Considerations on port-wine stains and their laser treatment
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Citation for published version (APA):

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Chapter 6


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Submitted.

This study was granted by OG 92/004 from the National Health Insurance Board.
Summary

Background
The flash-lamp-pumped pulsed-dye laser (FPDL) has been introduced as the treatment of choice for port-wine stains in childhood. This new treatment option has mostly been evaluated in terms of clearance. In this study we assess the consequences of the introduction of laser therapy in childhood.

Methods
We prospectively studied 43 consecutive patients (aged 4-12) with a facial port-wine stain. Two separate evaluation questionnaires, completed by the children's' parents, were distributed.

Results
Response rates were 38 (88%) and 41 (95%) respectively. Children went through a mean treatment period of 2.5 years. They had gained 38% reduction in the difference in color between the skin with the port-wine stain and the contralateral healthy skin. 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% of the parents felt the treatment to be a burden for their child. Eighty percent reported that their child shuddered when thinking of the laser treatment. Most respondents experienced purpura as a stigma. Overall, parents (85%) felt that it took a long time to gain optimal clearance.

Conclusions
The decision to apply children for FPDL therapy should not only be based on clearance or long-term psychological benefits. It is necessary to consider pain management, the impact of repetitive treatments, discomfort, anxiety, burden of treatment and the motivation of the child.
Introduction

Laser surgery has been introduced as a treatment of cutaneous vascular lesions. Advances in laser surgery in the last decade, particularly the development of the flash-lamp-pumped pulsed-dye laser (FPDL), have allowed children with a PWS (PWS) to have reconstructive and aesthetic therapeutic intervention during infancy. There is no clinical evidence of serious complications in children treated with the FPDL, in contrast with older techniques like the argon laser. FPDL is felt to be a low-risk procedure, even in patients with PWS who need multiple treatments.

Several retrospective studies have reported on the FPDL treatment of children. These studies did not unequivocally confirm better treatment results at early ages. We recently showed in a prospective study that children (0 – 18 year) did not show faster clearance of their PWS than adults (18-31 year). Since the FPDL came into use it has been assumed that infants and children can be treated safely, effectively and probably even more efficiently.

It has been hypothesized that children with a PWS can develop emotional and behavioral disorders. These children are perceived to be at risk for psychosocial problems because of their physical appearance and the effects of their congenital disfigurement on family functioning or child/caretaker interaction. Therefore it seems important to treat children as early as possible.

It is increasingly realized that the evaluation of new medical strategies in plastic and reconstructive surgery should be grounded on an evidence based approach. Outcomes of surgical therapies must also evaluate patient experience, emphasizing the patient’s perspective and quality assessment. Therefore the choice whether to use laser therapy in children with a facial PWS should not be centered on clearance of the PWS or on psychological distress alone. If limited clearance is reached other effects and more general remains of treating PWS in childhood become important.

It was our purpose to evaluate several specific issues of FPDL treatment procedure in children and the consequences for parents and children of the introduction of PWS treatment at early ages. The rationale for early treatment of PWS should be supported by an appraisal of these consequences. We will describe observed medical complications, additional medical care, days of absence from school of the children or work of the parents related to the laser treatment. We also evaluated, from a parent’s perspective, pain during treatment, anxiety of the children before treatment, the impact of postoperative purpura, burden of treatment and time needed to complete therapy.

Material and methods

Patients

Consecutive children (aged 4 to 12) with a previously untreated facial PWS were included in this prospective evaluation study. Parents applied their children for the FPDL treatment, after having learned from the new treatment option from the media. All patients were treated in the department of Plastic and Reconstructive Surgery in the Academic Medical Center at the University of Amsterdam, the Netherlands, between December 1991 and March 1995. Oral informed consent of the parents was obtained.
Patients were treated with a Candela SPTL-1 flash-lamp-pumped pulsed-dye laser (Candela Corp., Wayland, MA), an emission wavelength of 585 nm, a pulse duration of 0.45 ms, a repetition rate of 1 pulse per 3 seconds (0.33 Hz) and a circular 5 mm spot. Treatments were performed with a maximum spot overlap of 20% across the affected area. During each visit the largest possible area of the PWS was treated. In most pediatric patients a single treatment of the entire PWS required a number of visits. Within the 3.5 years study period, there was an average interval period of 8.6 weeks (sd 3.0) between consecutive visits.

All patients were initially treated in an outpatient setting. Prior to the actual start of treatment, a visit with an instructor was scheduled to explain the laser procedure to the children and to treat a test patch on the PWS to ascertain clinical response. Written information, including an explanation of the entire treatment was provided. Parents of patients participated in the process of choosing appropriate pain control, in which prevention of pain was the basic goal. During treatment of children a eutectic mixture of prilocaine and lidocaine (Emla®-cream) and/or nerve blocks were used. Emla® was applied by the parents to their children’s PWS one hour before the procedure. If those local analgesic methods were insufficient, midazolam (Dormicum®) was added for sedation. General anesthesia in a day-care unit was used if children became highly anxious or pains sensitive.

Treatment was to be discontinued if the PWS had reached total or optimal clearance. Optimal clearance was defined when three consecutive treatments did not result in further clearance. The difference in color between the healthy skin and the PWS was measured with a colorimeter. All measurements were performed with a Minolta chromameter type CR-300. Color measurements were taken before laser treatment started and after each following treatment of the entire PWS. Color differences were estimated, based on the color measurement of two identical areas in the face: one on the PWS and one on the contralateral healthy skin.

**Instruments**

The childrens’ parents received by mail two separate PWS specific questionnaires, to be completed by themselves. The use of parent/patient-based self-report follow-up questionnaires was thought to provide a reflection of the parents’ view of the laser treatment. In addition, questionnaires were to provide information on the medical care their children had received, independent of clinical opinion. Completion of both questionnaires required between 15 to 30 minutes. Both questionnaires were distributed to parents between January and May 1994. Initial non-responders were sent another questionnaire after 6 weeks.

With the first questionnaire data were gathered on the appearance of blisters, crusting and infections after treatment, and on extra medical care needed. Parents were asked to record time spent to take care of their children and days of absence and leave of parents and children related to treatment.
Additional data regarding parents’ and patients’ views of the FPDL treatment were collected through a second questionnaire. This instrument contained 6 items, each expressing an opinion on a specific issue of the treatment. These opinions were derived from semi-structured interviews with parents and patients having a facial PWS, conducted in a separate sample of patients and parents. The items covered the following subjects: pain during and following laser treatment, anxiety of the child, burden of treatment, stigma of the treated PWS area and the time span needed to finish treatment. Parents were asked to indicate their assessment on a five-point Likert scale anchored as “strongly agree”, “agree”, “not sure”, “disagree” and “strongly disagree”.

**Analysis**

Responses on both questionnaires were tabulated and evaluated using descriptive statistics. For the children the average number of days of absence from school or day-care was listed. For the parents the average time-investment and days absent from work due to one visit were recorded and reported. Responses to the five-point scale items of the second questionnaire were re-grouped into positive (4 or 5 on the used five item scale), intermediate (3 on the scale) and negatively (1 or 2 on the scale) answers.

**Results**

**Patients**

Forty-three consecutive children (aged 4-12) were approached for this study. Forty-one responded 15 male patients and 26 female patients. At the start of treatment the mean age of the included children was 6.4 years (sd 2.9); the mean surface area of their facial PWS was 98 cm$^2$ (sd 121).

**Treatment**

On average these children went through a treatment period of 2.5 years. The mean number of visits in this time period was 15 (sd 6), during which an average of 9 treatments (sd 3) of the entire PWS was performed. Overall patients gained a relative color improvement of their PWS of 38% (sd 24). None of the patients had completed therapy within the study period.

Figure 1 shows the decrease over time of the color difference between the PWS and contralateral healthy skin for two representative pediatric patients; both treated under general anesthesia. Treatment procedures and treatment outcome within these patients did not differ from the other children. Color differences were measured and determined after each treatment of the entire PWS. In both patients a non-linear relationship between PWS clearing and the number of treatments of the entire PWS could be observed. These two patients reached 57% and 59% clearance, respectively, each after 13 treatments of the entire PWS. The greatest decrease in color difference was achieved in the first five treatments of the entire PWS.

Thirteen children (32%) had to be treated with general anesthesia. Prior to treatment all other children had EMLA® applied to their PWS, occluded with Tegaderm®.
Table 1 represents the treatment characteristics of the children treated with and children treated without anesthetics. General anesthesia was introduced because some children became very frightened or sensitive to pain during the FPDL treatment and were already apprehensive upon arrival at the clinic for a repeat procedure. Parents actively participated in helping the child; by attending the introduction of general anesthesia and the recovery period. Additional hospitalization due to treatment did not occur. The introduction of general anesthesia made the treatment of the PWS in children more efficient. With the use of general anesthesia a larger number of pulses could be achieved and the whole PWS could be treated during 1 visit. The number of pulses given per minute during general anesthesia was approximately twice the number of pulses possible when treating without general anesthesia. (Table 1)

**Questionnaires**

The first questionnaire was returned by 38/43 (88%) parents of children and the second questionnaire by 41/43 (95%). All parents who responded to the first questionnaire also responded to the second questionnaire. On most visits both parents accompanied their child. During the first 24 hours after treatment parents attended their children and gave them extra care. For this purpose 39% of the mothers and 77% of the fathers took on average one day off from their work, mostly the day of therapy. In the week following treatment parents reported a mean extra time-investment of 40 minutes per day (sd 20) to give extra attention to the child and to take care of the treated PWS. Most parents (95%) reported that their children missed day care or (nursery) school after a visit. On average children were kept at home for 1 day after each visit. The parents of five young children (aged 4 to 5) reported additional medical care between two visits. They consulted their general practitioner, an ophthalmologist or a plastic surgeon because of blisters or vesicles on the treated area of the PWS. Two patients needed extra pain medication in the week following treatment.

Table 2 contains the response frequencies on the second questionnaire. Despite the use of EMLA® and general anesthesia, only a small number of parents (8%) saw the laser treatment as not painful. Fifty percent felt the laser treatment to be a burden for their child. Despite the careful introduction of the laser treatment and a preventive pain policy, 23% of the parents reported that their child was nervous before each visit. Eighty percent felt that their child disliked the idea of laser treatment. Most parents experienced the blue-grey discoloration of the PWS after each visit as a stigma for which the child is stared at. Purpuric areas were noted immediately after treatment and remained for 7 to 10 days, after which the skin returned to its former (red) color. Overall, 85% of the respondents felt that it took a long time until their child had gained optimal clearance and completed treatment.

**Discussion**

In this prospective study of children we found that the FPDL treatment of children (aged 4 to 12 years) with a facial PWS was not as easy, effective and efficient as has been hypothesized in the literature. Treatment required pain management,
sometimes necessitating general anesthesia, and multiple treatments for optimal PWS clearing. After each visit, temporary postoperative purpura was observed on the treated skin.

Our results may not unconditionally be generalized to other patients and settings. In this study, all children were submitted to treatment by their parents. This might have caused a selective referral bias. Mostly, parents decided how, when and where to seek therapy, often based on their personal anxiety level some felt they would have denied their child the possibility of medical care by not applying for treatment. Parents who are more concerned and involved or more distressed are more likely to refer a child with a PWS. Therefore a difference in parental coping may exist between referring and non-referring families.

It is difficult to evaluate objectively the pain in infants and children experienced during treatment. Behavioral observations, parents’ reports of children’s’ pain as well as children’s’ own reports can be used to assess the amount of pain. Unfortunately, little is known about the reliability of the cues parents use to assess pain. Although most parents are concerned that their children suffer pain, they may underestimate the amount of pain actually experienced by their children.

We found it necessary to use general anesthesia in 32% of the children. These children either became too frightened or experienced too much pain during treatment. General anesthesia made it possible to treat a larger surface area of the PWS during a single visit. A similar preventive pain management was reported by Rabinowitz et al. In contrast, Tan et al., Morelli et al. and Alster et al. saw no need for general anesthesia when treating young children.

Our study shows that numerous visits are needed to treat PWS in children. None of the forty-three children achieved complete clearance of their PWS within 2.5 years of therapy and multiple treatments. The outcome of treatment and the prognostic value of age, color, anatomical side, mean depth of the vessels and skin type, is still rather unpredictable. In our study the largest degree of fading was seen in the beginning of treatment. Thereafter the additional color reduction in the relative difference between the stain and the contralateral side gradually decreased.

Although it is stated that children with a facial PWS have an increased likelihood of developing emotional or behavioral disorders, there are still no longitudinal studies that demonstrate benefits of treatment to psychosocial functioning in the long-term. Results of recent studies suggest that children with PWS are not psychosocially maladjusted compared to nondisfigured children. Unfortunately the effect of a facial PWS on early socialization in childhood is still unknown. Compensatory parental coping may adjust the deficits of disfigurement, at least for the younger child. Further advances in laser technology could increase the efficacy of treatment. New lasers will administer more pulses per minute, each pulse covering a larger surface area (7mm). This may substantially shorten the duration of treatment and diminish the number of visits needed in the long-term. However the preventive pain policy in children will not be influenced by using a faster and more effective laser; general anesthesia will still be necessary.

We feel that our findings pose a challenge to people responsible for the decision whether or not to start treatment in children with PWS. The extended duration of treatment, requiring multiple painful visits, might be particularly stressful for children.
Due to their age, children may be difficult to motivate to act as a willing participant of therapy. Most young children are incapable of staying still for painful procedures, even if they have the procedure explained. There is evidence that children can suffer long-term emotional consequences from successive painful medical procedures. The decision whether to use anesthesia in children requires a careful balancing of two drawbacks within optimal pain management: a long and possible painful treatment duration without anesthesia versus a shortened treatment process involving the repeated use of anesthetics.

The goal of treating children with the FPDL should be to remove the PWS without causing physical scars or emotional trauma. Therefore the age at which therapy should be initiated must not be based on clinical outcome only but also on a considerate weighing of the expected psychological benefit and the discomfort of treatment. This will without doubt require careful investigation of the interplay between the PWS as a risk factor, and family, socio-environmental, and temperamental variables as protective factors. Consequently the question to treat or not to treat is not that simple. If the child becomes unmanageable during treatment or experiences (extreme) anxiety in the days surrounding the procedure due to the laser treatment or the use of anesthetics further therapy should be re-evaluated. Treatment of the PWS might even be delayed until it is important for the child itself, without pressure from parents or a referring doctor.

References


Table 1  Treatment characteristics.

<table>
<thead>
<tr>
<th></th>
<th>With general anaesthesia n=13</th>
<th>Without general anaesthesia n=28</th>
</tr>
</thead>
<tbody>
<tr>
<td>Visits per patient¹</td>
<td>12 (4)</td>
<td>16 (6)</td>
</tr>
<tr>
<td>Time per visit (minutes)¹²</td>
<td>85 (44)</td>
<td>24 (11)</td>
</tr>
<tr>
<td>Number of pulses per visit¹</td>
<td>709 (606)</td>
<td>123 (105)</td>
</tr>
<tr>
<td>Average number of pulses per minute¹³</td>
<td>12 (4)</td>
<td>5 (3)</td>
</tr>
</tbody>
</table>

¹ mean (standard deviation), ² preparation time included, ³ preparation time excluded

Table 2  Parent evaluation flash-lamp-pumped pulsed-dye laser treatment.

<table>
<thead>
<tr>
<th>Parents of children (n=41)</th>
<th>Not agree (%)</th>
<th>Slightly agree (%)</th>
<th>Agree (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>The laser treatment is quite painful</td>
<td>8</td>
<td>50</td>
<td>42</td>
</tr>
<tr>
<td>A few days before the laser treatment my child is getting nervous</td>
<td>52</td>
<td>25</td>
<td>23</td>
</tr>
<tr>
<td>My child shudders to think of the laser treatment</td>
<td>20</td>
<td>42</td>
<td>38</td>
</tr>
<tr>
<td>My child feels stared at with the blue-grey colour of the PWS after treatment</td>
<td>8</td>
<td>2</td>
<td>90</td>
</tr>
<tr>
<td>The laser treatment is not a burden for my child</td>
<td>50</td>
<td>10</td>
<td>40</td>
</tr>
<tr>
<td>It takes too long until my child has completed treatment</td>
<td>15</td>
<td>57</td>
<td>28</td>
</tr>
</tbody>
</table>
Figure 1  Color differences versus treatment of the entire PWS.

Patient 1

Patient 2
† Aged 7, surface area PWS 20 cm², treated between January '92 and February '95.

‡ Aged 10, surface area PWS 150 cm², treated between June '92 and March '95.

O No color measurement. The first measurement was taken before onset of treatment.