Is alternative cardiac surgery an option in adults with congenital heart disease referred for thoracic organ transplantation?†

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Abstract

OBJECTIVES: We analysed the outcomes of adults with congenital heart disease (ACHD) referred for thoracic organ transplantation who underwent non-transplant cardiac surgery as an alternative management option.

METHODS: Adult patients with congenital heart disease assessed for heart or heart–lung transplant were identified from the departmental database. A retrospective analysis of the medical records, transplant assessment data and surgical notes was carried out.

RESULTS: One hundred and twenty-six patients were assessed between January 2000 and July 2011. Non-transplant cardiac surgery was performed in 14 (11%) patients. There were nine males with a median age of 37 years (range 21–42). The patients can be divided into four subgroups [left-sided lesions (n = 4), right-sided lesions (n = 3), systemic right ventricle (n = 5) and Fontan circulation (n = 2)]. Surgical procedures performed were: relief of systemic obstructive/regurgitant lesions ± endocardial fibroelastosis resection (n = 4, three pulmonary vascular resistance >6 Wood units), correction of right-sided regurgitant/stenotic lesions (n = 3), ventricular assist device for patients with a systemic right ventricle (n = 5) and re-fashioning of the Fontan pathway (n = 2). There were two early (5 and 30 days) and three late deaths (64, 232 and 374 days) with a 1-year mortality of 28%. None of the deaths occurred in patients with a two-ventricle circulation and atrio-ventricular concordance. Nine patients are alive at a median of 433 days (range 204–2456). The New York Heart Association class has improved in all survivors by at least one class at 3 and 6 months (P = 0.004 and 0.003).

CONCLUSIONS: Alternative cardiac surgery can be undertaken in selected patients with ACHD referred for cardiopulmonary transplantation with a low mortality in patients with two ventricles and a systemic left ventricle. Ventricular assist devices carry a significant mortality in patients with a systemic right ventricle, although this offers a valuable palliation when there are no other options. The medium and long-term results are awaited.

Keywords: Transplantation • Adult congenital heart disease • Cardiac surgery • Ventricular assist device

INTRODUCTION

The number of adults with congenital heart disease (ACHD) continues to increase with advances in diagnostic, medical and surgical management [1, 2]. The majority of patients with a mild and moderate disease remain well; however, a significant proportion with a moderate and severe disease continues to deteriorate. Despite optimal medical management, patients in this group often require catheter-based intervention and conventional cardiac surgery, with some requiring consideration for either cardiopulmonary transplantation or end-of-life palliation.

The management of patients with a congenital heart disease referred for transplantation is dictated by a complex interplay of factors. These include: the putative life expectancy of the patient, the complexity of cardiovascular malformation, the presence and extent of pulmonary vascular disease, the pulmonary artery anatomy and the likelihood of preformed human leucocyte antigen (HLA) antibodies [3]. In addition, there are national and institutional considerations that can play a major part in determining the allocation of donor hearts and lungs, thus affecting the likely outcome of these patients. Alternative cardiac surgery can be considered in some of the patients where cardiopulmonary transplantation is not feasible, is too high-risk or a surgical procedure is required to enable transplantation to become a viable option.

In this retrospective observational study, we describe a single-centre’s experience of alternative cardiac surgery as a management option in the adult population (≥18 years) with ACHD referred for transplantation.
METHODS

All patients referred for ACHD transplant are admitted and formally assessed over a minimum of 3 days. Patients are then discussed at a joint congenital heart disease/cardiac transplant meeting and a decision made with regards to further management and whether the patient should be offered listing for transplant. Details of patients undergoing assessment are recorded in the departmental database.

Patients (aged ≥18 years) with congenital heart disease assessed for heart or heart–lung transplant were identified from the database and a retrospective analysis of the medical records, transplant assessment data and surgical notes was carried out on all patients referred for transplant assessment from 1 January 2000 to 31 July 2011. The outcome is reported as of 31 January 2012. Diagnosis, comorbidities, demographic data, surgical and medical procedures and other salient pre-assessment data were recorded.

Statistical analysis

The patients’ preoperative New York Heart Association (NYHA) status was compared with their status at both 3 and 6 months following surgery, using the Wilcoxon signed-rank test using SPSS version 19 software.

The Kaplan–Meier curves were generated and the $\chi^2$ test used to compare the subgroups to demonstrate the survival status using MedCalc version 10.0.2.

RESULTS

Patient population

One hundred and twenty-six patients were assessed for heart or heart–lung transplant over the study period (103 referred for heart transplant and 23 for heart–lung transplant). Non-transplant cardiac surgery was planned in 15 (12%) patients. One of these was a 22-year old male with Shone’s complex who had undergone four previous aortic valve replacements and a mitral valve replacement. He was referred as NYHA Class 3 with an ejection fraction of <30%. He died before the planned left ventricular overhaul could be undertaken [redo aortic valve replacement, mitral valve replacement and resection of endocardial fibroelastosis (EFE)]. Therefore, 14 patients underwent surgery (NYHA 3.29 ± 0.61). Nine patients were male with the median age of 37 years (range 21–42). Demographic data, diagnoses, preoperative status, reasons for non-transplant surgery, type of surgery and outcome are summarized in Table 1. Four patients had predominantly left-sided lesions, three patients had predominantly right-sided lesions, five patients had a systemic right ventricle, (three post-atrial inversion procedures, two with congenitally corrected transposition) and two patients had a Fontan-type circulation (Table 1). Details of these subgroups of patients and reasons within the groups for non-transplant surgery and the type of surgery are reported below.

Of these 14 patients, six could have undergone cardiac transplantation if a suitable matched organ had been available immediately (suitable pulmonary artery anatomy, pulmonary vascular resistance (PVR) <6 Wood units, no comorbidity precluding transplant). Among these six potential transplant candidates, four were considered too unwell to wait for an organ without intervention (i.e. non-transplant surgery carried out as a bridge to transplant or recovery). In one patient, conventional surgery was considered a better option than non-transplant surgery and in one, the panel reactive antibodies (PRAs) were so high that waiting for transplant was not a realistic option (PRA >90%). None of the remaining eight patients were suitable candidates for heart transplant at the time of the initial assessment even if a matched donor had been immediately available. The reasons for this were PVR >6 Wood units in six patients (one with PRA >90%), unsuitable pulmonary artery anatomy in one patient (with PRA >90%) and early remission from a malignancy in one patient (with PRA >90%).

Among these 14 patients, there were two early deaths (5 and 30 days) and three late deaths (64, 232 and 374 days) with an overall 1-year mortality of 28% (Fig. 1). Nine patients are alive at a median of 433 days (range 204–2456 days) from the surgery. Nine have undergone subsequent transplantation, with one patient actively listed for heart transplant. Except for the pre-operative death, there has been no mortality for those with a two-ventricle circulation and ventricular arterial concordance. All deaths have occurred in patients with a systemic right ventricle or a single-ventricle circulation (Fig. 2). All patients alive at 3 and 6 months following surgery demonstrated an NYHA improvement at 3 and 6 months ($P = 0.004$ and 0.003) (Table 1).

Left-sided lesions

Four patients had an original diagnosis of aortic valve disease (three aortic stenoses and one aortic regurgitation associated with Marfan’s syndrome). All had undergone at least two previous sternotomies for aortic valve replacement/repair and all had permanent or recurrent atrial fibrillation/flutter. All met the criteria for cardiac transplant based on symptoms and predicted survival. Three had a high PVR (>6 Wood units) at the time of the assessment due to chronically elevated left ventricular end-diastolic pressure/associated mitral regurgitation and one had associated EFE. Surgery, including atrial maze, was undertaken as outlined in Table 1. These three patients remain well, with an improvement in NYHA in all cases. A fourth patient had severely impaired left ventricular function following aortic root replacement 10 months previously and could not be listed for transplant in view of recent treatment for cervical carcinoma and high PRA. She underwent repair of the aortic homograft, tricuspid valve repair, atrial septal defect closure and left ventricular HeartWare® insertion as a bridge to recovery of the native cardiac function or as a bridge to reconsideration of the cardiac transplant pending sufficient time in remission from the malignancy.

Right-sided lesions

Three patients were referred with right-sided lesions, all with severe right ventricular dysfunction and all with impaired left ventricular function. Two patients had tetralogy of Fallot and had undergone corrective surgery and subsequent pulmonary valve replacements. Both had dilated right ventricles with severely impaired function from a combination of chronic tricuspid and pulmonary regurgitation. In one patient, tricuspid valve
Table 1: Patients undergoing conventional surgery categorized by primary anatomical lesion

<table>
<thead>
<tr>
<th>Patient</th>
<th>Anatomical classification</th>
<th>Initial diagnosis</th>
<th>Previous cardiac operations</th>
<th>Preoperative status</th>
<th>Why non-transplant surgery</th>
<th>Surgery</th>
<th>Status (days post-surgery) NYHA 3 and 6 months</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Left-sided lesions</td>
<td>AS EFE</td>
<td>(1) Aortic valvotomy (2) AVR metallic</td>
<td>NYHA 3 At home</td>
<td>PVR too high</td>
<td>AVR homograft, atrial maze, MVR, EFE resection</td>
<td>Alive (410) NYHA 2 and 2</td>
</tr>
<tr>
<td>2</td>
<td>AS</td>
<td></td>
<td>(1) AVR homograft (2) AVR homograft</td>
<td>NYHA 3 At home</td>
<td>PVR too high</td>
<td>AVR metallic, aortic root enlargement, atrial maze</td>
<td>Alive (2456) NYHA 1 and 1</td>
</tr>
<tr>
<td>3</td>
<td>AR (Marfan’s syndrome)</td>
<td></td>
<td>(1) AVR homograft (2) AVR metallic</td>
<td>NYHA 3 At home</td>
<td>PVR too high</td>
<td>Aortic root homograft, MVR, atrial maze</td>
<td>Alive (1199) NYHA 2 and 2</td>
</tr>
<tr>
<td>4</td>
<td>Supravalvular AS</td>
<td></td>
<td>(1) Aortic valvotomy, supravalvular AS repair (2) AVR metallic, supravalvular AS repair (3) AVR root, homograft</td>
<td>NYHA 3 Inpatient</td>
<td>PRA too high Early remission from malignancy</td>
<td>Aortic homograft repair, TVR, ASD closure, HeartWare® insertion</td>
<td>Alive (257) NYHA 2 and 2</td>
</tr>
<tr>
<td>5</td>
<td>Right-sided lesions</td>
<td>ToF Post-surgical LAD coronary artery occlusion</td>
<td>(1) Waterson shunt (2) ToF repair (3) Homograft PVR, LIMA to LAD, fistula closure (4) Biventricular pacing (5) Percutaneous PVR</td>
<td>NYHA 3 Inpatient*</td>
<td>PRA too high</td>
<td>Tricuspid valve replacement</td>
<td>Alive (598) NYHA 1 and 1</td>
</tr>
<tr>
<td>6</td>
<td>ToF</td>
<td></td>
<td>(1) ToF repair</td>
<td>NYHA 2 At home</td>
<td>Conventional surgery a better option</td>
<td>Tricuspid valve repair, PVR tissue, epicardial biventricular pacemaker TVR porcine valve</td>
<td>Alive (1041) NYHA 2 and 1</td>
</tr>
<tr>
<td>7</td>
<td>Ebstein’s anomaly</td>
<td></td>
<td>(1) TVR with pulmonary homograft, ASD and VSD closure (2) PA banding</td>
<td>NYHA 4 On ECMO</td>
<td>Too unwell to wait on ECMO</td>
<td></td>
<td>Alive (433) NYHA 2 and 2</td>
</tr>
<tr>
<td>8</td>
<td>Systemic right ventricle</td>
<td>ccTGA</td>
<td>(1) TVR metallic (2) ICD (3) ICD</td>
<td>NYHA 3 Inpatient on inotropes</td>
<td>PVR too high, mechanical support to reduce PVR</td>
<td>TVR metallic Mechanical support with Berlin heart</td>
<td>Dead (5)</td>
</tr>
<tr>
<td>9</td>
<td>TGA</td>
<td></td>
<td>(1) Right BT shunt (2) Mustard procedure</td>
<td>NYHA 3 At home</td>
<td>PVR and PRA too high</td>
<td>Insertion of HeartWare®</td>
<td>Dead (232) NYHA 2 and 2</td>
</tr>
<tr>
<td>10</td>
<td>TGA</td>
<td></td>
<td>(1) Mustard procedure</td>
<td>NYHA 4 Inpatient on inotropes</td>
<td>Too unwell to wait</td>
<td>Insertion of HeartWare®</td>
<td>Dead (64)</td>
</tr>
<tr>
<td>11</td>
<td>TGA Subpulmonary VSD</td>
<td></td>
<td>(1) Senning procedure</td>
<td>NYHA 4 Inpatient on inotropes</td>
<td>PVR too high</td>
<td>Insertion of HeartWare®</td>
<td>Alive (269) NYHA 2 and 2</td>
</tr>
<tr>
<td>12</td>
<td>TGA</td>
<td></td>
<td>(1) Senning procedure</td>
<td>NYHA 4 Inpatient on inotropes Post-partum</td>
<td>Too unwell to wait</td>
<td>Insertion of HeartWare®</td>
<td>Alive (204) NYHA 2 and 2</td>
</tr>
</tbody>
</table>

Continued
replacement, pulmonary valve replacement and biventricular pacing was thought to offer a better option than transplant. The second patient was undergoing regular admissions for intravenous diuretics and inotropes for right heart failure. She underwent previous percutaneous pulmonary valve implant and biventricular pacemaker insertion with ongoing severe tricuspid regurgitation. She was thought to be best served by transplant, however her PRA was 98% and therefore, tricuspid valve replacement was undertaken following a period of optimization (inotropes and diuretics). The other two patients who had undergone previous tricuspid valve replacement for Ebstein’s anomaly of the tricuspid valve had been assessed by their local team for cardiac transplant with increasing symptoms, deteriorating right and left ventricular functions and increasing tricuspid stenosis with pannus on the replaced valve. He presented acutely with thrombus occluding the tricuspid valve prosthesis and was transferred to us on veno-arterial extracorporeal membrane oxygenation (ECMO). He underwent redo tricuspid valve replacement with an Epic porcine 31-mm prosthesis from ECMO and continues to improve symptomatically at home.

Systemic right ventricle

Five patients had a systemic right ventricle, two with congenitally corrected transposition of the great arteries and three with transposition of the great arteries with previous atrial switch (two Mustard procedures and one Senning procedure). One patient underwent an insertion of a Berlin heart when he could not be taken off the bypass due to a low cardiac output state following a tricuspid valve (systemic atrio-ventricular valve) replacement. He remained vasoplegic, and despite a seemingly good function of the mechanical support, his blood pressure could not be maintained and he died 5 days later. The remaining four patients underwent HeartWare\textsuperscript{\textregistered} insertion with concomitant procedures (Table 1). Two of these patients’ PVR was too high for transplant at the time of referral and they underwent a mechanical support, both for survival and in an effort to reduce the left atrial pressure and PVR (bridge to reassessment for transplant or destination therapy). One patient was too unwell to wait for a suitable donor organ and HeartWare\textsuperscript{\textregistered} was inserted for survival to transplant. The fourth patient had a severe deterioration of the right ventricular function post-partum and HeartWare\textsuperscript{\textregistered} was inserted for survival to transplant or potential recovery of the right ventricular function. One patient, having been well, died at 2 months following HeartWare\textsuperscript{\textregistered} insertion from aspiration pneumonia following an ear, nose and throat procedure. Another died at 8 months with sepsis and impaired left (sub-pulmonary) ventricular function. The other two patients remain well in NYHA 1–2.

Single ventricle

Two patients with a Fontan pathway underwent Fontan revision to an extracardiac conduit. One patient was planned to be listed for transplant, as a Fontan revision was thought to be too high risk. However, he required anticoagulation reversal for a liver

<table>
<thead>
<tr>
<th>Patient</th>
<th>Anatomical classification</th>
<th>Initial diagnosis</th>
<th>Previous cardiac operations</th>
<th>Preoperative status</th>
<th>Why non-transplant surgery</th>
<th>Surgery</th>
<th>Status (days post-surgery) NYHA 3 and 6 months</th>
</tr>
</thead>
<tbody>
<tr>
<td>13</td>
<td>Single ventricle</td>
<td>Situs inversus with left atrial isomerism, double inlet right ventricle, double outlet right ventricle, TGA, pulmonary stenosis</td>
<td>(1) Classical BT shunt (2) Right-sided Glenn, right atrial to PA anastomosis (3) Systemic atrio-ventricular valve replacement metallic</td>
<td>NYHA 4 PRA 98%\textsuperscript{\textregistered}</td>
<td>PRA too high, PA anatomy not suitable</td>
<td>Fontan upgrade to extracardiac conduit with PA repair and maze procedure</td>
<td>Dead (374) NYHA 2 and 2</td>
</tr>
<tr>
<td>14</td>
<td>Tricuspid atresia with normally related great arteries, ASD, VSD</td>
<td>(1) Glenn shunt (2) Left BT shunt (3) Atrio-pulmonary Fontan (4) Takedown left BT shunt (5) Redo atrio-pulmonary Fontan</td>
<td></td>
<td>NYHA 3</td>
<td>Forced by Fontan thrombosis</td>
<td>Total cavo-pulmonary connection</td>
<td>Dead (30)</td>
</tr>
</tbody>
</table>


\textsuperscript{\textregistered}Recurrent admissions for diuresis, recent dialysis + inotropes.

\textsuperscript{\textregistered}Recurrent admissions for intravenous diuretics.
nodule biopsy (histology confirmed as benign). He subsequently deteriorated acutely with a massive thrombus formation in the atrio-pulmonary Fontan and, therefore, urgent Fontan revision was undertaken. He died with a low cardiac output and Fontan failure 1 month from the procedure. The second patient underwent Fontan revision in view of an unsuitable pulmonary artery anatomy and a PRA of 98%. There was an extensive thrombus in the atrio-pulmonary Fontan with a chronic complete occlusion of the stenosed left pulmonary artery. Conversion to an extracardiac conduit included thrombectomy and a long-segment gortex patch to the left pulmonary artery. She had significant improvement in her symptoms for several months and then was re-referred with increasing symptoms. Her pulmonary artery anatomy was sufficiently improved to consider transplant, but she died a year after the procedure on the transplant list following desensitization.

DISCUSSION

It is a measure of success of the management of children with congenital heart disease that increasing numbers of patients are reaching adulthood [1]. However, it is also becoming clear that not only the palliative but also some of the ‘curative’ procedures undertaken in childhood present with cardiopulmonary dysfunction later in life [4–6]. These can manifest as ventricular failure, pulmonary hypertension or debilitating cyanosis. Cardiopulmonary transplant continues to have a major role in the management of these patients with good results for those presenting with refractory symptoms and an estimated 1-year survival of <50% [7, 8]. The results of heart transplantation in patients with ACHD have the best long-term survival amongst all the cohorts undergoing cardiac transplantation [7, 8]. However, the data from the International Society for Heart and Lung Transplantation (ISHLT) registry also show that there is a higher early mortality associated with this group. This early risk reflects the challenges associated with complex reconstructions, re-operative surgery and the multiple comorbidities present in these patients including pulmonary vascular disease, chronic liver disease and coagulation disorders.

Given the paucity of donor hearts available, it is no surprise that the ISHLT data show that only 2% of adult hearts are considered for the diagnosis of ACHD [8]. This low proportion of transplants may reflect the competing factors, resulting in less risky transplants being preferentially undertaken.

The decision to perform either transplantation or alternative cardiac surgery in patients with ACHD is dictated not only by patient-related factors, but also by institutional and national considerations. Our management of these patients is highly influenced by the guidelines followed in the UK for heart transplantation [9]. In our experience, each patient referred for transplantation for ACHD follows a complex pathway, which is specific to that patient. However, in spite of the variability of diagnosis and management strategies, we can identify some common themes in managing these patients.

Patients with a high PVR pose a particular challenge. This challenge relates to either chronic left atrial hypertension, which is secondary to long-term obstruction from left heart outflow lesions, often in association with EFE, impaired systemic ventricular function, or a combination of these. With heart–lung transplant not a realistic option, due to the lack of availability of heart–lung blocks, one has little option but to consider non-transplant surgery for these patients. For those patients with aortic and/or mitral valve disease and/or EFE (Shone’s type), we have removed all residual obstruction with the option of mechanical support as either a bridge to recovery from that surgery, or in those whose ventricular function does not recover, longer term mechanical support to ensure a low left atrial pressure and potentially allow the PVR to fall (bridge to a low PVR and transplant or destination therapy). In fact, none of the patients with a high PVR associated with left-sided lesions required mechanical support, with mechanical support used only for left-sided lesions in a patient who could not be transplanted for other reasons (see below). Another subset of patients with high PVR are those with failing atrial inversion procedures. A range of non-transplant surgical strategies has been suggested with some success in carefully selected patients [10]. However, these options have not entered widespread practice and by the time transplant is considered, it is often too late. Our strategy for those with suitable PVR and who are not highly sensitized with HLA antibodies has been to actively pursue the transplant option with good outcomes when a donor becomes available. Currently, in the UK, these patients can be listed urgently if unwell enough to require inotropes. We have taken the view that those with a high PVR or too unwell to wait for an organ should be offered a mechanical support for the failing right ventricle in the same way that an...
equivalent cardiomyopathy patient would be. Our current strategy is to use a third-generation left ventricular assist device (VAD), i.e. HeartWare®. It is too early to draw any conclusions regarding whether it is possible to reduce the PVR using this strategy in this group of patients although the data from cardiomyopathy patients are promising [11, 12].

Patients who are highly sensitized due to the presence of HLA antibodies fare poorly compared with non-sensitized patients awaiting heart transplantation. Although we have carried out desensitization in two patients, desensitization is currently not considered as an option in adult patients requiring heart transplantation in the UK. The outcome is therefore adversely affected by longer waiting times to find a matched donor. We believe that a high-risk surgical treatment, if available, can be justified in this cohort.

The failing, non-systemic, right ventricle poses specific problems. The timing of pulmonary/tricuspid valve replacement/repair is an area of ongoing debate: balancing between intervening before irreversible right ventricular changes occur and carrying out unnecessary surgery [13–16]. Both the patients in this series had significantly dilated right ventricles with poor function and at least moderate impairment of the left ventricle. Both were beyond the point that surgical correction of the regurgitant lesions is usually advocated as evidenced by the fact that the referring surgical centre deemed valve replacement too high risk and referred for transplant. In one patient, conventional surgery was thought to be a better option and in the other patient, treatment was dictated by the patient being highly sensitized with HLA antibodies. A team approach with an admission for optimization preoperatively, repair/replacement of regurgitant valves, biventricular pacing and a gradual weaning of inotropes led to an extremely gratifying outcome for both patients. There has been significant recovery of ventricular function in both despite the starting point being ‘irreversible’. For patients with a Fontan-type circulation, success is well described for selected patients who have undergone upgrade of the circulation often with arrhythmia surgery and pacing [17, 18]. Both our patients had been turned down for this procedure by their referring units and we agreed that transplantation was the most appropriate option. However, in these two cases, the clinical situation necessitated carrying out the circulation upgrade and the risks are apparent from by the final outcome for both patients.

It could be argued that when primary transplant is not an option (high PVR, highly sensitized patients with HLA antibodies, unsuitable anatomy, too unwell to wait etc.), the decision-making process becomes easier. In these cases, the outcome of the non-transplant intervention can be compared directly with the outcome of continued medical/palliative treatment with the risk of waiting for transplant removed completely from the decision-making process. Although we have chosen to divide our patients into categories by diagnosis to illustrate the surgery undertaken, a more strategic way to divide the patients is into (i) those who can and should undergo heart transplantation but are too unwell to wait and will die without mechanical support; (ii) those who should have conventional surgery and not a transplant; (iii) those who cannot undergo cardiac transplantation and whose only options are conventional surgery, long-term mechanical support or palliation. Our management of the first of these is similar to that of patients with cardiomyopathy in the same situation: with escalating medical therapy and systemic VAD in suitable cases. The point at which the risk of conventional surgery outweighs the risk of transplant (including the wait for an organ) in patients who are ‘suitable’ for either is difficult to ascertain, leading to the overlap between the first two groups. The limited supply of donor organs and the responsibility to other patients on the heart transplant waiting list also needs to be given consideration. There are many patients who by any criteria are simply not suitable for heart transplant and it is this group of patients in particular that we feel that high-risk non-transplant surgery is justified.

When the opinion of the joint congenital heart disease/cardiac transplant meetings opinion is that non-transplant surgery should be undertaken, the patient is discussed with the referring unit to allow the option of the surgery being carried out locally. Invariably, the non-transplant surgery is turned down by the referring unit, not because of the complexity of the surgery, but because of the perceived need for mechanical support back up by both teams. This safety net that mechanical support seems to offer has allowed us to undertake surgery believed to be too high risk by other units. In fact, none of the patients with an anatomically correct circulation required a mechanical support following the correction of residual lesions (the patient undergoing mechanical support did so as part of the preoperative plan, as opposed to failure to come off bypass).

The difference in the outcome of these patients compared with systemic right ventricle and Fontan patients is so marked that grouping all patients together as we have performed in Fig. 1 may obscure the important learning points. The outcome is much better for the patients with an anatomically correct circulation as illustrated in Fig. 2 (P = 0.0028). The true extent of these cases for the UK is unknown, and it is not clear whether similar risk operations are being carried out by some units without a mechanical support back up or whether our series represents a small but unique population.

All of our patients had life-limiting symptoms and a high chance of dying in the next 12 months. It would be useful to illustrate the status of these patients using a scoring system. Several of these patients underwent relatively straightforward surgery with the high risk related to the patient’s preoperative condition, ventricular function and PVR, not the technical difficulty of the surgery itself. Although there are several scores available to compare the risk (Aristotle, RACHS and EuroSCORE etc.), our team and others have been frustrated by the lack of a suitable scoring system for ACHD patients.

Conclusions

The high mortality in patients with ACHD referred for cardiopulmonary transplantation without intervention is well recognized. We have described 14 patients in whom we carried out non-transplant surgery and have learned some important lessons. Patients with two ventricles and a systemic left ventricle can be operated on at an acceptable risk. Patients with two ventricles and a systemic right ventricle continue to have a high mortality despite conventional surgery and mechanical support, although some of the deaths in our series may have been avoidable with further experience. We believe that VAD is a reasonable alternative for these patients if there is no other option; however, in those suitable for transplant, this remains a bridging and not a destination therapy. Patients with a single ventricle, who have been turned down for Fontan upgrade by other centres, as expected, have a poor outcome from a ‘bail out’ procedure.
Although we are certain that the possibility of long-term VAD will be explored in this population as the technology develops, the only real options at present are cardiac transplantation or palliative medical therapy.

Conflict of interest: none declared.

REFERENCES


APPENDIX. CONFERENCE DISCUSSION

Dr L. Menicanti (Milan, Italy): This is an intriguing and interesting subject because, as you told us, the anatomical presentation can be very different. I think that the greatest message we can derive from your presentation is the fact that when there are two normally connected ventricles, the results you show us are very good, completely different than when there is a Fontan-like circulation or when the right ventricle is a systemic one.

So I would like to ask you, according to this experience, how do you consider the pulmonary resistance contraindications to treat this group of the patients with two normally connected ventricles? I mean, we can imagine that all patients with two ventricles should be operated with normal surgery. What do you think about this?

Mr Harper: If I could draw on the support of Dr Crossland for this question.

Dr D.S. Crossland (Newcastle-upon-Tyne, UK): The problem that we have got is that in the UK now, heart transplantation is dying, and is dying for ACHD patients in particular. There just are not enough donor organs to go around. So if we start transplanting patients with high pulmonary vascular resistance, above 5 Wood units is our usual cutoff, then the patients are not going to do well.

In fact all the things that we have tried to do in the past to try and make our congenital patients transplantable have failed for two reasons. One is that some patients die following transplantation, but secondly, even if what we do makes patients transplantable, patients still die on the waiting list. For example, one of the patients who survived a long time after Fontan upgrade subsequently went on to have desensitization. I know you have talked about pulmonary vascular resistance, and she was desensitized. It cost hundreds of thousands of pounds to desensitize her, and she died waiting for a heart. So despite making her transplantable, she could not be transplanted because of insufficient donor organs.

The patients who have a VAD put in to reduce pulmonary vascular resistance, once that pulmonary vascular resistance is down, still have to take the risk of going on the waiting list, and being on the waiting list is not a safe place because of the death rate on the waiting list in our patients.

Dr Menicanti: The other question that I would like to pose is, in the groups that you transplanted, what are the results of patients with two normally connected ventricles, are they comparable with normal surgery?

Dr Crossland: Our results were published in Heart three years ago, and our early mortality is relatively high in the same way as the ISHLT data. However, five to seven years out from transplant, the lines of the ACHD patient crosses the line from cardiomyopathy patients, and our survival beyond five years is as good as, and at ten years is better than patients with cardiomyopathy.

The Fontan patients are a big worry, though. We have recently transplanted eight Fontans and three of those have died, two acutely. The problem we have had with the Fontan patients is not the anatomical problems, because our surgeons are congenital surgeons, but they all seem to become vasoplastic following the operation. So it is a low systemic vascular resistance which we cannot seem to chase for some reason.

Dr M. Morshuis (Bad Oeynhausen, Germany): I have one short question. Is this the tip of the iceberg? What do you think?

Dr Crossland: We do not know what is happening to patients who are not referred, so we do not know if the operations we have done are brave or stupid or standard practice. We do not know which units are doing the operations themselves (without referring out of centre). We are a transplant unit, so our denominator here is patients who are referred for transplant. For example, I am sure everyone who is doing congenital transplantation knows that you will get a referral from one unit, and will think, my word, what are we going to do here because it is a patient who has got a Bi-V ICD, they have had a left ventricular overhaul, they tried a Glenn against high pulmonary vascular resistance and there are stents and coils everywhere. From other units, you get referrals for patients who do not even need a transplant. And so one of the problems with our data is the heterogeneous group created by the fact that our denominator is the status at referral. So I think it is the tip of the iceberg.

Dr Morshuis: And one other question concerns the failed Fontan: can it be done with an LVAD always, or are there also cases where you have to implant a BiVAD?

Dr Crossland: We have not put LVAD into any adult patients with a Fontan, but we have done four or five paediatric congenital single ventricle VADs which have all had an LVAD, but they had Fontan failure for different reasons. Patients who are failing because of high pulmonary venous pressure secondary to high EDP do very well on a VAD. Patients who are failing because of Fontan failure, so protein-losing enteropathy with high TPG, not high LA pressure, do less well on a VAD. And in these we have had to bail out and, fortunately, had an organ available quickly in children in that situation.

We will eventually have to put a VAD in an ACHD patient and we are still working out exactly how we would support the right side in that setting. But the first patient we do, we have already planned, will be a patient with

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EDP-driven Fontan failure as opposed to pulmonary vascular resistance Fontan failure. And if that works, the next stage will be to put some sort of VAD in on the right side for the other patients.

**Dr Morshuis:** And will all Fontans fail or not? Will some Fontans be 70 or 80 years of age? What is your opinion?

**Dr Crossland:** We have all got perfect patients who are doing extremely well with a Fontan. But I think by the time the patients we have now, our perfect patients, reach 80, then there will be a solution anyway in terms of mechanical support. So I am not worried about them getting to 80. I am worried about the non-perfect patients getting to 25.