





Liver transplantation in a patient with Budd-Chiari syndrome as a factor decreasing autonomic system dysfunction and risk of sudden cardiac death

Przeszczepienie wątroby u pacjenta z zespołem Budda-Chiariego jako czynnik poprawiający funkcję układu autonomicznego oraz obniżający ryzyko nagłego zgonu sercowego

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Abstract

The authors present the impact of liver transplantation in a patient with Budd-Chiari syndrome on the improvement of autonomic system function and decrease the risk of sudden cardiac death (SCD). The case shows that not only implantable cardioverter-defibrillator is a factor protecting patients with cirrhosis against SCD, but also liver transplantation itself.

Key words: liver transplantation, Budd-Chiari syndrome, cirrhosis, autonomic system dysfunction, HRV, sudden cardiac death

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Introduction

Budd-Chiari syndrome (BCS) is a rare condition characterized by impaired drainage of blood from the veins in the liver. This disorder can be asymptomatic, but it can also result in liver failure and cirrhosis leading to autonomic system dysfunction and an increased risk of sudden cardiac death (SCD). Heart rate variability (HRV) analysis on a 24-hour Holter electrocardiography (ECG) is commonly used to

assess the function of the autonomic system. It reflects the influence of the autonomic system on the work of the heart [1]. The HRV time-domain analysis considers the following parameters (among others):

- SDNN – the standard deviation of all RR intervals, describes the overall variability of sinus rhythm;
- rMSSD – the square root of the mean of the sum of the squares of the differences between consecutive RR intervals;

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– triangle index – total number of RR intervals divided by the number of RR intervals of the most common duration.

HRV spectrum power in frequency range analysis is also useful – it evaluates the following parameters: ultra-low frequencies [(ULF) 0–0.0033 Hz], very low frequencies [(VLF) 0.0033–0.04 Hz], low frequencies [(LF) 0.04–0.15 Hz], high frequencies [(HF) 0.15–0.4 Hz]. In patients with liver cirrhosis attention should be paid also to the length of the QT interval. Its extension often coexists and consequently may lead to ventricular arrhythmias and sudden cardiac death.

Case report

A previously healthy 29-year-old man was admitted to the hospital due to increasing epigastric pain. During diagnostics, elevated liver enzymes and hepatosplenomegaly without any other abnormalities were found. The complaints were resolved and the patient was discharged with the recommendation of a liver diet and a check-up at the Liver Disease Outpatient Clinic. Six months later symptoms of liver failure decompensation appeared: abdominal pain, jaundice, rapidly increasing ascites, moderate/severe general condition. Imaging and laboratory tests revealed:

thrombosis of the portal and superior mesenteric vein, increased parameters of haemoglobin, haematocrit, red blood cells and JAK2 V617F mutation. The patient was diagnosed with decompensated liver cirrhosis due to BCS, which occurred during polycythaemia vera and was qualified for a liver transplant. During the qualification, a cardiological consultation and a 24-hour Holter ECG were performed. The same test was repeated 6 months after the transplant. Based on the results, the function of the autonomic system and the risk of sudden cardiac death before and after liver transplantation were compared. For this purpose, was used the analysis of QTc interval and heart rate variability:

- HRV through time-domain variables and QTc assessment (Table 1);
- HRV through spectral-domain variables assessment (Table 2).

The obtained results show an improvement in parameters both in the time (SDNN from 51 to 169 ms, rMSSD from 5 to 19 ms, triangle index from 11 to 51) and frequency-domain analysis (decrease in LF and HF values, increase in LF/HF ratio). This means a marked change in HRV and thus an improvement in the function of the autonomic system. In addition, the QTc interval was shortened (from 446 to 407), reducing the risk of SCD in the mechanism of ventricular arrhythmias.

Discussion

The autonomic system controls the heart work in both health and disease. Through the vagus nerve and acetylcholine, it slows down its work directly affecting the sinus node, while through sympathetic impulses, adrenaline and noradrenaline – accelerates it (through beta receptors). Both components remain in a state of dynamic balance, guaranteeing that the rhythm rate is adequate to the needs of the organism. Autonomic dysfunction increases cardiovascular risk and risk of sudden cardiac death. One

Table 1. Heart rate variability time-domain variables and QTc

Parameter	Before transplantation	After transplantation
SDNN [ms]	51	169
rMSSD [ms]	5	19
Triangle index	11	51
QTc [ms]	446	407

SDNN – standard deviation of NN; rMSSD – root mean square of successive differences

Table 2. Heart rate variability spectral power in frequency range parameters before and after transplantation

Time (± 10 min)	LF [ms ²]		HF [ms ²]		LF/HF ratio	
	Before	After	Before	After	Before	After
21:25	1320	32	116	2	11.422	14.070
22:25	543	19	183	2	2.962	7.660
23:25	1481	25	153	3	5.845	8.175
00:25	308	31	160	3	1.926	11.221
01:25	2017	20	282	3	7.154	6.386
02:25	575	373	248	13	2.316	29.429
03:25	556	138	159	3	3.495	45.309
04:25	1204	267	159	4	7.560	67.718
05:25	1807	28	234	4	7.718	6.973

LF – low frequency; HF – high frequency

of the indicators of its function is HRV, which prognostic significance of the occurrence of life-threatening arrhythmias and sudden cardiac death has been proven in the group of patients after myocardial infarction. In these patients, decreased HRV parameters in the time-domain analysis were correlated with an increase in mortality – particularly in those with SDNN < 50 ms [2]. On the other hand, LF has a special predictive value in spectral assessment [3]. Such disorders can be seen in many diseases including the liver: hepatitis C [4], non-alcoholic fatty liver disease [5], primary biliary cirrhosis [6], cirrhosis of the liver in the course of other disorders. The described case shows how

the initial disease, which is polycythaemia vera, can lead to a decrease in HRV by inducing BCS and consequently cirrhosis of the liver. In cirrhosis, QTc prolongation is additionally observed, which intensifies the already increased cardiovascular risk of the patient. To protect against SCD, a cardioverter-defibrillator is usually implanted. However, as the present case shows, lowering this risk can also be achieved through liver transplantation. The literature describes similar observations after transplantation due to cirrhosis of the liver caused by other diseases than BCS [7]. This treatment significantly improves HRV parameters, and thus reduces the risk of sudden cardiac death.

Streszczenie

W artykule przedstawiono wpływ przeszczepienia wątroby u pacjenta z zespołem Budda-Chiari na poprawę funkcji układu autonomicznego oraz obniżenie ryzyka zgonu z powodu nagłego zatrzymania krążenia (SCD). Przypadek ukazuje, że nie tylko wszczepienie kardiowertera-defibrylatora jest czynnikiem chroniącym pacjentów z marskością wątroby przed SCD, ale może być nim również samo przeszczepienie wątroby.

Słowa kluczowe: przeszczepienie wątroby, zespół Budda-Chiari, marskość wątroby, dysfunkcja układu autonomicznego, HRV, nagły zgon sercowy

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Reference

- Berntson GG, Bigger JT, Eckberg DL, et al. Heart rate variability: origins, methods, and interpretive caveats. *Psychophysiology*. 1997; 34(6): 623–648, doi: [10.1111/j.1469-8986.1997.tb02140.x](https://doi.org/10.1111/j.1469-8986.1997.tb02140.x), indexed in Pubmed: [9401419](https://pubmed.ncbi.nlm.nih.gov/9401419/).
- Kleiger RE, Miller JP, Bigger JT, et al. Decreased heart rate variability and its association with increased mortality after acute myocardial infarction. *Am J Cardiol*. 1987; 59(4): 256–262, doi: [10.1016/0002-9149\(87\)90795-8](https://doi.org/10.1016/0002-9149(87)90795-8), indexed in Pubmed: [3812275](https://pubmed.ncbi.nlm.nih.gov/3812275/).
- Tsuji H, Venditti FJ, Manders ES, et al. Reduced heart rate variability and mortality risk in an elderly cohort. The Framingham Heart Study. *Circulation*. 1994; 90(2): 878–883, doi: [10.1161/01.cir.90.2.878](https://doi.org/10.1161/01.cir.90.2.878), indexed in Pubmed: [8044959](https://pubmed.ncbi.nlm.nih.gov/8044959/).
- Poliwczak AR, Białkowska J, Woźny J, et al. Cardiovascular risk assessment by electrocardiographic Holter monitoring in patients with chronic hepatitis C. *Arch Med Sci*. 2020; 16(5): 1031–1039, doi: [10.5114/aoms.2020.96600](https://doi.org/10.5114/aoms.2020.96600), indexed in Pubmed: [32863991](https://pubmed.ncbi.nlm.nih.gov/32863991/).
- Kumar MS, Singh A, Jaryal AK, et al. Cardiovascular autonomic dysfunction in patients of nonalcoholic fatty liver disease. *Int J Hepatol*. 2016; 2016: 5160754, doi: [10.1155/2016/5160754](https://doi.org/10.1155/2016/5160754), indexed in Pubmed: [28053786](https://pubmed.ncbi.nlm.nih.gov/28053786/).
- Keresztes K, Istenes I, Folhoffer A, et al. Autonomic and sensory nerve dysfunction in primary biliary cirrhosis. *World J Gastroenterol*. 2004; 10(20): 3039–3043, doi: [10.3748/wjg.v10.i20.3039](https://doi.org/10.3748/wjg.v10.i20.3039), indexed in Pubmed: [15378789](https://pubmed.ncbi.nlm.nih.gov/15378789/).
- Baratta L, Tubani L, Merli M, et al. Long-term effect of liver transplantation on cirrhotic autonomic cardiac dysfunction. *Dig Liver Dis*. 2010; 42(2): 131–136, doi: [10.1016/j.dld.2009.05.009](https://doi.org/10.1016/j.dld.2009.05.009), indexed in Pubmed: [19540819](https://pubmed.ncbi.nlm.nih.gov/19540819/).