



Dental Care for People with Osteogenesis Imperfecta

Osteogenesis Imperfecta (OI) is always associated with bone fragility. In addition, OI may affect the growth of the jaws and may or may not affect the teeth. About half of the people who have OI have teeth that appear normal, and their major concerns are routine care. However, the other half has a defect in the teeth called Dentinogenesis Imperfecta (DI), sometimes referred to as opalescent teeth or brittle teeth. These teeth may be discoloured, may chip or break easily, and will require special care.

About half of the people who have OI have a defect in the teeth called Dentinogenesis Imperfecta (DI), sometimes referred to as brittle teeth.

Oral cavity problems related to OI may include the following:

- A skeletal Class III malocclusion. The teeth do not correctly match up making biting difficult. This is caused by the size and/or position of the upper jaw or the lower jaw.
- An open bite. There is a vertical gap between some of the upper and lower teeth.
- Impacted teeth. The first or second permanent molars do not erupt, or they erupt out of the usual location (ectopic).
- Dental development. Tooth development may be delayed or advanced in some individuals affected by OI.
- Hypodontia
- MIH
- Caries



OI does not affect the presence or absence of gum disease (periodontitis).

Major Parts of the Teeth

The teeth are made up of four distinct parts.

- Enamel is the outside part of the crown. It is the hardest substance in the body and the point of contact for chewing.
- Dentine is the substance under the enamel that forms the rest of the crown and surrounding the pulp chamber and almost all of the root structure. It is similar to bone.
- The Pulp Chamber is the inner hollow part of the tooth containing blood vessels and nerves.
- The Dentinoenamel Junction (DEJ) is the term for where the enamel and dentine are attached to each other.

Dentinogenesis Imperfecta (DI)

Dentinogenesis Imperfecta can be part of Osteogenesis Imperfecta (OI type I) or it can be a separate inherited dominant trait without OI (OI type II). DI occurring with OI seems to run in families but can vary in severity from one member to another. DI has a variable effect on the colour, shape, and wear of both primary (baby) and permanent teeth, although the primary teeth are usually more affected than the permanent teeth.

DI may be diagnosed with the first baby tooth. If the tooth looks grey, bluish, or brown, DI should be suspected. Children should be taken to a dentist (if possible a specialist in paediatric dentistry) when the first teeth are erupting. This may happen as early as 6 months to 1 year of age. Radiographs, or X-rays, can be useful but may be difficult to obtain until the child is older. Sometimes there are changes visible on the X-rays that are not obvious just by looking at the teeth. Crowns appear bulbous and roots may be shorter and more slender than standard. Primary teeth are usually more affected than the permanent teeth.



General Care for People with OI and Without DI

A dentist should see a child with OI after the eruption of the first baby tooth, and by their first birthday at the latest. Baby teeth require care. They are important for chewing, speaking, holding space for the permanent teeth to grow in, and growth of the jaws. There appears to be minimal risk of jaw fracture from routine dental care and dental extractions. No particular precautions are needed other than those that would be taken anyway, such as support of a very thin lower jaw when an extraction procedure is being done.

Good oral care involves:

- Brushing as soon as the first tooth appears (usually at about 6 months of age), at least twice a day with fluoride toothpaste last thing at night and on at least one other occasion.
- Brushing at bedtime is important as it makes sure that the fluoride continues to protect the teeth while sleeping.
- Parents/carers should brush or help their child to brush their teeth until they are at least seven years old to make sure the teeth are cleaned properly, to supervise the amount of toothpaste used and to prevent licking or eating the toothpaste.
- For the maximum prevention of tooth decay for children aged 0-6 years use toothpastes containing 1350-1500 parts per million (ppm) fluoride.
- For children under three years old use a smear of toothpaste containing no less than 1000 ppm fluoride (see Figure 1).
- Children between three and six years old should use a pea-sized amount of toothpaste containing more than 1000 ppm fluoride (see Figure 2).
- Encourage your child to spit out the toothpaste after brushing and do not let them rinse out with water as this will wash away the fluoride and reduces how well it works- spit don't rinse.
- Sealants placed on the biting surface of the permanent molars in children may reduce the chance of developing cavities.

Figure 1

smear for 0-3
year olds



Figure 2

pea-sized blob for 3-6
year olds



General Care for People with OI Plus DI

Children with OI and DI need the same basic care as discussed above, but they also need to be monitored for cracking, chipping and wear of the teeth. Special care will be needed even with the baby teeth. Restorative treatment may be needed at some point, and includes preformed (metal) crowns or caps for the molar teeth. These crowns can be placed without drilling or local anaesthetic and are well tolerated by even young children (Figure 3). When, for any reason, crowns are not feasible, a “tooth colour” dental material may be used, such as composites or resin-modified glass ionomers. In any case, amalgam restorations should not be used because they impose an additional stress on the teeth.

Figure 3



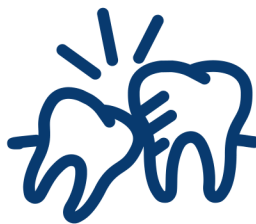
Regular care is needed so the teeth will last as long as possible and to prevent abscesses and pain. Brushing and cleaning has not been shown to cause damage, but will not make teeth affected by DI white. Sealants should be effective on teeth affected with DI as long as the enamel is intact. Older children and especially adolescents with DI are often embarrassed by their discoloured teeth. Different types of veneers can sometimes hide the problem. Veneers are cosmetic coverings typically placed on the outer surface of the upper anterior teeth.

Composite (white material) veneers are suitable for children and adults, as they require minimal tooth removal. Adults can have veneers or crowns placed to change the appearance of the anterior “smiling teeth.” Veneers can last for years and have the merit of being relatively inexpensive, versatile and are effective for hiding unsightly tooth colour. Bleaching is not usually recommended because the discoloration is not in the enamel, but has been shown to be effective, although takes longer than normal. If the teeth are wearing excessively, caps (also called crowns), will probably need to be placed on at least some of the teeth. More specialized treatment may be more appropriate for permanent teeth.

Treating Malocclusions with Orthodontia or Orthognaphic Surgery

A malocclusion is an abnormal relationship between the upper and lower teeth, which creates problems with how the teeth come together. This may be due to the relationship of the upper and lower jaws to each other, the alignment of the teeth, or both. This type of problem includes crooked teeth, "underbite," "overbite" and "open bite." Treatment usually involves braces, and is provided by an orthodontist. The particular treatment plan depends on the specific problem(s) with the bite and the teeth. If the malocclusion is caused by skeletal discrepancies, then orthognathic (jaw) surgery may be required as well. An orthodontist should examine each child with OI around the age of 7-9 years. At that time early orthodontic interventions in children who are developing a relatively small upper jaw compared to the lower jaw may help decrease the need for later orthognathicsurgery.

Although there are only a few case reports and no published studies regarding orthodontic treatment for people with OI, it seems to be safe to treat them if DI is not present. If DI is present, the orthodontist will have to decide if the enamel is strong enough for braces. Unfortunately, it is difficult to determine how strong the enamel is until it is tried. Referral to an experienced orthodontist is recommended for children with OI & DI who have malocclusion. If the malocclusion is due to a problem with the growth of one or both jaws, then a combination of orthodontic braces and orthognathic surgery may be used to align the teeth. There are a few published reports about these surgeries indicating good post-operative healing of the jaws. The same concerns that one would have with any surgery in people with OI, such as potential bleeding problems and reaction to general anaesthesia, still apply. Furthermore, the recent use of bisphosphonates to treat different bone disorderstriggers many additional questions regarding maxillo facial surgeries.



Treating Impacted Teeth

The dentist needs to consider if the impacted teeth should be left alone or extracted, or if an attempt should be made to move them into a functional position in the mouth. To move a tooth, a coordinated effort is needed between the paediatric dentist/oral surgeon and the orthodontist to surgically uncover the impacted tooth and glue an attachment onto the tooth so that light force from the braces can be used to bring the tooth into the proper position.

Dental Implants

Dental Implants are used to replace missing teeth. Theoretically it is possible to do this successfully for a person with OI and there is anecdotal evidence that this has been accomplished.

However, there are no controlled studies on the use of dental implants in people with OI and only a few case reports in the literature. The high failure rate, reported to be 50 per cent within 3 years of surgery is a concern.

Dental implants are somewhat like screws. In order to function, there must be enough bone in the jaw for the implant to be securely placed. After healing, a “post” is placed in the implant and an artificial tooth is attached. Good, strong healing around the implant is critical.

Bisphosphonates

An increasing number of children and adults with OI receive bisphosphonates as part of their treatment. There have been reports in medical journals suggesting a link between bisphosphonates and areas of dead bone (osteonecrosis), particularly in the jaw. Osteonecrosis could be caused by the type of bisphosphonate, the dose or the frequency of treatment. There have been no reported cases of bisphosphonate induced osteonecrosis of the bone (BON) for children with OI, it may be prudent to take precautions. People with OI taking a bisphosphonate should be closely monitored by a doctor and a dentist. Good oral hygiene along with regular dental care to prevent infections or periodontal (gum) disease lowers risk. When possible, required dental surgery should be scheduled prior to starting bisphosphonate treatment. Elective jaw surgery, including dental implants, should be avoided during intravenous bisphosphonate therapy. Extraction of third molars (wisdom teeth) should be deferred until more information is available.

Bisphosphonates work by reducing the remodelling rate in the skeleton. It is also not clear what effect bisphosphonates have on young children whose new teeth are erupting as they gro. There have been no reported contraindications for orthodontic treatment in children with OI, although tooth movement may take longer, extending the period of wearing braces. Similarly, the effect of bisphosphonates on the necessary remodelling surrounding dental implants is not understood.



Locating a Dentist

There is no national list of dentists who treat people with OI, but your OI team may have links with paediatric dentists in their units. Schools of dentistry, or the dental department at major medical hospitals, may be helpful in locating dentists who are familiar with OI and DI.

References

- Delivering better oral health: a quick guide to a healthy mouth in children (2017).
https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/601834/healthy_mouth_children_quick_guide.pdf
- Malmgren B, Andersson K, Lindahl K, Kindmark A, Grigelioniene G, Zachariadis V, et al. Tooth agenesis in osteogenesis imperfecta related to mutations in the collagen type I genes. *Oral Dis.* 2016.
- Rizkallah J, Schwartz S, Rauch F, Glorieux F, Vu DD, Muller K, et al. Evaluation of the severity of malocclusions in children affected by osteogenesis imperfecta with the peer assessment rating and discrepancy indexes. *Am J Orthod Dentofacial Orthop.* 2013;143(3):336-41.
- Johnson A, Hodgson TA. Dental extractions in children with osteogenesis imperfecta managed with bisphosphonates. *Oral Dis.* 2010;16 (6):571.

Notes

Scan to donate
to the BBS

If you found our factsheet helpful and would like to support the BBS in continuing to provide resources, you can donate by scanning our QR Code



Compiled by the Brittle Bone Society in collaboration with BBS Medical Advisory Board and POINT (Paediatric Osteogenesis Imperfecta National Team). The information in this leaflet is correct as at 31st July 2021 but we cannot guarantee that it will be accurate and current at any given time. This leaflet is not intended in any way to replace the advice of your doctor or other medical professional. Leaflets are available online at www.brittlebone.org. This information is available in accessible formats on request.

The Brittle Bone Society (BBS) is a registered charity in Scotland (SCO50854) and company limited by guarantee (SC677346), supporting the OI community throughout the United Kingdom and in Ireland.