PURPOSE
Physiotherapy role for inpatients with cystic fibrosis.

POLICY STATEMENTS
On admission to hospital all patients will be assessed by the physiotherapist within 24 hours. Physiotherapists have standing orders from Dr. Chilvers to treat all in-patients with Cystic Fibrosis

SITE APPLICABILITY
Inpatients

PRACTICE LEVEL/COMPETENCIES
Advanced Skill - Provides physiotherapy assessments using advanced skill, knowledge and clinical reasoning within assigned area. Applies advanced clinical knowledge and reasoning to the development and implementation of physiotherapy treatment programs in accordance with established standards of the College of Physical Therapists of BC.

DEFINITIONS
Cough swab
Throat swab
Sputum
Cystic fibrosis (CF)

EQUIPMENT
Equipment for airway clearance techniques are purchased by patients and families from the Physiotherapy Department though the CF clinic physiotherapist.
Replacement equipment is located in the physiotherapy department as well as current price list for equipment costs.
Compressors for aerosol medications are purchased through medical supply companies by families with recommendations by CF clinic physiotherapist.

PROCEDURE

<table>
<thead>
<tr>
<th>Rationale</th>
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<td>If patient is admitted for deterioration of chest status, physiotherapy is commenced three times a day. Treatments with the physiotherapist are done two times per day. The third treatment is done in the evening on their own or with family support if needed. Patients are encouraged to perform physiotherapy prior to meals to avoid vomiting.</td>
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1. As respiratory status improves, with decrease in sputum production, physiotherapy may be reduced to twice daily in consultation with physician
2. If patient is admitted for reasons other than deterioration of chest status, physiotherapy is then commenced twice daily in accordance with home routine
3. Mildly affected patients with cystic fibrosis
patients admitted for reasons other than deterioration of chest status may warrant only 1 – 2 daily treatments. This should be done after consultation with the physiotherapist attached to the CF clinic.

4. Sputum samples or cough swabs are collected on admission and then every Sunday during admission. It is the responsibility of the physiotherapist to obtain the samples. If the patient cannot expectorate any mucus a cough swab should be obtained.

5. Weekend physiotherapy treatment for patients with cystic fibrosis patients – refer to Appendix 2

6. Physiotherapy guidelines for treating patients with CF with B. Cepacia - Appendix 3 and Infection control policy IC.06.01 – Cystic Fibrosis

7. If the patient is to have ventolin prior to chest physiotherapy, the nurse is to administer this 15 minutes prior to the treatment time. If the patient is using the PEP mask, then the ventolin may be given during the PEP treatment. The physiotherapist will identify this exception to the nurse and write it on the Kardex. The physiotherapist may draw up the ventolin to give with the PEP treatment.

8. The following types of physiotherapy treatments may be used: Modified Postural Drainage with Percussion and Vibration, Autogenic Drainage, Positive Expiratory Pressure (PEP), Baby PEP, Active Cycle of Breathing or a combination of one or more techniques.

9. The physiotherapist treating the patient should consult with the family or CF clinic physiotherapist if unsure which technique the patient is performing. If the CF physiotherapist is unavailable, then check the patient's Cystic Fibrosis out-patient file located just outside Room K3-202 to ascertain which of the above chest physiotherapy techniques the patient is currently receiving at home.

**TREATMENT REGIMES**

1. **Postural Drainage with Percussion**
   
a) Postural Drainage positions are divided between the different treatment sessions so that each segment of the lung is drained once per day. All postural drainage positions are modified so that no head-down positions are used.
b) If patient is receiving chest physiotherapy three times a day, the postural drainage positions are performed as follows:
   i.) Morning (8-9:30): Anterior, and 2 lateral lower lobe segments.
   ii.) Afternoon (1-3:30): Right middle lobe, left lingular, and posterior segments of lower lobe.
   iii.) Evening (7-9): Apical, anterior, right posterior and left posterior segments of upper lobe.

   These positions may be changed at the discretion of the ward physiotherapist. It is recommended that newly diagnosed patients learning the technique maintain a consistent routine.

c) In each postural drainage position, the patient should be percussed for 3-5 minutes, followed by 3-4 vibrations on expiration and huffing if the patient is able. The patient is then requested to sit up and cough, expectorating any mucous produced. If the patient is quite congested, it may be necessary for the patient to sit up to cough and expectorate in the middle of a treatment position.

d) If a patient is receiving postural drainage with percussion twice daily, the following positions are performed together.
   i.) Morning – 4 lower lobe segments, right middle lobe and left lingular (6 positions)
   ii.) Afternoon – 4 upper lobe segments (4 positions)

2. **Autogenic Drainage**

   a) This technique is generally used in patients over 12 years of age, as patients younger than this generally do not have the concentration to perform this technique properly. However, it may be used in younger patients with the physiotherapist assisting.

   b) Patients are still required to perform 3 treatment sessions per day. Length of each treatment session will depend on the amount of secretions - minimum is 20 minutes, maximum is 45 minutes per session

   c) Autogenic Drainage is performed as per Autogenic Drainage Procedure.

   d) Patients performing Autogenic Drainage should be supervised during each of their treatment sessions by a physiotherapist.
The exception to this is on the weekend where they may only supervise one treatment sessions depending upon the patient’s condition and how well they perform autogenic drainage.

### 3. Positive Expiratory Pressure

| a) | This technique is generally used on patients over 6 years of age. Patients should be taught this physiotherapy technique in consultation with the CF Clinic Team and physiotherapist. |
| b) | PEP is typically completed three times per day. Two sessions are supervised by the physiotherapist. On the weekend, the physiotherapist need only supervise 1 – 2 treatment sessions depending upon the patient’s condition and how well they perform PEP. |
| c) | The positive expiratory pressure technique is performed as per the positive expiratory pressure procedure. |
| d) | The physiotherapist should check that the patient is attaining the correct PEP pressure of between 10–20 cm H₂O. This is done using the manometer. Resistance setting of the PEP device is individually determined and evaluated during CF clinic or during admission. |
| e) | In patients requiring ventolin, it may be given before or during the PEP treatment. |

### 4. Gym

| a) | Gym is held regularly through the week. It is considered part of the cystic fibrosis treatment protocol to improve exercise tolerance. It is always an adjunct to the assigned airway clearance technique. |
| b) | Gym time should be scheduled around IV antibiotics to allow the patient to be locked from IV during this time. |
APPENDIX 1 – Inhaled Medications Administered in Conjunction with Physiotherapy

ORDER OF INHALED MEDICATIONS AND AIRWAY CLEARANCE

ALL MEDICATION MUST BE ORDERED BY A PHYSICIAN.

1. Ventolin/Salbutamol: Bronchodilator, given prior to physiotherapy via metered dose inhaler (MDI) and aerochamber or nebulized with mask or mouthpiece; or given during physiotherapy airway clearance technique via PEP mask. Physiotherapist may request nurse to administer prior to physiotherapy.

2. Hypertonic Saline (HS): mixed with ventolin or given just after ventolin MDI. All patients on hypertonic saline also require ventolin, as HS can cause bronchospasm. Before HS is given to a patient they must have passed a hypertonic saline challenge test. The HS challenge test is completed with pulmonary function tests if the child is old enough or as per HS Challenge protocol (link below). Hypertonic saline is typically given twice a day with the morning and evening physiotherapy treatments. Ventolin is mixed with normal saline if a third treatment of ventolin is required.

3. Airway Clearance Technique (e.g. PEP or percussion and vibration or autogenic drainage): as per procedure outlined above

4. Pulmicort/Steroid: (e.g. Symbicort) to be given to patient after physiotherapy treatment. If a turbo inhaler is used the physiotherapist should ensure correct technique. If given by nebulizer, the nurse is to draw up the solution and either the nurse or physiotherapist to administer to patient.

5. Inhaled Antibiotic/Antifungal: (e.g. Colymycin) To be given after Pulmicort, nurse responsible for administration.

6. Dornase Alfa/Pulmozyme: Nebulized medication, typically given once per day after morning physiotherapy, post inhaled antibiotic, nurse responsible for administration.

NOTE: A Physiotherapist is legally only allowed to draw up and sign for ventolin but may administer medications which are adjuncts to physiotherapy treatments and includes ventolin, pulmicort and pulmozyme but not antibiotics which includes Colymycin. Physiotherapists do not have access to Pyxis for dispensing medications.

NEBULIZERS

The disposable ward nebulizers are to be replaced every 2 days as per hospital protocol. These should only be used for bronchodilator. If a patient requires any other inhaled medication, they are instructed to bring in their own non-disposable nebulizers, as they provide more efficient delivery of medications.

Patients’ own nebulizers are non-disposable and should not be replaced while in hospital. Instead, they need to be rinsed with water after each physiotherapy treatment.

A supply of non-disposable nebulizers (Pari LC Star Nebulizer) will be kept in the POD for the physiotherapist to provide for patients starting inhaled antibiotics during admission.
APPENDIX 2 – Weekend Physiotherapy for Inpatients with Cystic Fibrosis

Physiotherapy services are very limited on the weekend and statutory holidays. Patients with cystic fibrosis are scheduled to be seen by the physiotherapist once or twice daily depending on the acuity of the patient’s condition. For all other physiotherapy treatments, either parents are asked to assist with younger children, or if the patient is an adolescent they are responsible for performing their own physiotherapy treatment.

To assist with weekend treatment the physiotherapist will:

a) On Friday, inform the ward and the patient/parent of treatment times the patient will be seen over the weekend.
b) The physiotherapist will be responsible for collecting sputum on the Sunday.
c) Documentation of respiratory status (i.e. cough and sputum quality) is helpful for Monday CF rounds with weekday therapist.
d) If the physiotherapist is unable to see the patient at the assigned time (i.e. patient asleep/ refuses treatment or out on a pass) and she/he is unable to change treatment times, she/he will inform the nurse of the reason why and document it in the patient’s chart.
e) Physiotherapist on the weekend can be contacted on pager 41-01365 (wards) or 41-02126 (ICU therapist).
f) In order to maximise the use of the physiotherapist’s time with each patient it is beneficial if the RN ensures that their patients are awake and ready for physiotherapy at the time requested.

APPENDIX 3 – Physiotherapy Guidelines for Treating Patients with CF with B. Cepacia Complex

Please see BCCH CF Infection Control Policy (IC.06.01 – Cystic Fibrosis) on Burkholderia Cepacia Complex in patients with cystic fibrosis.

Policy states: “Patients with CF who are infected with bacteria from the Burkholderia cepacia complex shall be separated from other patients with CF (both infected with Burkholderia cepacia complex and culture negative)”.

Instructions are:

1. Any CF patient infected with B. cepacia complex should be placed on separate wards from other CF patients whenever possible.
2. CF patients with B. cepacia should not be cohorted i.e. should be separated from each other.
3. Patients with B. cepacia complex should not use the playroom, gym or classroom. No lag time needs to occur between the exit of one patient and the entrance of another patient to and from one of the above areas.

Specific Physiotherapy guidelines

1. As CF patients infected with B. Cepacia Complex are to be on separate units from other CF patients. If possible the ward physiotherapist on each unit should perform the physiotherapy treatment. If the ward physiotherapist is only treating one CF patient infected with B. Cepacia Complex and no other CF patients, they do not need to gown.
2. Good hand washing should be observed after treating any CF patient.
3. On a weekend or when there is limited staffing a physiotherapist may treat both CF patients with and without B. Cepacia. However, the Physiotherapist should gown and observe good hand washing technique when treating any patients with B. Cepacia Complex. If possible, the patient without B. Cepacia Complex should be treated before the patient with B. Cepacia Complex.

4. To minimise spread of infection, it is recommended that CF patients without B. Cepacia attend the physiotherapy gym session before the CF patients with B. Cepacia wherever possible.

RELATED DOCUMENTS

Please refer to other related policies/documents:
Hypertonic Saline Challenge in Infants with CF
Physiotherapy: Definition of a Cough Swab
Physiotherapy Patient Handout: PEP Mask
Physiotherapy Patient Handout: Autogenic Drainage
Physiotherapy Patient Handout: Percussion and Vibration Teaching for New Diagnosis (newborn)
Physiotherapy Patient Handout: Percussion and Vibration Teaching for New Diagnosis (older child)
Physiotherapy: Obtaining Sputum from a Child with CF on the Weekend
CF Infection Control Policy Reference Sheet

DOCUMENTATION

As per College of Physical Therapists of BC practice standards

REFERENCES

1. BC’s Children’s Hospital, Infection Control Policy IC.06.01 – Cystic Fibrosis
2. Canadian Cystic Fibrosis Foundation B. Cepacia Complex Policy.