What is osteoporosis?
Osteoporosis occurs when the struts which make up the mesh-like structure within bones become thin causing them to become fragile and break easily, often following a minor bump or fall. These broken bones are often referred to as ‘fragility fractures’. The terms ‘fracture’ and ‘broken bone’ mean the same thing. Although fractures can occur in different parts of the body, the wrists, hips and spine are most commonly affected. It is these broken bones or fractures which can lead to the pain associated with osteoporosis. Spinal fractures can also cause loss of height and curvature of the spine.

What is Osteogenesis imperfecta?
Osteogenesis imperfecta (OI) is an inherited condition. It is caused by abnormalities in the genes controlling the production of collagen, the fibrous framework of bone, which is important for its strength. 85% of cases have changes in the structure of their ‘Type I’ collagen, which is the major protein in bone. This leads to bones which are structurally weaker than normal and an increased likelihood of broken bones (fractures). It is estimated that 1 in 15,000 men and women in the UK have OI.

Abnormalities in other areas of the body containing collagen lead to additional problems in some people with OI, such as lax joints, fragile teeth, blue or grey sclera (whites of eyes) and bruising. Some people with OI are short in stature and some develop deafness, particularly in the teenage years or their twenties. OI can vary in severity from mild, in which the person may not be correctly diagnosed and children may simply be thought to be accident prone, through to severe, in which babies have multiple fractures even before birth. In both sexes and in all types of OI, the fracture rate tends to diminish in the teenage years. It can rise again in women after the menopause when bone density falls due to low levels of the hormone oestrogen, and also in men in later life. Bone density is the quantity of bone that, when measured, helps to indicate bone strength.

How is OI treated?
Bisphosphonate drugs are now regarded as an important component in the management of moderate to severe forms of OI. They are often given intravenously to affected children and in some cases oral bisphosphonates are used. All of the bisphosphonates work by slowing down the activity of bone cells that naturally break down bone. Studies have shown improvements in bone density, bone pain and in some studies a reduction in fracture frequency. Further research is needed though to assess the benefits of treating OI sufferers with this group of drugs particularly in children.

Drug treatment is only one aspect of the management of people with OI and it is important this is provided by a multidisciplinary team that includes physiotherapists and occupational therapists. Fractures need to be treated but the immobilisation period should be kept to a minimum as activity allows muscles and bones to stay as strong as possible. It is important also that affected individuals have a well-balanced diet with adequate calcium. Additional factors which can help to maintain healthier bones include avoiding smoking and keeping alcohol consumption within the recommended limits. For affected individuals who are considering having children, genetic counselling can help assess the risk of OI in future pregnancies.

Is OI the same as osteoporosis?
No. Osteogenesis Imperfecta is a form of brittle bones which is genetic in origin and is present from birth. People with OI have abnormal bones because the structure of the collagen in their bones is different. They may though have a lower than normal bone density as part of the problem. This is different from osteoporosis, however, where the bone that is present is normal but there is less of it than there should be. Both conditions are characterized though by an increased risk of fracture. OI can first come to light during or just after pregnancy and be confused with osteoporosis in pregnancy. In cases of mild OI it may also be confused with...
idiopathic juvenile osteoporosis (IJO), which is osteoporosis of no known cause in children. (For more information see our leaflets Pregnancy and osteoporosis and Osteoporosis in children).

Useful contacts

The Brittle Bone Society
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Email: bbs@brittlebone.org
www.brittlebone.org

The National Osteoporosis Society is the only UK-wide charity dedicated to improving the prevention, diagnosis and treatment of osteoporosis and fragility fractures. The Charity receives no Government funding and relies on the generosity of individuals to carry out its vital work.

For osteoporosis information and support contact our Helpline:

0808 800 0035
nurses@nos.org.uk

To become a member or make a donation:

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