Introduction

Adenotonsillectomy is one of the most common major surgical procedures performed in children in the US today with an annual expenditure of $500 million. The trend, however, is toward selective performance of adenotonsillectomy, with 259,000 operations performed in 1987... one quarter the 1970 figure. Recurrent infection has always been the number one indication for adenotonsillectomy, though an increasing percentage of procedures are performed for OSA. A study from Mt. Sinai showed a rise in the percentage of adenotonsillectomies performed for obstructive symptoms from 0% in 1978 to 19% in 1986.

History of Tonsil and Adenoid Surgery

Celsus, using a primitive scalpel, first authenticated removal of the tonsils in approximately 50 A.D. Paul of Aegina further contributed to the development of the procedure with his description of the tonsillectomy in 625. In the mid-eighteenth century, Caque of Rheims was the first to perform tonsillectomies on a regular basis.

Development of instruments for adenotonsillectomy was heralded by Philip Syng whose invention became the forerunner for the modern tonsillotome. The Sluder guillotine was first used by P.S. Physick in 1827 for removal of the uvula. In 1867, Wilhelm Meyer used a ring knife through the nasal cavities to extract the adenoids of a patient complaining of hearing loss and nasal obstruction.

In the early 20th century, Samuel Crowe refined adenotonsillectomy by addressing potential risks preoperatively, improving surgical techniques, addressing anesthetic concerns, and emphasizing postoperative hospitalization. Adenotonsillectomy quickly evolved into one of the most common surgical procedures performed, becoming popularized as a cure for numerous ailments, including anorexia, mental retardation, and enuresis. Eventually it became touted as a method of improving health in general, and the only indication for removal was mere presence.

In the 1930s and 1940s, the excitement began to wane as new studies showed a natural decline in the incidence of upper respiratory infections in children after the first few years in school and more
antimicrobial agents became available. In the 1950s, “managed care” stepped in as a major health care program refused to pay for the procedure without endorsement from a pediatrician or internist.

Today support for the procedure continues. Attitudes acquired during training, judgements drawn from clinical experience, randomized clinical trials showing its efficacy, and the discovery of new indications such as prevention of abnormal development of the facial skeleton and dentition from chronic mouth breathing and relief of life-threatening airway obstruction have kept adenotonsillectomy popular.

**Embryology**

The first eight weeks constitute the period of greatest embryonic development of the head and neck. The five branchial arches are encased by clefts externally and pouches internally. Each pouch, with a ventral and a dorsal wing, evolves into endodermal tissue, while clefts evolve into ectodermal tissue and arches become mesodermal tissue.

The tonsillar pillars arise from the 2nd and 3rd branchial arches while the epithelium of the palatine tonsils is derived from the 2nd pharyngeal pouch. Development of tonsil tissue begins in the 14th week when mononuclear cells invade the mucosa of the tonsillar fossa, condense and differentiate into tonsillar lymphoid tissue. From 3 to 6 months, invagination of surface epithelium coupled with programmed cell death create crypts. The tonsillar capsules develop during the 5th month, and functional germinal centers develop after birth.

The development of adenoid tissue begins in the 3rd month from the subepithelial infiltration of lymphocytes into the posterior nasopharynx. Soon after birth, the tissue is colonized with bacteria then enlarges in childhood in response to antigenic challenges. By early adulthood the tissue has normally regressed.

**Anatomy**

**ADENOID TISSUE:**

The adenoid tissue is positioned in the midline of the posterior nasopharyngeal wall immediately inferior to the rostrum of the sphenoid. It makes up the most rostral portion of the pharyngeal lymphoid tissue termed Waldeyer’s ring. The space created lateral to the adenoid and posteromedial to the eustachian tube orifice is termed the fossa of Rosenmueller. Gerlach’s tonsil is lymphoid tissue within the lip of the fossa of Rosenmueller, which extends into the eustachian tube. Inferiorly, the adenoid tissue abuts the superior margin of the superior constrictor or Passavant’s ridge.

The blood supply to the adenoid tissue arises from the ascending pharyngeal and sphenopalatine arteries. Branches of IX and X provide sensory innervation and are responsible for referred pain to the ear/throat with adenoid infection.

The surface of the adenoids differs from the tonsils in that the adenoids have deep folds and few crypts, while the tonsils have 10 to 30 crypts. Ciliated pseudostratified columnar surface
epithelium is important for mucociliary clearance. With chronic infection, this layer thins, resulting in impaired mucociliary clearance and stasis of secretions. Increased exposure time to antigenic stimuli then results in an increased inflammatory tissue response. Deep to this layer is stratified squamous tissue, which thickens with chronic infection. The deepest layer is the transitional layer and is responsible for antigen processing. Neither the tonsils nor the adenoid tissue possess afferent lymphatics.

**PALATINE TONSILS:**

The tonsils are nestled in fossae formed by the palatoglossus anterior pillar and the palatopharyngeus posterior pillar, with the superior constrictor muscle in the floor. Preservation of these muscular condensations and the overlying mucosa is critical in maintaining physiologic function of the palate postoperatively. Hypertrophy of the tonsils with extension into the nasopharynx may result in nasal obstruction; more commonly, extension inferiorly into the hypopharynx results in upper airway obstruction.

The tonsils are contiguous inferiorly with the lingual tonsils. The point of attachment, known as the plica triangularis, must be transected during tonsillectomy. In patients with marked hypertrophy, this extension is frequently quite large and can result in troublesome bleeding at the point of transection during tonsillectomy.

The tonsillar branch of the facial artery provides the majority of the blood supply to the tonsils. The ascending pharyngeal, descending palatine and the dorsal lingual branch of the lingual artery also contribute. In most people, the internal carotid artery lies two centimeters posterolateral to the deep surface of the tonsil; however in 1% of the population, it is found just deep to the superior constrictor muscle.

The nerve supply of the tonsils arises from IX and some branches of lesser palatine nerve via the sphenopalatine ganglion.

The luminal surface of the tonsil is covered by deeply invaginated stratified squamous epithelium. The base of the tonsil is separated from the underlying muscle by a dense collagenous hemi-capsule. The parenchyma consists of numerous lymphoid follicles dispersed just beneath the epithelium of the crypts.

**Immunology**

The tonsils and adenoids are important in the production of antigen-specific secretory IgA. A system of clefts covered by specialized epithelium allows intimate contact between antigens and immune competent cells. Antigens are transported by M cells in the specialized squamous epithelium to a tubovesicular system where they are captured by APC (antigen processing cells) and transported to the next layer, the extrafollicular area. This layer, rich in T-cells, contains abundant vasculature allowing circulating lymphocytes to gain access to the tonsils. The lymphoid follicle is encased by the mantle zone where mature lymphocytes reside. At the core of the lymphoid follicle is the germinal center where immunoglobulin production takes place by B cells.
The importance of tonsil and adenoid tissue in host immune defenses is unclear. Most studies have failed to demonstrate significant differences in immunoglobulin production in children with recurrent tonsillitis; furthermore, no adverse systemic immunologic effects have been proven in adults or children after adenotonsillectomy.

**Microbiology**

Tonsillopharyngitis may be caused by a number of bacterial pathogens and respiratory viruses. Studies have shown that the majority of acute infections are incited by viruses and may lead to secondary bacterial infection. Approximately 5 to 30% of acute infections are bacterial; H. influenza, S. aureus, Strept pneumo, strept pyogenes (group A beta-hemolytic streptococci or GABHS) are the most frequently cultured bacteria from patients with recurrent acute tonsillitis and tonsil hypertrophy. In addition, tonsil weight has been shown to be directly proportional to bacterial load (Brodsky et al 1988).

GABHS is the most important bacterial pathogen due to its potential sequelae, rheumatic fever and glomerulonephritis. Although the incidence of rheumatic fever is decreasing in the US, many developing countries show it as the etiology of 30-40% of all heart disease.

The prevalence of beta-lactamase-producing organisms (BLPO) isolated from patients with acute and recurrent tonsillitis is rising. In a study by Gaffney (1991) 39% of all cultured organisms in patients with recurrent tonsillitis were BLPO. Another study demonstrated that the prevalence of beta-lactamase-producing isolates of H. influenzae rose from 2% in 1980 to 44% in 1989 with the prevalence of Staph aureus increased from 6 to 40%. Also more commonly seen now are anaerobic (particularly *Bacteroides*) and polymicrobial infections.

**Non-neoplastic Diseases of the Tonsils and Adenoids**

**ADENOIDS**

Acute adenoiditis can be difficult to distinguish from a viral URI but is more likely to be associated with temporary snoring and usually lasts longer than a URI. Both recurrent and chronic adenoiditis are difficult to distinguish from sinusitis, however otitis media is more likely seen in conjunction with adenoiditis. The triad of hyponasality, snoring and mouth breathing normally indicates obstructive adenoid hyperplasia.

**TONSILS**

Tonsillar disease can also be divided into four categories: acute tonsillitis, recurrent acute tonsillitis, chronic tonsillitis, and obstructive tonsillar hyperplasia.

Patients with acute tonsillitis may have a sore throat, fever, dysphagia, tender cervical lymphadenopathy, and erythematous/exudative tonsils; a throat culture should be obtained to test for GABHS. The incidence of GABHS pharyngitis is lowest in the infant and peaks from 6-12 years of age. Another common cause of a sore throat is acute pharyngitis. A patient with acute pharyngitis will have diffuse erythema of the soft palate and pharyngeal wall but no involvement of the tonsils or lymph nodes.
The differential diagnosis of GABHS adenotonsillitis includes infectious mononucleosis, scarlet fever, diphtheria, tularemia, toxoplasmosis, and malignancy (lymphoma, leukemia, carcinoma). An ulcerative pharyngitis may be noted in children with agranulocytosis. Scarlet fever is caused by streptococci with erythrogenic toxin and is characterized by a sandpaper rash starting in the axillae, groin and neck that becomes generalized within 24h. The rash is preceded by fever, vomiting, headache, pharyngitis and abdominal pain. The tongue has a white coat through which red and edematous papillae project (white strawberry tongue); after several days, this layer desquamates and the papillae persist (red strawberry tongue). Infectious mononucleosis is caused by the Epstein-Barr virus (EBV) and is characterized by exudative tonsillitis, massive cervical lymphadenopathy, splenomegaly, fever, and sensitivity to ampicillin. The diagnosis should be suspected if sore throat and malaise persist despite antibiotic treatment and can be confirmed with a blood smear showing atypical mononuclear cells and a positive Paul-Bunnell test (elevated heterophile titer of infectious mononucleosis).

The physician should consider the diagnosis of chronic tonsillitis when the patient has a sore throat or pain with swallowing that last longer than 4 weeks. Associated symptoms include tonsilloliths, halitosis, excessive tonsillar debris, peritonsillar erythema, and persistent tender cervical lymphadenopathy. On exam, one may see erythema, dilated surface vessels and a smooth tonsillar surface. Malignancy should be suspected for persistent unilateral tonsillar enlargement with constitutional symptoms despite appropriate antibiotic treatment. Be especially suspicious when the child is immunosuppressed or has a history of previous malignancy.

Complications of GABHS pharyngitis include cervical adenitis, retropharyngeal or parapharyngeal abscess, peritonsillar abscess, inflammatory torticollis, hemorrhagic tonsillitis, and postanginal sepsis (Lemierre’s disease). Peritonsillar abscess (Quinsy) is caused by infection of crypts in the supratonsillar fossa and can result in infection of the parapharyngeal space if the superior constrictor muscle is penetrated. The condition is preceded by acute tonsillitis and is characterized by low grade fever, trismus, and difficulty swallowing secretions.

Nonsuppurative late complications of GABHS include rheumatic fever and glomerulonephritis. Streptococcal skin infections lead to acute glomerulonephritis but rarely to acute rheumatic fever. Not all rheumatic fever patients have a history of preceeding URI, and not all throat cultures are positive for streptococci, though most patients will have an elevated streptococcal antibody titer during the acute phase. Joint symptoms are the most common presenting complaint (75%) with pain disproportionate to the physical findings. The arthritis is migratory and normally resolves in 3-4 weeks. Carditis occurs in 40-50% and can be acutely fatal or cause long term valve damage. Mitral stenosis is the most common valvular problem. Chorea occurs in 10-15%, most often effects the face and upper extremities, and is normally self-limited to 3 months.

Obstructive tonsillar hyperplasia may cause loud snoring, dysphagia, and voice changes. In adults, excessive daytime sleepiness is the most common presenting symptom of OSA. In children, however, snoring is the most common presenting symptom and may be accompanied by poor mentation, decreased attention span, poor scholastic performance, and nocturnal enuresis.
Benign diseases afflicting the tonsils include hyperkeratosis (mycosis leptothrica), tonsilloliths, elongated styloid process (Eagle’s syndrome), and candidiasis. Hyperkeratoses are numerous white horny masses found on the tonsillar crypts, posterior/lateral pharyngeal walls, and lateral glossoepiglottic folds (wherever lymph tissue exists). Tonsilloliths are yellow, yellow-gray, gritty particles embedded in crypts; they are most commonly seen in adults, particularly those with a history of recurrent tonsillitis. Elongation of the styloid process or ossification of the stylohyoid ligament (Eagle’s syndrome) can cause irritation of the trigeminal, facial, glossopharyngeal, and vagus nerves. Symptoms include recurrent nonspecific throat discomfort, foreign body sensation, dysphagia, facial pain, and increased salivation. Symptoms tend to be worse during maximal deglution and deep breathing. Thrush (Candida albicans) may present as the initial manifestation of AIDS or in elderly or postradiation patients with decreased salivary gland output.

**Clinical Evaluation**

**ADENOIDS**

The triad of hyponasality, snoring, and mouth breathing normally indicates enlarged, obstructing adenoids. Other symptoms of adenoid disease include rhinorrhea, post-nasal drip, chronic cough and headache. The patient should be asked about possible allergies, GERD, and sinusitis. A complication of adenoidectomy is velopharyngeal insufficiency (VPI) characterized by hypernasal speech and regurgitation of fluids through the nasal cavity. Patients with a submucous or overt cleft palate or preexisting VPI are at risk for this and should be identified prior to adenoidectomy. Personal history of fluid regurgitation through the nose or a family history of VPI/cleft palate should be elicited.

On physical exam, patients may have “adenoid facies” (long face, flattened midface, open mouth) and hyponasal speech. Speech can be assessed with the pinch test, where the nose is pinched and released during nasally transmitted phrases such as “banana” and “ninety-nine”; a lack of change in pitch is seen with nasal obstruction. The incidence of submucous cleft palate is 1 in 1200. Signs include bifid uvula, abnormal palatal motion, or midline diastasis of muscles with a V-shaped notch. A nasal exam should be performed looking for signs of sinusitis or allergic rhinitis. The oral cavity and jaw should be inspected for displacement of the mandible downward and backward from chronic adenoid hyperplasia. These children may have an overbite, long face and crowded incisors.

Lateral neck films may be helpful and are ordered if the history and physical exam are not in agreement. They cannot assess small amounts of obstructing tissue or stasis of secretions obstructing the choanae and are highly dependent on patient positioning and cooperation.

The differential diagnoses of adenoid disease are allergies, GERD and sinusitis. If sinus disease coexists and medical therapy fails, one should first proceed with adenoidectomy. Studies have shown that 67% of children will respond with adenoidectomy alone. If symptoms persist 2 to 3 months after adenoidectomy, further investigation is warranted.

**TONSILLITIS**

In the Children’s Hospital of Pittsburgh adenotonsillectomy study, each episode of tonsillitis had to have one or more of the following: oral temperature of at least 38.3°C, enlarged (>2cm) or
tender cervical adenopathy, tonsillar or pharyngeal exudate, or positive culture for GABHS. Attention should be made to symptoms of failure to thrive and cor pulmonale from chronic tonsillar hypertrophy. Complications of GABHS such as poststreptococcal glomerulonephritis and rheumatic fever should be considered.

On physical exam, the size of the tonsils should be documented. The following grading system was developed by Brodsky et al. using the ratio of the width of the tonsils to the width of the oropharynx (between the anterior pillars):

- 0- in fossa
- +1- less than 25% occupation of oropharynx
- +2- 25-50%
- +3- 50-75%
- +4- greater than 75%

Care should be taken to depress the tongue anterior to the circumvallate papillae; inadvertently gagging the patient may give a false impression of tonsillar enlargement. The inferior extent of the tonsils should also be assessed via physical exam, flexible laryngoscopy, or AP/lateral neck films.

The surface of the tonsils should be examined for erythema or exudate. With recurrent and/or chronic infection, the number of crypts will diminish resulting in a smooth, glistening surface.

Patients should be examined for craniofacial, neuromuscular, or CNS anomalies as these conditions put patients at greater risk for airway obstruction and postoperative complications.

A throat culture testing for GABHS should be obtained, although it doesn’t need to be positive to treat if clinical suspicion is high. The rapid streptococcal antigen detection test can be read in 12 minutes and is highly specific but a negative result must be confirmed by culture. Both tonsils should be cultured; one may miss 25% GABHS otherwise. Post-treatment cultures are indicated for children at unusually high risk for rheumatic fever, who remain symptomatic, or who develop recurring symptoms. If a second episode of GABHS occurs within a 6-week period, obtain a follow-up throat culture 3 to 10 days after treatment completion.

If infectious mononucleosis is suspected, obtain a peripheral blood smear and a Paul-Bunnell test. After recovering from mononucleosis, the sore throat may persist for up to 6 months.

**OBSTRUCTIVE SLEEP APNEA (OSA)**

Many physicians use subjective criteria to diagnose sleep disordered breathing in children. A number of clinical studies have shown no correlation of the patient’s history to the sleep study findings. The best objective method to diagnose OSA is by multichannel polysomnography (PSG). The dilemma concerning PSG is that the current economic environment makes it difficult to obtain a PSG. Also, unless an esophageal pressure monitor is used, the diagnosis of upper airway resistance syndrome will be missed. The most common pattern in children is continuous partial obstructive hypoventilation. Therefore, PSG is only ordered if the history and physical exam are not in agreement or in children who are at unusually high risk for perioperative complications. Of note, the American
Thoracic Society has made a consensus statement that OSA exists if the apnea index is > 1 per hour. The actual clinical relevance of this number is unknown but it is dramatically different than the adult criteria.

**Medical Management**

Brodsky reports that adenotonsillar hyperplasia may respond to one month of antibiotics aimed against BLPO and anaerobes (augmentin, clindamycin). She says this protocol may reduce adenotonsillar size enough to relieve the obstruction for more than one year in 15 to 20% of children. This would be particularly useful in the older child or in a child at high risk for perioperative complications.

Penicillin is still the 1st line agent for acute adenotonsillitis, and in the face of a negative throat culture for GABHS, should still be used if clinical suspicion is high. For recurrent positive GABHS culture, noncompliance should be considered and an injection of PCN or a 2nd course of antibiotics aimed against BLPO and anaerobes (augmentin, clindamycin, or penicillin plus rifampin) is mandated. Interestingly, studies have shown that alpha-hemolytic streptococci are capable of inhibiting GABHS in vivo and in vitro. Ross et al (1993) showed that recolonization of pharynxes of patients with recurrent GABHS with normal throat flora (alpha-streptococci) reduced the incidence of new episodes of infection.

For an asymptomatic patient with a positive post-treatment culture for GABHS, repeated courses of antibiotics should be given only if the patient has a history of rheumatic fever or family members with a history of rheumatic fever.

Acute upper airway obstruction from adenotonsillitis (most commonly seen with mononucleosis) can be managed initially with a nasopharyngeal airway, steroids, and intravenous antibiotics; immediate tonsillectomy is indicated for patients who do not respond quickly to conservative management.

**Selection of surgical candidates**

**ADENOIDECTOMY**

The only absolute indication for adenoidectomy is airway obstruction with secondary cardiopulmonary complications (cor pulmonale). Relative indications include chronic nasal obstruction with rhinorrhea, obligate mouth breathing, failure to thrive, eustachian tube obstruction with otitis (secretory or acute) or conductive hearing loss, abnormal speech (hyponasality), and abnormal dental/facial development. In children with recurrent or chronic otitis media, adenoidectomy can be performed with the initial tympanostomy tube insertion if nasal obstructive symptoms are present or in children who continue to have problems with otitis media after the initial set of tympanostomy tubes have extruded. Paradise et al found only limited benefits of adenoidectomy with initial tympanostomy tube placement for recurrent otitis media and did not feel these were sufficient to justify the costs and risks. On the other hand, in a review of tympanostomy tube placement and adenoidectomy for persistent middle ear effusion, Gates (1994) found that tympanostomy tube surgery alone is associated with a higher rate of repeat surgeries, increased rate of otorrhea, and greater expense and human cost of illness than initial myringotomy with adenoidectomy.
Other indications include speech problems or severe orofacial/dental anomalies from adenoid hyperplasia as well as suspected neoplasia. Relative contraindications to adenoidectomy are palatal clefting and VPI; if adenoidectomy is deemed necessary in patients with these conditions, some surgeons may perform a limited lateral or anterior resection of the adenoid pad.

**TONSILLECTOMY/ADENOTONSILLECTOMY**

In the last 30 years, the indications for adenotonsillectomy have become better defined. The only absolute indications for tonsillectomy are unquestioned dysphagia, extreme discomfort in breathing, clinically significant obstructive sleep apnea and alveolar hypoventilation/cor pulmonale. Malignancy, and uncontrollable hemorrhage from tonsillar blood vessels are also an absolute indication for surgery but their occurrence is rare. Paradise et al’s (1984) study found that the incidence of throat infection in children with recurrent tonsillitis who underwent adenotonsillectomy was significantly lower during the first two years of follow-up. Recurrent tonsillitis was strictly defined as at least three episodes in each of 3 years or five episodes in each of 2 years or seven episodes in 1 year. Each episode needs to be documented by a physician, as their study showed patient histories to be unreliable. Patients with prior episodes of tonsillitis and prior PTA may be more likely to have a second PTA and therefore benefit from tonsillectomy. Recurrences are most likely within the 1st year and in patients younger than 40. Hyperplasia resulting in OSA, failure to thrive, cor pulmonale, speech disorders, or severe orofacial/dental anomalies with narrow upper airway are further indications for tonsillectomy. Cardiac valve disease, persistent sore throat, tender lymphadenopathy, halitosis, tonsillolithiasis, and streptococcal carrier state unresponsive to medical treatment in a child or household at risk for rheumatic fever are other reasons cited in the literature for tonsillectomy.

The American Academy of Otolaryngology-Head and Neck Surgery requires only 3 or more infections per year to justify removal. Decisions for or against tonsillectomy should be made on an individual basis. Considerations should be made for parents’ and children’s preferences, fears, and tolerance of illness; school performance in relation to illness-related absence; the accessibility of health-care services; out-of-pocket costs; and comorbid diseases and risk of perioperative complications.

**Special Preoperative Considerations**

**ATLANTOAXIAL SUBLUXATION**

Ten to 20% of patients with Down syndrome have an unstable transverse ligament of the atlas and are prone to atlantoaxial subluxation. Other conditions predisposing to subluxation include Arnold-Chiari, achondroplasia, and rheumatoid arthritis. Flexion, extension lateral neck films showing an atlas-dens interval of more than 4mm in children and 3mm in adults are diagnostic for subluxation. All patients with Down syndrome should be examined preoperatively for neck pain, hyperactive deep tendon reflexes, and clonus and with positive findings, should have flexion, extension lateral neck films before going to the operating room. Patients with Arnold-Chiari may have symptoms of hydrocephalus with nystagmus, hyperactive deep tendon reflexes, or gait disturbances; MRI should be obtained in these patients as well as those with achondroplasia.
BLEEDING DISORDERS

Leeding time, PT and PTT should be ordered preoperatively if the history or physical examination suggests a coagulation disorder. Routine preoperative laboratory screening has not been shown to be any more effective than a thorough history in predicting blood dyscrasias. Historical clues to a coagulation disorder include frequent bruises, bruises >5cm, hematomas under bruises, and epistaxis lasting longer than 20min. Hutchinson developed the following questionnaire to assess for bleeding disorders:

1. Has the patient ever bled for a prolonged period of time after biting the tongue, cheek, or lip?
2. Does the patient develop spontaneous bruises <4-5cm diameter?
3. Has the patient experienced prolonged bleeding after minor surgical procedures (circumcision, skin biopsy, dental extractions)? Has bleeding recurred 24 hours after cessation of hemorrhage?
4. What medications has the patient taken in last 10 days? ASA?
5. Any blood relatives with any known bleeding disorder? Anyone required blood transfusion?
6. Any systemic medical disorders (lupus, liver or kidney disease)?

Two of the most common coagulation defects in the general population are platelet dysfunction and von Willebrand’s disease. Many commonly used medications cause platelet dysfunction; ASA and piroxicam should be discontinued 2 weeks prior to surgery, naproxen 4 days prior, and all other NSAIDS for 3 days (Randall et al).

Von Willebrand’s disease affects 1% of the general population and is transmitted autosomal dominantly with variable expression. There are over 20 different types; all affect Factor VIII:vWF necessary in platelet activation. Type 1 is the most common (80-90%) with subnormal plasma levels of qualitatively normal vWF. Patients with this type will respond to desmopressin. Type 2 results in a defect in vWF whereas in type 3, vWF is completely absent. Neither of these types will respond to desmopressin. Elevated PTT, bleeding time, and decreased/absent vWF antigen are diagnostic for this disease. Measurements of these factors should be taken in response to desmopressin preoperatively.

Perioperative management consists of administration of desmopressin (0.3 microg/kg) IV over 30 minutes preoperatively (peak levels at 45-60min), 12 hours postoperatively, then every morning until the eschar has completely sloughed and the fossae are completely healed. Aminocaproic acid is given pre- and post operatively to counteract the high concentration of fibrinolytic enzymes in the oral cavity (Derkay et al).

Adverse effects of desmopressin include hyponatremia and tachyphylaxis. In these situations, desmopressin should be discontinued and substituted with cryoprecipitate or vWF-containing antihemophilic factor.

IDIOPATHIC THROMBOCYTOPENIC PURPURA

Acute idiopathic thrombocytopenic purpura (ITP) is the most common of the thrombocytopenic purpuras of childhood. Severe thrombocytopenia is likely caused by an immune mechanism related to
sensitization by viral infections and formation of platelet antibodies. The acute phase with spontaneous hemorrhages lasts for only 1-2 weeks, and by 9-12 months after the onset, 90% of children have regained normal platelet counts. Only about 2% of cases are chronic and refractory. No treatment is mandated in mild cases. For chronic cases, infusions of intravenous gamma globulin (IVIG) have been found to cause sustained rises of platelet count. Splenectomy is indicated for cases persisting beyond one year. Children needing surgery should have a CBC drawn one week prior, and if thrombocytopenia is present, IVIG should be given (400mg/kg for 4 days).

**Principles of surgical management**

Positioning and exposure are of paramount importance. Patients are placed in the Rose position with a shoulder roll; a McGiven or Crowe-Davis mouthgag with a properly-sized slotted tongue depressor is placed in the oral cavity and suspended from the Mayo stand. Red rubber catheter through the nares are used to retract the soft palate. A curved or straight Alyss clamp is used to retract the superior pole and dissection is started in the mucosa of the superior anterior tonsillar pillar. Finding the proper plane is essential to prevent excess bleeding; dissection is carried out along the subcapsular plane, with care taken to avoid the lingual tonsil inferiorly. The actual method of removal of tonsils, either by dissection or electrocautery, is probably not as important as attention to the detail of staying in the proper plane between the tonsillar capsule and its surrounding fossa. After extraction, the area is irrigated, the mouth gag is released for a few minutes, then the fossae are reexamined for further bleeding. Blood and secretions which may cause inadvertent reflex laryngospasm should be suctioned prior to extubation. The most important tactic to avoid excess bleeding is finding the proper plane of dissection. Prophylactic cautery of the upper, middle, and inferior poles has been shown in one study to decrease primary hemorrhage from 2.8 to 0.2% (Williams et al).

Important to adenoidectomy is adequate visualization and avoidance of the eustachian tube. Overaggressive dissection with injury to the orifice of the eustachian tube may result in scarring and obstruction.

**Principles of postoperative management**

The following are the AAO-HNS indications for twenty-three hour inpatient monitoring:

1. age younger than 3
2. Evidence of obstructive sleep disorder
3. Systemic disorders which put the patient at increased perioperative risk
4. poor socioeconomic situation or other situation which would limit the patient’s ability to quickly return to the hospital
5. other medical problems (major heart disease, bleeding disorder, airway disorder, mental retardation, Down syndrome, craniofacial disorder, cerebral palsy, congenital defect)
6. When the procedure is done for a peritonsillar abscess

The most common causes for unanticipated inpatient stays are emesis, dehydration, hemorrhage, obstruction, and pulmonary edema. Patients most at risk for postoperative respiratory problems are those with PSG-proven OSA (23% complication rate) and Down syndrome, cerebral palsy or congenital defects (27% complication rate). Children younger than age 3 are less likely to
cooperate with oral intake and more likely to have surgery for airway obstruction. They are more prone to postoperative complications, with 7% experiencing airway difficulties, 4% becoming dehydrated, and 1.5% having hemorrhage.

A couple of recent articles have questioned the AAO-HNS’s indications for inpatient monitoring.

Mitchell et al. reported that children younger than 3 who undergo an adenotonsillectomy for recurrent infections may be managed safely as an outpatient. Historically, children younger than age 3 were thought to be less likely to cooperate with oral intake. Biavati et al. recommended routine inpatient admission for OSA patients, if they had one of the following conditions: cerebral palsy, age ≤ 3 years, congenital heart disease, seizures and prematurity.

**Perioperative Complications**

The mortality rate for adenotonsillectomy is 1 in 16000 to 35000, mostly from anesthetic complications and hemorrhage.

The incidence of postoperative hemorrhage is 0.1 to 8.1%. Transfusion is required in 0.04% of bleeding patients and mortality occurs in 0.002% (with a higher mortality rate in patients with primary hemorrhage). Reasons include retained adenoid tissue, and damage to posterior pharyngeal wall muscle. There is an increased incidence in the winter and in patients older than 20.

Anesthetic complications include fire, laryngospasm, kinking of the endotracheal tube and iatrogenic extubation. In the presence of a large leak around the endotracheal tube, fire can occur and can be prevented with the use of wet sponges in the oropharynx. Towels should be used instead of paper drapes with adhesive to prevent accidental extubation when drapes are removed.

Patients with chronic obstructive sleep apnea are at risk for postoperative pulmonary edema. Long-standing partial airway obstruction from enlarged tonsils serves as a natural PEEP. With sudden relief of the obstruction/PEEP, transudation of fluid into interstitial/alveolar spaces occurs resulting in pulmonary edema. Initial management consists of reintubation and administration of PEEP with gentle diuresis.

Care must be taken to avoid extraction of loose teeth. A post operative chest x-ray should be ordered if any teeth are broken or missing.

Burns of the eyes and surrounding soft tissue can be avoided by covering the eyes and using a insulated bovie spatula blade. Towel clips can cause trauma to the face or eyes and should be placed with caution.

The duration and severity of sore throat had been shown in a few studies to be greater in older children and with use of electrocautery. The University of Pennsylvania group has shown with a randomized controlled study that perioperative/postoperative administration of antibiotics reduced pain, lassitude, fever, mouth odor and poor oral intake after tonsillectomy.
Eustachian tube injury during adenoidectomy can cause COME or patulous ET. Otalgia may be referred pain from IX but otitis should be ruled out in case of eustachian tube injury or edema.

Fever is an accepted complication in the 1st 36 hours but will increase insensible water loss and predispose to dehydration.

Dehydration results from nausea/vomiting secondary to anesthesia as well as swallowed blood and decreased oral intake secondary to pain. Younger children are especially prone as they are less cooperative and have less volume reserve. A single intraoperative dose of steroids has been shown to accelerate return to a normal diet (April et. Al.) and decrease post-operative nausea and vomiting (Pappas et. Al.)

VPI is usually transient, but clinically significant in 1 in 1500 to 3000. Approximately one third can be identified preoperatively as having an increased risk (and should be evaluated by speech pathology preoperatively). If VPI persists beyond 2 months, speech therapy is indicated. Beyond 6 to 12 months, palatal pushback or pharyngeal surgery may be necessary.

Nasopharyngeal stenosis results from contracture of Waldyer’s ring and is seen in syphilis and with excess mucosal removal during surgery. Fusion occurs of the posterior pharyngeal wall, tonsillar pillars, and soft palate. Treatment is difficult; options include a unilateral palatal mucosal flap or a laterally-based flap from the posterior pharyngeal wall.

Atlantoaxial subluxation is treated initially with immobilization and neurosurgical consultation. Grisel’s syndrome is vertebral body decalcification and laxity of the anterior transverse ligament secondary to infection/inflammation in the nasopharynx (caused by traumatic adenoidectomy or infiltration of anesthetic into the prevertebral space). Spontaneous atlantoaxial subluxation may occur 1 week post operatively and is characterized by pain/torticollis and an atlas-dens interval more than 4 mm in children, 3 mm in adults.

A mandibular condyle fracture can occur if attempt for better exposure results in subluxation of the mandible anteriorly.

Postoperative infection is rare; endocarditis and cervical osteomyelitis have been reported.

Trismus may result from inflammation of the pterygoids.

Regrowth is possible; 15 to 28% will have tonsil tags and 6% will experience recurrent pharyngitis.

**Management of Postoperative Bleeding**

Prevention of bleeding is optimal by staying in the proper plane of dissection and by obtaining hemostasis prior to termination of the procedure. Any amount of bright red blood coming from the mouth or the nose should alert the surgeon to the possibility of postoperative bleeding. Frequent swallowing, tachycardia, and coffee-ground emesis are indirect signs of bleeding. If the patient complains of intermittent bleeding but is not actively bleeding on presentation, he or she should be
admitted for overnight observation and administration of IV fluids. Coagulation testing should be obtained following a bleeding episode, and if abnormal, hematology consult and additional coagulation testing should be obtained. A CBC should also be obtained to assess the degree of hemorrhage and as a baseline in case of further bleeding. Minor bleeding may be controlled by gargling with ice water and application of afrin to the oropharynx. If bleeding persists and the patient is cooperative, an attempt can be made to cauterized the bleed with silver nitrate in the ER.

Patients who are uncooperative or bleeding heavily should be taken to the OR. Initially, any fresh clot should be removed then identified bleeding sites should be controlled with electrocautery or a suture tie. If these measures are not adequate, a suture ligature may be necessary. Extreme care must be taken as the tissue is friable and the suture tends to tear through the tissue and pull out. Attempting to place the suture deeper may lead to inadvertant laceration of a major vessel deep to the fossa and possible pseudoaneurysm. The combination of multiple bleeding episodes or uncontrollable bleeding with normal coagulation tests should raise the suspicion of a vascular abnormality that should be evaluated with angiography.

References