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# Crigler-Najjar Syndrome: Case Report

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## ABSTRACT

High levels of toxic bilirubin in your blood cause Crigler-Najjar syndrome, a rare genetic condition. Bilirubin is a substance that forms when red blood cells reach the end of their lifespan. Your liver breaks down bilirubin from a toxic substance to a nontoxic substance that you get rid of in your stool. If you're diagnosed with Crigler-Najjar syndrome, your liver can't break down bilirubin and it collects in your blood. As a result, Crigler-Najjar syndrome can cause life-threatening symptoms if left untreated.

***KEYWORDS:*** *crigler-najjar, bilirubin, choleliathaisis*

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## INTRODUCTION

Crigler-Najjar syndrome is a severe condition characterized by high levels of a toxic substance called bilirubin in the blood (hyperbilirubinemia). Bilirubin is produced when red blood cells are broken down. This substance is removed from the body only after it undergoes a chemical reaction in the liver, which converts the toxic form of bilirubin (called unconjugated bilirubin) to a nontoxic form called conjugated bilirubin. People with Crigler-Najjar syndrome have a build-up of unconjugated bilirubin in their blood (unconjugated hyperbilirubinemia). Crigler-Najjar syndrome is divided into two types. Type 1 (CN1) is very severe, and affected individuals can die in childhood due to kernicterus, although with proper treatment, they may survive longer. Type 2 (CN2) is less severe. People with CN2 are less likely to develop kernicterus, and most affected individuals survive into adulthood. Crigler-Najjar syndrome affects males and females in equal numbers. The incidence is estimated to be 1 in 750,000-1,000,000 people in the general population.

## CASE DESCRIPTION:

A 18 year-old female patient reported with yellowish discoloration of eyes past 4 weeks, high yellowish discoloration of urine, vomiting on and off past 3 weeks and also pain in upper abdomen non- radiating intermitted type of pain.

The physical examination revealed that yellowish discoloration is present all over the skin, sclera and all mucous membrane presented in yellowish colour. Baseline investigations showed haemoglobin of 8.4 g/dl, total leukocyte count 2500/mm3 and reticulocyte count 2.5% and peripheral smear revealed that evidence of hemolysis. Liver function tests revealed rise in total bilirubin of 46.38mg/dl, direct bilirubin 21.85mg/dl. Liver enzymes AST: 32 U/L, ALT:12 U/L. its indicating unconjucated hyperbilirubinemia. USG abdomen shows that cholelithasis with edematous GB wall- ? Cholecystitis and mild splenomegaly. The patient continuous to be monitored by multidisciplinary team.

**Fig.1: Crigler-najjar symptoms**

**DISCUSSION:**

Crigler- najjar syndrome is a rare inherited disorder affecting the metabolism of bilirubin. The diagnosis is based on clinical findings and confirmed by all base line investigations like haemoglobin, liver functions test, peripheral smear. Clinically patient had yellowish discoloration of all skin, eyes and mucous membrane. Cholecystitis treated by Endoscopic retrograde cholangiopancreatography. Patient also treated with antibiotics and oral bile salt therapy.

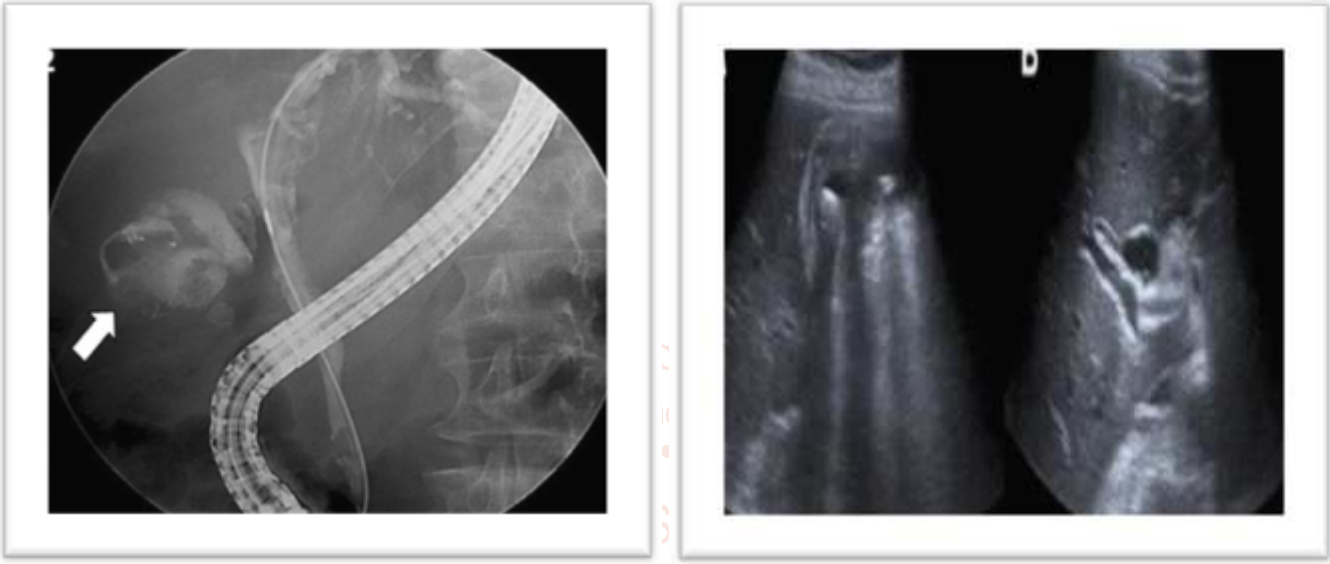

**Fig 2: a)Abdominal ultrasound showing gallbladder wall thickening, pericholecystic fluid and cholelithiasis. b) Endoscopic retrograde cholangiopancreatography showing multiple stones in the gallbladder.**

**CONCLUSION:**

In view of the wide clinical spectrum of this syndrome, the management of its modalities is not standardized. It recommended that patient will gone for plasmapheresis to removes toxic substances from the blood, liver transplant for healthy liver, Phenobarbital to prevent bilirubin from building up in blood. Crigler- najjar, if detected early in life, can be treated with good outcomes.

**CONFLICT OF INTEREST:**

None

**FUNDING:**

None

**CONSENT FOR PUBLICATIONS:**

Informed consent was obtained from the parents of the patients to publish this case in medical journal.

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