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Editorial comment

When Fontan circulation is not the only strategy

Cuando la circulación de Fontan no es la única respuesta

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At the start of the 20th century, the emerging specialty of cardiac surgery faced considerable challenges due to the lack of cardiopulmonary bypass, which hindered intracardiac repair procedures. This led the cardiac surgeons of the time to devise palliative operations to increase or decrease pulmonary flow based on the presence or absence of restricted pulmonary flow. Thus, on November 9th, 1944, Alfred Blalock, with the immeasurable help of his assistant Thomas Vivien, carried out the first systemic-to-pulmonary artery shunt to increase pulmonary flow. This procedure involved anastomosis between the subclavian artery and the pulmonary artery, radically altering the prognosis of cyanotic patients. Eight years later, in 1952, Muller and Dammann introduced pulmonary artery banding to reduce pulmonary flow in patients with unrestricted pulmonary flow and thereby prevent the development of pulmonary vascular disease. Although these advances altered the course natural history of these patients, the surgeons knew that their prognosis could only be modified through safe access to the intracardiac cavity that did not provoke cardiocirculatory arrest. After various attempts, the routine use of cardiopulmonary bypass was established in 1955, marking the start of modern cardiac surgery. Initially, efforts concentrated on repairing patients with biventricular circulation, and it was not until 1971 that Francis Fontan developed total right heart bypass for patients with single-ventricle physiology. Since then, Fontan surgery, with multiple modifications, has become the surgical procedure of choice for the management of patients with a univentricular heart.

Both the development of new surgical techniques and the adoption of new therapeutic advances for managing patients with congenital heart diseases have significantly enhanced survival. Indeed, in most developed countries, the adult population with congenital heart diseases already exceeds the pediatric population.[1] Alongside this improved survival, research in this field has exponentially increased, particularly in the last 20 years, as shown by the growing number of publications related to the topic. A simple PubMed search with the term "congenital heart disease" reveals that more than 7000 articles related to adult congenital heart diseases were published in 2022 alone. Of these, almost 600 articles concerned patients with Fontan circulation. However, most of these publications focus more on assessing the consequences of the residual lesions of the different surgical procedures and less on the natural history of the disease. Therefore, the recent article published by Gordon et al.[2] in *Revista Española de Cardiología* is crucial for understanding the natural history of patients with univentricular heart who, for different reasons, did not complete the Fontan circulation.

Before going any further, I must first congratulate the authors for focusing their attention on an important group of patients that is rarely seen in congenital heart disease clinics, at least in the Western world. The study by Gordon et al.[2] presents a descriptive study of 120 patients with univentricular circulation and restricted pulmonary flow without completed Fontan circulation, either due to medical contraindication or family preferences. The authors divided the patients into 3 groups: group 1, patients with restricted pulmonary flow (55 patients), most with native pulmonary stenosis (41; 74.5%) and the remainder with surgical banding of the pulmonary artery (14; 25.5%); group 2, patients with a cavopulmonary shunt (30 patients, the smallest group); and group 3, patients with a systemic-to-pulmonary shunt (35 patients).[2] Strikingly, even though the patients had a complex congenital heart disease, almost half (44.2%) had not previously undergone follow-up in a specialized congenital heart disease clinic. This indicates an unmet need in the 21st century to improve the education of both medical staff and patients themselves about the value of follow-up in clinics specialized in adult congenital heart diseases. Such follow-up is crucial, not only due to the complex nature of their disease, but also because research has shown that the absence of follow-up in specialized units is directly associated with increased mortality in this population.[3] In addition, even with a relatively short follow-up, it was interesting, albeit not surprising, to see that all patients in all groups experienced a progressive deterioration in ventricular function and functional class. Given that therapeutic options in most of these patients who develop major functional deterioration are typically limited to heart transplant, follow-up should be conducted in heart failure units specialized in the management of patients with congenital heart diseases. This allows for timely assessment for heart transplantation before multiorgan deterioration occurs. Although ventricular function is a major factor when evaluating these patients for heart or lung transplantation, such assessment is often prompted by functional deterioration and/or the onset of atypical symptoms of heart failure, which can be missed by cardiologists without experience of congenital heart diseases.

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In the study by Gordon et al.,[2] patients with a systemic-to-pulmonary shunt had the worst prognosis, with a higher risk of heart failure and/or arrhythmias, a greater deterioration of ventricular function during follow-up, and higher mortality. These outcomes were expected, given that half of these patients did not complete the Fontan surgery due to the presence of unfavorable anatomy, comprising hypoplastic pulmonary arteries, or due to the development of pulmonary hypertension before the procedure was completed. Both are high-risk factors for morbidity and mortality in these patients. While heart transplant is a therapeutic option for these patients, those with a hyperplastic pulmonary tree or established pulmonary arterial hypertension will need heart-lung transplantation, which has high short-term morbidity and mortality and worse mid-to-long–term survival. Given the poor prognosis of this group of patients, also reported by previous publications,[4] a treatment plan must be established that includes early referral to heart-lung transplantation units to assess the feasibility of transplantation, if required. As specialists in the management of patients with congenital heart diseases who develop heart failure, we often encounter these patients when the heart failure is already refractory or the renal failure is irreversible, limiting their possibilities for transplantation. In my opinion, this happens so frequently because it is assumed that transplantation is not a therapeutic option due to the complexity of the anatomy in this group of patients. Although this is true for many of these patients, it is important to stress that the eligibility of these patients for heart or heart-lung transplantation mainly depends on the experience of the transplant team in the referral center. Accordingly, it is crucial to refer them early so that, if transplantation is a viable option, it can be performed before irreversible multiorgan failure develops. In addition, patients not eligible for transplantation must be explained their mid-to-long–term prognosis and be offered psychological support and, when the time comes, palliative care.

The patients included in group 1 warrant special mention. This group included patients with well-balanced circulation, mostly natural. These patients have sufficient pulmonary flow to maintain appropriate oxygen saturation without the need for previous surgery. In the article by Gordon et al.,[2] this group of patients had the best prognosis, with greater survival and less ventricular function deterioration during follow-up. However, when it comes to extracting conclusions from the findings of the study, we should remember the survival bias in this population. Given that the study only included patients who survived to adulthood, we cannot conclude that neonates with univentricular heart and well-balanced circulation will always have better prognosis. What we can conclude—and this is important—is that survival in patients with univentricular heart who reach adulthood without previous surgery will be better than in children requiring a systemic-to-pulmonary artery shunt. Accordingly, given the unfavorable prognosis of patients with a systemic-to-pulmonary artery shunt, it seems reasonable not to perform the procedure in adults solely to improve their oxygen saturation.

Although the article by Gordon et al.[2] cannot and must not be used to make therapeutic decisions in pediatric patients with univentricular heart, it reminds us of a crucial consideration: in medicine, inaction is sometimes the best option and limiting the number of interventions can even improve patient prognosis. In this study, group 2 patients (with a cavopulmonary shunt without completed Fontan circulation) had a similar risk of death or heart failure to that of group 1, even though they had a higher risk of stroke. Given that patients with cavopulmonary shunt not completing Fontan circulation have a lower risk of typical Fontan-associated complications, such as liver failure and lymphatic abnormalities, the study by Gordon et al.[2] suggests that patients with well-balanced circulation and adequate oxygen saturation after a cavopulmonary shunt benefit in the long term from a conservative strategy that does not include Fontan circulation completion. While this is an interesting concept, given that that the absence of the intrinsic complications of the Fontan circulation may improve prognosis in these patients, the next step would be to identify which children would benefit from this strategy. This would require close collaboration among pediatric cardiologists, adult cardiologists, and cardiac surgeons to establish clinical studies addressing these types of questions. Perhaps it is time for pediatric and adult cardiologists to join forces to design studies that go beyond short-term prognosis and aim to identify the best therapeutic strategy for patients with univentricular hearts. Such a strategy should not only help them reach adulthood, but also ensure they undergo heart transplantation in the best condition possible.

Although Fontan circulation must still be the surgical procedure of choice for patients with univentricular heart, studies such as that by Gordon et al.[2] remind us that this is a palliative procedure and that further research must be conducted to improve the prognosis of these patients without closing the door to any therapeutic option.

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CONFLICTS OF INTEREST

None.

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