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Fulminant myocarditis and acute heart failure in the light of new American Heart Association 2020 guidelines. Mechanical cardiac support and endomyocardial biopsy. What should be first?

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Fulminant myocarditis is a rapidly progressive inflammatory process of the myocardium that may either end in spontaneous recovery or lead to hemodynamic instability, cardiogenic shock and arrhythmias resulting in a high risk of death [1, 2].

The 2020 American Heart Association (AHA) position paper summarises knowledge on fulminant myocarditis and highlights its diagnostic and therapeutic aspects [3]. Prompt initial assessment and seeking early signs of hemodynamic instability is extremely important in such cases. In the light of new AHA guidelines, presented herein is the history of a 27-year-old man diagnosed and treated successfully for fulminant myocarditis.

The patient, with no medical record, was admitted to Hospital with cardiogenic shock. The medical history revealed fever, weakness, vomiting and exercise intolerance for several days. Hypotonia with features of peripheral hypoperfusion was observed. Electrocardiogram showed sinus rhythm 120 bpm, right bundle branch block, ST-segment elevation in the precordial leads (Fig. 1A). Laboratory tests revealed: C-reactive protein 100 mg/L, procalcitonin 1.37 ng/mL, troponin T > 10000 ng/L, creatine kinase-MB 75 ng/L, N-terminal pro-B-type natriuretic peptide 16497 ng/L, lactates 4.1 mmol/L. In transthoracic echocardiography (TTE) a severe dysfunction of both ventricles with left ventricular (LV) ejection fraction (LVEF) 15%, LV end diastolic dimension 60 mm, LV outflow tract velocity time integral 8 cm and tricuspid annular plane systolic excursion 12 mm were found.

Immediate coronary angiography showed no significant stenoses. Based on the entire clinical picture a working diagnosis of fulminant myocarditis was made. Catecholamines were administered and an intra-aortic balloon pump (IABP) was introduced. Despite the treatment, cardiogenic shock persisted and the patient required mechanical circulatory support (MCS) with veno-arterial extracorporeal membrane oxygenation (VA-ECMO) (Fig. 1B). Moreover, mechanical ventilation and continuous renal replacement therapy were necessary due to multiorgan failure.

Unfortunately, the following TTE showed extensive right and LV failure with LV dilatation, LVEF decreased to 8%, with signs of echogenic blood and a lack of mobility of the aortic valve leaflets (Fig. 1C). The clinical picture reflected

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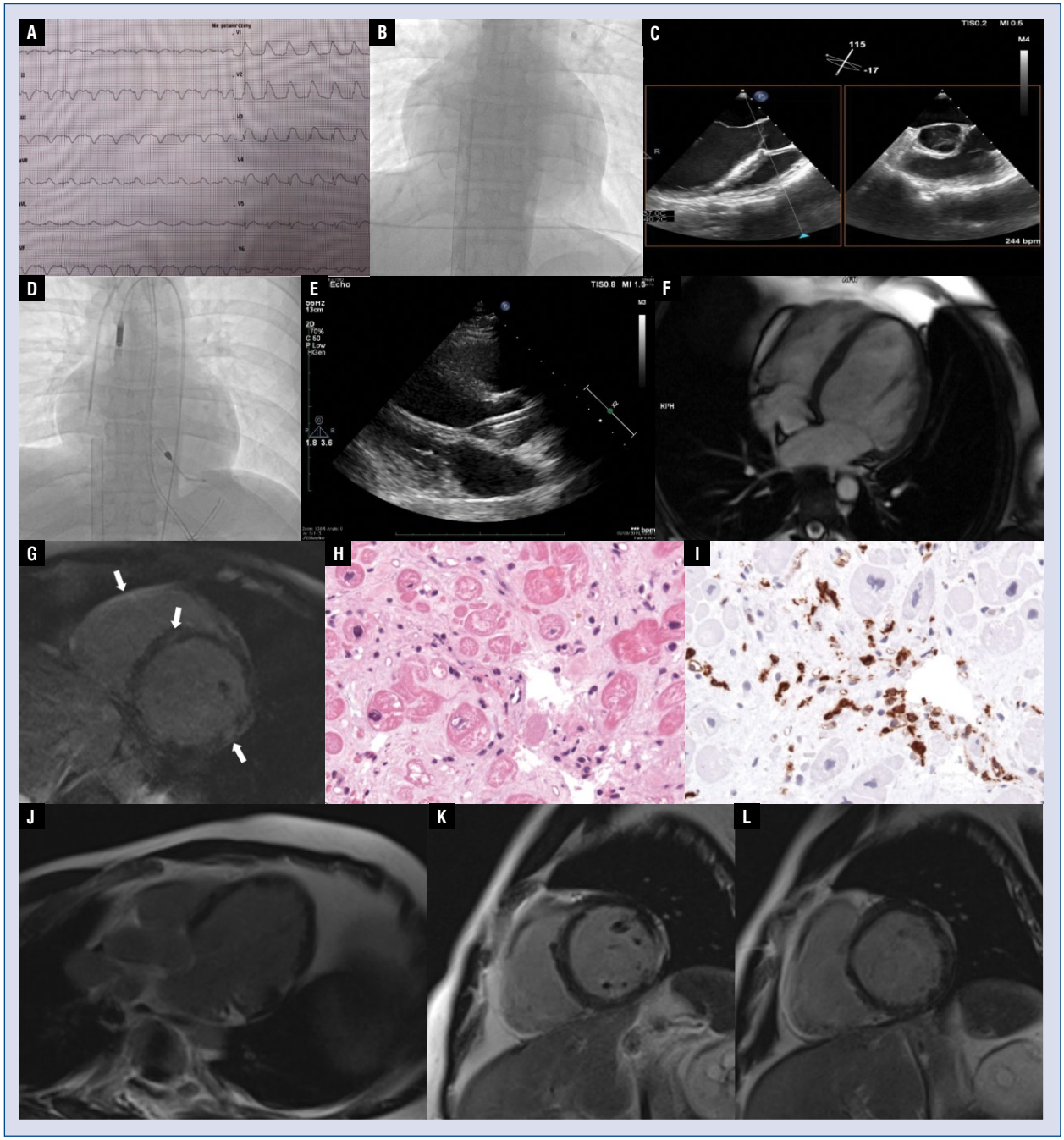

Figure 1. A. Electrocardiogram at admission; **B.** The implanted ECMO Cardiohelp, Maquet, Germany; **C.** Transthoracic echocardiography: left ventricle dilatation, presence of echogenic blood and lack of mobility of the aortic valve leaflets; **D.** Implanted Impella CP percutaneous intracardiac pump (Abiomed, Danvers MA) — angiographic image; **E.** Implanted Impella CP percutaneous intracardiac pump (Abiomed, Danvers MA) — echocardiographic image; **F, G.** Initial cardiac magnetic resonance (CMR): multiple, diffuse, intramuscular late gadolinium enhancement patterns in the myocardium of both ventricles (arrows); **H, I.** Endomyocardial biopsy: massive acute lymphocytic myocarditis, histological criteria — necrosis of adjacent muscle cells and a marked inflammatory infiltrate (Dallas criteria) as well as immunohistochemical criteria — an inflammatory infiltrate in the form of T lymphocytes at the amount of 46/mm^2; **J–L.** Follow up CMR.

a critical LV overload and therefore IABP was replaced with the Impella CP pump (Fig. 1D, E) to unload the LV.

During VA-ECMO, a hemostatic disturbance was observed with massive bleeding which required blood transfusions. In the following days

after the Impella CP implantation, TTE showed LVEF 30% and the patient's condition improved. Finally, it was possible to remove both, VA-ECMO and Impella CP.

Despite applied complex therapy, in the following days the serial TTE did not show any LVEF improvement. Short non-sustained ventricular tachycardias were observed. Therefore, the patient underwent cardiac magnetic resonance (CMR) that fulfilled the Lake Louis criteria, LVEF 23% (Fig. 1F, G).

Having immunosuppressive treatment in mind, endomyocardial biopsy (EMB) was performed, which revealed massive acute lymphocytic myocarditis (Fig. 1H, I). Simultaneously, virus infection was excluded in the myocardium by (RT-)PCRs, and therefore, steroid treatment could be initiated. Metyloprednizolon 500 mg/day i.v. for 7 days, then prednizon 65 mg/day p.o. for 4 weeks were administered. Then the dose was tapered-off every 5 days. Unfortunately, complex therapy (steroid and azathioprine) was not tolerated.

After 1 month of immunosuppressive therapy, the LV function improved. On the day of discharge, the patient's LVEF was 40% and his functional status was NYHA class II.

In a follow-up CMR performed 3 months after discharge Lake Louis criteria were not fulfilled, slight reduction of late gadolinium enchancement extend and LVEF 42% were observed (Fig. 1J–L).

Patients with fulminant myocarditis often require intensive care treatment with inotropic drugs and MCS. The latter usually allows achieving haemodynamic stabilisation. Recent data show that prolonged MCS has a positive effect on the cellular and molecular mechanisms responsible for myocardial remodeling, fibrosis, inflammation, and calcium metabolism, which increases the chance of recovery [4].

In the presented history, the IABP used in the first hours was ineffective and the patient required VA-ECMO [5]. The data indicate that the most commonly used MCS for fulminant myocarditis is VA-ECMO, which draws blood from the right atrium and pumps it into the aorta. At the same time, however, it increases the afterload of the damaged LV because it pumps blood in the opposite direction to the incoming blood flow from the heart. It puts additional strain on the LV and can lead to a dramatic reduction or even cessation of cardiac output with LV dilation. This condition generates excessive LV wall stress, impairs coronary flow and increases the risk of thrombus formation. In this situation rapid unloading of the LV is absolutely necessary. In our patient, the successive unloading was achieved by replacing IABP with the Impella CP which is a microaxial pump implanted percutaneously through the aortic valve into the LV. By pumping blood from the LV into the ascending aorta at a maximum rate of up to 4 L/min, it significantly reduces LV end-diastolic pressure and volume. The pump also allows for a reduction in VA-ECMO support, thus reducing the afterload. The combination of VA-ECMO with the Impella CP pump created a new name for this complex therapy — ECMELLA. According to the registry data, in patients treated this way, a significant reduction of in-hospital mortality and a higher rate of treatment success were observed [4, 6].

The AHA 2020 position statement emphasizes the importance of early imaging and invasive diagnosis. The timing of the latter, myocardial biopsy, depends on the anticoagulation used and the experience of the operator [3]. Performing EMB on admission was considered, however, due to the critical condition of the patient, massive multisite bleeding, complications related to VA-ECMO therapy, the EMB was performed after termination of MCS, during a stable period of the disease. Recent data showed a greater number of complications in EMB performed during MCS, albeit it led to histopathologic diagnosis. Researchers agree that the management of this patient population requires further refinement to improve procedural safety [7].

The presented report demonstrates that a patient suffering from fulminant myocarditis requires the cooperation of an interdisciplinary team. The main goal of treatment is the fastest possible stabilization of the cardiovascular system, therefore MCS should be the first procedure. MCS enables further diagnosis and adequate treatment. There is also evidence that it gives the inflamed myocardium a chance to regenerate. Taking in to consideration the benefits and risks, EMB should be performed as soon as possible, in order to initiate adequate treatment [8].

Conflict of interest: None declared

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