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OPIS PRZYPADKU
CASE REPORT

Electrocardiographic history of pulmonary arterial hypertension – from diagnosis, through pregnancy, to lung transplantation

Obraz elektrokardiograficzny tętniczego nadciśnienia płucnego – od diagnozy poprzez ciążę do przeszczepienia płuc

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ABSTRACT

The frequency and nature of electrocardiographic changes in patients with pulmonary hypertension varies significantly depending on the severity of the illness. In mild cases, electrocardiogram (ECG) may be normal. We present the consecutive ECGs of a patient with pulmonary arterial hypertension (PAH) in the 5-year course of the disease – from diagnosis to lung transplantation. The described case shows the variability of ECG changes in correlation with the clinical state of the patient. The observed ECG abnormalities in PAH are reversible and in the presented case ECG normalized after lung transplantation.

KEYWORDS

electrocardiography, pulmonary arterial hypertension, right ventricular hypertrophy

STRESZCZENIE

Częstość i charakter zmian w zapisach elektrokardiograficznych u pacjentów z nadciśnieniem płucnym znacząco różni się zależnie od ciężkości choroby. U pacjentów z łagodnym nadciśnieniem płucnym zapis elektrokardiograficzny (EKG) może być prawidłowy. W niniejszej pracy prezentujemy kolejne zapisy EKG pacjentki z tętniczym nadciśnieniem płucnym (*pulmonary arterial hypertension* – PAH) w 5-letnim okresie choroby – od postawienia diagnozy aż do przeszczepienia płuc. Opisany przypadek pokazuje różnorodność zmian w zapisach EKG, których nasilenie korelowało ze stanem klinicznym pacjentki. Obserwowane nieprawidłowości w zapisach EKG w przypadku PAH są odwracalne i w prezentowanym przypadku doszło do ich normalizacji po przeszczepieniu płuc.

SŁOWA KLUCZOWE

elektrokardiografia, tętnicze nadciśnienie płucne, przerost prawej komory serca

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INTRODUCTION

The electrocardiographic changes in patients with pulmonary arterial hypertension (PAH) may vary depending on the severity of the illness. In mild cases, electrocardiogram (ECG) may be normal. The most common findings in moderate and severe cases are right atrial and/or right ventricular hypertrophy (RVH), right ventricular (RV) strain, right axis deviation, right bundle branch block (RBBB), corrected QT (QTc) interval prolongation, sinus tachycardia and atrial arrhythmias [1]. We present the consecutive ECGs of a patient with PAH in the 5-year course of the disease.

MATERIAL AND METHODS

Standard 12-lead ECGs at rest, two-dimensional and Doppler transthoracic echocardiography (TTE) were performed on admission to hospital. The ECG recordings and TTE were assessed by an experienced cardiologist. There are several ECG parameters used to assess RVH. To evaluate the RVH features in the ECG of the presented patient, we used six well-known criteria [2,3,4]:

- R wave in aVR ≥ 0.5 mV
- R wave in V1 ≥ 0.7 mV
- rSR' pattern in V1–R' > 1mV (QRS < 120 ms)
- S wave in V5 > 1 mV
- S wave in V6 > 0.3 mV
- R wave in V1 + S wave in V5 or V6 > 1.05 mV.

RV strain was defined as T-wave inversion in the right precordial leads (V1-V3 ± V4) or inferior leads (II, III, aVF) or a S1Q3T3 pattern (a large S wave in lead I, a Q wave in lead III, and an inverted T wave in lead III).

CASE REPORT

A 30-year-old female was admitted to hospital due to dyspnoea at rest. Pneumonia, other pulmonary diseases, acute pulmonary embolism and connective tissue disorders were excluded. Sinus rhythm with right axis deviation, RVH and RV strain were present in the ECG at admission (Figure 1).

TTE showed a high probability of pulmonary hypertension. Right heart catheterisation confirmed the diagnosis of severe PAH with a positive response to vasoreactivity testing. Sildenafil and diltiazem were administered and led to improvement in the clinical condition of the patient. After 2-months of treatment the patient was in stable condition, but also 8-weeks pregnant. Our patient decided to maintain the pregnancy. After 6 months from diagnosis, at 25 weeks pregnant, she was in good condition, with no symptoms. The ECG is shown in Figure 2.

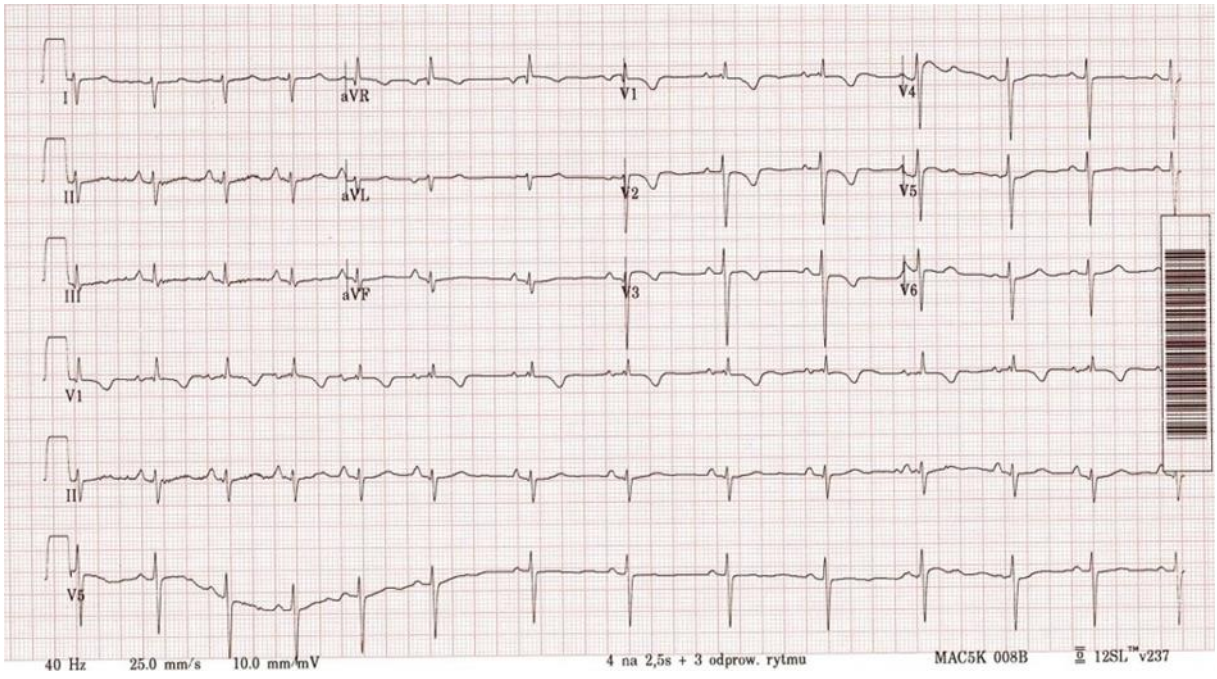

Fig. 1. Electrocardiogram (ECG) at diagnosis.
Ryc. 1. Zapis elektrokardiograficzny (EKG) w momencie rozpoznania.

The patient was in optimal clinical condition at 30 weeks of gestation. She was admitted to the hospital in the 31st week of gestation due to rapid clinical deterioration – rest dyspnoea and low blood pressure. In the ECG, we found sinus tachycardia, features of RVH and RV strain and slight prolongation of the QTc interval (Figure 3). TTE showed severe RV pressure overload. Right heart catheterisation revealed the progression of PAH and negative vasoreactivity testing.

The patient also developed pneumonia. Antibiotics and epoprostenol were administrated. An urgent caesarean delivery was necessary 4 days after admission due to bleeding from the genital tract. After delivery the patient was in a critical state, was treated with arteriovenous extracorporeal membrane oxygenation (ECMO) for 7 days due to deterioration of heart and respiratory failure. After 18 days in the intensive care unit the patient returned to the cardiology ward, where a Hickman tunnelled catheter was implanted for epoprostenol administration [5]. The ECGs after delivery are presented in Figures 4 and 5.

Two years after the initial diagnosis, the patient was in optimal condition – the World Health Organization functional class I (WHO-FC). Right axis deviation and incomplete RBBB were observed in the ECG without signs of RVH and RV strain (Figure 6).

Four years after diagnosis the patient started to rapidly deteriorate despite optimal treatment. Signs of RVH and RV strain appeared again in the ECG (Figure 7).

The patient met the criteria for lung transplantation (LTx). The procedure was performed 6 months after qualification. Normalization of the ECG was observed after the procedure (Figure 8). A summary of the ECG analyses, TTE parameters and haemodynamic features are shown in Table I.

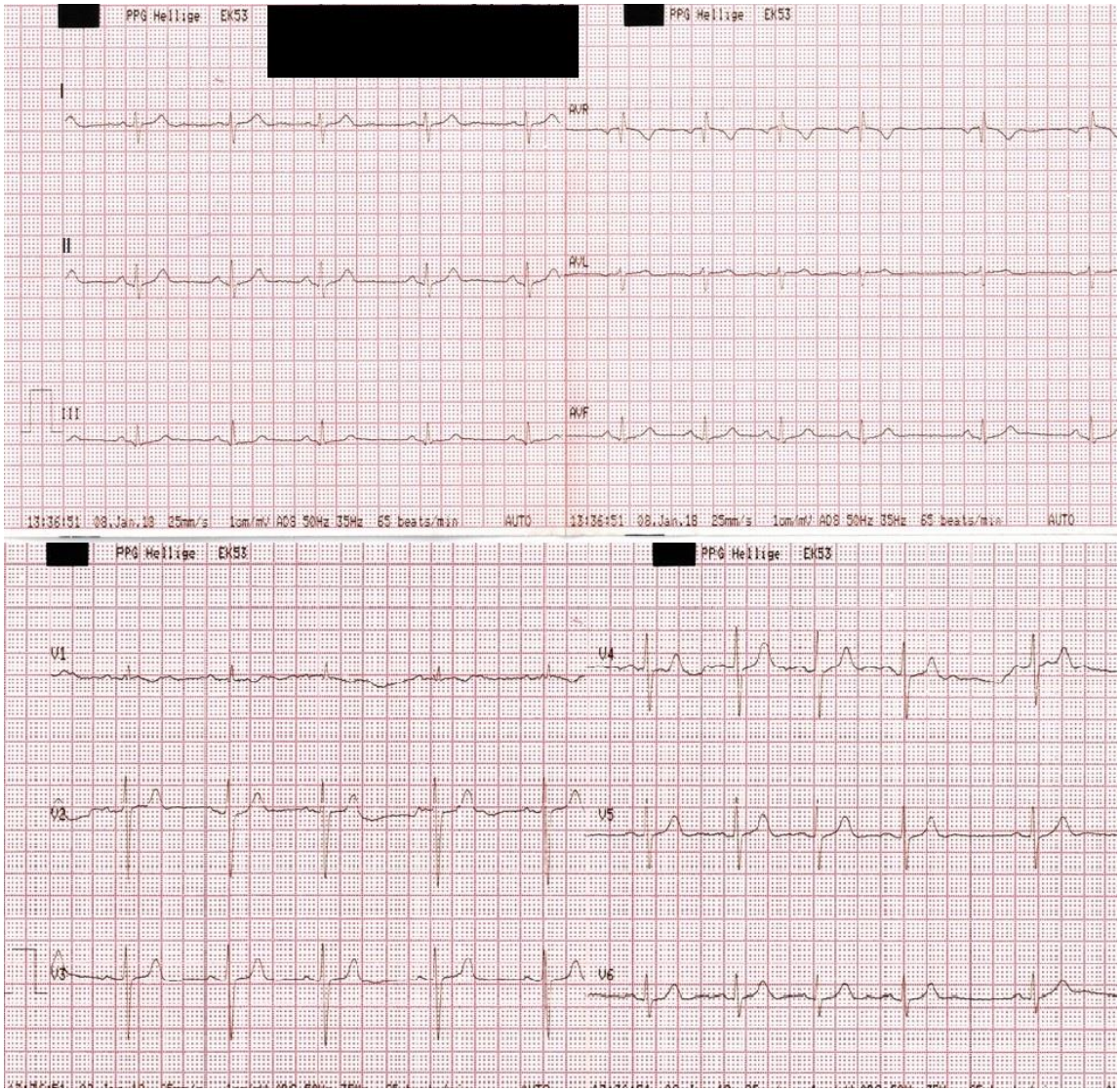

Fig. 2. Electrocardiogram (ECG) at 25-weeks of gestation.
Ryc. 2. Zapis elektrokardiograficzny (EKG) w 25. tygodniu ciąży.

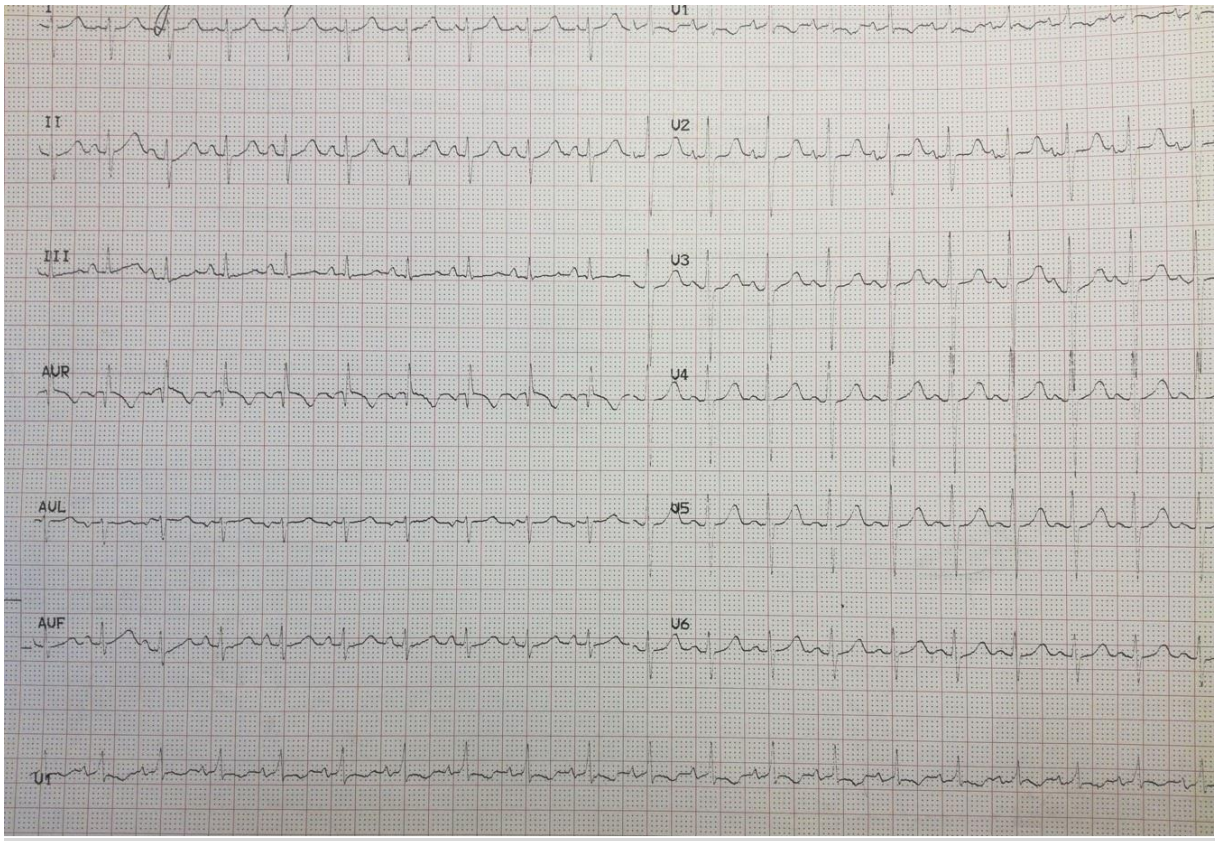

Fig. 3. Electrocardiogram (ECG) at 31st week of gestation.
Ryc. 3. Zapis elektrokardiograficzny (EKG) w 31. tygodniu ciąży.

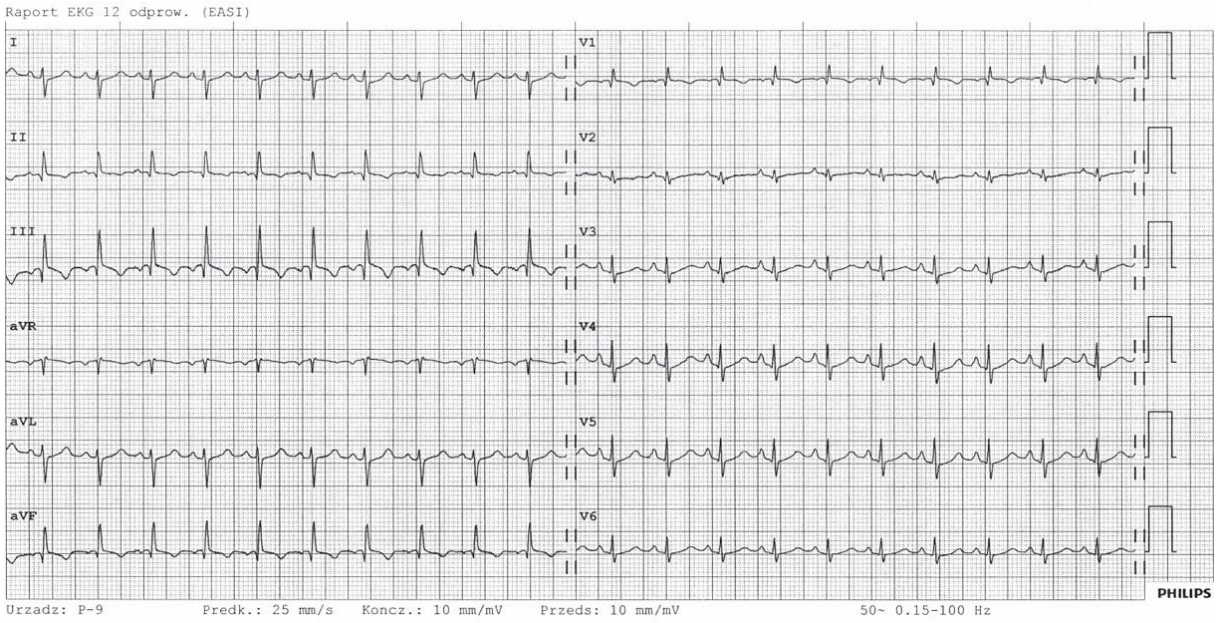

Fig. 4. Electrocardiogram (ECG) 2 days after delivery.
Ryc. 4. Zapis elektrokardiograficzny (EKG) 2 dni po porodzie.

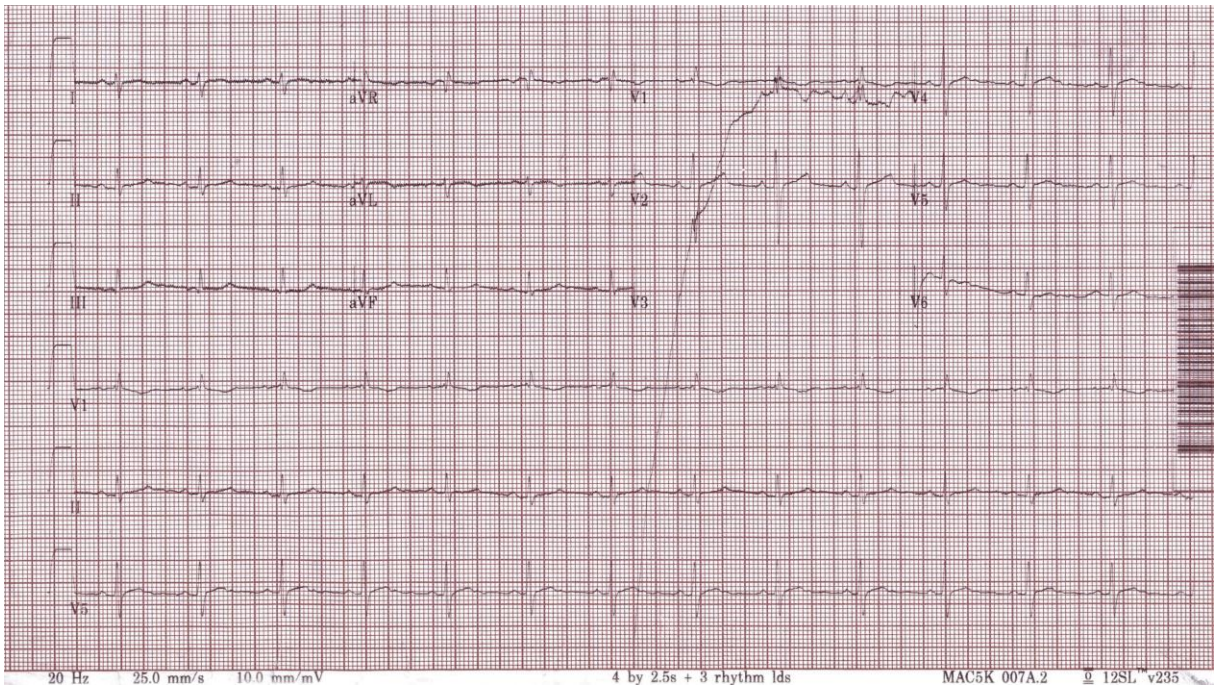

Fig. 5. Electrocardiogram (ECG) 1 month after delivery.
Ryc. 5. Zapis elektrokardiograficzny (EKG) miesiąc po porodzie.

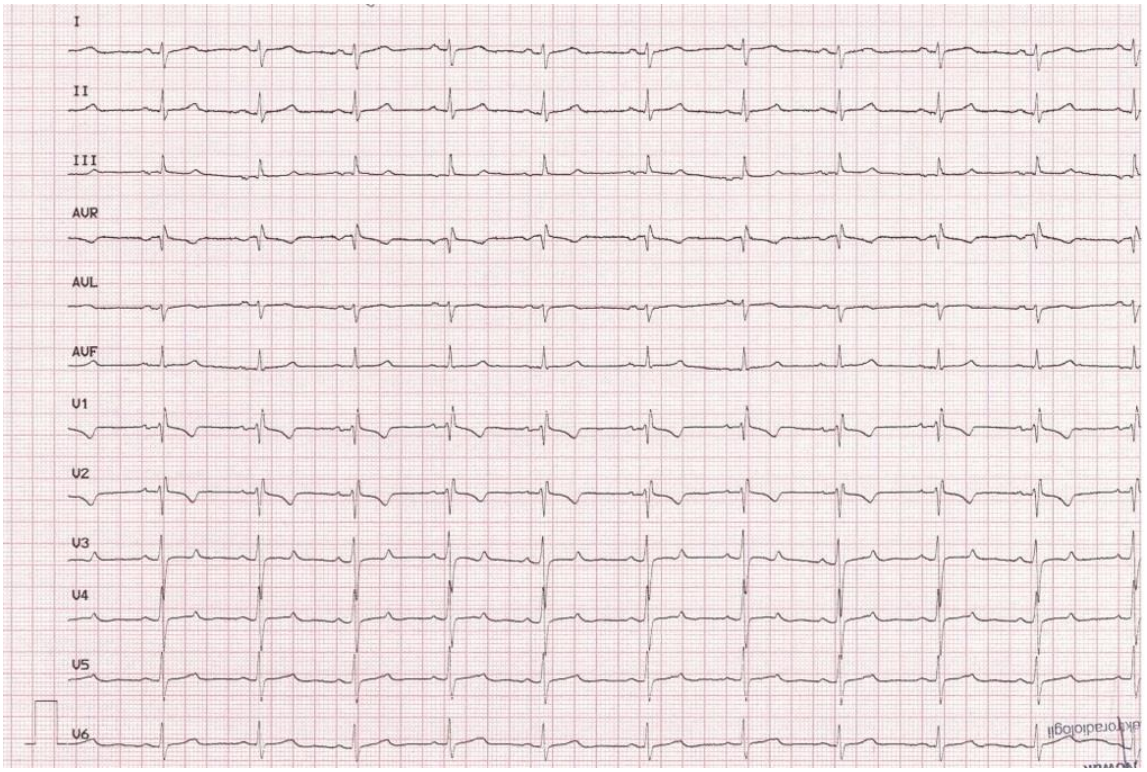

Fig. 6. Electrocardiogram (ECG) 2 years after diagnosis.
Ryc. 6. Zapis elektrokardiograficzny (EKG) 2 lata od rozpoznania.

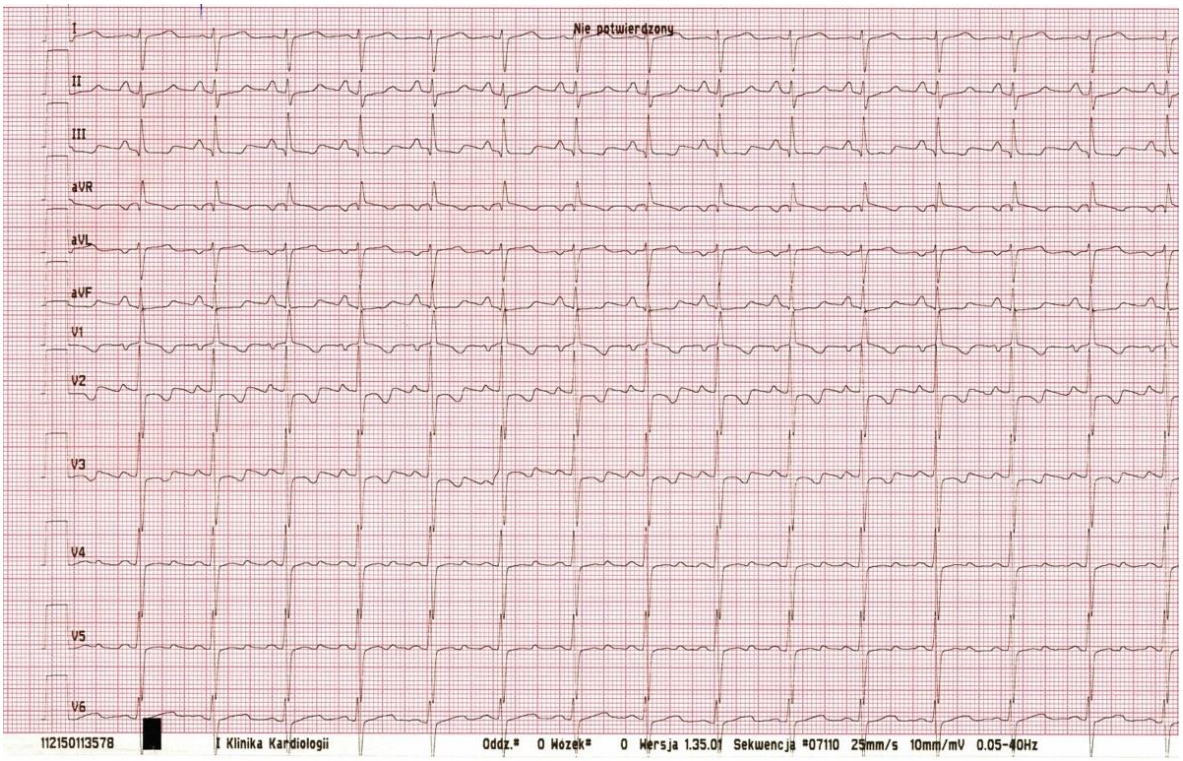

Fig. 7. Electrocardiogram (ECG) 4 years after diagnosis.
Ryc. 7. Zapis elektrokardiograficzny (EKG) 4 lata od rozpoznania.

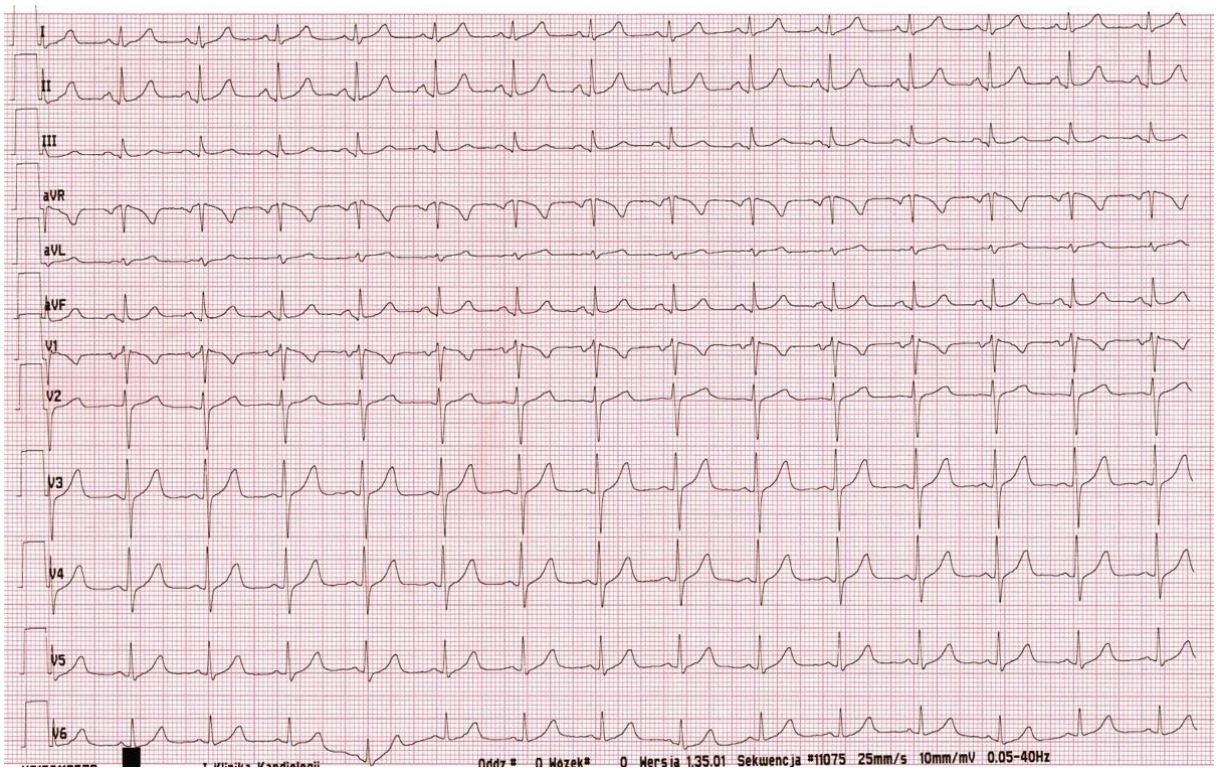

Fig. 8. Electrocardiogram **(**ECG) 4 months after lung transplantation.
Ryc. 8. Zapis elektrokardiograficzny (EKG) 4 miesiące po przeszczepieniu płuc.

Table I. Comparison of clinical, echocardiographic, haemodynamic and electrocardiographic features in course of disease
Tabela I. Porównanie parametrów klinicznych, echokardiograficznych i elektrokardiograficznych w przebiegu choroby

Variable	Diagnosis	25 wks (6 months after diagnosis)	31 wks	2 days after delivery	1 month after delivery	2 years after diagnosis	4 years after diagnosis	4 month after LTx
WHO-FC	IV	I	IV	IV	III	I	III	I
NT-proBNP (pg/ml)	1835	86	1579	NA	507	166	1638	NA
RHC								
mPAP (mm Hg)	56*	NA	54	61#	32	NA	74	NA
PVR (Wood Unit)	6.7	NA	11.4	NA	–	NA	15.4	NA
TTE								
RAA (cm^2)	13	14	19	20	17	15	23	12
TAPSE (mm)	16	–	13	12	–	17	15	20
RVSP (mm Hg)	108	78	115	120	87	62	114	NA
Pericardial effusion	no	no	no	no	yes	no	no	no
6 MWT (m)	0	570	NA	NA	NA	400	20	NA
ECG								
HR (bpm)	79	65	130	127	81	66	90	86
Axis	**right**	**right**	**right**	**right**	**right**	**right**	**right**	normal
1. P wave > 0.25 mV in lead II	no	no	no	no	no	no	no	no
2. Initial positivity of P wave in V1 or V2 ≥ 0.15 mV	no	no	no	**yes**	no	no	no	no
RVH parameters								
R wave in aVR ≥ 0.5 mV	**yes**	no	**yes**	no	no	no	**yes**	no
R wave in V1 ≥ 0.7 mV	no	no	**yes**	no	no	no	**yes**	no
rSR' in V1–R' > 1 mV (QRS < 120 ms)	no	no	–	–	no	no	–	no
S wave in V5 > 1 mV	**yes**	no	no	no	no	no	**yes**	no
S wave in V6 > 0.3 mV	**yes**	**yes**	**yes**	no	**yes**	no	**yes**	no
R in V1 + S in V5 or V6 > 1.05 mV	**yes**	no	**yes**	no	no	no	**yes**	no
Features of RV strain	**yes**	no	**yes**	**yes**	no	no	**yes**	no
IRBBB or RBBB	no	no	no	no	no	**yes**	no	no
QTc (N: ≤ 460 ms)	409	400	**464**	424	423	425	440	440
AF/AFl/AT	no	no	no	no	no	no	no	no

* vasoreactive; # via Swan-Ganz catheter

6MWT – six-minute walk test; AF – atrial fibrillation, AFl – atrial flutter; AT – atrial tachycardia; bpm – beats per minutes; ECG – electrocardiogram; wks – weeks of gestation; HR – heart rate; IRBBB – incomplete right bundle branch block; LTx – lung transplantation; mPAP – mean pulmonary artery pressure; NA – not available; NT-proBNP – N-terminal pro-brain natriuretic peptide; PVR – pulmonary vascular resistance; QTc – corrected QT interval; RAA – right atrial area; RBBB – right bundle branch block; RHC – right heart catheterisation; RV – right ventricular; RVH – right ventricular hypertrophy; RVSP – right ventricular systolic pressure; TAPSE – tricuspid annular plane systolic excursion; TTE – transthoracic echocardiography; WHO-FC – the World Health Organization functional class.

DISCUSSION

The present case focused on the electrocardiographic changes in a patient with PAH in the course of the disease – from diagnosis to LTx. It shows the dynamic ECG changes in correlation with the clinical state of the patient. Right axis deviation was present in all the ECGs until the LTx was performed. The presence of the features of RVH, RV strain and right atrial enlargement seems to be dependent on pulmonary haemodynamics. There were no significant atrial or ventricular arrhythmia during the observation.

ECG changes are not specific for PAH, however, several studies show a predictive value of ECG in adult PAH patients [6,7,8,9,10,11,12]. Igata et al. [6] demonstrate that the amplitude of RV1+SV5/6 inversely correlates with mean pulmonary artery pressure (mPAP) and right ventricular ejection fraction and patients with an amplitude greater than 16.4 mm had a worse prognosis during follow-up. The study on chronic thromboembolic pulmonary hypertension

patients revealed that the S waves in V5, R waves in V1 + S waves in V5, S waves in I, and the QRS axis were predictors of mPAP ≥ 30 mm Hg [7]. Change in mPAP was associated with a change in the amplitudes in the ECG after balloon pulmonary angioplasty (BPA) and improvement in an R in V1 + S in V5 predicts a lower functional class in a 6-month follow-up. The correlation between ECG changes after BPA and haemodynamics was also found in a study by Piłka et al. [13]. Other studies revealed that a decrease in the amplitude of the R wave in V1 during treatment indicates patients with better prognosis [8,10]. Michalski et al. [11] found a correlation with the ECG parameters during six-minute walk test (6MWT), echocardiographic and haemodynamic features of PAH. One study compared the initial ECG with the ECG close to death in patients with PAH, and similar to our presented case, significant progression of changes in the ECG was found [14]. In a study focused on patients with PAH secondary to connective tissue disease, up to 13% of the patients had normal ECG [12]. ECG changes in the paediatric PAH population also are related to the haemodynamic status and long--term prognosis [16]. ECG abnormalities are common in this population and features of RVH and right axis deviation correlated with pulmonary vascular resistance (PVR) and the transpulmonary gradient.

ECG evolution after LTx has not been extensively investigated. Most of the studies have focused on arrhythmias after the procedure and their prognostic values [17,18,19,20,21,22]. Atrial arrhythmias (atrial fibrillation, atrial flutter and atrial tachycardia) occurred in 11–30% of patients after LTx. Kim et al. [17] reported a worse one-year survival in those patients. In a case study of 4 patients with pulmonary hypertension after LTx, a leftward shift in the QRS axis, a decrease in the P-wave amplitude and a reduction in the RV force were observed [23]. Similar findings were described in another case report [24].

ECG is widely accessible, but its prognostic value in PAH patients is underestimated. ECG cannot be used as a screening tool for detecting PAH, because it may be normal, when mild elevation of mPAP is observed.

CONCLUSIONS

This case shows the variability of ECG changes in correlation with the clinical state of the patient. ECG abnormalities are not specific for PAH; nevertheless, they reflect the severity of RV pressure overload. ECG changes in PAH are reversible and in the presented case, ECG normalization was observed after LTx.

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Conflict of interest

The authors declare no conflict of interest

Author's contribution

Study design – K. Mizia-Stec, K. Bula
Manuscript preparation – K. Bula, K. Mizia-Stec
Literature research – K. Bula, K. Mizia-Stec, M. Grabka
Final approval of the version to be published – K. Mizia-Stec, K. Bula, M. Grabka

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