Symmetric Cephalothoracopagus “Janiceps” Twins: Sonographic Features

To the Editor: Conjoined twins can be classified on the basis of the site of the union; thus, 3 main types can be described: ventral union, dorsal union, and rarer forms of union. A ventral union is characterized by the fusion of the 2 embryos on the ventral side (eg, the abdomen). Dorsal union twins are joined on the dorsal aspect (eg, the vertebral column or occipital bone). Twins conjoined at the head and chest are called cephalothoracopagus twins. The cephalothoracopagus variety called “Janus” is characterized by the presence of 2 opposite faces, which are composite structures, half of which belonging to 1 twin and half to the other. We present the sonographic and anatomic features in a case of Janiceps twins.

A 33-year-old pregnant woman underwent her first ultrasound scan at 22 weeks’ gestation, with the diagnosis of joined twins. For ethical and religious reasons, she decided to avoid first-trimester screening procedures for fetal abnormalities (both structural and chromosomal). Sonographic features are described, with the help of the corresponding necroscopic characteristics (Figures 1 and 2). The twins were joined at the head and chest. The fusion of the cranial parts was symmetric. The single head showed 2 composite faces, 1 on each side (Figures 1, A and B, and 2, B and C). An enlargement of the cranial vault was noted. It is important to note that each face was well shaped, and the head resulted in a composite structure belonging half to 1 twin and half to the other. The fusion of the 2 heads involved the occipital regions, which were undetectable. The sphenoidal bone represented the center of the mass of the single head and was a deformed structure shared by the twins. One face had a common structure including a nose and mouth (proboscis), and the oral cavities were also complex and deformed because of the presence of a septum (Figures 1D and 2C). Two completely independent vertebral columns diverged inferiorly. A single common trachea was observed (Figure 1D). A fused thorax with 2 distinct hearts was seen. Although the heads were joined occipitally, the bodies were linked frontally, from the neck to the chest and the superior abdominal wall (Figure 2A). The arms and legs of the twins were well formed and independent (Figure 2A). The twins were female and had a common umbilical cord, rising from the abdominal wall at the midline, just before the separation of the 2 bodies (Figure 1C).

At 33 weeks’ gestation, because of preterm labor and polyhydramnios, the patient underwent a cesarean delivery. The twins died of acute respiratory failure soon afterward. Their weight was 4790 g, and a common umbilical cord rose from the lower abdominal wall (Figure 2A). No magnetic resonance imaging was performed during pregnancy or postmortem. An autopsy was avoided, following the last wish of the parents.

According to the “fission” hypothesis, this phenomenon occurs at the blastula stage, between the 13th and 15th days after fertilization. It has recently been proposed that conjoined twins result from secondary fusion of 2 separate embryos.1

Like all monozygotic twins, conjoined twins are always the same sex, both male or both female. It is estimated that 70% of conjoined twins are female.2

The subset of cephalothoracopagus twins is particularly interesting given its features at the midline.3 Chen et al4 reported a similar case but moreover displayed the shared circulation between the 2 separate hearts. In a follow-up study, 5 conducted a Doppler velocimetric study of cephalothoracopagus twins during the course of pregnancy. Even in that case, 2 hearts were seen. In both of these cases, polyhydramnios was present. Unfortunately, our case lacked first-trimester imaging.

It is important to note that in symmetric cephalothoracopagus janiceps twins, the face is well shaped but is shared by the twins and is therefore a composite structure belonging half to 1 twin and half to the other. In our case, the heads were joined at the occipital region, as they rotated 90° in opposite directions on their own thorax, thus resulting in 2 complete faces, located on the 2 opposite sites of a common head.
From a taxonomic viewpoint, the anatomic variety we found should be considered symmetric cephalothoracopagus twins, Janus type, orthogonal variety.

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References


Figure 2. Necroscopic characteristics of the joined twins. A, Janiceps twins observed on 1 side. B and C, Each face resulted in a composite structure belonging half to 1 twin and half to the other. The fusion of the 2 heads involved the occipital regions. The presence of a proboscis and the absence of a mouth and lips are shown in C.