## GUIDELINE STATEMENT

**CYSTIC FIBROSIS** The guideline outlines the management of patients with cystic fibrosis as required by the Children’s Rehabilitative Services Program, Arizona Health Care Cost Containment System, State of Arizona.

## PURPOSE

Clinical Practice Guidelines represent the minimum requirements for providing care for individuals with cystic fibrosis. Care and treatment should be provided in a manner that includes adherence to and consistency with each of the following Guidelines.

## DEFINITIONS:

**Children’s Rehabilitative Services (CRS):** An AHCCCS program for children with certain diagnoses which provides services using an integrated family-centered, culturally competent, multi-specialty, interdisciplinary approach.

**Multi-Specialty Interdisciplinary Clinic (MSIC):** The Specialty Medical Home for the members with diagnoses as designated by the Arizona Administrative Code (AAC) R9-7-202 (R9-22-1303, 10-1-2013).
I. PROCEDURAL GUIDELINES for POLICY COMPLIANCE

A. CRS Enrollment:
Children diagnosed with cystic fibrosis must be seen at a CRS Multispecialty Interdisciplinary Clinics (MSIC) site meeting these guidelines. Care provided to members under CRS in any other part of the state must be coordinated with the designated Cystic Fibrosis Team.

B. Interdisciplinary Team Membership:
The following Team Members must be present during MSIC sites and team conferences to review the patient information and determine the need to see the patient at a MSIC site. The Team Members must be available for inpatient consultation or coordination of care with inpatient staff:
- Child Psychologist
- CRS member / Caregiver
- Dietician/Nutritionist
- Primary Care Physician (Invited)
- Pulmonologist – Lead Physician
- Registered Nurse Coordinator
- Respiratory Therapist
- Social Worker

C. Available Personnel:
The following personnel must be available to the member at the MSIC Pulmonology Clinic:
- Advocate
- Child Life Specialist
- Educator
- Translator

D. Consultative Personnel:
The MSIC site must have access for consultation to specialists including, but not limited to the following:
- Allergist/Immunologist
- Anesthesiologist
- Angiographer
- Cardiologist, pediatric and adult
- Endocrinologist
- Gastroenterologist
- Geneticist / Genetic Counselor
- Infectious Disease Specialist
- Internist
- Nuclear Medicine specialist
- Otolaryngologist
- Pediatrician
- Pulmonologist
- Radiologist
- Surgeon, pediatric, general thoracic
- Urologist

E. Outreach Clinics:
Outreach Clinics are designed to provide a limited specific set of services including evaluation, monitoring and treatment in settings closer to the family than a MSIC site. Major treatment plan changes must be communicated to the MSIC site. Cystic Fibrosis Outreach / Field Clinics must include the following personnel.
• Clinic Coordinator  
• Pulmonologist  
• Respiratory Therapist

F. Community Based Services:
Community based services means all local services including provider agencies, schools, private physician offices, hospitals, and/or any other local setting.
The following community based services may be provided for patients with cystic fibrosis:
• Any other appropriate extension of services as approved by the Interdisciplinary Team and approved by the CRS Medical Director
• Lab Work
• Pharmacy Services
• Respiratory Therapy

G. Services to Adults with Cystic Fibrosis:
1. Clinic times separate from the member MSIC will be scheduled for adults
2. Child Life Specialist should be removed from the Team

H. Facilities and Services:
   i. Age-appropriate setting for all patients with cystic fibrosis
   ii. Defined age-appropriate services/i.e. Pediatrics, Adolescent Medicine and/or Internal Medicine
   iii. House Officers when the hospital is a teaching institution
   iv. Identified clinic area
      a. “Clean” Clinic separate area for members who have not developed colonization.
   v. Laboratories performing:
      a. Sweat test by quantitative pilocarpine iontophoresis as stated in the Cystic Fibrosis Foundation guidelines.
      b. Genetic Testing when meets criteria as per AHCCCS Medical Policy Manual Chapter 310 N.
      c. Pulmonary function tests, including ability to measure long volumes
      d. Daily, round the clock availability of:
          i. Bacteriology
          ii. blood chemistry
          iii. arterial blood gases
          iv. imaging studies including radiographic, ultrasonographic, and nuclear medicine studies.
   vi. Nutrition or Dietary Department
   vii. Pediatric and Adult Intensive Care Units
   viii. Respiratory Care Department
   ix. Respiratory therapy available 24 hours a day, 7 days a week
   x. Social Work Department

I. Other Criteria:
1. Written patient records to include, but not limited to:
   a. Documentation of positive sweat test and/or genetics testing
   b. Copies of outpatient clinic visits
   c. Discharge summaries
   d. Growth chart
   e. Laboratory data
2. Sweat Test results reviewed by Lead Physicians
3. Genetic Testing reviewed by lead physician and /or geneticist
4. Referral plan for procedures or services not available through CRS, such as oxygen at home, equipment, supplements, therapy vest, etc.
5. All outpatient clinic reports and discharge summaries sent to the referring physician
6. A plan for the care of transitioning to adult care and for adult patients
7. 24 hour coverage by a lead physician
J. Team/Staff Meetings:
1. Case Planning Meetings – a meeting of the specialist involved in the care and treatment of the member is to be held after each specialty clinic
2. Interdisciplinary Team Meetings / review and planning meetings (patient specific):
   a. Every three (3) months with the regular assessment
   b. Once a year for planning and review with the family
3. Staff Meetings at least annually to focus on issues of MSIC patient care and MSIC administration
4. Internal education meetings annually to focus on new information regarding the care and treatment of persons with Cystic Fibrosis
5. A yearly note by Social Services, Dieticians, and Child Life required.

K. Lead Physician Specialists:
Qualifications: The lead physician specialist for members with cystic fibrosis will be a pulmonologist, Pediatrician, or Internal Medicine with experience in the care of cystic fibrosis patients. Board certification in Pulmonary Medicine is recommended.

II. GUIDELINES FOR PATIENT SERVICE, EVALUATION, AND MONITORING FOR CYSTIC FIBROSIS
The purpose of this guideline is to promote a uniform level of care and teaching services at CF Centers, and to provide a general framework for good patient care. Their relevance to specific situations will depend on individual variations in clinical course and professional judgment as well as member and family input into treatment decisions based on their goals, cultural needs, strengths and barriers to treatment goals. In addition, this document should serve as a tool to assess programs, secure resources needed to enhance patient care and education, and guide the future growth and development of CF care.

A. Diagnosis:
   Goal: To provide accurate and timely diagnosis of CF.
   i. The diagnosis of cystic fibrosis is based on clinical and laboratory findings. These may include but are not limited to:
      a. chronic obstructive pulmonary disease,
      b. intestinal malabsorption,
      c. electrolyte loss through sweat,
      d. family history of CF,
      e. meconium ileus at birth,
      f. male infertility due to azoospermia,
      g. presence of staphylococcus aureus or mucoid pseudomonas aeruginosa in the respiratory tract.
   3. ii. Confirmation of the diagnosis currently requires
      a. two positive sweat tests, done on different days, by quantitative pilocarpine iontophoresis (Gibson-Cooke) according to Cystic Fibrosis Foundation approved methods
      A positive sweat chloride test is defined by sweat chloride measurements in excess of 60 milliequivalents per litre in an adequate sample of sweat (minimum of 75 milligrams in gauze or filter paper, or 15 microliters for the Wescor Collection system, collected over a 30 minute period). Repeat borderline sweat electrolyte measurements (40-60 meq/ml) require clinical correlation and judgment for diagnosis. 2
      b. 2 CF mutations known to cause disease.

B. Evaluation and Education of Newly Diagnosed Patients:
   Goal:
   i. To provide accurate assessment of physical and emotional status, and to begin patient family
Evaluation of the newly diagnosed patient should include
   a. medical,
   b. nursing,
   c. nutritional,
   d. psychosocial,
   e. respiratory, and physical therapy assessments as well as laboratory evaluation and genetic counseling.
   f. A comprehensive education program must be developed to promote
      1. optimum understanding of the disease,
      2. adherence to treatment plans and
      3. adequate coping with the demands of chronic illness.

C. Ongoing Patient Evaluation and Monitoring:
   **Goal:** To anticipate and treat physical and psychosocial problems and complications of the disease.

   At least four visits per year to the Clinic are recommended. The number of visits will vary with factors such as age, degree of illness; time elapsed from diagnosis, and distance from a clinic. Use of clinical scoring at every visit is encouraged. In addition, interim visits to the primary care physician for general pediatric care are necessary. The primary care practitioner has an important role in administering immunizations, evaluation and treatment of milder pulmonary exacerbation in consultation with the CF Center physician, advocacy and assessment of family dynamics. Every patient should be encouraged to be seen on a regular basis at a CF Clinic and by a physician in the community and records shared for coordination of care.

D. Respiratory Evaluation and Therapy:
   **Goal:** To achieve optimum respiratory status. To anticipate and treat progression and complications of pulmonary disease.

   i. Complete respiratory history and examination should be obtained at every visit (including nasal examination for nasal polyps).
   ii. Spirometry is recommended to quarterly and more often if clinically indicated and during hospitalizations for pulmonary exacerbation. Spirometry should be done at every for analysis at routine visits. Complete pulmonary function testing (including lung volumes) should be done at least once a year (performed according to American Thoracic Society Guidelines).
   iii. Arterial blood gases or pulse oximetry needs to be done at least annually on patients whose forced expiratory value in one second (FEV1) is less than 40% of predicted normal and additionally, when clinically indicated (i.e. exacerbation, oxygen therapy).
   iv. Respiratory tract culture and sensitivity should be done at least four times per year but preferably at each quarterly visit, before initiation of intravenous antibiotic therapy and at sick visits when clinically indicated.
   v. Chest roentgenogram should be obtained annually and on Pulmonary-related hospital admissions. Scoring of radiographs is encouraged.
   vi. All clinics should have written protocols for managing respiratory complications (hemoptyisis and pneumothorax).
   vii. Knowledge and performance of respiratory and physical therapy techniques should be evaluated annually.
E. **Gastrointestinal System/Nutrition:**

**Goal:** To anticipate and treat nutritional deficits and complications. The ultimate goal is to achieve optimum growth and nutrition.

i. Measurements should include height and weight, plotted on Guideline growth chart, for all patients, every visit. Other measurements such as triceps skin fold thickness and mid-arm circumference can be useful.

ii. Nutritional assessment should be carried out annually and when there is evidence of weight loss or poor weight gain. This assessment should include, but not be limited to:
   a. protein, fat, carbohydrate,
   b. vitamin and
   c. mineral intake.

iii. Other measures include:
   a. Assessment of pancreatic enzyme and
   b. vitamin supplementation and
   c. measurement of albumin and/or prealbumin levels at diagnosis, and when indicated;
   d. Abdominal examination with particular attention to liver and spleen size and consistency;
   e. A protocol for the management of diabetes mellitus;
   f. Laboratory measurements to include evaluation of
      i. metabolic and liver status,
      ii. complete blood count, and
      iii. fat soluble vitamin levels.

2 Am. Rev. Resp. Dis. 1987; 136; 1285-96

F. **Psychosocial Issues:**

**Goal:** To anticipate and treat social and emotional problems of patients and their families.

i. Psychosocial assessment should be carried out annually.

ii. The CF Clinic staff should be available for
   a. genetic counseling,
   b. crisis management,
   c. ongoing support and anticipatory guidance, when indicated.
   d. Sexuality, fertility and pregnancy should be discussed at age-appropriate intervals.

G. **Adult Issues:**

**Goal:** To ensure that the changing needs of the growing population of adult patients are met by caregivers.

Adult patients have specific needs different from those of the pediatric patients. Clinics should have clear plans for the care of adults including identification of appropriate caregivers and preparation for transition to adulthood and adult care. Adequate actions to address these needs include the

i. incorporation of adult care specialists in the clinic program,
ii. establishment of inpatient services in internal medicine wards,
iii. creation of transition teams or creation of parallel adult care teams.

The specific activities undertaken will depend on clinic size, geographic factors, local institutional idiosyncrasies and availability of specialists in the community.
Specific areas requiring services include:

i. obstetrics and gynecology,
ii. urology,
iii. cardiology,
5. CF Foundation Consensus Statement “Use of Pancreatic Enzyme Supplements for Patients with Cystic Fibrosis in the Context of Fibrosing Colonopathy”, March 1995