This leaflet explains about port wine stains and what to expect when your child comes to Great Ormond Street Children's Hospital for treatment.

**What do they look like and where can they occur?**

A port wine stain is a flat, red or purple mark on the skin that is present at birth. Very occasionally, over time, the port wine stain may become thicker, darken and develop a ‘cobblestone’ appearance with raised bumps and ridges.

Port wine stains can appear anywhere on the body, in most cases on one side of the body only, but occasionally on both sides. About 65 per cent of port wine stains are on the head and neck.

**What is a port wine stain and what causes it?**

A port wine stain is a vascular birthmark caused by abnormal development of blood vessels in the skin. A port wine stain is sometimes referred to as a capillary malformation.

The change in the blood vessels is caused by a mutation (change in a gene) occurring early in pregnancy while the baby is developing in the womb.

This change in the gene is not inherited (passed on from one generation to the next) and is not known to be related to anything that happened during pregnancy.

**How common are port wine stains?**

About three in every 1000 children has a port wine stain. Girls are twice as likely to have a port wine stain as boys, but we do not know why.

**How are port wine stains diagnosed?**

Port wine stains are clearly noticeable and quite different to other types of birthmark so no special diagnostic tests are usually needed.
Looking after your child’s port wine stain

Generally, port wine stains do not need any special treatment. However, they do need protection from the sun, before, during and after laser treatment. Use a high factor sun cream on all areas of exposed skin, and use a hat to protect your child’s face and/or an umbrella over the buggy or pushchair.

Are port wine stains associated with any other conditions?

Port wine stains involving the upper part of the face can be linked to the following conditions:

- **Glaucoma** – Children with a port wine stain around the eye have an increased risk of glaucoma. Glaucoma is raised pressure within the eye, which can lead to blindness if it is not treated. Treatment is usually by eye drops and occasionally an operation. A specialist eye doctor (ophthalmologist) should examine your child’s eyes to check for glaucoma regularly. You may notice when comparing to the normal eye, that the eye on the port wine side looks different. If the pupil looks larger, the eyelids are open further or the eye itself looks larger or more prominent than the other, please ask for your child’s eyes to be checked. This should be with a specialist eye doctor at your local hospital or specialist centre.

- **Sturge-Weber syndrome** – If your child’s port wine stain is on the skin around the eye, forehead or scalp, there is a chance that he or she may have a condition called Sturge-Weber syndrome. As well as the port wine stain affecting the skin, it may also involve blood vessels over the surface of the brain, which can cause seizures (fits or convulsions). If there is any suspicion that your child is at risk of Sturge-Weber syndrome, he or she will need to be checked over by a neurologist.
**Klippel Trenaunay syndrome** – A large port wine stain on the arm or leg might be associated with extra growth of that limb and is referred to as Klippel Trenaunay syndrome. This may need a multidisciplinary review by dermatologists, laser specialists, and general, orthopaedic and vascular surgeons. For more information, please see our *Klippel-Trenaunay syndrome* leaflet.

Port wine stains, particularly if they are on the face, can affect children psychologically, emotionally and socially. Our leaflet *Bringing up a child whose face looks different* contains lots of ideas and suggestions from the Birthmark team and parents of children with birthmarks.

**How can port wine stains be treated?**

Although it can be useful for a child with a port wine stain to see a specialist doctor soon after birth, treatment does not usually start later in infancy. There are currently two options for treating port wine stains: laser treatment and cosmetic camouflage.

Laser treatment, with a pulsed dye laser, is currently the treatment of choice for fading a port wine stain. It may also help the 'cobblestone' effect that can occasionally develop on facial areas in adulthood. When laser treatment is carried out at a specialist centre, experienced in dealing with vascular birthmarks in children, the results can be excellent and the side effects minimal. More information about laser treatment follows:

- **What is laser treatment and how does it work?**
  Laser treatment uses a narrow beam of light that is absorbed by the red colour in the blood vessels in the port wine stain. This is called selective photothermolysis, which means that the specific area (selective) of tissue containing blood vessels is treated (lysis) using light (photo) that in turn produces heat (thermo). Each
time the laser beam touches the skin, it treats a small area only a few millimetres across. We call this a laser ‘dot’ and most children have lots of ‘dots’ in one laser treatment session.

- **How long does laser treatment take?**
  In our experience, the best results occur when a series of treatments, usually between four and six, take place over a few years. The length of each treatment varies according to whether your child has treatment under general anaesthetic or local anaesthetic and the characteristics of his or her birthmark.

- **Test patch**
  The first stage in the treatment process is usually a ‘test patch’, which shows how well your child’s birthmark responds to the laser. Most of our children have a test patch, but if the port wine stain is very small, it might not be needed. The test patch involves having a few laser ‘dots’ on the birthmark itself. The ‘dots’ can be uncomfortable (children have told us that it feels like being flicked with a rubber band) so the area of skin being tested is usually numbed first using local anaesthetic cream. The treated area will be covered with a cold dressing afterwards to make it more comfortable.
  As with all laser treatment, your child’s skin will need to be looked after carefully after the test patch. For more information see our leaflet *After your child has had skin laser treatment*. A few months afterwards, when the skin has settled down, you will need to come back to the hospital so the results of the test patch can be checked and any future treatments planned.

- **Laser treatment**
  Once your child has had good result from a test patch, he or she can start to have regular treatment sessions. These involve many more ‘dots’ than the test patch, so some children have laser treatment under a general anaesthetic. As it is thought that repeated general anaesthetics (GA) in infancy may have an adverse effect on neurodevelopment, we do not start GA treatments until after the age of two years. Children can also have local anaesthetic treatment if the size of port wine stain, its location on the body and the number of ‘dots’ planned makes this practical.
What happens afterwards?

If your child had laser treatment using local anaesthetic, he or she will be able to go home immediately afterwards. If your child had a general anaesthetic, he or she will need to stay on the ward for a few hours afterwards until he or she has woken up completely.

Your child’s skin will look bruised and sore after each laser treatment. His or her skin will need to be looked after carefully in the weeks between each appointment. Full instructions are in our leaflet *After your child has had skin laser treatment*, which will be given to you after each treatment session.

What is cosmetic camouflage?

Cosmetic camouflage does not take away a port wine stain, but covers it up using a special kind of make up, which is water resistant. If cosmetic camouflage is used on the face, it’s best removed every night with cleansing cream, but if it’s elsewhere on the body it can be left for three or four days. In our experience, cosmetic camouflage is ideal for special events, such as parties, but most children do not bother to use it every day.

An appointment to learn how to use cosmetic camouflage is necessary and it can take up to an hour to find the right combination of shades to cover the port wine stain completely. Once you have learnt how to use cosmetic camouflage, you can order it on prescription from your family doctor (GP).

Are there any long-term problems associated with port wine stains?

In time, facial port wine stains can become thicker, darken and occasionally develop raised bumps and ridges, but these changes are less likely to arise if laser treatment has been carried out. Port wine stains have some specific long-term effects that may need treatment in the future:

- **Papules and pyogenic granulomas**
  - Port wine stains can sometime develop small blood vessel blisters called papules or pyogenic granulomas, which can bleed easily. If your child develops a papule, it is best treated early, before it develops into a pyogenic granuloma. They can be removed using laser treatment, cryotherapy (freezing) or surgery.
Soft tissue hypertrophy – Occasionally, the tissue beneath a port wine stain may enlarge; this is called ‘soft tissue hypertrophy’ and is most common around the lip.

What is the outlook for children with port wine stains?
If your child’s port wine stain was treated with a laser, you should see an improvement in its appearance, although it will not disappear completely. In our experience, 70 per cent of children with facial port wine stains having laser treatment will see an improvement in colour of about 70 per cent, after a course of four to six laser treatments.

A small number of children have a less good response to laser treatment, with little fading. The success of laser treatment varies depending on the part of the body affected and the depth of the abnormal blood vessels. In our experience, the laser works best on port wine stains that are bright pink or red and less well if the port wine stain is purple. Port wine stains on the face respond better than port wine stains on the trunk or legs.

Conclusion
Port wine stains can be treated at any age.
Where to get further information

- At Great Ormond Street Hospital
  Birthmark Unit, Great Ormond Street Hospital, London WC1N 3JH
  Tel: 020 7405 9200 ext 1113

- Support groups
  Birthmark Support Group
  BM The Birthmark Support Group, London, WC1N 3XX
  Tel: 07825 855 888
  Email: info@birthmarksupportgroup.org.uk
  Website: www.birthmarksupportgroup.org.uk

  Changing Faces
  The Squire Centre, 33-37 University Street, London WC1E 6JN
  Tel: 0845 4500 275 Fax: 0845 4500 276
  Web: www.changingfaces.org.uk  www.iface.org.uk