

Incidental diagnosis of Brugada syndrome in two girls hospitalized for pediatric inflammatory multisystem syndrome related to COVID-19 (PIMS-TS)

Magda Franke, Tomasz Marcin Książczyk, Radosław Pietrzak, Bożena Werner

Department of Pediatric Cardiology and General Pediatrics, Medical University of Warsaw, Warszawa, Poland

Correspondence to:

Prof. Bożena Werner MD, PhD,
Department of Pediatric
Cardiology and General
Pediatrics, Medical University
of Warsaw,
Żwirki i Wigury 63A,
02-091 Warszawa, Poland,
phone: +48 22 317 95 88,
e-mail:
bozena.werner@wum.edu.pl

Copyright by the Author(s), 2022

DOI: 10.33963/KPa.2022.0183

Received:

June 15, 2022

Accepted:

July 19, 2022

Early publication date:

August 4, 2022

SARS-CoV-2-related pediatric inflammatory multisystem syndrome (PIMS-TS) is a novel syndrome first described in England in May 2020 [1]. It affects children 2–6 weeks after COVID-19 infection whether symptomatic or not. The course of the disease varies from mild to requiring Intensive Care Unit support, with a 2% mortality rate reported [2]. An inherent symptom of PIMS-TS is high fever. Cardiac evaluation is crucial in PIMS-TS patients as the involvement of the cardiovascular system determines the course of disease and prognosis.

We present 2 cases of girls aged 5 (Patient 1) and 7 years (Patient 2). They were both admitted to hospital due to high unremitting fever, weakness, and mucocutaneous and gastrointestinal symptoms. Patient 2, presented with severe dyspnea and had a history of upper respiratory tract infection with a highly possible COVID-19 etiology 3 weeks earlier.

On admission, the girls presented with fever, polymorphic rash, and conjunctival redness. Patient 2 had hepatomegaly, pleural effusion, and cardiac enlargement on chest X-ray. Laboratory test results in both patients were typical of PIMS-TS with significantly elevated inflammatory parameters, cardiac biomarkers, lymphopenia, hyponatremia, and increased D-dimers. Following PIMS-TS diagnostic and treatment protocol, electrocardiography (ECG) and echocardiography were performed [3]. ECG showed abnormal repolarization with coved ST-segment elevation ≥ 2 mm, followed by negative T wave in V1 and V2 precordial leads. Such abnormalities are not characteristic of PIMS-TS.

The patients were treated for PIMS-TS in accordance with the Polish Pediatric Society

recommendations [3]. Immunomodulating therapy (immunoglobulins and glucocorticosteroids) and acetylsalicylic acid were administered. Patient 2 developed symptoms of cardiogenic shock with severely reduced left ventricular ejection fraction and required inotropic medication. They both recovered without complications. Their ECGs normalized once the fever resolved.

With the aforementioned ECG recordings, both patients met the criteria for the Brugada type 1 ECG pattern [4]. Brugada syndrome phenocopies were excluded [5]. Patients and their first-degree relatives underwent a thorough cardiological screening according to the Heart Rhythm Society/European Heart Rhythm Association/Asia Pacific Heart Rhythm Society (HRS/EHRA/APHRS) expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes [4].

Patient's 1 family's cascade screening led to the patient's father being subjected to an ajmaline challenge, which revealed a typical Brugada syndrome pattern. Moreover, the index-case paternal grandfather suffered a sudden cardiac death. The girl's genetic test with Next Generations Sequencing (NGS) testing showed a variant of unknown significance in the SCN5A gene. No other family members shared the disease. No significant events were reported in patient's 2 family. They are currently being investigated to exclude the disease in first-degree relatives of the patient. NGS testing is in progress. Both patients were then given lifestyle change recommendations, according to the HRS/EHRA/APHRS expert consensus statement [4]. However, both

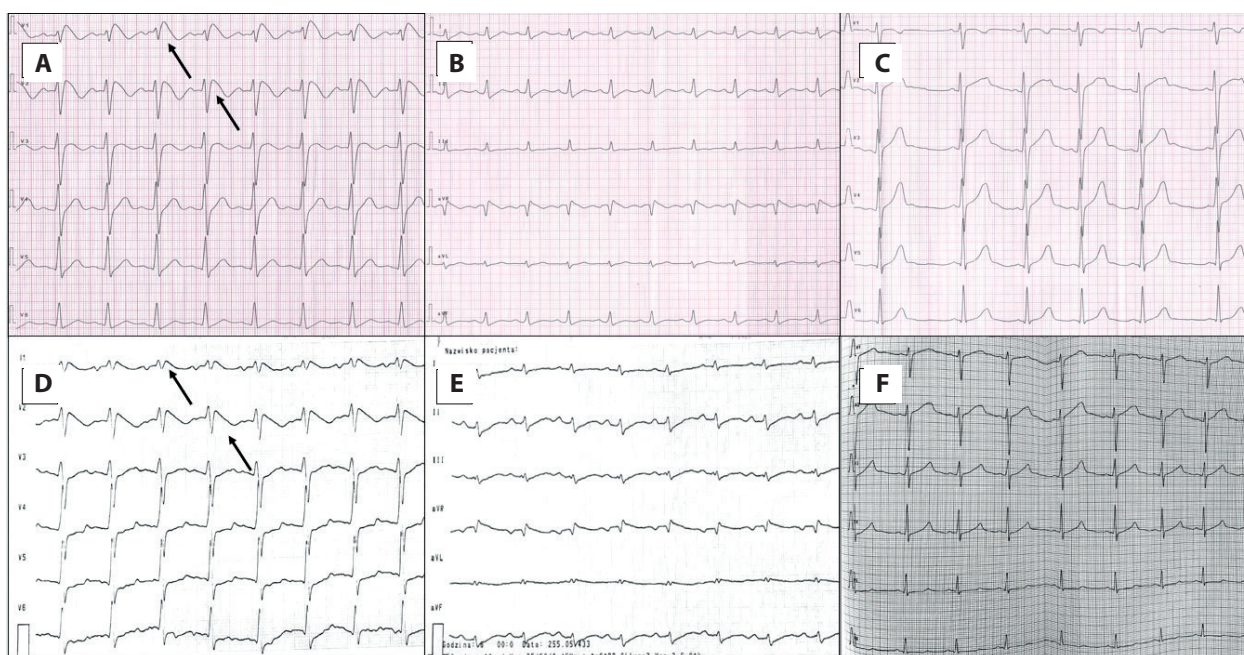


Figure 1. **A, B.** Patient 1 — ECG obtained during fever showing Brugada pattern (the black arrows) in the precordial leads. **C.** Patient 1 — normal ECG recording after defervescence, precordial leads. **D, E.** Patient 2 — ECG obtained during fever showing Brugada pattern (the black arrows) in the precordial leads. **F.** Patient 2 — normal ECG recording after defervescence, precordial leads

Abbreviations: ECG, electrocardiogram; PIMS-TS, pediatric inflammatory multisystem syndrome temporally associated with SARS-CoV-2

families were reassured about the low risk of life-threatening arrhythmias in asymptomatic and incidentally diagnosed patients.

The two cases presented a typical but severe course of PIMS-TS. High and long-lasting fever occurring in PIMS-TS exposed the abnormal electrocardiographic repolarization pattern which led to the diagnosis of Brugada syndrome. In children, ECG in fever is rarely performed, however, as stated, it may lead to a correct diagnosis.

Article information

Conflict of interest: None declared.

Funding: None.

Open access: This article is available in open access under Creative Commons Attribution-Non-Commercial-No Derivatives 4.0 International (CC BY-NC-ND 4.0) license, allowing to download articles and share them with others as long as they credit the authors and the publisher, but without permission to change them in any way or use them commercially. For commercial use, please contact the journal office at kardiologiapolska@ptkardio.pl.

REFERENCES

- Riphagen S, Gomez X, Gonzalez-Martinez C, et al. Hyperinflammatory shock in children during COVID-19 pandemic. *Lancet*. 2020; 395(10237): 1607–1608, doi: [10.1016/S0140-6736\(20\)31094-1](https://doi.org/10.1016/S0140-6736(20)31094-1), indexed in Pubmed: [32386565](https://pubmed.ncbi.nlm.nih.gov/32386565/).
- Patel JM. Multisystem Inflammatory Syndrome in Children (MIS-C). *Curr Allergy Asthma Rep*. 2022; 22(5): 53–60, doi: [10.1007/s11882-022-01031-4](https://doi.org/10.1007/s11882-022-01031-4), indexed in Pubmed: [35314921](https://pubmed.ncbi.nlm.nih.gov/35314921/).
- Okarska-Napierała M, Ludwikowska K, Jackowska T, et al. Approach to a child with Multisystem Inflammatory Syndrome associated with COVID-19. Recommendations by the Polish Pediatric Society Expert Group. Update — February 2021. *Pediatrics Polska*. 2021; 96(2): 121–128, doi: [10.5114/polp.2021.107395](https://doi.org/10.5114/polp.2021.107395).
- Priori SG, Wilde AA, Horie M, et al. HRS/EHRA/APHRS expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes: document endorsed by HRS, EHRA, and APHRS in May 2013 and by ACCF, AHA, PACES, and AEPCC in June 2013. *Heart Rhythm*. 2013; 10(12): 1932–1963, doi: [10.1016/j.hrthm.2013.05.014](https://doi.org/10.1016/j.hrthm.2013.05.014), indexed in Pubmed: [24011539](https://pubmed.ncbi.nlm.nih.gov/24011539/).
- Anselm DD, Evans JM, Baranchuk A. Brugada phenocopy: A new electrocardiogram phenomenon. *World J Cardiol*. 2014; 6(3): 81–86, doi: [10.4330/wjc.v6.i3.81](https://doi.org/10.4330/wjc.v6.i3.81), indexed in Pubmed: [24669289](https://pubmed.ncbi.nlm.nih.gov/24669289/).