Prenatal Diagnosis of Cephalothoracopagus Janiceps

Sonographic-Pathologic Correlation

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Conjoined twins are identical twins whose bodies are joined in utero and are extremely rare complications of monochorionic twinning. They occur with incidence that rates that range from 1 per 50,000 to 1 per 100,000 births; however, the incidence of the cephalothoracopagus variety is 1 per 58 conjoined twins.[1] The precise etiology of conjoined twinning is unknown, but the most widely accepted theory is that incomplete division of a monozygotic embryo occurs approximately 13 to 15 days after ovulation. Conjoined twins are classified according to site of union. The most common location is the chest (thoracopagus).

In this report, we describe a case of cephalothoracopagus janiceps with a fused head and thorax. This is the rarest form of conjoined twins with an incidence of 1 per every 3 million deliveries.[2] The cephalopagus janiceps type of twins is characterized by 2 faces each looking in opposite directions on a single fused head. The face of each fetus is split in the midline with half turned outward, so that each observed face is made up of the right face of one fetus and the left face of the other. The term *janiceps* is derived from Janus, the 2-faced Roman god of gates and doorways.[3] In the case of identical and symmetric faces caused by the orientations of the 2 notochordal axes that are perfectly ventroventral, they are called janiceps disymmetros.[4]

To our best knowledge, only 13 cases of janiceps twinning with prenatal diagnosis have been reported. A search of the PubMed for "janiceps" without any limitation revealed only 31 articles. Moreover, only 5 cases were prenatally described with 3-dimensional (3D) sonography.[2,5–7] The objective of this report was to describe prenatal diagnosis and delineate the fused structures using both 2-dimensional (2D) and 3D sonography.

Case Report

A 21-year-old pregnant woman, para 2001, attended the first antenatal care clinic of a primary hospital at 26 weeks' gestation. Initial sonography at the first visit revealed fetal abnormalities suggestive of conjoined twins with a single placenta and polyhydramnios. The patient was referred to our hospital, a tertiary center, for detailed sonography. The parents had no history of major diseases (eg, diabetes mellitus), twins, or congenital malformations. There was no history of induced ovulation, exposure to teratogenic medications, irradiation, or infectious diseases during pregnancy.

Abbreviations
3D, 3-dimensional; 2D, 2-dimensional

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Sonography showed 2 female fetuses with a fused head, thorax, and upper abdomen and polyhydramnios. The fusion involved the top of the head down to the umbilicus. The conjoined twins had a single fused head consisting of 2 faces, which were oriented away from each other at 180° to each other. Each face turned to the direction perpendicular to its body axis. The 2 faces were relatively symmetric. However, 1 face was normal, and the other was associated with cyclopia, a proboscis, and an abnormal nose as well as a poorly formed mouth (Figures 1 and 2). In the single cranial vault, duplicated cerebra, thalami, and brain stems and 4 lateral ventricles were shown. The twins had a fused thorax containing 2 separate hearts and vascular systems and a fused abdomen at the upper part. Each fetus had its own normal set of extremities, pelvic bones, and female genital structures. The single umbilical cord containing 4 vessels was inserted at a single fused umbilicus. Three-dimensional sonography showed surface-rendered images of the conjoined region, 2 pairs of limbs, and a unique craniofacial appearance of 1 normal face and 1 abnormal face as described above on 2D sonography but much more apparent and informative (Figures 3 and 4). Fetal echocardiography displayed 2 separate hearts with a relatively small left ventricle in 1 heart as well as a small aorta. The 2 aortic arches were joined together with an abnormal vessel. The volume data sets of 4-dimensional cardio–spatiotemporal image correlation could not well interpreted because of poor fetal posture and acoustic shadowing.

Induction of labor and vaginal delivery were successfully performed with intravaginal misoprostol. The conjoined twins had spontaneous delivery with a body weight of 900 g. Postmortem, the fetuses had gross anomalies compatible with prenatal the sonographic findings (Figures 2, 5, and 6). The autopsy revealed anthropometric measurements consistent with 26 weeks' gestation. The twins had ventroventrally fused trunks above the umbilicus with 4 upper and 4 lower limbs. The backs and extremities were externally unremarkable. Only 1 umbilical cord containing 4 vessels was inserted at a single fused umbilicus. The twins shared vessels from each aortic arch; 1 fetus had cardiac disproportion, a small left ventricle, a small ascending aorta, and a prenatally undetected ventricular septal defect. They had 2 tracheas with separated sets of lungs but only 1 esophagus, 1 stomach, and 1 duodenum, which was separated to each fetus at the jejunum. The jejunum was atretic in 1 fetus. Each had its own genitourinary tract. They had their own kidneys and uterus with

Figure 1. Two-dimensional sonograms of the janiceps fused head. Left, Cross-sectional scan at the orbital level. Top right, Oblique coronal view of the normal face. Bottom right, Coronal view of the abnormal face. C indicates cyclopia; and P, proboscis.

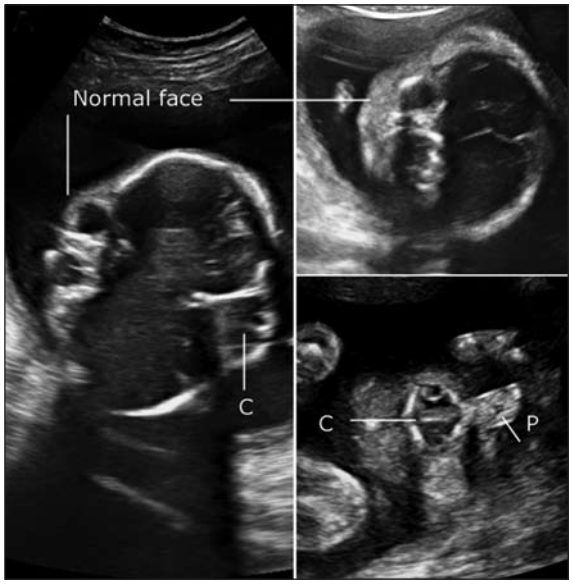
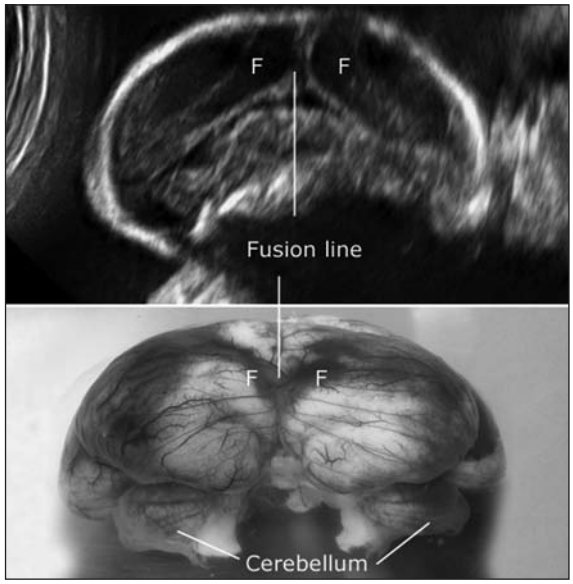


Figure 2. Prenatal 2D sonogram of the brain (top) and postnatal brain (bottom) showing ventral fusion of the forebrain (F)



normal female genitalia. Postmortem radiography showed 1 fused skull and 2 complete vertebral columns.

Discussion

Janiceps twins may be categorized into 2 subgroups: disymmetros and monosymmetros. If the 2 faces are symmetric, they are called janiceps disymmetros, in which the orientations of the 2 notochordal axes are perfectly ventroventral and the 2 faces on opposite sides of the head are identical and apparently normal. Monosymmetros twins, asymmetric unions, are associated with 1 rather "normal" anterior face and a rudimentary posterior face.[3,8] In janiceps disymmetros, the 2 faces are oriented away from each other, and each is at right angle to its axis of the notochord, whereas janiceps monosymmetros twins have 1 poorly developed notochord, resulting in 1 normal face and 1 poorly developed face, usually associated with varying degrees of differences and abnormalities.[9,10] The case presented here should be classified as disymmetros because the 2 faces were symmetric in size and location, although they were not identical because of abnormalities in 1 twin. In our case, the heads were joined at the frontal area, as they rotated 90° in opposite directions on their own body axis, resulting in 2 relatively complete faces on the 2 opposite sides of a single fused head. A half of the fused face belongs to 1 twin and the second half to the other twin. In our case, 1 twin was associated with a markedly abnormal face including a proboscis, absence of a nose, cyclopia or microphthalmia with a single palpebral fissure, and a malformed ear, but both had symmetrically duplicated cerebra, brain stems, cerebella, and spinal cords, as in previous reports.[9–11] Vascular abnormalities may play an important role in cerebral maldevelopment of cephalothoracopagus janiceps, primarily frontal abnormalities, cyclopia, severe gyral abnormalities, and focal areas of necrosis.[10] Fused forebrains are usually associated with varying degrees of holoprosencephaly.

The 2 faces of a single fused head must be differentiated from laterally joined diprosopus, or 2 faces, twins, in which both faces look forward, and each face is associated with just 1 twin. A distinguishing characteristic of parapagus twinning is that the duplicated structures retain their normal anterior/posterior orientation. In diprosopus twins, the duplicated structures are side by side, whereas janiceps twins are developmentally different in that both notochords face each other and are joined anteriorly or ventrally, not laterally. Each notochord contributes to the development of each face.

Figure 3. Coronal 3D sonogram of the relatively normal face.

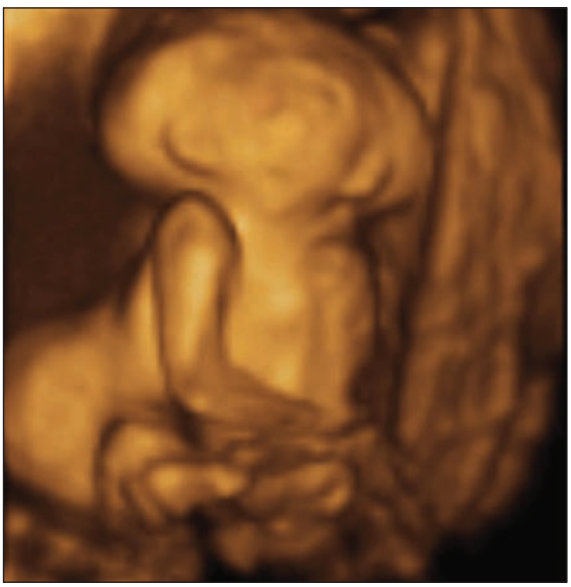
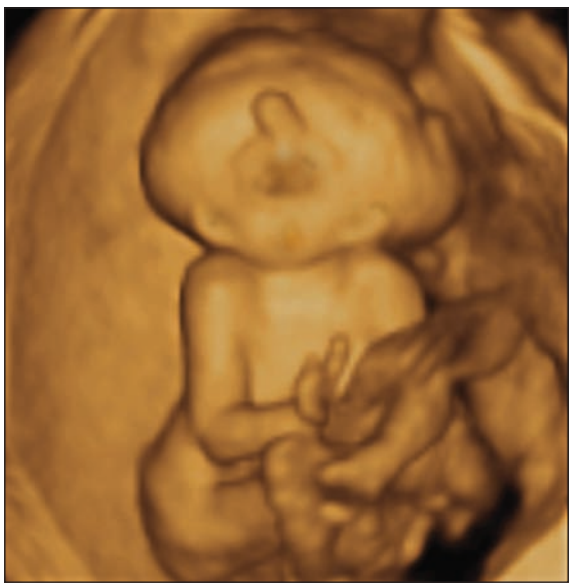


Figure 4. Coronal 3D sonogram of the abnormal face with cyclopia and a proboscis.



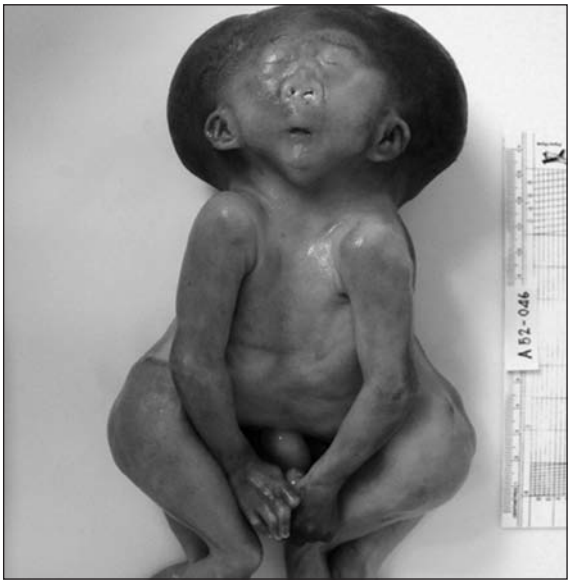


Figure 5. Postnatal coronal view of the relatively normal face.

Janiceps twins typically are joined in the sternal area and have relatively complete duplication of the lower body, as seen in the case presented here. Duplication of visceral structures is frequently seen, and the orientation is rather unpredictable because of the competing vertebral and facial axes.

Figure 6. Postnatal coronal view of the abnormal face with cyclopia and a proboscis.

Notably, 3D sonography has diagnostic advantages for evaluation of fetal structural abnormalities in its ability to store and retrieve a complete volume and to simultaneously display simultaneously surface image and the fetal skeleton, spine, and thorax. The combination of the 3D surface mode and 3D power Doppler imaging further gives a 3D impression of the fetal blood flow. The reliable diagnosis of janiceps twinning in the case presented here was based on the depiction of the 2 wide faces that were oriented 90° away from the body axis of the 2 vertebral columns of the single fused head on both 2D and 3D sonography. We believe that 2D sonography is the primary tool for diagnosis, but 3D sonography is very helpful as an adjunct for a detailed analysis of the degree of fusion and the extent of the anomalies. Three-dimensional sonography may clearly show janiceps twinning as early as 13 weeks' gestation.[5,12] Additionally, magnetic resonance imaging can be helpful for assessing the shared anatomy in some cases that are less deformed.[13] Although the prognosis is extremely poor, and surgical separation of the complex craniofacial defects is usually not offered, prenatal 3D sonography may depict the abnormalities for effective counseling of the parents. In our case, 3D sonography was very helpful for confirming the presence of the facial abnormalities and the degree of fusion. Three-dimensional sonography helped the parents observe 2 faces in an extremely wide frontal view on the opposite sides of the fused cranium.

In conclusion, we report a case of female conjoined cephalothoracopagus twins, janiceps disymmetros. The relatively symmetric fused head consisted of 2 faces looking in opposite directions. One face was relatively normal, whereas the other showed abnormalities. The prenatal diagnosis was made by 2- and 3-/4-dimensional sonography.

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