LRI Children's Hospital

Cystic Fibrosis Emergencies UHL Children's Medical Guidelines

Staff relevant to:	Medical & Nursing staff working within the UHL Children's Hospital.
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Written by: Reviewed by:	Erol Gaillard Imad Ahmed Vandana Pankhania
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1. Introduction and who this guideline applies to

1. Introduction and Scope

This guideline is intended to be a guide for medical and nursing staff in the management of certain emergencies seen in children with cystic fibrosis. As always, the appropriate paediatric life support guideline should be used in any unwell child.

Related documents

- Basic Life Support or Choking UHL Children's Hospital Guideline UHL C2/2016
- Infection Prevention UHL Policy UHL B4/2005
- Chest Drain Management UHL Children's Hospital Guideline UHL C41/2016
- Chest Drain Insertion UHL Children's Intensive Care Guideline UHL C41/2016
- Blood Transfusion UHL Policy UHL B16/2003
- Cystic Fibrosis Paediatric Prescribing UHL Children's Hospital Guideline UHL C35/2016
- Cystic Fibrosis - Inpatient Chest Exacerbation UHL Children's Medical Guideline UHL C36/2016
- Upper Gastrointestinal Bleeding UHL Children's Guideline UHL D2/2019

2. Haemoptysis in Cystic Fibrosis Patients

Haemoptysis is a common complication of Cystic Fibrosis (CF). Episodes of streaking (minor bleeding) are common, especially in adolescents, but massive haemoptysis (rare) should be considered a life-threatening emergency.[1] Approximately 1% of children with CF experience massive haemoptysis, with subsequent significant deterioration in lung function over the year following the episode. Most occur in late teens, and it is extremely rare in the under 10-years age group.[2]

Infection has been demonstrated to be the most common precipitating factor, with 90% of patients in one case series having a concurrent pulmonary exacerbation at the time of first episode.[3]

Definitions –Typically classified as scant (blood streaked sputum < 5ml), mild (5ml - 240 ml in one day) or massive (> 240 ml in one day or 2 days of greater than 100 ml).[4]

Management – Current management guidelines are based on expert opinion as there are very few trials and no randomised controlled trials assessing haemoptysis treatment efficacy.[5]

2.1 General Principles

1. All patients with haemoptysis warrant admission to hospital. All patients with massive haemoptysis should be admitted to either HDU or CICU.
2. Perform chest radiograph although it is not usually useful in localising source of bleeding
3. If on NSAIDs – STOP
4. If portacath present, consider possibility of pulmonary embolism.

2.2 Scant haemoptysis

- Streaky blood-stained secretions are not uncommon and is usually associated with an infective exacerbation. Sputum should be collected for culture and NPA/nasal swab sent for virology and antibiotics started empirically.
- Continue current physiotherapy regime but discuss with the CF Physiotherapist at the earliest opportunity

2.3 Mild Haemoptysis

1. This is usually associated with an infective exacerbation. Sputum should be collected for culture and NPA/nasal swab sent for virology. Antibiotics started empirically.

- For patients who have not previously isolated Pseudomonas, start IV Cefuroxime.
- Patients with previous isolates of Pseudomonas aeruginosa should be empirically started on IV Ceftazidime and Tobramycin. Before prescribing, check sensitivities if available of previous isolates.

For doses, please refer to the Cystic Fibrosis Paediatric Prescribing UHL Childrens Hospital Guideline which includes links to the CF drug formularies.

2. Check if patient is on vitamin K (usually on a combination product e.g paravit CF) If not then start the patient on oral vitamin K.

3. Start oral tranexamic acid. This has been used long term in children with CF and recurrent haemoptysis with some success.[6]

4. Physiotherapy should not be stopped; but may need to be adapted. The CF Physiotherapist should review and create a plan. Airway clearance techniques should minimise increases in intrathoracic pressure.

- Avoid the use of positive pressure techniques (internal, external or oscillatory) for 24-48 hours post bleed. Consider airway clearance techniques such as Active cycle breathing or autogenic drainage which utilise controlled coughing
- Minimising vigorous or excessive coughing.
- Continuing all inhaled therapies but consider stopping on an individual basis if identified as potential trigger to uncontrolled coughing or further bleeding.
- Cease physical exercise for 24-48hrs post bleed
- Maintain physical activity with careful monitoring of physiological status
- For children on NIV, stop NIV or, where considered too great a risk, consider temporarily reducing inspiratory pressures until bleeding settles
- Graded approach to reintroduce airway clearance techniques if no further bleeding - preferable to wait at least 24 hours prior to starting positive pressure, adjuncts or manual techniques (then only one at a time).
- Avoid forced expiratory manoeuvres (e.g. Lung function tests) at least 48 hours after last haemoptysis.

2.4 Massive Haemoptysis

This is a life-threatening medical emergency as patients can rapidly exsanguinate. Other causes of death from haemoptysis are asphyxiation (drowning in their own blood), hypovolaemia or a combination of the above. Airway protection and adequate volume resuscitation are the focus of emergency medical therapy.

1. Seek urgent help and resuscitate as per APLS principles i.e. Airway, Breathing, and Circulation

- Give oxygen, Optimise oxygenation and humidification
- If localised “gurgling” present on auscultation, lay patient gurgling side down
- 2 large bore IV cannulas and take bloods for FBC, Coagulation screen (including fibrinogen), Group and save/ cross match.

2. Give IV Vitamin K in all cases. If necessary, give blood and correct coagulation defects (FFP/Cryoprecipitate)

- Vitamin K (Phytomenadione) 250-300micrograms/kg max 10mg as single dose

3. Start IV Tranexamic acid

- IV Tranexamic acid- interval dosing or continuous infusion (Please see BNFc for doses.)

4. Start IV antibiotics with high dose Staphylococcus aureus + antipseudomonal cover (Tobramycin with either Meropenem OR Tazocin) . Patients colonised with MRSA should be discussed with microbiology, for advice on an appropriate antimicrobial agent.

5. Physiotherapy is stopped during resuscitation phase. Obtain advice of physiotherapist regarding restarting chest physiotherapy.

- Careful positioning (high side lying, bleeding side down).
- Cease all physiotherapy treatments and exercise. Avoid forced expiratory manoeuvres. Percussion is not advised and should be omitted for at least 48 hours
- In selected patients who need bronchial artery embolisation, in liaison with interventional radiologist, resume normal airway clearance and a graduated exercise programme following the procedure.
- Ensure adequate analgesia if chest pain limits effective airway clearance
- Psychological support and reassurance in resuming normal activity and treatments may be required.

Further Management Options

Most bleeds will stop with the above measures, but if massive bleeding persists, or if repeated bleeding occurs daily for 7 days with >100mls on 3 or more days:

- Admit to HDU or CICU
- IV Vasopressin / Terlipressin

○ In children < 12 years of age, use IV vasopressin at 0.3 units/kg (maximum 20 units) over 20 minutes followed by 0.3 units/kg/hour (maximum 1 unit/kg/hour) continued for 12 hours after bleeding has stopped and gradually wean over 24-48 hours (maximum duration 72 hours). It can lead to water intoxication and can cause bronchoconstriction.

○ For children >12 years, use IV Terlipressin (has fewer side effects); Dose is brand specific, please refer to BNFc for this. Variquel® injection and Glypressin® injection are both stocked in UHL. Variquel® can be used for a maximum of 72 hours and Glypressin® can be used for a maximum of 48 hours.

- Consider CT Bronchial arteriogram to show whether there are any hypertrophied bronchial arteries suitable for embolization.
- Selective bronchial angiography and bronchial artery embolisation (BAE) – This will need a referral to interventional radiologists (Please discuss with the Leicester

Radiology team and if unavailable may have to contact Birmingham Children's hospital (Dr Ian McCafferty/ Dr Simon McGuirk)

- Bronchoscopy is not generally very useful with haemoptysis. However rigid bronchoscopy may be considered to stop an acute large bleed, where it may be used to tamponade visible bleeding areas using a fogarty catheter or obtaining haemostasis with iced saline, topical adrenaline or thrombin glue.
- Oral atenolol has been used on an anecdotal basis – there is risk of associated bronchoconstriction. Starting dose is 0.5 mg/kg once daily (max 12.5 mg OD). Dose can be titrated up if necessary.
- Recombinant activated factor VII (rFVIIa) may have a role in patients with difficult-to-control haemoptysis or in children awaiting embolisation.8
- Lobectomy may be considered as a last resort

Recurrent Haemoptysis

- Where patients experience on-going haemoptysis despite BAE, identification and modification of potential triggers may be required while taking into consideration of individual needs.
- Where exercise has been identified as a trigger, avoid increasing HR more than 20-30 bpm above resting HR and/or < 120-130 bpm maximum HR and/or where resting HR >100 bpm limit to physical activity only keeping increase HR to a minimum during acute phase of recurrent haemoptysis.
- If tachycardia associated with salbutamol is identified as a trigger, consider reducing the dose or an alternative bronchodilator.
- Avoid or reduce caffeinated drinks.
- Ensure timely and optimal management of any respiratory exacerbation
- If continued concerns of haemoptysis episodes or further episodes of massive haemoptysis, consider repeat BAE.

3. Pneumothorax in Cystic Fibrosis Patients

Pneumothorax has long been recognised as a complication in Cystic Fibrosis (CF), with the first case report published back in 1966.[9]

The lifetime incidence of pneumothorax in people with CF has been reported to be 3.4% (3-19%) [4]. The mean age of occurrence has increased from typically being in the mid-teenage years to the early 20s[10]; when data from pre-1990 is compared with data from the 1990s. Overall, pneumothorax in people with CF is associated with poorer lung function and older age. Occurrence of pneumothorax indicates a poorer prognosis.

3.1 Presentation

Pneumothorax should be considered in patients presenting with –

- Increasing dyspnoea
- Unexplained chest pain
- Haemoptysis

Signs –

- Absent/distant sounding breath sounds on affected side
- Asymmetrical lung expansion
- Hyperresonance on affected side to percussion
- Trachea shifted away from affected side in tension pneumothorax

A chest X-ray (CXR) should be performed if a pneumothorax is suspected, although a CT thorax may be needed in uncertain or complex cases.

Size

- The size of a pneumothorax is **less** important than the degree of clinical compromise.
- The differentiation of a "large" from a "small" pneumothorax is defined as a visible rim of more than 2cms between the lung margin and the chest wall (at the level of the hilum).[5]
- Accurate pneumothorax size calculations are best achieved by CT scan.[10]

3.2 Management

- A small, asymptomatic pneumothorax may be managed conservatively [5,11]. However if the pneumothorax is large or the patient is decompensating or symptomatic regardless of the size, management should be as follows:
- Administer oxygen
- Analgesia with NSAIDs/Opiates
- Insert chest drain (in theatre/ PICU)
- Attach to underwater seal

If acute deterioration – perform emergency needle thoracocentesis

- Equipment – alcohol swabs, large IV cannula, a 5ml and 20ml syringe, 3-way tap
- Position – 2nd intercostal space, mid-clavicular line on side of pneumothorax
- Procedure[11,12]:
  - Place patient on continuous cardiac monitoring and pulse oximetry
  - Patients should be placed in 45-degree, sitting position
  - Palpate landmark (the upper border of the 3rd rib in the midclavicular line) and wipe with alcohol swab
  - Attach a 5ml syringe to the cannula
  - Carefully insert the needle at a slightly downwards angle into the chest wall whilst aspirating the syringe. Stop advancing once air is aspirated.
- In tension pneumothorax, often you will hear a pop or feel a change of resistance
  - Withdraw the needle while gently advancing the cannula downwards into position
  - Secure cannula with tape/tegaderm
  - Attach 3 way tap and 20ml syringe
  - Drain until no further drainage or to a maximum of 30ml/kg (max 2.5l)
  - Perform repeat CXR, a definitive chest drain is likely to be required still
- Empirical IV antibiotics should be considered if there is evidence of associated pulmonary exacerbations. (See Section 2.3)
- REMEMBER regular analgesia
- Physiotherapy Recommendations- Please discuss this with the CF physiotherapist/ physiotherapist on call.
  - Airway clearance should be continued, with avoidance of techniques that increase positive pressure in favour of more controlled techniques (e.g. Active cycle breathing. autogenic drainage).
  - Upper limb weightlifting (>5lb) should be avoided for two weeks post resolution and airway clearance is advisable prior to exercise to minimise the risk of coughing during exertion.
  - Inhaled therapies should be continued, in particular mucolytics to optimise airway clearance.

- NIV should be withheld or in circumstances where ventilatory requirements are such that it cannot be withheld, close monitoring in high level care settings should take place.

Further Management

If the lung does not re-expand, or there is continuing air leak, after several (>3) days, discuss with the paediatric surgical team. In some centres a 50% mortality has been reported in patients who have had a chest drain for more than one week. Pleurodesis or pleurectomy may make future lung transplantation difficult, but they are not absolute contra-indications. However, localised pleurodesis or thoracoscopic stapling may be preferable options.[13]

Patients should not travel by air until 7 days after full resolution on chest x-ray.
Those at higher risk of or recurrent pneumothorax should be advised accordingly.
Higher-risk groups, including those with cystic lung disease such as lymphangioleimyomatosis (LAM) and Brit-Hogg Dube (BHD) syndrome, should be advised accordingly
Patients with trapped lung and chronic air space opacification thought to present a low risk, should be evaluated in secondary care before travel.
No spirometry for 2 weeks after resolution.[5]

4. DIOS in Cystic Fibrosis Patients

Please discuss the management of severe DIOS with the surgical team

Distal intestinal obstructive syndrome (DIOS) is a common complication in CF patients. This results from the accumulation of viscid faecal material within the bowel which combines with thick, sticky mucus produced in the CF intestine. This adheres to the intestinal wall, commonly in the terminal ileum and caecum, making it fixed in position and difficult to remove[14]. The intestine may be completely blocked (complete DIOS) or only partially blocked (incomplete DIOS). The estimated prevalence of complete DIOS is 5 to 12 episodes per 1000 patients per year, with the figures for incomplete DIOS being higher[15]. In a child with previous DIOS, the recurrence risk can be as high as 77%[16]. There are often multiple contributory factors for DIOS including severe CF genotype, pancreatic insufficiency, Inadequate salt intake, dehydration and poorly controlled fat malabsorption.

4.1 Definitions[15]:

- Incomplete /Impending DIOS: Abdominal pain and/or distension and faecal mass in ileocaecum
- Complete DIOS: Abdominal pain and/or distension; faecal mass in ileocecum and complete intestinal obstruction as evidenced by vomiting of bilious material and/or fluid levels in small intestine on abdominal radiography

Differential diagnoses include - appendicitis, intussusception, volvulus, fibrosing colonopathy, biliary tract or gallbladder disease, acute pancreatitis, inflammatory bowel disease, ovarian cysts and urinary tract infections.
Another important differential diagnosis of DIOS is constipation, which is also common in patients with CF. In comparison to DIOS, symptoms of constipation have usually been longer standing and faecal material is distributed throughout the colon on a plain abdominal

X-ray. Constipation never presents with intestinal obstruction. However untreated constipation can progress to DIOS in CF patients.

4.2 Investigations:

- A plain abdominal x-ray (AXR) is usually all that is necessary to diagnose DIOS or constipation. Faecal loading throughout the colon, especially in the right iliac fossa suggest DIOS. Intestinal fluid levels confirm severe DIOS with obstruction; the differential diagnosis of a surgical cause of obstruction must always be considered.
- Urinalysis
- FBC, amylase, LFTs, CRP, urea and electrolytes
- If diagnosis in doubt, consider –
  - Abdominal US scan
  - Water soluble contrast enema can be used as both diagnosis and treatment.
  - A CT scan of the abdomen is rarely indicated, but in DIOS will show significant proximal small-bowel dilatation, with inspissated faecal material in the distal ileum.
  - Obtain a surgical review early, especially in cases of obstruction.

 Following resolution of acute episode, dietitian to review Creon dosing.

4.3 Management

Treatment of DIOS is still largely empirical as there are few randomized controlled trials to guide therapy, thus the following reflects best practice[13].

Incomplete/Impending DIOS – Severe but not obstructed (Severe abdominal pain, no obstruction i.e. no bilious vomiting, no peritonism)

1. Investigations not always necessary

2. Ensure adequate fluid intake – Set goals with the patient. Low threshold for IV fluids, especially if non-compliant/ vomiting.

3. Check dose / compliance / timing of enzyme supplements

4. Ensure adequate salt replacement to improve bile acid absorption in the terminal ileum and correct any bowel CFTR electrolyte imbalance.

5. Dietetic assessment including: fluid intake vs. requirement, fat intake vs. Creon intake, individual dosing at mealtimes and with snacks (which may require a food diary), fibre intake, method of taking enzymes and timing, compliance with laxatives, PPI, Creon and ensure adequate dietary fibre intake.

6. Consider acid reduction to enhance enzyme (Creon) performance
If not on a PPI add in Lansoprazole3.75mg for children 3kg-5kg
7.5mg for children 5kg<10kg
15mg for children 10<30kg
30mg for children >30kg

The below oral treatments are contraindicated in complete bowel obstruction

7. Klean Prep – polyethylene glycol
   - **Do not** administer just before bedtime due to risk of aspiration
   - **Do not** use in presence of bile stained vomiting
   - Can be given via NG tube. Despite taste, most patients prefer to take orally
   - Monitor CF diabetics undergoing this regimen for hypoglycaemia.
   - Administration –

- Add contents of 1 sachet to 1 litre cool water/fruit cordial.
- Start at 10ml/kg/hour for 30 mins then 20 ml/kg/hour for 30 mins (for oral administration calculate volume to be consumed in each 30 min)
- If well tolerated this can be increased to 25 ml/kg/hour.
- Maximum volume is 100 ml/kg or 4 litres (whichever is smaller) over 4 hours.
- First litre should be given under observation. Patients often respond within this time and need for further treatment should be reviewed.
- If treatment is to continue, patients must be reviewed after the first 4 hours.
- A further 4 hours treatment can be given if necessary
- Maximum daily dose should be 200 ml/kg or 8 litres (whichever is smaller), however in practice it is unusual to prescribe more than 4 litres a day. The standard adult dose is 2 litres on two successive days.

8**.** Oral gastrograffin
contains iodine and is highly osmotic. Gastrograffin may cause considerable fluid shift from the circulation to the bowel and serious complications have been reported, including, shock, perforation and necrotizing enterocolitis

- Patients must be well hydrated (with normal electrolytes) before, during and after treatment. In more severe cases, IV fluids/electrolyte correction may be required.
- The suggested fluids below are the minimum -
- <15kg: 15-30mls gastrograffin diluted with 45-90ml water or juice (i.e. 1:3 dilution)
- 15-25kg: 50mls gastrograffin diluted with 150ml water or juice
- >25kg: 100mls gastrograffin diluted with 200ml water or juice
- Repeat at 24 hours if no response but not if symptoms worsen.
- Encourage fluid intake for 3 hours after administration

9. Oral N-Acetyl Cysteine (NAC) – its use has been largely superseded by the above medications but may be tried (Consultant decision) should other agents fail. Acts as a mucolytic and can help break up the protein matrix of the inspissate.

- Administration –

1 month to 1 year 0.4-3g as a single dose

- 2-6 years 2-3g as a single dose
- 7-17 years 4-6g as a single dose
- Dilute 20% (200mg/ml) injection to 50mg/ml.
- Bitter taste of injection may be masked with cola, orange or blackcurrant juice.

Complete DIOS

Involve the paediatric surgical team urgently

1. Keep patient nil by mouth
2. IV fluids and NG tube on free drainage (drip and suck)
3. Urgent surgical review
4. Investigations as above
5. **Oral treatments cannot be given in the presence of bile stained vomiting**
6. Rectal Gastrograffin – discuss with CF consultant, surgical and radiology teams
   - Administer under radiological guidance only.

- Monitor for dehydration and perform a plain AXR at 1 hour to exclude massive dilation (toxic megacolon). If the latter is present, urgent paediatric surgical review is required
- IV access – Start IV fluids
- Administration –
  - Use same dose as oral (i.e. 15-30mls gastrograffin if weight is <15kg, 50mls gastrograffin if 15-25kg, 100ml gastrograffin if >25kg)

    but if age –
    1. <5 yrs: dilute to 5 times its volume with water
    2. >5 yrs: dilute to 4 times its volume with water

7. Colonoscopy or surgery is rarely required; although is indicated where above medical management has failed. Surgical options may involve laparotomy and enterostomy or even bowel resection.

Post-acute event management
Most DIOS patients will have more than one episode, therefore Laxatives e.g. Movicol or Lactulose (can cause abdominal pain and flatulence ay high dose, therefore movicol should be first preference) should be continued for 6-12 months post DIOS depending on bowel motions[16].
In addition, dehydration and fat malabsorption should be avoided to help prevent recurrence. Therefore, Steps 2-6 as per management of severe symptoms should be continued at discharge.

5. Haematemesis in Cystic Fibrosis Patients

Reports of the prevalence of liver disease in CF vary but cirrhosis has been reported in 24% CF patients and up to 50% in postmortem findings. However, symptomatic liver disease is uncommon, being reported as the cause of death in only 2% of CF patients. Incidence does not rise progressively but seems to peak during the second decade and is more common in males (3:1). There is no known genotype-phenotype correlation but there is a high familial concordance and strong association with certain polymorphisms that may be predictive of future disease.

Presentation of upper GI bleed in association with varices (secondary to portal hypertension) can be torrential and prompt resuscitation with an urgent assessment of ABC is a priority. Initial management is as per current APLS guidelines. Please refer to Upper Gastrointestinal Bleeding UHL Childrens Guideline available on INsite for further details on specific management.

Education and Training

Appropriate medical staff to receive APLS training.

Monitoring Compliance

What will be measured to monitor compliance	How will compliance be monitored	Monitoring Lead	Frequency	Reporting arrangements
Evaluate the occurrence of severe CF emergencies and effective management	Audit	Erol Gaillard	Yearly	Audit report
Reported patient safety incidents relating to this group of patients	Review of Datix forms	Dr Erol Gaillard	Annually	
Adherence to prescription guidelines	Review of Datix forms	Ms Vandana Pankhania	Annually	

Supporting References

1.Flume PA, Yankaskas JR, Ebeling M, Hulsey T, Clark LL. Massive hemoptysis in cystic fibrosis. Chest 2005;128:729–38.

2. Barben JU, Ditchfield M, Carlin JB, Robertson CF, Robinson PJ, Olinsky A. Major haemoptysis in children with cystic fibrosis: a 20-year retrospective study. J Cyst Fibros 2003;2:105–11.

3. Efrati O, Harash O, Rivlin J, et al. Hemoptysis in Israeli CF patients - Prevalence, treatment, and clinical characteristics. Journal of Cystic Fibrosis 2008;7:301–6.

4. Semin Respir Crit Care Med. 2019 Dec;40(6):804-809. doi: 10.1055/s-0039-1697639. Pulmonary Complications of Cystic Fibrosis. Garcia B, Flume PA.

5. Flume PA, Mogayzel JPJ, Robinson KA, et al. Cystic fibrosis pulmonary guidelines: Pulmonary complications: Hemoptysis and pneumothorax. Am J Respir Crit Care Med 2010;182:298–306; updated April 2019 https://www.cff.org/Care/Clinical-Care-Guidelines/Respiratory-Clinical-Care-Guidelines/Pneumothorax-and-Hemoptysis-Clinical-Care-Guidelines/

6. Hurley M, Bhatt J, Smyth A. Treatment massive haemoptysis in Cystic Fibrosis with Tranexamic acid. J R Soc Med 2011: 104: S49–S52.

7. Ther Adv Respir Dis. 2013 Aug;7(4):217-23. doi: 10.1177/1753465813482744. Beta-blocker management of refractory hemoptysis in cystic fibrosis: a novel treatment approach. Moua J, Nussbaum E, Liao E, Randhawa IS.

8. Lau EMT, Yozghatlian V, Kosky C, et al. Recombinant activated factor VII for massive hemoptysis in patients with cystic fibrosis. *Chest*. 2009 Jul;136(1):277-81. doi: 10.1378/chest.08-2948.

9. Flume PA. Pneumothorax in cystic fibrosis. Curr Opin Pulm Med 2011;17(04):220–225

10. Flume P, Strange C, Clark L et al. Pneumothorax in Cystic Fibrosis. Chest 2005;128(2):720-728

11. BTS Pleural Disease Guideline 2010. https://www.brit-thoracic.org.uk/document-library/clinical-information/pleural-disease/pleural-disease-guidelines-2010/pleural-disease-guideline-quick-reference-guide/

12. Advance Paediatric Life Support Manual. 6th Edition.

13. The Royal Brompton Hospital. Clinical Guidelines: Care of Children with CF. 2020. 8th Edition

14. Colombo C, Wilschanski M et al. Guidelines for the diagnosis and management of distal intestinal obstruction syndrome in cystic fibrosis patients. Journal of Cystic Fibrosis Volume 10 Suppl 2 (2011) S24–S28

15. Houwen RH, van der Doef HP, Sermet I, et al. on behalf of the ESPGHAN Cystic Fibrosis Working Group. Defining DIOS and constipation in cystic fibrosis with a multicentre study on the incidence, characteristics, and treatment of DIOS. J Pediatr Gastroenterol Nutr 2010;50:38–42.

16. World J Gastroenterol. 2015 Jan 7;21(1):318-25. doi: 10.3748/wjg.v21.i1.318. Long-term follow-up of distal intestinal obstruction syndrome in cystic fibrosis. Lavie M1, Manovitz T1, Vilozni D1,et al

17. Van der Doef HJ, Kokke FM, van der Ent CK, Houwen RJ. Intestinal Obstruction Syndromes in Cystic Fibrosis: Meconium Ileus, Distal Intestinal Obstruction Syndrome, and Constipation. Curr Gastroenterol Rep (2011) 13:265–27

Key Words

Cystic Fibrosis, Distal Intestinal Obstructive Syndrome, Haematemesis, Haemoptysis, Paediatric, Pneumothorax

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The Trust recognises the diversity of the local community it serves. Our aim therefore is to provide a safe environment free from discrimination and treat all individuals fairly with dignity and appropriately according to their needs.
As part of its development, this policy and its impact on equality have been reviewed and no detriment was identified.

Contact and review details	
Guideline Lead (Name and Title) Imad Ahmed Consultant in Paediatric Respiratory Medicine	**Executive Lead** Chief Medical Officer
Details of Changes made during review: 1. All patients with haemoptysis require admission to hospital 2. Discuss haemoptysis with the CF Physiotherapist at the earliest opportunity 3. Management options for severe haemoptysis 4. Amendment to the DIOS protocol	