

# A Case of Delivery of a Near Term Anencephalic Baby in an Unbooked Case: A Case Report

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**Abstract:** Anencephaly is a lethal defect characterized by absence of brain and cranium above the base of the skull and orbits. It can be diagnosed in the late 1st trimester, and with adequate visualization, virtually all cases may be diagnosed in the 2nd trimester when there is difficulty in measuring the biparietal diameter. Termination of pregnancy is offered to all the prenatally diagnosed cases. We describe the unbooked case of a woman, who presented at 34 weeks of pregnancy with pain abdomen with undiagnosed diabetes with anencephalic fetus on ultrasonography. The objective of this case report is to highlight the importance of an early ultrasonic diagnosis of anencephaly, which could help in thorough evaluation and active management. Also, the main role of folic acid in the prevention in neural tube defects.

**Keywords:** anencephaly, congenital malformations, delivery, folic acid supplementation, Hydramnios, pregnancy, termination, ultrasound.

## 1. Introduction

Anencephaly is a congenital malformation of the central nervous system that results in the failure of closure of the cranial end of the embryologic neural tube, usually occurs between the 23rd to 26th day of conception. Anencephaly represents 40% of neural tube malformations, which is the second leading cause of nervous system abnormalities after spina bifida. The prevalence rate is 1 / 1000. The diagnosis is made by the 1st trimester ultrasound between the 11th and 14th week. The causes are multifactorial (iatrogenic, toxic, metabolic, nutritional and exceptionally chromosomal).

## 2. Case Report

This case was reported and analysed in OBG department of Jamnabai General Hospital, Vadodara, and Gujarat, India. Mrs SP, 22 years old, rural geographic origin, the patient had no previous medical or surgical history, no notion of consanguinity, gravid 2, para 1. Her G1 was by vaginal delivery with episiotomy of a live born female weighing 3.20kg, G2 is the current pregnancy estimated at 34 weeks given by her last menstrual period. She was not registered at any healthcare facility, no antenatal checkups till date, no history of folic acid

in periconceptional period or in 1st trimester, no history of iron and calcium supplements, no history of injection tetanus toxoid, no routine antenatal blood tests including ultrasonography in any trimester. She presented to outpatient department at 34 weeks with complaints of pain abdomen. There was no significant significant past, obstetric or surgical history. On general examination, the patient was clinically stable, height 150cm, weight 65kg, no edema of lower limbs, blood pressure 110/60mmhg, pulse rate 88bpm, Spo2 98% and temperature 36.6. Obstetric examination: intermittent uterine contractions present, 33 cm uterine height, fundal height 36 weeks, fetus was longitudinal position, fetal heart rate was 142bpm, liquor seemed clinically on higher side evidenced by tensed abdomen and unable to identify the presenting part. On vaginal examination, cervix was 2 cm dilated with 20% effacement, intact membranes with breech presentation. Emergency ultrasound revealed a single intrauterine fetus, 33 weeks gestation in breech presentation, positive cardiac activity with anencephaly, with polyhydramnios Single vertical pocket (SVP) of 15cm with abnormal spinal curvature.



Fig. 1. Ultrasonography scan of the anencephalic fetus

Above is the ultrasonography scan of the anencephalic fetus (produced after permission from the couple). All the necessary antenatal investigations done, including random blood sugar which was 180mg/dl. HbA1c was done which was 6.2. After explaining the condition of the fetus to the patient and her

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relatives and after an informed and written consent labour induction was done with vaginal misoprostol tablets since she had an unfavourable bishop score. After 3 doses of 50mcg pv misoprostol, patient went into active labour and delivered a newborn female with birth weight 2.1kg apgar score was 5 and 7 at 1 and 5 mins respectively. A polymalformative syndrome: clubbed feet, low set ears, protruding eyeballs, hydrothorax, kyphosis of thoracic spine, thoracolumbar spina bifida were present. Death of the newborn occurred at 2 hrs of life. The mother was diagnosed as gestational diabetes and was put on insulin therapy and dietary advice given. She was then advised to follow up for further workup and management. Below are the actual photographs of the anencephalic baby with multiple anomalies (produced after permission from the couple).



Fig. 2. Anencephalic baby with multiple anomalies

### 3. Discussion

Anencephaly is one of the most common types of neural tube defects after spina bifida affecting approximately 1 in 1,000 pregnancies. A baby born with anencephaly is usually blind and deaf. Females outnumber males in cases of anencephaly. The female preponderance noticed by previous authors were David TJ 2 at 66%, Panduranga 3 at 56%, Aruna<sup>5</sup> recorded 55%. Many earlier studies reported cleft lip and palate as well as genital abnormalities to be more common in male anencephalic fetus. Neural tube defects (NTDs) are more common in insulin-dependent diabetes in the 1st trimester of pregnancy, the prevalence of these malformations varies from 4-15% (2.1% in general population). In absolute value, this risk is elevated because it increases from 2% for an HbA1C of 5.5 to 6% for an HbA1C of 96. Underlying diabetes which was undiagnosed could be one of the causes of anencephaly in our case. Antenatal diagnosis of anencephaly in the first trimester is most successful for fetal abnormalities. In the second trimester ultrasound, the typical appearance of anencephaly is a sign of “frog eyes”, due to the absence of visible brain tissue above the eye sockets<sup>2</sup>. Skeletal abnormalities found in previous studies were in the range of 1.7% by David TJ, 14.5% by Vare *et al.*<sup>8</sup>, to 20% by Tan *et al.*<sup>4</sup> Our case did not have an antenatal ultrasound done so it was not diagnosed earlier because termination of pregnancy at an earlier gestational age is less traumatic both physically and mentally for the patient and her family. Hydramnios is associated with anencephaly in 30-50% of cases as was present in our case. The contributing factors include secretion of cerebrospinal fluid into the amniotic cavity, lack of normal swallowing, lack of absorption of amniotic fluid by the hypoplastic lungs and excessive urine production due to lack of antidiuretic hormone. Hydramnios is the most common presentation of anencephaly before childbirth<sup>1, 2</sup>. Anencephaly is a uniform lethal anomaly. It appears to be of multifactorial origin, so women are advised to consume rich in folic acid at least 3 months before planning pregnancy. Deficiency of folic acid appears to be one cause of anencephaly in our case. Following studies carried out on primary and secondary prevention, Canadian, British and American organizations recommend that women of childbearing age consume 0.4mg to 0.8mg/day of folic acid to reduce the risk of anencephaly. For women who have already have an affected child, the recommended dose is between 0.8 to 4mg/day.

### 4. Conclusion

The antenatal diagnosis of anencephaly is mainly based on obstetric ultrasound. The prognosis of this anomaly is grim. It is a uniformly lethal defect. Termination of pregnancy is the most logical approach<sup>1</sup>. Unfortunately, in the absence of curative treatment, prevention is therefore essential and creating awareness among the people about the preventable causes of nutritional deficiency and the importance of early registration of pregnancy and antenatal ultrasound. Therefore, folic acid plays an important role in preconception or in the 1st trimester. The presence of associated abnormalities like spina bifida, clubbed foot, hydrothorax points to the fact that

anencephaly consists of more than one aetiological entity. Studies are required at molecular level to find its association with other anomalies. In Indian scenario, people from rural backgrounds still believe in reporting to hospital only if there arises a problem. This was an uncommon case in today's world where obstetric and ultrasound facilities are available at almost every health care facility and despite of which this patient did not register herself early because of which folic acid supplementation could not be given to her, her diabetic status was missed and diagnosis of anencephaly could not be made due to no antenatal ultrasound.

### 5. Compliance with Ethical Standards

Conflict of interest: the authors declare that they have no conflict of interest. Informed consent: informed consent was obtained from the couple for the case report and assurance was

given to maintain their privacy.

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