The evaluation and management of pediatric neck masses requires extensive knowledge of the anatomy and embryology of the head and neck, and pathogenesis of the specific disorders. Congenital masses, inflammatory and infectious lesions, benign neoplasms and vascular malformations may be encountered. Fortunately, malignant neck lesions in children are rare. A thorough history and physical examination along with a focused and coordinated work up are essential in the management of pediatric neck masses.

**Embryology and Anatomy**

During the first few weeks of development, the pharynx occupies the bulk of the foregut. As the buccopharyngeal membrane breaks down late in the third week, the mesodermal branchial bars begin to appear. These appear as u-shaped structures in the transverse plane, which fuse ventrally to form 6 pairs of pharyngeal arches. The fifth arch is small and quickly disappears. The arches are separated externally by ectodermal pharyngeal clefts and internally by endodermal pharyngeal pouches. Each arch contains an artery, a nerve, and muscular and cartilaginous structures, which give rise to most of the structures of the neck. The remaining cervical musculature gains contributions from cervical somites.

The first branchial arch contains cartilage with a dorsal portion, known as the maxillary process, and a ventral portion, known as Meckel’s cartilage or the mandibular process. Both of these structures eventually regress to leave only the malleus and incus. The ossification of mesoderm around Meckel’s cartilage gives rise to the mandible and the sphenomandibular and anterior mallear ligaments. The muscles of the first arch include the muscles of mastication (temporalis, masseter, and pterygoids), the tensor tympani and tensor veli palatini, the anterior belly of digastric and mylohyoid muscles. The nerve of the first branchial arch is the fifth cranial
nerve. Contributions of the first arch artery are thought to persist as the maxillary artery. The first pouch persists as the Eustachian tube, the middle ear and portions of the mastoid bone.

The second branchial arch cartilage is known as Reichert’s cartilage and its derivatives include the upper body and lesser cornu of the hyoid bone, the styloid process and stylohyoid ligament, and the stapes superstructure. Muscles of the second arch include the muscles of facial expression, platysma, posterior belly of digastric, stylohyoid, and stapedius. The nerve of the second arch is the seventh cranial nerve. Only very rarely does the artery of the second arch persist, as the stapedial artery, and courses through the crura of the stapes.

The derivatives of the third branchial arch include the greater cornu and inferior body of the hyoid. Muscles of the third arch include the stylopharyngeus and superior and middle pharyngeal constrictors. The artery contributes to the common carotid and proximal portions of the internal and external carotid arteries. The nerve of the third arch is the ninth cranial nerve. The third pouch contributes to the inferior parathyroids and thymus and thymic duct.

The fourth and sixth branchial arches fuse to form the laryngeal cartilages. The muscles of the fourth arch include the cricothyroid and inferior pharyngeal constrictors. The muscles of the sixth arch are the remaining intrinsic musculature of the larynx. The nerve of the fourth arch is the superior laryngeal nerve and the nerve of the sixth arch is the recurrent laryngeal nerve. On the right, the artery of the fourth arch contributes to the subclavian and to the aortic arch on the left. The sixth arch artery becomes the ductus arteriosus and pulmonary artery. The endodermal pouch of the fourth arch becomes the superior parathyroids and possibly the parafollicular thyroid cells.

As the branchial apparatus develops, the first and second arches grow in a cranial to caudal fashion, creating the epipericardial ridge. The epipericardial ridge contains the mesodermal rudiments of the sternocleidomastoid, trapezius, and the infrahyoid and lingual musculature. The nerves of the epipericardial ridge are the hypoglossal and spinal accessory. The proliferation of mesoderm in this area eventually causes overgrowth and narrowing of the third, fourth, and sixth arches into an ectodermal pit, known as the cervical sinus of His. This sinus eventually becomes obliterated. If it fails to be obliterated or is partially obliterated, a branchial sinus, cleft, or cyst of types II, III, or IV may develop.

The thyroid gland originates during the fourth week of gestation as endoderm of the floor of the mouth in between the tuberculum impar and copula of the first and second pouches. It descends as a bilobed diverticulum from the foramen cecum to pass in variable position to the hyoid bone to rest in the lower neck.

**Congenital Masses**

**Branchial Anomalies**

Developmental aberrations of the intricate branchial system give rise to branchial anomalies. These anomalies may exist as either cysts, sinuses, or fistulae. Generally, branchial clefts with external openings onto the skin are associated with the first and second arches, whereas the third and fourth arches are associated with internal openings.
First Branchial Cleft Cysts are classified as Type I or Type II. Type I is an ectodermal duplication anomaly of the external auditory canal. The cyst is lined by squamous epithelium without skin appendages. These lesions track parallel to the EAC and middle ear and often begin as fistulous tracts at the post auricular or pretragal area. Surgical excision in the noninfected state is the treatment of choice.

In Type II Branchial cleft cysts, the cyst or external opening is localized in the anterior neck, always superior to the hyoid bone. They have been traditionally found in a triangle formed from the EAC curving to meet the mid-hyoid and tip of the chin. The tract courses over the angle of the mandible, through the parotid gland, and terminates near the bony-cartilaginous junction of the EAC. The course of the tract in relation to the facial nerve is variable. 29% may pass medial to the nerve, and even may sometimes split around the nerve. Treatment of these lesions is surgical. Preoperative imaging with MRI to evaluate the lesion in relation to the parotid gland is helpful. The definitive excision requires identification of the facial nerve and a superficial parotidectomy is typically required to avoid inadvertent injury to the small and more superficially located facial nerve. Special care must also be taken at the upper end of the fistula in order to remove not only the portion of the tract between the skin and cartilage of the EAC, but also the flange of cartilage through which the tract passes.

Second Branchial Cleft cysts are the most common (90%) branchial anomalies. They usually present spontaneously as a painless, fluctuant mass of the anterior triangle in infants. These lesions may fluctuate after a URI. The tract courses deep to the second arch derivatives and superficial to the third arch derivatives. If an external opening is present, it is most consistently found at the anterior border of the SCM at the middle and inferior two-thirds junction. It then courses anterior to the SCM, superficial to the IX, XII nerve, to turn medial and pass between the internal and external carotid artery. The tracts typically terminate into the tonsillar fossa. Preoperative CT imaging will help delineate the pathway. The tract is surgically excised and may require a tonsillectomy.

Third Branchial Cleft Cysts are rare (<2%). They may present in a similar fashion as Second BCC. However, the internal opening is located within the pyriform sinus. The tract pierces the thyrohyoid membrane cephalad to the superior laryngeal nerve, lateral to the hypoglossal nerve, medial to the glossopharyngeal nerve, and posterior to the internal carotid artery. Surgical approach is usually via a standard thyroideectomy approach to fully visualize the recurrent laryngeal nerves and to adequately distinguish between a third and fourth BCC. Often, a partial thyroid lobectomy is required.

Fourth Branchial Cleft Cysts are extremely rare and a true fistula has never been reported. The potential path would lie between the fourth and sixth arch structures. The internal sinus opening would originate near the apex of the pyriform sinus, caudal to the superior laryngeal nerve and travel translaryngeal under the thyroid ala to emerge near the cricothyroid joint, and then descend superficially to the recurrent laryngeal nerve in the paratracheal region. Third and fourth branchial pouch anomalies have been recognized as a potential cause of acute suppurative thyroïditis in children. They present with fever, paratracheal fullness, and tenderness, typically on the left side. In contrast to hematogenous suppurative thyroiditis, the intraoperative cultures show a variety of aerobic and anaerobic organisms of pharyngeal origin.
For patients that fail antibiotic therapy, a hemithyroidectomy is recommended only after radiologic or endoscopic proof of a pyriform sinus opening is observed.

**Thyroglossal duct cyst**

The most common midline mass encountered is the thyroglossal duct cyst. As the bilobed thyroid diverticulum descends from the floor of the pharynx, a thyroglossal duct is created. Failure of involution of any portion of this tract can lead to a thyroglossal duct cyst. Arrest of normal descent of the gland can result in ectopic thyroid tissue. The pyramidal lobe, found in 40% of patients, is actually just the failure of closure of the inferior most portion of the thyroglossal duct. The majority of thyroglossal duct cysts present in children and adolescents as an asymptomatic midline mass at or below the hyoid bone. They commonly elevate with tongue protrusion. External sinuses involving the pharynx may rarely occur, and these may become infected and even fistulize. The preoperative workup of thyroglossal duct cysts has been a source of controversy. Some recommend checking overall thyroid function with a TSH level prior to excision. An ultrasound is probably the best study to document the presence of normal thyroid tissue. A thyroid scan is of value in patients who are hypothyroid, are unable to tolerate an adequate thyroid ultrasound, or fail to demonstrate a normal thyroid by ultrasound.

The primary management of thyroglossal duct cysts is with surgical excision. However, simple cyst excision results in a high rate of recurrence (50%). This procedure has been largely replace by the ‘Sistrunk procedure.’ During this procedure, the central portion of the hyoid is included in the en bloc excision of the cyst, and dissection is carried up into the tongue base. Recurrence rates with this procedure dropped to 4-6%. Patients who are at greatest risk for recurrence are those who have had recurrent infections, externally draining sinuses, or prior incision and drainage. For these patients, a modification of the Sistrunk procedure to include en bloc anterior dissection including portions of the strap muscles is advocated to reduce recurrence.

**Thymus gland anomalies**

The thymus gland originates from the third branchial pouch and descends into the lower neck and mediastinum. Ectopic thymus tissue can result from failure of descent of one or both lobes of the thymus. Failure of involution of thymopharyngeal ducts may persist as cervical thymic cysts. These are firm, mobile masses usually found in the lower aspect of the neck. The lesions usually present during the first decade of life due to the fact that the thymus tissue is of greatest relative size at birth and reaches greatest absolute size by puberty. Initial workup includes a plain chest radiograph and possibly a CT. Surgical excision is the treatment of choice.

**Dermoid and Teratoid Cysts**

Teratoid and Dermoid cysts are rare causes of neck masses. These developmental anomalies are composed of different germ cell layers. They are thought to possibly arise from isolation of pleuripotent stem cells during migration with resultant disorganized growth or from entrapment of germ cell layers at points of failed embryonic fusion lines. The lesions are classified according to their composition. Dermoid cysts are composed of mesoderm and ectoderm and may contain hair follicles, sebaceous glands, and sweat glands. They are often midline or paramedian, painless, and do not elevate with tongue protrusion. When presenting as
midline masses, they are frequently misdiagnosed as thyroglossal duct cysts even intraoperatively, as some may involve the hyoid bone. The treatment is simple excision in contrast to the Sistrunk procedure for thyroglossal duct cyst. Some have recommended intraoperative cyst aspiration or sectioning as a means of differentiating these two lesions.

Teratoid cysts and Teratomas contain all three germ layers. These lesions commonly present within the first year of life and are usually larger midline or paramedian masses than Dermoid cysts. Teratomas are distinguished by cellular differentiation enough to have recognizable organs or structures. The larger size of these lesions results in secondary aerodigestive compressive symptoms. There is an associated 20% incidence of maternal polyhydramnios which can often lead to the diagnosis with maternal ultrasound. In cases in which the lesion is anticipated to be large enough to cause immediate airway compromise during birth, the appropriate teams can be available to secure the airway. The well encapsulated cyst and poor vascularization found in these lesions enhance the likelihood of successful excision.

**Laryngoceles**

Laryngoceles are most frequently seen in the adult population, but can present as a lateral neck mass in the pediatric population. They are thought to form congenitally as a result of enlargement of the laryngeal saccule with distension and entrapment of air. Laryngoceles are classified as either internal, external, or combined. The internal laryngcele is confined solely to the larynx and is caused from distension of the saccule, typically to the false vocal cord and aryepiglottic fold. They typically present with hoarseness and respiratory distress and not as neck masses. The external laryngcele will protrude through the thyrohyoid ligament at the entrance of the superior laryngeal nerve. These lesions will present as a soft, compressible, lateral neck mass that may distend with increases in intralaryngeal pressures. If infected they are classified as laryngopyoceles. The combined type has features of both internal and external. CT scan will delineate the lesions. Asymptomatic laryngoceles in children require no surgical treatment. Symptomatic laryngoceles and laryngopyoceles should be excised. Internal laryngoceles may be excised or marsupialized endoscopically. For external laryngoceles without a significant internal component, simple excision after identification of the superior laryngeal nerve is appropriate. For a combined type, a lateral thyrotomy may be needed for more complete exposure.

**Vascular and Lymphatic malformations**

Pediatric vascular lesions of the neck can be classified as either hemangiomas or vascular malformations. Hemangiomas are the most common tumor of infancy and typically present within the first few months of life. They demonstrate a rapid growth and then a period a quiescence followed by a period of involution. 50% of children will have complete resolution of the lesion by the age of 5, increasing to 70% resolution by the age of 7. During the proliferative phase, the hemangioma is comprised of rapidly dividing endothelial cells with a significant presence of mast cells. Hemangiomas rarely cause bony or cartilaginous structure changes. CT with contrast or MRI with gadolinium will demonstrate the anatomy and vascularity of the lesion. Because of the natural course of involution, parental reassurance and periodic photo documentation are typically all that are required. After involution the skin exhibits mild atrophy, and surgical excision of this excessive tissue may be desired. Cavernous hemangiomas are
usually larger and are associated with deeper tissue involvement which makes them less likely to spontaneously resolve. Subglottic hemangiomas may be associated with cutaneous hemangiomas. It is imperative to perform an airway evaluation on any patient with cutaneous vascular lesions associated and stridor.

For hemangiomas which cause complications, high dose steroids can be used. They are most beneficial in young proliferating lesions. The usual dose is 2-3mg/kg/day of oral prednisone. Lesions that show no response in 7 days are not likely to respond, and therapy should be discontinued due to the side effects of long term steroid use. Local intralesional injections with triamcinolone and betamethasone may also be used for more focal lesions. The response to systemic and intralesional corticosteroid use has ranged from 30 to 60%. Surgical excision may be required for these complicated hemangiomas or those that are non-responsive to steroids. Subtotal or staged excision may be the best approach.

Vascular malformations are present during birth and grow proportionately with the child. They show progressive dilation of the venous structures. Vascular malformations can cause significant bony distortion and destruction. The Port wine stain is the most common vascular malformation. It is classified as a capillary type vascular malformation. Arteriovenous malformations are clinically distinguishable from other head and neck masses because they are pulsatile on palpation and may be associated with an audible bruit.

Lymphangiomas are lymphatic cysts that are isolated from their normal route of drainage into the venous system. The embryological development of the lymphatic system is theorized by the centrifugal theory and the centripetal theory. The centrifugal theory states that lymphatic channels grow outward from venous channels. The centripetal theory states that lymphatic channels grow independently of venous channels. Regardless of the theory, the lymphatic cysts become either totally or partially isolated from the venous system. Histologically, they are classified into capillary, cavernous, and cystic types based on the size of the lymphatic spaces. Cystic hygromas are large, soft, painless, and compressible masses that usually present by the age of 3. These lesions most commonly present in the posterior triangle where mass effect is limited. When they present in the anterior triangles, airway obstruction is more of a concern. CT is of greatest diagnostic value. Spontaneous regression is rare and surgical excision for obstructing lesions is the treatment of choice. Recurrence rates are generally high because of the poor encapsulation and dissection planes.

The larger size and obstructive potential of lymphangiomas and teratomas may lead to CHAOS (Congenital High Airway Obstruction Syndrome). Emergent airway management at the time of delivery is key for survival. High resolution fetal ultrasound may show flattened diaphragms, polyhydramnios, and a cervical mass. Appropriate team members include a maternal-fetal medicine specialist, neonatologist, anesthesiologist and an otolaryngologist. Management involves elective Caeserean section with establishment of the airway while still on placental oxygenation.
Other Benign Lesions

Plunging Ranula

A ranula is either a mucus retention cyst or a mucus extravasation pseudocyst arising from an obstructed sublingual gland. A simple ranula is confined to the oral cavity as a cystic unilateral mass of the floor of the mouth. A plunging ranula may pierce the mylohyoid and present as a paramedian or lateral neck mass with or without an obvious oral cavity ranula. They should be differentiated from Dermoid cysts and lymphangiomas. Cyst aspiration consistently reveals fluid with high levels of protein and salivary amylase. CT or MRI will demonstrate a uniloculated cystic mass arising from the sublingual space with extension into the submental, submandibular, or parapharyngeal space. Marsupialization has led to a high rate of recurrence of these lesions. Surgical treatment includes excision in continuity with the sublingual gland of origin.

Sternomastoid Tumor of Infancy

Sternomastoid tumor of infancy, also known as pseudotumor of infancy, is a fibrotic lesion of the distal sternocleidomastoid. These lesions usually present within the first 7 to 28 days of birth as a firm mass within the SCM. The patient may exhibit a head tuck to the ipsilateral side and chin pointed away from the lesion. An association with breach delivery and forceps delivery has been reported. The etiology is still unknown but it is theorized that hematoma formation in the SCM during traumatic delivery is replaced with subsequent fibrosis. Usually, aggressive physical therapy and range of motion exercises will resolve the lesion, with 80-100% regressing by the first birthday. Only rarely does an untreated case lead to long term torticollis and resultant dysmorphosis. Ultrasound will reveal a solid mass that is intrinsic to the SCM. Surgical division of the SCM is only recommended if physical therapy fails.

Infectious and Inflammatory lesions

Infection and inflammation of the cervical lymph nodes is the most common cause of pediatric neck masses. Palpable cervical lymph nodes are present in 40% of infants. Approximately 55% of pediatric patients have palpable lymph nodes that are not necessarily associated with an underlying systemic infection. Cervical nodes that are asymptomatic and <1cm in diameter may be considered normal in children under 12. The most common site of cervical adenitis is the submandibular or deep cervical nodes. Lymphadenitis may be of viral, bacterial, fungal, parasitic, or noninfectious etiology.

The most common bacterial cause of cervical adenitis is Staphylococcus aureus and Group A Streptococci. The cervical lymphadenopathy is usually tender and the patient may have associated symptoms of malaise and fever. The majority of the cases will respond within 4-6 weeks to a ten day course of a beta-lactamase resistant antibiotic. If there is failure to respond, a fine needle aspiration is warranted to exclude other diagnoses. Node suppuration may occur, and needle aspiration or incision and drainage of the necrotic lymph node may be required.

The parapharyngeal and retropharyngeal spaces are the most common neck spaces to be involved in pediatric neck abscesses. The infections are usually polymicrobial, including aerobic
organisms such as Staphylococcus, Streptococcus, Niesseria, and anaerobes such as Fusobacterium, Peptostreptococcus, and Bacteroides. CT scanning with contrast has greatly facilitated the diagnosis of deep neck space abscesses. It allows for precise anatomical localization and relation to such structures as the major cervical vessels can be assessed preoperatively. Surgical drainage may be either intraoral or external and depends on the location of the abscess. Abscesses that are confined within a capsule and that do not extend posterior or lateral to the great vessels may be safely drained via a transoral approach.

Lemierre’s syndrome is a septic thrombophlebitis of the internal jugular vein. It was classically seen as a complication of inadequately treated tonsillitis, but may also be seen with neck abscesses. Patients present with spiking fevers and fullness to one side of the neck. Treatment is with intravenous antibiotics. The use of anticoagulants is still controversial. Patients that demonstrate recurrent septic emboli may require surgical ligation or excision of the internal jugular vein.

Tuberculous and Nontuberculous mycobacteria (NTM) are other common causes of cervical adenitis (scrofula). In children, NTM is much more common than tuberculous mycobacteria. Patients with mycobacterial infection classically have a single large cervical lymph node. Later in the disease, the overlying skin may turn a characteristic violaceous color. For tuberculous adenitis a CXR is needed to rule out active pulmonary disease. A PPD is usually strongly reactive. Treatment usually is the same as for pulmonary TB with combinations of isoniazid, ethambutol, streptomycin, and rifampin.

NTM patients rarely exhibit fever or systemic symptoms. The CXR is usually normal, and PPD reactions are normal or only intermediate in reactivity. The treatment is less definitive than for that of tuberculous mycobacteria. NTM is notoriously resistant to traditional anti-tuberculous agents alone and requires a combination of typical and atypical agents. Unless total surgical excision can be obtained, chronic draining sinuses often develop and persist until spontaneous resolution occurs.

Cat Scratch Disease (CSD) is now known to be caused by Bartonella henselae. This bacteria can be cultured from 30-60% of cats. It can be transmitted to humans by a scratch. The clinical symptoms are self limited, unilateral, regional lymphadenopathy. Patients may also have low grade fevers and malaise. CSD is diagnosed by the Warthin-Starry stain which shows small pleomorphic gram-negative rods. Cultures may take several weeks. Serologic testing of IgM and PCR of Bartonella DNA are showing some promise and may help to reduce the need for open biopsy. The symptoms usually resolve over several weeks to months, but in 10% of the patients, the nodes may progress to suppuration and require incision and drainage. Antibiotic therapy has been unrewarding, but some anecdotal evidence has suggested benefit from macrolides and aminoglycosides.

Viral adenitis is likely the most common infectious process in the neck. Common pathogens include rhinovirus, adenovirus, and enterovirus. These conditions are usually self-limited and are associated with upper respiratory symptomatology. Infectious Mononucleosis is caused by Ebstein Barr Virus (EBV). It causes an exudative, almost necrotic tonsillitis and impressive cervical lymphadenopathy and sometimes hepatosplenomegaly. Heterophile antibodies (monospot) help confirm the diagnosis, but may remain negative early in the disease.
Titers of IgM and IgG are most specific. Treatment is usually supportive. In cases in which adenotonsillitis is so severe that airway symptoms emerge, steroid and antibiotic therapy may be necessary. Ampicillin and Amoxicillin have been associated with a rash in 90% of EBV patients and should be avoided. CMV and HIV cervical lymphadenitis present with similar symptoms to EBV. Treatments for the cervical lymphadenopathy are supportive as well.

Kawasaki syndrome is a multisystem vasculitis of unknown etiology occurring in young children under the age of 5. Diagnosis is clinical and requires five of six criteria including: fever greater than 5 days, conjunctival injection, reddening, or injection of the oral cavity, reddening or desquamation of the palms and soles, polymorphous erythematous rash, and cervical lymphadenopathy. The vasculitis is self limited but may cause permanent cardiac damage as a result of coronary artery aneurysms in about 20% of patients that are untreated. High dose aspirin is used in the acute phase, and intravenous immunoglobulin therapy reduces coronary aneurysm formation.

The diagnosis of pediatric neck masses can be quite challenging. Clinical features including the age of the patient, location and mobility of the mass, time course, and associated constitutional or head and neck symptoms can help narrow the differential diagnosis. Though most cases can be diagnosed with a thorough history and physical examination, imaging, serology, or biopsy may be necessary in selected cases.

**BIBLIOGRAPHY**


