

## PW1-11

### **Circulating Endothelial Cell Levels Decrease after Vasodilator Therapy and are a Biomarker of Deterioration in Pediatric Pulmonary Hypertension.**

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Background : Pulmonary vasodilators in general and prostacyclin therapy in particular have markedly improved the outcome of patients with pulmonary arterial hypertension (PAH). Endothelial dysfunction is a key feature of PAH and we previously described that circulating endothelial cells (CECs) could be used as a biomarker of endothelial dysfunction in PAH. We now hypothesized that PAH-specific vasodilator therapy might decrease CEC numbers.

Methods: We quantified CECs in peripheral blood from children with idiopathic PAH (iPAH, n =30) or PAH secondary to congenital heart disease (PAH-CHD, n =30), before and after treatment and during follow up. CEC were enumerated by immunomagnetic separation with mAb CD146-coated beads.

Results: CEC counts were significantly decreased in children after treatment with oral endothelin antagonists and/or PDE5 inhibitors. In 10 children with refractory PAH despite combination oral therapies, SC treprostinil was added and we found a further significant decrease in CEC count during the first month of treatment in every patient. We quantified CEC during 6 to 36 months follow-up after initiation of SC treprostinil and found that CEC count is modified according to clinical status.

Conclusions: CEC counts fall with vasodilator therapy in PAH and could also be used as biomarker of deterioration in refractory pediatric pulmonary hypertension treated with SC treprostinil.