SETTING THE STAGE
TIERS IN BRIEF
TIERS IN FULL

# TIERS OF CYSTIC FIBROSIS SERVICE IN BC

# (ADULTS AND CHILDREN)

Note: May 2024
Some terminology is changing in response to ongoing provincial Tiers of Service work. The previous Child Health BC "modules" are now called "companion guides," to emphasize their focus on operational and service planning considerations, such as responsibilities for pediatric care delivery, training, and quality improvement. Updates to this document are forthcoming.

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# Tiers of Cystic Fibrosis Services in BC (Adults and Children)

January 22, 2019

## Contents



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We encourage you to share these documents with others and we welcome their use as a reference. Please cite each document in the module in keeping with the citation on the table of contents of both of the documents. If referencing the full module, please cite as:

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# 1.0 Tiers of Service Framework and Approach (Context)

Utilizing a common language and methodology, the Tiers of Service framework:

- Recognizes that health services, while important, are one of several factors that contribute to overall well-being.
- Is informed by a review of frameworks/tools in other jurisdictions around the world.
- Facilitates system planning for clinical services, knowledge sharing/training and quality improvement/research. The responsibilities and requirements for each of these three areas are defined within the Tiers framework.

The Tiers of Service approach to system planning occurs through:

- Creation of a series of modules.
- Self-assessment based on the modules.
- System and service planning based on self-assessment results (provincial, regional and local planning).

Utilization of a Tiers of Service approach to planning Cystic Fibrosis (CF) services in British Columbia (BC) will help to strengthen the existing provincial system of CF services. Through collaborative efforts, variations in CF services can be reduced and the consistency and continuity of care provided to people (and their families) with CF across the province can be strengthened.

# 2.0 Setting the Stage for Development of Tiers of CF Services Module (Background)

## *2.1 Cystic Fibrosis*

CF is the most common, fatal genetic disease affecting Canadian children and young adults.[1] It is estimated that 1 in every 3,600 children born in Canada has CF.[2] In 2016, there were 436 unique patients seen in BC (from data collected on participating patients in the Canadian CF Registry). This number has increased to 518 patients in 2018 according to the numbers registered with the four specialty clinics in BC.

At present, there is no cure for CF which affects multiple organ systems but has its major impact on the lungs and the digestive system. While the severity of CF differs from person to person, in the majority, persistent and ongoing lung infection results in progressive destruction and loss of lung function, eventually leading to death.

Typical complications caused by cystic fibrosis include:[1]

- Difficulty digesting fats and proteins
- Malnutrition and vitamin deficiencies due to inability to absorb nutrients
- Progressive lung damage from chronic infections and aberrant inflammation
- CF related diabetes
- Sinus infections
- Osteopenia
- Liver disease
- Fertility issues
- Mental health challenges

Strategies to prevent or treat bacterial infections remain the key course of action to sustain patients' health status. In addition to antibiotics, patients must follow a daily routine of physical therapy to keep the lungs as free of congestion and infection as possible. Multiple medications are required to sustain health. Lung transplant is a possible option for those patients deemed suitable for transplant and for whom a transplant lung becomes available. Lung transplants for patients with CF living in BC are performed at Vancouver General Hospital (adults and older children) and Stollery Children's Hospital in Edmonton, Alberta (younger children).

Studies measuring psychological distress in individuals with cystic fibrosis have found high rates of both depression and anxiety. Psychological symptoms in both individuals with CF and parent caregivers have been associated with decreased lung function, lower body mass index, worse adherence, poorer health-related quality of life more frequent hospitalizations and increased healthcare costs. The International Committee on Mental Health in Cystic Fibrosis has recently published recommendations for screening and treatment of anxiety and depression in persons with CF.[3]

For persons with CF, life is a demanding daily routine of therapy and regularly scheduled visits to a CF clinic. Despite these demands, most individuals with CF lead productive lives in terms of education, physical activity, and social relationships. Eventually, however, respiratory and other disorders place increasing limits on daily life. Thanks to advances in research, clinical care and newborn screening, growing numbers of children with CF are surviving into adulthood. In Canada, the estimated median age of survival has increased from 26 years in 1986 to 53 years in 2016.

## *2.2 Cystic Fibrosis Services in BC*

Children and adults with CF receive specialized interdisciplinary care at one of 4 CF clinics in BC, each of which is within a hospital setting, has a university affiliation (University of British Columbia) and is accredited and regularly reviewed by CF Canada.

CF patients are seen by various healthcare professionals at each clinic visit and during inpatient hospital stays. The goal is for patients to attend clinic 4 times per year (more if unstable or in the pre-transplant phase) and to spend time with each team member as needed at each visit. Team members include a CF physician specialist (usually a respirologist), nurse, dietitian, respiratory therapist, physiotherapist, social worker, pharmacist and, ideally, a psychologist and/or psychiatrist. CF is a complex disease and requires treatment throughout a patient's life. In addition to the care provided by the CF Clinic team, patients are expected to have a primary care provider who is available to provide routine, non-CF related care.

With increases in the life expectancy of patients with CF, we now have more adults than children with CF in BC (the same is true in the rest of Canada). As patients age, the complexity of their disease invariably increases and this is compounded by age-related comorbidities previously not seen in people with CF. This changing demographic is compounded by the daunting task of

delivering equitable care across huge geographical distances with a broad urban and rural divide. Access to CF care is a challenge for individuals who live outside Metro Vancouver and Victoria.[5] However, the level of multidisciplinary expertise required to address the full range of CF care is such that it is not realistic to move to full-fledged CF Clinics in all HAs. Therefore, with increasing numbers of patients with CF, there is a need to re-look at the service delivery model and to broaden the availability of CF care across the province through partnerships with local healthcare providers and by using the latest in technology.[6] The existing CF Clinics will provide a foundation upon which to expand existing services and the Tiers of Service provides a forum for these discussions to occur.

## *2.3 BC Data: Patient Profile and Service Utilization*

The Canadian Cystic Fibrosis Registry (CCFR) provides a collection of national CF data. Participating CF patients who attend any of the accredited 42 CF clinics across Canada and provide consent are represented in this database. Given that the majority of CF patients attend one of these clinics and provide consent, the CCFR contains data on more than 90% of all Canadians diagnosed with CF.[4]

### 2.3.1 Patient Profile

#### 2.3.1.2 Numbers, ages and co-morbidities of patients with CF in BC

To provide a profile of people in BC living with CF, data was obtained from the CCFR for patients seen at the four BC CF clinics between 2010 and 2016 (the last year for which complete data is available). Refer to Table 1. Of note, there has been a 22% increase (358 to 436) in the number of unique patients care for in BC in the 7 year period (inclusive).

**Table 1: Patient Profile of People with CF in BC**

| Indicator | 2010-2016 | 2010 | 2016 | % Change, 2010 - 2016 |
|---|---|---|---|---|
| Number of unique patients with CF | | 358 | 436 | 21.8% |
| Number diagnosed by NBS in BC (since inception in 2009) | 69 (Average ~9/yr.) | | | |
| Number diagnosed as adult (≥18) | 53 (Average ~8/yr.) | | | |
| Median age of pts with CF | | | 24.25 yrs. 63.3%≥18yrs; 18%≥40yrs; 3.4%≥60yrs | |
| Transplants | 75 (Range 5-8/yr.) Lung 69; Liver 5 | | | |
| Deaths | 28 (Range 0-7/yr., almost all in adults) | | | |
| % with CF related diabetes | 25.6% | | | |

Sources: CF Profile: CCFR BC Report & Provincial Trend Analysis: 2017 (unpublished, Sameer Desai)

#### 2.3.1.3 Home Residence of Patients with CF in BC

The distribution of patients with CF across HAs is proportionate to the number of people living in each HA. See Table 2.

Table 2: Home Residence of People with CF in BC

| HA of Home Residence | Patients with CF | % Prov CF Total | % Prov Population in HA |
|---|---|---|---|
| Interior HA | 75 | 17% | 16% |
| Fraser HA | 163 | 38% | 37% |
| Van Coastal HA | 105 | 24% | 25% |

| HA of Home Residence | Patients with CF | % Prov CF Total | % Prov Population in HA |
|---|---|---|---|
| Van Is HA | 70 | 16% | 16% |
| Northern HA | 22 | 5% | 6% |
| Total | 435 | 100% | 100% |

Source: C Quon, B and Desai, S, GP Survey Data (Updated), March 20, 2017 (unpublished)

#### 2.3.1.4 Ethnicity

The vast majority of patients with CF in BC are Caucasian and given the genetics of CF this pattern is not anticipated to change significantly. See Table 3.

Table 3: Ethnicity of People with CF in BC

| Ethnicity | % |
|---|---|
| Asian | 0.95 |
| Caucasian | 94.08 |
| First Nations People | 0.95 |
| Hispanic | 0.19 |
| Other | 1.15 |
| South Asian | 1.53 |
| Two or more races | 0.19 |
| Not reported | 0.95 |

### 2.3.2 Service Utilization Data

#### 2.3.2.1 CF Clinics

Table 4 provides details on the activities within the 4 CF Clinics in BC. Of note is that between 2010 and 2016 there was a 22% increase in the number of patients, while the number of clinic visits increased by 51%. This difference is explained by the increasing complexity of patients related to the expansion of the NBS program, increased co-morbidities, the availability of new, expensive treatments and an increasing number of patients receiving organ transplants. With the steady growth of the CF population in BC, continued growth is expected.

Table 4: CF Clinics in BC (Pediatric and Adult)

| Focus | Clinic | Source of Referrals | Caseload | | Clinic Visits | | Frequency of Clinics | Outreach Clinics | Graduation/ Transition Clinics |
|---|---|---|---|---|---|---|---|---|---|
| | | | Case-load 2016 | % Growth, 2010 - 2016 | Visits/ Yr | % Growth, 2010 - 2016 | | | |
| Child-ren | Victoria General | Within HA | 25 | 19% | | | 2 clinic days (full days)/mo (1 day/mo is for children under 2 years) | | Royal Jubilee (orientation for transition patients) (2x/yr) |
| | BC Children's | Provincial | 148 | 14% | | | 6 clinic days (4 full days, 4 half days)/mo (the 4 half days are for children under 2 years) | Kelowna (3 x/yr, 1 - 1.5 days/visit)<br><br>Prince George (2-3 x/yr) | Graduation (transition) clinics: 2 - 3x/yr held at BCCH (involves staff from both BCCH and SPH). |
| Adults | St Paul's Hospital | Provincial | 224 | 20% | | | 12 clinic days (full days)/mo | Kelowna (3 x/yr)<br><br>Prince George (2 x/yr) | See BCCH above |
| | Royal Jubilee | Within HA | 47 | 62% | | | 2 clinic days (full days)/mo | | |
| **Total** | | | **444 (Note 1)** | **22%** | **1,931** | **51%** | | | |

Note 1: 8 patients were reported by more than one clinic in 2016.

Source: Canadian CF Registry, 2016

#### 2.3.2.1 Hospitalizations

Table 5 provides details on the number of hospitalizations of patients with CF in BC, the number of unique patients hospitalized and the number of hospital days. Of note is that despite the 22% increase in patients between 2010 and 2016, the number of hospitalizations, unique patients hospitalized and hospital days decreased by 5%, 2% and 9% respectively. These reductions in the need for inpatient hospital care reflect the significant advances that have been made in CF care and the 51% increase in outpatient visits provided by the CF clinics. Continuation of these trends, as the patient population grows and complexity of care increases, will require significant additional resources.

Table 5: Hospitalizations, Patients of all ages with CF Diagnosis, 2010 - 2016 (Source: Canadian CF Registry)

| Indicator | 2010 | 2016 | % Decrease, 2010 - 2016 |
|---|---|---|---|
| Hospitalizations (70.4% of admissions were for pulmonary exacerbations, 2015/16) | 243 | 230 | -5% |
| Unique patients hospitalized | 137 | 134 | -2% |
| Hospital days | 3,474<br>Avg. 14.3 days/hospitalization | 3,160<br>Avg. 13.7 days/hospitalization | -9% |

Source: Canadian CF Registry, 2016

#### 2.3.2.2 Home Intravenous Therapy Utilization Data

Table 6 shows the number of patients on home intravenous (IV) therapy, as well as the number of home IV courses and days. Of note is that while the total number of courses has not increased, the number of patients needing home IV treatment has and the length of treatment courses has been progressively increasing from an average of 16 days in 2010 to 21.7 days in 2016, a 35.6% increase. This reflects the increasing complexity of infections in patients with CF. There is strong demand from patients and their families for improved access to regional home IV services. Facilitating this demand will help provide patients with care closer to their home and help to further reduce the hospitalization rates.

Table 6: Home Intravenous (IV) Therapy, Patients of all ages with CF Diagnosis, 2010 - 2016 2016/17 (Source: Canadian CF Registry)

| Year | Unique Patients | Home IV courses | Home IV days |
|---|---|---|---|
| 2010 | 44 | 88 | 1,407 |
| 2011 | 50 | 100 | 1,396 |
| 2012 | 45 | 86 | 1,155 |
| 2013 | 43 | 63 | 1,210 |
| 2014 | 39 | 61 | 920 (see note) |
| 2015 | 41 | 63 | 1,327 |
| 2016 | 55 | 84 | 1,824 |
| **% Change, 2010-2016** | **25%** | **(4%)** | **30%** |

Note: Data is available only for 2 of the 4 clinics.

# 3.0 Development of Tiers of CF Service Module (Process)

## *3.1 Module Development*

Development of the module was led by the Provincial Strategic Initiatives area within the Provincial Health Services Authority (PHSA) and Child Health BC (CHBC). A small working group utilized the CF Standards of Care for BC[6] as well as other provincial and national documents and published literature to develop this *Tiers of Cystic Fibrosis (CF)* module. Development of the CF Standards of Care for BC involved a broad range of interdisciplinary CF providers and other experts and included a detailed search of the literature. The goal of this document is to translate the "care standards" into "service responsibilities and requirements" for the purposes of program planning.

Input into the draft module was provided by members of the Provincial CF Advisory Committee. This committee is comprised of representatives from each of BC's Health Authorities, the 4 CF Clinic medical directors (Respirologists and Pediatricians), a CF Clinic nurse coordinator, a family physician, directors/managers of CF/adult/pediatric and allied health services, Ministry of Health, CF Canada and a patient representative.

The final version was accepted by the Provincial CF Advisory Committee (November 30, 2018) and the Child Health BC Steering Committee (the latter from a pediatric perspective, December 7, 2018).

## 3.2 Module Scope

Includes:

- Outpatient services (hospital or community-based) that provide care to adults and children with CF in BC.
- Inpatient services that provide care to adults and children with CF in BC.

Excludes:

- General medical/surgical services provided to children[i] or adults in hospital or as outpatients that is not CF-specific.
- Emergency department services provided to children[ii] or adults that is not CF-specific.

## 3.3 Recognition of the Tiers

Because of the highly specialized nature of CF services and the relatively small volume of patients, CF services in BC are recognized primarily as Tier 6 (T6) services. While the 4 T6 services that currently operate in BC function relatively independent of each other, the future vision is to move toward a single Provincial CF Program which provides its services in four locally-based clinics (Vancouver and Victoria) with expanded models of care such as outreach clinics (CF Team provides "visiting" clinics at "host" site), virtual clinics (CF Team provides services using telehealth, etc.) and collaboration with centres/physicians who have an interest and appropriate resources to provide selective aspects of CF care. This module assumes this future vision.

[i] Refer to the Children's Medicine Services module at http://childhealthbc.ca/tiers-service/childrens-medical-services for services provided to children that are not specific to CF (note: a comparable model for adults has not yet been developed).

[ii] Refer to the Children's Emergency Department Services module at http://childhealthbc.ca/tiers-service/childrens-emergency-department-services for services provided to children that are not specific to CF (note: a comparable model for adults has not yet been developed).

The T6 CF services are supported by general service providers at T1 (outpatients) and T4 (inpatients). See Table 7. T1 and T4 services are grayed out because they are *general* and not *CF-specific* services. They are included in this module to show the continuum of services.

Table 7: CF Tiers of Service

| Tier | Description | Service | |
|---|---|---|---|
| | | Outpatient | Inpatient |
| 1 | Prevention and Primary Health Service | ✓ | |
| 2 | General Health Service | | |
| 3 | Focused Health Service | | |
| 4 | Comprehensive Non CF-Specific Inpatient Service | | ✓ |
| 5 | Regional Subspecialty Service | | |
| 6 | Provincial Subspecialty CF Service | ✓ | ✓ |

# 4.0 CF Tiers of Service (Description, Responsibilities and Requirements)

This module is organized into the following sections:

4.1 Clinical Services
- 4.1.1 Outpatient Services: Service reach and description; responsibilities; requirements
- 4.1.2 Inpatient Services: Service reach and description; responsibilities; requirements

4.2 Knowledge Sharing and Transfer/Training

4.3 Quality Improvement & Research

## *4.1 Clinical Services*

### 4.1.1 Outpatient Services

T1 is grayed out in the following charts because it is a *general* and not *CF-specific* service. It is included in this module to show the continuum of services.

#### 4.1.1.1 Service Reach and Description

| | Prevention & Primary Health Service | Provincial CF Subspecialty Service |
|---|---|---|
| | T1 | T6 |
| Service reach | Local community. | Provincial. |
| Service description | Care provided by a local care provider with general knowledge about CF. e.g., family physician, nurse practitioner, pediatrician, adult respirologist.<br><br>Service typically includes:<br>• General health maintenance & non-CF related care (e.g., vaccinations, musculoskeletal injuries, birth control counselling, care during pregnancy, etc.)<br>• Supportive & CF-related care, upon direction & with support from the patient's T6 CF Team or as outlined in the CF care plan (e.g., colonoscopy planning, management of pulmonary exacerbations, monitoring of patients on home IV therapy).<br><br>All CF patients regardless of where they live are expected to have a local primary care provider (PCP). | CF care provided by a specialized interdisciplinary CF Team to children/adults living throughout the province. The practice of individual team members aligns with the population served (i.e., practice of a Pediatric CF Team is exclusively or predominantly with children; vise versa for Adult CF Team).<br><br>Patients often have significant & multiple medical co-morbidities which require the services of multiple specialists/subspecialists.<br><br>The goal is for patients to be seen by a CF Team 4 times/yr throughout their life (more frequently if required). Visits may occur at a locally-based clinic (Vancouver and Victoria), outreach clinic (CF Team provides "visiting" clinics at "host" sites) &/or performed virtually (CF Team provides services using telehealth, etc.). |

### 4.1.1.2 Responsibilities

| | Prevention & Primary Service | Provincial CF Subspecialty Service |
|---|---|---|
| | **T1** | **T6** |
| CF screening & diagnosis | All ages:<br><br>PCPs:<br>• Refers children with clinical suspicion of CF to local pediatrician or T6 Pediatric CF Team for testing & follow-up.<br>• Refers adults with clinical suspicion of CF to local respirologist or T6 Adult CF Team for testing & follow-up.<br><br>Pediatrician/respirologist:<br>• Arranges for initial CF testing (sweat chloride test).<br>• Refers patients with positive or inconclusive CF test results to T6 Pediatric/Adult CF Team.<br><br>PCPs & pediatrician/respirologist:<br>• Supports patients/families throughout the screening & diagnosis process (providing education, psychosocial support, etc.) | Newborns:<br>• Oversees the provincial CF newborn screening program (NSP) which includes screening for CF.<br>• Contacts families of newborns with positive or inconclusive NBS CF blood test results. Arranges for initial assessment & sweat chloride testing.<br>• Conducts initial assessment & sweat chloride testing for newborns with positive or inconclusive NBS CF blood test results.<br><br>All other ages:<br>• Conducts initial assessment & arranges sweat chloride testing and CF genetics for patients referred from throughout the province with clinical suspicion of CF.<br>• Contacts patients/caregiver & PCP to review the results of the assessment & testing. Provides support, as required.<br><br>All ages:<br>• Performs & directs urgent & non-urgent constitutional genetic testing for the province. Arranges for extended genetic testing (CFTR) as necessary. For details, refer to Lab Tiers module at http://childhealthbc.ca/tiers-service/laboratory-pathology-transfusion-medicine-services. |
| Initial interdisciplinary assessment, education & management | | All ages:<br>• Conducts initial interdisciplinary assessment.<br>• In collaboration with the patient/family, develops & facilitates a plan of care.<br>• Provides treatment according to the guideline CF: Standards of Care for BC (2018). Typically includes:<br>  • Medical management<br>  • Infection prevention & management<br>  • Nutrition management<br>  • Airway clearance, exercise & inhalation therapy<br>  • Monitoring lung function<br>  • Education about CF & related issues<br>• Supports self/family management of CF (e.g., travel, sick day management, etc.)<br>• Refers & coordinates care with other subspecialty teams, as required.<br>• Provides regular updates to the child/adult's PCP. |

| | Prevention & Primary Service | Provincial CF Subspecialty Service |
|---|---|---|
| | **T1** | **T6** |
| | | • Helps to gain access to medications including Pharmacare Plan D, and through Pharmacare special authorization, other third party payments and non-formulary drugs. e.g., Health Canada. |
| Ongoing assessment, education & management | All ages:<br>• Provides general health maintenance & non-CF related care (e.g., vaccinations, musculoskeletal injuries, birth control counselling, care during pregnancy, etc.)<br>• Provides supportive & CF-related care, upon direction & with support from the patient's T6 CF Team (e.g., colonoscopy planning, management of pulmonary exacerbations, monitoring of patients on home IV therapy).<br>• Communicates changes in child/adult's condition &/or treatment plan to T6 Pediatric/Adult CF Team. | All ages:<br>• At each visit, assesses the well-being of the patient/family including:<br>  • Physical wellbeing (lung function, sputum, GI functioning)<br>  • Key indicators (FEV1, BMI)<br>  • Emotional status<br>  • Developmental stage<br>  • Adherence to prescribed therapies<br>  • Comorbidities & complications<br>• At least once/yr, conducts detailed review of the patient's status. Works with the patient/family to update the plan of care.<br>  • Arranges diagnostic & other testing at the intervals outlined in the guideline CF: Standards of Care for BC (2018). Takes action as required. For specifics, refer to applicable CF Guidelines for CF-Related Abdominal Pain, CF-Related Bone Disease, CF-Related Diabetes, Challenging CF, Pulmonary Exacerbations, New Acquisition of Pulmonary Bacteria, Nutrition Management & Screening & Treating Depression & Anxiety.[7-14]<br>• Refers patient/family to community resources/specialists/subspecialists, as required (subspecialists may be co-located or at T6).<br>• Provides regular updates (i.e., after reviews & inpatient admissions) to the patient's PCP on the status of the patient's CF treatment plan.<br>• Develops plan for ongoing monitoring. Where appropriate, works collaboratively with T1 providers to offer some aspects of CF care in the patient's local community (children/adults living in geographic areas outside the lower mainland & Victoria).<br>• Manages co-morbidities and CF-related complications and facilitates referral to designated subspecialty services.<br>• Provides regular updates to the child/adult's PCP.<br><br>Children:<br>• Supports children entering the school system who qualify for a Chronic Health Designation (applies to most CF pts who take pancreatic enzymes).<br>• Assist parents and school with creation of school care plan and additional information as requested. |

| | Prevention & Primary Service | Provincial CF Subspecialty Service |
|---|---|---|
| | **T1** | **T6** |
| Care planning | | All ages:<br>• In collaboration with patients/families, develops customized patient care plans. Updates annually and as required. Updates Keeps PCP updated of changes. |
| Care coordination | All ages:<br>• As directed & with support from the patient's T6 CF Team, arranges for tests, referrals, treatment etc locally. Keeps CF Team updated. | All ages:<br>• Acts as a link between the patient/family with primary & community services & the CF Clinic/hospital.<br>• Liaises with clinical, social, educational, employment & other community agencies, as required.<br>• Works with patients/families to develop strategies to navigate the health care, school & community systems.<br>• Arranges interdisciplinary CF Team and patient meetings at regularly scheduled intervals. |
| Nutrition counselling | All ages:<br>• Reinforces the nutrition-related information/teaching provided by the CF Team.<br>• Consults with CF Team for nutrition-related questions/issues. | All ages:<br>• At each clinic visit, assesses the patient's nutritional status, including review of nutritional intake, pancreatic enzyme status & absorption, management of metabolic complications & growth & nutritional status.<br>• At least once/yr, conducts a thorough dietary & nutritional assessment/review which may include a 3 day intake.<br>• Uses assessment findings to develop/update a specialized nutrition care plan.<br>• Provides nutrition education to patients/families. Education is age-specific & focuses on food choices & strategies to meet nutrition needs & promote adequate growth.<br>• Develops specific nutrition management plans for failure to thrive, pancreatic insufficiency, micronutrient deficiency, delayed gastric emptying, reflux, constipation & CF-related conditions such as distal intestinal obstruction syndrome, impaired glucose tolerance, CF related diabetes & bone disease.<br>• Provides information to patients/families about community-based nutrition resources, including tube feeding, access to food and culturally relevant resources. |

| | Prevention & Primary Service | Provincial CF Subspecialty Service |
|---|---|---|
| | **T1** | **T6** |
| Respiratory support | All ages:<br>• Reinforces the respiratory-related information/teaching provided by the CF Team.<br>• Consults with CF Team for respiratory/activity-related questions/issues. | All ages:<br>• At each clinic visit, assesses the patient's respiratory status, including respiratory signs & symptoms (cough characteristics, airway clearance technique), timing of inhalation therapy & technique, posture & musculoskeletal development, activity level & adherence with treatment plan.<br>• At each clinic visit, conducts procedures to measure lung function.<br>• At least once/yr, conducts a complete respiratory assessment including exercise and activity questionnaire and, if available, sub-maximal exercise testing.<br>• In collaboration with the patient/family, develops a respiratory treatment plan to maximize lung function (airway clearance, exercise & inhalation therapy). Considers age, disease severity, physical side effects or complications, social & domestic circumstances.<br>• Provides treatment or refers to external physiotherapy/respiratory specialists when needed. |
| Psychosocial support | All ages:<br>• Identifies patients with depression/anxiety.<br>• Provides ongoing treatment & management for patients with CF (provider identified &/or referred from CF Clinic).<br>• Provides ongoing psychosocial support for patient/family.<br>• Consults with CF Team for psychosocial-related questions/issues. | All ages:<br>• At each clinic visit, conducts brief psychosocial assessment. Screens the following annually for depression and anxiety using the PHQ-9 and GAD-7:<br>  ◦ Patients aged 12 and older<br>  ◦ Parent caregivers of children aged birth to 17 years<br>• Carries out clinical assessment for depression and anxiety when symptoms are reported or screening scores are elevated.<br>• Develops and implements treatment plan in close collaboration with the patient and caregiver and the patient's primary care provider. Refers to MH specialist(s) as required.<br>• For specifics, refer to CF Care Guidelines for Screening & Treating Depression & Anxiety.[14]<br>• Provides social, emotional & practical support at disease stages (e.g., CF diagnosis, first infection, hospitalization, transition to adult care, transplantation &/or EOL care) & developmental milestones (e.g., starting daycare/school/higher education, non-adherence, becoming a teenager, first relationship, living independently, death of a friend, higher education & parenthood).<br>• Refers to & advocates for practical resources in support of the patient/family (e.g., funding of equipment & travel to hospital, completion of forms. e.g., disability tax credit, other; appropriate housing), as required. |

| | Prevention & Primary Service | Provincial CF Subspecialty Service |
|---|---|---|
| | **T1** | **T6** |
| Medication management | All ages:<br>• Reinforces the CF-related medication information/teaching provided by the CF Team.<br>• Orders all non CF-related medications.<br>• As directed & with support from the patient's T6 CF Team, renews selected CF-related medications.<br>• Consults with CF Team for medication-related questions/issues. | All ages:<br>• At each clinic visit, conducts medicine reconciliation reviews, monitors signs/symptoms of adverse drug reactions & assesses adherence to medications.<br>• Ensures up to date medicine allergy/intolerance listing.<br>• Evaluates for drug interactions.<br>• At least once/yr, reviews patient's medication regime for appropriateness.<br>• Educates & counsels patients/families on medication administration and reconstitution as needed.<br>• Ensures the patient is registered for Pharmacare Plan D and assists patient/family to register for Fair Pharmacare plan, as required.<br>• Orders, manages & may dispense CF-related medications, vitamins, enzymes and, in many cases, nutritional supplements to patients/families. Arranges for the same to be dispensed for patients living outside the lower mainland/Victoria. Includes medications/nutritional supplements for patients receiving drugs administered under Health Canada's Special Access Program (SAP) may assist patients involved in clinical trials and/or specified research studies.<br>• Completes (or assists physician to complete) Special Authorization applications & tracks expiry/renewals. |
| CF related diabetes (CFRD) screening & management<br><br>For specifics, refer to CF Care Guidelines for CFRD.[9] | • For patients ≥ 18 years old, or sooner if symptomatic, works with the CF Team to arrange for annual CFRD screening.<br>• For patients diagnosed with CFRD, works with the CF Team to complete the following:<br>  • Refer to endocrinologist +/- Diabetes Clinic, as appropriate, upon diagnosis.<br>  • Provide initial education & treatment re medical management, nutrition management, regular physical activity, stress reduction strategies. Adjusts insulin as required (consults with CF Team as required).<br>  • Monitor the status of CFRD (e.g., blood work, blood pressure, etc.) Where appropriate, works collaboratively with PCP in patient's local community.<br>  • If patient on insulin pump (initiated through the Diabetes Centre), monitors treatment & adjusts insulin as required.<br>  • Screen for diabetes-related complications. Refers to appropriate personnel/resources.<br>  • Provide regular updates to CF Team. | • For patients ≥ 10 years old, or sooner if symptomatic, works with the PCP to arrange for annual CFRD screening.<br>• For patients diagnosed with CFRD, works with the PCP to complete the following:<br>  • Refer to endocrinologist +/- Diabetes Clinic, as appropriate, upon diagnosis.<br>  • Provide initial education & treatment re medical management, nutrition management, regular physical activity, stress reduction strategies. Adjusts insulin as required.<br>  • Monitor the status of CFRD (e.g., blood work, blood pressure, etc.) Where appropriate, works collaboratively with PCP in patient's local community.<br>  • If patient on insulin pump (initiated through the Diabetes Centre), monitors treatment & adjusts insulin as required.<br>  • Screen for diabetes-related complications. Refers to appropriate personnel/resources.<br>  • Provide regular updates to PCP.<br>• For specifics, refer to CF Care Guidelines for CFRD. |

| | Prevention & Primary Service | Provincial CF Subspecialty Service |
|---|---|---|
| | **T1** | **T6** |
| Home IV therapy | All ages:<br>• In collaboration with T6 CF Team, provides support for CF patients requiring home IV therapy. May include:<br>  • Acting as the Most Responsible Physician (MRP) for patients living outside the lower mainland/Victoria.<br>  • Assessing the status of patients while on home IV therapy to confirm treatment effectiveness.<br>  • Providing advice to patient/family to maximize treatment effectiveness (e.g., adequate nutrition, chest clearing activities)<br>• Provides regular updates to the T6 CF Team re treatment effectiveness & related issues.<br>• Consults with CF Team for IV therapy-related questions/issues. | All ages:<br>• Works with patients/families & involved providers to develop care plans for patients with CF who require home IV therapy.<br>• Makes arrangements for home IV therapy to occur.<br>• Monitors the status of children/adults receiving periodic care in their local community. Adjusts care plans as necessary. |
| Transition of adolescents from pediatric to adult CF services | Provides support for CF patients transitioning from pediatric to adult CF services, utilizing On TRAC transition services (www.ontracbc.com). | • Provides transition support to patients/families referred to the Pediatric CF Team.<br>• Develops transition plans & oversees & supports the transition process of youth, beginning at age 12yr (at peds centre) and ending around age 24 yrs at the adult centre. The actual transfer to the appropriate adult service/CF Team usually takes place in the patient's 18$^{th}$ year.<br>• For specifics, refer to CF Care Guidelines for Adolescent Transition from Pediatric to Adult Healthcare Services.[15] |
| Support for patients receiving transplants | Provides support to patients before & after transplant. | All ages:<br>• Refers patients to the BC Transplant (BCT) Team for transplant, as appropriate (double lung & liver are most common). If required, BC Transplant Team will arrange for out-of-province transplants.<br>• Continues to provide CF-care pre and post-transplantation (BCT arranges transplant-specific assessment, testing & follow-up). |

| | Prevention & Primary Service | Provincial CF Subspecialty Service |
|---|---|---|
| | **T1** | **T6** |
| Support for sexual health, family planning, pregnancy & childbirth | • Sexual Health: Discusses sexual health issues and refers to sexual health clinic as needed.<br>• Family planning/contraception: Provides education and contraception options as per patient need.<br>• Pregnancy: Works closely with the CF Clinic and other specialists throughout the pregnancy and postpartum. | • Sexual Health: Discusses sexual health issues as they relate to CF and refers to GP or sexual health clinic for general information and screening.<br>• Family planning/contraception:<br>  • Family planning/contraception advice: Where ever possible, refers patient to PCP or public health clinic.<br>  • Carrier testing for partners of CF patients: Directs partner to their GP with offer to assist GP, if needed, with completion of genetic testing requisition ( If possible provide partner with copy of the required documentation as few GP's would have this in their office).<br>  • Offers parents of a CF child expecting another child an early referral to the CF Newborn Screening Program.<br>  • If requested, makes referral to a fertility clinic of patient's choice. For male patients in particular, review the impact of CF on fertility.<br>• Pregnancy & childbirth:<br>  • Reviews female patient's level of CF health and safety re: carrying a child. If female patient can't carry a child safely, discusses other options such as adoption or surrogacy.<br>  • Reviews medications, in particular antibiotic choices, both before and during pregnancy and during breast feeding (CF pharmacist).<br>  • Reviews pancreatic enzymes and vitamins in terms of dosage and the need to switch brands for safety (dietitian or pharmacist). Follows closely before, during and after pregnancy to ensure proper nutrition and weight gain (dietitian).<br>  • CF-related diabetes (CFRD):<br>  • For patients without CFRD standard screening is carried out throughout pregnancy.<br>  • For patients with CF-related diabetes (CFRD), follows CFRD pregnancy screening protocol (dietitian). Refers to an endocrinologist with experience in CFRD (CF MD).<br>• Referrals:<br>  • Where available and in consultation with the patient's PCP, refers to Maternal Fetal Medicine (MFM) Clinic or perinatologist, as required (pre-conception and throughout the perinatal period).<br>  • Offers referral, in consultation with the patients PCP, to an Obstetrician with experience in CF.<br>• Delivery is recommended at a hospital providing T6 maternal services, but if not possible CF clinic works closely with the patient's PCP & T4 hospital closest to the patient's home residence. |

| | Prevention & Primary Service | Provincial CF Subspecialty Service |
|---|---|---|
| | **T1** | **T6** |
| Support for patients at end-of-life (EOL) | All ages:<br>• Participates in EOL care planning and care, as described in the care plan.<br>• Supports patients/families throughout the EOL process.<br>• Refers families to local bereavement &/or counselling resources as required.<br>• Keeps CF Team updated. | All ages:<br>• Works with patients/families, PCPs & other involved providers to develop plan for EOL care. Consults with other services (e.g., At Home Program, palliative team, Canuck Place), as required.<br>• Arranges for inpatient admission (palliative care, hospice, complex pain team, general inpatient unit) &/or outpatient palliative care, as described in the care plan.<br>• Refers families to local bereavement &/or counselling resources as required.<br>• Keeps PCP updated. |
| Resource for providers | | All ages:<br>• Procedures in place for providers *throughout the province* to receive CF-related advice:<br>a. If urgent, 24/7 by pediatric or adult CF subspecialist.<br>b. If non-urgent, within 2 business days by a member of the CF Team.<br>• Upon request, liaises with schools, camps, workplaces & other local care providers *throughout the province* to provide expert advice on the care of children/adults with CF.<br>• Develops resources and tools to support providers/teams in providing appropriate care to patients with CF. guidelines, protocols, education modules, etc. |

### 4.1.1.3 Requirements

| | Prevention & Primary Service | Provincial CF Subspecialty Service |
|---|---|---|
| | **T1** | **T6** |
| CF physicians & interdisciplinary team | Local care provider with general knowledge in CF care. e.g., family physician, nurse practitioner, pediatrician, adult respirologist. | Pediatric CF Team includes:<br>• **Pediatric CF physician specialist(s)<br>• *Clinic coordinator/RN(s)<br>• *Dietitian(s)<br>• *Physiotherapist(s)<br>• *Social Worker(s)<br>• *Pharmacist(s)<br>• *Respiratory Therapist(s)<br>• *Psychologist(s), as available<br>• Clerk(s)<br><br>*Team members:<br>• Are assigned to the CF Clinic & are present during scheduled clinics.<br>• Have expert knowledge of CF & chronic disease & are experienced in the care of children with CF.<br>• Maintain expertise through ongoing CF-related clinical experience & completing CF-specific continuing education (e.g., rounds, conferences).<br><br>**Pediatric CF physician specialist(s):<br>• Holds joint appointment with the UBC Department of Pediatrics & the local HA/hospital.<br>• Maintains active linkages (in-person &/or virtually using technology) with the UBC Department of Pediatrics & T6 CF pediatric physician specialists (e.g., participates in rounds, case reviews, journal club, etc.)<br>• Maintains specialized knowledge & skills through ongoing CF-related clinical experience & completing CF-specific continuing medical education (CME).<br><br>Adult CF Team includes:<br>• **Adult CF physician specialist(s)<br>• *Clinic coordinator/RN(s)<br>• *Dietitian(s)<br>• *Physiotherapist(s)<br>• *Social Worker(s)<br>• *Pharmacist(s)<br>• *Respiratory Therapist(s)<br>• *Psychologist(s), as available<br>• Clerk(s) |

| | Prevention & Primary Service | Provincial CF Subspecialty Service |
|---|---|---|
| | **T1** | **T6** |
| CF physicians & interdisciplinary team cont'd | | *Team members:<br>• Are assigned to the CF Clinic & are present during scheduled clinics.<br>• Have expert knowledge of CF & chronic disease & are experienced in the care of adults with CF.<br>• Maintain expertise through ongoing CF-related clinical experience & completing CF-specific continuing education (e.g., rounds, conferences).<br><br>**Adult CF physician specialist(s):<br>• Holds joint appointment with the UBC Department of Medicine (or other, as relevant) & the local HA/hospital.<br>• Maintains active linkages (in-person &/or virtually using technology) with the UBC Department of Medicine & T6 CF adult physician specialists (e.g., participates in rounds, case reviews, journal club, etc.)<br>• Maintains specialized knowledge & skills through ongoing CF-related clinical experience & completing CF-specific medical education (CME).<br><br>Adult & Pediatric CF Team:<br>Refer to CF Standards of Care for BC[6] for recommended staffing levels (staffing levels are prorated based on clinic caseloads). |
| Specialist/subspecialist physician interdependencies | | See Table 7 for speciality/subspecialist physician interdependencies (in inpatient section). |
| Facilities | | Locally-based CF Clinic (Vancouver & Victoria):<br>• Space assigned for Pediatric CF Clinic &/or Adult CF Clinic. Space may be shared with other pediatric (Pediatric CF Clinic) or adult (Adult CF Clinic) clinics.<br>• Space must be able to accommodate appropriate infection control measures & have an adequate number of exam rooms & centralized conference room for staff. Portable spirometry equipment is recommended.<br><br>Outreach Clinic "host" sites (i.e., T6 CF Team provides "visiting" clinics at "host" sites):<br>• Access to a radiology department & pulmonary function testing.<br>• Access to a microbiology laboratory for CF respiratory cultures; if not available, arrangements are in place to transport samples to CF Clinic laboratory.<br>• Access to appropriate exam rooms, similar to above requirements, and a centralized conference room.<br><br>Virtual care (i.e., T6 CF Team provides services using telehealth, etc.)<br>• Telehealth facilities available within the CF Clinic.<br>• Access to telehealth facilities & infrastructure in designated locations throughout the HA. |

| | Prevention & Primary Service | Provincial CF Subspecialty Service |
|---|---|---|
| | **T1** | **T6** |
| Facility interdependencies | | • Respiratory: Pulmonary function lab<br>• Laboratory:<br>  • Microbiology laboratory with ability to test for full panel of CF respiratory pathogens, including B. cepacia Complex.<br>  • Sweat testing facility (by accredited method)<br>• Radiology:<br>  • Nuclear medicine<br>  • Interventional radiology (IV access, bronchial artery embolization, ultrasound-guided thoracentesis, feeding tube placement, portovenous hepatic shunt insertion & IVAD insertion)<br>• Outpatient day lab (or similar) |
| CF Accreditation & minimum visit volumes | | Participates in Accreditation Site Visit Program by CF Canada. Refer to www.cysticfibrosis.ca/our-programs/healthcare/accreditation-site-visit-program.<br><br>Accreditation standards do not include a minimum visit volume but do consider volumes, geography, interdependencies, quality improvement processes, etc in granting CF clinic accreditation. Accreditation site visits occur every 4 - 7 years. Accreditors provide recommendations for improvements, based on collective, national knowledge. |

### 4.1.2 Inpatient Services

Notes:

1. While the majority of services identified in this section will be provided as inpatients, some may be provided as either an inpatient or outpatient service, depending on the acuity and complexity of the service required (e.g., home IV therapy, chest physiotherapy and nutritional counselling).
2. Given the multiple medical complexities of patients with CF, most patients who require inpatient services will require a minimum of T4 inpatient services. Occasional exceptions may occur, usually due to geography and transportation, in which treatments/procedures may be done on a case-by-case basis and in close consultation with the patient's specialty CF Clinic, by an inpatient service at a lower tier (e.g., T2 or T3). These exceptions are appropriate in situations in which the resources (trained personnel, equipment, etc) are available and will facilitate care closer to the patient's home.

T4 is grayed out grayed out in the following charts because it is a *general* and not *CF-specific* service. It is included in this module to show the continuum of services.

#### 4.1.2.1 Service Reach and Responsibilities

| | Comprehensive Non CF-Specific Inpatient Service | Provincial CF Subspecialty Service |
|---|---|---|
| | T4 | T6 |
| Service reach | Multiple local health areas/health service delivery area/health authority. | Province. |
| Care & management | For unplanned admissions, conducts initial assessment and initiates treatment of acute issues (e.g., lab work, sputum for culture, evaluates nutritional status, chest physio, blood glucose control, etc.) Communicates with patient's CF Team to discuss ongoing management &/or whether transfer to T6 is required.<br><br>For planned admissions (e.g., surgery), consults CF Team prior to admission re potential impact of treatment on the patient's CF.<br><br>In consultation with the T6 CF Team, provides ongoing inpatient care[3] to patients who:<br>• Have relatively common CF-related complications which usually involve a single system. Immediate deterioration is unlikely. | Same as T4 plus:<br>• Provides telehealth/telephone consultation to T4 providers re patients with CF and available to provide advice to the transplant teams as needed.<br><br>Provides inpatient care to children and adults who:<br>• Have highly complex &/or uncommon CF-related complications which may involve multiple systems. Complications may be life threatening & require admission to ICU. Multiple specialty/subspecialty teams may be involved. Examples:<br>  • Severe respiratory infection &/or respiratory failure (invasive or non-invasive ventilation often required).<br>  • Bowel obstructions.<br>  • Large volume hemoptysis (bronchial artery embolization often required).<br>  • Unstable CF-related diabetes. |

[3] Refer to Children's Medicine module at http://childhealthbc.ca/tiers-service/childrens-medical-services for specific activities with respect to children's inpatient care (e.g., monitoring, deteriorating/emergency situations, mental health crises, child & family teaching, discharge planning, parenteral fluid & medication management, nutrition management, psychosocial & spiritual support).

| | Comprehensive Non CF-Specific Inpatient Service | Provincial CF Subspecialty Service |
|---|---|---|
| | **T4** | **T6** |
| Care & management cont'd | • Course is predictable & condition can be treated using standard care algorithms/protocols. Examples:<br>  • Respiratory infection (most common) requiring IV antibiotics/therapies.<br>  • Bowel complication (2nd most common) requiring assessment, identification of appropriate therapy, management of pain & other symptoms &/or initiation of enteral feeding etc.<br>• Require admission for conditions unrelated to CF (e.g., acute appendix, gall bladder disease). CF is stable.<br><br>Collaborates with the patient's T6 CF Team & PCP about the discharge plan.<br><br>Meets inpatient care & infection control standards as outlined in the CF Standards for Care in BC (2018). | • Insufficient response to outpatient treatment.<br>• Failure to improve after admission to T4.<br>• Have been activated for transplantation.<br>• Require admission for conditions unrelated to CF (e.g., surgery). CF may require monitoring &/or interventions (e.g., TPN) during the inpatient admission.<br>• Requires palliative/EOL care.<br><br>Collaborates with the patient's PCP & other specialists involved in the patient's care about the discharge plan.<br><br>Meets inpatient care & infection control standards as outlined in the CF Standards for Care in BC (2018). |

### 4.1.2.2 Requirements

| | Comprehensive Non CF-Specific Inpatient Service | Provincial CF Subspecialty Service |
|---|---|---|
| | T4 | T6 |
| **Requirements** | | |
| Physicians & interdisciplinary team | Children:<br>• On-site pediatric unit.<br>• Pediatrician is the Most Responsible Physician (MRP) & is available on-call 24/7 & on-site as required. Pediatrician has general knowledge about CF care & common CF-related issues.<br>• Access to interdisciplinary inpatient team. Team includes nurses, social worker(s), child life specialist(s), RT(s), PT(s), OT(s), dietitian(s) & pharmacist(s). Team has general pediatric knowledge & skills, including knowledge/skills in the management of respiratory & GI-related symptoms. For specifics, refer to medical module at http://childhealthbc.ca/tiers-service/childrens-medical-services.<br>• Pediatrician and interdisciplinary team are encouraged to access T6 Pediatric CF Team via telehealth/telephone for CF-related advice/consultation.<br><br>Adults:<br>• On-site medical unit.<br>• General internist or subspecialist (e.g., respirologist) is the MRP & is available on-call 24/7 & on-site as required. MRP has general knowledge about CF care & common CF-related issues (usually respirologist).<br>• Access to interdisciplinary inpatient team. Team includes nurses, social worker(s), RT(s), PT(s), OT(s), dietitian(s) & pharmacist(s). Team has adult medicine knowledge & skills, including knowledge/skills in the management of respiratory & GI-related symptoms.<br>• General internist or subspecialist is encouraged to access T6 Pediatric CF Team via telehealth/telephone for CF-related advice/consultation.<br>• Access to T6 Adult CF Team via telehealth/telephone.<br><br>See Table 7 for a detailed list of physician interdependences. | Children:<br>• On-site pediatric unit.<br>• If CF physician is not the MRP, CF physician is readily available for patient care discussions. Attends on-site as needed.<br>• Pediatrician or designate (e.g., resident) available on-site 24/7.<br>• Access to interdisciplinary team. Team includes nurses, social worker(s), child life specialist(s), RT(s), PT(s), OT(s), dietitian(s) & pharmacist(s). Team has enhanced expertise in caring for patients with CF.<br>• Pediatric CF Team available to T4 for consultation. Actively involved in the inpatient assessment, care planning &, as required, provision of treatment.<br><br>Adults:<br>• On-site medical or medical subspecialty unit (e.g., respiratory focus).<br>• If CF physician is not the MRP, CF physician is readily available for patient care discussions. Attends on-site as needed.<br>• Access to interdisciplinary inpatient team. Team includes nurses, social worker(s), RT(s), PT(s), OT(s), dietitian(s) & pharmacist(s). Team has enhanced expertise in caring for patients with CF (e.g., respiratory focus).<br>• Adult CF Team available to T4 for consultation. Actively involved in the inpatient assessment, care planning &, as required, provision of treatment.<br><br>See Table 7 for a detailed list of physician interdependences. |

| | Comprehensive Non CF-Specific Inpatient Service | Provincial CF Subspecialty Service |
|---|---|---|
| | **T4** | **T6** |
| **Requirements** | | |
| Facilities | Inpatient unit must be able to accommodate appropriate infection control measures. Single-person rooms preferred. | Same as T4 plus:<br>• Inpatient unit has single-person room(s) for patients with CF.<br>• Inpatient unit has space/equipment for additional food storage & preparation.<br>• Inpatient unit has designated space for parents/caregiver(s).<br>• Space/equipment is available in reasonable proximity to the inpatient unit for exercise. |

**Table 8: Specialist/Subspecialist Physician Interdependencies**

✓24/7 = **Available for on-site consultation as needed.**
✓ M-F days = **Available for on-site consultation days M-F.**

| | Availability | | | |
|---|---|---|---|---|
| | Pediatrics | | Adults | |
| **Specialist in....** | **T4** | **T6** | **T4** | **T6** |
| CF physician (respirologist or pediatrician with CF expertise/ experience) | | ✓24/7 At a minimum, CF physician available days M-F; Pediatrician with general CF knowledge may supplement CF physician to complete 24/7 coverage | | ✓24/7 At a minimum, CF physician available days M-F; Respirologist with general CF knowledge may supplement CF physician to complete 24/7 coverage |
| Adolescent medicine | | ✓ M-F days | | |
| Allergy | | ✓ M-F days Pediatric or general allergist | ✓ M-F days | ✓ M-F days |
| Anesthesiology | ✓24/7 General anesthesiologist | ✓24/7 Pediatric anesthesiologist | ✓ 24/7 | ✓ 24/7 |
| Cardiology | | ✓ M-F days Pediatric cardiologist | | ✓ M-F days |
| Critical care medicine (CCM) | | ✓24/7 Pediatric CCM | ✓ 24/7 | ✓ 24/7 |
| Dermatology | | ✓ M-F days Pediatric cardiologist | | ✓ M-F days |
| Developmental pediatrics/child development & rehab | | ✓ M-F days | | |
| Endocrinology | | ✓M-F days Pediatric endo | ✓ M-F days | ✓ 24/7 |
| ENT | ✓24/7 General ENT | ✓24/7 Pediatric or general ENT | ✓24/7 | ✓24/7 |
| Gastro-enterology | ✓24/7 Adult GI | ✓24/7 At a minimum, pediatric GI available days M-F; Adult GI MD supplements pediatric GI to complete 24/7 coverage | ✓24/7 | ✓24/7 |
| General surgery | ✓ 24/7 Adult general surgeon | ✓ 24/7 Pediatric surgeon | ✓24/7 | ✓24/7 |
| Hematologist/ oncologist | | ✓ M-F days Pediatric hem/onc | | ✓ M-F days |

| | Availability | | | |
|---|---|---|---|---|
| | Pediatrics | | Adults | |
| **Specialist in....** | **T4** | **T6** | **T4** | **T6** |
| Infectious diseases (ID) | ✓Telephone consultation available 24/7<br>Adult ID | ✓24/7<br>Available on-site days M-F. Telephone consultation available outside these hours.<br>Pediatric or adult ID | ✓Telephone consultation available 24/7 | ✓Telephone consultation available 24/7<br>Adult ID |
| Medical genetics | | ✓ M-F days | | ✓ M-F days |
| Medical Microbiology | ✓24/7<br>Adult microbi | ✓24/7<br>Pediatric or adult microbi | ✓24/7 | ✓24/7 |
| Nephrologist | | ✓ M-F days<br>Pediatric neph | | ✓ M-F days |
| Neurologist | | ✓ M-F days<br>Pediatric neurologist | | ✓ M-F days |
| Obstetrics/gynecology | | | | T6 obstetrics services available 24/7 |
| Pain specialist | ✓ M-F days<br>Team serves children & adults | ✓ M-F days<br>Team serves children & adults or children only | ✓ M-F days | ✓ M-F days |
| Palliative care specialist | | ✓ M-F days<br>Pediatric specialist | | ✓ M-F days |
| Pediatrician | ✓24/7<br>On-call | ✓<br>On-site 24/7 | | |
| Radiology | ✓ 24/7<br>General radiologist | ✓24/7<br>At a minimum, pediatric rad available days M-F; General rad supplements pediatric rad to complete 24/7 coverage<br><br>✓24/7<br>Access to interventional radiology | ✓ 24/7 | ✓ 24/7, including interventional radiology |
| Psychiatry | ✓24/7<br>General psychiatrist | ✓24/7<br>At a minimum, C&Y psychiatrist available days M-F; Gen psychiatrist supplements C&Y psychiatrist to complete 24/7 coverage | ✓24/7 | ✓24/7 |
| Rheumatology | | ✓M-F days | | ✓M-F days |
| Transplant | | ✓24/7<br>Pediatric Team (BCT) | | ✓24/7<br>Adult Team (BCT) |

## *4.2 Knowledge Sharing & Transfer/Training*

T1 and T4 are grayed out grayed out in this chart because they are *general* and not *CF-specific* services. They are included in this module to show the continuum of services.

| Prevention & Primary Service | Comprehensive Non CF-Specific Inpatient Service | Provincial CF Subspecialty Service |
|---|---|---|
| **T1** | **T4** | **T6** |
| *Physicians & staff (interdisciplinary team):*<br><br>Accesses learning activities that support the maintenance of physician & staff competencies in CF. e.g., on-line access to guidelines/reference materials/continuing education courses & participation in HA & provincial learning activities relevant to CF & child/adult health (e.g., rounds and conferences). | Same as T1. | *Physicians & staff (interdisciplinary team):*<br><br>Same as T1 plus:<br><br>Organizes &/or participates in local, provincial, national/international activities/conferences that support the maintenance of physician & staff competencies in the care of children/adults with CF.<br><br>Organizes & participates in province-wide learning activities that support the maintenance of physician & staff competencies in the care of children/adults with CF. e.g., rounds and conferences.<br><br>Organizes & moderates CF "family information days" (for patients & families of CF patients living throughout BC).<br><br>Liaises with CF Canada, including disseminating information from local chapters and national office to CF Team members<br>*Students, residents & fellows:*<br>Creates opportunities to expose a broad range of undergraduate, graduate & post-graduate health care students to the care of children/adults with CF.<br><br>Provides outpatient experiences/placements in the care of children/adults with CF for a broad range of undergraduate, graduate & post-graduate health care students. |

## 4.3 Quality Improvement & Research

T1 and T4 are grayed out grayed out in this chart because they are *general* and not *CF-specific* services. They are included in this module to show the continuum of services.

| Prevention & Primary Service | Comprehensive Non CF-Specific Inpatient Service | Provincial CF Subspecialty Service |
|---|---|---|
| **T1** | **T4** | **T6** |
| *Quality improvement:*<br>• Participates in the provincial approach to evaluation / quality improvement of CF and contributes relevant data on indicators | *Quality improvement:*<br>• Same as T1. | *Quality improvement:*<br><br>*Provincial role*<br>• Develops a provincial approach to evaluation/quality improvement of pediatric/adult CF care, including electronic mechanisms for collection and analysis of data to support clinical care & quality improvement initiatives.<br>• Develops & disseminates guidelines/standards/pathways on CF-related care.<br>• Creates structures and processes to support active linkages (in-person &/or virtually using technology) amongst CF providers in BC.<br><br>*Role of locally-based clinics (Vancouver, Victoria)*<br>• Regularly reviews the quality of CF care provided to children/adults, including case reviews. Implements recommendations.<br>• Participates in the provincial approach to evaluation / quality improvement of CF care.<br>• Participates in professional development including attending meetings and conferences. e.g., the annual North American CF conference.<br>• Participates in the Canadian CF Registry. Obtains patient consents and updates registry data on an ongoing basis.<br>• Clinics are accredited by CF Canada and host periodic site visits by CF Canada.<br><br>*Research:*<br>• Actively participates in national and international CF groups.<br>• Conducts & supports other to conduct research in CF. |

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# Appendix 1 - Definition of Tiers

| Tier | Characteristics |
|---|---|
| T1: Prevention, primary and emergent health service | • Services available and accessible in most communities<br>• Service providers generally serve children AND adults<br>• Services target health promotion and common, low complexity health needs<br>• Service participates in regional and provincial quality improvement initiatives |
| T2: General health service | • Services are accessible to multiple communities and/or local health area<br>• Service providers generally serve children AND adults<br>• Services target health promotion and common, low complexity health needs<br>• Service participates in regional and provincial quality improvement initiatives |
| T3: Adult or child-focused health service | • Services available to multiple local health areas/health service delivery area<br>• Service providers specialize in children OR adults<br>• Service targets relatively common, medium complexity health needs<br>• Service participates in regional and provincial quality improvement initiatives |
| T4: Comprehensive health service | • Service is accessible to health service delivery area/health authority<br>• Service providers focus the majority of their time on children OR adults<br>• Service targets a broad range of medium complexity health needs<br>• Service offers general health experiences/placements for a broad range of undergraduate, graduate and post-graduate health care students and residents<br>• Service identifies relevant regional health equity indicators, and leads/participates in regional and provincial quality improvement initiatives |
| T5: Regional subspecialty health service | • Service is accessible to a health authority<br>• Most service providers work exclusively with children OR adults<br>• Service targets relatively common, high complexity health needs. Level of complexity usually does not require the availability of other on-site subspecialty teams<br>• Service offers general health experiences/placements for a broad range of undergraduate, graduate and post-graduate health care students and residents. Also offers placement for fellows in selected subspecialties<br>• Service identifies relevant regional health quality indicators and leads/participates in regional and provincial quality improvement initiatives<br>• Service participates in relevant health related research |
| T6: Provincial subspecialty health service | • Service is accessible to the province<br>• Most service providers work exclusively with children OR adults<br>• Service targets low incidence, high complexity health needs. Level of complexity often requires the availability of other on-site subspecialty teams<br>• Service offers general and subspecialty health experiences/placements for a broad range of undergraduate, graduate and post-graduate health care students, residents and subspecialty fellows.<br>• Service identifies relevant provincial health quality indicators and leads/participates in regional and provincial quality improvement initiatives<br>• Service conducts and supports others to conduct relevant health related research |