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Original Article

The Fontan operation 30 years later

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BASED ON THE REVOLUTIONARY CONCEPT OF THE dispensable right ventricle,[1] the Fontan operation stands as the extreme example of physiologic repair in pediatric cardiac surgery, serving as the common surgical denominator for the entire spectrum of functionally univentricular conditions. Also called ortho-terminal repair, it makes allowance for a circulatory arrangement that is in-series by effectively driving the pumpless systemic venous return directly through the pulmonary circulation. If this arrangement is to work, then two conditions must be fulfilled. First, there must not be any anatomical or functional hindrance along the whole Fontan pathway, from the caval venous inflows to the aortic outflow. Second, there must be a well-suited systemic pump, essentially recruited to the dual function of drawing blood from and, simultaneously, driving blood through, the lungs. This latter function, driving the blood through the lungs, takes place during the venous phase of the systemic output.

Over the last three decades, several modifications of the original procedure have been introduced with the purpose of alleviating the postoperative complications produced by systemic venous hypertension, and expanding the potential for circulatory adaptation to physical exertion. Despite the creation of the Fontan circulation achieving universal acceptance, some still object to the real efficacy of the arrangement in providing adequate long-term survival and quality of life. In its place, they advocate less invasive palliations. It would seem the time, therefore, for some reflections on the appropriateness of this "one fits all" operation.

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Staging and preservation of ventricular function

All patients with functionally univentricular hearts, whether or not they have been palliated by procedures such as creation of a systemic-to-pulmonary arterial shunt or banding of the pulmonary trunk, are forced to manage an intrinsically inefficient circulation as a result of the recirculating parallel pulmonary and systemic outputs.[2] The related chronic volume overload severely affects the morphologic and functional characteristic of the dominant ventricle. First, it alters myocardial development, with potentially irreversible changes in the myocytic structure, including myocytic apoptosis, and in comparable changes to cardiac collagen and elastin. Second, it induces a progressive ventricular dilation, with increasing biomechanical stress on the ventricular wall. The whole process eventually evolves into overt cardiac failure. On the other side, an operation such as the Fontan procedure that suddenly abolishes an excessive volume work may also induce an unfavorable geometrical adaptation of the systemic ventricle by drastically altering the relationship of its mass to volume. This process may cause relative ventricular hypertrophy and secondary diastolic dysfunction. The degree of the relative ventricular hypertrophy is even greater in the presence of a concomitant pressure overload if there is any obstruction within the aortic outflow tract.

From a clinical standpoint, the overall prognosis of the patients converted to the Fontan circulation was improved markedly after the introduction, in the mid-1980s, of the bidirectional cavopulmonary anastomosis, the so-called bidirectional Glenn shunt or hemi-Fontan operation, particularly if combined with a Damus–Kaye–Stansel anastomosis to bypass subaortic obstruction, should that be present.[3] It would appear that the bidirectional Glenn shunt, used as an intermediate stage towards creation of the

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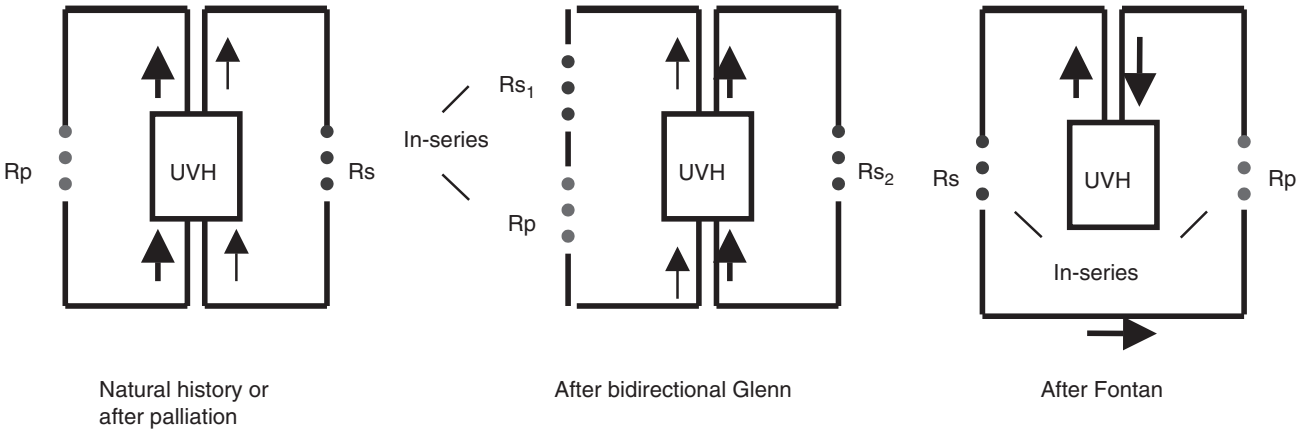


Figure 1.
Circulatory patterns in the presence of a univentricular heart. UVH: univentricular heart; Rp: pulmonary resistance; Rs: systemic resistance (Rs_1: upper district; Rs_2: lower district).

Fontan circulation, tapers the process of ventricular remodelling after volume unloading. More precisely, the degree of change in murall thickness, cavity volume, and the ratio of the thickness of the ventricular wall to the diameter of its cavity that it induces, is of significantly lesser extent than after the Fontan operation. Interestingly, in the setting of the Glenn shunt, while the superior caval venous flow is put in series with the pulmonary vascular bed, the upper and lower systemic hemi-circuits become, instead, arranged in parallel, with the arterial output for the upper body facing two levels of resistance, at both systemic and pulmonary levels (Fig. 1). We can speculate, therefore, that the systemic flow to the lower body, which confronts only the systemic resistance, is faster, particularly in a condition of increased demand for cardiac output. The relative increase in cardiac output maintains a condition of moderate volume overload and, obviously, occurs at the expense of lower systemic oxygenation. Only once the Fontan repair is completed, and the pulmonary and systemic circulations are totally placed in series, is the oxygenation normalized and the volume overload definitively abolished. With the completion of the Fontan repair, it is noteworthy that the additional decrease in ventricular volume is relatively limited and, therefore, easily tolerated.

At this point, the performance of the system, and particularly its adaptation to physical exertion, depends upon many factors, predominantly a well-preserved ventricular function. Hence, in preparation to, or in combination with, a Fontan repair, we must aggressively address any impairment of ventricular function secondary to one or more of the following three mechanisms: pressure overload as seen in the setting of subaortic obstruction, volume overload as produced by atrioventricular valvar regurgitation, or severe hypoxia as seen in the case of pulmonary arteriovenous fistulas. In this respect, normalization of ventricular function and/or the myocardial mass is more likely after removal of the insulting mechanism early in infancy than in later years.[4]

From atrial "pump" to total cavopulmonary connection

In 1988, Marc de Leval revolutionized the Fontan principle,[5] overcoming the old theory of the right atrium acting as a surrogate of the right ventricle, and being capable, to some extent, of propelling blood into the lungs. He introduced, instead, the new concept of the total cavopulmonary connection.[5] This consists of the combination of a bidirectional Glenn anastomosis with a tubular connection from the inferior caval vein to the pulmonary arteries. The inferior caval venous channel can be an intra-atrial tunnel, as in the original technique proposed by de Leval,[5] or an extracardiac conduit.[6] The advantages of completely tubular cavopulmonary connections stem primarily from conservation of the kinetic energies engendered by the systemic venous flows, which are no longer dispersed into spinning swirls stagnating in the atrial reservoir, but routed instead as concentric laminar wedges along conduits with cylindrical sections. As de Leval pointed out, the total cavopulmonary connection is technically simple, and can be reproduced with any anatomical substrate. It is constructed well away from the atrioventricular node, leaves the right atrial chamber at low pressure, reduces the risk of early or late arrhythmias, and minimizes the risk of atrial thrombosis. The total cavopulmonary connection is such an efficient system that many patients with failing atriopulmonary anastomoses, nowadays, are converted into the total connection rather than being referred for cardiac transplantation.

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Since the original report from de Leval et al.,[5] many studies using in vitro experiments and computational fluid dynamics have provided both qualitative and quantitative assessments of different layouts of the caval venous channels, and have led constantly to refinements in the design of the total cavopulmonary connection.[7,8] A great deal of controversy has revolved around the issue of how best to position the inferior caval venous channel with respect to the superior cavopulmonary anastomosis. Concepts like offsetting, flaring, velocity vectors, and so on have become all too familiar to us. Other entities, such as the "beneficial vortex", are now receiving attention.[9] As Dubini et al.[10] point out in their review, however, it is important to emphasize "that mathematical models should provoke, predict, and not take the place of new experimental observations".

Fenestration

Despite the hydrodynamic improvements introduced by the total cavopulmonary connection, significant morbidity may persist due to the effects of systemic venous hypertension, particularly in the splanchnic district, where an increased production of lymph may sometimes evolve into a vexing protein losing enteropathy. To cope with this scenario, in 1990, Bridges et al.[11] advocated the use of a calibrated fenestration in the inferior cavopulmonary conduit. The rationale underlying this was to allow for some right-to-left shunting at the atrial level, thus maintaining cardiac output during the high-pulmonary resistance phase in the early postoperative period. If necessary, the fenestration can be secondarily closed with an umbrella device in the cardiac catheterization laboratory.[11]

Many reports prove that the fenestrated total cavopulmonary connection has substantially mitigated the postoperative morbidity secondary to systemic venous hypertension, and has made it possible to repair a considerable number of candidates previously considered at high-risk or borderline for creation of the Fontan circulation. Interestingly, a recent prospective randomized study showed that fenestration improves the early outcome even in patients deemed at standard risk. It does this by reducing pleural drainage, and also reducing the duration of hospital stay and the need for additional procedures.[12] It follows that fenestration may prove to be beneficial in all patients considered for a Fontan operation.

One-and-a-half ventricle repair

An interesting subgroup is that of patients with two ventricles potentially amenable to septation, but with the right side of the heart presenting anatomical and/or functional attributes unsuitable to handle the entire cardiac output. In these cases, an alternative is the so-called one-and-a-half ventricle repair. Intuitively a more efficient solution than conversion to the Fontan circulation, this operation consists of a bidirectional Glenn anastomosis combined with the closure of the intracardiac communications, and with connection of the "inadequate" ventricle to the pulmonary arteries.[13] Notably, moderate hypoplasia of the right heart is a safe anatomic set-up for a pulsatile bidirectional Glenn anastomosis. The one-and-a-half ventricle repair has the advantage of reducing the surgical risk for biventricular repair and, compared to the Fontan circulation, it maintains a low right atrial pressure, pulsatile flow of blood to the lungs, and improves the systemic saturations of oxygen.[14]

Are there valid alternatives to the Fontan operation?

Overall results of the Fontan operation nowadays are generally satisfactory, both for survival and for quality of life, including a sizeable cohort of high performers. An even greater proportion of patients converted to the Fontan circulation, however, may still experience significant attrition and morbidity, including: myocardial systolic and diastolic dysfunction, systemic arterial and venous hemodynamic problems, diminished exercise capacity, arrhythmias, protein-losing enteropathy, retardation of somatic growth, dilation and insufficiency of the neo-aortic valve and root, thromboembolic complications, and below-average cognitive development.

It is legitimate, therefore, that some researchers, and particularly the Paris school, have speculated on alternative, less invasive, and possibly smarter solutions. This becomes even more justifiable if one considers that more radical solutions, such as ventricular septation or cardiac transplantation, are virtually impossible in most cases. Interestingly, we recently found that cardiac transplantation in patients with functionally single ventricles entails substantial early mortality, and that a bidirectional cavopulmonary anastomosis provides the best transition to subsequent transplantation. This experience, therefore, should always be kept in mind in the decision-making process towards completion of the Fontan circulation in candidates deemed to be at high-risk, since rescue transplantation in is unwarranted, it seems, in the setting of the failing Fontan circulation.[15]

In the dissertation preceding this review in this journal, Sidi pinpoints a number of convincing arguments in favor of his approach to the problem of treating patients with a functionally single ventricle, specifically the construction of the partial cavopulmonary circulation with an additional source of

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pulmonary blood flow.[16] Essentially, by keeping the abdominal district decongested, and by adding a supplemental source of flow to the lungs in addition to the bidirectional Glenn anastomosis, it is possible to assure adequate oxygenation for the patient, while abolishing most of the side effects of the Fontan circulation.

A crucial point for the surgeon is to be able to regulate the composite flow of blood to the lungs to obtain the magic level of a systemic saturation of oxygen between 85% and 90%, as suggested by Sidi. This corresponds to absence of ventricular volume overload, and "is usually sufficient to avoid polycythemia and hyperviscosity and to allow a decent intellectual life". Although fascinating, this theory may be hard to apply in common practice. Decisions about the size of the shunt, or the tightness of the pulmonary arterial band, cannot rely on specific rules, but rather depend on the experience and "savoir faire" of the surgeon. Furthermore, what is a perfectly adjusted flow of blood to the lungs today may become inadequate in the very near future, considering the rapid pace of growth of infants and young children. This may lead to a distressing series of fine-tuning interventions on the "additional" source of pulmonary blood flow, and should be taken into account in the equation of the life of any individual patient. I suspect that, to dilute reoperations, it would be natural to bargain on the amount additional pulmonary flow, accepting oversized accessory pulmonary arterial outlets, and falling back into the issue of overcirculation and ventricular overload. I am sure, however, that the approach advocated by Sidi may be the best for many patients deemed to be at high risk for construction of the Fontan circulation.

Conclusions

The ideal surgical treatment of patients with a functionally single ventricle has not yet been identified. Many sophisticated theories have been proposed for the "perfect" operation, but in common practice it is more appropriate to refrain from dogmatic approaches. Rather, any single patient should be considered on an individual basis, and the most appropriate "ad hoc" solution should be sought every time.

In most centers, this attitude would probably lean towards creation of a total cavopulmonary connection for most patients, and this is also my preference. Tt should be recommended, nonetheless, that patients enrolled in a protocol of staged ortho-terminal repair should be managed in a very timely fashion, and without letting too much time elapse between the stages. Furthermore, any additional problem potentially impacting on cardiac function, such as the development of atrioventricular valvar regurgitation, similar problems, should be addressed aggressively to maximize the chances of preservation of the organs, in particular the heart, lungs, and brain.

The cooperation between paediatric heart surgeons and bioengineers has certainly produced many fundamental contributions in the quest for the optimal ortho-terminal repair. It is unclear how much room remains for improvement in this field. We should probably never forget, however, that even the best arrangement of the Fontan circulation is conceptually closer to definitive palliation than to truly complete repair. Our efforts should probably be directed towards the search for additional sources of energy to recruit as neo-pulmonary ventricles.

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