Acrodermatitis enteropathica (AE) is a disorder of zinc metabolism that can be either congenital or acquired. Acrodermatitis enteropathica is also known as congenital zinc deficiency, Brandt syndrome, and Danbolt-Cross syndrome. The mineral zinc is an important co-enzyme and is structurally important in making gene regulation proteins.

Congenitally, it is an autosomal recessive disorder of zinc metabolism resulting from a defect in the small intestine’s ability to absorb zinc. It affects 1 in 500,000 births. There does not seem to be a predilection for race or sex. Symptoms start around the time of weaning of the infant.

One acquired form of AE can be seen post-bypass surgery when patients lack appropriate zinc supplementation. Another acquired form which is also a transient type, can be seen in infants which results from failure of zinc secretion in the mother’s breast milk, thus absence of zinc in the infant’s diet. Zinc binding factor is normally produced by the pancreas and secreted into breast milk, but may be lacking in this transient type of AE. This transient form results in a symptomatic infant prior to weaning, unlike in the congenital form.

A common manifestation of AE is pustular periorificial dermatitis. This presents as a blistering of the skin around the mouth, anus, and eyes. Hair loss may be evident as well. Other signs and symptoms include nail dystrophy, diarrhea, impaired immunity, and neurological deficit.

Diagnosis is typically made through identifying extremely low zinc levels in plasma, urine, or hair. More definitively, in the congenital form, genetic testing can reveal a defect in the SLC39A4 gene on chromosome 8q24.

Following repletion of zinc to therapeutic levels, skin lesions resolve rather quickly with good prognosis. Sometimes, near puberty, congenital AE may recur. However, appropriate lifelong supplementation with zinc sulfate allows patients to lead normal, healthy lives.