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Case Report

Pregnancy with Gilbert syndrome

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ABSTRACT

A rare case of Gilbert syndrome during pregnancy was reported at Narayana Medical College and Hospital's obstetrics department. Due to the fact that it was an extremely rare case with a typical presentation, it was properly studied and analysed to provide a thorough case report. A 36-week-pregnant primigravida who had been having nausea, vomiting, stomach pain, myalgia, and a yellowish discoloration of both the skin and sclera for the past five days presented to Narayana Medical College and Hospital. She consistently suffered having similar symptoms at 14 and 24 weeks. She had icterus, signs of dehydration, a uterus that corresponds to period of gestation, and a good fetal heart rate. Examination revealed that all of the obtained investigations were normal, with the exception of mild unconjugated hyperbilirubinemia and hypoglycemia. After the dehydration and jaundice were treated, the patient's symptoms improved, and she was discharged from the hospital after a week. Later she underwent an emergency caesarean section due to fetal distress after being admitted during pregnancy at term, and she gave birth to a 3 kg female baby. Mother and baby both were fine postoperatively. Due to its rarity, this case was documented. Almost all of the patients had decreased uridine diphosphate glucuronosyl transferase activity (UDPGT).

Keywords: Hyperbilirubinemia, Gilbert syndrome, UDP glucuronosyl transferase

INTRODUCTION

Gilbert syndrome is a benign and the most common inherited disorder of bilirubin metabolism characterized by recurrent mild unconjugated hyperbilirubinemia in the absence of hemolysis or underlying liver disease. Augustine Gilbert and Pierre Lerebullet first described Gilbert syndrome in 1901.[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, exercise, menstruation or stress precipitates Gilbert syndrome.[3] It is found in 7% of the general population. It is more common among men than women in the ratio of 2-7:1.[4,5]

CASE REPORT

A 36-week-old primigravida who had been experiencing severe vomiting, myalgia, headache, and yellowish dicolouration of sclera for five days prior presented to our emergency department. She was certainly able to appreciate foetal movements. No prior history of itching or light-colored stools. The patient revealed a history of such similar complaints at 14 and 24 weeks, and IV fluids were given as a conservative measure. Before becoming pregnant, she had no similar episodes. She was found to be aware, dehydrated, and suffering from icterus without pallor. Her blood pressure was 90/60 mm Hg. Dates and uterus matched. The foetal heart rate was 148 bpm. Investigations showed that the liver function test was normal, except for the serum bilirubin, which was 6 mg/dl

and the indirect, which was 5.5 mg/dl, and that urine ketone bodies were 3+, RBS was 54 mg/dl. Hepatitis virus testing came out negative. Reticulocyte count was normal. The scan revealed a single viable foetus that corresponded to dates with adequate liquor. The liver was normal and showed no abnormalities. IV fluids were administered to the patient to treat dehydration. Jaundice resolved on its own after the dehydration was corrected, and after 48 hours, the blood bilirubin level fell to 2 mg/dl. The patient showed symptomatic improvement and was discharged. She later presented to us at 38 weeks with pre-labour rupture of membranes. Due to foetal distress, she underwent an emergency caesarean section, giving birth to a 3 kg healthy female child. Following surgery, both mother and child were in good health. For five days, the baby had monitoring for hyperbilirubinemia. On the fifth day, both the mother and the infant were discharged. At six weeks, she came for a routine postnatal check-up, and things were good. The patient had constitutional symptoms, hypoglycemia, and jaundice, which were made worse by dehydration from vomiting. Upon evaluation, indirect bilirubin was elevated and there were no symptoms of hemolysis. After dehydration was treated, the patient's condition got better, and the jaundice disappeared on its own. These distinguishing characteristics assisted us in making the diagnosis of Gilbert syndrome. As a result, Gilbert syndrome must be ruled out whenever a patient has unconjugated hyperbilirubinemia linked to stress, an illness, or dehydration. Patients must be reassured of the diagnosis' benign nature, excellent prognosis, and normal life expectancy after it has been made.

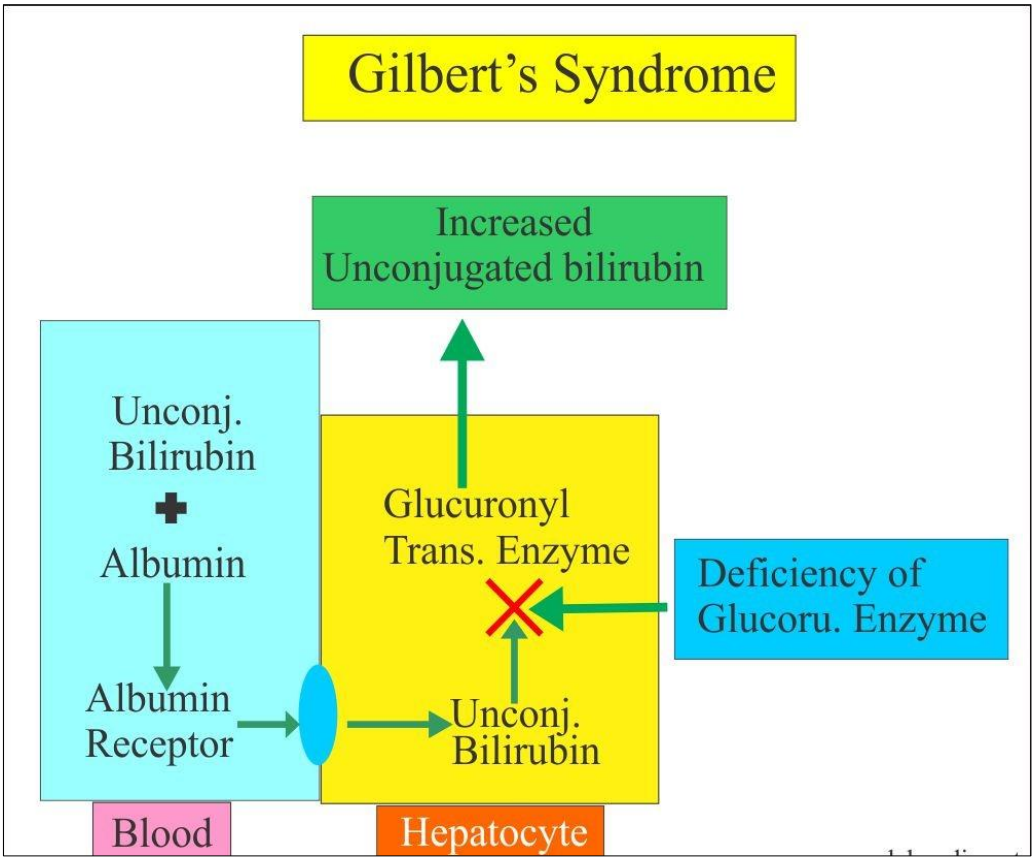

Figure 1: Mechanism of Gilbert syndrome.

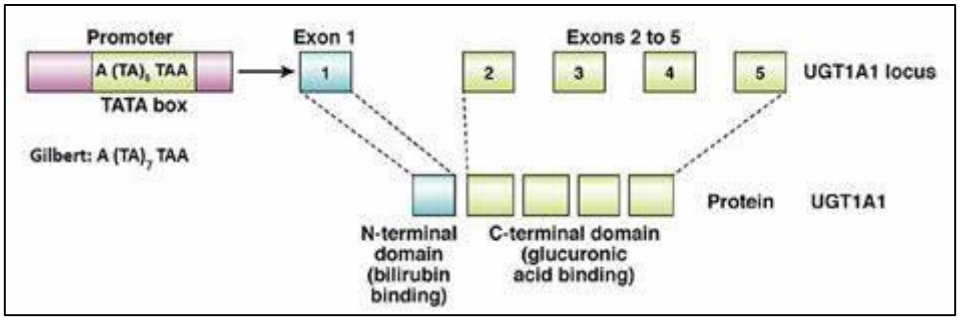

Figure 2: Genetic basics of Gilbert syndrome.

Table 1: Differential diagnosis of hereditary jaundice with normal liver chemistries and no signs or symptoms of liver disease.

Unconjugated hyperbilirubinemia			
Crigler-najjar syndrome			
	Gilbert syndrome	Type I	Type II
Incidence	<7% of population	Very rare	Uncommon
Inheritance mode	Autosomal Dominant	Autosomal Recessive	Autosomal dominant
Serum bilirubin total (mg/dl)	<3; ≤6	>20	<20
Defect hepatic UDPGT activity	Decreased	Absent	Markedly decreased
Age at onset of jaundice	Adolescence	Infancy	Childhood

DISCUSSION

Gilbert syndrome is a benign, frequently hereditary syndrome marked by recurring, mild unconjugated hyperbilirubinemia that does not include hemolysis or underlying liver conditions. Gilbert syndrome was initially defined in 1901 by Augustine Gilbert and Pierre Lerebullet. Gilbert syndrome is the most popular nomenclature for this ailment, despite the fact that other researchers have also used terms like constitutional hepatic dysfunction, hereditary haemolytic bilirubinaemia, and familial non haemolytic jaundice to describe it.[1] Gilbert syndrome is a congenital condition, however it is rarely identified before puberty. One theory is that puberty-related hormonal changes are to blame.[2] Gilbert syndrome is triggered by stress, fasting, or dehydration.[3] 7% of the general population have it.[4] In a ratio of 2-7:1, more men than women experience it.[5] By definition, the hyperbilirubinemia is mild and is less than 6 mg/dl. Gilbert's syndrome is strongly related with a genetic abnormality in the TATA box of the promoter region of the gene producing bilirubin UDPGT in Caucasians (Figure 1 and 2).[6]

Bilirubin is a natural antioxidant.[7] The antioxidant qualities of bilirubin suggest that Gilbert's syndrome may actually lower the risk of a number of age-related disorders. It's interesting to note that a recent study indicated that mortality rates in the general community for those with Gilbert syndrome were roughly half as high as for those without any signs of the disease.[8] Gilbert syndrome patients often have no symptoms in 30% of cases. Some people come with fatigue, nausea, loss of appetite, jaundice, vomiting, hypoglycemia, itching, and pain abdomen. These symptoms are typically driven on by stress, dehydration, or infection, however in this patient, dehydration was the cause. Mild jaundice recurs frequently in Gilbert syndrome, and this patient also experienced it at 14 and 24 weeks of pregnancy. We started by ruling out other inherited forms of unconjugated hyperbilirubinemia, such as Criggler Najjer syndrome, as our patient only had a 6 mg/dl increase in bilirubin rather than the 20 mg/dl increase seen in this syndrome (Table 1). Both infectious causes of jaundice and other causes of hemolysis were ruled out. Gilbert syndrome was diagnosed by exclusion. Gilbert syndrome had been identified by precisely separating and quantifying total serum as conjugated and unconjugated fractions using alkaline methanolysis and thin-layer chromatography. Similar results were seen by high-performance liquid chromatography (HPLC) of serum, which revealed considerably higher unconjugated bilirubin levels and much lower bilirubin monoglucuronides. To find genetic variations in the TATA box of the UDPGT1 gene utilising fluorescence resonance energy transfer, researchers have developed the polymerase chain reaction (PCR). The prognosis is good, and Gilbert syndrome is benign and self-limiting.

CONCLUSION

Gilbert syndrome must be ruled out whenever a patient exhibits unconjugated hyperbilirubinaemia linked to stress, an illness, or dehydration. Once the diagnosis has been made, the patient needs to be reassured of the condition's benign nature, good prognosis, and typical life expectancy.

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REFERENCES

1. Thomas DB. Zakim and Boyer's Hepatology A textbook of liver disease. 6th ed. Philadelphia: Saunders/Elsevier; 2012: 1101-3.
2. Kleinman R, Goulet O, Mieli-Vergani G, Sanderson I, Sherman P, Shneider B. Walker's Pediatric gastrointestinal disease. 5th ed. USA: PMPH; 2008: 770-2.
3. Arias IM. Inheritable and congenital hyperbilirubinemia. N Engl J Med. 1971;285(25):1416-21.

4. James DK, Steer PJ, Weiner CP, Gonik B. High risk pregnancy management options. 4th ed. Philadelphia: Saunders; 2010: 842.
5. Longo, Dan L, Fauci A, Kasper D, Hauser S, Larry J. Harrison's principles of internal medicine. 18th ed. McGraw Hill Education; 2011: 862.
6. Zusterzeel PL, Morsche RT, Raijmakers MT, Peters WH, Steegers EA. Gilbert's syndrome is not associated with HELLP syndrome. BJOG. 2001;108(9):1003-4.
7. Maruhashi T, Soga J, Fujimura N. Hyperbilirubinemia, augmentation of endothelial function, and decrease in oxidative stress in Gilbert syndrome. Circulation. 2012;126(5):598-603.
8. Horsfall LJ, Nazareth I, Pereira SP. Gilbert's syndrome and the risk of death: a population-based cohort study. J Gastroenterol Hepatol. 2013;28(10):1643-7.

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