Cardiorespiratory Responses to Exercise After the Fontan Operation

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Originally described for the treatment of tricuspid atresia, the concept of ventricular exclusion and direction of systemic venous blood from the right atrium immediately into the pulmonary artery has been applied to a variety of forms of single ventricle. Theoretically, this operation should eliminate the right-to-left shunt and decrease the volume of systemic ventricular blood flow. Fontan’s original description included a Glenn anastomosis, incorporation of a valve into the right atrium to pulmonary arterial connection, and insertion of a valve into the inferior vena caval–right atrial junction. There have been numerous modifications of the original operation. Many investigators think that a prior Glenn anastomosis is unnecessary and that insertion of valves into the inferior vena caval–right atrial junction and right atrium–pulmonary artery connection are unnecessary. The presence of a Glenn anastomosis results in less pleural fluid drainage after the Fontan operation. However, development of pulmonary arteriovenous fistulas can result from a Glenn anastomosis. In addition, the larger right lung receives the relatively lesser volume of superior vena caval venous return, whereas the smaller left lung receives the greater volume of inferior vena caval return.

Although the question of whether to include a subpulmonary ventricular remnant into the repair is still being debated, Lee et al could not show a difference in perioperative mortality or morbidity rates between the two techniques, and Rhodes et al could not demonstrate a difference in aerobic capacity and the cardiorespiratory responses to exercise. Of course, one cannot abandon, based on these two studies, the idea that incorporation of the ventricular remnant into the repair is advantageous. At this point, it is unclear whether incorporation of the right ventricle remnant affects the late development of arrhythmias, protein-losing enteropathy, or survival.

A unique aspect of Rhodes et al’s study in this issue of Circulation is the limitation of the selection of patients after Fontan operation for exercise testing to those who are doing well clinically (in New York Heart Association classification I). Prior studies included all patients, regardless of clinical status. One criticism of prior studies was that the reduced aerobic capacity of patients after Fontan operation was due in part to the inclusion of patients who had a less-than-ideal surgical result. The present study demonstrates limited aerobic capacity, even in patients with an ideal operative outcome. Of course, this is not particularly surprising because these patients still have only one functional ventricle; regardless of the quality of the surgical result.

Rhodes and his colleagues have confirmed other investigators’ findings of an abnormal cardiac output response to exercise after the Fontan operation. There is little disagreement that an abnormal stroke volume response, to a large degree, and chronotropic insufficiency, to some degree, account for this. However, systemic stroke volume is dependant on and perhaps limited by the volume of pulmonary blood flow. The absence of an effective subpulmonary ventricular chamber no doubt accounts for relatively reduced pulmonary blood flow. Indeed, our understanding of the determinants of pulmonary blood flow after the Fontan operation is limited. Presumably, respiratory fluctuations of the negative intrathoracic pressure, a skeletal-muscle pump system, and gravitational forces (relative to return of superior vena caval blood) contribute to promoting pulmonary blood flow. Although right atrial contraction contributes to antegrade systemic venous flow, it is not essential after the Fontan operation to provide adequate pulmonary blood flow.

Rhodes et al’s results are consistent with those of other investigators in demonstrating persistent hypoxemia, especially during exercise, after the Fontan operation. The mild hypoxemia persists despite the lack of apparent residual intracardiac right-to-left shunts using echo-Doppler and angiographic techniques. Presumably, this results from persistent intrapulmonary right-to-left shunt. Another potential source of right-to-left shunt exists if the surgeon leaves the os of the coronary sinus drain to the left of the atrial septum, a maneuver that might promote

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improved myocardial perfusion.\textsuperscript{8} The persistent right-to-left shunt accounts for the abnormal ventilatory equivalent for oxygen and ratio of dead space to tidal volume described by Rhodes et al.\textsuperscript{9}

The cardiorespiratory response to exercise after the Fontan operation has been well described; aerobic capacity improves after the Fontan operation but remains subnormal. There are, of course, many other important questions that remain about the Fontan operation. Does it improve long-term survival? Does it improve quality of life? What will be the incidence of serious arrhythmias, protein-losing enteropathy, pulmonary arteriovenous malformations, and pulmonary emboli among patients who have had a Fontan operation?

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