

# Coronary sinus ostial atresia: A rare associated anomaly that should be remembered in patients undergoing univentricular palliation

 Ayhan Çevik,  Murat Ertürk\*,  Şule Turgut Balcı\*\*,  
 Kanat Yalçın\*\*\*,  Ali Rıza Karacı\*

Departments of Pediatric Cardiology, and \*Cardiovascular Surgery,  
\*\*Anesthesiology, \*\*\*Faculty of Medicine, İstanbul Bilim University;  
İstanbul-Turkey

## Introduction

Coronary sinus ostium atresia associated with congenital heart diseases with single ventricular physiology is rare and detection of this anomaly is very important (1-3). In coronary sinus ostial atresia, the coronary sinus usually drains into an unroofed coronary sinus and the left atrium. However, it may drain into the right atrium via the Thebesian veins or, as presented in our case, drainage may only be present into the left persistent superior vena cava (SVC) (4-7). A case of a patient with tricuspid atresia who was diagnosed to have coronary sinus ostium atresia with retrograde drainage of the coronary veins into the left superior vena cava during diagnostic heart catheterization prior to the Fontan procedure is presented.

## Case Report

An 8-year-old boy was referred to İstanbul Bilim University Şişli Florence Nightingale Hospital with diagnosis of tricuspid atresia, ventricular septal defect, pulmonary stenosis, and hypoplastic pulmonary arteries. He underwent a right-modified Blalock-Taussig shunt at the age of 1 month, and right-sided bidirectional caval pulmonary anastomosis and ligation of the MBT shunt was performed when he was 1 year old. Pulmonary arteries were considered to be mildly hypoplastic on echocardiographic examination. A diagnostic cardiac catheterization was planned in preparation of the Fontan procedure. An innominate vein was seen during transthoracic echocardiography, but persistent superior vena cava was not detected. Dilated distal coronary sinus lumen with ostial atresia and retrograde coronary sinus drainage into the innominate vein via a thin left superior vena cava was detected during cardiac catheterization (Video 1). Pulmonary artery growth was felt to be inadequate for a successful Fontan operation, and medical follow-up with Fontan completion along with coronary sinus unroofing was planned to be performed later.

## Discussion

Coronary sinus (CS) ostium atresia associated with persistent left SVC in the absence of left atrial connection is a very rare cardiac anomaly. CS ostium atresia with unobstructed retrograde drainage via left SVC is an intrinsically benign anomaly. However, preoperative recognition and surgical intervention for this anomaly are necessary if ligation or division of the left SVC is indicated. If the left SVC is the only coronary venous drainage vein, coronary venous hypertension may progress after Glenn or Fontan procedure without division of the left SVC.

In the presence of a left persistent vena cava with a relatively normal diameter innominate vein, coronary sinus atresia via retrograde drainage should be suspected. Surgical ligation of the left persistent superior vena may cause death in undiagnosed patients (3, 4, 6). It has been previously described that surgical procedures (redirection or unroofing) can be performed to prevent complications after Glenn and/or Fontan operations (1, 3). In our patient, pulmonary artery growth was considered to be inadequate for successful Fontan operation, and medical follow-up with later Fontan completion along with coronary sinus unroofing was planned.

The relation between coronary sinus ostial atresia and accessory pathways causing supraventricular dysrhythmias has also been discussed in the literature (5-8). Our patient was on amiodarone therapy for the last 3 years because of recurrent supraventricular tachycardia attacks.

## Conclusion

We believe that coronary sinus atresia is a rare but very important congenital anomaly that should not be forgotten during preoperative assessment of patients with single ventricular physiology undergoing Glenn or Fontan procedures.

## References

1. Tadokoro N, Hoashi T, Kagisaki K, Shimada M, Kurosaki K, Shiraishi I, et al. Clinical Features and Surgical Outcomes of Coronary Sinus Orifice Atresia. *Pediatr Cardiol* 2016; 37: 387-91. [\[CrossRef\]](#)
2. Kaneko Y, Kobayashi J, Yamamoto Y, Tsuchiya K. Light-guided surgery to repair coronary sinus orifice atresia with left superior vena cava. *Eur J Cardiothorac Surg* 2008; 33: 939-41. [\[CrossRef\]](#)
3. Fulton JO, Mas C, Brizard CP, Karl TR. The surgical importance of coronary sinus orifice atresia. *Ann Thorac Surg* 1998; 66: 2112-4.
4. Petit CJ, Webb GD, Rome JJ. Creation of a coronary sinus to atrial communication in coronary sinus ostial atresia improves cardiac function after Fontan. *Catheter Cardiovasc Interv* 2007; 70: 897-9.
5. Okuyama Y, Oka T, Mizuno H, Sakai T, Hirayama A, Kodama K. A case of atrioventricular nodal reentrant tachycardia with atresia of the coronary sinus ostium. *Int Heart J* 2005; 46: 899-902. [\[CrossRef\]](#)

6. Jha NK, Gogna A, Tan TH, Wong KY, Shankar S. Atresia of coronary sinus ostium with retrograde drainage via persistent left superior vena cava. *Ann Thorac Surg* 2003; 76: 2091-2. [CrossRef]
7. Bozbuğa N, Balkanay M, Taş S, Erentug V, Yakut C. Atresia of coronary sinus ostium with coronary venous flow to left atrium. *Asian Cardiovasc Thorac Ann* 2003; 11: 261-2. [CrossRef]
8. Hidestrand PM, Kirkpatrick EC, Mitchell ME. An Unusual Case of Severe Stenosis of the Coronary Sinus Ostium in Association With Double Inlet Left Ventricle. *World J Pediatr Congenit Heart Surg* 2014; 5: 473-4. [CrossRef]

**Video 1.** Cardiac catheterization demonstrating dilated distal coronary sinus lumen with ostial atresia and retrograde coronary sinus drainage into the innominate vein via thin left superior vena cava.

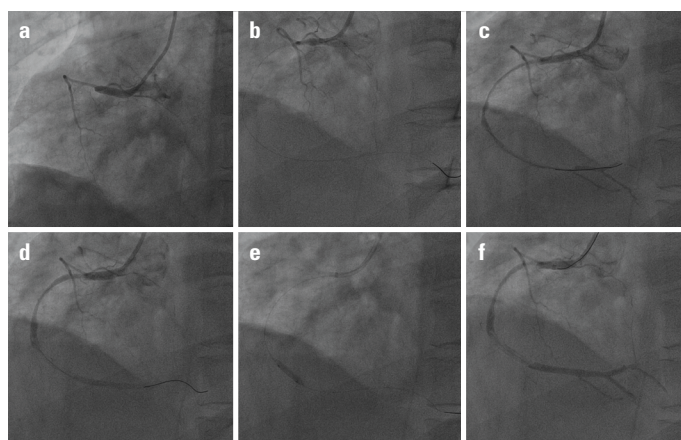
**Address for Correspondence:** Dr. Ayhan Çevik, İstanbul Bilim Üniversitesi Tıp Fakültesi Hastanesi, Çocuk Kardiyolojisi Bölümü, Abide-i Hürriyet Cad. İstanbul-Türkiye

Phone: +90 532 657 10 42

E-mail: ayhancevik12@hotmail.com

©Copyright 2018 by Turkish Society of Cardiology - Available online at [www.anatoljcardiol.com](http://www.anatoljcardiol.com)

DOI:10.14744/AnatolJCardiol.2018.84453



**Figure 1.** Angiography revealed occlusion of the right coronary artery (a). The blood flow did not improve after the crossing of the guidewire (b). There are still many thrombi and stenoses after aspiration (c, d). After balloon dilatation in the middle of the right coronary artery (e), the blood flow partially improved (f)

## Case Report

A 40-year-old male admitted to another hospital with fever for half a month and pain in lower limbs for 1 week. Ultrasound results revealed deep vein thrombosis of the lower extremity, and laboratory examination revealed a high level of troponin I.

The patient was transferred to our hospital. Electrocardiography revealed ST elevation in leads II and III, AVF and V1–3, Q wave was found in leads II and III and AVF. Troponin I level was 4.402 ng/mL. Therefore, a diagnosis of myocardial infarction was confirmed. However, the blood cell count revealed an extremely high white blood cell (WBC) count ( $81.9 \times 10^9/L$ ) and low platelet count ( $50 \times 10^9/L$ ) that indicated leukemia. The patient underwent bone marrow aspiration that confirmed the diagnosis of acute myelogenous leukemia (M4).

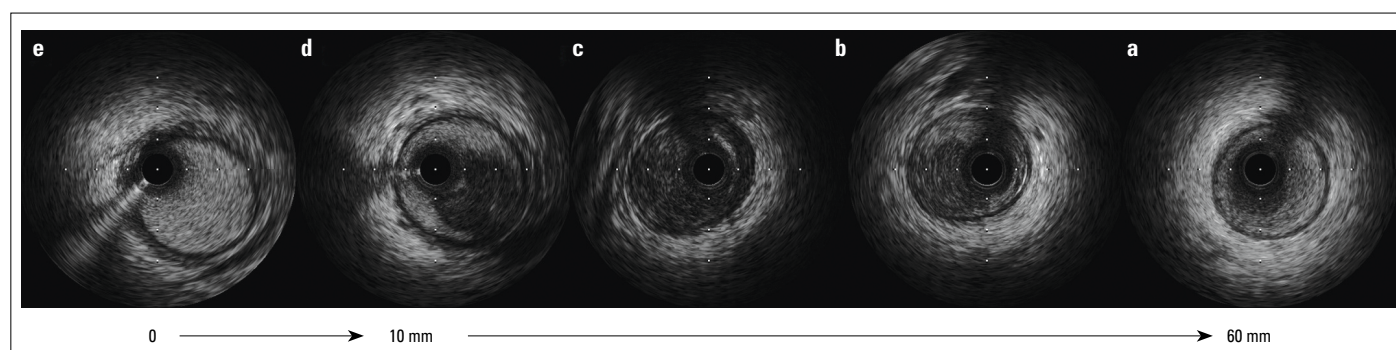
The patient then underwent percutaneous coronary angiography, which revealed an occlusion of the proximal right coronary artery (Fig. 1a). After the guidewire crossing, there was still no blood flow in the right coronary artery (Fig. 1b). Intravascular ultrasound was performed, which showed multiple thrombi in the

## Myocardial infarction caused by a leukemic clot: A case report

Jing Li, Liang Dong, Chang-ling Li, Ya-bin Liu, Li-jun Feng  
Department of Cardiology, The Second Affiliated Hospital, Zhejiang University School of Medicine, Hangzhou-China

### Introduction

Leukemia complicated with myocardial infarction is well known. However, myocardial infarction caused by a leukemic clot has been rarely reported. We present a case of myocardial infarction with right coronary artery occlusion due to the leukemic clot.



**Figure 2.** Intravascular ultrasound in the right coronary artery showed complete endarterium without atherosclerosis (a–e) but multiple thrombi (b, c, d) from distal to proximal