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Narrative Review

The Three Most Important Things to Tell Parents of a Newborn/Infant with a Congenital Melanocytic Nevus

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KEYWORDS

Melanocitic nevus,
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ABSTRACT

Congenital melanocytic nevi (CMN) are benign lesions which are typically classified according to their size. The significant attention given to CMN is justified by both aesthetic concerns and the risk of cutaneous melanoma. The author reports the most up-to-date estimates regarding the risk of melanoma and indicates the three most important things to communicate to the parents of a child with a congenital melanocytic nevus.

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Introduction

Melanocytic nevi (MN) are benign lesions composed of melanocytes, the cells responsible for producing melanin pigment. These melanocytes are typically found in clusters (thecae) arranged along the dermal-epidermal junction, in the demis or in both.

Congenital melanocytic nevi (CMN) develop *in utero* and are usually visible at birth. However, even those that appear in the first months of life (tardive nevi) are considered congenital, especially if their diameter exceeds 1.5 cm.

The significant attention given to CMN, even in childhood, is justified by both aesthetic concerns and the potential for them to evolve into melanoma.

Congenital melanocytic nevi are typically classified according to their projected adult size (1).

1) Giants: maximum diameter greater than 40 cm;
2) Large: maximum diameter between 20 and 40 cm;
3) Medium: maximum diameter between 1.5 and 20 cm;
4) Small: maximum diameter less than 1.5 cm.

The scaling factor used to predict adult size is determined by the anatomical location of the nevus. A CMN located on the head is predicted to grow by a factor of 1.7, on the lower limb by 3.3, and upper limb and torso by 2.8 (2).

Classification by size is justified by the significant differences in clinical, aesthetic, and therapeutic terms, as well as the varying risk of melanoma (Fig. 1).

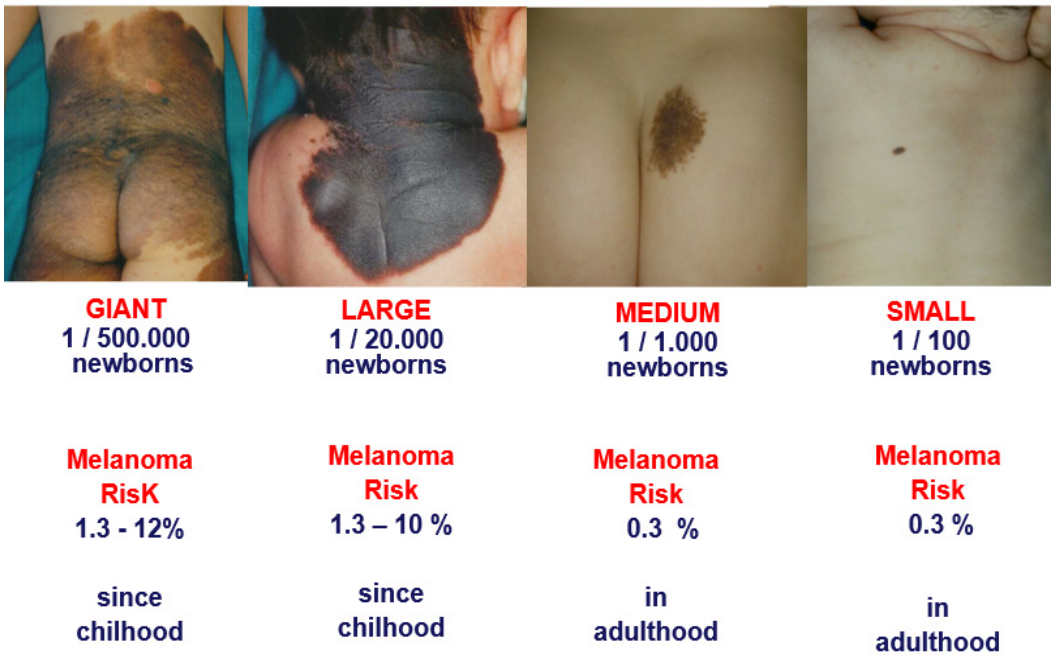

Fig. 1. *Congenital melanocytic nevi classified according to their size.*

Large and Giant Congenital Melanocytic Nevi (LGCMN)

The incidence of Large CMN is estimated at 1:20,000 births, while those Giant, which affect large skin surfaces, sometimes resembling a typical "garment" distribution, have an incidence of 1:500,000 births.

Their colour can vary from light brown to black and is often heterogeneous. The surface can be smooth or irregular, with clear or blurred edges, mostly being irregular. It is common to observe medium or small MN on their periphery or at a distance (*satellite nevi*). Some of these are congenital, while others are acquired.

Over time, LGCMNs can undergo significant changes. They increase in size harmoniously with somatic growth, become darker and more raised. They may cover themselves with terminal hair (sometimes present since birth) and frequently develop papular or nodular *pseudotumoral lesions*, due to deep infiltration of me-

lanocytic cells. In the first years of life, some LGCMNs tend to lighten, especially in skin folds or areas of friction. Those localized on the scalp are particularly prone to spontaneous involution.

LGCMNs, aside from being a potential aesthetic concern, though perhaps less impactful in today's era of widespread tattoos, can give rise to melanoma. Current estimates suggest that melanoma develops at the skin level in fewer than 5% of cases (3), but when considering nervous system involvement, the overall occurrence ranges from 1.3% to 12% of cases (4). Nevi located on the limbs and satellite ones do not appear to have a risk of melanoma.

Neurocutaneous melanocytosis (NCM) - characterized by an increased presence of melanocytic cells in the leptomeninges, brain, and spinal cord - is found in fewer than 7% of patients with LGCMNs and is believed to heighten the risk of melanoma (5).

In more than half of cases melanoma arises before the fifth year of life and originates from melanocytic cells deep within the dermis, leading to nodular lesions. Nodular melanoma is clinically elusive and challenging to detect, as it closely resembles benign nodular formations (*pseudotumoral lesions*) commonly found in LGCMNs. Consequently, diagnosis is often delayed, resulting in a poor prognosis.

LGCMNs should be surgically removed whenever technically feasible, with the primary goals of preserving functionality and achieving aesthetic improvement. However, the extensive size of these nevi often poses a significant limitation to surgical intervention. When direct suture excision, rotation flaps, skin expanders, or dermo-epidermal autografts are not viable options, alternative physical treatments such as dermabrasion, laser therapy, or curettage may be considered.

Neonatal curettage, performed within the first 4–6 weeks of life - when most nevus cells still have a superficial, junctional, distribution - is regarded as a reasonable treatment due to its relative effectiveness and ease of execution (6). The objective of all these treatments is to lower the risk of neoplasia rather than to eliminate it entirely, as numerous melanocytic cells remain in deep tissues and extracutaneous areas. At the same time, they offer the added benefit of aesthetic enhancement.

Careful clinical monitoring should always be scheduled, even following treatment. Due to the inherent challenges in clinical and dermoscopic evaluation - stemming from the frequent morphochromatic irregularities and the presence of hypertrichosis - follow-up should be conducted at specialized centers with expertise in managing LGCMNs.

LGCMNs, particularly those affecting the cervico-cephalic region, may be associated with leptomeningeal melanosis and neurological anomalies, such as congenital neurocutaneous melanosis. For early diagnosis, even in the absence of neurological symptoms, a thorough neurological examination and magnetic resonance imaging are essential.

LGCMNs affecting the lumbosacral region may be accompanied by spinal anomalies, including spina bifida and myelomeningocele.

Medium and Small Congenital Melanocytic Nevi (MSCMN)

These nevi constitute the vast majority of all CMNs and exhibit highly variable clinical appearances. While Medium CMNs are consistently identifiable as congenital - even in the absence of specific anamnestic information - due to their size exceeding 1.5 cm (or 1 cm in young children), Small CMNs cannot be reliably classified as congenital except at birth, as they are morphologically indistinguishable from many acquired melanocytic nevi.

Both Medium and Small CMNs can double in size and undergo changes in shape and color over time. These morphological alterations, particularly common during puberty, are not considered to have pathological significance. The potential development of melanoma within a Medium or Small CMN in adulthood is well-documented, as numerous cases have been reported in the literature. However, the quantitative assessment of this risk remains controversial. Various studies indicate a highly variable incidence of melanoma in MSCMNs, though most authors currently estimate that the risk does not exceed 1%, possibly similar to that of the rest of healthy skin (3, 4, 7).

Unlike LGCMNs, the majority of melanomas associated with MSCMNs develop in adulthood or, at the earliest, after puberty.The surgical removal of these nevi is primarily pursued for aesthetic reasons (Fig. 2) rather than for cancer prevention. In most cases, these procedures are straightforward and highly cost-effective, often performed on an outpatient basis or in a day hospital setting.

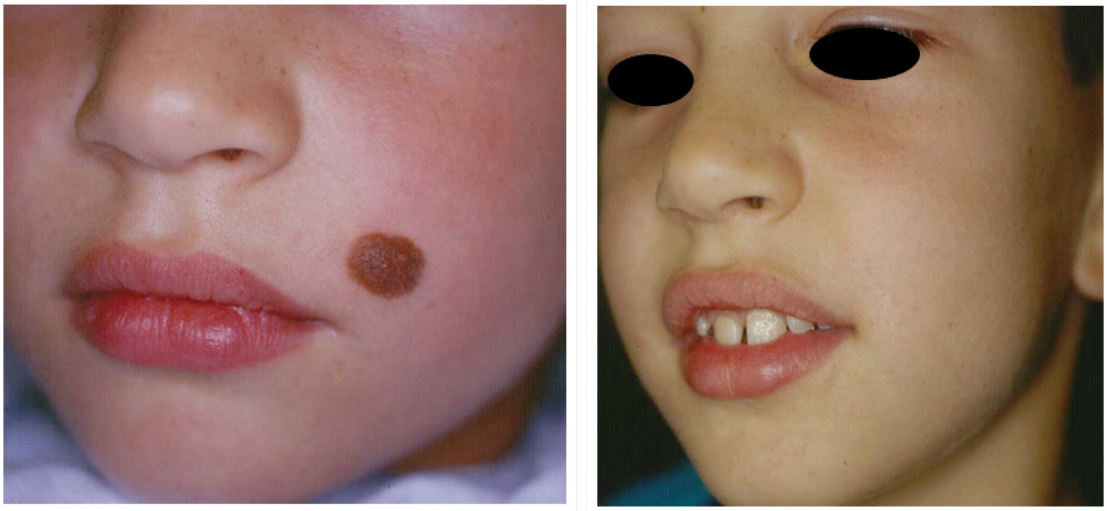

Fig. 2. *Surgical removal for aesthetic purposes.*

The three most important things to tell parents

1. Congenital melanocytic nevi are benign lesions with a very low risk of melanoma.

2. Large and Giant nevi require long-term monitoring, preferably at specialized centers. Medium and Small nevi, however, can be safely monitored by a pediatrician or dermatologist.

3. Surgical removal of Medium and Small nevi is optional but can be beneficial - even in preschool-aged children - if aesthetic concerns arise. However, the procedure should always be pursued for cosmetic improvement rather than as an oncological preventive measure, ensuring that aesthetic outcomes are not overlooked.

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