



Pamidronate infusions for Osteogenesis Imperfecta (OI): information for families

Pamidronate is a type of bisphosphonate, a medicine that prevents loss of bone mass. This information sheet from Great Ormond Street Hospital (GOSH) explains about pamidronate infusions for osteogenesis imperfecta (OI), how it works and what to expect when your child comes to GOSH for the infusion.

Bisphosphonates have been used for a variety of conditions in childhood where there is bone pain and immobility in association with osteoporosis and/or abnormal bone formation.

Pamidronate strengthens the bone by increasing bone density and corrects the imbalance between bone reabsorption and bone formation which occurs in osteogenesis imperfecta (OI).

Pamidronate infusions are most effective when they are given every six weeks in small children and three-monthly in older children. This may be reduced during or after puberty as the bone density may improve. We currently recommend continuing pamidronate treatment throughout childhood until growth is completed. Stopping treatment may potentially increase risk of fracture.

Are there any alternatives?

Bisphosphonate treatment strengthens bone reducing risk of fractures. In older adolescents there is a bisphosphonate tablet available called risidronate, which may be suggested instead of the pamidronate infusions. However this will be discussed with your consultant before starting.

What happens before the infusion?

You will already have received information about how to prepare your child for the infusion in your admission letter.

The person bringing your child to the test should have 'Parental Responsibility' for them. Parental Responsibility refers to the individual who has legal rights, responsibilities, duties, power and authority to make decisions for a child. If the person bringing your child does not have Parental Responsibility, we may have to cancel the test.

When you arrive on Kingfisher Ward in the morning (usually at 10am), a nurse will check your child and take some baseline observations of their temperature, pulse and breathing. These will be used to monitor your child during the infusion.

They will also take a blood sample for testing and insert a cannula (thin, plastic tube) into a vein, usually on the back of the hand or inside the elbow, so that the infusion can be given easily. A nurse will apply some local anaesthetic cream first so that their skin is numb. If your child is over five years old, they may like to have a cold numbing spray before the needle instead of the cream.

If your child has a central venous access device, such as a central venous catheter or implantable

port, this will be used to take the blood sample and infuse the pamidronate instead of a cannula.

In the afternoon, once the results of the blood test results are back and checked by a doctor, a nurse will start the pamidronate infusion.

Important

Vitamin D is important if your child is having treatment with Bisphosphonate drugs. Before these medications can be started, it is important we know their vitamin D level is above 50nmol/l. Blood samples will usually be taken either after your clinic appointment or if your child has a central venous access device, when this is flushed – usually on monthly basis.

If your child's vitamin D level is too low (under 50nmol/l), we will ask your family doctor (GP) to prescribe some higher strength vitamin D that is only available on prescription.

What does the infusion involve?

The pamidronate is usually in a plastic bottle with a tube connected to your child's cannula (or central venous access device). It will be infused gradually over four to six hours every 24 hours, giving three doses in total. The last infusion will usually finish in the evening of the third day of your child's admission. When the last infusion has finished, the nurses will check that your child is well enough to go home and remove the cannula or port needle.

Are there any risks or side effects?

Each person reacts differently to medicines, so your child will not necessarily experience every side effect mentioned. If you are concerned about any of these side effects, please ask the clinical nurse specialist, or doctor in clinic.

Side effects of pamidronate commonly include:

- **Flu-like symptoms including a high temperature, aches/pains and vomiting** – This affects the majority of children for 24 to 48 hours on day two of their first set of infusions. The nurses will give your child medicine to deal with symptoms as they occur. If they are severe, the third infusion may be delayed. This side effect does not usually happen with following infusions.
- **Low calcium levels** – Calcium levels tend to drop during the infusions and for three to five afterwards. This usually causes no symptoms but if it occurs, we will give your child calcium supplements in the form of dispersible or chewable tablets for a few days. Low calcium levels are usually diagnosed through blood tests, however in very rare circumstances, it can cause your child to have a small rise in blood pressure and shakiness in their hands.

If this happens while your child is at GOSH, tell your nurse. If it happens when you have gone home, please take your child to your nearest Accident and Emergency (A&E) department.

- **Delayed bone healing** – This is a potential risk after orthopaedic surgery. If your child requires surgery, we will usually reschedule their pamidronate infusion so that there is a gap of at least two weeks before and after surgery or until there is evidence of bone healing confirmed on an x-ray (usually taken around six weeks later).

What happens afterwards?

We will inform you in clinic when your child's next follow up appointment with us will be. When your child has had their infusion on Kingfisher ward, we will aim to give you their next admission date before you are discharged.

Dose regimens maybe modified depending on monitoring investigations and results of annual DEXA scans if your child weighs more than 10kg.

When you get home, call your children's community nursing team if you have any concerns about your child's central venous catheter or implantable port, or your family doctor (GP) if you have any general concerns.

If your child has a fracture or you suspect one, please attend your local Accident and Emergency (A&E) department and ask the nursing staff or

medical team to call the OI team to let us know or if they require any advice.

After the first two cycles of pamidronate, we will usually ask your local team to provide the infusions providing your child is tolerating the infusions well. Infusions given locally can be done as a day case admission, with no need to stay overnight.

We are unable to give the infusions at GOSH on a long term basis due to capacity. We work closely with local hospitals who can contact the GOSH team if they have any queries about the infusions.

If you have any questions, please contact the Clinical Nurse Specialist for Osteogenesis Imperfecta on 020 7405 9200 ext 5824 or call Kingfisher Ward on 020 7813 8221.