

## Linearly Arranged Pigmentary Cutaneous Pattern in a Child, Segmental Vitiligo vs. Hypomelanosis of Ito

**Yordania Velázquez Avila\***, Maritza Morales Solís, Recaredo Rodríguez Gundin

Provincial Teaching Pediatric Hospital "Martyrs of the Tunas", Las Tunas's University of Medical Sciences, Cuba

**\*Corresponding Author:** Yordania Velázquez

Avila, Provincial Teaching Pediatric Hospital "Martyrs of the Tunas", Las Tunas's University of Medical Sciences, Cuba.

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### Abstract

**Introduction:** Some dermatological diseases are still available with linear patterns. With hypopigmentation in childhood are the segmental vitiligo, which follows the dermatomes, but can follow the lines of Blaschko and hypomelanosis of Ito, which in turn follows the lines of Blaschko. These dermatoses are uncommon in dermatology practice.

**Objective:** To deepen the diagnostic elements that make it possible to differentiate two clinically characterized dermatoses by linear segmental hypopigmentation of blaschkoid type and treatment.

**Case Presentation:** The consultation of Genodermatosis in Las Tunas comes a child with achromic macules in left Hemibody, without other alterations. After being evaluated by several specialties (dermatology, genetics, pediatrics, ophthalmology and neurology), it is determined that only cutaneous involvement was performed, skin biopsy was done, which corroborated the diagnosis of segmental vitiligo.

**Conclusions:** The case is presented because segmental vitiligo is uncommon, follows a linear pattern that may be differentiated from another uncommon dermatosis, such as the hypomelanosis of Ito, and in the treatment it is important to provide support Psychological to the patient in order to obtain better results with the Melagenina Plus.

**Keywords:** Cutaneous Mosaicism; Vitiligo Hypopigmentation, Hipomelanosis Of Ito, Blaschko Lines

### Introduction

In dermatological practice it is common to find dermatoses in which the elemental lesions are distributed according to linear disposition patterns. Within the dermatoses that follow the linear distribution are those that affect traumatized or scar areas, called isomorphic phenomenon of Köebner, which follow Metameras nervous zosteriform type, which affect Dermatomes and Cutaneous mosaicismos [1]. Within the cutaneous mosaicism are those that follow the lines of Blaschko, of narrow bands or wide bands, in Chessboards, filodes, in patches or lateralized pattern [2]. In pediatric ages, within hypopigmented dermatoses presenting linear disposition patterns, segmental vitiligo and Ito hypomelanosis are found [3].

Vitiligo is an idiopathic acquired disease, which is manifested clinically by the progressive emergence of acrómicas macules [1] is a disease Degenerative skin in which melanocytes die; Consequently, it ceases to produce melanin in the area where cell death has occurred [4].

The word vitiligo seems derived from the Greek vitelius, which means calf, by the similarity of the white spots of this animal with the white spots of the vitiligo [5]. It has been reported since ancient times; there is a description in the classic Latin medical medicine Celsus during the second century. In studies based on the general population, vitiligo occurs in less than a half percent of the child population [6]. It is classified into non-segmental vitiligo and segmental vitiligo [5].

Segmental vitiligo involves only one segment of the body. It usually begins at early childhood, and is often associated with white hair [7]. It represents 10 to 15% of all types of vitiligo. It is defined by its unilateral and segmental distribution, or in the form of asymmetric bands of Acromic Macules. A single segment is generally involved, but two or more segments with IPSI or contralateral distribution have been described [6].

This is a multifactorial disease, in which certain factors act on genetically predisposed people [1]. Although there are 3 hypotheses to explain the pathogenesis of vitiligo: autocytotoxic, neurochemical and autoimmune theory [5]. In the segmental vitiligo usually occurs with a dermatomal pattern [8], although it can be disposed following the lines of Blaschko [9]. This observation leads to the neural hypothesis that proposes certain chemical mediators released from nerve endings, which can cause a decrease in the production of melanin [8]. Although the diagnosis is clinical, when performing histopathological study regardless of the mechanism, when using special stains for melanin, such as silver or dopa, it is observed that well established lesions show that there are no melanocytes [5].

The psychological impact of vitiligo is profound in childhood. Associated negative experiences may include fear of being questioned about one's own appearance, mockery and harassment, anxiety about the possibility of spreading the disease, interference with emotional maturation, depression, and interference with Socialization [6]. In these patients it is very helpful to use cognitive therapy, in which the main objective is the change of negative thoughts by others more adaptive [10].

In the world, for the treatment of recent onset vitiligo, improvement has been achieved with phototherapy and topical tacrolimus [7]. These therapies are not available in all areas of health in Cuba, however, for their treatment has been used Melagenina Plus.

It is a topical medicine, which was obtained as a result of the investigations carried out during the years 1968 to 1970 by the doctor Carlos Miyaes Cao. It isolates from placental tissue the Bioestimulina BIOPLA RE-761, which is a placental melanocitopoyético factor of protein nature, capable of producing stimulation of the production of melanocytes [5], and it was demonstrated in studies carried out, which has been effective in 86% of the cases treated [11].

The hypomelanosis of Ito refers to a set of neurocutaneous phenotypes in which there are hypopigmented macules that follow the lines of Blaschko, which can be associated or not to neurological defects, malformations and chromosomal anomalies. This disease is usually evidenced at birth. In clinical terms it is distinguished by hypopigmented macules, linear distribution or annular, following the lines of Blaschko-usually the trunk and the Extremities-without affecting the palms, plants and mucous, bilateral and symmetrical [12]. Three-quarters of the affected individuals present association with central nervous system anomalies, eyes, hair, teeth, skin, fingernails, musculoskeletal system, internal organs, including polycystic renal disease. Patients may manifest psychomotor or mental deterioration, autism, microcephaly, coarse facies, and dismórficas ears [13,14]. At the global level, the incidence is estimated at 1 x 8 000-10 000 inhabitants [14].

Blaschko lines are a linear cutaneous pattern described by Alfred Blaschko, German dermatologist, who, in the year 1901 presented at the 7<sup>th</sup> Congress of the German Society of Dermatology, his observations on a rare dermatological condition, which is distributed following some fairly defined patterns: V-shape in the upper part of the Back, with its vertex in the middle and downward; S-shape italicized in the abdomen; Linear shape in extremities; Or inverted, if you follow your path from the thorax; And, circular shape that flows into the nasal root in the face [15].

It is now known that Blaschko lines do not correspond to any underlying anatomical structure, whether arterial, venous, lymphatic or neural; Neither with the embryonic fusion lines, or with the dermatomes, but are those strokes determined by the migration of cutaneous cells during embryogenesis from the neural crest, and corresponds to Mosaicism cutaneous pigments [2].

Diagnostic criteria for Ito Hypomelanosis proposed by Ruiz-Maldonado (1992):

- **Fundamental criterion:** Existence of linear or patched cutaneous macules, of congenital character or of non-hereditary, very early acquisition, that extend by two or more parts of the body.
- **Higher criteria:** One or more abnormalities of the central nervous system, or one or more abnormalities of the musculoskeletal system.
- **Minor criteria:** Two or more congenital malformations outside the central nervous system, or musculoskeletal, and chromosomal anomalies.
- **Definitive diagnosis:** Fundamental criterion, plus one or more higher criteria, or 2 or more minor criteria [12].

Although skin biopsy is not diagnostic in the histopathological study with Hematoxylin-eosin staining, it may show skin that presents areas with melanic pigment reduction in basal cells along with normal areas [12]. Normal karyotype in blood does not rule out the existence of chromosomal anomalies [16].

Treatment requires multidisciplinary intervention, including dermatologists, neurologists, geneticists, ophthalmologists and Traumatologists, among others [12]. Periodic follow-up from the pediatric stage is important, and in this way it is possible to detect anomalies in time that can be corrected with early surgery or physical rehabilitation, depending on the characteristics of each complication. Treatment of skin blemishes is generally not specific, although the dermatologist advises sunscreen to reduce the risk of complications from sun exposure [17].

### Objective of the Study

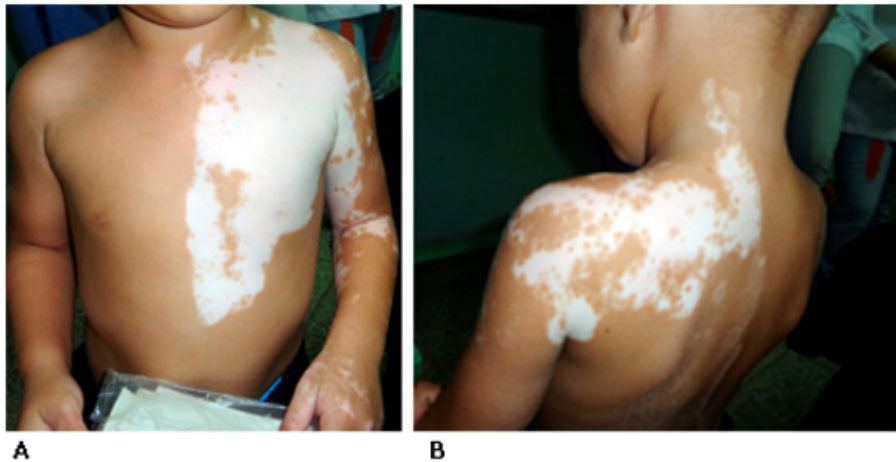
The objective of this presentation has been to deepen the diagnostic elements that make it possible to differentiate two clinically characterized dermatoses by linear segmental hypopigmentation, of the Blaschkoide type, and the treatment.

### Case Presentation

Male patient, 4 years old, skin Phototype II, of urban origin, with a history of good health, who from 3 years began to present white macules on the left side of the Hemithorax, initially small. It was valued by the dermatologist in his health area, and was treated with Melagenina Plus, with a discreet response and repigmentation. After about 6 months, after a depressive state, the response to the use of Melagenina stopped, and the macules continued to grow to converge in a large macula that occupied much of the anterior and posterior hemibody, as well as the upper left extremity. It was asymptomatic and not accompanied by other alterations or anomalies. With this painting it was referred to genetics of its area of health, and of this, to the consultation Multidisciplinary of Genodermatosis in the Tunas (dermatology, genetics, pediatrics, ophthalmology, orthopedics, neurology and psychology) with a probable diagnosis of hypomelanosis of Ito. Despite not showing extracutaneous manifestations or anomalies, we proceeded to perform ophthalmological examination, histopathological study and evaluation of the diagnostic criteria.

- Personal pathological antecedents: They did not refer.
- Family pathological antecedents: They did not refer.
- Reaction to drugs: They did not refer.

- Dermatological Examination: Monomorphic cutaneous picture, located in left Hemibody, including anterior and posterior region, in addition to upper left extremity, given by macula achromic with signs of repigmentation with skin color macules in the Interior of the macula acromic (Figure 1).
- Neurological examination and psychomotor development: Normal.
- Ophthalmological Examination: Normal Eye fund, without retinal alterations.
- Musculoskeletal exam: Normal.
- Skin biopsy: (17-7-11), which throws to the microscope, absence of some melanocytes in the basal layer, and inflammatory infiltrate lymphocyte perianexial. Informa: Vitiligo.



**Figure 1A and 1B:** Patient with segmental vitiligo. Notice you that it shows repigmentation signs with the use of the Melagenina Bonus, inside the stain acromic.

With the histopathological diagnosis, and taking into account the disposition of the macula and the absence of diagnostic criteria of Ito hypomelanosis, a segmental vitiligo was diagnosed.

Prior to retaking treatment with Melagenina Plus, cognitive psychotherapy was considered by psychology. Its implementation favored the patient assuming more positivist skills, and retreated with Melagenina Plus, and, without doubt, the results were more favorable.

## Discussion

It is a child who begins with hypopigmented macules showing a linear pattern located in left hemibody, which made think of the possibility of two pigmentary dermatoses appearing in childhood: segmental vitiligo and hypomelanosis of Ito, both dermatoses are uncommon in dermatological practice, if it is taken into consideration that the incidence of vitiligo worldwide is 2% of the population [8], and the Segmental form represents 10 to 15% of all types of vitiligo [6]; And in the case of Ito hypomelanosis, it is estimated that the worldwide incidence is 1 x 8000 - 10000 inhabitants [14].

Among these 2 dermatoses the common point is the Hipocromic macules that follow a linear pattern, but in the segmental vitiligo the disposition is following the dermatomes. Although it can follow the lines of Blaschko, it affects only the skin without other alterations,

and in the histopathology there is absence of melanocytes [5-7]. In contrast, the hypomelanosis of Ito usually occurs at birth, or at very early ages, usually appears associated with other anomalies, although they can occur without extracutaneous anomalies. It follows the pattern of the lines of Blaschko, tends to grow and then to stop, and in the Histopathological study are present the melanocytes, but there is reduction of the pigmentation [12,14,16].

In the present case, there were hypopigmented macules, of type acromic, following a linear pattern, without other alterations, with histopathological study that showed absence of melanocytes in the basal layer. It supported the diagnosis of segmental vitiligo its therapeutic response to the use of Melagenina, which stimulates melanocitopoyesis. The fact of presenting a macular disease that aesthetically affected the skin affected the psychological sphere of the child, and this could explain the decrease in the response in the first period of treatment with Melagenina; However, the use of cognitive psychotherapy favored the patient assuming positive aptitudes for his illness, and improved the response to treatment with Melagenina.

This study has coincided with the diagnostic criteria of segmental vitiligo posed by other authors, based on Hipocrómicas or segmental acrómicas macules, and the absence of melanocytes in Histopathology [6,7]. By comparing the criteria used by García F Aparicio In the presentation of a case hypomelanosis of Ito without extracutaneous involvement, which was characterized only by the presence of the Hipocrómicas macules [16]. The results of the histopathological study did not coincide with the present case, which reaffirmed the diagnosis of segmental vitiligo. In relation to the use of Melagenina Plus and the combination with psychotherapeutic techniques of cognitive therapy, the results were encouraging, coinciding with authors who support the use of these therapies [10,11].

### Conclusion

It is concluded that the case arising is uncommon, because segmental vitiligo follows a linear pattern that may be differentiated from another uncommon dermatosis, such as Ito hypomelanosis, and in the treatment it is important to provide psychological support to the patient To promote better results with Melagenina Plus.

### Conflict of Interest

The authors declare that they have no conflict of interest in the conduct of the study. The personal information on the patient is not included in the presentation by ethical reasons.

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