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Prosthetic Rehabilitation of a Patient with Apert Syndrome: A Case Report

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Abstract: *Apert syndrome is a rare, congenital, developmental disorder characterized by craniosynostosis, cone shaped calvarium, midface hypoplasia, ocular manifestations and symmetric syndactyly of hands and feet. These incapacities emphasize the imperative of multidisciplinary approaches, considered invariably at a very early stage including pedodontic and orthodontic management. A prosthetic management of the condition is arguably more economical than complex surgical treatments when the patient's demand is more on esthetic correction, especially in the advanced stage. The present case study is a detailed report of the treatment of a patient diagnosed with Apert syndrome distinguished with a deformity in the upper anterior region using removable prostheses.*

Keywords: *Apert syndrome; Craniosynostosis; Removable prostheses; Telescopic overdenture.*

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I. Introduction

Apert syndrome or Acrocephalosyndactyly is a rare congenital condition usually inherited as an autosomal dominant trait.[1]Craniosynostosis, cone shaped calvaria, acrocephaly, pharyngeal attenuation, symmetric syndactyly of the hands and feet, exophthalmia, ocular hypertelorism and hypoplastic midface with Class III malocclusion and systemic alterations are characteristic of this disorder.[2] As early as 1842, Baumgartner pioneered in the diagnosis of this syndrome and was followed by Wheaton in 1894. But the eponymous Eugene Apert, a French Pediatrician, was given the honour in 1906, as he described a series of 9 cases with similar attributes of the syndrome.[3]

Apert syndrome is estimated to affect 1 in 160,000 live births. It is most frequently caused by a de novo mutation in the male gameter. Two missense mutations in the fibroblast growth factor receptor 2 (FGFR2) gene on chromosome 10, have been found to account for the disorder in approximately 98% of patients.[4]Prosthodontic rehabilitation of the condition currently confronts a serious relapse as the preponderance of investigations reported on Apert syndrome concern the orthodontic, pedodontic or surgical management of the disorder. The present study proposes to discuss in detail the prosthetic rehabilitation of Apert syndrome identified on a patient with the aim of aesthetically reclaiming the dental deformation in the upper anterior region.

II. Case Report

An 18-year-old male patient reported to the Department of Prosthodontics, KMCT Dental College, Calicut, Kerala with the chief complaint of a lack of aesthetics in the upper front tooth region. Patient was a known case of Apert syndrome as seen in Figure 1a &b.Detailed history revealed that he was the first child of an apparently normal mother. No history of consanguineous marriage of his parents was reported. Patient's parents and 11yr old sibling did not manifest any related findings. The patient had normal development with minimal signs of mental retardation. His speech was found to be slightly incoherent. Patient had a history of bilateral cleft for which he had undergone surgery 2 years back. Clinical, extra oral, and intraoral examination revealed all abnormal features which where characteristic of Apert syndrome. In addition to that patient had missing teeth in relation to maxillary right and left central and lateral incisors which was extracted $1^{1/2}$ years back due to caries as seen in Figure2.

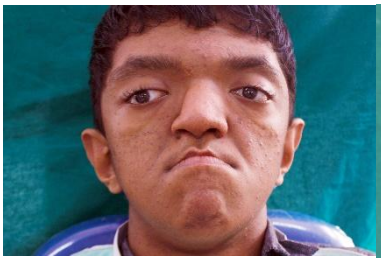

Fig 1a: Extraoral view : Frontal

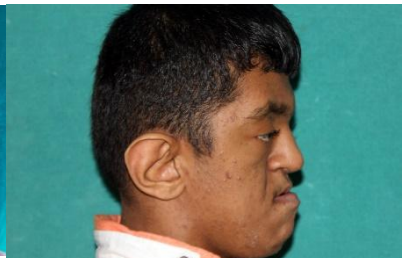

Fig 1b: Extraoral view : Lateral

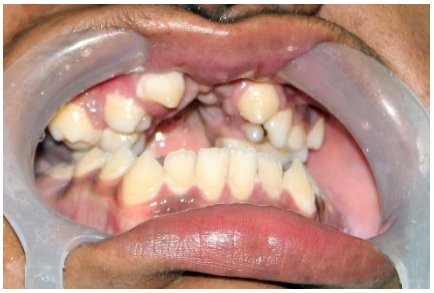

Fig 2: Intraoral view

Various treatment options like fixed partial denture, removable partial denture covering the edentulous area alone, removable partial denture covering the edentulous area and the existing natural dentition and implant supported overdenture were explained. Discussions with the patient's parents revealed that a procedure which gave credence to the facial upliftment as the necessity. The first two methods were discarded as it would not provide the required aesthetics and retention. Apert syndrome exhibits a progressive deterioration of the dental development as age advances. This validates the early commencement of treatment for the given condition. Therefore, after careful consideration of patient's mental instability, and dental development, for the time being a removable partial denture covering the existing natural dentition and edentulous area was suggested as a better treatment plan.

Patient's parents agreed with the treatment plan and informed consent were obtained from them. Accordingly the treatment was initiated with a primary impression made using irreversible hydrocolloid impression material. The minimal functional movements expected voluntarily from the patient have to be enabled artificially. Hence, the casts obtained were mounted on a semi-adjustable articulator during teeth arrangement. A face-bow transfer was done and maxillary cast was mounted as seen in Figure 3. For mounting the mandibular cast in centric relation with maxillary cast, an interocclusal record was made with aluwax and mounting was performed. Since the patient was unable to perform protrusive movements, the condylar angle of the patient was calculated on panoramic radiograph with the help of the software'SironaSidexis' as seen in Figure 4. The highest point on the articular eminence and lowest point on the glenoid fossa were taken as the 2 points of reference. The angles obtained were 37.1° on right and 33.9° on left side. Accordingly, it was set on the articulator.

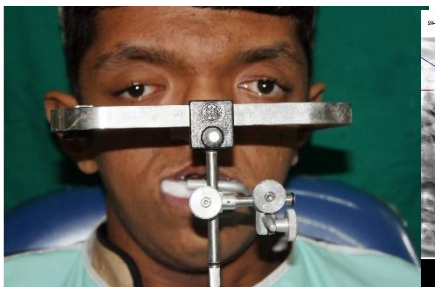

Fig 3: Facebow transfer

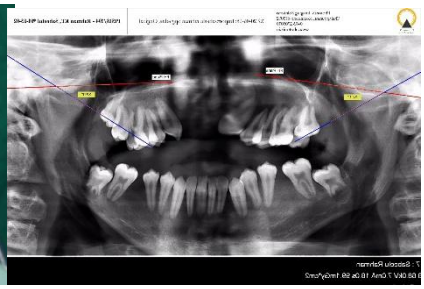

Fig4: Panoramic Radiograph

Following this, an occlusal rim was fabricated over the edentulous area and remaining natural teeth in the maxillary arch. The occlusal plane, visibility, and fullness was analyzed. Anterior teeth arrangement and try-in was performed followed by posterior teeth arrangement, ensuring a centric cusp contact on either side as seen in Figure 5. Functional movements were done on the articulator. After try-in it was sent to the laboratory for the fabrication of the denture. The denture was inserted in the patient's mouth and checked for any corrections required as seen in Figure 6 a & b. The patient as well as his parents were fully satisfied with the results as seen in Figure 7 a & b. Retention of the denture was achieved from the undercuts present on the right buccal and left palatal region.

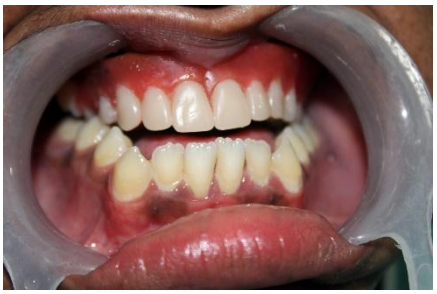

Fig 5: Try - in of the denture

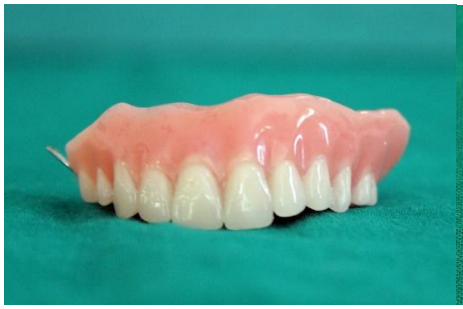

Fig 6a: Final Prostheses

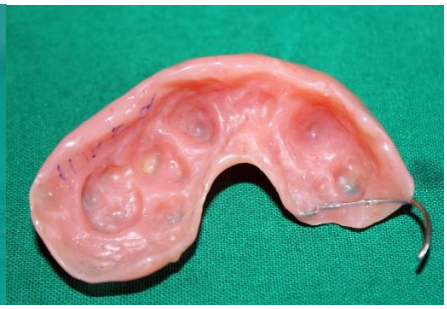

Fig 6b: Intaglio surface

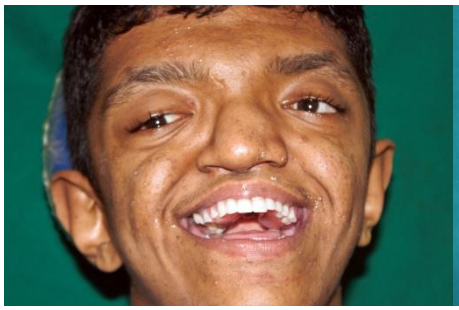

Fig 7a: Postoperative view: Frontal

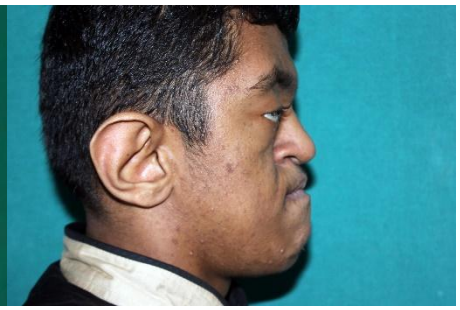

Fig 7b: Postoperative view: Lateral

The patient was successively reviewed post-treatment in a sequence of 1 day, 1 week and 1 month. Reviews evidently showed a psychological improvement in the patient. Patient was visibly socializing better and also initiated interactions with others. Application of fluoride was essential to alleviate the carious tooth over which denture was placed and a fluoride toothpaste was prescribed as well. It is important to note that the suggested treatment of removable partial denture is intended as a transitional implement which would assist the patient to adapt with the prosthesis. Such a transitional denture would enable the evaluation of esthetics achieved. In the event of the patient getting acclimatized to the current denture a telescopic overdenture is prescribed. This shall be the definitive treatment.

III. Discussion

The treatment of Apert syndrome must commence at birth or at the least in the pediatric stages and a life-long management supervised by a team of healthcare specialists must be ensured. The challenges significant to conventional dentistry is the complex relationship between the supporting basal maxillary and mandibular bones as well as the pseudo or real cleft palate.[5]The majority of these concerns are related with esthetics, breathing, bottle-feeding, and swallowing difficulties in infants. The dental management of this syndrome is marked by its unique and integrated treatment plans because each patient manifests unique physical and mental challenges that requires an individualized, comprehensive and collaborative care.

The present case exemplifies the centrality of individualized and tailored treatment plans for Apert syndrome. The patient was neither administered any treatment at birth nor been consulted for diagnosis throughout his childhood. Lack of long-term intensive care has resulted in the patient's loss of dentition. Therefore, orthodontic management falls counterproductive. Loss of dentition on the other hand, opens a way for prosthodontic rehabilitation. There are little or no cases reported regarding the prosthodontic management of Apert syndrome and therefore the present case study stands out as a pioneer in devising a treatment plan for the same. Considering the patient's pressing demand which was to achieve a presentable aesthetics in the facial area, an attempt is made here to prosthetically rehabilitate the oral and facial anomalies that accompany Apert syndrome.

IV. Conclusion

The present study significantly raises the bar of prosthetically rehabilitating Apert syndrome as it enables the amelioration of the most conspicuous yet under-acknowledged everyday challenge of coping with the syndrome – the chronic disfiguration of elementary facial features. It may do well to be aware of the old saying, "Appearances are deceptive", but in medical history as well as psychological research, the apparent aesthetics of appearance has a critical contribution towards the development of one's character, social skills and in consequence one's holistic well-being. This case study also demonstrates the psychosomatic implications of a successful prosthetic rehabilitation of Apert syndrome as the patient exhibited visible behavioural progress post reclamation of the aesthetics of his face.

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