would proceed; probably our cardiologist, would perform a percutaneous procedure to enlarge the atrial septum. But I cannot tell you numbers with regard to this study what it could be the clear indication to say we would do or not. In this group, that was not the case.

Dr Caldarone. Well, I can commiserate with you because when we look at our records and try to identify the subset of patients in whom a 2-ventricle repair was considered, it is hard to discern. So if we are going to really evaluate the use of hybrid management for 1-ventricle/2-ventricle decision making, as a community we are really going to have to identify those patients up front and say they are in this track and then monitor our outcomes to see how successful or unsuccessful we are. I think that is the only way we are ever going to really be able to assess this niche use of hybrid technology.

Dr Yerebakan. I absolutely agree with your comments and the scientific way to do it. My data also have the drawbacks of a retrospective design, so I am not really able to give clear data for your question.

Dr Sabine Daebritz (*Duisburg, Germany*). It is a very interesting approach. However, what is the rationale of doing this, particularly in the real hypoplastic left heart complex patients. What do you really gain? The decision is anyway very difficult, and there are no hard data to know whether a biventricular repair is going to work or not. The hypothesis is that it is probably most likely working in the neonate because there is some potential of even growth of the heart right after birth. So what do you really gain by adding this step in between?

And the other thing is, often you do not have a restrictive atrial septal defect, so how do you think you really promote growth of the left-sided structures, like in a very unbalanced atrioventricular septal defect or in a nonrestrictive atrial septal defect, if you do not have an intracardiac connection at the ventricular level, a ventricular septal defect.

Dr Yerebakan. So with regard to the second question maybe at first. I do not advocate that we really promote growth. I mean therefore to say we have to have hard data on this.

But what we see is when we apply the new discriminant score that was published in 2006 by Colan et al—this is with regard to your first question—what we gain with this approach is, you see, patients pretty much, as we expected, lie at the discriminant value of minus 0.65. So 18 patients would only be amenable for biventricular correction if we would opt for a decision in the neonatal period. In contrast, when we go to the time of the biventricular correction, this score goes up to 1.47, which means that the number of patients doubles. So we then have growth of the left ventricle structures, as we see with the score, and are sure, or let us say more sure, which patients would tolerate a biventricular correction.

Another reason why we choose this approach is there are data in the literature that tells us that in patients with complex heart disease, the brain maturation is delayed.

And second, the effects of a cardiopulmonary bypass and even hypothermic circulatory arrest in the newborn period has detrimental effects of the neurologic outcome. And what we have just—I am wrapping it up—just to show you, short results of the neurologic outcome; these are preliminary data. This is a study from the literature where we see that 40% of patients lie below the plus directional indicator of 70. In our patients, just preliminary, unpublished data, in 19 patients in comparison to a hypoplastic left heart syndrome patients with conventional repair, only 5% of our patients lie below the discriminate score of 70. And also with the minus directional indicator, this is the same case. So we have avoided neonatal major surgery, with cardiopulmonary bypass and we have at least seen in the preliminary data a satisfying neurologic outcome with this approach. But, as I said, this is just an alternative and only an institutional preference; nobody has to do it.

EDITORIAL COMMENTARY

Kicking the can down the road ... gently

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Hypoplasia of left heart structures presents along a spectrum, ranging from the most severe, mitral and aortic atresia, to the least severe, coarctation of the aorta with bicuspid aortic valve. Two alternative treatment strategies now exist, with assignment of a patient either to a biventricular circulatory arrangement or to a univentricular arrangement, depending the perceived functional adequacy of the left side of the heart. Thus a “continuous” disease must be

managed with “categorical” therapies. For many patients presenting in the middle of the spectrum, it is difficult to discern which strategy is most appropriate, although several predictive scoring systems to guide such decisions have been proposed.¹⁻³ Retrospective analysis suggests that inappropriate assignment may result in substantial morbidity, or even death.³ The report in this issue of the *Journal* by Yerebakan and colleagues⁴ from Giessen describes the outcome for a cohort of patients with varying degrees of left heart underdevelopment in whom the treatment assignment decision was delayed, with the patients initially undergoing a temporizing hybrid palliation in the neonatal period. No patients died in the interstage interval. Ultimately, 40 patients underwent biventricular reconstruction at a median age of nearly 7 months, with 5 deaths, 1 cardiac transplant, and 1 takedown to univentricular circulation, at a median follow-up of 7.9 years.

In an alternative approach suggested by the Boston group, initial palliation is a conventional first-stage Norwood-type operation, with either modified Blalock-Taussig shunt or right ventricle-to-pulmonary artery shunt, deferring the biventricular conversion until after the second or even third stage of palliation.^{5,6} Advantages of this approach include the opportunities to perform complex left-sided valve reconstruction, to resect endocardial fibroelastosis, and to enhance pulmonary blood flow, all before biventricular reconstruction. The Giessen approach does not permit these options, although approximately 10% of their patients had mitral or aortic reconstruction at the time of biventricular repair.⁴ Not surprisingly, the Giessen patients were substantially younger at biventricular conversion (median age of 6.7 months vs 42 months in the Boston experience). The Giessen group had also not previously undergone heart operations at the time of biventricular conversion, very different from the Boston patients, who had undergone a median of 3 heart operations before definitive [*sic*] repair.

An area of commonality between the 2 approaches is the focus on the atrial septum at initial palliation. The aim for both is to leave a restrictive interatrial communication after initial palliation. Indeed, no patient in the Giessen cohort underwent atrial septal intervention before biventricular conversion, guided by a threshold left atrial pressure of 15 mm Hg. The underlying hypothesis is that “forcing” blood to cross the left side of the heart, rather than the atrial septum, promotes growth of the mitral valve, left ventricle, and aortic valve. Comparison of echocardiographic measurements before the hybrid procedure with the same assessments before the biventricular conversion demonstrated a significant increase in aortic valve *z* scores (−2.13 to −1.21). The change in mitral valve *z* scores (0.52 to 0.98) was less impressive, however, and there was no significant change in the ratio of left to right ventricular length (0.90 to 0.92). Nonetheless, although the relative sizes of various left heart structures may

have changed only slightly, if at all, the impact on the discriminant score² (predicting suitability for biventricular repair) was a change from below the discriminant threshold of −0.65 to substantially above it by the time of biventricular conversion (−0.72 to 1.47).

With continued reference to the atrial septum, it is notable that at biventricular repair Yerebakan and colleagues⁴ chose to leave an atrial septal defect in 25% of the patients. Although it is not explicitly stated, here the concept was apparently away from forcing transmitral flow to encourage growth and toward prevention of early left atrial hypertension-induced pulmonary edema. This principle is not new, first appearing in the prebypass era in the management of patients with mitral stenosis⁷ and much later in the neonatal biventricular repair of left heart complex.⁸ The Boston group also reports leaving an atrial septal defect in most of their patients at the time of biventricular repair.⁶

For a neonate who does not obviously fit into either the biventricular or univentricular paradigm, this report suggests that assignment can be safely and successfully delayed for several months. The approach that Yerebakan and colleagues⁴ have described is simple, avoids exposure of a fragile neonate to cardiopulmonary bypass with or without circulatory arrest, does not require multiple operations before definitive repair, and seems to permit correct patient management strategy assignment in the vast majority of cases. For centers wishing to duplicate this experience, it must be remembered that the Giessen group has among the longest and largest experience in the world with the hybrid approach, and subtle insights and judgmental nuances derived from such an extensive experience are often difficult to detect and convey in retrospective analyses.

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