Prosthodontic Rehabilitation of a patient with Hutchinson Gilford Progeria Syndrome: a rare case report

Running Title: *Jain et al: Hutchinson Gilford progeria syndrome*

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Abstract:

Progeria is a rare genetic disease where symptoms resembling aspects of aging are manifested at an early age. Characteristic clinical findings of Progeria include abnormalities of the skin and hair in conjunction with characteristic facial features and skeletal abnormalities. The findings that are nearly interested in dentistry are delayed dentition, anodontia, hypodontia, or crowding of teeth. Psychological trauma is common in such patients because of the societal alienation they often face; hence, the successful completion of any treatment modality in such patients is affected by how well the Prosthodontist handles their psychological condition. This report follows the Prosthetic rehabilitation of a patient suffering from Hutchinson Gilford progeria syndrome with congenital anodontia , keeping special attention on psychological behaviour. Rehabilitation of such a compromised complete denture patient in a conventional manner possess many challenges. To overcome these challenges certain modifications in treatment procedure and materials used were made to fulfil the patient's functional, esthetic and psychological demands.

Keywords:

Anodontia, McCord, Progeria, Psychological.

INTRODUCTION

Hutchinson Gilford Progeria syndrome, first described by Hutchinson [1] and Gilford [2] is a combination of dwarfism, immaturity and pseudo senility.[3] Hutchinson–Gilford progeria syndrome is a rare, sporadic, autosomal dominant syndrome that involves premature aging, generally leading to death at early age due to myocardial infarction or stroke. The body of a child who has progeria actually ages 8 to 10 years for every year if he/she is alive. Ironically, children with progeria have an above average intelligence. It has a reported incidence of about one in four to eight million newborns with a total reported incidence of just over 100 in the century since its discovery. HGPs is not usually passed down in the families.[4] According to Polex and Hegele, since 1886 fewer than 100 cases of Hutchinson-Gilford progeria syndrome have been reported, with approximately 40 cases currently diagnosed (Polex & Hegele, 2004).[5]

The genetic basis of most cases of this syndrome is a change from glycine GGC to glycine GGT in codon 608 of the lamin A (*LMNA*) gene, which activates a cryptic splice donor site to produce abnormal lamin A; this disrupts the nuclear membrane and alters transcription. [6]

Progeria is a rare genetic disease where symptoms resembling aspects of aging are manifested at an early age. Characteristic clinical findings of Progeria disease include abnormalities of the skin and hair in conjunction with characteristic facial features and skeletal abnormalities. The characteristic facies show protruding ears, beaked nose, thin lips with centrofacial cyanosis, prominent eyes, frontal and parietal bossing with pseudohydrocephaly, midface hypoplasia with micrognathia and large anterior fontanel. The other reported anomalies are dystrophic nails, hypertrophic scars and hypoplastic nipples. The findings that are nearly interested in dentistry are delayed dentition, anodontia, hypodontia, or crowding of teeth.[7,8] Dental treatment in such patients overcomes the patient's functional, psychological, aesthetic and phonation problems. The purpose of this case report is to present a case of 18 year old male patient with Hutchinson Gilford progeria syndrome with anodontia in which Prosthodontic rehabilitation is done with complete denture.

CASE REPORT

A 18 year old male who was 143 cm tall and 44kg, reported in Department of Prosthodontics, Government college of Dentistry, Indore with chief complaint of difficulty in mastication due to total loss of teeth in both maxillary and mandibular arch. Family history revealed that he has a brother who has normal developmental growth. The family's medical history contained no report of similar case in the family. In the patient's dental history, he had congenital absence of teeth.

General examination-

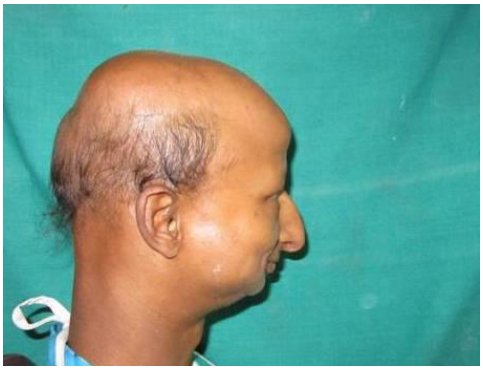
On general examination patient looked older than his age. He was moderately nourished and had a short stature. The skin was thin and atrophic with sparse subcutaneous fat.

Extraoral examination revealed convex profile, prominent forehead and frontal bossing. Scalp hair and eyebrows were lost and veins were prominent especially over the scalp. The nose was thin and beaked, causing a bird like face and senile appearance. Hypoplasia of facial bones, maxilla and mandible was present. There was a markedly retarded vertical and posterior growth of facial bones (fig.1&2). Mentally and emotionally, he was found to be normal.

Visual acuity-congenital microopthalmia, total corneal opacities and entropion were found in left eye.

Fig.1: Pre-operative photograph of the patient

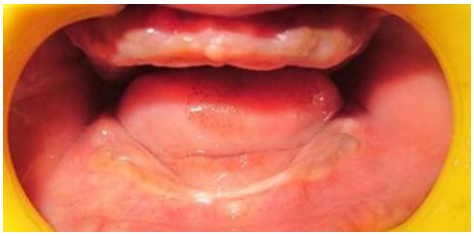
Fig.2: Profile view



Intraoral examination

Narrow, high palatal vault with complete anodontia and loss of vertical dimension was observed . Atrophy of alveolar process with micrognathia and retrognathia was present. There was relatively large tongue. Mucosa appeared normal on inspection and was confirmed by palpation. Class II Skeletal pattern was present with hypersalivation.(fig. 3)

Fig.3: Intraoral view showing resorbed mandibular ridge



Radiological Examination

OPG revealed poorly developed body and ramus of mandible, absence of dentition with loss of alveolar bone.(fig.4)

Lateral cephalogram showed markedly retarded vertical growth of jaws, short mandibular ramus length and obtuse mandibular angle.(fig.5)

Fig.4: Orthopantomograph of patient showing poorly developed mandible

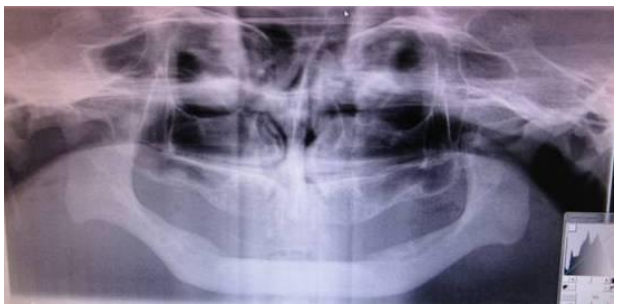
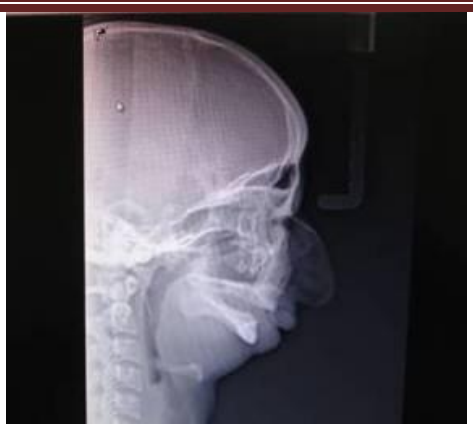


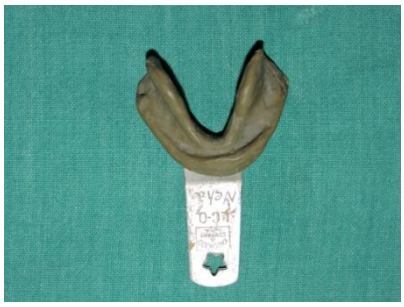
Fig.5: lateral cephalogram of the patient



Prosthodontic Management

A conventional complete denture was planned for the patient based on clinical and radiographic findings. Though the maxillary residual ridge growth was normal, there was increased bone loss in mandible. Poorly developed residual ridge in mandible can lead to inadequate retention and stability of complete denture. Therefore, care was taken at every step for providing adequate retention and stability to the complete denture. Preliminary impression of the edentulous maxilla was made with impression compound. Preliminary impressions of the edentulous mandible was made with a viscous mixture of two varieties of softened impression compound (3 parts impression compound + 7 parts greenstick compound) [McCord's Technique] providing optimum coverage of denture bearing area(fig.6).

Fig.6: Primary impression made using McCord technique



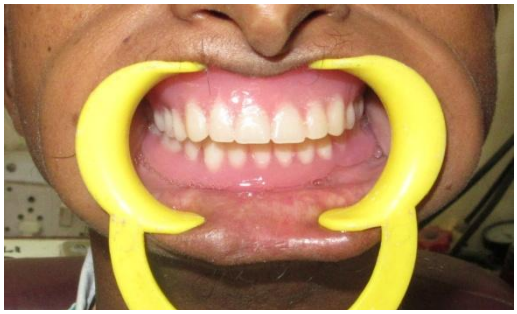
The impressions were washed and poured with the dental plaster. The custom tray was prepared with auto-polymerising acrylic resin (DPI –RR cold cure, Dental Products of India, the Bombay Burmah Trading Corporation limited) for making secondary impressions. The secondary impressions were made using green stick compound for border moulding (DPI Pinnacle Tracing Sticks,Bombay, India) and zinc oxide eugenol impression paste(DPI Impression Paste, Dental

Products of India) as impression material utilizing selective pressure impression technique .Master casts were made with Dental stone type III (Kalastone, Kalabhai Dental Private limited). Hanau spring Face bow was used to orient the patient's maxilla to terminal hinge axis of the mandible. Indirect mounting was done to mount the maxillary cast on semiadjustable articulator (Hanau Wide-Vue Arcon 183-2, Whip Mix, Louisville, Ky). After obtaining Protrusive interocclusal records, condylar guidance of 35º were set on Hanau wide vue articulator. Bennet angle was calculated using Hanau formula:

L= H/8+ 12

Patient had severe class II jaw relation because of poorly developed mandible and normal maxillary ridge development. A maximum effort was made to get an occlusion which was as close to balanced occlusion .20º semianatomic teeth were arranged on maxillary and mandibular occlusal rims with bilateral balanced occlusion for providing stability to the complete denture.There are different schools of thoughts regarding the arrangement of teeth in class II situation i.e. placement of extra canine in maxillary arch or exclusion of mandibular premolar from mandibular arch. In the present case, we preferred to exclude mandibular premolar as placement of extra canine in short maxillary arch will extend the maxillary teeth over maxillary tuberosity region.After evaluating the waxed up trial denture for function,esthetics and comfort in the try-in procedure,the denture was acrylized with heat polymerizing acrylic resin (Trevalon, Dentsply, Gurgaon ,India) . The interferences in the denture were eliminated and denture was given to the patient.(fig.7&8) Post insertion instructions were given to the patient regarding its maintenance, nutrition and hygiene. Patient was examined 48 hours later and then followed up on a routine basis. Patient had no notable complaints about the prosthesis. He stated that he was able to speak and function better. His facial appearance was improved. The patient has been using his prosthesis for 6 months and has not presented any complaints.

Fig.7 & 8: Intraoral and extraoral view after prosthetic rehabilitation



DISCUSSION

Hutchinson Gilford Progeria syndrome is a very rare genetic disorder where symptoms of aging are manifested at a very early age. The incidence of progeria has been estimated at about 1 in 8 million births. Boys outnumber girls in a 1.5:1 ratio, and there is a marked non-negroid racial prevalence amounting to 97% of cases. Diagnosis of Progeria rests on clinical presentation. Although there are very few cases of progeria have been reported in literature, several common features seem to be present in reported cases. Progeria mimics the aging process in many cases, however, some systems appear to be immature or dysplastic. The developments of primary and permanent dentitions are markedly delayed in these patients. Permanent dentition is often characterised by hypodontia or anodontia. This was a consistent finding in every case reviewed from a dental perspective. In this case report, the child presented with a congenital anodontia of teeth. Due to the abscence of teeth the alveolar bone growth was severely affected,especially in mandible.There was difficulty in mastication and facial aesthetics were poor. There was alveolar bone loss which poses problem in retention and stability of the complete denture. Rehabilitating is always a challenge for the prosthodontist in these cases . As the residual ridge resorbs , the tissues become unsupported and displaceable.The use of conventional impression technique may result in distorted impression. Therefore, McCord impression technique has been used in this patient. Also, decreased vertical height & reduced interridge distance cause difficulty in ideal teeth arrangement. Patient was given option for the fixed prosthesis, implant supported but the patient didn't agree for any surgical intervention. So, a complete denture was made in a conventional manner keeping an eye on the psychological behaviour. To gain the trust of the patient, each step of the treatment was explained in detail. Maintaining a pleasant conversational tone helped the patient to accept the treatment. By recording a detailed case history and taking care of all the steps in fabrication of complete denture, patient was successfully rehabilitated proshodontically. The patient was followed up for 6 months and the patient was satisfied with his esthetics and with his mastication.

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