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A Black Liver During Laparoscopic Appendectomy: Incidental Diagnosis of Dubin-Johnson Syndrome

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Abstract

Dubin-Johnson syndrome (DJS) is a rare, benign autosomal recessive disorder that involves chronic conjugated hyperbilirubinemia and a distinctive black pigmentation of the liver. It is typically asymptomatic and often discovered incidentally. We report a case of a 23-year-old male patient who presented with acute appendicitis and mild jaundice. Laboratory work-up found isolated conjugated hyperbilirubinemia with all other liver enzymes normal. Laparoscopic appendectomy was performed, and the liver was found to be diffusely black with a smooth surface, which led to suspicion of underlying hepatic pathology. Postoperative clinical evaluation and family history supported the diagnosis. This case highlights the importance of recognizing DJS intraoperatively to avoid unnecessary diagnostic and surgical interventions.

Categories: General Surgery, Internal Medicine, Gastroenterology
Keywords: anesthesia, black liver, dubin-johnson syndrome, general surgery, hereditary hyperbilirubinemia, hyperbilirubinemia, incidental findings, laparoscopy

Introduction

Dubin-Johnson syndrome (DJS) is a rare autosomal recessive disorder associated with chronic conjugated hyperbilirubinemia secondary to defective hepatocellular excretion of bilirubin [1]. It is caused by abnormal function of the multidrug resistance-associated protein 2 (MRP2) due to an ABCC2 gene mutation, which causes retention of conjugated bilirubin in the hepatocytes [1-2].

The clinical manifestations of DJS are usually mild, intermittent, non-pruritic jaundice with normal liver enzyme levels [3,4]. It is a condition that generally is not symptomatic and is often identified incidentally on imaging, surgery, or investigation for another condition [5]. The disease occurs in both genders, although it manifests earlier in males, generally in the second decade of life. It is more prevalent in Iranian and Moroccan jews [6].

One of the distinguishing characteristics of DJS is the dark pigmentation of the liver, which is black, slate-gray, or even indigo blue in appearance intraoperatively [7,8]. This dramatic discovery could warrant concern for important hepatobiliary disease in the surgical context, especially if it occurs unexpectedly.

Although it is well described in the literature, intraoperative recognition and challenges during emergency surgical procedures are underreported, especially in patients with acute abdominal conditions like appendicitis.

We present a case of acute appendicitis in which DJS was incidentally identified intraoperatively, highlighting the importance of intraoperative recognition of this rare but benign condition in emergency surgical settings.

Case Presentation

A 23-year-old male came into the emergency department complaining of right lower abdominal pain with nausea for one day. There was no history of vomiting or previous similar episodes. The patient denied any known history of liver disease.

His temperature was 37.8°C, heart rate was 96 beats per minute, blood pressure was 118/76 mmHg, respiratory rate was 18 per minute, and oxygen saturation was 98% on room air on examination. On an abdominal examination, tenderness and rebound tenderness were observed in the right iliac fossa. Mild scleral icterus was also noted.

Laboratory examinations showed leukocytosis with a white blood cell count of 13.2 × 10^3/µL. The total bilirubin level was 3.4 mg/dL, of which the direct bilirubin was 3.2 mg/dL. Table *1* shows the trend of liver function tests and creatinine at baseline and post-operative follow-up visits. Serologic testing for anti-HCV antibody, hepatitis B surface antigen (HBsAg), and HIV was negative. Alkaline phosphatase was not obtained at baseline as the clinical presentation was consistent with appendicitis with low suspicion for cholecystitis or choledocholithiasis.

Visit / timepoint	Total bilirubin (mg/dL) (0.2–1.3)	Direct bilirubin (mg/dL) (<0.30)	Indirect bilirubin† (mg/dL)	AST (U/L) (17–59)	ALT (U/L) (21–72)	Creatinine (mg/dL) (0.6–1.25)	Alkaline phosphatase (ALP) (U/L) (38–126)
Visit 1 (Baseline)	3.4	3.2	0.2	17.4	22.4	0.85	-
Visit 2 Postoperative day 7	2.9	2.2	0.7	16.3	20.0	0.75	67
Visit 3 Postoperative day 21	3.1	2.5	0.6	19.0	25.0	0.75	-

TABLE 1: Trend of liver function and renal parameters at baseline and follow-up

Serial trend of bilirubin fractions, liver enzymes, and renal function across follow-up visits, demonstrating persistent predominantly conjugated (direct) hyperbilirubinemia with normal transaminases and renal function throughout the course characteristic of Dubin-Johnson syndrome.

AST: aspartate aminotransferase; ALT: alanine aminotransferase; ALP alkaline phosphatase

Abdominal ultrasound showed a right iliac non-compressible tubular structure about 7.5 mm in diameter, which was compatible with a diagnosis of acute appendicitis. Several gallstones were observed in the absence of gallbladder wall thickening or pericholecystic fluid. The common bile duct was 4 mm, and there was no evidence of intrahepatic biliary dilatation. In view of conjugated hyperbilirubinemia, differential diagnoses included Rotor syndrome, biliary obstruction, viral hepatitis, and sepsis-related cholestasis. These were less likely, however, because liver enzymes were normal, there was no dilatation of the bile ducts, and the viral serology was negative. A comparison between DJS and Rotor syndrome derived from Thakkar et al. is shown in Table *2* [9].

Feature	Dubin-Johnson syndrome	Rotor syndrome
Inheritance	Autosomal recessive (ABCC2)	Autosomal recessive, digenic (SLCO1B1 and SLCO1B3)
Defective protein	MRP2	OATP1B1 and OATP1B3
Site of defect	Canalicular membrane (impaired excretion into bile)	Sinusoidal membrane (impaired hepatic uptake)
Serum bilirubin	~2-5 mg/dL, >50% conjugated	~2-5 mg/dL, >50% conjugated
Urinary coproporphyrin	Normal total; >80% isomer I	Total increased 2.5-5 times; ~65% isomer I
Hepatobiliary scintigraphy	Intense, prolonged liver uptake with delayed excretion	Poor or absent liver uptake with prominent renal excretion
Liver appearance	Black (pigment deposition)	Normal
Effect of pregnancy/estrogens	Jaundice may worsen	No change
Drug toxicity risk	Uncommon	Increased for drugs cleared by OATP1B1/1B3

TABLE 2: Comparison of Dubin-Johnson syndrome and Rotor syndrome

ABCC2: ATP-binding cassette subfamily C member 2; MRP2: multidrug resistance-associated protein 2; SLCO1B1/SLCO1B3: solute carrier organic anion transporter family members 1B1/1B3; OATP1B1/OATP1B3: organic anion transporting polypeptides 1B1/1B3

Emergency laparoscopic appendectomy was performed. In the operating room, the appendix was found to be acutely inflamed. Incidentally, the liver was found to be diffusely black in color with a smooth surface, with no evidence of biliary obstruction or cirrhosis. We did not perform cholecystectomy as the gallstones were asymptomatic and there is no definitive data to support prophylactic cholecystectomy in patients with DJS. The intraoperative images could not be retrieved due to technical issues with the laparoscopic equipment; however, the image in Figure *1* is reproduced from the New England Journal of Medicine (NEJM) after copyright permission and is similar to the intraoperative findings in our patient [10].

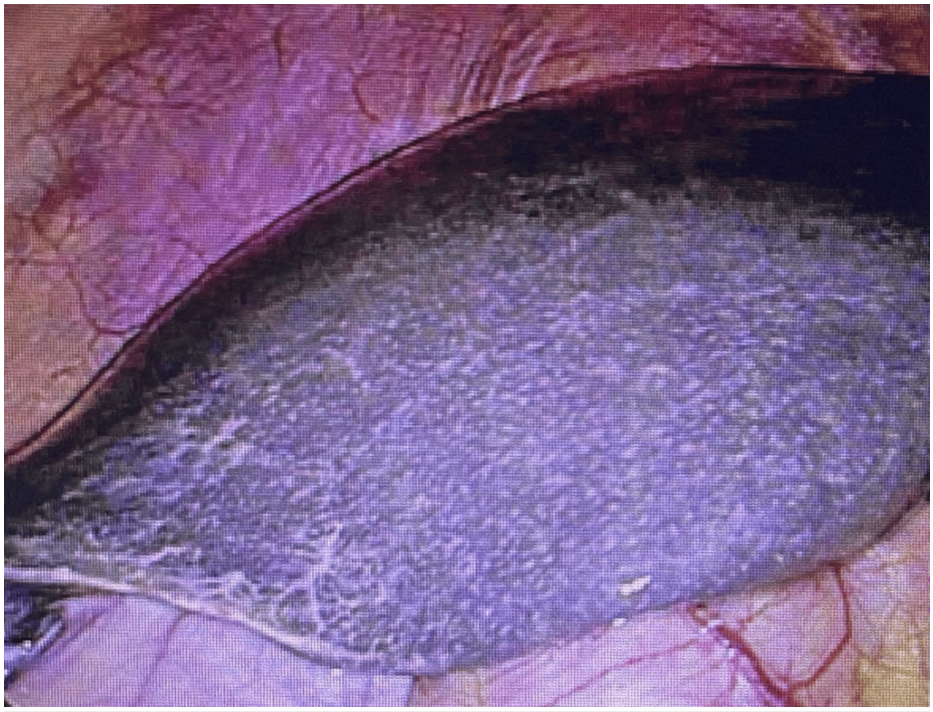


FIGURE 1: Image showing intraoperative diffusely black liver with smooth surface, similar with the present case

Reproduced with permission from the New England Journal of Medicine (NEJM): Morii K, Yamamoto T. Dubin-Johnson syndrome. N Engl J Med. 2016;375(1):e1 (doi:10.1056/NEJMicm1509529) [10]

Based on these findings, a presumptive clinical diagnosis of DJS was made. Remifentanil infusion and cis-atracurium IV were used for the laparoscopic appendectomy, and a steroid-bupivacaine mixture was deliberately withheld from peritoneal instillation at the end of the procedure to avoid adding an unnecessary hepatic drug burden.

The postoperative course was uneventful. The conjugated bilirubin remained elevated at postoperative day 21, which argues against appendicitis-related jaundice. Further family history revealed that several members of the family had intermittent scleral icterus, and there was a history of consanguineous marriage in the parents. The patient's younger brother also had similar laboratory findings of isolated conjugated hyperbilirubinemia. There was no history of pruritus in the family.

A probable diagnosis of DJS was made based on clinical signs, laboratory results, and intraoperative appearance of the liver. Liver biopsy and genetic tests were not conducted because they were not available and the condition was confirmed by family history. It may be safe to proceed with emergency surgery when black liver is seen intraoperatively, and liver biopsy may be deferred if the clinical and lab data suggest probable DJS.

Discussion

DJS is a rare hereditary disorder of bilirubin metabolism in which the liver continues to function normally but has a defect in the metabolism of conjugated bilirubin [1]. Impaired canalicular transport of conjugated bilirubin, caused by defects in the ABCC2 gene, leads to accumulation in hepatocytes [2,11].

A characteristic feature of DJS is the presence of a darkly pigmented liver, sometimes appearing black or blue, during surgery [7,8]. Deposition of melanin-like substances in hepatocytes results in this pigmentation, but not in structural liver damage or dysfunction. Although the liver looks different, it is structurally normal, and the clinical course is benign [3,4].

Laboratory data may demonstrate only isolated conjugated hyperbilirubinemia with normal transaminases, and patients with DJS are generally asymptomatic or have mild intermittent jaundice without pruritus [3,4]. These characteristics aid in the differential diagnosis of obstruction and hepatocellular causes of jaundice, but can be mistaken, particularly in acute surgical presentations [5]. We propose the following surgical and perioperative pathway for patients with DJS, as shown in Figure *2*.

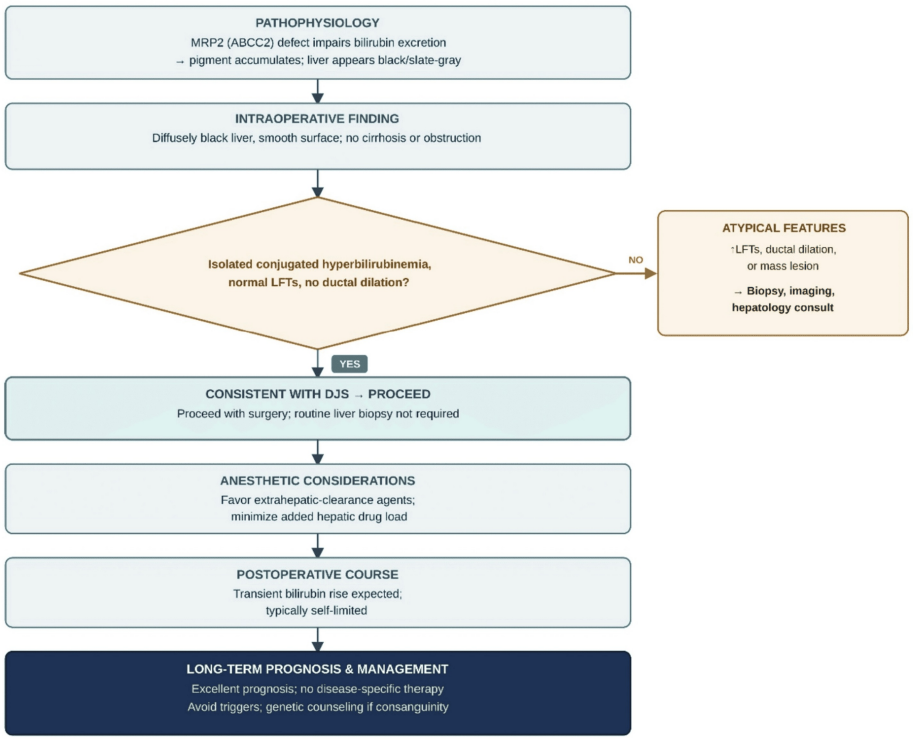


FIGURE 2: Proposed surgical and perioperative pathway for patients with Dubin-Johnson syndrome

In this case, the presence of acute appendicitis and jaundice suggested hepatobiliary disease at first. The diagnosis of DJS was supported by the presence of isolated conjugated hyperbilirubinemia, absence of biliary obstruction in conjunction with negative viral markers, and characteristic intraoperative finding of diffusely black liver.

The same findings were reported intraoperatively by Cebrián García et al. and Viñuela et al., when DJS was found as an incidental laparoscopic finding [5,7]. Its presence, however, in the context of acute appendicitis is still rare and thus significant. Miyakawa et al. described the anesthetic and postoperative course of a patient with DJS undergoing major esophageal surgery under fentanyl and nitrous oxide anesthesia; conjugated bilirubin rose markedly from the second postoperative day onward and ultimately required four days of plasmapheresis before resolving [12]. This underscores the importance of utilizing anesthetic agents with extrahepatic clearance in patients with suspected DJS. A transient rise in conjugated bilirubin following surgical stress is a well-recognized phenomenon, independent of the specific procedure performed. Gallstones have been reported previously in DJS although the exact etiology is unclear [13]. We did not perform cholecystectomy as there is no definitive evidence to support prophylactic cholecystectomy in asymptomatic cholelithiasis in patients with DJS.

There is no specific treatment for DJS, and the management is aimed mainly at reassuring the patient and avoiding unnecessary diagnostic and surgical procedures [1,4]. This case illustrates how critical it is to recognize the intraoperative finding of a black liver as probable DJS during surgery, especially in an emergency situation, in order to avoid additional procedures such as a bile duct exploration or liver biopsy.

Conclusions

Patients with isolated conjugated hyperbilirubinemia and incidental black liver should be evaluated for DJS. It may be safe to proceed with emergency surgery when black liver is seen intraoperatively, and liver biopsy may be deferred if the clinical and lab data suggest probable DJS.

Recognizing this benign condition is crucial to avoid misdiagnosis, unnecessary interventions, and to provide appropriate patient reassurance.

Additional Information

Author Contributions

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the

work.

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Disclosures

Human subjects: Informed consent for treatment and open access publication was obtained or waived by all participants in this study. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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