Inappropriate shocks from a transvenous implantable defibrillator caused by atrial fibrillation and a missed atrial septal defect in a patient with a modified Bentall procedure

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Received 7 March 2021; first decision 19 April 2021; accepted 5 October 2021; online publish-ahead-of-print 7 October 2021

Background
Inappropriate shocks have been reported in ∼1/3 of patients with implantable cardiac defibrillators (ICDs). We report an unusual case of inappropriate ICD shocks due to atrial fibrillation (AF) caused by a missed atrial septal defect (ASD) in a patient with a modified Bentall procedure.

Case summary
A 67-year-old Caucasian male, with an ICD and a history of a modified Bentall procedure 24 years ago, reported to our outpatient clinic with recurrent inappropriate ICD shocks due to episodes of fast AF. The transthoracic echocardiographic exam revealed two large aneurysms at the ostia of the coronary arteries. We performed further evaluation with transoesophageal echocardiogram and computed tomography (CT) angiography. The aneurysms measured on CT were 3.14 cm × 2.29 cm on the right ostium and 1.9 cm × 0.99 cm on the left. A large secundum type ASD of 1.5 cm, missed in all previous echocardiographic studies, was revealed. The therapeutic options of surgical closure of the ASD and repair of the aneurysms or a more conservative approach with percutaneous closure of the ASD and closer follow-up were discussed with the patient. The patient declined the surgical option due to high complication risk, and closure of the ASD with an Amplatzer device was performed 3 months later. A 3-year follow-up was uneventful.

Conclusion
It is of major importance to comprehensively and thoroughly assess patients before and after a surgical intervention to not miss other treatable conditions preoperatively and complications in the postoperative period.

Keywords
Modified Bentall • Aortic aneurysms • Atrial septal defect • Atrial fibrillation • ICD • Case report

ESC Curriculum
9.7 Adult congenital heart disease • 9.1 Aortic disease • 5.3 Atrial fibrillation • 2.1 Imaging modalities • 2.2 Echocardiography
Learning points
- It is of major importance to comprehensively and thoroughly assess patients before a surgical intervention, so as not to miss other treatable conditions preoperatively.
- Awareness of possible postoperative complications after surgery is of crucial importance. Knowing where to look and what to expect is important for recognizing late complications.
- Patients who have undergone aortic surgical procedures should be meticulously followed for late complications at the level of the anastomosis of the graft and valve. All significant structures should be systematically visualized.
- Unusual symptoms and arrhythmias in patients with defibrillators should be evaluated thoroughly.

Introduction
The modified Bentall procedure is the surgical repair of a proximal aortic aneurysm or dissection in combination with aortic valve disease. A composite conduit, consisting of a tube graft and a prosthetic valve, is used to replace the proximal ascending aorta, the root, and the aortic valve. In the original Bentall procedure, coronary arteries are directly reimplanted into the graft, whereas in the modified techniques, arteries are mobilized with a small rim of native aortic tissue, known as the ostial ‘buttons’, that are then attached to the graft. Here, we present an unusual case of inappropriate shocks due to atrial fibrillation (AF) in a patient who had undergone a modified Bentall procedure.

Timeline

| Date                  | Event                                                   | Treatment                                      |
|-----------------------|---------------------------------------------------------|------------------------------------------------|
| 24 years prior to presentation | Hypertension evaluation and diagnosis of a type A aortic aneurysm | Composite graft replacement with a modified Bentall technique |
| 10 years prior to presentation | Episodes of ventricular tachycardia                       | Implantable cardiac defibrillator implantation (ICD) |
| Presentation to outpatient clinic | Inappropriate ICD shocks due to atrial fibrillation and dyspnoea | Transthoracic echocardiography: diagnosis of ostial coronary aneurysms |
| Following 3 months | Dyspnoea on exertion                                      | Documentation of aneurysms and diagnosis of a secundum type atrial septal defect (ASD)—treatment options discussed with patient |
| 3 months after presentation | Ongoing symptoms, worsening dyspnoea                       | Percutaneous ASD closure with an Amplatzer device |
| 3-Year follow-up      | Uneventful                                               |                                                 |

Case presentation
A 67-year-old, Caucasian male with a history of a modified Bentall procedure and an implantable cardiac defibrillator (ICD) presented to our outpatient clinic due to recurrent shocks. Surgery had been performed 24 years prior due to an aortic root type A aneurysm and severe aortic regurgitation with a bicuspid aortic valve. The coronary arteries were directly reimplanted into the graft with the ‘button’ technique. A dual-chamber ICD was implanted 14 years later for secondary prevention due to episodes of unstable ventricular tachycardia (VT) with normal echocardiography and coronary angiography at the time; however, further details and clarification of the substrate of the arrhythmias are not available. He was hypertensive but had no family history of cardiac-related disease or sudden death, and his physical appearance was normal.

On presentation to our clinic, the patient was mildly cyanotic and reported dyspnoea on exertion. Interrogation of the device showed that the shocks were not caused by malignant ventricular arrhythmias but by episodes of AF with fast conduction entering the VF (ventricular fibrillation) treatment zone (Figure 1). The device was programmed in DDD mode with two zones of therapy; VT at 176 b.p.m. and VF >207 b.p.m. The transthoracic bedside echocardiographic exam revealed two large aneurysms at the ostia of the coronary arteries. Both atia were markedly enlarged, and there were signs of mild pulmonary hypertension. The left atrium on the long axis was 60 mm, and the right ventricular systolic pressure was 35 mmHg. Mitral and tricuspid regurgitation were mild. The right ventricle was dilated but had preserved good function, and the left ventricle was normal (Video 1). For further documentation of the diagnosis and a more detailed differential diagnosis between aneurysms and pseudoaneurysms, we performed a transesophageal echocardiogram (TOE) and then computed tomography (CT) angiography of the thoracic aorta and the coronary arteries. Two aneurysms, one on the right ostium and one on the left, were clearly shown in the ‘buttons’, the areas where the coronary arteries were attached to the composite aortic graft. Measurements on the CT scan were 3.14 cm × 2.29 cm for the right aortic aneurysm and 1.9 cm × 0.99 cm for the left aortic aneurysm (Figures 2 and 3). In addition to the ostial aneurysms, all coronary arteries were atheromatous, without significant stenosis. Surprisingly, a large secundum type atrial septal defect (ASD) of 1.5 cm was shown on TOE (Video 2) and CT (Figure 4) but missed on transthoracic echocardiogram due to the acoustic shadow of the prosthetic aortic valve. We have no information regarding preoperative imaging and cannot speculate on the
Due to normal left ventricular function and CT angiography, we did not perform any other tests for myocardial ischaemia. A surgical approach was discussed with a surgeon as an option for both closure of the ASD and repair of the aneurysms but was declined by the patient due to the high risk of complications. The alternative, more conservative approach of transcatheter closure of the ASD and close echocardiographic follow-up of the aneurysms was agreed upon 3 months later due to worsening symptoms. Closure of the ASD was performed with an Amplatzer septal occluder (Figure 5) and was uncomplicated. There has been an uneventful 3-year follow-up.

Discussion

This is an unusual case of a patient with a bicuspid aortic valve and type A aneurysm treated with a modified Bentall procedure complicated by two large aneurysms at the coronary ostia. The patient had an ASD, which was not noticed before the operation, which caused fast AF and subsequent discharge from the ICD. Investigations carried out after the inappropriate shocks brought the ASD and the coronary ostial aneurysms to our attention. The Bentall procedure and its modifications\(^1\) have continued to be considered the gold standard for treating aneurysms involving the aortic root. One of the rare but serious complications of the original Bentall technique is detachment of the coronary ostia with aneurysm and pseudoaneurysm formation at the anastomosis sites, both of which can be potentially lethal if they grow and rupture.\(^2-4\) True aneurysms are abnormal dilations of an artery due to thin vessel walls, but contain all wall layers.\(^5,6\)
contrast, false pseudoaneurysms are caused by damage to all three layers of the arterial wall, resulting in contained haematoma with turbulent flow and a neck that typically does not close spontaneously.\textsuperscript{5,6} They tend to silently increase in size over years and may result in a delayed diagnosis with an increased risk of rupture and mortality.\textsuperscript{2,4} Differential diagnosis between the two entities are often difficult since they have many characteristics in common, and the diagnosis is often made during surgery. The modified procedure with the button...
The technique is reported to have fewer complications at the coronary ostia,\textsuperscript{1,2} and the reports of true aneurysms in the literature and repairs are limited.\textsuperscript{7–10} However, the occurrence of aneurysms has not been completely eliminated, and ostial coronary aneurysms have been reported in patients with connective tissue diseases.\textsuperscript{9,10} Meijboom \textit{et al.}\textsuperscript{9} found coronary ostial aneurysms on magnetic resonance imaging angiography in 17 of 40 patients with Marfan’s syndrome who underwent surgery with the button technique.\textsuperscript{9} Leaving too much native aortic tissue in a too large opening in the graft seems to be the main cause for this type of aneurysm.\textsuperscript{9,10} As reported in Okamoto \textit{et al.},\textsuperscript{10} surgical repair and reconstruction of these rare aneurysms is challenging and requires specialized techniques to avoid mobilizing the coronary ostium from severe adhesions that previous surgery has caused and to reduce the tension on the anastomosis. In our patient, both CT angiography and echocardiographic studies showed that our patient had aneurysms at the coronary ostia that developed in the ‘button’ coronary reimplantation area.

Addressing the unexpected ASD that was discovered, it is known that ASDs are quite common, representing 15% of cardiac malformations at clinics for children and adults. Depending on size, symptoms can start early in life or, more frequently, later in adulthood.\textsuperscript{11} The most common presenting symptoms of ASDs are exertional dyspnoea or fatigue, whereas untreated patients may develop AF due to atrial dilatation, decompensated right heart failure, and finally cyanosis.\textsuperscript{11} Mortality and symptoms are significantly improved after surgical repair of ASDs, and patients with significant ASDs should therefore be offered elective closure once the diagnosis is made.\textsuperscript{11,12} Current indications for treatment include right atrial and ventricular dilatation, ASD minimum diameter >10 mm, and/or Qp:Qs >1.5:1.0.\textsuperscript{12} In our patient, closure was clearly indicated based on symptoms, dyspnoea, dilatation of the right atrium and the right ventricle, size of the

\textbf{Video 2} Transesophageal view of the ASD (atrial septal defect) with colour Doppler.

\textbf{Figure 4} This image shows the atrial septal defect on echocardiography and computed tomography angiography in the systemic arterial phase. (A) Left to right flow through the atrial septal defect in colour. (B) The colour Doppler of the left to right flow through the atrial septal defect. (C) A four-chamber view of the heart, which shows the dilated right chambers compared to the left due to the overload by the atrial septal defect. (D) The atrial septal defect flow on multiplanar reformation in the end-diastolic phase of the cardiac cycle. The slice thickness of the computed tomography reconstruction was 0.75 mm [scale of original image: (A and B) 28%, (C) 47%, and (B) 49%]. ASD, atrial septal defect; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.
ASD, and episodes of AF that caused inappropriate ICD shocks. The coexistence of ASDs with congenital aortic defects is extremely rare. We found only one report by Ermis et al. of an asymptomatic patient with a unicuspid aortic valve and a secundum type ASD, both diagnosed in adulthood. In our case, it is indeed remarkable and unknown why his ASD was missed on presurgical screening and on follow-up for years.

Conclusion

This is an unusual case of a patient with a bicuspid valve who had previously undergone a Bentall procedure with two missed serious problems, an ASD and two large ostial coronary aneurysms, that were discovered after inappropriate ICD discharges caused by fast AF. It is of major importance to comprehensively and thoroughly assess patients before and after a surgical intervention, so as not to miss other treatable conditions preoperatively and complications in the postoperative period.

Supplementary material

Supplementary material is available at European Heart Journal - Case Reports online.

Lead author biography

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Figure 5 The Amplatzer device in (A) the transthoracic study four-chamber view and (B) in the transoesophageal study (scale is 50% of the original). LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.
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