CLINICAL GUIDELINE FOR THE ADMISSION OF A CHILD OR YOUNG PERSON WITH CYSTIC FIBROSIS

1. Aim/Purpose of this Guideline

1.1. This guideline applies to all staff caring for children/young people admitted to child health with Cystic Fibrosis.

2. The Guidance

2.1 Record on Admission:

Weight (Plot on centile chart, needs to be repeated twice weekly, Mondays and Thursdays)
Temperature
Blood Pressure
Heart Rate & Pulse Rate
SaO₂ (needs to be measured daily and overnight on first night)
BM I hr post prandial and staggered over first three days

2.2 Investigations on Admission;

Blood for
- FBC
- U&E’s
- CRP
- IgE + Aspergillus RAST
- Aspergillus Precipitans
- Glucose + glycosulated Hb
- Pseudomonas serology (If not chronically colonised)

If Pre-op will need in addition;
- LFTs
- Clotting Studies
- Calcium
- X match may be required, check with surgeons

Sputum for Culture & Sensitivity
Virology
(In children in whom it is difficult to obtain sputum sample NPA post physiotherapy should be obtained with or without the use of nebulised 7% saline).

CXR should only be performed if patient has new localising signs.

POLICY UNDER REVIEW

Please note that this policy is under review. It does, however, remain current Trust policy subject to any recent legislative changes, national policy instruction (NHS or Department of Health), or Trust Board decision. For guidance, please contact the Author/Owner.
### 2.3 Physiotherapy

On admission the paediatric physiotherapist (or the duty physiotherapist at weekends) should be informed that the patient is in. The physiotherapist will organise the physiotherapy protocol for the duration of the patient's stay and spirometry. She will liaise with the nursing and medical staff as to the optimal timing of inhaled medication.
2.4 Dietician

The Cystic Fibrosis dietician should be informed that the patient has been admitted so that a dietetic review can be performed. If the child uses nutritional supplements please ensure that these are obtained from Pharmacy. Please document food and fluid intake as well as stool charts.

2.5 Infection Control

Isolation

Patients with cystic fibrosis should be admitted to single rooms in order that;
1. Their acquisition or respiratory viruses of pseudomonas from other patients on the wards is prevented
2. Cross infection between patients with different strains of pseudomonas etc is prevented.

If a patient is found or suspected to be colonised with Cepacia or MRSA he should be admitted to a single room with private washing and toilet facilities and strict barrier nursing procedures should be adhered to.

Stethoscopes

Each patient should have their own stethoscope for the duration of their stay. These stethoscopes should be thoroughly cleaned with alcohol wipes after the patient has been discharged home.

Spirometry Equipment

The spirometry equipment is cared for by the physiotherapist, a new bacterial/viral filter is used for each patient. Only expiratory measurements are performed. The surface of the spirometer is cleaned with an alcohol wipe between patients. The ward peak flow meters should not be used in patients with CF.

2.6 Antibiotic Therapy

Choice of antibiotic therapy should be guided by the most recent sputum result. The Public Health Laboratory should be contacted to ascertain which organisms are growing in the sputum and the antibiotic sensitivities of these organisms.

Staph Aureus

For the first two years of life patients with cystic fibrosis are placed on prophylactic Flucloxacillin in order to help limit the damage caused by staph aureus. In older children chronically colonised with staph aureus Flucloxacillin prophylaxis is continued.

Dosage schedule for prophylaxis;

Flucloxacillin

Primary prevention
125 mg bd 1month – 3yrs

Secondary prevention
50mg/kg (max 1 gm) bd 1month – 18yrs
Acute Exacerbations

0-2 years
When patients in the first two years of life are admitted for IV antibiotic therapy the dose of
Flucloxacillin is increased and given IV in addition Cefotaxime is added to provide cover
for other organisms such as haemophilus influenza.

Dosage schedules for IV therapy;
Flucloxacillin 25 mg/kg/dose qds as IV bolus over 5 minutes
Cefotaxime 50 mg/kg/dose tds diluted 10 times with 5% Dextrose or 0.9% saline and
given as an IV infusion over 30 minutes

> 2 years
IF COLONISED WITH STAPH AUREUS BUT NOT COLONISED WITH PSEUDOMONAS; use;
1. Flucloxacillin 25 mg/kg/dose qds po
2. Cefotaxime 50 mg/kg/dose (maximum dose 12 g daily) diluted 10 times with 5%
Dextrose or 0.9% saline and given as an IV infusion over 30 minutes
and 3. Tobramycin 10 mg/kg od (max. 660 mg) by infusion over 30 minutes
(levels and U&Es need to be performed just prior to the second or third dose and 9th
or 10th dose)
Trough < 1 mcg/ml
If the dose needs adjusting levels should be performed again before the second or third
dose post-adjustment.

Or if staphylococcus resistant to Tobramycin
Use a) Clindamycin 7 mg/kg/dose qds (max 600 mg) by IV infusion over at least 10
minutes (diluted to at least 6 mg/ml in 0.9% saline or 5% Dextrose)
(Clindamycin should be discontinued if patient develops diarrhoea.)
LFTs and FBC should be performed on 9th day of treatment.
Or b) Fucidin po Child 1 month-1 year 15mg/kg tds
Child 1-5 year’s 250mgs tds
Child 5-12 year’s 500mgs tds
Child 12-18 year’s 750mgs tds
Avoid or reduce dose in patients with liver disease.

IF NOT COLONISED WITH STAPH AUREUS OR PSEUDOMONAS

> 2 years
1. Cefotaxime 50 mg/kg/dose tds diluted 10 times in 5% Dextrose or 0.9% saline
as an IV infusion over 30 minutes, up to a maximum dose of 12 gm daily.
Adjust antibiotics as required when sputum result is available.
2.7 Treatment of Exacerbations in patients chronically colonised with pseudomonas

Patients are often admitted for the first few days of IV antibiotic therapy and intensive physiotherapy. If the patient would like to continue with IV antibiotics at home these are ordered via BUPA Healthcare. IV antibiotic courses last for two weeks but can be extended if necessary. During exacerbations patients increase their physiotherapy sessions at least to twice daily. (Please ensure that the patient/parent is aware that if a hospital supply is used to start home treatment then the BUPA Healthcare supply, when it arrives, should be used instead and not in addition). Oral macrolides should be discontinued during courses of IV antibiotic therapy. Nebulised Tobi, if used, may be continued but needs to be given in the morning and evening and the trough tobramycin levels taken predose in the afternoon (these should not be taken from the Port or long line).
Nebulised hypertonic saline (7 or 6%) 4 mls bd may be required to aid sputum retrieval but the first dose needs to be carefully supervised and many patients require pre-treatment with salbutamol therapy.

Choice of Antibiotics

Two IV anti-pseudomonal agents are administered. If the organism is sensitive Ceftazidime and Tobramycin are the first choice combination. Other antibiotics which can be used include Amikacin, Aztreonam, Imipenem, Meropenem, Ticarcillin. Piperacillin and Colomycin can also be used but only if the organism is multi-resistant as Piperacillin is associated with a high incidence of drug hypersensitivity reactions and IV Colomycin is associated with neuro and nephro toxicity.

Dosages

1st line
1. **Ceftazidime** 50 mg/kg/dose tds as an infusion over 30 minutes
   and 2. **Tobramycin 10 mg/kg od (max. 660 mg) by infusion over 30 minutes**
      (levels and U&Es need to be performed just prior to the second or third dose and 9th or 10th dose)
Trough < 1 mcg/ml
If the dose needs adjusting levels should be performed again before the second or third dose post-adjustment.

If the dose needs adjusting levels should be performed again before the third dose post-adjustment. Levels should be performed again on the 9th day. If the levels are high then U&E’s and creatinine should be measured.
2nd choice anti-pseudomonal agents

**Amikacin** 10 mg/kg/dose tds by slow IV bolus. (Up to a maximum of 500 mg every 8 hours)
Monitor levels and U&E’s before and one hour after the second or third dose if used tds.
Peak 15-30 mg/L
Trough < 10 mg/L
Or if used once daily:
15 mg/kg od diluted to 50 ml with 0.9% saline and infused over 30 min.
Trough < 5 mg/L 24 hours post second dose
Aim for the higher end of the therapeutic range when adjusting the dose if using tds. If the dose needs to be adjusted levels need to be performed again on the third dose post adjustment. Levels and U&E’s are again performed on the ninth day.

**Imipenem** 25 mg/kg/dose (max 1gm) qds by IV infusion over 20 minutes, Reconstitute each 500 mg vial with 100 ml 0.9% saline or 5% Dextrose and give over 20-30 minutes.

**Meropenem** 1 month-12years body weight under 50 kg 40 mg/kg/dose tds, body weight over 50 kg 2g tds. 12-18years 2g tds by IV infusion over 20 minutes. Reconstitute each 500 mg vial with 100 ml 0.9% saline or 5% dextrose

**Ticarcillin** 1 month-18 years body weight under 40 kg 80 mg/kg/dose tds
Adult or body weight over 40 kg 3.2 gm qds
(not as Timentin as Clavulanic Acid is associated with cholestatic jaundice) given as an IV infusion over 30 minutes after diluting in 100 ml 5% Dextrose or water for injections and given over 30 minutes. Adult or body weight over 40 kg 3.2 gm qds

3rd choice anti-pseudomonal agent

**Colomycin** < 60 kg 25,000 units/kg tds
> 60 kg 2 million units tds
by IV infusion over 30 minutes [dilute to concentration of 40,000 units/ml with 0.9% NaCl]

**Piperacillin** give with Tazobactam as Tazocin 4.5 gm tds diluted with 100 ml 0.9% saline IV infusion over 30 minutes in children of over 12 years of age only. (Should be given with an aminoglycoside if organism is sensitive as they have a synergistic effect).
2.8 Patients growing Pseudomonas in their sputum for the first time

Our current procedure is to give patients who have grown pseudomonas in their sputum for the first time for a four week course of oral ciprofloxacin 15mg/kg bd and nebulised colistimethate sodium 1 million units bd if under two years of age and 2 million units bd over two years of age. (for a second pseudomonas isolate a 3 month course of Ciprofloxacin and a three month course of nebulised Colomycin is the treatment of choice). Anna Rogers will organise the nebuliser if requested. If the patient is unwell a two week course of intensive physiotherapy and IV antibiotics is advisable especially if we do not feel that their treatment at home is optimal Two IV antipseudomonal agents are used and the usual combination of choice is Ceftazidime and Tobramycin but this choice is influenced by sputum sensitivities. Whilst in hospital blood is sent for pseudomonas serology, the physiotherapy techniques are reviewed and nebulised antibiotic therapy is commenced, i.e., Colomycin. Sputum is obtained for culture and sensitivity at the end of the course of IV antibiotics and if pseudomonas is no longer present in the sputum all the old spacehalers and nebuliser tubing are replaced.

Sputum samples are sent fortnightly from home. If three consecutive sputums have been clear of pseudomonas nebulised antibiotics are discontinued at the end of the three month period.

Sputum samples or cough swabs are sent monthly thereafter and if pseudomonas is isolated again a further 2 week course of IV Ceftazidime and Tobramycin is considered. These cycles are continued until the patient becomes chronically colonised.

Nebulised Tobramycin therapy is used in subjects who continue to deteriorate (see separate TOBI guideline).

2.8.1 Nebulised Therapy Antibiotics

<table>
<thead>
<tr>
<th>Antibiotic</th>
<th>Duration</th>
<th>Dose</th>
</tr>
</thead>
<tbody>
<tr>
<td>Colistin</td>
<td>1 month - 2 years</td>
<td>1,000,000 units bd</td>
</tr>
<tr>
<td>Injection solution used for nebulisation</td>
<td>2 – 18 years</td>
<td>2,000,000 units bd</td>
</tr>
<tr>
<td>Tobi</td>
<td>300 mg bd alternate months</td>
<td></td>
</tr>
</tbody>
</table>
2.9 Bronchodilators

β adenergic responsiveness should be tested in patients with cystic fibrosis on a regular basis both during and between exacerbations. Some patients with cystic fibrosis may show a deterioration in FEV₁ post treatment due to airway damage and these patients should not be given β adenergic agents. If an improvement in FEV₁ is seen even if this improvement is modest (e.g., 5-10% improvement) but reproducible then a β adenergic agent should be administered approximately 15 minutes prior to physiotherapy.

Bronchodilators

<table>
<thead>
<tr>
<th>Age</th>
<th>Salbutamol</th>
<th>mcg</th>
<th>Device</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;2½ years</td>
<td>Salbutamol</td>
<td>200</td>
<td>Aerochamber &amp; mask</td>
</tr>
<tr>
<td>2½ - 5 years</td>
<td>Salbutamol</td>
<td>200</td>
<td>Volumatic + mask</td>
</tr>
<tr>
<td>&gt; 5 years</td>
<td>Salbutamol</td>
<td>200</td>
<td>Accuhaler or Volumatic</td>
</tr>
</tbody>
</table>

Note:
1. Dry powder devices should not be given to pre-school children.
2. Children of 2½ to 5 years of age who can use the mouthpiece of the spacer device without a mask when not in respiratory distress may not be able to do so when unwell, therefore all children of 2½ to 5 years of age should be issued with a suitable mask
3. Only single doses of aerosol should be administered into the spacer device at a time, followed by 5 tidal breaths and then the dose repeated.
4. The MDI should not be actuated before the face mask or mouthpiece of the spacer device has been applied correctly.

Long acting β adenergic drugs such as serevent are prescribed for all patients of over 5 years of age with cystic fibrosis who also have asthma or exhibit β adenergic responsiveness. They are given via the same devices as salbutamol and administered after physiotherapy unless the patient is not using a short acting β adenergic agent, when they are administered 15-20 minutes before physiotherapy.
2.10 Vitamins

< 5 years  Dalivit /Abidec as prescribed.  
            + Vitamin E 50 mg/day

≥ 5 years  Multivitamins (BPC) 1 per day
            + vitamin E 100 mg/day

≥ 12 years Multivitamins 2 per day
            + vitamin E 200 mg/day

Vitamin A, E and D levels are measured yearly and dose is adjusted according to response.

2.11 Discharge planning

The CF nurse, community CF physiotherapists and GP need to be informed at least 24 hrs prior to a planned discharge and if the patient is going home on IV antibiotics the CF nurse, or her nominated deputy, needs to make sure that the patient has all of the equipment required for therapy. Medical staff are required to ensure that the patient has all the medications they require. If the child has been started on nutritional supplements they will also need to be provided on the TTO’s.
3. Monitoring compliance and effectiveness

<table>
<thead>
<tr>
<th>Element to be monitored</th>
<th>Compliance with admission procedure.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lead</td>
<td>CF team</td>
</tr>
<tr>
<td>Tool</td>
<td>Peer review and patient notes audit</td>
</tr>
<tr>
<td>Frequency</td>
<td>Annually or as required</td>
</tr>
<tr>
<td>Reporting arrangements</td>
<td>Via audit and guidelines</td>
</tr>
<tr>
<td></td>
<td>And dissemination to CF team</td>
</tr>
<tr>
<td>Acting on recommendations and Lead(s)</td>
<td>Required actions will be identified and completed in a specified timeframe</td>
</tr>
<tr>
<td>Change in practice and lessons to be shared</td>
<td>Required changes to practice will be identified and actioned within. A lead member of the team will be identified to take each change forward where appropriate. Lessons will be shared with all the relevant stakeholders</td>
</tr>
</tbody>
</table>
Equality and Diversity

3.1. This document complies with the Royal Cornwall Hospitals NHS Trust service Equality and Diversity statement.

3.2. Equality Impact Assessment
The Initial Equality Impact Assessment Screening Form is at Appendix 2.
## Appendix 1. Governance Information

<table>
<thead>
<tr>
<th>Document Title</th>
<th>Clinical Guideline for the admission of a child or young person with cystic fibrosis.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Date Issued/Approved:</td>
<td>30 October 2013</td>
</tr>
<tr>
<td>Date Valid From:</td>
<td>30 October 2013</td>
</tr>
<tr>
<td>Date Valid To:</td>
<td>1 October 2016</td>
</tr>
<tr>
<td>Directorate / Department responsible (author/owner):</td>
<td>Dr. A. Prendeville Paediatric consultant</td>
</tr>
<tr>
<td>Contact details:</td>
<td>01872 252017</td>
</tr>
<tr>
<td>Brief summary of contents</td>
<td>Clear guidance on admission procedures for children/young people with Cystic Fibrosis.</td>
</tr>
<tr>
<td>Suggested Keywords:</td>
<td>Cystic Fibrosis Child</td>
</tr>
<tr>
<td>Target Audience</td>
<td>RCHT PCH CFT KCCG</td>
</tr>
<tr>
<td>Executive Director responsible for Policy:</td>
<td>Medical Director</td>
</tr>
<tr>
<td>Date revised:</td>
<td>October 2013</td>
</tr>
<tr>
<td>This document replaces (exact title of previous version):</td>
<td>Cystic fibrosis admissions protocol</td>
</tr>
<tr>
<td>Approval route (names of committees)/consultation:</td>
<td>Audit and guidelines CF team Pharmacy</td>
</tr>
<tr>
<td>Divisional Manager confirming approval processes</td>
<td></td>
</tr>
<tr>
<td>Name and Post Title of additional signatories</td>
<td></td>
</tr>
<tr>
<td>Signature of Executive Director giving approval</td>
<td>{Original Copy Signed}</td>
</tr>
<tr>
<td>Publication Location (refer to Policy on Policies – Approvals and Ratification):</td>
<td>Internet &amp; Intranet Intranet Only</td>
</tr>
<tr>
<td>Document Library Folder/Sub Folder</td>
<td>Paediatrics</td>
</tr>
<tr>
<td>Links to key external standards</td>
<td></td>
</tr>
<tr>
<td>Related Documents:</td>
<td></td>
</tr>
</tbody>
</table>
Training Need Identified?  No

Version Control Table

<table>
<thead>
<tr>
<th>Date</th>
<th>Version No</th>
<th>Summary of Changes</th>
<th>Changes Made by (Name and Job Title)</th>
</tr>
</thead>
<tbody>
<tr>
<td>July 2011</td>
<td>V1.0</td>
<td>Initial Issue</td>
<td>Dr. Prendeville Consultant paediatrician</td>
</tr>
<tr>
<td>October 2013</td>
<td>V2.0</td>
<td>Re format. Minor changes to content wording.</td>
<td>Dr. Prendeville Consultant paediatrician Tabitha Fergus Deputy ward manager</td>
</tr>
</tbody>
</table>

All or part of this document can be released under the Freedom of Information Act 2000

This document is to be retained for 10 years from the date of expiry.

This document is only valid on the day of printing

Controlled Document

This document has been created following the Royal Cornwall Hospitals NHS Trust Policy on Document Production. It should not be altered in any way without the express permission of the author or their Line Manager.
### Appendix 2. Initial Equality Impact Assessment Form

Name of the strategy / policy / proposal / service function to be assessed (hereafter referred to as policy) (Provide brief description): Clinical guideline for the admission of a child/young person with cystic fibrosis.

<table>
<thead>
<tr>
<th>Directorate and service area: Child Health</th>
<th>Is this a new or existing Policy? existing</th>
</tr>
</thead>
<tbody>
<tr>
<td>Name of individual completing assessment: T. Fergus</td>
<td>Telephone: 01872 252800</td>
</tr>
</tbody>
</table>

1. Policy Aim*
   Who is the strategy / policy / proposal / service function aimed at?
   To provide clear guidance on care of a child or young person with Cystic Fibrosis when admitted to hospital.

2. Policy Objectives*
   Evidence based, standardised care.

3. Policy – intended Outcomes*
   “

4. *How will you measure the outcome?*
   Audit and case note inspection

5. Who is intended to benefit from the policy?
   Children and families.

6a) Is consultation required with the workforce, equality groups, local interest groups etc. around this policy?
   no

   b) If yes, have these *groups been consulted?

   C). Please list any groups who have been consulted about this procedure.

7. The Impact
   Please complete the following table.

<table>
<thead>
<tr>
<th>Equality Strands:</th>
<th>Yes</th>
<th>No</th>
<th>Rationale for Assessment / Existing Evidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td></td>
<td>x</td>
<td></td>
</tr>
</tbody>
</table>

Clinical Guideline for the admission of a child or young person with cystic fibrosis.
<table>
<thead>
<tr>
<th>Category</th>
<th>Yes</th>
<th>No</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex (male, female, trans-gender/gender reassignment)</td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>Race / Ethnic communities / groups</td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>Disability - learning</td>
<td></td>
<td></td>
</tr>
<tr>
<td>disability, physical disability, sensory impairment and mental health problems</td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>Religion / other beliefs</td>
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<td></td>
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<tr>
<td>Marriage and civil partnership</td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>Pregnancy and maternity</td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>Sexual Orientation, Bisexual, Gay, heterosexual, Lesbian</td>
<td>x</td>
<td></td>
</tr>
</tbody>
</table>

You will need to continue to a full Equality Impact Assessment if the following have been highlighted:
- You have ticked “Yes” in any column above and No consultation or evidence of there being consultation - this excludes any policies which have been identified as not requiring consultation. or
  - Major service redesign or development

8. Please indicate if a full equality analysis is recommended. | Yes | No | x |
9. If you are not recommending a Full Impact assessment please explain why.

No negative impact.

Signature of policy developer / lead manager / director T. Fergus

Date of completion and submission October 2013

Names and signatures of members carrying out the Screening Assessment

| 1. | 2. |

Keep one copy and send a copy to the Human Rights, Equality and Inclusion Lead, c/o Royal Cornwall Hospitals NHS Trust, Human Resources Department, Knowledge Spa, Truro, Cornwall, TR1 3HD

A summary of the results will be published on the Trust’s web site.

Signed T. Fergus

Date 29/10/13

Clinical Guideline for the admission of a child or young person with cystic fibrosis.  
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