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

Gastroşisizli Sefalotorakopagus Janiceps
Disimetrikus, Çok Az Görulen
Bir Yapışık İkiz Formu

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 disymmetricus with gastrochisis, 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, gastrochisis


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

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Division of the embryonic disc more than 13 days after fertilisation is usually incomplete and results in conjoined twins. 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. An increased risk was reported in black African population. An early and exact diagnosis of conjoined twins is a precondition for taking early and adequate action. The first prenatal diagnosis of conjoined twins by two-dimensional ultrasound was published in 1976. Due to improvement in ultrasound imaging it is now possible to diagnose conjoined twins as early as the first trimester.
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 monochorionicity, 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 disymmetrically and also fused from chest. Four arms and four legs were present (Figure 2). One fetus had gastrochisis 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.

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/monochorionic 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-omphalo-
pagus, account for 56% of conjoined twins.\(^3\) Cephalothoracopagus janiceps disymmetricus with gastroschisis is the rarest type of conjoined twins. This type has two faces, on the opposite si-
des 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 con-
joined 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 sec-
tion is based on the size of the fetuses and the like-
lihood 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 ap-
parently no hope for surgical separation due to the extensive fusion of vital organs.

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