IN BRIEF

Melanocytic Nevi in Children: Clinical Features and When to Worry

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INTRODUCTION

Melanocytic nevi, or as patients may more commonly refer to them, "moles," are a nearly ubiquitous pediatric condition. To avoid unnecessary procedures or worry, pediatricians should be aware of the clinical appearance, natural history, and common types of melanocytic nevi in this population, as well as truly concerning features of a pigmented nevus that should prompt referral to a dermatologist. This In Brief reviews a few of the most commonly seen melanocytic nevi and clinical features that should prompt referral.

MELANOCYTIC NEVI: WHAT IS NORMAL? (FIG 1)

Melanocytes are cells in the basal layer of the skin that produce melanin, the pigment responsible for an individual's skin tone. Acquired melanocytic nevi are benign proliferations of nevus cells (nests of melanocytes) that develop after birth. These lesions tend to arise in early childhood and increase in number as an individual ages, particularly around puberty (likely due to hormonal influence). They are usually round or oval, well demarcated, and uniformly pigmented. Light brown to black in color, they can range from flat to elevated dome-shaped papules, depending on the type of nevus.

Melanocytic nevi are generally classified as junctional, compound, or intradermal based on where in the skin the nest of melanocytic cells is found. As a person ages, a single nevus can (although does not always) undergo progression through these stages as the nevocytes migrate, first appearing as a flat macule (junctional nevus, melanocyte nests are in the epidermis) and gradually progressing into an elevated papule once adulthood is reached (compound nevus, nevocytes in the dermoepidermal junction, or an intradermal nevi, a "fleshier" nevi that contains these cells strictly in the dermis). This change is typically a normal and benign part of the life cycle of an acquired melanocytic nevus.

Genetics, race, and environmental factors all play a role in the number and distribution pattern of an individual. Sun exposure has also been linked with an increased number of melanocytic nevi, especially exposure, which is both significant and intermittent (eg, the child who vacations in Florida each year and receives a significant amount of sun exposure during that time).

AUTHOR DISCLOSURE: Drs Muser and Tamburro have disclosed no financial relationships relevant to this article. This commentary does not contain a discussion of an unapproved/investigative use of a commercial product/device.

Congenital Melanocytic Nevi. Moustafa D, Blundell AR, Hawryluk EB. *Curr Opin Pediatr.* 2020;32(4):491–497

No Biopsy Needed for Eclipse and Cockade Nevi Found on the Scalps of Children. Kessides MC, Puttgen KB, Cohen BA. *Arch Dermatol.* 2009;145(11):1334–1336

Pediatric Melanoma. Tracy ET, Aldrink JH. *Semin Pediatr Surg.* 2016;25(5):290–298

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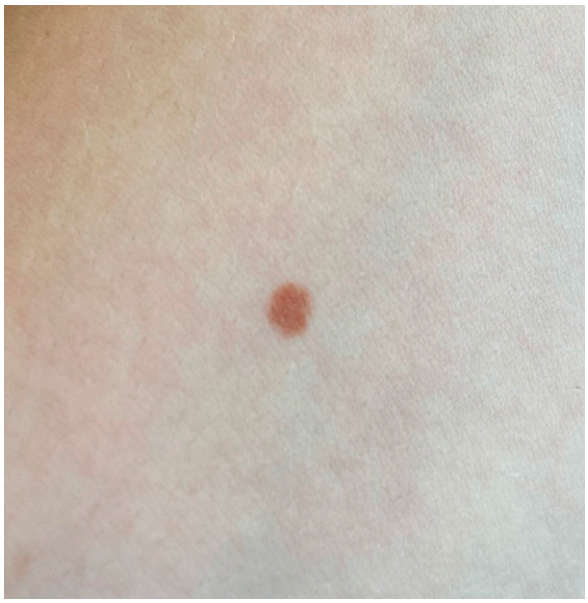

Figure 1. Flat, evenly colored, normal melanocytic nevus.

HALO NEVI (FIG 2)

A halo nevus is a relatively common lesion characterized by a central, pigmented macule or papule that develops a surrounding ring (or "halo") of pale depigmentation. This depigmented ring may cause some concern because apparent pigmentation change may indicate pathology, as seen in adult nevi. Typically, halo nevi are less than 1 cm in diameter, well defined, with a symmetrical and regular halo. Many of these nevi regress entirely, leaving a depigmented macule in their place; this is a normal course and is not cause for concern.

SPITZ NEVI (FIG 3)

Spitz nevi are also benign, acquired nevi, although their clinical features can sometimes make them difficult to distinguish from a melanoma. In fact, they were first characterized as "juvenile melanoma" by Sophie Spitz in 1948. These nevi are relatively rare and tend to occur in patients younger than 20 years.

Spitz nevi most often present as a rapidly growing pink or red papule on the head/neck and lower extremities. Because there are exceptions to every rule, these nevi can be amelanotic (do not contain melanin, therefore without pigment) or melanotic (contain melanin, therefore pigmented) and can affect any area of the skin. Given their significant clinical overlap with pediatric melanomas, typically, patients with this type of nevus should be referred to a dermatologist for continued monitoring. In children, this wait-and-see approach (consisting of periodic clinical and dermoscopic follow-up approximately yearly) is perfectly acceptable. Referral for surgical excision is reserved for suspicious nevi in children older than 12 years, and at any age if the lesion is showing concerning signs suggestive of malignant melanoma, such as large size (especially >10 mm), asymmetry, or ulceration.

ECLIPSE NEVI (FIG 4)

Another common nevus of which to be aware is an eclipse or "fried egg" nevus. This lesion is one of the most common scalp nevi in pediatric patients, although it may also be found on the trunk. When found on the scalp, it may be a marker of the child's propensity to develop an above average number of nevi later in life. It is characterized by a tan center (which may be elevated) and a brown (often ill-defined) rim, resembling a fried egg. It is worth noting that this can arise from an evenly colored, flat macule, which later progresses to the fried egg appearance. The dual tone and irregular border of this lesion may cause concern, although this is a benign lesion and does not require a biopsy or referral to a dermatologist for this finding alone.

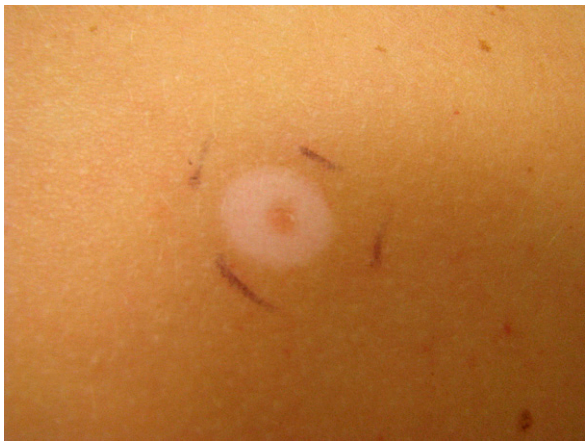

Figure 2. Halo nevus.

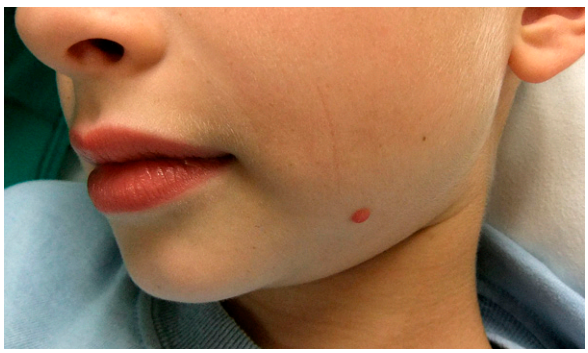

Figure 3. Typical Spitz nevus with a pink papular appearance on the face.

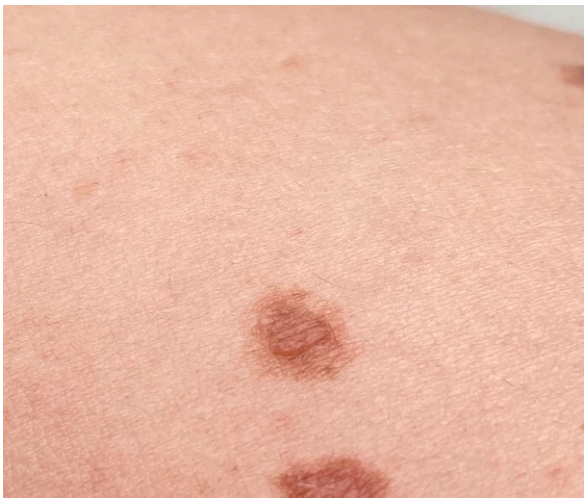

Figure 4. Eclipse nevi.

CONGENITAL MELANOCYTIC NEVI (FIG 5)

Congenital melanocytic nevi (CMNs) are proliferations of melanocytes that develop in utero and, therefore, present at birth or within the first few weeks of life. An estimated 1% of all newborns are born with a CMN, and they are classified based on projected adult size because these lesions grow proportionally with the child. A small CMN is projected to be smaller than 1.5 cm at its adult size; medium-sized, 1.5 to 19.9 cm; large, 20 cm or greater; and giant, greater than 40 cm.

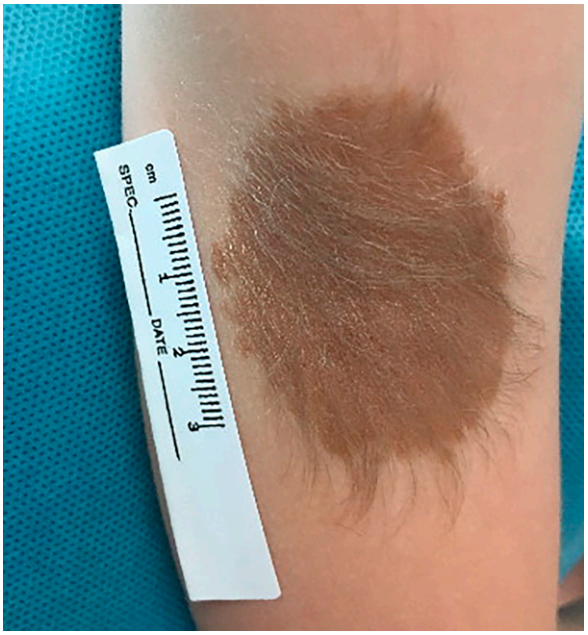

Figure 5. Hypertrichotic congenital melanocytic nevus with excessive hair growth.

Note that CMNs (especially large or giant CMNs) may be associated with extradermal complications, including central nervous system disorders, facial deformities, and endocrine abnormalities. CMNs can undergo many diverse, benign changes in clinical appearance that a pediatrician may observe as the patient grows, including pigmentary changes such as loss of pigment or speckling, textural changes such as becoming more verrucous or hypertrichotic (containing excessive hair growth), and the development of discrete nodules in the lesion termed *proliferative nodules*.

Although most of these proliferative nodules are benign, approximately 2% to 10% of large or giant CMNs undergo malignant transformation, and lesions on the axial body, scalp, and trunk carry an increased risk of malignant transformation compared with those on the limbs. Malignant transformation carries significant mortality: approximately 30% to 60% of these cases prove fatal. Pediatricians should be alert for new, deep subcutaneous or dermal nodules that rapidly grow within the borders of the CMN, especially with ulceration or bleeding, and should make appropriate referrals if this occurs.

PEDIATRIC MELANOMAS (FIG 6)

Malignant melanomas are considerably rarer in the pediatric population than in adult populations (with approximately 500 diagnoses annually), although the incidence continues to rise. Melanomas account for 8% of adolescent cancers and remain the most common skin cancer among patients younger than 20 years. Furthermore, pediatric melanomas (birth to early adulthood) have distinct clinical and pathological characteristics compared with melanomas in older populations and may go underrecognized due to the unique clinical patterns of presentation.

Pediatricians may be aware of the so-called ABCDE rule, a mnemonic used to aid in the diagnosis of

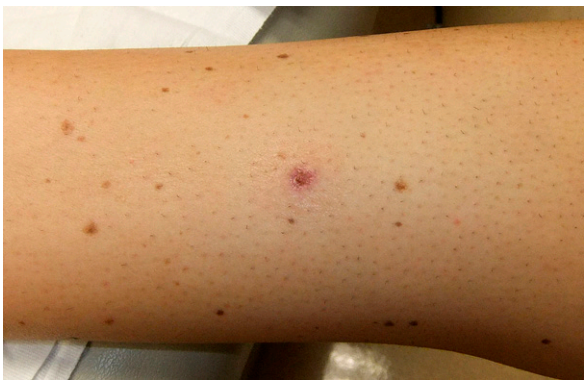

Figure 6. Biopsy-confirmed melanoma. Note the ulceration/bleeding present.

	"ABCDE rule"	Proposed ABCD Rule of Pediatric Nevi
A	Asymmetry	Amelanotic
B	Border Irregularity	Bleeding/bump
C	Color (ie, pigment is not uniform)	Color uniformity (ie, pigment is not uniform)
D	Diameter (>6mm)	De novo/any diameter
E	Evolving (ie, the lesion is changing size, shape, color, etc)	

Figure 7. ABCDE rule and proposed ABCD rule of pediatric nevi.

melanomas by paying careful attention to *A*symmetry, *B*order irregularity, *C*olor, *D*iameter (>6 mm), and *E*volution. Many parents and practitioners alike may be aware of this rule thanks to public health campaigns and may naturally apply them to monitoring their pediatric patients with nevi. Although this may be helpful in some cases, the classic ABCD hallmarks of diagnosis are not seen in 40% to 60% of pediatric melanomas. In fact, pediatric melanomas are frequently amelanotic, and most arise de novo rather than from a melanocytic nevi.

Pediatricians should instead be on high alert for a new, more uniformly colored, symmetrical, raised, and amelanotic lesion, particularly those presenting with bleeding. Because of these differences, an ABCD rule of pediatric moles that focuses on these aberrant findings has been proposed in the literature and remains to be verified with further research (Fig 7).

To identify the other nearly one-third of pediatric melanomas that arise from a precursor lesion, it is helpful to be aware of the most common presentations based on patient age. In infancy and the neonatal period, congenital melanocytic nevi and de novo melanoma predominate. De novo melanoma refers to a rare condition in which cells from a metastatic malignant melanoma in the mother are transmitted transplacentally to the fetus. All children born to mothers with a current diagnosis of metastatic melanoma should be examined by a pediatric dermatologist as soon as possible after birth. Childhood melanomas (prepubertal) tend to arise de novo, and once past puberty, patients are more likely to develop a melanoma from a dysplastic nevus (much like adults). Signs and symptoms of a malignant transformation in these lesions (as described previously herein) should prompt quick referral to a dermatologist for biopsy and excision if appropriate.

CONCLUSION

Knowledge of the clinical appearance, natural history, and truly concerning features of a pigmented nevus can help pediatricians identify which nevi require increased monitoring and specialist referral.

COMMENTS:

Melanocytic nevi are an entity that troubles pediatricians. We as clinicians want to balance the desire to not miss serious lesions and diagnose skin cancers and melanomas that are rare in childhood and yet not over-refer to dermatologists, which leads to increased cost and potential anxiety for the patient and family. This In Brief helps address some of this diagnostic uncertainty.

I have found as a pediatrician the benefit of having a relationship with a dermatologist or team of dermatologists to discuss patients with, and also the utility of sending photographs through the electronic record to gain their assistance to assess whether a lesion meets the criteria for referral and a closer inspection. This In Brief also reminds us of the importance of pediatric prevention strategies for good skin health regarding sun exposure and implementation of these strategies early in the life cycle: encouraging the use of sunscreen (at least SPF of 15 and reapplying every 2 hours and after swimming or sweating), wide-brimmed hats, and sunglasses; limiting sun exposure between 10 AM and 4 PM when possible; and covering the skin with clothing during peak sun exposure times.

–Janet Serwint, MD
Associate Editor, In Brief

ANSWER KEY FOR SEPTEMBER PEDIATRICS IN REVIEW

Early Puberty: 1. D; 2. C; 3. B; 4. B; 5. B.
General Hepatitis: 1. D; 2. B; 3.C; 4. B; 5. B.
Port-wine Birthmarks: Update on Diagnosis, Risk Assessment for Sturge-Weber Syndrome, and Management: 1. C; 2. D; 3. B; 4. A; 5. D.