Vascular Lesions

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KEYWORDS

• Vascular lesions • Vascular malformations • Hemangiomas • Port wine stains
• Lymphovascular malformations • Capillary malformations • Vascular defects

KEY POINTS

- Classification of vascular lesions based of off the biological behavior has greatly facilitated more accurate diagnoses, optimally defined treatment plans, and better outcomes.
- Treatment of vascular lesions has taken a more conservative surgical approach with reliance on select medical treatment options, which has greatly reduced morbidity and mortality resulting from extensive surgery.
- A multidisciplinary approach involving multiple surgical and pediatric subspecialties has led to advancement in both understanding and ideal treatment strategies of these lesions.

INTRODUCTION

The study of vascular lesions has spanned many medical and surgical subspecialties within academia over the last several decades. Much of the understanding regarding the nonsurgical management of these lesions has been predicated by pediatric subspecialties and complemented by advances in surgical management of these lesions and vice versa. Furthermore, because many of these lesions have involved the craniofacial region, this area has been thoroughly investigated by surgical subspecialties, including facial plastic surgery along with oral maxillofacial and craniofacial surgery.

Since the advent of the current classification system approved by the International Society for the Study of Vascular Anomalies in 1996 (and updated in 2014), relatively few advances in treatment of these lesions have occurred.[1] This classification system was derived from Mulliken and Glowacki's system,[2] which was published in 1982 and based on the biological behavior of the lesions. In general, the overlying theme has been toward more conservative surgical approaches and a greater reliance on the advances in medical management, which in turn has led to more effective treatment with a greatly reduced morbidity.

Specifically, the lesions can be divided into vascular tumors, including the more common infantile hemangiomas, rapidly and noninvoluting congenital hemangiomas, and kaposiform hemangioendotheliomas, among others; and vascular malformations, including low-flow venous, lymphatic, and capillary/port-wine stain subtypes, high-flow arteriovenous malformations (AVM), and combined complex capillary-venous, capillary-arteriovenous, and lymphaticovenous subtypes. In this article, the more common lesions, and specifically the classification, diagnosis, and management, are focused on, with an emphasis on an aesthetic approach regarding surgical technique when indicated (**Box 1**).

VASCULAR TUMORS: HEMANGIOMA

Considered the most common tumor of infancy and childhood, hemangiomas have an estimated

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Box 1
Vascular anomalies

Vascular tumors

- Infantile hemangiomas
- Congenital hemangiomas
  - Rapidly involuting
  - Noninvoluting
  - Partially involuting
- Kaposiform hemangioendothelioma
- Others

Vascular malformations

- Low-flow
  - Venous malformations
  - Lymphatic malformations
  - Capillary/port-wine stain malformations
- High-flow
  - Arteriovenous malformations
- Combined complex
  - Capillary-venous
  - Capillary-arteriovenous
  - Lymphaticovenous

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prevalence of 10% by age 1, with 30% evident at birth, and a female predilection of 4:1.[3] Ninety percent of all vascular tumors are infantile hemangiomas.[4] Although 40% to 60% involve the head and neck, 80% of these lesions tend to be solitary.[3] There are various syndromes that can be associated with hemangiomas, including PHACES (*p*osterior fossa malformations, *h*emangiomas, *a*rterial anomalies, *c*oarctation of the aorta and cardiac defects, *e*ye abnormalities, and *s*ternal defects), and other symptoms such as stridor in a patient with cutaneous hemangiomas should be further investigated along with the potential for visceral hemangiomas and hemangiomas overlying the lumbosacral spine due to the increased association with spinal cord and genitourinary anomalies.[3] Imaging studies, such as screening ultrasounds and confirmatory MRIs, should be strongly considered in these patients.[5]

Historically, treatment of these lesions involved the use of various modalities. Many investigators advocated for aggressive management with surgical excision, cauterization, carbon dioxide snow, surface radium, radioactive implants, external beam radiation, interstitial gamma radiation, and sclerosing agents. However, serious complications arose, such as malignant transformation after radiation therapy and poor cosmetic results due to significant scar formation secondary to resection and other ablative procedures. Conversely, proponents of more conservative benign neglect strategies argued that the natural course of most hemangiomas included involution with no residual deformity. The stark polarity in treatment algorithms was centered around the fact that vascular tumors and malformations were often categorized as similar or related entities—for example, the term capillary hemangioma was used to describe port-wine stains, which are capillary malformations, whereas strawberry nevi and cavernous hemangiomas were used to describe true hemangiomas. [5] This early misclassification of vascular lesions is what led to a lack of consensus in regards to management, which again was addressed by Mulliken and Glowacki's delineation of vascular tumors (hemangiomas) and malformations based on biological behavior, resulting in the current classification.[1]

CLINICAL PRESENTATION AND DIAGNOSIS

Infantile hemangiomas are usually visible within the first few weeks of life, whereas congenital hemangiomas are fully formed at birth and grow with variable intensity. Superficial hemangiomas are contained within the papillary dermis and are characterized by bright red, macular, or papular lesions with well-defined borders. The macular variety can be confused with port-wine malformations; however, over time the hemangioma will change in size, whereas the port-wine malformation remains relatively stable. Deep hemangiomas are contained in the reticular dermis or subcutaneous tissue and present as a blue, subcutaneous mass with bluish or colorless overlying skin depending on the depth. These hemangiomas may appear similar to lymphatic malformations on examination. Compound hemangiomas have features of both superficial and deep lesions (**Fig. 1**).[3,5]

Hemangiomas exhibit 2 distinct clinical stages, including proliferation and involution. Proliferation occurs during the first 12 months of life and occasionally as late as 18 months. An initial growth phase during the first few months of life followed by a subsequent growth phase at 4 to 6 months establishes a bimodal pattern of growth. Cosmetic deformity or functional obstruction of the eye,

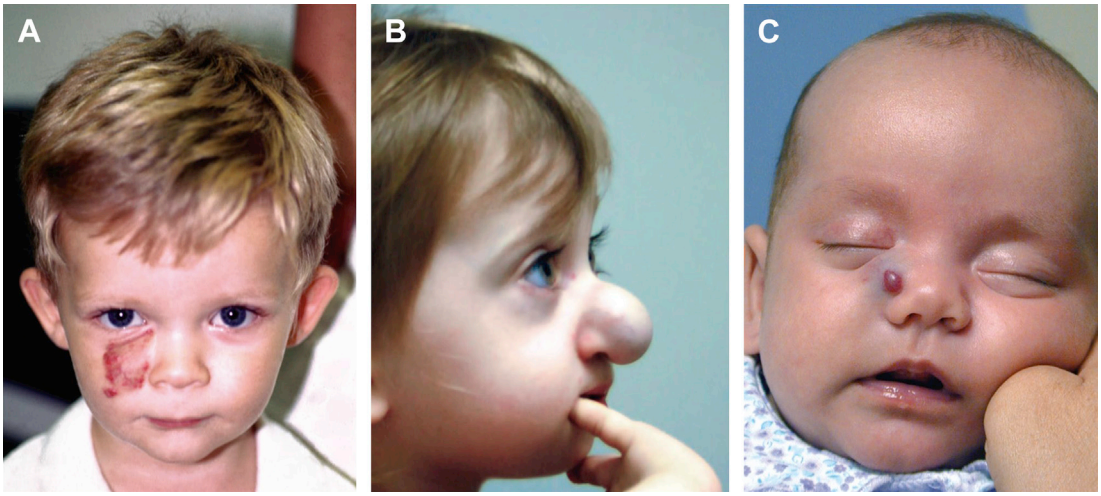

Fig. 1. (*A*) Superficial hemangiomas are bright red macular or papular lesions. (*B*) Deep hemangiomas present as bluish, subcutaneous masses. (*C*) Compound hemangiomas exhibit features of both superficial and deep hemangiomas.

nose, or airway typically arises during the first growth phase (**Fig. 2**).[3] Histologic features during the proliferative phase include plump proliferating endothelial cells and pericytes with barely perceptible vascular channels.[5] Involution, occurring over the first several years of life, is characterized by a decrease and then cessation in growth of the lesion. The lesion changes from bright red to dark maroon and eventually patches of ashen gray, evolving from a firm, tense consistency to a lobular, soft, compressible mass on palpation. Histologically, there is a gradual flattening of endothelial cells with progressive deposition of fibrous tissue and vessel ectasia, resulting in superficial telangiectasia and subcutaneous fibrofatty residuum (**Fig. 3**).[3]

Although imaging is not required for diagnosis, it can be useful to evaluate the extent of involvement in deeper lesions as well as rule out other vascular tumors. Ultrasound findings demonstrate a solid

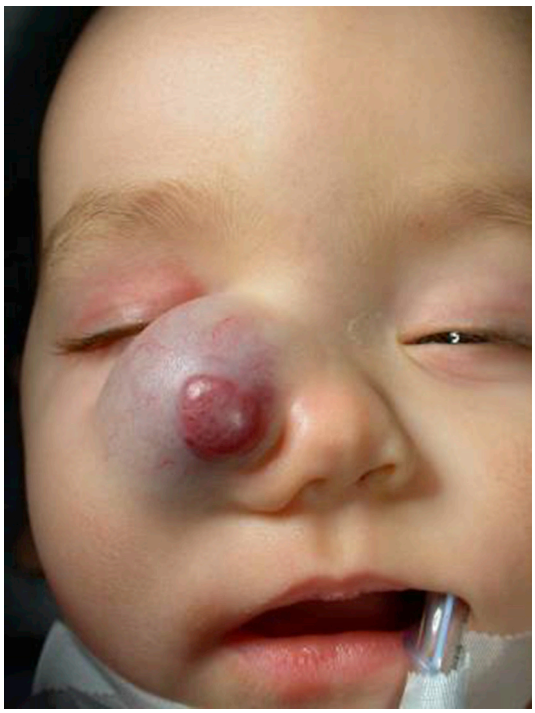

Fig. 2. Rapidly proliferating hemangioma in the same infant in **Fig. 1**C, 5 months later, unresponsive to intralesional steroids or PDL, now with astigmatism, partial visual obstruction, and impending amblyopia.

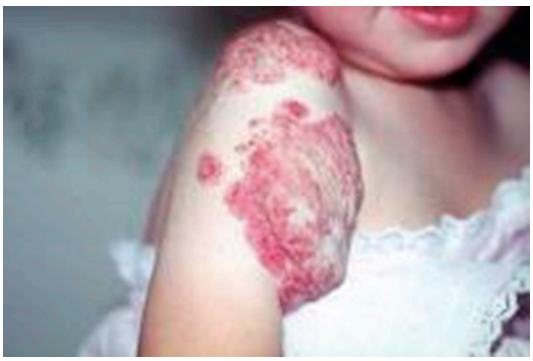

Fig. 3. Late involuting hemangioma demonstrating darkening skin color and patches of ashen gray. Significant subcutaneous fibrofatty residuum will require surgical excision.

mass with increased/high color flow within it; Doppler ultrasound can demonstrate the arterial feeder and venous drainage. MRI characteristics include lesions that exhibit enhancement on T2-weighted images, which are relatively isointense on T1-weighted images with homogenous contrast enhancement; internal serpiginous flow voids within the lesion in T2-weighted images represent the arterial feeder, which can be an important diagnostic clue.[4]

The diagnosis of infantile hemangiomas can also be assisted with the use of immunohistochemical markers because endothelial cells in these lesions stain strongly for the glucose transporter protein isoform 1, whereas most other vascular tumors, such as congenital hemangiomas, do not.[4]

The psychology of children with these lesions has also been studied. It is known that children begin to develop self-awareness at 18 to 24 months of age with a significant body image well under development by 3 years of age.[6] Individual studies by Williams and colleagues[6] and Dieterich-Miller and colleagues[7] comparing children 3 to 5 years of age with hemangiomas of the head and neck with unaffected children found that children with hemangiomas perceived that others valued them significantly lower compared with the unaffected group. Furthermore, parental interviews revealed reports of strangers questioning child abuse, children burying their faces or hiding lesions with their hair, and family and friends commenting openly on intervention.[6,7] Preventing the aforementioned scenarios makes diagnosis, classification, and effective treatment of these lesions extremely important in the pediatric population with respect to psychosocial affects.

TREATMENT: HEMANGIOMAS

Treatment of hemangiomas historically has involved various approaches, including surgical excision, steroids, and benign neglect; current therapy has evolved to include interventions such as laser therapy and propranolol. Review of the literature on pediatric facial hemangiomas revealed 25% to 40% will result in an unacceptable aesthetic outcome when no intervention is performed.[5] Finn and colleagues[3] found that of 50% of lesions that involuted early (before age 5), 19% resulted in a substantial aesthetic deformity, including residual scar, redundant skin, or telangiectasia, whereas of the 50% that involuted late (after age 5), 60% were cosmetically unacceptable.

The treatment algorithm is dependent on the classification of the lesion, and deciding when to observe or intervene is the most important decision (**Fig. 4**).[3] If observation is determined to be the approach, then unlike the practice of benign neglect, it must be an active process with routine monitoring every 3 months. This frequency of evaluation must be increased to every 2 to 4 weeks with demonstrated interval change or for a lesion in a cosmetically sensitive area. The decision to treat proliferating lesions should be based on the rate of proliferation, presence of or pending ulcerating, and aesthetic result in the setting of possible involution with no resulting deformity. Goals for therapy include reduction in size and induction of involution. The mainstay of interventions for these lesions includes intralesional or systemic steroids, pulsed dye laser (PDL), and surgical excision.

Hemangiomas that are involuting require monitoring for 8 to 12 months. Serial photography and questioning on history during early involution will reveal regression or provide information on the degree of stabilization of the lesion. Early involuters show regression by age 2, whereas later involuters do not have evidence of regression by this time. Hemangiomas that have not somewhat regressed in size by 8 to 12 months are most likely late involuters.[5] Early involuters are monitored every 6 months until 4 to 4.5 years of age, at which time surgical treatment for atrophic skin or fibrofatty residuum or PDL treatment for telangiectasia can be pursued. Approximately 20% of early involuters require additional intervention for an aesthetic deformity, whereas 60% of later involuters leave a cosmetically imperfect result.[5] The mainstay interventions for these lesions include observation, PDL, and surgical excision.

PDL therapy is indicated for more superficial hemangiomas that rapidly proliferate, ulcerate, or demonstrate pending ulceration, lead to nasal airway obstruction, or are located in more cosmetically sensitive areas (**Fig. 5**). Treatment of proliferating ulcerative hemangiomas is efficacious because of their inhibitory effect on the growth rate of the hemangioma, which allows re-epithelialization to occur.[3,5] Treatment is delivered at 4- to 6-week intervals until resolution of ulceration or cessation of proliferation. A wavelength of 595 nm is selected to allow deeper penetration into the thickened aspects of the hemangioma, and selective photothermolysis at this wavelength corresponds to the second and third absorption peak for hemoglobin and oxyhemoglobin, respectively.[3] Starting with a fluence around 9 J/cm^2 with a 5-mm hand piece or 8 J/cm^2 for a 7-mm hand

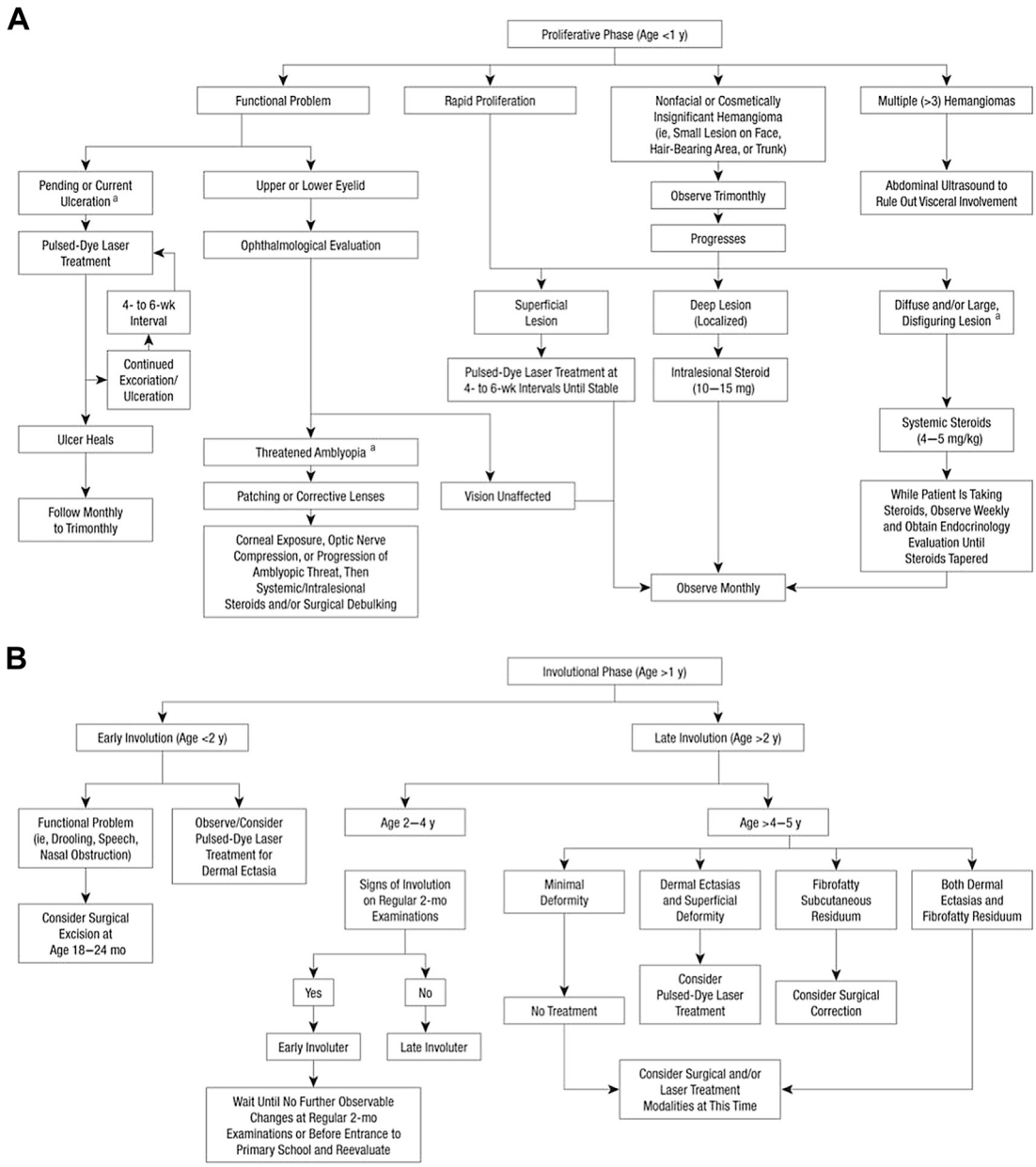

Fig. 4. Algorithm developed by the senior author (E.F.W.) for management of hemangiomas during the proliferative (*A*) and involutional phase (*B*). [a] Consider initiation of propranolol in infantile hemangioma. (*Adapted from* Batniji RK, Buckingham ED, Williams EF. An aesthetic approach to facial hemangiomas. Arch Facial Plast Surg 2005;7:301–6; with permission.)

piece, this should be increased until the desired degree of purpura is achieved. The addition of a cryogen spray via a dynamic cooling device (DCD) attached to the hand piece allows cooling of the superficial epidermis with continued penetration of laser energy to the lesion, theoretically resulting in less superficial thermal injury, scarring, and discomfort. It has been the authors' experience and observation that use of the DCD can diminish the effect of the laser on the lesion, and for select lesions, they do see better treatment response with deactivation of the DCD. One should be extremely mindful of fluence levels and tissue response if the DCD is deactivated because the potential for thermal injury is greatly increased. Of note, first-generation lasers have a pulse time of

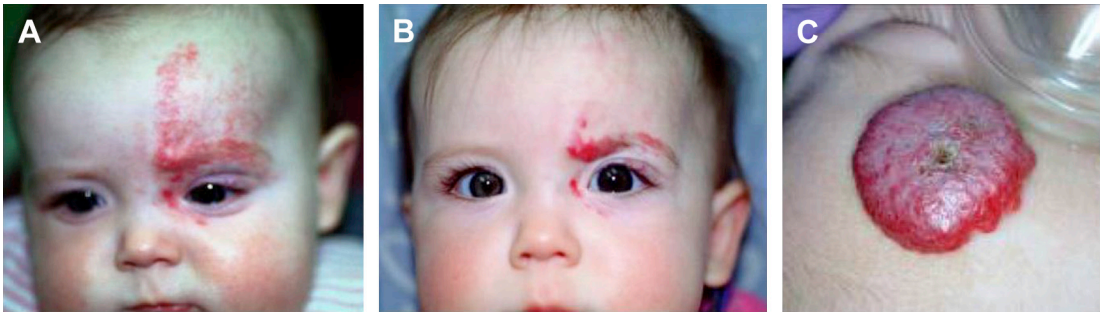

Fig. 5. Indications for PDL during the proliferative phase. (*A, B*) Prelaser and postlaser ablation of a superficial hemangioma in a potentially cosmetically disfiguring area. (*C*) Presence of ulceration or impending ulceration. Can also consider initiation of propranolol therapy if not medically contraindicated.

450 to 500 microseconds, which is significantly shorter than the thermal relaxation time of skin of 700 to 900 microseconds, thus increasing the safety profile for these lasers, but second-generation lasers have an increased pulse time of 1500 microseconds to increase efficacy for larger vessels.[3] As the PDL is relatively ineffective for deeper lesions due to the inability to penetrate tissue to the necessary depth, intralesional potassium titanyl phosphate or Nd:YAG lasers have been suggested; however, the concern for scarring and damage to underlying structures is greatly increased.

For deeper or more compound lesions, systemic or intralesional steroids can be used, especially for rapidly proliferating and aesthetically or functionally debilitating lesions. These medications were used historically but have been replaced by safer, more effective alternatives. Kenalog is used primarily for intralesional injection and is delivered to the bulk of the tumor. Ten milligrams per milliliter in a 1-mL syringe with a 27′ or 30′ needle is used to deliver a total of 10 to 15 mg depending on the size of the lesion, or roughly 2 to 3 mg/kg. The treatment is repeated every 4 to 6 weeks until the lesion stabilizes, decreases, or requires additional intervention. In order to prevent increased bleeding with the injection, ensure that the needle is passed through normal skin, avoiding the thinned epidermis involved with the lesion. If you opt for dual therapy with use of the laser, laser treatment should be performed first in order to avoid interference of injection-associated bleeding with accuracy of laser pulses. Systemic steroids can also be considered for proliferating hemangiomas. The optimal dosing regimen has not been completely delineated; however, increased responsiveness has been noted with higher dosing but at the expense of greater adverse effects. Side effects such as Cushingoid features, hypertension, hyperglycemia, immune suppression, behavioral disturbances, and growth retardation are observed and increased in likelihood with higher and prolonged steroid dosing. The authors propose an effective and safe regimen consisting of prednisone 4 mg/kg as a single starting dose. This dose is maintained for 3 weeks if stabilization or shrinkage of the lesion is observed or can be increased to 5 mg/kg daily, if tolerated, for a week if no response is noted. At around the 3-week follow-up, the steroid is tapered over 4 to 8 weeks. If rebound proliferation occurs during tapering, the dosage should be increased to the next highest level for an additional week, and then tapering can be reattempted. Ideally, this should be done in collaboration with the primary care physician or pediatrician and endocrinologist. Antireflux prophylaxis should be initiated and live vaccines avoided. The patient should be followed monthly, and the lesion re-evaluated after completion of the taper. A dose of 4 mg/kg daily can be resumed for 1 week if there is continued proliferation but should be tapered over 4 weeks if improvement is noted.[3,5] Although there are indications for steroid use, treatment has fallen out of favor because of the adverse side effects and has been replaced largely by the use of propranolol.

Propranolol, which is traditionally used by cardiologists in infants with tachyarrhythmias, has been shown to be an effective treatment for select hemangiomas.[8] Propranolol has been found to be rapidly effective for infantile hemangiomas and well tolerated, with the most frequently reported serious complications including hypotension, bronchial hyperreactivity due to inhibition of adrenergic broncodilation, hypoglycemia or hypoglycemic seizure, asymptomatic bradycardia, and hyperkalemia.[8] Commonly reported adverse effects include sleep disturbances, diarrhea, and gastroesophageal reflux disease.[8] The mechanism of action has not been clearly elucidated with

various proposed hypotheses, including vasoconstriction, decreased renin production, inhibition of angiogenesis, and stimulation of apoptosis proposed.[8] Consensus recommendations from 2011 encourage treatment of ulceration, ocular compromise, airway obstruction, and risk of permanent disfigurement and should be initiated and managed by a medical team familiar with hemangiomas and the medication in pediatric populations.[8] The dose is started low with escalation to a target dose of 1 to 3 mg/kg daily in 3 divided doses.[8] Initiation should be considered with inpatient hospitalization in infants less than 8 weeks of age or any infant with inadequate social support or comorbid conditions.[8] Peak effect on blood pressure and heart rate occurs 1 to 3 hours after administration.[8]

Surgical excision has also been an effective treatment option for carefully selected lesions. Planning is paramount when considering surgical intervention and should be considered during involution in order to minimize blood loss. Incisions should be placed in the junction of facial subunits or relaxed skin tension lines to conceal scars. Atrophic scar tissue and ulceration should be incorporated into the skin excision, and careful identification and management of feeding vessels with bipolar cautery will result in less blood loss and avoidance of blood products. Use of M-plasty technique can shorten incision lines (**Fig. 6**).[3,5] Ten percent of the lesion should be left behind if full excision will result in violation of aesthetic lines or an unsightly incision. The remaining lesion can be observed or treated with PDL (**Figs. 7** and **8**).[3,5] When considering treatment of hemangiomas in the periorbital region, management should include consultation with an ophthalmologist. Treatment is indicated with amblyopia or rapid proliferation (**Fig. 9**). Surgical management is reserved for functional concerns and nonresponders to steroid or laser therapy. If orbital structures are compromised, then a portion of the lesion is left behind and treated with intralesional steroid injections or PDL therapy.[5] Special consideration should also be given to nasal and lip hemangiomas with respect to placement of incisions within aesthetic subunits and avoidance of aggressive resection (**Fig. 10**).

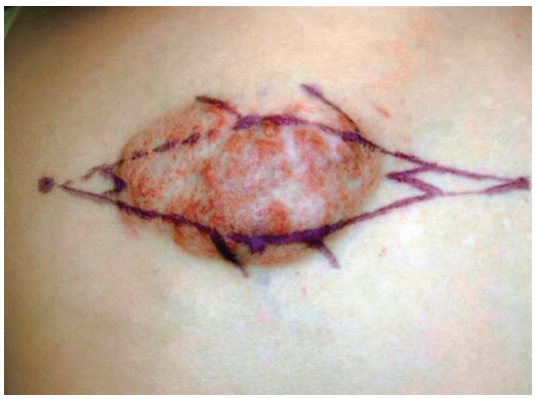

Fig. 6. Z-plasty is used for scar camouflage and M-plasty is used to shorten the length of the incision line.

VASCULAR TUMORS: KAPOSIFORM HEMANGIOENDOTHELIOMA

Kaposiform hemangioendothelioma is a rare vascular tumor that is present at birth or within the first few months of life, but may also present later in childhood.[4] It presents as an ill-defined, purpuric solid mass that is often painful and destructive—infiltrative growth differentiates this lesion from a hemangioma.[4] Histology reveals cells that form a slitlike lumina containing erythrocytes resembling Kaposi sarcoma.[4] It is associated with Kasabach-Merritt phenomenon in up to 50% of patients with a high mortality due to coagulopathy or complications from local tumor infiltration.[4] Imaging reveals a solid mass with ill-defined borders and variable echogenicity on ultrasound, whereas MRI usually demonstrates a lesion involving multiple soft tissue planes with skin, subcutaneous fat, muscle, and bony cortex infiltration.[4] Treatment is primarily medical because the tumor is usually too extensive for surgical resection.[9]

VASCULAR MALFORMATIONS

Vascular malformations are vascular lesions that are present at birth with a growth rate that is dependent on the principal type of vessel and proportional to the individual without spontaneous regression.[8] These lesions can experience rapid expansion when triggered by hormonal factors, trauma, or infection, but can also remain dormant with a presentation later in childhood or adulthood.[4] Histology reveals vascular spaces lined with flat, mature, mitotically quiescent epithelium with the subtype classified by the constituent vessel, presence or absence of arteriovenous shunting, and flow dynamics—specifically low flow versus high flow.[4]

Venous, lymphatic, and capillary malformations comprise low-flow lesions. Venous malformations present as soft, compressible, bluish lesions that infiltrate multiple tissue planes (**Fig. 11**). They may enlarge with Valsalva maneuvers and exhibit skin involvement. Spontaneous thrombosis and

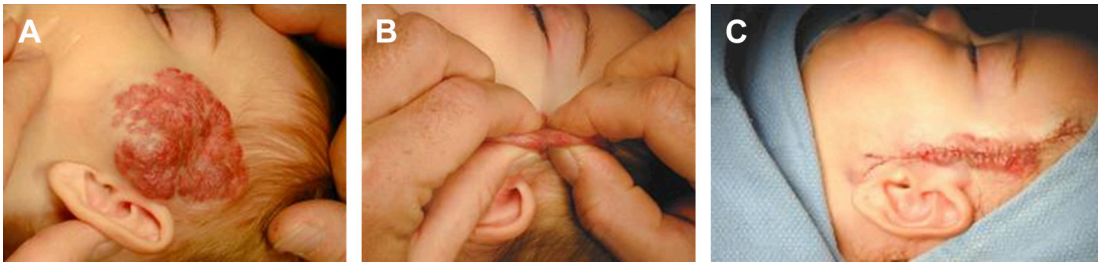

Fig. 7. (*A, B*) Significant tissue expansion effect of lesion. (*C*) Residual hemangioma after closure allows for postoperative involution and tissue creation with PDL.

thrombolysis can occur resulting in elevated D-dimer levels.[4] MRI is the best imaging modality and reveals hyperintense serpiginous lesions with phleboliths on T2-weighted images and possible fat or muscle interspersed between venous channels.[4] Easily compressible, spongelike networks of tubular structures with low velocity or no venous flow are demonstrated on ultrasound.[4] Lymphatic malformations present as soft, compressible cystic lesions filled with chylous material. They are classified as microcystic, macrocystic, or mixed. MRI varies depending on internal hemorrhage and inflammation, with high-signal T2-weighted images with gadolinium demonstrating mild peripheral enhancement without internal enhancement; ultrasound reveals no flow within major spaces.[4] Capillary vascular malformations (port-wine stains) present as flat lesions with a red to pink hue that may lighten during the first year of life, but eventually darken into a deeper red or blue lesion with thickening and nodularity.[10] These lesions are the most common type of malformations noted in 3/1000 births with an equal sex distribution and head and neck involvement in 80% of cases.[10] These lesions do not resolve spontaneously, grow proportionally with the patient, and are associated with syndromes such as Sturge-Weber (malformation distributed in the first trigeminal division, with possible V2 or V3 involvement, glaucoma, and central nervous system abnormalities) and Klippel-Trenaunay syndrome (malformation involving a unilateral lower extremity with associated hypertrophy, varicose veins, lymphedema and phleboliths).[10]

AVM comprise rare high-flow lesions with an unpredictable course, making treatment aesthetically challenging.[11] These lesions enlarge more readily because of higher inflowing pressures (absence of capillary transition between arterial and venous systems) and recruitment and collateralization, and as such, usually present as a pulsatile lesion with an associated bruit or murmur.[4] These lesions can lead to high-output cardiac failure in some cases.[4] The diagnosis should be made with MRI or computed tomographic angiography, and biopsy should be avoided due to the high risk of bleeding. MRI reveals prominent flow-related signal voids and allows for visualization of feeding and draining vessels, whereas Doppler ultrasound shows arterial flow within prominent high-flow draining vessels.[4] Histology exhibits beds of venules and arterioles intermixed with large-caliber arteries and veins.[4] The Schobinger scale can be used to classify the severity (**Table 1**).[4]

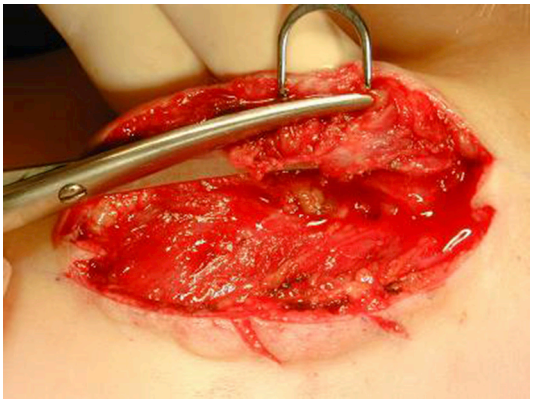

Fig. 8. Avoid overdeveloping the plane between skin and hemangioma to maintain a safe skin flap thickness. Residual hemangioma is left behind to ensure flap survival.

TREATMENT: VASCULAR MALFORMATIONS

Treatment varies depending on the type of lesion. Low-flow lesions can be treated effectively with sclerotherapy, PDL, and surgery in select lesions, whereas high-flow lesions are usually managed with a multimodality approach.

Percutaneous sclerotherapy can be used as a first-line agent for small low-flow vascular

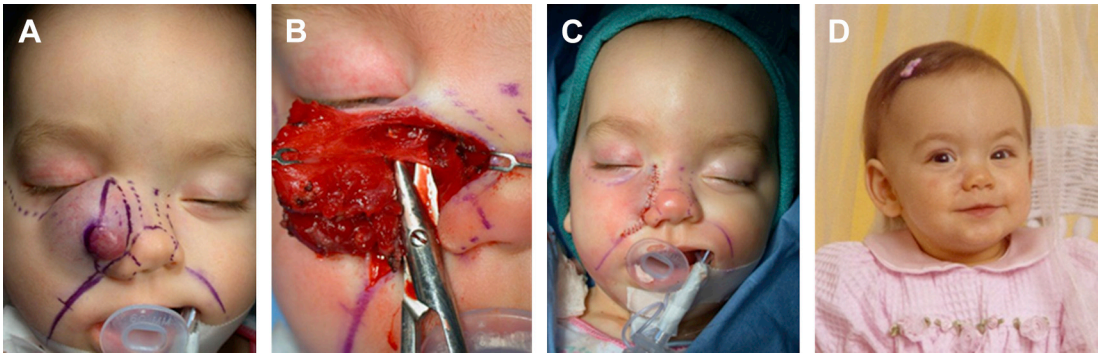

Fig. 9. (*A*) Preoperative photograph of the infant from **Figs. 1**C and **2** before surgical debulking during proliferation. (*B*) Surgical plane deep to the hemangioma is easily defined; rarely are deeper structures involved. (*C*, *D*) Immediate and delayed postoperative result.

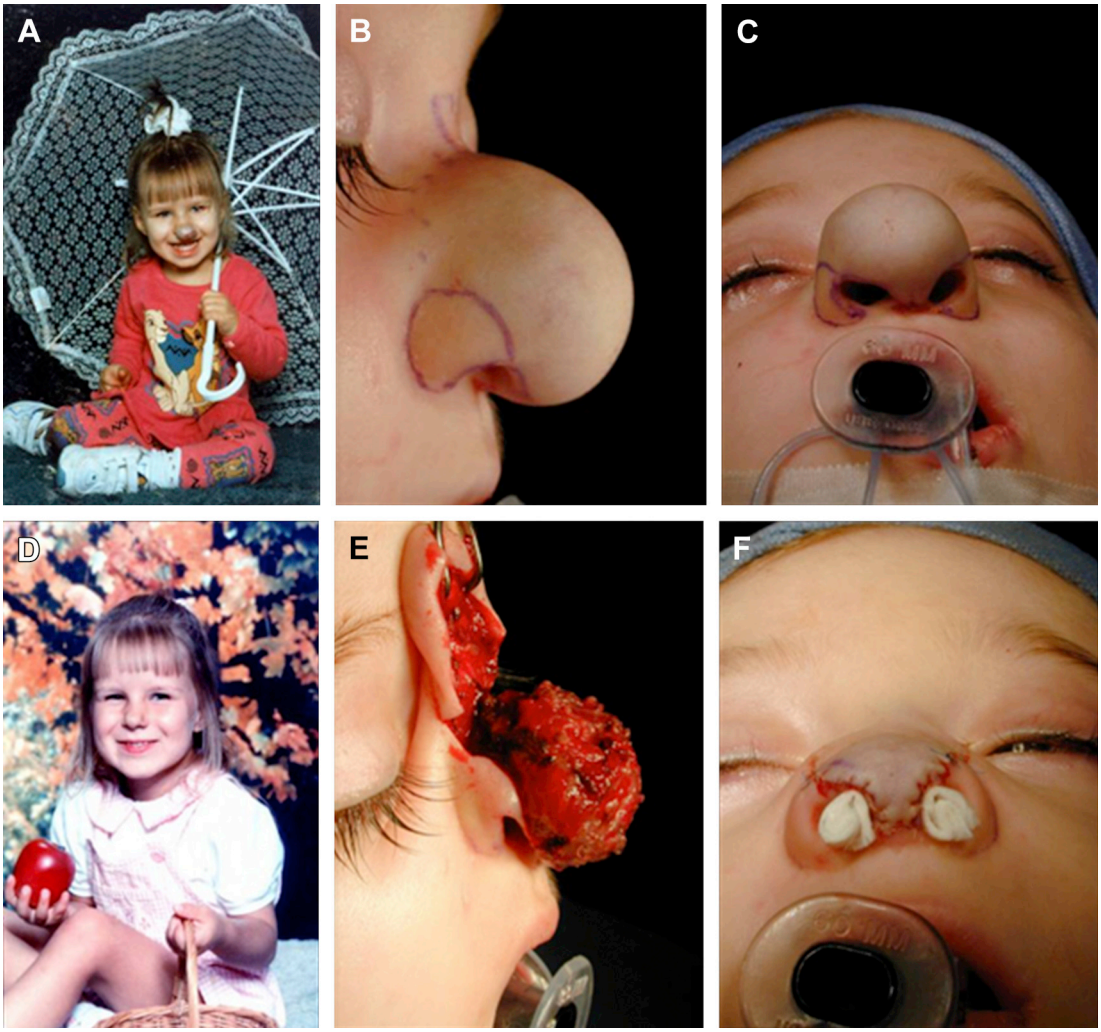

Fig. 10. External rhinoplasty approach to a deep hemangioma of the nose. (*A–C*) Preoperative views. (*D*, *F*) Postoperative views. (*E*) Intraoperative view with cutaneous flap elevated to demonstrate superolateral cutaneous extensions of marginal incision at the subunit junction.

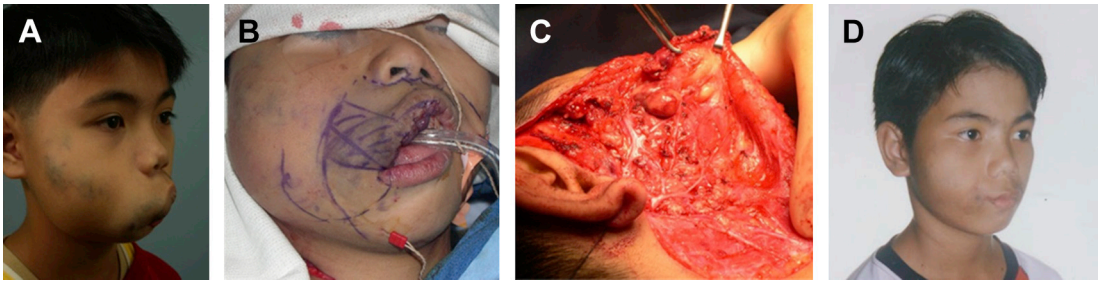

Fig. 11. Venous malformation resection. (*A*) Preoperative view. (*B*, *C*) Intraoperative views. (*D*) Postoperative view.

malformations or as a preoperative modality in an attempt to induce lesion regression before resection.[12–14] These treatments have been proposed to avoid significant morbidity and suboptimal cosmetic outcomes associated with surgical excision after failed conservative management. Sclerosants include ethanol, OK-432 (penicillin-killed *Streptococcus pyogenes*), sodium tetradecyl sulfate/sodium dodecyl sulfate (anionic surfactant), doxycycline, bleomycin, and gelatin adhesive, to name a few. They are injected in the malformation to induce endothelial damage, inflammation, thrombosis, and fibrosis and cause eventual destruction of the lesion.[12,13] Advantages include reduced risk of damage to nearby structures, such as nerves, absence of an incision, low risk of infections, and rapid recovery. Disadvantages include need for multiple treatments, limited application if lesions surround important structures, or ill effects from sclerosants that enter the systemic circulation.[12,13]

PDL treatment is particularly effective for low-flow capillary malformations or port-wine stains (**Fig. 12**). Before this, cosmetic camouflage and the argon laser were used to treat these lesions; however, the argon laser caused significant scarring. PDL has been established as the gold standard for treatment because it selectively targets the vascular chromophore, minimizing lesion-induced deformity with little morbidity.[10] Complications are rare but can include scarring, uneven pigment distribution, transient alopecia, cutaneous infections, and ocular injuries. It remains the authors' practice to treat no earlier than 6 months of age because this reduces the anesthesia risk with a plan for 3 to 5 treatment sessions approximately 6 weeks apart. The authors favor earlier laser therapy because mature lesions may be recalcitrant to laser intervention. It is important to educate the patient and family about the progressive, natural course of the disease, the need for repeated treatments, and the concept that port-wine stains cannot be completely eradicated, only lightened and flattened with the laser.[10] In fact, 50% of these lesions return to the initial appearance before treatment over time.[10] Preoperatively, outline the extent of the lesion before induction of anesthesia to ensure treatment of the lesion without treating the normal surrounding skin, because this can become more difficult to discern after anesthesia is administered. Ensure standard laser protocols for the patient (corneal shields or eye patches, surgical lubricant over hair-bearing structures to avoid damage) and staff

Table 1
Schobinger scale of severity of arteriovenous malformations

Stage	Stage Name	Description
I	Quiescence	Pink-bluish stain and warmth
II	Expansion	Enlarged swelling with pulsation, thrill, and bruit. Tense and tortuous veins
III	Destruction	Stage II with ulceration, bleeding, pain, and tissue necrosis
IV	Decompensation	Stage III with cardiac failure

Adapted from Tekes A, Koshy J, Kalayci TO, et al. S.E. Mitchell Vascular Anomalies Flow Chart (SEMVAFC): a visual pathway combining clinical and imaging findings for classification of soft-tissue vascular anomalies. Clin Radiol 2014;69:443–57; with permission.

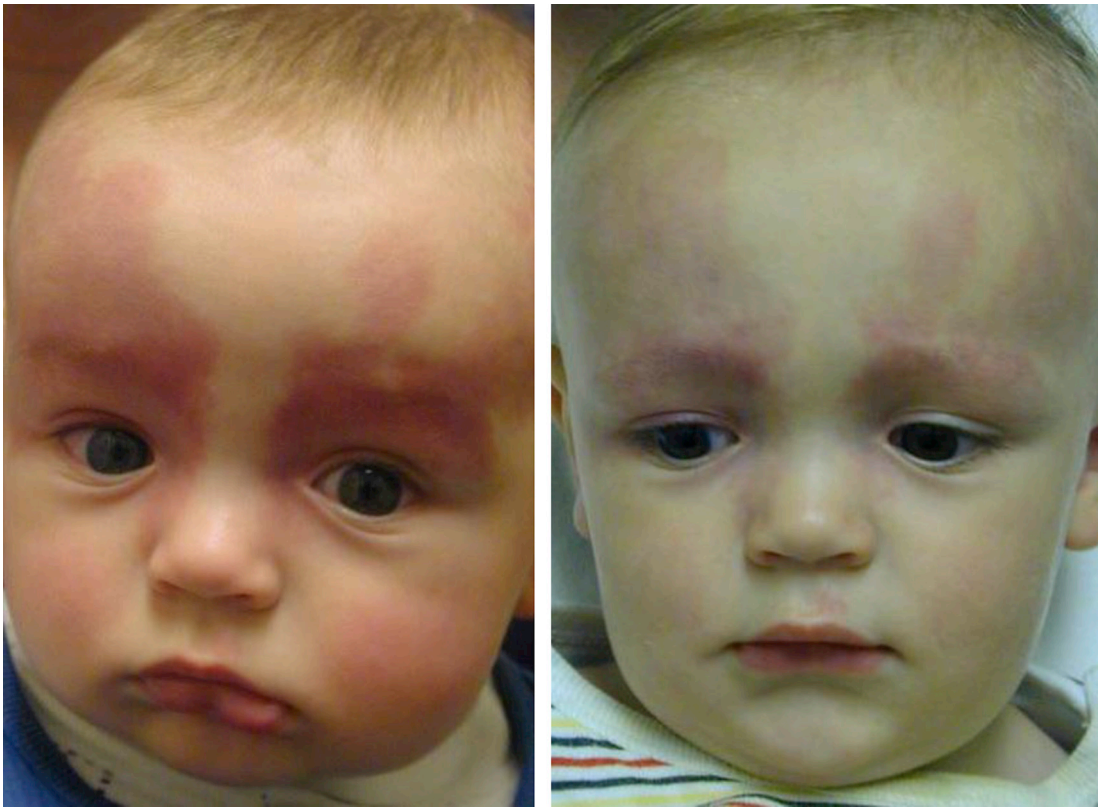

Fig. 12. Preoperative (6 months) and postoperative (1 year old) views of a child with bilateral V1 port-wine stains. There is noticeable lightening of the lesions after completing 3 laser treatments.

(safety glasses). By using a 5-mm hand piece with higher fluences (5–7 J/cm^2 for infants and patients with more sensitive skin, 8–12 J/cm^2 for patients >2 years of age), precise control can be attained at the periphery, whereas a 7-mm hand piece allows for a wider arc of coverage with lower fluences in the central areas. Target the perimeter of the lesion initially to disrupt the border of demarcation between normal and abnormal skin. Apply bacitracin as a dressing at the conclusion of treatment (**Fig. 13**).[10]

Treatment of high-flow lesions such as AVMs requires a multimodality therapeutic approach. Surgical resection is often the end result of management and is most effective with judicious resection and reconstruction using local and expanded tissue rearrangement. Preoperative embolization is highly recommended for complicated and extensive lesions that would otherwise be unsafe or surgically inoperable. The timing of surgical intervention is paramount as earlier intervention may minimize psychological distress and prevent growth of the lesion; however, older children are typically more tolerant of a potentially large operation. The focus surgically should be minimization of intraoperative hemorrhage via meticulous surgical principles with respect to vessel ligation and dissection. Once resected, the defect should be immediately reconstructed or allowed to heal by secondary intention if indicated (see **Fig. 13**).

SUMMARY

The ideal management strategy of vascular lesions rests on establishing the appropriate diagnosis, which historically was difficult until the advent of an effective classification system. Once the diagnosis has been secured, treatment of the lesion can follow a clear and usually successful algorithm. A multidisciplinary approach is paramount to successful management, and extensive surgical planning should be performed preoperatively if resection is anticipated. A conservative approach for the appropriate lesion has certainly reduced morbidity and optimized aesthetic outcomes. It is the surgeon's responsibility to exhaust all treatment strategies before considering operative management, unless early surgical intervention for established indications is necessary. With early

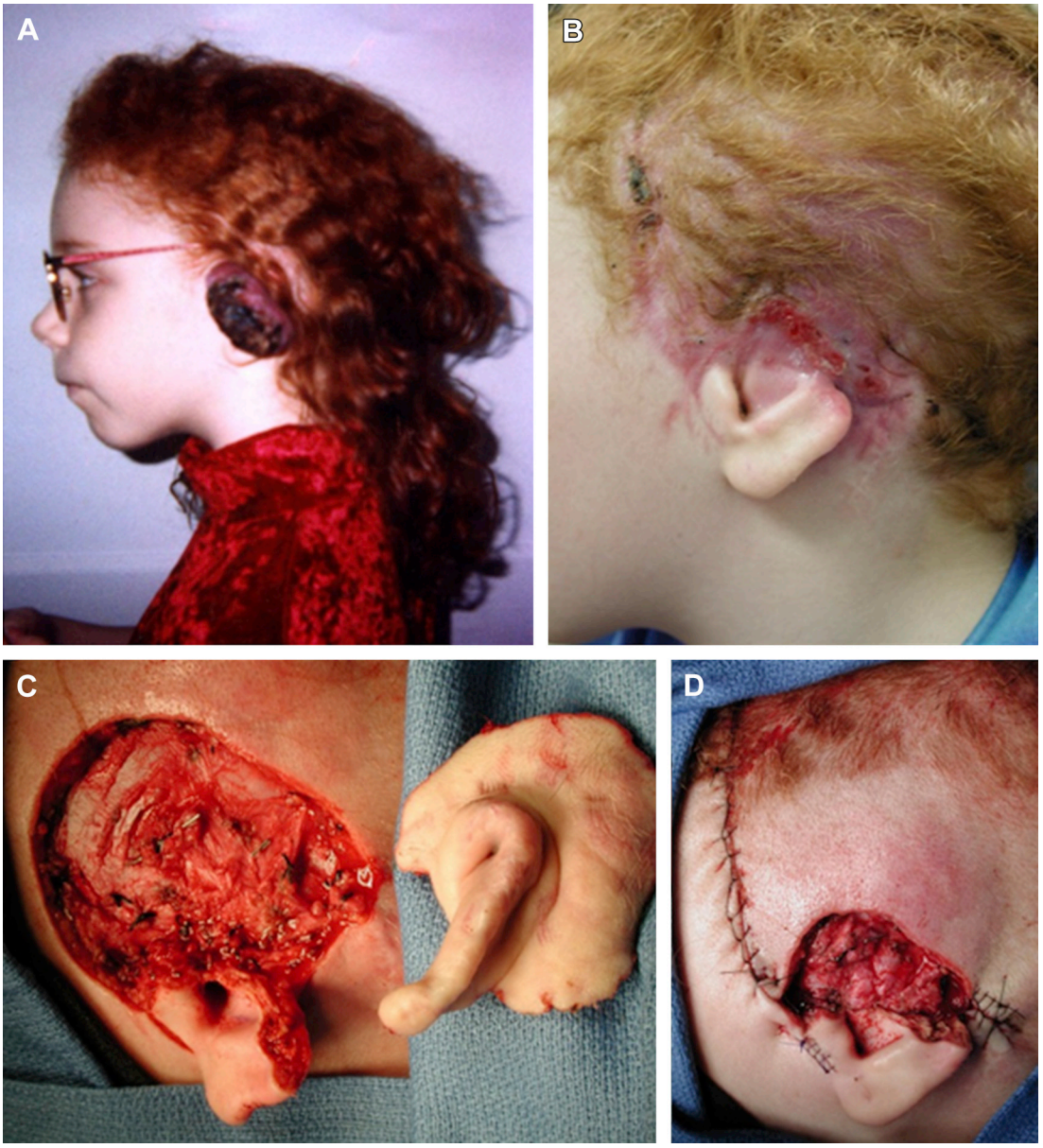

Fig. 13. (*A*) Preoperative view of left auricle AVM before the first round of embolization and surgical resection. (*B*) Postoperative view at 3 months after repeat surgical resection; complete closure of the wound by secondary intention occurred over a 3-week period. (*C*) Resected AVM lying adjacent to the surgical defect. (*D*) Surgical defect after rotation-advancement flap.

involvement of other specialists and the development of a comprehensive plan, safe and effective results can consistently be achieved.

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