Disorders of Salivary Glands

Douglas R. Sidell and Nina L. Shapiro

Contents

Introduction .................................................................................................................. 2
Anatomy of the Salivary Glands ........................................................................ 2
Diagnostic Evaluation ........................................................................................... 2
History ....................................................................................................................... 2
Physical Examination .............................................................................................. 2
Radiographic Imaging ............................................................................................... 3
Laboratory Studies .................................................................................................... 3
Biopsy ........................................................................................................................ 3
Sialoendoscopy ......................................................................................................... 4
Salivary Gland Pathology ....................................................................................... 4
Congenital Salivary Gland Pathology .................................................................. 4
Acquired Salivary Gland Pathology ..................................................................... 4
Mucoceles and Mucous Retention Cysts .............................................................. 6
Sialorrhea and Ptialism ............................................................................................. 7
Salivary Neoplasms .................................................................................................. 7
Malignant Neoplasms ............................................................................................... 8
Surgical Considerations ......................................................................................... 9
Conclusion and Future Directions ......................................................................... 9
Cross-References ..................................................................................................... 9
References ............................................................................................................... 9

Abstract

Primary salivary gland disorders are uncommon in the pediatric population. Infectious, neoplastic, granulomatous, and systemic disorders can occur, thereby resulting in a broad differential diagnosis. Several advances have been made over the past decade with regard to minimally invasive diagnostic and therapeutic procedures aimed to treat salivary gland pathology.
Introduction

This chapter aims to provide an overview of salivary gland pathology in children and to discuss current diagnostic and treatment modalities. A review of pertinent patient anatomy, physical examination findings, and surgical considerations is provided.

Anatomy of the Salivary Glands

The parotid salivary glands are the largest of the major salivary glands. They are located on each side of the lateral neck, resting anterior-inferior to the external auditory meatus, posterior to the mandibular ramus, and posterolateral to the masseter muscle. Each gland is composed of a deep and superficial lobe. The superficial lobe rests just below the superficial muscular aponeurotic system (SMAS), and its depth extends to the plane of the facial nerve. The facial nerve serves as the anatomic dividing point between the superficial and deep parotid lobes. The deep lobe of the parotid extends from the plane of the facial nerve superficially to the level of the stylomandibular ligament as its deep border. Also known as Stensen duct, the primary parotid duct traverses the masseter and empties its contents into the oral cavity via a ductal papilla adjacent to the second maxillary molar.

The submandibular glands are also paired and are located in the submandibular cervical triangle. This anatomic space is defined by the mandible superior laterally, the anterior digastric muscle anteriorly, and the posterior digastric muscle posteriorly. The roof of this space is the mylohyoid muscle and separates the submandibular triangle from the sublingual space. A portion of the submandibular gland extends around the posterior border of the mylohyoid, thus occupying a portion of the sublingual space. The submandibular duct is also known as the Wharton’s duct and exits the mouth adjacent to the lingual frenulum. This is easily seen on physical examination.

The sublingual salivary gland is the smallest of the major salivary glands. The sublingual gland occupies the sublingual space and is bound superficially by the oral mucosa and anterior-laterally by the mandible. The mylohyoid is the deep border of the sublingual space. The sublingual gland empties its excretory product into the ducts of Rivinus or into tributaries of the submandibular duct. The ducts of Rivinus are multiple in number and are located posterior to the opening of the Wharton’s duct, adjacent to the lingual frenulum.

Diagnostic Evaluation

History

A patient history should include the location, laterality, onset, and duration of the lesion. Pediatric salivary gland lesions may be congenital, appear shortly after birth, or manifest later in childhood. Congenital lesions include vascular anomalies such as lymphatic malformations and hemangiomas. Rapid growth or diffuse swelling in the setting of pain and fever suggests an inflammatory or infectious process and may present with bilateral tenderness and diffuse enlargement (Mehta and Willging 2006; Centers for Disease Control 1989; Cherry 2004). Other systemic disorders, including autoimmune disease, may also manifest with bilateral pathology. In contrast, slower growing, painless lesions that are isolated to a single salivary gland may indicate a neoplasm (Mehta and Willging 2006).

Physical Examination

A thorough physical examination includes inspection of the face for swelling, asymmetry, and overlying skin changes. Facial asymmetry may be the result of gland enlargement, facial nerve weakness, or both. Facial nerve weakness should always be graded and documented, and an etiology elicited. Although uncommon, facial nerve weakness in the setting of a salivary gland mass should be considered malignancy until proven otherwise.
Overlying skin changes may suggest a granulomatous process or malignancy; advanced malignancy with overlying skin changes is rare in the pediatric patient. The salivary glands should be palpated bimanually, and the salivary ducts should be compressed to express saliva. This technique allows the physician to identify isolated masses, assess tenderness, and identify obstruction or abnormal discharge from the salivary duct.

Radiographic Imaging

Salivary gland pathology in children does not always require radiographic imaging to appropriately diagnose or manage. In contrast to the adult patient, some imaging modalities may require general anesthesia to perform. This should be taken into consideration when weighing the risks and benefits of each study (Ilgit et al. 1992).

High-resolution ultrasonography has become increasingly popular as a painless, noninvasive technique for imaging head and neck pathology in children. Over the past decade, this has become the first-line imaging modality for evaluation of the parotid gland. The normal parotid gland has a homogeneous echogenicity. The facial nerve is not readily visualized, but its position can be deduced based on the location of the retromandibular vein. Lymph nodes present in the superficial parotid are easily visualized as oval or longitudinal hyperechoic masses; a longitudinal axis in excess of 6 mm or the lack of a hyperechoic hilum are abnormal findings and suggest a pathologic process (Garcia et al. 1998; Bialek et al. 2006). Inflammatory conditions, neoplastic processes, vascular anomalies, cystic lesions, sialadenosis, and salivary calculi can often be readily identified and characterized using high-resolution ultrasound (Sodhi et al. 2011). Due to the echogenicity of the mandible, the sublingual and submandibular glands are more difficult to evaluate using this technique.

High-resolution computed tomography (CT) with contrast enhancement is extremely effective in assessing salivary gland pathology and, in conjunction with magnetic resonance imaging (MRI), has effectively replaced plain radiographs and sialography. CT images allow one to distinguish lesions that arise within the salivary gland parenchyma from extrinsic pathology. CT imaging can also help characterize the behavior of a lesion by virtue of its involvement with surrounding structures such as the mandible or cervical vasculature.

Magnetic resonance imaging provides the highest level of detail of soft tissue structures and is considered the imaging modality of choice for investigating the nature of the lesion and its extent (Inarejos Clemente et al. 2018). It serves as an important imaging modality for pathology involving the deep lobe of the parotid gland and can be enhanced with gadolinium contrast dye when evaluating neoplastic, inflammatory, or vascular pathology. High-resolution 3-tesla MRI may also provide valuable information with regard to potential nerve involvement in the setting of a salivary malignancy (Mehta and Willging 2006; Freling et al. 1992).

Laboratory Studies

Laboratory studies are rarely necessary in the evaluation of salivary gland pathology. Serologic evaluation for the human immunodeficiency virus (HIV) can be performed in patients with bilateral parotid cysts (see “viral sialadenitis”). In the setting of suspected infection, material obtained by expressing ductal secretions may also be sent for culture and gram stain. Mumps titers may be performed if viral parotitis is suspected in the nonimmunized patient. Sjögren’s syndrome A and B antibodies (SS-A and SS-B, respectively) may be useful to evaluate for an autoimmune origin. Any patient with pulmonary symptoms or patients who are suspected of having extrathoracic tuberculosis may have a Mantoux TB test (also referred to as a purified protein derivative (PPD) test).

Biopsy

In adult patients who present with salivary gland masses, fine needle aspiration (FNA) is often performed in the clinic setting. In children, this procedure requires sedation under most circumstances and is uncommonly performed. FNA
allows the clinician to differentiate a salivary neoplasm from an inflammatory process and is capable of distinguishing benign pathology from malignant processes under most circumstances (Mehta and Willging 2006; Eisenhut et al. 1996).

**Sialoendoscopy**

The endoscopic evaluation and treatment of salivary disorders has gained popularity following its introduction in the 1990s. Since this time, the applications of sialoendoscopy have become more widespread and have rapidly expanded to include the pediatric population. As a diagnostic instrument, sialoendoscopy can readily identify salivary stones, ductal strictures, and chronic inflammatory processes. In the pediatric patient, some of the more common indications for sialoendoscopy include juvenile recurrent parotitis (JRP) and salivary calculi. Sialoendoscopy is therapeutic for both processes, allowing for the extraction of calculi and the mechanical irrigation of debris within the salivary ducts (Hackett et al. 2012; Schwarz et al. 2018).

**Salivary Gland Pathology**

**Congenital Salivary Gland Pathology**

**Branchial Cleft Anomalies**

Anomalies of the branchial apparatus that involve the salivary glands are most frequently branchial cysts, sinuses, or fistulae. Although a thorough discussion of branchial embryology is beyond the scope of this chapter, it is useful to note that branchial cleft anomalies are classified by the work classification. Work type I and type II first branchial cleft anomalies are capable of producing parotid masses. Work type I first branchial cleft cysts are often located within the preauricular soft tissues. These anomalies may run parallel to the external auditory canal and contain hair, skin, and sebaceous glands. They often lie in close proximity to the facial nerve. Work type II first branchial cleft cysts arise inferior to the preauricular soft tissues, commonly at the angle of the mandible.

These cysts or fistulae may course superficial or deep to the facial nerve and have the potential to terminate within the external auditory canal at the bony-cartilaginous junction (Rosa et al. 2008). Branchial cleft anomalies are frequently asymptomatic until they become infected or are involved in a traumatic injury, after which they may enlarge rapidly and come to clinical attention. Management frequently involves treatment of the acute inflammatory episode with antibiotics, followed by surgical excision.

**Vascular Malformations**

Vascular malformations are nonneoplastic, congenital vascular anomalies. Unlike vascular neoplasms such as hemangiomas, they have histologically normal endothelium and frequently grow commensurate with the child. Vascular malformations that involve the salivary glands include lymphatic malformations, venous malformations, and mixed malformations. Like branchial cleft cysts, vascular malformations are frequently asymptomatic until an infectious or inflammatory process ensues, after which they may rapidly enlarge. Treatment involves management of acute inflammatory episodes followed by nonsurgical therapy. Occasionally, complete excision is possible. Unfortunately, many vascular malformations are intimately involved with surrounding structures, thus precluding a purely surgical approach. Under many circumstances, serial partial excision or intralesional injections with sclerosing agents are the only suitable options. Recurrence or residual disease is common.

**Acquired Salivary Gland Pathology**

**Inflammatory Disease**

**Bacterial Sialadenitis**

Bacterial infection of the salivary glands is associated with acute enlargement of the gland in concert with pain, fever, poor oral intake, and generalized malaise. The parotid glands are most commonly affected by bacterial infections, followed by the submandibular glands and sublingual glands. Unlike viral infections, bacterial
Sialadenitis is almost always unilateral. In the neonatal period, 40% of bacterial sialadenitis occurs in premature infants (Mehta and Willging 2006).

Physical examination requires bimanual palpation of the gland while observing for purulent discharge from the duct. The pain associated with acute suppurative sialadenitis is often so severe that the child may not permit a thorough examination; however, ductal discharge can be sent for culture if an uncontaminated specimen is obtained. *Staphylococcus aureus* and *Streptococcus viridans* are the most common organisms implicated in acute suppurative sialadenitis, and antibiotic treatment is therefore directed at these organisms. The majority of patients will see resolution of the infection with oral antibiotics in conjunction with adequate hydration and oral hygiene. Because salivary stasis commonly contributes to the development of sialadenitis, additional treatment involves massage of the infected gland and the use of sialogogues such as sour candy to increase salivary production. Young patients and patients with severe infection may require hospital admission for intravenous antibiotics. Rarely, an abscess develops in the salivary parenchyma that requires drainage. This should be suspected in the presence of gland fluctuance on physical examination or unresolving infection despite appropriate treatment. Confirmation of the abscess and the extent of fluid loculation can be obtained using ultrasonography. Large parotid abscesses that closely approximate the facial nerve should be drained using a nerve-identifying technique. Superficial abscesses may be drained via a standard preauricular incision with subsequent incision of the SMAS parallel to the orientation of the facial nerve. Under all circumstances, facial nerve preservation is of paramount importance, and the patient must be counseled preoperatively with regard to the risk of facial nerve injury. Submandibular abscesses can be approached via a standard submandibular incision placed inferior to the marginal mandibular branch of the facial nerve. Blunt dissection can then be performed in an inferior to superior direction that is deep to the plane of the nerve, until all abscess contents are drained and abscess loculations are taken down. Fluid should always be sent for culture, and a drain should be placed under most circumstances (Mehta and Willging 2006; Rice 1991).

Sialendoscopy is a well-tolerated and effective procedure for the treatment of recurrent sialadenitis in children. Sialendoscopy and salivary duct irrigation have been shown to improve the frequency and severity of sialadenitis in patients with juvenile recurrent parotitis (Ogden et al. 2016).

**Viral Sialadenitis**

Viral infection of the salivary glands is a common cause of acute inflammation in the pediatric patient. Viral sialadenitis most frequently involves the parotid glands; however, it is usually less tender on presentation and is infrequently associated with fever, anorexia, and malaise. Viral sialadenitis is a self-limiting process and often lasts less than 3 weeks in duration. Treatment is conservative.

Although the echovirus, coxsackie virus, and Epstein-Barr virus are frequently implicated in viral sialadenitis, the mumps virus (paramyxovirus) and HIV (retrovirus) deserve special mention. In the late 1960s, the mumps vaccine was instituted as a component of a universal pediatric vaccination program. As a result, the virus has declined steadily in the United States over the past 50 years. Despite this, mumps remains one of the most common causes of viral sialadenitis worldwide. Other components of the disease include a spectrum of pancreatic, gonadal, and meningeal involvement (Centers for Disease Control 1989).

The HIV virus must also be considered in patients with bilateral viral sialadenitis. With the advent of current antiretroviral therapies, patients with HIV are now surviving for decades with low levels of viral burden. As a result, HIV-associated disease manifestations are being seen more regularly in the chronically infected population. HIV parotitis manifests as bilateral cystic parotid disease that is nearly pathognomonic for HIV. Fortunately, HIV parotitis is exceedingly rare in children. Treatment is conservative unless
malignancy is suspected (Mehta and Willging 2006; Cvetinovic et al. 1991).

**Chronic Sialadenitis and Sialolithiasis**

Juvenile recurrent parotitis (JRP) is a form of chronic salivary inflammation that is thought to be one of the most common causes of sialadenitis in children. JRP is characterized by recurrent painful inflammation. Diagnosis requires two episodes of sialadenitis per year and is characterized by structural changes within the gland, including acinar destruction and ductal stenosis (Hackett et al. 2012). Although it has been noted to persist into adulthood, JRP frequently resolves following puberty and has been effectively treated using sialoendoscopy.

Obstructive sialadenitis occurring secondary to salivary calculi is rare in children and occurs in the submandibular gland in the majority (80%) of cases. This predilection for the submandibular gland is due to the horizontal plane of the Wharton’s duct, the higher relative alkalinity of secretions, the greater viscosity of submandibular gland saliva, and the greater concentrations of calcium phosphate and calcium carbonate. As a rule of thumb, the majority of salivary stones in the submandibular gland are radiopaque, and the majority of salivary stones in the parotid gland are radiolucent. Management involves treating acute infection with antibiotics, adequate hydration, salivary gland massage, and the use of sialagogues. Surgical management often includes the use of sialoendoscopy with stone retrieval (Ogden et al. 2016). Submandibular stones in the floor of mouth may be retrieved by incision and extraction with marsupialization of the duct; however, ductal stenosis is a postoperative risk. In severe or recurrent cases, gland excision may be performed. Lithotripsy has also been used with some success in appropriately selected patients (McJunkin et al. 2009).

**Mucoceles and Mucous Retention Cysts**

The term mucocele is often used interchangeably with the term mucous retention cyst. A mucocele differs from the mucous retention cyst in that it does not have an epithelial lining. Mucoceles result from minor salivary ductal obstruction (often secondary to trauma) with subsequent enlargement of the minor salivary gland. They most commonly occur on the lower lip and are painless masses; however, due to the proximity to the anterior dentition, the lesions are frequently involved in a cycle of growth and rupture.

Ranulas are mucus retention cysts that arise secondary to obstruction of the sublingual gland. They result in a mass that occupies the floor of the mouth, often elevating the tongue. If the ranula enlarges, it may extend into the neck as a “plunging ranula” by traversing the mylohyoid. Plunging ranulas, oral ranulas, and labial mucoceles are each treated by surgical excision with removal of Mantoux skin test. In contrast, atypical tuberculosis occurring secondary to *Mycobacterium avium-intracellulare* is a common source of sialadenitis in the pediatric population. Although the disease does not often originate in the salivary parenchyma, periparotid or submandibular lymph node involvement is common (Mehta and Willging 2006; Rieu et al. 1990). Mycobacterial lesions frequently have overlying skin breakdown (with occasional spontaneous drainage) and have minimal tenderness on examination. Surgical excision may be performed, as antituberculosis drugs often have little effect on the course of the disease. Unfortunately, operative management can be exceedingly difficult due to the necrotic and inflammatory nature of the disease and the proximity to critical structures. Observation may be elected when lesions do not appear to be at risk for secondary infection or drainage. Nonsurgical therapy often includes the use of antibiotics, including azithromycin and rifampin. Spontaneous resolution has been observed. Other granulomatous diseases affecting the salivary gland include sarcoidosis and actinomycosis. They are uncommon in children (Mehta and Willging 2006).

**Granulomatous Disease**

Mycobacterial sialadenitis may result from either *Mycobacterium tuberculosis* (TB) or an atypical mycobacterial infection. Extrathoracic TB with salivary involvement is uncommon in children, but should be included in the differential diagnosis for those children with parotitis and a positive
the surrounding salivary tissue. This is a minor undertaking in the case of the minor salivary tissue associated with the labial mucocele, but requires sublingual gland excision in the case of the ranula. Care must be taken to avoid injury to the lingual nerve or submandibular duct. With the exception of select revision procedures to remove recurrent plunging ranulas, almost all mucoceles, including primary plunging ranulas, can be removed via a transoral approach. Under some circumstances, recurrent plunging ranulas have been managed with sclerotherapy.

Sialorrhea and Ptialism

Drooling can occur secondary to the overproduction of saliva (ptialism); however it is more commonly the result of the inability to control a normal quantity of saliva (sialorrhea). Ptialism can be the result of medications that stimulate the parasympathetic (or inhibit the sympathetic) nervous system, due to teething, or due to certain infectious processes. In contrast, sialorrhea is often the result of neuromuscular dysfunction. Under both circumstances, drooling can result in skin irritation and halitosis and also cause significant psychosocial problems (Kupferman et al. 2010; Crysdale 1994). Treatment is aimed at reducing the production of saliva and may include the use of systemic sympathomimetic medications or injections of botulinum toxin into the salivary glands. Surgical management may include parotid and submandibular duct ligation, submandibular gland excision, and tympanic neurectomy. All procedures have variable results, and a combination of treatments is frequently necessary to achieve success.

Salivary Neoplasms

Salivary gland neoplasms are rare in children and represent fewer than 10% of all pediatric head and neck tumors (Dombrowski et al. 2019).

Benign Neoplasms

Benign masses constitute the majority of salivary gland neoplasms in patients of all ages and represent more than 60% of salivary tumors in children (Bentz et al. 2000).

Hemangiomas

The most common salivary neoplasm in children is the hemangioma. Hemangiomas are a distinct form of vascular anomaly that is classified as a vascular neoplasm. Unlike the vascular malformation, vascular neoplasms arise shortly after birth, have histologically abnormal endothelium on microscopy, and have a nonlinear growth pattern. Overall, 80% of salivary hemangiomas occur in the parotid gland and comprise more than 90% of salivary neoplasms in the first year of life. An additional 18% of salivary hemangiomas arise in the submandibular gland, and the remaining 2% arise in the minor glands (Mehta and Willging 2006; Boyd et al. 2009). Overall, 20% of patients will have hemangiomas in multiple sites, and more than 50% of patients with a parotid hemangioma will also have a cutaneous hemangioma. Treatment is largely conservative. The natural growth pattern of the hemangioma dictates that most lesions proliferate during the first 6–9 months of life and involute gradually over the following 5–7 years. Lesions that cause functional or symptomatic impairments may be managed with nonsurgical systemic therapy aimed at inhibiting vascular proliferation. Over the past decade, the use of propranolol has revolutionized the management of infantile hemangiomas.

Pleomorphic Adenoma (Benign Mixed Tumor)

The pleomorphic adenoma is the most common nonvascular benign neoplasm to occur in pediatric salivary glands. Like hemangiomas, the vast majority of lesions arise in the parotid. Patients frequently present with a painless, firm, well-circumscribed mass that is discovered incidentally on physical examination or after being visualized by the child’s parents. On histology, the neoplasm is comprised of myxoid, stromal, and epithelial components, earning it the name “benign mixed tumor.” Ultrasound or MRI will demonstrate a well-circumscribed, nondestructive lesion with pushing borders. The neoplasm is most frequently unilateral and unifocal. The exception to this rule may occur following incomplete excision, after
which tumor spillage may cause multifocal recurrence that can be exceptionally challenging to treat (Malata et al. 1997).

Primary management includes complete surgical excision with a margin of normal salivary tissue to reduce the risk of recurrence. For superficial parotid lesions, a superficial parotidectomy with facial nerve preservation is performed to ensure excision of the neoplasm. Observation of the pleomorphic adenoma is not advised due to the increased risk of malignant degeneration that occurs over time (Mehta and Willging 2006).

**Warthin Tumor (Papillary Cystadenoma Lymphomatosum)**
The Warthin tumor is a benign neoplasm that may be seen in the parotid gland. It is an extremely rare lesion in the pediatric age group and frequently occurs in older, male patients. It is the most common benign salivary neoplasm to present bilaterally and may occur multifocally within the gland (Bentz et al. 2000). On physical examination, the mass is painless, slow-growing, and compressible due to the cystic component. Treatment includes complete surgical excision.

**Sialoblastoma**
Also known as “embryoma,” sialoblastoma is an embryonic neoplasm that is unique to children and frequently arises in the minor salivary glands. They are commonly diagnosed within the first year of life. Although they are histologically benign, they are locally aggressive, and a 25% incidence of malignant transformation with cervical metastases has been reported (Batsakis et al. 1988). Treatment includes complete surgical excision.

**Malignant Neoplasms**
Excluding vascular anomalies and infection, more than 50% of pediatric salivary masses are malignant. Fortunately, malignant salivary gland neoplasms are still rare in children, and the majority of malignancies are low-grade lesions. The most common malignant epithelial neoplasms identified in children include mucoepidermoid carcinoma and acinic cell carcinoma. Morse et al. (2018) studied the epidemiology of pediatric salivary cancer in 588 children, and mucoepidermoid carcinoma was identified in 40% of the patients and acinar cell carcinoma in 37% of patients. Rhabdomyosarcoma is the most common mesenchymal lesion to affect children and is frequently diagnosed in the first 1–2 years of life.

**Mucoepidermoid Carcinoma**
Mucoepidermoid carcinoma is the most common salivary gland malignancy in children, constituting about half of all malignant salivary gland tumors found in the pediatric population (Dombrowski et al. 2019). The age of presentation is usually between 9 and 16 years (Mehta and Willging 2006). The neoplasm is usually located in the parotid gland; however, submandibular gland and minor salivary gland lesions are possible. The behavior of the lesion is determined by the histologic grade, and a treatment plan is established accordingly. The histologic grade is established by the level of cellular differentiation and by the presence of mucinous elements. Low-grade, well-differentiated lesions have an abundance of mucinous elements. Treatment involves complete surgical excision with a margin of normal tissue. Adjuvant therapy is unnecessary, and overall survival is excellent (Batsakis et al. 1988).

High-grade mucoepidermoid carcinoma is uncommon in children. Lesions are locally and regionally aggressive. They demonstrate a paucity of mucinous elements and resemble squamous cell carcinoma on histologic analysis. Treatment requires wide local excision with a selective neck dissection. Adjuvant chemoradiation is often required, and recurrence-free survival is drastically reduced when compared to low-grade neoplasms (Mehta and Willging 2006; Ethunandan et al. 2003; Shapiro and Bhattacharyya 2006).

**Acinic Cell Carcinoma**
Acinic cell carcinoma is the second most common salivary carcinoma in the pediatric population. Lesions often arise in a similar age group as mucoepidermoid carcinoma and are frequently low grade. Metastases are exceedingly rare; however bilateral and multifocal lesions are possible. Treatment includes wide local excision of the lesion. Recurrence is common (Sato et al. 2005).
Adenoid Cystic Carcinoma
Adenoid cystic carcinoma is the second most common salivary malignancy in adults but is far less common in children. Like adults, it is characterized by neural and perineural invasion and is associated with a high recurrence rate. Management includes wide local excision, neck dissection for positive cervical disease, and adjuvant radiation therapy. Long-term surveillance is necessary due to the potential for distant metastases to arise decades after primary therapy has been completed (Mehta and Willging 2006).

Rhabdomyosarcoma
Rhabdomyosarcoma is the most common sarcoma to occur in the salivary glands. It is frequently diagnosed in the first year of life as a rapidly enlarging parotid mass. Histologically, the lesions are most often undifferentiated or embryonal types. Treatment is dependent on the tumor stage. Prognosis is improved significantly if complete excision is possible, including microscopic dissection.

Surgical Considerations
Although a comprehensive description of operative techniques is beyond the scope of this chapter, the surgical management of salivary gland lesions is usually performed using a nerve-sparing technique. The decision to sacrifice surrounding structures is dependent on the pathology of the lesion being treated. Neurovascular structures are not sacrificed unless they are directly involved by a malignant neoplasm.

Conclusion and Future Directions
The diagnosis and management of pediatric salivary disorders has made important advancements over the past decade. It is likely that innovations in noninvasive imaging modalities and minimally invasive endoscopic interventions will continue to revolutionize the management of pediatric salivary gland disorders in the future.

Cross-References
- Haemangiomas and Vascular Anomalies
- Lymphatic Malformations
- Principles of Pediatric Surgical Imaging
- Rare Malignant Tumors
- Rhabdomyosarcoma

References
Batsakis JG, Mackay B, Ryka AF, et al. Perinatal salivary gland tumours (embryomas). J Laryngol Otol. 1988;102:1007–11.
Bentz BG, Hughes A, Ludemann JP, Maddalozzo J. Masses of the salivary gland region in children. Arch Otolaryngol Head Neck Surg. 2000;126:1435–9.
Bialek EJ, Jakubowski W, Zajkowski P, Szopinski K, Osmolski A. Ultrasound of the major salivary glands: anatomy and spatial relationships, pathologic conditions & pitfalls. Radiographics. 2006;26:745–63.
Boyd ZT, Goud AR, Lowe LH, et al. Pediatric salivary gland imaging. Pediatr Radiol. 2009;39:710–22.
Centers for Disease Control. Mumps prevention. MMWR Morb Mortal Wkly Rep. 1989;38:338–92, 397–400
Cherry JD. Mumps virus. In: Feigen RD, Cherry JD, Demmler GJ, Kaplan SL, editors. Textbook of pediatric infectious diseases. 5th ed. Philadelphia: WB Saunders; 2004. p. 2305–14.
Crysdale WS. Drooling. In: Gates G, editor. Current therapy in otolaryngology: head and neck surgery. Hamilton: B.C. Decker; 1994. p. 426–9.
Cvetinovic M, Jovic N, Mijatovic D. Evaluation of ultrasound in the diagnosis of pathologic processes in the parotid gland. J Oral Maxillofac Surg. 1991;49:147.
Dombrowski ND, Wolter NE, Irace AL, et al. Mucoepidermoid carcinoma of the head and neck in children. Int J Pediatr Otorhinolaryngol. 2019;120:93–9.
Eisenhut CC, King DE, Nelson WA, et al. Fine-needle biopsy of pediatric lesions: a three-year study in an outpatient biopsy clinic. Dagn Cytopathol. 1996;14:43–50.
Ethunandan M, Ethunandan A, Macpherson D, et al. Parotid neoplasms in children: experience of diagnosis and management in a district general hospital. Int J Oral Maxillofac Surg. 2003;32:373–7.
Freling NJ, Molenaar WM, Verney A. Malignant parotid tumors: clinical use of MR imaging and histological correlation. Radiology. 1992;185:691.
Garcia CJ, Flores PA, Arce JD, Chuaqui B, Schwartz DS. Ultrasonography in the study of salivary gland lesions in children. Pediatr Radiol. 1998;28:418–25.
Hackett AM, Baranano CF, Reed M, Duvvuri U, Smith RJ, Mehta D. Sialoendoscopy for the treatment of pediatric salivary gland disorders. Arch Otolaryngol Head Neck Surg. 2012;138(1):912–5.
Ilgit ET, et al. Digital subtraction sialography techniques: advantages and results in 107 cases. Eur J Radiol. 1992;15:44.

Inarejos Clemente EJ, Navallas M, Tolend M, et al. Imaging evaluation of pediatric parotid gland abnormalities. Radiographics. 2018;38:1552–75.

Kupferman ME, de la Garza GO, Santillan AA, et al. Outcomes of pediatric patients with malignancies of the major salivary glands. Ann Surg Oncol. 2010;17 (12):3301–7.

Malata C, Camilleri I, McLean N, et al. Malignant tumours of the parotid gland: a 12-year review. Br J Plast Surg. 1997;50:600–8.

McJunkin J, Milov S, Jeyakumar A. Lithotripsy for refractory pediatric sialolithiasis. Laryngoscope. 2009;119 (2):298–9.

Mehta D, Willging JP. Pediatric salivary gland lesions. Semin Pediatr Surg. 2006;15:76–84.

Morse E, Fujiwara RJT, Husain Z, et al. Pediatric salivary cancer: epidemiology, treatment trends, and association of treatment modality with survival. Otolaryngol Head Neck Surg. 2018;159(3):553–63.

Ogden MA, Rosbe KW, Chang JL. Pediatric sialendoscopy indications and outcomes. Curr Opin Otolaryngol Head Neck Surg. 2016;24(6):529–35.

Rice DH. Non-neoplastic diseases of the salivary glands. In: Paparella MM, et al., editors. Otolaryngology. Philadelphia: WB Saunders; 1991.

Rieu PN, van den Broek P, Pruszczynski M, et al. Atypical mycobacterial infection of the parotid gland. J Pediatr Surg. 1990;25:483–6.

Rosa P, Hirsch D, Dierks E. Congenital neck masses. Oral Maxillofac Surg Clin North Am. 2008;20:339.

Sato T, Kamata SE, Kawabata K, et al. Acinic cell carcinoma of parotid gland in a child. Pediatr Surg Int. 2005;21:377–80.

Schwarz Y, Bezdjian A, Daniel SJ. Sialendoscopy in treating pediatric salivary gland disorders: a systematic review. Eur Arch Otorhinolaryngol. 2018;275(2):347–56.

Shapiro NL, Bhattacharyya N. Clinical characteristics and survival for major salivary gland malignancies in children. Otolaryngol Head Neck Surg. 2006;134(4):631–4.

Sodhi KS, Bartlett M, Prabhu NK. Role of high resolution ultrasound in parotid lesions in children. Int J Pediatr Otorhinolaryngol. 2011;75:1353–8.