

## Urticaria Pigmentosa (Mastocytosis)

### What is urticaria pigmentosa?

Urticaria pigmentosa (UP) is a skin rash. This kind of rash is not seen very often. The rash is made up of red to brown color spots that are flat or slightly raised. Sometimes the spots will blister. There may be just a few spots or many spots. The spots form hives when the rash is rubbed or scratched.

It is diagnosed by signs and symptoms such as a skin rash and itching.



### What causes it?

There are mast cells in most parts of the body. They help fight infection and make a substance called histamine. Mast cells release histamine in the skin. Histamine causes hives, itching and the red color of a rash. UP happens when there are more mast cells in the skin than normal. The exact cause for the extra mast cells is not known. Many things may trigger UP. A trigger is something that makes the rash worse.

### How does it affect my child?

The rash does not hurt, but may itch. A very large release of histamine can cause headaches, flushing, diarrhea, vomiting, wheezing (breathing hard with a whistling sound), increased heart rate, and a decrease in blood pressure. This is not common.

### How is urticaria pigmentosa treated?

A skin biopsy might be done. X-rays and blood tests may be done as well.

- **Use medicine as directed.** A medicine, called an antihistamine, is given by mouth. It is used to decrease the release of the histamine from mast cells. It may make the rash less itchy.
- **Take care of your child's skin.** If the rash gets dry, scaly or rough, put on a moisturizing cream or ointment 3 to 4 times a day.
- **Avoid things that trigger histamines and UP.** Triggers do not cause UP but they may bring out the rash or make the rash worse. A very large release of histamine in the skin can cause low blood pressure or even shock which is very uncommon. Some triggers include:
  - Stress.
  - Medicines, including some pain relievers.
  - Some foods.
  - Strong, hard rubbing of the skin.
  - Bee and wasp stings.

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## How long does it last?

Most children will outgrow UP as they get older. New spots may keep appearing as long as the condition lasts. There is no known treatment to keep new spots from appearing. The reddish-brown spots may fade as your child grows older but can last for months or even years.

## Important information

- Tell your child's primary care doctor, dentist, childcare provider and school that your child has UP. Give a copy of this sheet to these people. Keep a copy of this information handy in case of an emergency. Keep a copy in the glove box of your car, your purse, by your phone, etc.
- You can order a medical alert bracelet. It should say: **Urticaria Pigmentosa (Mastocytosis)**.
- Talk with your child's primary care physician about carrying an EpiPen<sup>®</sup> for emergencies.
- If your child has surgery or goes to the emergency room, tell the doctors that your child has urticaria pigmentosa. Give them a copy of this information sheet.

## What follow-up care is needed?

- Your child's doctor will tell you how often your child needs to be seen.

## Alert—Take your child to the Emergency Room if your child:

- Gets very red. This is called flushing.
- Is wheezing.
- Faints.
- Has a hard time breathing.

**ALERT:** Call your child's doctor, nurse, or clinic if you have any questions or concerns or if your child has:

- Diarrhea that does not go away.
- Special health care needs that were not covered by this information.

**This teaching sheet is meant to help you care for your child. It does not take the place of medical care. Talk with your healthcare provider for diagnosis, treatment, and follow-up.**