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EDITORIAL

The Fontan Enigma: Ventilating Between Lows and Highs

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The Fontan procedure is the final stage of palliation for patients with a single-ventricle congenital heart defect, or with a heart anatomy not compatible with a two-ventricle physiology. It is routinely performed in centers dedicated to congenital heart surgery.

In the normal circulation, cardiac output (CO) is mainly determined by the cardiovascular system. In contrast, after the Fontan procedure, the CO is highly dependent on the preload of the systemic single ventricle, and thus on the blood flowing through the lungs and a fenestration if present. This transpulmonary blood flow is influenced by both the pulmonary vascular resistance (PVR) and the transpulmonary gradient (TPG) (systemic venous pressure—common atrial pressure) [1].

Postoperative efforts focus on optimizing the transpulmonary blood flow by managing fluid status and PVR. One of the main objectives in the early postoperative period is to minimize the intrathoracic pressure by allowing spontaneous breathing and aiming for early extubation. Indeed, it has been shown that positive pressure ventilation (PPV) after the Fontan procedure impairs CO by increasing PVR and TPG. Even if multiple ventilator settings or devices have been proposed to minimize this side effect, there is no consensus on which intraoperative ventilation strategy should be adopted when it comes to the applied tidal volume (V_T). In this issue of *Pediatric Anesthesia*, Navaratnam et al. [2] assess this point in a randomized crossover trial including 12 cardiac surgical patients after a Fontan operation. Their primary endpoint is to evaluate the V_T effect on the TPG. They hypothesized that a low V_T ventilation strategy (5 mL/kg) compared to a high V_T ventilation (10 mL/kg) would lead to a TPG decrease of 2 mmHg or more. In addition to various hemodynamic and respiratory parameters, transesophageal (TEE) measurements of Fontan flow, systemic stroke volume, and pulmonary blood flow were analyzed as secondary outcomes. For the 10 patients in whom the primary endpoint was measured, there was no statistically significant difference in the measured TPG between the low and the high V_T group. Also, they found no significant differences in the secondary outcomes regarding stroke volume or Fontan flow between both groups. These results were obtained despite significantly higher peak and higher mean airway pressures in the group with high V_T ventilation.

Navaratnam et al. are to be complimented for having conducted this elegant study. However, their findings warrant some reflection in addition to the limitations highlighted by the authors. First, there is a wide variation in patients' ages ranging from 3 to 42 years. With advancing age, the cardiopulmonary physiology of single-ventricle patients changes. The Fontan procedure itself modifies the cardiopulmonary physiology. There are thereby substantial differences between a newly created Fontan circulation in a child and an already existing Fontan at a later age. Over time, there is a steady increase in systemic venous pressure, ventricular filling pressure, and most importantly PVR [3, 4]. Moreover, pulmonary abnormalities and lung function alterations are common in older Fontan patients. Adult Fontan patients have been shown to have reduced lung volumes with restrictive lung disease, resulting from different causes such as postural deviations, previous thoracotomies, number of surgeries, denutrition, scoliosis, and diaphragmatic dysfunction [5]. It is therefore plausible that PPV would induce more pronounced effects in older Fontan patients. Another issue is that there is a maturational change in the contribution of the superior vena caval flow to total cardiac output, and the distribution of venous return from the superior vena cava and inferior vena cava varies depending on age. A Doppler echocardiography study in 145 healthy children showed that the superior vena cava

contribution to CO will gradually decrease to reach adult values of 35% by age 6,6 years [6]. The effect of PPV on TPG and CO may therefore be less obvious in children compared to adults. All together, the results of this study should not be extrapolated to Fontan patients of all ages.

Second, Navaratnam et al. [2] showed a significant difference in mean and peak airway pressure between both groups. The lack of hemodynamic response despite these differences may indicate that the TPG is mediated mainly through significant changes in intrathoracic pressure and not airway pressure per se. As mean airway pressure is the area under the curve of the pressure versus time graph and as pulmonary blood flow is a continuous process occurring during inspiratory and expiratory phases, the effective mean airway pressure to which the patients were exposed during the 5′ ventilation sequence was the same in both groups, the lower mean airway pressure in the low V_T group being compensated by an increased respiratory rate in order to achieve the same minute ventilation. Third, Navaratnam et al. [2] compared low and high V_T ventilation, which both can negatively impact PVR and Fontan flow. Whether a ventilation strategy with a V_T of 7 mL/kg would result in the lowest PVR should be further evaluated.

This editorial is therefore a call for more research—not just on ventilation strategy, but on the essential principles of cardiopulmonary interactions governing Fontan physiology. In this unique physiology, every intervention—no matter how small—should compel us to a critical reassessment of the dogmas.

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The authors have nothing to report.

Conflicts of Interest

Mona Momeni is an associate editor for the journal *Pediatric Anesthesia*. Thierry Detaille has no disclosures.

Data Availability Statement

Data sharing not applicable to this article as no datasets were generated or analyzed during the current study.

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