Pulmonary vein isolation in a patient with achalasia and megaesophagus

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Introduction
Atrioesophageal fistula (AEF) is a rare but life-threatening complication of catheter ablation of atrial fibrillation (AF), thought to occur in as many as 0.03%–0.08% of cases. It is not known if patients with existing esophageal disorders, particularly those that dilate the esophagus, are at higher risk of AEF after pulmonary vein isolation (PVI). Achalasia is an inflammatory and neurodegenerative disorder of the esophagus characterized by increased tone and failure of relaxation of the lower esophageal sphincter as well as absent peristalsis of the esophageal body, often leading to progressive esophageal dilation and megaesophagus.

Here we present a case of a 73-year-old man with symptomatic paroxysmal AF and achalasia and marked esophageal dilation despite Heller myotomy who underwent PVI with Cryoballoon (Medtronic, Minneapolis, MN), pharmacologic esophageal protection, and a multisensor esophageal temperature (ET) monitor.

Case report
A 73-year-old man with a history of hypertension and chronic kidney disease was referred to the electrophysiology clinic for management of highly symptomatic paroxysmal AF that he developed 6 months after catheter ablation of cavitricuspid isthmus–dependent atrial flutter. The patient had a 10-year history of dysphagia and food regurgitation.

A diagnosis of achalasia was confirmed by esophageal manometry, demonstrating absent peristalsis and elevated esophageal relaxation pressure, and barium swallow, demonstrating a dilated esophagus with delayed esophageal emptying. He had undergone serial endoscopic esophageal dilations with only partial and transient relief of his symptoms. Six months prior, he underwent a laparoscopic Heller myotomy with simultaneous Dor fundoplication with significant improvement of his symptoms and esophageal size.

The patient’s AF had remained markedly symptomatic despite rate control with diltiazem. Flecainide had previously resulted in marked QRS widening, and at the time of referral he was being treated with amiodarone, despite which he still had paroxysms of AF.

Preprocedure computed tomography pulmonary venogram revealed a dilated and debris-filled esophagus measuring approximately 4.8 × 3.3 cm at the level of the left atrium (LA) with abutment and mass effect onto the LA (Figure 1). The anterior esophageal wall spanned the width of the posterior LA wall as well as the right pulmonary veins more than the left pulmonary veins. The patient’s gastroenterology and surgery teams were consulted and felt that his achalasia had been maximally treated, and his esophagus was unlikely to reduce any further in size with additional medications or interventions. The risks and benefits of catheter ablation were discussed with the patient, who agreed to proceed.

KEYTEACHING POINTS

- Pulmonary vein isolation is a procedure increasingly utilized as a rhythm control strategy in atrial fibrillation. Although this procedure is often successful at achieving a long period of arrhythmia-free survival, there are attendant risks, one of the most devastating of which is the development of a left atrioesophageal fistula.
- The risk for esophageal injury with catheter ablation for atrial fibrillation is likely elevated in patients with underlying structural anomaly in the esophagus.
- Risk for esophageal injury may be mitigated with a multimodal approach including multisensor esophageal temperature monitoring, conservative nadir esophageal temperature thresholds, and pharmacologic esophageal protection.
Given concern for esophageal injury, a Cryoballoon approach was chosen with a prespecified plan to discontinue ablation for esophageal cooling, based on prior studies routinely performing endoscopy post Cryoballoon ablation. The patient was continued on his omeprazole 20 mg by mouth twice a day and was prescribed sucralfate 1 g by mouth twice a day for 2 weeks before and after the ablation. Additionally, he received intravenous famotidine 20 mg at the start of the procedure. ET was monitored using a multisensor S-shaped ET probe (CIRCA S-CATH; CIRCA Scientific, Englewood, CO). Following administration of general anesthesia with endotracheal intubation, the ET probe was inserted and positioned under fluoroscopic guidance so that all 12 thermocouples spanned the LA, and esophageal position was consistent with computed tomography findings such that the probe lay more behind the right pulmonary veins (Figure 2). Following transseptal puncture and access to the LA, a Cryoballoon was advanced into the LA. Cryoballoon ablation of the LPVs (180 seconds × 2 applications for each vein) did not result in significant decrease in ET on any of the 12 sensors of the probe, and achieved entrance and exit block. Attention was turned to the right-sided pulmonary veins. Esophageal cooling was not noted during ablation of the right superior pulmonary vein (180 seconds × 1 application). Entrance and exit block was achieved on the first ablation, and a second application was not performed given proximity to the esophagus; time to isolation <30 seconds, -30°C achieved at 24 seconds, and thaw time of 13 seconds of the first application. During ablation of the right inferior pulmonary vein, ET decreased to a nadir of 30.2°C during the first 180 seconds of cryoablation (did not require premature termination), and subsequently dropped as low as 27.0°C during thaw phase. Entrance and exit block was also achieved on the first application, so a second ablation was not performed given esophageal cooling and similar cryoablation parameters to the right superior pulmonary vein. Entrance and exit block was confirmed for all 4 veins and there were no complications.

The patient was discharged the day after his procedure in normal sinus rhythm on amiodarone, diltiazem, rivaroxaban, and 2 more weeks of omeprazole and sucralfate. A postablation esophagogastrroduodenoscopy was not performed given the potential risks of esophageal instrumentation in the setting of achalasia, combined with the lack of fever, chills, focal neurologic symptoms, chest pain, or worsening dysphagia suggestive of AEF. Four weeks post procedure, he remained in sinus rhythm and had not had any AF-attributable symptoms, so his amiodarone was discontinued. Six months post procedure, he remains free of AF.

**Discussion**

One of the most feared and devastating complications of AF ablation is the development of an AEF, which is thought to arise in about 0.03%–0.08% of all cases. Although rare, it is of particular relevance owing to the high associated mortality rate—up to 55%. Achalasia may dispose towards a higher risk for such complications. With long-standing achalasia, the ultrastructure and histologic appearance of the esophagus becomes progressively deranged, with inflammation, significant atrophy of the muscle, and fibrosis. This may increase susceptibility to mechanical injury, although direct data on this topic are lacking. Achalasia may also be associated with increased risk for esophageal injury simply owing to proximity of the dilated esophagus to the LA, which is an independent predictor of esophageal injury in cardiac ablation. We present a case of a high-risk PVI in a patient with surgically repaired achalasia but a persistently enlarged esophagus.
esophagus where we attempted to mitigate risk with (1) use of Cryoballoon with conservative thresholds for nadir ET, (2) pharmacologic esophageal protection, and (3) multisensor ET monitoring.

While data exist demonstrating similar efficacy and overall complication rate between Cryoballoon ablation and radiofrequency (RF) ablation, direct comparisons between the 2 pertaining to esophageal injury and AEF are scarce. A study utilizing the MAUDE database suggests a remarkably low incidence of <1 in 10,000 for Cryoballoon-associated AEF, but may be limited by an underestimate owing to under-reporting. Owing to low incidence of AEF, its risk predictors are not well characterized, and are often extrapolated from studies of endoscopically-detected esophageal injury, which is not definitively a surrogate or precursor for AEF. A study of RF PVI patients undergoing postprocedure endoscopy demonstrated an incidence of 47% for any esophageal injury (including mild changes) and 18% for necrotic and ulcer-like changes. A similar study of Cryoballoon PVI demonstrated esophageal ulcerations in 17% of cases. We ultimately chose Cryoballoon for our procedure given the low incidence of AEF in the MAUDE database.

Although there is interest currently in intraprocedure ET monitoring to minimize the risk to the esophagus and help guide the creation of safer lesions, a recent meta-analysis of the available trials of ET monitoring did not demonstrate significant differences in the incidence of esophageal injury between the monitored and unmonitored groups. However, a significant association has been noted between nadir ET and incidence of esophageal injury as defined by esophageal lesions noted on postablation endoscopy. Miyazaki and colleagues noted no esophageal lesions in patients with nadir ET > 20°C. In the study by Ahmed and colleagues, only 1 out of 67 patients with nadir ET > 27°C developed an esophageal lesion. Mean ± standard deviation nadir ET for the 6 patients in that study developing esophageal lesions was 19.8°C ± 11.4°C compared to 31.7°C ± 4.5°C. We presumed that the abnormal esophageal wall in achalasia patients would be more susceptible to thermal injury, and based on the aforementioned studies, we prespecified a nadir ET of 27°C to abort ablation, which, fortunately, was never met.

In addition to intraprocedure ET, there is some evidence that gastroesophageal reflux may exacerbate direct ablation-related ulceration. Thus, prescription of proton pump inhibitor (PPI) may reduce the risk for fistula formation. Pre- and postprocedure PPI is generally felt to be reasonable, particularly in light of the overall excellent safety profile of these medications, and is a practice adopted by many centers performing AF ablation. The use of pre- or postprocedure sucralfate for esophageal protection has not been specifically studied, although cases of postablation esophageal ulcers have been successfully treated with PPI and sucralfate. For these reasons, we opted to treat the patient with both agents pre- and postprocedurally.

We considered, but did not use, esophageal deviation devices to move the esophagus away from ablation target sites. These devices were designed for the normal esophagus and no data exist for their safety or efficacy in achalasia. We had concerns regarding both ability to effectively maneuver a dilated esophagus (which would likely have a “trailing edge”) away from ablation sites, as well as the safety of exerting pressure on a dilated and remodeled esophagus that may have altered mechanical properties. With a megaeosophagus, the increased size of the lumen may preclude accurate measurement of ET with the use of conventional probes. Fukaya and colleagues recently reported a successful RF catheter ablation in a patient with achalasia and esophageal dilation despite endoscopic myotomy with the use of a steerable ET probe (Esophashtar, Japan Lifeline Co, Tokyo, Japan) that could be positioned in proximity to the ablation sites in order to more accurately detect worrisome temperature changes. As this ET probe was not clinically available to us, we opted for the use of a multisensor, S-shaped ET probe (CIRCA S-CATH; CIRCA Scientific, Englewood, CO) that would span more of the esophageal width, and may provide superior procedural temperature monitoring.

Conclusion
Further study is needed to clarify the role and mechanism of ET monitoring as well as to clarify methods to mitigate the risk for significant esophageal injury during catheter ablations for AF. A protocol for risk mitigation using Cryoballoon with a conservative temperature threshold for nadir ET during cooling, periprocedural pharmacologic esophageal protection, and multisensor esophageal temperature monitoring may be effective for patients at elevated risk for AEF, such as those with achalasia, who are undergoing AF ablation.

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