Abnormal laboratory findings prior to the diagnosis of protein-losing enteropathy after single ventricle strategy palliation.

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Introduction: Protein-losing enteropathy (PLE) occurs in up to 15% of patients palliated surgically with single ventricle strategy and is associated with a 5-year survival rate of only 49-88%. Protein loss is often accompanied by lymphopenia, which may lead to development of serious infections that contribute to the overall mortality of PLE patients. Early and adequate diagnosis leads to earlier treatment implementation, which may prevent complications.

Aim of the study: Aim of the study was to assess if and to what extent abnormal laboratory findings may precede clinical diagnosis of protein-losing enteropathy.

Material and methods: Between 1993 and 2013 13 patients with PLE following Fontan palliation were diagnosed in Children’s Memorial Health Institute. The results of laboratory tests prior to the development of symptomatic PLE were available in 8 patients (50% male) with single ventricle (left in 4, right in 4). Data including serum protein level and lymphocyte count at the time of clinical diagnosis and during asymptomatic period was collected retrospectively. Progress of the disease was assessed using matched-pair analysis.

Results: Last procedure of surgical palliation was performed at the mean age of 5,1 years. Mean age at the time of PLE diagnosis was 8,9 years. In all patients abnormal laboratory findings suggesting a possible onset of PLE were found prior to the clinical symptoms - an average of 10,1 months earlier. The laboratory results were compared between both periods - prePLE and PLE: mean serum protein level decreased to 44,9g/l and 40,11g/l (NS), mean lymphocyte count was 1,5G/L vs 1,05G/L (p=0,045), mean lymphocytes percentage in WBC differential was 26,9% vs 19,4% (p=0,018). Lymphopenia (below normal for age) was found in 5 patients before and in 6 after PLE diagnosis.

Conclusions: Abnormal laboratory findings such as hypoproteinemia and lymphopenia may precede PLE diagnosis and suggest probable PLE onset. Collected data indicates increasing abnormalities during asymptomatic period, especially in the WBC differential. Therefore, in patients after Fontan palliation, we suggest considering performing common, low-cost blood tests - serum protein level and lymphocyte count - in order to seek possible onset of PLE and prevent complications.