Cystic Fibrosis Infant Nutrition

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Newborn Screening

- Newborn screening is a nationwide program to find infants born with certain health conditions in order to begin treatment early to prevent serious, lifelong problems.

First few days of life
Heel prick
Blood spot on Guthrie Card
Prior to Newborn Screening
Severe CF Malnutrition at Diagnosis
(3 month old diagnosed during 2001 in a non-screening state)

Potentially fatal protein-energy malnutrition with salt depletion

Photo courtesy of Frank J. Accurso, MD
History of Cystic Fibrosis

- First described 1930s “celiac disease” and lung disease

- Dr. Dorothy Andersen Babies Hospital in New York
Salty Sweat

Dr. Paul di Sant’Agnese

Columbia Presbyterian Medical Center, 1937
Care of Infants Identified Through NBS

CF Care at an accredited care center

• Initial visit 24-72 hours of diagnosis
• Monthly visits to CF center (at a minimum) for the first 6 months

• *Sensitivity

At Initial Visit

• Diagnosis confirmed
• Basic genetic concepts
• Convey Difficult Facts
• Overview of symptoms
• Introduce Care Team
• Resources of Information
• Contact and follow up information
Initial Visit

- Knowledge about CF and factual information with straightforward answers (Tluczek, 2006).

- Education/support personalized to match the family’s immediate needs.

- Establish Coordination of Care with PCP with collaborative interdisciplinary team

- Need for supplemental salt

- Goal is $\geq 50^{\text{th}}$ percentile weight-for length
Pancreatic Insufficiency can Develop at Any Time

Bronstein et al, 1992
Recommendations: When (and When Not) to Start PERT

- Start PERT in infants with:
  - 2 CFTR mutations associated with PI or
  - lab evidence of PI or
  - unequivocal signs/symptoms of malabsorption*

- Do NOT start PERT in infants with 1 or 2 CFTR mutations associated with PS unless:
  - an objective test of pancreatic function indicates fat malabsorption or
  - unequivocal signs/ symptoms of malabsorption*

* pending lab results
Recommendation: PERT Dosing

• Initiate PERT at ½ to 1 capsule (“2,000-5,000 total lipase units”) at each feeding

• Adjust up to a dose of no greater than 2,500 lipase units/kg/feeding with a maximum daily dose of 10,000 lipase units/kg
Pulmonary Recommendations

- Infection Control (separate newly diagnosed)
- Smoke-Free environment
- Airway Clearance (initiate first few months of life)
- Baseline Chest x-ray with first few months
- Influenza prevention
- Microbiology (first culture by 1 month and quarterly; more often if symptoms)
- Antibiotic Therapy (staphyloccal and pseudomonas)
- Chronic Therapy
Early Disease

Early regional air trapping on expiration (arrows)


## Care Grid

<table>
<thead>
<tr>
<th>AGE AT VISIT</th>
<th>Day of Sweat test</th>
<th>24-48 hrs of dx</th>
<th>1 wk later or age 1 mo</th>
<th>2 mo*</th>
<th>3 mo*</th>
<th>4 mo*</th>
<th>5 mo*</th>
<th>6 mo</th>
<th>8 mo</th>
<th>10 mo</th>
<th>1 year</th>
<th>Every 2-3 months in the second year of life</th>
<th>24 mo</th>
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<td>INTERVENTIONS</td>
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<td>Encourage human milk feeding</td>
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<td>Start salt supplementation</td>
<td>1/3 tsp. salt</td>
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<td>1/4 tsp</td>
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<td>Start vitamins designed for CF patients</td>
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<td>History and physical with weight, length, OFC</td>
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<td>Teach / initiate P&amp;P</td>
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<td>Assess weight gain, caloric intake and PERT dose</td>
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<td>Serum electrolytes, BUN, creatinine</td>
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<td>Fill out “Who to call – where to Go” sheet</td>
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<td>Feeding/Behavior Anticipatory Guidance</td>
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<td>Referrals to community food resources</td>
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<td>Tobacco smoke exposure avoidance education</td>
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<td>Genetic counseling</td>
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Note: Some Centers may plan additional routine visits at 7, 9 and 11 months.
Consensus

• Phenotype more important than genotype
• CFF recommends genetic counselor discussion
• Communication with primary care to concurrently provide care
• Many infants with CRMS will be healthy
• Male higher risk of infertility
• Benefit from new treatments
• Update families as information becomes available
• Treat *P aeruginosa*. 
Infant Nutrition
## Nutrition for Newborn Infants with CF

<table>
<thead>
<tr>
<th></th>
<th>Birth-3 months</th>
<th>3-6 months</th>
<th>6 to 12 months</th>
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<tbody>
<tr>
<td><strong>Calorie goals</strong></td>
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<td></td>
<td>➢ 115-130 kcal/kg</td>
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<td>➢ 500-700 calories per day</td>
<td>➢ 100-110 kcal/kg</td>
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<td>➢ 700-800 calories per day</td>
<td>➢ 100 kcal/kg</td>
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<td>➢ 800-900 calories per day</td>
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<tr>
<td><strong>Weight gain goals</strong></td>
<td>25-30 g/day</td>
<td>15-21 g/day</td>
<td>10-13 g/day</td>
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<td><strong>Check vitamin levels</strong></td>
<td>X</td>
<td>X</td>
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<td><strong>Check Fecal Fat</strong></td>
<td>X (after 2 wks old)</td>
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<td><strong>Salt Supplementation</strong></td>
<td>1/8 tsp per day</td>
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<td>¼ tsp per day</td>
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(Borowitz et al: 2009)
Infancy is a time of dramatic metabolic demand: Healthy babies double their birth weight by 4 months of age and triple it by 1 year of age.
CF Infant Feeding Guidelines

Birth to 4 Months

• Rate of weight gain at 50th percentile or greater
• **Weight checks** every 2-4 weeks
• Infant formula or breast milk **every 3 hours**
• **Increase caloric density** of formula/breast milk at first sign of inadequate weight gain
• 1 ml per day of a **CF infant multivitamin** (can give 0.5 ml twice daily)
4-6 Months

- **Introduce** iron-fortified infant cereal, 2 servings/day. Use dry cereal made with formula or breast milk instead of water
  - If weight gain is low, **add** 1 tsp margarine per serving of cereal
  - **Continue** high calorie formula or breast milk
6-8 months

- Introduce pureed strained vegetables fruits, and meats as per infant feeding guidelines (one new food every 3-4 days)
- Add butter/margarine to food (1 tsp per 4 ounce serving)
- Add formula powder or instant breakfast powder to foods (1 Tbsp/serving)
- Begin finger foods such as cheerios and baby crackers
8-12 Months

- Critical age for introducing textures
- Mashed table foods with high caloric density
- Limit juice to 4 oz/day and give by cup instead of bottle
- Establish feeding schedule of 3 meals and 3 snacks per day
- Consider change to a 30 kcal/oz toddler formula
Our CF Center Infant Data
Percent of Infants with CF at > 50%ile weight for length at 12 mo
Feeding Patterns at 6 months of age

- % exclusively BF at 6 mo
- % fed BF/formula at 6 mo
- % exclusively formula fed at 6 mo
% of Infants >50%ile at 12 mo

- Exclusively BF Infants
- BF/Formula Fed Infants
- Exclusively Formula Fed Infants
The majority of infants in this group were exclusively formula fed

The majority of infants that meet the nutritional goal of >50%tile wt/l are pancreatic sufficient and exclusively formula fed
Maximizing Nutrition in the Exclusively Breast Fed Infant

- Frequent f/u in clinic to determine adequacy of weight gain
- Ensure enzymes are given with ALL feeds (even in the middle of the night)
- Ensure salt is given as directed
- Introduce bottle early, ≥3 months (also helps with salt intake)
- Pump and fortify breast milk to up to 28 cal/ounce
- Evaluate for zinc or iron deficiency if appetite seems poor and/or poor growth
- Evaluate need for acid-blocker
- Address concerns with constipation
References


• Hennekens and Buring, Epidemiology in Medicine, p. 327.
References continued


Questions and Discussion