

# Dichorionic twin pregnancy discordant for fetal anencephaly: a case report

## *Fetal anensefali ile komplike dikoryonik ikiz gebelik: Olgu sunumu*

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### Abstract

Dichorionic twin pregnancy discordant for fetal anencephaly is a serious condition that threatens the normal co-twin's life by causing polyhydramnios, preterm labor and sudden death of one or both of the fetuses. We report a case of dichorionic twin pregnancy discordant for fetal anencephaly delivered at the 32<sup>nd</sup> week of gestation because of preterm labor and nonreassuring fetal monitoring. The aim of this case report is to summarize management options in this situation.

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**Key words:** Anencephaly, twin pregnancy, discordant fetal growth

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### Özet

Fetal anensefali ile komplike dikoryonik ikiz gebelik polihidramnioz, preterm eylem ve bir ya da iki fetusun birden ani ölümü şeklinde sonuçlara yol açabilen potansiyel tehlikeli bir durumdur. Bu çalışmada 32. gebelik haftasında preterm eylem ve güven vermeyen fetal kalp hızı patterni nedeni ile doğum yapan fetuslardan birinin anensefal olduğu dikoryonik ikiz gebelik olgusu sunulmuş ve tedavi seçenekleri tartışılmıştır. (J Turkish-German Gynecol Assoc 2012; 13: 64-6)

**Anahtar kelimeler:** Anensefali, ikiz gebelik, uyumsuz fetal büyüme

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### Introduction

In twin pregnancies discordant for anencephaly, there are risks of development of polyhydramnios, severe preterm delivery and death of the anencephalic fetus. In dichorionic twins discordant for anencephaly, there are three management options: selective fetocide, serial ultrasound examination for polyhydramnios or expectant management (1). In this paper, we present our experience with a case of twin pregnancy discordant for anencephaly which was managed conservatively.

### Case Report

A-27-year old woman was admitted to our emergency unit with preterm contractions at 32<sup>nd</sup> week of gestation. She was hospitalized with the diagnosis of twin pregnancy and preterm labor. She was primary infertile and conceived with gonadotropine treatment and intrauterine insemination after 7 years of infertility. She was observed in another center with the diagnosis of preterm labor and was treated with tocolytic drugs and corticosteroids for fetal lung development. An ultrasound scan showed a dichorionic diamniotic twin pregnancy. Twin A's development was concordant to 30 weeks and it was structurally normal but twin B was anencephalic and concordant to 29 weeks. This situation was noticed at the 14<sup>th</sup> weeks of gestation, the couple was informed and the expectant management option was chosen. The amniotic

fluid index of the second fetus was increased (300 mm). Fetal heart rates of both babies were bradycardic (60-80/min), thus the patient was delivered by emergency cesarean section. The anencephalic female infant died soon after birth (birth weight 920 gr). The surviving male infant weighed 1520 gr and had an Apgar scores of 6 and 7 at 1 and 5 mins, respectively (Figure 1). The surviving infant was admitted to the neonatal intensive care unit (NICU) and given nasal continuous positive airway pressure for two days and supplemental oxygen for further day. During the NICU stay, the developing bronchopulmonary dysplasia was treated with synthetic surfactant therapy. At the 2<sup>nd</sup> day in NICU, icterus appeared and was treated with phototherapy. The infant was discharged after 26 days weighing 2010 gr.

### Discussion

Anencephaly, together with spina bifida, is the most common and multifactorial neural tube defect, occurring in about 1 in 1000 births (2). In a twin pregnancy when one of the fetuses is anencephalic, this situation multiplies the risks and complications twin pregnancies already have. Prenatal detection of anencephaly by ultrasound is possible in almost 100% of cases (3, 4). In singleton pregnancies, almost all of these conceptions are terminated since anencephalic infants have no chance to survive. However, in the case of multiple gestations the management is not that clear. In twin pregnancies complicated with one anencephalic fetus, there is



**Figure 1. Postnatal image of discordant anencephaly in dichorionic twins: Anencephalic and normal twins**

an increased risk of either neonatal death due to severe preterm delivery secondary to development of polyhydramnios or intrauterine death. Another major risk is development of discordant fetal growth. The prevalence of discordance for anencephaly is higher in monochorionic than in dichorionic twins (5, 6).

In their retrospective study, Ben Ami et al. asked the question "Is there an increased rate of anencephaly in twins?". In this study a higher rate of anencephaly was currently found among IVF-ICSI pregnancies compared to spontaneous pregnancies. This is attributed neither to assisted conception technique nor to recent folic acid supplementation, but rather to the twinning itself (7). In twin pregnancies the prevalence of anencephaly is higher than in singletons (5, 6).

After diagnosis of an anomaly in one fetus with a normal co-twin, the general management options are the following: abortion of both fetuses, continuation of pregnancy without intervention or selective fetocide of the abnormal twin. In the presented case, the family did not approve of the selective fetocide treatment. A multicenter study revealed that selective fetocide after 16 weeks' gestation is related with a higher risk of miscarriage (5%-14%), but it reduces the risk of severe preterm delivery in the remaining fetus before 16 weeks (8). In dichorionic twins, Vandecruys et al. recommend serial ultrasound examinations for early diagnosis of polyhydramnios, which can then be treated either by amniodrainage or selective fetocide (9). In the presented case, polyhydramnios was detected on admission and an emergency cesarean section due to the fetal bradycardia was performed at 32 weeks of gestation; and therefore amniodrainage was not performed. Leeker et al. emphasised that selective fetocide of the anencephalic fetus before 15 weeks of gestation prevents the development of polyhydramnios and might reduce the risk of prematurity and increase the chance for survival for the healthy fetus (1). In addition, there are a few studies carried out on expectantly managed dichorionic twins with one anencephalic fetus where the rates of pregnancy complications are similar to those reported in normal twin gestations (10, 11).

In monochorionic twin pregnancies, conventional selective fetocide is not possible because of the high risk of subsequent death of the normal co-twin, and treatment options are expectant management and selective fetocide by cord occlusion (1).

There are a few studies describing outcomes in twin pregnancies discordant for anencephaly. The largest ones are reviewed by Vandecruys et al. (63 cases) and Leeker et al. (43 cases) (1, 9). Neither group used selective fetocide for monochorionic twins and expectant management was the common choice. Survival rates of the normal co-twin in these studies were 90% and 88%, respectively. Lust et al. (11) introduced treatment options in 86 cases of monochorionic and dichorionic twin pregnancies complicated with anencephaly. The treatment options reported were selective fetocide or expectant management in all cases. In dichorionic twins, selective fetocide was performed by intracardiac instillation of potassium chloride, whereas in monochorionic twins, it was carried out by bipolar coagulation of the umbilical cord. No significant differences were found in survival of nonaffected twin between the management options in monochorionic and dichorionic twins although a statistically significant difference was found between the two groups in mean gestational age and birth weight at delivery supporting the selective fetocide. Survival rates in the dichorionic group were found as 94.1% and 95.1% for the fetocide and expectant management groups, respectively. In this study, the authors concluded that "While selective fetocide does not reduce perinatal mortality, it does result in significantly longer gestations and higher birth weight, and appears to be a choice in dichorionic twins discordant for anencephaly. In monochorionic twins, selective fetocide also increases birth weight, but in view of the complexity of this group, no clear recommendations can be made" (11).

As a result, dichorionic twin pregnancy discordant for fetal anencephaly is a rare case but may have serious consequences. Although both expectant management and selective fetocide have good outcomes, close follow-up of these patients is very important in management.

#### Conflict of interest

No conflict of interest was declared by the authors.

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