DermNet NZ

Authoritative facts about the skin from the New Zealand Dermatological Society Incorporated.

Acrodermatitis enteropathica

What is acrodermatitis enteropathica?

Acrodermatitis enteropathica is a rare genetic disorder characterised by diarrhoea, an inflammatory rash around the mouth and/or anus, and hair loss.

What causes it?

Acrodermatitis enteropathica is due to malabsorption of zinc through the intestinal cells. The precise cause is not known but it may relate to mutations in a gene (SLC39A4) that codes the zinc transporter protein, ZIP4. It is thought that the missing protein may be responsible for decreased zinc uptake and abnormal zinc metabolism.

To have congenital acrodermatitis enteropathica you must inherit two defective genes (one from each parent) i.e. the inheritance is autosomal recessive. If an individual receives one normal gene and one defective gene, the person will be a carrier for the disease, but usually will not show symptoms.

Who gets acrodermatitis enteropathica?

Symptoms usually occur in bottle-fed infants within a few days or weeks after birth and breast-fed infants soon after weaning. Both males and females are equally affected.

Zinc deficiency may also rarely arise in adults. Causes include:

- Necrolytic migratory erythema (glucagonoma)
- Inadequate zinc in the diet (especially in alcoholics and previously, with intravenous nutrition)
- Intestinal malabsorption (inflammatory bowel disease, intestinal bypass surgery)
- Excessive urinary loss of zinc (nephrotic syndrome)
- Low levels of albulin and high catabolic states (trauma, burns, extensive surgery)

What are the clinical features?

Skin findings include:

- Red and inflamed patches of dry and scaly skin, particularly around body openings such as the mouth, anus, and eyes, and the skin on elbows, knees, hands, and feet. It may look like atopic dermatitis.
- Patches evolve into crusted, blistered, pus-filled and eroded lesions.
- There is usually a sharp demarcation between the affected area and normal skin.
- Skin around nails becomes inflamed (paronychia) and there may be nail ridging.
- Diffuse hair loss on the scalp, eyebrows and, eyelashes.
- Secondary infection with *Candida albicans* or *Staphylococcus aureus*
- Red glossy tongue and mouth ulcers
- Impaired wound healing

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Other features of acrodermatitis enteropathica include:

- Conjunctivitis
- Sensitivity to light
- Loss of appetite
- Diarrhoea, mild or severe
- Irritability (babies cry and whine incessantly)
- Depressed mood
- Growth failure

Zinc

Zinc is an essential component of the diet. Zinc in human milk is more absorbable than that from infant formulas or cow's milk, hence the later onset of acrodermatitis enteropathica in breast-fed babies compared to formula-fed babies. Zinc is also found in meat, shellfish and wheat germ. Foods of plant origin are mostly low in zinc. Phytates present in cereals and soy, and high levels of calcium, can reduce the absorption of zinc through the duodenum.

Zinc is needed to assist metalloenzymes that are involved in many cellular processes throughout the body. These include the production of anti-inflammatory agents (cytokines and antioxidants) and the normal functioning of the brain.

Investigations

If zinc deficiency is suspected, the following investigations may be helpful:

- Serum/plasma zinc levels confirm the diagnosis (normal levels are 10.7 - 23.0 µmol/L)
- Urinary zinc excretion may be reduced
- Blood count may reveal anaemia
- Skin biopsy may show characteristic features

What is the treatment?

The inherited form of acrodermatitis enteropathica was usually fatal until the role of zinc was discovered in 1973. It should be treated with 1 mg/kg body wait of oral zinc supplementation per day for life. Zinc gluconate is better tolerated than zinc sulfate. Zinc can be given during pregnancy.

After zinc replacement the skin lesions heal within one to two weeks, diarrhoea ceases and irritability and depression of mood improve within 24 hrs.

Secondary bacterial and/or fungal infection of lesions require appropriate antibiotic therapy.

Although zinc is usually non-toxic, high doses for a long period can result in gastrointestinal symptoms, dizziness and copper deficiency, leading to anaemia.

Related information

References:

On DermNet NZ:

- Systemic diseases
- Necrolytic migratory erythema (glucagonoma)

Other websites:

- Acrodermatitis enteropathica images on the DOIA website
- Acrodermatitis enteropathica – emedicine dermatology, the online textbook

Books:

See the DermNet NZ bookstore

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