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# ICD Implantations in the Pediatric and Young Adult Population

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## 1. Introduction

Large clinical trials in adult patients at risk for sudden cardiac death (SCD) have proven the efficacy of implantable cardioverter defibrillator (ICD) therapy for both secondary and primary prevention of SCD (Moss et al., 1996). In 1989, the first use of ICDs in young patients was reported (Kral et al., 1989). Since this initial report, ICD therapy has become increasingly important as a treatment approach in the pediatric population, despite the lack of randomized pediatric ICD studies. Nevertheless, children still present only 1% of all ICD recipients. The first large study of ICD implantations in patients below the age of 20 years was reported by Silka et al in 1993 (Silka et al., 1993). Since then many retrospective cohorts describing the clinical outcome of pediatric ICD therapy have been published. (Eicken et al., 2006; Ten Harkel et al., 2005; Heersche et al., 2010). More recently, two large multicenter studies evaluating the clinical outcome of ICD therapy in pediatrics and adults with congenital heart disease, including more than 200 children were published (Berul et al., 2008; Von Bergen et al., 2011). These studies have similar results, showing that ICD therapy in children appears to be effective, but with a higher rate of inappropriate shock therapy and complications as compared with adult series. The growing number of ICD implantations in children has now been followed by a decrease in the number of complications (Burns et al., 2011). Despite the lack of large randomized trials, efforts have been made to develop guidelines for ICD therapy in children. Recommendations for ICD therapy in primary and secondary prevention of SCD in adults have been formalized in the 2006 ACC/AHA guidelines (Zipes et al., 2006). Class I indications in pediatric and congenital heart disease (CHD) patients include aborted SCD without reversible cause. Sustained ventricular tachycardia (VT) associated with structural heart disease may be an ICD indication if not amenable to ablation or surgical resection (Blom 2008; Berul 2009; Zipes et al., 2006). In the following we will discuss the role of ICD treatment in children and young adults within different disease entities with an increased risk of SCD: 1) primary electrical heart disease; 2) cardiomyopathy, either hypertrophic or dilated; 3) operated or unoperated congenital heart disease. Finally, we will discuss the effect of ICD therapy on the quality of life and the different modalities of programming and implantation.

## 2. Primary electrical heart disease

Among the various disorders, presently known as primary electrical heart disease, the long-QT syndrome is by far the most prevalent among young patients. The congenital long-QT

syndrome (LQTS) is a genetic channelopathy with variable penetrance and characterized by prolongation of the QT interval on the electrocardiogram. It is associated with increased propensity for polymorphic ventricular tachy-arrhythmias and SCD in young individuals with normal cardiac morphology (Goldenberg et al., 2010). To date, more than 500 mutations have been identified in 12 LQTS genotypes, with the LQTS type 1 (LQT1) and LQTS type 2 (LQT2) genotypes accounting for nearly 90% of identified cases. Although there is a continuous growth in the number of ICDs used to prevent fatal outcome associated with life-threatening arrhythmic episodes in LQTS patients the identification of patients that will profit most of ICD implantation remains a challenge. Long QT syndrome without symptoms is increasingly recognized as family members are screened. It has, however, been studied that the risk of death does not increase by the death of a sibling (Kaufman et al., 2008). It is, therefore, important that the decision to implant a defibrillator is based on the own risk factors of an LQTS patient (Kaufman et al., 2008). Previous studies of highly symptomatic patients were more worrisome. In the era of genetic testing and device implantation, overall mortality is low with treatment. Device therapy, although effective, is not without complications and should be reserved for high-risk patients (Etheridge et al., 2007).

The first-line treatment of patients with LQTS remains the use of beta-blockers. These have been shown to be very effective especially in LQT1 and LQT2 patients in the prevention of cardiovascular events. However, in large studies a substantial number of LQTS patients were not treated by beta-blockers before ICD implantation. (Goldenberg et al., 2010). In high-risk patients beta-blockers reduce the risk of adverse events by about 70% (Goldenberg et al., 2010). These findings underscore the fact that beta-blocker therapy should be routinely administered in all high risk LQT1 and LQT2 patients without contraindications as a first line measure (Goldenberg et al., 2010). Patients with syncope during beta-blocker therapy are at high risk of life-threatening events, and ICD therapy should be considered in these patients. The risk of beta-blocker failure is highest in young children and in women (Jons et al., 2010).

There is a continuous growth in the number of ICDs in this patient group (Schwartz & Crotti 2008). Potentially, life-saving therapies were rendered at a 5% to 6% per year rate among those selected for ICD therapy. Similar inappropriate shock frequencies were also noted. Secondary prevention, genotype, and QTc predicted those most likely to receive appropriate therapy. In a recent study about ICD implantations in LQTS patients Schwartz et al showed that especially among the LQT3 subtype many patients were asymptomatic before ICD implantation, and probably did not need one (Schwartz et al., 2010). They showed that some patients in whom an ICD was implanted the supposed high risk was questionable. Although the ICD implant frequency is greatest among LQT3 patients, the greatest save rate has occurred among LQT2 women who were assessed to be at high risk (Horner et al., 2010).

However, when used in a high-risk LQTS population, ICD therapy seems to be a safe option (Monnig et al., 2005). Risk factors for appropriate ICD shocks are longer QTc intervals and being survivors of a cardiac arrest. However, betablockers should always be added to ICD therapy, while some patients might benefit from additional antibradycardia pacing, prolonged detection time, and a rate-smoothing algorithm to prevent recurrent episodes (Monnig et al., 2005).

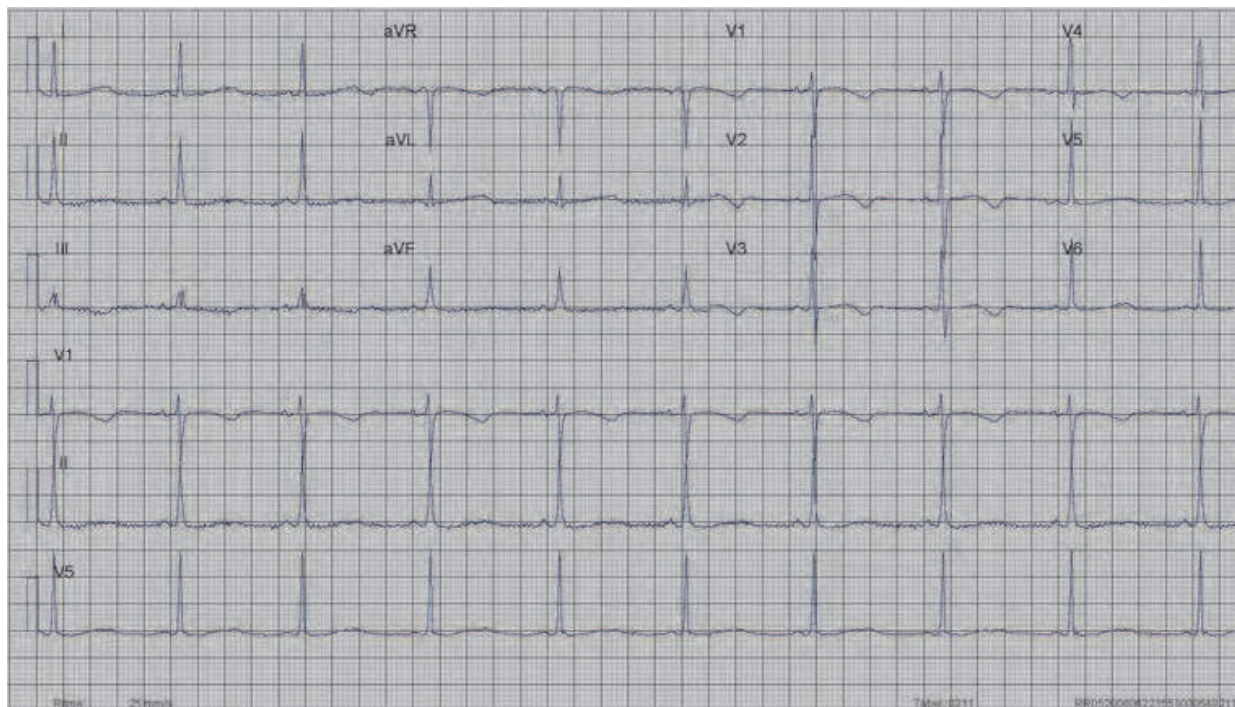


Fig. 1. Electrocardiogram of a 20-year old woman with the long-QT syndrome. A prolongation of the QT-interval of 520 msec is shown (normally less than 440 msec).

### 3. Hypertrophic cardiomyopathy

ICDs have proven effective in preventing SCD in young hypertrophic cardiomyopathy (HCM) patients with appropriate intervention rates of 11% for secondary and 4% for primary prevention, despite massive left ventricular (LV) hypertrophy, LV outflow obstruction, diastolic dysfunction or microvascular ischemia. Targeting candidates for prophylactic ICD therapy can be complex, compounded by the unpredictability of the arrhythmogenic substrate, the absence of a dominant risk factor, and difficulty in assembling randomized trials. However, a single major risk factor is often sufficient to justify an ICD, although additional markers and other disease features can resolve ambiguous decision-making. Nevertheless, the absence of all risk factors does not convey absolute immunity to SCD. However, when presenting with a normal electrocardiogram the patients seem at lower risk for cardiac events (McLeod et al., 2009).

Until now, there is, however, no consensus as to the indication for primary prevention with ICD implantation in children (Ostman-Smith 2010). Especially a high rate of inappropriate shocks up to 27% and a complication rate up to 17% necessitate careful considerations when an ICD implantation is to be planned in the pediatric age group (Ostman-Smith 2010).

The primary prevention risk factors in HCM are 1) family history of HCM related SCDs. When a high risk population that received primary prevention with ICD implantation was studied the number of appropriate shocks during follow-up did not differ between those with a family history of HCM related SCD, whether as an isolated risk factor or combined with other risk factors (Bos et al., 2010); 2) episodes of unexplained, recent syncope; 3) massive LV hypertrophy >30 mm; 4) nonsustained VT on ambulatory 24-hour Holter ECGs;



5) hypotensive or attenuated blood pressure response to exercise (Maron 2010). In addition, Gimeno et al recently showed that ventricular arrhythmias during exercise increased the risk of SCD as well (Gimeno et al., 2009). However, patients who survive cardiac arrest may remain asymptomatic for prolonged periods after the first cardiac event, underscoring the unpredictability of the arrhythmogenic substrate in these patients.

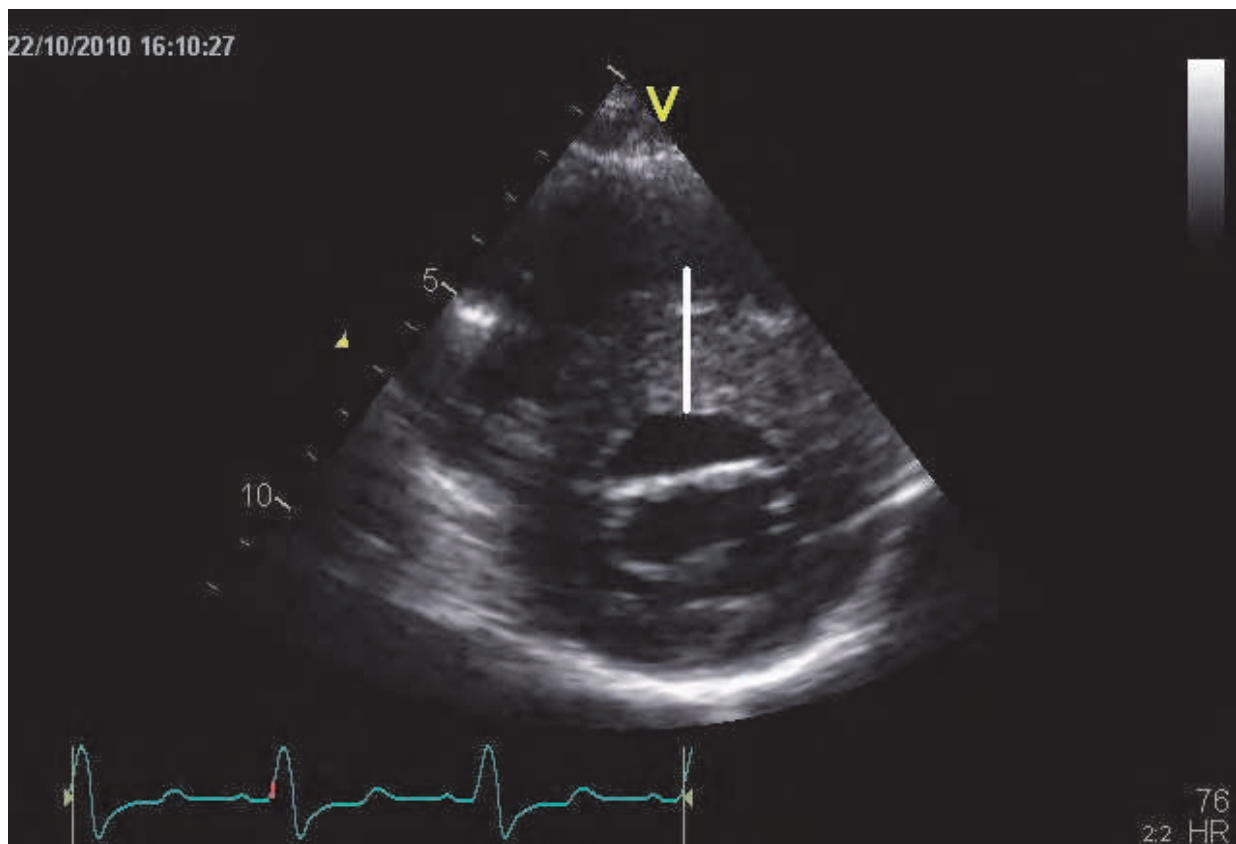


Fig. 2. Short axis of the left ventricle by echocardiography of a 14-year old boy with hypertrophic cardiomyopathy. There is a thickness of 30 mm (normally less than 11 mm).

In children, the risk of non-sudden cardiac death is as high as sudden cardiac death. Extreme left ventricular hypertrophy and a blunted blood pressure response to exercise are risk factors (Decker et al., 2009). Risk factors for SCD are extreme left ventricular hypertrophy on the electrocardiogram and a septal thickness over 190% of normal, with a sensitivity of 91% and a specificity of 78% (Ostman-Smith et al., 2005). Children may be at risk for SCD already at young age, and should therefore be screened early in hypertrophic cardiomyopathy families (Ostman-Smith et al., 2008).

When evaluating HCM patients who underwent ICD placement the number of complications is considerable (Lin et al., 2009). In the study of Lin et al 181 patients with a mean age of 44 years were investigated. During a follow-up period of 5 years, 65 patients (36%) had a total of 88 device related complications including 42 (23%) patients with inappropriate shocks. Younger age and atrial fibrillation were associated with an increased risk of inappropriate ICD discharges. It is concluded that the high incidence of complications should be taken into account when considering ICD implantation in a high-risk HCM patient (Lin et al., 2009).

#### 4. Congenital heart disease

Although most patients who are being operated for their congenital heart disease presently survive their surgery and lead a relatively normal life, a substantial portion of patients develop symptoms of heart failure or rhythm abnormalities in due time. In adult patients with ischemic or non-ischemic cardiomyopathy an LV ejection fraction below 30% is considered a clear indication for primary prevention with ICD implantation (Moss et al., 2002; Bardy et al., 2005). Risk stratification for primary prevention remains highly complex and is usually individualized, based on a variety of surgical, hemodynamic, electrocardiographic, and electrophysiologic factors (McLeod et al., 2010). However, some authors advocate that also for patients with congenital heart disease an LV ejection fraction below 30% as single risk factor is sufficient reason to implant an ICD (Silka & Bar-Cohen 2008), which is argued by others (Triedman 2008). Since the overall rate of SCD is 5 to 10 times lower than that observed in high-risk cardiomyopathy patients, the life expectancy in CHD patients is much larger. Furthermore, in CHD patients vascular access is often difficult, and the rate of device associated cardiac events and lead-failure is much higher, and they have variable cardiac anatomy. Therefore, the results of the large ICD trials cannot simply be extrapolated to the population of CHD patients. Other risk factors have to be taken into account before deciding to implant an ICD (Triedman 2008). With the increasing knowledge of rhythm abnormalities in congenital heart disease and the emergence of interventional electrophysiologic techniques some ventricular arrhythmias can be treated by catheter ablation (Walsh 2007). ICD implantation remains to be limited to otherwise untreatable rhythm abnormalities.

In a study of Yap et al more than 60% of the patients with an ICD and a congenital heart disease had a tetralogy of Fallot (Yap et al., 2007). However, the overall complication rate in the Fallot patients was higher as compared to other congenital heart defects, the number of inappropriate shocks was high (40%), while the number of appropriate shocks was low as compared to other patients (18% versus 33%). When compared with an older ICD population with dilated cardiomyopathy (DCM), the Tetralogy of Fallot patients were more likely to have experienced oversensing (45 vs. 13%;  $P < 0.02$ ), inappropriate anti-tachycardia pacing delivery (20 vs. 2%;  $P < 0.05$ ), and inappropriate cardioversion (25 vs. 4%;  $P = 0.06$ ) and less likely to receive appropriate therapies (Witte et al., 2008). On the other hand, Khairy et al studied 121 Fallot patients who received an ICD for primary (N=68) or secondary (N=53) prevention. During follow-up the number of appropriate shocks was considerable (30% of patients). This was, however, at the cost of a 5.8% yearly incidence of inappropriate shocks, and the occurrence of complications in 36 patients, mostly (70%) lead-related (Khairy et al., 2008). In Fallot patients with unstable ventricular tachycardia, their rhythm abnormalities may in some patients be treated by catheter ablation (Kriebel et al., 2007).

#### 5. ICD programming

In children with ICDs, the risk of inappropriate shock therapy is significantly higher as compared to adult studies. Pediatric ICD series with mean intervals of follow-up between 29 and 51 months have reported 20% to 50% inappropriate shock therapy. These shocks may have a negative impact on the quality of life, and can induce secondary arrhythmias.

	Tomaske 2008	Stephenson 2006	Kriebel 2006	Cannon 2006
Number	15	22	8	8
Age (yr)	12.5	8.9	0.3-8	1-29
Weight (kg)	36.5	25.5	4-29	
Follow-up (mo)	22	29	14.5	22
Appr Shocks	4	7	2	2
Inappr Shocks	3	4	0	3
Revision	4	7	2	3

Table 1. Outcomes of extracardiac ICD implantations in young patients and patients with congenital heart disease. Results are comparable to transvenous approaches.

Inappropriate shocks in children are mostly caused by sinus tachycardia or supraventricular tachycardia. High sinus rates up to 200 per minute are not uncommon in children and patients with congenital heart disease have a high incidence of atrial arrhythmias, mostly intraatrial reentrant tachycardia. Implantation of a dual chamber ICD system can potentially reduce the incidence of inappropriate discharges attributable to the misclassification of sinus tachycardia or supraventricular tachycardia as a ventricular event. In a recent multicenter study the difference of appropriate and inappropriate shocks in a single chamber or dual chamber device in patients <30 years old was investigated (Lawrence et al., 2009). The authors found no differences between single chamber or dual chamber systems regarding the occurrence of appropriate or inappropriate shocks, irrespective of underlying cardiovascular disorder or type of ICD system. (Lawrence et al., 2009). T-wave sensing is another important cause of inappropriate shock therapy especially in the group of patients with long QT syndrome. Exercise tests are required to obtain maximal heart rate and to evaluate T wave sensing during high heart rates. QRS discrimination or atrial discrimination algorithms can be helpful tools to prevent some of these problems.  $\beta$ -blockade can also be a practical therapy to avoid inappropriate ICD shock for supraventricular tachycardias or sinus tachycardia. However, the most important measure to reduce inappropriate shocks is careful ICD programming of the individual child with regard to detection rate and time immediately after ICD implantation. In a recent article several programming tips have been given (Khairy & Mansour 2011). Since sinus tachycardia and supraventricular tachycardia may have frequencies as high as 200 bpm, most young patients have a ventricular fibrillation zone programmed up to 220 bpm. Concerning detection times there is a compromise between overtreating otherwise self-terminating events and delaying therapy for potentially unstable arrhythmias. Detection times as long as 18 of 24 intervals or even 30 of 40 intervals have been advocated with no increase in adverse events. Antitachycardia pacing (ATP) has been shown to be effective in the majority of congenital heart disease patients, thereby lowering the number of shocks.

Although routine testing of the defibrillation threshold (DFT) is not necessary during follow-up of pediatric ICD patients, any clinical change or problem should be evaluated immediately and DFT testing should also be considered in this situation. (Theuns & Gold, 2010). In young children with subcutaneous ICD systems regular DFT testing should be considered during growth.

## 6. Complications

There seem to be differences in the frequency of ICD-related complications between children and adults. However, due to the small number of patients in pediatric series it is difficult to estimate the true incidence of ICD related complications in children. (Shah et al., 2009). Implantation procedure-related complications include pocket infection, pocket hematoma, microdislodgement requiring lead manipulation, hemothorax, superior vena cava syndrome, pneumonia, electromechanical dissociation requiring cardiopulmonary resuscitation and second degree burns from repeated external rescue shocks (Shah et al., 2009). Especially the number of infections seem to be higher in children as compared to adults (Link et al., 1999). The most common ICD system related adverse effect is the high incidence of inappropriate shocks and a high incidence of lead failures in children. Inappropriate shocks are usually related to the occurrence of sinus tachycardia, supraventricular tachycardia, T wave oversensing or QRS complex double sensing. Lead failures are strongly related to the size of the patient. The youngest and smallest patients have the highest risk of lead complications. Furthermore, higher levels of exercise and activity in the pediatric population may increase the amount of lead problems.

## 7. Mode of ICD implantation

In children or young patients with congenital heart defects it is often not possible to make use of routine transvenous ICD systems. Underlying causes are the size of the patient or structural heart diseases with residual intracardiac shunts and abnormal systemic venous pathways. Various modifications have been used to implant ICDs in patients with limited venous access (Cannon et al., 2006; Bove et al., 2010; Greene et al., 2004; Kriebel et al., 2006; Stephenson et al., 2006; Tomaske et al., 2008). These include subcutaneous patches, epicardial and pleural lead positioning, use of the subxiphoid incision, or direct transatrial approach. It is important that the technique used is individualized in the diverse population of children and young patients with congenital heart disease in need for ICD placement (Stephenson et al., 2006). Recently, Radbill et al compared the use of transvenous ICDs with nontransvenous systems in a group of pediatric and congenital heart disease patients (Radbill et al., 2010). Although no differences were reported considering appropriate or inappropriate shocks there was a significantly higher amount of ICD system failure in the nontransvenous group. System survival at 12, 24, and 36 months was 73%, 55% and 49% in the nontransvenous group compared to 91%, 83% and 76% in the transvenous group. Causes of system failure in the nontransvenous group included pace-sense lead failure, shock coil failure, generator migration, and loosened set-screw connection.

Recently, the search for avoidance of transvenous lead placement has resulted in the development of an entirely subcutaneous ICD system (Bardy et al., 2010). The system consists of a parasternal electrode and a left lateral thoracic pulse generator. In a trial of 55 adult patients it was shown as effective in terminating ventricular tachyarrhythmias with little complications during a 10-month follow-up period. This subcutaneous ICD system can also be used in older children and has been reported in two children of 10 and 12 years old (34 and 35 kg body weight) (McLeod and McLean 2010). In both children this device was implanted without complications, and no adverse events occurred during an 8- and 5-month follow-up period (McLeod & McLean 2010).



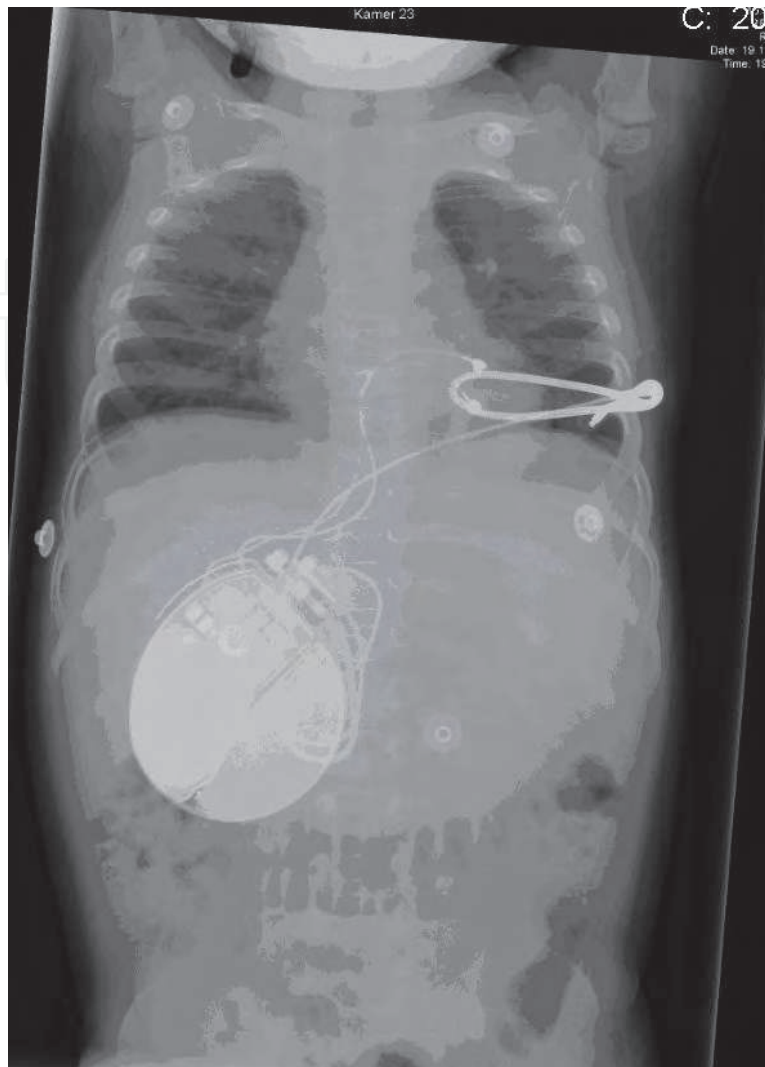


Fig. 3. Chest X-ray of a 6-month old child with an ICD. The active can is placed abdominally, and the pacing wires are placed epicardially. A subcutaneous patch is in use for eventual shocks.

Another approach is the placement of the ICD in the axilla. The usual infraclavicular placement of a transvenous ICD can cause a quite visible scar with a suboptimal cosmetic result. Furthermore, straps can rub and cause irritation at this site (Collins et al., 2009; Rausch et al., 2010). In a large retrospective review it was shown that there were no differences between the axillary and infraclavicular technique in implant characteristics, lead longevity, implant complications, lead fractures or dislodgements, inappropriate ICD discharges, or device infections (Rausch et al., 2010).

## 8. Home monitoring

Since 2000 a remote control system was introduced for the follow-up of ICD patients. Through this system data from the implanted device are transmitted to a website once a day as well as immediately following an arrhythmia. By this method changes in ICD function, lead problems, and asymptomatic arrhythmias can be detected earlier without the patient actually visiting the hospital. It has been shown that the events as send by the home

monitoring system show excellent comparability to the data as obtained by standard clinical evaluation (Perings et al., 2011). The home monitoring system can detect several, otherwise asymptomatic episodes enhancing prompt detection of for instance lead problems and facilitates management decisions (Varma et al., 2010). Most patients show a high level of acceptance and satisfaction of this new system after a mean follow-up period of 1 year, with only some refusing the home monitoring (Ricci et al., 2010). Until now, however, this system has mostly been used in the adult population, and data about the appropriateness in pediatrics are as yet lacking.

## 9. Quality of life

With the increasing number of ICDs in young patients the psychological effects has been studied as early as 1996 (Dubin et al., 1996). Nearly 75% of the patients felt their health was good to excellent with 40% reporting improvement since ICD implantation. All felt capable of performing daily activities (Dubin et al., 1996).

The implantation of an ICD has a great impact on the quality of life of these patients. These psychological effects have so far been investigated by many authors (Herrmann et al., 1997). The poor psychosocial outcome in these patients may be related to the underlying cardiovascular condition (Burke et al., 2003), or it may be the effect of surviving an out of hospital cardiac arrest, not related to the treatment received (Kamphuis et al., 2002). It has been found that the lasting psychological distress will not dissipate spontaneously or naturally and that psychosocial intervention may be warranted (Kamphuis et al., 2003). The most common psychological problems are depression and anxiety. Depressive symptoms persist over time, and are associated with a higher incidence of shock therapy (Suzuki et al., 2010). However, concerns about the ICD has been found a predictor of psychological morbidity independently of the number of shocks (Pedersen et al., 2005). On the other hand, Luyster et al found no relation between number of shocks and anxiety (Luyster et al., 2006). They found especially a higher level of perceived resource loss to be associated with higher levels of both anxiety and depression (Luyster et al., 2006).

The growing pediatric ICD population stresses the need to evaluate the quality of life in this group of ICD recipients. Adult patients with ICDs show significantly worse psychological and physical functioning. Behavioral changes have been reported such as reduced activity, avoidance, depression, and anxiety, especially in those who had received ICD shocks. These findings in adults could not be confirmed in a recent study in a group of pediatric ICD recipients. In this study, cardiac illness severity and ICD shock therapy were not significantly associated with anxiety, depression, quality of life, or family functioning.

Recently, we have also evaluated health-related quality of life study in 30 Dutch pediatric ICD recipients using different quality of life questionnaires. This study showed that pediatric ICD recipients showed more problems in the domains of motor functioning, sleep, work/school, negative emotions, and anxiety. Furthermore, worry and anxiety were significantly associated with the number of ICD shocks (unpublished data). These findings indicate that avoidance or reduction of inappropriate shocks is one of the most important steps to improve quality of life. Furthermore, it emphasizes the need of proper guidance and, if necessary, implementation of psychological interventions. Screening for cardiovascular diseases may have its own impact, Children who were genetically tested carriers showed to have a reduced psychological well-being (Smets et al., 2008).

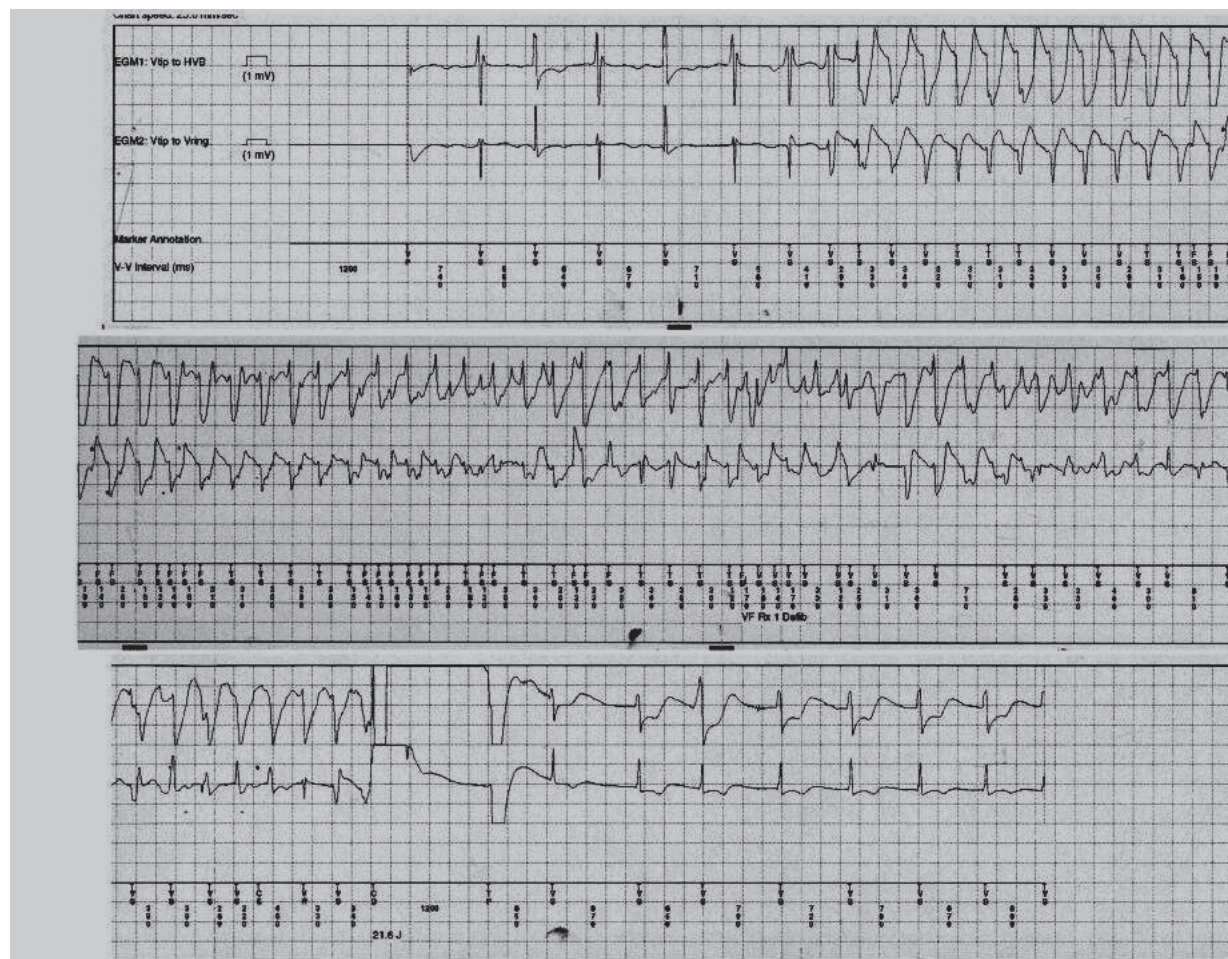


Fig. 4. An ICD strip of an appropriate shock is shown. Fast ventricular tachycardia is followed by a 21-J shock which terminated the tachycardia. This is followed by an escape rhythm with broad QRS complexes.

## 10. Conclusions

ICD implantations are increasingly performed in children and young adults with a variety of underlying cardiac disorders. Although the effectiveness of ICD therapy in this specific population is good, there are concerns about the high number of inappropriate shocks and lead problems. New developments include the complete subcutaneous device and home monitoring systems. Although the overall quality of life usually is good, several patients need psychological support, and this support has to be included in regular follow-up programs.

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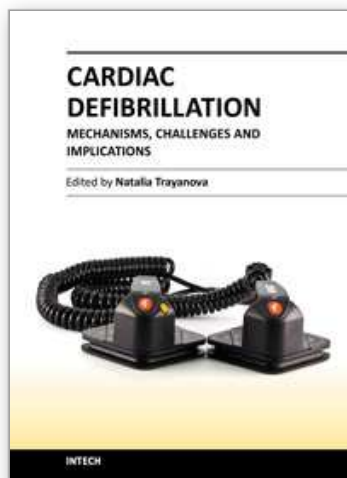
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The only known effective therapy for lethal disturbances in cardiac rhythm is deï-brillation, the delivery of a strong electric shock to the heart. This technique constitutes the most important means for prevention of sudden cardiac death. The efficacy 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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