Objectives:

1. Recognize clinical implications and potential complications of PWS based on their location.
2. Understand scientific advances and future directions in the management of PWS.

Introduction

Port wine stains (PWS) are capillary malformations occurring in approximately 0.3% of newborns. The head is the most common location, although this can develop anywhere on the body.

Our understanding of the pathogenesis and embryological origin has evolved in recent years. An important discovery was the finding of somatic mosaic mutations in GNAQ associated with syndromic and non-syndromic PWS. These findings along with studies of the distribution patterns of facial PWS suggest these malformations derive from the frontonasal prominence.

Extracutaneous complications

Facial PWS can be associated with neurologic, ocular and oral complications. The risk of these complications is determined by the specific location of the capillary malformation.
### Compensation

<table>
<thead>
<tr>
<th>Complication</th>
<th>PWS location</th>
<th>Work up / Evaluation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Neurologic</td>
<td>Forehead&lt;br&gt;Paramedian&lt;br&gt;Hemifacial</td>
<td>Symptomatic: Neurology evaluation&lt;br&gt;Asymptomatic: Consider MRI after 6-12 months, Consider EEG, Discuss with Neurology</td>
</tr>
<tr>
<td>Ocular</td>
<td>Periocular (upper or lower eyelid)</td>
<td>Regular Ophthalmology evaluation</td>
</tr>
<tr>
<td>Oral</td>
<td>Lower face&lt;br&gt;Perioral / oral</td>
<td>Dental evaluation&lt;br&gt;ENT evaluation&lt;br&gt;Speech</td>
</tr>
<tr>
<td>Psycho-social</td>
<td>Any location</td>
<td>Assess for symptoms of depression, anxiety and psycho-social complications</td>
</tr>
</tbody>
</table>

### Natural Course

In time, PWS can develop darkening, soft tissue hypertrophy, bony hypertrophy and vascular nodules. Tissue hypertrophy develops in up to two thirds of patients. Most studies show an average age of onset of hypertrophy in the mid-30’s to early 40’s, although it may develop at an earlier age. The incidence of hypertrophy increases with age.

### Treatment

While different technologies have been used to treat capillary malformations, pulsed-dye laser is considered the gold standard therapeutic modality. 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.

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.
**Pulsed dye laser treatment approach**

**When?**
- >1 month of age without sedation
- >6 months of age if sedation is required

**FDA warning**

**How often?**

Every 6-8 weeks. Increased frequency every 4-6 weeks has also been shown to be safe and effective.

**Number of treatments?**

Variable. The number of treatments is dependent on the extent of the PWS, color and specific location (medial cheek is more resistant to lightening).

**End-point?**

Complete clearance is only attained in <20% of patients. Lightening of the PWS of 70-80% represents an adequate outcome.

References:


