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CLINICAL REPORT

Prosthetic Rehabilitation of a Patient with Congenitally Deformed Ears

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Abstract Treacher Collins syndrome is a rare autosomal dominant congenital disorder characterized by craniofacial deformities and is found in about 1 in 50,000 births. This is a bilaterally symmetrical abnormalities derived from the first and second brachial arches and the nasal placode. Unfortunately, many of the new surgical techniques are extensive and compromise the patient's quality of life, not only function and esthetics but also the psychological status of the patient. These problems require prompt rehabilitation with surgery or prosthetic rehabilitation. This article presents a procedure in the basic fabrication of a prosthetic ear by a three-piece die technique.

Keywords Auricular prosthesis · Facial rehabilitation · Three-piece mould

Introduction

Ear prosthesis artificially restores the ear which has been lost due to radical cancer surgery, amputation, burns and/or congenital defects [1]. Treacher Collins syndrome is one of the developmental anomalies that affect the bones and other tissues of face. It is characterized by underdeveloped facial bones particularly hypoplasia of zygoma, micrognathia and anomalies of ears and eyes [2, 3]. In some people, cleft lip and cleft palate are also often observed. In severe cases, underdevelopment of the facial bones may restrict an affected infant's airway, causing potentially life-threatening respiratory problems. The clinical appearance of such affected individuals can be improved by rehabilitation with maxillofacial prosthesis and/or cosmetic surgeries. This will also improve the psychological status of the patient. In the literature, various methods have been suggested for the retention of the prosthesis [4–8]. This case report presents the clinical features, diagnosis and the procedure involved in rehabilitation of a case of Treacher Collins syndrome.

Case Report

A 19-year-old boy was brought to our institution with a chief complaint of missing ears and forwardly placed teeth. Extra-oral examination showed deformed ears which were flabby with no cartilage. Zygoma and mandible were underdeveloped. The patient had Angle's class II malocclusion and high arched palate. Patient is deaf and was following lip reading/speech reading, for which he was consulting the speech therapist. Radiographic examination (OPG, CT-scan, lateral cephalometric radiograph), exhibited under developed zygoma and absence of external auditory canals by using temporal bone CT scan using thin slices. Depend upon these clinical and radiographic findings; the patient was diagnosed as having Treacher Collins syndrome.

Clinical examination demanded the need of fabricating ear prosthesis to cover the deformed ears and a combination of orthodontic and surgical intervention to correct the malocclusion and facial deformity.

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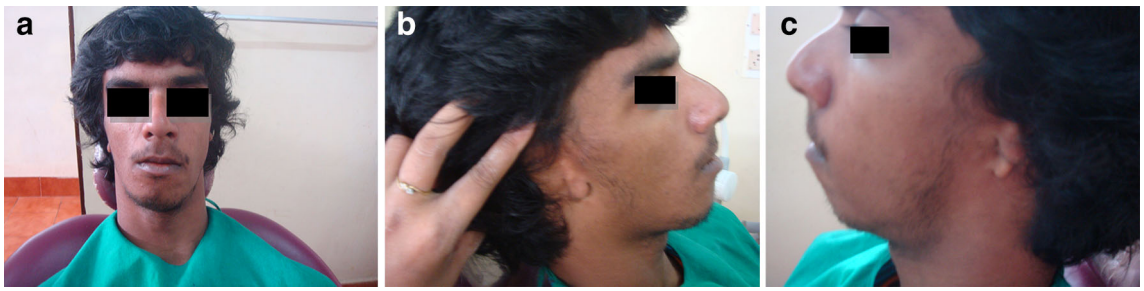

Fig. 1 **a** Preoperative view (frontal view). **b** Right side view. **c** Left side

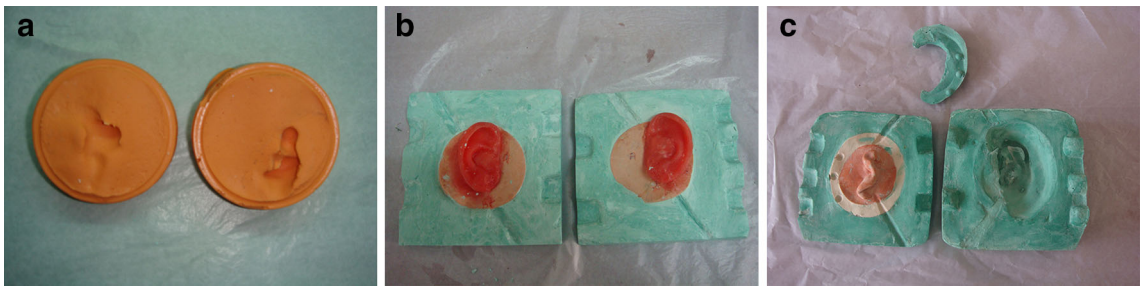

Fig. 2 **a** Impressions of the first part of the mould deformed ears. **b** Wax pattern adapted to first part of the mould. **c** Three part mould

Procedure

Impressions

The patient was seated in a dental chair in a supine position (Fig. 1a) and viewed the existing condition of both right and left ear (Fig. 1b, c). Impressions were made using alginate impression material (Zhermack Neocolloid alginate, Zhermack S.p.A, Italy) by standard impression procedure. After the setting of the impression, it was removed keeping in mind the angle of existing undercuts to prevent tearing. The impression was inspected for accuracy (Fig. 2a). The procedure was repeated for the other ear [8–10].

Sculpting Technique

The impressions were poured into working casts. As both the ears were deformed, patient's sibling's ears (donor) were selected to make the wax patterns. The impressions of donor ears were also made similar to the above mentioned procedure. Molten base plate wax was poured into the set impressions and allowed to cool. After the impressions were completely set, it was retrieved from the impression and adapted onto the cast (Fig. 2b) [6, 11, 12].

Try In

The wax patterns were tried individually on the patient (Fig. 3a–c), and modified to suite his facial appearance and the following were checked:

1. The fit of the prosthesis on the tissue.
2. The correct horizontal alignment with the opposing ear.

There should be two marks present:
(a) The junction of the helix with the side of the head.
(b) The junction of the lobe with the side of the head.
These points are observed by looking at the patient from the front elevation and lining the marks horizontally with the natural ear [9]. In this case report, we compared the patient's ear size, shape and alignment with that of his sibling's ear.

3. The projection of the ear in relation to the side of the head.
4. The integrity of the margins during simple jaw movements.

Investing Technique

The wax prosthesis was seated to the model and the leading edge thinned as much as possible so as to allow the silicone edges to feather into the natural skin. A three part mould was made using dental stone, which is necessary to achieve easy placement of the silicone (Fig. 2c) [9, 10].

Processing of the Prosthesis

Twenty grams of silicone elastomer (Cosmesil, Technovent; Leedes, UK) were measured on a white tile and 2 ml

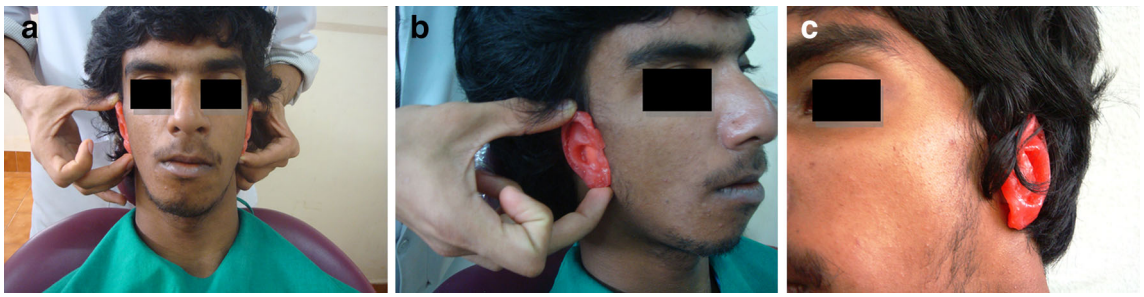

Fig. 3 **a** Wax pattern-Try in (frontal view). **b** Right side view. **c** Left side view

of catalyst was added and mixed to get a homogenous mix [9]. Basic intrinsic skin color was developed by slowly incorporating various silicone pigment tones as follows P416, P409, P105, P406, P106, and P117 (Cosmesil) into the major pool of resins. The blend was compared to the natural skin by occasional holding of a sample on the mixing spatula near the ear of the patient and the tone was increased by introducing small amounts of stronger resin pigments. After establishing the final matching blend of the basic color, all the three mould surfaces were packed with silicone and using keyed margins, the mould was accurately assembled to create the complete mould form. The mould was placed under bench press for 24 h for complete catalytic action to take place. Once the silicone prosthesis was processed, it was carefully retrieved. The flash was removed by trimming with a silicone trimming bur (Dreve-Dentamid) [13].

Extrinsic Coloring

The extrinsic skin color was developed by slowly incorporating Part A (Extrinsic Sealant P799) and Part B (Extrinsic Sealant P799) in 1:1 ratio initially and then, the following extrinsic stains P702d, P702f, P702g, P702c, and P217 were incorporated and the blend was compared to the natural skin. A small amount of extrinsic stain was applied to the external surface for added color tone enhancement. The extrinsic color application was completely allowed to air dry.

Retention and Care of the Prosthesis

Retention of the prosthesis was achieved by adhesives (Cosmedica PSA 1) which was painted thinly on to the fitting surface. After 1–2 min the adhesive will turn clear in color which gives the patient an indication that the prosthesis is ready to be applied (Fig. 4a–c). Patient was instructed to keep the skin surface clean and free of natural oil secretions to ensure proper adhesion of the appliance. He was also instructed to remove the prosthesis while sleeping and taking bath.

Discussion

Treacher Collins syndrome is an autosomal dominant disorder which affects an estimated 1 in 50,000 people, and is derived from the first and second branchial arches and the nasal placode [2, 3]. It is also called as Franceschetti–Zwahlen–Klein syndrome, Mandibulofacial dysostosis, and Zygoauromandibular dysplasia [2]. Patient with Treacher Collins syndrome is characterized by absent or unusually formed ears and sometimes, hearing loss which is caused by defects of the three small bones in the middle ear or by underdevelopment of the ear canal. People with this anomaly often have eyes that slant downwards, sparse eyelashes and eyelid coloboma [6]. Other findings are hypoplasia of zygoma, and the mandible is underdeveloped, resulting in retruded chin. Some patient may also show macrostomia and cleft palate with or without cleft lip and these facial features have been described as “bird-like or fish-like in nature” [2].

The facial appearance can be improved by either surgical or prosthetic rehabilitation. In this case report, auricular prosthesis, orthodontic treatment and orthognathic treatment were suggested to the patient to correct the ear deformity, malocclusion and skeletal discrepancy of jaws respectively.

Prosthetic rehabilitation is the most conservative method of correcting ear deformity, as the surgical reconstruction is complex in nature and also, it is challenging for the surgeons. The technique in this case report was followed by making the impression from donor’s ears (sibling’s) as both the ears were deformed and poured into the model. By comparing with the sibling’s ears, the wax patterns were sculptured. This technique of sculpturing gives a natural appearance of the wax pattern and it is time consuming.

The retention of the prosthesis was achieved by adhesives, which was applied to the fitting surface of the prosthesis. In the literature, various methods have been suggested for the retention of the prosthesis [4–8].

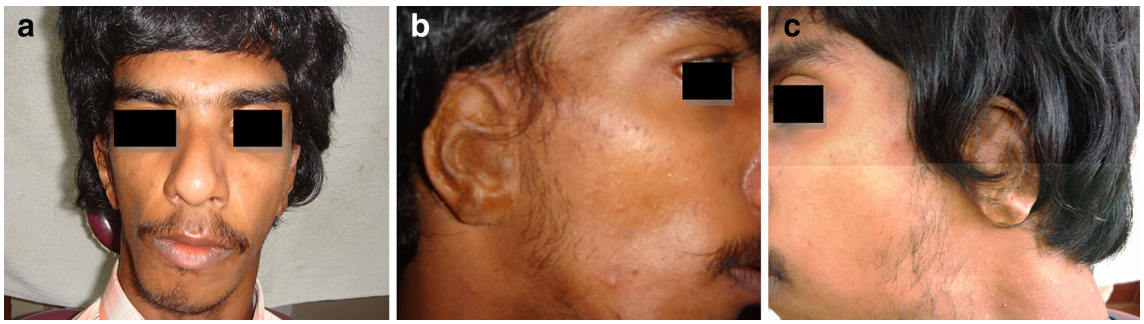

Fig. 4 **a** Postoperative. **b** Right side view. **c** Left side view

Conclusion

Management of Treacher Collins syndrome needs a multidisciplinary approach. The treatment plan must be explained to the patient and to the guardian to meet his/her need, considering the growth pattern, function and psychological development. This article presents a case report of successful rehabilitation of a 19-year-old boy who had congenitally deformed ears using silicone prosthesis by a three-piece die technique.

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