ESC European Society of Cardiology

European Heart Journal - Case Reports (2023) **7**, 1–5
https://doi.org/10.1093/ehjcr/ytad251

CASE REPORT
Cardiovascular imaging

Congenital membranous ostial stenosis of the left atrial appendage as a secondary finding in a patient with ST elevation myocardial infarction: a case report

David Hanke *, Ali Hamadanchi, P. Christian Schulze, and Marcus Franz

Department of Cardiology, University Hospital Jena, Am Klinikum 1, 07747 Jena, Germany

Received 8 August 2022; first decision 6 September 2022; accepted 17 May 2023; online publish-ahead-of-print 2 June 2023

Background Stenoses of the left atrial appendage (LAA) represent a common complication after incomplete surgical ligation. However, the idiopathic entity is very rare. So far, there is uncertainty about the thromboembolic risk and potential benefit of anticoagulation in these patients. We report on congenital ostial stenosis of the LAA as a secondary finding in a patient with myocardial infarction.

Case summary A 56-year-old patient presented with acute heart failure secondary to ST elevation myocardial infarction (STEMI) and eventually progressed to cardiogenic shock. A percutaneous coronary intervention and stent placement in the first diagonal branch and in the left anterior descending artery was performed in two sessions. There was a new onset of typical atrial flutter and paroxysmal atrial fibrillation with haemodynamically relevant tachycardia. Before synchronized electrical cardioversion, we performed transoesophageal echocardiography. Left atrial thrombi were ruled out. Surprisingly, we found membranous ostial stenosis of the LAA, resulting in a bidirectional flow pattern. After 28 days of treatment in the intensive care unit the patient had full clinical recovery.

Discussion Given the very rare cases of congenital LAA ostial stenosis, there is uncertainty about the thrombogenicity and the potential benefit of anticoagulation or even a percutaneous closure of the LAA. We discuss possible similarities regarding the thromboembolic risk of patients with an idiopathic narrowing of the LAA to patients with incomplete surgical ligation and patients with a device leak after percutaneous LAA closure. Congenital ostial LAA stenosis represents a clinically relevant condition and may be considered as a potential hazard for thromboembolism.

Keywords Case report • Atrial fibrillation • Left atrial appendage closure • Stenosis • Membrane • Anomaly • Transoesophageal echocardiography

ESC Curriculum 2.1 Imaging modalities • 2.2 Echocardiography • 3.2 Acute coronary syndrome • 5.3 Atrial fibrillation

Learning points

- Congenital ostial LAA stenosis is a very rare entity.
- Congenital LAA stenosis represents a clinically relevant condition and may be considered as a possible hazard for thromboembolism.
- There are several pathophysiologic mechanisms that might affect the thrombogenicity in LAA ostial stenosis.

* Corresponding author. Tel: +49 3641 9324534, Email: David.hanke@med.uni-jena.de
Handling Editor: Luca Arcari/Vincenzo Nuzzi
Peer-reviewers: Aisha Gohar; Marta Cvijic; Edgar Francisco Carrizales
Compliance Editor: Nicholas Weight
Supplementary Material Editor: Tom Wardill

Introduction

Secondary stenosis of the left atrial appendage (LAA) is a frequent complication of incomplete surgical ligation (in 35% of patients)[1,2] or as a consequence of device leak after percutaneous LAA closure (in 26% of patients).[3,4] Both scenarios are associated with an increased risk for thromboembolic events.

In contrast, congenital malformations of the LAA are very rare and, to the best of our knowledge, there are not more than 18 reported cases of complete LAA absence[5] and only one case of LAA hypoplasia.[6] Traceably, in those cases, the thromboembolic risk is considered to be reduced.

Focusing on primary congenital LAA stenosis, there are only 14 published cases.[7] This lack of data leads to uncertainty concerning the risk for thromboembolic events in such patients. Thus, it is of great clinical importance to describe all clinical constellations, in which LAA stenosis is detected.

We report on congenital ostial LAA stenosis as a secondary finding in a patient with subacute myocardial infarction resulting in cardiogenic shock and prolonged severe disease course.

Timeline

Day 1	Admission to the hospital with ST-elevation myocardial infarction, coronary angiography with percutaneous coronary intervention (PCI), and stent placement in the first diagonal branch (RD-1) of the left anterior descending artery (LAD)
Day 2	Second session coronary angiography: thrombotic occlusion of the LAD close to the RD-1 intervention site, PCI and stent placement in the LAD
Day 3	Progressive cardiogenic shock, implantation of percutaneous left ventricular mechanical circulatory support device (Impella® CP)
Day 4	Removal of the Impella® CP
Day 5	Onset of atrial flutter and paroxysmal atrial fibrillation with haemodynamically relevant tachycardia, transoesophageal echocardiography excluded LAA thrombus, but revealed a discrete membranous ostial stenosis of the LAA, synchronized electrical cardioversion was performed in total four times on this day and during the next days until stable sinus rhythm was achieved, supported by full titration of Amiodarone
Day 17	Ventricular fibrillation, cardiopulmonary resuscitation for 2 min, one defibrillation
Day 19	Implantation of an internal cardioverter defibrillator
Day 22	Cranial computer tomography because of a prolonged delirium, revealing bilateral former cerebral infarctions of the frontal cortex area
Day 28	Full clinical recovery, discharge, and outpatient rehabilitation

Case presentation

A 56-year-old male patient presented with acute heart failure. He was a heavy smoker with severe chronic alcohol abuse and a prior splenectomy secondary to trauma. Otherwise, clinical history including cardiovascular diseases was unremarkable. He began three days before admission with progressive dyspnoea. On initial evaluation, he was anxious and with an overall poor general condition. The pulse was regular and on chest auscultation, we found basal crackles.

The electrocardiogram showed sinus rhythm with normal heart rate and an R-loss from V1 to V3 as well as an ST-Elevation in the leads V2 and V3. Laboratory analysis showed a greatly increased troponin T (57 717 pg/mL reference < 16.8). Renal and hepatic function was almost normal [glomerular filtration rate (GFR): 86.7 µmol/L reference > 90; creatinine: 86 µmol/L reference 62–106; aspartate transaminase (ASAT): 0.47 µmol/L reference < 0.85; gamma-glutamyl-transferase: 2.1 µmol/L reference 0.17–1.19; bilirubin: 9 µmol/L reference < 21]. Transthoracic echocardiography revealed a dilated left and right ventricle with severely reduced systolic function (left ventricular end-diastolic diameter: 7 cm, interventricular septal thickness at end-diastole: 10 mm, left ventricular ejection fraction (LVEF): 10%, stroke volume 25 mL, right ventricular end-diastolic diameter: 5.1 cm, tricuspid annular plane systolic excursion: 9 mm) and apical akinesia. Moderate functional mitral and tricuspid regurgitation were noted and the systolic pulmonary artery pressure was estimated as 25 mmHg.

Coronary angiography performed on the day of admission revealed a moderate stenosis of the proximal left anterior descending artery (LAD) and an occlusion of the first diagonal branch (RD1). Percutaneous coronary intervention (PCI) with placement of a drug-eluting stent (DES) was performed successfully in the RD1 lesion, the LAD stenosis was left since it was assessed as non-significant. One day later, due to clinical decline, a second-look coronary angiography was performed. An acute thrombotic occlusion of the LAD close to the RD1 intervention site was detected and had to be interpreted, most likely, as a post-interventional complication. Of course, also coronary embolism had to be considered as an alternative cause. Nevertheless, another PCI with DES placement in the LAD was performed successfully. Unfortunately, on the third day, the patient went into cardiogenic shock with signs of hypoperfusion and peripheral vasoconstriction (Killip-Kimbal IV, Stevenson class C: cold and wet). Blood pressure was 96/80 mmHg under continuous infusion of noradrenaline (0.28 µg/min/kg). A chest X-ray displayed biventricular cardiac enlargement and pulmonary congestion; serum lactate was 4.8 mmol/L (Reference < 2.2 mmol/L). The patient rapidly went into multiple organ failure with impaired renal and hepatic function (GFR 9.6 mL/min, creatinine 530 µmol/L, ASAT 213 µmol/L, bilirubin 59 µmol/L), most likely caused by congestion. Due to anuria continuous veno-venous haemofiltration became necessary and the patient had to be intubated and required mechanical ventilation to control aggravating hypoxia.

Haemodynamic evaluation by right heart catheterization revealed a cardiac output of 1.1 L/min (Reference 4.5–5 L/min) and a cardiac index of 0.5 L/min/m^2 (Reference 2.5–4 L/min/m^2). We, therefore, decided, after excluding another coronary event, to place a percutaneous left ventricular mechanical circulatory support device (Impella® CP 3.5 L), which could be removed after approximately 24 h. Haemodynamic monitoring in the intensive care unit (ICU) was realized by pulmonary artery catheter and, due to the complexity of the case, was continued even after Impella® CP explantation for recompensation and tapering off the catecholamines.

During further disease course, new onset of typical atrial flutter and paroxysmal atrial fibrillation with haemodynamically relevant tachycardia up to 170 beats per minute occurred and, again, led to clinical decline with the need to re-escalate catecholamine doses. Since electrical cardioversion was intended, transoesophageal echocardiography (TOE) was performed to exclude LAA thrombus. Although thrombi or sludge could be ruled out by TOE, a discrete membranous ostial stenosis of the LAA was noticed as an incidental finding. The latter caused an accelerated bidirectional flow pattern at the LAA ostium with a velocity of up to 1 m/s, resulting in complete perfusion without stasis in any part of the LAA, despite typical atrial flutter (*Figures 1* and *2*, Supplementary material online, *Video S1*). We performed synchronized

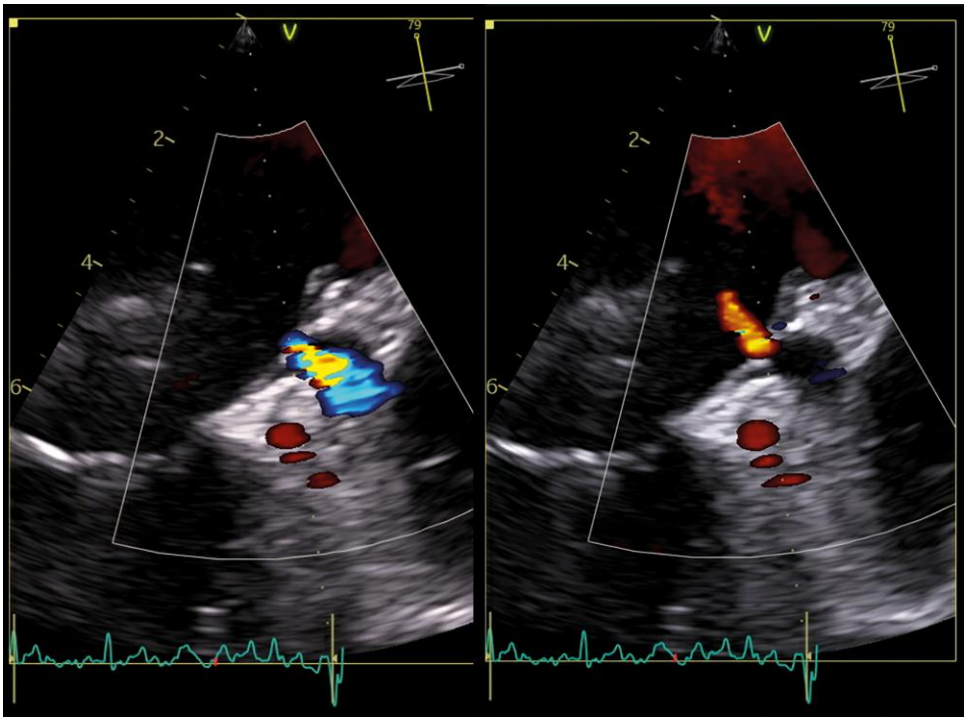

Figure 1 Transoesophageal echocardiographic image (midoesophageal, 79°) of a to (left) and fro (right) flow of blood across the membranous structure at the LAA orifice, endsystolic, respectively.

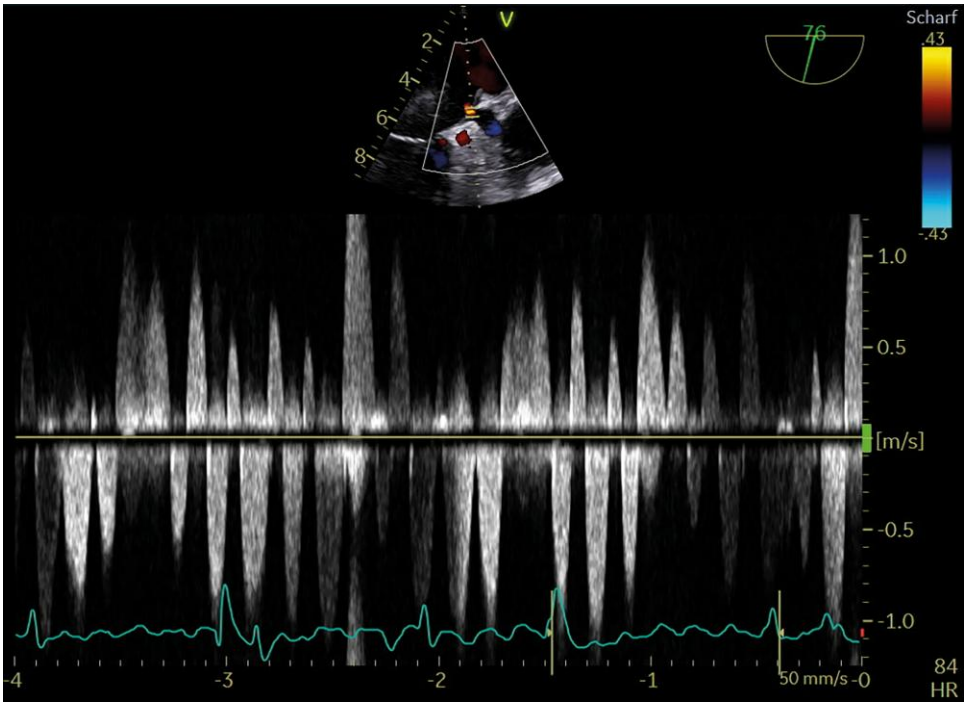

Figure 2 Transoesophageal echocardiographic image of pulsed wave Doppler displaying the to and fro flow at the LAA ostial narrowing with a frequency of 280–300/min according to the flutter frequency (midoesophageal, 79°).

electrical cardioversion, which had to be repeated four times, supported by full titration of antiarrhythmic medication with amiodarone (continuous intravenous amiodarone 900 mg/d up to 10 g, following a maintenance dose of oral 200 mg/d) to establish stable sinus rhythm.

With continuous veno-venous haemofiltration, mechanical ventilation, and application of noradrenaline up to 0.6 µg/min/kg, milrinone up to 0.49 µg/kg/min, and vasopressin up to 1 IU/h (maximum doses each), the patient's haemodynamics could be stabilized. The cardiac output normalized to 5.97 L/min and cardiac index to 3.05 L/min/m^2. During the severe course of the disease, cardiopulmonary resuscitation had to be performed once on day 17 because of an episode of ventricular fibrillation. For this reason and due to the severely reduced LVEF, an internal cardioverter defibrillator was implanted.

Since the patient was in persistent delirium, we also performed a cranial computer tomography, which revealed former bilateral frontal infarctions.

Fortunately, after a total of 28 days of ICU treatment followed by rehabilitation, the patient experienced full clinical recovery. The left ventricular function increased slightly to an LVEF of 25%. Laboratory parameters of hepatic function normalized and renal function recovered to a GFR of 78 mL/min. Because of paroxysmal atrial flutter and atrial fibrillation, we applied anticoagulation with unfractionated heparin during the ICU stay. The patient was discharged on oral treatment with apixaban (5 mg twice a day). In addition, the patient received a guideline-directed medical heart failure therapy including angiotensin-converting enzyme inhibitor (ramipril 2.5 mg), beta blocker (bisoprolol 5 mg), aldosterone antagonist (eplerenone 25 mg), and sodium-glucose cotransporter-2 inhibitor (empagliflozin 10 mg).

Discussion

We here report on a case of narrowed LAA ostium as an incidental finding in TOE. While stenoses of the LAA ostium are well-known complications after incomplete surgical ligation,[1] our patient never faced surgical or catheter-based interventions affecting the LAA. This condition has been previously described in patients with both atrial tachycardia[8] and sinus rhythm.[9,10] In a case reported by Coughlan and co-workers, the patient presented with a transient ischaemic attack.[9] A recently published study provides a review of the very few cases detected so far.[7] Here, in only one out of 13 patients, a thrombus formation was found in association with LAA ostial stenosis. However, in that particular case, the authors considered the LAA ostial stenosis not to be congenital but associated with rheumatic mitral stenosis.[11] It is conceivable that stasis after the stenosis results in thrombus formation. In our case, the TOE was done during typical atrial flutter resulting in an accelerated blood flow to and fro the LAA according to the flutter frequency (~280–300/min). It is well-known that perfusion of the LAA differs depending on the underlying cardiac rhythm (atrial fibrillation, atrial flutter, or sinus rhythm). Thus, it is conceivable that a higher frequency, e.g. due to atrial fibrillation, results in reduced blood flow across the narrowing, up to stasis and possible thrombus formation in the LAA. Katz and co-workers also described the high-velocity jet across the narrowing causing mechanical wall stress, resulting in fibrosis and therefore possible thrombus formation.[1] Even if the risk of thrombus formation behind the stenosis may be increased due to the above-described mechanisms, uncertainty remains on the questions, of how likely large thrombi can be released via narrow stenosis and then cause thromboembolic events.

The risk for thromboembolism is known to be increased in patients with LAA ostial stenosis after incomplete surgical closure.[1,12] In the idiopathic entity, given the very few cases, the hazard of thrombus formation and possible stroke remains unclear. Our patient showed no thrombi or sludge at the time when TOE was performed. Nevertheless, it is imaginable that the detected multiple former cerebral infarctions might be due to a former transient thrombus formation. The infarctions were located bilaterally in the frontal cortex, which could support the assumption of a cardioembolic cause. However, we cannot prove that cerebral lesions were caused by cardiac thromboembolisms since carotid atherosclerosis or any other causes must also be considered. By the way, echocardiography showed no signs of left ventricular thrombus formation, and a patent foramen ovale was ruled out.

A recent meta-analysis addressing interventional LAA closure revealed an association of persisting peri-device leaks at one year to a significantly higher stroke rate.[3] Alkhouli and co-workers analysed a large dataset of more than 50 000 patients with an occluder device.[4] Here, overall embolic and bleeding events occurred very rarely, but there were significantly higher rates of thromboembolic events in patients with a leak size of up to 5 mm. Interestingly, for larger leaks, there were no changes in thromboembolic risk. Whether this result is due to a pathophysiological mechanism resulting in a higher thrombogenicity remains unclear. The authors described a smaller narrowing causing higher blood flow and therefore a possible increased wall stress and fibrosis.[4] However, it is conceivable that for larger leaks no difference was found because of a higher rate of anticoagulation in these patients. Furthermore, given the very rare cases with a large leak, the study was probably underpowered for this question.

Regarding the rare idiopathic entity of LAA ostial stenosis, further investigations on how to treat these patients are needed. The possible benefit of oral anticoagulation or even a catheter-based closure using an occluder device needs to be clarified not only for patients presenting in sinus rhythm but also for those with atrial fibrillation. It is questionable if idiopathic LAA ostial stenoses can be considered similar to those resulting from partial LAA closure with respect to pathophysiology and potential consequences. Nevertheless, the risk for thromboembolic events in patients with congenital LAA ostial stenosis might be higher compared to patients in normal condition (sinus rhythm and an open LAA), but possibly still lower compared to patients with atrial fibrillation and an open LAA with normal anatomy.

So far, there are no guidelines for the therapy of idiopathic LAA ostial stenosis itself. Our patient was discharged with oral anticoagulation because of paroxysmal atrial fibrillation/flutter and a CHA2DS2-VASc-Score of 3, according to the current guidelines of the European Society of Cardiology.

Conclusion

Congenital ostial LAA stenosis is a very rare condition. The exact prevalence, pathogenesis, clinical significance, and possible therapeutic implications remain to be clarified. Nevertheless, the finding represents a clinically relevant condition and should be considered as a possible hazard for thromboembolism by the treating physician and whenever performing TOE to evaluate the LAA.

Lead author biography

Dr. David Hanke is a current resident in internal medicine and cardiology at University Hospital Jena (Germany). His research interests include echocardiographic imaging and haemodynamics of coronary artery disease.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text, have been obtained from the patient in line with the Committee on Publication Ethics (COPE) guidance.

Conflict of interest: None declared.

Funding: We acknowledge support by the German Research Foundation Projekt-Nr. 512648189 and the Open Access Publication Fund of the Thueringer Universitaets- und Landesbibliothek Jena.

Data availability

The data underlying this article are available in the article and in its online Supplementary material.

References

1. Katz ES, Tsiamtsiouris T, Applebaum RM, Schwartzbard A, Tunick PA, Kronzon I. Surgical left atrial appendage ligation is frequently incomplete: a transesophageal echocardiograhic study. *J Am Coll Cardiol* 2000;**36**:468–471.
2. Aryana A, Bhaskar R. Incomplete surgical ligation of the left atrial appendage-time for a new look at an old problem. *Ann Transl Med* 2017;**5**:141.
3. Reddy VY, Holmes DR, Doshi SK, Kar S, Singh S, Gibson D, *et al.* Peri-Device leak after left atrial appendage closure: impact on long-term clinical outcomes. *Circulation* 2021; **144**:E579–E579.
4. Alkhouli M, Du C, Killu A, Simard T, Noseworthy PA, Friedman PA, *et al.* Clinical impact of residual leaks following left atrial appendage occlusion: insights from the NCDR LAAO registry. *JACC Clin Electrophysiol* 2022;**8**:766–778.
5. Pourafkari L, Sadeghpour A, Baghbani-Oskouei A, Savadi-Oskouei S, Pouraliakbar H, Fazelifar AF, *et al.* Absent left atrial appendage: case report and review of the literature. *Cardiovasc Pathol* 2020;**45**:107178.
6. Hasegawa H, Ito T, Hourai R, Akamatsu K, Nomura Y, Miyamura M, *et al.* Hypoplastic left atrial appendage: a case report and literature review. *Am J Case Rep* 2021;**22**: e933260.
7. Sakatani Y, Ito T, Hasegawa H, Akamatsu K, Hoshiga M. Left atrial appendage ostial stenosis: a case report and literature review. *Am J Case Rep* 2021;**22**:e930510
8. Stern JD, Skolnick AH, Freedberg RS, Kronzon I. Isolated left atrial appendage ostial stenosis. *Eur J Echocardiogr* 2009;**10**:702–703.
9. Coughlan B, Lang RM, Spencer KT. Left atrial appendage stenosis. *J Am Soc Echocardiogr* 1999;**12**:882–883.
10. Ha JW, Chung N, Hong YS, Cho BK. Left atrial appendage stenosis. *Echocardiography* 2001;**18**:295–297.
11. Alsancak Y, Sezenoz B, Oncu F, Ucar M, Aydogdu Tacoy G. Left atrial appendage ostial stenosis associated with rheumatic mitral stenosis. *Anatol J Cardiol* 2015;**15**:E11–E12.
12. Kanderian AS, Gillinov AM, Pettersson GB, Blackstone E, Klein AL. Success of surgical left atrial appendage closure: assessment by transesophageal echocardiography. *J Am Coll Cardiol* 2008;**52**:924–929.