ADVANCED LIVER DISEASE AND PORTAL HYPERTENSION IN CYSTIC FIBROSIS

Patient Details:

This patient has cystic fibrosis with established liver disease and portal hypertension and is at risk of acute variceal gastro-intestinal haemorrhage.

General advice:

- The patient should have a Medi-Alert bracelet/necklace.
- The patient's blood group is:_____
- Aspirin and non-steroidal anti-inflammatory pain killers (eg Ibuprofen) must be avoided.
- Abstain from contact sports.
- The onset of haemorrhage may be associated with abdominal pain, vomiting blood, increased activity and movements of the bowels and possibly the passage of tarry, black, loose stools (melaena). Large bleeds may cause faintness, sweating, pallor and a high pulse rate.
- If any worrying symptoms arise, urgent transfer to the nearest Emergency Hospital Unit should be arranged.

Evaluation and management:

- Evaluate the patients condition (ABC). Administer high flow oxygen.
- Secure venous access and take blood for FBC, Coagulation, U&E, liver function, Glucose and Blood culture, Cross match for blood, FFP and cryoprecipitate.
- Monitor ECG, T, P, R, SaO2, BP and neuro-observations if encephalopathy suspected.
- Consider central line placement for CVP monitoring.
- If there are signs of hypovolaemia, resuscitate at once with blood. Normal saline 0.9% may be used until blood available. Use Group O Rh negative if necessary.
- Place a nasogastric tube (large bore – not silk). This should be aspirated at frequent intervals to identify persistent/recurrent bleeding. It can

provide early warning of re-bleeding and reduces the likelihood of vomiting.

· Place urinary catheter and monitor urine output.

· Correct any coagulation defect and arrange for platelet infusion if the patient has functional hypersplenism (platelets <50).

Drugs:

· Ranitidine 1-3 mg/kg/dose 8 hourly IV (max 50mg TDS)

OR

· Omeprazole 500mcg/kg intravenously daily (max 40mg BD)

AND

· Octreotide 3-5 mcg/kg/hour. Max dilution 25mcg/ml. Stable at room temperature for 48 hours. 1250mcg in 50mls saline 0.9% and run at Wt x 0.12mls/hr = 3mcg/kg/hr. Increase if bleeding is not controlled.

Ongoing bleeding consider:

· Terlipressin as IV bolus 500mcg for <7yrs, 1mg for 7-12yrs, 2mg >12yrs, doses can be repeated 4 hourly if bleeding continues. An alternative is Argipressin 0.3units/kg as an IV infusion over 30 minutes followed by an infusion at 0.3 units/kg/hour (max 1 unit/kg/hour). These drugs can be associated with allergic reactions and can increase gastro-intestinal symptoms including nausea, vomiting, abdominal pain, flatulence and so forth. Argipressin can cause a rise in blood pressure and ECG monitoring should be continued. Liver function would need to be monitored carefully.

· Endoscopy and sclerotherapy Consider only when patient is stable and discuss with liver unit/gastroenterologists.

· Sengstaken-Blakemore Tube. Only placed under controlled conditions eg following sclerotherapy when the patient is under general anaesthetic or in the intensive care unit. The patient would need to remain intubated and anaesthetized to protect the airway.

Associated liver failure:

Identify and treat predisposing causes including bleeding, infection, electrolyte disturbances (hypocalaemia and hyponatraemia). Administer oral Lactulose regularly to operate as a laxative and reduce the production of ammonia in the colon. Oral Neomycin may be used if Lactulose treatment fails. Ensure there is adequate carbohydrate administration (intravenously) to avoid the development of a catabolic state. Monitor blood glucose, ammonia, renal function and acid-base status.

DISCUSS THE PATIENTS MANAGEMENT WITH TERTIARY LIVER TEAM.

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