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Anatomic substrates for, and function of, the functionally univentricular circulation before and after surgical procedures

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WHATEVER THE SPECIFIC ANATOMY, THE Fontan circulation, be it created directly or subsequent to a bi-directional cavopulmonary anastomosis, transforms completely the pattern of circulation of the blood. In essence, it converts a network of circulations in parallel into one in series. The haemodynamic consequences are numerous. The most important and direct among them is, perhaps, the increase in afterload, as well as the reduction in preload, for the systemic ventricle. From the clinical point of view, the most immediate and relevant implication is the amelioration of cyanosis, this being the consequence of removing the common mixing of systemic and pulmonary venous blood within the heart.

In our first International Workshop,[1] entitled "The Univentricular Circulation: From Analysis of Fluid Dynamics to Surgical Procedures", held in Massa over the period 25 and 26 October 2002, we discussed the optimisation of the surgical results by application of experiments and techniques of computational modelling related to the functionally univentricular circulation. Those studies were mainly devoted to understanding, and possibly reducing, the losses of energy at the newly created surgical anastomoses. Achievement of these aims was anticipated to be the first step in optimising the flow of blood to the lungs in patients lacking a subpulmonary ventricle, and hence unsuitable for direct surgical creation of circulations in series.

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In our second International Workshop, held in 2004, again in Massa, we aimed to investigate the changes in the cardiac function when parallel circulations were changed to circulation in series, and the consequences of changes in preload and afterload on the size and shape of the heart. The progressive ventricular dilation, and the increasing tension of the walls of the heart, produced by the volume overload that exists prior to creation of the Fontan circulation, and the subsequent abrupt decrease that occurs after the surgical procedures, together with the increase in the ratio of mass-to-volume for the ventricles, are well-known phenomena. Their implications were already discussed in part during the First International Workshop, published as a supplement to the Journal in 2004.[1]

Two years later, the investigational approaches based on accurate in vitro experiments and computer simulations still suffer from major limitations. We have now come to acknowledge the capabilities of computer simulations in providing clinicians with quantitative data that permits very sophisticated comparison of the various surgical procedures. Work is still in progress, nonetheless, to extend their predictive capabilities to function as a tool for surgical planning and decision-making. But serious difficulties remain in modelling the complete range of feedbacks that occur in the clinical situation, both in physiological conditions and after surgical repair of complex malformations. Although computational models are available which include, for instance, the effects of the Starling law of the heart, of gravity on the venous system, and the baroceptive regulation,

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phenomenons like adaptation of the heart and the vasculature to abrupt changes in the flow of blood and patterns of pressure are still difficult to model. This mainly stems from the fact that we do not possess mathematical or quantitative relationships between the haemodynamic factors and the changes in the vasculature, and the size and shape of the heart that occur over time. This deficiency remains at the time of our second International Workshop, despite the continuing efforts of our joint research teams, which include physiopathologists, cardiologists, cardiac surgeons, and bioengineers.

In addition, the concept of remodelling itself, as established in adult cardiac pathology and surgery, may have a completely different meaning in paediatric cardiac surgery. Our patients are growing up, and even their normal hearts are undergoing appropriate increases in size. Such changes are required because of the need for a more powerful pump to cope with the increasing cardiac output in the face of increasing body weight and surface area. During this period, the heart exhibits extraordinary and unexpected capabilities in reproduction of cells and myocardial vascularisation.

The anatomico-structural changes seen in patients with the functionally univentricular circulation do not differ much from those in normal children of the same weight and age. These anatomico-functional changes, therefore, are probably better addressed as ventricular modelling, implying a normal evolutionary process, while reserving remodelling for description of those processes identified subsequent to pathological situations, or events induced by surgical procedures themselves. In this era of rapid evolution of diagnostic and surgical procedures, surgical timing has relevant and substantial effects on cardiac modelling and remodelling in determining the cardio-respiratory function and prognosis of these patients. It is our hope, therefore, that the multidisciplinary approach of our second International Workshop, now published in this second supplement, will help us better to understand one of the most difficult aspects of the functionally univentricular circulation. In closing our introduction, we express our deepest gratitude to Elisabetta Donnini, Roberto Bertolini, Giuliano Kraft, and Tiziano Carducci, for all their help in organising the workshop. We are also indebted to Fatima Bongiorni, who worked over and above the call of duty in collecting and revising the articles included within this supplement.

Reference

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