INTRODUCTION

Complete absence of the nose (arhinia) is an extremely rare congenital condition with very few cases having been reported in the literature. Many children born with this condition do not survive past infancy as the associated malformations are often not compatible with life. Rosen classified partial arhinia as that condition where the rhinencephalon is present, while in total arhinia, it is absent. Cases of partial arhinia have been described, but since all of these conditions are so rare their treatment modalities are numerous and controversial. Documentation of each patient will help further knowledge about the condition and guide clinicians towards the best treatment options for each.

The face develops from three processes: a frontonasal process which grows in the midline, and a maxillary process on either side of it. The nose develops between the third and eighth week of embryonic life beginning as a thickening of the ectoderm on both sides of the frontonasal process. These thickenings form the nasal (olfactory) placodes, which invaginate during the fifth week to form the nasal pits. Their outer edges become the lateral nasal prominences and the inner edges the medial nasal prominences. The formation of these nasal placodes develops under the inductive influence of the ventral portion of the forebrain, and thus arhinia is also often associated with absence of the rhinencephalon.

The bucco-nasal membrane then begins to descend posteriorly and marks the beginning of the nasal cavity, which consists of the nasal sacs and the anterior nostrils. The membrane descends further until the inferior portion of the nasal sacs reaches the nasopharynx. At this time the membrane then decays resulting in formation of the nasal choanae. If the septum between the nasal cavity and the nasopharynx does not decay, it will result in choanal atresia, while if the nasal sacs do not develop, the space becomes filled with unorganized tissue which eventually calcifies to form a bony plate. The nasal placodes on the frontonasal process develop into the nasal pyramid, and the nasal septum develops from the remnants of the frontonasal process which become compressed between the maxillary prominences.

Large defects of the midface present many problems for surgical reconstruction and may best be treated with a prosthesis. However good retention, and stability of the prosthesis is needed to ensure optimal functioning and to provide psychological benefits to the patient. This can be achieved by using adhesives, double-coated tapes, attachments to mechanical devices such as straps and spectacle frames, or utilizing available soft tissue undercuts. However all of these methods have limitations. Adhesives are weak, require daily removal for cleaning, provide retention for limited periods of time, may irritate the soft tissues or cause allergic reactions in sensitive patients, require good manual dexterity to replace the prosthesis in its correct position, and often cause damage and curling of the margins of the prosthesis. Mechanical devices are cumbersome and unaesthetic, and tissue undercuts may not always be available or suitably positioned for adequate retention. All of these inhibit the patient’s life style, as dislodgement of the prosthesis can easily occur - especially if the underlying soft tissues are mobile, or the patient is physically active.

The use of osseointegrated implants placed into facial bones has been advocated as a means of rehabilitating
CLINICAL REPORT

A 7-year-old female from a rural informal settlement in Mpumalanga presented with congenital total arhinia (Figure 1). A comprehensive examination was performed by a multidisciplinary team including otolaryngology and plastic surgeons, prosthodontists and occupational therapists. The patient was mentally normal and systemically healthy, but had delayed growth and cognitive skills for a child of her age. CT scans revealed rudimentary sinuses, a small nasopharynx, and bilateral absence of the nasolacrimal ducts. The medial canthi of both eyes were fissured and drooped inferiorly (coloboma palpebrae), and there was a large degree of strabismus in both eyes (Figure 2). The growth of the entire midface was retarded in both the vertical and the horizontal planes. Intra-oral examination and radiographs revealed a narrow maxilla with a bilateral posterior cross-bite and congenital absence of the maxillary primary and secondary lateral incisors. The patient had undergone a tracheotomy procedure at birth. The ostium was still patent and functioning well, even though it was very small. It was decided that surgical reconstruction would not be possible and that the most suitable treatment option for her would be to fabricate a nasal prosthesis.

Mechanical retention was not possible as there were no available soft tissue undercuts to help retain a prosthesis. Adhesives were deemed inappropriate as the supporting tissue bed was mobile especially when she spoke, smiled or chewed, and retention via spectacle frames was impossible due to the total absence of a nasal bridge, and the inferior position of her ears relative to her eyes. It was decided to provide the patient with a nasal prosthesis retained by extra-oral osseointegrated implants. A moulage impression of the face was taken using an irreversible hydrocolloid (PG.S. Alginate, Millners Dental Suppliers, Pty Ltd) which was backed by a layer of fast setting dental stone to prevent distortion before it was removed (Plastogum, Harry J Bosworth Co. Skokie, Illinois). A cast was poured and used to produce a diagnostic wax up of the future nasal prosthesis. The trial nose was adjusted on her face to ensure it sat in the correct position and had an aesthetically pleasing size and shape. This was then used to plan the desired implant position, and to fabricate a clear acrylic resin surgical stent (Figure 3). Surgery was performed via an intra-oral incision. It was anticipated to place three Straumann extra-oral implants in a triangular configuration in the maxilla. However at the time of surgery a midline suture was discovered which prohibited placement of the superior implant in the midline. This problem was overcome by using two implants, one on either side of the suture but as close to each other as possible. Thus a total of four implants were placed: two superiorly on either side of the midline, and two inferiorly below the future nasal openings. Four months later the implants were exposed and magnetic abutments (5.5mm long) were fitted to the fixtures. At this time it was also necessary to thin the subcutaneous tissues. The soft tissue was allowed to heal for a month; thereafter the final impression was taken using a vinyl polysiloxane material (Reporsil, Regular body, Dentsply Int.) An acrylic resin substructure which encased the magnet attachments was first fabricated (Dental ventures of America, Inc., Anaheim Hills, Calif manufacturer), and then embedded in the definitive silicone nasal prosthesis (Episil, Dreve Dentamid, Germany). An acrylic resin layer of fast setting dental stone to prevent distortion before it was removed (Plastogum, Harry J Bosworth Co. Skokie, Illinois). A cast was poured and used to produce a diagnostic wax up of the future nasal prosthesis. The trial nose was adjusted on her face to ensure it sat in the correct position and had an aesthetically pleasing size and shape. This was then used to plan the desired implant position, and to fabricate a clear acrylic resin surgical stent (Figure 3). Surgery was performed via an intra-oral incision. It was anticipated to place three Straumann extra-oral implants in a triangular configuration in the maxilla. However at the time of surgery a midline suture was discovered which prohibited placement of the superior implant in the midline. This problem was overcome by using two implants, one on either side of the suture but as close to each other as possible. Thus a total of four implants were placed: two superiorly on either side of the midline, and two inferiorly below the future nasal openings. Four months later the implants were exposed and magnetic abutments (5.5mm long) were fitted to the fixtures. At this time it was also necessary to thin the subcutaneous tissues. The soft tissue was allowed to heal for a month; thereafter the final impression was taken using a vinyl polysiloxane material (Reporsil, Regular body, Dentsply Int.) An acrylic resin substructure which encased the magnet attachments was first fabricated (Dental ventures of America, Inc., Anaheim Hills, Calif manufacturer), and then embedded in the definitive silicone nasal prosthesis (Episil, Dreve Dentamid, Germany). On delivery, the patient was instructed on hygiene procedures and maintenance of her prosthesis. Regular recall appointments were scheduled (Figures 4 & 5).

Six months later she returned with inflammation and hyperplasia of the skin around the inferior abutment on the left side. This was found to be due to a small piece of metal that had stuck to the magnet and acted as a spacer allowing an overgrowth of skin under the prosthesis. The hyperplastic skin was removed and at the same time a small cuff of resin was placed around the remaining magnets to prevent fu-
that will be used; planning the treatment sequence and manufacturing stages; the anticipated types, strength, frequency and direction of dislodging forces that will be imposed upon it, other areas available for retention and support, its short and long term functional requirements; the patient’s age, activity levels, financial status, dexterity and ability to maintain hygiene; and psychological status. In this patient the use of adhesive or any of the routine forms of mechanical retention were all unsuitable and it was decided to use osseointegrated implants to retain her nasal prosthesis. As the tissues in the midface region were mobile during function, magnetic abutments were chosen as these allow the prosthesis to easily re-attach as soon as the dislodging forces are removed. However, magnets must have differing orientations so as to be able to withstand forces directed from many different planes. They are also easier to access for hygiene and cleaning purposes.

In light of the child’s age there was concern that future bone growth in the midface region could impact on the implants – although very little was expected due to the absence of the nasal complex and the small size of her sinuses. Appositional growth in an anterior direction could lead to the implants becoming submerged. If this occurs, new longer transcutaneous abutments will be needed. However the possibility that they could become too deeply submerged must also be foreseen. This would necessitate their removal and replacement with new fixtures. Future lateral and vertical mid-facial growth may also occur which would alter the implants position relative to each other. This was another reason why it was decided to opt for magnetic retention rather than using bar attachments which would have splinted the implants together rigidly, impeding growth and movement. Disadvantages of magnets are that they require increase in bulk due to the substructure which may be a problem where there is limited space available, and problems with corrosion have also been reported.

Future treatment plans for the patient include: adaptation / remaking the prosthesis in accordance with growth of the rest of her face, or due to the inevitable material and colour deterioration that will occur over time; orthodontics to correct the cross bite; maxillofacial surgery to perform distraction-osteogenesis at the sutura if there is not sufficient growth of her mid-face; and plastic surgery to reconstruct the canthi of the eyes and the nasolacrimal ducts.

CONCLUSION

In this patient none of the regular mechanical retentive mechanisms were possible and osseointegrated implants were deemed to be the most suitable treatment option. Magnetic attachments were chosen as they allow easy placement and removal of the prosthesis. They are easier to clean than when adhesives are used, can be orientated in various planes to withstand dislodging forces from many directions, and they do not splint the implants together rigidly - especially in growing children. In addition, as the patient was from a rural area, adhesives were not suitable as she would not have been able to purchase these when needed, and they reduce the life-span of the prosthesis which would necessitate more frequent remakes. Success of the implant-retained prosthesis depends on patient motivation as well as careful design to ensure good access and visualization for hygiene purposes.

Declaration: No conflict of interest was declared

REFERENCES


Additional references (8-35) are available on
www.sada.co.za