

Treatment Recommendations for Infants with Cystic Fibrosis

Strength of evidence graded using the USPSTF grading system

Certainty of Net Benefit	Estimate of Net Benefit (Benefit minus Harms)			
	Substantial	Moderate	Small	Zero/negative
High	A	B	C	D
Moderate	B	B	C	D
Low	I (insufficient evidence)			

Question # / Recommendation

Strength of Evidence

Initial Diagnosis

1. The CF Foundation recommends that treatment for infants diagnosed with CF by NBS should be done at an accredited CF care center, with the goal on an initial visit within 24 – 72 hours of diagnosis (1 – 3 working days in absence of overt symptoms).

Consensus recommendation

Nutritional Recommendations

Pancreatic Function and Pancreatic Enzymes

2. For infants with CF under two years of age, the CF Foundation recommends that pancreatic function status should be measured by fecal elastase or coefficient of fat absorption in all individuals.
3. For infants with CF under two years of age, the CF Foundation recommends that pancreatic enzyme replacement therapy should be started.
 - in all infants with two CFTR mutations associated with PI.
 - in all infants with fecal elastase < 200 ug/g or CFA < 85% (in infants < 6 months of age), or other objective evidence of PI.
 - in all infants with unequivocal signs or symptoms of malabsorption, while awaiting confirmatory test results.
4. For infants with CF under two years of age, the CF Foundation recommends that pancreatic enzyme therapy should not be started in infants with one or two CFTR mutations associated with pancreatic sufficiency unless:
 - an objective test of pancreatic function indicates fat malabsorption; or
 - the infant has unequivocal signs or symptoms of malabsorption, while awaiting confirmatory test results.
5. For infants with CF under two years of age, the CF Foundation recommends that pancreatic enzyme replacement therapy be initiated at a dose of 2,000 – 5,000 lipase units at each feeding, adjusted up to a dose of no greater than 2,500 lipase units per kg per feeding with a maximum daily dose of 10,000 lipase units per kg.
6. For infants with CF under two years of age, as for patients of all ages, the CF Foundation recommends that generic, non-proprietary PERT should not be used.

Certainty: Low; Benefit: Substantial
Consensus recommendation

Certainty: Low; Benefit: Substantial
Consensus recommendation

Consensus recommendation

Certainty: Low; Benefit: Substantial
Consensus recommendation
Recommended in the CF Foundation Consensus Report on Nutrition for Pediatric Patients (35) and the European Consensus on Nutrition in Patients with CF (36)
Certainty: Low; Benefit: Moderate
Consensus recommendation
Recommended in the CF Foundation Evidence-based Practice Recommendations for Nutrition (consensus recommendation) (8)

Treatment Recommendations for Infants with Cystic Fibrosis – *continued*

Question # / Recommendation	Strength of Evidence
Nutritional Recommendations	
<p>Feeding, Vitamins and Micronutrients</p> <p>7. For infants with CF under two years of age, the CF Foundation recommends human milk as the initial type of feeding.</p> <p>8. For infants with CF under two years of age, the CF Foundation recommends that if infants are fed formula, standard infant formulas (as opposed to hydrolyzed protein formulas) should be used.</p> <p>9. For infants with CF under two years of age, the CF Foundation recommends that calorie-dense feedings should be used if weight loss or inadequate weight gain is identified.</p> <p>10. For infants with CF under two years of age, the CF Foundation recommends that positive feedings behaviors should be encouraged, such as by the provision of educational resources.</p> <p>11. For children aged 1 to 12 years with growth deficits, the CF Foundation recommends that intensive treatment with behavioral intervention in conjunction with nutritional counseling be used to promote weight gain.</p> <p>12. For infants with CF under two years of age, the CF Foundation recommends that multivitamins designed to provide at least the recommended levels of vitamins A, D, E, and K for patients with CF should be prescribed, beginning shortly after diagnosis.</p> <p>13. For infants with CF under two years of age, the CF Foundation recommends that blood levels of fat-soluble vitamins should be measured approximately two months after starting vitamin supplementation and annually thereafter; measure more frequently if values are abnormal.</p> <p>14. For infants with CF under two years of age, the CF Foundation recommends that a trial of zinc supplementation (1mg elemental zinc/kg/day in divided doses for six months) may be given to some infants who are not adequately growing despite adequate caloric intake and pancreatic enzyme replacement therapy.</p> <p>15. For infants with CF under two years of age, the CF Foundation recommends supplementation with ½ teaspoon table salt per day starting at diagnosis, increasing to ¼ teaspoon of table salt per day at 6 months of age.</p> <p>16. Patients ages 6 months to 2 years whose community water supply contains less than 0.3 ppm should be supplemented with fluoride 0.25 mg/d.</p> <p>17. For infants with CF under two years of age, the CF Foundation concludes that there is insufficient evidence to recommend for or against supplementation with linoleic acid.</p> <p>18. For infants with CF under two years of age, the CF Foundation concludes that there is insufficient evidence to recommend for or against supplementation with docosahexaenoic acid.</p>	
Pulmonary Recommendations	
<p>19. For infants with CF under two years of age, the CF Foundation recommends that a smoke-free environment be provided and that all caregivers are informed that cigarette smoke exposure harms children with CF.</p>	<p>Consensus recommendation</p>

Treatment Recommendations for Infants with Cystic Fibrosis – *continued*

Question # / Recommendation	Strength of Evidence
Pulmonary Recommendations	
Airway Clearance	
20. For infants with CF under two years of age, the CF Foundation recommends that airway clearance therapy be initiated in the first few months of life.	Certainty: Low; Benefit: Moderate Consensus recommendation
21. For infants with CF under two years of age, the CF Foundation recommends use of albuterol before percussion and postural drainage.	Certainty: Low; Benefit: Moderate Consensus recommendation
22. For infants with CF under two years of age, the CF Foundation recommends that the head-down position should not be used for percussion and postural drainage.	Certainty: Low; Benefit: Moderate Consensus recommendation
Infection Control, Surveillance and Treatment	
23. For infants with CF under two years of age, the CF Foundation recommends that newly diagnosed patients should be separated from other patients cared for in CF clinics until adequate infection control education has been provided to and is understood by the caregivers.	Certainty: Low; Benefit: Moderate Consensus recommendation
24. Infection control measures should be implemented in compliance with CF Foundation recommendations to minimize transmission of bacterial infections to infants.	Consensus recommendation Recommended in the CF Foundation Consensus Conference on Infection Control (95)
25. Annual influenza vaccination is recommended for infants with CF ≥ 6 months of age, all household members, and all health care providers caring for these infants. Household contacts and out-of-home caregivers of children with CF < 6 months of age also should receive annual influenza vaccine.	Consensus recommendation Recommended in American Academy of Pediatrics Guidelines (97) and CF Foundation Consensus Conference on Infection Control (95)
26. For infants with CF under two years of age, the CF Foundation recommends that use of palivizumab be considered for prophylaxis of respiratory syncytial virus.	Certainty: Low; Benefit: Moderate Consensus recommendation
27. For infants with CF under two years of age, the CF Foundation recommends that oropharyngeal cultures should be performed at least quarterly.	Certainty: Low; Benefit: Moderate Consensus recommendation
28. For infants with CF under two years of age, the CF Foundation recommends that bronchoscopy and bronchoalveolar lavage be considered in infants with symptoms or signs of lung disease, particularly those who fail to respond to appropriate intervention.	Consensus recommendation
29. For infants with CF under two years of age, the CF Foundation recommends against the prophylactic use of oral antistaphylococcal antibiotics in asymptomatic infants.	Certainty: Low; Benefit: Zero-negative Recommended in the CF Foundation Pulmonary Guidelines on Chronic Medications (88)
30. For infants with CF under two years of age, the CF Foundation concludes that there is insufficient evidence to recommend for or against active attempts to eradicate <i>Staphylococcus aureus</i> in asymptomatic infants.	Certainty: Low; Benefit: Unknown Grade: I recommendation
31. For infants with CF under two years of age, the CF Foundation concludes that there is insufficient evidence to recommend for or against active attempts to eradicate methicillin-resistant <i>Staphylococcus aureus</i> (MRSA) in asymptomatic infants.	Certainty: Low; Benefit: Unknown Grade: I recommendation
32. For infants with CF under two years of age, the CF Foundation recommends against the use of chronic antibiotics for prophylaxis to prevent <i>Pseudomonas aeruginosa</i> .	Certainty: Low; Benefit: Zero-negative Consensus recommendation
33. For infants with CF under two years of age, the CF Foundation recommends that new acquisition of <i>Pseudomonas aeruginosa</i> , 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.	Certainty: Low; Benefit: Moderate Consensus recommendation

Treatment Recommendations for Infants with Cystic Fibrosis – *continued*

Question # / Recommendation	Strength of Evidence
Pulmonary Recommendations	
Infection Control, Surveillance and Treatment	
<p>34. For infants with CF under two years of age, the CF Foundation recommends that infants who remain persistently colonized with <i>Pseudomonas aeruginosa</i> after two attempts at eradication be treated chronically with alternate-month tobramycin solution for inhalation.</p>	<p>Certainty: Low; Benefit: Moderate Consensus recommendation</p>
Diagnostic Testing	
<p>35. For infants with CF under two years of age, the CF Foundation concludes that there is insufficient evidence to recommend for or against use of pulse oximetry routinely as an adjunctive tool to detect disease.</p>	<p>Certainty: Low; Benefit: Small Grade: I recommendation</p>
<p>36. For infants with CF under two years of age, the CF Foundation recommends that pulse oximetry measurements be obtained in the infant with CF with acute respiratory symptoms.</p>	<p>Certainty: Low; Benefit: Substantial Consensus recommendation</p>
<p>37. For infants with CF under two years of age, the CF Foundation recommends that a baseline chest x-ray should be obtained within the first 3 to 6 months and once again within the first two years of life.</p>	<p>Certainty: Low; Benefit: Moderate Consensus recommendation</p>
<p>38. For infants with CF under two years of age, the CF Foundation recommends against the use of chest CT scans for routine surveillance.</p>	<p>Certainty: Low; Benefit: Zero-negative Consensus recommendation</p>
<p>39. For infants with CF under two years of age, the CF Foundation recommends that chest CT scans be considered in infants with symptoms or signs of lung disease who fail to respond to appropriate interventions.</p>	<p>Consensus recommendation</p>
<p>40. For infants with CF under two years of age, the CF Foundation recommends that infant PFTs be considered as an adjunctive tool to monitor respiratory status.</p>	<p>Certainty: Moderate; Benefit: Small Grade: C recommendation</p>
Chronic Pulmonary Therapies	
<p>41. For infants with CF under two years of age, the CF Foundation recommends that dornase alfa (recombinant human DNase) may be used in symptomatic infants.</p>	<p>Certainty: Low; Benefit: Moderate Consensus recommendation</p>
<p>42. For infants with CF under two years of age, the CF Foundation recommends that 7% hypertonic saline maybe used in symptomatic infants.</p>	<p>Certainty: Low; Benefit: Moderate Consensus recommendation</p>
<p>43. For infants with CF under two years of age, the CF Foundation concludes that there is insufficient evidence to recommend for or against the routine use of chronic azithromycin in patients colonized with <i>Pseudomonas</i>.</p>	<p>Certainty: Low; Benefit: Unknown Grade: I recommendation</p>
<p>44. For infants with CF under the age of two years without airway reactivity or asthma, the CF Foundation does not recommend use of inhaled corticosteroids to improve lung function or reduce exacerbations.</p>	<p>Certainty: Low; Benefit: Zero-negative Consensus recommendation</p>

Table adapted from *Cystic Fibrosis Foundation Evidence-Based Guidelines for Management of Infants with Cystic Fibrosis*