stimulation. Prostheses are replaced to accommodate the child's growth and increasing capabilities.

### Nursing Care Management

Prosthetic application, training and use are most successfully carried out in a center that specializes in meeting the special needs of these children, especially very young children and those with multiple amputations or missing limbs. Management involves a **prosthetist**, who specializes in the development, fitting, and maintenance of prosthetic limbs, and other health care providers, such as physical and occupational therapists. Parents need support and are encouraged to assist the child in making age-appropriate adjustments to the environment. Although these children need assistance, overprotection may produce overdependence, with later maladjustment to school and other situations.

## Osteogenesis Imperfecta

OI is a rare genetic disorder characterized by bones that fracture easily. Although inheritance follows an autosomal dominant pattern in most cases, rare autosomal recessive inheritance exists. Most types of OI have defects in the *COL1A1* or *COL1A2* genes, which code for polypeptide chains in type 1 procollagen, a precursor of type 1 collagen, which is a major structural component of bone. The error results in faulty bone mineralization, abnormal bone architecture, and increased susceptibility to fracture. There are at least 12 described types of OI, which accounts for significant disease variability. Clinical features may include varying degrees of bone fragility and deformity, short stature, blue sclerae, hearing loss, and dentinogenesis imperfecta (hypoplastic discolored teeth) (Marini and Blissett, 2013).

Classification is based on clinical features and patterns of inheritance (Box 29-6). Clinically, type I is the most common and mildest form with most fractures occurring before puberty. Stature is near normal and bone deformity is minimal or absent. Type II is the most severe and considered lethal in infancy. Type III OI is characterized by multiple fractures often present at birth, short stature, severe bone deformity and disability with a shortened life expectancy. Type IV is similar to type I although slightly more