Case study

Reconstruction of nongrowing hemifacial microsomia patient with custom-made unilateral temporomandibular joint total joint prosthesis and orthognathic surgery

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ABSTRACT

A case of hemifacial microsomia in a young male is presented. The ascending ramus and condyle was reconstructed utilizing virtual 3D planning with a custom-made total TMJ device (TMJ Concepts, USA) in combination with conventional orthognathic surgery. The alternative available reconstructive options are discussed and the advantages and disadvantages of the technique selected.

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1. Introduction

Hemifacial microsomia (HFM) is a complex congenital disorder, meaning it typically begins at birth or in the first months of life. HFM is essentially an abnormality of development of the embryonic first and second branchial arches and is the second most common craniofacial birth defect after cleft lip and palate. It occurs in approximately in 1:5000 or 1:6000 live births and displays a wide spectrum of abnormalities.¹ The disorder primarily affects the development of the lower part of the face, including the ears, mouth, and jaw area. It is usually unilateral and always asymmetrical. Synonyms, which are also in common usage, are craniofacial microsomia, oculo-auriculo-vertebral-dysplasia or facial-oculo-auriculo vertebral dysplasia. Goldenhar syndrome is a term liked by pediatricians but only really refers to a small sub group of HFM patients.

HFM occurs sporadically in most cases and can be considered a nonspecific symptom complex that is etiologically and pathogenically heterogeneous. Extreme variability of the expression is characteristic for the disorder.² The classification of HFM by Kaban identifies type I as a small mandible with normal temporomandibular joint (TMJ) morphology; type IIa as a ramus with abnormal

size and shape; type IIb as a ramus and TMJ with abnormal size, shape, and function; and type III as an absent condyle, absent ramus, and absent TMJ.³ This classification system may be the most useful to the surgeon in the preoperative evaluation because of its simplicity and inclusion of the TMJ anatomy and function.

A case of hemifacial microsomia is presented in a young male where a custom-made TMJ prosthesis was used along with orthognathic surgery to correct the facial asymmetry.

2. Case report

A 19-year old male suffering from left sided hemifacial microsomia was referred to the author's clinic, in 2014 for evaluation and treatment of his condition (Fig. 1a–c). Clinical and radiographical investigation revealed multiple skin tags, a HFM type IIb mandibular defect on the left side, deviated chin to the left and occlusal cant. The seventh cranial nerve was intact.

Treatment included orthodontic and surgical intervention. In order to reconstruct the mandibular defect on the left side a custom made TMJ prosthesis was planned along with conventional orthognathic surgery to correct the occlusal cant.

Orthodontic treatment was accomplished by means of bimaxillary fixed appliances for a period of 1.5 years. Once the presurgical orthodontic treatment was finished, a new CT scan with 0.625 mm slice thickness was obtained and sent to medical modeling (Colorado, USA) along with final dental casts set in to best possible occlusion. CT scan was segmented, and a 3D virtual skull model

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Fig. 1. (a) CT scan of the patient upon completion of orthodontic treatment and before surgery. (b) CT scan of the patient upon completion of orthodontic treatment and before surgery. (c) Clinical photo of the patient prior to surgery.

Fig. 2. Correction of the occlusal cant by means of Le Fort I osteotomy and contralateral sagittal split osteotomy.

was created. The occlusal cant was corrected by means of Le Fort I osteotomy and the mandible was advanced and set into final position by means of sagittal split osteotomy on the right side. Intermediate and final splints were fabricated based on the virtual planning (Fig. 2). These data were then sent to TMJ Concepts (Ventura, CA, USA) for planning of the custom made total TMJ prosthesis. Since the available bone stock in the ramus could not allow for any fixation because of the vicinity to the mandibular canal, an extended prosthesis was manufactured where the mentum was used for fixation of the prosthesis. A 3D steriolithographic model was created and sent to the surgeon for removal of any bony interference. Once the surgeon approved the 3D model, the prosthesis was waxed up on this model and manufactured subsequently. A hole was made in the condyle for a vertical suspension suture (Fig. 3).

In September 2015, the patient was taken to surgery. A right sagittal split ramus osteotomy was performed and the mandible mobilized and set into intermediate splint with IMF. Fixation on the right side was achieved by means of three bicortical screws which were instrumented transbuccally via a trocar. The incision was closed with 3/0 Vicryl running sutures. The oral cavity was sealed with Tegaderm (3M, Sweden). A different set of instruments was used to approach the left TMJ and submental area. A Left preauricular and a submental incision was performed and a tunnel was created between the two incisions by blunt dissection. The fossa part was secured to the zygomatic arch by multiple 2.0 mm

Fig. 3. Actual prosthesis with a hole made in the condylar head for a vertical suspension suture.

screws. The ramus part was placed through the submental incision, seated in the fossa and secured to the mentum by means of several 2.0 mm bicortical screws. A 2/0 PDS Vicryl suture secured the condyle to one of the fixation screws on the fossa part (Fig. 4). The incisions were closed in layers and no drain was used. At this stage the intermediate splint was removed and a Le Fort I osteotomy was performed. The maxilla was set into final occlusion with aid of the final splint and secured by means of 4 titanium plates (Synthes, Orthognathics) and multiple screws. IMF was released, occlusion was checked and the incision was closed with running sutures. Recovery was uneventful and the patient was discharged 48 h after surgery. A CT scan was performed postoperatively and confirmed adequate placement of the prosthesis. Sutures were removed a week later, guiding elastics were applied and the patient was advised on liquids and soft diet only.

At 4 weeks check-up there was obvious granulation tissue along the sagittal split incision on the right side and there was a mismatch in the occlusion. Mobility of the fragments was confirmed clinically and orthopantomogram revealed rotation of the proximal fragment on the right side. The patient was taken to surgery again and the incision was opened and debrided. Loos screws were removed and after IMF, fixation was achieved with a 4-hole Synthes Trauma 2.0 titanium plate (Synthes, USA).

Fig. 4. Fossa part fixated to the zygomatic arch. Ramus part is seated in the fossa. Note the suspension suture secured to one fixation screw on the fossa part.

Unfortunately this fixation showed to be inadequate since the patient needed another surgery just 2 weeks later. At this point, a submandibular incision was used to access the area and to debride it thoroughly. All loos bony fragments along with second and 3rd molars, which were deemed to be possible source of infection, were removed. The intraoral wound was closed in a watertight manner. IMF was achieved and a 2.5 mm reconstruction plate (Synthes, USA) was applied to the mandible and secured with multiple bi-cortical screws. The incision was closed in layers and again no drain was used. Healing was uneventful at this time. Upon 6 months check up, there was excellent occlusion and adequate symmetry, the scars were almost inconspicuous and maximum incisor opening measured 40 mm (Fig. 5a and b).

3. Discussion

The use of virtual surgical planning and computer-aided design/computer aided manufacturing has previously been reported to enhance the planning, increase accuracy and to reduce surgical time in a reconstruction case.

Treatment of hemifacial microsomia depends on the classification of the deformity, the severity of the deformity, the age of the patient and the wishes of the patient and their family.

It is rarely necessary to consider any surgery for the Type I mandible in childhood. Treatment of the deformity is generally going to be deferred to maturity and is likely to consist of either no surgery, genioplasty or conventional orthognathic surgery. Also type IIa can be treated by means of conventional osteotomies with or without grafting or distraction osteogenesis.³

Depending on the severity of the deformity and the wishes of the family, distraction osteogenesis can be considered to lengthen the mandibular ramus from the age of 3 or 4 years. However, any child who receives such lengthening during growth is likely to need further intervention over time.³

In type IIb and III HFM mandible, the ascending ramus, condyle, disc, and glenoid fossa are underdeveloped or may be absent. The aim of surgery in this case was to reconstruct the glenoid fossa, condylar head and part of the ramus, and to re-establish the horizontal and vertical mandibular deficiency, mouth opening, masticatory function and morphology.

Numerous reconstructive techniques have been described in the treatment of type IIb and III of HFM.^{4–6} Autogenous tissue grafts have been used for the treatment of ipsilateral mandibular hypoplasia, including costochondral, sternoclavicular, posterior and anterior iliac crest, free fibula, and second metatarsal.^{4–6} The costochondral graft has been popular for the reconstruction of the ipsilateral TMJ, particularly in young patients with types IIb and III

Fig. 5. (a) 6 Months postoperative frontal view. (b) 6 Months postoperative lateral view.

HFM owing to the growth center at the costocondral junction and for reconstruction of the glenoid fossa.⁷ However other studies show unpredictable growth of the graft creating a deformity necessitating another surgical procedure.^{8,9} In addition to the potential problem of overgrowth or no growth, these autogenous grafts (ribs, sternoclavicular grafts, or other bone grafts) when used in growing or nongrowing patients with HFM are subject to biophysiologic changes from functional loading (particularly with significant mandibular advancement due to counterclockwise rotation), creating unpredictable results related to long-term skeletal and occlusal stability because the grafts may bend, flex, fracture, and resorb. The workhorse for microvascular reconstruction of the mandible is the free fibula. The fibula free flap approach allows the possibility of using bone with/without skin for restoring a large defect. It has the advantages of consistent shape, ample length and distant location to allow a two-team approach. One major draw back of the fibula is it's restriction in terms of 3D shaping and also transfer of a fibula graft vastly increases surgery time and morbidity for the patient.

In this patient, the entire TMJ and part of the ramus had to be reconstructed. In addition the mandible had to be advanced and elongated on the ipsilateral side in order to correct the occlusal cant. In order to achieve all above we opted for an alloplastic reconstruction. Prosthetic TMJ reconstruction in cases with major mandibular defects or other abnormal anatomy usually requires custom-made devices. The TMJC (TMJ Concepts INC, Ventrura, CA, USA) device is a computer-assisted designed and manufactured

(CAD CAM) prosthesis constructed on a stereolithic three-dimensional (3D) model fabricated from the patient's computed tomography (CT) scan data. The fossa component consists of an ultra-high molecular weight polyethylene (UHMWPE) articulating surface and the ramus component is made of a machined alloyed titanium with a condylar head of chrome-cobalt-molybdenum (Cr-Co-Mb).¹⁰ Most TMJC devices installed over the years have been straight forward local TMJ reconstructions but more recently, they have been used to bridge increasingly large mandibular defects like the one presented here.^{11,12} There is theoretically a risk of condylar sag when using such a big piece of alloplastic device, especially in congenital deformity cases where the pterygo-masseteric sling is totally or partially absent. A modification which nowadays is standard in all extended TMJC devices is a smooth edged hole that has been introduced just inferior to the condylar head. The hole can be placed either medial to lateral or anterior to posterior depending upon the surgeon's preference. This allows for a vertical suspension suture to maintain the vertical position until the scar tissue is mature enough to support the mandible. We used 2/0 PDS suture, which was secured around one of the fixation screws of the fossa component.

Multiple surgeries were needed in order to stabilize the contralateral sagittal split osteotomy in this patient. We believe that this might be due to a combination of non-compliance by the patient as well as insufficient fixation. When the sagittal split was fixated with a load bearing plate, there were no further problems with healing.

Although successful in this patient, with increasing length and volume of the mandibular components it is likely that the risk of fatigue fracture will increase and further studies are needed to reveal this.

4. Conclusion

Alloplastic custom-made joints avoid donor site morbidity. When combined with virtual planning, they allow for a more precise anatomic and esthetic reconstruction and reduce operative time. The major disadvantages include the unknown life expectancy of the devices, loosening of the implant, high expense and limited availability.

Conflicts of interest

The author has none to declare.

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