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#### Abstract

First Arch syndrome or hemifacial microsomia one of the common patients of craniofacial deformity and malformation. The aetiology as known was genetic and hereditary factor. But the accepted theory as early occlusion of stab pedal artery of the first arch are acceptable.

Clinically the disease featured in three forms milled, moderate and severe form of the disease. The milled form appeared with underdeveloped mandible and presence of tags in the preauricular area as remnant of Meckel's cartilage, the moderate type featured as cleft in the angle of the mouth, underdeveloped masseter muscle, missing condyle, ear deformity and deficiency in growth of the mandible and midface.

The severe form of the disease is very wide cleft of the angle of the mouth with partial or complete absence of ear, missing of Temporomandibular joint (TMJ), missing of glenoid fossa and zygomatic root of temporal bone, with missing of upper part of ascending ramus of the mandible, with underdeveloped mandible and midface.

Kummoona chondro-osseous graft from iliac crest was used for reconstruction of the TMJ in children for restoring growth of the mandible and midface after correction of angle of the mouth by commissuroplasty and cartilage graft for building the glenoid fossa and zygomatic root of temporal bone by bone graft and platysma muscle flap for augmentation or building the atrophied masseter muscle before the cartilage bone graft building, this surgery should be carried out before the child go to school. Series of cases revealed success were reported.

Keyword: Kummoona; Chondro-Osseous Graft; TMJ; First Arch Syndrome

## Introduction

One of the common cases with facial malformation and deformities were reported are the First Arch dysplasia syndrome or nominated as hemifacial microsomia occurred as result of disturbance in growth of the 1<sup>st</sup> and 2<sup>nd</sup> branchial arches during embryonic development due to early occlusion of step dial artery. The hereditary and genetic factors should not be excluded.

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Other theories believed that first arch syndrome is congenital, and the defects caused by neural crest failure to migrate to form the first pharyngeal arches. It is well known that incidence was about 1/700 of new borne babies might be similar with cleft lip and palate incidence with variations of races.

There are three clinical features of first arch syndrome and they are either mild or moderate or severe.

The mild form appeared as underdeveloped mandible and overlying muscle with preauricular tags, the moderate type of the disease was featured with underdeveloped ear, mandible with overlying soft tissue, hypoplasia of the TMJ and microsomia.

The severe form of the disease featured by absence of the ear and glenoid fossa and temporomandibular joint (TMJ), zygomatic arch, missing of upper part of ascending ramus, with underdeveloped mandible, overlying soft tissue and severe microsomia.

The absence of the TMJ with missing condyle, was a problem facing all Craniomaxillofacial and plastic and reconstructive surgeons, the condyle represents and considered as growth centre for growth of the mandible and midface. The other theory is the facial growth was a demand for periosteal matrix of the facial skeleton as advocated by Moss, 1960.

There are two substitutes for restoring the growth of the mandible and midface either by reconstruction of the TMJ by costa-chondral graft or by Kummoona chondro-osseous graft.

The costa-chondral graft was practiced early by Sir Harold Gillis and Kennett, 1970 but David Poswillo did an experimental study 1974 on Me caca Iris Monkeys to demonstrate and prove that the Costa-Chondral graft can play an important role for restoring the growth of the condyle and mandible. It was a revolutionary work for Poswillo and the graft was extremely popular for all surgeons for reconstruction of the TMJ during the last 5 decades.

Kummoona advocated the chondro-osseous graft for reconstruction of the TMJ in 1986, this graft was more popular than costa-chondral graft because this graft can restore growth, remodelling and repair of the TMJ beside other favourite factors.

## **Case Study and Results**

There are great varieties of congenital and developmental and male formed syndromes effecting the face. The face represents the mirror of the body and its charisma featuring estimableness of both women's and men's and intelligence or dullness. People now a days very much concerned how they look to attract each other. The sadness of a family with new borne baby borne with such a deformity, it is a tragedy to them.

The managements of First Arch syndrome were not an easy task and required an experienced surgeon with a lot of knowledge and skill. There is no single technique to cover the managements of these cases with deformed half of the face but required a series of operations in stages started from childhood to after puberty.

Kummoona Platysma muscle flap which consist only of muscle was advocated for reconstruction of the atrophied or underdeveloped masseter muscle in hemifacial microsomia or first arch dysplasia syndrome [4].

The milled form of first arch showing underdeveloped mandible with absence or deformed condyle and underdeveloped masseter muscle, sometime the chin slightly twisted to the affected side, the moderate types of this disease or deformity showing microsomia and slight cleft of the angle of the mouth and slight deformity of the ear or presence of tags of cartilage in preauricular area. These tags may be a remanent of Meckel's cartilage with absence of the temporomandibular joint (TMJ) and missing of the upper part of ascending ramus of the mandible.

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The severe form of this first arch deformity is characterised by cleft of the angle of the mouth, absence of the ear or missing the helix and Anti helix, missing glenoid fossa, missing condyle and TMJ also zygomatic root of temporal bone and the mandible showing severe twisting to the affected side and underdeveloped midface [4].

The author advocated his own technique for reconstruction of helix and Anti helix of the ear by his flap Kummoona auricular-cervical flap we been able for reconstruction of missing of part of the ear in congenital or traumatic injuries and even in cancer cases was affected in this part of the ear [5].

We advocate many surgical techniques for managements of these cases during the last five decades. In mild cases less complicated surgery required but the moderate cases were planned first to remove the tags and remnants of Meckel's cartilages in preauricular area with commissuroplasty for repair and reconstruct microsomia and angle of the mouth and transfer platysma muscle flap for reconstruction of atrophied masseter muscle.

The second stage is by reconstruction of glenoid fossa by cartilage from the ear and reconstruction of missing zygomatic arch by piece of bone from the edge of iliac crest.

The third stage is by reconstruction of the TMJ by chondral-osseous graft harvested from iliac crest of 1 cm width and 4 - 5 cm length consist from cup of cartilage, 1 cm and an osseous element of about 3 - 4 cm fixed to the outer border of the ramus after good decortication of the graft and ascending ramus and the graft head should be in the new glenoid fossa, fixation was done by 3 holes made along the graft and ascending ramus by soft stainless steel wire of 0.5 mm. After 3 - 4 days we usually ask the child to eat soft food and gradually to normal masticatory process to enhance the growth potential of the graft and facial skeleton. The last stage is by reconstruction of the ear.

These surgical procedures should be done in the age of 5 - 6 years before the child attending school and the procedures should be done every 5 - 6 months between stages.

Alignment of teeth irregularities should be managed by orthodontic treatment after eruption of the permanent teeth and further surgery for correction of jaw relationship by an osteotomy after puberty.

Kummoona chondro-osseous graft proved to be an excellent substitute to the TMJ and more widely acceptable graft because the cartilage cap is rigidly attached to osseous element and the graft was rigidly attached to the mandible while the cartilage of the Costa-Chondral graft easily dislodged from the rib and the patients had intermaxillary (IMF) fixation for few weeks ends with spasm of muscles of mastication and unable to chew. The patients who were treated by Kummoona chondro-osseous graft were advised to chew food after few days [3].

Through experimental studies were done on rabbits by reconstruction of the TMJ by our technique of grafting and to prove the condyle contain growth centre.

Post-mortem and histological studies, this graft contains mesenchymal stem cells in the second granular layers of the graft and the graft has the potential to grow and to restore function and normal height of the face and further to that the graft can grow, repair and remodelling of the TMJ.

The histological examination of the graft showed 4 zones, the first layer contain a dense layer of fibrocartilage due to hard masticatory process of hard Rabbit food, the second layer showed several zones of active round cells of mesenchymal stem cells which represent the proliferator layer and the third layer showed a series of hypertrophic chondrocyte passed through series of changes with presence of an osteoid tissue, the third layer represent the differentiations of mesenchymal stem cells to chondrocyte and osteoblast, these changes represent the growth potential of the graft.

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The fourth layer was showing an osteoid tissue and bony trabeculae and bone marrow spaces in between.

Further experiment was done on rabbits to study the cellular changes that occurred in bone graft healing, we found after clot formation and platelets aggregation, platelets growth factor released (PGF) and mesenchymal stem cells from bone marrow, periosteum and overlying muscle and formation of large number of fibroblasts and osteoid tissue.

Recent studies on connective tissue for cell differentiation found the value of chondro-osseous junction of the graft to maintain growth, repair and remodelling of the graft due to intrinsic potential and to the presence of mesenchymal stem cells during endochondral bone formation in the graft and chondrocyte cells undergoes differentiation towards hypertrophy before they replaced by bone and bone marrow [2,6].

In our studies of Kummoona chondro-osseous graft they did found that a G-protein coupled receptor (CXCR4) predominantly expressed in hypertrophic chondrocyte, while its ligand chemokine stromal cells derived factor (SDF-1) is expressed in the bone marrow adjacent to hypertrophic chondrocyte.

By this research we proved the viability of our Chondral-Osseous graft and the most popular techniques and widely used graft for reconstruction of the TMJ.

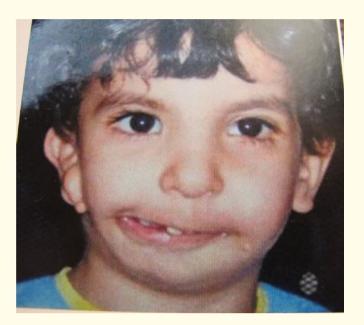
The first primary report of Kummoona chondro-osseous graft was published 1986 [1].

Series of cases were treated by this technique, figure 1 and 2 during the last 4 decades.



*Figure 1A:* Photograph of girl of 8 moths with First Arch syndrome of the right side of the face treated by another surgeon using a primitive distractor.

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*Figure 1B:* Photo of the same girl at age of 3 years showing deformity of the right side of the face with microsomia of the angle of the mouth.



*Figure 1C:* CT scans of three dimension showing sever deformity of the facial skeleton, showing missing of the glenoid fossa, zygomatic root of temporal bone, missing of temporomandibular joint (TMJ) and upper part of ascending ramus of the body of the mandible.

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Figure 1D: Photo showing Kummoona Platysma flap transferred for reconstruction of atrophied masseter muscle.



Figure 1E: Post-operative photo of the same child at age of 7 years after series of operations carried out for correction of First Arch syndrome of this child.

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Figure 2A: Photo of a child boy of 4 years with moderate type of First Arch syndrome showing tags in preauricular area with atrophy of masseter muscle and sever microsomia of the angle of the mouth.



*Figure 2B:* Post-operative photo after series of operations were carried out for correction of First Arch syndrome, this photo was taken after two years.

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## Conclusion

First arch syndrome was faced great difficulties for reconstruction and required an expert surgeon with great knowledge to carry a series of operations in different stages for application of platysma muscle flap for reconstruction of masseter muscle, Chondro-Osseous graft for reconstruction of the TMJ and auricular cervical flap for reconstruction of Helix and pre-Helix of the ear beside bone graft to zygomatic root of temporal bone and cartilage graft to build the glenoid fossa.

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