

Polyendocrinopathy

Endocrinopathy

[Endocrine-glands-path](#)

The hallmark of IPEX is the early onset of autoimmune endocrinopathies, typically manifest as diabetes or thyroiditis.

Type I Diabetes Mellitus

[\(1\) Wand Dyed-bead-pancreas](#)

IPEX syndrome is associated with diabetes in male infants. Diabetes mellitus type I is the most common endocrinopathy. Hyperglycemia may be seen at birth due to immune-mediated destruction of islet cells.

Enteropathy

Enteropathy

[Intestine-path](#)

Enteropathy is a common manifestation seen in infants with IPEX syndrome. It can cause failure to thrive. Manifestations may include intractable diarrhea, gastritis, vomiting, ulcerative colitis, celiac disease, and ileus.

Severe Diarrhea

[Severed Toilet](#)

Enteropathy can be featured as severe diarrhea with mucoid or blood. It can be aggravated by a diet change from breastfeeding to formula. This symptom can be life-threatening in patients.

Other Features

Dermatitis

[Dermatologist-examining-rash](#)

Dermatitis in IPEX syndrome manifests as an eczematous rash (mainly atopic dermatitis) that occurs in early infancy. Other manifestations may include alopecia, cheilitis, and onychodystrophy.

Nail Dystrophy

[Nail Disc-trophy](#)

An abnormal change in color, texture, shape, and growth of the fingernails or toenails characterizes nail dystrophy. This feature can also be seen in patients with IPEX syndrome.

Treatment

Bone Marrow Transplantation

[Bone Train-plant](#)

Patients with IPEX syndrome will need to be treated with supportive care. The standard treatment for this disorder is bone marrow transplantation, and if left untreated, it's often fatal.