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CASE REPORT

Poland syndrome: from the perspective of invasive electrophysiology in a pediatric patient – A case report

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Abstract

Introduction: Poland syndrome is a rare congenital condition characterized by the unilateral absence or hypoplasia of the pectoralis major muscle, often with ipsilateral upper limb malformations. Cardiac displacement may complicate invasive procedures. **Case report:** A 15-year-old female with Poland syndrome and recurrent palpitations presented with documented atrioventricular nodal reentrant tachycardia (AVNRT). During the electrophysiological study, mesocardia altered expected catheter positions. Ablation was successfully performed under fluoroscopic guidance. **Discussion:** This case highlights the need to recognize anatomical variants in Poland syndrome when planning cardiac interventions. Successful ablation is feasible with conventional techniques when adapted to individual anatomy, even without three-dimensional mapping systems.

Keywords: Poland syndrome. Congenital cardiac displacement. Mesocardia. Pediatric arrhythmia. Radiofrequency catheter ablation. Case report.

Síndrome de Poland: perspectiva da eletrofisiologia invasiva em paciente pediátrico – Relato de caso

Resumo

Introdução: A Síndrome de Poland é uma condição congênita rara caracterizada pela ausência ou hipoplasia unilateral do músculo peitoral maior, frequentemente acompanhada de malformações do membro superior ipsilateral. O deslocamento cardíaco pode complicar procedimentos invasivos. **Relato de caso:** Uma paciente de 15 anos com Síndrome de Poland e palpitações recorrentes apresentou taquicardia por reentrada nodal atrioventricular (TRNAV) documentada. Durante o estudo eletrofisiológico, a mesocardia alterou as posições esperadas dos cateteres. A ablação foi realizada com sucesso sob orientação fluoroscópica. **Discussão:** Este caso destaca a necessidade de reconhecer variantes anatômicas na Síndrome de Poland ao planejar intervenções cardíacas. A ablação bem-sucedida é viável com técnicas convencionais adaptadas à anatomia individual, mesmo sem sistemas de mapeamento tridimensional.

Palavras-chave: Síndrome de Poland. Deslocamento cardíaco congênito. Mesocardia. Arritmia pediátrica. Ablação por cateter com radiofrequência. Relato de Caso.

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Key points

What is known about this topic

- Poland syndrome includes unilateral pectoral muscle absence and frequent hand malformations.
- Thoracic anomalies such as mesocardia alter cardiac positioning, complicating invasive procedures.
- AVNRT ablation in children requires the precise localization of atrioventricular node structures.

What this case adds

- First report of AVNRT ablation in pediatric Poland syndrome with mesocardia.
- Fluoroscopyguided ablation was feasible despite altered anatomy, without 3D mapping.
- Individualized planning, integrating radiological and electrogram data, ensures safety and efficacy.

Introduction

Poland syndrome, first described in 1841, comprises a spectrum of anomalies primarily affecting the musculoskeletal structures of the thorax and upper limb, with an incidence of approximately 1:30,000 live births[1,2]. The hallmark is the complete or partial absence of the pectoralis major muscle, often associated with ipsilateral hand deformities. Cardiac displacement, such as mesocardia or dextrocardia, may occur and complicate imaging interpretation and invasive procedures[3,4].

Although arrhythmias are not typically considered part of the syndrome, isolated cases of atrioventricular nodal reentrant tachycardia (AVNRT) have been reported. To date, no description exists of an electrophysiological study and ablation in a pediatric patient with Poland syndrome and mesocardia. This case report aims to illustrate the anatomical challenges encountered during such a procedure and the adaptations required for successful ablation.

Case report

A 15-year-old girl with a known diagnosis of Poland syndrome was referred for recurrent palpitations. Her phenotype included left thoracic hypoplasia, partial absence of the pectoralis major muscle, athelia, and ipsilateral hand malformations involving metacarpal and phalangeal bones (Fig. 1). Echocardiography revealed a structurally normal heart with mesocardia.

The 24hour Holter monitoring documented episodes of supraventricular tachycardia with a long PR interval before onset, narrow QRS complexes with occasional aberrancy, and abrupt termination, consistent with AVNRT (Fig. 2). An invasive electrophysiological study (EPS) was indicated.

The EPS was performed under fluoroscopic guidance without threedimensional mapping systems. Due to mesocardia, standard catheter positions were altered: the right ventricular apex catheter was located to the right of the spine instead of the left, and the His bundle catheter was displaced from its typical site (Fig. 3). Despite this, His potentials were identified in the shifted location. Additional mapping in the left anterior oblique view showed displacement of the slow pathway region, requiring adjustments in catheter trajectory (Fig. 4).

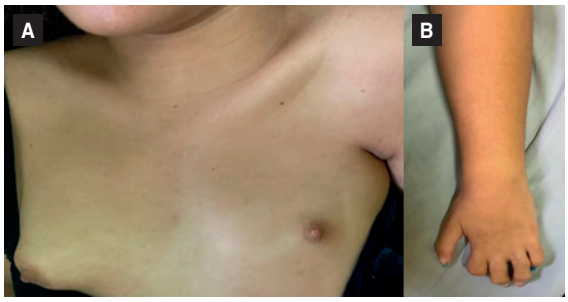

Figure 1. Clinical features of Poland syndrome. **A:** left thoracic hypoplasia with athelia. **B:** ipsilateral hand malformations (metacarpal and phalangeal anomalies).

Ablation was performed at the slow pathway region, identified by electrogram characteristics (Fig. 4). Radiofrequency energy was delivered (50 W, maximum 60 °C for 60 seconds) with an impedance drop of 10 Ω. An accelerated junctional rhythm appeared within seconds, and tachycardia was no longer inducible with or without isoproterenol. No complications occurred.

During six months of followup, the patient remained asymptomatic, with no arrhythmia recurrence on Holter monitoring or serial ECGs.

Discussion

This case demonstrates that Poland syndrome with mesocardia can significantly alter the expected position of intracardiac structures, challenging conventional catheter navigation during an EPS. Accurate localization of the His bundle and slow pathway, critical for AVNRT ablation, required the integration of fluoroscopic images with electrogram interpretation rather than reliance on fixed anatomical landmarks[5].

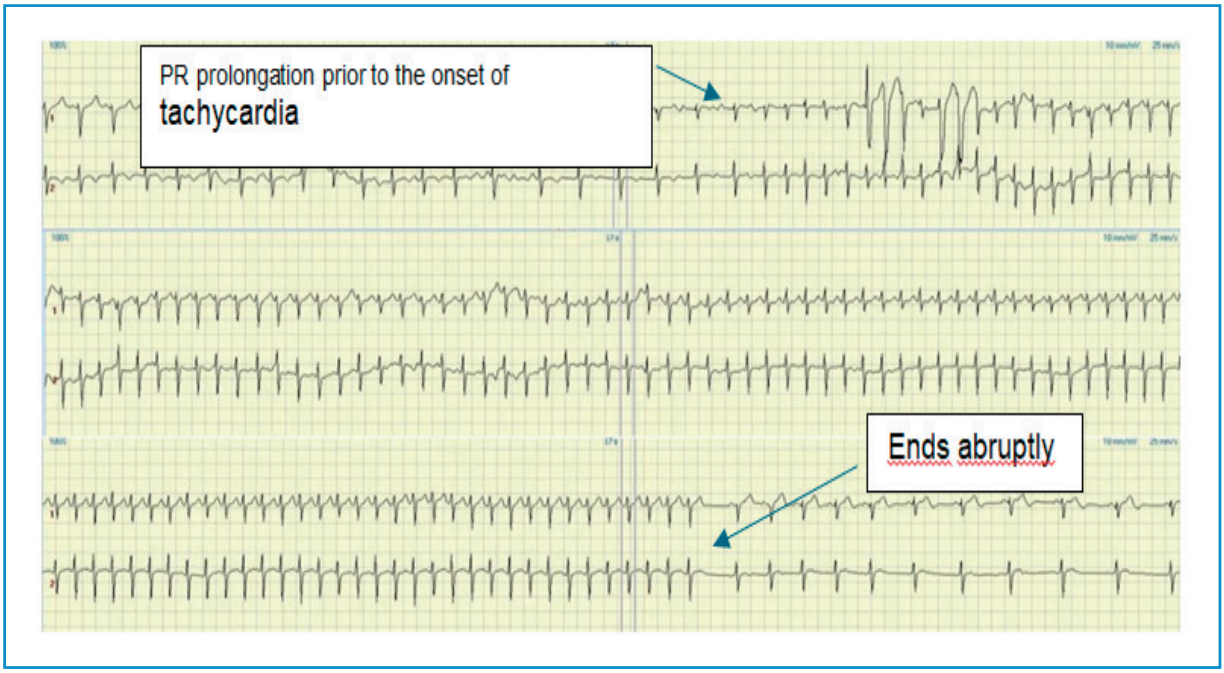

Figure 2. Holter recording showing AVNRT: prolonged PR interval before onset, narrow QRS with aberrancy, and abrupt termination.

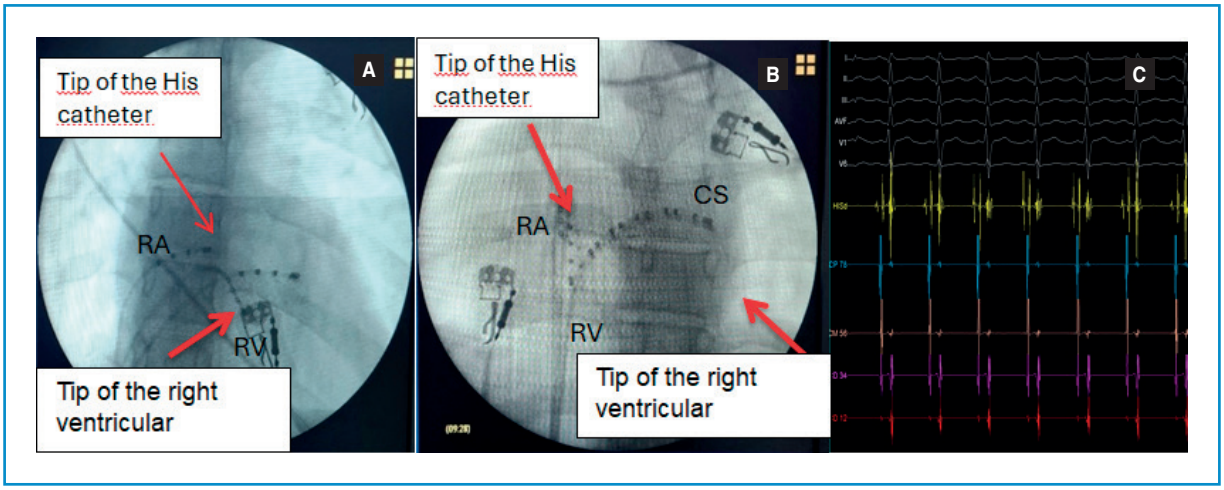

Figure 3. Catheter positions. **A:** normal levocardia (archival image), with His and right ventricular catheters left of the spine. **B:** present case, where mesocardia displaces catheters to the right of the spine. **C:** intracardiac electrograms showing His bundle recording (yellow).

Successful ablation in this patient was achieved using only fluoroscopy, without three-dimensional electroanatomical mapping. This finding aligns with previous reports of ablation in patients with thoracic anomalies such as dextrocardia, where procedural success depends on individualized adaptation rather than advanced technology[6]. However, the scarcity of published cases of Poland syndrome combined with arrhythmias underscores the need for awareness of such anatomical variants[7].

Clinically, this case emphasizes the importance of pre-procedural assessment of the thoracic anatomy in patients with congenital malformations. Multidisciplinary collaboration—including cardiology, radiology, and electrophysiology—can enhance safety and efficacy. While three-dimensional mapping systems and pre-procedural CT may offer additional precision, their absence does not preclude successful intervention when the operator is skilled in fluoroscopic navigation and electrogram analysis[8].

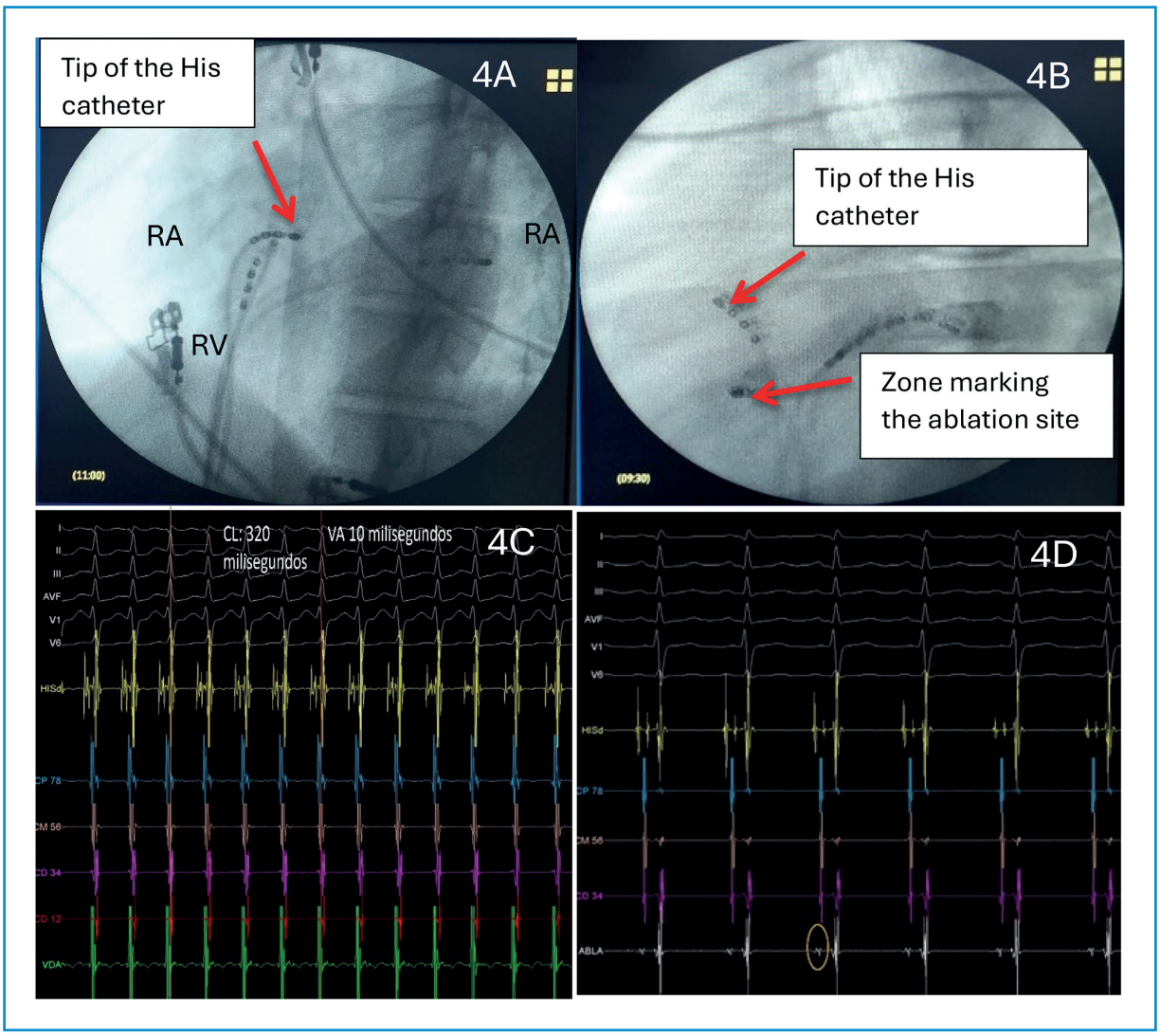

Figure 4. Left anterior oblique views. **A:** normal position (archival) with His catheter centered. **B:** present case, with displaced His catheter and slow pathway region (ablation site marked). Electrophysiological recordings. **C:** AVNRT with nearsimultaneous atrial and ventricular electrograms (VA interval 10 ms; cycle length 320 ms). **D:** slow pathway potential (marked with the oval).

The limitations of this report include its single-case nature, the lack of three-dimensional electroanatomical mapping, and a follow-up period limited to six months. These factors limit its generalizability, and further cases are needed to establish whether specific electrophysiological characteristics are consistently associated with Poland syndrome.

In conclusion, the recognition of mesocardia and other thoracic anomalies in Poland syndrome is essential for planning invasive cardiac procedures. With the careful adaptation of conventional techniques, successful AVNRT ablation is achievable, even in resource-limited settings.

Author contributions

E. Chávez-González is the lead author and chief operator for this case. A. E. Hernández-Castellón was co-operator; F. Rodríguez-González and J.M. Cruz-Elizundia were involved in the data collecton and literature review. The case was performed by E. Chávez-González, and A.E. Hernández-Castellón was involved in drafting the manuscript and editing the figures; all the other authors reviewed the manuscript.

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Conflicts of interest

None.

Ethical considerations

Protection of human subjects and animals. The authors declare that no experiments on humans or animals were performed for this research.

Confidentiality, informed consent, and ethical approval. The authors have followed their institution's confidentiality protocols, obtained informed consent from all patients, and secured approval from the Ethics Committee. SAGER guidelines have been followed as applicable to the nature of the study.

Declaration on the use of artificial intelligence. The authors declare that no generative artificial intelligence was used in the writing or creation of the content of this manuscript.

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