Pulmonary arterial hypertension in cavo-pulmonary shunts

Dear Editor,

The bidirectional Gleen shunting (an anastomosis between the superior vena cava and the right pulmonary artery) and the Fontan procedure (an operation that results in the flow of systemic venous blood to the lungs without passing through a ventricle) are done to treat several complex cyanotic congenital heart abnormalities such as tricuspid atresia, pulmonary atresia with intact ventricular septum, hypoplastic left heart syndrome, and double-inlet ventricle. Pathophysiology of the Fontan circulation leads to an obligatory systemic venous hypertension in which the superior and inferior vena cava and the mean pulmonary artery pressure are equal and generally in the range of 10 to 20 mmHg, 2 to 4 times that of normal.

Currently suggested criteria are not to offer bidirectional Gleen shunting *to patients with pulmonary vascular resistance (PVR) higher than 3* Wood units *and/or a mean pulmonary arterial pressure (PAP) higher than 18-20 mmHg. Nonetheless, other authors* increase this limit to *a mean PAP of* 25-30 mmHg especially if it is associated with unrestricted and high pulmonary flow[1]. However a Fontan operation can only be considered in very well selected patients and strict selection gives the best early and late results, with an operative mortality <5%. These criteria include a low PVR, a mean PAP <15 mmHg, a preserved ventricular function, an adequate pulmonary artery size, no relevant atrioventricular valve regurgitation, and a normal rhythm[2]. In fact, we should treat, surgically or percutaneously, any pulmonary artery stenosis, atrioventricular valve regurgitation or major aortopulmonary collateral arteries, before performing a Fontan surgery.

Also, several pulmonary vasodilators are now available, and many turn one patient from non-candidate to candidate to a *cavo-pulmonary shunt*[3]. Moreover, we may use a combination of drugs (endothelin receptor antagonists, phosphodiesterase inhibitors, and prostanoids) as they use different pathways for decreasing PVR. However, we should take into account that a decrease in PVR may favor the surgical approach but conversely a relatively mild increase in PVR may result in a low cardiac output, protein losing enteropathy, pulmonary thromboembolism and arrhythmias. To this is added the fact that PVR after the Fontan operation may increase slowly over time[4] possibly due to micro-emboli from a dilated right atrium or from the venous system, aging, obstructed airways caused by lymphatic dysfunction or lack of pulsatile pulmonary flow which may release endothelium-derived vasoactive molecules. For this reason we must weigh potential benefits against potential side effects when considering a cavo-pulmonary shunt in a patient with a borderline PAP.

Given these data we should choose other options, besides the Gleen and the Fontan procedure, in patients with *high risk* for adverse events. Among them we may find the placement of systemic-to-pulmonary shunts, although the benefit of increased pulmonary blood flow should be weighed against the risk of increased volume load to the systemic ventricle, heart/lung transplantation, or the initiation of pulmonary arterial hypertension treatment in order to improve exercise capacity[5]. Despite this, the longer-term effects of chronic use of phosphodiesterase type 5 inhibitors and endothelin-1 receptor antagonists in the Fontan population are not known.

Best regards,

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