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MULTI-DISCIPLINARY APPROACH IN THE MANAGEMENT OF CHRIST-SIEMENS-TOURAINE SYNDROME WITH ANODONTIA: A CASE REPORT OF TWO SIBLINGS

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Abstract:
Aim: The purpose of the case report was to highlight the clinical features and multidisciplinary approach to managing children suffering from Christ-Siemens-Touraine syndrome with complete anodontia and its prosthetic rehabilitation.

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Background: Christ-Siemens-Touraine syndrome, which is a recessive autosomal disorder, is the most common form of Heredity Ectodermal dysplasia in which two or more ectodermally derived anatomic structures fail to develop, thus leading to hypoplasia or aplasia of structures such as skin, hair, nails, teeth, nerve cells, sweat glands, parts of the eye and ear and other organs. **Case report**: A 7-year-old boy and his 6-year-old younger sibling were referred to the Department of Pediatric & Preventive Dentistry with the chief complaint of missing teeth in the whole mouth and difficulty in speaking and chewing. After thorough examination and evaluation, in order to improve appearance, mastication, and speech, removable complete maxillary and mandibular dentures were planned to be the best treatment choice.

Keywords: Christ-Siemens-Touraine syndrome, anodontia, Hereditary Ectodermal Dysplasia (HED), multidisciplinary approach

1. Introduction

Hereditary Ectodermal Dysplasia (HED) represents a heterogeneous group of hereditary diseases in which two or more ectodermally derived anatomic structures fail to develop. Hypoplasia or aplasia of structures such as skin, hair, nails, teeth, nerve cells, sweat glands, parts of the eye and ear, and other organs are the characteristic features of such patients. [1]

There are 154 patterns of ectodermal dysplasias, further divided into 11 subgroups and then classified according to the involved structures as described by Freire-Maia-Pinheiro. [2] The most common form of ED is the Christ-Siemens-Touraine syndrome, which is a recessive autosomal disorder. On the basis of severity, it can be further classified as Hypohydrotic Ectodermal Dysplasia (HED) or as Anhidrotic Ectodermal Dysplasia (AED). The three most important clinical signs are Hypotrichosis (lack of hair), Hypdontia (Absence or reduction in number of teeth) and Hypohydrosis (absence or diminution of sweat glands). [3] Systemic manifestations include skin changes, recurrent ocular infections, chronic rhinitis, dystrophic nails, epistaxis, dysphagia, dysphonia, alopecia, and atypical facies.[4] Dental findings include complete or partial anodontia of the primary or permanent dentition, peg-shaped incisors or canines and taurodontism. [5]. Fine wrinkling over pre-ocular skin, hyperpigmentation, midface hypoplasia and protuberant lips are frequently observed. Prominent lips, the disappearance of vermilion border and dry mucosa result in a characteristic old-age appearance. [6] For dental management of such patients, age, pattern of dysplasia, and alveolar ridge morphology play a significant role. Also, the oral rehabilitation of such cases requires a multi-disciplinary approach involving pediatric dentistry, orthodontics, prosthodontics, and oral and maxillofacial surgery. [7] Thus, the aim of this case report is to highlight the multidisciplinary approach to managing children suffering from Christ-Siemens-Touraine syndrome with complete anodontia and its prosthetic rehabilitation.

2. Case-Report

A 7-year-old boy and his 6-year-old younger sibling were referred to the Department of Pediatric & Preventive Dentistry with the chief complaint of missing teeth in the whole mouth and difficulty in speaking and chewing.

2.1 Family History

It was revealed that his maternal uncle has the same condition while his mother has peg laterals.

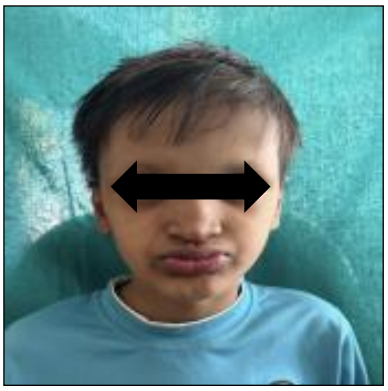

Figure A1: Frontal View

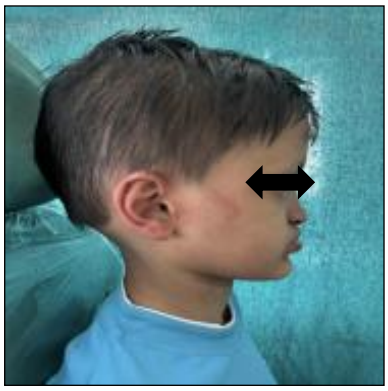

Figure A2: Lateral View

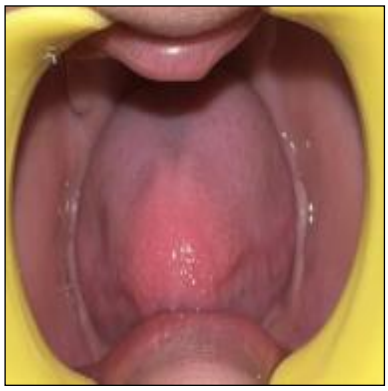

Figure A3: Intra oral

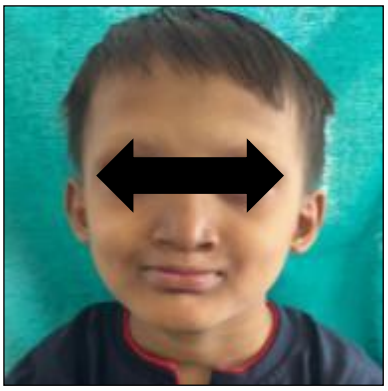

Figure B1: Frontal View

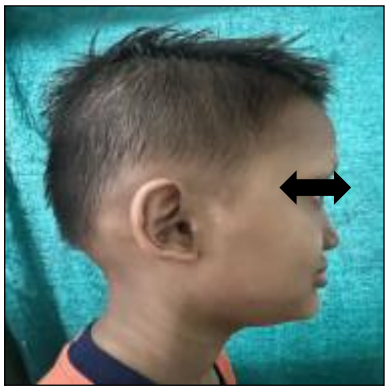

Figure B2: Lateral View

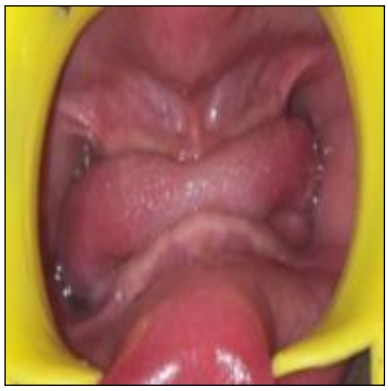

Figure B3: Intra oral

2.2 Intra-oral examination

Both the children showed complete absence of both primary and permanent dentitions. Complete anodontia was confirmed by taking OPGs as shown below. They showed thin alveolar ridge, reduced vertical bone heights and loss of sulcular depths too.

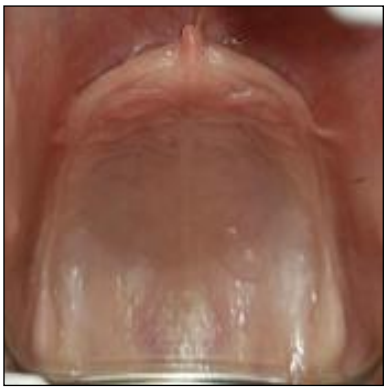

Figure A4: Maxillary Occlusal View

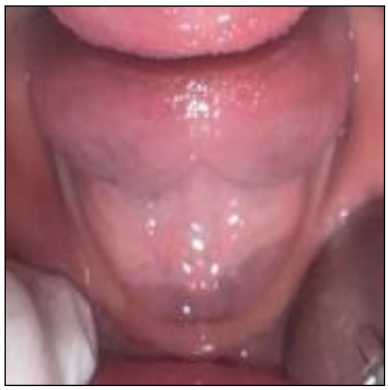

Figure A5: Mandibular Occlusal View

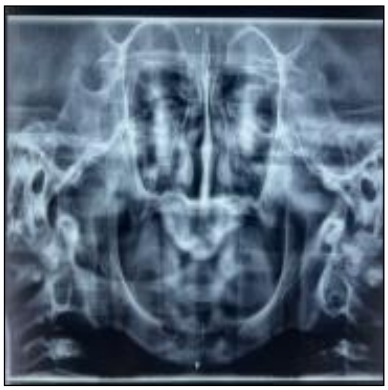

Figure A6: OPG Showing Anodontia

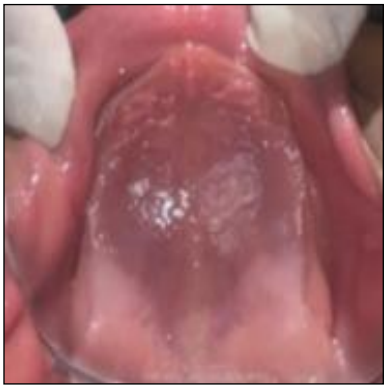

Figure B4: Maxillary Occlusal View

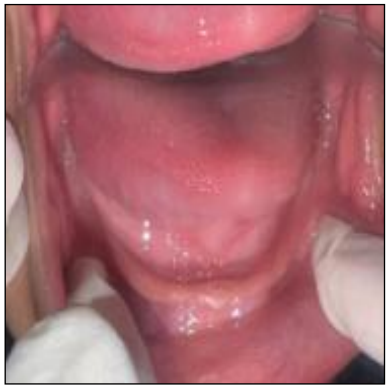

Figure B5: Mandibular Occlusal View

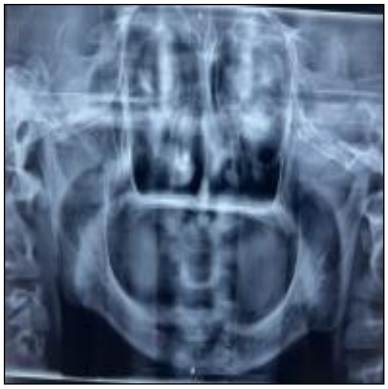

Figure B6: OPG Showing Anodontia

2.3 Extra-Oral Examination

Both the children exhibited the typical features of anhidrotic ectodermal dysplasia: saddle nose, soft, dry and light-coloured skin, increased pigmentation, as well as thin, linear wrinkles in the perioral region as shown in the above figures too. They do not sweat due to the absence of sweat glands, which makes them heat intolerant.

2.4 Management

Both patients were made familiar with dental treatment through counselling, modelling, and tell-show-do techniques in the pedodontics department. After thorough examination and evaluation, in order to improve appearance, mastication, and speech, removable complete maxillary and mandibular dentures were planned to be the best treatment choice. Construction of complete dentures was started following each and every routine step. Preliminary impressions were made using additional silicone impression material, followed by the fabrication of custom trays. This was followed by a secondary impression in the conventional manner.

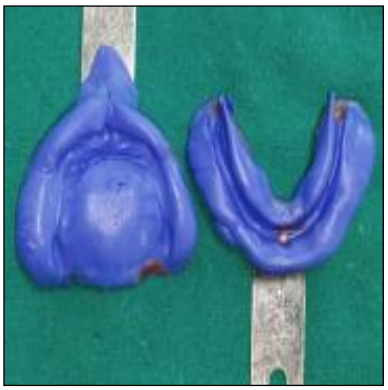

Figure A7: Primary Impression

Figure A8: Border Moulding

Figure A9: Secondary Impression

Figure B7: Primary Impression

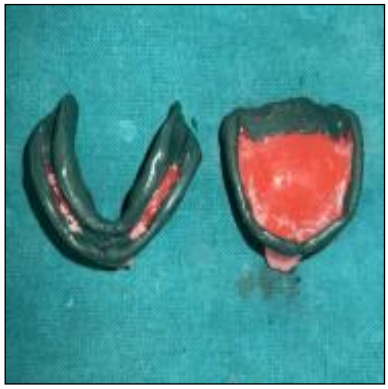

Figure B8: Border Moulding

Figure B9: Secondary Impression

After making the maxillo-mandibular records, the casts were mounted in an articulator. Bite registration was done, and teeth setting was done on the articulator. Primary tooth forms were selected for both 6 and 7-year-old patients in order to provide better static and dynamic occlusion. After the final insertion, routine instructions to maintain the prosthesis were given to children and parents. The patients were advised to maintain a soft diet for the first few days and to remove the dentures at night to promote the healing of the oral tissue. Emphasis was given to maintaining dentures and soft tissue hygiene. Despite the initial lack of compliance, children tolerated the dentures quite well. In order to accommodate growth and development, patients were scheduled for ongoing follow-up visits every 3 months for relining of the dentures.

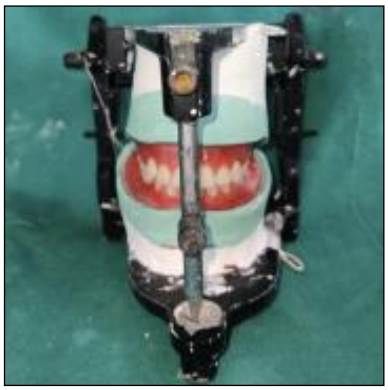

Figure A10: Jaw Relation

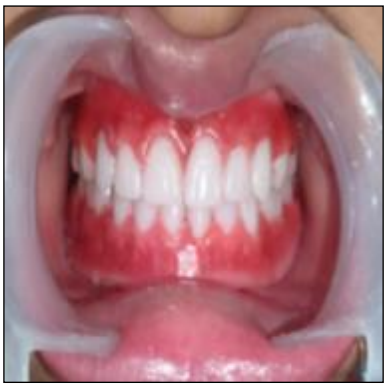

Figure A11: Try-in

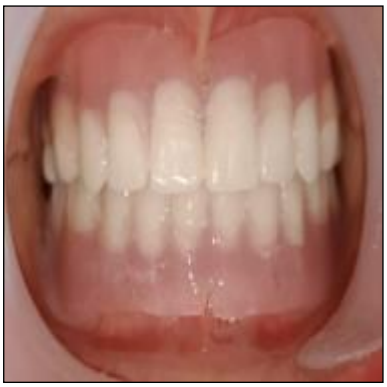

Figure A12: Post-Op View

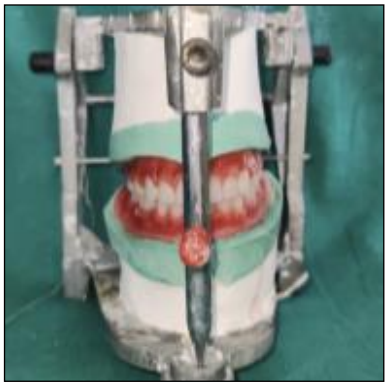

Figure B10: Jaw Relation

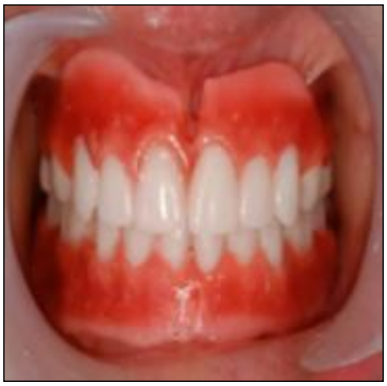

Figure B11: Try-In

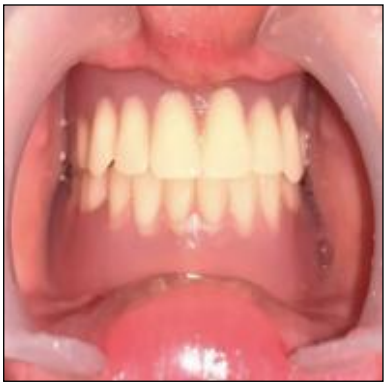

Figure B12: Post-Op

3. Discussion

Christ-Siemens-Touraine syndrome, or HED, is the most common type of ED. It occurs due to any form of genetic defect in ectodysplasin signal transduction pathways.[8] This genetic defect in developing epithelial cells like teeth, hair follicles, nails, and sweat glands can lead to dysplasia, hypoplasia, or aplasia of these structures during morphogenesis. Hypodontia, tooth malformations, and hypoplasia of teeth are seen in oral manifestations of these disturbances. [9] In the present case, the patient showed typical features of Christ-Siemens-Touraine syndrome in his face and oral cavity. Ectodermal dysplasia is generally communicated as an X-linked recessive trait in which the gene is carried by the female and manifested in the male. Carrier females tend to show negligible expression in the form of conical teeth or anodontia and reduced sweating in X-linked type of such syndrome. The unaffected female has a 50% chance of transmitting this disorder to her male children, and each female offspring has a 50% possibility of inheriting the defective gene and being a carrier. [8] Similar circumstances were seen in our cases where the mother was the carrier as she has conical teeth and her brother (patient's maternal uncle) has an indistinguishable condition like them. To maintain sagittal and vertical skeletal relationships during craniofacial growth and development as well as to improve esthetics, speech, and masticatory efficacy, oral rehabilitation of Christ-SiemensTouraine syndrome using complete or removable dentures is of utmost necessary. [9] Prosthodontic rehabilitation at an early age helps the child to adjust to the

prosthesis and develop a normal appearance. It also helps in improving the phonetics, mastication, and the temporomandibular joint function. When the patient's age is more than 12 years, an implant-supported denture is also suggested as an ideal reconstruction modality, but in most cases, it may not be possible without bone grafting. [10] In this case, as the patient's age was less than 12 years, complete denture fabrication was planned. The exact time to initialize prosthetic management is still controversial, where Till and Marquez [11] recommend that a first prosthesis should be given when the child starts school in order to benefit the child in better appearance and to let him adapt to the prosthesis. Due to unavoidable circumstances in our both cases, the patient could not receive any dental treatment prior to their referral to our institution. The conventional removable prosthesis is an appropriate choice for young children with ectodermal dysplasia as they require repeated replacement of prosthesis as the person grows.[12] It requires regular maintenance and should be changed when a reduced vertical dimension of occlusion and an abnormal lower jaw posture are seen due to growth.[13] Follow-ups should be made to match up to the developing jaws. There is a guideline-recommended for relining/rebasing prostheses in a growing patient. It should be done every 2-4 years; remaking a new prosthesis is recommended after 4-6 years.[14] Traditional fixed prosthodontic management cannot be applied in children with Christ-Siemens-Touraine syndrome as they have complete anodontia. [15] Dry oral mucosa, under-developed maxillary tuberosities and alveolar ridges are problematic factors for resistance and stability of the dentures in such patients. [16] Thus, when fabricating dentures for these patients, special attention should be given during impression-making, and care should be taken to obtain a wider distribution of occlusal loads by the denture base extensions. Since the oral management of these cases is often complex, particularly in pediatric patients, treatment is always administered by a multidisciplinary team that includes prosthodontics, pedodontics, orthodontics, and oral-maxillofacial surgery. So, in both cases, we carried out the management in association with the Department of Prosthodontics.

4. Conclusion

Management of Christ-Siemens-Touraine syndrome presents a unique challenge for dentists. So, a multidisciplinary approach is always required. The treatment of young edentulous patients with removable partial or complete dentures is widely acceptable, easy to fabricate, and cost-effective, and it improves the function, speech, esthetics, and psychosocial condition of such patients very effectively and efficiently. However, its long-term success depends on patient compliance, regular recall appointments and meticulous maintenance of oral and prosthetic hygiene.

Ethics Statement
Not applicable.

Funding Statement
This work is not financed.

Conflict of Interest Statement
The authors disclose no potential conflicts of interest.

About the Author(s)
Dr. Sonal Gupta is a professor and Head of the Department at KD Dental College, Mathura, India. She has published more than 100 articles in national and international journal and she's a national level speaker as well.
Dr. Utkarsh Singh is a post graduate student at KD Dental College, Mathura, India.

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