INTRODUCTION

Imagine the birth of a healthy, beautiful child as an occasion of excitement, anticipation, and incredible joy for parents and family alike. Although every parent is faced with fears and anxiety about the future and how this newborn will fare in the coming years, this is a time of great wonder and beauty. Now suppose that, on first look at the child, this picture is marred by the appearance of a deeply pigmented birthmark covering a major portion of the baby’s face, trunk, or extremity. This time of joy can be suddenly transformed into one of horror, concern, despair, and fear. This transformation is not often softened by the lack of readily available information about what the lesion is (if it is malignant or benign, and if there are other problems associated with it) and even less knowledge of what can be done to eliminate its presence and lessen the impact of the lesion on this child’s future. Unfortunately, there is often a delay in gaining this information.

Regardless of the size of the “birthmark,” early consultation with a knowledgeable physician is critical to the family’s ability to bond with their newborn, and that physician, whether he or she is a dermatologist or plastic surgeon, must be armed with the information to present the family with a straightforward, easily understood discussion of the nature of the lesion, the varied approaches to treatment, and an overview of the outcomes of the varied methods of treatment. If well presented in a compassionate manner, even the prescription for multiple surgeries over a many-year period can at least offer these new parents the hope that eventually their child will be able to integrate normally into the world. It is the purpose of this chapter to lay the groundwork for such a discussion and outline an approach to treatment based on experience with more than 200 large and giant nevi, as well as many more small to medium lesions, pigmented and nonpigmented, treated over a 17-year experience on an active pediatric plastic surgical service (with close ties to a pediatric dermatology service). The treatment plan outlined has evolved over this period based on careful consideration of the effectiveness of each different treatment modality in each body region with similar-size lesions. Refinements and modifications in treatment have come about through a desire to improve cosmetic and functional outcomes and to minimize the likelihood of late corrective surgery for problems encountered in the early treatment. We begin with an overview of the lesions treated and the indications for treatment, and then we concentrate on the pros and cons of different treatment modalities. Although we expect that these treatment “protocols” will continue to evolve over the coming years, we believe that this chapter represents a large step in the direction of dealing with these often problematic lesions.

INDICATIONS

Congenital nevus is any nevus present at birth. The word nevus refers to an abnormal or faulty growth in the skin that is generally synonymous with the term hamartoma. It is a nonspecific term that applies to a wide variety of cutaneous lesions. The term is derived from the Sanskrit root -gha and the Latin root -gen, both referring to birth. It is descriptive of a histologic clustering of cells into nests that share embryologic origin but in an ectopic location. Although most physicians and patients use the word nevus to describe pigmented lesions, this term applies to a variety of cutaneous malformations, (e.g., epidermal nevus, sebaceous nevus), many of them not present at birth.  

Most congenital nevi are made up of congenital melanocytic nevi (CMN). At birth, approximately 2.5% of children have pigmented lesions, and 1% of newborns have congenital melanocytic nevi. CMN result from an aberration in the normal development of the neuroblast cells. Melanoblasts, which are precursors of melanocytes, migrate during the early embryologic stage from the neural crest not only to the skin but also to mucous membranes, eyes, meninges, ears, mesentery, and the chromaffin system, where they differentiate between the eight and tenth weeks of gestation into dendritic melanocytes. When the normal differentiation and migration of those cells is disturbed, the result is an ectopic population of cells seen in nevus. Nevus cells in the dermis are classified into
types A (epithelioid) found in the superficial dermis, type B (lymphoid) found in the mid-dermis, and type C (fibrocytic) found in the deep dermis. They are distinguished from fibroblasts and Schwann cells by staining with immunoperoxidases, including the S-100 stain and myelin basic protein stain.23

CMN are characterized at birth by a thickened skin that may be smooth, rough, nodular, or verrucous with variations in shades of brown and blue throughout the lesion. Many of them are covered with hair that is coarse, larger, and more pigmented than the patient's normal hair (Figure 65-1). There are often multiple satellite pigmented papules beyond the periphery of the main lesion; some may be faint or barely visible at birth, increasing in depth of pigmentation over the first 6 to 12 months. As the infant grows, the involved areas become thicker and frequently darker, the surface is more irregular, and verrucous nodules frequently develop.23,40 Giant nevi located in the head and neck and posterior trunk may be associated with leptomeningeal melanocytosis and neurologic disorders, such as epilepsy.22 Those that overlie the vertebral column may have underlying spinal defects, such as meningocele or spina bifida. In some cases of giant nevus, computed tomography (CT) and magnetic resonance imaging (MRI) studies may reveal neural lesions, particularly in the temporal lobe, which may or may not be associated with a seizure disorder.14

CMN can be divided into three groups, depending on size (see Figure 65-1).20 Small congenital nevi are defined as those less than 1.5 cm in diameter, most of which are easily excised in a single operative procedure and are found in 1% of newborns.40 Medium congenital nevi are defined as those greater than 1.5 cm but less than 20 cm in diameter and are seen in 0.6% of newborns. Large and giant congenital nevi have been defined variously as those larger than 20 cm, those greater in total area than 100 cm², or those that cannot be completely

Figure 65-1. Congenital nevi, varying in size, depth of pigmentation, and hair growth. A, Multiple small nevi of the face in a child with giant nevus of the trunk, showing marked variation in thickness, color, and texture. B, Medium (7-cm) nevus of the flank with typical variegated pigment within the lesion. C, Giant nevus of entire back with dense pigmentation and hair growth.
excised without a significant deformity. Other authors have defined large and giant nevi as those measuring greater than 2% of total body surface (TBS) (the definition we have followed). This rare variant occurs in less than 1 in 20,000 newborns.

The differentiation of congenital from acquired melanocytic nevi carries an important treatment connotation. There is controversy about whether specific histologic changes can differentiate congenital from acquired melanocytic nevi. According to Mark et al., con genital melanocytic nevi differ histologically from acquired nevi. In contrast to acquired melanocytic nevi, a congenital nevus is generally a deeper lesion with melanocytes distributed far down into the dermis and sometimes even in the subcutaneous fat. The nevus cells tend to be grouped around adnexal structures, nerves, blood vessels, and lymphatics. These features, however, are not specific to congenital nevi and can occasionally be seen in acquired nevi.

One of the most important elements of the diagnosis of CMN concerns the likelihood of the development of melanoma (and for giant nevi, the possibility of degeneration into either melanoma or sarcoma). Although the association between large and giant congenital melanocytic nevi and malignant melanoma has been established beyond reasonable doubt, the magnitude of risk of malignant transformation has varied in the literature from as little as 2% to as high as 31%. Many of these studies were significantly skewed by the sample population examined. Quaba and Wallace, in their 1986 review of the subject, attempted to put the widely different figures into perspective, and they have calculated an 8.51% incidence of melanoma developing with larger than 2% of the TBS during the first 15 years of life. The opinion that complete prophylactic excision of all giant nevi should be accomplished in infancy and early childhood is well supported in the literature. Approximately 50% of malignancies that have occurred in giant nevi developed in the first 3 years of life, 60% by childhood, and 70% before puberty. The lifetime risk of melanoma in patients with giant CMN may be as high as 15%. The nature and magnitude of the association between small CMN and melanoma is still controversial. Obtaining accurate data on malignancy of small CMN is very difficult because establishing their congenital nature based on history taken by an older child or adult is often inaccurate. Careful statistical analysis estimates a 21-fold increased risk of melanoma for individuals with small CMN when nevi were ascertained by history, and a threefold to tenfold increase in risk when nevi were uncertain by histopathologic criteria (the presence of cytologically benign nevocellular nevi in the dermis immediately adjacent to invasive melanoma). Although history and histology are not infallible determinants and the strength of association is entirely dependent on methods used for ascertainment of CMN, the observed frequency of association is several orders of magnitude greater than expected based on surface area considerations and chance alone.

The lifetime risk of melanoma in patients with small CMN as stated by Rhodes and Melski lies between 0.8% and 2.6% by histologic method of determination and 4.9% by historic assessment. The risk of melanoma before puberty is 1:200,000 or near 0, with the risk increasing in the years following.

Aside from the potential for malignancy, the cosmetic deformity and psychologic impact of CMN on a child are profound. In certain instances the parents will reject and even abandon a newborn infant with this condition. Modern plastic surgical techniques can often greatly ameliorate the cosmetic defects, and parents should be given a realistic appraisal of this condition. The benefits both from a risk of malignant degeneration standpoint and aesthetic concern of excising the large and giant lesions before school age cannot be overemphasized because the procedures involved are often significantly better tolerated during infancy and early childhood.

There are several variants of congenital melanocytic nevi that are important because they can simulate melanoma histologically. Blue nevi, Mongolian spots, and the nevi of Ota and Ito are benign melanocytic nevi frequently present at birth that are characterized by proliferation of melanocytes that are located wholly in the dermis. They occur secondary to an arrest in embryonal migration of melanocytes bound for the dermal-epidermal junction. They most likely represent different stages of the same physiologic process.

MONGOLIAN SPOTS

Mongolian spots are flat, deep brown or blue-gray, often poorly circumscribed, large macular lesions with a predilection for certain locations and racial groups. They are often found over the lumbosacral region and buttocks. Occasionally they are seen in the upper extremities and shoulder. Infrequently they appear on the abdomen, chest, and rarely on the palms or soles. Approximately 96% of black infants, 46% of Hispanic infants, and about 10% of white infants are born with these lesions. They may be single or multiple and vary in size from a few millimeters to 10 cm or more in diameter. Although they may occasionally persist into adulthood, Mongolian spots are usually self-limited and therapy is unnecessary.

BLUE NEVUS

Blue nevi, although often seen in childhood, occasionally are present at birth. These lesions are histologic variants of the intradermal nevus found in the mid-dermis to deep dermis. They are divided into two types: simple and cellular. The common blue nevus is a small, round, or oval, blue-black to slate grey, smooth-surfaced, sharply circumscribed, mildly elevated nodule that is usually less than 1 cm in diameter. Although they may occur in any part of the body, they are most commonly seen in the extremities, especially the backs of the hands and feet, the lumbosacral region, and the head. They can be single or multiple, and women are twice as frequently affected as men. Cellular blue nevus are considerably less
common than ordinary blue nevi and are characterized by irregular shape, lighter color, and frequently size larger than 1 cm in diameter with predilection for the lumbosacral region.42 The individual cells in a blue nevus have an elongated dendritic appearance and may be associated with densely aggregated macrophages that have phagocytized the pigment. It is the presence of this dense pigmentation at the deeper layers of the dermis that impart the bluish coloration to these lesions.

Although the common blue nevi remain benign lesions, there is a low but distinct chance of malignant transformation in the cellular variety of this lesion.16,35 Although relatively few cases of malignant degeneration have been reported, these lesions may exhibit aggressive behavior and metastasize.16,35 For this reason, it is generally recommended that blue nevi be excised.

NEVUS OF OTA AND NEVUS OF ITO

The nevus of Ota and the nevus of Ito are clinical forms of blue nevus with a distribution correlated with specific dermatome patterns.23,28 Nevus of Ota was first described in Japan in 1939.28 Since then, multiple cases have been recorded from around the world, most commonly in Indians and blacks. Females account for 75% of all reported cases. Approximately 50% of lesions are congenital and 40% appear at puberty.31 Nevus of Ota is a flat, irregular, gray-blue patch of the face supplied by the first and second divisions of the trigeminal nerve, particularly the periorbital region, temple, forehead, malar region, and nose.28 More than 50% of patients will have involvement of the sclera of the ipsilateral eye and occasionally the conjunctiva, cornea, and retina. Mucous membrane involvement, especially the lips, pharynx, hard palate, and nasal mucosa, is occasionally seen. In about 5% of cases the nevus of Ota is found bilaterally. The nevus of Ito has the same features as the nevus of Ota, except that it is seen in the distribution of the posterior supraclavicular and lateral cutaneous branches to the shoulder, neck, and supraclavicular areas. Unlike Mongolian spots, nevi of Ota and Ito do not disappear spontaneously and hyperpigmentation frequently occurs after puberty. Although these lesions are benign, there have been at least 37 cases of associated melanoma reported in the English-language literature.40 Because microscopically melanocytes are found in the upper dermis, Q-switched ruby and argon lasers have been used for ablation of these lesions with excellent results.15

NEURAL NEVUS

The neural nevus is distinguishable histologically as a type C form of intradermal nevus.23 The cells of the neural nevus have a similar histologic appearance to Schwann cells and nerve organelles (pacinian and Meissner's corpuscles). They may in some cases resemble neurofibromatosis. These lesions frequently appear as smooth, lobulated, hairless nodules varying from light tan to dark black appearance. Some have a soft, flabby consistency, often hanging in folds (Figure 65-2). They are commonly found within congenital giant pigmented nevi, particularly in the midline region of the back. Some authors have suggested that there is a high association of neural nevus with malignancies in these areas,23,52 whereas some patholo-

Figure 65-2.  A, An infant with a giant nevus of the trunk showing the thickened folds of a neural nevus in the lumbosacral area. B, A rare bulky nevocytoma, a variant of neural nevus in the midst of a giant bathing trunk nevus.
gists believe that these lesions have been not infrequently confused with melanomas on histologic examination. Given the fact that there may be this increased association, it is recommended that these portions of a giant nevus be either excised early or at least biopsied if definitive treatment will be delayed.

**NEVUS SPILUS**

Nevus spilus is a sharply demarcated, flat brown patch, speckled with smaller dark brown to black-brown areas of pigmentation (Figure 65-3). This is a relatively common lesion found in 1% to 3% of the adult population with equal gender prevalence; it is frequently seen on the trunk and extremities. Although frequently present at birth, it may occur in infancy or childhood. Its size may vary from 1 to 20 cm in diameter, almost always solitary. Histologically, the epidermis shows increased pigment in the basal layer with an increased number of melanocytes throughout the lesion and elongation of rete ridges. The darker spots are melanocytic nevi of the junctional and compound types. Although there are reports of malignant transformation developing in nevus spilus, routine clinical observation is indicated unless suspicious changes are seen or if the lesion is clearly congenital in nature. Some of these lesions, particularly when large and in prominent locations, may warrant excision for aesthetic reasons alone.

**EPIDERMAL NEVUS**

Epidermal nevus is a benign hamartoma characterized by hyperkeratosis, acanthosis, and hypertrophy of the epidermis. Onset at birth occurs in 60% of cases; 80% are evident by 1 year of age and 95% by 7 years of age. It affects both genders equally, and lesions can be unilateral or symmetric, solitary, or extensive. Typical lesions are hyperpigmented, linear, velvety, or papillomatous. They are often found in the extremities, although they may occur anywhere in the body, frequently in a dermatomal distribution. Four variants have been described: Nevus verrucous is a localized, solitary lesion often present at birth, linear or oval in shape. Most are 2 to 3 cm in size, often noted on the trunk or extremities but may occur on the head or neck. Nevus unius lateralis is extensive, systematized forms of epidermal nevus that can often cover more than one half of the body. Ichthyosis hystrix refers to widespread epidermal lesions in irregular geometric patterns that follow Blaschko’s lines. Epidermal nevi in which inflammation is present are known as inflammatory linea verrucous epidermal nevus (ILVEN). They are erythematous, pruritic, scaling linear plaques that can be misdiagnosed as psoriasis. The association of epidermal nevi with other congenital anomalies is termed epidermal nevus syndrome (Figure 65-4). Typical anomalies include skeletal abnormalities (in approximately two thirds of patients, with kyphoscoliosis the most common), central nervous system abnormalities (in approximately one third of patients as hydrocephaly, mental retardation, or seizures), and ocular abnormalities (in approximately one fifth of patients as epibulbar dermoids and colobomata of the retina, iris, or lid). Less commonly, anomalies of the urogenital tract and cardiovascular system are seen. The risk of malignant degeneration with epidermal nevi is unknown but not believed to be common. Most tumors reported are low-grade, such as Bowen’s disease, keratoacanthoma, or basal cell carcinoma. In addition, affected individuals may show increase incidence of noncutaneous systemic malignancies. Although surgery may be effective for removal of localized lesions, it may provide little relief in the treatment of extensive (see Figure 65-4) lesions because surgical intervention is rarely indicated in infancy and the extent of involvement with thick, hyperkeratotic lesions later may even render tissue expansion of little benefit. It is also the lead author’s personal impression that excision of these lesions may for unknown reasons be associated with significantly greater risk of hypertrophic scarring, and at the very least this factor should be taken into consideration when deciding on the optimal treatment modality. Although cryosurgery, dermabrasion, or electrodesiccation and curettage may produce gratifying results initially, recurrences are common. At the same time, these later treatment approaches, if properly applied, may give benefit without the risk of unsightly and potentially deforming scarring. In some cases, reepithelialization after dermabrasion or dermaplaning may be followed by a flat appearance without discoloration that may remain as such for many years.

**SEBACEOUS NEVUS**

Nevus sebaceous was described in 1895 by the German dermatologist Jadassohn. It is a congenital hamartoma of sebaceous glands. They present as yellowish-tan, waxy or velvety plaques, most commonly located on the head and neck.

**Figure 65-3.** The typical features of a nevus spilus are visible in this large lesion of the lower lip and chin. The lightly speckled pigmentation is interspersed with melanocytic nevus along the vermilion and in darker spots within the lighter lesion.
Figure 65-4. A 10-year-old child with epidermal nevus syndrome. A, Anterior view with typical linear epidermal nevi of the shoulder and upper arm. B, Giant epidermal nevus of the back with tissue expanders in the back before reduction of the nevus. C, After partial excision of the back nevus and advancement of the adjacent flaps.

Figure 65-5. The varied presentation of a sebaceous nevus are evident in the two infants illustrated here. A, A giant sebaceous nevus of the scalp is seen in a child with sebaceous nevus syndrome who had a severe seizure disorder. B, A typical waxy, pinkish, orange nevus of the cheek and superior auricular sulcus.

(Figure 65-5). The lesion is usually solitary, round, oval, or linear, and varies in size from a few millimeters to several centimeters in diameter. Typically, lesions over the face and neck are linear where scalp lesions are oval, appearing as an area of alopecia. Although congenital in origin, some may not be apparent early and appear to arise in early childhood and rarely in adulthood. They are estimated to occur in 0.3% of births. Although familial cases have been described, they are usually sporadic without gender preference. As the child grows, hormonal changes during puberty bring on a series of changes of these lesions. By adolescence these lesions have often shown progressive thickening with the surface changing from waxy smooth to verrucous with hypoplasia of sebaceous gland associated with itching, drainage, and occasional intermittent inflammation. Large, linear sebaceous nevi in the head and neck
have been associated with a seizure disorder in 10% of cases.\textsuperscript{53} Even more extensive lesions, again particularly in the head and neck, may be associated with other manifestations and neurologic symptoms. Nevus sebaceous syndrome refers to this constellation of symptoms, seen in these patients, including sebaceous nevus, ocular dermoid, major ophthalmic abnormalities, mental retardation, skeletal abnormalities, and seizure disorders.\textsuperscript{10,13,31}

The primary indication for excision of sebaceous nevus is the well-recognized risk of malignant degeneration, which has been reported to be between 15% and 20%.\textsuperscript{13} The most common neoplasm arising from this disorder is basal cell carcinoma. Other tumors arising from lesions of nevus sebaceous include syringocystadenoma papilliferum; keratoacanthoma; leiomyoma; piloleiomyoma hyaladenoa; apocrine cystadenoma; squamous cell carcinoma; and, rarely, aggressive apocrine carcinoma and malignant eccrine poroma. Malignant degeneration, although on occasion reported during adolescence, rarely occurs before early adult life (the third decade).\textsuperscript{13}

Although the risk of malignant transformation is low in infancy and early childhood, larger lesions may be more readily excised during this period, avoiding the more complicated reconstructive procedures at a time that the child may not tolerate them as well. This approach may also avoid the embarrassment of the change in appearance and cellular activity (with increased thickness, discharge, etc.) that occurs in these lesions as a child goes through the hormonal changes of puberty. Treatment of extensive lesions often requires tissue expansion and is approached in a similar fashion to that described below for congenital pigmented nevi. However, unlike the latter, it is recommended that tissue expander placement be carried out through an incision beyond the border of the sebaceous nevus rather than within the lesion. At the same time, excision of the smaller lesions can easily be left until the child is old enough to undergoes the procedure under local anesthesia in an office setting.

**SPITZ NEVUS**

Although not a congenital nevus, this lesion is worth mentioning briefly because it is commonly seen in young children and may be confused with other congenital lesions. The Spitz nevus is a distinct histologic and clinical entity.\textsuperscript{18,23} Originally described as “juvenile melanoma” and readily confused with the latter lesion under the microscope (without the knowledge of the context in which the lesion is seen), these lesions have been reclassified as a variant of an acquired nevus. Typically appearing as a firm, pinkish, raised lesion that may even be confused with a vascular lesion, they may also be pigmented with wide variegation in color (Figure 65-6). They are quite common on the face but may even appear as large clusters of lesions in an area on the trunk or extremity. Histologically, these lesions exhibit clusters of nonpigmented spindle and epithelioid nevomelanocytes in the lower epidermis with elongated nevomelanocytes “raining down” into the upper dermis.\textsuperscript{18,23}

![Figure 65-6. A large cluster of Spitz nevi with typical firm, pink nodules, presented on this teenager's upper thigh.](image)

Although there is no evidence that Spitz nevus is a precursor of melanoma, these lesions may recur and enlarge rapidly if not completely excised, with the histologic appearance seeming to become more disorganized and increasing the concerns about the true nature of the lesion. This being the case, excision with a few-millimeter margin is recommended and has been effective in treating even recurrent lesions in our experience.

**OPERATIONS**

Although much of the following discussion of operations pertains to experience gained in the treatment of congenital pigmented nevi, the treatment of large sebaceous nevi, problematic epidermal nevi, etc., can follow the approaches described. Some variations are mentioned in the Indications section; others will be touched on here.

Over the past two decades, the approach to the management of congenital nevi has evolved. Cronin,\textsuperscript{12} Johnson,\textsuperscript{21} and others\textsuperscript{34} originally advocated dermabrasion and split-thickness excision of large nevi to diminish significant cosmetic deformity (Figure 65-7). Although these techniques may improve the appearance and decrease the overall “nevus cells load,” they do not address the nevus cells located in the deep dermis, around hair follicles, nerves, and lymphatics.\textsuperscript{40}

Curettage and a Q-switched ruby laser therapy have most recently been suggested for removal of CMN. Although these therapeutic approaches may give fairly favorable cosmetic results, they do not necessarily remove all nevus cells and
Figure 65-7. This figure demonstrates the effects of neonatal dermabrasion for treatment of a giant nevus of the face and the potential problem of late "bleed through" of the remaining deep nevus requiring subsequent excision and flap reconstruction. **A**, An infant with a giant nevus of the face. **B**, Dermabrasion of the face at 1 month of age. **C**, At age 3 years showing a reasonably good cosmetic result of the procedure. **D**, At 6 years of age the nevus is now visible with deep pigment throughout the area previously dermabraded.

thereby are not recommended. Either of these treatment modalities may be associated with late “bleed through” of the pigment from the residual deeper portions of the nevus occurring often when the child is approaching school. This presents a greater treatment dilemma than it would have had the definitive treatment been carried out earlier (see Figure 65-7). At the same time, there may be a role for these less invasive treatment modalities for very extensive lesions or those not lending themselves well to treatment. In these cases the ability to lighten the pigment may represent a significant advantage even if the effect of this treatment on risk of malignant change were not seemingly significantly altered.

The most appropriate treatment of CMN is based primarily on considerations of lesion size, location, histology, and patient age. Because the risk of malignant change in small CMN is extremely low during the first decade of life, our approach is to delay excision until the child is older and able to undergo the procedure under local anesthesia. Exceptions to
this rule would be if the lesion were located in an area where excision under local would be poorly tolerated (e.g., periorbital area) or if the lesion lies in a region that the complexity of the reconstruction would still require a general anesthetic even in an older child (e.g., lips, nasal tip or alar margin, eyelids). Most of these lesions can be excised primarily, but not infrequently those on the face may be better excised in two stages to minimize the length of the final scar and avoid the need for more complicated flap procedures (Figure 65-8). Dissection should be carried down to the underlying fascia to remove all nevus cells in most cases, except in the facial area where for smaller lesions, excision well into the underlying fat may be acceptable. Even medium-size lesions, because of the tissue requirements and the need to avoid distortion of key anatomic landmarks of the particular region, may require more complex methods of excision and reconstruction. These include serial excisions or reconstruction with skin grafts, local flaps, or flaps generated using tissue expansion (Figure 65-9). This may frequently be the case with lesions of the scalp, face, and distal extremities.

Giant CMN present a reconstructive challenge for both the novice and even many experienced plastic surgeons. Although complete early excision of giant CMN is often looked on as an insurmountable task, newer approaches to excision and reconstruction gained with further experience with tissue expansion, including varied methods of flap design to maximize the benefits of the tissue gained through expansion, along with the addition of microsurgical refinements, allow the excision to begin as early as 6 months of age and to be completed in early childhood. In many cases this evolution of technique has enabled excision of larger and larger lesions without the need for skin grafting. As we have gained more experience with the
Figure 65-9. Nevi of the periorbital area frequently require a combination of graft and flap techniques for the excision and reconstruction. This nevus of lids, canthus, and medial brow was excised in a single stage using a full-thickness skin graft in combination with an island flap reconstruction of the brow. The brow flap was then thinned. A, The nevus before excision. B, The result 1 year after excision. C and D, The result 3 years postoperatively.

technical aspects of these varied reconstructions and have been able to evaluate the outcomes of the varied treatment modalities, in each given area, we have continued to add refinements that should minimize the likelihood of poor aesthetic and functional outcomes, as well as to minimize the likelihood of additional procedures in later life to address those problems. We will look at treatment of large and giant nevi by body region, looking at the techniques used and which of the spectrum of techniques have proven most effective in each different area. Within the discussion of the operative planning for excision of congenital nevi of the head and neck we include some generalized discussion of expander type, placement, use of expanded full-thickness grafts, and postoperative management of the expander patient.

LARGE AND GIANT MELANOCYTIC NEVI OF THE SCALP

Tissue expansion is the primary treatment modality used for excision and reconstruction of large congenital nevi of the scalp, as well as a part of the treatment plan for most other lesions of
the head and neck. Unless dealing with nevi of greater than 50% of the scalp, when the initial options for reconstruction are limited, every effort should be made to orient hair (particularly along the anterior hairline) correctly. Although tissue expansion can begin as early as 3 months of age with no evidence of residual cranial malformation (although temporary molding may occur that will reshape in the months after expander removal), in patients in whom serial expansion will be required or in whom a large expander (500 ml or greater) will be positioned, expansion at 6 months of age is preferred. The most critical element in accomplishing early excision and avoiding complications is meticulous preoperative planning. Experience has shown that the spherical shape of the scalp and the ungiving nature of the galea necessitates the routine use of large or multiple tissue expanders. Even smaller lesions of the scalp may be treated with initial tissue expansion of the adjacent scalp to minimize problems in healing that result from undue tension. Tissue expanders are placed through incisions just within the margin of the nevus whenever possible (see Sebaceous Nevus above). The incisions are made after infiltration with 0.5% lidocaine with 1:200,000 epinephrine and are carried down into the subgaleal plane in the scalp and the subfrontal plane in the forehead. In general, dissection can be accomplished easily using malleable retractors under the flap. When difficulty is encountered in reaching over the curve of the scalp, dissection can be continued using a urethral sound. The dissection is carried to a dimension of at least 1 cm greater than the expander base in all directions. A narrow path is dissected for the injection port, which is placed far away from the tissue expander (a miniport or low-profile injection port is both well tolerated and readily injected in the preauricular region; the area of the mastoid prominence should be avoided) to allow for safe, full expansion. If necessary, the miniport is secured to the tissues with absorbable sutures (if working in a pocket large enough to see the port) or secured with a carefully placed suture (through the skin and around the tubing) and secured with a gauze bolster on the skin to avoid migration and accidental deflation of the expander. As in all cases in which tissue expanders are being used, after placement of the expander, the injection port is tested. Every effort should be made to avoid surface folds on the expander because this will put the overlying skin flap at risk for possible necrosis. Kinks or bends of the injection port should be prevented to avoid difficulties with inflation of the expander, and care should be taken when using large expanders to ensure that the injection port is far enough from the expander to prevent difficulty in expansion when the expander is approaching maximal size. This may entail adding additional tubing between expander and port in some cases. In addition, some standard (higher-profile) injection ports may prove too high a profile beneath the scalp of an infant or young child, thereby representing a risk to the overlying skin blood supply. These should be avoided. One or more 19-gauge butterfly intravenous (IV) tube drains are routinely placed in all patients (generally one drain per expander), and the needle end of the drain is placed into Vacutainer tubes at the completion of the case. Closure of the incisions is carried in two layers, with 4-0 clear nylon on the galea and 4-0 blue nylon on the scalp. Typically the sutures are left in place throughout the expansion process, only being removed if there is undue irritation.

Expansion is usually accomplished over a period of 10 to 12 weeks, expanding once per week in most cases but increasing to every fourth or fifth day when early expansion has been proceeding uneventfully. Expanders have varied in size from 100 to 550 ml, with most larger lesions being addressed with expanders in the 300-to 550-ml range. We have used rectangular expanders with no reinforced backing in virtually all cases. The amount injected will depend on the area expanded and size of the expander. In the scalp, expansion with 20 to 40 ml is routine once the initial weeks of expansion have demonstrated good healing and the parents are comfortable with the expansion routine (approximately two thirds of the expanders being injected by a family member in our more recent series).

The choice of flap, whether direct advancement, transposition, or rotation, will depend on the amount of tissue obtained, the location of the nevus, and recognition of differences in hair direction (Figure 65-10). Whereas early in our experience we, along with others, used a preponderance of advancement flaps, we have found an increasing benefit both in expedience of covering large areas with less need for serial expansion and improved reconstruction of hairline and hair direction using transposition flaps (see Outcomes section).

LARGE AND GIANT CONGENITAL MELANOCYTIC NEVI OF THE FACE (CHEEK, FOREHEAD, NOSE, EAR)

Although tissue expansion is routinely used for excision and reconstruction of large and giant nevi of the face, areas including the periorbital region (including eyelids), nose, and ears may be best treated with either nonexpanded or expanded full-thickness skin grafts (Figures 65-11 and 65-12). Where little or no normal adjacent tissue is available for expansion, these expanded full-thickness grafts can be used to cover complete facial aesthetic units. More recently, the use of the prefabricated flaps for the central facial and nasal area have improved final aesthetic and functional outcome over full-thickness grafts where immediately adjacent normal regional tissue is unavailable. These prefabricated flaps have the obvious advantage of the added thickness of subcutaneous tissue and improved skin characteristics over full-thickness grafts. Meticulous preoperative planning is critical to avoid the risk of distortion of the eyebrow and temporal hairline in treatment of large and giant nevi of the forehead and also to avoid distortion of the lateral canthus (see Figure 65-11), ectropion, distortion of the oral commissure, and hollowing of the cheek secondary to atrophy of the subcutaneous fat of the cheek for lesions of the midface. Expanders are placed on either side of nevi of the central forehead with 100- to 200-ml expanders used in most cases and flap advancement carried out along the brow. When advancing and rotating a flap from the hemiforehead to the opposite side, great care must be taken to avoid elevation of the brow on the side opposite the nevus and repeat expansion may be needed. Transposition flaps may provide an
additional means of avoiding brow elevation by interposing extra tissue between the hairline and brow. Expansion for excision of nevi of the cheek may be more readily accomplished, particularly when the lesion lies from the level of the lateral canthus medially, by a prior elevation and advancement of the flap with partial excision of the nevus without expansion first. This approach will allow better positioning of the expander to deal with the medial cheek, lip, and commissure area and thereby minimize the likelihood of distorting these areas.

The expansion of full-thickness donor sites for facial reconstruction after nevus excision has been accomplished using postauricular skin, clavicular skin (Figure 65-12), and submental skin (where bearded skin was desired for an upper lip graft in a single case). The donor site expansion is continued until there is enough tissue to harvest the full-thickness graft and close the donor site without tension. Preoperatively, a pattern of the aesthetic unit to be reconstructed is made using x-ray film and used to harvest the appropriate graft. Recognizing the potential difficulties of

Figure 65-10. Excision and reconstruction of large and giant nevi of the scalp is facilitated by use of transposition flaps rather than straight advancement flaps in many cases because of the ease of covering large areas and better orienting the flaps for optimal hair pattern. A, A large nevus of the occipitoparietal area. B and C, With single expander placed posteromedially, showing the outline of the flap and transposition of the flap. D, The result at 6 months after excision.
Figure 65-11. **A**, This large nevus of the medial cheek and infraorbital area was treated by both upward advancement of an expanded flap from below the lesion and advancement downward of some of the skin above the orbital rim. **B**, The result at 5 years after excision shows the persistent effects of an approach that allowed downward pull rather than lateral and upward pull of the flaps on the canthus. This distortion and relative tissue shortage can be hard to correct later.

Figure 65-12. This giant nevus of the face and adjacent temporal scalp required repeated expansion and a combination of flap and expanded graft reconstruction. **A** and **B**, Anterior and lateral views of the lesion. **C**, Initial expansion of the forehead and neck/postauricular skin with subsequent flap advancement into the forehead, island flap from the dome of the expanded scalp for the eyebrow reconstruction, and transposition of the neck/postauricular flap into the cheek.
getting a large full-thickness graft to take over the facial contours, meticulous hemostasis, central tacking sutures, and a suture dressing should be used. Xeroform gauze, cotton soaked in saline and mineral water, and adhesive Reston foam secured beyond the graft margins with skin adhesive and sutures is routinely used for extensive grafts. An alternative to an expanded full-thickness skin graft for medium-size lesions such as both lids or the entire anterior ear would be the harvest of the entire postauricular skin with closure of this donor site using a full-thickness skin graft from the groin.

The pigmentation difference in the latter graft being in the postauricular donor region is not a concern, and the benefits of a large, postauricular graft to the visible face are fully gained.

When applying full-thickness grafts on the face or extensive grafts (both full-and split-thickness on the trunk or extremity), movement between the graft and bed is further minimized by placing the child on a combination of analgesics and sedation, with diphenhydramine (Benadryl) and chloral hydrate (each of these drugs on a q8h regimen but alternated q4h). The graft is

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**Figure 65-12, cont'd.** D, The second expansion was carried out beneath the cheek flap and scalp and at the clavicular expanded skin graft donor site. E, After completion of the nevus excision (except ciliary margin) and flap and graft reconstruction. F, Result at 3 years postoperatively before revision of the eyelid. G, Result at 4 years after the excision.
examined in most cases at 3 to 5 days for full-thickness grafts and 5 to 7 days postoperatively for large split-thickness grafts on the posterior trunk.

**LARGE AND GIANT CONGENITAL PIGMENTED NEVI OF THE TRUNK**

Some of the largest congenital melanocytic nevi are seen over the trunk. They may involve either the anterior or posterior trunk, with the majority being seen in the posterior trunk, with capelike and bathing trunk distributions common. The treatment varies with each location.

Anterior trunk nevi, located in a transverse direction across the abdomen, may be excised and the defect may be closed in an abdominoplasty fashion either in a single procedure or in serial procedures. Dissection is carried down to the abdominal fascia, similar to an abdominoplasty, preferably with electrocautery to minimize blood loss and transfusions. If the nevus cannot be excised and the defect closed primarily in the first procedure, the risk of excessive tension and scar spread can be minimized by using tissue expansion. Large expanders (500 to 1000 ml) are routinely used and placed above the abdominal fascia either above, below, or along either margin of the lesion. Selection of tissue expander site is very critical because dissection in the immediate area of the breast buds should be avoided to prevent distortion of the position of the nipple-areolar complex and subsequent breast growth. Expander ports are typically placed down on the anterior thigh, where they can be readily palpated, or for expanders in the upper abdomen overlying the lower ribcage or sternum, where there is firm skeletal tissue beneath the port.

With careful planning and adequate expansion (frequently overexpanding the expanders), the need for extensive release of the capsule will be minimized. Under no circumstances should the capsule be excised because this will lead to a significant increase in blood loss and may well compromise the vascular supply to the flap. Even limited capsule release should be done very carefully to avoid injuring the blood supply to the skin flaps. In the majority of cases involving the anterior trunk, excisions are accomplished with direct advancement flap closure whether the flaps are expanded or not. Lesions involving the immediate breast or nipple-areolar area present a treatment dilemma. Excision in infancy or early childhood carries a significant risk of injury or even excision of the breast bud with subsequent breast deformity or absence. Attempting to expand the tissue in the same area and move it elsewhere may have similar consequences. It is our recommendation that these lesions be left alone until after thelarche (Figure 65-13). At this time the excision and reconstruction may be carried out more safely. Large nevi may be more easily excised and reconstructed using an expanded full-thickness graft for coverage rather than adjacent expanded skin. Lesions involving the entire breast are relatively rare, and few have been

**Figure 65-13.** This case demonstrates the use of an expanded full-thickness skin graft in treatment of a large nevus of the breast. This approach was chosen to ensure that the breast shape was not violated and to confine the scars to the breast above the inframammary fold. A, Shows the nevus was treated by a relatively ineffective partial excision in early childhood, then left for the definitive excision until after thelarche. B, Close-up view of the expanded graft on the breast 7 years after excision and graft. C, Both the breast and the donor site scar in the groin along the bikini tan line.
placed over the flanks allow for construction of large transposition "angel wing" flaps, which can be frequently advanced to cover most of the back and buttocks. Expanders are placed either cephalad or caudal to the nevus, typically using 500- to 550-ml expanders in infants. These expanders can be inflated to as much as 1200 ml each and provide substantial transposition flaps with excellent blood supply and the ability to cover considerably longer distances than the typical advancement flaps used by many surgeons. Careful design of these flaps to include large paraspinous perforators increases their length and survival. The routine use of hand-held Dopplers preoperatively to map out these vessels is very beneficial. Dissection of tissue expander pockets is as described above and is usually started at 6 months of age while the infant skin is most mobile. Expansion of the buttock and upper thigh skin used in combination with repeat expansion of previously expanded and transposed flaps will allow coverage of the entire buttock area with the scars at completion in a very favorable position to avoid late contracture problems. "Intermediate-size" giant nevi in the mid-back have in some cases been excised by others (who prefer to use expanders as little as possible) without the use of tissue expansion, but we believe that even in these cases the excision is simplified by the added tissue gained through the expansion process. In these instances the excision is accomplished with gradual recruitment of surrounding skin and serial excision.

The shoulder area along with the adjacent neck and deltoid area presents additional challenges of reconstruction in an area with varied contour, skin thickness, and a propensity for wide and often hypertrophic scarring. Expansion of the upper back and anterior chest to recruit tissue into this area may potentially distort the breast and carry the scars that much further down the back. Although excision and grafting would be considered by many as the treatment of choice, the contour defect that may remain, as well as the color and texture difference expected with an extensive graft, have made use of this option less than ideal. In an effort to address these concerns, we have applied microvascular techniques to our armamentarium of procedures for treatment of giant nevi. Given the size of the defects that may need to be covered in this area, we have elected to use either a preexpanded or nonexpanded free TRAM flap, recognizing that the flap will need contouring after inset (Figures 65-16 and 65-17). The contouring is done readily with liposuction in combination with final excision of the border of the nevus, which is left in place at the time of the initial flap inset. Although novel in approach, we believe that the outcome seems to warrant this innovation.

**LARGE AND GIANT CONGENITAL PIGMENTED NEVI OF EXTREMITIES**

Our management of CMN of the extremities has evolved over the last few years, as well. The choice of technique is determined by the location and extent of the lesion. Although tissue
Figure 65-15. Giant nevi of the mid-to low back with a bathing trunk distribution require a different flap design. A, This 3-month-old presented with involvement of the entire perineal and perianal area, buttocks, and back. B, Two 500-ml expanders were placed cephalad to the nevus at the same time some upward advancement of buttock/thigh skin was carried out without expansion. The expanders are shown expanded to approximately 700 ml each over 12 weeks. C, Expanded transposition flaps shown before placement of new expanders. D, After expansion of both the previous back flaps and the buttock/thigh flaps. E, The result after excision of remaining nevus except for a thin strip of nevus along one edge of the anal verge, left for excision once other scars were stable (to minimize risk of circumferential scar or wound problem during flap transfer).
Figure 65-16. This 19-year-old presented with a giant nevus of the shoulder and neck. It was felt that grafting would result in a poor aesthetic result and advancement of anterior skin from the breast area, and posterior skin from the back would carry risk of additional scarring. **A** and **B**, The lesion is shown from anterior and posterior views. **C** and **D**, The result after reconstruction with an expanded free TRAM flap, which was contoured after initial inset using liposuction. The contouring was done at the time the final excision of a rim of nevus, left bordering the inset flap. Note excellent contour of both neck and shoulder with this approach.

expansion has been a primary modality of treatment in the head and neck and is also readily accomplished in the trunk; tissue expansion for excision of nevi of the extremities has not been as readily accomplished. This fact is more the result of the limitations imposed by the geometry of the extremities and limitations this geometry imposes on flap design and movement than the greater likelihood on expander complications in the extremities. Although the risk of expander complications can be significantly reduced with careful planning, placement through a remote incision, and at times endoscopic expander placement, the greater likelihood of scars imposing a restriction on later movement and unsightly contour defects after reconstruction still present a significant concern. When used, expanders must be carefully selected to ensure placement without folds or edges that will compromise the overlying flap or risk breakdown of the incision used for the expander placement. Expander placement through a distant incision, possibly aided by the endoscope, may minimize the risk of expander exposure. The expanders will vary in size based on where on the extremity they are being placed, the size of the lesion to be excised, and the age of the child at the time of treatment. Expanders vary from 100 to 500 ml. This is an area in which the replacement of an expander in the same pocket with an even bigger expander and continuation of expansion before nevus excision may be of
great efficacy (so-called in situ serial expansion). This approach may involve significantly less risk of complication than attempting to reexpand the previously advanced skin flap with its adjacent scar.

Lesions of the deltid can be addressed by expanding the upper back/scapular area and transposing an epaulet type of flap into place. This will avoid potential narrowing of the proximal extremity and scarring into the axilla with limitation of movement. We have applied a similar principle to that mentioned for the shoulder and neck for the entire shoulder and upper neck using a large expanded free TRAM flap with good success. As we move further down the upper extremity, our previous choice for circumferential lesions was the use of either split-thickness or, more ideally, expanded full-thickness skin grafts. Again with this approach we are faced with the aesthetic problems of poor color, contour, and the potential functional problems of graft contracture and loss of range of motion. To that end, we have approached some of these lesions with either expanded or nonexpanded pedicle flaps from the flank/abdomen (Figure 65-18). These flaps can provide the supple skin with subcu-
taneous tissue necessary to avoid carrying scars up or down the arm (that might be needed to move adjacent expanded skin flaps) and leave the donor site in a relatively less visible area. Although this approach requires more staging than simple excision and skin graft, the ultimate outcome both aesthetically and functionally is superior. When it comes to large and giant nevi of the hand, skin grafts, particularly expanded full-thickness grafts, remain our treatment modality of choice. An expanded full-thickness skin graft can provide both excellent aesthetic and functional results in this area (Figure 65-19).

The lower extremity provides an even greater challenge than does the upper. Large but not giant nevi of the thigh generally lend themselves well to excision using expansion, at times carrying out a partial excision at the time of tissue expander placement. Tissue expansion is effective for medium to large lesions of the lower leg, as well, but the limitations of the area impose themselves more the farther distal the lesion. Expanded full-thickness grafts have provided a means of improving on
Figure 65-18. In an effort to improve on the aesthetic result of excision of a near-circumferential nevus of the arm, where a graft frequently leaves a contour defect, and expansion of flaps above and below may even if able to cover the defect require carrying scars above and below the area of nevus, a pedicled flap is planned from the adjacent flank. A, Lateral view of the nevus. B, Posterior view of the nevus and expander in the flank with outline of the proposed flap. C, Anterior view of the initial flap attachment. D and E, Two views of the inset flap after complete excision and flap inset. Note maintenance of soft tissue contour and absence of any scar outside the initial area of involvement.
Figure 65-19. An expanded full-thickness skin graft used for treatment of a giant nevus of the dorsum of a hand. A and B, Preoperative and postoperative views of the nevus and graft 5 years after reconstruction. C and D, Additional view of the grafted hand and the groin donor site.

Figure 65-20. The problem contour defect after excision of nevus to fascial level and graft (even full thickness) on the extremity. A, The nevus of the leg just distal to the knee is shown before excision. B, The result 1 year after coverage with an expanded full-thickness skin graft shows reasonable scar maturation and graft color but poor contour restoration.

the relatively poor aesthetics of split-thickness skin grafts in this area but still fall short of ideal, particularly when dealing with circumferential lesions (Figure 65-20). When planning skin graft reconstruction in these cases, it is recommended that the reconstruction be carried out in two procedures, addressing first the posterior and then the anterior aspect of the lesion. Scars are ideally placed along the mid-lateral and mid-medial lines of the extremity. With this sequence of procedures, an untoward flexion contracture after the posterior excision might be addressed as the anterior half of the nevus is excised. With time and more experience, we would hope that the principles of reconstruction with free tissue transplantation and prefabricated flaps may provide for improved aesthetic and functional results in the lower and upper extremities. As with the hand, treatment of large and giant nevi of the feet are best addressed using skin grafts, with expanded or nonexpanded full-thickness grafts being of some benefit in terms of long-term durability (Figure 65-21).
Figure 65-21. A and B, This stocking distribution giant nevus was treated initially with excision of the anterior aspect of the leg and foot and reconstruction with a split-thickness skin graft, then coverage of the plantar aspect of the foot and toes with an expanded full-thickness skin graft from bilateral lower abdomen and groin area. The result is shown 3 years after excision. C, A portion of the lesion of the plantar foot showed atypical cell changes before excision. D, Plantar view also shown at 3 years after reconstruction of the foot with some callous formation but stable, durable graft coverage.

OUTCOMES

The treatment of congenital nevi, whether pigmented or nonpigmented, follows similar principles based on size and location of the lesion, risk of malignant change, and age of the patient. Assessment of outcomes must consider issues of the effect of treatment on the risk of malignant change, the aesthetic appearance after reconstruction, and the potential functional consequences of the surgery. To a lesser extent, we also must look at the varied treatments in terms of how they affect hospital stay and how well they are tolerated by the patients.

In regard to the risk of malignant change, we would hope that continued evaluation of large series of patients with congenital pigmented nevi would one day answer the continued debate as to whether these lesions carry a risk of malignant degeneration or not. Although there are institutions that state that they have yet to see a case of malignancy in a giant congenital nevus, many series state otherwise and we ourselves have seen three patients develop malignancies in a giant nevus and go on to die from extensive metastases. With much variation in reporting, differing classifications of nevus by size, and variation in the age of the different populations followed, we may never have an answer. Certainly until we do have a more definitive answer, we must proceed on the premise that these
lesions present both a risk of malignant change and a significant stigma because of their appearance. Treatment must be directed at both of these concerns. Outcome data at this time is limited to subjective assessment of different treatment options and assessment of how effectively these different options accomplish the removal of the lesion. However, it may be of some significance that, as of today, none of the patients who have undergone excision in our series has developed a malignancy, with follow-up of the earliest patients being greater than 15 years. One child who underwent excision of a borderline deviation melanoma in infancy is healthy without nevus or tumor at 14 years after surgical excision of her giant nevus.

Complications of the surgical procedures described within this chapter are primarily related to use of tissue expansion. Within our early group of patients undergoing excision and "large segment" split-thickness skin grafting, we had no graft losses. Although we have not reviewed our expander complications specifically in nevus cases only, our numbers give a general idea of the low risk of adverse outcomes from expansion. In fact, given the inclusion of complex wounds in the series, we would expect the complication rate to be somewhat lower in the current group. In our review of expansion in 1982-April 4, 1999, 622 expanders were used in 412 patients, with 69% for treatment of nevi. The patients ranged in age from 12 days to 19 years, averaging 4.7 years. The complications were similar to all series on expansion in respect to infection, exposure, and expander leakage. The complication rate by number of patients was 16% and by number of expanders was 12%, with failure rates (need to seek another reconstructive technique to address problem) of 3% and 2%, respectively. Of the expanded full-thickness skin grafts, 2 of 15 in this series had areas of minor loss. These results strongly support the safety of this treatment modality.

Although we have seen a reduction in inpatient stays even for children undergoing extensive skin grafting procedures, we also have noted that hospital stays of greater than 23 hours are rarities for patients undergoing nevus excision with tissue expansion. With increased experience, we have seen continued reduction in the complications of tissue expansion; this reduction in complications is occurring at the same time that we have observed continued increase in the effectiveness of the expanded tissue. With parental involvement in the expansion process, greater than two thirds of the expansion are performed at home, thereby reducing trips to the clinic and easing the anxiety of the children undergoing expansion. This shift of responsibility has not been at the expense of any increase in complications. Finally, in comparison of our patient population with similar-size lesions in similar locations treated with different treatment modalities, we have had the opportunity to compare outcomes of these procedures from both an aesthetic and a functional point of view.

Let us summarize these thoughts. In 1988, we presented a protocol for treatment of large and giant congenital pigmented nevi (greater than 2% TBS) based on a comparison of different treatment modalities in different body regions. Each area—head and neck, trunk, and extremity—seemed to lend itself to different treatment options in accomplishing early nevus excision and reconstruction. To date, we have now treated 200 patients (from 1979 to 1997). Evaluation of this larger group has now demonstrated both the benefits of the previously described protocol and its weaknesses. Most of these are described above, but we should summarize them here.

We have seen little change in our approach to treatment of large congenital nevi of the head and neck. Tissue expansion is still the treatment modality of choice. However, as with other regions of the body, experience has demonstrated significantly better results in terms of hair direction, hairline restoration, and efficacy of coverage of larger surface areas (with reduced need for serial expansions) using expanded transposition flaps versus simple advancement of the expanded tissue. Although we have accepted the use of larger expanded full-thickness skin grafts for nevi of the combined periorbital area and medial cheek, we have confined the grafts as much as possible to the lids and have completed aesthetic unit reconstruction with expanded cheek flaps. Thoughtful integration of simple partial nevus excision and flap advancement or rotation before initial tissue expansion has seemed to reduce the need for serial expansion in the midfacial area and thereby has minimized some of the risks of expansion in this region. In addition, recent experience with prefabricated flaps, either carried on a transposed superficial temporal artery pedicle or transferred by microvascular technique from a more distant donor site, may further limit the need to use skin grafts in the reconstruction of some of these lesions (e.g., in the nasal region).

One of our greatest advancements has come in the treatment of giant nevi of the trunk, where improved flap design has allowed the excision of much larger nevi of the posterior trunk and buttocks. This is an area in which previously we believed there was little role for tissue expansion because of well-known problems with serial flap expansion and advancement on the back (i.e., increased likelihood of expander exposure and continuing decrease in the tissue gain). Previously, we recommended large segment grafting with nonmeshed split-thickness skin grafting. With continued experience in expansion and with a switch to the use of expanded transposition flaps, even the excision of giant nevi covering the entire buttocks, perineum, and perianal areas may be reconstructed without skin grafting.

The treatment of large and giant nevi of the extremities has been seen to improve as well, yet not to the extent seen in other regions. Although we accept the fact that large lesions, particularly those that are circumferential, are best treated with skin grafts, long-term follow-up of these patients has demonstrated a poor aesthetic result of many of these reconstructions and, on occasion, poor durability of the grafts, as well. We have just entered the era in which some of these problems are being addressed with use of pedicled flaps from the trunk and free tissue transfer from distant sites.

Although some surgeons might consider the innovations discussed in this chapter overkill for treatment of congenital nevi, we cannot diminish the psychologic impact of many of
these large and very visible lesions without aiming for optimal aesthetic restoration, as well as treatment of a potentially pre-malignant lesion. It is through continued outcomes research that these latter "innovations" and others will prove their value.

REFERENCES