Laryngotracheal stenosis is an umbrella description which encompasses a heterogeneous group of uncommon conditions that cause abnormal narrowing of the central airways from the supraglottic larynx to the main bronchi (Figure 39–1). The main symptoms of adult laryngotracheal stenosis are exertional dyspnea, effort intolerance, voice change, chronic cough and mucus, and, in a proportion of patients, added respiratory sounds. These sounds may resemble and, without a high index of clinical suspicion, can be readily mistaken for lower airway wheeze. The diagnosis of laryngotracheal stenosis, being a very uncommon cause of a very common clinical presentation, is often delayed and patients are frequently mislabeled diagnostically as “resistant asthmatics” and are treated incorrectly, in many cases for prolonged periods.

Laryngotracheal stenosis has been described since antiquity and its most common historical causes were infections and trauma. Iatrogenic laryngotracheal stenosis as a complication of tracheostomy insertion was recognized in the late 1800s. Laryngotracheal stenosis as a complication of translaryngeal intubation became increasingly prevalent from the middle of the twentieth century as a corollary of the birth and growth of intensive care medicine, which, at its inception and core, involved changing the management of acute respiratory failure from external negative-pressure iron-lung ventilation (Figure 39–2) to endotracheal intubation and positive-pressure ventilation. Despite significant advances in intensive care airway management, in the design of the tubes, in the meticulous attention that is being paid to their management, and in the earlier switch from translaryngeal to transtracheal ventilation, intubation and tracheostomy-related airway strictures remain the most common causes of laryngotracheal stenosis.

Other historical trends, which are beyond the scope of this chapter, have also influenced laryngotracheal stenosis disease patterns. As one example, introduction of systemic immune suppression in the 1980s and 1990s transformed the prognosis of patients with vasculitis from months to decades, and as a result, airway strictures, which are a long-term complication of avascular mucosal and cartilage necrosis, emerged as a disease entity. It occurs in approximately 15% of patients with granulomatosis with polyangiitis and now follows intubation-related strictures as the second most common cause of benign adult laryngotracheal stenosis.

A 49-year-old lady was admitted under the care of respiratory medicine with a working diagnosis of infective exacerbation of long-standing “difficult” asthma. She had been born uneventfully but had an emergency tracheostomy at 6 months of age for diphtheria and was successfully decannulated just before the age of 1 year. She was labeled a “wheezing” as a baby and toddler and a “difficult asthmatic” throughout childhood and adult life. She had numerous hospital admissions for “asthma attacks” in the decades that followed her tracheostomy decannulation.

On that particular admission, recognition of acute voice change by the medical team precipitated an otolaryngology consult for laryngeal evaluation. This resulted in the reevaluation of airway symptoms as
being more consistent with upper airway pathology, and the resulting computed tomography scan showed “a narrowing of the trachea. . . . This is quite focal extending over a vertical length of 5 mm. The trachea is narrowed to a minimal size of 9 × 15 mm. The appearance is that of scarring rather than of a soft tissue mass narrowing the trachea. The level of this narrowing is 15 mm below the cricoid cartilage.” A microlaryngoscopy and tracheoscopy was then performed and confirmed the diagnosis of an isolated lambdoid-pattern tracheal stenosis. She was offered a tracheal resection but declined. She was subsequently lost to follow-up of the local services and presented 10 years later, having had recurrent chest and throat infections and a refractory cough in the intervening years. She again declined the option of an open tracheal resection and did not present for regular follow-up upon discharge. Approximately 10 years later, aged 69, she presented to respiratory physicians with acute respiratory failure during an acute lower respiratory tract infection. She had been becoming increasingly effort intolerant, with worsening chronic cough and difficulties with expectoration of pulmonary secretions. She had no known cardiovascular morbidities.

On this occasion she had an in-office flexible laryngoscopy and tracheoscopy which confirmed the presence of a lambdoid-pattern tracheal stenosis with normal distal trachea and no evidence of malacic disease (Figure 39–3). She also had a maximum-effort flow-volume loop and whole-body plethysmography which showed an extrathoracic pattern of upper airway obstruction, normal lung volumes, and increased respiratory resistance (Figure 39–4).

The option of an endoscopic resection tracheoplasty had at this time become available within the unit and she elected for this course of treatment. The procedure was successfully undertaken (Figure 39–5 and Video 39–1). This led to normalization of her post-operative flow-volume loops (Figure 39–6), but somewhat surprisingly, she subjectively remained breathless and did not feel that she had derived any significant benefit from her surgery. She underwent an airway-focused cardiopulmonary exercise test (CPET) according to a protocol developed in partnership with the New Zealand Sleep and Respiratory Institute (www.nzrsi.co.nz) to evaluate the upper airway. She was able to exercise for 4.07 minutes and had a peak oxygen consumption of 22.5 mL/kg/min, which was 82% of her

Figure 39–2. Negative-pressure ventilation using the iron lung.
Figure 39–3. Four-shot flexible endoscopic views of the larynx, subglottis, trachea, and the carina showing a lambdoid-pattern tracheal stenosis.

Figure 39–4. Maximum-effort flow–volume loop and whole-body plethysmography prior to airway surgery. The blue portion of the loop is a maximum-effort expiration from full lung volume. The red component of the flow–volume loop shows expiratory flow limitation due to the presence of the stenosis. The green component of the loop shows the phenomenon of expiratory flow limitation which continues until full expiration, at which point a maximum-effort breath (gray line) is taken back to full lung volume. Spirometry shows normal forced expiratory volume in one second (FEV1), increased forced vital capacity (FVC), reduced peak expiratory and peak inspiratory flow rates (PEFR and PIFR), and an increased Expiratory Disproportion Index (EDI). Whole-body plethysmography showed normal lung volumes (total lung capacity [TLC], vital capacity [VC], inspiratory capacity [IC], functional residual capacity [FRC], expiratory reserve volume [ERV], residual volume [RV], and thoracic gas volume [Vtg]). The airway resistance was significantly increased (Raw) and admittance (Gaw).
predicted value. There were no desaturations and no cardiac ischemia. She reached a peak exercise power of 138 watts and stopped due to a combination of breathlessness and leg fatigue. She had a Borg dyspnea score of 0 at the start of the exercise and 9 at the end. She was able to raise her minute ventilation in response to increased exercise power and did not have a major fall in the expired partial pressure of oxygen, nor a major rise in the expired partial pressure of carbon dioxide (Figure 39–7). Her exercise flow-volume loops, performed by asking her to perform full inspirations at different exercise powers (Figure 39–8), showed an abnormal persistent breathing pattern, characterized by a failure to increase both the respiratory rate and per-breath flow rate in response to increased exercise demand, but instead taking deeper and slower breaths in order to increase minute ventilation. The signal for the change in breathing pattern was reaching expiratory flow maxima (Figure 39–8-5). She closely approached but did not reach the inspiratory flow maxima (Figure 39–8-8). She underwent a period of respiratory rehabilitation which led to resolution of her dyspnea symptoms and she remains asymptomatic over 1 year later.

Figure 39–5. Endoscopic resection tracheoplasty. The operation involves laser resection of herniated tracheal ring down to tracheal adventitia to allow wound contracture to proceed cranio-caudally. It is important to maintain both anterior and posterior mucosal bridges.

Figure 39–6. Maximum-effort flow-volume loops before and one week after endoscopic resection tracheoplasty showing significant improvements in both inspiratory and expiratory flow rates.

Figure 39–7. Airway-focused cardiopulmonary exercise test showing increases in minute ventilation, and expired partial pressures of oxygen and carbon dioxide. Supplied courtesy of Dr Andrew Veale at the New Zealand Respiratory and Sleep Institute.
Figure 39–8. Exercise flow-volume loops constructed within the maximum-effort flow-volume loop envelope showing an abnormal pattern of breathing consisting of failure to raise ventilatory flow rate in response to increased exercise demand. Supplied courtesy of Dr Andrew Veale at the New Zealand Respiratory and Sleep Institute.
DISCUSSION

Recognition and management of airway stenosis may have a marked impact on quality of life and it is important for a multidisciplinary approach (respiratory, otolaryngology, radiology, gastroenterology, speech pathology, physiotherapy) to be employed to ensure that a correct diagnosis (and therefore appropriate management plan) is reached.

Incidence of Intubation-Related Laryngotracheal Stenosis

Adult intubation-related laryngotracheal stenosis is estimated to occur in 1 in 204,000 adults per year. However, this number relates to patients who presented for treatment, and it is further estimated that as many as 80% of patients with this condition may actually remain undiagnosed.

Pathophysiology of Intubation-Related Laryngotracheal Stenosis

This condition is a response to injury caused by the presence of a translaryngeal or a transtracheal ventilatory conduit and, as such, the locations and patterns of stenosis are specific and consistent (Figure 39–9). It may also arise in the context of particular systemic diseases or where there is significant extra-esophageal reflux combined with airway instrumentation. A detailed description of normal and abnormal mucosal wound healing at a cellular level is beyond the scope of this chapter and has been provided by Sandhu and Nouraei.

Diagnostic Evaluation of Intubation-Related Laryngotracheal Stenosis

Correct diagnosis will enable specific targeted treatment and is the key to patient symptom relief.

Clinical History

In addition to a comprehensive general medical history, an airway history aims to answer a number of specific questions:

- How Have the Symptoms Evolved? There is no period of intubation time below which a significant airway injury is improbable, nor is there a period of intubation time above which such an injury becomes inevitable. Particular attention must therefore be paid to ascertaining a detailed history of surgical and critical care intubations, including, as was the case in the patient presented, events during early years of life. A significant mechanical obstruction, once established, will very likely have respiratory manifestations, but these manifestations will often have been attributed, sometimes for many decades, to other conditions like asthma or post critical care bronchopulmonary morbidity.

- As such, a detailed respiratory history preceding identification of laryngotracheal stenosis must be sought. One exception to this, which must be borne in mind, is development of new symptoms in a male patient with a history of neonatal intubation, at or around puberty due to what is a complex interaction between pre-existing but hitherto asymptomatic anatomic anomalies and the laryngeal growth spurt. In cases of relatively recent intubation events, the length of time from extubation to the onset of airway symptoms must be sought. It can take up to one year from extubation for maturation of an airway injury into a symptomatic stenosis but most patients with significant stenoses
become symptomatic within 3 months. Breathlessness is not a symptom that is primarily evaluated by laryngologists, and, as such, most tracheostomy-free breathless patients are referred with established diagnoses. An important exception to this, although not directly related to intubation-related laryngotracheal stenosis, is development of airway strictures in patients with granulomatosis with polyangiitis who are under otolaryngologist follow-up for nasal and sinus symptoms. To avoid potentially life-threatening diagnostic delays, new-onset dyspnea should be inquired about in the otolaryngological follow-up of vasculitis patients and appropriate investigations ordered.

What Are the Risk Factors for Intubation-Related Laryngotracheal Stenosis? There are a wide range of risk factors which influence occurrence of intubation-related laryngotracheal stenosis (Figure 39–10). An effort must be made to obtain details of the intubation event preceding development of the stenosis and particular attention must be paid to agitation and problems with sedation during intubation, persistent hypotension, use of inotropes, presence of immune suppression, endotracheal tube size selection, and time from endotracheal to tracheostomy ventilation.

With regard to tube size selection in particular, it is important to recognize that height and sex exert identical influence on adult trachea size and yet, in many cases, endotracheal tube size selection in adults is based on patient sex and not height and sex. As such, and particularly in relation to posterior glottic injury, an over-sized endotracheal tube may be a specific and iatrogenic etiological factor.

What Is the Likelihood of the Presence of a Different Airway Pathology? There are diagnostic pitfalls that may lead to misattribution of an airway stenosis as being intubation related. An airway stenosis may develop as a complication of a vasculitic process, principally granulomatosis with polyangiitis, or de novo in idiopathic subglottic stenosis. Stenosis progression may cause increasing but misattributed respiratory morbidity and precipitate respiratory failure, the treatment of which may then require endotracheal intubation or tracheostomy tube placement. A careful clinical history of vasculitis, which should be ascertained in all patients,
if necessary using a formal symptoms inventory instrument like the Birmingham Vasculitis Activity Score, and a precise understanding of the evolution of symptoms should reduce the likelihood of diagnostic misattribution. This pitfall may also be avoided by paying close attention to the precise stenosis geometry. The subglottis is not the narrowest part of the adult airway and only a small minority of long-term intubation-related airway injuries occur in the true subglottis. By contrast, both vasculitis-related and idiopathic stenoses frequently, although not exclusively, occur in the true subglottis, and the endoscopic appearance of the stenosis (see Figure 39–1) can provide significant clues about the underlying etiology.

What Are the Current Symptoms and Disability Levels? A laryngotracheal stenosis impacts multiple domains of patient symptom and often has a profound impact on well-being. The four principal symptom domains are dyspnea, voice, swallowing, and cough. Two additional domains—of airway and independence—also influence the patient’s overall quality of life. The ADV-CSI score (Table 39–1) provides a disease-specific symptoms inventory and complications grading systems both for initial evaluation and for outcomes assessment.

What Are Patient Expectations and Prognostic Considerations in Determining Management Strategies? The aim of laryngotracheal reconstructive surgery is to restore an airway lumen that can support the ventilatory demands of the patient while minimizing collateral injury to the voice and swallowing mechanisms. This aim is achievable in most patients who have intubation-related laryngotracheal stenosis, and indeed in many cases, this can be achieved using only minimally invasive surgical techniques. However, patients with extensive injuries, those with significant concurrent laryngeal and tracheal injuries, and patients with laryngeal stenosis who have borderline pretreatment swallow safety may need to be maintained with long-term luminal stents or a long-term tracheostomy. Likewise, in patients who have long-term neurological injury whose respiratory demands are likely to remain minimal in the long term, performing major open cervicomedialisternal surgery, while technically feasible, may not serve the best holistic interests of the patient, and treatment goal may need to shift toward creating an airway that will likely remain safe during intercurrent episodes of lower respiratory tract infection. The appropriate treatment goals and approach are frequently nuanced and should, in all cases, be a shared decision between the patient and the airway team.

Clinical Examination

Thorough evaluation is always warranted by the surgeon, even if the patient has been reviewed by other services.

General Examination. The most common clinical scenarios in which intubation-related laryngotracheal stenosis is encountered are a chronically breathless or tracheostomy-dependent patient with an established diagnosis, a failed or difficult critical care extubation/decanulation, and an acute clinical presentation with respiratory decompensation. In the acute settings, clinical examination follows standard intermediate and advanced life-support protocols. Management of an acutely compromised airway is discussed elsewhere in this volume. General examination aims to ascertain the degree of respiratory effort through assessing for tracheal tug, use of accessory respiratory muscles, chest recession, and stridor. Stridor should be elicited by asking the patient to take deep breaths through an open mouth while the neck is auscultated. Whether the stridor is inspiratory, expiratory, or biphasic should be documented. Oxygen saturation on room air needs to be measured. The patient should be examined for peripheral stigmata of abnormal scarring like keloid and hypertrophic scars, presence of syndromes associated with abnormal scarring like Turner’s or Noonan’s, connective tissue disorders like joint hypermobility associated with Ehlers-Danlos syndrome, and immune-related conditions like sarcoid nodules, saddle-nose deformity of granulomatosis with polyangiitis, and lobule-sparing ear inflammation of relapsing polychondritis.

Laryngotracheoscopy. Historically, airway assessment required an examination under general anesthesia using suspension laryngoscopy, and for some conditions, specifically for bilateral vocal fold immobility when palpation of the cricoarytenoid joints is required, this remains a minimum standard of care. For most tracheal conditions, however, an office-based laryngotracheoscopy is readily feasible. Key pharyngeal findings to document are presence of scars and hypertrophic secretions which can be a sign of abnormal swallowing. Stigmata of pharyngolaryngeal reflux, including inflammation of the respiratory mucosa within the postnasal space, cobblestoning of the posterior
Table 39–1. The ADV-CSI System and Classification of Airway Stenosis and Complications

Please indicate which of the five responses below best describes your level of breathlessness over the past two weeks. (only one response out of the five available options below).

**Dyspnoea**
1. I get short of breath only on strenuous exercise.
2. I get short of breath when hurrying on the level or climbing up a slight hill.
3. I walk slower than people of the same age on the level because of breathlessness, or have to stop for breath when walking at my own pace on the level.
4. I stop for breath after walking 100 yards or after a few minutes on the level.
5. I am too breathless to leave the house

Please indicate which of the five responses below best describes your voice over the past two weeks (only one response out of the five available options below).

**Voice**
1. I have had no problems with my voice
2. I have had some problems with my voice, for example the quality of my voice may vary throughout the day, or I have difficulty being heard in loud environments
3. I struggle to make my voice heard, particularly in loud environments
4. Despite my best efforts, I can only produce a weak voice/whisper and have difficulty being heard in a normal conversation/on the phone
5. I have no voice

Please indicate which of the five responses below best describes your voice over the past two weeks (only one response out of the five available options below). If you have had any episodes of being unable to breathe/having to go to hospital because of mucous plugs or crusting please since the last time you took this test, choose option 5.

**Cough / Mucus**
1. I have had no problems with coughing or with mucous in my airway or throat.
2. I do have a fairly regular cough and/or need to clear mucus, but it does not bother me.
3. I do have a bothersome problem with cough and/or mucus. For example:
   • Problems with cough/need to clear mucus causes me physical pain/discomfort (eg rib/throat pain).
   • Problems with cough/need to clear mucus has an impact on my social life.
4. I have a significant problem with coughing and/or mucus. For example:
   • I regularly have to clear clumps or mucus/crust from my throat/airway.
   • I have experienced at least one episode of my airway “blocking” due to mucous/crust which I had to clear with coughing/nebulising.
   • I have needed to see a doctor regarding my cough / mucous symptoms.
5. Since the last time I answered these questions, I have needed to call an ambulance / attend hospital in an emergency due to my airway blocking off with "mucous plugging" / "airway crusting."

Please indicate which of the five responses below best describes your use of devices (eg humidifiers or nebulisers) for your airway over the past two weeks (only one response out of the five available options below).

**Independence**
1. I have not needed to use any devices (eg nebuliser or humidifier) for my airway.
2. I have needed to humidify my airway (either by steam inhalation or by use of a humidifier), and/or use a saline nebuliser. I have not needed to do this more than 2-3 times a week.
3. I have needed to use a nebuliser or humidify my airway one or more times a day.
4. I have an internal airway stent in place.
5. I have a tracheostomy or a T-tube in place.
Please indicate which of the five responses below best describes your swallowing over the past two weeks (only one response out of the five available options below).

**Swallowing**

1. I have been able to eat and drink normally.
2. I have been able to eat a normal diet but with some difficulty. For example:
   - I have occasionally had to cough to clear my throat
   - I find some foods more difficult than others to swallow
   - It takes me longer to finish a meal than it does people around me
   - I sometimes cough when I drink liquids quickly
3. I have had significant swallowing difficulties. For example:
   - I cough to clear my throat, or do a double-swallow during most meals
   - I tend to eat soft or pureed foods, that are easier to swallow.
   - It takes me much longer to finish a meal than most people
   - Drinking fluids, frequently makes me cough.
4. My swallowing is a serious problem / is seriously abnormal. For example:
   - My diet consists almost entirely of semi-liquid / liquidized foods
   - I need to take a significant amount of the fluids I drink, as thickened fluids
   - I take regular dietary supplements -or- I receive a proportion of my diet
     through a stomach tube (PEG).
5. I am unable to swallow. I take all of my nutrition through a stomach tube (PEG).

Please indicate which of the five responses below best describes your overall sense of health and well-being over the past two weeks (only one response out of the five available options below).

**Overall Health**

In general, I would say that my health is:

1. Excellent
2. Very good.
3. Good.
4. Fair.
5. Poor.

**Modified Myer-Cotton grading system**

Grade 0. No or minimal (<10%) discernible stenosis
Grade 1. Discernible obstruction between 10 and 50%
Grade 2. Obstruction between 51 and 70%
Grade 3. Obstruction between 71 and 99%
Grade 4. No discernible lumen

**Dindo–Clavien classification of complications.**

Grade 1. Any deviation from the normal postoperative course without the need for pharmacological intervention 1 or surgical, endoscopic, or radiological interventions.
Grade 2. Required pharmacological treatment with drugs other than such allowed for grade 1 complications. Blood transfusion and total parenteral nutrition are also included.
Grade 3. Requiring surgical, radiological, or endoscopic interventions.
Grade 4. Life-threatening complications (including Central Nervous System complications) 2 requiring intermediate or intensive care management.
Grade 5. Death of a patient.

1. Allowed therapeutic regimens are: drugs as antiemetics, antipyretics, analgesics, diuretics, electrolytes, and physiotherapy. This grade also includes wound infections opened at the bedside.
2. Brain haemorrhage, ischaemic stroke, subarachnoid haemorrhage but excluding transient ischaemic attack.
pharyngeal wall, post-cricoid and inter-arytenoid edema, and glottic pseudosulcus should be ascertained and documented. The larynx should be examined for gross movement and a stroboscopic assessment of vocal vibrations should be performed.

The subglottis and proximal trachea can be examined without the need for topical anesthesia by performing a Laryngeal Distraction Maneuver (LDM) (Figure 39–11). This brings the glottic inlet and the trachea into bird’s-eye view of the endoscope. The patient is asked to take slow and deep breaths in order to demonstrate the subglottis and the trachea. The same position, if instead of deep inspiration is accompanied by a trumpet Valsalva maneuver, opens and demonstrates the post-cricoid space. Full tracheal examination can readily be performed by instillation of 3 to 5 mL of 2% lidocaine across the laryngeal vestibule and lumen. This can be administered through a mucosal atomization device (Figure 39–12), through the working channel of a laryngoscope, or through a thyrohyoid or cricothyroid injection. This procedure is safe to perform in the clinic but as with all airway interventions, full resuscitation facilities and personnel, and oxygen, must be available. A note of caution is performing this procedure in patients with laryngospasm, secondary breathing-pattern disorders, or high anxiety. In these patients, temporary inability to sense airflow through the larynx or trachea may cause claustrophobia and precipitate either laryngospasm or poorly controlled hyperventilation, which may then worsen airflow across a stricture. If a tracheoscopy is performed, then four standard views (see Figure 39–1V1-V4) should, as a minimum, be documented, and in patients with granulomatosis with polyangiitis or previous tuberculosis, the left and right main bronchi should also be visualized. These assessments may be readily performed using a standard flexible laryngoscope with video recording functionality.

**Figure 39–11.** The laryngeal distraction maneuver is accomplished by asking the patient to move forward and bend in the upper back, to put his or her chin down onto the chest and to look to the side. In this position, the larynx and trachea are brought into the bird’s-eye view of the endoscope and the epiglottis is pulled anteriorly. With careful maneuver it is possible to obtain full views of the subglottis without needing to administer anesthesia. The same position, when combined with a trumpet Valsalva maneuver, allows for visualization of the posterior cricoid region and the upper esophageal sphincter.

**Figure 39–12.** Use of a mucosal atomization device (MAD) to anesthetize the larynx and trachea to allow in-office laryngotracheoscopy.
Investigations

Supplementary investigations can provide stratification and prognostic assistance.

Blood Tests. All patients should undergo a complete blood count, basic blood biochemistry, and thyroid function tests. Inflammatory markers including erythrocyte sedimentation rate and c-reactive protein should be measured. An immune screen including angiotensin converting enzyme (ACE), anti-nuclear cytoplasmic antibody (ANCA) including myeloperoxidase (MPO) and proteinase 3 (PR3), and rheumatoid factor (RF) titers should be measured as screening for systemic inflammation and vasculitis. In very specialist contexts other tests like matrilin assays for relapsing polychondritis may be considered.

Flow Physiology. All patients should undergo a maximum-effort flow-volume loop at each visit to the airway unit (Figure 39–13) and should be supplied with a flow meter to measure at least peak expiratory flow at regular intervals (Figure 39–14).

Effort Physiology. Cardiopulmonary exercise testing (CPET) should become a routine test for evaluating laryngotracheal stenosis. It has proven utility in differentiating between causes of dyspnea. It can also, as shown in the case presented, identify breathing-pattern disorders and may come to find particular utility in reducing the likelihood of secondary breathing pattern disorders developing in patients with recurrent disease processes like idiopathic subglottic stenosis, or in patients with chronic marginal laryngeal airways. Other tests like the six-minute walk test or shuttle test may also provide information about effort intolerance but they provide significantly less information compared with CPET.

Cross-sectional Imaging. Cross-sectional imaging and in particular, computed tomography of the neck and chest, has historically used to establish the diagnosis of laryngotracheal stenosis. Imaging can now be reconstructed to provide 3-dimensional lumen views (Figure 39–15), and efforts are being made to investigate flow dynamics within reconstructed lumen geometries. In practice, most patients continue to undergo

Figure 39–13. Maximum-effort flow-volume loops before and after treatment of an obstructive intubation granuloma arising from the medical surface of the left arytenoid cartilage. Features of note are improvement in the inspiratory portion of the loop, increase in peak expiratory flow and “sharpening” of the “expiratory peak.”
Figure 39–14. Biweekly rolling average of peak expiratory flow rates of one patient with idiopathic subglottic stenosis, measured daily for over 3 years, following treatment of idiopathic subglottic stenosis with endoscopic laryngotracheoplasty with biological inhibition. Changes in peak flow rate clearly identify initial and subsequent treatment failures.

Figure 39–15. Computed tomography and 3-dimensional airway lumen reconstruction views from CT scans. The endoscopic image corresponds to the point of maximum narrowing of the stenosis.
CT scanning or are referred with cross-sectional imaging already performed. Cross-sectional imaging is useful in evaluating extraluminal causes of airway stenosis like vascular anomalies or compressive adenopathy. It also provides a useful screen of the lung tissue. It provides no information about mucosal status and in particular, presence of acute inflammatory stenoses, and as such, it does not by itself provide an adequate airway assessment and must, in all cases, be supplemented by direct visualization of the endoluminal mucosal surfaces.

**Voice Analysis.** Patients should undergo objective voice analysis. As a minimum, this should include perceptual voice assessment which may be performed using GRBAS or CAPE-V systems, and phonation time. Aerodynamic measures including transglottic flow rates and derived subglottic pressure may also provide insights. Strong consideration should be given to performing electroglottographic analysis of connected speech in all patients.\(^{39,40}\)

**Tests for Reflux.** There should be a low threshold for requesting 24-hour pH and impedance monitoring, especially for patients who are being considered for open laryngotracheal surgery.

**Swallowing Function Tests.** All patients who are being considered for open airway surgery, and all patients who are being considered for endoscopic surgery to the larynx, should undergo swallow function testing. A videofluoroscopic swallowing study (VFSS) is the preoperative investigation of choice and a minimum standard of care, while functional endoscopic evaluation of swallowing (FEES) may be preferred for monitoring swallowing safety following airway surgery.

**Tissue Biopsy.** Diagnosis of airway obstruction secondary to intubation-related laryngotracheal stenosis does not require histological confirmation. However, biopsies are useful in establishing diagnoses other than intubation-related stenosis, including vasculitis,\(^{19}\) noncaseating granulomata in sarcoidosis,\(^{41}\) Mickulicz cells in rhinoscleroma,\(^{42}\) apple green birefringence with polarized light examination in amyloidosis,\(^{43}\) immunofluorescence studies in mucosal pemphigoid,\(^{44}\) and different tumor pathologies. At present, the differentiation between acute inflammatory and mature fibrotic stenoses is endoscopic (Figure 39–16) but it is likely

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**Figure 39–16.** Left images show endoscopic and microscopic images of acute inflammatory airway stenosis. Right images show endoscopic and microscopic images of a mature fibrotic airway stenosis.
that as novel therapeutic approaches become available and scar tissue modulation becomes more tailored to the underlying abnormalities of the scar formation pathways, the role of biopsies to guide personalized treatment approaches will increase.

**Management of Intubation-Related Laryngotracheal Stenosis**

Extent of pathology and underlying etiology will help determine effective management strategies.

**Acute Inflammatory Obstructions Versus Mature Fibrotic Strictures.** The prime consideration in managing adult intubation-related laryngotracheal stenosis is whether the lesion is acute and inflammatory or chronic and fibrotic (Figure 39–17). Acute inflammatory lesions almost invariably respond to treatment with intralesional depot corticosteroids, judicious laser treatment, controlled radial dilation, and regular follow-up with repeat procedures to manage the evolution of the scarring process.

**Laryngeal Versus Tracheal Stenosis.** The next consideration is the exact location and pattern of the stenosis, what specific structures have been damaged, and whether or not the stenosis involves the larynx. Management of intubation-related bilateral vocal fold immobility continues to evolve. Vocal cordotomy or a partial arytenoidectomy are minimally invasive procedures but ones that can be associated with voice morbidity, airway violation of food and fluid, and a recurrence rate. A laryngotracheal reconstruction, consisting of a laryngofissure combined with costal cartilage graft, and placed under a post-cricoid mucosal flap for posterior glottic wound closure combined with intra-articular cricoarytenoid joint surgery represents a major paradigm shift from static glottic widening toward laryngeal remobilization and restoration of respiratory and phonatory laryngeal movements. The cricoarytenoid joint is unusual in that it can remain viable for up to 17 years after denervation and in cases of joint ankylosis, careful intra-articular division of adhesions or partial excision of the joint may restore laryngeal mobility. In this context, a comparison may be drawn between mobility of an abnormal cricoarytenoid joint and a Girdlestone hip. The principal approach for achieving laryngeal remobilization is through the use of the post-cricoid flap combined with intra-articular joint remobilization. This operation may be performed endoscopically in many cases or, in cases of damaged cricoid cartilage or unfavorable transoral access, may be performed as a combined glottic reconstruction via a laryngofissure combined with costal cartilage grafting that is placed under a post-cricoid mucosal flap for primary mucosal wound closure (Figures 39–18 and 39–19).

**Tracheal Stenosis Configuration.** There are a number of classification systems for laryngotracheal stenosis and the best known is the Myer–Cotton grade (see Figure 39–1M). This system characterizes the degree of cross-sectional obstruction as being less than 50%, between 51% and 70%, between 71% and 99%, and 100%. It was devised for pediatric airway surgery where the subglottis is the narrowest part of the airway and the severity of subglottic injury is reflected in the degree of cross-sectional narrowing. As such, in children, the degree of cross-sectional impairment provides information about disease prognosis. This is not the case in adults, where in practice, although the degree of cross-sectional obstruction is often documented, it does not provide the same prognostic information compared with pediatric cases. Figure 39–20 provides a framework for identifying and documenting individual components of a global approach to airway assessment. Specific injuries may be managed using specific procedures. For example, an isolated cricoid injury can be managed with a hyoid-on-sternothyroid flap or a crico-tracheal resection using the trachea to reconstruct the anterior cricoid arch. A lambda deformity may be managed using endoscopic resection tracheoplasty, as demonstrated in the case presented. Structural tracheal collapse, if resectable, should be managed with tracheal resection and unresectable lengths may be reconstructed with augmentation tracheoplasty or maintained with long-term stents. In selected cases, an injury may heal in a favorable position around a long-term soft silicon tracheal stent. There are very few, if any, indications for circumferential tracheal replacement for...
Figure 39–17. A decision tree and images of key surgical procedures used for managing intubation-related laryngotracheal stenosis.
Figure 39–18. Combined glottic reconstruction procedure for treating bilateral vocal fold immobility due to interarytenoid scarring in the presence of poor transoral access and/or injury to the cricoid cartilage, precluding transoral reconstructive laryngeal microsurgery.

Figure 39–19. Outcome of combined glottic reconstruction four months after treatment, showing restoration of laryngeal mobility, lowering of post-cricoid height, and healed tracheostomy scar.
benign intubation-related laryngotracheal stenosis and in particular, the bar for considering tracheal transplantation for this condition should be set extraordinarily high indeed. In that highly unusual scenario where any form of circumferential tracheal replacement for benign intubation-related laryngotracheal stenosis is contemplated, an autologous replacement using the Dartevelle or Olias procedures should be considered.

**Structural Collapse Versus Lamina Propria Fibrosis.** Patients with lumen-encroaching strictures within a normal cartilaginous framework warrant special consideration. An interesting trend has been the change in dominant pattern of intubation-related tracheal stenosis from a combined mucosal and cartilaginous injury that is associated with the collapse of the tracheal structure, to a lamina propria disease characterized by formation of a lumen-encroaching cicatrix within a largely intact tracheal cartilaginous framework (Figure 39–21). This may be a corollary of greater awareness of airway injuries by intensive care clinicians and as a result, better care of endotracheal tubes and earlier switch to tracheostomy. In this context, if a stenosis still develops, and particularly an injury that does not involve

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**Figure 39–20.** A framework for outcomes documentation in laryngotracheal stenosis.
**Figure 39–21.** Pathophysiology of laryngotracheal stenosis as a function of depth of injury.

**Intrinsic Pathologies**

Intrinsic laryngotracheal diseases include idiopathic subglottic stenosis and sarcoidosis. In these cases disease results from fibro-inflammatory expansion of submucosal tissues overlying normal cartilage and underlying normal mucosa. Granulomatosis with polyangiitis and some infections like tuberculosis or Klebsiella cause ischemic necrosis of the overlying mucosa with resulting mucosal cicatrix formation overlying normal cartilage.

The primary pathology is within the lamina propria which overlies a normal cartilage. There is sub-mucosal granulation tissue with chronic inflammation. Airway obstruction results from fibrotic thickening of the lamina propria.

**Laryngotracheal Injuries**

**Depth of Injury**

- **Epithelium-only**
  - Healing with Resolution
  - The commonest cause of laryngotracheal stenosis is airway injury due to endotracheal intubation. Injury results from a combination of ischemic necrosis, acid reflux, and infection. When the extent to the submucosa but not the underlying cartilage, a similar pattern of injury, namely, lumen obstruction due to fibrotic lamina propria thickening overlying normal or near-normal cartilage.

- **Epithelium and Lamina Propria**
  - More severe, deeper laryngotracheal injuries involve inflammation and damage to the underlying cartilaginous framework of the airway. This pattern of injury results in both mucosal fibrosis and cartilage degeneration and necrosis. The stenosis results from a combination of mucosal cicatrix formation and collapse of the tracheal structure.

- **Epithelium, Lamina Propria and Cartilage**
  - Lambdoid deformity of the tracheal is a specific variant of transmural injury in which all layers of the anterior tracheal walls are removed and, in a proportion of cases, the normal wound-healing response causes tracheal wall collapse.
cartilage, it is likely to be a manifestation of abnormal wound-healing response rather than an iatrogenic complication per se. As such, a tracheal resection is more likely to cause suture-line restenosis and recurrence. These patients are initially managed with planned and staged endoscopic laryngotracheoplasty (see Figure 39–17) and are observed for “serial regression of lesion.” This is attempted using the standard “endoscopic laryngotracheoplasty” procedure, which consists of suspension laryngoscopy using a Dedo–Pilling laryngoscope, intralesional injection of 60 to 80 mg of methylprednisolone acetate, cruciate laser incision at 5 watts continuous CO2 laser setting, and stenosis dilation using a controlled radial expansion balloon. Mitomycin C is generally not used due to its association with significant post-procedure pain and risk of acute airway crusting. If serial regression of lesion does not occur, then augmentation tracheoplasty, which aims both to introduce new tissue and to break the circumferential scar, is considered. Use of antimetabolite therapy for highly selected cases of rapidly recurrent laryngotracheal stenosis is favorably but cautiously considered. The recent year has seen an increase in modulating fibrosis using existing novel agents and existing medications in this new indication, and as genomic and transcriptomic analyses provide increasingly patient-tailored targets for modulating recurrent scar formation, it is likely that the role of novel agents and existing agents in novel indications will increase.

**Treatment Intent and Shared Decision Making**

A prosthesis-free airway capable of meeting the ventilatory demands of the patient, accompanied by normal voice and swallowing functions, is the goal of laryngotracheal reconstructive surgery. This goal is achievable in the majority of patients, and indeed for many patients it can be achieved using only minimally invasive surgical techniques. The main considerations relate to the stenosis configuration, patients’ general fitness to undergo surgery, and, when laryngeal surgery is considered, safety of swallowing and risk of aspiration (see Figure 39–20).

In some cases, like the patient presented, this can be achieved with a single endoscopic procedure lasting less than an hour. In other cases, definitive airway treatment may require more extensive open cervico-mediastinal surgery, with the ultimate expectation being long-term restoration of the airway without the need for ongoing maintenance treatments. However, not all patients and not all stenoses lend themselves to curative treatment. For example, patients with post-radiation laryngeal stenosis and patients with complex stenoses involving both the larynx and trachea may not be reconstructable. Caution must be exercised over blinkered or narrow decision making—once the patient and the surgeon have resolved to keep out a tracheostomy at all cost, with each maintenance procedure, the decision to consider either a tracheostomy or a laryngectomy becomes progressively more difficult to revisit, and target fixation may cause major patient harm.

Repeated hospital admissions for stenosis dilation and airway maintenance can reinforce the sick role, and dysfunctional breathing as a consequence of fluctuations in a patient’s ability to breathe consistently can rapidly develop. This can lead to significant physical and psychological morbidity and repeated emergency hospital admissions. Moreover, living with a chronically marginal airway places the patient at ongoing risk of acute-on-chronic respiratory failure, particularly during episodes of intercurrent respiratory infections, and this may ultimately prove fatal. In addition, endoscopic treatment for intubation-related laryngotracheal stenosis is less cost-effective than open surgery, and this will be particularly the case when treatment turns to an open-ended approach of multiple and repeated hospital admissions for secondary dysfunctional breathing principally to avoid a stent or a tracheostomy.

Professor Isaac Eliachar described the technique of a permanent tubeless tracheostomy for non-reconstructable laryngotracheal stenoses. A further option is the use of a Silver Negus tracheostomy, which may reduce the morbidity associated with tracheostomy maintenance. A long-term tracheostomy and an intraluminal airway stent are doubtless associated with reductions in the patient’s quality of life and are to be avoided if possible. However, if deemed necessary, they must not be seen as treatment failures by either the patient or the surgeon and their use should be considered early for selected patients.

**Outcomes Assessment**

Assessing what management pathways provide the best outcomes is difficult in airway stenosis, as quantifiable measures do not always equate to patient-rated success.

**Pathology-Based Stratification.** The primary consideration in relation to outcomes and what constitutes treatment success in laryngotracheal stenosis is the nature of the underlying pathology. For example, idiopathic subglottic stenosis is a progressive fibromatosis with a high
expectation of recurrence following endoscopic treatment. A lambdoid tracheal deformity, on the other hand, is a structural stenosis caused by contracture of the anterior tracheal wall following removal of a tracheostomy in the context of otherwise normal wound healing. The expectation following endoscopic treatment is therefore full and permanent resolution of the condition. Stenosis recurrence following treatment of a lambdoid tracheal deformity therefore constitutes a true treatment failure, whereas stenosis recurrence after endoscopic treatment of idiopathic subglottic stenosis is expected. As such, even though both conditions cause laryngotracheal stenosis, the appropriate statistical methodology for studying these two conditions varies, being actuarial analysis for lambdoid tracheal deformity and intervention-free interval for idiopathic subglottic stenosis.

**Defining Treatment Success.** Decannulation rate has been used to measure treatment success. However, in contemporary practice, many stenosis patients do not receive tracheostomies, and decannulation rate alone, in the absence of information about functional outcomes, does not provide a picture of treatment success that is even remotely adequate. There is a pyramid of outcomes which, at the lowest level, involve impact of surgery on stenosis anatomy. The next set of outcomes are physiological and measure the impact of the presence and treatment of the stenosis on flow limitation and effort tolerance. Toward this, minimum clinically important difference (MCID) values for flow-volume loops have been calculated. As demonstrated in the case presented, the relationship between anatomy, physiology, and symptoms is not always cut and since ultimately intubation-related laryngotracheal stenosis is a benign condition, what matters is the extent to which treatment leads to improvements in the patient’s symptoms and quality of life, that is, efficacy of treatment. Durability of treatment defines how long these improvements last, and patient safety assesses both incidence of general and disease-specific complications, and duration of time a patient is exposed to risk. For example, a patient with idiopathic subglottic stenosis may be treated with serial endoscopic treatments, including serial in-office steroid injections. This treatment approach has good efficacy, short durability, and low risk in terms of exposure to major complications of open surgery. It does, however, have a higher risk of exposing the patient to dysfunctional breathing in the longer term and to proximal stenosis migration and glottic fixation. A cricotracheal resection has over 90% long-term cure rate but it does expose the patient to complications of major surgery and places the patient at risk of permanent voice morbidity. An endoscopic laryngotracheoplasty with biological inhibition may likewise achieve long-term disease remission, but exposes the patient to significant short-term risk of chronic cough and mucus. The shared decision on surgical approach is informed by a multitude of factors related to upfront and long-term risks, morbidities associated with different domains affected by the disease and its treatment, and the array of outcomes tools used to measure treatment success and compare different approaches need to comprehensively assess the different domains that are affected by the disease and its treatment. A holistic approach to outcomes assessment in laryngotracheal stenosis depends on the integration and communication of the different types and probabilities of expected risk and benefit in the context of a shared decision-making process with the patient, and while excellent research is now being performed in this area, a significant body of work remains to be done.

**Outcomes Assessment Instruments.** As discussed above, outcomes assessment in laryngotracheal stenosis is an area that is undergoing rapid evolution at this time. Table 39–2 provides the range of outcomes assessment instruments that are currently deployed at the Robert White Centre for Airway Voice and Swallowing.

**Prevention**

A full discussion of the risk factors for the occurrence of intubation-related laryngotracheal stenosis and the
strategies deployed to reduce its incidence are beyond the scope of this chapter. The use of high-volume low-pressure endotracheal cuffs and careful monitoring of endotracheal cuff pressures during critical illness are now near-universal practices within modern intensive care medicine. Use of tubes which contain a subglottic suction port to reduce tracheal exposure to digestive enzymes is also becoming standard of care. While traumatic intubation, particularly in the settings of cardiopulmonary resuscitation, cannot always be prevented, good attention to sedation technique and management of agitation may reduce ongoing trauma. Hemodynamic instability and in particular prolonged hypotension reduce tracheal perfusion pressure and need to be carefully managed. Paying close attention to endotracheal tube selection and in particular taking account of patient height as well as sex in choosing the most appropriate tube size may be particularly helpful in reducing inter-arytenoid trauma.

A particularly pertinent consideration for the airway surgeon is the timing of switch from endotracheal tube to tracheostomy. A tracheal stenosis, if it develops, can be successfully managed in the overwhelming majority of cases and as demonstrated in this case and previously published, lamblid tracheal deformity which is a specific complication of tracheostomy placement is particularly amenable to treatment. By contrast, a mature interarytenoid scar causing bilateral vocal fold immobility is one of the most complex airway injuries to manage. While development of endoscopic and open laryngeal remobilization procedures is beginning to change the poor outcome of this injury, prevention remains far more desirable. Interarytenoid scarring exclusively occurs in translaryngeal intubation. Interestingly, patient height is an independent risk-factor for the development of this injury, and this reinforces the notion of a height-inappropriate, abnormally large endotracheal tube causing inter-arytenoid ulceration and stenosis formation. There is an expanding literature on the relative merits of early vs late tracheostomy within the broader intensive care context, but from an airway surgical perspective, an early tracheostomy, in the sole context of preventing a laryngeal injury, even at the expense of a higher risk of tracheal injury, is a desirable strategy.

**Screening for Intubation-Related Laryngotracheal Stenosis**

Intubation-related laryngotracheal stenosis fulfills the World Health Organization criteria (Table 39–3) as a condition that should be screened for. Its identification and treatment is wholly consistent with guidelines issued by the National Institute of Health and Care Excellence for rehabilitating adults following critical illness (CG83). An important factor to consider, however, is the numbers needed to treat. At or shortly

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**Table 39–2. Outcomes Assessment Instruments for Laryngotracheal Stenosis**

<table>
<thead>
<tr>
<th>When Assessed</th>
<th>Instruments Used</th>
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<tbody>
<tr>
<td><strong>Anatomy</strong></td>
<td></td>
</tr>
<tr>
<td>Distance from glottis</td>
<td>Every visit</td>
</tr>
<tr>
<td>Cross-section of the stenosis</td>
<td>Every visit</td>
</tr>
<tr>
<td><strong>Physiology</strong></td>
<td></td>
</tr>
<tr>
<td>Voice</td>
<td>Every visit</td>
</tr>
<tr>
<td>Flow</td>
<td>Every visit</td>
</tr>
<tr>
<td>Effort</td>
<td>Major treatments</td>
</tr>
<tr>
<td><strong>Symptoms</strong></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Every visit</td>
</tr>
<tr>
<td></td>
<td>Major treatments</td>
</tr>
<tr>
<td><strong>Quality of life</strong></td>
<td>Major treatments</td>
</tr>
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<td>EQ5D</td>
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following extubation, almost half of all patients have evidence of significant laryngotracheal injury, and yet fewer than 10% of these patients go on to develop significant long-term laryngotracheal stenosis. As such, acute intervention for every laryngotracheal injury would lead to significant overtreatment.

One possible and pragmatic approach is to perform at least a laryngoscopy on every patient within 48 hours of extubation (Figure 39–23). This can be done by the critical care team in partnership with the laryngologist. All patients with post-extubation stridor should be further assessed and if necessary treated. Among patients who are minimally symptomatic, patients with laryngeal injury, and in particular patients with inter-arytenoid ulceration should be acutely treated to reduce the likelihood of progression to bilateral vocal fold immobility due to inter-arytenoid scarring. Patients with evidence of significant but asymptomatic tracheal injury should be followed up with office tracheoscopy and flow-volume loops and only treated if they develop airway symptoms. All critical care follow-up clinics should have access to flow-volume loop testing and ideally to office laryngotracheoscopy. There is a clear progression in the natural history of intubation-related laryngotracheal stenosis from readily treatable acute fibro-inflammatory lesion

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**Table 39–3. Wilson and Junger Criteria for Screening Programs**

- The condition sought should be an important health problem.
- There should be an accepted treatment for patients with recognized disease.
- Facilities for diagnosis and treatment should be available.
- There should be a recognizable latent or early symptomatic stage.
- There should be a suitable test or examination.
- The test should be acceptable to the population.
- The natural history of the condition, including development from latent to declared disease, should be adequately understood.
- There should be an agreed policy on whom to treat as patients.
- The cost of case-finding (including diagnosis and treatment of patients diagnosed) should be economically balanced in relation to possible expenditure on medical care as a whole.
- Case-finding should be a continuing process and not a "once and for all" project.