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Management of congenital tracheal stenosis in infancy☆

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Abstract

Objective: Congenital tracheal stenosis (CTS) is a very infrequent malformation. Till recently, the outlook for these patients was dismal because medical management was the only way of treatment. Surgical and endoscopical techniques developed in the last years have improved the prognosis. We review the short- and long-term outcomes of a single institution experience in the management of children with CTS, comparing different treatment modalities. **Methods:** Between 1991 and 2004, 19 cases of CTS have been managed in our Unit. Respiratory symptoms varied from mild stridor on exertion to severe distress. Bronchoscopy was performed for diagnostic purposes in all cases; other imaging techniques (computed tomography (CT), magnetic resonance imaging (MRI), bronchography, angiography, doppler-ultrasound) were performed on an individual basis. According to clinical and endoscopical features, patients were classified into three groups. The following data have been studied in each case: sex, age at diagnosis and treatment, anatomical type, associated anomalies, treatment modality, complications, outcome and time of follow-up. **Results:** Ten boys and nine girls have been included in this study. Age at diagnosis ranged from 3 days to 7 years (median, 4 months) and 84% of cases showed associated anomalies. Five patients presented mild or no symptoms and have been managed expectantly. The other 14 cases were operated on because of persistent or severe clinical symptoms. The following procedures were performed: slide tracheoplasty ($n = 7$), costal cartilage tracheoplasty ($n = 5$), tracheal resection and reconstruction ($n = 3$), endoscopical dilatation ($n = 3$), stent placement ($n = 1$), and laser resection ($n = 1$). Three patients required two or more procedures and surgical survival rate is 78%. Overall mortality in the series is 21% and all survivors (15 patients) are asymptomatic or show mild symptoms with respiratory infections only. Follow-up is complete, ranging from 8 months to 12.3 years (mean, 5 years). **Conclusions:** Bronchoscopy is our preferred diagnostic tool. Selection of the type of treatment depends on the patient's clinical status and the anatomical pattern of the stenosis. In symptomatic cases with short-segment stenosis (<30% of total tracheal length), we prefer tracheal resection with end-to-end anastomosis; for long-segment stenosis (>30%), slide tracheoplasty is our procedure of choice.

Keywords: Tracheal stenosis; Congenital airway anomalies; Tracheal surgery; Bronchoscopy

1. Introduction

Congenital tracheal stenosis (CTS) is an infrequent structural obstructive lesion of the airway, representing between 0.3 and 1% of all laryngo-tracheal stenosis [1]. Until recently, the outlook for patients with CTS was dismal because an appropriate treatment was not available and many of them died as the result of acute airway obstruction. Fortunately, the prognosis of these patients has improved in the last decade due to the development of new surgical and endoscopical techniques that allow an early and effective reconstruction of the stenotic trachea. At the present moment, the challenge consists more in reducing the high morbidity usually associated to the treatment of this malformation rather than in achieving survival. Herein, we review the short- and long-term outcomes of a single institution experience in the management of children with CTS, comparing different treatment modalities.

2. Materials and methods

We have performed a retrospective study of patients with a diagnosis of CTS treated in our Pediatric Airway Unit between 1991 and December 2004. All patients showed respiratory symptoms varying from mild stridor on exertion to severe distress and apnea. The patients were managed by a multidisciplinary team including pediatricians, pediatric surgeons, cardiac surgeons, anesthesiologists, and trained nurses.

Bronchoscopy was performed for diagnostic purposes in all cases. Chest X-ray and echocardiography have also been

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made in every patient. Other imaging and diagnostic techniques such as computed tomography (CT) scan, magnetic resonance imaging (MRI), broncography, pulmonary function tests or angiography, were performed on an individual basis. Finally, gastroesophageal reflux (GER) was a matter of concern and a 24-h pH-monitoring test or an esophagogram was obtained in everyone.

Patients were classified into three groups (Table 1) according to their clinical status and the endoscopic findings. The following parameters have been compiled: sex, age at diagnosis, clinical group, anatomic type, associated anomalies, treatment modality, complications and their treatment, result, and time of follow-up. In addition, the following facts were studied in the group of surgically treated patients: age at surgery, surgical technique, use of cardiopulmonary by-pass, postoperative airway intubation or stenting, and length of hospital stay.

Choice of type of treatment was based on the following criteria: (1) clinical group, (2) anatomic type of stenosis (length and diameter), and (3) period in which the patient presented.

Associated cardiovascular malformations requiring surgical treatment were repaired simultaneously with the stenotic airway.

3. Results

Nineteen patients presented CTS in the period of study. There were 10 boys and 9 girls, and age at diagnosis ranged from 3 days to 7 years (median, 4 months). Fifteen patients (79%) showed a long segment stenosis (>30% total tracheal length) and four (21%) a short tracheal stenosis. In two cases the tracheal stenosis extended to a main bronchus. Associated anomalies were present in 84% of patients (Table 2) and severe GER, surgically treated, was included (31%). Eleven patients (57%) displayed more than one associated anomaly.

All the patients presented with stridor and in 26% it was considered severe, moderate in 42% and mild or occasional in 31% of cases. Respiratory distress was observed in 73% of patients. Five patients (26%), classified as clinical group I, have not required surgical treatment and are currently asymptomatic or only show mild symptoms. In the other 14 patients (73%), clinical groups II and III, 15 surgical procedures were performed: tracheal resection and reconstruction ($n = 3$), costal cartilage tracheoplasty ($n = 5$), and slide tracheoplasty ($n = 7$). In one case, two different techniques were used, a costal graft tracheoplasty initially and a resection of the restenosed segment afterwards.

Endoscopic techniques (dilatation, laser resection, and stenting) have been used mainly in the treatment of postoperative complications and in only one case we used them prior to surgery.

Survival rate is 79% (15 patients) and all survivors are asymptomatic or show only occasional symptoms. Follow-up is complete in all case, ranging from 8 months to 12.3 years (mean, 5 years). More specific details of the results are given for each type of treatment.

Table 1
Clinical classification of congenital tracheal stenosis

Group I: mild stenosis
Endoscopic findings: narrow posterior membranous trachea or complete tracheal rings with adequate diameter (4–6 mm in a small infant)
Asymptomatic or occasional symptoms
Group II: moderate stenosis
Endoscopic findings: complete tracheal rings
Symptomatic, no respiratory embarrassment
Group III: severe stenosis
Endoscopic findings: complete tracheal rings
Very symptomatic, respiratory embarrassment
Subgroups
A: no associated anomalies
B: associated anomalies

Table 2
Associated anomalies

Type of malformation	Number of cases
Gastroesophageal reflux	6
Tracheal bronchus	5
Pulmonary artery sling	5
Unilateral lung agenesia	3
Polidactylia	3
Atrial septal defect	3
Tracheal cartilaginous sleeve	2
Left superior Cava vein	2
Crouzon syndrome	2
Anorectal malformation	2
Ventricular septal defect	1
Double aortic arc	1
Hipospadias	1
Down syndrome	1
Other skeletal malformation	1

3.1. Medical management

Five patients, clinical group I, have been managed expectantly (Table 3; Fig. 1). Medical treatment consists of respiratory physiotherapy, antibiotics in case of infection, and a close survey. Two patients with Crouzon syndrome and congenital tracheal cartilaginous sleeve, with a diffuse tracheal stenosis, are included in this group. Three patients presented severe GER requiring surgical treatment. One patient died due to unrelated causes and the other four are alive and doing well. Mean follow-up is 42 months.

3.2. Surgical treatment

Fourteen patients, clinical groups II and III, were operated on (Table 3). Mean age at diagnosis was 8 months and 14 months when treated. Three surgical techniques have been used: resection and reconstruction, autologous costal cartilage tracheoplasty, and slide tracheoplasty.

Tracheal resection was performed in two patients with short segment stenosis (<30% total tracheal length) and cardiopulmonary by-pass was used during surgery in one patient because of a double aortic arc. Mean postoperative intubation and hospital stay were 12 and 33 days, respectively. The only complication was a partially obstructive intraluminal scar (mild restenosis) that was successfully resected endoscopically with YAG laser. Long-

Table 3
Results related to clinical classification and treatment

Clinical group	Number of patients	Treatment	Complications	Final result
I A	1	Medical	No	Good
I B	4	Medical	No	Good (100%)
II A	1	Surgical (CCT)	Yes	Good
II B	8	Surgical (5ST/2CCT/1TR&R)	Yes (37%)	Good (87%)
III A	1	Surgical (TR&R)	No	Good
III B	4	Surgical (2 CCT/2 ST)	Yes (75%)	Good (50%)

CCT, costal cartilage tracheoplasty; ST, slide tracheoplasty; TR&R, tracheal resection and reconstruction. Good result: asymptomatic patient or mild respiratory symptoms with infection.

term results have been good in both cases with a mean follow-up of 11.7 years.

Five patients with long stenosis were treated with a costal cartilage tracheoplasty. Early in our series, two patients died during the surgical procedure and another one in the early postoperative period. Cardiopulmonary by-pass was not used in the first two patients and the fatal outcome was due to ventilatory and hemodynamic complications.

The two survivors (40%) are currently doing well but complications have appeared during follow-up. One showed a restenosis that required endoscopic dilatation, surgical resection, and tracheal stenting with a silicone prosthesis (Dumon, Novathec) (Fig. 2). The other patient required endoscopic dilatation postoperatively, because of granulation tissue, and a temporary tracheostomy. Mean postoperative intubation was 30 days and average hospital stay was 61 days. Follow-up has been 12.5 years for the first case and nearly 7 for the second.

Seven patients have been treated with a slide tracheoplasty since 1996 (Fig. 3). All, except one, displayed a long segment stenosis and five had a left pulmonary artery sling. In addition, three patients had a tracheal bronchus. Cardiopulmonary by-pass was used during the procedure in every case and extubation was carried out in the operating room or early after the operation (mean, 12 h). Average hospital stay was 21 days and mean time of follow-up was 2.8 years. All patients are asymptomatic and the only postoperative complication was a wound infection with mediastinitis.

In summary, good results have been achieved in 78% of operated patients (n = 14) with a mortality rate of 21%.

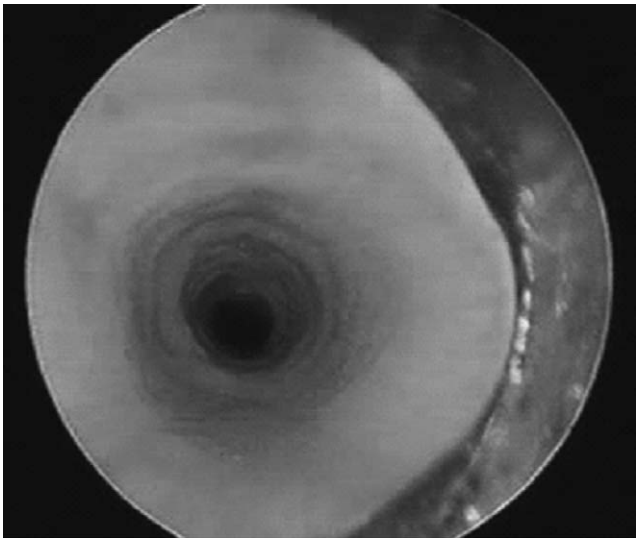

Fig. 1. Bronchoscopic view of a moderate tracheal stenosis (with complete rings) managed nonoperatively.

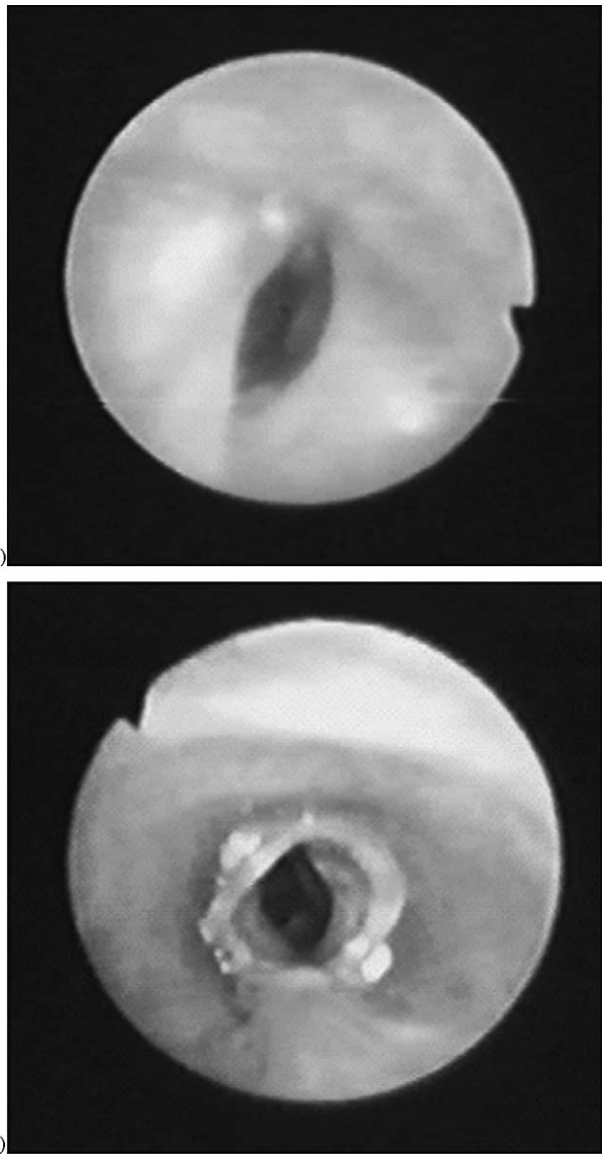

Fig. 2. (a) Endoscopic aspect of a restenosis after costal cartilage tracheoplasty. (b) An endoluminal silicone stent maintains the tracheal lumen.

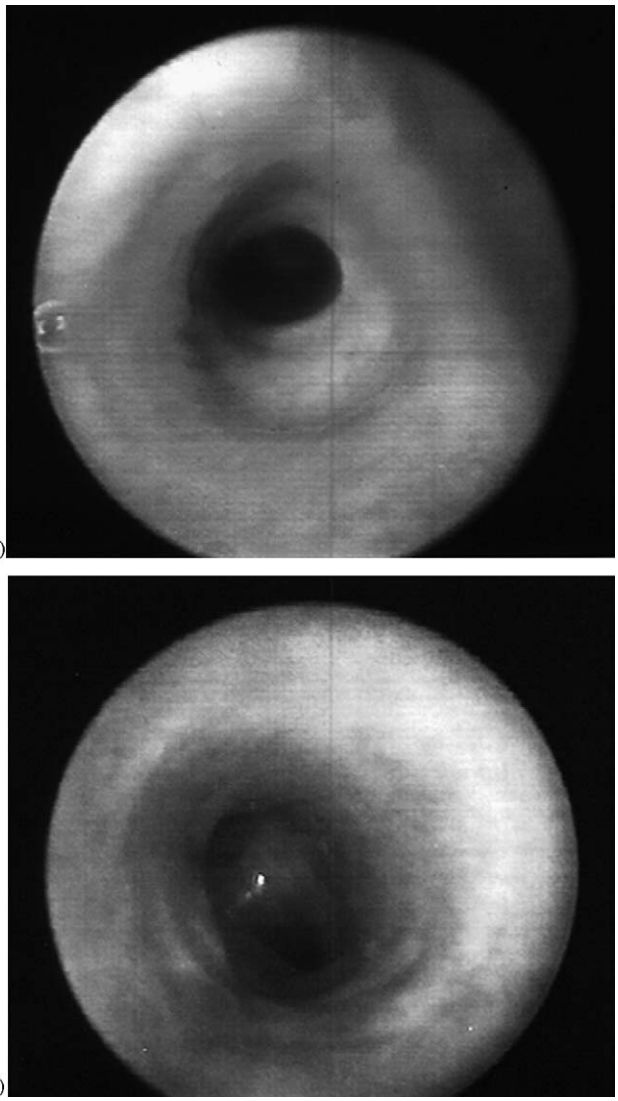

Fig. 3. (a) Diagnostic bronchoscopy of a severe CTS. (b) The same case after a slide tracheoplasty.

3.3. Endoscopic treatment

Endoscopic procedures were used mainly in the treatment of postsurgical complications (Fig. 2). Three patients required endoscopic techniques to treat complications such as restenosis (two cases) and granulation tissue formation (one patient). In the only case in which an endoscopic electroresection, followed by dilatation, was performed as the initial treatment, it was a failure. In one patient with a mild CTS, clinical group I, and severe GER which debutated with a bacterial tracheitis, bronchoscopy was effective in the debridement and cleaning of the airway.

4. Discussion

Most of the published data on CTS describe cases with complete tracheal rings and severe symptoms treated surgically [2,3]. Nevertheless, we have observed that some patients have mild stenosis with little clinical expression not requiring surgical reconstruction [4–6]. Anatomically, these stenosis may show either a tracheal ring with a narrow pars membranosa or a completely circular one with an adequate diameter (between 4 and 6 mm in small infants; Fig. 1). Rutter et al. [7] have tried a nonoperative management in a group of seven patients with complete tracheal rings achieving good results. Elliott et al. [8] describe CTS as a wide spectrum of pathologies with diverse clinical expression, from simple stridor to life threatening episodes of apnea. In addition, they state that CTS is not synonymous of complete tracheal ring although the association is frequent. These observations support our findings and experience published previously [4,6].

Clinical and radiological data point that the airway diameter of children with CTS grows with time [9]; so the type of treatment should be tailored to each particular case. Taking this into account, we have classified the patients according to clinical aspects, endoscopic findings, and the presence of associated anomalies (Table 1). Assembling the cases into three categories enables anatomo-clinical correlation and, with further experience, may allow group-related prognosis (Table 3) [6]. Tracheo-bronchoscopy is our preferred diagnostic tool. It is the most reliable procedure in establishing the type of lesion, its characteristics, location and extension. In addition, it allows the evaluation of the dynamic behavior and settles the amount of nonaffected airway. Spiral computed tomography with multiplanar reconstruction (CT/MPR) and magnetic resonance imaging are very useful in the diagnostic work-up of tracheobronchial anomalies. CT/MPR provides a good anatomic delineation of the airway, and MRI enables full assessment of the vascular structures and their relations to the adjacent trachea [10]. The increasing availability of these techniques makes bronchography unnecessary in most cases of CTS, so we no longer use it.

As it has already been stated, patients with mild or no symptoms may be treated in an expectant way. In symptomatic cases operative treatment is indicated. Othersen et al. [11] and Clement et al. [12] have tried laser division and dilatation of the stenotic rings, but as Wright [13] has pointed, there is very little experience and the technique is not reliable yet. In our series, the endoscopic procedures have been very useful in the management of postoperative complications, dilating and stenting restenosed tracheal segments and removing granulation tissue.

Surgical procedures fall into three categories: (1) tracheal reconstruction with autologous tracheal tissue (tracheal resection, slide tracheoplasty, and free tracheal autograft); (2) tracheoplasty with nontracheal autologous tissue (costal cartilage or pericardial patch); and (3) tracheal transplant with cadaveric homograft.

Tracheal resection with end-to-end anastomosis is the treatment of choice for short segment stenosis [2,14]. Although this procedure has been used in long stenosis, 50% or more of the tracheal length, excessive tension in the anastomosis considerably increases the risk of leakage and restenosis [15]. Most of the authors do not recommend this technique if the lesion involves more than 4–5 rings that is 30% of total tracheal length [16,17]. Longer stenosis are best managed by means of tracheoplasty. Several types of tracheoplasties have been described ever since Kimura

et al.'s [3] first report in 1982. Published data concerning this technique show a mortality rate ranging from 0 to 55%, and complications around 40% of cases [18,19]. We have used it in five cases and long-term results have been satisfactory only in two (Fig. 2).

Slide tracheoplasty, described by Tsang et al. [20] in 1989, constitutes a landmark in the treatment of CTS. Its main advantage is that tracheal reconstruction is performed using native tracheal tissue, so granulation tissue formation is minimal and stenting is avoided. Published results with this technique are outstanding with an overall mortality rate around 9% [8,17]. Moreover, recent experimental and clinical data have shown tracheal growth after slide tracheoplasty [21,22]. We have used this procedure in seven cases and the results have been excellent (Fig. 3). Indications for slide tracheoplasty are spanding, short stenosis and stenosis associated with tracheal bronchus can also be managed with this technique, as we did in our series. Moreover, Grillo et al. [17] have treated successfully a 'bridge' bronchus, in a 6-month-old patient, with this technique.

In one of the few studies that compare two types of techniques, Tsugawa et al. [23] conclude that the results obtained with slide tracheoplasty in 17 patients (76% of survival and 58% asymptomatic) are superior to those obtained with costal cartilage tracheoplasty in 12 operated cases (66% of survival and 50% asymptomatic). In addition, they recommend delaying surgery beyond the first year of life, if the clinical status allows it, since the postoperative course is much better. Data concerning the long-term outcome of patients treated surgically are scarce. Most of the reports have been published in the last decade and only in a few cases follow-up is extended. Grillo et al. [17] and Kutlu and Goldstraw [24] support the good long-term results of slide tracheoplasty, and Backer et al. [25] report a similar outcome in patients treated with a tracheal autograft. It seems that if the patient overcomes the short-term after the reconstructive surgery, the trachea grows and its function improves with time. Nevertheless, physiological studies are still necessary in order to confirm this clinical observation.

Management of patients with CTS is complex and requires a close cooperation between pediatricians, surgeons, anesthesiologists, and nursing staff. Selection of the most suitable treatment depends on the patient's clinical situation and the anatomic type of stenosis.

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Appendix A. Conference discussion

Dr A. Turna (*Istanbul, Turkey*): What was the surgical basis of costal cartilage tracheoplasty? Having said such a high mortality, would you recommend this technique for selective patients after this analysis?

Dr Anton-Pacheco: We do not recommend. We do not perform costal cartilage tracheoplasty anymore. These were the first cases of the series, 10 years ago, 12 years ago, and some of the patients were operated without cardiopulmonary by-pass. So we had ventilatory and hemodynamic problems during surgery. And we think that's the main problem, apart from that two of them were very sick when they were operated.

Dr Turna: In the patients with costal cartilage tracheoplasty, could you give the information about the immediate tracheal fistula?

Dr Anton-Pacheco: You mean the complications of the two survivors, or the complications of the three patients that died?

Dr Turna: I meant to say the perioperative complication, tracheal fistula.

Dr Anton-Pacheco: Well, we have one leak and mediastinitis and that patient died in the postoperative course. And the other two survivors had restenosis in one case and granulation tissue in the other.

Dr D. Branscheid (*Grosshansdorf, Germany*): I missed a little bit the view to the pretreatment of those kids. We have a lot of problems in that little children, they got a tracheostomy, they got conservative treatment before they got dilatation, they got stents, they got insufficient operations before they came then, definitively, to resection of the trachea which made them good.

Do you have in your series also some of those kids, and what do you recommend?

How is your cooperation with the pediatricians?

Dr Anton-Pacheco: We don't have patients that have been operated in other institutions, first, for the moment.

And then we classify the patients. There are some patients with very mild symptoms and so they are treated nonoperatively with physiotherapy, close surveillance and antibiotic therapy in case of infection. And then we follow them very, very narrowly. We repeat the bronchoscopies, and we see how they are. And if they become symptomatic, then they are operated on.