Developmental field reassignment in unilateral cleft lip: Reconstruction of the premaxilla

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In the 21st century the greatest stimulus for progress in cleft surgery will come from more a more accurate model of facial development and how clefts originate. Victor Veau[1] accurately predicted this: “All cleft surgery is merely applied embryology.” The revolution in developmental biology has not yet been incorporated into surgical practice. The drawings and terminology used in textbooks today are based on the work of Wilhelm His in the 1870's.[2] Cleft repairs are therefore designed based on the anatomy as it appears in the newborn. Measurements are taken and the tissues are geometrically manipulated. But the anatomy of a cleft as seen at the time of birth is far different from its original configuration in the embryonic face. From its onset at gastrulation, the clefting event unleashes pathologic processes that predictably alter the original embryonic anatomy over time to produce what we recognize as a cleft lip. Left uncorrected, these processes will remain operative throughout facial growth. This explains why geometric cleft repairs relapse over time, requiring revision.

Developmental field repair (DFR) is based upon the neuromeric model of craniofacial embryology.[3-4] The goals of DFR surgery are: (1) resolution of all pathologic processes of clefting (deficiency, displacement, division and distortion); (2) dissection along embryonic separation planes (subperiosteal release); (3) preservation of blood supply to the alveolar mucoperiosteum; (4) primary unification of the dental arch; and (5) reassignment of all developmental fields to their correct relationships. Before describing the surgical technique, certain basic concepts of field theory should be understood.

Craniofacial anatomy results from the assembly of specific, identifiable developmental fields. Fields are discrete units composed of ectoderm, mesoderm, endoderm and neural crest. At each level of the embryo all germ layers specific to that level can be referenced back to developmental zones of the neuraxis known as a neuromeres. These are named using the nomenclature of neuroembryology. The master plan of the entire embryo is determined by 6 prosomeres (prosencephalon-forebrain), 2 mesomeres (mesencephalon-midbrain), 12 rhombomeres (rhomboencephalon-hindbrain) and 31 myelomeres (spinal cord).[5-7]

RUBENSTEIN

Applications of neuromeric anatomy provide a potential embryonic “map” of all craniofacial structures with important implications for diagnosis and surgery. Exclusive of the cranial base (basiphenoid and posterior) and parietal bone, the craniofacial skeleton is made exclusively from neural crest. Thus the cell populations producing the ethmoid, presphenoid, premaxilla and vomer all originate in antero-posterior order from the neural folds in genetic register with the 1st rhombomere (abbreviated r1). The inferior turbinate, palatine bone, maxilla, alisphenoid (greater wing) and zygoma arise from the neural crest of the 2nd rhombomere (r2). The squamous temporal, mandible, malleus and incus are r3 neural crest bones.

Non-neural crest craniofacial bones come from paraxial mesoderm (PAM) lying immediately adjacent to the neural tube. PAM is divided into individual units shaped like popcorn balls called somitomes, each one in register with its corresponding neuromere. The first seven somitomes (Sm) contain the myoblasts for all muscles of the orbit and the first three pharyngeal arches. For example, the
mandible comes from r3 neural crest and all muscles originating from it arise from Sm3. Beginning with Sm8 all somitomeremes undergo a further transformation into somites, each having a dermatome, myotome and sclerotome. The first four somites (derived from Sm8-Sm11) produce the cranial base posterior to the sphenoid, the muscles of the tongue and part of the sternocleidomastoid and trapezius. These are called occipital somites.

Disturbances at a particular neuromeric level can affect individual or multiple fields to be deficient or absent. Thus, isolated cleft palate (unassociated with cleft lip) represents a deficiency state of the vomer. This occurs as a spectrum. As the vomer is progressively smaller, it lifts away from the plane of the palatal shelves and the cleft extends forward toward the incisive foramen. In mild cases of cleft palate associated with Pierre Robin sequence, a reduction in the horizontal plate of the r2 palatine bone is seen. Soft palate muscles are consequently normal but divided. As the pathology worsens, reduction in the horizontal plate of the r2 maxilla creates the well-known “horse-shoe” palate cleft. Submucous cleft palate, on the other hand, involves pathology in the 3rd pharyngeal arch. Somitomeremes 6 and 7 contain the myoblasts of levator, uvulus, palatopharyngeus and superior constrictor. These can be globally affected. Frequently, persistent VPI follows a seemingly simple palatoplasty, requiring further surgery. Failure to stratify cleft palate by embryologic mechanism explains much of the confusion currently extant in the speech and surgical literature.

Finally, Treacher-Collins syndrome provides an example in which all r2 developmental fields of the midface are affected: the maxillary, palatine and zygomatic bones are all small. The septum, vomer and premaxilla (being r1 structures) are unaffected. For this reason, the central midface projects normally while the dimensions of the palate, maxilla and zygoma are constricted.

Developmental fields form in a specific spatio-temporal sequence. Each one builds upon its predecessors. Making a face is much akin to assembling a house with magical pieces of Lego®, each one of which will grow over time. Imagine a Lego house made from 20 pieces (4 on the floor and 5 stories high). All pieces are growing independently. If a cornerstone piece is removed, the 19 remaining pieces undergo a deformation and the house tilts into the deficiency site. The missing Lego piece in cleft lip is the premaxilla. The physiologic basis of DFR is the reconstruction of the premaxillary field. This chapter presents the reconstructive application of these principles to the surgery of labiomaxillary clefts.

The pathologic anatomy of cleft formation
The pathologic anatomy of unilateral and bilateral labiomaxillary clefts stems from a tissue deficiency state localized to the lower lateral piriform fossa. The tissue at fault is the mesenchyme of the ipsilateral premaxilla. Neural crest stem cells responsible for synthesis of the presphenoid and ethmoid arise from the mesencephalic neural folds and are genetically identified with the 1st rhombomere (r1). Note that the basisphenoid is not neural crest in origin; it comes from paraxial mesoderm from somitomere 1 (Sm1). Sm1 lies just outside the neural tube at level r1 and is in register with it. Immediately caudal to this population are neural crest cells immediately above the rostral rhomboencephalon in register with the 2nd rhombomere (r2). The most rostral zone of r2 (herein referred to as r2’) is the likely source material for the vomer and PMx. The more caudal zone of r2 produces inferior turbinate (IT), palatine bone (P), maxilla (Mx), alisphenoid (AS) and zygoma (Z).

These cell populations migrate forward into the developing face in a strict temporospatial order. The sphenoid is laid down first, followed by the ethmoid. In like manner, formation of PMx is the prerequisite for the appearance of V. Formation of the PMx and V requires the pre-existence of the perpendicular plate of the ethmoid (PPE). The function of the PPE is to provide a cellular scaffold by which r2’ neural crest cells can reach the midline. [10] Figures 1-2] In holoprosencephaly (HPE) the PPE can be absent. PMx and V cannot develop correctly; a wide bilateral cleft results.[11-13]

The piriform fossa of humans and some high primates is assembled as the fusion of the frontal process of the premaxilla (PMxF) and the frontal process of the maxilla (MxF). In all other vertebrate skulls PMxF and MxF are readily visible as two distinct entities.[12] Evolution foreshortened the human snout. The two fields became superimposed, PMxF becoming telescoped internal to MxF. This lamination is responsible for the strength of the piriform rim. Plating of the piriform “buttress” in fracture repairs takes advantage of this bicortical anatomy.

**PIRIFORM**

The developmental field in which the PMx resides consists of an epithelium and a mesenchyme. Formation of the PMx results from interactions between these tissues. The
Figure 1: Developmental field map of the face. All neuromeres are color coded. Neural crest cells forming ethmoid, presphenoid, vomer and premaxilla share common embryologic origin from 1st rhombomere (violet). Maxilla, zygoma, alisphenoid come from 2nd rhombomere (blue). Nostril sill is union of r1 and r2. All craniofacial sutures contain neural crest cells. Craniofacial fields are neuroembryologic and follow suture boundaries.

Developed has several anatomic subcomponents, these are assembled in a strict sequence. In dental terminology the central incisor is called “A” and the lateral incisor is called “B.” A erupts before B. Therefore the central incisor field (PMxA) is biologically “older” than the lateral incisor field (PMxB). Neural crest mesenchyme flows forward along the previously-established PPE. It first encounters the epithelium corresponding to PmxA and then “spills over” into zone PMxB. The time sequence of dental eruption (central incisor A > lateral incisor B) is a manifestation of the relative biologic “maturity” of the mesenchymal field within which each tooth develops. The frontal process field (PMxF) is a vertical offshoot of PmxB; this subfield is the biologically “newest” tissue.

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Pathology affecting the PMx occurs as a spectrum based on this original developmental pattern. A deficiency state of the PMx will first occur in the most distal aspect of the frontal process (ie. at its most cranial extent). As the mesenchymal deficit worsens, frontal process will be reduced in a cranial-caudal gradient. “Scooping out” of the piriform rim results; the nasal lining is pulled down as well. This causes depression of the alar base and a downward-lateral displacement of the lateral crus. Biologic signals from PMxF regulate epithelial stability and therefore affect lip formation (vide infra). When the signal strength is minimally disturbed the lip is normal despite the piriform distortion. Therefore the former frustre manifestation of premaxillary deficiency is a cleft lip nose with a perfectly normal lip.[18]

Once the frontal process is eliminated, the deficiency state shows up in the lateral incisor field. Progressive degrees of PMx deficiency in the lateral incisor field cause incremental loss of alveolar bone. Normal alveolar development follows a gradient. It begins at the incisive foramen and progresses forward. Mild deficiency causes notching on the labial surface. As the deficiency worsens the notch deepens backward toward the incisive foramen. A critical lack of alveolar bone mass results in outright failure of lateral incisor development.

BMP-4 signals from this field are directly implicated in the mechanism of fusion between the lateral lip element and the prelabium.[17-18] BMP-4 emanating from PmxL forms a cranio-caudal chemical gradient. The strength of this gradient depends upon the total amount of available BMP-4; this in turn is proportional to the overall mesenchymal mass of PMXB. Reduction in mass of the lateral incisor field results in a diminution of the total BMP-4 signal. Lip fusion follows this same gradient. Mild weakness of the BMP-4 gradient will result in notching of the vermilion. As the situation worsens the extent of the lip cleft worsens in a cranial direction. The clinical spectrum of the so-called minimal cleft lip deformity faithfully reproduces this biologic sequence.[16]

In summation, variations in clefts involving the primary palate and the lip can be understood as interactions between deep plane fields of the premaxilla, the maxilla and superficial plane field of the lateral lip element. The mesenchyme of the lip has a different embryologic origin. It is genetically identified with the 2nd rhombomere. Neural crest cells from r2 provide the dermis and the subcutaneous tissue of the alar base, while paraxial mesoderm from the 2nd somitomere forms the anterior half of the squamous temporal bone and the cranial half of the parotid gland. All derivatives from level r2 can be mapped out within the sensory distribution of V2, the nucleus of which resides within r2.[19-24]

The developmental anatomy of the nose, prelabium and premaxilla has been previously described in terms of neurosensory theory by this author.[3-4,8,9] In contrast to the rest of the body, all dermis and submucosa of the head originates from neural crest cells, not from a dermatome associated with a somite.[25] Pre-dermal neural crest arises from three distinct zones of the embryo. The dermis of the forehead, nose and vestibular lining come from the caudal prosencephalon (above somites p4-p1). This prosencahalic neural crest (PNC) migrates forward like a gigantic glacier to occupy the neural folds above somites p6 and p5. The alar half of p6 and p5 creates the telencephalon (cerebrum). The basal halves of p6 and p5 plus all remaining somites (p4-p1) synthesize the diencephalon (epithalamus, thalamus and hypothalamus). The neural folds above p6 and p5 are “sterile.” They contain the pituitary, olfactory and optic placodes, but no neural crest. When PNC flows forward into this zone the placodes are activated and dermis is formed. Nasal vestibular lining from the cribriform plate forward to the internal nasal valve comes from p6 PNC. All remaining frontonasal dermis from the internal nasal valve forward to the hairline comes from p5 PNC.

PNC skin shares sensory innervation with the dura of the underlying frontal lobe. V1, the sensory nerve for this common zone has its nucleus with the 1st rhombomere (r1). The neuroanatomy is analogous for the rest of the face. Rhombomeres r2 and r3 contain neurons supplying all skin and dura innervated by V2 and V3. This is the embryologic basis for the treatment of migraine headaches with Botox® injected into peripheral trigger points.

Design of surgical incisions for cleft repair follows this embryology. The boundary between these two fields is sharply demarcated within the nasri. The skin of the anterior columella and philtrum is thus a p5 derivative. This skin is supplied by terminal branches of the internal carotid artery, the anterior ethmoid arteries and innervated by V1.[3] The skin making up the floor of the nose has a different origin. It extends from the base of the columella laterally and makes contact with the alar base. The medial (terminal) branch of the sphenopalatine artery innervates this skin. The innervation is from V2 and
is shared with the ipsilateral incisors. Continuity between the p5 skin of the lateral columella and the r2’ skin of the nasal floor makes possible the elevation of a skin-cartilage flap containing the medial crura with long skin flaps. Many years ago Vissarianov described this flap as a means of secondary cleft reconstruction.\textsuperscript{[26-27]}

Based on signals from the underlying premaxilla, the r2 alar base produces a tongue of tissue that makes contact with the r1 lateral prolabium. This skin bridge sets up the floor of the nose. It also provides a mechanical platform by which mesenchyme from the lateral lip element can make contact with the p5 mesenchyme of the prolabium. Lip closure thus occurs. This process involves mesenchymal “flow” from the lateral lip element toward the prolabium. For this to occur, the epithelium covering the lateral lip element, the r2-r1 skin bridge and prolabium must undergo a genetically-induced breakdown. BMP-4 produced by the premaxillary field causes de-repression of Sonic hedgehog (SHH), a gene localized to these overlying epithelia. The protein product of SHH causes epithelial breakdown. Thus, absence or deficiency of an appropriate BMP-4 signal will lead to restricted expression of SHH, abnormal persistence of epithelium and failure of mesenchymal fusion.\textsuperscript{[28-29]}

The volume of the PMx determines whether or not lip closure can occur. First, a small premaxillary field manufactures small amounts of BMP-4. The amount of BMP-4 produced is critical to produce the epithelial breakdown necessary to permit mesenchymal merger. Second, when the premaxilla is too small, the physical distance between it and maxilla will exceed a critical dimension. Epithelial bridge formation between the alar base and the prolabium cannot occur. If this critical distance exists at the level of the incisive foramen, a cleft of the secondary palate will form. This is because the horizontal repositioning of the palatal shelf from the maxilla must make contact with the vomer just posterior to the incisive foramen. The process is just like a zipper. If initial contact is not made, fusion of the palatal shelf to the vomer cannot take place. Even if initial contact is made, a secondary palatal cleft can still result due to displacement of the vomer away from the midline. The vomer can become warped by the inequality of growth forces on either side of the cleft. Thus, the zipper may get started anteriorly but, as the process proceeds posteriorly, when it encounters the deviated vomer, a palatal cleft will open up.\textsuperscript{[3]}

Reconstruction of the premaxilla

Cleft surgery that does not reconstruct the missing PMx does not solve the biologic problem. The pediatric face is a set of Lego\textsuperscript{8} pieces, all growing over time. If one piece of the set is missing, with subsequent growth the remaining pieces will collapse into the deficiency site. Only when all the Lego pieces are in place can harmonious facial development occur.

The premaxillary developmental field consists of lining (present in its totality) and mesenchyme (missing). Lining can be recreated by: (1) subperiosteal dissection of the primary palate (r2’ premaxilla and r2 maxilla); (2) subperiochondrial dissection of the p6 septum (sufficient to reduce the septum into the midline); (3) subperiosteal elevation of r2’ vomer to close the nasal floor (sufficient to reduce the septum into the midline); and (4) subperiosteal rescue of the r2’ nasal floor skin from the lateral prolabium. When these flaps are elevated and sutured, the primary palate becomes a box lined with neural crest stem cells, all of which carry membrane-bound BMP receptors.\textsuperscript{[29-31]}

Mesenchymal replacement can be undertaken using two basic mechanisms. Autogenous bone graft from rib or hip provides mesenchymal cells originating from paraxial mesoderm. Incorporation of the graft into the primary palate occurs by osteocondensation. Implantation of recombinant human bone morphogenetic protein-2 (rhBMP-2) in combination with an activated collagen sponge (ACS) results in morphogen-based recruitment of stem cells from the environment into the sponge. Stem cell concentration and differentiation into osteoblasts results in the formation of bone native to the site. This process is known as osteoinduction.\textsuperscript{[32]} Extensive pre-clinical work by Boyne demonstrated the ability of rhBMP-2 to form membranous bone, including reconstitution of surgically-created cleft palate defects in primates.\textsuperscript{[33-34]} Studies demonstrating efficacy in maxillary lift combined with absence of donor site morbidity resulted in FDA approval for this indication.\textsuperscript{[35]}

The technique of facial bone reconstruction using rhBMP-2/ACS implantation is known as in situ osteogenesis (ISO) and has been previously reported by this author and co-workers.\textsuperscript{[36]} Resynthesis of a 12 cm mandibular defect in a 9 year old demonstrated the ability of ISO to function effectively outside the range of blood supply associated with a critical-size defect.\textsuperscript{[37]} BMP-2 mediated osteoinduction is accompanied by extensive recruitment of blood vessels from local environment.\textsuperscript{[38]} For this reason selected bone defects can be resynthesized using ISO alone, without recourse to microsurgical tissue transplantation.
Distraction techniques have been successfully applied to ISO-regenerated bone. In a number #7 lateral cleft with a foreshortened mandibular body and absent ramus, distraction of the recipient periosteal chamber in a posterior and superior direction permitted synthesis of 3.5 cm of mandibular ramus with eventual articulation with the skull base via a pseudoarthrosis. Distraction-assisted in situ osteogenesis (DISO) will be an alternative treatment to rib grafts in the reconstruction of the Pruzansky III mandibular defects in craniofacial microsomia. The bone produced will be membranous. The dissection is less problematic. There is avoidance of unpredictable growth of the rib graft outside the natural periosteal environment. Finally, the chest wall donor site is obviated. Histology of ISO-produced membranous bone is indistinguishable from that of the recipient site in both mandible and maxilla.

Alveolar clefts are lined with mucoperiosteum containing neural crest stem cells. Blood supply is excellent and the dimensions modest. For this reason, this author and co-workers reported 50 alveolar clefts successfully treated with rhBMP-2. Precise surgical technique of implant placement and of soft tissue

**Figure 2:** Developmental anatomy of premaxilla. Spatio-temporal distribution of neural crest (arrow) demonstrates frontal process as most “recent” derivative. Clefting affects the premaxillary fields in reverse order; piriform fossa deficiency is therefore the “forme fruste” manifestation of a cleft.

**Figure 3:** Medial and lateral sections of the nasal cavity demonstrate the neuromeric origins of the bone fields. The biosynthetic pathway of the r2’ MNC lays down the premaxilla and vomer along the axis of the nasopalatine nerve and the medial branch of the sphenopalatine artery (the absolute terminus of the external carotid). Fields affected by the Tessier clefts are indicated by yellow stars, eg. the #3 cleft is a selective knock-out of the inferior turbinate. Because the p5 lacrimal bone is built upon it the #3 cleft destroys the lacrimal system. The cleft premaxilla is zone #2.
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Closure was emphasized in the current series of 200 cleft sites. Long-term outcome of 43 cleft sites was assessed at one year post grafting with 3-D CT. The series was comprised of 23 unilateral clefts (6 primary and 17 secondary) and 12 bilateral clefts (3 primary and 9 secondary). Complete transverse fill (unification of the dental arch) was achieved in all cases. Vertical fill was improved to 75-100% when an inert bulking agent (tricalcium phosphate) was included. For these reasons, primary cleft repair using developmental field reassignment technique can ideally be combined with ISO to achieve primary unification of the dental arch without donor site morbidity.

Philosophy of Developmental Field Reassignment (DFR): the 4 D’s
Faulty embryogenesis of the premaxillary field affects the position and shape of surrounding fields in four distinct ways: Division of force vectors causes unequal growth, Deficiency-related collapse of partner fields into the void,
The execution of DFR surgery is based upon the concepts of Sotereanos. Its guiding principles are five. (1) Correct all pathologic states in the first operation (as above). (2) Respect embryonic developmental planes during dissection, i.e. subperiosteal release of the soft tissue envelope for a tension-free closure. (3) Conserve blood supply to the periosteum, safeguarding it for future membranous bone synthesis. Fourth, align and unite the dental arch into a normal relationship. Fifth, reassign all developmental fields into correct relationships with each other. When properly executed, these principles result in the restoration of the functional matrix. When all the bone-forming soft tissue fields are spatially correct, all force vectors exerted upon bone will be correctly aligned. Subsequent growth of the face is directed back toward the normal.

A new algorithm: Is lateral nasal wall deficiency relative or absolute?
In keeping with Victor Veau, cleft repair is a constant exploration of nature’s experiments. Developmental field reassignment surgery is deliberately designed to address a tissue deficit of the lateral nasal wall and alveolus, the product of a congenitally small premaxillary field. That the lateral crus be entrapped cannot be in doubt. Its release into a normal position occasions a triangular tissue gap that must be filled. Proper airway reconstruction is the name of the game. At the same time, the alveolar cleft (a six-sided box) must be reconstructed. This requires flap coverage for its nasal surface (the “top” of the box). Can these two goals be accomplished with the same tissue? A skin graft alone (particularly anterior auricular skin and cartilage) will effectively support lateral alar crus advancement. It cannot provide vascular coverage for the alveolar cleft “roof.” At first blush, the LLC-NPP flap would seem big enough to accomplish both goals. After 7 years of work with this technique this author has concluded that paring from the prolabilum is not sufficient. Premaxillary field soft tissues are not only relatively deficient from the lateral nasal wall: an absolute tissue deficit exists. Use of the premaxillary tissue mismatched to the prolabilum is not always sufficient to solve the problem. The new algorithm of DFR surgery now calls for optional composite grafting of the lateral wall, followed by elevation and transposition of the LCC-NPP flap across the nasal surface of the alveolar cleft. The decision to graft is based upon the relative size of the defect versus that of the flap.

SURGICAL TECHNIQUE OF DFR
This operation consists of markings, a five-step dissection sequence and a five-step closure.

Preparation and marking
The DFR operation is comprehensive, providing simultaneous correction of the lip, nose and primary palate. A more extensive dissection is required. The
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**Figure 7:** Four-step nasal incision sequence achieves advancement of both medial and lateral crura. I have recently modified the infracartilagenous incision, making it discontinuous along the lateral crus to minimize contracture. **Nasal tip is reconstructed by simultaneous elevation of both the medial and lateral crura.** The Delaire sutures are shown. #1 is placed in the levator insertion into the SOO one finger-breadth lateral to the alar base. This centralizes the lip-cheek flap. The non-cleft side is done first to the anterior nasal spine (a drill hole is helpful), #2 is placed in the nasalis and is optional (decision made at the end of the case). It adjusts alar base position and nostril sill curvature. #3 suspends the oblique head of SOO from the columellar base. It establishes the “aesthetic drape” of the lip operation.

The patient takes longer to perform (about 3-4 hours for a unilateral cleft). For this reason, patients come to surgery at 4-6 months of age. Prior to operation the dental arch is prepared with splinting (a form of infant presurgical orthopedics); this is begun as early as two weeks after birth. The emphasis of presurgical splinting is: (1) promoting anterior growth of the retro-displaced cleft maxilla; and (2) maintenance of the space in the alveolar cleft. If satisfactory maxillary shelf repositioning does not result by age 3 months, a lip adhesion procedure is carried out. When satisfactory dental arch correction is achieved, DFR repair is performed. This usually occurs 3-4 months after the lip adhesion.

The patient receives antibiotics and corticosteroids for swelling. A central V2 block is performed at the pterygopalatine fossa using 0.25%Marcaine® (bupivacaine hydrochloride). Approximately 3-5 cc per side is sufficient (maximum dose for 0.25%bupivacaine being 1 cc/kg). At the end of the case, the central block is reinforced by blocking the infraorbital nerves with bupivacaine, 1-2 cc per side. The child returns to recovery pain-free. The initial block ensures that substance P (a critical mediator in the pain cascade released in response to surgical trauma) will not be produced. In the absence of substance P the entire postop pain response is altered.

The surgical fields of the unilateral cleft are defined as follows. **FIGURE (In neuromeric terms: A = p6 (red) + p5 (turquoise), B = r2‘ (pale gold), C is r2 (yellow) and D is r2 + r4 (yellow + orange). The prolabium is divided into two zones. Zone A is the true philtrum; it measures**
Figure 8: NPP-LCC flap has dual blood supply. Behind the surgical tape are: (1) lateral branches from the ipsilateral anterior ethmoid artery of the columella; and (2) vessels running along the dorsal surface of the lower lateral cartilage that anastomose the nasopatineal artery with the lateral nasal artery. In front of the tape, the nasopatine (medial sphenopatine) artery emerges from the septopremaxillary junction. This tissue, otherwise called the B flap, reconstructs the nasal floor (roof of the alveolar cleft) and the lateral nasal wall. This corresponds to the missing premaxillary field.

Figure 9: Dissection of alveolar cleft. Elevation and division of mucoperiosteal flaps provides closure of nasal floor and oral aspect of primary palate. R2’ premaxillary flaps (violet) are sutured to r2 maxillary flaps (yellow), providing 4 walls of the box. The cleft-side sliding sulcus flap closes off the front of the box. Backwall closure occurs when vomerine mucoperiosteum is elevated and sutured to the palatal shelf MxP. Septum is reduced and secured to midline. Cleft site “box” filled with osteoinductive cytokine rhBMP-2 or autogenous bone graft. This reconstructs the missing premaxillary Lego® piece.

Figure 10a: Anterior closure of unilateral alveolar cleft using sliding sulcus flaps. On the cleft side two incisions are required. (1) A medial-to-lateral L-shaped releasing incision (red line) is placed in the gingival sulcus. It runs from the cleft lateral to the buttress. There, it ascends vertically to the zygomatico-maxillary junction. The subperiosteal flap is elevated widely up to the infraorbital foramen. Because the periosteum is an intact sheet, the flap is stiff, being tethered to the orbital rim. (2) A counter-incision (green line) is begun at the level of the nasal floor and brought transversely from the piriform to the buttress. This releases the lower (alveolar) mucoperiosteal flap. It freely translates two tooth units medially. This closes the anterior wall of the alveolar cleft.

Figure 10b: Bilateral sliding sulcus flaps are illustrated. Although I started out with this technique, I no longer do a gingival release on the non-cleft side. A one-sided release is sufficient. On the non-cleft side, wide subperiosteal mobilization is combined with a full-thickness vertical releasing incision up the buttress. This creates a huge, mobile bipedicle flap. The entire soft tissue complex becomes centralized without tension. Nota bene: Surgeons working under extreme condition (very wide clefts in remote areas with no access to orthodontic management) bilateral sliding sulcus flaps are very effective. They will close any size defect.

D. It contains a sphincter layer, the deep orbicularis oris (DOO) and the dilator layer, the superficial orbicularis oris (SOO) muscles. DOO develops in conjunction with the oral mucosa while SOO develops with the skin. A layer of fat conveniently separates these two layers.²⁷⁻⁴⁸

The true destination of NPP soft tissue is in the nasal...
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Figure 11 (Case1): Bilateral cleft lip with right unilateral cleft of primary palate and bilateral clefts of secondary palate. Soft tissue relationships one year after DFR demonstrating stability of nasal tip projection. Comparison of CT scans at 3 months and 9 months show bony union of primary palate. Periapical view of alveolar cleft site shows reconstitution of the periodontal ligament, a neural crest structure. This is not seen with conventional iliac crest graft. Serial 3-D skulls taken 9 months apart support the Delaire dissection concept: wide subperiosteal undermining repositions the osteogenic soft tissue fields into a correct, centralized relationship. Subsequent osteosynthesis of membranous bone now occurs in a correct centric distribution. Developmental field reassignment promotes normal membranous bone formation after the surgical procedure.

floor and the lateral nasal wall. NPP represents the soft tissue envelope corresponding to the lateral zone of the premaxilla, PMxB + PMxF. Nasal floor soft tissue corresponds to the lateral incisor field, PMxB. Soft tissue of the lateral nasal wall between the inferior turbinate and

the lateral crus corresponds to the frontal process field, PMxF. Lateral wall soft tissue corresponds to the frontal process field, PMxF. Nasal floor soft tissue corresponds to the lateral incisor field, PMxB. The rationale of DFR surgery is to reassign the misplaced fields of the premaxilla.
into correct position. When the NPP flap is combined with the reflected mucoperiosteum from the margins of the bony cleft a surgical “pocket” is created. This site is subsequently filled with bone graft or a bone-producing cytokine (rhBMP-2) such that missing premaxillary fields are re-synthesized.

The NPP flap functions just like a “boxtop” to cover the alveolar cleft. It is almost always adequate. I am convinced that the “secret identity” of NPP is the original PMxB. Because of the cleft this tissue is “shipwrecked” on the premaxilla. Unfortunately, NPP is frequently insufficient to replace PMxF. Every time the lateral crus is released and advanced a soft tissue defect appears. The defect represents missing PMxF. In my clinical experience, this is almost always the size of the auricular cymba. A composite graft of anterior auricular skin and cartilage from the cymba is the most reliable replacement for this tissue. The cymba graft, in combination with the NPP flap, provides more than enough tissue to reconstruct the missing premaxilla.

Markings are carried out using a modification of the Millard system. (Paired anterior ethmoid neurovascular bundles define the true philtrum; this equals the width of the columella.). First to be marked is point 2, the junction of the normal philtral column with the white roll. The width of the columella at its base is then measured. This distance (usually 6-10 mm) is subsequently marked along the white roll medial to the philtral column as point 3. Distance 2-3 is the width of the philtrum; this contains the two anterior ethmoid arteries (approximately 4-6 mm apart). The midpoint of the philtrum (point 1 in the Millard system or nadir of cupid’s bow) is irrelevant. Point 13, defined by the bulge of the footplate of the medial crus on the non-cleft side, marks the “shoulder” of the columella.
Developmental field reassignment in unilateral cleft lip

The corresponding point 12 on the cleft side will be found displaced caudally and internally with respect to point 13. Total philtral height 2-13 should equal 3-12. Points 4 and 10 mark the midpoint of the alar bases on non-cleft and cleft sides, respectively.

On the lateral side, points 9 and 11 mark the terminus of skin within the nasal cavity. This is located just anterior to the inferior turbinate. Point 9 is the tip of flap D while point 11 is the tip of the future nostril sill C', so-called because it is in continuity with the alar base C. The dimensions of C' can be roughly mapped out by measuring the nostril sill on the non-cleft side. This is the distance from the midpoint of the non-cleft alar base (point 4) to the ipsilateral footplate point 13. Point 8 marks the natural transition of the white roll. Distance 8-9 should match 3-12.
Dissection sequence
Step 1. Lateral dissection: rescuing the nostril sill
The lateral lip is tensed with a single hook and the skin-mucosa margin is incised proceeding upward from 8 to 9, located just below and anterior to the inferior turbinate. From here the incision swings around laterally to 10, the midpoint of the alar base, (but not beyond it at this point). In this manner, the lateral lip flap D is separated from the alar base C. This step separates orbicularis from the nasalis. From point 9, a second, more internal, incision defines the triangular flap C’. The base width of C’ can be deduced by measuring the dimensions on the non-cleft side.

From the lip a lateral vermilion flap (L) is pared off and dissected down to the alveolar cleft margin. Proper paring of L includes a small strip of lip skin 2-3 mm wide because it is “rolled-in,” with an abnormal relationship to the underlying muscle. If the surgeon does not do this, the skin will pucker inward at the final closure. The L flap is the most optimal tissue of the entire DFR operation. It is best brought medially and interposed between the nasal vestibular lining and the B flap. It can also be used as a free graft to the lateral nasal wall. No tissue is sacrificed in DFR.

With its vermilion stripped off, the lateral lip margin is now entered and the orbicularis is split into its deep (constrictor) and superficial (dilator) components. The deep orbicularis oris (DOO) is shaped like the letter J and separated from the superficial orbicularis oris (SOO) by a layer of fat. The caudal margin of the two muscles defines the white roll and contains the labial artery.\(^{[47]}\)

Step 2. Medial dissection: the NPP-lateral columellar advancement flap
The medial margin of the cleft (zone B in our diagram) consists of probalabial tissue lateral to the true philtrum. This is in continuity with lateral columellar skin and the medial crus of the alar cartilage. Conveniently, the blood supply of these two units is a watershed permitting dissection of a very long flap. The B flap has two parts: (1) a skin/mucosa flap of the non-philtral philtrum (NPP) and (2) a chondrocutaneous flap containing the lateral columellar skin and medial crus (LCC). \[B = NPP + LCC\]

B has a rich blood supply. The NPP component is irrigated by the nasopalatine artery. The LCC component gets supply from 2-3 lateral branches of the ipsilateral anterior ethmoid. In addition, vessels in continuity with the lateral nasal system from the facial artery run along the surface of the alar cartilage itself. Once elevated, B is surprisingly long, reaching all the way across the cleft to the lateral nasal rim. [Figure B] is inset into the gap created by advancement of the p5 lateral vestibular lining (and the lateral crus). This achieves two important surgical goals: (1) cephalic advancement of the medial crus with increased tip projection; and (2) soft tissue augmentation of the lateral nasal wall. Inset of B permits release of the tethered lateral vestibular lining. Repositioning the lateral crus now occurs in what would otherwise be a Y-V pattern without the Y-V closure. Natural alar cartilage anatomy is accomplished without pinching together an already-deficient lining.

The future cleft-sided philtral column is determined by understanding the embryology of the prolabium. The true philtrum lies between points 2 and 3 and consists of two fields (each supplied by a separate branch of the anterior ethmoid artery). The philtral dermis comes from p5 prosencephallic neural crest and is innervated by V1. The width of the philtrum is roughly equal to that of the columella, generally measuring 6-10 mm. Because the philtrum develops on top of the r2’ premaxilla, it receives additional blood supply from the terminal branches of the sphenopalatine artery (SPA) emerging at the level of the septopremaxillary junction. Thus, the philtrum has a dual blood supply. All remaining prolabial skin and mucosa (the NPP) is an r2’ derivative, supplied by the nasopalatine artery and innervated by V2. The prolabium of a bilateral cleft thus consists of four developmental units based on
embryology, blood supply and innervation: two central philtral A fields and two lateral B fields.

The embryology of the columella is as follows. The anterior centrally-located skin comes from p5 and contains the paired anterior ethmoid arteries. On either side of that central swatch, the columellar skin extends backwards toward the septum. Just beneath the lateral columellar pillars lie the medial crura of the lower lateral cartilages (LLC). Because these pillars serve as the biologic "template" for the cartilage to form, dissection of skin away from the crura is extremely difficult. The LLCs are thus p5 neural crest derivatives. The upper lateral cartilages lie above the p6 vestibular lining and are therefore of p6 neural crest origin.
Correction of the cleft lip nose requires releasing the alar cartilage from two points of entrapment. The deficiency state of the premaxilla causes a mechanical deformation of perfectly normal p5 skin on both sides of the nasal introitus. In the lateral nasal wall, the r2’ skin overlying the premaxillary frontal process (PMxF) is reduced or absent. Consequently, p5 skin containing the lateral crus of the alar cartilage is dragged down into this “sinkhole” located just in front of the inferior turbinate. The lateral crus is therefore flattened. On the medial side, the deficiency of the r2’ premaxillary lateral incisor field (PmxB) creates a “sinkhole” into which the p5 lateral columnellar skin is displaced. This displaces the medial crus of the alar cartilage downward and inward compared with the non-cleft side.

The LCC-NPP flap is elevated as follows. Under tension, the NPP skin is elevated in continuity with the lateral columnellar wall. The lateral (internal) incision is made first, separating the skin from the mucosa all the way to the junction between the premaxilla and vomer. Although this incision will eventually be carried up in front of the septum, it is advisable to stop here. Tension can thus be maintained on the NPP while it is separated from the philtrum proper.

Next, a second incision, parallel to the first, ascends from point 3 to the base of the columnella. It then extends into the nose along the lateral sidewall of the columnella. Previously I would continue this incision along the side of the columnella (just anterior to the edge of the medial crus) and thence bring it directly into the nose as an infracartilaginous incision. This design made me concerned about possible scar contracture. At the suggestion of Dr. John Reinsch, I began to break up the incision by elevating a medially-based rectangular skin flap occupying the caudal half of the columnella. Just beneath the Reinsch flap lies the footplate of the lateral nasal cartilage. At its cephalic margin, the incision is resumed along lateral columnella. At the level of the intermediate crus the incision transitions beneath the soft triangle into a standard infracartilaginous incision and is then continued all the way to the piriform margin.

The anterior incision of the NPP-LCC flap is not sufficient to advance it. A second parallel incision is required. FIGURE It starts in the membranous septum about halfway up. It is then carried downward to the vomer and then re-directed forward to meet the lateral margin of the B flap. Note that the membranous septal incision provides immediate exposure of the cartilaginous septum.

The NPP-LCC flap now resembles a long boot, shaped like Italy. The “toe” of the boot extends along the prolabial margin. Beneath the “heel” of the boot lies the footplate of the alar cartilage. This landmark corresponds to point 12. Grasping the heel of B provides instant access to the medial aspect of the medial crus. This is a safe plane permitting dissection of the alar cartilage right into the nasal tip, with the following caveat. The B flap gets blood supply from the nasopalatine artery via 2-3 branches emerging at the junction of the premaxilla and vomer. Gentle spreading along the medial border of the cartilage will reveal these branches and preserve them. Additional blood supply descends along the skin. Formerly I would elevate these NPP-LCC flaps completely, never having an issue with ischemia. Now however, I think it prudent to preserve the nasopalatine branches when possible. This type of blunt dissection is more than sufficient to advance the nasal tip.

The septum is now dissected out and freed from the maxillary crest until it sits passively in the midline. As growth proceeds the centralized septum will no longer constitute an abnormal force vector tethering the nasal tip.

Step 3. Nasal dissection: “open-closed” rhinoplasty

At this juncture, a standard infracartilaginous incision is made. This incision is brought all the way to the piriform rim, following the natural fold between the nasal skin and the vestibular skin. The caudal extent of this incision terminates at the internal border of the triangular nostril sill flap B’. The exposure gained via the lateral columnella-infracartilaginous incision allows a complete dissection of the dorsal nasal skin envelope as described by McComb. The success of the McComb dissection is really an embryonic field separation. The deep layer comprises the p6 vestibular epithelium and p6 neural crest upper lateral cartilages, the blood supply to which comes from below via the ICA. The superficial layer is p5 nasal skin and the lower lateral nasal cartilages, the blood supply to which is also of ICA derivation. Interposed between these two layers, like a sandwich, is the SMAS layer of facial muscles derived from the 2nd pharyngeal arch (r4-r5). The myoblasts come from somitomere 4. The nasal muscles are compartmentalized by r4 neural crest fascia (the SMAS). The blood supply to this intermediate 2nd arch layer is from the facial artery (ECA). This mesenchyme provides an additional source of blood supply to the alar cartilages. Careful dissection of the lateral columnellar walls from the columnella discloses vessels running along the medial

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surface of the medial crura. These anastomose the lateral nasal vessels with the nasopanatine vessels.

Along the lateral nasal rim the dissection is carried right down to the piriform margin. The proper plane for separation is achieved by hugging the surface of the cartilage. The overlying r4 SMAS layer and the p5 skin layer are left in anatomic continuity. Very little bleeding is occasioned by this approach. As one reaches the piriform rim the periosteum is incised and stripped vertically. Subperiosteal elevation of the soft tissues lateral to the piriform rim preserves the facial artery arcade. The nasal skin envelope is then liberated cephalically all the way to the nasal bones.

Despite these maneuvers, the vestibular lining is still tight! The media crus has been completely released and advanced into the nasal tip but the lateral crus being remains splayed out and tethered. Releasing the lateral crus from the vestibular lining has been advocated in the past. This is technically difficult because it violates the embryology (recall that the alar cartilage arises as a neural crest response to a pattern embedded in the vestibular epithilium). The size and shape of the cleft-side alar cartilage has been demonstrated to be normal compared with the non-cleft side. Consequently, the p5 vestibular lining “program” must be normal as well. Studies at UNC demonstrate that the overall surface area of the repaired cleft nostril is reduced by 30%. The site and dimensions of the deficit correspond to the soft tissues of PMxBF.

The soft tissues of the alveolar cleft PMxBF can be found on the prolabium. The LCC-NPP flap effectively provides a vascularized “roof” for the bony cleft. Lateral nasal wall release “uncovers” a defect that frequently exceed the reach of LCC-NPP. The release begins at the inferior turbinate and continues along the junction of vestibular and nasal skin. At the apex of the lateral crus it becomes V-shaped. Due to the prior McComb dissection the lateral crus is advanced cephalically into symmetry with the normal side. The internal nasal valves will appear equal. Left behind is a raw spot roughly the size of the ear cymba. A composite graft using anterior skin and cartilage from the cymba provides a sturdy reconstruction for the airway. The LCC-NPP flap, inset without tension across the alveolus, comes to rest at the foot of the cymba graft.

Exit the turbinate flap

As the DFR evolved, I employed various solutions to patch the defect created by the vestibular release. One of these solutions made use of an anteriorly-based inferior turbinate flap as described by Noordhoff. Although this tissue worked adequately, I had several reservations about it: (1) the dissection is subtle and difficult to teach; (2) the tissue type is distinct and not native to the nasal rim; and (3) healing of the donor site can be accompanied by crusting and bleeding; and (4) it did not make embryologic sense. Proper dissection and inset of flap NPP combined with use of a composite for FTSG made the turbinate flap unnecessary. The key to getting the most out of NPP is to include the prolabial vermilion with the skin.

Step 4. Intraoral dissection: sliding sulcus S flap

The rationale and design of the sliding sulcus mucoperiosteal flap stem directly from pioneering work at the University of Pittsburgh by Soteranos. This technique involves a gingival release on the cleft side carried out from the alveolar cleft to the buttress. This permits wide subperiosteal dissection over the entire face of the maxilla below the infraorbital foramen as described by Delaire. A 45 degree backcut up the buttress is performed. The attached gingival is released. The flap is covered on its undersurface by a sheet of peristemum, rendering it rather stiff. The Soteranos maneuver is a mobilization of the S flap using a counter-incision in the periosteal sheet itself parallel to the gum line. The counter-incision, located half-way from the gingival margin to the infraorbital foramen, is made by just scoring through peristemum. It extends from the piriform margin straight lateral to the buttress. At this point it joins up with the previous backcut. These two incisions make a right angle: the periosteal counter-incision is transverse across the maxilla and the buttress incision is vertical. The combination of these two incisions releases the lower mucoperiosteal flap S from the upper mucoperiosteum attached to the orbit.

When S is released, it advances mesially about two tooth units. The margin of S that was freed from the lateral border of the cleft now extends across the alveolar cleft without tension and is sutured to the mucoperiosteum of the premaxilla. In this way, like a sliding door, the S flap seals up the anterior aspect of the alveolar cleft.

On the noncleft side, a similar subperiosteal dissection is done without recourse to a gingival release. A large bipedicle flap is created, permitting centralization of the previously lateralized soft tissue envelope. Note that when cleft lip occurs without an alveolar cleft, bilateral S flaps, elevated without gingival release, permit proper tension-free centralization of midface soft tissues.
The S flap then comes in two varieties, each of which has a specific benefit for the patient. The simple S flap is elevated in the subperiosteal plane across the entire face of the maxilla between two points of vertical release: the alveolar cleft and lateral wall of the buttress. It remains attached to the teeth. A gingival releasing incision is not required. By virtue of its ability to centralize the soft tissues of the entire midface, the simple S flap fulfills two important functions: (1) the creation of an esthetic soft tissue “drapery”; and (2) the biologically active osteogeneic potential of the mucoperiosteal stem cells is transferred forward, thus enabling bone deposition to take place in centrically for better facial skeletal projection. The drawback of the simple S flap is that it cannot produce a mucoperiosteal flap to cover over the anterior aspect of the alveolar cleft. In such cases, a rectangular mucoperiosteal flap can be harvested immediately lateral to the alveolar cleft and brought over for closure. This design works for narrow clefts.

The compound S flap (as described by Sotereanos) implies a full gingival releasing incision all the way from the alveolar cleft back to the tuberosity. This involves additional operating time. Gingival release from an alveolus containing erupted primary or secondary dentition is technically simple. The bone is solid and the surgical plane easy to follow. Infants are a different matter. Prior to the 4th month of life maxillary and alveolar bone is too soft to work with. DFR should be performed between ages 4–6 months. Dissection is facilitated using loupe magnification, a #15C or Beaver blade, an amalgam packer and Molt (#9) or Louisville periosteal elevators. A compound S flap can be mobilized two full tooth widths. The main advantage of the compound S flap is the dramatic increase in mucoperiosteal flap length it provides. This is generally the width of two dental units. The anterior aspect of the alveolar cleft is thus covered with stem cell-containing mucoperiosteum. Disadvantages of S flap relate to the temporary anatomic disruption of periocoronar attachments.

The S flap has distinct indications for use in primary and secondary cleft repair. These are of vital importance, particularly when one must operate under conditions that are less than ideal. In a primary cleft, whenever possible, pre-surgical orthodontic control of the arch should be accomplished. This may also involve lip adhesion. The entire effort is directed toward normalizing arch dimensions. In these cases, compound S flaps are not required. Closing the anterior wall of the alveolar cleft can be accomplished with a rectangular superiorly-based mucoperiosteal flap elevated directly off the lateral incisor. The space will fill in nicely. If one desires, the flap donor site can be grafted with Alloderm®.

Limited resources and difficult logistics in economically developing countries force surgeons treating cleft patients to adopt different and creative strategies, particularly when the alveolar cleft is wide. These children often have no access to pre-surgical orthopedics. They may never receive orthodontics. They do possess a ready source of bone graft from the rib. In such cases, achievement of a consolidated arch and elimination of oronasal fistula are reasonable goals for primary cleft surgery. The two key factors for success in these patients are: (1) ability to make a simple acrylic splint and secure it—2 mm screws work perfectly well; and (2) meticulous dissection of the alveolar cleft with attention to its vascular anatomy.

If a dental splint can be fabricated, lip adhesion can narrow the cleft down to dimensions permitting simultaneous DFR repair and alveolar grafting with a simple S flap. When this is not possible, a compound S flap will close the gap. In some very wide primary unilateral clefts I have used bilateral compound S flaps to successfully close gaps of 14-16 mm. Of course, this maneuver disconnects the frenulum from the midline. Postoperative remodeling of the alveolus reestablishes symmetry. In such cases, arch stability justifies the extensive intraoral dissection.

Secondary cleft reconstruction follows the same rules but with the proviso that gingival release incisions are fast and easy to accomplish. The floor of the alveolar reconstruction has already been provided by a pre-existent palatal repair. When orthodontic capability exists, such clefts should first be expanded to the original width prior to grafting. But when the patient has no access to ortho, S flap alveolar reconstruction can be combined with DFR to produce a permanent, aesthetically pleasing and physiologic result in one stage.

Step 5. Dissection of the primary palate
Beginning with the lateral nasal wall, the mucoperiosteum is elevated off the maxillary alveolar bone. The palatal margin is undermined as well. This results in a large mucoperiosteal envelope. This must be separated into nasal and oral components. This is done at the level of the “shoulder” of the alveolus. The incision can be extended backward a few mm along the edge of the palatal shelf. On the medial side, the septal mucoperichondrium and the vomerine mucoperiosteum are elevated in continuity.
The surgeon then proceeds forward to elevate the mucoperiosteum off the premaxilla. Separation of the envelope occurs at the “shoulder” of the premaxilla. A vomer mucoperiosteal flap is elevated and sutured to the nasal mucosa of the cleft maxilla.

A six-sided “box” is created. The medial and lateral walls are raw bone. The floor is oral mucoperiosteum. The anterior wall is the sliding sulcus mucoperiosteum. The roof is the nasal mucoperiosteum + the NPP flap. The posterior wall is the vomer flap. These flaps are loaded with undifferentiated mesenchymal stem cells (MSC). The cambium layer of the periosteum is especially rich in stem cells. All neural crest MSCs contain membrane-bound receptors for BMP. This pocket is now ready to be filled with rib graft (iliac crest graft in older children) or with an rhBMP-2/ACS implant.

Alveolar reconstruction, both primary and secondary, demands precise knowledge regarding the vascular anatomy of mucoperiosteum bordering the alveolar cleft. This is a neglected area of cleft surgery. The blood supply to the premaxilla is the most complex (and potentially treacherous). It is derived from three sources. (1) Premaxilla is an r2’ derivative. Like all mesenchymal structures caudal to r1, premaxilla is supplied from the external carotid. The vascular axis of the vomer and premaxilla is the medial sphenopalatine (nasopalatine) artery. PMx mucoperiosteum supplied by the SPA envelops the bone from above-downward, like a cloak thrown over a chair. (2) From below, PMx mucoperiosteum is continuous with that of the r2 maxillary palatal shelf (MxP), the arterial axis of which is the external carotid-based greater palatine artery. (3) A second anastomosis exists within the mucoperiosteum covering the anterior superior aspect of the premaxilla. This zone is in contact with the soft tissues of the prolabium, a p5 derivative supplied by the internal carotid-based anterior ethmoid arteries.

Premaxillary survival depends upon maintenance of sufficient contact between the bone and its enveloping mucoperiosteum. Alveolar cleft reconstruction cannot be accomplished without stripping away the vascular coverage of PMx, to a greater or lesser degree. Thus, premaxillary vascular anatomy dictates the direction in which the mucoperiosteum must be elevated. Surgical strategy for this maneuver is a direct consequence of the cleft type.

In unilateral palate clefts, the medial alveolar cleft margin is a continuous zone of external carotid from the SPA to the GPA. This permits downward dissection of the mucoperiosteum from the shoulder of the premaxilla or upward dissection from the oral margin. The premaxilla remains alive, based on its other sources. In bilateral palate clefts, no such palatal anastomosis exists. The mucoperiosteum can only be reflected upward. If this is done on both sides and if the prolabium is simultaneously elevated, premaxillary necrosis can ensue.

This principle is exactly the same as in probalial death reported by Millard, a combination of bilateral rotation backcuts (destroying the anterior ethmoid supply to the philtrum) and elevation away from the underlying premaxilla. In bilateral cleft lip and palate, the anatomy of both prolabium and premaxilla consists of paired angiosomes. Disruption can occur in one or the other with impunity, but not both.

Fortunately, the robust blood supply of the maxilla comes to our rescue. Injection studies demonstrate the maxilla to be supplied by the pharyngeal branch of the facial artery and facial branch of the ascending pharyngeal artery. It is independent of its mucoperiosteal cover. Meticulous dissection of the lateral alveolar cleft creates a continuous mucoperiosteal sheet uniting the lateral nasal wall with that of the hard palate. This contains a mirror-image external carotid anastomosis, this time between the medial sphenopalatine and greater palatine arteries. This can be stripped downward with impunity. It is a long flap, extending from just in front of the inferior turbinate all the way to the hard palate. The lateral alveolar mucoperiosteal flap is of sufficient size to close the entire floor of the alveolar cleft.

What then should be done for children BCL(P)? Must one sacrifice arch reconstruction to facial aesthetics or vice versa? The answer (no!) lies in a staged approach with careful dissection. Primary lip reconstruction must not violate the tissue plane separating the prolabium and mucoperiosteum. Even in a bilateral dissection, 50%of the premaxilla will remain perfused by both SPA and AEA vessels. Lateral nasal wall reconstruction with the cymba composite graft is appropriate. Liberation of the medial crura must be done without entering the territory of the medial sphenopalatine artery. If this appears difficult, nasal elevation and lip revision may have to be staged later. Alveolar cleft closure should be done by elevating the premaxillary mucoperiosteal flap upward and the maxillary mucoperiosteal flap downward.
Wide bilateral clefts require alternative strategies. Two principles to avoid trouble in these patients are: (1) meticulous primary closure of the nasal floor; and (2) delayed grafting of the alveolar cleft. A wide alveolar “roof” defect requires big nasal floor flaps. These are best harvested at the primary surgery, before other tissues get in the way. The key point here is to not cut the lateral mucoperiosteal flap until the medial flap is harvested from the premaxilla. Once the dimensions of medial flap are known, a superiorly-based lateral flap of sufficient size is elevated to complete the anterior nasal closure. At this point we are still left with a wide alveolar “floor” defect and not enough tissue to close it. What does one do? One waits it out, using a palatal acrylic splint (secured with pins or 2 mm screws) to maintain the transverse arch dimensions and prevent collapse.

6 months later, at palatoplasty, the long mucoperiosteal flaps elevated from the hard palate provided ideal coverage for the alveolar cleft “floor.” Bilateral compound S flaps are raised. These will adequately seal off the anterior face of the alveolar cleft. The graft (using rhBMP-2 or rib) is placed. Once again, care must be taken to prevent transverse collapse until the bone graft has hardened. Intraoral splinting is thus continued for another 2-3 months.

Secondary grafting in bilateral cases follows the same principles. The vascular anatomy may have been altered (primary surgery may have elevated the prolabium away from the premaxilla). In such cases some degree of revascularization to the premaxilla occurs over time. Nonetheless, dissection of the premaxillary mucoperiosteum must be sparing. All that is required is elevation of sufficient tissue to close the soft tissue defect and place the graft. Nasal tip elevation can be safely accomplished using DFR technique and cyma grafts. The key point is to preserve soft tissue continuity between the vomer, the septal mucoperichondrium and the premaxilla: this is the site of entry of blood supply into the premaxilla.

**Closure sequence**

**Step 1. Elevation of the nasal tip**

The lateral crus is stabilized into symmetry with the normal side using a transcutaneous 4-0 chromic mattress stitch. The V-shaped limbs of the releasing incision are closed. The donor site is reconstructed using a full-thickness retroauricular graft or a composite graft of anterior ear skin and cartilage. By trial and error I prefer the cyma as the donor site.

The tip is positioned anatomically using the Cronin nasal retractor (Padgett Instruments). The medial crura are battened together with 5-0 PDS. Suture suspension and modification of the alar cartilage can be readily executed by means of the open-closed approach as per the surgeon’s preference. The author’s approach is to avoid the establishment of normal field relationships. Wide dissection of the nasal soft tissues combined with release from their “piriform prison” allows all fields to be passively held in position by a nasal stent inserted at the conclusion of the procedure. The author finds the Koken-type silicon stents (Porex Corp, Newman, Georgia) easy to use. The stent is placed at the end of the surgery. Closure of the nostril incision starts at the intermediate crus working medially down to the Reinisch flap. One then proceeds laterally to the margin of the lateral nasal wall. This involves placing 2-3 sutures over 5 mm.

The remainder of the lateral nasal incision will be filled by inset of the B flap. Reconstruction of the frontal process of the premaxilla adds the missing tissue to the lateral nasal wall (see below). This frequently involves placement of a composite for full thickness graft into the defect. The medial crural complex is then elevated with respect to the septum with 4-0 vicryl. This also closes the membranous septum counter-incision.

**Step 2. Soft tissue reconstruction of the premaxilla**

The roof of the missing premaxillary field is reconstructed based on meticulous closure of the nasal floor. This is carried out using a mouth gag, starting anteriorly at the incisive foramen. Because the space is tight using 5-0 Vicryl® on a small P-2 needle is helpful. The medial vomerine and lateral nasal mucoperiosteal flaps are closed all the way posterior to the end of the vomer flap. This provides correct orientation for inset of the B flap. The posterior margin of B is sutured from medial to lateral along the newly-united nostril floor. The tip of B is eventually inset into the donor site of the lateral crural advancement flap. Next, the alar nasal skin flap C is sutured to the anterior margin of B. The surface area of the lateral nasal wall is now restored.

The floor of the missing premaxillary field is reconstructed when the medial and lateral mucoperiosteal flaps harvested from the walls of the alveolar cleft are turned down into the mouth using 4-0 vicryl. The cleft side sliding sulcus flap S is advanced and secured using 4-0 Vicryl PS-2 around the dental units. Three or four such sutures will suffice. Optional suspension of the sulcus to...
The purpose of this suturing is to control the height and curvature of the cleft side nostril. This is accomplished by connection of the nasalis to the anterior nasal spine. However this step is optional at this point. If performed, care should be taken to not tighten this and inadvertently narrow the nostril. When the remaining sutures are placed, a decision can be made if a Delaire 2 is warranted. I usually decide upon a Delaire 2 at the end of the case to accentuate curvature.

Orbicularis closure: Three or 4 sutures of 4-0 PDS are required for the DOO layer. The SOO layer is closed with 5-0 PDS making sure the loop is at the level of the dermal-epidermal junction.

Delaire 3: The oblique head of SOO sets the aesthetic drape of the lip. The 5-0 PDS is obliquely passed upward from the cephalic edge through the base of the columella and then back down to the SOO as a mattress suture.

Step 5: Final adjustments: finessing the nostril floor
After closure of the lip, the alar base C is, at time, compressed medially by the movement of the neighboring D flap (lateral lip element). If so, the alar base must be translocated laterally. This can be accomplished by excision of a crescent of skin from the lateral lip element. C is then elevated and secured to the lateral via a buried 5-0 PDS suture. At times it is necessary to elevate the ala completely in order obtain sufficiently lateralization. The tip of the nostril sill flap C’ is now sutured just posterior to the columellar shoulder. Continuity between the columellar shoulder and the nostril sill is now reestablished. Perialar suturing with inverted 5-0 PDS sutures restores the alar crease very nicely.

A final caveat concerns the lining of the lateral nasal defect. Experience with this technique shows that prevention of post operative contraction is paramount. This requires placement of a Porex® silicon conformer sized to the patient. The stent should be sutured in place with 4-0 nylon for 8 weeks. It is important that the B flap not be sutured to the sidewall of C with any tension. If tension exists I have found it prudent to use a small pinch graft of retroauricular skin.

DISCUSSION
Whenever a surgical procedure for a congenital condition in a growing child leads to a predictable pattern of relapse over time two, inescapable conclusions must be drawn: (1)
the biologic rationale of the procedure is incorrect; and (2) the anatomic pattern of relapse points an accusatory finger at the pathology. DFR surgery represents a new form of thinking about clefts by identification and rearrangement of specific developmental fields. The primary pathology of cleft lip is hypoplasia or absence of the distal premaxilla. DFR is designed to reconstruct the missing premaxilla using osteoinductive technologies for stem cell concentration and differentiation such as rhBMP-2 or by conventional osteoconductive technique (bone grafting). DFR incisions are made on the basis of vascular supply and embryology, not geometric manipulation. For this reason the design of DFR presents a series of specific solutions that speak to problems in cleft surgery hitherto inadequately addressed: distortion of the nasal envelope and septum, displacement of soft tissues away from the midline, the entrapped position of the medial crus and the “hidden” nostril sill. DFR treats mechanisms, not appearance.

The extraoral design of DFR is invariable; it is the same for both unilateral and bilateral clefs. Developmental fields are recognized, separated and rearranged into correct anatomic relationships. The intraoral design varies with cleft type. If no alveolar cleft is present, subperiosteal release and zygomaticomaxillary buttress backcut are sufficient to achieve centralization of the soft tissue envelope. Gingival release is not necessary. If a primary palate cleft exists, a complete sliding sulcus dissection with gingival release will allow for mesial translocation and coverage of the alveolar cleft.

DFR includes a straight-line repair, resulting in an anatomically correct philtral column. The distorted and seemingly foreshortened philtrum in clefts is familiar to all plastic surgeons. For many years, the rotation-advancement technique has been used to reorient the philtrum. So why is philtral rotation not required in DFR? The appearance of the philtrum in clefts stems from the overall malposition of the soft tissue in a falsely lateralized state. Thus, the philtrum and columella appear to be short, but are actually displaced into the nasal tip. DFR achieves de-rotation of the philtrum and columella by the following: (1) field separation; and (2) subperiosteal mobilization. The entire soft tissue complex in a unilateral cleft is laterally displaced “around the corner” of the premaxilla. When mobilized correctly off the bone it is brought forward around the curve of the bone and drops right into place.

The membranous bones of the maxilla and premaxilla are just a product of the soft tissue functional matrix that surrounds them. As the soft tissue grows, new bone is deposited and old bone resorbed according to the mechanical forces placed on the bone. The purpose of subperiosteal centralization in DFR is to change the biologic relationship of the bone product to the functional matrix. Over time, the former will adapt to the latter in its new, centralized position. Appreciation of the subperiosteal plane as the correct approach to surgically separate the functional matrix from its product (bone) ensures tension-free release and accuracy of muscle repair (individual muscle units can be identified by tugging on them from below). Thirty years of work by Delaire and others confirm the clinical accuracy and safety of this approach. Readers are also advised to study the pioneering work on primary bone grafting done by Rosenstein,[59-60] This 30 year follow-up of dental development in patients treated with careful presurgical orthodontics and rib grafting demonstrates clearly the safety and advantages of gaining control over the alveolar cleft at primary surgery.

The concepts and techniques of DFR are applicable to all forms of clefts. Because it separates out osseous pathology from soft tissue pathology, DFR is a true “cut as you go” technique. In secondary cleft surgery, DFR is capable of rescuing previously violated fields and reuniting them into their correct functional relationships. Sliding sulcus flaps function as elastic flaps as defined by Goldstein[61-62] and can readily be brought into the midline. Abbe flaps are virtually unnecessary, even in salvage cases involving total loss of the philtrum.

DFR is a practical application of developmental anatomy. It provides a means to analyze membranous bone formation and periosteal physiology. It embodies concepts of facial growth central to orthodontics. It makes use of neuromeric theory to map out and manipulate developmental fields. Proper implementation and study of cleft repair using DFR will provide a forum for dialog among craniofacial surgeons, orthodontists and developmental biologists.

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Cleft Lip and Palate Surgery

These references cover the bases of developmental field theory

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EPILOGUE FOR COLLEAGUES IN INDIA AND SOUTHEAST ASIA

I should like to depart at this point from traditional academic writing to address the reader in a very personal way. Many of you must treat children and adults with clefts under the most demanding of circumstances. Poverty, poor communication, distance from medical centers and difficulties in transportation negatively affect every aspect of your treatment plan. The same is true for your patients, no matter how motivated they might be. Time taken to bring a child in for treatment from a remote location may present an intolerable stress on families living in these circumstances. For many, if not most, of the world’s children with clefts, these factors make careful, well-regulated multidisciplinary care a distant mirage so easy to talk about, so difficult to achieve.

For these patients the surgical team must set priorities to accomplish the maximum physiologic correction with the minimum of surgeries. The goals at the first stage are: (1) a functional nasal airway; (2) a united dental arch without collapse; (3) subperiosteal dissection designed to promote centric facial growth; and (5) soft tissue correction using developmental field concepts to place scars into embryonic separation planes. The goals of the follow-up stage are: (1) meticulous three-layer closure of the soft palate; and (2) use of AEP (alveolar extension palatoplasty) flaps to prevent anterior fistula. Should the patient be unable to return for further work, this should provide at the very least an intact oral cavity in some degree of alignment, a working airway, reasonable speech and an appearance that is socially acceptable.

Developmental Field Reassignment cleft surgery was designed with these circumstances in mind. It is a “high intellect/low technology” method. The only structures missing are a lateral nasal wall soft tissue defect (equal to the cymba) and an alveolar bone defect (equal to the housing of the lateral incisor). Reconstruction of these vital elements is within the hands of all cleft surgeons. There are only two requirements for DFR to be properly performed: (1) in-depth anatomic knowledge of embryologic field anatomy; and (2) the technical ability to make and secure an acrylic palatal splint. How one achieves alveolar reconstruction is irrelevant. One may use cancellous graft (rib in young patients) + platelet enriched plasma. One may use rhBMP-2. What really matters is to follow biologic principles.

DFR was born in post-war Nicaragua, under the harshest of conditions. I was assigned there to work for the Pan American Health Organization (WHO) as a surgical consultant in 1990-1992. At that time, the backlog of plastic surgery cases was estimated at 10,000 patients. Problems were not only logistical but also the creation of manpower. Working with Professor Arturo Gomez at the Universidad Nacional de Nicaragua in Leon and with a small group of lay people, the first medical foundation in the history of Nicaragua was formed. Noel Icaza, an agricultural engineer, gave it the name Nicaplast. We traveled around the country, operating and working within various cities to create support for this apparently crazy idea. Nicaragua had descended into chaos. The gross national product was just above that of Haiti, making this formerly active exporting nation the second-poorest of all the Americas. Given this situation, the creation of resources to resuscitate reconstructive surgery seemed an impossible task.

At this point, Carlos and Vivian Pellas stepped in. These remarkable people, from a highly educated and wealthy family, survived a commercial airline crash in 1989. 170 perished at the scene. The Pellas were evacuated to Miami where they underwent numerous surgeries for burns and fractures including free tissue transfer. Upon recuperating they returned to Nicaragua determined to help children with burns and deformities. Their support for Nicaplast resulted in the establishment of new residency training at Unan-Leon, improved infrastructure and a complete modernization of the pediatric burn unit in the capital city, Managua. Partner relationships were constructed with Interplast, the University of Wisconsin, Physicians for Peace and the AO Foundation in Basel, Switzerland. Each one of these entities contributed toward expanded reconstructive surgical care and advanced education. In time, a new foundation, under the leadership of Vivian Pellas, was established. The Asociacion Pro Ninos Quemados de Nicaragua (APROQUEN) now has direct responsibility for burn care in that country.

The residency was particularly innovative. The longest in Latin America, it demands full general surgery + 3 years of plastic surgery, preparing one graduate every three years. Participation by the resident in all activities with visiting teams created a broad exposure. Mini-fellowships were created. Each graduate undergoes final oral examination in English with the participation of national and international
faculty. All three graduates remain in the public sector. Dr. Gustavo Herdocia work at UNAN-Leon while Dr. Mario Perez and Dr. Leonel Briceno staff the APROQUEN burn unit. Over 4000 surgeries and 100,000 outpatients have benefited from these efforts without a single dollar from abroad.

But human resources are not enough. While Nicaplast continues its valuable work in Leon, APROQUEN has surged forward as a modern-day model of a medical foundation with a bold strategy: the creation of a financial “engine” to finance the clinical work and cost of running a 1st-class burn center in an extremely poor country. In 1972 a terrible earthquake destroyed much of the capital city, including the main hospital for the country. Extensive outside aid was lost to corruption; the hospital was never rebuilt. 4 temporary structures built 35 years ago continue to provide care to the population. A high quality “center of excellence” was nonexistent.

Burns are a socioeconomic disease. Nicaragua has 1000 new burn cases per month, 10% of which require admission. Given the many needs in the health sector, public resources are grossly inadequate for these patients. Knowing this, APROQUEN partnered with other investors to build the most modern (albeit small) medical facility in the country, with state-of-the-art radiology, surgery and intensive care. The facility would serve the international community and private patients. It would be the very best in Central America. Trips to Miami would be avoided. But the key concept is that that APROQUEN would maintain 51% ownership... and a percentage of profits would thus directed to the burn service and reinvested for the foundation.

12 years ago I proposed this plan to Carlos and Vivian Pellias over rum on a muggy Managua night over several rounds of Flor de Cana Rum. In 2005 the Hospital Metropolitan Vivian Pellias opened its doors. HMVP exists because people cared deeply, refused to give up hope and worked unceasingly to make it happen. And the stunning fact of the entire Nicaragua experience is that it happened from within. Information regarding these foundations is available to all surgeons on the Internet.

I write this to you, the reader, to propose that improvements in cleft care can always be made, even under difficult circumstances. Our patients speak for themselves. The primary task is to make it possible for the national medical teams to achieve the logistical support required to provide the care. And, in this regard, innovative efforts in the private sector, backed by lay people with the desire to help, to bypass roadblocks and provide better service.

As I look backward and contemplate the evolution of developmental field theory and the DFR repair, I realize that it followed a number of steps, some forward and some backward. The driving force behind innovation comes from restless curiosity and the capacity to demand that anatomy must make sense. Nature is trying to tell us her secrets if we can only learn to listen. As we understand how the face is constructed, we will learn the sequence that leads to cleft formation. This inevitably will direct us toward better techniques and better results. As a cleft-affected person I know the reality behind the surgeries we perform for our patients. We must push forward to do better. Hopefully, DFR surgery will lead you to question old assumptions, strive for perfection and (most importantly) find your own innovations.