References

1. Clabby ML, Canter CE, Moller JH, Bridges ND. Hemodynamic data and survival in children with pulmonary hypertension. J Am Coll Cardiol 1997;30:554–60.
2. Sandoval J, Bauerle O, Gomez A, Palomar A, Guerra MLM, Furuya ME. Primary pulmonary hypertension in children: clinical characterization and survival. J Am Coll Cardiol 195;25:466–74.
3. Sandoval J, Bauerle O, Palomar A, et al. Survival in primary pulmonary hypertension: validation of a prognostic equation. Circulation 1994;89:1733–44.
4. Hopkins WE, Ochoa LL, Richardson GW, Trulock EP. Comparison of the hemodynamics and survival of adults with severe primary pulmonary hypertension or Eisenmenger syndrome. J Heart Lung Transplant 1996;15:100–5.
5. Wagenvoort CA. Open lung biopsies in congenital heart disease for evaluation of pulmonary vascular disease: predictive value with regard to corrective operability. Histopathology 1985;9:417–36.
6. Bush A, Busst CM, Haworth SG, et al. Correlations of lung morphology, pulmonary vascular resistance, and outcome in children with congenital heart disease. Br Heart J 1988;59:480–5.

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 hypertension 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.

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References

1. Clabby ML, Canter CE, Moller JH, Bridges ND. Hemodynamic data and survival in children with pulmonary hypertension. J Am Coll Cardiol 1997;30:554–60.
2. Bridges ND, Mallory GB, Huddleston CB, Canter CE, Spray TL. Lung transplantation in infancy and early childhood. J Heart Lung Transplant 1996;15:895–902.
3. Bridges ND, Spray TL. Lung transplantation in children. In: Karp RB, Laks H, Wechsler AS, editors. Advances in Cardiac Surgery, Vol. 8. St. Louis (MO): Mosby, 1996:73–7.
4. Bridges ND, Clark BJ, Gaynor JW, Spray TL. Outcome of children with pulmonary hypertension referred for lung or heart and lung transplantation. Transplantation 1996;62: 1824–8.