Tracheomalacia

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Tracheomalacia is a rare condition of the trachea involving a loss of rigidity of the cartilagenous wall of the airway with a resultant forward approximation of the membranous posterior wall, which narrows the lumen. The membranous wall can also become quite redundant, which further worsens the problem. The airway compromise is much worse in expiration when positive intrathoracic pressure further worsens the dynamic collapse of the airway. Symptoms from this condition include dyspnea on exertion, difficulty clearing secretions, and incessant cough. The cough is characteristic and allows the astute observer to make the diagnosis at the bedside. The cough, which is best described as seal-like, is presumably caused by the expiratory collapse and vibration of the floppy membranous wall against the anterior wall. Patients are often diagnosed with severe asthma and prescribed steroids to no avail.

Radiologic investigations

A variety of radiologic examinations can be helpful for diagnosing and monitoring tracheomalacia. A chest radiograph can be diagnostic in tracheobronchomegaly with tracheal air columns that are sometimes three times normal. Chest radiographs are not helpful in the typical adult case of chronic obstructive pulmonary disease (COPD) with suspected tracheomalacia. Fluoroscopy in the lateral position can be revealing in young children and in adults. Typical static (usually inspiratory) CT scans are likewise nondiagnostic because the trachea is often close to normal when intrapleural pressure is most negative during inspiration. CT scans can demonstrate other pathology that might relate to the airway (eg, vascular rings, substernal goiters). A combination of inspiratory and expiratory images from a CT scan is currently the easiest and most accurate way to make a noninvasive diagnosis of tracheomalacia. Aquino and colleagues recently reported a comparative CT study to detect acquired tracheomalacia using...
the differences in trachea size between inspiration and expiration [1]. On average, patients with tracheomalacia had a 44% reduction in cross-sectional area in expiration, whereas normal controls only had a 14% reduction. Based on receiver-operator curve analysis, a greater than 18% change in the upper trachea and a 28% change in the midtrachea were associated with a greater than 90% chance of having tracheomalacia. Figs. 1-3 illustrate CT scans of patients with adult acquired tracheomalacia.

Pulmonary function studies

Expiratory flow is severely diminished in tracheomalacia. The forced expiratory volume in 1 second (FEV1), forced vital capacity (FVC), and (most notably) the peak expiratory flow rate (PEFR) are markedly reduced. The flow volume curve confirms severe expiratory flow limitation with relative preservation of the shape of the inspiratory limb. Characteristically, the expiratory curve shows a low
maximal flow that quickly drops off close to the baseline, then a long plateau until end expiration [2]. The shape of the curve is similar to that seen in COPD patients and it is difficult to define the contribution of COPD versus tracheomalacia to airflow obstruction. Fig. 4 demonstrates the flow volume curve of a patient with tracheomalacia before and after membranous wall tracheoplasty.

4 Bronchoscopic evaluation

Bronchoscopy is the gold standard in the diagnosis of tracheomalacia because it is real-time and several provocative maneuvers can be done and immediately assessed. Bronchoscopy should be performed under local anesthesia with the

Fig. 4. (A) Preoperative flow-volume curve of a patient with adult idiopathic tracheomalacia demonstrating a reduced peak flow rate and a long plateau with minimal flow. (B) Postoperative flow-volume curve on the same patient (but on a different machine) demonstrating almost normal expiratory flow after membranous wall tracheoplasty.
patient alert and able to cooperate with instructions. The examination should be performed with quiet tidal breathing, varying degrees of forced expiration, and during coughing. The extent of malacia should be documented carefully. In the typical COPD patient, the malacic changes typically begin at the thoracic inlet and worsen as the carina is approached, then they lessen as the main bronchi are entered. Sometimes the main bronchi are severely involved, so it is important not to overlook the distal airways because this information is necessary to plan a corrective operation. In infants and young children, bronchoscopy is the diagnostic procedure of choice because radiologic techniques are insufficient to fully assess these small airways. Bronchoscopy is crucial after any sort of corrective operative procedure to ensure that the repair is technically satisfactory. Fig. 5 demonstrates bronchoscopic photographs of a patient with COPD-related tracheomalacia before and after membranous wall tracheoplasty.

**Congenital tracheomalacia**

Isolated congenital tracheomalacia can occur as a focal defect or (more commonly) as a diffuse weakness of the entire tracheobronchial tree. Congenital tracheomalacia most commonly occurs in association with esophageal atresia and tracheoesophageal fistula, and it usually presents shortly after repair of the esophagus. Tracheomalacia is sometimes present after division of a vascular ring, and if severe it requires repair. Tracheomalacia is most easily diagnosed just after division of the ring by performing intraoperative bronchoscopy before and after division of the ring. The treatment is usually to perform a pexy of segment of artery that is still compressing the airway [3-5]. Tracheobronchomegaly (Mounier-Kuhn syndrome) is a rare congenital diffuse dilatation and softening of the entire airway that usually presents in adults. Patients are usually misdiagnosed

Fig. 5. (A) Preoperative bronchoscopy on the patient from Fig. 1 confirming tracheomalacia during mild expiration. (B) Postoperative bronchoscopy on the same patient demonstrating marked improvement in the trachea.
with asthma and bronchitis. Patients usually have repeated episodes of respiratory tract infections and pneumonia and die prematurely.

Acquired tracheomalacia

Extrinsic compression

Chronic compression of the upper trachea is common in substernal goiters and is often a preoperative concern to the surgeon and anesthesiologist. There is no accurate test that can be done preoperatively that allows a determination as to whether or not malacia will be a postoperative problem. When the goiter is removed, the trachea almost always resumes a more normal configuration that allows a satisfactory airway. In the rare case in which there is an inadequate airway on trial extubation, the prudent course is usually to simply reintubate with an endotracheal tube and wait for several days to see if the airway will firm up. If this fails, treatment options include tracheostomy, insertion of a T-tube (the author's usual preference), or a tracheal resection (rarely required). Malacia of the trachea or main bronchus can also occur after late division of a vascular ring. Although it is not commonly done, intraoperative bronchoscopy allows immediate diagnosis of this problem and facilitates the decision as to whether or not pexy of the offending vessel is to be done and, if so, to what degree. Malacia of the main bronchus can occur (or, more correctly, become manifest) after treatment of the postpneumonectomy syndrome by mediastinal repositioning. The affected bronchus, which was previously stretched over the spine or aorta, does not spring back to normal shape and presents a difficult management problem. The problem is compounded by minimal pulmonary reserve and the foreign material in the pleural space, which makes open bronchus work less inviting. Options for treatment include aortopexy if there is a component of vascular compression, a long, custom-made T-tube (which acts as a stent), or resection of the malacic segment. The small subset of patients with postpneumonectomy syndrome is the group in which the author has had the most disappointing results, and it represents a real challenge.

Malacia associated with postintubation tracheal stenosis

Tracheomalacia occurs in postintubation stenosis from either ischemic necrosis of the cartilage from the balloon cuff or from chronic inflammation and infection of the tracheal wall. There is usually a subtle gradation of damaged trachea around a stenosis and the surgeon has little difficulty deciding what to save and what to remove. One difficulty comes in extensive resections in which one is tempted to preserve abnormal trachea to allow a resection and a malacic segment must be used. Minor degrees of airway softening can be tolerated, but one must avoid depending on an extremely malacic segment despite the illusion
of an adequate lumen in the trachea. Another situation in which malacia can be a factor is the segment of trachea between a tracheostomy stoma and the cuff stenosis more distal. This segment is often malacic if the tracheostomy is chronic, and this is sometimes underappreciated. If the whole segment of trachea (stoma, intervening segment, and cuff stenosis) is to be resected, the situation is straightforward. The more common scenario is that there is not enough trachea to resect all involved segments, then the intervening malacia comes into play. If the malacia is not bad, the stoma can be closed and these segments can be used in the reconstruction, which will reduce tension on the repair. If the malacia is severe, this segment must be resected and the stoma can either be closed or left open.

Relapsing polychondritis

Relapsing polychondritis is a systemic autoimmune disease that is thought to be the result of the destruction of cartilage by autoantibodies. Other cartilage besides the tracheobronchial tree can be affected, including the nose, ear, larynx, and articular cartilage. The destroyed cartilage is eventually replaced by fibrous scar tissue. As its name implies, the disease can wax and wane. Flares can be self-diagnosed by the patient with obvious inflammation and discomfort in the ears, nose, or joints. Most patients present with joint symptoms first. The disease can involve any segment of the airway and can skip segments to a degree. The disease affects men and women equally and is most common in the fourth decade of life. Women tend to have more airway involvement than men. The end result of cartilage inflammation can be a fixed obstruction, a soft, malacic airway, or some combination of the two. Treatment is primarily with anti-inflammatory agents. Nighttime continuous positive airway pressure (CPAP) can be helpful to provide airway support and relief of dyspnea [6]. If there is severe malacia of the proximal airways, a T or TY tube can provide good palliation.

Chronic obstructive pulmonary disease

Patients with advanced COPD often have a variable degree of atrophy of the cartilaginous support of the airway, which can lead to symptomatic tracheobronchomalacia if it is severe [7,8]. Why some patients are susceptible to this complication and most others are not is unclear.

The airway changes from its usual D shape into that resembling an archer's bow, with a long curve followed by flattening (or even a reverse curve) at the edges where the membranous wall begins. The membranous wall becomes increasingly redundant through time and leads to further flattening and obstruction of the airway. The symptoms that result are also compatible with COPD, making it difficult to suspect the diagnosis based on symptoms alone, which undoubtedly leads to difficulty in making a timely diagnosis. Most of the author's patients have symptoms suggesting tracheomalacia for years and have been
treated with steroids to no avail before the diagnosis was considered and made. Sorting out to what degree the symptoms are caused by COPD versus tracheomalacia is difficult because they are relatively similar. Some patients can describe an abrupt change in the nature of their symptoms. Treatment options include CPAP, silicone T-tube stents, and membranous wall tracheoplasty. The author does not recommend internal metal self-expanding stents because several patients have developed devastating complications from these stents including migration (not unexpected because the trachea is much larger than normal), tracheoesophageal fistula, obstructing granulation tissue at each end of the stent, and chronic stent infections. The author's general preference is to offer the patient a tracheoplasty if they are extremely symptomatic and in reasonable shape.

**Adult idiopathic tracheomalacia**

Rarely, adults without COPD present with this disease. It is unclear what (if any) predisposing factors lead to weakness of the airway cartilage. Relapsing polychondritis should be ruled out. Two of 18 patients that that author has operated upon have had this idiopathic variety.

**Membranous wall tracheoplasty**

In 1954, Herzog and Nissen introduced the concept of membranous wall tracheoplasty to improve expiratory airflow obstruction [7]. They first used bone grafts to plicate the floppy membranous wall and provide rigidity. Rainer later took up where they left off and introduced the use of homemade polypropylene sheets to plicate the membranous wall [8]. In some cases he used external polypropylene rings to try to add additional support to the airway. Of 12 patients, eight were long-term survivors and four significantly improved. At least four patients in his series died of late complications from the prosthetic material with erosion into the trachea, esophagus, and aorta [8]. For this reason, the author is loath to place external rings adjacent to the aorta. The polypropylene mesh available today is vastly superior to the atdimentary hard solid sheets available to Rainer, and the author has not had any late complications from mesh erosion. The author has performed membranous wall tracheoplasty in 18 patients through an evolution in use of material to plicate the posterior wall. Pericardium was used in two patients, Goreex in four patients, and polypropylene in 12 patients. The author now prefers polypropylene because the pericardium stretches with time and the Gorex (WL Gore & Associates, Inc., Flagstaff, AZ) does not incorporate into scar along the membranous wall, allowing fluid collections to build up. Good results were achieved in the majority of patients. The operation is done through a right thoracotomy in the fourth interspace. The airway is dissected out to expose the malacic segment, and the membranous wall is reed across its width with (typically) four mattress sutures (4-0 nonabsorbable), which are then brought
Fig. 6. Membranous wall tracheoplasty with numerous nonabsorbable mattress sutures fixing the posterior membranous wall to a sheet of polypropylene mesh to recreate the normal shape of the trachea. Note how the membranous wall is reefed because the mesh is cut to the normal width of the trachea, which imparts a normal bend to the cartilagenous wall (From Maddaus MA, Pearson FG. Tracheomalacia. In: Pearson FG, Cooper JD, Deslauriers J, et al, editors. Thoracic surgery, 2nd edition. New York: Churchill Livingstone; 2002. p. 324; with permission).

through the polypropylene mesh and tied. The sutures are partial thickness through the membranous wall to avoid mesh infections. The mesh is typically cut so it is 2 cm wide to recreate the normal width of the trachea. The length of the repair is dictated by the extent of the malacia as determined by the preoperative bronchoscopy. Bronchoscopy is performed as soon as the repair is complete to ensure a proper result. Fig. 6 illustrates the key aspects of the repair.

References


