Treatment of large and giant nevi

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Congenital melanocytic nevi (CMN) are composed of clusters of nevo-melanocytes that are generally present at birth but occasionally arise as late as several years. These lesions arise from melanocytic stem cells that migrate from the neural crest to the embryonic dermis and upward into the epidermis [1]. They may also migrate into the leptomeninges. Although small pigmented nevi are present in 1 in 100 births [2], large nevi are present in only 1 in 20,000 births [3], and the giant lesions are even less common [4]. As a result, most surgeons have little experience with them and little opportunity to develop a rational protocol for their treatments.

The appearance of these dark, often hairy lesions over a large portion of a newborn infant's face, trunk, or extremity is often devastating to parents who have been anxiously awaiting the birth of their child. Early consultation with a pediatric plastic surgeon or pediatric dermatologist can help educate the family and decrease the stress of the situation by providing concise information about the nature of the nevus, its natural history, and the options for its management. Families can accept even the news of a multiple-stage reconstruction over many years if it is presented in a compassionate manner.

In nearly 25 years of pediatric plastic surgery practice, the senior author has had the opportunity to treat over 270 children with large and giant pigmented nevi. This experience has afforded the opportunity to compare various treatment approaches and determine which techniques have been most effective in achieving optimal aesthetic and functional outcomes. This article discusses the rationale for treatment and summarizes the authors’ current thoughts on planning and accomplishing their treatment goals.

**Definitions of size**

It is important to have a frame of reference in discussing the treatment of CMN. Multiple definitions have been used, and without some uniformity it is difficult to compare different studies. The authors believe that the following definitions are becoming accepted and used in most studies: Small nevi are those measuring 1.5 cm or less, medium nevi measure from 1.5 to 19.9 cm, and large nevi are 20 cm or greater [5]. Giant nevi are a subset of large nevi that measure 50 cm or greater [4]. Another definition of large and giant nevi classifies them as those that cover 2% or more of total body surface [3,6,7].

During the first 6 months of life, some nevi can appear to “grow” significantly as tardive pigment becomes more visible. Some satellite nevi may become visible for the first time over the first 2 to 3 years (tardive CMN) [4]. After the first 6 months, the lesions grow proportionally to the particular area of the body involved. The diameter of the lesion grows by a factor of 1.7 times in the head, 3.3 in the thigh and leg, and 2.8 in the torso, arms, hands, and feet. The large nevi are at least 6 cm in diameter on the infant’s body and 9 cm on its head [4].
Rationale for and timing of treatment

Two immediate concerns face the family of a child with a large or giant nevus. The first is the risk of the child’s developing melanoma [3–9], and the second is the stigma of this very visible lesion and how it will affect the child’s psychological development. Although the exact risk of malignant melanoma may never be determined, the early treatment of these lesions, provided it leads to the necessary aesthetic and functional outcome, may alleviate concerns about the child’s appearance and significantly reduce the risk of malignant degeneration [6,7,10].

In the literature, the estimated risk of developing melanoma ranges from 2% to 31% [3–7]. The different populations and numbers in these studies explain the wide variance. In a retrospective study, Quaba and Wallace [3] examined patients with CMN covering more than 2% of the total body surface and found the melanoma risk to be 8.5% during the first 15 years of life. Sandmark et al [11] have quoted a risk of 6.7% in childhood. Marghoob et al [4] have quoted a lifetime risk of 4.5% to 9% for melanoma arising in large and giant CMN. Approximately 50% of the malignancies that develop in large CMN do so in the first 3 years of life [12], 60% by childhood, and 70% by puberty [12,13].

Another factor that needs to be considered and discussed with families is the issue of neurocutaneous melanosis (NCM). Recent reports have demonstrated the association of nevus cells in the leptomeninges in a percentage of children with large nevi in an axial orientation or those with an extensive number of satellite nevi [14–16]. Although symptomatic NCM is characterized by mental retardation, hydrocephalus, and seizures, many children are asymptomatic [14,15]. These children can be identified by T1 shortening in MR imaging. Foster et al [15] reported that 23% of at-risk patients had evidence of central nervous system involvement (melanotic rests within the brain and meninges) on MR imaging. Marghoob and Dusza [17] have seen this finding in only 3% of children in the Nevus Outreach Registry of over 600 patients. The latter figure coincides with the authors’ experience. Although the presence of a lesion on MRI does not typically alter the decision to treat or not treat a child with a large or giant nevus, the approach may be altered in cases of symptomatic NCM.

The rationale for early treatment of large and giant nevi has four components. These are (1) the presence of the greatest risk for malignancy in the first 3 years, (2) the elasticity and healing capacity of the skin in the early years, (3) the greater patient tolerance of surgery at this time, and (4) the psychological benefit [6,7]. Taking all this into account, and assuming the child is otherwise healthy, the authors begin treatment of the large and giant nevus by 6 months of age in most cases, provided they have seen the child from early infancy. Although many of the tissue-expansion procedures used in treatment of giant nevi can be applied to older children and selected adults, the intolerance for repeated procedures and the decreased elasticity of the skin may make the excision of extensive lesions impractical in older patients.

Treatment of large and giant nevi

The treatment of large and giant nevi is controversial [3–12]. Many feel that the risk of degeneration is too low to warrant the unsightly scars or grafts that may follow treatment. Others feel that, in the presence of NCM, the greatest risk lies within the central nervous system, so the excision of the cutaneous lesion can only have limited benefits. However, the appearance of these lesions clearly produces a stigma with significant psychological implications. Removal on this basis is often warranted. The challenge for the surgeon involved in treating these often complex lesions is to develop treatment modalities that not only accomplish the excision of all or most of the nevus but also lead to an optimal aesthetic and functional outcome. Where the second requirement cannot readily be met, it is best not to operate and to have the child closely followed by a dermatologist with experience with giant nevi.

Treatment choices include observation, dermabrasion [18] or curettage [19], and staged excision and reconstruction [6,7,10,12]. Some giant nevi are so extensive as to have no available normal “donor” tissue for reconstruction. In other cases, the family situation or lack of available resources may speak for a less “aggressive” approach. The treating physician should be well versed in the available treatment options, honest about the potential risks and outcomes of the various surgical modalities of treatment, and able to present these to the family or patient (when the latter is old enough to understand).

Dermabrasion [18] and curettage [19] are both techniques that have been applied in the neonatal period in an effort to remove the more concentrated population of nevus cells near the lesion’s surface. The technique can be effective in reducing the overall
nevus “cell load” but cannot fully remove the nevus, because of the well-known depth of nevus cells in CMN [20]. Although this treatment may result in significant lightening of the color of the lesion, it is quite common to see later “bleed-through” of the deeper nevus, with gradual darkening and reappearance of the lesion. This result may present a difficult treatment problem in visible areas like the face, where other techniques for excision may then be less well tolerated.

Many large and giant nevi can be excised completely or nearly completely with very acceptable aesthetic outcomes. In 1988, the authors presented an overview of their first 78 patients with large and giant nevi (2% of total body surface or greater), focusing on the issue of which treatment techniques most effectively allowed early excision and reconstruction [6]. Since that time, the senior author has treated an additional 193 patients, for a total of 271 patients, and during the intervening 16 years he has modified the treatment approach to address the challenge mentioned earlier. The optimal choice of treatment still varies by body region, and the remainder of this article summarizes the authors’ thoughts on these different treatment modalities.

Scalp

Tissue expansion is well recognized as the treatment modality of choice for excision and reconstruction of large and giant nevi of the scalp. As surgical experience increases and planning improves, larger nevi can be excised and the defects reconstructed with fewer serial excisions and better restoration of normal hair patterns. Rectangular expanders with remote injection ports are used here and in all other body regions, with the expanders in place for an average of 10 weeks. Expanders are typically injected weekly (increased to every 4 to 5 days in some cases). The typical expanders used in the scalp vary from 250 to 500 mL in size. Treatment starts as early as 6 months, with some cranial molding expected by the time the expanders are removed, but with no instance of long-term cranial deformity noted (remodeling usually occurs over 3 to 4 months). The most significant modification the authors have made in their technique from early cases to the present is the increased use of expanded transposition flaps rather than advancement flaps [21]. The benefit of this modification is most apparent in the use of the expanded occipital transposition flap for coverage of the entire parietal scalp and reconstruction of the temporal hairline and sideburn. Although it is not always possible following a single expansion, this approach has been successful in multiple cases and provides for an optimal hair direction and hairline.

Face

Large and giant nevi of the face present some of the greatest challenges in treatment of these lesions. Certainly, these are the most visible nevi with which the patient and family must deal and the ones that are most likely to be associated with significant psychological sequelae. They also represent the area where unsightly scarring is most readily visible; consequently, the planning and execution of the reconstructive plan must be very detailed. A description of all the nuances of treatment of facial nevi is beyond the scope of this paper; this topic has been covered previously in both the authors’ work [10] and that of Zuker et al [22]. What follows is a summary of the highlights.

Tissue expansion of the hemiforehead for unilateral lesions or the bilateral or lateral forehead for central lesions can very effectively treat even extensive lesions. Because many of these nevi involve the adjacent scalp, the combined “attack” on both of these regions often facilitates the excision and lessens the number of stages required. Flap advancement, rotation, and transposition each play a role in assuring that the hairline, eyebrow, and distance between the two are not disturbed. Nevi of the cheek are best reconstructed with expanded or nonexpanded postauricular/neck flaps. The reconstruction of the entire aesthetic unit of the cheek may require two expansions. Again, the use of a transposition flap significantly reduces the risk of downward traction and distortion of the lower eyelid and canthus, which are seen as common sequelae of direct advancement of expanded flaps from below the mandible to the cheek.

Expanded or unexpanded full-thickness skin grafts [23] have been used effectively for excision and reconstruction of nevi of the periorbital and eyelid area and, occasionally, of the nasal dorsum. Although single lids may be grafted from a postauricular donor site, a single large expanded full-thickness graft from the supraclavicular area can reconstruct eyelids, canthus, and the region between eyelid and brow without the multiple “seams” that follow the use of many smaller grafts. Recently, the authors have combined the excision of nevi of the lateral forehead and nasal dorsum, borrowing part of the expanded forehead flap to cover the na-
sal dorsum while advancing the remainder to the temporal area.

Extensive nevi of the central face (nose, lips, chin) are some of the most challenging that we have to deal with, and their treatment requires both ingenuity and a solid grasp of plastic surgery reconstructive techniques.

**Trunk**

As the authors’ series of cases enlarged and their opportunities to view the long-term outcomes of earlier cases increased, it became clear that the role of skin grafting has significantly decreased [7,21]. Physicians need to decide whether the aesthetic and functional outcome of a trunk covered with split-thickness grafts (or, even worse, with meshed split-thickness grafts) is an acceptable alternative to leaving the nevus untreated. If one follows the authors’ earlier approach of using large segments of nonmeshed split-thickness skin and confines its use to the back, then the aesthetic outcome may be acceptable. However, the authors confine use of this treatment to cases that are so extensive in size as to have no expandable skin available, and where particularly atypical features of the nevus prompt resection. Today, these cases are few and far between.

Tissue expansion can be very effective on the anterior trunk, provided that the lesion is confined either to the lower abdomen or central abdomen and that there is sufficient uninvolved skin above or above and below the nevus to expand. Expansion must be avoided in or around the area of the breast bud in females, and lesions of the breast should be left until after breast development, regardless of the psychological implications of delaying the treatment till that age.

The use of expanded transposition flaps has allowed excision of nevi of the upper back and buttock/perineal region, where previously it was thought that only skin grafting was possible [21]. Tissue expanders in the 500- to 750-mL range are used most commonly in infants and young children. Serial expansion with careful planning has made possible the excision of progressively larger nevi of the back and buttocks, with excellent outcomes. Another tool for reconstruction of giant nevi of the upper back, shoulders, and neck has been the expanded free transverse rectus abdominus myocutaneous (TRAM) flap, which can be positioned in the upper back and posterior neck or shoulder, then re-expanded, contoured, and draped about the neck and shoulders [24].

**Extremities**

Large and giant nevi of the extremities present a challenge that is still not fully met. In past years, the authors were willing to excise extensive circumferential nevi of the extremities (both upper and lower) and replace the nevus with either nonmeshed split-thickness skin grafts or large expanded full-thickness skin grafts. However, long-term follow-up of these patients typically demonstrated contour deformities and unacceptable aesthetic outcomes. Concerns were also expressed about both the durability of these grafts and their ability to keep up with normal extremity growth.

Tissue expansion has been of some help in treatment of smaller lesions, where there is available tissue proximal and distal to the lesion and the lesion is confined to a fairly small segment of the limb. The geometry of the extremity, as well as the limited flexibility of the skin (particularly in the lower extremity) makes regional expansion of limited use [24].

In the past decade, the authors have begun to find a way around these limitations, using large expanded transposition flaps from the scapular region to cover the upper arm and shoulder and expanded or nonexpanded pedicle flaps from the flank and abdomen for circumferential nevi from the elbow to the wrist. Expanded full-thickness skin grafts have been used effectively for the dorsum of the hand, with excellent aesthetic outcomes [23,24].

Although pedicled flaps are not readily available for coverage of more extensive lesions of the arm, thigh, or leg, the authors have had some success with expanded free flaps from the abdomen and scapular region [24]. These procedures have been used only in very carefully selected cases, and the optimum timing of these complex reconstructive procedures is still under consideration. Again, we must critically evaluate each case and be assured that the treatment chosen is likely to provide an aesthetic and functional outcome that is better than accepting the lesion and dealing with the associated psychological issues with appropriate support.

**Satellite nevi**

Satellite nevi may appear anywhere over the course of the first few years of life, and their number seems to correlate directly with the likelihood of NCM [16]. They may vary in size from small to medium lesions. To date, no case of melanoma has been reported arising in a satellite nevus [16,17]. With this in mind, it is generally agreed that the pri-
mary reason for excision is an aesthetic one. The authors generally excise some of the larger lesions early, often with serial excision, and leave the smaller lesions until the child expresses specific concerns about them. A significant benefit may also result from excising multiple satellite nevi on the face before the child enters his or her school years.

Summary

Although the exact risk of malignant degeneration may never be determined, there is still evidence that large and giant nevi carry this potential. Excision and reconstruction are warranted, provided that they can be accomplished with an optimal aesthetic and functional outcome, but experience has demonstrated that some extensive lesions are best left to observation by the dermatologist (with selective surgery if atypical areas arise). NCM is a well-recognized disorder whose full implications have yet to be determined, but children with large and giant nevi, particularly in axial distribution, should be evaluated. The same goes for those with multiple satellite nevi. The presence of NCM is not at present a contra indication to treating the cutaneous lesion.

Experience with a large population of children with large and giant CMN has demonstrated that thoughtful application of the full spectrum of reconstructive options, heavily weighted toward the use of tissue expansion (as well as expanded pedicled and free flaps) can result in total or near-total excision of many of these extensive nevi with predictably good outcomes (Figs. 1–4).

Fig. 1. (A,B) One-year-old child with giant nevus of forehead and scalp, with two expanders in place for first-stage excision of nevus following 12 weeks of expansion. (C) The expanded forehead flap and scalp flaps in position following excision of the greater part of the nevus. The remaining nevus is in the right lateral forehead and temporal region. (D) Appearance 9 months after excision of remaining nevus, with symmetric hairline and brow position.
Fig. 2. (A) Six-year-old child with large nevus of left medial eyelids and brow, with extension to canthus and lateral nose. (B) Defect following excision of the nevus. (C) Reconstruction with single large postauricular full-thickness skin graft (FTSG; donor site closed with additional FTSG from groin). Medial brow reconstructed with island flap from temporal region. (D) Eight months after the reconstruction, the full-thickness graft is blending in well and has fully reconstructed the complex contours of the defect and lids. The hair growth in the reconstructed brow is still sparse but appears to be increasing. As typically planned in these cases, the nevus of the ciliary margin is left in place to avoid potential functional disturbance of the lid and canalicular function.

Fig. 3. (A) A giant nevus of the back with atypical and variegated pigmentation is approached with placement of four expanders (2 – 500 mL above and 2 – 350 mL distal to the nevus). (B) The greater part of the nevus is excised following this first expansion, which was carried out over 12 weeks. The remaining nevus will be excised with a second round of expanders after 4 to 6 months of healing.
Fig. 4. (A) A 2-and-a-half-year-old with a giant nevus extending circumferentially around the forearm from elbow to wrist, with additional satellite nevus in the upper arm. (B, C) The arm is positioned against the flank and abdomen after expansion of the site and before placement of the arm within the expanded pedicle flap. (D) The arm is placed for 3 weeks within the expanded “tunnel,” and the pedicle is gradually tightened with through-and-through bolster sutures, gradually reducing the blood flow through the pedicle. (E) The volar surface of the reconstructed forearm with the healing donor site visible on the abdomen. Residual nevus is still present at the wrist and elbow and at the satellite. (F) The remaining border of nevus at the edges of the flap is excised. Clearly noted are the excellent contour and quality of the skin of the forearm gained with this approach.
References


