Vascular anomalies are disorders of abnormal vasculogenesis or lymphogenesis.\(^1,\)\(^2\) Vascular malformations are labeled according to the vessel type including venous malformations, lymphatic malformations, capillary malformations, arteriovenous malformations, and mixed types (lymphatico-venous malformations). Vascular malformations may also be divided into low flow and high flow lesions. Low flow lesions include most vascular malformations, including capillary (port wine), venous malformations, lymphatic malformations, and combined lesions. High flow lesions include arteriovenous malformations (AVMs), arteriovenous fistula (AVFs), or aneurysms, which all have a high flow arterial pedicle. Venous malformations, lymphatic malformations, and AVMs are the most common vascular malformations to affect the aerodigestive tract, but are less common overall than hemangiomas. All types of vascular anomalies may involve the airway, causing varying degrees of upper airway obstruction as well as dysphagia and bleeding.

Certain signs and symptoms may implicate airway involvement with a hemangioma or vascular malformation. The most common pathology to affect the airway is a hemangioma and usually one with segmental distribution. The most common symptoms include biphasic stridor, recurrent croup, and retractions. Flexible laryngoscopy performed in the outpatient setting may show vascular staining of the endolarynx and may reveal a subglottic mass, but it may be difficult to appreciate small subglottic or tracheal hemangiomas on awake flexible laryngoscopy. It is necessary to distinguish a vascular anomaly from other airway lesions such as a congenital cyst. This is accomplished with imaging and endoscopy.

**AIRWAY IMAGING**

At the initial presentation of a child with stridor, plain films of the airway will show a tapered subglottic airway and a subglottic mass with a subglottic hemangioma and are very useful for assessing the subglottis (Fig. 1). This is to be differentiated from subglottic stenosis or subglottic cysts. With these two pathologies, there is usually a history of intubation except with congenital subglottic stenosis, which is quite rare.

Sonography may be useful for defining vascular anomalies, but has limited resolution. Both contrast-enhanced CT/CT angiography (CTA) and MRI/MR angiography (MRA) are the most precise at defining vascular lesions and their extent of involvement in the aerodigestive tract. The injection of an arterial phase contrast agent is most important for demonstrating the vascular anomalies.
nature of the lesions and the extent of disease in the neck. However, one may have a limited view of the actual airway. MRI is most helpful for defining lymphatic malformations and determining microcystic versus macrocystic disease. Angiography is useful for determining the structure and blood supply of AVM, AVF, and aneurysm and allows for embolization before definitive surgery.

OPERATIVE ENDOSCOPY

Microlaryngoscopy and bronchoscopy (MLB) is an essential technique for confirming a diagnosis of an airway vascular anomaly such as a subglottic hemangioma. The technique is performed under general anesthesia with spontaneous ventilation. This allows for the most accurate assessment of the airway. First, flexible laryngoscopy is performed to study the dynamic characteristics of the larynx such as vocal cord mobility and epiglottic position, glossoptosis, and laryngomalacia. A flexible scope with side port suction is used to do this.

Microlaryngoscopy is performed with a rigid anterior commissure-type laryngoscope such as a Benjamin or Parsons placed in the vallecula (Fig. 2) and used to expose the endolarynx and subglottic airway. This will reveal a subglottic hemangioma just below the vocal cords (Fig. 3). Magnification from a Storz Hopkins telescope is used to visualize the subglottic airway and the scope can be passed down to the level of the carina. It is important to carefully look at the trachealis muscle in the back wall of the trachea for staining, fullness, and signs of vascular disease (Fig. 4). Tracheal hemangioma may be subtle and its treatment is challenging.

MANAGEMENT OF HEMANGIOMAS AFFECTING THE AIRWAY

Hemangiomas are often seen in infants in the head and neck region and may involve the upper aerodigestive tract and airway. Involvement of the airway may result in varying degrees of upper airway obstruction as well as dysphagia and bleeding. Based on the distribution of the hemangioma, one may subclassify hemangiomas into focal, multifocal, or segmental based on the distribution characteristics. Focal implies a solitary lesion, whereas multifocal implies multiple. Segmental lesions occupy a dermatomal distribution. Most
Hemangiomas are solitary; however, the distribution may have an impact on the prognosis and the potential for airway involvement. Segmental hemangiomas are more likely to have involvement of the aerodigestive tract relative to solitary lesions, and multifocal lesions are more likely to simultaneously involve the liver or other areas of the gastrointestinal system.

When there is involvement of the airway, the symptoms are often stridor, croup, and respiratory distress. It is important to characterize the severity of the respiratory distress and the characteristics of the stridor. An infant with no history of intubation with a croupy (barky) cough and biphasic stridor localizes the lesion to the subglottic larynx. In the case of an infant with skin hemangiomas, the airway lesion would likely be a subglottic hemangioma. Following physical examination, it is necessary to proceed to imaging and endoscopy to confirm a diagnosis.

Hemangiomas may be classified by thickness as superficial, deep, or compound, as well as by distribution as localized, multifocal, or segmental. A localized hemangioma would involve a solitary lesion in the skin or airway, whereas multifocal includes 6 or more, and segmental involves a distribution in a known anatomic region such as V3 segment of the fifth nerve. Generally, airway symptoms are more common with segmental hemangiomas, and the so-called “beard distribution” in V3 is associated with a 20% to 30% involvement of the upper airway. The classic and most common finding is subglottic hemangioma, which is most often laterally based or less commonly posterior.

Less common sites affecting the airway include postcricoid, tracheal, mediastinal, thoracic, and esophageal. They may stain the posterior trachealis muscle (see Fig. 4). Airway symptoms include biphasic stridor and recurrent croup. Diagnosis may be suggested by history and lack of another cause of subglottic stenosis, such as prolonged intubation. Plain neck films may suggest the diagnosis.

Treatment of airway hemangioma depends greatly on the severity of symptoms, degree of airway obstruction, and the position of involvement in the airway. The treatment options for infantile hemangiomas of the airway are as follows:

- Observation
- Steroids
- Interferon: Historic – spastic diplegia
- Laser: CO₂, KTP, Candela
- Tracheotomy: Discuss risk
- Open resection with or without cartilage augmentation
- Propranolol.

For mild cases with minimal or no symptoms, observation is all that is necessary. The size of a small hemangioma may not cause turbulent airflow or may involve nonendoluminal sites, such as the postcricoid region, which will cause no major airway symptoms. In such cases, no treatment is necessary, but close observation with flexible laryngoscopies in the ambulatory setting every 3 to 4 months is needed. However, it is important to understand that the symptoms may initially worsen as the hemangioma proliferates. Bilateral hemangiomas may impact the airway the most (Fig. 5).

Mild cases or those that present with sudden acute airway symptoms may benefit from a short course of corticosteroids. This may be helpful during acute exacerbations, such as with upper respiratory infections. In addition, aerosolized

![Fig. 4. Hemangioma staining of the posterior trachealis muscle.](image)

![Fig. 5. Bilateral subglottic hemangioma with total airway obstruction.](image)
When pharmacologic means are not sufficient alone, lasers, such as CO$_2$ or potassium-titanyl-phosphate (KTP), can be used to treat surface hemangiomas in the airway. In the 1980s and 1990s, it remained a workhorse for subglottic hemangiomas; however, experience has shown that the recurrence rate with this technique is high. In addition, the laser may need to be used multiple times to achieve a stable airway because it treats only the surface and may not be effective at all for bilateral or circumferential disease. In addition, overaggressive use of the laser may result in subglottic stenosis. In the same way, the microdebrider, as demonstrated by Pransky and Canto, may be useful for focal lesions amenable to laser ablation. Although this technique has some of the same limitations as the laser, it is easier to set up than the laser and has less chance of injury from its use.

Open resection and cartilage graft augmentation has been reported as an effective treatment for subglottic hemangiomas unresponsive to more conservative treatments. The indication is a posterior or laterally based subglottic hemangioma that has failed steroids or propranolol. The procedure starts with a microlaryngoscopy, where the airway is assessed and secured with a small endotracheal tube orally. At this point, the patient may be given a course of intravenous steroids that may decrease bleeding during the resection.

The patient is positioned with the neck hyperextended and a transverse incision is made in the neck at about the level of the cricoid cartilage. A superior and inferior subplatymal flap is elevated to expose the laryngotracheal complex and the thyroid notch. This may expose extralaryngeal hemangioma in the neck (Fig. 6). Stay sutures are placed on the cricoids, which is opened in
the midline. The incision is performed with a Beaver blade and extended up to just under the anterior commissure. It is important to avoid violating the AC unless necessary. Posterior-based lesions are incised in the direct midline and the lesion is carefully dissected off the cartilage plate of the posterior cricoid (Fig. 7) using otologic instruments such as the round knife, Rosen needle, and microtabe knife. An operating microscope may be prepped and used to facilitate visualization. Laterally based lesions may be approached by elevating a submucosal flap off the hemangioma from the laryngofissure down. When the lesion is resected completely, the flaps are sutured back into position using small vicryl. At this point, the airway is often stenotic and a small thyroid alar graft is harvested and used to augment the size of the subglottic airway (Fig. 8). The graft is harvested from the thyroid alar and usually measures no more than 5 to 6 mm in length and 2 to 3 mm in width in a small infant, but this is enough to offset the stenosis.

The wound is closed and the infant is kept nasally intubated for 5 to 7 days and then undergoes a microlaryngoscopy and bronchoscopy to evaluate the airway. If everything looks satisfactory, the patient is extubated the following day in the ICU. At CHOP, we have had 12 patients undergo this surgery, all with a remarkable resolution of their symptoms.

For life-threatening lesions that do not respond to any therapy, tracheotomy is a treatment option. Although this is usually a temporary measure, as hemangiomas will involute over time, it carries the risk of speech delays and complications related to the tracheotomy such
as mucous plugging or accidental decannulation. Therefore, the tracheotomy should be reserved as a last option.

In summary, a complete paradigm shift in the treatment of airway hemangiomas has occurred over the past several years. Newer technologies have replaced the CO₂ laser. Open resection has become an option for avoiding a tracheotomy and propranolol appears to be a promising therapy and may eventually supplant surgery and corticosteroids. The main take-home message is that many hemangiomas of the airway cannot be assumed to fully involute and many require definitive therapy. We recommend our treatment algorithm as follows:

- Initial airway symptoms: short course of steroids and then rapid taper at the same time as workup before starting propranolol (MLB, MRI, electrocardiogram [EKG], brain MRI, glucose).
- If no rebound, follow clinically for symptoms. If airway symptoms are present, consider trial of propranolol.
- If there is immediate rebound, consider early trial of propranolol. Get up to 2 mg/kg for 2 weeks and re-scope. If response is seen, then continue for 6 months and then taper.
- If no change on propranolol and focal or bilateral subglottic hemangioma, then consider open resection.
- If diffuse and no change on propranolol, consider tracheotomy and/or laser ablation.

**MANAGEMENT OF LYMPHATIC MALFORMATIONS AFFECTING THE AIRWAY**

Lymphatic malformations can vary from small isolated neck masses to extensive cervico-facial processes that infiltrate into large tissue planes. There is a staging system proposed by deSerres and colleagues that may be used to predict outcome. Stage I includes unilateral infrahyoid disease. Stage II is unilateral suprathyroid disease. Stage III is unilateral suprathyroid and infrahyoid disease. Stage IV is bilateral suprathyroid disease and stage V is bilateral suprathyroid and infrahyoid disease.

For treatment purposes, lymphatic malformations may also be subdivided into macrocystic and microcystic. By definition, macrocysts are larger than 2 cm and were formerly referred to as “cystic hygromas” and microcystic lesions were referred to as “lymphangiomas.” The stage and cystic structure of the lymphatic malformations is crucial in determining the ideal surgical approach.

Lymphatic malformations are often noted at birth and may be detected by prenatal 2-dimensional ultrasound.³ They occur most often in the cervicofascial region and they may be seen in the axilla, chest, and mediastinum as well. The cystic lesions can become infected and flare up during upper respiratory infections. Antibiotics and steroids may treat such flare-ups. Lymphatic malformations that infiltrate into the supraglottic portion of the larynx will cause severe airway obstruction. Extensive cervicofascial lesions will also secondarily involve the supraglottic larynx or regions of the pharynx causing airway compression. This may prevent safe extubation and lead to tracheotomy dependency.

Lymphatic malformations can be easily visualized by ultrasound, MRI, or CT scanning. In the interest of radiation reduction, CT should be reserved for acute lesion presentations such as airway compression or acute hemorrhage/infection of the lesion. On ultrasound, macrocystic lymphatic malformations appear as cystic cavities with internal septae and often debris. Microcystic lesions have small cystic cavities often so small that the lesion appears solid. Ultrasound is a useful imaging tool to assess acute hemorrhage and infection. Both hemorrhage and infection will cause the cyst contents to appear brighter than normal. In addition, in infection the cyst wall will demonstrate increased blood flow. Lymphatic malformations on CT appear as fluid-filled low-attenuation lesions, occasionally with fluid-fluid levels that can represent acute or subacute bleeding. Peripheral contrast enhancement of the walls may occur with bleeding or infection. The relationship of cervico-facial lesions to the airway can be elegantly demonstrated. Three-plane reconstruction can also enhance assessment of the true extrinsic effect of the lesion on the airway. On MRI, lymphatic malformations appear as multicystic masses that insinuate between tissue planes. These lesions demonstrate predominantly fluid-type characteristics on all MRI sequences with varying degrees of septation. MRI is the ideal imaging tool, as it can assess lesion characteristics in relation to the aerodigestive tract and the presence of hemorrhage or infection in multiple planes without radiation. Younger children do have to be sedated because of the time it takes to complete the study.

As previously mentioned, staging, extent of disease, age, associated symptoms, time to diagnosis, and completeness of excision strongly influence prognosis.¹⁷ The histologic pattern and length of time to treatment did not influence...
prognosis. The most important factor is the ability to perform complete surgical excision without significant morbidity. The best results are for those lesions that can be cleanly resected with minimal morbidity. These are most often those lesions that are isolated to the infrahyoid or posterior triangle position of the neck. More extensive lesions that infiltrate into vital structures may require staged resections that are quite challenging or other treatments such as sclerotherapy or laser ablation. Treatment options for lymphatic malformations include observation for small lesions, early stage I with no cosmetic or functional compromise. They may persist, but cause no problems and may be watched for expansion. The decision to operate would be based on age of the patients and concerns about future symptoms.

1. Surgical excision: For focal lesions (stages I to III) involving the neck, parotid, or mediastinum, show neck lesions that are well circumscribed and can be completely resected to reduce the risk of recurrence. More extensive lesions (III to V) and microcystic disease that is not amenable to sclerotherapy may be resected in staged fashion.³

2. Laser ablation for compressed lesions, especially those that appear on mucosal surfaces, such as tongue lymphatic malformations. The CO₂ or neodymium:yttrium aluminum garnate (Nd:YAG) laser may be used to ablate compressed tongue lymphatic malformations (Fig. 9).¹⁸

3. Radiofrequency ablation may be used for the same type of microcystic lesions of the tongue and supraglottic larynx. The advantage is the avoidance of thermal energy and complications associated with laser use.¹⁹

4. Microdebrider excision may be useful for the large supraglottic lesions and debulk lesions causing supraglottic airway obstruction. We have used it in piecemeal fashion to reduce supraglottic obstructive lesions (Fig. 10).

Sclerotherapy with a variety of sclerosing agents is the treatment of choice for more extensive macrocystic lesions that are not amenable to complete surgical excision. This would include large cervico-facial lesions or lesions that extend to regions that are not easily exposed such as skull base or parapharyngeal space (Fig. 11).²⁰ All have been shown to be effective with respect to treatment of lymphatic malformations with success rates ranging between 92% and 100% for macrocystic and 20% and 64% for microcystic lesions. Microcystic lesions tend to require more sessions than the macrocystic because of the presence of numerous small cysts. Different sclerosing agents have been used effectively for the treatment of lymphatic malformations, including doxycycline, ethibloc, absolute alcohol, sodium tetradecyl foam, bleomycin, and OK432.²¹–²⁸ Sclerotherapy of lymphatic malformations is generally performed with ultrasound guidance.

In our institution, for large cervico-facial lesions, we use doxycycline via catheter instillation and 3 consecutive treatments are performed at 24-hour intervals. The doxycycline is removed after 6 hours. The children are intubated and monitored in the ICU setting for the duration of the treatment (3–4 days). Preextubation MR imaging is performed to evaluate the residual extent of the lesion, inflammation, and airway status. The total dose of doxycycline used in our practice ranges from 150 to 200 mg per instillation in neonates to
a maximum of 1 g in older children. Neonates receiving more than 200 mg are more likely to experience side effects such as hypoglycemia, metabolic acidosis, and hemolytic anemia. Because of the high incidence of spontaneous infection together with an increased risk upon accessing the lesion, prophylactic antibiotics have been recommended immediately before sclerotherapy of lymphatic malformations.21–23

Tracheotomy may be necessary to secure the airway with lymphatic malformations that cause severe upper airway obstruction from involvement of the supraglottic larynx or massive cervicofacial lymphatic malformations. The tracheotomy may need to be placed in early infancy.

The EXIT procedure, or ex utero intrapartum treatment procedure, may be required to secure the airway during delivery for massive cervicofacial disease that presents on delivery making the securing of the airway difficult. Three-dimensional ultrasonography and fetal MRI can accurately diagnose prenatal lymphangiomas with a great deal of accuracy and predict delivery problems. Those lesions that completely obstruct the oropharynx may require an emergent airway at delivery and a planned EXIT procedure may be necessary.3

The EXIT is an extension of a standard caesarian section, where an incision is made in midline of the uterus. The baby is delivered on the placental circulation, and a fetal/airway team then establishes an airway either through endotracheal intubation or emergency tracheotomy. Once the airway is secured, the placental artery and vein can be ligated. Large lymphatic malformations that obstruct the oropharynx may make intubation difficult or impossible. In such cases, the EXIT procedure permits time to establish a safe airway. Because only a few centers in the United States and the world perform this highly specialized procedure, any baby with an obstructing mass discovered on ultrasound should be referred to such a center for evaluation.

**MANAGEMENT OF VENOUS MALFORMATIONS AFFECTING THE AIRWAY**

Venous malformations may involve a number of sites in the upper airway and may be quite extensive, like their counterpart the lymphatic malformations. They can involve the base of the tongue, pharynx, supraglottic larynx, and trachea. They most commonly occur in the supraglottic larynx followed by the trachea. In these locations, they can cause varying degrees of airway obstruction. The association with smooth and skeletal muscle may lead to compression of these lesions. Many lesions may remain small and require no treatment. Laryngotracheal involvement is rare. Large lesions may cause airway obstruction, voice change, and bleeding and hence require active intervention.

Venous malformations can be imaged with ultrasound, CT scanning, or MRI. Ultrasound with Doppler imaging provides information on the patency of the vessels, compressibility, and any areas of thrombus. Ultrasound is limited in regions adjacent to the airway. MRI with contrast provides 3-plane information regarding the spatial relationships of the lesion to the aerodigestive tract. On flow-sensitive sequences, areas of thrombosis can be identified. The relationship of the lesion to normal or abnormal outflow veins can be mapped.

These lesions usually do not have a distinct capsule and therefore complete surgical excision is usually not possible. The mainstay of surgical treatment for lesions affecting the airway involves a combination of laser photocoagulation with the laser including the CO₂, Nd:YAG, or the KTP.24,31 The laser is used in a noncontact fashion on the surface of the lesion and can be combined with sequential sclerotherapy.32 Most are approached with endoscopic exposure. Sequential sclerotherapy creates inflammation and sclerosis of venous channels with subsequent involution.

Low-flow venous malformations can also be treated percutaneously with a variety of sclerotherapy agents such as ethanol, sodium tetradecyl sulfate foam, ethibloc and polidocanol, and endovenous laser therapy.22,33–41 Sclerotherapy is performed with a combination of ultrasound and fluoroscopic guidance. Ultrasound provides good visualization of the lesion during needle access and fluoroscopy is used to monitor the sclerosant.
injection to decrease the risk of extravasation or undesired egress into normal veins. Sclerotherapy of lesions in the cervico-facial region should be performed with general anesthesia for airway protection. In addition, the injection of the sclerosant is painful and postprocedure edema may compromise the airway.

In our institution, the sclerotherapy agent of choice is sodium tetradecyl foam, the foam being created by adding a combination of oily contrast medium (ethiodol) and air. The foam consistency creates increased surface tension enabling greater contact with the vessel wall for a longer period than more liquid agents.

Ethanol is commonly used as an alternative agent in many centers. The recommended ethanol dosing is 1 mg/mL to a maximum of 50 mL, which in children should be conservatively limited to 0.5 mg/kg if possible. Serum ethanol levels in children have been recorded after sclerotherapy to levels significant for the risk of respiratory depression, cardiac arrhythmias, seizures, rhabdomyolysis, and hypoglycemia. In addition, coagulation disorders have also been described with the use of ethanol for sclerotherapy.

In general, on completion of the procedure edema will be present. The swelling will maximize over 1 to 2 days. Swelling and induration of the skin have been shown to predict both therapeutic effect and prolonged recovery in low-flow vascular malformations.

MANAGEMENT OF ARTERIOVENOUS MALFORMATIONS AFFECTING THE AIRWAY

These high flow vascular malformations can occur anywhere in the head and neck region and rarely affect the upper airway. Although they may occur in the trachea, bronchi, or larynx, they most commonly occur in the lung parenchyma. Pulmonary arteriovenous malformations are commonly associated with hereditary hemorrhagic telangiectasia and may be a source of hemoptysis. Diagnosis is performed through diagnostic angiography and treatments can be performed by embolization at the time of angiography. They can be followed by contrast-enhanced CT of the chest. Embolization can also precede surgical excision as a means for minimizing bleeding. In rare circumstances, surgical resection may be considered after preoperative embolization.

Arterial malformations are the most problematic and symptomatic of the vascular malformation group. Because of their size and location, most arterial malformations are inoperable or require extensive, potentially disfiguring resection. Transcatheter and percutaneous nidal embolization is now often the first therapeutic option and is an effective approach that can be used as a palliative procedure or as an adjunct to a surgical resection.

Arteriovenous malformations can be embolized either by direct percutaneous puncture into the nidus or by using an endovascular approach or a combination of both. Several agents have been used, including absolute alcohol, onyx, and adhesive glue. These are the most penetrating agents if the lesion is to be embolized for potential “cure.” If the embolization is preoperative, additional agents such as coils and polyvinyl alcohol particles are used to reduce lesional blood flow.

SUMMARY AND FUTURE DEVELOPMENTS

The opportunities for treatment of vascular anomalies of the airway will change dramatically as advances in new technologies emerge. We expect to see a major evolution in the care of patients with vascular malformations of the airway. Improvements in treatment will include newer surgical techniques, such as radiofrequency ablation or improved lasers, and new treatment modalities, such as new vasoactive drugs and highly selective beta blockade. In addition, early detection of novel molecular signals to determine aggressiveness and predict clinical behavior is likely to influence treatment. Last, interventional radiologists will continue to make advances in minimally invasive techniques. Open surgical techniques that we use today may become obsolete in the future.

REFERENCES


