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Cardiac and pulmonary physiology in the functionally univentricular circulation with reference to the total cavo-pulmonary connection

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CARDIAC AND PULMONARY PHYSIOLOGY IN patients with functionally univentricular hearts is markedly different from that found in the setting of the normal circulation. The physiology of both the heart and the lungs remains abnormal after palliative procedures, such as creation of systemic-to-pulmonary shunts, banding of the pulmonary trunk, or creation of the cavo-pulmonary connections. In this brief review, I will concentrate on two main aspects of physiology, namely oxygenation and haemodynamics. I will make comparisons between the normal circulation and the functionally univentricular circulation, the latter both before and after creation of a cavo-pulmonary connection. To finish, I will discuss the physiology of the cavo-pulmonary connection itself, and consider some clinical implications.

Oxygenation

The normal circulation is made up of two separate circuits in series. This arrangement produces optimal oxygenation, since all the deoxygenated blood returning through the caval veins is redirected to the lung, with none entering the arterial circulation. In other words, there are no shunts in the normal circulation. At rest, the arterial oxygen saturation is usually near to 100%, while the caval venous saturation is around 70%, the arterio-venous difference usually being in the order of 30%. This leaves considerable reserve for the oxygenation of tissues, either by increasing cardiac output or by additional extraction of oxygen. This relationship can be expressed through the Fick formula, which applies both for consumption of oxygen and capture of oxygen in the lungs, features that are equal on a steady state. Thus, consumption of oxygen is equal to the systemic flow multiplied by the arterio-venous difference in content of oxygen. The same consumption of oxygen can also be expressed as the flow of blood through the lungs multipled by the difference in content of oxygen between the pulmonary veins and the pulmonary arteries.

In the normal heart, the saturations of oxygen are the same in the aorta and the pulmonary veins, since there is no shunting from right-to-left. They are also the same in the pulmonary arteries and the mixed return from the caval veins, since there is no shunting from left-to-right. The flows in the systemic and pulmonary circuits are also equal, confirming that there is no shunt, and that the circulations are in series.

In the patient with a functionally univentricular heart, there is usually complete mixing of blood before arterial ejection. Due to this, the saturations of oxygen are equal in the aorta and in the pulmonary arteries. If the difference in systemic saturation of oxygen from normal is about 30%, and if the pulmonary venous saturation of oxygen is near to normal, at 100%, the relationship between the arterial saturation and the ratio between the flows to the lungs and the systemic circuit can be calculated as:

Aortic saturation of oxygen
= 100 − 30: lung/systemic flow ratio

Analysis of this relationship shows that a reasonable saturation of oxygen cannot be achieved without imposing a significant volume overload on the heart

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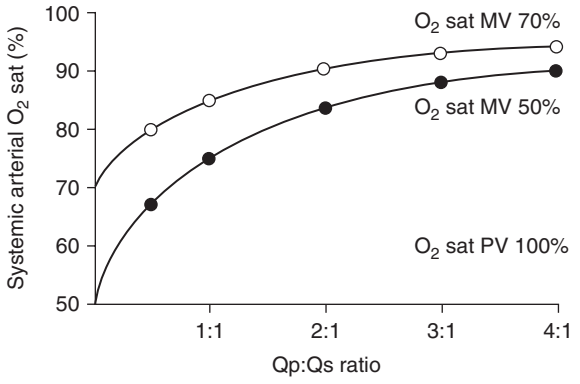

Figure 1.
Relationships between flows and arterial saturations in the patient with a functionally single ventricle, modified from the original concept of Rudolph. MV: mitral valve; PV: pulmonary valve; Qp: pulmonary flow; Qs: systemic flow; O_2 sat: oxygen saturation.

(Fig. 1). In order to achieve an arterial saturation of around 90%, the ventricular mass in the setting of the functionally univentricular arrangement needs to eject three times more blood to the lungs than to the systemic circuit. This is equivalent to a regurgitation fraction of around 65% for aortic or mitral valvar insufficiency.

Haemodynamics

In the normal circulation, it is possible to achieve different conditions of loading for the two ventricles. Indeed, in the normal circulation, the pressure is higher in systole and diastole in the left compared to the right heart. This is because of the major differences between the lungs, which have low vascular resistances, and the systemic vessels, with their high vascular resistances. It is also because of the different properties of the left ventricle, with its good contractility, and the right ventricle, with its good compliance.

I emphasize the great benefit of a circulation being able to have a higher preload in the left compared to the right ventricle, because the hydrostatic pressure threshold for oedema is much higher in the pulmonary capillaries, at up to 25 millimetres of mercury, compared to the caval venous capillaries, with the threshold at less than 10 millimetres of mercury. The independence of the conditions of loading permit increases in aortic pressure or cardiac output without exerting any significant increase in the caval venous pressures. The effect on the right heart is indeed minimal, since the increase in cardiac output has little effect on the right heart. There is a minimal rise in the caval venous pressure, because the right ventricle is very compliant, and there is also limited rise in the pulmonary arterial pressure, since the pulmonary arterial bed has the ability to decrease vascular resistance by recruitment of vessels together with passive vasodilation. Thus, there is no increase in the transpulmonary pressure gradient, and the increase in pulmonary arterial pressure is only a repercussion of the increase in capillary pressure due to the higher preload on the left ventricle.

In the patient with a functionally univentricular heart, as in those with large atrial or ventricular septal defects, the diastolic pressures are usually identical in both ventricles and atriums, unless there are abnormal atrioventricular valves and an intact atrial septum. And it is impossible, therefore, to increase the pulmonary venous or capillary pressure, because this is limited by the identical increase in the caval venous pressures. There is some protection against the development of pulmonary oedema, but a great restriction in the ability to increase the systemic output. In the normal circulation, the possibility of having a relatively high pressure in the pulmonary capillaries also provides a higher pressure in the pulmonary arteries. This allows perfusion of the upper part of the lungs, since the pulmonary arterial pressure is the addition of the capillary pressure and the pressure gradient across the pulmonary vasculature.

Determinants of pulmonary arterial pressure in the functionally single ventricle

As explained above, in the normal circulation, with the circuits in series, the pulmonary arterial pressure is the sum of the pressure gradient between the pulmonary arteries and veins and the capillary pressure, the latter being the repercussion of the left ventricular diastolic pressure. The flow through the lungs is equal to the cardiac output, and the pressure gradient is the flow multiplied by the pulmonary vascular resistances.

In a patient with a functionally single ventricle, the level of the pulmonary arterial pressure, particularly the systolic pressure, is the resultant of an eventual obstacle between the ventricular mass and the arterial trunks. When the systolic pressure is compared between the pulmonary arteries and the aorta, the systolic pulmonary arterial pressure is equal to the systemic aortic pressure in the absence of any stenoses. It is infrasystemic should there be pulmonary stenosis, but suprasystemic when there is aortic stenosis. The diastolic and mean pressures in the pulmonary arteries also depend on the pulmonary vascular resistances and pulmonary capillary pressure. Most importantly, the flow of blood to the lungs will depend on the ratio of the mean pulmonary arterial pressure gradient through the lung divided by the pulmonary arterial resistances.

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In other words, in a normal circulation, the pulmonary flow is fixed, and is the same as the cardiac output, while the pulmonary arterial pressure is variable.

In the patient with a functionally univentricular heart, in contrast, it is the pressure that is relatively fixed, depending of course on the specific anatomy, and the flow that changes, this depending on the ventricular pressure and vascular resistances.

Loading conditions and ischaemic threshold

The loading conditions of the dominant ventricle in a functionally univentricular heart can vary markedly, depending on the anatomy, and particularly on the presence or absence of pulmonary or aortic stenosis, atrioventricular valvar anomalies permitting regurgitation or stenosis, or atresia, and the integrity of the atrial septum; this latter feature being particularly important should the atrioventricular valves be abnormal.

I explained in the previous paragraphs the obligatory increase in preload that is needed to achieve acceptable arterial saturations. This increase in preload is accompanied by an increase in afterload, even if there is no increase in ventricular pressure, simply because the volume overload produces an increase in wall stress, which is equal to the product of pressure and volume. Due to the increase in ventricular systolic pressure, this increase in afterload is augmented by arterial hypertension or subaortic stenosis.

Along with the increase in work, there is an increase in the mass of the functionally univentricular heart in order to reduce the wall stress per gram of tissue. This is determined according to the Laplace law, which states that wall stress is equal to the product of pressure and diameter, but divided by mural thickness. This increase in mass affects the ischaemic threshold of the ventricular myocardium.

The potential myocardial ischaemia produced by the increase work and ventricular mass, implying an increase in the myocardial consumption of oxygen, is exacerbated by a decrease in the delivery of oxygen through the coronary arterial bed due to a decrease in the coronary arterial diastolic pressure gradient, this being the difference between the coronary arterial diastolic pressure and the diastolic ventricular pressure. This is particularly true in the setting of systemic shunts, or aortic regurgitation with a low diastolic aortic pressure and a high diastolic pressure in the dominant ventricle due to overloading, this combination eventually leading to myocardial dysfunction. In the setting of concomitant ischaemia, this then produces a vicious circle, with the ischaemia increasing the end-diastolic pressure and decreasing coronary arterial perfusion because of the lower pressure gradient for perfusion. In addition, coronary perfusion itself may be impaired by coronary arterial stenoses, as can occur in aortic atresia with retrograde coronary arterial flow through the long and narrow ascending aorta.

Hence, we can see that, because of the frequent prevailing anatomical and physiological conditions, the patient with a functionally univentricular heart has to contend with a combination of increased preload, the afterload challenge of increased myocardial mass, and additional potential myocardial ischaemia. The patient with complete mixing of the venous returns, therefore, is at a very high risk of deterioration. Such patients should not be left too long with this unfavourable physiology. There is, therefore, a solid physiological basis for creation of a cavo-pulmonary circulation, so as to bring desaturated blood directly to the lungs, thus increasing oxygenation without loading the functionally univentricular mass.

What about the cavo-pulmonary circulation in terms of its physiology?

The main consequence of creation of a cavo-pulmonary connection is the obligatory rise in venous pressure, at least to the level of the pulmonary arterial pressure. Although it is often argued that the venous bed has a high compliance, this is not true. There is certainly a high capacitance within the venous bed, but as soon as the veins are full, and there is stretching of their walls, their compliance is very low. In this setting, increase pressure leads very quickly to oedema.

The consequences of the increases in pressure, however, are different for the superior and inferior caval venous territories. In the territory drained by the superior caval vein, the hypertension is well tolerated. It does not affect the flow of blood to the brain, or result in interstitial oedema, unless the pressure is very high. It does, however, impact on the lymphatic drainage, leading eventually to chylothorax. A second problem is the creation of veno-venous fistulas between the high pressure territory draining to the superior caval vein, and the low pressure territory drained through the inferior caval vein, even after ligation of the azygos vein. Should venous hypertension develop in the territories drained by the inferior caval vein, is not well tolerated. It leads to hepatic pain and dysfunction, and portal hypertension, with the end-stage being an exudative enteropathy and enormous increase in production of lymph.

Another consequence of creation of a cavo-pulmonary circulation is that flow through the lungs is now non-pulsatile, and occurs at low pressures. This situation means that it is difficult to recruit pulmonary vessels at the apexes of the lungs, the

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flow does not dilate the arterioles, and the arrangement may facilitate the creation of pulmonary arterio-venous fistulas, especially if there is no flow from the liver to the lungs, as can occur in patients with left isomerism and interruption of the inferior caval vein.

And, because the entire flow has to pass through the lung without the driving force of a ventricle, there is the danger of the dominant left ventricle having inadequate preload should a total cavo-pulmonary connection be created without a fenestration. The limiting factor for cardiac output then becomes the amount of blood that crosses the lung without any propulsion apart from ventilation. In Paris, we have identified what we call the Fontan paradox. This is the fact that normal physiology requires pulmonary arterial pressures above 15 millimetres of mercury, and caval venous pressures below 10 millimetres of mercury. In the cavo-pulmonary circulation, however, the caval venous pressure is equal or above the pulmonary arterial pressure.

Clinical experience, nonetheless, shows that the haemodynamic situation can be acceptable, at least in the middle term, when caval venous and pulmonary arterial pressures are between 10 and 15 millimetres of mercury. The anticipated level of the pulmonary arterial pressure subsequent to creation of the cavo-pulmonary connection, therefore, should be the key factor in determining whether or not this type of surgery is contraindicated. Predicting the likely pulmonary arterial pressure after creation of a cavo-pulmonary shunt, however, is very difficult because of the dramatic change in the physiology between the two types of circulations. The pulmonary arterial pressure subsequent to creation of the cavo-pulmonary connection, as with circulations in series, is the sum of the capillary lung pressure, itself directly dependent on the diastolic pressure in the functionally univentricular mass, and the pressure gradient across the lungs, this depending on the new pulmonary vascular resistances and the cardiac output. Unfortunately, the dramatic change in loading conditions from a functionally univentricular situation to a cavo-pulmonary circulation affects considerably the pulmonary vascular resistances, which are much higher after creation of the cavo-pulmonary circulation because there is less vascular recruitment and less passive and active vasodilatation, meaning that there is less pressure and no pulsatility to dilate the arterioles. It also affects the lung capillary pressure in the pulmonary parenchyma through the ventricular diastolic pressure, the latter decreasing because of the reduction in flow. The decrease is difficult to quantify, since the compliance of the ventricle changes due to the acute change in the thickness of its walls. Furthermore, the curve for diastolic compliance is not linear but exponential. The errors inherent in the estimation are enormous in the patient with a very overloaded functionally univentricular heart. They are much less should the ventricle have been unloaded by significant pulmonary stenosis, or if the initial palliation has been achieved by creation of a partial cavo-pulmonary circulation.

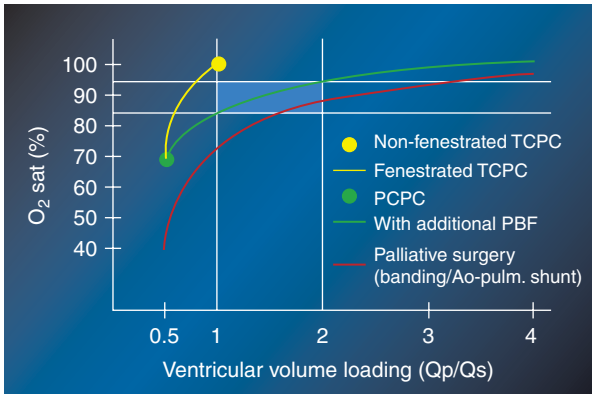

Figure 2.
Cartoon showing the relationships between the saturations of oxygen and the volume load exerted on the functionally univentricular heart by different palliative procedures. TCPC: total cavo-pulmonary connection; PCPC: partial cavo-pulmonary connection; PBF: pulmonary blood flow; Ao-pulm.: aorto-pulmonary; O_2 sat: oxygen saturation.

Nowadays, therefore, most centres do not advocate converting the patient with a functionally univentricular heart directly to the total cavo-pulmonary circulation. Instead, most recommend a transitional approach with a partial cavo-pulmonary circulation. The partial connection can be either by first creating the partial cavo-pulmonary circulation without any other flow of blood to the lungs that will preserve volume load to the heart. This approach will require early totalization of the cavo-pulmonary circulation because of cyanosis. The alternative is to provide an additional source of flow of blood to the lungs in addition to the partial cavo-pulmonary circulation. This option creates more overload but less cyanosis. This additional source of flow may come either directly from the functionally univentricular heart through a stenotic or banded pulmonary trunk, or through a systemic-to-pulmonary arterial shunt. In Figure 2, I have shown the saturations of oxygen, and the volume load exerted on the functionally univentricular heart, by different palliative procedures.

In Paris, we often leave this mixed circulation, made up of the cavo-pulmonary shunt together with direct flow, in place for quite some time, since the end result can be an acceptable level of cyanosis with no volume overload. We believe that this situation may "delay the clock" of a total cavo-pulmonary circulation or even be better for some patients as the

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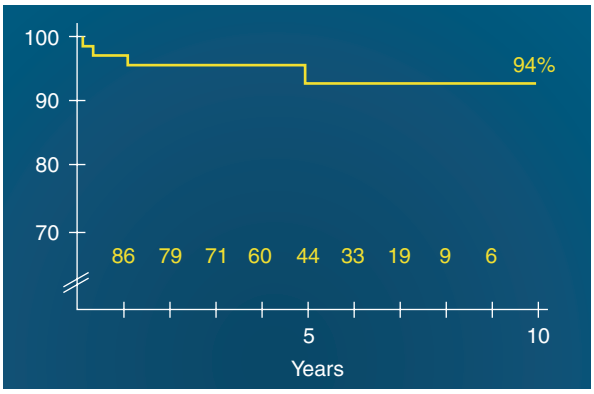

Figure 3.
The graph shows the actuarial survival for patients in Paris palliated using a cavo-pulmonary shunt in combination with direct flow of blood to the lungs.

definitive form of palliation, this notion being born out by our results in the middle term (Fig. 3). In closing, I reiterate the question as posed during the first workshop.[1] Which is worst, to be pink without a motor, like riding a bicycle, or to stay a little blue but with a motor, as driving on a motorcycle?

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