Congenital Melanocytic Nevi: Treatment Modalities and Management Options

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Congenital melanocytic nevi can be cosmetically disfiguring, give rise to melanoma, and suggest the presence of neurocutaneous melanocytosis. Management decisions must be tailored for each patient and each nevus, taking into consideration the risk for developing malignancy, risk for developing symptomatic neurocutaneous melanocytosis, cosmetic implications of having the nevus, cosmetic implications of any resultant surgical scars from their removal, adverse effects that the nevus may have on psycho-social development, and the adverse effects and long-term sequelae of any surgical intervention. The advantages and disadvantages of different modalities used in the treatment of congenital melanocytic nevi are discussed. Organizational flow diagrams are presented to help clinicians in managing patients with different sized congenital melanocytic nevi.
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Congenital melanocytic nevi (CMN) may be cosmetically disfiguring, can give rise to melanoma, can suggest the presence of neurocutaneous melanocytosis (NCM), and may occasionally be associated with other medical problems such as the tethered cord syndrome.[1,2] It is estimated that each year in the United States between 1% and 6% of infants are born with a CMN, which translates to between 40,000 and 240,000 infants born with a CMN annually.[1] Furthermore, between 3 and 17 million people in the United States have or have had a CMN (4,052,789 live births in the United States in 2000 X approximately 1% of newborns with congenital melanocytic nevi = 40,000 newborns with congenital melanocytic nevi. 284,797,887 people in the United States in 2000 X 1% newborns with a congenital melanocytic nevi = 3 million people with congenital melanocytic nevi [www.census.com]). Hence, physicians are frequently asked to render advice on the management of these nevi. This advice must be tailored for each patient and each CMN, taking into consideration the risk for developing malignancy, risk for developing symptomatic NCM, cosmetic implications of having the CMN, cosmetic implications of any resultant surgical scars from the removal of the CMN, adverse effects that the CMN may have on psycho-social development and the adverse effects and long-term sequelae of any surgical intervention.

CMN are present in a continuum of sizes ranging from small to very large. There are several definitions of what con-

stitutes a small, medium, and large CMN (Table 1).[1-10] Reasons for classifying CMN by size include the risk for developing melanoma, level of surgical complexity involved as well as other complications may be roughly proportional to their size. In this article, we elect to classify CMN as "large" if the greatest diameter of the lesion, in adulthood, is 20 cm or more; "medium" if its greatest diameter measures between 1.5 and 19.9 cm; and "small" if it measures less than 1.5cm. CMN, in general, enlarge in proportion to the growth of the skin covering the involved area of the body.[11] Thus, a 9-cm CMN on the head or a 6-cm CMN on the body of an infant will measure approximately 20 cm in adulthood.[2,12] Patients or guardians should be informed that it is currently not possible to predict which CMN will develop melanoma; however, most will not. Special mention regarding risk of melanoma in large congenital melanocytic nevi (LCMN) is warranted here. Although the risk for developing melanoma within a LCMN is distributed over the lifetime of the patient, it appears that the peak risk occurs before puberty. In contrast, the risk for developing melanoma in smaller CMN is highest after puberty.

Treatment Modalities

The decision for or against treatment of CMN remains controversial. Factors that influence this decision include size of the nevus, its location, its clinical appearance, ease of clinical follow-up, and its malignant potential. Thus, the management needs to be tailored for each patient; however, if active treatments are sought then the treatment modality chosen

Table 1 Size Definitions for Congenital Melanocytic Nevi (CMN)

Size	Definition	References
Small	A CMN less than 1.5 cm in greatest diameter.	Kopf et al[3]
	A CMN that can be completely excised and the defect closed primarily in a single operation.	Lanier et al[4]
Medium	A CMN 1.5 to 19.9 in greatest diameter.	Kopf et al[3]
	A CMN that can be completely excised, but the resulting surgical defect cannot be closed primarily; e.g., flaps, grafts, or tissue expanders are required.	Lanier et al[4]
Large (Giant)	A CMN 20 cm or more in greatest diameter.	Kopf et al[3]
	A CMN involving a major part of an anatomical area such as a face or hand.	Lanier et al[4] Greeley et al[5]
	A CMN that covers greater than 1% of the cutaneous surface surface. (Greater than 0.5% if on the head and neck).	Weinberg et al[6]
	A CMN on the head and neck that is at least the size of a palm. CMN in most other anatomical sites need to be at least twice the size of a palm.	Pers et al[7] Lorentzen et al[8]
	A CMN is 900 cm² or more in area	Greeley et al[5]
	A CMN that covers at least 5% of the body surface area	Swerdlow et al[9]
	A CMN that requires serial or staged excisions for its complete removal	Pilney et al[10]

should ideally address the risk of malignant transformation, achieve satisfactory cosmetic results, and maintain adequate function. Treatment interventions include full-thickness excisions, partial-thickness excisions, dermabrasions, curettage, laser treatment, and chemical peels. Improving the cosmetic appearance frequently requires the use of a combination of different treatment interventions. In terms of preventing the development of melanoma (prophylactic removal), any of the above-mentioned procedures will reduce the overall number of melanocytes, which theoretically should lower the risk of cutaneous melanoma. However, with the exception of full-thickness surgical excision, these procedures do not adequately address the risk for developing melanoma within the deep dermis or subcutis.[13] As with any treatment, the risk and benefits of each treatment modality should be discussed with the patient or guardian. Patients and their parents also need to be informed of the psychological and cosmetic burden frequently placed on patients with large or multiple smaller CMN. This burden may not be eliminated by surgery because the scars from surgery may also be cosmetically disfiguring, although it appears that most patients or their parents prefer the scars to the nevus.[14]

Surgical Excision

Most small CMN can easily be excised and the resulting defect repaired in a relatively simple manner. Larger lesions require individualization, depending on their size, location, and depth. Serial excisions,[15] tissue expanders,[16] and skin

grafts each have a place in the surgical management of LCMN.[17] The development of cultured epithelial autografts and biological agents to help wound healing (eg, Platelet derived growth factor, Keratinocyte growth factor, Epidermal growth factor, and cytokines) may eventually allow the excision of larger areas than would otherwise be possible, resulting in a reduction in the total number of procedures needed to remove very large CMN.[18] As with any treatment, the possible complications involved with surgery need to be considered, such as infection, bleeding, and risk of general anesthesia. One study, evaluating the risk for developing melanoma, risk of general anesthesia, and psychosocial factors, deemed that the best timing for surgical excision is between 6 and 9 months of age, or between 8 and 12 years of age.[19] Based on the age groups at highest risk for melanoma, prophylactic excision of LCMN should be performed early in life, whereas prophylactic excisions of small CMN can be delayed until later years.

Dermabrasion

Dermabrasion, which removes the epidermis and part of the dermis, eliminates the superficial nevus cells. Thus, this procedure may reduce the degree of pigmentation and improve the cosmetic appearance. The remaining deep dermal and subcutaneous nevus cells are eventually covered by scar tissue. The postdermabraded skin is usually thinner, more fragile, tender, and has reduced hair density.[20] In an attempt to remove as many nevus cells as possible, it has been suggested that dermabrasion be performed during infancy. This is based on the fact that dermabrasion becomes more difficult as the epidermal and dermal elements become more adherent with age and on the belief, held by some, that nevus cells migrate down into the deeper layers of the skin over time.[21]

Dermabrasion does not adequately address the issue of melanoma prevention in the deeper tissues. However, some believe that the reduced pigmentation after dermabrasion may allow for easier detection of color changes indicative of melanoma in the deeper layers of the CMN.[20]

Curettage

Treatment of CMN with curettage consists of curetting through a natural cleavage plane that separates the highly nevus populated upper dermis from the relatively less nevus populated deeper dermis.[22] Unfortunately, this cleavage plane is present only during the first few weeks of life, thus limiting the time frame within which this procedure can be performed with reasonably good results and raises the issue of whether the potential cosmetic success of the operation outweighs the risk of anesthesia at this young age.[19] On the other hand, because curettage of larger CMN can often be performed as a single procedure, it may offer a reduction in operative risk compared to multiple serial excisions.[23,24] When performed by experienced operators, curettage can result in acceptable cosmesis. One study showed that for non-scalp CMN the functional and cosmetic results from curettage were superior or equivalent to surgical excision.[22] The postcurettaged dermis is replaced by sclerotic and dense

connective tissue. Repigmentation of the scarred skin and the potential of masking subcutaneous melanoma developing from remnant nevus cells located in the deep dermis or subcutaneous tissue are disadvantages that must be considered.

Chemical Peel

Chemical peels with agents such as phenol have been utilized by some to treat CMN. Deep chemical peels can result in the reduction of the number of melanocytes and may be an option for those lesions that are surgically unresectable and cosmetically disfiguring.[25] CMN most suitable for treatment by chemical peel are those with lighter pigmentation and those with nevus cells confined to the epidermis and superficial dermis[25] (Imaging modalities such as ultrasound, magnetic resonance imaging (MRI), or optical coherence tomography may help in assessing the depth of penetration of nevus cells). Potential side effects of phenol include cardiac and renal toxicity, which need to be considered before embarking on this treatment modality.[26]

Laser

Lasers can be used in the treatment of some CMN with most patients requiring multiple laser treatments before achieving acceptable cosmetic results. Carbon dioxide (CO_2) lasers vaporize tissue resulting in scarring and thus should be considered a surgical procedure akin to dermabrasion.[27,28] Commonly used lasers that do not vaporize tissue include normal mode ruby, Q-switched ruby, Q-switched alexandrite (755 nm) and Q-switched neodymium:yttrium-aluminum-garnet (Nd:YAG) (532 and 1064 nm).[29,30] The Q-switched ruby laser is the most popular laser used to treat CMN. The specificity of the Q-switched ruby laser is due to its 694 nm wavelength, which is selectively absorbed by melanin.[31] In addition, the laser produces a 20-nanosecond pulse duration that approximates the thermal relaxation time for melanosomes, thereby confining the energy to the targeted cells and resulting in the thermal destruction of melanocytes.[32] Q-switched ruby lasers have recently been shown to lighten CMN that because of location, size, or depth of nevomelanocytes were not amenable to surgical excision. This form of treatment is attractive because of its low potential for scarring and its ability to decrease the pigmentation thereby improving the cosmetic outcome. Preliminary histological data show that treatment with Q-switched lasers can achieve significant reduction of papillary dermal melanocytes resulting in reduction of visible pigment.[33] Partial repigmentation, however, does occur in most patients resulting in a final pigment clearance of approximately 50%.[33] The degree of pigment clearance and melanocyte destruction can potentially be enhanced by using a combination of different lasers.[34,35] As with other methods that eliminate only the upper portion of the CMN, the risk of developing melanoma in the deep dermis is not addressed by laser treatments.[36] In addition, lightening of CMN may make it difficult to monitor the resultant lesion for signs of malignant transformation.[13]

Lasers can also be used to help eliminate the hypertrichosis that so commonly occurs on CMN. However, whether lasers are used for hair removal or nevus "removal" one must contemplate the possibility of adverse long-term sequelae. Lasers work by applying heat energy to melanocytes and it is currently unknown whether this heat energy can be potentially mutagenic.

Management Options

Normal changes in CMN can be anticipated. These may include changes in size, color, hair growth, and topography.[26,37,38] Although some alterations are normal, focal changes should be viewed with caution. Focal growths, pigment changes, ulceration, and tenderness are all signs that may suggest malignancy.

All patients with CMN and their parents should be instructed in the technique of self-skin examination, which should be performed on a monthly basis. They need to be educated on the warning signs of melanoma including change in color, size, shape, and symptoms. If a change is noted it should be brought to the attention of their physician. All patients should be instructed to avoid excessive ultraviolet light exposure and to use sun protective clothing and sunscreens. Patients with LCMN, whether excised or not, should be followed for life with complete skin examinations, review of systems, palpation of lymph nodes, and neurological examinations to search for primary or metastatic melanoma. In addition, the nevus and scars, if any, should be palpated for the detection of subcutaneous lumps.

Large Congenital Melanocytic Nevi

Patients with LCMN can develop cutaneous melanoma, noncutaneous melanoma, and NCM. Since malignancies can develop even after surgical removal of the CMN, it would be prudent for these patients to undergo periodic total-cutaneous and general medical examinations, including thorough neurological examinations. Comprehensive neurological examinations may help detect a subset of patients with NCM. Unfortunately, over 80% of patients with symptomatic NCM die because of their disease, 70% within the first decade of life.[39]

Screening brain MRI scans of patients with LCMN may help identify patients with NCM who are neurologically asymptomatic.[40,41] It is not yet known what proportion of asymptomatic patients with central nervous system abnormalities noted on MRI scans will eventually develop symptomatic NCM, but it is probably low.[42] Therefore, further studies are needed to determine the prognostic value and cost-effectiveness of screening MRI scans in this asymptomatic population. In addition to brain MRI scan, patients with large- or medium-sized nevi overlying the lumbosacral area may require an MRI scan of the spine to rule out spinal anomalies such as the tethered cord syndrome.[42-44] Finally, some physicians are investigating the possibility of following serum 5-S-cysteinyldopa levels in patients with LCMN.[45] A sudden rise in the 5-S-cysteinyldopa level may indicate the development of melanoma.

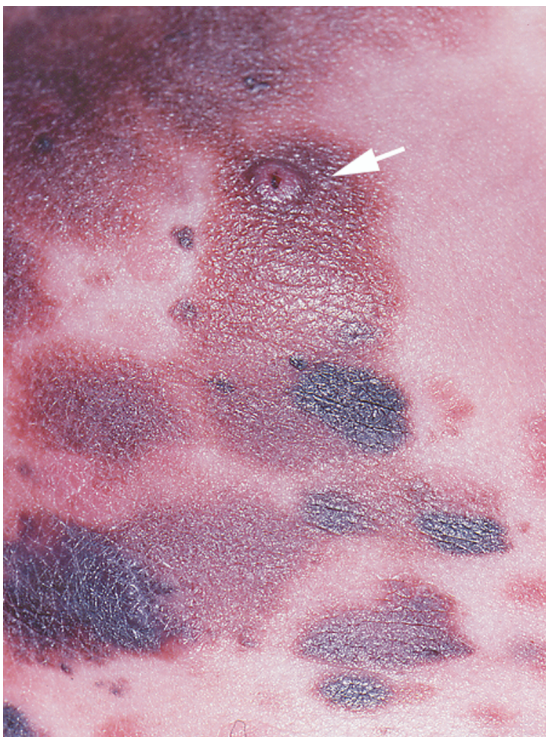Key issues regarding the management of larger-sized CMN
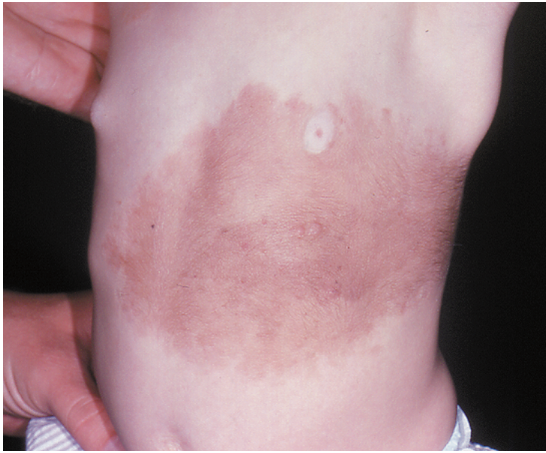
Fig 1 This homogeneous, light brown CMN has no suspicious characteristics and can easily be followed-up clinically.

are presented in Figures 7-11. The salient points of each diagram are discussed below.

Overview

A) Patients with LCMN (Fig 7), especially those with multiple satellite nevi, are at increased risk for developing NCM. Taking a thorough history and performing a neurological examination can help identify patients with symptomatic NCM. Some clinicians recommend obtaining screening MRI scans in asymptomatic patients at high risk for NCM (Fig 11).

However, other physicians disagree with routinely ordering screening MRI scans since currently there are no treatments available to prevent the development of asymptomatic NCM. Those clinicians in favor of obtaining screening MRI scans state that a negative MRI scan can often alleviate patient, parent, and physician anxiety about NCM and a MRI scan that is suggestive of NCM may occasionally alter decisions regarding timing of surgery.

Normal Neurological Examination and LCMN Without any Suspicious Areas

A) LCMN (Fig 8) that are clinically homogeneous and without irregularities (Fig 1) can be followed clinically or can be excised.

B) Clinical follow-up is performed with the aim of detecting changes developing within the LCMN that may herald melanoma. The presence of baseline clinical photographs of the skin can greatly assist the physician in following these lesions and aid in identifying subtle changes.[46] Changes that are suggestive of malignancy should be biopsied (Fig 9).

C) Cosmetic cover-up foundation/make-up such as Dermablend (Dermablend Corrective Cosmetics, New Jersey) can be used to help conceal exposed nevus and/or scars. These

Fig 2 The sudden appearance of a papule within this LCMN raised concern of melanoma and thus was biopsied. The biopsy revealed a benign proliferative nodule.

Fig 3 The clinical examination of thickened and rugous CMN can often be difficult. Prophylactic excision of these nevi should be contemplated.
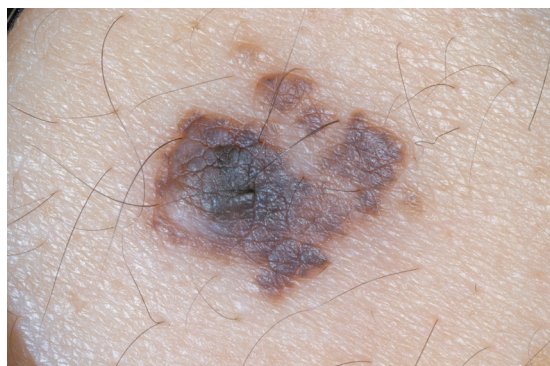
Fig 4 Patients with LCMN and multiple satellite nevi are at high risk for developing NCM.

nevus may be important issues for the patient and should not be overlooked.

E) Other treatment methods such as laser, dermabrasion, curettage, and partial excisions may be used in an attempt to improve the cosmetic appearance. The limitations of these procedures were discussed previously.

F) If prophylactic removal of a LCMN is considered then the planned excision should attempt, if feasible, to remove all of the involved tissue down to the underlying fascia. Consideration for cosmetic outcome and function needs to be taken into account when planning surgical excisions. Parent and patient expectations need to be addressed and they should be informed that complete removal of every nevomelanocyte is frequently impossible. Thus, patients and parents should be told that the excision is done in an attempt to lower the risk for developing cutaneous melanoma, but that it does not eliminate the risk. Melanoma can still develop de novo or within the residual nevus cells or from nevus cells in the central nervous system.[47]

G) Prophylactic excision of a LCMN should be undertaken between 6 and 9 months of age because the risk of melanoma is greatest in infancy and the risk of general anesthesia is greatest for patients under six months of age.[19] However, if the lesion is suspicious or develops features consistent with malignant degeneration then excision and biopsy should not be delayed.

H) Photographs of the nevus, satellite nevi, or residual nevus after partial excision can aid in the clinical follow-up examinations. The cutaneous examinations should include

products have an added benefit in that most also contain a sunscreen.

D) Surgical excision can be performed with the intent to improve cosmesis and/or to prevent the development of melanoma. The decision to perform surgery is often difficult and needs to weigh the risks and benefits of multiple issues such as degree of anticipated cosmetic improvement, extent to which it will lower the risk for developing melanoma, risks involved with anesthesia and surgery, degree of scarring, degree of functional impairment and degree of discomfort involved with the procedures. The cosmetics and improved self-image gained by removing the exposed portion of the

Fig 5 Prophylactic excision of irregular small CMN, such as the one depicted here, can easily be performed by simple elliptical excision.

Fig 6 Homogeneous and symmetric small to medium CMN are easy to follow clinically. Baseline photographs are useful for this purpose.

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                          │    LCMN     │
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              ┌──────────────────┴──────────────────┐
      ┌───────────────────┐              ┌───────────────────┐
      │ Neurological exam  │              │ Neurological exam  │
      │ normal (consider   │              │ suggestive of NCM  │
      │ screening MRI-Fig 11)│            └─────────┬─────────┘
      └─────────┬─────────┘                         │
       ┌────────┴────────┐                  ┌────────────────┐
 ┌──────────────┐ ┌──────────────┐          │     Fig 10     │
 │ LCMN without │ │ LCMN with    │          └────────────────┘
 │ any suspicious│ │ "atypical"   │
 │ areas        │ │ areas        │
 └──────┬───────┘ └──────┬───────┘
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   │  Fig 8  │      │  Fig 9  │
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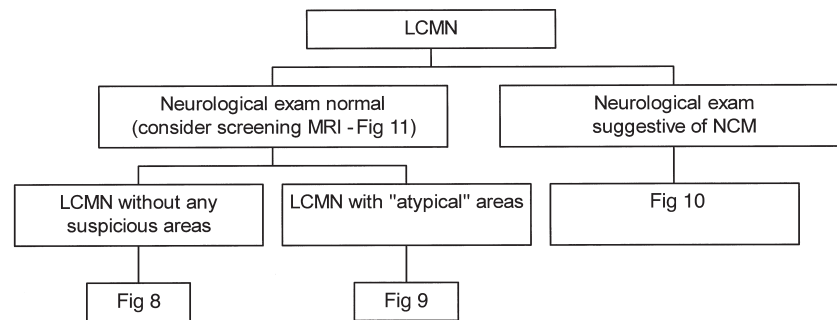
Fig 7 The overview for managing patients with LCMN (see text overview).

visual inspection of the skin and palpation of the nevus or scars overlying areas from where portions of the CMN were excised. Palpation allows the examiner to detect subcutaneous nodules that may be indicative of melanoma developing below the dermo-epidermal junction.

Normal Neurological Examination & LCMN With Atypical Areas

A) Clinically atypical areas within a LCMN (Fig 9) need to be biopsied (Fig 2).

B) If the biopsy is benign and the rest of the LCMN is easy to follow clinically, see Fig 8.

C) Prophylactic excision should be contemplated for LCMN that are atypical, nodular, or thickened (Fig 3). These lesions are difficult to follow clinically even in the presence of baseline photographs.

D) If prophylactic complete excision is not possible then attempts should be made to excise the most infiltrated, thickened or multinodular component of the nevus. The reason for this is that melanoma can be difficult to detect in these thickened areas.

E) Once the LCMN has been excised (partial or complete), the patient should be followed-up and the scars and remnant nevus areas should be inspected and palpated.

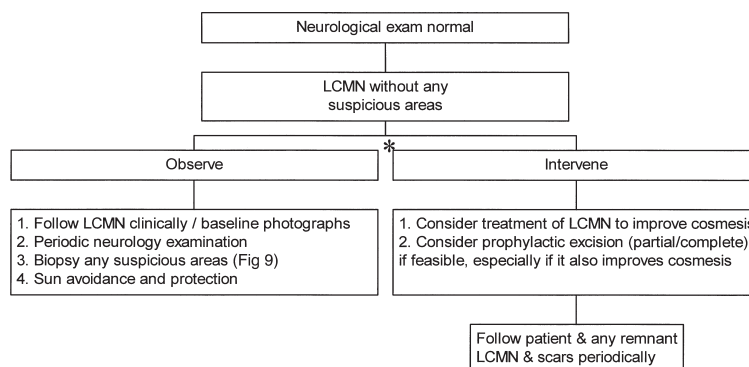F) A word of caution is warranted here. Biopsy of lesions within LCMN, particularly those found during early infancy, may resemble melanoma histologically but they behave in a clinically benign fashion.[48-50] The clinical and histological differentiation between true melanoma and its simulants can at times be difficult (Fig 2). Thus, if a diagnosis of melanoma is made it would be prudent to obtain a second opinion from a dermatopathologist experienced in the evaluation of pigmented lesions.[51]

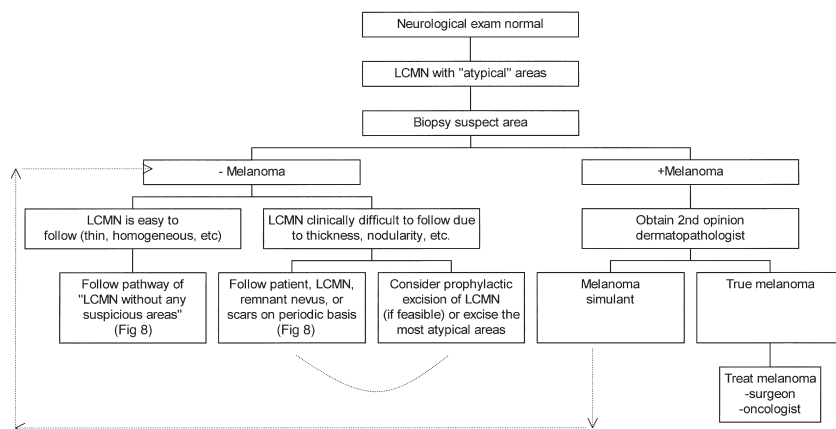## Neurological Examination Suggestive of NCM

A) Patients with symptomatic NCM (Fig 10) should be evaluated to determine whether their symptoms can be alleviated. Neurosurgery procedures such as placement of a ventriculo-peritoneal shunt may significantly improve the functional status of some patients. However, the ventriculco-peritoneal shunt may also provide a conduit for leptomeningeal melanocyte migration from the leptomeningies to the peritoneal cavity.[52]

B) Patients with symptomatic NCM whose symptoms are progressive tend to have a poor prognosis and thus should be spared aggressive prophylactic cutaneous surgery, at least until their status improves.

C) Patients with symptomatic NCM whose symptoms are improving may ultimately have a relatively good prognosis and thus can be managed as outlined in Figs 8 and 9.

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                    ┌──────────────────────────┐
                    │ Neurological exam normal  │
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                    ┌──────────────────────────┐
                    │ LCMN without any          │
                    │ suspicious areas          │
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      ┌──────────────┐                          ┌──────────────┐
      │   Observe    │                          │  Intervene   │
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 ┌───────────────────────────┐        ┌───────────────────────────────┐
 │1. Follow LCMN clinically / │        │1. Consider treatment of LCMN   │
 │   baseline photographs     │        │   to improve cosmesis          │
 │2. Periodic neurology exam  │        │2. Consider prophylactic        │
 │3. Biopsy any suspicious    │        │   excision (partial/complete), │
 │   areas (Fig 9)            │        │   if feasible, especially if it│
 │4. Sun avoidance and        │        │   also improves cosmesis       │
 │   protection               │        └───────────────┬───────────────┘
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                                              │ Follow patient & any │
                                              │ remnant LCMN & scars │
                                              │ periodically         │
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Fig 8 Flow diagram that may help in management decisions for patients with nonsuspicious LCMN and who have a normal neurological examination (see text). *Factors influencing therapeutic intervention include size, location patient anxiety, aesthetic and functional tradeoffs, etc (see text).

Fig 9 Flow diagram that may aid in the management decisions for patients with LCMN that have a normal neurological examination but who have clinically atypical areas within their LCMN (see text).

Screening MRI:

A) MRI scans (Fig 11) can be used as a screening tool to detect asymptomatic NCM in high risk individuals such as those with LCMN and many satellite nevi (Fig 4). The timing of when a screening MRI is performed may be very important. It appears that myelin protein can obscure subtle deposits of melanocytes.[42,53] Thus, it may be best to obtain a screening MRI in early infancy (birth to 4 months of age) before the brain has myelinized.

B) Individuals with a normal MRI scan can be managed as outlined in Figs 8 and 9.

C) Asymptomatic individuals with an MRI scan suggestive of NCM should be followed closely and elective surgeries delayed until the status of the patient is known. Preliminary data suggest that only a small proportion of these individuals will actually develop symptomatic disease.[42] Those with a good prognosis (ie, stable or improving MRI findings) can be managed as outlined in Figs 8 and 9. Those with a poor prognosis (ie, progressive changes on MRI scans or development of symptomatic NCM) should be managed as described in Figure 10.
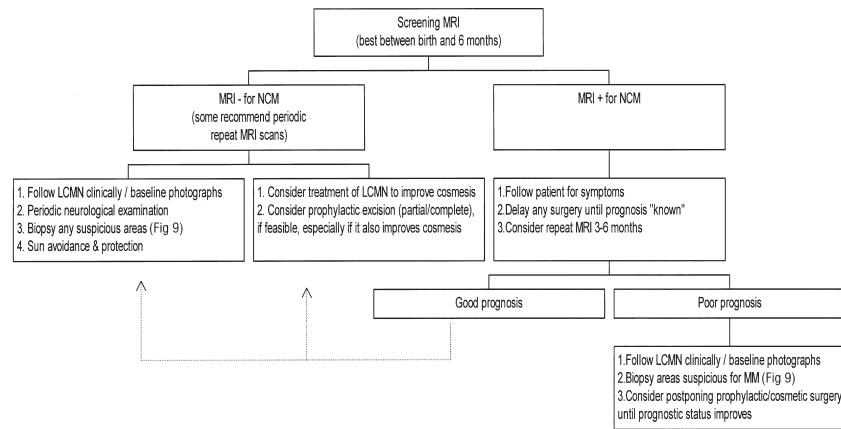
Small and Medium Congenital Melanocytic Nevi

An algorithm delineating an approach to the management of patients with small to medium CMN is presented in Fig 12. The management of small and medium-sized CMN remains controversial.[54] The lifetime risk for developing melanoma in smaller CMN is estimated to be between 0% and 5% (Table 2). Since it is rare for smaller CMN to undergo malignant transformation during childhood, many physicians agree that these lesions generally need not be considered for excision until later in life.[54] However, CMN under 20 cm in diameter that may warrant excision at an earlier age include relatively large medium-sized lesions and those with unusual morphologic features (Fig 5), such as thickened lobular or cerebriform appearance, that would otherwise compromise the ability to clinically follow the lesion.[54,55]

Homogeneous and symmetric smaller CMN (Fig 6) can be followed during childhood with routine physician follow-up, self-skin examinations, baseline photographs, and dermoscopy.[56] If a change is noted on any of the aforementioned examinations, the lesion should be evaluated for biopsy. The
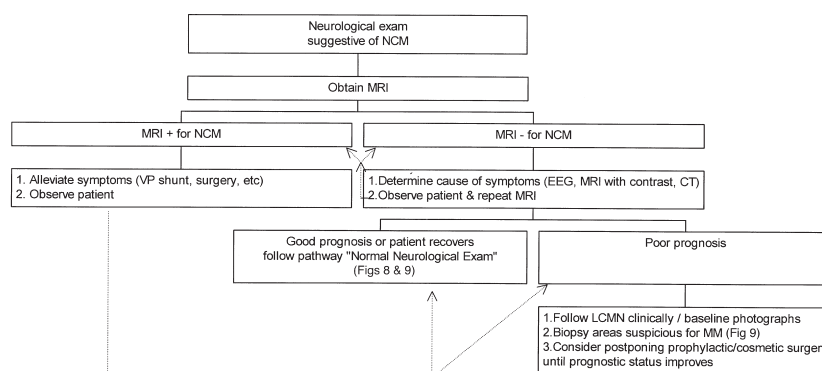
Fig 10 Flow diagram that may be used to guide the management of patients with LCMN that are suspected to have NCM (see text).

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                          ┌─────────────────────────┐
                          │       Screening MRI      │
                          │ (best between birth and  │
                          │        6 months)         │
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                   ┌───────────────┴─────────────────┐
         ┌──────────────────┐              ┌──────────────────┐
         │   MRI - for NCM   │              │   MRI + for NCM   │
         │ (some recommend   │              └──────────────────┘
         │ periodic repeat   │
         │    MRI scans)     │
         └──────────────────┘
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Fig 11 diagram boxes:

MRI - for NCM box branches to:
- 1. Follow LCMN clinically / baseline photographs
  2. Periodic neurological examination
  3. Biopsy any suspicious areas (Fig 9)
  4. Sun avoidance & protection
- 1. Consider treatment of LCMN to improve cosmesis
  2. Consider prophylactic excision (partial/complete), if feasible, especially if it also improves cosmesis

MRI + for NCM box:
1. Follow patient for symptoms
2. Delay any surgery until prognosis "known"
3. Consider repeat MRI 3-6 months

Branches to: Good prognosis | Poor prognosis

Poor prognosis box:
1. Follow LCMN clinically / baseline photographs
2. Biopsy areas suspicious for MM (Fig 9)
3. Consider postponing prophylactic/cosmetic surgery until prognostic status improves

Fig 11 Flow diagram for managing MRI results (see text).

decision to proceed with elective "prophylactic" excision can be delayed until the child can actively participate in the decision and is old enough to tolerate the procedure under local anesthesia. Often this is around eight years of age.

Small/Medium CMN

A) The risk for developing melanoma (Fig 12) appears to increase at puberty. Therefore, small to medium-sized nevi that are not clinically atypical or symptomatic can be clinically followed during childhood.

B) Nevi that are uniform, light colored, even textured, and without nodules can be photographed and followed periodically for change. Any nevus that has changed, becomes symptomatic, or develops a new growth (especially on its periphery) should be considered for excision. Nevi that remain stable need not be excised.

C) Nevi that are dark, multinodular, or clinically atypical should be considered for excision and biopsy.

D) Excision should be considered if examination of the lesion and the follow-up is burdensome to the patient or family. Prophylactic removal should also be considered if the patient or the parents' anxiety level is very high regarding the follow-up of these nevi or their potential risk for developing melanoma.

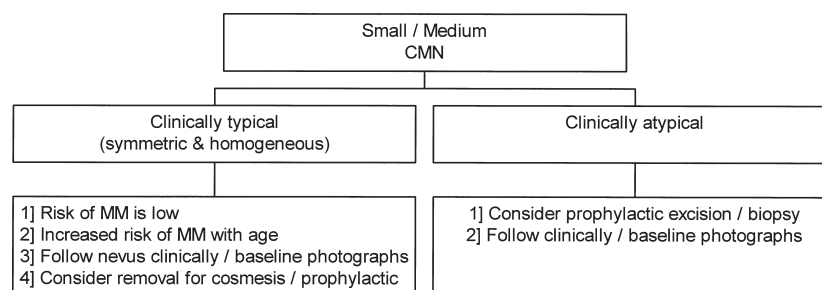E) If the nevus is not excised then serial photographs and periodic follow-up, especially after the age of 12, is recommended.

Conclusion

Although the precise risks for melanoma in patients with CMN are not known, including the added risk resulting from sun exposure, all patients and/or family members should be instructed on sun avoidance and sun protection. Routine examinations, aided by photographic documentation, may enable early diagnosis and treatment should melanoma supervene.[57]

The parents and (if appropriate) the patient should be informed of the controversy concerning the risk of developing melanoma and should be briefed on the anticipated cosmetic results from surgery since many have unrealistic expectations that "plastic surgery leaves no scars." All potential treatment modalities including surgery, dermabrasion, curettage, and lasers should be discussed and their associated risks and benefits outlined. The final decision on whether to follow-up clinically or to treat the CMN rests on the patient and family.

At times, psychological consultation is useful in gaining a better understanding of the impact that a CMN may have on psychosocial development. The patient and/or family might wish to contact a CMN support group such as Nevus Out-

Fig 12 diagram:

Small / Medium CMN branches to:
- Clinically typical (symmetric & homogeneous):
  1] Risk of MM is low
  2] Increased risk of MM with age
  3] Follow nevus clinically / baseline photographs
  4] Consider removal for cosmesis / prophylactic
- Clinically atypical:
  1] Consider prophylactic excision / biopsy
  2] Follow clinically / baseline photographs

Fig 12 Flow diagram that may be used to help in the management decision for patients with CMN < 20 cm in size (see text).

Table 2 Frequently Asked Questions Regarding CMN

CMN Size	Small & Medium	Large
1. What is the reported lifetime risk for developing melanoma?	0-4.9%	4.5-10%
2. Where do melanomas develop?	Within the CMN CNS (if ≥3 CMN present)	Within the CMN CNS Other (ie, retroperitoneal)
3. If melanoma develops within the CMN, where does it start?	Dermo-epidermal junction Peripheral edge of the CMN	Anywhere within the CMN including below the dermo-epidermal junction (ie, dermis)
4. What is the ease of diagnosing cutaneous melanoma?	Since melanomas tend to develop at the dermo-epidermal junction, they can be easily recognized clinically and diagnosed early	Since many melanomas develop deep within the CMN, the clinical recognition of early melanoma is difficult
5. When do most melanomas develop?	At or after puberty	Before puberty
6. Who is at increased risk for NCM?	Patients with ≥3 CMN	CMN located in axial locations (ie, head & neck, midline back) Patients with multiple satellite CMN
7. What are the management options?	Clinical follow-up Monthly self skin examination Cosmetic improvement (make-up, lasers, excision) Prophylactic removal	Clinical follow-up Monthly self skin examination Cosmetic improvement (make-up, lasers, excision, dermabrasion) Prophylactic removal
8. If prophylactic removal is decided upon, when should it be done and what methods are available?	Removal can be planned at anytime up to puberty Excision (simple or serial, with or without skin grafts or tissue expanders)	Treatment should be rendered early in life Excision (simple or serial, with or without skin grafts or tissue expanders) ?Curettage in neonatal period?
9. Aids to the detection of early melanoma? (if prophylactic excision is not an option or if portions of the nevus are not excised)	Clinical inspection Dermoscopy Baseline photographs used during follow-up examinations to help detect subtle changes Confocal laser microscopy	Clinical inspection Palpation Baseline photographs to help detect subtle changes ?Dermoscopy? ?Confocal laser microscopy?
10. What complications other than melanoma can develop?	Cosmetic issues due to CMN or surgical scars Psychosocial	Cosmetic issues due to CMN or surgical scars Psychosocial Other malignancies (ie, rhabdomyosarcoma) Symptoms (ie, pruritus, tenderness, skin fragility)
11. What lab tests or consultations should be considered?	Dermatologist Plastic surgeon Psychologist	Dermatologist Plastic surgeon Pediatric neurologist Psychologist MRI of brain (radiologist)

NOTE. All patients should be instructed on the importance of sun avoidance and sun protection.

reach (ww.nevus.org) or Nevus Network (www.nevusnetwork.org).

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