Ohio Department of Health • Children With Medical Handicaps

Standards & Outcome Measures

for Children With Chronic Pulmonary Disease

Developed By:
The Pulmonary Standards Committee
of Children With Medical Handicaps

Standards of Care for Children With Cystic Fibrosis

Definition

To provide state-of-the-art care for patients with cystic fibrosis, an autosomal recessive disease, diagnosed using the National Cystic Fibrosis Foundation criteria.

Goals

These standards are prepared to assure that children with cystic fibrosis achieve and maintain maximum habilitation or rehabilitation throughout life.

Care Needs

Medical, psycho-social, educational, vocational, recreational.

Team

Board certified pediatrician with cystic fibrosis training/experience; clinic coordinator; other physicians with appropriate pediatric training who have an interest in cystic fibrosis; registered nurse (may be clinic coordinator); CMH service coordinator; general pediatrician or primary care physician; geneticist; other on-site support services including social services, nutrition, and respiratory therapy.

Referral

Individuals with suspected or previously diagnosed cystic fibrosis should be referred to Children with Medical Handicaps' approved center that provides coordinated services. Hospitalizations for exacerbations of cystic fibrosis must be at cystic fibrosis centers. In cases of emergency admission to another facility, the patient should be transferred to a cystic fibrosis center when stable. All procedures requiring general anesthesia should occur at a CMH approved cystic fibrosis center. Some minor procedures with local anes-thetic (e.g., dental procedures) can be done at other facilities. All hospitaliza-tions must be approved by the CMH cystic fibrosis managing physician.

First Days of Diagnosis

Care needs met by cystic fibrosis team include the following: confirmation of diagnosis through appropriate testing; assessment of family needs; genetic counseling; development of therapeutic plan and overall therapy goals; education regarding airway clearance techniques, nutrition and respiratory care; emotional/family support; referral to appropriate community resources, including local Cystic Fibrosis Chapter; scheduling regular evaluations to include chest x-rays, respiratory cultures and pulmonary function tests; communication with primary care physician.

Content of Re-evaluation

As the child with cystic fibrosis grows and develops, additional needs become evident that require other disciplines and strategies. The following elements are recommended by age group without specifying the diagnostic or therapeutic interventions to be used. The listing is cumulative. A child entering the program for the first time would receive all items indicated through the actual age group as part of the initial evaluation. In addition, provision for appropriate and usual well-child care should be made in accordance with the American Academy of Pediatrics Standards of Child Health Care.

Emphases/Concerns of Re-evaluation

Toddler—Re-education in airway clearance techniques; counseling with regard to nutrition, pancreatic enzyme usage, vitamins, and supplements to maintain growth; prompt assessment and treatment for acute illness; promotion of family stability; appropriate laboratory tests.

Pre-school and Early School Age (3-9) -Review total schedule including therapies, medication, sleep pattern, postural drainage, school and recreational activities; education of the child about the disease; parent education to foster independence; initiate pulmonary function studies; encourage normalcy in school and extracurricular activities; beginning of self care.

Later School Age (9-12) - Beginning education of the child regarding sexuality, alcohol, drugs, and smoking.

Adolescent and Young Adult - Counseling with regard to financial and career planning and marriage; review of sexuality issues; referral to Bureau of Vocational Rehabilitation; and appropriate transition to adult services.

CMH Services

Requests for treatment services for children with cystic fibrosis must be made by a CMH physician provider from an approved cystic fibrosis center. Diagnostic services may be requested by a CMH approved pediatrician in consultation with an approved cystic fibrosis center. Service coordination services, where available, may be requested by a CMH-approved service coordinator at a participating center.

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Cystic Fibrosis Outcome Criteria

Outcomes	Process Measures
A. Quality Management	
1) Child's care for the cystic fibrosis condition is managed by an established multi-disciplinary team at an approved pediatric pulmonary center.	1 a) Referral made to the team at the time of diagnosis
	b) Treatment plan developed and implemented on the basis of team recommendations; plan updated at least annually.
	c) Family has access to appropriate providers with each visit.
	d) Care coordination is provided by the team; provision of care locally whenever possible.
	e) Complex diagnostic and surgical procedures restricted to major centers.
	f) Documentation of long and short term outcomes of the treatment plan.
	g) Family has access by phone to team members at times other than regularly scheduled appointment
	h) Team actively solicits family participation and collaboration in treatment plan.

2) Service coordination is provided at both the team and local levels.	2 a) The child has identified team and local service coordinators. b) Documentation of a comprehensive service plan, developed by the family/child, team and local service coordinators, which addresses the medical, social, vocational, educational and recreational needs of the child. c) The comprehensive service plan is updated at least twice yearly. d) The family has input into the comprehensive service plan and demonstrates an understanding of, and agreement with, the content of the plan.
3) Appropriate pediatric care is provided throughout childhood.	3 a) Has an identified primary care physician who is an extended member of the team. b) Physical exams and well-child care provided on a regular basis. c) Immunizations are current. d) Influenza vaccine given to client, family and other caregivers. e) Periodic monitoring for growth failure, delayed development or other problems. f) Mutual communication between the specialty team and primary care provider regarding the child's plan of care, progress and special needs/problems. g) Family demonstrates knowledge of child's routine health needs and treatment plan.
4) Preparation for transition to adult care is initiated at an age-appropriate level.	4 a) A transition plan is developed during the teenage years. b) The child and family have input into the transition plan. c) Qualified adult care providers (especially physicians) are available to assume care at the appropriate time.

B. Physiological

1) Maintains adequate nutritional status.	1 a) Documentation of periodic nutritional assessments. b) Documentation of weight for height (length) ratio per National Center for Health Statistics (NCHS) growth grids. c) Family demonstrates ability to give prescribed diet and supplemental feedings.
2) Child maintains adequate gastrointestinal function.	2 a) Parent/child recognizes signs and symptoms of bowel obstruction and other GI problems and appropriate actions to take. b) Parent/child demonstrates knowledge of appropriate use of enzyme supplementation and prescribed diet.

3) Child attains and maintains optimal pulmonary function.	3 a) Parent/child demonstrates knowledge of use of ventilatory aids and appropriate action to take if problems or malfunctions occur. b) Parent/child recognizes the sign and symptoms of pulmonary infections and identifies appropriate actions to take. c) Parent/child can identify actions and side effects of prescribed medications and can demonstrate correct administration. d) Periodic assessment of trends of pulmonary status (blood gases, oxygen saturation, peak flow etc.). e) Parent/child demonstrates knowledge of proper pulmonary hygiene technique.

C. Functional

1) Child achieves appropriate developmental milestones.	1 a) Participates in age appropriate activities.

D. Psychosocial

1) Psychosocial response to handicapping condition is age appropriate.	1 a) Achievement of age appropriate socialization skills. b) Parent/child have information about support groups/peer groups. c) Child demonstrates positive expressions of self esteem. d) Child participates in recreational activities according to level of tolerance.
2) Educational/vocational goals achieved.	2 a) Receives education in least restrictive environment with appropriate related services. b) Achieves age appropriate grade levels. c) Achieves employment/post-secondary educational placement.

E. Family Satisfaction

1) Parent/child achieves satisfaction with outcomes of treatment.	1 a) Parent/child have input into care plan. b) Parent/child have access to choice of providers. c) Parent/child demonstrate knowledge of cystic fibrosis condition, complications, treatment plan, available resources, parenting skills, and support opportunities. d) Parent/child receives information about genetic counseling services available.

Bronchopulmonary Dysplasia (BPD) Outcome Criteria

Outcomes	Process Measures
A. Quality Management	
1) Child's care for the BPD condition is managed by an established multidisciplinary team at approved pediatric pulmonary center.	1 a) Referral made to the team for initial evaluation within the first month of life.
	b) Treatment plan developed and implemented on the basis of team recommendations; plan updated at least annually.
	c) Family has access to appropriate providers with each visit to the team.
	d) Care coordination is provided by the team; provision for care locally whenever possible.
	e) Complex diagnostic and surgical procedures only provided in (restricted to) major centers.
	f) Documentation of long and short term outcomes of the treatment plan.
	g) Family has access by phone to team members at times other than regularly scheduled appointments.
	h) Team actively solicits family participation and collaboration in treatment plan.
2) Service coordination is provided at both the team and local levels.	2 a) The child has an identified team and local service coordinator.
	b) Documentation of a comprehensive service plan, developed by the team, family and local service coordinators, which addresses the medical, social, vocational, educational and recreational needs of the child.
	c) The comprehensive service plan is updated at least twice yearly.
	d) The family demonstrates an understanding of, and agreement with, the content of the plan.
3) Appropriate pediatric care is provided.	3 a) Has an identified primary care physician who is an extended member of the team.
	b) Physical exams and well-child care provided on a regular basis.
	c) Immunizations are current.
	d) Periodic monitoring for growth failure, delayed development, or other problems.
	e) Mutual communication, regarding the child's plan of care, progress, and special needs/problems, between the specialty team and primary care provider.
	f) Family demonstrates knowledge of child's routine health needs and treatment plan.

4) Transition from neonatal care to pulmonary management is initiated at an age appropriate level.	g) Influenza vaccine given to client, family, and other caregivers.
	4 a) A transition plan is developed jointly by neonatal and pulmonary providers.
B. Physiological	
1) Maintains adequate nutritional status.	b) The family has input into the transition plan.
	1 a) Documentation of periodic nutritional assessments
2) Child attains and maintains optimal pulmonary function.	b) Documentation of weight for height (length) ratio, per National Center for Health Statistics (NCHS) growth grids.
	2 a) Appropriate use of ventilatory assistance, oxygen, and tracheostomy as needed.
	b) Parent demonstrates knowledge of use of ventilatory aids and appropriate actions to take if problems or malfunctions occur.
	c) Parent recognizes the signs and symptoms of pulmonary infections and identifies appropriate actions to take.
	d) Parent can identify actions and side effects of prescribed medications and can demonstrate correct administration.
C. Functional	e) Periodic assessment of trends of pulmonary status (blood gases, oxygen saturation, peak flow etc.).
1) Child attains and maintains maximal motor skills.	
2) Child achieves appropriate developmental milestones.	1 a) Parent demonstrates knowledge of treatment plan to promote development.
D. Family Satisfaction	2 a) Periodic assessment of developmental progress
1) Parent achieves satisfaction with outcomes of treatment.	b) Child participates in age appropriate activities.
	1 a) Parent has input into care plan treatment.
	b) Parent has access to choice of providers.
	c) Parent demonstrates knowledge of BPD condition, complications, treatment plan, available resources, parenting skills, and support opportunities.

Children with Medical Handicaps
Pulmonary Standards Committee

Robert Stone, M.D., Chairperson
Carl Doershuk, M.D.
David Hufford, M.D.
Karen McCoy, M.D.
William Spohn, M.D.
Pierre Vauthy, M.D.
Robert Wilmott, M.D.
Gordon Young, M.D.

CMH Representatives

James Bryant, M.D.
Rosemary Feka, R.N.
Karen Lane, R.N., M.A.
Kathie Myers, R.N., B.S.N.
Ann Weidenbenner, M.S., R.D., L.D.

Children With Medical Handicaps
P.O. Box 1603
Columbus, Ohio 43216-1603
(614) 466-1700

Ohio Department of Health

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