

Pregnancy with Gilbert Syndrome – A Case Report

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ABSTRACT

A primigravida presented to us at 32 weeks of gestation with vomiting, myalgia and jaundice. On examination she had icterus, she was dehydrated, uterus was corresponding to dates and the fetal heart rate was good. On evaluation, all the investigations were normal except mild unconjugated hyperbilirubinaemia and hypoglycaemia. Based on the above findings we derived at a diagnosis of Gilbert syndrome. Dehydration due to vomiting aggravated her jaundice. On correcting her dehydration jaundice resolved, patient improved symptomatically and was discharged two days later. She was later admitted at term and underwent emergency caesarian section in view of fetal distress. Mother and baby were fine postoperatively and was discharged on the fifth postoperative day Gilbert syndrome is rare in obstetric practice. Virtually all patients have decreased activity of Uridine diphosphate glucuronosyl transferase (UDPGT). The case is reported due to its rarity.

Keywords: Hyperbilirubinaemia, UDP Glucoronyltransferase

CASE REPORT

A primigravida presented to our emergency room at 32 weeks with severe vomiting, myalgia, headache and yellowish discoloration of sclera since three days. She was appreciating fetal movements well. No history of itching or pale colour stools. The patient gave history of similar complaints at 16 & 24 weeks and was treated conservatively with IV fluids. She did not have similar episodes before pregnancy. On examination she was conscious, was dehydrated, had icterus without pallor. Her blood pressure was found to be 90/60 mm of Hg. Uterus corresponded to dates. The fetal heart rate was 148/mt. The investigations revealed urine ketone bodies 4+, RBS 54mg/dL, Liver function test was normal except Serum Bilirubin which was 6mg/dl, the indirect being 5.4mg/dL. Viral screening for hepatitis was negative. Reticulocyte count was normal. Scan showed a single live fetus corresponding to dates with adequate liquor, Liver was normal, No signs of obstruction. Patient was treated with I.V. fluids to correct dehydration. With correction of dehydration, jaundice resolved spontaneously, after 48 hours serum bilirubin decreased to 2mg/dL. The patient improved symptomatically and was discharged. She later presented to us at 38 weeks with prelabour rupture of membranes. In view of fetal distress she was taken up for emergency caesarian section and delivered a healthy female child of 2.8 kgs. Mother and baby were fine postoperatively. Baby was observed for hyperbilirubinaemia for five days. Both mother and baby were discharged on day five. Later she came at six weeks for a routine postnatal check up and was found to be fine. Our patient presented with constitutional symptoms, hypoglycaemia and jaundice which was aggravated due to dehydration following vomiting. On evaluation indirect bilirubin was raised and there were no signs of haemolysis. Patient condition improved on correcting dehydration and the jaundice resolved spontaneously. These salient features helped us to arrive at the diagnosis of Gilbert Syndrome.

DISCUSSION

Gilbert Syndrome is a benign often familial condition characterized by recurrent mild unconjugated hyperbilirubinaemia in the absence of haemolysis or underlying liver diseases. Augustine Gilbert and Pierre Lerebulet first described Gilbert syndrome in 1901. Although

various investigators have used other names for this disorder, such as constitutional hepatic dysfunction, hereditary haemolytic bilirubinaemia, and familial nonhaemolytic jaundice, Gilbert syndrome is the most commonly used name for this condition [1]. Gilbert syndrome is rarely diagnosed before puberty though it is a congenital disorder. Hormonal changes of puberty have been suggested as one explanation [2]. Dehydration, fasting or stress precipitates Gilbert syndrome [3]. It is found in 7% of general population [4]. It is common among men than women in the ratio of 2-7:1 [5]. The hyperbilirubinaemia is mild and by definition < 6mg/dL. In Caucasians a genetic defect in the TATA box of the promoter region of the gene encoding for bilirubin UDP-Glucuronyltransferase is tightly associated with Gilbert's syndrome [6]. Bilirubin is an endogenous antioxidant [7]. Gilbert's syndrome may actually reduce the risk of various age-related diseases because of the antioxidant properties of bilirubin. Interestingly, one recent study has found that mortality rates observed for people with Gilbert syndrome in the general population were shown to be almost half those of people without evidence of Gilbert syndrome [8].

Thirty percent of the patients with Gilbert Syndrome are usually asymptomatic. Some patients present with fatigue, nausea, loss of appetite, jaundice, vomiting, hypoglycaemia, itching and pain abdomen of which itching and pain abdomen were absent in our patient. These symptoms are usually precipitated by infection, dehydration or stress and in our patient these were precipitated by dehydration. In Gilbert Syndrome there is recurrent mild jaundice and our patient also presented with mild jaundice at 16 and 24 weeks. We first excluded other inherited causes of unconjugated hyperbilirubinaemia such as Crigler Najjer Syndrome, in this syndrome bilirubin is raised up to 20mg/dl whereas in our patient it was raised up to 6mg/dl only. The other causes of haemolysis and infective causes of jaundice were excluded. Gilbert Syndrome was diagnosed by exclusion.

Alkaline methanolysis and thin-layer chromatography have been used to diagnose Gilbert syndrome by accurately separating and measuring total serum as conjugated and unconjugated fractions. High-performance liquid chromatography (HPLC) of serum showed similar findings with significantly decreased bilirubin monoglucuronides (1.1% vs 6.2% in normal) and increased

unconjugated bilirubin (98.8 vs 92.6 in normals) [3]. The polymerase chain reaction (PCR) is a novel and rapid method of identifying genetic polymorphisms in the TATA box of the UDPGT1 gene using fluorescence resonance energy transfer. Gilbert syndrome is self-limiting and benign with good prognosis.

This case is reported for its rare incidence in pregnancy and its typical clinical features with which the patient presented and its benign nature. To conclude when any patient presents with unconjugated hyperbilirubinaemia associated with stress, infection or dehydration Gilbert Syndrome must be excluded. Once this diagnosis is made patient must be reassured of its benign nature, excellent prognosis and normal life expectancy.

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