

Introduction

These guidelines were constructed after a joint audit and review of practice at the three specialised paediatric hepatology centres in June 2016, and wider consultation with BSPGHAN Council, Liver Steering Group and Endoscopy Working Group. Although evidence based where possible, where not available they represent consensus. A detailed version of this guideline which includes review of evidence, together with reference to adult evidenced based guidelines is available. Section 4: management of acute variceal bleed: is designed for implementation at any hospital to which a child presents and can be used as a stand-alone guideline.

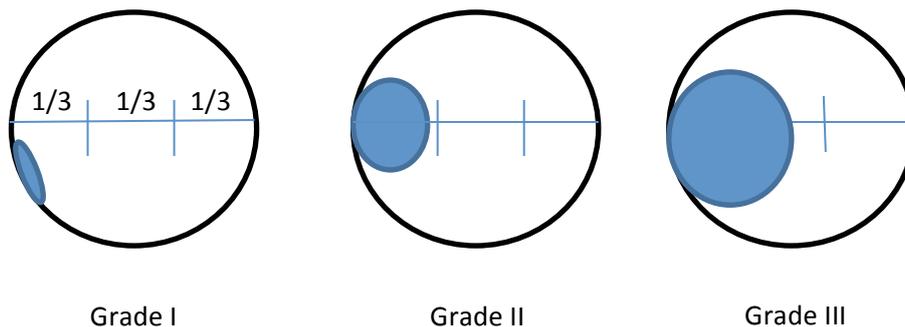
1. Grading of oesophageal varices

Grade I: these collapse on inflation of oesophagus with air

Grade II: these are varices between grades 1 and 3 ($< 1/3$ of diameter of lumen)

Grade III: these are large enough to occlude the lumen ($> 1/3$ of diameter of lumen)

Supplementary descriptive terms e.g., with stigmata, red spots, red wheals, vessels on vessels etc.



2. Treatment categories and definitions

- Primary prophylaxis of oesophageal varices:
 - treatment of oesophageal varices before bleeding has occurred
- Management of acute oesophageal variceal haemorrhage:
 - acute time frame interval is within 5 days from onset of initial bleed
- Management of oesophageal variceal re-bleed:
 - re-bleed is defined as bleeding after interval of more than 5 days since first bleed
- Secondary Prophylaxis of oesophageal variceal haemorrhage:
 - treatment of varices that have previously bled, but not during acute bleeding episode
- Primary prophylaxis of gastric variceal bleeding:
 - treatment of gastric varices before bleeding has occurred
- Management of active haemorrhage from gastric varices

3. Surveillance endoscopy and primary prophylaxis of oesophageal varices

3.1. Patient selection

- Primary prophylaxis / surveillance endoscopy NOT offered to children who could not be treated by band ligation due to their size (i.e. prophylactic sclerotherapy not performed)
- Children should be offered surveillance endoscopy if evidence of portal hypertension, irrespective of age, with splenomegaly and hypersplenism (platelets $<120,000 \times 10^9/L$ on two consecutive samples)

3.2. Perioperative management

- **3.2.1. Pre-theatre assessment:** Full blood count, coagulation, group and save
- **3.2.2. Threshold for blood products:** If platelets less than $50,000 \times 10^9/L$ have platelets available for infusion in theatre if intervention is needed
- **3.2.3. Which varices to treat prophylactically by band ligation:**

Prophylaxis with band ligation but *not* non-selective beta blockade is recommended

Grade 1 varices: no treatment

Grade 2 varices: consider band ligation of any grade 2 varix: treatment recommended if multiple varices or grade 2 with stigmata

Grade 3 varices: treatment by band ligation

- **3.2.4. Post endoscopy feeding and discharge**

If no treatment of varices needed: same day discharge

If treated by band ligation:

- Offer liquids initially and then if tolerated soft diet on same day as procedure.
- Discharge 6 hours post procedure can be considered unless:
 - First endoscopy requiring intervention
 - Procedure complicated by bleeding or airway complications
 - Platelets needed as $< 50,000 \times 10^9/L$ and band ligation performed
 - Journey time $>$ one hour by car: consider overnight stay locally

- **3.2.5. Medication post prophylactic band ligation**

Insufficient evidence to support use of sucralfate post band ligation, but sucralfate should be prescribed for 48 hours for all treated by sclerotherapy. Omeprazole: prescribe for two weeks, continued longer according to clinical need.

- **3.2.6. Follow up: interval to next endoscopy**

Clinical judgement should be used to determine interval, which may be influenced by underlying diagnosis, co-morbidity, severity of varices, and risk of repeated GA. The following is for guidance:

- Grade 3 and grade 2 varices (treated): repeat after 1-3 months, then 3-6 monthly if stable or improved. If no banding required at follow up endoscopy repeat after 12 months
- Grade 2 varices (not treated): repeat after 6 months, then yearly if stable and no treatment needed
- Grade 1: repeat 12 monthly, subsequently may reduce frequency to 24 monthly
- No varices: repeat after 2- 3 years

Section 4. Management of active variceal bleeding

Any child presenting with acute gastrointestinal bleed, whether haematemesis or melaena, should be admitted to the nearest hospital for stabilisation and initial management. Transfer to a centre that can offer appropriate diagnostic and therapeutic endoscopy should be arranged at the earliest opportunity. Variceal bleeding may be the first presentation of a child with portal hypertension, which may be secondary to underlying liver disease or a portal vein thrombosis without liver disease. Other causes of acute GI bleeding should be considered, and include battery ingestion, duodenal ulceration and Meckel's diverticulum (melaena only). This guidance is appropriate for a child in whom acute GI bleeding may be due to varices, with clinical suspicion of portal hypertension even if unproven. The protocol should be shared with referring hospitals so that management is instituted without delay. This guidance reflects the consensus management of the three National Paediatric Liver centres in Birmingham, Leeds and London (King's) with revision and endorsement from BSPGHAN during consultation process.

AIRWAY, BREATHING & CIRCULATION (ABC)

- ABC patient assessment
 - Airway: Ensure airway is patent
 - Breathing: Visually assess patient for breathing efficacy and effort. Obtain respiratory rate and oxygen saturation. Administer oxygen via a face mask if required
 - Circulation: Obtain heart rate, manual pulse, capillary refill and blood pressure. Start continuous monitoring of heart rate and oxygen saturation

FLUID RESUSCITATION and INITIAL MANAGEMENT

- Insert two wide bore peripheral intravenous cannulas and commence intravenous fluids: either crystalloid or colloid as clinically indicated and fluid bolus if required. Ensure sample for full blood count, coagulation and cross match is obtained ASAP.
- Aim initially to transfuse to haemoglobin level of 90g/L: commence transfusion slowly to reduce risk of increasing portal pressure and re-bleeding. Do not over-transfuse.
- If no blood group matched blood is available, use O Negative blood.
- Give platelets, Fresh Frozen Plasma (FFP), and cryoprecipitate where indicated (platelets $<100 \times 10^9/L$ or INR >1.5 and bleeding not controlled).

- Commence patient on 2/3 maintenance intravenous (IV) fluids
- Keep child nil by mouth
- Monitor blood sugars 2-4 hourly. Aim for a blood sugar of 4-8mmols/L. Consider changing to 10% glucose concentration in IV fluids if patient is hypoglycaemic
- Correct any electrolyte or pH abnormalities
- Strict fluid balance (monitor input and output)

INVESTIGATIONS

- Obtain blood gas (processed urgently) and blood sugar
- Bloods: FBC, U&Es, INR, renal, liver and bone profiles and blood cultures. Consider sending an ammonia sample (stored on ice) if encephalopathy suspected

PHARMACOLOGICAL THERAPY

OCTREOTIDE INFUSION

- Commence in children with possible varices as cause of gastrointestinal bleed.
- Use dedicated IV cannula

Octreotide dose:

Stat dose: 1microgram/kg IV over 5 minutes (maximum 50 microgram) followed by Infusion at 1-3microgram/kg/hour (max 50microgram/hour)

To prepare: dilute 500micrograms to 40ml with 0.9% sodium chloride (2ml/hour = 25microgram/hour)
Continue Octreotide Infusion until 24 hrs after bleeding is controlled, then wean slowly over 24 hours to reduce risk of rebound bleeding.

Octreotide has a short half- life, therefore re-site cannula immediately if drip tissues

OTHER DRUGS

- Intravenous antibiotics (co-amoxiclav or a cephalosporin or piperacillin / tazobactam depending on local guideline, however should include gram positive and negative cover and avoid local quinolone resistance).
- Intravenous omeprazole (1mg/kg od max 40mg OD) **or** pantoprazole (child <12years: 500mcgs/kg; max 20mg OD. Child >12years: 40mgs OD) **or** esomeprazole (0-1 month: 0.5mg/kg OD; 1-11 months: 1mg/kg OD; 1-11 years and <20kg: 10mg OD; 1-11 years and > 20kg 10-20mg OD; 12 years and above: 40mg OD.

If a proton pump inhibitor is not available, give
Intravenous ranitidine (1mg/kg TDS max 50mg TDS)

- Vitamin K (phytomenadione): 300microgram/kg as slow IV injection (max 10mg)

ADDITIONAL MANAGEMENT**NASOGASTRIC TUBE**

If a nasogastric tube in in situ, it may be used to aspirate and be put on free drainage. Do not insert a new nasogastric due to risk of further bleeding.

SENGSTAKEN TUBE

If bleeding continues and the patient is not responsive to all of the above management, consider placing a Sengstaken-Blakemore Tube. This however is RARELY needed. Patient must be intubated and transferred to intensive care setting prior to Sengstaken-Blakemore insertion. This should only be passed on an intubated child when rapid blood loss continues in spite of medical management. Control of the airway and volume replacement are essential. Once the tube has been passed well into the stomach, the gastric balloon is inflated initially with 40mls saline and 10mls contrast (available via radiographer) and maintained on skin traction.

DO NOT INFLATE OESOPHAGEAL BALLOON

Leave gastric and oesophageal lumen on free drainage. Portable check x-ray required immediately to assess the size and position of gastric balloon. Regular release of traction is essential to avoid skin injury from continuous pressure. The balloon is deflated at time of endoscopy.

| Weight of Child | SENGSTAKEN TUBE Size |
|-----------------|----------------------|
| 10-30 kg | paediatric size 14F |
| > 30 kg | paediatric size 16F |

For infants < 10kg in whom use of a Sengstaken tube is not possible because of size, a Foley catheter inserted orally may be effective. This however is NOT a licensed use of Foley catheter.

FURTHER ADVICE

On going management, including transfer, should be discussed with a centre that can offer endoscopic treatment at the earliest opportunity. Local arrangements and pathways should be in place, and may include referral to colleagues in paediatric surgery depending on local practice, to tertiary paediatric gastroenterology centre or to specialist liver service.

The following are the contact details of specialist hepatology centres for children: contact details of your referral centre could be inserted into a local version of this guideline.

BIRMINGHAM

The Liver Unit, Birmingham Children's Hospital

Consultant Paediatric Hepatologist: switchboard 0121 333 9999

Registrar: via switchboard 0121 333 9999 and request registrar phone or bleep 55200

Nursing team: Liver Direct 0121 333 8989 or email Liver.Direct@bch.nhs.uk

Ward: Liver Unit Ward 8: 0121 333 9066

Office Fax: 0121 333 8251

LEEDS

Children's Liver Unit, Leeds Children's Hospital.

Consultant Paediatric Hepatologist: switchboard 0113 2432799

Registrar: via switchboard or ward and request paediatric hepatology registrar (9am-5pm weekdays) OR Paediatric Specialty Registrar On Call (5pm-9am weekdays and weekends).

Clinical Nurse Specialist Team: 0113 3926151 / 3926138

Ward 50: tel 0113 3927450

Fax 0113 3925129 (Admin Office) or 0113 3923110 (Ward Doctors Office)

KING'S, LONDON

Paediatric Liver, Gastroenterology and Nutrition Centre, King's College Hospital, London

Phone 020 3299 9000

Fax 0202 3299 4228

Bleep 426 weekdays 9am-5pm

Phone 07866792368 (5pm-9am)

5. Secondary prophylaxis

Perioperative management is as for primary prophylaxis. There is currently insufficient evidence to recommend use of non-selective beta blockers in primary or secondary prophylaxis in children, unlike in adults. However, it is acknowledged that practice may vary.

6. Gastric varices

- **6.1. Prophylaxis**

No indication for endoscopic treatment as primary prophylaxis

- **6.2. Treatment of active bleeding**

Treatment of bleeding gastric varices include histoacryl glue injection. Haemospray may have a role in treating erosive gastritis. Use depends on local expertise and ongoing review of evidence.