

Addressing Stroke Risk in a Patient with CREST Syndrome and Atrial Fibrillation with Left Atrial appendage Occluder Device (WATCHMAN)

Ali Alkhayru^{1*}, Adria Madera-Acosta², Bassman Tappuni³, Belal Kaseer⁴, Kamal Shemisa⁵

¹Institutions: University of Iowa Health Care

²Institutions: Augusta University

³Institutions: Cincinnati Good Samaritan Hospital.

⁴Institutions: Cincinnati Good Samaritan Hospital.

⁵Institutions: Cincinnati Good Samaritan Hospital and Trihealth health system.

*Corresponding Author: Ali Alkhayru, MD Adult Hospitalist Program. Department of Internal Medicine University of Iowa Health Care

Received: December 21, 2020; Accepted: December 28, 2020; Published: January 06, 2021

Citation: Ali Alkhayru, Adria Madera-Acosta, Bassman Tappuni, Belal Kaseer, Kamal Shemisa (2021) Addressing Stroke Risk in a Patient with CREST Syndrome and Atrial Fibrillation with Left Atrial appendage Occluder Device (WATCHMAN). *J. Clinical Cardiology and Cardiovascular Interventions*, 4(1); Doi:10.31579/2641-0419/123

Copyright: © 2021 Ali Alkhayru, This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

Abstract

CREST syndrome is rare autoimmune disease causing calcinosis, Raynaud phenomenon, esophageal dysmotility, sclerodactyly and telangiectasias. We present a case of an eighty-two year old female with CREST syndrome who presented to our clinic with atrial fibrillation and prohibitive bleeding risk. Managing stroke risk in atrial fibrillation is essential to minimize the morbidity and mortality of the condition. Those with CREST syndrome presenting with recurrent gastrointestinal bleeding may require alternatives to anticoagulation. Recently, the left atrial appendage occluder device became widely used to manage patients at increased risk for bleeding. The device provides a safe and efficacious alternative in lowering atrial fibrillation associated stroke risk. Our patient underwent uncomplicated implantation of the left atrial appendage occluder device. She was closely monitored for one year where she remained stroke free and had one minor episode of gastrointestinal hemorrhage.

Keywords: crest syndrome; atrial fibrillation; watchman device; gastrointestinal hemorrhage

Short Title: CREST syndrome and stroke

Introduction

CREST syndrome is a variant of scleroderma that is characterized by calcinosis, Raynaud phenomenon, esophageal dysmotility, sclerodactyly and telangiectasia. Patients with CREST syndrome often suffer from cardiovascular diseases including coronary artery disease, myocardial fibrosis, primary pulmonary hypertension and arterial embolization [1].

Atrial fibrillation raises stroke risk. it is estimated that about 87% of strokes are ischemic in nature and atrial fibrillation is the underlying cause of 15-20 % of cases [2]. Management of this risk usually requires long-term anticoagulation if the CHA2DS2Vasc score is elevated (>2 in men and >3 women) [3]. However, in certain susceptible patient populations, long term anticoagulation can significantly raise the risk for spontaneous hemorrhage [4]. We present a case of a patient with CREST syndrome who suffered from atrial fibrillation and encountered recurrent bleeding due to multiple gastrointestinal telangiectasias.

Case report:

An eighty-two year old female with past medical history of CREST syndrome, diabetes mellitus, hypertension and permanent atrial

fibrillation presented to the cardiology clinic after hospitalization for atrial fibrillation. She was previously treated with a rhythm control strategy and while on amiodarone, underwent two successive cardioversions. She maintained sinus rhythm for three months after the second cardioversion but presented to the hospital with increasing shortness of breath and melena and was found to have recurrent atrial fibrillation and severe microcytic anemia with a hemoglobin concentration of 6.4 grams/dl.

On evaluation she was hemodynamically stable and in mild respiratory distress. She was transfused three units of packed red blood cells and apixaban was discontinued. The hemoglobin increased appropriately to 9 grams/dl after transfusion. Administration of the apixaban reversal agent, adexanetalfa, was therefore deferred. She was evaluated by our gastroenterologists with esophagogastroduodenoscopy (EGD) and a colonoscopy. The EGD revealed multiple small nonbleeding telangiectasias along the duodenum, cecum and ascending colon. These areas were successfully cauterized, and daily pantoprazole was recommended.

She was not a candidate for anticoagulation during this period. We estimated the stroke risk attributed to atrial fibrillation using the CHA₂DS₂VASc score which was five estimating a stroke risk of approximately 6.7% in the first year [3]. The HAS-BLED bleeding risk score was seven estimating a bleeding risk is 3.74% in the first year while on anticoagulation. Although resuming anticoagulation would have effectively addressed the stroke risk, she was high risk for recurrent and potentially life-threatening gastro-intestinal hemorrhage. Thus, we considered procedural options in the management of stroke risk associated with atrial fibrillation.

Treatment with the left atrial appendage occlusion device shortens the duration for anticoagulation to 45 days and thus minimizes the risk for bleeding. Because of recurrent bleeding, elevated stroke risk and risk of rebleeding, she was considered a suitable candidate for the left atrial appendage occluder device, (WATCHMAN device). A transesophageal echocardiogram demonstrated a left atrial appendage with “windsock” morphology, anatomically suitable for left atrial appendage closure. The right ventricular systolic pressure was 30 mmHg. She underwent successful deployment of the WATCHMAN device using standard technique (Figure 1, and Figure 2).

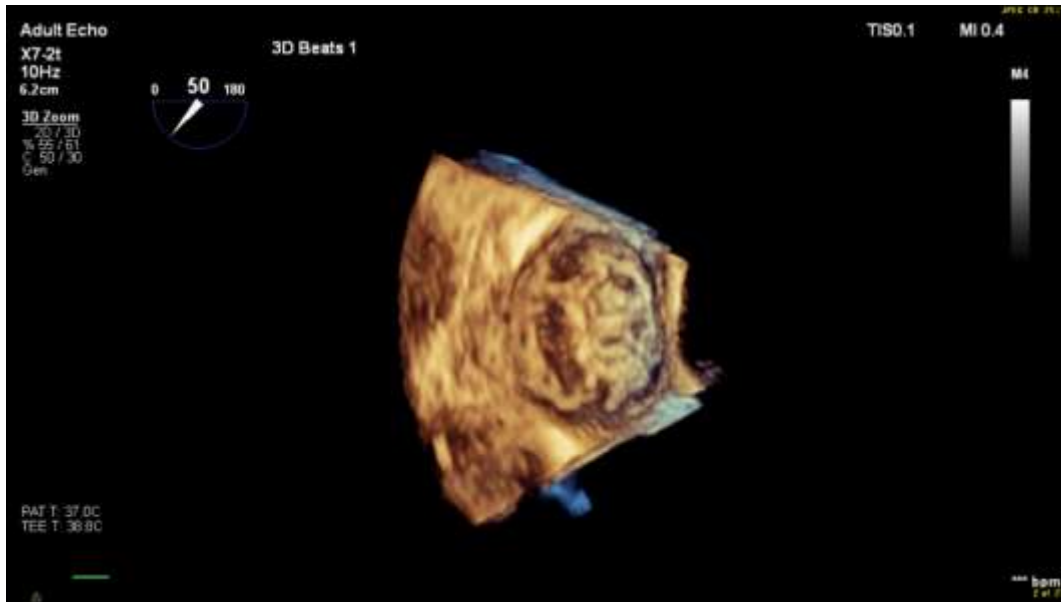


Figure 1: 3D Echocardiogram showing appendage occluder device with appendage visible.



Figure 2: Transesophageal Echocardiogram short axis view showing appendage occluder device.

She was then anticoagulated with warfarin for 45 days following the device placement and did not suffer gastrointestinal hemorrhage during that period [5]. She was followed for one year where she remained stroke free and had one minor episode of gastrointestinal hemorrhage.

Discussion:

Systemic sclerosis is a rare autoimmune connective tissue disease with varying severity. The condition falls into two major categories; diffuse or

localized scleroderma. The localized version is known as CREST Syndrome and has a fourfold higher prevalence among women compared to male counterparts [6]. Bleeding telangiectasias can cause severe gastrointestinal bleeding which have high mortality rate, up to 22% in some series. Women were 2.7 times more likely to have gastrointestinal bleeding due to telangiectasias [7]. Iron deficiency anemia due to severe acute upper gastrointestinal bleeding is common. Hence, Proper endoscopic evaluation is needed in patients with CREST syndrome who present with microcytic anemia. Recurrent bleeding from mucosal telangiectasia is possible [8].

Atrial fibrillation is widely encountered in medical practice. Its most feared consequence is stroke resulting in permanent neurologic sequelae [3]. The main scoring system used to predict thromboembolic stroke risk in patients with atrial fibrillation is CHA₂DS₂VASc score, a comprehensive scoring system using clinical variables to identify those at risk [1]. Patients with a CHA₂DS₂VASc score of 2 or higher require anticoagulation. Bleeding risk with anticoagulation can be estimated using the HAS-BLED bleeding score that incorporates known clinical risk factors that raise bleeding risk [10]. Anti-coagulation raises both the risk of intracranial bleeding and gastrointestinal bleeding [4]. Gastrointestinal bleeding is more common and is not an absolute contraindication to anticoagulation. Risk of recurrence and individual patient factors need to be carefully considered prior to resuming anticoagulation. Those with gastrointestinal telangiectasias are at higher risk for recurrent bleeding [11,12].

In our patient, we considered strategies to occlude the left atrial appendage and therefore reduce the duration of treatment with anticoagulation. The WATCHMAN Device is a minimally invasive procedure that can occlude the left atrial appendage. The device was non-inferior to warfarin as a strategy to lower stroke risk (2.3 events per 100 patient-years with WATCHMAN compared to 3.8 events per 100 patient-years with warfarin (rate ratio, 0.60; 95% credible interval, 0.41-1.05, noninferior margin rate ratio of < 2). The estimated relative risk reduction for stroke was as low as 1.1 in those with high CHA₂DS₂VASc score. The Watchman device has a high success rate of about 95% in sealing the left atrial appendage and shortens the period of anticoagulation to 45 days [13].

The parachute shaped implantable nitinol device is encased within a trans-septal access sheath and delivery catheter. After successful trans-septal puncture of the atrial septum, introduction of the WATCHMAN access system is followed by isolation of left atrial appendage with a pigtail catheter to allow for safe advancement of the WATCHMAN delivery system and finally deployment of the device.

When performing trans-septal puncture of the atrial septum, there should be careful consideration of patients at risk for pulmonary hypertension such as our patient. Pulmonary hypertension can occur in as many as 35% of patients with CREST syndrome. If untreated, transseptal puncture can lead to right to left shunting patients and cyanosis in such patients. Therefore, patients with CREST affected by pulmonary hypertension who undergo the procedure should be treated with an atrial septal occlude device (Amplatzer) in order to avoid such complications [14].

Conclusion:

WATCHMAN device has proven safety and efficacy and maybe the more appropriate option for stroke prevention in patients with atrial fibrillation and Crest Syndrome who are at higher risk of bleeding from gastrointestinal telangiectasias.

Teaching points:

- 1) Patients with CREST syndrome who suffer from atrial fibrillation may have increased risk for bleeding making treatment of atrial fibrillation challenging.
- 2) Stroke prevention in those with CREST syndrome and atrial fibrillation can be achieved safely with WATCHMAN device.

Disclosures:

Kamal Shemisa, MD, FACP, FACC: Speakers Bureau: Boehringer Ingelheim, Espiron, and Zoll Cardiac Devices.

Other Authors: Nothing to disclose.

Acknowledgements: None

List of Support/Grant Information: None

Statement of contribution: All authors contributed equally to the manuscript.

Word count: 1500 (Excluding Abstract).

References:

1. Mavrogeni S, Bratis C, Manoussakis M. Coronary artery abnormalities in CREST syndrome revealed by cardiovascular magnetic resonance imaging. *Can J Cardiol.* 2011;27(3):390.e5-390.e7. doi:10.1016/j.cjca.2010.12.052
2. January CT, Wann LS, Calkins H, et al. 2019 AHA/ACC/HRS Focused Update of the 2014 AHA/ACC/HRS Guideline for the Management of Patients With Atrial Fibrillation: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. *J Am Coll Cardiol.* 2019;74(1):104-132. doi:10.1016/j.jacc.2019.01.011
3. Lip GYH, Nieuwlaat R, Pisters R, et al. Refining clinical risk stratification for predicting stroke and thromboembolism in atrial fibrillation using a novel risk factor-based approach: The Euro Heart Survey on atrial fibrillation. *Chest.* 2010;137(2):263-272. doi:10.1378/chest.09-1584
4. Pisters R, Lane DA, Nieuwlaat R, de Vos CB, Crijns HJGM, Lip GYH. A Novel User-Friendly Score (HAS-BLED) To Assess 1-Year Risk of Major Bleeding in Patients With Atrial Fibrillation. *Chest.* 2010;138(5):1093-1100. doi:10.1378/chest.10-0134
5. Reddy VY, Sievert H, Halperin J, et al. Percutaneous left atrial appendage closure vs warfarin for atrial fibrillation a randomized clinical trial. *JAMA - J Am Med Assoc.* 2014;312(19):1988-1998. doi:10.1001/jama.2014.15192
6. Chiffot H, Fautrel B, Sordet C, Chatelus E, Sibilia J. Incidence and Prevalence of Systemic Sclerosis: A Systematic Literature Review. *Semin Arthritis Rheum.* 2008;37(4):223-235. doi:10.1016/j.semarthrit.2007.05.003
7. Duchini A, Sessoms SL. Gastrointestinal hemorrhage in patients with systemic sclerosis and CREST syndrome. *Am J Gastroenterol.* 1998;93(9):1453-1456. doi:10.1111/j.1572-0241.1998.00462.x
8. Chen JY, Zhang AD, Lu HY, Guo J, Wang FF, Li ZC. CHADS2 versus CHA2DS2-VASc score in assessing the

- stroke and thromboembolism risk stratification in patients with atrial fibrillation: A systematic review and meta-analysis. *J Geriatr Cardiol.* 2013;10(3):258-266. doi:10.3969/j.issn.1671-5411.2013.03.004
9. Zhu WG, Xiong QM, Hong K. Meta-analysis of CHADS2 versus CHA2DS2-VASc for predicting stroke and thromboembolism in atrial fibrillation patients independent of anticoagulation. *Texas Hear Inst J.* 2015;42(1):6-15. doi:10.14503/THIJ-14-4353
 10. Radaelli F, Dentali F, Repici A, et al. Management of anticoagulation in patients with acute gastrointestinal bleeding. *Dig Liver Dis.* 2015;47(8):621-627. doi:10.1016/j.dld.2015.03.029
 11. Lip GYH, Lowe GDO. ABC of Atrial Fibrillation: ANTITHROMBOTIC TREATMENT FOR ATRIAL FIBRILLATION. *BMJ.* 1996;312(7022):45-49. doi:10.1136/bmj.312.7022.45
 12. Boersma L V., Ince H, Kische S, et al. Efficacy and safety of left atrial appendage closure with WATCHMAN in patients with or without contraindication to oral anticoagulation: 1-Year follow-up outcome data of the EWOLUTION trial. *Hear Rhythm.* 2017;14(9):1302-1308. doi:10.1016/j.hrthm.2017.05.038
 13. Reddy VY, Doshi SK, Sievert H, et al. Percutaneous left atrial appendage closure for stroke prophylaxis in patients with atrial fibrillation 2.3-year follow-up of the PROTECT AF (Watchman left atrial appendage system for embolic protection in patients with atrial fibrillation) trial. *Circulation.* 2013;127(6):720-729. doi:10.1161/CIRCULATIONAHA.112.114389
 14. Battle RW, Davitt MA, Cooper SM, et al. Prevalence of Pulmonary Hypertension in Limited and Diffuse Scleroderma. *Chest.* 1996;110(6):1515-1519. doi:10.1378/chest.110.6.1515



This work is licensed under Creative Commons Attribution 4.0 License

To Submit Your Article Click Here: [Submit Manuscript](#)

DOI:10.31579/2641-0419/123

Ready to submit your research? Choose Auctores and benefit from:

- ❖ fast, convenient online submission
- ❖ rigorous peer review by experienced research in your field
- ❖ rapid publication on acceptance
- ❖ authors retain copyrights
- ❖ unique DOI for all articles
- ❖ immediate, unrestricted online access

At Auctores, research is always in progress.

Learn more www.auctoresonline.org/journals/clinical-cardiology-and-cardiovascular-interventions