Cystic Fibrosis

Care Guidelines for Challenging Cystic Fibrosis

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This document is based on a consensus of evidence and/or clinical expert opinion.

Adapted from Balfour-Lynn, I. M. (ed.) (2017). Clinical Guidelines: Care of Children with Cystic Fibrosis. *Royal Brompton Hospital. 7th edition. London: Royal Brompton Hospital.*

It is intended that this Care Pathway be reviewed and revised as needed by April 2020.

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Table of Contents

General Age Range Guide

Infant	Preschool	Child	Adolescent	Adult
0 to 2 years	2 to 6 years	6 to 12 years	12 to 18 years	≥ 18 years

Abbreviation Guide

ABPA . . . . Allergic bronchopulmonary aspergillosis
BMI . . . . . Body Mass Index
CF . . . . . Cystic Fibrosis
CFRD . . . . Cystic Fibrosis-Related Diabetes
CT . . . . . Computerized Tomography
CXR . . . . Chest X-ray
ENT . . . . . Ear, Nose, Throat specialty
GERD . . . . Gastroesophageal Reflux Disease
GI . . . . . Gastrointestinal specialty
IgE . . . . . Immunoglobulin E
LCI . . . . . Lung Clearance Index
MDR . . . . Multi Drug Resistant
MRSA . . . . Methicillin-resistant Staphylococcus aureus
MSK . . . . Musculoskeletal
NTM . . . . non-tuberculous mycobacteria
PA . . . . . Pseudomonas aeruginosa
PCTL . . . . Percentile
PERT . . . . Pancreatic Enzyme Replacement Therapy
PEx . . . . . Pulmonary Exacerbation
RAST . . . . Radioallergosorbent test

Introduction

Cystic Fibrosis Standard Care Guidelines for BC recommend patients have a CF clinic review at least 4 times per year. This review includes pulmonary function testing and assessment of BMI.

When the patient's lung function decline is more rapid than expected or the patient negatively crosses BMI percentiles, intensified clinical review is advised to identify potential causes and to direct treatment strategies. This includes activation of this protocol.

Clinical Features

Patients (both pediatric and adult) who present with any of the following require an intensified evaluation by a multidisciplinary CF Team:

- 10% decline in spirometry from best in previous year and not responding to treatment
- 3 or more courses of intravenous antibiotics annually
- Requires home oxygen therapy
- Nutritional failure
  - 0 to 24 months weight for length < 15th PCTL
  - 2 to 18 years BMI < 15th PCTL
  - Adults BMI < 20 (kg/m2) or BMI Z-score < 2
- Severe pulmonary exacerbation (PEx) such as large volume hemoptysis, pneumothorax, therapy resistant allergic bronchopulmonary aspergillosis (ABPA) or other cause of steroid dependency
- Patient says they are feeling well, but the objective findings indicate that they are not well
- Challenges with adhering to treatment

Possible Causes for Decline

The most likely causes of a rapid lung function decline or a decrease in BMI are:

a. Not adhering to the therapy

b. Infectious respiratory organism such as Pseudomonas aeruginosa (PA), Methicillin-resistant Staphylococcus aureus (MRSA), non-tuberculous mycobacteria (NTM), or other multi-drug resistant (MDR) pathogen

c. CF-related diabetes (CFRD)

d. Nutritional failure due to CFRD, malabsorption, bacterial overgrowth syndrome

e. Gastrointestinal Reflux Disease (GERD)

f. Allergy related issue such as asthma, ABPA

g. Chronic pain

h. Reduced or minimal physical activity

i. Comorbidity of chronic sinusitis

j. Psychological issues such as depression, anxiety, substance use

k. Access to or continuity of care issues

Assessment / Investigations

a. Non-adherence

- Review PharmaNet records – Are medications being picked up?
- Conduct a meeting with patient and/or family outside of regular clinic visits to discuss challenges and barriers (might involve counsellors with expertise in motivational interviewing):

Access and Funding barriers

- Worries about finances?
- Issues with pharmacy?
- Registered for Fair Pharmacare?
- Taxes completed last year (needed to renew Fair Pharmacare)?
- All special authorizations up to day?
- Any problems with extended health benefits plan coverage such as a lifetime cap?
- Ability to pay for medications?
- Aware of BC CF medication grant?

Adherence barriers

- Understanding of CF disease and what is needed for self-management? Rationale for medications? Anticipated outcomes?
- Airway clearance techniques? Difficulty doing? Time or lack of time? Perceived effectiveness? Treatment beliefs?
- Beliefs and preferences about therapy?

- Concerns about treatment burden? Cost of medications?
- Proper storage of medications?

Equipment barriers

- Able to pay for equipment? Access to funding sources?
- Appropriate equipment? Correct make and model? Correct nebulizer?
- Using equipment correctly?
- Replacing nebulizer regularly?
- Equipment cleaned correctly and regularly?
- Challenges to accessing and using new technology (such as e-flows)?

b. Respiratory

- Reassess sputum for PA, MDR bacteria (MRSA etc.), NTM, or fungi.
- If sputum is difficult to obtain, consider bronchoscopy for airway lavage.
- Perform CXR to rule out structural abnormalities. Consider CT chest imaging for best assessment of lung parenchyma.
- Ensure all chronic airway therapies are being used including:
  - Hydration therapy using hypertonic saline
  - Mucolytic therapy with Dornase alfa (Pulmozyme)
  - Inhaled antibiotics (for PA colonization)
- Ensure being treated with chronic macrolide antibiotic therapy (confirm that NTM has been ruled out).
- Repeat lung function tests with bronchodilator reversibility.
- Additional tests might include Lung Clearance Index (LCI), exercise testing, overnight O2 saturation, and transcutaneous carbon dioxide (TcCO2).

c. CF Related Diabetes

- Check fasting serum glucose levels along with repeat glucose tolerance testing, HbA1C.
- Consider home glucometer testing and record keeping multiple times per day.

d. Nutritional failure

- For low BMI, test for malabsorption and stool fat content. Reassess PERT compliance.

- If diarrhea prominent, check gastric emptying. Rule out celiac disease (baseline IgA and anti-TTG).
- Consider referral to Gastroenterology.

e. GERD

- Consider esophageal manometry or pH study.

f. Allergy related issues

- If aspergillus present, assess for ABPA.
  - Check IgE level, Aspergillus serum precipitins, Aspergillus-specific IgE, Aspergillus skin hypersensitivity testing, and/or radioallergosorbent test (RAST).
- Consider possibility of asthma.
  - Get pre and post bronchodilator spirometry or perform methacholine challenge.
  - Consider skin aeroallergy testing, particularly if airway hyper-responsiveness seen on pulmonary function tests.

g. Chronic pain

- Physiotherapist to perform a musculoskeletal assessment. Refer to a musculoskeletal (MSK) physiotherapist as needed.
- Determine need for analgesics.
- Refer to a chronic pain clinic as needed.

h. Minimal physical activity

- Physiotherapist to perform an incremental exercise test and determine level of fitness and barriers to exercise.

i. Sinusitis

- Review with sinus imaging (x-ray or CT).
- Consider ENT referral for nasopharyngoscopy.

j. Psychosocial issues

- Screen for anxiety and/or depression (General Anxiety Disorder-7, Patient Health Questionnaire-9). If score high, consider psychiatry assessment if possible.

Mental Health barriers

- Emotional Wellness Screening done? Up to date?
- Supports in place?
- Access to mental health services?
- Possibility of alcohol/narcotics/substance abuse or eating disorder?

k. Access or continuity of care issues

- If not regularly attending CF clinic (minimum 4 times/year):
  - Reason for non-attendance?
  - Lives locally or far from clinic?
  - Offered CF outreach appointments?
  - Offered CF telehealth appointments?
  - Aware of CF travel grant and BC Residency Program?
  - Aware of other travel assistance to clinic/hospital? (e.g. BC Ferries Medical Travel Assistance Program (TAP)/Medical Assured Loading, Hope Air, other?)
- If patient is newly transferred (transitioned) to adult healthcare:
  - Does adult clinic have appropriate medical history/detailed transfer summary from pediatric clinic?
  - Is patient attending (and engaged with) the adult clinic?
- If involved with other sub-specialists/specialized clinics (Endocrine, Pain Clinic, ENT, GI, Infectious Diseases):
  - Access Issues?
  - Has patient been referred?
  - Has appropriate follow-up occurred?
- If patient have been hospitalized:
  - Admitted locally or at CF centre?
  - Home IV?
  - Appropriate length of IV antibiotic treatment?

Interventions and Ongoing Monitoring

- Generally, patients with complicated CF disease should be seen on a more frequent basis, particularly to review diagnostic test outcomes and results from empiric therapeutic trials.
- Might need to consider rotating inhaled antibiotics, or oral outpatient antibiotics, and schedule regular IV antibiotic admissions.
- Need to review all of the investigative aspects with the full multidisciplinary CF Team and formulate an action plan with family.

End Notes

1 Balfour-Lynn, I. M. (ed.) (2017). Clinical Guidelines: Care of Children with Cystic Fibrosis. *Royal Brompton Hospital. 7th edition. London: Royal Brompton Hospital.*