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Case Report

Azygos Vein ICD Lead Implantation Lowers Defibrillation Threshold in a Patient with Hypertrophic Cardiomyopathy

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Abstract: A 14-year-old boy with hypertrophic cardiomyopathy (HCM) diagnosed at the age of 1 year and with massive left ventricular hypertrophy suffered an episode of ventricular fibrillation during mild effort. He underwent a dual-chamber implantable cardioverter defibrillator (ICD) implantation. The defibrillation threshold testing (DFT) was ineffective. Subcutaneous multi-coli arrays tunneled into the left postero-lateral position and connected to the superior vena cava (SVC) port of the dual-chamber ICD were added to increase the myocardial mass involved in the defibrillation shock pathway. A new DFT was unsuccessful. The patient was transferred to our hospital for myectomy. An epicardial defibrillation patch was placed on the left ventricular lateral wall, but again, DFT testing was ineffective using the right ventricular (RV) coil to lateral patch as shock pathway. Another epicardial defibrillation patch was then placed on the inferior wall. In this case, DFT testing was effective with a defibrillation pathway between the two patches and the can. In November 2015, a high shock impedance alarm was recorded through remote monitoring, thus compromising the safety of the ICD shock pathway. The patient underwent the implant of a new trans-venous defibrillation coil lead in the azygos vein. After few months, the patient developed symptomatic severe aortic regurgitation and underwent an aortic valve replacement. During the operation, DFT testing was performed and was successful. Our case illustrates that azygous vein ICD lead implantation is efficacious in HCM with massive hypertrophy and high DFT, and prompts further studies to systematically investigate its efficacy in this particular subgroup of the HCM population.

Keywords: hypertrophic cardiomyopathy; azygos vein defibrillation coil lead; defibrillation threshold testing

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Citation: Quarta, G.; Ferrari, P.; Giammarresi, A.; Malanchini, G.; Leidi, C.; Senni, M.; De Filippo, P. Azygos Vein ICD Lead Implantation Lowers Defibrillation Threshold in a Patient with Hypertrophic Cardiomyopathy. *Cardiogenetics* **2021**, *11*, 185–190. https://doi.org/10.3390/cardiogenetics11040019

Academic Editors: Giuseppe Limongelli and Lia Crotti

Received: 13 April 2021
Accepted: 12 July 2021
Published: 7 October 2021

1. Introduction

Hypertrophic cardiomyopathy (HCM) is the most frequent inherited heart muscle disease. The annual incidence of cardiovascular death, including sudden cardiac death (SCD), heart failure death and death due to thromboembolism, is around 1–2% in the adult population [1]. The implantable cardioverter defibrillator (ICD) has been proved efficacious in primary and secondary prevention of SCD [1]. We present a challenging case of a young patient with HCM and massive left ventricular hypertrophy (LVH) with high defibrillator threshold (DFT).

2. Case Presentation

A 14-year-old boy came to our medical attention for the first time in December 2008 following an episode of ventricular fibrillation (VF) during mild effort. He was previously diagnosed with HCM at the age of 1 year after a systolic murmur was detected. His growth was normal, but he developed massive LVH by the age of 13 years (maximal left ventricular wall thickness of 54 mm) and significant left ventricular outflow tract obstruction (LVOT) gradient of 50 mmHg. Patient was asymptomatic. Beta-blocker therapy was started (propranolol 20 mg three times a day). An ICD implantation was recommended but was declined by his parents.

In December 2008, the patient suffered a cardiac arrest due to VF while climbing a flight of stairs. He was successfully resuscitated with external defibrillation and was transported to a local A&E department where he suffered an electric storm with multiple episodes of VF requiring orotracheal intubation. The patient was extubated after 24 h, had no neurological complications, and underwent a dual-chamber ICD (Teligen, Boston Scientific, Inc., St Paul, MN, USA) implantation with a dual-coil defibrillation ventricular lead. The DFT was ineffective, and the induced VF was externally defibrillated. Subcutaneous multi-coli arrays (Boston Scientific, Inc., St Paul, MN) tunneled into the left postero-lateral position and connected to the superior vena cava (SVC) port of dual-chamber ICD were added to increase the myocardial mass involved in the defibrillation shock pathway (Figure 1). A new DFT was unsuccessful.

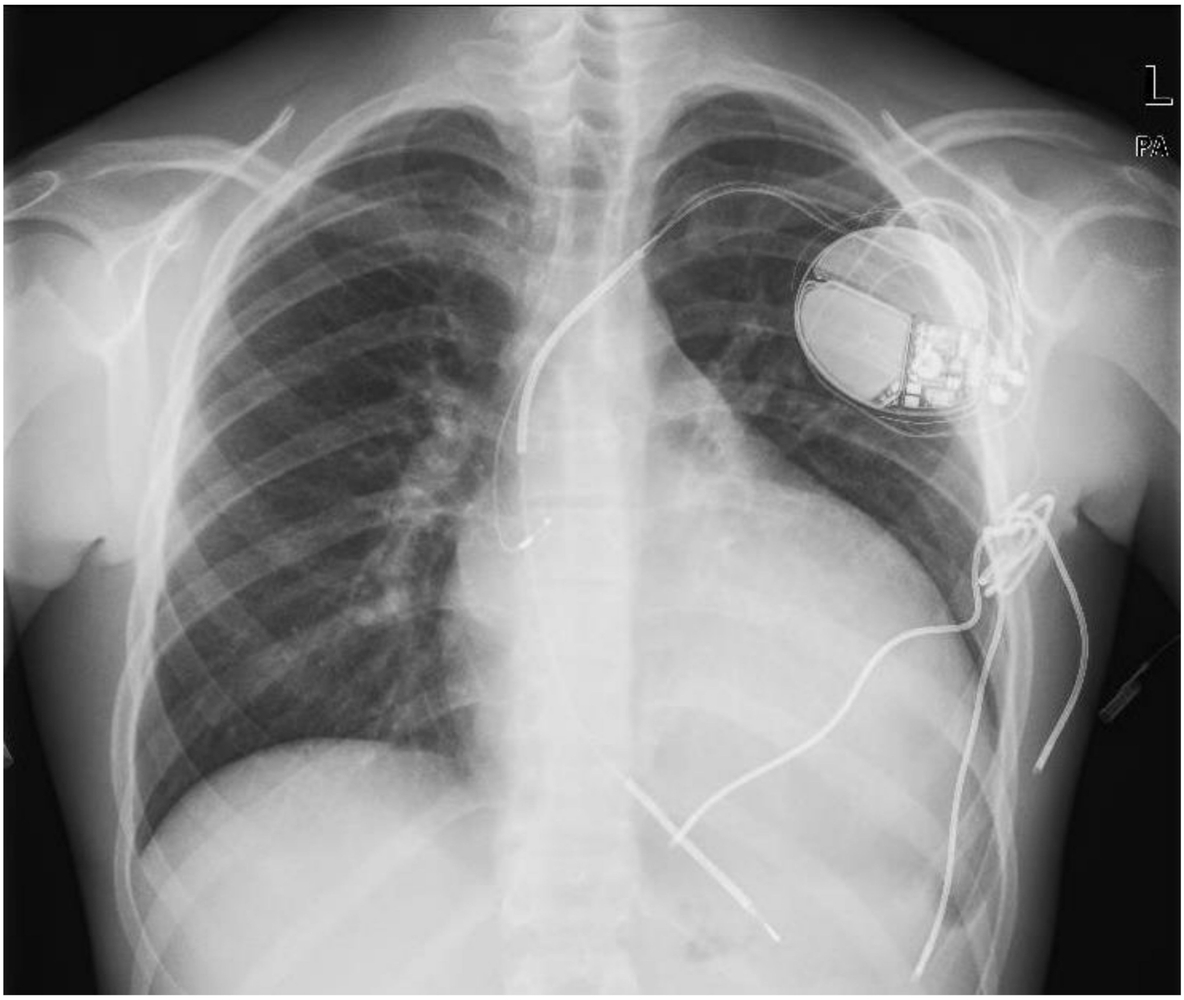

Figure 1. Anterior–posterior (AP) chest X ray showing dual-chamber ICD with a dual-coil defibrillation ventricular lead and subcutaneous multi-coli arrays tunneled into the left postero-lateral position and connected to the SVC port of ICD.

The patient was transferred to our hospital for myectomy. At the end of the operation, another DFT testing was performed. This time, both ICD shocks and external defibrillation were ineffective and only defibrillation with internal paddles directly connected to the heart was successful.

An epicardial defibrillation patch (Medtronic, Inc., ST Anthony, MN, USA) was placed on the left ventricular lateral wall, but again DFT testing was ineffective using the right ventricular (RV) coil to lateral patch as shock pathway. Another epicardial defibrillation patch was then placed on the inferior wall (Figure 2 left panel). In this case, DFT testing was effective with a defibrillation pathway between the two patches and the can. The subcutaneous multi-coli arrays were removed.

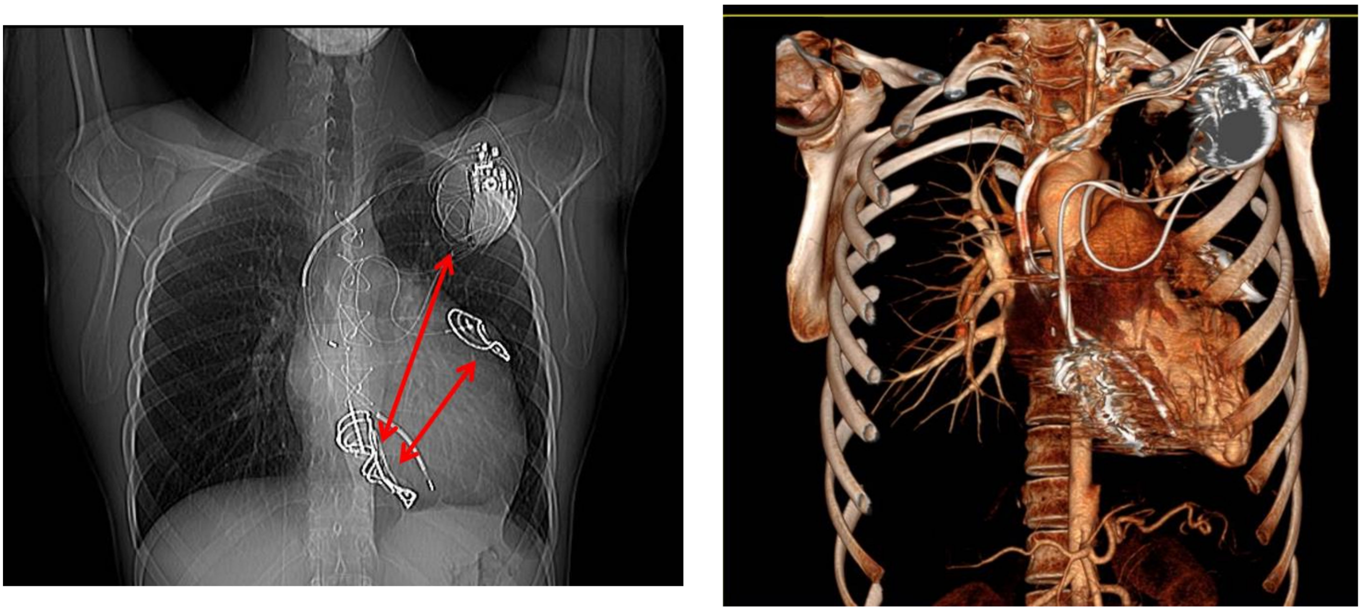

Figure 2. Left panel. AP chest X-ray showing dual-chamber ICD connected to two epicardial patches (lateral and inferior). The defibrillation pathway is between the two patches and the can (red arrows). Figure 2 right panel. CT scan shows strong adhesion of the patches with the underlying myocardium.

In September 2012, the left lateral epicardial patch showed high impedance (>200 ohm), indicating it was damaged. The inferior patch–can shock pathway was then programmed.

In November 2015, a high shock impedance alarm was recorded through remote monitoring, thus compromising the safety of the ICD shock pathway.

Several options were considered. The first one was surgical removal and replacement of the broken epicardial patches, but this was considered too dangerous due to possible strong adhesions of the patches with the underlying myocardium, with consequent risk of myocardial rupture (Figure 2 right panel).

The second option was performing a new DFT testing with the old endocardial leads, but it was considered unsafe, as it had already been ineffective during myectomy in 2008.

The third option was the implantation of a subcutaneous ICD, but unfortunately, screening ECG testing failed.

The last available option was to implant a defibrillation coil lead in the azygos vein.

The patient underwent the implant of a new trans-venous defibrillation coil lead (Medtronic, Inc. ST Anthony, MN) in the azygos vein (Figure 3). In order to maximize the defibrillation success, a new ICD (Inventra 7 VR-T, Biotronik, Inc., Lake Oswego, OR, USA) was implanted with a high nominal energy of 45 J (40.5 J delivered) from the first shock. After this implant, the connection of the high energy included the distal coil of the first endocardial lead and the new coil in azygos.

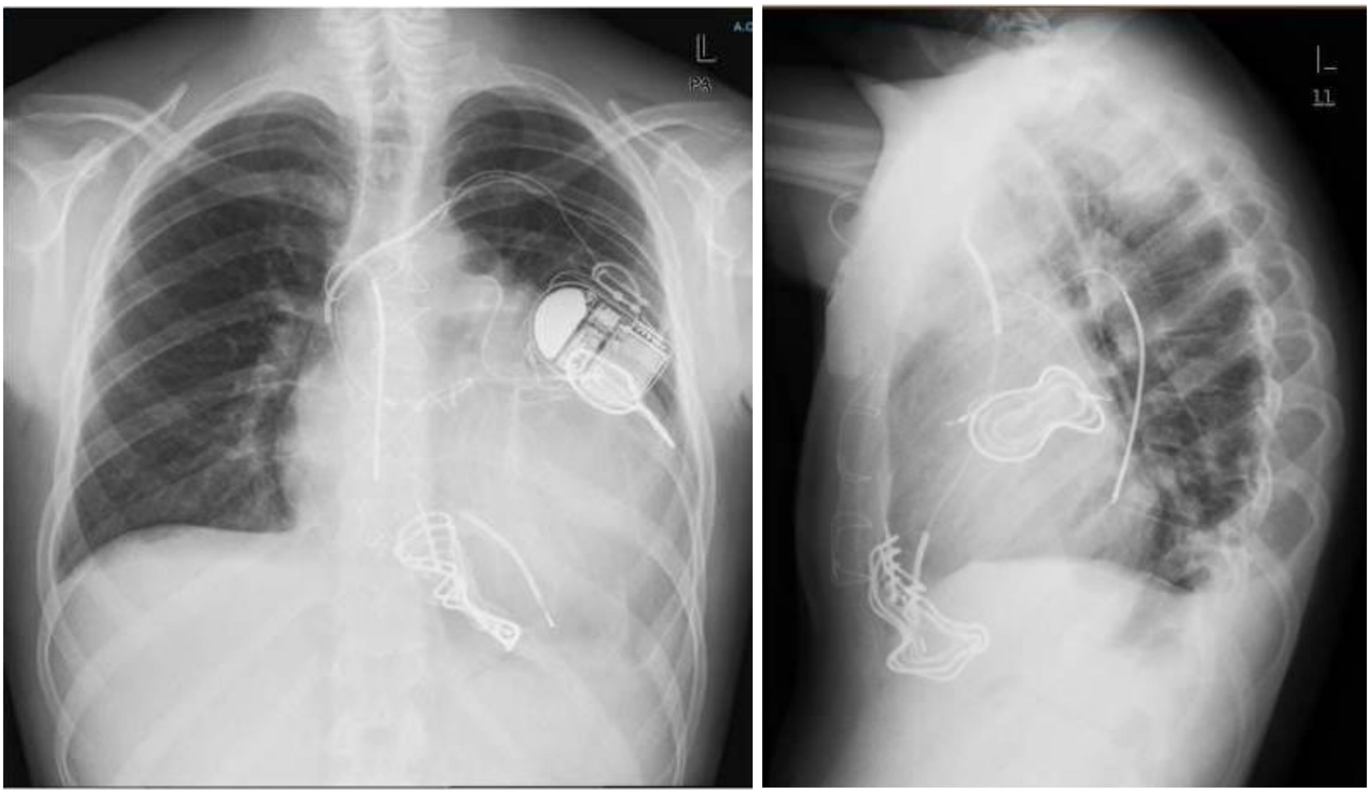

Figure 3. Lateral (left panel) and AP (right panel) chest X-ray after azygous vein ICD lead implantation. The shock pathway was formed by the old endocardial distal coil; the new floating coil was placed in the azygos vein and the can.

A new DFT testing was not performed immediately after the new implant. It was deemed too risky for the patient (last unsuccessful DFT required internal paddles shock); also considered were the controversy regarding the predictor value of DFT [2], the long arrhythmias event-free period after myectomy for the patient, the increased safety margin from the new device with the highest possible shock energy and the possibility to program different shock wave forms, including Bifasic-2 modality. In pre-market tests, bifasic-2 wave decreased DFT thanks to time-controlled waveform at 2 ms of the second tilt in the bifasic shock wave.

ICD was programmed with a VF zone above 200 bpm and a therapy including eight shocks at 45 J, automatic alternating polarity and a Bifasic-2 wave form. The shock pathway was formed by the old endocardial distal coil; the new floating coil was placed in the azygos vein and the can.

After few months, the patient developed symptomatic severe aortic regurgitation and underwent an aortic valve replacement. During the operation, DFT testing was planned with the back-up of open-chest defibrillation with internal paddles. VF induction was performed using a 1-J T-wave shock delivered after eight synchronized pacing pulses. VF detection zone was programmed above 188 bpm with a 6/8 counter criterion. Therapy included 45 J with normal polarity and Bifasic-2 waveform from the very first shock with the possibility to automatically invert the polarity in case of ineffective shock. VF was immediately induced, correctly recognized by the device and defibrillated with the first 45 J shock. The last ICD check performed in September 2020 showed stable parameters and no arrhythmic events.

3. Discussion

Our case illustrates that azygous vein ICD lead implantation is efficacious in HCM with massive hypertrophy and high DFT. ICD is highly effective in restoring normal rhythm in patients with HCM suffering life-threatening ventricular arrhythmias, with intervention rates of 10.6% per year for secondary prevention and 3.6% per year for primary prevention [3]. It has been shown that HCM patients have higher DFT and that DFT is linearly correlated with increasing left ventricular wall thickness [4], although these results have not been replicated in other studies [5]. Nevertheless, the clinical utility of DFT in predicting ICD efficacy has been questioned [2] in the general HCM population, but it is still advocated in the young and in patients with massive LVH [2]. In our HCM patient with massive LVH and previous episode of VF, high DFT required surgical implantation of two epicardial patches. When the latter broke, azygous vein ICD lead implantation allowed enough coverage of critical defibrillation mass with anterior–posterior direction to achieve a safe DFT. The azygos vein originates at the level of first/second lumbar vertebrae from tributaries of the renal and lumbar veins and runs along the rightward side of the vertebral column, joining the posterior aspect of the superior vena cava slightly superior to the upper border of the right main stem bronchus. The azygos vein has the great anatomical advantage of being posterior to the heart, and studies have demonstrated a DFT decrease with a combination shock pathway that includes a coil in the azygos vein [6,7]. Some case reports and case series [8–10] highlight the role of implantation of a defibrillator coil in the azygous vein as a valid strategy to reduce defibrillator threshold. This approach is reported to be feasible and safe [6]. Adding an azygos coil is successful in lowering the DFT in most cases [9]. This approach may be particularly useful with a right-side-placed ICD. It can be used alone or in combination with other external defibrillation devices such as subcutaneous arrays [10].

4. Conclusions

We presented a case of a young patient with HCM and massive left ventricular hypertrophy in whom the implantation of a defibrillator coil in the azygous vein was efficacious in reducing the DFT. Our case prompts further studies to systematically investigate its efficacy in this particular subgroup of the HCM population.

Funding: This research received no external funding.

Institutional Review Board Statement: Ethical review and approval were waived for this study due to the absence of recognizable external morphological aspects of the patient reported in the case.

Informed Consent Statement: Patient consent was waived due to the absence of recognizable external morphological aspects of the patient reported in the case.

Data Availability Statement: The data presented in this study are available on request from the corresponding author. The data are not publicly available due to privacy reason.

Conflicts of Interest: The authors declare no conflict of interest.

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