

# Bloch-Sulzberg syndrome

**Synonyms:** **Incontinentia pigmenti**, melanoblastosis Bloch-Sulzberg, Siemens-Bloch-Sulzberg syndrome, melanoblastosis cutis linearis sive systematica, naevus pigmentosus systematicus.

Named after Swiss dermatologist Bruno Bloch and American dermatologist Marion Baldur Sulzberg.<sup>[1]</sup>

## Genetics:

- OMIM: 308300 (<http://omim.org/entry/308300>)
- The disease is caused by a mutation in the gene of *IKBKG* (*NEMO*) in the Xq28 region.
- Inheritance: X-linked dominant, male lethal, variable expression in females (due to random X-chromosome inactivation).

## Characteristics:

- cinnamon dirty symmetrical skin spots on the sides,
- alopecia,
- dystrophy nails,
- defects of teeth,
- corneal opacity,
- microcephaly.



Deformation of teeth in a 3-year-old girl

## Links

## References

1. OLE DANIEL, Enersen. *Whonamedit - Bloch-Sulzberger pigment dermatosis* [online]. [cit. 2011-04-25]. <<http://www.whonamedit.com/synd.cfm/1762.html>>.

## References

- LAZOVSKIS, Ilmars – DOBIÁŠ, Václav. *Overview of clinical symptoms and syndromes*. 2. edition. Praha : Avicenum, zdravotnické nakladatelství, 1990. 0 pp. ISBN 80-201-0043-1.



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