Young Adult with Failed Fontan Circulation and Symptomatic Tachycardia:…….. *What is Your Next Move?*

There have been many dramatic changes in the ACHD population over the last 15 years. Continued growth and development of centers of excellence in the subsequent decades created a population of teenagers and young adults living with congenital heart disease.(Webb) [1] Care for the patient >18 years is divided amongst pediatric centres that continue to manage the cardiac lesions of adults living with congenital heart disease and established ACHD centers that accept transitioned patients. Regardless of the model, across North America there are insufficient resources at a provider level and many patients >18 years live far removed from a center, have lack of insight into the palliative nature of their paediatric cardiac surgery and perceive themselves as well or 'fixed', and have inadequate personal resources or insurance to regularly pursue care.(Reid and others) [2], (Wacker and others) [3], (Gurvitz and others) [4] In the case of the Fontan patient, the first presentation in several years to any health care institution frequently is the new onset of a tachyarrhythmia.

Fontan survivors represent the extremes of congenital cardiac care. They reflect the great successes of innovation and commitment in pediatric surgical centers across North America and yet we have created a generation of young adults with expectations of a normal life including post-secondary education and long term relationships including marriage and parenting. (Balint and others) [5]ACHD patients are a resilient group and their quality of life measures are correlated to their NYHA classification and "what they can do" rather than their specific anatomical defect. (Bang and others) [6], (Fteropoulli and others) [7] As more ACHD patients pass the threshold of 18 years and develop Heart Failure, patients will be very motivated to pursue anti-arrhythmic therapy to improve their quality of life.(Kovacs and Moons) [8], (Apers, Luyckx and Moons) [9]

In an adult practice, patients with Fontan and Glenn shunts with single ventricle physiology have been, for the most part, patients with tricuspid atresia, pulmonary atresia and DILV; morphological left ventricles as their systemic ventricles. Increasingly adult cardiac anaesthesiologists will see survivors past 18 years with the hypoplastic left heart syndrome (HLHS) living with systemic morphological right ventricles.(Iyengar and others) [10], (Hebson and others) [11] The two cohorts will present with heart failure over the decade, as the experience with systemic RV is that they fail in the 3rd and 4th decade of life, as we know with L-TGA and Mustard/Senning repairs of D-TGA.(Dobson and others) [12]

Fontan and Baudet first described the surgical correction of Tricuspid Atresia in 1971. (Fontan and Baudet) [13] Modifications include the lateral tunnel and extra cardiac conduit which both create a total cavopulmonary connection. There are several case series of successful conversion from conventional RA to PA connection to a cavopulmonary connection with concomitant arrhythmia surgery, with many of the studies having patients with a median age of late teens and early adulthood.(Mavroudis and others) [14], (Mossad, Motta and Vener) [15] The indications are refractory arrhythmias and hemodynamic anomalies. The goal is to improve the haemodynamics and loss of energy in the flow into the PA, and to prevent atrial dilatation and subsequent arrhythmias. Previously patients traditional RA to PA connections presented with right sided arrhythmias (flutter) and due to a dilated RA. With the

cavopulomnary connection the failing systemic ventricle and systemic AV valve regurgitation will produce more patients with fibrillation.(Mavroudis and others) [14]

Whether this lessens the burden of atrial arrhythmias and sudden death in later decades remains to be seen, as this population has had follow up for 10 to 15 years, and although 85% are free from SVT recurrence, they have not been followed as long as the previous version of the Fontan procedure.(Deal) [16]. A large follow up cohort in Boston found no difference in long term survival between RA to PA connection Fontans vs. total Cavopulmonary connections.(Khairy and others) [17]

The challenge for an anesthesiologist involved in the resuscitation of lateral tunnel Fontan or an extra cardiac connection is that there is no access to the atria and ventricle with a central line or temporary pacing wire. The burden of arrhythmias in this population will continue to rise and patients will present for cardioversion, EP mapping and ablation, and arrhythmia surgery in conjunction with conversion to a Cavopulmonary connection.

Patients with lateral tunnel Fontans and extra cardiac connections will have limited or no access to the atria and ventricles. Transconduit/baffle mapping is possible with radiofrequency transeptal techniques.(Correa and others) [18] A large series from Boston reported 50 of 89 adult Fontan patients receiving RFA transbaffle access had deaths(2) and adverse events which included hemoptysis (1), AKI needing ICU support(3) and persistent cyanosis form right to left shunting across the transbaffle access (2).(Correa and others) [18] This series used general anaesthesia whereas other centers provide conscious sedation. The procedures are lengthy for both the anesthesiologist and the patient, as the patient must not be too heavily sedated so that the EP physician can map the arrhythmia, and yet should be comfortable to remain still for several hours.

Cardioversions are fraught with complications, as there is no possibility of floating a temporary wire in the event of conversion to a bradyarrhythmia, and resuscitation drugs must traverse the pulmonary vasculature in the situation of manual positive pressure ventilation and hemodynamic collapse. An experienced adult cardiac anesthesiologist will have wide bore IV access and a clear plan A and Plan B mapped out for each cardioversion. Ideally cardioversion should occur in a central location in the hospital where ancillary personnel can be quickly recruited for assistance and supervised directly by senior staff. The ability to trans-cutaneously pace is an absolute. Ideally the procedure will occur in an environment where a cardiac surgeon is readily available. It is essential to rule out thrombus via TEE prior to the cardioversion with the cardiac anesthesiologist in attendance for both the TEE and cardioversion. Rate control may be the only option if thrombus is seen. Community physicians are challenged with the urgent/emergent presentations for rapid afib or aflutter and may be compelled to cardiovert the unstable patient with asymptomatic thrombus with disastrous outcomes neurologically.(Balling and others) [19], (Oechslin and others) [20]

Adults with congenital heart disease compromise only 2% of the heart transplants reported to the United Network for Organ Sharing in the United States from 1987 to 2006. In this report the 5 year survival in the Fontan patients was 60%, and 74% in the non-Fontan patients. (Patel and others) [21] A series of cardiac transplantation after the Glenn or Fontan procedure found the 30 day mortality of

single ventricle transplants to be very high (28%) and yet those that survive the initial postoperative period have favourable 1 year (71.5%) and 5 year (67.5%) survival.(Jayakumar and others) [22] The competition for the scarce resources with acquired heart failure patients will make transplant as a destination unlikely for most of this population. Many of the patients will have exclusions that will prohibit them for consideration for heart transplant, and if listed will spend a longer time waiting for a 'non-lung' donor due need of increased length of donor vessels.(Burchill and Ross) [23]

The proportion of the ACHD population living with heart failure in the third and fourth decades will increase and the anesthesiologist will be providing care for procedures that may not be cardiac surgery but cardiology interventions for improving their quality of life and lessening the burden of arrhythmias. Congenital cardiac anesthesiologists must show leadership in consulting and advising the adult cardiac anesthesiology community in providing care for this rapidly expanding and complex population.

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