

Dichorionic twin pregnancy discordant for anencephaly: two cases with different management

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Summary

Background: Prevalence of anencephaly in dichorionic twins is higher than in singleton pregnancies. The authors report two cases with two different management strategies. **Case 1:** Spontaneous dichorionic diamniotic twin pregnancy with the second twin diagnosed with anencephaly at 12 weeks gestation. Selective feticide was performed at the age of 13.2 weeks. Vaginal delivery occurred at 39 weeks, and birth weight was 2,850 g. **Case 2:** Dichorionic diamniotic twin pregnancy discordant for anencephaly in the second twin was diagnosed at 13 weeks gestation. An expectant management was decided. Preterm delivery occurred at 35 weeks due to hydramnios of the affected fetus, delivering a healthy newborn weighing 2,300 g and an anencephalic neonate who died immediately after delivery. **Conclusion:** Anencephaly should be diagnosed as soon as possible, idealistically at 11-13+6 weeks ultrasound (US) scan, in order to offer the most appropriate counselling to the parents, ranging from selective feticide or expectant management. This short series suggests that selective early feticide may increase gestational age and birth weight.

Key words: Anencephaly; Twin pregnancy; Prenatal diagnosis; Selective feticide; Expectant management.

Introduction

Anencephaly is a neural tube defect incompatible with life. Intrauterine death rate is about 25% and postnatal survival is usually less than 48 hours in these cases [1]. The incidence rates of anterior neural tube defects, anencephaly and encephalocele, are increased among twins compared with singletons [1-3]. Also, congenital anomalies that include neural tube defects are increased in monochorionic twins in comparison to dichorionic twins [1, 4].

During the first-trimester scan, many major fetal abnormalities, including anencephaly, can be reliably diagnosed [5]. In twin pregnancies discordant for anencephaly, the two main risks for the unaffected co-twin arise from either the spontaneous death of the anencephalic fetus or the development of polyhydramnios [1, 4, 6]. The diagnosis of such anomaly in the first trimester of gestation allows the health professional to establish the best strategy to minimize the risks, especially death or early premature delivery, of the normal twin. In dichorionic twin pregnancies discordant for anencephaly, the two management options are selective feticide (SF) by intracardiac injection of potassium chloride (KCl) or an expectant management with serial ultrasound (US) examinations [1, 6].

Counselling parents in this situation may be a difficult task. A first series suggested that the best management for these pregnancies was the expectant management with serial US in order to minimize the risks of miscarriage that may occur after SF. If polyhydramnios is detected amniocentesis or late selective feticide could be offered. However, this conclusion was based on the presence of

only one case of miscarriage in a series of nine cases managed with conservative management. In addition, in cases managed with conservative management, 57% developed polyhydramnios and, in total, 20% had late invasive procedures (amniocentesis or late SF) [6]. Risks of these procedures were not completely evaluated. On the contrary, in a further series and systematic review [7, 8], recommendation of SF was performed on the basis of a longer duration of gestation in this group without significant increased perinatal mortality.

Therefore counselling to these parents has not been completely resolved. This is an important issue, not only considering the health problem, but also the psychological impact of carrying an anencephalic fetus along gestation with the uncertain risk about how this condition may affect the healthy baby. On the other hand, moral and religious issues should be obviously respected.

The aim of the present study was to report a short series of two cases occurring in the same year, and therefore counselled in the same way but managed differently after knowing the parents' decision. A discussion about the risk and benefits of the two managements is presented. The authors describe the outcomes of both pregnancies and discuss the benefits and risks of each one of them. The authors aimed to know which one of these two managements could better improve the outcome of the normal twin.

Case Reports

Case 1

A 38-year-old primiparous woman with a spontaneous twin pregnancy attended this hospital for a 12 week scan. Abdominal two-dimensional (2D) US revealed a dichorionic diamni-

otic twin pregnancy with the second twin affected with anencephaly. The parents received counselling and decided to have a SF. The procedure was conducted with intracardiac injection of 1.5 cc of KCl at 13.2 weeks. Follow-up US were performed every two weeks, showing normal fetal growth, anatomy, and amniotic fluid volume. The maternal hematological and coagulation parameters remained normal. Vaginal delivery occurred spontaneously at 39 weeks. Birth weight was 2,850 g and Apgar scores at one and five minutes were 7 and 9, respectively, requiring resuscitation type I.

Case 2

A 39-year-old primiparous woman with a twin pregnancy conceived with in vitro fertilization (IVF) was controlled in this hospital. At 13 weeks gestation the abdominal 2D US revealed an anencephaly in the second twin. On this occasion, the parents decided not to perform SF, but an expectant management. US monitoring showed intrauterine growth restriction and moderate hydramnios in the affected twin. A magnetic resonance imaging (MRI) scan was performed at 28 weeks gestation in order to rule out central nervous system abnormalities in the healthy twin. Spontaneous preterm labour occurred at 34.4 weeks and the patient was hospitalized. A single course of steroids for lung maturation was given. Cesarean section was performed at 35 weeks gestational age due to breech presentation of the first twin. The first newborn weighed 2,300 grams, and Apgar scores at one and five minutes were 6 and 8, respectively. No congenital abnormalities were observed and he was admitted in the neonatal intensive care unit due to low weight. He was discharged without any complication at the third day after birth. The second twin weighed 1,640 grams and his Apgar scores at one and five minutes were 1 and 0, respectively. No anomalies other than anencephaly were observed.

Discussion

Neural tube defects, such as anencephaly, are increased in twin pregnancies compared with singletons [1-3]. It is not clear if the cause of this phenomenon could be attributed to the twinning or to the mode of conception. In a recent study of 43 pregnancies diagnosed with anencephaly, Ben-Ami *et al.* concluded that twin pregnancies conceived by assisted reproductive technology constituted a high-risk group for anencephaly OR = 24.6 (CI = 11.4-53.2). It is a possible synergistic effect of both twinning and assisted reproductive technology [2].

An early diagnosis of dichorionic twin pregnancies discordant for anencephaly between 11+0 to 13+6 weeks gestation should be performed to offer the parents management options of the pregnancy: expectant or SF [6].

There is controversy regarding which treatment is best, expectant management or SF. Data from dichorionic twin pregnancies discordant for anencephaly managed expectantly demonstrated that although the rate of either miscarriage or fetal death between 12 and 23 weeks is similar to that in dichorionic pregnancies with normal fetuses (about 1%) [6, 7], the rate of early preterm delivery is increased, probably associated with a high risk of polyhydramnios [1, 6, 8]. In these cases, polyhydramnios is caused by an impairment of the swallowing reflex of the anencephalic fetus

and the management in severe cases may include amniocentesis or late SF. Invasive management in late pregnancy has potential complications, such as rupture of membranes, chorioamnionitis or fetal death [6, 8, 9]. Vandercruys *et al.* observed, in a group of 35 dichorionic twin pregnancies discordant for anencephaly with expectant management, an incidence of 55% of polyhydramnios between 25 and 31 weeks gestation, requiring invasive procedures in 35% of them, which represents 20% of the total [6]. In the present case, the patient managed expectantly developed a moderate hydramnios and delivered at 35 weeks.

SF in dichorionic twins is performed by US-guided fetal intracardiac injection of KCl in the second trimester. Although there is no consensus regarding which is the best gestational age to perform the SF, most series recommend performing it as early as possible in order to reach the best perinatal outcomes with the lowest rate of fetal loss and extreme prematurity [10-12]. The overall pregnancy loss rate before 24 weeks of gestation after SF was 4% in a series of 200 patients [10] and 7.5% in the largest collaborative experience of 402 women [11]. Also, Evans *et al.* showed a trend towards increasing loss rates with advancing gestational age at the time of the SF with a risk of fetal loss rate from 5.4% between nine to 12 weeks, to 9.1% when it is performed later than 25 weeks [11]. In the present case, the SF was performed at 13 weeks without any complications. Therefore, the present authors recommend that SF be performed as early as possible to minimize the risks in pregnancy and the psychological impact on women.

Although a short series with SF did not find a reduction in the rate of early preterm delivery [6], a recent systematic review with 58 dichorionic twin pregnancies discordant for anencephaly concluded that while SF does not reduce perinatal mortality, it does result in significantly longer gestations and higher birth weight than expectant management, as observed when compared to the two present cases [9].

In conclusion, despite the fact that this series is short, it seems that there are more advantages with SF than with expectant management in dichorionic twin pregnancies discordant for anencephaly. Nonetheless, the mother has the ultimate decision to take risks and her personal decision should always be respected.

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