Transcatheter intervention for double-steal syndrome from isolation of the subclavian artery associated with patent ductus arteriosus

Isolation of the subclavian artery is a rare aortic arch anomaly, in which the left subclavian artery (LSCA) does not originate from the aortic arch and is connected to the pulmonary artery, through the arterial duct. A five-year-old girl, with left-arm claudication symptoms and a diagnosis of patent ductus arteriosus (PDA), was referred for interventional PDA closure. Blood pressure measurement showed that her right-arm systolic pressure was 30 mm Hg higher than that of her left arm. An unusual PDA was detected on echocardiography. During catheter angiography, a right-sided aortic arch was observed, and injection into the right vertebral artery (RVA) demonstrated a retrograde flow, down the left vertebral artery (LVA) to the LSCA, PDA, and pulmonary artery (Videos 1 and 2). The patient had a pathology resulting in double-steal syndrome, from the LVA to the left arm and the pulmonary artery. PDA closure was planned to eliminate the pulmonary artery steal. However, passing the PDA through the antegrade route was not possible. The PDA was closed with an Amplatzer duct occluder type II device, via the retrograde route (Videos 3–7). After 1 month, pain in the left arm was decreased. When coarctation is not detected in a patient with PDA, an isolated LSCA should be considered, particularly when the left upper extremity blood pressure is low. Due to subclavian steal syndrome, the PDA closure using the transcatheter intervention and disconnecting the subclavian artery from the pulmonary artery represents a safe therapeutic alternative to surgery in patients without critical extremity ischemia.

Informed consent: Informed consent was obtained from the patient’s parents.

Video 1. Catheter angiography, demonstrating a right-sided aortic arch. The left subclavian artery was opacified later.

Video 2. Catheter angiography, showing an injection into the right vertebral artery, which fills the left vertebral artery via a connection between the arteries, before forming the basilar artery. The left vertebral artery is filling both the left subclavian artery and the patent ductus arteriosus.

Video 3. Catheter angiography, demonstrating the right vertebral artery (RVA), which was reached retrograde from the arterial pathway. A 0.014” soft coronary guidewire was pushed forward from the RVA into the left vertebral artery, via the connection between the arteries.

Video 4. After passing the coronary guidewire in a retrograde manner, the wire was snared via an antegrade route in the pulmonary artery, and the arteriovenous loop was formed.

Video 5. Catheter angiography image, showing an injection into the subclavian artery with a 5F JR4 catheter sent over the created arteriovenous loop.

Video 6. Catheter angiography shows the antegrade injection into the pulmonary artery, which demonstrated the patent ductus arteriosus closure using Amplatzer duct occluder type II. The steal was decreased to the pulmonary artery.

Video 7. Catheter angiography, demonstrating the retrograde injection into the right vertebral artery, which fills the left vertebral artery and the left subclavian artery. The flow to the left subclavian artery increased.

© Bekir Yükcü, © Fatma Sevinç Şengül, © Alper Güzeltaş
Department of Pediatric Cardiology, University of Health Sciences, Istanbul Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training and Research Hospital; Istanbul-Turkey

Address for Correspondence: Dr. Alper Güzeltaş, Sağlık Bilimleri Üniversitesi, Istanbul Mehmet Akif Ersoy Göğüs Kalp ve Damar Cerrahisi Eğitim ve Araştırma Hastanesi, Çocuk Kardiyojloji Kliniği, Halkali/Küçükçekmece, İstanbul-Türkiye
Phone: +90 212 692 20 00
E-mail: alperguzeltas@hotmail.com
©Copyright 2020 by Turkish Society of Cardiology - Available online at www.anatolijcardiol.com
DOI: 10.14744/anatolijcardiol.2020.51481

Intra-atrial path of the right coronary artery: An infrequent and still unknown anomaly

Coronary congenital anomalies are rare. Most are benign, but some can cause myocardial damage or sudden death. With the advancement of imaging studies, its diagnosis is increasing. We present a series of three cases in which the right coronary artery has an intra-auricular path. We examined a middle-aged man and two women with no cardiovascular history. They consulted a doctor for chest pain. Conventional ergometry and transthoracic echocardiography were performed and revealed normal results. Due to persistence of the symptoms, a computed tomography of the coronary arteries is requested. There were no significant atherosclerotic lesions found; however, an abnormality was found. Anatomical characteristics were evaluated using conventional axial images and curved multiplanar reconstructions, with retrospective acquisition and single injection of contrast (Fig. 1 and 2). The abnormality is diagnosed by evaluating that any segment of the artery is completely surrounded by blood in the right atrium. The artery originates from the right Valsalva sinus, with the junction of the proximal and middle thirds being affected. It crosses...
the inferolateral portion inside the right atrium with an average length of 27.7 mm in three cases and a maximum depth of 5 mm. Our cases do not associate with other coronary anomalies. The symptoms (if any) are unknown. In a 2-year follow-up, no major cardiovascular events or cardiovascular death appeared. It is important to be aware that this anomaly is not visualized by coronary angiography. Apart from the lack of knowledge about its prognosis, its clinical importance may lie in the risk of accidental injury during endovascular procedures or in cardiac surgery.

Figure 1. (a, b) Coronal image with cardiac synchronization and contrast only in left cavities which demonstrates the middle/distal third of the right coronary artery inside the right atrium (arrow). Plane images 4C oblique images following the axis of the artery. The intra-atrial path of the artery with contrast completely surrounding it. (c) With contrast in one phase. (d) With contrast in two phases to opacify the right cavities.

Figure 2. Images with multiplanar reconstruction following the axis of the artery, demonstrating the intracavitary path of the artery. (a) With contrast in one phase. (b) With two-phase contrast to opacify the right cavities.

An 84-year-old female with history of bioprosthetic mitral valve replacement four years earlier presented with a progressively worsening dyspnea on exertion. A transthoracic echocardiogram (TTE) showed a mean gradient of 13 mmHg across the bioprosthetic valve (Fig. 1a). The TTE also noted a left ventricular outflow tract obstruction (LVOTO) gradient due to residual native valve tissue (peak gradient >130 mm Hg) (Fig. 1b, arrow) and an estimated right ventricular systolic pressure of 70 mm Hg. The transesophageal echocardiogram (TEE) corroborated that two out of three leaflets on the bioprosthetic valve had a significantly reduced motion (Fig. 1c and Video 1). In addition, a significant systolic anterior motion of the native mitral valve anterior leaflet (red arrows) was observed which had not been resected throughout the original surgery, resulting in a significant dynamic LVOTO (Fig. 1d and 1e and Video 1). She subsequently underwent redo bioprosthetic mitral valve replacement and resection of the native anterior mitral valve leaflet (Fig. 1f). The resected bioprosthetic valve revealed findings consistent with a degenerated valve prosthesis with calcified leaflets and significantly restricted motion (Fig. 1g, white arrows). Her postoperative course was unremarkable, and she was discharged on postoperative day 7.

Postoperative LVOTO may occur for a variety of reasons, including abnormal prosthetic position, hypercontractile ventricle, left ventricular hypertrophy, and a small ventricular cavity (1-3). Dynamic obstruction secondary to the preservation of native anterior mitral valve leaflet has also been outlined (our patient) (4, 5). This problem was likely exacerbated by the presence of a prosthetic stenosis. This case also highlights the importance of intraoperative TEE.

References

1. Bortolotti U, Milano A, Tursi V, Minarini M, Thiene G, Mazzucco A. Fatal obstruction of the left ventricular outflow tract caused by low-