Imaging the Neck in Children

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Abstract

Congenital anomalies are important to consider in the differential of head and neck masses in children. These lesions can present as palpable cystic masses, draining sinuses, or fistulae. Thyroglossal duct cysts are most common, followed by branchial cleft anomalies and dermoid cysts. Lymphatic malformations are typically found in the posterior neck and may not respect the fascial planes. Other vascular malformations can be differentiated based on their US, MRI, and CT features. Hemangiomas are by far the most common pediatric neck neoplasms, with teratomas and nerve sheath tumors as the other common benign masses. Head and neck malignancy is rare in children, with lymphomas, rhabdomyosarcomas, and carcinomas occurring in decreasing frequency. CT and MR imaging are excellent modalities for detection of deep-space infections; however they are not very reliable for the distinction between abscesses and inflammatory mass (cellulitis). The pediatric airway is primarily assessed by endoscopy, while imaging may offer additional information when needed, including virtual endoscopy. External and internal laryngotracheal trauma is best assessed with CT, which plays an increasingly important role in patient management. Dedicated lower-dose pediatric CT protocols and newer low-dose scanners are valuable in minimizing radiation exposure.

Practical Embryology of the Neck

The anatomic structures of the neck are predominantly derived from the branchial (pharyngeal) apparatus, which consists of paired arches, clefts, pouches, and membranes (Fig. 1). The branchial arches are the embryologic precursors of the ear and the muscles, blood vessels, bones, cartilage, and mucosal lining of the face, neck, and pharynx. Five pairs of arches form on either side of the pharyngeal foregut, in a craniocaudal sequence, from day 22 to 29 of the embryological development; they correspond to the primitive vertebrate gill bars (branchial arches) 1, 2, 3, 4, and 6. Arch 5 either never forms or promptly degenerates in humans. Each arch is composed of a central core of mesoderm, lined externally by ectoderm and internally by endoderm, and contains a central cartilage, muscle element, an arch-specific cranial nerve that innervates the muscle, and an aortic arch artery.

The surface ectoderm invaginates between the arches, creating four pairs of branchial clefts (or grooves), which are apposed by branchial pouches, formed by foregut endoderm evaginations. Therefore, the first and the second pharyngeal arches are separated by the first clefts and pouches, the second and the third arches by the second clefts and pouches, and so on. In fish, this process leads to gill slits, which never become perforated in humans. Instead, thin branchial membranes remain at the junction of each pharyngeal cleft and pouch. These membranes consist of three layers, with mesoderm separating the ectoderm from the endoderm (Sadler 2012; Grevelle and Tucker 2010).

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The first pharyngeal cleft gives rise to the epithelial lining of the external auditory meatus, and the tympanic membrane develops from the first pharyngeal membrane. All the other clefts and membranes become obliterated. The adult derivatives of branchial arches and pouches are summarized in Table 1.

![Fig. 1 Schematic sagittal section of the head and neck region of a 4-week embryo shows the pharyngeal pouches developing in between the branchial arches](image)

**Table 1 Derivatives of branchial arch components**

<table>
<thead>
<tr>
<th>Arch</th>
<th>Skeletal structures</th>
<th>Ligaments</th>
<th>Muscles</th>
<th>Nerves</th>
<th>Vessels</th>
</tr>
</thead>
<tbody>
<tr>
<td>First (mandibular)</td>
<td>Upper portions of the malleus and incus</td>
<td>Anterior ligament of the malleus, sphenomandibular ligament</td>
<td>Muscles of mastication, mylohyoid, anterior belly of the digastric muscle, tensor tympani, tensor palati</td>
<td>Trigeminal (maxillary and mandibular divisions)</td>
<td>Terminal branch of maxillary artery</td>
</tr>
<tr>
<td>Second (hyoid)</td>
<td>Lower portion of the malleus and incus, stapes crura, styloid process of the temporal bone, lesser horns, and upper part of the body of hyoid bone</td>
<td>Stylohyoid ligament</td>
<td>Muscles of facial expression, stapedius, stylohyoid, posterior belly of the digastric muscle</td>
<td>Facial</td>
<td>Stapedial artery</td>
</tr>
<tr>
<td>Third</td>
<td>Greater horns and lower part of body of hyoid bone</td>
<td>None</td>
<td>Stylopharyngeal</td>
<td>Glossopharyngeal</td>
<td>Common carotid artery, root of internal carotid</td>
</tr>
<tr>
<td>Fourth, fifth, and sixth</td>
<td>Thyroid cartilage, arytenoid cartilage, corniculate cartilage, cuneiform cartilage, cricoid cartilage</td>
<td>None</td>
<td>Pharyngeal and laryngeal muscles</td>
<td>Vagus (superior laryngeal and recurrent laryngeal branches)</td>
<td>Aortic arch, right subclavian, origin sprouts of pulmonary arteries, ductus arteriosus, roots of definitive pulmonary arteries</td>
</tr>
</tbody>
</table>

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First Branchial (Pharyngeal) Apparatus
The first pharyngeal arch divides into cranial maxillary and caudal mandibular swellings, which give rise to the upper and lower jaw, respectively. The cartilaginous core of the maxillary division is called palatopterygoquadrate bar, and that of the mandibular division is known as Meckel’s cartilage. The maxillary cartilage forms the incus and a small bone in the lateral orbital wall, while the mandibular cartilage gives rise to the malleus, anterior malleolar and sphenomandibular ligaments, and a small portion of the mandible. The maxilla, zygoma, temporal squama, and most of the mandible are all membranous bones, formed by direct ossification of dermal mesenchyme. Derivatives of the first branchial arch include the muscles of mastication, tensor veli palatini, tensor tympani, anterior belly of the digastric, and mylohyoid, all innervated by the mandibular division of the trigeminal nerve. The first pharyngeal pouch is the precursor to the auditory tube and tympanic cavity (Sadler 2012; Grevellec and Tucker 2010).

The basic structure of the face is created starting from the fourth gestational week by development and fusion of five prominences: the maxillary and mandibular swellings of the first arch plus an unpaired frontonasal process, on which paired lateral nasal placodes develop. Each nasal placode is then divided into medial and lateral nasal processes by the nasal pits, with a nasolacrimal groove being formed between the maxillary swellings and the lateral nasal processes. The inferior tips of the medial nasal processes fuse into the intermaxillary process, whose posterior extension becomes the primary palate. Posterior to the primary palate, the maxillary processes produce medial extensions called palatine shelves, which fuse with each other during the ninth gestational week, creating the secondary palate. The definitive palate is formed following fusion of the primary and secondary palates at the incisive foramen. Congenital facial defects, such as cleft lip and cleft palate, result from incorrect fusion of these different prominences (Sadler 2012; Grevellec and Tucker 2010).

The first arch also gives rise to the median tongue bud and to adjacent lateral lingual swellings, which form the mucosa of the anterior two thirds of the tongue. The intrinsic tongue muscles and the styloglossus, hyoglossus, and genioglossus are not derivatives of the pharyngeal apparatus, but arise from myoblasts that migrated from the occipital somites. The parotid gland develops from an ectodermal invagination in the crease between the maxillary and mandibular swellings. The blind dorsal end forms the gland, while the anterior extension becomes the duct. Endodermal invaginations give rise to the submandibular and sublingual glands in a similar fashion (Sadler 2012; Grevellec and Tucker 2010).

Second Branchial (Pharyngeal) Apparatus
The cartilage of the second pharyngeal arch, also called Reichert’s cartilage, forms the stapes, the styloid process, the stylohyoid ligament, and the superior portion of the hyoid bone with the lesser horns. The muscles derived from the mesoderm of the second arch are innervated by the facial nerve and include the posterior belly of the digastic, stylohyoid, stapedius, and the muscles of facial expression. With continued growth, the rapidly expanding second branchial arch extends caudally and overgrows the second, third, and fourth branchial clefts, enclosing them in a transient lateral cervical sinus, which soon completely disappears (Sadler 2012; Grevellec and Tucker 2010). The epithelium of the second pharyngeal pouch forms endodermal buds, which are precursors of the palatine tonsils. Later, the cells in the center of the buds die and slough, creating tonsillar crypts that are then rapidly infiltrated by lymphoid tissue.

Persistent remnants of the cervical sinus and second pharyngeal cleft are the precursors of a variety of anomalies associated with the second pharyngeal apparatus. During the second gestational month, the mandible and associated muscles migrate ventrally and inferiorly, while the pinna migrates dorsolaterally. This migration allows anomalous cell rests or unobliterated sinuses to assume the characteristic morphology and position that are associated with branchial anomalies (Sadler 2012; Grevellec and Tucker 2010).
Third Branchial (Pharyngeal) Apparatus

The cartilage of the third branchial arch forms the lower body and greater horns of the hyoid bone. The musculature derived from the third pharyngeal arch is limited to the stylopharyngeus muscle, the only muscle supplied by the glossopharyngeal nerve (Sadler 2012; Grevellec and Tucker 2010). The mucosa covering the posterior third of the tongue (the base of tongue) is a derivative of the hypopharyngeal eminence, a median structure arising from the third and fourth arches. The boundary between the first and third arch contributions, that is, between the anterior two thirds (oral tongue) and the posterior third (tongue base), is called terminal sulcus. The foramen cecum is a depression at the intersection of the terminal sulcus with the median sulcus, a midline groove in the oral tongue.

The thyroid gland develops from the thyroid diverticulum, a small amount of endoderm at the apex of the foramen cecum. The thyroid diverticulum migrates caudally at the end of a slender thyroglossal duct. The duct breaks down and the thyroid continues to descend to its normal adult anatomical position. A portion of the duct may occasionally persist as a thyroglossal cyst, or a fragment of the gland may detach during the migration, forming ectopic thyroid tissue that may be located anywhere along the tract (Sadler 2012; Grevellec and Tucker 2010).

At the ventral end of the third pouch, thymic primordia arise forming hollow tubes, which transform into branching cords. The cords lose their connection with the pharynx and descend to their definite location along the thymopharyngeal duct, where they fuse to form the bilobed thymus. In the dorsal portion of the third pouch, the inferior parathyroid glands are formed. They also detach from the pharynx and migrate inferiorly and medially to rest on the posterior inferior aspect of the thyroid lobes. Occasionally, the inferior parathyroid glands will migrate further inferiorly and may be found within the thorax in close proximity to the thymus (Sadler 2012; Grevellec and Tucker 2010).

Fourth Through Sixth Branchial (Pharyngeal) Apparatus

The fourth through sixth pharyngeal arches form the various laryngeal cartilages and muscles. The arytenoid, corniculate, cuneiform, and thyroid cartilages are derivatives of the fourth pharyngeal arch, whereas the cricoid cartilage arises from the sixth pharyngeal arch. The development of the larynx begins in the fifth week as the arytenoid swellings form in the sixth arch. The arytenoids also chondrify first, followed by the other cartilages during the seventh week. The epiglottis develops from the fourth pharyngeal arch, and its midline swelling, the hypobranchial eminence, gives rise to a small portion of the tongue base. The epiglottal cartilages chondrify in the fifth month, much later than the other laryngeal cartilages, and their origin is still not clear (Sadler 2012; Grevellec and Tucker 2010). The cricothyroid muscle, levator veli palatini, and muscles of the pharynx (except the stylopharyngeus) arise from the fourth pharyngeal arch and are thus supplied by the superior laryngeal nerve. The intrinsic laryngeal muscles are derived from the sixth pharyngeal arch and supplied by the recurrent laryngeal nerve (Sadler 2012; Grevellec and Tucker 2010).

The dorsal portion of the fourth pouch develops into the superior parathyroid glands, which migrate caudally and medially to rest along the dorsal surface of the thyroid gland, slightly superior to the pair derived from the third pouch. The parathyroid glands, thus, switch positions during their descent, and their names reflect the final relative locations. Just caudal to the fourth pouch, a small invagination appears during the fifth week of development, considered by many authors as a fifth pharyngeal pouch (Sadler 2012). Cells that form the ultimobranchial body, which immediately migrates to fuse with the dorsal wall of the thyroid gland, populate it. This structure gives rise to the parafollicular cells (C cells) that produce calcitonin (Sadler 2012; Grevellec and Tucker 2010).
Trachea

The laryngotracheal diverticulum (or lung bud) forms on the ventral wall of the pharynx, caudal to the fourth pharyngeal pouch, on day 22 of the embryological development. This precursor for the trachea and bronchi then continues to grow ventrocaudally. Esophageal atresia (EA) is thought to be due to slow esophageal endoderm proliferation; however, the cause of tracheoesophageal fistula (TEF), and the reason why the two defects are typically found together, remains controversial. The fundamental morphogenetic process appears to be a rearrangement of the proximal foregut into separate respiratory (ventral) and gastrointestinal (dorsal) tubes, which depends on the precise temporal and spatial expression pattern of multiple genes. Disturbance of this pattern disrupts foregut separation and underlies the development of tracheoesophageal malformations (Ioannides and Copp 2009).

Congenital Abnormalities

Branchial Apparatus Anomalies

Branchial (pharyngeal) apparatus anomalies are the second most common congenital neck pathology after thyroglossal duct cysts. It is estimated that they account for approximately 20 % of all pediatric cervical lesions. Drainage from an opening and repeated infections are the most common additional presenting symptoms (Goff et al. 2012; Bajaj et al. 2011). These anomalies result from abnormal embryogenesis of the branchial apparatus and may manifest as a combination of sinus (a single opening onto skin or mucosa), cyst (no communication with skin or mucosa), or fistula (open communication between skin and a mucosal surface). Sinuses, and especially cysts, tend to present in older children and adults and are more common than fistulas, which are typically found in infants (Bajaj et al. 2011; Schroeder et al. 2007). Branchial cysts (BC) are thought to result from incomplete obliteration of an epithelial-lined cervical sinus. A “branchial cleft sinus” connects to the skin, whereas a “branchial pouch sinus” connects to the pharynx, neither of them arising from the mesenchyme of the branchial arch. A true fistula is a communication between two epithelialized surfaces; a “congenital branchial fistula” should be present at birth and should communicate between a persistent pouch and a cleft. Most of the branchial fistulas reported in the literature are actually pseudofistulas, formed between a pouch remnant and a skin opening following an infection or a surgical incision, or are just sinuses. Branchial anomalies are classified according to their proposed pouch or cleft of origin. Those arising from the second branchial apparatus are most common, followed by the first branchial cleft; the third, and especially the fourth, are very rare. Bilateral anomalies occur in 2–3 % of cases and have a strong familial association (Goff et al. 2012; Bajaj et al. 2011; Schroeder et al. 2007).

Branchial apparatus anomalies are usually well visualized by ultrasound (US) and may to some extent be evaluated with pharyngoscopy and barium swallow studies, but computerized tomography (CT) and especially magnetic resonance imaging (MRI) are superior in determining the extent of these lesions and their relations with the surrounding structures (Guarisco and Fatakia 2008; Mukherji et al. 2000). Intact cysts have low CT densities, in the range of 10–25 HU, and are of high T2 and low T1 signal. CT and fluoroscopy with direct injection of contrast material provide the best visualization of fistulous tracts (Goff et al. 2012). The distribution of the lesions, internal openings, and relationship with parotid gland, carotid sheath, and submandibular gland are clearly demonstrated on CT cross-sectional or volume-rendering images (Sun et al. 2012). Intraoperative fistulograms are easy to perform and are a very useful tool in the management of these abnormalities (Guarisco and Fatakia 2008). Surgical removal may be difficult, and inadequate resection is likely to cause recurrence. It seems that anomalies without epithelial lining and those with multiple preoperative infections are much more likely to recur (Schroeder et al. 2007). The treatment of choice for all these lesions has historically been complete surgical excision; however, recent
studies have shown that less invasive procedures are promising for several anomalies (Goff et al. 2012). Certain controversial issues, such as the branchial origin of lateral cervical cysts and the differentiation between third and fourth branchial pouch sinuses, remain unresolved.

First Branchial Cleft Anomalies

They compose somewhere between 8 % and 19 % (higher percentages have been found in more recent studies) of all branchial cleft anomalies and usually present before 10 years of age. They are more common on the left side and females are affected twice as frequently as males. According to the location, they can be subdivided into Type I, adjacent to the external auditory canal (EAC), and Type II, at the angle of the mandible. Patients have histories of recurrent parotid abscesses, otorrhea, or skin pits near the angle of the mandible. The average delay between the presentation and adequate treatment is 4 years. The anomaly is most commonly found within the parotid gland, but may be situated anywhere from the floor of the EAC to the hyoid bone (Bajaj et al. 2011; Schroeder et al. 2007; Mukherji et al. 2000; D’Souza et al. 2002; Whetstone et al. 2006).

Imaging Studies

CT is probably the study of choice for evaluation of suspected first branchial anomalies. Lesions appear as unilocular cysts within the parotid gland, with a thin rim that does not significantly enhance after contrast administration, typically located within the inferior aspect of the superficial lobe of the parotid gland (Figs. 2 and 3). Other cystic masses may mimic this lesion, including retention cysts and sialoceles. The fistulas extend from the undersurface of the EAC, through the parotid gland, to an opening located between the sternocleidomastoid muscle, hyoid bone, and angle of the mandible (Mukherji et al. 2000; D’Souza et al. 2002; Whetstone et al. 2006). The fistulous communication and the bony defect in the floor of the EAC are best visualized on thin section coronal CT images with bone algorithm (Fig. 4) and with CT fistulograms (Sun et al. 2012; Mukherji et al. 2000; D’Souza et al. 2002; Whetstone et al. 2006).

The internal appearance of the cystic components varies with the protein content of the lesion. Pure cystic lesions show expected standard characteristics – low density on CT, hypointense signal on T1-weighted images, and hyperintense signal on T2-weighted images. With recurrent infections, an increase in both CT attenuation and T1 signal may be found, due to higher protein content. Also, the cyst wall may become thicker and show prominent contrast enhancement. During embryologic development, the parotid gland migrates posteriorly, whereas the facial nerve moves anteriorly. A first branchial anomaly has therefore a variable relation to the facial nerve, which should be determined prior to surgery,
as facial nerve injury is a recognized surgical complication (Mukherji et al. 2000; D’Souza et al. 2002; Whetstone et al. 2006).

Second Branchial Cleft Anomalies
Cysts are the most common form of second branchial anomalies with recurrent inflammation as the typical presenting symptom. These lesions are occurring in a bimodal age distribution, primarily in infants and young adults (James et al. 2007). The characteristic position of these lesions is in the triangle between the submandibular gland, sternocleidomastoid muscle, and carotid arteries (Fig. 5), but they may occur anywhere along the developmental pathway of the second branchial complex. Bailey classified second branchial cleft cysts into four types (Guarisco and Fatakia 2008; D’Souza et al. 2002) (Table 2) (Fig. 6). Recent reports suggest that bilateral second BCs may be found in almost 20 % of cases. Sclerotherapy and endoscopic excision of second BCs are promising less invasive treatment procedures (Goff et al. 2012; Bajaj et al. 2011).

Imaging Studies  The study of choice for imaging patients with BC is some form of cross-sectional imaging, whereas direct injection of the external orifice should be performed in patients with a sinus or
fistula. Direct opacification of a fistula or sinus characterized by a linear collection of contrast that ascends toward the upper neck on a CT or fluoroscopic fistulogram is essential prior to surgical resection. Standard CT and MRI may not visualize the tract; however they demonstrate the relationship to adjacent structures, which may also be done on source CT images following filling of the tract with contrast for CT fistulogram (Fig. 7).

On US, the cysts are sharply marginated, round to ovoid anechoic masses with a thin wall and posterior acoustic enhancement. They are compressible and displace the surrounding tissues. Fine internal echoes are occasionally found. On CT, these lesions present as thin-walled cysts, often located along the anterior aspect of the sternocleidomastoid muscle (SCM), but may be found anywhere from the anterior lower third of the SCM to the tonsillar fossa (Figs. 7 and 8) (Guarisco and Fatakia 2008; Mukherji et al. 2000; D’Souza et al. 2002). In most patients the tract extends through the carotid bifurcation and up to pharyngeal constrictor muscles. BC typically show little peripheral contrast enhancement, whereas a thicker rim of enhancement suggests prior infection (Fig. 9). Reticulation and stranding of the adjacent subcutaneous fat, in addition to an enhancing rim, point to an ongoing infection. The internal characteristics of these lesions can vary; the fluid in chronically infected cysts may be of increased CT density, suggesting higher protein content. BC may also contain internal septations.

Table 2  Bailey’s classification of second branchial cysts

<table>
<thead>
<tr>
<th>Second branchial cyst type</th>
<th>Location</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type 1</td>
<td>Just deep to the platysma, along the anterior surface of the sternocleidomastoid muscle. It is the most superficial</td>
</tr>
<tr>
<td>Type 2</td>
<td>Anteriorly and medially to the sternocleidomastoid muscle, lateral to the carotid space, and posterior to the submandibular gland. It is the most common. It probably arises from the persistent cervical sinus</td>
</tr>
<tr>
<td>Type 3</td>
<td>Medial extension between the internal and external carotid arteries to the lateral pharyngeal wall</td>
</tr>
<tr>
<td>Type 4</td>
<td>Deep to the carotid arteries in the pharyngeal mucosal space, at the tonsillar level</td>
</tr>
</tbody>
</table>

From Bailey (1929)
On MRI, these lesions typically demonstrate their cystic nature, with low signal on T1-weighted images and high signal on T2-weighted images (Fig. 8). Similar to CT, this appearance may vary, depending on the protein content of the cyst fluid. Highly proteinaceous fluid may have increased T1 and, possibly, decreased T2 signal intensity. The most common of these cysts (Bailey type 2) displace the submandibular gland anteriorly, push the carotid space vessels medially or posteromedially, and displace the SCM posterolaterally (Figs. 5 and 9). Bailey type 2 cysts are found in the same location as the jugulodigastric lymph node, and they may be mistaken for an enlarged necrotic node (alternatively in adults a cystic metastatic lymph node may be mistaken for a BC). Occasionally, a “beak sign,” considered a pathognomonic imaging feature of a second branchial cleft cyst, may be seen on CT or MRI. This sign refers to a curved rim of tissue pointing medially between the internal and external carotid arteries and is most commonly found in Bailey type 3 but also in Bailey type 2 cysts (Bajaj et al. 2011; Schroeder et al. 2007; Sun et al. 2012; Mukherji et al. 2000; Harnsberger 1995).

**Third and Fourth Branchial Cleft Anomalies**

Congenital anomalies of the third and fourth pharyngeal complex (Fig. 10) are reported to be rare, in the range of 3–10 % of all branchial lesions. Third branchial cleft anomaly usually presents as a fluctuant mass in the posterior cervical triangle. Although rare, they are the second most common congenital lesions of the posterior cervical space, after lymphatic malformations. A classic fistula of third branchial origin communicates externally along the anterior border of the sternocleidomastoid muscle (Fig. 11), similar to a second branchial arch anomaly. The sinus (or fistulous) tract ascends from the base (upper part) of the pyriform sinus, courses above the superior laryngeal nerve, passes through the thyrohyoid membrane, and
travels within the carotid sheath posterior to the carotid arteries, between the hypoglossal and glossopharyngeal nerves (Mukherji et al. 2000; Harnsberger 1995; James et al. 2007; Nicoucar et al. 2010; Rea et al. 2004; Mahomed and Youngson 1998).

They typically arise from the piriform sinus of the hypopharynx, almost always (80–97%) on the left side, and usually present as neck abscess or acute suppurative thyroiditis. Preoperative direct

Fig. 7  Second branchial cleft fistula. (a) Axial CT image following contrast injection into a small opening on the skin (arrow) along the lower neck shows subcutaneous extension of contrast agent (arrowhead). (b) Axial CT image at oropharyngeal level demonstrates cranial extension of contrast agent along the tract (arrow) toward the right palatine tonsil, where it leaks through an internal opening and accumulates in a dependent location (arrowhead). (c–e) Various 3D reconstructions of the CT data reveal the entire fistulous tract (arrows)
laryngoscopy reveals a pit within the apex of the piriform fossa. Children typically undergo multiple surgical procedures before the diagnosis is made. Fistula formation is typically iatrogenic, secondary to incision and drainage. Surgical excision involves ipsilateral thyroidectomy as the lesion passed through the thyroid gland. Endoscopic cauterization or sclerotherapy at the piriform opening may be an effective less invasive treatment for these lesions (Goff et al. 2012; James et al. 2007; Nicoucar et al. 2010; Rea et al. 2004; Mahomed and Youngson 1998).

These anomalies have been recently reevaluated and no lesions following the classical course of either a third or fourth branchial anomaly were identified. The clinical presentation of branchial sinuses arising from the piriform fossa is more in keeping with derivation from the thymopharyngeal duct (of the third pouch) than the hypothetical course of third and fourth branchial fistulae (James et al. 2007; Thomas et al. 2010).

Fig. 8  Second branchial cleft sinus. Coronal STIR MR image demonstrates a well-circumscribed hyperintense lobulated mass (arrow) in the left neck that extends toward the palatine tonsil (arrowhead)

Fig. 9  Infected second branchial cleft cyst. Contrast-enhanced axial CT shows a thick enhancing rim of a centrally hypodense mass lesion (arrow) in the typical location between the submandibular gland, carotid arteries, and SCM
Fig. 10 Theoretical course of the third and fourth branchial cleft anomalies according to the classic teaching. (a) Third branchial cleft anomaly: the cyst is located posterior to SCM and the tract typically ascends posterior to the internal carotid artery. It then courses medially to pass over the hypoglossal nerve and below the glossopharyngeal nerve and pierces the posterolateral thyrohyoid membrane to communicate with the pyriform sinus. c cyst, a internal carotid artery, h hypoglossal nerve, g glossopharyngeal nerve, m thyrohyoid membrane. (b) Fourth branchial cleft anomaly: the cysts are located anterior to the aortic arch on the left and the subclavian artery on the right, respectively. The sinus tract hooks inferiorly around the adjacent vascular structures and ascends to the level of the hypoglossal nerve posterior to the carotid arteries. It then loops over the hypoglossal nerve to pass deep to the internal carotid artery. c cyst, aa aortic arch, sa subclavian artery, h hypoglossal nerve.

Fig. 11 Third branchial cleft anomaly. Fluoroscopic fistulogram shows contrast material (arrowheads) ascending from the opening in the skin along the anterior aspect of the SCM.
Imaging Studies  The diagnosis of an isolated third branchial cleft cyst can be suggested by various imaging techniques. In a recent review article, barium swallow, direct laryngoscopy, and MRI were deemed the most useful diagnostic tools (Nicoucar et al. 2010). Characteristically, these lesions are situated in the lateral compartment of the neck, posterior to the common carotid artery and jugular vein, usually in the posterior triangle (Fig. 12), and they may extend inferiorly to the level of the thyroid gland (Fig. 13). There is no significant enhancement of the cyst wall, and the overlying SCM is displaced laterally. Lymphatic malformations are also found in this location and may have identical appearance on cross-sectional imaging studies (Mukherji et al. 2000; Mahomed and Youngson 1998).

Fistulous tracts arising from the fourth branchial complex are even less common. The opening of the fistulous tract is again found along the anterior border of the SCM. The tract then hooks inferiorly around the adjacent vascular structure derived from the fourth arch (left = aortic arch, right = subclavian artery). It then ascends in close association with the recurrent laryngeal nerve to the level of the hypoglossal nerve. The tract loops over the hypoglossal nerve and passes deep to the internal carotid artery along the tracheoesophageal groove, penetrates the thyrohyoid membrane inferior to the superior

![Fig. 12 Third branchial cleft anomaly](image1)

![Fig. 13 Third branchial cleft anomaly](image2)
laryngeal nerve, and communicates with the apex of the pyriform sinus (Mukherji et al. 2000; James et al. 2007; Nicoucar et al. 2010; Rea et al. 2004). At surgery, the relationship of the sinus tract to the superior laryngeal nerve and the pyriform sinus will differentiate between third and fourth BCs. However, more recent reports negate this claim and show that considerable overlap exists. Imaging and surgical findings together suggest that branchial anomalies at the piriform fossa are sinuses arising from the embryonal thymopharyngeal duct, since they do not follow the classic hypothetic course of third or fourth arch fistulas. The use of the term “third branchial sinus” may, therefore, be most appropriate for branchial lesions involving the thyroid gland (James et al. 2007; Thomas et al. 2010).

Thyroglossal Duct Cysts and Lingual Thyroid

Thyroglossal duct cysts (TGDC, TDC) are the most frequently encountered congenital lesion of the neck, accounting for 70 % of them, and are approximately three times as common as branchial anomalies (Zander and Smoker 2014). They develop from remnants of the thyroid anlage and may present at any age. Failure of involution of the thyroglossal duct (Fig. 14) predisposes to TGDC formation. The migration of thyroid primordium begins at the foramen cecum at the base of the tongue and then loops around the hyoid bone anteriorly and inferiorly, descending anteriorly to the thyrohyoid membrane to its final normal location. TGDC and thyroid ectopia are found in one of the four typical locations with respect to this embryologic course: the base of the tongue, adjacent to the hyoid bone, the midline infrahyoid portion of the neck, and, rarely, the lateral part of the neck. Cysts located adjacent to the tongue base are lined with stratified squamous epithelium, while those adjacent to the thyroid gland are lined with cells resembling the thyroidal acinar epithelium. More than half of all TGDC walls contain ectopic rests of thyroid tissue. Cyst formation is considered secondary to either inflammatory changes or persistent drainage of fluids from secretory epithelium (Zander and Smoker 2014).

These lesions present as nontender mobile midline masses. Rapid growth may result from an associated upper respiratory tract infection. Because TGDC are often attached to the tongue or hyoid bone, they characteristically move with tongue protrusion. Complete excision of the cyst with the Sistrunk procedure is the treatment of choice. The vast majority of TGDC are benign lesions. Coexistent malignancy may be found in 1 % of cases, usually as an incidental finding. Malignancy arises from ectopic thyroid tissue and is usually a papillary carcinoma, but squamous cell cancers may also occur (Branstetter et al. 2000;
Approximately 70–80% of patients with a lingual thyroid have no other functioning thyroid tissue. Additionally, one third of patients with only lingual thyroids have hypofunctionality (Rahbar et al. 2008).

Imaging Studies
US may easily establish the diagnosis if an anechoic midline mass with a thin outer wall is found in the anterior neck. However, this is seen in less than half of the cases. CT may be the imaging modality of choice since MRI is more likely to be degraded by respiratory motion artifact, especially in children. TGDC presents as a unilocular or multilocular cystic mass with or without peripheral enhancement. TGDC may arise anywhere along the course of migration of the embryonic thyroglossal duct and thyroid gland, with the majority of lesions located below the level of the hyoid bone (65%) and 20% at the tongue base (Figs. 15 and 16). TGDC may be slightly paramedian in location (Fig. 16), although it is usually found in the midline (Fig. 17). TGDC may be anterior to and posterior to or encompass the hyoid bone (Figs. 16, 17, and 18). Below the hyoid bone, TGDC are often centered within the strap muscles of the
neck (Fig. 19). Frequently, TGDC may consist of only small tract remnants, rather than a true cystic lesion (Zander and Smoker 2014; Waddell et al. 2000; Tumu et al. 2014). Recurrent infection may increase the attenuation of the cystic component and cause thickening of the enhancing rim (Fig. 20).

On MRI, TGDC typically presents as a fluid-containing lesion, of low T1 and high T2 signal intensity (Fig. 15). Sagittal MR images are very useful for determining the extent of the lesion prior to resection. A solid mass in association with a presumed TGDC usually represents ectopic thyroid tissue. Vallescular cysts may be indistinguishable from TGDC at the tongue base on imaging studies (Fig. 21) (Tumu et al. 2014). Malignancy occurring within TGDC also presents as a soft tissue mass that enhances with contrast and often contains calcifications that are best visualized on CT (Branstetter et al. 2000; Glastonbury et al. 2000). A $^{123}$I thyroid scan may be used to differentiate ectopic thyroid from malignancy, since the thyroid tissue accumulates the radiotracer, whereas malignant tumors are typically “cold” masses. Radionuclide scanning is also justified in cases where the thyroid gland cannot be detected in its normal location (Fig. 22) (Rahbar et al. 2008; Tumu et al. 2014).
Interestingly, a recent study concluded that the US was the preferred exam given its comparable diagnostic accuracy, ease of administration, and lower cost. The added risks of general anesthesia with MRI and ionizing radiation with CT were not considered justified in this clinical setting, as their diagnostic performance was equivalent or inferior to the US (Huoh et al. 2012).

**Dermoid Cysts**

The term “dermoid cyst” (DC) has been loosely used to identify epidermoid and dermoid ectodermal inclusion cysts. Both of these cysts arise from trapped pouches of ectoderm and are lined by squamous epithelium. Epidermoids are filled with debris from desquamated epithelium, which consists of keratin and some cholesterol from the breakdown of cell membranes. Dermoids have thicker lining that includes adnexal structures – sebaceous glands, sweat glands, and hair follicles. In addition to keratin and cholesterol, they also contain lipid material from sebaceous secretions. DC account for around 10 % of congenital neck masses, following TGDC and branchial anomalies (Pryor et al. 2005; Connolly and MacKenzie 1997; Al-Khabteeb and Al 2007). Dermoids may not be identified until the second or third decade of life, whereas epidermoids are much less common and typically present during infancy.
Fig. 21 Vallecular cyst. Midsagittal T2-weighted image shows a hyperintense round mass lesion (arrowhead) projecting posteriorly from the tongue base (Case courtesy of A. Tumu and M. Maheshwari, Children’s Hospital of Wisconsin, Medical College of Wisconsin, Milwaukee).

Fig. 22 Ectopic thyroid gland. (a) There is an oval lesion (arrow) that is slightly hyperintense to the adjacent strap muscles on axial T1-weighted image. (b) The lesion shows mild homogeneous enhancement on postcontrast coronal T1-weighted image. Nuclear medicine thyroid scan in the frontal (c) and lateral (d) projections reveals radiotracer uptake in the lesion (arrow), consistent with thyroid tissue. No other functional thyroid tissue is present.
The majority of DCs in the head and neck are found in the orbital area, with the oral cavity being the second most common location (Harnsberger 1995; Al-Khabteeb and Al 2007), but they may present along the midline all the way down to the suprasternal notch. DCs of the floor of the mouth are curiously free of hair compared to DCs in other locations (Guarisco and Fatakia 2008). They are enclaved during midline closure of bilateral first and second branchial arches and predominantly arise in the floor of the mouth as midline lesions. DCs may rarely be located on the dorsum of the tongue and along the palate. Sublingual DCs are centered above the mylohyoid muscle and present as submucosal lesions in the floor of the mouth. Because these lesions may elevate the tongue, they may be clinically confused with a ranula (which is very rare in children). Submental DCs are centered below the mylohyoid muscle and typically present as a swelling just above the hyoid bone. Since they are not fixed to the tongue or hyoid bone, these lesions, unlike thyroglossal duct cysts, do not move when the tongue is protruded (Pryor et al. 2005; Connolly and MacKenzie 1997; Al-Khabteeb and Al 2007; Koeller et al. 1999; Smirniotopoulos and Chiechi 1995). Most DCs are located superior to the mylohyoid muscle and will be removed intraorally, while lesions that are inferior to the mylohyoid muscle require an external submandibular approach (Harnsberger 1995; Koeller et al. 1999; Smirniotopoulos and Chiechi 1995).

Imaging Studies
On imaging, these lesions present as thin-walled unilocular midline masses within the submandibular or sublingual space. The presence of fat within the lesion is highly characteristic for a dermoid cyst. These lesions are usually sharply demarcated and may contain a thin enhancing rim, often with dystrophic calcifications. On CT, the central cavity is usually filled with homogeneous hypodense material. The coalescence of fat into small nodules may give a “sack of marbles” appearance, which is virtually pathognomonic for a dermoid in the floor of the mouth (Fig. 23) (Harnsberger 1995; Koeller et al. 1999; Smirniotopoulos and Chiechi 1995). Fluid-fluid levels may also be found. In a similar fashion, US shows that the mass filled with hypoechoic fluid contains multiple well-defined suspended hyperechoic nodules (Fig. 23). Dermoids have variable signal characteristics on T1-weighted images ranging from hyperintense to isointense relative to muscle, more commonly hyperintense due to the presence of lipids. They are usually hyperintense on T2-weighted images (Fig. 23).

Epidermoids show fluid attenuation on CT and are T1 hypointense and T2 hyperintense, following the signal intensity of fluid. They are of characteristically very high signal on DWI, due to a combination of very high T2 signal and diffusion characteristics similar to soft tissues. MRI depicts the exact location of DC with respect to mylohyoid muscle. Sagittal and especially coronal T1-weighted images are the most valuable for planning surgical resection.

Vascular Malformations
The International Society for the Study of Vascular Anomalies (ISSVA) adopted in 1996 the modern system of classification, dividing vascular abnormalities into two broad categories: (i) vascular tumors and (ii) vascular malformations. Vascular tumors are true neoplasms characterized by increased mitotic activity and cellular hyperproliferation. Vascular malformations are not neoplasms but products of abnormal vascular morphogenesis and are subcategorized and named by the primary constituent endothelial subtype: arterial, venous, capillary, and lymphatic. Combined/mixed/complex vascular malformations are described by their constituents, such as arteriovenous or venolymphatic. Despite these efforts, the use of colloquial terms remains widespread, perhaps with even greater frequency than the ISSVA terms. Inconsistent and incorrect usage of these terms continues, potentially leading to inappropriate clinical management (Mallucci 1999; Baer et al. 2014). Capillary malformations are
diagnosed clinically and are often untreated. Of note, they can be associated with Sturge-Weber syndrome, when localized in the dermatome of the ophthalmic division of the trigeminal nerve. We will discuss lymphatic, venous, and arterial malformations. For guiding treatment, these malformations may also be divided according to their relative flow velocities (high or low flow), though in reality flow velocity exists on a continuum. Vascular malformations grow proportionately with the child, but can

Fig. 23 Sublingual dermoid cyst. (a) Contrast-enhanced axial CT image shows a lobulated nonenhancing well-circumscribed hypodense mass containing multiple small nodules of even lower attenuation. (b) Sagittal US image shows corresponding suspended hyperechoic nodules within the hypoechoic fluid – the “sack of marbles.” (c) Axial T2-weighted image with fat saturation at a level similar to (a) again reveals multiple globules within the midline lesion that is larger to the left. (d) Midsagittal T1-weighted image demonstrates that the mass lesion (arrows) is below the tongue (arrowhead), consistent with sublingual location. (e) Postcontrast coronal T1-weighted image with fat saturation shows minimal peripheral enhancement of the mass, which is located above the peripherally displaced mylohyoid muscles (Case courtesy of B. Campbell, Medical College of Wisconsin)
change in conspicuity with hormonal changes, accidental or iatrogenic trauma of the lesion, or even with the Valsalva maneuver (Mallucci 1999; Baer et al. 2014).

Lymphatic Malformations
Lymphatic malformations (LMs) constitute about 5 % of all benign lesions of infancy and childhood. As with all vascular malformations, LMs are present at birth, and 90 % of the cases are diagnosed by 2 years of age. The extracranial head and neck are the most common sites of occurrence for LM (75–80 %) (Harnsberger 1995; Mallucci 1999; Baer et al. 2014; Borecky et al. 1995). The terms “lymphangioma” and “cystic hygroma” are no longer recommended by the ISSVA to describe LMs but are still frequently used in the literature and clinical practice. LMs are subcategorized into macrocystic, microcystic, and mixed. The pathophysiology behind their formation is unclear, but proposed mechanisms include sequestration of lymphatic tissue (preventing normal drainage into the venous system), abnormal lymphatic budding, and failure in the development of normal venolymphatic drainage channels (Baer et al. 2011).

Macrocystic LM (cystic hygroma) is by far the most common form, typically located in the posterior cervical space, followed by the submandibular region. Cystic hygromas consist of large dilated cystic spaces, lined by endothelium and separated by minimal amount of intervening stroma. Most cystic hygromas are asymptomatic painless masses, of variable size. They tend to grow proportionately with the child, but some may spontaneously regress (probably <5 %), sometimes even completely. Some of these lesions may extend into the axilla and mediastinum as well as across the midline. Large cystic hygromas may lead to life-threatening airway compression. Trauma or infection may induce a sudden increase in size, with airway compromise as a possible complication (Harnsberger 1995; Baer et al. 2014).

Surgical treatment of LMs is often difficult, as these benign masses can infiltrate and extend deep into the surrounding tissues. Over the last two decades, intraleisonal injection of a sclerosing agent (typically OK-432) has become the first-line therapy, which may be preceded by percutaneous aspiration of the cysts (Mitsukawa and Satoh 2012). Even prenatal sclerotherapy appears to be a safe and effective intrauterine treatment for large macrocystic fetal neck LM (Mikovic et al. 2009). Microcystic lesions are notoriously difficult to treat, and recurrence is common.

Imaging Studies
On US, these lesions usually appear as multilocular cystic masses with septa of variable thickness. The echogenic portions correspond to clusters of abnormal lymphatic channels. Prenatal US typically shows cystic hygroma as an anechoic septated mass in the posterior neck (Tekşam et al. 2005).

MRI (Fig. 24) is the preferred modality because it shows best the full extent of the lesion and its relationship to the adjacent soft tissues. T2-weighted images give the best contrast between the lesions and normal tissues, providing precise preoperative mapping, and they can also identify whether the larynx and trachea are partially or completely compressed on fetal MRI (Tekşam et al. 2005; Rahbar et al. 2005). Postcontrast T1-weighted images usually do not show enhancement, but sometimes may allow for better delineation of the mass and show enhancement of the internal septations. Cystic hygromas typically appear as circumscribed, sometimes multiloculated masses, without a visible wall. They typically have a homogeneous appearance, consistent with fluid-filled cystic spaces, with low to intermediate signal intensity on T1-weighted images and high signal on T2-weighted images (Baer et al. 2014; Borecky et al. 1995; Fung et al. 1998).

On CT, cystic hygromas have low density, sometimes similar to water (Fig. 25). The lesions may be heterogeneous with hyperintense areas on T1-weighted images and CT (Figs. 24 and 25), associated with clotted blood products or high lipid content. Fluid-fluid levels are indicative of recent hemorrhage and represent a common and characteristic finding in LMs (Fig. 26).
Fig. 24  Macrocystic lymphatic malformation. (a) Axial T2-weighted image shows a large lesion of the left neck with a very high signal intensity indicative of fluid. The lesion contains internal septations (arrows). (b) Coronal STIR image demonstrates the craniocaudal extent of the multilocular cystic lesion. Flow voids (arrowheads) correspond to major vascular structures that are running through the mass. (c) Axial T1-weighted image reveals that some of the loculated cysts are of higher signal intensity (arrows), consistent with increased hemorrhage and increased protein content. Flow voids of the carotid arteries (arrowheads) are also noted.

Fig. 25  Macrocystic lymphatic malformation. Nonenhanced axial CT image shows a well-demarcated cystic mass lesion in the right neck that extends from the posterior triangle to the retropharyngeal space and contains a central hyperdensity indicative of hemorrhage.
Microcystic LMs often have a more infiltrative appearance, and discrete cystic spaces can be difficult to appreciate (Fig. 27). The cystic cavities of LM do not enhance, but the septa do. In microcystic LMs, enhancing septa can be so closely packed that postcontrast MR imaging may produce the illusion of an enhancing mass (Baer et al. 2014). Extension to the retropharyngeal space is often seen, and cystic hygromas are the most common retropharyngeal space masses in childhood (Figs. 24 and 25). They commonly arise in the posterior triangle of the neck and may extend into the mediastinum. These lesions usually do not displace normal structures, but large cystic hygromas may show considerable mass effect. Partially resected or repeatedly infected lesions may demonstrate an enhancing wall or prominent internal septations. The absence of feeding arteries or draining veins generally allows for differentiation of lymphangiomas from hemangiomas.
Venous Malformations

Venous malformations (VMs) are the most common vascular malformations, for which terms “venous angioma” and “cavernous hemangioma” have been often used in the literature (although incorrectly, as they are not neoplasms). They can be located anywhere in the body – in mucosal, intradermal, subcutaneous, intramuscular, and intraosseous locations as well as within the internal organs. Approximately 40% of VMs are found in the head and neck, where their clinical presentation depends on lesion size and depth. Superficial VMs are classically painless blue or purple lesions that are compressible on examination and increase in size with dependent positioning, crying, or the Valsalva maneuver. Deeper VMs can be trans-spatial and result in remodeling of adjacent bone, but in contrast to high-flow malformations, VMs should never have a palpable thrill or auscultatory bruit. VMs are not aggressive; however larger lesions can result in disfigurement and dysfunction secondary to the mass effect. Hemorrhage and thrombophlebitis are also common (Baer et al. 2014; Legiehn and Heran 2008).

First-line treatment options for VMs include compression garments, sclerotherapy, embolization, and laser therapy. Absolute ethanol (95–98%) is the most commonly used and most effective sclerosant but has the greatest potential for toxicity. To avoid egress into the systemic circulation, large draining veins may be occluded with embolization coils, temporary balloons, or external compression before injection. The procedure may be repeated every 8–10 weeks until results are adequate. Surgical resection is mostly reserved for lesions that cannot be treated otherwise (Baer et al. 2014; Legiehn and Heran 2008).

Imaging Studies

US is the initial diagnostic tool for evaluation of suspected vascular malformations and can usually characterize lesions as high or low flow, which is valuable for both diagnosis and treatment selection. VMs are often compressible and contain phleboliths, essentially pathognomonic within a vascular abnormality, appearing as shadowing echogenic foci. Lesions commonly appear as poorly margined, hypoechoic vascular channels without substantial intervening echogenic parenchyma (Baer et al. 2011, 2014). Very large or deep suspected vascular anomalies are best visualized with MR imaging or CT. VMs are often trans-spatial and T2-weighted images are best suited for the evaluation of their extent. On T1-weighted images, VMs are as hypo- or isointense to muscle with a lobular appearance. VMs can have imaging features very similar to hemangiomas, but in many cases they can be distinguished by the presence of “venous lakes” (representing larger vascular channels) that are of homogeneous high T2 signal and phleboliths, small foci of very low signal on all pulse sequences within venous lakes (Figs. 28 and 29). Venous lakes are moderately to markedly T2 hyperintense, not as bright as simple cysts or lymphatic malformations but more hyperintense than hemangiomas. Smaller channels have intermediate signal intensity, while heterogeneity can be produced by intralstional thrombus or hemorrhage (Legiehn and Heran 2008; Dubois and Alison 2010). Vascular flow voids are not seen with VMs due to their low-flow velocity. Heterogeneous delayed enhancement is typically present on postcontrast images, and MR venography may be useful.

On CT, VMs are multilobular lesions that are heterogeneous but predominantly isodense with muscle. Phleboliths that are characteristic of VM are well seen as small, rounded calcified foci. Remodeling of adjacent bone is also clearly identified on CT. Direct injection of contrast into the lesion with catheter venography is useful in defining lesion extent, communication with normal draining veins, and as a guide for fluoroscopic injection of sclerosing agents (Baer et al. 2011, 2014).

Arterial Malformations

Arterial malformations are a group of high-flow vascular anomalies that include arteriovenous malformations (AVM) and arteriovenous fistulas (AVF). The categories of AVM and AVF are not
mutually exclusive, as they all operate as shunts to some degree. AVMs are characterized by abnormal connections between feeding arteries and draining veins with the absence of a normal intervening capillary network, and 70% are found in the craniofacial region and neck. When fully formed, they often produce an audible bruit and/or palpable thrill. Lesions can enlarge following trauma, instrumentation, and partial surgical resection. They can also grow rapidly with hormonal variation, which may explain their common presentation during puberty. A wide variety of complications from AVMs and AVFs are possible, resulting from malperfusion, arteriovenous shunt surgery, mass effect, and hemorrhage. Syndromes associated with arterial malformations include Rendu-Osler-Weber. The treatment of AVMs centers on obliteration of the vascular nidus, usually by endovascular embolization with or without subsequent surgical resection (Baer et al. 2011, 2014; Dubois and Alison 2010).

Fig. 28 Venous malformation. Axial contrast-enhanced CT image shows a conglomerate of enlarged vascular structures (arrow) within and adjacent to the right masseter muscle, corresponding to venous lakes.

Fig. 29 Venous malformation. Axial T2-weighted image reveals an irregular and discontiguous mildly hyperintense lesion (arrows) with rounded internal areas of very low signal, suggestive of phleboliths (arrowhead).
Imaging Studies

On US, an AVM will appear as a poorly defined and heterogeneous lesion often surrounded by fat. Doppler sonography will identify a network of vessels with arterial waveforms and increased diastolic flow as well as arterialized draining veins (Baer et al. 2014; Dubois and Alison 2010). If a suspected AVM appears to have intervening parenchyma, other diagnoses such as hemangiomas or other hypervascular neoplasms should be considered.

MR imaging and CT angiography are ideal for identification of the entire course and extent of AVMs, including detection of arterial or intranidal aneurysms, as well as venous varices. As with other high-flow lesions, the abnormal vessels appear as flow voids on spin-echo MR sequences, while postcontrast CT and MR images demonstrate arterial and very early venous enhancement. The large network of feeding arteries and enlarged draining veins has a characteristic “bag of worms” appearance. CT has the advantage of better showing osseous extension and intraosseous complications (Fig. 30). Unlike VMs, AVMs are generally not associated with venous lakes. Real-time visualization with catheter angiography allows a clear distinction between arteries and veins and is essential for treatment planning (Baer et al. 2011, 2014).

Fig. 30 Arterial malformation. Axial contrast-enhanced CT image with bone algorithm and windowing demonstrates multiple enlarged vascular structures in the left face with associated expansion of the left maxilla (arrows)

Laryngotracheal Congenital Anomalies

A variety of congenital anomalies arise within the laryngotracheal airway, with stridor being the most common presentation. Symptoms may also include hoarseness and aphonia, less frequently aspiration and other feeding disorders. All newborns and infants with such symptoms should be thoroughly evaluated, which is usually best achieved by endoscopy. However, imaging studies may be used as a screening tool, avoiding the risk of general anesthesia (Tumu et al. 2014; Smirniotopoulos and Chiechi 1995). In addition to obtaining static images, fluoroscopy can visualize lesions that show dynamic changes with respiration. Functional swallowing studies provide valuable information in cases with feeding disorders. Multidetector CT can be used for virtual noninvasive endoscopy. Radiologic studies may be helpful in establishing the diagnosis, but their primary role is to determine the extent of the disease and to detect possible synchronous lesions (Tumu et al. 2014; Wiatrak 2000).

At birth, the larynx remains higher at the C2–C3 level and will eventually settle at the C4–C5 level later in life. The airway above the vocal cords is lined with resilient squamous epithelium, whereas below the
vocal cords, the airway is lined with fragile ciliated columnar epithelium, which is more susceptible to damage and postinflammatory narrowing. The level of the cricoid ring is the narrowest portion of the pediatric airway that is circumferentially encompassed by stiff cartilage. As the caliber of the pediatric airway is already small, even minor changes are substantially amplified (Tumu et al. 2014).

**Laryngomalacia**

Laryngomalacia is the most common congenital laryngeal abnormality and is the most common cause of stridor in infants. The abnormality appears to be related to flaccidity and incoordination of the soft tissues and cartilages of the supraglottic larynx. During inspiration, there is collapse of the supraglottic structures (epiglottis, arytenoids, and aryepiglottic folds) leading to airway obstruction. The condition is typically benign and self-limited, resolving by 18–24 months of age. Other coexistent abnormalities of the larynx are frequently present, and about 80 % of the cases are associated with gastroesophageal reflux. The exact cause of laryngomalacia is still unclear; some studies suggest that essentially all children with laryngomalacia have efflux of gastric acid to the pharyngeal level. In rare severe or prolonged cases, supraglottoplasty may be required, during which the obstructive supraglottic tissue is removed by either laser or other surgical techniques (Olney et al. 1999).

**Imaging Studies**

Frontal and lateral plain films have historically been the imaging modality of choice, showing characteristic anterior bowing and inferior displacement of the aryepiglottic folds during inspiration (Tumu et al. 2014; Walner et al. 1999). However, recent studies showed that plain films have a very low sensitivity for laryngomalacia, as well as for tracheomalacia, and are therefore of questionable value (Walner et al. 1999; Tostevin et al. 1995). Fluoroscopy is a better option, since laryngomalacia is a dynamic process. Spiral CT with virtual endoscopy provides excellent noninvasive assessment, especially in evaluating the extent of the lesion and possible associated anomalies (Tumu et al. 2014).

**Vocal Cord Paralysis**

Vocal cord paralysis (VCP) is the second most common developmental laryngeal anomaly. It may be bilateral or unilateral, with the left side affected more frequently. Bilateral VCP may be idiopathic; however, it is typically associated with abnormalities of the central nervous system, usually a posterior fossa malformation, particularly Chiari malformations. Unilateral VCP is usually caused by a peripheral lesion, the most common causes being congenital cardiac anomalies, surgical repair of a tracheoesophageal fistula or congenital heart defect, and traumatic delivery (Wiatrak 2000).

**Imaging Studies**

Limited abduction of the vocal cord with respiration or crying is seen on fluoroscopy. In unilateral paralysis, imaging studies show a thinner cord and larger laryngeal ventricle on the affected side, and the ipsilateral pyriform sinus is also larger. The role of imaging in most cases of vocal cord paralysis is to detect the presence of an offending lesion. CT is the preferred imaging modality in the evaluation of unilateral VCP, and contrast enhancement is necessary to opacify the vascular structures. Evaluation of the entire course of the recurrent laryngeal nerve is required, so coverage must extend to the level of the aortopulmonary window, when there is left VCP. CT is also useful for evaluating patients who have undergone medialization procedures, since it visualizes the positions of both the vocal cord and the implant. MRI of the brain, including the posterior fossa and skull base, should be performed for possible brain lesions in cases of bilateral VCP. In addition to Chiari malformations, hydrocephalus and posterior fossa tumors may result in bilateral VCP (Castillo and Mukherji 1996a).
Subglottic Stenosis

Congenital subglottic stenoses can be due to extrinsic mass effect or idiopathic, which are part of a continuum of congenital laryngeal abnormalities and usually classified as membranous and cartilaginous. Today, the incidence of acquired subglottic stenosis secondary to prolonged intubation or, less frequently, laryngeal trauma is higher than that of the congenital form. The patients present with respiratory distress and stridor in the early neonatal period. Diagnosis is made when the diameter of the lumen of the subglottic airway in the cricoid region is \(<4\) mm in full-term neonates or \(<3\) mm in premature infants (Tumu et al. 2014; Wiatrak 2000; Hartnick and Cotton 2000). Acquired stenosis is usually more severe but both are treated similarly. Management includes laryngotracheoplasty and splitting of the cricoid in severe cases. Balloon dilation is a more recent highly effective and low-risk method, which may be used as an alternative or adjunct to traditional reconstructive procedures (Wentzel et al. 2014).

Imaging Studies

Stenosis of the subglottic air column may be seen on frontal and lateral plain films of the neck, sometimes having a typical hourglass appearance. This narrowing extends inferiorly for 10–15 mm and does not change with the phase of respiration. However, the sensitivity and specificity of radiography are questionable since croup may have a similar appearance (Tumu et al. 2014; Wiatrak 2000; Walner et al. 1999; Tostevin et al. 1995).

CT is the imaging modality of choice if the extent of the narrowing cannot be fully appreciated by endoscopy (Fig. 31). Volumetric CT is especially useful due to short imaging time and reduced motion artifacts and may obviate the need for sedation. Contrast injection is not needed. Circumferential soft tissue thickening in the subglottic region may extend inferiorly to involve the trachea. The full extent of involvement should be determined and the luminal diameter at the level of the most severely affected area measured.
Laryngeal Webs
Most laryngeal webs are located at the level of the true vocal cords, usually in the region of the anterior commissure (Fig. 32). They may also occur in the region of the posterior commissure, resulting in interarytenoid fixation.

Imaging Studies
Lateral radiographs may show the thickness of the webs, which are typically seen as shelves of soft tissue immediately inferior to the true vocal cords (Hartnick and Cotton 2000). Sagittal and coronal reformations of CT data allow for precise measurement and mapping of the webs. Again, imaging studies are also used to detect the presence and extent of associated laryngeal abnormalities, such as subglottic stenosis.

Laryngeal Cysts
Laryngeal cysts are benign, fluid-filled lesions which probably occur due to obstruction of mucous glands. They may occur anywhere in the supraglottic larynx, the vallecula, or the tongue base. The symptoms and radiographic appearance depend on their size and location.

Imaging Studies
On cross-sectional imaging, laryngeal cysts are well-circumscribed nonenhancing submucosal masses projecting into the supraglottic airway. They are of low density on CT and of high T2 signal intensity on MRI. The T1 signal intensity is variable, as in other cystic lesions, depending on the amount of protein, including the presence of hemorrhage or infection.

Tracheomalacia
Tracheomalacia refers to processes that cause abnormal narrowing or compression of the trachea during respiration. It includes multiple acquired and congenital abnormalities that lead to either external compression or intrinsic laxity. Primary tracheomalacia (TM) is characterized by abnormal flaccidity of the trachea, leading to collapse during expiration, caused by weakness and softening of the supporting cartilage and abnormal widening of the posterior wall. TM usually presents in infants independently of laryngomalacia, but may be associated with it, as well as with bronchomalacia. It is usually a self-limited process that resolves by the second year of life, similar to laryngomalacia. Secondary tracheomalacia is due to extrinsic compression of the trachea by a mass or a vascular structure, most commonly the innominate artery or a double aortic arch. It may also occur in patients with tracheoesophageal fistula, commonly after surgical repair (Tumu et al. 2014; Wiatrak 2000).
Imaging Studies

The initial imaging study has traditionally been frontal and lateral radiograph of the airway, which may show narrowing of the tracheal air column (Fig. 33). However, studies have demonstrated a poor correlation of radiographs with endoscopy in cases of primary tracheomalacia (Walner et al. 1999; Tostevin et al. 1995). The tracheal airway decreases by more than 50% during expiration, which may be visualized by fluoroscopic examination.

Diagnosis is suggested on imaging when the tracheal walls are not perpendicular or when there is contour irregularity of the trachea (Fig. 34). Dynamic airway imaging can be performed with fluoroscopy to evaluate the tracheal caliber in real time (Tumu et al. 2014). Dynamic MRI during the respiratory cycle in sagittal and axial planes may also be performed; real-time axial imaging is especially helpful for evaluation of the degree of tracheal collapse during expiration. CT has the additional advantage of detecting focal areas of dystrophic calcification that may be present within the tracheal cartilages, and non breath-held 3D CT virtual bronchoscopy appears to be a reliable diagnostic tool (Fig. 35). Imaging
plays a major role in patients suspected of having secondary tracheomalacia. Plain chest films may detect vascular compression, and barium swallow can suggest a vascular ring by showing a persistent impression on the posterior esophagus. Contrast-enhanced CT of the chest is a sensitive and specific study to evaluate for the presence of compressive mass lesions (Tumu et al. 2014; Inoue et al. 1998).

**Laryngotracheoesophageal Cleft**

Laryngotracheoesophageal cleft (LTC) is a very rare congenital anomaly resulting in an abnormal communication between the larynx and hypopharynx. LTC is associated with other congenital abnormalities, and an associated tracheoesophageal fistula is found in 20 % of patients. The anomalous communication may be localized to the posterior aspect of the cricoid cartilage or be more extensive and involve the entire common wall between the trachea and esophagus. Patients with LTC usually present in the first weeks to months of life with a variety of symptoms associated with feeding difficulties and respiratory distress (Castillo and Mukherji 1996a; Carr et al. 1999).

**Imaging Studies**

Chest films may show parenchymal opacities, consistent with aspiration pneumonia. The diagnosis of LTC is confirmed with a barium swallow study that demonstrates contrast material simultaneously filling the esophagus and trachea (Castillo and Mukherji 1996a). The study is best performed with the patients placed in the prone position. 3D CT has been used to delineate the lower limit of the cleft, when bronchoscopy is not possible.

**Esophageal Atresia and Tracheoesophageal Fistula**

Esophageal atresia (EA) and tracheoesophageal fistula (TEF) are major congenital malformations affecting 1:3,500 live births. About 35 % of patients are born prematurely, and coexisting anomalies are present in up to 50 % of them. The most common patterns of anomalies are summarized by the acronyms VATER (vertebral defects, imperforate anus, tracheoesophageal fistula, radial and renal dysplasia) and VACTERL.
vertebral, anal, cardiac, tracheal, esophageal, renal, and limb deformities). The presence of at least three of the listed anomalies is diagnostic for VACTERL.

Patients can present with isolated esophageal atresia or with various configurations of fistulous connections with the trachea. TEF has been classified into five separate types (Castillo and Mukherji 1996a) (Table 3). The most common form is a blind-ending esophageal pouch proximally and a fistula connecting the trachea with the distal pouch (Fig. 36). The H-type fistula is important clinically because it can be occult with chronic symptoms, as the esophagus is not atretic. In this form of TEF, both esophagus and trachea are patent, but there is a small fistulous connection distally (Fig. 37). These patients can present with recurrent infections and gastroesophageal reflux leading to aspiration (Tumu et al. 2014).

**Imaging Studies**

Plain films typically demonstrate lack of a stomach air bubble in neonates with complete esophageal atresia. The presence of a gaseous abdomen and air in the stomach in patients with EA is indicative of a

### Table 3 Classification of tracheoesophageal fistula

<table>
<thead>
<tr>
<th>Type</th>
<th>Features</th>
<th>Frequency (%)</th>
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<tbody>
<tr>
<td>Type I</td>
<td>Isolated esophageal atresia</td>
<td>5–10</td>
</tr>
<tr>
<td>Type II</td>
<td>Proximal fistula</td>
<td>1</td>
</tr>
<tr>
<td>Type III</td>
<td>Distal fistula. Esophageal atresia with a blind-ending upper pouch and a fistula connecting the trachea with the distal pouch</td>
<td>85</td>
</tr>
<tr>
<td>Type IV</td>
<td>Proximal and distal fistulas</td>
<td>2</td>
</tr>
<tr>
<td>Type V (H – type)</td>
<td>Without esophageal fistula</td>
<td>6</td>
</tr>
</tbody>
</table>

From Castillo and Mukherji (1996b)

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**Fig. 36** Tracheoesophageal fistula (TEF). (a, b) Fluoroscopy with barium shows contrast material injected into the proximal esophagus (E) spreads into the tracheobronchial tree (T), with direct visualization of the fistula (arrows)
If EA or TEF is suspected, an attempt to pass a pediatric feeding tube through the nose into the stomach should be made. A coiled nasogastric tube in the proximal pouch is indicative of EA. Air may be injected into the nasogastric tube in order to better visualize the proximal pouch. Care must be taken not to perforate the proximal pouch or advance the tube into the bronchial tree, as could occur in TEF with a proximal fistula. Contrast may be injected into the proximal pouch in order to confirm the diagnosis and to identify a proximal fistula, and it also aids

**Fig. 37** H-type TEF. Barium fluoroscopy shows that while the esophagus is patent (*arrow*), the contrast material does spread into the trachea (*arrowhead*) (Case courtesy of A. Tumu and M. Maheshwari, Children’s Hospital of Wisconsin, Medical College of Wisconsin, Milwaukee)

**Fig. 38** TEF. (a) Barium fluoroscopy shows a blind-ending proximal esophageal pouch (*arrow*) consistent with esophageal atresia. (b) Plain film of the chest and abdomen shows looping of the nasogastric tube (*arrow*) due to esophageal atresia. Presence of air in the stomach and gaseous abdomen are indicating a communication between the trachea and the distal pouch communication between the trachea and the distal pouch (Fig. 38). If EA or TEF is suspected, an attempt to pass a pediatric feeding tube through the nose into the stomach should be made. A coiled nasogastric tube in the proximal pouch is indicative of EA. Air may be injected into the nasogastric tube in order to better visualize the proximal pouch. Care must be taken not to perforate the proximal pouch or advance the tube into the bronchial tree, as could occur in TEF with a proximal fistula. Contrast may be injected into the proximal pouch in order to confirm the diagnosis and to identify a proximal fistula, and it also aids
in determining the side of the aortic arch. Only 0.5–1.0 ml of aqueous barium should be applied via the feeding tube under pressure while slowly withdrawing the tip of the feeding tube. Horizontal beam fluoroscopy should be used with the patient in prone position, as this allows barium to opacify the ventral wall of the esophagus, thus maximizing the ability to visualize the fistula (Tumu et al. 2014; Castillo and Mukherji 1996a).

CT and MRI are helpful for identifying and evaluating the multitude of associated abnormalities that are known to occur with EA and TEF. Early reports of 3D CT and virtual bronchoscopy in the assessment of EA and TEF have demonstrated complete agreement with operative and/or bronchoscopic findings (Lam et al. 2000; Fitoz et al. 2000); however according to a more recent study, CT was associated with significant exposure to ionizing radiation while not being helpful for surgical decision making (Mahalik et al. 2012). The combination of polyhydramnios and absent stomach bubble on prenatal US offers only a modest positive predictive value for esophageal atresia, whereas fetal MRI appears to be accurate for establishing or ruling out this diagnosis and should be considered in fetuses who are at high risk based on ultrasound findings (Langer et al. 2001; Salomon et al. 2009).

**Congenital Tracheal Stenosis**

Congenital tracheal stenosis (CTS) is a rare anomaly often associated with other systemic anomalies, such as tracheoesophageal fistula, pulmonary hypoplasia, and anomalous great vessels. CTS is caused by abnormal cartilage which completely encircles the involved tracheal region. The normal horseshoe-shaped rings are replaced by complete or near-complete cartilaginous rings, which result in the absence of the soft and pliable posterior membranous wall (Castillo and Mukherji 1996a). There is no absolute correlation between luminal diameter and prognosis, and therapeutic decisions are difficult for intermediate cases (Mimouni-Benabu et al. 2012).

**Imaging Studies**

Historically, radiographs have been the initial modality of choice for imaging patients suspected of having CTS. These studies may show a narrowed tracheal lumen. However, radiographs have a tendency to underestimate the narrowing (Castillo and Mukherji 1996a).

CT has replaced tracheobronchography as the preferred imaging modality for the evaluation of patients who are suitable candidates for surgical intervention. In CTS, a completely round tracheal air column replaces the normal appearance of trachea, with flattened posterior margin. The caliber of the trachea typically measures less than 3 mm. The cartilages are not yet calcified and therefore are not visualized by CT. Virtual bronchoscopy reconstructions provide a unique and accurate view of the stenotic portion of the tracheal air column (Fig. 39). CT also evaluates for the presence of other anomalies that are known to occur with CTS (Lam et al. 2000). It seems that simulations with computational fluid dynamics based on CT data may provide an objective method to evaluate the severity of symptoms and may help guide treatment (Mimouni-Benabu et al. 2012).

**Neoplasms**

**Teratoma**

Teratomas are tumors arising from misplaced embryonic germ cells. They contain tissues which are either foreign to the site of origin or represent more than one embryonic germ layer. The designation of teratoma is appropriate even for lesions with tissues derived from only a single embryonic germ layer, if divergent differentiation is demonstrated histologically. Teratomas that arise in the head and neck commonly contain neuroepithelial and thyroid elements and are typically found in the cervical region and the
nasopharynx (Smirniotopoulos and Chiechi 1995; Green et al. 1998). These are usually large masses that are clearly evident at birth, and many of them are detected at routine screening prenatal sonography. The vast majority of teratomas in children are benign lesions; however, malignant teratomas do occasionally occur, and the risk of malignancy is increased in patients who have undergone prior resection. Because of their large size, they may be associated with considerable morbidity. Affected infants have symptoms of respiratory obstruction due to deviation and compression of the trachea. The high incidence of maternal polyhydramnios may be due to the inability of the infant to swallow the amniotic fluid. These lesions and associated airway obstruction are diagnosed in utero by prenatal US and MRI, enabling successful performance of EXIT (ex-utero intrapartum treatment) procedures, which in turn secure the fetal airway while preserving fetomaternal circulation (Figueiredo et al. 2010).

**Imaging Studies**

Teratomas may be difficult to distinguish from cystic hygromas, both clinically and radiologically. CT is the preferred modality for imaging patients with suspected cervical teratomas due to short imaging time, which takes on added importance in infants with respiratory obstruction (Fig. 40). Compared to MRI, CT may also be more reliable in detecting calcifications within the lesion (Figs. 40 and 41). The radiographic appearance of a large, bulky, multiloculated, and heterogeneous mass with focal areas of low attenuation on CT and high signal on T2-weighted MR images is characteristic for teratoma (Fig. 42). The tumors are commonly located adjacent to the thyroid and are often surrounded by an enhancing rim. These lesions may extend to the contralateral side and into the thoracic inlet. The presence of intralesional fat and calcification is a typical finding that is also common in dermoids; however, dermoids are unilocular masses. Careful follow-up, including serial alfa-fetoprotein (AFP) measurements and imaging, is required for all patients who have undergone previous treatment due to increased risk of malignancy, regardless of the location of the lesion and the histologic findings (Smirniotopoulos and Chiechi 1995; Green et al. 1998; Figueiredo et al. 2010).

**Hemangiomas**

The term “hemangioma” is perhaps the most inconsistently used in describing vascular anomalies. It is often applied to capillary and venous malformations in addition to true hemangiomas. The term
“hemangioma” should be reserved for anomalies that fulfill the following criteria: (1) present at or soon after birth, (2) experience a period of proliferation, and (3) eventually involute to some degree. There are two categories of hemangiomas: the infantile hemangioma, which is very common, and the congenital hemangioma, which is rare. Hemangiomas are primarily diagnosed clinically. The role of imaging is to confirm the diagnosis, exclude malignancies with similar clinical presentations, identify lesion extent, and identify potential complications of tumor growth (Mallucci 1999; Baer et al. 2011, 2014).

**Infantile Hemangiomas**

Infantile hemangiomas (IHs, also known as hemangioma of infancy, common infantile hemangioma, juvenile hemangioma, capillary hemangioma, or just hemangioma) are the most common neoplasms of the head and neck in children. They can occur anywhere in the body, with the head and neck being the most common location (60 %). They are reportedly present in 1–2 % of all neonates and 10–12 % of infants, with an increased incidence in girls, whites, twins, and premature births. Most affected children

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**Fig. 40** Teratoma. Contrast-enhanced axial CT image shows a rim-enhancing heterogeneous mass containing scattered calcifications (arrowheads) in the low neck, displacing and narrowing the airway (arrow)

**Fig. 41** Teratoma. (a, b) Contrast-enhanced axial CT images show a large heterogeneous cervical mass (arrow) containing small calcifications (arrowhead)
will have a solitary hemangioma, but as many as 20% will have five or more lesions. Benign neonatal hemangiomatosis is characterized by multiple small cutaneous hemangiomas, while the diffuse neonatal hemangiomatosis includes additional multiple visceral lesions. A few syndromes are associated with IHs, most notably PHACE (posterior fossa abnormalities, hemangiomas, arterial abnormalities, aortic coarctation and cardiac anomalies, and eye abnormalities) and PHACES (additional sternal clefting) (Baer et al. 2011, 2014). It has been recognized that IHs are not associated with Kasabach-Merritt phenomenon and consumptive coagulopathy (Enjolras et al. 1997).

Gene-expression profiles have provided insight into the origins of these tumors, including the discovery that endothelial cells from common infantile hemangiomas express glucose transporter isoform 1 (GLUT1) gene throughout their life span (North et al. 2000). A minority of these lesions are notable at birth and they are frequently very subtle. IHs typically present within the first year of life (usually in the neonatal period), show rapid growth, and then undergo involution by adolescence. Endothelial proliferation with high mitotic activity is found histologically during the proliferating phase (phase I). This can last 4–10 months and is followed by a quiescent period and then a prolonged period of involution (phase II, involuting phase), which can last 6–10 years, when the lesion is replaced by fat and fibrotic tissue. IHs generally involute completely by late childhood (phase III, involuted phase) (Baer et al. 2011, 2014).

Superficial lesions are clinically manifested as nontender soft masses with discoloration, but deep lesions may not be clinically apparent. Superficial IHs are more common and imaging studies are usually not necessary. Deep hemangiomas are typically single lesions located within the muscles of mastication, parotid space, and in the submandibular and sublingual spaces. These tumors do not respect the fascial planes and multiple lesions may be encountered. Most IHs are small, do not result in dysfunction, and can be managed conservatively. Treatment is considered for rapidly growing and symptomatic lesions, as well as those that may result in functional compromise or lead to permanent cosmetic deformity. In these cases diagnostic imaging studies are valuable in identifying lesion location and extent (Baer et al. 2011, 2014).

**Imaging Studies**

The role of imaging in suspected hemangiomas is to rule out alternative tumors, determine lesion extent, and establish a baseline before treatment. Both US and MRI are reasonable first-line diagnostic tools in the workup of suspected hemangiomas. For small, isolated, cutaneous lesions, sonography is probably better as it is more cost-effective and obviates the need for sedation or general anesthesia. The characteristic imaging features of a hemangioma depend on its phase of evolution. On US, IHs appear as well-
demarcated tumors of variable echogenicity. During the proliferating phase, they contain numerous large vessels with relatively high flow velocity at color Doppler, which decrease during the later phases. While vascular malformations may also have large vessels and high flow velocities, they do not contain the parenchymal component (Baer et al. 2011, 2014; Dubois and Alison 2010).

The imaging modality of choice for large or deep lesions is MRI, which shows masses that are typically isointense to muscle on T1-weighted images and moderately to very bright on T2-weighted images (Fig. 43). Phase I IHs are sharply circumscribed and fairly homogeneous, showing intense homogeneous enhancement after contrast administration, which is best depicted with fat-suppressed T1-weighted images that provide clear delineation from surrounding muscles and fat. Small “satellite” lesions may be seen immediately adjacent to or at some distance from the dominant mass. They commonly contain prominent vascular structures, which are depicted as flow void on spin-echo sequences and flow-related enhancement on gradient-echo sequences (Figs. 43, 44, and 45). MR angiography (as well as CTA and catheter angiography) may identify the large feeding vessels (Fig. 45). Once involution starts in later phases, IHs become low-flow lesions, without prominent vessels. They tend to show a more heterogeneous enhancement as well as T1 and T2 signal intensity, related to perivascular fibro-adipose deposition. Nonenhancing areas of high T1 signal reflect fatty replacement of the tumor. Findings of rapid growth associated with hemorrhage, or with compromise of the airway, should prompt therapeutic interventions.

If a suspected hemangioma appears as a network of vessels without any enhancing tissue component, an alternative diagnosis of vascular malformation should be considered. On the other hand, if there are relatively few vessels within an enhancing mass or perilesional edema is present, other neoplasms should be considered. On CT, proliferating IHs appear as well-delineated masses with a very prominent and early vessel-like contrast enhancement (Fig. 44). Involuting lesions may contain areas of fat attenuation and

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**Fig. 43** Infantile hemangioma. (a) Axial T2-weighted image with fat saturation shows a very bright mass lesion (arrow) just anterior to the right masseter muscle. (b) Internal flow voids (arrow) are better seen on the coronal STIR image. (c) Coronal T1-weighted image demonstrates that the lesion is isointense with the muscles. (d) Corresponding postcontrast image with fat saturation reveals a strong homogeneous enhancement of the mass.
**Fig. 44** Infantile hemangioma. (a) Contrast-enhanced axial CT image shows bilateral masses in the parotid space (arrows) that demonstrate a dense vessel-like contrast enhancement. (b) Axial T2-weighted image with fat saturation at a similar level shows mildly increased signal of the lesions that extend into the parapharyngeal space. Note internal flow voids.

**Fig. 45** Infantile hemangioma. (a) Coronal STIR image reveals a large mildly hyperintense mass in the left neck that contains flow voids. (b) Sagittal T1-weighted image through lesion shows its relative isointensity. (c) Postcontrast dynamic MR angiogram demonstrates a very early and strong lesion enhancement. (d) Corresponding postcontrast T1-weighted image again shows the enhancing mass and its flow voids.
show heterogeneous enhancement. Phlebolith is not a feature of hemangiomas, and their presence suggests a venous malformation (Baer et al. 2011, 2014; Dubois and Alison 2010).

**Congenital Hemangiomas**

Congenital hemangiomas (CHs) are generally solitary and account for only 3% of all hemangiomas. In contrast to IHs, these lesions do not express GLUT-1 gene, and they complete their proliferating phase in utero so that CHs are always present at birth. They are subdivided into two types: the rapidly involuting CH (RICH) and the noninvoluting CH (NICH). RICHs usually complete their regression within 14 months of life, while without treatment NICHs will persist into adulthood (Baer et al. 2014; Krol and MacArthur 2005).

**Imaging Studies**

The differentiation of CH and IH is based primarily on clinical information. Congenital hemangiomas are rare lesions that are fully grown at birth and either involute rapidly or never spontaneously involute. These lesions share several imaging characteristics, particularly during the proliferating phase, but there are exceptions. CHs at birth may have very high flow velocities, similar to those of arteriovenous malformations but without a true shunt. Furthermore, CHs may have aneurysms and intravascular thrombi that are generally not seen with IHs (Baer et al. 2014; Dubois and Alison 2010).

**Subglottic Hemangioma**

Subglottic hemangiomas (SHs) are infantile hemangiomas located in subglottic region. These are typically asymptomatic at birth; however, as they enlarge rapidly during the first 6–12 weeks of life, SHs can present mimicking croup. Patients can also have concomitant skin findings; characteristic cutaneous hemangiomas in a beardlike distribution are found in approximately 20%. When multiple or atypical hemangiomas are identified, screening for other associated congenital abnormalities should be performed (O-Lee and Messner 2008; Orlow et al. 1997).

The diagnosis of SH is made at endoscopy. Photographic documentation is helpful as biopsy poses the risk of hemorrhage. Treatment is dependent on the severity of symptoms as these lesions typically resolve spontaneously and involute later in childhood, as do IHs in other locations. The dramatic efficacy of β-blockers on IH has radically changed the prognosis. Surgery should now only be performed for difficult, refractory cases, or in the presence of absolute contraindications to β-blockers. Long-term steroid therapy is also no longer indicated, and propranolol should be recommended as a first-line single-agent therapy in infantile airway hemangiomas (Leboulanger et al. 2011; Peridis et al. 2011).

**Imaging Studies**

Frontal and lateral radiographs of the neck classically show an asymmetric soft tissue narrowing, just inferior to the true vocal cords (Fig. 46), in contrast to the symmetric narrowing seen with congenital subglottic stenosis. However, the narrowing may be symmetric in half of the cases of SH, and plain films have low sensitivity for these lesions (Walner et al. 1999; Cooper et al. 1992). CT and MRI are able to precisely delineate the mass and define its extent. SHs are seen as enhancing, polypoid soft tissue masses of high signal intensity on T2-weighted images (Fig. 47) that extend into the airway (Tumu et al. 2014).

**Recurrent Respiratory Papillomatosis**

Squamous papilloma is the most common laryngeal tumor in children, and multiple papillomas are referred to as recurrent respiratory papillomatosis (RPP). The juvenile form (JRRP) is usually diagnosed under 5 years of age and is more aggressive than the adult form. RRP is caused by the human papilloma virus (HPV) and is associated with maternal condylomata acuminata (genital warts), with the same
subtypes, 6 and 11, being most commonly responsible for both RRP and genital warts (Derkay and Wiatrak 2008). Although many newborns are likely exposed to HPV, only a few develop JRRP and the clinical course of the disease varies from one child to another. HPV-11 genotype and younger age at onset of JRRP are predictors of increased disease severity (Niyibizi et al. 2014).

After changes in voice and hoarseness, stridor is the second most common presenting symptom, first inspiratory, and then biphasic. Dyspnea, acute respiratory distress, and chronic cough may also occur. The average time from onset of the symptoms to diagnosis ranges from 1 to 8 years, as the affected children may be erroneously diagnosed with recurrent croup, asthma, hemangioma, or tracheomalacia. RRP typically arises from the larynx, but supraglottic or subglottic extension is frequent, and the entire respiratory tract may be involved (in less than 2 % of patients). Distal spread may result in the formation of multiple lung nodules, which may cavitate.

The diagnosis is based on endoscopic findings and biopsy. RRP appear as irregular, cauliflower-like pedunculated masses that consist of a vascular connective tissue core covered by stratified squamous

**Fig. 46** Subglottic hemangioma. Frontal radiograph shows a short segment asymmetric narrowing of the airway (arrow) caused by soft tissue just inferior to the true vocal cords.

**Fig. 47** Subglottic hemangioma. (a) Axial T2-weighted image with fat saturation reveals a very bright midline mass lesion (arrow) along the posterior wall of the airway at the level of the cricoid cartilage. (b) Postcontrast T1-weighted image with fat saturation at a similar level demonstrates strong lesion enhancement (arrow) (Case courtesy of A. Tumu and M. Maheshwari, Children’s Hospital of Wisconsin, Medical College of Wisconsin, Milwaukee).
epithelium with abnormal keratinization. The disease is frequently recurrent, with an average of almost 20 surgical procedures (such as debulking with microdebriders and balloon dilation) per child. When surgical therapy is needed more frequently than four times in 12 months or there is disease outside the larynx, adjuvant medical therapy should be considered. Various intralesional or systemic medical treatments include cidofovir, quadrivalent HPV vaccine, and bevacizumab (Sidell et al. 2014). Although basically benign, RRP causes significant morbidity and occasional mortality. The natural history of RRP is quite variable and unpredictable, including spontaneous regression, persistence and dissemination of lesions causing airway compromise, and in rare instances progression to carcinoma (<1 % in JRRP, usually HPV-11 genotype) (Derkay and Wiatrak 2008).

**Imaging Studies**

RRP is characterized on plain films by nodular, sometimes pedunculated, soft tissue masses involving the laryngeal and tracheal air column. Distention of the air in the oropharynx above the soft tissue masses may be present (Castillo and Mukherji 1996a).

Cross-sectional imaging plays no role in the initial diagnostic workup, but is useful for follow-up of extralaryngeal spread of RRP. CT is the modality of choice for evaluating pulmonary extension. On CT, these lesions appear as multiple nodular soft tissue masses that narrow the airway (Figs. 48 and 49). These lesions may enhance with contrast. Larger recurrent lesions may spread transbronchially, with subsequent formation of lung masses and cavities. The pulmonary findings consist of multiple bilateral parenchymal nodules, which may be solid or cavitory (Fig. 48), and solid lesions may eventually cavitate. An enlarging solid lesion is suggestive of malignant transformation to squamous cell carcinoma (Castillo and Mukherji 1996a; Derkay and Wiatrak 2008; Niyibizi et al. 2014).
Fibromatosis Colli

Fibromatosis colli (sometimes referred to as sternocleidomastoid or sternomastoid tumor of infancy) is an uncommon benign congenital fibrous pseudotumor of the sternocleidomastoid muscle, which presents clinically as a firm, well-circumscribed mass and is characterized by diffuse or focal muscle enlargement. The condition is characteristically seen within the first weeks of life and is often associated with a history of birth trauma. It is a self-limiting process that frequently does not require therapeutic intervention, but in some patients tightening of the muscle results in torticollis (Fig. 50). Management includes observation and physiotherapy to prevent or reverse torticollis and the craniofacial asymmetry that can result. Physiotherapy provided by parents also involves them directly in the care of their newborn (Lowry et al. 2010).

Imaging Studies

US, CT, and MRI demonstrate a fusiform mass localized to the sternocleidomastoid muscle, with clear surrounding fascial planes and without lymphadenopathy or any other associated abnormalities (Figs. 50 and 51). The signal intensity of the lesion may be slightly lower on T2-weighted than on T1-weighted images due to the presence of fibrous tissue within the mass (Ablin et al. 1998).

Aggressive Fibromatosis

Aggressive fibromatosis (AF), also referred to as desmoid tumor, is a rare monoclonal, fibroblastic proliferation arising in musculoaponeurotic structures. AF is a tumor of intermediate malignancy, with a strong potential for local invasiveness and recurrence. It is most commonly found in the abdomen, while it originates from the head and neck in less than a quarter of cases, typically around the mandible. The tumor generally presents as painless swelling during the first 2 years of life, often fixed to the underlying tissues, but not to the skin. It infiltrates adjacent muscles and vessels and destroys the bones. Histologically, it may be confused with low-grade fibrosarcoma and is associated with a postsurgical local recurrence in up to 70 % of cases.

Recently collected data suggest that these tumors warrant a wait-and-see strategy (clinical-radiological observation, without any treatment), since their natural history is often characterized by lengthy periods of
**Fig. 50** Fibromatosis colli. (a, b) Contrast-enhanced axial CT images at different levels show a nonenhancing homogeneously enlarged left sternocleidomastoid muscle (SCM) (arrows), which is isointense with the normal contralateral SCM (arrowhead). (c) Reconstructed coronal CT image again shows thickened but otherwise unremarkable left SCM, as well as left-sided torticollis.

**Fig. 51** Fibromatosis colli. (a) US shows a diffusely swollen SCM of homogeneous echogenicity (arrow). (b) Contrast-enhanced axial CT scan shows that the enlarged right SCM (arrow) is homogeneous and of normal attenuation values.
stability or even regression, and that treatment should be reserved for tumors with aggressive growth or serious symptoms (Peña et al. 2014).

**Imaging Studies**

On CT, aggressive fibromatosis appears homogeneous and slightly hyperattenuating relative to adjacent normal muscles and shows contrast enhancement (Fig. 52). The masses are typically of an elongated shape with a rim of surrounding fat. MRI demonstrates lesions that are isointense to muscle on T1-weighted images, usually T2 hyperintense, and with strong enhancement after contrast administration. Characteristic nonenhancing low-signal-intensity bands on all MR sequences correlate with dense collagenous stroma (Fig. 53) (Rhim et al. 2013). These findings, along with the absence of peritumoral edema in a mass that infiltrates the surrounding tissues, are highly suggestive of the diagnosis (Flacke et al. 1999).

**Lipoblastoma and Lipoblastomatosis**

Lipoblastoma and lipoblastomatosis are rare benign tumors of embryonic fat that occur in infancy and early childhood, usually arising from the limbs or the trunk. The embryonic fat-cell precursors, the lipoblasts, may still be normally found after birth in the axilla, mediastinum, retroperitoneum, and prevertebral space. These areas are the most common sites for the occurrence of lipoblastomas (Hussein et al. 1999). Two thirds of these lesions are focal and encapsulated lipoblastomas, whereas the remaining one third are diffuse and locally infiltrative, hence the term lipoblastomatosis. Patients with focal lipoblastoma are unlikely to require further surgery after initial resection. Patients with lipoblastomatosis may have recurrent disease and should undergo close follow-up (Bruyeer et al. 2012).

**Imaging Studies**

Radiographically, these are nonenhancing septate lesions containing areas of fat (Fig. 54). With MRI, on T1-weighted images this embryonic fat is bright, however of lower signal compared to the characteristically hyperintense normal fat (Castillo and Mukherji 1996a). These features may allow differentiation from lipoma (Fig. 55) on imaging studies. Infiltration of the surrounding soft tissue and fascial planes may be present in the diffuse form. Large, advanced lesions may extend into the mediastinum and displace surrounding structures. It appears that, after chemotherapy, increase in T1 signal occurs, consistent with maturation of lipoblasts (Hussein et al. 1999; Bruyeer et al. 2012). Recurrent lesions are best imaged with MRI to assess extent and plan reconstruction if necessary.
Neurogenic Tumors

Neurofibromas and schwannomas are by far the most common neurogenic tumors arising in the neck (Weber et al. 2000).

**Neurofibroma**

Neurofibromas are almost exclusively found in patients with neurofibromatosis type 1 (von Recklinghausen’s disease). These tumors are often multiple and usually associated with other cutaneous and systemic manifestations of the disease. They are neoplasms of Schwann cells and axons, which infiltrate and expand the nerves, characteristically in the neural foramina of the spine but may also be found in the neck (Fig. 56), along the skull base, or in the parotid region. MRI is the best modality for defining the extent of these lesions, which are isointense to muscle on T1-weighted images and hyperintense on T2-weighted images and enhance variably (Fig. 57). The use of fat suppression is recommended as the tumors frequently spread into fat-containing spaces, most commonly the parapharyngeal space (Weber et al. 2000).

**Schwannoma**

Schwannomas are less common than neurofibromas. They may also originate in the neck, most frequently along the nerves in the carotid space, and may rarely occur at multiple sites. These tumors are typically
well-defined tubular structures that show intense contrast enhancement, both on MRI and CT (Fig. 58). They are usually of intermediate signal intensity on T1-weighted images, T2 hyperintense, bright on ADC maps, and of low density on CT. They frequently have a heterogeneous appearance due to cystic (Fig. 59) or fatty degeneration. Schwannomas may be associated with neurofibromatosis type 2.

**Neuroblastoma**

Neuroblastoma (NB) is the most common extracranial pediatric solid malignant tumor and the most common neoplasm in infancy. Children 2–4 years of age are most frequently affected, and 90 % of cases present before 10 years of age. The vast majority of NB involving the head and neck are metastatic lesions from primary abdominal tumors, with only a small fraction of primary tumors. The most common metastatic sites include the skull, orbit, and cervical lymph nodes. NB arises from undifferentiated neural crest cells, which are considered the precursors of the sympathetic nervous system. Cervical involvement usually occurs along the course of the sympathetic chain. Affected individuals characteristically present

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**Fig. 54** Lipoblastoma. (a) Coronal T1-weighted image shows a left lateral neck well-circumscribed mass (arrow) that is very bright, slightly less hyperintense compared to the subcutaneous fat. The lesion also contains internal septations (arrowheads). (b) With fat saturation pulse on this axial postcontrast T1-weighted image, the nonenhancing mass is again slightly darker compared to the adjacent subcutaneous fat. The mass is displacing the submandibular gland (arrow) (Case courtesy of P. Tortori-Donati, Genoa, Italy)

**Fig. 55** Lipoma. Contrast-enhanced axial CT image demonstrates a well-defined left neck mass (arrow) that is isointense with the subcutaneous fat and without internal septations.
Fig. 56 Neurofibromas in a patient with neurofibromatosis type 1. Coronal STIR image through the neck and cervical spine shows multiple mildly hyperintense heterogeneous lesions, some of which are extending through the left neural foramina.

Fig. 57 Neurofibroma, sporadic. (a) Contrast-enhanced axial CT scan shows a well-defined hypodense nonenhancing mass (arrow) arising from the right carotid space and displacing the vascular structures. (b) Coronal STIR image shows mildly increased signal intensity of the homogeneous lesion (arrow). (c) Axial T1-weighted image at a level similar to the CT scan demonstrates isointensity of the mass compared to muscles. (d) Corresponding postcontrast T1-weighted image shows minimal and patchy lesion enhancement (arrow).
with a firm, nontender neck mass and unilateral Horner’s syndrome or heterochromia of the iris. The prognosis of these tumors has been increasingly favorable (Abramson et al. 1993).

**Imaging Studies**

In the head and neck, NB usually presents either as a large cervical lymph node or a solid bone metastasis in the orbit or temporal bone, sometimes bilateral, which is not infrequently the initial presentation of NB. The tumor demonstrates vascular compression and frequently extensive bone destruction, commonly with a prominent spiculated new bone formation resembling osteosarcoma (Fig. 60). NB is of increased signal intensity on T2-weighted images and shows contrast enhancement on MRI and CT. Similar to other small blue cell tumors, it has characteristically very low signal on ADC maps, due to decreased diffusion of water molecules (Fig. 60). This is in contrast with ganglioneuroma, which shows increased diffusion with high ADC signal (Gahr et al. 2011). Internal calcification and growth along the expected course of the sympathetic chain are sometimes found and are indicative of the diagnosis (Fig. 61).
Primary paraspinal tumors in the neck arise along the spinal column, where they may invade the intervertebral foramina and produce cord compression (Fig. 62) (Siegel and Jaju 2008).

Lymphomas

Lymphomas are the most common neoplasms of the head and neck in the pediatric population, accounting for 10–15 % of all pediatric malignancies. They are highly curable, with 5-year survival rates of up to 95 % for Hodgkin lymphoma and 82 % for non-Hodgkin lymphoma (NHL). These results have focused recent attention on reducing the burden of treatment-related morbidity while maintaining the excellent outcomes. As the late effects of radiotherapy, including secondary tumors, were recognized, successive protocols were developed that reduced its use, so that the current treatment protocols for NHL are chemotherapy based with radiotherapy virtually eliminated. In contrast, current pediatric Hodgkin lymphoma protocols continue to use radiotherapy (Frew et al. 2013).

Hodgkin Lymphoma

Hodgkin lymphoma (also Hodgkin disease, HD) is a lymphoproliferative malignancy predominantly affecting adolescents and young adults and is rarely seen in children under 5 years of age. Males are more frequently affected than females (2:1). In 95 % of patients, HD arises in lymph nodes. The region of

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Fig. 60  Metastatic neuroblastoma. (a) Contrast-enhanced axial CT image shows an osteolytic mass of the right temporal bone with spiculated new bone formation (arrow). (b) Axial ADC map at a similar level reveals low signal intensity of the mass (arrow), which is darker than the brain.

Fig. 61  Neuroblastoma. Contrast-enhanced axial CT image reveals a mildly enhancing mass (arrow) in the lower right neck containing small calcifications (arrowhead). The abnormality is found along the location of the sympathetic chain. The finding could correspond to both metastatic and primary neuroblastoma.

(Abramson et al. 1993). Primary paraspinal tumors in the neck arise along the spinal column, where they may invade the intervertebral foramina and produce cord compression (Fig. 62) (Siegel and Jaju 2008).
Waldeyer’s ring is rarely involved in patients with HD. While extranodal primary sites are unusual, systemic involvement may result from progression of disease.

**Non-Hodgkin Lymphoma**

NHL is, unlike HD, most commonly seen in children between 2 and 12 years of age. Predisposing conditions that increase the likelihood of developing NHL include various congenital and acquired immunodeficiency states, and males are more commonly affected. In contrast to HD, extranodal sites are much more common and, in the head and neck, include the nasopharynx, parotid, skin, and bone. The majority of patients with NHL have advanced disease at the time of presentation. More than 90 % of pediatric childhood NHLs are high-grade lymphomas belonging to four major histologic subtypes: Burkitt lymphoma/leukemia (BL), diffuse large B-cell lymphoma, lymphoblastic lymphoma, and anaplastic large cell lymphoma.

BL is almost exclusively seen in pediatric patients and is also the most common (40 %) NHL type to occur in children and adolescents. The endemic form is characterized by a distinct geographic distribution in Africa. Almost all patients with this form have high antibody titers to Epstein-Barr virus (EBV), with the face and jaw being the most commonly affected areas. In contrast, the sporadic (American) form of BL does not have a strong link with EBV, and facial and mandibular masses are rare (Frew et al. 2013).

**Imaging Studies**  CT is generally the preferred technique for initial evaluation of patients with nodal masses and MRI for extranodal lesions within the head and neck. Involved lymph nodes are homogeneous and variable in size, with an average diameter from 2 to 10 cm. They may enhance slightly to moderately, or even not at all (Figs. 63 and 64); they may display necrosis before and after treatment and contain calcification posttreatment. The presence of necrosis before treatment is uncommon even in large nodes. Lymphadenopathy is found in multiple locations, usually with a dominant large node or a cluster of nodes. Differentiation of lymphoma from other causes of lymphadenopathy is not reliable with conventional cross-sectional imaging.
Involvement of the extranodal soft tissues is characterized by a homogeneous mass slightly hyperintense on T2-weighted images, most commonly with mild postcontrast enhancement (Fig. 65). It typically demonstrates low signal on ADC maps, corresponding to reduced diffusion of water molecules. The margins of the lesion may be either well defined or, less commonly, spreading into the surrounding structures. Characteristically, the adjacent bone is remodeled from chronic mass effect, although a more aggressive pattern of bone destruction may occasionally be found (Castillo and Mukherji 1996a; Abramson and Price 2008).

PET/CT has become the modality of choice for both staging and treatment response evaluation in patients with lymphoma, as it shows metabolic activity of the lesions and not just their size (Fig. 66). Today, FDG PET is used for tailoring treatment intensity in children with HD within the framework of international treatment optimization protocols. In contrast, the role of this method in children with NHL is not well defined (Kluge et al. 2013). Efforts are now largely focused on decreasing the diagnostic radiation exposure to these patients, by utilizing reduced or radiation-free modalities, such as PET/MRI and diffusion-weighted whole-body MRI, as well as on refining the surveillance imaging strategies (Littooj et al. 2014).
Fig. 65 Burkitt lymphoma. Postcontrast sagittal T1-weighted image shows a well-defined mildly enhancing mass arising from the palatine tonsil (*asterisk*) that deforms and compresses the adjacent structures.

Fig. 66 Burkitt lymphoma. (a) A large mass (arrow) that cannot be separated from the left SCM is seen on this contrast-enhanced axial CT image. There are areas of enhancement within the lesion (arrowhead). (b) Coronal PET/CT image from the neck acquisition shows a very high FDG uptake (corresponding to metabolic activity) within the lesion. There is a central area without metabolic activity. (c) Coronal PET/CT image from the body acquisition reveals multiple additional osseous lesions (arrows) with very high FDG uptake.
Sarcomas and Carcinomas

Rhabdomyosarcoma

Rhabdomyosarcoma (RMS) is the second most common malignancy and the most common soft tissue malignant tumor in the pediatric head and neck, where 35–40% of cases occur. There is a bimodal peak for age of onset (2–5 years and 15–19 years) and a sex predilection for males (2:1).

Histologically, RMS has been classified into four separate types. The embryonal and botryoid forms are more common in infants and children, whereas the alveolar pattern is seen predominantly in adolescents and has the worst prognosis. The fourth variety, the well-differentiated pleomorphic form, is usually found in adults. Embryonal (ERMS) and alveolar (ARMS) are the two major RMS subtypes that are distinct in their morphology and genetic makeup. The prognosis depends strongly on tumor size, location, staging, and child’s age. In general, ERMS has a more favorable outcome, whereas the mortality rate remains high with ARMS, due to its aggressive and metastatic nature. The most common location of RMS is the orbit; it also occurs in the masticator space and other locations. These tumors can arise in any tissue, except for the bone. Head and neck RMS can be divided into parameningeal, orbital, and nonorbital-nonparameningeal. Orbital RMS carries the best, while parameningeal involvement has the poorest prognosis, and its presence should be detected preoperatively (Robson 2010; Lloyd and McHugh 2010). The treatment typically involves a combination of surgery, radiation, and chemotherapy. Advancements in surgical and radiotherapy techniques have reduced patient morbidity, whereas new chemotherapeutic protocols have improved local disease control and overall survival (Gillespie et al. 2006).

Other Sarcomas

Fibrosarcoma (FS) is the second most common pediatric sarcoma, with 15–20% of cases involving the extracranial head and neck at the sinonasal region, face, cheek, larynx, and hypopharynx. Metastatic spread is infrequent. Osseous sarcomas, primarily Ewing’s sarcoma, have a predilection for the face and jaw of children. Patients with advanced forms of neurofibromatosis are at increased risk for developing neurofibrosarcoma. These lesions tend to be more aggressive in patients affected with neurofibromatosis compared to those that occur sporadically.

Nasopharyngeal Carcinoma and Squamous Cell Carcinoma

Unlike adults, in whom this neoplasm accounts for 80% of head and neck malignancies, squamous cell carcinoma (SCCA) comprises only 4% of lesions involving the upper aerodigestive tract in children, with the nasopharynx as the most common site. Similar to Burkitt lymphoma, nonkeratinizing nasopharyngeal carcinoma (NPC) has a strong serologic association with Epstein-Barr virus. At times, histologic differentiation among SCCA, neuroblastoma, and NHL may be difficult. The majority of patients have cervical nodal metastases at the time of presentation, frequently including level 5 nodes. Nasopharyngeal carcinoma may also demonstrate intracranial spread either by direct invasion of the skull base, extension along the carotid artery, or perineural spread along the third division of the fifth cranial nerve.

Thyroid Carcinoma

The incidence of thyroid carcinoma (TC) has significantly decreased since the association between TC and radiation exposure was first noted in the 1950s. However, children who are exposed to low doses of radiation are still at increased risk for both benign and malignant neoplasms of the thyroid gland. Cold nodules on 123I thyroid scan in children have a higher frequency of malignancy than in adults, up to 29%. TC is also one of the secondary neoplasms that can occur in patients previously treated for malignancy.
**Imaging Studies**

The radiographic findings of various sarcomas and carcinomas are often nonspecific. Radiographically, these tumors are aggressive lesions, which often infiltrate and invade surrounding soft tissue and bony structures. The lesions are often solid and demonstrate postcontrast enhancement. Both benign and malignant tumors may enhance with contrast; therefore, this is not a reliable differentiating characteristic. RMS is usually hyperintense on T2-weighted images and may have fairly well-defined margins, presumably due to its rapid growth (Figs. 67 and 68). It shows a variable internal structure and enhancement, with characteristic internal ring-enhancing areas resembling bunches of grapes that may be seen in some cases (Hagiwara et al. 2001). RMS frequently arise from the muscles, so a masticator space mass in a pediatric patient is most likely RMS, unless it arises from the bone, in which case Ewing’s sarcoma should be considered. Focal areas of cortical destruction are frequent with Ewing’s sarcoma, commonly seen as subtle channels extending through the cortex at CT or MR imaging, a finding that reflects the underlying pathologic appearance. A sharp and defined margin of the bone lesion, optimally visualized on T1-weighted images, is the most significant feature of Ewing’s sarcoma in distinction from osteomyelitis (Fig. 69) (Henninger et al. 2013). Benign fibro-osseous lesions may also present as large masses arising from the jaw. The distinguishing imaging features include sharp margins and ground glass appearance on CT (Fig. 70).

The important role of imaging is in precisely defining the extent of a lesion and other possible associated findings, such as lymphadenopathy. MRI is the preferred technique for evaluation of the upper aerodigestive tract tumors, due to excellent soft tissue detail and multiplanar capabilities (Fig. 71). MRI is clearly superior to CT in detecting perineural tumor spread and may also be more reliable in

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**Fig. 67** Rhabdomyosarcoma (RMS). (a) Coronal STIR image shows an apparently well-circumscribed, very bright mass (arrow) in the left masticator space. (b) Sagittal T1-weighted image through the lesion demonstrates its isointensity (arrow) and extension into the foramen ovale (arrowhead). This finding is consistent with parameningeal RMS. (c) Axial CT image with bone window again demonstrates widening of the left foramen ovale (arrowhead). (d) Postcontrast axial T1-weighted image shows a relatively mild enhancement of the lesion (arrow).
detection of laryngeal cartilage and tracheal wall invasion. Thin-section CT with bone algorithm is the imaging modality of choice for early detection of skull base erosion. CT may also be superior to MRI for the assessment of nodal involvement (Castillo and Mukherji 1996a; Robson 2010; Lloyd and McHugh 2010).

**Inflammatory Disorders**

**Adenotonsillar Hypertrophy**
Adenotonsillar hypertrophy is considered the most common cause of upper airway obstruction in children. It typically involves the palatine tonsils and the adenoids. Lymphoid tissue grows rapidly in the first few years of life, and the tonsils peak in size between 4 and 7 years of age. Enlargement of the adenotonsillar tissues can be either due to idiopathic hyperplasia or secondary to inflammatory stimuli.
The symptoms are mainly due to the mass effect leading to airway obstruction and blockage of eustachian tube drainage. Acute manifestations include mouth breathing, nasal congestion, snoring, and obstructive sleep apnea. Adenotonsillectomy is performed when conservative treatment is not successful (Tumu et al. 2014).

**Fig. 69** Ewing’s sarcoma. (a) Axial CT image with bone window demonstrates an irregular expansile lesion (arrow) of the left mandible. (b) Axial T1-weighted image shows loss of normal bone marrow signal, which is replaced by the mass, along with cortical destruction. (c) Coronal STIR image shows heterogeneous hyperintensity of the lesion, which (d) demonstrates enhancement (arrow) on coronal postcontrast T1-weighted image.

**Fig. 70** Ossifying fibroma. Contrast-enhanced axial CT image reveals that the right mandibular ramus is replaced by a well-circumscribed round mass lesion with peripheral calcification and internal areas of ground glass appearance.
Infectious Cervical Adenitis

Enlarged cervical lymph nodes in children are most commonly due to an associated infectious process. Infectious agents spread into the surrounding tissues and drain into the afferent lymphatic channels of the lymph nodes, leading to activation of lymphocytes with eventual enlargement of the nodes. The activated lymph nodes are often referred to as reactive nodes, whereas suppurative adenitis stands for an infected node that has undergone liquefaction necrosis. The likelihood of developing cervical adenitis, especially suppurative forms, decreases with age. Upper respiratory viral infections are the most common cause of lymph node enlargement in children. Bacteria are the usual culprits for suppurative cervical adenitis, with *Staphylococcus aureus* and Group A *Streptococcus* as the leading etiological agents (Castillo and Mukherji 1996a). Unlike *Mycobacterium tuberculosis*, surgery appears to be the treatment of choice for adenitis caused by other mycobacteria (Caruso et al. 2009).

The syndrome of periodic fever, aphthous stomatitis, pharyngitis, and cervical adenitis (PFAPA) is the most common periodic fever disease in children. The exact pathogenesis of this autoinflammatory syndrome is unknown, and the overall prognosis is excellent. On the other hand, surgery leads to immediate complete resolution of symptoms in almost all patients, so that tonsillectomy and adenoidectomy may be offered as part of the treatment options (Licameli et al. 2012).

Imaging Studies

Contrast-enhanced CT is the preferred imaging modality for the evaluation of suspected suppurative cervical lymphadenitis. In addition to shorter acquisition time, reduced cost, and better overall patient compliance, the appearance of involved lymph nodes may at times be confusing on MRI. If lymph nodes are to be evaluated with MRI, postcontrast T1-weighted images with fat saturation should be included, as they seem to be the most reliable.

In general, the cervical lymph nodes in children are larger than in adults and normal internal jugular chain nodes may reach 25 mm in diameter. Intact retropharyngeal nodes are seen in most pediatric patients as round homogeneous structures of the same signal intensity as the other lymphoid tissues, mildly hyperintense on T2-weighted images. Infection is an extremely unusual cause of enlarged facial or parotid nodes. Enlarged facial lymph nodes can occur in patients with lymphomas or recurrent tumors that drain to these nodes. Enlarged parotid lymph nodes are usually seen with underlying systemic abnormalities or malignant neoplasms (Castillo and Mukherji 1996a).
Early involvement by infection is characterized by homogeneous enlargement, loss of the fatty hilum, and increased enhancement of the involved lymph node. Liquefaction necrosis within the center of enlarged nodes gives an appearance of a low attenuation focus surrounded by enhancement (Fig. 72). Low attenuation is also present in an infected node in the stage prior to liquefaction; this phase has been termed the presuppurative phase, and attempted aspiration of contents at this stage is unsuccessful. Patients affected with mycobacterial infections characteristically present with multiple matted bilateral nodes in the lower neck and frequently have associated pulmonary disease and hilar adenopathy.

**Deep-Neck Infections**

Pediatric deep-neck infections involving the retropharyngeal or parapharyngeal spaces are potentially life-threatening processes that may be secondary to lymphadenitis, due to parapharyngeal spread of a tonsillar abscess, or from direct traumatic inoculation. The characteristic symptoms are fever, dysphagia, limited range of motion in the neck, neck mass, and a leukocyte count greater than $15 \times 10^9/L$. Tonsillar abscess is most commonly seen in teenagers, while retropharyngeal infection almost exclusively occurs in young children, under 10 years of age (Chang et al. 2010).

**Imaging Studies**

The radiologic evaluation should determine the size and location of the infection and attempt to distinguish whether the process is an inflammatory mass (also known as cellulitis or phlegmon) or an abscess. The initial imaging modality has traditionally been a lateral neck radiograph. The presence of prevertebral widening, loss of the normal cervical lordosis, and air in the prevertebral soft tissues are suggestive of an underlying infection. However, the location and the extent of the process cannot be determined, and abscess cannot be differentiated from cellulitis. The sensitivity and specificity of radiographs for deep-space infections are also relatively low (Fig. 73).

Contrast-enhanced CT permits precise localization of infections involving the deep neck; however, it is not very reliable for differentiation between abscess and cellulitis (Smith et al. 2006; Malloy et al. 2008). Cellulitis typically has an appearance of swelling or loss of parapharyngeal soft tissue planes (Fig. 74), while a homogeneous area of low attenuation surrounded by ring enhancement is suggestive of an abscess (Fig. 75). The sensitivity of CT for abscess is very high (in the range of 81–100 %), but abscesses may be overcalled, as the specificity is much lower (53–80 %). The overall accuracy of CT for a drainable...
The accuracy of CT in distinguishing a drainable collection from inflammatory mass is particularly poor in the retropharyngeal space with reported specificity of even less than 50% (Cheng and Elden 2013). This may be due to a relatively rare abscess formation with necrotizing retropharyngeal adenitis in children. It seems that wall irregularity may be a better predictor of the presence of pus than contrast enhancement. Although narrowing of the ipsilateral internal carotid artery may seem dramatic on imaging studies, it typically represents a benign common finding (Boucher et al. 1999). A potential space found anterior to the retropharyngeal space is often referred to as the “danger space” and is inferiorly continuous with the posterior mediastinum. Infections in the retropharyngeal space may enter the danger space because of their close proximity and then extend into the mediastinum (Harnsberger 1995). The inferior extent of infections involving the retropharynx is best determined with MRI (Fig. 77). It is the technique of choice for assessing the epidural space involvement in pre- and paravertebral space infections and complements CT in the evaluation of infections reaching the skull base.
Epiglottitis (Supraglottitis)

Epiglottitis is an emergency as it can lead to life-threatening airway obstruction. This acute supraglottic infection used to be caused by *Haemophilus influenzae* type B; however, its incidence has decreased thanks to vaccination. Epiglottitis is now typically related to nontypable *Haemophilus influenzae*, *Haemophilus parainfluenzae*, *Staphylococcus aureus*, and *Streptococcus pneumoniae* (Loftis 2006).

Epiglottitis is, in fact, a systemic disease with supraglottic involvement being a major feature of the process. This disease usually occurs in children between 3 and 6 years of age, although recent reports suggest that its incidence is increasing in younger patients. The newborn is protected because of immunity to the capsular antigen acquired from the mother. This passive immunity resolves by 3 months of age, and the child’s own immune system does not produce significant amount of antibodies until 3–4 years after
birth. This window of diminished antibody levels partly explains the high incidence of infections in this age group.

Patients with epiglottitis characteristically present with rapid onset of symptoms, including high fever, sore throat and respiratory distress, difficulty swallowing, muffled voice, often with sitting upright, and drooling. Blood cultures are frequently positive in these patients. Swelling of the epiglottis, aryepiglottic folds, and false vocal cords is characteristically found on clinical examination, with resultant narrowing of the supraglottic airway. The secretions in patients with epiglottitis are often thick and tenacious, and mucous plugging of an already compromised airway may cause sudden respiratory arrest. Airway manipulation should be avoided because it can lead to complete airway occlusion from further irritation. Management typically includes rapid intubation, intravenous antibiotics, and steroids (Tumu et al. 2014; Castillo and Mukherji 1996a; Loftis 2006).

**Imaging Studies**

A lateral radiograph of the soft tissues of the neck is used to confirm the diagnosis and exclude an aspirated foreign body. However, radiographs may be of little value in the primary evaluation of epiglottitis. If imaging is to be performed in the radiology department, personnel able to perform intubation should accompany the patients. There is no role for CT and MRI at initial presentation. Furthermore, these studies may actually be life-threatening as placing the patient in the supine position will further reduce the caliber of an already narrowed airway. Radiographs show a thickened epiglottis that is often two to three times increased in size (Fig. 78). Swelling of the free edge of the epiglottis is a characteristic finding, often referred to as the thumb sign. The aryepiglottic folds are also thickened, and the oropharynx may be overdistended (Tumu et al. 2014; Loftis 2006).

**Croup (Laryngotracheobronchitis)**

Croup (laryngotracheobronchitis, laryngotracheitis) is the most common cause of pediatric stridor. This acute upper respiratory tract infection is usually of viral etiology (most commonly parainfluenza type 1) and occurs in children between 6 months and 3 years of age. The onset is gradual, and patients typically present with a characteristic barking “seal-like” cough and stridor. The infection results in diffuse edema of the mucosa of the subglottic region and trachea. Two characteristics of the subglottitis make it especially
vulnerable: this is the narrowest portion of the respiratory tract in children under 3 years of age and the only portion of the upper respiratory tract surrounded by a complete cartilaginous ring. Croup is typically managed conservatively but may rarely require intubation, as severe cases may progress to complete laryngeal obstruction. Treatment usually consists of a single-dose oral corticosteroid or nebulized epinephrine in more severe cases (Tumu et al. 2014; Castillo and Mukherji 1996a; Loftis 2006).

Imaging Studies
Imaging is usually not needed for diagnosis, but it helps confirming the clinical suspicion. The imaging method of choice is frontal and lateral radiographs of the neck. The additional role of imaging is to exclude other causes of stridor, such as bacterial epiglottitis or a foreign body, as tracheal irregularity. It should be kept in mind that the radiographs have reportedly low sensitivity and specificity and should therefore always be interpreted in the context of a clinical setting.

The characteristic plain film findings are the loss of normal subglottic angles with a long segment of airway narrowing, resulting in the classic steeple sign/inverted V appearance of the subglottic region on frontal radiographs (Fig. 79). Lateral radiographs are helpful for excluding other causes of stridor; in addition to epiglottitis, intraluminal membranes and tracheal irregularity may be seen in patients with exudative tracheitis (Tumu et al. 2014; Castillo and Mukherji 1996a).

Trauma
Laryngotracheal trauma (LTT) is uncommon in children, but if undiagnosed it may be life threatening. Most cases of LTT involve the subglottic and upper tracheal regions. The glottic region, which includes the true vocal cords and arytenoid and thyroid cartilages, is the second most common location. Injuries of the supraglottic structures including the false vocal cords, aryepiglottic folds, epiglottis, and hyoid bone are rare in children. This is due to the fact that, in infants and young children, the larynx is more mobile and flexible and in a higher position than in adults. Thus, the mandible acts as a protective agent against crush injuries involving the anterior neck. With age, the larynx descends in the neck and the incidence of
supraglottic injuries increases in older children. LTT has been classified into internal and external types of injury, with different causative factors and demographics (Castillo and Mukherji 1996a; Hackett et al. 2012).

**Internal Injuries**
The most common cause of internal LTT is prolonged endotracheal intubation. Acute injuries consist of edema, erosions, and ulcerations and usually occur at the medial surface of the arytenoids, the cricoarytenoid articulation, the posterior commissure, and the anterior surface of the cricoid lamina. Traumatic intubation may also lead to dislocation of the arytenoid cartilages and cricoid fracture. Scarring and fibrosis may accompany healing and reepithelialization of the injured laryngeal mucosa. Intubation granulomas, glottic webs, submucosal cysts, and vocal cord immobility may eventually develop. Endotracheal intubation is the most common cause of acquired subglottic stenosis. Stenosis caused by prolonged intubation may develop immediately after extubation or may be delayed by as much as 90 days. Stenosis caused by tracheotomy is of similar appearance and typically seen 1–2 cm distal to the stoma. Other causes of internal injury include ingestion of caustic substances (toxic smoke inhalation) and aspiration of foreign bodies. Affected patients typically present with stridor, hoarseness, cough, dyspnea, and difficulty in clearing secretions (Hackett et al. 2012).

**Imaging Studies**
Internal injuries with laryngeal stenosis are best imaged with CT (Figs. 80 and 81), and intravenous contrast is not needed. Characteristic CT findings are partial or circumferential soft tissue thickening involving the subglottis and/or trachea. Important information includes the caliber of the residual lumen and the extent of the involvement, and this may be well shown by virtual endoscopy (Fig. 81). Deformity of the air column or dystrophic calcifications of the cricoid cartilage or tracheal rings are suggestive of associated laryngomalacia or tracheomalacia.

Frontal and lateral neck radiographs are obtained in patients with clinical presentation consistent with a laryngotracheal foreign body. A significant proportion of the foreign bodies, most commonly food particles such as peanuts, may be radiolucent and not seen on radiographs. Indirect signs of aspiration,
such as subglottic swelling and airway narrowing, may indicate foreign body location in those instances (Tumu et al. 2014; Castillo and Mukherji 1996a).

**External Injuries**

External trauma represents a spectrum of injuries from ecchymosis to major laryngotracheal injury. The risk of laryngotracheal injury is higher with blunt trauma, and the most common cause is motor vehicle accidents. In cases of direct blow, the larynx is crushed between the inciting force and the rigid spine, resulting in fracture of the cartilages or rupture of the membranous tracheal wall. Tracheal injury may also result from severe hyperextension of the cervical spine, causing a marked increase in the intraluminal pressure, which, if severe enough, may result in laceration or transection. Severe forms of trauma are often
associated with injuries to one or both recurrent laryngeal nerves or esophageal lacerations. Penetrating oropharyngeal injury is much less common and usually occurs when a child falls with a foreign body, such as a pencil or lollipop, inside the mouth. The resultant blow to the lateral soft palate can compress the internal carotid artery or the internal jugular vein between the foreign body and the spine (Castillo and Mukherji 1996a; Hackett et al. 2012).

Patients with external LTT injuries present with respiratory difficulties, hoarseness, and subcutaneous emphysema. Hemoptysis and odynophagia may also be present. Physical examination may reveal neck or chest contusions or lacerations and loss of the normal cricoid and thyroid prominence. Surgical exploration of the neck used to be mandatory in the management of penetrating neck injuries (with platysma penetration), resulting in a number of unnecessary operations. The clinical practice has over years moved toward selective exploration in clearly indicated cases. Imaging now plays a vital role and is widely used to limit nontherapeutic explorations, improve diagnostic accuracy, and reduce morbidity (Vick and Islam 2008).

**Imaging Studies**
In the acute setting of external trauma, radiographs may display findings suggestive of LTT, including soft tissue swelling of the involved portion of the airway, subcutaneous air, and fracture or displacement of the hyoid bone. The films should also be evaluated for the presence of foreign bodies.

CT is the study of choice in patients with external laryngeal trauma. The role of CT is to demonstrate the extent of injury, the degree of airway compromise, and the integrity of the larynx. The presence of subcutaneous air is indicative of airway laceration affecting either the larynx or trachea. Severe trauma may be associated with subluxation or fracture of the hyoid bone. The normal segmentation of the hyoid bone should not be mistaken for fractures. Severe trauma may also result in disruption of the thyroepiglottic ligaments, resulting in subluxation of the epiglottis. Other cartilaginous fractures may be very difficult to detect in the noncalcified pediatric larynx. The cricoid cartilage is the foundation of the larynx, and its status following severe trauma is one of the most important prognostic factors regarding function in the posttraumatic larynx. Cricoid fractures result in deformity and collapse of the cricoid ring, manifested as a deformity of the airway (Figs. 82 and 83). Displacement of the arytenoid cartilage is often associated with foreshortening and paramedian position of the involved true vocal cord, which sometimes makes differentiation with an isolated cord paresis difficult (Castillo and Mukherji 1996a; Becker et al. 2013). The use of 3D volume rendering with or without virtual endoscopy improves assessment of thyroid and hyoid bone fractures, arytenoid luxations, and laryngotracheal narrowing (Becker et al. 2013).
In the setting of severe penetrating oropharyngeal injury, MR angiography is the preferred modality for detection of possible associated thrombosis or dissection of the internal carotid artery or the internal jugular vein. If CT is performed, CTA with intravenous contrast should be acquired to clearly visualize the vascular structures.

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