Introduction

When examining the patient with a neck mass, the initial consideration should be the patient’s age group – i.e. pediatric, young adult, and adult. Within each group the relative incidence of congenital, inflammatory, and neoplastic disease should be considered as most neck masses fit into one of these categories. Pediatric patients generally exhibit inflammatory more frequently than congenital neck masses, with the incidence of neoplastic masses being relatively low. This is similar in the young adult age group. In adults, especially older, the first consideration is generally neoplastic with less emphasis on inflammatory and much less on congenital. After age, the location of the neck mass is a very important consideration. This is especially true in the congenital neck mass where they usually occur in consistent locations. The most common congenital masses found in the lateral neck include branchial cleft cysts and laryngoceles. Lesions in the midline include thyroglossal duct cyst, dermoid cyst, thymic cyst, and teratomas. Hemangiomas and the vascular malformation lesions typically involve large areas of the neck without true localization to one specific area. The esophageal duplication cyst, a rare congenital mass, also deserves attention.

Branchial Cleft Cysts.

The embryology associated with the branchial apparatus in the development of vital head and neck structures is complex. It is aberrations from this finely controlled system that branchial cleft cysts, sinuses, and fistulas arise. The branchial apparatus begins its developmental course at the second week of fetal life and is generally felt to be complete at the sixth to seventh week of life. Appearing at the lateral aspect of the foregut, the branchial apparatus consists of five arches (mesodermal) that are made by an external ectodermally lined groove (branchial cleft) and an internal endodermally lined concavity (branchial pouch). Each branchial arch, branchial cleft, and branchial pouch have well defined structures derived from them. The five branchial arches are numbered in a cranial to caudal fashion; however, the fifth branchial arch does not appear on the surface of the branchial apparatus and is, by convention, called the sixth arch.
Each branchial arch contains a cartilaginous skeleton, muscle rudiment, nerve, and artery. For instance, the mandible develops from cartilage of the first arch, while the second and third arches give rise to the hyoid bone. The fourth and sixth arches are forerunners to the laryngeal skeleton. Regarding the artery associated with each arch, the first and second arch arteries typically degenerate. A persistent stapedial artery is an example of failure of the second branchial arch artery to degenerate. The proximal aspect of the internal carotid artery, external carotid, and common carotid is derived from the third branchial arch. The distal aspect of the internal carotid artery comes from the dorsal aorta. The arch of the aorta as well as the proximal aspect of the right subclavian artery is felt to come from the fourth arch. Finally, the sixth arch artery on the left persists as the ductus arteriosus and pulmonary artery on the right side. Regarding the nerve / muscle unit, the trigeminal nerve (CN V) and associated musculature such as the those involved in mastication, anterior belly of the digastric, mylohyoid, tensor veli palatini, and tensor tympani all come from the first branchial arch. The second arch has the seventh cranial nerve associated with it and the following musculature – muscles of facial expression, platysma, stapedial, and posterior belly of the digastric. The glossopharyngeal nerve (CN IX) supplies the third branchial arch – innervating the stylohyoid muscle and superior constrictor. Finally, the fourth arch is supplied by the vagus nerve while the sixth branchial arch by the spinal accessory.

In the development of the branchial apparatus, the first and second arches grow in a caudal fashion; meanwhile, there is an impressive growth of mesoderm that forms the epipericardial ridge. The epipericardial ridge contains the mesodermal rudiments of the sternocleidomastoid, trapezius, and the infrahyoid / lingual musculature. Because of this mesodermal growth, the third and fourth arches recess into a deep ectodermal pit -- forming the cervical sinus of His. Over time there is so much growth of the surrounding structures that the opening into the cervical sinus eventually becomes narrowed to a small channel which is then referred to as the cervical duct. This will eventually be obliterated. Branchial cysts arise from retained portions of this cervical sinus. Branchial fistulas, epithelial lined tracts connecting the skin to the lumen of the foregut, may arise because of persistence of the cervical sinus in addition to breakdown of the branchial plate or the closing membrane between clefts and pouches. In contrast, the branchial sinus is seen as an epithelial lined tract opening onto the skin or the lumen of the foregut and ending blindly into the deep tissues of the neck. This occurs because of a persistent cervical duct or breakdown of the closing membrane.

As development continues the branchial clefts and pouches are also obliterated with the exception of the first branchial cleft. In fact, portions the first branchial cleft deepens to form the external auditory meatus. Briefly the structures derived from the first branchial pouch includes the eustachian tube, the tympanic cavity, the mastoid antrum and cells, and the tympanic membrane, while the second branchial pouch forms the palatine tonsils and supratonsillar fossa. From the third pouch come the inferior parathyroid, the thymus, and the piriform fossa. Finally, the fourth branchial pouch produces the superior parathyroids.

**First Branchial Cleft Cysts.** These branchial anomalies are further subclassified into type I and II. The type I first branchial cleft anomaly is *ectodermal* in origin and is considered to be a duplication of the external auditory canal, while the type II anomalies are considered to be duplication errors of the membranous external auditory canal and the pinna that contain *ectodermal* elements (skin) and *mesodermal* elements (cartilage). In general, the first branchial
anomalies compromise 1-8% of all branchiogenic anomalies. In the type I mass, they appear in the periauricular region as a cyst, sinus, or a fistula connecting to the skin and the external auditory canal. These lesions are typically lateral to CN VII and may be embedded in the parotid tissue.

In contrast, the type II first branchial cleft cysts are typically located just inferior or posterior to the angle of the mandible. Because these lesions are intimately associated with the parotid gland, these may lie lateral, medial, or in between the branches of the facial nerve.

Clinically, the patient with a first branchial cleft anomalies may present with a parotid mass, parotitis, or ototrauma / otalgia if there is a tract with the external auditory canal. The treatment of these lesions is surgical. Pre-operatively, it may be helpful to outline a sinus tract or fistula with radiopaque dye to confirm its course. In addition, several authors advocate MRI to evaluate the lesion in relationship to the parotid gland. Especially with the type II first branchial cleft anomalies, a superficial parotidectomy is typically required to avoid inadvertent injury to the facial nerve as well as removing the entire lesion. Portions of the cartilaginous external auditory canal may need to be excised along with involved skin.

**Second Branchial Cleft Cysts.** This represents the most common type of the branchiogenic anomalies. Again, failure of the cervical sinus to obliterate results in the persistence of an epithelial lined space that may communicate either or both internally or externally. These typically present spontaneously as a painless, fluctuant mass below the angle of the mandible and anterior to the sternocleidomastoid. These can suddenly change size after a common upper respiratory infection. The diagnosis is usually evident as few other cystic masses occur in this region. Anatomically, the tract passes superiorly, lateral to the carotid artery, CN IX, and CN XII. It courses superior and lateral to CNIX to turn medial to pass in between the internal and external carotid artery. The tracts typically terminate close to the middle constrictor muscle or may have an internal opening into the tonsillar fossa. The treatment again is surgical. Again, pre operative radiographic studies such as MRI / CT may be useful to demonstrate the lesions location to vital structures.

**Third Branchial Cleft Cysts.** These are very rare branchiogenic anomalies. They appear as a mass anterior to the sternocleidomastoid. The tract ascends lateral to the common carotid artery, passes posterior to the internal carotid artery, superior to CN XII, and inferior to CN IX. It then coursed medially to pierce the lateral aspect of the thyrohyoid membrane to open into the piriform sinus. Typically, it pierces the thyrohyoid membrane superior to the internal branch of the superior laryngeal nerve. Treatment is surgical.

**Fourth Branchial Cleft Cysts.** Fourth branchial cleft anomalies are extremely rare with only a number of cases actually reported in the literature. When these appear as left sided lesions, the tract begins at the apex of the piriform sinus. They then descend to exit the pharynx caudal to the superior laryngeal nerve, the cricothyroid muscle, and the thyroid cartilage. The tract courses inferiorly, lateral to the trachea and the recurrent laryngeal nerve. The tract then falls into the thorax and loops around the aortic arch before ascending in the neck posterior to the common carotid artery. The tract crosses CN XII before descending again to open into the skin at the anterior aspect of the lowest portion of the sternocleidomastoid. In the right sided lesion, the tract
passes anteriorly beneath the subclavian artery before passing superiorly into the neck. Because of the rarity of these lesions, there has not been a standard established for surgical management. Some authors feel that complete exposure of the mediastinal and cervical components is unnecessarily aggressive and likely not indicated.

It has become recognized that both third and fourth branchial anomalies have been associated with recurrent acute suppurative thyroiditis in children. Because these anomalies are almost always present on the left side, the child may present with left paratracheal tenderness, fullness, and with associated fever. Cultures will typically demonstrate E.Coli, Klebsiella, Proteus, and Clostridium. In cases of suppurative thyroiditis that fails antibiotic therapy, hemithyroidectomy with closure of the internal piriform sinus tract opening is recommended.

**Laryngoceles.**

While not a common congenital neck mass, laryngoceles may be seen as a laterally located lesion. Although seen in all age groups, these are found most frequently males in the fifth and sixth decade of life. The causative factor of laryngoceles is felt to be either a congenital enlargement of the saccule or acquired by increased, sustained intralaryngeal pressure (i.e. trumpet player). There are three types of laryngoceles—internal, external, and combined. The internal laryngocele is found entirely within the larynx and typically extends posteriorly and superiorly to the false vocal cord and aryepiglottic fold. As it is intraluminal, it does not present as a neck mass. The external laryngocele is seen as a lateral swelling in the neck and passes superiorly through the opening of the thyrohyoid membrane at the entrance site of the superior laryngeal nerve and vessels. Finally, the combined laryngocele has features of both internal and external. Clinically, presenting symptoms includes lateral neck mass, dysphagia, cough, dyspnea, and occasionally a gurgling sensation as the dilatation releases the contained air. On physical examination, the external and combined laryngoceles enlarges as intralaryngeal pressures are elevated. These masses are easily compressible. Additionally, these may present as an acute cervical inflammation if the sac becomes secondarily infected, filled with a purulent fluid, forming a laryngopyocele. Besides the classic clinical picture, a CT scan will outline the lesion.

The management of internal and external laryngoceles is surgical, usually through an external approach. Especially in the adult patient, it is important to perform a thorough endoscopic examination rule-out any underlying obstructive pathology that may be creating a check-valve within the ventricle. In the adult, there is an estimated incidence of an obstructing carcinoma ranging from 2 – 18%. For the external laryngocele, an external approach is required. The cyst mass must be traced to its exit site at the thyrohyoid membrane with careful identification of the superior laryngeal nerve. If there is no significant internal component, the cyst may be ligated without entering the larynx. For a combined laryngocele, however, a limited lateral thyrotomy may be indicated for complete exposure. For small, isolated internal laryngoceles, several reports have shown success with transoral laser excision with a bivalved laryngoscope.

**Teratomas and Dermoid Cysts.**

Both teratomas and dermoid cysts are seen as developmental anomalies involving pluripotential embryonal cells. Two theories have been suggested to explain these lesions – (1) isolation of
pluripotent cells during embryogenesis and subsequent disorganized growth of these cells and (2) the germ layers may be buried into deeper tissue at points of failed fusion lines. Teratomas have been found to occur in 1:4000 births; however, less than 10% actually affect the head and neck region. More commonly, these masses are found in the sacrococcygeal, mediastinal, retroperitoneal, and gonadal regions. In the head and neck region, the most commonly involved sites include the orbit, nose, nasopharynx, oral cavity, and neck.

To clarify confusion, these lesions classified into four groups that recognizes both the germ layer of origin and the degree of tissue organization. Based on this criteria, there are four characteristic lesions – (1) dermoid cyst, (2) teratoid cyst, (3) teratoma, and (4) epignathi. Dermoid cysts are composed of mesoderm and ectoderm and may contain hair follicles, sebaceous glands, and sweat glands. These lesions are midline, painless masses, typically found in the submental region. These may be distinguished from a thyroglossal duct cyst in that they do not elevate with tongue protrusion. Although they are almost uniformly located below the mylohyoid muscle, these lesions may be large enough to protrude into the anterior floor of mouth, resembling a ranula. Treatment is surgical excision. Teratoid cysts are composed of ectoderm, endoderm, and mesoderm. The lining of the cyst ranges from a stratified squamous epithelium to a ciliated respiratory epithelium. The characteristic feature of the teratoid cyst is the poor degree of differentiation of all the germ layers. Teratomas are also composed of all of the germ layers. In contrast to the teratoid cyst, however, there is cellular differentiation such that recognizable organs may found within the masses. Maternal polyhydraminos has been associated with cervical teratomas as it has been documented in 18% of the obstetric record in mothers of involved children. For cervical teratomas, these generally appear during the first year of life as a midline neck mass. By virtue of their size, especially if present at birth, these lesions can cause respiratory and feeding difficulties. Often, ultrasound will detect these lesions in utero allowing a team to be assembled to secure the airway after birth. The teratoma may then need to be resected once the airway has been secured. While most teratomas are generally considered benign lesions, there is a 20% incidence of malignancy arising in these lesions. Malignant teratomas are very rare in the cervical region (estimated at less the 1%) but much more common in the gonadal and retroperitoneal sites. For the rare malignant cervical lesions, surgical excision followed by multiple drug chemotherapy or radiation therapy is required. Despite the combined modality, the overall prognosis is very poor. Epignathi contain all the germ layers with a marked degree of differentiation. The tissue is so highly developed that it contains full fetal organs, limbs, etc… These lesions are rarely compatible with life.

Thymic Cysts.

Seen rarely as a neck mass, these masses arise from thymic remnants along its course from the mandible to the midline of the neck. The thymus gland is derived from the inferior aspect of the third pharyngeal pouch at the sixth week of fetal life. It descends into the thorax by the ninth week maintaining its relationship with the pericardium. The inferior parathyroid glands are also derived form the third branchial pouch but from the superior aspect. While the thymus gland descends, there remains a connection to the third branchial pouch, referred to as the thymopharyngeal duct. This duct normally degenerates and is completely obliterated by the eight or ninth week. It is the persistence of this duct that explains the presence of cervical thymic cysts. These lesions may be found midline or in the lateral neck reflecting the overall course of
descent near the angle of the mandible to the midline of the neck. Surgical excision is the treatment.

**Thyroglossal Duct Cysts.**

The thyroglossal duct is the most common mass encountered in the midline of the upper neck. These masses present in the midline anywhere along the thyroid's fetal descent from the foramen cecum to the normal location of the thyroid gland. Beginning at the seventeenth day of gestation, the thyroid primordium begins its inferior descent from the foramen cecum, passing anterior, posterior, or through the hyoid bone in its path to the lower neck. It maintains a stalklike connection to the base of the tongue which is gradually resorbed. Any thyroid tissue that remains in along this area has the potential for developing into a thyroglossal duct cyst. The usual presentation is a midline mass that enlarged rapidly after an upper respiratory infection, generally in young children, although they can be found in adults as well. In review of 1316 cases, 31% were under the age of 10, 20.4% were in their 20's, 13.5% were in their 30’s, and 34.6% were older than 40. These masses typically measure between 2 to 4 cm and can be seen to elevate on tongue protrusion. Carcinoma has been found to arise, albeit rarely (<1%) in a thyroglossal duct cysts. Cancer within this lesion has been found to occur more frequently in females than males, generally in the third to sixth decade of life. Typically, the diagnosis is made at the time of pathological examination. The pathologic diagnoses in these cases are papillary adenocarcinoma, follicular adenocarcinoma, mixed papillary and follicular adenocarcinoma, adenocarcinoma, and squamous cell carcinoma.

The preoperative workup of the patient with a presumed thyroglossal duct cyst has been a source of controversy in the literature. The controversy exists whether a study is indicated as well as which study is best to prevent inadvertent post-operative hypothyroidism by removing the only functioning thyroid tissue. It has been estimated that ectopic thyroid tissue presenting as a thyroglossal duct cyst is between 1 – 2%. The options that have been proposed includes: (1) no preoperative studies, (2) no preoperative studies but look for thyroid tissue during surgery, (3) radioisotope scanning of the thyroid tissue on all patients before surgery, (4) other imaging modalities (US namely), and (5) radioisotope screening on those patients with a history suggestive of hypothyroidism, abnormal thyroid function tests, etc. Tunkel et. al. tackled this clinical challenge and presented an elegant algorithm. Signs and symptoms of hypothyroidism are sought in the patient. If normal an ultrasound is recommended to search for normal thyroid tissue. Intraoperatively, the authors suggest palpating or visualizing the thyroid gland, if feasible. For the patient with clinical signs of hypothyroidism, a radioisotope scan is indicated according to the authors.

Treatment is surgical excision, namely through the Sistrunk procedure. This has been the standard surgical approach since the 1920’s as it takes into consideration the knowledge of embryology and developmental anatomy of the thyroglossal duct cyst. The operation includes excision of the cyst in continuity with the midportion of the body of the hyoid and a block of muscle that theoretically includes the thyroglossal duct remnant dissected up to the foramen cecum. Local excision is not advocated because of the high recurrence rate, estimated as high as 40% in some reviews. In comparison, the recurrence rate when following the Sistrunk procedure is between 1 – 4%.
Hemangioma and Vascular Malformations.

The understanding of head and neck vascular anomalies has been hampered by confusing classification schemes. This confusion has been responsible for improper diagnosis, illogical treatment, and misdirected research efforts. Interdisciplinary communication has also been limited due to each medical specialty’s variant of classification. Based on biological behavior, Mulliken and Glowacki in 1982 divided the cervicofacial vascular anomalies into two major groups – hemangioma and vascular malformations. The vascular malformations were then further divided based on histology and velocity of blood flow.

There are certain characteristics that distinguish a hemangioma form a vascular malformation. Vascular malformations are present at birth and grow proportionally with the child; whereas, a hemangioma is generally not at birth, rather undergoes a later stage of rapid growth followed by regression. At a cellular level, the hemangioma, during the proliferative phase, is comprised of rapidly dividing endothelial cells with a significant presence of mast cells. Vascular malformations do not show the evidence of cellular proliferation – rather there is a progressive dilatation of vessels of abnormal mural structure. In regards to surrounding structures, hemangiomas do not typically cause bony or cartilaginous changes. In contrast, slow flowing vascular malformations can cause significant craniofacial distortion from bony hypertrophy, while the high-flow vascular malformation causes bony erosion and destruction.

**Hemangioma.** The hemangioma is the most tumor of infancy. There is an overall incidence of 10 - 12% in premature (1500 – 2500 grams) and full term infants, in contrast to an increased incidence of 23% in low birth infants (less than 1000 grams). There is a female predilection of 3:1. Eighty percent of the lesions are isolated. Laryngotracheal involvement is seen in up to 50% of those children with a head and neck lesion. Most hemangiomas are detected within the first six weeks of life, presenting as either a superficial or a deep lesion. The superficial hemangioma is characterized by a raised skin lesion measuring from .5 cm to 5.0 cm with a vivid red color; whereas, the deep hemangioma has overlying normal skin with a deeply located bluish hue. The deep seated hemangiomas may be difficult to distinguish from a lymphangioma. Diagnosing these lesions is typically based on the clinical presentation; however, radiographic imaging with doppler flow ultrasound, MRI, CT, and angiography are helpful tools in assisting the diagnostic process and mapping the lesion.

Clinically, hemangiomas undergo a proliferative phase in which the lesion rapidly grows for the first 8 - 12 months. This is then followed by a gradual involutional phase over the ensuing years. It has been clinically shown that in over 50% of the children will have resolution of the lesion at the age of 5, increasing to over 70% resolution at the age of 7. There is continued improvement in the remaining children until 10 to 12 years of age. Because of the natural course of resolution without any medical or surgical intervention, the physician should not attempt a course of aggressive treatment. In fact, parental reassurance and periodic photodocumentation is typically all that is required. The fibrofatty residual tissue is all that may need to be excised at a much later date.

There are, however, certain complicating situations associated with hemangiomas that require intervention.
**Visual Obstruction.** Obstruction of vision from a hemangioma (i.e. upper eyelid, cornea) may impair the ability of the child to develop binocular vision. Distortion of the cornea may further result in astigmatic ambylopia.

**Subglottic Hemangioma.** There may be a potentially life threatening lesion that presents after the first six weeks of life. The child typically presents with biphasic stridor but may present in frank respiratory distress secondary to the overwhelming size of the lesion. Direct laryngoscopy is the gold standard for diagnosis which typically shows a soft, easily compressible mass.

**Ulceration and Bleeding.** Local complications of ulceration and recurrent hemorrhage from the lesion may require treatment. Typically, conservative local care is all that is required. Extensive or refractory ulceration may be an indication for a trial of pharmacologic therapy.

**Congestive Heart Failure.** This life threatening complication is usually seen in a child with multiple cutaneous hemangiomas and visceral involvement – typically the liver. High output congestive heart failure may also occur with an isolated, large cervicofacial lesion. Despite multimodality treatment, the overall mortality rate is estimated to be as high as 54%.

**Kasabach-Merritt Syndrome.** This syndrome is characterized by a platelet trapping coagulopathy occurring early in the proliferative stage. The involved skin may become tense, red, and shiny secondary to intrallesional hemorrhage. Hematologic evaluation typically reveals a profound thrombocytopenia (2000 – 40,000/mm³), reduced fibrinogen, and elevated PT / PTT. There is an alarming risk of hemorrhage into the GI tract, peritoneum, pleural space, or central nervous system. The mortality rate has been estimated at 30 – 40% despite aggressive management.

The treatment of complicated hemangiomas falls into three areas – laser ablative, pharmacologic, and surgical excision.

**Corticosteroids.** The decision to use high dose corticosteroids for a complicated case should be made early as there have been clinical reports that young proliferating lesions respond more favorably than the older lesion. Typically, the usual dose is 2 – 3 mg/kg/day of prednisone given orally as there has been no conclusive data showing intravenous is more efficacious than oral. The response to corticosteroids is typically seen early – measured in several hours to a few days. The lesion itself will begin to lighten, soften, and reverse its growth. It has been suggested that a lesion that fails to respond in 7 days been deemed "unresponsive" and further treatment is not indicated. For the responsive lesion, the steroid dose should be tapered slowly over several weeks and kept on a maintenance dose at least until the natural involutional stage has commenced. Intrallesional injections with triamcinolone acetate (40 mg) and betamethasone (6 mg) have been useful for protuberant hemangiomas of the face – upper eyelid and nasal tip. The injections may be spread apart every 4 to 6 weeks. The response rates to systemic and intrallesional corticosteroid use has ranged from 30 – 60%.

**Alpha Interferon.** While not completely elucidated, interferon has prominent antiangiogenic properties and has been recently introduced into the management of difficult hemangiomas. Especially in patients with life threatening complications, such as Kasabach-Merritt syndrome,
has been documented to have a favorable response even in those lesions not responding to corticosteroids.

**Laser Ablation.** In the patient with a subglottic hemangioma, watchful waiting is the typical clinical course. For more symptomatic children, corticosteroids at 2 – 3 mg/kg/d have been shown to have a response rate of approximately 60%. CO2 laser ablation has been advocated in those not responding to steroid therapy. Care must be taken with the rare circumferential lesion – it should be removed in stages to prevent subglottic stenosis. For the eccentric lesion, however, these may usually be removed in one surgical procedure without adverse sequelae.

**Surgical.** Surgical excision is typically reserved for the removal of the fibrofatty tissue that remains after the initial lesion involutes. For obstructing hemangiomas not responsive to pharmacotherapy, however, excision may be required. The surgeon should be cautioned at aiming more toward a subtotal resection rather than violating normal tissue. For children with severe psychosocial distress from the lesion, subtotal / staged excision may also be indicated. These children must be carefully selected.

**Vascular Malformations.** This is a large group of lesions that encompasses capillary, venous, lymphatic, and arteriovenous malformations. To reiterate, in contrast to the hemangioma, these lesions do not undergo spontaneous regression. They are present at birth and grow proportionally with the child. These lesions, however, may expand rapidly secondary to trauma, infection, hormonal changes, or embolic intervention.

The "port wine stain" is the most common vascular malformation. It represents a capillary malformation since it histologically contains a prominent number of dilated capillaries and venule sized vessels in the superficial dermis. The anomaly, while present at birth, attains a very marked purplish appearance and may become nodular with progressive vascular ectasia. The Sturge-Weber syndrome is classic in the finding of capillary hemangiomas along the distribution of the trigeminal nerve (namely V1). In this syndrome, there is concomitant capillary, venous, and arteriovenous malformations of the leptomeninges which contribute to progressive atrophy and degeneration of the cerebral cortex.

The argon laser has gained popularity in treating the capillary hemangioma in adults, especially those with darker skin and a thick lesion. The argon laser produces significant thermal thrombosis of the lesion with later fibrosis, lightening and flattening of the skin. Because of the thermal damage, its use in children and lighter skinned patients has been limited. In contrast, the flashlamp pulsed tunable dye laser may be useful in younger children, in patients with lightly colored, thinner lesions, and in patients with extremely heat sensitive skin. It has less morbidity and less chance of hypopigmentary changes. In some instances, surgical excision may be necessitated with reconstruction using locoregional flaps, skin grafting, or tissue expansion.

Venous malformations were referred to as cavernous hemangiomas or varicose hemangiomas. Histologically, these lesions are characterized by largely dilated, ectatic vascular channels with a normal endothelial lining and areas of thrombosis. There may also be a component of capillary or lymphatic anomalies within the venous malformation. These lesions are commonly found on the lip or cheek within the head and neck region. They soft, compressible, nonpulsatile mass
which refills rapidly after releasing. Venous malformations may also appear as an intramuscular or intraosseous mass. These occur most commonly in the masseter muscle and the mandible. Radiographically, the intraosseous lesions demonstrate a pathognomonic “honeycombing” or “soap bubble” appearance. The low flow nature of these lesions predisposes them to phleboliths which may be the cause of recurrent pain. In the treatment of venous malformations, surgical resection, often subtotal, may be indicated for large or symptomatic lesions, which results in bulk reduction and improved contouring. Vital structures are preserved during surgical resection. For these lesions, good results have been reported with sclerosing agents injected into the epicenter of the lesion. The arterial inflow and venous outflow are occluded prior to injection.

Lymphatic malformations are uncommon congenital lesions, although the head and neck region is the most common site. Classically, the cystic hygroma is the prime example of the lymphatic malformation. Histologically, lymphatic malformations are composed of multiple dilated channels lined by a single layer of endothelium. Often there is a notable hemorrhage within the lymphatic spaces. A classification of lymphatic malformations has been described. Type I lesions are located below the mylohyoid, while type II lesions are located above the mylohyoid. Radiographically, the type I lesions tend to have relatively sharp demarcation of the cystic areas, which appear to be well circumscribed and discreet. In contrast, the type II lesions are iso-dense that are poorly defined and show obscured muscle and fatty planes. Treatment is based on surgical excision, with the degree of success far greater with the type I lesions than that of type II. For the type I lesions, complete resection is usually possible and may be performed within the first 9-12 months of age. In contrast, the type II lesions are approached with reasonable expectations, often requiring multiple procedures and highly unlikely total resection. In general, most authors accept surgical management at 5-6 years of age. In cases where the lesion is not amenable to safe dissection, i.e. the supraglottis and tongue, the CO2 laser has been useful for resection.

High-flow vascular anomalies of the head and neck are characterized by arteriovenous malformations. There is microscopic evidence of multiple arteriovenous shunts with thick walled, irregular caliber arterial vessels with concomitant venous changes consisting of hypertrophy, intimal thickening, and sclerosis secondary to the high flow. Clinically, these lesions are brightly erythematous lesions of the skin with an associated thrill to palpation and bruit to auscultation. These lesions, as most vascular malformations, may change suddenly in response to minimal trauma, infection, or hormonal changes. Clinically, these lesions may cause problems with skin necrosis and craniofacial destruction secondary to shunting of the blood from these structures. In addition, there is a significant compressive and erosive effect. Because of these high-flowing lesions, heart failure may be a coincident morbidity. For the symptomatic lesion, pre-operative evaluation of the lesion with MRI should be undertaken along with angiography which allows superselective embolization of the lesion, if so indicated. Embolization does not reduce the extent of the resection, rather assists in controlling intra-operative blood loss. Long term success, unfortunately, depends on total resection of the involved tissue with often complex reconstruction, i.e. microvascular tissue transfer.

**Esophageal Duplication Cysts.** Although rare, the esophageal duplication cyst may present as a midline neck mass. The esophageal duplication is felt to occur because of disturbed recanalization with abnormal foregut rests forming cysts. The lining of the duplication cyst may
be squamous, respiratory, intestinal or mixed with the secretory epithelium causing gradual enlargement of the cyst. Additionally, gastric acid secreting mucosa, which is often found in these structures, may produce severe complications if ulceration and hemorrhage occur. The predominating presenting symptoms are generally stridor, recurrent pneumonia, or cough. The neonate, however, may present with frank respiratory distress secondary to obstruction from the mass. For the infrequent person presenting as a neck mass, it is typically near the midline and may reach immense proportions, depending on the activity of the secretory mucosa. Interestingly, these cysts do not communicate with the esophagus but may communicate with the vertebral column. In evaluating these lesions, the CT scan is invaluable in observing the lesion. The literature indicates that endoscopic and barium swallow studies have limitations in the diagnosis and preoperative planning of the esophageal duplication cyst. Endoscopically, a submucosal mass with normal overlying mucosa is typically found, making it difficult to distinguish from other lesions, such as a leiomyoma. This is similar for the barium swallow. While technically difficult, endosonography has been used to assist in the diagnosis. For the rare lesion that communicates with the spinal canal, myelography is typically performed. Treatment is surgical excision. Depending on the location of the cyst, the approach may require a cervical, thoracotomy or laparotomy for appropriate removal. For the neurenteric communication, these must be carefully dissected and closed with neurosurgical assistance.

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