Facial Port Wine Stains

Port wine stains (PWS) are capillary malformations with an estimated incidence of 3 per 1000 live births. As with other vascular anomalies, the location of the capillary malformation determines the potential risk of extracutaneous manifestations and local complications.

Potential complications of facial port wine stains include:

- Association with neurocutaneous syndromes (Sturge-Weber syndrome)
- Ocular complications
- Hypertrophy
- Oral involvement
- Negative psycho-social impact

Traditionally, port wine stains located in the distribution of the first branch of the trigeminal nerve (V1) have been associated with a risk for Sturge-Weber syndrome (SWS). Recent studies along with identification of specific genetic mutations propose that port wine stains are likely caused by faulty embryological development of facial vasculature. These studies propose that port wine stains on the forehead should be considered at risk for SWS.

Recommended evaluations of facial PWS:

- PWS on the forehead:
  - Presence of neurologic symptoms \(\rightarrow\) neurologic evaluation with further imaging
  - Absence of neurologic symptoms \(\rightarrow\) consider central nervous system imaging between 6-12 months of age.

- Periocular PWS \(\rightarrow\) ophthalmology evaluation to rule out glaucoma

- Oral involvement \(\rightarrow\) evaluate teeth development and speech development

- Psychosocial impact \(\rightarrow\) facial PWS can negatively impact quality of life. Depression and anxiety are common among adults with facial PWS. Consider mental health assessment.

Treatment of PWS

While different technologies have been used to treat capillary malformations, pulsed-dye laser is considered the treatment of choice. Pulsed-dye laser causes selective photocoagulation of the capillary malformation vessels. Appropriate counseling of parents and patients is important to establish realistic expectations of therapy. Different studies have shown that treatment at a younger age leads to higher rates of clearance, reduced number of treatments and lower incidence of hypertrophy and complications.
PDL treatment recommendations:

- **Age to start**
  - 1 month old if no sedation required
  - 6 months old if sedation required
- **Frequency of treatment** → every 6-8 weeks
- **Number of treatments** → variable, depends on location and extent of capillary malformation
- **End point**
  - Complete clearance is attained in less than 20% patients
  - 70-80% lightening is considered a very good response
- **Safety**
  - Appropriate ocular protection when PDL is done on periocular / eyelid area
  - Consider decreasing PDL fluence on eyelid and neck

**Future directions**

Different therapeutic modalities are being studied to enhance treatment efficacy. Rapamycin (sirolimus) has been shown to have anti-angiogenic effects. When used in combination with pulsed dye laser, rapamycin (off-label use) may enhance the treatment efficacy in part by preventing re-vascularization after laser injury.

**References:**


