

Management of Small and Intermediate Congenital Nevi: A Nationwide Survey in Italy

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Key Words

Melanoma · Melanocytic nevus · Small congenital nevus · Intermediate-sized congenital nevus · Survey · Dermoscopy · Skin cancer unit

Abstract

Objective: Small- and intermediate-sized congenital nevi (SCN and ICN) undergo periodic clinical monitoring or surgical excision. We analyzed the management of SCN and ICN in the Italian hospital network. **Methods:** A nationwide survey of a representative sample of centers was conducted. Data were analyzed grouping centers by melanoma incidence into high-volume (>25 diagnoses per year) and low-volume (≤25 diagnoses per year). **Results:** In the pediatric population, 11% of SCN and 22% of ICN are excised, the remainder undergoing clinical monitoring at intervals of 6 months to 2 years (SCN) and of 6 months to 1 year (ICN). In adults, 24% of SCN and 41% of ICN are excised. Clinical monitoring of SCN varies from 6-monthly (most common among low-volume hospitals) to every 2 years; preferred strategies for ICN are follow-up at 1 year (51%) or follow-up at 6 months (42%). For prophylactic surgery, complete excision is preferred. **Conclusions:** The Italian hospital network values

management and treatment of SCN and ICN. In most cases natural evolution prompts prophylactic excision. Clinical examination is an important monitoring tool, though follow-up frequency depends on the clinician's experience and practice.

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Background

In clinical practice, congenital nevi are arbitrarily divided by diameter into small-sized congenital nevi (SCN, ≤1.5 cm), intermediate-sized congenital nevi (ICN, 1.5–20 cm) and large/giant congenital nevi (>20 cm) [1–3]. The incidence of congenital melanocytic nevi is 0.2–6% [4–9]. The prevalence of congenital nevi in Italy ranges from 1 to 1.4% [10]. In a recent Italian survey, the prevalence of ICN was 0.67%, similar to the rate in other published European studies [9].

All congenital nevi can be considered potential melanoma precursors [11]. However, only large congenital nevi are associated with a substantial quantifiable risk of malignant transformation because most studies show an incidence rate of melanoma in SCN and ICN of <1% [1,

12–18]. Based on these data, early and complete surgical excision of giant congenital nevi is advisable while the excision of SCN and ICN remains controversial. For this reason several strategies for the management and treatment of SCN and ICN have been suggested: (1) in the absence of atypical or symptomatic features, periodic clinical monitoring alone; (2) clinical monitoring until puberty and subsequent prophylactic excision; (3) periodic clinical monitoring and surgical excision if suspicious changes develop; (4) surgical excision of a nevus with atypical or dubious features; (5) prophylactic excision of a congenital nevus that does not present atypical characteristics if the patient presents with a high level of anxiety; (6) periodic clinical monitoring of congenital nevi using computer-assisted methods and comparison of images starting from puberty [19]. In most cases surgery includes full-thickness excision, dermabrasion, curettage, laser treatment, or superficial or deep shave [19].

We assessed the frequency of periodic clinical monitoring, prophylactic excision rate and the most frequently used surgical treatment for SCN and ICN in adults and children using a specific questionnaire forwarded to Italian hospitals.

Methods

Briefly, a nationwide survey of clinicians responsible for the diagnosis, therapy or follow-up phases of melanoma care in Italian hospitals was conducted. Italian hospitals with ≥ 200 beds ($n = 285$) were subdivided into 145 hospitals with 200–399 beds and 140 hospitals with ≥ 400 beds and a proportionally stratified random sample ($n = 120$ centers), stratified by number of beds and geographic distribution, was selected. Two or three clinicians were interviewed at each center, resulting in approximately 250 interviews and a predicted margin of error – 95% confidence level – of 7.7%.

Based on the findings, centers were grouped by number of new melanoma diagnoses per year into low- and high-volume centers, around the median value of 25. Variables were analyzed in the total sample/total Italian hospitals, and comparisons were made between high- and low-volume centers using Pearson's χ^2 test and the zeta test at 95% confidence level. Detailed methods are presented elsewhere in this issue [20].

Results

When managing SCN in children, 11% of Italian hospitals perform prophylactic excision, with no difference between high- and low-volume centers, while the remaining 89% propose periodic clinical monitoring (table 1). Monitoring frequency varies from 6 months to 2 years

Table 1. Management of SCN not removed prophylactically in children at Italian hospitals

Examination schedule used	Type of center		
	high-volume (n = 56)	low-volume (n = 62)	all (n = 118)
Centers not excising	50 (89%)	55 (89%)	105 (89%)
Follow-up schedule in centers not excising prophylactically			
As needed	8%	4%	6%
Every 6 months	25%	60%*	43%
Every year	43%	30%	36%
Every 2 years	20%**	4%	12%
In adolescence	2%	0%	1%
Other	2%	2%	2%

Centers are grouped according to yearly melanoma diagnoses into high-volume (>25) and low-volume (≤ 25) centers. Such lesions are not removed prophylactically in 89% of high-volume and 89% of low-volume centers.

* $p = 0.001$; ** $p = 0.01$.

with a small number of hospitals recommending additional examinations only when required (6%) or in adolescents (1%). Monitoring every 6 months is recommended more often in low-volume hospitals than in high-volume hospitals (60 vs. 25%; $p < 0.001$), while monitoring at yearly intervals is recommended by 43% of high-volume hospitals versus 30% of low-volume hospitals. We also found a difference between high- and low-volume hospitals concerning monitoring at 2-year intervals (20 vs. 4%; $p = 0.01$).

In children, prophylactic excisions are performed twice as frequently for ICN as for SCN (11 vs. 22%) (table 2). If periodic monitoring is performed, low-volume hospitals generally monitor every 6 months (61 vs. 25%; $p < 0.001$ vs. high-volume centers) while high-volume centers tend to monitor at 1-year intervals (58 vs. 34%; $p = 0.005$). Overall, monitoring at 2-year intervals is less common for ICN compared to SCN (mean 5 vs. 12%).

When treating children, complete surgical excision is recommended on average in 62% of hospitals (71% of high-volume hospitals versus 55% of low-volume hospitals) (table 3). Other surgical treatments, such as curettage (6%), excisional laser (4%) and laser ablation (1%), are recommended in 11% of hospitals, with no significant differences between categories.

In 11% of Italian hospitals, prophylactic surgery is not performed, and in 14% it is not indicated, particularly in low-volume hospitals (23%) compared to high-volume

Table 2. Management of ICN not removed prophylactically in children at Italian hospitals

Examination schedule used	Type of center		
	high-volume (n = 56)	low-volume (n = 62)	all (n = 118)
Centers not excising	46 (82%)	56 (73%)	105 (78%)
Follow-up schedule in centers not excising prophylactically			
As needed	10%	2%	6%
Every 6 months	25%	61%*	44%
Every year	58%**	34%	45%
Every 2 years	7%	3%	5%

Centers are grouped according to yearly melanoma diagnoses into high-volume (>25) and low-volume (≤25) centers. Such lesions are not removed prophylactically in 46 of 56 high-volume centers (82%) and 45 of 62 low-volume centers (73%).

* p = 0.001; ** p = 0.005.

Table 4. Management of SCN not removed prophylactically in adults at Italian hospitals

Examination schedule used	Type of center		
	high-volume (n = 56)	low-volume (n = 62)	all (n = 118)
Centers not excising	41 (73%)	48 (78%)	90 (76%)
Follow-up schedule in centers not excising prophylactically			
As needed	10%	5%	8%
Every 6 months	33%	54%*	44%
Every year	50%	41%	44%
Every 2 years	7%	0%	4%

Centers are grouped according to yearly melanoma diagnoses into high-volume (>25) and low-volume (≤25) centers. Such lesions are not removed prophylactically in 73% of high-volume and 78% of low-volume centers.

* p = 0.04.

hospitals (5%), while 2% of centers refer patients to other structures for this service. More hospitals perform prophylactic excision of SCN in adults (24%) than in children (11%), with no significant difference between high- and low-volume hospitals (table 4). The remaining 76% of hospitals recommend periodic clinical monitoring.

The interval for clinical monitoring varied from 6 months to 2 years, with a small percentage suggesting additional examinations only when deemed necessary

Table 3. Procedure for prophylactic treatment of congenital melanocytic lesions in children at Italian hospitals

Procedure	Type of center		
	high-volume (n = 56)	low-volume (n = 64)	all (n = 120)
Complete surgical removal when possible, also using successive partial excisions	71%	55%	62%
Periodic clinical follow-up	11%	10%	11%
Surgical curettage	5%	7%	6%
Laser excision	4%	5%	4%
Laser ablation	2%	0%	1%
Self-inspection	2%	0%	1%
Photoprotection	2%	0%	1%

Centers are grouped according to yearly melanoma diagnoses into high-volume (>25) and low-volume (≤25) centers.

Table 5. Management of ICN not removed prophylactically in adults at Italian hospitals

Examination schedule used	Type of center		
	high-volume (n = 56)	low-volume (n = 62)	all (n = 118)
Centers not excising	33 (60%)	37 (58%)	70 (59%)
Follow-up schedule in centers not excising prophylactically			
As needed	6%	6%	6%
Every 6 months	40%	43%	42%
Every year	51%	51%	51%
Every 2 years	3%	0%	1%

Centers are grouped according to yearly melanoma diagnoses into high-volume (>25) and low-volume (≤25) centers. Such lesions are not removed prophylactically in 89% of high-volume and 89% of low-volume centers.

(8%). Clinical monitoring every 6 months is more common in low-volume hospitals (54 vs. 33%; p = 0.04), whereas more high-volume hospitals perform yearly clinical monitoring (50%). Clinical monitoring at 2-year intervals is performed only in high-volume hospitals (7%).

In adults, the number of hospitals performing excisions for ICN (41%) is nearly double that for SCN (24%) (table 5). Prophylactic surgery in adults consists of complete excision in 78% of hospitals, overall (table 6). Other

Table 6. Procedure for prophylactic treatment of congenital melanocytic lesions in adults at Italian hospitals

Procedure	Type of center		
	high-volume (n = 56)	low-volume (n = 64)	all (n = 120)
Complete surgical removal when possible, also using successive partial excisions	83%	73%	78%
Periodic clinical follow-up	7%	10%	9%
Surgical curettage	4%	10%	7%
Laser excision	4%	5%	5%
Laser ablation	2%	0%	1%
Self-inspection	2%	0%	1%
Photoprotection	2%	0%	1%

Centers are grouped according to yearly melanoma diagnoses into high-volume (>25) and low-volume (≤25) centers.

treatment modalities such as surgical curettage (7%), excisional laser (5%) and laser ablation (1%) are performed in 13% of hospitals. Only 1% of centers refer patients to other structures for prophylactic treatment. 9% of Italian hospitals offer only clinical examination and no prophylactic treatment. Periodic clinical monitoring is performed in 60% of hospitals overall, most often with monitoring at 1-year intervals (51%) or short-term follow-up of 6 months (42%). Monitoring at 2-year intervals is used in only 3% of high-volume hospitals, and not at all in low-volume hospitals.

Discussion

Management of giant congenital nevi should always be initiated as early as possible using the most adequate surgical strategy; on the other hand, the excision of SCN and ICN, to reduce the risk of associated melanoma, is controversial [21].

In 1982, Rhodes et al. [22] reported an 8.1% incidence of melanoma in SCN. Similar results (7.9%) were published by Betti et al. [23] in a study of 190 cases of melanoma. In addition, in a large study of 1,954 cases of primitive melanomas, an association with melanocytic nevi was found in more than half of cases (58%); of these, 38% had a histological pattern consistent with congenital nevus [24]. These findings support the hypothesis that SCN are precursors of melanoma. According to Hoss and

Grant-Kels [25], SCN may have a small but definite malignant potential. These data induce many dermatologists to perform 'prophylactic surgical excision', and many clinicians agree that removal is advisable after puberty due to their increased risk of developing melanoma.

However, the absolute risk of melanoma associated with SCN and ICN reported in several studies appears to be low [1, 12–17, 26, 27]. This compares with a general lifetime risk of melanoma in the United States of 1.97% [14]. Therefore, unnecessary surgical treatment could be avoided and a 'wait and see' approach is appropriate. Nevertheless, the true lifetime risk of melanoma may be underestimated toward younger average age of patients in many studies [18]. Therefore, both SCN and ICN can be monitored using integrated clinical and instrumental evaluation; these congenital nevi should be photographed and observed for any changes throughout the patient's lifetime. Dermoscopy appears useful also in the evaluation of superficial congenital nevi [28]. Particularly, symmetric and homogeneous SCN can be monitored through periodic clinical examination and dermoscopy, comparisons over time and self-examination [28]. If suspicious changes occur, the nevus should undergo surgical excision.

Monitoring frequency depends on management strategy, which is based on patient age and type of congenital nevus. Factors influencing treatment decisions include nevus size, location, clinical appearance and possible changes during monitoring over time. We found differences in the diagnosis and/or treatment of SCN and ICN among Italian hospitals and speculate that they are due to variability in how Italian clinicians view the potential risk of malignant transformation. Clinical monitoring of congenital nevi is important for identifying potential malignant transformation, but the appropriate follow-up frequency remains controversial. Periodic clinical monitoring of SCN and ICN in children is managed differently in high-volume hospitals, where yearly monitoring is prevalent, compared to low-volume hospitals where a 6-month interval is more common. Increased monitoring in low-volume hospitals results in an organizational commitment to diagnostic procedures and criteria for efficiency and effectiveness. It is also interesting that low-volume hospitals tend to monitor adults with SCN every 6 months, but to monitor ICN at 1-year intervals (table 4). This means that in centers where periodic monitoring every 6 months is preferred, SCN are monitored more frequently than ICN, even though the latter appear to pose a higher risk of malignant transformation.

Regardless of melanoma case volume, twice as many centers perform prophylactic surgical excision of ICN compared to SCN. This pattern is similar in both children and adults, but the number of centers performing excisions in adults is much higher, reflecting the orientation of many clinicians that surgical excision is advisable after puberty due to an increased risk of melanoma [1, 19]; in Italian patients prophylactic surgical excision was reported in 35% of SCN and in 61% of ICN. In most cases surgery consists of full-thickness excision, dermabrasion, curettage, laser treatment or either superficial or deep shave excisions. Each of these procedures reduces the number of melanocytes, resulting in a reduced theoretical risk of melanoma; however, with the exception of full-thickness excision, these techniques do not eliminate the risk of melanoma originating in the dermis or subcutaneous tissue. Moreover, the impact of surgical prophylaxis on the risk of melanoma, and hence the ratio of cost-benefit in terms of survival, are difficult to evaluate [26], although both exert an important influence on health-care policy. Population-based data from the Swedish medical birth register reveal a surgical excision rate of 40% for (mainly small) congenital nevi and no cases of

melanoma after a median follow-up of 14 years [29]. For this reason, each treatment should be discussed with the patient to analyze the potential risk of malignant transformation and to obtain satisfactory cosmetic and functional results.

Our results indicate that the Italian hospital network pays great attention to the management and treatment of both SCN and ICN. While the effectiveness of SCN and ICN removal at reducing the incidence of melanoma requires further investigation, a randomized trial would not be ethical. Therefore, periodic clinical follow-up remains an important tool for monitoring of congenital nevi and early detection of melanoma, although monitoring frequency is established according to the individual clinician's experience and practice.

Disclosure Statement

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