



Childhood Mastocytosis

(Urticaria Pigmentosa, Mastocytoma, Diffuse Cutaneous Mastocytosis)

What is Mastocytosis?

Mastocytosis is a group of disorders in both children and adults caused by the presence of too many mast cells in a person's body. Mast cells are located in skin, the linings of the stomach and intestine, as well as other sites and may play an important role in helping defend these tissues from disease. By releasing chemicals such as histamine, mast cells attract other key players of the immune defense system to areas of the body where they are needed. Histamine is involved in reactions such as allergies and hives.

There are several different forms of Cutaneous Mastocytosis. Some are Cutaneous (skin) and others are systemic (involve internal body organs). Cutaneous (skin only) Mastocytosis is the most common form. It is also called **urticaria pigmentosa** (if there are several lesions) or **solitary mastocytoma** (if there is a single lesion). CM mostly affects children. Diffuse cutaneous mastocytosis is a rare familial (inherited) form of mastocytosis in which all of the skin is affected.. These childhood forms of mastocytosis should NOT be confused with the more severe adult forms.

Systemic mastocytosis is caused when mast cells collect in the tissues and can affect organs such as the liver, spleen, lymph nodes, and bone marrow. This tends to occur in adults and is rare in children.

What are the symptoms of childhood mastocytosis?

Chemicals released by mast cells cause hive or allergy symptoms including:

- Itching
- Abdominal cramping
- Diarrhea
- Facial flushing
- Skin lesions which swell, blister, or become red
- Wheezing (less common)
- Episodes of low blood pressure and faintness (uncommon)

How is childhood mastocytosis diagnosed?

Doctors can often diagnose these conditions by seeing the characteristic lesions which are light pink-brown to yellow-brown in color, stay in the same location (do not come and go) and become red or swollen when rubbed. A small skin sample (biopsy) is sometimes needed to confirm the diagnosis.

How is mastocytosis treated?

Several medicines help treat the symptoms of mastocytosis.

- Antihistamines (such as Benadryl) to help with itching or skin reactions
- Medicines (such as cromolyn sodium) to relieve persistent abdominal cramping or diarrhea