

British Columbia Provincial Newborn Screening Program

Guidelines for Management of the Infant Diagnosed with Cystic Fibrosis

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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 have a responsibility to provide the highest standard of care so that a new generation of children with CF may fully benefit from future CF cures**

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 for CF commenced in the fall of 2009. In planning for appropriate resource allocation, the CF screening algorithm was used to predict the number of children required to attend BC Children's Hospital (BCCH) for further assessment and sweat testing. It is estimated that 101 children will be required to attend BCCH for evaluation each year. Of these, at least 17 infants will be diagnosed with CF.

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 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

On reviewing the distribution of British Columbia's population, it is envisaged that the majority of patients will be from the lower mainland; however, there will be families identified from Vancouver Island and Northern/Interior BC. To ensure continuity of care and equitable resource provision, all patients in BC will be contacted by the Newborn Screening Nurse (Figure 2). 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. Following the sweat test, the outcomes are as follows.

Lower Mainland

Sweat Test Outcome:

- Negative: Discharged to the primary care provider. Consult letter supplied.
- Positive: Transferred to BCCH CF clinic for training/education. Consult letter given to the primary care provider.
- Borderline: Transferred to BCCH CF clinic for further assessment: Consult letter given to the primary care provider.
- Insufficient: Repeat sweat test arranged

Vancouver Island

Sweat Test Outcome:

(Applies to infants with one mutation found on genetic screening that require sweat testing. Those with two mutations are referred directly to the Victoria Clinic for evaluation and follow up)

- Negative: Discharged to the primary care provider. Consult letter supplied.
- Positive: Referred to Victoria CF clinic for training/education: Consult letter given to the primary care provider
- Borderline: Referred Victoria CF clinic for further assessment: Consult letter given to the primary care provider
- Insufficient: Repeat sweat test arranged

Northern BC/Interior

Sweat Test Outcome:

- Negative: Discharged to the primary care provider. Consult letter supplied.
- Positive: Transferred to BCCH CF clinic for training and education: Consult letter given to the primary care provider.
- Borderline: Transferred to BCCH CF clinic for further assessment: Consult letter given to the primary care provider
- Insufficient: Repeat sweat test arranged

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 identify and train local physiotherapists, dieticians, social workers and other members of the allied health team to assist in the care of the newborn infants. This will ultimately lead to the development of a CF clinical network.

The proposed follow-up schedules for infants outside of the lower mainland are highlighted in Figure 3.

Multidisciplinary Roles for the newborn infant

For the majority of infants with CF the initial evaluation, diagnosis and teaching will be performed at BCCH. To ensure continuity, appropriate management, and avoid duplication of roles; team members will have input at specific times. This may alter dependent on the infant's clinical status as detailed below.

Initial Infant assessment

Who: Newborn Screening Nurse and Physician +/- Social worker

Role: Formal clinical assessment

Basic genetic counseling provided

Identification of psychosocial needs

Post Diagnosis: Visit 1

Who: CF Multidisciplinary Team

Role: CF Diagnosis counseling

Identification of psychosocial needs

Investigations as per protocol

Nutritional Assessment

Physiotherapy Assessment

Provide options for financial support

Initiation of pancreatic enzymes as indicated, prophylactic antibiotics,

Vitamin D & E.

CF Teaching

Who: CF Multidisciplinary Team

Role: As per protocol

Medication Review

Telephone Follow-up

Who: Newborn Screening Nurse and/or Dietician

Role: Review clinical status

Review medication

Review feeding/enzyme dosage

Liaison/feedback from local Pediatrician

Local Pediatrician Follow-up

Who: Identified Pediatrician/Family Physician

Role: Clinic assessment as per Performa/clinical resource pack

Weight and cough swab

CF clinic visit

Who: Newborn Screening Nurse and CF Multidisciplinary team

Role: Full review as per clinic sheet

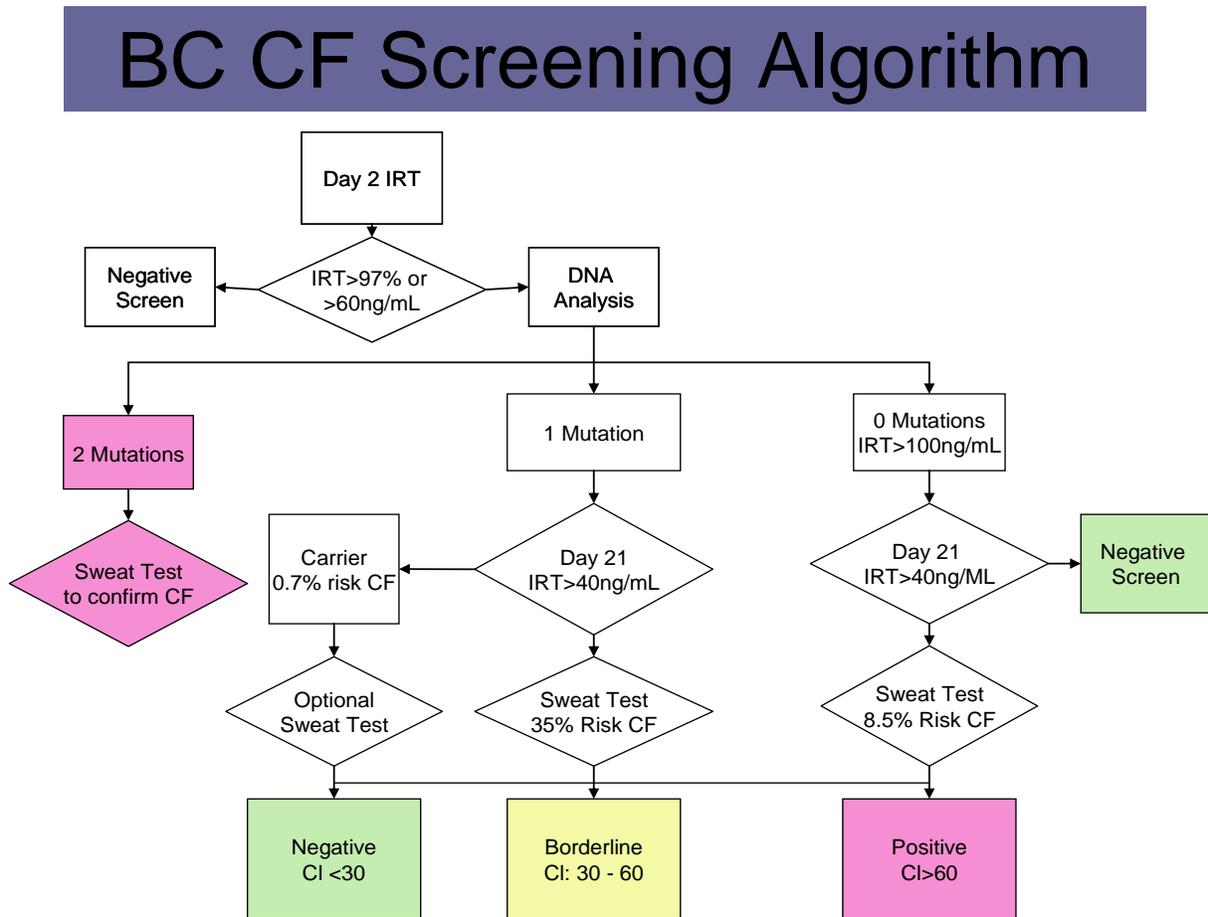
Cough swab and weight

Additional investigations as per initial evaluations.

At 1 year of age or when clinically appropriate full handover of infant from newborn screening clinic nurse to general CF nurse.

Figure 1

Newborn Screening Algorithm



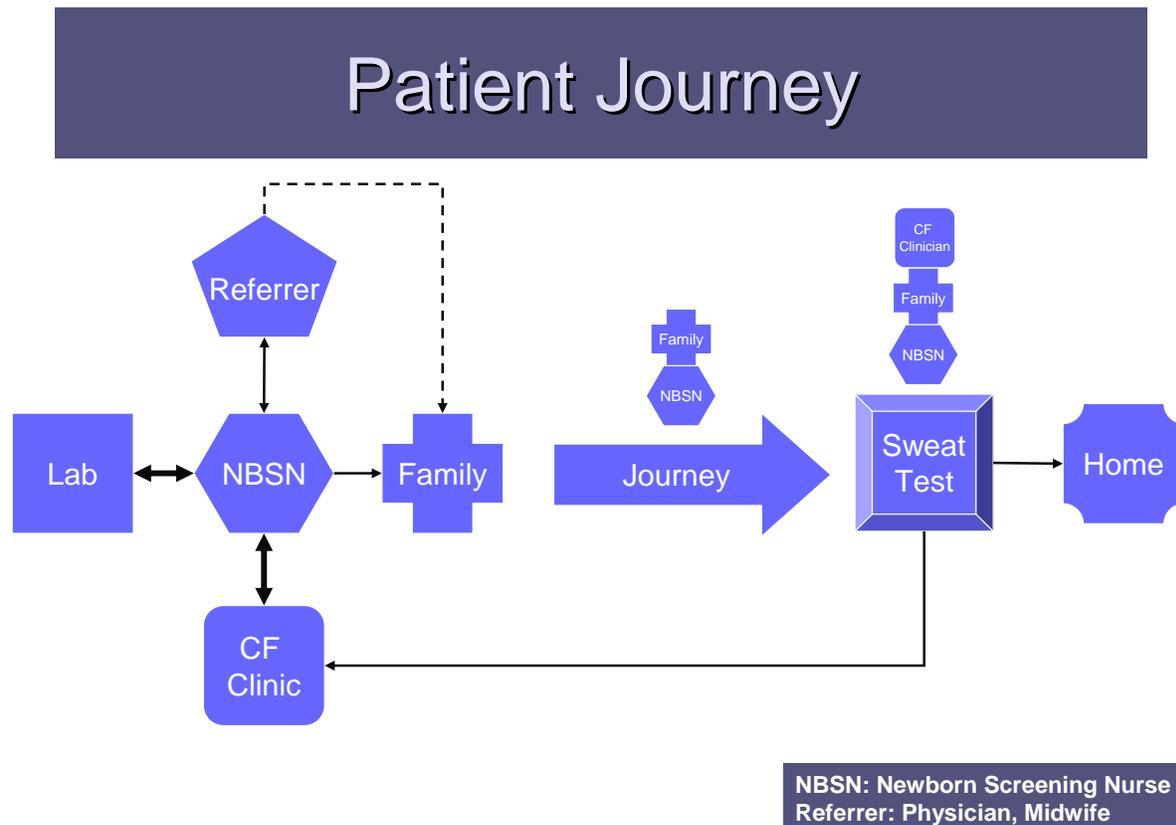
*3-5% False Negative Rate

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

*Sweat test borderline range is 40-60 for children 6 months or older

Figure 2

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 3

Schedule for Infants diagnosed with Cystic Fibrosis

Diagnosis, Education and Follow-up for Newborn Infants with CF from BC					
Week	Mon	Tues	Wed	Thurs	Fri
1		■ ■	■	■ ■	■
2	■	■	■		■
3		■			
4		■			
Month					
2		■			
3		■			
4		■			
5		■			
Key					
■ ■	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 consultation in conjunction with a local GP or pediatrician visit, as deemed appropriate by the CF team/family.

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 may be appropriate after diagnosis to discuss the implications for family members and perform genetic testing, if desired.
- Information to be provided at each clinic visit: See Table I

Topic 2: Nutrition

It is recommended that:

- Pancreatic functional status be measured by fecal elastase in all infants at the initial visit and, if normal, 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 II).
 - 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 1000 lipase units/kg/meal with a maximum of 8 doses in 24 hours. 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. Doses may be titrated up to a maintenance dose of 2500 lipase units/kg/meal with a maximum daily dose of 10 000 lipase units/kg/day.

Topic 3: Feedings, Vitamins and Micronutrients

- Human milk is preferred as the initial type of feeding

- For infants with CF under two years of age who drink formula, standard infant formulas (as opposed to partially or extensively hydrolyzed protein formulas) should be used.
- 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 patients with CF should be prescribed by the CF physician, shortly after diagnosis if albumin levels are within the normal range(>35g/l).
- Blood levels of fat-soluble vitamins should be measured at diagnosis, approximately two months after starting vitamin supplements, 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 begin eating sufficient amount of solid foods (approximately 8-9 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 usually performed 2 – 3 times daily. Several airway clearance techniques may be used which include modified percussion and postural drainage, 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.
- For infants with CF under one year of age, the use of palivizumab is recommended for prophylaxis of respiratory syncytial virus in keeping with the recommendations of the BC RSV Task Force.
- It is recommended that monthly cough swabs be taken for bacterial surveillance. Additional cough swabs may be required if clinically indicated.
- The prophylactic use of oral anti-staphylococcal antibiotics is recommended for the first year of life.
- 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 two unsuccessful attempts at eradication, with *Pseudomonas aeruginosa* should be treated chronically with inhaled anti-pseudomonal agent and oral Azithromycin.
- 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.

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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 ↓													
CARE ISSUES													
Discuss Diagnosis		Either visit	X	C	C								
Encourage Breast Feeding		Either visit	X	X	X	X	X	X					
Start Pancreatic Enzymes		Either visit	X	C	C	C	C	C	C	C	C	C	C
Start Salt Supplementation		X								X	R		R
Start vitamins for CF patients		Either visit											
Start Staph prophylaxis			X										
weight, length, HC; plot on WHO growth chart		Either visit	X	X	X	X	X	X	X	X	X	X	X
Vitals, SpO2 & physical exam		X	X	X	X	X	X	X	X	X	X	X	X
Teach and initiate ACT			X										
Assess weight gain, intake, salt supplementation and enzyme dosage			X	X	X	X	X	X	X	X	X	X	X
Assess respiratory systems		X	X	X	X	X	X	X	X	X	X	X	X
PCP Anticipatory Care & Immunizations, RSV			X			X		X		X		X	
DIAGNOSTIC TESTING													
Sweat Test		X	X*										
Fecal Elastase		X								C		C	C
Cough Swab		X			X	X	X	X	X	X	X	X	X
Chest X-ray						X						X	C
Vitamin A, D, E & Zinc		X				X						X	
Serum Lytes,CBC, BUN, Cr		X								X		X	
LFTs, Albumin, Coags Bilirubin (conj/unconj)		X								X		X	
Urine Na(>30mmol/L),Cr, K			X			X				X	9mos	X	
EDUCATION													
Infection Control			X	X	X					X		X	
Who to Call/Where to Go			X										
CF Patient Data Registry			X										
CFResearch/Outlook			X							X		X	
Pathophysiology of CF		X	X	X	X	C	C	C	C	C			
Genetic Implications of CF		One of these visits											
Feeding Behaviour/Guidance			X			X	C	C	C	X	X	X	C
Review ACT Techniques			X	X	X	X	X	X	X	X	X	X	
Smoke Exposure Avoidance			X										
Review Baseline Testing		X	X	X									
Review Medication List		X	X	X	X	X	X	X	X	X	X	X	X
CF Team/Roles		X	X	X	X								
Immunizations/RSV prophylaxis			X										
Supports/Resources		X	X	X	X	C	C	C	C	C	C	C	C
Pharmacare/Plan D		X											

Table 2: 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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III. Clinical Resources for the Newborn Infant Diagnosed with Cystic Fibrosis

Initial Evaluation

New Diagnosis Cystic Fibrosis



Date of Assessment:

Reason for Dx: - NBS - Mec Ileus - Family Hx - FTT - Other: _____

1. History

Date of Birth:	Place of Birth:		
1st IRT Date:	Result:	ng/mL	
2nd IRT Date	Result:	ng/mL	
Genotype:	PHN:		
Sweat Test- Date:	Sweat Cl-	mmol/L	Wt- grams
Repeat Sweat- Date:	Sweat Cl-	mmol/L	Wt- grams
Prenatal/Birth History:			
Apgar:	Birth HC:		(cm)
Birth Weight:	Birth Length:		(cm)

2. Demographics

Mother:	Age:	Occupation:
Father:	Age:	Occupation:
Marital Status:		
Address:		
Phone: (H)	(Cell)	
Email:		
Pharmacy:	Phone:	Fax:
Family Doctor:	Phone:	Fax:
Paediatrician:	Phone:	Fax:

3. Family Main Concerns

-
-
-
-

4. Genogram/Family History (including physical and mental illness)

Sibling(s) requiring sweat test? Yes () No ()

Name:	DOB:	PHN:	
	Date:	Sweat Cl:	mmol/L
Name:	DOB:	PHN:	
	Date:	Sweat Cl:	mmol/L
Name:	DOB:	PHN:	
	Date:	Sweat Cl:	mmol/L

5. Family Psychiatric History:

Any family history of depressive illness (including post partum depression), anxiety disorders, substance misuse? If yes or there is clinical concern consider using the Montgomery-Asberg Depression Scale.

Informal screen for psychiatric symptoms in parents:

Level of concern: High ___ Low ___ None ___

6. Social Screen

Reviewed by - Nurse - Social Worker

Are expenses related to the child's medical situation a concern at this time?

- Yes ___ Travel ___ Accomodation ___ Meals ___ Child Care
- No

Identify areas where there is additional support needed:

- Emotional Support - Financial Support - Baby-sitting/Respite - Other _____

Discuss: _____

Assessment of Knowledge – New Diagnosis Teaching Guide

C=Initial Teaching Completed &
Family Demonstrates Basic Working Knowledge
F=Follow up Required/More Teaching Needed
N=Not Addressed
NN=See Nurses Notes for Expanded Information
N/A=Not Applicable



Teaching Aids: Pamphlets and Handouts for the Parent Education Binder

1. Understanding CF – pamphlet	11. Managing Nutrition & Malabsorption in CF - Booklet
2. Infection Control - pamphlet	12. Vitamins and CF - Handout
3. CF in Canada - pamphlet	13. Healthy Diet in CF - Handout
4. Information on CF Clinic at BCCH – Handout	14. A Parent Guide to Giving Enzymes to Infants - Handout
5. The CF Team - handout	15. Managing Lung Problems in CF – Booklet
6. Who to call and Where to Go to Care for your Infant with CF – Handout	16. Cystic Fibrosis and Physiotherapy - Booklet
7. Carrier Testing for CF – pamphlet	17. Disability Tax Credit - Handout
8. Beginning CF Care: A Guide for Parents of Young Children - Booklet	18. Social Work Package
9. Your Child and Cystic Fibrosis - Handout	19. BC Assoc. Government Grant Policy for Financial Assistance with CF Meds - Handout
10. Nutrition for Infants with CF – Handout	20. BC Assoc. Government Grant Policy for Financial Assistance with CF Travel and Accommodation - Handout

PATIENT NAME:	Initial Consult	1st Visit	1st Ed Day	2nd Ed Day	3rd Ed Day	5th Visit	6th Visit
DATE OF VISIT							
CF EDUCATION							
(i) Testing: Baseline							
• Sweat Test							
• Stool							
• CXR							
• Throat Swab C&S							
• Blood Test							
• Urine Sodium							
(ii) Testing: As Indicated							
• Bone Age							
• Hearing Test							
• Gastric Emptying							
• Gastric Reflux Scan							
• CT Scan							
• Bronchoscopy							

	Initial Consult	1 st Visit	1 st Ed Day	2 nd Ed Day	3 rd Ed Day	5 th Visit	6 th Visit
(iii) Genetic Implications							
• Siblings Tested?							
• Carrier Testing							
• Referral for Genetic Counseling							
(iv) Pathophysiology of Systems Affected							
• Respiratory							
- Normal Lung Development							
- How CF affects the lungs							
• GI							
• Liver							
• Sweat Glands							
• Reproductive							
(v) Infection Control							
•							
•							
(vi) Prevention of Hyponatremia/Dehydration							
(vii) Medications: Rationale; Dosage; How to Administer							
•							
•							
•							
•							
•							
• Decongestants vs Saline Wash/Bulb Suction							
• Probiotics/Antibiotics							
(viii) CF Team/Roles							
• Physiotherapy							
- Role of airway clearance							
- Teach airway clearance techniques							
- Role of exercise							
- Role of mucolytics							
(ix) Government Grants							
• Medication							
• Travel/Accommodation							
(x) Immunization in CF							
• Regular Immunizations							
• RSV Prophylaxis							
• Flu Shot							

	Initial Consult	1st Visit	1st Ed Day	2nd Ed Day	3rd Ed Day	5th Visit	6th Visit
(xi) CF in the Future							
• Current CF Research							
• Future Treatments/Outlook							
• Lung Transplant							
• Pregnancy/Reproduction							
• CF Related Complications- CFRD, Osteoporosis							
CF NAVIGATION							
(i) Out Patient CF Clinic-Physical Space, How Clinic Runs, Timing, Correspondence							
(ii) Pharmacy							
• How to Choose One							
• Process of Filing Prescription							
• Drug Coverage							
(iii) Parking							
(iv) Clinic Summary- Expect Mail							
RELATIONSHIP BUILDING							
(i) When and Who to Call- CF Team							
(ii) Role of GP/ Pediatrician							
SUPPORT/CF AND THE FAMILY							
(i) Assist with Identifying/Mobilizing Support Systems							
(ii) Able to Express Grief							
(iii) Risk for Post Partum Depression							
(iv) Role of Child and Family Psychiatrist							
(v) Explore Potential for Sibling Jealousy/Setting Boundaries							
(vi) Introduce CCFF							
(vii) Provide Link with another CF parent, if wishes							
REGISTRATION							
(i) Plan D-PharmaCare							
(ii) Fair PharmaCare							
(iii)CCFF Patient Data Registry							
• Consent Form Given/Explained							
• Consent Form Returned							

IV. Program Summary

Summary

The implementation of the Cystic Fibrosis (CF) newborn screening program is now complete and screening in BC 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 pro-active 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 identify and train local physiotherapists, dieticians, social workers and other members of the allied health team to assist in the care of the newborn infants at a local level. This will lead to the development of a provincial CF clinical network for the delivery of care to the infant under the leadership of BC CF Clinics. The construction and development of a regional team will foster inter-professional relationships for ongoing communication and collaboration

It is envisaged that, through the provincial CF clinical network, care will be provided for newborn infants with cystic fibrosis infant that not only meets but exceeds provincial and national standards

Discipline Specific Newborn Screening Resources

I. Nutrition

Nutritional Recommendations for the Infant Diagnosed with Cystic Fibrosis

Written by: Barbara Bell, RD

Updated by: Christine Loong, RD

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

All children and adults 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)

Most 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 from opened gel caps, divided to 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

Children 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 children with CF.

Neonates: Breastfed newborns require the standard Vitamin D supplementation of 10 µg/ (400 IU) daily.

At diagnosis: Most infants with CF will be started on water soluble Vitamin E supplements of 10 IU/kg daily.

When stable: Once albumin levels are assessed to be normal(>35g/l), infants will be prescribed a multivitamin supplement including higher levels of vitamins A, D, E & K.

Recommended: AquADEK (multivitamins with selenium and zinc)

Practice Point

- Parents should be advised of careful handling with AquADEK 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 $>30\text{mmol/L}$.

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 milk is the best source of nutrition for all babies. A CF infant can be successfully breast fed and benefit from the advantages of breast milk. The mother-infant bonding is valuable as mom adjusts to the 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.
- Domperidone may be considered for a breast feeding mother if milk supply is a concern.
- 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 is a first recommendation, and thereafter meats. Vegetables, fruits, other foods can be introduced following the usual infant feeding guidelines.

When regular solid feeds are established salt may be added by sprinkling onto meals and the infant's salt solution 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:

Borowitz D, Baker RD, Stallings V. Consensus Report on Nutrition for Pediatric Patients With Cystic Fibrosis. *J Pediatr Gastroenterol Nutr* 2002; 35:246-59.

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.

Satter, E. The feeding relationship. *J am Diet Assoc* 1986;86:352-6.

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

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

The usual enzyme dosage range is 500 - 2500 units of lipase/kg/meal, up to a maximum of 10000 units of lipase/kg/day. In an infant, the total daily dose would be divided among the number of times the infant feeds. Infant dosing is initiated at 1000 units of lipase/kg/feed to be given immediately prior to each feed up to a maximum of 8 feeds per day. 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.

Contents of Coated Pancreatic Enzymes on Pharmacare Plan D Formulary			
Brand Name	Lipase (units)	Amylase (units)	Protease (units)
Pancrease MT 10	10000	30000	30000
Pancrease MT 16	16000	48000	48000
Cotazym ECS 4	4000	11000	11000
Cotazym ECS 8	8000	30000	30000
Cotazym ECS 20	20000	55000	55000
Creon 5 Minimicrospheres	5000	16600	18750
Creon 10 Minimicrospheres	10000	33200	37500
Creon 25 Minimicrospheres	25000	74000	62500

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 > 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, patients with cystic fibrosis also require vitamin supplementation, particularly the fat soluble vitamins – vitamins A, D, E, and K.

Infants should be assessed prior to starting vitamin supplementation, as vitamins can be potentially toxic in the CF infant.

When fat-soluble vitamins are initiated in an infant, usually not all 4 vitamins are needed right away.

AGE	Vitamin A (IU)	Vitamin E (IU)	Vitamin D (IU)	Vitamin K (mg)
0 – 12 months	1500	40 – 50*	400	0.3-0.5
1 – 3 years	5000	80 - 150	400 - 800	0.3-0.5
4 – 8 years	5000 – 10000	100 - 200	400 - 800	0.3-0.5
>8 years	10000	200 - 400	400 - 800	0.3-0.5

*In infants up to 1 years of age, to ensure adequate absorption, the water soluble Vitamin E preparation should be prescribed Aquasol E®. In infants, Vitamin E is dosed at 10 units/kg up to a maximum of 50 units in the first year of life.

Adapted from the Consensus Report on Nutrition for Pediatric Patients with Cystic Fibrosis. J Pediatr Gastroenterol Nutr, Vol. 35, No. 3, September 2009

Once all laboratory values indicate that it is safe (Albumin > 35g/l) for the infant to start the fat-soluble vitamins A, D, E and K, a single product may be prescribed to the patient. There are currently two vitamin products formulated containing the fat soluble vitamins. Both products are available through the Health Canada Special Access program:

- AquADEKs® Liquid, Chewable tablets, Gel capsules
- SourceCF® Liquid, Chewable tablets, Gel capsules

For infants, the pediatric liquid formulation would be prescribed:

Age	AquADEKs® Pediatric Liquid	SourceCF® Pediatric Drops
0-12 months	1 mL daily	1 mL daily
1-3 years	2 mL daily	2 mL daily

Both AquADEKs® and SourceCF® products are on part of Pharmacare Plan D: 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)

Antibiotic Prophylaxis

Antibiotics are used to prevent the growth of a common organism called Staphylococcus Aureus in the first year of life.

Staphylococcal prophylaxis is based on the evidence of limited studies, but it is our recommendation to use it in all patients in the first year of life.

Cloxacillin is recommended in the first month of life or until the liver enzymes, bilirubin and albumin normalize in the infant. Cloxacillin is not acid stable, and is therefore poorly absorbed in the presence of food. Adherence to Cloxacillin is

difficult in an infant, especially if feeding every 2 -3 hours, and Cloxacillin is to be administered on an empty stomach. When the infant is over 1 month of age and liver enzymes normalized, then the baby is switched to Septra®. Unless there is an intolerance or allergic reaction to Septra®, staphylococcal prophylaxis is continued for the first year of life. Cephalexin is not recommended for staphylococcal prophylaxis, as one randomized control trial found an association with Pseudomonas colonization in the group being treated continuously with cephalexin.

- Cloxacillin 50 mg/kg per day divided BID on an empty stomach
- Septra 8 mg/kg of trimethoprim/40 mg/kg of sulfamethoxazole per day divided BID (equivalent to 1 mL/kg/day) used in patients > 1 month old
 - NOTE: avoid using in neonates as the sulfamethoxazole displaces bilirubin from the albumin binding sites increasing the risk for kernicterus

References:

AquADEKs® Product Information:

<http://www.yasooproducts.com/aquadeks/supplementfacts.html>

Borowitz D, Baker RD, Stallings V. Consensus Report on Nutrition for Pediatric Patients With Cystic Fibrosis. J Pediatr Gastroenterol Nutr 2002; 35:246-59.

Pharmacare Cystic Fibrosis Plan D:

<http://www.health.gov.bc.ca/pharmacare/plans/index.html>

SourceCF Product Information: <http://www.sourcecf.com/nutritionals.htm>

Smyth AR and Walter S. Prophylactic anti-staphylococcal antibiotics for cystic fibrosis. Cochrane Cystic Fibrosis and Genetic Disorders Group Cochrane Database of Systematic Reviews. 11, 2010.

Stutman HR et al. Antibiotic prophylaxis in infants and young children with cystic fibrosis: A randomized controlled trial. J Pediatr. 2002; 140:299-305.

Weaver LT et al. Prognosis in cystic fibrosis treated with continuous flucloxacillin from the neonatal period. Arch Dis Child. 1994;70:84-9.

III. Physiotherapy

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

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

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 both alter ventilation and allows gravity to assist in the movement of mucus from the small airways to the large airways.
- Percussion and Vibration; the caregiver provides an external mechanical force through percussion and vibration over specific lobes of the 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 large Therapy ball.

Assisted huffing.

- The caregiver applies an external force over the rib cage only during expiration to increase the expiratory flow rate. This may also 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 including 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:

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.

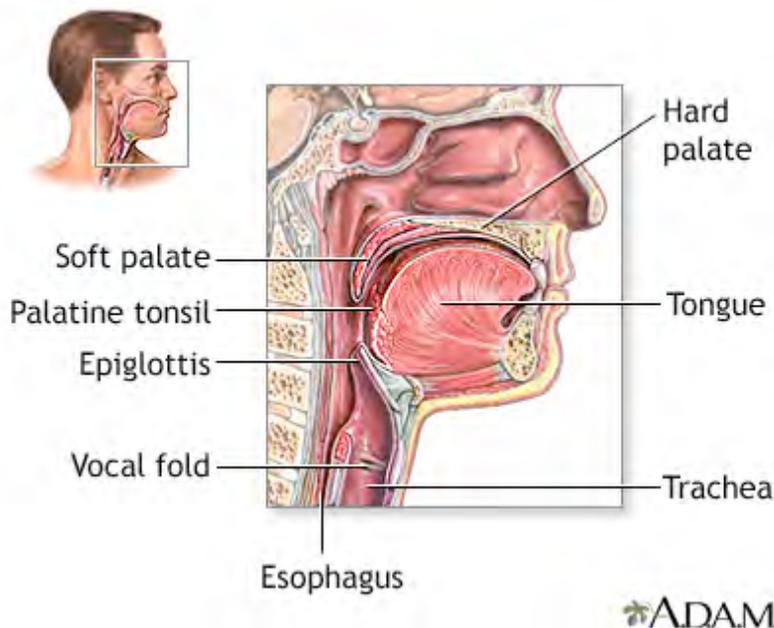
*The following document describes the difference between a cough swab and throat swab. The cough swab is the ideal laboratory test, in patients who do not produce sputum, for cystic fibrosis patients who require frequent microbial surveillance of the lungs not the throat.

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 culture¹.

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 saline¹. 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.



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

1. Use of cough swabs in a cystic fibrosis clinic

A C Equi^a, S E Pike^b, J Davies^a, A Bush^a ^aDepartment of Paediatric Respiratory Medicine, Royal Brompton & Harefield NHS Trust, Sydney Street, London SW3 6NP, UK, ^bDepartment 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.

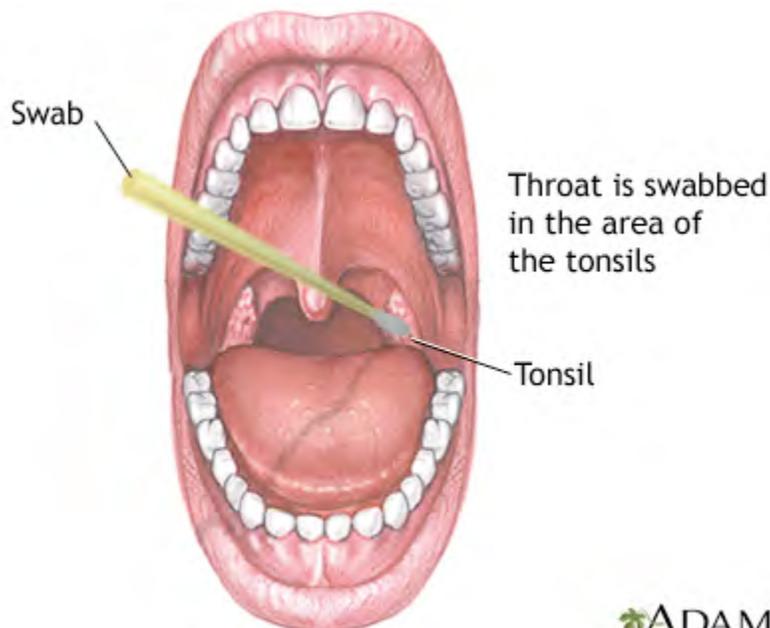
Definition of a Throat swab culture

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.



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

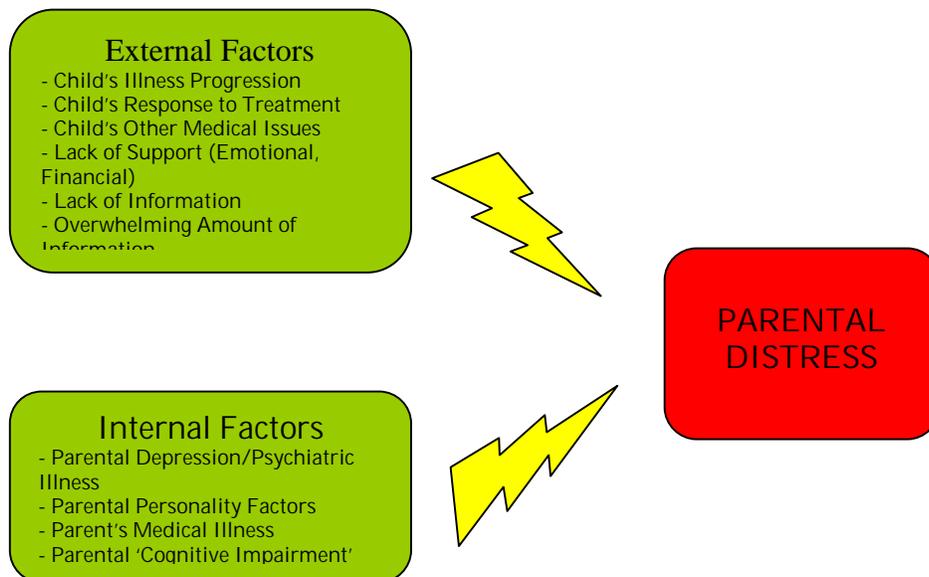
In major depression, these symptoms are persistent over weeks to months, and are a significant change for them. If these symptoms are seen in the first few weeks after diagnosis, they should be put in the context of the enormous impact this diagnosis will have on any parent of a newborn. However, if they persist they may indicate a Depressive Episode.

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 are shown below:



We also suggest that vigilance for and informal assessment of psychiatric signs and symptoms in parents of newborns diagnosed with CF are 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/madrs_print1.htm)

is a clinically useful scale which includes the more specific indications of Depression.

3. Psychiatric Consultation

We recommend if possible having a paediatric psychiatrist available to consult with the CF team, not only at the time of initial assessment but on an on-going basis. Ideally this consultant can do 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 provide or help arrange treatment for parents with these difficulties.

V. Social Work

NEWBORN SCREENING PROGRAM
CYSTIC FIBROSIS CLINIC

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

Parental Counseling Preferences:

A study from Wisconsin University Hospital (Tluczek) described the need to assess the parents' preferences for counseling and noted two dimensions: factual information and emotional support. *This study found that if "Counseling matched parents' preferences it reduced distress while mismatched counseling tended to increase parents' worry about their infant."* It was also noted that parental preferences changed several times during the first year after diagnosis. There were times when the need for factual information exceeded the need for emotional support and vice versa. It is important for the health care professional to observe the parents' cues and to focus on what appears to be important to them at that time.



Social Work Financial Assessment: Newborn Screening Program

Date: _____

Interviewed: _____

Name: _____ Unit Number:

_____ Birth Date: _____

Address: _____

- Parental Status: Couple
 Single Parent
 Blended Family
 Joint Custody
 Single Custody

Guardianship Information _____

MCFD

Consent on File

Contact Information:

Maternal Name:

Home Tel: _____ Work Tel: _____ Cell:

Paternal Name:

Home Tel: _____ Work Tel: _____ Cell:

Sibling Name(s) & Ages & Residence:

Family Doctor: _____ Pediatrician:

Current
Concerns: _____

Action Plan

Social Worker: _____
Local: _____

Parental Employment Status

Maternal: Employed- F/T or P/T
 Self-Employed
 At Home
 Unemployment Insurance
 Income Assistance
 Student
 Other _____

Paternal: Employed- F/T or P/T
 Self-Employed
 At Home
 Unemployment Insurance
 Income Assistance
 Student
 Other _____

Benefit Status (Registered and/or Eligibility Reviewed)

Extended Health Benefits: Maternal – Yes/ No Sunshine Bundle
 Discussed Paternal – Yes/ No Sunshine Bundle

Fair PharmaCare: Aware: Yes/ No In Process Registered
 Discussed

Disability Tax Credit: Aware: Yes/ No In Process Registered
 Discussed Disability Plan Disability Grant

First Nations Benefits: Patient: Yes/ No Status

Maternal: Yes/ No Status # _____

Paternal: Yes / No Status # _____

First Nations Contact Information:

Financial Distress Minimal Moderate Severe

Major Financial Issues Facing Family at Present

Financial Resources

Income Assistance: N/A
 Income Assistance
 In Process
 Registered
 Transportation/Accommodation
 MSP Premium Assistance
 Medical Services
 Healthy Kids Program

MCFD Special Health Needs: N/A In Process Registered To Apply

Accommodation: Rental Assistance Program
 BC Housing
 First Nations

- Transportation: Hope Air
 Wings of Hope
 T.A.P.
 First Nations
 Shriner's Bus
 Northern Health Bus
 Interior Health Bus

Income Tax: Transportation Medications Accommodation

At Home Program: N/A In Process Yes

Contact: _____

Respite Services: N/A Yes

Organization: _____

Community Living Program: N/A In Process To Apply Yes

Contact: _____

Support Organizations

- Variety Club
- Red Cross
- Kinsmen
- Rotary Club
- Lions Club
- C F Foundation
- Membership
- Medication Grant
- Transportation Grant
- Accommodation Grant

Wish Organizations

- Make a Wish
- Children's Wish
- Starlight Foundation
- Sunshine Dreams



Social Work Social Support Assessment: Newborn Screening Program

Family Genogram

Social Support

Who provides you with support when you most need it?

Maternal _____

Paternal _____

Current Perceptions

Maternal

1. What is the toughest part of this situation right now?

2. When you have gone through a difficult time before what helped you through it?

Current Stress Scale Low 1 2 3 4 5 High
Stressors: _____

3. What could the CF team offer you that would best meet your needs right now?

Paternal

1. What is the toughest part of this situation right now?

2. When you have gone through a difficult time before what helped you through it?

Current Stress Scale Low 1 2 3 4 5 High
Stressors: _____

3. What could the CF team offer you that would best meet your needs right now?



Social Work : Questions to Consider
Linda MacNutt MSW RSW
604) 875-2345 ex 7013

- Who are the members of the CF team?

- In summary, what do we understand about the diagnosis of cystic fibrosis?

- Who do we want to tell about our child's diagnosis?
For example: family, friends, day care, school

- What information do we want to share right now?

Mother: _____

Father: _____

Others: _____

Social Work References:

Newborn Screening for Cystic Fibrosis: Parent's Preferences Regarding Counseling At the Time of Infants' Sweat Test; Tluczek,A et al, (Wisconsin-Madison) Journal of Genetic Counseling, Vol. 15, No. 4., August 2006

Appendix: Information and Resources for Families

Online Resources

1. BC Family Residence Program: <http://www.bcfamilyresidence.gov.bc.ca/>
2. Cystic Fibrosis Canada: www.cysticfibrosis.ca
3. BC Children's Hospital Cystic Fibrosis Clinic: www.bcchildrens.ca. Click on 'services', then 'specialized pediatrics' then 'cystic fibrosis'

Handouts

1. Cystic Fibrosis and the Care of My Child- What is it All About?
2. Nutrition for Infants with Cystic Fibrosis: An Educational Guide for Families
3. How to Give Enzymes to Babies
4. Salt Supplementation
5. Introduction of Solids: A Guide for Infants with Cystic Fibrosis
6. Vitamins in CF
7. The CF Newborn Screening Program Support Resources: A Parent's Guide to BC Children's Hospital

Cystic Fibrosis and the Care of My Child- What is it All About?



An Education Resource for Families

You will get a learning binder at one of your first visits to the CF Clinic. You should bring this binder with you to all clinic visits and education sessions for the first few months after diagnosis. There is some paper inside so you can write down questions or comments throughout your experience.

You will be given a number of pamphlets, handouts, and booklets to help with your learning. The following is a list of resources you will be receiving during your CF education.

1. Understanding CF – pamphlet		11. Managing Nutrition & Malabsorption in CF- Booklet	
2. Infection Control - pamphlet		12. Vitamins and CF – Handout	
3. CF in Canada – pamphlet		13. Healthy Diet in CF – Handout	
4. Information on CF Clinic at BCCH- Handout		14. A Parent Guide to Giving Enzymes to Infants – Handout	
5. The CF Team- Handout		15. Managing Lung Problems in CF- Booklet	
6. Who to call and Where to Go to Care for your Infant with CF- Handout		16. Cystic Fibrosis and Physiotherapy - Booklet	
7. Carrier Testing for CF- Pamphlet		17. Disability Tax Credit- Handout	
8. Beginning CF Care: A Guide for Parents of Young Children - Booklet		18. Social Work Package	
9. Your Child and Cystic Fibrosis - Handout		19. BC Assoc. Government Grant Policy for Financial Assistance with CF Meds	
10. Nutrition for Infants with CF - Handout		20. BC Assoc. Government Grant Policy for Financial Assistance with CF Travel and Accommodation- Handout	

There are many topics to cover at time of diagnosis. Listed below are the items we would like to review during the first month after diagnosis. It is alright if you do not understand it all right away. You will hear this information many times over the coming year and you can always ask for more time with the team.

Remember, as caregivers you are very important members of the team. We want to hear all your questions and concerns.

Management of Cystic Fibrosis:

1. What sort of tests will be done on my child?
2. How does CF affect the body? What does that mean for my child both now and in the future?
3. Infection Control – what is it and why is it important?
4. What types of medications has/will my baby be started on?
5. Should my child receive all the regularly scheduled vaccinations?
6. Should my child receive the flu shot? How about me and my family?
7. What is RSV? What is Synagis/Palivizumab? Does my child need that too?
8. What does the future of CF look like? What sort of current research is being done?
9. When do I call the CF Clinic and when do I make an appointment with my family doctor or pediatrician?
10. When should we come straight to emergency?
11. What is the Patient Data Registry? How and by whom is the information used?
12. Will the CF Team share information with my family doctor or pediatrician?

Cystic Fibrosis & My Family:

1. What are the genetic implications of my child having CF?
2. Who are the people on the CF Team? Who should I call when I have a question or a problem?
3. What is the BC Government Grant? How can I apply?
4. How does the clinic run? How often do we need to come?
5. Where can I park when I come to clinic or the hospital?
6. I got some mail from you after our clinic visit. What is that for?
7. How might our child's CF affect our other children?

Supports Systems:

1. Who is the Cystic Fibrosis Foundation? What sort of information and support can they provide?
2. Can I go to my regular pharmacy to get my child's medications? Do I get any special drug coverage?
3. What are Fair PharmaCare and Plan D? How do I register?
4. Is there someone available to help me if I am having trouble coping with all the changes and feelings around my child's diagnosis?

There are some resources available online as well. If you choose to go on the internet we recommend you choose well established credible sites to gather your information. We suggest you try these ones:

- GeneTests (hosted by the National Center for Biotechnology Information (NCBI))
www.genetests.org
- Canadian Cystic Fibrosis Foundation
www.cysticfibrosis.ca
- Canadian Cystic Fibrosis Foundation (CCFF), Vancouver & Lower Mainland Chapter
www.cfvancouver.ca/
- For a listing of contacts for other CCFF chapters in BC:
cfvancouver.ca/index.php?option=com_content&task=view&id=108&Itemid=57

Nutrition for Infants with Cystic Fibrosis: An Educational Guide for Families

Written by Barbara Bell, RD

Nutrition is important for all children and especially for a baby with Cystic Fibrosis. Your baby needs the same feedings as others do – breast milk (or formula) for the first six months then solid foods added after 6 months of age.

Children with CF may have problems digesting and absorbing foods and also require higher amounts of energy (calories). They need extra vitamins and salt.

These special nutrition treatments for CF will help your baby to grow and gain weight well.

Pancreatic Enzyme Supplements.

Once CF is diagnosed other tests determine how well your baby can digest food. Most children with CF require enzyme supplements to be taken with every feeding.

Enzymes are essential for the body to digest foods and use the nutrients for health and growth. Your baby can be well nourished when enzymes are given before every feed.

The dose taken at each feeding will be adjusted as your baby takes more milk and food. Your CF team will provide advice on how to give enzymes by spoon with a small amount of applesauce. It is important to give the correct dosage just before starting to feed.

Vitamins

Multivitamin supplements are recommended for all children and adults with CF and will be prescribed for your child.

In the first few weeks after diagnosis the prescription may be for Vitamins D and E only. It is the standard for all breastfed infants to be supplemented with Vitamin D. Most infants with CF need extra Vitamin E from the time of diagnosis.

Once settled on enzymes the prescription for multivitamins will start. Special multivitamins are used to meet the extra requirements for CF notably for the fat soluble vitamins A, D, E and K which can be lost by malabsorption. Your baby's vitamins will be in liquid form and you will be given directions how to give the drops once or twice daily.

Salt

Infants and children with CF lose more salt than others through their sweat everyday. Loss of salt occurs even faster with fever, in hot weather or when very active (playing sports).

Older children add salt to their food everyday. However, for infants salt must be given daily using a special solution.

A recipe for salt solution will be provided to you. The solution may be mixed with expressed milk or formula. Your CF team will advise you how to give the salt.

When your baby takes solid foods (after age 6 months) salt may be sprinkled onto meals.

First Six Months

Breast milk is the best food for all babies. Exclusive breast feeding your child with CF will give your baby all the nutrition needed until six months of age. If not breast fed, most babies with CF can grow well on regular infant formula.

Your baby may need more energy (calories) than others and so need more milk and drink more often. (If you are a breast feeding mother you need to eat well yourself to keep your energy and strength up!)

Solid Foods

Solid foods should be introduced to your baby's diet at six months of age. All babies need a source of iron so give infant cereals which are fortified with iron as a first food then later introduce meats. Pureed vegetables and fruits add variety. Encourage your baby when giving new foods and remember it often takes several times of trying before a child accepts and likes a new taste.

Your baby needs the same good foods as other infants and will learn to eat by the usual steps of starting with purees fed from a spoon then learn to chew more chunky foods, use fingers to pick up foods and eventually use a spoon to self feed. As with all children learning to eat, expect your child will be messy!

Most baby foods contain no added salt so you will be advised to add salt to meals.

Small amounts of butter, margarine or oil may be added to meals for extra energy, but not too much or it might reduce appetite for other foods. Other high calorie additions such as cream or milkshakes are not needed for infants. Those extras may be helpful for an older child who is active in sports. Your CF dietitian can guide you.

Your young child needs basic healthy, family meals and so should not need to be specially catered to or "spoiled".

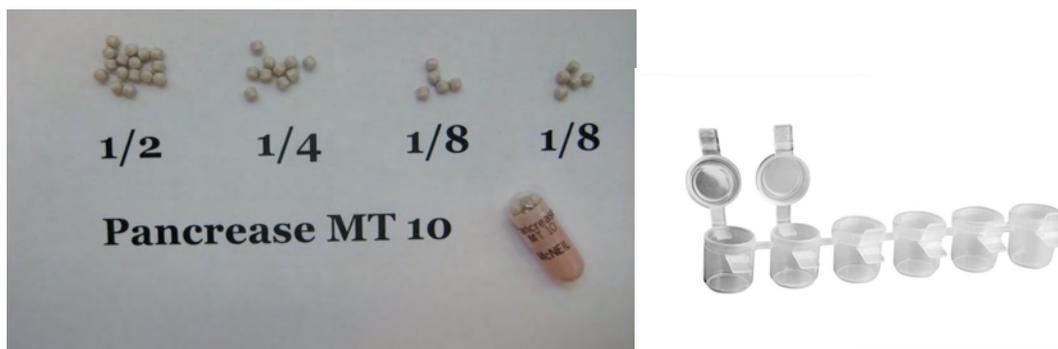
The extra needs of CF can be met by the addition of salt and regular use of important foods including meats, cheeses, yogurt and, later, eggs and whole milk.

Nutrition is important for health and growth and will always be part of your child's CF care. Enjoyment and relaxation at mealtimes is a great foundation for health eating.

How to Give Enzymes to Babies

Everyone needs substances called *pancreatic enzymes* to help their bodies digest and absorb the food they eat. About 85% of people with Cystic Fibrosis do not make enough of these enzymes. For this reason, your baby has been prescribed pancreatic enzymes. This handout gives you the information you need in order to successfully give your baby enzymes before each feed.

1. Use a small baby spoon with a flat, rubbery soft surface.
2. Open the capsules onto a clean, flat surface. Eyeball enzymes to get the right dosage. You do not need to count the beads. It may be helpful to measure the doses in the morning and put each dose in a separate compartment of a container with a lid. Paint pots from the craft store or a multi-dose medication container can work well for this. There is less waste and you always have a supply of enzymes to take with you.



3. Mix enzymes with a small amount of apple sauce immediately before you give them. The enzymes will begin to break down if left sitting in the apple sauce for too long. How much applesauce? It may take a little practice to get this right. Usually 1 teaspoon or less is enough to help swallow the beads. Too much applesauce is more work for your baby and "fills up" the tummy.

Only mix enzymes in applesauce because:

- It protects your baby's mouth and throat from damage caused by early breakdown of the enzymes
- It helps the enzymes to get to the intestine before dissolving so they can do their work on the food.
- It is easy for your baby to swallow.

- Most babies like the taste and most babies will not have an allergy to apples.
- It's not too sweet.

It is normal for babies to not like new foods. Keep offering the applesauce to your baby many times, such as 12 to 15, before deciding your child doesn't like it.

4. Do not give enzymes mixed with other medications.
5. Beads should never be mixed with breastmilk or given with dairy products. You can breastfeed your baby after your baby has swallowed the enzymes. Once your baby is old enough to have dairy products you can give them with meals after the enzymes have been swallowed.
6. Put the applesauce and enzyme mix on the tip of the baby spoon and aim for the middle of the baby's mouth. Too far back will bring out the gag reflex and too far forward will cause the tongue to push the enzymes out. Wet your baby's mouth first with a small syringe of water if she or he is having trouble swallowing the medication.
7. Before starting to feed your baby check that all the beads have been swallowed by carefully looking into your baby's mouth. Use your finger to find any beads that have not been swallowed and give them again. If baby is having trouble swallowing them, you can use a syringe to give a small amount of water in the corner of the mouth or offer a soother to wash the rest down.
8. If you are breastfeeding, enzymes can irritate your nipples. You may decide to put Vaseline on your nipples before feeding time.
9. Give only the amount of enzymes your doctor has prescribed. Enzymes are considered 'active' for two hours after swallowed. There is no need to give enzymes twice in a two hour period. *For example, you give enzymes only once in a two hour period even though your baby had breastfed three times during the two hours.*
10. Keep track of enzyme doses on a calendar. Babies are often up during the day and during the night. This can make it difficult to track how much enzyme they have had in each 24 hour period. Keep a chart of enzymes doses, feedings and how many times your baby pees or poos. This can be helpful for clinic visits.

Other things to watch for....

Some babies react to the enzymes during the first month. Mouth sores, skin irritation and diaper rash may develop. Your baby's bottom may become red or raw

What helps?

- Use a barrier cream to protect your baby's bottom. A zinc-based diaper cream works well. You may also try leaving the diaper off for periods of time during the day to air out the area and help it to heal.
- Let your CF clinic know if any of these symptoms become concerning.

Enzyme Q &A

Q. How do I safely store enzymes?

- Check the expiry date printed on the bottle. Enzymes lose their effectiveness if they are past their expiry date.
- Keep the bottle tightly closed, and store in a cool, dry place. Do not refrigerate.

Q. What else is important to know about enzymes?

- Know the name and strength of the enzymes prescribed to your child, as well as the number she or he should take before meals and snacks.
- The number of bowel movements your child has every day is one sign of how well she or he is digesting food. Call the CF Clinic about any significant change in the number and type of bowel movements.
- As your child grows larger he or she will need to take more enzymes. When the amount or type of enzymes is changed, your child may need 3 to 5 days to adjust to that change.

Q. If my child has a snack 1 hour after eating his or her meal, should I give more enzymes?

A. No. The enzymes that your child took before his or her meal work for about two hours. Never give enzymes twice in a two hour period. Never give more than the maximum daily amount prescribed by your CF Clinic.

Q. Are there foods or drinks that do not need enzymes?

A. Yes. Your child does not need to take enzymes when he or she drinks fruit juice or eats plain fruit or vegetables. However, meals and snacks that have protein, carbohydrates and fats in them do need enzymes to be digested. Your child needs these types of foods to get the calories they need.

Q. Should my child take fewer enzymes if he or she is going to eat a small meal or more enzymes if she or he is going to eat a very fatty meal?

A. No. Do not change the number of enzymes your child takes from day to day.

Q. What should I do if we are going to be away from home for a meal?

A. Carry enzymes with you at all times so you have them in case your child needs to eat. Be sure to check expiry dates if you keep enzymes away from home.

Call the CF clinic if your baby has:

- Tummy pain or increased gas
- A more distended or firm tummy
- Bowel movements that different than usual for your baby. They may be looser or harder, smell bad or be oily. This may be a sign that your CF doctor needs to make an adjustment to the enzyme dose.

CF Clinic Nurse's Line: 604-875-2345 ext 2623 or 7005



Nutrition Services

SALT SUPPLEMENTATION GUIDELINES FOR CYSTIC FIBROSIS

Your child requires extra salt (NaCl) while being fed mostly breast milk, formula or infant foods. The simplest way of providing this salt is as a salt solution which can be given by dropper or can be mixed with breast milk, formula or food.

As your child gets older, and eats more table foods and drinks whole milk the liquid salt supplement can be stopped, usually by the age of 1 year. You will then need to give salty foods or add extra table salt to foods every day.

GUIDELINES

_____ needs _____ ml of the salt solution daily.

Give this in _____ doses of _____ mL of salt solution daily

Recipe for salt solution

1. Mix 1 level teaspoon salt (5 mL) with 40 mL of boiled cooled water.
2. Store in covered container in the refrigerator and use as needed.

Note: This recipe for salt solution provides 2.5 mEq. Sodium /1.0 mL.
The usual goal for salt supplementation: 2 - 4 mEq /kg/day

Your Dietitian is _____

Call 604-875-2345 Local _____ if you have any questions



Introduction of Solids: A Guide for Infants with Cystic Fibrosis

Between the ages of 4-6 months, your child may be ready for solids. Signs that your baby is ready are:

- Can sit and support his or her head up
- Shows interest in food by watching or opening mouth
- Tongue no longer pushes out food

If your baby is showing these signs, and with the recommendation from your physician or dietitian, your child is ready to experience a new variety of tastes and textures!



Tips to Starting Solids:

1. Choose a time of the day when the baby is alert, interested and most likely to try something new
2. Offer solids **AFTER** breast or formula feeding
3. Start with 1-2 teaspoons of iron fortified infant cereal (such as rice, barely or oat) mixed with breastmilk or formula into a thin puree consistency. Gradually increase amount to 4 - 8 Tbsp (60 - 125 mL) daily.
4. Offer 1 new food at a time
5. **Wait 3-5 days** before introducing another new food item. This makes it easy to tell if your baby is allergic to any food.
6. Next, introduce meat (such as chicken, beef, veal, salmon, lentils tofu, egg yolk). Gradually increase to 6 Tbsp (100 mL) daily.
7. You can add a small amount of fat such as oil, margarine or butter to solids to allow your child to get used to the taste, while providing extra energy without reducing appetite or intake. It is suggested to add 1 teaspoon (5mL) of fat to every $\frac{1}{2}$ cup (125 mL) of solids
8. Once your baby is eating iron-rich foods, you can start to introduce pureed fruits and vegetables and gradually increase to 4 - 8 Tbsp (60 - 125 mL) daily
9. Gradually increase the amount of solids per feed and increase the frequency of solid feeds to 3-4 times per day. You can combine foods such as pureed meat with carrots and cereal once your child has tolerated these foods.

*Do not put cereal or solids in a bottle

**Always stay with your baby while he or she is eating or drinking

Avoid the following foods for infants:

- Foods that can cause choking such as: popcorn, peanuts, nuts, hard candies, hard raw vegetables, large seeds, jelly beans, chips, ice cubes, globs of peanut butter
- Honey - may cause botulism poisoning. Do not give for infants under 1 years of age
- Egg White - may cause an allergic reaction. Avoid for infants under 1 year of age.
- Cow's milk - breastmilk or formula does not need to be substituted for cow's milk until 1 year of age.
- Unpasteurized dairy products

How much to offer

Your baby knows the best of how much food his or her body needs. Their appetite will vary with each meal and from day-to-day. Signs of hunger are opening mouth for spoon or getting upset when food is taken away. Babies will shut their mouth, turn their head away or push food away when they have had enough and are full. Listen to your child's hunger and fullness cues and avoid forcing your baby to eat when they are full.

Salt

- Continue salt solution as recommended by your physician or dietitian. Eventually your child will be weaned off or discontinued the solution as solid intake increases
- Add sprinkle of salt to each spoonful of solids
- If you're preparing homemade baby food, can puree foods with salty broth



Bowel Movements

Your infant's stools will start to change with introducing solids. Typically stools will become more firm and brown in colour. Frequency of bowel movements will decrease as your child gets older. Contact the CF clinic if your child is showing signs of malabsorption (such as oily, foul smelling, more frequent stools, and increase in appetite) or other change in stools such as constipation or diarrhea.

Fluids

Continue to breastfeed or offer formula with every feed. Sips of water may be offered in a cup after meals or in between meals and snacks. Limit juice to $\frac{1}{4}$ - $\frac{1}{2}$ cup (60 - 125 mL) daily, diluted and served in a cup.

Textures

Initially the texture of your child's foods will be thin puree. As solid intake progresses, the texture of the food will be thicker and lumpier. Eventually the texture of foods can be soft, mashed, diced and can introduce crunch foods that dissolve (such as mum-mum's).

From 9-12 months of Age

1. Offer solids first **BEFORE** breast or formula feeding
2. Your baby can continue to eat the same meals at same mealtimes. Texture of the food can be soft, mashed or diced and can offer finger foods. Signs you can tell if your child is ready for finger foods are the ability to mash and chew with their gums and being able to grab and hold objects with thumb and fore finger.
3. Can try offering cottage cheese, plain high fat yogurt and small pieces of hard cheeses (cheddar, gouda, marble, mozzarella)
4. Let your baby try to self-feed with their fingers or with a spoon. Let them have fun playing and exploring with their food, even if it's messy!

Some finger food ideas to give to your child are:

- Soft and small pieces of fruit such as peaches, ripe banana, mango, melon, avocado, squished blueberries, soft pears
- Scrambled egg yolks
- Small pieces of soft, cooked vegetables such as carrots, peas, broccoli, green beans, squash
- Tofu cubes
 - **try coating fruit, vegetable or tofu in cheerio/cereal dust or wheatgerm to make it easier to pick up
- Sweet potato fries
- Soft cooked pasta such as cut up spaghetti, rotini, penne
- Small pieces of cheeses such as mozzarella, cheddar, marble
- Pieces of shredded meats such as cooked chicken, fish, beef or turkey
- Pieces of toast, roti or tortilla spread with cream cheese or butter/margarine
- Oat rings cereal

Vitamins and Mineral Supplements

Continue with AquADEK solution 1 mL daily and other supplements that have been recommended by the CF team or your healthcare provider.

Sample Meal Plan

Below is a suggested sample meal plan to be used as a guideline only as every infant's meal pattern and amount of solids will vary. As your child's intake of solids increases, intake of breastmilk or formula will decrease.

Mealtime	6-8 months	9-12 months
Early Morning	Breastmilk or Formula	Breastmilk or Formula
Breakfast	<ul style="list-style-type: none"> • Breastmilk or Formula <u>before</u> solids • 1 tsp - 2 Tbsp iron fortified infant cereal • 1 tsp - 2 Tbsp cooked egg yolk • 1 tsp - 2 Tbsp mashed fruit • 1 tsp - 2 Tbsp tofu 	<ul style="list-style-type: none"> • 2-3 Tbsp iron fortified infant cereal • 2 Tbsp plain high fat yogurt • 2 Tbsp cooked egg yolk • 2-4 Tbsp soft, cubed or diced vegetable or fruit (squash, pears) • Breastmilk or Formula <u>after</u> solids
Morning Snack	Breastmilk or Formula	<ul style="list-style-type: none"> • Strips of whole wheat toast with butter • Grated apple • Breastmilk or Formula <u>after</u> solids
Lunch	<ul style="list-style-type: none"> • Breastmilk or Formula <u>before</u> solids • 1 tsp - 2 Tbsp iron fortified infant cereal • 1 tsp - 2 Tbsp mashed chicken • 1 tsp - 2 Tbsp mashed vegetable (yam, peas) 	<ul style="list-style-type: none"> • 2 -3 Tbsp iron fortified infant cereal • 2 - 4 Tbsp cut up meat • 2 -6 Tbsp soft cubed or diced vegetable and fruit (avocado, carrot, peaches) • Breastmilk or Formula <u>after</u> solids
Afternoon Snack	Breastmilk or Formula	<ul style="list-style-type: none"> • Small cheese cubes and salted crackers • Breastmilk or formula <u>after</u> solids
Dinner	<ul style="list-style-type: none"> • Breastmilk or Formula <u>before</u> solids • 1 tsp - 2 Tbsp iron fortified infant cereal • 1 tsp - 2 Tbsp mashed beef • 1 tsp - 2 Tbsp mashed fruit (peaches, pears) 	<ul style="list-style-type: none"> • 2 - 3 Tbsp iron fortified infant cereal • 2 - 4 Tbsp diced cooked chicken • 2 - 6 Tbsp soft, cubed or diced vegetable (green bean, cantaloupe, banana) • Breastmilk or Formula <u>after</u> solids
Bedtime Snack	Breast milk or Formula	Breast milk or Formula

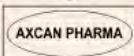
Reference: Healthlink BC – What to Feed Your Baby: Sample Meals for Baby in the First Year

CF Health Matters



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Vitamins and Cystic Fibrosis

Why do people with Cystic Fibrosis need to take vitamins?

Many people with CF have problems when they eat food, making it hard for their intestines to soak up nutrients, *especially* fat. To help with this, enzymes are taken with meals and snacks. Vitamins A, D, E, & K are unique because they need fat in order to be 'soaked up' by your body, they are called "fat-soluble" vitamins. Even when someone takes enzymes they can still lose fat in their stool. If they are losing fat then they are probably also losing vitamins. If you don't take your vitamins, over time, you may develop low levels in your blood. This is known as a *vitamin deficiency*.

You may wonder why it is important for someone with CF to take their vitamins if they don't have a problem when they eat. To understand why, you must realize that vitamins are important to your body when you are healthy, but they even more important when you are sick. People who have CF may find they get sick more often than people who don't have CF. To help their bodies fight an infection they need more vitamins than someone without CF. We ask people with CF to take a vitamin pill because it is very hard to get enough vitamins from food alone. Our focus is often the *fat soluble* vitamins, but it is important to remember that all vitamins are needed for a healthy body.

What do the different fat soluble vitamins do and what will happen if I do not get enough?

Vitamin A

Vitamin A has many important roles in the body. It helps you see at night and lets your eyes adjust when you move from a dark place to a bright place. If you have a vitamin A deficiency then your eyes may not be able to get used to changes in light. There have been people with CF who have reported having this problem. If you think this is a problem speak to your CF doctor. Vitamin A also helps make skin healthy and helps fight infection when you get sick. During childhood it helps you grow. It is found in eggs, liver, tomatoes, milk, and some fruits & vegetables. For people with CF, food sources are not enough to prevent vitamin A deficiency, therefore a vitamin supplement is usually recommended.

Vitamin D

Vitamin D comes from two sources. It is made by the skin when it is exposed to sunlight, and it is found in the foods we eat. It helps your body absorb calcium, and helps move calcium from your blood to your bones. Without enough vitamin D your bones can weaken and the chances of breaking a bone become greater. While foods such as eggs and cereals contain vitamin D, the main source is milk. A vitamin supplement is usually recommended to make sure you get enough every day.

Vitamins and Cystic Fibrosis

If you want more information on Vitamins and CF, please ask a member of your CF team.

Vitamin E

Vitamin E has become very popular because of its role as an *antioxidant*. As you grow your body goes through many changes. As your body changes it produces harmful products that can destroy your healthy cells. Vitamin E helps get rid of these harmful products and this is why it is called an *antioxidant*. A deficiency of vitamin E can affect your body's ability to protect itself when you get sick. For people with cystic fibrosis this may affect their lungs and their ability to breathe. Foods that contain vitamin E include plant oils, margarines, and some fruits & vegetables. Diet alone cannot prevent a vitamin E deficiency in people with CF, therefore a supplement is usually recommended.

Vitamin K

Most of the vitamin K we need comes from our diet. It is found in green vegetables, plant oils, and margarine. We also have healthy bacteria in our intestines that produce vitamin K. Even though the bacteria in your intestine is healthy, it can be destroyed by the antibiotics you may take for a lung infection. This can affect the amount of vitamin K you get on a daily basis. Vitamin K is very important because it helps your blood clot. When you cut yourself, the time it takes for your cut to stop bleeding is affected by how much vitamin K you have in your body. The longer it takes to 'clot' the less vitamin K you may have. Vitamin K is also important for proper bone growth in children. To prevent a deficiency many people with CF take a supplement that contains vitamin K.

Why should I take vitamins?

People with CF are at a very high risk for getting vitamin deficiencies. These deficiencies are serious and can include symptoms such as night blindness, bone fractures, back pain, and bleeding. They may also effect how quickly you recover from a lung infection. You can prevent these deficiencies just by taking your vitamins recommended by your CF clinic. To make sure get the most from vitamin supplements it is best to take them with food and enzymes.

Dietary Sources

A liver
egg yolk
dairy products
green leafy or yellow
vegetables

D liver
fish
fortified milk and margarine
egg yolk

E vegetable oils
nuts
eggs
wheat germ

K green leafy vegetables
liver
vegetable oils
bacteria in the intestine also
make vitamin K.



The CF Newborn Screening Program Support Resources: A Parent's Guide to BC Children's Hospital

1. I may have trouble getting to the hospital, is there any assistance to help our family?

There are some transportation services for patients and parents available, some of these resources may require a medical not or a social work referral. Financial assistance may also be available subject to a brief financial review by the organization itself

- TAP forms
- Shriners Care Cruiser 1-800-661-5437 (KIDS) www.shriners.bc.ca
- Hope Air 1-877-346-4673
- Wings of Hope (604) 231-6580
kidshorizon.wingsofhope@aircanada.ca
- First Nations Travel Insurance

2. Is there assistance for accommodations?

BC Family Residence Program -- Accommodation Assistance Program

All families whose children require medical care at BC Children's Hospital are encouraged to contact the BC Family Residence Program. To qualify for assistance with accommodation costs, families must:

- be residents of BC and have medical insurance coverage under the Medical Services Plan of BC;
- have a child who is receiving medical care at BC Children's Hospital; and
- live outside Metro Vancouver.

Families are encouraged to contact the program directly Monday to Friday 8:30-4:30 toll free at 1-866-496-6946 for accommodation assistance.

3. Where can I find parking?

There are pay parking lots with "patient only" locations reserved for patients from 8am to 2:30 pm weekdays. A pamphlet has been enclosed with more directions.

4. Where can I find a place to eat?

On the hospital grounds there is the Shaughnessy Café in B1. It is open Monday to Friday 7am to 7pm and on Saturday and Sunday 8am to 5:30 pm. There are microwaves and vending machines in this area.

There are also three coffee bars: Second cup in the main lobby of Children's Hospital, Starbuck's in the Ambulatory Care Building and Tim Horton's in the BC Women's Hospital lobby.

There is a Safeway and restaurants in the Strip Mall at 25th and Oak Street.

5. How do I find my way around?

The parking pamphlet has a small map of the hospital grounds. There is an information desk in the main lobby of the Children's Hospital.

6. I need to check my email and the internet during the day.

The Family Resource Library is located on the second floor of the Ambulatory Care Building. It is open Monday to Friday from 10am to 4pm