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Debates

Rebuttal

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As I had hoped and anticipated, the previous contributions have shown that Len and I are involved in a debate, but not an argument. As I am asked in this rebuttal to respond to some of the very important points that Len has made, I shall do so. I think that he was very accurate in distilling, from the study of outcomes over 15 years in Philadelphia, the message that late attrition was quite low. Of the first cohort in that study, undergoing surgery way back in 1984, less than one-third survived. As those of you who are familiar with the study now, it has improved steadily, so that now at least four-fifths of those undergoing first stage palliations survive. But what is really important is that those children who got out to 5 years usually got out to 10. And nearly all of those that got out to 10 years were alive at 15 years. I have already pointed to evidence that rates of survival with respect to initial surgical palliation are improving steadily at centers throughout the United States of America, and also around the world. So that, were it not for the shortage of donor organs, we could get together again in a decade or two from now and debate the merits of these two forms of therapy based on a comparison of the late morbidities associated with the creation of the Fontan circulation, such as sinus nodal dysfunction, protein losing enteropathy, thromboembolic phenomenons, and so on, to those events occurring at later stages associated with organ replacement, such as graft atherosclerosis, lymphoproliferative disorders, late rejection, and so on.

Another of Dr Bailey's observations that is of considerable interest is that, so far, the incidence of re-transplantation in those undergoing neonatal replacement of the heart at Loma Linda seems to be as high, or higher, than the incidence of late eventual transplantation in the few large series of reconstructive surgery extending over the same period of time. But clearly there is a role for cardiac transplantation among a sub-group of patients treated initially with staged reconstructive surgery who go on to develop poor function of the systemic morphologically right ventricle. And clearly, there is a role for palliative reconstructive surgery even at those centers most strongly committed to replacement of the heart as primary therapy.

But the piece of information shared with us today in connection to cardiac transplantation that is most interesting to me came not from the debate between Len and me. It was a slide shown to us by Mark Boucek of Denver Children's Hospital. They investigated the impact of several variables on neuro-developmental outcome in their very fine program of transplantation for neonates with hypoplastic left heart syndrome. The factor with the greatest negative impact on neuro-developmental outcome was not surgical technique, nor the duration of cardiopulmonary bypass, nor hypothermic circulatory arrest. Instead, the variable that most directly impacted on neuro-developmental outcome was the waiting time to transplantation. This tells us that these neonates with functionally single ventricles, and ductal dependancy of the systemic circulation, are at continuing risk for important morbidity despite the very best supportive medical therapy that we have to offer. So the Norwood procedure has the additional advantage that it can be performed very early in life, usually within a few days of initial diagnosis and stabilization. Only rarely, as in cases with intact atrial septum, is it a surgical emergency. In addition, nonetheless, only rarely must one wait as long as several weeks with supportive medical therapy before proceeding with initial reconstructive surgery.

Dr Bailey also made the very important point that patients with hypoplastic left heart syndrome

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subjected to reconstructive surgery ultimately face a lifetime with a circulation based upon the morphologically right ventricle fulfilling a systemic role. Several studies published in recent years have amplified the observation made by Bill Norwood and myself over 10 years ago,[1] specifically that patients with a systemic morphologically right ventricle were at no disadvantage with respect to early survival after the Fontan operation when compared to those with a systemic morphologically left ventricle. More recently, Spray and associates[2] went on to show that the diagnosis of hypoplastic left heart syndrome is not a risk factor for mortality after the Fontan procedure. Perhaps as importantly, recent studies from the exercise laboratories at the Children's Hospital of Philadelphia,[3] performed on patients operated on by Norwood and myself in the early 1990s, have shown that patients converted to the Fontan circulation who experienced early removal of the volume load from the systemic ventricle, as is the case in virtually all reconstructive surgical patients with hypoplastic left heart syndrome, ultimately perform better under exercise testing than patients with a functionally single ventricle of similar age who underwent surgery to unload the volume of the ventricle later in life.

At this point in the symposium, I indulged myself by showing video tape of a floor exercise and balance beam program performed by a 9-year-old competitive gymnast with hypoplastic left heart syndrome who had undergone staged reconstructive surgery in infancy. When asked about the exercise capacity of patients with the Fontan circulation living at higher altitudes, I showed a video tape of her performance "at higher altitude" on the uneven parallel bars. Never was a presentation tainted by a more anecdotal or less scientific piece of evidence. But the personal energy of this child, her sense of determination and self-esteem, were palpable to all who watched. Children like this one with hypoplastic left heart syndrome are unconvinced that there are finite limits to their quality of life.

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