CASE REPORT / OLGU SUNUMU

DOI: 10.4274/tjccp.galenos.2023.41636
Turk J Clin Cardiov Perfusion 2023;1(3):111-114

Successful Extracorporeal Cardiopulmonary Resuscitation in a Case of Severe Tracheal Stenosis after Congenital Heart Surgery

Konjenital Kalp Cerrahisi Sonrası Ciddi Trakeal Stenozlu Bir Olguda Başarılı Ekstrakorporeal Kardiyopulmoner Resüsitasyon

Hacer Uçmak[1], Fevzi Kahveci[1], Hasan Özen[1], Emrah Gün[1], Edin Botan[1], Anar Gurbanov[1], Burak Balaban[1], Merve Havan[1], Ergun Ergün[2], Nazan Çobanoğlu[4], Zeynep Eyileten[3], Tanıl Kendirli[1]

[1]Ankara University Faculty of Medicine, Department of Child Health and Diseases, Division of Pediatric Intensive Care, Ankara, Turkey
[2]Ankara University Faculty of Medicine, Department of Pediatric Surgery, Ankara, Turkey
[3]Ankara University Faculty of Medicine, Department of Cardiovascular Surgery, Ankara, Turkey
[4]Ankara University Faculty of Medicine, Department of Child Health and Diseases, Division of Pediatric Pulmonology, Ankara, Turkey

Abstract

Extracorporeal cardiopulmonary resuscitation (E-CPR) is a modeling of extracorporeal membrane oxygenation performed during standard cardiopulmonary resuscitation within 5-10 min. Recently, E-CPR use has increased in all age groups. We present a 10-month-old male patient with tracheal stenosis accompanying congenital heart disease who underwent E-CPR because of severe tracheal stenosis. Interestingly, his upper airway anomaly was asymptomatic before heart surgery. His severe tracheal stenosis caused significant respiratory distress and cardiac arrest under mechanical ventilation after early cardiac surgery. We improved E-CPR and then tracheostomy in the after period. Upper airway anomalies are accompanied by congenital heart disease, and they remain asymtomatic until any process to there. E-CPR is a unique and life-saving method for severe and unresponsive cardiac and airway troubles after congenital cardiac surgery.

Keywords: Extracorporeal membrane oxygenation, extracorporeal cardiopulmonary resuscitation, congenital tracheal stenosis, tracheostomy

Öz

Konjenital kalp rahatsızlığına eşlik eden trakeal darlıklı hastada, operasyon sonrası dirençli solunum yetmezliğine bağlı arrest gelişen, ekstrakorporeal kardiyopulmoner resüsitasyon (E-CPR) uygulanarak venoarteriyel ekstrakorporeal membran oksijenasyonu (VA-ECMO) bağlanan hastanın kurtarıcı tedavisini sunulmuştur. Konjenital kalp [Ventriküler septal defekt (VSD) ve patent duktus arteriyozus] nedeniyle takip edilen bir yaş erkek hasta, VSD ve PDA kapatıldıktan sonra takip için çocuk yoğun bakıma (ÇYB) yatırıldı. ÇYB'ye kabulünden sonra mekanik ventilatör (MV) desteği altında yeterince basınç uygulanmasına rağmen ventilasyonu sağlanamıyordu. Ventilasyon sorunlarının bir nedeni de endotrekal tüp (ET) olabileceği düşünülerek tüpü revize edildi. Yaşına göre daha küçük boyutlu ET'yle solunum yolu güvenliği sağlandı. Kalp cerrahisi öncesi hastanın solunum sıkıntısı olmadığı, ancak anestezi uzmanının ET yerleştirmek için tekrarlayan girişimlerinin olduğu öğrenildi. Bronkoskopi yapılarak konjenital trakeal stenozu olduğu izlendi. Yatışının üçüncü gününde zorlu ventilasyon nedeniyle pnömotoraks gelişip göğüs tüpü yerleştirildi. Dördüncü gününde hastanın önce solunum, sonrasında kalp durması nedeniyle 8 dakikalık e-CPR uygulanarak VA-ECMO'ya bağlandı. ECMO kurulumun 14. gününde ekstübe edilerek hastanın solunumu sürekli pozitif nazal hava yolu basıncı (N-CPAP) ile desteklendi. Awake (uyanık) ECMO'yla 2 gün daha takip edilerek dekanüle edildi. Bir ay N-CPAP'la solunum desteği alan, solunum sıkıntısı devam eden hastaya 2-3 haftada bir tekrarlayan tanısal ve terapödik amaçlı bronkoskopi yapıldı. ET yerleştirilmesine sekonder subglottik darlık gelişen hastaya dilatasyon yapıldı. Konjenital trakeal darlığa yönelik operasyona (gelişimi tamamlanıncaya) kadar solunum yolunu güvenliği için hastaya 60. gününde trakeostomi açıldı. ECMO, tedavilere dirençli dolaşım ve/veya solunum yetmezliği olan çocuklarda hayat kurtarıcı müdahaledir.Yaşam beklentisi, nörolojik sağaltımı iyi olacağı öngörülen hasta imkan dahilinde E-CPR sonrasında ECMO bağlanıp organın iş yükü azaltılır. Awake ECMO ile de ekstübe takip edilen hastalarda volu-barotravma, enfeksiyon, ET bağlı solunum yolu hasarları gibi MV bağlı komplikasyon riski en aza indirilebilir. Ayrıca sedatif ve kas gevşetici ihtiyacı da olmayacağı için hasta nörolojik olarak daha iyi değerlendirilip bu ilaçlara bağlı görülebilecek diğer yan etkiler azalacaktır.

Anahtar Kelimeler: Ekstrakorporeal membran oksijenasyonu, ekstrakorporeal kardiyopulmoner resüsitasyon, konjenital trakeal stenoz, trakeostomi

Address for Correspondence/Yazışma Adresi: Hacer Uçmak, Ankara University Faculty of Medicine, Department of Child Health and Diseases, Division of Pediatric Intensive Care, Ankara, Turkey
Phone: +90 555 566 03 09 **E-mail:** prisca.danderfluff@yahoo.com **ORCID ID:** orcid.org/0000-0003-2927-2360
Received/Geliş Tarihi: 03.09.2023 **Accepted/Kabul Tarihi:** 05.12.2023

Introduction

Extracorporeal membrane oxygenation (ECMO) is a temporary support system that maintains life by providing gas exchange and hemodynamic support. ECMO provides ventilation and circulatory functions in cases of respiratory and circulatory failure that is unresponsive to standard therapies. Extracorporeal cardiopulmonary resuscitation (ECPR) is a relatively new and difficult ECMO method that is settled ECMO during unresponsive standard cardiopulmonary resuscitation (CPR) whom there are life expectancy patients (1). For the first time, ECPR was recommended for children in 2020 Pediatric Advanced Life Support. The usage of ECMO in children has been increased and beneficial since the 1970s when ECMO was introduced. This is related to technological improvement and pediatric intensive care unit (PICU) physicians, cardiovascular surgeons, ECMO nurses, and perfusionists' experiments. All ECMO counts, age group distributions, and outcomes have been reported as annually by the Extracorporeal Life Support Organization (2).

Tracheal stenosis is defined as obstruction of the trachea due to a congenital or acquired cause. Congenital causes include conditions such as tracheomalacia, cardiovascular anomalies, and congenital tumors. Acquired tracheal stenosis usually occurs after intubation, and its incidence varies between 0.6-21%. Tracheal stenosis that develops after intubation occurs because of excessive pressure applied by the endotracheal tube (ETT) to the trachea and formation of local ischemic necrosis and granulation tissue in the cartilage tissue. Tracheal stenoses that develop in this manner are usually stenoses at the subglottic level. The longer the intubation period, the higher the risk of developing stenosis; however, this complication can occur even in short-term intubations (3,4).

Here we present a patient who had no respiratory problems in the pre-operative period and was operated for congenital heart disease. He developed severe tracheal stenosis in the early postoperative period and survived with ECPR.

Case Report

A 10-month-old male patient was admitted to the PICU after ventricular septal defect (8 mm) closure and patent ductus arteriosus repair surgery. The patient's aortic cross-clamp and bypass times were 89 and 123 min, respectively. The patient was admitted to the intensive care unit with inotropes of milrinone (0.375 mcg/kg/min) and epinephrine (0.1 mcg/kg/min). It was observed that the lung air inlet and outlet were bad immediately after the patient came to the intensive care unit. ETT tube revision was performed considering that the ETT obstructed the patient whose peripheral oxygen saturation was below 96%. However, it was observed that the age-appropriate ETT (3.5 cuff) could not be placed into the trachea and that the end of the tube was just below the vocal cord. The patient could be intubated with a three-cuffed ETT, and it was learned that he was re-intubated 3 times during the operation. When the patient was admitted to the intensive care unit, blood gases were normal and pressure-controlled SIMV mode was preferred in mechanical ventilation (MV) (rate 24/min, peak inspiratory pressure 24 cm H_2O; positive end-expiratory pressure 6 cm H_2O, fraxioned inspiratory oxygen 40%). During follow-up, the patient, who did not need inotopes and remained respiratory stable under a mechanical ventilator, was extubate on the 1st postoperative day. However, after extubation, the patient's respiratory effort increased, there were significant intercostal and subcostal retractions, bilateral lung sounds were not heard, and he was re-intubated in the 1st hour of extubation. After re-intubation, the patient could not be ventilated and was switched to volume-controlled MV with a tidal volume of 10 mL/kg. On the same day (3rd day of PICU admission), a left pneumothorax was detected and a chest tube was inserted.

Because of recurrent extubation failure, the patient underwent bronchoscopy. We detected a structure completely filling the lumen in the subglottic area, and the diameter of the trachea was severely narrow, and the tracheal mucosa was irregular (Figure 1). Before the upper lobe bronchus separated from the right main bronchus, the the tracheal bronchus was observed, and the bronchus was found to be brochomalasiac. It was observed that the right upper lobe bronchus was two-segmented, and mild malacia was observed in the right middle lobe lateral segment. The bloody secretions was observed in all lower airways from the trachea. During bronchoscopy, granulation tissue was excised and tracheal dilatation was achieved.

On the 4th day of PICU admission, the patient's clinical situation deteriorated and cardiopulmonary arrest developed. CPR was performed for 8 min, and the patient who could not return of spontaneous circulation (ROSC) had difficulty in ventilating. We decided to perform VA ECMO establishing to the neck area. The patient's ROSC (ECPR) time was 55 min. The patient's ECMO settings were rotation per minute 1750/min and liter per minute 500 mL/min. Hemodynamically stabilized in the first 3-5 min of ECMO administration, and his ventilation improved. No abnormality was detected in the blood parameters measured after ECPR. We extubate the patient on the 16th day of ECMO run and placed him on a noninvasive mechanical ventilator (NIMV). The patient was successfully decannulate on day 19th of ECMO under NIVM.

While he was followed up with NIMV support for approximately 1 month, the patient who had respiratory arrest on the 49th day of hospitalization in the intensive care unit was intubated. The patient, whose respiratory distress continued under IMV, was evaluated by ear nose and throat experts, who found no

structural defect in the upper airway. We performed cervical and thoracic computed tomography and found significant narrowing in the trachea (Figure 2). We decided to perform tracheostomy for the prolonged and recurrent upper airway problems, which caused recurrent cardiac arrest. On the 60th day of hospitalization, tracheostomy was performed, and afterwards, he was connected to a home-type mechanical ventilator. On the 95th day of hospitalization, he was transferred to the pediatric chest ward on 95th day of PICU admission with normal vital signs.

In the patient's long follow-up, 13 bronchoscopy-guided dilatation procedures to the subglottic area were performed. The patient's tracheostomy was closed when he was 35 months old, and he is currently being followed as an outpatient without oxygen and any respiratory support.

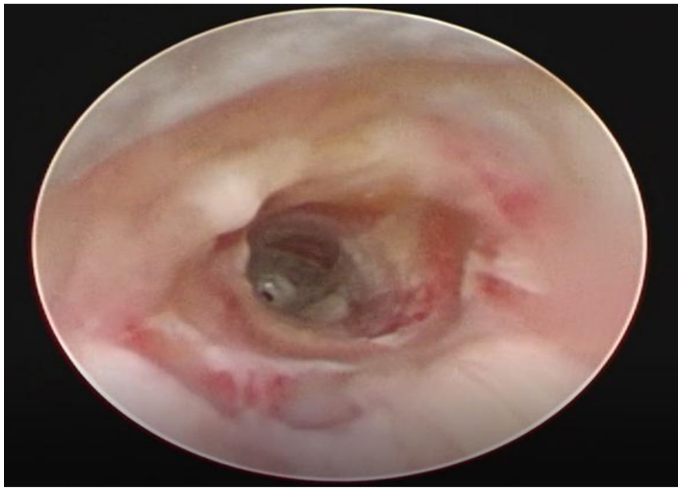

Figure 1. Patient's bronchoscopic image. There are severe narrowed trachea and fibrin covered mucosa due to severe tracheal stenosis

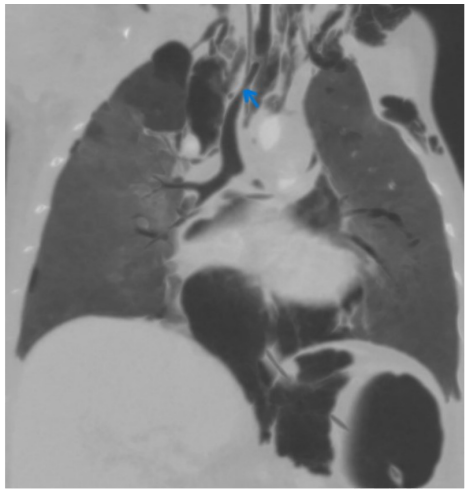

Figure 2. Patient's cervical and thorax computed tomography image. Long and severe narrowed trachea is seen on longitudinal section and it is marked by arrow

Discussion

Tracheal stenoses are airway anomalies with an incidence of 1/50,000 and are classified as acquired or congenital, extrinsic or intrinsic, short segment or long segment stenoses. Congenital tracheal stenoses, type I; long segment stenosis, most or all trachea stenotic. Type II: Funnel-shaped stenosis of different lengths and localization. Type III: Short segment stenosis occasionally observed under an abnormal right upper lobe bronchus. Type IV: It is classified as an abnormal right upper lobe bronchus and a bronchus extending horizontally to the rest of the lung (5,6). Because catch-up growth has been observed in stenotic tracheas over the years, conservative approaches are applied in mild cases and surgical procedures are applied according to the narrow segment length in symptomatic cases (resection/anastomosis, slide/patch tracheoplasty). Postintubation tracheal stenosis was first described by Cooper and Grillo (7). and 90% of acquired tracheal stenoses are caused by damage caused by the cuff pressure of the intubation tube. With the application of excessive pressure to the trachea by the intubation tube cuff, initially simple edema develops, followed by mucosal ulceration, perichondritis, and granulation tissue stenosis, which occurs with local ischemic necrosis of the tracheal cartilage (6-8). While stenoses are mostly observed in the subglottic region in infants and young children, the lesion is usually located in the trachea in older children (9). In a series published by Maeda et al. (9), acquired and congenital mixed subglottic stenosis was found in 4 cases, and subglottic stenosis due to laryngeal trauma was found in 1 case (10). Our case also had congenital stenosis, and repeated unsuccessful extubation attempts led to tracheal injury and then stenosis. The patient was followed up with V-A ECMO support until lung ventilation improved. In addition, the presence of the right tracheal bronchus detected by thorax CT was compatible with type 3 tracheal stenosis. A history of unsuccessful extubation of intubated cases or the presence of respiratory findings approximately 2 months after extubation suggest stenosis. Grillo stated that the risk increased in patients who were intubated for longer than 48 h and had symptoms before 2 years (5).

Isolated congenital tracheal stenoses nda in 10-30% of patients. It is often associated with other extrathoracic and cardiovascular anomalies. Cardiovascular anomalies are seen in 70% of patients nda re accompanied by pulmonary artery sling, patent ductus arteriosus, atrial septal defect, ventricular septal defect, double aortic arch, partial pulmonary venous return anomaly, Fallot tetralogy, and tricuspid atresia (10). Extrathoracic anomalies include gastrointestinal, renal, and skeletal anomalies (3). Our patient had a cardiological ventricular septal defect and patent ductus arteriosus.

ECPR is most often activated in children with heart disease and in hospital cardiac arrest when cardiac function does not improve within 5-10 min of conventional CPR. The ideal activation time of ECPR has not been precisely defined. There is increasing interest in reducing exposure to narcotic, sedative, and neuromuscular blocking agents and keeping ECMO patients alert and active to maintain muscle strength and shorten recovery time. Recently, the method of extubation from MV has emerged, especially for patients with respiratory failure and for patients followed under ECMO as a bridge therapy to lung transplantation (11-14). However, extubation management option notification during ECMO in PICU's is not sufficient. Our patient, who had cardiac arrest after respiratory arrest on the 4th day of PICU admission, was taken to ECMO via E-CPR. He was followed up intubated under ECMO for approximately 16 days, then extubate after ensuring his alertness and good muscle strength, and finally decannulate after being followed up with awake ECMO for 3 days. The first-line examination in imaging methods for airway stenosis is anteroposterior chest and lateral neck X-ray. After detecting tracheal stenosis by radiography, tomography and magnetic resonance imaging are among the tests used to confirm the diagnosis. Evaluation with bronchoscopy can give an idea about the degree of stenosis. Endoscopic interventions provide information regarding the type of lesion, location of stenosis or erosion, and accompanying tracheal anomalies. The treatment of tracheal stenosis after intubation is surgical, with a success rate of 95%. If the degree of stenosis is severe, time can be gained for dilatation by opening a temporary tracheostomy. In our case, due to severe tracheal stenosis, the granulation tissues were excised; however, because the stent was not placed in the congenital stenosis area in children, after the development of the tissues was achieved, the stenosis area was excised and end-to-end anastomosis was performed. It was decided that this could be done. Temporary tracheostomy was performed on the patient to prevent possible respiratory problems during this period.

In conclusion, our patient, who was initially asymptomatic with congenital tracheal stenosis accompanying congenital heart, had symptoms that emerged after the heart surgery because of an intubation attempt for cardiac surgery. Successful E-CPR was performed because of difficult airway-associated untreatable respiratory troubles and recurrent cardiac arrest. In order to prevent possible barotrauma/volutrauma of MV in mostly adult patients, awake ECMO, which allows extubated follow-up while under ECMO run, was successfully applied in our case. Temporary tracheostomy was performed in our case in order to allow time for the development of tissues.

Ethic

Informed Consent: Consent was obtained from the family for this case report.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: E.E., Z.E., N.Ç., Concept: T.K., Design: F.K., Data Collection or Processing: H.U., E.G., E.B., Analysis or Interpretation: H.U., H.Ö., A.G., Literature Search: H.U., B.B., M.H., Writing: H.U.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

References

1. Wyckoff MH, Greif R, Morley PT, Ng KC, Olasveengen TM, Singletary EM, et al. 2022 International Consensus on Cardiopulmonary Resuscitation and Emergency Cardiovascular Care Science With Treatment Recommendations: Summary From the Basic Life Support; Advanced Life Support; Pediatric Life Support; Neonatal Life Support; Education, Implementation, and Teams; and First Aid Task Forces. Resuscitation 2022;181:208-288.
2. The Extracorporeal Life Support Organization ECLS 'International Summary of Statistics. https://www.elso.org/registry/internationalsummaryandreports.aspx
3. Grillo HC, Donahue DM, Mathisen DJ, Wain JC, Wright CD. Postintubation tracheal stenosis. Treatment and results. J Thorac Cardiovasc Surg 1995;109(3):486-492.
4. Stauffer JL, Olson DE, Petty TL. Complications and consequences of endotracheal intubation and tracheotomy. A prospective study of 150 critically ill adult patients. Am J Med 1981;70(1):65-76.
5. Grillo HC. Congenital and acquired tracheal lesions in children. In: Grillo HC (eds) Surgery of the trachea and bronchi. BC Decker Inc, Ontario, 2004:174-178.
6. Cantrell JR, Guild HG. Congenital Stenosis of the Trachea. Am J Surg 1964;108:297-305.
7. Cooper JD, Grillo HC. The evolution of tracheal injury due to ventilatory assistance through cuffed tubes: a pathologic study. Ann Surg 1969;169(3):334-348.
8. Cotton RT. Prevention and management of laryngeal stenosis in infants and children. J Pediatr Surg 1985;20(6):845-851.
9. Maeda K, Ono S, Baba K. Management of laryngotracheal stenosis in infants and children: the role of re-do surgery in cases of severe subglottic stenosis. Pediatr Surg Int 2013;29(10):1001-1006.
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):527-536.
11. Thiagarajan RR. Extracorporeal Membrane Oxygenation for Cardiac Indications in Children. Pediatr Crit Care Med 2016;17(8 Suppl 1):S155-S159.
12. Nosotti M, Rosso L, Tosi D, Palleschi A, Mendogni P, Nataloni IF, et al. Extracorporeal membrane oxygenation with spontaneous breathing as a bridge to lung transplantation. Interact Cardiovasc Thorac Surg 2013;16(1):55-59.
13. Mohite PN, Sabashnikov A, Reed A, Saez DG, Patil NP, Popov AF, et al. Extracorporeal Life Support in "Awake" Patients as a Bridge to Lung Transplant. Thorac Cardiovasc Surg 2015;63(8):699-705.
14. Olsson KM, Simon A, Strueber M, Hadem J, Wiesner O, Gottlieb J, Fuehner T, Fischer S, Warnecke G, Kühn C, Haverich A, Welte T, Hoeper MM. Extracorporeal membrane oxygenation in nonintubated patients as bridge to lung transplantation. Am J Transplant 2010;10(9):2173-2178.