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Letter to the Editor

We read with interest the manuscript entitled "The real fate of pulmonary arteries after bidirectional superior cavopulmonary anastomosis: is there a need for concern?" that appeared in the January 1999 issue of *Cardiology in the Young*.[1] This article begins with the premise that the growth of the pulmonary arteries may be impaired following the bidirectional superior cavopulmonary anastomosis. The authors emphasize the importance of this concept, as they indicate that pulmonary arterial size may influence outcome at the time of the Fontan procedure. Having stated this hypothesis, the authors suggest that a more accurate method for measuring pulmonary arterial dimensions is needed, and that use of magnetic resonance imaging may help resolve issues related to pulmonary arterial growth and management in this population. They conclude that " . . . the fate of the pulmonary arteries after bidirectional cavopulmonary anastomosis is still unknown . . . and thus, there remains a need for concern."

We disagree with the premise of this article and, therefore, had difficulty accepting their conclusions. Specifically, we do not believe that the fate of the pulmonary arteries after bidirectional cavopulmonary anastomosis is an unknown. Since the inception of the two-staged approach to completion of the Fontan circulation in the late 1980s, thousands of children born with functionally single ventricles have undergone a cavopulmonary anastomosis and subsequent Fontan procedure, and are proof that there is adequate growth of the pulmonary arteries. We say adequate, because the goal of this approach is to reduce morbidity and mortality, and it is evident that this strategy has been successful in achieving improved results.[2]

It is a paradox that so much emphasis has been placed upon the size of the pulmonary arteries and their growth, when, indeed, this is not the central issue. What is the evidence that pulmonary arterial size has any relevance? There are a few early articles that concluded that size, along with architecture and pulmonary vascular resistance, are three important risk factors at the time of the completion.[3–4] Recent articles, nonetheless, have concluded that size of the pulmonary arteries is not a risk factor.[5–8] Since small pulmonary arteries are frequently seen in conjunction with architectural problems, it is possible that size never was an independent risk factor. And now that pulmonary arterial stenoses can be ballooned or stented in the catheterization laboratory,[9] the risk attributable to architectural problems has also been ameliorated.

It is widely held that pulmonary vascular resistance, pulmonary arterial size, and pulmonary architecture are also risk factors for the cavopulmonary anastomosis, a concept that has been carried over from the literature related to the Fontan circulation. There is no evidence that pulmonary arterial size has any relevance as a risk factor for the cavopulmonary anastomosis. Indeed, surgeons have recognized that this operation provides an opportunity to improve both size and architecture by augmenting the branches of the pulmonary trunk. What are the risk factors for the cavopulmonary anastomosis? In a recent review[10] of 400 patients, the only factor found to be predictive for increased mortality was an urgent operation requiring concomitant procedures. Jacobs and his colleagues[10] concluded that many traditional risk factors appear to have been neutralized by interposition of the hemi-Fontan operation. Having said this, the literature is replete with articles that have focused on pulmonary arterial growth following the cavopulmonary anastomosis.[11–13] Most of these papers conclude that pulmonary arterial growth is "impaired" following this operation. It is not surprising that pulmonary arterial growth is reduced, since the ratio of pulmonary to systemic flows is less than one. Whether growth is impaired or not is a matter of debate. Several authors have advocated the use of accessory sources of pulmonary blood flow, and have shown that this will enhance pulmonary arterial growth.[14] We do not doubt that the addition of an extra source of blood flow will result in additional growth. Given the preceding discussion, however, this seems to be pursuing a questionable goal. What is the point of growing large pulmonary arteries, when pulmonary arterial size is no longer viewed as a risk factor?

We have recently reviewed our own experience with patients undergoing the cavopulmonary anastomosis specifically to evaluate the effect of accessory pulmonary flow on survival.[15] The results of this study are shown in Figure 1, and indicate that the provision of accessory flow was associated with

significantly higher mortality at mid-term follow-up ($p < 0.02$). The results also demonstrate that the probability of achieving a successful Fontan circulation was 74% with accessory flow, versus 92% without accessory flow ($p < 0.01$). Parenthetically, no patient was declined for the Fontan procedure due to "small pulmonary arteries." We concluded that the addition of accessory pulmonary blood flow at the time of the cavopulmonary anastomosis has an adverse impact on short and mid-term outcome.

Thus, Slavik and his colleagues[1] began their paper with the premise that pulmonary arterial growth is impaired following construction of the cavopulmonary anastomosis, a concept that we do not believe is well-justified. The authors built upon this premise by suggesting that we need better methods of measuring pulmonary arteries, and that provision of an accessory source of pulmonary blood flow may facilitate pulmonary arterial growth. The first of these ideas we question in its importance, and the latter idea we generally view as detrimental based on our own data and experience. The authors conclude that there is need for a "prospective, multi-institutional study involving sequential non-invasive magnetic resonance imaging assessment of pulmonary arterial development." This is a logical conclusion based on their point of departure, but is only as valid as that original premise. We believe that there still are important controversies regarding the management of patients with functionally single ventricle, but pulmonary arterial size should not be considered one of them.

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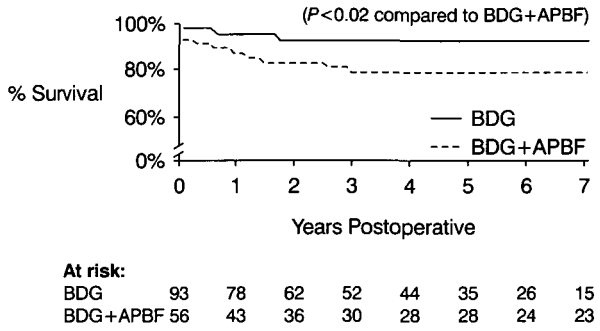

Figure 1.
A graph showing survival curves for patients undergoing a bidirectional Glenn (BDG) procedure along with those having a bidirectional Glenn in the presence of an accessory source of pulmonary blood flow (APBF).

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Reply to Letter

The letter was shown to the authors, who responded as follows:

Thank you for the opportunity to reply to the letter of Mainwaring and Lamberti related to our previous paper.[1] We state categorically that there is no premise in our article that "growth of the pulmonary arteries may be impaired following the bidirectional superior cavopulmonary anastomosis". The real premise is that the group of patients undergoing the bidirectional superior cavopulmonary anastomosis cannot be treated as a homogenous cohort, and that there are several methodological issues which make difficult the direct comparison of the data published thus far. Unfortunately, we do not share the views of Mainwaring and Lamberti. It remains our opinion that the true fate of pulmonary arteries following the superior bidirectional cavopulmonary anastomosis is important and unknown. We do not believe that the statement "thousands of children born with functionally single ventricles have undergone a cavopulmonary anastomosis and subsequent Fontan procedure" is the real proof of the adequate growth of pulmonary arteries. The detailed analysis of the largest series to date showed the strong association between patients having smaller pulmonary arteries and "early failure or persistent effusions" after the modified Fontan operation in an otherwise low-risk subset of patients.[2] Every institution selects its patients very carefully before their inferior caval venous blood is redirected into the pulmonary arteries. Despite this policy, every institution has its fair share of the patients in whom the Fontan procedure has failed. There is also a wide variation in the timing of the Fontan procedure. Moreover, we believe that many institutions, including ours, have some patients deemed unsuitable for the completion of the so-called total cavopulmonary connection due to the prohibitively high pulmonary vascular resistance and transpulmonary gradient subsequent to construction of the superior bidirectional cavopulmonary anastomosis. Unfortunately, it is not usual, with the rare exception,[3] to discuss in detail patients falling in these categories in the series published to date. Some of these patients will have underdeveloped *peripheral* pulmonary arteries.[3] This was the reason why we suggested that pulmonary arteries be measured at a relatively distal site, as we are aware that almost any distortion or hypoplasia of the central pulmonary arteries can be corrected surgically at the time of the superior bidirectional cavopulmonary or Fontan procedures.

It was not the aim of our paper to discuss the influence of pulmonary arterial size on the outcome of the superior bidirectional cavopulmonary anastomosis. Neither, as far as we are aware, is it the object to produce pulmonary arteries that are larger than normal following this anastomosis. We feel strongly that the patients undergoing the superior bidirectional cavopulmonary anastomosis should be divided into two haemodynamic subgroups, and only then that the fate of the pulmonary arteries be assessed. It is obvious from our previous study,[4] and from the review of other series,[5–7] that the patients with increased pulmonary blood flow at the time of their superior bidirectional cavopulmonary anastomosis are likely to have pulmonary arteries that are larger than normal. If these patients undergo the completion of the Fontan operation early, the size of their pulmonary arteries is unlikely to be a cause for concern, or even a risk factor. It is our opinion that the uncertainty dwells with the real fate of the relatively underdeveloped pulmonary arteries in those patients who have restriction of the flow of blood to the lungs at the time of the cavopulmonary anastomosis.

We congratulate Mainwaring and Lamberti concerning the review of patients undergoing a cavopulmonary anastomosis at their institution.[8] It is not obvious from their publication, however, which criterions for selection were used in their series for retention of the accessory source of pulmonary blood flow subsequent to the cavopulmonary anastomosis, nor the size of the pulmonary arteries at any stage of their palliation. Their results contravene some of the previously published data on the role of competitive flows of blood to the lungs following the bidirectional cavopulmonary anastomosis.[9,10] Could the primary selection of the

patients in whom an accessory source of pulmonary flow has been retained play any role in their worse overall outcome? It is, nonetheless, reassuring to know that the authors did not decline any patient for the Fontan procedure due to "small pulmonary arteries". We also assume that none of their patients died after the Fontan procedure because of the presence of "small pulmonary arteries".

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The letter was also shown to Tom Karl, who added an editorial comment to the initial review. He commented:

The authors of the letter have published before on the outcome for patients with a cavopulmonary shunt, and they are knowledgeable in this area. I would agree with them that the statement made by Slavik and colleagues regarding the fate of the pulmonary arteries after a bidirectional cavopulmonary anastomosis was perhaps exaggerated, since certainly a lot is already known at this point. As I emphasised in my original editorial, however, it would be impossible to calculate the effect of pulmonary arterial size on the outcome of the Fontan procedure unless patients with all possible dimensions of the pulmonary arteries were subjected to this operative procedure. Since there is a selection process and there always has been, it would equally be an overstatement for Mainwaring and Lamberti to claim that pulmonary arterial size has no effect on the outcome of the Fontan procedure. The point made by Mainwaring and Lamberti about neutralisation of some risk factors by interposition of the bilateral cavopulmonary shunt helps us identify patients who should not have a Fontan operation. This is different than subjecting all patients with a cavopulmonary shunt to a Fontan operation, and seeing if they do better than patients who do not have a cavopulmonary shunt as a preliminary operation.

To add to this confusion, we must also take into account the fact that many units no longer offer an elective Fontan operation to patients who have a stable circulation with a cavopulmonary shunt. The probability of achieving the Fontan circulation, therefore, is no longer a universally accepted end-point for judging the success of palliation. Thus, the argument is a bit more complex than stated by Mainwaring and Lamberti, although they certainly have written an interesting commentary which is worthy of widespread attention.

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