C16. PULMONARY HIPERTENSION ASSOCIATED WITH MAJOR THALASSAEMIA.

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Although the quality of life of patients with major thalassemia and its duration were dramatically improved in recent years, cardiac complications still represent an important cause of morbidity and mortality. Pulmonary hypertension appears to be a relatively common complication in patients with thalassemia major, echocardiography monitoring revealed elevated pulmonary systolic pressure. The causes of pulmonary hypertension are not always known, the most important being associated with thalassemia: chronic hypoxia, hemolysis, iron deposits in the liver, heart and lung, increased tendency of microthrombosis and splenectomy.

Annual heart evaluations performed for patients with thalassemia major revealed several cases with different degrees of pulmonary hypertension, two of them severe, symptomatic by decreased exercise tolerance, dyspnea to small efforts, mild leg edema. Both patients were older than 40 years, associated chronic hepatitis C, secondary osteoporosis, hypogonadism, splenectomy and cholecystectomy, and inconsistent transfusion history with long periods without blood transfusions.

For one of the cases was performed cardiac catheterization to confirm the diagnosis and initiate specific vasodilator therapy with sildenafil, and oral anticoagulant therapy, which are yet well tolerated. The other case is pending confirmation of diagnosis and for both patients decided to intensify the transfusion regime and the chelation therapy supported.