University Hospitals of Leicester NHS
NHS Trust

LRI Children's Hospital

Inpatient management guideline for chest exacerbations in children with Cystic Fibrosis

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 Guideline applies to

This document is a short guideline for managing children with Cystic Fibrosis (CF) admitted to one of the UHL Children's wards for a chest exacerbation, usually on an elective/ semi-elective basis. The duration of admission is usually for 2 weeks, but the child may in some cases be managed with home antibiotics after spending the first few days in hospital. Most of the following guideline is based on the 2022 standards of care document for children with Cystic Fibrosis in the UK https://www.cysticfibrosis.org.uk/sites/default/files/2022-10/Standards%20of%20care_interim%202022.pdf. During inpatient admissions, ensure meticulous attention is paid to infection control standards.

Related documents

- Infection Prevention UHL Policy B4/2005
- Cystic Fibrosis Paediatric Prescribing UHL Childrens Hospital Guideline C35/2016
- Cystic Fibrosis Emergencies UHL Childrens Medical Guidelines C64/2015

2. Admission of CF Children

2.1 Wards

All children must be admitted to a cubicle/side-room with its own toilet or nearby allocated toilet. Certain respiratory pathogens are difficult to treat and can be associated with poorer prognosis in CF patients. These include Pseudomonas aeruginosa, Burkholderia cepacia complex (BCC), Non-Tuberculous mycobacteria (NTM) and Methicillin Resistant Staph Aureus (MRSA).

Not more than one patient can be admitted on the same ward unless approved by the CF Team.

We avoid admissions to ward 12 because many patients there are colonised with Pseudomonas aeruginosa. This does not apply to CF patients needing HDU care.

2.2 Admission documentation

The following should be documented in admission notes (please use CF admission document, Appendix 1):

- Reason for hospital attendance.
- Most recent positive sputum culture result with full sensitivities.
- Previous isolation of BCC, NTM or MRSA (for infection control).
- Most recent & patient's best FEV_1 & FVC (over the last year).
- Other complications of CF, including but not limited to ABPA, CF related diabetes.
- Full drug history including allergy. Please remember to write up the regular drugs on the inpatient prescription chart. Ensure all oral nutritional supplements/ feeds and flushes are also prescribed. See prescribing information available on INsite/MEDUSA.

**Note: Nebulised aminoglycosides / nebulised colistin is usually stopped if IV aminoglycosides are prescribed (tobramycin / amikacin).

- Social issues, including but not limited to, school attendance and exposure to environmental/ tobacco smoke.
- Examination findings including-
  Weight (kg & centiles) and Height (cm & centiles), BMI, Head circumference in <1 year old children, temperature, respiratory rate, heart rate, blood pressure, Oxygen saturation in air or oxygen (include O_2 flow rate if relevant).

- Systemic examination including Respiratory system and gastrointestinal, specifically liver size and presence of right iliac fossa mass. Remember to examine ENT including Nasal polyps and tonsillar enlargement.

2.3 Admission Investigations

- Spirometry (next working day if coming in Out of hours).
- Admission bloods (ensure appropriate pain relief), try to organise at the same time as long line insertion/ portacath access.
  - FBC, U&E, CRP, LFT'S, bone profile, Mg
  - Add in following if not performed in the last 3 months or in case of clinical suspicion (e.g. wheezy, patchy changes in chest X Ray, coughing up brown/ black bits) – Immunoglobulins including Total IgE, Specific IgE for aspergillus and aspergillus precipitins.
  - Add in appropriate additional bloods if due for annual review
- Cough swab/ Sputum.
- NPA for virology if clinically indicated.
- Urinalysis if on steroids/ recent weight loss.
- Chest X Ray is not routinely done. It is done ONLY if clinically indicated (e.g. suspicion of pneumothorax/ ABPA).

3. Treatment

- In planned admissions, the choice of IV antibiotics is made at the CF MDT in the presence of microbiologist and CF consultants, the decision is based on previous drug history, microbiology and antimicrobial susceptibility. This is communicated to the CF Fellow/respective ward junior doctors before admission.
- Generally, we use at least 2 antibiotics, usually an aminoglycoside + other class of antibiotics. The choice depends on previously isolated organism and previous drug history. The only exception to this is patients who have not previously isolated pseudomonas, where IV cefuroxime is usually used as a first agent.
- Children with fungal lung disease may need to be started on IV antifungals. This should be a decision made by the CF MDT with microbiology consultation. If Children are started on IV AmBisome, they should be prescribed IV fluids overnight (full maintenance for 12 hours).
- Twice daily physiotherapy input is essential
- Addition of nebulised hypertonic saline / DNAse should be considered, if appropriate, following discussion with CF physiotherapist and consultants.

- If admitted out of hours for reasons other than chest exacerbation, please refer to Cystic Fibrosis Emergencies UHL Childrens Medical Guidelines. If admitted out of hours for a chest exacerbation, please review the recent microbiology and start on appropriate antibiotics. Please ensure that the CF team is informed of the admission the next morning.

4. Further investigations during admission:

- Weekly blood test at a minimum- U+E's LFTs and FBC
- Twice weekly weight and review by dietitian.
- Weekly spirometry
- Daily BP and urinalysis if on oral / IV steroids
- Daily review of results, blood and microbiology prior to reviewing patient.
- If child has had raised HbA1c, is on steroids or if recent pulmonary deterioration or weight loss, consider blood glucose monitoring/CGM.
- Regular sputum / cough swab (at least twice weekly while admitted)
- Consider Overnight oximetry early in admission, especially if FEV_1<50% or resting SaO_2 <92%
- Minimum twice daily observations including SaO_2.

Important! Monitoring of drug levels:

- For children receiving once daily Tobramycin, Pre- dose Tobramycin levels before second dose and eighth dose along with U+Es.
- For children receiving once daily amikacin, levels to be taken pre 2nd dose, then every 3 days for duration of antibiotic course.(See Cystic Fibrosis Paediatric Prescribing UHL Childrens Hospital Guideline). For three times a day amikacin dosing, seek advice from the CF paediatric pharmacist.

** Note: DO NOT take blood from the long line or portacath.

5. Discharge

A full discharge summary must be done on Nervecentre for all admissions. Ensure the following is documented on discharge:

- General conclusions about the admission
- Results awaited
- Weight on admission & discharge
- Spirometry results (FEV_1, FVC) on admission & discharge
- SaO_2
- Follow up plan
- Plan for tests necessary at home
- Date of next admission if elective (3 monthly IV antibiotics, monthly IV steroids)
- Document treatment received during current admission including name, dose and duration of antibiotics, microbiology and important results during this admission.
- Drugs on discharge (including any weaning):

- If on nebulised Colistin (Colomycin / Promixin) or Tobramycin (TOBI/Bramitob) then on discharge this will need to be restarted if stopped during admission.
- Continue all the long term medications unless any changes are made by the CF team.
- Most children will go home on oral antibiotics. CF doses are usually severe infection dose.(See Cystic Fibrosis Paediatric Prescribing UHL Childrens Hospital Guideline)
- Clarithromycin is a common antibiotic used in CF infective exacerbations, this SHOULD NOT be used in children who are on CFTR modulators (Ivacaftor, Orkambi, Symkevi, Kaftrio).

- The CF Team will make a decision as to what take home medication will be required prior to discharge.
- Ensure that TTO' s are written up at least one day before planned discharge date and inform CF Pharmacist/paediatric pharmacy team who can arrange for the medications to be dispensed.

3. Education and Training

None

4. Monitoring Compliance

What will be measured to monitor compliance	How will compliance be monitored	Monitoring Lead	Frequency	Reporting arrangements
Antibiotic monitoring	Internal audit/service evaluation/peer review	Dr Gaillard	2 yearly audit & evaluation	Departmental audit group

5. Supporting References

6. Key Words

Cystic fibrosis, Inpatient

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.

All documents within PAGL are also published automatically on the Trust's external website, **unless** such publication would enable a person to endanger their own health/safety or that

of another person. An example of this is the Anti Ligature Policy, which is not available on the Trust's external website due to a national Patient Safety Alert.

CONTACT AND REVIEW DETAILS	
Guideline Lead (Name and Title) Erol Gaillard Consultant in Paediatric Respiratory Medicine and UHL CF Paediatric Centre Director	**Executive Lead** Chief Medical Officer
Details of Changes made during review: 1. Removed stipulation of wards for patients with infections as more inpatients with PCD and bronchiectasis. Decisions made on a case by case basis. 2. Updated to the latest guideline documents. 3. Added weekly bloods 4. It is important to note that most of the care is delivered by the CF team. Most involvement of the general paediatric team relates to a) CF emergencies and b) drug monitoring.	

Appendix 1

MEDICAL ADMISSION NOTES	
Patient information	
Name : Address : Date of Birth : Hospital No :	Consultant : Admission date : Discharge date : Ward : Admitting doctor : Signature :

Reason for admission:					
General	☐Well	☐Unwell			
Cough	☐Nil ☐Wet	☐Occasional ☐Dry	☐Frequent ☐Both	☐Constant Onset :	☐Nocturnal
Sputum	☐No	☐Swallows	☐ < 10 ml	☐ 20 ml	☐50 ml
Colour	☐Clear	☐Cream	☐Yellow	☐Green	☐Brown ☐Blood
Wheeze	☐Nil	☐Rare	☐Frequent	☐ Constant	☐ With exercise
Nocturnal	☐Nil	☐Occasional	☐Frequent		
Nose	☐Normal	☐Blocked	☐Runny	☐Hay fever	☐Nocturnal
URTI	☐Yes	☐No	Onset :		
Appetite	☐Good	☐Moderate	☐ Poor		
Abdo pain	☐Nil	☐Occasional	☐Frequent	☐Reflux	Frequency:
Bowels/day	☐<1	☐1-2	☐3-4	☐>5	
Stools	☐Normal	☐Pale	☐Loose	☐Bulky	☐Fatty ☐Hard
Vomiting	☐Nil	☐Occasional	☐Frequent	Heartburn	☐Yes ☐No
Any other symptoms:					

CF complications:	☐Yes	☐No
Tick all complications that apply:		
☐CFRD	☐ABPA	☐Asthma
☐Liver related	☐Arthritis/Arthropathy	☐Nasal polyps
☐GORD	☐Pancreatitis	☐DIOS
☐Sinus disease	☐Hearing loss	☐ Septicaemia
☐GI Bleed	☐Haemoptysis	☐Pneumothorax
☐Rashes	☐Atopy	
☐*Other – specify:*		

Last admission	
Date :	Reason:
Treatment given and duration:	

Microbiology (Past 12 months)							
	Date	Date	Date	Date	Date	Date	Date
Bacteria							
Viral							
Fungi							
Other:							

Medications				
Name	Dose	Times	Route	Device/ any comments

Allergies :

Examination					
Cough	☐ Dry	☐ Wet click	☐ Wet		
General	☐ Well	☐Unwell	☐ Ill		
Clubbing	☐ Nil	☐ Mild	☐Moderate	☐Severe	
Nose	☐Normal	☐Abnormal	☐ Polyps	☐URTI	
Mouth/Throat	☐Normal	☐Candida	☐URTI		
Chest shape	☐Normal	☐Other:			
Crackles	☐Absent	☐Present			
Wheeze	☐Absent	☐Present			
CVS					IV Access:
Abdomen					Cannula ☐Leaderflex ☐
Distension	☐Nil	☐Moderate	☐Severe		Inserted By:
RIF mass	☐Absent	☐Present			Date:
Liver	☐Normal	☐Abnormal	cms palpable		Port/Hickman☐
Spleen	☐Normal	☐Abnormal	cms palpable		Inserted(mm/yy):

Observations			
Temp	HR	RR	Sats
BP(mm Hg)	Blood sugar		
Weight(kg)	Height (cm)	BMI(kg/m2)	HC

Blood tests	*Date*	*Interpretation*
Full blood count		
Renal function tests		
Liver function tests		
Bone profile		
CRP		
Aspergillus markers		
Other		

Drug monitoring		*Date*	*Results and interpretation*
Tobramycin	Day 2		
	Day 8		
Amikacin			

Microbiology		
Samples	Date	Results

Lung function			
Date	FEV1 in L (%pred)	FVC in L (%pred)	FEV1/FVC

Any other investigations		
Investigations	Date	Result

Management plan

- Nurse in isolation
- Plot height and weight
- Inform dietitian, physiotherapist (even over holidays) and diabetes team if on insulin
- Specific treatment to start:

- Continue usual medications. Stop Colomycin if on Tobramycin
- IV access
- Investigations:
  - Bloods at admission (FBC, U+Es, LFT, Bone profile, Mg)
  - Sputum / Cough swab M, C & S
  - Spirometry
  - Consider CRP, Immunoglobulins, Aspergillus IgE, Aspergillus precipitins,
    - NPA, Clotting, CXR if clinically indicated
  - Monitor BM (If age >10yrs, initial high random sugar/HbA1c, or on oral steroids)
  - Monitor levels if on Tobramycin and Amikacin

Doctor's name: ***Signature:***

Other notes

Name: ***Signature:***

Physiotherapy

Name: ***Signature:***

Dietitian

Name: ***Signature:***

Nursing

Name: ***Signature:***

Discharge plan