Management of Cutaneous Juvenile Hemangiomas

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Abstract

Past descriptions of vascular lesions often confused vascular malformations and hemangiomas using interchangeable definitions which led to inappropriate treatment and inconsistency in the medical literature. The work of Mulliken and Glowacki systematically delineated the difference between hemangiomas and the multiple varieties of vascular malformations. Hemangiomas are in fact as the suffix “oma” suggests true tumors exhibiting cellular proliferation on histology. Additionally, treatment has been clouded by numerous reports in the literature of near universal complete spontaneous resolution and conversely arguments that every lesion demands treatment. More recently, thoughtful studies of the natural course of hemangiomas have been completed, algorithms for intervention versus observation proposed, and effective safe treatment plans devised. The following article will be presented in two sections. The first section will review the diagnosis and natural history of hemangiomas as appreciated in modern literature and compare this to a review of historical articles. This information will then be used to describe a rational and thoughtful algorithm for observation or intervention and recommend appropriate treatment options. Special emphasis will be given to surgical technique and several cases of late involuting hemangiomas of the face will be presented.

KEYWORDS: Hemangioma, classification, management algorithm, vascular birthmark, cutaneous

Past descriptions of vascular lesions often confused vascular malformations and hemangiomas using interchangeable definitions, which led to inappropriate treatment and inconsistency in the medical literature. In many classic articles, the term capillary hemangioma was used to describe what is now known as a port-wine malformation and the terms strawberry nevus and cavernous hemangioma were used to describe what is recognized today as a true hemangioma. The work of Mulliken and Glowacki systematically delineated the difference between hemangiomas and the multiple varieties of vascular malformations. 1 Waner and Suen suggested further that describing true hemangiomas as capillary or cavernous confuses these terms with earlier descriptions and creates the illusion that these lesions are separate entities when in fact they are not and are only differentiated by their location within the soft tissue. Instead, they suggest using the simple classification

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Table 1 Hemangioma Terminology

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<th>Historical Terms</th>
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<tr>
<td>Strawberry nevus, capillary</td>
<td>Superficial hemangioma</td>
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<td>Cavernous hemangioma</td>
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<td>Capillary cavernous hemangioma</td>
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system of superficial hemangioma to describe those growing within the papillary dermis, deep hemangioma to describe those growing within the reticular dermis or subcutaneous tissue, and compound hemangioma if the lesion possesses both levels of involvement. This simplified sensible classification, which is easily assignable with clinical inspection, will be used throughout the remainder of this manuscript (Table 1).

In fact, hemangiomas are, as the suffix -oma suggests, true tumors exhibiting cellular proliferation on histology. They are cellular proliferations of endothelial cells and associated pericytes. Just as the diagnosis and classification of hemangiomas have been confusing, so too have the true natural history and propensity for persistent cosmetic deformity. Additionally, treatment has been clouded by numerous reports in the literature of near universal complete spontaneous resolution and, conversely, arguments that every lesion demands treatment. More recently, thoughtful studies of the natural course of hemangiomas have been completed, algorithms for intervention versus observation proposed, and effective safe treatment plans devised. The following discussion will be presented in two sections. The first section will review the diagnosis and natural history of hemangiomas as appreciated in modern literature and compared with a review of historical articles. This information will then be used to describe a rational and thoughtful algorithm for observation or intervention and recommend appropriate treatment options. Special emphasis will be given to surgical technique, and several cases of late involuting hemangiomas of the face will be presented.

Epidemiology and Natural History of Hemangiomas

Hemangiomas are the most common neoplasm of infancy and childhood, with an estimated prevalence of 10% by 1 year of age. Thirty percent of hemangiomas will be evident at birth, and the majority of others will present within the first few weeks of life. Approximately 40 to 60% of hemangiomas involve the head and neck, and 80% are solitary lesions. Within the head and neck, a pattern of presentation along embryonic fusion planes has been suggested, and a female sex predilection of 3 to 4:1 exists. The vast majority occurs spontaneously, but quoted risk factors include premature birth weight of 1000 to 1500 g and chorionic villus sampling. In the rare patient presenting with four or more cutaneous hemangiomas, consideration should be given to the possibility of visceral involvement, and a screening ultrasound should be performed. If further delineation of visceral involvement or a question in diagnosis persists, magnetic resonance imaging will assist in further assessment. The presentation of stridor in an infant with a cutaneous hemangioma should prompt an evaluation of the airway. Additionally, extensive facial hemangiomas have been associated with posterior fossa malformations, arterial anomalies, coarctation of the aorta and cardiac defects, and eye abnormalities; the acronym PHACE has been proposed.

Hemangiomas usually present within the first few weeks of life. The diagnosis can nearly always be made by a history and physical exam with description of a small red papule or blue subcutaneous lesion that has been growing with variable intensity since presentation. Superficial hemangiomas appear as a bright red macular or papular lesion with well-defined borders (Fig. 1A); the macular variety may initially be confused with a port-wine malformation, but time will easily differentiate the two because of the hemangioma's inevitable change in size and the port-wine malformation's relatively stable size. Deep hemangiomas, because of the intervening dermal layer, appear as a subcutaneous mass with bluish or colorless overlying skin depending on depth (Fig. 1B). At times, deep hemangiomas may appear similar to lymphatic malformations upon inspection; a careful history will nearly always reveal the correct diagnosis, however. If the diagnosis is in question, an ultrasound or magnetic resonance imaging scan will usually serve to differentiate a hemangioma from another vascular entity. Many hemangiomas that present with a combination of the above features should be termed compound (Fig. 1C).

A hemangioma's life cycle is characterized by two distinct clinical stages: proliferation and involution. A third stage, postinvolution, has been described; however, this is not actually a stage but instead a lack of completion of the involutorial stage. Proliferation occurs during the first 12 months and occasionally as late as 18 months. The growth pattern varies greatly in both timing and severity from one lesion to the next; however, a bimodal growth pattern is frequently observed: the first occurs during the first few months of life and a second between 4 and 6 months of age. It is during the first growth phase that functional and cosmetic concerns such as ulceration, nasal and visual obstruction, or frank airway obstruction may occur, thus stimulating referral to a specialist (Fig. 2). Histologically, proliferating hemangiomas are characterized by plump proliferating endothelial cells with barely perceptible vascular channels.
Proliferation is invariably followed by involution, which by definition follows the completion of proliferation. The onset of involution is characterized clinically by a definite decrease and then cessation in the growth of the lesion. The cutaneous component changes from a bright red color to a darker maroon, and eventually patches of an ashen gray color ensue and spread (Fig. 3). Upon palpation, the tumor will evolve from the original firm, tense consistency to a lobular, soft, compressible mass. Histologically, the plump endothelial cells give way to a gradual flattening until late involution, when flat, inactive endothelial cells predominate. At the same time, the vascular channels become more obvious until large ectatic capillary-like vessels are seen. Variable mast cell infiltration is evident in proliferation and involution. Eventual progressive deposition
of perivascular fibrofatty tissue, together with a decrease in the number of vascular channels and production of ectatic vessels, manifests clinically as superficial telangiectasias and at times a subcutaneous fibrofatty residuum.\(^1,2\) As with proliferation, involution occurs at drastically differing rates. However, some degree of prediction regarding the rate of involution and completeness of its course is possible. The literature and clinical experience suggest that lesions that are located in the central face or that are not significantly involuting by age 2 or 3 are more likely to leave cosmetic deformities.\(^8\)

**NATURAL HISTORY OF HEMANGIOMAS IN THE HISTORICAL LITERATURE**

The rate and completeness of involution have been debated for several decades. Many articles published in the mid-20th century argued that nearly all hemangiomas eventually involuted to the point that no residual deformity existed.\(^3-8,12\) During that same time period, other authors advocated aggressive numerous interventions for the treatment of hemangiomas.\(^13-20\) Later in the 20th century, the earlier evaluations as to the natural history of hemangiomas were reevaluated, and this new information has often been quoted as rationale for intervention in the management of hemangiomas.\(^21,22\) An analysis of the data from these earlier articles and methods of treatment is warranted to form the basis of a reasonable discussion of whether intervention in the care of hemangiomas is indicated or not. Articles published prior to Mulliken and Glowacki’s work often discussed what we now differentiate into hemangiomas and vascular malformations as similar, related entities. Even though different definitions were used, several authors gathered detailed information regarding the natural history of hemangiomas. Several of these will be discussed in further detail.

Lister in 1938 described beautifully the clinical presentation of “strawberry naevi” as superficial, deep, or involving both regions, as now suggested by Waner and Suen.\(^2\) He followed 92 “naevi” for 7 years, reporting that “No exception has been found to the rule that naevi which grow rapidly during the early months of life subsequently retrogress and disappear of their own accord, on the average about the fifth year of life. It follows that drastic measures for the destruction of these naevi are inadvisable, and treatment should be expectant and conservative”.\(^9\)

Bowers and colleagues in 1960 performed perhaps the most objective review and prospective long-term observation of the natural history of hemangiomas and attempted to identify factors that affected the eventual outcome. They divided 169 nevi into two groups: “cured,” defined as “gone without a trace, or so inconspicuous that a thin dusting with powder would make it invisible,” and “imperfect,” defined as “anything short of cured.” Eighty-three of 165 nevi (49%) were cured at the end of 3 years. At 7 years, 72% were reported as cured. He further observed that a nevus remaining unimproved after the end of 3 years is unlikely to recover completely by about 7 years, but that early improvement does not always lead to early cure. Interestingly, size and ulceration did not seem to affect outcome; however, a definite trend existed for poor outcome in lesions of the mucosal lip.\(^8\)

**Figure 2** Same infant in Fig. 1C 5 months later with rapidly proliferating hemangioma not responsive to intralesional steroids or pulsed-dye laser therapy, now with astigmatism and partial visual obstruction threatening amblyopia.

**Figure 3** Late involuting hemangioma demonstrating deepening of color and spreading of ashen gray color of skin. Significant subcutaneous fibrofatty residuum exists requiring surgical excision.
During the time period of the previous authors' publications, a definite difference of opinion existed regarding the management of hemangiomas. Those just presented represented one ideology, and others advocated various forms of treatment. Lewis advocated injecting hemangiomas with a sclerosing solution consisting of sodium morrhuate, procaine, and hyaluronidase. No comparison between observed and treated patients was done; however, several photographic examples are presented with apparent excellent results most notably in a nasal tip lesion and large central mucosal lip lesion.\(^{11}\) Kieln advocated following hemangiomas for the first year except in cases where "growth is infiltrating into the adjacent areas and perhaps threatening invasion of the orbit or nasal cavity, with possible danger to vision, breathing or swallowing." Surgery was planned after involution ensued; however, no guidelines were offered to determine which lesions warranted surgery.\(^{12}\) Matthews relied primarily on diathermy for strawberry marks (superficial hemangiomas) and sclerosing with supersaturated sodium chloride for cavernous (deep) hemangiomas with caution to inject under the skin and never into the skin, as it will inevitably necrose.\(^{14}\) Other methods of treatment used for hemangioma in the past include cautery, carbon dioxide snow, surface radium, radioactive particles (thorium-X), X-rays, interstitial gamma radiation, injection of boiling water or ethamolin froth, or continuous compression.\(^{16-20}\)

The available treatment options of the day all had serious side effects, and several authors commented on the disastrous results observed with treatment.\(^{9,10}\) Additionally, the available anesthesia carried its own significant risk. The diagnosis and treatment were also complicated by the confusing variable definitions given to various vascular lesions and a general lack of understanding of the histological basis for the lesions. Given the lack of consistency in diagnosis and description in the literature, lack of dependable reproducible treatment and safe anesthesia, coupled with the evidence that the far majority of hemangiomas eventually completely involuted, a strong opinion developed that the appropriate treatment for hemangiomas was merely observation. This came to be known as benign neglect. With the publication of Mulliken and Glowacki, a new understanding developed regarding the origins and differentiation of hemangiomas and vascular malformations.\(^{1}\) This allowed for consistent definition in the literature and the ability to more accurately follow the course of these differing entities.

### COMPARISON OF HISTORICAL WORKS AND RECENT PUBLICATIONS

Finn, working with Glowacki and Mulliken, produced an article evaluating the clinical application of their new classification system. As part of that article, they followed 159 hemangiomas to "complete involution." They described an excellent cosmetic result as "no redundant skin, scar, or telangiectasia." Of the 159 patients followed, 96 produced an excellent result (60%), and 63 produced a less than excellent result (40%). Seventy-nine of 159 lesions involuted by 5 years of age (50%), and 80 involuted after 5 years of age (50%). These will be called "early" and "late" involutors. Of the 79 early involutors, 64 produced an excellent result (81%), and 15 produced an "imperfect result" (19%). Of the 80 late involutors, 32 produced an excellent result (40%), and 48 produced an imperfect result (60%). In summary, 60% of all lesions produced an excellent result and 40% did not. Half involuted early and half, late. Eighty-one percent of those that involuted early produced excellent results and 19% did not; 40% of those that involuted late produced excellent results and 60% did not. In the authors' summary to this article, they state that "80% of lesions involuting after age 6 do so 'imperfectly', with residual scar, redundant skin or telangiectasia, as opposed to 38% 'imperfect' result for lesions involuting before age 6."\(^{21}\) This statement has been often quoted\(^{22}\); however, based upon the data presented in the paper and analyzed herein, its origin is not apparent.

The modern argument for treating hemangiomas rather than benign neglect has been based on the observation that many of the hemangiomas do not regress completely, that is, without cosmetic sequelae. Authors have quoted Finn in support of this and also reasoned that the previous works\(^{13,14,18-12}\) either overestimated the number of hemangiomas that would eventually involute completely or accepted a less than perfect cosmetic outcome. Table 2 summarizes the authors' stated results that have been presented. The proportion of early versus late involuting lesions appears to be

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<td>No. of Hemangiomas Followed</td>
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<tr>
<td>Lister (1938)(^9)</td>
<td>92</td>
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<tr>
<td>Wallace (1952)(^{13})</td>
<td>411</td>
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<td>Simpson (1959)(^{14})</td>
<td>140</td>
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<tr>
<td>Bowers et al (1960)(^9)</td>
<td>169</td>
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<tr>
<td>Finn et al (1983)(^{19})</td>
<td>159</td>
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around 50%. The number of lesions actually progressing to a perfect outcome is of greater debate but appears to be between 60 and 75% on critical review. Clearly 100% of lesions, as some authors suggested, do not resolve to a point where no evidence of the lesion exists. In addition to these findings, studies of the psychological impact of hemangiomas on children and families have emerged and provided additional guidance that should be considered when determining care.

PSYCHOLOGY AND RATIONAL FOR TREATMENT

Children begin to develop self-awareness at 18 to 24 months of age. A significant body image is well under development by the age of 3 years. A comparison of children aged 3 to 5 years affected by head and neck hemangiomas with unaffected children found a difference between the groups. The study found that children with hemangiomas perceived that others valued them significantly lower compared with the unaffected group. Additionally, interviews of the mothers reported strangers raising the question of child abuse, children burying their faces or hiding their lesions with their hair, and family and friends commenting on doing something about it. A further study confirmed the negative psychological impact of hemangiomas on the child and family and relief of these stresses with proper counseling and intervention if warranted.

During the period that benign neglect dominated most practitioners’ philosophies, the methods of treatment available may not have improved the result even in those lesions that did not regress on their own accord. This fact certainly influenced the opinion of earlier authors to support the philosophy of benign neglect. Recent advances in anesthesia, laser technologies, medical treatment, and surgical methods, however, not only allow effective intervention to treat those lesions that would otherwise leave a grossly unacceptable result in years to come but also permit safe treatment to prevent children from suffering the psychological impact of living with a gross facial or body deformity. Careful, thoughtful evaluation of past studies in the authors’ opinion now clearly shows that somewhere between 25% and 40% of hemangiomas will leave an unacceptable cosmetic result that could be improved with medical or surgical intervention.

TREATMENT OPTIONS FOR HEMANGIOMAS

The decision to treat a hemangioma must be carefully thought out and discussed with the parents of the child. The goal of the evaluation should be to attempt to determine which hemangiomas are likely to involute with a perfect result and which ones will likely leave a cosmetic deformity. Two phases of treatment are possible: during proliferation, which will hopefully affect the ultimate size of the hemangioma and induce involution, and during involution. Williams and colleagues presented a retrospective review of 168 patients with hemangiomas and developed an algorithm for management. This algorithm will be the primary basis for the following discussion (Fig. 4A, B).

Attempting to determine which proliferating hemangiomas will result in cosmetically unacceptable outcomes may be impossible. Fortunately, the treatment modalities primarily used during proliferation have few side effects or complications, and the goals of therapy are therefore to attempt to reduce the eventual size of the lesion and induce involution. Only those hemangiomas that are life-threatening, of significant functional concern, or overwhelmingly large warrant more aggressive therapy during proliferation. The treatment options commonly used for proliferating hemangiomas include observation, intralesional steroids, systemic steroids, pulsed-dye laser, and surgical debulking. Other therapies, including interferon α2a, interferon α2b, argon laser, and neodymium doped: yttrium aluminum garnet (Nd:YAG) laser, are available options. Enolías reported on 25 cases of “alarming” hemangiomas treated with systemic steroids. The three patients with liver involvement all died during or following therapy. Interferon α2a and interferon α2b have been recommended for the treatment of these types of complicated hemangiomas, including those with airway obstruction or visceral involvement, with good response in many patients. However, spastic diplegia has been reported in several patients with interferon α2a, and interferon’s use should be reserved to those severe life-threatening situations where other modalities are not an option or have failed. Argon, potassium-titanyl-phosphate (KTP), and Nd:YAG laser are recommended for use by some authors for the treatment of hemangiomas, but have been mostly surpassed by the introduction of flashed lamp pulsed-dye laser due to its effectiveness in the treatment of superficial hemangiomas and unparalleled safety. A role may still exist for the use of intralesional laser treatments, however, the safety of this modality in regards to scar production and damage to deeper anatomic structures has been of concern.

MANAGEMENT OF PROLIFERATING HEMANGIOMAS

The decision to treat a proliferating hemangioma is based upon the rate of proliferation, cosmetic significance of the location, presence of ulceration or pending ulceration, or impending or present functional problem. Lesions involving the eyelids or the visual axis deserve special attention and will be discussed in more detail.
later. Because at least 60% of hemangiomas will involute with no cosmetic deformity, perhaps the most important aspect of treating the proliferating hemangioma is determining when to observe. Observation should be considered an active treatment similar to laser and surgery, and the term benign neglect should be abolished. Lesions that do not fulfill the qualification to warrant further intervention should be followed closely at least every 3 months, and if the lesion is in a cosmetically sensitive area, every 2 to 4 weeks if necessary to attempt to determine its growth characteristics. Open and thoughtful dialogue must occur with the parents, and they should be encouraged to follow up and have the child treated more frequently as necessary for their own comfort as well as for the best interest of the child. Never should a physician be coerced into intervening to alter the natural course of a hemangioma because of the parent's demands. It is important to treat the family with educational materials outlining the natural course and reassure them that the treatment goals are in the best interest of the child. Just because modern science has given the tools to intervene safely does not make it a requirement. In Williams and coworkers' treatment of 168 patients in a high-risk tertiary referral environment, only 15 (7.8%) received steroids, either intraleSIONAL or systemic; 61 (36%) received pulsed-dye laser treatment; and 41 patients (24%) underwent surgery, usually during involution. Interestingly, Bowers et al in 1960 found a 25% imperfect resolution rate following involution, a nearly exact correlation to the percentage of patients requiring surgical intervention by Williams et al in 2000.

After interviewing the parents, hemangiomas in the proliferative phase that are undergoing rapid growth, have ulcerated, or on inspection are pending ulceration are considered for treatment with the pulsed-dye laser. Additionally, any superficial proliferating hemangiomas of the face or other cosmetically sensitive area should be carefully considered for this treatment. The tunable pulsed-dye laser produces a wavelength of 585 to 600 nm; this wavelength corresponds to the second absorption peak of hemoglobin and the third absorption peak of oxyhemoglobin, allowing for selective photothermalysis. At the shorter wavelength of 585 nm, most of the energy is absorbed by the first 0.5 to 1 mm of tissue, thereby limiting the response to the most superficial component of the hemangioma. As the wavelength is increased from 585 to 600 nm, the laser light will reach almost twice the depth, theoretically resulting in a more favorable response for hemangiomas that, although still superficial by definition, display thickened papular growth. Currently, a wavelength of 595 nm is used to treat most lesions. First-generation lasers used a pulse length of 450 to 500 microseconds, significantly shorter than the thermal relaxation time of skin of 700 to 900 microseconds, and therefore very safe. Second-generation lasers increased the pulse time to 1500 microseconds in an effort to increase clinical efficacy for larger vessels. This laser has also proven to be extremely safe. The addition of a cryogen spray to the pulse-dye laser allows cooling of the superficial epidermis while allowing continued penetration of laser energy to the lesion and effective thermal ablation. The dynamic cooling device (DCD) is attached to the hand piece of the pulse-dye laser and delivers a brief (20 to 30 millisecond) liquid cryogen pulse. Theoretically, the DCD allows cooling of the epidermis, and therefore less thermal injury results, with reduced risk for scarring and less discomfort. Clinically, when using the device, the purpura produced by the start-safe parameters is greatly reduced, allowing treatment at a fluence 2 to 3 J/cm² higher than start-safe parameters, theoretically improving results. Experience will determine the appropriate fluence to be used with each lesion; the starting fluence with a 5-mm hand piece is usually ~9 J/cm², and this is adjusted upward until the desired degree of purpura is reached (Fig. 5). If a 7-mm hand piece is used, the fluence should be started ~1 J/cm² lower.

Several articles have been published on the use of the pulsed-dye laser for the treatment of superficial proliferating hemangiomas. Glassberg in 1989 published the first article presenting a single case example of a large superficial hemangioma. Haywood treated 39 superficial hemangiomas with cutaneous height above the surrounding skin of no more than 5 mm and no deep component. The mean age at first treatment was 13 weeks, the first sign of involution were seen at an average of 19 weeks, and complete resolution was seen in 61.5% of treated hemangiomas at an average age of 39 weeks following a mean of 2.4 treatments. Although no control group was provided and so the data must be interpreted with caution, this treatment appears to induce resolution of at least a select group of hemangiomas much earlier than historical data would suggest. Additionally, Lacour used the pulsed-dye laser for the treatment of
and kaposiform hemangioendothelioma and not juvenile hemangiomas; visceral hemangiomas; and those undergoing multiple simultaneous treatments.

Although admittedly this retrospective review has limitations as pointed out in the article, several useful conclusions were apparent. Administration of prednisone more than 2 to 3 mg/kg per day resulted in a 75% response. Dosage greater than 3 mg/kg per day resulted in 94% response, but greater adverse effects (51%). Lesser dosing resulted in fewer adverse effects but also fewer responses, and rebound growth occurred in 70% of patients. Potential side effects include Cushingoid features, growth retardation, gastroesophageal reflux, peptic irritation and ulceration, fluid and electrolyte disturbances, hypertension, hyperglycemia, behavioral disturbances, and immune suppression. Despite these possibilities, no life-threatening adverse effects were observed and many authors reported no adverse effects.

Many proposed regimens of steroid dosage and tapering have been used, and as stated, the optimum regimen is unknown. A regimen that has proven effective and safe follows. A starting dose of prednisone 4 mg/kg per day in a single dose if possible is used. The patient’s hemangioma is then reevaluated in 1 week for steroid response. If shrinkage or stabilization is noted at this time, the treatment is maintained at the initial dose for 3 weeks. If no response is seen, an increase in dosage to 5 mg/kg per day may be attempted and reexamined in 1 week. On follow-up exam at 3 weeks, the steroid is tapered over 4 to 8 weeks. The patient is observed closely by the primary care physician or pediatrician for antireflux management (appropriate doses of a H2 blocker or proton pump inhibitor are recommended) and frequent well-baby checks. The patient should be observed by an endocrinologist as well. A follow-up exam is performed every 4 weeks or sooner if parents note problems. During treatment, no live vaccines are given, and parents are informed that the infant is immunocompromised. If during tapering rebound proliferation is noted, the dosage should be increased to the next highest level for an additional week and then tapering should be attempted again. One month after steroid treatments are discontinued, the patient is reevaluated. If the hemangioma exhibits proliferation, systemic steroid treatments are resumed at a dose of 4 mg/kg per day. The hemangioma should be reevaluated at 1 week and, if improvement has occurred, systemic steroids are tapered over 4 weeks (Fig. 6A,B).

MANAGEMENT OF INVOLUTING HEMANGIOMAS

Involution will begin usually no later than 12 months but occasionally as late as 18 months of age. As the lesion enters involution, it becomes necessary to observe...
the process for 8 to 12 months. As explained earlier, hemangiomas will involute at different rates, with ~50% doing so by age 5 and 50% more slowly than that (Fig. 7). As the hemangioma is monitored during early involution, it becomes apparent from serial photography and parent questioning whether the lesion is remaining stable in size or has begun to regress. Those that show regression by the age of 2 are classified as early involuters, and those not showing regression are classified as late involuters. Those lesions that are determined to be early involuters are monitored approximately every 6 months until age 4 or 4½ when if substantial deformity still exists, surgical therapy is offered for atrophic skin or fibrofatty residuum or laser therapy is offered for telangiectasias to attempt to resolve the abnormality completely prior to entrance in primary school. Approximately 20% of early involuting lesions will fall into this category, and 80% will resolve completely without any abnormal residuum.21 Hemangiomas that have not shown any regression in size after 8 to 12 months are determined to be late involuters. These are offered surgical excision or laser treatment around the age of 2 because a majority of these lesions will leave a substantial cosmetic deformity by the time of entrance to primary school. By intervening at the age of 2, the child has not yet begun to fully develop a body image, and little recognition of the treatment will likely persist past early childhood. The overall goal of intervention in the natural cycle of the hemangioma is to identify as readily as possible those lesions that will most likely leave a cosmetic deformity as the child progresses to school age, where it is likely to cause psychological detriment, and then to intervene in such a manner to leave the child with little if any noticeable remnant of the ordeal either physically or mentally.

When undertaking surgical excision of a hemangioma, several surgical principles are important to consider. First and foremost, unlike other vascular lesions, hemangioma excisions more resemble that of a benign tumor than a vascular lesion. Although no capsule exists around the hemangioma, an easily definable surgical plane does. This plane can be followed, and with careful identification and management of a few feeding vessels, very little bleeding should be encountered. Bipolar cautery is recommended, and although laser or “hot” scalpels may be used, they are certainly not necessary tools. Blood products are not necessary with the exception of perhaps a very large proliferating hemangioma related to the orbit as described. Placement of incisions should be done so as to include any abnormal atrophic scar tissue from previous ulceration and so that they lie in the junction of facial units, subunits, or relaxed skin tension lines. The incision line can often be shortened with the use of an M-plasty at one or both extremes of the incision (Fig. 8A,B). An advantage is gained when excising hemangiomas in the 2- to 3-year-old time frame because the tumor itself acts as a tissue expander, and adequate tissue is nearly always available to be advanced as necessary to primarily close the excision (Fig. 9A). When the hemangioma has an extensive superficial component and the entire lesion cannot be excised without violating aesthetic lines or making an unduly large incision, it is always better to leave hemangioma behind and rely on the natural involutorial process to complete the result. Routinely ~10% of the hemangioma is left behind and allowed to undergo involution to prevent overresection and tissue void (Fig. 9B,C). The pulsed-dye laser can be used postoperatively to effectively create tissue by converting telangiectatic skin

Figure 6  (A) Infant with rapidly proliferating and ulcerated central lip hemangioma. (B) Same infant after several weeks of systemic corticosteroids, showing decrease in size of hemangioma and Cushingoid features.

Figure 7  Graph of early versus late involuters.
Figure 8  (A) Large, late involuting upper lip hemangioma. (B) Use of M-plasty to shorten superior limb of incision avoiding extension into the nasal sill.

Figure 9  (A) Demonstration of significant tissue expander effect and determination of safe excision for primary closure. (B) Planned incision making no attempt to excise the entire hemangioma and allowing for postoperative involution and tissue creation with pulsed-dye laser. (C) Operative closure with residual hemangioma evident but also significant improvement.
or skin with residual hemangioma to normal–appearing skin. Mulliken recently published an article describing the use of a purse-string stitch to excise the hemangioma initially and gather the surrounding tissue, taking advantage of the tissue expander phenomenon. Following this, if necessary, a second operation is planned to remove the scar created by the purse string. This method may prove useful for lesions placed in the central cheek or forehead as many of the examples in the article illustrate; however, its use when incisions may be placed in facial unit junctions may be of less benefit.

Once the surgical plan has been made, the tissue is injected with local anesthetic with a vasoconstricting agent. The incision is usually carried down through skin and into the substance of the hemangioma, where it is transected by using a small hemostat to bluntly separate the tissue and a bipolar cautery to divide. No attempt is made to raise a skin flap over the substance of the hemangioma as the dermis is often involved, leading to an overly thin flap with the risk of necrosis. Once the deep aspect of the hemangioma is encountered, an easily dissectible tissue plane is the rule. The lesion is then separated from the surrounding tissue and any feeding vessels appropriately dealt with. As the superficial aspect of the lesion is encountered again, a portion is intentionally left on the undersurface of the skin flap, and no attempt to identify a tissue plane is made. Any undesirable tumor bulk that is left on the undersurface of the skin flaps is then thinned to a safe thickness with scissors and the residual cauterized with a bipolar cautery to induce further involution (Fig. 10). It appears and has been mentioned by other authors as well that surgery seems to influence the speed of involution even on any residual left after a debulking, although this cannot be substantiated by any scientific study.

Hemangiomas of the nasal tip, lip, and periorbital tumors pose an additional challenge to excision and reconstruction. Bowers et al demonstrated with near significance that hemangiomas of the lip fail to involute more often than other areas. Many of the same surgical principles already mentioned apply but have specific application to the lip anatomy. As hemangiomas grow within the red or white lip, expansion of the lip occurs. Removal of the lesion requires that both the vertical and horizontal proportion of the red and white lip be adjusted to their normal anatomic relationship. Once again, conservatism is the rule and overresection should be avoided. It is advisable to not resect orbicularis muscle unless absolutely necessary, as this often leads to volume loss within the lip, which cannot be easily replaced (Figs. 11A and 12A,B). Placement and reconstruction of the white roll is essential in lip reconstruction as malposition of this border is obviously perceptible.

Perhaps no other region may be affected or invite ridicule and social isolation more than the nasal tip. Hemangiomas in the nasal region tend to proliferate rapidly, often reaching large size relative to the native nasal skeleton, and will often splay if not cause frank deformity to the lower lateral cartilages. These lesions will often be classified as late involutes, and because of the psychological impact of allowing involution to go on into the late elementary school ages, they are often amenable to surgical excision. Thompson compared eight patients with nasal tip hemangiomas managed without surgery with 11 patients with surgery via various methods and concluded that conservative management provided superior results. Since that time, however, greater understanding of nasal anatomy and subunits has been gained and applied to reconstructive nasal surgery. Several approaches to the excision of nasal hemangiomas have been described with varying advantages, disadvantages, and results. The procedure chosen must be able to accomplish the following goals. The incisions must be placed within the aesthetic subunit junctions of the nose; they must provide access for complete removal of the

Figure 10  (A) Thinning of skin flaps to safe thickness, still leaving small residual tumor.

Figure 11  View of planned excision of intraoral portion of red lip vertical and horizontal excess in conjunction with deep hemangioma.
hemangioma, and they must allow for both horizontal and vertical trimming of the redundant nasal soft tissue envelope for repositioning. Care must be taken when performing the dissection to preserve adequate thickness to the nasal soft tissue flap for survival. Once again it is not necessary to define a tissue plane between the skin and the hemangioma. Conversely, the deep aspect of the lesion will provide an identifiable dissection plane above the nasal cartilaginous skeleton that must be preserved. Once the hemangioma is removed, the alar cartilages should be repositioned and sutured to place the domes in anatomic approximation. Following this, the soft tissue envelope is repositioned, trimmed, and sutured into position (Figs. 13A,B and 14A,B).

Periorbital lesions represent an entire body of literature, and prompt consultation with ophthalmologic and oculoplastic colleagues should ensue. Hemangiomas are the most common tumors of the orbit in children. In addition to their cosmetic significance, they carry a more ominous threat of visual disability. Because of this threat, treatment in proliferation, either medical or surgical, is often warranted.

**Figure 12** (A) Preoperative photo of large upper lip hemangioma. (B) Several years later demonstrating cosmetic outcome.

**Figure 13** (A) Lateral view of large nasal tip hemangioma with alar subunit defined. (B) Lateral view with cutaneous flap elevated demonstrating superior cutaneous extensions of marginal incision in proposed subunit junction.

**CONCLUSION**

Pediatric vascular lesions including hemangiomas and other vascular malformations have been called by many different names and confused in the literature for many years. This confusion in terminology also led to misdiagnosis and inconsistency of treatment. Mulliken and Glowacki fortunately clarified the different endothelial characteristics of these lesions, allowing them to be classified as true hemangiomas or vascular malformations. This microscopic classification scheme hence has allowed the clinical course and treatment of hemangiomas to be followed with clarity. Unfortunately, hemangiomas are still at times referred to in the literature as *capillary* or *cavernous*, terms that were used in the past to describe differing vascular lesions. Hemangiomas are a single clinical entity whose presentation only differs because of their depth within the soft tissue. The terminology of superficial, deep, and compound to differentiate these different clinical presentations of the same pathological entity is therefore sensible.

Hemangiomas are the most common neoplasm of infancy and early childhood, and their clinical course is usually completely benign both functionally and cosmetically. However, historical papers describing the natural history of hemangiomas prior to the advent of
newer medical, laser, and surgical treatments probably overestimated the complete resolution of these lesions. When these studies are compared with more recent projects, the natural history of hemangiomas appears unchanged, but the interpretation of the data is quite different. It becomes apparent on review that during the involutorial stage of the hemangioma, two groups emerge: those that will resolve by the age of 5 and those that will not. About 50% of hemangiomas will fall into each group; it is recommended that these hemangiomas should be referred to as early and late involuters. Additionally, a difference in cosmetic outcome exists between groups with ~20% of early involuters versus 60% of late involuters leaving a cosmetically imperfect result. With newer medical technologies, the expectation of attainable outcomes should be elevated. Psychological studies of children with facial hemangiomas bolster the argument that in certain cases intervention in the natural history of the hemangioma should be undertaken.

Intervention during proliferation should be offered for rapidly proliferating lesions in cosmetically sensitive areas, ulceration or pending ulceration, or impairment of function. The most urgent reason to intervene during proliferation is impending visual impairment. The available options for intervention are currently many and have been even more numerous in the past; however, the mainstay of therapy consists of oral or intralesional corticosteroids, pulsed-dye laser, and surgical excision. Once a hemangioma has begun to involute, determination should be attempted to place the lesion into the category of early or late involution. It must be emphasized that 60 to 75% of hemangiomas will involute without any cosmetic sequela, and for that reason every attempt should be made to identify those lesions that do not need any further intervention. However, once it becomes apparent that a cosmically un-desirable outcome is likely, surgical or laser therapy should be offered. Surgery if undertaken is not an unduly hemorrhagic affair. Definable surgical planes are readily apparent, and complete excision is not necessary or even desirable. Minute amounts of blood loss are easily controlled with standard surgical techniques. Surgical planning is paramount, and principles of facial units, subunits, and relaxed skin tension lines should be observed. Abnormal skin through which ulceration or atrophic scarring exists should be excised without effort to include all skin involved by the superficial component of the hemangioma. The normal involutorial process, which may be encouraged by surgery and the use of the pulsed-dye laser postoperatively, will actually produce additional normal skin as time passes.

Hemangiomas should not be routinely disregarded and assumed to resolve of their own accord. Physicians must be reeducated regarding the usefulness of active intervention for these tumors even if that intervention consists of thoughtfull monitoring and family education. When indicated, referral to a physician experienced in treatment should be commonplace early in the evolution of the process and not only when an unacceptable residuum exists in an already impacted child.

REFERENCES


