

Sildenafil for successful recruitment of lost pulmonary capillaries after Fontan surgery

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Introduction: The status of pulmonary circulation is essential for outcome after Fontan procedure. Elevated pulmonary pressure, vascular resistance, small size of pulmonary arteries and significant artery branch stenosis reflect some of the main problems.

Case report: A 5 year 8 months old boy (17,1 kg, 112 cm) with complex single ventricle morphology and coactation underwent fenestrated Fontan completion and stent implantation because of left pulmonary branch stenosis. 1 year later angiography and MRI revealed nearly no blood flow to the left lung with functional loss of capillary vascular structures. 4 months after initiation of oral Sildenafil therapy repeated MRI and angiography proved better hemodynamics, recovered lung perfusion and recruitment of the capillary vascular bed. There was significant clinical benefit.

Conclusion: Our case emphasizes that in case of functional loss of capillary vascular bed in non-pulsatile pulmonary perfusion recruitment of arteriolo-capillary bed is possible by oral treatment with PDE 5 inhibitor. Sildenafil was well tolerated without any adverse effects and caused significant hemodynamic and clinical improvement. It seems to be a safe and serious therapeutic option in case of functional loss of arteriolo-capillary bed.



Left: SVC-angiography with patent LPA stents but no flow to the left lung. Right: Recovered arteriolo-capillary bed after sildenafil treatment.