

Prosthetic treatment of a seven-year-old child with suspected Osteogenesis Imperfecta

Sonia Bhat*

*MDS, Senior Lecturer, Department of Prosthodontics, KLE Dental College and Hospital, Bangalore, India. Contact: sonubhat@rediffmail.com

Abstract:

This is a case report of a 7 yr old child patient with suspected osteogenesis imperfecta. Although the clinical findings suggest osteogenesis imperfecta, a conclusive diagnosis could not be arrived at. The patient was treated with removable partial dentures for his missing maxillary and mandibular anterior teeth. In school going children peer pressure is high and poor looks invite ridicule. Such patients are likely to have psychological problems in the long run. Hence it is prudent to give them an aesthetic treatment option which is easy to maintain and does not impede future growth and future dental treatments. Since radiographs confirmed the absence of few of the permanent teeth, patient was put on a regular recall check up programme for timely intervention and correction of intraoral problems likely to occur in future.

Introduction:

As we are advancing into the next century, our diagnostic and treatment tools have evolved at a rapid pace. Over the last decade, with the completion of human genome project, a lot of light has been shed on various aspects of the genetic occurrences.

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Case report

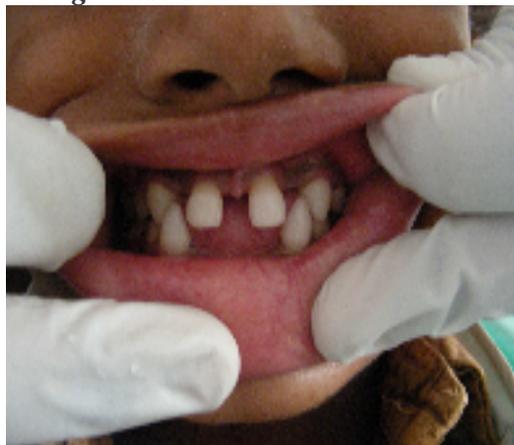


Figure 1 : Intraoral View



Figure 4 : Hand Articulated Secondary Cast

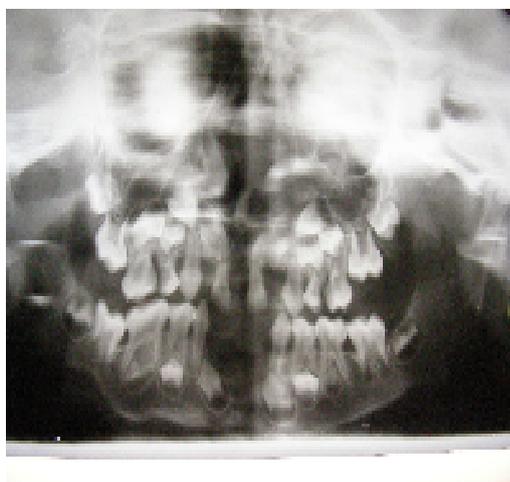


Figure 2 : Panoramic Radiograph



Figure 5 : Post-insertion intraoral view



Figure 3 : Custom Trays



Figure 6 : Post-operative frontal

However it is intriguing to note that, inspite of these advancements, everyday many more diseases surface, which have no treatment or logical explanation. This article is a case report of a 7 yr old child patient for whom, a conclusive diagnosis for his existing condition could not be arrived at but nevertheless dental treatment was rendered.

Case Report:

A 7 year old male child reported with the chief complaint of missing upper and lower deciduous front teeth. Patient had a slurred speech due to the same.

A natal history revealed premature birth at 7 months of intrauterine period. His parent reported delayed attainment of motor milestones. Head control was attained at 10 months of age, he was sitting with support at 1 year and sitting without support at 2 years of age. Standing with support was achieved at 3 years and without support at 3 and half years of age. He started walking only at 5 years of his age. There was history of repeated fractures of upper and lower limbs since he started walking. Dental history revealed delayed eruption of deciduous teeth and absence of permanent lower incisors and upper lateral incisors bilaterally. The Family history was negative for similar complaints including the patient's 10 year old elder sister.

Physical examination revealed the child, had a well proportioned body. He was of an average intelligence. It was observed that patient's height was just 107 cm much

smaller than boys of his age. He had a deformity of right upper limb at the elbow due to fracture at age of 5 year. Extraoral examination revealed an orthognathic profile with competent lips. Intraoral examination showed bilaterally missing upper deciduous lateral incisors, bilaterally missing lower central and lateral incisors (Fig 1). There was partial eruption of all the four permanent 1st molars. Patient had a deep bite and a diastema was noted between the maxillary deciduous central incisors. Although the tongue was normal in size, it was positioned anteriorly with the tip resting on the lower anterior edentulous ridge. Phonetic analysis showed poor pronunciation of "S" and "T" syllable.

Panoramic radiograph of the jaws revealed missing permanent lower central and lateral incisors bilaterally while the maxillary permanent central and lateral incisors bilaterally, appeared to be fused (Fig 2). Rest of the permanent dentition exhibited incomplete root development.

On the basis of the reported history and findings of clinical examination, a provisional diagnosis of Osteogenesis imperfecta was arrived at. Therefore a chromosomal analysis was planned and performed. The patient's chromosomal analysis revealed no abnormality and showed him to be a normal male Karyotype 46XY. After the genetic analysis, the treatment was aimed at addressing the patient's need for improved speech and aesthetics.

Prosthodontic Management

After evaluating all the treatment options (Fixed partial denture, Removable partial denture, Implants) it was decided to fabricate removable partial dentures to replace the missing teeth.

Primary impressions of upper and lower arch were made with stock, metallic perforated size 0 trays using irreversible hydrocolloid. Using these impressions primary casts were poured with dental stone. On the primary cast custom trays with autopolymerizing acrylic and a modeling wax spacer were fabricated (Fig 3). For secondary impressions making, border molding was carried out with low fusing compound and secondary impression were made with Irreversible hydrocolloid. Since the maxillary and mandibular cast articulated well, jaw relation recording was not deemed necessary (Fig 4). For trial denture, autopolymerizing acrylic resin denture base were made and teeth were arranged in modeling wax. It was decided to arrange a single central incisor between the upper deciduous central incisors to close the diastema and improve the speech. Also bilaterally missing upper lateral incisors were replaced. In the lower arch all the four incisors were replaced. At the time of trial of waxed up dentures, aesthetics, speech and occlusion were assessed and adjusted to the patient's satisfaction.

Stainless steel (24 gauge) wire was used to make retentive clasps on the abutments and the trial dentures were cured with heat cure acrylic. The final partial dentures were trimmed and polished. On the day of insertion the fit, occlusion, aesthetics and speech with the partial dentures was

assessed (Fig 5). Both the child patient and his parent were instructed in removal and placement of the partial dentures. He was instructed on hygiene maintenance and asked to avoid biting on any hard object with his artificial anterior teeth. No speech training was done at this appointment. Parents were advised to motivate the child to speak more often and read aloud from his text books at home.

First recall appointment at 1 week that the patient was comfortable with the partial dentures and could easily remove and place the partial dentures from his mouth. The patient was noticed to be smiling more often than the previous appointments (Fig 6). Second recall check up at 3 weeks showed significant improvement in child's speech. His pronunciation of the letters S, T and D had improved and this improvement was also evident to the parents.

Discussion:

Hypodontia is a relatively common occurrence in deciduous dentition. It may or may not be associated with a syndrome¹. In this case patient exhibited symptoms of osteogenesis imperfecta namely missing permanent teeth, fragile bones and delayed motor milestones. According to National institute of arthritis and musculoskeletal disease, USA, more than two hundred heritable disorders of connective tissue have been recognized with Osteogenesis Imperfecta being one of them. It has been classified from Type I to Type IV by Silence et al classification². Each of the four types is further subdivided on the basis of absence or

presence of dentinogenesis imperfecta³. The disorder is characterized by varying degrees of fragile bones, retarded growth, bone deformities, tooth abnormalities, blue sclerae, and hearing loss²⁻⁴. The incidence of dentinogenesis imperfecta has been reported to be greater than 80% in the primary dentition. The above described case is more likely to be a Type I Osteogenesis Imperfecta, wherein normal collagen is produced by the body but in lesser amounts, making the bone fragile. The initial fractures in these patients are noted when they start walking or standing which was also true in our case.

Dental treatment encompasses replacement of missing teeth and correction of malformed teeth. The dentist has the option of treating the patient with adhesive composites to correct the malformed teeth. However when dentinogenesis imperfecta exists adhesive treatment prognosis is questionable although it has met with some success^{3,5}. Fixed prosthodontic treatment should be initiated with caution in these cases, given the nature of dentin being brittle. Removable partial dentures can be used more successfully in these cases as this treatment option is the least invasive of the lot. A removable prosthesis not only allows addition of artificial teeth if more natural teeth are lost in future but also, causes no hindrance to the eruption of permanent teeth beneath. While fabricating partial dentures, development of proper speech can be ensured by giving correct form, dimension and position of the various parts of the prosthesis in their relation to the tongue cheek and lips⁶. Also the clinician must consider not only

space maintenance and masticatory function but also aim at restoring the function of speech while fabricating such a removable appliance for children⁷.

In school-going children, peer pressure is a dominant factor in social interactions. A poor dentition invariably leads to ridicule and isolation of the child and such a child grows to be poorly adjusted personality in the society⁹⁻¹⁰. For these reasons the significance of appropriate dental treatment instituted at the opportune time should be never underestimated. Also with regular recall the dentist may be able to predict any future problems and initiate suitable preventive programmes as may be deemed necessary. However among this plethora of choices for the child, his/her parental involvement is paramount in the overall management of such cases. In conclusion, it can be stated that timely intervention is the key aspect of treating any child patient. With regular recall check up and treatment several problems could be prevented or corrected at an earlier stage so that, from a functional perspective, speech and masticatory ability are not affected and from psychological point of view such a child will adjust well to the society.

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