Prenatal Diagnosis of Total Type 1 Vertical Craniopagus With 3-Dimensional Sonography

Conjoined twins, the most striking anomaly of monozygosity, have always aroused extensive interest in both the medical community and the general population, owing to their rarity and unusual presentations. In 1977, Fagan first reported the prenatal diagnosis of conjoined twins by sonography at 32 weeks’ gestation. Today, with the developments in technology, the prenatal diagnosis of conjoined twins is possible even in the first trimester. Here we present a case of craniopagus and the certain subtype with its 3-dimensional sonographic view, which is an extremely rare entity.

A 31-year-old woman, gravida 2, para 1, was referred to our perinatology unit at 18 weeks’ gestation for second-opinion sonography, with the suspicion of “pregnancy of Siamese twins.” Her personal and family histories were unremarkable, and she had a nonconsanguineous marriage. Also, none of her relatives had twin or triplet pregnancies. Her first pregnancy follow-up was uneventful, and she had a healthy boy.

Grayscale sonography revealed one common continuous cranium, absence of the falk cerebri in both twins, flattened cerebral hemispheres, and a transverse dural septum (Figure 1A). Three-dimensional sonography showed that the shape of the common cranium was like a stovepipe, and the faces of both fetuses were in the same directional axis, with an intertwin axial rotation less than 40° (Figure 1B). According to Stone et al, the final diagnosis of this condition was total type 1 vertical craniopagus. The parents chose pregnancy termination but denied fetal pathologic evaluation, after our counseling about the postnatal prognosis of craniopagus twins. Postabortal examination confirmed that the heads shared an extensive surface area on both parietal parts of the calvarium, with the faces looking at the same plane with a slight rotation (Figure 1C).

Despite the many hypotheses postulated during the past centuries, both the pathogenetic mechanism of conjoined twinning and the cause of monozygotic twinning itself are as yet unexplained. Conjoined twins are generally defined by the site of their most prominent union, which is ventral and dorsal in 87% and 13%, respectively. The abnormality is named with the suffix pagus, which means fixed. Types and distributions of ventral unions are cephalopagus (11%), thoracopagus (19%), omphalopagus (18%), ischiopagus (11%), and parapagus (pelvis and variable trunk; 28%), and types and distributions of dorsal unions are craniopagus (5%), as in our case, rachiopagus (vertebral column; 2%), and pygopagus (sacrum; 6%).

In craniopagus twins, the face, foramen magnum, and vertebrae are not primarily involved; the skulls are usually joined in roughly homologous regions, but asymmetries are common; and both vertical and nonvertical or angular forms are found. The thorax and abdomen are separate, and each twin has its own umbilicus and umbilical cord. It was also noted that the unions may involve any portion of the head and may include underlying structures such as meninges, venous sinuses, and the cortex. An infinite variation of configurations can occur according to the attachment location and degree of individual rotation of one head in relation to the other. Although some other twinning variations, such as cephalopagus, parapagus diprosopus (joined laterally, two faces on one head with one body), and rachiopagus, include fusion of the head, only craniopagus twins are attached solely at the calvaria.

The numerous phenotypic variations of craniopagus twinning and their postnatal outcomes necessitated the development of a classification system. To us, the most useful one is that in which craniopagus twins are initially classified as partial or total. Craniopagus twins are named partial when a limited surface area is affected and total when an extensive surface area is shared, with widely connected cranial cavities. In this classification system, total
cranioptagus types are also subclassified as frontal, parietal (vertical), temporalparietal, and occipital. The further subclassification of parietal (vertical) cranioptagus is defined as well, depending on the basis of the degree of rotation of one head in relation to the other. Type 1 vertical cranioptagus twins face the same direction; type 2 twins face opposite sides of the conjoined skull, the long axis of one head being rotated relative to that of the other through an angle of 140° or greater; and type 3 twins have an intermediate angle of rotation of the long axis of one head on that of the other.

According to the definitions above, our case’s final diagnosis was total type 1 vertical (parietal) cranioptagus. The couple chose pregnancy termination after our disclosure about the postnatal and post-multisurgical progress, pointing out 33% as the mortality rate, 17% as the severe disability rate, and 42% as the rate of mild-to-moderate or normal outcomes in twins with this subtype. Unfortunately, the parents’ refusal of postabortal pathologic, radiologic, and genetic examinations restricted us from reporting more detailed data about our case.

Today, in general practice, every effort is made to have earlier diagnoses of fetal abnormalities. Our case, an example of second-trimester cranioptagus, can be classified as one with a late diagnosis, after comparison to a case with cranioptagus diagnosed at 10 weeks and another case with cephalopagus diagnosed at 13 weeks.

We conclude that every effort should be made to determine the certain types and subtypes of cranioptagus twins for proper prenatal counseling. Additionally, 3-
dimensional sonography may be more accurate than 2-dimensional sonography alone for defining an earlier diagnosis and the definite type of craniopagus, in which the extent of the shared calvarial area and direction of the faces are the major determiners.

Cem Yasar Sanhal, MD, Murat Ozekinci, MD, Inanc Mendilcioglu, MD, Mehmet Sakinci, MD, Mehmet Simsek, MD

Department of Gynecology and Obstetrics (C.Y.S., M.O., I.M., M.Sa., M.Si.)
Department of Perinatology (C.Y.S., I.M., M.Si.)
Akdeniz University
Antalya, Turkey
doi:10.7863/ultra.33.1.179

References