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CASE REPORT

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Successful complex percutaneous intervention in patient with Fontan circulation and severe heart failure: A case report

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Abstract

We report the case of a successful complex percutaneous intervention in a patient with Fontan circulation and severe heart failure. The patient presented with cyanosis; Fontan conduit stenosis was detected, and the fenestration was patent. The complex interventional procedure allowed for a long-term stabilization of the patient's condition.

KEYWORDS

case report, congenital heart disease, Fontan circulation, heart failure, percutaneous intervention

1 | INTRODUCTION

The Fontan operation was introduced in 1968 and has become the definitive, palliative treatment for patients with functionally univentricular heart. The surgery separates pulmonary and systemic circulation by the creation of a venous-to-pulmonary connection for the passive delivery of deoxygenated blood into the lungs and utilization of the single functional ventricle to maintain systemic circulation. Establishment of a Fontan circulation typically requires multiple stages of repair.[1]

Over the past several decades, management of patients with congenital heart disease (CHD) has evolved continuously. Out of several methods that developed significantly, cardiac catheterization is one of the most important tools used in patients with CHD. Having evolved from a diagnostic tool to a therapeutic option for many conditions that previously required cardiac surgery, percutaneous

interventions are now an integral and indispensable part of the management of pediatric and adult CHD.[2]

The development of heart failure (HF) is a common problem in CHD, with an incidence of 20–50% in the adult population. In patients with a Fontan circulation, virtually all will develop some form of HF in the years following the operation.[1]

We report a unique case of a successful complex percutaneous intervention in a patient with a Fontan circulation and severe heart failure. A highly abbreviated version of the clinical case was presented in the Clinical Cases session during the EuroEcho 2021 conference.[3]

2 | CASE REPORT

A 17.5-year-old woman with tricuspid atresia (TA) was admitted to the Department of Cardiology and Congenital Diseases of Adults for cardiovascular evaluation and treatment optimization. She had a history of multiple palliative procedures, including a Blalock-Taussig shunt surgery in the first month of life, a bidirectional Glenn procedure 1 year later and an extracardiac conduit (Gore-Tex) Fontan procedure with fenestration at the age of six. She also had a dual-chamber cardiac pacemaker with epicardial electrodes, which had been implanted in the second year of her life due to third-degree atrioventricular block and has been replaced 4 times since then. Unfortunately, she did not have regular follow-up visits at the reference center in the years 2017–2021, resulting in a significant gap in her cardiology care. In addition, her last cardiac catheterization was performed shortly before the Fontan procedure (in 2009) and she missed her scheduled control catheterization (during which the closure of fenestration would have been considered).

On admission, she reported no symptoms at rest and was unaware of the severity of her illness. However, on physical examination, she had clinical features of central and peripheral cyanosis. Her oxygen saturation was 88% with a heart rate of 72 bpm and blood pressure of 112/65 mmHg. Her pharmacological treatment consisted of carvedilol 6,25 mg, lisinopril 2.5 mg, and spironolactone 25 mg per day.

Routine laboratory tests showed hemoglobin at 15.9 g/dL, red blood cells at 4.96×10^6/μL, hematocrit at 45.4%, and white blood cells at 6.27×10^3/μL. Alanine transaminase (ALT), aspartate transaminase (AST), C-reactive protein (CRP), ionogram, lipid profile, albumin level, and estimated glomerular filtration rate (eGFR) were normal. NT-proBNP was significantly elevated (1046 pg/mL).

Transthoracic echocardiography (Figure 1 and Video S1, Video S2) showed left ventricular hypertrophy with severe systolic dysfunction and an ejection fraction of 20%–25%. A large atrial septal defect and a ventricular septal defect were found. The function of the mitral and aortic valves was normal; the tricuspid valve was absent, and the pulmonary valve had degenerated leaflets. The shunt between superior vena cava and right pulmonary artery (RPA) [Glenn shunt] and between inferior vena cava and RPA [Fontan conduit] were visualized. The flow in the former was 0.34/0.47 m/s with a respiratory variation. The flow in the Fontan conduit was 0.27/0.62 m/s with a respiratory variation. Possible degeneration of the Fontan conduit was detected—the specialist suspected stenosis of the conduit. The fenestration was visible and patent. In addition, there were prominent trabeculations and multiple intertrabecular recesses in the apex of the left ventricle. These findings suggested a co-occurrence of noncompaction cardiomyopathy.

Twenty-four-hour Holter electrocardiography showed a paced rhythm (with atrial sensing-ventricular pacing domination) and ventricular arrhythmia with 5 episodes of non-sustained ventricular tachycardia (maximum of 8 consecutive beats) in addition to 5549 ventricular extrasystolic beats. There were only 10 supraventricular extrasystolic beats.

Cardiac catheterization is recommended for all patients after Fontan operation in case of cyanosis (2020 ESC Guidelines for the management of adult congenital heart disease; recommendation class I, level C[1]). Therefore, the patient was qualified for the procedure. During the catheterization, a narrow (11.2 mm) extracardiac Fontan conduit was found along with a 4 mm fenestration (Figure 2 and Video S3). This finding confirmed the suspicion from the transthoracic echocardiography—the Fontan conduit was stenosed. There was no thrombosis or calcification

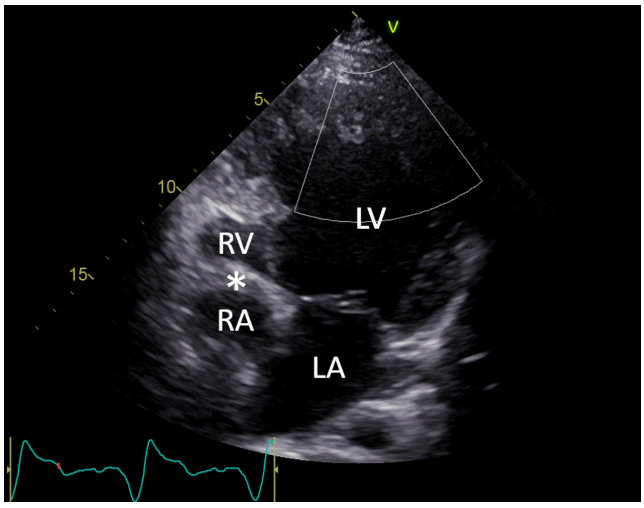

FIGURE 1 Transthoracic echocardiography, apical view. Visible left ventricular hypertrophy, hypoplastic right ventricle and absent tricuspid valve (marked as *). RV, right ventricle; RA, right atrium; LV, left ventricle; LA, left atrium.

typical of homografts. The main pulmonary artery (PA) was anatomically connected to the ventricular outflow tract of a hypoplastic right ventricle, but it was wide and a hypoplastic RV was strongly contrasted. The pressures measured in the Fontan circulation were within normal limits—in the Fontan conduit: 16/13/13 mmHg; in the superior vena cava (SVC): 10/12/11 mmHg; and in the left pulmonary artery (LPA): 14/8/10 mmHg (systolic pressure, mean pressure, and diastolic pressure, respectively). Based on these findings, an appropriate treatment plan was implemented. During the same procedure, two covered stents were implanted in the Fontan conduit (CP COVERED 45 mm/39 mm with use of Balloon-in-Balloon 18/45 mm catheter) and the main PA was closed with use of the Amplatzer Vascular Plug II 20 mm set (Figure 3 and Video S4). The pressures measured in the Fontan circulation after the procedure were still within normal limits: in the Fontan conduit: 14/14/13 mmHg; in the SCV: 13/13/13 mmHg; and in the LPA: 17/9/13 mmHg; the systemic blood pressure was 93/68 mmHg.

There were no postprocedural complications. The patient's oxygen saturation improved to 94%, and no features of cyanosis were found. She admitted that her exercise tolerance had improved. After the procedure, she was started on warfarin anticoagulant therapy with a target INR of 2–3. Postprocedural echocardiography showed a dilated Fontan conduit with a visible stent (Figure 4 and Video S5); the flow was 0.22/0.27 m/s with a respiratory variation. The fenestration was not visible. The closure device was visualized in the main pulmonary artery.

The patient's blood was collected for genetic testing with her informed consent. We performed the genetic cardiomyopathy panel, which included genes correlated with hypertrophic cardiomyopathy (HCM): myosin binding protein C3 (MYBPC3); myosin heavy chain 7 (MYH7); troponin I3, cardiac type (TNNI3); troponin T2, cardiac type (TNNT2); actin alpha cardiac muscle 1 (ACTC1); myosin light chain 2 and 3 (MYL2 and MYL3); phospholamban (PLN); and tropomyosin 1 (TPM1). Genes correlated with

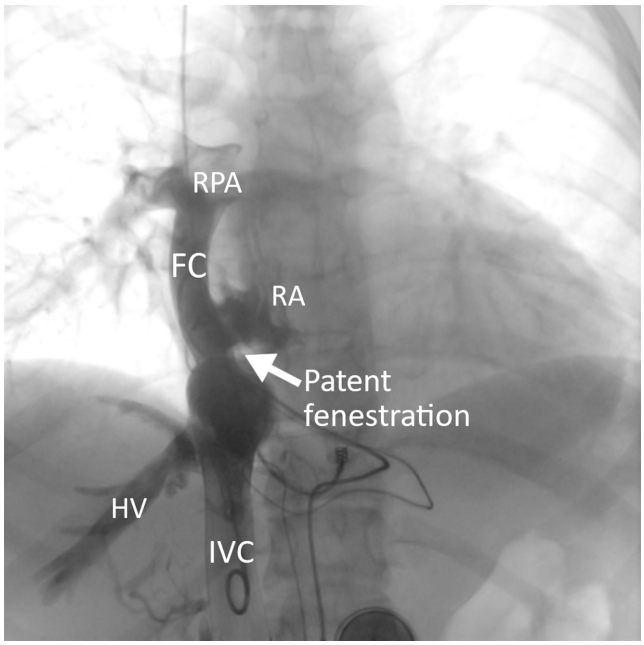

FIGURE 2 Fontan conduit angiography before stenting: a narrow (11.2 mm) extracardiac Fontan conduit and 4 mm fenestration were visualized. FC, Fontan conduit; RPA, right pulmonary artery; RA, right atrium; IVC, inferior vena cava; HV, hepatic veins.

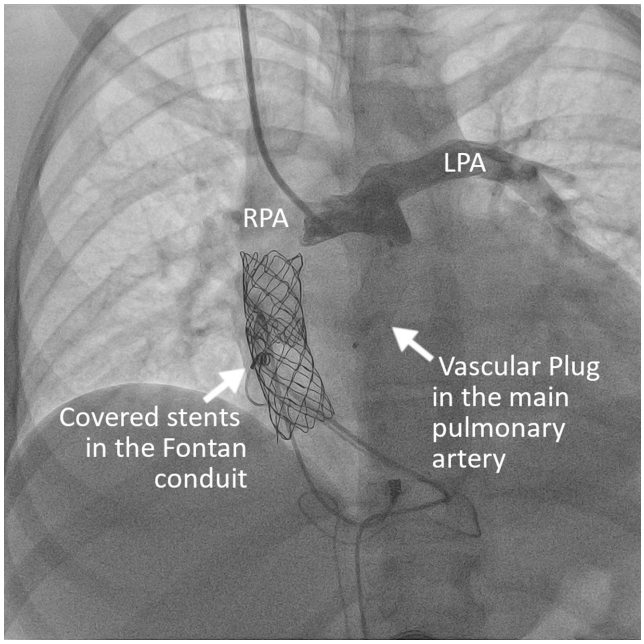

FIGURE 3 Angiogram at the end of the procedure: Fontan conduit after stenting, fenestration and the main pulmonary artery closed. RPA, right pulmonary artery; LPA, left pulmonary artery.

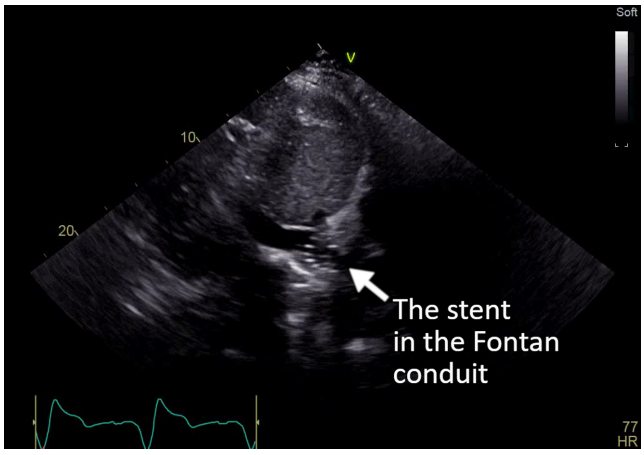

FIGURE 4 Transthoracic echocardiography, subcostal view: the Fontan conduit with the visible stent.

left ventricular noncompaction (LVNC) were also tested: actin alpha cardiac muscle 1 (ACTC1—associated with LVNC type 4); myosin heavy chain 7 (MYH7—associated with LVNC type 5); troponin T2, cardiac type (TNNT2—associated with LVNC type 6), tropomyosin 1 (TPM1—associated with LVNC type 9); and myosin binding protein C3 (MYBPC3—associated with LVNC type 10). All of these tests were negative.

The patient had a follow-up hospitalization 3 months later. Her condition was stable with no exacerbation of symptoms and an EF of 20%. Due to the severity of the patient's condition, she was referred to the transplant center, where she was admitted another 3 months later. There, after a thorough evaluation (including a cardiopulmonary exercise test [CPET]), she was found to be in acceptable condition and the heart transplant was withheld.

One and a half years after the percutaneous procedure, the patient had her latest follow-up hospitalization in our center. She was in good condition, reported no dyspnea, and had no signs of cyanosis. Her treatment had been modified in the outpatient care: sildenafil was started (25 mg/day), and pacemaker parameters were modified (atrioventricular delay was increased). Since then, she reported an improvement of her symptoms. The typical signs of heart failure were absent, and the oxygen saturation was 96%. Transthoracic echocardiography showed severe systolic dysfunction with an EF of 23%, and the flow in the Fontan conduit was 0.29/0.81 m/s with a respiratory variation. The fenestration was still fully occluded by the covered stents, and the closure device in the main pulmonary artery was positioned correctly. The CPET showed that the patient's peak oxygen uptake (VO2peak) was 1.08 L/min and oxygen uptake per kilogram was 18 mL/kg/min, which was comparable to her previous results. In the 6-minute walk test (6MWT), she walked 570 meters. Considering all these facts, the patient's pharmacotherapy was modified with the introduction of dapagliflozin (10 mg/day). In conclusion, the interventional treatment proved to be successful as it allowed for long-term stabilization of the patient's condition and significantly improved her quality of life.

3 | DISCUSSION

Tricuspid atresia (TA) is a complex congenital heart disease that presents with cyanosis in the neonatal period. It accounts for approximately 1% of all cases of CHD, and its prevalence is reported to be ~0.1 per 1000 live births.[4,5] In patients with tricuspid atresia, the right ventricle is hypoplastic; therefore, such a condition can be described as hypoplastic right heart syndrome (HRHS).[6] Patients with TA require multiple cardiac surgeries to maximize longevity. Creation of the Fontan circulation is the definite, palliative management of the univentricular heart. It is a method of redirecting systemic venous blood directly into the pulmonary arteries without passing through the right ventricle. Establishment of a Fontan circulation usually involves multiple stages of repair.[1] In the described case, the patient was diagnosed with tricuspid atresia. She underwent a standard series of procedures in order to create the Fontan circulation. This palliative surgery allowed her to survive and have a relatively good quality of life.

Heart failure can be defined as the inability of the heart to meet resting and exercise demands at low filling pressures. By such a definition, essentially all patients with Fontan circulation have a physiological form of chronic heart failure from the first day after the operation.[7] The 10-year survival rate may approach 90%, but it should be appreciated that a premature decline in cardiovascular performance, with reduced survival, is inevitable even in the best Fontan patients.[1] Systolic ventricular function is relatively preserved in the first decades after the Fontan procedure but declines over time. For example, a systolic dysfunction is present in 40% to 60% of patients with Fontan circulation undergoing evaluation for heart transplantation.[7] In HF, assessment of the functional status of patients is important for their diagnosis, management, and prognosis. Unfortunately, the assessment of functional status in patients with congenital heart disease can be challenging. For example, the New York Heart Association (NYHA) functional classification, which is one of the most commonly used evaluation tools in HF, has limited practicality in CHD. In fact, none of the available heart failure classification grading scales has been validated in CHD patients yet.[8] Congenital heart disease is by definition present from birth, and patients adapt their daily activities to their abilities. Patients with CHD make lifelong adaptations to their cardiovascular disease and its slow progression, so they may not be aware of the true extent of their exercise intolerance. This may lead to underestimation and underreporting of the severity of their physical limitations.[8,9] Because they are accustomed to a certain level of physical activity, patients with CAD may be virtually unaware of the limits of their exercise tolerance. In the described case, despite the clinical features of central and peripheral cyanosis, the patient was unaware of the severity of her disease. It was only after the successful percutaneous procedure, when the patient's oxygen saturation increased, that she noticed a subjective improvement in her exercise tolerance.

Treatment of symptomatic patients with a failing single ventricle in a Fontan circulation should always be initiated cautiously, taking into account the labile balance between ventricular preload and systemic afterload.[1] Cardiac catheterization is an important procedure

in Fontan patients as it can be used as a diagnostic tool and for interventional treatment. According to the latest ESC guidelines for the management of adult CHD, in selected adult patients it may be appropriate to consider device closure of a fenestration if there is significant cyanosis, but this may also worsen the patient's condition. In addition, catheter intervention may be required in cases of flow obstruction or anomalous vascular connections.[1] An unobstructed Fontan pathway is essential for optimal hemodynamics, as the Fontan circulation lacks a sub-pulmonary ventricle and it relies on low resistance, "passive" flow through the venous system and pulmonary vascular bed.[10] Unfortunately, Fontan conduit stenosis is a well-known complication in Fontan patients.[11–13] In the study by Patel et al., the authors observed a significant decrease in Fontan conduit cross-sectional area (CSA) over a mean follow-up period of 10 years in the population of 158 patients with extracardiac Fontan.[12] The mechanism of this stenosis was theorized by Hagler et al.: at the time of Fontan operation, patients are very young and a relatively short 18–20 mm conduit must be implanted in order to avoid compression of other structures. Years later that same conduit becomes significantly stretched with diffuse narrowing. Therefore, the authors suggest that all Fontan patients should be reevaluated for conduit obstruction at least 10 years after implantation and periodically thereafter.[11] Significant stenosis of the Fontan conduit requires appropriate management. Stenting of the obstructed extracardiac conduit has become a promising alternative to a surgical treatment.[13] In our patient, the 18 mm Gore-Tex extracardiac conduit was implanted during the Fontan operation at the age of 6 years. After 12 years, the same conduit became significantly stretched (due to patient's growth), which resulted in its narrowing up to 11.2 mm. The patient was qualified for cardiac catheterization, during which two stents were implanted in the stenosed Fontan conduit; additionally, the fenestration and main PA were closed. Because the patient presented with cyanosis, closure of the fenestration was considered desirable—the use of covered stents allowed for complete closure. The main PA was closed because blood from the Fontan circulation was unnecessarily flowing into the hypoplastic RV, posing a risk of thrombosis. This treatment proved successful as the Fontan conduit stenting and fenestration closure eliminated the patient's cyanosis and improved her exercise tolerance.

Lifelong and regular follow-up at an expert center is essential for all patients with CHD. According to the guidelines, adult Fontan patients should be followed in specialized adult congenital heart disease centers, usually at least annually, with echocardiography, ECG, blood tests, and exercise testing.[1] Unfortunately, in the described case the patient had a four-year gap in cardiology care, which resulted in a worsening of her functional status. The occurrence of such gaps in care is a serious problem in CHD; in the study by Gurvitz et al., the authors found that a > 3 year gap in cardiology care was identified in 42% of 922 adult CHD patients.[14]

In conclusion, the described case illustrates that the treatment of CHD is challenging because each patient has a unique morphology and physiology of the congenital defect. With the remarkable development of percutaneous techniques, more and more patients are receiving optimal, personalized treatment for their disease. A wide range of available techniques and devices allows interventional cardiologists to implement non-standard solutions to complex CHD problems. In the described case, a complex percutaneous intervention was performed with a success. A 1.5-year follow-up showed a long-term stabilization of the patient's condition and proved the effectiveness of the procedure. It is also important to emphasize that a lifelong and regular follow-up in an expert center is essential for all patients with CHD. Unfortunately, the gaps in cardiology care in patients from this group are a serious problem that all clinicians and health system managers should be aware of.

AUTHOR CONTRIBUTIONS

Andrzej Wittczak: Data curation; writing – original draft; writing – review and editing. **Paweł Dryżek:** Data curation; resources. **Marek Maciejewski:** Project administration; supervision. **Anna Kula-Mazurek:** Investigation; resources; validation. **Tomasz Moszura:** Data curation; project administration; resources; supervision. **Agata Bikiewicz:** Data curation; investigation; resources; validation; visualization. **Agata Magdalena Bielecka-Dabrowa:** Conceptualization; formal analysis; funding acquisition; project administration; supervision; writing – review and editing.

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CONFLICT OF INTEREST STATEMENT

The authors declare no conflict of interest.

ETHICS STATEMENT

The study is in compliance with the Declaration of Helsinki.

SUPPORTING INFORMATION

Additional supporting information can be found online in the Supporting Information section at the end of this article. The following supporting information can be

downloaded: Video S1—Transthoracic echocardiography, apical view; Video S2—Transthoracic echocardiography, apical view 2; Video S3—Fontan conduit angiography before stenting; Video S4—Angiogram at the end of the procedure; Video S5—Transthoracic echocardiography after the procedure, subcostal view, the Fontan conduit with visible stent.

DATA AVAILABILITY STATEMENT

Individual participant data that was presented in this article after deidentification will be available for researchers who provide a methodologically sound proposal. Proposals may be submitted after 9 months and up to 36 months following article publication.

CONSENT STATEMENT

The authors have confirmed during submission that patient consent has been signed and collected in accordance with the journal's patient consent policy.

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