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Hypomelanosis of Ito

Hypomelanosis of Ito is also known as 'incontinentia pigmenti achromians'. It is considered to be a neurocutaneous syndrome.

What is hypomelanosis of Ito?

Hypomelanosis of Ito is a rare genetic disorder characterised by:

- Streaky, patchy, whorl-like, or linear hypopigmented macules occurring on any part of the body
- Palms, scalp and soles of the feet are usually not affected
- Swirling patterns around the trunk and linear patterns down the legs and arms are referred to as Blaschko's lines
- Lesions first appear as small 0.5–1 cm hypopigmented or white macules that merge to form larger patches
- Macules cover more than two dermatomes and are often on both sides of the body
- Patches are not symmetrical

Hypomelanosis of Ito



Associated abnormalities are thought to occur in 30–50% of patients with cutaneous lesions. These include:

- Seizures
- Mental retardation
- Hearing abnormalities
- Tooth or mouth problems
- Visual problems
- Orthopaedic problems

What is the cause of hypomelanosis of Ito?

Hypomelanosis of Ito is believed to be due to chromosomal mosaicism and sporadic mutations. It is not an inherited disorder as the chromosomal defect occurs after conception. The specific gene(s) involved has not been confirmed.

Some researchers suggest that hypomelanosis of Ito may not be a disease in itself, but a symptom of other

disorders.

How is the diagnosis made?

Hypomelanosis of Ito is present at birth and is usually picked up by a dermatologist, paediatrician or neurologist during the first or second years of life. Thorough history taking and physical examination with attention to neurological and ophthalmological findings are necessary to detect associated abnormalities.

Although recent research has estimated that 30–50% of patients with hypomelanosis of Ito have associated abnormalities, some studies report rates of associated neurological abnormalities as high as 75–94%.

What is the treatment for hypomelanosis of Ito?

The cutaneous effects of hypomelanosis of Ito do not require treatment. In many cases, the skin may develop pigment over time and blend in well with normal skin. Patients that are conscious of their appearance can use [cosmetic camouflage](#).

Associated abnormalities require appropriate medical treatment.

Related information

References:

- Book: Textbook of Dermatology. Ed Rook A, Wilkinson DS, Ebling FJB, Champion RH, Burton JL. Fourth edition. Blackwell Scientific Publications.

On DermNet NZ:

- [Skin pigmentation](#)
- [Birthmarks](#)

Other websites:

- [Hypomelanosis of Ito](#) – emedicine dermatology, the online textbook
- [HITS \(UK\) Family Support Network](#)

Books about skin diseases:

See the [DermNet NZ bookstore](#)

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DermNet does not provide an on-line consultation service.

If you have any concerns with your skin or its treatment, see a [dermatologist](#) for advice.

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