Abdominal pain and jaundice in patients with sickle cell disease: Clinical guideline

(See also S11.2, Abdominal sequestration guidelines: Link here)

Introduction

There are currently no randomised trials to inform the management of hepatobiliary complications in patients with sickling disorders.

Pigment gallstones due to ongoing haemolysis are common in sickle cell disease, occurring in at least 30% of children. They are often asymptomatic but can precipitate painful abdominal crises and the girdle syndrome. They can also cause:

- Acute cholecystitis (<10% children and adults with SCD, despite high frequency of gallstones)
- Chronic cholecystitis
- Biliary colic
- Obstruction of the common bile duct
- Acute pancreatitis

Investigations

- Liver function tests including ALT, Bilirubin, Alkaline phosphatase, Albumin,
- Serum Amylase
- Plain abdominal X-ray (as many as 50% of stones may be radio-opaque)
- Abdominal ultrasound

Further abdominal imaging may be required where the cause of pain remains unclear.

Differential diagnosis of upper abdominal pain in patients with sickling disorders

- Biliary colic; Cholecystitis
- Hepatitis (viral)
- Acute intrahepatic cholestasis (acute tender hepatomegaly, with features of obstructive jaundice)
- Peptic ulcer
- Vaso-occlusive episodes
- Hepatic sequestration (enlargement of liver without other explanation, and drop in Hb >20 g/L)
- Splenic sequestration
- Chest syndrome

Management of acute cholecystitis

The mainstay of management involves:

- Analgesia
- Intravenous hydration and correction of electrolyte imbalance
- Antibiotics. Please consult local antibiotic guidelines

Referral for surgery

- For patients with recurrent cholecystitis, referral for consideration of cholecystectomy is advised.
- Where asymptomatic choledocholithiasis is identified, referral for cholecystectomy and removal of bile duct stones should be considered after a single episode, since this may be complicated by acute cholangitis, or biliary pancreatitis.
- For acute painful choledocholithiasis, request inpatient surgical opinion for ERCP or emergency surgery.

Preparation for surgery

While transfusion is not typically required for patients undergoing routine laparoscopic cholecystectomy, there is always the risk of conversion to an open laparotomy. Patients should be prepared by top-up transfusion where possible (ensure final Hb does not exceed 110 g/L), or exchange where top-up is not feasible due to high a baseline haemoglobin or at high risk of SCD related complications.
Peri-operative transfusion guidelines:http://nssg.oxford-haematology.org.uk/red-cell/documents/transfusion-and-exchange/S23-2-perioperative-transfusion-in-scd.pdf

Surgical teams should be provided with a copy of the current network perioperative guidelines for patients with SCD, which includes advice on transfusion and anaesthetic risks in patients. Ensure early input from anaesthetists, especially in relation to analgesia management.http://nssg.oxford-haematology.org.uk/red-cell/documents/acute-management-sickle-cell-disease/S23-1-perioperative-care-of-patients-with-scd.pdf

Intrahepatic cholestasis

Some patients experience episodes of severe hyperbilirubinaemia (conjugated + unconjugated) with moderately raised alkaline phosphatase, associated with fever and hepatic pain in the absence of demonstrable stones. These episodes may be due to severe intrahepatic sickling.

- Analgesia (with care as most opiates are metabolised in the liver)
- Hydration
- Antibiotics (see local guidelines for recommendation)
- Monitor liver function tests, and as for girdle syndrome/hepatic sequestration
- Hyperhaemolysis +/- sequestration may supervene, requiring frequent transfusion
- Early consideration of exchange transfusion is recommended.

Thames Valley SHT Specialist Referrals

Mr Zahir Soonawalla, Consultant Hepato-biliary Surgeon
Nuffield Department of surgery, Churchill Hospital, Old Road, Headington, Oxford OX3 7LE, , Zahir.Soonawalla@ouh.nhs.uk

Southampton SHT Specialist referrals

Gastro-intestinal surgery team.

References

Expert panel report on the management of Sickle Cell Disease, NHLBI, Bethesda, Maryland, USA, 2014

Standards for the Clinical Care of Adults with Sickle Cell Disease in the UK © Sickle Cell Society 2018

Authors:
Dr Wale Atoyebi, Clinical Lead for Haemoglobinopathies

Review

Name	Revision	Date	Version	Review date
Wale Atoyebi	Pre-peer review	Jan 2013	1.0	Jan 2015
Deborah Hay	Routine review	Aug 2015	1.2	Jan 2017
Deborah Hay	Routine review	Jan 2019	1.3	Jan 2020
Noemi Roy	HCC, plus update	Apr 2021	2.0	Apr 2023
Wale Atoyebi	Full review	May 2024	3.0	May 2026