Considerations on port-wine stains and their laser treatment
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Summary

Chapter 1

This chapter gives a definition of port-wine stain (capillary vascular malformation) and describes how patient history and time course can distinguish a PWS from a hemangioma. Psychosocial mechanisms are identified which may have an impact on persons with a visible anomaly. An overview is given of treatment options for vascular birthmarks including their disadvantages and the view on flash-lamp-pumped pulsed-dye laser treatment at the beginning of our study.

Chapter 2

Questioning of patients applying for laser treatment on prevalence of vascular malformations in their family showed a much higher family tendency (19.6%) for port-wine stains (PWS) than could be expected, based on PWS incidence studies (0.3-0.6%). Interesting pedigrees of affected families could be drawn. However, clear patterns of inheritance could not be found in most instances. These data are put in the context of new developments in the field of angiogenesis and genetics. Genes are detected which show the defect in the origin of vascular anomalies in Rendu-Osler-Weber disease and certain venous malformations where the endothelium seems normal but the coding of muscular wall and matrix is insufficient. Our next step has to go in the direction of DNA material in search of candidate genes.

Chapter 3

We prospectively studied 100 previously untreated patients with a PWS in head or neck. They were divided in four age groups (0-5, 6-11, 12-17, 18-30 years), before treatment with the flash-lamp-pumped pulsed-dye laser. The outcome measure was lightening of the PWS (reduction of difference in color between the skin with the stain and contralateral healthy side skin) as measured with a colorimeter after an average of five treatments (range, three to seven) of the entire lesion.

Of the 100 patients, 11 could not be included in the analysis. The average initial sizes, locations, and colors of the PWS were similar among the groups. When all 89 patients were analyzed together, the average reduction in color between the skin with the PWS and contralateral healthy skin was 40 percent. Difference between age groups in the average reduction in color differences were not significant (P=0.26).

We did not confirm the hypothesis that treatment of PWS at an early age is more effective than laser treatment at a later age.

Chapter 4

Children with a facial PWS can be treated with the flash-lamp-pumped pulsed-dye laser. We addressed the following questions: which newborns are at risk to develop
ophthalmologic and neurologic symptoms and at what age can these problems be expected?

Among a group of 198 patients between 0-30 years of age medical history was obtained. Children < 6 years of age were referred for ophthalmologic and neurologic screening, when not checked earlier. Location and extent of the PWS were recorded according to the division of the ophthalmic and cervical nerves. Involvement of the eyelids was separately noted.

Overall, 31 patients had neuro-ophthalmologic sequelae. When the PWS was localized on both upper and lower eyelid 55 percent had involvement of the accompanying eye. No eye problems were seen when the eyelid was free of this birthmark. Patients with a PWS extending over V1,2,3 had in 46% neurologic problems. When the PWS was located in V3 and C2,3 eye pathology was never seen. Seizures were almost always present before the age of one year. Eye involvement was diagnosed from birth to 28 years of age.

Advise for screening and follow up should be given depending on location and extent of the PWS. This counts for ophthalmologic and neurological sequelae. Unfortunately we were not able to follow patients from birth until adulthood. Only a very long follow up can give the exact answer on the above-mentioned question when eye pathology will develop.

Data regarding prevalence of neurologic pathology may be influenced by the fact that parents of children with a bad neurologic prognosis already from birth are less likely to present their children for tunable-pulsed-dye laser therapy.

Chapter 5

To assess the impact of a facial PWS on behavioral problems in children and on health related quality of life in adults a survey by questionnaire was done. All patients applied for treatment with the flash-lamp-pumped pulsed-dye laser and were not treated before. Two standard general instruments were used, as well as a more specific PWS questionnaire. Children’s (4-12 years) parents received the Child Behavior Checklist (CBCL). Adolescents and adults (13-31 years) received a short version of the RAND Health Insurance Questionnaire from the Medical Outcome Study (MOS-24). A specific PWS questionnaire was sent to both age groups.

The results of the CBCL were compared to a Dutch reference group, from children with constipation and compared to another group of children with a PWS. The results of the MOS-24 were compared with those of a reference population without chronic illness. Children’s parents reported no significant problem behavior. Adults reported low scores for mental health, self perceived health and vitality/health. Adolescents and adults showed statistically significant negative consequences of their PWS in social contacts compared to children (P<0.001, Mann Whitney).

This study confirms the presence of psychological problems in patients with a PWS.

Chapter 6

Introduction of the flash-lamp-pumped pulsed-dye-laser has mostly been evaluated in terms of clearance. We assessed the consequences of the introduction of laser therapy in
childhood. A study was performed in 43 consecutive patients (aged 4-12) with a facial PWS who started treatment. Two separate evaluation questionnaires, completed by the children's parents, were distributed. Response rates were 88% and 95% respectively. Children went through a mean treatment period of 2.5 years. Thirteen (32%) children could not be treated without general anesthesia. Parents reported an extra time-investment in the week following treatment. Most children (95%) missed two days of school for each visit. Only a small number of parents did not perceive the laser treatment as painful; 50% felt the treatment to be a burden for their child. Purpura after treatment were experienced as a stigma. Overall, parents (85%) felt it took a long time to gain optimal clearance. When considering laser treatment for a child, one can not neglect by the additional consequences: pain management, the impact of repetitive treatments, discomfort, anxiety, and the motivation of the child.

Chapter 7

A group of seven patients is described which were all referred for flash-lamp-pulsed dye-laser treatment. At examination, these patients had capillary-venous malformations known as: sinus pericranii. This entity is made up by a combination of certain features: extracerebral vascular malformation, with intracerebral vascular connections. An intracerebral venous malformation can be coexistent. Diagnostic procedures and therapeutic options are described.