Fontan-type operations in patients with functionally univentricular hearts have been performed for almost 40 years. Since the first description of the Fontan operation, short- and medium-term operative survival has been good in the current era due to careful patient selection, modifications in surgical techniques, and advances in postoperative care. However, a number of serious long-term complications affect most patients. Nonetheless, the prevalence of univentricular patients is increasing as the number of late survivors increases. Caregivers will therefore be evermore faced with the challenge of recognizing and managing the Fontan related complications and patients with failing Fontan physiology.

This review discusses the current surgical management of patients with functionally univentricular heart (UVH) and long-term sequelae of the univentricular circulation based on literature on outcome after the Fontan procedure.

**Introduction**

Birth prevalence of children with a functionally univentricular heart has recently been reported to be 3 to 4 per 100 of children born with congenital heart defects, yielding a birth prevalence of 0.2 to 0.3 per 1000 live births.\(^1\) The definition of this heterogenic patient group has been much debated, using either anatomical or functional definitions. Much of the surgical literature over the last 30 years, however, uses a functional definition of UVH; namely, that UVH is characterized by hypoplasia of one of the ventricles making it incapable of independently supporting either the pulmonary or the systemic circulation or making surgical biventricular repair impossible.\(^2\) UVH might be divided into: single left ventricle in which the right ventricle is rudimentary (e.g. double inlet left ventricle (DILV)), tricuspid atresia (TA), severe Ebstein’s Anomaly, and some patients with pulmonary atresia with intact ventricular septum (PA/IVS) and single right ventricle (e.g. hypoplastic left heart syndrome (HLHS), mitral atresia (MA), and complex forms of double outlet right ventricle (DORV)).

The natural history of UVH is generally poor with a reported survival rate of less than 50% at one year and 10% at ten years.\(^3\) However, the introduction of palliative treatment as originally described by Fontan and Baudet in 1971\(^4\) has dramatically changed the short- and medium-term prognosis for UVH.\(^5\) The technique has evolved and is today applied as a Total Cavo-Pulmonary Connection (TCPC).\(^6\)

**Surgical Management**

The TCPC operation is generally performed in three stages.\(^7\) The concept of staging and improved surgical techniques have reduced morbidity and mortality considerably during the past decades.\(^8-11\)
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- Covered Stent to Eliminate Fontan Baffle Leak & Stenosis
- Native Coarctation of Aorta - Stent Implantation
- Pulmonary Artery Flow Restrictors
- Transcatheter Valve
- Intra-operative PA Stent
- Perventricular Muscular VSD Device Closure
- Closure of Septal Defect Using Real Time 3D Echo Guidance
- Perventricular Membranous VSD
- Hybrid Stage I Palliation for HLHS PA Bands and PSA Stent
- Intraoperative Aortic Stent for CoA
- Intraoperative LPA Stent Using Endoscopic Guidance
- Creation of ASD after PA Bands & PDA Stent for HLHS in a Preemie
- Perventricular Implant of Edwards Valve Stent in the Pulmonary Position
- Closure of Septal Defect Using Real Time 3D Echo Guidance
- High Frequency Ultrasound Creation of ASD
- PmVSD Closure
- Hybrid Stage I Palliation for Complex Single Ventricle in a 1.4 kg Neonate
- Transcatheter Implantation of Implantable Melody Valve
- Perimembranous VSD Closure with Amplatzer Membranous VSD Occluder
- Stent RPA, Pre-Stent Conduit & Melody Valve Implant
- Transeptal LHC Assessment of Residual Coarctation
- Coarctation Stenting Using a Covered CP Stent
- Closure of ASD Under ICE Guidance
- PFO Closure
- Transcatheter Closure of Atrial Septal Defect (ASD)
- Closure of ASD under TEE Guidance
- Transcatheter Closure of Moderate PDA
- Pulmonary Angioplasty
- Hybrid Stage Palliation
- ADOI or MVSD Closure of PDA
- VSD Closure Using a PDA Device
- Balloon Pulmonary Valvuloplasty
- Coil or Vascular Plug II closure of Coronary Fistula
- Device Closure of ASD
- Stent Placement Across Recurrent Coarctation
- Stenting RVOT on 7-day Old Newborn
- Transfemoral AVR for AS
- ASD Closure with Septal Occluder
- Stenting of Coarctation of Aorta
- Transapical AVR for Aorta Stenosis
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First stage is in the neonatal period and aims to regulate pulmonary blood flow, either by banding of the main pulmonary artery (MPA) in children with excessive pulmonary blood flow, or by discontinuation of the MPA and insertion of an aortopulmonary shunt, i.e. modified Blalock-Taussig shunt. For children with systemic outflow obstruction or HLHS the Damus-Kaye-Stansel anastomosis and Norwood-type procedure will augment the systemic blood flow while pulmonary blood flow is controlled by a Blalock-Taussig shunt or a right ventricular-pulmonary artery conduit. Furthermore, malformations such as obstruction of the aortic arch and restrictive foramen ovale have to be addressed during first stage.

Second stage is performed in order to volume unload the functional ventricle. It includes an anastomosis between the superior vena cava and the pulmonary artery (bidirectional Glenn anastomosis (BDG)), and simultaneous discontinuation of the MPA and removal of the Blalock-Taussig shunt. The importance of ventricular wall size thickness has been realized to be crucial to the success of the TCPC operation. When ventricular volume overload is suddenly removed after the TCPC operation, geometric alterations occur in the face of relatively unchanging ventricular mass but considerable decrease in cavity size. These sudden changes may impair diastolic ventricular function and result in potentially lethal low cardiac output stage postoperatively. The BDG is therefore performed to allow early reduction of the volume overload of the functional ventricle and a gradual remodeling of ventricular geometry before completion of the TCPC operation. Staging the TCPC operation has resulted in a decrease in morbidity and mortality. Timing of the BDG procedure has been discussed in several studies, with many highlighting the benefit of performing an “early” BDG procedure between two and six months of age.

In the third and last stage the TCPC circulation is completed, which separates the systemic and pulmonary venous return and provides pulmonary blood flow without a ventricular pumping chamber. The original Fontan operation consisted of an anastomosis between the right atrium and the central pulmonary arteries (RA-PA connection). Follow-up, however, revealed a high incidence of postoperative atrial arrhythmia and thromboembolic events due to atrial enlargement. The technique has evolved through a lateral tunnel procedure and is today in most centres performed as an extracardiac tunnel procedure by insertion of a Gore-tex tunnel between the inferior vena cava and the pulmonary artery. Thereby atrial manipulation is minimized and atrial incision and suture lines eliminated. However, advocates of the lateral tunnel approach are concerned about the possible increased likelihood of thrombosis or stenosis in an external conduit and the lack of growth potential.

Insufficient long-term data are available to differentiate the outcome between these two approaches, however, early and intermediate results seem to be in favor of the extracardiac conduit. Unlike BDG, no consensus exists about the optimal timing of the TCPC operation. Some centers advocate early intervention to minimize the effect of persistent cyanosis on patient growth, those of volume overload on the ventricle, and the risk of paradoxical embolism. However, very young infants have shown higher values of pulmonary vascular resistance, lower oxygen saturation and a higher prevalence of arrhythmias after TCPC than older ones. Furthermore, several groups demonstrated that TCPC cannot be considered a definitive solution because of progressive, long-term attrition in terms of mortality and morbidity. A recent study showed, that the TCPC operation can be performed safely in older patients (>7 years) without affecting operative and medium-term follow-up results. However, most centers perform the TCPC operation at 2 to 3 years of age.

Fenestration

A surgical fenestration between the total cavopulmonary connection and the atrium improves survival in high-risk patients. The fenestration functions by allowing right-to-left shunt resulting in an increased cardiac index, albeit with lower than normal oxygen saturation. Besides improved survival in high-risk patients, a fenestration also reduces the duration of hospital stay, the duration and volume of chest tube drainage. It has been suggested that a fenestration might be a benefit for late morbidity. Right-sided thrombus formation and paradoxical embolization are, theoretically, more likely to happen after a fenestrated repair. However, several reports have been unable to confirm this observation. In most centers closure of the fenestration are performed by implanting a device during cardiac catheter procedure. The necessity, proper timing, and clinical benefit of fenestration closure remain unknown. After fenestration closure, improved oxygenation, reduced need for anticoagulation, and improved somatic growth, but higher incidence of tachyarrhythmias have been reported.

Mortality

Perioperative and early mortality after the TCPC operations have decreased markedly over the past three decades. In the report by Khairy et al., 82.6% of the early survivors were alive and had not had a heart transplantation 15 to 20 years later. It is of interest that the study found no significant difference in life expectancy beyond the postoperative period between the more obsolete, hemodynamic less efficient RA-PA connection and the direct cavo-pulmonary artery connection. Furthermore, data from this series indicated that the three most common causes of late death were thromboembolism, heart failure and sudden death.

Survival at ten years ranges from 75 to 94% and at 20 years from 68 to 87% (Table 1). Though, most of the retrospective studies in Table 1 include relatively heterogeneous patient groups, with heterogeneous treatment strategies, one can appreciate that early and late mortality was higher for patients operated on in the earlier era than for patients operated on more recently.

Dominant Right Ventricle Compared to Left Ventricle

Several groups have reported that patients with a dominant right ventricle (RV) may be at increased risk for post-TCPC complications. A recent study, however, found that early and intermediate results were similar between patients with a dominant RV and a dominant left ventricle (LV).

Experience from congenital corrected transposition and atrial switch for D-transposition (Senning/Mustard) has shown that the systemic RV has a substantial long-term failure rate. Several factors may explain the generally equivalent event-free survival in TCPC patients of the dominant LV and RV. First, mortality due to rhythm disturbances, protein-losing enteropathy (PLE), stroke, and thromboembolic...
episodes may overwhelm the impact of ventricular morphology. Second, the staged approach to single ventricle palliation may outdo the impact of ventricular morphology. Specifically, HLHS is the most common single-ventricle diagnosis, and these patients are subject to a treatment algorithm that benefits from the increased experience with the diagnosis. Patients with HLHS undergo predictable neonatal palliation, then undergo relatively early Glenn and TCPC operation that both relieves volume overload and improves the oxygenation. These patients may be better prepared for the TCPC procedure than many patients with dominant LV, who may have variable anatomy, may present outside of the neonatal period, and consequently may be subjected to longer periods of volume overload or cyanosis. However, follow-up time in studies identifying similar behavior of the single RV or LV is relatively short. With time and accumulation of additional patients, a difference in mortality and morbidity may become apparent.

### Arrhythmias

The incidence of both atrial tachy- and bradyarrhythmias increases with time after TCPC completion. These arrhythmias include sinus node dysfunction (SND) as well as supraventricular tachycardias such as junctional ectopic tachycardia, atrial ectopic tachycardia, intra-atrial reentry tachycardia. The incidence of arrhythmias ranges from 5% to 77% depending on time of follow-up and type of Fontan/TCPC procedure (Table 1). Thus, the incidence of supraventricular tachycardia (SVT) following TCPC surgery increases steadily with the postoperative interval, with about 50% of patients experiencing problematic SVT by the 20th year of follow-up. Arrhythmias may result from increased atrial pressure, or be incisional. Much of the research on arrhythmias after TCPC operation has focused on identifying predictors for atrial arrhythmias, and many have focused on surgical strategy as a possible critical predictor. Thus, SND and SVT following TCPC is believed caused by injury of the sinus node or disturbances of its blood supply. Functional rhythm and loss of atrioventricular synchrony result in reduction of cardiac output, and SND may predispose patients to atrial reentry tachycardia. One of the theoretical advantages of the extracardiac TCPC operation is the minimization of incisional lines and the avoidance of atrial wall stress that may predispose the patient to SND and SVT. In a multivariate analysis Azakie et al. showed a lateral tunnel as

### Table 1: Results of the Fontan Procedure

<table>
<thead>
<tr>
<th>Author</th>
<th>Design</th>
<th>Number of patients</th>
<th>Year of operation</th>
<th>Fontan procedure</th>
<th>Survival</th>
<th>SVT</th>
<th>SND</th>
<th>Thromboembolism</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kim SJ</td>
<td>Retro</td>
<td>200</td>
<td>1996-2006</td>
<td>Ec: 100%</td>
<td>10 year: 92%</td>
<td>5,0%</td>
<td>11%</td>
<td>6,5%</td>
</tr>
<tr>
<td>Blaufox AD</td>
<td>Retro</td>
<td>521</td>
<td>1990-2007</td>
<td>RA-PA: 14%</td>
<td>9,6%</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>d’Udekem Y</td>
<td>Retro</td>
<td>305</td>
<td>1980-2000</td>
<td>RA-PA: 50%</td>
<td>10 year: 91%</td>
<td>20 year: 84%</td>
<td>10 year: 15,5%</td>
<td>10 years: 3,1%</td>
</tr>
<tr>
<td>Pace NC</td>
<td>Retro</td>
<td>65</td>
<td>1990-2008</td>
<td>Lat: 20%</td>
<td>10 year: 94%</td>
<td>Follow-up: 5,7 year ±5,4: 13,8%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Tweddel JS</td>
<td>Retro</td>
<td>256</td>
<td>1994-2007</td>
<td>Lat: 32%</td>
<td>10 year: 94%</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ocello S</td>
<td>Retro</td>
<td>100</td>
<td>1997-2003</td>
<td>Ec: 100%</td>
<td>Follow-up 3,3 year ±1,7: 94%</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Kirklin JK</td>
<td>Retro</td>
<td>112</td>
<td>1988-2005</td>
<td>Lat: 55%</td>
<td>10 year: 83%</td>
<td>15 year: 71%</td>
<td>Lat: 15 year: 46%</td>
<td>15 years: 5%</td>
</tr>
<tr>
<td>Khairy P</td>
<td>Retro</td>
<td>261</td>
<td>1973-?</td>
<td>RA-PA: 61%</td>
<td>10 year: 75%</td>
<td>20 year: 68%</td>
<td>25 year: 54%</td>
<td></td>
</tr>
<tr>
<td>Ono M</td>
<td>Retro</td>
<td>121</td>
<td>1984-2004</td>
<td>RA-PA: 23%</td>
<td>20 year: 87%</td>
<td>20 years: 77% (including VT)</td>
<td>22%</td>
<td>9%</td>
</tr>
<tr>
<td>Stamm C</td>
<td>Retro</td>
<td>220</td>
<td>1987-1991</td>
<td>Lat: 100%</td>
<td>10 years: 9%</td>
<td>5 years: 4%</td>
<td>5 years: 12%</td>
<td>10 years: 21%</td>
</tr>
<tr>
<td>Haas GS</td>
<td>Retro</td>
<td>45</td>
<td>1990-1997</td>
<td>Ec: 100%</td>
<td>Follow-up: 5,3 years: 100%</td>
<td>13%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Coon PD</td>
<td>Retro</td>
<td>592</td>
<td>1987-1999</td>
<td>RA-PA: 12%</td>
<td>1 year: 8%</td>
<td>8 years: 16%</td>
<td>10 years: 18%</td>
<td></td>
</tr>
</tbody>
</table>

TCPC patients developing atrial tachycardia usually experience electrical problems as well as significant hemodynamic abnormalities. Thus, attention to only the arrhythmia with medication or ablation, or attention to only the hemodynamic problem with TCPC revision often shows recurrent tachycardia.

Deal et al. showed good results in terms of low operative mortality, low risk of recurrent tachycardia, and marked improvement in functional status after TCPC revision with arrhythmia surgery (isthmus ablation, modified right atrial maze and Cox-maze III) in TCPC patients with frequent recurrence of atrial tachycardia and significant hemodynamic problem.

Thromboembolic complications

Thromboembolic events (TE) (systemic venous and arterial) are a major cause of early and late mortality in TCPC patients. The reported incidence of TE in these patients varies from 3% to 25% depending on study design, the imaging technique, and the duration of the follow-up period. Although the etiology for this high risk is not well defined, possible explanations include venous stasis, coagulation abnormalities, dehydration, protein-losing enteropathy, and arrhythmias. Interestingly, there is no clustering of thromboembolic complications around a specific period after Fontan. They start to occur during the postoperative period, and the risk appears to be persistent throughout a patient’s lifetime, with no plateau.

Coon et al. reviewed transthoracic echocardiograms in 592 TCPC patients. They found no difference in thrombus formation between atrio pulmonary and lateral tunnel TCPC procedure, or between fenestrated and non-fenestrated TCPC patients. A recent study found no difference in TE between lateral tunnel and extracardiac tunnel procedures; however, more results comparing these two procedures is needed.

Abnormalities in the haemostatic system have been found. Procoagulant factors (factors II, V, VII, IX, and X; plasminogen; fibrinogen), as well as anticoagulant factors (protein C and antithrombin III), were lower than normal controls prior to the BDG and prior to the TCPC completion. Increased platelet reactivity prior to the TCPC completion has also been shown. A recent report, however, found no significant differences in thromboelastography (a global whole-blood assay of coagulation) in pediatric TCPC patients compared with healthy children.

There is no consensus concerning the postoperative mode and duration of prophylactic anticoagulation, since no controlled randomized studies have been performed comparing different therapeutic strategies. Many centers recommend acetylsalicylic acid for uncomplicated TCPC patients, while Coumadin is reserved for patients with additional risk factors as arrhythmia, protein losing enteropathy, or ventricular dysfunction.

Cardiopulmonary Function

There have been numerous studies of aerobic capacity after the TCPC operation, all showing a substantial reduction of exercise capacity measured as peak oxygen uptake (VO₂) in a cardiopulmonary exercise test (Table 2).

Giardini et al. demonstrated that exercise capacity progressively declined in TCPC patients over time. They studied, longitudinally, 53 TCPC patients with cardiopulmonary exercise test (average 3.2±1.1 test for each patient). Time intervals between tests ranged from 1 to 16 years (average 7.7±4.0 years). Overall, peak VO₂ decreased at a rate of -2.6±2.7%/year. Left ventricular morphology appeared as a predictor of the rate of decrease of peak VO₂ at multivariate analysis (p=0.0001), thus the progressive decrease in exercise capacity is slower in patients with a morphologically LV. A similar decrease in peak VO₂ over time was found by Müller et al in adult TCPC patients, whereas exercise capacity stayed unchanged during childhood.

Minamisawa et al. showed that programmed exercise training could be safely performed to improve aerobic performance and oxygen utilization in TCPC patients. Although the improvement was a modest one, the improvement in oxygen use and participation in appropriate exercise may allow patients to lead a more active life.

Protein-losing Enteropathy

PLE is a devastating complication after TCPC operation that responds poorly to present treatment protocols. PLE is defined as an abnormal loss of serum-proteins into the lumen of the gastrointestinal tract. The cause of PLE is unknown, and pathophysiology is not well understood. Possible explanations might be decreased cardiac output, increased systemic venous pressure, inflammation and pulmonary vascular dysfunction. Its clinical symptoms include peripheral edema, ascites, pleural- or pericardial effusion, diarrhea, and malaise. In laboratory tests a low and falling S-albumin and S-total protein and a high α1-antitrypsin in the stool is found. Time from TCPC operation to development of PLE is, in the literature, reported from 1 month and forward. The prevalence of PLE is reported from 1-15% and increasing with age and 5-year survival rate 46-59%. No effective treatment is available. Standard treatment includes diet (high protein, low fat), inotropic and afterload reducing agents, anti-inflammatory agents (corticosteroids), unfractionated heparin, and diuretics and albumin infusions. However, these agents have only shown sporadic effect.

Case reports suggest pulmonary vasodilation as a potential medical treatment.

Despite treatment PLE is associated with a very high mortality rate. Further investigation is needed to determine the exact mechanism of the disease and to develop new therapeutic approaches.

Quality of Life and Daily Activity

Daily activity is an important lifestyle factor for both children and adults, however, studies measuring daily activity in TCPC patients are sparse. Müller et al. investigated exercise capacity, quality of life and daily activity in 57 TCPC patients. Despite a reduced exercise capacity (Table 2), the patients seemed to be fairly active, many of them showing normal daily activity pattern. However, children younger than nine years were most prominently curtailed in physical activity compared to their healthy peers, perhaps due to overprotection. Later in adolescence, daily activity was mainly...
related to exercise capacity that was found to decline from early adolescence on. Self-reported quality of life, concerning physical functioning did not correlate to objectively measured exercise capacity.65 Blaloux et al. investigated quality of life in 521 TCPC patients using the Child Health Questionnaire (CHQ) Parent Report65. Mean CHQ physical (CHQ-p) and CHQ psychosocial (CHQ-ps) summary scores were lower than those for healthy controls: CHQ-p was 45.4 ± 11.8 versus 53.0 ± 8.8 for historical controls and CHQ-ps was 47.3 ± 10.9 versus 51.2 ± 9.1 for historical controls.

In another study, reduced habitual physical activity was associated with lower perceived general health, but no other aspects of quality of life.66 The same authors, in a recent multicenter study, found only weak association between parent-estimated functional health status and exercise capacity and ventricular characteristics (echocardiography and magnetic resonance imaging) in 511 TCPC patients 6 to 18 years of age.67

Failure of the TCPC Circulation

Even after excellent early results, TCPC patients remain at risk of TCPC circulation failure. The most common manifestations of failing TCPC physiology are progressive ventricular dysfunction, PLE, arrhythmia, TE, progressive hypoxemia, and elevated pulmonary vascular resistance.7, 68 So far there are no diagnostic tools allowing early prevention of declining univentricular circulation. Although the prognosis for UVH has changed considerably during the last decades, there still remains a continuing decrement of functional status during long-term follow-up. In failing TCPC, both TPCC takedown by conversion into BDG or orthotopic heart transplantation are possibilities.

Conclusions

Staged palliation and major improvements in the surgical techniques, have resulted in a decreasing mortality rate and increased prevalence of UVH patients. Despite these improvements, significant morbidity remains after the TCPC completion, including myocardial dysfunction, arrhythmias, TE, PLE and diminished exercise capacity. Therefore, the TCPC operation remains a palliative procedure and an imperfect solution. The persistent long-term morbidity document the obvious need for regular long-term surveillance of these patients as well as prospective follow-up studies.

Future Perspective

As the number of late survivors from the TCPC operation increases, caregivers will be evermore faced with the challenge of recognizing and managing complications secondary to the TCPC circulation. Early diagnosis and management of functional sequelae and complications associated with the TCPC physiology is required for improving the long-term outcome and quality of life.

Persistent investigations into new surgical modifications of the TCPC circulation as well as investigations into possible risk factors and treatment to TCPC failure are needed in order to continue improved outcome in the UVH patients.

Table 2. Aerobic Capacity After TCPC Operation

<table>
<thead>
<tr>
<th>Author</th>
<th>n</th>
<th>( \text{Vo}_{2\text{max}} ) mL/Kg/minute</th>
<th>% normal</th>
</tr>
</thead>
<tbody>
<tr>
<td>Müller J66</td>
<td>50</td>
<td>25.0</td>
<td>59.7</td>
</tr>
<tr>
<td>Harrison DA71</td>
<td>30</td>
<td>14.8</td>
<td></td>
</tr>
<tr>
<td>Ohuchi H72</td>
<td>105</td>
<td></td>
<td>59</td>
</tr>
<tr>
<td>RV morphology</td>
<td>33</td>
<td>25</td>
<td>55</td>
</tr>
<tr>
<td>LV morphology</td>
<td>47</td>
<td>27</td>
<td>63</td>
</tr>
<tr>
<td>Biventricule</td>
<td>15</td>
<td>25</td>
<td>55</td>
</tr>
<tr>
<td>Paridon SM73</td>
<td>411</td>
<td>26.3</td>
<td>65</td>
</tr>
<tr>
<td>Ovrouski S74</td>
<td>43</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Children (≤15 years)</td>
<td>28</td>
<td>27.9</td>
<td>62</td>
</tr>
<tr>
<td>Adults</td>
<td>15</td>
<td>22.9</td>
<td>53.5</td>
</tr>
</tbody>
</table>

LV: left ventricle; RV: right ventricle. \( \text{Vo}_{2\text{max}} \): maximal oxygen uptake.

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Abstracts from “Evolving Concepts in the Management of Complex Congenital Heart Disease II” - Part II

Objective
- Case presentations of representative adult CHD first diagnosed as an adult
- Case presentations of adults with CHD with postoperative issues
- Review the modern outlook and management of Eisenmenger Syndrome

Abstract

Surgical advances in CHD over the past 60+ years allowed >85% survival into adulthood. Adults with CHD now exceed 1 million in the U.S. of which >1/3rd are moderate-to-complex. Operation does not imply cure. Residua and sequelae (separate from complications), whether mild or severe are common. Residua (defects not addressed at the time of surgery) include long-term systemic RV or single ventricle function. Sequelae (abnormalities resulting from intervention) include severe pulmonary regurgitation or arrhythmias in tetralogy of Fallot, longevity of conduits and valves, or atrial arrhythmias in atrial baffle or Fontan patients. Myocardial dysfunction due to inadequate myocardial protection, extensive patches or incisions, or long-standing pressure or volume overload is a common concern.

Not all have been or will be operated. Some may not be recognized until the natural history results in symptomatic presentation in the adult years (e.g. CCTGA, Ebstein). In those that were inoperable, Eisenmenger Syndrome (ES) was more common in the earlier eras. Survival of ES is determined by the type of underlying CHD and the medical complications of cyanosis. Cyanosis has profound hematologic consequences and is a multi-system disorder.

References
There are two main treatment algorithms for the symptomatic neonate – the biventricular repair and the single ventricle pathway. In the current era, both surgical strategies continue to have high early mortality (10-30%) even in the most experienced hands, when compared to the majority of other complex congenital heart anomalies operated on in the first year of life, where the early mortality has been uniformly reduced to <5%.

There is general agreement that management of the child, adolescent and young adult should be focused on tricuspid valve repair (as opposed to replacement); almost always this operation is elective and early mortality is and should be low. While there is no general agreement as to the ideal method of repair, it is my impression that the “cone reconstruction” provides the most anatomic repair and should be applied in the majority of circumstances.

Management of the older adult (>50 years) with Ebstein’s Anomaly has its own set of technical challenges – often markedly enlarged hearts with concomitant atrial tachyarrhythmias, possible atherosclerotic coronary artery disease, or other acquired valve abnormalities. In addition, the presence of medical comorbidities such as diabetes or hypertension is also common, making perioperative management more difficult. The durability of the porcine bioprosthesis in this age bracket makes tricuspid valve replacement a reasonable alternative when valve repair is not possible, since durability and freedom from reoperation has been shown to be very good. The need for concomitant procedures such as the maze procedure, coronary artery bypass grafting, etc. also needs to be considered.

References

complex operations is acceptable at 2%. Development. Using current techniques surgical mortality for these hypoplastic pulmonary arteries and MAPCAs keeps up with somatic vascular resistance, allowing a high rate of intracardiac repair with low possible and is achieved early in infancy promotes low pulmonary artery system that includes as many lung segments as both surgical and interventional techniques is an important component of Ongoing evaluation of the pulmonary vasculature with appropriate use of rare. Pressure did not rise over time, and growth of unifocalized MAPCAs is Meier survival at 1, 5, and 10 yrs was 91%, 86%, and 86%. RV/LV pressure was 0.44 in the first half, and 0.35 in the second half. Late patients, according to strict morphologic and physiologic criteria. Staged unifocalization via thoracotomy, were performed in a minority of initial operations, such as creation of main PA to aortic anastomosis, or unifocalization using native tissue anastomoses. Intracardiac repair is performed as early as possible, but only when low right ventricular pressure is assured.

Our total experience involves 507 patients. There were 3.8 +/- 1.4 MAPCAs per patient. The median diameter of the central pulmonary arteries (PAs) was 1.5 mm. PAs were absent in 23%. Cardiac morphology was pulmonary atresia with VSD in 94%. Median age at first surgery was 8 months. Initial operation was complete bilateral unifocalization using native tissue anastomoses. Intracardiac repair was also achieved at initial operation in 56%, and in 83% within 2 yrs. Alternative initial operations, such as creation of main PA to aortic anastomosis, or staged unifocalization via thoracotomy, were performed in a minority of patients, according to strict morphologic and physiologic criteria. Early mortality was 9.7% in the first half of the experience, and 2.0% in the second. In patients achieving intracardiac repair, the median RV/LV pressure was 0.44 in the first half, and 0.35 in the second half. Late follow-up was 81% complete up to 12 yrs with 6.3% mortality. Kaplan-Meier survival at 1, 5, and 10 yrs was 91%, 86%, and 86%. RV/LV pressure did not rise over time, and growth of unifocalized MAPCAs is well documented. Documented occlusion of unifocalized MAPCAs was rare.

Ongoing evaluation of the pulmonary vasculature with appropriate use of both surgical and interventional techniques is an important component of the overall management. Surgical creation of a single compartment pulmonary artery system that includes as many lung segments as possible and is achieved early in infancy promotes low pulmonary vascular resistance, allowing a high rate of intracardiac repair with low right ventricular pressure. Under this management, growth of both hypoplastic pulmonary arteries and MAPCAs keeps up with somatic development. Using current techniques surgical mortality for these complex operations is acceptable at 2%.
Request for Research Applications on Pediatric Cardiomyopathy

The Children’s Cardiomyopathy Foundation (CCF) is pleased to announce the availability of one-year research grants for studies focused on pediatric cardiomyopathy. The purpose of CCF’s Annual Research Grant Program is to advance knowledge of the basic mechanism of the disease and to develop more accurate diagnostic methods and improved therapies for children affected with cardiomyopathy. Please visit CCF’s website www.childrenscardiomyopathy.org (click on Research/Grants & Awards) for guidelines and to view past grant awards.

Opportunity: The Children’s Cardiomyopathy Foundation (CCF) is inviting investigator-initiated research proposals for innovative basic, clinical, population, or translational studies related to the cause, diagnosis, or treatment of primary cardiomyopathy in children under the age of 18 years. CCF’s grant program is designed to provide seed funding to investigators for the testing of initial hypotheses and collecting of preliminary data to help secure long-term funding by the NIH and/or other major granting institutions.

Eligibility: Principal investigator must hold an MD, PhD or equivalent degree and reside in the United States or Canada. The investigator must have a faculty appointment at an accredited US or Canadian institution and have the proven ability to pursue independent research as evidenced in publications in peer-review journals.

Funding: Funding is available in the range of US$25,000 to US$50,000 for one year of total direct costs. Following the completion of the proposed study, a second year of funding may be an option for relevant study extensions.

Application Process: CCF grant guidelines and application forms are downloadable off the website. The 2010 deadline for application submission is September 3, 2010 with final award decisions to be made by January 2011.

Selection Process: Grant award decisions are made through a careful peer-review process led by CCF’s Medical Advisors and reviewed by CCF’s Board of Directors. Scientific excellence and relevance to primary forms of pediatric cardiomyopathy are the main criteria for selecting research projects to support.

Bypass Procedure Used During Infant Heart Surgery Does Not Impair Later Neurological Outcomes

Congenital heart defects (CHD) are the most common birth defects in humans, affecting 8 per 1000 live births with one third of affected children requiring intervention in early infancy. Increasing numbers of survivors combined with developmental expectations have led to a growing identification of neurobehavioral symptoms in some survivors. A study now suggests that a cooling technique often used in heart operations does not impair neurological outcomes.

Congenital heart disease and its treatment were originally thought to potentially increase neurologic injury in these patients. The technique of deep hypothermic circulatory arrest (DHCA) is used in order to repair these congenital cardiac defects by providing a bloodless surgical field, which may facilitate completion of the best physiologic repair, and decrease the duration of blood exposure to the bypass circuit. However, it involves a period of reduced blood flow in the brain. Cooling is a protective mechanism to reduce metabolism of the brain and other organs during periods of low blood flow.

Stephanie Fuller, MD, a cardiothoracic surgeon at The Children’s Hospital of Philadelphia, presented these research findings in the annual meeting of the Society of Thoracic Surgeons in Fort Lauderdale, Fla. According to the study, DHCA does not impair language skills, attention, and other neurocognitive abilities in school-age children.

Dr. Fuller added “use of DHCA as a support technique during cardiac surgery in infancy has many advantages; it is not necessary to sacrifice these advantages merely to avoid use of DHCA. Our study adds to the growing literature showing no adverse influence of limited periods of DHCA. New support techniques must be carefully evaluated prior to wide-spread acceptance to confirm they are not inferior to conventional management strategies.”

For more information, visit www.chop.edu.

Silver Nanoparticles May One Day be Key to Devices That Keep Hearts Beating Strong and Steady

Diamonds and gold may make some hearts flutter on Valentine’s Day, but in a University...
at Buffalo laboratory, silver nanoparticles are being designed to do just the opposite.

The nanoparticles are part of a new family of materials being created in the laboratory of SUNY Distinguished Professor and Greatbatch Professor of Advanced Power Sources Esther Takeuchi, PhD, who developed the lithium/silver vanadium oxide battery. The battery was a major factor in bringing implantable cardiac defibrillators (ICDs) into production in the late 1980s. ICDs shock the heart into a normal rhythm when it goes into fibrillation.

Twenty years later, with more than 300,000 of these units being implanted every year, the majority of them are powered by the battery system developed and improved by Takeuchi and her team. For that work she has earned more than 140 patents, believed to be more than any other woman in the United States. Last fall, she was one of four recipients honored in a White House ceremony with the National Medal of Technology and Innovation.

ICD batteries, in general, now last five to seven years. But she and her husband and co-investigator, SUNY Distinguished Teaching Professor of Chemistry Kenneth Takeuchi, PhD, and Amy Marschilok, PhD, UB Research Assistant Professor of Chemistry, are exploring even-better battery systems, by fine-tuning bimetallic materials at the atomic level.

Their research investigating feasibility for ICD use is funded by the National Institutes of Health, while their investigation of new, bimetallic systems is funded by the US Department of Energy (DOE).

So far, their results show that they can make their materials 15,000 times more conductive upon initial battery use due to in-situ (that is, in the original material) generation of metallic silver nanoparticles. Their new approach to material design will allow development of higher-power, longer-life batteries than was previously possible.

These and other improvements are boosting interest in battery materials and the revolutionary devices that they may make possible.

“We may be heading toward a time when we can make batteries so tiny that they -- and the devices they power -- can simply be injected into the body,” Takeuchi says. Right now, her team is exploring how to boost the stability of the new materials they are designing for ICDs. The materials will be tested over weeks and months in laboratory ovens that mimic body temperature of 37 degrees Celsius.

“What’s really exciting about this concept is that we are tuning the material at the atomic level,” says Takeuchi. “So the change in its conductivity and performance is inherent to the material. We didn’t add supplements to achieve that, we did it by changing the active material directly.”

Even though batteries are an historic technology, they are far from mature, Takeuchi notes.
Fixing a heart from birth through adulthood takes big teams working together. So we examined the needs of leading clinicians when designing our hybrid solutions. The result: our Infinix™-i with 5-axis positioners and low profile detectors, stays out of the way, but right where needed, providing the best possible access to patients. To lead, you must first listen.

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