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Only Hepatic Venous Blood Closes Intrapulmonary Shunts after Cavopulmonary Connection

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Objectives: Intrapulmonary shunts in congenital heart disease lead to significant morbidity and mortality. The exact pathophysiology is still under discussion although the most favourable hypothesis involves the lack of an unknown liver factor in pulmonary perfusion. This study compares therapeutic options of intrapulmonary shunts in patients with single ventricle.
Methods: We retrospectively evaluated nine patients with intrapulmonary shunts and a functional single ventricle, four of them in combination with heterotaxy.
Results: All patients had surgical management by means of a Fontan operation or cavo-pulmonary connection. In each of the patients the hepatic venous blood did not reach the lung or was angiographically unequally distributed between the lungs. The side with diminished hepatic venous blood flow showed a more frequent appearance of shunts. After the Fontan operation or cavopulmonary connection the shunts evolved after different periods of time, ranging from months to several years. Patients treated only with pulmonal vasodilators (oxygen or sildenafil) showed no substantial increase in oxygen saturation. Of the four patients treated successfully, in two the Fontan circulation was completed by total cavopulmonary connection. One had already a Fontan circulation, but hepatic venous blood was unequally distributed due to stenosis of the left pulmonary artery which was managed successfully by stenting. And one patient needed pneumonectomy due to massive shunts in one lung.
Conclusion: The appearance of intrapulmonary shunts in congenital heart disease is linked to pulmonary perfusion without hepatic venous blood. The only causative therapy seems to be redirecting hepatic venous blood to the lungs. If this is not possible, other therapy options, such as selective pulmonary vasodilation, provide low chances of success.