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Ventricular dysfunction of the functionally univentricular heart: management and outcomes

Marc Gewillig

Gasthuisberg University Hospital, University of Leuven, Leuven, Belgium

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THE VENTRICLE OF A FUNCTIONALLY univentricular heart is dilated, hypertrophic, and hypocontractile, certainly after the completion of a Fontan circulation (reviewed in Reference[1]). It is difficult, however, to determine the reasons for these abnormal ventricular conditions. The abnormal size of the ventricle, along with the less than adequate function, can be due to the congenital malformation itself, the previous surgical conditions, or the very abnormal working conditions of the ventricle during the various stages of palliation. Moreover, because of the varying morphology of the pumping ventricle in patients with a functionally univentricular arrangement, it is difficult to assess size, shape, hypertrophy, stress, strain, contraction and relaxation, both in cross sectional and serial follow-up.

The management of cardiovascular compromise in the setting of a Fontan circuit can be very frustrating, especially if the therapeutic aim is to improve cardiac output by treating ventricular dysfunction. As I will discuss, this is often an inappropriate, not to mention ineffective, strategy. The fact that most conventional therapeutic strategies are unsuccessful suggests that the ventricle in the patient with the Fontan circulation operates under conditions different from those found in classic cardiology. The generally poor response to the classic strategies for treatment, therefore, has eroded the reputation of the ventricle in a Fontan circuit.

Correspondence to: Marc Gewillig, Professor of Pediatric and Congenital Cardiology, Gasthuisberg University Hospital, University of Leuven, B-3000 Belgium. Tel: +32 16 34 38 65; Fax: +32 16 34 39 81; E-mail: marc.gewillig@uzleuven.be

Interpreting the functionally single ventricle

A key question is whether the dominant ventricle in the Fontan circuit is a dilated small ventricle, or rather a large ventricle trying to adapt to different, unusual, and very abnormal circulations.

Traditionally, the ventricle of the functionally univentricular heart has always been compared to the left ventricle, the latter considered normal for body surface area. Normalizing for body size is widely accepted in medicine. When assessing a pump like the heart, however, any engineer would adjust or normalize for "due output". In an individual with a normal circulation, there is a well-determined relationship between output and age or body surface area. This "normal" relationship in the patient with a functionally univentricular heart will long have been relinquished during the various palliative stages, including creation of the Fontan circuit itself.

Preload related to age or body surface area

When preload is related to a left ventricle normal for body surface area, any enlargement of the ventricular cavity is considered to be due to dilation. A hypothetical patient with a functionally univentricular heart is born with a ventricle submitted to a fetal volume overload of about 230% of normal, which is shortly thereafter augmented by a shunt procedure to around 280%[2,3] (Fig. 1). Chronic volume overload of this magnitude will damage the ventricle. The patient slowly outgrows his shunt, thereby gradually reducing the volume overload to around 250%. A second shunt may be needed, a strategy typically employed prior to the 1990s, again increasing the volume overload to about 280%. As this patient

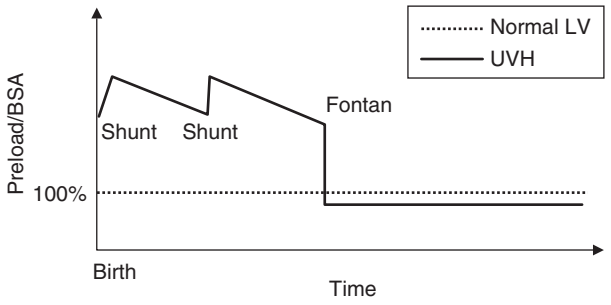

Figure 1.
Preload normalized for body surface area (BSA) versus time in the setting of a normal left ventricle (LV – dotted line), and in a hypothetical patient with a functionally univentricular heart (UVH – solid line), before and after construction of two shunts and a Fontan procedure. Such analysis assumes the ventricle to be big due to dilation, not because of overgrowth.

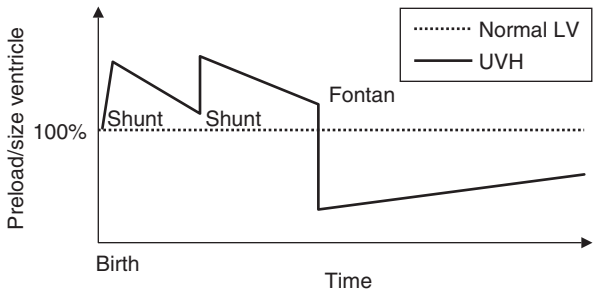

Figure 2.
Preload normalized for ventricular size versus time in the setting of a normal left ventricle (LV), and in the same hypothetical patient with a functionally univentricular heart (UVH) as considered in Figure 1, before and after two arterio-arterial shunts and a Fontan procedure. The analysis depicted in this figure assumes the ventricle to be big due to overgrowth, not because of dilation.

again outgrows the shunt, a Fontan circuit is created, reducing the volume preload to about 70% of normal for body surface area.[4,5]

Thus, when viewed in relation to a left ventricle normal for body surface area, this ventricle is already dilated at birth. It is enormously overloaded when first shunted, becomes even more dilated while reshunted, and remains significantly dilated but mildly underused when the Fontan circuit is completed. If we take this view, therefore, the ventricle is large due to stretch and dilation, and not as a result of overgrowth.

Preload related to ventricular size

The same sequence in the same hypothetical patient looks very different when preload is related to the size of the ventricle, which reflects chronic output (Fig. 2). For the sake of simplicity, let us first assume the ideal situation, in which all initial dilation during the adaptation of the ventricle is converted in time into appropriate overgrowth. Such a patient is then born with a functionally univentricular heart appropriate in size for the previous fetal output, representing a volume load of 100% of normal for ventricular size. The preload of the ventricle is increased shortly after birth by a shunt procedure to around 150%. The patient slowly outgrows the shunt, and adapts the ventricle, thereby gradually reducing the volume overload to around 100% for the size of the ventricle. A second shunt may be created, increasing the volume overload again to 150%. Eventually, the Fontan circuit reduces the volume load to 25% of its "due" preload. Thus, when viewed in relation to the size of the ventricle, this ventricle is normally proportioned for its output at birth, mildly overloaded when shunted, adapts gradually to the high preload, remains overgrown, but becomes significantly, and sometimes critically, underloaded and underused. It is thus oversized at the time of completion of the Fontan circulation.

Discussion

The concept as discussed above, in the second version, is appropriate for the "ideal" patient, in whom all dilation is compensated and converted into overgrowth, and thus adapted to the volume load. In reality, some inappropriate and irreversible dilation of the ventricle will occur, resulting in volume preload during the Fontan circulation of between 25% of its due preload should there be no dilation but exclusive overgrowth, and 70% in the setting of exclusive dilation and no overgrowth. The self-evident end result is systolic and diastolic ventricular dysfunction in the setting of the Fontan circulation.

Needless to say, and as discussed in my introduction, ventricular dysfunction can also be due to the congenital malformation itself, or due to the previous surgical manipulations such as bypass, and so on. The ventricle may now enter a vicious cycle, whereby the low preload results in poor ventricular filling, and thus lowered cardiac output, with creep to a further "compacted" state, with eventually continuously declining cardiac output. This phenomenon of progressive "disuse hypofunction" is well known. A chronic preload of less than 70% of the "due" preload impacts progressively and negatively on left ventricular function.[6]

Much of the ventricular dysfunction subsequent to completion of the Fontan circulation is due to severe underloading of an overgrown ventricle. This explains the lack of success of classic therapeutic strategies. An empty ventricle will not contract better or pump more when whipped, but only when filled more adequately! The best theoretical treatment, after avoiding such unloading, is to augment ventricular preload. Possibilities for achieving such augmentation,

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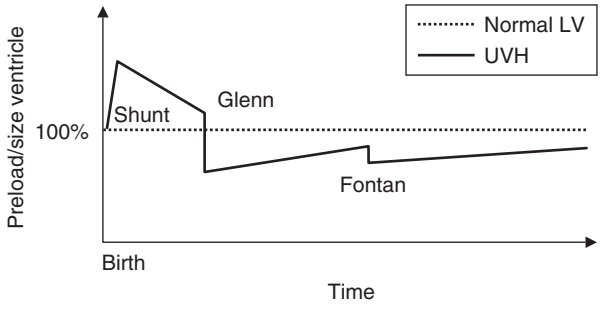

Figure 3.
Preload normalized for ventricular size versus time in the setting of a normal left ventricle (LV), and in the same hypothetical patient with a functionally univentricular heart (UVH) before and after an aorto-pulmonary shunt, followed by a partial cavo-pulmonary Glenn shunt, and later by a Fontan type procedure. This strategy avoids any excessive unloading.

however, are very limited. It may be possible to optimize the Fontan connection, to lower pulmonary vascular resistance, or to create a fenestration. Mitral and aortic valvar regurgitation, or creation of an arterial-pulmonary venous connection, do increase preload for the ventricle, but not without potentially significant side effects for the Fontan circuit.

The best therapeutic strategy, therefore, consists of avoiding excessive volume overload, thereby avoiding later excessive unloading. Excessive volume overload is prevented by early banding, creation of a rather small neonatal arterio-arterial shunt, or by the early placement of a partial cavo-pulmonary shunt. On the other hand, a stepwise partial cavo-pulmonary shunt prior to the completion of the Fontan circulation (Fig. 3), with appropriate fenestration, and avoidance of concomitant repair of an atrioventricular valve, may avert excessive underloading.

Both dilation and overgrowth are mechanisms leading to a big ventricle. Hypocontractility of the ventricle in the Fontan circuit may be in part due to dysfunction, but also in part due to underfilling for body surface area, and certainly for ventricular size. All reports so far fail to distinguish between these components, but simplify matters by considering only the aspect of dilation. In order better to assess and understand the ventricle that is enlarged, we certainly need methods to split up the potential components for its increase, such as growth, overgrowth, dilation, stretch, and reorientation of the myocytes, each with a reversible and an irreversible component! Only then we will understand how far we may go in overloading and underloading a ventricle.

Low cardiac output

The most frequent clinical problem in the patient with a Fontan circulation is low cardiac output. A cardiologist faced with any patient in a low output state, and with decreased systolic or diastolic cardiac function, will in all likelihood conclude that the heart is the limiting factor of the circulation. Having reached this conclusion, it is not surprising that therapeutic strategies are frequently chosen with the aim of improving ventricular function.

The situation is completely different in patients with the Fontan circulation. There is ample evidence that the limiting factor for cardiac output in the majority of such patients lies within the pulmonary vascular resistance, and only exceptionally within the Fontan connection itself, or within the dysfunctioning ventricular mass.[7] For the cardiologist, nonetheless, it remains a challenge not to blame the ventricle for the low output state, especially in the perceived setting of ventricular dilation and dysfunction. The conventional treatment of the low output state will then be very frustrating, as the ventricle is not the limiting factor. Several studies have shown little impact, either acute or chronic, of medication such as inotropics, afterload reductors, vasodilators or beta-blockers on ventricular function or cardiac output. Cardiac pacing is only beneficial in the setting of severe bradycardia or atrioventricular dissociation.

How can we avoid or treat a low output state in patients with the Fontan circulation? The management of such patients over the past few years was directed at maximal preservation of ventricular function, by avoiding excessive overloading and underloading. This strategy has been successful. The ventricle after completion of the Fontan circuit may now show some overgrowth, dilation, and mild dysfunction, but mostly within acceptable limits. In contrast, preservation of ventricular function may, in some patients, have created another problem, namely inadequate growth of pulmonary vessels. Adequate growth of pulmonary vessels requires a mild to moderate overflow. A Fontan circuit with underdeveloped pulmonary vessels, even in the presence of an excellent ventricle, is doomed to fail! The ideal volume load, acceptable for the ventricle and necessary for adequate pulmonary growth, still needs to be determined.

If avoidance of a low output state has failed, treatment is very difficult and frustrating. The management may include optimizing the Fontan connections, optimizing the pulmonary arteries by means of balloon dilation or stenting, obliteration of any residual arterio-arterial shunt, such as a patent arterial duct or shunt, or flow through collateral channels, increasing the tension of oxygen by moving the patient to higher altitude or providing supplementary oxygen, using pulmonary vasodilators, or varying the pattern of breathing. Fenestration will do no more than increase output at the expense of saturation. If all fails, transplantation can be considered, but at a higher

risk than expected because of multiple adhesions after multiple sternotomies, because of the usual cachectic state that renders the patient vulnerable for infections, and because of the increased pulmonary vascular constriction at the time of transplantation.[8]

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