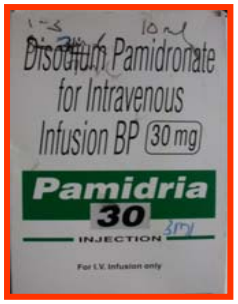
Care of Patients with Osteogenesis Imperfecta

There is no medical treatment for OI. However, judicious use of intravenous and oral bisphosphonates have found to reduce the incidence of fractures.

Cyclical Pamidronate and Zolendronic are given every four months to maintain the bone strength. Dietary supplements like calcium and Vit D3 are also given to supplement nutrition. Exercises and physical activity are useful to maintain muscle strength.



In summary, early correction of deformity in the lower limbs using rods to align the bone with cyclical bisphosphonate treatment and exercises gives us the best chance to prevent long term disability.

In certain types of OI, where the disease is very severe, only supportive treatment may be possible.

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Comprehensive Management of Osteogenesis Imperfecta

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Osteogenesis Imperfecta

OI or Fragile Bone Disease is a leading cause of pathological fracture in infants and children. The fracture occurs with triavial fall or injury.

Some children have typical features of OI: like hyperlaxity of tissues, hyper-mobile joints, blue sclerae, dental problems and deformities of limbs.

OI is caused by mutation in Type I collagen gene and four main clinical types have been described based on the location of mutation.

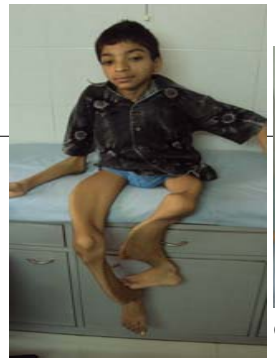
Dentinogenesis Imperfecta is a feature of some types.

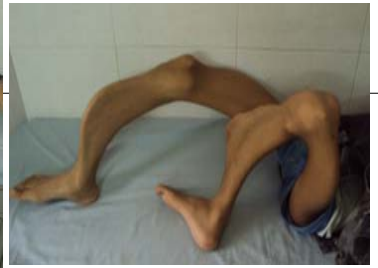
Some children present with multiple fractures and deformities. The fractures may occur with simple or trivial force. Radiographs show a ground-glass medullary cavity and thin cortices. There is associated osteopenia.

Routine haematological markers are normal and urinary markers of increased bone turnover like n-telopeptides and hyroxyproline are increased.

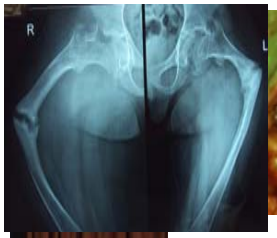
The multiple fractures cause bending of the main wieght-bearing bones such as tibia and femur and once deformity develops the vulnerability to fractures increase further. This leads to a vicious cycle of repeated fractures and progressive bending of the bones rendering the child completely disabled.

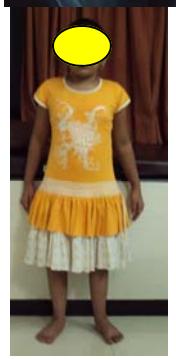
The walking potential of a child is inversely related to the amount of bone deformity. Lack of ambulation, and physical inactivity causes weakening of the muscles as well.

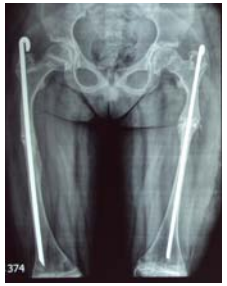




Child with severe OI. He has not received any treatment.



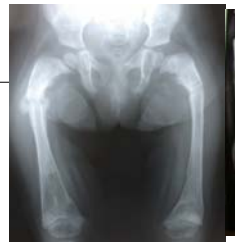


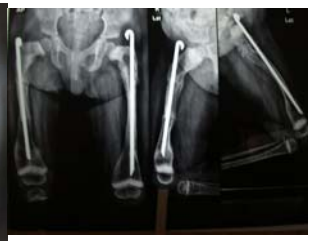


Typical femur deformity in a nine year old girl with OI.

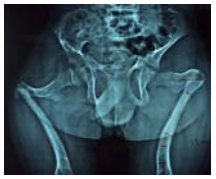
Post - femur rodding using Rush rods. The bone and limb alignment is restored which helps the child to walk.

There are two different types of rod available: non-elongating (n-e) rods and elongating rods aka telescoping rods. The n-e rods such as Rush Rod are used for children with canal diameter < 5mm or if the bones don't allow insertion of telescoping rods.





Two year old child with OI and bilateral femur rodding using Rush Rods.



10 year old boy with femur canal diameter of only 3mm. Rush rods of 2mm size used in both femur.

The incidence of fractures decreases once the child reaches skeletal maturity. Often multiple surgeries are required in childhood to maintain correct bone and limb alignment.

Telescoping Rod (Fassier—Duval) in both tibia and femur in a 9 year old boy. These rods need special equipment and expertise.

F-D rods lengthen with growth, decreasing the need for repeated rod exchanges.