

Discordance for fetal anencephaly in a dichorionic twin pregnancy: A case report

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ABSTRACT

Anencephaly is a frequent and easily diagnosed condition affecting twin pregnancies with higher incidence than singletons. Discordance for anencephaly in twins raises management dilemmas about the best approach to the situation. The authors report a successful case of a 33 years old woman who had a dichorionic twin pregnancy resulting from ICSI, with discordance for anencephaly. Diagnosis was performed at 13 weeks during first trimester ultrasound screening. A selective feticide with intrathoracic injection of KCl at 13 + 2 weeks was performed with no complications. The normal fetus proceeded with an uneventful pregnancy with spontaneous vaginal delivery at 40 weeks, weighting 3110 g. Management of twin pregnancies discordant for fetal anencephaly is far from being consensual. Two options are considered: selective feticide or expectant management, with serial ultrasound surveillance. In the latter option, amniodrainage or selective feticide may become options if polyhydramnios develops later in pregnancy. Therapeutic strategy requires an individual approach considering chorionicity, gestational age and all the risks of miscarriage, preterm labor and fetal demise to which these pregnancies are particularly exposed.

Keywords: Anencephaly; Twin Pregnancy; Multifetal Pregnancy Reduction

1. INTRODUCTION

Anencephaly is one of the most common and multifactorial fetal anomalies [1,2]. The widespread introduction of ultrasound screening at 11 + 0 to 13 + 6 weeks of gestation allowed an earlier diagnosis of major fetal anomalies such as anencephaly [1,2]. Twin pregnancies present with a higher prevalence of anencephaly, with monochorionic twins having a higher incidence of discor-

dance than dichorionic [3]. Twin pregnancies resulting from assisted reproductive treatments (ART) are particularly affected by this condition [4].

Management of twin pregnancies discordant for anencephaly is controversial, with different options depending on chorionicity and gestational age [3,5]. Balancing the risk of selective feticide and miscarriage *versus* the risk of polyhydramnios and preterm labor becomes a challenge.

The authors report a case of dichorionic twins discordant for anencephaly successfully managed with selective feticide.

2. CASE REPORT

A thirty-three-year-old woman, with a history of primary infertility, presents with a dichorionic/diamniotic twin pregnancy resulting from ART ICSI (intracytoplasmic sperm injection). On first trimester ultrasound a fetal discordance for anencephaly was diagnosed (**Figure 1**). After adequate obstetric and genetic counseling and given all the information about the risks involving this pregnancy, the parents opted for selective feticide. The affected twin has been terminated by ultrasound guided intrathoracic injection of 2.5 mL potassium chloride (KCl) using a 22G needle, at 13 + 2 weeks. Cardiac assistolie was confirmed at the end of the procedure. At the same occasion, a chorionic villus sampling was performed for both fetuses, revealing a chromosomally normal fetus and a 45,X (3)/46,X, + mar(9) karyotype for the anencephalic fetus. The procedures had no complications and the normal twin proceeded with a normal and uneventful pregnancy. The onset of labor occurred spontaneously at 40 weeks, and an apparently healthy female baby was delivered with 3110 g and an Apgar score of 9 and 10 at 1st and 5th minute, respectively. The normal twin had a favorable and healthy neonatal evolution. Pathologic evaluation of the placentas and the unborn twin confirmed dichorionicity and revealed a 15



Figure 1. Fetal acrania on first trimester ultrasound.

weeks fetus with a major open neural tube defect (anencephaly) and syndactyly of 2nd to 3rd fingers and 4th to 5th fingers of both hands.

3. DISCUSSION

Anencephaly is a uniformly lethal and untreatable condition that occurs more frequently in twin pregnancies than in singletons, with monochorionic twins having a higher incidence of discordance than dichorionic [3]. Ultrasound diagnosis is desirable in 100% of cases and is possible as early as by the 11th week [1,2,6]. Anencephaly is a condition for which termination of pregnancy is a common option in singleton pregnancies. In twin pregnancies, discordance for anencephaly provides a management dilemma. The main problem of these pregnancies is that 50% will course with polyhydramnios, varying from mild to severe, eventually requiring repeated therapeutic amniodrainage [3,6,7], and an increased preterm labor risk with delivery of both normal and affected fetuses [3,8,9]. Although selective feticide seems to be the obvious option in case of discordant anomaly in twin pregnancies, it may in fact increase the risk of miscarriage and lead to demise of the co-twin, particularly in monochorionic twins, so that expectant management may be considered for lethal anomalies [6,10].

For dichorionic twins, as in the reported case, two options are taken: selective feticide or an expectant attitude with serial ultrasound evaluations. The miscarriage risk associated to fetal selective termination depends on the gestational age and ranges from 5% to 14%, depending on whether the procedure is carried out before or after 16 weeks of gestation [5]. In the reported case the option was for selective feticide which occurred at 13 + 2 weeks. The use of intrathoracic injection of KCl is a well-established procedure, with a success rate near 100% [10]. An advantage of early selective feticide is to reduce the risk of severe preterm delivery, achieving a higher average gestational age at birth [5,9].

There are few small published series on this issue.

Lipitz *et al.* reviewed a limited number of dichorionic twin pregnancies discordant for anencephaly conducted expectantly, with relatively good outcomes [7]. For 14 dichorionic pregnancies the average gestational age of birth was 35 weeks, with only 6 cases developing mild polyhydramnios [7]. Sebire *et al.*, in 1997, analyzing a total of 24 cases, did not find a conclusion on the best approach for discordance for anencephaly [3]. Both authors considered expectant management to be a reasonable option in dichorionic twin pregnancies.

Leeker and Beinder, reviewing 6 cases, reported one successful case with selective feticide and a poor outcome in two expectantly managed dichorionic pregnancies presenting with premature rupture of membranes (PROM) at 28 and 32 weeks respectively (severe polyhydramnios developed during second trimester on the latter case) [8].

More recently, Vandecruys *et al.*, reviewed the dilemmas in the management of early diagnosis of discordance for anencephaly and concluded that, for dichorionic twin pregnancies, expectant management is the best option, associated with the possibility of amniodrainage or selective feticide if polyhydramnios develop after 24 weeks [6], in order to minimize the risk of miscarriage and maximize the chance of a live birth after 33 weeks.

The largest systematic review performed on the subject was reported by Lust *et al.*, analyzing a total of 86 cases (58 dichorionic and 28 monochorionic). In dichorionic twins selective feticide was performed in 17 cases and expectant management in 41 and no statistic difference was found in perinatal survival. Nevertheless, all the other results clearly favor selective feticide, with significantly longer pregnancies (39 vs 34.9 weeks) and higher birth weight (2922 g vs 2474 g) comparing to expectant management. Besides the lower incidence of preterm delivery, a higher rate of vaginal delivery and a reduction in care costs were achieved in the selective feticide group. Therefore, the authors consider selective feticide as the best treatment strategy for dichorionic twins discordant for anencephaly [9].

For monochorionic twins expectant management remains the main option. The presence of placental anastomoses do not allow the injection of KCl or any other toxic agent, so that selective feticide in monochorionic pregnancies must be performed by umbilical cord laser or bipolar coagulation of the affected fetus, which is a technique more difficult to perform [10]. Intrauterine death of the anencephalic fetus increases the risk of death or severe brain demise of the co-twin [5,9-12] so it requires a treatment strategy different from dichorionic. Lust *et al.* had results favoring the selective feticide in monochorionic pregnancies with significantly higher birth weights and a tendency toward a longer gestation when compared to expectant management, but with

approximately the same survival rates for both groups. However, the complexity of monochorionicity and the small number of cases did not allow the authors to attest the best approach for monochorionic pregnancies [9].

Dichorionic, as well as monochorionic twin pregnancies discordant for anencephaly are still requiring an individual approach considering chorionicity/amnionicity, time of gestation and potential risk for pregnancy complications. Selective feticide as early as possible, ideally before 16 weeks, seems to be an acceptable and preferable option for dichorionic twin pregnancies affected by discordance for anencephaly [5,9].

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