Acute myocardial infarction in a patient with congenitally corrected transposition of the great arteries and complex coronary anatomy—a case report

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Background
Congenitally corrected transposition of the great arteries (ccTGA) is a rare congenital heart anomaly with atrioventricular and ventriculoarterial discordance that is often associated with other cardiac and coronary artery anomalies. Here, we report a case of a patient with ccTGA and non-ST elevation myocardial infarction (NSTEMI) with challenging coronary anatomy that was treated with stress-perfusion cardiac magnetic resonance imaging (spCMR) guided percutaneous coronary intervention (PCI).

Case summary
A 46-year-old male smoker with ccTGA, dyslipidaemia, diabetes Type 2 managed with dietary restrictions and a family history of premature myocardial infarction, presented with typical chest pain, elevated cardiac troponin levels and ECG-changes indicative of ischaemia. The patient was diagnosed with NSTEMI and underwent initial urgent coronary angiography (CA) without apparent significant stenosis, although the right coronary artery (RCA) could not be selectively investigated. The patient had coronary anatomy 1R-2LCX according to the Leiden convention, which is the usual anatomy in patients with ccTGA. Despite this, CA was challenging due to the different anatomy compared with individuals with normally positioned great vessels. The patient remained highly symptomatic with chest pain at moderate exertion. To improve identification of the anatomic location and extent of ischaemia, we performed spCMR with adenosine. This revealed a limited septal infarction (likely embolic) in the right ventricle and reversible ischaemia in two inferior right ventricular segments. A second angiography, selectively investigating RCA demonstrated a significant stenosis in the distal RCA that was successfully treated with a drug-eluting stent. Fractional flow reserve (FFR) measurements of the left coronary arteries demonstrated hemodynamically non-significant stenosis. The patient’s symptoms resolved, and he remained asymptomatic at one month follow-up.

Discussion
This ccTGA patient had multiple risk factors for coronary artery disease and presented with NSTEMI. Diagnosis and treatment were challenging due to complex cardiac anatomy and associated different origins of the coronary arteries. We highlight the importance of careful evaluation of the coronary anatomy and functional testing using for example spCMR and FFR to target the culprit coronary vessel(s) in ccTGA complicated by NSTEMI.

Keywords
Case report • Acute myocardial infarction • Congenitally corrected transposition of the great arteries • Coronary artery anomalies

ESC Curriculum
2.1 Imaging modalities • 2.3 Cardiac magnetic resonance • 3.2 Acute coronary syndrome • 3.4 Coronary angiography • 9.7 Adult congenital heart
Learning points

- In patients with congenitally corrected transposition of the great arteries (ccTGA), aortography during the index coronary angiography or computed tomography coronary angiogram before a planned coronary angiography may be needed due to different anatomy or anomalous origins of coronary arteries.
- Stress-perfusion cardiac magnet resonance imaging is useful when anatomical and haemodynamic information is needed in selected cases of acute coronary syndrome with complex congenital heart disease.

Introduction

Congenitally corrected transposition of the great arteries (ccTGA) is a rare congenital heart anomaly accounting for 0.5% of all congenital heart diseases. It is characterized by double discordance: The right atrium is connected to left ventricle (LV) that in turn supports the posteriorly originating pulmonary trunk. Conversely, the left atrium is connected to the right ventricle (RV) that in turn supports the anteriorly originating aorta. This ventricular inversion puts the RV and tricuspid valve against the systemic pressure, commonly resulting in progressive deterioration in RV function.

Furthermore, ccTGA is often associated with other cardiac anomalies such as ventricular septal defect and pulmonary artery stenosis. On the other hand, up to 10% of ccTGA cases are isolated without other associated anomalies and can remain asymptomatic for several years. As a result, these patients may grow up and develop diseases common in the general population such as coronary artery disease (CAD) and diabetes.

Significant coronary artery anatomic variations associated with ccTGA have been described in the literature. Left dominant coronary circulation with the left circumflex artery (LCX) and the intermediate artery originating from the left sinus through separate ostia, and the left anterior descending artery (LAD) originating from the proximal part of the right coronary artery (RCA) through a single ostium from the right coronary sinus were reported. In these patients, revascularization may be particularly challenging in the setting of acute myocardial infarction.

Given the rare condition, there are not many reports on how to manage ccTGA patients with anatomic variations of coronary arteries and myocardial infarction. Furthermore, knowledge about special characteristics this population may have is still lacking. Therefore, we report a case of acute myocardial infarction in a patient with ccTGA in the fifth decade of his life.

Timeline

| Date       | Event                                                                 |
|------------|----------------------------------------------------------------------|
| 1976       | Congenitally corrected transposition of the great arteries (ccTGA) diagnosed at birth |
| 2015       | Transoesophageal Echocardiography displayed atrial septal defect (ASD) 22 × 16 mm with significant left-right shunt. Patient complained of exertional dyspnea and demonstrated reduced exercise capacity. |
| 2015       | Amplatzer device implantation for ASD                                  |
| 3/8/2020   | Hospital admission with non-ST elevation myocardial infarctionEchocardiography and primary coronary angiography (inconclusive) |
| 6/8/2020   | Stress-perfusion cardiac magnetic resonance imaging (spCMR) performed  |
| 7/8/2020   | Multidisciplinary heart team discussion and secondary angiography with spCMR and fractional flow reserve targeted percutaneous coronary intervention |

Case report

A 46-year-old male smoker with ccTGA and moderately impaired systolic function of the systemic RV, atrial septal defect (ASD) secundum closed with Amplatzer device at the age of 41 (Figure 1A), diabetes Type 2 managed with dietary restrictions, dyslipidemia and a family history of premature CAD, presented with one week history of exertional chest pain with accelerating symptoms the last 2 days before admission. At the time of admission to the hospital, the patient did not take any prescribed medications. High-sensitive cardiac troponin T levels were 620 ng/L (reference: <15 ng/L) at presentation and ECG showed novel anterior ischaemic changes (Figure 2A). The patient was diagnosed with NSTEMI and received a loading dose of acetylsalicylic acid (500 mg) and ticagrelor (180 mg) within hours after presenting to the emergency department followed by 75 mg qDay and 90 mg q12 hr, respectively. A chest computed tomography scan excluded aortic dissection and pulmonary embolism. The patient was transferred to a tertiary heart hospital for further management including urgent coronary angiography (CA).

Upon arrival, a physical examination was unremarkable: Blood pressure was 110/78 mmHg, heart rate regular at 65 bpm, respiratory rate was 14 breaths per minute and saturation 97% (without oxygen supplement). Body temperature was 36.9°C. High-sensitive cardiac troponin T was elevated with a clear rise and fall day 1–3 (620–580–20 ng/L). The patient remained highly symptomatic with recurrent chest pain already at light exertion for which he received nitroglycerin spray with prompt effect.

A transthoracic echocardiogram (TTE) revealed reduced systolic function of the systemic RV with right ventricular ejection fraction (RVEF) 29%, and global hypokinesia more pronounced in the apex, but the reduced image quality made it difficult to exclude regional hypokinesia in all segments. A previous TTE from 2018 showed RVEF of 30%, and right ventricular end-diastolic diameter of 60 mm. The present TTE was therefore considered unchanged compared with the one 2 years ago. Both TTEs also showed mild tricuspid valve regurgitation and normal longitudinal function of the subpulmonary LV.

An initial urgent CA with radial access did not display any obvious significant stenosis. CA was challenging due to the different way coronary arteries originated from the transpositioned aorta compared with individuals with normally aligned great vessels. The left main artery (LM) originated from the right sinus of Valsalva and was difficult to engage. LAD and its diagonal branch had atherosclerotic plaque formation of unclear significance and fractional flow reserve (FFR) attempts were unsuccessful due to engagement difficulty. LCX was rudimental and could not be investigated selectively. Non-selective images and
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Aortography images were taken instead and they did not show stenosis in the proximal segments of RCA (Figures 3A and B). To better illustrate the coronary anatomy, we provide a schematic visualization of the coronary anatomy in normal hearts and our patient (Figure 4).

The procedure was abandoned at this stage. Since the patient was symptomatic and a conclusive revascularization strategy was not achieved, we decided to further evaluate the anatomical location and target vessel of any ischaemia using stress-perfusion cardiac magnetic resonance imaging (spCMR) with adenosine. spCMR was considered superior to other imaging modalities due to its capability to demonstrate the anatomical and functional burden of ischaemia in patients with complex congenital heart anatomy.

spCMR showed a recent anteroseptal infarction at the basal segment of the systemic RV (Figure 1B) and reversible ischaemia in two segments at the basal and mid-ventricular inferior segments of the systemic RV (Figures 1C and D). The ischaemic region composed about 7% of RV which was mildly dilated and normotrophic with global hypokinesia (more pronounced in the septum) and RVEF 28%. At multidisciplinary heart team conference, it was decided to do another try with CA targeting the RCA.

A second CA with right femoral access could engage RCA selectively with a 6 french Judkins left 4 (JL4) catheter and revealed a large vessel with significant stenosis 90% in the distal RCA proximal of its bifurcation (Figure 3C). RCA also had two right ventricular branches, of which the distal one had atheromatous changes in its orifice. Thin collateral flow

Figure 1 Cardiac magnetic resonance images of the patient. (A) Systemic RV and Amplatzer ASD device indicated by arrow. (B) Late gadolinium enhanced short axes view at the basal level showing systemic RV, LV, and a minimal anteroseptal infarction (likely embolic) with microvascular obstruction (arrow). (C) Perfusion image of the systemic RV at rest. (D) Perfusion image of the systemic RV during quantitative spCMR with adenosine. Arrows highlight hypoperfused infero-septal segments of the RV. We want to point out that the RV seems dilated during stress (D) compared with the rest image (C). However, there was no right ventricular dilatation when comparing right ventricular volumes in four-chamber view and therefore this is likely explained by patient movement or excessive breathing during stress and not by right ventricular dilatation.
from RCA to the left coronary system was also shown. The LM was engaged with left coronary bypass angiographic catheter and internal mammary artery catheter. FFR measurements were taken of both LAD and diagonal arteries and showed haemodynamically non-significant stenosis (LAD-FFR 0.85, diagonal-FFR 0.82). FFR measurement is a diagnostic method used in CA to determine the significance of a coronary artery stenosis by measuring the pressure differences at hyperaemia before and after the stenosis using a pressure wire, and it predicts the percentage improvement in flow with percutaneous coronary intervention (PCI) where values > 0.80 are associated with non-significant stenosis. PCI was performed at the RCA stenosis using JL4 guide catheter. After balloon predilatation, a 3.5 × 16 mm drug-eluting stent was implanted and post dilated with non-compliant balloons 4.0 × 12 mm and 4.5 × 8 mm to 20–22 atmosphere. The final angiographic image of the RCA after PCI showed a flow of grade 3 according to the thrombolysis in myocardial infarction (TIMI) flow grading system (Figure 3D). At discharge, the patient was offered smoking cessation counselling and prescribed acetylsalicylic acid 75 mg qDay, Ticagrelor 90 mg q12hr, atorvastatin 80 mg qDay, metformin 500 mg q12hr, empagliflozin 10 mg qDay, and enalapril 5 mg qDay.

The systemic RV dysfunction was unchanged compared with 2 years before the NSTEMI described in this case suggesting that the acute myocardial infarction was not solely responsible for systemic RV dysfunction. However, the poor systemic RV function is of concern, and follow-up plan for this patient included repeated cardiac magnetic resonance imaging (CMR) for quantification of RV function, NT-proBNP and symptom evaluation. Unfortunately, there is no evidence-based pharmacological treatment for RV dysfunction in cCTGA patients.3

At six-week’s follow-up, the patient had a NT-proBNP of 188 ng/L (compared with 384 at admission), symptoms consistent with NYHA class II and no exertional chest pain. TTE showed RVEF 30%, right ventricular end-diastolic diameter 61 mm, normal longitudinal function of the subpulmonary LV, and mild tricuspid valve regurgitation.

Discussion

Patients with isolated cCTGA can grow up asymptomatic and acquire diseases common in the general population. In this case, the patient had several cardiovascular risk factors that contributed to the development of CAD and acute myocardial infarction. Moreover, the deviating anatomy was a challenging factor leading to delayed revascularization. Due to extreme rarity, there are only a few reported cases with cCTGA and CAD. In addition, coronary artery anomalies and associated cardiac anomalies can pose challenges in diagnosing and treating this type of patients, highlighting the importance to enrich our knowledge about these rare cases.

It is worth mentioning that the anteroseptal right ventricular infarction had an appearance in spCMR that was likely embolic, but with unclear embolism source. It was characterized by a distinct focal late gadolinium enhancement signal with a transmural oedema and a core of microvascular obstruction. This pattern was typical for embolic myocardial injury. Furthermore, a thrombotic injury would likely cause a subendocardial lesion and we did not observe atherosclerosis in the coronary vessel targeting the affected RV segment. Together, this suggests that the anteroseptal myocardial infarction was due to an embolic event. Several mechanisms may explain the embolic insult but atrial fibrillation and paroxysmal
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Supraventricular tachycardias are common in ccTGA. Although the mechanism is unknown in this particular patient, asymptomatic paroxysmal atrial tachycardia is the most likely explanation.

The usual coronary artery anatomy in patients with ccTGA, double outlet RV or pulmonary atresia, and usual atrial arrangement was described in a gross morphological study of 14 hearts. RCA was described to arise from sinus 1 (right-handed facing), as seen from the non-facing aortic sinus from a surgical point of view (Figure 4B). While LM and its branches were described to arise from sinus 2 (left-handed facing).

Moreover, 20 specimens with ccTGA, double outlet RV, and right ventricular aorta with pulmonary atresia were examined for coronary artery anomalies. Approximately 45% of specimens had coronary artery anomalies, such as LAD arising from RCA with LCX arising alone directly above the intercoronary commissure, and a single trifurcated coronary artery.

Our patient most likely had the usual coronary artery anatomy with 1R-2LCX formation according to the Leiden convention (Figures 4B and D). The usual coronary artery anatomy in normally aligned great arteries is shown in Figures 4A and C. However, we could not be sure about the exact coronary artery anatomy of our patient because computed tomography coronary angiogram (CTCA) was not performed. CTCA is the best method to show the exact origin of the coronary arteries, but it lacks the ability to provide hemodynamic information. We therefore chose, giving the urgent situation, to do a spCMR instead. We

![Figure 3](image-url) Coronary angiogram of the patient. Left (A) and unselective proximal right (B) coronary artery at the initial coronary angiography. (C) Right coronary artery with significant stenosis in distal portion (indicated by arrow). (D) Right coronary artery after PCI.
were able to reconstruct an image from the magnetic resonance imaging displaying the origins of the coronary arteries (Figure 5). However, this visualization is limited by the lack of many anatomical details such as the cusps and the sinuses of Valsalva which are better visualized by CTCA. Limited availability, comparatively long study times and claustrophobia are other disadvantages of spCMR. On the other hand, spCMR has the ability to provide anatomic and hemodynamic information, as well as the viability of the infarct area should this information be needed.

Further, it can be challenging with catheter engagement of the coronary arteries in ccTGA patients due to different anatomy or anomalous origins. In left anterior oblique view, engaging LM and RCA, in normal hearts, is performed through the left and right aortic sinuses respectively. Whereas in ccTGA patients with the usual coronary artery anatomy, the LM originates anteriorly from the right sinus, and the RCA originates from the posterior sinus. It can be therefore useful to use aortography during the index CA or a CTCA before a planned CA to investigate the origins of coronary arteries.

Finally, patients with isolated ccTGA suffer in the first place from gradual deterioration and dilatation of the systemic RV. Therefore, ischaemic events related to the right coronary circulation should be diagnosed and treated promptly to avoid further decline in RV function.
Conclusion

We presented this case with ccTGA, challenging coronary artery anatomy and multiple risk factors for CAD that suffered an acute myocardial infarction in the systemic RV that was treated successfully with PCI after identification of the suspected target vessel using advanced imaging.

Lead author biography

Mr. Fahd Asaad is a specialist in cardiology with special interest in electrophysiology at Karolinska University Hospital in Stockholm, Sweden. He graduated from medical school in Damascus University, Syria, and after doing his residency in cardiology at Damascus hospitals he moved to Sweden and began his journey in cardiology in the field of electrophysiology and he began his research in atrial fibrillation.

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 patient has provided oral and written consent to publish this case report in accordance with COPE guidelines.

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