

organs; complications from surgery; pulmonary infections; and recurrence of obstructive bronchiolitis, which decreases transplanted lung function.

There is increasing focus on the use of CFTR pharmacotherapy to act as correctors and potentiators to override the CFTR defect and maintain adequate airway surface liquid layer, as well as to correct abnormal chloride and sodium channels to reduce mucus production. Ataluren (PTC124) and an agent labeled VX-809 are currently being examined ([Panesar, 2011](#)). These pharmacotherapeutic approaches have been shown to offer clinical benefits for persons with delta F508 mutation ([Cuthbert, 2011](#); [Kim Chiaw, Eckford, and Bear, 2011](#)). With advances in technology, parents and adolescents are challenged to set future goals that may include college, careers, social relationships, and marriage. Concurrently, they are faced with increasing morbidity and higher rates of CF complications as they grow older.

Nursing Care Management

Assessment of the child with CF involves comprehensive assessment of all affected systems with special focus on the pulmonary and gastrointestinal systems. Pulmonary assessment is the same as that described for asthma, with special attention to lung sounds, observation of cough, and evidence of decreased activity or fatigue. Gastrointestinal assessment primarily involves observing the frequency and nature of the stools and abdominal distention. The nurse should also be alert to evidence of growth failure (e.g., weight loss, muscle wasting, pallor, anorexia, decreased activity [from baseline norm]). Family members are interviewed to determine the child's eating and eliminating habits and to confirm a history of frequent respiratory tract infections or bowel obstruction in infancy.

The nurse assesses the newborn for feeding and stooling patterns, which may indicate a potential problem, such as meconium ileus. The nurse also participates in diagnostic testing, such as the initial newborn screening, DNA analysis, or sweat chloride test.

The uncertainty, fear, and initial shock associated with the diagnosis are overwhelming to parents. They must face the impact of the chronic, life-threatening nature of the disease and the prospect of intensive treatment, for which they must assume a