Aortopexy for Tracheomalacia Via Partial Sternotomy

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This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

ABSTRACT

Tracheomalacia (TM) is a disease that causes the airway obstruction of the tracheal lumen as a result of the structural disorder of the tracheal cartilage. We present a 4-month-old patient who developed ventilator depended TM after repair of esophageal atresia with tracheoesophageal fistula. Aortopexy and intraoperative flexible bronchoscopy were first performed via partial sternotomy in Turkey for this patient. He was weaned from ventilatory support and extubated at the first and discharged at the 8th post-operative day. Partial sternotomy is performed in the supine position, thus it allows for intraoperative flexible bronchoscopy permitting to check for the adequacy of the aortopexy.

Keywords: Tracheomalacia; aortopexy; esophageal atresia; partial sternotomy; flexible bronchoscope.
1. INTRODUCTION

Tracheomalacia (TM) is the weakness of the tracheal lumen resulting in anteroposterior collapse during expiration or coughing. This situation may effect the trachea locally or totally\[1-3\]. While TM is mostly associated with oesophageal atresia (EA) with tracheo-oesophageal fistula (TOF), vascular ring and innominate artery compression, pectus excavatum or large vessel anomalies, it can also be seen after prolonged intubation or tracheostomy[1,4-6]. Clinical symptoms include recurrent pneumonia, stridor, failure to wean from ventilation, and life-threatening conditions such as apnea and cyanotic ‘death’ spell[1-3]. With this case report, we wanted to share our experience in an infant with TM who could not be weaned from mechanical ventilation after EA+TOF repair. This infant is the first reported case of successful aortopexy done via partial sternotomy (PS) in Turkey. To date, an article describing aortopexy for TM has not been found in the Turkish Medical Index.

2. CASE PRESENTATION

A 4-month-old male baby weighing 4.1 kg was referred to our pediatric surgical department for suspected TM. Baby had been operated by our pediatric surgical team at the first day of life for EA + TOF repair and multiple trials to extubate the child had failed (due to respiratory distress, apnea and deterioration of the general condition, he was always reintubated) and as a result he had become ventilator dependent. Computed tomography (CT) scan of the thorax showed consolidation in the lung involving the apical segments of right upper lobe and demonstrated that the lower trachea above the carina was obliterated. It was decided to perform flexible bronchoscopy (FB) after weaning in the operation theater. He was operated at the 123th day of life for TM. The patient was already intubated with 3.5 French uncuffed tracheal tube. Spontaneous breathing efforts of the patient were not suppressed for the dynamic evaluation of TM. First, FB was inserted into the trachea through the tracheal tube and oxygenation was achieved via FB. The intubation tube was then pulled over the bronchoscope allowing the patient to be extubated for the dynamic evaluation of the trachea. While spontaneous breathing movements were controlled with FB, anterior tracheal wall near the carina was observed to collapse with exhalation (Fig.1). FB revealed significant TM. No significant residual pouch or diverticulum at the right posterior trachea related to previous TOF repair was detected. Therefore, aortopexy was planned in this same session and the tracheal tube was reinserted. A suprasternal skin crease incision was made and a 3 cm partial sternotomy (PS) was performed and partial thymectomy was done to expose the upper part of the pericardium, the innominate artery and the aorta. Etibond and polypropylene sutures(5/0) were placed between the anterior wall of the aorta at the innominate artery level and posterior sternal fascia (Fig.2). After the sutures were tied a last peroperative control of the trachea was done via FB to control the adequacy of aortopexy. Trachea was seen not to collapse(Fig.3). Sternal edges were approximated by 3/0 polypropylene sutures and the wound was closed. He was weaned the intubation within the first postoperative day without oxygen requirement. All the respiratory problems of the patient improved and the life-threatening conditions such as apnea and respiratory distress were not observed after aortopexy and the child was discharged from the hospital at the 8th post-operative day. There was no problem in 18 months of follow-up period.

![Fig. 1. The tracheal wall was seen to collapse with exhalation during preoperative flexible bronchoscope](image-url)
Fig. 2. Partial sternotomy and sternal sutures

Fig. 3. The tracheal wall was seen not to collapse after aortopexy during intraoperative flexible bronchoscopy

3. DISCUSSION

The incidence of TM is 1: 2100. Primary or congenital TM typically occurs in prematures and heals spontaneously up to 1 year of age [7]. While over 90% of children with EA + TOF have various forms of TM, 2-25% of them require aortopexy [8,9]. Severe TM is seen in 11–33% of EAs, while TM is more common in patients with EA + TOF than isolated EA [3,8,10].

Pathology in TM is due to congenital weakness of the tracheal wall, elliptical deformity and congenital disorder of the cartilage rings, the muscle length and width of the posterior wall increase and the ratio of the cartilage trachea to
membranous trachea below 4.5: 1 and the trachea from “C” form to “U” form. As a result, the posterior membrane enters the tracheal lumen during expiration, causing airway collapse[3,10,11]. In mebranous trachea of children with EA, 80% esophageal muscle and squamous epithelium were observed. This rate is only 2% in normal children [12]. In addition; dilatation of the proximal esophageal pouch compresses the trachea in utero and alters its normal development [13]. The association of TM and gastro esophageal reflux (GER) has been shown in 59-63% of EA + TOF patients. Both conditions may worsen the symptoms of the other. GER worsens the clinic with airway obstruction, vagal reflex and upper esophageal pouch distension. However, there is no consensus on the time of fundoplication [1,14].

Vocal cord dysfunction, laryngeal clefts, tracheal diverticulum, tracheal septonosis and rings, recurrent TOF, intrathoracic vascular anomalies, lung parenchymal disease, esophageal strictures, esophageal dysmotility, GER, neurologic and cardiac conditions should be excluded in the differential diagnosis [5].

Multidetector CT shows in detail 2 and 3 dimensional anatomical imaging of all thoracic structures [3]. In the diagnosis of TM, vocal cord dysfunction, laryngeal clefts and TEF, FB has the advantages of direct visual inspection and dynamic evaluation. In addition, FB causes less damage to the airway and is more comfortable than rigid bronchoscopy or endotracheal intubation [2,3,6,15].

With the growing of the child and the cartilage becoming harder, TM resolves spontaneously until 1-2 years and surgery is not required in 70% of cases [1-3,16]. Hypertonic saline nebulizers, inhaled ipratropium bromide, low-dose inhaled steroids can be used to treatment for mild and moderate TM. They help to airway problems by softening, thining secretions, and reducing inflammation [17]. However, aortopexy is required in 30% of patients due to serious life-threatening symptoms [15]. The main indications for aortopexy are dying spells, recurrent pneumonia, airway obstruction and inability to extubate. The main indications for aortopexy are dying spells, recurrent pneumonia, airway obstruction and extubation. Among these symptoms, the most serious indication for aortopexy is “dying spell”. These patients should be kept in the hospital until they are treated [2,3,15,16].

Although many approaches have been tried so far, 72% of surgeons preferred left anterior thoracotomy and the rest 28% used other approaches [2]. Aortopexy has been the mainstay of surgical therapy for pediatric intrathoracic TM. The vessels and other structures on the airway move forward anteriorly with the removal of thymus and aortopexy. After this procedure, Thus, the trachea is indirectly displaced forward and tracheal lumen is always kept open even though posterior tracheal membrane intrusion during expiration [1-3,15,16]. External splinting shoul be preferred with autologous and prosthetic materials in cases with very long segment TM or in cases where aortopexy is unsuccessful [5].

While more than 80% of the patients recovered after aortopexy, 4% of the patients had worsening symptoms and 6% died [2]. Although similar results were obtained with thoracoscopic techniques, operation time was longer in this method than open surgery [17]. The difficulty in lateral approaches, whether with thoracoscopy or thoracotomy, is that the surgeon has to approach midline structures from one side and, when necessary, pull them to reach opposite side structures [6,15].

4. CONCLUSIÓN

PS allows direct viewing of the aorta, other big vascular structures, the pretracheal fascia and the trachea. With the direct viewing of the vascular structures, more secure placement and direct hanging of the aorta without angulation according aortopexies performed by right or left thoracotomy can be made with more effective elevation. PS is performed in the supine position, it permits intraoperative FB and this shows the adequacy of the aortopexy or distortion of the trachea. It also provides easy access to large vessels if bleeding occurs.

DISCLAIMER

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CONSENT

As per international standard or university standard guideline informed consent has been collected and preserved by the authors.
ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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