External Stabilization of Long-Segment Tracheobronchomalacia Guided by Intraoperative Bronchoscopy

Siegfried Hagl, MD, Heinz Jakob, MD, Christian Sebening, MD, Peter van Bodegom, MD, Klaus Schmidt, MD, Eugen Zilow, MD, Franz Fleischer, MD, Herbert Ulmer, MD

Departments of Cardiac Surgery, Pediatric Cardiology, and Anesthesiology, University Hospital and Clinic for Thoracic Medicine, Heidelberg, Germany

Methods. From July 1992 to April 1995, 7 children (age range, 4 months to 4 years; mean age, 19 months) and 1 adult (age, 46 years) were operated on for severe respiratory insufficiency. In 4 cases of congenital tracheal instability, 2 children had associated type IIIb esophageal atresia. Both children with esophageal atresia had previous operations (two and three times, respectively): 1 child had aortopexy and division of a patent ductus arteriosus and another child had distal tracheal resection elsewhere, both without relief of malacia. All children were intubated and ventilated since birth for 11 to 15 months. Secondary tracheobronchomalacia due to vascular compression was seen in 4 patients caused by double aortic arch (n = 2) and persisting ligamentum arteriosum after previous ligation of a patent ductus arteriosus (n = 2), with 1 child ventilated thereafter for 5 months. Operation was performed with the aid of extracorporeal circulation in all patients but 1, and consisted of transection of vascular rings and persistent ligamentum Botalli (n = 5), closure of multiple ventricular septal defects (n = 1) and extensive mobilization of the tracheobronchial tree as well as the great arteries. External stabilization of the severely dysplastic distal trachea (n = 6) or left main bronchus (n = 2) was achieved by suspending the malacic segment within an oversized and longitudinally opened ring-reinforced polytetrafluoroethylene prosthesis. Multiple plegeted sutures were placed extramucosally to the dysplastic tracheal wall and the dyskinetic pars.
membranacea, as well as to the polytetrafluoroethylene prosthesis in a radial orientation. Guided by simultaneous video-assisted bronchoscopy, reexpansion of the collapsed segments was achieved by gentle traction on the sutures while tying.

Results. Stenosis-free tracheobronchial reexpansion was achieved in all patients, as seen on repeated bronchoscopies during hospitalization and thereafter. All patients were extubated within 1 to 12 days after the operation. There was one late death, unrelated to the procedure, in a 3 1-month-old child 20 months after the operation. All other patients are free of stridor and in excellent clinical condition 21 to 54 months (mean, 38 months) thereafter.

Conclusions. The presented method of bronchoscopically guided external tracheobronchial suspension within a ring-reinforced polytetrafluoroethylene prosthesis immediately relieves severe malacia of the trachea or main bronchi in infants as well as adults without necessitating resection. Midterm preliminary data suggest that growth potential of the affected segment exists within the oversized polytetrafluoroethylene prosthesis.

Introduction

See also page 1421.

Severe airway obstruction at different levels of the system may result from congenital or acquired instability of the tracheobronchial wall. Congenital malformations such as hypoplasia or dysplasia of the tracheobronchial wall elements, absence or immaturity of the cartilaginous frame, or an excessively wide and flaccid pars membranacea with hypotonia of the myoelastic elements are important pathoanatomic substrates for severe dysfunction of malacic airway segments [1-3]. Secondary instability of tracheobronchial segments may result from anomalous vascular anatomy with or without associated tracheal abnormalities. Abnormal topography of the vascular or the airway system, vascular rings, or pseudoring formation can lead to deformation of the cartilaginous rings and can produce tracheal or bronchial malacia by direct pulse pressure transmission from adjacent arteries to the airway wall, causing destruction of the wall [4, 5].

Increased collapsibility and the presence of tracheomalacia is often associated with esophageal atresia and tracheoesophageal fistula. Abnormal mechanical forces directly transmitted from an adjacent structure like an esophageal pouch may play a major role in the pathogenesis of this disease [2, 6, 7]. Chronic inflammation and damage after tracheostomy and long-term ventilation are additive factors.

Both congenital and acquired severe long-segment tracheomalacia and bronchomalacia continue to be a challenge in thoracic surgery. The degree of compression and dysfunction of the airway segment determines the severity of symptoms. Recurrent retention pneumonias and atelectasis may lead to life-threatening respiratory failure requiring long-term mechanical ventilation.
Even successful surgical decompression of the airway system in cases with vascular anomalies often does not correct severe obstruction and may indicate repeated tracheoplastastic and other reconstructive procedures [8, 9]. Besides those procedures, the most widely applied method to treat severe tracheomalacia still is aortopexy, often followed by tracheostomy with long stenting tubes, which are left in situ for the first several years of life, thus leading to physical and psychosocial problems for these children and necessitating alternative methods of tracheal stabilization and reconstruction [6, 10, 11].

The present article aims to describe a technique for repair of long-segment airway instability and reports the midterm clinical outcome of 8 patients undergoing this type of operation.

Patients and Methods

Between July 1992 and April 1995 a total of 18 patients were operated on for relief of major airway obstruction, both isolated and associated with congenital heart defects, in our department. In 14 cases, vascular compression syndromes (Table 1) and in 4 cases congenital airway instability were the underlying disease.

View this table: Table 1. Vascular Tracheobronchial Compression Syndromes, 3/92 to 1/97 (14 Patients)

In a subgroup of 8 patients (age range at operation, 8 months to 46 years) severe long-segment instability of the airway system was present or persisted after complete surgical decompression. Four patients suffered from congenital or primary instability of the trachea. Two of these patients have had corrective operations for esophageal atresia, with successive repair of a laryngeal cleft in 1 case. The other infant underwent segmental tracheal resection elsewhere. The second group of 4 patients presented with tracheomalacia (n = 2) and bronchomalacia (n = 2) secondary to vascular compression syndromes. To illustrate the wide variability of the pathology and the clinical course, the case histories of these patients are summarized in Tables 2 and 3.

View this table: Table 2. Patients With Congenital Intrathoracic Tracheal Wall Instability (7/92-1/97)
Tracheobronchial malacia was defined as almost complete collapse (more than 70%) of the trachea and main bronchus for at least half of the intrathoracic tracheal length. These findings had to be consistent in computed tomographic (CT) scanning of the chest as well as in bronchoscopy. All CT scans (Siemens Somatom Plus, Erlangen, Germany) were performed under deep sedation in apnea with the endotracheal tube temporarily pulled back to the cricoid to avoid stenting of the trachea during scanning. All bronchoscopies were performed with rigid bronchoscopes, pulled back as far as possible to allow spontaneous breathing under deep sedation. Inspection of the unstented tracheobronchial system was done with a 5-degree angled Hopkins optic (Wolf, Tuttlingen, Germany).

To quantify the degree of tracheal collapse and reexpansion before and after the procedure, we used the method of Griscom and associates [L2, 13], which shows that anteroposterior diameter and transverse diameter of the trachea are strongly correlated to body height. In 4 patients with tracheal collapse, measurements of the narrowest point before and after the correction could be performed using Griscom and associates’ formula:

\[ bH (tracheal dimension in \text{ cm}) = a + 1.0 \times m \times \text{height in \text{ cm}} \]

where \(a\) is the intercept and \(b\) is the slope of a resulting straight line on a log-log graph using base e.

Surgical Technique
A standard median sternotomy was performed. The right thymus lobe was resected and the pericardium opened. The aorta, the aortic arch, the supraaortic vessels, and the pulmonary artery were dissected free. The ligamentum arteriosum was divided. By anterior leftward retraction of the aorta the entire trachea was exposed from the neck to the bifurcation. The topographic relationship of vascular structures and the airway system was examined. This maneuver was supported and guided by video-assisted bronchoscopy localizing the site and extent of narrowing. If present, vascular abnormalities were corrected during standard extracorporeal circulation; the aim was to separate vascular and airway structures completely. At this step it was important to withdraw or better remove the endotracheal tube to unmask "stenting effects." If severe airway instability persisted despite complete decompression, the trachea, the bifurcation, and the main bronchi were dissected free. Care was taken not to strip the tracheal or bronchial wall from its long-axis vascular supply. A segment of a ring-reinforced polytetrafluoroethylene (PTFE)-prosthesis (20 mm in diameter for tracheal and 14 ram for main bronchial stabilization) was longitudinally cut open and placed around the unstable airway segment. The pars membranacea was then fixed by two or three rows of single sutures at the posterior aspect of the prosthesis (Fig 1E3). Multiple rows of Teflon-pledgeted braided polyester sutures were then placed extramucosally in the tracheal or bronchial wall and passed through the PTFE prosthesis in a radial fashion (Fig 2fl). To ensure sufficient stability of the malacic portions within the prosthesis the suture rows had to be placed at least 1 cm beyond the transition zones to healthy tracheal tissues on all sides.
After closure of the prosthetic tube with a continuous suture line, the single sutures were gently tied over the prosthesis. Adapting the tension on each suture allowed not only reexpansion of the malacic segment but restoration of normal geometry of the unstable segment to be easily achieved with guidance from the videoendoscopic view of the suspended area. The free suspension of the affected tracheal, carinal, or bronchial portion within an oversized framework aimed to give sufficient space to allow growth with time. To prevent graft erosion into the esophagus or the great arteries a PTFE membrane was placed between the different structures.

The procedure preferably was performed during normothermic cardiopulmonary bypass, which enabled extubation and inspection of the extent of the collapsibility of the major airway system during the operation, but was also done without extracorporeal circulation in cases where isolated tracheomalacia was present.

Select Case Histories

PATIENT 2.

A 15-month-old boy was born prematurely (2,500 g) and operated on on his first day of life for
esophageal atresia (type IIIb) with closure of his tracheoesophageal fistula in another center.

Postoperatively, the child could be extubated but had to be reintubated several hours later for respiratory decompensation due to massive secretion retention. Bronchoscopy revealed segmental intrathoracic tracheomalacia, for which the patient underwent tracheal segmental resection at 5 months of age elsewhere. Several attempts to extubate the child thereafter failed and were accompanied by massive secretion retentions and episodes of respiratory arrest necessitating emergency reintubation and cardiopulmonary resuscitation, from which the patient recovered without sequelae. For the following 10 months the child remained intubated and mechanically ventilated via a tracheostomy.

Bronchoscopy documented persisting tracheomalacia with a dyskinetic pars membranacea 3 cm in length, beginning 1 cm above and ending 2 cm below the end-to-end anastomosis down to the carina. The anastomosis was slightly covered with granulation tissue, which could be passed with a 6-mm bronchoscope. Total remaining length of the trachea after previous resection was 8 cm. The endotracheal tube had to be used to internally stent the malacic distal portion of the trachea.

The patient was transferred to our center at 15 months of age and underwent external tracheal suspension over a length of 3.5 cm through a midsternal approach with the use of cardiopulmonary bypass. The postoperative course was uneventful, and the child was extubated under endoscopic control on the 10th day. One month after the tracheal suspension, the previous tracheostomy was electively closed by a plastic procedure using a muscular flap with immediate postoperative extubation. A few days later the patient was discharged home and currently is in excellent condition and leading a normal life.

PATIENT 5.
A 4-year-old girl had previously undergone ligation of a patent ductus arteriosus at the age of 2 years elsewhere. Over the past 2 years the girl suffered from recurrent and progressive respiratory distress requiring intermittent mechanical ventilation. Episodes of total left lung atelectasis and recurrent left-sided septic retention pneumonias were observed. Bronchoscopy revealed a severe, pulsating long-segment stenosis of the left main bronchus over a distance of 3 cm. At the age of 4 years, the patient underwent reoperation at our center. After midline sternotomy, the left main bronchus was found to be severely compressed between the distal aortic arch posteriorly and the slightly distorted left pulmonary artery anteriorly. The upper part of the "ferrule" was formed by the scarry remnant of the ligamentum arteriosum. Due to extreme adhesions in this area, the operation was performed on partial cardiopulmonary bypass. After transection of the firm remnant of the ligamentum arteriosum and subsequent extensive mobilization of the aortic arch, the left main bronchus, and the pulmonary artery, the freed left main bronchus showed marked wall instability and malacic portions over a distance of roughly 3 cm without tendency to reexpansion. Thus the left main bronchus was suspended within a 14-mm ring-reinforced PTFE prosthesis according to the technique outlined above (Fig 2CS).

The postoperative course was uneventful, and extubation was possible the morning after the operation. Follow-up now extends to 54 months with the girl in excellent condition. Follow-up CT
scan and bronchoscopy were done in June 1997.

PATIENT 6.
A 46-year-old woman had suffered from respiratory obstructive disease with stridor, intermittent severe respiratory distress attacks, and mild dysphagia for as long as 40 years, which was misdiagnosed as "asthma" for all the years.

Because of increasing intensity and frequency of severe dyspneic attacks, the patient underwent tracheoscopy, CT, and magnetic resonance scans, as well as angiographic examination, at the age of 46 years. These studies showed a right aortic, retroesophageal arch in relation to tracheal compression. Endoscopy revealed a pulsating, marked compression stenosis at the left posterolateral aspect of the distal trachea (more than 5.5 cm in length), which was malacic; the lumen was reduced by almost 70%.

At operation, performed through a midline sternotomy with cardiopulmonary bypass, the ascending aorta and the right-sided aortic arch followed a sharply curved anteroposterior course, then turned to the left behind the esophagus. The separate origins of the supraaortic arteries showed a normal branching pattern. The left subclavian artery arose with a typical Kommerel's diverticulum and coursed to the left. The ligamentum arteriosum was left-sided and showed a diverticuloid deformity on the aortic wall and was under apparent tension. The trachea at its distal third was compressed by the abnormally coursing aortic arch, the left pulmonary artery and the ligamentum Botalli forming a vascular sling complex.

After transection of the ligamentum Botalli the ligated stumps separated by 3 cm, but the trachea was not entirely freed from its compression because the abnormally coursing aortic arch system still exerted a residual "traction." In addition, the tracheal segment, severely compressed over a length of 5.5 cm, was clearly malacic and did not reexpand after the previous decompression maneuver. Thus hypothermic circulatory arrest at 19°C nasopharyngeal temperature was initiated and the aortic arch was transected just proximal to the left subclavian artery, and doubly oversewn with a 4-0 continuous polypropylene suture. A 5-cm-long, collagen-impregnated, 24-mm vascular prosthesis was inserted side-to-end between ascending aorta and the anteriorly translocated distal aortic arch segment.

Despite effective decompression of the trachea a 3.5-cm unstable segment persisted due to lack of several cartilaginous rings on the right anterolateral and left posterolateral aspect. This tracheal segment was then suspended within a 20-mm PTFE prosthesis (Figs 2A, 2B). This allowed full tracheal reexpansion, documented by intraoperative tracheoscopy.

The postoperative course was uneventful besides a left recurrent nerve palsy, which persisted over time. Extubation followed at postoperative day 1 under endoscopic control.

Mild dysphagia occurred, probably corresponding to a slight residual compression of the esophagus at the "crossing site" of the lower descending aorta (approximately 35 cm distal to the dental line). This symptom slightly regressed under conservative treatment.
Forty-nine months later (December 1996), actual tracheoscopy and CT scan examination demonstrated a widely patent airway system. The patient is in good condition and free of stridor but still plagued by a mild hoarseness and dysphagia.

**Results**

There was no operative mortality and no procedure-related morbidity. One patient (patient 3) with a very complex cardiac malformation and a severe therapy-resistant dystrophy died 20 months after successful restoration of airway stability and several palliative procedures of acute aspiration and cardiac failure. At autopsy the trachea was widely patent.

All patients were extubated within 12 days after the operation (see Tables 2, 3EIII). The midterm results after a mean follow-up of 38 months (range, 21 to 54 months) of the entire group are encouraging. All patients are in excellent condition. There is no evidence of obstruction or dysfunction of the airway system at any level.

Follow-up CT scans could be obtained in 4 of 8 patients after an interval of 2 to 48 months. In 1 patient (patient 1) a repeat CT scan 18 months after the first postoperative CT scan (3 months) became available (Fig 3s). In this child, growth of the trachea within this 1.5-year interval could be demonstrated. The narrowest anteroposterior diameter increased from 6.6 mm to 8.6 mm, and the transverse diameter from 10.7 to 11.6 mm. In all 4 patients the mean preoperative narrowest tracheal anteroposterior diameter increased from 1.8 mm (range, 1 to 2.2 mm) to 11 mm (range, 8.6 to 14.4 mm), and the narrowest transverse diameter from a mean of 4.5 mm (range, 2 to 8 mm) to 13 mm (range, 9 to 18.5 mm) (Table 4®).

*Fig 3. (Patient 1.) (A) Serial thoracic computed tomographic scans from the cricoid (I) to the tracheal bifurcation (V) demonstrating severe long-segment tracheal stenosis. (B) Follow-up computed tomographic scans in the same patient 3 months postoperatively, demonstrating a widely patent distal trachea.*
Comment

Tracheobronchial malacia may lead to life-threatening dysfunction of the airway system at different levels, especially in infants. Although the underlying pathology and pathogenesis seems to be clearly understood for acquired tracheobronchial malacia secondary to vascular compression or other mechanical forces, the etiology of congenital airway wall instability is not. Early diagnosis and relief of tracheobronchial compression and early restabilization are essential to prevent severe structural damage of the airway system by mechanical and chronic inflammatory mechanisms, besides the devastating physical and psychosocial stress caused to these patients and their parents.

Controversy still remains about the optimal treatment, ranging from conservative therapy to reconstructive procedures including resection, plasty, or graft interposition techniques [14–18]. Internal stenting by prolonged intubation after tracheostomy still is the most popular, but child-unfriendly procedure [6]. Dramatic immediate effects of modern endotracheal stenting with self-expandable metal airway-stents have been reported [19]. However, these endotracheal stents do cause constant pressure on the mucosa as well as the weak cartilage, which leads to at least chronic inflammation and resolution of cartilage [20]. Furthermore, there is a high risk of granuloma build up, perforation, and dislocation, especially in the growing child. All these children do need frequent bronchoscopies and repeated exchange of expensive stents, without the likelihood of a definite solution of their problem.

As a surgical option, a variety of procedures have been used to reconstruct or restabilize nonexpanding or malacic tracheobronchial segments in the past [9]. Long-segment resection and an end-to-end anastomosis can be performed in infants despite diminutive structures and the lower mechanical resistance of the infant trachea and the very reactive mucosa [14]. However, persistence of a dysfunctional pars membranacea, the tendency of recurrent stenosis at the anastomotic site due to tension, and reduced growth potential do cause considerable morbidity and often require repeated surgical interventions using these techniques. Tracheoplasty procedures as described by Idriss and associates [21] or Tsang and colleagues’ [22] sliding plasty technique do focus on noncollapsible tracheal stenoses and do not apply to the specific pathology of dysplasia of all circumferential tracheal structures.

Surgical external stabilization by aortopexy or aortotruncopexy is a well-established technique for short-segment malacia, especially in infants after tracheoesophageal fistula repair [11, 23], but not in long-segment tracheobronchial instability according to our experience. Late results of such splinting procedures in pediatric patients with this type of pathology are sparse. One reason for
failure could be that preservation of the tracheal cylindrical structure as a whole was not achieved.

Various forms of autografts and xenografts have been used to replace tracheal segments, both experimentally and clinically [8, 24-26]. However, the results are controversial and unsatisfactory, at least in infants and small children.

Recently, Elliot and coworkers [27] presented impressive results using homograft implants in 5 desperately ill infants, with an 80% success rate. However, the authors themselves caution to use this technique only in cases where alternative modes of treatment have failed. Silicone intraluminal stenting for 12 weeks, granulation tissue build-up, immunologic side effects including infection and anastomotic dehiscence, and the loss of viable cartilage producing a fibrotic, stiff tube have to be kept in mind.

Our surgical technique of resuspension of all affected tracheobronchial wall structures within an oversized PTFE prosthesis guided by videoendoscopic on-line control immediately rebuilds a normal tracheal geometry without affecting the mucosa. We first applied the technique in 1992 in a 4-year-old patient (patient 7 in Table 3S) with severe stenosis of the left main bronchus due to persisting external compression caused by a scarred ligamentum arteriosum after ligation of a patent ductus arteriosus 2 years before. In this patient as well as in all others, restabilization of the malacic segment was achieved using this technique (see Fig 2cIII). The very reactive mucosa remained untouched and therefore no granulation tissue build-up was observed in any patient. This seems especially important in the proximity of the bifurcation. The natural barrier between the airways and other mediastinal structures is respected. The risk of infection should be lower compared with resection and plasty techniques. In the entire group under study there was no infection documented.

This probably is a main reason that all our patients could be successfully weaned from the ventilator within 1 to 12 days even after periods of mechanical ventilation up to 15 months or severe respiratory distress for 40 years misdiagnosed as asthma. In addition, all patients but the 1 child who died of cardiac failure due to a complex cardiac malformation 20 months after successful tracheal suspension do lead a completely normal life without rehospitalizations other than for routine follow-up. Overcorrection of the tracheal lumen could be another reason for the unanimously smooth postoperative course in all patients, reducing airway resistance significantly and facilitating conversion to spontaneous breathing in children with weakened respiratory muscles after long-term intubation. Of concern certainly was the hypothetical prosthetic erosion of adjacent vascular structures or the esophagus. However, wrapping of those structures with a thin PTFE membrane was protective, as demonstrated by midterm follow-up CT scans that did not show vascular or esophageal compromise in 4 patients to date.

The second major concern was the preservation of growth potential of the freely suspended airway segments within the oversized PTFE prosthesis (minimum diameter,' 20 mm). To date, from this limited experience and midterm follow-up there is no evidence of restrictive or obstructive disease. Overcorrection to an anteroposterior diameter of a mean of 11 mm and a transverse mean diameter of 13 mm seems to warrant a sufficient lumen even for adulthood, where symptomatic stridor is to
be expected with diameters less than 6 mm. Whether tracheal growth can be generally anticipated as demonstrated in patient 1, with an increase in anteroposterior and transverse diameters of 2.2 and 0.9 mm within a 1.5-year observation period to current diameters of 8.6 and 11.6 mm, respectively, remains to be seen. However, this appears to be a substantial advantage compared with circumferential "onlay techniques" as advocated experimentally by Shaha and coworkers [28], in which no growth could be expected.

To qualitatively assess the strength of the fibrous tissue layer between the tracheal outer surface and the surrounding prosthesis, probably imaging techniques other than the CT scans that we used will become necessary. To date, no signs of nutritive disorders or degeneration of wall structures could be demonstrated during the follow-up examinations. At all follow-up broncoscopies, an intact mucosa and normal cartilaginous ring structures within the suspended areas were observed, indicating the restoration of tracheobronchial wall stability. We hypothesize that this process is comparable with observations in infants with localized tracheal instability due to innominate artery compression, where the initially collapsing tracheal segment was observed to regain wall stability within half a year after aortopexy performed early in life.

We conclude that this technique of circumferential free suspension of long-segment tracheobronchial malacia within an oversized PTFE prosthesis is a promising tool in treating different and complex tracheobronchial pathologies from infancy to adulthood.

Footnotes


Address reprint requests to Dr Hagl, Department of Cardiac Surgery, University Hospital, INF 110, 69120 Heidelberg, Germany

References