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Lev's Disease Presenting with Complete Atrioventricular Block in a Patient with Severe Aortic Stenosis: A Case Report

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Abstract

Lev's disease is an age-related degenerative conduction disorder characterized by progressive fibrosis and calcification of the His-Purkinje system and adjacent structures. It typically manifests in elderly patients as advanced atrioventricular (AV) block and is often associated with calcific valvular disease. This case report describes a 74-year-old man with a history of stable, non-obstructive coronary atherosclerosis who presented with dizziness and was found to have complete AV block with a ventricular escape rhythm of 36 beats per minute. Transthoracic echocardiography demonstrated severe aortic stenosis with an aortic valve area of 0.96 cm². Contrast-enhanced computed tomography revealed extensive aortic valve calcification extending into the interventricular septum, raising strong clinical-radiological suspicion of Lev's disease due to the anatomical proximity of the His bundle. Surgical risk stratification using Society of Thoracic Surgeons and European System for cardiac operative risk evaluation II indicated an elevated operative risk. Given the patient's advanced age, severe symptomatic aortic stenosis, and persistent conduction abnormality, the multidisciplinary Heart Team recommended transcatheter aortic valve implantation (TAVI) rather than surgical replacement. The patient underwent successful TAVI followed by dual-chamber pacemaker implantation for persistent AV block and remained pacemaker-dependent with a stable rhythm and resolution of symptoms during follow-up. This case underscores that degenerative conduction disease may remain clinically silent until irreversible AV block occurs. Unlike most reports describing new-onset conduction disturbances after TAVI, our patient already presented with complete AV block and imaging demonstrated extension of septal calcification involving the conduction system. Although histopathological confirmation and prior serial electrocardiogram were unavailable, this case highlights the potential role of advanced imaging in identifying patients at increased risk for permanent pacemaker dependence, thereby enabling more accurate risk stratification and guiding closer follow-up.

Keywords: Advanced imaging systems, aortic stenosis, atrioventricular block, cardiac computed tomography, lev's disease, transcatheter aortic valve implantation

INTRODUCTION

Severe calcific aortic stenosis (AS) is the most common valvular heart disease in the elderly population; its prevalence increases markedly with age, and it is associated with high morbidity and mortality if untreated.^[1] Classical symptoms include dyspnea,

angina, and syncope; however, conduction abnormalities are also frequently observed due to the extension of calcification into the interventricular septum and His-Purkinje system. Advanced atrioventricular (AV) block is one of the most serious complications and often requires permanent pacemaker implantation.^[2,3]

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Degenerative conduction system disease, historically described as Lenegre’s and Lev’s diseases, is characterized by progressive idiopathic fibrosis and calcification of the His bundle and bundle branches and typically presents with AV block in elderly patients. In the context of severe AS, the coexistence of valvular calcification and degenerative conduction system disease may accelerate the progression to complete heart block, creating both diagnostic and therapeutic challenges.^[4-6]

We present an elderly patient with severe calcific AS who developed advanced AV block attributed to Lev’s disease. This case highlights the clinical interplay between degenerative valvular and conduction system pathology and underscores the importance of early recognition and management in high-risk older adults.

CASE REPORT

A 74-year-old male with a history of stable, non-obstructive coronary atherosclerosis (no prior myocardial infarction, percutaneous coronary intervention, or coronary artery bypass surgery) presented to the emergency department with dizziness. A 12-lead electrocardiogram (ECG) revealed complete AV block with a ventricular escape rhythm at 36 beats per minute (Figure 1). Transthoracic echocardiography demonstrated a preserved left ventricular ejection fraction (60%), mild mitral and tricuspid regurgitation, and severe AS with an aortic valve area of 0.96 cm² (Figure 2). The coexistence of high-grade AV block and severe calcific AS raised clinical suspicion for Lev’s disease.

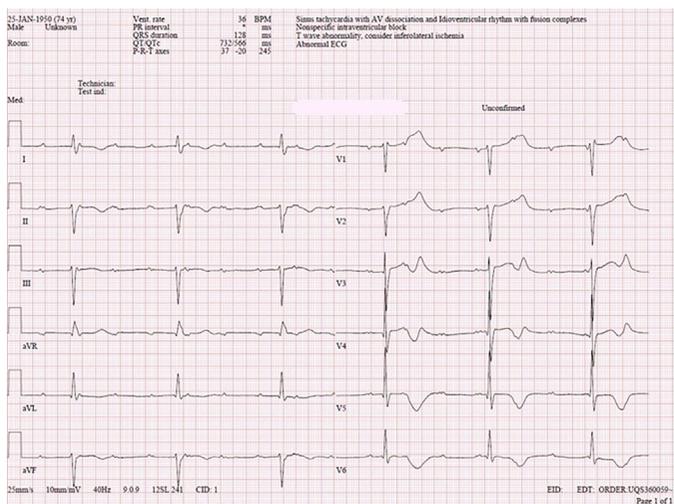


Figure 1. Admission electrocardiogram showing complete atrioventricular (AV) block with a ventricular escape rhythm at 36 beats per minute. There is AV dissociation, with P-waves occurring independently of the QRS complexes. The QRS duration is prolonged (128 ms), consistent with non-specific intraventricular conduction delay. T-wave abnormalities in the inferolateral leads suggest possible ischemic changes

Contrast-enhanced thoracic computed tomography (CT) revealed extensive calcification of the aortic valve extending toward adjacent cardiac structures, including the interventricular septum (Figure 3). Given the anatomical proximity of the His bundle to the membranous septum, conduction system involvement was strongly suspected. Since histopathological confirmation was not feasible, the diagnosis of Lev’s disease was considered on a clinical-radiological basis.

Surgical risk stratification indicated an STS-predicted operative mortality of approximately 4%, a combined morbidity and mortality risk of approximately 13%, and a European System for Cardiac Operative Risk Evaluation II of 5.28%. Considering the patient’s advanced age, severe symptomatic AS, and persistent high-grade conduction abnormality, the multidisciplinary Heart Team determined that surgical valve replacement carried excessive risk and recommended transcatheter aortic valve implantation (TAVI) as the preferred strategy.

With temporary transvenous pacing support, a CoreValve/Evolut prosthesis was successfully implanted. Final angiography showed no paravalvular leak, and the procedure was completed without complications. Despite hemodynamic improvement following valve replacement, the complete AV block persisted. A permanent dual-chamber pacemaker was subsequently implanted.

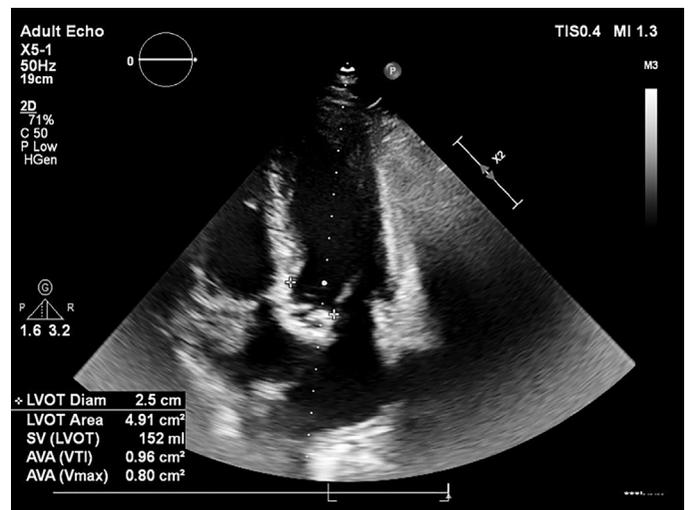


Figure 2. Transthoracic echocardiographic image demonstrating severe calcified aortic stenosis. The LVOT diameter was measured at 2.5 cm, resulting in a calculated area of 4.91 cm² and a stroke volume of 152 mL. Although this value appears relatively high for severe AS, it may reflect preserved systolic function and should be interpreted in clinical context. The aortic valve area was calculated as 0.96 cm² (VTI method) and 0.80 cm² (Vmax method), confirming severe stenosis

LVOT: Left ventricular outflow tract, VTI: Velocity time integral, AS: Aortic stenosis

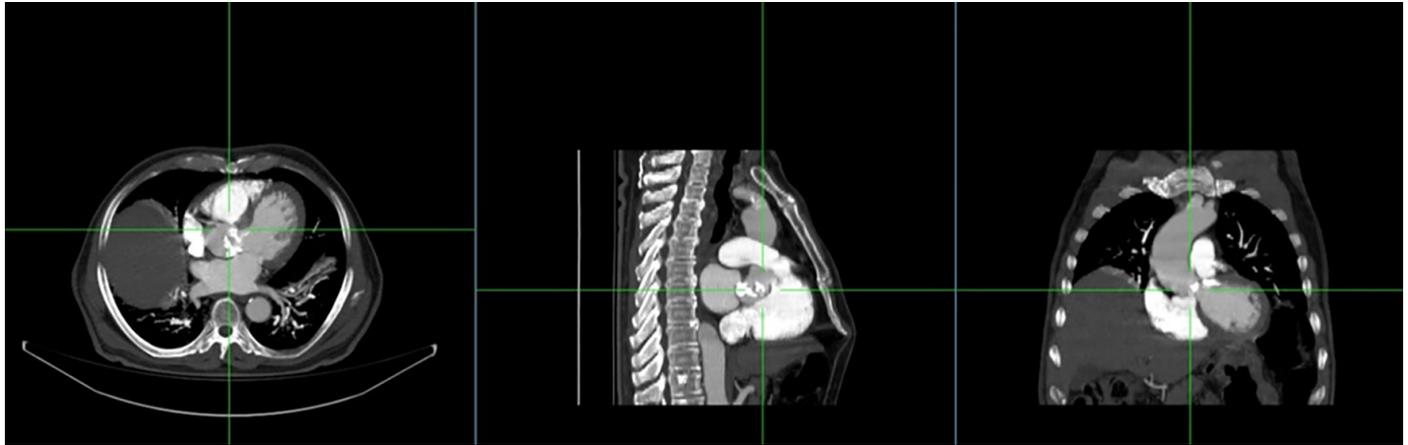


Figure 3. Contrast-enhanced thoracic computed tomography demonstrating severe calcification of the aortic valve with extension toward the interventricular septum. Multiplanar reconstructions (axial, sagittal, and coronal views) show calcific infiltration from the aortic root into adjacent cardiac structures, raising the possibility of conduction system involvement due to proximity to the His bundle. These findings support the suspicion of a degenerative conduction disorder coexisting with severe aortic stenosis

The patient's recovery was uneventful, with complete resolution of symptoms and maintenance of stable cardiac rhythm. At 1- and 3-month follow-up visits, the patient remained pacemaker-dependent, clinically stable, and free of recurrent symptoms.

Written informed consent was obtained from the patient for the publication of this case report and the accompanying images.

DISCUSSION

This case highlights the intricate relationship between severe AS and advanced AV conduction disturbances, raising a clinical-radiological suspicion of Lev's disease. Lev's disease is characterized by acquired fibrocalcific degeneration of the His-Purkinje system in elderly patients, and should be distinguished from Lenegre's disease, which typically affects younger individuals and is a primary fibrotic process without calcification.^[5,6] Unlike ischemic conduction disease, in which scarring is usually confined to areas of prior infarction, Lev's disease involves diffuse calcific infiltration extending into the interventricular septum, as demonstrated by the CT findings in our patient. Although histopathological confirmation would provide definitive proof, it was not pursued in our case due to the limited feasibility of obtaining conduction system tissue, the increased risk of periprocedural complications, and the lack of additional therapeutic benefit, as avoiding potential harm was prioritized. In clinical practice, the diagnosis relies on the integration of clinical features with advanced imaging findings; in this case, these findings were suggestive of Lev's disease.

A unique aspect of this case is that the patient presented with complete AV block prior to intervention. Most reports of conduction disturbances in severe AS emphasize new-onset AV block following TAVI. Our findings suggest that in some cases,

the substrate for conduction failure preprocedurally, and that advanced imaging can non-invasively demonstrate septal calcification that directly involves the conduction axis. This highlights the value of CT in risk stratification, complementing echocardiography by not only characterizing valvular pathology but also anticipating electrical complications. Verhemel et al.^[7] recently demonstrated that CT-derived parameters such as septal calcification burden and membranous septum length predict permanent pacemaker implantation after TAVI. Similarly, Pagnesi et al.^[8] reported that patients requiring pacemakers after TAVI had higher mortality and rehospitalization rates, underscoring the prognostic importance of conduction system assessment.

Electrocardiographic surveillance is another pragmatic tool for detecting progressive conduction disease.^[9] Serial ECGs may document gradual PR prolongation or bundle branch involvement, which can signal advancing conduction system degeneration.^[9,10] Unfortunately, our patient lacked longitudinal ECG documentation a limitation that reduces diagnostic certainty and has been explicitly acknowledged. In such scenarios, electrophysiological studies could theoretically quantify conduction reserve and confirm diffuse His-Purkinje involvement; however, invasive assessment was not pursued because of the patient's frailty and the urgent need for valve intervention.

From a therapeutic standpoint, management must address both mechanical obstruction and electrical instability. TAVI is increasingly favored in elderly high-risk patients with severe symptomatic AS, but it does not reverse degeneration of the intrinsic conduction system.^[11] Indeed, conduction disease often persists or worsens after valve implantation, necessitating permanent pacemaker therapy. Recent registry data indicate that 8-25% of patients require new pacemaker implantation

post-TAVI, particularly those with pre-existing conduction abnormalities or septal calcification.^[3,12,13] Importantly, long-term studies show that pacemaker dependence after TAVI is associated with higher mortality and adverse outcomes, reinforcing the clinical relevance of early identification.^[13]

CONCLUSION

This case underscores that degenerative conduction disease, such as Lev's disease, may remain clinically silent until irreversible AV block occurs. Unlike most reports describing new-onset conduction disturbances after TAVI, our patient had already presented with complete AV block, and CT demonstrated septal extension of valve calcification, strongly suggesting direct involvement of the conduction system. This finding indicates that advanced imaging before TAVI could help identify patients at higher risk of pacemaker dependence, allowing more accurate risk stratification and closer follow-up.

Ethics

Informed Consent: Written informed consent was obtained from the patient for the publication of this case report and the accompanying images.

Footnotes

Authorship Contributions

Surgical and Medical Practices: Ö.F.Y., C.K., Ö.K.M., H.S., U.A., Concept: Ö.F.Y., C.K., Ö.K.M., H.S., U.A., Design: Ö.F.Y., C.K., Ö.K.M., H.S., U.A., Data Collection or Processing: Ö.F.Y., C.K., Ö.K.M., H.S., Literature Search: Ö.F.Y., C.K., Ö.K.M., H.S., Writing: Ö.F.Y., C.K.

Conflict of Interest: No conflict of interest was declared by the authors.

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