

Myths and Misconceptions

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The Risk of Melanoma in Small Congenital Nevi

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A 2-year-old boy presents with a 4 × 3-cm, darkly pigmented nevus on his back. His mother states that this lesion has been present since birth. Both parents noticed that the nevus has become slightly larger but deny any other changes in its appearance. The boy is otherwise healthy. He has no allergies and does not take any medication. There is no family history of skin disease or cutaneous malignancy. The parents are concerned about the melanoma risk in their child's congenital nevus.

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Approximately 1% of all infants have congenital melanocytic nevi (CMN). CMN are pigmented lesions present at birth or arising shortly thereafter.¹ CMN are classified according to their largest estimated diameter in adulthood: small (<1.5 cm), intermediate (1.5–19.9 cm), and large (≥20 cm).² Large CMN comprise ≥5% of the body surface area in infants, children, and preadolescents.³ Classic microscopic description of CMN includes nevus cells in the lower two thirds of the dermis, occasionally extending into the subcutis; between collagen bundles distributed as single cells or cells in single file or both; and the lower two thirds of the reticular dermis or subcutis associated with appendages, nerves, and vessels.⁴ Large CMN usually have these classic microscopic findings of CMN, intermediate CMN may or may not show these features, and small CMN most often do not show these microscopic features.⁵ Also, there is significant overlap in histologic features between congenital and acquired nevi.²

The risk of developing a melanoma in small CMN is very controversial. Some of this uncertainty stems from difficulty with the histologic and clinical definition of these lesions, and also from lack of good long-term follow-up studies. An early study by Illig et al.⁶ suggested an increased risk of developing a melanoma in CMN <10 cm in diameter. All of the melanomas detected in this study arose superficially in the skin and developed after puberty. Two recent studies^{7,8} failed to detect an increased

risk of developing a melanoma in small CMN. The most recent study of 2,198,619 newborns with 3922 CMN of varied sizes failed to reveal an increased risk of developing a melanoma in CMN of any diameter and, therefore, does not advise prophylactic excision of any CMN.⁹ It has been postulated that, in general, prophylactic removal of small CMN is not essential because almost all melanomas arising in small CMN are of the epidermal variety and can be followed-up by clinical observation.⁵ If excision is to be performed, it can be safely done after puberty.

Some researchers recommend individualized follow-up of small CMN according to the lesion appearance, the location of the lesion, a patient's racial background, and strong family history of melanoma.² They postulate that if the lesion is uniform, light to medium in color, of smooth texture, and absent nodules, then such a lesion can be followed-up with clinical observation. If the lesion is more deeply pigmented or unevenly textured, then such lesion should be excised. Unfortunately, the clinical appearance characteristics that are associated with an increased risk of developing melanoma are conjecture, and there are no clear data supporting these recommendations. It is also suggested that a strong family history of melanoma may warrant an excision of small CMN, although there are no data relating the risk of melanoma in CMN to positive family history of developing melanoma in an acquired nevus.² Excision of these nevi in

nonmelanoma-prone populations (e.g., Asians and African-Americans) is not warranted.¹⁰

The risk of developing melanoma in small CMN is controversial. The management of these nevi is based on appearance, loca-

tion, histologic features, racial background, and strong family history of melanoma. A recent study failed to detect an increased risk of developing a melanoma in CMN of any diameter and therefore does not advise prophylactic excision of any CMN.⁹

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