On infantile hemangiomas
Hoornweg, M.J.

Citation for published version (APA):
Hoornweg, M. J. (2012). On infantile hemangiomas

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.
The impact of infantile hemangiomas

Marije J. Hoornweg
Martha A. Grootenhuis
Mark J.C. Smeulders
Chantal M.A.M. van der Horst
Introduction

Infantile Hemangiomas (IH) are the most common tumors of infancy. They are recognizable by their typical natural history characterized by two distinct clinical phases: proliferation and involution. The proliferation phase is typical because most IH are not visible at birth, but a red papule or teleangiectasia develops somewhere in the first months after birth. During the first few months of life sudden spontaneous rapid growth takes place up until 18 months of age. In the beginning of the following involution phase, the growth stabilizes and color intensity reduces. The bright red color becomes grey. The consistency will evolve from firm to a soft compressible mass by palpation. The involution phase starts at 12-18 months of age and can continue for the next 5 to 10 years.

Although the subsequence of steady proliferation and complete involution is typical for most IH, in 10% to 20% (1;2) of cases complications arise during the early to mid-proliferation phase, as growth is especially rapid and difficulties can arise suddenly en unexpectedly. The presence of an IH at certain locations poses a serious risk due to bleeding, ulceration or local pressure that hampers proper development of adjacent tissue. Of such latter complications obstruction of the eye with subsequent vision problems (54 to 80% of all periorbital IH (3)) or cartilage damage of the nose or ear are notorious. Ulceration occurs in 15 to 25% of IH in a referral centre especially located on the lips and anogenital area (4;5). In such cases, treatment is preferential and may lead to more parental and patient stress. Similarly, treatment may be pursued after the involution phase if the IH does not resolve spontaneously, as in 6% to 96% of cases some form of sequelae occur, as the remaining fibrofatty tissue can cause excessive skin, the ulcerated wounds may have led to scar tissue or remaining pigmentation and deformed cartilage, also the IH may have caused permanent vision damage. Treatment of the involuted IH will focus on the above mentioned sequelae with the goal to improve the cosmetic or functional result and consists of surgery to remove excessive skin or correct contour deformity, or laser treatment to correct teleangiectasias. Algorithms for treatment of IHs in different phases exist (6;7) and different forms of treatment have been a target of clinical studies (6;8;9). What becomes clear from these studies is that having an IH is felt as a burden for both children and their parents, and treatment may increase the burden as it often involves frequent hospital visits and sometimes painful bandage changes for these young patients. As such, IH have a psychological impact on parents, caretakers and children. Although the psychological impact of IH on child and parents have been widely accepted as an important factor to consider during treatment and clinical follow-up (6) evidence on this subject is anecdotal. The present review aims to evaluate the available evidence on the total impact of having an IH to aid physicians seeing children with IH to anticipate and monitor for signs and consequences in affected children and their parents.
Psychological Impact and Quality of Life

Children with an IH are seldom clinically ill, but their physical deformities may affect their behavior and well-being (10-14). In previous studies it has been shown that children with craniofacial deformities like cleft palate have more difficulties when making new contacts (13;14) and children with giant congenital melanocytic naevi are at increased risk of social behavioral and emotional problems (12). The major difference between these physical deformities and IHs is their typical natural history. IHs are not permanent. The child’s body image is usually poorly developed until the age of three, and during the entire proliferation phase it mainly seem to be parents that are shocked by the unexpected IH and its subsequent growth in their otherwise completely healthy child, This may have an impact on children and parents. The presentation of IHs has an enormous variety due to localization, size, possible complicated course and possible treatment. A one centimeter, non-complicated IH on the torso has a different influence compared to a 10 centimeter ulcerating peri-orbital IH causing pain and amblyopia. Therefore we expect diversity in impact of IHs.

Impact is considered in terms of the following domains: quality of life, psychological consequences and emotional impact. The concept of health related quality of life (HRQOL) has been introduced for evaluation the overall consequence of a disease for patients. Measurement of HRQOL is based on the assessment of the individual’s perception of the impact of medical and nonmedical issues regarding physical, mental and social functioning that has been used in the last decades to evaluate impact of several diseases and treatments. Several measurement tools exist, and have been validated (15-18). Both questionnaires and interviews are used in the studies. As such, HRQOL may be a helpful tool in understanding the impact of IHs. However, only a few valid tools exist to measure the HRQOL, especially for children with disfigurements and their families. In the present review we include all aspects of HRQOL of IHs.

Methods

We reviewed the articles concerning Health Related Quality of life, and social and psychosocial consequences of IHs. Searches were conducted in the National Library of Medicine’s PubMed database as well as the Cochrane Library using the keywords IH(s), (health-related) Quality of Life, psychosocial (impact), (parental) adaptation. All potentially eligible studies were retrieved and evaluated according to the inclusion- and exclusion criteria (Table 1, See page 102)
Chapter 8

Table 1 Inclusion criteria for the articles.

| Articles after 1982; introduction of terminology by Mulliken |
| Clear definition (according to article Mulliken 1982) used of infantile hemangiomas |
| English language of abstract |
| Key words: (infantile) hemangioma(s), (health-related) Quality of Life, psychosocial (impact), (parental) adaptation. |

Results

We found 11 potentially eligible studies that were retrieved (19-29). (Table 2). Study number 2, 3, 8, 10 and 11 were excluded for further review in this article. Article 2, 8, 10 and 11 were (incomplete) summaries of psychosocial impact of vascular anomalies that mainly provided practical suggestions without reporting actual data, article 3 included all vascular birthmarks without separating the IHs, and article 8 used a too wide definition for IHs. Table 2 lists data from all the retrieved articles. These studies included a total of 322 cases and after exclusion of article 2, 3 and 8 a total of 303.

Table 2 Summary of the literature review on impact of infantile hemangiomas on children and their parents.

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Journal</th>
<th>Age(yrs)children</th>
<th>Only IHs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Williams, EF</td>
<td>2003</td>
<td>Arch Facial Plast Surg</td>
<td>0-8</td>
<td>Yes</td>
</tr>
<tr>
<td>Weinstein, JM</td>
<td>2005</td>
<td>Lymp res and Bio</td>
<td>-</td>
<td>No</td>
</tr>
<tr>
<td>Sandler, G</td>
<td>2009</td>
<td>Aust Fam Physician</td>
<td>-</td>
<td>No</td>
</tr>
<tr>
<td>Hoornweg, MJ</td>
<td>2009</td>
<td>JPRAS</td>
<td>1-15</td>
<td>Yes</td>
</tr>
<tr>
<td>Dieterich-Miller, CA</td>
<td>1992</td>
<td>Pediatr Dermatol</td>
<td>3-5</td>
<td>Yes</td>
</tr>
<tr>
<td>Tanner, JL</td>
<td>1998</td>
<td>Pediatrics</td>
<td>0.5-8</td>
<td>Yes Only facial</td>
</tr>
<tr>
<td>Kunkel EJ</td>
<td>1994</td>
<td>Psychosomatics</td>
<td>0.83-9</td>
<td>Yes</td>
</tr>
<tr>
<td>Dieterich-Miller, CA</td>
<td>1992</td>
<td>Child health care</td>
<td>-</td>
<td>No</td>
</tr>
<tr>
<td>Snyder, H</td>
<td>2010</td>
<td>Cleft Palate-Craniofacial J</td>
<td>2-3</td>
<td>No</td>
</tr>
<tr>
<td>Cohen, SG</td>
<td>2005</td>
<td>Adv Nurse Pract</td>
<td>-</td>
<td>No</td>
</tr>
<tr>
<td>Lande, RG</td>
<td>2001</td>
<td>Facial Plastic surgery of North America</td>
<td>-</td>
<td>No/??</td>
</tr>
</tbody>
</table>
Overall, findings are difficult to interpret. An enormous variety of presentation of IHs, due to size, location and possible complications, led to an inhomogeneous group of children with a IH. Subdivisions were made based on visibility and complicated course of the IH (22;27;29). All included studies are hospital-based groups. Compared to population-based groups, hospital-based groups included more IHs located 1) in the head/neck region, 2) subcutaneously and/or with a complicated course (30).

Evaluation was done using different tools. Of those tools, the different questionnaires used by Dieterich-miller (21), Kunkel (23) and Hoornweg (22) were all validated. They were completed by interview, in person, by phone or written. In Table 2 the questionnaires are listed. Tanner et all (27) performed interviews of the parent in ethnographic in style, intended to understand the parents’ experience from their point of view. A qualitative analysis was undertaken and parental statements were coded to a list of core categories; 1) parental emotion and adaptation, 2) experiences with public reactions, 3) parent-child interactions and 4) expressed (dis)satisfaction with medical care. Williams et al (29) performed an interview by phone using a 38 point

<table>
<thead>
<tr>
<th>Children (C) and/or Parents (P)</th>
<th>Type of research / instrument</th>
<th>Number of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>P</td>
<td>Interview by telephone using a 38-point questionnaire.</td>
<td>39</td>
</tr>
<tr>
<td>-</td>
<td>Review on quality of life in vascular anomalies.</td>
<td>-</td>
</tr>
<tr>
<td>P</td>
<td>Interview in person with open ended questions</td>
<td>19</td>
</tr>
<tr>
<td>C + P</td>
<td>1. Medical determinants form, 2. Quality of life was measured with the TNO-AZL Preschool Quality of Life questionnaires compared with a control group. 3. A specific IH questionnaire based on clinical experience.</td>
<td>201</td>
</tr>
<tr>
<td>C + P</td>
<td>The Joseph’s preschool and primary self-concept test (JPPST) and Achenbach child behavior checklist (CBCL) compared with a control group and an informal interview with open-ended questions.</td>
<td>19</td>
</tr>
<tr>
<td>P</td>
<td>Interviews in person (13) or by phone (12) in ethnographic style.</td>
<td>25</td>
</tr>
<tr>
<td>P</td>
<td>A psycho-educational/self-help pilot group for parents (6 couples) using the Mental Health Inventory (MHI), a 44-item questionnaire.</td>
<td>6</td>
</tr>
<tr>
<td>-</td>
<td>Provision of practical suggestions</td>
<td>-</td>
</tr>
<tr>
<td>C</td>
<td>Child Behavior Checklist/2-3</td>
<td>13</td>
</tr>
<tr>
<td>-</td>
<td>Provision of practical suggestions</td>
<td>-</td>
</tr>
<tr>
<td>-</td>
<td>Provision of practical suggestions</td>
<td>-</td>
</tr>
</tbody>
</table>
questionnaire. Ten questions contained the attitudes that the family and child had toward the IH and five questions covered the emotional response to treatment. These 15 questions could be responded in a 5 point scale (strongly agree, agree to strongly disagree) and assessed the emotional and psychological effects that the IH had of the family. The origin of this list was not mentioned. Hoornweg et al (22) had a similar ‘IH specific questionnaire’. The items were developed from clinical experience by a team of researchers, dermatologist, plastic surgeons and psychologists. All studies collected IH data such as natural history, phase of the IH, size, location, and possible complication and treatment interventions.

No significant differences were found between parents and children with and without IHs with regard to impact of IH on the long term. The Child Behavior Checklist (CBCL), which was used by Snyder and Dietrich-Miller (21;26) revealed no difference between children with a IH and a control groups. Parental ratings of children were also within the range of acceptable behavior in the Joseph’s preschool and primary self-concept test (JPPST) (21). Even the Quality of life, which was measured with the TNO-AZL Preschool Quality of Life questionnaire for children (TAPQoL) and the TNO-AZL Children’s Quality of Life questionnaire, parent- (TACQoL-PF) and child form (TACQoL-CF) showed no difference with a control group (22). Kunkel et all (23) used the General Well Being Scale of the Mental Health Inventory (MHI) showed in their group of 6 couples a significant difference compared with a control group. The ‘psychological well-being’ subscale was statistical significant though all other subscales (anxiety, depression, loss of emotional control, general positive effect, life satisfaction, psychological distress) were not significant.

The specific questionnaires however including impact on parents and interviews of parents did show a trend toward increased fears, anxieties and feelings of disbelief (21-23;25;27;29). Wiliams et all (29) used an interview by telephone using a 38-point questionnaire covering the child’s birth history, natural history of the IH, physician encounters, treatment interventions and the family’s and child’s emotional attitudes toward the IH and related treatment (the last item were 15 questions with answer options on a 5 point scale). Tanner (27) also used interviews by telephone and in person. They were ethnographic in style. By qualitative analysis all interviews were coded to a list of core categories; 1. Parental emotion and adaptation: 2. Experiences with public reactions: 3. Parent-child interactions: and 4. Expressed satisfaction/dissatisfaction with medical care. Sandler (25) and Dietrich-miller (21) used open ended questions in an interview in person. Hoornweg et al (22) used a specific IH questionnaire based on clinical experience. The overall outcomes show a negative emotional effect of the IH on the child, their parent and rest of the family. Reactions from relatives and friends were often acceptable. Comments from other young children but especially strangers did
not remain so innocent. The question of child abuse is unfortunately sometimes heard, exact percentages are not known. There was a trend of visible IH to have greater impact (21;22;27). Though Kunkel et all (23) could not correlate site of the IH with increased parental distress. Again this was a very small group; 6 children.

**Discussion**

Although the literature on management of IH is ample, there is a lack of knowledge on the psychological sequelae of this disease. It is important for physicians seeing children with IHs to anticipate and monitor for signs of psychosocial impairment in both children and their parents.

Small differences in self esteem and feelings of grief are shown, but none of the studies show significant differences between children with and without an IH at older age. There are, however, two exceptions to this conclusion. The first exception is that children with visible IHs tend to have less self esteem and more psychosocial problems. Although samples are small, they could indicate that psychosocial problems may be related to size and visibility of the IH. The second exception that needs attention is that parents themselves, acknowledging that their children were too young to appreciate their own condition (22;29), had difficulty in coping with their own worries and with reactions from the community. The impact of IHs on parents was big as was shown from the reactions that parents gave in the IH-related questionnaires. This resulted from fear, feelings of guilt and most importantly negative reactions from family, friends or outsiders. Being accused of child abuse is, unfortunately, an often heard comment. Again visibility is of great importance for these reactions and differences between non-visible and visible IHs were evident. Parents of children with a IH with a complicated course seem also to be at risk for more feelings of guilt and fear. This could be explained by the troubles of numerous hospital visits, of seeing their child in pain of an ulcerated IH or worries for e.g. the vision of the child. Several studies have shown that parents can have major QOL problems related to the illness of their child (31) and this should not be neglected.

The impact on children with visible and big IH and the impact of IH on the parents do show the eligibility to advocate a careful management program around IHs. All included studies point out the importance of a good relationship between parents, child and physician. Depending on the complexity of the IH this could be the general-practitioner, or specialist. Most IH need – due to their natural history – no treatment at all. This ‘wait and see’ policy however must be carefully thought out and discussed with the parents of the child. Some parents harbor unusual, almost irrational, expectations. Extended information of the natural history, photographs of long-term outcomes can help set realistic expectations. A good relationship between parents and doctor is absolutely required. Important is the consistently and clarity toward parents and child about expected course, possible complications and necessary treatment.
Chapter 8

There is a need for follow up and to adequately monitor the psychosocial impact that these lesions could have, and to provide resources to those who might benefit from psychological intervention. Special care needs to be given to visible IHs or IHs with a complicated course. These children and, equally or sometimes even more important, their parents need extra attention in helping them to cope and adjust to the disease. This implicates screening of children and their parents ‘at risk’ in clinical practice, and more attention for family functioning. Monitoring, identifying and discussing HRQOL issues by providing patient reported outcomes (PRO’s) in clinical practice should be considered. Studies suggest that discussing PRO’s improves communication between physicians and patients and facilitates early recognition of HRQOL problems (32;33). The use of a web-based program contributes to an improved use of PRO’s in clinical practice. Children or their parents (depending on the age of the child) can complete the HRQOL questionnaires at home and pediatricians can retrieve the PRO’s of these children directly from this website during the doctor’s visit (34). Apart from a generic HRQOL questionnaire, disease-specific questions about body image and self esteem could be added.

In conclusion, we underscore the importance of evaluating the psychological role that IHs may have on the entire family unit.
Reference List

Chapter 8


