



Great Ormond Street Hospital for Children NHS Foundation Trust

# Zoledronic acid infusions for osteogenesis imperfecta (OI): information for families

**Zoledronic acid is a type of bisphosphonate, a medicine that prevents loss of bone mass. This information sheet from Great Ormond Street Hospital (GOSH) explains about zoledronic acid infusions for osteogenesis imperfecta (OI), how they work and what to expect when your child comes to GOSH for an 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.

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

Zoledronic acid infusions are most effective in children when they are given every six months. This may be reduced during or after puberty as bone density may improve. We currently recommend continuing zoledronic acid treatment throughout childhood until growth is completed.

Stopping treatment may potentially increase risk of fracture. As of September 2017, we are only using zoledronic acid in children over five years old.

## Are there any alternatives?

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

In children less than five years old pamidronate is used – see our information sheet for further details.

## What happens before the infusion?

Your family doctor (GP) or paediatrician will be asked to prescribe some calcium supplements for your child to take for one week before the infusion and one week afterwards. It is important that your child takes this as zoledronic acid is stronger than pamidronate so will lower the calcium levels quicker.

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 infusion.

When you arrive on Kingfisher Ward around 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 to record as a baseline to compare with the next morning. A doctor will listen to your child's heart and lungs to ensure they are fit to have the infusion.

The nurses will insert a cannula (thin, plastic tube) into a vein, usually on the back of the hand or inside of 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 zoledronic acid instead of inserting a cannula.

In the afternoon, once the results of the blood test results are back and checked, a nurse will start the zoledronic acid 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?

Zoledronic acid usually comes in a plastic bottle (burette) with a tube connected to your child's cannula (or central venous access device). It will be infused gradually over 45 minutes. The infusion

will usually finish in the afternoon of your child's admission.

For children who have had pamidronate infusions previously, the first dose will be a standard dose for their age given every six months, three months after the last dose of pamidronate. If your child has not had pamidronate before, the first dose will be a half dose followed by a full standard one six months later.

All children will have the first and second dose as an overnight admission. The following morning, the nurses will check that your child is well enough to go home and remove the cannula or de-access 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 your doctor, nurse or pharmacist. Side effects of zoledronic acid 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 the day after their first infusion. The nurses will give your child medicine to deal with symptoms as they occur. If they are severe, the second infusion may also be a half dose. This side effect does not usually happen, or is less severe with following infusions.
- **Low calcium levels** – Calcium levels tend to drop following the first infusion. This usually causes no symptoms but, if it occurs, we will give your child double calcium supplements in the form of dispersible 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 or tingling in their hands. If this happens at

home, after an infusion at GOSH, you should take your child to be seen at your local hospital, who can contact the OI team for advice.

- **Delayed bone healing** – This is a potential risk after orthopaedic surgery. If your child requires surgery, we will usually reschedule their zoledronic acid 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.
- **Tracking line** – During the infusion, in rare cases, children and young people can have red tracking line appear where the zoledronic acid is being given. This does not usually require any treatment. However, the cannula may need to be replaced into another vein.

### What happens afterwards?

We will inform you in clinic when your child's next follow up appointment with us will be. When you get home, call your children's community nursing

team if you have any concerns about your child's central venous access device 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. For orthopaedic management and advice your A&E can contact our on call orthopaedic team through switchboard and ask for the orthopaedic Registrar or Consultant on call.

After the first two cycles of zoledronic acid, we will 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.**