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Dermoscopy of Benign and Malignant Neoplasms in the Pediatric Population

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Dermoscopy is a noninvasive technique that enables visualization of subsurface colors and structures within the skin that are imperceptible to the naked eye. The dermatoscope allows the physician to examine both the macroscopic and microscopic primary morphology of skin lesions, identify subtle clinical clues, confirm naked-eye clinical diagnoses, and monitor treatment progress while posing little threat to the young patient. Dermoscopic findings have been formulated into diagnostic criteria that assist experienced clinicians in differentiating benign and malignant neoplasms. In this review, clinical morphology of melanocytic nevi and melanoma in the pediatric population is examined and the relevant dermoscopic findings and histopathologic correlates that aid in the diagnosis and management of these lesions are described.

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Children, like their adult counterparts, often present to the dermatologist with pigmented lesions that are new or changing. Unique to the pediatric population, however, is that they are in a dynamic growing phase of life. One sign of this dynamic phase is manifest by the development, growth, and occasional involution of nevi. In addition, children with certain genetic syndromes, such as epidermolysis bullosa, and basal cell nevus syndrome have unique pathophysiologies that necessitate careful evaluation and follow-up of their pigmented skin lesions.

Melanocytic neoplasms encountered during childhood may be divided into 3 distinct classes; congenital nevi, acquired nevi, and melanoma. Congenital melanocytic nevi (CMN) consist of nevi that are clinically evident at birth and nevi-manifesting congenital features that become clinically apparent shortly after birth (ie, tardive CMN). Nevus spilus and segmental speckled-lentiginous nevus are also considered CMN. Melanocytic nevi, such as junctional nevi, compound nevi, dermal nevi, blue nevi, and Spitz nevi, are classified as acquired melanocytic nevi if they develop many months to years after birth.

Although rare, the incidence of pediatric melanoma is increasing, and it has become imperative that clinicians include melanoma in the differential diagnosis of atypical pigmented and even amelanotic lesions in children. It is important to acknowledge that melanoma can develop in healthy children, as well as those with underlying genetic or immunologic disorders, such as xeroderma pigmentosum. Approximately one-half of all pediatric melanomas arise in a *de novo* fashion, whereas the other 50% emerge in association with preexisting lesions. Nearly 30% of childhood melanomas arise within giant CMN, and roughly 20% develop in association with acquired melanocytic nevi.

Clinical features of melanoma in children can be very subtle and may mimic Spitz nevi and angiomas, often resulting in the missed opportunity to perform a biopsy. Retrospective studies suggest that at the time of diagnosis, up to 60% of pediatric melanomas are of intermediate thickness, which may in part be attributable to a delay in the clinical diagnosis. By contrast, many benign CMN and acquired nevi can manifest clinical features resembling melanoma, leading to unnecessary biopsies leaving scars on young patients. Therefore, any technique that increases the sensitivity for detecting melanoma while at the same time improving specificity would be highly valued by patients, parents, and clinicians. One technique that can improve a physician's ability to detect melanoma and avoid superfluous biopsies is dermoscopy.

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Table 1 Dermoscopic Structures and Their Histopathological Correlates

Dermoscopic Structures	Definition	Histopathological Correlation
Pigment network (reticulation)	Grid-like network consisting of pigmented "lines" and hypopigmented "holes."	Melanin in keratinocytes and/or melanocytes along the epidermal rete ridges
Pseudonetwork	In facial lesions, diffuse pigmentation interrupted by nonpigmented follicular openings, appearing similar to a network.	Pigment in the epidermis or dermis interrupted by follicular and adnexal openings of the face
Structureless (homogenous) areas	Areas devoid of dermoscopic structures and without regression. These areas can be pigmented or nonpigmented. if the area is uniformly dark, it is referred to as a "blotch" (see below)	Lack of melanin or presence of melanin in all layers of the skin
Dots	Small, round structures less than 0.1 mm in diameter that may be black, brown, gray or bluish	Aggregates of melanocytes or melanin granules. Black dots represent pigment in the upper epidermis or stratum corneum. Brown dots represent pigment at the dermoepidermal junction. Gray-blue dots represent pigment in the papillary dermis
Peppering	Tiny, blue-gray granules	Melanin deposited as intracellular (mostly within melanophages) or extracellular particles in the upper dermis
Globules	Round to oval structures that may be brown, black or red with diameters greater than 0.1 mm.	Nests of melanocytes in the dermis.
Streaks (pseudopods, radial streaming)	Radially arranged, projections of dark pigment (brown to black) at the periphery of the lesion.	Confluent junctional nests of melanocytes.
Blotches	Dark brown to black, usually homogenous areas of pigment that obscure underlying structures.	Aggregates of melanin in the stratum corneum, epidermis and upper dermis.
Regression areas	White, scar-like depigmentation (lighter than the surrounding skin, shiny white under polarized dermoscopy) often combined with or adjacent to blue-gray areas or peppering.	Scar-like changes: thickened fibrotic papillary dermis, dilated blood vessels, sparse lymphocytic infiltrates and variable numbers of melanophages.
Blue-white veil	Irregular, confluent blue pigmentation with an overlying white "ground glass" haze	Aggregation of heavily pigmented cells (usually melanoma cells) and/or melanophages in combination with compact orthokeratosis of the stratum corneum and acanthosis (thickened epidermis).
Vascular pattern	Type of Vascular structure	See in
	Arborizing vessels	Basal cell carcinoma
	Irregular hairpin vessels	Melanoma
	Dotted vessels (red dots and globules)	Melanoma, clark nevi, spitz nevi, bowen's disease and psoriasis
	Glomerular vessels	Melanoma, bowen's disease, venous stasis
	Irregular linear-polymorphous vessels	Melanoma
	Milky red areas	Melanoma, spitz nevi, clark nevi
Milia-like cysts	Round whitish or yellowish structures that shine brightly (like "stars in the sky") under nonpolarized dermoscopy.	Intraepidermal keratin cysts
Comedo-like openings	"Blackhead"-like plugs on the surface of the lesion	Concave clefts in the surface of the epidermis, often filled with keratin.
Fingerprint-like structures	Thin light brown parallel running lines	Probably represent thin, elongated pigmented epidermal rete ridges.
Ridges and fissures	Cerebriform surface resulting in gyri (ridges) and sulci (fissures). Confluence of adjacent comedo-like openings will create a fissure.	Wedge-shaped clefts of the surface of the epidermis often filled with keratin (fissures).
Moth-eaten border	Concave invaginations of the lesion border	—
Leaf-like areas	Brown to gray-blue discrete bulbous structures resembling a leaf pattern	Large, complex nodules of pigmented basal cell carcinoma in the upper dermis
Spoke-wheel-like structures	Well circumscribed brown to gray-blue-brown radial projections meeting at a darker brown central hub	Nests of basal cell carcinoma radiating from the follicular epithelium
Large blue-gray ovoid nests	Large, well-circumscribed areas, larger than globules.	Large nests of basal cell tumor in the dermis
Multiple blue-gray globules	Round well circumscribed structures which, in the absence of a pigment network, suggest basal-cell carcinoma	Small nests of basal cell tumor in the dermis
Lacunae	Red, maroon or black lagoons	Dilated vascular spaces
Parallel patterns	On acral areas, parallel rows of pigmentation following the furrows (nevi) or ridges (melanoma) of the dermoglyphics	Pigmented melanocytes in the furrows (Crista limitants) or ridges (Crista intermedia) of acral skin

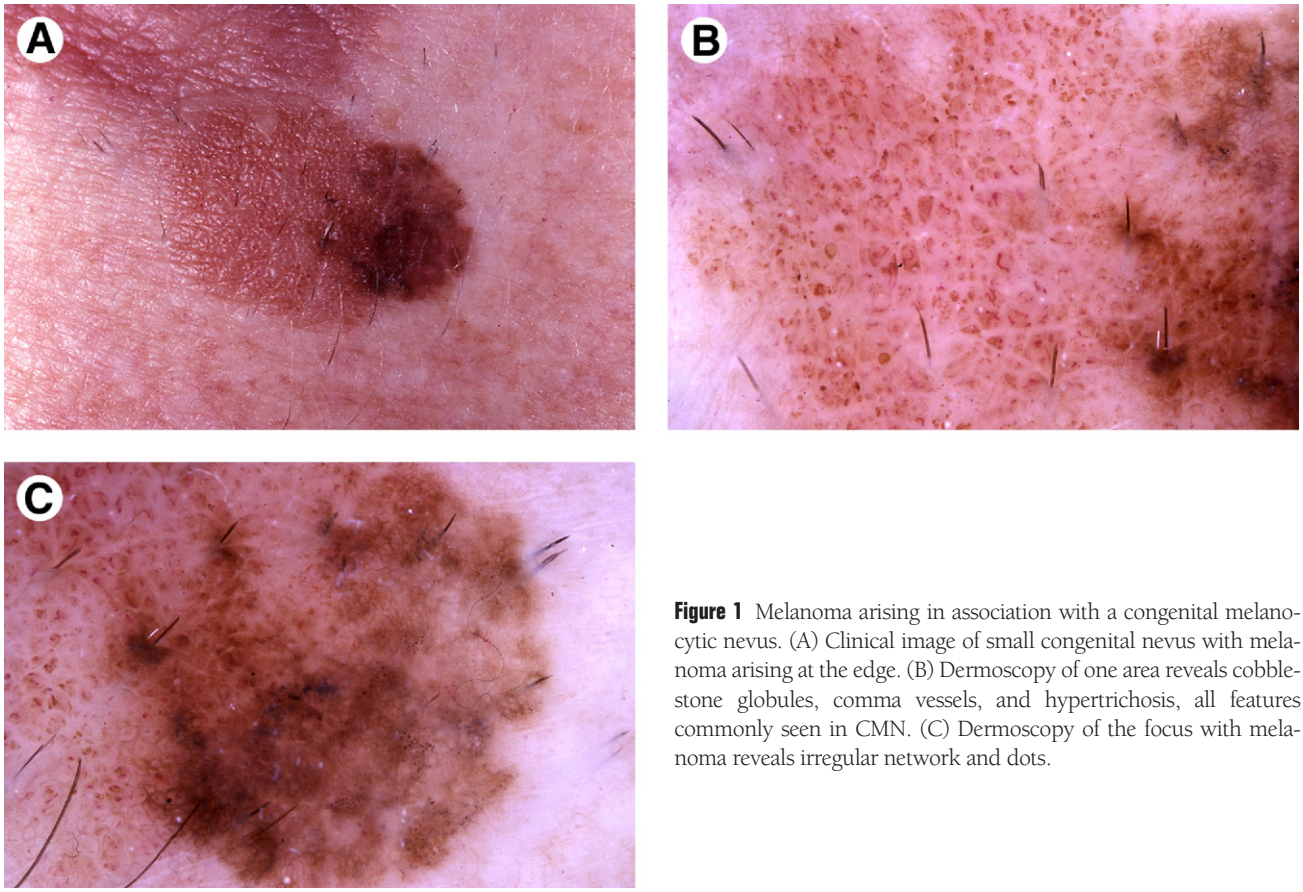

Figure 1 Melanoma arising in association with a congenital melanocytic nevus. (A) Clinical image of small congenital nevus with melanoma arising at the edge. (B) Dermoscopy of one area reveals cobblestone globules, comma vessels, and hypertrichosis, all features commonly seen in CMN. (C) Dermoscopy of the focus with melanoma reveals irregular network and dots.

Dermoscopy serves as a unique and harmless tool to aid in the cutaneous evaluation of lesions in children. Its painless modality and curiously bright lights intrigue the pediatric patient rather than intimidate. The dermatoscope allows the clinician to examine structures in the epidermis and reticular dermis otherwise invisible to the naked eye. The presence or absence of certain structures and colors and their relative distribution within a lesion can be used to guide management decisions, including whether to perform a biopsy. For example, the diagnostic accuracy of Spitz nevi with dermoscopy is 93% compared with only 56% with naked eye examination.[1] Other studies have shown that examination of nevi with dermoscopy decreases the need for surgical removal of suspect lesions from 15.6% to 9%, when compared with unaided visual assessment alone.[2]

Dermoscopy may either be polarized or nonpolarized. In nonpolarized dermoscopy the lens of dermatoscope must come in direct contact with the skin with oil, alcohol, or water used as a liquid interface. Polarized dermatoscopy, by contrast, does not require instrument-to-skin contact or an immersion liquid. When one is examining anxious children who resist and fight against any physical contact, polarized dermoscopy often should be the instrument of choice. In this article the dermoscopic features of benign and malignant neoplasms in the pediatric populations are reviewed, with special focus on its use in genodermatoses. The reader is referred to Table 1 for a review of basic dermoscopic terminology.

Melanocytic Lesions and Melanoma

Congenital Melanocytic Nevi

CMN have been arbitrarily classified by size into small (<1.5 cm), intermediate (1.5-19.9 cm), and large (>20 cm).[3] Small- or medium-size CMN are usually light to dark brown, have a smooth surface, and a well-demarcated border. Over time, they may darken or lighten and acquire coarse hairs. Some CMN may show variation in color, have irregular borders, and present an uneven surface.[4]

Knowledge of the dermoscopic structures and patterns found in CMN can assist the follow-up of these lesions and detection of features suspicious for melanoma (Fig. 1). Most small- and medium-sized CMN are fairly homogeneous in appearance, both clinically and dermoscopically, whereas large CMN are often heterogeneous, having multiple islands of color and an irregular topography; however, each "island" within the large CMN tends to be fairly uniform.[5] Combinations of 2 main dermoscopic patterns, reticular and globular, are present in congenital nevi. CMN with a prominent reticular pattern usually are found on the lower extremities, whereas those with a prominent globular pattern tend to occur on the head, neck, and torso.[6]

"Reticular network" describes a honeycomb-like arrangement of dark pigment lines; pigment lines correspond to elongated hypermelanotic rete ridges and the clear spaces

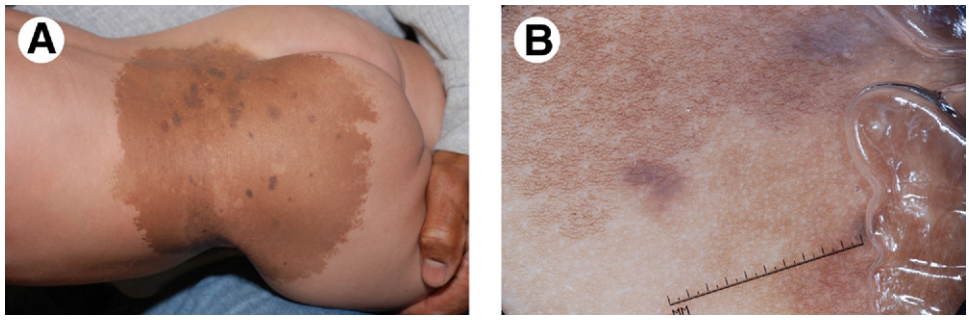

Figure 2 Large CMN. (A) Clinical image. (B) Dermoscopic image displaying a reticular pattern.

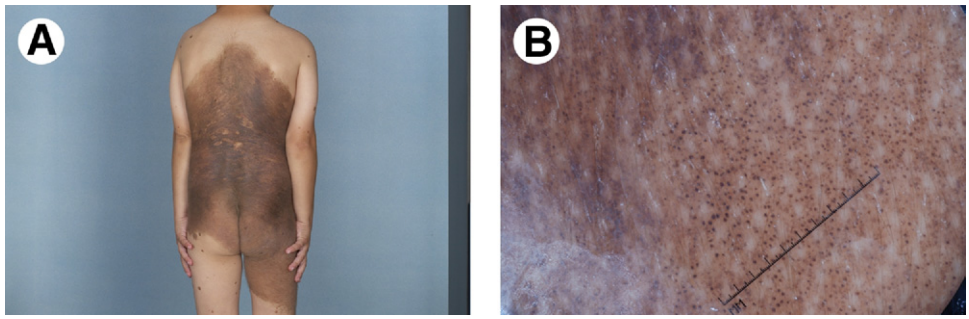

Figure 3 Large CMN. (A) Clinical image. (B) Dermoscopic image displaying a globular pattern.

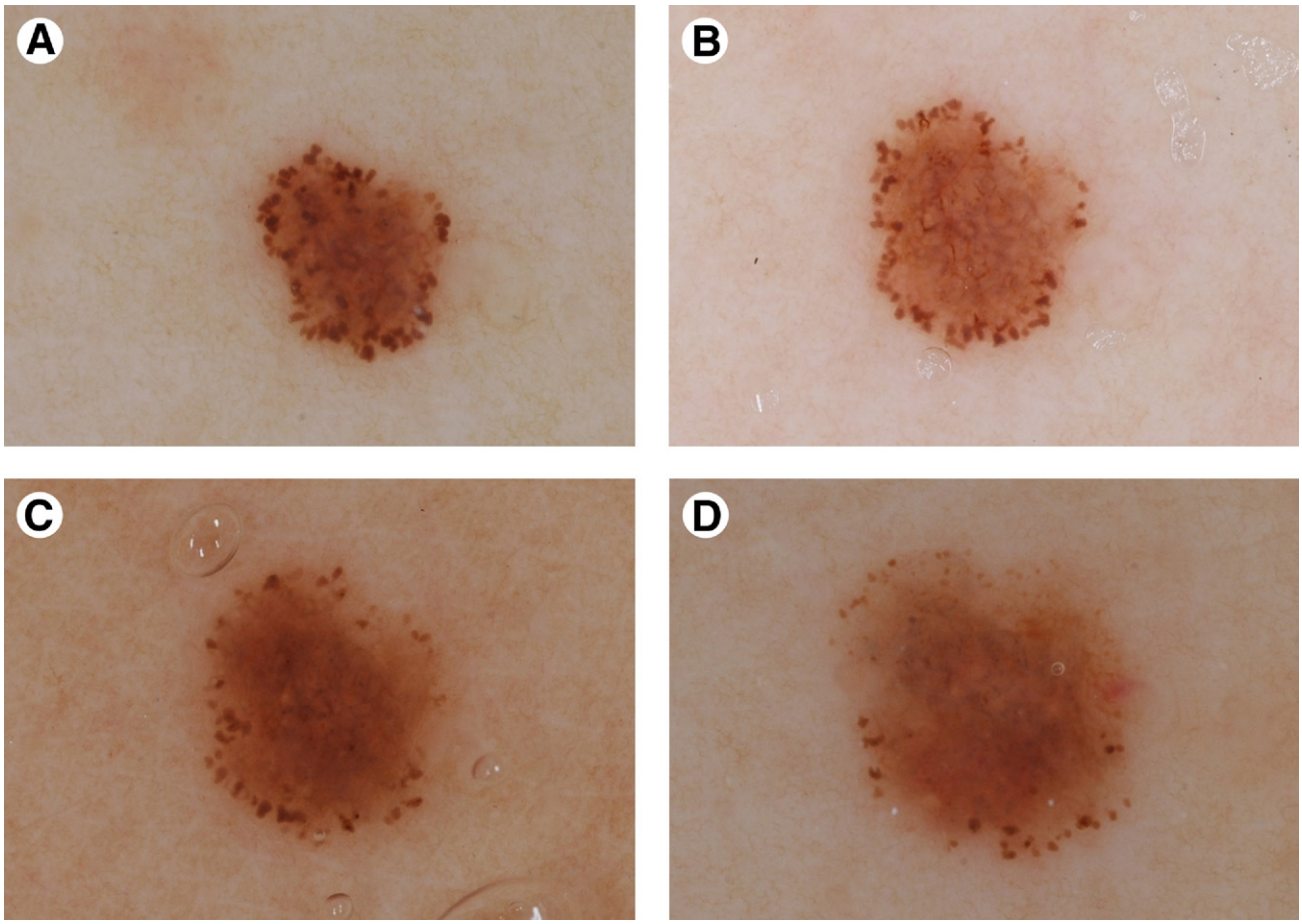

Figure 4 Actively, growing nevus. Dermoscopic image of peripheral globular pattern. Panels A-D were taken at baseline, 2,4, and 10 months, respectively.

between the lines correspond to the suprapapillary plate (Fig. 2).[7] "Globulars" refer to sharply circumscribed, round-to-oval aggregated structures that represent nests of melanin-containing nevus cells within the dermis (Fig. 3).[7]

Other dermoscopic structures frequently observed in CMN include (1) milia-like cysts, (2) hypertrichosis, and (3) perifollicular pigment changes. In addition, almost 70% of CMN reveal vascular structures under dermoscopy,[6] most commonly comma vessels, dotted vessels, and serpentine vessels.[5]

Acquired Melanocytic Nevi

Environmental and genetic factors have been linked to the development of nevi.[8] Acquired nevi usually manifest dermoscopically with a reticular network (diffuse or patchy), peripheral reticular network with central hyperpigmentation, peripheral reticular network with central hypopigmentation, or homogeneous pattern.[9,10] However, it is not uncommon to also see acquired nevi with a globular pattern.

Dermoscopic and histologic evaluation of one subset of growing acquired nevi show peripheral globules and nevo-melanocytic junctional nests at the perimeter of a nevus, respectively (Fig. 4). These lesions tend to grow in a symmetric, centrifugal fashion, with peripheral globules becoming progressively sparser and eventually disappearing. Once the lesion has matured, the peripheral globules are no longer visible and the nevus manifests a reticular or homogeneous pattern.

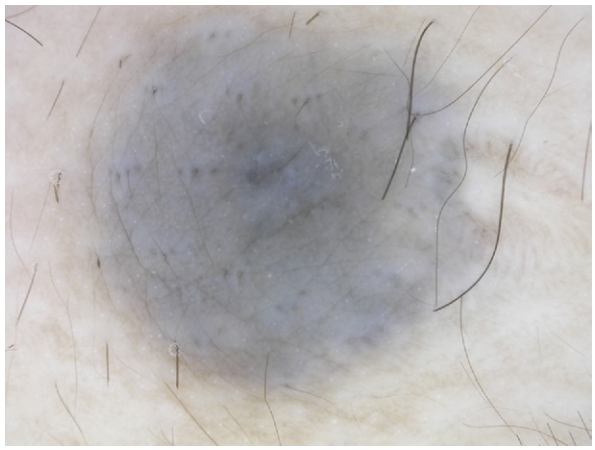

Figure 5 Blue nevus. Dermscopic image exhibiting homogenous blue-gray pigmentation.

It is important to acknowledge that because many nevi in children and young adults have not yet undergone senescence,

Table 2 Benign Dermoscopic Patterns

Global Pattern	Definition
Reticular diffuse	Diffuse homogeneous network with uniform thickness and color of lines. The holes of the network are of relatively uniform size. The network tends to fade at the periphery. This pattern can be seen in congenital nevi, especially those located on the lower extremity, and in acquired nevi.
Reticular patchy	Homogeneous network with uniform thickness and color of lines. However, the network is not contiguous because of the presence of homogeneous structureless areas. This pattern can be seen in congenital nevi, especially those located on the lower extremity, and in acquired nevi.
Peripheral reticular with central hypopigmentation	A relatively uniform network at the periphery of the lesion with a central homogeneous and hypopigmented structureless area. This type of acquired nevus is more common in fair skin phenotypes.
Peripheral reticular with central hyperpigmentation	A relatively uniform network at the periphery of the lesion with a central homogeneous and hyperpigmented blotch. This type of acquired nevus is more common in darker skin phenotypes.
Globular	Globules of similar shape, size and color distributed symmetrically throughout the lesion. This pattern is seen most commonly in congenital nevi.
Peripheral reticular with central globules	A relatively uniform network at the periphery of the lesion with a central globule. This pattern is seen most often in nevi with a congenital histopthology pattern.
Peripheral globules with central network or homogeneous area	The central component of this type of nevus is either reticular or homogeneous. Relatively uniform globules surround the entire perimeter of the lesion. This represents an acquired growing nevus that has not yet undergone senescence.
Homogeneous tan, brown, or blue pigmentation	Primarily diffuse structureless pattern with or without reticular networks fragments and/or remnant globules. When tan in color they usually represent acquired nevi in fair skin phenotypes. When brown they usually represent congenital nevi. When blue in color they represent blue nevi.
Two component	These lesions reveal 2 different patterns (reticular-globular, reticular-homogeneous, or globular-homogeneous). One-half of the lesion manifests one pattern whereas the other half of the lesion manifests a different pattern.
Multicomponent	Symmetrically distributed globules, reticulation, blotches, dots, veil, regression structures and/or structureless areas (3 or more of these must be present).
Starburst	Streaks (radial streaming or pseudopods) present around the perimeter of the lesion giving the appearance of an exploding star.

their nevi will undergo change.[11] However, these changes usually involve the entire nevus, such as enlarging or becoming darker. In contrast, focal changes developing in nevi should always be evaluated carefully with a low threshold for biopsy. Other benign nevus patterns are described in Table 2.

Blue Nevi

Blue nevi are benign neoplasms composed of dermal melanocytes that are believed to have arrested their migration in the dermis en route to the Dermo-epidermal junction during normal melanocyte migration in fetal life.[12] They typically develop during childhood or adolescence. Several clinical variants of blue nevi have been described.

Dermoscopic features of the common blue nevus are quite specific.[13] Common blue nevi have homogenous blue-gray, well-circumscribed pigmentation that is diffuse and structureless, ie, lacking network, dots, globules, and vessels (Fig. 5).[5] The presence of any structures, such as globules or various colors that make the lesion appear heterogeneous, should prompt suspicion of melanoma or a combined nevus.

Nevus Spilus (Speckled Lentiginous Nevus)

Nevus spilus is a patch of hyperpigmentation speckled with darker macules (*nevus spilus maculosus*) or papules (*nevus spilus papulosus*) that represent a variant of a congenital melanocytic nevus.[14-16]

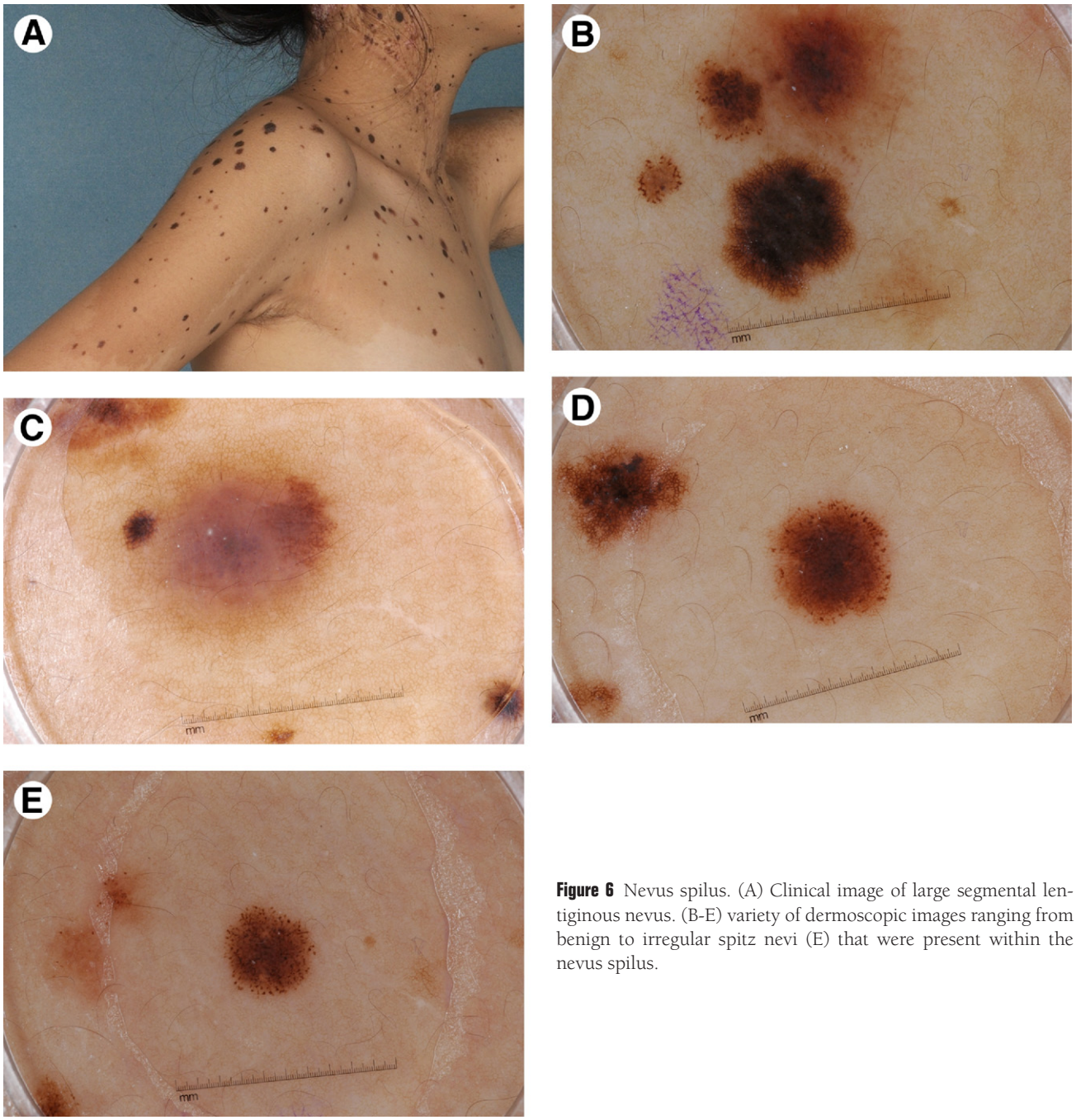

Figure 6 Nevus spilus. (A) Clinical image of large segmental lentiginous nevus. (B-E) variety of dermoscopic images ranging from benign to irregular spitz nevi (E) that were present within the nevus spilus.

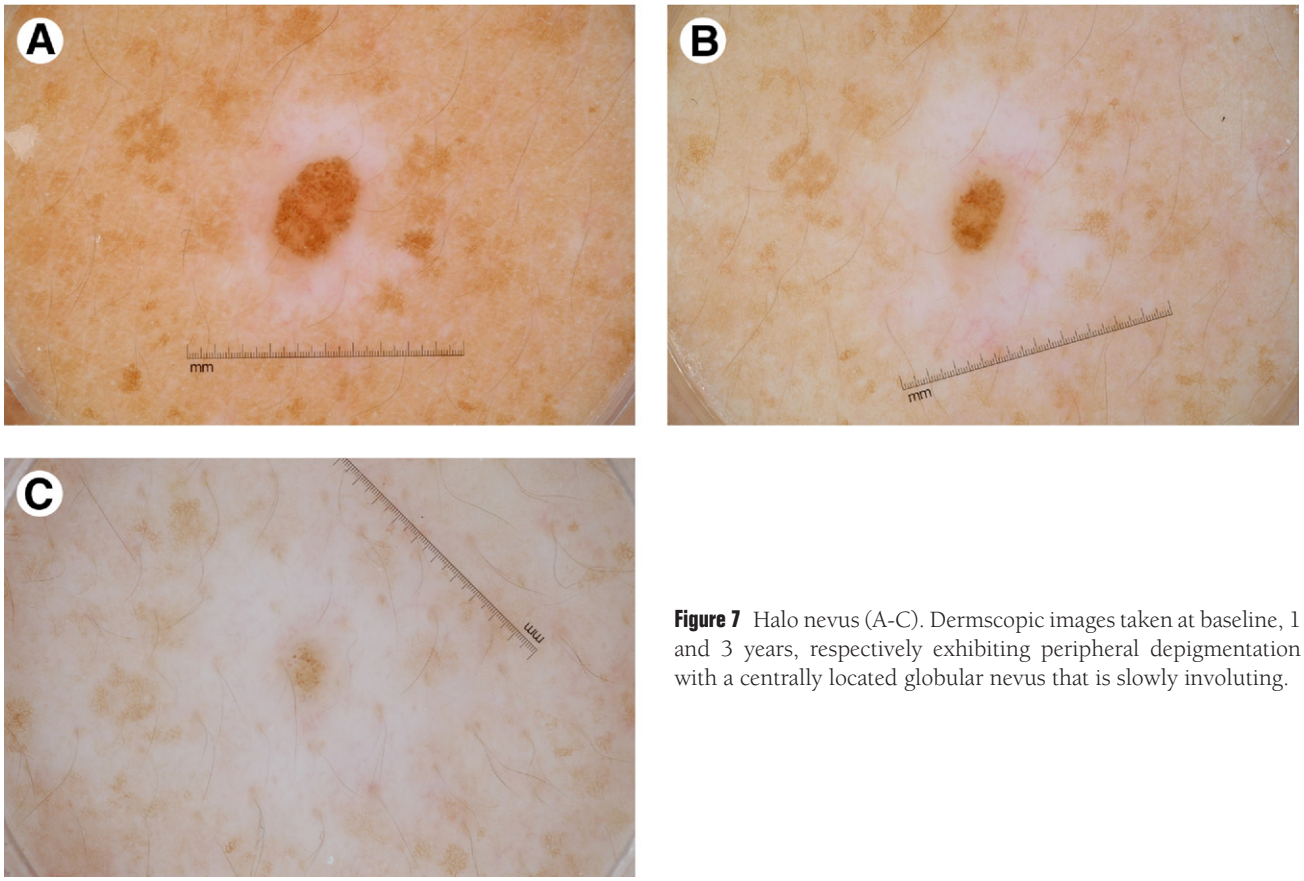

Figure 7 Halo nevus (A-C). Dermscopic images taken at baseline, 1 and 3 years, respectively exhibiting peripheral depigmentation with a centrally located globular nevus that is slowly involuting.

Nevus spilus has been referred to as a "melanocytic garden"[17,18] because many types of melanocytic proliferations may develop within a nevus spilus, including Spitz nevi, blue nevi, junctional nevi, compound nevi, intradermal nevi, and melanoma (Fig. 6).[19] Under dermoscopy, the background of a nevus spilus can appear homogeneous tan in color or can manifest a faint and delicate network. The isolated darker or pink macules and papules within the nevus spilus often reveal dermoscopic features consistent with the type of nevus present (ie, Spitz, blue nevi, compound nevi, etc.).[19]

Halo Nevi (Sutton's)

Although the halo phenomenon is usually found in conjunction with the benign halo nevus, there have been reports of the halo effect in melanoma.[20,21] Approximately 80% of halo nevi exhibit a dermoscopic globular and/or homogenous pattern in the center of the lesion with a surrounding halo of depigmentation (Fig. 7).[22] A light-brown and pink central area with dotted vessels is seen occasionally in nevi that have completely regressed.[22] In one study, halo nevi were sequentially followed with digital images, and it was observed that 51.5% exhibited a decrease in halo size, whereas 27.3% showed an enlargement of halo size.[22] However, all nevi within the halo exhibited a reduction in nevus area. Despite changes in size of the halo or the nevus, the dermoscopic features of the nevus remained unchanged (Fig. 7).

Spitz Nevus

Spitz nevi may take on different appearances in children, at times presenting as a pink-red papule or a jet black papule; they may occur anywhere on the body but have a predilection for the head and neck.[12] Since its original description as a lesion that was mistaken histologically for a melanoma, Spitz nevi have been perceived as a unique biological entity with characteristic histologic features.[23]

The diagnostic accuracy for Spitz nevi has increased from 56% to 93% when dermoscopy is used.[1] Several dermoscopic patterns can be appreciated. The patterns include thick (atypical) reticular, atypical globular, star burst, homogeneous (pink or with a black lamella), negative pigment network, and atypical/multicomponent.[5,24] The archetypal Spitz nevus pattern remains the star burst pattern (Fig. 8A) and it is seen in at least half of Spitz nevi. In the star burst pattern, likened to the image of an exploding star, the lesion has a homogenously distributed pattern of streaks, pseudopods and/or globules around the perimeter of the lesion. The center of these nevi usually displays a homogeneous pattern ranging in color from blue-gray to brown or brown-black. When present, the star burst pattern has been reported to allow a diagnostic sensitivity of 96%.[25]

The negative network pattern, with or without chrysalis-like structures, is another common pattern encountered in Spitz nevi (Fig. 8B). The negative network consists of light

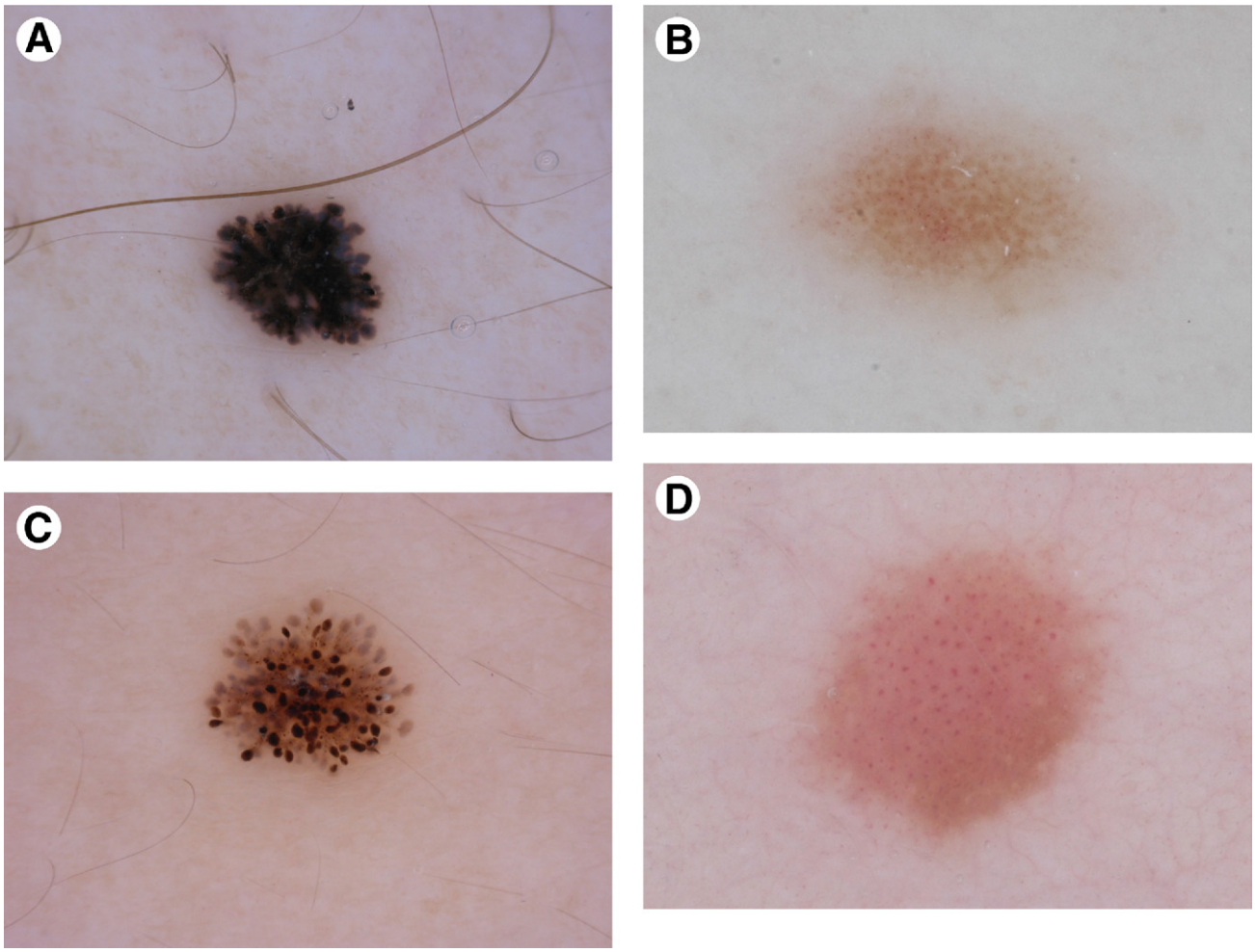

Figure 8 Spitz nevi. (A) Dermoscopic pattern exhibiting star burst pattern. (B) Dermoscopic image of negative pigment pattern. (C) Dermoscopic image of globular pattern (D) Dermoscopic image of pink spitz nevus with dotted vessels. (Images A, and C are courtesy of Iris Zalaudek, MD; images B and D are courtesy of Dr Marghoob.)

areas making up the "grid" of the network, with the dark areas presenting as the "holes." The chrysalis-like structures consists of bright white orthogonal lines, which likely correspond to altered papillary dermal collagen. Because collagen is birefringent, it should come as no surprise that the chrysalis structures can easily be seen with polarized light dermoscopy, and it is almost impossible to see them with standard nonpolarized light dermoscopy.

Approximately 22% of Spitz nevi manifest a globular pattern.[26] The globular pattern consists of globules varying in size and color, from brown-black to blue-gray (Fig. 8C). These globules tend to be distributed throughout the lesion with prominence of central gray-blue pigmentation. The homogenous pattern usually consists of a pink or black (black lamella) lesion. Although in most homogeneous Spitz tumors one cannot discern any dermoscopic structures, in some pink Spitz nevi one can see dotted or irregular vessels (Fig. 8D). At times, pink Spitz nevi that possess dotted or irregular vessels cannot be differentiated from amelanotic melanoma.[5] In addition, there exists another group of Spitz nevi that manifest an atypical pattern. These lesions are often asymmetric and/or appear disorganized, and many of them will have an asymmetric multicomponent pattern and will have features commonly associated with melanoma.[27] It is currently clinically impossible to differentiate an atypical Spitz nevus from melanoma.[5]

Melanoma

During the last 30 years, the incidence of melanoma in the 10- to 19-year age group in the United States has increased by 3%.[28] Because of the varied clinical presentations, the diagnosis of melanoma in children remains a challenge.[29] The 2 most common histologic subtypes of melanoma diagnosed in children are the superficial spreading and nodular. Superficial spreading melanomas grow slowly and often manifest at least some of the clinical ABCD features (ie, asymmetry, border irregularity, color variations, and dimension) and dermoscopic structures listed in Table 3.[5] In contrast, nodular melanomas tend to grow rapidly and they are often amelanotic and symmetric; lacking the clinical ABCD features of melanoma. Nodular melanomas have a broad presentation, from hypopigmented and erythematous papules to fungating red nodules.

Because the pigment in amelanotic melanomas is partially

Table 3 Histopathologic Correlates of Dermoscopic Structures Found in Superficial Spreading Melanoma

Dermoscopic Structure	Description	Histopathologic Correlate
Atypical pigment network	Reticulated grid of brown lines with broadened, thickened, or darkened areas that may end abruptly at the periphery	Atypical lentiginous or nested melanocytic proliferation along the dermoepidermal junction
Streaks	Linear, radially oriented pigmented projections at the periphery of a lesion	Confluent junctional nests of pigmented melanoma cells at the periphery (radial growth phase)
Negative pigment network	The "negative" of the pigment network, consisting of hypopigmented lines making up the grid, and dark areas filling up the "holes"	Thin elongated rete ridges and large tubular melanocytic nests within a widened papillary dermis
Chrysalis	Fine, white, shiny streaks within a lesion, visible only under polarized light dermoscopy	Remodeled or new dermal collagen
Atypical dots and globules	Dark, punctuate, or round to oval structures of varying shape, size, color, and distribution within a lesion	Junctional or dermal nests of melanocytes; dots can also represent pagetoid nests
Irregular blotches	Dark areas of diffuse pigmentation with irregular shapes, sharp margins, or eccentric locations	Melanin pigmentation throughout the epidermis or dermis or both
Blue-white structures over raised areas	White areas, blue areas, or both, overlying raised or thick portions of a lesion	Compact orthokeratosis overlying melanophages, melanocytes, or free melanin in the papillary dermis
Blue-white structures over flat areas	White areas, blue areas, or both, overlying flat or thin portions of a lesion	Fibrosis within the papillary dermis and melanosis (melanophages and free melanin "dust" within the dermis)
Atypical vascular structures	Milky red areas, dotted, linear, or twisted red structures with differing sizes	Tumor-induced angiogenesis
Peripheral brown structureless areas	Peripherally arranged light brown or tan areas of variable shape lacking perceptible structures	Flattening of the rete ridges with pagetoid spread of atypical melanocytes

or entirely absent, dermoscopy must rely on evaluating the vascular structures, if present, for clues to the diagnosis. In an effort to avoid compressing blood vessels from pressure placed on the skin during direct contact dermoscopy, ultrasound gel remains the ideal interface medium. However, because polarized dermatoscopes do not require direct skin contact, these instruments remain the ideal ones for visualizing blood vessels in skin lesions.[5] A biopsy should be strongly considered for any nodular pink lesion manifesting dotted vessels, linear irregular vessels, or serpentine vessels (Fig. 9C).[5]

Differentiating Benign Nevi from Melanoma

Melanocytic lesions in which the dermoscopic structures are organized and symmetrically distributed and belong to one of the known benign dermoscopic patterns listed in Table 2 have an exceedingly low risk for being melanoma and may safely be monitored. Rarely, melanomas will display a symmetric and organized pattern; however, this apparently banal configuration will not adhere to one of the known benign patterns as listed in Table 2 (Fig. 9B). All other lesions should be viewed with caution, especially if any melanoma-specific structures listed in Table 3 are seen (Fig. 9A).

Another important fact to remember is that melanomas developing in children are often nodular and/or amelanotic. These melanomas are often difficult to diagnose, but the presence of irregular blood vessels under dermoscopy can correctly identify these melanomas (Fig. 9C).[30] Finally, the greatest melanoma masquerader encountered in the pediatric population is the Spitz nevus. any dermoscopic structure specific to melanoma can on occasion be seen in Spitz nevi (Table 3).[31] However, in typical Spitz nevi these structures are arranged symmetrically (for example, pseudopods arranged symmetrically around the entire perimeter of the lesion, forming a star burst pattern), whereas in melanoma and some atypical Spitz nevi these structures are present focally and in an asymmetric distribution.

Genetic Syndromes

Basal Cell Nevus Syndrome

Basal cell nevus syndrome (BCNS), also known as Gorlin–Goltz syndrome, is an autosomal-dominant inherited disease characterized by a spectrum of developmental and cutaneous abnormalities, including early onset of nevoid basal cell hamartomas, carcinomas and volar pits.[32] Dermoscopy can confirm a diagnosis of BCNS by aiding in the early detection of basal cell carcinomas (BCCs) and acral pits.[33,34] Under dermoscopy, BCCs have an absent pigment network and presence of any one of the following features: arborizing vessels (Fig. 10), ulceration, blue-gray ovoid nests, leaf-like areas, spoke wheel areas, multiple nonaggregated blue-gray globules, or shiny white areas.[34-36]

Dermoscopy can also identify the acral pits. A pit usually has an irregular shape and a pale, pink or flesh colored base (Fig. 11A). Dermoscopically, the pits appear to be small,

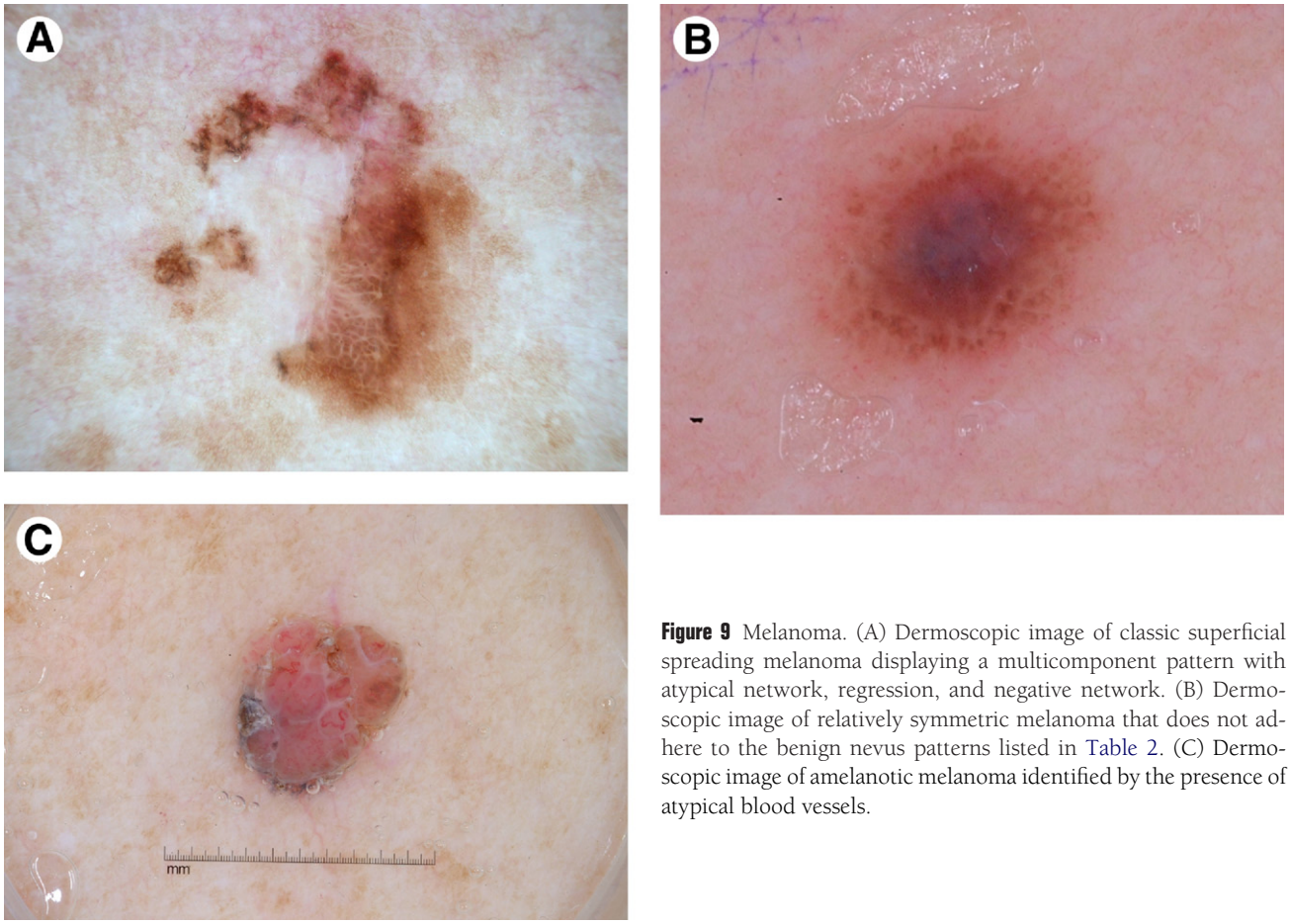

Figure 9 Melanoma. (A) Dermoscopic image of classic superficial spreading melanoma displaying a multicomponent pattern with atypical network, regression, and negative network. (B) Dermoscopic image of relatively symmetric melanoma that does not adhere to the benign nevus patterns listed in Table 2. (C) Dermoscopic image of amelanotic melanoma identified by the presence of atypical blood vessels.

irregularly shaped, sharply bordered depressions with red dots or globules linearly aligned along the acral furrows (Fig. 11B).[33] Histologically, red dots/globules represent the blood vessels in the underlying papillary dermis. These blood vessels are usually not visible in nonsyndromic patients due to the dense keratin layer of volar skin, however; pits in BCNS lack this dense keratin layer, allowing one to see the papillary dermal vasculature.[33] Dermoscopy has become useful for monitoring treatment response to imiquimod therapy. Leaf-like areas and spoke wheel areas have been shown to regress within several weeks, whereas blue-gray ovoid nests disappear only after a couple of months. The disparity in response rates likely reflects the depth of the BCC tumor islands: spoke wheels are seen in superficial BCCs and ovoid nests represent BCC tumor islands at a deeper histologic level.[37] It has been shown that complete disappearance of BCC-specific dermoscopic patterns is a good indicator of response to local treatment.[38] By contrast, early recurrence of BCC or lesions resistant to treatment can be identified by the degree of regression of dermoscopic features or by the reappearance of BCC-specific structures.[37-39]

Xeroderma Pigmentosum

Xeroderma pigmentosum (XP) is a rare autosomal-recessive disease characterized by marked photosensitivity and an in-

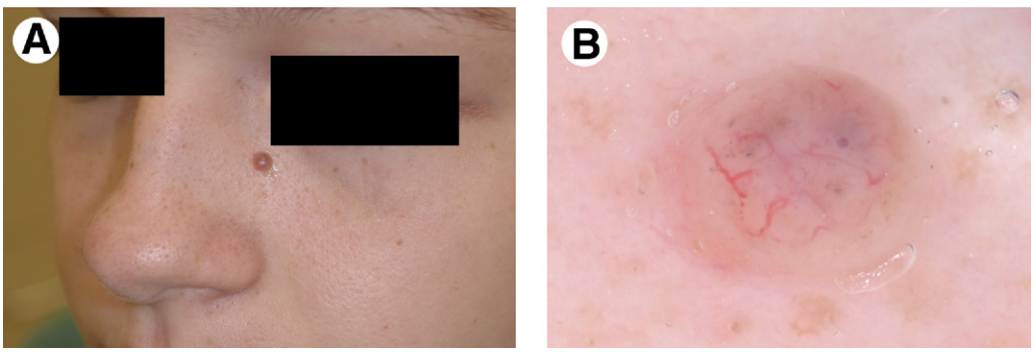

Figure 10 BCC. (A) Clinical image. (B) Dermoscopic image showing arborizing vessels highlighted.

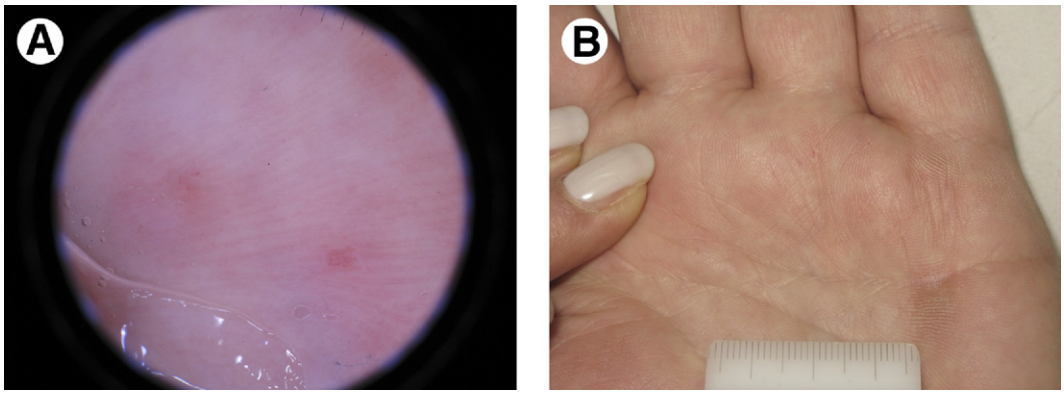

Figure 11 Acral Pit. (A) Clinical image. (B) Dermoscopic image of the pit reveals linear red dots in the furrows.

creased risk of early-onset skin cancer. Malvehy et al investigated the use of dermoscopy in 2 siblings with XP, specifically to study the dermoscopic patterns of benign and malignant lesions in these patients.[40] Skin tumors in XP patients exhibited dermoscopic patterns similar to those found in nonsyndromic patients, see Tables 2 and 3.[41] Dermoscopy was advantageous in these patients because it helped differentiate malignant neoplasms from background actinic damage, such as solar lentigenes and poikiloderma (Fig. 12).[41]

Epidermodysplasia Verruciformis

Epidermodysplasia verruciformis (EV) is a rare autosomal-recessive genodermatosis characterized by persistent human papillomavirus (HPV) infection and susceptibility to squamous cell carcinoma.[40] Patients typically present in childhood with flat, verrucous lesions on the face and extremities and hypopigmented macules and papules on the trunk resembling Pityriasis versicolor.[40]

Segura et al were the first to report the dermoscopic features of EV lesions.[42] They described patients with various growths that were accompanied by histologic changes compatible with EV namely keratinocytes infected with HPV. Dermoscopy of the *Tinea versicolor*-type lesions revealed a hypopigmented and erythematous area with a whitish, scaly surface. The pathology of this lesion revealed "an acanthotic epidermis with a prominent granular layer with dense kerato-hyaline granules and keratinocytes with clear cytoplasm and perinuclear halos" (Fig. 13).[42]

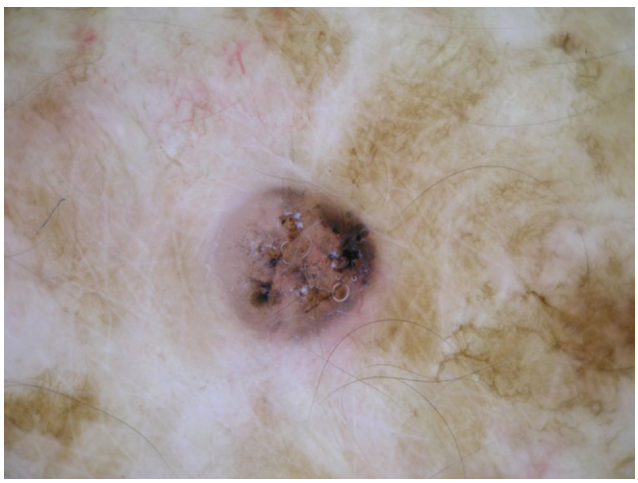

Figure 12 Xeroderma pigmentosum. Pigmented BCC in a patient with XP.

Epidermolysis Bullosa Hereditaria

Patients with any form of epidermolysis bullosa (EB) may develop large atypical melanocytic nevi, termed "EB nevi."[43] These nevi manifest clinical and dermoscopic features suggestive of melanoma, including asymmetry, irregular borders, heterogeneous color, and regressive changes.[43-45] EB nevi are dynamic in nature and appear to change color, enlarge rapidly, and give rise to small satellite nevi surrounding the primary nevus.[44,46] They tend to form in sites of previous bullae or erosions on the extremities or overlying bony prominences.

In a recent study of EB nevi, digital dermoscopy was shown to be useful in differentiating EB nevi from melanoma.[47] EB nevi frequently demonstrate a multicomponent pattern consisting of randomly distributed dots and globules (corresponding to nevus cell nests), an atypical pigment network, and structureless areas, each of which can also be seen in melanoma (Fig. 14). Occasionally, they may also reveal milia cysts.[46] However, these nevi usually lack any of the other melanoma specific structures listed in Table 3.[48]

In a 20-year prospective study of 86 EB nevi, none progressed to melanoma. Although no malignant transformation of an EB nevus has been reported to date, the number of EB nevi studied so far is not large enough to completely rule out the possibility.[44]

These nevi may be monitored by the use of baseline clinical and dermoscopic photography. Any concerning changes in these lesions can then result in further evaluation through confocal microscopic evaluation or biopsy. Moreover, dermoscopy may help select sites for small incisional biopsies within large EB nevi. Given that patients with EB tend to have sensitive skin and possibly impaired wound healing, dermoscopic selection of biopsy sites may be a preferred method

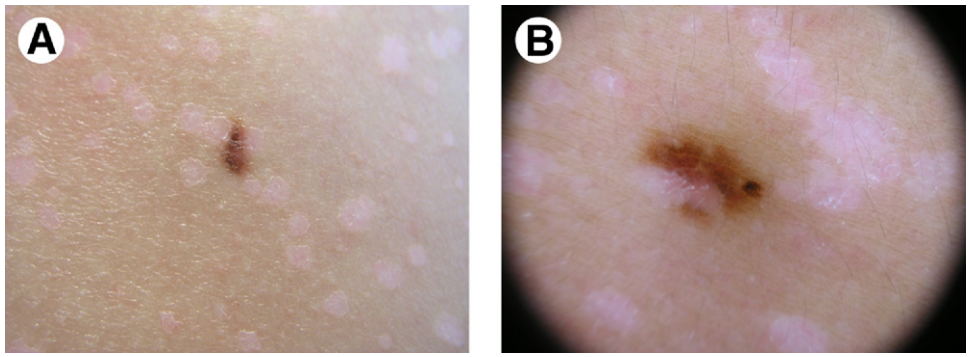

Figure 13 Epidermodysplasia verruciformis (EDV). (A) Clinical image of *Tinea versicolor*-like lesions and a nevus. (B) Dermoscopic image of benign nevus in a background of whitish, scaley macules. These macules are caused by HPV infection in a patient with EDV. They can sometimes be misdiagnosed as *tinea versicolor*.

over prophylactic total excision, which may create larger and more slowly healing wounds in these patients.

Of note, the overall disorganized architecture seen under dermoscopy might be explained by 2 main theories of EB nevogenesis. The first purports that repetitive disruption of the basement membrane may prime local nevus cell nests to halt senescence and begin proliferation.[45,47] Stress of the skin tissue may be an initiating event in tumorigenesis in general.[49] The second theory suggests that, within an EB blister, there exists a fluid-filled cavity containing free-floating, viable melanocytes that have detached from preexisting nevi due to defects in adhesion proteins.[50] These melanocytes may be stimulated to proliferate and develop into "EB nevi."[45,50-52]

In addition, squamous cell carcinoma is a well-documented complication in patients diagnosed with recessive dystrophic EB and less commonly junctional EB.[53-56] Cutaneous squamous cell carcinomas are recognized under dermoscopy by exhibiting glomerular vessels and a scaly surface.[57]

Conclusions

The art of pediatric dermatology requires a physician to be clinically attuned to pediatric skin disease as well as aware of parental and patient anxieties. A dermatoscope poses neither physical nor emotional harm and serves as the ideal looking glass to examine a child's skin. The visual information afforded by dermoscopy equips the clinician with an enhanced diagnostic capability to help confirm a suspected diagnosis or prompt other diagnostic testing.

The evaluation of melanocytic neoplasms in children is important given the increasing incidence of melanoma in this population. Dermoscopy can facilitate the diagnosis of melanoma while it is still thin and more readily curable. A multifaceted approach increases the sensitivity and specificity for diagnosing melanoma, and includes a thorough clinical history, visual examination, and dermoscopic evaluation of suspicious skin lesions. The decision whether to observe a clinically atypical lesion or to perform a biopsy can often be made with improved confidence based on the presence or absence of various features observed on dermoscopic evaluation. While there is overwhelming evidence demonstrating the benefits of dermoscopy (meta-analysis showed that the diagnostic accuracy for melanoma increases 49% when dermoscopy is used), it is only used by 20%-25% of dermatologists in the United States. A better understanding of the utility of dermoscopy combined with increased education during medical training is likely to transform dermoscopy into a more widely used tool. Like a stethoscope, it may become standard equipment among primary care physicians, medical specialists, and those in medical training. Dissemination of dermoscopy among medical professionals would likely have a significant impact on the early detection and curative treatment of melanoma.

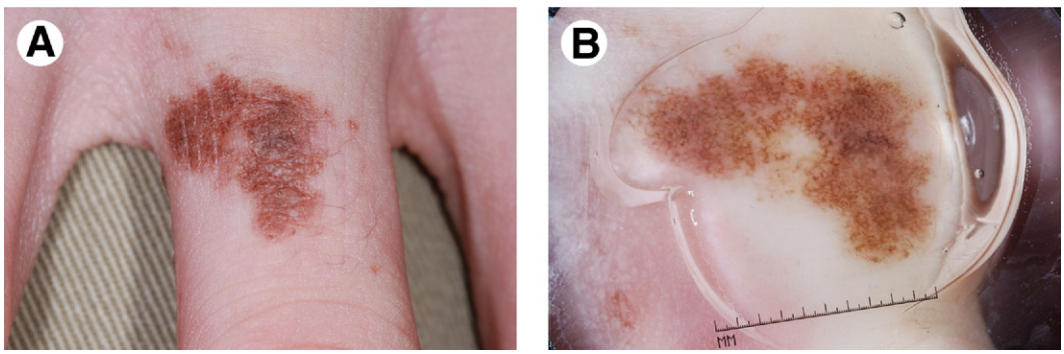

Figure 14 EB nevus on the abdomen. (A) Clinical image. (B) Dermoscopic image of EB nevus showing irregular reticulation, globules at the periphery, and bluish-white veil (asterisk).

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