CASE REPORT

Percutaneous Closure of Atrial Septal Defects in Patients with an Aberrant Retroaortic Coronary Artery: Is It Safe?

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ABSTRACT

Coexistence of an ostium secundum type atrial septal defect and a coronary artery anomaly with an aberrant retroaortic course is a rare congenital anomaly that may potentially complicate percutaneous atrial septal defects (ASD) closure. If the anterosuperior rim of the defect is deficient, the abnormally located coronary artery may be compressed or distorted by the implanted device causing myocardial ischemia, arrhythmias, and eventually sudden cardiac death. Due to the potential occurrence of these fatal cardiac events, diagnosis of an aberrant coronary artery with a retroaortic course must be established before percutaneous ASD closure. In this report, two patients with this rare association are described in whom percutaneous closure of the defect was feasible and uneventful. The importance of a careful periprocedural, noninvasive echocardiographic coronary artery imaging is emphasized, and the rational for percutaneous atrial septal defect closure in this unusual anatomic arrangement is proposed.

Key Words. Atrial Septal Defect; Coronary Artery Anomaly; Percutaneous Closure; Device Implantation

Introduction

Congenital coronary artery anomalies are a rare group of congenital heart malformations. They may include stenosis or atresia of the coronary ostium, anomalous origin of one coronary artery from the main pulmonary artery (usually the left), coronary artery fistulae, and anomalous origin of one coronary artery from the opposite facing sinus. In this regard, anomalous origin of the circumflex artery from the right coronary artery or from the right aortic sinus with an aberrant retroaortic course is occasionally seen in routine selective coronary angiography. The approximate incidence of this variant is 0.67% of all coronary anomalies.[1] On the other hand, secundum atrial septal defects (ASD) are one of the most common congenital heart diseases, and percutaneous closure has been proven to be superior to surgical closure with regard to morbidity, hospital length of stay, and rapid hemodynamic improvement.[2,3] The rare association between an ASD and an abnormal coronary artery arising from the opposite aortic sinus with a retroaortic course may have important clinical implications because the anterosuperior (retroaortic) rim of the defect is in close relation to the aberrant artery. Indeed, this association has been suggested as a potential contraindication for device implantation due to the risk of coronary artery compression or deformation caused by the implanted device resulting in myocardial ischemia, arrhythmias, and sudden cardiac death.[4–6]

In this report, we describe two patients with a secundum ASD associated with an aberrant retroaortic circumflex artery arising from the right aortic sinus in whom percutaneous closure of the defect was feasible and uneventful.

Case Reports

Case 1

A 6-year-old boy weighing 33 kg was referred to our clinic for evaluation for possible percutaneous

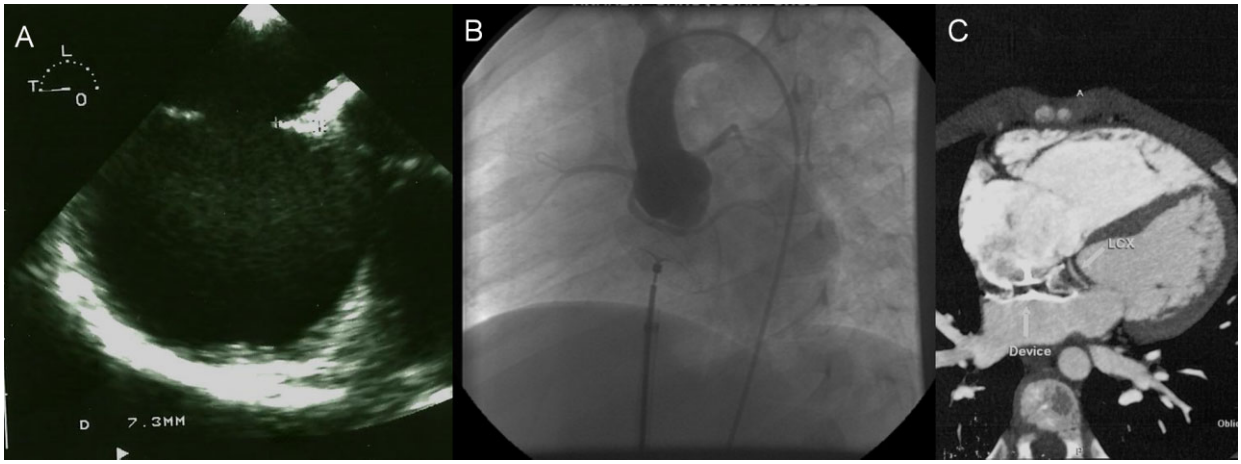

Figure 1. (A) Two-dimensional four-chamber transesophageal image at 0° depicting the ostium secundum type atrial septal defect is visualized. The anterosuperior rim of the defect measuring 7.3 mm is also shown. (B) An 18-mm Occlutech Figulla device is visualized in situ prior to release. A nonselective coronary angiography (LAO 60°) shows a left circumflex coronary artery arising from the right coronary sinus with a retroaortic course and no interference with the coronary artery flow by the implanted device. (C) A multislice cardiac CT image showing the close relationship between de implanted device and the abnormal originated left circumflex coronary artery is shown. (LCX: left circumflex coronary artery).

closure of an ostium secundum ASD. He had history of recurrent upper tract respiratory infections and mildly decreased exercise tolerance. Physical examination revealed the classical findings of an ASD. A 12-lead ECG (Fukuda Denshi Inc., Redmond, WA, USA) showed sinus rhythm, right atrial enlargement, and incomplete right bundle branch block. His chest x-ray demonstrated mild cardiomegaly and increased pulmonary vascular markings. A transthoracic echocardiogram (Philips Inc., Amsterdam, The Netherlands) showed an ostium secundum ASD measuring 15 × 12 mm in diameter, right atrial and right ventricular dilatation, and an abnormal coronary artery running behind the aortic root. Pulmonary pressure was estimated at normal levels. The origin of the abnormally located coronary artery was not clear. Cardiac catheterization under general anesthesia was performed, and an aortic root angiogram (Siemens AG, Munich, Germany) showed an abnormal origin of the circumflex coronary artery arising from the right aortic sinus with an aberrant retroaortic course. A two-dimensional color Doppler transesophageal echocardiogram (Esaote S.p.A., Genoa, Italy) performed before the intervention demonstrated an ostium secundum type ASD measuring 16 × 12 mm in diameter with adequate rims around the defect (anterosuperior rim: 7.3 mm) (Figure 1A). An abnormal origin of the circumflex coronary artery arising independently from the right aortic sinus with a retroaortic course was also delineated. The stretched diameter of the defect was estimated at 18 mm using the "stop-flow technique." An 18-mm Occlutech Figulla device (Occlutech, Helsingborg, Sweden) was implanted uneventfully. Prior to device release, an aortic root angiogram was repeated and showed no interference with the coronary artery flow by the implanted device (Figure 1B). The ECG remained unchanged during the intervention. The device was released, and the patient had an uneventful recovery with hospital discharge the following day. A multislice cardiac CT (Toshiba Aquilion 64, Toshiba Medical Systems, Otawara, Japan) was obtained 13 months after the intervention and showed the device in optimal position in close relationship with the retroaortic circumflex coronary artery (Figure 1C). Vessel compression or deformation was ruled out. After 23 months from the intervention, the clinical course of the patient has been uneventful with no symptoms of coronary ischemia either at rest or during exercise. An exercise test was still not performed due to the age of the patient.

Case 2

A 19-year-old boy weighing 90 kg was transferred to our cardiac catheterization laboratory for percutaneous closure of and ostium secundum ASD. He had been diagnosed 9 months earlier because of a new discovered heart murmur. He complained of increasing shortness of breath, fatigue, and persistent headaches. Physical examination revealed the classical findings of an ASD. The 12-lead ECG

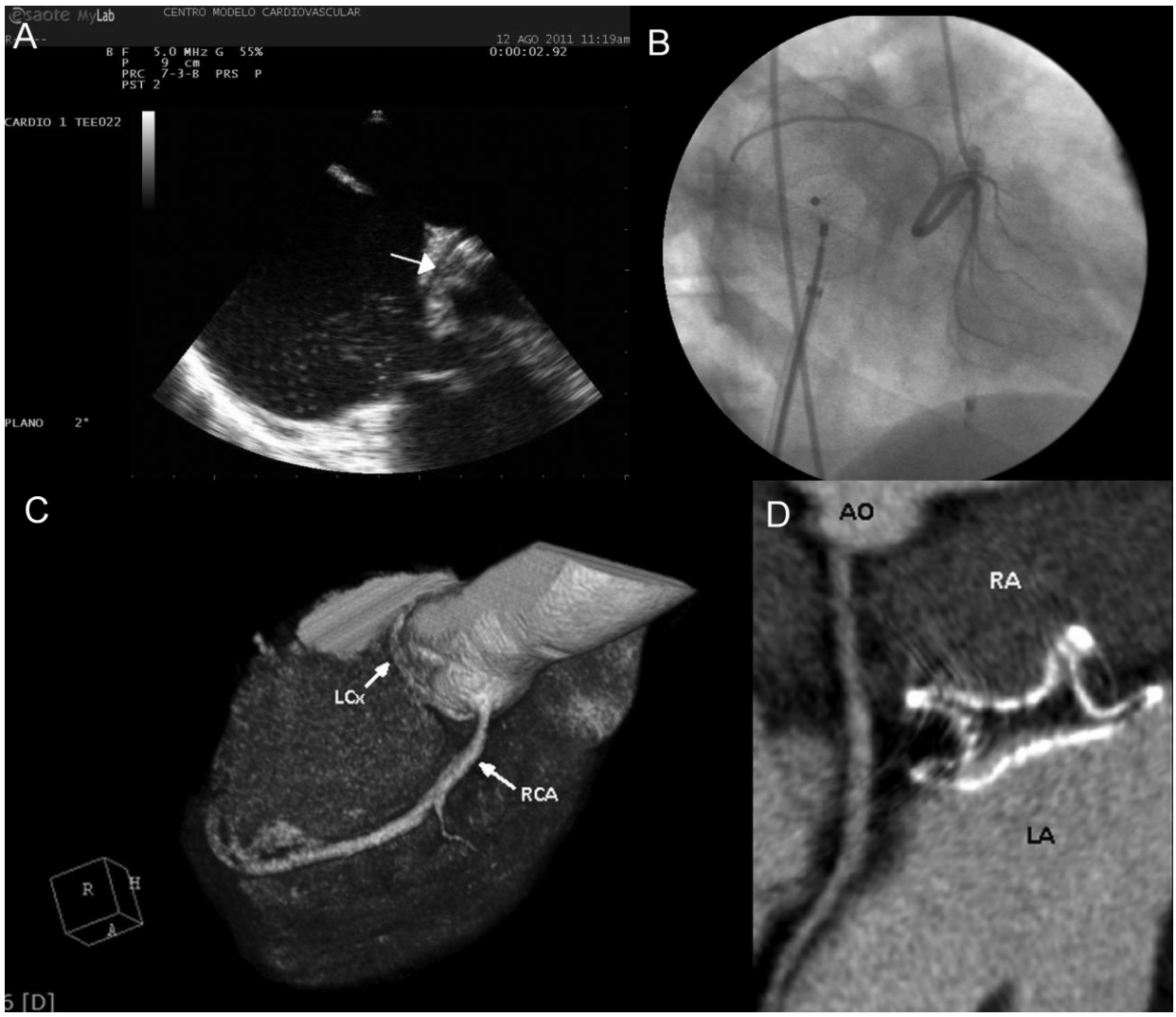

Figure 2. (A) A modified long-axis transesophageal echocardiography view is shown where a linear image running behind the aortic root suggestive of coronary anomaly is present (arrow). An ostium secundum type ASD is also visualized. (B) A 26-mm Amplatzer Septal Occluder is seen in situ still attached to the delivery cable. A selective angiogram shows the left circumflex coronary artery originated independently from the right aortic sinus with a retroaortic course. The distance between the abnormal coronary artery, and the implanted device is visualized as well as the absence of any coronary flow compromise. (C) A multislice cardiac CT with 3D reconstruction of the aortic root shows in a posterior view the separated origin of the circumflex coronary artery from the right aortic sinus with a retroaortic course. The entire right coronary artery is also visualized. (D) A multislice cardiac CT image shows the relationship between the abnormally independently originated circumflex coronary artery from the right aortic sinus and the implanted device. There is neither compression nor distortion of the coronary course. LCx, left circumflex coronary artery; RCA, right coronary artery; AO, aorta; LA, left atrium; RA, right atrium.

showed sinus rhythm, right atrial enlargement, and right ventricular hypertrophy. Chest x-ray displayed a slightly enlarged cardiac silhouette, a prominent main pulmonary artery segment and increased pulmonary vasculature. A transthoracic echocardiogram demonstrated a 19 × 18 mm ostium secundum ASD, right chambers dilatation, and normal pulmonary artery pressures. During the intervention under general anesthesia, a transesophageal echocardiogram depicted a 22 × 19 mm defect with adequate rims (retroaortic rim: 11.6 mm). A previously undetected abnormal linear structure running behind the aortic root suggestive of a coronary anomaly was also shown (Figure 2A). Arterial access was obtained, and a selective coronary angiography demonstrated a separated origin of the circumflex coronary artery from the right coronary sinus with a retroaortic

course. The defect was balloon sized, and the stretched diameter using the "stop-flow technique" was 24.8 mm. A 26 mm Amplatzer Septal Occluder (AGA, St. Jude Medical, Plymouth, MN, USA) was implanted uneventfully. After confirmation of proper positioning and before device release, a repeat selective coronary angiography was obtained and ruled out any circumflex coronary artery distortion or compression (Figure 2B). The ECG remained unchanged during the procedure. The device was released, and the patient had an uneventful recovery. A multislice cardiac CT (Toshiba Aquilion 64) was performed 7 months after the procedure with the aim of defining the relationship between the device and the aberrant retroaortic circumflex coronary artery (Figure 2C). This study showed a correctly positioned device with no compression or distortion of the aberrant circumflex artery (Figure 2D). After 14 months, an exercise stress test was performed and reported as normal at maximal physical effort. The patient has not had any signs or symptom of possible myocardial ischemia since ASD closure.

Discussion

An association between some coronary artery anomalies and adverse cardiac events such as myocardial ischemia, syncope, arrhythmias, and sudden cardiac death has been reported.[7–9] In these rare cases, the anomalous coronary artery (especially the left main) usually has an aberrant course between the aorta and the right ventricular outflow tract. During exercise and stress, with increased aortic blood pressure and flow augmentation through these vessels, compression of the abnormally located coronary artery (especially in systole) may ensue resulting in ischemia. On the other hand, an anomalous origin of the circumflex artery from the proximal right coronary artery or from the right aortic sinus with a retroaortic trajectory usually has a more benign clinical course, being diagnosed as an incidental finding.

Percutaneous ASD closure has evolved from a surgical procedure requiring cardiopulmonary bypass to a percutaneous, catheter-based procedure usually requiring only an overnight hospital stay. Although rare, complications have been described including erosion, device embolization, malfunction, and arrhythmias.[2,3] Long-term clinical outcomes have been excellent: good quality of life, functional class improvement, and ventricular remodeling have been the rule after the procedure.

Accepted anatomical and hemodynamics criteria for performing device closure of ASD have been already established. However, the identification of patients with coronary anomalies that could lead to postinterventional fatalities is a major diagnostic challenge in the absence of clinical manifestations. These anatomic variants may neither be diagnosed during the echocardiographic assessment nor during the interventional procedure itself because coronary angiography is not routinely performed.

In 2003, Casolo et al.[4] described a case in which a young man was diagnosed with a circumflex coronary artery originating from the right sinus of Valsalva only after device closure of a patent foramen ovale. A maximal ergometric effort stress test was normal before the procedure and became abnormal with evidence of decreased coronary reserve months after the procedure. Using three-dimensional coronary magnetic resonance imaging, it was evident that the anomalous coronary artery was compressed between the device and the aortic root. The patient refused surgical repair and improved symptomatically after medical treatment.

In 2008, Scholtz et al.[5] published a case report in which a middle-age woman had a diagnosis of an ostium secundun ASD associated with a coronary anomaly with one single coronary artery originating from the right aortic sinus and a retroaortic course of the main left coronary artery. The ASD was device closed, and due to the close anatomic relationship between the ASD and the left main coronary artery, a coronary angiography was repeated before releasing the device. It demonstrated systolic compression of the left coronary artery by the left atrial disc of the device. Although there was not any acute ECG or hemodynamic changes, the device was removed.

Recently, Bijulal et al.[6] described a similar case in which the circumflex coronary artery originated from the right sinus of Valsalva with a retroaortic course associated to a deficient anterosuperior rim of the ASD. This association was thought to be too risky for percutaneous closure, and the patient was sent for surgical repair.

Based on this and previous reports, it seems that the detection of a retroaortic coronary artery becomes crucial before planning percutaneous ASD closure. Transthoracic and transesophageal echocardiography were accurate to diagnose such abnormality in this report. However, the examiner should keep this uncommon association in mind when selecting a patient for ASD closure. In this

rare scenario, the implanted device may potentially compromise the coronary flow due to compression/distortion of the abnormally located vessel resulting in myocardial ischemia, fatal arrhythmias, and sudden cardiac death. Patients with an ASD and a deficient anterosuperior (retroaortic) rim may be even at a higher risk to develop such complications, especially if a device is oversized with splaying behind the aorta. Probably, in this circumstance, a less rigid type of device may be considered or device closure should not be undertaken at all. On the other hand, if there is enough anterosuperior rim (>7–8 mm, which is the radius of an Amplatzer and Occlutech device) and the edges of the discs (especially the left) remain somewhat distant to the abnormally located coronary artery after implantation, the risk of coronary compression related to the device is unlikely. However, due to the known inevitable minor changes in the position/orientation after release for both devices here implanted (Amplatzer and Occlutech devices), we wonder if an additional postrelease coronary angiography assessment would be appropriate. Although not used in this report, we speculate that three-dimensional transesophageal echocardiogram is a better imaging modality to accurately depict the anterosuperior rim and determine the distance between the edge of the device and the aberrant coronary artery. In addition, it also seems mandatory to assess myocardial ischemia before device release. Moreover, the role of compressive forces of the devices after implantation and the possibility of utilizing less rigid types of devices in this particular setting (i.e., Helex Septal Occluder, Gore Inc, Newark, DE, USA; Atriasept device, Cardia Inc, Eagan, MN, USA; Nit Occlud ASD-R device, pfm Medical, Cologne, Germany) needs to be taken into consideration. In this report, there were no ECG changes after device positioning, and the coronary arteries had no distortion as evaluated by aortic root or selective coronary angiograms before device release. However, both patients reported herein were under general anesthesia and nonphysiologic conditions with decreased oxygen consumption. In addition, no stress test was employed to assess the impact of increased heart rate, blood pressure and oxygen consumption, and possible changes in aortic dimensions in systolic and diastole in relation to device motion on possible coronary compression or distortion. As such, the best way to rule out myocardial ischemia in this rare scenario remains to be defined. We speculate that three-dimensional rotational angiography may be useful to evaluate external coronary compression by the device. Finally, although the presence of chest pain during exercise is probably sensitive enough to raise the diagnosis of myocardial ischemia, the role of additional screening tools such as exercise test, nuclear medicine, and stress echo to detect silent ischemia in the asymptomatic patient during follow-up remains to be determined.

This manuscript describes a rare association that may potentially complicate percutaneous ASD closure. The diagnosis of an aberrant coronary artery with a retroaortic course cannot be overlooked when closing an ASD percutaneously. The importance of a careful periprocedural, noninvasive echocardiographic coronary artery imaging is emphasized. The rational for percutaneous closure of this type of defect in this unusual anatomic arrangement is also proposed.

Conclusions

In selected cases, percutaneous closure of a secundum ASD associated with an aberrant retroaortic coronary artery is feasible and safe. It is mandatory to evaluate in detail the anterosuperior or retroaortic rim of the defect. If there is sufficient rim, device closure of the defect seems to be uneventful, especially if no coronary compression is observed on aortic root or selective coronary angiograms before device release. If this rim is severely deficient, device insertion should probably not be undertaken due to the potential risk of coronary artery compression or deformation resulting in myocardial ischemia. Longer follow-up and larger number of patients are required to assess the long-term outcomes after device closure of an ASD in this particular and unusual anatomy.

Author Contributions

1. Dr. Alejandro Peirone: conception, design, analysis and interpretation of data as well as drafting, revising critically for intellectual content, and final approval.
2. Dr. Alejandro Contreras: design, analysis and interpretation, art-imaging, revising critically intellectual content, and final approval.
3. Dr. Pedro Zangroniz: analysis and interpretation, art-imaging, revising critically intellectual content, and final approval.
4. Dr. Carlos Pedra: conception, design, analysis and interpretation of data as well as drafting, revising critically for intellectual content, and final approval.

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