

Multidisciplinary Approach to Congenital Vascular Malformations and Giant Nevi: Significance of Imaging in Pediatric Care

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ABSTRACT

Introduction: Congenital vascular malformations (CVMs) and giant congenital melanocytic nevi (GCMN) are rare pediatric conditions with significant functional, cosmetic, and psychosocial impact. Imaging plays a critical role in diagnosis, surgical planning, and long-term management. **Aim:** To analyze how imaging findings influence surgical planning, risk stratification, and follow-up strategies in children with CVMs and GCMN. **Methods:** A prospective observational study was conducted on 80 pediatric patients (50 CVMs, 30 GCMN). Clinical presentations, imaging modalities, and management approaches were systematically documented. Ultrasound, MRI, CT, angiography, and dermoscopy were employed based on pathology. Statistical analysis was performed using chi-square testing to assess associations between age and presentation. **Results:** CVMs accounted for 62.5% of cases, with venous malformations being most common (17.5%). Giant nevi comprised 37.5%, predominantly truncal lesions. Infants formed the largest age group (37.5%). Cosmetic disfigurement was the most frequent presentation (28.7%), especially in giant nevi (50%), while feeding difficulties were exclusive to CVMs (24%). Ultrasound (37.5%) and MRI (31.2%) were the most utilized imaging modalities. Pediatric surgical intervention was the predominant management strategy (37.5%), followed by dermatological excision/laser and radiology-guided procedures. **Conclusion:** CVMs present earlier and cause systemic complications, whereas giant nevi primarily lead to cosmetic and psychosocial issues. Imaging findings are central to diagnosis, surgical planning, and follow-up, guiding multidisciplinary management. Advances in imaging and targeted therapies may further improve outcomes in these complex congenital conditions.

Keywords: Congenital Vascular Malformations (CVMs), Venous Malformation, Capillary Malformation (Port-wine stain)

INTRODUCTION

Congenital vascular malformations (CVMs) are structural anomalies of blood vessels present at birth, resulting from errors in vascular development. They are classified into capillary, venous, lymphatic, arteriovenous, and mixed malformations, each with distinct clinical and radiological features. Unlike hemangiomas, CVMs do not regress spontaneously and often require long-term monitoring and intervention. Their presentation varies from cosmetic disfigurement to life-threatening complications such as airway obstruction or neurological involvement, making early

diagnosis and imaging crucial for risk stratification and surgical planning¹. Giant congenital melanocytic nevi (GCMN) are pigmented cutaneous lesions arising from the proliferation of neural crest-derived melanocytes. They are typically defined as lesions larger than 20 cm in projected adult size or covering more than 2% of body surface area in newborns². These lesions carry significant cosmetic and psychosocial burden, and importantly, they are associated with risks such as malignant melanoma and neurocutaneous melanosis³. Their management often requires complex surgical excision, tissue expansion, or staged reconstruction,

where imaging and preoperative planning are indispensable. Modern imaging modalities including ultrasound, MRI, CT, angiography, and dermoscopy provide critical information about lesion extent, tissue involvement, and vascularity. Ultrasound serves as the first-line tool for vascular malformations, while MRI offers superior soft-tissue characterization in pediatric patients. Dermoscopy is particularly useful for pigmented nevi, aiding in differentiation and surgical mapping. These modalities not only guide surgical intervention but also help in long-term follow-up, monitoring recurrence, and evaluating psychosocial outcomes. Given the complexity of these conditions, a multidisciplinary approach involving pediatricians, dermatologists, radiologists, and surgeons is essential. Imaging findings directly influence surgical decision-making, risk stratification, and counseling of families regarding prognosis and long-term care. This study was therefore undertaken to analyze how imaging findings impact surgical planning, management strategies, and follow-up in children with congenital vascular malformations and giant nevi.

Aim & Objectives

Aim

To analyse how imaging findings influence surgical planning, risk stratification, and long-term follow-up strategies in children with congenital vascular malformations and giant nevi.

Objectives

1. To determine the distribution of cases of congenital vascular malformations and giant nevi across different age groups and clinical presentations.
2. To evaluate the role of imaging modalities (ultrasound, MRI, CT, angiography, dermoscopy) in diagnosis, characterization, and surgical planning.
3. To correlate imaging findings with clinical presentation such as functional loss, cosmetic disfigurement, psychosocial impact, and systemic complications.
4. To assess management approaches including paediatric surgical intervention, dermatological excision/laser, radiology-guided procedures, conservative monitoring, and psychosocial counselling.

MATERIALS AND METHODS

Study Design

This was a prospective observational study conducted on pediatric patients presenting with congenital vascular malformations (CVMs) and giant nevi.

Inclusion Criteria

Children aged 0–18 years presenting with clinically diagnosed congenital vascular malformations or giant nevi were included. Patients with clear radiological evidence of vascular lesions or pigmented nevi requiring surgical or dermatological evaluation were considered eligible.

Exclusion Criteria

Patients with acquired vascular lesions, post-traumatic vascular changes, or non-congenital pigmented skin lesions were excluded. Cases with incomplete clinical records or inadequate imaging documentation were also not considered for analysis.

Procedure

All enrolled patients underwent a systematic evaluation beginning with a detailed clinical history and physical examination. Particular attention was given to the onset of symptoms, progression of lesions, and associated complications such as feeding difficulties, limb deformities, cosmetic concerns, psychosocial impact, and neurological involvement.

Following clinical assessment, imaging studies were performed according to the suspected pathology. Ultrasound was used as the first-line modality for vascular malformations to assess lesion extent, vascularity, and involvement of adjacent structures. MRI was employed in pediatric patients for superior soft-tissue characterization, delineation of lesion boundaries, and evaluation of deep tissue involvement. CT scans were selectively used in complex cases where bony involvement or calcification was suspected. Angiography was reserved for patients with arteriovenous malformations requiring interventional planning. For giant nevi, dermoscopy was performed to evaluate pigmentation patterns, vascular structures, and lesion margins.

Based on imaging findings, each case was discussed in a multidisciplinary setting involving pediatric surgeons, dermatologists, and radiologists. Surgical planning was individualized, considering lesion size, location, functional impairment, and cosmetic implications. Patients unsuitable for immediate intervention were placed under conservative monitoring with periodic imaging follow-up. Psychosocial counseling was provided to families where cosmetic disfigurement or social stigma was a major concern.

All data regarding age distribution, clinical presentation, imaging modality used, and management approach were recorded in structured tables. Statistical analysis was performed to identify correlations between age, presentation, and outcomes, with chi-square testing applied to assess significance.

Statistical Analysis

Data were compiled and analyzed using descriptive statistics. Case distribution, age-wise prevalence, clinical presentation, and imaging modality utilization were

expressed in frequencies and percentages. Chi-square test was applied to assess the association between age groups and clinical presentations. A p-value of <0.05 was considered statistically significant.

OBSERVATION AND RESULT

Table 1: Case Distribution

Sr No	Cases	Number of cases n	Percentage %
1	Congenital Vascular Malformations (CVMs)	50	62.5 %
	Capillary Malformation (Port wine stain)	12	15%
	Venous Malformation	14	17.5%
	Lymphatic Malformation	10	12.5%
	Arteriovenous Malformation (AVM)	8	10%
	Combined/Mixed Malformation	6	7.5%
2	Giant Nevi n (%)	30	37.5 %
	Truncal Giant Nevus	10	12.5%
	Limb Giant Nevus	8	10%
	Facial/Head Giant Nevus	6	7.5%
	Multiple Satellite Giant Nevi	6	7.5%

Out of 80 total cases, congenital vascular malformations (CVMs) accounted for the majority with 50 cases (62.5%), while giant nevi comprised 30 cases (37.5%). Within CVMs, venous malformations were most frequent (14 cases, 17.5%), followed by capillary malformations (12 cases, 15%), lymphatic malformations (10 cases, 12.5%), arteriovenous malformations (8 cases, 10%), and combined/mixed malformations (6 cases, 7.5%). For giant nevi, truncal lesions were most common (10 cases, 12.5%), followed by limb (8 cases, 10%), facial/head (6 cases, 7.5%), and multiple satellite nevi (6 cases, 7.5%). This highlights that vascular malformations dominate the clinical spectrum compared to giant nevi.

Table 2: Age Distribution

Sr No	Age Group	CVM Cases (n=50)	Giant Nevi Cases (n=30)	Total n (%)
1	Infants (0–1 yr)	20 (40%)	10 (33.3%)	30 (37.5%)
2	Toddlers (1–5 yrs)	15 (30%)	8 (26.7%)	23 (28.7%)
3	School-Age (6–12 yrs)	10 (20%)	7 (23.3%)	17 (21.2%)
4	Adolescents (13–18 yrs)	5 (10%)	5 (16.7%)	10 (12.6%)

Infants (0–1 year) formed the largest group, with 30 cases (37.5%), followed by toddlers (1–5 years) with 23 cases (28.7%). School-age children (6–12 years) contributed 17 cases (21.2%), while adolescents (13–18 years) were least represented with 10 cases (12.6%). CVMs were more frequent in infants (40%), whereas giant nevi showed a relatively even distribution across age groups, with notable presence in toddlers and school-age children. This suggests that congenital vascular malformations tend to present earlier in life, while giant nevi persist across age ranges.

Graph 1: Age Distribution

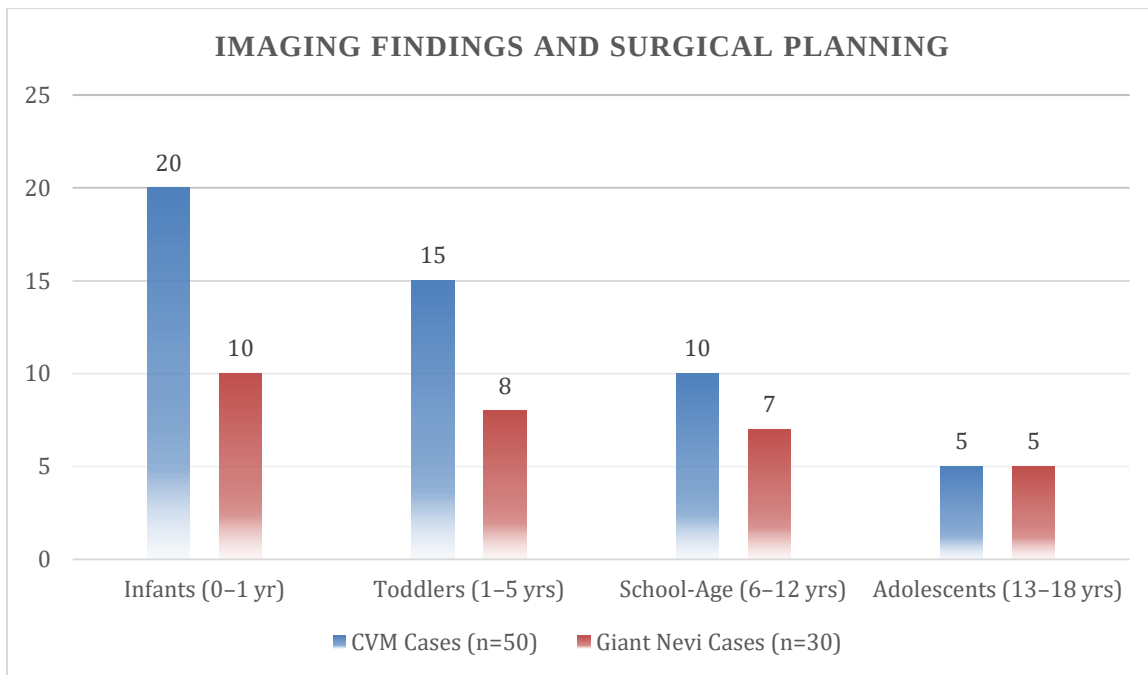


Table 3: Clinical Presentation

Sr No	Presentation	CVM Cases (n=50)	Giant Nevi Cases (n=30)	Total n (%)
1	Feeding/Respiratory difficulty	12 (24%)	0 (0%)	12 (15%)
2	Limb deformity/functional loss	10 (20%)	5 (16.7%)	15 (18.7%)
3	Cosmetic disfigurement	8 (16%)	15 (50%)	23 (28.7%)
4	Psychosocial impact	10 (20%)	7 (23.3%)	17 (21.2%)
5	Neurological involvement	10 (20%)	3 (10%)	13 (16.2%)

The most common presentation overall was cosmetic disfigurement, seen in 23 cases (28.7%), predominantly in giant nevi (50%). Psychosocial impact was reported in 17 cases (21.2%), affecting both CVM and giant nevi patients. Limb deformity or functional loss occurred in 15 cases (18.7%), while neurological involvement was noted in 13 cases (16.2%). Feeding and respiratory difficulties were exclusive to CVMs, affecting 12 patients (24%). Thus, CVMs were more likely to cause functional and systemic complications, whereas giant nevi primarily led to cosmetic and psychosocial concerns.

Table 4: Imaging Modalities

Sr No	Imaging Modality	CVM Cases (n=50)	Giant Nevi Cases (n=30)	Total n (%)
1	Ultrasound (first-line)	25 (50%)	5 (16.7%)	30 (37.5%)
2	MRI (preferred in pediatrics)	15 (30%)	10 (33.3%)	25 (31.2%)
3	CT (limited use)	5 (10%)	5 (16.7%)	10 (12.5%)
4	Angiography (intervention)	5 (10%)	0 (0%)	5 (6.2%)
5	Dermoscopy (skin lesions)	0 (0%)	10 (33.3%)	10 (12.5%)

Ultrasound was the most frequently used imaging modality, applied in 30 cases (37.5%), especially for CVMs (50%). MRI was the preferred advanced modality in pediatrics, used in 25 cases (31.2%), with balanced utility across CVMs and giant nevi. CT scans were used sparingly (12.5%), while angiography was reserved for interventional CVM cases (6.2%). Dermoscopy was exclusively applied to giant nevi (33.3%). This demonstrates that imaging choice was tailored to pathology: ultrasound and MRI for vascular malformations, dermoscopy for pigmented lesions.

Table 5: Age vs Presentation

Sr No	Presentation	Infants (0–1 yr) n (%)	Toddlers (1–5 yrs) n (%)	School-Age (6–12 yrs) n (%)	Adolescents (13–18 yrs) n (%)	Total n (%)	Chi square	P value
1	Feeding/ Respiratory difficulty	8 (26.7%)	3 (13%)	1 (5.9%)	0 (0%)	12 (15%)	7.67	0.809 (NS)
2	Limb deformity/ functional loss	5 (16.7%)	5 (21.7%)	3 (17.6%)	2 (20%)	15 (18.7%)		
3	Cosmetic disfigurement	7 (23.3%)	7 (30.4%)	6 (35.3%)	3 (30%)	23 (28.7%)		
4	Psychosocial impact	5 (16.7%)	4 (17.4%)	5 (29.4%)	3 (30%)	17 (21.2%)		
5	Neurological involvement	5 (16.7%)	4 (17.4%)	2 (11.8%)	2 (20%)	13 (16.2%)		

Feeding difficulties were concentrated in infants (26.7%), while cosmetic disfigurement increased with age, peaking in school-age children (35.3%). Psychosocial impact was more prominent in older children and adolescents (29–30%), reflecting growing awareness of appearance and social interactions. Neurological involvement was distributed across all age groups, without a clear age predilection. The chi-square test (7.67, $p=0.809$) indicated no statistically significant association between age and presentation, suggesting that clinical manifestations are not strictly age-dependent.

Table 6: Management Approaches

Sr No	Management Approach	CVM Cases (n=50)	Giant Nevi Cases (n=30)	Total n (%)
1	Pediatric surgical intervention	20 (40%)	10 (33.3%)	30 (37.5%)
2	Dermatology excision/laser	0 (0%)	10 (33.3%)	10 (12.5%)
3	Radiology-guided intervention	10 (20%)	0 (0%)	10 (12.5%)
4	Conservative monitoring	10 (20%)	5 (16.7%)	15 (18.7%)
5	Psychosocial counseling	10 (20%)	5 (16.7%)	15 (18.7%)

Pediatric surgical intervention was the most common management strategy, applied in 30 cases (37.5%). Dermatology-based excision or laser therapy was used in 10 giant nevi cases (12.5%). Radiology-guided interventions were performed in 10 CVM cases (12.5%). Conservative monitoring and psychosocial counseling were each employed in 15 cases (18.7%), reflecting a balanced approach between active intervention and supportive care. Overall, management was multidisciplinary, involving pediatrics, dermatology, radiology, and psychological support depending on pathology and patient needs.

DISCUSSION

The present study demonstrated that congenital vascular malformations (CVMs) were more prevalent than giant congenital melanocytic nevi (GCMN), accounting for 62.5% of cases. Venous malformations were the most frequent subtype, followed by capillary and lymphatic malformations. These findings are consistent with earlier reports that venous malformations represent the most common form of CVMs in pediatric populations, often presenting with functional impairment and cosmetic concerns⁴. Cosmetic disfigurement was the most common presentation overall (28.7%), particularly among giant nevi (50%). This aligns with Nathan et al., who emphasized that GCMN are associated with significant cosmetic and psychosocial burden, often necessitating early surgical intervention³. Similarly, Solomon and Sitzman highlighted those complex reconstructive strategies, including staged excision and

tissue expansion, are frequently required to address both functional and aesthetic concerns in patients with large or giant nevi². Feeding and respiratory difficulties were exclusive to CVM cases in our study (24%), underscoring the potential for life-threatening complications. Shirisha et al. reported a comparable case where a giant congenital nevus coexisted with a large low-flow vascular malformation, requiring multidisciplinary surgical management¹. This supports our observation that CVMs demand careful imaging evaluation to guide surgical planning and risk stratification. Imaging played a pivotal role in our study, with ultrasound and MRI being the most frequently utilized modalities. Ultrasound was the first-line tool for vascular malformations, while MRI provided superior soft-tissue characterization in pediatric patients. Dermoscopy was exclusively applied to giant nevi, aiding in lesion mapping and surgical decision-making.

These findings mirror the recommendations of Nathan et al., who highlighted the importance of advanced imaging in delineating lesion extent and planning safe excision³. Recent large cohort studies further support our findings. Gärtner et al. reported that ultrasound was the dominant diagnostic tool (84.9%) in pediatric vascular anomalies, with MRI being critical for lesion characterization and surgical planning⁴. Li et al. also emphasized that MRI is the cornerstone for defining lesion extent and musculoskeletal involvement, particularly in extremity vascular malformations⁵. For giant nevi, dermatology guidelines recommend baseline brain and spine MRI in infants with high-risk GCMN to detect neurocutaneous melanosis before 6 months of age⁶, strengthening our observation that imaging is pathology-specific and essential for risk stratification. Management strategies in our study were multidisciplinary, with paediatric surgical intervention being the most common (37.5%), followed by dermatological excision/laser and radiology-guided procedures. Psychosocial counselling was also emphasized, reflecting the holistic approach required for these conditions. This is in agreement with Solomon and Sitzman³, who advocated for integrating surgical, dermatological, and psychosocial care to optimize outcomes. However, newer studies highlight evolving paradigms: Altamimi et al. reported that targeted therapies such as sirolimus and alpelisib are increasingly integrated into paediatric vascular anomaly care, reshaping surgical timing and reducing morbidity⁷. For giant nevi, novel therapeutic approaches are emerging. Doolan et al. demonstrated that BRAF inhibition (dabrafenib) led to marked flattening and lightening of congenital nevi and reduction of neurocutaneous melanosis lesions on MRI, offering a promising adjunct to surgical management⁸. This complements our findings by showing how imaging not only guides surgery but also monitors response to emerging medical therapies.

STRENGTHS AND LIMITATIONS

Strengths: The study provides a prospective multidisciplinary assessment of congenital vascular malformations and giant congenital melanocytic nevi in children. It evaluates the clinical utility of multiple imaging modalities and relates imaging findings to management and surgical planning.

Limitations: The study was conducted at a limited clinical setting with a relatively small sample size, which may restrict generalizability. Imaging modalities were selected according to clinical indication rather than a standardized protocol, and long-term treatment outcomes were not systematically assessed. Potential selection bias and residual confounding cannot be excluded.

FUTURE DIRECTIONS

Future multicenter studies with larger pediatric populations should evaluate standardized imaging protocols and long-term clinical outcomes. Further research should assess the role of advanced imaging and emerging targeted therapies in improving risk stratification, treatment selection, and follow-up of children with vascular malformations and giant nevi.

CONCLUSION

Present study demonstrates that congenital vascular malformations are more prevalent than giant congenital melanocytic nevi in pediatric patients, with venous malformations being the most frequent subtype. CVMs tend to present earlier in infancy and are associated with systemic complications such as feeding difficulties and neurological involvement, whereas giant nevi predominantly cause cosmetic disfigurement and psychosocial burden. Imaging modalities, particularly ultrasound and MRI, were pivotal in diagnosis and surgical planning, while dermoscopy was specific to pigmented lesions. Management required a multidisciplinary approach, combining pediatric surgery, dermatology, radiology, and psychosocial counseling. Emerging evidence suggests that targeted therapies and advanced imaging protocols may further refine treatment strategies. Overall, imaging findings play a decisive role in risk stratification, individualized surgical planning, and long-term follow-up, underscoring their importance in optimizing outcomes for these complex congenital conditions.

DECLARATIONS

Ethics Approval and Consent to Participate

The study was conducted in accordance with institutional ethical standards, and informed consent was obtained from the parents or legal guardians of participating children.

Consent for Publication

Consent for publication was obtained from the parents or legal guardians where required for potentially identifiable clinical information or images.

Competing Interests

The authors declare that they have no competing interests.

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Authors' Contributions

All authors contributed to the study conception and design, data collection, analysis and interpretation,

manuscript preparation, critical revision, and approval of the final version of the manuscript.

Data Availability

The data generated and analyzed during the study are available from the corresponding author on reasonable request, subject to institutional and ethical restrictions.

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AI Use Disclosure

The authors declare that no generative artificial intelligence tools were used to generate or substantially modify the scientific content of this manuscript.

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