Successful TAVI Despite Sudden Low Output and Ventricular Fibrillation in a Patient with Cardiac Amyloidosis

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Management of patients with cardiac amyloidosis and concomitant high-grade aortic stenosis is challenging. Here, we report the case of a 79-year-old man with transcatheter aortic valve implantation (TAVI) complicated by low cardiac output during release of a Medtronic Evolut R 34 mm valve. After initiation of mechanical circulatory support, the TAVI valve was successfully implanted despite ongoing ventricular fibrillation. The ventricular fibrillation was successfully treated with injection of potassium into the aorta, with subsequent defibrillation and ventricular pacing. Moreover, pharmacological management of transient severe LV dysfunction and high-grade mitral regurgitation finally led to a favourable course.

Keywords: aortic stenosis; cardiac amyloidosis; ventricular fibrillation; transcatheter aortic valve implantation; heart-lung machine

Introduction

Treatment of patients with cardiac amyloidosis and concomitant aortic stenosis is particularly challenging. If transaortic valve implantation (TAVI) is performed in these patients, complications may arise from the structural and functional alterations in the myocardium. Interdisciplinary teamwork in professional teams is key to the successful management of these patients, as demonstrated by the case reported herein (Figure 1).

Case Presentation

History of Presentation

A 79-year-old white man presented to our hospital with shortness of breath upon exertion (New York Heart Association class III). These symptoms had
lasted for 2 years, thus severely impairing his quality of life.

**Past Medical History**

In 2009, the patient had an inferior myocardial infarction and received percutaneous coronary intervention (PCI) with a drug-eluting stent in the right coronary artery. Moreover, he experienced permanent atrial fibrillation. His current medications comprised clopidogrel, apixaban, metoprolol, amlodipine, ramipril, simvastatin and torasemid.

After a stable course, he reported progressive shortness of breath upon exertion, as described above. Thus, in 2020, echocardiography was performed, and showed high-flow, high-gradient aortic stenosis (Vmax 4.4 m/s; Pmax/mean 76/43 mmHg; aortic orifice area 0.8 cm²). Moreover, left ventricular hypertrophy with a maximum wall thickness of 21 mm in the left ventricular septum was observed. His left ventricular function was slightly impaired (45%). No other relevant valvular disease was present, and his systolic pulmonary artery pressure was 29 mmHg + central venous pressure. Subsequent skeletal scintigraphy revealed typical findings consistent with cardiac transthyretin amyloidosis (Figure 2). In October 2021, repeated coronary angiography revealed relevant stenosis in the left anterior descending coronary artery, which was treated with PCI and implantation of a drug-eluting stent. After PCI, the patient’s symptoms persisted.

Therefore, he was carefully evaluated by our heart team (EuroSCORE I: 20.96; EuroSCORE II: 6.25) and was deemed a candidate for TAVI.

**Pre-Procedural Planning**

The electrocardiogram showed atrial fibrillation, a heart rate of 83 bpm and complete left bundle branch block with a QRS duration of 124 ms. Important blood values were creatinine 1.3 mg/dL (reference: 0.6–1.4); potassium 4 mmol/L (reference: 3.5–5); NT-proBNP 7988 pg/mL (reference: < 1800); and TSH 1.38 mU/L (reference: 0.27–4.2). Computed tomography angiography revealed no relevant contraindications for transfemoral access. Because of the bicuspid anatomy, substantial calcification and an annulus diameter of 28.6 mm, a decision was made to implant a Medtronic Evolut R 34 mm valve. For predilatation, a 26 mm balloon (VASC III) was chosen. According to our institutional protocol, the procedure was planned under analgosedation with local anaesthesia and intravenous remifentanil, after evaluation of the patient’s individual characteristics.

**Procedural Workflow**

Establishment of temporary pacing, femoral access and placement of a safari wire in the left ventricle (LV) proceeded without any complications. After predilatation of the aortic stenosis with a 26 mm
balloon (Video 1A), the patient experienced transient hypotension, which resolved spontaneously. After insertion of the Evolut R 34 mm valve into the native annulus, the patient developed prolonged hypotension and subsequent low cardiac output unresponsive to intravenous catecholamines (210 μg noradrenaline and 200 μg suprarenine) via the central venous catheter. Thus, immediate cardiopulmonary resuscitation was initiated, the patient was intubated, and mechanical circulatory support (ECLS) was established via the left femoral artery and vein. The electrocardiogram showed ventricular fibrillation, which did not resolve after several defibrillation attempts. Consequently, a decision was made to implant the TAVI under ECLS and ongoing ventricular fibrillation (Video 1B). After successful implantation, the valve was postdilated with a 28 mm balloon because of residual aortic insufficiency. Acceptable results (Video 1C) were achieved but were difficult to interpret, because no antegrade flow but only retrograde flow from the ECLS was observed via the femoral vessels. However, ventricular fibrillation remained unresponsive to defibrillation despite previous administration of amiodarone. Therefore, a decision was made to administer potassium (3 g) into the aorta via the pigtail catheter. Repeated defibrillation led to asystole, and ventricular pacing via the temporary pacemaker was commenced. At that point, echocardiography indicated highly impaired left ventricular function as well as high-grade mitral insufficiency (Videos 2A and 3A). These findings improved after pharmacological treatment with intravenous milrinon (3 mg) (Videos 3B and 3C). Subsequently, stable hemodynamics was observed, and the patient

Figure 2  Skeletal Scintigraphy Showing Typical Findings Compatible with Cardiac Transthyretin (ATTR) Amyloidosis.

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Figure 3  Procedural Workflow.
CPR, cardiopulmonary resuscitation; LV, left ventricle; ECLS, extracorporeal life support; MVR, mitral valve regurgitation; TAVI, transcatheter aortic valve implantation; TOE, transoesophageal echocardiogram; VF, ventricular fibrillation.
was successfully weaned from ECLS. He was finally transferred to the intensive care unit in stable condition (Figure 3).

Post-Interventional Course

The patient was stabilized in the intensive care unit (length of stay 12 days) and required a pleural puncture as well as intravenous diuretics because of fluid retention. His post-operative hyperactive delirium was ameliorated starting from day 3. Post-TAVI echocardiography showed an ejection fraction of > 55% and an overall acceptable result, with a Pmean of 16 mmHg and mild aortic regurgitation. No mitral regurgitation or pericardial effusion was observed. After being transferred to the general ward, the patient was successfully discharged home 26 days later. His discharge medications comprised a combination of 47.5 mg metoprolol twice per day; 2.5 mg amlodipine once per day; an angiotensin-converting enzyme inhibitor (5 mg ramipril once per day); an oral anticoagulant (5 mg apixaban twice per day); and cholesterol-lowering treatment (20 mg simvastatin once per day). He has been followed up for 14 months and is able to cope with daily life (functional New York Heart Association class II).

Discussion

The appropriate management of patients with cardiac amyloidosis and concomitant high-grade aortic stenosis has been controversial. Recent publications have suggested that TAVI is a viable option after careful evaluation of each patient [1–4]. However, in these patients, in comparison to patients without cardiac amyloidosis, several particular aspects must be considered. Left ventricular wall thickening and myocardial stiffness due to cardiac amyloid deposition, and subsequent loss of normal cardiac structure and function, may lead to LV dysfunction [5] and impaired responsiveness to vasoactive drugs. In our opinion, responsiveness to vasoactive drugs was particularly relevant to the complications in our case. The hypotension and low cardiac output after pre-dilatation of the aortic valve were probably due to the aortic regurgitation, in addition to severe diastolic dysfunction in the presence of severe LV hypertrophy and cardiac amyloidosis. Although the TAVI was successfully implanted under ECLS, neither stabilization of the heart rhythm nor an improvement in LV function was observed. In contrast, severely impaired LV systolic function and high-grade mitral regurgitation were seen. In our patient, careful pharmacological management was essential, and led to improved LV function and the disappearance of mitral regurgitation. Moreover, the electrical stability of the heart is severely impaired in cardiac amyloidosis [5]. This impairment might have accounted for the prolonged ventricular fibrillation in our patient, which was treatable only with injection of potassium into the aorta (i.e., cardioplegia), with subsequent asystole after repeated defibrillation followed by ventricular pacing via the temporary pacemaker. Importantly, successful management of such cases requires highly professional multi-disciplinary teams. Although TAVI has become an established treatment option in patients with aortic stenosis, which enables short procedure times and very high rates of success, a broad spectrum of patients undergo TAVI. The present challenging case illustrates the advantages of performing TAVI in a hybrid operating room with standby mechanical circulatory support. Careful pre-procedural planning and identification of potentially challenging cases are essential to provide the optimal environment for complication management.

Conclusion

This case report demonstrates that complications during TAVI in a patient with high-grade aortic stenosis and concomitant cardiac amyloidosis can be successfully managed with an interdisciplinary team. In our case, the three key components were a) implantation of the TAVI during ECLS and ventricular fibrillation, b) injection of potassium into the aorta with subsequent ventricular pacing after successful defibrillation and c) pharmacological management of transient severe LV dysfunction and high-grade mitral regurgitation.
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Patient Consent
The patient consented to the use of his medical information and data.

Conflict of Interest
The authors declare no conflicts of interest.

REFERENCES

Supplementary Material
Supplementary material is available at the following locations.
- Video 1A https://youtube.com/shorts/As2n6SNbiME
- Video 1B https://youtube.com/shorts/3ubMLmis6jE
- Video 1C https://youtube.com/shorts/RHB2XWfmtBw
- Video 2A https://youtu.be/Dg7X1wfVwU4
- Video 3B https://youtu.be/lcp0Ma1-4jo
- Video 3C https://youtu.be/bhm5mKjn4oI