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Original Article

The partial cavo-pulmonary circulation with an additional source of pulmonary flow. An alternative to the total cavo-pulmonary circulation in patients with a functionally single ventricle

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MOST PAEDIATRIC CARDIOLOGISTS THINK THAT the optimal treatment for patients with functionally single ventricle is the relatively early construction, at from 2 to 7 years of age, of the total cavo-pulmonary circulation. This approach is based on the desire to produce an acyanotic child, such an outcome fostering the belief that the patient is cured. When using this therapeutic approach, the total connection is usually preceded by earlier creation of a partial connection, achieved by creating a bi-directional anastomosis between the superior caval vein and the right pulmonary artery at the age of from 3 to 9 months, without providing additional flow of blood to the lungs. This leads to severe cyanosis by the age of 3 years, when the flow increases in the inferior caval vein, and is the signal for early completion of the total connection. In this policy, the absence of additional flow to the lungs is justified by the fear of altering the ventricular function essential for the success of the total connection, because of the volume, and eventually the pressure, overload of the additional flow on the functionally single ventricle,which may impair its compliance and/or contractility, or be responsible for atrioventricular valvar regurgitation. These alterations in myocardial functions would then contraindicate, or increase, the mortality and morbidity of the total connection, producing deterioration in its long-term results.

Our approach in Paris is entirely different, and is based on the hypothesis that the non-physiologic circulatory pattern of the total connection has, in itself, a progressive deterioration over the years that may be worse than the moderate cyanosis and volume overload achieved in the setting of a well-calibrated partial connection supplemented with additional flow. We believe that, by avoiding or postponing as long as possible the total connection, we will delay the morbidity of this type of circulation. In other words, "setting the clock" as late as possible will improve the long-term outcome of these patients. We believe that the total connection should not be considered a therapeutic option, other than one of last resort just before cardiac transplantation.

In this review, I will justify our concept mainly on theoretical considerations, since we, as others, lack long-term follow-up with the alternative approaches. To our knowledge, no one has yet produced a scientific randomised study comparing the use of these two different therapeutic approaches in patients with functionally single ventricles. I will, however, discuss some mid-term results that support our policy and philosophy.

Theoretical considerations

Relationship between volume overload and saturation in the functionally single ventricle

In Figure 1, borrowed from Abe Rudolph, I show the relationship between the combined systemic and pulmonary blood flow, the cardiac output of the functionally single ventricle, and the systemic saturation at rest in a palliated patient, with complete mixing of pulmonary and systemic venous return at two levels of systemic arterio-venous differences. In this situation, a good arterial saturation can only be achieved by placing a significant volume load on the functionally single ventricle.

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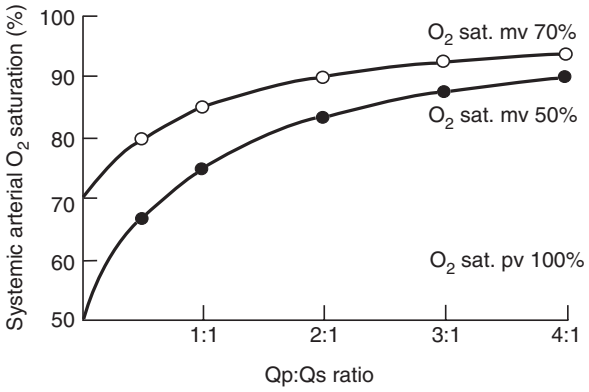

Figure 1.
Relationships between flows and cardiac output in the patient with a functionally single ventricle – modified from the original concept of Rudolph. mv: mitral valve; pv: pulmonary vein; Q_p: pulmonary flow; Q_s: systemic flow.

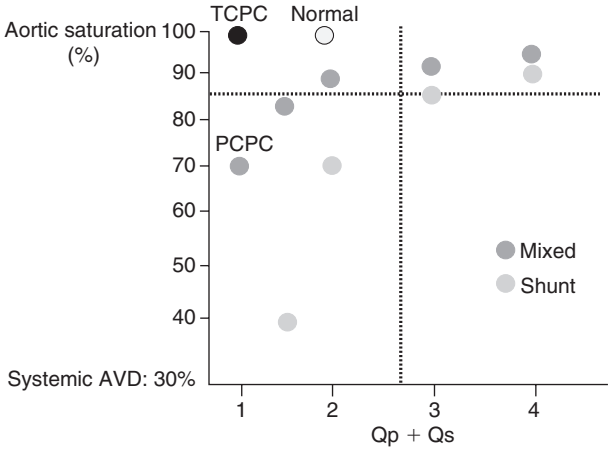

Figure 2.
The concept of palliation of the patient with the functionally univentricular heart. TCPC: total cavopulmonary connection; PCPC: partial cavopulmonary connection; AVD: arteriovenous difference; Q_p: pulmonary flow; Q_s: systemic flow.

In Figure 2, I show a theoretical representation of a different form of palliation based on the Fick principle, assuming that the pulmonary venous saturation is 100%, that the arteriovenous difference in saturations of oxygen through the tissues is 30%, and that the ratio of return from the superior and inferior caval veins is unity. Looking at this figure, we see that, if we accept a systemic saturation between 85% and 90%, one that is usually sufficient to avoid polycythemia and hyperviscosity and which allows a decent "intellectual" life, there is no volume overload in the situation of a partial connection with an added flow, whereas this was not possible in the situation of complete mixing.

Adverse effects of the total cavo-pulmonary connection

Although the total connection is a very efficient circulation for haematosis, since the volume load to the ventricle is minimal for perfect oxygenation, it is also a very pathological circulation, particularly when looking at the pulmonary, caval venous, and lymphatic circulations. It is also pathological for the systemic circulation, especially when considering the preload of the functionally single ventricle.

Optimal pulmonary circulation can be achieved by pulsating blood with a mean pressure of at least 15 millimetres of mercury in order to recruit the upper parts of the lungs. Non-pulsatile low pressure will concentrate the flow of blood to the bases of the lungs and, in the long term, it may damage the entire vasculature, leaving very little reserve in the face of lowering pulmonary vascular resistances. It may also, in the long term, facilitate the development of arteriovenous fistulas. We all know that cardiac transplantation in the setting of the Fontan circulation is a difficult situation for the donor right ventricle, although the pulmonary arterial pressure is relatively low, and that the pulmonary circulation stays abnormal in these situations.

Caval venous pressures should be low for proper functioning of the organs, ideally at values well below 15 millimetres of mercury, and probably below 10 millimetres. This is especially true for the organs draining via the inferior caval vein, and even more for those draining via the portal vein. Indeed, the liver and gut are very sensitive to increased venous pressures, with a major increase in production of lymph and eventual major oedema, or even vascular damage and protein-losing entheropathy. In the areas drained by the superior caval vein, because of the protected cerebral circulation due to auto-regulation of flow, the consequences are minor except for the impairment of lymphatic drainage, since this is also through the superior caval vein. It is, however, mandatory with a partial connection to ligate the azygos vein in order to avoid drainage from the superior to the inferior caval vein, instead of to the right pulmonary artery. It is not uncommon, nonetheless, that veno-venous communications develop through other pathways between the high pressure territories drained by the superior caval vein and the low pressure territories drained by the inferior caval vein. Lymphatic circulation in the setting of the total connection is characterised by the combination of an increased production of lymph due to inferior caval venous hypertension, and difficulties in drainage due to superior caval venous hypertension. It is likely that this association is very deleterious on the lymphatic circulation in many organs.

Preload of the functionally single ventricle in the setting of the total connection relies on the flow of

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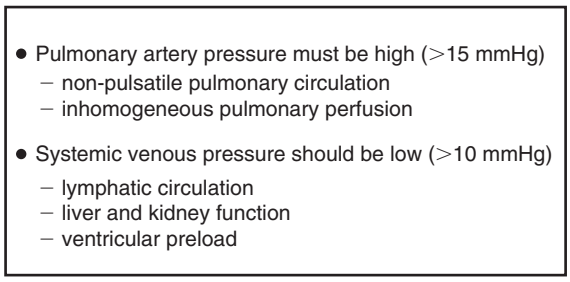

Figure 3.
The Fontan paradox as seen in patients with functionally univentricular heart.

blood passing through the lungs as a glider, with no motor from the right ventricle, pushed by respiration and sucked by the active relaxation of the ventricle. This situation is obviously very threatening regarding arrhythmias, myocardial, or pulmonary events, and is life-threatening due to low output and hypotension. We call it the Fontan paradox (Fig. 3). It is impossible in patients with a total cavopulmonary connection to achieve a physiological perfusion of the lungs, which requires pressures above 15 millimetres of mercury, with a physiological venous circulation requiring low inferior caval venous pressures below 10 millimetres of mercury, since in this situation inferior caval venous pressure is, at the best, equal to the pressures in the pulmonary arteries.

Clinical facts

We have limited experience of patients with a total cavo-pulmonary connection. From the literature, nonetheless, we know that deterioration of clinical state with time is well demonstrated for the Fontan circulation. In case of failures, the only possibility is heart transplantation, and these patients are far from ideal candidates for this option. It is possible and likely, however, that the total cavopulmonary connection is better than the classical Fontan operation, and this has been our experience during the last years, with better haemodynamics and less arrhythmias.

Our peri-operative mortality for the Fontan procedure in the 1980s, in 80 patients, was around 10%, with a late mortality or transplantation also of around 10%. There was also high morbidity, with 15% of patients having severe arrhythmias, 12% needing reoperation or interventional catheterisation for leak or obstruction, 2% having cerebral damage, and 1 patient having protein losing enteropathy. In those with a total cavopulmonary connection performed as a primary procedure not as a conversion from a failing Fontan, our mortality was 4% for a total of 25 patients, but the morbidity was still high over a short period of follow-up. Our policy now is systematically

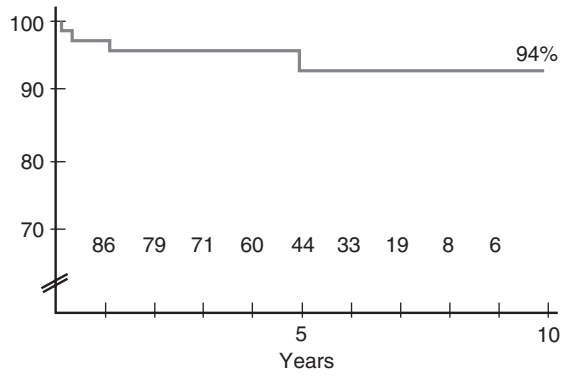

Figure 4.
Results in Paris with the bi-directional cavopulmonary anastomosis + arterial pulmonary blood flow/potential Fontan candidates: actuarial survival.

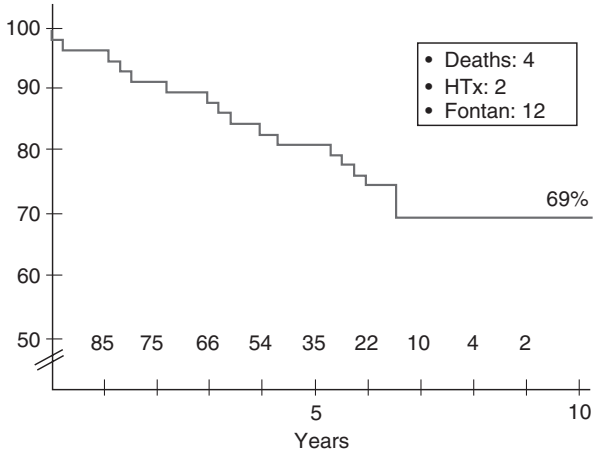

Figure 5.
Results in Paris with the bi-directional cavopulmonary anastomosis + arterial pulmonary blood flow/potential Fontan candidates: actuarial survival with bi-directional cavopulmonary anastomosis + arterial pulmonary blood flow.

to convert those with a classical Fontan procedure to a total cavopulmonary connection, but the results with the failing Fontan were not very good.

The partial cavopulmonary connection with an additional source of pulmonary flow

We have much more experience with this combination, albeit that it has not yet been published in the peer-reviewed literature. In the 1990s, 104 patients, including 14 with a contra-indication for a total cavopulmonary connection, have been palliated with a partial connection combined with an added source of pulmonary flow. The results are shown in Figures 4 and 5. This strategy has a low peri-operative mortality of 3%, a very low late mortality of 1%, and very low

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	BCPA with APBF	TCPC
Late mortality	–	1 (4.3%)
Normal EKG	16 (94%)	16 (70%)
Significant arrhythmia	–	3 (13%)
Chronic medication	2 (12%)	11 (48%)

Figure 6.
Late clinical results in our patients with functionally univentricular hearts, comparing the bi-directional cavopulmonary anastomosis (BCPA) with arterial pulmonary blood flow (APBF) against the total cavopulmonary connection (TCPC).

	BCPA with APBF	TCPC	
Peak exercise (W)	91 ± 17	103 ± 15	NS
O_2 sat. (%) at rest	84 ± 4	93 ± 4	$p < 0.001$
O_2 sat. (%) at peak exercise	64 ± 8	85 ± 6	$p < 0.001$

Figure 7.
Findings following exercise testing in patients with functionally univentricular hearts, comparing the results of the bi-directional cavopulmonary anastomosis (BCPA) with arterial pulmonary blood flow (APBF) against the total cavopulmonary connection (TCPC).

morbidity compared to the results using the total cavopulmonary connection (Fig. 6).

The functional status of the patients was acceptable postoperatively, with aortic saturations around 85%, and exercise limitation not significantly different from patients with the Fontan circulation (Fig. 7). There was no evident deterioration of ventricular function, or development of atrioventricular regurgitation, except for two patients with a loose band on the pulmonary trunk, with high superior caval venous pressures, who underwent reoperation immediately. The real difficulty with these patients is the calibration over the longer term of the extra flow. We have the choice in these patients between adapting the flow, usually by constructing a systemic shunt, or performing a relatively late total cavopulmonary connection. This last option has been our

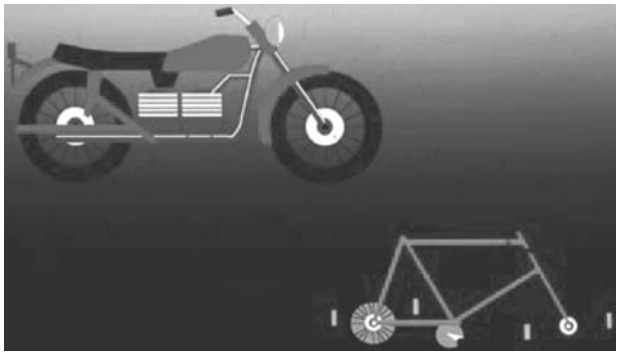

Figure 8.
The analogy between the partial and total cavopulmonary connections in terms of whether or not there is a motor under the lungs. Is it better to have a blue motorcycle or a pink bicycle?

favoured one of late, because of the acceptable mid-term results of the total cavo-pulmonary connection.

With this strategy, we are currently performing a total cavopulmonary connection in about one-third of our children around puberty, when the added flow becomes insufficient for adequate saturation. At this age, we have had no problems with ventricular function. We do not know if the other patients, who are well calibrated and not complaining after full growth, will remain good candidates for a total connection in the long term, or if they will be candidates for a long stable life or for late cardiac transplantation.

In conclusion, we feel that, since there is no ideal situation for patients having a functionally single ventricle because of the very hazardous long-term prognosis of the total cavopulmonary connection, it might be more efficient to have mild cyanosis with a motor for the pulmonary circulation in the fashion of a blue motorcycle, instead of a having a patient without a motor under the lung, who is a pink bicycle (Fig. 8). We feel that the partial cavopulmonary connection with an additional source of pulmonary flow gives good results in the mid-term, and does not impair the results of an eventual total cavopulmonary connection performed later in life. This strategy may well improve the long-term prognosis by avoiding or postponing for many years the non-physiological circulation of the total cavopulmonary connection.

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