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In December 2009, the Federation of International Respiratory Society (FIRS), with leadership representing the major international respiratory societies, announced that 2010 would be designated The Year of the Lung. The announcement launched a collaboration to implement public awareness campaigns around the globe.

“At that meeting it was announced that all the respiratory societies in the world and the World Health Organization would embrace The Year of the Lung to create awareness about lung airways and sleep disorders,” John Walsh explains. “Much of the focus is on Chronic Obstructive Pulmonary Disease (COPD) because it is the fourth leading cause of death in America and the second leading cause of disability. One individual in the U.S. dies of COPD every four minutes.”

The organizers knew that a global educational initiative such as The Year of the Lung would be a massive undertaking. However, various respiratory organizations in Europe, Latin America and Asia have taken this opportunity to make presentations to legislative bodies in their respective countries to promote awareness of lung disease. Although COPD is the main umbrella, tuberculosis is a focus in less developed countries, and the more rare diseases such as Alpha-1 Antitrypsin Deficiency, pulmonary hypertension, ARDS and others are also receiving exposure, as are lung cancer and asthma.

“Focusing on so many lung diseases over the period of a year is a real challenge,” Mr. Walsh says. “There are some campaigns that are focused on exposures and risk factors for developing symptoms and exacerbating the progression — not only tobacco but also environmental issues.”

The COPD Foundation’s strategy for The Year of the Lung was to select a different topic as their focus for each month. The focus in August was Women’s Lung Health because of the disparity in diagnosis of COPD between men and women. For the first time, more women have died of COPD over the last two years than men.
“If a man is diagnosed with asthma, he’s more likely to get a complete diagnosis of COPD than a woman. The gender disparities in healthcare are accentuated with lung disease,” Mr. Walsh explains.

The September focus for The Year of the Lung was the importance of maintaining influenza vaccines. October 14 was designated World Spirometry Day and November 12 World Pneumonia Day.

“The primary message from both the Alpha-1 Foundation and COPD Foundation during The Year of the Lung is that if a person has a diagnosis of COPD, it’s imperative that he or she gets tested for Alpha-1; and if a person is symptomatic for COPD, they need to see their physician and get a spirometry test,” Mr. Walsh says. “We are encouraging everybody with Alpha-1 to take this opportunity to get out there and raise awareness — talk to Better Breathers clubs and people with COPD across the country and encourage people to be tested.”

In conjunction with The Year of the Lung, the Alpha-1 Foundation has also launched a family testing initiative. “It’s All In The Family” explains the genetic factors involved in Alpha-1 and answers questions people may have about being tested. (Please visit the website for more information: www.alpha-1foundation.org/alphas/?c=06a-lts-all-in-the-family.)

His Personal Journey

John Walsh, his twin brother and two sisters watched their mother suffer with lung disease. At the time, nobody knew about Alpha-1.

“She was a brave woman,” John recalls. “She didn’t really let us know the extent of her symptoms. She worked harder than many people I’ve seen who are less affected than she was. She was remarkable. I think subconsciously there was a reason I never smoked or worked in places with environmental exposures because I saw Mom on oxygen as we were growing up.”

She died at age 46, about the time Alpha-1 was first identified in 1963. John and his brother were 13, and there were few physicians around who knew much about COPD, let alone Alpha-1.

The Walsh brothers were diagnosed at age 35 with allergy-induced, adult-onset asthma — the most common misdiagnosis for people with Alpha-1. It took five years between that initial diagnosis and the full Alpha-1 diagnosis at age 40. The full diagnosis is largely due to the vigilance of John’s sister-in-law in seeking answers as to why her husband was getting so many lung infections, to the extent that he was hospitalized with pneumonia three times in five years. One of their sisters was also eventually diagnosed homozygote (ZZ).

John began his augmentation therapy in 1993, and today his lung capacity is at 45 percent. One of his biggest challenges is that his life’s work involves 250 days of travel a year, and most of that in airplanes. He recognizes the danger that presents to a person with Alpha-1, but his passion about the work won’t allow him to avoid doing what needs to be done. He does take serious precautions — infusing religiously every week, keeping his pneumococcal vaccine and flu shot up to date, washing his hands frequently and carrying anti-bacterial wipes. He also uses oxygen when he travels or exercises, and hooks up to a percussion ventilator at night. He recognizes that maintaining his health isn’t just for him — he’s the front line for thousands of people impacted by COPD and Alpha-1.

“We all have to do our part,” he says simply. “I’ve found that this is what I’m supposed to be doing. I’m honored to be in the position to help the Alpha-1 Foundation and COPD Foundation grow. We are making an impact, and it’s rewarding to see that.”

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Influenza Overview

The flu vaccines approved for the 2010-11 season protect against three strains of influenza, including the 2009 H1N1 pandemic flu strain, which has been incorporated into both the nasal spray and injection vaccines.

You can begin protecting yourself and your family against influenza by getting vaccinated every year. The Centers for Disease Control and Prevention (CDC) expanded its 2010-11 influenza vaccination recommendations beyond high-risk groups. They now recommend all people over six months of age get the annual flu vaccine.

Influenza is a viral infection in the nose, throat and lungs. About 10 to 20 percent of Americans get the flu each year. Of those, about 200,000 people go to a hospital and 36,000 people die because of the flu and complications.

Influenza may cause fever, cough, sore throat, a runny or stuffy nose, headache, muscle aches and tiredness. Most people feel better after one or two weeks; but for some, the flu leads to serious diseases, such as pneumonia.

If you have alpha-1, your lungs are vulnerable to pollutants and infections, while individuals with lung disease such as emphysema are more prone to flu (influenza) and pneumonia. More importantly, the damage that can occur in your lungs from alpha-1 antitrypsin deficiency can be triggered by inflammation from a bacterial or viral infection. However, vaccines can truly help protect your lungs from these conditions.

If you are living with alpha-1, it is recommended that you receive a flu vaccine every year and a pneumococcal vaccine every five to six years. If you have established liver disease, your physician may also recommend vaccinations for hepatitis A and B.
People who may have a higher risk of getting the flu or complications include:

- Children aged 6 months up to 19 years
- Adults ages 50 years and older
- Women who are or will become pregnant during the flu season
- People living in nursing homes or long-term care facilities
- Individuals with chronic health problems
- Healthcare workers who have direct contact with patients
- Caregivers and household contacts of children less than 6 months old

It is best to get the vaccine as soon as it becomes available each fall, but you can also get it any time throughout the flu season. The vaccine is available by shot or by nasal spray, and works by exposing your immune system to the flu virus. Your body will build up antibodies to the virus to protect you from getting the flu. The flu shot contains dead viruses. The nasal-spray vaccine contains live but weakened viruses. However, you cannot get the flu from either vaccine.

You can also reduce your risk of catching the flu by washing your hands frequently, which stops the spread of germs. Eating healthy, exercising and getting enough sleep also play a part in preventing the flu because they help boost your immune system. If you are sick, cough and sneeze into your elbow, and wash your hands often to prevent giving the flu to others.

Some people should not get the flu shot. If you fall into one of these categories, you should talk to your doctor before being vaccinated:

- People who have had an allergic reaction to a flu shot in the past
- People with an allergy to eggs
- People who previously developed Guillain-Barré Syndrome within six weeks of getting a flu shot
- Children younger than 6 months of age
- People who have a moderate or severe illness with a fever should wait until symptoms subside before receiving the flu shot

If you are pregnant during flu season, you cannot get the nasal-spray vaccine. However, it is recommended that women who will be pregnant during flu season get the shot. Pregnancy can increase your risk for complications from the flu.

**Pneumococcal Disease**

Pneumococcal disease is an infection caused by a type of bacterium called *Streptococcus pneumoniae*. It can cause pneumonia, bloodstream infections or meningitis. *Streptococcus pneumoniae* bacterium kills more people in the United States each year than all other vaccine-preventable diseases combined. Antibiotics used to be effective, but the bacterium is becoming more resistant to antibiotic treatment, making immunization increasingly important.

Pneumococcal pneumonia is the most common type of bacterial pneumonia, and it is especially severe in adults over 65 and in people who have chronic medical problems such as alpha-1, asthma, emphysema, COPD, heart disease, diabetes, alcohol abuse, kidney failure, cancers and weakened immune systems from disease or drug treatments.

Pneumococcal meningitis is the leading cause of bacterial meningitis in the United States. Death rates are approximately 20-50 percent in adults. It can also result in permanent damage to the brain and nervous system, learning deficits and deafness.
The pneumococcal vaccine is recommended for all adults 65 or older and people age 2 through 64 with any of the following:

- Chronic cardiovascular disease
- Chronic pulmonary disease
- Cigarette smokers
- Chronic liver disease, including persons with long-term alcoholism
- Diabetes mellitus
- CSF leaks
- Functional or anatomic asplenia
- Immunosuppressive conditions
- Organ or bone marrow transplantation
- Therapy with alkylating agents, antimetabolites or systemic corticosteroids
- Chronic renal failure or nephrotic syndrome
- Cochlear implants

A second dose (revaccination) of the pneumococcal polysaccharide vaccine (PPSV) is recommended for persons age 2 through 64 years with any of the following:

- Functional or anatomic asplenia
- Immunosuppression
- Transplant(s)
- Chronic renal failure
- Nephrotic syndrome
- Adults age 65 years or older who received the first dose prior to age 65

Dosage recommendation for children 2 to 9 years old is every three years; for people age 10 and up, revaccination is recommended every five years.

Hepatitis A and B

The hepatitis A and B vaccine is especially important for those with established liver disease.* Hepatitis is an inflammation of the liver and can have different causes. Parents, caregivers or significant others need to be aware of any indication of possible complications related to liver disease. Symptoms may include:

- Increased abdominal swelling or edema of the extremities
- Coughing up or vomiting bright red blood
- Blackish, purplish or dark-colored stools
- Confusion, irritability, disorientation or lethargy
- Little or no urine or dark (cola or tea color) urine
- No appetite
- Itching or increased itching
- Change in appearance of jaundice

Once liver injury is identified, the first course of action is to evaluate lifestyle habits for potential liver toxins. Substances in some medications may be harmful to your liver, including prescription and over-the-counter medications with acetaminophen, or alcohol and nutritional supplements such as vitamins, herbs and protein drinks.

Summary

It is important to receive a flu shot annually and the pneumococcal vaccine every five to six years. The lungs are vulnerable to pollutants and infections, and the use of these prophylactic vaccinations is vital to alpha-1 patients. You should consult with your doctor about which vaccines are best for you.

*Always consult your physician before receiving any vaccination.
Common Myths Regarding Organ Donation

Reprinted with permission from the United Network for Organ Sharing (UNOS) website.

There is a severe organ shortage in this country. Despite continuing efforts at public education, misconceptions and inaccuracies about donation persist. It’s a tragedy if even one person decides against donation because they don’t know the truth. Below is a list of the most common myths followed by the actual facts.

**Myth:** If emergency room doctors know you are an organ donor, they won’t work as hard to save you.

**Fact:** If you are sick or injured and admitted to the hospital, the number one priority is to save your life. Organ donation can only be considered after brain death has been declared by a physician. Many states have adopted legislation allowing individuals to legally designate their wish to be a donor should brain death occur, although in many states organ procurement organizations also require consent from the donor’s family.

**Myth:** When you’re waiting for a transplant, your financial or celebrity status is as important as your medical status.

**Fact:** When you are on the transplant waiting list for a donor organ, what really counts is the severity of your illness, time spent waiting, blood type and other important medical information.

**Myth:** Having “organ donor” noted on your driver’s license or carrying a donor card is all you have to do to become a donor.

**Fact:** While a signed donor card and a driver’s license with an “organ donor” designation are legal documents, organ and tissue donation is usually discussed with family members prior to the donation. To ensure that your family understands your wishes, it is important that you tell your family about your decision to donate life.

**Myth:** Only hearts, livers and kidneys can be transplanted.

**Fact:** Needed organs include the heart, kidneys, pancreas, lungs, liver and intestines. Tissue that can be donated include the eyes, skin, bone, heart valves and tendons.

**Myth:** Your history of medical illness means your organs or tissues are unfit for donation.

**Fact:** At the time of death, the appropriate medical professionals will review your medical and social histories to determine whether or not you can be a donor. With recent advances in transplantation, many more people than ever before can be donors. It is best to tell your family your wishes and sign up to be an organ and tissue donor on your driver’s license or an official donor document.

**Myth:** You are too old to be a donor.

**Fact:** People of all ages and medical histories should consider themselves potential donors. Your medical condition at the time of death will determine what organs and tissue can be donated.
Myth: If you agree to donate your organs, your family will be charged for the costs.

Fact: There is no cost to the donor’s family or estate for organ and tissue donation. Funeral costs remain the responsibility of the family.

Myth: Organ donation disfigures the body and changes the way it looks in a casket.

Fact: Donated organs are removed surgically, in a routine operation similar to gallbladder or appendix removal. Donation does not change the appearance of the body for the funeral service.

Myth: Your religion prohibits organ donation.

Fact: Most major organized religions approve of organ and tissue donation and consider it an act of charity.

Myth: There is real danger of being heavily drugged, then waking to find you have had one kidney (or both) removed for a black market transplant.

Fact: This tale has been widely circulated over the internet. There is absolutely no evidence of such activity ever occurring in the U.S. While the tale may sound credible, it has no basis in the reality of organ transplantation. Many people who hear the myth probably dismiss it, but it is possible that some believe it and decide against organ donation out of needless fear.

**2010 Alpha-1 Consumer Teleconference Series**

Coram’s teleconference calls are a great way to learn about topics that affect alpha-1 consumers. The calls feature leading experts from the healthcare and patient communities. There is never a charge to participate, and you can call from the comfort of your own home.

**How to Join a Call**

**Call:** Toll-free 866.418.5399
Enter access code 7167731529 when prompted

**Time:** Tuesdays
7:00 p.m. EST / 4:00 p.m. PST

**OCTOBER 26, 2010**
**Topic:** Be the Driver at Your Doctor’s Appointment — How to Make the Most of Your Visits
**Speaker:** Kathy Johnson, RN
Alpha-1 Advocate Supervisor, Coram Specialty Infusion Services

**NOVEMBER 16, 2010**
**Topic:** To Be Announced
**Speaker:** To Be Announced

**DECEMBER 2010**
No call — have a great holiday season!
Susan Cornejo and Malinda Forshage came to Coram at the same time last March as Patient Relations Specialists in the expanded Alpha-1 Breathe Easy program. Both brought with them 20+ years experience working with the public. Susan held a management position in the Department of Social Services with the State of Colorado for 22 years, and Malinda was a self-employed school photographer for 27 years.

“Susan and I enhance each other,” Malinda says. “We didn’t just come in off the street. We’ve both had experiences in our lives that made us a good fit for this job.”

“The function of a Patient Relations Specialist is to build meaningful and lasting relationships with [Coram’s alpha-1] patients,” Susan explains. “We do that by following up with patients on a monthly or quarterly basis to 1) see how their condition is; 2) check on their augmentation therapy; and 3) answer any questions they may have. That can lead to a multitude of things.”

Both women have qualities essential to the position: they are both organized, they are problem solvers and most of all they are empathetic. And that’s where the similarities end. Although warm and caring, Susan is calm, collected and has a more clinical background. Malinda is just as efficient, but she has a big-as-Texas personality and tends to wear her heart on her sleeve. Both approaches work for them and their patients.

Prior to taking early retirement from the State, Susan oversaw the implementation of the Older Americans Act, monitoring the 16 Area Agencies on Aging (the Triple A’s) throughout the state.

“Those agencies are responsible for providing services such as congregate meals, home delivered meals, transportation, the ombudsman program, in-home services and the senior community employment program,” she explains. “In a management position, I monitored the Triple A’s to ensure that those services were being provided statewide.”

Her job also included helping seniors steer through the bureaucratic systems of Medicare, Medicaid, Social Security Disability, COBRA and other programs.

“I had worked for the State government programs, so I had the contacts to help people navigate through that maze. That also benefited me during the four years I spent as a patient advocate at a hospital.”

The quality of calm command helped Susan manage a group of patient advocates during her hospital experience and also benefits her current patients. If they are frustrated about their insurance coverage or their care, she can settle them down in order to identify and rectify the problem.

“The ultimate goal of this job is taking it full circle,” she says. “We identify the issues and get them addressed, and then go back to the patient and tell them what we’re going to do.”

“You have to be genuinely compassionate and understanding. You have to listen to know what the patient’s needs are, and sometimes that’s all they want. They just want somebody to talk to, someone who will listen to them and be understanding, especially if there’s frustration involved.”

Susan COrnejo
Malinda Forshage

Malinda echoes Susan’s commitment to listening, identifying problems patients are experiencing and solving them. The difference is in their styles.

“I want my patients to imagine us sittin’ in comfortable chairs and just talkin’,” she says with a distinct Texas drawl. “I want them to feel like they can tell me anything.

“Our patients can make us laugh — some are really funny — and they can make us cry because we know they’re in the later stages of life. The people who are down, if you can make them laugh a couple of times during the conversation, you’ve given them something to smile about after you hang up.”

Malinda’s people skills stem from her business as a school photographer for 27 years. She went to a different school each day and not only interacted with her photo subjects but also with school principals, superintendents, teachers and parents.

“I had several classes where I took their kindergarten photo and then took their picture as they crossed the stage at graduation.”

When she moved to Colorado to be closer to her son and his family, she sold all her professional equipment.

“I got rid of it all and bought a nice digital camera to take pictures of my grandchildren,” she says with a grin in her voice.

Malinda has spent extensive time since coming to Coram learning about alpha-1 and how it affects patients. One of the things she learned were the stages many people go through when they are first diagnosed — denial and then depression.

“Since they’re not with other alpha-1 patients, they feel like they’re the only one, and some of them are,” she says. “Many patients don’t know that depression is normal, and they keep it to themselves. If I encounter that, I let them know it’s normal and that we have information that can help them. If they’re not in a support group or have people like us calling them, it’s like they’re on an island by themselves.”

That’s one of the key roles for both Susan and Malinda: listening to their patients to identify the true issues. Sometimes a product delivery problem or an Social Security Disability Insurance situation is just that and they work to fix it. Other times, those aren’t the real issues; they are symptoms of deeper concerns. Susan and Malinda have the level of maturity and empathy to dig beneath the surface and find out what the true concern is, and the ability to help the patient work through it.♦
Affordable Care Act Saving Money for Medicare and Taxpayers

August 10, 2010 — Medicare will see savings of nearly $8 billion by the end of 2012 due to the Affordable Care Act, the Centers for Medicare & Medicaid Services (CMS) reported today. In a study examining the impact of the law and actions taken to achieve its goals (available at www.cms.gov), CMS reports that savings to Medicare will rise to more than $575 billion over the next decade.

These reforms include new provisions that will improve the quality of care, develop and promote new models of care delivery, appropriately price services, modernize the health system, and fight waste, fraud, and abuse. The Affordable Care Act is projected to more than double the life of the Medicare Trust Fund, extending its life from 2017 to 2029. Without enactment of the Affordable Care Act, the nation’s already excessive healthcare spending would have reached unsustainable levels within the next few decades.

Source: alpha1.org

Engineered Lung Experiment Successful

June 24, 2010 — A Yale University-led team of scientists reports that it has achieved an important first step in regenerating fully functional lung tissue that can exchange gas, which is the key role of the lungs.

Lung tissue is difficult to regenerate because it does not generally repair or regenerate beyond the microscopic level. The only current way to replace damaged adult lung tissue is to perform lung transplantation, which is highly susceptible to organ rejection and infection, and achieves only 10 percent to 20 percent survival at 10 years.

The Yale team’s goal was to see if it was possible to successfully implant tissue-engineered lungs, cultured in vitro, that could serve the lung’s primary function of exchanging oxygen and carbon dioxide. They took adult rat lungs and first removed their existing cellular components,
preserving the extracellular matrix and hierarchical branching structures of the airways and vascular system to use later as scaffolds for the growth of new lung cells.

Lead author Laura Niklason, MD, PhD, professor and vice-chair of the Departments of Anesthesiology and Biomedical Engineering at Yale University and a member of Yale Medical Group, said, “We succeeded in engineering an implantable lung in our rat model that could efficiently exchange oxygen and carbon dioxide, and could oxygenate hemoglobin in the blood. This is an early step in the regeneration of entire lungs for larger animals and, eventually, for humans.”

Niklason says that for this technology to be applicable to patients, it is likely that years of research with adult stem cells will be needed to repopulate lung matrices and produce fully functional lungs.

Source: Science Express

**WHO Chief Says H1N1 Flu Pandemic is Over**

August 10, 2010 (Reuters) — The H1N1 pandemic is over and the global outbreak turned out to be much less severe than was feared just over a year ago, according to the head of the World Health Organization (WHO).

WHO director-general Margaret Chan rebutted criticism that the United Nations agency had hyped the first pandemic in more than 40 years, whose mildness left some Western governments holding huge stockpiles of unused vaccines.

The Hong Kong public health expert said the world had been lucky the H1N1 virus had not mutated into a more deadly form and that a safe vaccine developed in record time remained effective against it.

“We are now moving into the post-pandemic period. The new H1N1 virus has largely run its course,” Chan said.

The swine flu virus will continue to circulate as part of seasonal influenza for years to come, requiring health authorities to remain vigilant, she told a news conference. It still threatens high-risk groups including pregnant women who would benefit from vaccination.

Stockpiled H1N1 vaccines remain effective against the strain and so far the virus has not developed widespread resistance to the antiviral oseltamivir, the best treatment.

The WHO’s downgrading of the H1N1 outbreak to “post-pandemic” was based on recommendations by external influenza experts who conducted an earlier review.

Source: www.nlm.nih.gov (Written by Stephanie Nebehay)

**The NHC Applauds New Regulations**

August 4, 2010 -- The National Health Council (NHC) released the following statement in support of the White House announcement of new federal regulations designed to strengthen patients’ rights
to appeal health insurance denials. The following statement is from Myrl Weinberg, President, NHC:

“People with chronic conditions make up the largest segment of users of healthcare in this country. They require care and treatment that will allow them to live the healthiest, most productive lives possible. It is essential that they be able to obtain and retain coverage and care that meet their individual health care needs.

The steps taken today by the Obama Administration will provide patients with an understandable, predictable internal and external appeals process. It’s the right thing to do. It’s a fair solution for patients — particularly those living with chronic diseases and disabilities.

“We are also pleased that $30 million has been allocated for strengthening state-based consumer assistance offices. This is a critical step to improving patient awareness of their rights.”

Source: alpha1.org

FDA Warns Consumers, Pharmacists and Wholesalers Not to Use Stolen Advair Diskus® Inhalers

July 16, 2010 — The FDA is warning the public that certain Advair Diskus inhalers stolen from a distribution warehouse in 2009 have been found in some pharmacies. The safety and effectiveness of the stolen inhalers cannot be assured and they should not be used.

Advair Diskus (fluticasone propionate and salmeterol inhalation powder) is an inhaler used to treat patients with asthma and chronic obstructive pulmonary disease.

The products were reported stolen in August 2009 from a GlaxoSmithKline warehouse near Richmond, VA. The inhalers found recently were the first from the stolen lots to be found in commerce. However, more stolen product may still be on the market and the FDA continues to aggressively investigate the matter.

Stolen medicine may be harmful because it may have

Alphas in New York Fire Department Continue to Rapidly Lose Lung Function

July 30, 2010 — Some New York City firefighters who were “first responders” to the World Trade Center destruction on September 11, 2001, continue to have “World Trade Center cough.”

And those with mild or moderate alpha-1 antitrypsin deficiency (alpha-1) continued to lose lung function at a faster rate than normal for at least four years after 9/11, according to a study this month in CHEST, the journal of the American College of Chest Physicians.

David Prezant, MD, Chief Medical Officer of the New York Fire Department, was the principal investigator for the study. Mark Brantly, MD, director of the alpha-1 lab at the University of Florida, is a co-author of the study reported in CHEST.

Source: alpha1foundation.org
been stored at the wrong temperature or humidity or other improper conditions, may degrade or lose potency, become contaminated, or may have been tampered with or handled improperly while outside of the legitimate supply chain.

Source: copdfoundation.org

Foundation Thanks Alphas for Joining “Virtual Lobby Day”

August 4, 2010 — Many Alphas from across the United States joined the first “Virtual Lobby Day” held by the American Association for Respiratory Care (AARC) August 4.

On Virtual Lobby Day, patients and patient advocacy organizations joined respiratory therapists and other professionals to send personal messages to Congress. The focus of the messages was for Congress to support the Medicare Respiratory Therapy Initiative (HR 1077 and S 343).

These bills would provide Medicare reimbursement for respiratory therapists working with physicians, but not necessarily under the physician’s direct supervision.

Source: alpha1foundation.org

Prenatal Smoke Tied to Poorer Asthma-drug Response

August 10, 2010 (Reuters Health) — Studies have shown that children whose mothers smoked during pregnancy may have an increased risk of developing asthma. Now new research suggests they may also get less benefit from the inhaled steroid medications used to prevent asthma attacks.

In a study of more than 1,000 children between 5 and 12 years old with mild-to-moderate asthma, researchers found that those who had been exposed to smoke in the womb had less of a response to the inhaled corticosteroid budesonide (Pulmicort®) than children with no prenatal exposure to smoking.

Overall, both groups of children improved with the medication. However, children with prenatal smoke exposure had 26 percent less of an improvement in their “airway responsiveness.”

Inhaled steroids are the mainstay of therapy for persistent asthma, helping to prevent attacks of coughing, wheezing and breathlessness. One of the ways that physicians measure whether a patient is responding to inhaled steroids is by testing airway responsiveness. In this study, children with prenatal exposure to maternal smoking had less of an improvement in airway responsiveness after starting budesonide — and some had no improvement at all, said Dr. Benjamin A. Raby of Brigham and Women’s Hospital in Boston, one of the researchers on the study.

Lung transplantation may be a last resort for the most severe cases of chronic obstructive pulmonary disease (COPD) and alpha-1 antitrypsin deficiency (alpha-1) lung disease, but it may bring a sigh of relief. Over the past 20 years, lung transplantation has gone from an experimental procedure to an accepted treatment option for many diseases, including alpha-1 and COPD. Approximately 4 percent to 5 percent of patients waiting for a lung transplant, and 7 percent of patients transplanted, have alpha-1 while 27 percent of lung transplant patients were diagnosed with COPD. Today, patients receiving a lung transplant can benefit from an improved quality of life and a higher survival rate due to the fact that approximately 85 percent of all lung transplant recipients survive the first year, nationally, and those who survive the first year are more likely to survive three years or longer. Although it may be the last resort, lung transplantation could be the life-saving operation needed for some with severe COPD or alpha-1 lung disease.
What to know before a transplant:
Currently there are almost 1,800 patients waiting for an available donor lung or lungs, and perhaps half of those waiting will receive a transplant within a year. The decision to pursue a lung transplant follows careful screening by a lung transplant center and a number of transplant specialists. Many factors impact a patient’s opportunity for a transplant and how long they wait, including blood type, body size, tissue type, weight and other conditions. Although each transplant center sets their own criteria, there are certain requirements generally accepted to determine if a patient will be a transplant candidate. The criteria may include:

- A diagnosis of severe end-stage lung disease for which there is no alternative treatment or all other options were exhausted without success
- No other chronic medical conditions (e.g. heart disease, kidney disease, liver disease)
- Progressive disability and/or loss of mobility
- A life expectancy of two years or less
- Understanding of and willingness to accept the risks of surgery as well as willingness and ability to comply with all post-transplant responsibilities

Candidates undergo a pre-transplant screening process to evaluate their overall health and ability to tolerate the surgery. If any risks are identified, strategies may be enforced to minimize these risks before or after a transplant. In addition, patients are prioritized on the transplant wait list according to their lung allocation score — a system put in place to help assure that the patients with the most likely to benefit from a lung transplant have priority. A higher score gives a higher position on the wait list. Since the implementation of the new system, the number of patients dying while on the wait list has dropped significantly.

What to know after a transplant:
After the transplant, the immune system will begin to see the transplanted organ as foreign and attempt to reject the organ. It is therefore essential to follow the prescribed medications (some may be intravenous [IV]) as directed for life — *a lung transplant is a life-long commitment*. Not taking the medications has been directly associated with transplant rejection, lung deterioration and even death. There are many side effects that will likely be experienced with these medications. However, most side effects can be managed and many go away or lessen as medication doses are reduced.

Most transplant recipients will experience a rejection episode as the body attempts to reject the organ. To treat a rejection episode, the doctor will change medications and doses as needed while

Lung transplantation does not cure alpha-1. The liver will continue to produce the same alpha-1 antitrypsin levels, even after a successful operation. It is therefore important for those with alpha-1 to discuss with their physician the value of maintaining augmentation therapy after a successful transplant.
constantly monitoring the patient’s white blood cell count and lung function. A rejection episode can typically be reversed so it is essential to continually watch for and report any signs of rejection. These signs may include:

- A fever
- Flu-like symptoms
- Shortness of breath or difficulties breathing
- Signs of poor lung function
- Chest pains or tenderness around the incision area

Sometimes, in the early stages of a rejection episode, a patient may have no obvious symptoms. Therefore, the doctor may order additional tests (e.g. chest x-ray, CT scan, biopsy) to look for any unseen signs.

Transplant patients are also more susceptible to infection because of the required medications (which reduce the body’s ability to fight infection), exposure risk during surgery and the inability to cough after surgery due to a decreased cough reflex. Patients are closely monitored and any infections are treated aggressively — which may include antibiotics. Patients should be aware of signs of infection and report them right away to their doctor, who will evaluate the need for other treatments to prevent any future complications.

Transplant patients are also educated on their post-transplant responsibilities. Such patient education may include:

- Medicine dosing and compliance
- How to help manage side effects
- Signs and symptoms of rejection
- Signs and symptoms of infection
- Exercise, nutrition and other self-care guidelines
- Short- and long-term healthcare requirements

**What to know about Coram:**

During every stage of a lung transplant, many patients need additional home infusion therapies before and/or after a transplant. For example, nutrition support (both tube feeding and IV) may be prescribed to strengthen a patient before the transplant or promote healing and recovery afterwards. Augmentation therapy (e.g. Aralast® and Zemaira™) for those with alpha-1 lung disease is recommended before a transplant and may continue afterwards to maintain the lung’s protection. Additional post-transplant infusion needs will likely include antibiotic, antiviral and antifungal therapies, immunoglobulin therapy (IVIg or SCIg) and/or pain management.

It is important to ensure your healthcare provider understands the entire lung transplant procedure, and can work with you throughout the transplant process. As a leading provider of home infusion services and having almost 20 years of transplant management experience, Coram can work with you and your doctor throughout the transplant process to support you through a complete and successful lung transplant procedure.

**Where to learn more:**

- www.transplantliving.org
- www.unos.org
- www.ustransplant.org

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As one of the longest-serving home care providers to the alpha-1 community, Coram understands the multitude of questions, concerns and challenges that patients face every day. To provide optimum long-term assistance for alpha-1 patients, Coram is pleased to introduce its new alpha-1 support program called Breathe Easy.

The Breathe Easy program is an expansion of Coram’s current alpha-1 advocacy program. It is designed to offer the individualized resources, information and advocacy that patients need to achieve success with their augmentation therapy. Through the Breathe Easy program, patients have the advantage of a coordinated team that will assist them in making vital lifestyle adjustments, providing answers to disease and therapy questions, locating community resources, addressing financial and insurance coverage issues and even having their infusion and/or oxygen therapy managed while they travel.

With Coram, you can breathe easy knowing we are committed to you and your care. For any questions regarding the Breathe Easy program, please contact Coram’s alpha-1 hotline at 866.FOR.A1Pi (866.367.2174).

Customer Service
Ongoing contact from a customer relations specialist to answer questions and address therapy or service related issues.

Peer Mentoring Program
On-demand counseling and educational support by a specially trained alpha-1 advocate.

Alpha-1 Resource Network
Customized alpha-1 information, tools and support items sent to your home.

Insurance and Financial Support
• Referrals to non-profits for premium assistance
• Referrals to non-profits for state high-risk insurance pools
• COBRA counseling and assistance
• Enrollment in manufacturer assistance programs

The Alpha-1 Advocate Quarterly Newsletter
The newsletter is provided free of charge to the alpha-1 community.

Alpha-1 Consumer Teleconference Series
A monthly, toll-free alpha-1 community conference call series for consumers and their caregivers and provided free of charge.

Alpha1Source.com
A website devoted exclusively to providing helpful information and resources to alpha-1 consumers and healthcare providers.
Whom to Contact for Help

If you have any questions or need assistance, please feel free to contact our alpha-1 advocates.

Barbee Bennington
Midwestern and Western States
Alpha-1 Consumer Advocate
Toll-free 800.355.5180 x 50
Email barbee.bennington@coramhc.com

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