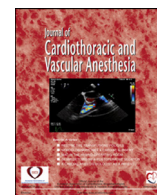


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Editorial

Assessing the Fontan Parturient; One Size Does Not Fit All



This invited editorial is related to: Ref.: Ms. No. JCVA-D-22-00216R2 Case series conference: Anesthetic management in parturients with Fontan physiology

“If you have choices choose the best. If you have no choices do your best!” Nishan Panwar

FONTAN SURGERY has been used to palliate a heterogeneous group of congenital heart lesions that share in common a functionally single ventricle. The introduction by Francis Fontan in the late 1960s was a major improvement over the then-available procedures, and greatly enhanced overall survival in this patient population. The resulting single-ventricle physiology is complex, and far from uniform, ranging from individuals with almost normal exercise capacity to patients who are severely ill with multiorgan dysfunction also referred to as “failing Fontan.” However, with advancements in surgery and overall care, the contemporary expected 30-year survival is now >80%, an estimated 70,000 patients are alive worldwide, and it is easy to predict that this population will increase significantly in the near future.¹ Thus, an increasing number of women with Fontan palliation will reach child-bearing age. The ethical challenges of preconception counseling and decision-making are significant, with increased maternal and fetal risks throughout pregnancy, delivery, and extending into the postpartum period. Thus, evidence-based guidance on how to best care for the parturient with such complex physiology is of the essence.

In this issue, Clark et al. reported a retrospective single-center case series reporting the maternal outcomes of 8 parturients with Fontan physiology following 13 pregnancies that resulted in live births from 1999 to 2020. In their series, 77% were delivered via caesarian section, with the remainder being forceps-assisted vaginal deliveries. All patients received neuraxial anesthesia (combined spinal-epidural, or epidural) with conversion to general anesthesia required in one patient due to a failed block. Most notable was the high rate of complications (54% of pregnancies) reported in this series, including 2 pregnant patients experiencing cerebrovascular accidents, 2 with

worsening cyanosis, and 1 with an acute reduction in ejection fraction. Postpartum hemorrhage occurred in 23% of pregnancies, and was reported to have occurred largely due to placenta accreta, abruption, or infarction.

The findings of this case series add to the available outcomes data for parturients with Fontan circulation, with some obvious limitations due to the small sample size. In a recent larger systematic review, Garcia Ropero et al. summarized the outcomes from 6 studies in parturients with Fontan circulation, including 255 pregnancies in 133 women, resulting in 115 live births.² The most common complications reported in this larger series were postpartum hemorrhage (14%), supraventricular arrhythmia (8.4%), and heart failure (3.9%).² In another recent large meta-analysis of maternal and neonatal outcomes of pregnancies in women with CHD, the rate of neonatal mortality and maternal morbidity increased with the severity of CHD.³ The subgroup analysis in Fontan parturients found arrhythmias (10.3%), heart failure (6.1%), and postpartum hemorrhage (11.2%) as maternal adverse events.³ For neonatal adverse events, maternal history of Fontan palliation carried the highest risk for cesarean section, miscarriage/spontaneous abortion, preterm delivery, and small for gestational age.

There seems to be a surprising consistency in the reported maternal and neonatal adverse events in Fontan parturients. The question for the anesthesiologist involved is how anesthetic management can help mitigate or help prevent some of these known adverse events. Unfortunately, what these reports have in common is that good evidence-based data to help guide decision-making with regard to the anesthetic management in the Fontan parturient are, for the most part, lacking. There are many variations in the underlying congenital heart disease anatomy, pathophysiology, and surgical technique, as well as individual adaptation to the Fontan physiology with all possible sequelae. The complexity and variability of post-Fontan long-term outcomes and disease progression are far too complex to allow making generalized recommendations for anesthetic management based on these few observational and practice-based studies. For example, in the study by Clark et al., all 8 parturients had a systemic left ventricle, with mostly

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preserved function. In contrast, the preoperative information available in the systemic review by Garcia Ropero et al. reported pregestational cardiovascular issues including arrhythmias, significantly reduced ejection fraction, and heart failure in many patients.² It is important to consider the heterogeneity in pathophysiology that accompanies patients with Fontan in the evaluation of peripartum risk, and how this relates to anesthetic management. For example, it is obvious that the described anesthetic management of patients in the series of Clark et al with almost normal ejection fraction will be different from patients with preexisting cardiovascular compromise including severely decreased cardiac function. It unlikely is based on the overall low number of patients who can be studied, as well as ethical concerns, that good data derived from prospective, randomized, controlled studies will become available any time soon.

As a first step, evaluating a Fontan parturient for anesthesia management should include a valid risk assessment model specifically for patients with Fontan, which captures the heterogeneity of disease and long-term sequelae in this patient population. The anesthesia provider should consider the overall long-term outcomes data for this cohort. Data from The Australian and New Zealand Fontan Registry have provided a comprehensive look at morbidity and mortality in adults with Fontan circulation.^{4,5} Patients with atriopulmonary Fontan have the worst long-term prognosis, including overall survival, Fontan failure, arrhythmia and the need for pacemaker placement, as well as reintervention.⁴ Time since the Fontan creation should be considered also in the risk assessment, as the rate of long-term complications increases with time since operation.⁶ The systemic ventricle, function, and aortic valve regurgitation all have to be considered.^{4,7}

Another point eloquently highlighted by the commentaries in the series by Clark et al is the multidisciplinary approach. Less commonly discussed though, and highly relevant for anesthetic management, is the reported high rate of postpartum hemorrhage. It seems contradicting, that patients with Fontan who have an increased risk of thrombosis often requiring life-long anticoagulation also have an increased risk of bleeding. This is yet another hint highlighting the complexity of patients with Fontan. Pregnancy is a hypercoagulable state and affords an increased risk for thrombosis, and, yet, Fontan parturients are at risk for both thrombosis and hemorrhage. Therefore, anticoagulation management at the time of delivery, and in particular how it affects anesthesia management, is complex weighing the risk of peripartum hemorrhage versus thromboembolic events. Additionally, patients with elevated Fontan pressures often experience exaggerated surgical blood loss due to the high venous pressures, which may be of particular consequence in patients requiring cesarean section.

The anesthetic care of patients with Fontan is rather complex, and ideally should be reserved for providers routinely caring for adults with congenital heart disease. However, given the growing number of patients surviving with single-ventricle physiology desiring pregnancy, it is likely that the management of these parturients in more diverse settings will increase. In the absence of evidence-based best practice recommendations, all we can base our anesthetic management on are observational data and comprehensive reviews on perioperative anesthetic considerations for patients with Fontan.⁸ Without clear guidance on what best choice anesthetic to pick, we must do our best as anesthesiologists to help improve outcomes in patients who are high risk and their newborn children.

Conflict of Interest

None.

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