mean of 18 mEq/L. A chloride concentration greater than 60 mEq/L in a child 6 months old or older is diagnostic of CF, a concentration between 40 and 59 mmol/L is indeterminate and a repeat test should be performed in 1 to 2 months (Nicholson, 2013). In some situations, DNA testing may be substituted for the sweat test and may be performed when the sweat test indicates the possible presence of CF. The presence of a mutation known to cause CF on each CFTR gene predicts with a high degree of certainty that the individual has CF; however, multiple CFTR mutations may also be present and detected with DNA assay.

Chest radiography reveals characteristic patchy atelectasis and obstructive emphysema. PFTs are sensitive indexes of lung function, providing evidence of obstructive airway disease. Other diagnostic tools that may aid in diagnosis include stool fat or enzyme analysis. Stool analysis requires a 72-hour sample with accurate recording of food intake during that time. Radiographs, including a contrast enema, are used for diagnosis of meconium ileus.

Therapeutic Management

Improved survival among patients with CF during the past two decades is attributable largely to antibiotic therapy and improved nutritional and respiratory management. Goals of CF therapy are to (1) prevent or minimize pulmonary complications, (2) prevent chronic pseudomonas infection, (3) ensure adequate nutrition for growth, (4) encourage appropriate physical activity, and (5) promote a reasonable quality of life for the child and the family. A multidisciplinary approach to treatment is needed to accomplish these goals.

Management of Pulmonary Problems

Management of pulmonary problems is directed toward prevention and treatment of pulmonary infection by improving ventilation, removing mucopurulent secretions, and administering antimicrobial agents. Many children develop respiratory symptoms by 3 years old. The large amounts and viscosity of respiratory secretions in children with CF contribute to the likelihood of respiratory tract infections. Recurrent pulmonary infections in children with CF result in greater damage to the airways; small