

# Recurrent anencephalic stillbirths: A rare case presentation

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## ABSTRACT

Anencephaly is one of the three major lethal fetal anomalies. Termination of pregnancy is offered for all prenatally diagnosed cases, but some parents choose to continue with pregnancy with full obstetric intervention. I describe the case of a woman who had two pregnancies with anencephalic stillbirths in the third trimester. There is a paucity of well-defined standards of care and management for pregnancies that are carried to the third trimester with this malformation. This case raises important practical questions: Management strategies of recurrent pregnancies with anencephaly, best supportive care during, and after birth and counseling the expectant parents.

**Keywords:** Anencephalic birth management, care plan, recurrent anencephaly

## Introduction

Anencephaly is a severe congenital neural tube defect that is incompatible with life. The prevalence remains at 2.1/100,000 live births<sup>[1]</sup> with a 1<sup>st</sup>-year mortality rate of 100%.<sup>[2]</sup> Data suggest a predominance in females and Caucasians with a multifactorial polygenic etiology.<sup>[3,4]</sup> In utero diagnosis can be made reliably by testing maternal serum  $\alpha$ -fetoprotein levels and ultrasonographic imaging of fetal cranial vault.<sup>[5]</sup> While a large number of parents decide to terminate the pregnancy on diagnosis, a few families choose to continue even after counseling about poor prognosis and survival rates.<sup>[6]</sup>

Previous studies have highlighted the effects of pregnancy loss due to fetal anomalies on expectant mothers such as post-traumatic stress, depression, and grief.<sup>[7,8]</sup> However, the psychological impact of continuing with pregnancy in lethal fetal anomalies is understudied. Furthermore, there is a paucity of well-defined standard of care and management strategies for pregnancies with anencephaly, especially when parents decide to carry out the pregnancy. This poses significant practical challenges for health-care professionals who must be ready for all possible outcomes. This paper describes a rare case of recurrent anencephaly carried to the third trimester and discusses some of the issues relevant to clinical practice.

## Case Report

A 32-year-old, gravida 2, para 1 Caucasian Muslim woman presented to our unit at 10 weeks of pregnancy with a history of stillbirth of an anencephalic infant at the 26<sup>th</sup> week of gestation 3 years before. A first-trimester screen, including  $\alpha$ -fetoprotein test and chorionic villus sampling for chromosome analysis to determine the karyotype, was offered but this was declined by

the mother even with the understanding of the risks involved. She had taken folic acid as advised during the first trimester. Prenatal ultrasonographic scan performed at 18 weeks showed an anencephalic fetus with polyhydramnios. No other structural fetal defects were identified. The expectant mother was counseled by a multidisciplinary team (MDT) consisting of an obstetrician and a neonatologist. The mother decided to continue the pregnancy. A decision was reached with the mother not to perform cesarean section delivery. It was agreed with the parents that after birth inflation breaths could only be applied if signs of life were present and sustainable unsupported, feeding through orogastric tube could be started after birth if in infant's best interest initially with water or dextrose before using milk. Any resuscitation or invasive supportive attempts beyond this were deemed inappropriate. A written informed consent was obtained from the patient.

The mother gave birth to a male stillborn infant at 39 weeks 3 days gestation in a specialized obstetric unit. Detailed neonatal physical examination of the baby was undertaken by an experienced pediatrician. Physical examination showed a flattened posterior skull with a large occipital defect through which hemorrhagic red tissue was protruding. There were no other associated congenital malformations.

Counseling and investigations were offered both on first and second pregnancy to determine the cause of anencephaly; however, the mother declined postnatal cytogenetic evaluations of the placenta, umbilical cord blood, and fetal skin samples. Family history did not reveal parental consanguinity or anencephaly. The mother was discharged with full bereavement support, follow-up and advice about future pregnancy. The coroner's office was not informed as anencephaly was regarded as the natural cause of death.

## Discussion

This is a rare case of recurrent anencephaly leading to a stillbirth late in the third trimester. Physicians usually provide directive advice in favor of termination of anencephalic pregnancy as it is regarded incompatible with sustained life. Therefore, preparation for all possible outcomes including carrying out the pregnancy to the third trimester and survival of infant after birth is not considered. It is also assumed that expectant mothers would be followed up with necessary investigations and advice after the first pregnancy to prevent future instances. This case challenges both of these assumptions. Although the mother received. Through appropriate counseling at first presentation and termination support at diagnosis, she decided to carry out the pregnancy. The MDT team prepared for all possible outcomes, including the unassisted survival of the infant, and had an agreed care plan, which was distributed to all relevant staff. The degree and extent of intervention that would be in the best interest of the infant were explained to the family. These included the number of inflation breaths if there were any signs of life, nutrition support, and possible investigations such as echocardiogram and renal ultrasound scan. In addition, the mother received full bereavement support care. The mother had prepared the means to care for the baby including clothes and diapers which were used after birth. The mother was satisfied with the care process and also with the outcome of the pregnancy. This seems to support the literature that continuation of pregnancy in lethal fetal defects may have psychological benefit to women compared to termination.<sup>[9]</sup>

The psychological benefits could be due to several reasons. First, the time from diagnosis to birth gives the opportunity to come to terms and accept the eventual outcome. For some mothers, it may also be more socially and religiously acceptable to continue with pregnancy than opt for termination. Furthermore, in this case, the mother had prepared all the essential items for after birth. The stillborn infant was clothed,

and his photographs were taken. He also received a full funeral ceremony.

## Conclusion

This case shines a light on clinically important practical issues in recurrent anencephalic pregnancies carried to term. It highlights the need for a thorough and non-directive discussion with parents with full involvement of a MDT to prepare for all possible outcomes.

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