

A stone was lifted from her heart: pericardial constriction in 28-year-old patient with Mulibrey nanism

„Kamień spadł jej z serca” — zaciskające zapalenie osierdzia u 28-letniej pacjentki z niskorosłością Mulibrey

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MuLiBrEy nanism (also known as Perheentupa syndrome) is an autosomal recessive congenital disorder. A patient with Perheentupa syndrome has growth retardation and multiple organ manifestations in muscles, liver, brain, and eyes. Over 80% of patients are Finnish. A regular component of this syndrome is pericardial constriction. A 28-year-old patient was firstly diagnosed with an ovarian tumour. Due to secondary liver failure caused by heart failure, she could not undergo gynaecological surgery. She was qualified to pericardiectomy as a first stage before gynaecological operation. On physical examination, the patient had 33.2 kg weight and 1.38 m height with body mass index 17.5 kg/m². Other symptoms included craniofacial dysmorphism, hepatomegaly (liver palpable 5 cm below the right costal arch), and heart failure corresponding to New York Heart Association class IV. Her coexisting diseases were: right ovary tumour, hypothyroidism, and hyperparathyroidism. Her treatment history included left ovariectomy in 2007, and right nephrectomy caused by a Wilms' tumour in 1988. Chest X-ray showed thin ribs and rounded heart with calcifications (Fig. 1). Spinal chest computed tomography showed pericardial calcification in anterior and inferior surfaces of the heart (Fig. 2). Transthoracic echocardiography performed in difficult conditions showed good left ventricle function with ejection fraction around 55%; the inferior vena cava was expanded to 20 mm as an effect of diastolic heart failure. The patient was qualified to pericardiectomy. During sternotomy calcified pericardium (Fig. 3) with deposits up to 8 mm thick was removed from the right ventricle, right atrium, vena cava superior, and front and lateral side of left ventricle (Fig. 4). A few small calcifications were left on the lower heart surface, due to their dangerous localisation. On the third postoperative day the patient developed symptoms of hepatic encephalopathy and kidney insufficiency. The patient was transferred for gastroenterological treatment. After seven days of treatment the patient went home with the recommendation systematic medical control. They are only 110 patients described as having this disease worldwide. This case is extremely rare but also extremely interesting. We did not find other case reports of Mulibrey nanism with pericardiectomy in Poland. This case demonstrates safe pericardiectomy in patients with Mulibrey nanism. We should bear in mind that liver and kidney complications are particularly common in patients with this syndrome.

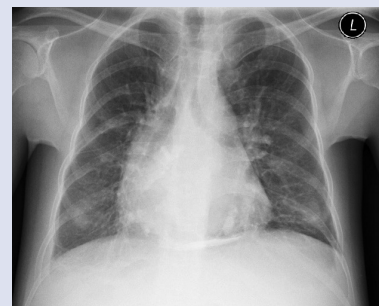


Figure 1. Chest X-ray shows thin ribs and rounded heart; arrow shows calcification



Figure 2. Chest computed tomography scan; arrow shows calcification in anterior and inferior surface

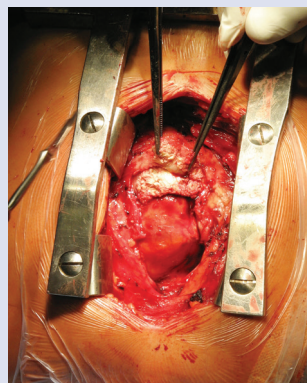


Figure 3. Pericardiectomy

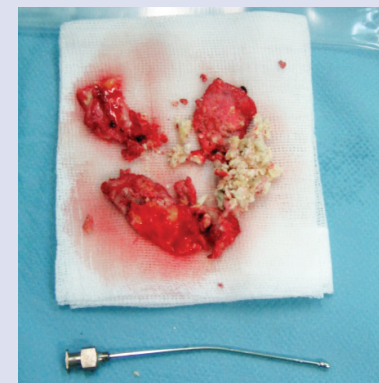


Figure 4. Removed calcified pericardium

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