Left atrial appendage device closure in an adult patient with univentricular heart: First reported case

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Introduction

Recent advances in surgical and medical therapies for congenital heart disease have resulted in a growing adult congenital heart disease (ACHD) population with various long-term complications including arrhythmias, ventricular dysfunction, and valvular heart disease. Atrial fibrillation (AF) is among the most common arrhythmias encountered in the complex ACHD population, imposing detrimental hemodynamic effects and significant thromboembolic risk. Left atrial appendage (LAA) closure devices, such as the Watchman device (Boston Scientific, St. Paul, MN), have been widely used in patients with nonvalvular AF for stroke prevention, with multiple clinical trials demonstrating comparable safety and efficacy to anticoagulant therapy. However, limited information is available regarding the use of these devices in complex ACHD patients. The heterogenous morphologic abnormalities in the ACHD population impose unique challenges with percutaneous LAA device implantation. We report the first case of a Watchman implantation in a patient with single-ventricle physiology who had persistent LAA thrombus despite multiple prolonged regimens of anticoagulation.

Case report

A 32-year-old woman with tricuspid atresia and pulmonary atresia with severely hypoplastic right ventricle presented with increasing dyspnea and progressive exercise intolerance. She underwent palliative central shunt placement in infancy with atrial septectomy; her course was later complicated by pulmonary vascular disease, resulting in unsuitability for subsequent Fontan palliation. At age 25, she developed symptomatic AF refractory to multiple antiarhythmics, including sotalol and amiodarone. She subsequently underwent 2 catheter ablations, with brief success. On the third ablation attempt, she was found to have a distal LAA thrombus on transesophageal echocardiography (TEE) that was persistent despite multiple prolonged anticoagulation regimens including warfarin, oral factor Xa inhibitors, and low-molecular-weight heparin. Given the presence of a persistent thrombus with risk for embolism, LAA closure or ligation was considered. Prior thoracotomy precluded the use of minimally invasive LAA ligation strategies with the LARIAT (SentreHEART, Redwood, CA) or AtriClip (AtriCure Inc, Westchester, OH) device. The risk of redo open heart surgery for standalone LAA ligation was prohibitively high. After a shared process of decision making, it was decided to proceed with LAA closure using a Watchman device.

Preprocedure TEE and cardiac computed tomography (CT) revealed severely dilated common atrium (post atrial septectomy), as well as severely dilated left ventricle and windsock-shaped elongated LAA (4.8 cm) with ostium measuring 2.7 cm, and a mural thrombus noted on the distal aspect of the LAA measured 15 × 8 mm (Figure 1). In preparation for the procedure, the patient was taken off oral anticoagulation and was admitted for 48 hours of intravenous heparin that was stopped 4 hours prior to the procedure.

KEY TEACHING POINTS

- Atrial arrhythmia is a leading cause of morbidity in the adult congenital heart disease (ACHD) population and is associated with a high risk of thromboembolism despite absence of traditional risk factors.
- Left atrial appendage (LAA) closure devices could present an alternative or additional therapeutic option to chronic anticoagulation in the ACHD population, particularly in patients with persistent left atrial thrombus, and in those with underlying cognitive impairment.
- Percutaneous LAA closure can be safely performed in the presence of distally located “chronic” appendage thrombus with technical modifications.

KEYWORDS
Atrial fibrillation in single ventricle; Left atrial appendage closure device; Single ventricle; Watchman device; Univentricular heart

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right femoral venous access was obtained, a double-curve Watchman delivery sheath was advanced over a 0.32 guide-wire through the inferior vena cava to the common atrium. The lack of interatrial septum obviated a transseptal puncture. However, without the natural “fulcrum” provided by the interatrial septum (as with usual Watchman implantations requiring transseptal puncture), the delivery sheath was very unstable. Additionally, obtaining sheath coaxiality with the appendage was particularly challenging, given the relatively superior trajectory of the appendage. As such, we were unable to engage the LAA with the commonly used angled pigtail catheter via the delivery sheath. A deflectable decapolar catheter (Dynamic XT; Boston Scientific) was introduced through the Watchman delivery sheath and used to engage the LAA ostium, over which the sheath was advanced. The catheter was then replaced with angled pigtail for further adjustment of the delivery sheath position as well as LAA angiography. To avoid thrombus dislodgment, the pigtail as well as the sheath were always kept proximal to the thrombus, allowing implantation with “thrombus trapping,” which was feasible given the length of the LAA and the distal location of the thrombus in the LAA. The sheath was further advanced into the LAA with extreme counterclockwise torque to achieve more coaxiality with the LAA. Contrast was injected through the sheath with low pressure at a position away from the thrombus (Figure 2A). Although contrast injection in the LAA with known thrombus is always a concern and imposes a risk for thrombus dislodgment, considering the location of the thrombus being distal in the LAA and attached to the lateral mural wall, the risk of embolization was thought to be minimal with careful contrast injection directing the tip of the angiographic catheter away from the location of the thrombus and with injecting minimal amount of contrast.

Angiographic and TEE measurements suggested the use of a 33 mm Watchman 2.5 device that was deployed successfully with an excellent ostial position, adequate compression of approximately 15% as well as stability, and no significant residual flow into the LAA (Figure 2B–2F). There were no periprocedural complications. Repeat TEE and cardiac CT in 3 months revealed a well-positioned Watchman device with no significant leak around the device. The residual mural
thrombus in the distal LAA aspect was noted by cardiac CT and appeared smaller (4 × 6 mm) (Figure 3). After the procedure, she was started on antiplatelet therapy of clopidogrel 75 mg twice daily along with oral anticoagulation with apixaban 5 mg twice daily for 6 weeks. Thereafter, she was continued on aspirin 81 mg once daily along with low-dose apixaban 2.5 mg twice daily. Although the standard for post-LAA device closure includes a short course (6 weeks) of anticoagulation followed by dual antiplatelet therapy for 6 months post LAA closure, we opted to continue with aspirin and low dose of anticoagulation, considering underlying complex hemodynamics of palliated single ventricle with Eisenmenger physiology and complete dependence of pulmonary blood flow on the central shunt between ascending aorta and left pulmonary artery.

Discussion
We present an innovative application of LAA closure device technology in an adult patient with univentricular heart and AF with a persistent LAA thrombus. To the best of our knowledge, this is the first report of Watchman device implantation in a complex ACHD patient with single-ventricle physiology.

With the growing number of congenital heart disease patients reaching adulthood, numerous long-term complications such as atrial arrhythmias have been observed, with associated morbidities including worsening ventricular function and thromboembolic events. AF is the second most common atrial arrhythmia (29%) in the ACHD population after intra-atrial reentrant tachycardia (68%). The management of AF in adult patients with single-ventricle physiology is complex and includes different management strategies of rate control, rhythm control, and catheter ablations combined with effective anticoagulation regimen. Despite the relatively younger age of ACHD patients with absence of typical risk factors for thromboembolism, they remain at a considerable high risk for thromboembolic complications. Therefore, specific risk factors tailored to underlying anatomical and physiological status should be reviewed when considering chronic anticoagulation, such as chronic cyanosis, systemic ventricular dysfunction, the prothrombotic nature of single-ventricle physiology, and persistent interatrial shunting. In our patient, underlying chronic cyanosis with common atrium physiology (mixing of systemic and pulmonary venous returns at the atrial level) and systemic ventricular dysfunction in the setting of AF portended a high risk of thromboembolism.

LAA device closure has emerged as a stroke prevention alternative to oral anticoagulants, with multiple trials demonstrating noninferiority to systemic anticoagulation in high-risk patients. It is important to note that these trials did not include those with ACHD; as such, there is currently limited data regarding the feasibility of using LAA devices in complex ACHD patients. The variable, complex anatomy and limited operator experience impose a challenge for most
implanters. Therefore, a multidisciplinary approach and detailed preprocedural multimodality imaging evaluation at a center with a comprehensive ACHD program is crucial before considering LAA device closure in these cases. The utility of LAA closure for stroke prevention could potentially be an attractive alternative to full-dose, longer-term oral anticoagulation in the ACHD population, particularly with the high prevalence of cognitive and psychological problems in ACHD patients compared to the general population, which could impact compliance. Further studies through national registries and multicenter collaborative networks are warranted to explore the safety and efficacy of LAA device closure in the ACHD population and to formulate an anticoagulation regimen as well as perioperative imaging protocols.

One of the relative contraindications for performing percutaneous LAA device closure is the presence of LAA thrombus owing to the risk for thrombus dislodgment during implantation, with subsequent devastating complications. Nevertheless, few reports have demonstrated the safety and feasibility of percutaneous device closure in patients with persistent LAA thrombus despite appropriate anticoagulation therapy or in patients with contraindication to anticoagulation. If percutaneous LAA closure is considered in cases with known or suspected LAA thrombus, every effort should be made to avoid contrast injection in the LAA. In our case, given the complexity of the underlying congenital heart disease and particularly unusual orientation of the LAA in relation to the common atrium, it was felt that the benefit of minimal contrast injection outweighs the risk of thrombus dislodgment with directing of the tip of the angiographic catheter away from the location of the thrombus and with injection of minimal amount of contrast.

The use of a cerebral protection system was considered in this patient; however, we decided against using such a device given the underlying fragile hemodynamics and the need for catheter manipulation in the aortic arch, which, in the presence of central shunt, carries a risk for compromising pulmonary blood flow, which would have been devastating in our patient, giving complete dependence of pulmonary blood flow on the shunt flow. Furthermore, the risk for thrombus dislodgment was thought to be low considering the stability of the thrombus over serial imaging and the location in the distal end of an elongated appendage.

**Conclusion**

We report the first case of LAA closure device implantation in a patient with complex ACHD and a persistent appendage thrombus. LAA closure with Watchman device can be considered as an effective alternative or additional management strategy to standard anticoagulation in selected

![Image of LAA closure device implantation](https://example.com/image)
ACHD populations after a careful individualized approach by a multidisciplinary team.

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