

Primary tracheal tumors: experience with 14 resected patients

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Abstract

Objective: Primary tracheal tumors are rare. Management includes interventional endoscopy, surgery and radiotherapy. **Methods:** Between 1987 and 1996, 14 patients treated by resection and reconstruction of the trachea and bifurcation for primary tracheal tumors were retrospectively analyzed. **Results:** The most common histological finding was adenoid cystic carcinoma ($n = 7$), followed by a squamous cell carcinoma ($n = 2$), a mucoepidermoid carcinoma ($n = 2$), a carcinoid tumor ($n = 1$) and two benign tumors (xanthogranuloma, pleomorphic adenoma). Various reconstruction techniques were used and one prosthesis was implanted. Eight of the patients required preoperative Nd-YAG laser recanalisation. Six were treated by postoperative external beam radiotherapy, in three cases combined with endoluminal brachytherapy. Two major postoperative wound-healing impairment at the anastomosis occurred. Four minor wound-healing disorders were successfully treated by interventional endoscopy. Two patients died postoperatively with mediastinitis respectively with bilateral pneumonia. A local recurrence was observed in only two cases. At the last follow-up in January 1998, nine patients were still alive. We observed five long-term survivors (>6 years) with an adenoid cystic carcinoma or mucoepidermoid carcinoma. **Conclusions:** Extensive segmental resection of the trachea is the treatment of choice for primary malignant and occasionally for benign tracheal tumors. Interventional endoscopy is a part of modern tracheal surgery. © 2001 Elsevier Science B.V. All rights reserved.

Keywords: Trachea; Tracheal resection; Tracheal tumors

1. Introduction

Standards based on experimental and clinical studies on segmental tracheal resection have been established in the last four decades [1]. Surgical techniques were developed for both adult and pediatric patients [2]. Today we consider tracheal surgery to be indicated in two cases: (1) tumorous tracheal stenoses mainly caused by a locally advanced lung cancer with infiltration of the bifurcation or by primary and secondary tracheal tumors; (2) congenital tracheal stenoses in children or those acquired after trauma or long-term intubation. Modern tracheal surgery includes not only resection and reconstruction techniques but also interventional bronchoscopy in the diagnostics, preoperative preparation, emergency treatment, postoperative control of results and management of postoperative complications.

Primary tracheal tumors are rare. There is a 1:100 ratio of tracheal to bronchial tumors in autopsy material. They represent only 2% of upper airway tumors [3]. The majority

of tracheal tumors in adults are malignant but benign in 90% of children [4]. Tumors can grow in every parietal layer of the trachea and thus greatly vary histologically. The adenoid cystic carcinoma is the most common type in adults, followed by squamous cell carcinomas and carcinoids [4]. 40–50% of the tumors are located in the lower trachea and bifurcation, 30–35% in the upper and only 10–15% in the middle trachea [5]. Resecting the lower trachea or bifurcation presents a technical challenge for surgeons and anesthesiologists [6,7].

Secondary tracheal involvement due to infiltrating tumors of the larynx, thyroid, esophagus, and lungs is more common than primary tumors. Even mediastinal lymph node enlargement caused by metastasizing may lead to considerable tracheal compression and life-threatening suffocation. Distant mucosal metastases are very rare and have been specifically observed in connection with malignant melanomas. Lung cancer, which have spread to the distal trachea as T4 tumors, are treated according to the principles of lung cancer surgery and are the most common indication for resection of the distal trachea [8].

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2. Materials and methods

We retrospectively examined 14 patients (six men, eight women) who underwent tracheal tumor resection from 1987 to 1996 in our Department of Thoracic Surgery by the senior author. The median age was 55.5 years (range 34–72). We included patients with benign and malignant primary tumors located between the cricoid cartilage and the tracheal bifurcation. Resection techniques, complications and long-terms results were of primary importance in this study. A total of 251 tracheal resections were performed in the same period, the majority for lung cancer infiltrating the distal trachea or benign stenoses. All patients were postoperatively submitted to bronchoscopy every 3 months during the first year and every 6 months or whenever necessary thereafter. The last follow-up examination was in February, 1998. The follow-up is complete (median 33.4 months, range 5 days–120 months days).

2.1. Diagnosis

All patients were symptomatic. The most common symptoms were coughing ($n = 12$), dyspnea ($n = 8$), stridor ($n = 8$) and hemoptysis ($n = 4$), whereas changes in voice quality ($n = 2$) and chest pressure ($n = 1$) were rather rare. Some patients had two or more symptoms. The median course from the onset of symptoms to diagnosis was 18 months (range 0–120 months). All patients were examined with chest roentgenography, rigid bronchoscopy and computed tomograms as well as function tests. A median stenosis grade of 50% (range 0–90%) was observed in preoperative rigid bronchoscopy. The majority of tumors were located in the middle and lower tracheal section. The longitudinal spread of the tumor was a median 41 mm (range 15–80 mm). All patients underwent a preoperative chest CT and a lung function test.

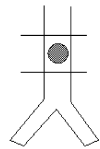
2.2. Therapy

Preoperative endoscopic intervention was necessary in eight patients due to bleeding or subtotal obstruction (eight laser ablations (Nd-YAG-Laser), one combined with forceps ablation). One female patient was submitted to external beam irradiation with 60 Gy before surgery.

A combined cervico-mediastinal access was used in eight patients (upper trachea $n = 1$, middle trachea $n = 5$, lower trachea $n = 1$, bifurcation $n = 1$). Sternotomy alone was sufficient in three patients (lower trachea $n = 1$, bifurcation $n = 2$). A right posterolateral thoracotomy was done in three patients (bifurcation). The extent of the tracheal resection was a median of 55 mm (40–80 mm). The resections of the trachea and the bifurcation are shown in Fig. 1. In five patients, an end-to-end anastomosis was carried out for reconstruction. As shown in Figs. 2 and 3, in seven patients (four bifurcation resections, three tracheal segment resections), the left bronchus was implanted into the bronchus intermedius for tension-free anastomosis. In one case (Fig.

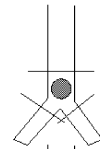
Segmental resection

7



Bifurcation

6



Sleeve pneumonectomy

1

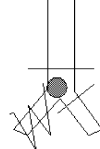


Fig. 1. Resection of the trachea and bifurcation in primary tracheal tumors ($n = 14$).

4), the carina was reconstructed after isolated bifurcation resection. Implantation of a Neville bifurcation prosthesis was required in one case with a resection length of 80 mm. In order to prevent disturbances of blood supply, posterior dissection of the trachea is avoided. The anastomosis is performed with the use of interrupted sutures, monofilament 3-0 or 2-0 with knots tied on the outside. We use high-frequency jet ventilation during anastomosis. Anastomosis is covered with a flap of vascularized tissue only to separate it from innominate artery.

2.3. Histological findings and radicality

All operative specimens were checked for the absence of tumor tissue in the resection margin by frozen section under surgery. The final histological examination revealed a malignant tumor in 12 cases (seven adenoid cystic carcinomas, two squamous cell carcinomas, two mucoepidermoid carcinomas and one carcinoid) and a benign tumor in two (one pleomorphic adenoma and one xanthogranuloma). The resection was macroscopically and microscopically radical (R0) in nine patients. Microscopic residual tumor tissue at the resection margin (R1) was found in four patients and macroscopic (R2) in one. Mediastinal lymph node metastatic spread was also detected in four patients (adenoid cystic carcinoma $n = 3$, carcinoid $n = 1$).

3. Results

3.1. Postoperative course and adjuvant treatment

The patients were extubated after 7 h (median), range 3–120 h following surgery. Three patients had a complication-free course. Eleven patients experienced complications which are listed in Table 1. Four patients with minor wound healing disorders were successfully treated by endoscopic procedures (tracheal stent implantation, laser abla-

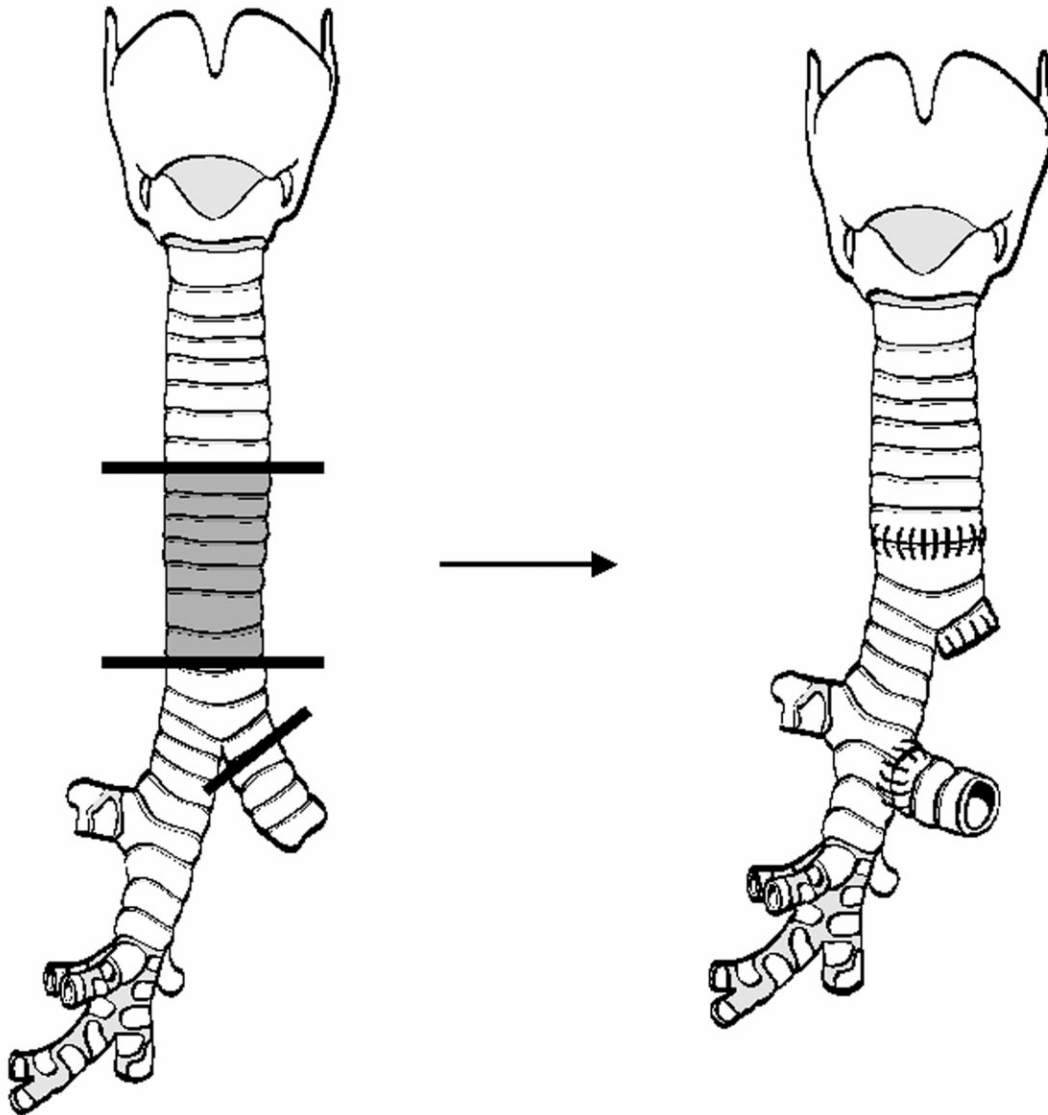


Fig. 2. Reconstruction in extended resection of the trachea: extended segmental resection of the middle part of the trachea. The right lung is mobilized and the distal trachea is elevated to reach the proximal part. The left main bronchus is divided and anastomosed to the bronchus intermedius ($n = 3$).

tion of granulation, bouginage). Another patient following isolated bifurcation surgery developed a cicatricial stenosis after wound healing disorder that required a right superior sleeve lobectomy 8 weeks later. Two patients died of surgical sequelae while hospitalized (mediastinitis following implantation of a tracheal prosthesis in the presence of anastomotic insufficiency; bilateral pneumonia with secretion retention).

Postoperative external beam radiotherapy was performed in six patients, in three cases combined with endoluminal high-dose-rate (HDR) brachytherapy.

3.2. Long-term results

At the last follow-up (28 February, 1998), nine patients

were still alive and recurrence-free: three adenoid cystic carcinomas (R0), two mucoepidermoid carcinomas (R0), one squamous cell carcinoma (R0), one carcinoid (R1) and two benign tumors. Median survival of patients with adenoid cystic carcinomas ($n = 7$) was 25 months (5 days to 8 years). Three of these patients achieved long-term survival: 5.6 years without local recurrence and good function, 8 years without local recurrence and good function and 7.1 years with lung metastases and local recurrence which appeared in the 4th postoperative year and was treated by stent implantation. Moreover, two patients are alive recurrence-free with a mucoepidermoid carcinoma and good functional results 9.4 and 9.9 years, respectively after surgery.

Two patients experienced a local recurrence (one patient

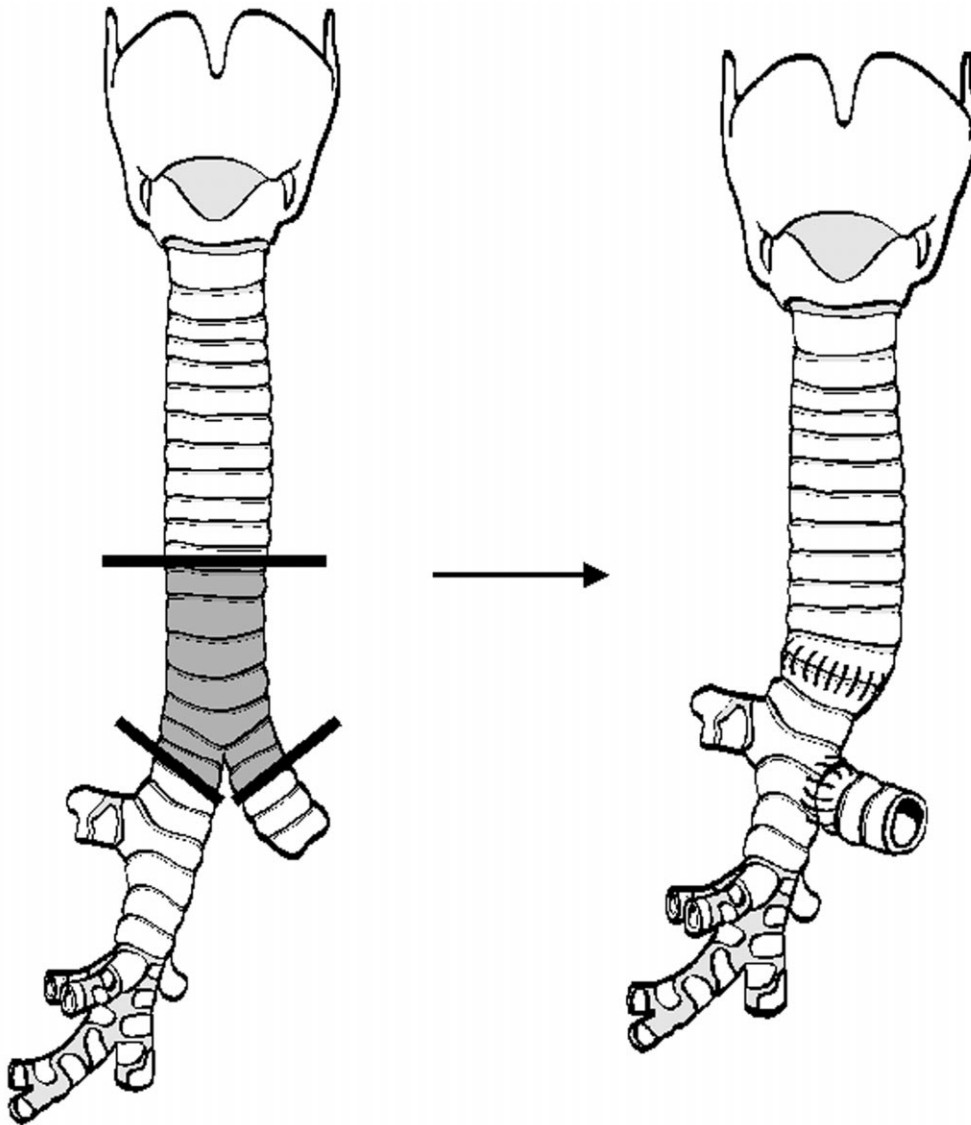


Fig. 3. Reconstruction in extended resection of the trachea and bifurcation: in extended resection of the distal trachea and the carina, reconstruction can be performed by implanting the left main bronchus into the bronchus intermedius ($n = 4$).

with a squamous cell carcinoma of the lower trachea, preoperative radiotherapy, R1 situation, died after 13.5 months of the local recurrence without metastases; one patient with an adenoid cystic carcinoma of the bifurcation, R1 situation, postoperative percutaneous and endoluminal radiotherapy, died after 7.1 years of a local recurrence with pulmonary metastases). A tracheal stent was implanted to prevent tracheal obstruction. One female patient with a squamous cell carcinoma died after 25 months from osseous and cranial metastases without local recurrence. The long-term results are summarized in Table 2.

4. Discussion

Primary tracheal tumors are rare. In large studies, the

relative incidence of tracheal tumors compared to bronchial tumors is under 2% [3]. However, there is only a relative incidence of 0.6% in our thoracic surgery patient population. Clinical symptoms such as coughing, dyspnea and stridor are unspecific and insidious and these tumors are often slow-growing. We can confirm the long delay in making the diagnosis observed by Perelman et al. [3]. He reported a mean delay of 25 months in benign and 8 months in malignant tumors.

Bronchoscopy plays a central role in the diagnostics as well as in therapy of tracheal tumors. Local complications such as bleeding, fistulas and subtotal obstruction may be treated on an emergency basis under general anesthesia with rigid instruments [9]. When rapid recanalisation or hemostasis are required endoluminal laser therapy with the Nd-YAG laser has proven to be effective [10,11]. Eight of 14

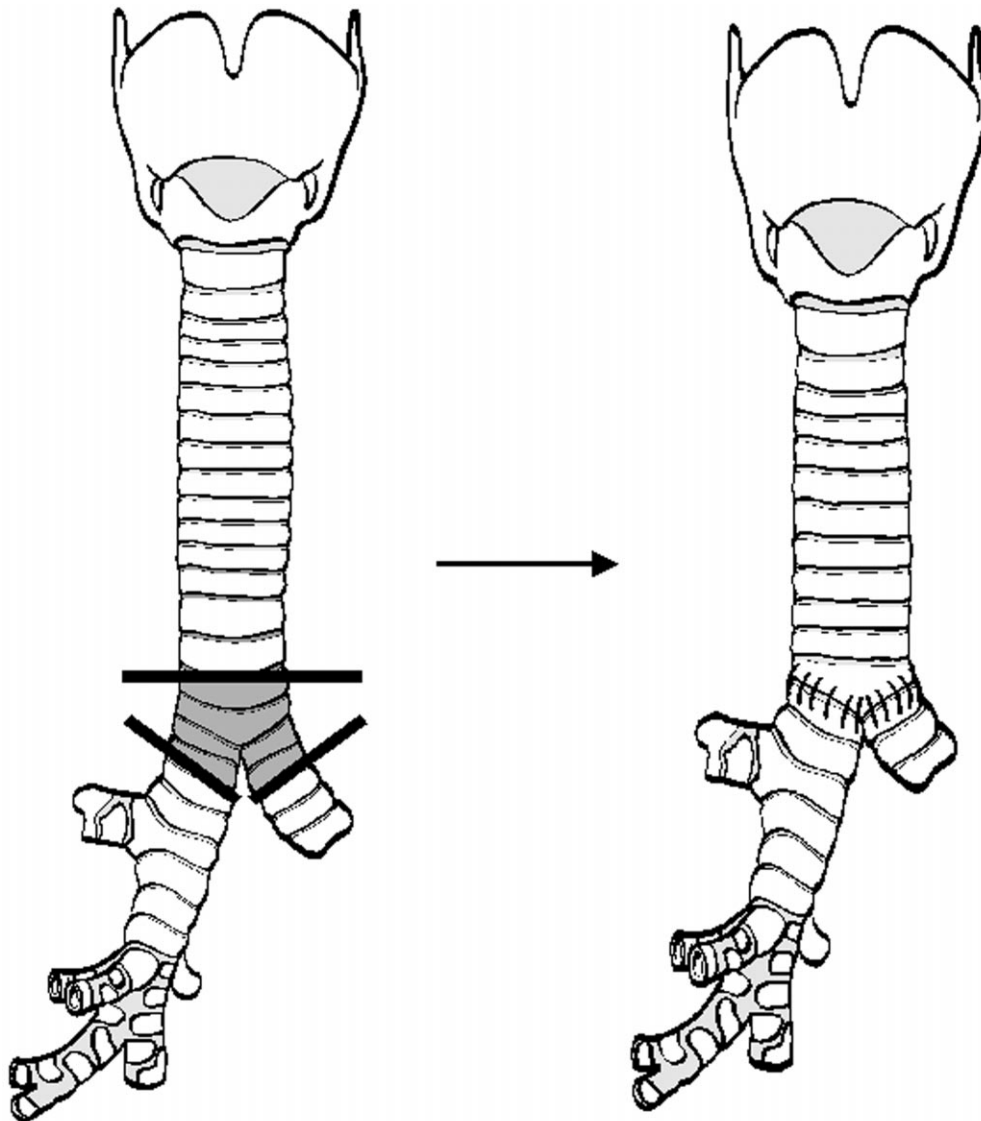


Fig. 4. Reconstruction in resection of bifurcation: in limited resection of the distal trachea and the carina, a neo-carina may be fashioned ($n = 1$).

patients were preoperatively recanalised for continuation of diagnostic measures. However, laser therapy alone has only a palliative character in malignant tumors because in most cases the tumor, perforates all parietal layers into the mediastinum like an iceberg [12]. Therefore resection is the only potentially curative treatment for primary malignant tracheal tumors [4,5,12]. Furthermore, the aim of bronchoscopy in the postoperative phase is to diagnose and treat secretion retention and anastomotic wound healing disorders. Most of the wound healing disorders can be treated successfully by repeated laser granulation ablation, bouginage or stent implantation. One case of a cicatricial stenosis of the right superior lobe required a sleeve lobectomy. Interventional bronchoscopy also plays a decisive role in tumor therapy [11,13]. Improved local tumor control is possible by saturation of the radiation dose. In three patients the external beam

radiotherapy was combined with endoluminal high-dose-rate (HDR) brachytherapy: in two of them (carcinoid and mucoepidermoid carcinoma) definitive local control was assessed. Another patient with an adenoid cystic carcinoma and R1 had 4 recurrence-free years. In adenoid cystic carcinoma as well as in patients with lymph node involvement or non complete tumor resection, we perform an adjuvant external beam radiotherapy. Doses of 56 Gy after R0-resection are delivered. Single doses were 2 Gy/day, 5 days/week. Radiotherapy should follow surgery after 3–6 weeks. In patients with microscopic residual tumor, a dose of 64 Gy is given. However in these cases we prefer an additional endoluminal HDR brachytherapy. HDR (median 15 Gy, single dose 3–5 Gy to 1 cm from the source axis) is delivered 2 weeks after external beam radiotherapy of 50 Gy [13].

The development of modern mobilization procedures

Table 1
Complications after surgery (in 11 out of 14 patients with resection for primary tracheal tumors)

	<i>n</i>	Note
<i>Minor complications</i>		
Secretion retention	7	Tracheotomy (<i>n</i> = 2)
Other minor complications (arrhythmia, thrombosis, gastritis)	5	
Minor wound healing disorders at the anastomosis	4	Successful endoscopic treatment
Restenosis (by granulation)	3	Interventional bronchoscopy
Recurrent nerve palsy	1	
<i>Major complications</i>		
Respiratory distress	2	1 bilateral pneumonia with septicaemia and exitus letalis; 1 ventilator therapy, extubation after 5 days
Wound healing disorders at the anastomosis	2	1 cicatricial stenosis, secondary sleeve lobectomy 8 weeks later; 1 fistula and mediastinitis after implantation of prosthesis, exitus letalis

enabled the resection of more than half of the trachea and the restitution of continuity [14]. It is possible to achieve a tension-free anastomosis following infra- or suprahyoid release of the larynx [15], mobilization of the mediastinum and pericardial circumscision. In extensive resection of the bifurcation or the middle trachea, the left bronchus is divided and implanted in the bronchus intermedius (Figs. 2 and 3). By combining the different procedures, 60–70% of the trachea can be removed. For this reason, implantation of prosthetic materials should be avoided because they may disturb healing of the anastomosis with subsequent mediastinitis and fatal bleeding of the innominate artery. We were not able to confirm the favorable results reported by Neville et al. [16,17]. Implanting a prosthesis is only justifiable in exceptional cases, if unexpectedly the defect cannot be closed during surgery despite all mobilization maneuvers.

The most radical tumor removal has to be accomplished by surgery. Thus, there is the demand to make the frozen section diagnosis of the resection margins during surgery

[4]. Adenoid cystic carcinomas have the property of circumferentially and longitudinally spreading far beyond the visible tumor border into the mucosa and lymphatics. In an R1 situation, the question arises during surgery whether to extend the resection with the risk of anastomotic tension or to leave the R1 situation. Grillo and Mathisen [5] demonstrated that R1 resection has no adverse effect on wound healing or long-term results. Maziak and Pearson [18] confirms these observations that long-term survival is not significantly decreased (90 months in R1 vs. 119 months in R0). Regnard et al. [19] had similar findings. Adenoid cystic carcinomas are very radiosensitive and slow-growing so that it is justified to accept an R1 situation. Our own results confirm these observations: we did not find a higher rate of wound healing disorders with R1 resection. Adenoid cystic carcinomas have a favorable long-term prognosis but very late recurrence may occur. Perelman and Koroleva [3] report a 66% 5-year and a 55% 10-year survival probability in 56 resected patients. Grillo and Mathisen [5] achieved a 5-year survival probability of 75% in 41 patients. Eschappasse [12] obtained disease-free survival of 3–9 years in 5 of 19 patients. Maziak et al. [18] and Pearson et al. [20] also reported a 5-year survival of 79%, 10-year survival of 51% and disease-free survival of 20 years. Regnard et al. [19] in his large multicenter study observed a 73 and 57% respective survival for 5 and 10 years. Squamous cell carcinomas have a poorer prognosis. Perelman and Koroleva [3] found a 15% 5 year survival probability in 20 patients, and Grillo and Mathisen [5] estimate the 5-year survival probability to be 35%. Regnard et al. [19] found 47% 5-year survival and 36% 10-year survival.

In conclusion, primary tracheal tumors are relatively rare so that, with a few exceptions, there is only a small number of cases reported internationally. However, there is a consensus that resection is the best prerequisite for local tumor control in malignant, and occasionally benign, primary tracheal tumors. By extensive mobilization procedures, 60–70% of the adult trachea can be resected, but prosthetic material should not be used for reconstruction when at all possible. Fatal anastomotic leakage is a rare complication. Endoluminal brachytherapy improves local tumor control, especially in nonradical surgery. High-precision endoscopy with the

Table 2
Long-term results in patients resected for primary tracheal tumor (*n* = 14, 1987–1996)

Histology	<i>n</i>	Radicality	Survival	Recurrence	Alive and recurrence-free
Adenoid cystic	7	R0 (<i>n</i> = 4)	25 months in median (5 days to 8 years)	Brain (<i>n</i> = 1)	<i>n</i> = 3
		R1 (<i>n</i> = 2)		Local + lung (<i>n</i> = 1)	
		R2 (<i>n</i> = 1)			
Mucoepidermoid	2	R0 (<i>n</i> = 2)	9.9 and 9.4 years	–	<i>n</i> = 2
Squamous cell	2	R0 (<i>n</i> = 1)	27 months and 13.5 months	Local (<i>n</i> = 1)	<i>n</i> = 1
		R1 (<i>n</i> = 1)			
Carcinoid	1	R1	4.5 years	–	<i>n</i> = 1
Benign tumor	2	R0	11 months and 40 months	–	<i>n</i> = 2

possibility of interventional procedures is a pre- and post-operative component of modern tracheal surgery.

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