

PFTs, dyspnea scores, and perceptions of well-being and has reduced the viscosity of sputum.

Nebulized hypertonic saline (7%) has been shown to be effective in improving airway hydration and increases mucus clearance in patients with CF; this treatment, however, causes bronchospasm and may not be recommended for patients with severe disease (Furnari, Termini, Traverso, et al, 2012). The use of hypertonic saline with hyaluronic acid can decrease the incidence of bronchoconstriction, cough, and throat irritation (Furnari, Termini, Traverso, et al, 2012).

Physical exercise is an important adjunct to daily ACT. Exercise stimulates mucus excretion and provides a sense of well-being and increased self-esteem. Any aerobic exercise that the patient enjoys should be encouraged. The ultimate aim of exercise is to increase lung vital capacity, remove secretions, increase pulmonary blood flow, and maintain healthy lung tissue for effective ventilation.

Pulmonary infections are treated as soon as they are recognized. In CF patients, characteristic signs of pulmonary infection—fever, tachypnea, and chest pain—may be absent. Therefore, a careful history and physical examination are essential. The presence of anorexia, weight loss, and decreased activity alerts the practitioner to pulmonary infection and the need for an antibiotic regimen. Aerosolized antibiotics (such as tobramycin, aztreonam, and colistin) are beneficial for patients with frequent pulmonary exacerbations and are administered in 2- to 4-week cycles or on an ongoing basis to prevent colonization with *P. aeruginosa*.

IV antibiotics may be administered at home as an alternative to hospitalization. The use of peripherally inserted central catheters (PICCs) for the administration of antibiotics in children with CF is a viable option with limited complications and fewer needle punctures to obtain blood specimens and to maintain often lengthy treatment with parenteral antibiotics. Alternatively, an implanted vascular access device offers the advantage of access for blood draws and antibiotic infusion. When pulmonary function does not improve with outpatient management, hospitalization may be recommended for continued antibiotic therapy and vigorous ACT. Periodic hospitalizations for preventive IV antibiotic therapy and percussion and postural drainage occur less frequently than in the past due to limited evidence to support this practice and concern