Policy for the Management of Port Wine Stains
The CCG policy has been reviewed and developed by the Treatment Policies Clinical Development Group in line with the groups guiding principles which are:

1. CCG Commissioners require clear evidence of clinical effectiveness before NHS resources are invested in the treatment;
2. CCG Commissioner require clear evidence of cost effectiveness before NHS resources are invested in the treatment;
3. The cost of the treatment for this patient and others within any anticipated cohort is a relevant factor;
4. CCG Commissioners will consider the extent to which the individual or patient group will gain a benefit from the treatment;
5. CCG Commissioners will balance the needs of each individual against the benefit which could be gained by alternative investment possibilities to meet the needs of the community
6. CCG Commissioners will consider all relevant national standards and take into account all proper and authoritative guidance;
7. Where a treatment is approved CCG Commissioners will respect patient choice as to where a treatment is delivered; AND
8. All policy decision is considered within the wider constraints of the CCG’s legally responsibility to remain fiscally responsible
Category: Not Routinely Commissioned.

Port Wine Stains

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.

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. 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.

Port wine stains are clearly noticeable and quite different to other types of birthmark so no special diagnostic tests are usually needed. Generally, port wine stains do not need any special treatment. However, they do need protection from the sun. The patient should use a high factor sun cream on all areas of exposed skin, and use a hat to protect the patient’s face and/or an umbrella over the buggy or pushchair.

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

Glaucoma: Patients 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 the patient’s eyes to check for glaucoma.

When comparing to the normal eye, the eye on the port wine side may look different. If the pupil looks larger, the eyelids are open further or the eye itself looks larger or more prominent than the other, the patient’s eyes should be checked by an ophthalmologist (specialist eye doctor).

Sturge-Weber syndrome: If the port wine stain is on the skin around the eye, forehead or scalp, there is a chance that the patient 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 the patient is at risk of Sturge-Weber syndrome, the patient will need to be reviewed 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, and general, orthopaedic and vascular surgeons.

**Treatment Options for Port Wine Stains**

There are 2 options for treatment of port wine stain:

1. **Camouflage Makeup**
2. **Laser Treatment**

1. **Camouflage makeup**

Skin camouflage products effectively cover the affected area. Changing Faces provides education by trained volunteer practitioners on the use and application of cosmetic camouflage creams and powders, and people may self-refer.

Details of the nearest skin camouflage service can be found at www.changingfaces.org.uk.

Cosmetic camouflage creams can be used on any part of the body. The aim is to provide natural-looking cover. They are waterproof, and may remain on the body for up to 4 days, and on the face for 12–18 hours.

Three brands of camouflage product, in a range of shades, are included in the Birmingham, Sandwell and Solihull APC Formulary, and the prescription must be endorsed 'ACBS'.

- Covermark classic foundation (10 shades) and Covermark finishing powder.
- Dermacolor camouflage cream (100 shades) and Dermacolor fixing powder.
- Keromask masking cream (9 shades) and Keromask finishing powder.

2. **Laser treatment**

Laser treatment for a port wine stain, may lighten the affected area of skin. NICE IPG90 (2004) states the following: Laser treatment is often recommended for lesions near the eyes or orifices, or if lesions bleed, ulcerate or become infected. However, external laser treatment of these vascular abnormalities may not be effective because the laser beam does not penetrate far beneath the skin.

There are multiple laser modalities, each having different settings that can be used for port wine stain. The pulse dye laser is considered the current gold standard. The evidence review produced for the CCG policy review (please see CCG website for full evidence review) suggests that the pulse dye laser is clinically effective in reducing the colour of port wine stains in the short term. The evidence review also suggests that other laser modalities may show clinical effectiveness in reducing the colour of port wine stains within trial settings, and combinations of laser and other adjunctive
may augment the action of pulse dye lasers, although there is no strong systematic review/guidance in this area.

The pulse dye laser was generally considered safe, although long term outcomes are not well studied. Short term adverse effects are common which vary from pain, skin crusting and bullae.

There was limited evidence investigating quality of life after treatment. One qualitative study found that the majority of patients felt that pulse dye laser therapy improves PWS colour over time with just under half of them feeling satisfied with treatment, but questionnaire response rates were around 50%. Most patients believed that the laser treatment did not improve their social interaction but it did help reduce their worry about their appearance.

There was no evidence found investigating the cost effectiveness for the treatment of port wine stains. The overall application of these findings to clinical practice is limited and further research and investigation is warranted, especially around long term outcomes, quality of life and cost effectiveness.

**Eligibility Criteria.**

Patients with port wine stains involving the upper part of the face should be appropriately referred for further investigation to ensure complications of port wine stain, as outlined above, are identified and managed appropriately.

Patient may access camouflage make-up through their GP as described above.

**Laser treatment is not routinely commissioned for the treatment of port wine stains.**

This is because there is insufficient clinical evidence to support the use of laser treatment as an effective intervention for port wine stain, further research is warranted, particularly around long term outcomes, quality of life and cost effectiveness.

This means the CCG will only fund the treatment if an Individual Funding Request (IFR) application proves exceptional clinical need and that is supported by the CCG.
Guidance


Kelly KM1, Choi B, McFarlane S, Motosue A, Jung B, Khan MH, Ramirez-San-Juan JC, Nelson JS.OI Description and analysis of treatments for port-wine stain birthmarks.


Berit C. Carlsen, MD, PhD et al. (2017) *A Randomized Side-by-Side Study Comparing Alexandrite Laser at Different Pulse Durations For Port Wine Stains* Department of Dermatology, Bispebjerg University Hospital, 2400 Copenhagen, NV, Denmark, Boom Laser Clinic, 2850 Boom, Belgium (2017)

