure 3). Autopsy revealed one heart with hypoplastic ventricles and one circulatory system, two immature respiratory organs. The twins had separate livers, spleens, esophagus, stomach, duodenum, terminal ileums and colons. The separate genitourinary tracts appeared male.



FIGURE 1: Two-dimensional ultrasound images of the cephalopagus twins at 13 weeks of gestation.

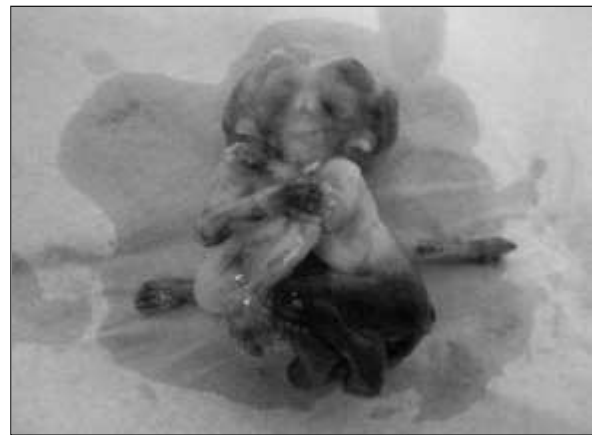


FIGURE 2: Macroscopic view of the cephalothoracopagus Union of the conjoined twins prior to autopsy.

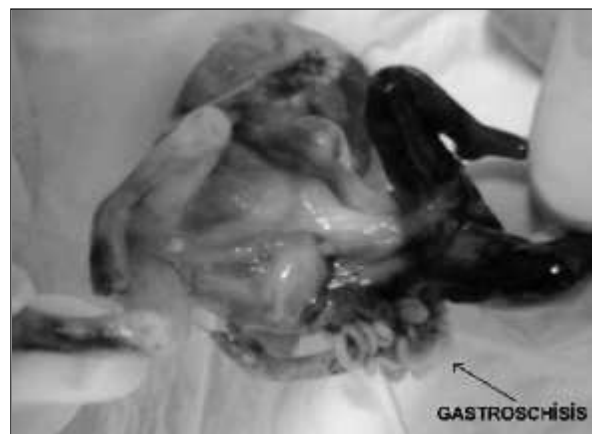


FIGURE 3: Macroscopic view of the gastroschisis of twins.

DISCUSSION

Conjoined twins form exactly like identical twins (because they are identical after all), but at some point during the stage where the single egg splits, the process stops, and the twins develop attached to one another. Identical twins will only be conjoined if they separate after the 12th day of conception. The twins will share a chorion, amnion and a placenta, making them monoamniotic/ mono-chorionic and subject to TTTS (Twin to Twin Transfusion Syndrome) and cord entanglement (two reasons why many are miscarried). No one knows why conjoined twins occur, although genetic and environmental factors have been explored. Conjoined twins are always of the same

sex. The most common types of conjunction, thoracopagus, omphalopagus, and thoraco-omphalopagus, account for 56% of conjoined twins.³ Cephalothoracopagus janiceps dissymmetricus with gastroschisis is the rarest type of conjoined twins. This type has two faces, on the opposite sides of the head, with one face usually being rudimentary. Other findings include separation of the lower abdomen and pelvis resulting in four arms and four legs. The conjoined twins presented here, fused from the head to upper abdomen, demonstrated four upper and four lower extremities, and two faces, each on opposite sides of the conjoined head. The prenatal diagnosis of conjoined twins is important for optimal obstetric management, including decisions regarding

mode of delivery to minimize maternal and fetal morbidity and mortality. The prognosis for conjoined twins is poor, thus when the diagnosis is made before viability, the option of terminating the pregnancy by vaginal delivery can be offered; 75% of such twins are stillborn or die within 24 h of delivery.² Later in pregnancy, the decision regarding vaginal delivery versus Cesarean section is based on the size of the fetuses and the likelihood of survival.⁸

In this case, 2D ultrasonography demonstrated that the facial, truncal and abdominal features are characteristic of cephalothoracopagus twins. The pregnancy was terminated because there was apparently no hope for surgical separation due to the extensive fusion of vital organs.

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