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Case Report

Endoscopic Repair of Refractory Tracheoesophageal Fistula with a Cardiac Septal Occluder Device in a 12 Years-Old Patient

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Abstract

Purpose: Esophageal atresia (EA) and tracheoesophageal fistula (TEF) are rare anomalies in neonates that must be surgically repaired by esophageal reconstruction with or without ligation of the fistula. Recurrent tracheoesophageal fistula (rTEF) occurs in 3-15% of primary surgical repairs in esophageal atresia; it is associated with recurrent hospital admissions and up to 27% short term mortality. Dependable reparation very often proves difficult by standard surgical techniques. Using oesophageal fully covered self-expandable metal stents in adult patients yields a <50% efficacy and other endoscopic techniques such as occlusion by clips or glue show no better results. A minimally invasive alternative is the use of vascular plug septal occluders. We report the efficacy of endoscopic placement of a cardiac septal occluder (CSO) in a paediatric patient. Clinical case: A 12-year-old female with recurrent (rTEF) and refractory tracheoesophageal fistula (refTEF) was subjected to an refTEF closure procedure via endoscopic placement of a cardiac septal occlusion device. Conclusion: Debate regarding the gold standard of rTEF treatment closure a hot debate but flexible endoscopy is an accepted alternative. This report describes the successful fixing of a refTEF using a cardiac septal occluder. After four weeks follow up, no re-incidence of the tracheoesophageal fistula was detected. The results advocate for the endoscopic closure of refractory tracheoesophageal fistula with cardiac occluders in children thus establishing a promising therapeutical alternative in refTEF in paediatric population patients.

Keywords

Tracheoesophageal Fistula, Recurrent Tracheoesophageal Fistula, Refractory Tracheoesophageal Fistula, Endoscopic Interventional Therapies, Treatment, Children, Cardiac Septal Occluder

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1. Introduction

Recurrent tracheoesophageal fistula (rTEF) is not an uncommon complication after surgical repair of congenital tracheoesophageal fistula (TEF). The recurrence of esophageal fistula occur in 3-15% of patients and its surgical management is associated with recurrent hospital admissions, high morbidity and a mortality of 27% [1, 2].

Numerous non-surgical techniques are used for rTEF closure, but the most common are injection of sealant into the fistula tract, de-epithelialization with diathermy, or a combination of both [3, 4] as well as Chemocauterization with brushing and application of trichloroacetic acid [5]. The surgical technique for rTEF closure is based on the application of patches or flaps between the esophagus and trachea although not always with optimal results [6, 7]. But when an rTEF was treated by endoscopy and becomes patent again, the degree of complexity for its closure increases considerably either by endoscopy or even more by surgery. Therefore, at this time the fistula should be named call it a "Refractory tracheoesophageal fistula" (refTEF). A minimally invasive alternative consists of the endoscopic placement of a cardiac septal occluder (CSO) [8, 9]. The purpose of this report is to demonstrate that refTEF closure can be achieved by the endoscopic placement of a cardiac septal occluder in a paediatric patient with refractory TEF.

2. Clinical Case

A 12-year-old female diagnosed with atresia type III of the Gross classification at birth was surgically intervened at 24 hours of life for oesophageal plasty and closure of TEF. At the age of 6 months, she presented with bronchospasm and repeated pneumonias so a digestive and respiratory endoscopy was performed. The result reported a rTEF of 2mm in diameter at 3cm from the carina which was resolved by stamm type gastrostomy for exclusive enteral feeding. Two endoscopic de-epithelialization sessions were performed with argon plasma + fibrin sealant to close the rTEF. The esophagogram corroborated the closure of the rTEF. The patient remained asymptomatic for a period of 8 years. At the age of 12 years, she began with recurrent pneumonias again; a new endoscopic and fluoroscopic study demonstrated a refractory tracheoesophageal fistula of 2 mm in diameter by 2.3 mm in length twenty cm from the dental arch in (Figure 1 and 2a, 2b). Following interdisciplinary discussions refTEF closure by cardiac occluder was considered as the best minimally invasive and non-surgical option for the patient. After explaining the benefits and risks of this procedure, informed consent was obtained from the parents.

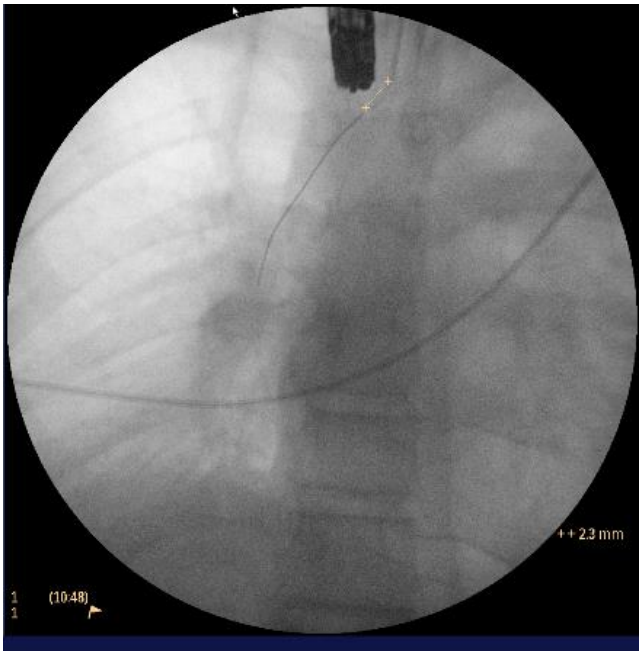

Figure 1. *Fistula length by fluoroscopy.*

3. Endoscopic Procedure

Under general anaesthesia, a bronchoscope (Fuji EB-530T OD5.8mm/WCh:2.8mm) was inserted through the orotracheal cannula, FTE was cannulated with 0.035 inches in diameter hydrophilic Jagwire™ guidewire from the trachea to the esophagus. One end of the guidewire was retrieved through the endoscope (Fuji EG-530FP OD8.5mm/WCh:2.8mm) inserted through the esophagus, with a foreign body forceps (Figure 2).

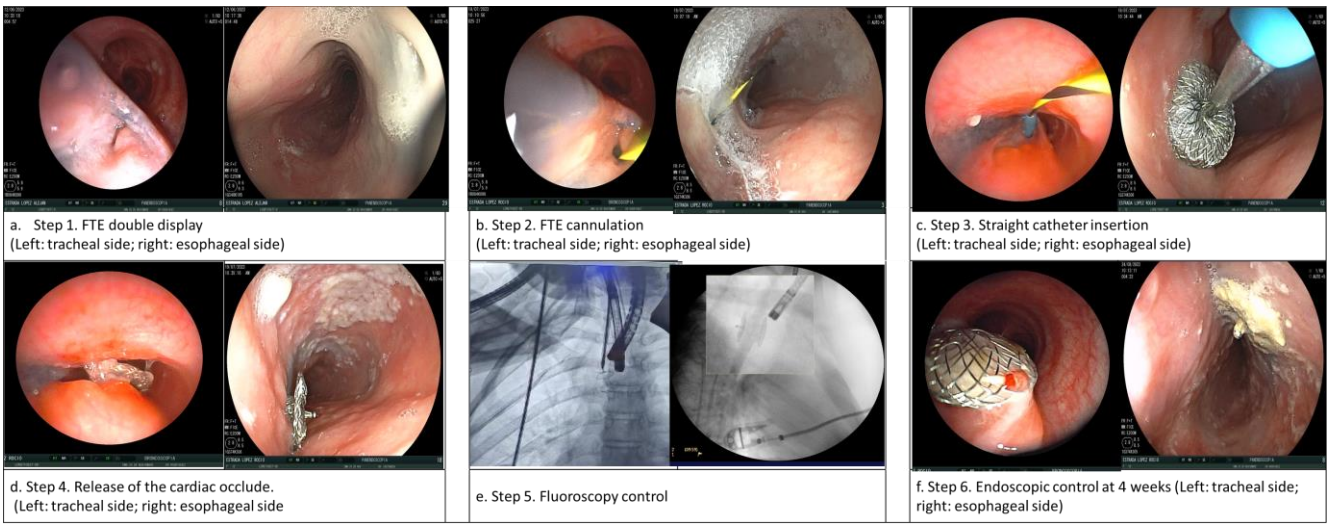

Figure 2. *Endoscopic procedure. Placement of cardiac septal occluder step by step.*

The double endoscopic approach was maintained and under fluoroscopic vision, a 6 fr catheter was introduced through the working channel of the gastroscope positioning the occlutech mVSD Occluder™ (Figure 2). The first disc of the device was released into the tracheal lumen and the second disc was subsequently released into the oesophageal lumen. The patient remained asymptomatic and a radiographic control which showed an adequate esophageal and tracheal lumen (Figure 3), the patient was discharged. The successful occlusion of the defect was confirmed 4 weeks later by an esophagogram that corroborated the absence of contrast medium passage to the tracheobronchial tree; the adequate position of the device was also verified by endoscopy (Figure 2) and no fistula was visible on either side. The patient has been followed monthly for a year, oral liquids are well tolerated and oral food intake has been resumed (Figure 4).

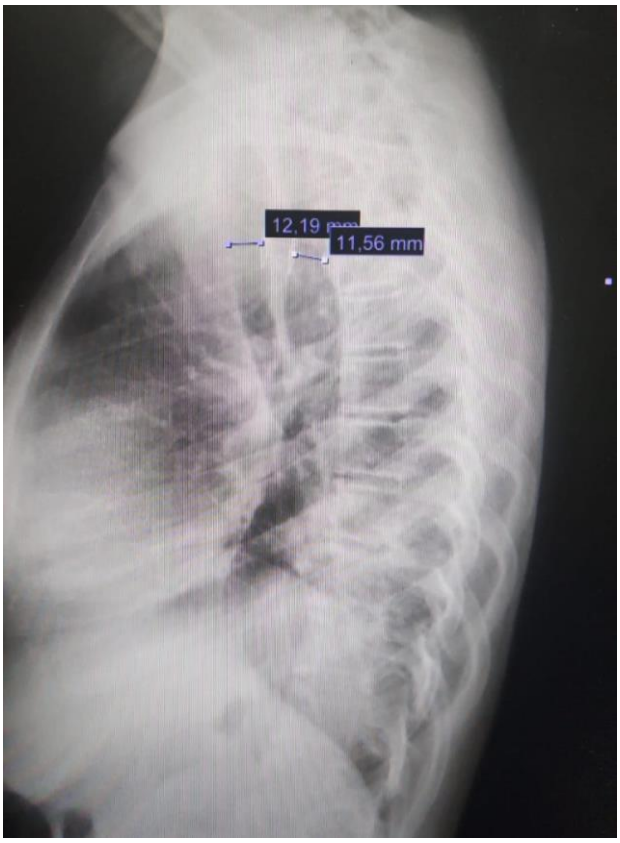

Figure 3. *Lateral chest x-ray with measurement of the tracheal lumen 12.19mm and esophageal lumen 11.56mm, cardiac occluder in adequate position.*

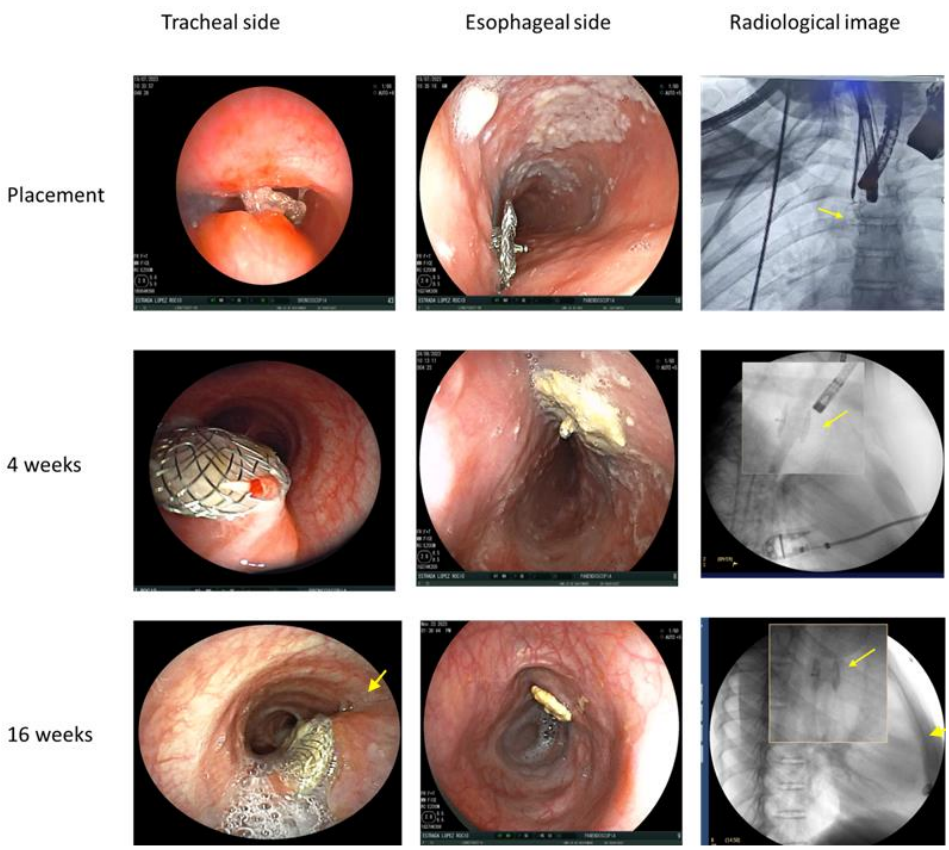

Figure 4. *Endoscopic and fluoroscopic follow-up of the patient. Healing tissue is observed at week 16 (see arrow). Radiological images show the proper position of the cardiac occluder (see arrow).*

4. Discussion

Esophageal atresia, caused by incomplete embryonic compartmentalization of the foregut commonly occurs with a tracheoesophageal fistula [9]. It is thought-provoking to keep in mind that esophageal malformations are the product of abnormalities during embryogenesis where members of the forkhead-box family of transcription factors, as well as the SRY-box transcription factor SOX2, are preferentially expressed in embryonic fibroblast the former and epithelial subpopulations the latter [10, 11]. The Fox family of transcription factor are up-regulated in patients with tracheoesophageal fistulas [12] but transcription factor SOX2 is a vital regulator of stem cell activity in developing human tissues [13] and its mutation has been associated to tracheoesophageal fistula [14]. Therefore, the relevance of the cellular mechanisms associated with TEF development must be kept present as the numerous include the so-called epithelial-mesenchymal transition that is pivotal in various gastrointestinal cancers [15].

TEFs are difficult to manage in children due to the anatomical and surgical complexity as well as associated comorbidities such as cardiac anomalies, renal anomalies, or vertebral anomalies [16] and the biopsychosocial impact of the inability to feed orally. After primary repair of TEF, 5-10% of cases recur [5]. Surgical reintervention in children with TEFs is associated with a high rate of adverse events [6] so endoscopic intervention: (endotracheal stent, de-epithelialization with diathermy -laser, argon plasma -, chemical sealant with cyanoacrylate or trichloroacetic acid, fibrin glue, or the combination of both) [17, 24], represents a less invasive option with low mortality but highly variable success rate [18, 19].

The decision to use a cardiac occluder device was based on its success in adult population [7, 20] where technical and clinical success is 100% and 92%, respectively [21]. This device produces an inflammatory response that favours closure of the defect [8], preserves airway patency, prevents pneumonia due to the abnormal communication between esophagus and trachea, and allows oral feeding. Nevertheless, mucus retention and formation of granulation tissue must be frequently evaluated [16]. The Occlutech mVSD occluder that we used has a biocompatible surface made of Nitinol, a nickel-titanium alloy, wire that decreases the amount of granulation tissue. The application of Nitinol occluders generates a granulated reaction with less inflammatory and more fibrous tissue 3 month after implantation [22] thus suggesting and beneficial immune response. A recent report suggests the use of a CT scan with holographic 3D reconstruction of the fistula in order to improve the coverage of the fistula margins [23]. Although the application of this device in paediatric patients is endorsed by the

present report larger multicentre studies are necessary to improve the method and patient selection.

5. Conclusion

TEF closure using a cardiac septal occluder is a feasible, safe and effective modality for the endoscopic closure of refractory TEFs in children. Further studies are needed to launch such a promising therapeutical alternative in paediatric patients. The nomenclature in the surgical/endoscopic treatment of TEFs should be reconsidered as there is a significant clinical difference between the approach of a recurrent versus a refractory TEF as the latter implies at least two previous closure attempts, either surgical and/or endoscopic, that imply a treatment challenge.

Abbreviations

rTEF: Recurrent Tracheoesophageal Fistula
TEF: Tracheoesophageal Fistula
refTEF: Refractory Tracheoesophageal Fistula
CSO: Cardiac Septal Occluder

Conflicts of Interest

The authors declare no conflicts of interest.

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