Oral Health Related Quality of Life in Children with Osteogenesis Imperfecta

Submitted by
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2017-2020

Declaration

I, Jasmine May Cachia Mintoff confirm that the work presented in this thesis is my own. Where information has been derived from other sources, I confirm that this has been indicated in the thesis.

Jasmine May Cachia Mintoff

Acknowledgments

I am very grateful to Dr Susan Parekh for all her help, support, encouragement and guidance during my time at the Eastman Dental Institute, without whose help I would have been unable to complete this project. I would also like to thank Dr Megan Riddington for her assistance throughout this experience especially whilst data collecting at Great Ormond Street Hospital. I would also like to thank Mr S Harden for his help with the statistics. I would particularly like to thank my parents for their constant support and encouragement during my studies and chosen career path, without them I would not have completed this project. Finally, I would like to thank all the participants, without whom this study would not have been possible.

Abstract

Background:

Osteogenesis Imperfecta (OI) is a genetic condition resulting from a mutation in the genes involved in the modification or biosynthesis of collagen. The study involved two parts and took place in the OI department at Great Ormond Street Hospital (GOSH).

Aims:

To investigate the oral health related quality of life of children aged eight to sixteen-years-old with OI.

Service Evaluation:

Methodology:

Five questions designed to assess if children with OI are receiving adequate dental care.

Results:

86 participants (43% female, 57% males, mean age 8.31)

78 (91%) of children were receiving some form of dental care.

49 (57%) of participants reported one or more dental concerns.

27/41 (66%) of the mixed dentition group reported at least one type of concern; most frequently appearance [18/41 (44%)].

Conclusions:

Dental concerns in children with OI are present.

It is important to have ongoing dental care to address concerns.

Questionnaire:

Methodology:

Ethical approval and consent were obtained

The Child Oral-Health Impact Profile – Short Form (COHIP-SF) was used, with demographic and qualitative data

Children aged 8 – 16 years participated between January – October 2019

Statistical analysis with parametric and non-parametric tests

Results

106 children participated (44 female, 62 male, mean age 11.93 years)

COHIP-SF Data

A higher COHIP-SF score indicates better OHRQoL (maximum score, 76).

The median score was 59. Children reporting mild OI had higher median score (62) than those reporting severe OI (55) [P=0.087]. When comparing mixed (<12 years, n=46) vs permanent dentition (>=12, n=60), no statistically significant difference in OHRQoL was seen [P= 0.977].

The 3 COHIP-SF domains are Oral Health, Functional and Socio-Emotional Well-Being. There was no significant difference between severities for each domain.

Perceived severity was not associated with significantly lower OHRQoL scores for females [P=0.125] or males [P=0.406].

Limited data on the presence of Dentinogenesis Imperfecta did not impact overall score [P=0.109] but was significant in the Oral Health domain [P=0.033].

Qualitative Data

Common themes were the need for braces (straightness/ gaps between teeth), discolouration, pain and function.

Conclusions:

This study confirmed that children with OI have dental concerns, including oral health, functional and socio-emotional well-being. This was related to severity of OI.

Compared to the general child population, children with OI had similar OHRQoL scores.

Impact Statement

Osteogenesis Imperfecta (OI) is a relatively common genetic condition characterised by bone fragility affecting many of the body's systems and can affect quality of life. As part of this condition, patients can also be affected by dentinogenesis imperfecta (DI) or malocclusion, particularly in the more severe types of OI. Initially, a service evaluation was carried out at the OI department at Great Ormond Street Hospital (GOSH). This service evaluation aimed to clarify what dental concerns children with OI are aware of in themselves. Oral Health Related Quality of Life (OHRQoL) is an important public health concern and there is currently limited evidence on how OI affects a child's OHRQoL. The second part of this study used a questionnaire aimed to address this gap.

The service evaluation assessed 86 children. It found that children with OI commonly have aesthetic dental concerns and have had trouble accessing primary dental care because their general dentist or local orthodontist was concerned about patient management given the complexities of their medical condition.

The second part of the study used the short form of the Child Oral Health Impact Profile (COHIP-SF) questionnaire, which was distributed at the weekly OI clinics at GOSH. The COHIP-SF questionnaire is divided into three domains assessing Oral Health well-being, Functional well-being and Socio-emotional well-being.

A total of 106 children completed the questionnaire. Results showed that OHRQoL scores are worse in children with severe OI than those with mild OI, however this was not significant. Overall, 50% of children felt they had discoloured teeth at least some of the time, and 65% felt they had crooked or spaced teeth at least some of the time. The presence of DI also has an effect on OHRQoL in the oral health well-being domain with those having DI having a significantly lower OHRQoL than those without DI.

The service evaluation indicated a need to reassess the pathway between the Eastman Dental Hospital (EDH) and GOSH for referral of OI patients and this is part of the action plan moving forwards. The OHRQoL section of this study will benefit the OI community as it disputes the assumption that clinical severity of OI is linked to OHRQoL and therefore overall quality of life. It shows that dental care should be tailored to each child on an individual basis depending on their oral condition. The results of the COHIP-SF reinforce that it is not only a child's general health but also their oral health which impacts their quality of life. Additionally, the need to educate general dentists and orthodontist on the importance of treating children with OI or referring to an appropriate specialist early on is clear. Education can be done by

presentation of results at conferences or organisation of lectures and information sheets through dental bodies such as the British society of Paediatric Dentistry or the Faculty of Dental Surgery. Further expansion of this to include other OI centres, both nationally and internationally would provide a more representative sample and ensure results are even more meaningful.

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List of Abbreviations

OI	Osteogenesis Imperfecta
OHRQoL	Oral Health Related Quality of Life
DI	Dentinogenesis Imperfecta
MRI	Magnetic Resonance Imaging
ONJ	Osteonecrosis of the Jaw
COHIP	Child Oral Health Impact Profile Questionnaire
COHIP-SF	Short form of Child Oral Health Impact Profile Questionnaire
POQL	Paediatric Oral Health Related Quality of Life Questionnaire
COIDP	Child Oral Health Impact on Daily Living Profile
SOHO-5	Scale of Oral Health Outcomes for 5 year old Children
PEDsQL	Paediatric Quality of Life Inventory
CPQ 11-14	Child Perception questionnaire for 11-14 year olds
MOHRQoL-C	Michigan Oral Health Related Quality of Life questionnaire for Children
ECOHIP	Early Child Oral Health Impact Profile Scale
WHO	World Health Organisation
YPAGroup	Young Persons Advisory Group
GOSH	Great Ormond Street Hospital for Children
EDH	Eastman Dental Hospital
SD	Standard Deviation
HRQoL	Health Related Quality of Life

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1. Literature Review

1.1 Introduction

Osteogenesis Imperfecta (OI) is a genetic condition resulting from a mutation in the genes involved in the modification or biosynthesis of collagen. OI can affect many of the body's systems including bones, teeth, eyes and joints (Van Dijk and Sillence, 2014). Oral Health Related Quality of life (OHRQoL) is an important part of general health care, and there is currently limited available literature about the oral health-related quality of life of children with OI. This oversight is what will be addressed by this study.

A questionnaire based research study identified the oral health-related quality of life of children with OI, in order to help us understand their specific concerns, and tailor our dental treatment to manage this more effectively.

1.2 Osteogenesis Imperfecta

Osteogenesis Imperfecta (OI), also known as 'brittle bone disease' (Forlino et al., 2011), is a genetic condition resulting from a mutation in the genes involved in the modification or formation of collagen (Byers, 1989). It affects Type I collagen, and can be associated with Dentinogenesis Imperfecta (DI), a genetic disorder of teeth (Okawa et al., 2017).

1.2.1 Prevalence

OI is a relatively common condition, and the literature cites the prevalence between 0.3-0.7 in 10,000 live births in the United States of America (Stevenson et al., 2012), 1.5 in 10,000 live births in the Danish population (Folkestad et al., 2016), and 0.5 in 10,000 live births in Finland (Kuurila et al., 2002). There is no available data on the prevalence of OI in the United Kingdom. There are several subtypes, and the prevalence of the different types of OI is also not available as many or the rarer types may only have a few known cases. The primary feature of all types of OI is varying degrees of bone fragility (Okawa et al., 2017). This is due to a low bone mass or reduced bone density. The fragility of the bones means they are more susceptible to fracture, deformity and deficient growth (Forlino et al., 2011). Other Clinical features include blue sclera, early hearing loss, DI, hypermobility of the joints, short stature and skeletal deformity (Van Dijk and Sillence, 2014), as shown in Figures 1.1, 1.2 and 1.3 and Table 1.1. These will be discussed further in section 1.2.6.

Figure 1-1 - Showing: A – Oblique mid-diaphyseal fibula fracture (open arrow) and a communitive diaphyseal tibia fracture (arrow); B – severe kyphoscoliosis with thoracal deformation, severe shortening and bowing of arms and legs

Images taken from Osteogenesis Imperfecta: A Review with Clinical Examples (Van Dijk et al., 2011).

Figure 1-2 - Showing blue sclera in OI

Image taken from Osteogenesis Imperfecta and Blue Sclera (Mitaka 2018)

Figure 1-3 - showing hypermobility of the finger joints

Image taken from Osteogenesis Imperfecta Type I: A Case Report (Ren et al., 2014)

1.2.2 Classification

OI was first classified in 1979 and included four subtypes of OI (Van Dijk and Sillence, 2014; Okawa et al., 2017). Later, other subtypes were added due to the clinical and radiographic variability seen in the types of OI (Van Dijk and Sillence, 2015). The current genetic classification, showing genetic variation, is shown in Table 4.1 (Marini et al., 2017). Using the old classification, type OI is I as the most common, followed by Types III and IV (Okawa et al., 2017).

Types I, III and IV can be further subdivided depending on whether they have DI or not (Okawa et al., 2017). In these types of OI, the presence of DI indicated an increased risk of bone fractures (Van Dijk and Sillence, 2014). In recent years the classification has been further subdivided to allow for genetic variations (Forlino et al., 2011).

The types of OI typically have been given severities based on clinical features. These usually are (Van Dijk et al., 2014):

- Type I – Mild
- Type II – Lethal
- Type III – Severe
- Type IV – Moderate
- Type V – Moderate
- Types VI-XX – Vary depending on clinical presentation however tend to be more severe (Marini et al., 2017, Moosa et al., 2019).

When classifying by genetics, one can divide OI into 20 main types. The severity of the OI varies across the types and can be either Autosomal Dominant (AD) or Autosomal Recessive (AR); an X-linked type has also been discovered. The four classical Sillence OI types were AD, and in all cases COL1A1 or COL1A2 genes were defective. Phenotypically they range from mild non-deforming (type I), moderately deforming (type IV), progressively deforming (type III) and lethal perinatal (type II). OI type V is the most common type of OI after the four classical Sillence types I-IV (Marini et al., 2017).

Although there is no data to support the prevalence of the different types of OI, approximately 77% – 85% of cases of OI are types I-IV (Marini et al., 2017; Moosa et al., 2019). Therefore, this paper will focus on the original Sillence classification of OI types I-IV.

1.2.3 Diagnosis

Diagnosis of OI is made based on several parameters. These include clinical evaluation of a child who has several bone fractures early in life, family history of OI, radiographic appearance and genetic testing (Marini et al., 2017). Bone deformity, bone mineral density scans and bone biochemistry should also be assessed when diagnosing OI (Palomo et al., 2017). Genetic assessment can be useful not only to aid diagnosis, but it can also give information about the risks of passing the condition on to offspring and identifying other family members with the same affected genes. Genetic testing is currently done by sequencing the DNA for specific target genes (Palomo et al., 2017).

1.2.3.1 Differential Diagnoses

Many childhood skeletal bone conditions could be part of the differential diagnosis of OI. These include Bruck syndrome, Osteoporosis-pseudoglioma syndrome (Glorieux, 2008; Van Dijk et al., 2011), Juvenile Paget's disease (Glorieux, 2008), Cole-Carpenter syndrome, Hajdu-Cheney, Gerodermia osteodysplasia and idiopathic juvenile osteoporosis (Van Dijk et al., 2011). Child abuse may also be mistaken for OI without a family history of OI (Glorieux, 2008; Van Dijk et al., 2011). Diagnosis can be determined by proper history taking and clinical examination. Assessing for blue sclera, dentinogenesis imperfecta, short stature can help come to the diagnosis of OI, whilst signs of progressive skeletal deformity, joint contractures cerebral calcifications or premature loss of teeth will exclude this diagnosis (Byers et al., 2006). This can be of significance in the milder types of OI given the relative rarity of OI when compared to child abuse. If incorrectly diagnosed, there may be long standing consequences for the families involved and even a lasting distrust of healthcare professionals (Singh et al., 2011). According to a study by Marlowe et al., approximately 2-5% of children suspected of child abuse are misdiagnosed cases of children with OI (Marlowe et al., 2002).

1.2.4 Genetics

Generally, around 90% of the primary responsibility for the changes occurring in OI are due to mutations in genes COL1A1 and COL1A2 in OI types I-IV, (Marini et al., 2017; Jain et al.,

2019). However, another study by Bardai et al. assessed 598 individuals with OI; by sequence analysis of the DNA; and quoted the frequency of these genes as lower, at 77%. The second most commonly found gene for OI was IFITM5 causing type V OI (Bardai et al., 2016).

COL1A1 and COL1A2 are the genes that code for the production of type I collagen (Van Dijk and Sillence, 2014). Specifically, the α-chains, α1 (I) and α2 (I), forming type I collagen are encoded by these genes. The type I collagen molecule is made up of 3 polypeptide chains, two proα1 (I) and one proα2 (I) chains. These form the procollagen heterotrimer which in turn forms Type I collagen (Forlino et al., 2011). Correctly entwined chains have a glycerine amino acid at every third position. Figure 1.4 shows a schematic representation of the COL1A1 and COL1A2 genes and normal versus mutant type I collagen.

When structure or quality of Type I collagen is altered due to mutations in OI, it can cause a range of phenotypes from subclinical to lethal (Forlino et al., 2011). Quantitative defects cause a milder type of OI, whilst structural defects are responsible for the more severe types of OI (Marini et al., 2017). In OI, the glycine amino acids are commonly affected by point mutations (Glorieux, 2008; Palomo et al., 2016b), creating a blend of abnormal and normal collagen. This produces a range of phenotypes which are difficult to predict, depending on which alpha chain is affected, the position of the glycine affected and where the substitution occurs. Another type of mutation is when a stop codon is created within the COL1A1 gene before it is necessary, these phenotypes are easier to predict in and most mutations result in type I OI (Glorieux, 2008).

It is important to bear in mind that the genetics of OI types V-XVIII are distinctly different from types I-IV, as different genetic mutations are present. In addition to this, the genes involved encode for different proteins, which have only recently been identified as necessary for the development of bone (Marini et al., 2017).

OI type V is of AD inheritance and the IFITM5 gene is affected. Phenotypically it shows distinct histology of irregularly arranged lamellae (Glorieux et al., 2000; Marini et al., 2017). Type VI OI also shows distinct histology with the normal birefringent pattern of the lamellae missing (Glorieux et al., 2002; Marini et al., 2017). However, it is AR and has a mineralization defect due to a mutation in the SERPINF1 gene. OI types VII, VIII and IX are of recessive inheritance and the defective genes cause 3-hydroxylation defects. The phenotype ranges

from moderate to lethal and the genes affected are CRTAP, LEPRE1 and PPIB. OI type X has a defect in the SERPINH1 gene and causes a severe type of OI, while type XI has a defect in the FKBP10 gene and is progressively deforming. OI type XII is a severe phenotype and shows a high bone mass. The gene encoding for it is the BMP1 gene. The gene defect causes C-Propeptide cleavage. PLOD2 mutations also cause a type of moderate to severe OI with progressive joint contractures however the type has not yet been specified. Types X, XI, VII and XII are all AR and are due to compromised collagen processing and cross-linking. Types XIII to XVIII are all due to altered osteoblast differentiation and function and are encoded for by the genes SP7, TMEM38B, WNT1, CREB3L1, SPARC and MBTPS2. The mode of inheritance varies from AR in the severe types XIII to XVII, an additional AD form of type XV and an X-linked recessive inheritance for type XVIII which is a moderate to severe type of OI (Marini et al., 2017). Type XX is AR and progressively deforming. It is likely due to hypomorphic alleles of the MESD gene (Moosa et al., 2019).

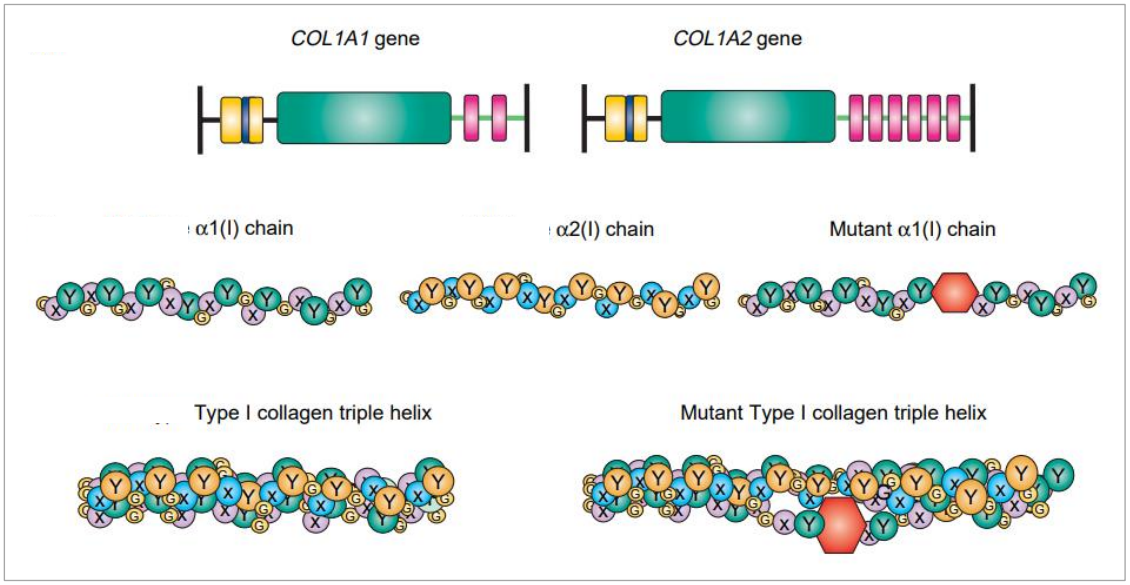

Figure 1-4 - Schematic representation of the COL1A1 and COL1A2 genes and normal versus mutant type I collagen.

Adapted from emerging therapeutic approaches for osteogenesis imperfecta (Millington-Ward, McMahon, & Farrar, 2005)

Table 1-1 - Showing genetic classification, adapted from Marini et al., 2017 and Moosa et al., 2019.

Mutated gene	Encoded protein	OI type based on the genetic classification	Inheritance	OMIM	Clinical characteristics	DI
Impairment of collagen synthesis and structure						
COL1A1 or COL1A2	Collagen α1(I) (COL1A1) or α2(I) (COL1A2)	I	AD	166200	Characterized by blue sclera, near-normal stature and late-onset hearing loss	Yes
COL1A1 or COL1A2	Collagen α1(I) (COL1A1) or α2(I) (COL1A2)	II	AD	166210	Perinatal lethal form	No
COL1A1 or COL1A2	Collagen α1(I) (COL1A1) or α2(I) (COL1A2)	III	AD	259420	Progressive deforming variety, blue sclera at birth but normalises, dentinogenesis imperfecta	Yes
COL1A1 or COL1A2	Collagen α1(I) (COL1A1) or α2(I) (COL1A2)	IV	AD	166220	Characterized by white sclera, short stature, bone deformity and dentinogenesis imperfecta, which is more severe than type I but less severe than type II and type III	Yes

Compromised bone mineralization						
IFITM5	Bone-restricted interferon-induced transmembrane protein-like protein (BRIL; also known as IFM5)	V	AD	610967	Normal-to-severe skeletal deformity, intraosseous membrane ossifications, radio dense band and radial head dislocation, normal-to-blue sclera and sometimes hearing loss	No
SERPINF1	Pigment epithelium-derived factor (PEDF)	VI	AR	613982	Moderate-to-severe skeletal deformity, the presence of osteoid, fish-scale appearance of lamellar bone pattern and childhood onset	No

Abnormal collagen post-translational modification						
CRTAP	Cartilage-associated protein (CRTAP)	VII	AR	610682	Severe rhizomelia (disproportion of the length of the proximal limbs) with white sclera	No
P3H1 (previously known as LEPRE1)	Prolyl 3-hydroxylase 1 (P3H1)	VIII	AR	610915	Severe rhizomelia with white sclera	No
PPIB	Peptidyl-prolyl cis–trans isomerase B (PPease B)	IX	AR	259440	Severe bone deformity with grey sclera	Yes
Compromised collagen processing and crosslinking						
SERPINH1	Serpin H1 (also known as HSP47)	X	AR	613848	Severe skeletal deformity, blue sclera, dentinogenesis imperfecta, skin abnormalities and inguinal hernia	Yes
FKBP10	65 kDa FK506-binding protein (FKBP65)	XI	AR	610968	Mild-to-severe skeletal deformity, normal to-grey sclera and congenital contractures	No
PLOD2	Lysyl hydroxylase 2 (LH2)	XII; Also know as Bruck's Syndrome 2 and Osteogenesis	AR	609220	Moderate-to-severe skeletal deformities and progressive joint contractures	No

		Imperfecta with Congenital Joint Contractures				
BMP1	Bone morphogenetic protein 1 (BMP1)	XIII	AR	614856	Mild-to-severe skeletal deformity and umbilical hernia	No

Altered osteoblast differentiation and function						
SP7	Transcription factor SP7 (also known as osterix)	XII	AR	613849	Severe skeletal deformity with delayed tooth eruption and facial hypoplasia	No
TMEM38B	Trimeric intracellular cation channel type B (TRIC-B; also known as TM38B)	XIV	AR	615066	Severe bone deformity with normal-to-blue sclera	No
WNT1	Proto-oncogene Wnt-1 (WNT1)	XV	AR AD	615220 Unknown	Severe skeletal abnormalities, white sclera and possible neurological defects	No
CREB3L1	Old astrocyte specifically induced substance (OASIS; also known as CR3L1)	XVI	AR	616229	Severe bone deformities	No
SPARC	SPARC (also known as osteonectin)	XVII	AR	616507	Progressive severe bone fragility	No
MBTPS2	Membrane-bound transcription factor site-2 protease (S2P)	XVIII	XR	301014	Moderate-to-severe skeletal deformity, light blue sclera, scoliosis and pectoral deformities	No

Truncating and frameshift mutation						
MESD	Mesoderm development gene, previously called MESDC2	XX	AR	607783	Progressively deforming type seen with parental consanguinity. Bisphosphonate treatment appears to have limited affect. Osteopenia, skeletal deformity, blue sclera, hypodontia	No

1.2.5 Bone fragility

Bone fragility is the most important clinical feature in OI. Typically fractures occur with little or no trauma due to the extreme fragility of the bones, particularly in the more severe types of OI. Children with mild OI have fewer fractures than those with more severe types of OI (Brizola et al., 2017). Fractures occur due to a reduced bone density or bone mass and abnormalities in the material properties of the bone (Marini et al., 2017). Bone turnover is anomalous in these patients as bone resorption is greater than bone formation (Van Dijk and Sillence, 2014). In healthy bone, 65-70% of the bone mass comes from minerals. The rest of the bone mass is water and proteins. The majority of OI cases appear to have normal mineralisation processes and functioning. However, in OI, the collagen content can be reduced by 20% (Bishop, 2016).

The variation in bone composition compared to normal bone is similar across the types of OI. Histologically, a reduced volume of cortical bone and reduced cortical width are seen in OI types I-IV, VII and VIII (Marini et al., 2017). The bones in OI are characterised mainly by their brittleness which is due to increased mineralisation density and an increased number of non-enzymatic cross-links. The loose bone matrix and abnormal bone architecture can also lead to increased fragility (Bishop, 2016). This increased mineral content is not affected by bisphosphonate treatment. As the particle size of the minerals are unchanged, the extracellular space available for water molecules between the collagen particles is less than in normal bone. Combined with altered cross linkages of the collagen, a stiffer, more brittle bone is seen in OI. Finally, due to the increased brittleness of the bone, energy is absorbed and dissipated less efficiently, increasing the risk of fracture due to its lack of elasticity (Marini et al., 2017).

The implications of bone fragility are that persons with OI are more likely to suffer bone fractures. Whilst fractures can occur at any stage of a person's life, they are most common in children. It seems that the more severe the type of OI the younger the person will be at first fracture (Brizola et al., 2017).

1.2.6 Further features of OI

1.2.6.1 Hearing Loss

As previously mentioned, hearing loss can be a common feature in OI. It can be seen across all types of OI and, tends to be a progressive due to sensorineural and conductive deficiencies (Forlino et al., 2011). The condition often resembles otosclerosis and

abnormalities of aural bone structure are often seen, including deformities of the temporal bones, fractures of the ossicles in the middle ear and abnormalities of the labyrinth (Hald et al., 2018).

The affects often start later in life typically by the fourth decade (Hald et al., 2018), and by 50 years old approximately 50% of OI patients have hearing loss according to a Scottish study (Forlino et al., 2011). Results from Finland are similar giving a 52% prevalence of hearing loss, with type I OI more likely to suffer from hearing loss than those with types III and IV (Kuurila et al., 2002). However, a study by Hald et al in 2018 shows that adults with type III OI always have hearing loss, whilst those with type I OI are more likely to suffer from loss of hearing than those with type IV (Hald et al., 2018). The study by Hald et al. boasts a slightly larger sample size of OI types III and IV and may therefore show the more likely trend.

In adults, sensorineural hearing loss is most common however many suffer from a mixed type of hearing loss as conductive hearing loss may begin much earlier (Hald et al., 2018). Approximately 5% of children with OI also show signs of conductive hearing loss (Hald et al., 2018). Treatment is typically with the use of hearing aid, however surgery may be indicated in severe cases (Forlino et al., 2011).

1.2.6.2 Eyes

Disorders of type I collagen can also affect the eyes, the staple ophthalmic sign of OI is blue sclera which results from the abnormal way the light reflects off the collagen in the sclera (Pillion et al., 2011). Collagen is also the most prevalent protein in the cornea and uveal tissues, therefore defects such as reduced corneal thickness, myopia and astigmatism may be seen in OI (Hald et al., 2018). Retinal detachment can also occur in patients with OI and may cause additional morbidity due to the increased risk of accidents resulting in trauma. Thus, regular ophthalmic review is recommended in all persons with OI (Paolo et al., 2018).

1.2.6.3 Neurological features

Certain neurological features may also be linked to OI, including macrocephaly; which is relatively common; syringomyelia, basilar invagination and hydrocephalus (Forlino et al., 2011; Brizola et al., 2017). The most significant of these is basilar invagination which can lead to brainstem distortion due to an infolding of the skull base (Forlino et al., 2011), this can be seen in figure 1.5. The current hypothesis is that the skull base settles over

the cervical spine due to the softness of the bone, resulting in this complication. The severity of the invagination is assessed based on the relationship between the foramen magnum and the Tip of Dens (Castelein et al., 2019).

Although it is a rare complication, due to the severity of the consequences, it is regularly monitored by Magnetic Resonance Imaging (MRI) in children with OI, and early intervention is important to delay progression. Occipitocervical bracing is the management method of choice. Serious cases may warrant surgery, however 80% will once again start to progress in the first six years (Forlino et al., 2011).

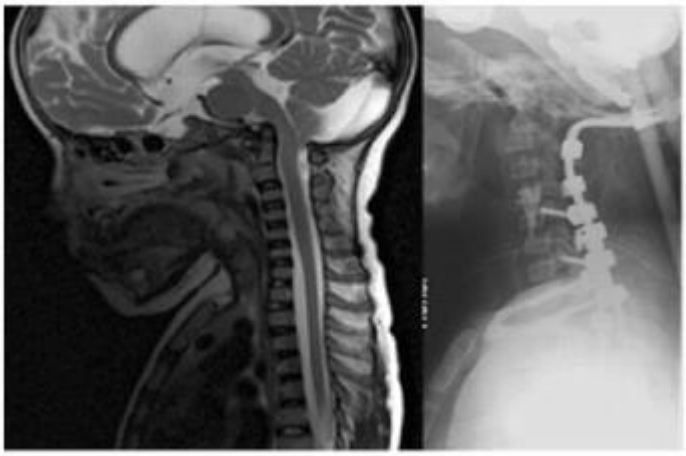

Figure 1-5- Showing a 6-year-old boy with basilar invagination, before and after treatment

Image taken from Complex spine deformities in young patients with severe osteogenesis imperfecta: current concepts review (Castelein et al., 2019)

1.2.6.4 Growth and the skull

Many people with OI also have stunted growth resulting in the typical short stature, considered a hallmark of the more severe types of OI, most especially in types IV and III (Jain et al., 2019). Despite this, endocrine analysis is usually normal in these children (Forlino et al., 2011). Scoliosis, vertebral and long-bone fractures, kyphosis and bone deformities can all be causes for short stature in OI. However, lack of height is seen also in cases without significant bone deformities; reasons for this could be that the matrix and cellular abnormalities associated with OI affect final height, as well as reduced responsiveness to growth hormone (Jain et al., 2019).

Wormian bones are another typical feature of OI, typically diagnosed from routine skull radiographs (Dahan, 2016). These can be seen in figure 1.6 below. They are present in approximately 30% of children with OI. They are found between the fontanelles and sutures of the skull and are classified as supernumerary bones. Although they are

generally associated with OI and other bony disorders, wormian bones may also be present in the general population (Brizola et al., 2017).

Figure 1-6 – Showing Wormian bones in the skull of a four year old girl with OI.

Image taken from Wormian Bones in Osteogenesis Imperfecta: Correlation to Clinical Findings and Genotype. (Selmer et al., 2010)

1.2.6.5 Scoliosis

Scoliosis is a spinal deformity commonly found across all types of OI. Different papers quote different results for prevalence of scoliosis in OI. According to Liu et al., it may vary from 26% - 74.5% (Liu et al., 2017); whilst Castelein et al quote the prevalence at 25% in five year olds, increasing to up to 80% in adolescents (Castelein et al., 2019). The spinal curvature of the scoliosis also varies widely, from 7˚ to 105˚, and 73.7% of scoliosis cases are mild and are usually curvatures of under 40˚. This spinal deformation, although rare before 6 years old, will progress rapidly after this age or after the spinal curvature is larger than 50˚ (Liu et al., 2017). Risk factors for spinal deformity include severity and type of OI, motor milestone achievements, vertebral fractures and deformities, ligamentous laxity and weakness of the muscles. These all lead to a cycle of increasing scoliosis and inhibited vertebral growth (Castelein et al., 2019).

In OI type I, when scoliosis develops it is usually idiopathic in nature. In types III and IV OI, progressive scoliosis is common, it begins in childhood and continues through to adolescence. In the past, scoliosis could be a cause of death in these patients, secondary to chest wall deformity (Van Dijk and Sillence, 2014). Scoliosis could also lead to pulmonary insufficiency (Forlino et al., 2011; Sato et al., 2016). The implications are that early surgical treatment is necessary when the curvature of the back is more than

60˚. Scoliosis in OI patients does not appear to be related to vertebral compressions fractures (Forlino et al., 2011).

Bisphosphonates do not reduce the incidence of scoliosis but may have a limited effect on the progression of the spinal curvature. Although conventional scoliosis may be treated with a brace, there is some evidence that in OI, this may affect the chest wall morphology and pulmonary function (Castelein et al., 2019).

1.2.6.6 Respiratory and Cardiac

Although management and treatment of children with OI has improved significantly in recent years, respiratory or cardiovascular concerns care still common causes of mortality in OI. Children with severe OI can also suffer from recurrent pneumonia and this is thought to be secondary to scoliosis, rib fractures or other skeletal anomalies. With a spinal curvature of over 60˚, a serious decline in pulmonary function is noted. This is particularly common in OI types III and IV. Eventually, respiratory issues can lead to *cor pulmonale*. Some cardiac anomalies are also present however these are more prevalent in adults. These can include valvular insufficiency, atrial septal defects, ventricular wall thickening and aortic root dilatation. The latter being the most common cardiac manifestation. There is speculation that the changes are due to an abnormally stiff myocardial tissue caused by the collagen mutation (Forlino et al., 2011).

1.2.6.7 Joint laxity

Although joint laxity or hypermobility is a common feature in OI, the range of motion of the joints varies between types of OI. Generalised joint hypermobility is common in OI type I, contrary to OI types III and IV which show reduced range of movement (Engelbert et al., 1998). Studies show that the increased range of motion seen in type I OI decreases over time (Forlino et al., 2011). The hypermobility seen in type I OI can lead to unstable knees and feet due to knee hyperextension, flat feet and hip extra-rotation (Monti et al., 2010).

1.2.6.8 Obesity

Children with OI may also be prone to a higher body weight than their healthy peers. Although there is no pathological reason for this, a study on the nutritional needs of children with OI quote that 31% of OI children had a high percentage of body fat compared to non-OI children (14%). Children with OI type III are more prone to obesity

(83%) than children with type I OI (42%) (Chagas et al., 2012). This is also seen by Jain et al where BMI in children with type III OI is higher than in children with types I and IV OI (Jain et al., 2019). The implications of a high fat percentage in children with OI are that, apart from the general issues of being overweight seen in the general population, they have increased risk of reduced motor function and thus consequences on rehabilitation after fractures (Palomo et al., 2016b).

1.2.6.9 Fatigue

Fatigue is a well-known symptom in OI (Monti et al., 2010; Van Dijk and Sillence, 2014; Arponen et al., 2018), which may be related their hypermobility. Chronic pain causing disturbed sleep may also contribute to this (Marr et al., 2017). A study by Arponen et al found that 96% of adults with OI report fatigue and this is higher than the average for the general population. They also found a positive correlation between pain episodes and fatigue (Arponen et al., 2018). Van Brussel et al found that children with OI report less subjective fatigue after exercising to improve muscle force, however the level of exercise should be ideal for the patient and maintained for long-standing effects (Van Brussel et al., 2008).

1.2.7 Management of OI

Currently, there is no known cure for OI. Management is based on age, severity of the OI, functional status and treatment of symptoms (Palomo et al., 2017; Rousseau et al., 2018). Treatment is based on pre-emptively strengthening functional capabilities and physiotherapy to maximise mobility. Physiotherapy is also given post-orthopaedic surgery together with rehabilitation treatment when surgery becomes necessary (Glorieux, 2008). Occupational therapists also play a role in management of OI to promote self-care skills and help with overcoming their functional limitations such as low muscle strength and range of motion (Montpetit et al., 2015). If a child only has a mild case of OI, restrictions such as avoiding contact sports, regular physiotherapy and management of fractures is usually sufficient. The more severe types of OI require early orthopaedic interventions to improve mobility, bone deformity and scoliosis (Palomo et al., 2017).

Many children with OI start treatment early on in life. The standard management is intravenous bisphosphonates. This medication is use to help increase bone density and reduce the chance of fractures (Dahan, 2016).

1.2.7.1 Bisphosphonate treatment

Bisphosphonates work by reducing osteoclastic activity which thus affects how the bone repairs and maintains itself, as well as how it remodels (Rousseau et al., 2018). In the body, pyrophosphate binds to hydroxyapatite in bones, bisphosphonates are synthetic analogues of these chemicals (Zeitlin et al., 2003). The bone resorption is inhibited as these drugs draw osteoclasts into the bone and thus reduce their activity. It also causes a reduction in the osteoclast precursor formation (Christou et al., 2013). The bone mass of children receiving bisphosphonate treatment will increase due to the relative rise in the activity of osteoblasts (Seikaly et al., 2005). The treatment also results in reshaping of vertebral bodies and a reduction in fracture rates (Montpetit et al., 2015).

Bisphosphonate treatment for children with OI was first introduced in 1987. Since then its use has increased exponentially. These drugs can be administered orally or intravenously and work by inhibiting osteoclastic function in the bones. This decrease in bone resorbing activity helps to increase bone density (Glorieux, 2008). The bisphosphonates used for treatment are generally the nitrogen-containing bisphosphonates and these inhibit the mevalonate pathway (Christou et al., 2013).

Mainstay treatment with bisphosphonates involves cyclical treatments of pamidronate every one to four months by intravenous infusion. This is the best studied protocol and the dose is administered at 1mg per Kg body weight per day over three days, with a treatment time of four hours each day (Palomo et al., 2016a). This treatment leads to a reduction in chronic bone pain and an increase in vertebral bone mineral mass. Despite worries to the contrary in the early days of this treatment, there was been no evidence of reduced growth in children with moderate to severe OI (Glorieux, 2008). Treatment is extended over several hours and several days to minimise the effects of renal toxicity leading to glomerular and tubular injury. This is commonly seen in the treatment of oncology patients with a higher dose of the pamidronate infusion. Incidence of renal toxicity in adult patients has been quoted at 9.3% for pamidronate (Palomo et al., 2016a).

Other bisphosphonates used include zoledronate infusions (Montpetit et al., 2015) and oral bisphosphonates such as alendronate and risedronate (Ward and Rauch, 2013). Bisphosphonate treatment can be started early in life and is often continued through puberty until the patient has reached maturity. Although bisphosphonate treatment for adults with OI is not as effective as in a growing child, some benefits may still be seen (Glorieux, 2008). Temporary symptoms of muscle pain, malaise, diarrhoea, fever,

nausea and bone pain may be evident the first three days after an intravenous infusion of bisphosphonates and are commonly only associated with the first dose (Christou et al., 2013).

New protocols are under investigation have been found where the dose was increased to a single dose of pamidronate to 2mg per Kg body weight over two hours up to a maximum of 60mg every four months, with similar effects on bone density and safety as traditional protocols. This protocol will lessen the burden on families who need to attend for regular infusions of bisphosphonates (Palomo et al., 2016a).

1.2.7.1.1 Osteonecrosis of the jaw

The use of bisphosphonates in adults, for other conditions, has been associated with osteonecrosis of the jaw (ONJ) following dental treatment. This condition has typically been seen in persons with a history of malignancy, generally over 60 years of age (Maines et al., 2012). According to the American College of Rheumatology, ONJ 'occurs when the jawbone is exposed and begins to starve from a lack of blood. Most cases of ONJ happen after a dental extraction (Bolster, 2017). In cases where bisphosphonate related ONJ has been reported, the high dose of bisphosphonate administered caused a reduction in blood supply to the wound (Rousseau et al., 2018). Although cases of osteonecrosis of long bones have been reported in children undergoing chemotherapy of leukaemia, no cases have ever been reported in the jaw following bisphosphonate treatment in children (Maines et al., 2012).

Osteo means bone and necrosis means death. As the name indicates, the bone begins to weaken and die with ONJ which usually (but not always), causes pain'(Bolster, 2017). However, current studies have shown that there have been no cases of ONJ in children on bisphosphonate treatment (Christou et al., 2013). There is however the possibility that this treatment can cause problems over time and so it is important that children with OI have good dental health to try and avoid these possible complications (Maines et al., 2012). In addition, some types of OI are associated with DI, which can also significantly impact oral health.

1.2.7.2 Orthopaedic treatment

In severe cases of OI, rodding of the long bones is carried out to help correct deformities. This is usually done when a child first attempts to stand but prior to walking. The rods also serve as preventive protection of the legs once the child starts walking (Zeitlin et al.,

2003). The rods serve to straighten bowed long bones. Repeated fracture can cause deformities and long periods of immobilisation, requiring further functional rehabilitation (Palomo et al., 2017). Other surgically placed aids for straightening deformed bones such as plates and screws should not be used in children with OI as they can cause additional stress on the bones and further increase risk of fractures. The two types of rods used are rigid or telescopic rods. Telescopic rods are a controversial topic and may result in repeat surgeries if they do not elongate as required. They may also cause impaction of the growth plate. Risks with regular rods include fracturing of the rods or bent nail screws which may also require a repeat surgery to correct (Marini et al., 2017).

The upper limbs may also require rodding, but this is only considered in extreme cases where there is severe functional disability or repeated fractures (Zeitlin et al., 2003). However, this form of treatment is becoming more popular as the awareness of the impact of upper limb deformities on self-care is now recognised. Again, in these cases conventional or telescopic rods can be used, however the use of elastic rods of Kirschner wires are also common treatment options (Marini et al., 2017). Corrective surgery of the spine in cases with progressive scoliosis is also sometimes indicated (Zeitlin et al., 2003). This is frequently carried out close to puberty and in cases where the Cobb angle has progressed beyond 40-50 ˚. In these cases, progression is likely to continue, and specialist surgical management is indicated. Surgical management can involve spinal fusion with or without Harrington rod placement, segmental pedicle screw fixation and HALO-gravity treatment.

1.2.7.3 Future therapies

Other possible future therapies for children with OI are transplantation of bone marrow stromal cells or gene therapy (Glorieux, 2008). Bone marrow transplants using multipotent cells in mice receiving human stem cells has been attempted and showed improved bone mechanics. The next stage will be *in vivo* trials to assess the safety of this treatment in children with OI (Marini et al., 2017). Whilst gene therapy is the most desirable form of treatment as it can potentially cure this condition, current research cannot as yet achieve this (Glorieux, 2008). Gene therapy would probably involve allele replacement or silencing to cure the condition (Maines et al., 2012). Other options may include anti-sclerotin therapy which promotes the recruitment and activity of osteoblasts. There have been some promising results in studies on mice (Marini et al., 2017).

1.3 Dentinogenesis Imperfecta

Approximately 50% of children with OI have dental anomalies (Patel et al., 2015); the most common of which is Dentinogenesis Imperfecta (Rousseau et al., 2018).

Dentinogenesis Imperfecta (DI) is a hereditary disorder of dentine. The incidence has been reported at 1 in 8000 and classification is based on the Shields Criteria (Clark et al., 2019). There are three types of DI; Type I is associated with OI and the COL1A1 or COL1A2 genes. DI types II and III are due to mutations of the dentine sialophosphoprotein gene and contrary to type I DI are generally not related to any syndromes. (Teixeira et al., 2008; Clark et al., 2019). Type III DI was first found in Maryland and Washington DC in a tri-racial isolate population as is thought to be a variation of type II DI (Takagi and Sasaki, 1988). Types I, III and IV OI commonly have DI (Rios et al., 2005). Dentine, like bone is formed by the secretion of an extracellular matrix by odontoblast or osteoblast cells and subsequent deposition of minerals. Ninety percent of the extracellular matrix is type I collagen (Opsahl Vital et al., 2012; de La Dure-Molla et al., 2014). Carbonate-substituted hydroxyapatite crystals make up the mineral content of both bone and dentine, this is progressively built up into fibrils assuming the final matrix framework (Opsahl Vital et al., 2012).

The severity of DI associated with OI varies. Prevalence of DI varies with OI type across the literature, from 21% to 73% and is most commonly found in OI types III and IV (Rousseau et al., 2018). Teixeira et al, 2008 report that DI is seen in approximately half of OI cases (Teixeira et al., 2008). According to Marçal et al, 2019, more severe cases of OI are more likely to have DI than the milder cases (Marçal et al., 2019). Although DI can be found in both the primary and secondary dentition, there are variations in prevalence and severity between the dentitions. DI is more severe in the primary than the secondary dentition in children with OI and DI (Teixeira et al., 2008; Clark et al., 2019). As the severity of DI tends to be less in the permanent dentition, it may often go unnoticed. It is also important to keep in mind that in patients diagnosed with DI, an unreported, underlying diagnosis of OI should be considered (Teixeira et al., 2008).

Teeth affected by DI have several characteristic features; most significantly a greyish discolouration due to incomplete development of the dentine. This gives them an opalescent appearance (figure 1.7) (O'Connell and Marini, 1999; Patel et al., 2015). The discolouration, although typically of grey hue can vary from shades of yellow to blue and brown (Rios et al., 2005).

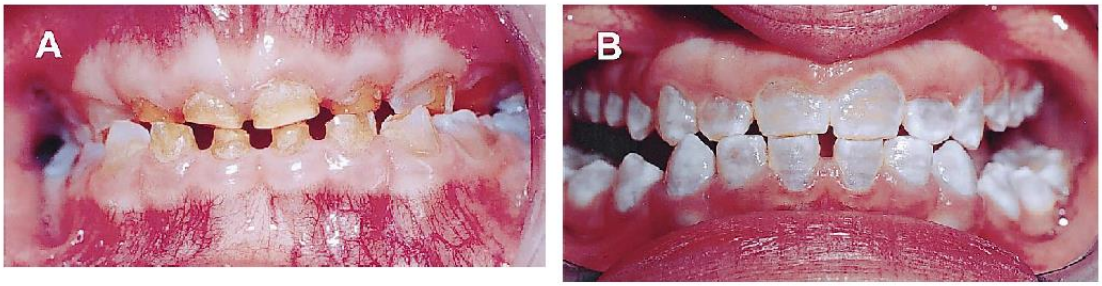

Figure 1-7 - A, Primary dentition affected by yellow/brown DI shows attrition and enamel fractures. B, Permanent dentition with yellow/brown DI shows discoloration throughout crowns of all teeth. Bilateral posterior open bite is present, and occlusion is only on anterior teeth, as shown.
Images taken from Evaluation of oral problems in an osteogenesis imperfecta population (O'Connell & Marini, 1999)

The crowns are often bulbous in shape with a well-defined constriction at the cementoenamel junction (O'Connell and Marini, 1999; Patel et al., 2015). In the primary dentition, the pulp chambers are originally quite large but will calcify early (Rousseau et al., 2018). Narrow roots with obliterated pulp chambers and root canals can also be seen radiographically, as shown in figure 1.8 (O'Connell and Marini, 1999; Teixeira et al., 2008). The thin roots can cause an increased incidence of root fracture during root canal treatment and the pulpal obliteration makes root canal therapy more difficult to carry out (Okawa et al., 2017).

Figure 1-8 - Panoramic radiograph of 14-year-old patient shows classic radiographic features of DI (bulbous crowns, pulpal obliteration, and short roots). In addition, impaction of both upper second molars is evident.

Images taken from Evaluation of oral problems in an osteogenesis imperfecta population (O'Connell &

Small scales studies seem to indicate that teeth affected by DI do not have an increased incidence of caries. It seems that the carious lesion progresses slowly because of the unusual structure, and reduced number of the dentinal tubules (Rousseau et al., 2018; Clark et al., 2019). However, a recent study looking at 319 children with OI, disagrees with this and found that the presence of DI increases the risk of developing caries (Ma

et al., 2019). This study is well designed and has much a much larger sample size than previous studies, indicating this is most likely to be true. Caries may also be detected later as pain is a late symptom due to pulp canal obliteration following secondary dentine deposition (Clark et al., 2019).

Due to the developmental defect, the enamel-dentine bond is poor and thus often causes flaking away of the enamel and subsequent tooth wear (O'Connell and Marini, 1999; Okawa et al., 2017) as seen in Figure 1.7A. This exposes the defective dentine leading to further tooth wear (Clark et al., 2019). Vulnerability to breakdown varies but tends to be worse in the primary dentition. It is important to bear in mind that the enamel of teeth affected by DI is normal (Rousseau et al., 2018). The pattern of enamel chipping is incisal in the anterior teeth and occlusal in the posterior teeth, with buccal and lingual wear also seen in all teeth. The underlying dentine is soft and thus the subsequent tooth wear occurs. In the primary dentition this can be so severe that the occlusal surfaces of the teeth become flush with the gingiva as seen in figure 1.7A (Rios et al., 2005).

The discolouration of dentine results in an opalescent grey-brown appearance of the teeth, and patients with DI may suffer from aesthetic concerns due to the colour of their teeth.

Histologically, normal dentine contains phosphophoryn, a protein which makes up most of the dentine matrix. This protein is secreted by full differentiated healthy odontoblasts. In DI, there is a deficiency in the amount of phosphophoryn thus the teeth affected are less mineralised (Takagi and Sasaki, 1988). Additionally, dentine in DI is abnormal and shows missing, abnormally structured or reduced numbers of dentinal tubules (Rios et al., 2005).

1.3.1 Treatment of DI

Early diagnosis and treatment for DI is important due to the rapid loss of tooth structure (Clark et al., 2019). The treatment of DI is aimed at improving aesthetics and reducing sensitivity (Rios et al., 2005). Maintaining occlusal height and adequate jaw development for the eruption of permanent teeth is also recommended (Clark et al., 2019). Restorations with composite resins, veneers and crowns can be carried out. Unfortunately due to the defective tooth structure, the bonding of resins to teeth affected by DI may be compromised however clinically it appears to show successful results in most patients. In cases of severe discolouration, full coverage restorations may be required (Rios et al., 2005).

Prior to carrying out any dental treatment, full examination, both clinically and radiographically is essential. This will ensure adequate diagnosis of any pulpal calcification, thin roots and abnormal tooth structure. In the paediatric patient, clinical restorability and long-term prognosis must be decided for each tooth prior to treatment. This also ensures proper treatment planning. One must also be aware of the potential loss in vertical dimension of occlusion and take measures to maintain it when necessary (Rousseau et al., 2018).

It is currently thought that treatment should be carried out early to help prevent some of the deterioration associated with DI, particularly in the primary dentition (Rousseau et al., 2018). As with all patients, treatment should start with a plan for caries prevention according to the Department of Health prevention toolkit for high risk patients including oral hygiene instructions, fissure sealant placements, fluoride application and diet advice (Munday, 2008). This advice is seconded by Clark et al in their 2019 paper (Clark et al., 2019). Full coverage restorations such as pre-formed metal crowns are ideal as they will help to prevent tooth fracture and decay (Rousseau et al., 2018; Clark et al., 2019). Conventional or intracoronal restorations are more poorly retained in teeth affected by DI and may increase vulnerability to fracture (Rousseau et al., 2018). In the permanent dentition, if teeth show signs of post-eruptive breakdown, then full coronal coverage is indicated. If the teeth are of poor prognosis, treatment options are limited and may require early extraction with space maintenance (Rousseau et al., 2018).

It is important to note, that although not all persons affected by OI also have DI, some features of DI may be present in the dentition and thus extra care should be taken when treating these patients (Rousseau et al., 2018).

1.3.2 Other Dental Concerns

1.3.2.1 Malocclusions

Another noted oral manifestation of OI is malocclusion, particularly anterior overbites (Okawa et al., 2017), class III malocclusions and anterior or lateral open bites (Rizkallah et al., 2013). As well as a possible increase in the incidence of caries and posterior crowding (Schwartz and Tsipouras, 1984). Some of these can be seen in figure 1.9.

In a study of 40 OI patients with types III and IV OI, the incidence of Class III malocclusions in the OI population is significantly higher than in the general population,

70% compared to 3-8% (O'Connell and Marini, 1999). In another study with a larger variety of OI types (I, III, IV, V), 30% of the entire cohort was found to have malocclusions, with class III malocclusions being the most prevalent. The lower percentage of malocclusions seen in this study could be due to assessing all types of OI as one cohort instead of dividing them by OI type as above (Clark et al., 2019). Jabbour et al, also found class III malocclusions in all patients with OI types V and VI however the numbers of these patients was very small at three overall and so the results are not conclusive (Jabbour et al., 2018).

The cause of the increased incidence of malocclusions has been related to inhibited maxillary growth and mandibular prognathism together with dento-alveolar abnormalities due to the bone abnormalities in OI (Clark et al., 2019).

The complexity of these malocclusions imply a need for orthodontic correction, possibly with the need for orthognathic surgery (O'Connell and Marini, 1999; Clark et al., 2019). It has been suggested that the size of the head, which may be abnormal in OI patients, may contribute to this (O'Connell and Marini, 1999). A different study places the incidence of class III malocclusions in all types of OI at 30%. The incidence is types III and IV OI is also reported as 70-80% in this study (Okawa et al., 2017). Given the frequency of class III malocclusions, an increased incidence of crossbites is not unexpected. Interestingly, there seems to be an occurrence of approximately 46% of posterior open bites in children with OI older than nine years old (O'Connell and Marini, 1999). Lateral, or posterior open bites are a rare finding in the general population (Rizkallah et al., 2013).

The literature does not yield a large amount of data regarding orthodontic treatment and orthognathic surgery in children with OI. Several case reports of patient's treatment with orthognathic surgery or rapid maxillary expansion show no adverse reactions due to the bisphosphonate therapy. Successful bonding of brackets to teeth affected by DI has also been reported (Hartsfield et al., 2006; Rosen et al., 2011; Ierardo et al., 2015).

1.3.2.2 Ectopic/impacted teeth

An increased incidence of impaction of the first and second molars is also found in patients with OI (Schwartz and Tsipouras, 1984). Impacted and missing teeth can be found in persons with type III OI. Ectopic eruption of the dentition may also be present. It has been suggested that this may be due to the bisphosphonate treatments as the

natural bone resorption mechanism around the dental follicle may be disturbed (Rousseau et al., 2018).

1.3.2.3 Hypodontia

Agenesis of a variable number of teeth in different types of OI has been reported in several studies including Malmgren et al, 2017 who reported it at 17% and O'Connell et al 1999 at 10% (O'Connell and Marini, 1999; Malmgren et al., 2017). This can be seen in figure 1.9 below.

Figure 1-9 - Radiographs and clinical photographs of a patient with OI type V who has hypodontia and a class III maloclussion. Image taken from Genotype and malocclusion in patients with osteogenesis imperfecta (Jabbour et al., 2018)

Treatment should be planned based on the patient's severity of dental and medical conditions. It can be either by conventional orthodontics or may require surgery. Surgery should be limited if possible as healing and success may be influenced by bisphosphonate treatment and by the severity of OI (Rousseau et al., 2018).

Given the amount of dental concerns patients with OI can potentially suffer from, the affect these concerns can have on their quality of life is important to consider. The effect of both OI and dental concerns in OI on quality of life, and oral health related quality of life will be discussed fully in sections 1.4 and 1.5 respectively.

1.4 Quality of life in OI

The concept of quality of life was developed after the World Health Organisation (WHO) gave a definition of health which not only related to the physical, but also the mental and social condition of a person. This definition was 'The state of complete physical, mental and social well-being and not merely the absence of disease or infirmity' (de Wit and Hajos, 2013). Quality of life thus also relates to a person's perception of their health and how a particular condition will affect their quality of life. Implying that whilst physical disability plays an important part, the perception of how the disability affects a person's life might differ from individual to individual (Bagramian and Inglehart, 2002). Quality of life linked to health conditions has now been investigated for many years. With the realisation that prolonging life or removing disease is not necessarily sufficient if it does not make one's life better, measures to determine health-related quality of life began to evolve (Locker and Allen, 2007). WHO defines quality of life as 'the individuals' perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns' (Dahan, 2016). This trend naturally progressed into the dental field where oral health related quality of life began to be measured (Locker and Allen, 2007).

OI can have a serious impact on the lives of both the children as well as their parents. The impact on quality of life begins at diagnosis and affects the parents particularly. Parents may experience devastation; on finding out their child has a chronic condition; or relief; due to a reduction in uncertainty caused by frequent trauma or a misdiagnosis of abuse (Hill et al., 2019). Many may require psychological support which is not commonly given according to a study by Dogba et al, 2016 (Dogba et al., 2016).

Studies show that children who live with OI are constantly aware of the possibility of fracturing of their bones and this alters their behaviour compared to children without OI (Dogba et al., 2013). This constant awareness will affect many aspects of a child's life as it will affect the way they interact with other children and the world around them. The constant awareness that fractures may occur will prevent them from fully participating in activities which other children without OI may be able to undertake. Parents will also be affected as they will constantly watch out for their child and worry about the possibility of fractures. Early life can also be a challenge as parents may worry that handling their child may cause breaks or trauma (Hill et al., 2019). These children also experience challenges in their daily lives such as getting into cars or reaching for objects, particularly those children who have more severe types of OI and are wheelchair bound. The number of medical appointments also means that these children miss days from school and this

has an impact on their education (Dogba et al., 2013). Children with OI also seem to feel that their independence may be reduced due to their condition with parents being overprotective to reduce the risk of fractures and pain (Hill et al., 2014).

Interestingly, a systematic review of papers related to quality of life of children with OI showed that although there were lower scores in the physical sphere of quality of life, quality of life related to the psychosocial was similar to that of the general population. Bisphosphonate treatment also seems to have an important effect on quality of life with children reporting and increased quality of life however the results are inconclusive due to imperfections in study designs (Dahan, 2016). The effects of bisphosphonate on the child's quality of life will also impact the parent's confidence as they feel the improvement in their child's well-being (Hill et al., 2019).

Parents' lives are impacted in different ways than the child's. Many families find that one parent will need to stay at home to help take care of their child and take them to their medical appointments. This often can have a financial burden on the family. The parents will also be concerned about the possibility of fractures occurring and will often be required to modify their behaviours and their plans in a way that accommodates their child's safety (Dogba et al., 2013). Parents may also feel an emotional burden due to the amount of everyday care, health appointments, fractures and the effort to keep their child safe which results in feelings of lack of control, helplessness and unpredictability of when problems can occur. Parents who have OI themselves often feel guilty in passing the hereditary condition on to their offspring (Hill et al., 2019).

A recent study by Hill et al in 2014 attempted to identify quality of life issues in children with OI using an interview-based system as a preliminary stage in developing a QoL measure for children with OI. The authors found that there was a difference in themes that should be included for children with OI. The aim of this study was to see how OI impacts on QoL and well-being in the paediatric OI population through a series of interview with children with OI, their parents and their health-care professionals. The author interview 10 children between the ages of 7 and 18 years old, 10 parents and 5 health-care workers including physiotherapists, occupational therapists, a nurse and a consultant. The participants were recruited when attending the metabolic clinic. The majority of children chose to have the interviews conducted without their parents present and interviews ranged from 13 to 52 minutes depending on the child's age.

This study identified 6 main themes which affect the quality of life of children with OI. These themes are being safe, reduced function, pain, fear, independence and isolation

or being different. This study highlights that although functional disability can be an issue in OI, many times, issues related to quality of life have a different source. QoL measures specific to a particular condition will detect a change in QoL within the population with different interventions and treatment modalities, however, it is then not possible to compare with different populations (Hill et al., 2014)

The Hill et al., 2014 study found that Reduced function appeared to cause the largest problems after an injury, as did pain. It was also found that several participants mentioned needle phobia when talking about fear which is particularly relevant to dentistry. Older children felt their independence was stifled due to overconcerned parents and parents tended to agree that they found it difficult to let go. Although the themes were generally similar to those in other QoL measures, fear and safety related to fractures is particular to this group and showed the need for a specific QoL measure. (Hill et al., 2014).

In the United Kingdom (UK), the Paediatric Osteogenesis Imperfecta National Team (POINT) take care of these children. This team includes clinicians, specialist nurses, occupational therapists, physiotherapists and others, and they all contribute to maintaining the health of these children. There are four specialist centres for children with OI in the UK, and Great Ormond Street Hospital (GOSH), where this study is to be conducted, is one of them. The Brittle Bone Society is the national charity for children with OI, and it provides emotional and financial support to families of children with OI.

This section covers why children with OI may have a reduced quality of life. However, dental concerns or pain are not mentioned when referring to quality of life in OI. As discussed in section 1.3, there are several dental considerations in OI including DI, malocclusions and missing teeth. It is important for these children to have holistic support and therefore looking at their Oral health related quality of life is an important consideration in this.

1.5 Oral health quality of Life

Oral health related quality of life was first described in the 1980s, compared to health related quality of life which was described in the 1960s. The delay in recognising the importance of oral health related quality of life can be attributed to the fact that until recently, the idea that oral conditions could affect general health was not a popular one (Bennadi and Reddy, 2013).

Oral health related quality of life has been defined as 'the impact of oral disorders on aspects of everyday life that are important to patients and persons, with those impacts being of sufficient magnitude, whether in terms of severity, frequency or duration, to affect an individual's perception of their life overall' (Locker and Allen, 2007). It was later described as 'a multidimensional construct that reflects (among other things) people's comfort when eating, sleeping, and engaging in social interaction; their self-esteem; and their satisfaction with respect to their oral health'. This definition seems to resonate better with the association of oral health related quality of life with function, psychology, social life and experience of pain (Bennadi and Reddy, 2013). Oral care has had an impact on quality of life since its conception as people frequently visit the dentist to ameliorate their quality of life for the alleviation of pain or the improvement of aesthetics. This impact on quality of life has determined the way dentistry has developed and its focus on the reduction of pain and painful dentistry with the use of drugs such as local anaesthetics as well as the development of cosmetically acceptable courses of treatment (Bagramian and Inglehart, 2002).

Oral health-related quality of life has recently become an important part of clinical dentistry and dental research. In 2003, the World Health Organisation (WHO) recognised it as a vital part of general health. (Sischo and Broder, 2011). Since then several OHRQoL questionnaires have become available, for adults and children. The importance of oral health is evident in everyday life, adequate oral functioning allows eating, speaking and socializing (Manapoti et al., 2015). A systematic review of several available oral health related quality of life measures claims that although many of the available measures are patient-centred and incorporate aspects of daily living, there is great variation in what criteria are used to establish oral health related quality of life in the measures (Locker and Allen, 2007).

When evaluating indices measuring oral health related quality of life it is important to bear in mind that they must have the following properties:

- Validity
- Appropriateness and acceptability
- Reliability
- Responsiveness to change
- Interpretability (Bennadi and Reddy, 2013)

Although categorical measures of oral health related quality of life have been mentioned, many questionnaires involve multiple items. Questionnaires are also being developed to tailor to specific conditions; such as dental anxiety or oral cancer; or specific groups; such as the elderly or children. The number of questions and the format of the question also varies amongst the various measures available (Bennadi and Reddy, 2013).

Differences have been noted between oral health related quality of life in children and in adults. This indicates that there is a need for different measure to be developed for measuring oral health related quality of life in children. The first child specific measure was the Child perception questionnaire (CPQ). The first measure to incorporate both positive and negative aspects of oral health was the Child Oral Health Impact Profile (COHIP). The assumption is thus that not only can it measure the absence of a condition but also whether treatment or intervention has a positive effect on oral health related quality of life (Genderson et al., 2013a).

There are important oral health aspect to OI. A thorough search of the literature showed that the impact of oral health related quality of life of children with OI has not been investigated. Several papers assessed the health related quality of life in adults (Widmann et al., 1999; Widmann et al., 2001; Seikaly et al., 2005; Kok et al., 2007; Balkefors et al., 2013; Dahan, 2016) and children (Fano Virginia et al., 2013; Hill et al., 2014), however none were related to oral health related quality of life.

Other medical conditions have been investigated with respect to oral health related quality of life including, cleft lip and palate (Bos and Prahl, 2011; Eslami et al., 2013; Ward et al., 2013), congenital bleeding disorders (Salem and Eshghi, 2013), Trisomy 21 Syndrome (AlJameel, 2015), Sickle Cell Disease (da Matta Felisberto Fernandes et al., 2016) and cystic fibrosis (Patrick et al., 2016). The Oral Health-Related Quality of Life (OHRQoL) of children with OI has not been investigated to date.

For the purposes of this study, only questionnaires related to Child oral health related quality of life were considered. There are many oral health related quality of life questionnaires available. These included:

- Child Oral Health Impact Profile (COHIP) (Broder et al., 2007)
- Short form of the Child Oral Health Impact Profile (COHIP-SF) (Broder et al., 2012)
- Paediatric Oral Health-Related Quality of Life Questionnaire (POQL) (Huntington et al., 2011)
- Child Oral Impacts on Daily Performance (COIDP) (Yusuf et al., 2006)
- Scale of Oral Health Outcomes for 5 year old children (SOHO-5) (Tsakos et al., 2012)
- Paediatric Quality of life Inventory (PEDsQL) (Varni et al., 2001)
- Child Perception Questionnaire 11-14 year olds (CPQ 11-14) (Jokovic et al., 2006)
- Michigan Oral Health-Related Quality of Life Questionnaire – Child Version (MOHRQoL-C) (Filstrup et al., 2003)
- The Early Childhood Oral Health Impact Scale (ECOHIP) (Pahel et al., 2007)
- WHO oral health questionnaire for children (World-Health-Organisation, 2013)

All the questionnaires were assessed in detail to determine their suitability for this study. While all the above questionnaires have been validated and have their uses, the COHIP-SF is the questionnaire which best suits the purposes of this study. This questionnaire was chosen for several reasons.

Firstly, it is a questionnaire which has been validated for children as young as 8 years old and has been used in similar studies related to other conditions when assessing oral health-related quality of life. These include conditions such as craniofacial anomalies and cystic fibrosis (Ward et al., 2013; Patrick et al., 2016). This is useful as the final results can be compared with those of other studies, to assess for any related trends between conditions. COHIP allows for comparison between age groups unlike questionnaires such as the CPQ which has two different questionnaires for children older or younger than 11 years old. The short form of the questionnaire was chosen to ensure the children participating were not overwhelmed by the number of questions and could therefore answer the questionnaire quickly and without too many problems (Broder et al., 2012).

2. Aims

2.1 Primary Aim

To investigate the oral health related quality of life of children aged eight to sixteen years old with Osteogenesis Imperfecta

2.2 Secondary Aims

To assess patient's thoughts on their dental health care and identify key areas of concern with their dental health through a service evaluation.

To identify areas of concern in children with OI which are specific to this group and thus be able to tailor the information we provide towards their specific needs through questionnaire based research.

3. Service Evaluation

3.1 Methodology

Prior to beginning the questionnaire-based part of the study, a service evaluation was carried out in the OI department at GOSH. GOSH for children has a highly specialised OI service which sees children with OI in their catchment area for routine check-ups from birth until at least 16 years old. It is one of five OI centres in the UK. The service evaluation comprised of a series of five questions. The questions were designed to assess if children with OI attending review at GOSH are receiving adequate dental care. It was also used to assess their thoughts on their dental health care and identify key areas of concern with their dental health and dental care.

Drafts of the questions for the service evaluation were developed and edited by the research team. The final version (Version 3) was used in the clinical setting.

The GOSH OI department and EDH paediatrics department have a long-standing relationship where each child seen with OI is referred for a dental assessment at the EDH as children with OI often have DI or other dental concerns.

The standard to be reached in this service evaluation is that every follow up patient seen by the OI team at GOSH has been referred to the EDH. Service evaluation registration was carried out through the clinical audit team at GOSH and was approved. As this was a service evaluation ethics approval was not necessary for this part of the project.
A pilot study was carried out on the 5th of October 2018 to assess ease of understanding and reception of the questions with a total of five responses. The questions appeared to be well received generally however a slight change was carried out – this edit was to add 'Not applicable (N/A)' to question three (see below).

The service evaluation was carried out at GOSH during the OI clinics which run on Fridays and every third Wednesday of the month. This ran from the 5th October 2018 until the 19th December 2018.

All persons who attended the OI clinic for regular assessment at the OI clinics were asked a short series of questions. These were:

Q1) Are you/ your child under the care of a local dentist? Yes/ No

Q2) Have you/ your child been seen by a dentist at the Eastman Dental Hospital? Yes/ No

If yes, are you/ your child still under their care now? Yes/ No/ Don't Know

Q3) Did you/ your child find your appointment at the Eastman Dental Hospital helpful? Yes/ No/ N/A

Q4) Do you/ your child have any concerns about:

The appearance of your teeth - Yes/ No (If yes, in what way:)

The way you bite - Yes/ No

Pain in your teeth - Yes/ No

Pain in your jaw - Yes/ No

Q5) Have you/ your child had trouble accessing dental care and/ or treatment? Yes/ No

If yes, where?

The question sheet used can be found in Appendix 1.

The researcher attended the clinic and asked the child or the parent the questions at the appropriate time during their routine appointment.

The data collected was inserted into an MS Excel spreadsheet and simple statistical analysis was carried out using excel to obtain the necessary results.

3.2 Results

The service evaluation was carried out from October - December 2018 and in this time, 87 patients attended the OI service. One family was not asked to participate due to concerns regarding social services and safeguarding which prevented the student researcher from being present during the patient's appointment, resulting in 86 participants. The age ranged from two months to 18 years old with a mean age of 8.31. 57% were male and 43% were female. From these patients, the majority had attended for a routine follow up appointment (77), while only nine were new patients.

The questions were asked during the routine appointments and most of them were answered by the patients' parents. However, when it came to the question about what dental concerns the child had, many of the older children also responded to the question themselves.

Questions 1,2 and 3:

1. *Are you/ your child under the care of a local dentist?*
2. *Have you/ your child been seen by a dentist at the Eastman Dental Hospital? If yes, are you/ your child still under their care now?*
3. *Did you/ your child find your appointment at the Eastman Dental Hospital useful?*

Out of the 86 participants, 72 (84%) children were under the care of a local dentist, from these 72, 23 (32%) were also seen at the Eastman Dental Hospital whilst 49 (68%) were only seen by their local dentist. Six (7%) children were seen only at the Eastman Dental Hospital and did not have a general dentist. Overall, 29 (34%) children were seen at the Eastman dental Hospital. Approximately half (48%; n=14) of the children seen at the Eastman Dental Hospital where still under their care. From the patients seen at the Eastman Dental Hospital, 86% (n=26) claimed to have found this appointment useful.

Question 4: Do you/ your child have any concerns about:

The appearance of your teeth (if yes, in what way)
The way you bite
Pain in your teeth
Pain in your jaws

Many of the children who were asked this question as part of this service evaluation did not have any concerns regarding the appearance of their teeth. 33/86 (38%) children had aesthetic concerns while 53 did not. For those children who did have concerns about

the appearance of their teeth, the reasons varied and were grouped into orthodontic concerns (eg. alignment, gaps, overbite), discolouration (eg. grey, brown, yellow), hypomineralisation, structural concerns, hypodontia, cavities or no reason. This variety and its distribution can be seen in Figure 3.1 below. The most common type of concerns were orthodontic concerns at 43% (n=14), followed by concerns due to discolouration at 33% (n=11).

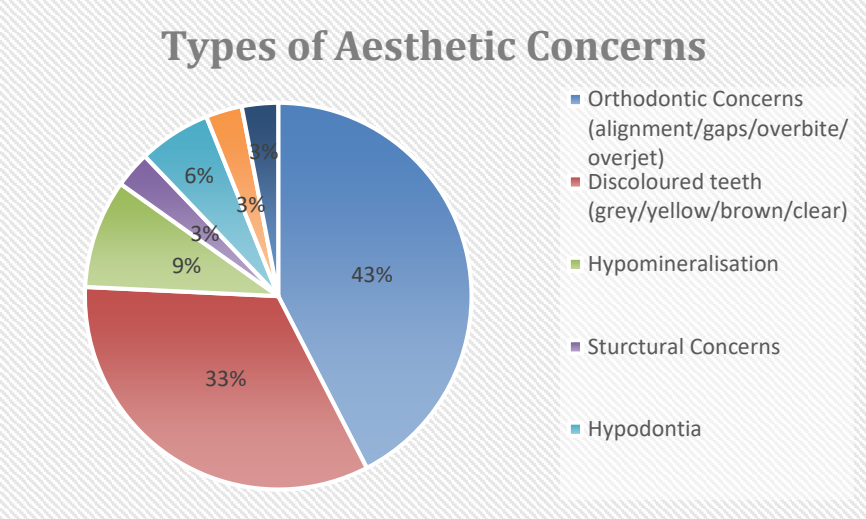

Figure 3-1 - Chart showing different types of dental aesthetic concerns patients with OI had

The children were divided into age groups depending on dental development. Studies show that older children are more bothered by aesthetics and malocclusion, thus we divided the children by age to see if this held true for our study. The age groups were five years old or young, between six and twelve years old, and twelve years old or older. This division showed that aesthetic concerns were most prevalent in the mixed dentition phase with 44% of these children having aesthetic concerns (n=18), whilst they were the least prevalent in the permanent dentition phase at 28% (n=5). The graph below shows this distribution (Figure 3-2).

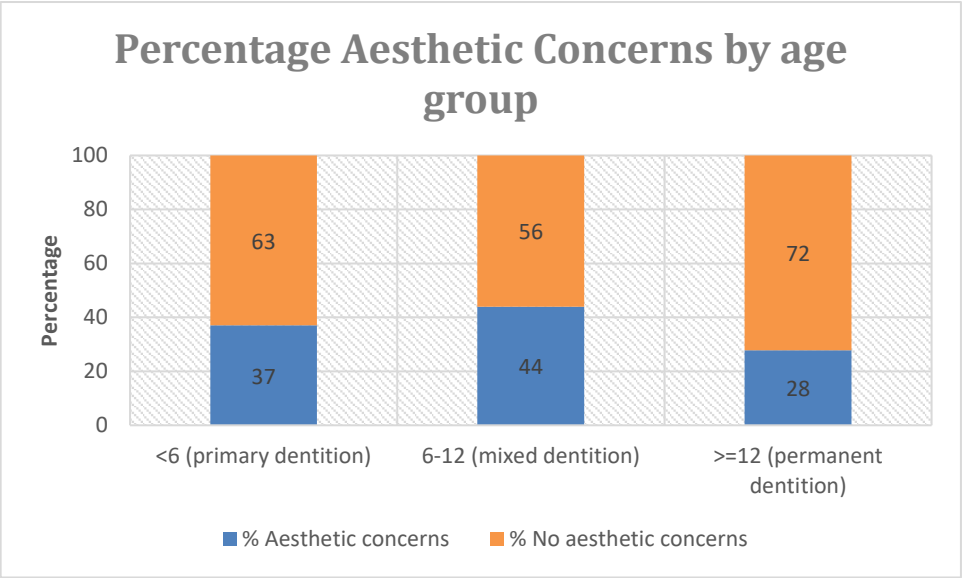

Figure 3-2 - Graph showing aesthetic concern distribution by age group

Other types of concerns included concerns about biting, pain in the teeth and pain in the jaws. These were not common with only 16 children having concerns about biting, 21 children, which is less than 25%, having pain in their teeth and 4 children having pain in their jaws. The total percentage of children having any type of dental concern collated to 57% (49 children). Table 3-1 shows the percentage of all concerns overall and by age group.

Table 3-1 – Table showing distribution of all concerns, overall and by age group

	Percentage			
	Aesthetic concerns	Biting concerns	Pain in teeth	Pain in jaws
Total	38%	19%	24%	5%
<6 (primary dentition)	37%	4%	19%	4%
6-12 (mixed dentition)	44%	22%	27%	2%
>=12 (permanent dentition)	28%	33%	28%	11%

Figure 3.3 below shows the distribution of types of concerns by age group. Again, we see that the majority of concerns are found within the mixed dentition group. The exception being pain in the jaws which was more prevalent amongst the permanent dentition group.

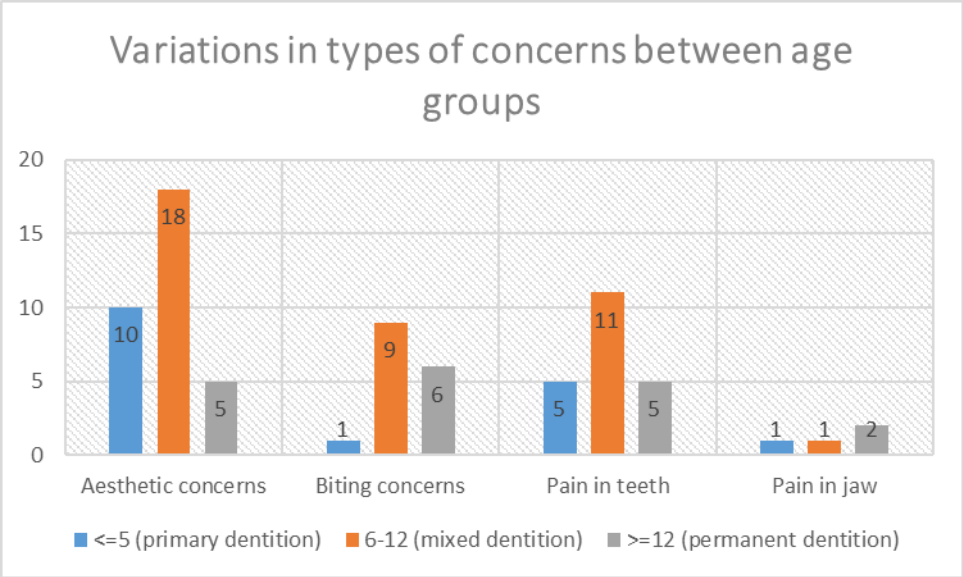

Figure 3-3 Graph showing all types of concerns distributed by age group

Question 5: Have you/ your child had trouble accessing dental care and/ or treatment? If yes, where?

The majority of parents stated they had not had trouble accessing dental care or dental treatment (78/86, 91%). The eight children who had trouble were asked what issues they had experienced, and the results are shown in Figure 3.4. Those who had trouble accessing care at the GDP stated this was due to the dentist's concern about treating a child with a medical history of OI; for those who had trouble with the Eastman Dental Hospital this was due to problems in contacting the hospital to book an appointment. For the final group, those who had trouble accessing care at the orthodontist, this was due to concerns providing orthodontics when the child was undergoing bisphosphonate treatment.

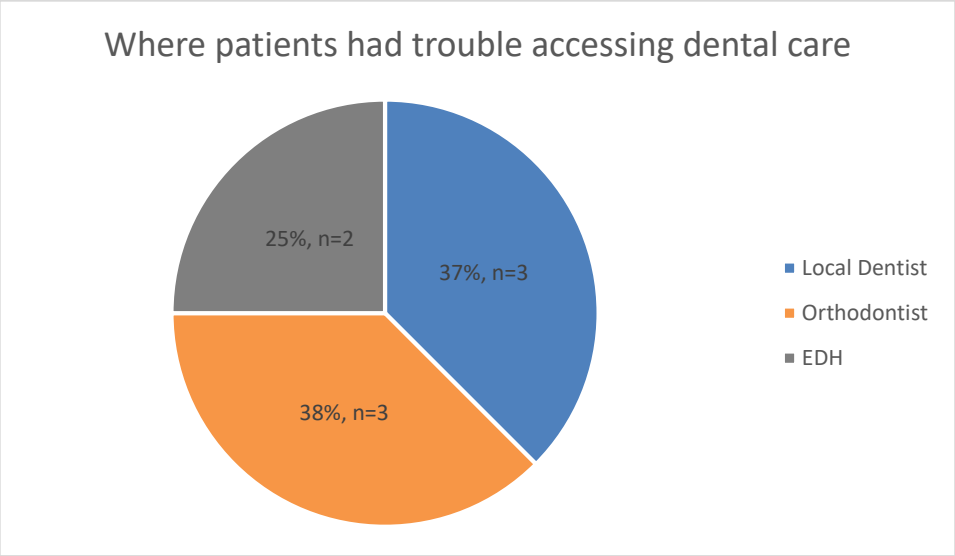

Figure 3-4 - Chart showing where patients had trouble accessing dental care.

This service evaluation showed that there were dental needs in the OI population therefore this demonstrated the need for the study assessing OHRQoL in children with OI.

3.4 Discussion

This service evaluation was carried out to assess dental concerns of children seen at the OI department at GOSH, as well as to ensure they were receiving adequate dental care.

In the United Kingdom, the British Society of Paediatric Dentistry recommends that children first see a dentist at the appearance of milk teeth and continue to attend for regular check-ups (Stevens, 2018). The American Dental Association and American Academy of Paediatric Dentistry recommend the first dental check-up is within 6 months of the first teeth erupting (ADA, 2013). This would imply that all children over 1 year old should be seeing a local dentist. In this service evaluation only 83% of participants were under the care of a local dentist, however 10% where one year old or less, and 23% where 3 years old or less. For these children, parents may have felt they were too young to take to the dentist and had as yet not been registered with a general dentist. This implies a need to continue to educate people on the importance of a dental check-up by one (BSPD, 2020), whether they have OI or not.

All OI patients should be referred to the EDH for dental assessment, as part of their care pathway. Results of this service evaluation show that this pathway is not being utilised the way it should. Only 33% of the patients seen for their routine OI appointment had been seen at EDH. A limitation of this study may be that if the child was referred for an appointment at the EDH several years previously and then discharged due to no dental issues, the family may not have remembered the appointment given the multitude of other appointments necessary for a child with OI. Regardless, this number was unlikely to make up for the remaining 66%. Another reason for the children not yet being seen at the EDH is that the OI team may have considered the child too young for a dental check-up. This highlights the need for an official standard to be setup with the OI team for referral to EDH at an appropriate age. Ideally when several of the primary teeth have erupted (three years old) and again when the permanent teeth are partially present in the mouth (eight years old). These ages are appropriate to ensure DI is diagnosed and treated early in the primary dentition and again in the permanent dentition. In the permanent teeth the presence of malocclusion should also be noted, and interceptive orthodontics carried out when necessary. This would make any future service evaluations on the subject more appropriate. The standard for this service evaluation (every follow up patient seen by the OI team at GOSH has been referred to the EDH) has not been reached. Investigation into the referral pathway and adapting the pathway to ensure all children are referred when appropriate is necessary. It is also important that both the OI team at GOSH and the paediatric clinicians at the EDH are aware of the

importance of this relationship and encourage the routine use of this pathway, ensuring children with OI receive the best possible dental care.

The results of this service evaluation showed that although many of the patients attending GOSH for their OI appointment see their local dentist, most of them (66%) had not been seen at EDH. Up to 50% of patients with OI can also have DI (Rios et al., 2005) whilst malocclusions are present in 30% of patients with OI (Okawa et al., 2017). These numbers highlight the importance of dental care in patients with OI. Given that 86% of families seen at EDH found the appointment useful, it is a shame that only 33% were seen by the dental team at EDH.

When asking the questions, responses where usually answered by the parents or guardian, however in some cases, older children also replied. This may give a more accurate representation of the situation as parental proxy reports may not always give the correct perception of their child's feelings (Tsakos et al., 2012). It has been found that parents can underestimate their child's anxiety or functioning (Alcantara et al., 2017), therefore reports from the child may give a more representative result.

Overall, 90% of the patients responding to the service evaluation were follow-up patients. This would be expected as all children diagnosed with OI in the GOSH catchment area are seen on the OI clinic from diagnosis until they are transferred to adult services from the age of 18 (GOSH, 2020). Diagnosis of OI usually happens early in life and has been quoted at a median of 38 months (Brizola et al., 2017). The median age of children in this service evaluation was 8 years old.

Another point of note is that 86% of families who were referred to the EDH for dental consultation found the appointment useful. No other information to determine the reason for this was gathered however, one can speculate that that this was because these families felt their dental concerns were addressed through these consultations.

When asked if the child had concerns regarding different aspects of their teeth (appearance, pain, biting), the majority felt they had aesthetic concerns (n=33). The most common aesthetic concerns were orthodontic concerns (16%, n=14) and discolouration (13%, n=11). Overall, 16% of the participants had orthodontic concerns, which is lower than other studies reporting incidence of malocclusion in the OI population. Malocclusion is present in 30% for the OI population as a whole (Clarke et al., 2019) but varies depending on the type of OI with types III and IV OI having a higher percentage (Jabbour et al., 2018). The reason for the discrepancy between the population in this service

evaluation and the literature may be twofold, firstly the average age of our population was 8.31 years, which may be too young for orthodontic concerns. Another reason may be that these children, whilst having orthodontic issues, where not concerned by them due to their overall physical condition.

Only 13% (n=11) of the participants brought discolouration up as a concern. In the general OI population, the prevalence of DI, which is the most likely cause of dental discolouration in OI, ranges from 21-73% (Rousseau et al., 2018). The low numbers of children concerned about discolouration may be due to a proportion of older children who only had DI in the primary dentition, or the DI not being severe enough to cause an aesthetic concern.

Older children tend to be more concerned about their dental appearance. When dividing by age group according to the phase of dentition (primary dentition, mixed dentition and permanent dentition), it was found that aesthetics was most concerning for the mixed dentition group, with 44% (n=18) of these having aesthetic concerns. This number was only 28% (n=5) in the permanent dentition group. The reason for this distribution could be twofold, first of all the small number of children in the permanent dentition phase (twelve years old or older) at n=18. The second reason could be that these children already have had dental treatment carried out. This point should be investigated further.

Other concerns investigated were concerns with biting, pain in the teeth and pain in the jaws. For the former two, again the mixed dentition group were the most affected. The latter was most prevalent in the permanent dentition phase however was only experienced by four participants overall, two in the permanent dentition (a 16 year old female and 14 year old male), and one each in the primary dentition and mixed dentition groups. The child in the primary dentition group was a one year old male. This pain would have been reported by his parents however is unlikely to be the case as most jaw pain is due to temporomandibular disorder which is more common in older children and is often caused by stress and anxiety. A recent study has shown that very few people (12%) with OI have temporomandibular disorders (Bendixen et al., 2018). Jaw fractures in children with OI is also uncommon (Kobayashi et al., 2016). The small number of patients in each of this groups is a limitation of the service evaluation and the results may not be representative of the whole OI population.

Trouble accessing care at any dentist due to their medical condition is disappointing and stressful for families. It is important to educated dentists on treating patients with OI and ensuring that if they feel incapable of treating the patient, they refer them to the

appropriate specialist practice. Education can be done by targeted campaigns and webinars organised together with national groups for dentists such as the General Dental Council, The British Society of Paediatric Dentistry or the Faculty of General Dental Surgery and the British Dental Association. Another way to educate dentists is to present our findings at national dental conferences and study days. Delay in treating dental problems in children with OI can lead to pain, increased aesthetic concerns and functional problems. If the child has DI, delay in treatment can require more extensive treatment at a later stage, adding to the appointment burden of these children (Rousseau et al., 2018). For those children whose orthodontist was unwilling to treat them due to their bisphosphonate therapy, again it is important to ensure that orthodontists are aware that it is safe to treat these children. There are several reported cases of successful orthodontic treatment in children on bisphosphonate therapies and children with DI so their orthodontic treatment should not be delayed (Hartsfield et al., 2006; Rosen et al., 2011; Ierardo et al., 2015). Again, if the local orthodontist is concerned, referral to a secondary care orthodontic clinic may be indicated.

This service evaluation indicated the need to reassess the pathway for children with OI to ensure all these children are referred and receive appropriate care. It also highlighted that children with OI do have dental concerns, and many feel they benefit from care at a specialist practice. This justified the need to begin a larger study assessing the OHRQoL in children with OI to assess trends and ensure we can tailor their dental treatment to any particular dental needs that may arise.

4. Questionnaire

4.1 Methodology

The main focus of this study is a mixed qualitative and quantitative questionnaire based study involving children with OI aged between eight and sixteen years old, who are receiving care from the OI clinic at Great Ormond Street Hospital (GOSH). The questionnaire assessed the oral health related quality of life in children with OI.

Prior to handing out the questionnaire, a thorough search of the literature was carried out using online resources including PubMed, ProQuest, Cochrane Library, Elsevier, Up-to-date and Medline (Ovid). These were accessed using the University College London Library resource 'Explore' available to the student researcher.

The search was related to oral health related quality of life in children with OI. The important key words used included:

'Oral health related quality of life'
'OHRQoL questionnaires'
'Osteogenesis imperfecta'
'Quality of life'
'Children'
'Paediatrics'
'Bisphosphonates'
'Dental concerns'

Whilst several papers were found pertaining to health related quality of life in adults or children with OI and oral health related quality of life in children with other medical conditions, no studies on oral health related quality of life of children with OI were available at the time. This research project seeks to contribute to this field of study.

The literature search also included searching for validated questionnaires associated with Oral Health related quality of life in children. For the purposes of this study, only questionnaires related to Child oral health related quality of life were considered.

The Short Form of the Child Oral Health Impact Profile (COHIP-SF) questionnaire was chosen for several reasons including validity and reliability; its use to assess OHRQoL in children with other concerns such as craniofacial abnormalities, Clefts of the lip and

palate and orthodontic concerns; and the fact that it is designed to be carried out by the child and not the parent. Direct answers from the children was ideal in this situation to assess the child's perspective. The fact that this questionnaire has been used in other studies to assess different conditions meant comparisons could be carried out when data analysis was done. This questionnaire is validated for children aged eight to sixteen years old and therefore this informed our inclusion criteria.

The COHIP-SF was adapted to add demographic data and information about the participant's OI. Two qualitative questions and five general questions about the child's view on their dental status were also added at the end of the questionnaire. These were added to confirm whether the COHIP-SF questions were accurately reflecting participants OHRQoL. The questionnaire can be found in Appendix 2.

The information leaflets, for children and for parents, consent form and assent form were developed and edited by the research team. To ensure adequate understanding and accessibility of the documents to young children, the documents were taken to the Young Persons Advisory Group (YPA group) at GOSH. The YPA Group is a group of children aged between nine to twenty years old who hold meetings at GOSH once a month and give feedback to researchers carrying out medical research involving children. The meeting was attended by the student researcher and chief investigator on the 7th July 2018.

A short presentation giving background information about OI and the rationale for the project was given by the student researcher. The documents were then distributed, and the children worked in groups of three to four children to come up with feedback. The children gave useful feedback on the information leaflet, generally agreeing that whilst easy to read and understand, a second information leaflet should be made for teenagers with more mature language. Their feedback resulted in the development of a second children's information leaflet for children aged thirteen to sixteen years old. The original information leaflet was targeted towards children aged eight to twelve years old. They also suggested the separation of each section of questions to different pages to reduce confusion and improve aesthetics of the document.

Subsequently, the final versions of all documents were developed. These can be found in appendices 2 – 7 as follows:

- Appendix 2 – Questionnaire
- Appendix 3 – Consent form
- Appendix 4 – Assent form

- Appendix 5 – Child information leaflet, 8-12-year olds
- Appendix 6 – Child information leaflet, 12-16-year olds
- Appendix 7 – Parent information leaflet

Ethical approval was obtained from the North of Scotland (2) Research Ethics Committee on the 14th November 2018 (reference number: 18/NS/0129). Health Research Authority approval was obtained on the 19th November 2019. Internal approval to carry out the project was also obtained from the Research and Development department at GOSH to confirm capacity and capability to undergo the study. This was obtained on the 8th January 2019. Finally, the student researcher was required to obtain an honorary research contract at GOSH and this was finalised on the 10th January 2019.

Questionnaires were first handed out on the 11th January 2019.

Participants were recruited from the OI clinics at GOSH, during routine scheduled appointments. The study was scheduled to run for one year from the 16th January 2019, or until approximately 100 valid questionnaires were collected. The questionnaires and all relevant documents were handed out at the dedicated OI clinics at GOSH on Fridays and Wednesdays, by the student researcher, Jasmine Cachia Mintoff, or a clinical member of the OI team.

Each questionnaire was given an identification number which was written on the cover page and first page of the questionnaire. The participants are first assessed for eligibility based on the criteria below:

Inclusion Criteria

- Children between the ages of eight to sixteen years old
- Children capable of understanding the questionnaire
- Children with OI
- English speaking participants or those for whom a translator can be booked/ a translator is present at appointment

Exclusion Criteria

- Children whose parents are unable to give informed consent

If the child fit the inclusion criteria, they were asked if they wish to participate in the study. Information leaflets were handed out to the child and their parent to explain about the study. Any questions from the parents or child were answered and then the child and parents were given adequate time to decide if they wished to participate or not. The voluntary nature of the study was emphasised, and written consent was obtained. Two copies of the consent form were handed out and signed so the parents could keep a copy of the consent form. An assent form was also filled in and signed by the child.

The first 10 questionnaires obtained were piloted to assess for any obvious issues. Two minor changes were added to the questionnaire,

1. The addition of the participants date of birth to the cover page
2. The addition of a question on ethnicity in the first section of the questionnaire.

The study then continued as detailed earlier.

The questionnaire was then completed at the appointment. The whole process took between 10-15 minutes. As the questionnaire was completed at the appointment, no follow-up was necessary. The completed consent forms and questionnaires were placed into an A4 envelope and sealed by the participant prior to returning it to the person asking for participation.

Once the completed consent forms and questionnaires were returned the cover page of the questionnaire and the consent forms were separated from the rest of the questionnaire to maintain anonymity. The questionnaires and consent forms were stored separately in locked rooms at the Eastman Dental Hospital University College London site.

4.1.1 Sample size

Sample sizes are calculated depending on the type of study, the confidence interval required, the acceptable margin of error and the expected response distribution. For a confidence interval of 90%, a margin of error of 5% and a response distribution of 50%, the required sample size is 106 responses in a population of 172. The plan was therefore to recruit approximately 100 participants to the study. As the study is questionnaire based, the sample size should be as representative as possible. The population size was determined from the GOSH OI patient database and included all children with OI fitting the age criteria of eight to sixteen years old.

When needed, patient data could be added to the correct questionnaire using the identification number written on each questionnaire. This was necessary for those participants who did not know what type of OI they had and to assess. This information was obtained from the patients records and was used in data analysis.

4.1.2 Statistics

Ethnicity was grouped according to the Office of national statistics groupings (Office for National Statistics, 2016) depending on how the participants self-identified.

Statistical data was obtained using the statistical software MS Excel and SPSS to achieve descriptive statistic results. Simple statistical analysis varied but included basic average and percentage calculations and comparative graphs. Distribution was tested using the Shapiro-Wilks test and QQ plots. The Shapiro-Wilks test is one which tests the null hypothesis that the sample comes from a normally distributed population using the formula seen in equation 1 (Deviant, 2020c).

Equation 1 – Shapiro Wilks Test - Where x_i = the ordered random sample values and a_i = constants generated from the covariances, variances and means of the sample (size n) from a normally distributed sample

$$W = \frac{\left(\sum_{i=1}^{n} a_i x_{(i)}\right)^2}{\sum_{i=1}^{n}(x_i - \overline{x})^2}$$

A Q-Q plot is a scatterplot created by plotting two sets of quantiles against one another. If both sets of quantiles came from the same distribution, we should see the points forming a line that's roughly straight.

For normally distributed data, a t-test (Equation 2) (Stats Direct Ltd, 2016) and standard deviation was be used, and for data which was not normally distributed, the non-parametric Mann Whitney U test (Equation 3) (Deviant, 2020b) and the interquartile range was used.

Equation 3 – T-Test - Where x = means, s^2 = pooled standard error of the two groups, and n = number of observations

$$t = \frac{\overline{x}_1 - \overline{x}_2}{\sqrt{(s^2(\frac{1}{n_1} + \frac{1}{n_2}))}}$$

Equation 2 – Mann Whitney U test - Where R = sum of mean ranks in the sample, and n = number of items in the sample

Cronbach's Alpha was used to assess internal consistency of the results. This test is used for Likhert scales such as the one used for the COHIP-SF section of the questionnaire. The equation can be seen in Equation 4 (Deviant, 2020a).

Equation 4 – Cronbach's Alpha - Where N = number of items, c = average covariance between item-pairs and v = average variance

$$U_1 = R_1 - \frac{n_1(n_1+1)}{2}$$

or

$$U_2 = R_2 - \frac{n_2(n_2+1)}{2}$$

4.1.3 Thematic Analysis

For the final section of the questionnaire, the qualitative data gathered will be analysed using the framework method which looks for common themes within the data to allow for analysis. This method is carried out by inputting all the data into a spreadsheet, familiarisation with the content and applying labels or 'codes' to each participant's responses depending on what is important within the qualitative information. The codes are then grouped into clear themes which can be analysed (Gale et al., 2013). The final identified themes will then be compared to the COHIP-SF scores to further validate the findings.

4.2 Results

4.2.1 General information

During the period of 11th January – 25th October 2019, a total of 112 eligible children attended the OI clinics. Out of these children, five children did not participate, and 2 questionnaires were invalid for the following reasons:

- Three families refused to answer the questionnaire;
    - In one family the daughter refused to answer
    - In the second family the parents were unwilling to spend the time to participate
    - The third family did not give a reason.
- Another family felt that the child would not be able to fill the questionnaire out appropriately due to learning difficulties.
- One child attended with his grandfather who did not have parental responsibility and as such could not give valid consent for the child to answer the questionnaire.
- Of the 108 participants,
    - Two were filled in by the same family twice
    - One questionnaire had the consent form signed by a grandparent who did not have parental responsibility.

Thus, a total of 106 questionnaires were included for data analysis in this study.

Feedback from families completing the questionnaire was good as many of them expressed a keen interest in the study and felt it could be important to help their children. This is clearly seen by the high level of participation in this study (n - 106/112, 94%).

4.2.2 Demographic Data

The average age of patients was 11.93 (range of eight to sixteen years old). The gender distribution was biased towards males at 58% (n=62), the remaining 42% (n=44) were females. No children chose the ‘other’ category for gender.

Ethnicity was left as an open question and all answers were grouped into five groups according to the Office for National Statistics. The majority of the participants self-reported as white – 54% (n=57). Three percent (n=3) responded as multiple or mixed ethnicities, 14% (n=13) reported as Asian or British-Asian, 7% (n=7) as Black/ Black-

British/ British-Caribbean and 8% (n=9) as other. 15% of the respondents (n=16) did not answer this question.

With respect to subjective severity of OI, the majority of cases self-reported as mild. Table 4.1 shows how the children rated their severity of OI.

Table 4-1 - Table showing subjective severity of OI

Self-reported severity of OI	Numbers	Percentages
Mild	55	52
Mild to moderate	27	25
Moderate to severe	11	10
Severe	7	7
No Answer	6	6

Two-thirds of the children participating knew what type of OI they had (n=70, 66%). Of those children who did not know what type of OI they had, the information was obtained from their medical records by their OI consultant and included in the results. The distribution of OI types can be seen in figure 4.1 below.

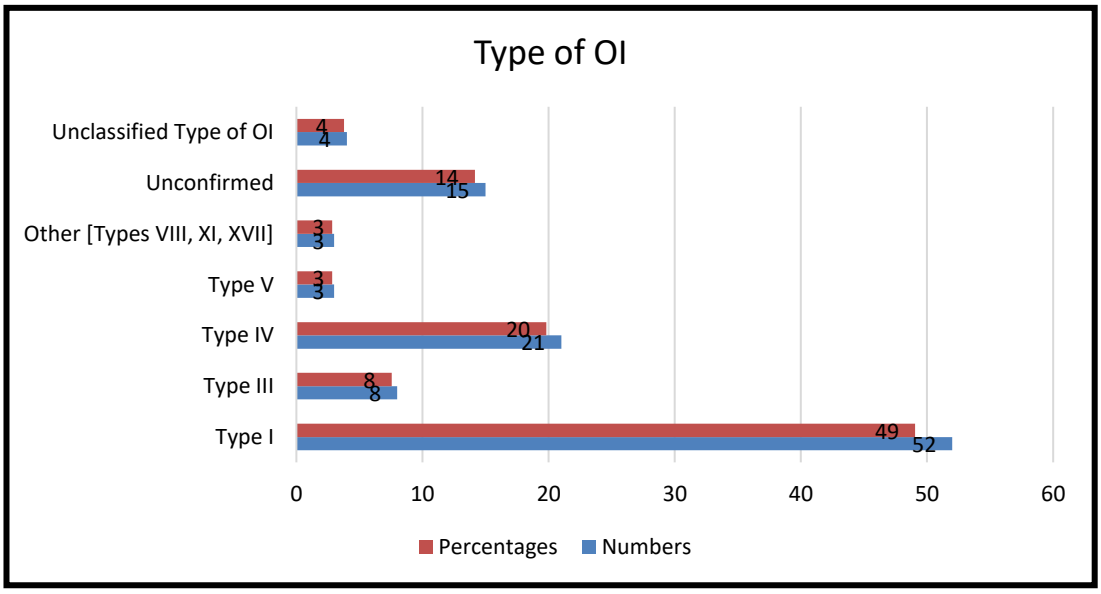

Figure 4-1 - Graph showing types of OI, numbers and percentages

Table 4-2 below shows demographic data overall, and according to gender for all questions.

Table 4-2 – Summary of Demographic Data

	Overall		Females		Males	
	Number	**Percentage**	**Number**	**Percentage of Females**	**Number**	**Percentage of Males**
Numbers	106	100%	44	42% (overall)	62	58% (overall)
Age						
Average Age	11.93	n/a	12.00	n/a	11.81	n/a
Age Range	8 to 16					
Less than 12 years old	46	43%	16	36%	30	48%
12 years old or older	60	57%	28	64%	32	52%
Ethnicity						
Asian	14	13%	4	9%	10	16%
Black	7	7%	4	9%	3	5%
Mixed	3	3%	0	0%	3	5%
White	57	54%	24	55%	33	53%
N/A	16	15%	7	16%	9	15%
Other	9	8%	5	11%	4	6%
Self-reported severity of OI						
Mild	55	52%	18	41%	37	60%
Mild to Moderate	27	25%	15	34%	12	19%
Moderate to Severe	11	10%	6	14%	5	8%
Severe	7	7%	3	7%	4	6%
N/A	6	6%	2	5%	4	6%
Type of OI						
Type I	52	49%	19	43%	33	53%
Type III	8	8%	5	11%	3	5%
Type IV	21	20%	8	18%	13	21%
Type V	3	3%	1	2%	2	3%
Other types [VIII, XI, XVII]	3	3%	1	2%	2	3%
Unconfirmed if have OI	4	4%	1	2%	3	5%
Unclassified Type	15	14%	9	20%	6	10%
DI Status						
Has DI	14	13%	6	14%	8	13%
Does not have DI	34	32%	15	34%	19	31%
Unknown	58	55%	23	52%	35	56%

4.2.4 COHIP- SF Questions

4.2.4.1 Internal consistency

Internal consistency of the questionnaire was checked using SPSS and calculating Cronbach's alpha. For the COHIP-SF section of the questionnaire the Cronbach's alpha was 0.821 indicating a high level of internal consistency.

4.2.4.2 Overview

Participants answered each question considering how they felt in the last three months. A score was assigned to each category (never = 0, almost never = 1, sometimes = 2, fairly often = 3, almost all of the time = 4). Scores for negatively worded questions were reversed to ensure consistency. Out of 19 questions, only two questions were positively worded:

- Have you ever been confident because of your teeth, mouth or face?
- Have you ever felt that you were attractive (good looking) because of your teeth, mouth, or face?

The remaining 17 questions were negatively worded and therefore the scores needed to be reversed.

The scores of each question were then added up to obtain a value for oral health related quality of life. The higher the score, the better the quality of life and thus a lower score reflected a lower quality of life. The maximum possible score was 76.

The COHIP-SF questionnaire is divided into three domains, Oral-Health, Functional Well-Being and Socio-Emotional Domain, reflecting an aspect of the child's life contributing to their overall OHRQoL. The Oral Health Domain contained questions pertinent to different concerns which could arise in the oral cavity including pain or discolouration. The Functional Well-Being domain included questions about whether their oral state affected their ability to function in their daily lives, such as affecting their ability to eat and speak. Finally, the Socio-Emotional Well-Being domain related to interactions with other children or effect on their mood such as being teased or feeling anxious.

4.2.4.2 Testing for Normality

To check whether the data was normally distributed, a Shapiro-Wilks normality test was carried out on the data and QQ plots were generated. This was done for the overall COHIP-SF scores and for each individual domain using SPSS. For the overall COHIP-SF scores, the functional well-being domain and the socio-emotional well-being domain the data was not normally distributed. However, the scores in the oral-health well-being

domain were normally distributed. Due to this, non-parametric Mann Whitney U tests were carried out for the overall COHIP-SF, functional well-being and socio-emotional well-being domains statistical analysis, whilst a t-test was used for the oral-health well-being domain. Table 4-3 below shows the Shapiro-Wilks test for each of the data sets.

Table 4-3 – Table showing tests for normality for overall COHIP-SF scores and within each domain.

Shapiro-Wilk Tests of Normality			
	Statistic	Degrees of freedom	Significance (P-Value) If <0.05 Data is not normally distributed
Overall COHIP-SF Scores	0.935	106	0.000
Oral health Well-being Domain	0.978	106	0.076
Functional Well-being Domain	0.890	106	0.000
Socio-emotional Well-being Domain	0.913	106	0.000

Figures 4-2, 4-3, 4-4 and 4-5 show the QQ plots for the overall COHIP-SF data as well as for each domain.

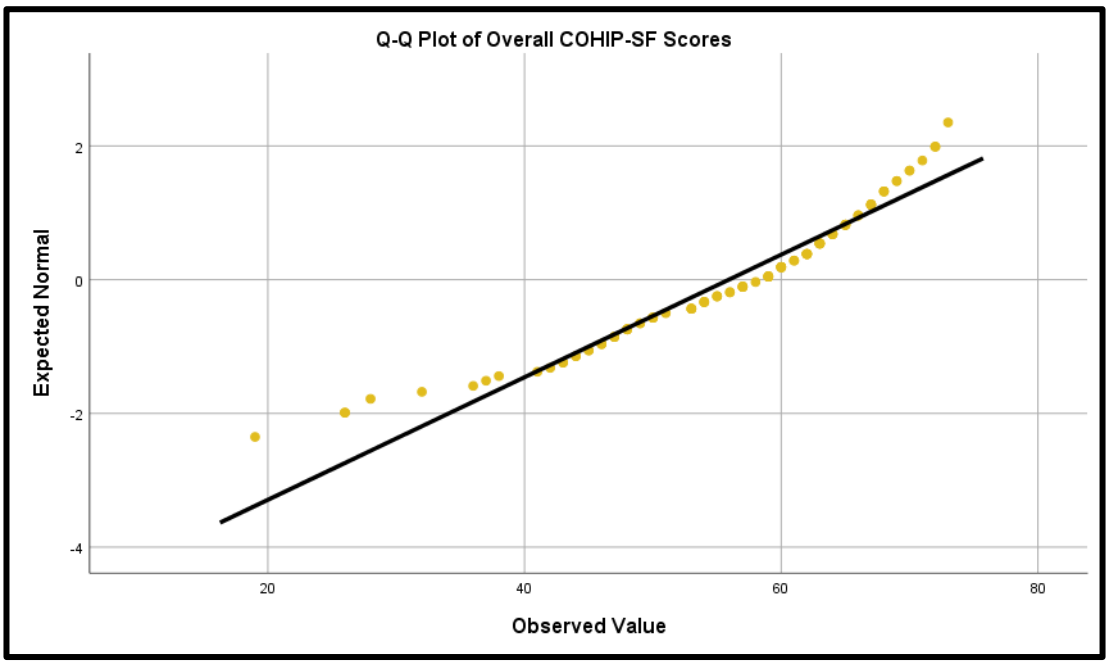

Figure 4-2 – QQ plot showing that overall COHIP-SF scores are not normally distributed

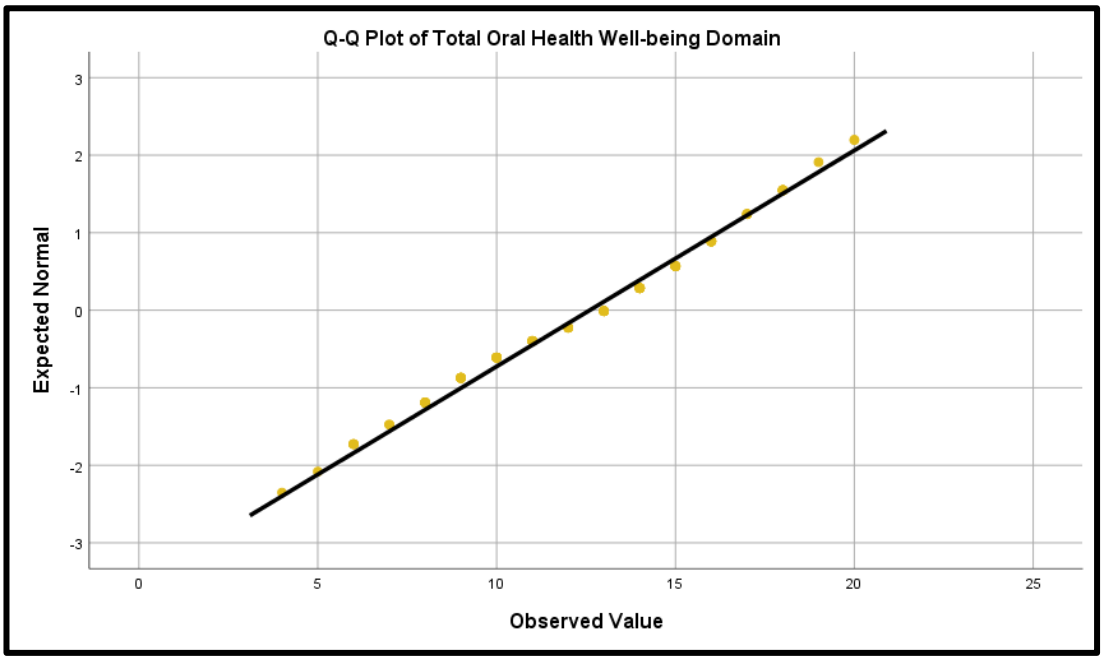

Figure 4-3 - QQ plot showing that total oral health well-being domain scores are normally distributed

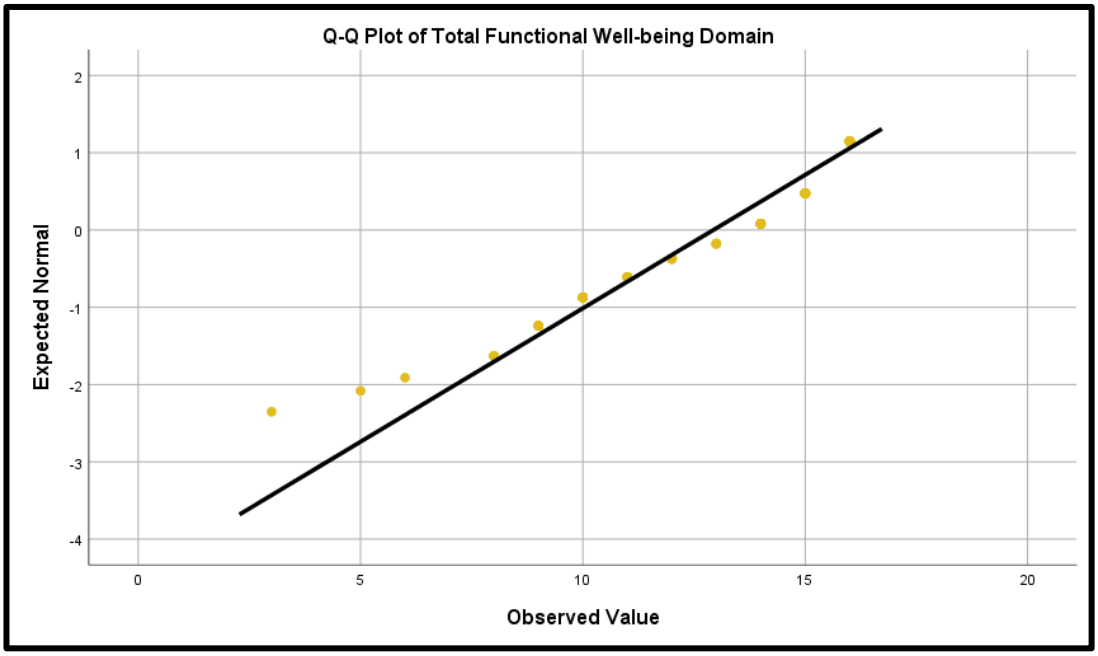

Figure 4-4 - QQ plot showing that overall total functional well-being domain scores are not normally distributed

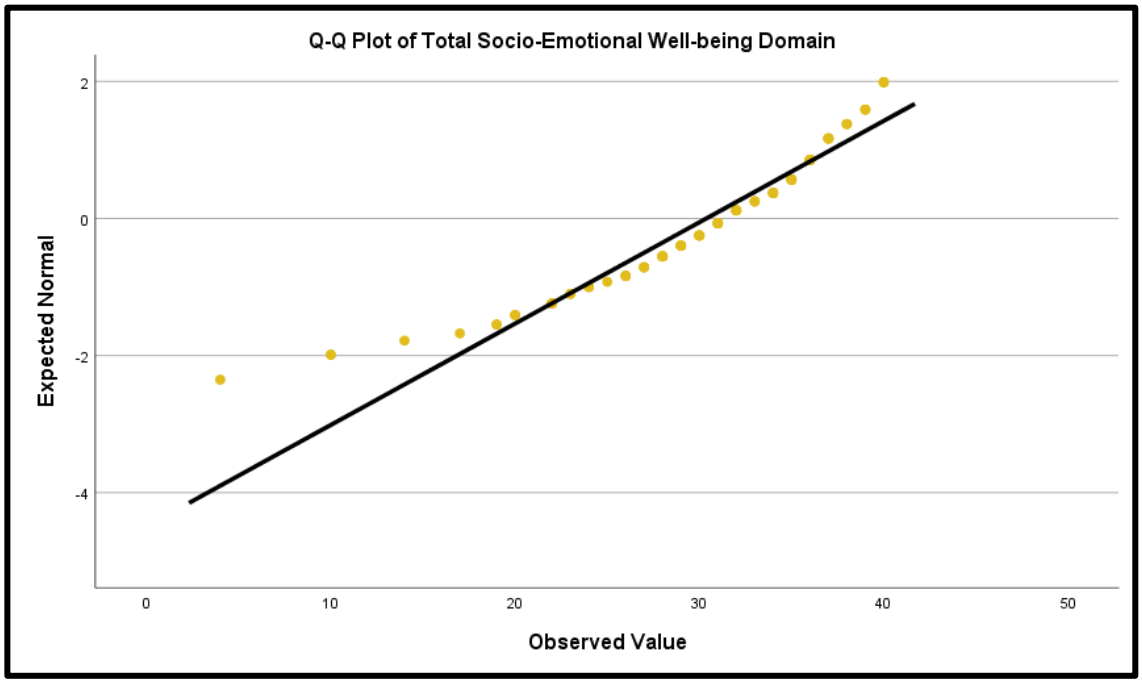

Figure 4-5 - QQ plot showing that total socio-emotional well-being domain scores are not normally distributed

4.2.4.3 Unanswered Questions

Out of a possible 2014 answers (19 questions for 106 participants) only 15 questions were not answered (<1%). Missing answers did not appear to show a bias towards age, gender or ethnicity however the number of missing answers in this section was too small for significant results. Further details can be found in Table 4-4 below.

Table 4-4 – Table showing details of unanswered questions

Category	Numbers
General	
Total number of participants not answering at least one question	12
Total number of unanswered questions	15
Participants not answering one question	9
Participants not answering two questions	3
Most frequently unanswered question: “Have you ever felt that you were attractive (good looking) because of your teeth, mouth, or face?”	5
Gender	
Females	5
Males	7
Ethnicity	
White	7
Asian	2
No answer	2
Black	1
Age groups	
12 years old or older	6
Younger than 12 years old	6
Self-reported Severity of OI	
Mild	3
Mild to Moderate	5
Moderate to Severe	2
Severe	1
No answer	1

4.2.4.4 Overall COHIP-SF Results

The total number of questions in the COHIP-SF was 19. The overall data in this group was not normally distributed as mentioned in section 4.2.4.2 above, therefore the Mann Whitney U test was used for data analysis. The median score was 59 with an interquartile range of 15. The COHIP-SF scores ranged from 19 at lowest to 73 at the highest.

Age was not statistically significant [p = 0.977] in the OHRQoL scores with children younger and older than 12 years old scoring similar average results. There was no statistically significant difference between the COHIP-SF scores between genders either,

p-value 0.155, however figure 4-6 below shows males score slightly better overall than females. Table 4-5 shows the statistical data for age groups and gender.

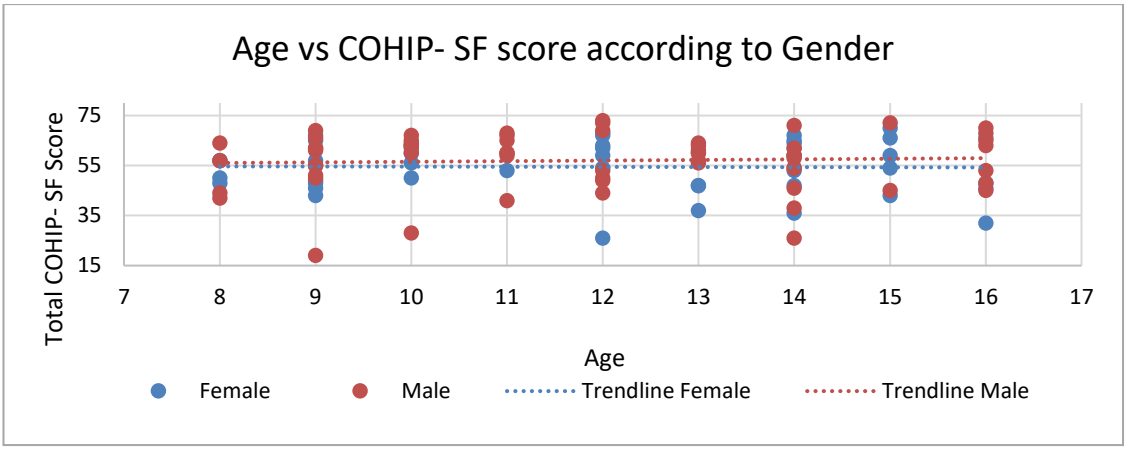

Figure 4-6 – Graph showing total COHIP-SF scores according to age and gender

Table 4-5 – Table showing statistical data for overall COHIP-SF for gender and age groups

	Numbers	Median	Interquartile Range
Gender			
Female	44	56.5	15.3
Male	62	60.0	12.5
Age Groups			
Younger than 12	46	58.0	13.0
12 years or Older	60	59.0	16.5

For further analysis, the data was divided according to subjective perception of the severity of their OI. Table 4-6 below shows the statistical data according to severity of OI.

Table 4-6 – Table showing statistical data for overall COHIP-SF by subjective severity of OI

Self-reported Severity of OI	Numbers	Median	Interquartile Range
Mild	55	62.0	12.0
Mild to Moderate	27	59.0	14.5
Moderate to Severe	11	57.0	11.5
Severe	7	55.0	18.0
No Answer	6	49.5	2.5

Using overall COHIP-SF scores, the participants perceiving a more severe OI had a lower COHIP- SF score with a score of median of 55 with an interquartile range of 18. Those claiming a mild type of OI had a median score of 62 and an interquartile range of 12. However, there was no statistically significant difference between the COHIP-SF scores of those who felt they had mild OI and those who felt they had severe OI [p = 0.087]. Females with mild OI had a median of 62, whilst males had a median of 61.

Table 4-7 – Table showing statistical data according to gender for self-reported severity of OI

	Female			Male		
	Number	Median	Interquartile Range	Number	Median	Interquartile Range
Mild	18	62.0	12.5	37	61.0	12.0
Mild to Moderate	15	54.0	12.5	12	60.0	12.8
Moderate to Severe	6	53.0	10.3	5	63.0	6.0
Severe	3	48.0	13.5	4	56.0	10.5
N/A	2	42.5	5.5	4	50.5	3.5

There was no statistical difference in COHIP-SF scores between the genders who self-reported as having mild OI [P = 0.767]. The same was true for those reporting severe OI with a P value of 0.857, however in this case the males had a slightly higher median (56) than the females (48). Overall, apart from those who self-reported as having mild OI, males scored higher than females as can be seen in Table 4-7. Figure 4.7 is a graph showing the distribution of COHIP-SF scores according to subjective severity of OI.

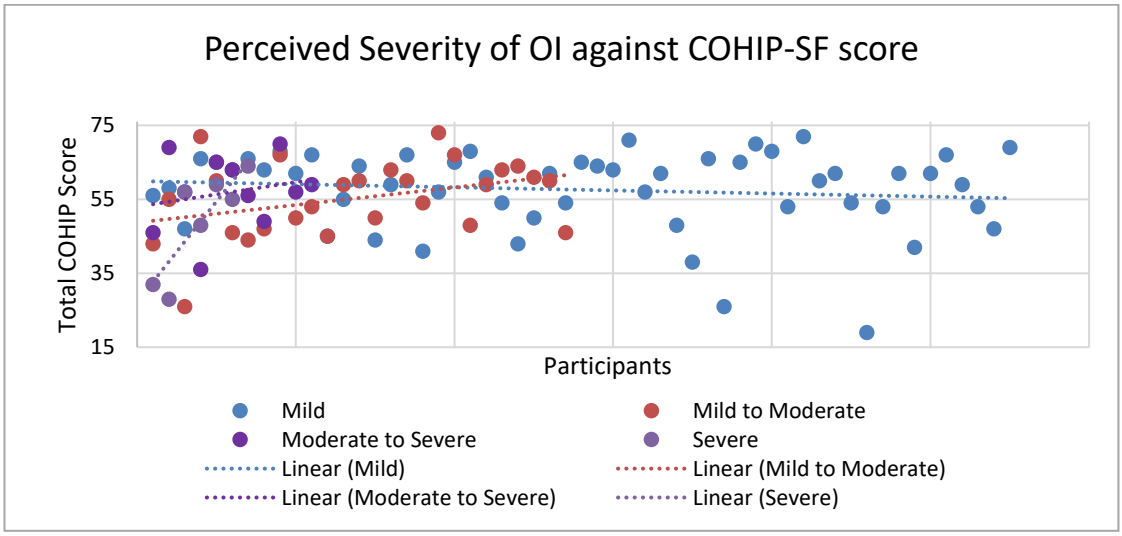

Figure 4-7 – Graph showing severity of OI vs COHIP-SF Scores

Age groups also did not show any statistically significant difference between mild and severe perception of OI. When comparing mild to severe in under 12-year olds, p = 0.191. In those children over 12 a p-value of 0.290 was calculated for mild against severe OI. Gender also did not show statistical significance between self-reported severity of OI. In females the p-value between mild and severe OI was 0.125, whilst for males this was p = 0.406.

From the types of OI in this cohort, type V had the highest median COHIP-SF score (67) whilst those whose OI types were unclassified and those with 'other' OI types [types VIII, XI, XVII] had the lowest medians (48.5 and 55 respectively). Table 4-8 below shows the statistical data according to type of OI.

Table 4-8 - Table showing statistical data for overall COHIP-SF by Type of OI

Type OI	Numbers	Median	Interquartile Range
Type I	52	59.0	15.5
Type III	8	58.0	10.8
Type IV	21	57.0	14.0
Type V	3	67.0	6.5
Other [Types VIII, XI, XVII]	3	55.0	12.0
Unconfirmed	4	60.0	11.0
Unclassified Type of OI	15	48.5	8.0

Figure 4.8 shows the range of total COHIP-SF scores for each of the types of OI as well as the median for each type of OI. From this graph, participants with type V OI had the

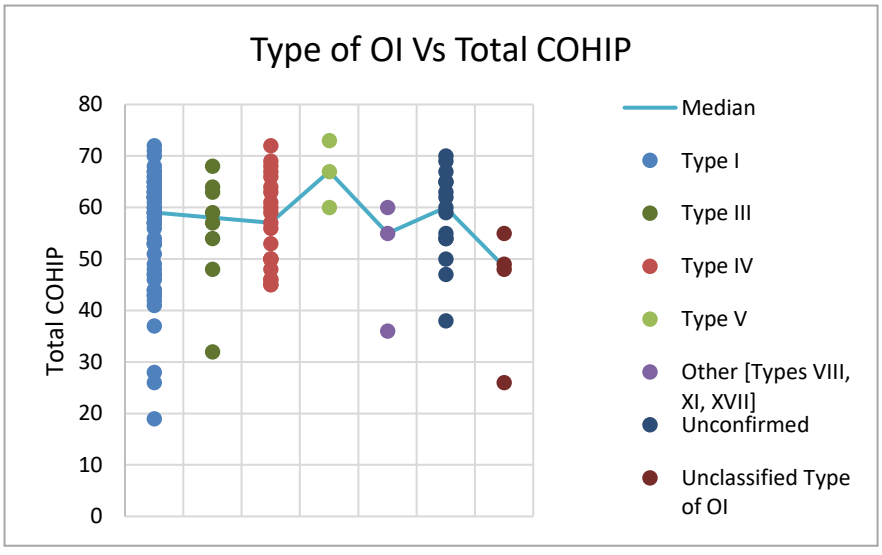

Figure 4-8 – Graph showing range and median of COHIP-SF scores for each type of OI

highest median whilst those whose OI type was unclassified scored the lowest. The difference however was not statistically significant [p = 0.614].

There was no statistically significant difference between mild or mild to moderate perception of OI and moderate to severe or severe perceptions of OI according to OI type. (Type I p-value = 0.195; Type III p-value = 0.393; Type IV p-value = 0.841; p-value could not be calculated for other types of OI due to insufficient numbers). There were limited numbers in some of the types of OI which could skew the p-value.

Figure 4-9 below shows the types of OI divided by subjective severity.

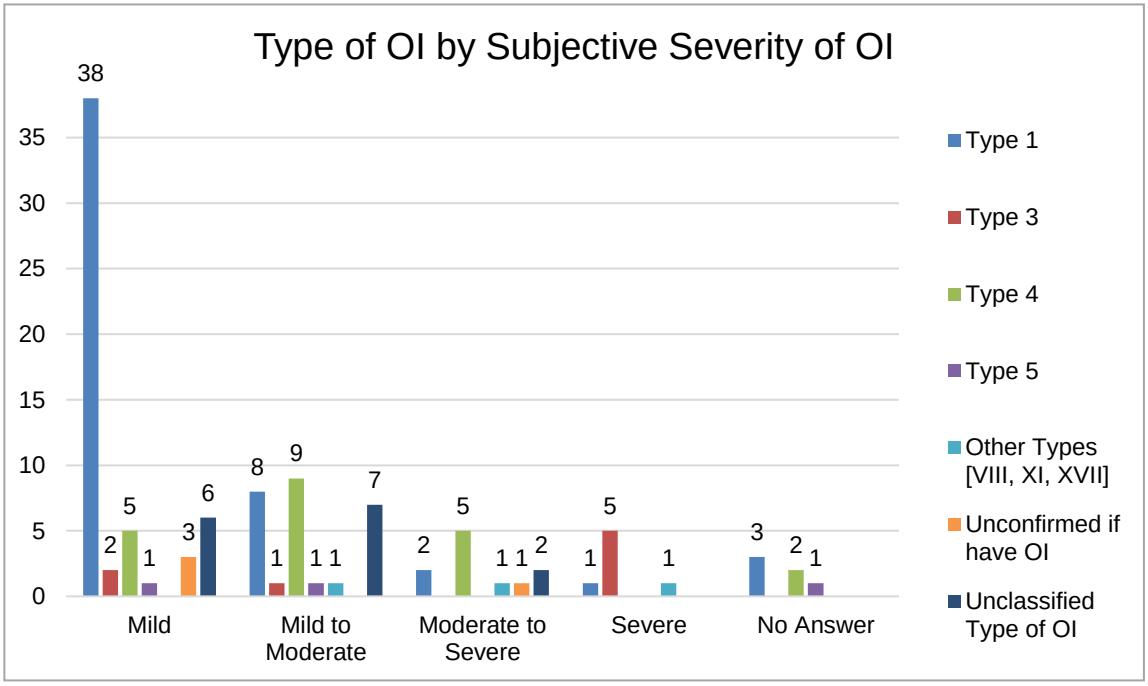

Figure 4-9 – Figure showing subjective severity of OI by type of OI

There was limited data about whether the participants had DI with only 13% (n=14) known to have DI. Thirty two percent (n=34) did not have DI and there was no data on the remaining 55% (n=58) of participants. Table 4-9 below shows the statistical data for the presence of DI.

Table 4-9 – Table showing statistical data for overall COHIP-SF depending on presence of DI

	Numbers	Median	Interquartile Range
No DI	34	60.5	17.5
DI	14	56.5	9.0
Unknown	58	57.5	15.3

Figure 4-10 shows which types of OI had DI

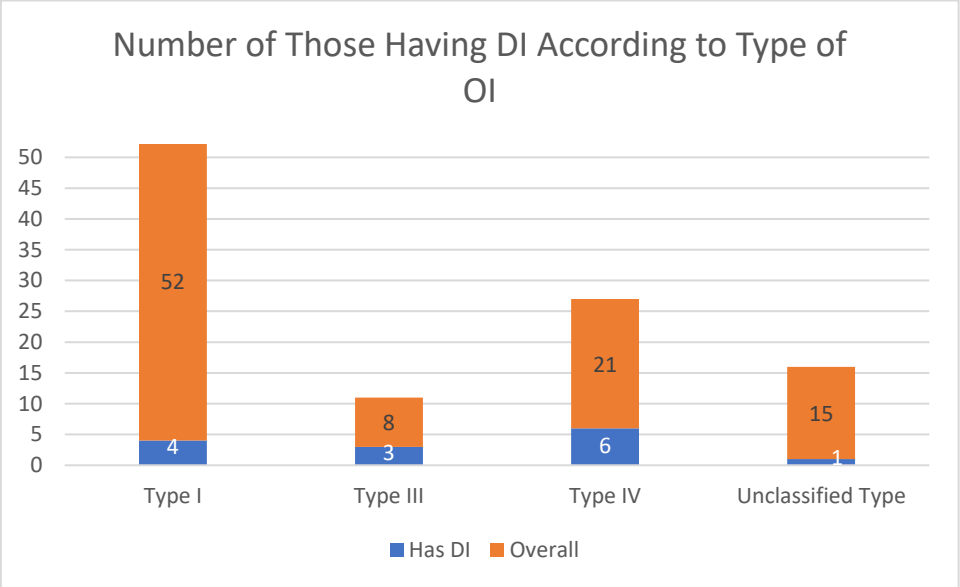

Figure 4-10 – Graph showing Number of Type of OI for those having DI

The presence of DI did not impact overall OHRQoL scores [P-value 0.109]. However, it did impact in individual domains and this will be discussed in later sections.

The question, 'Have you ever felt that you were attractive (good looking) because of your teeth, mouth, or face?' scored consistently low with 35% (n=37) choosing the lowest scoring response. Twenty eight percent (n=30) chose 'sometimes' and only 19% (n=20) chose the most positive response. Another question which achieved low overall scores was 'Have you ever had crooked teeth or spaces between your teeth?' with 29% (n=31) choosing 'almost all of the time', 21% (n=22) choosing 'sometimes' and 22% (n=23) choosing 'never'.

Questions which scored consistently high, with over 60% of participants choosing the most positive response included:

- 'Have you ever been unhappy or sad because of your teeth, mouth or face?'
- 'Have you ever felt worried or anxious because of your teeth, mouth or face?'
- 'Have you ever not wanted to speak/read out loud in class because of your teeth, mouth or face?'
- 'Have you ever avoided smiling or laughing with other children because of your teeth, mouth or face?'
- 'Have you ever had trouble sleeping because of your teeth, mouth or face?'
- 'Have you ever been teased, bullied or called names by other children because of your teeth, mouth or face?'

- ‘Have you ever felt that you look different because of your teeth, mouth or face?’
- ‘Have you ever been worried about what other people think about your teeth, mouth, or face?’

The question with the most positive responses was ‘Have you ever been teased, bullied or called names by other children because of your teeth, mouth or face?’ with 86% (n=91) of participants responding with ‘never’, 9% (n=10) responding with ‘almost never’ and only 5% (n=5) responding with ‘sometimes’. Figure 4-11 shows the responses of well scoring questions grouped into agree (Almost all the time/ fairly often), sometimes and disagree (almost never/ never).

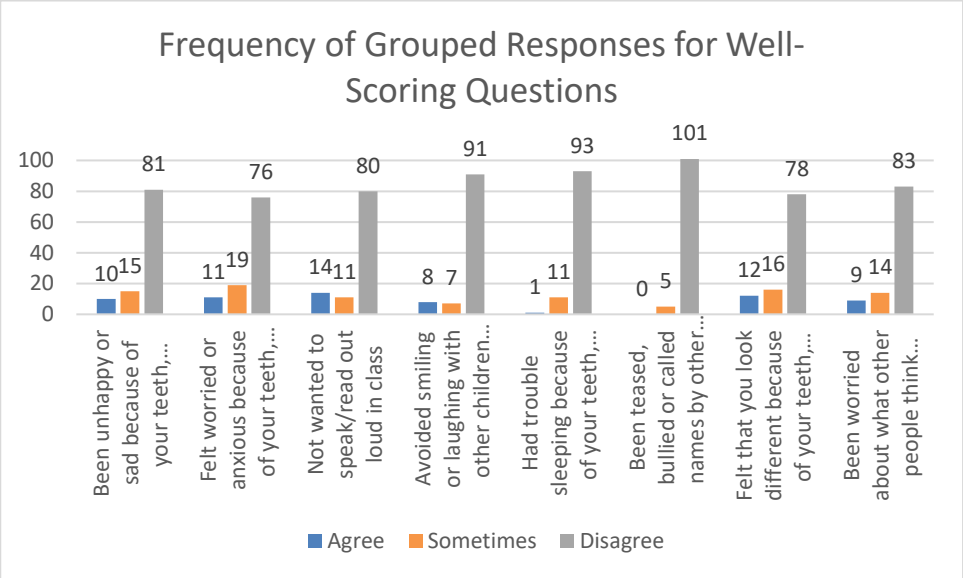

Figure 4-11 - Graph showing how high scoring questions scored overall

Children with OI commonly have DI or malocclusions. The breakdown of responses to the questions ‘Have you ever had crooked teeth or spaces between your teeth?’ and ‘Have you ever had discoloured teeth or spots on your teeth?’ according to type of OI can be seen in Table 4-10 below. In this table, the responses were grouped into:

- Agree/ sometimes when responses were almost all of the time, fairly often or sometimes
- Disagree when responses were almost never or never

Table 4-10 – Showing results for two questions by type of OI

	Had crooked teeth or spaces between your teeth	Had discoloured teeth or spots on your teeth
Type I (N = 52)		
Agree/ sometimes	71%	46%
Disagree	29%	54%
Types III/ IV (N = 29)		
Agree/ sometimes	69%	41%
Disagree	31%	38%
Type V (N = 3)		
Agree/ sometimes	67%	33%
Disagree	33%	67%

4.2.3.5 Oral Well-Being Domain

The oral well-being domain consisted of five questions. As the data in this domain was normally distributed, the t-test was used to calculate the p-values. The mean score for the Oral-Health domain of the questionnaire was 12.60 out of 20, with a SD of 3.59. The scores ranged from four to 20. The most frequently unanswered question was “Have you ever had crooked teeth or spaces between your teeth?” with two respondents failing to answer this question. This was also the lowest scoring question with 29% of participants (n=31) responding with almost all of the time. The highest scoring question in this domain was “Have you ever had bleeding gums” with 42% (n=44) responding with never. Table 4-11 shows the breakdown of questions by response and some basic data from the Oral Health Well-being Domain.

Table 4-11 – Table Showing breakdown of questions by response and basic data for Oral Health Well being domain

Question [Have you ever…]	Scoring (Score)- Scores reversed unless indicated otherwise					Statistical data			
	Almost all of the time (4)	Fairly Often (3)	Sometimes (2)	Almost Never (1)	Never (0)	Mean	Standard Deviation	Range	Number (n=106 unless otherwise specified)
Had pain in your teeth/Toothache	0	6	40	33	26	2.75	0.9	1 to 4	105
Had crooked teeth or spaces between your teeth	31	16	22	12	23	1.81	1.53	0 to 4	104
Had discoloured teeth or spots on your teeth	17	11	25	13	40	2.45	1.48	0 to 4	106
Had bad breath	1	9	33	25	38	2.85	1.04	0 to 4	106
Had bleeding gums	3	10	33	15	44	2.83	1.16	0 to 4	105
Total	N/A	N/A	N/A	N/A	N/A	12.6	3.59	4 to 20	n/a

There was no statistically significant difference between male and female scores for this domain [p = 0.161]. There was also no significant difference between children aged young than 12 or 12 years old and older [p = 0.083] however, there was a significant difference in the oral health well-being domain between those who had DI and those who did not [p = 0.033]. Table 4-12 below shows further information on these groups.

Table 4-12 – Table showing statistical information on gender, age and DI status for Oral Health Well-being Domain

	Numbers	Mean	Standard Deviation	P-Value
Gender				
Male	62	13.02	3.42	0.161
Female	44	12.02	3.78	
Age				
Younger than 12	46	11.91	3.37	0.083
12 years old or older	60	13.13	3.68	
DI status				
Has DI	14	11.57	3.57	0.033
Does not have DI	34	12.85	3.23	

When comparing OHRQoL scores in different ethnicities, there was little difference in the mean, SD and means as can be seen in Table 4-13 below. The greatest differences were between the Asian/ British-Asian group and the mixed ethnicities group however this difference was not significant [p = 0.085].

Table 4-13 – Table showing data according to ethnicity in Oral Health Well-Being Domain

Ethnicity	Numbers	Mean	Standard Deviation
White	57	12.44	3.75
Multiple/ Mixed ethnic group	3	16.00	3.46
Asian/ British-Asian	14	11.93	3.47
Black/ Caribbean/ Black-British	7	12.57	3.31
Other	9	14.44	2.74
No answer	16	12.13	3.46

When dividing according to subjective severity of OI, those with severe OI had the lowest mean scores, whilst those with mild OI had the highest mean scores, this difference was

not significant [p = 0.125]. The rest of the data for OHRQoL by severity of OI can be seen in Table 4-14.

Table 4-14 – Table showing data according to subjective severity of OI in the Oral Health Well-being Domain

Subjective severity of OI	Numbers	Mean	Standard Deviation
Mild	55	13.15	3.73
Mild to Moderate	27	12.74	3.21
Moderate to Severe	11	12.09	3.86
Severe	7	10.86	3.08
No Answer	6	10.00	3.03

When dividing according to type of OI, OI types V had the highest mean at 15.33. The lowest mean was in the group of those who's OI was unconfirmed (10.00). The difference between these two groups was significant [p = 0.026]. There was no statistical difference between OI types I and V in the Oral Health Well-being domain [p = 0.206]. Table 4-15 shows the data for types of OI in the Oral Health Well-being domain.

Table 4-15 – Table showing data according to type of OI in the Oral Health Well-being Domain

Type of OI	Numbers	Mean	Standard Deviation
Type I	52	12.44	3.85
Type III	8	12.63	4.07
Type IV	21	12.67	3.35
Type V	3	15.33	2.31
Other [Types VIII, XI, XVII]	3	11.00	4.36
Unconfirmed	4	10.00	2.16
Unclassified Type	15	13.53	2.92

4.2.3.6 Functional Well-Being Domain

The next domain is the functional well-being domain and consists of four questions. The data in this domain was not normally distributed therefore a Mann Whitney U test was used for data analysis. The median score in this domain was 14, with an interquartile range of four. The range of scores in this domain was from three to 16, the highest achievable score in this domain.

The highest score was achieved by 26 participants (25%), 16 males and 10 females with an average age of 12.85 years. The majority of these participants (n=16/26, 62%) self-reported as having mild OI, six reported mild to moderate OI (23%), one reported moderate to severe (4%), one severe (4%) and two (8%) did not answer this question. Only one participant who scored 16 in this domain was confirmed to have DI.

The participant who scored the lowest (3) was a 16 year old female with severe OI (type III) who does not have DI. The participant scoring the next lowest was a 9 year old male with type I OI and no DI who scored 5.

Two participants did not answer one question each in this section. These questions were ‘Have you had difficulty eating foods you would like to because of your teeth, mouth, or face?’ and ‘Have you had trouble sleeping because of your teeth, mouth, or face?’. The lowest scoring question was ‘Have you ever had difficulty keeping your teeth clean?’ with 7 participants (7%) choosing ‘almost all of the time’. In general, this section had high results with over 85% of participants choosing one of the three most positive responses for all four questions. The breakdown of this, as well as some statistical data can be seen in table 4-16 below.

Table 4-16 – Table showing breakdown of scores by question and statistical data for functional well-being domain

Question [Have you ever…]	Scoring (Score) - Reversed unless otherwise indicated					Statistical Data			
	Almost all of the time (4)	Fairly often (3)	Sometimes (2)	Almost never (1)	Never (0)	Median	Interquartile Range	Range	Number (n=106 unless otherwise specified)
Had difficulty eating foods you would like to because of your teeth, mouth, or face	4	6	22	10	63	4	2	0 to 4	105
Had trouble sleeping because of your teeth, mouth, or face	0	1	11	10	83	4	0	1 to 4	105
Had difficulty saying certain words	0	6	21	16	63	4	1.75	1 to 4	106
Had difficulty keeping your teeth clean	7	7	28	13	51	3	2	0 to 4	106
Total	N/A	N/A	N/A	N/A	N/A	14	4	3 to 16	N/A

There was no statistically significant difference between male and female scores for this domain [p = 0.716], although once again males scored slightly higher. There was also no significant difference between children aged young than 12 or 12 years old and older [p = 0.407]. Those who had DI and those who did not had quite different results with those having DI scoring considerably lower than those without. However, the difference was still not statistically significant [p = 0.982]. For those who were under 12, there was a significant difference between those who had DI and those who did not [p = 0.039]. For those 12 or older, there was no significant difference [p = 0.164]. Table 4-17 shows further information on these groups.

Table 4-17 – Table showing statistical information on gender, age and DI status for Functional Well-being Domain

Category	Numbers	Median	Interquartile Range	P-Value
Gender				
Male	62	14	5.00	0.716
Female	44	12.5	3.75	
Age Groups				
Younger than 12	46	13	5.00	0.407
12 years old or older	60	14	4.00	
DI Status				
Overall - Has DI	14	11	3.50	0.982
Overall - Does not have DI	34	14	3.75	
Less than 12 – Has DI	7	11	4.00	0.039
Less than 12 – does not have DI	16	14	2.00	
12 or older – Has DI	8	13	2.00	0.164
12 or older – Does not have DI	26	16	5.50	

When comparing OHRQoL scores in different ethnicities, the lowest scoring ethnicity was Asian/ British-Asian with a median of 11 whilst the highest scoring ethnicity was Multiple/ Mixed ethnicities with a median of 16, the difference between these two groups

was not statistically significant [p = 0.121]. Table 4-18 below shows statistical data according to ethnicity for this domain.

Table 4-18 – Table showing data according to ethnicity in Functional Well-Being Domain

Ethnicity	Numbers	Median	Interquartile Range
White	57	14	4
Multiple/ Mixed ethnic group	3	16	0.5
Asian/ British-Asian	14	11	5.5
Black/ Caribbean/ Black-British	7	14	4
Other	9	14	2
No answer	16	14	5

When dividing according to subjective severity of OI, those with severe OI had the lowest median scores, whilst those with mild OI had the highest median scores. The difference in OHROQoL scores between these two groups was not statistically significant for this domain [p = 0.335]. The rest of the data for OHRQoL by severity of OI can be seen in Table 4-19.

Table 4-19 – Table showing data according to subjective severity of OI in the Functional Well-being Domain

	Numbers	Median	Interquartile Range
Mild	55	14	4
Mild to Moderate	27	14	5
Moderate to Severe	11	13	4
Severe	7	11	4.5
No Answer	6	13	5

Type V OI had the highest median from the types of OI in this domain (16). For this domain, the lowest scores were for the other types of OI [types VIII, XI and XVII] with a median of 10. The difference between these two groups of OI was statistically significant with a p-value of 0.043. The rest of the statistical data for types of OI in this domain can be seen in Table 4-20.

Table 4-20 – Table showing data according to type of OI in the Functional Well-being Domain

Type of OI	Numbers	Median	Interquartile Range
Type I	52	13.5	5
Type III	8	14.5	2
Type IV	21	14	4
Type V	3	16	0
Other [Types VIII, XI, XVII]	3	10	1.5
Unconfirmed	4	11.5	3.5
Unclassified Type	15	15	2

4.2.3.7 Socio-emotional Well-Being Domain

The final domain is the socio-emotional well-being domain this is the largest section with ten questions. Here the data was once again not normally distributed and thus Mann Whitney U tests were carried out for data analysis. The median score in this domain was 31, with an interquartile range of 7.75 and a range from four to 40, the highest achievable score in this domain.

Only four participants (4%) achieved this score, three participants were male and one was a female, and the average age was 10.5 years. All self-reported as having mild OI. The types of OI included Type I, Type IV and unclassified type and none were confirmed to have DI.

The most frequently unanswered question from the COHIP-SF was in this domain and was the question ‘Have you ever felt that you were attractive (good looking) because of your teeth, mouth, or face?’ with five participants not responding to this question. The question ‘Have you been confident because of your teeth, mouth or face?’, which is along a similar theme was the second least answered question with three participants choosing not to answer this question. One participant did not answer both of these questions. These two questions were also the lowest scoring questions. For these questions the scores were reversed with a score of zero corresponding to ‘never’ and a score of four corresponding to ‘almost all of the time’. For the question ‘Have you ever felt that you were attractive (good looking) because of your teeth, mouth, or face?’, 37 participants (35%) chose never, whilst for the question ‘Have you been confident because of your teeth, mouth or face?’ had 19 participants (18%) chose never. The breakdown of this as well as some statistical data can be seen in table 4-21 below.

Table 4-21 – Table showing breakdown of scores by question for socio-emotional well-being domain (highlighted rows = positively worded questions)

Question [Have you ever…]	Scoring (Score) - Reversed unless otherwise indicated					Statistical Data			
	Almost all of the time (4)	Fairly Often (3)	Sometimes (2)	Almost Never (1)	Never (0)	Median	Interquartile Range	Range	Number (n=106 unless stated otherwise)
Been unhappy or sad because of your teeth, mouth, or face	4	6	15	16	65	4	1	0 to 4	106
Missed school for any reason because of your teeth, mouth or face	1	2	30	21	52	3	2	0 to 4	106
Been confident because of your teeth, mouth or face (Scores not reversed)	34	13	30	7	19	2	2.5	0 to 4	103
Felt worried or anxious because of your teeth, mouth, or face	7	4	19	12	64	4	2	0 to 4	106
Not wanted to speak/read out loud in class	7	7	11	15	65	4	1	0 to 4	105
Avoided smiling or laughing with other children because of your teeth, mouth, or face	4	4	7	11	80	4	0	0 to 4	106
Been teased, bullied or called names by other children because of your teeth, mouth, or face	0	0	5	10	91	4	0	2 to 4	106

Felt that you were attractive (good looking) because of your teeth, mouth, or face (Scores not reversed)	20	8	30	6	37	2	3	0 to 4	101
Felt that you look different because of your teeth, mouth, or face	5	7	16	12	66	4	2	0 to 4	106
Been worried about what other people think about your teeth, mouth, or face	8	1	14	12	71	4	1	0 to 4	106
Total	N/A	N/A	N/A	N/A	N/A	31	7.75	4 to 40	N/A

There was almost no difference in OHRQoL scores between males and females for this domain, the medians were 32 and 31 respectively. This was not statistically significant [p = 0.400]. Although children older than 12 scored slightly higher (median 33) than those younger than 12 (median 31), the difference was not statistically significant [P= 0.073]. In this cohort, participants who had DI had a lower score in this domain than those without and the difference was not statistically significant [p = 0.381]. Table 4-22 shows further information on these groups.

Table 4-22 – Table showing statistical information on gender, age and DI status for Socio-Emotional Well-being Domain

	Numbers	Median	Interquartile Range	P-Value
Gender				
Male	62	32	7	0.400
Female	44	31	9	
Age groups				
Younger than 12	46	33	7	0.073
12 years old or older	60	31	8.5	
DI status				
Has DI	14	28	6.25	0.381
Does not have DI	34	31	8	

When comparing OHRQoL scores in different ethnicities, again the lowest scoring ethnicity was Asian/ British-Asian with a median of 30, whilst the highest scoring ethnicity was Multiple/ Mixed ethnicities with a median of 34, however the difference between these two groups was not statistically significant [p = 0.197]. Table 4-23 below shows statistical data according to ethnicity for this domain.

Table 4-23 – Table showing data according to ethnicity in Socio-Emotional Well-Being Domain

Ethnicity	Numbers	Median	Interquartile Range
White	57	31	6
Multiple/ Mixed ethnic group	3	34	4
Asian/ British-Asian	14	30	14.5
Black/ Caribbean/ Black-British	7	29	6.5
Other	9	32	8
No answer	16	34	5.8

When dividing according to subjective severity of OI, those with severe OI had the lowest median scores, however, those with moderate to severe OI had the highest median scores. The difference in OHROQoL scores between the mild and severe OI cases was not statistically significant for this domain [p = 0.105]. There was a greater statistical difference between the severe and moderate to severe scores however this again was not statistically significant [p = 0.069]. The rest of the data for OHRQoL by severity of OI can be seen in Table 4-24.

Table 4-24 – Table showing data according to subjective severity of OI in the socio-emotional Well-being Domain

Severity of OI	Numbers	Median	Interquartile Range
Mild	55	32	8
Mild to Moderate	27	29	8
Moderate to Severe	11	34	5.5
Severe	7	29	6
No Answer	6	26.5	4

Once again, the type V OI group had the highest median (37) score from the types of OI in this domain. The lowest scores were in the group who were unconfirmed as having OI with a median of 28. The difference between these two groups of OI was not statistically significant with a p-value of 0.114. The rest of the statistical data for types of OI in this domain can be seen in Table 4-25.

Table 4-25 – Table showing data according to type of OI in the Socio-emotional Well-being Domain

Type of OI	Numbers	Median	Interquartile Range
Type I	52	32	8
Type III	8	31	5.75
Type IV	21	31	8
Type V	3	37	4.5
Other [Types VIII, XI, XVII]	3	30	7.5
Unconfirmed	4	28	10
Unclassified Type	15	32	7

4.2.5 Final Section

The third and final part of the questionnaire titled ‘The Last Few Questions’ included a series of seven questions, three qualitative questions, two dichotomous questions and two which were qualitative.

4.2.5.1 Additional questions

Table 4-26 shows the quantitative questions along with the breakdown of their scores and some statistical data.

Table 4-26 – Showing scores and statistical data for quantitative questions in the final section

Question	Scoring - Number (Percentage)					Median	Interquartile Range	Range	Number (n = 106 unless otherwise specified)
How much does the condition of your teeth, mouth, or face affect your life overall?	*Never (0)*	*Almost never (1)*	*Sometimes (2)*	*Fairly Often (3)*	*Almost all of the time (4)*				
	43 (40%)	34 (32%)	30 (28%)	5 (5%)	1 (1%)	1	1.5	0 to 4	103
How important is the health of your teeth and mouth?	*Not at all (0)*	*Not Much (1)*	*Fairly Important (2)*	*Quite a lot (3)*	*Very (4)*				
	2 (2%)	5 (5%)	16 (15%)	18 (17%)	64 (60%)	4	1	0 to 4	105
How often do you think about your teeth and smile?	*Never (0)*	*Rarely (1)*	*At Least Once or Twice a Month (2)*	*At Least Once or Twice a Week (3)*	*Every day (4)*				
	22 (21%)	27 (25%)	9 (8%)	13 (12%)	35 (33%)	2	3	0 to 4	106

Question 1: How much does the condition of your teeth, mouth, or face affect your life overall? For this question the majority of participants, 40% (n=43) responded with ‘never’, whilst only 5% (n=5) responded with ‘fairly often’ and 1% (n=1) responded with ‘almost all of the time’. As the total scores were quite small, the median and interquartile range did not differ greatly between comparison groups and can be seen in table 4-27.

Table 4-27 – Showing statistical data by category for the question 'How much does the condition of your teeth, mouth, or face affect your life overall?'

Category	Total	Median	Interquartile Range	Range
Gender				
Male	62	1	2.0	0 to 4
Female	44	1	2.0	0 to 3
Age Groups				
Younger than 12	46	1	2.0	0 to 4
12 years old or older	60	3	1.3	0 to 3
DI status				
Has DI	14	1	2.0	0 to 4
Does not have DI	34	1	1.0	0 to 3
Ethnicity				
White	57	1	1.0	0 to 3
Multiple/ Mixed ethnic group	3	0	0.5	0 to 1
Asian/ British-Asian	14	1.5	1.0	0 to 4
Black/ Caribbean/ Black-British	7	0	0.5	0 to 1
Other	9	0	1.0	0 to 2
No answer	16	1	2.0	0 to 3
Severity of OI				
Mild	55	1	1.0	0 to 3
Mild to Moderate	27	1	2.0	0 to 3
Moderate to Severe	11	1	1.0	0 to 2
Severe	7	1	3.0	0 to 4
No Answer	6	0	0.8	0 to 3
Type of OI				
Type I	52	1	1.0	0 to 4
Type III	8	1	1.0	0 to 3
Type IV	21	1	1.0	0 to 2
Type V	3	2	0.0	0 to 0
Other [Types VIII, XI, XVII]	3	1	1.0	1 to 3
Unconfirmed	4	0	0.5	1 to 3
Unclassified Type	15	1	1.0	0 to 2

The Mann Whitney U test was carried out on the COHIP-SF scores of those who felt that the condition of their oral cavity and face affected their life (fairly often/almost all of the time) and

those who did not (never). Those in the former group had a statistically significantly lower COHIP-SF score than those in the latter with a p-value was 0.025.
The data from this question was tested for normality using the Shapiro Wilks test and was found to be not normally distributed. When comparing scores for this question using Mann Whitney U tests, there was no significant difference between genders [p = 0.754], age groups [p = 0.254], DI status [p=0.196] or self-reported mild vs severe of OI [p = 0.321].

Question 2: How important is the health of your teeth and mouth?
As seen in table 4-26, over half the participants, 60% (n=64) felt that the health of their teeth and mouth was ‘very’ important. Conversely to the preceding question only 6% (n=6) responded with ‘not much’ or ‘not at all’ to whether they felt the health of their teeth and mouth was important. Statistical information for this question can be found in table 4-28.

Table 4-28 – Table showing statistical data for the question 'How important is the health of your teeth and mouth?'

Category	Total	Median	Interquartile Range	Range
Gender				
Male	62	4	1	0 to 4
Female	44	4	1	1 to 4
Age Groups				
Younger than 12	46	4	1	1 to 4
12 years old or older	60	4	2	0 to 4
DI status				
Has DI	14	3	2.0	1 to 4
Does not have DI	34	4	1.0	0 to 4
Ethnicity				
White	57	4	1	0 to 4
Multiple/ Mixed ethnic group	3	4	0	4 to 4
Asian/ British-Asian	14	4	2	0 to 4
Black/ Caribbean/ Black-British	7	4	1	2 to 4
Other	9	4	1.25	2 to 4
No answer	16	3.5	2	1 to 4
Severity of OI				
Mild	55	4	1	1 to 4
Mild to Moderate	27	4	1	0 to 4
Moderate to Severe	11	3	2	0 to 4
Severe	7	4	1.5	1 to 4
No Answer	6	4	2	2 to 4
Type of OI				
Type I	52	4	1.0	0 to 4
Type III	8	4	1.0	1 to 4
Type IV	21	4	1.0	0 to 4
Type V	3	3.5	0.5	3 to 4
Other [Types VIII, XI, XVII]	3	4	0.0	4 to 4
Unconfirmed	4	2.5	1.3	2 to 4
Unclassified Type	15	4	1.5	1 to 4

There was no statistically significant difference between the COHIP-SF score of those who felt oral health was important and those who felt it was less so [p-value = 0.120], however, those who felt their oral health was important had a lower median COHIP-SF score (57.5) than those who did not (63). The Shapiro Wilks test again confirmed that the results for this question were not normally distributed. The Mann Whitney U test was used to test significance between gender [p-value = 0.948], age [p-value = 0.187], DI status [p-value = 0.236] and self-reported severity of OI [p-value = 0.541]. None of these were found to be significant.

Question 3: How often do you think about your teeth and smile?

There was a fairly even distribution of how often participants thought about their teeth and smile with the most popular answer being ‘Everyday’, 33% (n=35) (Table 4-25). Statistical data for this question can be found in table 4-29.

Table 4-29 – Table showing statistical data for Question 3

Category	Total	Median	Interquartile Range	Range
Gender				
Male	62	1.5	4	0 to 4
Female	44	2	3	0 to 4
Age Groups				
Younger than 12	46	2	3	0 to 4
12 years old or older	60	2	3	0 to 4
DI status				
Has DI	14	3.5	3.000	0 to 4
Does not have DI	34	3	4.000	0 to 4
Ethnicity				
White	57	3	3	0 to 4
Multiple/ Mixed ethnic group	3	2	2	0 to 4
Asian/ British-Asian	14	3	2.75	1 to 4
Black/ Caribbean/ Black-British	7	0	1	0 to 4
Other	9	2	2	0 to 4
No answer	16	1.5	1.5	0 to 4
Severity of OI				
Mild	55	2	2.5	0 to 4
Mild to Moderate	27	3	3	0 to 4
Moderate to Severe	11	1	2.5	0 to 4
Severe	7	3	2.5	0 to 4
No Answer	6	4	3	0 to 4
Type of OI				
Type I	52	2	2.8	0 to 4
Type III	8	3	2.0	0 to 4
Type IV	21	1	4.0	0 to 4
Type V	3	2	2.0	0 to 4
Other [Types VIII, XI, XVII]	3	0	2.0	0 to 4
Unconfirmed	4	2	2.5	0 to 4
Unclassified Type	15	2	2.5	0 to 4

There was a significant difference in COHIP-SF scores between those responding with 'Everyday' and those responding with 'never' [P-value = 0.040]. Those who thought about their teeth more often scored lower (median = 55) to those who never did (median = 60.5).

The answers to this question were not normally distributed (Shapiro Wilks Significance Value – 0.000) and again the non-parametric Mann Whitney U test showed no significant difference between genders [p-value = 0.207], age [p-value = 0.982], DI status [p-value = 0.472] and perceived severity of OI [p-value = 0.358].

Questions 4 and 5:
Have you talked with your family or other important people in your life about how you feel about your teeth and smile?
Have you talked with the dentist or doctor about how you feel about your teeth and smile?

Table 4-30 – Showing statistical data for yes/no questions

Question	**Scoring - Number (Percentage)**			**Number (n = 106 unless otherwise specified)**
Have you talked with your family or other important people in your life about how you feel about your teeth and smile?	*No*	*Yes*	*No Answer*	
	68 (64%)	33 (31%)	5 (5%)	101
Have you talked with the dentist or doctor about how you feel about your teeth and smile?	*No*	*Yes*	*No Answer*	
	68 (64%)	37 (35%)	1 (1%)	105

The final two questions in this section dealt with communication with family or medical and dental professionals about the participant's feelings regarding their teeth and smile. The majority of participants did not speak to anyone about this with 64% (n=68) responding with 'no' to both questions. In both cases, the male to female ratio in responses was similar.

Those participants who spoke to their families about their teeth and smile scored lower than those who did not in the COHIP-SF and this difference was significant. Whilst those who spoke to their dentist also scored lower than those who did not, the difference was not significant. Statistical data can be seen in table 4-31.

Table 4-31 – Showing data for questions 4 and 5

				COHIP-SF Data		
	Overall (numbers)	**Males - N (% of response)**	**Females - N (% of response)**	**Median**	**Interquartile Range**	**P-Value**
Have you talked with your family or other important people in your life about how you feel about your teeth and smile?						
Yes	33	16 (48%)	17 (52%)	53	17	0.001
No	68	43 (63%)	25 (37%)	60	10	
Have you talked with the dentist or doctor about how you feel about your teeth and smile?						
Yes	37	19 (51%)	18 (49%)	53	16	0.104
No	68	42 (62%)	26 (44%)	59	11	

4.2.5.2 Qualitative Questions

Two qualitative questions ended the questionnaire.

Question 1: What one change to your teeth or smile would make the biggest difference in your life? And how would things be different for you?

The first question was answered by 61% of participants (n=65). The answers provided by the participants were analysed and divided into 5 themes:

- Aesthetics
- Confidence
- Function
- Hygiene
- Pain or Caries

The aesthetic theme was further subdivided into:

- Orthodontic concerns (n=31)
- Discolouration (n=9)
- Both orthodontic and colour concerns (n=6)
- Others (n=4).

Several participants, n=4 (4%) mentioned more than one change they would wish for. The number of participants within each theme can be seen in Table 4-33. Table 4-32 shows some quotes for each theme and subtheme.

Table 4-32 – Showing quotes for patients by theme and subtheme

Theme	Subtheme	Patient ID	Quotes
Aesthetics	Colour	19	Them to be one colour
		41	Multi-coloured
	Orthodontics	11	To make them less crooked, gappy and align all my teeth
		99	I would like straight teeth rather than wonky
	Both Orthodontics and Colour	17	I would want them coated white and have braces
		23	Whitening. no gaps. straighter
	Other	39	The chip on my lower incisor
Confidence		26	To make my teeth connect properly. It would be different because I would be more confident in smiling
Function		48	Healthy and strong teeth would be very important in order to be able to eat what I like and not have to worry about damaging my teeth
Hygiene		74	To be cleaner
Pain or Caries		18	It would make a difference if my teeth would stop hurting. I would be able to eat properly
Multiple Themes	Pain and Aesthetics	6	I don't want fillings so teeth don't hurt, I want my teeth to grow quicker. I want my side tooth to grow straight so I don't have weird teeth
	Aesthetics and confidence	43	Braces to straighten my teeth. They make me self-conscious sometimes

When comparing COHIP-SF scores of those participants who gave a response and those who did not, those responding had a lower median COHIP-SF score (55) than those who did not (60) and the difference was significant [p-value = 0.035]. Statistical data for this question can be seen in Table 4-33. Those participants fitting into multiple themes had the lowest COHIP-SF scores. For three participants the themes were aesthetics and confidence and the themes

for the fourth participant were aesthetics and pain. These four participants were males whose COHIP-SF scores were 55, 46, 41 and 38, all below the overall median (59). Two self-reported as mild OI and two as mild to moderate OI. For two participants, the type of OI was unclassified and was Type I OI for the other two.

Overall, from the 14 participants known to have DI, 7 had aesthetic concerns and 1 has confidence issues.

Table 4-33 – Showing statistical data for the first qualitative question

			COHIP-SF		
	Numbers	**Percentage**	**Median**	**Interquartile Range**	**P-Value**
No Answer	41	39%	60.0	11.0	0.035
Total answered	65	61%	55.0	16.0	
Aesthetic	50	47%	57.0	15.5	
Confidence	2	2%	63.0	3.0	
Function	4	4%	59.0	11.8	
Hygiene	2	2%	47.5	4.3	
Pain/ Caries	3	3%	68.0	20	
Multiple	4	4%	43.5	5.8	

From the 40 participants (38%) whose aesthetic concern was orthodontics (both orthodontics alone or orthodontics and colour), there was data on DI status for 17 of them. Overall, the median for orthodontic concerns was 54, for those with orthodontic concerns and DI it was 50 and for those with orthodontic concerns but no DI it was 60 and the difference was not significant [p-value = 0.245]. When carrying out the same comparisons for those who mentioned colour as a concern, the median for those with DI was higher (50) than those without DI (40.5). This difference was not significant [p-value = 0.730].

Table 4-34 gives more data on the orthodontic and discolouration concerns.

Table 4-34 – Showing Data for orthodontic and discolouration concerns

	Orthodontic concerns	Discolouration concerns	Both orthodontic and discolouration concerns
Number	40	16	6
Median	54.0	52.0	47.5
Males (N)	20	8	4
Females (N)	20	8	2
Average Age	11.9	12.0	12.3
Severity of OI			
Mild	23	7	2
Mild to moderate	10	3	2
Moderate to severe	5	3	2
Severe	1	1	0
No answer	1	2	1
Type OI			
Type I	21	7	1
Type III	2	2	1
Type IV	10	5	2
Type V	1	0	0
Other [Types VIII, XI, XVII]	1	1	1
Unconfirmed	2	0	0
Unclassified Type	3	1	0
DI Status			
Yes	4	5	2
No	13	4	2

When dividing according to the COHIP-SF domains, there was no significant difference between those who responded and those who did not in any of the domains. This is shown in table 4-35.

Table 4-35 – Data for Question 1 according to COHIP-SF domains

	Median	Interquartile Range	P-Value
Oral Health Well Being Domain			
Responded	13	6	0.755
No Answer	14	6	
Functional Well Being Domain			
Responded	14	4	0.875
No Answer	14	6	
Socio-Emotional Well Being Domain			
Responded	31	9	0.155
No Answer	42	6	

Question 2: 'Is there anything else about your teeth, mouth or face that you think is important? Please tell us what it is.

The second question was less well responded with 78% (n=83) not answering the question. There was a significant difference between COHIP-SF scores of those who responded and those who did not [p-value = 0.030] with those responding having a lower COHIP-SF score. For this question, the responses were again divided into themes. The themes and quotes for each theme can be seen in Table 4-36.

Table 4-36 – Showing quotes for each theme for question 2

Theme	Patient ID	Quotes
Aesthetics	11	Brings my chin forward and looks displeasing when I smile
Hygiene	32	Cleaning properly
	35	About your breath
Pain/ Caries	41	Pain, very much pain
Confidence	58	That you see feel good about them
Other	74	Gag reflex

The most commonly occurring themes were aesthetics and keeping their teeth clean (hygiene). Table 4-37 below shows statistical data according to theme. 'Other' had the lowest overall COHIP-SF median score, from the two respondents one felt his gag reflex was important, and the second that his teeth were taking too long to 'grow'.

Table 4-37 – Showing statistical data for second qualitative question

			COHIP-SF Scores		
	Number	**Percentage**	**Median**	**Interquartile Range**	**P-Value**
No Answer	83	78%	59.0	11.0	0.030
Total Answered	23	22%	49.0	14.0	
Aesthetics	9	8%	50.0	17.0	
Hygiene	9	8%	53.0	12.0	
Pain/ Caries	2	2%	48.5	20.5	
Confidence	1	1%	48.0	0.0	
Other	2	2%	46.0	2.0	

Table 4-38 shows further statistical data for question two. It shows numbers for those who did and did not respond to 'Is there anything else about your teeth, mouth or face that you think is important? Please tell us what it is' regarding gender distribution, self-reported severity of OI, type of OI and DI status. More males than females responded to this question. There was no bias towards self-reported severity of OI or type of OI. There was no significant difference between those responding who had DI and those who did not have DI [p-value = 0.659].

Table 4-38 – Table showing data for question 2

	Responded	No Answer
Number	23	83
Median	49	59
Males (N)	14	48
Females (N)	9	35
Average Age	11.7	12
Severity of OI		
Mild	13	42
Mild to moderate	5	22
Moderate to severe	2	9
Severe	3	4
No Answer	0	6
Type OI		
Type I	9	43
Type III	1	7
Type IV	5	16
Type V	0	3
Other [Types VIII, XI, XVII]	3	0
Unconfirmed	3	1
Unclassified Type	2	13
DI Status		
Has DI	3	11
Does not have DI	11	23

4.3 Discussion

Children with Osteogenesis Imperfecta (OI) have co-morbidities including dental issues such as Dentinogenesis Imperfecta (DI) and malocclusion. The service evaluation of children attending the OI clinics at Great Ormond Street Hospital (GOSH) confirmed that children have dental needs, which emphasised the importance of assessing Oral Health Related Quality of Life (OHRQoL) in these children which had not been explored previously.

This study began in December 2017, but data collection did not start until January 2019. The reason for this delay was that obtaining ethical approval took almost one year due to the changes happening with the implementation of GDPR in 2018. Once ethical approval was obtained in November 2018, it took a further 2 months to obtain approval and a research contract from the local research and development department at GOSH.

The importance of evaluating OHRQoL for OI is gaining recognition around the world, as evidenced by a Canadian study from the Shriner's Children's Hospital (Najirad et al., 2018) which came out late in October 2018. This study assessed 138 children with OI using the CPQ 8-11 and CPQ 12-14. There is no mention of whether the questionnaire was piloted, and severity was assumed depending on type of OI. Participants were recruited over two years through the Brittle Bone Consortium which is a group of centres specialising in OI across North America. The sample size is similar to our study, we recruited 106 participants over one year. The types of OI seen in the Montreal study were types I, III, IV, V and VI. Comparisons with our study will be discussed in the relevant sections.

4.3.1 Demographics

As part of the discussion we wanted to see if our sample was representative of the OI population and so we wanted to compare variables such as gender, severity of OI, types of OI and DI. We also asked participants about ethnicity; however, we were unable to compare this to the Montreal study as they did not assess ethnicity (Najirad et al., 2018). OI has not been seen to discriminate between ethnicity or race (Marini et al., 2015). In this study 31% of the cohort did not identify as white. When comparing this proportion to the United Kingdom (UK) it showed a high proportion of ethnic minority groups in this study. In the UK the proportion of people who do not identify as white is 13% (GOV.uk, 2020). The large proportion of minority groups may affect the overall results of the questionnaires as people from different ethnicities may have different ideas or priorities on what affects their quality of life.

Males and females are equally likely to be affected by OI (Dahan et al., 2016). This was reflected in our sample of OI patients, 58% of whom were male.

It was interesting to see that self-reporting did seem to correlate with severity. For example, 75% of children with type I OI said they had mild OI, and likewise the majority of children with type III reported severe OI. To the best of my knowledge there are no other studies correlating patient perceived severity of OI to type of OI. This is a gap in the literature which may impact both OHRQoL and HRQoL.

The rarer types of OI (Types VI-XX) make up around 12% of cases, whereas in our sample it made up only 3% (Marini et al., 2017). However, these discrepancies may be because of the small sample size (n = 106) and those with unclassified or unconfirmed OI in 18% of our cohort.

In this study 13% of the participants had Dentinogenesis Imperfecta (DI) which is lower than the overall prevalence of DI (22-25%) reported in two recent studies (Najirad et al., 2018; Hald et al, 2018). However, there was missing data on DI for 55% of participants, so this proportion must be interpreted cautiously. Ideally, we should have asked participants whether they had DI or not. For those participants whose DI status was known, this was found by inputting patient names into the electronic records system at the Eastman Dental Hospital (EDH) and checking if a diagnosis of DI was established or discarded. One possible way to identify the DI status in the remaining patients would have been to obtain the details from their medical records, however, due to COVID-19, access to the participants details was limited as they were locked in a safe room which I could not access during the data analysis stage. All cases with DI had OI types I, III or IV, except for one participant who had DI but did not have their OI type confirmed.

4.3.2 COHIP-SF

This study used the short form of the Child oral health impact profile (COHIP-SF). The questionnaire asked participants to reply to each question referring to how they felt in the last three months. This was clearly stated in large fonts on the cover sheet in an example question. An error in the wording on the third page was noted, asking participants how they felt in the past year rather than the last three months. This was not picked up by the researcher, research team, the Young Persons Advisory Group or most importantly any of the participants. Given that none of the participants queried this, probably because the example was much more

prominent than the small print preceding the actual questions, in all probability the answers referred to the previous three months rather than one year.

This questionnaire, or the longer form of the same questionnaire, has been used by other researchers to assess the oral health related quality of life in children, including malocclusions (Broder et al., 2012; Anthony et al., 2018), and a high caries population (Genderson et al., 2013b), Cystic Fibrosis (Patrick et al., 2016), Orofacial Clefts (Bos and Prahl, 2011; Eslami et al., 2013; Agnew et al., 2017), and the general paediatric population (Broder et al., 2012).

The COHIP and COHIP-SF questionnaires ask researchers to reverse scores for negatively worded questions so that a higher score reflects a better oral health related quality of life. This was a particularly tricky part of data analysis as all questions except two in the COHIP-SF are negatively worded. This means 17 questions needed recoding. One would expect that for ease of use the two positively worded questions would have their scores reversed and a lower score would mean a better OHRQoL. This part of the analysis requires particular attention to understand and has not always been done well by other researchers. Anthony et al. in 2018 used COHIP-SF to assess malocclusion, however they reversed positively worded questions and stated a lower score meant a lower OHRQoL. This makes comparison of results with this study difficult as their numbers are completely reversed to those in this study (Anthony et al., 2018). Other studies using the long form of the COHIP have also chosen to analyse their data in the same was as Anthony et al., with a lower score reflecting a better OHRQoL (Patrick et al., 2016; Eslami et al., 2013). Two studies chose to reverse positively worded questions then recode, meaning high scores indicated a better OHRQoL (Agnew et al., 2017; Genderson et al., 2013) making comparison easier.

4.3.2.1 Age

As in other studies using the COHIP or COHIP-SF to assess OHRQoL (Patrick et al., 2016; Agnew et al., 2017), the participants were divided into two age groups: those younger than 12 years old and those aged 12 and older. These groupings were chosen to broadly reflect those children in the mixed dentition phase and those in the permanent dentition phase.

The trend amongst medically compromised children seems to be a lower OHRQoL as they get older. One possible reason is that older children are more aware of their facial appearance and how it differs from that of their peers. A similar trend has been seen in children with orofacial clefts (Agnew et al., 2017; Bos et al 2011), cystic fibrosis (Patrick et al., 2016) and malocclusion (Anthony et al., 2018). In this study the median score in this OI cohort was slightly higher in the older group this was only by 1 score (58 vs 59) and was not significant [P-value

0.977]. This could perhaps be because of a small sample size or due to the fact that children with OI often have dental concerns from a young age, particularly if they have DI in the primary dentition. Further work needs to be done on this.

Najirad et al. (2018) also measured OHRQoL in children with OI but using a different questionnaire: the CPQ for ages 8-10 and 11-14 years old. They found that older children had significantly lower scores (Indicated poorer OHRQoL) if they had severe OI, compared to those with mild OI, while there was no difference in the younger cohort. The CPQ for ages 8-10 and 11-14 have different numbers of items so they were not able to compare between age groups. The COHIP-SF allows us to compare groups, we found that OHRQoL scores were not significantly different between age groups. When comparing types of OI within age groups, there was no significant difference for our population. However, in the older group, those with type III OI did score slightly lower than those with type I OI, because there were only 5 patients with OI type III in this group (Severe), the results may not be representative.

4.3.2.2 Gender

Unlike the Montreal study (Najirad et al., 2018), we assessed the effects of gender. and found that there was no significant difference in COHIP-SF scores, overall [P-value 0.155], or within the different COHIP domains. Similarly, Bos et al, Agnew et al, and Patrick et al did not find a significant difference in OHRQoL scores between genders for cleft lip and palate and cystic fibrosis patients (Bos and Prahl, 2011; Patrick et al., 2016; Agnew et al., 2017). There was a significant difference between genders for malocclusion patients in a study conducted on patients in Zambia by Anthony et al. (2018) with females scoring lower than males overall. This could be due to increased aesthetic awareness in females with significant malocclusions (Anthony et al., 2018).

4.3.2.3 Self-Reported Severity of OI

As previously mentioned, participants were asked how they perceived the severity of their OI from mild to severe. Generally, those with mild OI had a better, but insignificant, overall COHIP-SF score than those with severe OI. Reasons for this could be that the sample size of those perceiving their OI as severe was small (n = 7) compared to those perceiving their OI as mild (n = 55). There was no significant difference with those reporting mild to moderate or moderate to severe OI either.

It was surprising that there was not significant difference between those with mild and severe OI and shows that severity of OI may not necessarily relate to severity of dental concerns. Children with OI have other complex social factors affecting their life such as their ability to play with their peers, frequent visits to hospitals or clinics. Fear of fractures or feeling a lack of

independence may colour the way a child perceives their OHRQoL, particularly if they feel different to their peers and this may account for why severity of OI is not significantly correlated to overall OHRQoL.

Within the COHIP-SF domains, those with severe OI always scored the lowest, however, this difference was not significant. Interestingly, those who did not answer how they perceived the severity of their OI (n = 6) tended to score even lower than those with severe OI, in all but the functional well-being domain. It could be that those with more severe OI are less comfortable talking about their condition or it could be a coincidence because of the small sample size, this should be viewed with caution. In the socio-emotional well-being domain, those with self-reported moderate to severe OI scored slightly higher overall than those with mild OI. This could be because given the moderate severity of their OI, the emotional support given to these children is sufficient to address their socio-emotional concerns. It is well documented that a good social and emotional support system can increase health-related quality of life (Strine et al., 2008), the same may hold true for OHRQoL however further research needs to be done about this.

4.3.2.4 Type of OI

There was no significant difference in COHIP-SF scores between type I OI (mild) and types III and IV (severe), but type V OI had the highest scores, both overall and across all domains. Type V is generally held to be a moderate type of OI without DI, although malocclusions and crossbites are common in this type (Retrouvey et al., 2018). In this cohort, there were only 3 participants with type V OI. These were all in the older age group and all of these individuals self-reported their severity as mild or mild to moderate. The small number of participants with type V OI may not be representative of the entire type V OI population. The lowest scoring type for overall COHIP-SF score were those whose type of OI was unclassified. These are likely to have a variety of types from type V to type XX and the diagnosis can only be determined by genetic testing. Those participants who were unconfirmed for OI scored the lowest in the oral health and socio-emotional well-being domains. However, from those whose diagnosis of OI was confirmed, all those with other types of OI [Type VIII, IX, XVII] scored the lowest in all three domains. As mentioned above, the other types of OI tend to be more severe and this could account for their low scores (Marini et al., 2017). The literature also tells us that anecdotally, those whose OI is unclassified have higher bone fracture rates than those whose type of OI has been determined (Patel et al., 2015).

4.3.2.5 DI Status

COHIP-SF scores were also compared between those who had DI and those who did not. It appeared that DI did not significantly impact overall OHRQoL scores. However, in the oral

health well-being domain, there was a significant difference between those who had DI and those who did not. This makes sense as it would be expected that questions such as 'Have you ever had discoloured teeth or spots on your teeth?' would have scored low due to the discolouration seen in DI. From those patients who had DI, the majority (43%) agreed that they felt this way fairly often or almost all of the time, and a further 36% felt this way sometimes. This confirms that participants with DI did notice the discolouration of their teeth. In the functional well-being domain, there was no significant difference between those with DI and those without, however, those in the under 12 year old age group scored significantly lower if they had DI than if they did not. Again, this would make sense as DI is more common in the primary dentition and the loss of tooth structure may have an effect on the oral functioning of these children. Again, this needs to be viewed with caution because there were only 14 participants who had DI. This is a limitation as we should have known who in our sample had DI, but it is worth noting that in the only other study looking at OHRQoL in children with OI, even though they had a higher proportion of children with DI, they did not compare between groups.

4.3.2.6 Additional Information on Overall COHIP-SF Results

Two questions of particular importance for dental issues in this population are 'Have you ever had crooked teeth or spaces between your teeth?' and 'Have you ever had discoloured teeth or spots on your teeth?'; as children with OI commonly have DI and malocclusion For both questions, the majority of children felt they had discoloured (50%) or crooked or spaced teeth (65%) at least some of the time.

From the eight questions scoring consistently high (over 60% of the participants chose the most positive score), seven were from the socio-emotional well-being domain and one was from the functional well-being domain.

It reflects well on our society that 86% of the participants felt they have never been bullied, and none of the children felt they were bullied often or almost all of the time because of their face and mouth.

When looking at the participants who had the highest COHIP-SF scores and therefore best OHRQoL, none of them reached the maximum achievable score of 76. The highest COHIP-SF scores were four males over the age of 12 years old who self-reported with mild or mild to moderate OI. Reasons for this may be that these young men were unconcerned with their appearance. The literature shows that whilst boys are aware of their dental aesthetics, they are less likely to be affected by their appearance (Rodd et al., 2011). Another factor may be

that none of these participants were confirmed to have DI, which may have had an impact on their OHRQoL.

A 9 year old boy with self-reported mild OI (type I) had the worst OHRQoL with a score of 19. The next lowest scores were 26 for two participants and 32 for one participant. Two were male, and all were older than 12 years old. Again none of these participants had DI. A nine year old child mild OI would be expected to have a higher OHRQoL score. This shows that quality of life is subjective and is not related to severity or type of OI. This child also reported that he thinks about his teeth everyday and has spoken to both his family and his doctor or dentist about the condition of his teeth. He also reported having discoloured teeth 'almost all of the time'. This suggests that whilst he did not have DI, he may have had another dental anomaly causing discolouration which was affecting his OHRQoL such as enamel defects (Arsan et al., 2016, Bontemps et al., 2017). Ideally, interviews or focus groups would have given a more definitive answer. However, the study was carried out on a busy clinic, which gave the participants limited time to answer and we did not wish to overburden families or clinicians.

Data on caries prevalence and other dental anomalies within this cohort may have given further information as to why certain participants scored lower than others. Ideally all patients would have also had dental records and we would have been able to cross reference for caries and dental anomalies to have richer data for this group, but unfortunately the service evaluation showed that the pathway is incomplete.

Another limitation of this study is that we did not ascertain whether participants were undergoing intravenous bisphosphonate therapy. Bisphosphonate therapy, the ability to walk, or needing a wheelchair may also have affected the OHRQoL score. The study by Najirad et al., showed that all persons with types III and IV OI were on bisphosphonate treatment and had lower scores than the children with type I OI. They did not compare the OHRQoL of people with type I OI on bisphosphonate therapy and those not undergoing treatment, this would have been an interesting point to note. Studies have shown that children undergoing bisphosphonate therapy have delayed dental development and eruption times (Malmgren et al., 2020). This may affect their OHRQoL because if they retain primary teeth affected by DI for longer, the tooth wear and loss of vertical dimension of occlusion may affect their OHRQoL.

4.3.2.7 Domains

When looking at individual domains, the oral health well-being domain scores were relatively low. Those participants scoring highest in this domain achieved the highest possible score of 20, and both where males with type I OI, aged 14 and 16 years old respectively. The lowest

score was four and belonged to a 9 year old male with type I OI. Periodontal concerns do not appear to be much of a problem for children with OI; the highest scoring question in this domain was 'Have you ever had bleeding gums?' with the majority responding never.

The functional well-being domain scored particularly high with 25% of the participants reaching the maximum achievable score (16). The gender distribution and average age of the group achieving 16 was similar to the overall cohort, and the majority of these self-reported mild OI. This seems to indicate that type of OI, presence of DI, malocclusion or other dental concerns these participants may have did not appear to affect oral function.

Overall, participants scored well in the socio-emotional well-being domain with four participants scoring a 40/40. From the 57% of participants scoring the median score or higher, there were more males than females, which may imply that females are overall more aware of socio-emotional aspects of their face and mouth such as confidence, anxiety, smiling and feeling unhappy or sad. From the four participants scoring a 40, all self-reported mild OI and there was no bias toward a specific type of OI. This domain also had the question which both scored lowest and was least responded: 'Have you ever felt that you were attractive (good looking) because of your teeth, mouth or face?'. This question was one which the young person's advisory group mentioned may make children uncomfortable due to the wording. Unfortuantely we could not change the question based on their recommendation as it was part of the validated COHIP-SF questionnaire. It may be necessary for the authors of the questionnaire to re-evalute the wording of this question. The lowest score in the socio-emotional well-being domain was a four and was achieved by a 14 year old male with mild OI whose overall COHIP-SF score was one of the lowest scores at 26 out of 76. The next two lowest scores in this domain were the nine year old boy scoring lowest overall, and a 10 year old boy with DI. This implies that persons who scored low in the socio-emotional domain generally had a lower overall score and thus a lower OHRQoL, it is interesting that oral health and function are less likely to affect overall OHRQoL than socio-emotional aspects.

4.3.2.8 Comparison with other populations

When comparing the OI population COHIP-SF scores to other paediatric populations, it is difficult to calculate significant differences due to a difference in distribution between populations. To carry out non-parametric tests, the whole data set for each population is needed and this is not readily available for comparison with our study. When comparing the overall median to means in different populations, there are similarities in COHIP-SF scores between the OI population and those in a general paediatric population, orthodontic population (Broder et al., 2012), and a high caries population (Genderson et al., 2013b). However, the

paediatric population with craniofacial abnormalities (Broder et al., 2012) and a paediatric cleft lip and palate population (Agnew et al., 2017) have lower scores compared to the the OI population. The OI population scored comparatively low in the oral health well-being domain with a similar score to the caries population (Genderson et al., 2013) and the cleft lip and palate population (Agnew et al., 2017). However, children with OI were the highest scoring population in the functional well-being domain and the socio-emotional well-being domain. This may imply that children with OI do not have increased functional limitations due to the potential presence of DI or malocclusions and overall are well-taken care of emotionally. This socio-emotional support may be because a clinical psychologist is part of the OI team at GOSH who helps these children through their social and emotional concerns. Frequently, children with OI do not get the psychological support they require (Dogba et al., 2016) so having a clinical psychologist as part of the OI team is of great importance.

Comparison with the study by the Montreal group (Najirad et al., 2018) was not possible overall for two main reasons. The first is that this study used a different OHRQoL questionnaire, the CPQ. As mentioned previously, this questionnaire has two sets of questions for children aged 8-11 and 12-14, therefore an overall score for the whole population is not achieved. Overall, the older cohort in the Montreal study scored lower in the functional domain. This is different to the cohort in this study. Reasons for this could be that the population of children with more severe types of OI was larger in Najirad et al.'s study or that the questions asked in the CPQ were sufficiently different to obtain a different result to the COHIP-SF scores.

4.3.3 Final Section

4.3.3.1 Additional Questions

When looking at the last section of the questionnaire, the first three were:

1. How much does the condition of your teeth, mouth, or face affect your life overall?
2. How important is the health of your teeth and mouth?
3. How often do you think about your teeth and smile?

The responses to these questions reflected how participants scored on the COHIP-SF section of the questionnaire. Those participants who felt their oral condition affected their life or their oral health or thought about their teeth often, scored lower in the COHIP-SF than those who did not. For the first and third questions, this difference was statistically significant. The results obtained help to validate that the COHIP-SF questionnaire gave an accurate representation of the OHRQoL of the participants.

The two dichotomous questions dealt with whether participants spoke to their family or dentist or doctor about how they felt about their oral status. For both questions, the majority did not communicate with either family or health professionals. It was interesting to note that those participants who spoke to their family about their oral condition scored significantly lower in the COHIP-SF than those who did not [P-value 0.001], whilst those who spoke to their dentist, whilst still scoring lower, did not score significantly so. Those who replied they spoke to both their family and their health professional had a lower median score than those who spoke only to their family or only to their dentist.

Another point of note is that although more participants spoke to their dentist or doctor about their teeth, those children with a lower OHRQoL were more likely to speak to their family than their dentist. This may show that if a child is concerned about their oral health, they would rather express this to their family. This could be because if they feel self-conscious about their appearance, they are more comfortable with their family rather than a stranger. Another possibility is that children respond to their parents' reactions, and if they express their concerns to their parents and their parents catastrophise the situation, this may translate into the child have a worse perception of their own OHRQoL. Parental pain catastrophising leading to an increase in dental, or non-dental pain in children is well documented (de Castro Morias et al., 2018; Coric et al., 2014). A questionnaire on how parents or guardians viewed their child's OHRQoL may have given an indication of this and may be a consideration for similar studies carried out in the future.

When looking at the participants with the lowest and highest COHIP-SF scores, trends can be seen with these final questions. The nine year old boy with a COHIP-SF score of 19, felt that the condition of his teeth, mouth and face affected his life fairly often and his oral health was very important. He said he thought about his teeth everyday and spoke to both his parents and his dentist or doctor about the condition of his teeth. Again, this implies that perhaps he has a dental anomaly, which is not DI, which is affecting the condition of his teeth. Although there are no direct links between OI and other dental anomalies, there have been cases where persons with OI have had amelogenesis imperfecta (Arsan et al., 2016; Bontemps et al., 2017) and further research may need to be carried out to assess prevalence of other dental anomalies in children with OI. The other three lowest scoring participants felt their oral condition affected their life, their oral health was fairly or very important and thought about their teeth at least once or twice a week. All three spoke to their family about their dental condition and only one did not speak to their doctor or dentist about how they felt about their teeth.

Conversely, those children with the highest COHIP-SF scores never thought about their teeth and didn't feel like the condition of their teeth affected their life overall. There were varying degrees of agreement about the importance of their oral health ranging from very to not at all. The majority did not speak to their family or dentist about the condition of their teeth.

4.3.3.2 Qualitative questions

The last two qualitative questions were asked to gauge what the participant's main concerns were and what they would change about their oral status. The first point of note is that those with a lower OHRQoL went on to comment on the factors feeding into it, and those participants who did respond to these questions showed a significantly lower COHIP-SF score than those who did not. Only 20 participants answered both questions and these had a lower median COHIP-SF score than those who answered neither question. It is interesting that those who did not answer either question had the same median score as the overall population whilst those who chose to answer both questions had a significantly lower score. Aesthetics were by far the largest theme that arose from the framework analysis for the first question.

A point of interest is that, whilst not significant, those participants for whom colour was a concern and had a definitive diagnosis of DI, had a higher OHRQoL score than those who had a colour concern and no DI. Reasons for this may include:

- Other possible dental anomalies which have a bigger impact on perceived OHRQoL
- That children with OI having dental anomalies other than DI have been overlooked and left untreated
- That the children with OI and DI are seen more frequently by a dentist and feel better taken care of from a dental perspective.

The second qualitative question asked participants to tell us anything they felt was important with their teeth. Whilst response was poor, it was encouraging that out of those who did respond 39% mentioned oral hygiene and keeping their teeth clean.

When looking at how participants scoring lowest and highest in the COHIP-SF part of the questionnaire responded to the qualitative questions, none of those with the highest scores (73, 72, 72 and 71) had any comments about teeth or smile, barring one who mentioned 'add more enamel' and 'regular check-ups'. Conversely, with those who scored lowest (19, 26, 26, and 32), all except one mentioned changes they would make to their teeth including:

- 'White straight teeth'
- 'If it can be straight and don't have any gaps in my teeth'

- ‘It would make a difference if my teeth would stop hurting. I would be able to eat properly.’

Although data is still limited, there is information on health related quality of life (HRQoL) in children with OI. Studies have found that children with more severe types of OI (Types III and IV), had significantly lower HRQoL scores than those with milder (Type I) OI. They also found that children with OI scored lowest in the physical domains of HRQoL tools (Dahan et al., 2016; Song et al., 2019). Comparing these to our study, we see that whilst children with types III and IV OI did score lower than those with type I OI, the difference was not significant. This is likely due to the small number of participants with either type III or IV OI (n = 29) compared to those with type I (n = 52). Dahan et al. also found that children undergoing bisphosphonate treatment had a better HRQoL than those not having treatment (Dahan et al., 2016) and that socio-economic status was lower in children with OI than in healthy children (Song et al., 2019) which may also have an effect on HRQoL and OHRQoL. Assessing these two factors may have given more information to this study.

During the study, we wanted to see if our results were representative of OI populations outside the United Kingdom (UK), I therefore attended conference in Amsterdam where I presented a poster about this project (Appendix 8). At the conference we networked with a dentist from Vienna who was interested in expanding this study to his local OI service. After email correspondence, it was established that he would use the German version of the COHIP-SF with his population and we began the process of obtaining an amendment to our Ethical approval. My supervisor also contacted the four other OI centres in the UK and we were expanding nationally with them and again looking at ethics approval amendments for all this. Unfortunately, the COVID-19 pandemic caused this work to be put on hold.

Quality of life, and oral health related quality of life are dynamic constructs which are subject to change depending on a child’s moods, and perceptions of the day (Najirad et al., 2018). As the questionnaires were handing out at GOSH before a routine appointment, some anxiety about the appointment ahead or some negative feelings towards needing to be in a hospital environment may have coloured the children’s opinion of their OHRQoL. OHRQoL questionnaires give us an indication of the OHRQoL at the time, and whilst this may be accurate overall, it is tentative.

This study shows us that whilst compared to other medically compromised populations of children, OHRQoL in children with OI is not low, there are still dental concerns which need to be addressed. The COHIP-SF OHRQoL questionnaire, whilst useful, may not have questions

tailored towards the specific needs of children with OI. As Najirad et al. points out, tools developed to assess OHRQoL based on the needs of those with specific conditions will give more accurate perceptions of a child's OHRQoL (Najirad et al., 2018).

5. Conclusions

The COHIP-SF questionnaire results show that Oral Health Related Quality of life (OHRQoL) is highly subjective and whilst children with self-reported severe OI had worse OHRQoL scores than those with mild OI, the difference was not significant. Age and Gender were not indicators of better or worse OHRQoL in children with OI. The presence of dentinogenesis imperfecta was significant in the oral-health well-being domain, and in the functional well-being domain for those children younger than 12 years old. Low scores in the socio-emotional domain was an indicator of worse overall OHRQoL.

From the service evaluation we concluded that families of children with OI who have been seen by paediatric dental specialists found the appointments useful. Some families have had trouble accessing dental care locally due to their general dentists or orthodontists concern over treating children with OI. Further education of general dentists and orthodontics is necessary and may be done by presentation of findings at conferences, development of information leaflets or formal lectures.

Both the service evaluation and the COHIP-SF questionnaire agree that children with OI have dental concerns and these concerns are often aesthetic in nature, particularly involving colour or malocclusion.

6. Future work

Further research is needed on the OHRQoL of Children with rare bone diseases, particularly OI. Extending this study to a national and perhaps international cohort would give a better understanding of the dental and oral issues with this condition. As mentioned in the discussion, plans were already underway to extend this study nationally to the other OI centres in the UK and internationally to Vienna. As the effects of the COVID-19 pandemic ease, and research in this context is once again possible, we will continue to pursue these avenues of expanding the study. We are also interested in making connections with the EDH's sister hospital, the 'Karolinska Institutet' in Sweden and rolling out the study to this population too. This international expansion of the study will provide the OI literature with a large sample of data regarding OHRQoL in children with OI and as such the results will be more representative and more meaningful. Learning from this study, we adapt the demographics section of the questionnaire to include data on the participant's DI status. We would also plan to publish a paper in a journal with the results of this study and present our findings at national and international dental and OI conferences.

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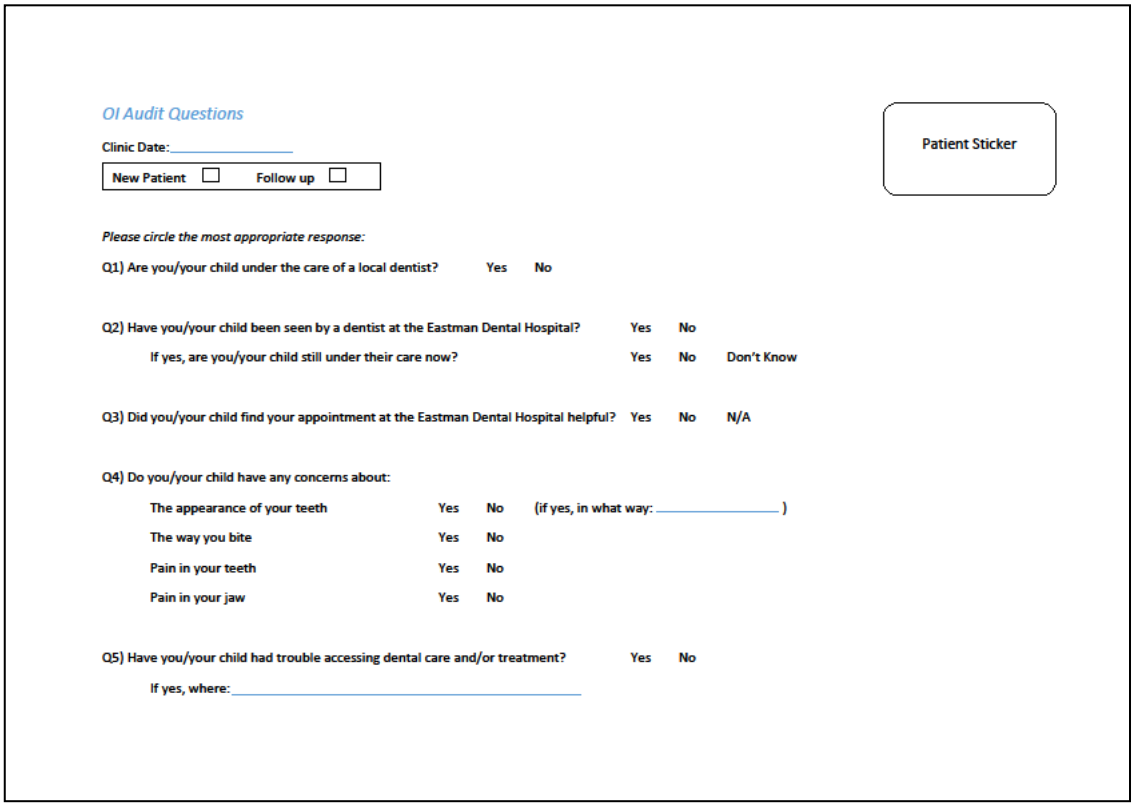
8. Appendices

Appendix 1 – Service Evaluation Questions



OI Audit Questions

Clinic Date:_____

New Patient ☐ Follow up ☐

Patient Sticker

Please circle the most appropriate response:

Q1) Are you/your child under the care of a local dentist? Yes No

Q2) Have you/your child been seen by a dentist at the Eastman Dental Hospital? Yes No

If yes, are you/your child still under their care now? Yes No Don't Know

Q3) Did you/your child find your appointment at the Eastman Dental Hospital helpful? Yes No N/A

Q4) Do you/your child have any concerns about:

The appearance of your teeth Yes No (if yes, in what way: _____)

The way you bite Yes No

Pain in your teeth Yes No

Pain in your jaw Yes No

Q5) Have you/your child had trouble accessing dental care and/or treatment? Yes No

If yes, where:_____

Appendix 2 – Questionnaire

eastman DENTAL INSTITUTE

Great Ormond Street Hospital for Children NHS

UCL

Date:_____

Patient Sticker

Date of birth: _____

Thank you for helping us with our study.
We would like to ask you a few questions about how you feel about your teeth and smile.

Example: During the past 3 months, how often have you felt shy because of your teeth, mouth, or face?

If you have felt shy because of your teeth, mouth, or face then choose the appropriate response. If you felt shy for other reasons choose "Never."

Never	Almost Never	Sometimes	Fairly Often	Almost all of the time
☐	☐	☑	☐	☐

A little bit about you

How do you describe yourself:		Boy ☐	Girl ☐	Other ☐
How old are you?		Age _____		
How would you describe your OI? (please circle)				
Mild	Mild to Moderate	Moderate to Severe	Severe	
Do you know what type of OI you have? (please circle)		Yes	No	
If yes, do you have	Type 1 ☐	Type 3 ☐	Type 4 ☐	Type 5 ☐
	Type 8 ☐	Other (please give details if known)		
		_____		
Ethnicity: _____				

A little bit about your teeth

Please tick ✓ the appropriate box (one box) for each question below.
In the last year have you ever:

		Never	Almost Never	Some-times	Fairly Often	Almost all of the time
1	Had pain in your teeth/Toothache	☐	☐	☐	☐	☐
2	Had crooked teeth or spaces between your teeth	☐	☐	☐	☐	☐
3	Had discoloured teeth or spots on your teeth	☐	☐	☐	☐	☐
4	Had bad breath	☐	☐	☐	☐	☐
5	Had bleeding gums	☐	☐	☐	☐	☐
6	Been unhappy or sad because of your teeth, mouth, or face	☐	☐	☐	☐	☐
7	Missed school for any reason because of your teeth, mouth or face	☐	☐	☐	☐	☐
8	Been confident because of your teeth, mouth or face	☐	☐	☐	☐	☐
9	Had difficulty eating foods you would like to because of your teeth, mouth, or face	☐	☐	☐	☐	☐
10	Felt worried or anxious because of your teeth, mouth, or face	☐	☐	☐	☐	☐
11	Not wanted to speak/read out loud in class	☐	☐	☐	☐	☐
12	Avoided smiling or laughing with other children because of your teeth, mouth, or face	☐	☐	☐	☐	☐
13	Had trouble sleeping because of your teeth, mouth, or face	☐	☐	☐	☐	☐
14	Been teased, bullied or called names by other children because of your teeth, mouth, or face	☐	☐	☐	☐	☐
15	Felt that you were attractive (good looking) because of your teeth, mouth, or face	☐	☐	☐	☐	☐
16	Felt that you look different because of your teeth, mouth, or face	☐	☐	☐	☐	☐
17	Had difficulty saying certain words	☐	☐	☐	☐	☐
18	Had difficulty keeping your teeth clean	☐	☐	☐	☐	☐
19	Been worried about what other people think about your teeth, mouth, or face	☐	☐	☐	☐	☐

The last few questions

		Never	Almost Never	Some-times	Fairly Often	Almost all of the time
1	How much does the condition of your teeth, mouth, or face affect your life overall?	☐	☐	☐	☐	☐
		Not at all	Not much	Fairly important	Quite a lot	Very
2	How important is the health of your teeth and mouth?	☐	☐	☐	☐	☐

3 How often do you think about your teeth and smile?

Every day	At least once or twice a week	At least once or twice a month	Rarely	Never
☐	☐	☐	☐	☐

Have you talked with your family or other important people in your life about how you feel about your teeth and smile?	Yes ☐	No ☐
Have you talked with the dentist or doctor about how you feel about your teeth and smile?	Yes ☐	No ☐

What one change to your teeth or smile would make the biggest difference in your life? And how would things be different for you?

Is there anything else about your teeth, mouth or face that you think is important? Please tell us what it is.

Thank you!

Jasmine May Cachia Mintoff Dr Susan Parekh Dr Megan Riddington

Appendix 3 – Consent form

Patient Identification Number for this study: IRAS: 248033
Form version: 4 Date: 24/09/2018

CONSENT FORM

(For parents / guardians for their child's participation)

Project Title: Oral Health Related Quality of Life in Children and Young People with Osteogenesis Imperfecta

Research Team Jasmine May Cachia Mintoff Dr Susan Parekh
Dr Megan Riddington

1. I confirm that I have read and understood the information sheet (version [1]) for the above study and have had the opportunity to ask questions ☐
2. I confirm that I have had sufficient time to consider whether or not I wish for my child to be included in the study ☐
3. I understand that my child's participation is voluntary, and that they free to withdraw at any time without giving any reason. I also understand that doing so will not affect my child's medical care or legal rights in any way ☐
4. **I understand that relevant sections of my child's medical notes and data collected during the study, may be looked at by individuals from University College London, from regulatory authorities or from the NHS Trust, where it is relevant to my taking part in this research. I give permission for these individuals to have access to my records.** ☐
5. I agree for my child to take part in the above study. ☐

Name of Parent/Guardian	Signature	Date
Name of Person Taking Consent	Signature	Date

Any comments or concerns?
If you have any comments or concerns during the study you may discuss these with the investigator Dr Susan Parekh, on telephone 0203 456 1022 or email s.parekh@ucl.ac.uk

If you wish to go further and complain about any aspect of the way you have been approached or treated during the course of the study, you can write or get in touch with the Complaints Manager at GOSH Hospitals. Please quote the reference number at the top this consent form.
Telephone: 020 7813 8402
E-mail: complaints@gosh.nhs.uk

Consent Form IRAS: 248033 Version 4 (24/09/2018)
Page 1 of 1

Appendix 4 – Assent form

Centre Number:
Patient Identification Number for this study:
IRAS: 248033

Form version: 2.0
Date: 24/09/2018

Assent Form

(For child to agree to participation)

Project Title: Oral Health Related Quality of Life in Children and Young People with Osteogenesis Imperfecta

Research Team Jasmine May Cachia Mintoff
Dr Susan Parekh
Dr Megan Riddington

Please read and then tick the boxes:

1. Have you read the information sheet about this research? ☐
2. Has someone explained this research to you? ☐
3. Do you understand what this research is about? ☐
4. Have you asked all the questions you want to? ☐
5. Have you had all of your questions answered in a way you understand? ☐
6. Do you understand that it's OK to stop taking part at any time? ☐
7. Are you happy to take part in this research? ☐

If you are happy to take part in this study, please write your name and the date below.

Thank you ☺

Name: _____

Date: _____

Appendix 5 – Child information leaflet – 8-12 years old

Oral Health Related Quality of Life of Children with Osteogenesis Imperfecta

Participant Information Leaflet

UCL eastman DENTAL INSTITUTE

We are carrying out a project and need your help. We would like to talk to you about your teeth and mouth. Please read this leaflet. You can also discuss it with your mum or dad.

Why are we doing this project?

We would like to know how you feel about your teeth and mouth, and to know if having Osteogenesis Imperfecta (OI) affects your teeth.

Do I have to take part?

No. It is completely up to you, and you don't have to take part. If you change your mind while answering the questions you can stop at any time.

What will I have to do if I take part?

If you decide to help us we need you to answer some questions. This should only take around 10-15 minutes. There are no right or wrong answers. We are just interested in learning how you feel.

Why is it good for me to take part?

The information you give us will help us to learn how children with OI feel about their teeth and mouth.

What Will happen to my work?

We hope to publish the information to help other professionals who take care of children with OI. We will make sure that all information about you is kept a secret. No one will know who you are or that you have taken part.

Did anyone else check that it is ok to do the study?

Yes. A special group of people called the 'Research Ethics Committee' has checked the project to make sure it is ok to carry out.

If you do not understand something or have any questions to ask us, you can always email us at s.parekh@ucl.ac.uk

Thank you

IRAS: 248033
Version 3 (25/09/2018)

Appendix 6 – Child information leaflet – 13-16 years old

Oral Health Related Quality of Life of Children with Osteogenesis Imperfecta

Participant Information Leaflet 13-16 years old

UCL eastman DENTAL INSTITUTE

We are carrying out a research project and we would like to talk to you about your teeth and mouth. Please read this leaflet. You can also discuss it with your parents.

Why are we doing this project?

We would like to know how you feel about your teeth and mouth, and to know whether having Osteogenesis Imperfecta (OI) affects your teeth.

Do I have to take part?

No. Taking part is optional and is up to you and your parents. If you decide to take part, remember you are free to withdraw from the project at any time.

What will I have to do if I take part?

If you decide to take part you will be asked to answer a short questionnaire. This should only take around 10-15 minutes. There are no right or wrong answers. We are just interested in learning how you feel.

If you do not understand something or have any questions to ask us, you can always email us at s.parekh@ucl.ac.uk

Why is it good for me to take part?

The information you give us will help us to learn how children with OI feel about their teeth and mouths.

What will happen to my work?

We hope to publish the information in a scientific journal, and the Brittle Bone Society website. We will also be making leaflets for children with OI. We will make sure that all information about you is confidential.

Did anyone else check that it is ok to do the study?

Yes. A group of people called the 'Research Ethics Committee' has checked the project to make sure it is ok to carry out.

Appendix 7 – Parent information leaflet

Additional Details:

University College London is the sponsor for this study based in the United Kingdom. We will be using information from your child and/or your child's medical records in order to undertake this study and will act as the data controller for this study. This means that we are responsible for looking after your information and using it properly.

Your rights to access, change or move your information are limited, as we need to manage your information in specific ways in order for the research to be reliable and accurate. If you withdraw from the study, we will keep the information about you that we have already obtained. To safeguard your rights, we will use the minimum personally-identifiable information possible.

You can find out more about how we use your information by contacting Dr. Susan Parekh.

Great Ormond Street Hospital will collect information from your child and/or your child's medical records for this research study in accordance with our instructions.

Great Ormond Street Hospital will keep your name, and contact details confidential and will not pass this information to University College London. Great Ormond Street Hospital will use this information as needed, to contact you about the research study, and make sure that relevant information about the study is recorded for your care, and to oversee the quality of the study. Certain individuals from University College London and regulatory organisations may look at your medical and research records to check the accuracy of the research study. University College London will only receive information without any identifying information. The people who analyse the information will not be able to identify you and will not be able to find out your name, or contact details.

UCL HOSPITALS eastman DENTAL INSTITUTE

If you require a large print, audio or translated copy of this document, please contact us on (020)34561023. We will try our best to meet your needs.

If you wish to discuss this study with a member of the research team or an independent expert who is not part of the research team, please ask Dr Susan Parekh

Thank you for taking the time to read this leaflet.

Chief Investigator's Contact Details:

Dr. Susan Parekh
Tel: 020 3456 1022
s.parekh@ucl.ac.uk

Unit of Paediatric Dentistry
The Eastman Dental Hospital
256 Gray's Inn Road
London, WC1X 8LD

Study Investigators:

Dr. Susan Parekh
Dr. Megan Riddington
Jasmine May Cachia Mintoff

UCL

ORAL HEALTH RELATED QUALITY OF LIFE OF CHILDREN WITH OSTEOGENESIS IMPERFECTA

Parent Information Leaflet

Invitation

You and your child are being invited to participate in this research study. This research study makes up part of a post-graduate thesis investigating the oral health related quality of life of children with osteogenesis imperfecta. Before you make a decision, it is important that you know why the research is being done and what you will be asked to do. Please take time to read the following information carefully and discuss it with others if you wish. Ask us if there is anything that is not clear or if you would like more information.

Purpose of Study

Osteogenesis Imperfecta (OI) is also known as brittle bone disease. Children with OI are prone to fractures of their bones, and it can also affects their teeth and jaws. Children with OI may have specific concerns about their teeth, but no research has been done on their oral quality of life. With this study we hope to gain an insight into how OI may affect oral health, so that we can improve the dental care we provide.

Why has my Child Been asked to participate in this study?

Your child has been asked to participate in this study since your child lives with osteogenesis imperfecta and meets the inclusion criteria for our study.

Does my Child Have to Take Part?

No. If your child decides to take part, you will be asked to sign a consent form and your child will be asked to sign an Assent Form. The decision to take part is optional and it is entirely up to you and your child.

What if my Child no Longer wants to Take Part?

You are free to withdraw at any time without giving a reason and the standard of care your child receives will not be affected in any way.

What will my Child Need to do if he/she Takes Part?

Your child will be asked to answer a short questionnaire. This should only take around 25-30 minutes depending on how much your child wishes to say. There are no right or wrong answers. We are just interested in hearing your child's experience. You will not need to do anything else. If you have not decided whether to take part by the end of your appointment, you can choose to answer the questionnaire at your next appointment.

What are the Disadvantages or Risks?

There are no risks anticipated. None of your child's answers will affect your child's treatment in any way.

What are the Possible Benefits?

We cannot promise the study will help you directly, but the information we gather from this study will help us to better understand how children with OI view their teeth and oral health. This will help us to give them the best treatment tailored to their specific needs

What will Happen With the Results?

We hope to publish the results of this study. We also hope to liaise with the Brittle Bone Society to give them our results. All confidential information will be coded and your child will not be identifiable in any way.

Will Taking Part in the Study Remain Confidential?

Yes. No one other than the person (your child's Osteogenesis Imperfecta team) and the study investigators will be aware of your participation in the study.

Will the Information Obtained in the Study be Confidential?

Yes. All information collected from you and your child during the research will remain strictly confidential and will be seen only by the investigators of this study (as named on this sheet). The safety and security of the data will be the responsibility of the chief investigator (Dr. Susan Parekh).

Who has reviewed the Study?

The North of Scotland (2) Research Ethics Committee has reviewed the study. If you would like a copy of the results.

What If I have any Concerns?

If you have a concern about any aspect of this study or the conduct of members of the research team, in the first instance you should speak to Dr. Susan Parekh or members of the research team who will do their best to answer your questions.

If you remain unhappy and wish to complain formally, you can do this through the normal hospital complaints procedure by contacting the NHNN Patient Advice and Liaison Service (PALS@ucln.nhs.uk). Their office is based on the ground floor, at the NHNN near the pharmacy and library. Opening hours are 0900-1600, Monday-Friday (except public holidays). Telephone number: 020 3447 3042.

Who is funding and organising this study?

This study is organised and led by Dr. Susan Parekh, the Chief Investigator and is funded by University College London. Sponsor for the study is University College London.

Appendix 8 – Poster from Amsterdam Conference

P96 -

ORAL HEALTH RELATED QUALITY OF LIFE IN CHILDREN WITH OSTEOGENESIS IMPERFECTA

Cachia Mintoff[i], J., Parekh[i], S., Riddington[ii], M., DeVile[ii], C.

Great Ormond Street Hospital for Children NHS Foundation Trust

UCL

BACKGROUND

Children with Osteogenesis Imperfecta (OI) can have significant dental concerns, such as Dentinogenesis Imperfecta (DI) and malocclusion.

Oral health related quality of life (OHRQoL) is an important part of clinical dentistry, associating oral health with function, psychology, social life and experience of pain.

OBJECTIVES

- To investigate oral health-related quality of life (OHRQoL) in children aged 8-16 years with OI

METHOD

- A validated questionnaire, The Child Oral Health Impact Profile–Short Form (COHIP-SF) was used to measure OHRQoL. A higher COHIP-SF score represents higher OHRQoL (max score = 76)
- Ethical approval was obtained
- Children meeting the inclusion criteria completed the questionnaire after informed consent
- Z-test statistics carried out using MS excel

KEY POINTS

1. **Children reporting severe OI had lower OHRQoL scores**
2. **Males reported slightly better OHRQoL than females**
3. **This study confirms children with OI have oral concerns**

RESULTS

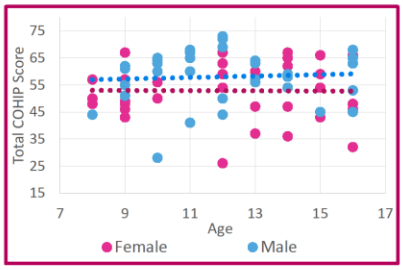
- 83 participants (46 males, 37 females)
- Mean age 12.01 years.
- 39 (47%) reported mild OI, 25 (30%) 'mild to moderate', eight (10%) 'moderate to severe' and seven (8%) 'severe' and 4 (5%) did not respond

COHIP-SF

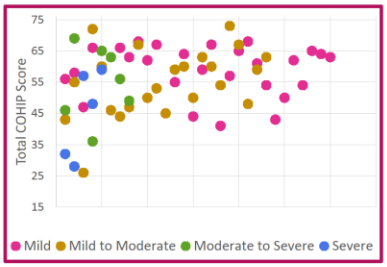
- The overall mean for the COHIP-SF was 55.7/76
- Individual domain-mean OHRQoL was:
  - Oral Health Well-Being: 12.8/20
  - Functional Well-Being: 12.9/16
  - Socio-Emotional Well-Being: 30.3/40

CONCLUSIONS

- Children with OI reported similar OHRQoL scores to general paediatric populations[1] and a high caries population[2]
- Children with OI reported lower scores in the Oral Health Domain than the Functional well-being and socio-emotional domains.
- On average, the more severe a child's perception of their OI, the lower the OHRQoL.
- This agrees with another study assessing OHRQoL in children aged 11-14 years old with OI[3]



Graph 1: COHIP Scores for males and females with trend lines showing a slightly higher COHIP (better OHRQoL) in males



Graph 2: COHIP Scores vs Patient perceived severity of OI

SCAN ME

For more info scan the QR code

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FOR FURTHER INFORMATION, PLEASE CONTACT:

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ii. Great Ormond Street Hospital: megan.riddington@gosh.nhs.uk

eastman DENTAL INSTITUTE

Adherence to Acceptance Criteria for Emergency Patients Audit

A Clinical Audit

Submitted in partial fulfilment of the requirements for the Degree of

Doctor of Dentistry (Paediatric Dentistry)

UCL

Submitted by

Jasmine May Cachia Mintoff

Word count: 7495

UCL Eastman Dental Institute,

256 Gray's Inn Road,

London WC1X8LD,

Acknowledgment

I would like to thank Ms Suzanne Dunkley, and all the team at the Eastman Dental Hospital for their help with this audit. I would particularly like to thank Samad Ali and the administration team for helping me develop the standard operating procedure flowcharts.

Abstract

Background:

Departmental Emergency Acceptance Criteria (EAC) are Swelling and Dental Trauma**.** No emergency slots exist, and increasing numbers of patients booked as add–ons, and not always seen within the recommended time-frame

Aims:

Establish if emergency patients:

- adhere to EAC
- Are seen within two days

Standard:

- **100%** of emergency patients meet EAC
- **100%** of patients seen within two days

Method:

Retrospective audit.

- Cycle 1(C1) – paper notes (01/07/2018 – 31/09/2018)
- Cycle 2 (C2) – electronic records (01/10/2019 – 31/12/2019)

Results:

- 64 patients C1 & 49 patients C2
- Age range 0.3-18 years
- Patients meeting EAC improved - 59% (C1) to 86% (C2)
- C1 reasons for not meeting EAC: oncology referrals, lost anterior and posterior restorations/prosthesis, neonatal teeth, pain and caries. In C2 this reduced to denture problems and lost gold chain.
- Number of Patients with same day appointments worsened: 51% in C1 vs 42% in C2; as well as within two days 82% in C1 vs. 69% in C2.

Recommendations and action plan:

After C1:

- Adapted EAC for new patients; introduced different EAC for current patients
- Developed standard operating procedure (SOP) flowcharts based on SDCEP guidelines
- Updated triaging form
- Departmental teaching on triaging patients using new SOP

After C2:

- Face-to-face teaching for administrative team using SOP
- 'Emergency slots' on new patient clinics

Conclusions:

- Number of emergency patients booked who met EAC improved
- All emergency new patients met EAC in C2.
- Time standard not met reflecting changes to clinic scheduling over the audit time period, therefore emergency slots created.
- Re-audit in one year

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1 Introduction and Background

1.1 Emergency dental care

Emergency dental care is a mainstay of any dental service. Urgent dental care is the responsibility of NHS England (Marshman, 2019). At present, there are no national standards agreed upon for emergency dental care and different practices may interpret emergency treatment differently depending on availability of practitioners, remuneration considerations and the time at which the emergency occurs (SDCEP, 2017).

The National Health Service (NHS) England defines emergency dental care as care for patients who require dental treatment immediately to prevent risk of health complications (NHS England, 2015). Dental treatment which can cause risk to life include submandibular cellulitis and may need to be treated in a hospital (Worsley et al., 2017). Urgent dental care is for those patients who need to be seen quickly but the condition is not threatening to general health. Dental emergencies include excessive bleeding after an extraction, swelling of the facial tissues due to dental infection or trauma to the teeth or jaws. Urgent dental care is care for severe facial pain which cannot be controlled by regular analgesia and acute dental or soft tissue infection (NHS England, 2015).

The Scottish Dental Clinical Effectiveness Programme (SDCEP), also developed guidance on emergency or urgent dental treatment and they propose timescales in which these patients should be seen. Emergencies should be seen within 60 minutes for assessment, urgent cases should be seen within 24 hours and non-urgent cases should be seen within 7 days (SDCEP, 2017).

According to the NHS England, emergency dental care for paediatric patients can be provided by any dentist with a training level equivalent to Dental Foundation Training. Severe trauma to dental tissues or the alveolar arches may require treatment by a specialist paediatric dentist or paediatric dental consultant and should be referred to these clinicians as soon as possible. Possible injuries include avulsions, moderate to severe luxation injuries or trauma to several teeth (NHS England, 2018).

Many dental emergencies will be due to caries or trauma. According to the 2013 Child Dental Health Survey, 13% and 14% of five- and fifteen-year olds respectively have severe dental caries. Similarly, 12% of twelve-year olds and 10% of fifteen-year olds have suffered from some type of dental trauma to their permanent incisors (NHS England, 2015).

There is limited data about the number of dental emergencies for paediatric patients. However according to Worsley et al., in 2017, between 2014 and 2015, 3.7 million people in England received urgent dental care by NHS dentists (Worsley et al., 2017). A study carried out in the West midlands in 2019 found that 7.7% of all dental treatments courses delivered by NHS dentist was for emergency dental care (Woodman et al., 2019). Another review found that younger adults are more likely to use emergency services and the highest use is in those between 19 and 29 years old (Marshman, 2019).

1.2 Impact of Dental Emergencies

Dental emergencies most frequently occur when a patient is in pain or has had trauma. These dental concerns can have an impact on the patient, the patient's family as well as the service providing emergency care.

1.2.1 Impact on the Patient

Untreated caries, infection or trauma can lead to reduced oral health well-being, as well as a reduction in function, and socio-emotional well-being. These all lead to a reduced oral health related quality of life (Martens et al, 2018).

Appropriate emergency care can alleviate symptoms and reduce the long-term implications of poor oral health on both the patient and the society in which they live. Societal impacts can include missed school or work (Worsley et al., 2017).

Caries can impact a child both in the long and short term as children with caries in the primary dentition are more likely to suffer caries in the permanent dentition. Caries also has an effect on growth as it can negatively affect weight and height in a growing child (Ayhan et al 1997; Abanto et al., 2004). Studies show that caries affects quality of life and can affect:

- Eating, both amount of food consumed and the types of food (Low et al, 2000)
- Sleeping, disturbed sleep, waking up at night or problems falling asleep (Low et al, 2000)
- Schooling, affecting cooperation, concentration and interaction with other children (Low et al, 2000)
- Cognitive development (Abanto et al., 2004)

Studies also show that traumatised teeth impact oral health related quality of life with children less willing to smile, laugh and clean their teeth. Like caries, it also impacts their eating habits. Dental trauma can also have a long term impact on aesthetics, occlusion and function. Thus timely, and appropriate treatment is vital in cases of dental trauma (El-Kalla et al, 2017).

1.2.2 Impact on Families

When a child is in distress due to dental pain, the impact on a parent can also be significant. Parents most frequently experience distress and guilt over the child's dental health (Corrêa-Faria et al., 2018; Abed et al., 2019). If a child's sleep is disturbed, parents sleep is also affected, restricting normal life behaviours (Abed et al., 2019; Abanto et al., 2012). They are also required to take time off work to care for their child and take them to the dentist which can additionally have a financial impact on the families (Corrêa-Faria et al., 2018). Significantly, it also worries parents that the dental trauma may offer their child few opportunities in life (Abanto et al., 2012).

1.2.3 Impact on Dental Services

According to NHS England in 2018, evaluation of the impact of emergency dental care should be carried out as part of emergency dental services planning. Including impacts on A&E departments and dental access centres (NHS England, 2018). However, in spite of this there is little research available on the impact of emergency dental care on dental services, whether in practice or at a secondary level.

There is however data on the impact of dental emergencies on emergency services. A study by McCormick et al in 2013 found that in a hospital in Virginia, between 2007 and 2009, 4.3% of emergency department visits were due to dental concerns. This study also found that emergency dental patients contributed to overcrowding and increased waiting times for other emergency patients (McCormick, 2013). This was also found by a study in Australia in 2014 in which almost 1% of emergency patients seen were for dental reasons (Verma et al., 2014).

This emphasises the importance of a clearly publicised pathway for dental emergencies to ensure appropriate signposting to emergency dental services.

1.3 General Dental Emergency Pathways

1.3.1 Triaging patients with dental concerns

Typically, when patients feel they have a dental emergency, the initial contact is by telephone (SDCEP, 2017). NHS Direct, the predecessor to NHS 111, assessed 12.5 million telephone calls in 2010/2011, and 8% of these were for dental problems. NHS 111 now directs dental problems to a dental nurse for dental triaging (Marshman, 2019). Patients are triaged depending on their need for treatment and assigned into one of three groups: those requiring emergency dental care, those requiring urgent dental care and those requiring routine care (SDCEP, 2017).

According to the Cambridge English Dictionary, triage is defined as 'the process of examining problems in order to decide which ones are the most serious and must be dealt with first'. In the case of dental emergencies, this is to prioritise patients needing emergency dental care and ensure they are directed to the correct dental or emergency service. Dental triaging is particularly important for those patients who do not attend the dentist regularly.

1.3.2 Accessing Emergency Dental Care

In a review of urgent dental care in the UK from 2019, data collection showed that patients 'shop around' for dentists when they need to seek emergency treatment. Reasons include dentists or doctors unable to see emergency patients in a timely manner, as well as the cost associated with urgent dental care. Many patients choose to attend A&E or general medical practitioners which are free or charge in the hopes of obtained a prescription to help alleviate symptoms (Marshman, 2019), however, this is not relevant for paediatric patients as their care is free. Other barriers to care may include availability of paediatric dentists and long waiting lists for NHS treatment.

According to the NHS website, emergency dental treatment should be accessed by contacting one's own general dentist or using the NHS 111 telephone service which can provide self-help advice and refer to an available NHS emergency dentist. It also provides details on cost of emergency treatment and when it is necessary to attend hospital for emergency treatment (NHS England, 2020).

Local dentists may be able to provide emergency appointments and should be a patient's first point of call. If the dental emergency is out of hours, a recorded voicemail message should indicate where patients can access out-of-hours emergency care (Marshman, 2019). If the

general dentist is unable to provide emergency dental treatment, NHS 111 can connect patients to an emergency dentist for any necessary treatment (NHS England, 2017).

For treatment at a secondary dental service, patients should be referred to the dental hospital by their general dentist, general medical practitioner or the community dental service for specialist treatment. Different hospitals have different criteria for emergency treatment. Some examples include:

- Guy's and St Thomas' Dental hospital - Walk-in A&E service for dental trauma and acute dental infections (Guy's and St Thomas' NHS Foundation Trust, 2020)
- King's College Hospital – Appointment based self-referral emergency service for swelling, infection and dental trauma (telephone to make appointment)
- University College London Hospital (UCLH) – Walk-in casualty service for swelling or dental trauma (Eastman Dental Hospital) (UCLH, 2020)

1.4 Eastman Dental Hospital Paediatrics Department Emergency Pathway

Prior to this audit, no standard operating procedure or official policy was in place for emergency patients at the EDH paeds department. All emergency or walk-in patients were booked under the PFA02 clinic code which is not assigned to a specific clinician.

Acceptance Criteria for emergency patients are children aged 16 year or younger who present with

- Facial Swelling
- Dental Trauma

Patients booked under these codes have either been referred as urgent by their General Dental Practitioner, an emergency dentist or the Community Dental Service, or they have accessed the UCLH website and found the criteria for walk-in casualty patients. Local agreement in the department is that emergency patients should ideally be seen within 48 hours of contacting the department.

Emergency patients went down one of two pathways:

1. Walk-in patients:
    - Inform reception of emergency and give reception team referral letter if available

- They are asked to wait in waiting area whilst receptionist/ dental nurse speaks to consultant in charge on clinic
- Consultant in charge will triage/ speak to patient/ family and decide if emergency treatment is required
- If emergency treatment is required, an appointment is booked under PFA02 code or onto a specific clinic code if an appointment is available, and patient is seen immediately.

2. Telephone emergency patients:
    - Patients telephone and are in contact with administration team. They are asked for details of emergency and whether a referral form is available
    - Administration team speaks to consultant in charge on clinic
    - Consultant in charge will triage/ speak to patient/ family and decide if emergency treatment is required
    - If emergency treatment is required, an appointment is booked onto PFA02 code or onto a specific clinic code if an appointment is available and patient is seen within 48 hours where possible

Over the summer of 2018, many emergency patients were seen. As these patients are booked under the PFA02 code and did not have assigned times, the emergency patients were absorbed into existing clinics.

1.4.1 Impact of emergency patients at the EDH

As the emergency patients were being treated by clinicians who already had full lists of booked patients, several issues were seen to arise within the department. The impact of several emergencies a day led to:

- Overbookings and double bookings
- Busier clinics which impacted treatment options due to time management
- Overworked clinicians who sometimes worked into their lunchbreaks and late to ensure patients were seen

1.5 Auditing Emergency Services

According to SDCEP guidelines, emergency dental services should be audited regularly. Audit should ensure:

- Arrangements for emergency dental treatment are examined and reflect good practice
- Appropriateness of the triaging system (SDCEP, 2017)

Further information on why this audit was carried out can be found in section 2.1.

2 Aims and Standards

The aims of this audit were twofold. The primary aim was to establish whether emergency patients seen in the dental department at the Eastman Dental Hospital (EDH) adhered to the department's emergency acceptance criteria (EAC). The secondary aim was to assess if these patients were seen within two days. According to SDCEP guidelines, dental clinics that provide an emergency service should regularly audit their service as well as the appropriateness of the triaging of patient attempting to access emergency dental care (SDCEP, 2017). Therefore, the standards of this audit were set at, 100% of emergency patients are meeting the department's EAC and 100% of emergency patients are seen within two days.

2.1 Why this audit?

An increasing number of emergency patients were attending the paediatric dental department of the EDH over the summer of 2018. As the department had no dedicated emergency appointments, all emergency patients were being absorbed into existing clinics. This led to overbooked clinics and overworked dentists, busy clinics risk impacting the quality of care provided. Concern was also expressed that some emergency patients were not being seen in a timely manner. It was therefore important to audit this service to ensure only patients that met the EAC were seen in a timely manner, and therefore ensure there is no compromise to quality of care provided.

2.2 Previous Audits on the emergency service at the Eastman Dental Hospital paediatrics department

A search through the audit database of the EDH paediatrics department from 2016 revealed that this service has not been audited recently. This highlights the importance of carrying out this audit.

3 Materials and Methods

This audit was registered and approved by the audit committee of the EDH. Retrospective analysis of emergency patients seen within a three-month period, for each cycle, was carried out.

For the first cycle, carried out between July and September 2018, paper notes were used for data collection. Patients chosen were those booked under the PFA02 clinic code on the electronic scheduling system, CareCast. Notes were retrieved from the medical record office and stored in a locked room during data collection to ensure maintenance of confidentiality. During this time period, 64 patients were booked however only 49 of these notes were available for retrieval from medical records. The rest were unavailable as they were with other clinicians in different departments, or the secretaries for writing letters.

The second cycle was carried out between October and December 2019, just over a year later. Due to the switch to electronic health records using EPIC, as well as moving into a new building, the audit could not be carried out exactly 1 year later. For this cycle, the patients chosen were once again those booked under the PFA02 code on EPIC; as well as those booked under specific clinician codes where 'Emergency' was chosen as reason for attendance. In this case, all notes were electronic, thus all 49 emergency patients seen within this time frame were used for data collection.

Data collection was done using a proforma created for this purpose and all data was then inputted into MS excel for data analysis.

3.1 Data Collection Proforma

The proforma was made up of four main sections. Section one dealt with patient demographics including name, hospital number, date of birth, age and gender. Section two detailed the emergency, and included type of emergency, when they were seen, and how long between initial contact and patient being seen.

Section three included data about the clinician who dealt with the emergency, medical needs of the patients and the source of the emergency referral.

The final section dealt with interventions carried out at the emergency appointment as well as the outcome of the emergency appointment.

The complete Proforma can be found in Appendix 1.

4 Results Cycle 1

All data was inputted into MS excel for data analysis. Forty-nine patients from cycle 1 were analysed, the remaining 15 paper notes were unavailable. These notes were unavailable because they were either at other clinicians in different departments or they were in the care of the secretaries for patient letters to be typed out and sent to the patient's parents.

4.1 Demographic Data

Gender distribution was approximately equal, with 49% males and 51% females. The age ranged from six months to 19-years-old with an average age of nine-years-old. When dividing into age groups, the majority of dental emergencies were patients between the ages of four and seven years old with 15 patients (31%) falling within this category. Table 4-1 shows how emergency appointments differed between the different age groups.

Table 4-1 – Table showing distribution of dental emergency appointments by age groups.

Age groups	**numbers**	**percentage**
<=3	8	16
4-7	15	31
8-12	13	27
13-16	11	22
>16	2	4

4.2 Type of Emergencies

As mentioned earlier, EAC for the paediatric department of the EDH are swelling and dental trauma. Over half, 59%, of patients seen (n=29) followed the department EAC, 10% (n=5) had swellings and 49% (n=24) suffered from dental trauma. The remaining 41% (n=20) of emergency patients did not fit the EAC.

From the 24 trauma patients, 42% (n=10) suffered trauma to the primary dentition, 54% (n=13) suffered trauma to the permanent dentition and 4% (n=1) suffered trauma to both primary and permanent teeth.

Those patients who did not fit the EAC showed a variety of reasons for attendance as seen in table 4-2.

Table 4-2 – Table showing reasons for attendance not adhering to EAC

Reasons for Attendance Not Adhering to EAC	**Numbers**	**Percentage (of Total)**
Oncology referral before starting oncology treatment	4	8%
Lost/ Fractured restoration/ RBB	6	12%
Pain	7	14%
Caries	1	2%
Sinus	1	2%
Neonatal teeth + ulcer	1	2%

From the emergency patients seen, 59% (n=29) were new patients, 69% (n=20) of the new patients followed the EAC, whilst 31% (n=9) did not. From the new patients not adhering to EAC, reasons for non-adherence included caries, lost restoration, neonatal teeth, pain, sinus and oncology referrals. All 8% (n=4) of the oncology referrals were new patients. From the patients who attended as an emergency but were already EDH patients, 45% (n=9) followed the EAC, whilst the remaining 55% (n=11) did not. These included five lost restorations and six patients who were in pain. Figure 4-1 shows a graphical representation of the male to female ratio of patients fitting EAC.

Considering age, emergency appointments due to trauma were most prevalent in the 8 – 12-year-old age group at 29% (n=7). Followed by those 3-year olds or younger and the 4 – 7-year-old age groups both at 25% (n=6). Children in the 13 – 16-year-old age group had the least traumas (21%, n=5).

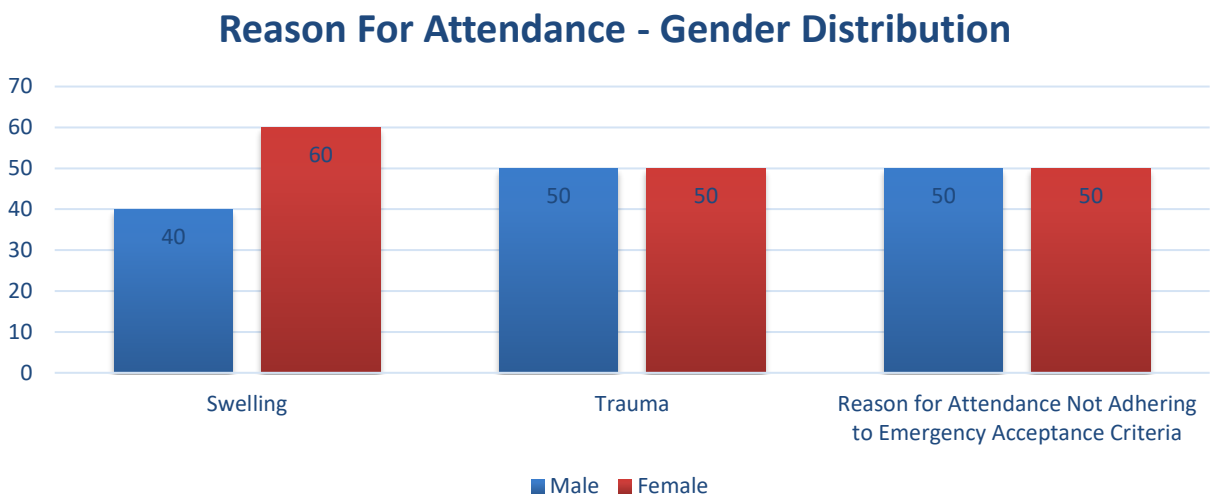

Figure 4-1 - Graph showing gender distribution depending on reason for attendance

4.3 Time taken to see Patient

The local agreement at the paediatric department is that emergency patients are seen within two days from first contact. The majority of patients, 82% (n=40), were seen within this time frame. Figure 4-2 shows the date difference from patient contact to patient being seen for all patients.

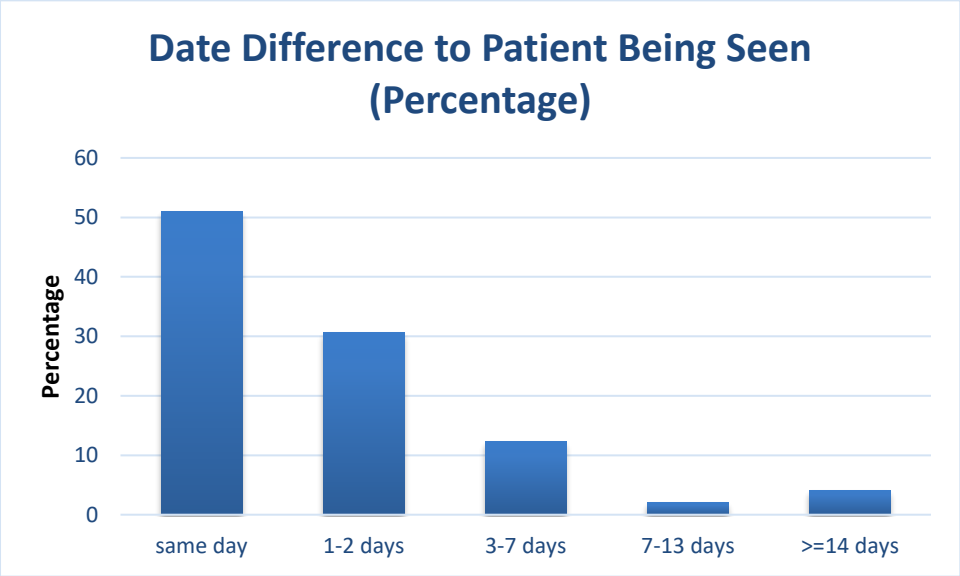

Figure 4-2 – Graph showing Date Difference from first contact to patient seen for emergency appointment

Diagnoses of patients seen in a longer timeframe were:

- 3-7 days: 2 Pain, 1 Caries and 3 Trauma
- 7-13 days: Trauma

- After two weeks: 1 Swelling and 1 Trauma (permanent tooth)

4.4 Grade of Clinician Seeing Patient

In the department, grades of clinicians include: Speciality Dentists, Dental Core Trainees (DCTs), Speciality Registrars (StRs), Postgraduate students, and Consultants.

Half (50%, n=24) of emergency patients were seen by the postgraduate students, consultants saw 23% (n=11). Distribution of patients according to grade of clinician can be seen in Figure 4-3.

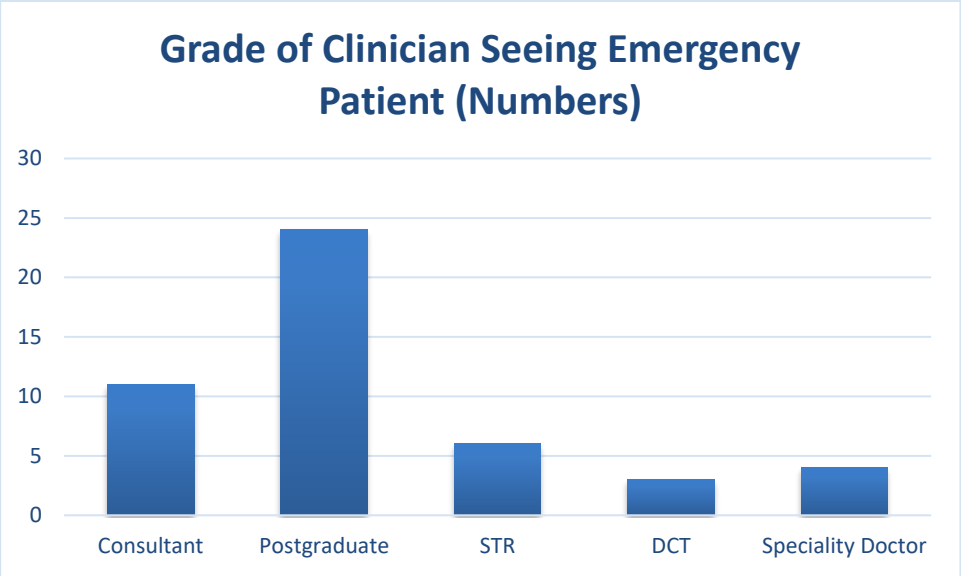

Figure 4-3 – Figure Showing Distribution of emergency patients by clinician

4.5 Medical History

Seventy-three percent (n=36) of emergency patients were fit and healthy. The remaining 27% (n=13) had a variety of medical conditions which can be seen in Table 4-3.

Table 4-3 – Table showing distribution of medical conditions seen

Medical Conditions	Numbers	Percentage from Total	Percentage from Medically Compromised
Oncology	5	10%	38%
Asthma	4	8%	31%
Autism	1	2%	8%
Cardiac	2	4%	15%
Epilepsy	1	2%	8%

Four of the Oncology patients were referred by the University College London Hospital (UCLH) oncology team for assessments before starting oncology treatment. From these four patients, one was discharged and three were booked in for follow-up appointments. The other oncology patient was an existing patient who attended due to pain.

4.6 Referral for Emergency Appointment

The majority of emergency patients (n=21, 43%) were general dental practitioner (GDP) referrals. From these 21 patients, 15 patients (71%) were new patients, whilst 6 patients (29%) were already registered with EDH. Self-referral was the next most common at 29% (n=14), these were all existing EDH patients. 8% (n=4) were new patients referred by the community dental service (CDS) and the remaining 20% (n=10) were referred by ‘other’. The distribution from the ‘other’ group can be seen in table 4-4.

Table 4-4 – Table showing where patients in 'other' category were referred from

Referred From	Number
Hospital (non-specific)	1
Unknown	2
UCLH MaxFax	1
UCLH Oncology	4
A&E	2

4.7 Intervention at Emergency Appointment

Interventions carried out were divided into eight groups:

- Antibiotics
- Suturing
- Surgicel Placement
- Endodontic Treatment
- Temporary restoration for caries
- Trauma Management
- No Treatment
- Other

Division of interventions by group can be seen in Figure 4-4.

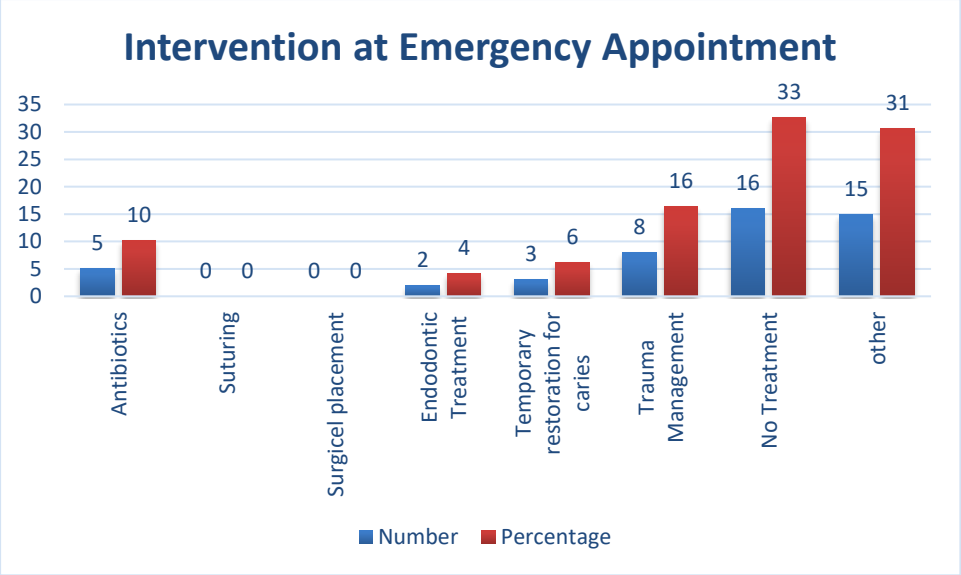

Figure 4-4 – Graph showing Interventions at emergency appointment

Most commonly, patients received no treatment (33%, n=16). From these, 2 patients had swellings and were placed on an emergency GA list, 11 patients suffered from dental trauma and trauma assessments were carried out, but no active treatment was given. The final 3 were patients that did not meet the EAC.

'Other' Interventions ranged from extractions (n=6, 40%) to incision and drainage (n=1, 7%). The variation of treatments can be seen in Figure 4-5.

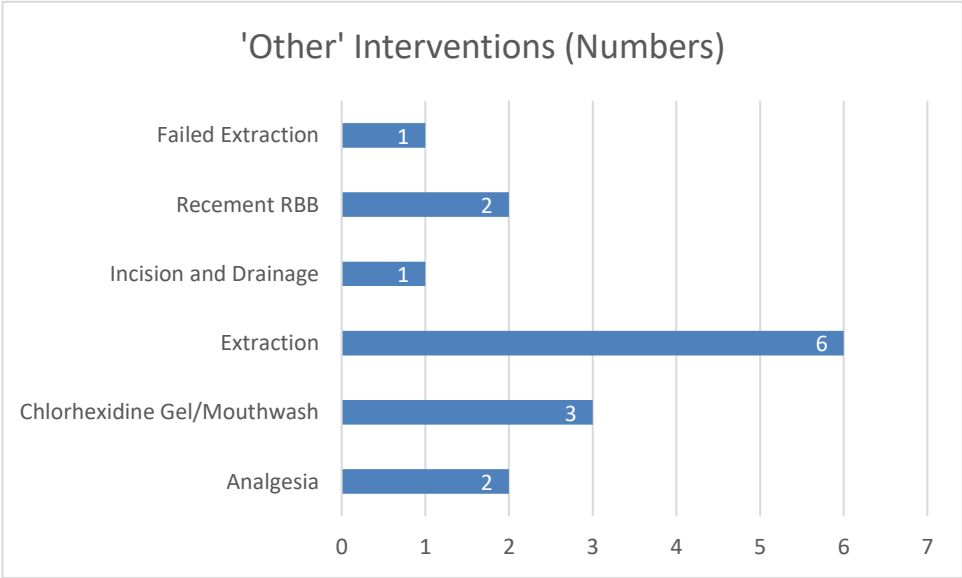

Figure 4-5 – Graph showing Variation in 'Other' Interventions

Those patients who had swelling (n=5, 10%), had a variety of interventions. Two were prescribed antibiotics, two had no treatment but were placed on a general anaesthetic waiting list, and one had the swelling incised and drained.

Figure 4-4 also shows that only 16% of patients (n=8) had trauma management. As mentioned previously, 49% (n=24) patients attended with trauma as a reason for the emergency appointment. The interventions for trauma patients can be seen in Table 4-5.

Table 4-5 – Table showing interventions for trauma at emergency appointment

Management	**Primary Teeth**	**Permanent Teeth**	**Mixed Primary and Permanent Teeth**
Other	2 (Extractions)	1 (Chlorhexidine gel/ mouthwash)	0
Antibiotics	2	0	0
No Treatment	6	5	0
Trauma Management	0	6	1
Endodontic treatment	0	1	0

4.8 Outcome of Emergency Appointment

Most patients, 76% (n=37), attending for emergency appointments were given a follow-up appointment at the EDH. 14% (n=7) were placed on a GA waiting list and the remaining 10% (n=5) were discharged. Of the patients put on the GA waiting list, all were under the age of seven years old and 3 suffered from medical conditions. Table 4-6 shows data on patients placed on GA waiting list.

Table 4-6 – Table Showing Patients Whose Outcome was a General Anaesthetic

Age (years)	Medical History	Reason for Emergency
2	Healthy	Swelling
3	Healthy	Trauma
4	Asthma	Trauma
5	Autism	Does not Adhere to EAC - Caries
5	Healthy	Does not Adhere to EAC - Pain
6	Healthy	Does not Adhere to EAC - Pain
7	Cardiac condition	Swelling

5 Action Plan – Cycle 1

After cycle one was completed, four action points were decided upon.

5.1 Adapt EAC

The EAC would be adapted to differentiate between new and existing EDH patients.

For new patients the following were added:

- oncology referrals
- neonatal teeth causing problems

For existing patients, the following were added:

- Pain
- Lost anterior restoration or bridge
- Broken Denture

Exclusion criteria were also emphasised as follows:

- Lost posterior restoration with no pain
- Discharged patients without a new referral
- Patients older than 16 years old

5.2 Development of Standard Operating Procedure

Standard operating procedure (SOP) flowcharts were developed together with the administration team to ensure ease of use. SDCEP guidelines regarding triaging of emergency patients was referred to during development. They were discussed at the monthly departmental clinical governance meeting and approved for use. Following this they were disseminated to all clinicians and administrative staff via email.

The two flowcharts developed are Figures 5-1 and 5-2.

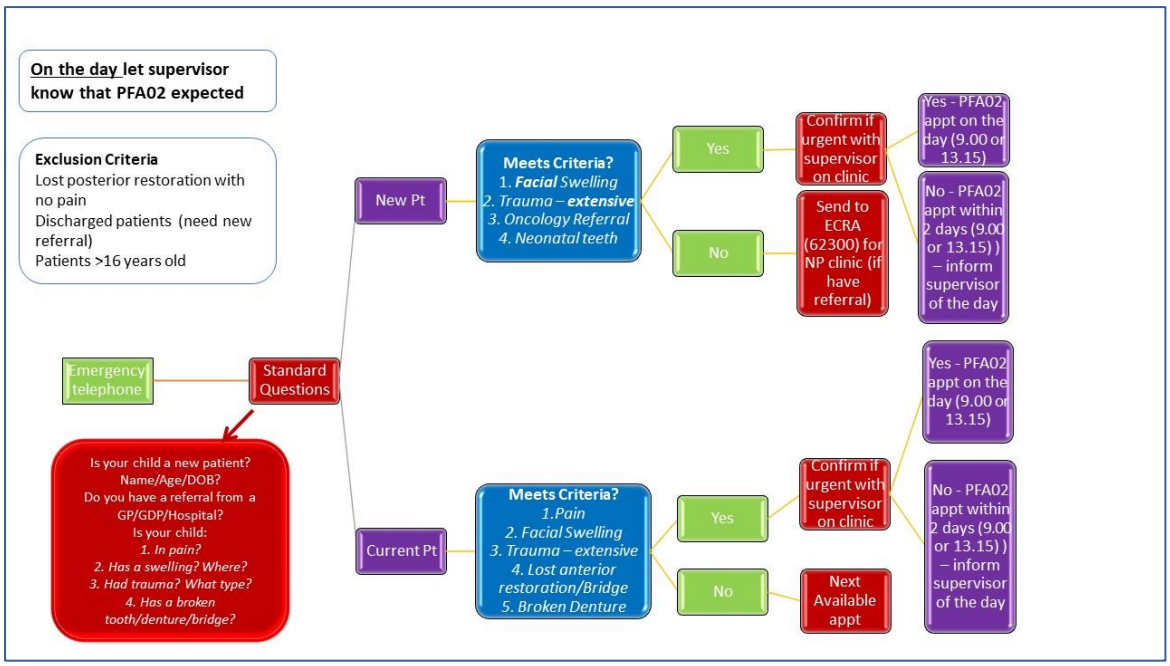

Figure 5-1 – Figure showing flowchart for emergency telephone call patients

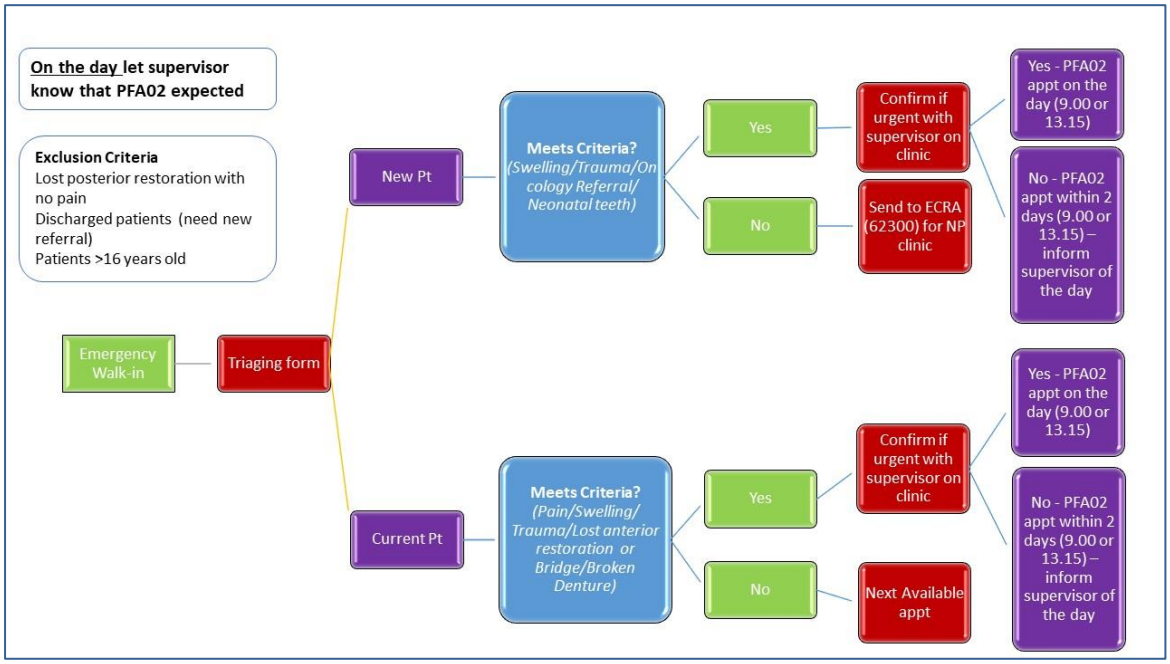

Figure 5-2 - Figure showing flowchart for walk-in emergency patients

5.3 Updating of Triaging Form

The department had a triaging form which had not been updated since 2007 (Appendix 2). This form was meant to be used for walk-in patients to make more efficient use of administrative and clinician time. This form was no longer being used.

The triaging form was updated to make it shorter and more user friendly. It was again discussed at the monthly clinical governance meeting and approved for use. This form is available at the administrative welcome desk to be given to parents who attend with children as walk-in patients. The new form can be found in Appendix 3.

5.4 Teaching on Pathways for Emergency Patients

After the first cycle, the results were disseminated at the monthly clinical governance meeting and a teaching presentation was given to the clinical and administrative staff regarding the new protocols, the updated triaging form and the SOPs. The SOPs were also shared with the administration team via e-mail from their administrators.

6 Results Cycle 2

Forty-nine patients from cycle 2 were analysed, these notes were obtained from clinical records of patients booked under the PFA02 code in EPIC (34 patients) and those that had 'Emergency' as a reason for attendance booked under other clinician codes (15 patients). The first result of note is that less patients were seen as emergencies during cycle 2 – 49 overall compared with 54 from cycle 1.

6.1 Demographic Data

In cycle 2, slightly more male (61%, n=30) than female (39%, n=19) patients attended as emergencies. The age ranged from one year to eighteen years old with an average age of 10.2. When dividing into age groups, the majority of dental emergencies were in patients between the ages of 8 - 12 and 13 - 16 with 33% (n=16) falling into both these groups. Table 6-1 shows how emergency appointments differed between the different age groups in cycle 2 compared to cycle 1.

Table 6-1 – Table showing distribution of dental emergency appointments by age groups in cycles 1 and 2.

Age groups	**Cycle 2**		**Cycle 1**	
	Numbers	**Percentage**	**Numbers**	**Percentage**
<=3	4	8	8	16
4-7	11	22	15	31
8-12	16	33	13	27
13-16	16	33	11	22
>16	2	4	2	4

6.2 Type of Emergencies

In cycle 1, the EAC at the paediatrics department of the EDH were swelling and dental trauma, in cycle 2 the new criteria was added. Overall, 86% of patients seen (n=42) followed the department EAC for emergency patients. From these, 18% (n=9) had swellings and 27% (n=13) had dental trauma. All new patients (100%, n=14) followed the EAC, whilst 80% of

follow-up patients (n=28) followed the EAC. The breakdown according to new patients and follow up patients can be seen in Tables 6-2 and 6-3.

Table 6-2 – Table showing type of emergency appointment for follow-up patients.

Follow-up patients	**Numbers**	**Percentage**
Broken/Lost Denture	3	9%
Lost Bridge	1	3%
Lost Restoration	9	26%
Pain	7	20%
Swelling	4	11%
Trauma	6	17%
Not Protocol	5	14%

Table 6-3 - Table showing type of emergency appointment for new patients.

New Patients	**Numbers**	**Percentage**
Swelling	5	36%
Trauma	7	50%
Oncology	1	7%

Figure 6-1 shows the comparison of new and existing patients meeting EAC in cycles one and two.

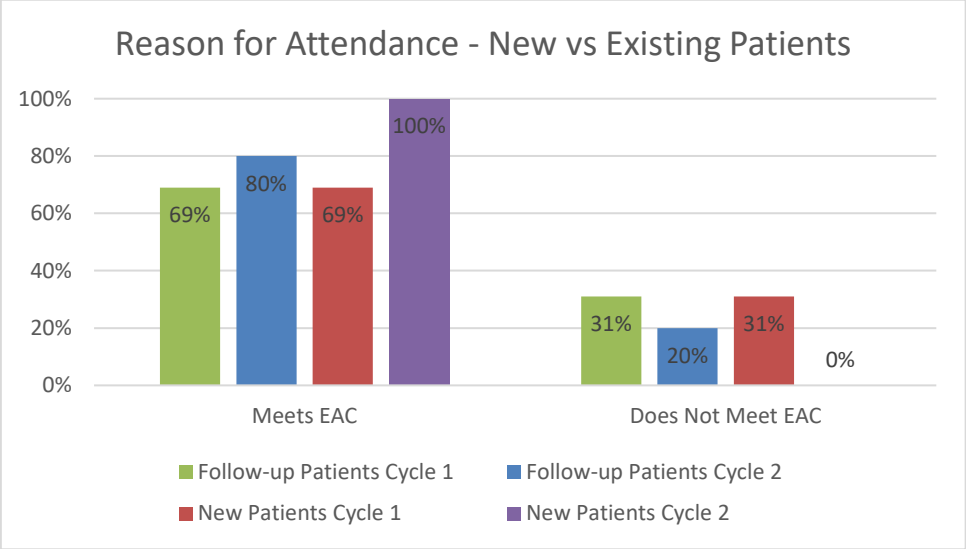

Figure 6-1 – Graph showing comparison between cycle 1 and cycle 2 for patients meeting EAC.

From the 13 trauma patients, 23% (n=3) suffered trauma to the primary dentition, and 77% (n=10) suffered trauma to the permanent dentition.

Reasons for attendance not adhering to emergency protocols are:

- Denture – Other (80%, n=4)
- Lost gold chain (20%, n=1)

When assessing male to female ratio of emergencies fitting EAC, we see that there were slightly more males attending with trauma (69%, n=9) than females (31%, n=4) and for patients not adhering to EAC, with 80% (n=4) male and having issues with their denture, and 20% (n=1) female (lost gold chain). Swelling also showed a slight skew, this time in favour of females at 78% (n=7) (Males - 22% n=2).

Dividing by age, once again trauma was most prevalent in the 8 – 12-year-old age group at 38% (n=5). Followed by the 4 to 7-year olds with 31% (n=4).

6.3 Time taken to see Patient

In cycle 2, the time taken to see patients was worse than cycle one. The majority of patients, 69% (n=33), were still seen within this time frame, however this was reduced from the 82% in cycle one. In cycle two, 42% (n=20) of patients were seen the same day they made contact

compared to the 51% (n= 25) in cycle 1. Figure 6-2 shows the date difference from patient contact to patient being seen for all patients.

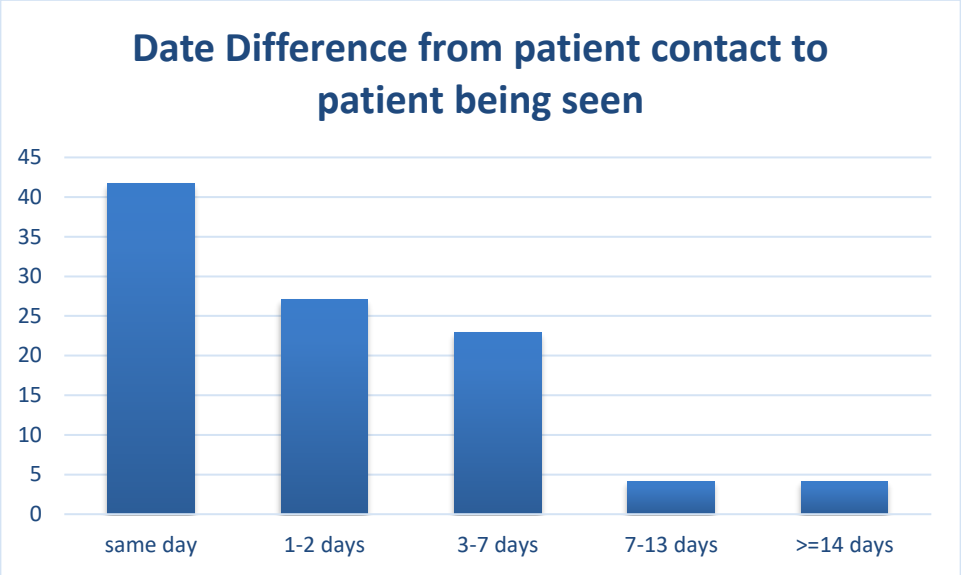

Figure 6-2 – Graph showing Date Difference from first contact to patient seen for emergency appointment

6.4 Grade of Clinician Seeing Patient

The majority of the emergency patients, 33% (n=16) were seen by consultants, followed by the postgraduates at 23% (n=11). The distribution of emergency patients seen by clinician, compared to cycle 1, can be seen in Figure 6-3.

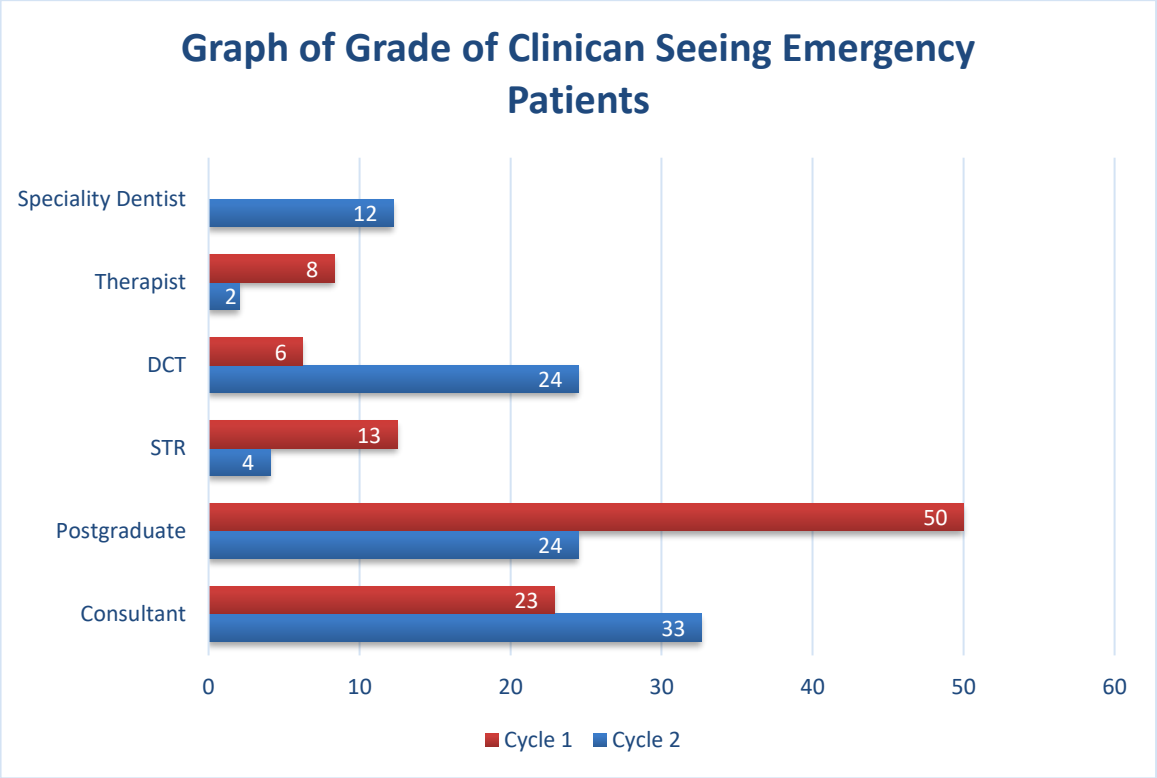

Figure 6-3 – Figure Showing Distribution of emergency patients by clinician – Comparing cycles

6.5 Medical History

71% (n=35) of patients were fit and healthy compared to 73% (n=36) from cycle 1. The remaining 29% (n=14) had a variety of medical conditions which can be seen in Table 6-4.

Table 6-4 – Table showing distribution of medical conditions seen

Medical Conditions	Numbers	Percentage out of Total	Percentage out of Medically Compromised
Oncology	2	4%	14%
Asthma	4	8%	29%
Kidney	2	4%	14%
Others	5	10%	36%
Epilepsy	1	2%	7%

'Others' conditions included:

- Chronic Pancreatitis
- Eczema
- Diabetes mellitus
- Anorexia
- Hip Dysplasia

6.6 Referral for Emergency Appointment

Unlike cycle 1, the majority of patients in cycle 2 were self-referred (59%, n=29). From these, all except one (57%) were already patients of the EDH. The next most common referral was by the GDP (29%, n=14). From these, 10% (n=5) were follow-up patients whilst 18% (n=9) were new patients. The rest of the patients were referred in the 'other' group. The distribution of referees from the 'other' group compared with cycle 1 can be seen in table 6-5.

Table 6-5 – Table showing where patients in 'other' category were referred from

Referred From	**Cycle 1**	**Cycle 2**
Hospital (non-specific)	1	2
Unknown	2	0
UCLH MaxFax	1	2
UCLH Oncology	4	0
Internal Referral	0	1
A&E	2	0

6.7 Intervention at Emergency Appointment

The most common intervention was ‘Other’ interventions at 45% (n=22). These treatments can be seen in Table 6-6.

Table 6-6 – Table showing other treatments carried out

Intervention	**Numbers**
Impressions	5
Restorations	7
Extractions	1
Referral to Orthodontics	1
CBCT	1
Fissure Sealants	1
Reattach gold chain	1
Recement preformed metal crown	1
Rebond bridge	1
Separator placements	1
Adjust/fit denture	2

From those 11 children (22%) receiving no treatment, one was a 2-month-old with an eruption cyst, 3 were in pain, 5 had a swelling and 2 had trauma. From those with a swelling, 4 were placed on a general anaesthetic list and one was given a follow-up appointment. From the trauma patients, one was discharged back to their GDP and one was given a follow-up appointment.

Division of interventions by group can be seen in Figure 6-4.

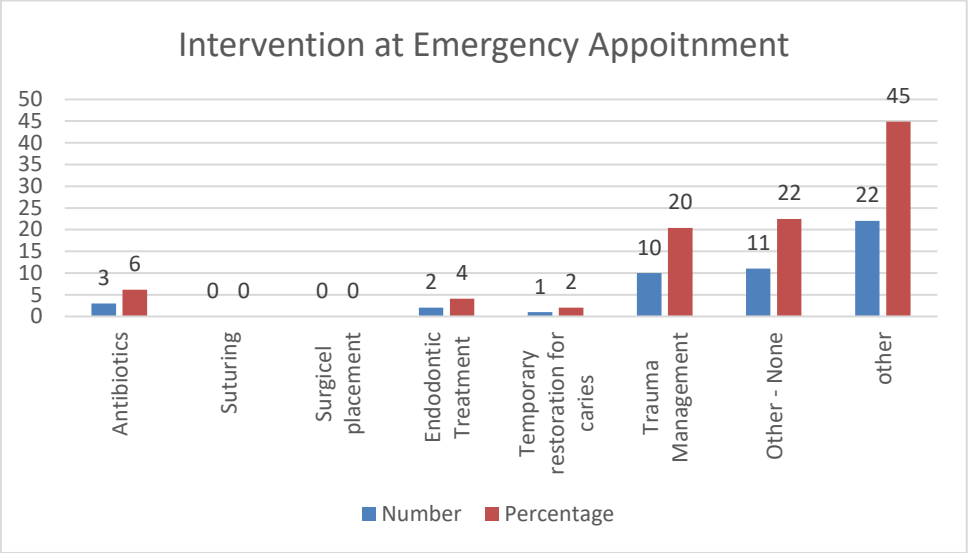

Figure 6-4 also shows that only 20% of patients (n=10) had trauma management. As mentioned previously, 27% (n=13) patients attended with trauma, and the interventions carried out for these patients can be seen in Table 6-7.

Table 6-7 – Table showing interventions for trauma at emergency appointment

Management	**Primary Teeth**	**Permanent Teeth**
Other	0	1 (Extraction)
No Treatment	0	2
Trauma Management	3	7

6.8 Outcome of Emergency Appointment

As in cycle 1, for cycle 2, 78% (n=38) of patients were given follow-up appointments. Similarly, the second most common outcome was placement on a GA list (16%, n=8) and the remaining 6% (n=3) were discharged. Table 6-9 shows the patients placed on GA waiting list.

Table 6-8 – Table Showing Patients Whose Outcome was a GA

Age	Medical History	Reason for Emergency
1	Healthy	Trauma
3	Healthy	Does not Adhere to EAC - Pain
6	Healthy	Swelling
7	Healthy	Swelling
9	Asthma	Does not Adhere to EAC - Pain
9	Healthy	Swelling
10	Healthy	Swelling
13	Healthy	Swelling

7 Action Plan – Cycle 2

After cycle 2, a three-point action plan was developed

1. Further face to face teaching of the whole team, both administrative and clinical staff. Going over the SOP flowcharts and triaging form.
2. Addition of ‘emergency sessions’ each week to reduce the burden of emergency patients on booked clinics
3. Annual re-audit

8 Discussion

This two-cycle audit of emergency patients in the paediatric dental department showed improvement in some areas between the two cycles. This positive change shows quality improvement within the department and a willingness to change to improve patient care.

It is important to note that two significant changes occurred in the department between the two cycles and these may have affected the outcomes of some of the results. The first change was one from paper notes to electronic health records. The second change was that the dental hospital moved location in October 2019, at the beginning of cycle 2.

The first improvement between the two cycles is that less emergency patients attended in cycle 2 (49) than in cycle 1 (54). This change arose because of the action plan implemented after cycle 1, particularly the development of a standard operating procedure (SOP) which made triaging of emergency patients by administrative staff more efficient. The location change may have impacted the number of emergency patients seen however, clinics were still being carried out and emergency patients were signposted to the department and not turned away.

Prior to this audit, there was no SOP for emergency patients in the department. Establishing this SOP not only reduced the number of emergency patients seen, but also reduced the number of cases which did not adhere to the emergency acceptance criteria (EAC).

In cycle 1, only 59% of patients seen adhered to the EAC, which improved to 86% in cycle 2. When breaking down into new patients and existing patients, an improvement was again seen between the cycles, particularly with new patients meeting EAC increasing to 100%. The reasons for not reaching the standard could have been that patient's known to the EDH clinicians felt they would benefit from emergency treatment, as well as that 4 of these patients had issues with dentures replacing anterior teeth and the argument may be made that aesthetic concerns arising from not having a front tooth can affect quality of life (El-Kalla et al, 2017).

Of note in both cycles is that children older than 16 years old were seen, despite the fact that the department should only see children under the age of 16. All four of these young adults were previous patients of the department and had fractured anterior restorations or lost a resin bonded bridge previously provided by the department.

This audit compares to some of the literature about dental emergencies and gender, Martens et al found that the male to female ratio for dental emergencies was 1.28:1 and Muller et al

found it at 1:1; for this audit, cycles 1 and 2 had ratios of 1:1 and 1.6:1 respectively (Martens et al., 2018; Muller et al., 2017). The percentage of trauma emergency patients different considerably between cycle 1 (49%) and cycle 2 (27%). A possible reason for this could be that cycle 1 took place over the summer months and children may have been more likely to be playing than during the school term. When it comes to male to female ratios of the trauma patients, these are 1:1 and 1.6:1 in cycles 1 and 2 respectively. A review of the literature in 2008 states that males are at least double the risk of suffering dental trauma than females. However, it goes on to say that this ratio is decreasing as traditional gender games are becoming gender neutral (Glendor et al., 2008).

In cycle 1, 4 patients were referred from UCLH Oncology department. Prior to any oncology treatment, it is essential that patients have a full oral and dental assessment to remove oral infections and develop a preventive regimen. Extractions should be carried out at least 10 days before beginning chemo- or radiotherapy (Kumar et al., 2018). For these reasons, I felt it was important to add oncology referrals to the acceptance criteria for new patients to ensure they have dental treatment carried out in a timely manner prior to commencing oncology treatment.

In cycle 1, the majority of patients were seen within 2 days. Whilst still maintaining a majority, the number of patients seen within this timeframe, reduced to 69% in cycle 2. Reasons for this could include the change in scheduling system when switching to electronic health records, as well as administrative troubleshooting when booking patients due to the confusion associated with moving into a new building. According to SDCEP guidance, all emergency and urgent care should be seen within 24 hours. However, this guidance is aimed at primary care clinicians, not referral-based services such as the EDH (SDCEP, 2017).

In cycle 1, 43% of patients were GDP referrals, this reduced to 29% in cycle 2. The reason for this could be that new patients referred from their GDPs were being triaged more effectively and less patients not adhering to EAC were given emergency appointments. In both cycles, those referred by the ‘other’ category were all patients with acute emergencies and included dental trauma, swelling, neonatal teeth causing ulcers and an eruption cyst or oncology referrals. All these patients were seen within 24 hours.

Appointment outcomes ranged from follow-up appointments to discharging the patient. However, in both cycles, the majority of patients were seen for a follow-up appointment. Many of the children placed on a GA waiting list were young children with acute swelling or trauma, or pain due to caries. Reasons for placement on the GA list included age, co-operation and complexity of treatment.

The improvements gained between the two cycles of this audit highlights the importance of a well-planned SOP for emergency patients. In cycle 2 the time taken to see patients was further from the standard than was hoped. The action plan after cycle 2 seeks to address these shortcomings by further emphasising the current SOPs to all staff members, both clinical and administrative, as well as by emphasising the need for dedicated emergency slots on existing clinics to reduce the burden of emergency patients on existing clinics.

9 Conclusion

The audit showed that gains are still necessary to reach the standards chosen, however implementation of the SOP for management of emergency patients increased the numbers to:

- 100% of new patients meeting EAC.
- 80% of existing patients meeting EAC.

The department still needs to improve the time taken to see emergency patients; however, the implementation of emergency slots should address this issue.

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Appendix 1

Eastman Dental Hospital - Paediatric Department

Adherence To Acceptance Criteria for Emergency Patients Audit – Booked under PFA02 Code

Patient's Sticker

Age_____

Gender: ☐ Male

☐ Female

☐ Other

Unknown

Type of emergency: ☐ Swelling

☐ Dental trauma

Not according to protocol; details:_____

Date appointment was booked:_____

Date of appointment: _____

Grade of clinician seeing patient:

☐ Consultant

☐ Postgraduate

☐ STR

DCT

Speciality Doctor

Is the patient special needs or medically compromised: ☐ Yes ☐ No

If yes, details:_____

Referred by:

☐ GDP

☐ CDS

GP

Self-referred

☐ Other: _____

Intervention at emergency appointment:

☐ Antibiotics

☐ Suturing

☐ Surgicel placement

☐ Endodontic Treatment

Temporary restoration for caries

Trauma Management (e.g. Splint/Fragment reattachment etc.)

Other: _____

Outcome of appointment:

☐ Follow-up appointment – outpatient

☐ Placement on GA list

☐ Referral to another department/hospital (which:_____)

Discharge

Other: _____

Appendix 2

Old Triaging Form

February, 2007

Department of Paediatric Dentistry, Eastman Dental Hospital

Casualty Triage Form

Date / / **Time** _____ **am / pm** **Nurse** _____

Name of patient _____ **DOB / /**

Age ___ **Gender – M / F**

Reason for attendance = _____

(eg Caries, Trauma, Swelling, Fractured restoration, with PAIN** – yes or no)

Registered with GDP – Yes / No **If yes, contacted – yes / no**

Referral letter – Yes / No

Eastman patient – Yes / No **Hospital no =** _____

History of present complaint = _____

COMPLAINT	Question	Answer
Caries	When noticed?	
	Eating?	
	Sleeping?	
	Medication?	
Trauma	When?	
	Where?	
	How?	
	Anyone else involved?	
	AVULSION?	
	Deciduous / Permanent / Both	
	Tooth stored in anything? Since when?	
Swelling	When noticed?	
	In mouth / face / both?	
	Medication?	
	Temperature?	
Fractured restoration	When?	
	How?	
	Sharp edge?	
Other		
** Pt in PAIN?? Yes / No	Details	

Any patient specifics eg – interpreter, wheelchair, special needs _____

Clinician discussed with = _____

Outcome = _____

1

Appendix 3

New Triaging Form

Department of Paediatric Dentistry, Eastman Dental Hospital
Casualty Triage Form

Date:_____ **Time:** _____ **Nurse:**_____
New patients: Yes/No
Name of Patient:_____
DOB:_____ **Age:** _____ **Gender:** M/F

Reason For Attendance:

Trauma	*Oncology referral*	Fractured restoration with pain
Swelling	Fractured anterior	Neonatal teeth
Pain	restoration	Broken Denture

Other: _____

Registered with GDP:	**If Yes, Contacted GDP?**	**Referral Letter available:**
Yes/No	Yes/No	Yes/No

COMPLAINT	QUESTION	ANSWER
Pain	Since When?	
	Eating and sleeping?	
	Medication?	
Trauma	When?	
	Where?	
	How?	
	Anyone Else involved	
	Avulsion? (tooth completely out of mouth)	
	Adult/Baby tooth?	
	Tooth/Fragment stored in liquid? What and for how long?	
Swelling	Since When?	
	Medication?	
	Temperature?	
Fractured Restoration	Where?	
	Pain?	
	When?	
	How?	
	Sharp edge?	
Other	Any Relevant details?	

Patient Specifics? (eg. Interpreter, wheelchair, special needs)

_____

Outcome: Emergency slot(PFA02)/Next available appointment

Caries Case

A Clinical Case Report

Submitted in partial fulfilment of the requirements for the Degree of

Doctor of Dentistry (Paediatric Dentistry)

UCL

Submitted by

Jasmine May Cachia Mintoff

Word count: 1928

UCL Eastman Dental Institute,

256 Gray's Inn Road,

London WC1X8LD,

SUMMARY

A.A., a 4 year old girl, was referred to the Paediatric Department by the community dental service due to dental pain and multiple carious teeth. On presentation, A.A. had no pain and was co-operative for examination. She had a total of 8 cavities in her primary teeth.

Dental treatment was carried out under inhalation sedation. It included a combination of prevention, extractions and preformed metal crowns.

PATIENT DETAILS

Initials:	A.A.
Gender:	Female
Age at presentation:	4 years, 4 months
Age at last review:	5 years, 8 months

PRE-TREATMENT ASSESSMENT

History of presenting complaint

- C/O: nil
    - No history of swelling, analgesia required to sleep on multiple occasions
    - No antibiotics

Relevant Medical History

- Born full term
- Immunisations up-to-date

Drug History

- None
- No known drug allergies

Dental History

- CDS
- Regular attender
- No treatment carried out

Social History

- Attended with father
- Lives with parents and 5 sisters
- Reception year at school

Diet

- Regular fizzy drinks and milkshakes
- Fruits as a snack
- One bottle at night - milk

Oral Hygiene

- Brushes twice daily, supervised with 1000ppm Fluoride toothpaste
- Rinses not spits

Habits

- None

CLINICAL EXAMINATION

Extra-Oral Examination

- NAD

Intra-Oral Examination

- Soft tissues: NAD
- Fair oral hygiene
- Occlusion – Full primary dentition, spacing
- Teeth present:

E	D	C	B	A	A	B	C	D	E
E	D	C	B	A	A	B	C	D	E

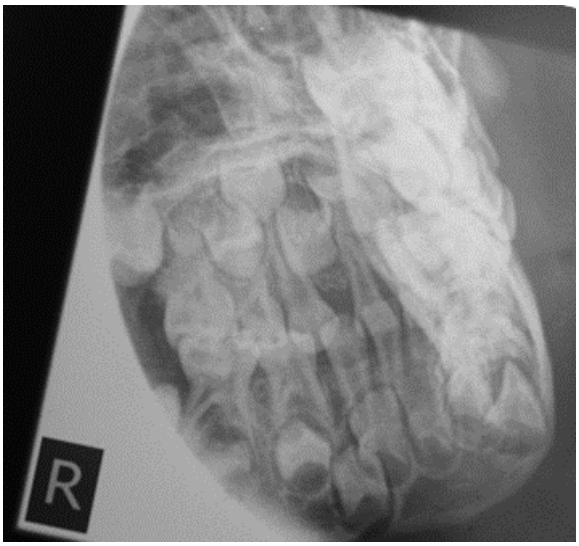
- Carious Teeth:

E	D							D	E
E	D							D	E

Radiographic Examination (21/05/2018)

Radiographs taken:

- R+L Bimolar radiographs
- Justification – Caries



Figures 4 and 5: Right (Grade 2) and Left (Grade 1) Bimolar Radiographs –

Radiographic Findings:

- No furcation Lesions
- Caries on URE (MO), URD (DO), ULD(DO), ULE (MO), LLE (MOD), LLD (DO), LRD (DO)
- Deep caries on LRE (MOD)

PRE-TREATMENT PHOTOGRAPHS (27/09/2018)

Figure 1: Anterior View

Figure 2: Maxillary Occlusal View (Mirror View)

Figure 3: Mandibular Occlusal View (Mirror View)

DIAGNOSTIC SUMMARY

- Early childhood caries
- Anxious young patient

AIMS AND OBJECTIVES OF TREATMENT

- Improve oral hygiene
- Restore Oral Health
- Promote positive attitude towards dentistry
- Monitor development of permanent dentition

PROVISIONAL TREATMENT PLAN

1. Acclimatisation and prevention
    a. Establish a preventive regimen consistent with the Department of Health preventive toolkit
    b. Use of non-pharmacological behaviour management
2. Restorations
    a. Preformed metal crowns (PMC) using Hall technique on URE, URD, ULD, ULE, LLE, LLD, LRD
3. Extraction LRE under Inhalation sedation (IS)
4. Maintenance and Follow-up
    a. Clinical review every 6 months
    b. Radiological review every 6 months
    c. Fissure sealants on the first permanent
5. A.A.'s father was made aware of the need for a general anaesthetic if co-operation in the dental chair was lost

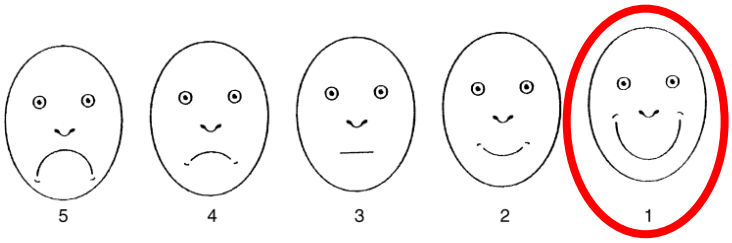
TREATMENT PROGRESS AND DENTAL MANAGEMENT

Visit 1: 21/05/2018

- Patient attended the new patients' clinic with father
- Medical history was taken
- Extra-oral and intra-oral clinical examination
- Radiographs:
    - Right & Left bimolars
- Treatment options discussed with patient's father
- Preventative advice given as per department of health toolkit –
    - Brushing twice a day – 1450ppm Fluoride toothpaste
    - Supervised
    - Spitting not rinsing after brushing
    - Reduce frequency and amount of sugars
    - Stop night-time bottle
- Fluoride varnish on all teeth

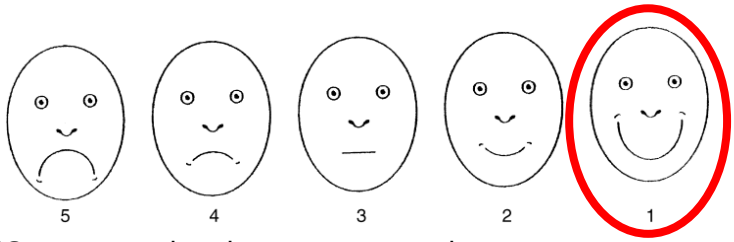
Visit 2: 27/09/2018

- C/O: nil
- Toothbrushing advice reinforced
- "Tell-Show-Do" behavioral management technique
    - URD, ULD:
        - Topical LA placed interdentally with microbrush
        - separators placed distal to URD, ULD
- Post-operative instructions were given
- Treasure card was given
- Facial Image Scale (FIS):



Visit 3: 9/10/2018

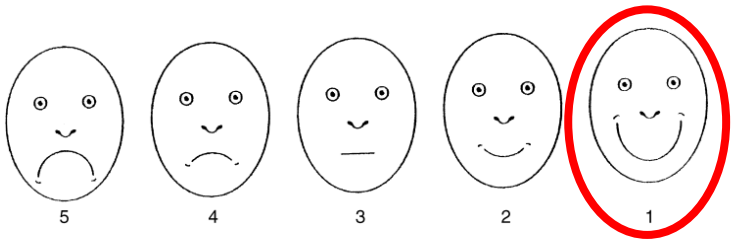
- C/O: nil
    - both separators were lost on day before appointment
    - sufficient space when contacts flossed
- URD, ULD – Hall crowns
    - PMC size 5 for URD, Size 4 for ULD
    - Topical LA



    - PMCs cemented and excess removed
    - Contact points flossed
- LRD, LLD – separators
        - Topical LA
        - separators placed distal to LRD, LLD
- Stickers given
- FIS:

Visit 4: 16/10/2018

- C/O: nil
- O/E:
    - LRD separator was lost in waiting room
    - sufficient space when contacts flossed
- LRD, LLD – Hall crown
    - PMC size 4 chosen for both teeth
    - Topical LA
    - Cemented as before
- URE, ULE – separators
        - Topical LA placed interdentally with microbrush
        - separators placed mesial to URE, ULE
- Stickers given
- FIS:

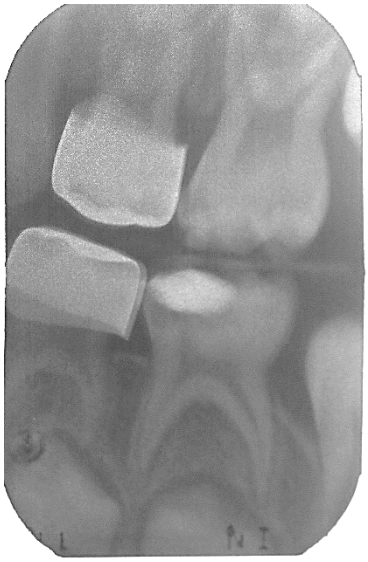


Visit 5: 23/10/2018

- Patient was not brought for this appointment and there was no answer when parents were called. A new appointment was booked.
- Parents contacted later that day and we were informed separators had fallen out.

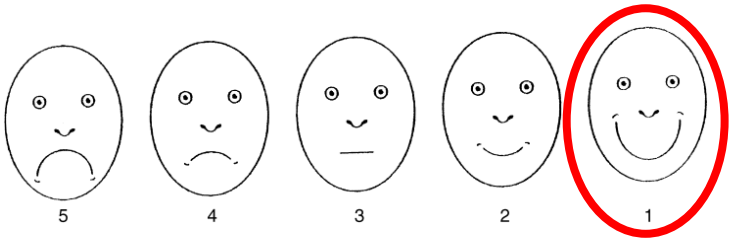
Visit 6: 29/01/2019

- C/O: pain on eating/sweet LRE
- O/E:
    - LRE significantly broken down
    - No evidence of swelling or sinus at present
    - LLE – large temporary restoration placed by GDP
- URE, ULE – separators
        - Topical LA
        - separators placed mesial to URE, ULE
- New right and left vertical bitewing radiographs were requested
    - Justification: Caries
    - LRE showed furcation pathology
    - LRD PMC tilted
    - LLE showed large cavity however dentine bridge was present over the pulp chamber



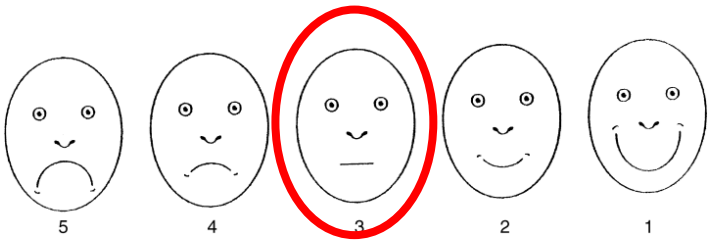
Figures 6 and 7: Right and Left Vertical Bitewing Radiographs (Grade 1) – 29/01/2019

- Discussed options for treatment with father:
    - Extraction of the LRE under IS
    - PMC LLE and monitor
- Fluoride varnish on all teeth
- Stickers given
- FIS:



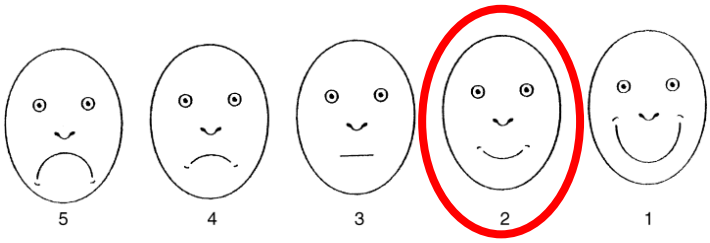
Visit 7: 5/02/2019

- C/O: pain LRE
- O/E:
    - both separators were lost 1 day prior to appointment
    - sufficient space when contacts flossed
- URE, ULE – Hall crown
    - PMC size 3 chosen for both
    - Topical LA
    - PMCs cemented on and excess material removed
    - Contact points flossed through
- Patient became agitated due to pressure when cementing on PMCs
- Consent for treatment under inhalation sedation signed
- Stickers given
- FIS:



Visit 8: 7/03/2019

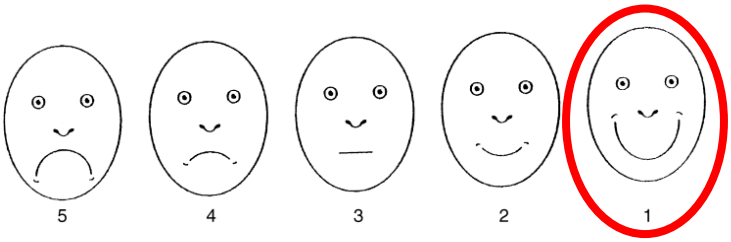
- C/O: nil
- Confirmed patient had breakfast
- “Tell-Show-Do” behavioral management technique.
- LRE – Extraction
    - IS → 70% O2, 30% N2O
    - Topical LA and 2.2ml lignocaine with adrenaline, 1:80 000 using the Wand®
    - Elevators and forceps extraction
    - Haemostasis achieved
    - 100% O2 for 5 minutes – Normal recovery
- Post-operative instructions were given, verbal and written
- Treasure was given
- Patient praised for good behaviour
- Stickers
- FIS:



Visit 9: 9/04/2019

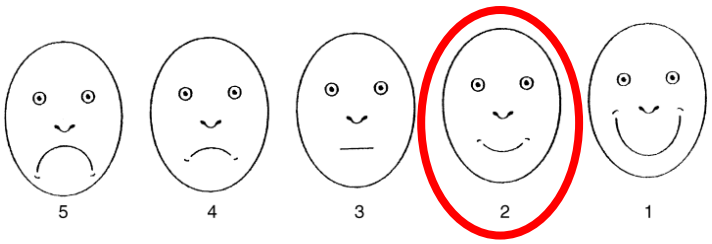
- C/O: Lost PMC LRD
- O/E:
    - LRD PMC missing
    - sufficient space when contacts flossed
- LRD Hall crown
    - PMC size 4 chosen
    - Topical LA
    - PMC cemented as before
- LLE – separator
        - Topical LA placed interdentally with microbrush
        - separators placed mesial to LLE
- Stickers given

- FIS:



Visit 10: 10/04/2019

- C/O: Lost separator that morning
- LLE Hall crown
    - PMC size 4 chosen
    - Topical LA
    - PMCs cemented as before
- Patient began agitated due to pressure when cementing crown
- Clinical photographs taken
- Fluoride varnish on all teeth
- Stickers given



Visit 11: 11/11/2019 (6 month Review)

- C/O: nil
- O/E:
    - Mobile LRA
- Prevention reinforced
- Fluoride varnish on all teeth

POST-TREATMENT PHOTOGRAPHS (10/04/2019)

Figure 8: Anterior View

Figure 9: Maxillary Occlusal View

Figure 10: Mandibular Occlusal View

LONG TERM TREATMENT PLAN AND FUTURE CONSIDERATIONS

- Monitor restorations.
- Fluoride varnish application every 3 months.
- Radiographic reviews.
- Fissure seal first permanent molars when they erupt.

DISCUSSION AND REFLECTION

Overall therapy was successful, and the final outcome was a caries free child who was learning to have a positive attitude towards dental care and treatment. At initiation of therapy, the patient had no previous dental experience, therefore it was important to introduce dental treatment gradually using non-pharmacological behavioural management techniques such as 'Tell, Show, Do' to reduce fear of the unknown. All treatment was completed chairside, avoiding general anaesthesia.

Prevention

Prevention is one of the most important parts of dental treatment and it should be the starting point of any treatment plan. Prevention advice followed the Department of Health's 'Delivering better oral health: an evidence based toolkit for prevention' (3rd edition) (1).

In this case the patient was already brushing her teeth twice a day with a toothpaste containing over 1000ppm Fluoride. We encouraged her father to change to a 1450ppm fluoride as she is high caries risk and encouraged spitting rather than rinsing after brushing. I advised A.A.'s father to supervise or help with brushing using the Modified Bass Technique (1).

Aside from having a high sugar diet, A.A. was still having a bottle of milk at night after brushing her teeth. I advised against this explaining how this can contribute to caries and the dental advice is to stop this practice after the age of one year. I also encouraged a reduction in the frequency and amount of sugars in A.A.'s diet (1).

Behaviour Management

It is widely accepted that behavioural management techniques are the cornerstone of paediatric dentistry. Without management of a child's behaviour, dentistry cannot be carried out efficiently (2).

The aim of non-pharmacological behavioural management techniques is to build a rapport with the child and their parent thus allowing high quality dentistry to be carried out. One of the traditional behaviour management techniques is desensitisation, where a child is gradually introduced to dental procedures (2). I used this technique with A.A., I initially introduced her to simple treatment (separators and preformed metal crowns) and eventually she was able to cope with an extraction under local anaesthetic with the help of inhalation sedation.

I decided to use IS as a pharmacological behavioural management adjunct to treatment for the extraction of the LLE as this was A.A.'s first extraction and she was anxious. Nitrous oxide is a safe method of managing a child's anxiety. It works quickly and is rapidly expelled from the body's systems at the end of treatment. There are minor side effects including nausea and vomiting but neither of these were experienced by A.A. during any of the interventions (3).

The Facial Images Scale was used to assess A.A.'s anxiety at the end of treatment at each appointment. The Facial Images Scales is a validated dental anxiety scale for children (4).

Treatment

When A.A. first presented, she was not in any pain, thus the initial treatment was simple restorations with PMCs using the Hall Technique. This technique does not necessitate the use of tooth preparation or local anaesthetic and evidence shows that the hall technique has a success rate of 97% after 5 years (5). Upon reflection I could speculate that A.A. may not have required separator placement prior to cementation of the PMCs as they were almost always lost between appointments. However, radiographs showed tight contacts and the separators seem to have been lost after being in place for several days. This could be due to the patient picking them out once the contact points had loosened.

Painless dentistry is vital to ensure patient co-operation and comfort (6). When it came to the choice of local anaesthetic for the extractions, we considered several parameters. As A.A. was only four years old, lignocaine was the anaesthetic of choice. This was delivered via buccal infiltration using the Wand® computerized delivery system. Studies have shown that anesthesia via the Wand® is less painful, and better accepted when delivered slowly and well controlled, than traditional methods of delivering local anesthetic (6).

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Medically Compromised Child Case

A Clinical Case Report

Submitted in partial fulfilment of the requirements for the Degree of

Doctor of Dentistry (Paediatric Dentistry)

UCL

Submitted by

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Word count: 2017

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SUMMARY

J.O., a 6 year-old boy, was referred by his Consultant Paediatrician with dental pain and caries. Medically, he has Osteogenesis Imperfecta (OI) Type I and is on intravenous bisphosphonate therapy. On presentation, J.O. had caries, trauma and gingival recession.

Dental treatment was carried out under inhalation sedation and local anaesthetic and included restorations, preformed metal crowns and extractions. Prevention advice, habit stopping advice and topical fluoride application were also provided.

PATIENT DETAILS

Name:	J.O.
Gender:	Male
Age at presentation:	6 years, 2 months
Age at last review:	8 years, 4 months

PRE-TREATMENT ASSESSMENT

History of presenting complaint

- Dental pain – lower left side
    - No history of swelling
    - Occasional analgesia to sleep
    - No antibiotics
- Discolored anterior tooth
    - Trauma – playing with friends 1.5 years previously
    - No pain

Relevant Medical History

- Born: 35 weeks
- Incubator – 2.5 weeks
- Jaundice at birth
- Type 1 OI; Diagnosis at 11 months (COL1A1 mutation)
- Yearly reviews at Specialist OI Service
- Multiple fractures including:
    - Legs, 3 times
    - Thumb
- Glue-ear
- Immunisations up-to-date

Drug History

- Vitamin D, Syrup
- Iron Supplements
- Pamidronate IV infusions, 20mg for 2 days every 3 months, Local Hospital

Dental History

- Not registered at GDP

Social History

- Lives with mother (pregnant) and sister (24 months)
- Mainstream school, Year 1

Family History

- No family history of OI

Diet

- Breast fed for 8 weeks, bottle fed for 1 year
- Low Sugar Diet

Oral Hygiene

- Brushes twice a day, 1000ppm NaF, supervised, rinsing

Habits

- None reported

CLINICAL EXAMINATION

Extra-Oral Examination

- No blue sclera or short stature

Intra-Oral Examination

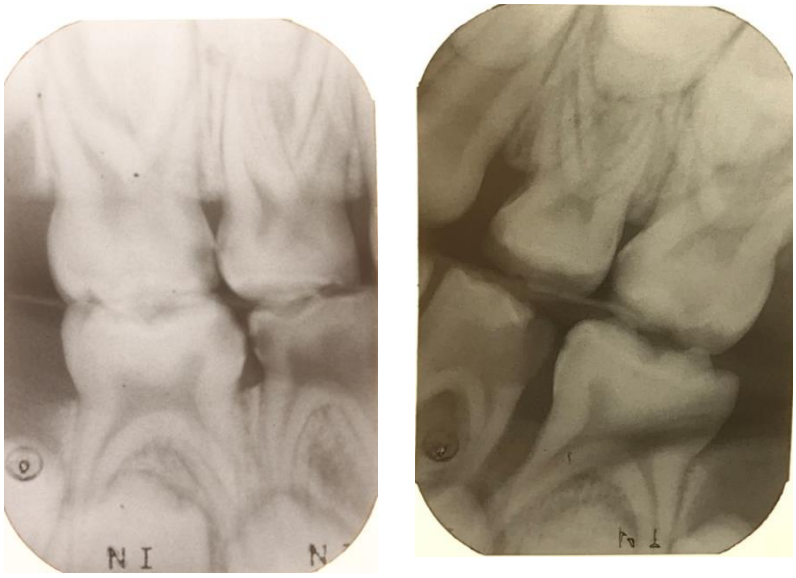
- Soft tissues: Sinus LLD
- Gingival recession upper incisors
- Good oral hygiene – Debris index score 0.3 (Greene and Vermilion, 1964)
- Occlusion – Full primary dentition, spacing
- No discolouration or tooth wear suggesting DI
- Anterior spacing with AOB
- Teeth present (Caries):

$E^{(MOD)}$	$D^{(DO)}$	$C^{(B)}$	B	A	A	B	$C^{(B)}$	$D^{(DO)}$	$E^{(MOD)}$
$E^{(MO)}$	$D^{(DO)}$	C	B	A	A	B	C	$D^{(DO)}$	$E^{(MO)}$

RADIOGRAPHIC EXAMINATION (11/12/2017)

Radiographs taken:

- Vertical Bitewings
- Upper Standard Occlusal
- Justification – Caries assessment/ Trauma



Figures 1 and 2: Right (Grade1) and Left (Grade 2) Vertical Bitewings

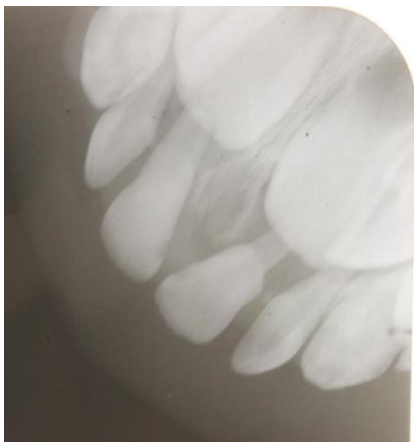


Figure 3: USO (Grade 2)

Radiographic Findings:

- Intreradicular radiolucency associated with LLD
- Caries on URE, ULE, LLD, LRD, LRE
- No radiographic features of DI
- Anomaly/ resorption: apical 1/3 ULA
- Accepted artefact over LLE crown

PRE-TREATMENT PHOTOGRAPHS (11/12/2017)

Figure 4: Anterior View

Figure 5: Maxillary Occlusal View (Mirror)

Figure 6: Mandibular Occlusal View (Mirror)

Diagnostic Summary

- Caries affecting all primary molars and upper canines
- Non-vital LLD
- Trauma ULA
- Gingival recession
- Dental anxiety

Aims and Objectives of Treatment

- Alleviate Pain
- Restore Oral Health
- Promote positive attitude towards dentistry
- Monitor development of permanent dentition
- Manage safely in line with medical needs
- Monitor gingival recession

Treatment Plan

- Acclimatisation and prevention – Consistent with the Department of Health preventive toolkit
- Liaise with medical team about precautions for extraction needed
- Rehabilitation using Inhalation sedation and local anaesthetic:
    - Extraction of the LLD
    - Preformed metal crowns (PMC) on URE, ULE, LRD, LRE
    - Composite restorations on URD, URC, ULC, ULD, LLE
- Maintenance and follow-up:
    - Regular clinical review
    - Radiological review every 6 months
    - Monitoring of ULA, clinically and radiographically
    - Monitor gingival recession
    - Fissure seal first permanent molars

J.O.'s mother was made aware of the need for a general anaesthetic if co-operation was lost

TREATMENT PROGRESS

For all appointments:

- Attended with mother
- Non-Pharmacological behaviour management (NPBM)
    - ‘Tell-Show-Do’
    - Positive reinforcement
    - Treasure card sticker

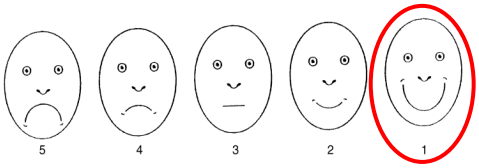
- Pharmacological behaviour management
    - LA – Articaine 4%, 1:20000 adrenaline
    - IS – titrated to patient (70%/30%)
- Facial Images Scale (FIS)

Visit 1: 11/12/2017

- Assessment as above
- Prevention as per toolkit – brush with 1450ppm Fluoride toothpaste twice a day, brushing should be supervised, spit not rinse after brushing and 3 monthly fluoride varnish
- Letter to OI consultant

Visit 2: 8/01/2018

- Correspondence from OI team – happy to proceed with dental treatment
- Non-pharmacological behaviour management
- IS and LA – Extraction LLD



Visit 3: 7/02/2018

- Ulcer noted LRC gingival margin – mother revealed patient had nail scratching habit (Figure 7)
- Full history of gum picking habit – advised to stop habit
- IS – attempted PMC LRE – did not seat well
- LA – PMC removed, GIC in cavity
- Co-operation lost

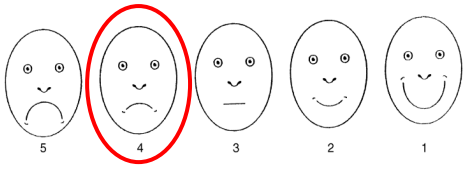


Figure 7: Ulcer between LRB and LRC (7/02/2018)

Visit 4: 13/02/2018, Emergency

- Facial swelling associated with LRE, temperature 37.2ºC
- Radiographs (Figure 8) + Clinical Photograph (Figure 9)
- Caries has now extended to pulp in LRE
- Decision to give antibiotics and rebook for extraction with IS (amoxicillin 500mg TDS, 5/7 oral suspension)
- Advised to attend A&E if not resolved

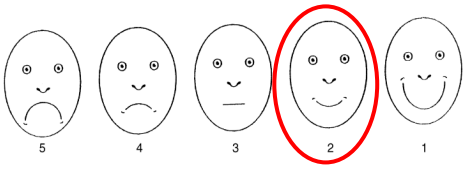
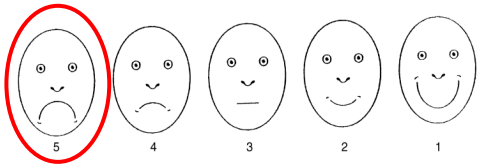


Figure 8: Buccal swelling around LRE (13/02/2018)

Figure 9: Right Bimolar (Grade 1)

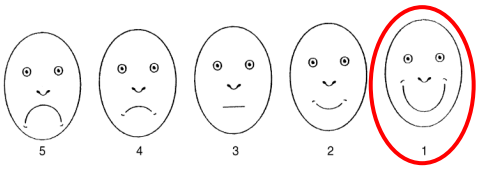
Visit 5: 21/02/2018

- NPBM, IS and LA
- XLA LRE – cried
- PMC LRD
- C/O pain therefore 250mg of Calpol® was administered post operatively



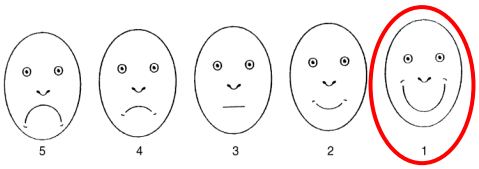
Visit 6: 07/03/2018

- NPBM, IS and topical LA
- Separators URE, ULE



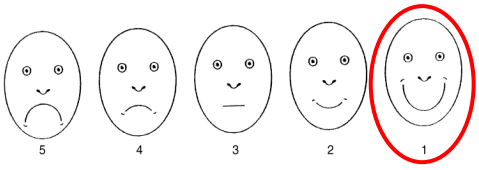
Visit 7: 12/03/2018

- Attended at wrong time, decision to do treatment this time (mother pregnant – difficult to travel) without IS (not available)
- Topical LA
- PMC URE, ULE



Visit 8: 16/04/2018

- IS
- Composite restorations – Buccal URC, ULC
- Fluoride varnish
- Discussion with mother: Cavities URD, ULD, LLE – shallow and self-cleaning – monitor



Visits 9, 10: 4/7/2018, 6/08/2018

- Patient not brought
- Mother gave birth/ new born in hospital

Visit 11: 15/08/2018, Review

- Lost restoration ULC
- Gingival recession noted together with inflammation
- Reinforced advice to stop gum picking habit
- Mobility URA, ULA – radiograph (Figure 10) revealed ULA close to exfoliating; query pathological resorption ULA – decision to monitor and await exfoliation due to lack of symptoms and history of traumatic extraction

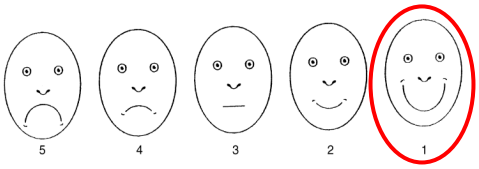


Figure 10: USO (Grade 1) radiograph (15/08/2018)

Visit12: 17/10/2018

- Patient reduced gum picking habit (Figure 11)
- Composite restoration – Buccal URC
- Fluoride varnish

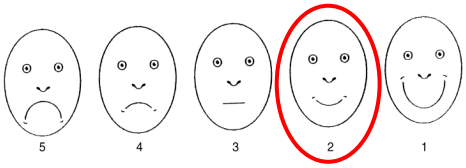


Figure 11: Anterior view showing gingival recession on primary canines and incisors (17/10/2018)

Visit 13: 19/03/2019

- Patient not brought as having IV Bisphosphonate infusions

Visit 14: 02/04/2019, Review

- Recession progressing
- Mobile LLC – radiograph (Figure 12) – no pathology
- Reiterated importance of stopping habit

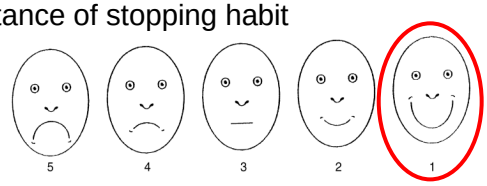
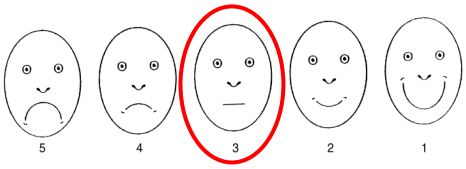
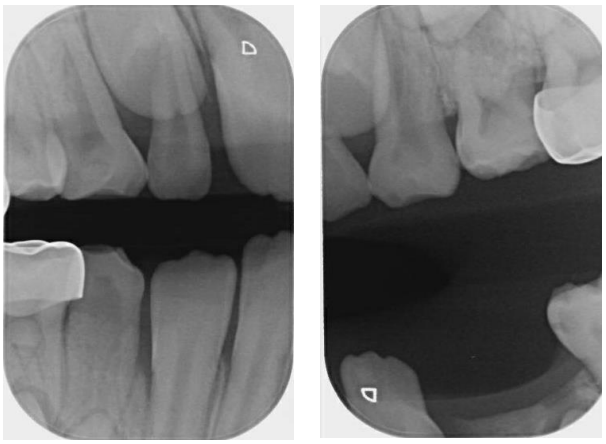


Figure 12: PA radiograph (Grade 1) (02/04/2019)

Visit 15: 30/10/2019, Review

- Bitewing radiographs taken (Figures 13, 14)
- New caries: ULD, LLE
- Staining UL6, LR6
- Attempted FS UL6 with GIC – patient could not cope
- <u>New Treatment plan</u> under IS:
    - PMC ULD, LLE
    - FS UL6, LR6
    - Fluoride varnish

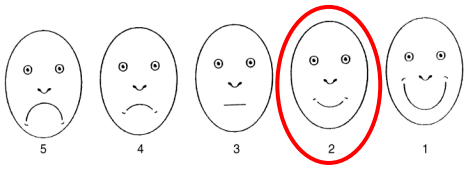




Figures 13, 14: Bitewing radiographs (Grade 1) (30/10/2019)

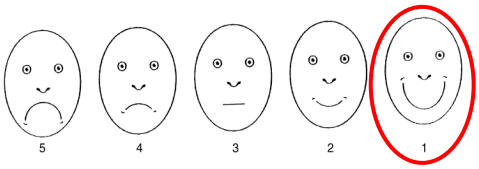
Visit 16: 18/11/2019

- GIC on UL6 in situ – decision to leave there
- IS + Topical LA
- PMC ULD (size 2 E crown chosen), LLE
- FS LR6



Visit 17: 19/02/2020, Review

- No new clinical cavities
- Fluoride varnish
- Final clinical photographs



POST-OPERATIVE PHOTOGRAPHS (26/02/2020)

Figure 15: Anterior View

Figure 16: Maxillary Occlusal View

Figure 17: Mandibular Occlusal View

LONG TERM TREATMENT PLAN AND FUTURE CONSIDERATIONS

Caries:

- Monitor caries as high risk – clinical and radiographic
- Maintain fissure sealants on first permanent molars

Periodontal:

- Monitor habit and recession
- Consider referral to OI team clinical psychologist/ CAMHS

Developmental:

- Monitor space loss from extractions

DISCUSSION AND REFLECTION

J.O. has Type I OI without dentinogenesis imperfecta. I liaised with J.O.'s OI consultant prior to starting treatment and confirmed there were no additional precautions needed for his extraction. Although guidelines for oral health management of patients at risk of medication-related osteonecrosis of the jaw (MRONJ), state that any person on bisphosphonates for over 5 years is classified as high risk (1), no cases of MRONJ have been reported in children with OI. If extractions are required, the recommendations are to proceed as normal without additional antibiotics or antiseptic prophylaxis (1).

Prevention

Prevention is particularly important in a medically compromised child. Prevention advice followed the 'Delivering better oral health: Toolkit for prevention' for children who are a high caries risk (2). Advice was given on preventing and stopping gum picking was also needed. Advice included habit stopping techniques such as putting a sock over his hands, unpleasantly flavoured nail polish and positive reinforcement techniques implemented by his mother when he does not scratch during a pre-determined amount of time.

Behaviour Management

Behaviour management was challenging in this case. Ideally, I would have used techniques such as desensitisation to allow familiarisation with the dental clinic, however due to pain and infection from the LLD, I prioritised the extraction. To help alleviate anxiety during the extraction, I used non-pharmacological behaviour management techniques such as 'Tell-Show-Do'. I found it to be effective in helping alleviate some of J.O.'s anxiety.

IS was used as an adjunct to alleviate anxiety and I noticed that this helped him relax and accept treatment initially (3).

Despite this, after his third visit with IS/LA, J.O. became distressed. Part of treating children is understanding when the child needs a break. The decision not to continue treatment at this appointment ultimately helped J.O. have a more positive outlook towards treatment and built his trust. His mother was also preparing for the birth of her third child under 6, and this may have affected his overall behaviour and ability to cope.

Treatment

For J.O.'s first extraction (LLD), the decision to use articaine was made. According to a recent review, there is no difference in pain perception during administration of anaesthesia with articaine or lignocaine, however, children report less post-operative pain after dental treatment with articaine. Additionally, there is no difference in adverse effects post-operatively between the two types of local anaesthesia (4). Given this was J.O's first experience of treatment I felt that this option would ensure a pain-free first appointment.

J.O.'s posterior cavities were managed with PMCs using the Hall Technique which is quick, simple and painless. The evidence shows that the hall technique has a success rate of 97% after 5 years (5) I could have placed conventional crowns or intra-coronal restoration, but decided that Hall crowns were more suitable in light of his anxiety.

Whilst placing Hall crowns one appointment which did not go according to plan. On his third visit, J.O. became distressed after I attempted to remove a PMC from his LRE which I had not seated properly. On reflection I feel that I probably chose the wrong size crown as it got stuck on the buccal aspect of the LRE. This brought home the importance of correct size selection for hall crowns, as this made a relatively straightforward appointment quite challenging and upset J.O.

At last review all teeth treated with PMCs showed no pathology.

Gingivitis Artefacta

J.O.'s gingival recession progressed over the year. Although the family initially denied any habits, slowly a history of gum picking and rubbing was revealed. Using clinical photography to monitor the recession aided in communication with the family, when advising them to stop the habit. Gingivitis artefacta minor is a form of unconscious self-injurious behaviour involving gum picking with fingernails or sharp objects, often indicating an underlying emotional concern. This habit can prove potentially destructive as it causes gingival recession (6).

I felt it was particularly important to address this problem in the primary dentition, to reduce the risk of irreversible damage to the permanent dentition periosteum.

Reasons for J.O.'s habit could include the birth of his new sister, or stress due to his medical treatment. If the problem persists, referral to OI team psychologist or CAMHS for support may be indicated.

Lessons Learnt

- Importance of recognising possible causes of gingivitis artefecta
- IS allowed treatment to be provided successfully and avoided the need for GA

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Dental Trauma Case

A Clinical Case Report

Submitted in partial fulfilment of the requirements for the Degree of

Doctor of Dentistry (Paediatric Dentistry)

UCL

Submitted by

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Word count: 2000

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SUMMARY

A.J. was referred by his GDP after failure of treatment following dental trauma to the UL1. Repeated reattachment of fragment led to subsequent loss of vitality and the need for root canal treatment. Despite his young age, A.J. coped well with treatment in the chair.

PATIENT DETAILS

Name:	A.J.
Gender:	Male
Age at presentation:	7 years
Age at last review:	8 years, 6 months

PRE-TREATMENT ASSESSMENT

History Of Presenting Patient's Complaint

- C/O: UL1 abscess
- When: September 2018
- Where: Youth club
- How: Slipped playing dodgeball
- Action taken:
    - At GDP:
        - UL1 fragment reattached by emergency dentist on the day
        - Debonded and re-attached three times
        - Two days prior to first attendance, swelling of lip and antibiotics prescribed
- At the Department:
    - Patient presented with a buccal abscess associated with UL1.
- Other signs/symptoms: Nil.

Relevant Medical History

- Nil relevant.
- No known allergies.

Social History

- Lives at home with both parents, sister
- Attends primary school, year 2

Dental History

- Regular attender
- Brushes twice daily, 1450ppmF toothpaste, unsupervised, spits.

CLINICAL EXAMINATION

Extra -Oral Examination

- Normal TMJ
- Lymphadenopathy left side
- Swelling of upper lip and left cheek
- No temperature

Intra -Oral Examination

Soft Tissues

- Gingival swelling around UL1

Oral Hygiene

- Good – plaque score 0.2

Dentition

- Early mixed dentition

6	E	D		C	2	1	1	2	C	D	E	6
6	E	D	C	2	1		1	2	C	D	E	6

- No caries
- UR1 partially erupted
- Fractured UL1

Occlusion

- Class I molar occlusion, anterior teeth not in occlusion

Specific Teeth Examination

Tooth	**UR2**	**UR1**	**UL1**	**UL2**
EPT	+	+	-	-
Ethyl Chloride	+	+	-	-
Colour	normal	normal	Grey	normal
Sinus	no	no	no	no
Tender to percussion (TTP)	no	no	yes	yes
Percussion sound	no	no	no	no
Pain in sulcus	no	no	yes	yes
Mobility	no	no	yes	yes

GENERAL RADIOGRAPHIC EXAMINATION (3/09/2018)

RADIOGRAPHS REQUESTED:

- Long Cone Periapicals (LCPA) of UR2-UR1-UL1-UL2.
- Upper Standard Occlusal (USO), vertical angle to exclude root fractures.

- Justification: Trauma Assessment

Figure 1, 2, 3: LCPA and USO (Grade 1)

Radiographic findings:

- UR1: Immature root with open apex
- UL1: Immature root with open apex; Periapical radiolucency
- No root fractures

PRE-TREATMENT PHOTOGRAPHS

Figure 4, 5: Anterior and extra-oral views (3/09/2018)

Figure 6: Anterior and extra-oral view (19/09/2018)

DIAGNOSTIC SUMMARY

Tooth	UL1
Hard tissues and pulp	Complicated crown fracture Pulp necrosis Apical abscess

AIMS AND OBJECTIVES OF TREATMENT

- Remove source of pain/ infection.
- Establish a preventative regime according to Department of Health toolkit.
- Restore oral health (function and aesthetics), maintaining UL1 in dentition.
- Establish a positive attitude towards dentistry.

TREATMENT PLAN

Emergency (immediate) treatment plan

- Antibiotics prescribed due to non-healing facial swelling of the left-side of lip
- Warned: attend A&E if swelling increased

Intermediate treatment plan

- Root Canal treatment (RCT) of UL1
- Replace restoration and bleaching of UL1
- Establish a preventative regime according to the toolkit.

Long term treatment plan

- UL1:
    - Poor Long-term Prognosis: immature root, risk of fracture
    - If lost then maintain space until adulthood when restorative options will be discussed.

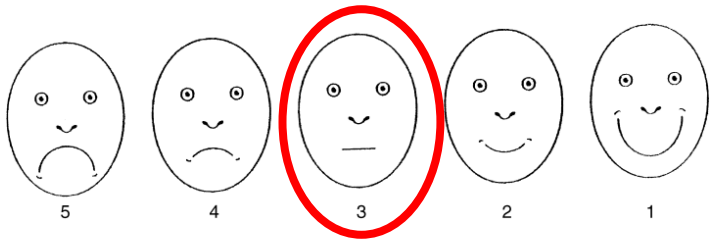
Review the pulpal and periapical status of UR2 -UL2

TREATMENT UNDERTAKEN

- No medical history changes throughout treatment
- Facial index scale to assess anxiety
- Attended appointments with father

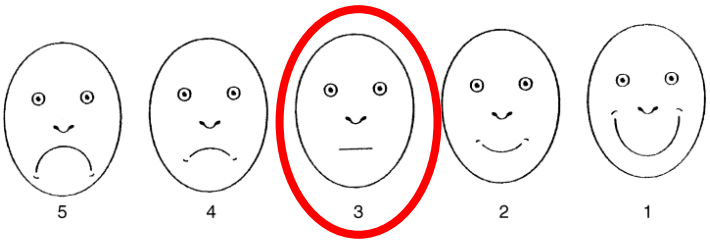
Visit 1: 3/09/2018 New-patient clinic

- C/O: Pain UL1 due to abscess.
- Dental and radiographic examination as previously described
- UL1 – discussed guarded long-term prognosis due to immature root
- Treatment options discussed with parents for UR1:
    1. RCT under Inhalation sedation (IS) and local anaesthesia (LA);
    2. RCT under LA.
- Family opted for option 2.



Visit 2: 19/09/2018

- C/O: Sore UL1
- Topical and LA
- Dry-dam isolation as UL1 not yet fully erupted
- UL1:
    - Access gained
    - Necrotic pulp remnants removed – barbed broach
    - Working length (WL) PA taken at 21mm – short by 2mm, established at 23mm
    - Irrigated with NaOCl 2.5% and agitated with syringe tip to 21mm
    - Canal dried with paper points
    - Packed with thick paste of non-setting calcium hydroxide ($Ca(OH)_2$)
    - Interim IRM access restoration
- Post-operative radiograph – Well filled canal



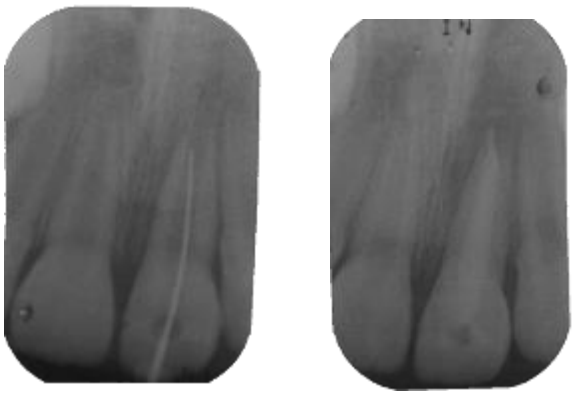
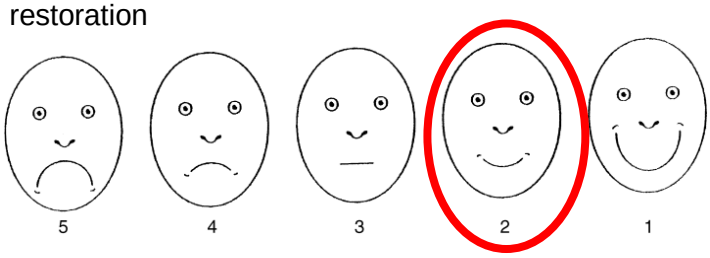


Figure 7, 8: WLPA of UL1 and post-operative PA UL1 (Grade 1)

Visit 3: 17/10/2018 – Patient attended late

- C/O:
    - Fracture of reattached fragment – restored with GIC by GDP
    - Sore UL1
- Dry-dam isolation
- Irrigated with NaOCl 2.5% as before
- Filing using k-files size 40
- Tenderness felt at apical 2mm
- Canal dried with paper points
- Packed with $Ca(OH)_2$
- Interim IRM restoration



Visit 4: 7/11/2018

- C/O: Spontaneous intermittent pain
- Topical and LA
- Dry-dam isolation
- Copious irrigation with NaOCl 2.5% as before
- Gates gladdens bur size 6
- WL PA taken at 21mm (fractured incisal fragment) - Short by 1mm, established at 22mm
- Filing using k-files size 40-60
- Tenderness felt at apical 2mm
- Canal dried with paper points
- Packed with thick paste of $Ca(OH)_2$
- Interim IRM restoration
- Post-operative radiograph - well filled root canal

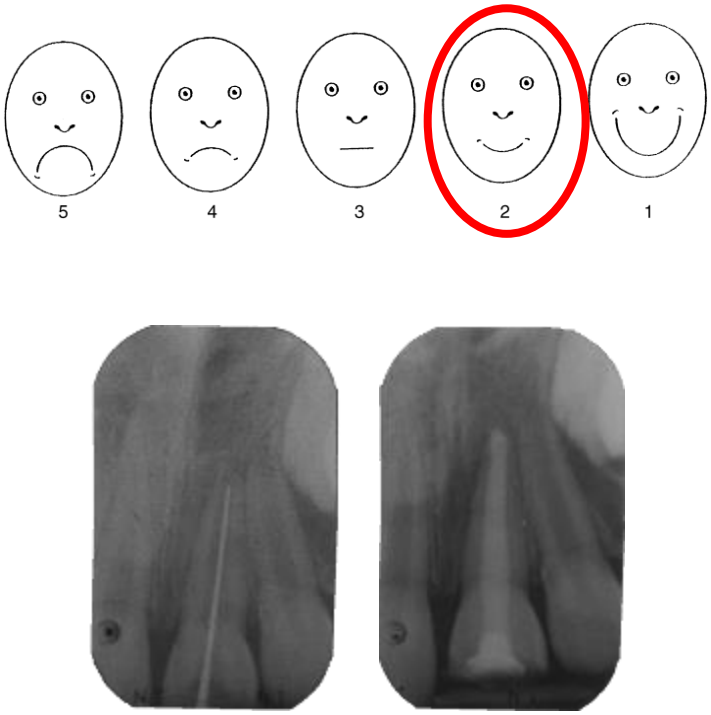
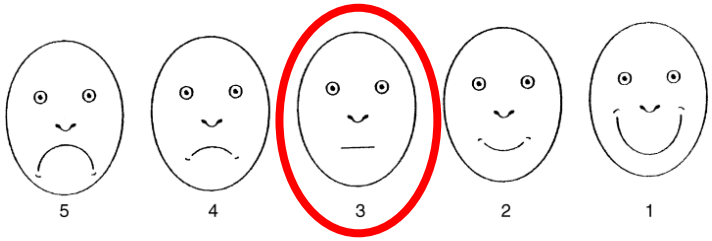


Figure 9, 10: WLPA and post-operative PA UL1 (Grade 1)

Visit 5: 5/12/2018

- Dry-dam isolation
- Access gained and WL confirmed at 21mm with master-cone radiograph – WL confirmed, barrier
- Irrigation using NaOCl
- Canal dried with paper points
- 4mm MTA plug at 21mm
- Check radiograph – Well condensed MTA at 21mm
- Damp cotton pellet and interim IRM restoration – patient began to become tired and fidgety
- Sensibility tests:

Tooth	**UR2**	**UR1**	**UL1(RCT)**	**UL2**
EPT	+	+	n/a	+
Ethyl Chloride	+	+	n/a	+
Colour	Normal	Normal	Grey	Normal
Sinus	No	No	No	No
TTP	No	No	No	No
Percussion sound	No	No	No	No
Pain in sulcus	No	No	No	No
Mobility	No	No	No	No



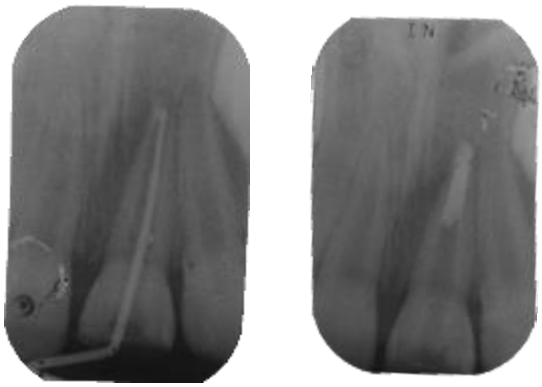


Figure 11, 12: Master-cone GP PA of post-MTA placement PA UL1 (Grade 1)

Visit 6: 09/01/2019

- Patient did not attend.

Visit 7: 13/02/2019

- Dry-dam isolation
- Access gained and cotton pellet removed
- Irrigation with saline and cleaning of walls with tepe brush
- Canal dried with paper points
- Root canal sealer placed on the walls and canals filled to cemento-enamel junction (CEJ) with GP using Calamus®
- Radiograph – Well condensed GP, slightly cervical
- Excess GP removed to 2mm below CEJ, GIC layer placed over GP
- Crown form chosen
- Acid etch, bonding agent, composite Shade A1 using crown form.
- Polished with polishing burs and soflex discs
- Upper alginate impression – construction of bleaching tray
- Pre-bleaching clinical photograph

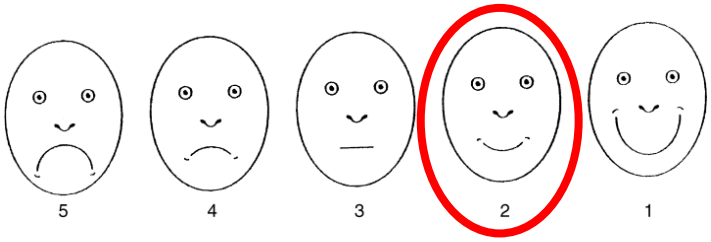


Figure 13: Post-GP PA of UL1 (Grade 2)

Visit 8: 13/03/2019

- Shade of UL1 taken – C2/C3
- Access cavity opened, GP confirmed to below CEJ
- Bleaching tray tried in
- Patient and father shown how to clean cavity and use tray

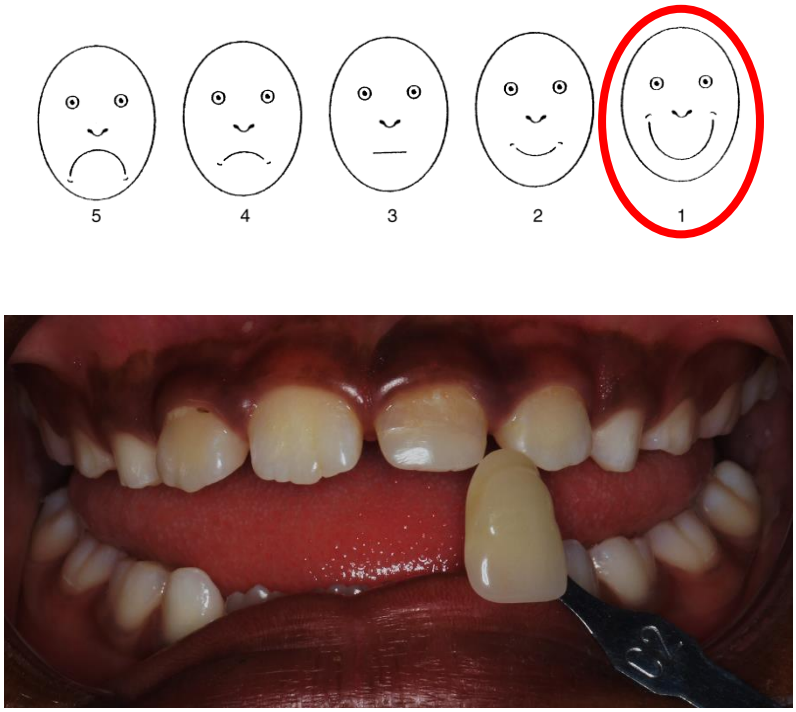
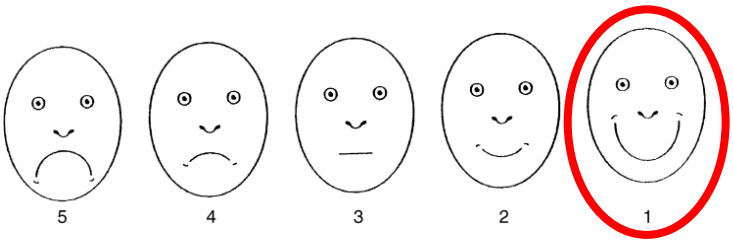


Figure 14: Anterior view photograph pre-bleaching

Visit 9: 26/03/2019

- C/O: temporarily stopped bleaching for 2 days – Gum sensitivity
- Cavity not cleaned daily. Reiterated importance of keeping cavity clean
- Shade of UL1 taken – C1
- Further bleaching required

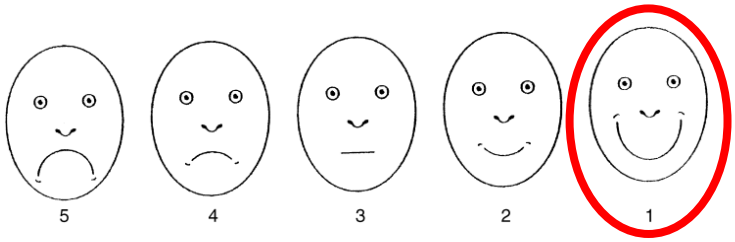


Visit 10: 09/04/2019

- Shade UL1 – A1
- Cavity clean and good OH
- Dry-dam isolation
- Cavity cleaned using water and a tepe brush
- White GP placed in access cavity, restoration lost during cleaning
- Crown form size L1 chosen
- Acid etch, bonding agent, composite Shade A1 using crown form
- Polished with polishing burs and soflex discs
- Patient and father happy with aesthetics
- Sensibility tests

Tooth	**UR2**	**UR1**	**UL1**(RCT)	**UL2**
EPT	+	+	n/a	+
Ethyl Chloride	+	+	n/a	+
Colour	Normal	Normal	Grey	Normal
Sinus	No	No	No	No
TTP	No	No	No	No
Percussion sound	No	No	No	No
Pain in sulcus	No	No	No	No
Mobility	No	No	No	No

- Clinical photographs taken
- Review in six months



Visit 11: 6 month review (28/10/2019)

- Sensibility tests

Tooth	**UR2**	**UR1**	**UL1(RCT)**	**UL2**
Ethyl chloride	+	+	N/a	+
Electric pulp tester	N/a	+	N/a	N/a
Colour	Normal	Normal	Normal	Normal
Sinus	No	No	No	No
TTP	No	No	No	No
Percussion sound	No	No	No	No
Pain in sulcus	No	No	No	No
Mobility	No	No	No	No

- Unable to get correct results for EPT test – faulty EPT
- PA radiographs UR2 to UL2 taken – no evidence Periapical pathology

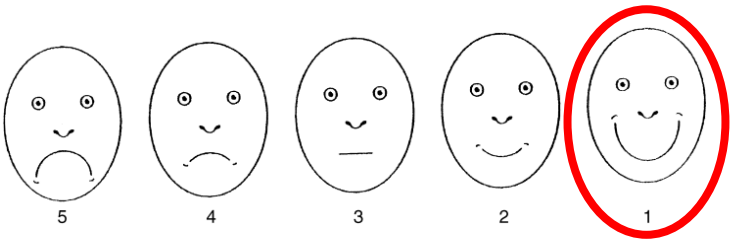


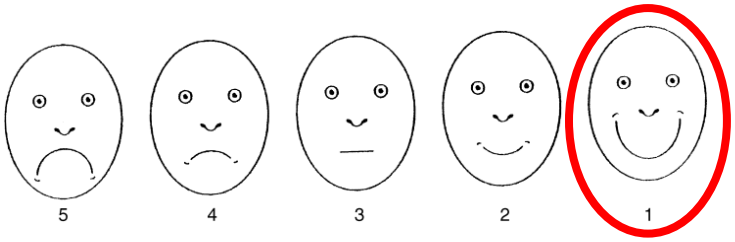
Figure 15: PA of UL1 (Grade 1)

Visit 12: 03/02/2020 – Final Review

- Sensibility tests

Tooth	**UR2**	**UR1**	**UL1(RCT)**	**UL2**
Ethyl chloride	+	+	N/a	+
Electric pulp tester	N/a	+	N/a	N/a
Colour	Normal	Normal	Normal	Normal
Sinus	No	No	No	No
TTP	No	No	No	No
Percussion sound	No	No	No	No
Pain in sulcus	No	No	No	No
Mobility	No	No	No	No

- PA radiographs UR2 to UL2 taken – no periapical pathology



Fiigure 16: PA of UL1 (Grade 1)

POST-TREATMENT PHOTOGRAPHS (09/04/2019)

Figure 17, 18, 19: Anterior view photograph, upper and lower occlusal photographs

LONG TERM TREATMENT PLAN AND FUTURE CONSIDERATIONS

- Monitor UR2-UL2

DISCUSSION AND REFLECTION

Trauma Management

Approximately 25% of school-age children experience dental trauma. In this case, A.J. suffered from a complicated crown fracture of the UL1. Crown fractures are the most common type of dental trauma in the permanent dentition [1].

Enamel-dentine fractures have a low risk of pulp necrosis if managed efficiently. In this case, the dentine tubules were left exposed following repeated debonding of the reattached fragment. This could be the reason the UL1 underwent pulp necrosis so soon after the trauma. Another potential reason is concomitant luxation injury. When extra-oral abscess and cellulitis is present, short courses of antibiotics are advised, as in this case, where the patient was prescribed amoxicillin for three days. I was concerned about non-resolution of the abscess and prescribed an alternative antibiotic, Metronidazole, which targets anaerobic bacteria [2]. Extirpating the root canal at the first appointment would have been ideal, however due to tenderness to percussion, mobility and pain causing limited co-operation, extirpation was postponed until resolution of the abscess.

At the time of trauma, the tooth had an immature root. This complicates root canal therapy for several reasons, including divergent and thin root walls and open apices [3]. The gold standard restoration for the apical portion of roots with open apices is MTA, as it forms a seal at the apex of the tooth, providing an ideal apexification material. It is also a biocompatible material with good physiochemical properties [3].

Prior to endodontic treatment, the UL1 showed grey discolouration. Despite his age, A.J. was bothered by the appearance of the UL1 after treatment, so internal-external bleaching was carried out to improve its aesthetics. Inside-outside bleaching is demonstratively as effective as conventional methods of internal bleaching such as the walking bleach, yet the lower concentration of bleach reduces the risk of external cervical resorption. Inside-outside bleaching can be safely used to treat children [4]. However, according to the ‘EU Cosmetics Regulation’, dental bleaching cannot be carried out on children younger than 18 years old. The General Dental Council, UK, encourages dentists to carry out bleaching in children younger than 18 years old for the purpose of treating dental disease [5].

Behaviour Management

A.J. had only just turned seven when he presented for treatment. As such, complex treatment proved challenging. To help A.J. cope with treatment, behaviour management techniques such as ‘Tell-Show-Do’ and positive reinforcement were used. Appointments were kept as short as possible to ensure A.J. did not tire during treatment. Good behaviour was rewarded with stickers.

The Facial Images Scale used to assess A.J.’s anxiety demonstrates increasing acceptance to treatment as appointments progressed [6].

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