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ORAL HEALTH STATUS OF CHILDREN WITH OSTEOGENESIS IMPERFECTA

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The experimental protocol was approved by the Ethics Research Committee of the Dental School of the National and Kapodistrian University of Athens (NKUA) (396/ 20_12_2018) and a written informed consent was obtained from all the participants and their parents. The study was conducted according to the declaration of Helsinki.

Pre- phase

This study was conducted to investigate the oral health status of children with OI, a genetic disease of the connective tissue, which is also the most common form of primary osteoporosis, and compare it with healthy individuals. Information from this research would be really helpful in understanding the needs of these young patients and, as dentists, planning a preventive program and treatment protocol for them. It would also be interesting to see the oral health status of OI patients in our country, since there are no relevant data, to our knowledge.

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Introduction

Definition, epidemiology

Osteogenesis imperfecta (OI) - also known as brittle bone disease - is a heritable disorder of connective tissue metabolism, characterized by susceptibility to bone fractures (Andersen et al, 1989).

OI was at first described in 1788 by a Swedish army surgeon, Olaus Jakob Ekman, who reported a family with hereditary bone fragility in his thesis on congenital osteomalacia. The term “osteogenesis imperfecta” was introduced by Willem Vrolik, a Dutch professor at the Athenaeum Illustre of Amsterdam, who suggested that OI is probably a genetic rather than an acquired disease (Dijk et al, 2011).

Osteogenesis imperfecta is a rare disease with a birth prevalence of approximately 0.3–0.7 per 10,000 births in Europe and the United States. However, the studies that estimate the prevalence of OI often include the more severe types of the disease and not the more-subtle ones that become apparent after birth. There is no difference in the prevalence between sexes (Marini et al, 2017).

OI types

In the late 1970s David Sillence and his colleagues proposed a classification for osteogenesis imperfecta and classified the disease into four distinct types based on the distinctive clinical presentation, radiographic characteristics and patterns of inheritance. There were two known genes at the time, causing the disorder, i.e. COL1A1 and COL1A2. Nowadays, there are more than 15 genes responsible for similar phenotypes. Therefore, this classification was expanded based on distinguishing clinical features or based on the genetic cause, where new genes were given additional type numbers (Marini et al, 2017). According to Van Dijk and Sillence DO, the new clinical OI classification comprises five types; the already known I, II, III and IV types plus type V, which has a dinstict phenotype (Van Dijk and Sillence 2014) (see Table 1).

New OI Sillence classification

- Type I: is the mildest form, has a triad of clinical characteristics: fractures, blue sclera, and hearing loss. Fractures often begin with ambulation and decrease after puberty. These individuals have minimal bone deformity, near normal stature and rarely have dentinogenesis imperfecta (DI).
- Type II is perinatally lethal. Affected infants have short, bowed long bones with crumpling from in utero fractures, blue/grey sclerae, and a large, soft cranium. Radiographs reveal undertubulated long bones. The most common cause of death is respiratory failure, associated with small thorax, rib fractures, pneumonia, frequent pulmonary infections and perhaps with intrinsic collagen related abnormalities of lung tissue.
- Type III (progressively deforming) is the most severe, non-lethal form. Affected individuals may sustain numerous fractures. Most have triangular facies, frontal bossing, blue/grey sclerae, DI, vertebral compressions and scoliosis. Many have platybasia or basilar invagination. They have extremely short stature; about half have “popcorn” formation at femoral growth plates (sclerotic lines seen on radiographs representing growth plate fragmentation).
- Type IV (moderately severe) has a broad phenotypic range, overlapping types I and III. Affected individuals incur dozens of long bone fractures, but most achieve ambulation. Scleral hue, DI, basilar impression, hearing loss and final stature are variable (Forlino et al, 2011).
- Type V is of moderate severity and it’s distinct from the above types, because of the hypertrophic callus formation after a fracture, limitations in the range of pronation/supination of the forearms, associated with a radiologically apparent calcification of the interosseous membrane and a tendency towards anterior dislocation of the radial head.

Several versions of an alternative classification have been proposed, in which the recessive types are folded into the original Sillence numeration based on clinical phenotype; these classifications vary in whether the histologically defined types V and VI are retained or also classified clinically. The alternative classification results in children with defects in the same gene (ie LEPRE1) being classified as different types because of clinical variability (ie type II for lethal cases, or III for severe survivors).

Table 1: New Sillence clinical classification of OI and genes implicated in the pathogenesis (Van Dijk and Sillence, 2014)

OI syndrome names	Type	Gene	MIM	Locus	Protein product	Inheritance
(A)						
Non-deforming OI with blue sclerae	1	1. *COL1A1*	#166200	17q21.33	Collagen alpha-1(I) chain	AD
		2. *COL1A2*	#166200	7q22.3	Collagen alpha-2(I) chain	AD
Common variable OI with normal sclerae	4	1. *COL1A1*	#166220	17q21.33	Collagen alpha-1(I) chain	AD
		2. *COL1A2*	#166220	7q22.3	Collagen alpha-2(I) chain	AD
		3. *WNT1*[a]	#615220	12q13.12	Wingless-type MMTV integration site family, member 1	AD
		1. *CRTAP*	#610682	3p22.3	Cartilage-associated protein (CRTAP)	AR
		2. *PPIB*	#259440	15q22.31	Cyclophilin B (CyPB)	AR
		3. *SP7*	#613849	12q13.13	Osterix	AR
		1. *PLS3*		Xq23	Plastin 3	XL
OI with calcification in interosseous membranes	5	1. *IFITM5*	#610967	11p15.5	Interferon-induced transmembrane protein 5	AD
(B)						
Progressively deforming	3	1. *COL1A1*	#259420	17q21.33	Collagen alpha-1(I) chain	AD
		2. *COL1A2*	#259420	7q22.3	Collagen alpha-2(I) chain	AD
		1. *BMP1*	#614856	8p21.3	Bonemorphogeneticprotein 1	AR
		2. *CRTAP*	#610682	3p22.3	Cartilage-associatedprotein (CRTAP)	AR
		3. *FKBP10*	#610968	17q21.2	Peptidyl-prolyl cis-transisomerase FKBP10	AR
		4. *LEPRE1*	#610915	1p34.2	Prolyl 3-hydroxylase 1 (P3H1)	AR
		5. *PLOD2*	#609220	3q24	Procollagen-lysine, 2-oxoglutarate	AR
		6. *PPIB*	#259440	15q22.31	5-dioxygenase 2	AR
		7. *SERPINF1*	#613982	17p13.3	Cyclophilin B (CyPB)	AR
		8. *SERPINH1*	#613848	11q13.5	Pigment-epithelium-derived factor (PEDF)	AR
		9. *TMEM38B*	#615066	9q31.1	Heat shock protein 47 (HSP47)	AR
		10.*WNT1*	#615220	12q13.12	Trimeric intracellular cation channel B (TRIC-B)	AR
		11.*CREB3L1*		11q11	Wingless-type MMTV integration site family, member 1	AR
					Old Astrocyte Specifically induced substance (OASIS)	AR
Perinatally lethal OI	2[b]	1. *COL1A1*	#166220	17q21.33	Collagen alpha-1(I) chain	AD
		2. *COL1A2*	#166220	7q22.3	Collagen alpha-2(I) chain	AD
		1. *CRTAP*	#610682	3p22.3	Cartilage-associated protein (CRTAP)	AR
		2. *LEPRE1*	#610915	1p34.2	Prolyl 3-hydroxylase 1 (P3H1)	AR
		3. *PPIB*	#259440	15q22.31	Cyclophilin B (CyPB)	AR

Diagnosis

OI can be diagnosed through the patient's history and clinical (based on skeletal and/or extraskeletal manifestations) as well as radiographic examination. Collagen testing, molecular testing by DNA sequence analysis, and prenatal testing by ultrasound or amniocentesis may also confirm the disease (Muhney et al, 2007). Specifically, a genetic test can be very advantageous in locating possible mutations, determining the risk of recurrence in the same family (dominant vs recessive OI), and identifying other members of the family that are affected but have subtle clinical phenotype (Trejo et al, 2016).

Differential diagnosis

Differential diagnosis varies with the severity of OI and age of the patient. Prenatally, in ultrasounds, severe OI may be confused with thanatophoric dysplasia, achondrogenesis type I, or camptolic dysplasia, all of which demonstrate relatively large heads and short limbs. Type III OI may need to be distinguished from infantile hypophosphatasia, which presents with severe osteoporosis and micromelia. Type IV and more severe type I OI may be confused with primary juvenile osteoporosis or other secondary causes of osteoporosis in childhood, such as hypogonadism or malignancy. The major differential diagnosis of type I OI is child abuse (Fotiadou et al, 2016).

In infants, the differential diagnosis of OI includes prematurity, hypophosphatasia, child abuse, arthrogryposis multiplex congenital, Bruck syndrome, geroderma osteodysplasticum, Osteoporosis Pseudoglioma (OPPG) Syndrome and vitamin and mineral deficiencies.

In childhood as well as adolescence, OI may be confused with mucolipidosis type II (I-Cell Disease), idiopathic juvenile osteoporosis, exercise-related osteoporosis, hypophosphatasia (HPP), Ehlers–Danlos syndrome (EDS), malabsorption syndromes, disuse osteoporosis or iatrogenic osteoporosis (Sutton, 2014).

Aetiology

Approximately 90% of OI is caused by autosomal dominant inherited mutations in either the COL1A1 or COL1A2 genes encoding type 1 collagen. Mutations that lead to functional null alleles (decreased collagen protein production) generally result in less severe disease than those which lead to structurally abnormal collagen. Also, since type 1 collagen is comprised of 2 α1-chains and 1 α2-chain, mutations in COL1A1 are generally more deleterious than those in COL1A2, since the A1 collagen chains make up two thirds of each collagen fibril complex. The most commonly observed structural collagen abnormalities occur when the sterically hindered glycines in the collagen fibril are replaced by a larger amino acid, which then disrupts the folding process of the collagen into a triple helical structure. The substitution results in an abnormally shaped collagen protein with inhibited right folding. The abnormal collagen works in a dominant-negative fashion to disrupt the type 1 collagen helix assembly and weakens the extracellular matrix. Depending on where the glycine substitution occurs, the resultant collagen product stability can range from mild to severe with consequently different clinical outcomes (Thomas et al, 2016). Furthermore, there are also other osteogenesis imperfecta-causing, noncollagenous genes, encoding proteins involved in collagen biosynthesis, or transcription factors and signaling molecules related to bone cell differentiation and mineralization, and are associated with an autosomal recessive (most commonly), dominant or X-linked inheritance (Rossi et al, 2019).

Recessive OI with moderate to lethal phenotypes is caused by defects in genes whose products interact with type I collagen. Most recessive cases have null mutations, causing absence of proteins involved in collagen prolyl 3-hydroxylation (CRTAP, LEPRE1 and PPIB) 3–8, or helical folding (FKBP10 and SERPINH1) (Forlino et al, 2011).

Clinical features

The severity of clinical features of OI at birth ranges from no clinical features to prenatally lethal skeletal abnormalities (Dijk et al, 2011).

Skeletal manifestations

OI is characterized by osteopenia and fractures, most commonly long bone fractures as well as others, such as vertebral fractures (Thomas et al, 2016). Fracture rates tend to decrease after adolescence, but can then increase again later in life. They are usually caused by bone fragility or by acquired bone fragility due to muscle wasting and immobilization (Fotiadou et al, 2016). Limb deformity including bowing can be present both prior to fracture as well as resulting from fractures (Thomas et al, 2016). In the long bones, bending and thinning of the diaphyses can be seen. Fractures usually occur in the concave aspect of the deformity and can recur after healing. Severe residual angulation of a healed fracture may be encountered. In toddlers with severe forms of OI (mainly type III), the long bones may appear thick and broad with a "bamboo cane appearance." Other common long bone deformities constitute the "shepherd's crook" deformity of the femur (anterolateral bowing) and the "sabre shin" deformity of the tibia (anterior bowing) (Fotiadou et al, 2016).

Patients with OI present with altered facial characteristics which are related to the severity of the OI type. OI patients usually present with a triangular face, protrusive bitemporal bone and prominent frontal bone, an overhanging occiput, and a relatively larger head circumference (Chang et al, 2006). The Wormian skull bones are found in approximately 60% of affected individuals and are found to correlate with the genotype. Short stature is common and may correlate with the gene mutated, the location of the mutation in the gene, and the amino acid substitution. Scoliosis and kyphosis are also common (Thomas et al, 2016).

Eye manifestations

One of the most common clinical features of OI is the color of the sclerae, which can be normal- white, gray -or light/dark blue. The color may be darker in infants and lighten with age. Blue sclerae is found in almost half of the types of OI and is probably related to the thin central corneal thickness. The thin corneal thickness may also predispose to glaucoma. Other eye manifestations seen in patients with OI are cataracts, ectopia lentis and presbyopia (Thomas et al, 2016).

Pulmonary manifestations

Scoliosis and rib fractures in patients with OI can often cause obstructive pulmonary disease and show progressive decline in pulmonary function parameters. These pulmonary complications are the main cause of death in osteogenesis imperfecta. Thus, in cases of vertebral or chest wall deformities, a spirometry test should be performed for diagnosing possible restrictive or obstructive disease. Recent studies show that pulmonary function diminishes in osteogenesis imperfecta with age and lung disease is not necessarily associated with scoliosis (Marini et al, 2017).

Cardiovascular manifestations

In OI the abnormal type I collagen, which is a major component of the extracellular matrix of cardiac valves and the aortic wall, can cause various cardiac anomalies such as aortic root dilatation (Marini et al, 2017), valvular insufficiency, aortic root dilatation, atrial septal defects and septal and posterior left ventricular wall thickening (Forlino et al, 2011). In cases of scoliosis, chest deformity, heart murmurs, or any cardiac or pulmonary symptomatology echocardiography should be performed and the patient should be examined by cardiologists (Marini et al, 2017).

Bleeding diathesis

Another manifestation of OI is platelet dysfunction and vessel fragility due to tissue fragility and a bleeding diathesis and these conditions can cause subdural and epidural haematomas to patients, even after a slight trauma and a higher risk of surgical complications (Marini et al, 2017).

Hearing loss

Hearing loss is a common secondary feature of OI as it can appear in almost 50% of the patients by 50 years of age (Thomas et al, 2016) and is progressive as well as mainly bilateral (Forlino et al, 2011). Hearing loss is usually caused because of otosclerosis, bony changes (Marini et al, 2017), atrophy, fractures of the ossicles and neural degeneration (Thomas et al, 2016). It may appear in three types: conductive, sensorineural or mixed, with the initial conductive deafness evolving into the mixed (conductive and sensorineural) type. All patients with osteogenesis imperfecta should

be evaluated for hearing loss on a regular basis, starting in childhood, and referred for hearing aids, stapes surgery or cochlear implants as needed (Marini et al, 2017).

Other manifestations

Neurologic sequelae are also common in children with OI. They may develop macrocephaly or hydrocephalus, conditions that require shunting. Approximately 8-25% develop secondary basilar invagination, a disorder characterized by infolding of the skull basiocciput with resultant upward translocation of C1 into the posterior fossa. Many of these patients don't have any clinical symptoms while others may present with sleep apnea, headache, ataxia, nystagmus, cranial nerve palsies, and even quadriparesis. Additionally, about 30% of OI patients may have hypercalciuria, which is associated with an increased kidney stone risk (Thomas et al, 2016).

Oral manifestations

OI can affect both teeth and jaw development and growth, causing various disorders in the stomatognathic system. The most frequently occurring dysfunctions include sucking and swallowing disorders, improper structure of the masseter muscle, and biomechanical temporomandibular joint disorders (Smolag et al, 2017). Underdeveloped nasomaxillary complex (hypoplastic maxilla) in all 3 planes of space leads to counter-clockwise rotation of the mandible, causing skeletal discrepancies between the jaws. This discrepancy is translated into dental malocclusion in all dimensions, namely sagittal (dentoskeletal class III, anterior crossbite), vertical (anterior and posterior open bites), and transverse (lingual posterior crossbite). In addition, dentinogenesis imperfecta, taurodontism, agenesis, tooth impactions (mostly premolars and second molars), as well as the ectopic eruption of teeth, contribute to malocclusion in this population. The incidence of class III malocclusion is high and associated with anterior and posterior open bites, as well as crossbites. Orthodontic and orthognathic surgery interventions may be contraindicated in the OI population because of the poor quality and quantity of bone and teeth, and use of bisphosphonates (Najirad et al, 2020).

One of the most common dental finding in patients with OI is the presence of dentinogenesis imperfecta (DI). DI has three types and from those, type 1 is associated

with OI (Majorana et al, 2010). In DI the enamel is normal with normal or infrequently decreased mineral content (Vital et al, 2012), but dentin is affected and histologic analysis shows microscopic disturbances as early as the tooth germ stage (Schwarz et al, 1984). Specifically, the initially formed mantle dentin is normal while the rest of the dentin has an irregular, dysplastic texture with amorphous areas, abnormal number and structure of dentin tubules, embedded cells, and occasionally interglobular dentin. Large spots of nonmineralized matrix and even zones without any dentinal tubules are present (Vital et al, 2012). The light diffraction through the defective dentin–enamel junction (Vital et al, 2012) gives those DI teeth a characteristic discoloration that ranges from grey- brown to opalescent blue (Majorana et al, 2010). This discoloration is not associated with the type of the OI but it has been shown that teeth with yellow- brown discoloration may experience more enamel fractures and attrition than those with grey discoloration (Forlino et al, 2011). The crowns may appear normal, bell- shaped (Schwarz et al, 1984) or bulbous with a distinct cervical constriction and the teeth are susceptible to severe attrition. The radiographic examinations show short, narrow roots and pulpal obliteration due to dentine hypertrophy and this condition appears before or just after eruption (Chetty et al, 2016). Generally the primary dentition is more severely affected than the permanent possibly due to the more rapid formation of the primary teeth and the greater expression of collagen during the embryonic developmental stage (Vital et al, 2012). The degree of dental involvement may vary within a single dentition (Schwarz et al, 1984).

Other atypical dental features have been reported in OI, such as increased rate of heterotopic eruption of first and second molars and a high prevalence of permanent tooth agenesis (10% to 22%) (Vital et al, 2012).

Craniofacial and dental disturbances are more pronounced in OI types III and IV, while type I shows almost normal characteristics (Jabbour et al, 2018).

In cases of patients with OI undergoing treatment with bisphosphonates there might be an effect on the teeth. Bisphosphonates are well established medications used to inhibit the function of osteoclasts and prevent bone resorption. Eruption of any tooth requires resorption of the alveolar bone, and in question of permanent teeth other

than molars, also resorption of the roots of the pre-existing primary teeth. Since osteoclasts are responsible for the resorption of both, bisphosphonate treatment can be anticipated to delay tooth eruption (Vuorimies et al, 2017). Also, prolonged treatment with high doses of bisphosphonates has been associated with jaw osteonecrosis. To date, there is no report of such an event in children with OI or other causes of osteoporosis receiving bisphosphonates; however, regular and meticulous dental follow up is advised.

Management- treatment

Management is symptom-based and depends on the type and severity of complications (Marini et al, 2017). For the successful management of an OI patient a multidisciplinary team should be formed, consisting of orthopedic surgeons, rehabilitation physicians, endocrinologists, physical therapists, pediatricians and dentists as well as other specialists when needed (eg. cardiologist, otorhinolaryngologists etc.) (Dijk et al, 2011). The goal of the treatment is for the patient to gain bone strength, prevent further fractures, provide comfort from pain and improve vertebral morphology (Martin et al, 2007). Management consists of pharmacological treatment, orthopedic treatment, physical medicine, dental treatment, treatment for hearing loss, and prevention of primary (e.g. basilar impression) and secondary (e.g. problems due to general medical problems) complications.

Pharmacological Treatment

Bone strength depends on bone material properties (quality), bone mass (amount) and bone architecture (distribution). The disorganized, hypermineralized bone matrix in osteogenesis imperfecta is not directly altered by any currently available pharmacological therapy. However, both anti-resorptive (for example, bisphosphonates) and anabolic (for example, growth hormone) therapies might improve bone mass (Marini et al, 2017).

Oral and intravenous bisphosphonates are commonly prescribed for all OI types. The main rationale for bisphosphonate therapy is based on several clinical trials that showed improvements of bone mineral density in individuals with OI. Nitrogenous

bisphosphonates disrupt osteoclast formation, survival and cytoskeletal dynamics, and non-nitrogenous bisphosphonates initiate osteoclast apoptosis. The use of growth hormone to affect short stature in types III and IV OI is still under active investigation (Dijk et al, 2011).

All bisphosphonates (BPs) are structural analogues of pyrophosphates and, when administered, either orally or intravenously (IV), they are adsorbed onto bone surfaces being slowly released into the bone matrix: here they bind to hydroxyapatite and inhibit osteoclastic activity. Thus, the action of BPs may last many years, even after stopping the drug. IV pamidronate is the most commonly used BP; recently, zoledronate has been proposed and applied, due to the fastest IV infusion and superior potency (Contaldo et al, 2020).

The options for anti-resorptive therapy are not limited to bisphosphonates. Osteoclast inhibition can also be achieved with denosumab, a drug based on an antibody against RANKL. On a bone histological level, denosumab seems to have a similar effect on growing children as intravenous pamidronate. However, denosumab has a much shorter duration of action than bisphosphonates, which can be seen as an advantage because it allows better control of the duration of antiresorptive action. (Trejo et al, 2016).

The administration of pamidronate to children with OI has proven a signal advance in the treatment of the disorder as it can increase the bone density for a period of about 2 to 4 years, decrease the fracture rate of about 50%, improve vertebral and relief from musculoskeletal pain (Martin et al, 2007).

Growth hormone has also been administered to both type I OI and type III/IV children in several clinical trials and it has been shown that treatment with standard doses of rGH can result in significant increases in linear growth (Forlino et al, 2011).

There is evidence that prolonged BPs therapy is associated with osteonecrosis of the jaws (BRONJs). This is a serious consideration, especially for adult patients who need invasive dental procedures, because the BPs inhibit osteoclast resorption and they have anti-angiogenic properties (Contaldo et al, 2020). The danger for BRONJ is more reasonable for the IV BPs rather than oral BPs probably due to their bioavailability.

Only 1% of the dose of an oral BP is absorbed by the gastrointestinal tract, while almost 50% of the IV BPs are absorbed by the bone matrix. The risk of osteonecrosis of the jaw is highly correlated with the type of the bone and the turnover rate. Trabecular bones present a higher rate of turnover than cortical bone and BPs have a higher affinity for bones with higher turnover rates. Since the jaw bone consists mainly of alveolar bone which is trabecular in nature and has a high turnover, it is expected to have an increased risk for BRONJ (Hennedige et al, 2013). What is not yet clear, is if BRONJs occur in children/adolescents affected by OI under BPs treatment. Literature reports no evidence, but the reasons for this difference with respect to adults still need to be elucidated (Contaldo et al, 2020).

Orthopedic Treatment

Orthopedic rehabilitation of patients with OI includes surgical as well as nonsurgical management and aims to treat the fractures, the pain and the deformities. The surgical management consists of lower extremity, upper extremity and spine surgery, and is usually combined with medical treatment and pre-surgery and post-surgery rehabilitation (Muhney et al, 2007).

For lower extremity surgery the most common procedure is treatment of deformities with osteotomies in order to straighten the bone with intramedullary rodding (insertion of a metal rod into the medullary cavity of a bone to provide strength and alignment) (Marini et al, 2017). Rodding corrects the usual bowing of bones, providing support and thus preventing fractures. The use of expandable rods that lengthen as the child grows is a better choice as they don't need replacement like the non-expandable ones (Muhney et al, 2007). Plates and screws are not recommended because of fracture risk above or below the plate due to stress. Upper extremity surgery is performed more frequently than in the past because upper extremity deformities are now understood to affect self-care, whereas they were previously considered as purely cosmetic (Marini et al, 2017). Nonsurgical management consists of braces and splints, casts and harnesses in order to immobilize fractured bones and provide comfort to the patient (Dijk et al, 2011).

Physical Therapy (Rehabilitation)

Rehabilitation is a very important part of the management in patients with OI. From the early stage of infancy, parents should be taught on how to hold and handle the affected baby. Rehabilitation is crucial and very beneficial during infancy and early childhood as it provides strength and promotes function as well as independence especially when overprotection or fear of movement prevents skills acquisition (Etich et al, 2020). Physical therapy is also helpful after an injury, a fracture or surgical procedures and acts in combination with pharmacological treatments (Marini et al, 2017). Children with OI are recommended to participate in weight-bearing physical activities regularly. However, activities which are characterized by a high fracture risk (falling from heights, trampolines, contact sports) should be avoided. For persons with moderate/severe OI, physical therapy may be recommended, and for the most severely affected, aquatherapy may be beneficial (Thomas et al, 2016).

Treatment for Hearing Loss

Hearing loss often occurs in adults with OI and it initially concerns conductive hearing loss, but as the hearing loss progresses, a significant sensorineural component emerges. Surveillance for hearing loss is advised after adolescence every 3–5 years. Initially, hearing aids will be sufficient. As the hearing loss progresses, stapedectomy (surgical procedure of the middle ear performed in order to improve hearing) can be considered for which successful outcomes have been reported; however, long-term hearing restoration may be unsatisfactory due to fragility of the ossicular middle ear structures. Cochlear implantation has been used because of the sensorineural hearing loss, but data are too limited to draw conclusions on its effectiveness (Dijk et al, 2011).

Treatment for Basilar Invagination

Basilar invagination is a rare complication occurring in adults with OI type III when the top of the C2 vertebra migrates upward which may lead to (partial) closure of the foramen magnum with hydrocephalus, pressure on the brain stem, syrinx formation, and hindbrain herniation, requiring ventricular shunt placement or surgery. Only prolonged orthotic immobilization has been proven to stabilize symptoms and arrest progression (Dijk et al, 2011).

Pregnancy and Mode of Delivery

Pregnant women with OI with skeletal deformity and short stature should be closely monitored in high-risk prenatal care clinics for the safety of the mother and the fetus which can also be affected (Dijk et al, 2011).

Dental Treatment

The aim of the treatment in these patients is a) to remove sources of infection or pain, b) restore aesthetics and c) protect posterior teeth from wear and the treatment plan depends on the age of the patient and severity of the problem (Barron et al, 2008). A child with OI should be examined by the dentist as soon as the first tooth erupts to diagnose whether dentinogenesis imperfecta exists. The diagnosis will be determined mainly with the clinical examination (tooth discoloration and opalescence) because radiographic examination might be difficult due to child's young age and cooperation (Muhney et al, 2007).

In the primary dentition sealants should be placed on posterior teeth to prevent occlusal dental caries. For aesthetic purposes, composite facings or strip crowns can be used but only when the attrition is not severe. Bonding with resin has been shown to be successful. Stainless steel crowns are usually placed in primary teeth to protect them from attrition and reestablish vertical dimension. However, if the dental wear reaches the level of the gingivae, the treatment option is fixed partial dentures, partial removable dentures, complete dentures, overdentures or even extraction if the teeth cannot be restored (Muhney et al, 2007).

As the permanent teeth erupt, they should be closely monitored in relation to the rate of tooth wear with intervention only if necessary. The goal of treatment is minimal tooth preparation until the child reaches adulthood. Cast occlusal onlays on the first permanent molars and eventually the premolars, help to minimize tooth wear and maintain the occlusal vertical dimension. Teeth with short thin roots and marked cervical constrictions however are poor candidates for crowns. A common dental finding in patients with OI is the development of abscesses due to pulp obliteration. In these cases, if conventional pulp therapy is not successful, extraction of the affected teeth is recommended (Barron et al, 2008). In OI children due to facial skeletal

anomalies a visit to an orthodontist is necessary as early as the age of seven. In some cases, orthognathic surgery may also be indicated in addition to orthodontic treatment (Muhney et al, 2007).

It is noted that patients with OI are prone to develop latex allergy and this should be determined before treatment. Dentists should also be cautious when treating a patient with OI because of the risk in causing fractures. Therefore, a comfortable placement of the patient on the dental chair using pillows or padding and gentle handling is important. Educating the OI children and their environment on proper oral hygiene practices and disease prevention strategies is crucial (Muhney et al, 2007).

Aim

The aim of the study was to record the oral health status of children with OI and compare the dental findings with healthy individuals of the same age and gender.

Specific objectives were:

a) The recording of the state of health of the teeth, the soft tissues of the mouth, the oral hygiene and the temporomandibular joint (TMJ) and the occlusion.

b) The radiographic evaluation of the hard dental tissues, of the dental age and the abnormalities in the formation of the teeth and their roots.

All of the above findings were compared with those of healthy controls.

The contribution of the present study will be the systematic recording of the oral health status of children with OI in Greece, findings that so far have not been reported in the international literature and also to raise awareness of both health professionals (dentists, pediatricians) and patients and their families about the need to ensure their dental health for a better quality of life.

Methodology

Study design

This is a cross sectional comparative study between children with osteogenesis imperfecta (OI) and healthy children. The experimental protocol was approved by the Ethics Research Committee of the Dental School of the National and Kapodistrian University of Athens (NKUA) (approval number: 396) and a written informed consent was obtained from all the participants and their parents. The study was conducted according to the declaration of Helsinki.

Sample size

The sample of this comparative cross-sectional study consisted of 40 children in the OI group and 38 in the healthy children group.

Sample selection

The children of the OI group were patients of the Department of Bone and Mineral Metabolism, Institute of Child Health, Athens, Greece, while the healthy children were selected from the Postgraduate Paediatric Dental Clinic, Dental School NKUA, matched for age and gender. Exclusion criteria for both groups were uncooperative patients and lack of parental consent and for the control group also the presence of general health problems.

Data collection

Demographic data (chronological age and sex, parental education level, area of residence), children's dental history, brushing frequency and dietary habits (consumption of sugary snacks and sweetened beverages) were collected from the parents, using self-administered questionnaires. The educational level of the mother and the father of each participant was recorded separately on a 1-10 scale (1=no school to 10=MSc/ Phd graduates) and a combined variable was constructed adding the scores of the two parents.

Dental examination was carried out at the Postgraduate Paediatric Dental Clinic, NKUA with the use of a dental probe, a periodontal probe and a mouth mirror, by the same examiner. Periodontal health was assessed by using the Löe & Silness plaque (PI) and

gingival (GI) indices at six Ramfjord teeth (Goldberg et al, 1985). All teeth were then cleaned with a prophylaxis brush and assessment of dental caries was based on the ICDAS II, converted consequently to dmft/DMFT -the highest ICDAS score was used for the conversion to dmft/DMFT (Gugnani et al, 2011). Missing teeth were scored only if they were extracted due to caries. Restorations were also recorded.

The teeth were also evaluated for possible dental anomalies based on the DDE index (Clarkson et al, 1989) and attrition using the BEWE index (Bartlett et al, 2008).

TMJ disorders were also evaluated based on the TMD pain screener (Gonzalez et al, 2011) and the teeth were also checked for possible pathological mobility and presence of abscesses.

Information about the occlusion was obtained by recording the molar and canine relationship based on the Angle classification, the presence of crossbite (anterior or posterior) and the need for orthodontic treatment using the orthodontic needs index-IOTN (Richmond et al, 1994).

Panoramic radiographs were also evaluated for possible tooth anomalies related to osteogenesis imperfecta such as agenesis, microdontia, cervical constriction, bulbous crowns, pulpal obliteration, intraradicular lesions, taurodontism or impaction.

Statistical Analysis

Descriptive statistics, Student's t-test, x2, Spearman correlation coefficient rho and hierarchical multivariate regression analysis were used to analyze the data. Level of significance was set to $P \leq 0.05$.

Results

The mean age of the children in the OI group was 9.5 (SD: 4.55) years and in the healthy children group was 9.04 (SD: 2.43) years. Most children in both groups, lived in urban areas and their parents were mostly high school graduates or had a university diploma. There were no statistically significant differences between the groups for all demographic variables (Table 1). In the OI group, 8 children had primary, 20 mixed and 12 permanent dentition while in the control group 3 children had primary, 33 mixed and 2 permanent dentition.

About one fifth of the children in both groups had never been to the dentist before. Children in both groups brushed mostly once or twice per day with no difference between groups, while only one child in the OI group and two in the healthy group used dental floss. Participants in both groups consumed sugary snacks mainly between meals, while most did not have sugary drinks on daily basis. Nevertheless, healthy children consumed daily, significantly more sugary snacks ($p=0.007$) and believed that their oral hygiene routines were better compared to their counterparts in the OI group ($p= 0.01$) (Table 2).

Periodontal status was similar between the groups, but more children in the OI group had caries on permanent teeth (marginal significance, $p=0.05$) and higher Decayed, Missing, Filled Permanent Teeth (DMFT) scores ($p<0.001$). In primary teeth, the distribution ($p=0.9$) and severity of dental caries ($p=0.75$) was similar in the two groups. Twenty five percent of the OI children had enamel dental defects and 10% had dentinogenesis imperfecta. In the healthy group, 18.4% had enamel defects. Radiographic defects were found in 45% of the OI children and they were mainly taurodontism and cervical constriction while in the healthy group 31.6% presented defects and they were mainly taurodontism and impaction (Table 3).

The orthodontic status of the children is presented in Table 3. At the time of the examination, 6 children in the OI and 4 in the healthy group were under orthodontic treatment or they had completed treatment in the past. Nevertheless, significantly more OI children had posterior crossbites ($p=0.04$), less OI children had increased overbite ($p=0.03$) and more were in need of orthodontic treatment ($p=0.03$). Finally,

OI children had slightly higher TMD Pain scores (p=0.01), but none of the children in both groups had clicking.

Bivariate correlations indicated that age (rho = 0.379, p=0.002), having OI (rho=0.524, p<0.001), urban residency (rho = -0.265, p=0.03), sugar consumption between meals (rho = 0.269, p= 0.028), presence of bleeding/calculus (rho= 0.331, p=0.006) and need for orthodontic treatment (rho = 0.364, p=0.003), significantly correlated with DMFT.

When entered in multiple regression models, having OI and sugar consumption between meals significantly predicted the DMFT, after controlling for age. There were no significant correlations between dmft and the examined parameters.

Table 1. Descriptive statistics and differences of demographic data between OI and healthy children

		OI (N=40)	**Healthy** (N=38)	P value
		% or mean (SD)	% or mean (SD)	
Gender (boys)		52.5	52.6	0.99
Age		9.5 (4.5)	9.04 (2.4)	0.58
Region (urban)		75	78.9	0.37
Father education				
	Basic	60	55.6	0.91
	Higher	26.7	33.3	
	Post-graduate	13.3	11.1	
Mother education				
	Basic	40	47.4	0.69
	Higher	46.7	47.4	
	Post-graduate	13.3	5.3	

Table 2. Descriptive statistics and differences of oral hygiene and dietary habits between OI and healthy children

	OI (N=40) %	**Healthy** (N=38) %	P value
First visit to the dentist (yes)	23.1	13.2	0.4
Brushing frequency			
2/day	30	28.9	0.23
1/day	42.5	52.6	
Few/week	27.5	16.4	
Sugary snacks consumption			
Daily	35	44.7	**0.007***
Few/week	65	55.3	
Sugary drinks consumption			
Daily	5	0	0.24
Few/week	12.5	26.3	
Rarely	82.5	73.7	
Consumption time (between meals)	82.5	89.5	0.57
Opinion on oral hygiene			
Good	22.5	52.6	**0.01***
Moderate	65	44.7	
Poor	12.5	2.6	

*statistically significant

Table 3. Descriptive statistics and differences of oral health parameters between OI and healthy children

		OI (N=40) % or mean (SD)	**Healthy** (N=38) % or mean (SD)	P value
		Dental caries		
dmft		3.19(3.81)	3.47(3.21)	0.75
Caries free (primary dentition)		22.5	23.7	0.9
DMFT		5.31(4.9)	1.74(1.83)	**<0.001***
Caries free (permanet dentition)		17.5	36.8	**0.05***
		Periodontal status		
OHI-s		2.02(0.83)	1.73(0.82)	0.12
CPI	No disease Bleeding Calculus	10 42.5 47.5	26.3 42.1 31.6	0.12
		Dental defects		
Dental defects (clinical)				
	No defects	65	81.6	**0.03***
	Enamel hypoplasia	25	18.4	
	Odontogenesis imperfecta	10	0	
Presence of radiographic dental defects		45	26.3	0.13
		Orthodontic status		
Crossbite				
	No	65	87	**0.04***
	Anterior	5	5	
	Posterior	30	8	
Permanent molar relationship	Class I Class II Class III	69 31 0	59 35 6	**0.003***
Overbite		1.88 (1.25)	2.64(1.74)	**0.03***
Overjet		1.84(2.05)	2.69(1.82)	0.06
Need for orthodontic tx (yes)		75.9	39.5	**0.03***
		TMJ status		
TMD Pain score		4.97(0.16)	4.22(1.84)	**0.01***
Midline deviation		0.62(0.83)	0.34(0.47)	0.08

*statistically significant

Table 4. Hierarchical multiple regression analysis on DMFT

	β-coefficient	95% CI	P value
Step 1			
Constant	-2.841		
Age	0.678	0.464, 0 .893	<0.001
Step 2			
Constant	-2.969		
Age	0.639	0.467, 0.811	<0.001
Having OI	3.492	-4.751, -2.233	<0.001
Between meals sugar consumption	3.060	1.254, 4.865	0.001

R^2=0.380 (P<0.001) for Step 1, R^2=0.616 (P<0.001) for Step 2

Discussion

This cross-sectional study showed that children with OI have moderate oral hygiene and significantly higher caries rates in primary and permanent dentition compared to the healthy children and diet plays an important role as a risk indicator for the development of dental caries. OI children had also more orthodontic problems and radiographic dental defects, while dentinogenesis imperfecta, was reported in only 10% of our study group.

The information about oral hygiene in OI patients in the literature is limited, however O' Connell and Marini found that the OI patients in their study had no serious periodontal health problems (O' Connell and Marini, 1999). Representative national data show that 12 yo Greek adolescents have moderate oral hygiene and brush one/day (Vadiakas et al, 2012), as the children and adolescents in this study. It seems that the medical condition is not relevant to the periodontal health and the oral hygiene practices, but follows the pattern of the children of the same age. Vettore and colleagues report that children with rare genetic diseases affecting skeletal development, such as OI, presented with poor oral hygiene and poor patterns of dental attendance compared to healthy children and that is also possibly linked to the increased levels of dental caries that they present (Vettore et al, 2020). In our group one out of five children had never visited the dentist before, conforming the above finding.

In the present study the healthy group had similar caries status to the national average (Diamanti et al, 2021), but we found that prevalence and severity of dental caries in permanent teeth was significantly higher in OI children compared to healthy children, while in primary teeth it was similar. These high caries rates do not exclusively correlate with the disease, but with the patient's dietary habits as well. Diet probably plays an important role as a risk indicator for the development of dental caries in the permanent teeth. However there are no studies in the literature that correlate sugar consumption by OI children with dental caries, although it is very well established that increased and frequent sugar consumption between meals is a risk factor for caries development. Regarding caries status, in a study of families with OI type I, it was found that the DMF ratio increased with the age of the individuals and was not related to

the presence of DI (Schwatz et al, 1984). Ma et al, (2019) also reported the caries experience of OI patients and showed that it varied with age and was more frequent in older patients compared to younger ones, as in the present study. The combination of poor oral hygiene, diet, increased prevalence of dental defects and low rates of visits to the dentist, have probably contributed to the caries status in our OI group.

Our findings that OI children have more orthodontic problems and specifically crossbite and class II malocclusion and have completed orthodontic treatment in the past, when compared to healthy participants, are in accordance with the international literature. This higher prevalence of malocclusion was also reported by Prado et al in their 2020 meta-analysis stating that Angle Class III malocclusion and anterior crossbite occur at higher rates in OI individuals compared to those without OI, due to maxillary retrognathia and/or maxillary hypoplasia in these patients.

OI children had also slightly higher TMD Pain scores, but none of the children in both groups had clicking. This finding agrees with the study of Bendixen et al. who showed that temporomandibular pain disorders are relatively rare in the OI population and if present, it is mostly present in more severe OI cases. However, the higher TMD Pain score in the OI group is probably linked to the fact that the orthodontic problems of these patients such as class III malocclusion, posterior open bite and crossbite are contributors to worse oral health related quality of life and specifically can cause functional disparities (Najirad et al, 2020).

Dentinogenesis imperfecta (DI) is one of the most common dental characteristics in OI patients but its prevalence is unknown because it depends on the OI type and the severity of the disease. In our study DI was clinically evident in 10% of the OI children and only in those with primary dentition. The prevalence of DI in patients with OI reported in other studies varies considerably, ranging from 28% to 73%, depending on the methodology of each study. In one of our patients with OI and DI, where transition to the mixed dentition had started, there were no signs of this defect in her permanent teeth. This finding is in agreement with the current literature supporting that permanent teeth are not usually affected as much as the primary ones (Forlino et al, 2011). This was also observed in another study where in some patients no involvement of the permanent dentition was observed even though the primary teeth

were affected (O' Connell et al, 1999). Furthermore, Forlino et al, (2011) suggested that very mild types of DI could be diagnosed only microscopically and could be missed radiographically. Another study also suggested that about one half of all OI patients show no obvious clinical signs of DI but it can be detected radiologically or histologically (Vital et al, 2012). Thus it is difficult to assess in details the exact prevalence of DI, unless this is clinically evident.

In our study 45%, of the OI patients had radiographic defects and mostly taurodontism and cervical constriction, findings that have widely seen in the literature among others (Thuesen et al, 2018, O'Connell and Marini, 1999). The need of radiographic examination for complete oral health evaluation in OI patients is highly indicated because the literature suggests that the teeth in OI patients with no clinical evidence of DI or any other manifestation may have radiographic defects. This was supported by Gage et al. who found that almost all teeth from OI patients are biochemically abnormal and also O' Connell and Marini who showed that teeth clinically unaffected by DI exhibited radiographic evidence of DI to a varying degree (O' Connell and Marini, 1999).

A key strength of the present study was the fact that, to our knowledge, in Greece there is no information about the oral health status of children with OI, so this study will contribute to the understanding the oral health problems of OI patients and thus the establishment a preventive protocol for them. To date, existing publications have focused mainly on case reports or series of case reports. The few observational studies examine usually individual dental characteristics of patients with OI and do not include a control group with healthy individuals to compare their findings. The comparison to a control population adds value to the findings of this study, because it highlights the specific features and dental issues of the paediatric OI population. Another advantage was the fact that the department of Bone and Mineral Metabolism of the Institute of Child Health is a key referral center, accepting paediatric patients with metabolic bone disorders from all over Greece for comprehensive evaluation of bone health. Furthermore, the mean dmft index of our control group was very close to the mean dmft score of the children of this age in Greece and that helped us make a reliable comparison (Diamanti et al, 2021).

However, a number of limitations in the present study have to be taken into account, too. The cross-sectional design of our study, does not allow for detection of specific risk factors for the oral health problems. Nevertheless, our results can give us a fairly good idea about indicators that will make a preventive program more targeted for these patients.

It is recommended that further research should be undertaken to investigate the association of medical parameters of these patients with the oral health factors. This would be useful for understanding how OI and its different types can affect the oral health and for helping the dental team organize individualized preventive and therapeutic protocols for OI patients in the future.

Conclusions

Osteogenesis imperfecta may be a rare disease but this study shed new light on the oral health status of OI children in Greece and indicates the need for incorporating the oral health evaluation at diagnosis.

In this study, patients with OI had more caries on permanent teeth and higher DMFT scores than healthy children and diet is probably an important risk factor. Furthermore, OI children had more orthodontic problems and dental defects, findings that are in accordance with the international literature. Nevertheless, dentinogenesis imperfecta, which is reported as the most common dental finding in patients with OI, was observed in only 10% of our study group. Results of this study indicate that OI patients need careful dental examination starting at an early age and targeted oral health preventive programs to ensure good oral health for a better quality of life.

Summary

Osteogenesis imperfecta (OI) is a metabolic bone disease that affects the connective tissue and specifically the collagen; it is the most common form of primary osteoporosis. Patients with osteogenesis imperfecta present with skeletal abnormalities, increased risk of fractures, muscle weakness, hearing loss, and a variety of oral findings. The aim of the study is to record the oral health status of children with OI and compare the dental findings with those of healthy individuals of the same age and gender.

Methodology

The sample of this comparative cross-sectional study consisted of 40 children in the OI group and 38 in the healthy children's group. The children of the OI group were patients of the Department of Bone and Mineral Metabolism, Institute of Child Health, Athens, Greece, whereas the healthy children were selected from the Postgraduate Paediatric Dental Clinic, Dental School NKUA, matched for age and gender. Exclusion criteria for both groups were uncooperative patients and lack of parental consent and for the control group wasthe absence of general health problems. Demographic data, children's dental history, brushing frequency and dietary habits were collected from the parents, using self-administered questionnaires. Dental examination was carried out at the Postgraduate Paediatric Dental Clinic, NKUA and information was collected on periodontal health, dental caries, occlusion, dental anomalies and TMJ. Panoramic radiographs were also evaluated for possible tooth anomalies related to the disease and estimation of dental age. Descriptive statistics, Student's t-test, x2, Spearman correlation coefficient rho and hierarchical multivariate regression analysis were used to analyze the data. Level of significance was set to $P \leq 0.05$.

Results

The mean age of the children in the OI group was 9.5 (SD: 4.55) years and in the healthy children group was 9.04 (SD: 2.43) years. Most children in both groups, lived in urban areas and their parents had basic to higher education level. There were no statistically significant differences between the groups for all the demographic variables.

About one fifth of the children in both groups had never been to the dentist before. Children in both groups brushed mostly once or twice per day with no difference between groups, while only one child in the OI group and two in the healthy group used dental floss. Participants in both groups consumed sugary snacks mainly between meals, while most did not have sugary drinks on daily basis. Nevertheless, healthy children consumed daily, significantly more sugary snacks (p=0.007) and believed that their oral hygiene routines were better compared to their counterparts in the OI group (p= 0.01).

Periodontal status was similar between the groups, but significantly more children in the OI group had caries on permanent teeth (p=0.05) and higher DMFT scores (p<0.001). In primary teeth, the distribution (p=0.9) and severity of dental caries (p=0.75) was similar in the two groups. Twenty five percent of the OI children had enamel dental defects and 10% had dentinogenesis imperfecta. In the healthy group, 18.4% had enamel defects. Radiographic defects were found in 45% of the OI children and they were mainly taurodontism and cervical constriction while in the healthy group 31.6% presented defects and they were mainly taurodontism and impaction.

At the time of the examination, 6 children in the OI and 4 in the healthy group were under orthodontic treatment or they had completed treatment in the past. Nevertheless, significantly more OI children had posterior crossbites (p=0.04), less overbite (p=0.03) and more need of orthodontic treatment (p=0.03). Finally, OI children had slightly higher TMD Pain scores (p=0.01), but none of the children in both groups had clicking. The mean estimated age from the orthopantomograms was 11.13 (SD: 3.55) in the OI and 9.86 (SD: 2.15) in the healthy group which was not statistically different. Bivariate correlations indicated that age (rho = 0.379, p=.002), having OI (rho=0.524, p<0.001), urban residency (rho = -0.265, p=0.03), between meals sugar consumption (rho = 0.269, p= 0.028), presence of bleeding/calculus (rho= 0.331, p=0.006) and need for orthodontic treatment (rho = 0.364, p=0.003), significantly correlated with DMFT. When entered in multiple regression models, having OI and between meals sugar consumption significantly predicted the DMFT, after controlling for age. There were no significant correlations between dmft and the examined parameters.

Conclusion

In this study, patients with OI had more caries on permanent teeth and higher DMFT scores than healthy children and diet is probably an important risk factor. Furthermore, OI children had more orthodontic problems and dental defects, findings that are in accordance with the international literature. Nevertheless, dentinogenesis imperfecta, which is reported as the most common dental finding in patients with OI, was observed in only 10% of our study group. Results of this study indicate that OI patients need careful dental examination starting at an early age and targeted oral health preventive programs to ensure good oral health for a better quality of life.

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Appendix

ΕΡΕΥΝΗΤΙΚΟ ΠΡΩΤΟΚΟΛΛΟ

ΑΤΕΛΗΣ ΟΣΤΕΟΓΕΝΕΣΗ ΚΑΙ ΣΤΟΜΑΤΙΚΗ ΥΓΕΙΑ ΣΤΗΝ ΠΑΙΔΙΚΗ ΗΛΙΚΙΑ: ΠΕΡΙΓΡΑΦΙΚΗ ΜΕΛΕΤΗ

ΕΝΤΥΠΟ ΚΑΤΑΓΡΑΦΗΣ ΟΔΟΝΤΙΑΤΡΙΚΗΣ ΕΞΕΤΑΣΗΣ

Αύξων Αριθμός

| |

Ον/μο Εξεταζομένου: _____ Ημ/νία Γέννησης: **……. / ……. / …….**

Στοιχεία Επικοινωνίας: _____

Α. ΔΗΜΟΓΡΑΦΙΚΑ ΣΤΟΙΧΕΙΑ

1. Τόπος διαμονής: ……
    1. Αστική περιοχή
    2. Ημιαστική περιοχή ☐
    3. Αγροτική περιοχή

2. Επίπεδο εκπαίδευσης μητέρας
    1. Δεν πήγα σχολείο
    2. Μερικές τάξεις Δημοτικού
    3. Απολυτήριο Δημοτικού
    4. Απολυτήριο Γυμνασίου
    5. Απολυτήριο Μέσης Επαγγελματικής Σχολής
    6. Απολυτήριο Λυκείου ☐
    7. Φοίτησα σε ανώτερη-ανώτατη σχολή
    8. Πτυχιούχος ανώτερης σχολής
    9. Πτυχιούχος ανώτατης σχολής
    10. Κάτοχος διδακτορικού-μεταπτυχιακού διπλώματος

3. Επίπεδο εκπαίδευσης πατέρα
    1. Δεν πήγε σχολείο
    2. Μερικές τάξεις δημοτικού
    3. Απολυτήριο δημοτικού
    4. Απολυτήριο γυμνασίου
    5. Απολυτήριο μέσης επαγγελματικής σχολής
    6. Απολυτήριο λυκείου ☐
    7. Φοίτησα σε ανώτερη-ανώτατη σχολή
    8. Πτυχιούχος ανώτερης σχολής
    9. Πτυχιούχος ανώτατης σχολής
    10. Κάτοχος διδακτορικού-μεταπτυχιακού διπλώματος

Α. ΟΔΟΝΤΙΑΤΡΙΚΟ ΙΣΤΟΡΙΚΟ Ημερομηνία

- ***Παρούσα κατάσταση***

1.Τελευταία Οδοντιατρική Επίσκεψη	1. 1-3 μήνες πριν 2. 6 μήνες πριν 3. 1 χρόνο πριν 4. 2 χρόνια πριν 5. >3 χρόνια πριν	☐
2.Αιτία Επίσκεψης	1. Τραύμα 2. Πόνος 3. Τακτικός Έλεγχος	☐
3.Συχνότητα Βουρτσίσματος	1. 1 φορά τη βδομάδα 2. 1φορά/ 2 μέρες 3. 1 φορά τη μέρα 4. 2 φορές τη μέρα	☐
4.Χρήση νήματος	0. Όχι 1. Ναι	☐
5.Χρήση στοματικού διαλύματος	0. Όχι 1. Ναι	☐
6.Χρήση γέλης χλωρεξιδίνης	0. Όχι 1. Ναι	☐
7.Συχνότητα κατανάλωσης γλυκών	1. Καθημερινά 2. 3-4 φορές τη βδομάδα 3. 1-2 φορές τη βδομάδα	☐
8.Συχνότητα κατανάλωσης αναψυκτικών	1. Καθημερινά 2. 3-4 φορές τη βδομάδα 3. 1-2 φορές τη βδομάδα 4. Σπάνια	☐
9.Χρόνος κατανάλωσης γλυκών/αναψυκτικών	1. Μετά το γεύμα 2. Μεταξύ των γευμάτων	☐
10.Πώς θα αξιολογούσατε την συνολική στοματική σας υγιεινή	1. Καλή 2. Μέτρια 3. Κακή	☐

Β. ΚΛΙΝΙΚΗ ΟΔΟΝΤΙΑΤΡΙΚΗ ΕΞΕΤΑΣΗ Ημερομηνία

<u>ΕΞΩΣΤΟΜΑΤΙΚΗ</u>

ΚροταφογναθικήΔιάρθωση:

- TMDPain screener

Ερώτηση 1. Κατά τις τελευταίες 30 ημέρες είχατε πόνο στη γνάθο και στην περιοχή του προσώπου και πόσο διήρκησε;

α. Καθόλου πόνο
β. Ο πόνος ήταν παροδικός
γ. Ο πόνος ήταν συνεχής ☐

Ερώτηση 2. Κατά τις τελευταίες 30 μέρες είχατε καθόλου πόνο ή ακαμψία στην περιοχή των γνάθων όταν ξυπνάγατε;

α. Ναι
β. Όχι ☐

Ερώτηση 3. Κατά τις τελευταίες 30 μέρες άλλαξε την ένταση ή την μορφή του πόνου η μάσηση σκληρών τροφών;

α. Ναι
β. Όχι ☐

Ερώτηση 4. Κατά τις τελευταίες 30 μέρες άλλαξε την ένταση ή την μορφή του πόνου η διάνοιξη του στόματος ή μετακίνηση των γνάθων μπροστά ή προς τα πλάγια;

α. Ναι
β. Όχι ☐

Ερώτηση 5. Κατά τις τελευταίες 30 μέρες άλλαξαν την ένταση ή την μορφή του πόνου δραστηριότητες όπως το τρίξιμο των δοντιών ή η μάσηση τσίχλας;

α. Ναι
β. Όχι ☐

Ερώτηση 6. Κατά τις τελευταίες 30 μέρες άλλαξαν την ένταση ή την μορφή του πόνου άλλες δραστηριότητες όπως ομιλία, χασμουρητό, κτλ.;

α. Ναι
β. Όχι ☐

α : 0 βαθμοί, β: 1 βαθμό, γ. 2 βαθμούς

ΣΚΟΡ ☐

- Παρουσία Ήχων 0. ΟΧΙ 1.ΝΑΙ ☐
- Παρέκκλιση από την μέση γραμμή
  - Απουσία
  - > 2 mm
  - …… mm ☐

ΕΝΔΟΣΤΟΜΑΤΙΚΗ Ημερομηνία

1. Μαλακοί Ιστοί

2. Στοματική Υγιεινή

OHI-s

DI–s

16 11 26

46 31 36

Καλυπτόμενη επιφάνεια μύλης

0 = Απουσία πλάκας
1 = $^1/_3$ ή λιγότερο
2 = $^1/_3$ έως $^2/_3$
3 = Περισσότερο από $^2/_3$
X = δεν καταγράφεται

CI-s

16 11 26

46 31 36

Καλυπτόμενη επιφάνεια μύλης

0 = Απουσία τρυγίας
1 = 1/3 ή λιγότερο
2 = 1/3 έως 2/3 ή υποουλική ή και τα δύο
3 = Περισσότερο από 2/3 ή παχιά συνεχής ζώνη υποουλικής ή και τα δύο
X = Δεν καταγράφεται

OHI-s=DI-s+CI-s=

0-1.2	***Good***
1.3-3	***Fair***
>3.1	***Poor***

3. Περιοδοντική Κατάσταση(CPI)

17/16 11 26/27

47/46 31 36/37

0 = Υγιές	3 = Θύλακος 4-5 mm
1 = Αιμορραγία	4 = Θύλακος ≥6 mm
2 = Τρυγία	x = Εκτημόριο με ένα ή κανένα δόντι

CPI=0	***Καμία ένδειξη νόσου***
CPI=1	***Αιμορραγία σε >1δόντια***
CPI=2	***Τρυγία σε >1 δόντια***

4. Οδοντική Κατάσταση

Ημερομηνία

Α.Δ.	ΤΕΡΗΔΟΝΑ					ΠΑΘΟΛΟΓΗ ΚΙΝΗΤΙΚΟΤΗΤΑ
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43/83						
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45/85						
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47						

ΤΕΡΗΔΟΝΑ- ICDAS

0 = Υγιής
1 = Μικρή και εντοπισμένη αλλαγή χρώματος αδαμαντίνης μετά από παρατεταμένο στέγνωμα (5 sec)
2 = Εκτεταμένη οπτική αλλαγή χρώματος σε υγρή επιφάνεια
3 = Λύση συνέχειας της αδαμαντίνης
4 = Γκρι αδαμαντίνη με ή χωρίς κοιλότητα
5 = Ανοιχτή κοιλότητα με εκτεθειμένη οδοντίνη
6 = Τερηδόνα με εκτεταμένη απώλεια αδαμαντίνης μασητικά ή όμορα
7= Έμφραξη χωρίς τερηδόνα
8= Προληπτική κάλυψη
9= Ανοξείδωτη στεφάνη
10- Εξαγωγή λόγω τερηδόνας
11= Λείπει για οποιαδήποτε άλλη αιτία εκτός τερηδόνας
12= Κάλυψη οπών και σχισμών

ΠΑΘΟΛΟΓΙΚΗ ΚΙΝΗΤΙΚΟΤΗΤΑ

1= Ναι
2= Όχι

ΑΛΛΑ

ΟΔΟΝΤΙΚΕΣ ΑΝΩΜΑΛΙΕΣ ΑΔΑΜΑΝΤΙΝΗΣ ΟΔΟΝΤΙΝΗΣ																	
Α.Δ.	**ΕΙΔΟΣ**					**ΟΔΟΝΤΙΚΗ ΑΠΟΤΡΙΒΗ**					**ΑΠΟΚΟΛΛΗΣΗ ΑΔΑΜΑΝΤΙΝΗΣ**					**ΧΡΩΜΑ**
	Ε	**Μ**	**Α**	**Π**	**Γ**	**Ε**	**Μ**	**Α**	**Π**	**Γ**	**Ε**	**Μ**	**Α**	**Π**	**Γ**	
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44/84																
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ΕΙΔΟΣ

1= Άσπρη αδιαφάνεια

2= Κίτρινη/καφέ αδιαφάνεια

3= Υποπλασία (οπές)

4= Υποπλασία (ράβδωση)

5= Υποπλασία (ποσοτική απώλεια αδαμαντίνης)

6= Άλλο

ΟΔΟΝΤΙΚΗ ΑΠΟΤΡΙΒΗ (ΔΕΙΚΤΗΣ BEWE)

1= Όχι διάβρωση

2= Αρχική απώλεια της φυσιολογικής υφής της οδοντικής επιφάνειας

3= Σαφής βλάβη, απώλεια σκληρών ιστών <50% της επιφάνειας

4= Απώλεια σκληρών ιστών ≥50% της επιφάνειας

ΑΠΟΚΟΛΛΗΣΗ ΑΔΑΜΑΝΤΙΝΗΣ

1= όχι φθορά σε οδοντίνη

2= οδοντίνη μόλις ορατή ή εκτεθειμένη

3= έκθεση οδοντίνης > 1/3 της επιφάνειας

4= έκθεση πολφού ή δευτερογενής οδοντίνη

Ημερομηνία

5. Ορθοδοντική Αξιολόγηση- Σύγκλειση

Οδοντοφυΐα 1. ΝΕΟΓΙΛΗ 2. ΜΙΚΤΗ 3. ΜΟΝΙΜΗ ☐

Ασθενής υπό ορθοδοντική θεραπεία 0. ΟΧΙ 1. ΝΑΙ ☐

Σταυροειδής σύγκλειση 0. ΟΧΙ 1. ΠΡΟΣΘΙΑ ☐

2. ΟΠΙΣΘΙΑ ΕΤΕΡΟΠΛΕΥΡΗ 3. ΟΠΙΣΘΙΑ ΑΜΦΟΤΕΡΟΠΛΕΥΡΗ

ΜΟΝΙΜΟΣ ΦΡΑΓΜΟΣ

Τάξη κατά Angle 1. Ιη τάξη 2. IIη τάξη/ υποκατηγορία 1

3.IIη τάξη/ υποκατηγορία 2 4. IIIη τάξη

ΔΕΞΙΑ ☐ ΑΡΙΣΤΕΡΑ ☐

ΝΕΟΓΙΛΟΣ ΦΡΑΓΜΟΣ

2ο νεογιλοί γομφίοι	**Δεξιά**	**Αριστερά**
1. Εγγύς σκαλοπάτι 2. Ίδιο κατακόρυφο επίπεδο 3. Άπω σκαλοπάτι		

Νεογιλός Φραγμός με διαστήματα	
Άνω	
Κάτω	
0. ΟΧΙ 1.ΝΑΙ	

Ημερομηνία

Ακολουθεί η αξιολόγηση ανάγκης ορθοδοντικής θεραπείας με τα κριτήρια της συνιστώσας οδοντικής υγείας του τροποποιημένου **Δείκτη Ανάγκης Ορθοδοντικής Θεραπείας**:

1. **ΣΤΑΥΡΟΕΙΔΗΣ ΣΥΓΚΛΕΙΣΗ (ΟΠΙΣΘΙΑ/ΠΡΟΣΘΙΑ) ΜΕ ΠΑΡΕΚΚΛΙΣΗ ΜΕΓΑΛΥΤΕΡΗ ΤΩΝ 2χιλ. ΜΕΤΑΞΥ ΚΕΝΤΡΙΚΗΣ ΣΧΕΣΗΣ-ΜΕΓΙΣΤΗΣ ΣΥΓΓΟΜΦΩΣΗΣ** (πλαγιολίσθηση, προολίσθηση)	**ΟΧΙ** □ **ΝΑΙ** □
2. **ΟΛΙΓΟΔΟΝΤΙΑ** (η οποία επιβάλλει είτε ορθοδοντική θεραπεία πριν την προσθετική αποκατάσταση είτε σύγκλειση των χώρων με ορθοδοντικές δυνάμεις)	**ΟΧΙ** □ **ΝΑΙ** □
3. **ΔΙΑΤΑΡΑΧΕΣ ΑΝΑΤΟΛΗΣ ΔΟΝΤΙΩΝ** (εξαιρουμένων των τρίτων γομφίων) λόγω συνωστισμού, κακής θέσης, παρουσίας υπεραρίθμων δοντιών, υπερπαραμονή νεογιλών ή οποιασδήποτε άλλης παθολογικής αιτίας	**ΟΧΙ** □ **ΝΑΙ** □
4. **ΟΡΙΖΟΝΤΙΑ ΠΡΟΤΑΞΗ** σε χιλ., με θετικό ή αρνητικό (αν παρατηρείται πρόσθια σταυροειδής) πρόσημο Σε **θετική οριζόντια πρόταξη,** είναι μεγαλύτερη από **6χιλ**.; Σε **αρνητική οριζ. πρόταξη**, ξεπερνάει τα -3,5χιλ; Σε **αρνητική οριζ. Πρόταξη** (από -1 έως -3,5χιλ), παρατηρούνται επιπτώσεις στη μάσηση στην ή στην ομιλία;	**_____χιλ.** **ΟΧΙ** □ **ΝΑΙ** □ **ΟΧΙ** □ **ΝΑΙ** □ **ΟΧΙ** □ **ΝΑΙ** □
5. ΚΑΤΑΚΟΡΥΦΗ ΠΡΟΤΑΞΗ σε χιλ., με αρνητικό πρόσημο όταν παρατηρείται πρόσθια χασμοδοντία **Σε αυξημένη κατακόρυφη πρόταξη**, παρατηρείται τραυματισμός περιοδοντίου υπερώια των άνω τομέων ή χειλικά των κάτω τομέων**;**	**_____χιλ.** **ΟΧΙ** □ **ΝΑΙ** □
6. Παρατηρείται **ΠΡΟΣΘΙΑ Η ΠΛΑΓΙΑ ΧΑΣΜΟΔΟΝΤΙΑ**, μεγαλύτερη των 4 χιλ.;	**ΟΧΙ** □ **ΝΑΙ** □
7. **ΠΑΡΑΤΗΡΕΙΤΑΙ ΣΕ ΚΑΠΟΙΟ ΔΟΝΤΙ ΑΝΩΜΑΛΙΑ ΘΕΣΗΣ** μεγαλύτερη των 4 χιλ.;	**ΟΧΙ** □ **ΝΑΙ** □
ΑΝΑΓΚΗ ΟΡΘΟΔΟΝΤΙΚΗΣ ΘΕΡΑΠΕΙΑΣ (σημειώνεται «ΝΑΙ» αν υπάρχει έστω και μία θετική απάντηση στα παραπάνω 7 κριτήρια)	**ΟΧΙ** □ **ΝΑΙ** □

Γ. ΑΚΤΙΝΟΓΡΑΦΙΚΟΣ ΕΛΕΓΧΟΣ

Α.Δ.	ΤΕΡΗΔΟΝΑ ICDAS 4-5					ΑΝΩΜΑΛΙΕΣ ΔΙΑΠΛΑΣΗΣ	ΔΟΚΙΜΑΣΙΑ ΖΩΤΙΚΟΤΗΤΑΣ ΠΟΛΦΟΥ (Μόνο στα δόντια με ενασβεστίωση μυλικού θαλάμου)
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ΑΝΩΜΑΛΙΕΣ ΔΙΑΠΛΑΣΗΣ

1. Αγενεσία

2. Μικροδοντία

3. Περίσφιξη σε αδαμαντινοοστεϊνική ένωση

4. Βολβοειδές σχήμα μύλης

5. Ενασβεστίωση μυλικού θαλάμου <50%

6. Ενασβεστίωση μυλικού θαλάμου ≥50%

7. Διαταραχή διάπλασης μήκους ρίζας

8. Διαταραχή διάπλασης πάχους ρίζας

9. Διαταραχή διάπλασης ακρορριζίου

10. Ακρορροζική αλλοίωση

11. Μεσορριζική αλλοίωση

12. Έγκλειστα δόντια

13. Ταυροδοντισμός

ΑΠΟΣΤΗΜΑΤΑ

0= ΟΧΙ
1= ΝΑΙ

ΑΛΛΑ