Type IV

A: Mild to moderate bone fragility; normal sclerae; normal teeth; short stature; variable deformity; autosomal dominant inheritance

B: Same as A except dentinogenesis imperfecta instead of normal teeth; approximately 6% of cases of OI

Type V: Clinically similar to type IV; hyperplastic callus; collagen mutation negative

Type VI: Sclerae and dentition normal; moderate to severe bone fragility; diagnosis by bone biopsy because of similarities to other types

Types VII and VIII (recessive form): Clinically overlap types II and III but have white sclerae, rhizomelia, and small to normal head circumference; severe osteochondroplastica and short stature in survivors. Type VII is associated with *CRTAP* gene, and type VIII is associated with the LEPRE1 genetic mutation.

OI, Osteogenesis imperfecta.

*Two thirds of cases are type I.

†This classification is based on that proposed by Sillence DO, Senn A, Danks DM: Genetic heterogeneity in osteogenesis imperfecta, *J Med Genet* 16(2):101–116, 1979, which originally included OI types I to IV. Additional types have been described but are not included herein.

Therapeutic Management

The treatment for OI has historically been primarily supportive; although patients and families are optimistic about new research advances. The use of bisphosphonate therapy with IV pamidronate to promote increased bone density and prevent fractures has become standard therapy for many children with OI. However,