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Case report

# Sympathetic storm and rhythm of death "Spiked Helmet" in the Takotsubo cardiomyopathy. Case report

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## ABSTRACT

The "Spiked Helmet" sign (SHS) has been described in critically ill patients and is associated with a high risk of death. We present the case of a young individual with Marfan syndrome, who, 72 hours after the postoperative period from a ruptured abdominal aortic aneurysm, developed a Takotsubo cardiomyopathy and the electrocardiographic manifestation of SHS. In this case, the factors that may justify the presentation of this electrocardiographic pattern are the thoraco-abdominal surgical intervention and Takotsubo cardiomyopathy, which together activated the sympathetic system, triggering the clinical-electrocardiographic manifestation.

**Keywords:** Electrocardiography; Takotsubo Cardiomyopathy; Sympathetic Nervous System (Source: MeSH-NLM).

## RESUMEN

### Incendio simpático y ritmo de la muerte *Spiked Helmet* en la miocardiopatía de Takotsubo. Reporte de caso

El signo electrocardiográfico *Spiked Helmet* (SHS) ha sido descrito en pacientes críticamente enfermos asociándose con alto riesgo de muerte. Presentamos el caso de un joven con síndrome de Marfan, quien a las 72 h del periodo posquirúrgico de un aneurisma de aorta abdominal roto, presentó un cuadro compatible con miocardiopatía de Takotsubo y la manifestación electrocardiográfica de SHS. En este caso, los factores fundamentales que pueden justificar la presentación de este patrón electrocardiográfico son la intervención quirúrgica toraco-abdominal y la miocardiopatía de Takotsubo, que en conjunto activaron el sistema simpático de manera intensa desencadenando esta manifestación clínico-electrocardiográfica.

**Palabras clave:** Electrocardiograma; Miocardiopatía de Takotsubo; Sistema Nervioso Simpático (Fuente: DeCS-Bireme).

## Introduction

The electrocardiogram is a widely used tool due to its versatility, regardless of the scenario. There are specific conditions in which the electrocardiogram can guide the suspicion of potentially life-threatening pathologies beyond the heart. In 2011, the Spiked Helmet Sign (SHS) was described [1], and it has been reported in various clinical cases, with similarities regarding the critical nature of the underlying pathology. Recently, a systematic review of all reported cases was published [2]. To date, there are different theories about its presentation, all agreeing on excessive sympathetic activation; however, the pathophysiology is not completely understood [2-4].

## Case report

A 16-year-old male patient with Marfan syndrome (MS) presented in February 2023 with aortic dissection (Stanford A) extending to the celiac trunk, accompanied by severe aortic insufficiency. Additionally, the patient had an abdominal aortic aneurysm (AAA) measuring 4.7 cm in diameter. A modified Bentall-De Bono procedure was performed using a valved mechanical tube.

At 3 months, the patient experienced a rupture of the AAA with a retroperitoneal hematoma, leading to the decision to replace the thoraco-abdominal aorta with a straight Dacron graft. During the postoperative period, at 12 hours, surgical re-exploration was required due to bleeding. At 72 hours, the patient showed hemodynamic deterioration, multi-organ failure, and electrocardiographic changes consistent with SHS **(Figure 1 A and B)**.

The echocardiogram showed segmental wall motion abnormalities, akinesia of the apical segment of the septal and lateral walls, hyperkinetic basal segments, and an ejection fraction of 45%, findings consistent with a Takotsubo pattern **(Figure 2, Video 1)**. Cardiac biomarkers were elevated, with a peak troponin T of 239 ng/L and NT-proBNP of 34,245 pg/mL at 24 hours post-discovery. The patient's critical and unstable clinical condition precluded transfer for coronary angiographic study, given the low pretest probability for coronary artery disease. Additionally, the patient was not a candidate for mechanical support (such as intra-aortic balloon pump or extracorporeal membrane oxygenation), leading to the decision to optimize pharmacological support, replacing dobutamine and norepinephrine with levosimendan and vasopressin.

During the following 36 hours, the patient experienced episodes of non-sustained ventricular tachycardia. Forty-eight hours after the reported electrocardiographic changes, the ST segment returned to its baseline form, although a prolonged QT interval (520 ms and QTc by Bazett: 542 ms) was noted **(Figure 1C)**. There were no alterations in serum electrolyte levels, and the cardiac biomarkers showed a downward trend, along with a gradual resolution of the concomitant multiple organ failures.

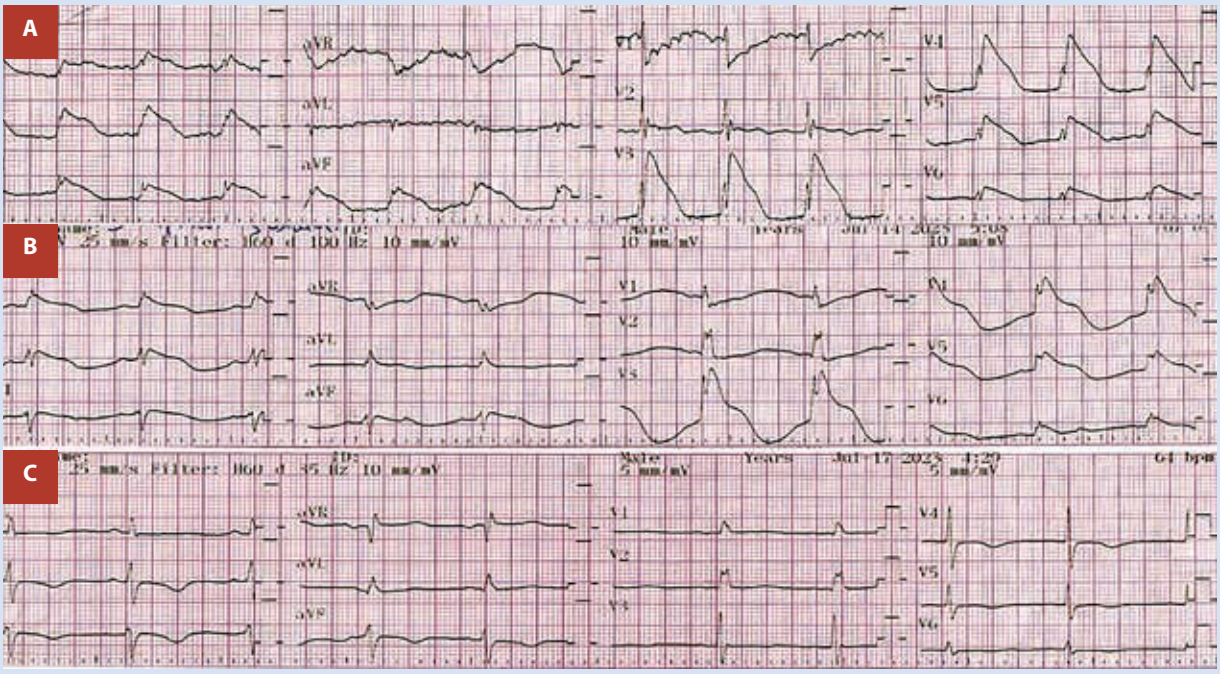


**Figure 1.** Spiked Helmet Sign that persisted for 48 hours from diagnosis. **(A, B)** Tall and peaked R waves with convex ST segment elevation in the inferior leads (II, III, and aVF) and anterior leads (V3 to V6) up to 6 mm. **(C)** Subsequent electrocardiographic tracing.

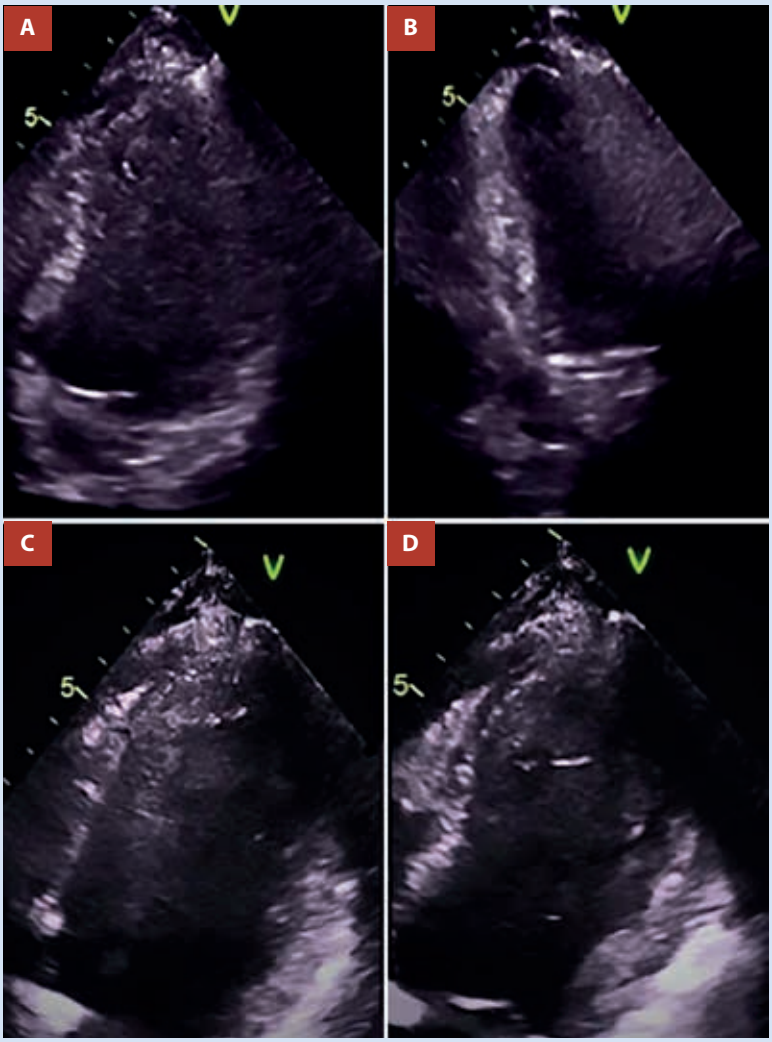


**Figure 2.** Transthoracic echocardiogram. In the two-chamber view **(A-B)**, akinesia of the apical segment of the anterior and inferior walls was observed, and in the four-chamber view **(C-D)**, akinesia of the apical segment of the inferior septal and lateral walls was noted..

The patient was extubated on the seventh day post-intervention and was discharged sixty-nine days after admission with optimal medical treatment for heart failure due to persistent ventricular dysfunction. During outpatient follow-up, a coronary CT scan revealed no coronary lesions; additionally, the echocardiogram showed reversal of the wall motion abnormalities, and the ejection fraction returned to its initial baseline value. At the time of writing this report, the patient remains stable and in good functional class.

## Discussion

The presented case is novel due to the association of the SHS electrocardiographic pattern concomitant with Takotsubo cardiomyopathy, which complicated the evolution of a thoraco-abdominal surgical procedure **(Figure 3)**. Undoubtedly, in any critically ill patient, ST-segment elevation raises alarms due to its immediate relation to acute myocardial infarction; however, ST-segment elevation does not always represent an acute myocardial infarction.

The SHS was described by Littmann *et al.* in 2011 [3] as an electrocardiographic marker associated with poor prognosis due to the potential risk of imminent death it implies. From an electrocardiographic perspective, its image resembles the helmet of German soldiers from World War I, and it is described as an apparent ST-segment elevation but with an upward displacement beginning before the appearance of the QRS complex [4,5]. The leads in which it appears can correlate with the underlying cause. In our case, its presence in the inferior leads would be associated with elevated intra-abdominal pressure [6,7], and the involvement of the anterior precordial leads would be related to Takotsubo syndrome.

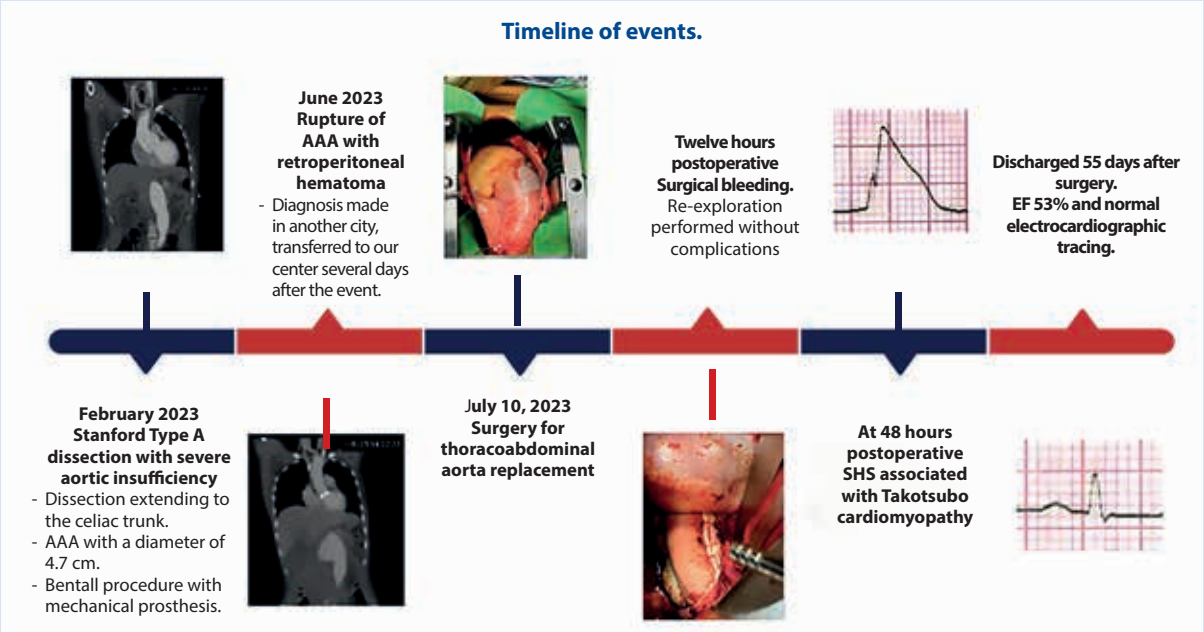


**Figure 3.** Timeline of events.

AAA: Abdominal Aortic Aneurysm; EF: Left Ventricular Ejection Fraction; SHS: Spiked Helmet Sign

To date, the mechanism behind SHS is not fully understood. The literature describes it in patients with critical conditions where elevated intra-abdominal and intrathoracic pressure is evident, as well as in patients with hemorrhagic cerebrovascular events [8]. In other scenarios, sympathetic discharge, pulsatile stimulations in the diaphragm, and Takotsubo cardiomyopathy could explain it [9,10]. In our case, several factors could justify the presentation of this pattern, such as the thoraco-abdominal surgical intervention and Takotsubo cardiomyopathy, with excessive sympathetic activation playing a particular role [11]. This is further supported by the subsequent electrocardiogram finding of a prolonged QT interval. The QT interval provides information about the state of ventricular depolarization and repolarization; thus, an acquired long QT interval indicates the presence of electrical instability, serving as a marker for malignant arrhythmias and sudden death.

One of our limitations was not performing an urgent coronary angiography (CCG), despite it being one of the fundamental criteria for diagnosing Takotsubo cardiomyopathy. Due to our strong suspicion of this condition, we opted to use the echocardiogram as the first imaging tool, in which the typical pattern of apical ballooning was observed.

Performing a coronary cineangiography (CCG) could have helped differentiate between the current condition and an acute myocardial infarction (AMI). However, given the patient's age, the probability of an AMI was low, and the increase in troponins was relatively modest compared to the levels of NT-proBNP. Although this does not completely rule out the possibility of coronary artery disease, the patient's hemodynamic instability supported the medical decision not to proceed with additional studies, as these would not have significantly altered the course of treatment.

In the literature, 39 patients have been described as presenting with this sign, which has been associated with a mortality rate of 59%. Additionally, this report could be the fourth case that associates SHS with Takotsubo cardiomyopathy [2].

Despite being associated with high mortality within the first 24 hours of its presentation [2], in our case, targeted intervention addressing the underlying cause was able to change the patient's prognosis. Similarly, there have been cases described where early correction of the underlying cause improved patient survival [8].

Therefore, we conclude that SHS is associated with critical conditions and a high risk of death, related to intense sympathetic activation and, as in our case, can be associated with Takotsubo cardiomyopathy.

## Ethical aspects

This case report has been conducted in strict compliance with the ethical principles established by the Declaration of Helsinki. Informed consent was explicitly obtained from the patient prior to data collection and manuscript preparation. The patient was informed about the nature of the study and its academic objectives. Written permission was granted by the patient for the publication of anonymous data from his case, including images and any other relevant information for understanding the case. The patient's identity has been protected at all times, and details that could allow identification have not been disclosed.

## Author Contributions

**PM:** conceptualization, methodology, supervision, Project administration. **MG:** data curation, writing - review & editing. **EK:** writing - original draft, investigation. **MB:** investigation, visualization. **CL:** validation.

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