Osteogenesis imperfecta (OI)
Characterized by abnormal bone development with low bone mass and increased bone fragility

- **Dominant mutations** affect synthesis & structure of type 1 pro-collagen
- **Recessive mutations** affect post-translational processing or tracking of type 1 pro-collagen

Association with **dentinogenesis imperfecta type 1** with COL1A1 and COL1A2, SERPINH1, FKBP10 mutations

Primary and permanent dentitions both involved in DI type I
Primary dentition more severely affected
- Tooth discoloration
- Reduced enamel thickness
- Vertical enamel fracture
- Associated short roots and bulbous crowns

Bisphosphonates
Primary action as osteoclast inhibitors used for over 20 years in osteogenesis imperfecta
- Increase bone hardness
- Reduce fracture risk

To date
- No reports of any effect of bisphosphonates on tooth development in OI
- No reports of any adverse effects of bisphosphonates on incidence of osteonecrosis of jaw (ONJ) in children or adolescents

Patients and methods
3 Caucasian children with severe forms of OI (Sillence classification type 3, more recently classified as Progressively Deforming OI with normal sclerae)

COL1A1/2 mutations
All presented with peri-natal multiple long bone fractures

Treatment
Zoledronic acid 0.04mg/kg/dose
- All received intravenous zoledronic acid infusions
  - 4 monthly from the first week of life
  - Infusions administered to at least age 6-8 years
  - Increasing intervals between treatment cycles based on improving bone mineral density

Dental examination

Primary dentition
Abnormal teeth
- Brown discolouration of all teeth increased wear
- Loss of dental height over 2-3 years
- Increased rate of tooth loss

Secondary dentition
Almost white teeth normal colour and appearance no signs of reduced enamel or dentine

Results

Mechanism of action of bisphosphonate on tooth formation?
Possibly due to altered lamellation of collagen ?? within teeth exposed to bisphosphonate

Conclusions:
Very early treatment of dentinogenesis imperfecta associated with osteogenesis imperfecta can result in improved dental health and stronger teeth