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CASE REPORT
Congenital heart disease

A case report of atrial fibrillation in early adulthood: dig deeper

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Background Atrial fibrillation (AF) is a common cardiac arrhythmia, which is often associated with underlying risk factors and undiagnosed conditions, including congenital heart disease. Atrial septal defects (ASDs) come to mind, albeit arrhythmias usually present later in life. We present herewith a young patient with cor triatriatum sinister (CTS), with some delay in establishing the diagnosis, following new onset AF in early adulthood.

Case summary A 31-year-old man presented with pre-syncope and coryzal symptoms and was newly diagnosed with AF in the context of an intercurrent viral illness. After treatment with oral anticoagulation and successful outpatient cardioversion, he was discharged from cardiology review. Two years later he re-presented with exercise intolerance and a 12-lead electrocardiogram revealing recurrence of AF. Subsequent investigation with transthoracic echocardiography revealed the underlying congenital cardiac defect of CTS, together with an ASD and patent foramen ovale. After corrective surgery, which involved membrane resection, a Cox-maze procedure and ASD closure, sinus rhythm was restored and at follow-up the patient had returned to baseline function.

Discussion Young patients presenting with new onset AF should undergo thorough cardiovascular assessment to identify treatable causes and reversible risk factors. Cor triatriatum sinister is a rare congenital anomaly that may present in adulthood and give rise to symptomatic AF. Surgical correction including a Cox-maze procedure in our patient resulted in restoration of sinus rhythm and a return of the patient's baseline functional status and improved quality of life.

Keywords Atrial fibrillation • Early adulthood • Cor triatriatum sinister • Case report

ESC Curriculum 5.3 Atrial fibrillation • 7.5 Cardiac surgery • 9.7 Adult congenital heart disease

Learning points

- Atrial fibrillation in early adulthood can be a marker of underlying undiagnosed conditions and should prompt thorough cardiovascular assessment in addition to symptomatic management.
- Cor triatriatum sinister is a congenital anomaly of the left atrium, which infrequently presents in adulthood and can be surgically corrected with good outcomes.

Introduction

Atrial fibrillation (AF) is the commonest and most clinically significant sustained cardiac arrhythmia in the general population.[1] The Global Burden of Disease project estimated the prevalence of AF in 2016 at around 46.3 million worldwide,[2] a number expected to rise owing to an ageing population, with age the commonest risk factor for AF. The prevalence of AF in younger individuals <40 years, on the other hand, is markedly lower at <0.1%,[3] and unlike older individuals with AF many do not have overt predisposing cardiovascular disease.

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Previously, these patients were deemed to have 'idiopathic' or 'lone' AF, terms historically used to describe AF in individuals under 60 years of age without clinical or echocardiographic evidence of cardiopulmonary disease.[4] This classification of AF has been abandoned as increasing knowledge of AF-related risk factors and diagnoses have been identified.[5] These include positive family history, binge alcohol drinking, athlete status, and structural heart disease such as congenital heart defects.[6]

Cor triatriatum is a rare congenital cardiac malformation characterized by a thin fibromuscular membrane within either the left (sinister) or right (dextrum) atrium, which divides the respective atrial chamber into two.[7,8] Cor triatriatum sinister (CTS) first presenting in adulthood is relatively uncommon. In this report, we describe a case of AF in a young adult in association with CTS.

Timeline

Dec 2019	Patient presented to local emergency department with coryzal illness and syncope. Electrocardiogram (ECG) showed new onset AF. Discharged with outpatient cardiology clinic follow-up
2 weeks later	Cardiology clinic review. The patient was temporarily given rivaroxaban 20 mg daily to prepare for elective electrical cardioversion 8 weeks later
Feb 2020	Successful outpatient DCCV. Discharged from follow-up
July 2021	Patient presented to GP with 6-month history of lethargy. NTproBNP raised. Re-referred to local cardiology service
1 week later	Electrocardiogram showed recurrence of AF and transthoracic echocardiography revealed a new diagnosis of CTS with patent foramen ovale (PFO) and an atrial septal defect (ASD). Initiated on oral anticoagulation with rivaroxaban 20 mg daily and bisoprolol and referred to a tertiary ACHD centre
Oct 2021	ACHD clinic review and referral to congenital cardiac surgeon
Nov 2021	Patient underwent surgical membrane resection, Cox-maze procedure and ASD closure with restoration of sinus rhythm

Case presentation

A 31-year-old male patient, previously fit and active, presented to the local emergency department with coryzal symptoms and syncope whilst walking upstairs. At clinical examination, heart sounds were deemed unremarkable and the chest clear on auscultation. A 12-lead ECG showed new onset rate-controlled AF. Blood tests revealed lymphopenia (0.7×10^9/L) and a raised C-reactive protein of 45 mg/L with normal electrolyte levels and thyroid function. The patient was resuscitated with intravenous fluids and discharged home with an outpatient cardiology clinic appointment. At cardiology review, both stroke risk (CHA_2DS_2-VASc score 0) and bleeding risk (HAS-BLED score 0) were low and the patient was initiated on oral anticoagulation with rivaroxaban 20 mg daily before elective electrical cardioversion 8 weeks later. Symptoms subsequently resolved, and after 4 weeks anticoagulation was stopped and the patient was discharged with a diagnosis of AF in the context of an upper respiratory tract viral illness. The patient's underlying congenital cardiac malformation remained undiagnosed at this point. Two years later, the patient represented to his general practitioner with a 6-month history of fatigue, breathlessness, and reduced exercise tolerance. Laboratory tests revealed a raised N-terminal pro B-type natriuretic peptide of 992 ng/L (0–400 ng/L), prompting a re-referral to the local cardiologist. Physical examination at cardiology assessment revealed resting oxygen saturations of 96% on air, no signs of cardiovascular decompensation, right ventricular lift, exaggeration of the pulmonary component of the second heart sound, and a soft flow murmur at the left sternal edge with no audible diastolic murmurs. A 12-lead ECG showed a recurrence of AF (*Figure 1A*). The patient was initiated on oral anticoagulation with rivaroxaban 20 mg daily and bisoprolol 2.5 mg daily. A transthoracic echocardiogram (TTE) revealed a membranous structure within the left atrium (*Figure 1C*), dividing the chamber into two parts, with a small communication (12 mm) in the membrane and continuous blood flow across it, with a peak gradient of 15 mmHg and mean gradient of 6 mmHg suggestive of moderate obstruction. The proximal high-pressure chamber within the left atrium was dilated and received drainage from all pulmonary veins. There was also a small ASD between the high-pressure chamber and the right atrium, and a PFO between the distal low-pressure chamber and the right atrium. The left ventricle was normal in size and function [ejection fraction (EF) 56%]. The right ventricle was normal in size with mild systolic impairment [tricuspid annular plane systolic excursion (TAPSE) 13 mm] and an estimated systolic pressure of 30 mmHg. Cardiac computed tomography revealed normal coronary artery anatomy without disease and confirmed normal pulmonary venous drainage. The patient was referred to an adult congenital heart disease centre and 4 months later underwent corrective surgery; resection of the fibromuscular membrane, resection and oversewing of the left atrial appendage, closure of the ASD, and Cox-maze surgical cryo- and radiofrequency ablation. A post-operative ECG revealed normal sinus rhythm. A subsequent TTE revealed no residual membrane in the left atrium (*Figure 2*), laminar flow within the left atrial cavity, no residual ASD or PFO, normal left ventricular size and systolic function (EF 68%), mildly dilated right ventricle with impaired longitudinal function (TAPSE 10 mm) but preserved radial function, mild to moderate tricuspid regurgitation with estimated right ventricular systolic pressure of 38 mmHg, and no pericardial effusion. The patient was discharged home in sinus rhythm. At 3-month follow-up, the patient had remained in sinus rhythm, felt improved and had returned to baseline functional status. He will discontinue anticoagulation at 6 months from surgery.

Discussion

We herewith report a case of new onset AF in a young adult related to underlying CTS in whom sinus rhythm was restored and symptoms resolved after surgical correction of the lesion. Our case highlights that AF in early adulthood is a marker of underlying undiagnosed conditions and thus requires the clinician to 'dig deeper', providing thorough cardiovascular assessment as well as symptomatic management, to avoid missing causative diagnoses. The ESC guidelines recommend a 'standard package' for the diagnostic evaluation of AF, which includes a complete medical history and assessment of symptoms, 12-lead ECG, laboratory tests (thyroid and kidney function, electrolytes, and full blood count), and transthoracic echocardiography for assessment of biventricular size and function, left atrial size, and structural abnormalities.[9] In this case, a more thorough initial diagnostic evaluation may have avoided delay in identifying the causative underlying cardiac malformation CTS, a rare congenital heart defect accounting for 0.1–0.4% of all patients with congenital heart disease, which normally presents in childhood.[10] Cor triatriatum sinister is characterized by the presence of a fibromuscular membrane within the left atrium, which divides it into a posterior chamber that receives the pulmonary veins and an anterior chamber

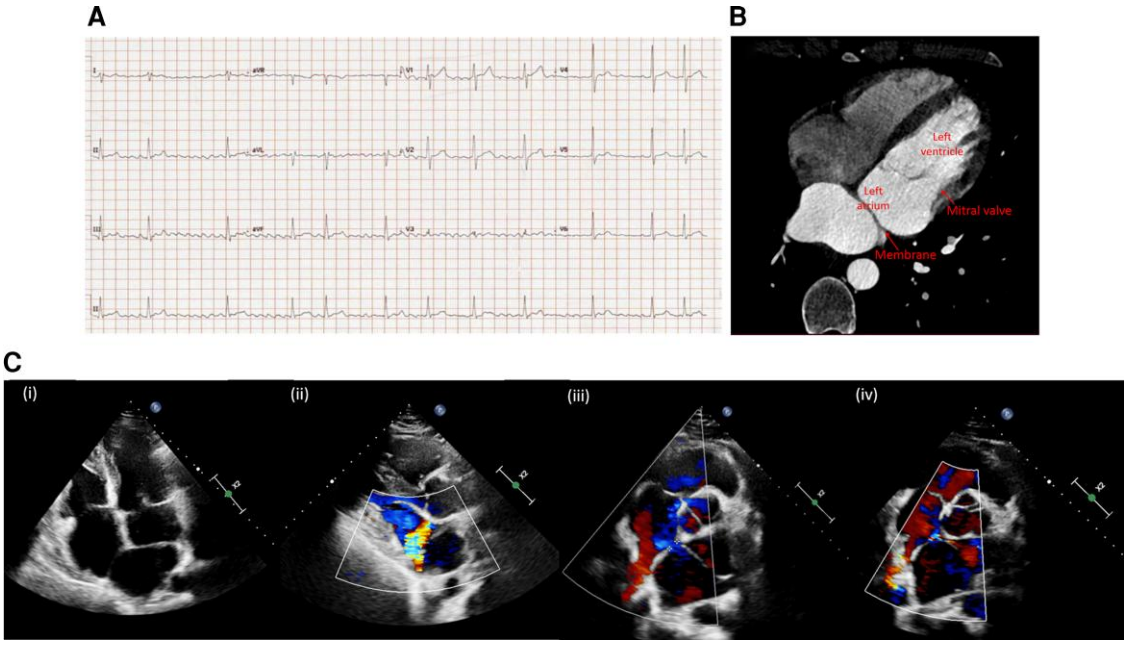

Figure 1 (*A*) Electrocardiogram showing recurrence of atrial fibrillation with left posterior fascicular block. (*B*) Computed tomography image showing fibromuscular membrane dividing the left atrium into two chambers. (*C*) Transthoracic echocardiographic images: (i) apical four-chamber view and (ii) parasternal long axis view with colour Doppler, showing membrane in left atrium causing obstruction with turbulent flow. Parasternal short-axis views with colour Doppler showing (iii) atrial septal defect between proximal left atrial chamber and right atrium and (iv) patent foramen ovale between distal left atrial chamber and right atrium.

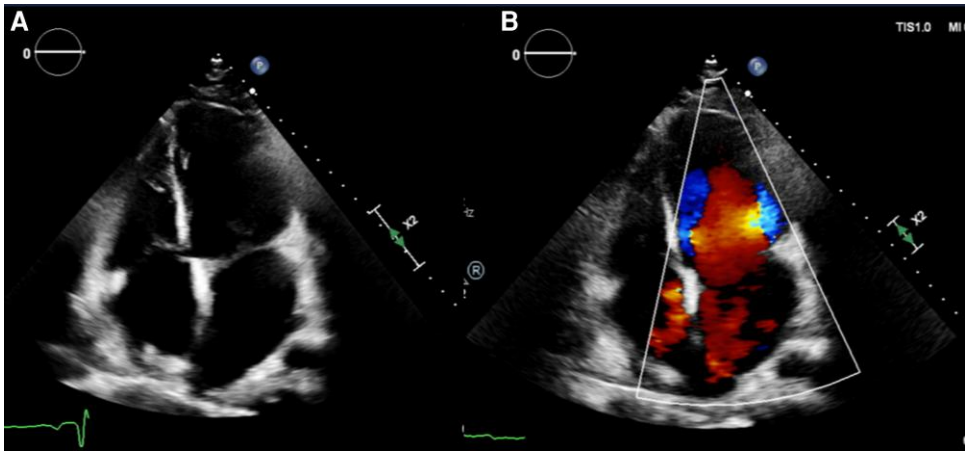

Figure 2 Post-operative echocardiographic images: (*A*) apical four-chamber view showing successful resection of the fibromuscular membrane, (*B*) apical four-chamber view with colour Doppler showing laminar flow within the left atrium.

which communicates with the left atrial appendage and atrioventricular junction.[7,8] Embryologically, CTS is thought to arise from incomplete resorption of the common pulmonary vein thus leaving a membrane or ridge of tissue within the left atrium.[11] Defects in the dividing fibromuscular membrane allow communication between the two chambers, the size of which determines the time of presentation, nature, and severity of symptoms.[12] Larger less obstructive communications are more likely to be tolerated well for several years with few or no symptoms, and in this case the lesion was 12 mm in diameter with moderate obstruction to blood flow. In addition, blood flow through defects in the membrane occurs in both systole and diastole, and thus even higher degrees of obstruction can be tolerated for longer periods of time.[11] Later, conversion to a symptomatic state in adulthood may be precipitated by fibrosis and calcification of the membranous defect resulting in progressive obstruction, or the development of mitral regurgitation or AF, the latter evident in this case.[11,13] When symptoms do arise, the presentation mimics mitral stenosis, with pulmonary venous obstruction and raised pulmonary capillary pressures,

causing pulmonary oedema and symptoms of breathlessness and orthopnoea, or symptoms related to AF. In a recent systematic review including 171 cases of CTS presenting in adulthood, the majority of patients were symptomatic at the time of diagnosis ($n = 141$; 82.5%), most with symptomatic AF ($n = 56$; 32.8%).[14,15] The definitive treatment for CTS is surgical resection of the membrane through midline sternotomy with concomitant intervention for AF, giving an excellent survival and low AF recurrence rates.[16]

Conclusion

New onset AF should prompt a thorough clinical assessment to identify risk factors and potential new underlying cardiac diagnoses, which can be treated with excellent outcomes. Cor triatriatum sinister is a rare congenital anomaly, which infrequently presents in adulthood and can give rise to a clinical presentation, which mimics mitral stenosis, including the development of AF. Patients should be referred to and managed at a tertiary adult congenital heart disease centre, where surgical correction should provide excellent short- and long-term outcomes.

Lead author biography

Dr Abigail Masding graduated from the Hull York Medical School in 2013. She is currently a Cardiology specialist registrar in Adult Congenital Heart Disease and has an interest in medical education.

Supplementary material

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

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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 has been obtained from the patient in line with COPE guidance.

Conflict of interest: None declared.

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