

## SUBSPECIALIST CONSULT

## Management of the Child With Cystic Fibrosis

All 50 states require newborn screening for cystic fibrosis, and pediatricians play an important role when a newborn is diagnosed. The positive screens from your state laboratory will include referral options for confirmatory testing and specialty care at an accredited Cystic Fibrosis Center or CF center affiliate in your area. Pediatricians remain the patient's "medical home," help to coordinate this specialty care, and remain in close communication with the patient, families, and specialists at a CF center as these children grow.

Optimal outcomes come from assessment and treatment by a multidisciplinary team at an accredited CF center. Encourage your CF patient and families to be seen there regularly. You want those specialists on board to help you take optimal care of the child. Monitoring at least every 3 months is recommended and is called "expectant" or "proactive management" for an individual with CF. A more thorough assessment occurs at least annually.

Pediatricians are essential, in particular, when patients live far from a CF center. We have patients who live a 3-hour drive from our center at the University of Chicago. In some states, patients may live hundreds of miles from a center and can be seen for specialty care only once per year.

There are some specialist outreach efforts as well for children who live in

extremely remote, rural areas: Clinicians in Seattle fly to Anchorage, Alaska, to monitor and treat children with CF.

If a sick child with CF comes as a new patient to your office and the records are not available, you should always feel free

to call the closest CF center to speak to one of the specialists there and eventually refer them for care. Physicians at CF centers work in collaboration with primary care physicians who are the "go to" professionals, often treating other members of a CF child's family.

Consultation and referral are important. Pediatricians tend to be very, very busy and see 20-40 patients a day in their office for a wide range of indi-

cations, including earaches, stomach aches, and well-child exams. But if you are taking care of a sick child with CF who is just not getting better with antibiotic or other treatment, definitely communicate with a CF care provider.

Many pediatricians we know well in the Chicago area will call us up and say, "I have so-and-so in my office. They are coughing a little bit more. I know you sent a letter before saying I should try this particular antibiotic. Is there anything else I should do at this point?" They stay in close touch, and we always have a center physician or nurse available to speak with them.

Regular assessment by a pediatric pulmonologist is part of the CF center care. If a child has an increased cough and they

have certain bacteria such as *Pseudomonas*, we aggressively treat them so they don't get worse. We monitor lung function and obtain frequent respiratory cultures.

In addition, up to 90% of these children and adolescents develop digestive problems and may benefit from consultation with a gastroenterologist. Some patients develop sinus-related problems and referral to an ENT may be warranted. Accredited centers are required to provide these specialists, as well as dieticians, social workers, and respiratory therapists, who are experienced and knowledgeable about CF and who are involved in providing a care plan for children with CF.

Pediatricians play an important role in the facilitation of regular testing of children and adolescents with cystic fibrosis. At a minimum of once a year, they require blood work and chest x-rays. Many patients do their annual visits in the summer because testing takes almost a whole day. The lab work and chest x-ray copy (on a CD) can then be brought to CF physicians on the day of that "annual" visit. Depending on the patient's insurance, it may be less expensive and more convenient for the family for the pediatrician to coordinate this annual testing at the local community hospital. Lung function testing typically needs to be done at the center.

We also now do an oral glucose tolerance test for CF-related diabetes in any patient older than 10 years. That often can be done locally as well. Patients with CF may develop a specific type of diabetes. Most patients with CF have pancreatic in-

sufficiency, which results in problems with digestion and the need to take medication (pancreatic enzymes) with every meal. Over time, scarring of the pancreas results and insufficient insulin may be produced.

The Cystic Fibrosis Foundation, which approves and accredits CF centers, is a great source of information for patients and families. Advise parents to visit [www.cff.org](http://www.cff.org) for reliable and up-to-date information. This site provides locations of all of the accredited CF centers in the United States, information on clinical research studies, and general medical information on CF for families and patients. It is important to educate families to avoid reading too much on CF blogs or online chat or discussion groups. Sometimes the information found there is inaccurate or out of date. The median survival for children with cystic fibrosis was 12 years in 1970, 24 years in 1985, and currently it's 38 years. We have a very large clinic for adults with CF at our center and here, as well as all over the country, there are individuals who are 40, even 60, years of age with CF. We haven't found a cure for this disease, but the management has improved markedly over the years. Much of this improvement has been as a result of coordinated care at CF centers. ■

DR. LESTER is professor of pediatrics and section chief of pediatric pulmonary medicine at the University of Chicago, and director of the Cystic Fibrosis Center there. She said she had no relevant financial disclosures.



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## Many Asthmatic Kids Sidelined by Exercise-Related Symptoms

BY DOUG BRUNK

FROM THE ANNUAL MEETING OF THE AMERICAN ACADEMY OF ALLERGY, ASTHMA, AND IMMUNOLOGY

SAN FRANCISCO – Coughing, shortness of breath, and other exercise-related respiratory symptoms substantially limit the ability of children with asthma to participate in normal physical activities.

In addition, many asthmatic children who experience such symptoms are not using short-acting bronchodilators as recommended in national guidelines.

Those are key findings from a national survey of parents that set out to evaluate the impact of exercise-related respiratory symptoms on physical activities of children with asthma.

"Kids with asthma need to be active to be healthy," Dr. Nancy K. Ostrom said in an interview during a poster session at the meeting. "A lot of them have exercise-induced bronchospasm that needs to be recognized and treated. This is a critical issue for health in the United States right now. If you can't be physically active, you are not going to be able to be fit."

The telephone survey included 516 parents of children or adolescents with current asthma contacted between Dec. 8, 2009, and Jan. 3, 2010, said Dr. Ostrom, codirector of the San Diego-based Allergy and Asthma Medical Group and Research Center. The study by Dr. Ostrom and her colleagues, known as the EIB Landmark Survey, consisted of 84 questions structured to provide data on demographics, symptoms experienced, and impact of exercise-induced bronchospasm (EIB) on daily function.

## VITALS

**Major Finding:** Nearly half of parents of children with asthma (46%) reported that their child experienced four or more exercise-related respiratory symptoms, such as coughing and shortness of breath. Adolescents were more likely than younger children to avoid activities because of exercise-related respiratory symptoms (32% vs. 22%, respectively).

**Data Source:** A national telephone-based survey of 516 parents of children or adolescents with current asthma.

**Disclosures:** The study was sponsored by Teva Respiratory. Dr. Ostrom disclosed that she has served in one or more capacities as a consultant, clinical investigator, or speaker for multiple companies, including Teva, Glaxo-SmithKline, and Merck.

More than three-quarters of parents of children with asthma (79%) reported that their child experienced at least one exercise-related respiratory symptom, and nearly half (46%) reported that their child experienced four or more such symptoms. The three most common symptoms were coughing (62%), shortness of breath (61%), and wheezing (53%).

Adolescents were more likely than younger children to avoid activities because of exercise-related respiratory symptoms (32% vs. 22%, respectively). At the same time, parents of children aged 4-12 years reported that asthma limited their child's ability to participate either "a lot" or "some" in sports/recreation (30%), normal

physical exertion (21%), and other outdoor activities (26%). The corresponding figures for adolescents were 21%, 24%, and 54%, respectively.

"That's a huge number of kids with potential impact," Dr. Ostrom commented. "You tend to learn exercise habits when you're a child, not when you're an adult."

Adolescents were also more likely to be limited "a lot" in sports competition, compared with younger children (13% vs. 7%, respectively).

Use of bronchodilators such as albuterol before exercise was infrequent. Only 23% of children and adolescents with asthma took bronchodilators "always" or "most of the time," 19% took them "sometimes," 15% took them rarely, and 42% never took them (1% was unknown).

In their poster, the researchers stated that adherence to controller therapy and prevention of exercise-related symptoms with short-acting bronchodilators "should be optimized per current treatment guidelines."

One way to achieve that goal, Dr. Ostrom said, is to ask parents and asthmatic children during office visits if they ever experience shortness of breath, coughing, or other respiratory symptoms during or shortly after physical activity. "The important question is, 'Does that keep you from what you want to do or what you should be doing from a health standpoint?'" she said. "Exercise is critical. These health habits need to begin in childhood."

Dr. Ostrom acknowledged certain limitations of the study, including the fact that it surveyed parents, not the children directly. ■