

# Rehabilitation of a Treacher Collins Syndrome Patient with Adhesive Retained Bilateral Auricular Prosthesis

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## Abstract

**T**reacher Collins syndrome is a rare autosomal dominant congenital disorder characterized by craniofacial deformities like micrognathia underdeveloped zygoma, malformed or absent ear. Rehabilitation of a patient with craniofacial deformities should be done as early as possible in order to reduce the psychological impact on the patient's, ideal treatment is autogenous reconstruction or use of alloplastic implants for surgical reconstruction. But if this treatment is not affordable to patient due to financial constrain then, prosthetic rehabilitation is the next best treatment option. Retention of auricular prosthesis can be achieved by use of spectacles, head bands, adhesives or implants. The following case report describes the fabrication of an adhesive retained auricular prosthesis for replacement of a bilateral ear defect in a patient who had congenitally missing ears for last 18 years.

**Key Words:** Treacher Collins syndrome, Bilateral auricular defect, Silicon prosthesis, Adhesive retained.

## Introduction

Treacher Collins syndrome is an inherited developmental disorder with a prevalence estimated to range between 1 in 40,000 to 1 in 70,000 of live births<sup>1,2</sup>. The particular gene that causes Treacher Collins syndrome is called TCOF1. This gene, located on chromosome 5 is responsible for facial development. In about half of all cases, TCOF1 spontaneously changes at conception but what triggers the mutation is unknown.<sup>3</sup>

In seventh century, replacement of the exterior part of auricle was termed epithesis.<sup>4</sup> Surgical correction and prosthetic rehabilitation are the two options available for correction of auricular defects. Auricular reconstruction can be done surgically by implantation with autogenous cartilage grafts. Tissue engineering is a newer method for potential clinical application in auricular reconstruction.<sup>5</sup> If the patient are not a candidate for surgical reconstruction because of high operative risk, severely compromised tissue or financial constrain then prosthetics replacement is the next best option. Prosthetic replacement of missing facial tissues has several advantages over surgical reconstruction. The process is relatively inexpensive, allows for periodic evaluation and cleaning of the site. It is a short process and the maxillofacial clinician has complete control of color, shape and position of the prosthesis. Disadvantages include possible

irritation of the tissue site, need for periodic remake and depending on adhesives or some other form of retention.<sup>6</sup> These prostheses are retained with different methods of retention such as medical adhesives, anatomical undercuts, and mechanical devices like spectacles, hair bands, magnets and implants. Since the introduction of percutaneous endosseous implants for use with bone conduction hearing aids in 1977, implants have acquired important role in the prosthetic rehabilitation of patients with craniofacial defects.<sup>7</sup> Implants can vastly improve the retention and stability of a facial prosthesis. The co-ordinated participation of surgeon and maxillofacial prosthodontist is required in presurgical planning to determine the number, type and positioning of the implants in the defect.

This case report is a brief description of fabrication of adhesive retained silicon prosthesis for a Treacher Collins syndrome patient who had bilateral auricular defect.

## Case Report

A 18 year old patient was referred to Department Prosthodontics with a history of Treacher Collins syndrome and missing bilateral ears. On examination the patient was found to be short stature, low weight, micrognathis and facial asymmetries were observed like deviated nose and lip which were due to surgical reconstruction of cleft lip & palate. External parts of auricles were totally missing only the auricular tubes were present at a distance of 5 cm from angle of the mandible (Fig. 1). Otorhinolaryngologist evaluation found conductive hearing loss. Mental status was found to be normal with developmental delay due to undiagnosed hearing loss. The patient was undergoing speech and hearing therapy. Family history did not reveal any significant data. Keeping in mind the psychological impact of the deformity on the patient's development, adhesive retained silicone prosthesis was planned as an interim option, till the patient could afford for implant retained auricular prosthesis.

## Procedure

The wax pattern of the ear was obtained by 'donor technique'. The patient mother was used as donor for auricular impression. After applying petrolatum to adjacent hair, impression was obtained in irreversible hydrocolloid (Tropicalgin; Zermach Inc, California), with the donor lying on her side in a supine position in a large plastic ring. (Fig. 2). Two impression were made of right and left auricle. And hard baseplate wax (AL-

192A, Technovent USA) was melted and poured into the impressions to obtain the two auricular wax pattern. Then petrolatum was applied to the adjacent hair of the patient and the wax pattern thus obtained were used as a tray for impression with irreversible hydrocolloid of the defect & for evaluation of the final orientation of the prosthesis on the face (Fig. 3). Condylar movements were examined to check for mobility of tissue that may affect the placement of the margin, tissue coverage and retention of the prosthesis<sup>8</sup> Master cast was poured and the wax pattern was readapted to the working cast and sculpted to mimic the contours of the normal ear. Surface texture on the pattern was created by using a damp sponge (Fig. 4).

The processing was done in three part mould. V-shaped notches were prepared on all four corners of the working cast. After applying a separating medium (Coe-sep, GC, USA), the posterior under-surface of the sculpted ear was registered by placing a mix of type III stone posteriorly along the greatest dimension of the helix and lobe enclosing all of the posterior aspect of the form. After the material had set, escape channels for silicone material were prepared by placing strips of wax (Fig. 5). After application of another layer of separating medium, the entire working cast with the second part of the mold was boxed and type III stone was poured to completely cover the rest of the wax form and the outer surface of the second part of the mold. Dewaxing was done after the mold had set. The three parts of the mold were separated (Fig. 6).

Packing was done in room temperature vulcalising silicone material (Cosmesil; medical grade, Technovent Co,UK) was used for fabrication of the prosthesis. Intrinsic shades were used for matching the skin tone of the patient. The evaluation of the shade was done under natural light. After applying separating medium (soap solution), a little darker shade than the patient's basic skin tone was brushed onto the mold surface in places where shadows were observed. The three parts of the mold were approximated into a tightly fitting mold assembly after generously coating the entire mold surface with the basic color silicone. The final prosthesis was allowed to cure for 24 hours before retrieving it for finishing. Final color matching was done with extrinsic pigmentation (Cosmesil; Silicon coloring kit, Technovent Co,UK) in presence of patient. The mold was preserved for use when the patient would require a replacement. The final prosthesis was then

placed in position using medical adhesive (Fig.7 & 8).

Medical adhesive (Edge adhesive, Cosmesil; Technovent Co,UK) was applied in a thin film on the tissue surface of the prosthesis and on cleaned skin surface and allowed to dry. The prosthesis was then placed in position by applying slight pressure. The guardian of the patient was taught how to place and remove the prosthesis. They were also instructed to keep the skin clean and free of oil secretions. While cleaning the prosthesis they were advised to ensure complete removal of the adhesive both on the skin and the prosthesis.

**Discussion**

The current approach for Treacher Collins syndrome (TCS) clinical deformities seeks functional and esthetical corrections as well as psychosocial support. Multi-disciplinary approach, including otorhinolaryngologists, craniofacial surgeons, ophthalmologists, speech therapists, psychologists and pediatric dentist, is the most appropriate way to manage these patients. In addition to anatomical and physiological anomalies, TCS patients carry a social stigma because of its severe facial deformities.<sup>9</sup>

Prosthetic rehabilitation of auricular defects is performed due unsuitability of the patient for surgery, due to surgical complications<sup>10</sup> or because of economic constraints.<sup>11</sup> Retention of prosthesis can be achieved by mechanical means or by adhesives or implants. Mechanical retention may be by way of tissue undercuts or skin tunnels, attachment to spectacles or

headband<sup>12</sup>. Implants are the preferred mode of retention for the auricular prosthesis as they have better patient acceptance and treatment satisfaction.<sup>13</sup> Recent advances in techniques, including a new generation of Computed Tomography scanner and three dimensional (3D) systems facilitate the production of mirror image of auricular prosthesis with high level of accuracy, alleviating most of the limitations of conventional prosthesis.<sup>14</sup> Limitation to its use is high cost. Development in the field of tissue engineering has resulted in the formation of new tissue equivalents of bone and cartilage that will augment the outcome of prosthodontic rehabilitation in future<sup>15</sup>. Evidenced based studies pertaining to the value of facial prosthetics have to be addressed to better understanding of the economical, functional and psychological burden of having a facial ablative procedure involving the orofacial, ocular, auricular and nasal tissues. Psychologic problems in children with craniofacial deformities have included lack of emotional attachment between parent and child, inadequate development of peer relationships, and the experience of shame related to a poor body image.<sup>10</sup> While delaying the implant treatment, we fabricated an adhesive retained auricular prosthesis to be used by the patient for a temporary period.

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**Fig. 1**

**Fig. 2**

**Fig. 3**

**Fig. 4**



**Fig. 5**

**Fig. 6**

**Fig. 7**

**Fig. 8**

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