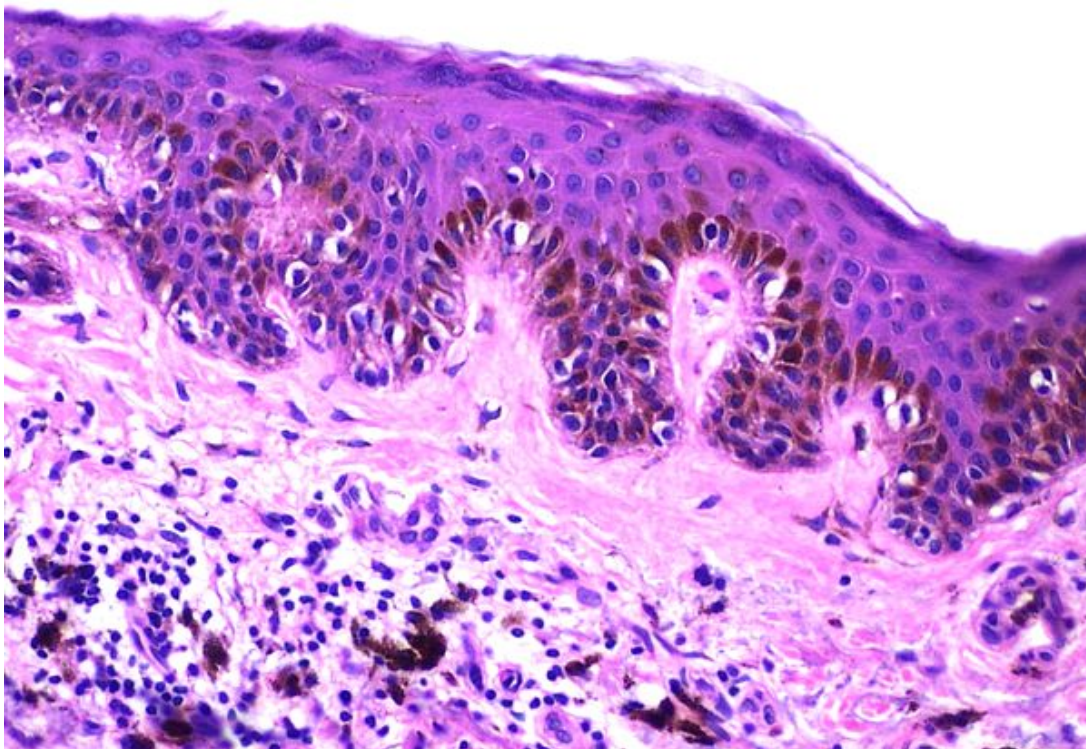


## Pigmented Skin Lesions in Children (Melanocytic Nevus, Clonal Nevus and Halo Nevus) — Symptoms and Treatment

[See online here](#)

**Benign pigmented skin lesions are very common in children and can be generally classified into congenital or acquired nevi. Congenital nevi can be small, medium or large. Large nevi are usually associated with a higher risk of malignant transformation into melanoma. Acquired melanocytic nevi can be classified into typical melanocytic nevi, clonal nevi, or halo nevi. Cytological atypia can be observed in congenital and acquired nevi and is associated with an increased risk of malignancy. Large nevi or nevi with cytological atypia should be completely excised to lower the risk of malignant transformation.**



### Overview

Nevi are brown colored flat or elevated skin lesions and can be seen anywhere on the body.

Pigmented skin lesions in children are quite common and can be **classified into malignant or benign skin lesions**. Malignant melanoma is rare in children.

**Pigmented Melanocytic nevi are hamartomatous collections of melanocytes** which are derived from the epidermal melanoblasts or dermal Schwann cells. These melanocytic nevi can be either congenital or acquired after birth.

**A clonal nevus is a benign condition that is characterized by focal proliferation of pigmented dermal melanocytes**. The main distinction between this type of nevi and the typical melanocytic nevi is the fact that the latter arises from the epidermal melanocytes and are usually multiple.



An example of a Halo Nevus (mole with white 'halo' around it), zoomed in.

**A halo nevus is a typical pigmented melanocytic nevus that is surrounded by an area of skin hypopigmentation**, hence the term halo.

Finally, an **atypical melanocytic nevus is a condition that is characterized by the presence of cellular atypia within the nevus**. These dysplastic nevi carry an increased risk of malignant transformation into melanoma.

## Epidemiology of Pigmented Skin Lesions in Children

**Congenital melanocytic nevi are very common** and can be identified in up to 1% of newborns. On the other hand, acquired melanocytic nevi usually start appearing after the first year of life and peak in number during the second or third decades of life.

Large congenital melanocytic nevi are of special interest because of their association with an increased risk of malignant transformation of melanoma. The estimated incidence of large congenital melanocytic nevi in newborns is around 1 in 20,000. If malignant transformation happens, it usually occurs during childhood.

**Other types of nevi, such as clonal nevi and halo nevi, are usually acquired after birth**. Halo nevi are usually solitary lesions, but multiple halo nevi have been seen in up to 25% of the patients.

The prevalence of atypical melanocytic nevi in the general population is estimated to be around 7%, but this figure can be as high as 59% in patients with confirmed malignant melanoma.

# Pathophysiology of Pigmented Skin Lesions in Children

**The exact trigger for congenital or acquired melanocytic nevi formation is largely unknown.** One possible hypothesis suggests that a pigmented nevus first forms from epidermal melanoblasts within the epidermal layer and then drops off into the dermis. Other hypotheses suggest that pigmented nevi form within the dermis and then migrate upwards into the epidermis.

Despite our poor understanding of the etiologies and the pathogenesis of congenital or acquired melanocytic nevi, we could identify several risk factors for the development of an increased number of pigmented nevi. **The most commonly encountered risk factors in children are prolonged sun exposure, the use of cortisone, corticotropin, growth hormone or chemotherapy and immunosuppression.** Studying pigmented nevi is important because their number in the body and their size positively correlate with the risk of malignant melanoma.



**Image:** "5-day-old girl (head to right) with giant congenital melanocytic nevus over back, neck, buttocks and left thigh. Histopathological findings showed nevomelanocytes throughout the thickness of the dermis and extending into the subcutaneous tissue. No other complications were reported." by S. Sharma , N.L. Sharma, V. Sharma - Giant Bathing Trunk Naevus with Multiple Congenital Melanocytic Naevi. The Internet Journal of Pediatrics and Neonatology. 2012 Volume 14 Number 1. DOI: 10.5580/2aa3. License: [GFDL](#)

Large congenital melanocytic nevi are of special interest to us because they carry a significantly higher risk of malignant transformation compared to smaller nevi and because of their association with the central nervous system involvement.

Clonal nevi are at a borderline condition between truly malignant melanoma and benign pigmented skin lesions. Clonal nevi usually carry p53 mutations in their core melanocytes.

Recent studies on halo nevi aimed to address two important questions. Why this type of nevi is associated with a higher risk of malignant transformation and why they have a halo appearance. The reason for the skin hypopigmentation around the nevus is believed to be the consequence of an immune response to the proliferating melanocytes. Activated cytotoxic T-cells are believed to release a cytotoxic substance that destroys the melanocytes around the forming nevus.

**The presence of atypical melanocytic nevi is indicative of a complete skin examination to rule out the presence of malignant melanoma.**

## Clinical Presentation of Pigmented Skin Lesions in Children



**Image:** "Congenital melanocytic nevus. Brown papule on the nose, which developed shortly after birth. The brownish exophytic lesion is well circumscribed." by M. Sand, D. Sand, C. Thrandorf, V. Paech, P. Altmeyer, F. G. Bechara - M. Sand, D. Sand, C. Thrandorf, V. Paech, P. Altmeyer, F. G. Bechara: Cutaneous lesions of the nose. In: Head & face medicine Band 6, 2010, S. 7. License: [CC BY 2.0](https://creativecommons.org/licenses/by/2.0/)

Newborns who are born with a congenital melanocytic nevus usually present with small nevi that can be missed. Large and intermediate-sized congenital melanocytic nevi are usually easy to identify and **present with dark-colored or pigmented skin lesions over the face, trunk, or extremities**. These lesions are usually present at birth.

The diameter of the pigmented skin lesion should be determined because of the clear association between the size of the nevus and the risk of malignant transformation. **Small nevi usually have a diameter less than 1.5 cm, medium nevi have a diameter between 1.5 and 19.9 cm and any nevus that is above 19.9 cm is considered as large.** The risk of malignant transformation in small and medium-sized nevi is believed to be like baseline risk of malignant melanoma.

Infants who have seizures might have neurocutaneous melanosis if they have a large congenital melanocytic nevus. The most common cutaneous sites for congenital nevi in children with central nervous system involvement are the head, neck, and face.

Acquired melanocytic nevi usually have different clinical features from congenital nevi. They usually have a verrucous surface and can be pedunculated. Clonal nevi usually appear light brown to blue or black in color and usually are well-demarcated and focal. Halo nevi are usually solitary and have an area of skin depigmentation around them.

The presence of cytological atypia within a nevus cannot be confirmed with the appropriate histological examination and molecular testing. However, certain risk factors increase the risk of cellular atypia in acquired nevi such as the presence of malignant melanoma, or the presence of an abnormally looking nevus that changes over time in size and character.

# Diagnostic Workup for Pigmented Skin Lesions in Children

When a child presents with a pigmented skin lesion, the main goal of our approach should be to **determine the malignant potential of the lesion**. The most important risk factors for malignant transformation are the size and number of nevi.

In addition to the clinical features of congenital and acquired nevi, **certain advanced diagnostic studies can be used to confirm the diagnosis of a benign skin lesion over a malignant melanoma**.



Polarized light dermatoscope.

Dermatoscopy can reliably differentiate between benign pigmented nevi and malignant melanoma. Benign pigmented nevi usually have a uniform and regular network of pigmentation, do not have a blue or white veil, rarely have vascular patterns, do not have a regressive center, and can be cystic. On the other hand, malignant melanomas usually have a dark and irregular network of pigmentation, can have a blue or a white veil, have areas of regression and scarring, and might have irregular vascular patterns. A cystic formation is not seen with malignant melanomas.

When a biopsy is obtained from a clonal nevus, the nevus is usually found to be located within the dermal layer. The focal proliferation of dermal melanocytes is usually observed. Melanin content is usually increased within the epidermal melanocytes and keratinocytes which explains the dark color of these nevi. These lesions usually do not show any dysplastic features or cytological atypia. Halo nevi usually have lymphocytic and histiocytic infiltrates on microscopic examination.

Children with large congenital melanocytic nevi should undergo brain and spine magnetic resonance imaging to exclude central nervous system involvement.

## Treatment of Pigmented Skin Lesions in Children

**Small congenital melanocytic nevi do not require any specific treatment** except for close monitoring for changes in the lesions' characteristics. Any lesions that develop asymmetry, their border become irregular, change in color or increase in diameter should be excised and a biopsy should be obtained for histologic examination.

Prophylactic lesionectomy is usually recommended for large or medium-sized pigmented nevi in children. Large scalp pigmented nevi should be removed during infancy if possible. Delayed surgical intervention is associated with an increased risk of recurrence, scarring, and poor cosmetic results.

**It is recommended to completely remove clonal nevi despite being benign.**

Children with halo nevi should be provided with a full-body examination because higher numbers of halo nevi are clearly associated with an increased risk of malignant melanoma.

Atypical nevi that are changing in color, size, shape or border should be completely excised and a histologic examination should be performed to assess the degree of cytological atypia and the presence of molecular abnormalities such as p53 mutations. Small atypical nevi with minimal atypia might be excised without a free margin or might be followed-up closely looking for any changes.

## Prognosis of Pigmented Skin Lesions in Children

Halo nevi are benign with very low chances of morbidity. The only concern is cosmetic appearance.

Melanoma can develop from atypical nevus but chances are as low as 2 per 200,000 population. Risk of developing melanoma is higher in multiple atypical nevi lesions as compared to single or few lesions. Still, the personal family history of melanoma is more predictive than the number of nevi.

## Prophylaxis of Pigmented Skin Lesions in Children

Patients with nevi should be educated to avoid exposure to sunlight, UV tanning beds and are advised to use sunscreens to minimize harmful effects of UV rays,

Patients and family members should be educated to monitor the lesions regularly and should be guided to report in case of severe abnormal symptoms like a sudden increase in the size of the lesion, bleeding, itching, and ulceration.

## References

Russak JE, Dinulos JGH. Pigmented Lesions in Children. *Seminars in Plastic Surgery*. 2006;20(3):169-179. doi:10.1055/s-2006-949120

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