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Review

Anesthetic and perioperative care of high-risk adults with congenital heart disease: Managing ventricular dysfunction and minimal reserve

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ABSTRACT

The number of adults with congenital heart disease has increased dramatically in recent years owing to advances in the surgical and medical care of pediatric congenital heart disease, and these patients present with increasing frequency for both cardiac and non-cardiac surgical interventions. By adulthood, many demonstrate varying degrees of ventricular dysfunction and are at high risk for perioperative morbidity and mortality. Due to their limited cardiopulmonary reserve, physiologic insults are poorly tolerated and must be minimized throughout the perioperative period. An understanding of the altered cardiopulmonary physiology, unique complications, and overarching management goals for ACHD patients will allow anesthesiologists to coordinate optimal perioperative care for this challenging patient population.

1. Introduction

Over the past four decades, increasing success in the management of pediatric congenital heart disease (CHD) has resulted in dramatic increases in the number who survive to adulthood [1], and adults now outnumber children with CHD [2], accounting for approximately 66% of the overall CHD population [3]. Since 2000, the prevalence of CHD has increased most dramatically in patients 26–40 years old [3]. But many CHD lesions are treated with long-term palliative strategies that are not truly curative, and many adult congenital heart disease (ACHD) patients require cardiac reintervention and eventually develop progressive heart failure as a late sequela of their initial CHD.

These patients are at high risk of morbidity and mortality when they present for cardiac reoperations, electrophysiology interventions, noncardiac surgery [4], and obstetric care [5]. Anesthesiologists will encounter these patients with increasing frequency, and provider survey data suggest that anesthesiologists have limited knowledge of and comfort with this patient subgroup [6].

This review will discuss an approach to the perioperative management of high-risk ACHD patients: those with compromised ventricular function and limited physiologic reserve. It will include a discussion of specific anesthetic agents, but it will focus on a more comprehensive approach to managing these patients within the model of the anesthesiologist as the coordinator of complex perioperative concerns for challenging patients.

2. The paradigm of limited reserve

There is a widely held but mistaken impression about the care of high-risk cardiac patients, namely that there is some ideal or benign combination of medications that can guarantee a safe anesthetic. Cardiac anesthesiologists and others who frequently care for high-risk patients have access to the same pharmacologic tools as they do when they care for low-risk patients.

The core challenge for high-risk patients is to devise an anesthetic strategy that minimizes the chances of destabilizing a patient with minimal reserve. With few exceptions (i.e. patients presenting in extremis for an emergent intervention), even the sickest patients with the most severely depressed ventricular function present for an anesthetic having achieved some homeostatic equilibrium of adequate circulatory function. The key difference between such a patient and a patient without cardiac disease is the physiologic reserve that he or she has to tolerate additional insults during the perioperative period: hypoxia, hypercarbia, hypotension, pain, sympathetic surges, abnormal pressure conditions (positive pressure ventilation, pneumoperitoneum, etc.), blood loss, arrhythmias, and fluid shifts. In caring for these high-risk patients, we are tasked not with fixing their tenuous – though, at the moment, adequate – circulation, but with putting them in the best possible position to enable that circulation to withstand those perioperative stressors.

3. The risk: understanding the dangers in ACHD

The general and not surprising observation has been made that taken as a whole, ACHD patients are at greater risk of morbidity and mortality during an anesthetic, even for noncardiac surgery [4],

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and ample anecdotal evidence exists to demonstrate the potential perils of anesthetizing ACHD patients without appropriate forethought and preparation. However, richer outcome data to risk-stratify patients and guide interventions to reduce risk are lacking. Even in the absence of robust data, though, expert opinion provides some guidance.

One may recognize that for many CHD lesions, cardiovascular decompensation is not a matter of "if" but "when." In all but the simplest defects, surgical interventions that restore a phenotypically normal circulation and that are described as a "fix" or a "repair" are more accurately conceptualized as highly effective and stable palliative procedures, and while compromise may not become evident for several decades, even moderately complex CHD may result in substantial morbidity in adult life. Most patients with a systemic right ventricle, complex structural CHD, and especially those with single-ventricle physiology can expect eventual cardiovascular decompensation. This point was made early on by Dr. Fontan himself, who described in a 1990 paper that the expected outcome after a "perfect Fontan" operation would include a substantial risk of early mortality, a long plateau phase of clinical stability, and then a late failure phase beginning 10–15 years from the Fontan procedure in which a significant proportion of patients succumb to the eventual consequences of the abnormal circulation that has maintained them adequately for many years (see Fig. 1) [7]. In this paradigm, a failing circulation is not a sign of substandard care or unusual decompensation — but an expected (if poorly temporally predicted) eventual occurrence.

The challenge in managing ACHD patients who have begun to show either subacute or overt evidence of decompensation, then, is to recognize where the patient is on the spectrum of that downward trajectory and identify the best strategies to support them.

Cardiac dysfunction and end-organ compromise often are underrecognized in ACHD patients, in part because of a self-conception that they have no functional limitations, which may result from their youth, tendency to regard themselves as normal and healthy, and their lack of any comparative life experiences with normal cardiac physiology. While substantial functional impairments exist in the ACHD population [8], patients underappreciate and underreport them [9]. For instance, ACHD patients underreport exercise limitation, and substantial impairment is often apparent if assessed with exercise testing. A cohort of 335 ACHD patients followed at the Royal Brompton in London (average age 33 years, with the majority describing themselves as "asymptomatic") demonstrated an average peak oxygen consumption of only 48% of that seen in age-matched controls without ACHD. This impairment was on par with that demonstrated by a comparison cohort of acquired heart failure patients (average age 58 years) [10]. Likewise, end-organ compromise is common: for instance, restrictive lung disease [11] and renal insufficiency [12] each have been demonstrated in approximately 50% of a broad panel of young ACHD patients, the majority of which described themselves as fit and healthy.

In particular, the systemic RV and single-ventricle patients may have less reserve even in the presence of a baseline echocardiogram that shows normal resting systolic function. Stress echocardiography and exercise testing may provide additional information about reserve, but is not always logistically possible or clinically appropriate, especially if one cannot demonstrate how it will change management.

3.1. The systemic right ventricle

A number of patient subgroups have a systemic right ventricle, with the morphologic right ventricle (RV) of a two-chambered heart functioning as the pump to the systemic circulation. Such patients include those with L-transposition of the great arteries, D-transposition treated with Senning or Mustard atrial switch procedures, and certain patients with double-outlet right ventricles or

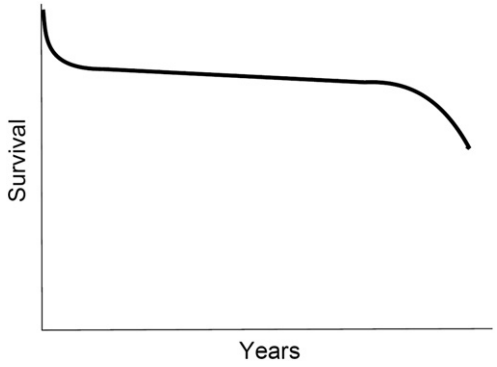

Fig. 1. Schematic representation of the pattern of survival after complex palliation or single-ventricle repairs. Note: this figure is intended to represent qualitative patterns shared by many lesions and not exact quantitative survival statistics, which vary by patient group and time period.

heterotaxy syndrome. By the time that they reach adulthood, the majority of these patients will display some amount of systolic dysfunction, volume overload, tricuspid regurgitation (TR), and/or arrhythmias. The largest study to date of adults with systemic RVs found that 68% had RV dysfunction, 64% had TR, 20% had tachyarrhythmias, and the average RV end-diastolic volume was 40% greater than the average LV end-diastolic volume [13]. Despite these marked physiologic perturbations, most of these individuals were outpatients who had not required repeat operative interventions. The high frequency of significant circulatory abnormalities seen in this seemingly well-compensated patient subset demonstrates the often tenuous status of the systemic RV patient and ease with which they may be tipped toward instability following even small perioperative insults.

Optimization of heart rate, tight regulation of volume status, and preservation of inotropic state are cornerstones of perioperative management for systemic RV patients. Maintenance of sinus rhythm is critical whenever possible, as arrhythmias may lead to critical reductions in RV filling. Hypotension should be avoided and RV perfusion maintained, as any offense to an already tenuous RV may lead to a challenging cycle of decreased ejection, further dilation, worsened TR, and decreased cardiac output. Patients with previous atrial switch procedures are also at risk for paradoxical emboli during conditions of RV compromise, as elevated right atrial pressures can convert a stable, small left-to-right shunt to a right-to-left shunt via baffle leaks.

3.2. Patients with a single ventricle

As a direct result of the expansion of the Fontan procedure from limited use for tricuspid atresia in the 1970s to widespread use for a range of lesions with improved survival, the adult Fontan population has grown dramatically over the past decade. As post-Fontan children grow into adulthood, they are prone to predictable complications of their altered circulation including marked right atrial dilation, atrial arrhythmias, baffle leak, pulmonary venous obstruction, and baffle or conduit obstruction [14,15]. These complications not infrequently necessitate repeat operative intervention for conduit revision or, more recently, conversion to an extracardiac conduit. Well-compensated adult Fontan patients will also increasingly present for non-cardiac interventions and warrant close management by the anesthesia provider.

Maintaining an adequate transpulmonary gradient and preventing increases in pulmonary vascular resistance (PVR) are of upmost importance in the adult Fontan patient. Blood flow from the right atrium to the pulmonic circulation is entirely passive in