The wide spectrum of β-thalassaemia intermedia induced pulmonary hypertension: The possible role of specific pulmonary arterial hypertension therapy

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Pulmonary hypertension (PH) has been reported as a frequent complication in patients with β-thalassaemia intermedia (TI). According to the 5th world symposium on PH, PH due to haemolytic anaemias is classified in group 5. The use of pulmonary arterial hypertension (PAH) specific therapy in these patients is based on data derived from other forms of PH, expert opinion, and small series or case reports.

We report 2 patient cases with TI and PH; the 1st patient was a 43-year-old female and the 2nd patient a 31-year-old female. Both patients had an history of splenectomy and had received occasionally blood transfusions.

The right heart catheterization (RHC) revealed pre-capillary PH (mPAP: 31mmHg, PAWP: 6mmHg and PVR: 3,6Wood Units for the 1st patient, mPAP: 46mmHg, PAWP: 9mmHg and PVR: 5,2Wood Units for the 2nd patient).The perfusion lung scan was without signs of Chronic Thromboembolic Pulmonary Hypertension.

We treated the 2 patients with PAH specific therapy (Bosentan) together with intensification of haemoglobinopathy-directed therapy (regular blood transfusions, chelation, hydroxyurea). Both patients presented significant clinical and haemodynamic improvement. For the 1st patient, we decided to stop Bosentan and continue only with the haemoglobinopathy-directed therapy. Two years later, she remains in NYHA-FC I and has normal haemodynamic parameters in the follow-up RHC. The 2nd patient decided on her own to discontinue Bosentan and maintain the transfusion program. One year later, her clinical status was severely deteriorated as well as the haemodynamics derived from the new RHC. Thereafter, double specific PAH therapy including Bosentan and Sildenafil added to her treatment, followed by clinical and haemodynamic improvement.

Reporting these two cases with the same initial approach and different outcome, we highlight the multifactorial mechanisms of PH development in patients with TI, the importance of optimal chelation transfusion therapy and the need of PAH specific therapy in some patients.