

# Eustachian Valve Endocarditis in a Patient With Charcot-Marie-Tooth Disease

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## Abstract

Endocarditis is a serious infectious disease of the endocardial surface of the heart, predominantly involving the heart valves, and it results from the colonization and proliferation of microorganisms within the bloodstream. The condition primarily affects individuals with underlying cardiac abnormalities or those who have undergone invasive procedures. Symptoms may include pyrexia, fatigue, arthralgia, and new cardiac murmur. We present a case of a young male patient who had recently undergone surgery and developed eustachian valve endocarditis (EVE), a condition scarcely described in the literature.

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**Categories:** Cardiology, Internal Medicine, Infectious Disease

**Keywords:** trans-esophageal echocardiogram, echo, right infectious endocarditis, staph aureus bacteremia, eustachian valve endocarditis

## Introduction

Eustachian valve endocarditis (EVE) is a rare but potentially fatal infection of the eustachian valve, an embryological remnant of the inferior vena cava valve, within the right atrium of the heart. This typically occurs through seeding from bacteremia with an initial distant focus of infection or a recent invasive procedure [1]. Due to its rarity and diverse clinical presentation, the diagnosis and management of EVE can be challenging. In this report, we describe a case of EVE in a patient with no previous cardiac history who presented with fever and chills after an operation. We highlight the importance of early recognition and appropriate management of this unusual but potentially life-threatening condition.

## Case Presentation

A 35-year-old male with a history of Charcot-Marie-Tooth (CMT) disease presented with complaints of fever and chills. He had recently undergone total left knee arthroplasty (TKA) and noticed two days of worsening erythema and drainage around the surgical site. He denied any knee pain, chest pain, shortness of breath, cough, or intravenous (IV) drug, or alcohol use. Vital signs revealed a temperature of 37.4 °C, a blood pressure of 103/63 mmHg, and a heart rate of 87 beats per minute. His physical exam was remarkable for severe erythema extending from his left knee to his left foot with numerous open sores. The patient's laboratory investigations are shown in Table 1.

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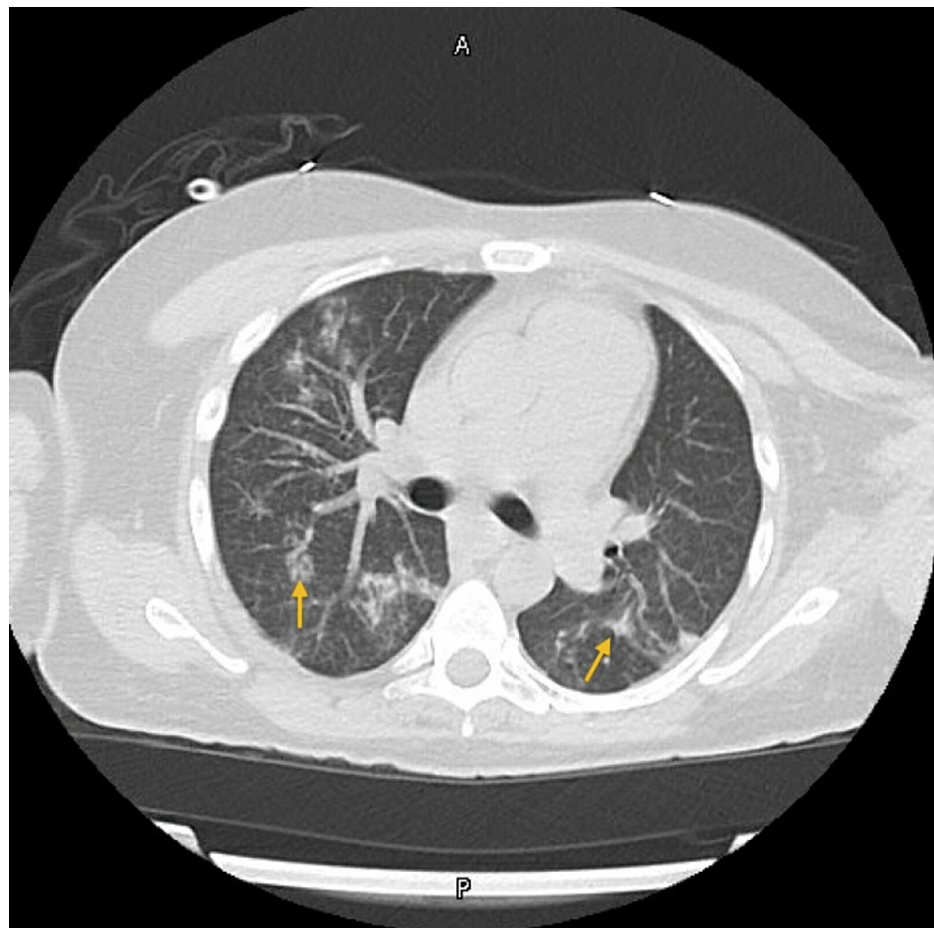
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Investigations	Results	Reference range
Complete blood count		
White blood cells	12.9 x 10 <sup>9</sup> /L	4.0–10.5 x 10 <sup>9</sup> /L
Hemoglobin	9.2 g/dL	12.0–16.0 g/dL
Platelet count	296 x 10 <sup>9</sup> /L	140–400 x 10 <sup>9</sup> /L
Basic metabolic panel		
Sodium level	138 mmol/L	133–147 mmol/L
Potassium level	3.7 mmol/L	3.5–5.6 mmol/L
Chloride	107 mmol/L	96–110 mmol/L
Carbon dioxide	11 mmol/L	20–32 mmol/L
Anion gap	20 mmol/L	5–15 mmol/L
BUN	113 mg/dL	5–27 mg/dL
Creatinine	3.85 mg/dL	0.40–1.40 mg/dL
Glucose level	93 mg/dL	60–100 mg/dL
Venous blood gas		
pH	7.30	7.35–7.45
CO <sub>2</sub>	28 mmHg	35–45 mmHg
Other laboratory values		
Lactate	1.1 mmol/L	0.5–2.0 mmol/L
ESR	125 mm/hr	0.0–12.0 mm/hr
CRP	466 mg/L	0.2–10.0 mg/L

TABLE 1: Laboratory investigations

BUN: blood urea nitrogen; ESR: erythrocyte sedimentation rate; CRP: c-reactive protein

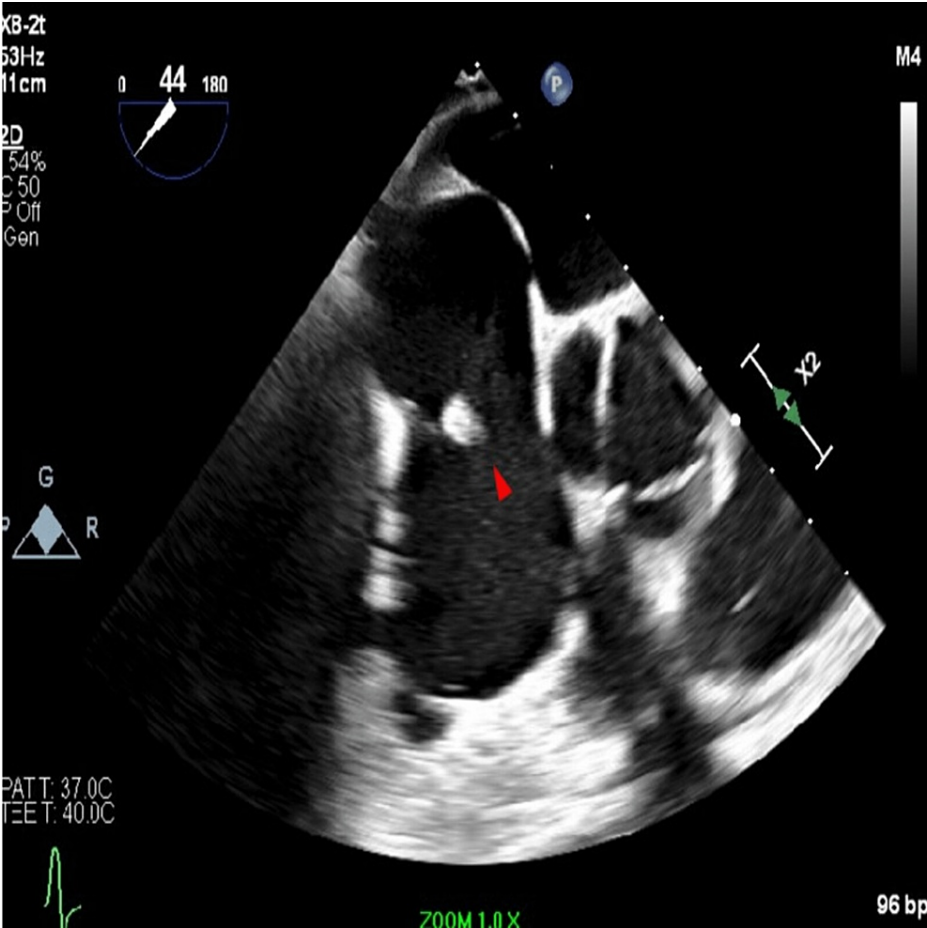
CT with contrast of the left knee revealed a large fluid collection along the anterior aspect of the left lower extremity, with generalized edema suspicious for cellulitis. Chest CT revealed multiple nodular lung infiltrates, suspicious for pulmonary septic emboli (Figure 1).



**FIGURE 1: CT chest showing nodular infiltrates concerning for septic emboli (arrows)**

CT: computed tomography

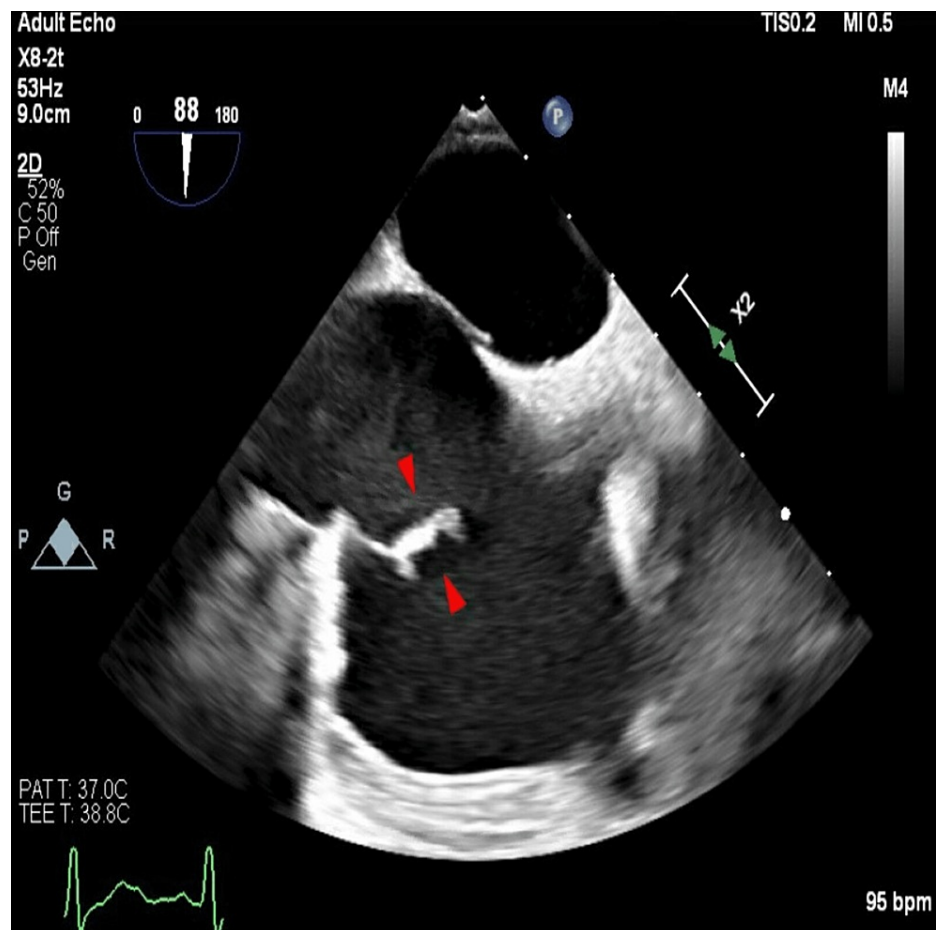
The patient underwent an urgent incision and drainage of the left knee, resulting in the aspiration of 20 mL of dark brown purulent fluid. Blood and joint aspirate cultures grew moderate *Staphylococcus aureus* (*S. aureus*) sensitive to methicillin (MSSA). Joint fluid culture revealed a white blood cell count of  $95,000/\text{mm}^3$  (reference range:  $13\text{--}180\text{ mm}^3$ ) and it was negative for crystals. The patient was given IV fluids, and nafcillin and admitted to the ICU due to MSSA bacteremia secondary to left knee septic arthritis and left lower extremity cellulitis. The patient had persistent bacteremia based on blood cultures and showed signs of shock including hypotension and decreased urinary output. Further investigation with a transthoracic echocardiogram (TTE) uncovered a mobile right atrial mass (Figure 2).



**FIGURE 2: TTE showing a mobile right atrial mass attached to the valve**

TTE: transthoracic echocardiogram

TTE was followed by a transesophageal echocardiogram (TEE), which revealed an unusually thickened eustachian valve within the right atrium visible with apparent fibrinous aggregate or vegetation, suspicious for endocarditis (Figure 3).



**FIGURE 3: TEE showing a thickened eustachian valve within the right atrium visible with apparent fibrinous aggregate or vegetation, suspicious for endocarditis**

TEE: transesophageal echocardiogram

The patient was deemed not a candidate for percutaneous aspiration of the vegetation. He improved hemodynamically and was transferred from the ICU to the medical rehabilitation unit where he completed a six-week course of nafcillin. He was scheduled for an outpatient follow-up TEE.

## Discussion

EVE is a rare condition that is unfortunately not well-documented in the literature. The incidence of EVE is estimated to be around 3% in right-sided infective endocarditis [2]. However, the condition is associated with significant morbidity and mortality. Early diagnosis and appropriate management are crucial for a good outcome. In fact, it has been reported that embolic events and in-hospital death occur in roughly 21% and 17% of cases respectively [3]. Patient populations that are at increased risk include IV drug users and those with previously implanted hardware. In addition, uncontrolled diabetes is a serious risk factor and is also associated with poor prognostic outcomes [4]. In our patient, the risk of infection was heightened as CMT can cause profound sensory loss leading to severe and difficult-to-treat infections [5].

*S. aureus* is the usual culprit in EVE, accounting for roughly 50% of the cases, although there have been reports of Gram-negative bacteria also playing a role in the pathology [6]. Diagnosis of EVE can be challenging due to its rarity and nonspecific symptoms. Echocardiography is a valuable diagnostic tool, as it can visualize the eustachian valve and detect any vegetations or masses on the valve, which are suggestive of infection. Recent studies have shown that TEE is a more sensitive modality for right-sided endocarditis than TTE, being able to detect smaller vegetations and complications such as valve perforations [7]. Our patient underwent both TTE and TEE, and the diagnosis was made based on TEE findings.

Treatment of EVE typically involves a combination of targeted antibiotic therapy and surgical intervention, which may include valve replacement or repair. While surgical intervention for vegetation removal is also an option, it requires the patient to be temporarily put on cardiopulmonary bypass [8]. Percutaneous

intervention is evolving as a new treatment option for these difficult cases. This method carries less risk as the cardiopulmonary bypass is not required and has been shown to reduce the size of the vegetation by as much as 60% [9]. However, percutaneous aspiration could also cause displacement of the vegetation into the pulmonary circulation, causing distal emboli and bleeding-related complications [10]. Unfortunately, our patient did not qualify for either of these therapies due to his concurrent infections and comorbidities. He showed improvement on antibiotics and was eventually discharged.

## Conclusions

EVE is a rare but serious condition that can lead to significant morbidity and mortality. Clinicians should maintain a high index of suspicion for EVE in patients with a history and findings of infective endocarditis, especially in those with nonspecific symptoms. It is an often-missed diagnosis. Early diagnosis and appropriate management are essential for a favorable outcome. Multidisciplinary care, including consultation with infectious disease specialists for recommendations on antibiotics and physiatry for rehabilitation, might be needed in patients requiring a prolonged hospital stay or those with extensive disease.

## Additional Information

### Disclosures

**Human subjects:** Consent was obtained or waived by all participants in this study. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

## References

1. Sawhney N, Palakodeti V, Raisinghani A, Rickman LS, DeMaria AN, Blanchard DG: Eustachian valve endocarditis: a case series and analysis of the literature. *J Am Soc Echocardiogr*. 2001, 14:1139-42. [10.1067/mje.2001.114012](https://doi.org/10.1067/mje.2001.114012)
2. San Román JA, Vilacosta I, Sarriá C, Garcimartín I, Rollán MJ, Fernández-Avilés F: Eustachian valve endocarditis: is it worth searching for?. *Am Heart J*. 2001, 142:1037-40. [10.1067/mhj.2001.119125](https://doi.org/10.1067/mhj.2001.119125)
3. Habib G, Lancellotti P, Erba PA, et al.: The ESC-EORP EURO-ENDO (European Infective Endocarditis) registry. *Eur Heart J Qual Care Clin Outcomes*. 2019, 5:202-7. [10.1093/ehjqcco/qcz018](https://doi.org/10.1093/ehjqcco/qcz018)
4. Biezma MI, Muñoz P, De la Villa S, et al.: Infective endocarditis in diabetic patients: a different profile with prognostic consequences. *J Clin Med*. 2022, 11:3-7. [10.3390/jcm11092651](https://doi.org/10.3390/jcm11092651)
5. Barisic N, Claeys KG, Sirotković-Skerlev M, Löfgren A, Nelis E, De Jonghe P, Timmerman V: Charcot-Marie-Tooth disease: a clinico-genetic confrontation. *Ann Hum Genet*. 2008, 72:416-41. [10.1111/j.1469-1809.2007.00412.x](https://doi.org/10.1111/j.1469-1809.2007.00412.x)
6. Fan J, Le PT, Jones BD: Eustachian valve endocarditis. *Proc (Bayl Univ Med Cent)*. 2019, 32:572-3. [10.1080/08998280.2019.1646595](https://doi.org/10.1080/08998280.2019.1646595)
7. Hasan S, Blanco A, Faluk M, Nasser H: Eustachian valve endocarditis: a case report on an under diagnosed entity. *J Community Hosp Intern Med Perspect*. 2020, 10:145-8. [10.1080/20009666.2020.1742474](https://doi.org/10.1080/20009666.2020.1742474)
8. Almanfi A, Nabous I: Percutaneous aspiration of vegetation in tricuspid valve infective endocarditis: a possible novel treatment option. *JACC Case Rep*. 2022, 4:1151-5. [10.1016/j.jaccas.2022.03.032](https://doi.org/10.1016/j.jaccas.2022.03.032)
9. George B, Voelkel A, Kotter J, Leventhal A, Gurley J: A novel approach to percutaneous removal of large tricuspid valve vegetations using suction filtration and veno-venous bypass: a single center experience. *Catheter Cardiovasc Interv*. 2017, 90:1009-15. [10.1002/ccd.27097](https://doi.org/10.1002/ccd.27097)
10. Moriarty JM, Liao M, Kim GH, Yang E, Desai K, Ranade M, Plotnik AN: Procedural outcomes associated with use of the AngioVac System for right heart thrombi: a safety report from RAPID registry data. *Vasc Med*. 2022, 27:277-82. [10.1177/1358863X211073974](https://doi.org/10.1177/1358863X211073974)