CF: Information for Case Managers

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About your speaker
25+ years in pulmonary
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Grandmother of 2 year old twins who are cute, cute, cute!
Cystic Fibrosis

* 101
* Where we are today
* What’s made the difference?
* Cost
* Hopes for the future
Register Now!
April 13, 2012

Updates in Pediatric Pulmonary Care XII

Continuing Education Credits pending approval through Suwannee River AHEC

Other topic highlights include:

- Advances in CF Testing
- Clinical Insights Improving Patient Outcomes in CF
- Sleep Disorders: Got Sleep!
- New Directions in CF Management
- Health Literacy
- Recent Developments in Exercise Testing

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Featuring:
Michael Boyle, MD FCCP
Johns Hopkins University
Cystic Fibrosis Center

Dr. Boyle will present during a special morning & lunch plenary session.

Take advantage of this learning opportunity & register today.

Register at www.ahecregistration.org & select Suwannee River region

Special registration rates:
40.00 - Full Day including CE credit
10.00 - Lunch Plenary Session Only

Families of the PPC are invited to attend as our guest, but must register selecting this option.
A chronic life-shortening, genetic disorder that primarily affects the lungs and the gastrointestinal tract
Lung disease is the predominant cause of illness and death (68.3%)
Lower Respiratory Manifestations

- Persistent cough/wheeze
- Recurrent pneumonia
- Abnormal PFT’s
- Abnormal CXR
- Organisms
- Sputum production
Blocked pancreatic duct
* Malabsorption
* Obstruction
* Bloating/Discomfort
* Large, greasy stools
* Prolapse
Inheriting CF
Autosomal Recessive Gene

Mom
CF/N

Dad
CF/N

CF/CF
CF/N
CF/N
N/N

Same risk with EVERY pregnancy!
1 in 3200 Caucasians 94.7%

1 in 15000 African-Americans 4%

Hispanic 6.9%

rare in Native Americans & Asians
Respiratory 20.5%
Abnormal Stools 7.8%
Electrolyte Imbalance
Mec Il/Intestinal Obst 8.8%
Neonatal screening 57.5%
Rectal prolapse 0.9%
Nasal polyps/sinus 3.5%
FTT/Malnutrition 9.7%
Family History 13.4%
Prenatal screening 3.6%
Where could you come in?
Percent of New Diagnoses by Newborn Screening

1990-2009
Costs

Families
- Psychosocial
- Time
- Money

CF Centers
- Staff
- Care management
- Prior authorizations

Payors
- More and more costly medications
- Equipment
Patients & Families

- Medications
  - 2-3 hours/da
  - Copays
  - Access

- Airway clearance
  - 1 hour/da
  - Costly? Cumbersome?

- Hospitalizations
  - Time/money/work
  - loss/pain/sleep disturbance

- Clinic visits
  - Time/money/fear

- Emotional drain of chronic illness/fear/restrictions
CF Care Centers

- CFF Accreditation Requirements
  - Interdisciplinary team
  - Facility requirements
  - Data reporting
  - Standards of care: know them... use them!
  - QI projects
  - National meetings

- Barriers to care
  - Prior authorizations
  - Access to care/assistance
  - Economy
  - Family factors
Third party payors

* Determination of “medical necessity”
  * Science
  * Art
  * Kitchen sink
* Increasing cost of new drugs/therapies
* Cost shifting to providers?
Mechanics of success
Patients Evaluated by a Respiratory Therapist, by CF Center

Lung function testing, airway clearance, correct administration of medications is pivotal.

The national rate is 89.2 percent (green bar).
The range is 20 to 100 percent.
Patients Evaluated by a Social Worker,
by CF Center

The national rate is 77.3 percent (green bar).
The range is 0 to 100 percent.
Lung function and nutrition are clearly linked.

The national rate is 86.0 percent (green bar).
The range is 7 to 100 percent.
Physicians and Nurses?
Family Partners
still optional... but recognized for their value
Overall, 44.5 percent of patients get four or more respiratory cultures a year.
Age Specific Prevalence of Respiratory Organisms, 2009

- *P. aeruginosa* 51.7%
- *H. influenza* 16.0%
- *B. cepacia complex* 2.7%
- *S. aureus* 51.3%
- *S. maltophilia* 12.7%
- *MRSA* 23.7%
- *Any Staph* 65.8%
**Common Complications vs. Age**

- **Bone Disease**
- **Diabetes (CFRD)**
- **Depression**
- **Arthritis/Arthropathy**

**Complication** | **Percentage of People with CF (%)**
--- | ---
Gastroesophageal reflux (GERD) | 26.7
Distal Intestinal Obstructive Syndrome (DIOS) | 4.2
Asthma | 22.2
Overall, 74.4 percent of patients had two or more pulmonary function tests in 2009.
Patients 6 to 18 Years With $\geq 4$ Visits, $\geq 1$ Culture, and $\geq 2$ PFTs, by CF Center

The national rate is 72.9 percent of patients who meet all three criteria (green bar).
The range is 7.7 to 100 percent.
The national rate is 71.5 percent (green bar). The range is 0 to 98.6 percent.

The CDC’s Advisory Committee on Immunization Practices recommends influenza vaccination for all patients with CF ≥ 6 months of age.²
The national rate is 74.7 percent (green bar). The range is 14.3 to 100 percent. The CFRD guidelines recommend screening patients \( \geq 14 \) years of age with at least a random blood sugar measurement\(^3\).
The national rate is 78.2 percent (green bar). The range is 0 to 100 percent.
The national median weight-for-length percentile is 47.8 (green bar). The range is 9.5 to 82.9. A center must have five patients < 2 years old to be represented. The goal established by the nutrition guidelines committee is 50th percentile weight-for-length by 2 years of age.
Median CDC Weight Percentile vs. Age by Birth Cohort

Figure 32
The national rate is 84.4 percent (green bar). The range is 23.1 to 100 percent.

A center must have five patients < 2 years old to be represented.
The national rate is 72.2 percent (green bar). The range is 16.7 to 100 percent.

The nutrition guidelines committee recommends the use of nutritional supplements in addition to usual dietary intake for children with growth deficits.

°Gastrostomy, nasogastric, jejunostomy, and parenteral feeding.
The national median length of treatment for a pulmonary exacerbation in patients < 18 years is 15 days (green bar). The range is 5.5 to 26.3 days. A center must have five patients with an exacerbation treated by IV therapy to be included in this graph.
The national rate is 77.5 percent (green bar). The range is 20.5 to 100 percent. Of all patients taking Pulmozyme® at any encounter during the year, 13.7 percent use it twice a day [BID use by disease severity: severe (< 40) is 27.6 percent, moderate (40-69) is 19.2 percent, mild (70-89) is 11.5 percent, and normal (≥ 90) is 7.1 percent].

*At any encounter in the year.
Chronic Inhaled Tobramycin (TOBI®) Use* in *P. aeruginosa*

Positive Patients $\geq 6$ Years, by CF Center

The national rate is 68.7 percent (green bar). The range is 18.2 to 100 percent.

*At any encounter in the year.
The national rate is 67.5 percent (green bar). The range is 9.1 to 100 percent. Patients were considered eligible if they met the selection criteria used in the U.S. Azithromycin trial.15

*At any encounter in the year.
The national rate is 46.2 percent (green bar). The range is 0 to 93.3 percent.

*At any encounter in the year.
Cayston
(Aka inhaled Aztreonam)
New in 2010
Case management

- Where to get the medications
- What equipment is needed
- Where tests can be done and how to order them
- Funding sources
- What is most necessary?...if you have to choose
- Resources for food, travel, copays
- Coordinating tests (annual labs, bone scans, lung functions, x-rays/CT, GI testing, referrals, home care, pharmacy services, audiology)
- Cohorting clinics
- Solving school issues
- Summer activities
- Helping families find a balance
Building blocks of success?

- Blood work
- Vitamins
- zithromax
- Insurance companies
- Family!!
- Nutrition Supplements
- Clinic Visits
- Tuneups
- School
- Copay assistance
- Nurses
- Nutritionist
- Social Worker
- Physicians
- Respiratory Therapists
- Individualized school plan
- Pulmozyme
- TOBI
- Hypertonic Saline
- Airway Clearance
- Exercise
- Enzymes
“You have to be really careful that a chronic illness doesn’t become your whole life”

From the parent of a child with a chronic illness
Median Predicted Survival Age, 1986-2010
Over 5 Year Bands

Year:
- 1986-1990
- 1991-1995
- 1996-2000
- 2001-2005
- 2006-2010

Median Survival Age (Years):
- 24
- 28
- 32
- 36
- 40
Predicted age of survival 38.3 years

47.4% of patients are over 18 years

Range of ages is 1-74 years
Number of CF Patients Who Had a Lung Transplant Each Year, 1990-2010

Year


Number of Transplant Patients

0 40 80 120 160 200
Characteristics of Adult CF Patients 18 Years and Older

**Education**
- 34.1% Some College
- 29.5% College Graduate
- 24.9% High School Diploma
- 7.9% Less Than High School
- 3.6% Masters or Doctoral Level Degree*

*New Field in 2010

**Employment**
- 33.2% Full-time
- 25.3% Student
- 17.4% part-time
- 15.8% Disabled
- 8.5% Unemployed
- 1.0% Retired

**Marital Status**
- 56.0% Single
- 39.2% Married/Living Together
- 4.6% Separated/Divorced
- 0.3% Widowed
Hopes for the future

Simpler drug delivery

A pill to treat the defect
ΔF508 Mutations (%):
- Homozygous ΔF508: 47.8
- Heterozygous ΔF508: 39.4
- No ΔF508 or both unidentified: 12.8

Mutation frequency (24,019 genotyped patients, 48,038 alleles):

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The drug has been shown to be remarkably effective at diminishing symptoms and limiting disease progression. The yearly cost for a prescription of Kalydeco is slated to be $294,000.
Building blocks of success?

- Clinic Visits
- Nurses
- Enzymes
- Nutritionist
- Social Worker
- Physicians
- Nutrition Supplements
- Blood Work
- Vitamins

LIFE!

FAMILY!!!