Tracheomalacia and Tracheobronchomalacia in Children and Adults*

An In-depth Review

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Tracheomalacia and tracheobronchomalacia are disorders that are encountered in both pediatric and adult medicine. Despite increasing recognition of these disease processes, there remains some uncertainty regarding their identification, causes, and treatment. This article is intended to be a comprehensive review of both the adult and pediatric forms of the diseases, and includes sections on the historical aspects of the disorders, and their classification, associated conditions, histopathology, and natural history. We also review the various modalities that are used for diagnosis as well as the state of the art of treatment, including airway stent placement and surgical intervention. (CHEST 2005; 127:984–1005)

Key words: bronchomalacia; esophageal atresia; tracheobronchomalacia; tracheoesophageal fistula; tracheomalacia

Abbreviations: CPAP = continuous positive airway pressure; EA = esophageal atresia; FEF = forced expiratory flow; MAC = major airway collapse; TBM = tracheobronchomalacia; TEF = tracheoesophageal fistula; TM = tracheomalacia

M alacia refers to “softness” and, in medical terminology, generally refers to cartilage or bone.1 Tracheomalacia (TM) refers to a weakness of the trachea, frequently due to reduction and/or atrophy of the longitudinal elastic fibers of the pars membranacea, or impaired cartilage integrity, such that the airway is softer and more susceptible to collapse. By virtue of its compliance, the normal intrathoracic trachea dilates somewhat with inspiration and narrows with expiration, as a reflection of the difference between intrathoracic and intraluminal pressures.2–4 In TM, there is accentuation of this physiologic process, such that exaggerated changes in tracheal diameter occur.5 The majority of cases of TM are intrathoracic in nature1 such that excessive narrowing is most prominent when intrathoracic pressure is substantially greater than intraluminal pressure, as it is during forced expiration, cough, or the Valsalva maneuver.6,7 Various degrees of tracheal collapse, and therefore airway obstruction, can result from this narrowing.4,8 Less commonly, extrathoracic or cervical TM is seen. In these cases, negative intrapleural pressures are transmitted to the extrathoracic trachea due to pleural reflections such that the upper airway collapses during inspiration.1 TM may be localized to one portion of the trachea or may involve the entire trachea. If the mainstem bronchi are involved as well, the term tracheobronchomalacia (TBM) is employed. It should be noted that, historically, many authors have used TM and TBM interchangeably, especially in studies found in the pediatric literature. Thus, we have paralleled this practice where necessary. The term bronchomalacia is used to describe the isolated weakness and easy collapsibility of one or both of the mainstem bronchi.
without tracheal involvement. It is much less common than TM and TBM, and is therefore mentioned only briefly in this review.

Both TM and TBM are becoming more frequently recognized and treated in children and adults, a pattern that may be related to better diagnostic imaging and increased awareness of the condition among clinicians. Despite this, it is still thought to be a relatively underdiagnosed condition, and there remains confusion as to the appropriate treatment. For this article, an exhaustive review of the literature from the late 1800s to the present was undertaken using multiple medical literature search engines and the resources of Harvard Medical School, and the search included references in multiple languages. We herein review the history of pediatric and adult TM and TBM, the classification of these disease processes, the most common signs and symptoms, and the current methods for their diagnosis and treatment.

Pediatric TM and TBM

History

The earliest references to the disease we now call TM are from the 1930s and 1940s, when clinicians described congenital thoracic vascular abnormalities that resulted in tracheal obstruction. In 1948, Gross and Neuhauser described an infant with cough, wheezing, cyanosis, and spontaneous hyperextension of the head. Her trachea was compressed by an anomalous innominate artery that, despite surgical correction, had structurally deformed the trachea. One year later, Evans described several infants with symptomatic tracheal narrowing and advocated radiography as the method of choice for diagnosis. In 1950, Kirklin and Claggett suggested that prolonged pressure from anomalous vasculature can lead to the softening of tracheal rings. In 1952, Holinger et al. described the classic clinical features of malacic collapse of the airway lumen on expiration in three infants with dyspnea, stridor, and marked cyanosis. They noted that the symptoms disappeared with the passage of a bronchoscope through the flaccid walls of the trachea. They later suggested that surgery should be performed early in cases of vascular anomalies to “take full, effective advantage of the early growth potential of the constricted trachea.” In 1963, Baxter and Dunbar defined TM as a condition in which the tracheal wall weakens from a softening of the supporting cartilage and by hypotonia of the supporting myoelastic fibers, resulting in easy collapsibility. In addition, they divided the disease into primary and secondary forms. In 1968, Gupta et al. reported the first case of congenital isolated bronchomalacia in a 3-year-old child with a normal trachea but in whom the left mainstem bronchus collapsed to such a degree that the child required pneumonectomy for treatment.

Classification

In children, TM and TBM have been classified in various ways, including by macroscopic appearance, into “generalized” and “localized” disease. However, we adhere to the more commonly used division into congenital disease (also called primary) and acquired disease (also called secondary).

Congenital or Primary Forms

TM is the most common congenital anomaly of the trachea. It can be an isolated finding in healthy infants, but it is more commonly seen in premature infants. It is believed to be a consequence of the inadequate maturity of tracheobronchial cartilage, either from premature delivery or from an innate immaturity despite normal gestation. Some authors have reported no gender predominance in the primary form of the disease, whereas others have reported a definite male predominance.

Disease processes that result in the formation of an abnormal cartilaginous matrix of the trachea are included as primary forms of TM. These associated diseases, which include polychondritis and chondromalacia, result in dysmaturity of the collagen fibers and weakness in the tracheobronchial tissue. In addition, TM is clearly associated with the mucopolysaccharidoses, such as Hunter syndrome and Hurler syndrome, as well as with many other genetic syndromes (Table 1).

The most common associated disease is tracheoesophageal fistula (TEF), which some authors have classified as an acquired form rather than a congenital form of the disease. We and others think that TEF is more appropriately included as a primary form of the disease because the weakness of the trachea is not caused by compression from other structures but, instead, is due to an inherent weakness of the involved segment of the trachea. Some think that the congenital form of TM is due to a faulty division of the foregut into the trachea and esophagus during embryonic development, with the trachea receiving too much tissue during embryologic separation. This explanation accounts for the pathologic finding of a widened posterior membranous wall as well as the association with TEF and with esophageal atresia (EA). In 75% of pathologic specimens from infants born with TEF and EA, the circumference of the tracheal cartilage is reduced and the membranous trachea is widened, resulting in...
easy collapsibility.\textsuperscript{32} Blair and colleagues\textsuperscript{23} also have reported the presence of esophageal muscle in the posterior wall of the trachea in infants with TEF, EA, or both. On the basis of these findings, it has been have proposed that idiopathic primary TM may actually be the result of a faulty foregut division that was not severe enough to cause TEF or EA but did result in an inherent weakness of the trachea.\textsuperscript{23}

Mustard et al,\textsuperscript{33} Moes et al,\textsuperscript{34} and Cohen\textsuperscript{35} independently recognized that, although the innominate artery was often implicated in cases of secondary TM, angiographic and surgical data suggested normal positioning of the artery in many cases. We and others\textsuperscript{29} think that TM in this situation occurs from an inherent tracheal weakness that manifests as a pulsatile movement of an abnormally pliable trachea in the area juxtaposed to a normally placed artery. This condition has been labeled the "pulsatile" form of TM by some authors and should be considered a primary form of the disease.\textsuperscript{31}

In the majority of healthy and even premature infants, primary TM is a self-limiting disease. Most infants outgrow the condition by the age of 2 years.\textsuperscript{17,21,36} In the patients with connective tissue disorders and congenital syndromes, the effects of TM often persist and may even be fatal.\textsuperscript{26,37}

### Acquired or Secondary Forms

Acquired or secondary TM is caused by the degeneration of normal cartilaginous support from a variety of causes.\textsuperscript{17} Acquired TM is more common than the congenital form. There is a male predominance of the acquired form, but the reason for that is unknown.\textsuperscript{16,22} It commonly results from protracted endotracheal intubation,\textsuperscript{38} with the associated insults of increased airway pressure, oxygen toxicity, and recurrent infections. Premature infants with respiratory distress syndrome seem to be the most predisposed to this condition,\textsuperscript{21,39} as their supporting structures are not yet mature. As more and more premature infants are treated for respiratory distress syndrome, the incidence of TM may very well increase.

Tracheotomy also predisposes the patient to the development of TM.\textsuperscript{40} Not only is the stoma vulnerable, but the cuff site and the impingement point at the distal end of the tube may also be involved.\textsuperscript{4} The mechanisms of action could be pressure necrosis, impairment of the blood supply, infection, or mucosal damage caused by friction.\textsuperscript{31} In addition, TM is associated with recurrent tracheobronchitis.\textsuperscript{4,42}

A smaller percentage of secondary TM cases is caused by external tracheal compression from various structures (Table 1). Compression affects the integrity of the tracheal wall and increases the

### Table 1—Diseases Associated With TBM\textsuperscript{*}

<table>
<thead>
<tr>
<th>Disease Description</th>
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<tbody>
<tr>
<td>Primary (congenital)</td>
<td>Normal infants (idiopathic or primary TM proper)</td>
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<td></td>
<td>Prematurity</td>
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<td>Pulsatile collapse with normal innominate artery</td>
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<td>Congenital abnormalities of the cartilage</td>
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<td>Dyschondroplasia/chondromalacia/chondrodysplasia</td>
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<td>Polychondritis</td>
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<td>Ehlers-Danlos syndrome</td>
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<td>Congenital syndromes associated with TM/TBM</td>
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<td></td>
<td>Mucopolysaccharidosis</td>
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<td>Hunter syndrome</td>
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<td>Hunter syndrome</td>
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<td>CHARGE syndrome</td>
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<td>VATER anomaly</td>
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<td>Triosomy 9</td>
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<td>Triosomy 21</td>
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<td></td>
<td>Atelosteogenesis type 1</td>
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<td></td>
<td>Antley-Bixler syndrome</td>
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<tr>
<td></td>
<td>11p13 deletion</td>
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<td>12q11 deletion</td>
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<td></td>
<td>16–22 translocation</td>
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<td>Hallermann-Streiff syndrome</td>
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<td>Blackfan-Diamond syndrome</td>
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<td>Williams-Campbell syndrome</td>
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<td>Kniest dysplasia</td>
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<td>DiGeorge syndrome</td>
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<td>Deletion of 12q</td>
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<td>Larsen syndrome and Larsen-like syndromes</td>
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<td></td>
<td>Brachmann-de Lange syndrome</td>
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<td>Camptomelic dysplasia</td>
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<td>Pierre-Robin syndrome</td>
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<td></td>
<td>Congenital anomalies associated with TM/TBM</td>
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<td></td>
<td>Tracheoesophageal fistula</td>
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<td></td>
<td>EA with or without laryngeal cleft</td>
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<td></td>
<td>Bronchopulmonary dysplasia</td>
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<tr>
<td>Secondary (acquired)</td>
<td>Prolonged intubation</td>
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<td>Tracheotomy</td>
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<td></td>
<td>Severe tracheobronchitis</td>
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<td>Resulting from compression</td>
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<td>Vascular</td>
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<td>Double aortic arch</td>
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<td>Abnormal take-off of the innominate artery</td>
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<td>Anomaly of left pulmonary artery</td>
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<td>Right aortic arch</td>
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<td></td>
<td>Aberrant right subclavian</td>
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<td>Enlarged pulmonary veins</td>
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<td></td>
<td>Cardiac</td>
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<td>Left atrial hypertrophy</td>
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<td>Enlarged left atrium</td>
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<td>Skeletal</td>
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<td></td>
<td>Scoliosis</td>
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<td>Pectus excavatum</td>
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<td>Tumors and cysts</td>
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<td>Tenotomy</td>
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<td>Cystic hygromas</td>
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<td>Hemangiomomas</td>
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<td>Bronchogenic cysts</td>
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<td>Enterogenous cysts</td>
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<td>Thymoma</td>
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<td>Thymus enlargement</td>
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<td>Goiter</td>
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<td>Lymphatic malformation</td>
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<td>Lymphoema</td>
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<td>Neuroblastoma</td>
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<td>Infection</td>
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<td>Abscess</td>
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<td>Posttraumatic</td>
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</table>

\textsuperscript{*}CHARGE = colobomata of the eyes, heart defects, choanal atresia, retardation of growth, genital hypoplasia, and ear abnormalities; \textsuperscript{986} VATER = vertebral defect, anal atresia, tracheoesophageal fistula, esophageal fistula, and radial/renal dysplasia.
compliance over that in the adjacent tracheal tissue. Although some lesions are severe enough to cause persistent obstruction of the airway, many cause only transient collapse with an increase in intrathoracic pressure. The structures implicated in the development of secondary TM through compression include cardiovascular abnormalities, such as double aortic arch,\(^8\) anomalous left pulmonary artery,\(^{16,51,52}\) right aortic arch with left ligamentum arteriosum,\(^{44}\) and left atrial hypertrophy.\(^{53}\) Skeletal disorders, such as scoliosis,\(^{54}\) and pectus excavatum,\(^{55}\) may cause secondary TM. Last, space-occupying lesions, such as goiters,\(^{21,56,57}\) tumors,\(^{23,29,58}\) abscesses,\(^{29}\) and cysts,\(^{8,29,59,60}\) may compress the trachea. Even after surgical correction of the compressive structure, tracheal weakness and collapse may persist, occasionally requiring additional evaluation and treatment.\(^{29,61}\)

**Associated Conditions**

There are many conditions that are associated with TM. Cardiovascular abnormalities are found in 20 to 58% of patients with TM.\(^{8,23}\) These abnormalities include patent ductus arteriosus,\(^8\) atrial or ventricular septal defects,\(^8,23\) abnormalities of the aortic arch,\(^{62,63}\) hypoplastic left heart, hypoplastic right heart,\(^{64}\) Tetralogy of Fallot,\(^{23}\) dextrocardia,\(^{23}\) and valvular stenosis.\(^{8,23,64}\) Up to 52% of infants with TM have associated bronchopulmonary dysplasia.\(^{21,39,65–70}\) About half of infants with TM have gastroesophageal reflux disease.\(^{21,71,72}\) Reflux has been seen in up to 78% of patients with life-threatening TM and may require fundoplication,\(^{23}\) either simultaneously with TM surgical correction\(^{73,74}\) or sequentially to it.\(^{75,76}\) In addition, subglottic stenosis,\(^{21}\) laryngomalacia,\(^{1,21,77}\) and vocal cord paralysis\(^{8,21}\) may be seen. TM can occur in the setting of an immature autonomic nervous system.\(^{5,78}\) Neurologic impairment is seen in 8 to 48% of infants with TM.\(^{8,21,79}\) Mair and Parsons\(^8\) have also reported severe developmental delay in 26% of infants with TM.

**Histopathologic Characteristics**

The pioneers in the histology of the trachea in TM are Wailoo and Emery,\(^{80}\) who in 1980 compiled data from 1,000 sequential autopsies of children of all ages. Of the 74 children who had no known congenital disease but did have histories of respiratory symptoms prior to death, many had an increased tracheal internal perimeter and an increased cartilage length compared to the mean values of the investigators’ “normal” population. With greater statistical significance, the membranous trachea measurements were also larger than the mean, but the ratio of cartilage to muscle was reduced. As the proportion of cartilage would signify rigidity, this decreased ratio would imply easier collapsibility of the trachea.

Wailoo and Emery\(^{81}\) later studied the membranous tracheas of 560 children and found that the transverse muscle was consistently uniformly arranged; however, there was considerable variability in the longitudinal muscle fibers. These fibers are located deep to the transverse fibers and are most consistently found in the lower trachea, suggesting its importance in preventing collapse, particularly in the intrathoracic trachea. The lowest incidence of longitudinal muscle was found in the preterm infants (31.4%) and the highest was found in children > 1 year of age (54.0%), suggesting an increase in the muscle with age.

In a separate study, Wailoo and Emery\(^{32}\) studied 40 tracheas obtained postmortem from children with TEF’s and found that the most common abnormality was a deficiency of cartilage with a concomitant increase in the length of membranous muscle. There is a decrease in the normal 4.5:1 ratio of cartilage to muscle in 68% of children with TEF. This resulted in a loss of the normal horseshoe shape, potentiating the anteroposterior collapse of the walls during respiration. Endoscopic correlations of the findings of Wailoo and Emery\(^{32}\) have been observed in patients with TM and fistula.\(^{82}\)

Mair and Parsons\(^8\) have proposed a “major airway collapse” (MAC) classification system that incorporates histologic features and endoscopic appearance. MAC type 1 would represent congenital tracheal collapse, without airway compression. This group of patients would therefore include premature infants, those with EA, TEF, the mucopolysaccharidoses, and Larsen syndrome. MAC type 2 would be reserved for those patients with tracheal collapse caused by extrinsic compression, such as that from vascular anomalies, cysts, teratomas, hemangiomas, thymus enlargement, or goiter. MAC type 3 refers to acquired malacia arising from prolonged ventilatory support, tracheotomy, or severe tracheobronchitis. Mair and Parsons\(^8\) thought that the term tracheomalacia should be reserved for cases in which the posterior membranous wall is widened and its ratio to the anterior tracheal cartilaginous wall approaches the 2:1 ratio.

**Incidence and Natural History**

The incidence of the congenital form of TM in children at large is not commonly reported, but one study\(^{83}\) has estimated it to be 1 per 1,445 infants. The incidence of the congenital form of TM caused by
associated congenital diseases parallels the incidence of those diseases and is therefore smaller. In a retrospective study of 664 diagnostic bronchoscopies performed in a university setting, 15.4% of infants studied had evidence of TM. However, in children ≤3 years old undergoing bronchoscopy specifically for respiratory distress, TM has been shown to be present in up to 30% of patients. In a study of 50 infants with TM, 24 (48%) were considered to have primary TM and 26 (52%) to have secondary TM. Ninety-six percent of the infants with the secondary TM were premature infants with respiratory distress syndrome requiring prolonged mechanical ventilation.

With the advent of modern neonatal respiratory care, a growing number of premature infants are being treated for respiratory distress syndrome and bronchopulmonary dysplasia. In addition, the number of bronchoscopic procedures in the United States has increased greatly over the past decade, resulting in an increase in the number of documented cases of TM. The number of false-positive cases that this annual increase reflects is unclear, as the techniques that are used for diagnosis have not been standardized.

TM is associated with substantial morbidity and mortality. It may go unrecognized or misdiagnosed as asthma or other respiratory conditions, thereby contributing to the morbidity of the disease. The clinical severity of TM and TBM ranges from mild to life-threatening. The mortality rate from severe TM is as high as 80%.

Symptoms

Classically, the signs and symptoms of TM have been described as occurring not at birth but instead appearing insidiously during the first week to months of life. A more recent study noted that the first respiratory symptoms of congenital TM occur at birth in as many as 95% of infants. Expiratory stridor and cough, often described as barking or brassy, are the most commonly reported symptoms (Table 2). If there is malacia of the extrathoracic trachea, inspiratory stridor can be found. The cough is most likely caused by the juxtaposition of the anterior and posterior walls of the trachea, resulting in recurrent vibrations and irritation of the anterior and posterior walls of the trachea, resulting in recurrent vibrations and irritation of the airway. Affected children may display noisy, medium-pitched to high-pitched breathing. In addition, recurrent respiratory distress, cyanosis, spontaneous hyperextension of the neck, and breath-holding spells have been reported. The “baggpipe sign,” an expiratory sibilant note that persists after the end of visible expiration, has also been described.

Table 2—Symptoms of Pediatric Tracheomalacia

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<th>Symptom</th>
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<td>Stridor</td>
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<td>Barking cough</td>
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<td>Respiratory distress</td>
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<td>Wheeze</td>
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<td>Anoxic spells</td>
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<td>Cyanosis</td>
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<td>Bradycardia</td>
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<td>Tachyarrhythmias</td>
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<td>Spontaneous hyperextension of the neck</td>
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<td>Prolonged expiratory phase</td>
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<td>Breathholding spells</td>
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<tr>
<td>Failure to thrive</td>
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<tr>
<td>Increased work of breathing</td>
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<tr>
<td>Sternal, substernal, and intercostal retractions</td>
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<tr>
<td>Recurrent pulmonary infections</td>
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<tr>
<td>Reflex apnea</td>
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<tr>
<td>Respiratory arrest</td>
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<td>Cardiac arrest</td>
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Sternal, substernal, and intercostal retractions can be seen as the work of breathing increases. Symptoms may be exacerbated by activities that increase the child’s respiratory efforts, including coughing, feeding, and crying. These activities can increase intrathoracic pressure until the extraluminal pressure exceeds the intraluminal pressure and the trachea collapses. Most of these infants have impaired clearance of secretions as a result of luminal closure during cough. Squamous metaplasia and impaired mucociliary clearance develop, which contributes to the development of respiratory infections and recurrent pneumonia. It is important to mention that airway obstruction can occur with anesthesia both in patients known to have or suspected to have TM and in patients who were previously asymptomatic.

Infants with TM, especially those with vascular compression, are often described as having “feeding difficulties.” These difficulties include varying degrees of dysphagia, regurgitation, cough, and cyanosis, and are thought to be secondary to anatomic and reflex mechanisms. When the trachea is compressed by a full esophagus, intermittent respiratory obstruction and arterial desaturation can occur, interfering with normal feeding and resulting in poor weight gain. These children may continue to feed despite cyanosis, therefore parents are instructed to interrupt feeding at frequent intervals.

“Reflex apnea,” a term coined by Fearon and Shortreed, is also called “death attacks,” “dying spells,” and “apneic spells” in the literature. The process is thought to be a reflex that is triggered when the trachea is stimulated with secretions or a bolus of food in the esophagus, leading to respiratory arrest. In fact, affected infants cease respiration during bronchoscopy through the same reflex mech-
anism. In 1978, Davies and Cywes\textsuperscript{89} reported 15 respiratory arrests in a patient with repaired EA, distal TEF, and TM. These respiratory arrests can lead to cardiac arrest,\textsuperscript{23,100} and up to two thirds of children with reflex apnea die from their disease.\textsuperscript{8}

A clinical severity rating has been proposed for children with TM caused by TEF,\textsuperscript{29,77} and it has been applied to all patients with TM.\textsuperscript{101} Children with “mild” cases have respiratory difficulties associated with infectious processes such as croup or bronchitis. These children often have difficulties with retained secretions. Children with “moderate” cases present with more of the classic findings, including stridor, wheezing, recurrent respiratory infections, and even cyanosis associated with exacerbations. Children with “severe” cases present with stridor during tidal breathing, marked sputum retention, and upper airway obstruction, reflex apnea, and even cardiac arrest.

**Diagnosis**

TM and TBM must be differentiated from difficult-to-control asthma, intraluminal obstruction, foreign body aspiration, and other diseases. One must have a high index of suspicion for the disease. As Gupta et al\textsuperscript{18} suggested 30 years ago, the uncommon diagnosis of TM may be due to our failure to suspect it. Sotomayor et al\textsuperscript{86} reviewed a series of children with TM in which the mean delay from symptom onset to correct diagnosis was 6 months. Mair and Parsons\textsuperscript{8} found similarly that the delay from the onset of symptoms to diagnosis ranged from 1 to 144 weeks, with a mean of 37 weeks in their series of 38 infants and toddlers. Blair et al\textsuperscript{23} reported that the mean age of treatment in their 25 patients with TM and TBM was 7 months, but the range was 1 to 96 months.

Many methods have been used through the years to diagnose TM. Obtaining an adequate history and performing a physical examination are paramount. If the condition is mild enough, pulmonary function testing may be helpful. The flow-volume loop classically reveals flow limitation on the expiratory component and a reduced midexpiratory/midinspiratory ratio.\textsuperscript{102}

Radiographs (including comparative inspiratory and expiratory views),\textsuperscript{17} tomograms,\textsuperscript{4} tracheograms or bronchograms,\textsuperscript{4,16} and fluoroscopy\textsuperscript{4} have all been used in the diagnosis of TM. In the 1970s, many authors\textsuperscript{103,104} thought that all children with stridor should be evaluated radiographically. The most recent data suggest that the sensitivity of plain radiographs for diagnosing TM is only 62\%, using microlaryngoscopy and bronchoscopy as the reference standards.\textsuperscript{105} In addition to plain radiography, barium esophagography is useful for evaluating associated disease processes, including TEF, EA, and reflux disease.

Prospective studies have evaluated the value of CT scans in diagnosing airway lesions in children. Frey et al\textsuperscript{106} found 13 airway abnormalities by endoscopic evaluation, 11 of which (84.6\%) were also found by CT scan. This high sensitivity coupled with the fact that CT scanning is rapid, noninvasive, and often requires no sedation in children, has led to its use as an initial test in those children with stridor or suspected TM. In the past decade, helical CT scanners, as well as multidetector helical CT scanners, have been developed. The speed of multidetector scanners, which permit imaging of the entire central airways in only a few seconds, is particularly appealing for use in the pediatric population.\textsuperscript{107} Although still exposing the patient to ionizing radiation, strict adherence to pediatric guidelines for reducing the CT scan dose provides high-quality images without the use of excessive radiation exposure.\textsuperscript{108} Moreover, because of the inherently high contrast between the air-filled trachea and the adjacent soft-tissue structures, it is likely that even further dose reductions can be employed, as demonstrated in a recent animal model study by Boiselle et al.\textsuperscript{109} Another advantage of CT scanning is that the data can be reconstructed into two-dimensional and three-dimensional images, including virtual bronchoscopy and external rendering. Although a preliminary report by Konen et al\textsuperscript{10} has suggested that virtual bronchoscopy may play a complementary role to conventional bronchoscopy in the evaluation of children with central airways disorders, more research is necessary in this area to determine the precise role of this emerging technique.

Although CT scanning provides excellent anatomic detail of the trachea and adjacent structures, this technique exposes patients to ionizing radiation and requires iodinated contrast material administration to fully assess the mediastinal vascular structures. Because of its lack of ionizing radiation, its ability to evaluate vascular structures without iodinated contrast material, and direct multiplanar imaging capabilities, MRI is the preferred method for evaluating extrinsic airway abnormalities, including vascular compression syndromes\textsuperscript{111–113} (Fig 1). As CT scanning and MRI technology evolve, it is likely that they will play an even larger role in imaging the pediatric airway and its surrounding structures.

Despite the amazing advances in imaging the airways, it remains difficult to image the airway during dynamic maneuvers in very small children because of their inability to cooperate with breathing instructions. Thus, for this reason and those described above, many experts still think that diagnos-
tic endoscopy is mandatory.\textsuperscript{29,36,82} We think that endoscopy is an invaluable, essential tool and that it should remain the preferred method for evaluating the airways. This belief is somewhat controversial in pediatric populations because endoscopy is invasive and requires that the child be spontaneously ventilating. If the child is paralyzed, heavily sedated, or receiving positive-pressure ventilation through any means, airway collapse may be missed. Therefore, general inhalation anesthesia, which maintains spontaneous ventilation without endotracheal intubation, is often employed. The anesthetic is entrained along with oxygen into the side port of the rigid bronchoscope. Once the vocal cords are inspected and anesthetized with topical agents, the trachea and bronchi are evaluated, with careful attention to their shape, configuration, and mucosal contours. As the anesthesia is reduced and the patient begins to move and cough, the dynamics of the airway can be assessed fully.\textsuperscript{27}

A visualized decrease in the diameter of the trachea of $>50\%$ is considered to be abnormal. This percentage is based primarily on studies in the 1960s by Wittenborg et al,\textsuperscript{2} who used trachograms to show that changes in the diameter of the trachea were minimal with quiet breathing in healthy children. However, with exertional respiratory efforts, such as crying or struggling, tracheal collapse reduced the diameter to between 20\% and 50\% of normal. The majority of infants with TM have a $>75\%$ collapse, with complete tracheal collapse in up to 33\% of infants.\textsuperscript{2} In addition to the typical anterioposterior collapse, indistinct tracheobronchial rings are a described finding.\textsuperscript{36}

\textbf{Treatment}

In many children with TM, intervention is not necessary. As the child grows, the tracheal cartilage strengthens and stiffens. The symptoms often resolve in children with mild-to-moderate TM by age 1 year\textsuperscript{7} or 2 years.\textsuperscript{17,47,85} Therefore, conservative therapy in milder cases is preferred,\textsuperscript{31} and includes the treatment of respiratory infections, humidified oxygen therapy, and pulmonary physiotherapy.\textsuperscript{31,87} For children who do not recover spontaneously or who have life-threatening symptoms, a variety of treatment options are available.

There have been many inroads in the treatment of TM in the last 3 decades. In the past, the mainstay of therapy in patients with severe TM was tracheostomy and long-term mechanical ventilation. Pioneering physicians recognized early that standard tracheostomy tubes were not always long enough to stent the involved segment of the trachea. Physicians developed elongated tracheostomy tubes or advanced thin-walled tubes through a standard tracheostomy to mechanically stent the distal trachea.\textsuperscript{16,114–117} The problems with this approach included the need to change the length of the tube as the child grew, recurrent bronchospasm, the negation of the glottic mechanism of increasing intratracheal pressure, and difficult decannulation.\textsuperscript{64} The associated complications of additional tracheal injury and recurrent infections also decreased the popularity of this approach and spurred the search for alternatives.

The percentage of infants and children who still require tracheostomy for TM varies from 12 to 62\%.\textsuperscript{8,21,31,118,119} In 1994, Jacobs et al\textsuperscript{21} reviewed 50 cases of TM and reported that tracheostomy was required in 75\% of premature infants and in 39\% of full-term infants. Of those, 71\% were able to undergo decannulation after an average of 30 months without further intervention.\textsuperscript{21} In 1992, Duncan et al\textsuperscript{120} reviewed a total of 44 home tracheostomies and found that 14 (32\%) were for TM. With the advent and implementation of other forms of therapy, this percentage is likely to decrease.

Continuous positive airway pressure (CPAP) is an effective treatment for infants with moderate-to-severe TM, TBM, or bronchomalacia.\textsuperscript{101,121–128} Bronchoscopy\textsuperscript{121,125} and fluoroscopy\textsuperscript{86} have shown that CPAP maintains airway patency during tidal breathing. By essentially creating a pneumatic stent, CPAP prevents the collapse of the airway throughout the respiratory cycle.

Tidal-breathing flow-volume measurements have been used to evaluate the changes in airway obstruction with the use of CPAP.\textsuperscript{101,123} The level of CPAP is often based on these evaluations, but more commonly it is based on clinical response.\textsuperscript{122,124,127,129}

\textbf{Figure 1. MRI of vascular entrapment of the trachea. The arrow refers to the narrowed segment.}
Kanter et al\textsuperscript{121} used up to 19 cm H\textsubscript{2}O pressure, and, with the aid of esophageal measurements, found that esophageal pressure did not exceed 3 cm H\textsubscript{2}O and that circulation was not compromised. Davis and colleagues\textsuperscript{101} measured full forced expiratory flow (FEF)-volume curves at CPAP levels of 0, 4, and 8 cm H\textsubscript{2}O in infants with TM and in healthy control subjects. In both groups, FVC did not change with increasing levels of CPAP, but inspiratory capacity decreased and functional residual capacity increased. The FEF values at functional residual capacity increase with increasing levels of CPAP, but the FEF values at 50\% and 75\% of expired volume did not differ with different levels of CPAP or between the groups. The authors concluded that CPAP affects forced flows primarily by increasing lung volume. Panitch et al\textsuperscript{128} agreed with their predecessors that the measurement of tidal mechanics was important in determining the appropriate level of pressure. However, they placed new emphasis on the mechanics during forced expiratory maneuvers (such as coughing and crying) because airway collapse is more likely to occur during such maneuvers.

Despite these advances, CPAP has some disadvantages, including lag in the commencement of oral feedings, retardation of speech and language, and potential developmental delay.\textsuperscript{21,122} Jacobs et al\textsuperscript{21} reported that CPAP was required for an average of 21.4 months by premature infants and 22.0 months by the full-term infants in their series. Depending on the estimated duration of treatment, CPAP can be considered a primary treatment or as an adjuvant to other therapies.\textsuperscript{122}

In patients who have the most severe forms of TM that are unresponsive to medical management or who present with life-threatening symptoms, surgical intervention may be necessary. Indications for surgery include recurrent pneumonia,\textsuperscript{23,87,130} intermittent respiratory obstruction,\textsuperscript{23} and the inability to extubate the airway.\textsuperscript{23,87,130,131} Dying spells, the most concerning of all symptoms, is an indication for continued hospitalization and definitive surgical intervention.\textsuperscript{23,76,130}

During the past decade, aortopexy has become the procedure of choice for the treatment of severe TM.\textsuperscript{23,87} Gross and Neuhauser\textsuperscript{12} were the first of many investigators\textsuperscript{6,23,24,31,34,94} to describe the use of aortopexy specifically for treating TM caused by a vascular anomaly. In this procedure, the ascending aorta is approached traditionally through a right-sided anterior thoracotomy at the third intercostal space\textsuperscript{23,132} or through alternative routes.\textsuperscript{7,133–136} Traction on the sutures placed in the wall of the aorta juxtaposes it to the undersurface of the sternum, which also pulls the anterior wall of the trachea forward. This mechanical fixation of the aorta widens the anterioposterior dimensions of the trachea and prevents collapse.\textsuperscript{23,87} It is important to note that aortopexy is not always successful in relieving the collapse,\textsuperscript{23,87} and therefore additional surgical approaches have been devised.\textsuperscript{130,137–141}

Of note, patients with airway malacia warrant special consideration with respect to anesthesia.\textsuperscript{142,143} In the 2003 review by Austin and Ali,\textsuperscript{1} the authors point to the following two main goals of the anesthesiologist: preventing airway collapse and air trapping with the use of CPAP/positive end-expiratory pressure; and minimizing cough. The authors discuss\textsuperscript{1} the safe and effective use of topical anesthesia, general anesthesia with inhalational agents, general anesthesia with IV agents, endotracheal tubes, and laryngeal mask airways in experienced hands.

External splinting with autologous materials, most commonly a resected rib, and prosthetic materials, such as silastic membrane-reinforced crystalline polypropylene and high-density polyethylene (Marlex; Phillips Petroleum; Bartlesville, OK) mesh, have been used to support the flaccid trachea.\textsuperscript{23,47,74,131,144,145} Support can be provided either by suturing a rigid support directly to the membranous trachea through a posterolateral right thoracotomy or by wrapping the supporting material around three quarters of the tracheal circumference through an anterior cervical or median sternotomy approach.\textsuperscript{23} In animal studies\textsuperscript{144} and in limited human studies,\textsuperscript{146} this procedure did not adversely affect the growth of the trachea. However, it is an invasive procedure that may not adequately treat distal bronchial lesions and may not be tolerated well by patients with complicated conditions.\textsuperscript{87} Treatment with tracheoepoxy,\textsuperscript{147,148} tracheal resection,\textsuperscript{31,149} and tracheal reconstruction\textsuperscript{150} have been attempted with limited success.

Internal tracheal stent placement with a silicone prosthesis was first attempted in 1965 by Montgomery.\textsuperscript{151} Since then, a wide variety of stents have been used, including those made of silicone and of metal.\textsuperscript{64,87,152} The obvious advantages of stents are their less invasive nature and the shorter surgical recovery times. Filler et al\textsuperscript{153} described 16 patients with airway obstruction, 8 of whom had TBM as their primary diagnosis. All eight children were successfully treated with stents, including two children with dying spells.\textsuperscript{153} Furman et al\textsuperscript{154} also reported relief of airway obstruction and the liberation from mechanical ventilation with the use of stents; however, the formation of granulation tissue was a common complication, and one death in the series may have been related to stent placement.\textsuperscript{154}

Metal stents have the advantages of minimal thickness and easy deployment. However, complications and limitations include granulation tissue formation,
difficult removal, death, stent fracture, the need for additional stent placement, migration, and the need to further dilate the stent as the child grows. An advantage of the silicone stents is the relative ease of both the removal of the stent and the deployment of a larger stent if it is required as the child grows. Removability is especially important, as children stand a reasonable chance of becoming asymptomatic once their airways grow. Currently, stents are used in limited situations in which conventional therapy has failed. However, stents currently in development, such as resorbable biopolymer stents, may address the limitations of current stents. With these new developments, stent placement may become a more attractive treatment option in the future.

In conclusion, conservative treatment is preferred in milder cases of TM. Secondary TM can develop if primary TM is treated with tracheostomy, mechanical ventilation, or both. Where conservative measures fail, the therapy must be tailored to the individual patient. To date, no perfect operation or procedure exists for the treatment of TM.

**ADULT TM AND TBM**

**History**

In 1897, Czyhlarz was the first to describe the postmortem finding of an unusually large trachea and bronchi. The first clinical report of isolated tracheal enlargement in an adult was by Mounier-Kuhn in 1932. Lemoine was the first to use bronchoscopy to document acquired tracheal enlargement in the adult in 1949. Additional case reports of TM in adults began to appear in the 1950s. Ferraris described two patients with acquired TM who both reported “expiratory dyspnea,” the inability to clear secretions, and recurrent respiratory infections. Both had been labeled and treated as asthmatic patients. Herzog and Nissen described five patients with acquired TM, all of whom had stridor, cough, and expiratory obstruction. Endoscopically, all five patients had tracheal obstruction caused by the ballooning of the posterior membranous wall into the tracheal lumen during expiration. Four patients were treated surgically by placing a bone graft in the membranous trachea to prevent collapse.

**Classification**

A few classification schemes for adult TM have been proposed. Classification by macroscopic clinical findings is sometimes employed, with lateral wall narrowing being called the “saber-sheath type” or “fissure shape,” and the anterioposterior wall narrowing being referred to as the “crescent type” or “scabbard shape.” Some clinicians have included a third macroscopic designation for the “circumferential” narrowing or have labeled this appearance as a combination of the crescent and saber-sheath types. Feist et al and others classified TM into congenital forms (as in the Mounier-Kuhn disease) and acquired forms, such as those resulting from tracheostomy, chest trauma, chronic irritation, inflammation, mechanical anatomic factors, or malignancy. We have adhered to the latter classification schema, as we did in the pediatric section (Table 3).

**Congenital or Primary Forms**

In the pediatric section, we discussed the congenital diseases, such as polychondritis, that weaken the trachea. Some of these conditions allow the survival of affected children into adulthood, therefore these adults would be considered to have congenital TM. There is one additional congenital condition that is not seen in children, but is found in the adult population. Idiopathic giant trachea, or tracheomegaly, is a rare condition that is characterized by atrophy of longitudinal elastic fibers and thinning of the muscularis mucosa. This combination allows the trachea and central bronchi to dilate, with a transition to the normal diameters of the peripheral airways.

**Table 3—Classification of Adult TM**

<table>
<thead>
<tr>
<th>TM Description</th>
<th>Genetic, such as polychondritis (also see pediatric table)</th>
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<tbody>
<tr>
<td>Primary (congenital)</td>
<td>Idiopathic “giant trachea” or Mounier-Kuhn syndrome</td>
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<tr>
<td>Secondary (acquired)</td>
<td>Posttraumatic</td>
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<td></td>
<td>Postintubation</td>
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<td></td>
<td>Posttracheostomy</td>
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<td></td>
<td>External chest trauma</td>
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<td>Post-lung transplantation</td>
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<td></td>
<td>Empysema</td>
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<td></td>
<td>Chronic infection/bronchitis</td>
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<td></td>
<td>Chronic inflammation</td>
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<td></td>
<td>Relapsing polychondritis</td>
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<td></td>
<td>Chronic external compression of the trachea</td>
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<td></td>
<td>Malignancy</td>
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<td>Benign tumors</td>
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<td>Cysts</td>
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<td>Abscesses</td>
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<td></td>
<td>Aortic aneurysm</td>
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<td></td>
<td>Vascular rings, previously undiagnosed in childhood</td>
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</table>
Idiopathic giant trachea bears the name of the clinician, Mounier-Kuhn, who originally described the radiographic and endoscopic appearance of a giant trachea in a patient with recurrent lower respiratory tract infections. As of 1988, 82 cases of Mounier-Kuhn disease had been described in the literature and to date, well over 100 cases have been described. The number of recognized cases has increased with the increased awareness of the disease and with the advent of CT scanning because the condition may not be apparent on plain chest radiographs. The diagnosis can be made when the right mainstem, left mainstem, and trachea exceed 2.4, 2.3, and 3.0 cm, respectively, as these measurements represent 3 SDs above the upper limit of normal in adults. The cause is unknown; however, in 1965, Johnston and Green described tracheobronchial diverticulitis or cysts. The literature also contains several reports of relapsing polychondritis, a disease that is characterized by recurrent inflammation and the destruction of the cartilaginous structures (tracheobronchial chondritis). In fact, respiratory tract involvement in relapsing polychondritis occurs in up to 56% of cases, but the respiratory symptoms are found on presentation in only 14% of cases. Patients with respiratory complications have a worse prognosis and poorer response to corticosteroids. Last, 26 cases of vascular rings have been diagnosed in adults, including double aortic arch and right aortic arch with an aberrant left subclavian artery and ligamentum arteriosum. As in children, these vascular malformations can cause TM and respira-

Acquired or Secondary Forms

A variety of processes can cause secondary TM and TBM in adults (Table 3). A sentinel review article by Feist et al in 1975 incorporated the data from the 1950s and 1960s and described the causes of adult TM. The authors identified tracheostomy or intubation with endotracheal tubes as the most common cause of secondary TM. Tracheostomy may lead to tracheal stenosis; however, it can also lead to frank weakening of the tracheal wall as a result of the destruction and loss of the supporting cartilage. This postintubation malacia is most commonly ≤ 3 cm in length and is segmental in nature. Although some injuries heal, factors such as recurrent intubation, the duration of intubation, and the use of high-dose steroids may predispose patients to developing progressive TM. The area of weakness can be seen not only at the stoma site but also at the inflatable cuff site. Occasionally, an additional abnormality may appear at the point where the tip of the tracheostomy tube impinges on the tissue. Possible causes of tracheal weakness in these areas include pressure necrosis, impairment of blood supply, and recurrent infections, as well as friction on, and inflammation of, the mucosa. Other forms of postransplantation TM may result from any injury causing a loss of cartilage from the trachea, including external trauma and surgery, such as lung transplantation. The TM that occurs in these situations should be differentiated from the less collapsible areas of anastomosis that may occur from trauma or surgery, which does not result in a weakness of the trachea.

Other authors also have suggested that chronic inflammation and irritants, such as cigarette smoke, are the most important contributors to the development of TM. A substantial proportion of patients with severe emphysema have some degree of malacia. The weakening of the tracheal wall may be related to the recurrent injury from cigarette smoke that leads to the emphysema, or it may merely be an extension of the peripheral hypermobility of the airways. Patients with chronic bronchitis may also have TM, which is thought to be secondary to the insult of recurrent infections, with or without cigarette smoking.

Additional etiologies for secondary TM exist. Chronic compression of the trachea resulting in TM most commonly results from benign mediastinal goiter, but it can also result from other sources of compression, including malignancies, abscesses, and cysts. The literature also contains several reports of relapsing polychondritis, a disease that is characterized by recurrent inflammation and the destruction of the cartilaginous structures (tracheobronchial chondritis). In fact, respiratory tract involvement in relapsing polychondritis occurs in up to 56% of cases, but the respiratory symptoms are found on presentation in only 14% of cases. Patients with respiratory complications have a worse prognosis and poorer response to corticosteroids.

Last, 26 cases of vascular rings have been diagnosed in adults, including double aortic arch and right aortic arch with an aberrant left subclavian artery and ligamentum arteriosum. As in children, these vascular malformations can cause TM and respira-
tory symptoms, including dyspnea, bronchitis, recurrent pneumonia, and stridor.183

Histopathologic Characteristics

Little is known about the histologic changes occurring in adult TM. In autopsy studies,150,184 the pars membranacea is dilated and flaccid, with anterioposterior narrowing of the bronchial lumen. Kienener et al185 described atrophy of the longitudinal elastic fibers of the pars membranacea, as did Herzog.186 Ikeda et al187 also described “fragmentation” of the tracheal cartilage. Jokinen et al176 also noted that the number of longitudinal fibers in the pars was clearly reduced throughout the entire length of the trachea above the bifurcation in one patient with TM compared to that in a control population. The number of longitudinal fibers did not differ from that of control subjects at the level of bifurcation or at the beginning of the mainstem bronchi.176 In this study, the total mucopolysaccharide, elastin, and collagen content in the cartilage of the trachea and bronchi were also similar.176

Incidence and Natural History

The occurrence of TM and TBM in the adult population is not uncommon. Although the Mounier-Kuhn form of the disease may have a genetic basis, the overwhelming majority of adults with TM and TBM have the acquired or secondary forms of the diseases. The true incidence of TM and TBM in adults is unclear because reports have been based on selected populations, instead of the population at large. In the late 1970s, Nuutinen5 and Jokinen et al176 greatly expanded the literature on acquired adult TM. Their data indicated that acquired TM was a disease of the middle-aged and elderly, most commonly seen in men > 40 years of age. Jokinen et al176 reported finding TM in 50 of 214 patients (23%) with a history of chronic bronchitis who were examined bronchoscopically.175 Herzog186 reported TBM in 16 of 1,500 patients (1%) undergoing bronchoscopy for various respiratory symptoms. In 1977, Jokinen et al176 also reported bronchoscopic findings for 2,150 Finnish patients with a range of symptoms and found that 94 patients (4.5%) had some form of malacia. Of these 94 patients, TM was diagnosed in 21 (22%), TBM in 59 (62%), and isolated bronchomalacia in 14 (15%). TM was seen much more commonly in men (82%) compared to women (18%), possibly reflecting the increased smoking prevalence in men at the time the study was conducted. It was seldom found in individuals without some evidence of obstructive lung disease.188 The most recent data on the incidence187 of airway collapse was > 50% in 542 of 4,283 patients (12.7%) with from pulmonary disease who underwent bronchoscopy. In that study, 72% of patients were aged 50 to 80 years.

TM is progressive in some patients,5,162,170 Jokinen et al176 performed repeat bronchoscopies on 17 of their patients with TM and TBM, and found that severity had progressed in 13 patients. Nuutinen189 reported a longitudinal study of 94 patients with TM and TBM with an average follow-up period of 5.2 years. Of those patients who had undergone repeat bronchoscopy, TM had progressed into TBM in six of nine patients, and bronchomalacia had progressed to TBM in all five patients. In no patient did the malacia improve. Some patients remained stable; however, the conditions of the majority of patients with mild-to-moderate disease worsened.

Symptoms

The main symptoms of TM in adults are dyspnea, cough, sputum production, and hemoptysis.5 These symptoms are nonspecific and are often attributed to emphysema, chronic bronchitis, cigarette smoking, or asthma. In fact, the patients may have coexistent emphysema, chronic bronchitis, or, less commonly, asthma or bronchogenic carcinoma.5 These patients may exhibit evidence of collapse of the upper airway during forced exhalation. There may be inspiratory wheezing or stridor. An associated barking cough, which has been likened to a barking seal, has also been reported. In addition to the more nonspecific symptoms, patients may also report dyspnea,150 episodic choking, chronic cough,180 hemoptysis,191 recurrent pulmonary infections,190 and even syncope associated with forced exhalation or cough.164,192,193 Unmasking of the disease with anesthesia194 and progressive hypercapnic respiratory failure195 have been reported. Some patients are asymptomatic until stressed by infection such as bronchitis or pneumo-

Diagnosis

The trachea and bronchi are normally flexible and compliant, and their diameters change during normal respiration. During inspiration, the airways dilate and lengthen, and during expiration, they narrow and shorten. Weakening of the tracheal wall accentuates these changes, and excessive changes in diam-

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eter may deform segments of the trachea, the entire trachea, and even the left and right mainstem bronchi. Because this deformation is a dynamic process that is accentuated by forced expiration, routine anteroposterior and lateral chest radiographs often show no abnormality. However, if TM is not isolated, but is caused by compression from other structures, such as mediastinal goiter, this deformation may be seen on plain radiographs.4

In an earlier time, tracheograms and bronchograms, which required the introduction of radiopaque material into the trachea, were performed to outline the bronchial tree and to evaluate the size of the structures.4 Cinetracheograms were used in the hopes of seeing “tracheal flutter,” and fluoroscopy was also used to estimate tracheal diameter. However, due to improved sensitivity, the convention became direct visualization by bronchoscopy to document a narrowing of at least 50% in the sagittal diameter. Some authors189 further classified the severity of malacia into mild, moderate, and severe. The malacia is deemed to be mild if the obstruction during expiration is to one half of the lumen, moderate if it reaches three quarters of the lumen, and severe if the posterior wall touches the anterior wall.189

Today, bronchoscopic visualization of dynamic tracheal or bronchial collapse remains the reference or “gold standard” for diagnosing TM. Although some studies use straining, coughing, and other maneuvers to elicit airway wall collapse, the expiratory effort to achieve collapse has never been standardized. Endoscopic visualization may be performed with either a rigid or a flexible bronchoscope. Flexible bronchoscopy is generally preferred because the patient is able to breathe spontaneously and follow commands to perform deep breathing, forced exhalation, and cough maneuvers to elicit the collapsibility of the airways (Fig 2). On direct visualization, the membranous trachea is widened and may even be redundant. There is obvious collapse of the airway during forced exhalation, and in some cases the airway lumen is completely obliterated as the membranous trachea collapses to the cartilaginous rings.

Emerging data196,197 suggest that dynamic CT scan images, although not the reference standard, are useful in diagnosing TM. Stern et al198 published data on the normal trachea during forced exhalation. They defined the normal range of intrathoracic tracheal diameters and cross-sectional areas during forced maneuvers, and showed that tracheal narrowing was about 50% in a patient with TM vs 35% in 10 healthy men. They then recommended using a cutoff of ≥ 70% narrowing on forced expiration as a diagnostic threshold for TM.198 Most other investigators, however, have employed the criteria of > 50% narrowing as a criteria for TM.191,196,199–203 Future studies of dynamic tracheal measurements involving a larger number of subjects of varying ages and both genders are necessary to more fully elucidate the normal range of tracheal collapsibility in the general population.

Although a diagnostic criterion of > 50% reduction of the cross-sectional area of the trachea during expiration has been widely used in both bronchoscopy and CT scan studies, it is important to note that the use of end-expiratory imaging rather than dynamic expiratory imaging may require a lower threshold criterion for diagnosing TBM. In this case, a different criterion for TM may be applied.

![Figure 2. Bronchoscopic view of TM. Left, A: normal status during inhalation. Right, B: near total collapse during quiet exhalation.](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/22023/ on 06/25/2017)
regard, Aquino et al\textsuperscript{197} compared cross-sectional area and coronal and sagittal diameters of the trachea between inspiration and end-expiration in 10 patients with TM and 23 healthy patients. Receiver operating characteristic curve analysis revealed that if a change between inspiration and expiration in the upper trachea exceeded 18\% and the change in the midtrachea exceeded 28\%, the probability of TM was between 89\% and 100\%.

The development\textsuperscript{204} of multidetector CT scanners, which permit imaging of the entire central airways in only a few seconds, allows for volumetric imaging of the airways during a single, dynamic, forced exhalation maneuver. Using this method, Gilkeson et al\textsuperscript{196} reported agreement between dynamic expiratory CT scan findings and collapsibility seen during bronchoscopy. Similarly, in a 2003 study that included 10 patients with bronchoscopically proven TBM, Zhang et al\textsuperscript{203} showed that dynamic expiratory CT scanning is highly sensitive for detecting malacia. These investigators also showed that a low-dose CT scan technique is comparable to a standard-dose technique for measuring the tracheal lumen during the dynamic expiratory phase of respiration. One year later, Zhang et al\textsuperscript{205} used dynamic expiratory volumetric CT scanning to compare 10 patients with bronchoscopically confirmed TM to 10 control subjects. Air trapping was seen at a higher frequency (TM patients, 100\%; control subjects, 60\%) and was more severe in the patients with TM\textsuperscript{205}.

Suto and Tanabe\textsuperscript{206} used dynamic MRI during forced expiration and cough to compare the collapsibility of the trachea in patients with TM to that of healthy subjects by using a "collapsibility index," originally proposed by Shepard et al\textsuperscript{207}. These investigators showed that a coughing maneuver elicited a significantly greater degree of collapse than forced end-expiration. Because of its lack of ionizing radiation, MRI has the potential advantage of allowing repeated assessments of the trachea during multiple respiratory maneuvers. Future studies comparing CT scanning and MRI are necessary to determine the relative sensitivities and specificities of these methods for diagnosing TM with conventional bronchoscopy as the "gold standard."

Although still in their youth, multiplanar and three-dimensional CT scan reconstructions, including virtual bronchoscopy, are promising imaging methods for the evaluation of TM. Although axial images suffice for assessing the airways that are perpendicular to the axial plane (e.g., the trachea and bronchus intermedius), they are less than ideal for evaluating airways that course obliquely (e.g., the mainstem bronchi). Paired end-inspiratory and dynamic expiratory virtual bronchoscopy images provide an important complement to axial CT scan images for these portions of the airway. Interestingly, in the study by Gilkeson et al\textsuperscript{196} virtual bronchoscopy images occasionally obviated the need for conventional bronchoscopy in patients who had relative contraindications to this procedure. With regard to multiplanar CT scan reformations, paired end-inspiratory and dynamic-expiratory sagittal images are helpful for displaying the craniocaudal extent of excessive tracheal collapse at end-expiration (Fig 3).

It is important to pay careful attention to technical parameters when performing CT scan examinations for assessing TM. For example, Burke et al\textsuperscript{208} described a series of patients with airway obstruction

![Figure 3. Two-dimensional CT scan airway reconstruction of the trachea. Left, A: the trachea during inhalation. Right, B: segmental tracheal collapse (arrow) during exhalation.](image-url)
evaluated by virtual endoscopy and bronchoscopy in which patients underwent imaging only at end-inspiration. Although the virtual technique was excellent at defining and measuring fixed lesions, it missed half of the laryngeal TM cases. The low sensitivity for detecting malacia in this study likely relates to the fact that these investigators performed CT scanning only at end-inspiration. Their results underscore the need to perform dual-phase (end-inspiratory and dynamic expiratory imaging) for assessing TM. Future studies of dual-phase virtual bronchoscopy are necessary to determine the precise role of this emerging technology in the diagnosis of TM.

Pulmonary function studies may be useful in evaluating a patient with suspected TM, but they are not diagnostic. Spirometry commonly reveals obstruction in proportion to the severity of malacia, however, this is not a universal finding. The pattern is usually that of a decreased FEV₁ and a low peak flow rate with a rapid decrease in flow (Fig 4). Airway resistance is almost always elevated. There is relative preservation of the shape of the inspiratory limb, Herzog, Gandevia, and Campbell et al. have reported that patients with tracheobronchial collapse frequently have a characteristic pattern on forced expiratory spiromgrams. In addition to a near complete absence of the usual sloping phase of the mid-portion of the curve, there may be a “break” or notch in the expiratory phase of the flow-volume loop. This break, first described by Koblet and Wyss, is presumed to be the point of MAC after the emptying of dead space air volume. It is also seen in moderate-to-severe emphysema patients who may or may not have TM, and it is difficult to determine the relative contributions of lung disease and TM if the conditions coexist. Flow oscillations, defined as a sequence of alternating decelerations and accelerations of flow, are often seen on the expiratory curve. In a large survey of 2,800 flow-volume loops, the incidence of these oscillations was 1.4%. These oscillations can be caused by redundant pharyngeal tissue, as in obstructive sleep apnea syndrome, structural or functional disorders of the larynx, neuromuscular diseases, and TM. Campbell and Faulks showed by esophageal balloon pressure measurements that the intrathoracic pressure was still rising sharply, while the expiratory velocity began falling rapidly from its peak. This condition is an exaggeration of the normal pressure-flow relationship observed during the development of flow limitation. Decreased peak flow is thus characteristic of the obstruction and hypercollapsibility of the airways.

Treatment

Many adults with TM and TBM do not require therapy because the finding may be incidental, and these diseases may not be causing symptoms. In a patient with symptomatic TM, care is initially supportive, unless the situation is emergent or progressively worsening. As TM frequently occurs in patients who also have COPD, the obstructive disorder optimally should be treated first. Bronchospasm must be controlled as it results in large pressure swings in the thorax, thereby worsening the degree of collapse of the malacic tracheal segments. This increased airway resistance and work of breathing can lead to respiratory failure. Once COPD has been controlled, a functional pulmonary baseline should be documented so that any response to an intervention for TM or TBM can be objectively evaluated.

If conservative measures fail, or if the patient is in critical condition, noninvasive, positive-pressure ventilation can be used in the short term to keep the airway open and to facilitate secretion drainage. Ferguson and Benoist have reported that nasal CPAP ventilation increases functional vital capacity and that increased levels of CPAP reduce dynamic

![Figure 4. Classic flow-volume loops for TM. There is a rapid decline in the maximal expiratory flow after a sharp peak associated with the collapse of central airways due to negative transmural pressure. Normal inspiratory profile is associated with dilation of the central airways during treatment with positive transmural pressure.](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/22023/ on 06/25/2017)
airway collapse. The notching on the flow-volume loop indicating airflow obstruction was abolished with the use of CPAP. In the hospital, the patient might require therapy with continuous nasal pressure for a short “recovery” time before transitioning to intermittent use until it is no longer needed.

In selected patients, surgery may be employed. Tracheostomy alone may be effective because the tracheostomy tube might either bypass the malacic segment or the tube itself might splint the airway open. If the patient has generalized and extensive TM, a longer tube may be necessary, as most of the more commonly used tubes are too short to prevent distal collapse, despite providing adequate proximal stenting. A tracheostomy also provides easy access for treatment with positive-pressure ventilation if required to maintain an open airway. Unfortunately, as in the pediatric population, tracheostomy may aggravate the underlying disorder and is, therefore, not a first-line treatment.

Herzog and Nissen and Nissen were the first to suggest surgical support for the pars membranacea by using a bone graft. Subsequently, a variety of prosthetic and autologous materials have been used as external stents. Amedee et al described 16 patients with TM who were treated with tracheal implantation of from one to three biocompatible ceramic rings. The procedure was successful in all patients, and reintubation or tracheostomy was not necessary in the 6.4 years of follow-up. Additionally, three patients who had been tracheostomy-dependent prior to the procedure tolerated decannulation after the surgery.

Recently, surgical placation of the posterior wall of the trachea with crystalline polypropylene and high-density polyethylene mesh has received attention. In that procedure, access is obtained through a right posterolateral thoracotomy, and the mesh is fashioned into a 2.5-cm wide strip, which is sutured to the posterior membranous wall. Thereafter, 2-cm sheets of mesh can then be sutured to the right and left mainstem bronchi. At our institution, this procedure is offered to patients who are good surgical candidates and in whom central airway stenting has improved symptoms. Conventional resection and reconstruction can be considered for treatment of focal malacia of the trachea. Last, although tracheal replacement has been talked about for > 50 years, to date no reliable substitute has been found.

An array of stents can be used to keep the airway open mechanically. Early attempts at stent placement for TM used stents and tubes that were designed for other purposes, namely, tracheostomy or esophageal intubation. As in pediatric patients, both rigid bronchoscopy and fluoroscopic guidance techniques have been used in adults. Some centers have also reported using flexible bronchoscopy with self-expanding stents in adults.

Metal stents have been used to manage airway obstruction resulting from many causes. They are easily placed by flexible bronchoscopy, are visible on plain radiographs, expand dynamically, and preserve mucociliary function. The most common complications resulting from the use of metal stents are the formation of granulation tissue, which may or may not require intervention, and breakage over time, which can cause severe problems including airway obstruction, airway perforation, and death. Additionally, embedded stents make future options such as surgical interventions difficult or impossible. These disadvantages, coupled with the fact that metal stents can usually not be removed easily, do not make them a first choice for patients with TM. Silicone stents, on the other hand, are easily inserted, repositioned, and removed. Placing these stents re-

![Figure 5](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/22023/ on 06/25/2017)
quires rigid bronchoscopy and general anesthesia. Although silicone studs on the surface of the stent retard migration, stent migration is still possible and may be heralded by a new cough. This problem requires direct visualization and repositioning, removal, or replacement of the stent. Hybrid stents and biodegradable stents are now being developed, but their utility for this disorder has not yet been evaluated.

Both subjective and objective means have been used to evaluate the efficacy of stent placement for benign airway stenoses. TM is included in many of these series, but there are no large series on TM alone. To describe the benefits of stent placement, clinicians have used improvement of respiratory symptoms, clearing of infectious processes, and lack of stent complications as end points. Researchers have also used bronchoscopic techniques and imaging techniques to show stent patency as a marker of success.

Most patients report immediate improvement in their respiratory symptoms once the stent is placed. Stents can immediately improve airflow dynamics in patients with benign airway obstruction, including TM but success has not been universal. Gotway et al also reported long-term pulmonary function data with stents placed for both stenotic and malacic lesions. At a mean of 15 months after stent placement, values for FEV₁, midexpiratory phase of FEF, and peak flow parameters had declined, despite the patients’ ongoing subjective improvement. The long-term improvement for the TM patients with relapsing polychondritis and idiopathic was better in the series by Gotway et al than it was in that of 11 patients with TM due to lung transplantation. This has been described previously and is not surprising, because 7 of the 11 patients in the study described previously received a single lung transplant for emphysema (the native lung would contribute to the degree of airflow obstruction). O’Donnell et al measured tracheal transmural pressure with esophageal balloons, assessed the cross-sectional shape of the trachea, and measured the critical pressure required to produce maximum expiratory flow in TM patients before and after stenting. They hypothesized that the critical pressure for flow limitation occurs before central airway collapse. In such circumstances, stenting the airway is unlikely to improve maximal expiratory flow. Therefore, a low critical pressure may be a marker for therapeutic failure. If airway stenting does not improve symptoms or the functional baseline of the patient, the stents should be removed to avoid any stent-related complications. At our institution, airway stenting is used mainly to identify the individual who is most likely to benefit from permanent or surgical airway stabilization. If improvement is present and the patient is a good surgical candidate, surgical tracheoplasty may be a preferred goal (Fig 5). If the patient improves but declines surgery, long-term stenting can be utilized, most commonly with silicone stents. Very rarely will metal stents have a role in the long-term management of this benign disorder. An algorithm of our approach to the patient with TM is outlined in Figure 6.

**Conclusion**

TM and TBM are becoming more commonly recognized and treated in children and adults. The causes and, therefore, the treatments vary, so a working knowledge of the options is important. Noninvasive imaging technology is increasingly being employed for diagnosis, and novel treatments, such as definitive surgical placation and stabilization with removable stents, are becoming alternatives to conservative interventions, such as CPAP. As a result of the complexity of these conditions and the options for treatment, these patients may best be assessed and managed individually in centers specializing in complex airway disorders.

![Figure 6. Treatment algorithm for adult TM.](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/22023/)
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REFERENCES

12 Gross RE, Neuhauer EBD. Compression of the trachea by an anomalous innominate artery; an operation for its relief. Am J Dis Child 1948; 75:570–577
34 Moe CA, Izuwaka T, Trusler GA. Innominate artery compression of the trachea. 1975; 101:733–738
41 Johnston RF, Green RA. Tracheobronchiomegaly: report of five cases and demonstration of familial occurrence. Am Rev Respir Dis 1965; 91:35–50
45 Snelling CE, Erb HI. Double aortic arch. Arch Dis Child 1933; 8:401–408
46 Heck HAJ, Moore HV, Lutin WA, et al. Esophageal-aortic erosion associated with double aortic arch and tracheo-
58 Krivchenia DI, Slepov AK, Chumakov LF. Intrathoracic compression of the respiratory airways by mediastinal teratomas in children. Lil Sprava 2001; 4:92–95
63 Bradham RR, Sealy WC, Young WG. Respiratory distress associated with anomalies of the aortic arch. Surg Gynecol Obstet 1968; 126:9–14
91 Griffiths H, Doull I, Williams RG, et al. Tracheomalacia and
107 Boiselle PM, Ernst A. Recent advances in central airway imaging. Chest 2002; 121:1561–1600
111 Fanst RA, Remley KB, Rimell FL. Real-time, cine magnetic resonance imaging for evaluation of the pediatric airway. Laryngoscope 2001; 111:2187–2190
114 Page BA, Klein EFJ. Tracheal stent as an aid in weaning from mechanical ventilation in tracheomalacia. Anaesthesiology 1977; 47:300–301
116 Levine PA, Sasaki CT, Kirchner JA. The long tracheostomy tube: alternative management of distal tracheal stenosis. Arch Otolaryngol 1978; 104:108–110


Czyharz ER. Ueber ein pulsionindiskret der trahe mit bemerkungen über das verhalten der elastischen fasern an normalen tracheen und bronchien. Zentralbl Allg Pathol 1897; 18:721–728

Mounier-Kuhn P. Dilatation de la trachee: constatations radhii-graphiques et bronchoscopiques. Lyons Medical 1932; 150:106–109

Lemoine JM. Dyspnees et retraction of the trachee and des grosses bronches. Semin Hop Paris 1949; 25:3984–3987

Pohl R. Universal conditions of malacia at the level of the tracheobronchial system (universelle erweichungszustande am tracheobroncialsystem (tracheo-bronchopthia malacica). Fortschr Geb Rontgenstr Nuklearmed 1951; 74:40–43

Herzog H, Nissen R. Relaxation and expiratory invagination of the membranos portion of the infratractic trachea and the main bronchi as cause of asphyxial attacks in bronchial asthma and the chronic asthmoid bronchitis of pulmonary emphysemata. Schweiz Med Wochenschr 1954; 84:217–221


Rahim AAA, Ahmed ME, Hassan MA. Respiratory complications after thyroidecotomy and the need for tracheostomy


186 Herzog H. Respiratory stenosis of the trachea and great bronchi by looseness of the membranous portion; plastic chip repair. Thoraxchirurgie 1958; 5:281–319


192 Demajumdar R, Rajesh PB. Have we got the full picture? J Laryngol Otol 1998; 112:785–789


