

## CF Guidelines - Haemoptysis

### Haemoptysis and CF:

Streaky haemoptysis is common with chronic infection but may indicate an exacerbation of respiratory symptoms so sputum should be cultured and a course of antibiotics considered. Haemoptysis must be differentiated from haemetemesis. The source of haemoptysis is usually from areas of chronic airway inflammation. The volume of haemoptysis is always difficult to assess but any volume of about 250ml – 300ml per 24hrs (approximately 1 cupful) is regarded as potentially life threatening. Some series record in-hospital mortality of up to 80% among adult patients with > 500ml over 24 hrs. The rate of bleeding is more important than the total volume over 24hrs. The cause of death from haemoptysis is usually asphyxiation rather than volume loss and only 30ml - 50ml may from a fatal clot in the trachea. Any patient with brisk haemoptysis or a volume of about 1 cupful over 24hrs should be admitted. Most significant haemoptysis comes from bronchial arteries which arise from the bronchial arterial circulation derived from the aorta at systemic pressure. In CF the bronchial arteries can be significantly hypertrophied and may be suitable for embolisation. The patient may sometimes experience a gurgling sensation which can be a localising symptom indicating the bleeding site. The patient is likely to be very scared - reassurance is essential and sedation may be necessary. Primary management is resuscitation if needed (incredibly rare) - lay patient on side (gurgling side down), give oxygen. There is no evidence to suggest that stopping Dornase Alfa is necessary. Physiotherapy may have to be adapted - seek advice from the physiotherapist. In CF haemoptysis, remember the possibility of pulmonary embolism and this may have arisen from a port-a-cath.

### Initial investigations:

- Hb & Platelets.
- Coagulation.
- Group and save or cross-match blood,
- Sputum culture
- Chest X-ray can show new

For adults, a CT Bronchial arteriogram is the investigation of choice to show whether there are any hypertrophied bronchial arteries suitable for embolisation and should be arranged as soon as possible after admission with massive haemoptysis. This involves liaison with a pulmonary radiologist to ensure that the timing of scanning is coordinated with maximal contrast density in the aorta (rather than the pulmonary artery as for a CTPA). If hypertrophied bronchial arteries are present (> 2mm in adults) discuss with interventional radiologists to decide whether transfer to centre with facilities for embolisation is appropriate – Exeter and Plymouth both have experience. If bronchial arteries are not present then management is predominantly medical rather than interventional.

### Initial management:

In massive haemoptysis, patients are usually, understandably, extremely anxious. This requires reassurance and may need cautious sedation. Consider Benzodiazepam and/or opiates. As significant bleeding is usually at systemic pressure consider hypotensive agents in adults if significantly hypertensive. Give blood and correct coagulation defects if necessary (IV vitamin K/FF/Cryoprecipitate). Start intravenous antibiotics. Infection often triggers haemoptysis and blood in the bronchial tree frequently becomes infected after a short period of time. Review records and record most recent FEV1 when well – this may determine whether surgical options are

feasible. Inform thoracic surgeons and ITU of patient's admission. Continue with gentle regular physiotherapy, but omit chest clapping for 24hrs.

#### Further management:

- Mild (< 100ml per day)/Moderate (> 2 episodes of 100ml/day per week):
  - Oral Tranexamic Acid - 15mg/kg/td - 25mg/kg/td - max 1gm/dose - for 5 days.
  - IV Tranexamic Acid - 10mg/kg/td.
  
- Massive (> 300ml/24hrs) or if repeated bleeding occurs over a short period - daily for 7 days with > 100ml on 3/7, consider the following:
  - Tranexamic Acid as above, plus IV Terlipressin. 2mg then 1mg - 2mg every 4-6 hours until bleeding is controlled, (maximum duration 72 hours). Not to be used in pregnancy. Fewer side effects than Vasopressin.
  - Tranexamic Acid as above, plus IV Vasopressin (Argipressin) is occasionally useful - the paediatric dose is 0.3 units/kg over 20 minutes followed by 0.3 units/kg/hour. The adult dose is 20 units in 100ml 5% dextrose given over 15 mins followed by an infusion of 0.2 units/min for 36 hours. It can lead to water intoxication and can cause bronchoconstriction.
  - Tranexamic Acid as above, plus Octreotide. 50mcg IV bolus units over 15 minutes intravenously.
  - Tranexamic Acid as above. Then Octreotide infusion : dilute 400mcg in 20mls of 5% dextrose and infuse at 2.5ml per hour through a syringe driver. Can be increased to 5ml per hour if necessary and can be continued for up to 2 days. (The infused drug is only stable for 8 hours).
  - Nurse upright and give an ice cold drink (vasoconstricts bronchial arteries).
  - Humidified Oxygen

#### Further investigations:

- Bronchoscopy is rarely useful in the acutely bleeding patient. If you are considering this procedure initially try flexible, then consider a rigid, under general anaesthetic. With massive haemoptysis, go straight to rigid bronchoscopy. This can be technically very difficult but may allow clot removal (beware precipitating further bleeding), tamponade of bleeding site using a Fogarty catheter, or haemostasis with thrombin glue or iced saline lavage/ vasoconstrictor lavage.
- Selective bronchial angiography and embolisation can only be carried out by experienced specialists, currently in Exeter and Plymouth. Numerous dilated tortuous bronchial arteries are often identified some of which may take origin from aberrant sources. Actual source of bleeding is difficult to discern but generally a number of large vessels (>2.5mm) are embolised using variable sized gel foam pledgets. Great care to avoid spinal artery (with consequent paraplegia) and other systemic artery embolisation is necessary. Post embolisation pain requiring narcotic analgesia and transient dysphagia may occur. This is not a cure and many patients develop new vessels within months or years that may bleed and so require further embolisation.
- Lobectomy may be considered as a last resort. Thoracic surgeons will need as much information as possible on previous lung function and performance status. A bronchoscopy to confirm the lobe from which blood is originating is essential prior to surgery.

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