Perioperative Challenges in Adults with Congenital Heart Disease: A Pediatric Perspective.

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In his assessment of the current health care of adults with congenital heart disease (CHD), Dr. G. Webb stated the following [1]. “The care of adult patients with congenital heart defects in the United States is spotty at best, and needs to improve greatly if the needs of these patients are to be met. The care of American children with congenital heart defects is generally excellent. Pediatric cardiac services are well established and well supported. The care of adults with congenital heart disease (CHD) is well established in only a few American centers. While there are an increasing number of clinics, they are generally poorly resourced with relatively few patients. If located in adult cardiology programs, they are usually minor players. If located in pediatric cardiac programs, they are usually minor players as well. Training programs for adult CHD (ACHD) caregivers are few, informal, and poorly funded. To improve the situation, we need perhaps 25 well-resourced and well-established regional ACHD centers in the United States.” Most ACHD centers in North America and Europe do not meet their national guideline standards [2-7].

Currently, a sizable proportion of North American institutions that self-report to be ACHD centers are associated with pediatric hospitals (International Society for Adult Congenital Heart Disease, http://www.achaheart.org/). Most outpatient and inpatient services and surgery are conducted in facilities that provide both pediatric and adult care [2]. A recent pan-Canadian survey of quality of ACHD care reported 8 of the 15 ACHD clinics were part of a university center that had an established pediatric cardiac surgery program. These ACHD clinics had larger numbers of yearly clinic visits, annual percutaneous volumes and surgical volumes compared to clinics without an affiliated pediatric cardiac surgery program at their site. They also tended to have larger numbers of registered patients, larger numbers of active patients, more dedicated nursing support and more physicians with ACHD Fellowships [3].

National practice guidelines generally concur regarding the provision of anesthesia services to ACHD patients. “Surgery in ACHD patients (including anesthesia and intensive care) is very different from conventional adult cardiac surgery, and this provides a strong case for concentrating resources into specialist units for both treatment and training. It should be appreciated that even minor non-cardiac surgery may carry a high risk; consultation with specialists, and careful preoperative planning and intraoperative monitoring, are vital to avoid complications [8].” “Surgical procedures that require general anesthesia or conscious sedation in adults with moderate or complex CHD should be performed in a regional ACHD center with an anesthesiologist familiar with ACHD patients. An ACHD center should have several cardiac anesthesiologists [9].”

From a pediatric cardiac anesthesiologist’s perspective, optimal perioperative management requires an understanding that ACHD patients are not simply “big children with CHD”. These two patient groups are dissimilar for the following reasons. (i) Adults are naturally different from children; (ii) ACHD presents some unique management challenges; and (iii) non-CHD comorbidities differ between adults and children.

When children with CHD attain adulthood and independence, they have to tackle a variety of new life-issues. Transition of medical care refers to a shift in the responsibility of health care management from the family to the patient. Often this is poorly done and there is substantial loss of patients to follow-up [10]. Specialized ACHD clinics are hard to find and waiting times for ACHD services may be long [3]. Non-compliance with medical therapy is a common problem. Adults with CHD may have felt unwell their entire life and may suffer from depression [11]. Perceived parental overprotection is associated with heart-focused anxiety in adults with CHD [12]. Older patients may be living with the diagnosis of incurable heart disease. Many adults with congenital heart disease have a specific need for advice about socio-legal questions (insurance, provision for old age, severe disability), types of education (school, study, occupation), employability, physical resilience (e.g. performance ability, sporting activity), acquisition of a
driver’s license, suitability to fly and often also issues concerning sexuality, pregnancy and inheritance of heart defects [13]. Many patients wish to discuss end of life matters with their health professionals [14]. Awareness of these psycho-social factors is important when interacting with the ACHD patient.

There are several reasons why CHD in adults is not the same as CHD is children. Some elderly patients with Eisenmenger’s syndrome have unrepaird septal defects that were deemed inoperable during an earlier era. Certain lesions (eg: recurrent pulmonary vein stenosis, hypoplastic left heart syndrome) have (or had) a high infant mortality and are under-represented in the current adult population. Adults may have undergone surgical procedures that later fell from favor. Examples include the atrial switch (Mustard, Senning), Potts’ and Waterston aorto-pulmonary shunts and the atrio-pulmonary Fontan. Adults are likely to develop the late cardiovascular complications of CHD because their hearts have been subjected to multiple cardiac surgeries and have endured abnormal pressure and volume loads, cyanosis, hypoxia and dysrhythmias for a long time. For example, the prevalence of atrial fibrillation and ventricular arrhythmias increases markedly after 45 years of age in patients with repaired Tetralogy of Fallot [15]. In earlier times, biventricular repairs (eg: Tetralogy of Fallot) were delayed until late childhood and this may limit the potential for ventricular remodeling. There are gender-related differences in CHD that are less apparent in children. For example, women with CHD are more likely to develop pulmonary hypertension and less likely to have infective endocarditis [16].

Complications commonly associated with ACHD include heart failure [17], arrhythmia [18, 19], infective endocarditis [20], impaired pulmonary function [21], pulmonary hypertension [22], neurological impairment, cerebrovascular accidents [23] and thromboembolic events. Some patients (for example, those palliated with the Fontan procedure) are prone to other organ injury such as liver, gut or kidney dysfunction from poor systemic output, chronic venous congestion, hypoxia, drug effects, abnormal coagulation, sepsis and other factors. There may also be complications related to surgery, other invasive diagnostic and therapeutic cardiac procedures and heart or heart-lung transplantation [24]. CHD is associated with non-cardiac anomalies and genetic disorders and there may be complications related to these conditions. For example, patients with Down syndrome have an increased risk of Alzheimer’s disease [25].

In addition, adults with CHD may have concomitant non-cardiac disease such as obesity, hypertension, coronary artery disease, diabetes mellitus and chronic kidney disease.

Considering the factors outlined above, it is not surprising that ACHD guidelines recommend that the anesthesiologist should have expertise in adult cardiac anesthesia and the management of CHD. Physicians with this expertise are in short supply and may not be available. In such a situation, pediatric cardiac anesthesiologists are often asked to provide ACHD care because they are familiar with the management of CHD. Currently, it appears cardiac surgery outcomes are best when a congenital heart surgeon performs the heart surgery [26-28]. Some studies suggest the cardiac surgery should occur in a pediatric hospital [26] while others note a high incidence of perioperative complications in the pediatric setting [29] and a higher mortality compared to care in an adult hospital [28]. Information is lacking about outcomes after non-cardiac surgery in ACHD patients.

Typically, a pediatric cardiac anesthesiologist becomes involved because (i) the adult patient with CHD is undergoing a cardiac procedure, performed by a CHD specialist (surgeon or cardiologist) at a pediatric hospital; or (ii) the pediatric anesthesiologist has been asked by his adult hospital colleagues to assist in the management of an ACHD case that will be performed at an adult hospital. When the procedure is to occur in a pediatric hospital, the pediatric anesthesiologist should comprehensively review the ACHD case and decide whether she/he is comfortable managing its non-pediatric aspects. If necessary, an anesthesiologist with adult expertise should be consulted. This is, of course, labor intensive but seems an appropriate strategy until each ACHD center has built up a cadre of experienced ACHD anesthesiologists.

The perioperative management of ACDH patients in a pediatric hospital requires considerable pre-planning and coordination of effort. All equipment and pharmaceuticals should be appropriate for the patient’s size and age. Special attention should be paid to items that are seldom used but important - for example, code carts, ECMO backup, ICU emergency sternotomy sets, etc. Nursing, physician and staff
education and collaboration are vital. With proper preparation, care for ACHD patients at our pediatric hospital has anecdotally been a positive experience for patients and providers; others report a similar finding [30].

More problematic for our institution, is the sharing of patient management between the adult and pediatric hospitals. Although physically connected, there are many impediments to seamless transfer of care. These include cumbersome administrative processes, non-compatible computer systems, limited accessibility to patient data and electronic documentation, different hospital equipment and pharmacy support, dissimilar clinical protocols and styles of practice. Communication channels between pediatric and adult services may be mediocre because they are infrequently used. To overcome these issues, it is our practice to involve an adult cardiac anesthesiologist in a pediatric hospital procedure if the ACHD patient is to receive early postoperative care in the adult hospital. Again, this is labor intensive but well worth the effort.

Many ACHD patients will require anesthesia for non-cardiac procedures. There appears to be minimal indication for performing these procedures in a pediatric hospital. Far better to use adult hospital facilities and adult surgeons and anesthesiologists, with a pediatric cardiac anesthesiologist consulted on CHD perioperative management.

Like many other ACHD centers, the process at our institution (Stanford University, CA) is still evolving. The ACHD clinic is located within the adult hospital and staffed with ACHD-trained cardiologists. ACHD cardiac surgery is performed at the pediatric hospital by the pediatric CHD team. After initial postoperative care by pediatric cardiologists and intensivists in the pediatric CVICU, patients are transferred to an adult hospital ward and managed by ACHD cardiologists. Other invasive procedures are done at the adult hospital with collaborative involvement of CHD and adult specialists. The longer-term proposal is to move all management to the adult hospital and utilize pediatric CHD expertise as required.

A recent editorial [4] comments that it will be a long journey before reaching the end of the road to better ACHD care. Although most agree there is a need to strengthen the human resource infrastructure of ACHD care in the USA, how and when that will occur remains unclear. Until there is a full complement of personnel competent at ACHD care, it will often be pediatric subspecialists (including pediatric cardiac anesthesiologists) who are asked to provide their services.

References


