

# Diagnosis of male dicephalus parapagus dibrachius conjoined twins discordant for anencephaly at early weeks of pregnancy. Case report

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## Abstract

We report a rare case of male dicephalus conjoined twins discordant for anencephaly diagnosed by ultrasound at 13 weeks. The prognosis of conjoined twins depends on type of fusion and presence of associated structural defects. High incidence of poor prognosis in postnatal, termination of pregnancy recommended in cases where dicephalic twins are detected early in utero, especially if one of them has discordance for anencephaly.

**Keywords:** dicephalus, anencephaly, ultrasound, pregnancy, prognosis

## Introduction

Conjoined twins are rare and complex complication of monozygotic twinning (MC). The incidence of conjoined twins is reported as approximately 1/30.000-100.000 and 1/600 of twin births<sup>(1)</sup>. In particular MC twins, have an increased risk of congenital anomalies than singleton pregnancies. More common congenital anomalies are observed up to now; neural tube defects (particularly anencephaly), holoprosencephaly, cloacal dystrophy and sirenómelia. As interesting, chromosomal abnormalities are virtually absent in conjoined twins<sup>(2)</sup>.

Two-dimensional ultrasound (2D) and three-dimensional ultrasound (3D) are reliable methods diagnosis of conjoined twins in early prenatal period<sup>(3)</sup>. Conjoined twins with the classification of 2D ultrasound are too difficult because of their complex three dimensional structures. 3D ultrasound, due to reveal its complex anatomical spatial relationships, may be useful in determining the complex of fetal anatomy of conjoined twins<sup>(4)</sup>.

Early prenatal diagnosis of conjoined twins allows better counseling of the parents regarding the management options, including continuation of pregnancy with post-natal surgery, termination of pregnancy or selective fetocide in cases pregnancy<sup>(5)</sup>.

We reported the prenatal diagnosis of a case of male dicephalus parapagus conjoined twins discordant for anencephaly via ultrasound and autopsy findings at the 13th week of gestation.

## Case Report

A 27-year-old, multigravid healthy woman was referred for the first scan at 13 week of gestation. Her previous pregnancy

was selectively terminated because of anencephalic fetus. Conjoined twins were diagnosed via 2D abdominal sonography at 13 week of gestation. Sonographic examination revealed single fetus with two heads facing each other and one of them had anencephaly, two necks, a single thorax, two upper and lower limbs were visualized. There was single umbilical cord and no amniotic membrane. Two vertebral column, single stomach, single liver, single fetal heart, single urinary bladder were also observed (Figure 1). 2D ultrasound findings were confirmed with transvaginal ultrasound, 3D, and color doppler. Only one heart and liver was examined by the color



Figure 1. Transvaginal ultrasonography of dicephalus conjoined twins discordant for anencephaly. Single fetus with two heads and one of them had not calvarium, two necks, a single thorax, separate two vertebral column, single stomach and single fetal heart are visualized

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**Figure 2.** Clinical images of the external examination of the dicephalus conjoined twins discordant for anencephaly. Two discordant head with one normal and the other anencephalic and also two necks, a single thorax, two upper and lower limbs with no anomaly and male external genitalia were observed

doppler ultrasound. The parents decided to terminate the pregnancy. After delivery, clinical external examination and postmortem autopsy examination of the conjoined twins confirmed ultrasonographic diagnosis (Figure 2). According to these findings, dicephalus parapagus dibrachius conjoined twins discordant for anencephaly was diagnosed.

## Discussions

The etiology of discordant and concordant anomalies of monozygotic and conjoined twins is unknown and still discussed. Congenital malformations usually occur in almost all sets of conjoined twins and 60-70% of them involve structural abnormalities not associated with the area fusion. Despite genetic identity, conjoined twins often have discordant anomalies<sup>(6)</sup> and these often occur in the twin on the right<sup>(7)</sup>. Most commonly, these anomalies include neural tube defects, cardiovascular system anomalies and orofacial clefts<sup>(8,9)</sup>. Schinzell et al. reported that conjoined twinning and anencephaly develop on the same etiological factors; this relationship may be resulted lack of embryonal migration or fusion<sup>(2)</sup>.

Dicephalus conjoined twins discordant for anencephaly is rare in literature. Our case is parapagus dicephalus di-

brachius; and as it is supported in literature, right fetus has anencephaly and that is not associated with the area fusion. Additional anomaly wasn't observed. Diagnosed at early weeks of pregnancy dicephalus parapagus conjoined twin discordant for anencephaly has not been reported in literature<sup>(5)</sup>. Although we found three dicephalus parapagus conjoined twins discordant for anencephaly in literature, none of these cases was not diagnosed at early pregnancy period<sup>(10,11,12,13)</sup>. Also, our case was male fetus despite predominance female fetus in literature<sup>(14)</sup>.

## Conclusions

As far as we know this case is the first of dicephalus conjoined twins discordant for anencephaly twins of male sex diagnosed at first trimester. The point we want to emphasize is that in conjoined twin cases, the presence of concordant/discordant congenital anomaly, and determination of organ sharing level should be evaluated by ultrasonography in early prenatal period and accurate diagnosis should be maintained, which is critical for pregnancy management in the later period. This offers an opportunity for adequate counseling and, if desired, earlier termination of pregnancy associated with minimal maternal risks. ■

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