Giant congenital melanocytic naevi: definition, malignant transformation and treatment modalities
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Citation for published version (APA):

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Results of early curettage of giant congenital melanocytic naevi; a report of 8 cases and a review of the literature

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Sillevis Smitt JH
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Van der Horst CM

Published in:
Abstract

**Background.** Aim of this study was to investigate the long term cosmetic and oncologic results occurring after early curettage of giant congenital melanocytic naevi (GCMN).

**Methods.** Neonates with GCMN treated with curettage within four weeks of age and a minimum follow-up of two years were evaluated at the outpatient department. Scar formation was evaluated by means of the Patient and Observer Scar Assessment Scale (POSAS). Biopsy specimen were analysed.

**Results.** In nine years eight neonates were treated. Mean follow-up period was 5.6 years. Six (75%) patients developed re-pigmentation of the curetted skin, hypertrichosis returned in five cases. One patient developed hypertrophic scars, all others formed good scars. None of the patients developed a melanoma. Biopsy specimen showed naevus cells in the skin after curettage.

**Conclusion.** In 50% of the patients curettage is followed by severe re-pigmentation. Since this treatment does not remove all naevus cells, long term follow-up remains important to detect malignant transformation.
Introduction

By definition, congenital melanocytic naevi (CMN) are present at birth, all other being acquired. Divisions into size groups are clinically relevant because the problems associated with CMN vary according to size.¹ The larger lesions are obviously more disfiguring and pose a greater threat to the psychological well-being of the patient. Also, the risk of malignant transformation varies with size.¹ Therapeutic options depend on size and may pose major management problems. Ideally, the excision removes all naevus cells. Unfortunately, this is not always feasible due to the size and deep spread of Giant CMN (GCMN); in those instances, superficial ablative techniques such as laser therapy, dermabrasion or curettage are indicated. The primary aim of curettage treatment is the improvement of the cosmetic result; also, it is hoped that the reduction of numbers of naevus cells reduces the risk of malignant transformation.

Aim of this study was to investigate the longterm cosmetic and oncologic results of curettage of GCMN.

Operative technique

Johnson was the first to observe the phenomenon of a natural cleavage plane in newborns between the upper dermis, containing the majority of naevus cells and melanocytes, and the deeper dermis within the naevus.² He reported a case of a newborn with a CMN of the scalp being delivered with forceps. Part of the naevus was accidentally removed with the instrument and the resulting skin defect healed spontaneously. On this evidence, Johnson treated the remaining naevus with dermabrasion, and pigmentation did not return with a follow-up of 17 years. In 1987 Moss proposed a technique of curettage before six months of age but preferably as early as is feasible.³ Curettage is said to become progressively more difficult to perform as the dermal component of the naevus becomes more coherent with increasing age.

Under general anaesthesia, curettage might be performed as an one-stage procedure in neonates. With a sharp curette, the naevus is scraped from its centre to the periphery of the naevus in the direction of the maximum relaxation. The cleavage plane is more easy to find when curettage is performed early in life, preferably within two weeks.⁴,⁵

Patients and Methods

During a nine-year period, starting in 1996, a subset of 93 patients with GCMN were treated at the department of Plastic, Reconstructive and Hand Surgery of the Academic Medical Centre of Amsterdam in the Netherlands. GCMN were defined as CMN larger
than one percent in the face/neck region, and more than two percent elsewhere on the body.\(^6\)

Of these 93 patients with GCMN, covering 1 to 30% of the total body surface, ten patients were treated with curettage by the last author within one to four weeks after birth. Curettage was only indicated for GCMN located on the back, abdomen, trunk, thorax or extremities which could not be excised with primary wound closure after tissue expansion. All patients were photographed pre- and postoperatively by a medical professional photographer.

For this study we only included patients with a minimum follow-up period of two years, so our study population consisted a total of eight patients. Distribution of location of GCMN over the body, age at treatment and sex are shown in Table 1. The newborns underwent one or two sessions of curettage, depending on the size of the lesion. We limited the curetted area to less than 15% of the total body surface at any procedure, in order to limit the blood loss. Pre- and postoperative haemoglobin (Hb) concentration was determined in order to assess the possible need for blood transfusion. If, mainly at the borders of the GCMN complete curettage could not be performed, the lesional periphery was further excised. Per-operative biopsies were taken for histopathologic evaluation after curettage. The patients were hospitalised for at least one week postoperatively, to optimize pain medication and wound care. Pain medication was given according to our hospital protocol and depended on the weight of the patient. The first few days the wound was covered with Opsite®, after this silversulfadine (Flammazine®) crème was used until re-epithelialisation was realised. No systematic antibiotics were given.

The patients included for this study were invited once more to our outpatient department for clinical follow-up and especially for evaluation of all aspects of scar formation after curettage. For the latter we obtained the validated Patient Observer and Scar Assessment Scale (POSAS ).\(^7\)

**Table 1. Distribution of age, sex and location of GCMN**

<table>
<thead>
<tr>
<th>Sex</th>
<th>Location GCMN</th>
<th>Age start</th>
<th>Location curettage</th>
</tr>
</thead>
<tbody>
<tr>
<td>f</td>
<td>thigh, abdomen, buttocks, back</td>
<td>4 weeks</td>
<td>back, buttocks</td>
</tr>
<tr>
<td>m</td>
<td>thigh, urogenital</td>
<td>3 weeks</td>
<td>thigh</td>
</tr>
<tr>
<td>f</td>
<td>thorax, back, legs</td>
<td>2 weeks</td>
<td>thorax</td>
</tr>
<tr>
<td>f</td>
<td>leg</td>
<td>1 week</td>
<td>leg</td>
</tr>
<tr>
<td>m</td>
<td>buttocks, thigh, urogenital</td>
<td>2 weeks</td>
<td>buttocks</td>
</tr>
<tr>
<td>f</td>
<td>thigh, buttocks, urogenital, leg</td>
<td>4 weeks</td>
<td>buttocks</td>
</tr>
<tr>
<td>m</td>
<td>abdomen</td>
<td>2 weeks</td>
<td>abdomen</td>
</tr>
<tr>
<td>m</td>
<td>urogenital, thigh</td>
<td>1 week</td>
<td>thigh</td>
</tr>
</tbody>
</table>
Scar assessment

The POSAS consists of two scales; the observer scale and the patient scale. When the child was eight years or younger, the parents scored the scale. Both scales contain six items. The two observers rated scar (resulting from curettage) vascularity, pigmentation, thickness, relief, and surface area and pliability. The patient scale contains six questions applying to pain, itching, colour, pliability, thickness, and relief. Each of the six items on both scales has a 10-step score, with 10 indicating the worst imaginable scar or sensation. The total score of both scales consists of adding the scores of each of the six items range 6 (normal) to 60 (worse scar). In addition the observer and patient gave their overall opinion on the appearance of the scar, again a 10-step scale.

Because re-pigmentation was an important outcome in this study, we listed this item separately in Table 2.

<table>
<thead>
<tr>
<th></th>
<th>observer POSAS</th>
<th>patient POSAS</th>
<th>Re-pigmentation</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>15 / 4</td>
<td>22 / 4</td>
<td>8</td>
</tr>
<tr>
<td>2</td>
<td>19 / 5</td>
<td>19 / 5</td>
<td>8</td>
</tr>
<tr>
<td>3</td>
<td>21 / 5</td>
<td>24 / 4</td>
<td>6</td>
</tr>
<tr>
<td>4</td>
<td>15 / 3</td>
<td>20 / 4</td>
<td>3</td>
</tr>
<tr>
<td>5</td>
<td>23 / 6</td>
<td>24 / 6</td>
<td>8</td>
</tr>
<tr>
<td>6</td>
<td>12 / 3</td>
<td>11 / 3</td>
<td>2</td>
</tr>
<tr>
<td>7</td>
<td>19 / 6</td>
<td>24 / 5</td>
<td>9</td>
</tr>
<tr>
<td>8</td>
<td>14 / 5</td>
<td>16 / 5</td>
<td>5</td>
</tr>
</tbody>
</table>

POSAS scores: the first number of the POSAS Observer and the first number of the POSAS Patient represents the total amount of adding the six separate items each with a 10-step score. For the observers these are vascularity, pigmentation, pliability, thickness, surface and relief. For the patients the six items are pain, itching, color, pliability, thickness and relief. The lowest total score is six and reflects with normal skin, whereas the highest score 60 reflects the worst imaginable scar. The second number of the POSAS Observer and Patient represent the overall opinion on the appearance of the scar. Again a 10-step scale was used in which 10 corresponded to the worst imaginable scar. The last item represents the re-pigmentation scores: 0-10; 1-4 mild re-pigmentation, 5-7 moderate, 8-10 severe.

Results

Seven out of the eight patients were seen in our outpatient department. For geographic reasons in one patient the questionnaires were send by mail and the photographs were taken in another academic centre. Our mean follow-up period was 5.6 years (range 2.3 years to 9.5 years). None of the patients needed blood transfusion after the curettage and the haemoglobin (Hb) drop was minimal.
Table 2 shows the sum of the POSAS scores was almost completely made up by the item pigmentation. As seen in Table 2 these numbers vary between 15 and 24, which is a moderate scar. Due to the small number of patients, statistical analyses could not be performed. In addition, judgement of parents and observers was comparable (Table 2). One patient developed hypertrophic scars due to infection, but all others formed good, pliable and soft scars after curettage. Two patients had mild re-pigmentation, two moderate and four patients had extensive re-pigmentation (Table 2). Re-pigmentation first appeared two months after curettage, with a mean of three months. Patients in figures one and two show that severe re-pigmentation. Hair growth returned in five cases. Histopathologic biopsy specimen post curettage showed that intradermal pigmented nevus cells were present in all cases. None of our patients developed a malignant melanoma or non-melanomatous malignant tumor during follow-up.

Discussion

Histology

Essentially, CMN show the histological features of the far more common acquired naevi, the main difference being their larger size and the more extensive spread of naevus cells in the skin superficial dermis and along adnexae. Early studies suggested that the congenital naevus does not yet involve the deep reticular dermal tissue at birth but that this develops during the first year of life.8-10 Others noticed the importance of nevus size in determining the extent of dermal involvement already in the early postnatal period.11-13 Curettage of GCMN at a very early age has been based upon the supposedly superficial location of naevus cells and the finding of an easy cleavage plane between the upper and lower dermis during first weeks of life. Moss noted that this cleavage plane is not present in the normal skin and that it is not located at the junctional area of the naevus.3 Mark et al. mentioned in 1973 that in the reticular dermis and subcutis, naevus cells in CMN are usually disposed as single cells between collagen bundles and fat cells, but also found naevus cells in the hair follicles which are surrounded by collagen sheaths with anchors in the subcutis, and in erector pili muscles.14

In fact, in our experience the histological features of GCMN in infancy are variable, and may include the presence of large numbers of naevus cells arranged in densely cellular masses within the dermis. Such cellular masses, which contain little collagenous stroma, present a plausible explanation for the plane of cleavage found by the surgeon, since in contrast to normal collagen-rich reticular dermal tissue, such naevus cell masses provide little resistance to the physical trauma of the surgeon. The plane of cleavage would thus lie in the level where cellularity of the naevus is high; this is in accordance with
the presence of many naevus cells in the post-resection biopsies mentioned above. The disappearance of the “cleavage” plane after a few weeks of life can be explained by the gradual emergence of more collagen-rich stroma within the main mass of the naevus. Recently, the explanation for malignant transformation for GCMN has been sought in genetic mutations (BRAF oncogene and NRAS mutations). Michaloglou showed in vitro and in vivo in our own CMN BRAF (V600E) expressing melanocytes. Nakato et al. detected BRAF mutation in acquired naevi and small CMN. Larger congenital naevi tend to harbour NRAS mutations rather than BRAF mutations. Very recently De Raeve did not show oncogenic BRAF mutation in cured GCMN and excised MCMN, which could support her idea of removing the chance of malignancy with curettage. There is no evidence that the risk of malignant transformation is different between small and medium-sized CMN, but GCMN are definitely associated with increased risk of developing melanoma. This difference could, however, be accounted for by the fact that there are so many more naevus cells in a large congenital naevus.

De Raeve mentioned in her study that naevus cells in the superficial component of the GCMN, were more proliferative, and this component was more vascular compared with its deep component and with MCMN, which were not removed after curettage. However, curettage or indeed any trauma inducing a wound healing response, deeper naevus cells can be re-activated to proliferate. It is conceivable that such re-activated deep naevus cells which have recommenced proliferation can transform to malignancy, although the chance of this occurrence is probably unknown.

Treatment results

Our study for the first time used a validated scare scale to evaluate the results of treatment. In only one patient re-pigmentation did almost not occur, and four patients (50%) developed severe re-pigmentation. Except for the patient without re-pigmentation, all other patients underwent additional surgery because it was not possible to perform satisfactory complete curettage at the borders. All parents agreed that the postoperative

<table>
<thead>
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<th>Author</th>
<th>patients</th>
<th>location</th>
<th>age at treatment</th>
<th>results</th>
</tr>
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<tbody>
<tr>
<td>Moss</td>
<td>10</td>
<td>5 bathing trunk</td>
<td>2 wk - 6 months</td>
<td>3 re-pigmentation, 6 satisfactory</td>
</tr>
<tr>
<td>de Mey</td>
<td>6</td>
<td>unknown</td>
<td>unknown</td>
<td>5 satisfactory</td>
</tr>
<tr>
<td>Kay</td>
<td>1</td>
<td>bathing trunk</td>
<td>&lt; 2 weeks</td>
<td>not satisfactory</td>
</tr>
<tr>
<td>Casanova</td>
<td>9</td>
<td>back, abdomen, buttocks</td>
<td>&lt; 7 weeks</td>
<td>6 satisfactory</td>
</tr>
<tr>
<td>De Raeve</td>
<td>19</td>
<td>back, scalp, arm, leg, abdomen</td>
<td>&lt; 2 weeks</td>
<td>6 moderate and 3 severe repigmentation</td>
</tr>
<tr>
<td>Zaal</td>
<td>8</td>
<td>back, abdomen, buttocks, legs</td>
<td>&lt; 4 weeks</td>
<td>4 severe re-pigmentation</td>
</tr>
</tbody>
</table>
period is very difficult and must not be underestimated, especially if the curettage area is in or near the diaper region.

The first treatment results of curettage were reported by Moss et al. in 1987.\textsuperscript{3} (Table 3). Moss stated that it is more difficult to find a cleavage plane at the age of six months and advised to perform curettage before that age.

De Mey and colleagues reported satisfactory results of five out of six children.\textsuperscript{19} In this study, the age of patients at treatment and the size and location of CMN are not specified. Kay published a case-report of an unsuccessful neonatal curettage of a classic “bathing trunk” GCMN within two weeks of life.\textsuperscript{20} The procedure was stopped because there was no easy plane of cleavage. This is also in line with our experience, although treated during first weeks of life, neonates with thick and hairy GCMN do not always have an easy and clear cleavage plane and curettage can not be performed properly. In literature and also in our study population the patients with elevated and hairy GCMN, showed hypertrichosis and re-pigmentation after curettage.

Casanova et al. reported data on nine neonates.\textsuperscript{21} Treatment was deemed ineffective in three patients, in whom there was no cleavage plane. They mentioned that naevus cells persist in the deep dermal layers and may come to the surface, such that close monitoring is needed long after curettage.

De Raeve published in a period of 14 years, a total amount of 19 neonates treated by curettage.\textsuperscript{4,5,17} She is a fervid supporter of curettage and advocates to perform the curettage during the first two weeks of life. In her last report De Raeve presented data which support the idea that curettage of GCMN in neonates has the potential for lowering the risk of developing cutaneous melanoma by not only obtaining an important numerical reduction of naevus cells but also removing the ‘active’ melanocytes.\textsuperscript{17} However, the surgical tissue trauma can to some extent re-activate naevus cells, which is macroscopically seen by the presence of re-pigmentation, and curettage is therefore best regarded first and foremost as a cosmetic treatment.

Apart from curettage, there are two other incomplete removal treatments of GCMN; dermabrasion, and laser therapy.\textsuperscript{22,23} The disadvantage of dermabrasion is that it is followed by re-pigmentation if preformed (too) superficially, but produces hypertrophic scares when performed too deeply. The same is true for laser treatment which has been justified on the basis of the superficial position of the majority of the pigment in GCMN. Again the deeper naevus cells in the deep dermis and subcutaneous fat remain.\textsuperscript{20} Also, the naevus cells are repeatedly damaged by the laser treatment and thereby repeatedly inducing re-activation of the deeper located naevus cells. Among Dutch dermatologists there is now consensus that pigmented lesions should not be treated with laser. Nowadays, even the group of Ostertag does not use the laser for treatment of GCMN anymore because of severe repigmentation.\textsuperscript{23}
In our opinion, any kind of incomplete treatment of congenital naevi is indicated only when complete excision is impossible. The reduction of the chance of malignant transformation is the key argument why surgical excision remains the standard of care.

In conclusion, early curettage resulted in our series in 50% severe re-pigmentation in the long term. The disappearance of the “cleavage” plane after a few weeks can be explained by the gradual emergence of more collagen-rich stroma within the main mass of the naevus. The presence of re-pigmentation shows that it is unrealistic to aim at removal of all naevus cells with curettage. Long term follow-up and close monitoring of pigmentation remains essential to detect malignant transformation at an early stage.

**Acknowledgments**

The authors like to thank PPM van Zuijlen for providing the POSAS scale.
**Figure 1a.** GCMN of a four weeks old girl covering most part of the back and diaper region (25% of the total body surface). Patient 1 in table I and II

**Figure 1b.** Results of curettage four months postoperatively performed at three weeks of age. Curettage at the borders was not possible. Only islands of pigmentations returned.

**Figure 1c.** Results of curettage after a follow-up of five years. Note the severe re-pigmentation and hypertrichosis.
Figure 2a. Results of curettage three weeks postoperatively performed at one week of age. No pigmentation has returned. Patient 2 in table I and II.

Figure 2b. Results of curettage four months postoperatively. Islands of pigmentations are visible.

Figure 2c. Result of curettage two years postoperatively with severe re-pigmentation.
References


