

## Research Article

# Feasibility and Long-Term Results of Versajet® Hydro-surgery Technique as First Treatment of Neonates Affected by Giant Congenital Melanocytic Nevi (GCMN): A Single Center Ten-Year Experience

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## Abstract

Giant Congenital Melanocytic Nevus (GCMN) is defined as a melanocytic congenital lesion that will reach a diameter  $\geq 20$  cm in adulthood, and that presents as a brown lesion, with flat or mammilated surface, well-demarcated borders and hypertrichosis. It may associate with severe complications such as malignant melanoma, neurocutaneous melanosis, and psychosocial disorders. Treatment may include surgical and non-surgical procedures, psychological intervention and/or clinical follow-up. The only absolute indication for surgery is the development of a malignant neoplasm on the lesion. A retrospective observational study was conducted including all newborns (6 patients, median age 14 days, range 8-25 days) that underwent Versajet® hydrosurgery during the first month of life for GCMN at our Center in the period between November 2009 and December 2020. Treatment modalities, early and late complications, cosmetic results (with an average five years follow-up), malignant transformation and psychosocial impact were examined. The effectiveness of Versajet® hydrosurgery varied according to the site and extension of the lesions. Compliance to treatment was close to 100%. This study aimed to assess the short and long-term results of Versajet® hydrosurgery as initial treatment of GCMN in newborns. It showed to be safe and effective both at short- and long-term follow-up. However, patients reported a negative impact of the residual disease on their social life. Neurocutaneous melanosis is considered the most challenging associated condition to manage.

**Keywords:** Giant Congenital Melanocytic Nevi (GCMN); Melanoma; Neurocutaneous melanosis; Versajet® hydrosurgery

## Introduction

Giant Congenital Melanocytic Nevi (GCMN) is pigmented lesions of the skin existing at birth but not inherited. They represent benign proliferations of abnormal melanocytes caused by a somatic mutation of melanocytic cells during embryofetal development, between the 5<sup>th</sup> and 24<sup>th</sup> weeks of gestation [1]. It is believed that a morphological error occurs in the neuroectoderm during embryogenesis, leading to unregulated growth of melanoblasts, the precursor cells of melanocytes [2], derived from the neural crest and then migrated to different locations including skin, mucosa, eyes, meninges. To be defined as giant, their minimum diameter should be equal to 1% of the total body surface, if localized to the head, or to 2%, if located in other parts of the skin [3]. They can affect any region of the body but their most frequent location is the lower back, followed by limbs

and head [4]. GCMN are rare lesions: the birth frequency ranges from about 1:20000 to 1:500000 newborns [2]. GCMN usually present as brown lesions with well-defined borders and hypertrichosis [5].

The main concerns for GCMN were about 1) their potential risk of degeneration in malignant melanoma, 2) the association with neurocutaneous melanosis and 3) possible cosmetic disfigurement with consequent aesthetic/psychosocial problems.

The lifetime risk of melanoma in patients with GCMN range from 3% to 8% [6,7]. The 50% of diagnosed cases of melanoma arises in the first three years of life. Risk factors for malignant transformation include size ( $>20$  cm), presence of satellite lesions, and early onset [8]. Cutaneous melanoma in those patients with GCMN presents some peculiarities; unlike other melanomas that arise in the epidermis, indeed, the melanocytic neoplasia associated to GCMN, in up to two thirds of cases, occurred in the dermis or in deeper layers [9,10]. One of the serious complications of GCMN is Neurocutaneous Melanosis (NCM), a rare syndrome with just over 100 symptomatic cases described in the literature, characterized by deposits of benign or malignant melanocytic cells along the leptomeninges, the spinal cord, and the cerebral cortex or within the brain parenchyma that typically develops within the first 2-3 years of life. Although the location of these cells is similar to that occurring under physiological conditions (in up to 85% of individuals), the extent of infiltration differentiates these patients from the normal population [1,11-13]. Poor prognosis with high mortality rate affects symptomatic patients. Symptoms of NCM may be related to increased intracranial pressure, mass lesions, or spinal cord compression.

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The diagnostic criteria for NCM currently accepted are those of Kadonaga & Frieden [13]:

1. Presence of one GCMN or three or more Congenital Melanocytic Nevi (CMN), associated to meningeal melanosis or meningeal melanomas.
2. No evidence of cutaneous melanoma, except in patients in whom meningeal lesions are histologically benign.
3. No evidence of meningeal melanoma, except in patients with skin lesions subjected to histological examination, and confirmed as benign.

The incidence of NCM in high-risk children has been estimated to range from 17% to 41% [14] and the presence of neural melanosis represents itself a risk factor for melanoma. Some cases of depigmentation have been reported around the nevus region (halo phenomenon) and/or in skin areas distant from it, interpreted as an autoimmune phenomenon [15-17].

Nevus cells presence may also lead to the derangement of the normal skin architecture, leading to cutaneous fragility, erosions and superficial ulcerations [18-20]. Furthermore, smaller pigmented lesions scattered over the skin surface ("satellite lesions") are common, with an incidence of approximately 78% of the cases [20-22]. It's widely recognized worldwide that the risk of complications is proportional to the diameter of the nevus [11,20].

Lesions evolution is very variable and, sometimes, unpredictable: over time, indeed, GCMN can undergo a process of darkening or lightening, develop a more heterogeneous or homogeneous pigmentation, increase hair growth, acquire a more irregular surface or, more rarely, spontaneously regress [23-25].

GCMN represent a great reconstructive challenge for paediatric and plastic surgeons: the risk of degeneration early in life would impose a surgical radicality, often impossible to achieve due to GCMN extent and because developing cerebral melanoma is at least as high as that of cutaneous melanoma. To date, as stated in the English literature, the indication for treatment of CMN has been largely limited to the aesthetic improvement of the patient's appearance as well as removal of suspicious portions of CMN [7].

There are two surgical approaches to GCMN: 1) full thickness excision and reconstruction (serial excision, tissue expansion, local flaps and grafting) and 2) non-excisional procedures. During the first two weeks of life the skin layers are still well separated and the melanocytes are still mainly in the surface layers because they have not yet completed their migration to the deeper layers of the skin: this particular feature makes these cells easy to remove in the first month of life by different non-demolition treatments such as dermoabrasion, curettage, laser or the most recent hydro-surgery technique. There are as yet no prospective studies or systematic retrospective analyses in terms of long term Versajet® hydrosurgery treatment outcomes, both in terms of safety, effectiveness and parental satisfaction.

The aim of our study was to fill this void by assessing the feasibility and distant results of Versajet® hydrosurgery technique as first treatment of neonates affected by GCMN.

## Materials and Methods

Between November 2009 and December 2020, we treated 6 newborns for GCMN in the Pediatric Surgery Unit of the University

Hospital Sant'Anna of Ferrara, with Versajet® Hydrosurgery System. For each patient, demographic and disease-related data with GCMN classification, including recent consensus classification [26], were collected, as well as number of surgical sessions, duration and modality of postoperative resuscitation, effects and outcome after this treatment, side effects and patient's compliance to treatment, long-term outcome and parent's satisfaction. After Versajet® Hydrosurgery all patients were followed for an average duration of five years (range 10-1.5 years). This retrospective study was approved by the ethics committee at Ferrara Sant'Anna University Hospital.

Patients included 3 females and 3 males from different regions of Italy, age of the first hydrosurgical session ranged from 8 to 25 days (median 14 days). All patients underwent a complementary assessment by an early brain and spinal cord Magnetic Resonance Imaging (MRI), with neither contrast nor anaesthesia, combined with a close lifelong follow-up with dermoscopy and incisional biopsies of any suspicious areas during the procedure, and dermatological evaluation once a year to rule out respectively NCM and melanoma. There are no evidence-based guidelines specifying follow-up intervals for CMN of any size: we performed an annual evaluation.

Dermabrasion/curettage of GCMN is first documented in 1987 by Moss. He described the existence of a natural cleavage plane between the superficial dermis, containing the majority of nevus cells, and the deeper dermis, clearly identifiable until the second-third week of life [27]. This technique, using a sharp curette, allows the removal of the most active melanocyte component, reducing the risk of malignant transformation. It must be applied early, within the first three weeks of life, because this is the best time to locate the cleavage plane. Versajet® Hydrosurgery System employs a high-pressure jet of saline water, parallel to the skin surface. This high-speed jet leads a Venturi effect that allows the surgeon to separate and at the same time remove the skin cells [28]. The initial power of the instrument was conventionally set at 3-4 but it can be increased gradually as needed depending of tissue resistance [29]. Sterile saline was heated at body temperature to prevent cooling of the child. Haemostasis was obtained by simple compression or electro coagulation.

Surgery was done under general anaesthesia. If the preoperative dermatoscopic examination revealed suspect areas, they were biopsied before proceeding with hydrosurgery. Mepitel type silicone dressing coated with Connettivina cream was immediately applied over all shaved skin areas. After hydrosurgery the newborns were managed in the neonatal intensive care unit as severely burned patients, with fluid replacement and frequent dressing changes under light sedation, until the wounds have healed.

### Case 1

Our first patient, D.I.E., was a newborn girl with a giant congenital melanocytic nevus of 28 cm × 17 cm covering almost all of her back and the upper part of the right gluteus extending anteriorly to the right breast line. She had also 4 smaller disseminated nevi all over her body. Twelve days after birth she was treated with Versajet® Hydrosurgery and 13 days later she needed a second session for peripheral portions of the nevus. Histopathologic exam revealed no evidence of malignancy. The gadolinium-enhanced MRI identified no signs of neuromelanosis or neurological tumor. Nine months later, she underwent partial surgical excision combined with reconstruction through skin expansion. No signs of malignancy were revealed up to the age of 10 (Figure 1).



**Figure 1:** (A-B): Intraoperative procedures at the age of 12 and 25 days respectively; (C): Appearance 6 days after the last Versajet® Hydrosurgery; (D): Skin expanders for reconstruction's preparation after surgical excision; (E): Patient at the age of 10 years.

### Case 2

The second case was a boy, M.E., with a GCMN on the back, extended to the proximal part of the lower limbs. The skin surface appeared irregular with hyperpigmented areas and erosions. A lot of disseminated smaller nevi were present all over the body. Eight days after birth dermabrasion was performed with Versajet® Hydrosurgery. A second treatment was performed at the age of 25 days. He required blood and plasma transfusions; skin and blood infection occurred soon after the second procedure, but it was otherwise successfully treated with antibiotics. Histopathologic exam revealed no evidence of malignancy. The early MRI identified no signs of neuromelanosis but after 5 years brain deposits of melanin appeared. At 8 years follow up no neurological signs are still evident (Figure 2).

### Case 3

The third patient was a boy, C.M., with a GCMN extended on the back from the shoulders to the sacral region, associated to several satellite nevi. The skin surface appeared irregular with hyperpigmented areas. Thirteen days after birth he was treated in the back region using Versajet® Hydrosurgery. The patient required blood and plasma transfusions postoperatively due to blood loss. Histopathologic exam revealed no evidence of malignancy. Three months after procedure, he was hospitalized for seizures; so, he underwent brain MRI that showed three lesions in the cerebellar and temporal area. Three years later, another neurological melanocytic lesion compares. Actually, he suffers from an autism spectrum disorder with frequent seizures resistant to antiepileptic therapy (Figure 3 and 4).

### Case 4

The fourth patient was a female, Q.S., with a GCMN extended on the back from the neck to the sacrum with anterior development associated to many satellite nevi. Sixteen days after birth the patient

was treated in the back region using Versajet® Hydrosurgery. She required blood and plasma transfusions during surgical procedures and developed a severe skin infection, successfully treated with antibiotics. Histopathologic exam revealed no malignancy but identified a high index of mitotic and proliferative activity. At early MRI, melanin deposits appeared in both cerebellar hemispheres, in the pontine region and near the amygdala. Two years later, the patient underwent surgical correction of hydrocephalus and experienced epileptic insults. At the age of 4 years, repigmentation, hypertrichosis and keloids were present, otherwise with no evidence of malignancy (Figure 5).

### Case 5

The fifth patient was a newborn girl, B.A., with a GCMN covering the right side of the face. Twenty days after birth, she was treated with Versajet® Hydrosurgery, without complications. Histopathologic exam revealed no evidence of malignancy. On early MRI no signs of neuromelanosis were documented. At the age of 3.5 years old the area is characterized by intense hypertrichosis and slight hyperpigmentation but the baby had a good psychomotor evolution (Figure 6).

### Case 6

The last patient was a newborn boy, T.A., with a GCMN of the back and satellite nevi limited to the back and the subscapular region, with no other pathological conditions. Twenty-four days after birth, he was treated with Versajet® Hydrosurgery, without complications. No malignancy was revealed at the histopathologic exam. On early MRI no signs of neuromelanosis were documented. We only have a ten months follow-up that shows back repigmentation without hypertrichosis or hypertrophic scar formation (Figure 7).

A questionnaire survey (Figure 8) was carried out in our institute from August 2021 to January 2022. The examination focused on



**Figure 2:** (A-B): Intraoperative procedures at the age of 8 and 25 days respectively; (C): Appearance 3 months after the last Versajet® Hydrosurgery; (D): Patient at the age of 8 years.





Figure 3: Intraoperative images.

psychological and social impact of these skin lesions on children and their parents, and on the satisfaction degree of the multidisciplinary care received through the years. The survey administration method was a structured interview sent out by mail at patient's families. A cover letter introduced the questionnaire. Responses returned by mail or fax. Survey's collected data were completely anonymously analyzed by statistical analysis with orthogram.

### Results

For each patient, clinical features, treatment modalities and its results are reported and summarized in Table 1. Mean age at referral to our Pediatric Surgery Unit was 12 days old (range of 19-6 days old). 3/6 patients (50%) were referred for evaluation within the first 2 weeks of life. 5/6 (>80%) patients then underwent Versajet® hydrosurgery. Surgical procedure was performed within a mean time of two days from referral and at a mean age of 14 days old (ranges of

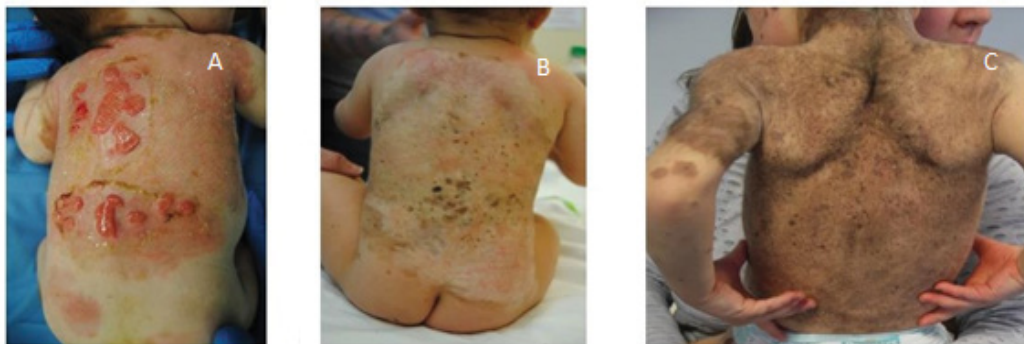


Figure 4: (A): Appearance 5 days after Versajet® Hydrosurgery; (B): Patient at the age of 3 months; (C): Patient at the age of 4 years.



Figure 5: (A): Birth age; (B): Intraoperative procedures at the age of 16 days; (C): Appearance 4 months after Versajet® Hydrosurgery; (D): Patient at the age of 1 year; (E): Patient at the age of 4 years.

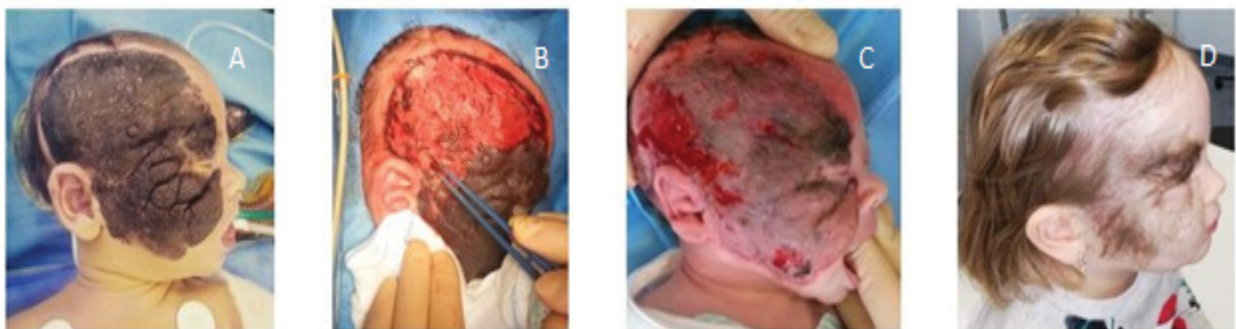


Figure 6: (A-B): Intraoperative procedures at the age of 20; (C): Appearance 10 days after Versajet® Hydrosurgery; (D): Patient at the age of 3 years (d).



Figure 7: (A) Birth age; (B) Intraoperative procedures at the age of 24; (C) Appearance 10 months after Versajet® Hydrosurgery.

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1. On a scale of 0 to 5, where 0 indicates very bad and 5 indicates very well, how do you judge the aesthetic result of the giant congenital melanocytic nevus treatment?  
 0  
 1  
 2  
 3  
 4  
 5

2. On a scale from 0 to 5, where 0 indicates very much and 5 indicates not at all, how much does the child's aesthetic appearance influence his social life?  
 0  
 1  
 2  
 3  
 4  
 5

3. On a scale of 0 to 5, where 0 indicates very poor and 5 indicates very good, how do you rate the quality level of received care?  
 0  
 1  
 2  
 3  
 4  
 5

Figure 8: Questionnaire administered to the patient's family.

21-28 days old). Only one (20%) of the five patients who underwent hydrosurgery had the requirement for planned second look surgery after 17 days to complete the treatment and get better results on the peripheral portions of the nevus. In our neonatal case series Versajet® hydrosurgery system was successfully applied by simply decreasing the flow direction reaching an angle of 4°, opening the disposable gun flow of about 8 mm and healing the water jet [28]. All patients after dermoabrasion were hospitalized in our Neonatal Intensive Care Unit for a period ranging from 2 to 4 weeks. Of the 6 patients of our series, just 1/6 (16.6%) underwent additional unplanned surgery with skin expansion and second-stage excision/coverage.

Complications are listed in Table 1 and are divided into early and late complications. Severe skin infection related to curettage was dominant in the early complications group, it occurred in 2/6 patients

(33.3%) but it was successfully managed with antibiotics. Partial repigmentation and hypertrichosis were seen in all patients (100%), while keloid scars were noted in just 1/6 (16.6%). Of all treated patients, just 1/6 (16.6%) needed unplanned delayed additional surgery, that consisted in the application of skin expanders, to improve cosmetics.

The overall cosmetic result was very satisfying in 2/6 patients (33.3%) and acceptable in the remaining 67% of cases. After the initial surgical procedure, annual/biannual dermatological follow-up was joined by 5/6 (>80%) patients with a compliance to therapy close to 100%. There has been no incidence of cytological atypia or malignant transformation in our case series at follow up. A high rate of neurocutaneous melanosis was documented at MRI (60%), symptoms appeared in half of affected patients. Early MR screening, without contrast, was successfully performed by using a “feed and swaddle” technique in all the patients (100%).

The questionnaire survey was applied in 5/6 patients at a distance of a median of six years (range 3.5-10 years). Five completed questionnaires were returned with a response rate of 100%. In 3/5 patients (60%), self-esteem was significantly improved after surgery at a median of 7.5 years (range 4.5-10 years). Even the quality of the care received was judged to be fully satisfactory in 80% of cases. Despite this good parental feedback, social impact of the disease was however significantly high for affected children, even after surgery (5/5 patients replied that the disease interfered negatively enough or greatly in their social life) (Table 2) (Figure 9).

### Discussion

Treatment of patients with GCMN is complex and still controversial. One of the open questions is whether to recommend Versajet® hydrosurgery as first approach to neonates with GCMN and with which modalities. The efficacy of Versajet® hydrosurgery treatment for GCMN is well documented and our series confirms it; however, there are still very few data concerning treatment modalities, postoperative management and long-term outcomes of newborns undergone hydrosurgery for GCMN. In particular, no prospective or systematic retrospective studies on long-term Versajet® hydrosurgery treatment outcomes have been published to date, that is why our data, despite arising from a small series of patients, are noteworthy.

Early MR screening by using a “feed and swaddle” technique, avoiding general anaesthesia, was successfully adopted in all our neonate patients; furthermore, in this age range, neural melanosis is not obscured by myelination and can be visualized without contrast. The percentage of patients affected by NCM in our small series is higher (>60%) than those reported in English literature. We

**Table 1:** Patient’s clinical features, treatment modalities, complications and evolution.

	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6
<b>Classification</b>	G2, S1, trunk, right gluteal region, lateral C1, R1. NO. H1	62, 52. back chest abdomen (bathing trunk). C2, R2, N1, HO	G2 52 left shoulder and back C2, R2, NO. HO	G2, 51, Back, chest. abdomen "bathing trunk", C2, R2, NO. HO	GO, 50. Half face. CO. R2, N2. H2	G2 51. tunk, C2. R2, NO. HO
<b>Satellite Lesions</b>	Lower limbs	Upper limbs, head	Upper limbs, head	Lower limbs	None	Back, subscapular region
<b>Brain Melanin Deposite (MRI)</b>	No	Appeared at the age of five years Asymptomatic	Yes, symptomatic Epilepsy psyco-motor deficit	Yes, symptomatic hydrocephalus (ventricle-peritoneal shunt). epilepsy	No	No
<b>Age at First Procedure (Days)</b>	12	8	13	16	21	24
<b>N. of Procedures</b>	2	2	1	1	1	1
<b>Transfusions</b>	No	Yes: blood and plasma	Yes: blood and plasma	Yes: blood and plasma	No	Yes: plasma
<b>N. of Medication in Sedation</b>	2	3	3	3	4	
<b>Further Surgical Treatment</b>	Skin expanders	No	No	No	No	No
<b>Short-term Complications</b>	No	Cutaneous infection	No	Cutaneous infection	No	No
<b>Long-term Complications</b>	Repigmentation hypertrichosis, increase of satellite lesions	Repigmentation hypertrichosis, increase of satellite lesions, hypertrophic scarring and keloids	Repigmentation hypertrichosis, hypertrophic scarring atopic dermatitis, increased of brain melanin deposit	Repigmentation hypertrichosis, increase of satellite lesions, hypertrophic scarring and keloids	Repigmentation hypertrichosis,	Repigmentation
<b>Melanoma Development</b>	No	No	No	No	No	No
<b>Family Satisfaction</b>	Very good	Good	Good	Good	Very good	Good

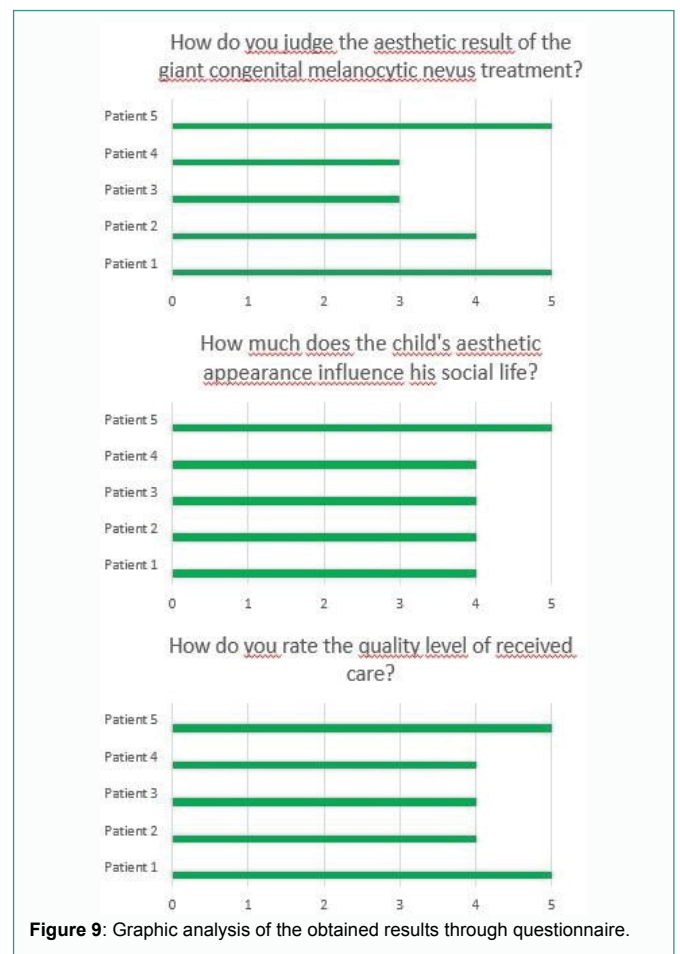
**Table 2:** Answers from patient’s family to the questionnaire.

	Question n.1	Question n.2	Question n.3
Patient 1	Very well	Slightly	Very good
Patient 2	Well	Slightly	Good
Patient 3	Discreet	Slightly	Good
Patient 4	Discreet	Slightly	Good
Patient 5	Very well	Not at all	Very good

confirmed that the presence of NCM strongly influences prognosis and quality of life of patients with GCMN. To date, available therapies just heal symptoms and we can only make an early diagnosis. An early diagnosis was possible in all patients of our study.

The risk of melanoma onset in the GCMN patients is estimated to be between 0.7% and 2.4% and it is maximum in the first 3 years of life [6,8]. Incidence of melanoma remains high up to 5 years and then decreases without ever cancelling itself out [2]. None of our 5 patients showed cytological atypia or malignant transformations at follow-up. The old principle of “nevus removal at any cost” to rule out malignancy with full thickness excision and reconstruction was followed since 2010, when Krengel et al. [7] and Arad and Zuker [30] showed that indication to treatment should be limited to an aesthetic improvement as well as removal of suspicious areas. Prophylactic removal of GCMN does not eliminate the risk of melanoma because radical excision, especially for larger nevi, it is almost impossible to achieve. Neither NCM nor risk of central nervous system melanoma is impacted by surgical approaches.

Some author suggested that destructive techniques, such as dermabrasion, may make melanoma more difficult to detect because of resulting fibrosis and scarring [30]. Data emerging from our series seem to disprove the idea that dermoabrasion makes melanoma difficult to identify. A follow-up program including a close lifelong follow-up with dermoscopy and incisional biopsies of any suspicious



**Figure 9:** Graphic analysis of the obtained results through questionnaire.



areas has shown to be safe and effective to rule out malignancies.

Today the overwhelming majority of GCMN are treated soon after birth to improve aesthetic appearance and to mitigate the stigmatization of those affected. Prophylactic full-thickness surgical excision associated with long-term monitoring is recommended but it is often not feasible. It can be associated with several risks, such as multiple procedures, repeated general anaesthesia, anxiety associated with treatments, postoperative pain, and scar appearance. When conventional full-thickness excision of GCMN is not achievable, a cosmetic benefit can be attained using alternative techniques called non-excisional or destructive treatments [31,32]. Conventional destructive techniques are anyway burdened by the risk of scarring, wound infection and repigmentation. The goal of a conservative non-excisional treatment should be to reduce scar formation removing as much pigmentation as possible without reaching subcutaneous tissue.

In our series, in agreement with the international recommendations for traditional dermoabrasion on GCMN, most patients started treatment as soon as they could at an average age of 14 days, but the 2 children, who had a slight delay, also obtained good results. The number of Versajet® hydrosurgery sessions varied and was dependent on the extent of the disease, whereas its effectiveness, clinically evaluated during and at the end of treatment, appeared not to be dependent on the nevus size but on the presence of micro and macroscopically deeper pigmented skin lesion. 3/6 patients (50%) of our series needs blood and plasma transfusion in the immediate postoperative period. Just 1/6 (16.6%) has built up a hypertrophic scar. This patient who experienced keloid was the only one in which the biopsy has given the histological result of an intradermal nevus. Unlike the other cases, in which the nevus was composed, probably the macroscopically deeper aspect of the lesion may have led, in the intraoperative setting, to a more aggressive attitude with the instrument which could therefore have stimulated the scar reaction in a pathological way.

Concerning early complications we reported a high rate of skin infection (2/6 patients, 33%). Antibacterial and antifungal prophylaxis is administered before surgery, but the infectious risk may also be related to the patient's low level of immune defenses as a newborn. In our series, Versajet® hydrosurgery treatment was well tolerated, and compliance to therapy was close to 100%, even considering the frequent postoperative ambulatory medications. Tolerance was improved by having a multidisciplinary approach with easy access to different physicians at the same time (dermatologists, psychologists, neuropaediatrics).

An early analysis can give a skewed view of a treatment's value. Long-term follow up is an important part of evaluating any medical condition or treatment because clinically important findings can arise several years after cure is done. Long-term follow up was obtained in 5/6 patients. About our long-term outcomes, 4/5 patients (80%) experienced a snow surface that has undergone repigmentation and 3/5 (60%) showed hypertrichosis at an average of 4 years after procedure. The problem of repigmentation has been definitely scaled down since 2019 when Polubotou and kinsler [33] showed that 1) superficial removal techniques do not alter the final colour of CMN and that 2) many CMN lighten over time without intervention and final CMN colour in childhood is significantly associated with the individual's normal skin colour and genetically determined. About hypertrichosis, laser permanent hair removal is now a day's thought to be the definitive solution but it requires serial treatments, difficult to

tolerate by infants; furthermore, histologic and dermoscopic changes have been noted in nevi after laser treatment [34]. For all these reasons, we postponed hair removal until adolescence. In our recent survey study, despite parents reported a "good" level of satisfaction about both aesthetic results and assistance quality in the overall management of the disease, we registered a poor impact of the disease in social life.

## Conclusion

Our results on a small group are in line with what is reported in English literature with 54% of affected adolescents reporting an "extremely large" impact on their skin-related quality of life. The impacts of CMN on a child's social relationships and emotional function are not foreseen at birth but referral to psychologists should be strongly recommended for all affected children to address quality of life and emotional and behavioural health. Shared decision-making can support patients and families in making choices about treatments, focusing the discussion on both patient and family preferences and medical evidence. Potential complications must be weighed against potential improvement in the patient's appearance, improvement in function if associated impairment exists, and psychosocial benefit (or lack thereof).

We are aware of the limits of our study, deriving above all from the small number of the sample examined, but our findings can be considered encouraging and should promote further multicentric studies to reach more reliable conclusions.

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