

# Teaching NeuroImages: Hypomelanosis of Ito

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A 2-year-old girl presented with developmental delay. Examination revealed height, weight, and head circumference above the 95th percentile, appendicular hypotonia, right hemibody hypopigmented lesions following the lines of Blaschko, and right hypopigmented iris (figure, A and B). EEG showed disorganized background. Genetic microarray was normal.

The skin lesions are characteristic of hypomelanosis of Ito. Presentation is variable, and behavioral problems and seizures are common.<sup>1</sup> Fifty percent have an IQ <80.<sup>2</sup> Males are often infertile.<sup>2</sup> X-chromosomal abnormalities, primarily mosaicism, have been reported.<sup>2</sup> Neuroimaging occasionally shows abnormalities contralateral to the skin lesion.<sup>2</sup> Differential diagnosis includes vitiligo, incontinentia pigmenti, tuberous sclerosis, and other phakomatoses.

## AUTHOR CONTRIBUTIONS

Aunali S. Khaku, MD: study concept and design, drafting and revising the manuscript, analysis or interpretation of data, guarantor of study. Vishnumurthy Shushrutha Hedna, MD: critically revising the manuscript for intellectual content. Anuranjita Nayak, MD: study concept and design, drafting and revising the manuscript, analysis or interpretation of data.

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## DISCLOSURE

The authors report no disclosures relevant to the manuscript. Go to [Neurology.org](http://Neurology.org) for full disclosures.

## REFERENCES

1. Assogba K, Ferlazzo E, Striano P, et al. Heterogeneous seizure manifestations in hypomelanosis of Ito: report of four new cases and review of the literature. *Neurol Sci* 2010;31:9–16.
2. Hsieh DT, Sarnat HB. Clinical summary: hypomelanosis of Ito. In: *Medlink Neurology* [online]. Available at: <http://www.medlink.com/medlinkcontent.asp>.

**Figure 1** Photograph showing unilateral hemibody hypopigmented whorled lesions following the lines of Blaschko pathognomonic for hypomelanosis of Ito (incontinentia pigmenti achromians)



The lines of Blaschko were described by Alfred Blaschko in 1901 after mapping 140 patients with linear skin lesions. The lines trace the migration of embryonic cells.

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