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Biotin responsive dermatoses

What is Biotin?

Biotin is an essential micronutrient. It is one of the B vitamins and is also known as Vitamin H – 'H' stands for 'haut', the German word for skin. Biotin is found in many foods, and is made by bacteria, fungi, algae and certain plant species. There are particularly high amounts of biotin in brewer's yeast and in Royal Jelly. Human tissues do not make biotin, although it is made by the bacteria that live in the large intestine.

Biotin acts as a cofactor for the function of carboxylase synthetase enzymes. Carboxylases are enzymes that catalyze the addition of a molecule of carbon dioxide to another compound to form a carboxyl group, necessary in the manufacture of fatty acids. Biotin is also involved in making glucose, some amino acids and in energy production.

What are the biotin responsive dermatoses?

These are either acquired or inherited problems that resolve when extra biotin is taken.

Biotin deficiency can be acquired by:

- Prolonged ingestion of raw egg whites. Avidin, a glycoprotein in egg white, binds biotin specifically and tightly and prevents its absorption. Avidin is denatured by cooking so cooked egg whites do not cause biotin deficiency.
- Long term 'parenteral' nutrition given by intravenous infusion can also lead to biotin deficiency.

There are two rare inherited biotin responsive dermatoses.

- Holocarboxylase synthetase deficiency (diagnosed in neonates).
- Biotinidase deficiency (diagnosed in older infants)

Both have autosomal-recessive inheritance. This means two abnormal genes are required to cause disease, one coming from each parent. One-in-four of the parents' male and/or female children will be affected by the disease, but it is unlikely in other family members.

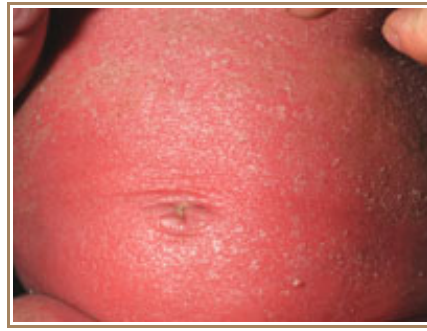
Holocarboxylase synthetase catalyzes bonding of biotin to the carboxylases. Deficiency of holocarboxylase synthetase results in a greater requirement for biotin and multiple carboxylase deficiency. This results in an accumulation of organic acids in the urine.

Biotinidase is required to salvage biotin for reuse during cellular turnover. It is also needed to release bound ingested biotin for absorption. A shortage of the enzyme results in a greater requirement for the vitamin than normal.

What are the clinical features?

The inherited conditions usually present in the first few weeks of life. The major features of biotin responsive dermatoses are rash and neurological problems. The rash is red and scaly and sometimes there are erosions. It develops over most of the body and may resemble [psoriasis](#), [seborrhoeic dermatitis](#) or [ichthyosis](#). It is especially prominent around the eyes, nose, mouth, ears and genitals. Alopecia ([hair loss](#)) and conjunctivitis (red eyes) may be present.

Baby with Holocarboxylase Synthetase Deficiency



Progressive severe neurological symptoms develop in the untreated inherited conditions. These include seizures, developmental delay, poor feeding and death.

For adults with acquired Biotin deficiency the rash is similar but neurological symptoms include hyperaesthesia (sensitive skin), paraesthesia (pins and needles), depression and muscle pain.

How are these conditions diagnosed?

Holocarboxylase synthetase deficiency and biotinidase deficiency are diagnosed in the neonatal and infant period respectively by finding high levels of organic acids in urine. Blood tests can then be done on white cells and plasma to measure enzyme levels to confirm the diagnosis.

Acquired biotin deficiency is diagnosed on clinical grounds and response to treatment.

What treatment is available?

A trial of biotin should be given in those with suspected or confirmed biotin deficiency. Usually a dose of 10mg–20mg per day will be adequate. Biotinidase deficiency usually responds rapidly and dramatically to as little as 10mg per day. Holocarboxylase synthetase deficiency may require much larger doses. There are reports of poor response even to 200mg/day.

Does biotin supplementation help any other skin conditions?

There are unconfirmed reports that biotin supplements may also be helpful for a rare congenital hair disorder called the 'uncombable hair syndrome', also known as 'spun-glass hair' and 'cheveux incoiffables'. Although it has been added to many cosmetic hair products, it is uncertain whether it is of any benefit for those with normal hair.

Biotin 300 micrograms daily may also be useful for thin, brittle fingernails and those with [lamellar splitting](#).

Biotin may be useful in at least some cases of [Leiner disease](#).

Related information

Other websites:

Emedicine dermatology, the online textbook:

- [Biotinidase Deficiency](#)
- [Biotin Deficiency](#)

Books about skin diseases:

See the [DermNet NZ bookstore](#)

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