



HYPOMELANOSIS OF ITO

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Hypomelanosis of Ito, also known as incontinentia pigmenti achromians, is a rare disorder characterized by a whorled pattern of light patches on the skin. Abnormalities of other organ systems are often seen in conjunction with the skin findings, particularly the central nervous system. This condition is present from birth but may not be recognized until the first or second year of life. Females are more often affected than males. Sporadic gene mutations are believed to be the cause of hypomelanosis of Ito; therefore, it is not considered an inherited disorder. The specific genes involved have not yet been identified.

The skin findings associated with hypomelanosis of Ito include small patches of pale or white skin that merge to form larger, asymmetric patches arranged in whorled or linear patterns. These patterns follow the **lines of Blaschko** which represent a developmental pattern of the skin. The pale or hypopigmented areas can be found in any part of the body including the head, face, neck, trunk and extremities. Typically a v-shape pattern is seen over the face and spine, an s-shape pattern is seen over the trunk, and a linear pattern is seen over the extremities. Additionally, it is important to note that the hypopigmentation is not preceded by vesicular or verrucous stages as this is a hallmark of another disorder that appears similar to hypomelanosis of Ito, known as incontinentia pigmenti.

Hair abnormalities can also be seen with hypomelanosis of Ito and can include changes in hair color, hair breakage and/or loss as well as hair with a white-gray color.

Individuals with hypomelanosis of Ito may have other anomalies including birth defects (cleft palate, or limb, hand, foot or face abnormalities), mental retardation and seizures most commonly. Other associated features of the condition include developmental delay, deafness, musculoskeletal defects, heart abnormalities, kidney problems, and abnormalities of the eyes and teeth.

Hypomelanosis of Ito can be diagnosed by a dermatologist based on clinical findings. The use of a Wood's lamp can help enhance the pattern of hypopigmentation. A **biopsy**, as well as genetic testing may also be performed to help aid in confirmation of the diagnosis. Due to the association with central nervous system and eye abnormalities, neurologic and ophthalmologic exams should also be performed. Other consultations and imaging should be performed based on the patient's history.

Currently there is no treatment available for the skin findings of hypomelanosis of Ito. Make up can be used to cover up the pale or white areas. Over time, skin pigmentation may develop and blend with the normal skin. Other associated abnormalities require appropriate medical treatment.

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