Pediatric Facial Plastic and Reconstructive Surgery

James D Smith, Robert M Bumsted

Chapter 5: Vascular Lesions of the Head and Neck

James D Smith

Vascular lesions are the most common congenital abnormality seen in humans. One in three infants will be born with a vascular mark of some type. Lesions range from small, barely noticeable vascular birthmarks to large, deforming lesions that may be quite grotesque and even life-threatening. Many fade rapidly, but others may increase in size rapidly.

Birthmarks have played a part in folklore for thousands of years. The question is whether maternal thoughts, desires, or trauma can produce a mark in the unborn child. In antiquity it was felt that ungratified desires, such as craving a certain food, like a strawberry, could produce a similarly shaped birthmark in the unborn child, leading to the term *strawberry hemangioma.* The notion that mothers could imprint their babies carred into the medial field through the Renaissance and affected medical teaching into the late 19th century. There were a few brave souls who argued against the theory of maternal impression and concluded that birthmarks were the result of faulty embryological development and not maternal impressions. Unfortunately, there is still the lingering suspicion by some people that maternal impressions may occur, which is perhaps reinforced by parental convictions. With this lingering doubt, the moth of a newborn child presenting with a vascular malformation may have feelings of guilt that she has done something wrong during the pregnancy and thus marked her child for life.

Birthmarks have had an interesting effect on art and literature. Mulliken and Young found a paucity of portrait paintings showing vascular birthmarks. The only one found was a portrait by Picasso in 1906 that showed a woman with a lymphatic anomaly of the lower cervical area. In literature the vascular birthmark has been associated with tragedy in stories by Shakespeare, Hawthorne, and V S Pritchett. For some reason people seem to shrink away from someone so marked by nature. Even today Mikhail Gorbachev, who has a visible vascular birthmark on his forehead and scalp, had it airbrushed out by newspaper photographers in Russia. In the USA it is the political cartoonists' favorite trademark when depicting Mr Gorbachev. Other important people have learned to turn their head in photographs so their birthmark is not visible. Birthmarks today continue to be a source of embarrassment for those individuals affected by these vascular lesions.

Emotional Aspects

It is natural for all parents to expect a perfect, normal baby at the end of a 9-month pregnancy. When this expectation is not realized, there is an emotional impact on the parents. If the lesion is in the highly visible area of the head and neck, or is large, there will be a major challenge to the parents' self-esteem and ability to parent. Their response will have a critical effect on the child's emotional adjustment to the birthmark. Many times the parents' reaction may be overprotective and not allow the child to develop his/her own potentials. As in many other cases of serious illness or congenital deformities, the stress of a large vascular

birthmark on the family unit may be so great that the family will break up. One of the parents will become so obsessed with the affected child that the needs of the spouse and other children are neglected. This will increase the stress and guilt feelings on the affected child, who may feel it is his/her fault the family has split up.

Harrison has divided personality development in an affected child into six stages. In infancy, the reaction is mainly on the part of the family. There is guilt on the mother's part, who blames herself for what she has done or eaten, feeling that maternal impressions do occur despite assurances to the contrary. With hemangiomas, only 30% are visible at birth, but within a few months all hemangiomas rapidly expand and grow, creating a great deal of anxiety in the mind of the parents. They want something done "now" to restore their "normal" child. During this time, when a new baby is normally shown off, the parents are reluctant to dress up and show off their new child for fear of negative reactions from friends and strangers. This sense of shame will affect the parenting process.

The toddler stage is from 12 to 24 months of age. At this stage the child will first become aware of the vascular lesion as the child begins to develop his/her own self-awareness. At this age all children begin to fear separation from parents, clinging at bedtime and being shy with strangers. This is a normal developmental stage, but if parents do not understand this they may interpret this reactions as frustration about the birthmark and press for early removal. Helping parents understand that these are normal developmental conflicts rather than concerns about the birthmark will help them deal with this stage in a more relaxed manner.

In the preschool period the child starts to identify with the parent of the same gender, but there are also feelings of inferiority and inadequacy in relationship to the adult competition. Bodily defects, such as a vascular birthmark, may become a focus for feelings of inferiority and badness, and as a result the child will act sad or regress to babyish behavior. The child may vacillate between positive attachment to the birthmark and feelings of inferiority or badness because of the birthmark. As a consequence the child may vacillate between wishing for surgery to get rid of the birthmark and positive feelings about his/her body, which the birthmark is part of, and not wanting to have it off. At this point the surgeon may want to counsel the parents to delay surgery and seek counseling for the child.

In the school-age child there is the beginning of separation from home. In the school situation, the child must develop competence in the academic arena, in sports, and in making friends. The birthmark may get mixed up in these developmental conflicts. The child may believe that the birthmark is the reason he cannot make friends or is not doing well in school. For the parents it is also a difficult time to sort out how much is related to the birthmark and how much is a lack of competence in peer relationships.

In the first 1 to 2 years of school the child seems to be accepted, but by age 7 to 8 years teasing and isolation become more acute with significant stress on the child's psychological development.

The next stage is adolescence, when there is the normal struggle with sexuality and a need to become more independent. It is a normal time for high anxiety in relationships, a need for peer acceptance, and concern for personal appearance. The presence of a vascular

birthmark compounds all of these problems and feelings. Rejection by the opposite sex is felt to be secondary to the birthmark. The normal child-parent conflict is intensified with the child possibly feeling rejected by his/her parents or blaming them. The normal embarrassing incidents of adolescence are compounded and made worse by the physical mark.

As one approaches and enters adulthood some acceptance will take place, but still there is the acute feeling that people are staring or rejecting one because of one's appearance. It may result in withdrawal or isolation to protect oneself. With help, the person can overcome these feelings, but there is still the desire to be rid of this abnormality. This leads to the hope by all of these patients, no matter what their age, that there will be a new miracle treatment to rid them of this unwanted birthmark. They are often willing to try any new treatment and will be happy with minimal improvement that the physician feels is a poor result at best.

Classification

The classification of vascular lesions has been an area of confusion for centuries with descriptive, anatomico-pathological, and embryological classifications. It has led to a confused body of literature and nomenclature using terms such as hemangioma, cavernous and capillary hemangioma, port-wine stains, nevus flammeus, lymphangioma, cystic hygroma, and hamartoma, just to state a few. In 1982, Mulliken and Glowacki proposed a histological classification based on the correlation of cellular features of the vascular lesions of infancy and childhood with the physical examination and natural history. On the basis of cell kinetics they identified two major types of birthmarks: hemangiomas, those demonstrating endothelial hyperplasia; and malformations, those with normal endothelial turnover. Table 1 compares the older terminology for vascular birthmarks to that proposed by Mulliken and Young.

Table 1. *Translation from old terminology for vascular birthmarks into hemangioma or malformation*

Hemangioma	Old terminology	Malformation
Capillary	Capillary	
Strawberry	Strawberry	
	Port-wine	Capillary
Capillary-cavernous	Capillary-cavernous	
	Cavernous	Venous
	Venous	Venous
	Hemangio-lymphangioma	Lymphatic
	Lymphangioma	Lymphatic
	Arteriovenous	Arteriovenous

With this classification the term *hemangioma* would be reserved for those vascular lesions that are common in infancy, which have a rapid growth phase characterized by hypercellularity and endothelial multiplication. This proliferation may result in a large cell mass that will necessitate the dilatation and formation of new feeding and draining vascular channels. These channels on angiography will give the appearance of a high-flow vascular lesion. There are also a few hypercellular tumors of vascular origin in adults that meet the

definition of hemangioma.

The second category, vascular malformations, exhibit a normal rate of endothelial cell turnover throughout their natural history. They are by definition congenital lesions, present at birth, although they may not all be obvious at that time. Some may be a vascular malformation or abnormality of the vessel wall that presents in adolescence or adulthood as the result of progressive ectasia. These vascular malformations grow with the child but may have gradual or sudden expansion secondary to flow/pressure changes, collateral formation, or hormonal modulation. These vascular malformations may be subdivided into capillary, venous, arterial, arterial-venous fistulae, and lymphatic anomalies. The capillary, venous, and lymphatic, or a combination of these, are "low-flow" lesions and the arteriovenous lesions are "high-flow".

Table 2. *Characteristics of vascular birthmarks*

Hemangioma	Malformation
Clinical	
Usually nothing seen at birth, 30% as red macule	All present at birth; may not be evident
Rapid postnatal proliferatin and slow involution	Commensurate growth; may expand as a result of trauma, sepsis, hormonal modulation
Female:male 3:1	Female:male 1:1
Cellular	
Plump endothelium, increased turnover	Flat endothelium, slow turnover
Increased mast cells	Normal mast cell count
Multilaminated basement membrane	Normal thin basement mebrane
Capillary tubule formation in vitro	Poor endothelial growth in vitro
Hematological	
Primary platelet trapping: thrombocytopenia (Kassabach-Merritt syndrome)	Primary stasis (venous); localized consumptive coagulopathy
Radiological	
Angiographic findings: well-circumscribed, intense lobular-parenchymal staining with equatorial vessels	Angiographic findings: diffuse, no parenchyma Low-flow: phleboliths, ectatic channels High-flow: enlarged, tortuos arteries with arteriovenous shunting
Skeletal	
Infrequent "mass effect" on adjacent bone; hypertrophy rare	Low-flow: distortion, hypertrophy, or hypoplasia High-flow: destruction, distortion, or hypertrophy.

The appeal of this simplified classification is that it is based on an accurate history, physical examination, and follow-up, but does not require complicated diagnostic techniques or biopsies. Table 2 presents the characteristics distinguishing hemangiomas from vascular malformations during infancy and childhood.

Hemangioma

Natural History

Hemangiomas are the most common tumor of infancy. About one-third will be visible at birth as a small macular spot or rarely as a full-size hemangioma. They are more common in fair-skinned families and occur in a 3:1 ratio female to male. There does not appear to be a familial incidence. Abotu 60% occur in the head and neck region with 25% in the trunk area and 15% on the extremities.

Hemangiomas have two distinctive phases. During the first 1 to 6 months of life there is a rapid proliferation and expansion of the hemangioma. At this time there is a proliferation of endothelial cells that form syncytial masses with and without lumina. In the early stages this lesion will consist of solid masses of proliferating cells where lumen formation is difficult to appreciate, but later vascular spaces are not so compressed and capillary-sized lumina may be seen lined with plump endothelial cells. The proliferative stage is the hallmark of hemangiomas and the most distinctive feature in separating them from vascular malformations. During this phase if the lesion is in the superficial dermis, the skin will become raised with varying shades of crimson color and should be referred to as a superficial hemangioma. If the lesion involves the deep dermis and subcutaneous tissue, the skin over it will be smooth with normal color, but with a bluish hue from the underlying hemangioma and will frequently have dilated veins or telangiectatic vessels on the surface. This should be referred to as a deep hemangioma. As might be suspected many hemangiomas will present with involvement of both areas and as such will have clinical characteristics of both, but the histological pattern of proliferative endothelial cells is consistent throughout the tumor.

The involution phase begins at about 6 to 10 months of age and may be divided into an early and late stage. After the initial phase of rapid growth, which may last a few weeks to a few months, the hemangioma stabilizes and for a short period of time seems to grow at the same rate as the child. As involution begins to occur the color changes from a bright crimson to a more purple color. During this time there may still be some remaining areas of proliferation while involution is beginning in the center of the lesion. The surface begins to take on a grayish mottled appearance, especially near the center. The lesion begins to soften, the skin becomes wrinkled, bleeding and ulceration cease to be a problem, and the lesion no longer swells when the child cries. As involution proceeds it seems to spread in a centrifugal pattern, starting inthe center and proceeding to the edges. As involution is completed the long-term result is variable. In some the skin will be near normal with mild atrophy and a few telangiectatic vessels; in others there may be significant atrophy with the appearance of scarfring. In some there is almost an empty sac of atrophied wrinkled skin, whereas the remaining may have fibrofatty infiltration leaving a mass with varying degrees of abnormal skin covering the surface. Histologically, there is decreased endothelial cellularity and progressive deposition of perivascular fibrofatty tissue. The vascular channels decrease as fibrosis increases and the remaining vessels dilate, with flattened, more normal-appearing

endothelial cells. Most clinical studies would indicate that complete resolution occurs in over 50% of children by age 5, in over 70% by age 7, and continued improvement until puberty. Usually the earlier involution begins, the better the finall appearance will be.

Differential Diagnosis

The most important differential diagnosis is between hemangioma and vascular malformation. The clinical history is the most singly helpful distinguishing feature. Hemangiomas are not normally seen at birth, but within the first 1 to 2 months appear and rapidly grow at a rate beyond the child's growth. A vascular malformation will be present at birth and grow commensurately with the child. The early color of the hemangioma is usually bright crimson, and as involution proceeds it changes to dark purple, to a lilac, and ends as a yellowish to whitish color. Vascular malformation have a consistent color that is dependent on the depth of location and whether it is arterial, venous, capillary, or lymphatic. On palpation the hemangioma will have a firm or rubbery feel, whereas the venous malformations will be soft and compressible. In some circumstances the differential diagnosis will be more clear with time and the patient may need to be followed. In deep lesions, especially of the cheek or parotid area, a hemangioma is more likely to be confused with a lymphatic malformation, as both are rubbery to cystic on palpation and noncompressible. Hemorrhage into a lymphatic malformation may further confuse the picture. Both may rapidly expand under certain circumstances. In these circumstances I have found simple aspiration with a 20- or 22-gauge needle to be helpful in the differential diagnosis. If one gets clear, straw-colored fluid, the diagnosis of a lymphatic malformation is confirmed. Obviously pure blood would indicate a hemangioma or vascular malformation. Aspiration is relatively safe in this situation since pressure is easily applied to control bleeding. This is in contrast to potential hemangiomas of the viscera, especially the liver, where aspiration should never take place.

Another lesion that may occasionally be mistaken for a hemangioma is a pyogenic granuloma. It is a reactive proliferative lesion that histologically resembles a hemangioma and may be called a "capillary hemangioma" by the pathologist. These lesions appear suddenly, usually in children and occasionally in adults. They may occur on the cheek, eyelids, extremities, or on the mucosal surface of the oral mucosa or nose. They are usually granulomatous-type lesions that bleed easily and are pedunculated. They will occasionally occur ina capillary venous malformation (port-wine stain). These may be treated by removaland cauterization of the base with silver nitrate. If they recur, laser therapy or excision and closure may be necessary.

Radiographs are not usually needed for the differential diagnosis, but in deep lesions may be helpful. Computed tomography (CT) with contrast will help differentiate hemangiomas from vascular malformations. During the proliferative phase of hemangiomas the lesion will have a well-circumscribed homogeneous pattern, whereas during involution it will bemore heterogeneous with a lobular architecture. Venous malformations demonstrate a heterogeneous pattern with occasional calcifications. Lymphatic malformations show cystic elements with enhancement of the septa. The CT will also show the extent of the tumor, especially if resection is being contemplated.

Magnetic resonance imaging (MRI) scans are also helpful in distinguishing these lesions. Hemangiomas have high-flow characteristics with solid tissue of intermediate intensity

on T1-weighted images and appear bright on T2-weighted appearances of high flow with flow voids and lack of solid tissue, which helps differentiate them from a hemangioma. Venous malformations are characterized by a low-flow pattern that has an intermediate intensity on a T1 image slightly greater than muscle and are very bright on T2 images. With gadolinium the venous channels enhance. The MRI is especially helpful in identifying multiple venous malformations in muscle. Lymphatic malformations have low intensity compared to muscle on T1 images, and on T2 images are bright. They may exhibit either cystic spaces with varying signal intensities or multiseptated spaces. Combined lesions may share characteristics of both types of malformations. The MRI may obviate the necessity for arteriograms in most low-flow lesions.

Angiography is not usually indicated in head and neck hemangiomas unless for some reason surgery is being contemplated or there is some unusual circumstance. The characteristics of a hemangioma are a well-circumscribed mass and intense, persistent tissue staining in a lobular pattern. Some hemangiomas have high blood flow, which suggests arteriovenous shunting and may be confused with tissue shunting seen in artiovenous malformations.

Complications

Several complications may occur in hemangiomas, usually during the proliferative phase. The physician must be alert to these as some may produce long-term or permanent disabilities.

In rapidly growing superficial hemangiomas the epidermis may be lost and cause ulcerations and/or bleeding. The bleeding is usually venous oozing or more rapid bleeding from a small punctate area. Both of these are easily controlled with pressure, but may be frightening to the parents, especially when there is an abrasion and bleeding while the child is asleep, leaving blood over the bedclothes. In rare circumstances these ulcerated areas may become infected, producing a septicemia or extensive necrosis and slough of soft tissue.

Rapidly enlarging hemangiomas surrounding or in the orbit may permanently affect vision. A lesion, such as in the upper eyelid, which obstructs vision for even short periods of time, may result in permanent effects on vision, such as amblyopia or failure to develop binocular vision. Periorbital hemangiomas may place pressure on the growing cornea causing distortion, which may produce astigmatism and/or myopia. Strabismus may result from paralysis secondary to infiltration of the muscles or secondary to amblyopia. All infants with hemangiomas in the orbital or periorbital area should have ophthalmological consultation and close follow-up. In lesions that are affecting vision, a more aggressive treatment plan may be necessary.

Another life-threatening area of obstruction is the subglottic area. These infants will develop biphasic stridor in the first 1 to 2 months of life, in contrast to a congenital subglottic stenosis in which there will be stridor from birth. The symptoms rapidly become worse and are worse with crying. They will respond temporarily to racemic epinephrine or subcutaneous epinephrine. About 50% will exhibit a cutaneous hemangioma elsewhere on the body. The diagnosis may be suspected by the appearance of a subglottic swelling on x-ray. It is confirmed by direct laryngoscopy and bronchoscopy with the typical findings of a soft

compressible subglottic mass that may or may not have telangiectatic vessels or a bright red appearance. The mass is not usually circumferential, but comes from the posterior and/or lateral walls. In contrast the congenital subglottic stenosis is firm, not dilatable and circumferential.

Other potential areas of obstruction in the head and neck are the nose and external ear canal. Theoretically a hemangioma that obstructs both sides of the nose could produce airway problems in the infant, who is an obligate nasal breather. In practice, total obstruction is rare and when it does occur it is slow enough for the infant to adapt to mouth breathing. However, one must be aware that these infants have the potential for significant sleep apnea secondary to this obstruction, which in severe cases may interfere with feeding and result in failure to thrive or right heart failure. If a hemangioma obstructs the external auditory canal the child will have a conductive hearing loss. This is not a problem if the hemangioma involutes and the canal opens up, unless it is bilateral. If it is bilateral then some type of amplification should be used by 6 to 8 months or there will be a delay in the child's speech development.

Skeletal distortion is a concern, but is relatively rare in hemangiomas. It is much more likely to occur in vascular or lymphatic malformations.

Bleeding from a superficial ulcerated lesion is unusual and is easily controlled. Of much more concern is the rare condition of Kasabach-Merritt syndrome. This is a generalized clotting disorder that manifests itself with petechiae, ecchymosis, or internal bleeding in a child with a large hemangioma. There appears to be platelet trapping during the proliferative phase, which results in a profound thrombocytopenia. This is a potentially fatal condition from bleeding into the hemangioma with pressure on vital structures or internal bleeding into the viscera or central venous system. Infants with a large hemangioma (greater than 5 cm in diameter) are at a greater risk and should be evaluated with a complete blood count including a platelet count. If thrombocytopenia is present, coagulation studies including prothrombin time (PT), activated partial thromboplastin time (PTT), fibrinogen, fibrin degradation products, and fibrinopeptide A should be obtained to look for evidence of a consumptive coagulopathy. The other serious complication to be watchful for in the Kasabach-Merritt syndrome is infection and septicemia. If this is suspected, wound and blood cultures should be obtained.

A final possible complication is congestive heart failure. This is rare in hemangiomas isolated to the head and neck, but may occur if they are large or multiple or there are associated visceral hemangiomas. It is felt this is the result of high output failure secondary to multiple hemangiomas requiring large amounts of blood. The infant will usually present at 2 to 8 weeks of age with congestive heart failure, hepatomegaly, and anemia. There is a high mortality rate associated with this condition. It is treated medically and if this is unsuccessful, consideration must be given to embolization and/or surgical intervention.

Treatment

One can imagine the fright and concern that new parents experience as suddenly from nowhere or from a small innocuous red birthmark there is the appearance of a rapidly expanding crimson tumor that is disfiguring their newborn child. In rare cases a large portion of the head and neck may be involved. The frightened parents seek medical help to arrest the growth of the tumor and restore their child to normal. The concerned physician may make

recommendations that are detrimental to the child if he is unaware of the natural history of hemangiomas versus vascular malformations. Concern that this represents a highly malignant tumor may lead to injudicious or unnecessary surgery. Parents are concerned that everything be done and may pressure the physician into doing something. It makes a sympathetic and concerned physician who is willing to listen to the parents' concerns and reassure them that watchful waiting is usually the best treatment. Care must be taken to watch for potential complications such as eye involvement, congestive heart failure, or coagulopathies and treat these entities expeditiously.

Historical

For centuries multiple types of therapy have been attempted to rid the newborn of this bright red stain. In antiquity various methods of folklore were used, such as rubbing the spot with the afterbirth, having the mother lick it, etc. Physicians for centuries have attempted ligation and/or excision and continue to be the source of reports for treating large "hemangiomas". The observation that hemangiomas that ulcerate would go on to heal and involute led to the widespread use of artificial ulceration, using agents such as astringents, caustics, refrigerants, and various bacterial and viral inoculums to produce an infection and scarring. As galvanic electricity became available in the late 19th century, electrolysis and thermocautery were widely used and were the forerunners of today's more sophisticated laser technology. Sclerosing agents were used widely in the 19th and early 20th centuries, and continue to be used in various forms today. Radiation therapy was widely used in the 1930s to 1950s. The rapidly proliferating hemangioma is very sensitive even to small dosages of 300 to 600 R, but there are well-documented risks of associated malignancies years later. As a result, the consensus is that radiation therapy does not have a place in the treatment of hemangiomas today.

Compression therapy was first advanced in the early 1800s and has been more recently suggested for the treatment of hemangiomas involving the extremities.

Spontaneous involution received some notice, but it was Lister's report of a prospective study in 1938 that documented the natural history of 93 hemangiomas that grew rapidly in the first few months of life, then went on to regress and disappear by about 5 years of age. Since this significant observation of spontaneous involution, the literature is replete with numerous articles debating which lesions will and will not involute. Unfortunately, the confusion is enhanced by the inconsistent terminology combining clinical findings with pathological descriptions of capillary, cavernous, and capillary-cavernous hemangiomas. The classification described by Mulliken and Glowacki limiting the term *hemangioma* to those rapidly proliferating lesions in the infant, which are the ones that will most likely undergo spontaneous involution no matter how fast they grow or how large they become, leads to a rational treatment plan for these unfortunate children.

Contemporary Management

Observation

Watchful waiting as a tumor is enlarging on a daily basis is most difficult and alarming for both parents and physicians. The former want something done and the latter wish

to do something. Inactivity in the face of a rapidly progressive tumor is galling for a surgeon whose life is spent doing something to improve patients' well-being. For those surgeons attuned to the importance of appearance it is particularly difficult to stand by and watch a beautiful infant rapidly become distorted and disfigured by this devastating lesion while the anguished parents are pleading for something to be done. One must temper this natural inclination to do something with the knowledge that hemangiomas will involute and any treatment we recommend must guarantee a better or more rapid outcome than spontaneous involution. Our overriding motto should be "do no harm".

At the initial consultation the physician must be willing to take the time to listen to the parents and explain the pathogenesis and natural history of hemangiomas in layman's terminology. It is particularly helpful if he has personal or textbook photographs demonstrating the stages of spontaneous involution. Special care must be taken to reassure the mother that it is not her fault or some activity or eating pattern that produced this growth. It is necessary to assure the parents that this is a benign tumor and that the need for active intervention is unlikely. During this initial visit one must be sensitive to the desperate feelings of the parents and if one senses doubts, to offer a second opinion from a reliable and knowledgeable physician. Many parents will go from physician to physician until they find someone who is willing to actively treat their child's growing tumor. During this initial visit photographs and measurements should be taken to document changes in the future.

Finally, close follow-up to reassure the parents and watch for possible complications should be arranged. The parents should be encouraged to call if they have concerns. If there is concern for vision or airway problems, the appropriate consultations should be arranged. Depending on the size and location, return visit intervals may be every 1 to 3 months during the proliferative phase and as long as 6 to 12 months once the lesion has stabilized and involution is starting.

Local Complications

If complications occur, the patient will need to be seen on more frequent intervals. The most common local complication is during the proliferative phase when ulceration with or without bleeding may occur. The ulceration from epithelial breakdown in a superficial hemangioma is treated with local care including gentle cleaning, topical antibiotic ointment, and a nonadherent dressing changed at frequent intervals. This may be an avenue for cellulitis and septicemia, which should be treated with the appropriate systemic antibiotics. It may take several weeks for the area to reepithelialize.

Bleeding from an ulceration or punctate area is another troublesome complication. Parents are particularly concerned, since they feel bleeding may become serious and uncontrollable. The bleeding can be controlled by gentle pressure with a nonadherent pad for at least 10 min. If bleeding continues, the child should be brought to the office or emergency room where cautery with silver nitrate or electrocautery will solve the problem. I have found it useful to teach the parents how to control the bleeding with pressure and then use a silver nitrate stick to cauterize the bleeding point. In a short period of time this ceases to be a problem. Neither ulceration or bleeding are indications for surgical removal, but persistent local bleeding may be an indication of a systemic coagulopathy and should be evaluated.

Steroid Therapy

Steroids were first used to treat thrombocytopenia associated with hemangiomas, but it wasn't until 1963 that Zarem and Edgerton noted a coincidental shrinkage of the hemangioma as well. Reported response rates have ranged from 30% to 90%. The exact mechanism of action by steroids is not clear. It is clear that the rapidly proliferating hemangioma is much more responsive than the stable or involuting lesion. It may be that the steroids have a direct effect by causing vasoconstriction of the channels and sinusoids of the immature hemangioma or by somehow modulating the control of endothelial proliferation. At the present time there is a great deal of research going on regarding angiogenetic factors that may shed light on the mechanism of steroid action, as well as provide new medical agents that may be used to treat hemangiomas. Crum et al have noted that there may be more active inhibition of angiogenesis by steroids in the presence of heparin or heparin fragments.

Mulliken gives the following guidelines for the use of systemic steroids:

1. A rapidly growing lesion that seriously distorts facial features.

2. A lesion where there is recurrent bleeding, ulceration, or infection.

3. A lesion that interferes with essential, normal, physiological functions (breathing, vision, eating, or hearing).

4. Large or multicentric hemangiomas causing bleeding (secondary to thrombocytopenia) and/or high cardiac output failure.

He recommends oral prednisone at 2 to 3 mg/kg/day for 2 to 3 weeks. In 7 to 10 days some response should be noted with decrease rate of growth, color change, and/or softening of the lesion. If there is no effect, steroid therapy should be discontinued. If there is a response the dosage should be tapered and if possible switched to an every-other-day dosage. Pulse therapy of 4 to 6 weeks is used and then a rest period, depending on the response of the lesion. In some cases there may be a rebound phenomenon that will require further courses of prednisone. Direct intralesional injection of long-acting steroids has also been advocated by some authors to minimize the long-term effect seen with systemic steroids. The efficacy of both methods seems to be comparable depending on the lesion, location, and report. Intramuscular or intravenous Decadron has a rapid effect on some subglottic hemangiomas.

Side effects of short-term high-dose steroids are minimal, but there are temporary effects such as slowing of growth, decreased appetite, hypertension, facial edema, and increased risk of infection, especially for otitis media and pneumonia. Because of these potential complications, it is advisable to use steroid therapy in conjunction with a pediatrician or pediatric endocrinologist.

Chemotherapy

There are sporadic reports of the use of chemotherapy in the treatment of hemangiomas. Hurvitz et al reported success with using cyclophosphamide in hemangiomas

not responsive to other treatments.

Orbital and Periorbital

Hemangiomas of the upper eyelid may affect vision by producing astigmatismmm from pressure on the cornea itself or amblyopia. Large hemangiomas that occlude vision, even for short periods of time, may result in permanent visual loss. It is imperative that these infants be examined and followed by an ophthalmologist.

Rarely a small hemangioma may be surgically resected, but most of the time these lesions are full thickness in the lid and steroid therapy is the treatment of choice in the proliferative phase. Direct intralesional injection of a long-acting steroid, such as triamcinolone acetate or Depomedrol, has been used extensively. The technique is to use a 27-gauge needle with multiple punctures to spread the material throughout the lesion. A maximum of 40 mg of triamcinolone should be used at any one time. If the lesion extends into periorbital tissue there is a risk for hemorrhage or hematoma and risk to vision. In these situations systemic steroids are preferred. During treatment patching of the noninvolved eye may be necessary to minimize the development of amblyopia and strabismus.

Subglottic Hemangioma

Subglottic hemangiomas are a relatively rare cause of upper airway obstruction in infants. A child with cutaneous hemangiomas who presents with stridor and upper airway obstruction should be suspect for the presence of a subglottic hemangioma. The diagnosis is confirmed by direct laryngoscopy and bronchoscopy with the typical appearance of a bluish-red compressible mass or biopsy of the lesion. In an excellent review article, Shikhani et al reviewed all of the reported cases from 1913 to 1985 summarizing the myriad of treatments used in the past. Because of the acute airway obstruction, most patients have ended up with a tracheotomy. Treatments essentially mirror those used for the cutaneous lesions including sclerosing agents, cautery, open surgery, and observation. In the past, tracheotomy and observation were the recommended therapy. With involution by 2 to 3 years of age the child could be decannulated. Unfortunately, as pointed out by Shikhani et al, some series report over a 50% mortality rate secondary to complications of the tracheotomy. With better education of caregivers, better tracheotomy tubes, and in-home monitors, this figure has been significantly reduced, but there is still a risk plus the problems with wearing a tracheotomy tube and delayed speech development. Radiation therapy has the same risks as for cutaneous lesions and is not recommended. Open surgery in the infant larynx for subglottic hemangioma should also be condemned. Intralesions steroid injections have been used with and without tracheotomy. If they are used without tracheotomy the child must remain intubated for 5 to 10 days.

In the early 1970s the CO_2 laser began to be used for laryngeal lesions and the logical extension was to use it for subglottic lesions. The CO_2 laser works well, but there is the risk of cicatricial stenosis if the lesion is large or circumferential. Fortunately most subglottic hemangiomas involve posterior and one or both lateral walls. They should be treated segmentally, allowing for reepithelialization. No more than one-third of the circumference should be treated at one time. With recent experience using the neodymium:yttrium-garnet (Nd:YAG) laser for oral cavity and supraglottic submucosal hemangiomas, we have been

encouraged to try it on a subglottic hemangioma. A noncontact probe was used at 40 W and 0.3 sec. The patient was left intubated for 3 days to allow for swelling to decrease. In one case the results have been excellent, but obviously more cases are necessary to make a strong recommendation for the Nd:YAG laser over the CO_2 laser.

Congestive Heart Failure

Congestive heart failure is rare in children with hemangiomas limited to the head and neck. However, if a child presents with congestive heart failure and a head and neck hemangioma, care must be taken to look for other systemic hemangiomas, especially of the liver. If this is the case, steroid therapy is the first choice of therapy followed by angiography and embolization of the lesion if possible. If this is not successful or not indicated, surgical intervention may the only alternative.

Radiotherapy

Although radiotherapy is not recommended, it may be necessary in some desperate situations such as for hepatic hemangiomas.

Coagulopathy

The Kasabach-Merritt syndrome, which is characterized by thrombocytopenia, is usually a self-limiting condition, but still may be fatal. Many children may tolerate a low platelet count, but if they become symptomatic with a bleeding problem, treatment must be instituted. Steroids are the first line of therapy and some would recommend using it with heparin. Platelet infusions are only a temporary measure since the problem is platelet entrapment. In those who fail steroid therapy, Hurvitz et al have recommended cyclophosphamide treatment.

Surgical Therapy

An attractive idea is that early surgical removal of the hemangioma may prevent its rapid growth. The fallacy of this approach is that it may always remain small and the resultant scar may look worse than allowing the natural course of events to occur, or the transformation may be a field event beyond the bounds of simple excision from the very onset. As stated earlier surgical intervention is seldom necessary, and then one must weigh the potential surgical outcome against the natural history of the lesion.

Rarely is surgery indicated for head and neck hemangiomas during the proliferative phase, although reports continue to appear in the literature where this is being recommended. Significant complications, including skin slough and facial nerve paralysis, are reported. Deep hemangiomas of the parotid, which expand rapidly, may create the concern that this represents a malignancy. Batsakis has pointed out that malignant tumors of the parotid gland are medical curiosities, but that hemangiomas that undergo involution make up the majority of tumors of the parotid gland in infants. Rapid growth is not an indication for surgery.

Early childhood, ie, before the child enters school, may be an opportune time to do resections for redundant skin or fibrofatty tissue residuals after involution has taken place.

Care must be taken to make these resections in relaxed skin tension lines to minimize scarring. Hemangiomas of the tip of the nose, sometimes called the "Pinocchio nose", are particularly obvious and will benefit from surgical intervention. These may be approached from an external rhinoplasty incision or a direct excisional approach.

Final scar revision, removal of excess fibrofatty tissue, and cosmetic touch-ups should be left until complete involution takes place. Ideally one would prefer to do this before puberty when hypertrophic scarring is more of a problem. General plastic and reconstructive principles and techniques must be used to obtain the best functional and cosmetic results. Abnormally pigmented or textured skin should be removed in serial excisions or in some areas by tissue expansion techniques for skin coverage of excised areas.

Laser Therapy

Various lasers have been used in the treatment of vascular lesions for nearly 20 years. The argon laser with a wavelength of 480 nm is preferntially absrobed by red cells. It has been useful in treating port-wine stains, but does not penetrate deep enough to affect a proliferating hemangioma. Other potential lasers are the copper vapor laser and the flashlamp pumped dye laser. Each has their own specific characteristics and place of use. Waner and Suen have advocated the use of the copper vapor or flashlamp pumped dye laser in treating the early proliferative phase of superficial hemangiomas. They point out the low scar rate, less than 1%, and the effectiveness of controlling lesions in the small numbers they have treated so far. Obviously more cases and longer follow-up are necessary to see if this treatment should be used in all cases.

The Nd:YAG laser has been successful in treating a variety of vascular malformations. It has a wavelength of 1.050 nm with the ability to penetrate up to 1 cm, but does produce skin changes and scarring. It may have a place in the treatment of bleeding or ulcerated lesions where skin changes are inevitable. However, where the epithelium is intact, the scarring left by the laser may be worse than the skin left by involution. I have used it in a few situations where it was obvious there was redundant skin that would later have to be excised in the hopes of hastening involution and decrease bleeding.

Vascular Malformations

Pathogenesis

The peripheral vascular system arises in situ from the primitive mesenchyme cells, but they are surrounded by perocytes and smooth muscle cells that arise from neuroectoderm. There is a complex development that takes place in three stages.

First, there is a period of an undifferential capillary network composed of interconnected blood lakes, but with no identifiable arterial or venous channels. At about 48 days in the human embryo, separate venous and arterial stems appear on either side of the capillary system in what is called the retiform stage. The final stage occurs by 2 months of life and involves the gradual replacement of these immature networks by mature vascular channels.

It is less clear how the lymphatic system develops. The most widely accepted theory is the "centrifugal theory", which postulates that the lymphatic system arises from a process of budding from the embryonic venous system to form five lymphatic sacs: two jugular, two posterior, and one retroperitoneal. Endothelial sprouting from these sacs form the mature peripheral lymphatic system at about the 9th weel of development.

Many theories have been advocated as a cause of vascular malformations. There is some evidence based on the patterns of port-wine stains, hydrosis over malformations, etc, that there is a defect in the autonomic nervous system through the perivascular cells, which have contributed to vascular malformations. Multifactorial aberrations of normal development may also cause vascular malformations. Sequestered or maldeveloped areas of primitive capillary sets could result in venous or combined lymphatic and venous malformations. Failure of regression of arteriovenous communicators in the primitive rete could produce arteriovenous malformations. Abnormal dynamics in the developing system can also explain the particular morphology of some malformations. In lymphatic malformations the deep lymphatics are normal, but there is a failure of the cutaneous lymphatics to communicate with them, so it would appear there is a maldevelopment or sequestration of parts of the developing lymphatic system that form the lymphatic malformations.

In evaluating vascular birthmarks, the first step is to separate true hemangiomas from vascular malformations. At this point vascular malformations may be divided into "low-flow" and "high-flow" malformations. "Low-flow" anomalies may be further divided into capillary, lymphatic, and venous lesions; "high-low" anomalies are arteriovenous malformations that may have microscopic or macroscopic shunts. Each of these categories will be discussed separately.

Clinical Evaluation

Vascular malformations are much less common than other vascular birthmarks such as hemangiomas and vascular stains. Pratt examined 1.096 neonates and found a 1% incidence of hemangiomas, 10% incidence of capillary marks on the eyelids, forehead and nose, and a 42% incidence of macular birthmarks (nevus flammeus nuchae) in Caucasian infants. There were only 5 out of the 1.096 who had a true vascular malformation. Although vascular malformations are relatively rare, they persist throughout life, whereas the nuchae stains and capillary marks fade and hemangiomas involute.

At the initial evaluation it is sometimes difficult to distinguish hemangiomas from vascular malformations. Malformations will grow with the patient and are histologically stable. They do not regress, but under certain circumstances will enlarge, extend, or cause future problems, such as arteriovenous fistulas, and a few are progressive malformations. Some will enlarge secondary to hormonal stimulation such as pregnancy or taking birth control pills. Arteriovenous fistulas may not manifest themselves until adulthood. With this variability in mind, the patient must be followed to evaluate the clinical course of the lesion to make a better prognosis. In most circumstances if the lesions are stable, not affecting function or a significant cosmetic problem, a wait-and-see attitude is indicated. If this approach is to be taken, it is important that the patient be evaluated for other potential involvement such as the viscera, extremities, central nervous system, cardiovascular system, hemostatic abnormalities, and, in older patients, the possibility of a malignant lesion. It is not

within the scope of this chapter on vascular lesions of the head and neck to cover all the described syndromes and lesions of vascular etiology in detail as this information is available in textbooks devoted to the subject, such as the excellent work by Mulliken and Young.

Investigative studies are not normally necessary for vascular lesions of the head and neck. Clinical examination and longitudinal follow-up are adequate for the majority of these lesions unless there is a clear indication that management of the condition will be influenced by the studies. Our desire to make an academic diagnosis should not be an indication for expensive or unnecessary investigations. On the other hand, if active treatment is necessary, thorough investigation is mandatory before any intervention is carried out.

First of all, one must determine if there are any hematological abnormalities. A history of bleeding, recurrent thrombosis, or persistent anemia should raise suspicion that there is an abnormality associated with the lesion. As previously discussed, with the profound thrombocytopenia seen with some hemangiomas, a less severe localized consumptive coagulopathy with normal or near-normal platelet count may occur in some vascular malformations. These patients may be stable, but under the stress of surgery the coagulopathy may become clinically significant with the possibility of localized and systemic bleeding. Personally, I have seen this occur in two patients with lymphatic malformations. These patients should be evaluated with a complete blood count, including a peripheral blood smear and clotting studies including bleeding, prothrombin and partial thromboplastin time, fibrinogen levels, fibrin degradation products, and fibrinopeptide levels. These studies should be done preoperatively as well as postoperatively.

The mainstay of radiological evaluation for vascular lesions has been the angiogram, but with newer modalities such as digital subtraction angiography (DSA), computed tomography (CT), and magnetic resonance imaging (MRI), more useful information is obtained. Although plain x-rays may be useful in evaluating skeletal structures of the extremities involved with vascular lesions, they are seldom indicated in the head and neck. CT with and without contrast is much more useful in evaluating the skeletal effects of a vascular malformation of the head and neck. Not only is good skeletal detail seen, but spatial relationships with location and size of the lesion may be visualized and some idea of its vascularity may be determined if contrast is used.

Although MRI does not give good bony detail, soft tissue detail seen in a three-dimensional plane is excellent. With its highly sensitive densitometry characteristics, it is useful in distinguishing tissue planes, fat, mucus, fluid-filled cysts, lymphatic and venous malformations, as well as giving some indication of whether a lesion is high flor or low flow. Combined, CT and MRI give very useful information if surgery is contemplated or as a noninvasive technique to follow the growth characteristics of deep lesions. They may replace the necessity of an angiogram in some, but not all, situations, especially high-flow lesions. They may be useful in directing the angiographer.

Angiography is a more invasive technique with higher risk, but with newer contrast agents and improved catheters and techniques it has a very acceptable morbidity rate. The angiogram will give the physician vital information as to the extent of the lesion, the feeding and draining vessels, and whether embolization, thrombosis, or occlusion are viable treatment options. It will help distinguish if a lesion is high flow or low flow and will differentiate an

arteriovenous (AV) malformation from a venous malformation.

Capillary Malformations (Port-Wine Stains)

Vascular birthmarks with a typical vivid reddish stain of the skin have been referred to as port-wine stains for centuries. In antiquity it appeared as if a deep red wine had been spilled on the patient's skin; hence the name. The Latin term for these vascular lesions is *nevus flammeus.* In the terminology used for this chapter taken from Mulliken and Young, these birthmarks would be classified as capillary malformations. However, the term *port-wine stain* is so ingrained in medical literature that we will bow to this usage. The term *capillary hemangioma* is confusing and should be abandoned as it is frequently used to describe hemangiomas discussed earlier. Capillary malformations or port-wine stains are not proliferative lesions and thus should not be called hemangiomas.

Port-wine stains are present at birth and remain throughout life without involution. The skin discoloration is usually evident at birth or shortly thereafter. There is equal sex distribution and the incidence is approximately 0.3% of all births.

Histologically these lesions are characterized by ectatic capillary to venular-sized channels located in the papillary and upper reticular dermis. The walls are thin and lined by flat, normal-appearing endothelium with normal cell turnover.

The port-wine stain is macular and sharply demarcated. It grows in proportion to the growth of the child. If the lesions are near mucous mebranes there may be involvement of the underlying mucosa as well. Most patients will describe color changes with emotion, internal and external temperature changes, and exercise. The color ranges from a pale pink to a deep red. As the child matures, the pale pink color will deepen to a darker red in early adulthood, to a deeper purple in middle age. As the patient ages, the surface becomes more irregular and corrugated with multiple nodular lesions of ectatic vessels or venous lakes.

Port-wine stains are associated with many other malformation syndromes, especially on the trunk or extremities. In the head and neck area one may see other underlying developmental defects such as a meningoencephale or arteriovenous fistula.

The Sturge-Weber syndrome is one of the more common and feared vascular malformation complexes associated with port-wine staining. It is characterized by vascular anomalies of the upper face dermis (port-wine stain), and choroid and ipsilateral leptomeninges. The dermal lesion will always involve the V1 dermatome, but may extend to V2 and V3 as well. In many cases there will be hypertrophy of the underlying connective tissue and skeleton, giving the patient an asymmetrical, sometimes grotesque appearance that is progressive through puberty. Focal or generalized seizures are common presenting symptoms of neurological involvement. Some of these patients will have varying degrees of mental retardation. In rare circumstances there may be other neurological findings of hemiparesis, hemisensory disturbances, and growth disturbances of the contralateral extremities.

CT examinations will show intracranial calcifications, sometimes even in infancy. Arteriograms will show capillary and venous anomalies of the leptomeninges. Other

anomalous abnormalities may be seen and may be responsible for the progressive degeneration and atrophy of the involved cerebral hemispheres seen in some patients.

Patients with port-wine stains of both V1 and V2 are at risk for the development of glaucoma. Those infants suspected of having Sturge-Weber syndrome or port-wine stains surrounding the orbit should have ophthalmological evaluations for glaucoma early, and then on a regular basis at 6- to 12-month intervals. The glaucoma may be refractory to medical therapy and require surgical treatment.

The history of treatments for port-wine stains is similar to the range of treatments for hemangiomas, both being equally ineffective with potentially serious side effects. Scarification by various methods has been one of the more frequently described treatments. Various methods of cutting, puncturing with needles, electrocautery, ultraviolet light, various forms of radioactive materials and radiation therapy, cryotherapy and even sandpaper, have been described. If effective at all, these treatments traded scarring for the stain. Tattooing has been used for many years, but the results are inconsistent, leaving a blotchy papular surface with an unnatural, fixed appearance that does not blend well at the edges, and in children may result in hypertrophic scarring.

One of the most acceptable forms of therapy without side effects is cosmetic camouflage. (The two most popular products are Covermark and Dermablend, which may be purchased at many large department store cosmetic counters.) It is a sophisticated blend of opaque, waterproof cream that is covered by an appropriate shading cream. The biggest disadvantage is the time it takes to apply (20 to 30 min daily), and the cost. Teenage girls and adult women are more likely to make the commitment of time necessary to use this method. I have had personal experience with one 10-year-old boy who seemed to be well adjusted to his port-wine stain, but after his mother used some makeup to cover this he became much more outgoing and self-assured.

Excision is another alternative for small areas or in facial aesthetic units. If primary closure is possible, a linear scar in relaxed skin tension lines may be an excellent alternative. If larger areas are involved, serial excision, skin expansion techniques, or full- or split-thickness skin grafts may be used to resurface the area. The disadvantage of skin grafts are their unpredictable pigmentation, scar hypertrophy at the edges, recurrence in the graft or at its edges, and abnormal skin texture. For these reasons skin grafts are usually discouraged. However, some patients are willing to trade these deficiencies for the bright red or purple port-wine stain they have lived with all their lives.

Laser treatment for port-wine stains was first reported by Goldman and Rockwell in 1973. In the 1970s the argon laser became the laser of choice for treating port-wine stains. It has the advantage of emitting light in the blue-green spectrum that is preferentially absorbed by red hemoglobin and melanin and penetrates only 1.0 to 1.5 mm beneath the surface of the skin. As the light energy penetrates the skin, it is changed to heat-producing thrombosis with collapse of the vessels and damage to the dermal collagen. The adnexal elements of the skin, sweat glands, and hair follicles survive, whereas there is superficial necrosis of the epidermis and dermis. The end result is a controlled dermal scar with epidermal regeneration from the adnexal elements and a decrease in the size and number of blood vessels, which leaves the skin smoother and lighter in color.

Side effects of the argon laser treatment are atrophic and hypertrophic scarring. The former is an acceptable complication and to a certain degree is the expected result from the dermal scar produced. Hypertrophic scarring is much less acceptable as it creates a raised, irregular surface that is cosmetically unacceptable and may actually be physically uncomfortable from itching and irritation. It is more common in children under 12 years, and in all ages in the nasolabial fold and upper lip. Unfortunately, in children in whom physicians would like to use the argon laser, the results are poorest, with either little or no response or hypertrophic scarring.

The technique for the argon laser is to do a test patch 1.0 to 1.5 cm square at the edge of the lesion in the most inconspicuous spot or in an area that may be excised with a good cosmetic result if there is hypertrophic scarring. If there is a good result the lesion may be treated in stages. The area to be treated is anesthetized with 1% Xylocaine without epinephrine. One characteristic is that the anesthetic very rapidly absorbs, so that only an area that can be treated in 5 to 10 min is anesthetized at one time. Each session will last 1 to 1.5 hr, depending on the area to be treated. We have tried narrow stripes, wide stripes, and checkerboard patterns, but have ended up choosing to treat anatomic units, which seems to give the best cosmetic result. Reepithelialization comes from the remaining adnexal units, not from the edges of the treated areas. The problem with the geographic patterns was the difficulty of blending the egdes between the treatment areas. They tended to have hyperpigmentation at the edges that left a visible pattern result. Following treatment, the skin is covered with an antibiotic ointment daily until the surface reepithelializes, which usually takes 7 to 14 days. The patient should avoid direct sunlight as much as possible, and should use a good sunscreen for 6 to 8 months. The final result will be visible in 3 to 4 months, although further lightening may take place over the next 12 to 18 months. The expected result is a lightening of the color, and in the those with thickened or an irregular skin surface, a significant smoothing. Although many times the results are mediocre in the eyes of the surgeon, most patients are happy with any improvement, especially women who use cosmetics for camouflage. If there is significant lightening of the lesion they can use less makeup with a better result.

Other lasers have been used to treat port-wine stains. The CO_2 laser vaporizes the lesion including the epidermis, dermis, and ectatic vessels. This is a nonspecific burn and is essentially the same as using dermabrasion. Scarring is significant. In general, it is not a recommended treatment.

The most recent laser to be used is the tunable dye flashlamp laser. It is tuned at 576 nm, which should be the ideal wavelength for maximum absorption by the ectatic vessels. In clinical experience it seems to work better on children, patients with light port-wine stains, patients who might scar easily, and patients who have not responded well to the argon laser. It does seem to require more treatments than the argon laser, but if the overall results are better with less scarring and a better textured skin, it will be worth it.

Laser therapy seems to be the best treatment available for port-wine stains today, but newer lasers, especially the copper vapor laser, and refined techniques hold promise that our results will continue to improve.

Telangiectatic Stains

Nevus flammeus neonatorum is a very common entity that may be confused with the uncommon port-wine stain. It has several synonyms including stork bite, salmon patch, and angel's kiss. These lesions occur most commonly on the forehead or nape of the neck and are pink, irregular, macular spots that blanch with pressure. The majority fade during the first year of life, but some may persist into adolescence and even into adulthood. Rarely there may be a familial incidence.

There are several other syndromes that include capillary-lymphatic malformations, hyperkeratotic vascular stains, angiokeratomas, and congenital telangiectasis, each with their own characteristics. Again, for a more complete description of these entities one is referred to Mulliken and Young's text on vascular birthmarks.

A final telangiectatic condition is the unusual familial syndrome of Rendu-Osler-Weber or hereditary hemorrhagic telangiectasia. It is inherited in an autosomal dominant pattern with an incidence of 1 to 2 per 100.0000. These lesions are characterized by discrete, bright red maculopapules, 1 to 4 mm in diameter, that appear on the mucosal surfaces of the nose, lips, and oral cavity, as well as on the face, palmar surfaces of the hands, nailbeds, and conjunctiva. They are also found on the mucosal surfaces of the gastrointestinal tract, urinary tract, tracheobronchial tree, vagina, and the parenchyma of the liver and central nervous system. The lesions may occur in childhood, but more commonly appear after puberty in the third and fourth decade. The condition is usually progressive with age and the patient's symptoms become more difficult to control. The vascular papules tend to ulcerate and bleed, leading to recurrent and difficult to control epistaxis, hemoptysis, hematuria, and melena. There is a family history in 50% to 70% of patients. However, many patients are unaware of the inherited syndrome, but just assume it was a family characteristic to have frequent nosebleeds or other bleeding problems. Severe complications may occur if the patient develops pulmonary or hepatic arteriovenous fistulae with high-output cardiac failure. Bleeding into the brain or spinal cord will produce neurological symptoms.

The most troublesome problem for most patients is epistaxis. It may occur spontaneously at socially awkward times such as during meals, with exercise, during sexual intercourse, and with emotional stress. As the patient becomes older and the bleeding becomes more severe, the patient may become incapacitated from anemia, cardiac failure, and the necessity for frequent hospitalizations for treatment and blood transfusions. Frequent blood transfusions, if necessary, increase the patient's risk to hepatitis and AIDS.

Treatment of the epistaxis starts with simple measures of pressure, superficial cautery with silver nitrate or electrocautery, packing with Gelfoam and thrombin, or various hemostatic agents such as Avitine or Instat. One should avoid gauze packing as it produces more bleeding when it is removed. Care must be taken with overzealous cautery as it may produce septal perforations. As these methods fail, one can progress to the use of the Nd:YAG laser, which is quite effective on the visible lesions, but as new ones appear it will need to be repeated. Systemic estrogen or estrogen creams have been recommended, but there is conflicting information as to their efficacy. If the lesions are surgically accessible through an alaotomy, one may remove the mucosa of the septum and floor of the nose, leaving perichondrium, and cover it with a split-thickness skin graft. The problem is that the lesions

may recur through the graft or at its margins, and there will be crusting and sometimes and odor associated with the skin graft.

Vascular spiders are small lesions with a central arteriole that has superficial vessels radiating from it. With pressure on the lesions using a glass slide, one may see central pulsations, and, as it is released, a centrifugal rush of blood to the edges. These lesions may appear on the face, arms, hands, and fingers. They occur in children and/or adults as well as temporarily during pregnancy. Because they appear during pregnancy and in patients with liver failure, there is evidence that these are hormonally modulated by estrogen. These lesions may be treated with the argon laser or a needlepoint cautery to destroy the central arteriole, although recurrences are not unusual.

Venous Malformations

Venous malformations are developmental abnormalities of veins that are dysmorphic in configuration and structure. At times they may be combined with capillary or lymphatic malformations. They may be further classified as localized, diffuse, deep, or superficial.

Venous malformations may present as skin or mucosal varicosities, spongy masses, or as diffuse lesions spreading through multiple tissue planes. If the lesion is deep, the overlying skin may be normal or have a bluish hue. More superficial lesions will have a deep purple color.

On a physical examination the lesions are soft and compressible, but nonpulsatile. After compression they refill slowly. Patients note that with straining or in a dependent position they will fill and this may be associated with a vague feeling of pressure or discomfort. They will grow in proportion with the child, but may enlarge rapidly following trauma or coincident with puberty or pregnancy. Occasionally, after direct trauma, they may develop arteriovenous shunting and develop into an arteriovenous fistula. In older lesions phleboliths from thrombosis may be palpable or visible on x-rays.

Histologically these lesions will have large venous spaces (formally referred to as cavernous hemangiomas) lined with flat, normal, epithelial cells and sometimes evidence of recent and old thrombi.

Venous malformations may involve muscles, especially the masseter muscle. If they are deep they may be mistaken for unilateral masseter muscle hypertrophy, even on a CT scan. MRI is particularly useful in separating out differences in these soft tissues and will help make the diagnosis. There is also a rare infiltrating intramuscular vascular lesion that has been called an intramuscular hemangioma or angiolipoma. It should not be confused with venous malformations.

Venous malformations of the facial bones are rare, but they may cause skeletal hypertrophy and bleeding around teeth. X-rays show a radiolucent defect with a honeycomb appearance.

Therapy for venous malformations has suffered from the same plethora of modalities as other vascular lesions. These again include irradiation, electrocoagulation, cryotherapy,

intravascular magnesium, and various sclerosants.

Before embarking on therapy, it is important to determine if the lesion is in the low-flow category or the high-flow. If there is any question, angiography should be carried out. The anatomical boundaries must be determined, either by MRI or angiography. One must determine if there is any localized intravascular coagulopathy that could lead to devastating bleeding at the time of resection. This must be evaluated as suggested previously with coagulation studies looking not only at the PT and PTT functions, but also studies for a consumptive coagulopathy.

If possible, total excision is the treatment of choice, but this is possible only in small to medium-sized lesions that are well demarcated. Unfortunately, surgery may be limited to partial resection or contour resection only. One of the frustrating points in surgical excision is that as the lesion is dissected and the blood supply is interrupted, the whole area collapses to a small amount of tissue and the edges are so ill-defined that it is impossible to tell if total excision has taken place.

Subtotal resection may lead to postoperative expansion of the remaining lesions, but it is worth the risk if there is significant improvement in contour, cosmetic abnormalities, or feelings of pressure and discomfort. In some cases removal of skeletal lesions may be indicated.

Selective angiography and embolization has been used, but runs the risk of necrosis of the adjacent soft tissue and overlying skin as reported by Demuth et al. Persky has reported a sclerosing technique for direct injection of contrast media and 95% ethyl alcohol into low-flow venous malformations with good results. Multiple treatments could be performed, and surgical resection of residuals was carried out in some cases.

The Nd:YAG laser has been used successfully for the treatment of mucosal malformations. In our own experience this method has been quite successful for pure venous malformations, especially if they are ectatic. The CO_2 laser has been more successful in treating the variegated combined venous-lymphatic malformations seen on the dorsum of the tongue.

Arteriovenous Malformations

Of all the vascular anomalies, arteriovenous malformations are the most feared and difficult to treat. Arteriovenous malformations are high-flow lesions that enlarge not by cellular hyperplasia but by hemodynamic mechanisms. Fortunately they are rare lesions in contrast to low-flow anomalies.

Cervicofacial arteriovenous anomalies may be noted soon after birth and grow proportionally with the child, but many will not be visible until years later when they become symptomatic. Rapid expansion may occur after local trauma, attempted excision or ligation, or with hormonal changes at puberty and during pregnancy.

The presenting complaint will frequently be a pulsatile tinnitus or buzzing heard best at night when it is quiet. There may be intermittent episodes of stabbing pain. In lesions

closer to the surface, sudden bleeding from a tooth socket or epistaxis may unmask an unsuspected lesion. On examination, the overlying skin may have a mild bluish-red hue with elevated temperature. There will be a palpable thrill with a bruit on auscultation. Congestige heart failure may be present if the arteriovenous fistula is large or of long standing. Arteriovenous shunting may diminish blood flow to adjacent areas and cause ischemic pain and/or necrosis of the skin. Recurrent severe episodes of bleeding may be life threatening. Some lesions may cause local bony destruction or involve the bone itself, producing radiolucent or multiloculated areas similar to low-flow anomalies. If these involve the mandible or maxilla, they may be manifested by a loose tooth with massive bleeding when it is extracted or bleeding around the tooth. As in other vascular anomalies, there is always the danger of disseminated intravascular clotting secondary to thrombotic consumption or local destruction of clotting factors.

The pathophysiology of arteriovenous fistula starts with a direct connection between the arterial and venous circulation. With direct flow from a high-pressure to a low-pressure system, two circuits develop, one with a high resistance through the capillary bed and one with alow resistance directly from the artery to the vein. As more and more blood passes through the low resistance circuit, the arteries enlarge and become tortuous, whereas the veins undergo commensurate hypertrophy to accommodate the flow. As the fistula enlarges, the veins become pulsatile as there is direct blood flow from the artery unimpeded by the capillary bed. As this flow increases, blood pressure in the artery beyond the fistula decreases and eventually there is a reversal of flow as the collateral circulation develops, and this supplies the fistula as well. As this happens, blood may be "stolen" from the area supplied by the distal artery, producing ischemia of the area and possibly leading to necrosis. As the shunting increases in volume, the systemic blood pressure would drop, but this is compensated for by increasing blood volume and cardiac output. If shunting reaches a critical point, the patient may suffer from high-output cardiac failure.

The treatment of arteriovenous malformations is hazardous and the results are frequently disappointing. If the patient is asymptomatic, it is best to follow the patient closely. Intervention is indicated for life-threatening problems such as hemorrhage, pain, pressure ischemic ulcerations, or heart failure. Treatment consists of embolization, surgical excision, or a combination of both.

In these high-flow lesions angiography is imperative. Angiograms are difficult to interpret because of the rapid blood flow. Contrast media may be preferentially swept through proximal arteries and veins, failing to demonstrate more distal arteriovenous fistula and collateral shunting. The dye may be so diluted in large venous lakes at the center of the lesion that minor arterial feeders are hidden. It is important that the intracerebral circulation also be evaluated to assess shunting between the internal and external carotid circulation.

In the operative therapy for these lesions, it has been well shown by multiple authors that proximal ligation of the feeding vessels is not only useless, but will result in disaster, especially in the cervicofacial area where there is such a rich collateral circulation. The lesion may undergo a rapid increase in size as collaterals are picked up and develop from the contralateral as well as the ipsilateral circulation and the internal carotid system. Ligating proximal vessels precludes the use of embolization in the future.

Embolization would appear to have the same risks as proximal ligation, but the goal is to embolize the center of the lesion first and work outward. It may be a curative, palliative, or preoperative effort. Ideally, embolization should be a preoperative effort to decrease blood flow for the surgical resection, which should be planned for 48 to 72 hrlater. If surgical resection is impossible because of the location and/or structures involved, embolization may be attempted as a curative procedure. The goal is to block as many shunts as possible, decreasing the size of the lesion and hoping that collateral circulation will be slow to develop. It is imperative that the venous lakes at the center of the lesion, especially if they are in bone, be obliterated as well, or they will act as sumps attracting new blood supply. If the embolization attempt is meant to be curative, we prefer to use vascular springs that are permanent and will not pass through into the distal circulation. In bony lesions, a direct stick into the venous lakes is possible, packing them with multiple springs. For preoperative efforts, absorbable materials such as Gelfoam are used. Palliative embolization may beused for relief of symptoms such as pain and bleeding, and when the lesion cannot be removed surgically.

As we have condemned proximal ligation of the arterial supply, the goal, if surgery is contemplated, should be en bloc resection as if for a tumor. Embolization will not reduce the amount of resection needed, but the goal is to make the blood loss manageable. Several other techniques may be used such as temporary mattress sutures around the periphery, temporary occlusion of feeding vessels with percutaneous transcatheter balloons or temporary clamps, or profound hypotensive anesthesia. Large lesions of the scalp, ear, and midface that have failed multiple therapies may require large resections of tissue with resurfacing by regional myocutaneous flaps or distant free flaps. Even with the best of intentions, these frightening lesions frequently recur. However, these patients may be helped for a significant length of time, and some will be cured.

Lymphatic Malformations

The lymphatic anomalies suffer from the same semantic problems as the other vascular anomalies that we have discussed. To be consistent with the terminology suggested by Mulliken and Young, as used throughout this chapter, all lymphatic anomalies would come under lymphatic malformations. This change in terminology, although technically correct, is difficult to use, since common usage of terms, such as lymphangioma and cystic hygroma are so entrenched in our medical literature and everyday language. Even recent articles that espouse the classification suggested by Mulliken and Young will revert to common usage terms, creating confusion as to the conditions they are describing and treatment they are suggesting. In the past, the term *lymphangioma* and *cystic hygroma* have been confusing and have been suggested by some to represent distinctly different entities. Today it is generally accepted that they are the same entity. For our purpose we will consider lymphatic malformations to be lymphatic anomalies consisting of spaces that may be unilocular, multilocular, or diffuse, lined with flattened epithelium, and containing a pale yellow fluid. There may be combined lesions with venous and lymphatic components, which are referred to as lymphatic-venous or venous-lymphatic malformations. As in venous malformations, these lesions do not undergo cellular proliferation as in a neoplasm. In general, they grow commensurate with the growth of the child, but may undergo rapid expansion during upper respiratory infections or with hemorrhage into the lesion.

It is generally accepted that lymphatic anomalies arise as a result of maldevelopment of the primitive anterior, posterior, or retroperitoneal lymph sacs. This could be a failure of these sacs to form a venous connection or some interruption of the drainage path of the lymphatics. The lack of egress of the lymphatic fluid produces large cysts or more compact anomalies if present in a restrictive tissue environment.

Lymphatic anomalies have an equal sex distribution. They are most common in the cervicofacial region, but also occur in the axilla, extremities, and trunk. These lesions occur at about the 6th to 9th week of gestation and may be detected as early as the 12th week by ultrasound. In a review article by Cohen et al, it was found that if a lymphatic malformation was detected in the antenatal period, 73% had elective termination of the pregnancy and 22% resulted in fetal death in utero. The most common location was in the posterior nuchal region. Some of those detected had spontaneously resolved by birth. There is a well-documented correlation between these cystic lymphatic lesions in the fetus and various chromosome abnormalities, especially Turner's syndrome. If this condition is found on ultrasound, amniocentesis for karyotyping is indicated.

The majority of lymphatic malformations are detected in the newborn nursery or during the first year of life, but Kennedy reported 45% of his cases were over age 20. These lesions may present as large multiloculated cystic structures to small cutaneous or mucosal blebs. The large cystic lesions may transluminate, and do not collapse with pressure as does a venous malformation. Large lesions may compress or distort the parapharyngeal airway and trachea, and extend into the mediastinum. Lesions may involve the orbit or periorbital tissue causing exophthalmus and muscular problems or visual disturbances similar to those seen in the hemangiomas of the orbital area.

Cellulitis of lymphatic malformations is a relatively common occurrence. The lesion will become tense, warm, and erythematous with rapid enlargement. This rapid enlargement may be life threatening if it intrudes on the airway by direct extension or extrinsic pressure, especially if the tongue or laryngeal structures are involved.

It is possible for the lymphatic abnormality to diffusely involve the tongue musculature, resulting in various degrees of macroglossia. Sometimes the mucosal surface of the tongue and/or cheek will be covered with small cystic or hemorrhagic vesicles that will intermittently bleed. Skeletal hypertrophy occurs with lymphatic malformations adjacent to the skeletal structures, especially of the mandible and maxilla.

Although growth in proportion to the child is characteristic, the question of spontaneous regression or improvement has not been answered. There are sporadic reports of spontaneus involution that is slowly progressive over a period of 2 to 6 years.

The treatment of lymphatic malformations is even more controversial than the other vascular anomalies. History is filled with the usual plethora of attempted modalities that were tried in other vascular lesions. These include a multitude of sclerosing agents, cautery, and irradiation. Because of the wide variety of presentations, to do nothing may be the least harmful. In general, the main therapeutic thrust is a well-planned surgical excision.

Two conditions that require emergent intervention are airway obstruction and infection. Airway obstruction is usually secondary to tongue involvement with the tongue being either infiltrated or pushed up and posterior by cystic lesions in the floor of the mouth. Because of the tongue position, these infants may be very difficult to anesthetize and intubate without losing control of the airway. Aspiration or incision of the cysts in the floor of the mouth may give temporary relief, but usually they are so multiloculated that significant decrease in size is not obtained. If there is significant airway obstruction present, a tracheotomy must be performed, which has its own unique risks for mortality. Some lesions may become so invasive that the naso-oro-hypopharyngeal areas will become partially obstructed. If oral alimentation is compromised, a gastrostomy may be necessary to prevent a failure-to-thrive condition.

Infection is the second complication requiring emergent treatment. In some cases a viral upper respiratory infection may cause rapid enlargement, but without cellulitis. If cellulitis occurs, it is usually from oropharyngeal organisms and should be treated as such. Penicillin should cover most oral organisms, but in children there may be a significant number of infections with staphylococci and hemophilus. In the acute stages, the child may be septicemic with a high fever, as well as with the localized cellulitis. In this situation the child is usually quite sick and should be hospitalized for blood cultures, aspiration and culture of the cyst fluid, and intravenous antibiotics. With the rapid changes in antibiotics, a specific recommendation may soon be out of date, and it would be best to consult with a pediatric infectious disease specialist for the best antibiotic to cover the most common organisms. At present we tend to use one of the cephalosporins or clindamycin. Once infected, these patients seem to be more susceptible to repeated infections. In this situation if there is just cellulitis without systemic signs, a prescription for oral antibiotics may be sufficient, to be used when early symptoms appear. If infections become frequent, one may be able to prevent recurrences with the use of prophylactic antibiotics such as those used for recurrent otitis media and sinusitis.

Surgical excision is the treatment of choice in symptomatic lymphatic malformations. Surgical excision should be preceded by good planning. In large lesions, radiological evaluation with either CT or MRI should be used to determine the extent of the lesion. They can be evaluated quite well with either or both modalities, giving the surgeon the perspective of what vital structures are at risk and if there is extension into the mediastinum. If lesions are relatively small, circumscribed, and easily outlined on palpation, x-rays are not needed.

Timing is controversial. If lesions are small and not causing complications, a wait-and-see approach is indicated. This is particularly true for parotid lesions, where a differential diagnosis between a hemangioma that will involute and a lymphatic malformation may be difficult without invasive studies. Since there are sporadic reports of regression, this hope may be held out to the parents, but with the warning that this outcome is unusual. Allowing time to pass and the child to grow may make surgical excision safer for the child in terms of fluid and blood loss, as well as easier in that vital structures such as nerves and blood vessels will be larger.

In large cystic lesions that are rapidly enlarging with the potential for causing complications, especially airway compromise, the surgeon's hand may be forced even in the neonate or first few months of life. These are usually cystic lesions involving the anterior

cervical and submaxillary triangles, but may also extend into the posterior cervical triangle. The rationale for approaching these lesions early is to prevent the necessity for a tracheotomy and/or gastrostomy. If the lesion is bilateral, staged excision may be best. The surgeon must be prepared for a prolonged, tedious dissection. It should not be added to a long surgical list at the end of the day, or one will be tempted to compromise the dissection. These lesions may be approached from a variety of incisions, depending on the extent and location of the lesion. Many times a horizontal midneck incision will suffice to remove lesions from the mandible to the clavicle, especially if extended across the midline or more posterior. If better exposure is needed, a superior and/or inferior limb of a half H may be used. The superior limb of the half H may be extended upward in the preauricular area to complete a parotid incision, if a parotidectomy becomes necessary. A classical McFee or inferior and superior horizontal incisions are also very cosmetic. Care must be taken to find and preserve vessels and nerves. Basically, the technique is similar to a conservation lymphadenectomy (Bocca technique) where all anatomic structures are preserved. One only needs to review the literature to see the high rate of complications such as facial nerve paralysis, Horner's syndrome, and paralysis of cranial nerves X, XI, or XII to see why people would recommend no surgery. The ramus mandibularis must be found and preserved, as well as the greater auricular nerve, sympathetic trunk, VII, X, XI, and XII cranial nerves, phrenic nerve, brachial plexus, and cervical nerves. The sternocleidomastoid muscle and internal jugular vein may usually be preserved and obviously the carotid artery must be preserved. As the dissection is carried out, the cysts may be accidentally ruptured or may have to be transected to preserve vital nerve or vascular structures. Although the goal is complete resection, this is frequently impossible. An attempt is made to dissect as much of the shiny cyst wall as possible, even after rupture has taken place. If the lesion extends into the floor of the mouth or substance of the tongue, an attempt is made to dissect these cysts as completely as possible, and where not possible, to at least open them. I have found it helpful in some of these deep cysts to lightly fulgurate the lining with the cautery.

Parotid involvement combined with a cervical lesion or as an isolated lesion needs speciall attention. Usually the entire parotid is involved, although the majority of the gland (80%) is lateral and inferior to the facial nerve. If one is forced to treat an infant or very young child, the facial nerve may be very small. Microscopic dissection or loupe magnification should be used to reduce risk to this vitally important nerve. Tedious, meticulous dissection is the watchword. If the physician does not have the experience or patience to do this, the patient should be referred. The physician must *do no harm!* Cysts in the deep lobe are dissected as best as possible without injuring the nerve, or are opened with a hemostat.

When the resection is completed, there is the question of how to drain the surgical site. In general, I prefer to use suction drains. One must be careful to have controlled any open vascular channels associated with the lesion. Upon occasion it seems that there is a localized coagulopathy condition leading to either significant bloody drainage and/or a hematoma. In those situations, reinspecting the wound and penrose drains may be necessary. One must account for and replace fluid loss, especially blood and protein, if there are large amounts of drainage in a small infant. Drainage may be necessary for several days to prevent a postoperative seroma or reaccumulation of lymphatic fluid. If there is a relatively rapid recurrence of cystic structures at the edge of a resection, reoperation is indicated. If the child has had cellulitis prior to surgery, prophylactic antibiotics should be used.

In those patients who have large lesions that have been observed without noticeable regression, surgical excision is indicated at age 1 to 5 years, depending upon its size and location. The same surgical principles as previously described should be used.

In patients with significant macroglossia demonstrated by extension outside the oral cavity with mandibular and/or maxillary distortion, a reduction glossectomy is necessary. This may be done with vertical and/or transverse wedge resections. One must be careful to preserve nerve function and enough muscular bulk to avoid interference with deglutition and speech. Mandibular or maxillary distortion may require orthognathic and/or orthodontic procedures.

Other treatment modalities may be used as adjunctive procedures. The CO_2 laser is useful in vaporizing lymphatic involvement of the larynx and hypopharyngeal structures. The Nd:YAG laser may be used on combined venous-lymphatic malformations, but does not work as well in pure lymphatic lesions where there is no pigment to absorb the laser rays unless high power-density ratios are used. Vaporization of the lymphatic lesions of the tongue and oral mucosa is better accomplished with the CO_2 laser.

There are sporadic reports using chemotherapeutic agents in the treatment of lymphatic malformations. Ogita et al have used a sclerosing agent OK-432 for direct injection into the lesion. In nine patients treated, eight had complete resolution of the lesion. Cyclophosphamide has also been reported as having a good response in two cases.

Summary

In summary, vascular anomalies are difficult lesions to treat. However, if careful evaluation is carried out and an appropriate diagnosis is made, the appropriate therapy may be chosen.