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**CASE REPORT**
*Arrhythmias/electrophysiology*

# Case report of an S-ICD implantation for secondary prevention in a patient with complex congenital heart disease, dextrocardia, and situs solitus

**Felix Wiedmann [1,2,3], Raffaele De Simone[4], Peter Rose[4], Matthias Karck[4], Matthias Gorenflo[5], Norbert Frey[1,2,3], and Constanze Schmidt [1,2,3]***

[1]Department of Cardiology, University Hospital Heidelberg, Heidelberg, Germany; [2]DZHK (German Centre for Cardiovascular Research), Partner Site Heidelberg/Mannheim, University of Heidelberg, Heidelberg, Germany; [3]HCR, Heidelberg Centre for Heart Rhythm Disorders, University Hospital Heidelberg, Heidelberg, Germany; [4]Department of Cardiac Surgery, University Hospital Heidelberg, Heidelberg, Germany; and [5]Department of Pediatric Cardiology and Congenital Heart Diseases, University Hospital Heidelberg, Heidelberg, Germany

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**Background** Dextrocardia is a congenital anomaly in which the apex of the heart is abnormally located on the right side of the chest. Situs solitus describes viscera that are in the normal position, with the stomach on the left side. In these patients, implantation of transvenous implantable cardioverter-defibrillator (ICD) can be limited by anatomical abnormalities commonly associated with this condition.

**Case summary** We present the case of a young female patient with absent right atrioventricular connection, morphologically left systemic ventricle, muscular restrictive ventricular septal defect, and dextrocardia with situs solitus who was indicated for secondary prophylactic ICD implantation after resuscitation for polymorphic ventricular tachycardia. Due to a bilateral bidirectional Glenn anastomosis, transvenous access via the vena cava superior to the right ventricle could not be achieved. For this reason, we successfully implanted a subcutaneous ICD (S-ICD) with an individually optimized right parasternal electrode position. Potential complications of epimyocardial implantation via re-thoracotomy could thus be circumvented.

**Discussion** In patients with complex congenital heart disease, the S-ICD is an effective method of preventing sudden cardiac death. Our case report demonstrates the feasibility of left S-ICD implantation even in the presence of dextrocardia with situs solitus.

**Keywords** Case Report • Complex congenital heart disease • Dextrocardia • ACHD • S-ICD • Situs solitus

**ESC Curriculum** 5.10 Implantable cardioverter defibrillators • 5.6 Ventricular arrhythmia • 9.7 Adult congenital heart disease

## Learning points

- In patients with complex congenital heart disease lacking the option of transvenous access, the subcutaneous ICD (S-ICD) is an effective method of preventing sudden cardiac death.
- Left-sided S-ICD implantation may be possible even in the presence of dextrocardia with situs solitus.

* Corresponding author. Tel. +49 6221 56 8187, Fax. +49 6221 56 5724, Email: Constanze.Schmidt@med.uni-heidelberg.de
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## Introduction

Dextrocardia with situs solitus can be observed in approximately 7,500–29,000 people worldwide.[1] Dextrocardia is a congenital anomaly in which the heart is abnormally located on the right side of the chest. Situs solitus describes viscera that are in the normal position, with the stomach on the left side. Similar to the general population, these patients are likely to benefit from an implantable cardioverter-defibrillator (ICD) for primary or secondary prevention of sudden cardiac death due to ventricular arrhythmias.[2]

In contrast to dextrocardia with situs inversus, dextrocardia with a normal abdominal situs is associated with a high incidence of associated congenital cardiac anomalies.[3] Therefore, placement of transvenous leads may not be anatomically feasible and can further be associated with an increased risk of arterial thromboembolism due to intracardiac shunt lesions.[4]

We present the case of a young female patient who required an ICD for secondary prophylaxis of sudden cardiac death after resuscitation for polymorphic ventricular tachycardia (VT). Because of the complex anatomic situation and extensive previous surgery, both transvenous and epimyocardial electrode placements were considered rather unfavourable. While subcutaneous ICDs (S-ICDs) are generally considered a good option to overcome the limitations of transvenous ICDs (TV-ICDs) delivery systems in complex anatomic conditions, published data on implantation of S-ICDs in dextrocardia are limited to a few case reports.[5,6]

## Timeline

| | |
|---|---|
| September 1982 | Birth |
| September 1982 | First heart surgery: pulmonary artery banding |
| June 1993 | Second heart surgery Damus–Kaye–Stansel anastomosis (pulmonary artery to aorta) and bidirectional bilateral Glenn anastomosis (vena cava to pulmonary artery) |
| February 2012 | Stent implantation in a pulmonary arterial bifurcation stenosis |
| February 2012 | Coil occlusion of two venous collaterals from the area of the left superior vena cava and V. mammaria |
| August 2012 | Occlusion of the right mammary artery due to aortopulmonary collaterals using an Amplatzer Vascular Plug 4 |
| June 2016 | Coil closure of a sequestral artery to the right lung |
| December 2021 | Resuscitation due to polymorphic VT |
| January 2022 | S-ICD implantation for secondary prevention |

## Case summary

We report the case of a 39-year-old female patient with complex congenital heart disease who was scheduled for secondary prophylactic ICD implantation after resuscitation for documented polymorphic VT over 2 min. Due to absent right atrioventricular (AV) connection, morphologically left systemic ventricle, muscular restrictive ventricular septal defect, and dextrocardia with situs solitus, this patient received Damus–Kaye–Stansel anastomosis of the pulmonary artery with the aorta and bidirectional bilateral Glenn anastomosis at the age of 11 (see timeline). At the age of 30 years, a stent was implanted for pulmonary arterial bifurcation stenosis, and the patient received a coil closure of two venous collaterals from the area of the left superior vena cava and the mammary vein. Further, occlusion of the right mammary artery with aortopulmonary collaterals to the right lung was performed with an Amplatzer device (*Figure 1A*). Her chronic medication is listed in *Table 1*. After successful extrahospital resuscitation for polymorphic VT, the patient was intubated, mechanically ventilated, and externally cooled to 36 °C for 24 h. The cardiovascular examination did not show any other pathological findings. Laboratory tests revealed no secondary causes for the occurrence of the arrhythmia. Echocardiography, limited by a significantly restricted acoustic window, revealed a functionally singular ventricle with left ventricular morphology and preserved global function without the possibility of exact quantification. A 12-lead electrocardiogram (ECG) demonstrated sinus rhythm with inversion of all complexes (global negativity) in lead I and aVL, a right axis deviation, as well as an abnormal precordial progression, consistent with dextrocardia (*Figure 2A*); The QRS width was 132 ms. Paroxysmal atrial fibrillation was documented in the medical history. Beyond secondary prophylactic ICD indication, the patient had no indication for atrial or ventricular pacing or cardiac resynchronization therapy. Transvenous access to the right ventricle through the right atrium was not possible due to the bidirectional bilateral Glenn anastomosis (*Figure 1B*). Because of the absent right AV connection, a transfemoral approach was avoided, too. Furthermore, due to the increased risk of thromboembolism in the presence of severe intracardiac shunt lesions, it was decided to abstain from using transvenous leads as far as possible.

The surgeon's opinion was that epimyocardial electrode placement is associated with a significantly increased perioperative risk because of the extensive prior surgery. Based on this, the option of an S-ICD implantation with a customized electrode position was discussed with the patient. A standard left-side generator position was preferred due to the preference of the operator and the right-handed patient. Surface electrogram-based eligibility screening was performed using the automated screening tool. The electrodes were placed in an anatomically adapted right parasternal position (2 cm parasternal). The screening was performed in supine sitting and standing positions (*Figure 2B*). For this individually optimized electrode position, the primary (lead III) and secondary (lead II) vectors demonstrated feasibility in all positions, whereas the alternative vector (lead I) was functional only in the supine position (*Figure 2B*). After a thorough explanation and sufficient time for reflection, the patient gave her consent to the implantation of an S-ICD system. The implantation of a subcutaneous defibrillator (Boston Scientific Emblem MRI S-ICD A209) was successfully performed in analgosedation with a 20° elevation of the upper body to optimize venous reflow. The intermuscular device pocket was located in the natural

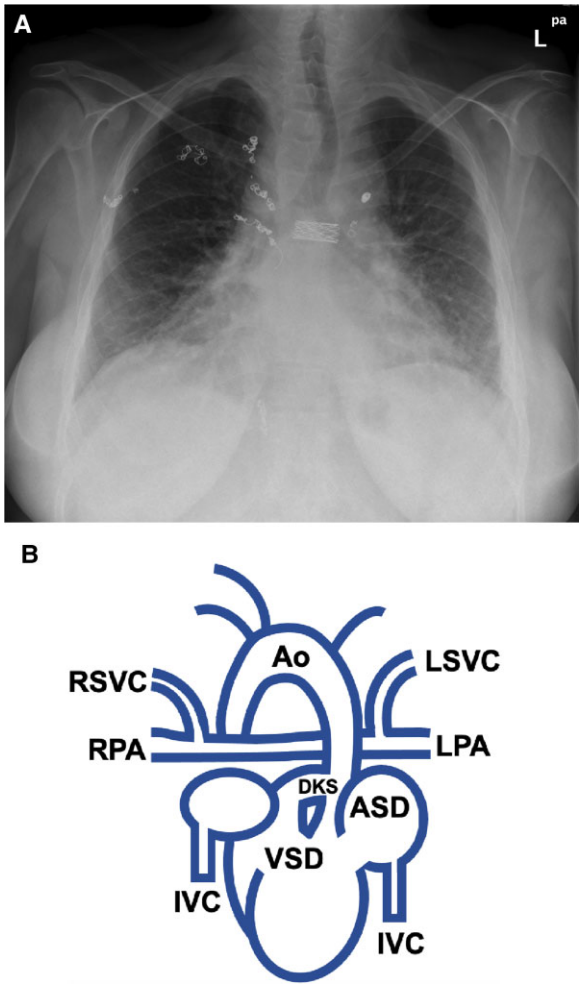


**Figure 1** (*A*) Chest X-ray posteroanterior (pa) view showing dextrocardia with situs solitus. (*B*) Hypothetical schematic of the patient's cardiovascular anatomy. Ao, aorta, ASD, atrial septal defect; DKS, Damus–Kaye–Stansel anastomosis; IVC, inferior vena cava; LPA, left pulmonary artery; LSVC, left superior vena cava; LV, morphologically left ventricle; RPA, right pulmonary artery; RSVC, right superior vena cava; and VSD, ventricular septal defect.

space between the left latissimus dorsi and the serratus anterior muscle (*Figure 3*). The defibrillation electrode was positioned 2 cm lateral and parallel to the right sternal border. As predicted by the eligibility screening, the primary and secondary vectors showed adequate signal quality. Upon completion of the device implantation, conversion testing was performed after induction of ventricular fibrillation (VF) with a sensing configuration of the secondary vector (Lead II) (*Figure 2C*). VF was successfully terminated by one single 65-J shock, and the shock impedance yielded 72 Ω. The patient has not experienced any device-related complications nor received any device therapy after one month of follow-up. The S-ICD is well tolerated, and sensing vectors show no relevant changes when compared to the time of implantation; the SMART Pass filter function was activated on the day of implantation.

**Table 1 Chronic medication**

| | |
|---|---|
| Apixaban | 5 mg b.i.d. |
| Bisoprolol | 5 mg b.i.d. |
| Digitoxin | 0.07 mg q.d. |
| Levothyroxine | 75 µg q.d. |
| Pantoprazole | 40 mg q.d. |
| Ferrosanole | 100 mg q.d. |
| Folic acid | 0.4 mg q.d. |

b.i.d., twice a day; q.d., once a day.

## Discussion

Since approval by European and North American regulatory agencies in 2009 and 2012, the clinical use of S-ICD devices has expanded significantly.[7] The S-ICD was designed with the goal of reducing lead-related complications, commonly encountered in transvenous systems.[7] Advantages of S-ICD over TV-ICD have been well demonstrated and include the absence of risk of pneumothorax, vascular injury or cardiac perforation, less radiation exposure during implantation, lower incidence of systemic infections, lower risk of lead-related complications such as lead fracture, lower morbidity associated with lead extraction, and cosmetic advantages, especially in female patients.[7,8] Unlike TV-ICDs, an S-ICD does not allow for antibradycardia pacing, cardiac resynchronization therapy, or antitachycardia pacing.

S-ICDs have proven successful in terminating VT or VF with success rates at first shock similar to TV-ICD.[7] The patient case described illustrates very well that an S-ICD is a suitable option to overcome the limitations of TV-ICD systems in complex anatomical conditions. The use of an S-ICD has already been demonstrated in adult patients with dextrocardia and dextrocardia with Fallot-tetralogy[5] as well as dextrocardia and situs inversus.[6] Our case also demonstrates the technical feasibility of left-sided S-ICD implantation in patients with dextrocardia and situs solitus.

Since the incidence of screening failure in adults with congenital heart defects (17–21%) is significantly higher than in the general adult population (7–10%), it is essential to perform surface electrogram-based eligibility screening.[9,10] There are case reports in which electrocardiographic eligibility screening indicated the feasibility of right-sided S-ICD system implantation after conventional screening from the left side failed.[11] Overall, however, there is little data for a right-sided aggregate position in S-ICD patients, which ultimately led us to choose a left-sided position.

The guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death of the European Society of Cardiology (ESC) as well as the Association for European Paediatric and Congenital Cardiology (AEPC) recommend the S-ICD as a reasonable option for TV-ICD in patients who meet the indication for an ICD for primary or secondary prevention of sudden cardiac death.[2,12]

The present case clearly shows that an S-ICD should be considered the device of choice in patients with complex congenital heart

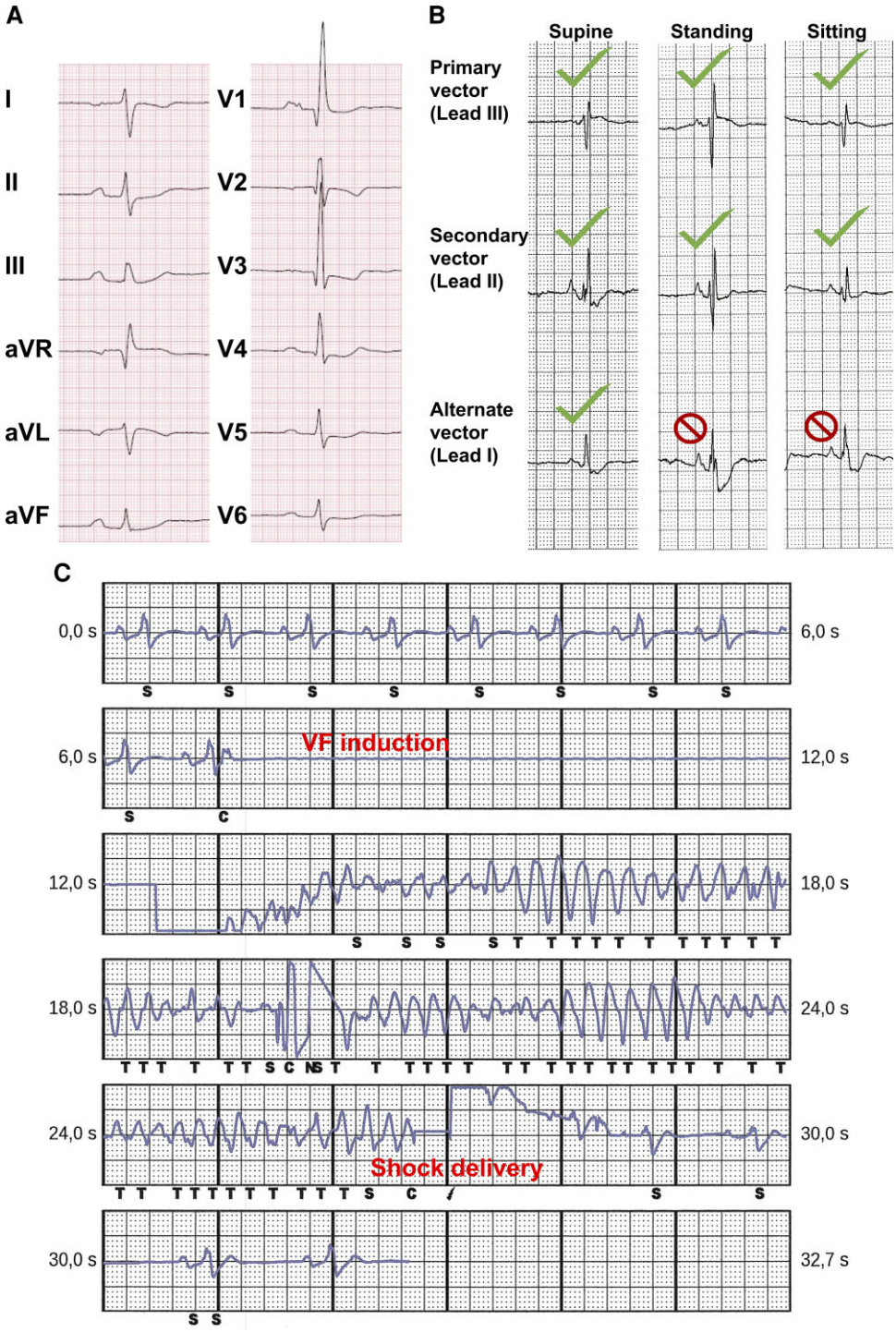


**Figure 2** (*A*) Surface ECG of the patient. (*B*) Electrocardiographic eligibility screening in supine, standing, and sitting positions as indicated. The test results are visualized as inserts. (*C*) Documentation of the perioperative conversion test. Secondary vector with two-fold amplification. After induction of ventricular fibrillation (VF) via delivery of a 50 Hz burst, the arrhythmia is adequately detected and effectively terminated by a 65-J shock. The time to therapy is approximately 14 s. S, sensing of a regular QRS complex, T, tachy-sensing, N, detection of noise, and C, start of capacitor charging.

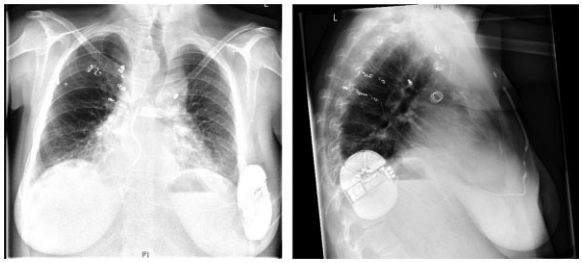

**Figure 3** Chest X-ray posteroanterior and lateral view after S-ICD implantation with an individually optimized right parasternal electrode position.

disease. However, studies with larger patient populations and long-term follow-up periods are needed to demonstrate long-time efficacy and safety of S-ICD systems in patients with congenital heart disease.

## Conclusions

In patients with complex congenital heart defects disease, lacking the option of transvenous access, the S-ICD is an effective method of preventing sudden cardiac death. Our case report demonstrates the feasibility of left S-ICD implantation even in the presence of dextrocardia with situs solitus.

## Lead author biography

Dr Felix Wiedmann was born in Germany in 1987. He received his M.D. degree from Heidelberg University in 2014 where he is currently employed as a cardiologist and basic scientist at the Department of Cardiology, University Hospital Heidelberg. His doctoral thesis, focusing on the 'Cardiac roll of two pore domain potassium channels' was awarded with the Kaltenbach scholarship (German Heart Foundation) and the Otto-Hess scholarship (German Cardiac Society). His research interests cover heart rhythm disorders and their underlying mechanisms. Since 2022 he is receiving a scholarship from the Clinician-Scientist-Program of the German Cardiac Society.

## Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

**Slide sets:** A fully edited slide set detailing these cases and suitable for local presentation is available online as Supplementary data.

**Consent:** The authors confirm that written consent for submission of the case report including images and associated text has been obtained from the patient in line with COPE guidance.

**Conflict of interest:** None declared.

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