Atrioventricular block as the initial presentation of calcified bicuspid aortic valve

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Case Report

Abstract

BACKGROUND: Bicuspid aortic valve (BAV) is one of the most common and important congenital heart disorders in adults. If a patient with congenital disorders is not diagnosed early, the patient's disease may progress to a severe condition and thus diagnosis of the main disorder will be rendered difficult.

CASE REPORT: A 34 year-old male patient referred to a referral medical care unit for cardiac electrophysiological study with cardiac shock due to complete heart block 3 months ago and he underwent Dual-Chamber permanent pacemaker (PPM) implantation. Thick and calcified bicuspid AV with invasion to interventricular septum, moderate to severe valve insufficiency (AI), severe aortic valve stenosis (AS), and dilated ascending aorta were observed at his echocardiography. Aortic valve replacement (AVR), aneurysm of ascending aorta, root replacement with tube graft (Bentall Procedure), and also a 3 chambers intracardiac defibrillator (ICD) were used. After 2 weeks of operation, he was discharged and at the first post-hospitalization visit (1 week later), his cardiovascular condition was acceptable.

CONCLUSION: Thick calcified aortic root is a less studied and potential contributing risk factor for AV block after AVR. Therefore, in candidates of aortic valve replacement, considering conductive disorders, especially in patients with calcified valve, is mandatory. Irreversible AV block requiring PPM implantation is a rare condition following AVR.

Keywords: Atrioventricular Block, Bicuspid Aortic Valve, Calcified Valve

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Introduction

Bicuspid aortic valve (BAV) is one of the most common and important congenital heart disorders in adults.[1] BAV can occur along with other co-morbid heart abnormalities which are secondary to BAV and make its diagnosis easier due to further cardiac assessment.[2] Aortic valve stenosis (AS), aortic valve insufficiency (AI), aortic valve calcification, and aneurysm of aorta (AA) can complicate BAV and can induce patients symptomatic especially with aging. Moreover, they can raise the mortality rate of BAV and also BAV repairing process.[1,3,4] BAV as a congenital disorder is diagnosed at childhood, but it is rarely reported at old age. Moreover, with ageing aortic root or ascending aorta disorders may complicate BAV.[5] Therefore, when a patient with congenital disorders is not diagnosed early, the patient's disease may progress to a severe condition, making it difficult to diagnose the main disorder. We would like to report an adult case of calcified BAV presented initially with atrioventricular (AV) block.

Case Report

This case was a 34 year-old male patient referred to our medical care center (Baqiyatallah Hospital, a referral medical care unit for cardiac electrophysiological study) with cardiac shock due to complete heart block [Echocardiography (ECG) findings: pace rhythm (Figure 1)]. He had a history of AV block (ventricular rate: 30, atrial rate: 60) 3 months prior to referral and he had undergone dual-Chamber permanent pacemaker (PPM) implantation. At echocardiography, which is

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shown in figure 2, he had severe left ventricular enlargement, less than 30% ejection fraction, normal right ventricular size with mild dysfunction, mild to moderate mitral valve regurgitation (2+), thick and calcified bicuspid AV with invasion to interventricular septum (with area of 1.4*1 cm), moderate to severe AI, severe AS, and dilated ascending aorta (aneurysm = 5 mm).

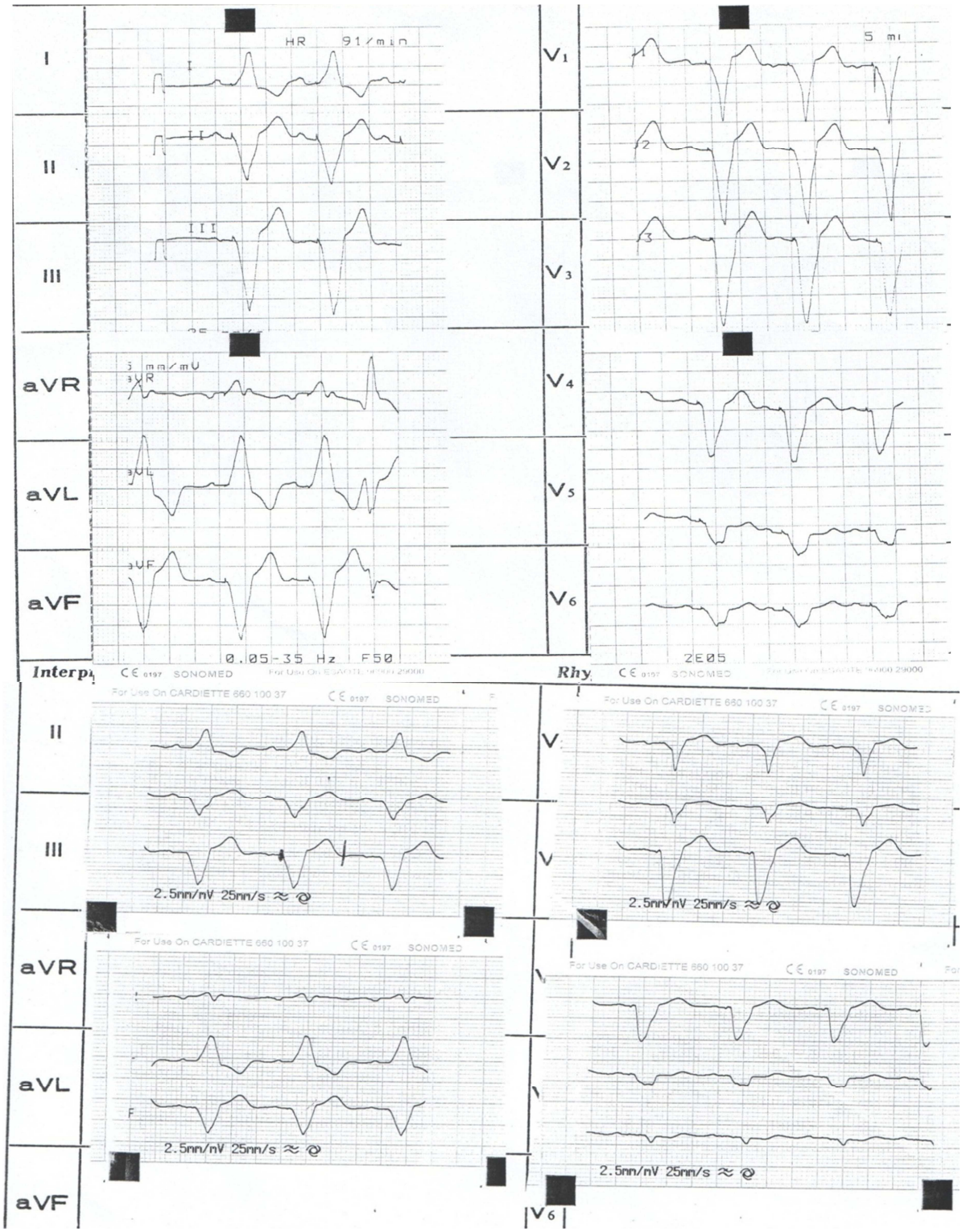

Figure 1. Electrocardiography study at admission (pace rhythm)

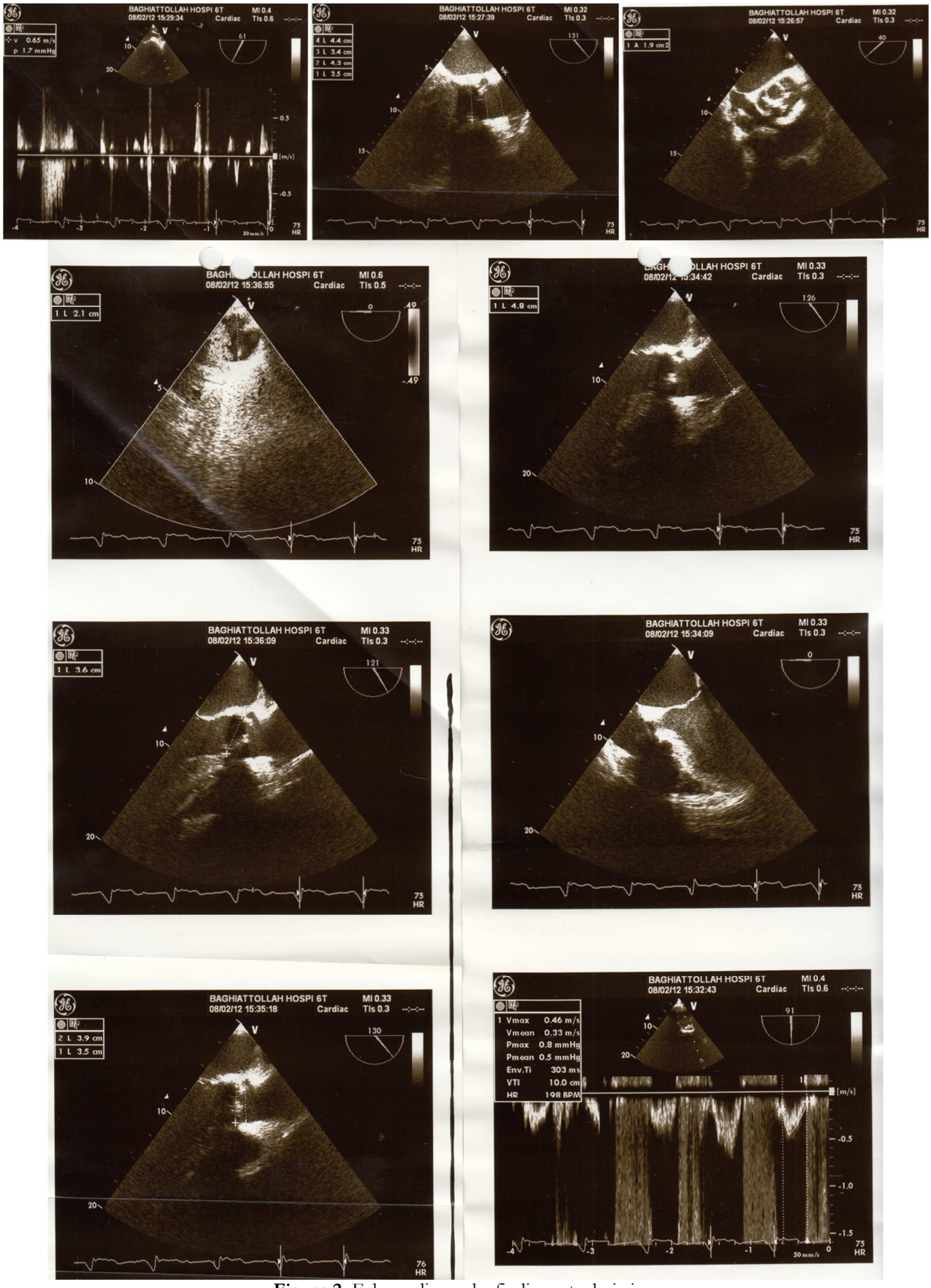

Figure 2. Echocardiography findings at admission

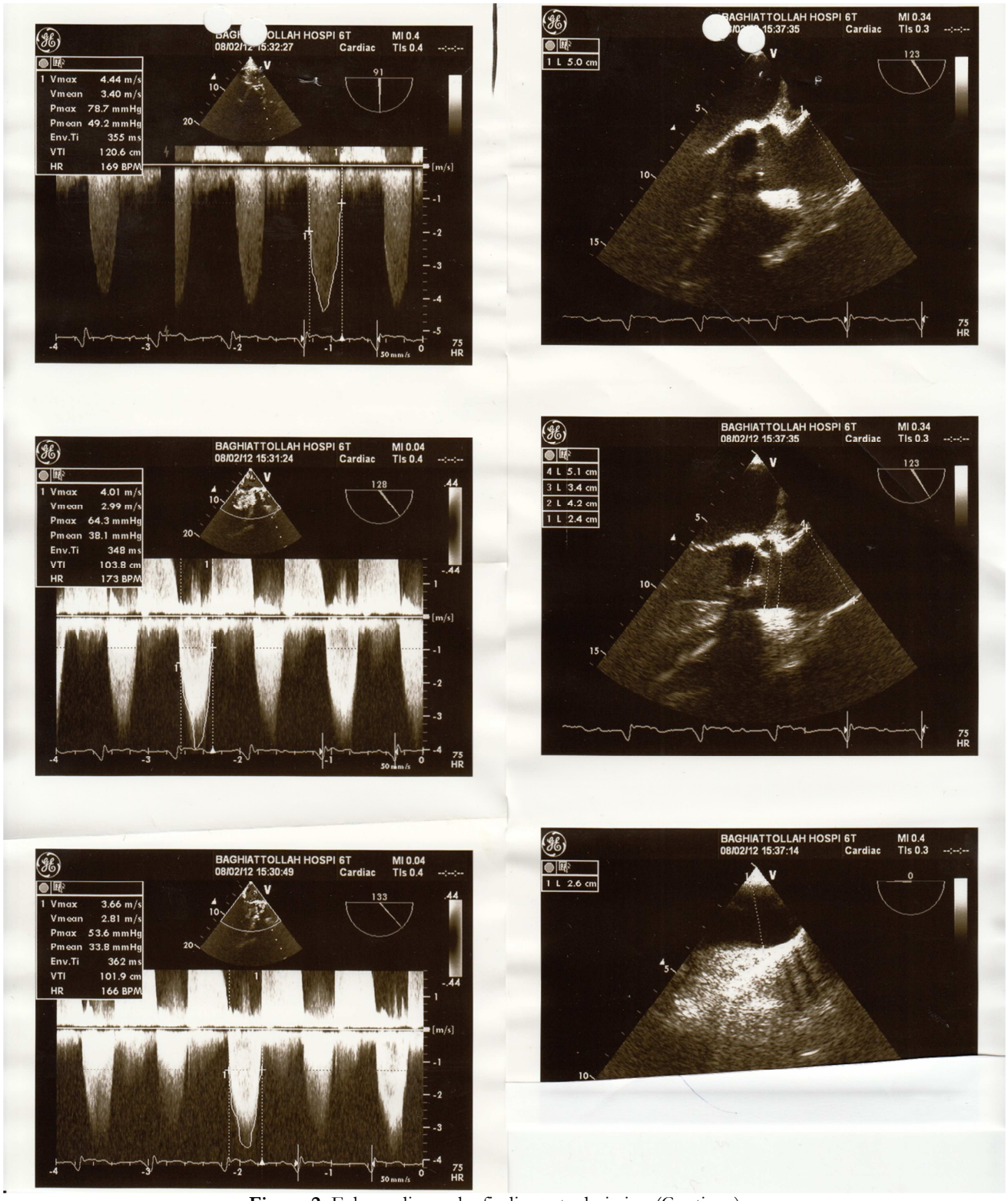

Figure 2. Echocardiography findings at admission (Continue)

The aortic valve properties in echocardiography included aortic valve velocity-time integral (AV VTI) = 120 cm, left ventricular outflow tract velocity-time integral (LVOT VTI) = 11 cm, aortic valve pressure gradient (AVPG) = 78 mmHg, aortic valve mean gradient (AVMG) = 50 mmHg, Anulus = 2.6 cm, valsalva sinus = 4.2 cm, and ascending aorta = 5.0 cm. After admitting the

patient to intensive care unit and stabilizing his general medical condition, the patient was referred to a cardiac surgeon for aortic valve replacement (AVR), replacement of ascending aorta, and root aneurysm with tube graft (Bentall procedure) which is a standard procedure in such cases.[6] Furthermore, a 3 chambers intra-cardiac defibrillator (ICD) was used for him. After 2 weeks of operation, he was discharged and at the first post-hospitalization visit (one week later), his cardiovascular condition was acceptable; there was no considerable dysfunction in implanted valve and pacemaker.

Discussion

There are many reports about the rare presentation of congenital aortic valve abnormalities. In 1978, Edwards et al. report 11 (9%) cases of BAV among 119 cases of fatal dissecting aneurysm of the aorta.[7] They concluded that: "Compared to an estimated incidence of bicuspid aortic valve of about 1 to 2% in the population, the high incidence among subjects with dissecting aneurysm suggests a causative relationship between BAV and aortic dissecting aneurysm."[7] Moreover, they stated that the most common background among patients with fatal dissecting aneurysm of the aorta and BAV is hypertension (73%), but our case neither hypertension nor dissection of aorta.[7] In one of the oldest reports (1953), Gore found a 24% incidence of BAV among 38 cases of dissecting aneurysm of the aorta.[8] Regarding previous reports, it seems that BAV is an associated congenital abnormality among patients with dissecting or aneurismal lesion of the aorta. Suzuki et al. reported a 52-year-old man with aortic valve stenosis and calcification complicated with complete AV block.[9] They concluded that the cause of complete AV block was a considerable progressive calcification involving the conduction system. In addition, they recommended that "the generator implantation be performed in the several days after the operation because of destruction of the pacemaker function by counter shock for arrhythmia in the early post-operative period".[9] The function of the pacemaker in our cases was followed after surgery and it was rearranged before discharge.

Some congenital disorders such as Marfan's syndrome can be associated with these two cardiac disorders (BAV and aneurismal lesion of the aorta), but it is not permanently constant such our reported case.[7] On the other hand, the higher incidence of AV block as a conductive abnormality is associated with aortic valvular stenosis and regurgitation.[10] In these cases with BAV, infectious endocarditis is more frequent than a normal valve, but in our case there is no evidence for endocarditis.[11] Thick calcified aortic root is a less studied and is a potential contributing risk factor for AV block after AVR.[10] Therefore, in candidates of aortic valve replacement considering conductive disorders, especially in patients with calcified valve, is mandatory. Irreversible AV block requiring PPM implantation is a rare condition following AVR, but in some cases, such as our reported case, AV block can be an initial presentation of disorders of the aortic valve.[12] Moreover, due to aortic root abnormalities, such as dissection and aneurysm, which was frequently reported in these cases, more accurate diagnostic methods such as computed tomography (CT) angiography is suggested before further interventions.[13]

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Conflict of Interests

Authors have no conflict of interests.

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