

# Successful Outcome of Pregnancy in $\beta$ -thalassemia Major Individual

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## Abstract

Thalassemia major also called as Cooley's anemia, it has a codominant inheritance and pathology lies in decreased synthesis of beta chains resulting in increased production of alpha chains, which subsequently leads to red cell destructions, ineffective erythropoiesis, and anemia. A 23-year-old primigravida, married life 1 year (height 158 cm, weight 61 kg) with thalassemia major presented in the antenatal care outpatient department at Santokba Durlabhji Memorial Hospital with 6 weeks gestation. The patient had severe form  $\beta^0/\beta^+$  thalassemia major, and her thalassemia was identified at the age of 2 years. Both parents were thalassemia trait, and her sibling had normal hemoglobin pattern. Her antenatal period was managed well, with blood transfusion almost every week. She presented to labor room with 35+6 weeks of gestation in labor at 11 pm and not willing for normal delivery therefore under general anesthesia cesarean section was done and an alive female baby with birth weight 2.6 kg was delivered at 11:50 pm with Apgar 8 at 1 min and 10 at 5 min. Surgery was uneventful, and blood loss was minimal. This case report aims to highlight important issues associated with pregnancy in  $\beta$ -thalassemia major patient and obstetric management.

**Key words:** Blood transfusion, Chelation therapy, Gestation, Hemoglobin

## INTRODUCTION

The  $\beta$ -thalassemias are distributed widely across the Indian sub-continent, Mediterranean region and throughout the Southeast Asia, and can also occur sporadically in many ethnic groups. According to statistics, 45% of the world population is affected by thalassemia; out of this population, in India, 3.5% are carriers of thalassemia.  $\beta$ -thalassemia minor does not influence the pregnancy outcome in the negative way significantly.<sup>1</sup>

Since late 1970s, the pediatric and hematological management of patients with  $\beta$ -thalassemia major has improved significantly due to advances in medical care, ready availability and experience in extensive blood transfusions

and iron chelation therapy. However, complications such as pulmonary hypertension, thromboembolic complications, overwhelming postsplenectomy sepsis, and the development of hepatocarcinoma may reduce survival in this group of patients.<sup>2</sup> Even with these medical advances a case of  $\beta$ -thalassemia major carrying a pregnancy to term and with a pregnancy to term and with a successful outcome is rarity.

According to the recent discoveries of psychoneuro endocrinology showing the relation between biological function mediated by the immune system and psychological status, therefore evaluation of the psychological life will have to be included within the regular clinical investigations on thalassaemic pregnant women.<sup>3</sup>

## CASE REPORT

A 23-year-old primigravida, married life 1 year (height 158 cm, weight 61 kg) with thalassemia major presented in antenatal care (ANC) outpatient department at

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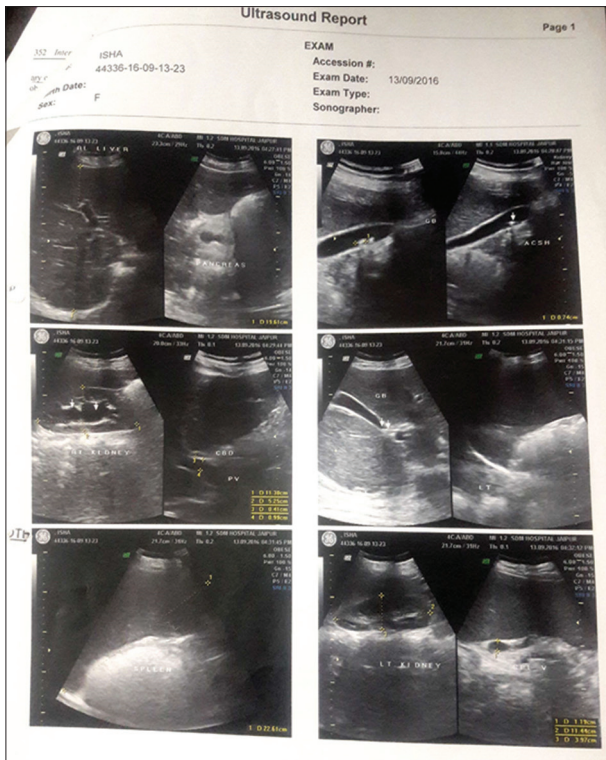
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**Figure 1: USG showing hepatosplenomegaly with spleen 22.6cm with dilated splenic vein**

Santokba Durlabhji Memorial Hospital with 6 weeks gestation. The patient had severe form  $\beta^0/\beta^+$  thalassemia major, and her thalassemia was identified at the age of 2 years. Both parents were thalassemia trait, and her sibling had normal hemoglobin pattern. The patient had received lifelong blood transfusions along with chelation therapy regularly since 2 years of age at Santokba Durlabhji Memorial Hospital. Her developmental milestones had been normal, with menarche at the age of 12 years followed by regular menstrual periods. Pregnancy was spontaneously conceived, her partner had a normal hemoglobin pattern, and therefore fetus was not subjected for prenatal diagnosis for thalassemia. The patient had normal thyroid function pre-pregnancy but her thyroid-stimulating hormone (TSH) in the first trimester was 5.83, therefore, she was put on tablet Eltroxin 50  $\mu$ g OD, on this her TSH was  $<2$  in next trimesters. Other ANC checkups were normal. Her nuchal translucency scan, double marker, and congenital anomaly scan were normal. Before pregnancy, she had a blood transfusion at every 14<sup>th</sup> or 15<sup>th</sup> day to maintain hemoglobin at 10 g/dl, in pregnancy she had transfusion weekly. In her second trimester at 24+3 weeks, she was admitted for 3 days in view of high-grade fever with hemoglobin 8.6 g/dl, which was later diagnosed as malaria, managed accordingly.

There was evidence of thalassemic facies and hepatosplenomegaly with spleen measuring about 22.6 cm with dilated splenic veins about 11.9 mm and enlarged liver with coarse echocardiography (ECHO) texture. As soon her pregnancy was confirmed, her cardiac function was assessed by ECHO which was normal. Her lifelong care had been managed by pediatric specialist at our center. Her ANC was jointly managed by obstetric and pediatric hematological departments at our institution. The chelation therapy, desferrioxamine, was stopped when pregnancy had been confirmed due to the risk of teratogenicity and fetal iron deficiency in late pregnancy and restarted 1 month after delivery.

At 34 weeks gestation, repeat reference done with a cardiologist and cardiac evaluation done which was within normal limit and prophylactic steroids given to her for fetal lung maturity. She presented to labor room with 35+6 weeks of gestation in labor at 11 pm and not willing for normal delivery therefore under general anesthesia cesarean section was done and an alive female baby with birth weight 2.6 kg was delivered at 11:50 pm with Apgar 8 at 1 min and 10 at 5 min. Surgery was uneventful, and blood loss was minimal.

The post-operative period was uneventful. Her post-operative hemoglobin was 9.7 g/dl, and there were no postpartum complications, and both mother and baby were discharged after 3 days. She started her routine follow-up with pediatric hematologist post-delivery. Iron-chelating therapy was restarted after 1 month, and routine hematological and cardiovascular review was done.

## DISCUSSION

Thalassemia major also called as Cooley's anemia, it has a codominant inheritance and pathology lies in decreased synthesis of beta chains resulting in increased production of alpha chains, which subsequently leads to red cell destructions, ineffective erythropoiesis, and anemia. Since repeated blood transfusions are required in thalassemia major, there is iron overload which in turn causes deposition of iron in the hypothalamus and the pituitary causing reproductive axis failure leading to delayed puberty, delayed sexual development, and infertility.

Improvements in managing  $\beta$ -thalassemia major have allowed many patients to survive beyond puberty. Fertility can be impaired as a result of iron overload related hypogonadism, although assisted reproductive

techniques and advances in treating iron overload have increased the number of successful pregnancies in such patients.

## CONCLUSION

This case report aims to highlight important issues associated with pregnancy in  $\beta$ -thalassemia major patient and obstetric management. Our patient had a successful cesarean delivery under general anesthesia as a result

of careful monitoring and treatment of her condition antenatally and closes multidisciplinary approach to ensure optimal management of her pregnancy.

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