Introduction: the total cavopulmonary connection (TCPC) is the only surgical palliation in children with complex congenital heart defects and single ventricle physiology. The prerequisite for the passive pulmonary blood flow are low mean pulmonary artery pressure (MPAP) and pulmonary vascular resistance and preserved ventricular function. High risk patients prior to TCPC are those with MPAP >15mmHg. The use of selective pulmonary vasodilator drugs, such as phosphodiesterase-5-inhibitor Sildenafil, may lower the MPAP to acceptable value and make it possible to complete TCPC. The purpose of this retrospective study was to evaluate the effect of Sildenafil in patients with single ventricle physiology and elevated MPAP pre- and post TCPC.

Methods: from January 2011 to February 2014, 30 patients underwent TCPC. Eleven children with no morphological factors for elevated MPAP were treated with Sildenafil prior to TCPC. All had previous palliations in early infancy, 7 with pulmonary artery banding (PAB) and 4 with Blalock-Taussig shunt. All patients had hemodynamic study prior to TCPC. The indication for starting Sildenafil was MPAP >15mmHg measured invasively after Glenn procedure in ICU (4 patients) or during preoperative catheterization (7 patients). The treatment was maintained for 12±10months (3-34months) at oral doses of 2-3mg/kg/day. In 8 patients with persistently elevated MPAP or unfavorable hemodynamic data after TCPC, Sildenafil was continued. They had follow-up catheterization.

Results: patients with higher preoperative MPAP are those after PAB, elevated end-diastolic pressure and ventricular dysfunction. The Sildenafil therapy allowed TCPC completion in all. In the follow up postoperative catheterization, a significant decrease in MPAP (18 ± 1mmHg vs. 13±2mmHg, p=0,023) was found. An improvement in the ejection fraction (58 ± 3% vs. 67 ± 4, p=0,002) and ventricular end-diastolic pressure (VEDP) (13±3mmHg vs. 10±2 mmHg, p=0,048) was observed. Postoperative hemodynamic data in 3 patients allowed Sildenafil discontinuation. Satisfactory results were achieved after a minimum of 6 months Sildenafil treatment course duration. Minor adverse effects were observed in one patient, abolished after lowering the dose.

Conclusions: our preliminary result demonstrates that Sildenafil treatment improves hemodynamic parameters in high risk TCPC patients. A randomized trial can identify safety, effectiveness and long-term effect.