Abdominal sequestration and girdle syndrome in sickle cell disease: Clinical guideline (HN-506 –d2)

Splenic Sequestration
Although splenic sequestration is more common children it can also occur in adults. It may be precipitated by fever, dehydration or hypoxia. Rapid sequestration of red cells can lead to sudden anaemia and even death from hypoxic cardiac failure with pulmonary oedema. In some patients it may have a more insidious onset and can be recurrent.

Symptoms
• Abdominal pain (pulling legs up to abdomen)
• Abdominal distension
• In some cases, sudden collapse

Signs
• Rapidly enlarging spleen (may or may not be painful)
• Pallor, shock (tachycardia, hypotension, tachypnoea)
• +/- Fever due to associated sepsis

Investigations
• FBC and retics (raised in sequestration, absent in aplastic crisis)
• Blood cultures and other infection screen, as clinically indicated
• Parvovirus B19 serology (differential diagnosis is aplastic crisis)
• Cross-match half the patient’s estimated blood volume immediately
• Ensure blood is subject to extended phenotyping but do not let this delay the supply of blood

Management: immediate
• Resuscitation with fluids.
• Emergency top-up transfusion, if necessary with O Rh negative (‘flying squad’) blood
• Broad-spectrum antibiotics

Management: following episode
• Consider a hypertransfusion regime for 2-3 months
• Consider splenectomy if recurrent (> 1 episode)

Hepatic Sequestration
Although less widely documented, a similar process of acute hepatic sequestration may be seen, associated with a rapid reduction in haemoglobin and an appropriate reticulocytosis.

Symptoms
• Right hypochondrial pain, abdominal distension
• +/- Fever due to associated sepsis
Signs
• Enlarging tender liver, increasing jaundice
• Collapse/shock is less common than with splenic sequestration

Investigations
• Bilirubin may be very high; remaining liver function tests may not be altered
• Exclude gallstones/cholestasis by ultrasound
• Blood cultures and other infection screen, as clinically indicated

Management
• May need urgent top-up transfusion.
• IV antibiotics
• If the patient develops tachypnoea, or develops chest signs, then check ABG and treat for acute chest syndrome.

Girdle (or mesenteric) syndrome
May be said to be present when there is an established ileus, with vomiting, a silent distended abdomen, and distended bowel loops with fluid levels on abdominal x-ray. Some hepatic enlargement is common, and it is often associated with bilateral basal lung consolidation (early acute chest syndrome).

Differential diagnosis is wide and includes many intra-abdominal surgical emergencies (e.g. splenic and hepatic sequestration, acute appendicitis, pancreatitis, cholecystitis, biliary colic, splenic abscess, ischaemic colitis, peptic ulcer etc).

Well-localised or rebound tenderness, board-like rigidity or lack of movement on respiration is suggestive of this diagnosis. Ultrasound may be helpful. If surgical intervention is contemplated, exchange transfusion should be performed prior to laparotomy: this can be started pending clarification of the diagnosis.

Investigations
• Full blood count
• Renal and liver biochemistry
• Serum amylase to exclude pancreatitis
• Erect chest x-ray (a CXR may need to be repeated every 1-2 days)
• Oxygen saturation
• Abdominal ultrasound - as indicated
• Serum amylase to exclude pancreatitis.
• Group and crossmatch for likely exchange transfusion

Management
• Intravenous rehydration
• Analgesia
• Antibiotics as per local microbiological guidelines
• Request urgent surgical opinion
• Exchange red cell transfusion is indicated for the girdle syndrome and for acute surgical intervention for its differentials. Exchange is best arranged early in this
Adult Haemoglobinopathy Service


- With signs of ileus/obstruction, keep patient fasted, and consider nasogastric aspiration if vomiting

**Monitoring**
- Document abdominal girth (at umbilicus) 1-4 hourly
- Document liver size at least twice daily
- Document chest examination at least twice daily
- Monitor SaO2 in air and O₂, HR, RR, BP 1-4 hourly
- Measure capillary blood gases if SaO2 in air <95%, or 3% below well baseline, as per guidelines for acute chest syndrome

**References:**
Draft Standards for the Clinical Care of Adults with Sickle Cell Disease in the UK © Sickle Cell Society 2017


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