

**SNS COLLEGE OF PHYSIOTHERAPY
COIMBATORE-35**

COURSE: BPT

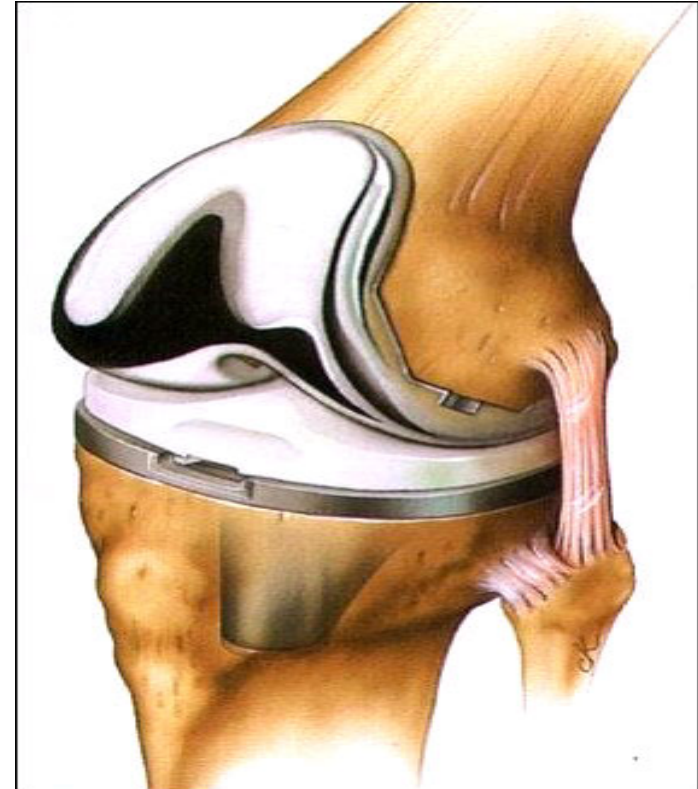
SUBJECT: CLINICAL ORTHOPAEDICS

TOPIC: OSTEOGENESIS IMPERFECTA

UNIT: I

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OSTEOGENESIS IMPERFECTA



DEFINITION

- It is a hereditary condition characterized by deafness, blue sclera, laxity of joints with age.

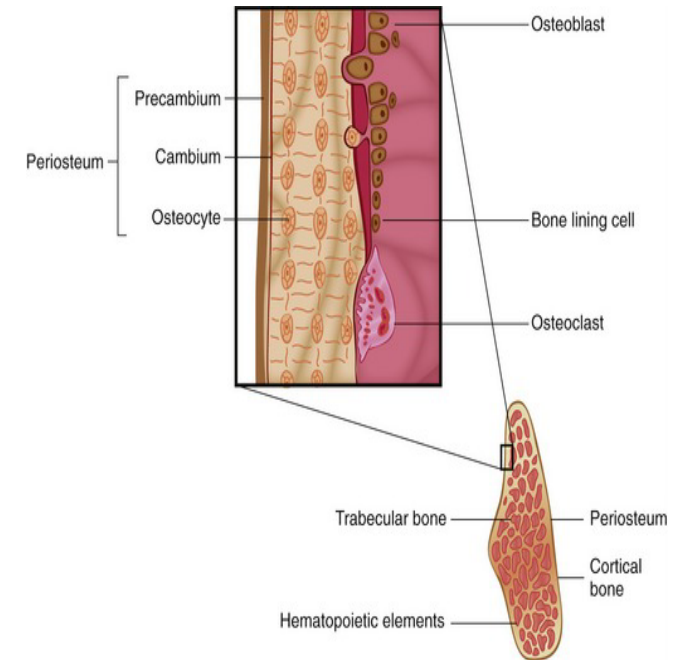


Etiology

- The etiological factors could be
- heredity
- Mendelian recessive—in prenatal cases
- Mendelian dominant—in postnatal cases

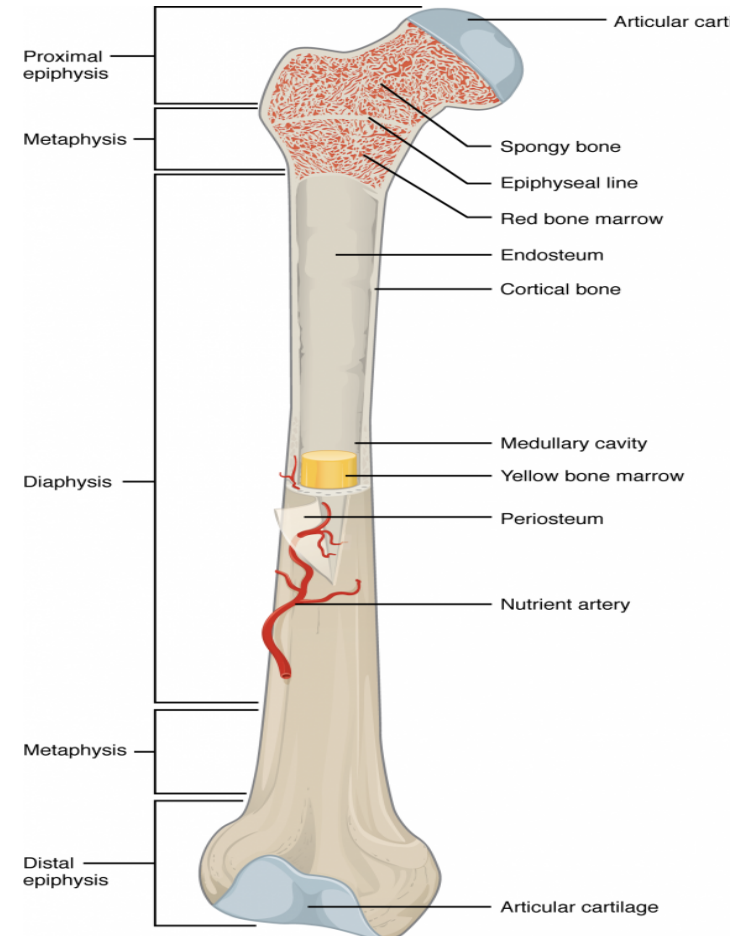
PATHOLOGY

- Primary defect is failure of osteoblast formation during enchondral ossification.
- Features of bones are:
 - Periosteum is thick but the cambium layer is thin
 - Bone is short and thin and the epiphysis is bulbous.



PATHOLOGY

- Cortex is thin and medullary contents are fatty and fibrous
- Bones break easily but heal well with abundant callus.
- Fracture is usually subperiosteal and heals by periosteal bone formation.



CLINICAL FEATURES

- The patient presents with blue sclera, dentinogenesis imperfecta and generalized osteoporosis .
- Blue sclera is seen only in 92 percent of cases, while the other two features are seen in almost all cases.



CLINICAL FEATURES



CLINICAL FEATURES CONTD..

- Dentinogenesis imperfecta is a genetic disorder that affects tooth development, causing discolored, translucent, and weaker teeth, making them prone to wear, breakage, and loss
- Osteoporosis gives rise to bowing and multiple fractures.
- Fractures are usually due to trivial trauma but surprisingly heal well.

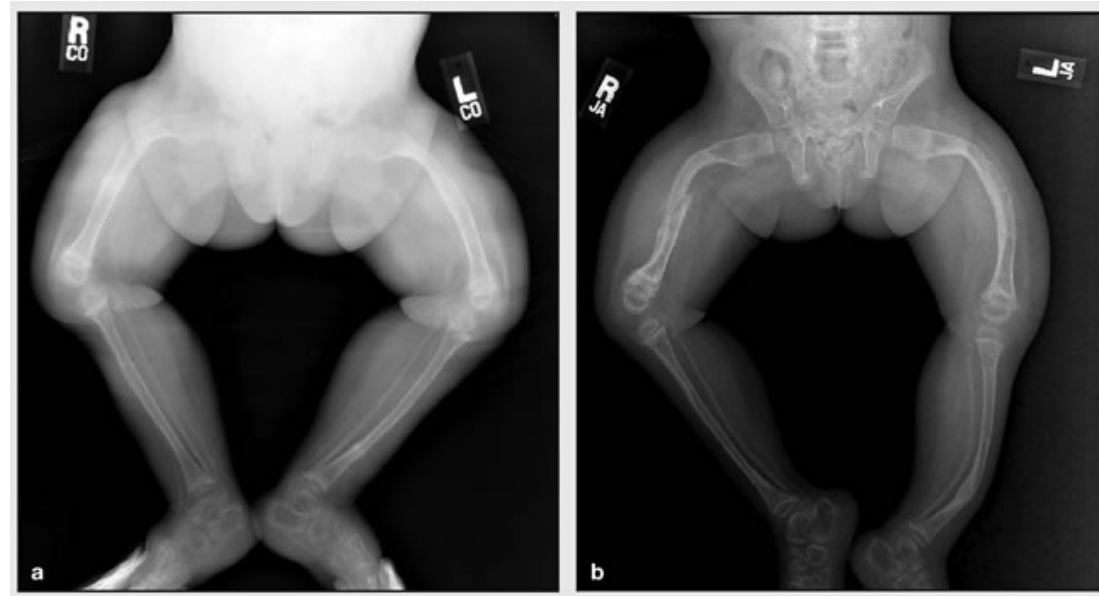
INVESTIGATIONS

Laboratory tests-no specific laboratory test.

Prenatal determination -- amniocentesis and estimation of inorganic pyrophosphate.

X-ray of the affected limbs.

INVESTIGATIONS CONTD..

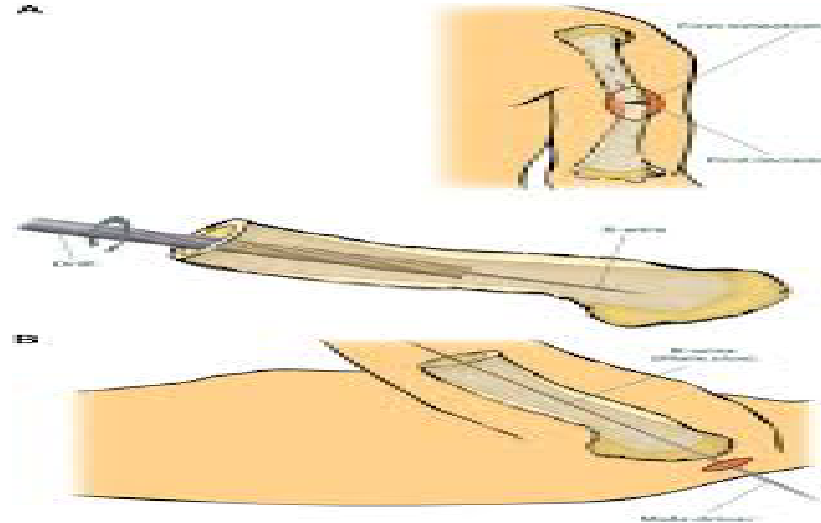


TREATMENT

- Principles
- Protect the child until the tendency of the fracture lessens as age advances.
- Administer vitamins, estrogens and androgens.
- Operate in infantile type as the tendency to fracture is much higher and hence the treatment of choice is multiple osteotomies with intramedullary nailing.

Bailey and Dubow's methodu

- Telescopic medullary rod is used which elongates as growth occurs.



William's retrograde nailing

- It is done by fixing an extension to the distal end of the rod and driving the nail through the heel.



THANK YOU...