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ORIGINAL ARTICLE

Prosthetic management of an 11-year-old patient with hereditary ectodermal dysplasia and partial anodontia – a case report.

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ABSTRACT
Objective: To illustrate the problems of partial anodontia and replacement of missing teeth and bone in patients with ectodermal dysplasia.
Method: The illustrative case is an 11year old boy with tuberculoid shaped 11, 21 and 16 as the teeth present in the upper arch with anodontia of lower arch.
Result: Upper removable partial denture and lower removable complete denture was fabricated and fitted satisfactory.
Conclusion: The psychological and social embarrassment suffered by children with ectodermal dysplasia associated with missing teeth can be greatly improved with early prosthetic rehabilitation with removable prostheses. Further management with fixed prosthesis may be undertaken as they grow older.

Key words: Ectodermal dysplasia, Prosthetic treatment.

INTRODUCTION

The ectodermal dysplasias are a heterogeneous group of over 150 syndromes characterized by disturbances in the formation and function of ectodermally derived structures, mainly hair, nails, teeth and sweat glands.[1] Dental findings may consist of total anodontia of either the primary or permanent teeth. Cone-shaped teeth frequently occur. The maxilla and mandible grow independently and develop to normal size and shape. The absence of salivary glands is not a common finding and most patients do not complain of xerostomia.[2]

Earlier reports[3,4] have described the most common type of ectodermal dysplasia as anhidrotic ectodermal dysplasia because of the lack of sweat gland function. Due to partial absence of sweat and sebaceous glands, the phenotype has smooth, soft dry and thin skin. Fine linear wrinkles and increase pigmentation are often present around the eyes and mouth.

The mode of inheritance of hypohydrotic ectodermal dysplasia can be either autosomal dominant (AD) autosomal recessive (AR) or X-linked recessive (XLR) and the gene locus is X q^{13}-$q^{21.}$ It is commonly X linked recessive with full

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expression in males; female carriers have a minimal expression in 60-70% of cases and usually show mild manifestation restricted to minimal hypodontia such as absent maxillary lateral incisors or presence of conical teeth.[6]
All racial groups have been afflicted by this condition. Attempts have been made in the past to develop objective diagnostic criteria based on the number and distribution of sweat pores and the number of sweat produced. Structural and biochemical characteristic of the hairs, dermatologlypic analysis, characteristics of the lacrimal secretion and the distribution and pattern of scalp hair have also been used. However, universally acceptable standards have not been provided[7]. An important factor is that the facial appearance of affected individuals is so characteristic that unrelated patients may be mistaken for siblings.

This case report describes the dental abnormalities of ectodermala dysplasia as seen in an 11 year old male patient and the challenges of restoring aesthetics and function with removable prosthesis.

Case Report
Master T.O. was 11 years old when he presented at the Prosthetic out-patient clinic of Lagos University Teaching Hospital (LUTH) accompanied by his mother requesting for dentures to replace his missing teeth. The patient became aware of his condition when he started primary school but his mother did not think it was necessary to seek treatment at that time. On admission into secondary school at the time of presentation he urged his mother to bring him for fabrication of dentures because he did not want to suffer and go through the embarrassment of his younger days in primary school in secondary school.

The mother claimed that the patient was a full-term baby delivered in the same hospital (LUTH) and that the only teeth in his mouth were the only set of teeth that has ever erupted into his mouth. The patient cannot tolerate heat and hardly sweats. The mother's youngest male sibling has the same condition. The parents were not consanguineous.

The patient has scanty fine scalp hair and eye brow, with fine linear wrinkles around the eyes. His skin is smooth and dry. He has protuberant lips with fissuring of the angles of the mouth. His finger nails were normal **(Fig 1a,1b)**

Figure 1a: shows the facial features of the patient.

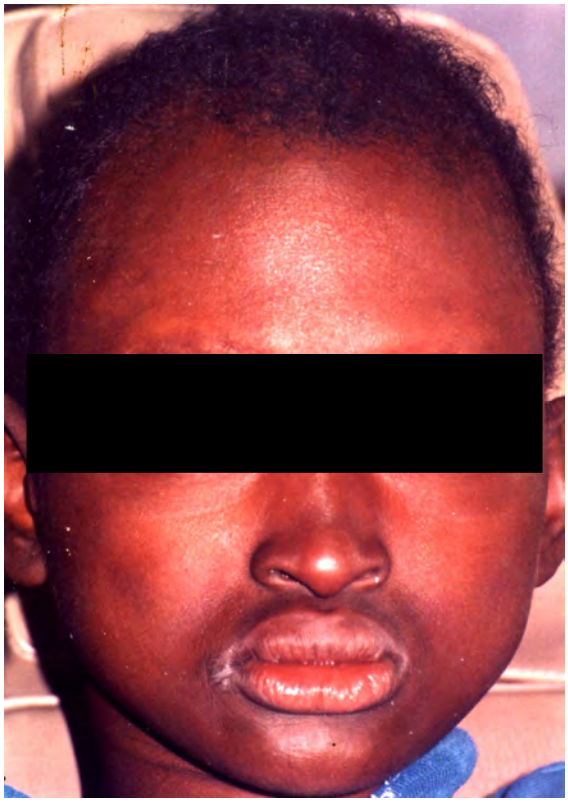
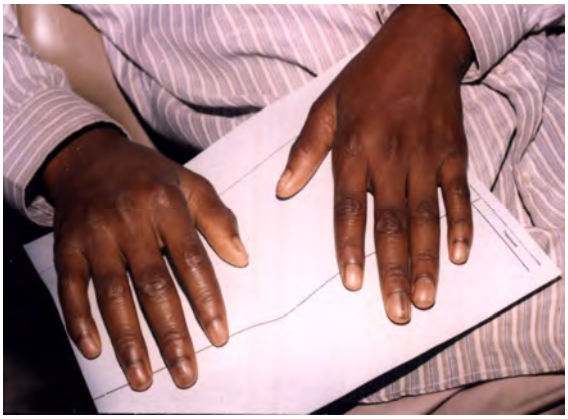
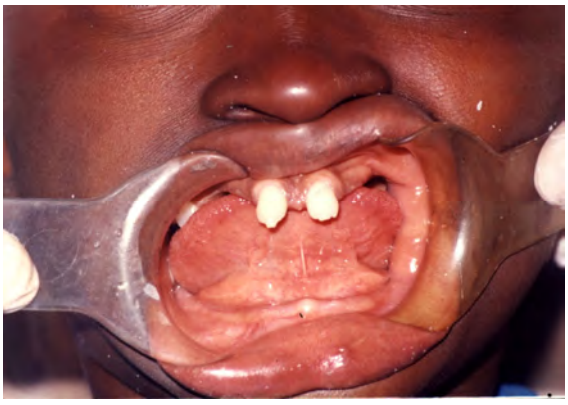


Figure 1b: shows the fingers with normal nails.



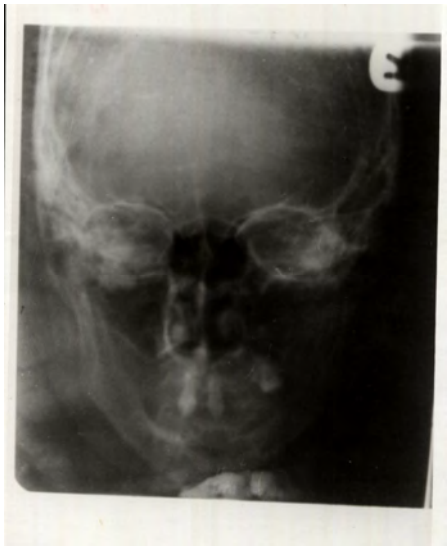
Intraorally, present in the upper jaw anteriority were two tuberculoid shaped central incisors (11,21) and posteriorly on the right a malformed first molar (16). The lower jaw was completely edentulous. **Fig 2.**

Figure 2: Tuberculoid shaped upper central incisors and upper right first molar and edentulous mandible.



The palatal vault was high arched, the upper and lower alveolar ridge heights was low. Posterior-Anterior radiographs of the jaws did not reveal any buried teeth. **Fig. 3.**

Figure 3: Posterior –Anterior radiograph of the jaws which did not reveal any buried teeth.



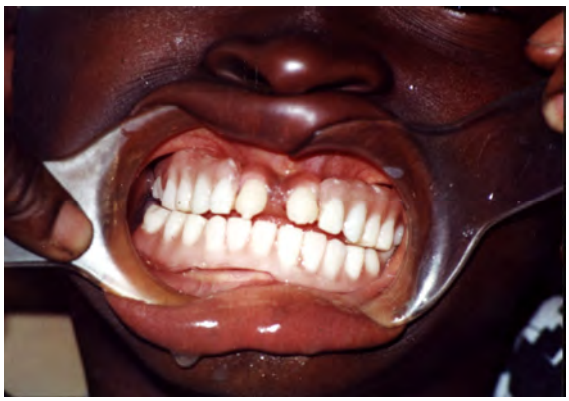
Examination of the 47 year of mother did not reveal any extra oral anormally. Intraorally the 12, was pegshaped, 13, 23 was congenitally missing, retained 63. Lower 42,41,31,32 were missing, patient could not explain how they got missing but has been wearing a lower removable partial denture replacing 42,41,31,32 for 33 years.
Based on the clinical findings a diagnosis of Hereditary Ectodermal dysplasia was made.

Treatment
Upper and lower alginate impressions were taken, using stock trays which was poured in dental stone. Special trays were fabricated using self curing acrylic resisns. A two staged secondary impression were taken using green stick impression material for border moulding and zinc oxide eugenol impression paste for the wash impression.

Record blocks were constructed to register his jaw relations. The rest vertical dimension was established using

the Willis gauge. Acrylic denture teeth were set up at the determined occulusal vertical dimension. The trial dentures were aesthetically pleasant, retentive and stable. The dentures were processed using heat curing acrylic resin. The dentures were eventually fitted satisfactorily. **Fig4.**

Figure 4: Upper removable partial and lower complete dentures in occlusion.



The patient has attended recall visits monthly for one years during which slight relieve of pressure areas has been carried out and there has been no new complaints. Patient has since been referred to the conservative clinic for the composite veneer restorations of 11,21 and porcelain fused to metal crown on 16. A definitive cobalt-chromium partial denture will subsequently be fabricated using the crowned 16 as an abutment. The need for continuous recall visits was greatly emphasized.

DISCUSSION

A diagnosis of hereditary ectodermal dysplasia was made from the history and clinical findings. Previous studies [7,8-] in Nigerians however made a further diagnosis of the anhidrotic ectodermal dysplasia type based on microscopical examination of finger tips for sweat pores and skin biopsy findings. The goals of management of this patient with ectodermal dysplasia is to restore the facial appearance to as near as possible. This helps the patient's psychological development and help establish a normal childhood for the patient.[9] This is the most important management in early childhood.

A precious case study[10] reported obvious and dramatic change in the emotions of a 2 year 8 months old girl after the fittings of complete dentures; she became more co-operative, friendly and willing to participate in the game with other children in the neighbourhood. Our patient also demonstrated such a joyous emotion and excitement on the day of try-in of the trial dentures and was disappointed when he realized that he was not going to go home with the trial dentures.

Partial and total anodontia of both deciduous and permanent teeth in patients with ectodermal dysplasia is very common. Our patient did not have any deciduous dentition. Age is not a barrier to prosthetic management, as early in life as possible treatment may be commenced and children have been seen to cooperate well with dental procedures.

Challenges of replacement with removable prostheses are that of providing retentions stability and ensuring support. There was marked reduction in the denture bearing area of the two arches, this problem was overcome by conservative management using Boucher's[11] impression procedure which ensures peripheral seal and intimate tissue contact.

Stability was a major problem because of high muscular activity of the tongue which usually undergo lateral spread due

to absence of the teeth. Due to the fact that occlusal dynamics is essentially very new to our patient, the dentures were fabricated at a very low occlusal table using teeth that are narrow buccolingually and almost cuspless.

To improve aesthetics the tuberculoid shaped incisors were to be restored with composite veneers and a porcelain fused to metal crown was to be placed on the upper right molar to further improve the retention of a clasped upper removable partial denture.

Some authors[12,13] have proposed different treatment modalities, timing and treatment protocol for meeting the patient's functional and aesthetic needs as they grow into adulthood. The prosthetic rehabilitation is a life long management as reports[14] have illustrated with cases that have been managed for over a 20 year period. Early treatment starts with a replacement of missing teeth with simple acrylic based removable partial or complete dentures. The teeth present may be crowned or restored with composite veneers to help with the retention of the denture and also to improve aesthetics.

As the child grows and there is further concomitant growth of the maxilla and mandible, several other treatment options such as sulcus deepening to improve areas of contact of denture with supporting tissues, ridge augmentation with bone grafts and implant fixed Prosthesis [2,15,16] have been carried out with remarkable success.

CONCLUSION

Ectodermal dysplasia is both physically and emotionally devastating to patients affected. With proper prosthetic management early in childhood, the quality of life can be improved. It is important that these patients be seen and treatment commenced at an early age to aid in their social interactions.

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