A unique case of embolic myocardial infarction in a 22-year-old woman with Fontan circulation

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SÚHRN

Embólia koronárnych artérií je menej častou príčinou akútneho infarktu myokardu. Predstavujeme kazuistiku 22-ročnej pacientky s dvojvtokovou ľavou komorou, defektom komorového septa a pulmonálnou stenózou po Fontanovej operácii s akútnym spodným STEMI na embolickom podklade. Okrem známych rizík vedúcich k trombotickým komplikáciám spojeným s Fontanovskou cirkuláciou, medzi potenciálne predisponujúce faktory trombembolizmu u tejto pacientky patrili vytvorenie trombu v dolnej dutej žile a nedostatočná antikoagulačná liečba. Táto kazuistika predstavuje jednu z prvých dokumentovaných embolických príčin STEMI u pacienta s Fontanovskou cirkuláciou. Doteraz nie je vytvorený konsenzus, že antikoagulačná terapia warfarínom je u pacientov s Fontanovskou cirkuláciou superiórna voči aspirínu v primárnej prevencii trombembolických príhod.

Keywords:
Anticoagulation therapy
Fontan circulation
Myocardial infarction
Trombembolic event

ABSTRACT

Coronary artery embolism is an uncommon cause of acute myocardial infarction. We present a case of a 22-year-old patient with double inlet left ventricle (DILV), ventricular septal defect and pulmonary stenosis after Fontan repair with an acute embolic inferior ST-segment elevation myocardial infarction (STEMI). Apart from already known risk factors of thrombotic complications associated with Fontan circulation, additional predispositions in this patient included thrombus formation located in the inferior vena cava and the lack of anticoagulation therapy. This is one of the first reported embolic causes of STEMI in a patient with a Fontan circulation. Up to date, there is no consensus that anticoagulation therapy with warfarin is superior to aspirin in primary prevention of thromboembolism in patients with Fontan circulation.

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Introduction

Fontan circulation is associated with a higher risk of thromboembolic events. However, the incidence of embolic myocardial infarction (MI) is very rare. We represent a unique case of a young patient after fenestrated Fontan repair, presenting with embolic inferior STEMI. The antithrombotic treatment strategy plays a pivotal role in primary and secondary prevention of thromboembolism.

Case report

We report a case of a 22-year-old woman with double inlet left ventricle, pulmonary stenosis, and ventricular septal defect who underwent superior cavo-pulmonary shunt (Glenn) at the age of one year and atrio-pulmonary (Fontan) connection at the age of 4 years. Recurrent supraventricular tachycardia necessitated multiple electrophysiological studies, which led to ablation of the posteroseptal accessory pathway at the age of 11 years.

In 2017, she referred to the emergency department for acute chest pain radiating to the neck. Examination showed no signs of heart failure, blood pressure was 100/60 mmHg. The ECG showed significant ST segment elevations in limb leads II, III and aVF (Fig. 1).

A transthoracic echocardiogram (TTE) revealed preserved left ventricular ejection fraction (LVEF 50%), hypokinesis of inferior wall, trivial mitral and tricuspidal regurgitation, suspicion of thrombus formation of width 5 mm and length 38 mm in inferior vena cava (IVC) (Figs 2, 3). Her chronic medical therapy included sotalol (2× 80 mg) and acetylsalicylic acid (1× 100 mg). She was transported to the East Slovak Institute of Cardiovascular Diseases and was medicated according to the STEMI guidelines with heparin and dual antiplatelet therapy. Her creatine kinase peaked to 5.72 μkat/l, MB fraction of creatine kinase peaked to 0.46, high sensitive troponin T (hsTn-T) peaked to 0,390 μg/l. NT-proB-type natriuretic peptide was 137,8 pg/ml and D-dimers 0,36 mg/l. The complete blood count was normal. The patient's symptoms, TTE findings with regional wall motions abnormalities and elevation of hsTn-T over 99 percentile were consistent with the definition of acute STEMI. Selective coronary angiography showed normal findings. No coronary artery anomalies were detected (Fig. 4) which raised the possibility of spasm or thromboembolism. Serial electrocardiograms showed the typical changes of acute myocardial infarction (Fig. 5).

Ultrasonographic examination of lower extremities ruled out the possibility of deep venous thrombosis. Cardiac magnetic resonance imaging (CMR) was performed and revealed a patent Fontan pathway, however no thrombus was detected and the pulmonary arborization was normal. The left ventricular ejection fraction was 50%, but the late gadolinium enhancement showed post-ischemic changes after transmural myocardial infarction of the inferior wall (Fig. 6). The patient had a complete hematological examination, which excluded thrombophilia. Recommended therapy on discharge was: acetylsalicylic acid 100 mg, clopidogrel 75 mg, proton pump inhibitor 20 mg, sotalol 2× 80 mg, cardilan 175 mg and atorvastatin 80 mg. Because of the second type of MI and the thrombus formation in IVC documented during the first TTE, antithrombotic therapy was changed in outpatient care for warfarin. During the three years of follow-up, the patient has no signs of thrombotic recurrence.

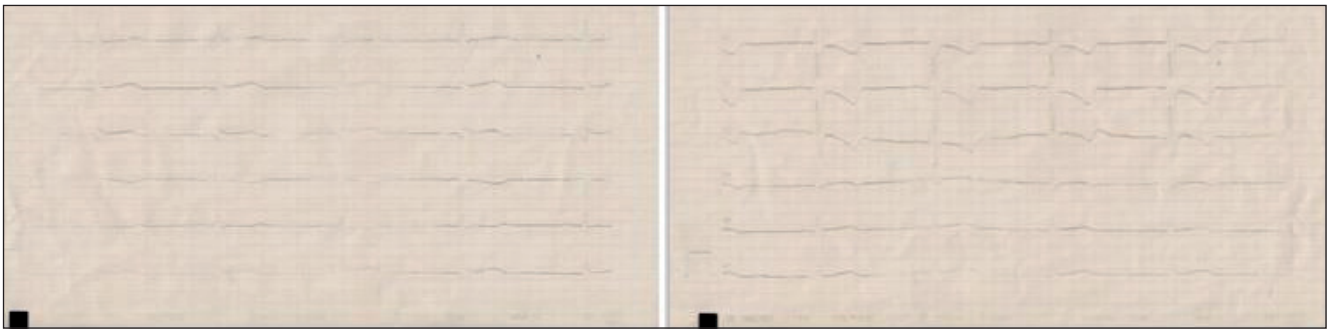

Fig. 1 – ECG showed signs of inferior STEMI.

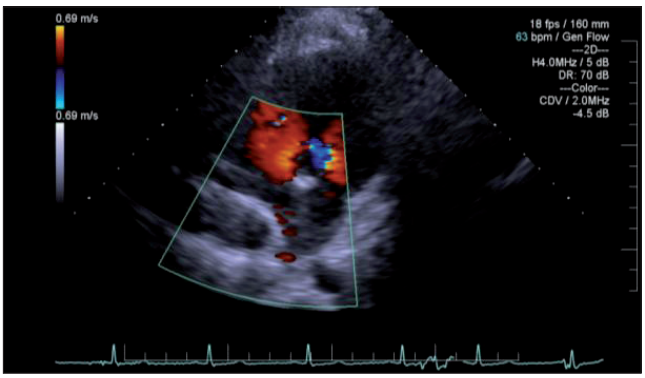

Fig. 2 – Echocardiography – four-chamber view.

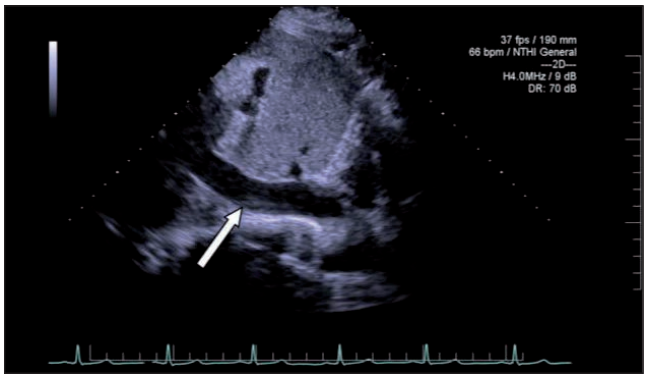

Fig. 3 – Echocardiography – thrombus formation located in inferior vena cava.

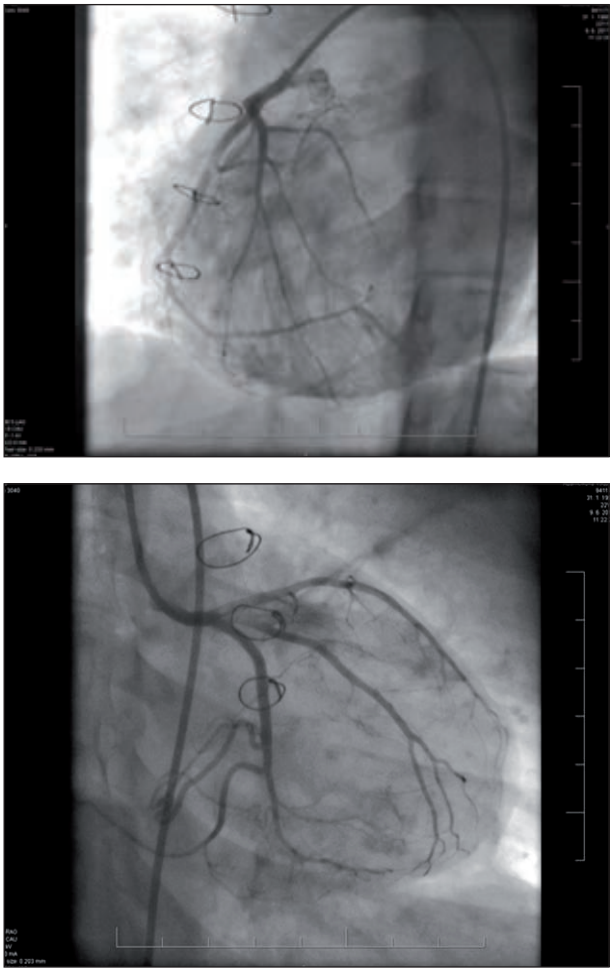

Fig. 4 – Selective coronary angiography – normal finding.

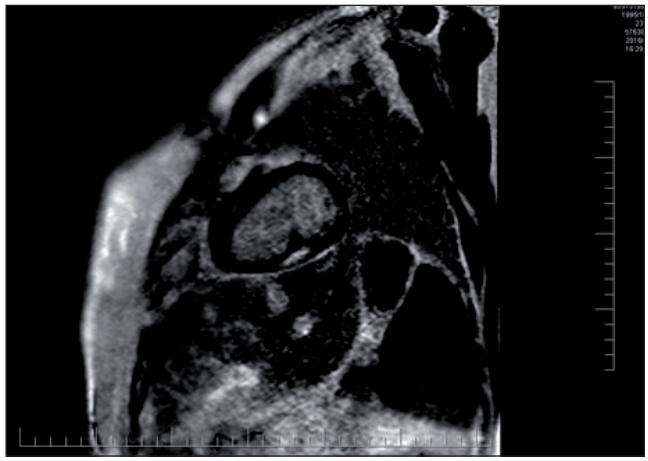

Fig. 6 – Cardiac magnetic resonance imaging (MRI), the late gadolinium enhancement showed the signs of past inferior transmural ischemia.

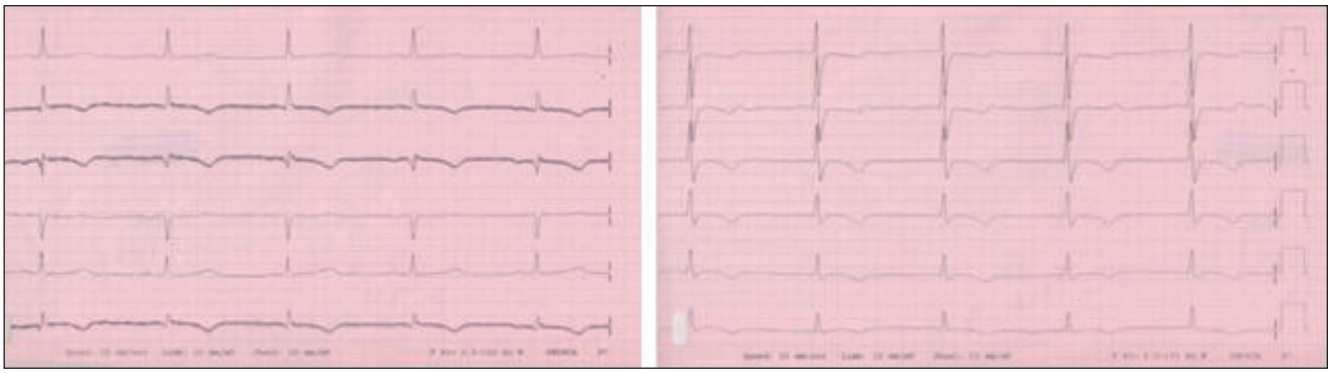

Fig. 5 – ECG after selective coronary angiography.

Discussion

The Fontan procedure was first performed in 1968 in a patient with tricuspid atresia and has transformed the lives of children born with single-ventricle physiology.[1] The earlier Fontan connected the right atrial appendage to the pulmonary artery (PA). The current procedure is an extracardiac conduit going from the inferior vena cava to the PA, and a Glenn connection joining the superior vena cava to the right PA.[2] Most of patients with Fontan circulation are surviving into adulthood. However, due to the abnormal circulation, they have an increased risk of complications. The frequent late complications are arrhythmias, thromboembolism, protein-losing enteropathy, heart failure and plastic bronchitis.[3,4]

The most common etiological factors of thromboembolism are endothelial dysfunction, abnormal blood flow, hypercoagulability, presence of a right-to-left shunt.[5] The recognised locations of thrombus embolisation are caval veins, right atrium, pulmonary artery, venous chamber and cerebral arteries.[6] MI due to embolic occlusion of coronary arteries post-Fontan surgery is very rare and was described only in a few patients; with repaired hypoplastic left heart syndrome, with history of double outlet right ventricle, with ventricular septal defect, with pulmonary atresia and severe right heart hypoplasia who underwent extra-cardiac Fontan operation. Table 1 shows a summary of published case reports dealing with this topic.

We present a unique case of inferior embolic STEMI in a 22-year-old patient with fenestrated Fontan circulation due to double inlet left ventricle, ventricular septal defect and pulmonary stenosis. The potential source of embolism was the thrombus formation located in the IVC. We suppose that after the heparin and dual antiplatelet treatment in the acute phase of MI, the coronary

Table 1 – Summary of case report's findings of embolic MI incidence after Fontan procedure

Author	Basic diagnosis before FP	Type of MI	Age in admission	Antitrombotic treatment used before admission/ after discharge	Recurrence of trombembolic event reported
Hastings et al.[14]	Pulmonary atresia, severe right heart hypoplasia	Inferior STEMI	19 yo	Aspirin/Combined aspirin and warfarin therapy	No
Deshpande et al.[15]	Tricuspid atresia, coarctation of the aorta, single ventricle	Anterolateral STEMI	17 yo	NA/NA	NA
Subahi et al.[16]	Hypoplastic left heart syndrome	Anterolateral STEMI/the only one with stent implantation	10 yo	Aspirin/aspirin, clopidogrel and warfarin	No
Shamoon et al.[17]	Hypoplastic left heart syndrome	Inferolateral STEMI	24 yo	NA/aspirin, ticagrelol and warfarin	NA
Noonan et al.[18]	Hypoplastic left heart syndrome	Inferior STEMI	3 yo	None/aspirin and warfarin	NA
Wilson et al.[19]	Atresia of tricuspid valve, ventriculoparterial discordance, coarctation of the aorta	Anterior STEMI	3 yo	Aspirin/low dose aspirin and warfarin	Yes
Meier et al.[20]	Double outlet right ventricle, ventricular septal defect and straddling atrioventricular valves	Lateral STEMI	21 yo	NA/NA	NA

MI – myocardial infarction; FP – Fontan procedure; NA – not available; STEMI – ST segment elevation myocardial infarction; yo – years old.

bed was without any residual thrombotic occlusions. Moreover, MRI and ultrasound examinations did not confirm any residual vascular or intracardiac thrombus formation. However, late gadolinium enhancement showed post-ischemic signs of inferior transmural MI. The thrombus formation could have been caused by the sluggish circulation and stagnation as a result of the absence of the right ventricular pump. Alterations of coagulant factors are also one of the explanations, but the hematological examination did not reveal any anomaly. To date, however, there has been no reported difference in the incidence of thrombotic events between fenestrated and non-fenestrated Fontan circuits in recent trials.[7] Based on the patient's medical records obtained from her pediatrician, she underwent indicated fenestrated Fontan procedure. Fenestration of Fontan circuit decreases postoperative morbidity and mortality rates in high-risk patients. It appears to result in a lower incidence of arrhythmia. Moreover, baffle fenestration performed at the time of Fontan surgery improves short-term outcomes in standard-risk patients by decreasing pleural drainage, length of hospital stay, and need for additional postoperative procedure.[8,9] The closure of Fontan fenestration was not indicated. A persistent fenestration may be a benefit, as lower central venous pressure may decrease the risk of exercise intolerance, protein losing enteropathy, plastic bronchitis and bradyarrhythmias. In previous publications, fenestration closure was not associated with higher event-free survival.[10]

The thromboprophylaxis after Fontan repair is very important to prevent thromboembolism.[11] Up to date, there was no significant evidence proving that anticoagulation with warfarin is superior to aspirin preventing thromboembolic events in patients with Fontan circulation.[12,13] However, in our case, there was thrombus formation, while on antiaggregation monotherapy with aspirin. Therefore, the treatment with warfarin was started and during a period of three years of anticoagulation therapy, there was no evidence of any thrombotic recurrence. Anticoagulation monotherapy was chosen instead of simultaneous treatment with warfarin and aspirin used in previous reported cases, because of the higher risk of bleeding. In case of future recurrence of thromboembolism despite adequate antithrombotic treatment, the consideration of fenestration closure may be needed in secondary prevention of other embolic episodes. Specific fenestration management guidelines might help in future decision making.

Conclusion

The main goal of this case report is to highlight the potential need of anticoagulation therapy instead of antiplatelet treatment in primary and secondary prevention of thromboembolic complications after Fontan repair. There is a need for a large multicentered randomized control trials comparing the two treatment strategies and focusing on the perspective of new oral anticoagulants. Secondary, we want to emphasize, that acute MI of embolic origin is one of the very rare and life threatening complications of Fontan circulation, even in very young patients, regardless of the primary diagnosis requiring Fontan operation in childhood.

Conflict of interest
Authors declare no conflict of interests.

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