CASE REPORT

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PEDIATRIC CRANIOFACIAL DISFIGUREMENT WITH AUTOLOGOUS RIBS BONE GRAFT RECONSTRUCTION



Anne Saputra,^{1*} Wayan Niryana,² Made Suka Adnyana³

ABSTRACT

Pediatric reconstruction of the cranial defect is a challenging task, the standard reconstruction method has been bone grafting. The reconstruction of complex facial defects should satisfy both aesthetic and functional requirements. In the case of large defects, the use of craniofacial prostheses using autogenous bone is the material of choice because of its potential for revascularization and its osteoconductive properties.

A 3-year-old patient has facial disfigurement as result from bone deficiency following anterior skull base tumour resection. To minimize the associated functional and cosmetic problems, a number of reconstructive options are available to the surgeon including the use of autogenous and alloplastic implants. A computed tomography (CT) 3-dimensional reconstruction scan showed a large craniofacial defect as residual radical skull base tumour resection. A transcranial approach by a neurosurgeon and plastic surgery was performed to reconstruct the defect using autologous rib bone graft. Six months after the reconstruction surgery, a defect of the craniofacial was narrowing without cranial nerve deficits.

Complex reconstructions of extensive defects in craniofacial area can be achieved using autologous bone grafts. They yield reasonable functional and aesthetic outcomes and noticeably improves the quality of life.

Keyword: reconstructive surgery, autologous bone graft, craniofacial defect

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INTRODUCTION

Carcinomas of the ethmoid, frontal, or maxillary sinuses sometimes invade the anterior skull base. It is necessary to perform en-bloc resection for this invasive carcinoma according to the concepts of surgical treatment for head and neck cancer. The anterior skull base consists of two parts, the orbital roof as the lateral portion and the roofs of the frontal sinus, ethmoid sinus, and/or sphenoid sinus as the central portion. Selective reconstructive options for the anterior skull base. A dura defect is repaired by a fascia lata or a pericranial flap. After the dura has been tacked up, reconstruction of the anterior skull base is performed simultaneously with augmentation of the defect of extracranial structures.

The anterior skull base is a common location of many intradural and extradural cranial and/or facial pathologies. The transnasal approach was first applied to resect a large frontal meningioma involved with the ethmoid sinus.¹ Since the introduction of this approach, numerous modifications have been made, aiming to lessen brain retraction, widen exposure of the tumours and better functional and cosmetic outcomes.² The development of skull base surgical approaches has improved the treatment of malignant tumours and other lesions of the anterior skull base and allowed successful resection of many tumours once considered inoperable. As a result, a remarkable increase in the survival rates of the patients with malignant tumours of the anterior skull base is achieved after proper modern surgical treatment. Also, appropriate reconstruction and isolation of the anterior cranial fossa from the contaminated areas is a critical surgical step for the prevention of ascending infection, thus, decreasing the rates of the postoperative morbidity and mortality.³

Craniofacial reconstruction (CFR) of complex facial defects should satisfy both aesthetic and functional requirements. CFR with implants varies significantly based on the cause of the surrounding anatomical structures and function. The properties of such implants are tied closely to the material(s) and method(s) used to construct them. Desired properties such as biocompatibility, bioactivity, toxicity, yield strength, flexural modulus, implantation complexity, and infection risk are all defined by the choice of material and, as such, implant material consideration remains a key factor in cranial and craniofacial reconstruction.⁴ Of these properties, infection remains a core concern in regards to implant failure and patient health.⁵

The most common alloplastic implants include titanium bone plates and screws, which though well tolerated, are associated with delayed complications including implant extrusion or fracture.⁶ Sources of

¹General Surgery Resident Faculty of Medicine Udayana University, Sanglah General Hospital Denpasar, Bali, Indonesia

²Department of Neurosurgery Faculty of Medicine Udayana University, Sanglah General Hospital Denpasar, Bali, Indonesia ³Department of Plastic Surgery and Reconstruction Faculty of Medicine Udayana University, Sanglah General Hospital Denpasar, Bali, Indonesia.

*Correspondence to: Anne Saputra, Department of Neurosurgery, Udayana University -Sanglah General Hospital, Kesehatan St. 1, Denpasar-Bali, Indonesia 80114. anne.saputra@gmail.com free non-vascularised bone grafts include calvarium, rib, ilium, tibia, fibula, scapula, and radius.7 Their usefulness has, however, been limited by early bone resorption and infection.8 Vascularised grafts are now the state-of-the-art for bone replacement in the craniofacial region,⁹ as they are reliable, resistant to radiation and infection, and allow the placement of dental implants. Their disadvantages include high cost, the need for specialized training and equipment as well as significant donor site morbidity.10

CASE REPORT

A 3-year-old girl presented with facial disfigurement as result from bone deficiency following anterior skull base spindle cell tumour resection. The girl was otherwise healthy, without any motoric and sensory deformities. A 3-dimensional computed tomography (CT) scan reconstruction showed a large complex defect on the fronto-orbital roof. The cardiovascular, gastrointestinal, and central nervous systems were normal. Liver function,

kidney function, and hemostatic tests were normal, without associated congenital anomalies.

Three months after resection of an anterior skull base spindle cell tumour, the brain CT scan showed a large defect on the fronto-orbital without any residual mass tumour. A frontal approach by a neurosurgeon was performed to excise the tumour and close the dura mater. The bone defect on the fronto-orbital roof and frontal calvaria continued with gradual craniofacial reconstruction using



Figure 3 Skull base spindle cell tumour (before resection)



Clinical before resection anterior skull

Figure 1 base spindle cell tumour



Figure 2

Facial disfigurement after skull base tumour resection



Skull base tumour (post resection) Figure 4





Facial disfigurement with CT scan 3D Figure 5 reconstruction (before reconstruction)



Figure 6 The defect of orbital roof



Figure 7 Autologous ribs bone graft

autologous ribs graft which fixates by surgical miniplate and wire.

Six months after the initial surgery, a defect of the fronto-orbital roof was narrowing and without cranial nerve deficits.

DISCUSSION

Extended defects in the midfacial area may result from tumour resections and less frequently from severe trauma.¹¹ Malignancies in the frontal sinus, in particular, are often detected at a late stage; therefore, tumour infiltration of adjacent structures, such as the orbit, ethmoidal cells, and the outer skin, is common at the time of diagnosis.¹² The resection of these tumours may result in extended defects with broad connections between the orbit and frontal sinus.

Reconstructions for these defects and the rehabilitation of patients require the use of various technical modalities. The primary goals are a separation of the functional units and obtaining the best possible restoration of the facial appearance.¹³ From a pathological point of view, a two-stage





(a). Ribs bend to be a superaorbital roof,(b). Fixate with miniplate and screw,(c). parts of ribs graft to the reconstruction of calvaria and applied with bone cement artificial

reconstruction is more reasonable for most tumour patients because the bony and complex resection margins in the midface do not allow frozen section diagnosis.¹⁴ On the other hand, large defects in the midface sometimes require immediate reconstruction because anatomical vital structures cannot be left uncovered.

Historically, the first clinical trial of a bone graft was reported in 1670 when a xenograft, canine bone, was used to repair a skull defect in a peasant in Russian.¹⁵ Bone graft has been used in craniofacial reconstructive surgery in a variety of ways and in a multitude of locations. Most surgeons have favourite techniques for inlay or on-lay grafting. Inlay bone graft is useful in osteotomies because they demonstrate little resorption. Autogenous bone graft to fill osteotomy sites and to promote early consolidation are routinely utilized by most surgeons.

Bone grafting plays a central role in craniofacial surgery, in both reconstructive and aesthetic realms. Bone grafts are used to fill bony defects, impart structural support, and augment deficient projection in the craniofacial skeleton. Although commonly thought of as a static entity, bone is in a state of dynamic equilibrium and has the ability to regenerate, remodel, and replace itself. Unlike most tissues, bone has the potential to heal with virtually no scars. These unique properties make bone wellsuited for grafting applications.



Figure 9 Follow up CT scan 3D, 6 months post reconstruction

Although nonvascularized autogenous bone grafts (i.e., free grafts) are the gold standard for craniofacial reconstruction, recent attention has been directed at vascularized grafts, allografts, bone substitutes, and osteoinductive factors in grafting applications. The use of free non-vascularised bone grafts to replace large segments of facial bone has been superseded by microvascular techniques in developed countries where such skills and facilities are readily available and outcomes are more predictable.¹⁰ Vascularised grafts are less likely to get infected or resorb as compared with non-vascularised ones. In developing countries, however, the older reconstructive techniques still have a place provided steps are taken to minimize graft failure.

The surgeons' preference for different grafting procedures in reconstructing many head and neck defects and deformities of variable aetiology depends on many factors such as the age of the patient, nature of the defects; with their consequent, structural, functional, and cosmetic effects; available resources, and personal experience and training. Despite significant advances in biomedical engineering, the perfect graft material has to be attained. The rib grafts are among the sources of free non-vascularised bone and cartilage grafting materials that have versatility in craniomaxillofacial reconstruction.

For the reconstruction of extensive craniofacial bony defects, autogenous bone is the material of choice because of its potential for revascularization and it has osteoconductive and osteoinductive properties.¹⁶ It can be used as corticocancellous blocks, compressed particulate marrows, cortical grafts, or free flaps. Donor site choice is strictly dependent on defect size and localization from general conditions of the patient and from potential morbidity at the donor site. Donor sites include ilium, tibia, rib, mandibular symphysis, maxillary shaft, retromolar area, calvaria, and scapula.¹⁷

The favourite donor sites for craniofacial reconstruction are the calvarium, rib, and iliac bone. It seems that there is less resorption of cranial bone that is the case rib or iliac bone.¹⁸ The difference between the resorption rates of cranial and rib or iliac bone comes from the different proportions of the cortical and cancellous components. Cranial bone has some disadvantages, such as technical difficulties in manipulation, brittleness, and limitations in the amount of cancellous bone. As for the rib graft, because of the contour of the split rib, the ease of bending, and it's relative thinness, it would seem to be one of the ideal materials for augmenting and contouring the restoration for the craniofacial region. However, complications of the donor site are common, 12% involve pneumothorax. The iliac bone is adequate when a large bone segment involving cancellous bone is needed, but the persistent pain is a common problem. A split-thickness calvarial autogenous graft is the material of choice CFR but children under the age of 6 years may need another bone source due to lack of the skull thickness.

Demineralized bone can be used for a craniofacial defect with minimal tissue reaction and remarkable little osteoclastic activity. Eight to twelve weeks after the implantation of demineralized bone, new bone growth was noticed in histologic evaluations. More bone and endothelial growth were noticed on the dural aspect of the calvaria and its continuity with the implant surface.

The healing of autogenous bone grafts parallels that of fracture repair. An important similarity in bone graft healing is that a substantial portion of the biological activity originates from the host. This occurs because most viable osteocytes within the graft itself necrose shortly after transplantation, rendering the graft relatively inert. Nonetheless, substantial biological interactions still remain between graft and host. This important biological interplay contributes to the final outcome of graft take.

Osteogenesis refers to the process that occurs when surviving osteogenic cells from within the graft produce new bone. This mechanism of graft healing relies on the transplanted osteogenic cells to retain viability and produce osteoid. These cells are believed to be derived from the periosteum, endosteum, marrow, and intracortical elements of the graft.¹⁹ The role of osteogenesis as a mechanism of the new bone formation during non-vascularized bone graft healing, however, is thought to be of lesser significance than that of osteoconduction

Nonvascularized bone grafts heal through a predictable sequence of events. Bone grafts initially undergo partial necrosis, followed by an inflammatory stage. During this phase, much of the grafted bone is replaced by new bone as a consequence of interactions between osteoclasts and osteoblasts, which are delivered through invading blood vessels. The term *creeping substitution* is used to describe this slow vessel invasion and bony replacement, a process formally known as *osteoconduction*. This mechanism may be conceptualized by envisioning the graft as a scaffold on which new bone is deposited.

Because of the surgical disruption of host soft tissues and the recipient bony bed, hematoma formation around the graft occurs shortly after bone graft transplantation. During this early stage, a small minority of cells on the graft's surface are able to survive, primarily as a result of plasmatic imbibition.²⁰ An inflammatory reaction focused around the graft ensues after hematoma formation and lasts for 5 to 7 days. The inflammatory tissue is then reorganized into a dense fibrovascular stroma around the graft, and the onset of vascular invasion occurs at 10 to 14 days.²¹ Vascular invasion brings additional cells with osteogenic potential into the graft,²² as the interstices of the old bone act as a directive matrix. As osteoblasts deposit new bone, osteoclasts resorb necrotic bone and pave the way for the graft to be penetrated by vascular tissue.

Osteoinduction refers to the process by which active factors released from the grafted bone (e.g., BMP) stimulate osteoprogenitor cells from the host to differentiate and form new bone. Three phases of osteoconduction have been described: chemotaxis, mitosis, and differentiation. During chemotaxis, bone inductive factors direct the migration and activity of osteogenic cells via chemical gradients. The inductive factors then stimulate these osteoprogenitor cells to undergo intense mitogenic activity, followed by their differentiation into mature, osteoid-producing cellular elements (i.e., osteoblasts). Ultimately, the cells become revascularized by invading blood vessels and are incorporated as new bone. The ultrastructural character of the bone graft (i.e., cancellous versus cortical) determines the ability of revascularization to take place and, therefore, significantly impacts the process of incorporation.

Cancellous bone grafts are more rapidly and completely revascularized than cortical bone

grafts.²³ The large spaces between trabeculae in cancellous grafts permit the unobstructed invasion of vascular tissue and the facile diffusion of nutrients from the host bed. This is thought to promote osteogenic cell survival, imparting increased osteogenesis when compared with cortical grafts. Osteoprogenitor cells, brought in by the invading vessels, differentiate into osteoblasts and deposit a layer of new bone around the necrotic trabeculae. An osteoclastic phase ensues, wherein the entrapped cores of dead bone are resorbed. Cancellous bone grafts are completely revascularized and ultimately replaced with new bone over several weeks to months.

Ultimately, all of these changes are applications of Wolff's Law, which states: "Remodelling of bone occurs in response to physical stresses -or to the lack of them- in that bone is deposited in sites subjected to stress and is resorbed from sites where there is little stress".²⁴ In essence, a bone's form follows its function. LaTrenta *et al.* support these theories, reporting that inlay bone grafts in a dog model maintained greater volume and weight than on-lay grafts, and citing favourable remodelling forces of the inlay position.²⁵

Autogenous bone grafts are considered the gold standard for reconstructing craniofacial bone defects, the nature of which dictates the type of graft used. Inlay bone grafts are used for the treatment of bone gaps, whereas on-lay bone grafts are used to restore bone projection. Cancellous bone grafts are well-suited for inlay bone grafts because they revascularize quickly and stimulate significant new bone formation through osteoinduction. Conversely, cortical bone grafts are often used as on-lay bone grafts in cases of volume deficiency (e.g., malar augmentation). These grafts survive without complete resorption and retain some mechanical strength after transplantation.

Allogenic bone grafts refer to the transplantation of bone from genetically nonidentical individuals. To date, bone allografts have been plagued by their unpredictable rates of resorption and bone formation. Allografts also carry the coincident risk of disease transmission. In 1997, the U.S. Food and Drug Administration (FDA) implemented an extensive donor screening protocol with the hope of reducing the transmission of HIV and Hepatitis B and C viruses (Screening and Testing of Donors of Human Tissue Intended for Transplantation, 1997). The same factors that reduce immunogenicity, however, also de-activate the osteoinductive factors that are so critical to survival. In addition, deep freezing (-70°C) and freeze-drying -the most common methods of preservation- may significantly alter the mechanical properties and strength of the graft

Among alloplasts titanium has been used extensively in the last 20 years due to its malleability, lightweight, and bioinert and non-magnetic properties.²⁶ Others include polymethylmethacrylate,²⁷ hydroxyapatite,²⁸ and more recently bioceramics.²⁹ Because of the increased rate of infection, extrusion, and intense foreign body reaction associated with the use of alloplasts, cranioplasty using autologous split rib and calvarium has been universally accepted as the preferred option in adults and pediatric patients.^{30,31}

CONCLUSION

Bone grafts are the building blocks of the craniofacial surgeon. To optimize their use, a clear understanding of graft physiology is required. Clinical success with bone grafting may be maximized with reverence to the established variables affecting free graft survival. Modern science has also provided bone graft alternatives and adjuvants in the form of bone substitutes and recombinant human growth factors. In addition, the phenomenon of mechano-transduction may take on new applications in de novo osteogenesis.

Although these modalities represent exciting adjuncts in the field, they have not, as of yet, uniformly supplanted the autogenous, nonvascularized bone graft as the gold standard in craniofacial skeletal reconstruction. A critical appraisal of these novel techniques will be required in the ongoing quest to restore the craniofacial skeleton.

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