airways are destroyed, causing bronchiectasis.

The most common pathogens responsible for pulmonary infections are *Pseudomonas aeruginosa, Burkholderia cepacia, S. aureus, H. influenzae, Escherichia coli,* and *Kiebsiella pneumoniae*. *P. aeruginosa* and *B. cepacia* are particularly pathogenic for children with CF, and infections with these organisms are difficult to eradicate. In addition, children with CF who are chronically colonized with these organisms have poorer survival rates than children who are not colonized. Colonization and infection with methicillin-resistant *Staphylococcus aureus* (MRSA) has emerged as a critical factor in lung infection and pulmonary function in patients with CF (Muhlebach, Miller, LaVange, et al, 2011). Patients with MRSA require multiple antibiotic regimens. Fungal colonization with *Candida* or *Aspergillus* organisms in the respiratory tract is also common in CF patients.

Airway clearance therapies (ACTs) are an essential part of CF management and include percussion and postural drainage, positive expiratory pressure (PEP), active-cycle-of-breathing technique, autogenic drainage, oscillatory PEP, high-frequency chest compressions (HFCCs), and exercise. Studies have demonstrated that no particular ACT has any advantage over the other in relation to outcomes of sputum production; however, it is recommended that individualized assessment occur to determine the best ACT for each patient.

ACTs such as percussion and postural drainage are usually performed on average twice daily (on rising and in the evening) and more frequently if needed, especially during pulmonary infection. Percussion and postural drainage is especially useful for infants and young children. Patients with CF have been found to regress when conventional percussion and postural drainage is discontinued.

The **Flutter mucus clearance device** is a small handheld plastic pipe with a stainless-steel ball on the inside that facilitates removal of mucus (Fig. 21-11). It has the advantage of increasing sputum expectoration and being used without an assistant. Handheld percussors may be used to loosen secretions. Another method to clear mucus is HFCC in which the child temporarily wears a mechanical vest device that provides high-frequency chest wall oscillation.