Considerations on port-wine stains and their laser treatment
van der Horst, C.M.A.M.

Citation for published version (APA):

General rights
It is not permitted to download or to forward/distribute the text or part of it without the consent of the author(s) and/or copyright holder(s), other than for strictly personal, individual use, unless the work is under an open content license (like Creative Commons).

Disclaimer/Complaints regulations
If you believe that digital publication of certain material infringes any of your rights or (privacy) interests, please let the Library know, stating your reasons. In case of a legitimate complaint, the Library will make the material inaccessible and/or remove it from the website. Please Ask the Library: http://uba.uva.nl/en/contact, or a letter to: Library of the University of Amsterdam, Secretariat, Singel 425, 1012 WP Amsterdam, The Netherlands. You will be contacted as soon as possible.
A port-wine stain (PWS) is a vascular birthmark, consisting of ectatic dermal blood vessels. The affected vessels can show a progressive ectasia, which sometimes leads to nodularities and a cobblestone pattern in the skin (1). These birthmarks occur in approximately 0.3-0.6% of newborns with an equal sex distribution (2). The stigma of a disfiguring facial birthmark may have a substantial effect on social and psychosocial development in children and adults (3). Bleeding either spontaneous or because of pyogenic granulomas can occur in adults. Early treatment of the PWS, when leading to disappearance or diminishment of the size and color of the lesion could improve psychological development of the children and may prevent progression of the lesion.

**Definition**

‘Port-wine stain’ is a generally used name for a capillary vascular malformation, present at birth and persistent through life. This stain is also called: nevus flammeus. The malformation is growing commensurately with the child. Patient history and appearance make a distinction possible between hemangioma, which is not present at birth and shows a fast growth, followed by almost complete resorption. Stork bites and salmon patches are capillary malformations present at birth on specific areas (glabella, nape of the neck, eyelids) which fade away during the first years of life (4, 5).

Throughout life distention of these vessels of a PWS can lead to thickening of the malformation. Associated hypertrophy of the affected area can be present from the start. In a facial PWS the ipsilateral eye and brain may be involved. Such a combination is called the Sturge-Weber syndrome (6, 7).

**Etiology**

Vascular malformations consist of a group of inborn errors of vascular morphogenesis, which include any type of venous and lymphatic malformation, telangiectasias, and capillary malformations such as salmon patches, stork bites, and PWS (5). All capillary malformations exhibit mature, ectatic capillaries with flat endothelium in the dermis, primarily in the superficial plexus. It has been suggested that PWS are caused by a disturbance of the neural crest (8): normally neural crest cells give rise to vasomotor nerve cells, which were shown to be lacking in the perivascular tissue in PWS (9, 10). Hence, innervation is altered, which is supposed to cause a dysfunction in vasoconstriction, and, so, to lead to vasodilatation. This was confirmed in other studies, which showed a reduced response of PWS to both vasodilating and vasoconstricting stimuli (11). In the other capillary malformations as salmon patches and stork bites, the cause is supposed to be a defect in maturation of the cutaneous sympathetic innervation (12). Familial occurrence of PWS has been infrequently reported (13-17). There is no clue whatsoever for its molecular genetic background.
Psychology

There is evidence that, in general, an attractive appearance is associated with social competence and interpersonal ease. This notion is present in the “What is beautiful is good” theory of Dion and Berscheid (18). The cultural ideal of physical attractiveness and fitness is still spreading (19). In these times where ‘everything’ seems available, having a visible stigma can be experienced as unacceptable. Psychological morbidity in adult patients with PWS is often mentioned in the literature. However, standard psychological screening has not shown any difference compared to controls.

Therapeutic modalities

Many methods have been used to reduce the visibility of the birthmark. Ionizing radiation has been used without positive effects. Unfortunately for patients, treated with this modality we now see skin carcinomas develop in the formerly treated area (20). Cryotherapy did not have the intended effects and caused scarring. After operative removal of PWS recurrent vessels were often seen in the edges of the former excision. Reconstruction of the defect created after excision was done with skin grafts and/or free flaps which both have their own drawbacks, especially in the face, like (hypertrophic) scarring and color difference to surrounding skin. Tattooing of PWS leads to a static color without the possibility of adaptation to the environment (blush). It is an option for some patients with PWS but cannot be used to hide the cobblestone appearance. In the 1980s the argon laser became the laser of choice for treatment of adult patients (21). In children, however, serious scarring was reported with this technique (22). Children with a PWS under 18 years of age could not be treated until the development of the flash-lamp-pumped pulsed-dye laser. This laser was introduced in 1985. The flash-lamp-pumped pulsed-dye laser was the first laser specifically designed for cutaneous vascular lesions. It is based on the principles of selective photothermolysis: the wavelength of the laser and the duration of the pulse were chosen to produce thermal injury that remains confined to the targeted PWS vasculature (23).

Treatment of PWS with the flash-lamp-pumped pulsed-dye laser was now available for children. Better results with early treatment were reported by some, but not unequivocally confirmed by others (24). It was hypothesized that in young children the skin was thinner than in adults, and the size of the PWS was relatively smaller. As a consequence fewer treatments were thought to be necessary to achieve optimal clearance. Yet there, there are also disadvantages in treating early. The flash-lamp-pumped pulsed-dye laser treatment can be experienced as painful. The sensation of one pulse on the skin is described as “when someone lets go a rubber band against your skin”. In general 50-100 pulses per treatment session are given. These treatments of the complete stain have to be repeated 10-20 times to get optimal clearance (25). Pain is reduced with eutectic mixture of lidocaine and prilocaine (Emla® creme), applicated one hour before treatment, with nerve block and cooling. Increasing anxiety in children made sedation and/or general anesthesia inevitable (26).
Since the first results of port-wine stain treatment in children with the tunable flash-lamp-pumped pulsed-dye laser were described in 1989, we were often confronted with parents applying for treatment of their new born children with a facial PWS. We had some experience in this form of laser treatment since 1990. Confronted with a young patient population we had questions that were not answered in textbooks or literature on the subject at that time.

**Aim of the thesis**

The aim of this thesis is to address the following questions:

1. Is there evidence for a family tendency for PWS?
2. Is treatment of PWS at a young age more effective than treatment at an older age?
3. Is there a relation between location of the PWS and occurrence of ophthalmologic and/or neurologic problems in patients with a facial PWS. Until when and how often should ophthalmologist and/or neurologist check patients at risk?
4. When trying to find the right age to treat patients with PWS, psychosocial adjustment in different age groups is an important issue. Is there a difference in adjustment between children and adults?
5. Complete clearance of PWS through flash-lamp-pumped pulsed-dye laser treatment is seldom achieved. Treatment has to be repeated several times, can be painful, or has to be performed under general anesthesia. How do children and their parents appraise the flash-lamp-pumped pulsed-dye laser treatment?

**Outline of the thesis**

Chapter 1 gives a definition of PWS as described in this study. Background on treatment and history of treatment options are given.

Chapter 2 reviews the results of a search for a positive family history for PWS among 280 consecutive patients enrolled with a port-wine stain.

Chapter 3 describes the design, conducting and results of a prospective study of 100 patients with a facial port-wine stain treated with the flash-lamp-pumped tunable pulsed-dye laser.

Chapter 4 focuses on ophthalmologic and/or neurologic pathology related to PWS in the head and neck region among our patients who presented themselves for laser treatment.

Chapter 5 discusses the results of a questionnaire answered by children (parents of children) and adults with a facial port-wine stain. The questionnaire contained questions about psychological and social items concerning patients' appearance.
Chapter 6 describes the answers on a questionnaire where satisfaction with the results of laser treatment and treatment itself were evaluated.

Chapter 7 reports on a group of patients who were referred for dye laser treatment. These patients had a different diagnosis and were not eligible for pulsed dye laser therapy. Seven patient histories and their diagnostic work-up and treatment are reported.

Chapter 8 presents summary and conclusions of our study and indicates directions for further research.

References


