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Review Article

Dental Concerns of Children with Fragilitas Osseum - A Short Review

Nirmala SVSG[1]* and Saikrishna Degala[2]

[1]*Professor, Department of Pedodontics and Preventive Dentistry, Narayana Dental College and Hospital, Nellore, India*

[2]*Professor and HOD, Department of Oral and Maxillofacial Surgery, JSS Dental College and Hospital, Mysore, India*

***Corresponding Author:** Nirmala SVSG, Professor, Department of Pedodontics and Preventive Dentistry, Narayana Dental College and Hospital, Nellore, India.

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Abstract

Osteogenesis imperfecta (OI) comprise a heterogeneous group of diseases characterized by susceptibility to bone fractures with inconsistent severity and in most cases, with assumed or proven defects in collagen type I biosynthesis. Incidence is approximately 1 per 20,000 live births. It is autosomal dominant (AD) inheritance. The clinical manifestations observed in patients with OI are abnormal bone formation, growth deficiency, bone fragility, blue sclerae, hearing losses, skin thinness, joint laxity, hypermobility, and dentinogenesis imperfecta. Delayed eruption pattern, primary dentition may demonstrate evidence of attrition, class III dental malocclusion with a high occurrence of anterior and posterior crossbites and open bites. Impacted first or second molars, missing teeth, and ectopic eruption occur more frequently in this population. Easiness of fracturing bone or teeth and difficulty intubation due to patients short necks, large tongues, or thoracic deformities are concerns when surgery is required. Primary teeth are usually treated with stainless steel crowns to protect the dental complex and to reestablish a vertical dimension. Discolored permanent teeth can restore employing porcelain or composite veneers and full. Sealants must be placed on posterior teeth to avoid occlusal dental caries. Bonding with resin is flourishing. In severe cases, teeth may require extraction and replace with fixed partial dentures, partial removable dentures, complete dentures, or overdentures depending upon the treatment required. We present about etiology, clinical features along with dental management of children with osteogenesis imperfecta.

Keywords: *Children; Bisphosphonate; Collagen; Oral Health; Osteogenesis Imperfecta*

Introduction

Osteogenesis Imperfecta (OI) is a heterogenous disorder resulting from both quantitative and qualitative defects in type I collagen [1]. The first scientific description of OI dates from 1788. Other Names for this Condition is Brittle bone disease, Fragilitas ossium, OI, Vrolik disease [2]. The incidence of approximately 1 per 20,000 live births [3]. OI does not seem to have a predilection for any particular race and Gender.

Mutations in the COL1A1, COL1A2, CRTAP and P3H1 genes cause osteogenesis imperfecta. Mutations in the COL1A1 and COL1A2 genes are responsible for more than 90 percent of all cases of osteogenesis imperfecta [4].

Classification

Four major types of osteogenesis imperfecta are often distinguished on the basis of clinical features and disease severity, according to the classification suggested by Sillence [5].

Features	Type I	Type II	Type III	Type IV
Mode of Inheritance	Autosomal dominant inheritance	Autosomal dominant inheritance	Autosomal recessive inheritance	Autosomal recessive inheritance
Incidence/ prevalence	A prevalence of 3 - 4/100,000 and an incidence of 3.5/100,000	Incidence is about 1-2/100,000 prevalence data are not available due to early lethality	Prevalence of 1 - 2/100,000 An incidence of 1.6/100,000	Believed to be a rare entity abnormalities are present at birth in approximately 50% of patients
Stature	Normal Stature	------	Short	short
Fracture	Most of the fractures occur during the preschool years and are less common after puberty	Exhibits extreme bone fragility and frequent fractures, which may occur during delivery	Fractures may be present at birth	Fractures present during childhood and decreases after puberty
Deformity level	Moderate	Severe	Progressively deforming	Mild to moderate
Congenital fractures	No	Yes	Usually	Rarely
Sclera	Blue sclera	Blue sclera	Normal or pale blue at birth; fades with age	Normal or pale blue at birth; fades with age
Joint hyper mobility	yes	yes	Yes	Variable
Hearing loss	Present in 60% of cases	NA	NA	Present in 42% of cases
Age	For type I, the age of onset is variable. This form most commonly appears during the preschool years when the child is starting to stand.	Type II occurs in utero.	Abnormalities are present at birth (i.e. abnormalities develop inutero) in more than 50% of patients. Fractures are frequent during the first 2 years of life	Abnormalities are present at birth in approximately 50% of patients. onset of this form is during infancy or the preschool years
Clinical features	Possible dentinogenesis imperfecta	Perinatally lethal	Dentinogenesis imperfecta	Possible dentinogenesis
Radio-graphic features	Skull shows Wormian bones and back reveals codfish vertebrae (adults). Extremities show thin cortices. Osteopenia is present.	Skull shows under mineralization with plaques of calcification and back reveals platyspondyly. Extremities are severely deformed; Broad, crumpled and bent femurs are seen. Small beaded ribs are pathognomonic of IIA and pectus excavatum in IIB.	Skull shows Wormian bones, frontal bossing, and micrognathia and back reveals codfish vertebrae, kyphoscoliosis, and platyspondyly. Extremities show flared metaphyses ("popcorn like" appearance [childhood]), bowing, and thin cortices. Other features include thin ribs, severe osteoporosis by dual energy X-ray absorptiometry (DEXA).	Skull may or may not show Wormian bones. Back reveals Codfish vertebrae. Extremities show thin cortices. Other features include protrusio acetabuli

Table 1: *Showing clinical features of OI.*

Clinical features: OI is a disease in which there is enlarged fragility of bones and a tendency to fracture with only slight trauma. Cortical bone is thinner than usual due to distressed osteoblastic activity and cancellous bone has wide spaces and superior trabeculae. After a fracture, the callus forms usually, but it may replace with bone, which is even further lower to the original. The deficiency is one of mesoderm and as well as bone, other tissues exaggerated. Such a patient may have blue sclera due to its thinness or transparency, lax ligaments with a tendency to dislocations, and a disturbance of dentine formation (dentinogenesis imperfecta). Some develop deafness from otosclerosis as adults [8].

There appear to be two slightly different types of the disease, congenital and a later variety, tarda, or osteopsathyrosis. In the primary class, fractures may occur early in the utero and heal with the bones in an abnormal position. As a result, there may be considerable deformity at birth. Fractures also occur during delivery, and the infant may sustain several involving long bones, ribs, and skull. By adolescence, there may be an accumulated history of a dozen or more episodes. This type is probably due to an inherited recessive character [9].

In osteopsathyrosis, the fragility does not become apparent until the child has passed his first year or later, and as a generalization, the later the manifestations appear, the less severe the condition. As with the congenital type, there is an improvement at puberty. This variety inherited as a dominant trait, and half of the second generation may expect to possess it [10,11].

Children with OS, Class III dental malocclusion take place in 70% - 80% of types III and IV of the OI population, with a broad incidence of anterior and posterior crossbites and open bites [12]. Chang., *et al*. did a study to identify craniofacial characteristics in OI, which demonstrate that the OI patient had a more prominent Class III occlusal relationship, prognathic mandible, more extensive facial divergence. Shorter face heights, defective sagittal growth of the maxilla and mandible, a flattened cranial base angle, impaired cranial base growth, and more forward rotation in mandibular growth compared with the controls [13].

The characteristic radiographic findings of DI type I are bulbous crowns, constriction of the cementoenamel junction and obliteration of pulp. Slight to marked attrition of the occlusal surface and roots are short and slender [14].

In 2009, the International Nomenclature Group for Constitutional Disorders ICHG of the Skeleton (INCDS) proposed that the OI syndromes classified as five different groups rely on phenotype alone (Table 2) [15].

New OI classification/OI type	Phenotype
1/I	Mild, non-deforming
2/II	Severe, seen as perinatal and lethal forms
3/III, VI, VIII, IX, X, Bruck syndrome Type 1	Moderately severe, progressively deforming
4/IV, IV, VII, XI, XII, XIII	Moderate
5/V, osteoporosis pseudoglioma syndrome, idiopathic juvenile osteoporosis, Bruck syndrome Type 1 and Type 2	Moderate, calcification of the interosseous membranes seen

Table 2: *The international nomenclature group for constitutional disorders ICHG of the skeleton 2009.*

A significant difficulty in the dental care of a patient with osteogenesis imperfecta is the bone condition and a simple extraction can result in fracture of the mandible or alveolus. Any patient observed to have blue sclera should be carefully questioned for any history of fracture, as more than half of them suffer from bone fragility. During extractions, the tooth should cautiously luxate, employing small amplitude movements and nominal force. The child should be prepared himself/herself contented in the dental chair. The majority of these children have healthy intellectual development and are capable of co-operating with the treatment [16].

In these cases, prophylactic premedication is essential before highly invasive dental procedures (e.g. extractions and periodontal surgeries) may be a sensible approach. However, any apparent probable advantage of antibiotic prophylaxis ought to weigh beside the known risks of antibiotic toxicity, allergy, and development, selection, and transmission of microbial resistance [17].

The ease of fracturing bone or teeth and difficulty intubation due to patients' short necks, large tongues, or thoracic deformities are concerns when surgery is required [18].

Dislocation

During conservation and wide mouth-opening, it is as well to bear in mind the liability to disruption. While these may quickly reduce, the neck of the mandibular condyle is both slender and fragile [19].

Tooth structure

A proportion of patients with osteogenesis imperfecta also suffer from dentinogenesis imperfecta (opalescent dentine). In this case, the teeth appear more translucent than regular with a brownish or grey-brown discoloration. Although the enamel seems to be healthy, it chips badly, and attrition is a feature. Radiographically, the roots may be short and slender, having narrowed suddenly at the neck, and the pulp canals may be occluded or so tine as to be challenging to identify. Periapical infection is relatively common and obliteration of pulp chamber and root canals; hence root canal therapy usually impossible. The teeth are brittle and extractions hazardous for this reason. Histologically, the dentine is abnormal with occluded tubules at the periphery, but so deranged centrally that there are vascular inclusions. There are defective calcification and lack of cement substance [20].

It has observed that the typical radiographic appearance of slender roots, occluded pulp canals, and clinical discoloration may be less noticeable when accompanied by the bone disease than when occurring alone.

Treatment

Management involves surgical and medical treatment of skeletal abnormalities, and treatment of other complications. More innovative approaches based on gene and cell therapy, and signaling pathway alterations, is under investigation.

The aim of dental care should be towards preventing the need for extractions at least until after puberty when the bone fragility improves. Account must take, however, of those cases in which dentinogenesis imperfect makes this very difficult both aesthetically and by severe attrition. Sealants must be placed on posterior teeth to avoid occlusal dental caries. Bonding with resin is successful [21].

Some of the dental professionals may be reluctant to provide services for individuals with OI for fear of causing fractures. In most cases, no peculiar care is necessary, except for ensuring patient comfort. Using beanbags, pillows, or padding in the dental chair is instrumental. Clinicians need to be gentle while doing the treatment and avoid spontaneous movements or excessive force [12,22].

Children with OI suffer from restrictions in day-to-day living, which may give rise to emotional and psychological problems for both the parents as well as children. Parents or caregivers may be overprotective or have low coping strategies, and they may undergo despair. Psychiatrists suggest that parents of children with OI seek support from organizations or groups whose members experience similar problems [23,24].

Medical treatment

There is no cure for OI. Orthopedic rehabilitation and medical management of the symptoms are the only treatment. Splints, casts, and harnesses provide immobilization of fractured bones. Ambulation may take account of crutches, braces, walkers, or a wheelchair. Physical therapy helps make stronger weak muscles. Swimming, walking in safe environments [23]. Bisphosphonates slow down bone resorption, therefore, improving bone mass. Likewise, they alleviate chronic pain and acquire enhanced mobility [25].

In proceedings with a record of multiple bone fractures, OI ought to distinguish from assumptive child abuse. To this end, the clinician must prevail in excellent medical and family histories. Although the incidence of OI is infrequent, the being of the blue sclera, aberrant teeth, hearing problems, osteoporosis, wormian bones, joint laxity, and short stature can be well thought out as positive findings of OI, as their proximity in child abuse is rarefied. If the child has DI, the clinician should rule out the diagnosis of OI [26,27].

Conclusion

Early diagnosis of OI is vital to prevent subsequent physical and psychological damage to such patients. So, the pediatric dentist can play a starring role in the primal diagnosis of OI and impart for improvement of quality of life of children with OI. It is essential that educational services on correct oral home care and disease prevention strategies taught to the parents as well as caregivers.

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