

A case of dubin johnson syndrome in twin pregnancy

Abstract

Background: Dubin Johnson syndrome is a variety of inherited hyperbilirubinemia and is recognized by a low elevation of conjugated bilirubin without other signs of liver damage.

Objective: To present perinatal results of a patient diagnosed with Dubin Johnson syndrome.

Methods: The clinical file of a patient diagnosed with Dubin Johnson syndrome during pregnancy, treated at the Instituto Nacional de Perinatología was reviewed.

Results: 34-year-old patient with no history of fever, pruritus, malaise or abdominal pain; Due to the macroscopic characteristics of the liver during a laparoscopic cholecystectomy at 23 years of age, a biopsy was taken which confirmed the diagnosis of Dubin-Johnson syndrome. She began prenatal control at 17 weeks gestation. Serial bilirubin determinations ranged between 3.4 and 4.5 mg / dl. Intrauterine growth restriction was presented in one of the fetuses.

Conclusion: Dubin Johnson syndrome is a benign condition, the disease must be differentiated from other conditions that present hyperbilirubinemia specifically in pregnancy in order to stratify management.

Keywords: pregnancy, dubin johnson, syndrome

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Introduction

In 1954 Dubin and Johnson described a clinical entity characterized by idiopathic chronic jaundice with a specific pigment identified in the liver.¹ This relatively rare condition is a variety of hereditary hyperbilirubinemia and is recognized by a low elevation of conjugated bilirubin without other signs of liver damage. It is the result of a mutation that leads to inappropriate excretion of bilirubin from the hepatocyte. The condition is benign, has no long-term consequences and does not require medical management.² The case of a woman who had a multiple pregnancy is presented, having as a background the diagnosis of Dubin-Johnson syndrome nine years before the current pregnancy.

Methods

The clinical file of a patient diagnosed with Dubin Johnson syndrome during pregnancy, treated at the Instituto Nacional de Perinatología was reviewed.

Case report

34-year-old patient with no history of fever, pruritus, malaise or abdominal pain; No tobacco or alcohol consumption. No history of blood transfusion, intravenous drug abuse, chronic drug use or occupational hazards. Laparoscopic cholecystectomy at 23 years of age where given the macroscopic characteristics of the liver, a biopsy was taken which confirmed the diagnosis of Dubin-Johnson syndrome. From the current pregnancy, at 27, we do not know the history in relation to jaundice. She started prenatal control at our institution when she was pregnant with 17 weeks of gestation twin

pregnancy. Serial bilirubin determinations ranged between 3.4 and 4.5 mg/dl. Intrauterine growth restriction was diagnosed in one of the fetus without hemodynamic compromise. The pregnancy was interrupted at 38 weeks gestation by caesarean section where two live newborns with weights of 1878 and 2862 gr were obtained, both presented physiological jaundice that resolved spontaneously

Discussion

Dubin-Johnson syndrome originates from a mutation in the ABCC2 gene which provides instructions to produce a protein called MRP2 (multidrug resistance protein 2) which works as a transporter protein and is essential for excretion of conjugated bilirubin outside hepatocyte within the bile duct system; Conjugated bilirubin accumulates in the hepatocyte and the bilirubin levels in the blood rise.³ Dubin Johnson syndrome is rare, typically manifested in adolescence or young adult stage and is found in all races with a similar prevalence.⁴

The hepatic architecture is normal but there is accumulation of a dark pigment that is not iron or bile changing its color macroscopically. Hyperbilirubinemia when found is an incidental finding. In women, the condition may be subclinical and only present with jaundice when contraceptives are used or when they are pregnant, with very few reports being the association between Dubin Johnson syndrome and pregnancy.⁵⁻¹⁰ The clinical examination of patients is normal. Itching is not a symptom. Patients have predominantly conjugated hyperbilirubinemia and concentrations typically range between 2 and 5mg/dL. Other liver function tests such as transaminases, alkaline phosphatase, gamma-glutamyltransferase, coagulation tests and bile acids are normal. Liver biopsy is not recommended to establish the diagnosis.

It is a benign condition that does not progress to fibrosis or cirrhosis and does not require treatment. The differential diagnosis includes obstructive conditions of the biliary tract, acute or chronic liver damage, Rotor syndrome, Gilbert's syndrome, Crigler-Najjar syndrome and porto-systemic shunt, when discovered during pregnancy, intra-hepatic cholestasis, acute fatty liver of pregnancy and HELLP syndrome should be ruled out. Viral hepatitis is the most common cause of jaundice in pregnancy, followed by intra-hepatic cholestasis of pregnancy. In the neonatal period the disease is rarely present and should only be suspected when there is unexplained hyperbilirubinemia of moderate to severe range and long evolution.^{2,4,10}

Conclusion

Dubin Johnson syndrome is a benign condition, the disease must be differentiated from other conditions that present hyperbilirubinemia specifically in pregnancy in order to stratify management.

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Conflicts of interest

The authors declare there are no conflicts of interest and nothing to disclose.

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