



## Inappropriate Defibrillator Shock Due to Oversensing. What to Do?

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### ABSTRACT

In spite of advanced designs of implantable cardioverter defibrillators (ICD) lead fracture remains the most frequent complications. ICD interrogation is mandatory for the diagnosis of all type of lead failures.

We report the case of an 18 years old boy, diagnosed with Jarvell and Lange Nielsen syndrome since he was 2 years. He received first Nadolol then he got ICD and a prophylactic left sympathectomy after he had a cardiac arrest. One year later, the ICD started delivering shocks, appropriate at first then inappropriate due to oversensing.

Long QT syndrome is a rare congenital disorder characterized by repetitive episode of syncope and cardiac arrest due to polymorphic ventricular tachycardia. The treatment is mostly pharmaceutical with B-blockers. The indication for an ICD is in class I for patients who survived cardiac arrest, and for patients who experience recurrent syncopal events under B-blockers. On the other hand, in case of contraindicated or not tolerated medical therapy, ICD is recommended as a class IIa, according the ESC guidelines of 2022.

An ICD must be interrogated frequently and have its measurements reviewed in order to detect lead dysfunction. The detection of high impedance suggests conductor fracture along with inappropriate defibrillator shocks delivered by the ICD.

Oversensing with normal pacing impedance is the initial electrical abnormality with either conductor fracture.(4)

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The fractured lead will either be left inside the patient or removed, depending on specific factors related to the patient and the ICD. (5)

**KEYWORDS:** LQTS Jarvel and Lange Nielson syndrome Inappropriate shock ICD

### INTRODUCTION

Sudden cardiac death continues to pose a life-threatening challenge.

The lack of symptoms in channelopathies causes a big concern to clinicians, making the identification of genetic factors of a significant importance.

Long QT syndrome is known as abnormally prolonged ventricular repolarization due to defects in cardiac potassium and sodium channels.

In spite of advanced mastermind sophisticated designs of implantable cardioverter defibrillators (ICD) lead fracture remains the most frequent complications. ICD interrogation is mandatory for the diagnosis of all type of lead failures.

### CASE REPORT

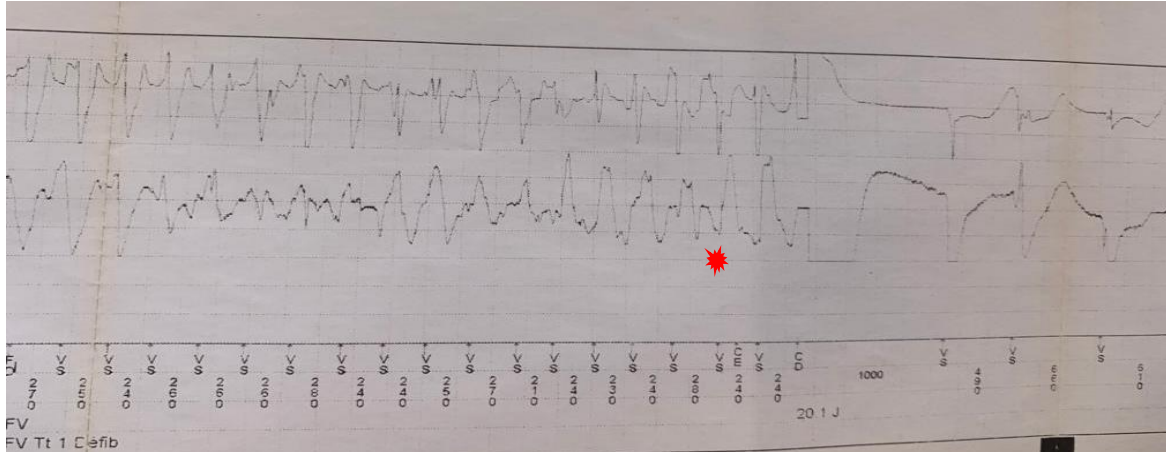
We report the case of an 18 years old boy, diagnosed with Jarvell and Lange Nielsen syndrome since he was 2 years. At the age of 1 deafness has been diagnosed, later that year, he

started fainting leading the diagnosis of long QT syndrome. TTE showed no structural defects, and blood analysis was normal. A karyotype was performed, revealing mutation in the KCNG1 gene which is responsible of type 1 LGTS. He received as a treatment Nadolol. However, he faints the moment he forgets his medication until one day he had a cardiac arrest. At that moment the decision of implantable cardioverter defibrillator was made. He got his device at the age of 12 years old, 3 months after that, he had prophylactic left sympathectomy.

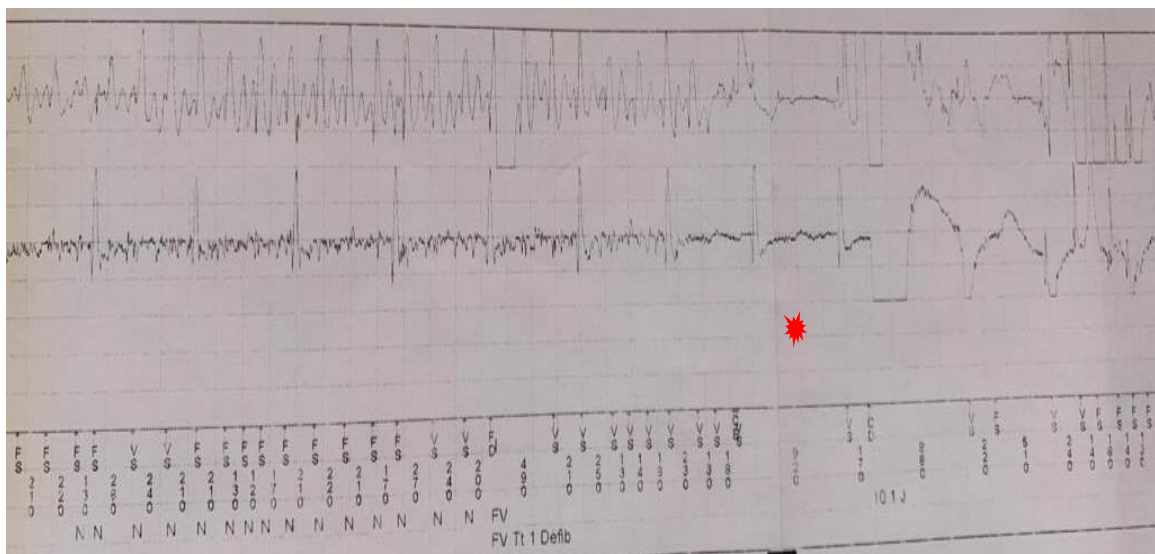
One year later, the ICD started delivering shocks. The boy skin around the ICD was normal with no signs of swelling or palpable hematoma. A chest X ray was performed, it showed no leads defects. After that we interrogated our device many times. We found first appropriate shocks due to non-sustained ventricular tachycardia (Figure 1 ) Then the next time higher impedance was found, until they became inappropriate

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shocks due to oversensing signals (Figure 2 ). At this step we decided to deactivate ICD therapy.



**Figure 1: The EGM shows Vfib, and an appropriate shock delivered (marked with the red asterisk)**



**Figure 2: The near filed EGM is clean with normal ventricular sensing confirming diagnosis noise-related oversensing. An inappropriate ICD shock was delivered (marked as red asterisk)**

The tachycardia cycle length having wide fluctuations with non-physiological rates

More to that, some intervals are too short to represent successive ventricular depolarizations.

### DISCUSSION

The heart is considered as an electromechanical pump, electrically triggered by an action potential. Which propagates across myocytes therefore, creates changes across the membrane.

Channelopathies are diseases that result from ion channel dysfunction, mostly caused by mutations. (1) They can either be inherited or acquired from de novo mutations, drugs or autoimmune attack of ion channels. Mutations can alter ion channel functions at any steps.

Long QT syndrome is a rare congenital disorder characterized by repetitive episode of syncope and cardiac arrest due to polymorphic ventricular tachycardia. The EKG shows QT

interval prolongation. (2) Out of the 12 forms of congenital LQTS only 3 forms have been characterized. The EKG reveals prolonged QT interval. The QTc corrected for heart rate can be calculated ( $QTc = QT \text{ interval} + \text{square root of the RR interval}$ ).

Usually asymptomatic, but when triggered it can lead to life threatening arrhythmias such Torsades de points, or ventricular fibrillation.

The treatment is mostly pharmaceutical with B-blockers. The indication for an ICD is in class I for patients who survived cardiac arrest, and for patients who experience recurrent syncopal events under B-blockers. On the other hand, in case of contraindicated or not tolerated medical therapy, ICD is recommended as a class IIa, according the ESC guidelines of 2022.

Studies have proposed that prophylactic left thoracic sympathectomy should be considered in case of recurrent episodes of syncope under maximal pharmacological treatment and for patients who suffer arrhythmia storms in the

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presence of an ICD. (3) According to the ESC guidelines of 2022, left thoracic sympathectomy is indicated as a class I if ICD is contraindicated or in case of recurrent ICD shocks.

For young active individuals, the decision of implantation of an ICD should be strongly considered as such as none procedure is immune from failure One of the most repeated complications of having an implantable device is lead fracture mainly due to chest trauma or weightlifting. Actually, lead fractures often take place next to the subclavian venous entry site due to compression between ribs and the clavicle.

An ICD must be interrogated frequently and have its measurements reviewed in order to detect lead dysfunction. The detection of high impedance suggests conductor fracture along with inappropriate defibrillator shocks delivered by the ICD.

The malfunctioning lead will generate electrical artifacts, which will be misinterpreted by the ICD as ventricular fibrillation, triggering defibrillation shocks.

Oversensing with normal pacing impedance is the initial electrical abnormality with either conductor fracture.(4)

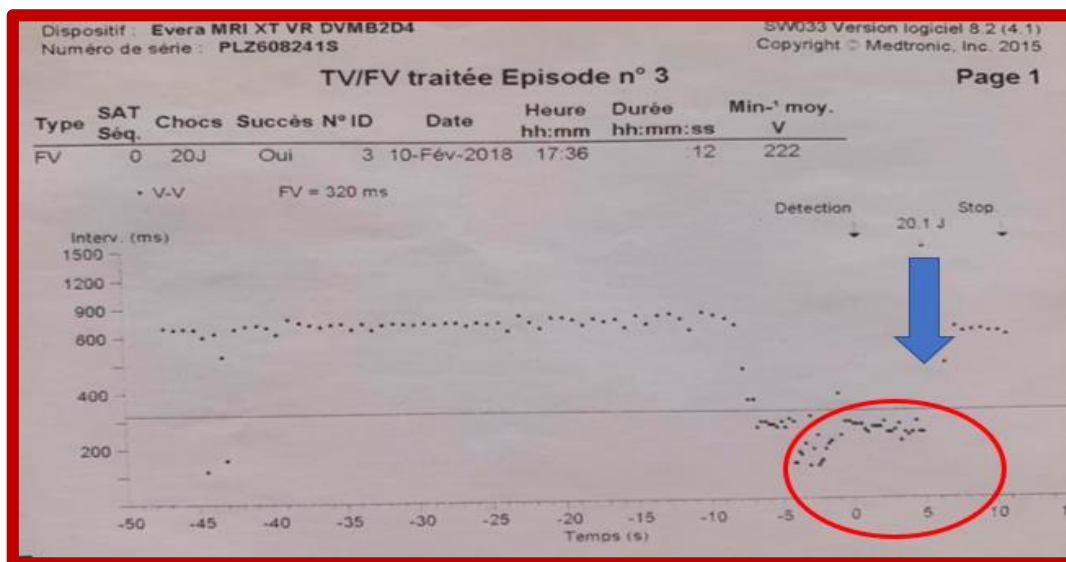
The fractured lead will either be left inside the patient or removed, depending on specific factors related to the patient and the ICD. (5)

The disadvantages of adding new leads without removing the failed lead include: multiple leads crossing the tricuspid valve, lead to lead interaction and increased risk of future lead related problems. However only experienced operators in high centers with surgical backup should perform extractions. (4)

### CONCLUSION

Those devices are not immune to failure. All shocks appropriate or inappropriate are associated with increased mortality. Lead fracture is a serious matter which necessitates placement of new leads or a subcutaneous ICD with or without removal of the fractured lead.

### Appendix1:



Dispositif : Evera MRI XT VR DVMB2D4  
 Numéro de série : PLZ608241S  
 Date de la visite : 16-Jul-2018 12:29:07  
 SW033 Version logiciel 8.2 (4.1)  
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**Rapport mesures pile et sondes** Page 1

Dernière interrogation : 16-Jul-2018 12:29:07

<b>Longévité résiduelle</b>	<b>16-Jul-2018</b>
Estimée à :	10.6 ans
Minimum :	9.7 ans
Maximum :	11.4 ans
RRT	> 5 ans (basée sur l'interrogation initiale)
<b>Tension pile</b>	<b>16-Jul-2018</b>
Tension	3.04 V (RRT=2.73V)
<b>Dernière charge</b>	<b>13-Mai-2018</b>
Temps de charge	3.7 s
Energie	0.0 - 13 J
<b>Compteur d'intégrité de la détection</b>	<b>Depuis 22-Jan-2018</b>
Intervalle V-V courts	16
<b>Impédance sonde</b>	
Stimulation VD (Bipolaire)	570 ohms 16-Jul-2018



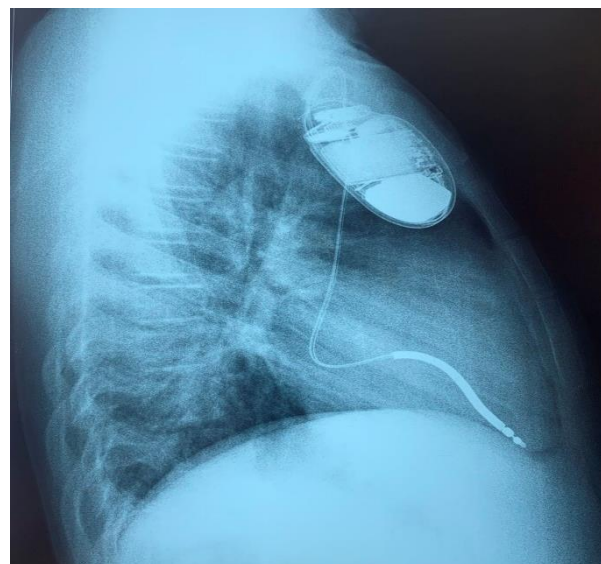
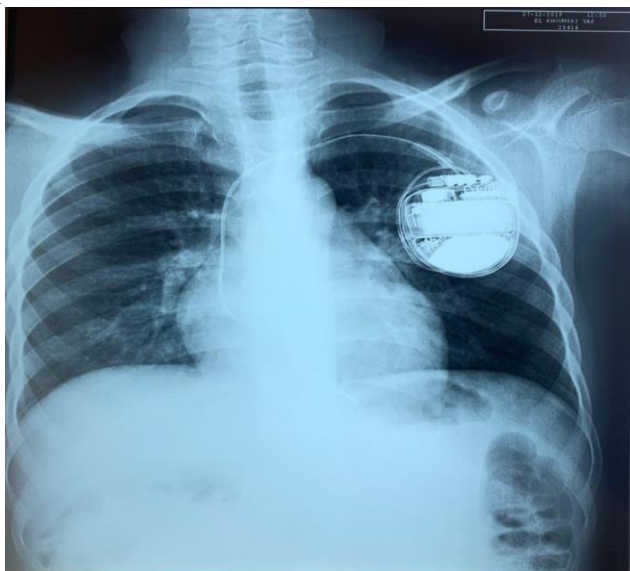
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Numéro de série : PLZ608241S		Copyright © Medtronic, Inc. 2020	
<b>Final : Résumé session</b>		<b>Page 2</b>	
<b>Etat dispositif (Implanté : 19-Jan-2017)</b>			
		<b>VD(6935M)</b>	
Impédance de stimulation		>3000 ohms	
Impédance de défibrillation		VD = >200 ohms	
Polarité stimulation		Bipolaire	
Contrôle du seuil		0.250 V à 0.40 ms	
Date mesure		10-Mai-2023	
Amplitude/durée d'impuls. programmées		2.00 V / 0.40 ms	
Onde R mesurée		4.9 mV	
Sensibilité programmée		1.20 mV	

Upper panel shows the detection of many episodes of tachycardia considered as Vfib until the ICD delivered a shock of twenty Joules.

Middle panel is the first ICD interrogation after receiving the first shock; it shows impedance of pacing lead at 570 ohms. Lower panel is the latest ICD interrogation showing higher shock impedance higher than 200 ohms, and pacing impedance higher than 3 000 ohms.

### Appendix 2:



The Chest X ray showed no lead defects.

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