

tient age, underlying cardiovascular disease or use of antiarrhythmic drug therapy, but not so after adjustment for left ventricular ejection fraction, left atrial diameter or duration of atrial fibrillation. We feel that our results justify the conclusion that "patients who undergo electrical cardioversion display a greater degree and longer duration of mechanical atrial dysfunction than those who convert pharmacologically or spontaneously" (1).

We agree with Falk et al. that some of the patients who underwent electrical cardioversion may have had attempts at pharmacologic cardioversion in the past. By the same token, some of the patients who had pharmacologic cardioversion in our study had also undergone previous attempts at cardioversion. Previous history of cardioversion attempts was not specifically monitored in our study; hence, we are unable to comment on the actual number of study patients who had had previous attempts at cardioversion. To say that "the study is predominantly a comparison of patients who responded to pharmacologic agents compared with those in whom pharmacologic cardioversion failed" is therefore erroneous.

To the best of our knowledge, there is only one previous study that showed that duration of atrial fibrillation <14 days (vs. >6 weeks) is associated with faster recovery of atrial function (2). In that particular study, no multivariate analysis was performed to account for differences between groups of patients who had atrial fibrillation <14 days versus >6 weeks. For this reason, we are not convinced that atrial fibrillation duration <14 days should be a standard reference division point for all future studies with regard to atrial fibrillation. Until more definitive information is available on the impact of duration of atrial fibrillation on atrial function recovery, any dichotomization of this variable will have to be "arbitrary." The division of atrial fibrillation duration in our study is identical to the inclusion criterion used by Falk et al. in their study addressing postcardioversion atrial function (3). Due to uncertainty about the exact duration of atrial fibrillation in many cases, it is not feasible to use duration of atrial fibrillation as a continuous variable.

KISHORE J. HARJAI, MD  
FREDDY ABI-SAMRA, MD

Department of Cardiology  
Ochsner Clinic  
1514 Jefferson Highway  
New Orleans, LA 70121-2483

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## Before Predicting Survival in Children With Pulmonary Hypertension and Congenital Heart Disease . . .

In the recent article by Clabby et al. (1), the authors provide very interesting data on the predictive value of hemodynamic data with

respect to survival in pediatric patients with primary and secondary pulmonary hypertension. In primary pulmonary hypertension, such data have been reported earlier (2,3). In patients with plexogenic arteriopathy and congenital heart disease, these data are scarce (4).

The design of this study has led to the pitfalls of highly selected patient population and small sample size in relation to the used analysis techniques, and consequently the results have to be interpreted with great caution. The most serious problem with the study design is the composition of group 2. This group includes patients with repaired and unrepaired heart defects, which leads to two comments on the interpretation of the data:

1. In patients with persistent pulmonary hypertension after adequate surgical repair of a heart defect, hemodynamic variables are comparable to those in patients with primary pulmonary hypertension. In contrast, in patients with unrepaired heart defects or residual shunts, hemodynamic variables cannot be interpreted in the same way. The authors suggest that the variable "mean right atrial pressure (mRAP)  $\times$  pulmonary vascular resistance (PVR)" represents the pressure load on the right ventricle and indicates how well the right ventricle handles that pressure. However, in children with large intracardiac communications, with or without transposition of the great arteries, increased PVR is not simply a pressure load for the right ventricle and ventricular dysfunction does not necessarily lead to increased mRAP.

2. More important, the authors conclude that survival in children with pulmonary hypertension in the presence of congenital heart defects, as predicted by the suggested hemodynamic variables, can be used to determine the optimal timing of lung transplantation. However, before predicting survival or scheduling lung transplantation, the question to be addressed is whether the pulmonary arteriopathy has progressed to the irreversible stage. Pulmonary plexogenic arteriopathy in children with left to right shunts is a reversible disease until a so-called "point of no return" is reached, at which the process has become irreversible, and progression will occur even if the heart defect is corrected (5). In the reversible stage, the treatment of choice will obviously be surgical correction of the heart defect. It can be speculated that the pulmonary vascular disease might have been still reversible in some of the study patients: In 80% of group 2 patients, the heart defect was unrepaired or palliated, or residual defects were present, whereas right to left shunting was present in only 29%. Hemodynamic or histologic evaluation is currently used to assess the progression of pulmonary vascular disease. However, criteria to predict in which patients pulmonary vascular disease will progress despite surgical correction have a broad gray zone (5,6). Therefore, before including these children in a study to predict survival in patients with irreversible pulmonary vascular disease, we should be able to determine the reversibility of this disease process more accurately.

In other words, one does not want to predict the optimal timing for lung transplantation in children who may be cured by surgical correction of their heart disease.

R. M. F. BERGER, MD  
R. GEIGER, MD  
F. J. MEIJBOOM, MD  
*Sophia Children's Hospital  
University Hospital Rotterdam  
Dr. Molewaterplein 60  
3015 GJ Rotterdam, The Netherlands*

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### Reply

We appreciate the interest of Berger and colleagues in our report. We agree with their comments regarding the differences that exist between patients with and without intracardiac defects. These differences almost certainly affect the rate at which symptoms and unstable physiology develop in patients with elevated pulmonary vascular resistance. In interpreting our results, the reader must keep in mind that survival in our study (1) was measured from the date of cardiac catheterization, not from the date of birth or the date of diagnosis. The multivariable analysis takes into account the presence or absence of structural heart disease as a potential predictor of survival. It also takes into account the pulmonary/systolic flow ratio (Qp/Qs)—a reasonable indicator of the presence or absence of a residual shunt lesion, when the pulmonary vein and systemic arterial oxygen saturations are known, as required by our entry criteria. The result of the analysis indicates that the predictive power of the hemodynamic variables is significant at 1 or 2 years of follow-up, with or without structural heart disease and with or without a residual shunt.

We also agree that the child with pulmonary vascular disease must undergo thorough evaluation, including drug testing, to elicit reversibility of pulmonary vascular disease; that a surgically correctable cause of pulmonary hypertension must be sought and excluded in all cases; and that lung transplantation should only be considered when all other therapeutic options have been explored (2,3). The evaluation of children for lung transplantation is extensive, and the result of that evaluation is never determined by any one value. However, it is also clear that a substantial proportion of children with pulmonary hyper-

tension are referred for transplantation when they have very advanced disease and cannot be expected to survive until organs become available (4). Thus, it behooves us to try to determine how we might best predict survival in these patients. We do not argue that every child included in our study was or will be a candidate for lung transplantation; rather, we argue that every child in our study had, by definition, pulmonary hypertension and that their survival at 1 and 2 years of follow-up was significantly related to their hemodynamic status. We suspect that when medical management results in a favorable change in hemodynamic status, it also improves predicted survival, but that question could not be addressed by our study design.

Finally, we agree (as stated in our report) that our study has limitations. Any retrospective, multicenter study must be viewed as less than definitive. We limited our patients to those for whom the data had been collected meticulously enough to be deemed reliable. The sample size is small. Nevertheless, in the context of the existing body of knowledge, we stand by our conclusions. We hope that our work will stimulate increased interest and work in this area—particularly, the evaluation and management of children with nonprimary forms of pulmonary hypertension—and thus to more data and better decision making.

NANCY D. BRIDGES, MD, FACC

*Cardiology Division*

*Children's Hospital of Philadelphia*

*University of Pennsylvania School of Medicine*

*34th Street and Civic Center Boulevard*

*Philadelphia, Pennsylvania 19104*

MARTHA L. CLABBY, MD

*Cardiology Division*

*Saint Louis Children's Hospital*

*Washington University School of Medicine*

*1 Children's Place*

*Saint Louis, Missouri 63110*

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