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

Ayhan Çevik, Murat Ertürk*, Şule Turgut Balçı**, Kanat Yalçın***, Ali Rıza Karacı*
Departments of Pediatric Cardiology, and *Cardiovascular Surgery, **Anesthesiology, ***Faculty of Medicine, İstanbul Bilim University; Istanbul-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 Istanbul 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.

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**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 Kardiyoloji Bölümü, Abide-i Hürriyet Cad. İstanbul-Türkiye
Phone: +90 532 657 10 42
E-mail: ayhancevik12@hotmail.com
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