Extravascular defibrillator implant in a patient with Poland syndrome

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Introduction

We present a case of a young man with Poland syndrome who underwent extravascular implantable cardioverter-defibrillator (EV-ICD) implantation.

Poland syndrome is a rare congenital condition characterized by unilateral absence or underdevelopment of chest wall muscles that may be accompanied by upper limb abnormalities, which often include absence of 1 pectoralis major muscle.[1,2] With a traditional transvenous defibrillator, the lead is secured to the chest wall by a nonabsorbable suture attaching the suture sleeve to muscle, including the fascia.[3] The anatomic abnormalities associated with Poland syndrome lead to challenges for implantation of cardiac devices with transvenous leads and can require alternative approaches. Previously reported techniques include anchoring the device to the periosteum of the first rib[4] and tunneling to position the device in the pre-fascial plane of the abdominal cavity.[5]

Case report

We present a case of a 34-year-old man with a clinical diagnosis of Poland syndrome with an obvious chest wall deformity (Figure 1) and complete absence of his left pectoralis major muscle (Figure 2) who was referred for a primary prevention ICD. He had recently been diagnosed with a dilated cardiomyopathy. There was a strong family history of dilated cardiomyopathy associated with premature sudden cardiac death.

With his abnormal anatomy, we anticipated that a traditional left-sided intravascular ICD implant would be difficult. A right-sided implant was considered; however, compared with a standard left-sided implant, there is a significantly different shock vector, and there may be a higher defibrillation threshold.[6] The EV-ICD allows for both antitachycardia pacing and defibrillation to be performed via a substernal lead that is secured to abdominal muscles and a pulse generator placed in the left midaxillary line.[7] After considerable thought, we elected to implant an EV-ICD in this patient (Figure 3).

Fluoroscopy and external landmarks were used to place the lead in a substernal location via a subxiphoid location. The pulse generator was placed in a left lateral location (Figure 3). Dissection in this area was more difficult than would be typically expected, but we were unsure whether this was related to the patient's underlying Poland syndrome. At the end of the case, defibrillator threshold testing was performed, and ventricular fibrillation was induced and terminated successfully with a single 30-J high-voltage shock.

KEY TEACHING POINTS

- Poland syndrome is a rare congenital syndrome that provides significant challenges for traditional transvenous defibrillator implantation.
- The EV-ICD is novel defibrillator that provides pacing as well as defibrillation therapy.
- The EV-ICD may be feasible in patients who present with anatomy that may make transvenous lead placement difficult.

Discussion

The EV-ICD is a novel defibrillator that is now commercially available and in clinical use. It may provide up to 40 J energy for defibrillation therapy, and in addition to this can provide pacing therapy either through antitachycardia pacing to treat monomorphic ventricular tachycardia, hemodynamic support after a shock, or short-term pause prevention pacing.[8] In this case, the patient's anatomic abnormalities would have created challenges for transvenous lead insertion, meaning that an extravascular system was preferable.

Conclusion

The EV-ICD is feasible and should be considered in patients with Poland syndrome.

KEYWORDS Extravascular defibrillator; EV-ICD; Poland syndrome; Implantable cardiac defibrillator; ICD; Sudden cardiac death

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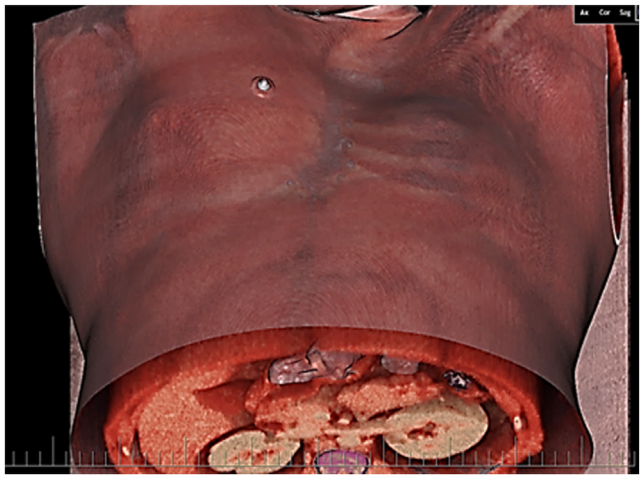

Figure 1 Three-dimensional reconstruction from computed tomography scan demonstrating left chest wall deformity.

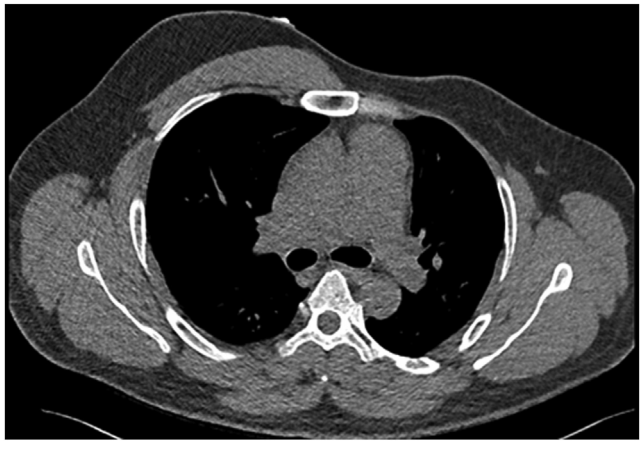

Figure 2 Axial slice from computed tomography scan demonstrating absence of left pectoralis major.

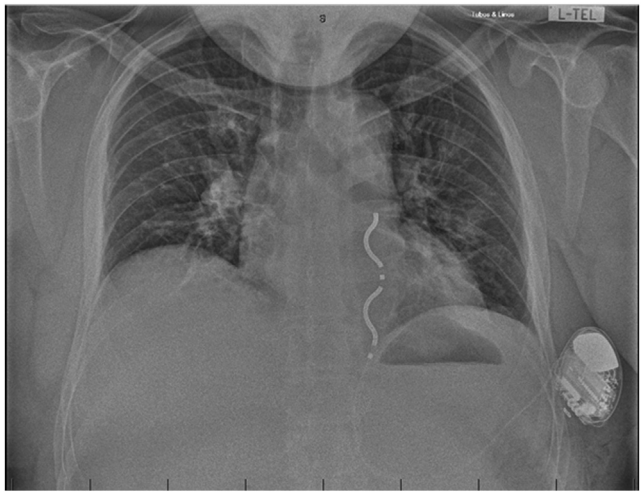

Figure 3 Postprocedural chest radiograph showing the substernal defibrillator lead and pulse generator in a left lateral location.

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References

1. Kennedy KR, Wang AL. Poland syndrome. N Engl J Med 2018;378(1):72. https://doi.org/10.1056/nejmicm1709713.
2. Baldelli I, Baccarani A, Barone C, et al. Consensus based recommendations for diagnosis and medical management of Poland syndrome (sequence). Orphanet J Rare Dis 2020;15(1):201. https://doi.org/10.1186/s13023-020-01481-x.
3. Burri H, Starck C, Auricchio A, et al. EHRA expert consensus statement and practical guide on optimal implantation technique for conventional pacemakers and implantable cardioverter-defibrillators: endorsed by the Heart Rhythm Society (HRS), the Asia Pacific Heart Rhythm Society (APHRS), and the Latin-American Heart Rhythm Society (LAHRS). Europace 2021;23(7):983–1008. https://doi.org/10.1093/europace/euaa367.
4. Brown Kristen, Johal Gurpreet, Cavalieri Stuart. Poland syndrome: a novel approach to pacemaker placement. Chest 2019;156(4 Suppl 2019):A673. https://doi.org/10.1016/j.chest.2019.08.653.
5. Jones SO, Wiggins NB. An alternative pacemaker site in patient with Poland syndrome. EP Lab Digest 2019;19(8):38–39.
6. Pope MT, Paisey JR, Roberts PR. Defibrillation threshold testing for right-sided device implants: a review to inform shared decision-making, in association with the British Heart Rhythm Society. Arrhythm Electrophysiol Rev 2023;12:e10. https://doi.org/10.15420/aer.2022.38.
7. Friedman P, Murgatroyd F, Boersma LVA, et al. Extravascular ICD pivotal study investigators. efficacy and safety of an extravascular implantable cardioverter-defibrillator. N Engl J Med 2022;387(14):1292–1302. https://doi.org/10.1056/NEJMoa2206485.
8. Thompson AE, Atwater B, Boersma L, et al. The development of the extravascular defibrillator with substernal lead placement: a new frontier for device-based treatment of sudden cardiac arrest. J Cardiovasc Electrophysiol 2022;33(6):1085–1095. https://doi.org/10.1111/jce.15511.