1. Introduction

Since the first implant in 1980, the implantable cardioverter defibrillator (ICD) has become the first line therapy for sudden cardiac death and it’s technology has developed from a non-programmable device into a sophisticated multi-programmable, multi-functional device with extensive diagnostic and therapeutic options [1,2]. Programmable options of the new generation ICDs include arrhythmia detection, tachyarrhythmia therapy, pacing function, and stored intracardiac data.

An ICD consists of pulse generator, endocardial electrode, and defibrillation coils. ICD senses the rhythm and detects ventricular tachycardia (VT) and/or ventricular fibrillation (VF) and may deliver the following antitachycardia therapies [2,3]:

1. Anti-tachycardia pacing (ATP),
2. Low energy cardioversion,
3. High energy defibrillation.

All ICDs are programmed several heart rate zones to detect tachyarrhythmia. The VF zone is programmed to a rapid heart rate, often with a heart rate of ≥180 bpm. Monomorphic and stable VT may allow to program other therapy zones such as ATP. ATP may allow termination of a tachyarrhythmia via pacing impulses that are faster than the underlying VT rate and prevent defibrillator shocks (Figure 1-2) [2,4].

Randomized clinical secondary prevention trials have demonstrated the effectiveness of ICD therapy for arrhythmic death and total mortality in survivors of cardiac arrest [5]. Syncope of undetermined origin with inducible ventricular tachyarrhythmias is also indication for ICD implantation. Low incidence of sudden cardiac death and high incidence of appropriate ICD therapy has been shown in patients with syncope of undetermined origin and inducible ventricular tachyarrhythmias at follow-up [6]. Prophylactic therapy in patients at risk for
sudden cardiac death (primary prevention) was first based on the results of MADIT, MUSTT, and MADIT II, then later expanded upon results from SCD-HeFT [7-9]. Multiple randomized ICD trials reported that ICD benefit increases dramatically with time [10]. Indications for ICDs have expanded considerably since approval of the first ICD in 1985 [11]. Table 1 and 2 summarizes the Class I and Class II indications for ICD implantation in adults [11,12]. In this section we discuss ICD-related problems including implantation of device detection and therapy of VT/VF.

<table>
<thead>
<tr>
<th>Class I</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Survivors of cardiac arrest due to VT or VF not due to a reversible cause.</td>
</tr>
<tr>
<td>2. Spontaneous sustained VT in association with structural heart disease.</td>
</tr>
<tr>
<td>3. Syncope of undetermined origin with clinically relevant, hemodynamically significant</td>
</tr>
<tr>
<td>sustained VT or VF induced at EPS.</td>
</tr>
</tbody>
</table>

![Figure 1. Stored ventricular electrogram showing a successful anti-tachycardia pacing (ATP) therapy, converting a rapid ventricular tachycardia to sinus rhythm.](image1.png)

![Figure 2. Stored atrial and ventricular electrogram showing a successful anti-tachycardia pacing (ATP) therapy, converting a rapid ventricular tachycardia to sinus rhythm.](image2.png)
Emergencies of Implantable Cardioverter Defibrillator

Class I

4. Patients with LVEF less than or equal to 35% due to prior MI who are at least 40 days post-MI and are in NYHA functional Class II or III.

5. Patients with nonischemic DCM who have an LVEF less than or equal to 35% and who are in NYHA functional Class II or III.

6. Patients with LV dysfunction due to prior MI who are at least 40 days post-MI, have an LVEF less than or equal to 30%, and are in NYHA functional Class I.

7. Nonsustained VT due to prior MI, LVEF less than or equal to 40%, and inducible VF or sustained VT at EPS.

Abbreviations: DCM, Dilated Cardiomyopathy; EPS, Electrophysiologic Study; LVEF, Left Ventricular Ejection Fraction; MI, Myocardial Infarction; NYHA, New York Heart Association; VF, Ventricular Fibrillation; VT, Ventricular Tachycardia.


Table 1. Indications of ICD Implantation in Adults

Class IIa

1. Unexplained syncope, significant LV dysfunction, and nonischemic DCM.

2. Sustained VT and normal or near-normal ventricular function.

3. ARVD/C who have 1 or more risk factors* for SCD.

4. HCM who have 1 or more major risk factors† for SCD.

5. Brugada syndrome who have had syncope.

6. Brugada syndrome who have documented VT that has not resulted in cardiac arrest.

7. Catecholaminergic polymorphic VT who have syncope and/or documented sustained VT while receiving beta blockers.

8. Long-QT syndrome who are experiencing syncope and/or VT while receiving beta blockers.


10. Cardiac sarcoidosis, giant cell myocarditis, or chagas disease.

Class IIb

1. Nonischemic heart disease who have an LVEF of less than or equal to 35% and who are in NYHA functional Class I.

2. Familial cardiomyopathy associated with sudden death.

3. Long-QT syndrome and risk factors for SCD.
4. LV noncompaction.

5. Syncope and advanced structural heart disease in whom thorough invasive and noninvasive investigations have failed to define a cause.

Abbreviations: ARVD/C, Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy; CABG, Coronary Artery Bypass Grafting; DCM, Dilated Cardiomyopathy; EPS, Electrophysiologic Study; HCM, Hypertrophic Cardiomyopathy; LVEF, Left Ventricular Ejection Fraction; NYHA, New York Heart Association; SCD, Sudden Cardiac Death; VF, Ventricular Fibrillation; VT, Ventricular Tachycardia.

* The risk factors include induction of VT during electrophysiological testing, detection of nonsustained VT on noninvasive monitoring, male gender, severe RV dilation, and extensive RV involvement.

† The major risk factors include prior cardiac arrest, spontaneous sustained VT, spontaneous nonsustained VT, family history of SCD, syncope, LV thickness greater than or equal to 30 mm, and an abnormal blood pressure response to exercise.


Table 2. Indications of ICD Implantation in Adults

1.1. Implantable cardioverter defibrillator related problems

In the early years, ICD’s generator was implanted in the abdomen and epicardial leads and patches were placed via thoracotomy associated with significant morbidity and mortality [2]. With the new generation of endocardial leads and significant reduction in size of the devices, the procedure can be performed under local anaesthesia. Moreover, ICD generator is implanted in the pectoral region and endocardial lead is positioned at the right ventricular apex [2,3].

The ICD related problems can be classified as mechanical complications and pacing system malfunction including detection and therapy of VT/VF (Table 3). Mechanical complications are related to implantation procedure such as pneumothorax, hemothorax, subclavian artery puncture, thrombosis, pocket hematoma, infection, lead dislodgement, and myocardial perforation [3,13,14].

Venous access can be administered via the cephalic, subclavian, or axillary vein for ICD implantation and pneumothorax is mostly associated with the blind puncture approach of the subclavian vein. Review of randomized clinical trials showed that the incidence of pneumothorax was low and observed approximately 0.9% [13].

<table>
<thead>
<tr>
<th>Mechanical Complications</th>
<th>Pacing system malfunction</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pneumothorax</td>
<td>Shocks delivered by the device</td>
</tr>
<tr>
<td>Hemothorax</td>
<td>• Inappropriate ICD Shock</td>
</tr>
<tr>
<td>Subclavian artery puncture</td>
<td>• Electrical Storm</td>
</tr>
<tr>
<td>Thrombosis</td>
<td></td>
</tr>
<tr>
<td>Pocket hematoma</td>
<td>Ineffective Therapy</td>
</tr>
</tbody>
</table>
Mechanical Complications | Pacing system malfunction
--- | ---
Infection | Drug Effects on ICD
Lead dislodgement, and myocardial perforation | ICD-Pacemaker Interactions

Table 3. Implantable cardioverter defibrillator related problems

Pocket hematoma is not directly life threatening, but it may increase the risk of infection. Pocket hematoma was observed approximately 2.2% in clinical trials [13,15]. Infection is a rare complication of pacemaker and ICD implantation, with the incidence ranging from 0.8% to 5.7%. Clinical manifestations include skin infection, pocket infection, and endocarditis. Staphylococcus aureus and S. epidermidis are the most common organisms [3]. Endocarditis has a fearful mortality rate as high as 27% [16]. Randomized clinical trials demonstrated that infection rates decreased with routine use of antibiotic prophylaxis at the time of implantation or generator change and hence routine prophylaxis with antistaphylococcal antibiotics is recommended [3,17].

Venous thrombosis in the subclavian vein and superior vena cava was observed in 14% of pacemaker and ICD implantations in a prospective trial. Clinical manifestations vary from subclinical to superior vena cava obstruction syndrome [18]. Intracardiac thrombosis generally occurs in the right atrium around the lead. The size and location of the thrombus determine the clinical manifestation. Intravascular and intracardiac thrombosis may cause acute or recurrent pulmonary embolism. Treatment is usually initiated with intravenous heparinization and followed by oral anticoagulation [3].

Lead dislodgement with myocardial perforation is a rare complication with an incidence of <1% and it is defined as perforation of a device lead through the myocardium. Clinical manifestations vary from asymptomatic to sudden cardiac death [14].

Pacing system malfunction can be grouped into following categories [2,3,19].

1. Shocks delivered by the device
   a. Inappropriate ICD Shock
   b. Electrical Storm
2. Ineffective Therapy
3. Drug Effects on ICD
4. ICD-Pacemaker Interactions

2. Shocks delivered by the device

Although randomized clinical trials have demonstrated the effectiveness of ICD therapy, both appropriate and inappropriate ICD shocks are painful, physically and mentally disturbing, and potentially arrhythmogenic. In addition, inappropriate shocks are associated with a higher risk of all-cause mortality [20,21]. Even appropriate ICD shocks increase mortality, mostly due to progressive heart failure [22]. Most single shocks are appropriately
delivered to therapy of VT/VF, but multiple shocks are more often classified as inappropriate [2]. When a patient presents with an ICD shock, history, physical examination, and device interrogation should be performed. Stepwise approach for patients with ICD shock is essential and the first step is to differentiate a real tachycardia from a device recorded tachycardia originating from another source (Figure 3) [2,19].

**Figure 3.** Stepwise Algorithm for Patients with ICD Shock. Adapted from van Erven L, Schalij MJ. Arrhythmias: Troubleshooting implantable cardioverter-defibrillator related problems. Heart 2008;94(5):649-60. Abbreviations: EMI, Electromagnetic interference; ICD, implantable cardioverter defibrillator; SVT, supraventricular tachycardia; VF, ventricular fibrillation; VT, ventricular tachycardia.
2.1. Inappropriate ICD shock

Inappropriate shock is the most common adverse effect observed among ICD patients and the incidence varies from 13% to 22% [21-24]. Inappropriate shock may be caused by misdiagnosis of supraventricular tachycardia (SVT) as VT/VF or inappropriate sensing originating from internal and external sources (Table 4) [2,21]. These sensed events must occur at a rate higher than the programmed cut-off rate [2]. Atrial fibrillation (AF) is the most common cause of inappropriate shock, followed by sinus tachycardia, atrial flutter, atrial tachycardia and less commonly other types of SVT, like atrioventricular nodal reentry tachycardia [19,25,26]. History of AF, younger age, no statin use, interim appropriate shocks, a maximal heart rate during exercise close to the detection interval, and a low cut-off rate for VT-detection have been found to be independent risk factors for inappropriate shock [21,26].

Various algorithms, including tachyarrhythmia stability, sudden onset, and morphology of rhythm have been developed to discriminate between SVT and ventricular arrhythmias. Sinus tachycardia is differentiated from VT based on sudden onset criterion. While sinus tachycardia increases gradually, VT has a sudden increase in ventricular rate. But this criterion might not differentiate slow VT from the sinus tachycardia due to minimal cycle length difference [27,28]. Stability algorithm based on regularity of VT is an effective discriminator to differentiate AF from VT; however, AF with faster ventricular rate can be regular and VT can show some irregularity. Ventricular arrhythmias have different origin and wave form, resulting in a morphology district from supraventricular rhythm. These criteria help to discriminate between SVT and ventricular arrhythmias [2]. It is important to remember that these discriminators don’t work in the VF zone and hence adjusting detection intervals and therapy zones more carefully may reduce recurrent shocks. In addition, the use of medications and/or appropriate ablation procedure may prevent recurrences of SVT [19,29].

<table>
<thead>
<tr>
<th>Tachycardia Related Causes</th>
<th>Non-Tachycardia Related Causes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Supraventricular Tachycardia</td>
<td>Oversensing of Intracardiac Signals</td>
</tr>
<tr>
<td>• Atrial Fibrillation (most common)</td>
<td>• Oversensing of P or T Wave</td>
</tr>
<tr>
<td>• Sinus Tachycardia</td>
<td>• Double Counting of R Wave</td>
</tr>
<tr>
<td>• Atrial Flutter</td>
<td>• Sensing of Pacemaker Artifact</td>
</tr>
<tr>
<td>• Atrial Tachycardia</td>
<td>Oversensing of Extracardiac Signals</td>
</tr>
<tr>
<td>• Other Types of Arrhythmia (like AVNRT)</td>
<td>• Conductor or Connector Defect</td>
</tr>
<tr>
<td></td>
<td>• Myopotentials</td>
</tr>
<tr>
<td></td>
<td>• Electromagnetic Interference</td>
</tr>
</tbody>
</table>

Abbreviations: AVNRT, Atrioventricular Nodal Reentry Tachycardia; ICD, implantable cardioverter defibrillator.

Table 4. Causes of Inappropriate ICD Shocks
When the device interrogation doesn’t reveal a real tachycardia, the device recorded signals originate from intracardiac or extracardiac sources (Table 4) [2,30]. Oversensing of P or T wave and double counting of R wave are easily diagnosed from intracardiac electrocardiograms. T-wave oversensing occurs more frequently during exercise. A high T wave amplitude, a low R wave amplitude and younger age may contribute to T wave oversensing as a cause of inappropriate shocks. T wave oversensing and double counting of R wave can be managed by reprogramming the sensitivity level of ICD [2,30]. Device-related causes of inappropriate shocks include inappropriate sensing due to lead/device malfunction or dislodgement. Conductor defect or connector problems are associated with postural changes. Impedance may be within normal limits. On the other hand, if lead impedance is high, conductor fracture should be suspected, while low lead impedance is associated with insulation defect. Insulation defect may lead to oversensing of signals and inappropriate shocks. Upper extremity isometric exercises, deep breathing may lead to diaphragmatic oversensing and cause inappropriate shocks [2,19]. Electromagnetic interference (EMI) is recognised as high frequency, low amplitude signals that refers to noise on the ventricular channel from environmental sources. Potential sources of EMI are listed in Table 5. It is believed that using household appliances, such as televisions, radios, microwaves, toasters, and electric blankets do not effect functions of ICDs. A detailed history is useful in the diagnosis of this problem [2,19,31].

<table>
<thead>
<tr>
<th>Non-Medical Sources</th>
<th>Medical Sources</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Electronic Article Surveillance Devices</td>
<td>• Transthoracic Cardioversion</td>
</tr>
<tr>
<td>• Cellular Phones</td>
<td>• Magnetic Resonance Imaging</td>
</tr>
<tr>
<td>• Metal Detector Gates</td>
<td>• Radiation Therapy</td>
</tr>
<tr>
<td>• Arc Welding</td>
<td>• Electrocautery</td>
</tr>
<tr>
<td></td>
<td>• Lithotripsy</td>
</tr>
</tbody>
</table>

Table 5. Sources of Electromagnetic Interference

2.2. Electrical storm

Electrical storm refers to the occurrence of three or more episodes of VT or VF in a 24-hour period and it is associated with episodes of ATP (Figure 4) and/or multiple shocks (Figure 5) [3,32]. A patient with electrical storm should be immediately hospitalized and monitorized to determine the appropriateness of the shocks. Intravenous amiodarone is the first line therapy and the first step is to evaluate reversible causes of VT/VF such as electrolyte abnormality and ischemia. All clinicians should keep in mind that proarrhythmic effects of antiarrhythmic drugs may also lead to electrical storm. Intravenous β-blockers to suppress adrenergic stimulation, intubation and sedation, anti-ischemic therapy, intra-aortic balloon pump and ventricular assist devices for hemodynamic support have been used to suppress the ventricular arrhythmias. The ICD can be interrogated to ascertain programmed parameters to attempt pace termination of VT. Catheter ablation of VT/VF may be used as a last treatment way in selected patients [3,32,33].
**Figure 4.** Continuous intracardiac electrogram recordings from interrogation of an ICD in a 56-year-old man with sustained VT. Electrical storm terminates with four episodes of ATP therapy.

**Figure 5.** Continuous intracardiac electrogram recordings from interrogation of an ICD in a 62-year-old man with sustained VT. Electrical storm terminates with three episodes of high-energy defibrillation.
3. Ineffective therapy

Ineffective therapy refers to delayed or absent ICD therapy delivery during VT/VF and it can be lethal [2,3]. Undersensing and lack of detection of VT/VF, and device related problems are the main causes of ineffective ICD therapy. Lead malfunction or displacement, generator malfunction, exposure to EMI, use of antiarrhythmic drug, and pacemaker-ICD interaction may cause undersensing of VT/VF. High cut-off rate for detection, battery depletion with prolonged charge time, exposure to EMI, and slower VT may obstruct the detection of VT/VF [3]. Consequently, device failure have resulted in unnecessary deaths. Although these deaths may be infrequent, patients with an ICD should be evaluated to assess the battery status, charge time, lead integrity and function, and underlying rhythm every three months and after each exposure to EMI [3,34]. Alternatively, if remote monitoring is used, the office follow-up may be spaced out according to the recent HRS/EHRA guideline [35].

4. Drug effects on ICD

The primary indications to initiate antiarrhythmic agents in patients with ICD are suppression of the burden of the arrhythmias. Antiarrhythmic drug use has been reported to range from 49% to 69% in various trials [36]. Adding concomitant therapy with antiarrhythmic drugs to ICD patients results in following beneficial effects [3,36,37]:

1. Reduction in frequency of ICD therapy by decreasing the episodes of VT/VF or making them nonsustained,
2. Reduction of VT rate to allow ATP therapy,
3. Reduction in the frequency of recurrence of SVT to prevent inappropriate shocks,
4. Reduction in the defibrillation threshold (DFT),
5. Improvement in quality of life.

Although evidence supports above beneficial effects, the potential deleterious effects of antiarrhythmic drugs on ICD function include [3,36,37]:

1. Increased ICD discharge due to proarrhythmias or making them sustained
2. Slowing of the VT rate below cut-off detection rate for VT
3. Changing the QRS morphology that causes sensing alteration
4. Elevation of the DFT.
5. Elevation of the pacing threshold
6. Heart failure exacerbation

Drug effects on the pacing threshold vary with the class of drug and there is little or no effect on pacing threshold except class IC agents, especially flecainide [38]. The most potentially dangerous effect of drugs on ICD function is an increase in DFT that can lead to ineffective ICD therapy [3,36]. Generally, the drugs that block fast inward sodium channel and shorten the action potential duration can cause an increase in DFT, while potassium channel blocking drugs that prolong the action potential duration (e.g., sotalol) tend to decrease DFTs and these drugs are favorable choices in patients with high DFTs [3]. Class IA
drugs do not seem to have significant effects on the DFT whereas class IB drugs that block fast inward sodium channel (lidocaine and mexiletine) have consistently shown elevation in DFT [3,36,39]. The effect of class IC agents on the DFT is less clear. As opposed to the class I agents, class III agents tend no effect or to lower the DFT except amiodarone. Intravenous amiodarone had no effect on DFT, while oral amiodarone was associated with an increase in DFT in animal and human studies and hence DFT should be re-evaluated whenever antiarrhythmic agents especially amiodarone is administered [3,36].

5. ICD-pacemaker interactions

ICD-Pacemaker interactions involve only patients with separate PM and ICD systems implanted, which is a very rare scenario nowadays. The potential adverse interaction between ICD and pacemaker include [3,40]:

1. ICD effect on pacemaker function,
2. Pacemaker effect on ICD function.

Defibrillation shocks can lead to transient failure of pacemaker sensing and pacing because of exposure of the myocardium to high current density [40]. After an ICD shock, pacemaker function needs to be evaluated as reprogramming or damage to the PM system can occur. Bipolar pacing systems may be less sensitive to this issue. Another potential adverse effect is pacemaker reprogramming during ICD interrogation; this can be prevented by keeping adequate spatial separation between the pacemaker and ICD generators. In patients with ICDs and separate pacemakers, the pacing stimulation can cause ICD oversensing or undersensing and hence result in inappropriate therapy or failure to deliver therapy during VT/VF [3,40]. In addition, the electrical artifact between the pacemaker and the ICD lead can be oversensed by the ICD and cause inappropriate shocks [3].

6. Conclusion

Evolution of ICD technology allows accurate detection and therapy of arrhythmias, while also reducing unnecessary shocks and ineffective therapy. However, patients with ICD-related problems are increasingly encountered worldwide due the growing number of implantations. Regular outpatient follow-up is crucial to find out these problems before occurrence of clinical manifestations.

Consequently, clinicians, who may not be electrophysiologists, should use stepwise approach (Figure 3), gathered from the patient’s history, physical examination, and device interrogation as most problems can be solved by simple ICD reprogramming and/or a change in medical therapy.

Author details

Baris Bugan
Malatya Military Hospital, Cardiology Service, Malatya, Turkey
7. References


Implantation of Cardiac Pacemakers and Antiarrhythmia Devices) Journal of the American College of Cardiology. 51: e1-e62.


