

# Implantable-Cardioverter Defibrillator in Pediatric Population

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## 1. Introduction

Implantable cardioverter-defibrillator is largely used as an effective treatment for potentially lethal arrhythmias in adult population. On the contrary, just 1% of devices are implanted in pediatric population worldwide. In our series, 4% of defibrillators implanted between 2000 and 2010 were in children under 18 years of age.

During the last two decades, prevention and treatment decisions in pediatric population have been commonly made on adult data, including device therapy recommendations. Indeed, most pediatric data comes from single-center series and case reports, with no specific clinical trials focused on this population.

The incidence of pediatric sudden cardiac death is estimated at 1.3-8.5:100.000 patients-years (William et al., 1998). Survival of out-hospital cardiac arrest is as low as 8-9% and neurological sequels remain high (Driscoll et al., 1985). Sudden cardiac death in childhood and adolescence is associated with three main cardiovascular conditions: congenital heart diseases, cardiomyopathies, and genetic arrhythmia syndromes. In this setting, implantable defibrillator has become an effective antiarrhythmic therapy in a large variety of structural cardiovascular abnormalities and primary electrical diseases.

Device implantation in children is challenging because of peculiar patient characteristics as age, weight, vascular access and potential somatic growth. Therefore, procedural approach and site of implantation, therapeutic algorithms and early and long-term complications differ from adults. Transvenous approaches could be difficult because of small venous system (Radbill et al., 2010). Abdominal implantation of generator and epicardial leads are frequently needed. Early physical activity and impaired sterile conditions added to individual characteristic are related to more frequent procedural complications (Shah, 2009).

Device dysfunction, i.e. inappropriate therapies may appear frequently, between 11% and 50% of cases, in the settings of sinus and supraventricular tachycardia or T wave oversensing (Korte et al., 2004). Several algorithms have been used to reduce inappropriate discharges, as QRS discrimination or cardiac rate stability (Barry et al., 2001). Lead complications are related to high physical activity and body surface, significantly higher in patients with body area under 1.2 m<sup>2</sup> (OR 4.5) (Shah, 2009).

Psychological impact of defibrillator implantation and discharges during follow up may lead into more frequent depression and anxiety symptoms than in adult patients (Sears et al., 2011). Screening of inherited arrhythmic conditions in relatives of children carrying a defibrillator may be useful to detect potential risk in these relatives. Tomaske et al., 2011, reported 22% appropriate shocks in defibrillators implanted for primary prevention in this population.

## 2. Sudden cardiac death in children

Sudden cardiac death is defined as an abrupt, unexpected death occurring within 1 hour from the onset of cardiovascular symptoms. In young people, it typically occurs within a few minutes of symptoms onset. Aborted cardiac arrest includes cardiac resuscitation restoring spontaneous circulation. Excluding the Sudden Infant Death Syndrome, that affects children under 1 year, with an incidence around 1-1.5/1.000 infants, sudden death in a young person is a rare event (Gajewski et al., 2010). The estimated incidence of pediatric sudden cardiac death ranges from 1.3 to 8.5 per 100,000 children in the United States (Driscoll et al., 1985) (William et al., 1998). Approximately 20-25% of the deaths occur during sports (Liberthson et al., 1996). In patients with congenital heart disease, this rate increases to 100 deaths per 100,000 patients (O'Connor et al., 1998). Early cardiopulmonary resuscitation and extended availability of automatic external defibrillators could prevent about a quarter of pediatric sudden deaths (Gajewski and Saul, 2010). Since most sudden deaths have a cardiovascular cause, it is theoretically possible to identify patients at risk prior to the event and prevent it (Haskell et al., 2010).

Causes of sudden cardiac death	Relative incidence (%)
Hypertrophic cardiomyopathy	36
Increased cardiac mass	10
Coronary arteries anomalies	24
Marfan's Syndrome	6
Congenital heart disease	5
Myocarditis	3
Dilated cardiomyopathy	3
Arrhythmogenic right ventricular dysplasia	3
Ischemic heart disease	2
Commotio cordis	<1

Table 1. Causes of sudden cardiac death in children. Taken from Maron et al. JAMA. 1996.

Most young people with sudden cardiac death have an underlying heart condition, with hypertrophic cardiomyopathy, coronary artery anomalies, arrhythmogenic right ventricular dysplasia and long QT syndrome being commonest in most series (Silka et al., 1991, Maron et al. 1996a).

Hypertrophic cardiomyopathy is the most common cause of sudden unexpected death in childhood, significantly higher in the 8- to 16-year age range than in the 17- to 30-year (Maron et al., 1996a). Disease prevalence is as high as 1 per 500 in young adults (Maron et al., 1996b), (Corrado et al., 1998). Carriers of a genetic mutation may have little or no hypertrophy, especially earlier in life. Sudden death is often exertional and secondary to malignant ventricular arrhythmias. Lipophilic betablocker, disopiramid and implantable cardioverter-defibrillator have demonstrated to increase survival in this population.

Arrhythmia in children with dilated cardiomyopathy is one major clinical manifestation of the disease. The occurrence of arrhythmia is associated with the left ventricular size and heart function and includes ventricular ectopy (Han et al., 2011). An underlying myocarditis is found in 2-15% of patients, rising to 45% in a series of patients under 2 years, with other 25% affected by endomyocardial fibrosis (Meune et al. 2006). Other conditions as infectious, metabolic and neurological diseases have been described as causes of dilated cardiomyopathy. 20-25% of cases are inherited. Dilated cardiomyopathy is progressive, often clinically silent in childhood, and sudden cardiac death may occur prior to development of heart failure symptoms.

Left ventricular hypertrabeculation/noncompaction is a genetic myocardopathy affecting line-Z skeletal and cardiac contractile proteins. In children, it is found in 0,01% of echocardiographic explorations, meaning 10% of paediatric cardiomyopathies (Pignatelli et al., 2003). In pediatric population, diagnosis is usually made within first three months of life. Sustained or non-sustained ventricular tachycardia is seen in 40% of patients, and in 14% of patients QT interval is prolonged. Ventricular fibrillation is more frequent in children than in adults (Stöllberger et al., 2010). Almost 20% of patients with ventricular tachycardia or fibrillation have a normal systolic function. Data about long-term follow-up of patients with implanted cardioverter-defibrillator is necessary since indication for prophylactic implantation is still unclear.

The incidence of sudden death in patients with congenital heart disease is about 100/100,000 patient-years (O'Connor et al., 1998). It is higher in cyanotic and left heart obstructive lesions, may be due to arrhythmic, embolic or circulatory phenomena. Certain congenital defects have a higher risk of acquired arrhythmias following repair. The risk of sudden death appears to increase with age and time from surgery. Specifically, tetralogy of Fallot is associated with high incidence of ventricular tachycardia and 0.5% to 6% risk of sudden cardiac death (Gajewski et al., 2010). Patients with both single-ventricle physiology status-post Fontan, and transposition of the great arteries status-post atrial switch also have high acquired arrhythmia rates with increased incidence of sudden cardiac death. These two congenital cardiac conditions may lead to the implantation of a cardioverter-defibrillator as a primary prevention strategy.

Arrhythmogenic right ventricular dysplasia is a rare cause of sudden cardiac death in the United States, but is reported as the most common cause of sudden cardiac death in the young athletes in Italy (Maron et al., 2009), (Corrado et al., 2009). It is a heritable, progressive cardiomyopathy characterized by fatty and fibrous replacement of the

myocardium, causing thinning of right ventricular free wall. Although both drug therapy and catheter ablation are occasionally successful, implantation of a defibrillator is usually recommended for patients with significant symptoms.

A variety of conditions can cause primary arrhythmia in young people: Long QT Syndrome, Brugada Syndrome, Catecholaminergic Polymorphic Ventricular Tachycardia, Wolff-Parkinson-White Syndrome, and Congenital Complete Heart Block. Although there are cases in which sudden cardiac death is the first symptom, recurrent syncope often precedes malignant events (Proclemer et al., 2009). Fortunately, the surface 12-lead ECG is abnormal in most cases.

The congenital form of Long QT syndrome is a familial genetic disorder occurring about 1 in 2,500–3,500 individuals (Vincent et al., 1992). It manifests primarily as ventricular repolarization abnormalities caused by cardiac ion-channel mutations. For symptomatic patients, the presenting symptom is usually syncope, due to torsade-de-pointes ventricular tachycardia. The syncope may occur with specific triggers, such as stress, swimming, and loud auditory stimuli, or it may occur when the child is relatively bradycardic, at resting or sleeping (Schwartz et al., 2001). The specific phenotype (LQTS1, LQTS2 and LQTS3) can be predicted from the genetic mutation and may help in the assessment of risk for sudden death or response to therapy (Tester et al., 2005). Main therapy remains beta-blockade, which is less effective for LQTS3. If symptoms recur under beta-blocker therapy, implantation of a cardioverter-defibrillator is generally indicated.

Brugada syndrome is an inherited arrhythmogenic syndrome related to life-threatening ventricular arrhythmia due to a mutation in genes encoding sodium-channels (Miyamoto et al., 2011). Family sudden death history does not predict higher ventricular arrhythmia susceptibility (Delise et al., 2010). Treatment is limited to ICD implantation when symptoms like syncope occur.

Catecholaminergic polymorphic ventricular tachycardia is a genetic arrhythmogenic disease caused by mutations in genes encoding sarcoplasmic calcium ion-channels (Tester et al., 2006). Ventricular ectopy induced by exercise or emotional stress is typically observed. The onset of symptoms typically occurs in childhood and adolescence. Left untreated, Catecholaminergic Polymorphic Ventricular Tachycardia is lethal in 30–50% of patients (Leenhardt et al., 1995). Although beta-blockers are the recommended therapy, many patients present with recurrent arrhythmic symptoms and may need a defibrillator.

### **3. Cardioverter-defibrillator in pediatric population**

#### **3.1 Indications**

For the last decade, use of implantable cardioverter-defibrillator in children has increased dramatically. The number of pediatric implants per year has augmented by three-fold. The mean age at implant has decreased significantly (from 13.6 to 12.2 years), and the percentage of patients younger than 5 years of age receiving an implantable defibrillator tended to increase up to 10% (Burns et al., 2011). A large variability in the number of implants per center and year is observed and this situation may have implications for competency and training.

Specific pediatric recommendations have been included in the ACC/AHA/HRS 2008 Guidelines for Device-Based Therapy of Cardiac Rhythm Abnormalities (Ebstein et al., 2008):

Class I	<ul style="list-style-type: none"> <li>• ICD implantation is indicated in the survivor of cardiac arrest after evaluation to define the cause of the event and to exclude any reversible causes. Level of Evidence : B</li> <li>• ICD implantation is indicated for patients with symptomatic sustained VT in association with congenital heart disease who have undergone hemodynamic and electrophysiological evaluation. Catheter ablation or surgical repair may offer possible alternatives in carefully selected patients. Level of Evidence : C</li> </ul>
Class IIa	<ul style="list-style-type: none"> <li>• ICD implantation is reasonable for patients with congenital heart disease with recurrent syncope of undetermined origin in the presence of either ventricular dysfunction or inducible ventricular arrhythmias at electrophysiological study. Level of Evidence : B</li> </ul>
Class IIb	<ul style="list-style-type: none"> <li>• ICD implantation may be considered for patients with recurrent syncope associated with complex congenital heart disease and advanced systemic ventricular dysfunction when invasive and noninvasive investigations have failed to define a cause. Level of Evidence : C</li> </ul>
Class III	<ul style="list-style-type: none"> <li>• ICD therapy is not indicated for patients who do not have a reasonable expectation of survival with an acceptable functional status for at least 1 year, even if they meet criteria specified in the Class I, IIa, and IIb recommendations above. Level of Evidence : C.</li> <li>• ICD therapy is not indicated for patients with incessant VT or VF. Level of Evidence : C</li> <li>• ICD therapy is not indicated in patients with significant psychiatric illnesses that may be aggravated by device implantation or that may preclude systematic follow-up. Level of Evidence : C</li> <li>• ICD therapy is not indicated for NYHA Class IV patients with drug-refractory congestive heart failure who are not candidates for cardiac transplantation or CRT-D. Level of Evidence : C</li> <li>• ICD therapy is not indicated for syncope of undetermined cause in a patient without inducible ventricular tachyarrhythmias and without structural heart disease. Level of Evidence : C</li> <li>• ICD therapy is not indicated when VF or VT is amenable to surgical or catheter ablation (e.g., atrial arrhythmias associated with the Wolff-Parkinson-White syndrome, RV or LV outflow tract VT, idiopathic VT, or fascicular VT in the absence of structural heart disease). Level of Evidence : C</li> <li>• ICD therapy is not indicated for patients with ventricular tachyarrhythmias due to a completely reversible disorder in the absence of structural heart disease (e.g., electrolyte imbalance, drugs, or trauma). Level of Evidence : B</li> </ul>

Table 2.

Indications for implantation of cardioverter-defibrillator in children over last two decades are based on clinical trials designed and performed for adult population. Indications are shifting from secondary to primary prevention. In fact, secondary prevention implants decreased significantly when compared to primary prevention from 77% to 45% (Burns et al., 2011). In the Spanish Registry of Cardioverter-Defibrillator implantation, prophylactic implantation increased from 2006 to 2008 for Arrhythmogenic Right Ventricular Dysplasia and Brugada syndrome, with no increase for Long QT syndrome and hypertrophic

cardiomyopathy (Peinado et al., 2008). Primary prevention of sudden cardiac death remains a challenge in which individualized decisions play a mayor role. The low use of cardioverter-defibrillator in pediatric population difficult the assessment of cardioverter-defibrillator survival benefit and long term results.

In 1991, Silka et al. reported a series of 177 patients under 20 years in which a implantable cardioverter-defibrillator was indicated. 75% patients were survivors of sudden cardiac death, 10% had drug-refractory ventricular tachycardia and 10% syncope with positive electrophysiology study for arrythmia-inducibility. Almost 60% had an overt cardiovascular disease, whereas 26% had primary electrical condition and 18% congenital cardiopathy. Hypertrophic and dilated cardiomyopathies, transposition of great arteries and tetralogy of Fallot were commonest structural cardiac diseases. Systolic function was normal in 54% patients, and 48% had some degree of systolic function impairment. Von Berger et al., 2010 reported an updated registry of 210 cardioverter-defibrillators implanted in patients under 30 years-old in seven institutions between October 1992 and January 2007. Heart disease was categorized as electrical (n=90, 42%), cardiomyopathic (n=62, 30%), or congenital heart disease (n=58, 28%).

In the Dutch Registry from 1995 to 2006 (Heersche, 2010), 45 cardioverter-defibrillator were implanted in children and young patients. According to indication, sudden death and ventricular tachycardia patients were similar to the American registry, with a higher rate of electrical conditions (55%) and prophylactic indication (17%). In the electrical disease group, 56% had Long QT syndrome, 25% Idiopathic Ventricular Fibrillation and 19% Brugada Syndrome. Ten Harkell et al., 2006, reported another series of 23 Dutch pediatric defibrillator patients. 22% defibrillators were epicardial and 88% transvenous. The generator was placed in an abdominal position in 35% patients, whereas it was placed in the subpectoral region in 65%. There was no early mortality. Median hospital stay was 5 days.

In our series, 11 cardioverter-defibrillators were implanted between 1995 and 2010 in patients under 18 years, 4% of all implanted defibrillators. 80% defibrillators were implanted for secondary prevention (Ventricular Fibrillation 60%, Ventricular Tachycardia 20%) and 20% were implanted for primary prevention. Cardiac conditions were 40% Long QT Syndrome, 20% congenital cardiopathies, 10% hypertrophic cardiomyopathy, 10% hypertrabeculation/ Noncompaction cardiomyopathy and 20% had no overt structural heart disease. By age, long QT Syndrome was more frequent between younger patients, whereas transposition of Great Arteries was the commonest underlying cardiopathy in 15-18 years-age group. 50% were single chamber devices and 50% dual chambers.



Fig. 1. Comparison of three series of cardioverter-defibrillator implantation according to clinical presentation. Silka et al., 1991. Heersche et al., 2011. Granada, our series.

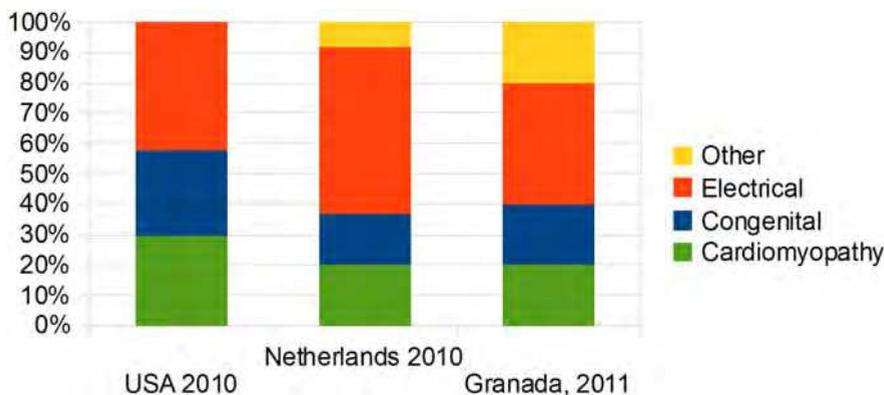


Fig. 2. Comparison of implantable cardioverter-defibrillation cardiac conditions in largest series. USA: Von Berger et al., 2010. Netherlands: Heersche et al., 2010. Granada, our series.

The Italian Registry included pediatric defibrillator use in inherited arrhythmogenic diseases from 2001 to 2006 (Proclemer et al., 2009). For primary prevention, 30% defibrillators were implanted in Hypertrophic Cardiomyopathy, 16% in Arrhythmogenic Right Ventricular Dysplasia, 17% in idiopathic ventricular arrhythmia (they included Brugada Syndrome in this group) and 16% in the Long QT Syndrome. There were 52% single-chamber, 44% dual-chamber, and 5% triple-chamber cardioverter-defibrillators.

In terms of defibrillation energy required in pediatric patients, available data includes only external resuscitation devices. The recommended energy dose had been established in 2 J/kg for the last 30 years, but recent reports may indicate that higher dosages may be more effective and safe. In 2005, the European Resuscitation Council recommended 4 J/kg as initial dose, without escalation for subsequent shocks (Haskell et al., 2010 & Sandroni et al., 2011).

### 3.2 Implantation techniques

Despite the increasing use of implantable cardioverter-defibrillator in congenital heart patients, specific challenges and implications related to implantation and follow-up are continuously observed. The variability and complexity of congenital pediatric patients make device management a highly individualized art. There are technical issues related to implantation since vascular access and device characteristic may not be suitable for pediatric patient (Chun et al., 2008). Although advances in implantable cardioverter-defibrillator technology are constantly made, the optimal cardioverter-defibrillator implantation technique for pediatric patients has not been established yet. The implantation of cardioverter-defibrillator in pediatric patients has many peculiarities, and there is little information on implant methodology for this population. A statement on training pathways for implantation of cardioverter-defibrillators and cardiac resynchronization therapy devices in pediatric and congenital heart patients was published in 2008 (Saul et al., 2008).

Transvenous implantation in children presents multiple challenges, related to patient body surface and weight, physical activity, increased risk of infections, and long life expectancy. There are no specific electrodes for small vessel diameters, with the consequent risk of venous thrombosis, nor devices adapted to their body surface. The creation of an atrial loop might allow the "elongation" of the lead with the growth. Concerns have been raised about Long-term leads patency, ventricular and valvular dysfunction, venous integrity, cosmetic results and psychological factors. System survival rates are estimated at 91% for the first year from implant, 83% at 24 months and 76% at 36 months (Rabdill et al., 2010).

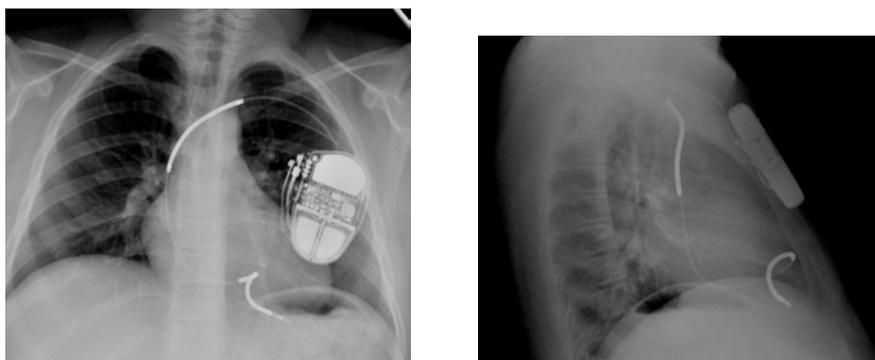


Fig. 1. and 2. Frontal and lateral Rx of a dual-chamber cardioverter-defibrillator in a 10 years-old patient with Brugada Syndrome.

Nontransvenous implantable cardioverter-defibrillator systems include pericardial and subcutaneous coils as alternative approaches in selected pediatric and congenital heart patients who are not candidates for transvenous leads. These nontransvenous systems are more commonly used in younger patients, with smaller body surface area, intracardiac shunts and concurrent thoracotomy surgery or affection of tricuspid valve.

Intrapericardial placement of an ICD coil system can be carried out through a subxiphoid approach and pericardial window without thoracotomy (Tomaske et al., 2008 & Bové et al., 2010). This technique is independent from child size or cardiac status. The defibrillation coil lead is actively fixated in the transverse sinus under fluoroscopic guidance, and the generator placed in a subrectus pocket in the upper abdomen through the same incision. Epicardial system is effective in treating ventricular arrhythmia without inappropriate discharges and no perioperative complications nor early or late deaths have been reported (Hsia et al., 2009). Controversy remains about defibrillation thresholds, since Stephenson et al., described high defibrillation thresholds with epicardial leads (Stephenson et al., 2006) More recently, Silvetti reported, for a 20-months follow-up, impedance stability and acceptable defibrillation thresholds (5-15J) (Silvetti et al., 2007).

Endocardial and epicardial steroid-eluting leads have comparable electrical performances, especially in absence of other congenital heart defects and previous heart surgery, although endocardial pacing shows the best outcomes and should be the first choice in children over 10-15 kg (Chun et al., 2008). System survival is significantly shorter in nontransvenous than in transvenous systems at 12, 24, and 36 months (survival rates at 73%, 55%, and 49%, respectively) (Rabdill et al., 2010). In fact, nontransvenous systems have demonstrated to be an independent predictor of system failure.

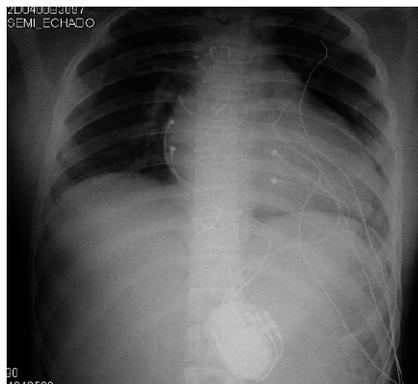


Fig. 3. Dual-chamber epicardial leads cardioverter-defibrillator in a male 6 years-old patient with Tetralogy of Fallot.

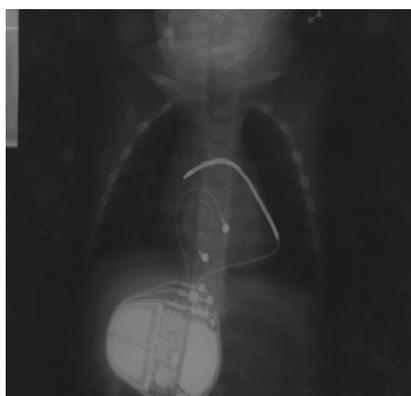


Fig. 4. Single-chamber epicardial cardioverter-defibrillator in a 2 years-old female child with Long QT Syndrome. See abdominal generator implantation, transverse sinus defibrillation coil and epicardial sense and pacing leads.

One increasing option is totally extracardiac implantation. A subcutaneous implantable defibrillator does not require a lead placed on or in the heart (McLeod et al., 2010). It may become an option for children suffering from chronic complications related to transvenous or epicardial leads and inappropriate shocks. High defibrillation thresholds at implant and follow-up are seen (Stephenson et al., 2006). Changing device position from abdominal to a supradiaphragmatic site may solve unsafe elevated discharge impedance and defibrillation threshold during follow-up (Berrueto et al., 2010). The best device configuration reported by Bardy et al., 2010, consisted of a parasternal electrode and a left lateral thoracic pulse generator. This configuration results as effective as a transvenous ICD for terminating induced ventricular fibrillation, albeit with a significantly higher mean energy requirement (35J vs. 11J). 100% ventricular fibrillation detection and 98% cardioversion effectiveness in two consecutive tests confirms its good performance. 100% appropriate shocks have been reported for treating ventricular tachycardia during 10-months follow-up. A low rate of adverse events confirms its safety.

The wearable cardiac defibrillator is an alternative for patients at risk for sudden death who do not fulfill standard criteria for defibrillator implantation or in whom the risk:benefit ratio is equivocal (Everitt et al., 2010). Careful patient selection and education result essential to ensure safety, as noncompliance with wear is common.

### 3.3 Procedural complications

Implantation procedure complications appear between 14% and 26% (Alexander et al., 2004), (Shah et al., 2009) (Stefanelli et al., 2004). These include pocket infections, pocket hematoma, microdislodgement requiring lead manipulation and electromechanical dissociation. Early electrode dislocation may need reintervention. In young patients, transvenous leads of implantable defibrillator can cause vascular obstruction up to 13%, mainly asymptomatic (Bar-Cohen et al., 2006). Local infection increasing rate may be due to early activity resume and impaired sterile conditions of wound (Link et al., 1999). Most pocket infections are related to local contamination at the time of implantation. Cardiac perforation (Morrison et al., 2009), hemothorax or superior Vena Cava syndrome have been described as implantation early and late complications (Alexander et al., 2010). Postpericardiotomy syndrome is described related to epicardial leads (Stefanelli et al., 2002). No pediatric death has been reported related to implantation procedure.

## 4. Long-term follow up

### 4.1 Therapy history

Pediatric defibrillator recipients have significant appropriate shock rates. Antitachycardia pacing therapy is rarely effective and often harmful in young ICD recipients, because this therapy is effective in monomorphic ventricular tachycardia, a rare arrhythmia among



Fig. 5. Appropriate shock delivery for ventricular fibrillation in an 11-years-old female child with hypertrophic cardiomyopathy. See instability in the cycle length of the sensed ventricular electrograms.

children. In most patients, programming ICD for only VF therapy is sufficient. A significant increased rate of appropriate discharges was found in defibrillator devices placed for secondary prevention (52%) versus primary prevention (14%) at 5 years (Von Berger et al., 2011). Therefore, the benefits of an implantable cardioverter-defibrillator remain greater in secondary than in primary prevention patients. In patients with nontransvenous systems, up to 23% receive appropriate shocks (Rabdill et al., 2010). In the Dutch registry, rate of appropriate shocks were reported at 31%, with a significant difference according to patient age (55% for patients under 12 years, 9% for patients between 13 and 18 years old). No difference has been reported in secondary prevention related to age, with rates of approximately 38% appropriate shocks for both groups (Heersche et al., 2010). In a registry of Long QT syndrome from 2002 to 2009, at least 1 appropriate shock was received by 28% of patients during 4 years mean follow-up.

## **4.2 Device-related complications**

Inappropriate discharges, lead-related complications and generator anomalies are the commonest adverse events occurring during follow-up. Lead complications are related to high physical activity and body surface, significantly higher in patients with body area under 1.2 m<sup>2</sup> (OR 4.5) (Shah, 2009). Lewandoski et al., 2010 reported 21% complications requiring surgical intervention. In our series, we describe 20% of inappropriate discharges, 20% lead complications and 10% generator anomalies. In the Long-QT-syndrome registry from 2002 to 2009, adverse events occurred in 25% (Schwartz et al., 2009). Serious psychological sequel may reach 43% of patients (Lewandowski et al., 2010).

### **4.2.1 Inappropriate therapy**

Inappropriate discharges are frequent, some of them caused by suboptimal pre-discharge programming of the device (Lewandowski et al., 2010). Reported rates vary sharply in infant series, from 11% to 50% (Botsch et al., 2007), being better defined in adult series (20-30%). Inappropriate shocks occur in the setting of sinus and supraventricular tachycardia, QRS double-sensing or T-wave oversensing (Korte et al., 2004). Lewandoski et al., 2010 reported inappropriate therapy resulting from T-wave over-sensing in 14%, sinus tachycardia in 5%, fast atrial fibrillation in 8%, and lead insulation disruption in 1%. Several algorithms have been used to reduce inappropriate discharges, as QRS discrimination or cardiac rate stability (Barry et al., 2001). In a multicentric series of 210 young defibrillator recipients from seven institutions, no differences were found in the risk of inappropriate discharges between primary and secondary prevention defibrillators, both rates estimated at approximately 35% within 5 years from implant (Berul et al., 2008). In the Dutch registry, 27% shocks deliveries were inappropriate. In patients with nontransvenous systems, up to 18% receive inappropriate shocks (Rabdill et al., 2010). Congenital patients have higher risk of inappropriate discharges (Williams et al., 1998). A higher rate of inappropriate shocks has been reported in the setting of lead failure than in other conditions causing inappropriate therapies.

### **4.2.2 Lead and generator specific complications**

As a general rule, less leads implanted in children, less complication will occur in the future, and the simplest system (generally, single-chamber), the better outcome (Silvetti et al., 2009).

The second most frequent ICD system-related adverse effect in the pediatric population is ICD lead failure. Lead failures requiring programming or revision interventions have been reported in the range of 7–30% at median follow-up of 2 years in the pediatric literature (Stefanelli et al., 2002 & Berul et al., 2008).

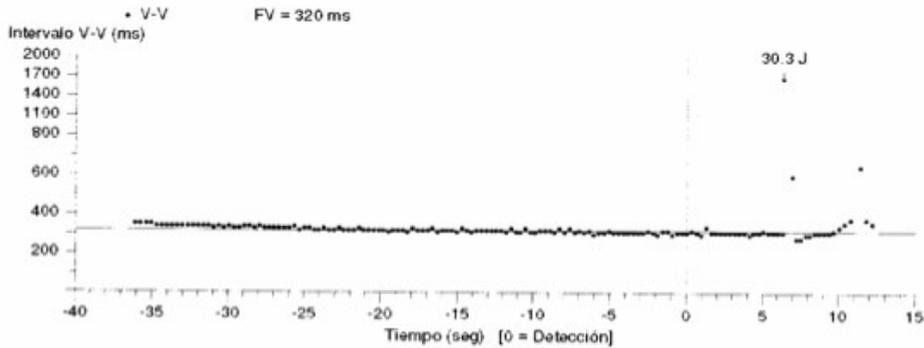


Fig. 6. Inappropriate shock during sinus tachycardia. See the progressive increase of heart rate, stability of cycles length and no further effect of defibrillation discharge.

In the Netherlands Registry, overall complications occurred in 17% patients, 87% related to lead failure (Heersche et al., 2010). Rate of total unanticipated interventions in the nontransvenous group is estimated at 18 per 1,000 person-months versus 6 per 1,000 person-months in the transvenous group (Rabdill et al., 2010). Survival rates for defibrillator leads in children are reported in 89.6% at 5-year, when implantation is made by an expertise operator. Lead failures as lead fraction and insulation failure (Bennett & Tung, 2010) occur mostly within the second year of implantation (Lewandowski et al., 2010). An increase in size was associated with higher risk for lead failure as the proximal shocking electrode ends to become stretched and distorted, leading to lead failure.

Given the finite longevity of current lead designs, lead extraction is an eventuality for a significant subset of pediatric defibrillator patients. Generator elective replacement is the most frequent indication for generator change in the majority of pediatric series.

Longevity is estimated at more than 9 years for a single-chamber defibrillator without permanent pacing, 7 years for a dual-chamber defibrillator pacing 50% of time and 6 years for a resynchronization-defibrillator (Bonney et al., 2010), in clinical practice the predicted generator survival is hardly accomplished.

Apart from lead durability, main indications for removal are vascular obstruction (that requires simultaneous revascularization), increased thresholds, and lead dislocations

(Welisch et al., 2010). Other complications affecting generator are prolonged charge time, early battery depletion, and malfunction during implant testing.

In Dutch series of Harkell et al., 2006, generator replacement was necessary in 18% of patients between 28 and 54 months from implantation. Procedural complications rates are low, according to a review of 203 lead extractions carried out between 2002 and 2008 (Cecchin et al., 2010). No procedure deaths were seen in this series, although removal of non-functional leads bears the risk of vascular disruption and embolizations. Of this series, 60% of patients had structural heart disease and successful simple extraction was only achievable in 29% of patients (requiring just a nonlocking stylet). Complex extraction techniques include radiofrequency-powered sheath (Zartner et al., 2010). Successful extraction was performed in 80% of all leads and 94% of complex extraction leads (Cecchin et al., 2010). Complications were observed in 5% of patients. Older leads, intraventricular location, and polyurethane insulation were associated with an increased probability of complex extraction. Procedure and x-ray duration correlated to time from lead implantation.

Cardiac device endocarditis is an infrequent, but potentially lethal complication. Hematogenous seeding of *Staphylococcus aureus* from a distant focus is the most common etiology in late infections. Cure is achievable in the large majority of patients under an aggressive antimicrobial regime and complete device removal. When the intravascular portion of the lead system cannot be aseptically separated from the pocket, removal of the entire system is essential (Shah, 2009). After device explantation and long-term standard antibiotic treatment to decrease risk for recurrent endocarditis, reimplantation requires additional caution (Mihalcz et al., 2008).

Industry advisories and recalls have an adverse economic, psychosocial and physical impact on pediatric defibrillator patients. Between 2000 and 2005, 25% of implanted defibrillators were affected by industry advisories or recalls (Mahajan et al., 2008), which meant 22% patients undergoing explantation after three years from implant. Just 2 of 89 explanted devices were defective, with loose headers as the unique failure observed.

#### **4.2.3 Progressive increase in defibrillator thresholds**

Failure of first cardioverter-defibrillator shock to terminate ventricular tachycardia or fibrillation was reported in 7% of pediatric defibrillator recipients during follow-up, mainly due to chronic rise in defibrillation thresholds (Stefanelli et al., 2002). Rates of significant changes of defibrillation thresholds range 3,2 to 12% (Stephenson et al., 2006 & Brodsky et al., 1999). Epicardial and subcutaneous systems are more likely to present with this complication than transvenous systems.

#### **4.2.4 Electrical storm**

Real incidence of electrical storm in pediatric patients is unknown, although Alexander et al., 2004 reported consecutive appropriate shocks in 6% patients. Antiadrenergic medical therapy and amiodarone have been used to treat this complication. Morbidity and hospitalization are direct consequences of this complication.

#### **4.2.5 Death rates**

The majority of reported deaths in the pediatric ICD patients appear to be related to intractable arrhythmias. Silka et al., 1991 reported 4% sudden death, 1% due to recurrent

ventricular arrhythmias. Alexander et al., 2004 observed 2% sudden deaths in patients with cardioverter-defibrillator, one of them due to intractable ventricular arrhythmia.

<b>Adverse events reported in cardioverter-defibrillator pediatric patients</b>	<b>%</b>
Implantation-related: Pocket complications: hematoma, infection Lead dislocation Cardiac perforation Hemothorax Venous thrombosis	14-26     13
Mid and long term follow-up: Inappropriated shocks Lead failure Generator failure (recalls, advisories) Increased defibrillation thresholds Electrical storm Death	 11-50 7-30  3-12  2

Table 3. Early and late complications observed in pediatric defibrillator patients. From Shah, 2009. Blank-spaces refer to unknown rates.

## 5. Cost-effectiveness study

Although more common in adult population, cost-effectiveness studies in pediatric patients are scarce. Because of differences in heart failure etiology, sudden death rates, and defibrillator complication rates, addition of a prophylactic cardioverter-defibrillator to conventional medical management has resulted not cost-effective in children with dilated cardiomyopathy, poor ventricular function, and symptomatic heart failure (Feingold et al., 2010). Total costs were estimated at \$433,000 for the defibrillator strategy and \$355,000 for the medical management. Although quality adjusted survival was greater in the defibrillator group, the defibrillator strategy was cost-effective only when the annual probability of sudden death exceeded 13%. The low sudden death rates in this population may justify the results. No data is available for other cardiac conditions that may benefit more clearly from the implantation of a cardioverter-defibrillator in pediatric population.

## 6. Quality of life in children with implantable cardioverter-defibrillator

Psychosocial and quality-of-life outcomes in pediatric patients with implantable cardioverter-defibrillators are poorer than in adult population. Anxiety and depression are highly related to defibrillator therapies. Shock-related anxiety is suspected to be particularly common (Sears et al., 2011). The PedsQL, the Device Severity Index, the ICD and Avoidance Survey provide data about Quality-Of-Life. Pediatric defibrillator patients have similar Quality-Of-Life outcomes to chronic ill children, with exception of lower physical Quality-Of-Life. Parent-observed reports show lower psychosocial and physical QOL than reported by children themselves. Up to 85% of children present with avoidance behaviors from

cardioverter-defibrillation implantation, with female children avoiding places more than male. Similar to adult samples, female patients reported lower psychosocial, physical, and cardiac Quality-Of-Life scores. Differently from other series, Sears et al. did not find discharges and medical severity affecting Quality-Of-Life negatively (Sears et al., 2009).

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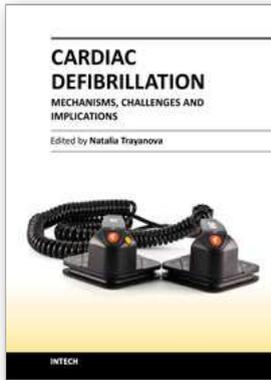
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## **Cardiac Defibrillation - Mechanisms, Challenges and Implications**

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The only known effective therapy for lethal disturbances in cardiac rhythm is defibrillation, the delivery of a strong electric shock to the heart. This technique constitutes the most important means for prevention of sudden cardiac death. The efficacy of defibrillation has led to an exponential growth in the number of patients receiving implantable devices. The objective of this book is to present contemporary views on the basic mechanisms by which the heart responds to an electric shock, as well as on the challenges and implications of clinical defibrillation. Basic science chapters elucidate questions such as lead configurations and the reasons by which a defibrillation shock fails. Chapters devoted to the challenges in the clinical procedure of defibrillation address issues related to inappropriate and unnecessary shocks, complications associated with the implantation of cardioverter/defibrillator devices, and the application of the therapy in pediatric patients and young adults. The book also examines the implications of defibrillation therapy, such as patient risk stratification, cardiac rehabilitation, and remote monitoring of patient with implantable devices.

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