

# BRITISH COLUMBIA PROVINCIAL NEWBORN SCREENING PROGRAM

Guidelines for Management of  
the Infant Diagnosed With  
Cystic Fibrosis

Revised November 2017

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## Part One: Program Overview

### Mission Statement

- To continue to improve CF care, quality of life and survival, for infants and children diagnosed with CF through newborn screening in BC
- To shift health care focus from reactive care to proactive care
- To instill in families and health care providers a strong working relationship and collaboration with the two pediatric CF Clinics in BC
- To provide the highest standard of care so that a new generation of children with CF may fully benefit from future CF treatments
- To provide personalized care which is family centered and includes the family as a valued partner of the healthcare team.

# I. Introduction and Process Map for the BC Newborn Screening Program

## Management of Newborn Infants Diagnosed with Cystic Fibrosis:

*An Introduction and Process Map for the BC Newborn Screening Program*

### Newborn Screening for Cystic Fibrosis

The development and implementation of the CF newborn screening algorithm was finalized (Figure 1) and screening all newborns born in BC and the Yukon for CF commenced in the fall of 2009. Over 65 infants have been diagnosed with CF during the first seven years of the program (Figure 2).

To maximize the benefit of screening, the aim has been to see all infants within the first month of life, before significant morbidity and malnutrition occur. We have found that it is possible to manage and educate these infants and families in an ambulatory setting. This promotes bonding and minimizes the disruption to the family unit at such a sensitive time. However, this decision may be influenced by the clinical status of the child, the geographical location of the family, and family's ability to achieve the educational and therapeutic standards required.

### Geographical Distribution

There are two accredited CF clinics located in British Columbia (BC):

- Vancouver: BC Children's Hospital (BCCH) follows patients who live in mainland BC and the Yukon.
- Victoria: Victoria General Hospital follows patients who live on Vancouver Island.

To ensure continuity of care and equitable resource provision, all infants with inconclusive CF newborn screening results will be contacted by the Newborn Screening Nurse to arrange follow up (Figure 3).

The initial assessment and sweat test will be undertaken at BCCH to ensure rapid testing and to maximize quality assurance. The exceptions are infants who have two mutations on genetic screening and live on Vancouver Island. These infants will be followed by the Victoria CF Clinic and should be seen there for initial assessment and sweat testing to ensure continuity of care.

## Follow up schedule for newly diagnosed infants

Once infants have been identified, treatment should be initiated in a timely manner. Over time there will be a shift in the CF care paradigm from reactive to pro-active management, with the clinical focus being on nutritional wellness and microbial surveillance. Previously published recommendations, for care of the newborn infant with CF, state that infants should be seen on a monthly basis. The BC Newborn Screening Program aims to see all newly diagnosed infants weekly for the first month and then monthly if clinically well. Monthly visits will continue for the first year of life. After this children will be seen in the CF Clinic every 2-3 months.

In the lower mainland this is achievable and practical, but the frequency of follow up will have a huge social and economic impact on families from the interior, Northern BC and Northern Vancouver Island. To ensure 'accessibility' to services for all BC residents, the aim is to involve local pediatricians and family doctors as point of care contacts for interim review of newly diagnosed infants. The BC CF clinics are working with Child Health BC to offer outreach services to both northern BC and the BC interior.

The proposed follow-up schedule for infants diagnosed with CF is highlighted in Figure 4.

## Multidisciplinary Roles for the newborn infant

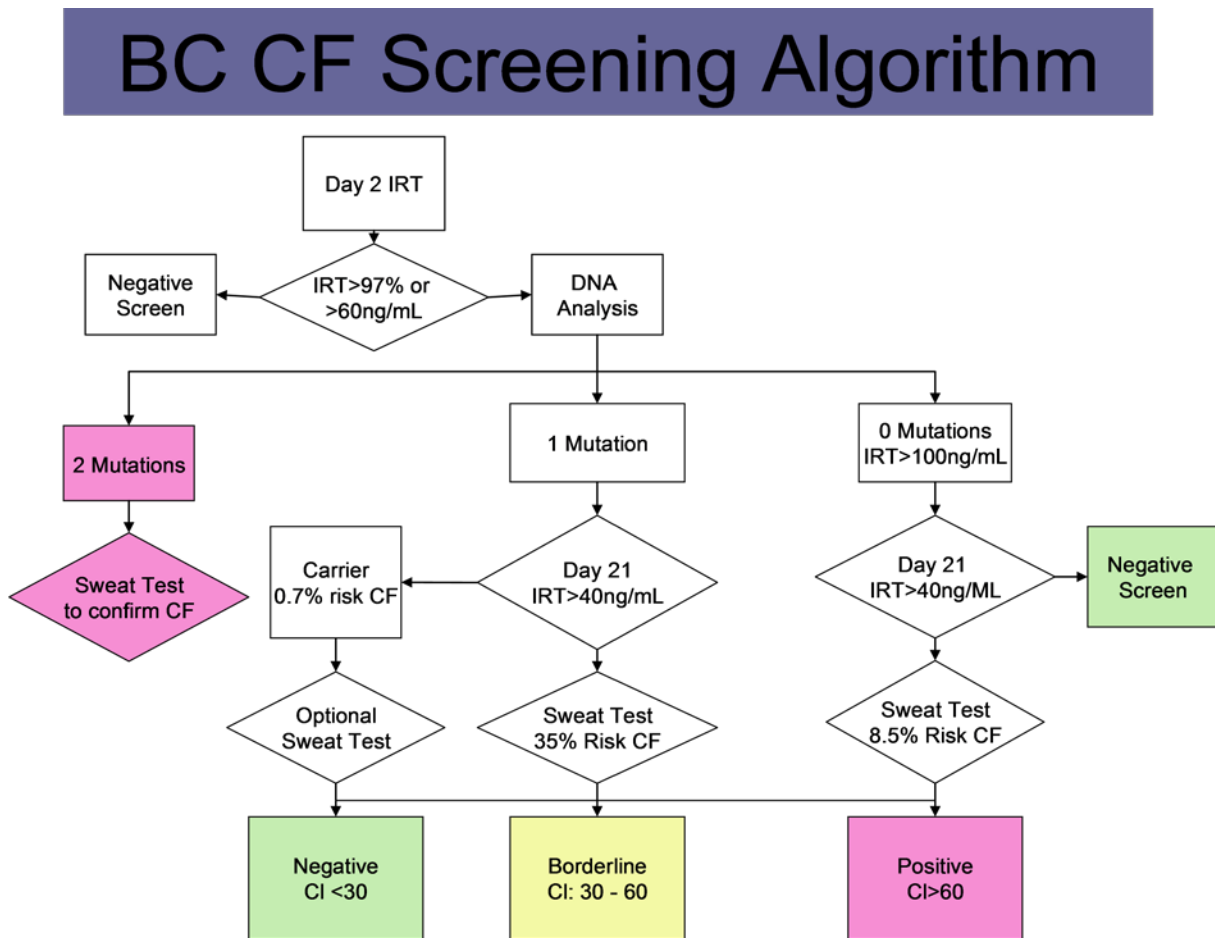
The CF care team is comprised of:

- Physician
- Nurse
- Physiotherapy
- Dietician
- Pharmacist
- Social Work
- Psychologist/Psychiatrist

“CF clinics provide specialized multidisciplinary care for individuals with cystic fibrosis. CF patients are seen by various healthcare professionals at each clinic visit, and during hospitalization. This multidisciplinary approach optimizes the care delivered – regular interaction with healthcare professionals at clinic visits that include all team members offer convenient, comprehensive care and promote a long-term association with CF clinical care. Cystic fibrosis must be treated throughout life, and it is important that affected individuals develop a comfortable, trusting relationship with clinic personnel.” (quoted from CF Canada website)

- See more at: <http://www.cysticfibrosis.ca/our-programs/healthcare/how-cf-care-is-delivered#sthash.rUoLD6d3.dpuf>

Figure 1:  
Newborn Screening Algorithm



\*3-5% False Negative Rate

Positive Predictive Value: 6% (2010), 11%(2011)

\*Sweat test borderline range is 30-60 for children 6 months or older

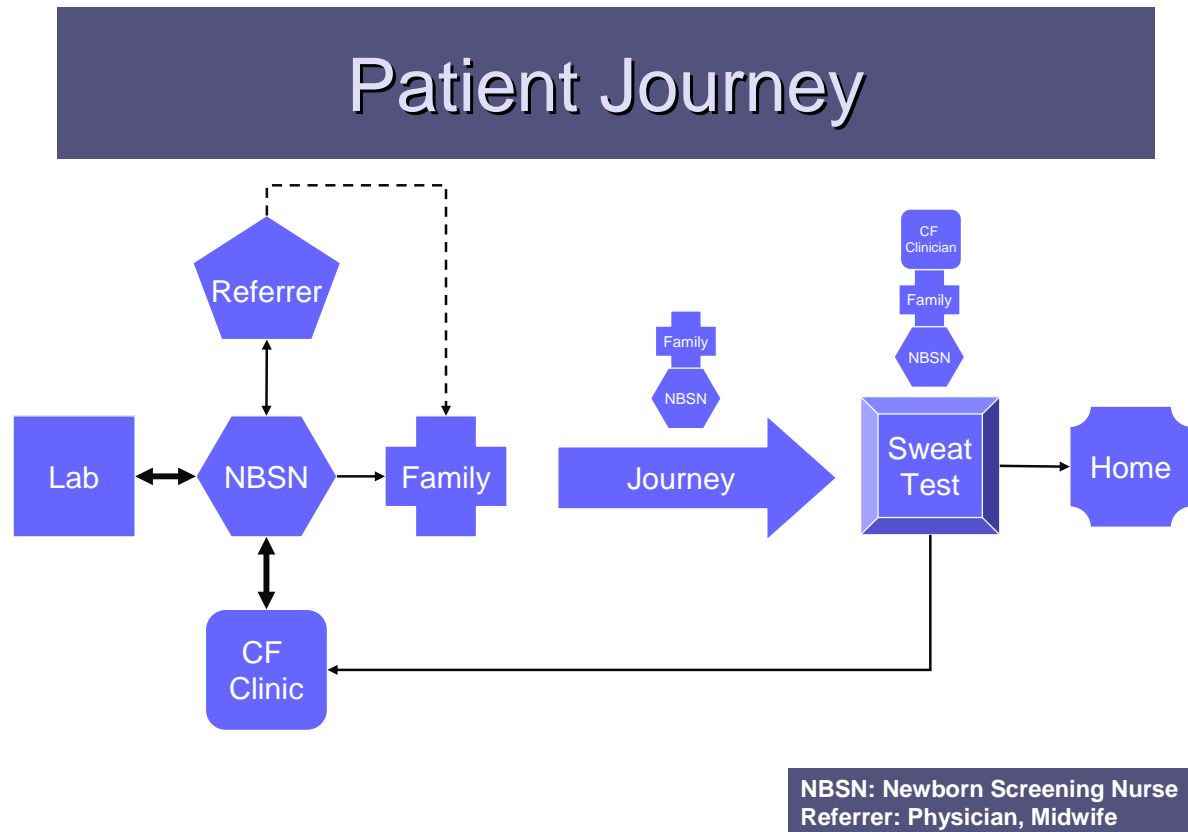
\*DNA analysis increased from a 39 mutation panel to 139 mutation panel in October 2016. Genetic sequencing, if necessary and approved by MSP, is currently shipped out of province to Edmonton for processing.



**Figure 2:  
Summary of CF Newborn Screening 2010-2016**

BC CF Screening Outcome	2010	2011	2012	2013	2014	2015	2016
Initial Screens	44 727	44 347	44 494	44 813	45 262	44 155	45 593
DNA Screening (Top 3% or >60ng/mL)	1412	1423	1447	1462	1358	1449	1458
Follow-Up Required	136	128	171	175	127	164	132
High IRT & 2 Mutations	9	12	7	7	13	6	9
High IRT & 1 Mutation	97	94	114	123	78	96	84
CF Confirmed	0	1	4	2	1	2	3
High IRT & 0 Mutations	24	34	57	52	36	62	48
CF Confirmed	0	0	0	0	0	0	0
Total CF Confirmed	8	13	11	9	16	8	10
False Negative	1 (mec illeus)	1	2	1	2	1	2 (1 born in 2010)
False Positive	1 (corrected as carrier)	0	0	0	0	0	0

**Figure 3:**  
Screened Infant journey



1. Infant identified by lab
2. Newborn screening Nurse (NBSN) contacts referrer, liaises with CF Team and lab
3. NBSN arranges sweat test and acts as point of contact/counselor for family
4. Family seen by NBSB and CF Physician prior to sweat test; Consultation
5. Sweat results given to family; if positive then follows CF team algorithm
6. Negative result: Discharged home and letter supplied to primary provider
7. Borderline result: All results reviewed by CF team and follow up CF clinic
8. Insufficient sweat: Repeat sweat test arranged by NBSN

**Figure 4:**  
**Schedule for Infants diagnosed with Cystic Fibrosis**

<b>Diagnosis, Education and Follow-up for Newborn Infants with CF from BC</b>					
<b>Week</b>	<b>Mon</b>	<b>Tues</b>	<b>Wed</b>	<b>Thurs</b>	<b>Fri</b>
<b>1</b>		■ ■	■	■ ■	■
<b>2</b>	■	■	■		■
<b>3</b>		■			
<b>4</b>		■			
<b>Month</b>					
<b>2</b>		■			
<b>3</b>		■			
<b>4</b>		■			
<b>5</b>		■			
<b>Key</b>					
■ ■	Sweat test and investigation; Diagnosis and explanation				
■	CF Counseling; Basic nutrition				
■	Intensification of Therapy; CF Education				
■	CF Clinic Visit *				
■	Telephone Follow up CF clinic				

\* week 3 may be a telephone/telehealth consultation, as deemed appropriate by the CF team/family.

## Cystic Fibrosis Screen Positive, Inconclusive Diagnosis (CFSPID)

A consequence of newborn screening is that babies may be identified as having CFSPID (Cystic Fibrosis screen positive inconclusive diagnosis). A diagnosis of CFSPID is given when there is an abnormal newborn screening result for CF but the child does not meet all the criteria for a CF diagnosis: 1 or more CF causing gene mutations or signs and symptoms of CF disease AND one or more positive sweat chloride tests (>60mmol/l).

CFSPID is a category where an infant is screen positive with raised immune reactive trypsinogen and may have one of two scenarios:

- A. Two CF gene mutations are identified, of which one or no mutations are CF causing. The sweat test is normal(<30mmol/l).
- B. One or no CF mutations are found and sweat test results are borderline (30-60mmol/l).

The frequency of CFSPID has been reported to be 1 CFSPID diagnosed for every 3-5 CF diagnoses. About 20 babies per year are diagnosed with CFSPID in Canada.

The natural history of this condition is unknown and is currently being studied. Around 10% of these infants will later be diagnosed with classical CF between the ages of 2 to 5 years. It is also known that some patients may not be diagnosed until adulthood, by which time, may have developed significant bronchiectasis.

Reports from cross sectional observational studies have described CFSPID infants as having mild or no symptoms. The vast majority will be pancreatic sufficient. CFSPID infants are at risk of acquiring pseudomonas and will need to have appropriate eradication.

In Canada and British Columbia all babies with CFSPID are followed in the CF clinic. Currently the frequency of follow up is as per the table below:

	Visit 1 0-2m	Visit 2 6m	Visit 3 12m	Visit 4 18m	Visit 5 24m	Annual F/up Visits	Interim Visit Q6 months	Visit 6 Years
CFTR Sequencing	X							
History and physical exam	X	X	X	X	X	X	X	X
Anthropometric measurements	X	X	X	X	X	X	X	X
Throat swab	X	X	X	X	X	X	X	X
Sweat chloride test (CFSPID)	X	X	X	X	X	X		X
Sweat chloride test (CF)	X				X			
Fecal elastase/fecal fat	X		X		X	X		X
Bloodwork*	X		X		X	X		X
Chest x-ray	X		X		X	X		X
Abdominal ultrasound	(only if clinically indicated)							X
Urine Sodium	X	X	X		X	X		X
Infant/preschool PFT/MBW/LCI	X						X	X
Standard PFT						X**		X

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## II. Recommendations for Management of the Infant Diagnosed with Cystic Fibrosis

### BC Cystic Fibrosis Clinics' Recommendations for Management of the Infant Diagnosed with Cystic Fibrosis

#### Topic 1: Initial Diagnosis

- Diagnosis and treatment of infants with CF should be done at an accredited CF Care Center as soon as possible after the initial newborn screening test.
- Initial Intensive Education Days should be performed at an accredited CF Care Center as soon as possible after confirmation of diagnosis.
- Basic genetic information will be provided. A referral to a genetic counselor should be offered after diagnosis to discuss the implications for family members and perform genetic testing, if desired.

#### Topic 2: Nutrition

##### It is recommended that:

- Pancreatic functional status be measured by fecal elastase in all infants at the initial visit, at 6 months of age and twice yearly thereafter (if results are abnormal or significant weight loss is seen fecal elastase should be repeated more frequently).
- Pancreatic Enzyme Replacement therapy be started in
  - Infants with unequivocal signs or symptoms of malabsorption, while awaiting confirmatory test results
  - All infants with two CFTR mutations associated with Pancreatic Insufficiency (see table of mutations for guidance-Table 1).
  - In all infants with fecal elastase <200 ug/g or other objective evidence of pancreatic insufficiency

- Pancreatic enzyme replacement therapy should be initiated at a dose of 500-1000 units lipase/kg/feed. Total daily dose can be titrated up to a maximum of 10 000 lipase units/kg/day. Doses should be given no sooner than 2 hours apart.

### Topic 3: Feedings, Vitamins and Micronutrients

Exclusive feeding of breastmilk or standard infant formula will meet CF nutrition requirements for the first 6 months of age

- Partially or extensively hydrolyzed protein formula should be considered for infants with food protein (such as cow's milk) allergy or intolerance or extensive bowel resection.
- Positive feeding behaviors should be encouraged and educational resources should be provided to families. This includes:
  - Providing a positive feeding environment that has limited noise, light, and other distractions.
  - Introducing solid foods and allowing self-feeding skills to develop at 4-6 months of age.
  - Managing neophobia (the reluctance to try new foods) by presenting foods up to 10-12 times before determining that a child does not like them.
  - Providing positive reinforcement to appropriate eating behaviors (e.g. trying new foods, eating independently) and minimizing attention to poor eating behaviors (e.g. food refusal)
- Multivitamins designed to provide at least the recommended levels of vitamin A, D, E and K for infants with pancreatic insufficiency should be prescribed by the CF physician at diagnosis).
- Blood levels of fat-soluble vitamins should be measured at diagnosis, at 6 months of age, and annually thereafter. It is advised to check levels more frequently if values are abnormal. Prothrombin time should be monitored as a measure of vitamin K levels in the bloodstream.
- Infants with CF are recommended to start sodium supplementation of 2-4 mEq/kg/day until they are eating sufficient amount of solid foods (approximately 12 months months of age).
- Consider sodium as a factor affecting nutritional status and weight gain. Monitor urine and serum sodium at time of diagnosis and as clinically indicated. Sufficient sodium status is urinary sodium >30mmol/L.

## Topic 4: Pulmonary

- A smoke-free environment is recommended for the infant and all caregivers must be informed that cigarette smoke exposure harms children. Families struggling with smoking cessation should be advised of local resources for assistance (i.e. programs, medications, and/or counseling).
- Parents should be informed that exposure to wood stoves or campfires can cause bronchospasm in CF infants with increased airway reactivity.

## Topic 5: Physiotherapy

### It is recommended that:

- Airway clearance therapy is initiated at time of diagnosis and is usually performed 2 – 3 times daily. Several airway clearance techniques may be used which could include modified postural drainage with percussion and vibration, , assisted autogenic drainage or baby positive expiratory pressure.
- There is regular surveillance of the infant's overall development as malnourishment may delay developmental milestones.
- The physiotherapist provides anticipatory guidance regarding physical strategies that promotes an active lifestyle and development of a healthy respiratory system.
- If inhaled medications are required, the caregivers should be instructed in the correct technique of inhalation therapy.

## Topic 6: Infection Control, Surveillance and Treatment

- It is recommended that infants with CF be separated from other CF patients cared for in a designated CF baby Clinic. Adequate infection control education (i.e. appropriate hand hygiene and cough etiquette) to be provided and understood by all caregivers.
- All new diagnosis exams and teaching should be conducted in a designated area to prevent cross-colonization of potential pathogens.
- Follow up visits with the CF Team should be done in a designated CF baby clinic.
- Appropriate cleaning and disinfecting of devices for inhaled medications, by parents and the health care team, should be done in both healthcare and other settings to prevent acquisition of potential pathogens.



- During hospitalization, infection control measures should be followed according to local hospital guidelines. CF patients should always be admitted to a private room with a private bath.
- Infection control measures should be implemented in compliance with BC CF Clinic and CF Canada recommendations to minimize transmission of bacterial infections.
- The annual influenza vaccination is recommended for infants with CF greater than or equal to 6 months of age, all household members, and all health care providers caring for these children. Household contacts and out of home care givers of children with CF less than 6 months of age also should receive the annual influenza vaccine.
- Children and infants with CF should receive all recommended regularly scheduled vaccinations at the appropriate intervals. A CF specific vaccination schedule is available on the BCCDC website under Immunization Manual, Section III, subsection 2.6 – Cystic Fibrosis ([http://www.bccdc.ca/resource-gallery/Documents/Guidelines%20and%20Forms/Guidelines%20and%20Manuals/Immunization/Vaccine%20Info/BCCH\\_CysticFibrosisVaccineLetter.pdf](http://www.bccdc.ca/resource-gallery/Documents/Guidelines%20and%20Forms/Guidelines%20and%20Manuals/Immunization/Vaccine%20Info/BCCH_CysticFibrosisVaccineLetter.pdf)). This schedule includes additional protection from Pneumococcal infection at 6 months and 2 years of age.
- For infants with CF under one year of age, the use of palivizumab is recommended for prophylaxis of respiratory syncytial virus. Applications should be submitted to the BC RSV Task Force. Current practice guidelines may be accessed on the Child Health BC website: <http://www.childhealthbc.ca/search/content?keyword=rsv>
- It is recommended that monthly cough swabs be taken for bacterial surveillance. Additional cough swabs may be required if clinically indicated.
- If an infant under 1 year of age is culture positive and symptomatic the infant should be treated.
- If an infant under 1 year of age is culture positive and asymptomatic, please consult the CF clinic.
- If an infant under 1 year of age is culture positive for staphylococcus aureus, the infant should be treated regardless of symptoms.

- It is recommended that new acquisition of *Pseudomonas aeruginosa*, defined as initial acquisition or new acquisition after 'successful' eradication therapy, should be treated with anti-pseudomonal antibiotics and increased airway clearance, regardless of the presence or absence of symptoms.
- Those who remain persistently colonized, after three unsuccessful attempts at eradication, with *Pseudomonas aeruginosa* should be treated chronically with inhaled anti-pseudomonal agent and oral Azithromycin.
- It is recommended that new acquisition of MRSA, defined as initial acquisition or new acquisition after 'successful' eradication therapy, should be treated with anti-MRSA antibiotics and increased airway clearance, regardless of the presence or absence of symptoms.
- It is recommended that bronchoscopy and bronchoalveolar lavage be considered in infants with symptoms or signs of lung disease and/or who fail to respond to treatment.
- Environmental infection control precautions recommend avoidance of certain situations which may put the infant at risk. This includes hot tubs in which *Pseudomonas* bacteria grows easily and the warm moist environment surrounding them.

### Topic 7: Diagnostic Testing

- A baseline chest x-ray is recommended at 3 months and 1 year. Additional chest x-rays will be done if clinically indicated.
- The use of chest CT scans for routine surveillance is not recommended because they require sedation and expose patients to unnecessary doses of radiation.

### Topic 8: Chronic Pulmonary Therapies

- Inhaled mucolytic agents will be prescribed by the CF Clinician if indicated.
- For infants with CF **without** airway reactivity or asthma, the use of inhaled corticosteroids to improve lung function or reduce exacerbations is **not** recommended.

### Topic 9: Psychosocial Needs

- Parents should be encouraged to invite supportive friends, family or caregivers that will be regularly involved in the child's care to clinic visits.
- Emotional wellness screening and resources

**Table 1: CFTR Mutations Associated with Pancreatic Insufficient (PI) and Pancreatic Sufficient (PS) Phenotypes**

Usually PI-associated CFTR Mutations		Usually PS associated CFTR Mutations
F508del	Y122X	R117H
G542X	1898+5G>T	R347P**
G551D	3120+1G>A	3849+10kbC>T
N1303K	E822X	A455E
W1282X	2751+2T>A	A455V
R553X	296+1G>C	R334W**
621+1G>T	R1070Q-S466X*	G178R
1717-1G>A	R1158X	R352Q
R1162X	W496X	R117C
I507del	2789+5G>A	3272-26A>G
394del1TT	2184insA	711+3A>G
G85E*	1811+1.6kbA>G	D110H
R560T	1898+1G>A	D565G
1078delT	2143delT	G576A
3659delC	1811+1.6kbA>G	D1152H
1898+1G>T	R1066C	L206W
711+1G>T	Q890X	V232D
2183AA>G	2869insG	D1270N
3905insT	K710X	
S549N	1609delCA	
2184delA		

\* May also be associated with PS

\*\* May also be associated with PI

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Additional resources may be found on CFTR2.com

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### III. Clinical Resources for the Newborn Infant Diagnosed with Cystic Fibrosis

#### Routine Monitoring and Care Recommendations for the Infant Diagnosed with CF:

Table I. Routine Monitoring & Care Recommendations for the Infant Diagnosed with CF													
X= do at this visit; C=consider doing at this visit				R=Review at this visit									
AGE AT VISIT →	Day of Sweat Test	24-48 hrs of dx	Ed Days (Wk 2)	Clinic Visit (Wk 3)	Clinic Visit (Wk 4)	2 mos	3 mos	4 mos	5 mos	6 mos	Monthly Clinic Visit (at 7-11 mos)	1 yr	Every 2-3 months after 1st year of life
INTERVENTIONS ↓													
<b>DIAGNOSTIC TESTING</b>													
Sweat Test	X		X*										
Fecal Elastase	X									X		X	C
Cough Swab	X				X	X	X	X	X	X	X	X	X
Chest X-ray						X						X	Q6 mos
Vitamin A, D, E & Zinc	X									X		X	
Serum Lytes,CBC, BUN, Cr	X									X		X	
LFTs, Albumin, Coags	X									X		X	
Bilirubin (conj/unconj)													
Urine Na(>30mmol/L),Cr, K			X			X		X		X	Q 2mos.	X	annual

#### Bloodwork for Newly Diagnosed Infants – 1<sup>st</sup> week

BUN, Creatinine, Lytes
AST, ALT, GGT, Bilirubin
Amylase, Albumin, Pre Albumin
Vit A, E, and 25 OH Vit D
Alk phos, Ca, Mg, PO4 Zn
IRT
Hb, CBCD
PT &INR

#### Bloodwork for Infants aged 1

BUN, Creatinine, Lytes
AST, ALT, GGT, Bilirubin
Amylase, Albumin, Pre Albumin
Vit A, E, and 25 OH Vit D
Alk phos, Ca, Mg, PO4 Zn
IRT
Hb, CBCD
PT &INR
Uric Acid
Cholesterol
Iron
IgA, IgM, IgG, IgE
Anti-Pseudomonal Antibodies

## IV. Program Summary

The implementation of the CF newborn screening program commenced in November 2009.

In order to capitalize on the benefit of screening, the aim is to see all infants within the first month of life before significant morbidity and malnutrition occurs. This early identification of infants will allow a shift in care from reactive to proactive management with the clinical focus being on respiratory, nutritional and microbial surveillance.

Recommendations for the care of newborn infants with CF have been developed and state that infants should be seen on a monthly basis. BC Children's Hospital (BCCH) and Victoria General Hospital CF clinics have developed guidelines to assist in this and ensure equitable care is received by all BC infants.

To ensure 'accessibility' to services for all newborns, the intention is to involve local pediatricians and/or family physicians as point of care contacts for interim review of newly diagnosed infants. BCCH and the Victoria General Hospital CF clinics are working with Child Health BC to support regular outreach clinics to bring CF care closer to home for those families living in northern and interior BC.

It is envisaged that care will be provided for newborn infants with cystic fibrosis that not only meets but exceeds provincial and national standards

# Part II: Discipline Specific Newborn Screening Resources

## I. Nutrition

### Nutritional Recommendations for the Infant Diagnosed with Cystic Fibrosis

*Written by: Barbara Bell, RD and Christine Loong, RD*

Infants with cystic fibrosis can be expected to achieve their growth potential and be well nourished with therapies to manage the gastrointestinal manifestations of the disease and special diet modification to meet the high nutrient requirements.

The majority of infants will have pancreatic insufficiency which may be present at birth or develop over time. Pancreatic enzyme replace therapy (PERT) enables the digestion of macronutrients thus addressing the malabsorption of both macro and micronutrients.

All infants with CF require supplementary salt to replace losses from sweat and supplementary vitamins. Energy and protein needs are elevated due to factors including malabsorption, infection and laboured respiration.

Estimated requirements for CF are 2 times the dietary reference intake (DRI) for protein and 1.2 - 1.5 times DRI for energy.

The recommended CF diet is high calorie, high protein, and high fat with added salt and multivitamin supplements.

This diet is quite opposite to what is considered a healthy diet for the general population. Families and others need to understand that what is “healthy” for the CF individual may differ from “healthy foods” for others.

### **Pancreatic Enzyme Replacement Therapy (PERT)**

Majority of infants will require pancreatic enzyme replacements to enable digestion and absorption of nutrients.

Enzyme preparations are available as enteric coated spheres encased in gel capsules. Older children will swallow the caps intact. Infants require the spheres to be taken at the required dose and mixed with a small amount of applesauce to be administered on the tip of a flat spoon.

Enzymes must be given with feeds, immediately prior to feeding. It takes practice and patience to establish a successful technique. Parents benefit from demonstrations and (repeated) supervision by a pediatric nurse.

### **Practice Points:**

- The enteric coated spheres must not be crushed or broken (or chewed). Re-feed any enzyme spheres which have been spit out to ensure the full dose is swallowed. A thorough mouth check (using a finger) is important to avoid the risks of any spheres remaining in the infant's gums or mouth.
- Applesauce is the recommended vehicle to administer enzymes. Baby's natural tongue thrust may cause parents to assume a dislike and they may resort to trying other foods (with no better success). Advise persisting with applesauce.
- Any enzymes which linger in the mouth, on lips or face (if spit out) or on mom's nipples can break down and irritate the skin possibly causing fissures. The consequence is very uncomfortable (especially for a nursing mom) and may disrupt feeding. The mouth check is very important with every feeding. Application of Vaseline around the baby's mouth offers further protection and Vaseline on mom's nipples will protect her also.
- Enzymes may remain active for up to 2 hours; therefore, repeat dosing is not advised if baby feeds again within 2 hours of taking enzymes.
- Enzyme dose prescription will be adjusted over time based on the infant's growth, volume of feeds and symptoms of malabsorption.
- See also: A Parents Guide for Giving Enzymes to Infants (appendix).

### **Vitamins**

Infants with CF require higher than normal amounts of vitamins, most notably the fat soluble vitamins A, D, E & K. Multivitamin supplementation is prescribed for all infants with pancreatic insufficiency..

### **Practice Points:**

- Fat soluble vitamin should be administered with pancreatic enzymes for optimal absorption
- Parents should be advised of careful handling with fat soluble vitamin supplement as it can stain clothing, carpets and furniture.

### **Salt**

Infants and children with CF are at risk of hyponatremia and hypochloremia due to salt loss in sweat. Breast milk and infant formulas will not meet the high needs

of salt for CF. Sufficient salt status can be measured with a urinary sodium >30mmol/L.

Supplementation of sodium and chloride is required to balance the excessive losses. Once established on solid foods, salt can be added to the child's meals. However, for infants, salt must be given in liquid form.

The recipe for home preparation of a salt solution is provided to parents (appendix). A prescribed dose is given by dropper or syringe or mixed with expressed breast milk or formula.

\*Highest risk occurs when the infant is sweating more than usual i.e. with fever, in hot environments or when over bundled. Vomiting and diarrhea also increase the risk of salt depletion. Parents must be advised to contact the physician at the onset of fever, vomiting, irritability or weakness.

**Practice Points:**

- Given the taste of the salt supplementation, some infants reject the solution. The daily dose may be divided and spread over the day in 2 - 4 doses mixed with expressed breast milk or formula to dilute the taste.
- Ensure adequate salt supplementation for infants in situations that can increase sweat and salt losses, such as over-bundling, and living in or travelling to a hot climate
- **\*\*Prompt medical attention** to potential hyponatremia and hypochloremia is essential for an infant presenting with fever, vomiting or diarrhea.



## First Six Months

Prior to diagnosis and the start of pancreatic enzyme replacement therapy, many CF infants will have large or voracious appetites and feed very frequently (ever 30 to 90 minutes). Breast feeding moms may be exhausted and some may think of discontinuing nursing. Support for breast feeding is important to provide at diagnosis.

Breast feeding on one side only during a single feeding session is advised to ensure the infant receives hind milk for maximum calories and satiety.

If not breast fed, most babies can grow well on regular cow's milk-based infant formula. The use of concentrated formulas, hydrolyzed formulas, soy formulas or low iron formulas is not indicated unless in certain circumstances. Encourage ad lib bottle feeding based on the infant's appetite and satiety cues.

Hunger, frequency of feeding and volumes consumed will change when enzyme therapy begins. It is helpful to advise parents in advance they will observe the child feeding less yet can expect better weight gain as the enzymes allow the milk to be better digested and utilized.

### **Practice Points:**

- Exclusive breast milk or formula feeding will meet the CF requirements for the first six months.
- Ensure formula preparation directions are followed correctly to provide the accurate standard dilution and use of proper hygiene and storage.
- Enzymes are given by spoon with applesauce before feeding.
- Enzymes must not be mixed into a bottle of milk.

## Solid Foods

Guidelines for the introduction of solid foods are basically the same for the CF infant as for others. Breast milk or formula is indicated for the first six months. Signs of readiness are ability to support the head, sit with support, showing more interest in food, and being hungry after a good feed. The infant is then developmentally and physiologically ready for diet progression.

A source of iron is needed so use of iron-fortified infant cereal and meats should be introduced first. Vegetables, fruits, other foods can be introduced following the usual infant feeding guidelines.

Salt should be sprinkled onto solids, then introduction of high salt foods is encouraged when age appropriate. At approximately 12 months of age, when the infant has sufficient intake of solids, salt solution can be discontinued.

Small amounts of fats (butter, margarine, oil) may be added to meals to increase the caloric density and to have the child develop a taste for the extra fat.

The amount of added fats should be limited in the infant's diet to not reduce appetite for other foods. It is suggested to add 1 teaspoon (5 mL) of fat (such as butter, margarine, canola or olive oil) for every ½ cup (125 mL) of solids. Other high calorie additions such as cream or milkshakes are not indicated for infants.

**Practice Points:**

- Solids should be introduced between 4-6 months of age. Parents must be assured that early introduction is counterproductive as the solids displace the more nutrient dense milk intake.
- As with the recommendation for all infants, honey, cow's milk, and foods that can cause increased risk of choking (such as hard foods or large pieces) should not be introduced before the age of 12 months.
- On line CF forums and other information sources often lead parents to over-loading calories into the young child's diet using large amounts of cream, milkshakes, Pediasure and even describe tube feeding options. Parents can get caught up in pushing calories and lose perspective on the goal of developing age appropriate eating.
- "Empty calorie" foods such as juice, pop, candy, etc. are not advised for the CF (or any) child as a means of boosting energy intake.

## A Healthy Eating Environment

Nutrition is very important for the health and long-term prognosis of a CF child. When parents are made aware of the importance of their child's diet, they can be very motivated to ensure the child eats as well as possible. Ongoing nutrition counseling by health care providers seeks to enable parents and children to strategize for the many challenges that can occur which range from the universal 'what to feed', finding time to plan and prepare, dealing with certain food refusals, dawdling at meals, school lunches, etc. to the physical challenges presented by the disease which disrupt appetite, intake or absorption.

Health professionals must recognize the pressure that can be put on parents by closely monitoring growth and reinforcing the importance of good nutrition.

The focus and emphasis on diet coupled with anxiety about the disease can lead to stressful mealtimes and negative interactions between parent and child. Parents should be reassured that hovering over their child, coaxing them to eat, catering to their demands, or spending long period of time over a meal is not helpful and will often exacerbate negative mealtime behaviour.

Positive feeding behaviors should be encouraged. Studies on children with CF show providing positive reinforcement to appropriate eating behaviors (e.g. trying new foods, eating independently) and minimizing attention to poor eating behaviors (e.g. food refusal) results in better intake. Proactive nutrition counseling helps enable families to develop and maintain a healthy eating environment.

### Practice Points:

- Give praise or positive comments for positive mealtime behavior.
- Establish a meal time routine and limit meals to 20 – 30 minutes. Grazing should be discouraged.
- Encourage an eating environment where the child eats together with the family. All distractions such as television, toys, etc. should be removed during meals.
- Feeding strategies should be consistently approached from all caregivers.

### References

1. Borowitz D, Baker RD, Stallings V. Consensus Report on Nutrition for Pediatric Patients With Cystic Fibrosis. *J Pediatr Gastroenterol Nutr* 2002; 35:246-59.
2. Crist W, McDonnell P, Beck M, et al. Behaviour at mealtimes and the young child with cystic fibrosis. *J Dev Pediatr* 1994;15:157-161.
3. Satter, E. The feeding relationship. *J Am Diet Assoc* 1986;86:352-6.
4. Stallings VA, Stark LJ, Robinson KA, Feranchak AP, Quinton H. Evidence-based Practice Recommendations for Nutrition-Related Management of Children and Adults with Cystic Fibrosis and Pancreatic Insufficiency: Results of a Systemic Review. *J Am Diet Assoc* 2008;108:832-9

## II. Pharmacy

### BC Children's Hospital CF Clinic Pharmacy Recommendations for the Infant Diagnosed with Cystic Fibrosis

**Written By: Eva Cho, BSc. Pharm. (ACPR)**

#### Pancreatic Insufficiency

Infants diagnosed with pancreatic insufficiency will require enzyme supplementation in order to optimize nutritional intake and growth. Infants will be screened for pancreatic insufficiency, and if found to be pancreatic insufficient, then pancreatic replacement enzyme therapy will be initiated.

In an infant, the total daily dose would be divided among the number of times the infant feeds. Infant dosing is initiated at 500-1000 units of lipase/kg/feed to be given immediately prior to each feed. Doses should be given no sooner than 2 hours apart. Infant doses can be titrated up to a daily maximum of 10000 units of lipase/kg/day divided among the total daily feeds.

There are different types and brands of enzymes available on the Canadian market. Please note that these brands are not interchangeable, as the contents of the enzymes differ. For enzyme replacement therapy, coated enzymes are recommended over non-coated powdered enzymes due to increased gastric irritation and enzyme inactivation by stomach acid. The following table lists the coated enzyme preparations that are covered by Pharmacare's Cystic Fibrosis Plan – Plan D.

<b>Contents of Coated Pancreatic Enzymes on Pharmacare Plan D Formulary</b>			
<b>Brand Name</b>	<b>Lipase (USP units)</b>	<b>Amylase (USP units)</b>	<b>Protease (USP units)</b>
Pancrease MT4	4200	17500	10000
Pancrease MT 10	10500	43750	25000
Pancrease MT 16	16800	70000	40000
Cotazym ECS 8	10800	42000	45000
Cotazym ECS 20	25000	100000	100000
Creon Minimicrospheres Micro	5000*	5100*	320*
Creon 10 Minimicrospheres	10000*	11200*	730*
Creon 25 Minimicrospheres	25000*	25500*	1600*

\*enzyme amount expressed as Ph. Eur units

Close monitoring is required to ensure that the patient's dose is appropriate.

### Monitoring for Efficacy:

- Weight gain and growth
- Decreased diarrhea, abdominal cramping and flatulence
- Decreased steatorrhea

### Monitoring for Side Effects:

Common:

- Abdominal pain, nausea (with initiation of therapy, and with dosage titration)
  - Start low, and increase doses slowly to improve tolerance
- Constipation
  - May require reassessment of dose (is dose too high?)
- Diarrhea
  - May require reassessment of dose (is dose too low?)
  - Monitor for diaper rash/perianal irritation, sore bottom
- Oral mucosal irritation
  - Do not crush, chew or leave enzyme beads in the mouth. Sweep child's mouth after enzyme administration to ensure no beads remain.
- Headache, dizziness
- Allergy/hypersensitivity to porcine (pork) protein – rash, pruritis, urticaria, anaphylaxis

Rare:

- Colonic strictures (reported at high doses doses > 6000 units of lipase/kg/meal)
- At high doses: hyperuricemia → monitor for signs of joint swelling, painful joints, cloudy urine

### Vitamin Supplementation

In addition to dietary supplementation with formulas to ensure adequate nutrition, infants with pancreatic insufficiency also require vitamin supplementation, particularly the fat soluble vitamins – vitamins A, D, E, and K.

AGE	Vitamin A (IU)	Vitamin E (IU)	Vitamin D (IU)	Vitamin K (mg)
0 – 12 months	1500	40 – 50*	400-500	0.3-0.5

There are currently two vitamin products formulated containing the higher content of fat soluble vitamins. Both products are available only through the Health Canada Special Access program:

- **MVW Complete Formulation Multivitamins<sup>®</sup>** Liquid, Chewable tablets, Gel capsules
- **DEKAs Vitamins<sup>®</sup>** Liquid, Chewable tablets, Gel capsules

For infants, the pediatric liquid formulation would be prescribed:

Age			<b>MVW<sup>®</sup> Pediatric Drops</b>
0-12 months			0.5 mL daily

For full prescribing information, please refer to the product website.

Currently only the MVW<sup>®</sup> products are a benefit under the Pharmacare Plan D: Cystic Fibrosis Formulary. DEKAs products may be added to formulary. For the most current information, please refer to the Pharmacare website for Cystic Fibrosis Formulary.

Despite being supplemented with the combination products, some children may still be deficient in the fat soluble vitamins, and may require additional supplementation of that specific vitamin as indicated by laboratory monitoring. Other supplements that may be required if insufficient in diet are calcium, magnesium, iron, and zinc.

### Salt Supplementation

This is important in breast fed or formula fed infants with CF to prevent hyponatremic alkalosis (Pseudobartters Syndrome). Patients with CF are at increased risk for hyponatremia as a result of salt loss through the skin, especially in the hot summer weather.

- Salt supplementation: 2-4 mEq/kg/day (likely requiring higher end of dosing range)

## References

1. Borowitz D, Baker RD, Stallings V. Consensus Report on Nutrition for Pediatric Patients With Cystic Fibrosis. *J Pediatr Gastroenterol Nutr* 2002; 35:246-59.
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3. MVW<sup>®</sup> Complete Formulation Product Information: <http://mvwnutritionals.com/>
4. Pharmacare Cystic Fibrosis Plan D: <http://www2.gov.bc.ca/gov/content/health/practitioner-professional-resources/pharmacare/pharmacies/product-identification-numbers/cystic-fibrosis-supplies-pins>
5. Tangpricha V et al. An Update on the Screening, Diagnosis, Management and Treatment of Vitamin D Deficiency in Individuals with Cystic Fibrosis: Evidence-Based Recommendations from the Cystic Fibrosis Foundation. *J Clin Endocrin Metab*. 2012; 97(4):1082-93.

## III. Physiotherapy

### BCCH CF Clinic Physiotherapy Recommendations for Management of the Infant Diagnosed with Cystic Fibrosis

*Written By: Nicole Lee-Son, BScPT and Maggie McIlwaine, PhD.*

Physiotherapy assists with the prevention and retardation of lung damage in the infant diagnosed with cystic fibrosis. Lung damage in cystic fibrosis is the result of several contributing factors including increased secretion retention, airway obstruction, inflammation and infection.

Prior to newborn screening, the diagnosis of cystic fibrosis was primarily made based on symptomatic presentation. This presentation often included respiratory compromise and lung changes on chest radiography. Identifying infants with cystic fibrosis through newborn screening allows the initiation of physiotherapy treatment prior to the manifestation of respiratory symptoms.

The objective of physiotherapy treatment for individuals with cystic fibrosis is to assist with mucus clearance from the lungs and maintain normal development.. There are several airway clearance techniques that can be considered for physiotherapy treatment for the infant with cystic fibrosis. At BC Children's Hospital, Modified Postural Drainage with Percussion and Vibration is primarily used for children under the age of 5 years. Assisted Autogenic Drainage and Baby Positive Expiratory Pressure may also be used. Exercise and activities that encourage gross motor development are also encouraged to assist with lung health and development. Where indicated, the correct use of inhaled medications such as bronchodilators or mucolytics and mode of delivery is taught to caregivers.

Physiotherapy treatment for the infant with cystic fibrosis highly relies on the commitment of the caregiver. Initiating physiotherapy techniques upon diagnosis allows the caregiver to become attune with the respiratory status of the infant. If the caregiver performs physiotherapy on a daily basis, the caregiver will likely be able to detect minute changes from baseline allowing for appropriate care. Airway clearance techniques become part of the infant's daily routine which sets an important precedence as the infant grows into adulthood.

#### Modified Postural Drainage with Percussion and Vibration

- Modified Postural Drainage; the infant is placed in specific positions that utilize ventilation patterning to assist with mucus clearance.
- Percussion and Vibration; the caregiver provides an external mechanical force through percussion and vibration over specific areas of the ribcage and lung to assist with mucus clearance.



### Assisted Autogenic Drainage

- The caregiver applies an external force over the ribcage and lungs to alter the volume and flow rate of inspiration and expiration of the infant to assist with mucus clearance. This may be assisted by use of a Therapy ball.

### Assisted huffing.

- The caregiver applies an external force over the rib cage only during expiration to increase the expiratory flow rate which assists with mucus clearance. This may be assisted by use of a Therapy ball.

### Baby Positive Expiratory Pressure

- The caregiver assists the infant through use of a facemask to breathe out against a resistor which creates positive expiratory pressure. This is combined with assisted huffing, which assists with mucus clearance.

### Exercise and Gross Development

- The baby's development is assessed at each clinic visit to ensure they are performing age appropriate activities.
- Specific ideas for the infant with cystic fibrosis include activities that both promote gross motor development and encourage the infant to take deep breaths. Thus activities that focus on stretching or opening the chest and trunk rotation are encouraged.

At present, families are taught Modified Postural Drainage with Percussion and Vibration as the initial physiotherapy airway clearance technique. Treatment starts immediately after diagnosis and consists of a total of 9 Modified Postural Drainage positions in which percussion and vibration is performed for 3-5 minutes in each position. Physiotherapy sessions are performed 3 times a day and each session addresses 3 of the 9 positions.

### References

1. Lannefors L, Button BM, McIlwaine M. Physiotherapy in infants and young children with cystic fibrosis: current practice and future developments. *J R Soc Med* 2004;**97**(Suppl. 44):8-25.

**\*The following document describes the difference between a cough swab and throat swab. If a patient is unable to expectorate sputum, the cough swab becomes the ideal laboratory test for cystic fibrosis patients who require frequent microbial surveillance of the lungs.**

## Definition of a Cough Swab

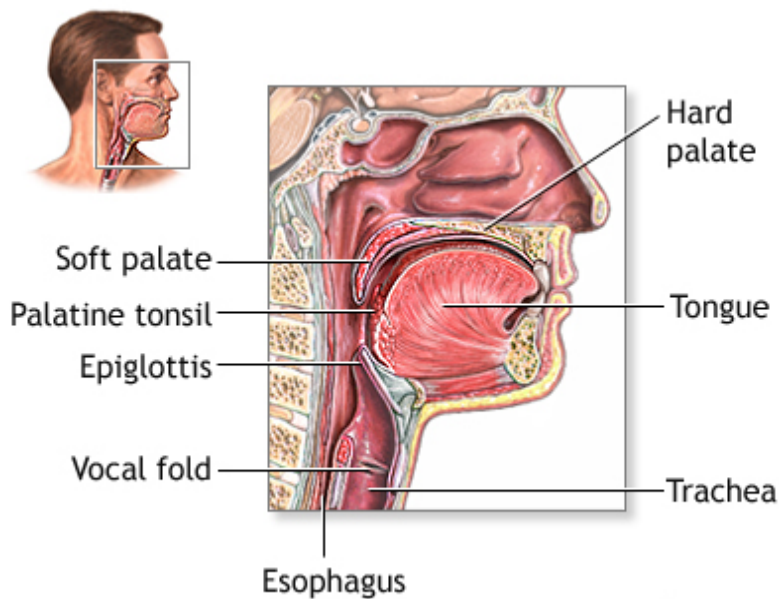
A cough swab is a laboratory test done to identify germs in the lungs that may cause infection in the lungs. It is a strong predictor of a sputum culture<sup>1</sup>.

### How the test is performed

The ability of the test to detect pathogens in the lungs is enhanced if the cough swab is performed after a Physiotherapy session or the administration of hypertonic saline<sup>1</sup>.

The patient should be instructed to tilt their head back and open their mouth wide. A sterile cotton swab is placed at the back of the throat under the Uvula, not touching the posterior pharynx. The patient is then instructed to cough. They need to resist gagging and closing the mouth while the swab is in their mouth.

If the patient is too young to co-operate with a cough, then you may place the swab against the posterior pharynx and stimulate a cough.



ADAM.

Structures of the throat include the esophagus, trachea, epiglottis and tonsils.

## 1. Use of cough swabs in a cystic fibrosis clinic

A C Equi<sup>a</sup>, S E Pike<sup>b</sup>, J Davies<sup>a</sup>, A Bush<sup>a</sup> <sup>a</sup>Department of Paediatric Respiratory Medicine, Royal Brompton & Harefield NHS Trust, Sydney Street, London SW3 6NP, UK, <sup>b</sup>Department of Physiotherapy, Royal Brompton & Harefield NHS Trust .Dr  
Busha.bush@rbh.nthames.nhs.uk

### Abstract

We audited prospectively 322 cough swabs taken from cystic fibrosis children and compared cough swabs with concomitant sputum samples in 30 expectorating patients. A positive cough swab is a strong predictor of sputum culture. However, a negative cough swab does not rule out infection. Persistent symptoms should be further investigated.

## 2. Clinical value of obtaining sputum and cough swab samples following inhaled hypertonic saline in children with cystic fibrosis.

[Ho SA](#), [Ball R](#), [Morrison LJ](#), [Brownlee KG](#), [Conway SP](#). [Pediatr Pulmonol](#). 2004 Jul;38(1):82-7.

### Abstract

Prompt detection and treatment of lower respiratory tract infection are essential in the management of patients with cystic fibrosis (CF), who often have signs or symptoms of respiratory infection without any pathogens being isolated from sputum or cough swab specimens. The aims of this study were to assess the efficacy and clinical value of obtaining sputum and oropharyngeal cough swab samples following induction with hypertonic saline (HS) in this group of patients. Forty-three outpatients with CF, mean age 7.2 years (range, 1.8-12.9 years), were recruited over a 2-year period. Nebulized salbutamol was administered, followed by 6% HS. Sputum was preferentially obtained before and after HS induction if possible. If the patient was not able to expectorate, oropharyngeal cough swabs were taken instead. Four patients were able to expectorate sputum before and 19 after HS induction. The procedure was tolerated in 41 of 43 patients. Pathogens were isolated from 13 patients' HS-induced samples, but not from their corresponding preinduced specimens, and 4 patients' preinduced specimens cultured organisms which were not identified from their HS-induced samples. Significant changes were made in the management of 13 (30.2%) patients directly resulting from the positive culture of pathogens only from HS-induced samples. Cultures from oropharyngeal cough swab or expectorated sputum specimens following inhalation of HS provide additional microbiological information which is of clinical value and may lead to changes in patient management.

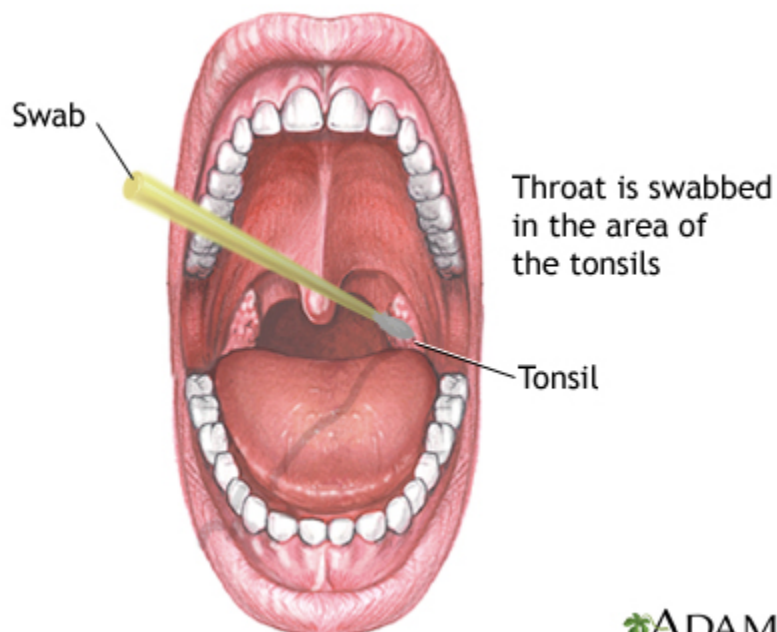
## Definition of a Throat swab culture

A throat swab culture is a laboratory test done to identify germs that may cause infection in the throat. It is most often used to diagnose strep throat.

### How the Test is Performed

The patient should be instructed to tilt their head back and open their mouth wide. The health care provider rubs a sterile cotton swab along the back of their throat near the tonsils. The patient needs to resist gagging and closing the mouth while the swab touches this area.

The health care provider may need to scrape the back of the throat with the swab several times. This helps improve the chances of detecting bacteria.



ADAM.

A throat swab can be used to determine if Group A Streptococcus bacteria is the cause of pharyngitis in a patient.

M. McIlwaine

## IV. Psychiatry

### BCCH CF Clinic Recommendations for Detecting and Managing Psychiatric Difficulties in Parents of Infants Diagnosed With Cystic Fibrosis

**Written By: Patrice Dunn, MD,FRCP (Paediatric and Family Psychiatrist)**

Psychiatric disorders, in particular Depression and Anxiety Disorders, are very common in the general population and higher in patients experiencing major stressors. As this would include parents of a CF baby diagnosed through newborn screening, it is recommended that the initial assessment by the CF team include the following:

#### 1. Personal and family psychiatric history

These are the most powerful predictors of psychiatric disorders, in particular for Depression. This information is perhaps best obtained when a general medical family history is taken, and we suggest it include as a minimum histories of:

- a) Depressive illness, including Post-Partum Depression
- b) Anxiety disorders
- c) Substance Misuse

#### 2. Informal Assessment

For indications suggestive of a psychiatric disorder in the parents, we recommend that this be done by the NBS nurse, but that all CF team members inform the NBS nurse of worrying changes in parental behavior or emotional presentation. While most parents will find this a stressful time, there are some highly specific signs of a Major Depression which differentiate it from situational depressive symptoms.

These include:

- a) Loss of appetite, often with weight loss - the person has to force themselves to eat.
- b) Anhedonia – the loss of enjoyment in things that they usually enjoy.
- c) Inability to feel – the person has a sense of having little or no feeling for friends and family.
- d) Difficulty initiating any activities – the person has to force themselves to do even routine tasks.
- e) Early-morning wakening – the person wakes very early and is unable to get back to sleep.

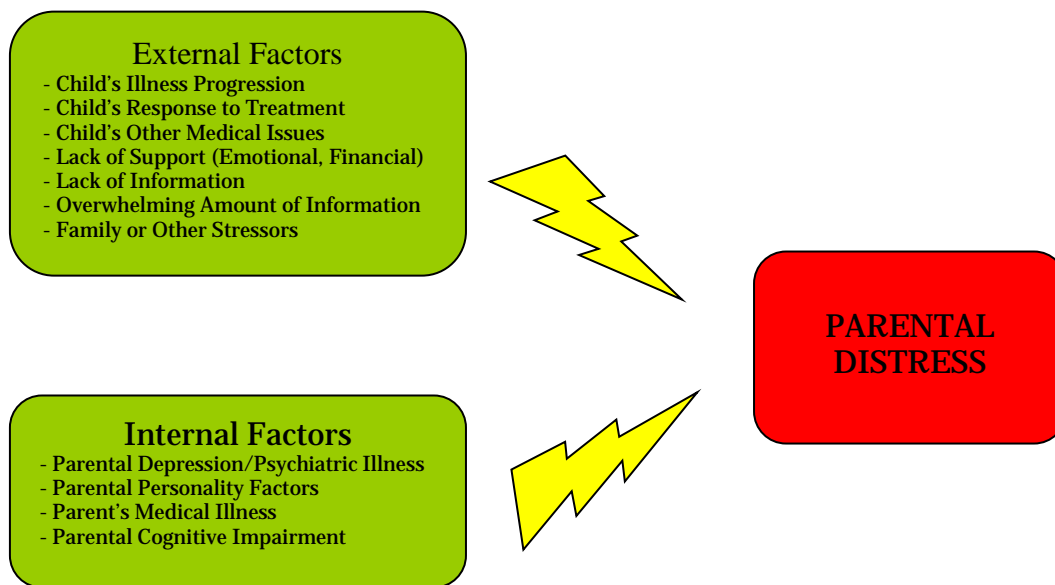
These symptoms are persistent over weeks to months, and are a significant change for the person. Should these symptoms be seen in the first few weeks after diagnosis, they should be put in the context of the enormous impact this diagnosis will have on any parent of a newborn.

There are also some **Very Serious** signs & symptoms which require urgent intervention whether or not depression is present:

- a) Suicidal thoughts and especially plans.
- b) Thoughts of harming their baby.

If this informal assessment raises any concerns about a possible Major Depression in a parent we would suggest they contact their family doctor, and may try to arrange referral to a psychiatrist.

**Some of the factors we have found to contribute to parental distress:**



We also suggest that vigilance for and informal assessment of psychiatric signs and symptoms in parents of newborns diagnosed with CF be part of **all** clinic visits. Specifically regarding Depression, while we find that increased awareness and knowledge about this illness by the team is most effective at picking it up early, there are numerous rating scales available which are used to screen for Depression and Post-Partum Depression. Most such scales are designed as research tools, but the Montgomery-Asberg Depression Rating Scale ( [http://www.psy-world.com/madr\\_s\\_print1.htm](http://www.psy-world.com/madr_s_print1.htm) ) which is a **clinically** useful scale which includes the more specific indications of Depression.

### **3. Psychiatric Consultation**

We recommend if possible having a Paediatric & Family Psychiatrist available to consult with the CF team, not only at the time of initial assessment but on an ongoing basis. Specifically regarding psychiatric difficulties in parents, it is ideal to have this consultant available for indirect consultation with the team, to assist in determining whether psychiatric difficulties are present, and to assist the team in providing optimal support for the family as well as to provide or help arrange treatment for parents with these difficulties.

# V. Social Work

## Social Work Services with NBS Infants and Their Families

**Written By: Linda MacNutt & Tami Kolb, Social Work, BCCH**  
**Edited by Amy Schellenberg, Nurse Clinician, BCCH**

“Good medical care is vital, but unless the root social causes that undermine people's health are addressed, the opportunity for well being will not be achieved.” - *WHO Commission on the Social Determinants of Health (2006)*

## Important Considerations and Assessment Tools

### Social Work Role:

- I. Family Assessment
- II. Family Intervention
- III. Team Member

### I. Family Psychosocial Assessment

- Names & Ages of Parents & Siblings
- Occupations
- Living Arrangements – where do they live & with whom
- Financial Issues
- Social Relationships/Support
- Sources of Stress
- Comprehension
- Mental Health History
- Belief Systems – religious, cultural & medical
- Treatment Challenges
- Family Needs/Preferences
- Other Risk Factors

### Barriers to Learning, Coping & Adherence

1. Single parent
2. Young or first time parents
3. Financial constraints
4. Parental Mental Health - post- partum depression, anxiety
5. Substance Usage
6. MCFD involvement / Child Protection Concerns
7. Parental cognitive limitations making it difficult to process information
8. Relationship Issues
9. Lack of Support
10. Belief System – medical, spiritual, cultural, philosophical
11. Poor Coping Skills
12. Parental Physical Health



## II. Family Intervention

### 1. Supportive & Adjustment Counseling -- Emotions

- Variety of Emotions
- Grieving Process
- Guilt -- both rational and irrational
- Living with the unknown – control issues
- Normalizing feelings
- Encouraging the family to enjoy their baby
- Reactions of Family & Friends
- Stressing importance of self-care

### 2. Social Support Interview

- What have you been told?
- When did you know it?
- What do you understand?
- Who to tell?
- What information to tell?
- What do you need right now?

### 3. Clinical Management of Stress, Shock and Uncertainty

- Personal resources for: self, each other, children, extended family, friends
- Assessment of hopefulness/helplessness
- Community resources
- Hospital resources

### 4. Assessment of Information Seeking Sources

- Health professionals: primary/specialty services
- Family, friends, friends of friends
- Online

### 5. Financial Assessment

#### Funding, Education, Handouts, Pamphlets

- Practical resources – travel related resources
- Financial – Fair PharmaCare, extended benefits, CF Foundation Grant
- Appropriate Handouts

## III. Liaising with the Cystic Fibrosis Team Members

- Importance of a multidisciplinary team approach

## Newborn Screening Program

### New Diagnosis: Adapting Family Life to a New Diagnosis

#### 1. Information/Exploration

- Education: learning needs
- Comprehension
- Assessment of current preferences

#### 2. Coping with a New Diagnosis

- Parental expectations/hopes/goals
- Potential stress points: medications, physiotherapy, diet, hospitalizations, travel, etc.
- Financial impact/financial resources
- Shared parenting: “who, what, how & why?” working and communicating with the health care system

#### 3. Telling Your Family: Immediate & Extended

- “Giving the news”: how, when & where?
- Managing the impact
- Education
- Comprehension
- Support

#### 4. Stress Management/ Resilience

- Resilience inventory: individual & parental normalization challenges
- Social support inventory
- Financial resources
- Community, provincial, national & international organizations

#### 5. Your Child’s Growth

- Developmental milestones: physical, cognitive, behavioral, emotional

#### 6. Parental Challenges

- Normalization
- Coping, vicarious hope/despair = stress point interventions
- Adherence to medical regimes
- Developmental milestones in relation to CF

#### 7. Navigating the Health Care System

- Being an advocate
- Who & when to talk to a team member
- Partnership/ Team -- Never alone
- Navigating the changes – both system and child’s health

## Mental Health Screening

Mental health is a very important part of overall health. In 2015, the U.S. CF Foundation and European CF Society published a consensus statement for screening and treating depression and anxiety in CF. This was based on a large research study which showed that there are increased rates of both depression and anxiety in individuals with CF and their caregivers (<http://www.cysticfibrosis.ca/about-cf/virtual-education-program-for-patients-and-caregivers/webinar-new-international-guidelines-on-mental-health-in-cf/>).

We are presently screening caregivers of babies 3 months following diagnosis and when the infant is 1 . If indicated, screening will be done more frequently. Screening is conducted by administration of 2 brief validated questionnaires, GAD-7 and PHQ-9. Based on the results, supportive resources and/or referrals are made.

