Tracheal Stenosis Successfully Treated without Surgery in Preterm Infant

Yae Eun Chung, MD¹,
Ran Lee, MD, PhD²,
Jae Sung Son, MD, PhD²,
Hye Won Park, MD, PhD²

¹Department of Pediatrics, Konkuk University Medical Center, Seoul;

²Department of Pediatrics, Konkuk University School of Medicine, Seoul, Korea

Congenital tracheal stenosis is a rare disease that can be life threatening, especially when early identification is missed. A premature twin showed progressive respiratory symptoms at birth and tracheal stenosis, with difficulty intubating. Chest computed tomography scan revealed diffuse tracheal stenosis and left pulmonary artery (LPA) sling. Tracheal ballooning under fluoroscopic guide was successfully performed at 5 days old and maintained with endotracheal intubation until 7 months old. The patient underwent surgery for LPA sling and is doing well without any respiratory symptoms following extubation. Herein, we report the first case of congenital tracheal stenosis in a preterm infant who was successfully treated with tracheal ballooning and conservative management in Korea.

Key Words: Tracheal stenosis, Infant, Premature birth, Dilatation

Introduction

Congenital tracheal stenosis is a rare disease, which can be life threatening, especially in the case of diffuse tracheal stenosis and when clinical recognition is delayed. Many cases of congenital tracheal stenosis have been associated with congenital heart defects including left pulmonary artery (LPA) sling.¹⁻³

Although diffuse bronchial stenosis cases treated with surgery have been reported in Korea,³ tracheal stenosis surgery for premature infants is difficult due to the small body size and limited space for manipulation. Tracheal ballooning may be a life-saving technique for premature infants below the suitable weight for surgical intervention.

Herein, we report on the case of a premature infant with both congenital tracheal stenosis and LPA sling. To our knowledge, this was the first case of fluoroscopically guided balloon dilation and conservative management of tracheal stenosis in premature infants in Korea.

Case

The preterm twins were delivered by cesarean section at 34⁺⁶ gestational weeks of pregnancy. The first baby weighed 1,846 g (10th percentile) and the second baby weighed 1,160 g (<10th percentile). Both twins had an Apgar score of 8 points at 1 minute and 9 points at 5 minutes after birth. Quadruple test for Edwards syndrome was performed with an abnormal result and subsequent amniocentesis was normal in both patients.

The first baby required oxygen after delivery and had retained lung fluid on chest radiograph. In the first day of life, he showed desaturation and chest retraction, so high flow nasal cannula (HFNC) was applied. On the 3rd day of life, the patient was in good condition while maintaining care at the flow rates of 4 L/minute and 0.21 of FiO₂ through HFNC and weaning off was planned by gradually lowering the HFNC setting. However, 6 hours after weaning off HFNC, tracheal intubation and ventilator care were required due to aggravation of respiratory distress

Received: 29 April 2020

Revised: 26 June 2020

Accepted: 30 June 2020

Correspondence to

Hye Won Park, MD, PhD
Division of Neonatology,
Department of Pediatrics, Konkuk University Medical Center, Konkuk University School of Medicine, 120-1 Neungdong-ro, Gwangjin-gu, Seoul 05030, Korea

Tel: +82-2-2030-7558

Fax: +82-2-2030-5122

E-mail: 20110673@kuh.ac.kr

Copyright© 2020 by The Korean Society of Perinatology

This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (<http://creativecommons.org/licenses/by-nc/4.0/>), which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided that the original work is properly cited.

with severe chest retraction, frequent and repeated episodes of desaturation, increased oxygen requirement and tachypnea about 70 times/minute. Chest x-ray examination confirmed the opacification of the right hemithorax.

During the intubation attempts, a 2 mm endotracheal tube could not advance beyond the clavicle level after passing the vocal cord and a 6 Fr fiberoptic bronchoscopy could not pass the narrow region of the trachea. Given these results, tracheal stenosis was suspected, and diagnostic evaluation was required. A computed tomography scan of the neck and three-dimensional images at 5 days old showed diffuse tracheal stenosis with a tracheal ring in the distal half of the trachea (Fig. 1A, B). Internal diameter was 1.21 mm at the narrowest point of the stenotic segment. The longest stenosis was approximately 2–3.5 cm long (from the seventh cervical vertebral level to the carina), with two different focal stenoses above the carina and near the right main bronchus (Fig. 1B). The LPA that originated from the posterior aspect of the right pulmonary artery was also detected in the heart CT scan (Fig. 1C). Tracheal stenosis was long in length and within the thoracic cavity, so tracheostomy or tracheoplasty during cardiac bypass surgery were not feasible treatment options.

Fluoroscopically guided balloon dilation was performed using a tracheal balloon 5 mm in diameter and 2 cm in length from the clavicle level to the right main bronchus (Fig. 2). Once dilated, the endotracheal tube could advance to the level of the carina.

Due to restenosis of the trachea just above the carina from previous endotracheal tube dislocation, the tracheal ballooning was required six times before successful extubation. The patient was mechanically ventilated until 30 days old, and was then maintained without ventilator support but with an endotracheal tube as a stent reaching the carina until 7 months old to prevent restenosis of the trachea above of the carina.

The patient was successfully extubated at 7 months old, and chest CT scan was performed 10 days later to assess the degree of improvement in tracheal stenosis (Fig. 3A). The stenosis with

Fig. 2. Fluoroscopically guided balloon dilation was done. (A) Diffuse tracheal stenosis revealed during ballooning. Diffuse tracheal stenosis (block marked) from the level of the clavicle to the carina revealed during ballooning. (B) Focal stenosis near the right main-stem bronchus. Focal stenosis near the right main-stem bronchus and tracheal narrowing was found at the waist of the balloon (an arrow).

Fig. 1. Chest computed tomography (CT) scan at 5 days old showed the diffuse tracheal stenosis and left pulmonary (LPA) sling. (A) Tracheal stenosis on the chest CT scan. Coronal view of the chest CT scan showing the tip of the endotracheal tube (an arrow) reaching the inlet of the thorax and the narrowing lower half of the trachea. (B) Long segmental stenosis on the reconstructed view. Reconstructed view showing the longest stenosis of approximately 2–3.5 cm from the seventh cervical vertebral level (a blue arrow) to the carina (arrowhead), with two focal stenoses above the carina and near the right main bronchus (a red arrow). (C) LPA sling surround the right main bronchus. Axial view showing the LPA (an arrow) branching from the posterior part of the right pulmonary artery and surrounding the right main bronchus. Tracheal stenosis was observed by tracheal compression between the right main pulmonary artery and LPA.

the narrowest diameter was located 2 cm above the carina and had widened from 1.21 mm to 2.55 mm. The patient had no severe respiratory distress and was discharged 31 days post-extubation without additional balloon dilation or ventilation support. At discharge, the weight and height of the infants were less than third percentile in the percentile by age group.

If respiratory distress symptoms appeared after discharge, additional tracheal ballooning was to be considered. However, no additional procedures were performed, as the patient was in good overall condition with the exception of intermittent coughing and mild respiratory distress. Sufficient oral intake was possible via feeding bottle and the patient showed steady weight gain. In the most recent chest CT scan performed at 5 years old (Fig. 3B), the narrowest bronchial diameter located 1.5 cm above the carina was 3.0 mm, approximately 0.5 mm wider than the previous CT scan performed at 7 months old. Pulmonary function tests were not performed. The patient was tested for development at the 37-month-old test. The results showed normal motor skills and cognitive skills but, delayed in the language ability including 21 months for receptive language and an 11 months of age for expressive language. It was possible that the patient had external uterine growth restrictions (weight less than third percentile at discharge) and that he was a premature infant, which may have affected developmental delays. Although he has been hospitalized several times with pneumonia due to viral infection,

but the patient is currently free of complications at the age of 7 without additional procedure for tracheal stenosis during follow-up in an outpatient clinic.

Discussion

Tracheal stenosis is defined as the reduction of luminal diameter by more than 50% of the normal tracheal caliber.⁴ The incidence of congenital tracheal stenosis is estimated to be approximately 2 in 100,000 newborns.⁵

Severe tracheal stenosis can be fatal with respiratory failure occurring shortly after birth; early clinical recognition and treatment at birth improve the prognosis and survival. However, clinical recognition is challenging as chest X-ray findings are non-specific, especially in preterm infants who frequently show signs of respiratory distress.⁶ Common clinical findings in tracheal stenosis include difficulty in intubating and persistent lobar collapse on chest X-ray.^{6,7} In this case, tracheal intubation was difficult and the chest X-ray found pleural effusion. Chest CT scan at 5 days old showed the internal diameter of the trachea at the narrowest point was 1.21 mm, corresponding to an approximately 70% reduction from the average tracheal diameter of 3.75 cm in term babies.⁸

Congenital tracheal stenosis can be caused by external com-

Fig. 3. Chest computed tomography (CT) scan was obtained after extubation. (A) Improved tracheal narrowing on the chest CT scan at 7 months old. At 7 months old the tracheal narrowing (luminal diameter of the narrowest point 2.55 mm) (an arrow) had improved. (B) Improvement on the chest CT scan at 5 years old. At 5 years old, the narrowest bronchial diameter located 1.5 cm above the carina was 3.0 mm, approximately 0.5 mm wider than the previous CT scan performed before discharge.

pression from the surrounding vascular structures or by cartilage rings without the normal posterior membranous wall. Chest CT scan of the patient showed ring shaped attenuation surrounding the tracheal lumen, indicating the presence of cartilage making tracheal rings. LPA sling can cause tracheal stenosis via external compression of the trachea; however, this patient showed long segment tracheal stenosis independently of LPA sling (Fig. 1B). In this patient, a 4 mm endotracheal tube could pass through the dilated trachea to the carina. Balloon dilation for tracheal stenosis in premature infants has been reported in other countries,^{6,9,10} however, most cases involved focal stenosis or required tracheoplasty after ballooning, and one case did not survive. Moreover, the use of an expandable metallic stent after tracheal ballooning has been reported,¹¹ but metallic stents for preterm infants are not available in Korea.

In Loukanov's report,¹ tracheal resection and end-to-end anastomosis was performed in patients with localized congenital tracheal stenosis. However, since the stenosis was not focal in the patient, end-to-end anastomosis was not a feasible treatment option. In this case, we planned the slide tracheoplastic surgery once the appropriate body weight was achieved; however, since the bronchoscopy exam at 7 months old showed no tracheal stenosis except for two small granulomas around the carina, surgery was not performed. Cheng et al.⁸ suggested that conservative management was not suitable for patients with long segment tracheal stenosis or when the tracheal luminal was reduced by more than 40% of the normal tracheal diameter. This patient showed long segment tracheal stenosis with narrowing length of 3 cm, and narrowing of the tracheal lumen to 30% of the average diameter in full-term babies. In this case, the patient was treated with conservative management instead of surgical airway repair by pericardial patch tracheoplasty, tracheal autograft or slide tracheoplasty, after tracheal ballooning and prolonged endotracheal intubation.^{2,3} Similar to the findings of Cheng et al.⁸, tracheal growth was observed in this patient with long segment tracheal stenosis.

To the best of our knowledge, this was the first case of tracheal stenosis treated with tracheal ballooning in a premature infant

in Korea. This case suggests that conservative treatment after tracheal ballooning may be a suitable treatment option even in long segment or severe tracheal stenosis cases in preterm infants.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

References

- 1) Loukanov T, Sebening C, Springer W, Ulmer H, Hagl S. Simultaneous management of congenital tracheal stenosis and cardiac anomalies in infants. *J Thorac Cardiovasc Surg* 2005;130:1537-41.
- 2) Fraga JC, Gabra HO, Calkoen EE, McLaren CA, Roebuck DJ, Elliott MJ. Long segment congenital tracheal stenosis in twins successfully treated by slide tracheoplasty. *J Pediatr Surg* 2009;44:640-3.
- 3) Yang JH, Jun TG, Sung K, Choi JH, Lee YT, Park PW. Repair of long-segment congenital tracheal stenosis. *J Korean Med Sci* 2007;22:491-6.
- 4) Backer CL, Mavroudis C. Congenital Heart Surgery Nomenclature and Database Project: vascular rings, tracheal stenosis, pectus excavatum. *Ann Thorac Surg* 2000;69(4 Suppl):S308-18.
- 5) Krause U, Rödel RM, Paul T. Isolated congenital tracheal stenosis in a preterm newborn. *Eur J Pediatr* 2011;170:1217-21.
- 6) Hauf SM, Perlman JM, Siegel MJ, Muntz HR. Tracheal stenosis in the sick premature infant. Clinical and radiologic features. *Am J Dis Child* 1988; 142:206-9.
- 7) Wolman IJ. Congenital stenosis of the trachea. *Am J Dis Child* 1941;61: 1263-71.
- 8) 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;41:1203-7.
- 9) Hartnick CJ, Anupindi SA, Fan CM, Ryan D, Haver K. Fluoroscopic-guided balloon dilation of tracheal stenosis in a 26-week premature infant. *J Pediatr* 2004;145:566.
- 10) Messineo A, Forte V, Joseph T, Silver MM, Filler RM. The balloon posterior tracheal split: a technique for managing tracheal stenosis in the premature infant. *J Pediatr Surg* 1992;27:1142-4.
- 11) Maeda K, Yasufuku M, Yamamoto T. A new approach to the treatment of congenital tracheal stenosis: balloon tracheoplasty and expandable metallic stenting. *J Pediatr Surg* 2001;36:1646-9.