The subcutaneous ICD (S-ICD) in congenital heart disease – what lessons have we learnt so far?

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Introduction:
The S-ICD offers a considerable alternative to conventional systems in patients with congenital heart disease (CHD), as it avoids vascular access and thoracotomy.

Methods:
S-ICDs were implanted in a heterogeneous group of 15 patients with structural CHD (minimum age 8.9 y, height 131.0 cm, weight 28.0 kg). The majority presented with challenging hemodynamic and anatomical features, such as missing anatomical vascular access (e.g. univentricular anatomy and Glenn anastomosis), tricuspid valve dysplasia, existing ventricular assist device (VAD), or chronic left-to-right shunt. In 4 patients S-ICD was combined with epicardial pacing. Lead and device positioning had to be modified according to anatomic variations such as dextrocardia. 14 devices were implanted intramuscular or submuscular.

Results:
Follow-up was up to 7 years. Cosmetic results and clinical tolerance was good even in subjects below 30 kg. 7 patients received appropriate shocks. 4 patients with RBBB presented with t-wave-oversensing (TWOS) due to altered t-wave morphology under higher heart rate. 2 patients consequently received inappropriate shocks in sinus tachycardia under exercise. The other 2 patients received “pseudo-appropriate” shocks (treated episodes that were misinterpreted by device resulting from TWOS, though patient had clinical benefit from shock); one with hemodynamically relevant SVT and the other with slow VT. Subsequently, TWOS could be successfully addressed by modified programming and firmware updating. Combined epicardial pacing was feasible and safe although potential interactions may occur, as experienced in one patient with false (unipolar) programming and unreasonable template acquisition. One patient with appropriate shocks due to monomorphic VTs developed subsequently slow VTs below the programmable shock zone (170 bpm) and drug-induced chronotropic incompetence. Another patient with subcutaneous implantation and progressive obesity suffered from late device migration. The two latter patients had to be switched to transvenous systems. The VAD patient died of unknown reason.

Conclusion:
S-ICD therapy is feasible even in patients with complex CHD without transvenous access or systemic intracardiac shunting. Thorough pre-implantation screening combined with modifications in lead or device placement and adapted programming strategies are indispensable to allow proper function even in severely distorted CHD anatomy. Post-implantation exercise testing is mandatory to address TWOS, especially in RBBB. Cosmetic and functional results are excellent if protective implantation techniques are applied; nevertheless, the bulky size of the current device generation precludes its use below a bodyweight of approximately 25-30 kg. Despite successful application with combined (epicardial) pacing, the lack of antibradycardia and antitachycardia pacing represents the other major limitation towards use in CHD.