# Medical Expertise

"Development of the European Network in Orphan Cardiovascular Diseases"
„Rozszerzenie Europejskiej Sieci Współpracy ds Sierocych Chorób Kardiologicznych"

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## CASE SUMMARY

The authors presented a case of a 25 years old male patient referred to the Centre for Rare Cardiovascular Diseases with a suspicion of pulmonary hypertension based on echocardiographic evaluation showing elevated right ventricular systolic pressure of 40 mmHg (25 + 15 mmHg). He complained of recurrent ankle edema. He declined limitation of exercise capacity. His past medical history revealed an episode of an unspecific leg muscle pain with concomitant transient increase of creatinine kinase in 2013. Consecutive neurological and rheumatological evaluation was with no abnormalities. He had a history of neutropenia and thrombocytopenia since childhood. 50% decrease of factor VII activity was diagnosed also in childhood. He remained under close follow-up of angiological evaluation due to the presence of stiff distention of inferior vena cava (IVC) of 22mm. No arterial or venous abnormalities of inferior body vessels were ever diagnosed on usg. Physical examination revealed marfan-like physiognomy with height of 197cm, astigmatism, arachnodactyly and cavoid feet. However, no diagnosis of marfan syndrome was made based on the Ghent criteria. He also had keloid lesions on the skin on chest and shoulders. His laboratory evaluation revealed low level of WBC of 2750. No other abnormalities were detected. Cardiopulmonary exercise test showed normal exercise tolerance. Echocardiography and CMR showed normal size and function cardiac chambers and valves, no pericardial effusions and dilated IVC of 20mm diameter, stiff. Cardiac catheterization revealed elevated cardiac output of 11 L/min, cardiac index of 4,6 l/min/$m^2$, elevated O2 saturations on the venous system up to 90% and no signs of pulmonary hypertention. No signs of fistulae were found on catheterization neither on angiography. Abdominal CT showed dilation of the IVC, left renal vein compression by the aorta and superior mesenteric artery, dilated in proximal part; Number of dilated, tortuous veins present (fistulae?/collateral circulation?) in the region of spleen, left kidney and pancreas; enlargement of spleen. The authors are uncertain whether the diagnosis of hypercinetic circulation is correct? Whether treatment with beta-blocker should be continued? And should selective left renal artery angiography be performed?

## DISCUSSION

Hyperkinetic circulation (HC) is characterized by an elevated resting cardiac index beyond the normal range of 2.5 to 4.0 L/min per m2, ineffective blood pressure, increased heat rate, and decreased systemic vascular resistance. There is a number of physiologic circumstances that can substantially increase cardiac output: excitement, anxiety, or stress; exercise; Pregnancy; Fever.

Pathologic conditions resulting in HC include arteriovenous fistulas (systemic arteriovenous dialysis fistula, hereditary hemorrhagic telangiectasia, hepatic hemangiomas, giant cutaneous hemangiomas) hyperthyroidism, anemia, beriberi (vitamin B1 or thiamine deficiency), dermatologic disorders (eg, psoriasis), renal diseases, hepatic diseases, skeletal disorders (eg, acromegaly, McCune-Albright syndrome, Paget disease, multiple myeloma), sepsis or carcinoid syndromes. In some circumstances, especially in patients with underlining cardiac diseases symptoms of heart failure may occure. This state of high-output heart failure is defined as elevated CO, decreased systemic vascular resistance and the presence of chronic activation of the sympathetic nervous system and renin-angiotensin-aldosterone axis, increased serum vasopressin (antidiuretic hormone) concentrations, and chronic volume overload gradually causing ventricular enlargement and left ventricular remodeling.

Typical findings of HC include warm extremities, wide pulse pressure, bounding pulses, a hyperkinetic heart to palpation, and a systolic flow murmur.

The diagnosis is made upon right heart catheterization.

Therapy is aimed at correcting the cause of HC.

## EXPERT'S OPINION

The diagnosis of HC in this patient made upon RHC seems to be correct. Further diagnosis of the cause of HC is necessary. Active search for arteriovenous fistulas is advised. Selective angiography of left renal vein should be performed. The patient should also be consulted with orthopedist, endocrinologist, hematologist and dermatologist. Genetic consultation may also be required. Treatment with betablockers may be continued, however close follow-up should be introduced.

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