CHAPTER 121
Plastic and Reconstructive Surgery

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Introduction

In most parts of the African continent, plastic and reconstructive surgery remains among the least developed surgical specialties, with training opportunities available only abroad. As a result, this specialty still depends largely on the abilities of nongovernmental organisations (NGOs), philanthropic individuals, and organised surgical safaris/camps to meet the reconstructive needs of the majority of the populace. Because of the overwhelming needs for this service, general surgeons and the few paediatric surgeons provide this care. It is therefore essential that the surgeon managing paediatric reconstructive surgical problems be well informed on the spectrum of presenting pathologies, as well as the available options and resources for their treatment.

This chapter highlights the most common reconstructive topics peculiar to Africa. It is not intended to be a comprehensive text, and the reader is referred to appropriate texts, including those mentioned at the end of this chapter. Illiteracy, superstition, poverty, and armed conflicts continue to hamper the evolution of health care for all in Africa into the 21st century. In addition, children with disabilities or deformities are still hidden away from school and society. Most public transport, health, and education systems remain disability-unfriendly. Surgeons can become agents of change; by providing quality surgical care and educating the relatives and community, children with disabilities or deformities can become socially acceptable. Educational opportunities will give them the vehicles to self-actualisation.

A good understanding of embryology and anatomy are indispensable for the management of paediatric plastic and reconstructive surgical problems. Embryology is important because congenital aberrations and malformations form the majority of these problems; surgical restoration of form depends on a good knowledge of normal anatomy. As a caution to the surgeon, the patient and relatives of the patient, an appreciation of the limitations of current surgical techniques in achieving desired results is essential. Patients and their relatives must be involved in the decision-making process and informed on available options. Realistic goal-setting is a crucial part of reconstructive surgery. The physician must also gently inform the patient and relatives of the need for long-term follow-up and/or therapy. The surgeon seeks to restore form, function, and cosmesis, but this is not always possible, and part of the process of goal-setting must include agreeing on which of these aspects are the most feasible and most important to achieve. Staged procedures and their results should also be carefully relayed to the patients and relatives.

Whenever a congenital malformation is presented to the clinician, the history taken must include, amongst others:

• prenatal and postnatal history;

• maternal and paternal age;

• family medical history;

• consanguinity;

• previous maternal abortions; and

• teratogens.

A search must be made for other malformations in the child. Failure to diagnose a cardiac anomaly preoperatively may result in the death of a child from a non–life-saving procedure.

Skin Grafts

In many instances in surgical practice, relatively superficial or simple wounds cannot be closed primarily; skin grafts are a good choice for wound closure in such circumstances. Skin grafts may be split- or full-thickness (Table 121.1), depending on the amount of dermis included. Split-thickness grafts consist of the epidermis and varying amounts of dermis, whereas full-thickness grafts carry the entire dermis as well. Skin grafts carry with them adnexal structures.

Table 121.1: Features of split-thickness versus full-thickness grafts.

Parameters	Split-thickness graft	Full-thickness graft
Adnexal structures	Contains sweat glands, sebaceous glands	Contains sweat glands, sebaceous glands
Donor site	Donor sites heal primarily from skin adnexal structures	Requires closure or a split-thickness skin graft for closure
Available donor sites	Large amounts can be harvested	Limited donor sites. Size limited because of donor defect.
Graft "take"	Takes readily	Take is more precarious
Cosmesis of graft/color match	Poor	Fair to good
Primary contracture	Minimal	Significant
Secondary contracture	Significant (up to 40%)	Minimal

Primary contraction is the recoil of skin grafts immediately after harvest. It depends on the amount of elastin present in the dermis—the thicker the dermal layer, the greater the degree of contraction. This is not of clinical significance. Secondary contracture, in contrast, involves myofibroblastic activity. The thinner the graft, the greater the resulting secondary contracture.

The donor site must be well prepared to receive the skin graft for a successful "take". Skin grafts will not take on infected beds, exposed bone (without periosteum), cartilage (without perichondrium), or tendon (without paratenon). The only exception to this general rule is the facial skeleton. Skin grafts take well on temporal and orbital bones even without periosteum.

Meshing skin grafts increases the size of the available graft, but results in a poor cosmetic ("pebbled") appearance. The skin grafts must be well applied to the recipient bed. A moist environment enhances graft take, and thus wet bolster dressings should be applied to maintain close apposition, as well as to maintain a moist environment. Haematomas and seromas compromise skin grafts if not well immobilised.

If the recipient bed is bleeding excessively, the harvested skin may be stored in saline and applied the next day in the ward. This is an especially useful manoeuvre when operating time is limited. Skin stored in saline in a refrigerator is viable for up to one week.

Flaps

Unlike skin grafts, flaps have their own blood supply. For more complex, unstable, or deep wounds, skin grafts are inadequate. More tissue is required to fill these defects, and thus flaps are used. The flaps used may be pedicled or free, random or axial. Depending on the needs of the recipient, the defect, and available tissue, flaps may be cutaneous, myocutaneous, or even osteomyocutaneous.

Vacuum-Assisted Closure

Negative-pressure therapy, also called vacuum-assisted closure (VAC), was begun as a method of treatment of chronic wounds that were resistant to standard treatment modalities. Subsequent studies indicated that VAC therapy was a useful adjunct in the treatment of patients with lower-extremity injuries with exposed bone, tendon, and orthopaedic hardware. To date, negative-pressure therapy has been used for the management of all types of complex wounds and related conditions, including gastrointestinal fistulas, dehisced sternotomy, and abdominal wounds. Standard VAC therapy is expensive. VAC involves a piece of foam with an open-cell structure inserted into the wound, a drain placed on the foam, and the entire construct covered by a transparent adhesive plastic sheet secured to healthy wound margins. The drain is connected to a vacuum machine with a disposable reservoir.

The mechanism of action remains controversial, with many mechanisms proposed, including the acceleration of wound debridement, maintenance of a moist wound environment, and removal of oedema fluid. The VAC further improves wound perfusion, stimulates formation of healthy granulation tissue, and decreases bacterial colonisation.

The use of negative-pressure therapy significantly reduces the number of wound dressing changes as well as the associated pain.

Negative-pressure therapy is contraindicated in malignant wounds, grossly infected or necrotic wounds, and in wounds with inadequate haemostasis.

Because of the expense involved in procuring VAC kits, many hospitals in the developing world have developed novel negative-pressure therapy systems. Successful use of wall suction or suction machines (that must be turned on and off to simulate interrupted negative-pressure) therapy with gauzes packed into the wound and clean food-wrapping plastic available in the local supermarkets have been reported.

Tissue Expanders

Tissue expanders increase the surface area of local tissue, facilitating performance of reconstructive procedures and closure of defects that would otherwise require complicated procedures. Progressive inflation increases the overlying tissue and avoids the need for transfer of distant tissue, maximising aesthetic results.

Tissue expansion requires at least three stages: (1) the initial insertion; (2) an intermediate period of inflation to a predetermined size over a period of weeks; and (3) a final procedure, during which the prosthesis is removed, the flap is developed, and it is used to cover the defect. Leaving the injection port outside the skin avoids the pain of repeated injections through skin in subsequent visits. The amount instilled at each visit is determined by the amount that either blanches the skin or causes pain, whichever comes first.

Deformities resulting from burns, trauma, and irradiation, as well as congenital defects such as Poland's syndrome, are all amenable to coverage by use of tissue expansion. Myocutaneous flaps and full-thickness donor sites may all be pre-expanded to provide more tissue, both for the recipient site, as well as to facilitate primary donor closure, especially with free flaps. The resulting cosmesis is significantly enhanced. Unfortunately, tissue expanders are relatively expensive and have not yet found widespread use across Africa.

Several comments can be made regarding complications involved with tissue expansion.

- Infection may result, either from contamination of the prosthesis at the time of surgery, or later from exposure of the implant.

- Expander exposure is usually due to technical problems related to poor prosthesis placement or aggressive expansion. Remove the expander if exposure or infection occurs early in the course of expansion. If either occurs much later, the procedure is completed as planned.

- Discomfort and pain associated with expansion are usually transient and can be managed by using analgesics.

- The lower extremity, back, buttock, and neck are highly prone to complications.

- Skull deformation is temporary and resolves with time after the expander is removed.

- Implant deflation may occur if the prosthesis accidentally gets disconnected from the filling port.

- Seromas and haematomas are common, and are best prevented by the use of a closed/suction drain at the time of prosthesis insertion.

Vascular Lesions

Vascular lesions may be divided into haemangiomas and vascular malformations. Management of congenital vascular lesions remains a major challenge because their therapy carries a substantial risk of recurrence, morbidity, and—depending on location and size—even mortality. The wide spectrum of presentation extends from innocuous birthmarks to life-threatening conditions, such as the aggressive kaposiform haemangioendothelioma and tufted angioma (both associated with the Kasabach-Merritt phenomenon). Surgical extirpation is associated with significant bleeding, requiring massive transfusion. The lesions are therefore managed only in centres that are equipped to handle the expected difficulties and complications. Table 121.2 presents the most common sites and clinical evolution of vascular lesions.

Table 121.2: Vascular lesions.

Lesion	Most common sites	Clinical evolution
Haemangioma	Skin and viscera	Initial proliferation followed by involution. Size, location, and shunting are main concerns.
Kaposiform haemangioendothelioma	Skin and other tissues	Associated with Kasabach-Merritt phenomenon.
Tufted angioma	Skin	Slow growth. May present with tenderness or Kasabach-Merritt phenomenon.
Angiosarcoma	Skin and liver; pulmonary metastases possible	Degeneration of hepatic haemangioendothelioma or vascular malformation into angiosarcoma.
Congenital haemangioma	Skin	Spontaneous regression may occur; rarely, mild thrombocytopenia.
Eccrine angiomatous hamartoma	Skin (limbs)	Benign but painful.
Pyogenic granuloma	Oral, nasal cavities	Associated with trauma. Recurrence can occur in up to 15% upon excision.

Haemangiomas

Haemangiomas are the most common tumours of infancy and childhood, affecting approximately 12%, 1.4%, and 0.8% of Caucasian, African, and Asian infants, respectively, by the age of 1 year. The

diagnosis is made from the patient's history and physical examination findings. There is a slight female predominance, and 60% of the lesions occur in the head and neck region.

Mulliken and Glowacki developed a biological classification of congenital vascular lesions based on the degree of cellular turnover, histology, clinical findings and natural history. They described two major groups: infantile haemangiomas and vascular malformations. The natural course of haemangiomas is defined by two distinct clinical stages: proliferation and involution. Except for the occasional precursor lesion, haemangiomas are not evident at birth. Proliferation occurs during the first 12 months of life, whereas involution begins at 1 year, peaks at 2 years, and gradually diminishes thereafter. Proliferation involves an increase in the size of the lesion. In contrast, involution occurs by cellular apoptosis, clinically characterised by a gradual decrease in size.

The behaviors of congenital haemangioma and noninvoluting haemangioma differ from that of classic haemangioma.

Significant complications may occur in up to 20% of patients with haemangiomas, including:

- ulceration (the most common complication);

- compression of vital structures, depending on location;

- high-output cardiac failure;

- bleeding; and

- Kasabach-Merritt syndrome.

Investigations

A full haemogram, chest radiograph with ultrasound (US) scan, computed tomography (CT) scan or magnetic resonance imaging (MRI) may be requested, as indicated and based on availability.

Management depends on the location of the lesion, its clinical effects, and the age of the child. Conservative management, consisting of observation, serial photographs, and regular review with reassurance, is the preferred mode of management.

Complicated haemangiomas may require any combination of the following:

- compression therapy;

- intralesional steroids (some response in 50–90% of cases);

- interferon α-2a;

- chemotherapy;

- intralesional sclerosing agents (hypertonic saline, glucose);

- cryotherapy;

- arterial embolisation;

- radiation therapy; and

- surgical intervention.

Surgical intervention is indicated in cases of rapid growth, ulceration, haemorrhage, or deterioration of haemodynamic status (as in high-output cardiac failure or in secondary ischaemic complications caused by high-flow arteriovenous shunting). The mass effect of a lesion due to its proximity to vital organs (e.g., orbit), or compression on the trachea or auditory canal are other indications for surgical intervention. A cosmetically severe deformity and intervention designed to improve quality of life (as in lesions located in eyelids, vermillion border, nasal tip (the so-called "Cyrano de Bergerac" deformity), and the eyebrow) would also be considered relative indications.

Resection is possible only in small lesions. Surgical debulking is all that can be performed in larger lesions. In much larger lesions, compartmentalisation is necessary. Multiple sutures are placed so as to create many segments, which are then subsequently injected with sclerosing agents, leading to a process of initial swelling, inflammation, and a gradual decrease in the size of the lesion.

Vascular Malformations

Vascular malformations are generally evident at birth and grow at a rate commensurate with the growth of the child. Vascular malformations include:

1. *Capillary malformations*, which remain present for life and have no tendency toward involution. Presentation depends on size and location.

2. *Venous malformations*, which are usually present at birth but may not be clinically apparent. They are bluish, soft, and easily compressible, presenting as diffuse and extensive or localised lesions. Small cutaneous lesions may be treated with intralesional injection with 1% sodium tetradecyl decanoate.

3. *Intracranial arterial malformations*, which are more common than extracranial lesions, and usually present in the head and neck region. They are usually present at birth, but may not be noticeable.

4. *Lymphatic malformations*, which are the most common congenital vascular aberrations. They may be the result of hypoplasia of lymphatic trunks and lymph nodes, presenting as lymphoedema, or may present as superficial or deep single or multiple cystic lymphatic malformations. The latter are divided into macrocystic (formerly cystic or cavernous hygromas) and microcystic (formerly lymphangioma or lymphangioma circumscriptum) lymphatic malformations. Cystic lymphatic malformations are always present at birth, but may not become apparent until later in life. Surgical excision is the mainstay of management.

5. *Arteriovenous malformations*, which are rare and clinically apparent at birth. They have a high propensity to bleeding and recurrence. Spontaneous rupture accompanied by life-threatening haemorrhage may also occur. Depending on size and location, embolisation followed by surgical excision is the best form of management.

6. *Pyogenic granuloma* (lobular capillary haemangioma), which is a benign lesion that occurs on skin and mucosal surfaces of the proximal aerodigestive tracts, but has also been reported to occur subcutaneously, intravenously, in the small bowel, colon, rectum, burn scar, and thigh. It may arise at any age, but is more common in childhood.

7. *Nasopharyngeal angiofibroma*, which is the most common benign tumour of the nasopharynx, and may present with recurrent epistaxis, nasal congestion, hearing loss, or even cranial nerve palsies, usually in prepubertal and adolescent males. A clinical exam with radiographic or CT/MRI scans will confirm the diagnosis. Embolisation followed by surgical excision is the standard mode of therapy, although radiotherapy and chemotherapy have also been used.

Other classifications divide vascular lesions into "low flow" (capillary, venous, and lymphatic) and "high flow" (arterial and arteriovenous) lesions.

Craniofacial Surgery

An appreciation of the dynamics of the growing skeleton is essential for the surgeon venturing into management of paediatric craniofacial pathology. A child is not a small adult; even though a child's anatomy may appear similar to that of an adult, the behavior and response of the growing skeleton to trauma, whether accidental or iatrogenic, is considerably different. Such trauma may thus result in poor craniofacial cosmesis, form, and function, including mastication and deglutition. An understanding of the importance of craniofacial anatomy and cosmesis, as well as the major role it plays in defining an individual's identity, is central to the success of craniofacial surgery. Where possible, a team approach in craniofacial surgery is recommended to obtain optimal results. This team may include the following specialists: a paediatrician, the surgeon, a neurosurgeon, a dentist, a speech therapist, and a

nurse. Other specialists may be co-opted as needed. In practice, such a team is not feasible in most of Africa, and a more realistic approach would be to invest as many of the skills in the key members of the team—the surgeon and the nurse—as possible.

Most paediatric craniofacial problems are not merely surgical: they span the entire life of the individual, having their maximum impact in childhood and early adulthood. Appreciably, therefore, where possible, a team approach is required throughout life. Due counselling, with involvement of the patient and family in the decision-making process, is essential for the patient to gain self-esteem and fit into the various phases of life.

Definitions

Definitions of developmental aberrations will help in gaining a better understanding and appreciation of the need for a broad team approach in handling paediatric craniofacial problems.

- A *malformation* is a morphological defect of a part of the body consequent to an abnormal developmental process. Generally, malformations initiated late in foetal development are simple, whereas those initiated early (e.g., cleft lip, cleft palate, and microcephaly) are more complex and severe.

- *Deformation* is an abnormal form or position of a body part caused by nondisruptive mechanical forces. Deformations usually occur late in foetal development and may improve spontaneously once the deforming force is removed.

- *Disruption* is a defect of a part of the body resulting from mechanical interference with the normal developmental process (e.g., amniotic bands leading to amputations). The resulting defect depends on the timing of the mechanical insult. Disruptions are rare and occur sporadically.

- A *sequence* malformation is the result of multiple defects occurring consequent to a single presumed structural anomaly. An example is the Pierre Robin sequence, in which the primary event is thought to be a micrognathia resulting in a relative macroglossia, and consequently a failure of the palatal shelves to fuse.

- A *syndrome* encompasses groups of anomalies that contain multiple malformations and/or sequences, such as Treacher-Collins syndrome.

- An *association* is the nonrandom occurrence of a group of anomalies in multiple individuals. The constituent groups of anomalies must not be a sequence or syndrome. Examples are the CHARGE (Coloboma, Heart defect, Atresia choanae, Retarded growth, Genitourinary anomalies, Ear malformations) and VATER (Vertebral defects, imperforate Anus, Tracheo-Esophageal Fistula, and Renal) associations.

Airway Problems

Many congenital craniofacial anomalies have associated airway, hearing, and speech problems. Nasal obstruction is a major problem in infants younger than 3 months of age because at this stage in life they are obligate nose breathers. Affected infants have paradoxical cyanosis: cyanosis at rest with resolution upon crying. These babies need oral airways. Babies with micrognathia or maxillary hypoplasia should be nursed prone and with a nasopharyngeal airway. A tracheostomy may be performed as needed. Adenotonsillectomy may increase the size of the airway and improve ventilation.

Speech Problems

Speech disorders present mainly as hypernasality (as in velopharyngeal incompetence (VPI) and craniofacial anomalies associated with hearing loss); hyponasality (as in nasal obstruction); and hoarseness (as in VPI or after endotracheal intubation). These problems improve with speech therapy, or require surgery.

Common Presentations

Examples of common presentations include the following syndromes and sequences:

- The *Treacher-Collins syndrome* has an autosomal dominant inheri-
tance, with bilateral abnormalities of first and second branchial arches resulting in maxillary, mandibular, and zygomatic hyplasia. Affected children have downward-slanting eyes with colobomas of the lower eyelid and no eyelashes. Choanal atresia may be present, and airway control is therefore potentially difficult.

- The *Pierre Robin sequence* is a triad of micrognathia, glossoptosis, and cleft palate. Twenty percent of patients are syndromic and 80% are not. Airway management is a major concern, and failure of a nasopharyngeal airway may necessitate a tracheostomy.

- *Apert and Crouzon syndromes* are autosomal dominant and exhibit craniosynostosis, hypertelorism, exopthalmos, and maxillary hypoplasia. Whereas children with Crouzon syndrome have normal intellect and normal extremities, those with Apert syndrome have complex symmetrical syndactyly of the hands and feet as well as cervical vertebral anomalies, and they may be mentally retarded.

- The *Goldenhar syndrome* (the oculo-auriculo-vertebral complex) is defined by a unilateral craniofacial malformation involving the first and second branchial arches. Most cases occur sporadically. Clinically, these children have facial asymmetry, unilateral ear deformities, and vertebral malformations. They also have upper eyelid colobomas. Facial weakness occurs in 10–20%.

- The *Binder syndrome* (maxillofacial dysplasia) is characterised by a peculiar arrhinoid facies—affected children have no frontal sinus. They also have abnormal nasal bones with nasal mucosal atrophy, an absent anterior nasal spine, and maxillary hypoplasia with malocclusion.

- The *CHARGE association* involves four or more of the following presentations, from which it derives its name: coloboma, heart defect, choanal atresia, retarded growth, genitourinary anomalies, and ear malformations.

Cleft Lip and Palate

Cleft lip and palate have been described in more than 300 syndromes. Intrauterine diagnosis of cleft lips is possible from the second trimester of pregnancy. Cleft palates are more difficult to recognise with ultrasound. Postnatal diagnosis is clinical.

Other facial clefts have been described. The Tessier classification is the most commonly used system of classification. However, due to the rarity of these clefts, they will not be considered here.

Embryology

The primary palate is anterior to the incisive foramen and develops from mesodermal proliferation and fusion of the median nasal and maxillary processes during the 4th to 7th week of gestation. The secondary palate forms posterior to the incisive foramen in the 6th to 9th weeks of gestation. Palatal shelves change from a vertical to horizontal position and fuse in the midline. The tongue must first move antero-inferiorly to permit this fusion. Failure to move leads to palatal clefting, as in the Pierre-Robin sequence. A cleft lip may increase the likelihood of a cleft palate because the tongue may get trapped.

The secondary palate develops from the lateral maxillary processes that fuse in the midline. Thus, cleft lip and palate occur at 4–6 weeks of gestation, due to mesenchymal deficiency, whereas isolated cleft palate forms later, at 7–12 weeks, from partial or total failure of midline fusion.

Epidemiology

Most reports indicate that combined cleft lip and palate is the most common anomaly of this type (50%), followed by isolated cleft palates (30%), and isolated cleft lips (20%). Onyango and Noah reviewed 309 children managed in a single institution in Kenya and reported more isolated cleft lips than cleft palates. Accurate data for the African continent is lacking; however, estimates suggest there may be up to 400,000 children with clefts in Africa. Diabetic mothers and older paternity may be risk factors for the development of orofacial clefting.

Classification

Many different classifications are in use. The cleft lip may be unilateral or bilateral, complete or incomplete. Cleft lips occur as isolated anomalies or in combination with cleft palates. The cleft palate, likewise, may be uni- or bilateral, complete or incomplete. Submucous clefts and bifid uvulae are examples of incomplete palatal clefting.

Pathophysiology

The child with a cleft lip is unable to seal in air or fluid, when talking or eating. Malocclusion results from failure of lip seal, as well as due to intrinsic deformities of the alveolar process and teeth. In the child with a cleft palate, the nasal and oral cavities are in continuity, and air and fluid escape through the nose on attempted speech or drinking.

Associated problems

Children born with cleft lips and/or cleft palates are faced with the following potential problems: abnormal midface development, velopharyngeal incompetence, speech disorders, eustachian tube dysfunction with repeated middle ear infections, hearing and feeding difficulties, and malocclusion. Malocclusion occurs in almost all of these patients. Other significant dental anomalies include supernumerary teeth (20%), dystrophic teeth (30%), and missing teeth (50%). These problems contribute to poor cosmesis and may lead to a poor self image if not properly addressed, resulting in significant psychological difficulties.

Primary repair of cleft lip

Anatomically, there is lack of continuity of skin, muscle, and mucous membrane of the lip with associated nasal deformity. The skin, muscle, and mucous membrane should be repaired to restore lip continuity, symmetrical length, and function. Bilateral cleft lips are repaired simultaneously. In patients with cleft lip and palate, primary nose repair should include closure of the nasal floor. The use of presurgical orthodontics permits the palate to be moulded into anatomical position and may improve facial growth, leading to an easier and tensionless surgical repair.

Lip adhesion is a temporising measure that may improve breast-feeding as well as palatal anatomy; however, the definitive repair is more difficult because of scarring. This surgery is performed at about 2 weeks of age.

In primary cleft lip repair, the "rule of tens" is often used:

• 10 weeks of age;

• 10 pounds in weight, implying a healthy baby; and

• haemoglobin of 10 g/dl.

Early repair improves speech but may impair facial growth, leading to midfacial retrusion. Surgical repair alone, may improve VPI in up to 75% of patients. The rest require speech therapy, with surgical intervention being reserved for the few who fail to improve.

Although many procedures have been described, most surgeons use Millard's rotation-advancement flap technique. Common surgical complications include dehiscence, infection, and a prominent scar.

Primary repair of cleft palate

A palatal obturator may be used in high-risk cleft palate patients, in those who defer surgery, and in selected palatal fistulas that have repeated recurrences. The prosthesis is changed as the child grows.

Surgical repair is best performed around the age of 10 months, before development of intelligible speech. A number of techniques are currently in use, with the trend towards less scarring and less tension on the palate. Although push-back palatoplasty has been used widely, the Bardach or Veau-Wardill two-flap techniques are now popular. Scarring of the palate may cause impaired midfacial growth, leading to alveolar arch collapse, midface retrusion, and dental malocclusion. If surgery is delayed until the age of 18 months, facial growth may be less affected, but feeding, speech, and socialisation may suffer significantly.

Secondary procedures

Secondary procedures performed in cleft lip and palate patients, as indicated, include:

• *Pharyngoplasty* for velopharyngeal incompetence. The child may however develop stenotic side ports or continue to suffer from VPI. Of interest is the fact that some children with no VPI may develop incompetence after adenotonsillectomy.

• *Alveolar bone grafting* to close the palatal bony defect is delayed until the age of 6–12 years. The iliac crest is the most commonly used donor site. A palatal obturator may be used in patients with persistent fistulas.

• Midfacial advancement is achieved by using a *Le Forte osteotomy*. Potential complications include maxillary necrosis from vascular impairment, infection, and malocclusion.

• *Rhinoplasty* is used for the flattened nose and short columella, which frequently require secondary procedures.

Craniosynostosis

Craniosynostosis is the premature fusion of one or more cranial sutures, resulting in a partial or complete growth arrest of that suture, and leading to a skull deformity that is defined by the fused suture. Cranial growth is restricted perpendicular to the fused suture, leading to a corresponding compensatory growth parallel to this suture. The resultant cranial deformity causes a corresponding facial deformity. Some types of deformities are more common in certain populations than in others. In North Africans, for example, oxycephaly occurs with a high frequency.

Classification

Craniosynostosis syndromes (e.g., Crouzon, Apert, Pfeiffer, and Carpenter syndromes) are primarily due to genetic anomalies. Management of the child with syndromic craniosynostosis should include suspicion of hydrocephalus, increased intracranial pressure, mental retardation, and visual anomalies.

Table 121.3: Causes of secondary craniosynostosis disorders.

Disorder	Causes
Haematologic disorders	Sickle cell anaemia, thalassaemias
Metabolic disorders	Rickets, hyperthyroidism, mucopolysaccharidoses
Malformations	Microcephaly, shunted hydrocephalus, encephalocele
Teratogens	Valproic acid, diphenylhydantoin, etc.
Foetal head constraint	Uterine abnormalities, multiparous pregnancies

Table 121.4: Craniosynostoses classification.

Suture involved	Comments	Deformity
Metopic	Rare	Trigonocephaly and hypertelorism
Saggital	Most common form	Scaphocephaly
Coronal, unilateral		Frontal plagiocephaly
Coronal, bilateral		Brachycephaly/turricephaly
Lambdoid, unilateral or bilateral	Most posterior plagiocephaly due to deformational plagiocephaly	Posterior plagiocephaly
Multiple sutures	Rare	Cloverleaf deformity (Kleeblattschädel deformity)

Craniosynostosis can be primary or secondary. Primary craniosynostosis has no underlying identifiable abnormality, whereas secondary craniosynostosis occurs in association with other anomalies or identifiable causes (Table 121.3).

An anatomic classification of craniosynostosis by suture involved is shown in Table 121.4.

Management

The surgical procedure of choice for a specific type of craniosynostosis is age-dependent. The younger the patient, the less extensive or demanding the procedure. Strip craniectomies are generally performed in children younger than 6 months of age.

The differentiation between lambdoid craniosynostosis and deformational plagiocephaly is important, but may be a diagnostic difficulty. Deformational plagiocephaly, also called positional head deformity, is a benign cranial deformity resulting mainly from sleeping and nursing predominantly in one position (supine). There is no synostosis. There is a temporal association between increased deformational plagiocephaly and the "Back to Sleep" campaign in the West that was designed to combat sudden infant death syndrome (SIDS). Other causes include *in utero* compression and congenital torticollis. Most deformities are mild and resolve spontaneously, and the rest can be managed by the use of a moulding helmet.

Surgical intervention in craniosynostosis aims at correction of the craniofacial deformities and control of increased intracranial pressure, which is present in up to 30% of patients with two or more fused sutures.

Complications

Common complications of surgery for craniosynostosis include:

- *Haemorrhage:* Children do not tolerate blood loss well because of their small volumes, and loss must be replaced volume for volume.

- *Cerebrospinal fluid (CSF) leak:* Dural tears may occur. These should be sought for and repaired. If noted postoperatively, an initial conservative management may be attempted, but if this fails, definitive surgical repair must be performed.

- *Ocular complications:* Optic nerve injury or direct eye trauma is avoided by careful technique.

- *Infections:* These are rare, but potentially life-threatening should they occur.

- *Death:* Mortality should be rare.

Maxillofacial Trauma

In the management of paediatric maxillofacial trauma, tissue handling, both soft and especially bony, must be gentle, so as to ensure optimal restoration of mastication, deglutition, and cosmesis. The availability of resorbable plates and screws for use in craniofacial bone fixation has significantly reduced the morbidity associated with the need for hardware removal after fracture consolidation; however, the cost is prohibitive. Titanium plates and screws are the standard of care; however, wire fixation, although time consuming, is effective and affordable and does not require removal.

The craniofacial skeleton serves three main functions:

1. It offers structural support for muscles and other and soft tissues.

2. It protects important sensory organs.

3. Facial bones collapse from trauma and thus absorb most of the energy that would otherwise be transmitted to the brain.

In children younger than 5 years of age, the cranial skeleton is much larger than the facial skeleton; hence, there is a lower incidence of midfacial and mandibular fractures and a higher incidence of cranial injuries in this age group. With increasing age and facial growth, the midface and mandible become more prominent, with a concomitant increase in the incidence of facial fractures and a decrease in cranial injuries.

Incidence and aetiology

Social, cultural, and environmental factors vary from one country to another. These significantly influence the incidence and aetiology of craniofacial trauma. As examples, violence is the main cause of paediatric maxillofacial trauma in South Africa and Zimbabwe, and road traffic injuries account for 11% and 50% of paediatric craniofacial trauma in Zimbabwe and Libya, respectively.

Age-related activities in children also influence the incidence and aetiology of trauma. Younger (preschool) children usually sustain low-velocity trauma related to falls around the home, whereas school-age children are more likely to sustain high-velocity trauma, usually related to sports but also to road traffic injuries. Child abuse occurs in all cultures and across all paediatric ages.

Facial fractures in children occur less frequently than in adults. Facial fractures are frequently minimally displaced because of the thicker layer of adipose tissue, elastic bones, and flexible/deformable sutures. Lack of facial skeleton pneumatisation and the presence of tooth buds within the jaws are other contributing factors to a lower incidence of displaced fractures. However, as noted above, trauma to the growing skeleton may adversely affect long-term cosmesis. It is difficult to predict the long-term effects of any trauma because growth may serve to improve long-term results, as has been noted in occlusal realignment and compensatory condylar growth after condylar fractures.

Acute management

The initial presentation of a paediatric craniofacial injury may be so dramatic as to upset management priorities. The ABCs of trauma care must be recalled and used at all times; irrespective of the drama, these principles help save lives.

The ABCs of trauma care are:

A. *Airway:* Secure the airway and protect the cervical spine. This is especially important because craniofacial trauma may be associated with C-spine trauma (5–10%). Major facial trauma is also associated with the risk of upper airway obstruction. The obtunded patient will have no control of the tongue.

B. *Breathing:* Ensure adequate ventilation.

C. *Circulation:* Control bleeding. Life-threatening haemorrhage occurs in up to 5% of Le Fort fractures. Fracture reduction and direct pressure will generally control bleeding. Anterior and posterior nasal packing may be performed by using a urinary catheter passed through the nose into the nasopharynx, where it is inflated, then pulled back snugly.

D. *Disability:* Conduct a thorough neurological examination (e.g., Glasgow Coma Scale) to establish level of consciousness as well as any neurological deficits.

Administer tetanus toxoid to all trauma patients. Additionally, animal bite victims should be given the antirabies vaccine.

Cover avulsed tissues or organs (teeth, ears, nose, etc.) with saline-soaked gauze. Avulsed deciduous teeth may be discarded. Permanent teeth should be replaced and held in place for a little while. If a tooth has been out for more than 6 hours, it is unlikely to survive and may be discarded.

CT scans are the gold standard radiographic investigation in paediatric craniofacial trauma. Plain radiographs are not as helpful as they are in adults, particularly in the midface region, where poorly developed sinuses and tooth buds occupy space and obscure skeletal anatomic landmarks. The orthopantomogram (OPG) is a good tool in looking at mandibular fractures.

Because the plain radiograph may be the only tool available, paediatric radiograph atlases provide references for normal anatomy. Where radiographic examination is unavailable, clinical examination should reveal most of the injuries, with surgical exploration and repair as indicated. The index of suspicion is high, with clinical examination alone, however, and missed or untreated injuries are likely to result in significant morbidity, including poor facial cosmesis.

Nasal injuries

Nasal injuries may be associated with trauma to adjacent structures; thus, naso-ethmoid fractures may be missed if the apparent "simple" nasal injury is ignored. Also, septal perforation may complicate a missed septal haematoma that may also interfere with midfacial growth. Nasal fractures should be reduced and splinted at the earliest opportunity.

Mandibular trauma

Most mandibular fractures are simple, undisplaced or minimally displaced, and may be managed with maxilla-mandibular fixation. Minimal interference with the growing mandible and contained tooth buds will lead yield excellent results. The mandible is the final facial bone to complete growth, and injury to the condyle may result in facial asymmetry and malocclusion from arrested mandibular growth. Temporomandibular ankylosis and loss of mouth opening are further complications. If there are no other associated facial bone injuries, most condylar injuries are probably best managed conservatively, although this is controversial. There is currently no evidence that **open reduction, internal fixation** (ORIF) improves outcomes. Whatever the management, early institution of mouth-opening exercises are important to prevent temporomandibular joint (TMJ) fibrosis or ankylosis.

Maxillary trauma

Because sinuses develop between the ages of 6 and 12 years, the fracture patterns in children are different from those in adults. Thus, Le Fort fractures increase with increasing aeration of the maxillary and ethmoid sinuses. Displaced midfacial injuries, however, should be managed with open reduction and internal fixation, irrespective of age.

Orbital "blow out" fractures

Orbital "blow out" fractures are common in children. Failure to recognise and treat these injuries early will result in diplopia, enophthalmos, and extraocular muscle entrapment. These complications are difficult to treat. A high index of suspicion and early exploration and repair are mandatory due to the rapid rate of cicatrisation and consolidation seen in children.

Soft tissue trauma

As a general rule, soft tissue cuts and lacerations of the face and head (scalp) tend to bleed profusely. Because of good vascularity in this region, wounds heal well, and surgical wound infections are rare. Management consists of the following principles:

1. Wounds may be closed primarily safely, up to 24 hours after trauma.

2. Copious wound irrigation and minimal debridement are all that is required. Excessive debridement will lead to large defects that require skin grafts or other procedures, resulting in larger scars and inferior cosmetic results.

3. Degloved flaps that are still attached by a skin bridge should be preserved and replaced. Subsequent debridement will reveal how much of the flap is viable.

4. The suturing technique and material are equally important determinants of the scar. Intradermal sutures should be used to reduce tension on the skin sutures. Skin is closed by using a subcutaneous absorbable suture or with 5.0 monofilament, which must be removed on postoperative day 5 to avoid a "railroad" scar.

Animal bites are a common cause of facial trauma in children. The animal may bite off any of the parts of the face (ears, nose, or lips). At presentation, the piece of tissue may be missing or still attached. The same principles outlined above are used in managing animal bites, and if presentation is early, primary closure is attempted.

Complications

Problems commonly found in craniofacial trauma include:

- *CSF rhinorrhea*, which may be managed conservatively with head elevation at 30° to 45°.

- *Epistaxis*, for which, if anterior packing fails, additional posterior

nasal packing should be done. Packing should not be left in for more than 48 hours because of a significant increase in infections. If packing needs to be left in for longer, then prophylactic antibiotics are used and the packing must be removed within 5 days.

- *Infraorbital nerve damage* may occur with Le Fort 3 fractures.

- *Facial nerve paralysis* may occur in petrous temporal bone fractures.

- *Subcutaneous emphysema* is evidence of fracture of one or more sinuses.

- *Anosmia* is the result of fracture of the cribriform plate.

- *Ocular and visual disturbances* occur from orbital fractures.

- *Epiphora* is evidence of damaged lacrimal ducts.

If surgical principles are followed, complications are very few because of the healing capacity of the craniofacial skeleton. Whereas nonunion is unlikely, malunion is the result of inadequate reduction. Most ocular trauma , brain injury, and death are causally related to the initial trauma.

In Africa, late presentation of patients is a particularly common occurrence. Reconstructive surgery must then be offered. This is frequently difficult and expensive, and it may require multiple stages. Referral to a competent centre may be preferable, as resources required at this stage may be unavailable locally.

Noma

Noma, variously known as gangrenous stomatitis or cancrum oris, primarily affects orofacial structures. There is a strong correlation between noma and poverty, malnutrition, poor oral hygiene, and infectious diseases (primarily malaria and measles). These conditions are widespread in sub-Saharan Africa, particularly within strife-ridden communities. It has thus been aptly called "the face of poverty", and afflicts mainly children in developing countries. The World Health Organization (WHO) estimates 140,000 new cases of noma each year, mostly from sub-Saharan Africa, with a mortality rate of up to 90%. The few reports from developed countries involve patients with debilitating diseases, including HIV, diabetes mellitus, and haematological disorders.

Aetiology

Although bacteriological studies are inconclusive on causative organisms, most experts agree that noma is the result of a mixed bacterial infection involving aerobic and anaerobic organisms. Fusiform bacilli such as *Fusiformis fusiformis* and *Borrelia vincenti* are frequent isolates. Other isolates include *Prevotella melaninogenica*, *Corynebacterium pyogenes*, *Fusobacterium nucleatum*, *Bacteroides fragilis*, *Bacillus cereus*, *Prevotella intermedia*, and *Fusobacterium necrophorum*.

To date, there is no effective mode of treatment, and thus prevention plus the treatment of noma sequelae remain the mainstay of care. Broader-spectrum antibiotics covering gingival flora, including aerobes and anaerobes, are now recommended, but the treatment has to be started as soon as possible.

Clinical evolution

Noma presents initially as a painful blister on the gingiva that becomes indurated, leading to massive oedema. Subsequent necrosis, ulceration, and slough separation leave underlying bone exposed. The slough is cone-shaped, with the base being in the oral cavity. This is an important consideration at the time of reconstruction because the size of the skin defect alone is misleading. The final result is an ugly midfacial defect that then scars and contracts with time if the patient does not succumb. Initial management in the acute stage consists of fluid resuscitation, nutritional rehabilitation, and administration of broad spectrum antibiotics. Any predisposing illnesses and/or infections are managed appropriately. Wound care consists of gentle debridement and dressing changes. Physiotherapy will help prevent trismus and the resulting TMJ ankylosis.

Survivors of the acute stage suffer severe destruction of the midface, including lips, cheeks, maxilla and mandible, nose, and occasionally the orbit. Progressive scarring, trismus, oral incontinence, and mandibulo-maxillary synostosis leave the patient malnourished, severely disfigured, and with speech difficulties. The psychosocial difficulties experienced by these children and their families should not be underestimated.

Surgical treatment

Reconstruction in noma surgery is a difficult undertaking; collaboration with a team versed with noma care is helpful. Modified Montandon's classification is a simple, comprehensive, and practical clinical classification. It is useful in formulating surgical intervention. The addition of a fifth grouping completes this classification:

1. Localised lip, commissure, or cheek defect

2. Upper lip and nose amputation

3. Lower lip and mandible amputation

4. Large defects (e.g., lips, cheek, palate, maxilla, orbital floor)

5. Bilateral defects (may be any combination of the above)

Free flaps offer the reconstructive surgeon considerable versatility. Single-stage reconstruction with appropriate tissue bulk and the avoidance of additional facial scars and incisions are important advantages. Free flaps, however, are labor- and technology-dependent, are expensive, and demand considerable technical skills. It is encouraging to note that microvascular reconstruction has been successfully performed in suboptimal environments, proving that these skills and technology can be modified to suit the operating theatre conditions found in most developing countries.

Loco-regional flaps provide excellent color match, minimise the number of surgical procedures, and are generally fail-safe, requiring simple postoperative care. Thus, although ideal, the use of free flaps for noma reconstruction is not essential, and loco-regional flaps can give very good results if well chosen.

It is important that the surgeon and patient realise that even though cosmesis may be improved significantly by surgery, functional outcomes may be poor at best.

Craniofacial Tumours of Bony Origin

Craniofacial tumours affecting children are too numerous to be discussed in this chapter, so only the most common bony lesions encountered in Africa are discussed here. The general subject of tumours is covered elsewhere in this book (Chapters 102–109).

Burkitt lymphoma

Burkitt lymphoma was initially described as "African jaw lymphoma". It affects children, shows rapid growth, and may involve the mandible or the maxilla. Burkitt is *not* a surgical disease—it is a purely medical disease, and is effectively treated using chemotherapeutic agents. It shows excellent response to prednisolone, with a rapid decrease in volume evident within 24 hours of administration. A fine needle aspirate for cytological analysis or a core biopsy is all that is required for diagnosis.

Dentigerous cyst

A dentigerous (follicular) cyst is the most common developmental cyst, comprising about 20% of all developmental cysts. On an OPG, it is a radioluscent lesion with well-defined, sclerotic margins encircling an unerupted tooth. Most of these cysts are asymptomatic, but when they are large, they may cause the displacement or resorption of adjacent teeth and cause pain. Drainage and marsupialisation or enucleation is done for large or symptomatic cysts.

Fibrous dysplasia

With fibrous dysplasia (FD), normal bone is replaced by fibrous connective tissue due to a defect in osteoblast differentiation and maturation. This genetically based sporadic bone disease may affect single or multiple bones (monostotic or polyostotic, respectively). Twenty-five percent of monostotic FD occurs in the head and neck, and the maxilla is more often affected than the mandible. The polyostotic variant forms 15% of the total, and 50% of polyostotic lesions occur in the head and neck region. Cranial polyostotic FD may occur concurrent with café-au-lait spots and endocrinopathology (hyperthyroidism and precocious puberty in females), constituting McCune-Albright's syndrome.

FD clinically presents as a painless swelling with jaw asymmetry, usually in childhood or early adolescence, during the periods of greatest skeletal growth. Presentation depends on the specific site involved: hearing loss and facial palsy in temporal bone involvement; facial asymmetry and malocclusion with mandibular involvement; or pain, paraesthesia, and cranial nerve lesions with skull base involvement. Frontal bone involvement results in hypertelorism, with massive cranial bony involvement resulting in lionlike facies, known as "leontiasis ossea".

Cherubism, a variant of FD, is an autosomal-dominant disorder of variable penetrance. It is more severe in boys. The involvement of maxilla and mandible is symmetric. Some regression may occur after adolescence.

Radiologically, FD most commonly has a ground-glass pattern. FD lesions generally do not cross the midline and tend to be limited to one bone.

Management

Management of tumours of bony origin may consist of observation, conservative surgery involving shaping or contouring the dysplastic bone, or radical surgical excision and reconstruction. The choice depends on a combination of factors:

- age of the patient;

- growth rate of the lesion;

- extent and location;

- degree of cosmetic deformity; and

- level of functional impairment.

The aim of surgical treatment is to correct or prevent functional deficits and restore as near-normal facial cosmesis as possible. If possible, surgery should be deferred until skeletal maturity. Successful staged excision of extensive lesions, especially in the head and neck region, followed by multistaged reconstruction has been reported.

Accelerated growth or aggressive lesions require early surgical intervention with en bloc resection and bone graft reconstruction. The rate of transformation to malignancy is less than 1%, which has also been reported to occur after radiation therapy and is therefore contraindicated.

Chondrosarcomas and osteosarcomas are malignant lesions that occasionally affect the head and neck region. On the African continent, even in the best centres, these lesions generally have a poor prognosis.

Neurofibromatosis

Neurofibromatosis covers a number of distinct disorders that share many features but clearly differ from one another. These include neurofibromatosis type 1 (Nf1), neurofibromatosis type 2 (Nf2), and others, as recommended in 1987 by the National Institutes of Health (NIH) Consensus Conference on Neurofibromatosis.

Neurofibromas arise from nerve sheath cells and consist of a mixture of Schwann cells, fibroblasts, perineurial cells, and mast cells. Nf1, formerly known as von Recklinghausen disease, affects about 1 in 3,000 people. It has autosomal dominant inheritance with complete penetrance, variable expression, and a high rate of new mutation. The NIH Consensus Conference on Neurofibromatosis diagnostic criteria for Nf1 include neurofibromas, café-au-lait macules, skin-fold

freckling, skeletal dysplasia, Lisch nodules, optic gliomas, and a first-degree relative with Nf1. A positive diagnosis is based on two or more of these signs.

Skeletal involvement in patients with Nf1 may include macrocephaly, dysplasia of the greater wing of the sphenoid, vertebral dysplasia, scoliosis, pseudoarthrosis, and a short stature, amongst others. About 50% of patients with Nf1 have learning disabilities as well as some behavioral problems. There is also a heightened risk of epilepsy and headaches. These patients have an increased risk for developing malignant tumours, such as malignant peripheral nerve sheath tumours, leukemia, and rhabdomyosarcoma.

Nf1 lesions that appear early in childhood can be disfiguring and cause psychological discomfort. Most are not resectable because of their diffuse nature. They are difficult to deal with even if only an agreeable cosmetic result is all that is desired. Conservative management is recommended unless a lesion presents with sudden enlargement, functional deficits, pain, or bleeding. In the craniofacial region, however, the smallest neurofibroma lesion produces a prominent abnormality, whereas the larger ones can result in hideous deformities in the child, leading to ostracisation from society. Surgical excision and reconstruction should be offered for lesions of the craniofacial region. Surgical debulking is associated with significant bleeding.

Lymphoedema

Lymphoedema is the accumulation of protein-rich interstitial fluid within the skin and subcutaneous tissues as a result of lymphatic dysfunction. The accumulated fluid provokes an intense fibrous reaction, further reducing lymphatic transport and leading to progressive swelling of the limb. Limb induration, fibrosis, and repeated attacks of cellulitis set up a vicious cycle that, untreated, leads to ulceration and chronic nonhealing wounds. Normal unidirectional lymph flow is due to a combination of factors: intrinsic lymphatic muscle contractions, muscle pump action, presence of valves, and negative intrathoracic pressure. The lymphatic system functions to clear proteins and lipids from the interstitial space into the vasculature by use of differential pressures. Lymph flows into the venous system at the neck. Lymphoedema results from a decrease in the number of functioning lymphatics below a critical level, or from an increased lymphatic load.

Lymphoedema is confined to the subcutaneous compartment; the deep muscle compartments remain uninvolved. The excess protein serves as a medium for bacterial multiplication, and the resultant infection adds to the lymphatic dysfunction. Recurrent soft tissue infection is one of the most troublesome aspects of chronic lymphoedema management.

Causes

Lymphoedema may be divided into primary or secondary, based on cause.

Primary lymphoedema is of unknown aetiology or congenital lymphatic dysfunction. Primary lymphatic obstruction may be caused by several anatomic abnormalities, including lymphatic hypoplasia and functional insufficiency or absence of lymphatic valves. It is further subclassified based on the age of onset or lymphangiographic findings (Table 121.5).

Table 121.5: Subclassification of primary lymphoedem

Age of onset	• Birth: lymphoedema congenita (e.g., Milroy's disease)
	• Adolescence: lymphoedema praecox
	• Adulthood: lymphoedema tarda
Lymphangiographic findings	• Lymphatic aplasia
	• Lymphatic hypoplasia
	• Varicosity of subcutaneous lymphatics

Secondary lymphoedema is more common and has a known precipitating cause. It may result from surgical, infectious, inflammatory, neoplastic, or traumatic causes. Filariasis is a common parasitic cause of lymphoedema in Africa, usually presenting in adulthood.

Clinical Presentation

The oedema generally begins distally and progresses proximally over months to years. Initially soft and pitting, it becomes nonpitting secondary to fibrosis. Peau d'orange changes in the skin (due to fibrosis), the Stemmer sign (inability to tent skin over the toes), and the blunted appearance of the digits of the involved extremity are the classic signs of lymphoedema.

Differential diagnoses include: chronic venous stasis, myxoedema, and lipoedema. These are distinguished on the basis of history and physical examination.

Management

It is important to discuss with the patient and guardians the fact that lymphoedema has no cure. Conservative management should be initiated immediately when the diagnosis is made. This should focus on the avoidance of infection and the application of compression to maintain or to reduce limb volume. Elastic compression garments, for example, have been shown to lead to a 30–40% reduction of oedema. Improved skin care avoids infection and prevents the development of associated skin changes.

Other nonsurgical therapies include treating infection with antibiotics and improving protein lymphatic transport by stimulating macrophage proteolysis (benzopyrones) and reducing oedema (diuretics), among other therapies. *Wuchereria bancrofti* and *Brugia malayi* infections should be eliminated with diethylcarbamazine, and antihistamine and/or anti-inflammatory agents may be used to control the allergic reactions to the dying parasite. Lymphangitis must be treated quickly when it occurs, and the patient may require lifelong antibiotic prophylaxis to prevent further episodes, which may occur in up to 25% of patients.

Surgical treatment options may be grouped together into:

• re-creation or imitation of lymphatic channels;

• bridging the lymphoedematous area to normal lymphatic areas; and

• resecting or debulking of lymphoedematous tissue.

The procedure may be physiologic, attempting to re-establish lymphatic drainage, or excisional—procedures that debulk the limb by removing excess skin and subcutaneous tissue. These procedures aim at improving the cosmetic appearance, preserving the function of a limb, and preventing infection episodes. Whereas the re-creation of lymphatics requires microsurgical capability, debulking is a much simpler surgical procedure.

The Homans' or modified Sistrunk procedure is easy to perform and results in up to a 50% reduction in limb volume. Excision of subcutaneous tissue is carried out in two stages over a 2- to 3-month period, excising half of the limb circumference at each stage. The skin and part of the subcutaneous tissue are preserved. The Charles procedure has no durable results and is mentioned here only for historical reasons.

Complications

Recurrent lymphangitis and cellulitis, progression of skin changes, oedema, fibrosis, and ulceration with chronic nonhealing wounds constitute the natural progression of untreated lymphoedema. The limb becomes heavy, may be painful, and in extreme cases, the patient may be unable to ambulate.

Genital lymphoedema is a significant problem in some patients. The management is similar to that of limb lymphoedema. Surgical excision is indicated only for cosmetic or functional reasons, such as where the lymphoedema interferes with micturition or causes significant discomfort with ambulation or dressing. Solitary lesions of any size may be excised.

Pressure Ulcers

Pressure ulcers involve tissue loss resulting from constant pressure. Decubitus ulcers occur in areas with underlying bony prominences—sacrum, trochanter, heel—when the subject is lying down. Three to 4.5% of all hospitalised patients will develop a pressure sore at some time during their hospitalisation. Geriatric nursing homes have the highest prevalence of pressure ulcers (35–64%). A number of theories have been proposed to explain pressure ulcer development. Currently, the accepted mechanism involves a direct effect by one or more extrinsic, or primary, factors (e.g., pressure, shear, friction), propitiated and modified by a number of intrinsic, or secondary, factors (e.g., infection, impaired mobility). Although bacteria do not cause pressure ulcers, they contribute to tissue breakdown and delayed healing. Keeping bacterial counts at less than 10^4 per gram of tissue allows for wound healing.

Aetiology

Patient immobility, resulting in prolonged contact with objects, may be due to long bone fractures, paralysis (congenital (e.g., spina bifida), trauma, neoplasm, infection, iatrogenic injury, or radiation), and loss of consciousness. Interestingly, cerebral palsy patients do not develop pressure sores, even when placed in wheelchairs for a long time.

Classification

Pressure ulcers are staged and graded depending on the depth of involved tissue (Tables 121.6 and 121.7). Whereas ulcer staging helps nursing planning to help reverse progression, ulcer grading aids in determining the type and volume of tissue required for the reconstruction.

Table 121.6: Pressure ulcer staging.

Stage	Lesion
I	Hyperaemia ≥ 1 hour after pressure relief
II	Blister or other break in the dermis with or without infection
III	Subcutaneous destruction into muscle with or without infection
IV	Involvement of bone or joint with or without infection

Table 121.7: Pressure ulcer grading.

Ulcer grade	Tissue involved
I	Limited to epidermis and superficial dermis
II	Adipose tissue
III	Muscle ± infection
IV	Bone and/or joint ± infection

Source: National Pressure Sore Advisory Panel Consensus Development Conference, 1989.

Management

Pressure ulcers add a significant burden in terms of cost of care and morbidity of the patient. Uncontrolled wound infection may lead to sepsis and death. Thus, prevention remains the most effective means of pressure ulcer management. Consequently, basic skin care and pressure dispersion by timely position changes must be taught to parents and guardians of those at risk. Also important is the use of topical antibiotics, such as silver sulfadiazine. Many paraplegic children remain ulcer-free until they start going to school. Thus, teachers and peers at school need to be taught about pressure ulcer prevention so that they can reinforce it at school.

Once an ulcer has developed, its management depends on several factors. As with all reconstructive surgical procedures, the setting of achievable goals, by both the patient/family and the surgeon, must form the first step in the process. For patients with potentially reversible underlying diseases in whom the goals are to restore normal function or prolong life, aggressive efforts at nutritional resuscitation, treatment of infection, physical therapy and surgical intervention are warranted. In contrast, for chronically or terminally ill patients with advanced or recurrent ulcers, the goal is to provide comfort only.

The severity of the ulcer is evidenced by its grade (see Table 121.7). Grade III and IV ulcers require flap coverage; skin grafts and primary closure will fail. Free flap coverage may be preferable, depending on skills and personnel.

Patient idiosyncrasies and family support in nursing care are important factors. The older child may develop depression because of low esteem, among other reasons. As a result, the child moves less, and thus develops a pressure ulcer. In contrast, the motivated and goal-oriented child is less likely to develop a pressure ulcer. Family interest and involvement form an important and integral part of patient care. Poor family support implies poor nursing care. Any surgery performed will therefore most certainly fail, and conservative management would be preferable.

The cost of managing a pressure ulcer is significant. To protect a flap after coverage, weight and pressure dispersion may require special equipment, including silicone pillows and wheelchairs. These issues must be discussed before initiating surgical treatment.

The goals of surgical intervention include the following:

• reduction of protein loss through the wound;

• prevention of progressive osteomyelitis and sepsis;

• avoidance of progression to secondary amyloidosis and renal failure;

• minimisation of rehabilitation costs;

• improved patient hygiene and appearance;

• simplification of nursing care, as well as improving patient capability for self-care in the future; and

• averting future development of Marjolin's ulcer.

Technique

The technique in treating pressure ulcers includes the following:

• excision of the ulcer, surrounding scar, underlying bursa, and soft tissue calcification, if any;

• radical removal of the underlying infected bone;

• padding of bone stumps and filling the dead space with fascia or muscle flaps;

• resurfacing with large regional pedicled flap or free flap as indicated;

• in flap design, never sacrificing muscles that are still innervated; and

• grafting the donor site of the flap with thick split skin, if needed.

In doing so, the following guidelines are important:

• Use the simplest, but appropriate method of closure.

• Design the flap as large as possible; place the suture line away from the area of direct pressure.

• Be sure the flap design does not violate adjacent flap territories.

• If sensate flap is available, use it.

• Always have a second, and third, coverage option, should the first fail.

Regional flap options are shown in Table 121.8.

Table 121.8: Regional flap options.

Ulcer regions	Flap options
Trochanteric ulcers	Tensor fascia lata (TFL), vastus lateralis, anterolateral thigh flap
Sacral ulcers	Gluteal flaps
Ischial ulcers	Gluteal flaps, V-Y hamstring flap, posterior thigh flap

Complications

Complications of surgical intervention for pressure ulcers include haematoma, infection, dehiscence, recurrence, and heterotopic ossification of muscle flaps.

Microsurgery in Africa

The African continent is replete with reconstructive surgical needs, but the manpower to meet this is either inadequate or totally lacking. The College of Surgeons of East, Central, and Southern Africa has made significant strides in helping to train surgeons in various specialties, including plastic and reconstructive surgery.

The operating microscope has revolutionalised reconstructive surgery and thus needs to be part of the skills that trainees develop. The immediate cost of the microscope is significant, but the benefits to the surgeon and patients are immense. Procedures that would normally require multiple stages are performed in a single stage, significantly saving the morbidity, cost, and time lost in otherwise multiple-staged procedures. Note that the skills and equipment are easily available on the Indian subcontinent, which has patients whose needs and financial capabilities are equivalent to those in Africa.

Key Summary Points

1. An understanding of embryology is critical to the management of paediatric congenital malformations.

2. Children are not **"small adults"**; their physiology and anatomy differ from that of adults in a number of significant ways.

3. In reconstructive surgical interventions, goal-setting with the patient and the patient's relatives and guardians is an essential part of the management.

4. Surgeons managing children with disabilities or congenital malformations should participate in and/or lead campaigns of community education and awareness to help these children become integrated into their individual communities.

5. The long-term cosmetic and functional results of neglected maxillofacial trauma in children can be devastating. An early referral or consultation is important. Mobile telephony and the Internet can be used well to this end.

Suggested Reading

Andreev A. The Bulgarian School for Diagnosis and Treatment of Vascular Malformations and the leading role of Stefan Belov. Int J Angiol 2002; 11:46–52.

Argenta LC, Morykwas MJ. Vacuum assisted closure: a new method for wound control and treatment: clinical experience. Ann Plast Surg 1997; 38: 563–577.

Banwell PE, Musgrave M. Topical negative pressure therapy: mechanisms and indications. Int Wound J 2004; 1:95–106.

Baratti-Mayer D, Pittet B, Montandon D, et al. Noma: an "infectious" disease of unknown aetiology. Lancet Infect Dis 2003; 3:419–431.

Bauer J, Phillips LG. MOC-PSSM CME article: pressure sores. Plast Reconstr Surg 2008; 121(1 Suppl):1–10.

Brown MD, Johnson TM. Complications of soft tissue expansion. J Dermatol Surg Oncol 1993; 19:1120–1122.

Chao JJ, Longaker MT, Zide BM. Expanding horizons in head and neck expansion. Oper Tech Plast Reconstr Surg 1998; 5:2–11.

Cheville AL, McGarvey CL, Petrek JA, Russo SA, Taylor ME, Thiadens SR. Lymphedema Management. Semin Radiat Oncol 2003; 13:290–301.

Chidzonga MM. HIV/AIDS orofacial lesions in 156 Zimbabwean patients at referral oral and maxillofacial surgical clinics. Oral Dis 2003; 9:317–322.

Crysdale WS, Gaffney RJ. Malformations and syndromes. In: Bluestone CD, Stool SE, Kenna MA, eds. Pediatric Otolaryngology, Vol I, 3rd ed. Saunders, 1995, Pp 51–70.

Damstra RJ, Mortimer PS. Diagnosis and therapy in children with lymphoedema. Phlebology 2008; 23:276–286.

Enweon WU. Epidemiological and biochemical studies of necrotizing ulcerative gingivitis and noma (cancrum oris) in Nigerian children. Arch Oral Biol 1972; 17:1357–1371.

Marchac D, Renier D. Craniosynostosis. World J Surg 1989; 13:358–365.

Millard DR. Cleft Craft, Vol 1. Little Brown, 1976, Pp 165–173.

Miraliakbari R, Mackay D. Skin grafts. Oper Tech Gen Surg 2006; 8:197–206.

Montandon D, Lehmann C, Chami N. The surgical treatment of noma. Plast Reconstr Surg 1991; 87:76–86.

Mooney MP, Siegel MI. Understanding craniofacial anomalies: etiopathogenesis of craniosynostoses and facial clefting. Wiley-Liss, 2002.

Msamati BC, Igbibi PS, Chisi JE. The incidence of cleft lip, cleft palate, hyrocephalus, and spina bifida at Queen Elizabeth Central Hospital, Blantyre, Malawi. Cent Afr J Med 2000; 46:292–296.

Mulliken JB, Glowacki J. Hemangiomas and vascular malformations in infants and children: a classification based on endothelial characteristics. Plast Reconstr Surg 1982; 69:412–420.

National Institutes of Health Consensus Development Conference Statement (1988), Neurofibromatosis. Arch Neurol 1988; 45:575–578.

Nicole M, Wymann E, Hölzle A, Zachariou Z, Iizuka T. Pediatric craniofacial trauma. J Oral Maxillofac Surg 2008; 66:58–64.

Nthumba PM. The donkey, the fox, the gun and a lip. Internet J Surg 2005; 6(2). Available at: http://www.ispub.com/ostia/index.php?xmlFilePath=journals/ijs/vol6n2/lip.xml.

Nthumba PM. Bilateral thigh flaps: a case report and review of literature. East & Cent Africa J Surg 2007; 12(2): 82-87.

Nthumba PM. Giant pyogenic granuloma of the thigh: a case report. J Med Case Reports 2008; March 31(2): 95.

Nthumba PM. "Blitz surgery": Redefining surgical needs, training and practice in Sub-Saharan Africa. World J Surg 2010; 34(3):433–437.

Nthumba PM, Bird G. Marjolin's ulcer in a spina bifida patient. A case report. East & Cent Afric J Surg 2010; 15(2):127-129.

Nthumba PM, Carter LL. Visor flap for total upper and lower lip reconstruction: a case report. J Med Case Reports 2009; June 9(3): 7312.

Odeku EL, Martinson FD, Akinosi JO. Craniofacial fibrous dysplasia in Nigerian Africans. Int Surg 1969; 51:170–182.

Onyango JF, Noah S. Pattern of clefts of the lip and palate managed over a three year period at a Nairobi hospital in Kenya. East Afr Med J 2005; 82:649–651.

Posnick JC, Costell BJ, Tiwana PS. Pediatric craniomaxillofacial fracture management. In: Miloro M, ed. Peterson's Principles of Oral and Maxillofacial Surgery. BC Decker, 2004, Chap 27, Pp 527–545.

Rockson SG. Lymphedema. Am J Med 2001; 110:288–295.

Ross JH, Kay R, Yetman RJ, Angermeier K. Primary lymphedema of the genitalia in children and adolescents. J Urol 1998; 160:1485–1489.

Ruggieri M. The different forms of neurofibromatosis. Childs Nerv Syst 1999; 15:295–308.

Sørensen JL, Jørgensen B, Gottrup F. Surgical treatment of pressure ulcers. Am J Surg 2004; 188:42S–51S.

Suleiman AM, Hamzah, ST, Abusalab, MA, Samaan, KT. Prevalence of cleft lip and palate in a hospital-based population in Sudan. Int J Paediatr Dent 2005; 15:185–189.

Tessier P. Anatomical classification of facial, cranio-facial and latero-facial clefts. J Maxillofac Surg 1976; 4:69–92.

Viljoen DL, Versfeld GA, Losken W, Beighton P, Optiz JM, Reynolds JF. Polyostotic fibrous dysplasia with cranial hyperostosis: new entity or most severe form of polyostotic fibrous dysplasia? Am J Med Genet 2005; 29:661–666.