

Surgical treatment of congenital tracheal stenoses*

Tratamento cirúrgico das estenoses traqueais congênitas

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Abstract

Objective: To analyze the outcomes of patients undergoing repair of congenital tracheal stenosis. **Methods:** This was a retrospective review of congenital tracheal stenosis patients treated between 2001 and 2007 at the University of São Paulo School of Medicine *Hospital das Clínicas* in São Paulo, Brazil. **Results:** Six boys and one girl (age at diagnosis ranging from 28 days to 3 years) were included. Five of the patients also had cardiac or major vessel malformations. The stenosis length was short in three patients, medium in one and long in three. The techniques used were pericardial patch tracheoplasty in three patients, resection and anastomosis in two, slide tracheoplasty in one and vascular ring correction in one. One patient died during surgery due to hypoxia and hemodynamic instability, and one died from septic shock on postoperative day 11. Other complications included pneumonia, arrhythmia, stenosis at the anastomosis level, residual stenosis, granuloma formation and malacia. The mean follow-up period was 31 months; four patients were cured, and one required the use of a T-tube to maintain airway patency. **Conclusions:** Congenital tracheal stenosis is a curable disease. However, its repair is complex and is associated with high rates of morbidity and mortality.

Keywords: Tracheal stenosis/congenital; Tracheal diseases; Surgical procedures, operative.

Resumo

Objetivo: Analisar os desfechos dos pacientes submetidos ao reparo de estenose congênita de traqueia. **Métodos:** Análise retrospectiva dos pacientes com estenose traqueal congênita tratados no Hospital das Clínicas da Faculdade de Medicina da Universidade de São Paulo entre 2001 e 2007. **Resultados:** Seis meninos e uma menina (idade ao diagnóstico entre 28 dias e 3 anos) foram incluídos. Cinco pacientes apresentavam malformações intra-cardíacas e/ou de grandes vasos associadas. A extensão das estenoses foi curta em três pacientes, média em um e longa em três. As técnicas utilizadas foram traqueoplastia com enxerto de pericárdio em três pacientes, ressecção e anastomose em dois, traqueoplastia em bisel em um e correção de anel vascular em um. Um paciente morreu no intraoperatório por hipóxia e instabilidade hemodinâmica e outro no 11º dia pós-operatório por choque séptico. Outras complicações observadas foram pneumonia, arritmia, estenose na anastomose e estenose residual, malácia e formação de granulomas. O tempo médio de seguimento pós-operatório foi de 31 meses; quatro pacientes ficaram livres da doença e um necessitou de tubo T para manter a via aérea pérvia. **Conclusões:** A estenose congênita de traqueia é uma doença curável. Entretanto, seu reparo é complexo e está associado a taxas de morbidade e mortalidade significativas.

Descritores: Estenose traqueal/congênita; Doenças da traqueia; Procedimentos cirúrgicos operatórios.

Introduction

Congenital tracheal stenosis is a rare and potentially lethal malformation including a variety of entities that, although pathogenically distinct, are all associated with a reduction in airway diameter.⁽¹⁾ As of 1999, fewer than 200 cases had been described in the literature.⁽²⁾

The diagnosis is difficult because the initial signs and symptoms, especially those of a respiratory nature, can present a broad spectrum of severity—ranging from mild stridor to severe respiratory failure. The symptoms typically occur in the first months of life, after a bacterial

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infection or after intubation, when the fragile respiratory system of such patients decompensates due to the reduction in airflow.^(2,3)

The diagnosis is frequently delayed due to the rarity and diversity of manifestations of the disease. In addition, other malformations often divert the attention of the team of attending physicians. Experience and clinical suspicion are essential for accurate and early diagnosis. Unfortunately, prenatal diagnosis is still not possible, since the accuracy of intrauterine examination remains limited.

Congenital stenosis and its treatment are classically associated with high rates of morbidity and mortality. Endoscopic and surgical techniques developed in recent decades have contributed to better outcomes in such patients.⁽¹⁾ However, a highly specialized team and individual management are essential for good results. No technique—resection and anastomosis; pericardial patch tracheoplasty; or slide tracheoplasty—is accepted as definitive, and no surgical technique corrects all of the anatomic variants of this disease.⁽⁴⁻⁷⁾ The treatment choice is a controversial issue, and the approach adopted at each facility varies, mainly depending on the clinical status of patients and the characteristics of the stenosis.⁽⁸⁾ The objective of the present study was to analyze the evolution and outcomes of patients undergoing repair of congenital tracheal stenosis at our facility.

Methods

This was a retrospective study of patients treated between January of 2002 and December of 2005. Patient charts were reviewed for the following data: gender; age; comorbidities; symptoms; surgical procedures; complications; and outcomes. The present study was approved by the Ethics Committee of the University of São Paulo School of Medicine *Hospital das Clínicas*.

All patients underwent bronchoscopy, together with CT of the chest and cervical region, as well as transthoracic echocardiography. Those tests were performed to confirm the diagnosis of stenosis, establish its characteristics and investigate the presence of associated cardiac or major vessel malformations. The patients were classified based on the stenosis length (as measured by bronchoscopy and CT) as follows: short (less than one third of the total tracheal length); medium (less than two thirds of the total

tracheal length); and long (two thirds or more of the total tracheal length). This classification, suggested by Elliot et al.⁽⁹⁾ in 2003, is similar to that devised by Cantrell & Guild, in which congenital stenoses are divided into three types: segmental stenosis; funnel-shaped stenosis; and generalized hypoplasia. The classification by Elliot et al. was chosen because it was considered simpler to use in clinical practice.

At our facility, the surgical strategy for the treatment of congenital tracheal stenosis is based on the stenosis length. In the cases in which the stenosis length was short (less than one third of the total tracheal length), the patients were treated with resection and tracheal reconstruction with end-to-end anastomosis (Figure 1a). Patients with long-segment stenosis underwent tracheoplasty with autologous pericardium, a procedure that consists in making a longitudinal incision on the anterior wall of the whole stenosed segment in order to enlarge the tracheal lumen and covering the resulting defect with a pericardial flap; this flap is fixed to the mediastinal tissue by sutures in order to prevent airway collapse (Figure 1b). Medium length-segment stenoses were treated by slide tracheoplasty, in which, initially, the stenosed area of the trachea is divided axially. After this section, the posterior wall of the cranial segment and the anterior wall of the distal portion are divided by means of median longitudinal incisions. Subsequently, the proximal and distal portions are slid and sutured, promoting tracheal shortening and enlargement of tracheal lumen (Figure 1c). All of these techniques have been previously described.^(10,11) Extracorporeal circulation was used on a case-by-case basis, and correction of cardiac or vascular anomalies was performed when necessary, preferably during the same operation.

Results

Six boys and one girl were included. The median age at diagnosis was 2 months (range, 28 days to 3 years). The patients presented a wide variety of respiratory signs and symptoms, the most common being dyspnea, cyanosis and wheezing. Two patients had respiratory infection at diagnosis.

Based on the bronchoscopy and CT findings, the stenosis length was classified as short in three patients, as medium in one and as long in three. The following intracardiac anomalies were

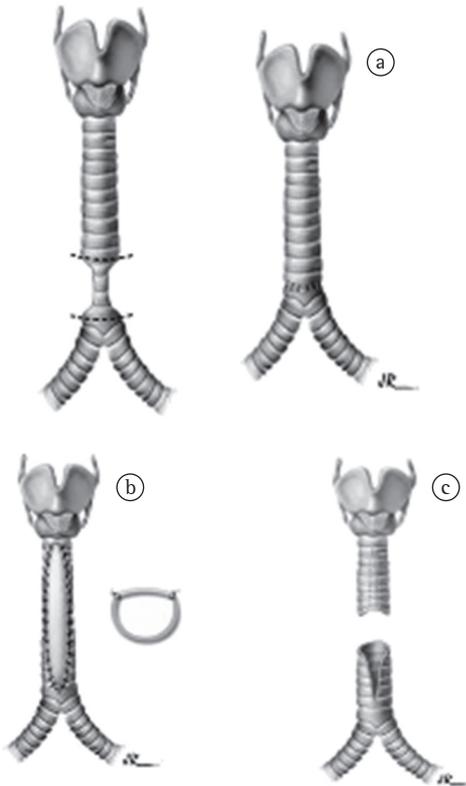


Figure 1 - In a), resection and end-to-end anastomosis: The stenotic segment is resected, and both tracheal stumps are anastomosed. In b), pericardial patch tracheoplasty: After a longitudinal incision in the stenotic trachea, a pericardial patch is sutured to the edges of the incision in order to constitute the anterior wall of the trachea, now with an enlarged lumen (detail). In C, slide tracheoplasty: First, a cross-sectional incision is made in the middle of the stenotic segment. Subsequently, an anterior and a posterior incision are made, and the edges are sutured, shortening the trachea and increasing the airway diameter.

found: tetralogy of Fallot in one patient; and interventricular communication accompanied by interatrial communication in another. Other congenital anomalies were as follows: dextrocardia and left superior vena cava, both in the same patient; double aortic arch (type 1A/Mayo classification) in one; and imperforate anus in one. Two patients presented pulmonary artery sling. The demographic characteristics of the patients are described in Table 1.

Surgical treatment was necessary in all cases, since all seven of the patients were highly symp-

tomatic. The three patients with long-segment stenosis underwent pericardial patch tracheoplasty. Extracorporeal circulation was used in two patients who presented cardiac malformations, which were corrected during the same operation. One of the patients died of severe intracardiac anomalies occurring during the procedure. In the remaining patients, the immediate postoperative complications were pneumonia (in one) and arrhythmia (in one). During the medium- and long-term follow-up period, one of those two patients remained asymptomatic, without the need for additional procedures. The other developed pronounced granulation in the trachea, accompanied by malacia at the patch site and residual stenosis at the carina. Various endoscopic procedures were needed in order to maintain airway patency. Granulomas were resected, dilatations were performed, and, finally, a T-tube was implanted. At the time of the final data collection, the T-tube remained in place, the patient was asymptomatic, and periodic endoscopic follow-up examinations were being performed in an outpatient setting.

Slide tracheoplasty was performed in one patient with tracheal and subglottic stenosis. In that case, extracorporeal circulation was also used. The patient developed extensive tracheal necrosis, together with tracheal fistula and pneumopericardium. In addition, the patient developed pulmonary atelectasis and pneumonia, dying on postoperative day 11 from sepsis secondary to infection at the surgical site.

Resection and end-to-end anastomosis were used in two of the patients with short-segment stenosis. One of those two had a cerebrovascular accident and developed renal failure in the postoperative period. However, despite a long stay in the intensive care unit, that patient was discharged in good clinical condition after 120 days. That same patient had stenosis at the anastomosis site, and two endoscopic dilatations were needed in the first 6 months. Subsequently, no other procedures were necessary. For the other patient, the postoperative course was excellent, without complications.

One of the patients presented congenital tracheal stenosis due to extrinsic vascular compression. In that patient, the anomalous aortic arch was ligated and sectioned. After the arch had been sectioned, the extrinsic compression ceased to exist—as confirmed by

Table 1 – Characteristics of the patients studied (patients included in the table by date of surgical procedure).

Patient	Gender	Age	Other diagnoses	Type of tracheal stenosis	Surgical procedure	Other procedures	ECC
01	F	3 years	PAS	Short-segment stenosis (PAS)	Resection and anastomosis	Pulmonary artery reimplantation	Yes
02	M	28 days	PAS, IAC, IVC, imperforate anus	Long-segment stenosis	Pericardial patch tracheoplasty	Pulmonary artery reimplantation	Yes
03	M	2 mos.	GER	Long-segment stenosis	Pericardial patch tracheoplasty	None	No
04	M	1.3 mos.	TF, GER	Long-segment stenosis	Pericardial patch tracheoplasty	Correction of TF	Yes
05	M	29 days	Dextrocardia, LSVC	Medium length-segment stenosis	Slide tracheoplasty	None	Yes
06	M	3.7 mos.	DAA	Short-segment stenosis (DAA)	Vascular ring correction	None	No
07	M	2 mos.	None	Short-segment stenosis	Resection and anastomosis	None	No

ECC: extracorporeal circulation; PAS: pulmonary artery sling; IAC: interatrial communication; IVC: interventricular communication; mos.: months; GER: gastroesophageal reflux; TF: tetralogy of Fallot; DAA: double aortic arch; and LSVC: left superior vena cava.

intraoperative bronchoscopy—and, therefore, no other additional tracheal procedures were necessary. The postoperative course was favorable, without complications.

The mean length of hospital stay was 45 days, and, excluding the inpatient deaths, the mean postoperative follow-up period was 31.6 months (range, 13-84 months). Our 30-day survival rate was 71%. By the end of the data collection period, all five of the surviving patients were asymptomatic and completely free of disease, although, in one case, the T-tube remained in place (Table 2).

Discussion

In congenital tracheal stenosis, the onset of symptoms can be quite dramatic. In our study, all patients presented at least one severe respiratory event prior to treatment. The presence of congenital malformations (especially cardiac malformations) makes diagnosis difficult. In this context, clinical suspicion is fundamental and should be followed by sophisticated assessment. All patients underwent echocardiography, bronchoscopy and CT of the chest.⁽⁹⁾ Those tests allowed us to confirm the diagnosis, as well as to characterize the stenosis and the malformations. In our study, echocardiography revealed cardiac malformations in two patients (28.5%), and chest CT detected vascular anomalies in four

(57%)—one patient presented malformation of the pulmonary artery and cardiac malformation.

There is no standard procedure for the treatment of congenital tracheal stenosis, and there have been no controlled studies comparing the different methods described (which would be quite difficult to compare due to the rarity of the disease, as well as to the severity of its distinct manifestations and associated diseases). The conservative approach is an option recommended for cases in which the stenosis is short and the diameter of the stenotic segment is more than 60% of the normal tracheal diameter.⁽¹²⁾ Such cases have been described in the literature. However, in the present study, none of the patients met those criteria and therefore all underwent surgical treatment.

The morbidity and mortality rates associated with the surgical treatment of congenital tracheal stenosis are high, and despite the growing worldwide experience, the latest publications show that the mortality rate is 18% in patients undergoing pericardial patch tracheoplasty and 24% in those undergoing slide tracheoplasty.^(13,14) In our study, the overall surgical mortality was 28.5%. This higher mortality rate might be due to the fact that our sample comprised children in whom the condition was more severe, children who were underweight and children whose diagnosis was delayed. The results of a recent meta-analysis tend to confirm this suspicion,

Table 2 – Surgical treatment results.

Patient	Hospital stay, days	Postoperative complications	Reintubation	Outcome	Follow-up
01	120	Dialytic renal failure	Yes	Outpatient decannulation	7 years
02	NA	NA	NA	Intraoperative death	
03	55	Pneumonia, multiple intubations	Yes	Outpatient decannulation	26 months
04	25	Arrhythmia, granuloma	Yes	Outpatient T-tube use	18 months
05	NA	Infection at the incision site, hypoxia, atelectasis.	Yes	Death on postoperative day 11	
06	17	None	No	Outpatient decannulation	17 months
07	11	None	No	Outpatient decannulation	13 months

NA: not applicable.

since two significant risk factors for morbidity and mortality were identified in such children: being less than 1 month of age and presenting associated intracardiac malformations.⁽¹³⁾ In the present study, 57% of the patients were at high risk according to these factors (two patients were less than 1 month of age, and two presented intracardiac anomalies), confirming the severity of the cases comprising our sample.

It is of note that few studies have addressed short-segment stenosis,⁽⁹⁾ which suggests that this is an uncommon characteristic. However, three of our patients had short-segment stenosis. The therapeutic option in these cases was for resection of the stenosis and end-to-end anastomosis (except for one patient with a vascular ring). Although some authors discourage the use of this procedure, in our study, we found that late evolution was favorable, suggesting that resection and anastomosis are a good option for this specific population.^(15,16)

Based on the experiences mentioned in the literature, we realized that complications are common after surgical treatment of congenital tracheal stenosis. The consequences of this fact can be measured by the long mean length of hospital stay found in the present study. Such complications are typically related to associated cardiac malformations, infectious profiles (especially pneumonia or mediastinitis) and tracheal reconstruction (restenosis, malacia or granulation tissue formation).^(4,5,14) In our study, two patients had severe complications leading to death: one died during surgery (this patient had developed hemodynamic instability and hypoxemia, and weaning from mechanical ventilation had been impossible); and one died on postoperative day 11 from septic shock. The patient who died during the procedure presented the two

factors previously described as increasing the risk for an unfavorable outcome: being less than 1 month of age; and presenting severe intracardiac anomalies (single ventricle and pulmonary stenosis). Two patients had postoperative tracheal stenosis, and, in both, the stenosis was treated endoscopically without difficulties. One of the patients presented reduction in the diameter of the anastomotic line, which evolved well after two dilatation sessions. Another patient presented residual stenosis at the carina and developed pronounced granulation tissue along the cranial edge of the suture. The residual stenosis was treated with hydrostatic balloon dilatation, the granulation tissue was resected by rigid bronchoscopy, and a T-tube was used in order to maintain the airway. It is of note that, during the follow-up period, the distal stenosis stabilized, and it was possible to progressively decrease the caudal extent of the T-tube.

The present study represents the experience of a referral facility for pediatric laryngotracheal and cardiac surgery in the treatment of congenital tracheal stenosis, a complex entity that requires specialized treatment and an adequate infrastructure. Our results show that, although surgical treatment of congenital tracheal stenosis is possible, it is associated with significant morbidity and mortality in high-risk patients.

References

1. Antón-Pacheco JL, Cano I, García A, Martínez A, Cuadros J, Berchi FJ. Patterns of management of congenital tracheal stenosis. *J Pediatr Surg.* 2003;38(10):1452-8.
2. Lang FJ, Hurmi M, Monnier P. Long-segment congenital tracheal stenosis: treatment by slide-tracheoplasty. *J Pediatr Surg.* 1999;34(8):1216-22.
3. Hoffer ME, Tom LW, Wetmore RF, Handler SD, Potsic WP. Congenital tracheal stenosis. The otolaryngologist's

- perspective. *Arch Otolaryngol Head Neck Surg.* 1994;120(4):449-53.
4. Tsugawa C, Nishijima E, Muraji T, Satoh S, Takamizawa S, Yamaguchi M, et al. Tracheoplasty for long segment congenital tracheal stenosis: analysis of 29 patients over two decades. *J Pediatr Surg.* 2003;38(12):1703-6.
 5. Backer CL, Mavroudis C, Dunham ME, Holinger L. Intermediate-term results of the free tracheal autograft for long segment congenital tracheal stenosis. *J Pediatr Surg.* 2000;35(6):813-8; discussion 818-9.
 6. Backer CL, Mavroudis C, Dunham ME, Holinger LD. Reoperation after pericardial patch tracheoplasty. *J Pediatr Surg.* 1997;32(7):1108-11; discussion 1111-2.
 7. Kim HK, Kim YT, Sung SW, Park JD, Kang CH, Kim JH, et al. Management of congenital tracheal stenosis. *Eur J Cardiothorac Surg.* 2004;25(6):1065-71.
 8. Antón-Pacheco JL, Cano I, Comas J, Galletti L, Polo L, García A, et al. Management of congenital tracheal stenosis in infancy. *Eur J Cardiothorac Surg.* 2006;29(6):991-6.
 9. Elliott M, Roebuck D, Noctor C, McLaren C, Hartley B, Mok Q, et al. The management of congenital tracheal stenosis. *Int J Pediatr Otorhinolaryngol.* 2003;67 Suppl 1:S183-92.
 10. Idriss FS, DeLeon SY, Ilbawi MN, Gerson CR, Tucker GF, Holinger L. Tracheoplasty with pericardial patch for extensive tracheal stenosis in infants and children. *J Thorac Cardiovasc Surg.* 1984;88(4):527-36.
 11. Tsang V, Murday A, Gillbe C, Goldstraw P. Slide tracheoplasty for congenital funnel-shaped tracheal stenosis. *Ann Thorac Surg.* 1989;48(5):632-5.
 12. Cheng W, Manson DE, Forte V, Ein SH, MacLusky I, Papsin BC, et al. The role of conservative management in congenital tracheal stenosis: an evidence-based long-term follow-up study. *J Pediatr Surg.* 2006;41(7):1203-7.
 13. Chiu PP, Kim PC. Prognostic factors in the surgical treatment of congenital tracheal stenosis: a multicenter analysis of the literature. *J Pediatr Surg.* 2006;41(1):221-5; discussion 221-5.
 14. Airway Reconstruction Team. Recent challenges in the management of congenital tracheal stenosis: an individualized approach. *J Pediatr Surg.* 2005;40(5):774-80.
 15. Beierlein W, Elliott MJ. Variations in the technique of slide tracheoplasty to repair complex forms of long-segment congenital tracheal stenoses. *Ann Thorac Surg.* 2006;82(4):1540-2.
 16. Hasaniya N, elZein CF, Mara S, Barth MJ, Ilbawi M. Alternative approach to the surgical management of congenital tracheal stenosis. *Ann Thorac Surg.* 2006;82(6):2305-7.

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