Juvenile Osteoporosis

Osteoporosis literally means “porous bone." It is a disease characterized by too little bone formation or excessive bone loss and an increase in the risk of fractures. Osteoporosis is called a silent disease because it progresses without any symptoms until a fracture occurs. It usually affects people later in life, and is most common in women after menopause.

Osteoporosis is rare among children and adolescents. When it occurs, it is usually caused by an underlying medical disorder or by medications used to treat such disorders. This is called secondary osteoporosis. It may also be the result of a genetic disorder such as *osteogenesis* ***imperfecta.*** Sometimes there is no identifiable cause of juvenile osteoporosis. This is ***known*** as ***idiopathic juvenile osteoporosis (IJO).***

No matter what causes it, juvenile osteoporosis can be a significant problem, as it occurs during the prime bone-building years. From birth through young adulthood, up to about 30 years of age, bone formation predominates, resulting in a steady accumulation of bone mass. Most bone mass, in fact, is accumulated by early adulthood (Matkovic, 1994; Teegarden, 1995). After the mid-thirties, bone mass typically begins to decline slowly, speeding up in women after menopause. Both genetic and lifestyle factors (e.g., diet and physical activity) influence the development of peak bone mass and the rate at which bone is lost.

Secondary Osteoporosis

Secondary osteoporosis can affect both adults and children, and results from an underlying (primary) disorder or therapeutic activity. Juvenile arthritis (JA) 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 JA have bone mass that is lower than expected, especially near the arthritic joints. Sometimes, ***medication*** used to treat the primary disorder may reduce bone mass. For example, certain drugs such as prednisone (glucocorticoids) used to treat JA may affect bone mass. Finally, some ***behaviors*** associated with the primary disorder may lead to bone loss or a reduction in bone formation. For example, a child with JA may avoid physical activity (which is necessary for building and maintaining bone mass) because it may aggravate his or her condition or cause pain.

In cases of 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 also consider prescribing it.

Disorders, Medications and Behaviors That May Affect Bone Mass:

Primary Disorders Medications Juvenile arthritis Anti-convulsants (e.g., for epilepsy) Diabetes mellitus Corticosteroids (e.g., for rheumatoid arthritis, asthma) Osteogenesis imperfecta Immunosuppressive agents (e.g., for cancer) Hyperthyroidism Hyperparathyroidism **Behaviors** Cushing’s Syndrome Prolonged inactivity or immobility Malabsorption syndromes Inadequate nutrition (especially calcium, vitamin D) Anorexia nervosa Excessive exercise leading to amenorrhea Kidney disease Smoking Alcohol

abuse

Idiopathic Juvenile Osteoporosis

Idiopathic juvenile osteoporosis (IJO) is diagnosed after excluding other causes of juvenile osteoporosis (i.e., primary diseases or medical therapies known to cause bone loss, as discussed above). IJO was first identified in the medical literature in 1965 (Dent and Friedman). Since then, fewer than 100 cases have been reported.

This rare form of osteoporosis typically occurs in previously healthy children just before the onset of puberty. The average age of onset is between 8 and 14 years, but it may also occur in younger children during periods of rapid growth. The most notable feature of IJO is that it can remit within two to four years.

Clinical features. The first sign of IJO is usually pain in the lower back, hips, and feet, often accompanied by difficulty walking. There may also be knee and ankle pain, and fractures of the lower extremities. Physical deformities may be present, such as kyphosis (abnormal curvature of the thoracic spine), loss of height, a sunken-chest, or a limp. These physical abnormalities are sometimes reversible after the IJO has run its course.

X-rays of children with IJO often show low bone density, fractures of the weight-bearing bones, and collapsed or misshapen vertebrae. However, conventional X-rays may not be able to detect osteoporosis until significant bone mass has already 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.

Treatment. Early diagnosis of IJO is important, although there is no established medical or surgical therapy for the disease. In fact, there may be no need for treatment, as IJO usually resolves spontaneously. The basic strategy of treatment is to protect the spine and other bones from fracture until remission occurs. This is accomplished through supportive care, which may include physical therapy, use of crutches, and/or avoidance of unsafe weight-bearing activities. Some medications that are used to treat osteoporosis in adults have also been given children with IJO. Examples include bisphosphonates and calcitriol. The physician may try a medical therapy if the problem is severe and not resolving spontaneously.

Prognosis. As mentioned above, patients with IJO can experience a complete recovery within two to four years. Growth may be somewhat impaired during the acute phase of the disorder, but normal growth resumes--and catch-up growth often occurs--thereafter. In some cases, IJO can result in permanent disability such as kyphoscoliosis or even collapse of the rib cage.

Distinguishing IJO from Osteogenesis Imperfecta

Osteogenesis imperfecta (OI) is a genetic disorder characterized by bones that break easily, often from little or no apparent cause. Most forms of OI are caused by imperfectly formed bone collagen, the result of a genetic defect. There are at least four distinct forms of the disorder, representing extreme variation in severity from one person to another. For example, a person may have as few as ten or as many as several hundred fractures in a lifetime. While the prevalence of 01 in the United States is not known, the best estimate suggests that about 20,000 people are affected by this disorder.

The clinical features of OI vary greatly from person to person; there is also great variation in their severity. The most common features of OI include:

- bones that fracture easily
- family history usually present
- small stature common
- blue sclera ("whites" of the eyes) common
- possible hearing loss
- possible dental problems

The features that most often distinguish OI and IJO are the family *history* and *blue sclera commonly* found in cases of OI. There are also radiographic differences: patients with OI often have Wormian bones (irregular bone patterns in the skull).

The Bottom Line

★ Secondary osteoporosis is best addressed by treating the primary disorder and/or using the lowest effective dose of an osteoporosis-inducing medication.

★ Idiopathic juvenile osteoporosis is quite rare. It is often suspected after a series of fractures not caused by serious trauma. The condition usually resolves itself within two to four years, and permanent disability is uncommon.

★ Juvenile osteoporosis can be most easily distinguished from osteogenesis imperfecta by the lack of family history and the absence of blue sclera.

For more information about weight-bearing exercise and other ways to prevent osteoporosis, contact the Osteoporosis and Related Bone Diseases-National Resource Center,

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