Juvenile Osteoporosis

Osteoporosis literally means "porous bone." This disease is characterized by too little bone formation, excessive bone loss, or a combination of both. People with osteoporosis have an increased risk of fractures. It is most common in older people, especially older women.

Osteoporosis is rare in children and adolescents. When it does occur, it is usually caused by an underlying medical disorder or by medications used to treat the disorder. This is called secondary osteoporosis. Sometimes, however, there is no identifiable cause of osteoporosis in a child. This is known as idiopathic osteoporosis.

No matter what causes it, juvenile osteoporosis can be a significant problem because it occurs during the child's prime bone-building years. From birth through young adulthood, children steadily accumulate bone mass, which peaks sometime before age 30. The greater their peak bone mass, the lower their risk for osteoporosis later in life. After people reach their mid thirties, bone mass typically begins to decline—very slowly at first but increasing in their fifties and sixties. Both heredity and lifestyle choices—especially the amount of calcium in the diet and the level of physical activity—influence the development of peak bone mass and the rate at which bone is lost later in life.

Secondary Osteoporosis

Secondary osteoporosis, which can affect both adults and children, results from another primary disorder or therapy. Some examples are included in the box on the next page.

As the primary condition, juvenile idiopathic arthritis (also known as juvenile rheumatoid arthritis), provides a good illustration of the possible causes of secondary osteoporosis. In some cases, the disease process itself can cause osteoporosis. For example, some studies have found that children with juvenile idiopathic arthritis have bone mass that is lower than expected, especially near the joints affected by arthritis. In other cases, medication used to treat the primary disorder may reduce bone mass. For example, drugs such as prednisone, used to treat severe cases of juvenile idiopathic arthritis, negatively affect bone mass. Finally, some behaviors associated with the primary disorder may lead to bone loss or reduction in bone formation. For example, a child with juvenile idiopathic arthritis may avoid physical activity, which is necessary for building and maintaining bone mass, because it may aggravate his or her condition or cause pain.
For children with secondary osteoporosis, the best course of action is to identify and treat the underlying disorder. In the case of medication-induced juvenile osteoporosis, it is best to treat the primary disorder with the lowest effective dose of the osteoporosis-inducing medication. If an alternative medication is available and effective, the child's doctor may consider prescribing it. Like all children, those with secondary osteoporosis also need a diet rich in calcium and vitamin D and as much physical activity as possible given the limitations of the primary disorder.

**Idiopathic Juvenile Osteoporosis**

Idiopathic juvenile osteoporosis (IJO) is a primary condition with no known cause. It is diagnosed after the doctor has excluded other causes of juvenile osteoporosis, including primary diseases or medical therapies known to cause bone loss.

This rare form of osteoporosis typically occurs just before the onset of puberty in previously healthy children. The average age at onset is 7 years, with a range of 1 to 13 years. The good news is that most children experience complete recovery of bone.

**Clinical features.** The first sign of IJO is usually pain in the lower back, hips, and feet, often accompanied by difficulty walking. Knee and ankle pain and fractures of the lower extremities also may occur. Physical malformations include abnormal curvature of the upper spine (kyphosis), loss of height, a sunken chest, or a limp. These physical malformations are sometimes reversible after IJO has run its course.

X rays of children with IJO often show low bone density, fractures of weight-bearing bones, and collapsed or misshapen vertebrae. However, conventional x rays may not be able to detect osteoporosis until significant bone mass already has been lost. Newer methods such as dual energy x-ray absorptiometry (DXA), dual photon absorptiometry (DPA), and quantitative computed tomography (CAT scans) allow for earlier and more accurate diagnosis of low bone mass. These noninvasive, painless tests are a bit like x rays.

**Treatment.** There is no established medical or surgical therapy for juvenile osteoporosis. In some cases, no treatment may be needed because the condition usually goes away spontaneously. However, early diagnosis of juvenile osteoporosis is important so that steps can be taken to protect the child's spine and other bones from fracture until remission occurs. These steps may include physical therapy, using crutches, avoiding unsafe weight-bearing activities, and other supportive care. A well-balanced diet rich in calcium and vitamin D is also important. In severe, long-lasting cases of juvenile osteoporosis, some medications called bisphosphonates, approved by the Food and Drug Administration for the treatment of osteoporosis in adults, have been given to children experimentally.
**Prognosis.** Most children with IJO experience a complete recovery of bone tissue. Although growth may be somewhat impaired during the acute phase of the disorder, normal growth resumes—and catch-up growth often occurs—afterward. Unfortunately, in some cases, IJO can result in permanent disability such as curvature of the upper spine (kyphoscoliosis) or collapse of the rib cage.

**Distinguishing Juvenile Osteoporosis from Osteogenesis Imperfecta**

Osteogenesis imperfecta (OI) is a rare genetic disorder that, like juvenile osteoporosis, is characterized by bones that break easily, often from little or no apparent cause. However, OI is caused by a problem with the quantity or quality of bone collagen resulting from a genetic defect.

Because most children with OI never attain normal bone mass, they suffer from secondary osteoporosis as well. There are several distinct forms of OI, representing extreme variations in severity. For example, a person with OI may have as few as 10 or as many as several hundred fractures in a lifetime. Although the number of people affected with OI in the United States is unknown, the best estimate suggests a minimum of 20,000 and possibly as many as 50,000. The clinical features of OI and their severity vary greatly from person to person. Many individuals with OI have some, but not all, of the clinical features. Children with milder OI, in particular, may have few obvious clinical symptoms. The most common features of OI include:

- bones that fracture easily
- ligament laxity (hypermobile joints) and low muscle strength
- family history of OI (present in about 65 percent of cases)
- small stature in moderate and severe types
- sclera ("whites" of the eyes) tinted blue, purple, or gray in about 50 percent of cases

- possible hearing loss in late childhood or early adulthood
- possible brittle teeth (known as dentinogenesis imperfecta).

The features that most often distinguish OI from juvenile osteoporosis are the **family history** of the disease and the **blue, purple, or gray sclera** commonly found in patients with OI. Distinguishing between OI and IJO may require genetic testing or, in some cases, bone biopsy.