

# Giant Congenital Melanocytic Nevi

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**Background:** Giant congenital melanocytic nevi are rare lesions with the potential to regress into malignant melanoma and/or neurocutaneous melanosis. Appropriate investigations include a screening magnetic resonance imaging scan, neurologic evaluation, and serial clinical observations looking for the development of these complications. Numerous excisional and nonexcisional options have been described for the management of giant congenital melanocytic nevi.

**Methods:** A MEDLINE search was performed to obtain all relevant citations.

**Conclusions:** To successfully treat these complex lesions, the plastic surgeon must understand the disease process, the natural history and complications, and the options for treatment. (*Plast. Reconstr. Surg.* 124 (Suppl.): 1e, 2009.)

**G**iant congenital melanocytic nevi are rare, disfiguring lesions present at birth associated with complications of malignant melanoma and neurocutaneous melanosis. These lesions often invoke significant psychological distress in the parents and require a thorough discussion with the treating physician regarding the natural history, complications, and available treatment options. The treatment of giant congenital melanocytic nevi can be divided into nonexcisional options (i.e., dermabrasion, laser ablation, curettage, chemical peel) and excisional options (i.e., staged or en bloc excision with primary closure, skin graft, flap, skin substitute, or tissue expansion reconstruction). The current review provides an overview of the definition and epidemiology; histology, embryology, and genetic basis; clinical characteristics and evaluation; natural history and complications; and options in management of giant congenital melanocytic nevi.

## DEFINITION AND EPIDEMIOLOGY

Approximately one in 100 infants is born with a congenital melanocytic nevus<sup>1</sup>; however, the incidence of giant congenital melanocytic nevi has been estimated at one in 20,000 live births.<sup>2</sup> A melanocytic nevus is a pathologic accumulation of melanocytes of neuroectodermal origin in an ectopic location.<sup>3,4</sup> Melanocytic nevi can be classified into nevi appearing at birth (i.e., congenital melano-

cytic nevi) or nevi appearing after birth (i.e., acquired nevi). Giant congenital melanocytic nevi are always present at birth; however, an entity referred to as “early-onset nevus” may present in infancy exhibiting properties similar to those of congenital melanocytic nevi.<sup>5,6</sup>

Giant congenital melanocytic nevi have been described historically by various nomenclatures, varying with anatomical location. More recently, there have been several definitions proposed for giant congenital melanocytic nevi, including congenital melanocytic nevi affecting a significant portion of an anatomical area (i.e., face and hand),<sup>7</sup> congenital melanocytic nevi measuring 20 cm or more in greatest diameter,<sup>8</sup> congenital melanocytic nevi covering more than 1 percent total body surface area in the head and neck region or 2 percent total body surface area elsewhere in the body,<sup>9,10</sup> congenital melanocytic nevi greater than 100 cm,<sup>11,12</sup> and congenital melanocytic nevi that are unable to be excised in a single operation.<sup>13</sup> Bittencourt et al.<sup>13</sup> have appropriately defined “giant” as congenital melanocytic nevi measuring (or predicted to reach) 20 cm or greater in diameter in adults. This number correlates with a diameter estimated at 6 cm on an infant’s body and 9 cm on an infant’s head.<sup>15,16</sup>

## EMBRYOLOGY, GENETICS, AND HISTOLOGIC CHARACTERISTICS

Neural crest melanoblasts migrate to numerous sites in the body, including the skin, mucous mem-

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branes, leptomeninges, mesentery, eyes, and ears between the eighth and tenth weeks of gestation.<sup>17</sup> The melanoblasts subsequently differentiate to dendritic melanocytes. Most nevi are classified as acquired because they migrate postnatally to their characteristic dermal and epidermal locations. Abnormalities with neuroectodermal development and arrest during migration or differentiation result in the formation of a congenital melanocytic nevus.<sup>4,11,18</sup>

The genetic basis of congenital melanocytic nevi is not well defined and has been theorized to involve hepatic growth factor/scatter factor. The development and migration of neuroectodermal cells are regulated by hepatic growth factor/scatter factor, and it has been demonstrated that hepatic growth factor/scatter factor is found in large quantities in congenital melanocytic nevi.<sup>19</sup> Overexpression of hepatic growth factor/scatter factor results in abnormal accumulations of melanocytes in the skin.

Histologic characteristics that distinguish congenital melanocytic nevi from acquired nevi include (1) the presence of nevus cells within the lower two-thirds of the dermis and within the subcutaneous tissue (2); nevus cells extending between collagen bundles of the reticular dermis as single cells or cords of cells (3); extension of nevus cells around and into sebaceous glands, hair follicles, eccrine and apocrine sweat glands, blood vessel walls, and nerves (4); a perivascular and perifollicular distribution of nevus cells simulating an inflammatory reaction; and (5) arrector pili muscles that are infiltrated or distorted by nevus cells.<sup>20</sup> Giant congenital melanocytic nevi can have nevus cells that extend beyond the subcutaneous tissue and into the fascia or muscle.<sup>4</sup>

### CLINICAL PRESENTATION AND EVALUATION

Giant congenital melanocytic nevi most often occur on the trunk, followed by the extremities and the head and neck.<sup>6</sup> The clinical presentation often changes with age; a hairless, pale brown, flat lesion at birth may evolve over months and years to have hyperpigmentation and color variegation, hypertrichosis, erosions or ulcerations, a verrucous texture, and nodularity representing neurotization of the lesion.<sup>12,20</sup> Roughly 80 percent of patients with giant congenital melanocytic nevi have solitary or multiple satellite nevi associated with them dispersed over the extremities, trunk, or head and neck.<sup>21</sup> The differential diagnosis for giant congenital melanocytic nevi includes epidermal nevus, nevus sebaceous, capillary malformations, and macular stains.

A multidisciplinary approach should be advocated in the treatment of patients with giant congenital melanocytic nevi. Work-up should include a thorough, regular examination by a dermatologist and input by plastic surgeons early in the treatment course. Continued follow-up is imperative following treatment, because many patients will have positive margins and/or satellite nevi that must continually be monitored for signs of malignant transformation. A clinical psychologist who is familiar with child development should be made available for consultation in treating children of school age who are undergoing treatment, because peer pressure may make certain treatment options more challenging.<sup>22-24</sup>

### NATURAL HISTORY AND COMPLICATIONS

The natural history of giant congenital melanocytic nevi is one of expansion in proportion to the body's growth. Giant congenital melanocytic nevi may endure changes in size, color, topography, and hair development.<sup>6,23</sup> Dark, coarse hair may develop within the first 1 to 2 years of life.<sup>24</sup> Surface pigment may fade with time, and at approximately 10 years of age these lesions can become elevated and verrucous, with hyperkeratosis and coarser hair.<sup>24,25</sup> These changes may stabilize in adulthood. Malignant transformation can be diagnosed by focal growth, ulceration, tenderness, dark pigmentation, pruritus, pain, or bleeding.<sup>24,26</sup>

Complications of giant congenital melanocytic nevi include irritating symptoms (e.g., pruritus, ulceration), psychosocial dysfunction related to the aesthetic appearance, a prolonged treatment course with the potential for surgical complications, malignant degeneration in the form of cutaneous melanoma, and extracutaneous melanocyte deposits. Whereas extracutaneous involvement in mucosal or retroperitoneal locations usually manifest only after malignant transformation, leptomeningeal involvement may present with seizures and other clinical findings associated with symptomatic neurocutaneous melanosis, even without malignant transformation.<sup>27-29</sup>

Giant congenital melanocytic nevi produce significant distortion of form (in addition to function) to the involved anatomical site, often associated with severe psychosocial distress to patients and their families. Local changes that can develop within the lesion include nodularity, ulceration, and hypertrichosis, and systemic involvement can include spinal dysraphism, scoliosis, and limb abnormalities.<sup>24,28,29</sup> The two most concerning complications include malignant melanoma and neurocutaneous melanosis.

### Malignant Melanoma

Plastic surgeons focus primarily on cutaneous melanoma; however, malignant transformation can also occur in extracutaneous melanocytic deposits.<sup>26,28</sup> These areas include mucosal involvement in the gastrointestinal tract, in central nervous system depositions of neurocutaneous melanosis, and in the retroperitoneum. Previous studies reported the risk of melanoma in patients with giant congenital melanocytic nevi to range from 1.8 to 45 percent.<sup>30–35</sup> More recent retrospective reports have suggested the risk of melanoma develops in 2.8 to 8.5 percent,<sup>31,32</sup> and two prospective studies revealed melanoma in 3.3 and 3.8 percent of their respective cohorts.<sup>16</sup> Risk factors for the development of melanoma in a congenital melanocytic nevi include multiple lesions (three or more), large size (diameter >20 cm), and younger age (3 to 5 years).<sup>14,24,32</sup> Fifty percent of diagnosed cases of melanoma in giant congenital melanocytic nevi arise in the first 3 years of life, 10 percent arise later in childhood, and an additional 10 percent arise by puberty.<sup>31–35</sup> Therefore, the vast majority (70 percent) of cases with malignant transformation do so by age 13 years. For this reason, surgical excision is best performed in early childhood. A recent study revealed that patients with giant congenital melanocytic nevi had a 51.6 percent higher risk of developing malignant melanoma compared with the general population.<sup>36</sup>

Features suggestive of malignancy on histologic evaluation include dysplasia of nevus cells, pagetoid nevus cell spread, and proliferative dermal nodules.<sup>20,35</sup> Characteristics of malignant nodule degeneration include spread of melanocytic aggregations outside and lateral to surrounding nevus cells, high mitotic rate, high-grade atypia, inflammatory infiltrate, cell necrosis, and perineural invasion.<sup>20</sup>

To date, there are no available staging or treatment strategies for melanoma in the pediatric population, as such treatment recommendations are based on adult guidelines.<sup>37</sup> Cases of diagnosed melanoma require appropriate staging beginning with a biopsy to determine the lesion's histology (i.e., thickness) and nodal status through sentinel node evaluation and/or complete nodal biopsy of appropriate lesions. Treatment and prognosis are subsequently based on the disease stage (i.e., tumor grade and nodal status).

### Neurocutaneous Melanosis

Neurocutaneous melanosis is characterized by benign and/or malignant melanocytic proliferations in the central nervous system in patients having

a giant congenital melanocytic nevus or three or more smaller melanocytic nevi.<sup>15</sup> Neurocutaneous melanosis results from an error in embryonic neuroectodermal development, with dysregulated proliferation and migration of melanoblasts in the central nervous system.<sup>37</sup> Neurocutaneous melanosis may become symptomatic independent of malignant transformation; intracranial hemorrhages or proliferation of the ectopic melanocytes may result in patients presenting with signs and symptoms of hydrocephalus, seizures, developmental delay, cranial nerve palsies, or a tethered spinal cord.<sup>38–41</sup>

Neurocutaneous melanosis has a propensity to manifest at two peak ages. During infancy, patients with neurocutaneous melanosis may present with increased intracranial pressure, hydrocephalus, or developmental delay<sup>27</sup>; whereas in the second to third decades, patients may present with space-occupying lesions, increased intracranial pressure, or spinal cord compression.<sup>29</sup> Risk factors for the association of neurocutaneous melanosis include the presence of multiple satellite nevi (>20),<sup>37</sup> and congenital melanocytic nevi in a midline location over the trunk and calvaria.<sup>6</sup> In patients with risk factors for neurocutaneous melanosis, a screening magnetic resonance imaging scan of the central nervous system should be ordered between 4 and 6 months of age, before normal brain myelination obscures visualization of melanin deposits.<sup>22,39,40</sup>

Diagnosed cases of symptomatic neurocutaneous melanosis should be referred for further medical and surgical management, including ventriculoperitoneal shunt placement, radiation therapy, surgical excision, or immune modulation.<sup>41</sup> Symptomatic neurocutaneous melanosis suggests a grave prognosis, with death usually occurring within 2 to 3 years of diagnosis.<sup>38</sup> Patients with asymptomatic neurocutaneous melanosis may have a similar lifetime risk for the development of melanoma in the central nervous system as patients with cutaneous giant congenital melanocytic nevi.

The management of asymptomatic neurocutaneous melanosis identified on screening magnetic resonance imaging remains controversial.<sup>40–43</sup> We recommend that these patients be followed with serial magnetic resonance imaging scans during childhood to determine whether there is a change relative to the baseline magnetic resonance imaging scan. Unless such changes can be documented, treatment of cutaneous giant congenital melanocytic nevi should not be altered in the setting of asymptomatic neurocutaneous melanosis.<sup>6</sup>

## TREATMENT OPTIONS

The rationale for treatment of giant congenital melanocytic nevi in early childhood includes the risk of degeneration to malignant melanoma, the psychological benefit to operating in the preschool age years, and patient tolerance for reconstruction, particularly with the use of tissue expanders.<sup>44</sup> Indications for surgical excision of giant congenital melanocytic nevi include surgical resection in cases of established melanoma, prophylactic excision to decrease the melanocytic load, reduction and control of symptoms, aesthetic improvement, psychosocial well-being, and maintenance of function.<sup>22,24,42,44,45</sup> The clinical presentation of a verrucous, hairy, nodular, and occasionally ulcerative giant congenital melanocytic nevus makes screening for malignancy a challenge.<sup>20</sup> Several authors suggest initiating surgical intervention at 6 months of age, with completion by school age.<sup>44,45</sup>

The treatment strategy in the overall management of giant congenital melanocytic nevi remains controversial.<sup>44-63</sup> Some specialists believe the risk of malignant degeneration is so insignificant that total surgical removal is not warranted, whereas others maintain the need for complete surgical excision over several stages. Regardless of one's view of the risk of malignant transformation in giant congenital melanocytic nevi, strong reasons for their surgical removal remain: (1) giant congenital melanocytic nevi produce significant psychosocial distress because of the aesthetic appearance of the lesion; and (2) the skin integrity within giant congenital melanocytic nevi is often poor, with repeated excoriation and ulceration. In the setting of neurocutaneous melanosis, some feel that excision of the cutaneous lesions is of less concern; however, these authors often overlook potential differences in management between symptomatic and asymptomatic lesions. Many treatment options have been postulated for giant congenital melanocytic nevi, including dermabrasion,<sup>46</sup> carbon dioxide laser ablation,<sup>50,62,63</sup> chemical peels,<sup>51</sup> curettage,<sup>47</sup> and excision with various methods of reconstruction. These methods include serial excision,<sup>6</sup> split-thickness skin graft,<sup>26</sup> expanded full-thickness skin graft,<sup>52</sup> tissue expansion,<sup>6,44,54,60</sup> free tissue transfer,<sup>54</sup> and skin substitutes.<sup>55-59</sup>

## NONEXCISIONAL METHODS

Nonexcisional methods for the treatment of giant congenital melanocytic nevi remain controversial, given the inability to obtain a histologic tissue diagnosis and difficulty in posttreatment surveillance. Curettage and dermabrasion have been previously described, whereas chemical peeling and

laser ablation are currently the most common non-surgical modalities. Although these methods may decrease the absolute number of nevus cells, they remove the most superficial of all cells. Criticism of these techniques is that nevus cell density remains highest in the deep dermis, making it more difficult to monitor changes in the residual nevus cells because the more superficial component of any regional population of nevus cells has been removed.

## Chemical Peeling

Phenol chemical peeling was advocated by Hopkins et al.<sup>51</sup> for congenital melanocytic nevi excessively large for excision or lesions for which excision would lead to undesirable scarring (i.e., face). They used the Baker formula for 20 patients with giant congenital melanocytic nevi, beginning with a test area, followed by formal treatment with areas having favorable responses.<sup>61</sup> The number of peels required to achieve the treatment outcome of optimal aesthetic appearance, judged by lightening of the pigmentation or resolution of the lesion, was 2.6 (range, one to nine). The authors noticed improved outcomes in 15 of 20 patients, with the greatest improvement in lightly pigmented, superficial lesions. Their indications were extremely large lesions, lesions in delicate aesthetic locations, parental preference, or cases for which surgery would result in functional impairment. Other authors have had less success with chemical peeling, reporting a high rate of scarring and acceptable results in only seven of 17 patients.<sup>3</sup>

## Lasers

Several lasers have been used for the management of giant congenital melanocytic nevi, including the carbon dioxide, ruby, argon, and erbium:yttrium-aluminum-garnet.<sup>50,62,63</sup> The laser described most often recently has been the 10,600-nm carbon dioxide laser. The carbon dioxide laser's target chromophore is water, and the mechanism of action is a thermal injury to the epidermis and partial dermis with removal of the melanocytic depositions by means of selective photothermolysis. After laser ablation, the tissue heals from the remaining dermal elements and dermal appendages. Authors preferring the use of the carbon dioxide laser offer that the risk of melanoma in giant congenital melanocytic nevi is small, and when melanoma develops, it often occurs in sites distant from the giant congenital melanocytic nevi.<sup>50</sup> Horner et al.<sup>63</sup> used the carbon dioxide laser in 12 patients in childhood with considerable aesthetic improvement; however, half of their patients developed hy-

peritrophic scarring. Reynolds et al.<sup>50</sup> primarily used the carbon dioxide laser in children in the first year of life, with acceptable aesthetic results, although they did not comment on treatment complications. Kornberg et al.<sup>64</sup> described an entity called “pseudomelanoma” of a partially excised pigmented lesion that recurred having atypical cells that histologically resembled a superficial spreading melanoma. Several *in vitro* studies have further concluded that the risk of malignant transformation increases with exposure of melanocytes to laser energy.<sup>65–68</sup>

### EXCISIONAL METHODS

Family education regarding the surgical procedure, complications, and wound and scar management is essential. It has been advocated that surgery should be offered after puberty for lesions that are bothersome for the child, such that the child’s input can be incorporated into the treatment plan.<sup>69,70</sup> Others differ, initiating treatment at 6 months of age, with completion before school age.<sup>48,49</sup> In deciding the optimal surgical timing, it must be acknowledged that half of malignancies within giant congenital melanocytic nevi have been reported during the first 3 years of life. Our preference for the treatment of giant congenital melanocytic nevi is for surgical excision to begin as early in life as can be tolerated by the patient, well before age 3 years. A greater challenge than resection of the nevus is resurfacing of the residual defect, for which several options exist.

#### Serial Excision

If the lesion is amenable to excision in three stages or less, serial excision is often the reconstructive option selected.<sup>6</sup> Serial excision is much less labor-intensive, has better patient tolerance, and has far fewer potential complications than do other reconstructive methods. A 6-month period of convalescence is preferred before successive serial excisions to allow adequate tissue relaxation and scar maturity. However, the majority of giant congenital melanocytic nevi cannot be treated by serial excision alone.

#### Skin Grafting

Reconstruction of the defect following excision of giant congenital melanocytic nevi can reliably be covered by split-thickness or full-thickness skin grafts (expanded or nonexpanded). Full-thickness grafts are preferred because they offer optimization of aesthetics with less contracture. The superior results possible through tissue expansion have rendered split-thickness skin graft-

ing less desirable. We reserve the latter technique for cases in which there is a paucity of donor sites or restricted bordering anatomical regions that limit tissue available for expansion.<sup>6</sup>

#### Tissue Expansion

If the lesion is not amenable to treatment with serial excision in three stages or less, tissue expansion is the preferred modality. Tissue expansion should be considered as a valuable adjunct to traditional reconstructive options, because expanders can be used to create an expanded full-thickness skin graft; expanded advancement, rotation, or transposition flaps; or an expanded free flap.<sup>6</sup> Given such diversity with tissue expansion, it has evolved as a first-line treatment for giant congenital melanocytic nevi. Tissue expansion recruits and creates tissue having the same qualities of normal skin, with similar color, texture, hydration, durability, and hair production.<sup>53</sup> Tissue expansion is generally performed over a 3- to 6-month interval, with inflation performed at weekly intervals.<sup>6</sup> With proper teaching, parents can often perform expander filling in the home setting. During surgery, expanders are removed and a capsulotomy is performed, allowing maximal mobility of the expanded flap. The maximum excision of the lesion is performed and the expanded skin is maneuvered to fill the defect. If additional nevus remains, tissue expanders can be reinserted for staged serial expansion; we prefer waiting at least 3 months before reinsertion of expanders to allow adherence of the previous advanced tissue flaps.

Complication rates from tissue expansion range from 13 to 20 percent.<sup>71–76</sup> Complications are highest for lower extremity expansion, for younger children, and for multistage expansion.<sup>54,71,72</sup> Complications that do not delay the expansion process have been classified as minor (i.e., pain during expansion, seroma, scar widening, and altered aesthetics during expansion); complications that delay reconstruction have been classified as major (i.e., hematoma; infection; expander exposure, extrusion, or failure; and flap ischemia). Well-educated and compliant families who actively participate in the expansion process produce the most favorable results.<sup>71</sup>

#### Skin Substitutes

Skin substitutes offer several advantages as compared with autogenous tissues, including an absence of donor-site morbidity, a readily available and unlimited supply, and decreased operating room time. However, these materials also have several limitations, including fragility, absence of normal skin



**Fig. 1.** A 9-month old boy presented with a giant congenital melanocytic nevus of the upper back, neck, and scalp (*above, left*), for which expanders were placed in the scalp and back (*above, right*). Residual nevus following intermediate stages of tissue expansion (*center, left*). The anterior upper chest is expanded to develop expanded transposition flaps to resurface the involved areas of the anterior, lateral, and posterior neck (*center, right, and below, left*). The remaining nevus will be treated with placement of a final tissue expander in the back below the residual nevus (*below, right*).

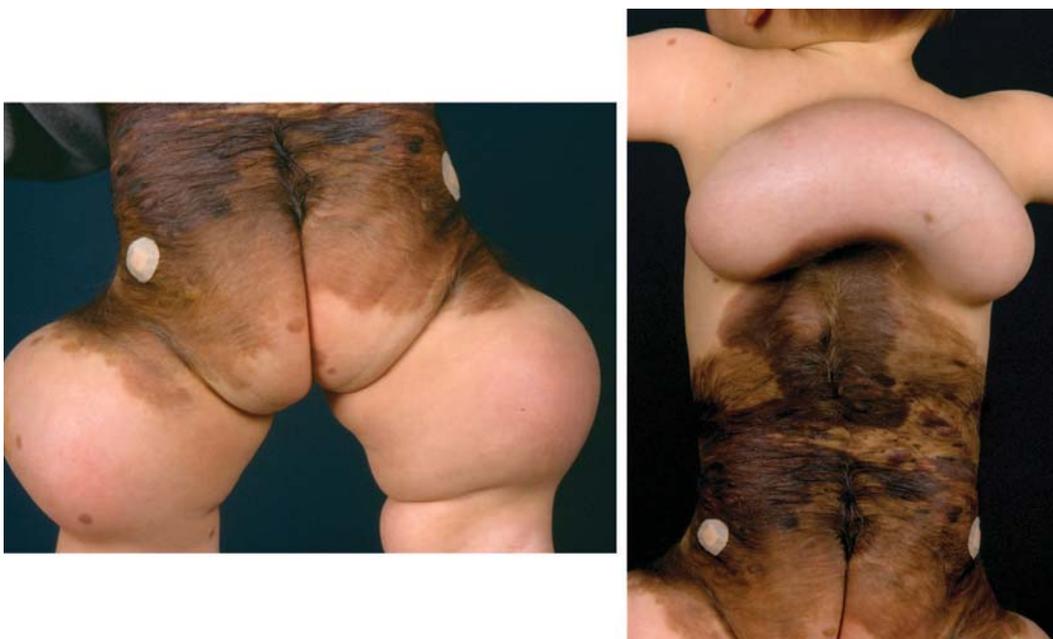
properties, decreased survival in a poorly vascularized bed, increased rates of infection, inferior aesthetic outcome, and high cost.

Several recent reports have suggested dermal, epidermal, or bilayer skin substitutes for the treatment of tissue defects created from the excision of giant congenital melanocytic nevi.<sup>55-59</sup> Dermal substitutes include AlloDerm (LifeCell Corp., Branch-

burg, N.J.) and Integra (Integra Life Science Corp., Plainsboro, N.J.). Epidermal substitutes include Apligraf (Organogenesis, Inc., Canton, Mass.; and Novartis Pharmaceuticals, East Hanover, N.J.), a bilayer substitute, and cultured epithelial autografts (Epicel; Genzyme Tissue Repair Corp., Cambridge, Mass.). Integra has been the most frequently reported, and is a bilayered matrix, with the dermal



**Fig. 2.** A 1-year-old girl presented with a circumferential giant congenital melanocytic nevus of the torso, buttocks, and thighs in a bathing trunk distribution.



**Fig. 3.** Expanders were placed to resurface adjacent tissues of the upper back and bilateral thighs.

layer consisting of porous, cross-linked bovine collagen and shark chondroitin-6-sulfate, that acts as a matrix for the migration of capillaries and fibroblasts from the wound bed. This synthetic layer is eventually replaced by the patient's own collagen. The epidermal layer consists of a polysiloxane (silicone) polymer that acts as a protective layer against trauma and infection, prevents dehydration, and maintains wound homeostasis.<sup>55</sup> After a 3- to 4-week period, the silicone epidermal layer is removed and the dermal layer is resurfaced with a split-thickness skin graft.

No large, prospective studies have been published for the treatment of giant congenital melanocytic nevi with skin substitutes. However, several case reports have described the use of Integra followed by split-thickness skin grafting once the neodermis becomes stable and neovascularized.<sup>57-59</sup> These authors advocate the use of skin substitutes to treat the defect created by excision of giant congenital melanocytic nevi in the setting of limited donor sites. However, results currently achieved with skin substitutes provide inferior form and function compared with reconstruction with expanded flaps from adjacent tissue.

## SURGICAL MANAGEMENT OF SPECIFIC ANATOMICAL REGIONS

### Trunk

Tissue expansion is the mainstay of treatment for giant congenital melanocytic nevi of the trunk. The lesion can be excised and the expanded skin

advanced, transposed, or rotated, and the process can be accomplished in stages as needed (Fig. 1).<sup>6,44</sup> Giant congenital melanocytic nevi of the abdomen are also amenable to tissue expansion facilitated by an expanded abdominoplasty flap if the patient has sufficient subcutaneous tissue in the abdominal wall (Figs. 2 through 4). Reconstruction of giant congenital melanocytic nevi of the breast region requires careful dissection to prevent injury to the developing breast bud. As such, serial excision and tissue expansion have provided the best modalities for the management of breast lesions. Asymmetry of the breast following reconstruction is optimally treated with a tissue expander (until the breast is fully developed) and subsequently exchanged for a permanent implant.<sup>77</sup>

### Extremities

Giant congenital melanocytic nevi of the extremities can be treated with expanded flaps, grafts, or free tissue transfer.<sup>49</sup> In the proximal aspect of the upper and lower extremities, expanded transposition flaps from the groin and upper back can be used to reconstruct defects created by resection of giant congenital melanocytic nevi. Expanded transposition flaps offer acceptable contour and scar position, less secondary scar contracture, less tension on the distal flap inset, and a larger area of nevus that can be excised. Expanded and delayed "pocket" flaps have been used for lesions excised from the forearm and hand (Fig. 5). Expanded and delayed free transverse rectus abdominis musculocutaneous flaps can



**Fig. 4.** The final outcome for the patient shown in Figures 2 and 3, after multiple stages of tissue expansion.

be used for the distal brachium and distal thigh/proximal lower leg, including the elbow and knee joints.<sup>49,52,53</sup> The distal foot and hand can be resurfaced with expanded full-thickness skin grafts harvested from the abdomen and groin.<sup>77</sup>

### Scalp and Face

Giant congenital melanocytic nevi of the scalp are best treated with tissue expansion. The scalp can be reliably expanded to cover defects measuring half of the hair-bearing scalp without visible thinning



**Fig. 5.** An 18-month-old boy presented with a giant congenital melanocytic nevus with circumferential involvement of the left forearm and hand (*above, left*) to be resurfaced with an expanded lower abdominal flap (*above, right*). Photograph showing circumferential excision of the congenital melanocytic nevus in the forearm and hand to the level of the metacarpophalangeal joints (*center, left*), followed by placement of the extremity in the expanded abdominal flap (*center, right*). Final appearance of the abdominal donor site and the extremity after the involved areas of the index and middle fingers were resurfaced with an expanded full-thickness skin graft (*below*).

or alopecia.<sup>76,77</sup> Some authors feel that calvarial molding that occurs following scalp expander placement remodels over 3 to 4 months,<sup>45</sup> whereas others offer that the molding and ridging following expander removal interfere with expanded flap mobility and therefore manage the calvarial contour deformity with intraoperative burr recontouring.<sup>75,77</sup>

The treatment of giant congenital melanocytic nevi of the face is challenging because of postreconstruction unsightly scarring; as such, the planning and choice of reconstruction requires careful

attention to facial aesthetic subunits.<sup>78,79</sup> Giant congenital melanocytic nevi involving the forehead are best treated with tissue expanders if less than two-thirds of the forehead is involved; more extensive forehead involvement requires distant tissues, and reconstruction of the entire aesthetic subunit with an expanded full-thickness skin graft is an acceptable option.<sup>78</sup> Giant congenital melanocytic nevi of the cheek can be addressed with expanded cervical flaps and/or an expanded postauricular flap if additional tissue is required (Figs.



**Fig. 6.** A 2-year-old boy presented with a giant congenital melanocytic nevus involving the scalp and face.



**Fig. 7.** (Left) Expanders were placed to reconstruct adjacent tissues in the scalp, forehead, and neck. (Right) There was residual nevus following intermediate stages of tissue expansion.



**Fig. 8.** The final outcome of the patient shown in Figures 7 and 8 following completion of tissue expansion.

6 through 8). Expanded full-thickness skin grafts are the best option for the periocular and eyelid regions and are generally taken from the supraclavicular region to ensure proper color match. A giant congenital melanocytic nevus transgressing multiple nasal aesthetic subunits is best addressed with an expanded forehead flap.<sup>78,79</sup>

### CONCLUSIONS

Giant congenital melanocytic nevi provide a difficult diagnostic and reconstructive challenge requiring careful pretreatment evaluation, excisional or nonexcisional treatment, and lifelong patient monitoring and follow-up. With proper treatment, patients and their families can expect a decreased risk of melanoma, reduction of symptoms, improved aesthetics and psychosocial sequelae, and maintenance of function.

The modality selected for the treatment of giant congenital melanocytic nevi should not obscure the surveillance of residual nevus for signs of malignant transformation. Until adequate data can be provided to the contrary, nonexcisional techniques such as chemical peels, curettage, laser, or dermabrasion should be avoided. The latter techniques leave nevus cells behind, particularly in the deeper layers of the dermis, and may obscure future clinical monitoring. In addition, many children and adults who had undergone such treatments have experienced unsatisfactory aesthetic outcomes, such as extensive diffuse granuloma formation, many years after curettage in infancy. The absolute risk of developing cutaneous melanoma in patients with giant congenital melanocytic nevi may be difficult to estimate, because approximately half of the patients select surgical excision for prophylactic or cosmetic reasons. How-

ever, a summation of studies in the published literature reporting the development of malignant melanoma following surgical excision of giant congenital melanocytic nevi estimates a risk of 7.5 percent without surgery and a risk of 0.6 percent with surgery. This suggests that surgical removal of giant congenital melanocytic nevi certainly has a prophylactic effect in the prevention of malignant melanoma.<sup>80</sup>

If surgical resection of giant congenital melanocytic nevi is undertaken, achievement of a histologic deep margin of resection is necessary such that subsequent reconstruction will not mask residual nevus cells. Despite the complex and multistage treatment, tissue expansion has surfaced as the modality that best addresses the principles of treatment and offers the optimization of aesthetic and functional postoperative outcomes. Skin substitutes are not a first-line intervention at the present time but should be considered in the management of giant congenital melanocytic nevi with limited or unusable donor sites. Future research in tissue engineering and gene therapy may offer enhanced outcomes and alternative reconstructive modalities.

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