Nasal reconstruction of a patient with complete congenital arhinia:

A clinical report

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ABSTRACT

Complete congenital arhinia is a rare embryonic disorder of unknown etiology. This is a clinical report of prosthetic nasal rehabilitations done in the early childhood and adolescent stages of a patient with complete congenital arhinia. Additive

manufacturing techniques for creating pre-surgical planning models to assist in the creation of new nasal passages is also described. These rare cases can be successfully rehabilitated if patients are sufficiently motivated and there is meticulous planning and collaboration from a multidisciplinary team.

Keywords: congenital arhinia; additive manufacturing.

INTRODUCTION

Complete congenital arhinia is an extremely rare embryological disorder of unknown aetiology, with both sporadic and familial cases having been reported.[1,2] There are less than 50 cases reported in the medical literature,[3] and it is usually associated with other craniofacial abnormalities such as underdeveloped maxillae, cleft lips and cleft palates.[2,4-8] Some associated conditions which may impact patient prognosis include microphthalmia, hypertelorism, absent nasolacrimal ducts,[9] rudimentary nasal bones, Treacher Collins syndrome,[10] and central nervous system defects.[7]

Pathogenesis, though poorly understood, involves disturbances in the complex pathway of embryological development. These may include a lack of invagination of the nasal placodes, reduced growth of the median and lateral nasal processes.[1] premature fusion of the median nasal processes, failure of resorption of the nasal epithelial plugs, or abnormal migration of the neural crest cells.[1,5]

CASE HISTORY

Ten years ago, a 7-year-old female patient from a rural settlement presented with complete congenital arhinia. She was systemically healthy, but had delayed growth and cognitive skills for her age. The medial canthi of both eyes were fissured and

drooped inferiorly (coloboma palpebrae), and she had bilateral strabismus. There was delayed growth of the midface vertically and horizontally. Intraorally we noted a narrow maxilla, bilateral posterior cross-bite, and congenitally missing maxillary primary and secondary lateral incisors. Computed tomography (CT) scans also revealed impacted upper permanent central incisors, rudimentary sinuses, a small nasopharynx, and bilateral absence of the nasal passages and nasolacrimal ducts. As mechanical retention was not possible, it was decided to rehabilitate the patient with an implant-retained nasal prosthesis.[11]

Ten years later, after failing to return for regular follow-up appointments, the patient returned to the clinic without her prosthesis. On clinical examination, the implants and surrounding tissue appeared healthy, but the patient's eyesight was severely impaired, as a result of bilateral teary eyes and cortical lens opacities (Fig. 1). Intraoral examination revealed the absence of upper incisors and inadequate plaque control (Fig. 2), while CT scans confirmed that all implants were still integrated, along with impacted upper central incisors, and absent lateral incisors, nasal passages, and maxillary sinuses (Fig. 3). To address her functional and aesthetic concerns, it was decided to surgically create and maintain new nasal passages, followed by the construction of a new implant-retained nasal prosthesis once adequate healing had occurred.

PROCEDURE

First nasal prosthesis

An implant-retained nasal prosthesis was planned for the patient due to the absence of soft tissue undercuts for mechanical retention, and a mobile supporting tissue bed precluding the use of adhesives. An alginate moulage was made of the face and a

stone cast was poured. This cast was used to produce a diagnostic wax up of the future nasal prosthesis. This trial nasal prosthesis was adjusted on the patient's face to ensure correct positioning, size and shape. It was also used to manufacture a clear acrylic resin surgical stent to guide ideal implant placement. Four titanium implants (Straumann; Straumann AG) were placed via an intra-oral approach on either side of the midline. Two were placed superiorly and two were placed inferiorly below the future nasal openings as close to each other as possible. These implants were exposed after four months and 5.5 mm long magnetic abutments were placed on the fixtures. Following a further month of soft tissue healing, a final vinyl polysiloxane impression (Reprosil; Dentsply Corp) was made of the area. An acrylic resin substructure to house the magnets was made first, which was then enclosed in the definitive silicone nasal prosthesis (Episil; Dreve Dentamid GmbH). The patient and her mom were instructed on oral and fixture hygiene procedures and proper maintenance of the prosthesis. Regular recall appointments were scheduled in advance. The patient was advised on the use of sunhats when venturing outdoors to protect against colour deterioration from sunlight exposure in a climate such as that found in South Africa.

Additive manufacturing, surgical reconstruction and second nasal prosthesis

On returning to the clinic after 10 years and having lost her first nasal prosthesis, clinical examination revealed that her eyes were very teary due to absent nasolacrimal ducts preventing adequate drainage. In order to functionally and aesthetically rehabilitate the patient, new nasal passages and nasolacrimal ducts had to be surgically created, in addition to the manufacture of a new nasal prosthesis.

A CT scan revealed a close proximity of the maxilla and palate to the cranium which posed a serious risk of intra-operative perforation or uncontrollable bleeding

(Fig. 4). To assist with pre-surgical planning and surgical stent manufacture, accurate three dimensional (3D) models were needed. This could only be done by manufacturing a physical prototype of the skull in a process known as rapid prototyping (RP). This involves the rapid manufacture of physical prototypes, as the name implies, by utilising 3D computer aided design (CAD) data. Additive manufacturing (AM) decomposes the 3D computer model data into thin cross-sectional layers which are then stacked together into 3D forms, via selective laser sintering or 3D printing.[12] Computer aided design and computer aided manufacture (CAD/CAM) of 3D models requires data acquisition, data processing and manufacturing. The data, in this case, was acquired from the patient's CT scans at 1 mm slice intervals (resolution). This data is available as DICOM files (digital imaging and communications in medicine), which needs to be converted into .stl files (standard triangulation language), to be fed into an AM machine. Engineers at the centre for rapid prototyping and manufacture (CRPM) in South Africa then imported the DICOM files into a segmentation program (Mimics; Materialise NV), which segmented the obtained image. This differentiated between bone and soft tissue to provide a clearer representation of the area of interest (Fig. 5), and produced the necessary .stl files. Two pre-operative models were produced in a polyamide material (PA2200; EOS GmbH) on a selective laser sintering machine (P385; EOS GmbH). This polyamide material is nylon based and specifically formulated to meet food and drug administration (FDA) requirements for medical use, and is available in powder form which is spread by a roller over the surface of a build cylinder of the AM machine. A piston in the build cylinder moves down one object layer thickness at a time to accommodate a new layer of powder, while a laser beam traces over the surface of tightly compacted powder, thus elevating its temperature to melting point.

The particles are fused together forming a solid mass in the form of the 3D models, an example of which is shown in Figure 6.

Surgically, it was decided to perform a Le Fort I osteotomy to create space for new nasal passages. Intra-operatively the two inferior implants were sacrificed but the remaining two were still sufficiently integrated and capable of supporting a new nasal prosthesis. The newly created nasal passages were lined with an autogenous skin graft from the thigh and packed with bismuth iodoform paraffin paste impregnated gauze (BIPP) for two weeks to maintain the graft in place during healing. A dacryocystorhinostomy procedure was also performed to create nasolacrimal ducts to allow for tears to drain from the patient's eyes into the newly created nasal passages. After healing, the BIPP was removed and a nasal stent was inserted.

Once the surrounding tissues were sufficiently healed, a final vinyl polysiloxane impression (Reprosil; Dentsply Corp) was made of the area, and a new nasal silicone prosthesis was made (Episil; Dreve Dentamid GmbH), in the same manner as the first prosthesis (Fig. 7).

There were no intra- or post-operative complications and healing was uneventful. The patient initially experienced hypernasal speech due to an increased airflow through the newly created nasal passages. She was referred for speech therapy, and showed a marked improvement within the first month. The patient was again instructed on hygiene procedures and proper maintenance of the prosthesis, which included the limitation of exposure to sunlight. Regular yearly recall appointments were scheduled, to which the patient has adhered to.

DISCUSSION

Congenital arhinia is usually associated with other facial and /or general abnormalities, and may have a genetic predisposition. The patient exhibited complete congenital arhinia as well as a lack of lacrimal sacs, small underdeveloped eyes, missing olfactory bulbs, severe class III malocclusion and mild mental retardation.

Retention and stability of facial prostheses is problematic, relying on adhesives, mechanical retention via hard and soft tissue undercuts, or attachment to mechanical devices. Adhesives could not be relied upon due to the mobility of the underlying soft tissue bed and unavailability of these products in rural settlements, and the lack of any undercuts precluded any mechanical retention. Rehabilitation was then done by an implant-retained nasal prosthesis. A major concern was that any future growth in her midfacial region would impact on the implants, but very little growth was expected in this area due to the absence of her nasal complex and the small size of her sinuses. Anterior appositional growth could lead to the implants becoming submerged, but this could be corrected with longer transcutaneous abutments. Lateral and vertical mid-facial growth could alter the position of the implants relative to each other, which is why magnetic retention was chosen over bar attachments, as these would of splinted the implants together rigidly impeding future growth and movement.

Future treatment plans for the patient include regular yearly recall appointments, and remaking the prosthesis if there is implant failure, or material and colour deterioration which is sometimes inevitable over time in a sunny climate.

Roelfs et al. (1984) stated that, “Self-image and self-perception, especially of a person’s deformities, can have vastly differing impacts on quality of life, depending on the person’s adaptability and level of acceptance, personality, other sources of

stress, and social support mechanisms".[13] However, they looked at patients with acquired and not congenital defects. The latter have never had prior experience of life with a "normal" facial appearance and may not suffer from the same feelings of loss, but may have other social and psychological problems. Clinicians need to be aware of these needs and not just focus on the provision of an anatomical substitute.

CONCLUSION

This case presents nasal rehabilitation treatment, over two phases, for a patient with complete congenital arhinia with many associated anatomical malformations. It highlights how new developments in imaging and surgical techniques were used to address many of the patient's long-standing, and previously untreated functional and aesthetic problems. It is hoped that these vast improvements will also impact positively on the patient's general emotional and social well-being.

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FIGURES

Fig. 1: Patient at 19 years of age showing healthy implants and surrounding soft tissues, and bilateral teary eyes.

Fig. 2: Intraoral view showing absent upper incisors and poor plaque control.

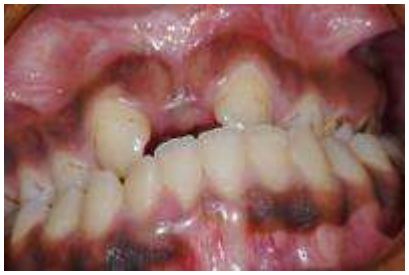

Fig. 3: CT scan noting integrated implants, impacted upper central incisors, and absent lateral incisors, nasal passages, and maxillary sinuses.

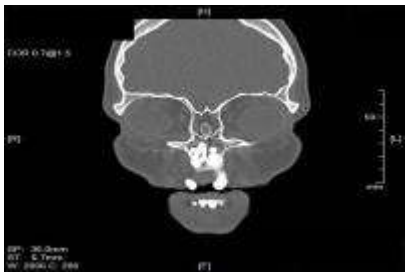

Fig. 4: Yellow arrow indicates planned nasal passage. Red arrow indicates close proximity of planned nasal passage to brain.

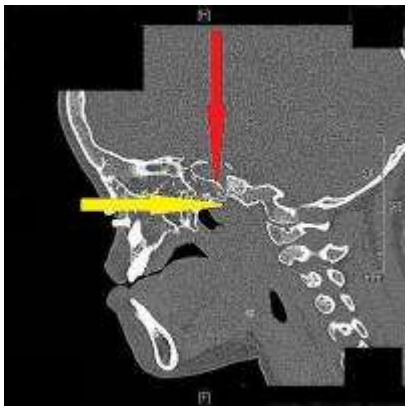

Fig. 5: 3D computer model after segementation of CT image by Mimics.

Fig. 6: Planning model verifying close proximity of palate to base of skull.

Fig. 7: Final magnetically retained silicone nasal prosthesis.