Prosthetic Management of chirst- Siemens-Touraine Syndrome - A case report

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Abstract

Ectodermal dysplasia is a rare hereditary disorder. It appear to be inherited as a X-linked recessive trait, with the most common condition among the ectodermal dysplasia's being hypohidrotic (anhidrotic) ectodermal dysplasia also known as Chirst - Siemens - Touraine syndrome . Such patients are characterized by the clinical manifestations of Hypodontia, Hypotrichosis, Hypohidrosis and a highly characteristics facial physiognomy.

This article reports about the Prosthodontic management of a typical case of chirst - Siemens - Touraine syndrome.

Key words - Ectodermal dysplasia, Hypohidrotic ectodermal dysplasia,chirst - Siemens - Touraine syndrome.

Introduction

The National Foundation of Ectodermal Dysplasia (NFED) defines Ectodermal dysplasia (ED) as a genetic disorder in which there are congenital birth defects (abnormalities) of two or more ectodermal structures . These structures include skin, hair, nail,teeth, nerve cells, sweat glands,parts of eye and parts of other organ.1 Chirst - Siemens - Touraine syndrome is mainly inherited through a X - linked recessive trait considered to be a traid of Hypohidrosis, Hypotrichosis, Hypodontia and characteristic facies2.Hypohidrosis/ anhidrosis means,complete or partial absence of sweat glands making skin smooth, thin, dry with reduced or absence of sweating and often presents with episodes of hyperthermia or unexplained fever. Hypotrichosis is characterized by sparse and short hair of scalp,eyebrow, and eyelashes and represent lanugo.The mustaches and beard are usually normal in appearance.3The most common oral characteristic is hypodontia or anodontia, reflecting the complete suppression of the dental ectoderm.A few teeth may be present but with retarded eruption.because of the lack of teeth and resultant loss of vertical dimension, the lips are protuberant, the vermilion border is indistinct, and the alveolar process does not develop in the absence of the teeth and hence is missing.4,5Incisors, canines and premolars, when present, often have conical crowns and the oral mucosa often appears dry.

Case report

A 20 year old male patient reported to the department of Prosthodontics with the chief complaint of missing teeth and wanted to replace them since he had difficulty in eating food.

On extra oral examination revealed frontal bossing,depressed nasal bridge and protuberant lips due to absence and mal developed teeth.

(Fig;1) .The patient also presented with sparse

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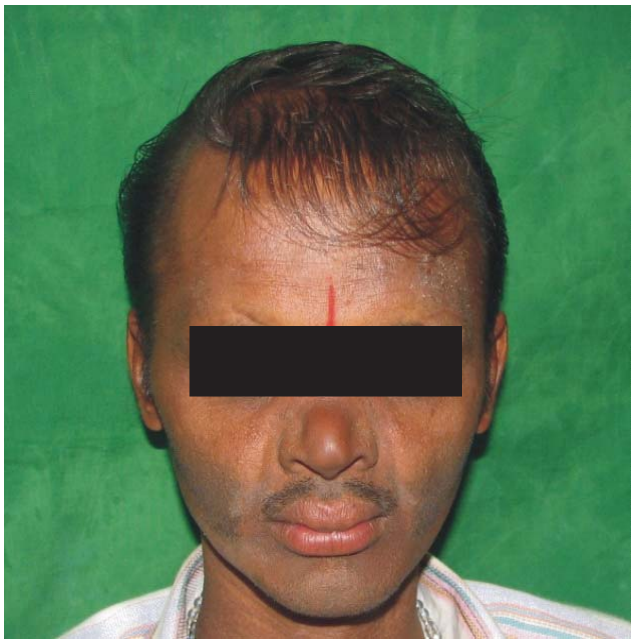
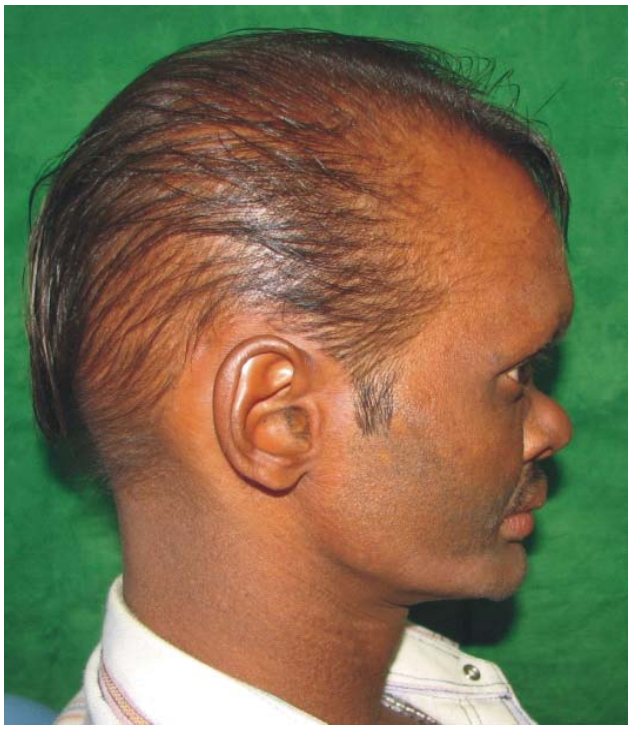


Fig 1) Extra oral view (Frontal)



The skin appeared scaly, dry, thin with hyperpigmentation around the eye and the mouth (Fig 2)

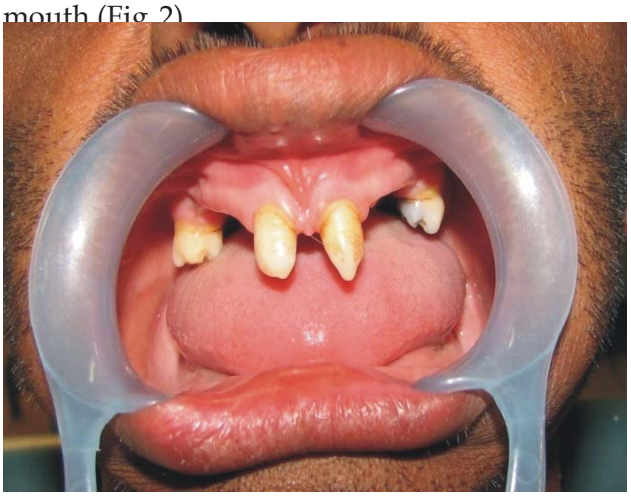


Fig 3) Intra oral view showing maxillary teeth

There was no positive family history.His parents did not exhibit evidence of ectodermal dysplasia and none of his relatives were known to have a condition similar to the patient.

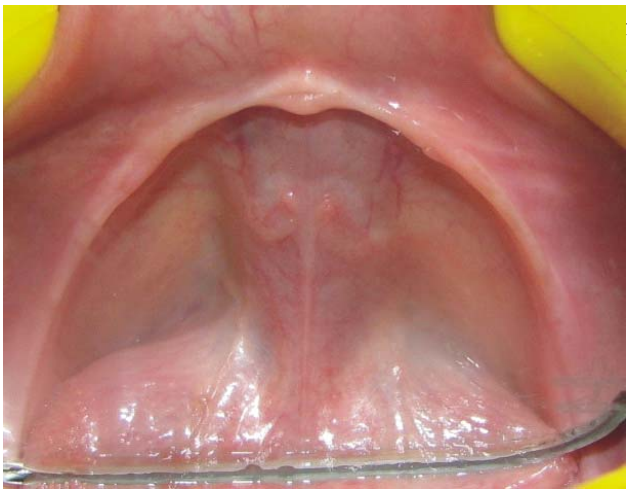
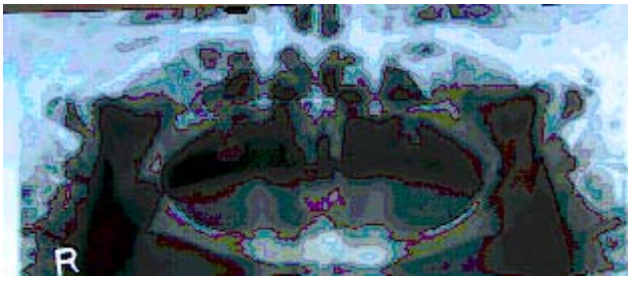


Fig 3a) Intra oral view showing mandibular knife Edge ridge .

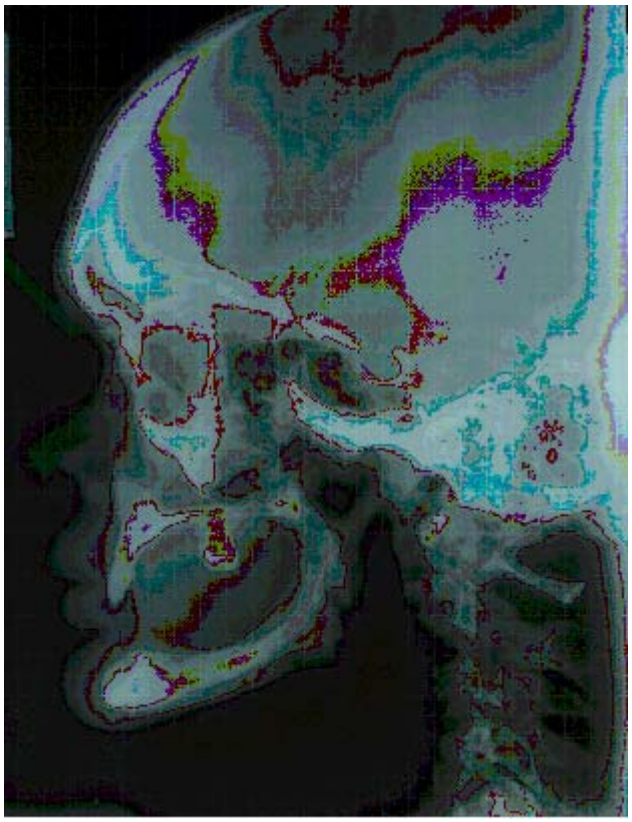
The canines were conical shape.The palate was shallow and the oral mucosa was healthy with slight dry appearance.The lower jaw was

edentulous with thin and knife edge ridge.(Fig3a). The tongue was relatively large, but no signs of macroglossia could be dectected. The flow of saliva appeared to be reduced

Panoramic radiograph



Lateral cephalogram



The vertical dimension of occlusion (VDO) was reduced as evidenced by deep folds in the commissures of the mouth and by the patients thin lips.

By considering the presence of triad of hypohidrosis,hypotrichosis and hypodontia, diagnosis of Christ -Siemens-Touraine syndrome was made.

Treatment plan was explained and informed consent was obtained by the patient.

Materials and Methods

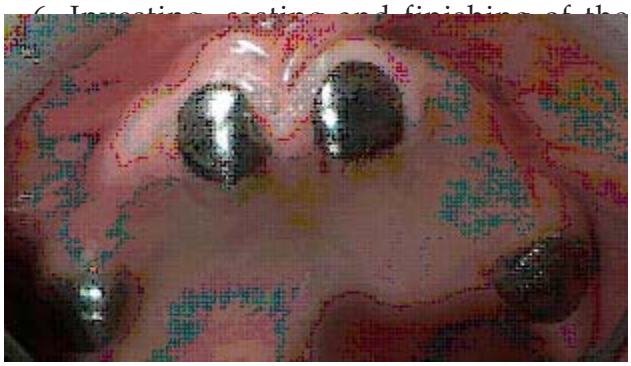
1.Primary impression of upper and lower arch was made with irreversible hydrocolloid alginate (Zelgan, Dentsply India) impression material to obtain a diagnostic cast .

2.A tentative jaw relation was recorded. The casts was mounted on the semi-adjustable articulator which allows for a dynamic evaluation of the interarch relations which was reduced.

3.All the abutment teeth ie (2 permanent maxillary canines and 2 permanent 2nd maxillary molar) were endodontically treated

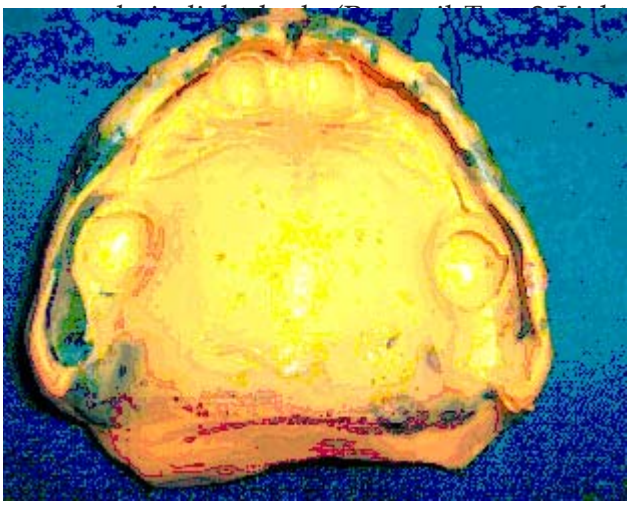
4. The individual teeth was prepared to receive medium metal copings. The coronal length was reduced to about 3mm.Maximum reduction of tooth structure was accomplished on the facial surface of the anterior tooth (canines).

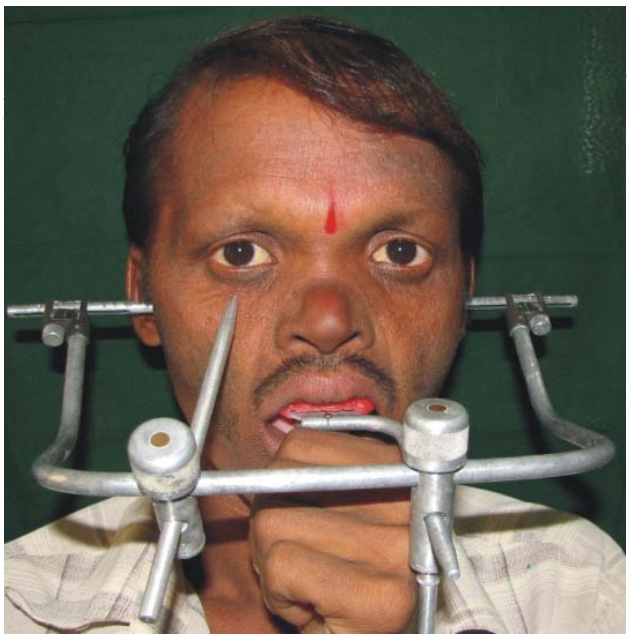
5.Rounded contour was provided with no undercuts and a chamfer finish line was given at the gingival margin.The occlusal or incisal surface was convex or dome shaped.



7. Special tray was fabricated on the primary

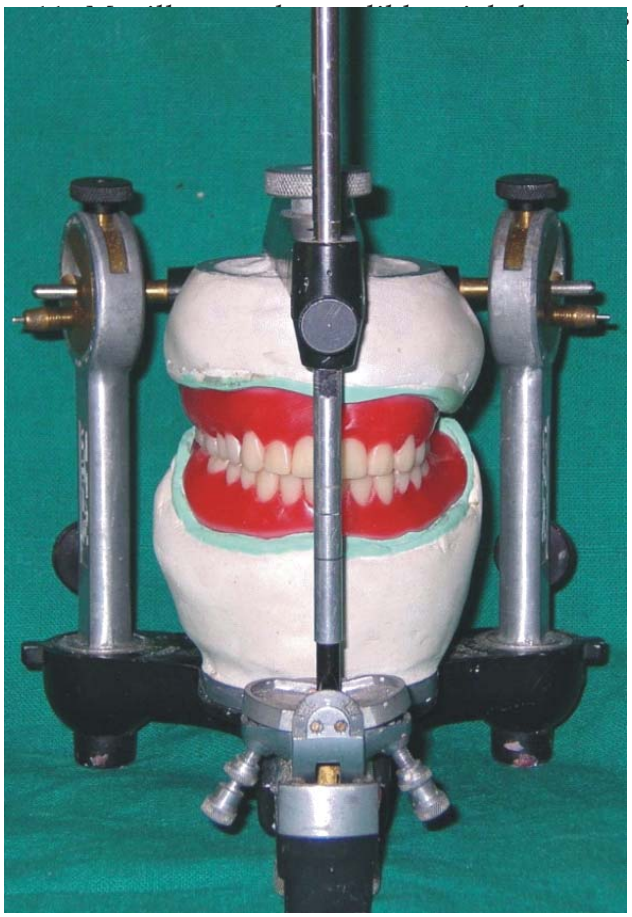
cast for maxillary and mandibular arch, border moulding was done with low fusing impression compound(Aslate India) and finial impression





9.Jaw Relation was recorded.

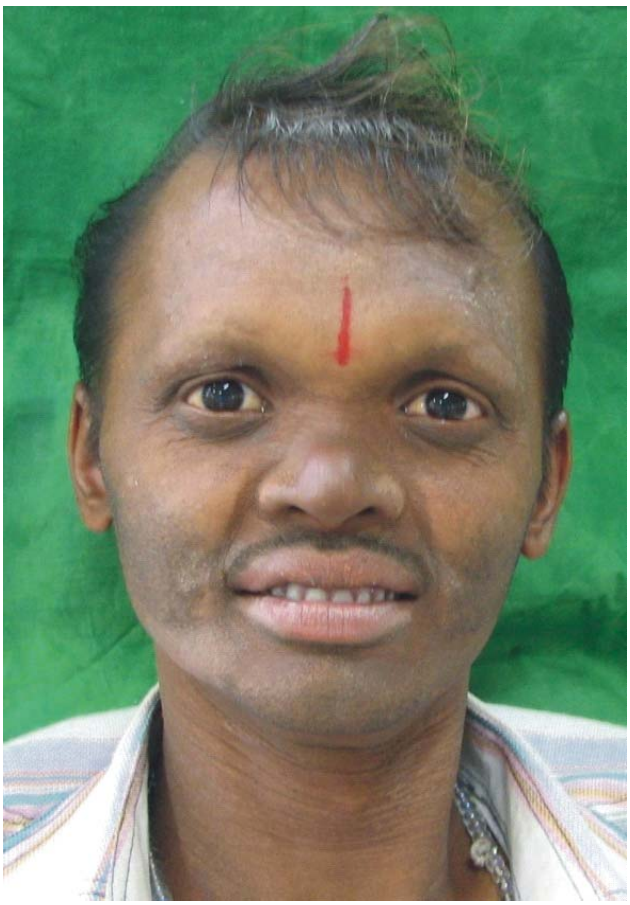
10..Non-anatomic teeth were selected because the mandible was underdeveloped and the alveolar ridge was thin and knife edge ,hence to prevent the occlusal load on the mandible which could lead to further resorption of the bone in mandible.



12. After the approval of the tooth arrangement by the patient and his parents, the waxed dentures were processed in a High impact heat-polymerized denture base resin (Lucitone 199,Dentsply).

13. Fit of the completed denture was verified.

The complete denture was delivered to the patient (Fig 10)



Instruction to the patient.

The patient was instructed on the proper maintenance of the oral tissues,the abutment teeth and the prosthesis.

Patient follow-up

The patient was put on a 24-hour follow-up schedule for any adjustments. The patient was then scheduled for 1month,3 month and 6 month follow up-appointments.

Discussion

Treatment for young patient with anhidrotic ectodermal dysplasia require the team approach. For the patient in this clinical report ,the 4 permanent teeth were endodontically treated to provide an interocclusal distance more favourable for the placement of the artificial teeth in an esthetically acceptable position .An understanding of the ectodermal dyslasia patients psychosocial status is crucial to any prosthodontic treatment effort. The unesthetic appearance that accompanies Ectodermal Dysplasia syndrome often has a negative psychological effect on the patient.7

Complete denture prosthesis can provide acceptable results for esthetics, psychologic support and function, however, underdevelopment of the alveolar ridges in the patient with hypohidrotic ectodermal dysplasia makes denture retention and stability difficult to achieve.Endosseous implants can also be considered as an alternative treatment.8,9,10

Conclusion

This clinical report describes the characteristics and restoration with the overlay complete denture in a young male patient with Christ - Siemens - Touraine syndrome .The overlay complete denture improved denture retention, stability and support and preserves the alveolar bone. For the patient described, the treatment improved esthetics and oral function and established a more favorable occlusion. The patient's social confidence also improved significantly as a result of the dental treatment.

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