

## Facts about Osteogenesis Imperfecta

### Overview of Bisphosphonate Use in Children Living with OI

Bisphosphonate therapy, such as intravenous pamidronate or zoledronic acid, has become a standard treatment to increase bone density in children with moderate and severe OI. In most of the studies of people living with osteogenesis imperfecta, bisphosphonates led to a beneficial increase in bone density (measured by DEXA scan). Determining when to start, the most effective dose, and how long to treat are issues that continue to be studied.

There are a growing number of families who find it more convenient to have the drug administered at home by a nurse with direct contact with the treating physician. This is not applicable for the first infusion. Logistical details vary by clinic and physician, and must be discussed on a case-by-case basis. To receive a copy of commonly used protocols for your clinic staff or to discuss with your child's care team, please email [Bonelink@oif.org](mailto:Bonelink@oif.org). Below are a series of frequently asked questions and answers from the OI Foundation's Medical Advisory Council (MAC) about bisphosphonate therapy in children living with OI.

**What is a bisphosphonate?** Bisphosphonates are a group of medicines that are used to slow down normal bone breakdown, allowing bone mass to increase. There are several versions of bisphosphonate compounds, which vary based by the mode of administration and potency. The commonly used versions of bisphosphonate therapy in OI can be broken down into two categories:

- Intravenous (IV) Administration
  - Pamidronate (Aredia®)
  - Zoledronic acid (Zometa®/Reclast®)
- Oral Administration\*
  - Aledronate (Fosamax®)
  - Risedronate with calcium (Actonel®/Atelvia®)

*\*Oral bisphosphonates are typically not used with children as they are less effective, and compliance more difficult to maintain.*

**What is the difference between intravenous pamidronate treatment and intravenous zoledronic acid treatment?** The first input of bisphosphonate use was with IV pamidronate treatment, which requires a three-day infusion every 3-4 months. Zoledronic acid is given intravenously like pamidronate but only requires one 30-60 minute infusion every 6 months. Zoledronic acid has slightly different characteristics including its potency. For convenience, zoledronic acid treatment should be considered due to a smaller requirement of volume, less reaction at first infusion, and less frequent and briefer periods of infusion.

**How long might a child with OI need to stay on bisphosphonate treatment?** DEXA bone scans and blood tests will help plan the dose and duration of treatment. While some individuals taking bisphosphonates on various research protocols are reported to have reached average bone density, it is not possible to predict how any particular individual will respond to the drug. When some of these individuals were taken off bisphosphonates, their bone density gradually decreased over several years. However, it is also important to note that increased bone density does not necessarily translate into increased bone strength. Furthermore, bone density is just one aspect that is modified by the drug- there is also pain relief.

**Should treatment be discontinued for an osteotomy or a fracture?** Scheduled bisphosphonate treatments should not be adjusted in the case of a fracture, but they should around an osteotomy. The drug can be safely administered up to two days before surgery because medication has then cleared from the bloodstream. Reactivation of treatment should be timed with the healing of the osteotomy site- usually 4-5 months- but it may vary due to bone turnover activity.

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*This fact sheet was prepared with assistance from Francis Glorieux, OC, MD, PhD, Chair of the OI Foundation's Medical Advisory Council, 6/1/2018.*

Decision on continuation of treatment or changing to a lower dose regimen is the responsibility of the treating physician and should be made on a case-by-case basis. It is recommended to discontinue treatment when growth is completed.

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