

CYSTIC FIBROSIS CLINICS AS A MODEL OF INTERPROFESSIONAL CARE

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INTRODUCTION

Cystic fibrosis is one of the most common serious genetic disorders and there are approximately 30,000 CF patients in the U.S. CF is a chronic and complex disease that affects many systems, including the respiratory and GI tract. Improvements in early diagnosis (now primarily made by newborn screening) and supportive care have improved survival for CF patients and most CF patients now survive into adulthood. Among chronic illnesses, CF care is unique in that most care is received in CF Foundation accredited care centers (>120) which are multidisciplinary, peer-reviewed, and follow evidence and expert-based guidelines for diagnosis and care. Because of the complexity of cystic fibrosis and the multiple systems involved, numerous medical specialists participate in CF care centers including adult and pediatric specialists in pulmonology, gastroenterology, infectious diseases, endocrinology, genetics, as well as others. In addition, CF center care teams typically include nurses, NP's, nutritionist/dieticians, RT, PT, pharmacists, and genetic counsellors.

CF FOUNDATION CARE CENTERS

CF care centers provide CF care to CF patients and follow CF Foundation guidelines for team organization, services provided, and are peer-reviewed before accreditation is awarded. CF centers generally range in size from 50 to 100+ patients, and must offer a mechanism for transition of pediatric CF patients to adult CF care. Adult CF care centers are generally less well developed than pediatric care centers but with over half of current CF patients over 18 years of age, adult CF care centers will be increasingly important as sites of CF care. CF centers receive a small grant from the CF Foundation to help support the infrastructure and administrative cost of the CF care center but the CF Foundation does not provide direct support for patient care costs which are generally provided by insurance, Medicaid or other forms of support. In addition to

providing patient care, most CF care centers participate in CF clinical research. Most CF patients and families participate in the CF Registry, a large registry of CF patients which includes information on genetic mutations, clinical features like lung function, cultures, height and weight percentiles, CF therapies, respiratory tract cultures, and presence of CF-related complications such as diabetes. The CF Registry provides the basis for public reporting of CF Center outcomes which are provided on the CF Foundation website and are available to families and patients.

CARE DISCIPLINES: ROLES

Physicians:

Center director

The CF center director is responsible for organizing and administering the CF care center and is usually a specialist in pulmonology or gastroenterology. Adult programs are required to have directors who are trained in adult medicine.

Pulmonologist

Since pulmonary complications are the major morbidity in CF, the majority of CF clinicians are specialists in pediatric and adult pulmonary medicine.

Gastroenterologist

GI complications are very common in cystic fibrosis and having GI specialists to deal with difficult GI complications, including liver disease, persistent difficulty in gaining weight, and chronic GI symptoms despite pancreatic enzyme supplementation is important. Cystic fibrosis-related liver cirrhosis is one of the most common indications for liver transplantation and patients who require liver transplant generally have milder lung disease and their subsequent follow-up generally requires follow-up with the liver transplant team to deal with frequent transplant related complications.

Endocrinologist

CF-related diabetes mellitus begins to appear around age 10 and continues to increase in prevalence throughout adulthood so that up to 50% of adult patients have CF-related diabetes or abnormal glucose tolerance. Poor control of CFRD can worsen lung function, weight gain, and survival so follow-up with an endocrinologist familiar with CFRD is generally recommended.

ENT

Chronic sinus disease is universal in cystic fibrosis and many patients require sinus surgery and removal of nasal polyps (which can cause nasal obstruction, worsen sinus disease, and affect appetite).

Hospitalist

Many inpatients with cystic fibrosis are cared for by hospitalists, who need to be familiar with CF care guidelines and infections common in CF.

Infectious disease specialists Chronic lung infection is universal in cystic fibrosis and generally include infections with organisms such as Staphylococcus aureus (including methicillin-resistant strains), Pseudomonas aeruginosa, and Stenotrophomonas maltophilia. More resistant strains of Pseudomonas, Burkholderia species, atypical Mycobacterial infections often require involvement of infectious disease specialists.

Genetics

Geneticists have been involved in setting up universal newborn screening for cystic fibrosis in all states and with over 1900 mutations described in the CFTR protein that is abnormal in CF, genetic counseling for CF patients is a complex topic. Prenatal screening and counseling is also provided for couples at risk for having a child with cystic fibrosis.

Nurses

All CF centers have a coordinator and in most centers a nurse experienced in cystic fibrosis provides this role. Nurses also work in the CF care center clinics, inpatient units, communicate with patients and families after clinic, and help deal with insurance, medication refills, and intercurrent illnesses.

Social worker

Cystic fibrosis is a complex, stressful disease for families and the social worker provides a critical role in helping patients and families deal with issues of insurance coverage, divorce and family stress, support for siblings, and other important issues that complicated medical care.

Respiratory care

The majority of CF patients utilize some sort of daily lung secretion mobilization technique (chest physiotherapy) and RT's often teach and supervise these techniques. Routine assessment of lung function (pulmonary function testing) is done at every CF clinic visit and is often done by a RT or pulmonary function technician with specialized training in lung function. Lung function testing can be done even in infants. Many medications used in CF patients are delivered by aerosols and RT's teach families how to use these devices.

Physical therapy

In some centers, chest physiotherapy is performed by PT rather than RT, and PT's in the clinic assess patient physical activity, encourage routine exercise, and look for chest wall deformity, joint complications, and other factors that may impact function.

Nutritionist/dietician

Good growth and nutrition is critical to the newborn with cystic fibrosis, and the goal of CF care centers is to achieve normal growth (BMI of 50th percentile). Dieticians counsel new families on the importance of a high fat, high calorie diet, and fat-soluble multivitamin supplements. Most patients require pancreatic enzyme supplementation and dieticians assist in calculating the correct dose of pancreatic enzyme supplements. In older patients, supplemental feedings are often needed and may include appetite

stimulants, NG feedings, and G-tube/nocturnal feedings in order to achieve optimal weight gain, or maintain weight in older patients. There is a direct relationship between BMI percentile and lung function.

Genetics counselor

Genetics counselors work with geneticists and CF teams to help families; genetic counseling is particularly important in states where genetic information is provided at the time of newborn screening. Many CFTR mutations found at newborn screening are associated with no clinical disease and individuals who are carriers for CFTR mutations generally are well and it is important to counsel these families.

Child life

Child life focuses on reducing the stresses associated with procedures such as IV placement, blood drawing, bronchoscopy, G-tube placement, and other procedures that CF patients frequently require. Hospitalizations for

Pharmacist

CF patients generally have a complex regimen of chronic medications to maintain lung health and adequate growth and pharmacists can be helpful in education of families. Most CF patients require regular admission to the hospital for IV antibiotics and pharmacists assist with inpatient planning, measuring antibiotic drug levels, and avoiding harmful drug interactions or toxicity. In some CF centers pharmacists also work in the clinic to review outpatient medications.

Clinic staff

The efficient flow of CF patients through the CF clinic depends upon having an efficient clinic team of coordinators, patient assistants and other staff to minimize wait time and help minimize the time patients and families have to wait in clinic.

APPLICATION OF CLINICAL MICROSYSTEMS TO CF CARE TEAMS

Clinical microsystems are multidisciplinary teams of frontline health care workers and the concept of clinical microsystems was developed primarily at Dartmouth Medical School.(REF) Clinical microsystem theory is ideal for a disease and care system like cystic fibrosis care and an excellent tool for analyzing CF care center teams has been developed by the CF Foundation and made available to CF care teams (REF).

DATA AND DATA TRANSPARENCY IN CYSTIC FIBROSIS CARE CENTERS

The CF Registry provides CF care centers with a wealth of data on CF outcomes. In addition to a center's individual outcomes, center data are provided as a bar graph compared to all CF care centers. In addition the CF Center's data for several key measures are provided over time so that trends in improvement (or worsening) are shown. This provides important feedback for CF care teams on their performance and the CF Foundations mandates that the CF center director share the results of the CF Foundation Registry report with all members of the CF care team. In addition to care

teams, most CF care centers now share the CF registry with patients and families and/or patient/parent advisory groups.

CF FOUNDATION QI TRAINING

The CF Foundation has provided a unique focus on quality improvement training for CF care teams and has funded a series of Learning and Leadership QI Collaboratives to train CF care team members in QI. The resources developed by CF care teams are made available by the CF Foundation on the internet and the annual CF care conference includes numerous sessions on quality improvement initiatives by discipline.

TRANSITION TO ADULT CARE

One of the biggest challenges in CF care has been

CLINIC FLOW

A number of tools have been adopted by CF center teams to assist in clinic flow and making maximal use of patient and family time in clinic:

Team huddle: the team huddle is generally done before clinic to review the patients to be seen in the clinic that day, what studies are planned and if there are specific problems that need to be addressed.

Post clinic conference: After patients are seen the post clinic conference provides an opportunity for all team members to hear about all the patients, and discuss issues that have come up during the busy CF clinic visit

Team retreat: The team retreat away from the CF center gives the CF team an opportunity to focus on improving procedures, reviewing CF registry data to highlight outcomes (e.g. CF-related diabetes screening, use of recommended CF therapies, influenza vaccine rates) that need improvement; updates n research and review of clinical topics by team members are often done during retreats; many patient/parent advisory groups also participate in team retreats

QI projects: Improved outcomes requires the application of improvement methods, such as PDSA (plan-do-study-act) rapid improvement cycles and measurement of outcomes over time using run charts and other visual displays of results

Clinic board: A complex clinic with multiple caregivers benefits from a central communication board showing what team members need to see (or have already seen) the patient, what tests are required, and other details of the visit.

Pre-clinic review: A review of the clinic is essential to planning the team huddle and having an efficient clinic. This is usually done by a nurse or nurse coordinator 1-5 days prior to the actual clinic.

CF Care and primary care/health homes

CF care centers generally do not provide primary care and work with primary care practitioners (pediatricians, family practice, internal medicine) who provide immunizations, routine care, and care for non-CF related complications. Many CF patients live a long distance from the CF care center and it is impractical for them to travel to the CF care for routine care. CF patients can develop non-CF related complications such as heart disease, renal complications, etc. that are best screened for and cared for by primary care practitioners. Some complications such as diabetes which are frequently cared for by primary care practices, have significant differences in patients with cystic fibrosis and require a different approach best provided by a specialist with experience in CF and familiar with evidence based guidelines for CF.