

# What is a port-wine stain (also known as a port-wine birthmark)?

A port-wine stain is a type of birthmark made of dilated small blood vessels in the skin. It is also called a capillary malformation.

This type of birthmark is usually present at birth. It can appear as light red or darker red to purple discolorations on any part of the body but is most common on the forehead, nose, cheek and chin. Port-wine stains usually grow in proportion to the growth of the child. Unlike hemangiomas, a more common type of vascular birthmark in children, port-wine stains are flat, do not grow quickly, and do not go away on their own.

For most children with this type of birthmark, there are no other associated health problems. In a small group of children, port-wine stains can be associated with brain and eye problems. This occurs in a condition called Sturge-Weber syndrome.

Port-wine stains can occur on other parts of the body including arms and legs and can be associated with overgrowth of the soft tissues and bones underlying the stain.

Over time, port-wine stains become darker red or purple in color and the involved skin may get thicker. The teeth, gums, and jaw underneath a port-wine stain may slowly enlarge over time, which often requires surgery.

## **▼** WHAT CAUSES PORT-WINE STAINS?

In the past few years, an important discovery was made about the cause of port-wine stains. In most children, a small genetic change occurs in the birthmark in a gene called GNAQ. Port-wine stains occur spontaneously, and are not inherited from parents.

### **PORT-WINE STAIN FACTS**

- » Port-wine stains occur in 1 out of 200 children.
- » Port-wine stains affect only one side of the body in 85% of cases.
- » Only about 10% of patients with a port-wine stain in certain areas of the face will have Sturge-Weber syndrome.

# **HOW ARE PORT-WINE STAINS TREATED?**

Laser therapy with a **pulsed dye laser (PDL)** can help lighten the color of the port-wine stain and may prevent darkening and thickening of the stain with time. The laser works by targeting a part of red blood cells called hemoglobin. When the laser hits the skin, the energy from the laser is absorbed by the red blood cell, which causes it to become hot and, in turn, destroys the red blood cell and the surrounding abnormal dilated blood vessels. Complete clearance of the port-wine stain is difficult, however, even with laser treatment.

Generally 4 to 8 laser treatment sessions are performed on the skin, about 6-8 weeks apart. Some experts believe that starting treatment before 1 year of life can yield better results because the skin of a young infant is thinner, allowing the laser to penetrate more effectively. Stains on the extremities do not respond to the pulsed dye laser as well as stains on the face or neck.

Some patients describe the pulse of the laser as similar to a rubber band snapping against the skin. Depending upon the size and location of the stain, laser therapy may be performed without anesthesia, with topical anesthesia, or under general anesthesia. This is an important point to discuss with your doctor.

Immediately after the laser treatment, the port-wine birthmark will look bruised and may feel sore. Redness, swelling and itching may also occur immediately after the procedure and last for a few days. An ice pack may be applied to reduce discomfort. The bruising may last for 2-3 weeks. Although rare, blistering of the skin may occur. Protection of the treated area from the sun is important to avoid brownish discoloration of the skin after the bruising has resolved. It is also necessary to minimize tanning, which can decrease the usefulness of laser treatments. The risk of scarring from the pulsed dye laser is very small. With time, the remaining stain can begin to darken again and retreatment may be necessary.

### **PULSED DYE LASER FACTS**

For every 100 port-wine stains treated with laser treatment:

- » 80% will get at least 50% lighter.
- » 10% will disappear completely.
- » 10% will have no change.



The Society for Pediatric Dermatology 8365 Keystone Crossing, Suite 107 Indianapolis, IN 46240 (317) 202-0224 www.pedsderm.net

The Society for Pediatric Dermatology and Wiley Publishing cannot be held responsible for any errors or for any consequences arising from the use of the information contained in this handout.

© 2016 The Society for Pediatric Dermatology

Contributing SPD Members: Muhammad Amjad Khan, MD Tess Peters, MD, MSc Ki-young Suh, MD

Committee Reviewers: Brandi Kenner-Bell, MD Andrew Krakowski, MD

**Expert Reviewer:** Lawrence Eichenfield, MD