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Improving Influenza Vaccine Compliance Through Patient Education for Patients With Cystic Fibrosis

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Cystic fibrosis (CF) is an autosomal recessive disorder that affects about 30,000 children and young adults in the United States. More than 1000 new cases are diagnosed each year, with only 10% of patients with newly diagnosed disease being 18 years of age or older. A sweat test performed at a CF center is the standard method for diagnosis; however, genetic testing confirms the diagnosis. The median age of survival for patients with CF has risen to 36.8 years, which is an increase of 5 years in the last 4 years (Beall, 2006).

Complications from chronic lung disease are the leading cause of mortality in the CF population. Standardization of care and additional CF therapies are factors in the improvement of the length and quality of life for patients with CF. According to the Cystic Fibrosis Foundation *Clinical Practice Guidelines* (1997), all patients with CF older than 6 months of age should receive the influenza vaccine annually. Thus, encouraging flu vaccinations through patient education and monitoring of vaccination status is an important way to prevent deterioration of lung function in patients with CF.

PATHOPHYSIOLOGY OF CF

Patients who have CF inherit a mutation causing the abnormal expression of the protein CF transmembrane regulator (CFTR). The mutations can be homozygous or heterozygous. The most common mutation is delta 508. CFTR is involved in the transport of electrolytes across the cell membrane. CFTR acts as a chloride channel present on the surface of many types of exocrine tissue, including those lining the airways, bile ducts,

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pancreas, sweat ducts, and vas deferens (Rowe, Miller, & Sorscher, 2005).

Lung damage is progressive and causes more than 95% of deaths from CF. The typical features of CF lung disease are mucus plugging, chronic inflammation, and infection. Mucus plugging in CF results from both the increased production of mucus and altered physicochemical properties of the mucus. There are an increased number and size of mucus-secreting airway cells coupled with alterations in CF mucus, which is dehydrated and viscous because of abnormal chloride secretion and sodium absorption (Behrman, Kliegman, & Jenson, 2003).

Inflammation is another typical feature of CF. Abnormal cytokine profiles, such as deficient interleukin-10, promote chronic inflammation and contribute to long-term lung damage. The airways of individuals with CF also have increased numbers of neutrophils. The byproducts of neutrophil metabolism cause direct damage to structural proteins, such as elastin, as well as inducing airway cells to produce interleukin-8, which attracts more neutrophils and worsens the vicious cycle of inflammation and the destruction of respiratory tissue elasticity. Neutrophils also cleave immunoglobulin G and other immune components important for the phagocytosis of pathogens and directly stimulate more mucus secretion (McCance & Huether, 2002).

Pathogens commonly causing lung infections in patients with CF include *Staphylococcus aureus, Pseudomonas aeruginosa, Burkholderia cepacia, Mycobacterium avium-intracellulare,* and occasionally nontypeable *Haemophilus influenzae, Moraxella catarrhalis, Xanthomonas maltophilia, Alcaligenes xylosoxidans, Enterobacteriaceae*, the fungus *Aspergillus* (Steele, 2006), and *Stenotrophomonas maltophilia* (Lang et al., 2004).

Children and young adults with CF are at an increased risk of pulmonary complications if they contract influenza. They have an increased incidence of endobronchial infections for reasons that are poorly understood, because there is no systemic immune defect (McCance & Huether, 2002). The most likely cause is the CF airway environment, which favors bacterial colonization within viscous airway secretions because of the individual's inability to remove secretions adequately from the airways (Rowe et al., 2005). Conversely, infection with the influenza A virus may lead to patients' lungs becoming chronically colonized with *P. aeruginosa* bacteria (Bhalla, Tan, & Smyth, 2006). Infections with viruses, including the influenza virus, lead to 20% of episodes of increased bronchial infections in patients with CF.

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INFLUENZA

Influenza viruses spread from person to person via respiratory-droplets. The incubation period is typically 1 to 4 days, with an average of 2 days. For the majority of the population, symptoms last 3 to 7 days, although a cough and symptoms of fatigue may last as long as a week. Influenza is potentially serious in the very young because of a lack of immunity, in the elderly because of decreased resistance to infection and a poorly operating immune system, and in those with underlying medical conditions such as CF, which render them at greater risk for infections (Harper, Fukada, Uyeki, Cox, & Bridges, 2005). The primary way to reduce the incidence of influenza in the United States is to vaccinate those people most at risk, their household contacts, and health workers with either inactivated or live attenuated influenza vaccine. Ideally, this is done by administering the vaccine to people at their place of work, during hospitalizations, or during routine health care office visits before the start of the flu season. The Advisory Committee of Immunization Practices recommends strong efforts to increase vaccination rates, including reminder/recall systems and standing order programs (Harper et al.).

INFLUENZA VACCINE

The 2005-2006 trivalent vaccine virus strains were A/California/72004(H3N2)-like, A/New-Caledonia/20/99(H1N1)-like, and B/Shanghai/361/2202-like antigens. Available influenza vaccine gave approximately 70% to 80% protection and reduced the likelihood of severe infection. Most vaccinated children had high titers that were protective against the strains in the vaccine, but the antibody response among high-risk children was usually somewhat lower. Vaccines are repeated each year because the principal surface antigen hemagglutinin of influenza viruses A and B undergoes changes by point mutation (antigenic drift) or by exchange of whole DNA segments encoding for this protein (antigenic shift) (Bhalla et al., 2006).

There are a few adverse reactions to and contraindications for the influenza vaccine reported by the National Vaccine Information