Extracorporeal Membrane Oxygenation Support of the Fontan and Bidirectional Glenn Circulations

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Background. **Extracorporeal membrane oxygenation can provide effective mechanical circulatory support for the failing circulation in children. Patients with failing Fontan and bidirectional Glenn physiology present additional challenges both for extracorporeal membrane oxygenation cannulation and support. We report our institutional experience in patients with cavopulmonary connections who received extracorporeal membrane oxygenation.**

Methods. **We performed a retrospective review of 20 patients with cavopulmonary connections (14 Fontan and 6 bidirectional Glenn) who were supported with extracorporeal membrane oxygenation from a single, large pediatric tertiary care center.**

Results. **Of the 20 patients, ten were supported and decannulated successfully (50%) (two after cardiac transplantation), but only six (30%) are alive at follow-up. Of the 14 Fontan patients, seven (50%) were withdrawn from extracorporeal membrane oxygenation or died within 48 hours of decannulation due to lack of myocardial recovery or severe neurologic injury. All four adult-sized (> 40 kg) Fontan patients were withdrawn from extracorporeal support. The seven Fontan patients who were successfully decannulated survived to discharge, and five (35.7%) are alive at follow-up (median 35 months; range, 7 to 140 months). Of the six bidirectional Glenn patients, five died before hospital discharge and the lone survivor has neurologic injury at follow-up.**

Conclusions. **Patients with failing Fontan and bidirectional Glenn physiology present significant challenges to successful extracorporeal membrane oxygenation support. While the morbidity and mortality rates are high, there are select patients for whom extracorporeal support can be effective and lifesaving as a short-term resuscitative intervention.**

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Extracorporeal membrane oxygenation (ECMO) is an important mechanical support for the failing circulation in pediatric patients with both congenital and acquired heart disease [1–5]. Since ECMO support of patients with congenital heart disease was first reported in the 1970s [6, 7], ECMO has been used effectively for a variety of indications including preoperative hemodynamic support, low cardiac output after cardiopulmonary bypass (CPB), unexpected cardiac arrest, and as a bridge to cardiac transplantation [5, 8]. Survival of cardiac patients supported with ECMO has varied by institution, indication, and cardiac diagnosis [3, 9, 10]. Patients with single ventricle physiology and cavopulmonary connections following either a bidirectional Glenn (BDG) or Fontan operation present additional challenges for cannulation and circulatory support with ECMO. From 1986 to July 2002, the cumulative survival to hospital discharge of all cardiac patients supported with ECMO reported by the Extracorporeal Life Support Organization (ELSO) was 39%. During the same period, ELSO reported only a 24.8% survival for patients with Fontan and BDG physiology [11].

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The literature contains little specific data on patients with cavopulmonary connections supported with ECMO. Previous reports include a single case report [12] and descriptions of different institutional experiences with cardiac patients on ECMO that include small subsets of patients with Fontan or BDG circulations [9, 13–18] (reviewed in Table 1). The purposes of this study were to describe our institution's cumulative experience with Fontan and BDG patients who were supported with ECMO, including the indications for ECMO, details of cannulation, subsequent interventions, complications, and clinical outcomes, and to use our experience as a possible guide to improve management and outcomes.

Patients and Methods

Retrospectively, we reviewed the medical records of all patients with Fontan or BDG physiology supported with ECMO at Children's Hospital Boston. Permission was obtained from the hospital's institutional review board for medical record review. Patient characteristics recorded included age and weight at the time of cannulation and anatomic cardiac diagnoses. The patients' courses were documented, including indications for ECMO, site(s) of cannulation, duration of support, events during ECMO support including any subsequent inter-

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Table 1. Published Literature of Fontan and BDG Patients Supported With ECMO

Author	Year	Series size	Fontan/BDG patients
Kanter et. al [13]	1987	13 patients	1 Fontan, died
Klein et. al. [14]	1990	39 patients	4 Fontans, 4 died
Ziomek et. al. [15]	1992	24 patients	3 Fontans, 2 survived including 1 Fontan takedown on ECMO
Saito et. al. [12]	1993	1 patient	Fontan, survived
Dalton et. al. [16]	1993	29 patients	3 Fontans, 2 survived including 1 transplant from ECMO
Kulik et. al. [17]	1996	64 patients	18 cavopulmonary connections, 3 survived
Jaggers et. al. [18]	2000	35 patients (all infants)	1 BDG, died
Aharon et. al. [9]	2001	50 patients	2 Fontans, 2 survived; 1 BDG, died

BDG = bidirectional Glenn; ECMO = extracorporeal membrane oxygenation.

ventions, circuit complications, and follow-up. Patient outcomes were categorized as successful decannulation from ECMO (including cardiac transplantation), death due to withdrawal from ECMO support, survival to hospital discharge, and survival during follow-up. Due to the heterogeneous nature of this patient population, descriptive statistics alone were used to analyze the patients' courses and clinical outcomes.

Results

From December 1984 through June 2002, ECMO support had been used in 20 patients with cavopulmonary connections (14 Fontan and 6 BDG), which represents 8.4% of our total ECMO experience with cardiac patients. Eighteen of 20 patients were cannulated for venoarterial ECMO. Two patients were cannulated for venovenous ECMO, and one of these two patients was subsequently converted to venoarterial ECMO. Patients were cannulated onto a crystalloid primed circuit by our rapid response protocol as previously described [19]. The median age at cannulation for the Fontan patients was 4.4 years (range, 1 to 42 years) and the median weight was 14.5 kg (range, 7.9 to 82 kg). The median age at cannulation for the BDG patients was 17.1 months (range, 4.7 to 26.9 months) and the median weight was 8.4 kg (range, 4.4 to 10.5 kg). The anatomic diagnoses included hypoplastic left heart syndrome (n = 5), hypoplastic right ventricle (n = 5), heterotaxy variant (n = 4), double inlet left ventricle (n = 2), Ebstein's anomaly (n = 1), Shone's complex (n = 1), unbalanced atrioventricular canal defect (n = 1), and double outlet right ventricle with scimitar syndrome (n = 1).

Indications for ECMO

FONTAN. Indications for ECMO in the Fontan patients included myocardial failure (n = 8) or respiratory failure (n = 2) immediately following a fenestrated Fontan operation, progressive myocardial failure in patients who had previously undergone a Fontan procedure (n = 3), and cardiorespiratory arrest during an intervention in the cardiac catheterization laboratory (n = 1). Details of patient status before ECMO cannulation, including indications for ECMO, hemodynamic parameters measured, echocardiographic data, and inotropic support, are shown in Table 2.

The ten patients who were placed on ECMO after a fenestrated Fontan operation had all undergone a preoperative evaluation including echocardiography and cardiac catheterization that indicated adequate myocardial function, low pulmonary vascular resistance (< 4 Woods units), and pulmonary artery anatomy suitable for a successful Fontan operation. Any significant aortopulmonary or venovenous collateral circulation that might compromise a successful Fontan circulation was coil embolized at this time. Three were adult patients who had undergone a right atrial-to-pulmonary artery Fontan operation at an earlier age, and were converted to a lateral tunnel fenestrated Fontan circulation. One adult Fontan patient failed to wean from CPB because of severe ventricular dysfunction and was transitioned to ECMO in the operating room. Seven patients had postoperative myocardial failure with elevated Fontan pathway pressures ranging from 18 to 24 mm Hg and systemic hypotension, acidosis, arrhythmias, and(or) severe cyanosis. There was clinical evidence of premature fenestration closure in two of these seven patients. The median number of days from surgery to ECMO cannulation in these patients was two days (range, 0 to 11 days). One Fontan patient with postoperative respiratory failure and cyanosis secondary to respiratory syncytial virus (RSV) bronchiolitis was cannulated seven days postoperatively. We initially used venovenous ECMO, but this patient was transitioned to venoarterial ECMO for persistent severe hypoxemia. The other patient placed on ECMO because of respiratory failure had undergone conversion to a fenestrated lateral tunnel Fontan. He was noted to have a large baffle thrombosis on postoperative day 14, and after thrombolytic therapy was initiated he developed a large pulmonary hemorrhage and hypoxemia that could not be reversed with aggressive mechanical ventilation. He was therefore cannulated for venoarterial ECMO on postoperative day 16.

Three patients with progressively failing Fontan physiology and myocardial dysfunction underwent ECMO cannulation with the intention of using ECMO as a bridge to cardiac transplantation. These patients had been previously palliated with a fenestrated Fontan operation (2, 3, and 10 years prior) and subsequently developed congestive heart failure. All three patients underwent repeat catheterization to address reversible causes

Table 2. Indications for ECMO, Pre-ECMO Hemodynamics, ECHO Findings, and Inotropic Support for Fontan and BDG Patients

FONTAN

Pt	Age (yr)	Diagnosis	Indication	Inotropes	CPR	Pre-ECMO Hemodyn (mm Hg)	ECHO Dysfxn	AVVR	ECMO Wean	Hosp D/C
1	1.7	Hypoplastic RV	Postop low CO	Dopa, epi	20″	PAp 18, CAp 13	Severe	None	Y	Y
2	2.6	HLHS	Postop low CO, sepsis	Dopa, amrinone	None	PAp 18, CAp 10	Mild	Mild	N	N
3	2.2	DORV/heterotaxy	Postop low CO	Dopa, dobut, amrinone	None	PAp 24, CAp 14	Mod	Mild	Y	N
4	37.2	Hypoplastic RV	Postop arrhythmia	Dopa, phenyl	45″	PAp 20, CAp 10	Mod	Mild	N	N
5	1.0	DILV	Postop cyanosis	Dopa, epi	None	PAp 22	Mild	Trivial	Y	Y
6	2.2	DORV/heterotaxy	Postop fen closure	Dopa	None	PAp 24, CAp 16	Mild	Mod	Y	Y
7	4.0	DORV/scimitar	Postop fen closure	Dopa, epi	None	PAp 21, CAp 18	Mild	Mild	Y	Y
8	31.6	Hypoplastic RV	Postop pulm embolus	Dopa, epi	None	CVP 35	Mild	Mild	N	N
9	1.7	Hypoplastic RV	Postop RSV pneumonia	Dopa, milrinone	None	PAp 23, CAp 10	Severe	None	Y	Y
10	42.3	Hypoplastic RV	Failed CPB wean	Dopa, epi	None	NA	NA		N	N
11	4.3	HLHS	CHF, arrest	Dobut	20″	PAp 23, CAp20	Severe	Mild	N	N
12	5.3	HLHS	CHF, aortic thrombosis	Dopa, epi	None	CVP 19	Severe	Mild	Y	Y
13	14.5	DORV/heterotaxy	CHF, arrest	Dobut, milrinone	43″	PAp 16, CAp 5	Severe	Severe	N	N
14	4.4	Pulm atresia/heterotaxy	Cath event, arrest	None	30″	PAp 5, CAp 2	None	Trivial	Y	Y

BDG

Pt	Age (mos)	Diagnosis	Indication	Inotropes	CPR	Pre-ECMO Hemodyn (mm Hg)	ECHO Dysfxn	AVVR	ECMO Wean	Hosp D/C
1	4.6	DILV	Postop cyanosis	Dopa, epi	None	PAp 23, CAp 11	Severe	Mild	N	N
2	23.0	Ebstein's anomaly	Postop low CO, block	Dopa, epi	None	CAp 12	Severe	Severe	Y	N
3	9.6	HLHS/Shone's	Postop low CO	Dopa, epi	None	CVP 22	Severe	Trivial	N	N
4	21.4	Unbalanced AVCD	CHF, arrest	Milrinone	36″	PAp 19, CAp 16	Severe	Severe	N	N
5	26.9	HLHS	CHF, arrest	Dobut	45″	PAp 23, CAp 16	Mod	Severe	Y	Y
6	12.8	HLHS	Cath event, arrest	None	35″	PAp 14, CAp 8	Mild	Severe	Y	N

AVCD = atrioventricular canal defect; AVVR = atrioventricular valve regurgitation; BDG = bidirectional Glenn; CAp = common atrial pressure; cath = catheterization; CHF = congestive heart failure; CO = cardiac output; CPB = cardiopulmonary bypass; CPR = cardiopulmonary resuscitation; CVP = central venous pressure; DILV = double inlet left ventricle; dobut = dobutamine; dopa = dopamine; DORV = double outlet right ventricle; dysfxn = ventricular dysfunction; ECHO = echocardiogram; epi = epinephrine; fen = fenestration; hemodyn = hemodynamics; HLHS = hypoplastic left heart syndrome; mod = moderate; mos = months; N = no; NA = not applicable; PAp = pulmonary artery pressure; postop = postoperative; Pt = patient; pulm = pulmonary; RSV = respiratory syncytial virus; RV = right ventricle; Y = yes; yr = year.

of their decline such as the development of new collateral circulation. At the time of cannulation for ECMO, all three had been inpatients receiving inotropic infusions while awaiting cardiac transplantation. Two of these patients suffered a cardiac arrest and ECMO cannulation occurred during cardiopulmonary resuscitation (CPR). The third patient developed progressive low cardiac output with acidosis, and a large ascending aortic thrombus was demonstrated with echocardiography. He was subsequently cannulated for ECMO due to progressive myocardial failure, presumably from myocardial ischemia.

One Fontan patient had an acute cardiac arrest during fenestration closure at an elective cardiac catheterization that was caused by an air embolus to the coronary arteries. The patient had ventricular fibrillation unresponsive to conventional resuscitation maneuvers. Cannulation for ECMO was accomplished in the catheterization laboratory during CPR.

BDG. Indications for ECMO in the BDG patients included severe low cardiac output after cardiac surgery (n = 3), progressive myocardial failure (n = 2), and acute decompensation in the catheterization laboratory (n = 1) (see Table 2). All patients who required ECMO following the BDG operation had undergone a preoperative evaluation including echocardiography and cardiac catheterization to coil embolize any significant aortopulmonary or venovenous collateral circulation. Only one patient was considered high risk for a BDG on the basis of a preoperative pulmonary vascular resistance of 5.9 Woods units. All three postoperative BDG patients had elevated central venous or pulmonary artery pressures ranging from 20 to 23 mm Hg with cyanosis and(or) systemic hypotension before cannulation. The two BDG patients who underwent ECMO cannulation for progressive myocardial failure were awaiting cardiac transplantation and were receiving inotropic infusions. Both patients had an acute cardiac arrest and were cannulated during CPR. The BDG patient, cannulated during cardiac catheterization, became cyanotic and bradycardic during a hemodynamic study to assess his candidacy for Fontan surgery. He acutely deteriorated during catheter measurement of his distal pulmonary artery pressure. He was unresponsive to conventional resuscitative maneuvers and also underwent cannulation during CPR.

Cannulation and ECMO Flows

FONTAN. Cannula insertion sites in the Fontan patients included the femoral vessels alone (n = 5), neck vessels alone (n = 3), neck plus femoral vessels (n = 3), open chest alone (n = 2), and combined open chest and femoral vessels (n = 1). The distal tips of the cannulae were directed as centrally as possible towards the superior vena cava (SVC)-Fontan baffle-inferior vena cava (IVC) junctions. Table 3 provides the details of ECMO cannulation. The median maximal ECMO flow obtained for the 14 Fontan patients was 85 ml/kg/min (range, 56 to 120 ml/kg/min). Five of these patients needed placement of additional arterial and(or) venous cannulae to provide adequate ECMO circuit flow; two patients required the placement of a second venous cannula, one patient had the existing femoral venous cannula upsized, one patient had both an additional arterial and venous cannula placed, and one patient was converted from venovenous to venoarterial ECMO. Five of six patients with femoral arterial cannulae required insertion of an arterial bypass cannula (8Fr) to provide adequate perfusion to the distal extremity. Choice of cannulation site was limited by known vessel occlusion in eight of the 14 patients.

BDG. The sites of cannula insertion in the BDG patients was via both the neck and femoral vessels (n = 3), open chest alone (n = 2), and the neck plus open chest (n = 1) (see Table 3). The median maximal ECMO flow obtained for the five BDG patients supported with venoarterial ECMO was 133 ml/kg/min (range, 90 to 163 ml/kg/min). The two patients cannulated for heart failure and one patient cannulated for cardiac arrest during cardiac catheterization had venous cannulae placed in the SVC via the neck, as well as in the IVC via a femoral vein, to achieve adequate flows. The femoral vein was the site of initial venous cannulation, followed by the SVC in two of three of these patients. The third patient was first cannulated from the neck, followed by the femoral vein. Of the three postoperative patients, two had the venous drainage cannula placed directly into the right atrium through the open chest. The third postoperative patient was supported with venovenous ECMO for cyanosis. The venous drainage cannula was placed in the neck and the inflow cannula was placed in the right atrium through the open chest, thus relying on the patient's own myocardium to generate adequate cardiac output. Only one of BDG patients had a femoral arterial bypass catheter (4Fr) to treat distal leg ischemia. However, this was ineffective and the femoral artery cannula was removed and a new arterial cannula was placed in the carotid artery. Choice of cannulation site was limited by known vessel occlusion in three of the six BDG patients.

ECMO Management and Subsequent Interventions

The median duration of ECMO support was 85.5 hours (range, 12 to 368 hours) for the Fontan patients and 92 hours (range, 48 to 317 hours) for the BDG patients. All 20 patients were supported using a roller pump ECMO circuit without vacuum assist. Venous drainage was augmented by elevating the height of the patient's bed. After ECMO cannulation, inotropic support was successfully weaned in ten of 14 Fontan patients and in all six BDG patients. However, the four adult-sized Fontan patients ($>$ 40 kg) all received escalating doses of inotropic agents to maintain adequate mean arterial blood pressures on ECMO, due to limitations in the ECMO flows attained (less than 70 ml/kg/min in three of four patients). The patients who had been cannulated during CPR or near-arrest circumstances were mildly cooled to a core temperature 35°C for 12 hours after cannulation as a protective maneuver to preserve end-organ function. Ultrafiltration was used to augment extravascular fluid

Table 3. Cannulation Sizes and Sites, ECMO Flows, and Complications of Fontan and BDG Patients Supported with ECMO

Fontan Pt	Initial Cannulation			Additional cannula(e) (Fr)	Maximal flow ml/kg/min	Known occlusions	Hours	Complications
	Wt (kg)	Outflow (Fr)	Inflow (Fr)					
1	12	IVC-chest (28)	AO-chest (14)	N	120	LFV/RFV	21	
2	10	RFV (19)	RCCA (16)	N	80	RIJV/LFV	12	Inadequate venous return
3	11	SVC-chest (17)	AO-chest (12)	N	118	None	218	
4	66	LFV (21)	LFA (14)	LFA bypass	53	RFV	96	Neurologic injury, MSOD
5	7.9	LFV (12)	LFA (8)	LFA bypass	71	RFV/RIJV	134	Neurologic injury
6	9.3	LIJV (14)	LCCA (12)	N	108	None	108	
7	11.7	RIJV (15)	RCCA (14)	LFV (15)	103	LFA	72	
8	60	RIJV (27)	RFA (19)	RFA bypass	67	RFV/LFV	47	MSOD
9	9.1	DL-RFV (15)	None	*VV→VA*, LFA (15), LFA bypass	82	LIJV/LCCA	368	Sepsis
10	82	SVC-chest (24)	AO-chest (20)	N	56	LFA injury	15	Profuse bleeding, MSOD
11	14	RIJV (14)	RCCA (12)	N	100	None	133	Neurologic injury
12	15	RIJV (17)	RCCA (14)	N	87	None	26	
13	43	RFV (23)	RFA (15)	LFV (23), LFA (15), RFA bypass	116	None	163	MSOD
14	17	RFV (15)	LFA (12)	Upsized RFV (19)	81	None	75	

BDG Pt	Initial Cannulation			Additional cannula(e) (Fr)	Maximal flow ml/kg/min	Known occlusions	Hours	Complications
	Wt (kg)	Outflow (Fr)	Inflow (Fr)					
1	4.4	RIJV (10)	CA-chest (16)	N	*VV ECMO*	RFV/LFV	93	
2	10.5	CA-chest (23)	AO-chest (14)	N	133	None	91	Sepsis, MSOD
3	6.2	CA-chest (18)	AO-chest (14)	N	97	None	84	
4	9.2	RIJ (12)	RCCA (12)	RFV (10)	163	LFV/small RFV	317	Circuit thrombosis, MSOD
5	9.4	RFV (10)	RFA (8)	RIJ (10), RCCA (12), RFA bypass	149	LIJV/LFA	143	Neurologic injury
6	7.5	RFV (10)	RFA (8)	LSCV (8)	89	None	48	Neurologic injury

AO = aorta; CA = common atrium; DL = double lumen; Fr = French size; IVC = inferior vena cava; kg = kilograms; LCCA = left common carotid artery; LFA = left femoral artery; LFV = left femoral vein; LIJV = left internal jugular vein; LSCV = left subclavian vein; min = minute; ml = milliliter; MSOD = multisystem organ dysfunction; N = no; Pt = patient; RCCA = right common carotid artery; RFA = right femoral artery; RFV = right femoral vein; RIJV = right internal jugular vein; SVC = superior vena cava; VA = venoarterial; VV = venovenous; Wt = weight.

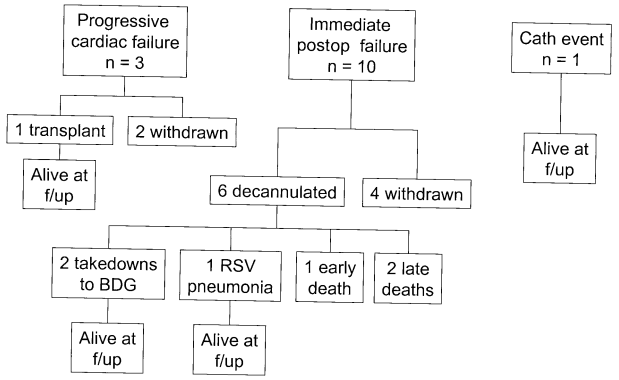

*Fig 1. Outcomes of Fontan patients supported with extracorporeal membrane oxygenation. (*BDG = *bidirectional Glenn;* cath = *catheterization;* f/up = *follow-up;* postop = *postoperative;* RSV = *respiratory syncytial virus.)*

removal in patients on ECMO for greater than 36 hours (7 Fontan, 3 BDG).

Interventions undertaken during or early after ECMO included three cardiac catheterizations (one hemodynamic study and one pulmonary artery stent in two Fontan patients, and a hemodynamic study in a BDG patient) and seven surgical procedures. The surgeries included three takedowns of the cavopulmonary anastomosis (two Fontan patients converted back to a BDG, both of whom survived, and one BDG patient who died in the operating room during shunt placement), one tricuspid valvuloplasty (BDG), one right coronary artery thrombectomy (BDG), and two heart transplants (1 Fontan, 1 BDG). Only two of the five patients placed on ECMO as a bridge to cardiac transplantation survived to be transplanted.

Complications

Complications included refractory bleeding (1 Fontan), circuit thrombosis (1 BDG), seizures or neurologic injury (3 Fontan, 2 BDG), and sepsis (1 Fontan, 1 BDG) (see Table 3). Despite the placement of additional cannulae to maximize ECMO flow and systemic perfusion, four Fontan and two BDG patients developed progressive multisystem organ dysfunction on ECMO. Three patients were withdrawn from ECMO due to irreversible multisystem organ dysfunction or circuit complications while awaiting cardiac transplantation.

Outcomes

Of 20 total patients, ten (50%) were decannulated successfully, eight (40%) survived to discharge, and six (30%) are alive at a median follow-up of 35 months (range, 7 to 140 months). Three of eight patients (38%) (2 Fontan, 1 BDG) cannulated during CPR survived. Of the 14 Fontan patients, eight patients (57%) were decannulated, with one early death within 48 hours of decannulation (Fig 1). All four adult-sized Fontan patients were withdrawn from ECMO for progressive multisystem organ dysfunction. Three of four Fontan patients who had additional cannulae placed to generate better ECMO flows were successfully decannulated and survived to discharge. The seven Fontan patients who were successfully decannulated all survived to discharge, and five are alive at follow-up (36%). The surviving Fontan patients include the two with failing circulations taken down to BDG in the immediate postoperative period, of whom neither has had reattempts at a Fontan operation. The Fontan patient transplanted from ECMO is alive at follow-up and thus far is free from significant complications after heart transplantation. The patient with postoperative respiratory failure from RSV bronchiolitis and the patient with the air embolus at catheterization also survived. These two surviving Fontan patients are functioning well without cardiovascular symptoms (New York Heart Association Class 1). The two late deaths of Fontan patients after discharge include one patient discharged to hospice care with severe neurologic injury. The second patient developed late progressive pulmonary vein stenosis and also died in hospice care.

Of the six BDG patients, the lone survivor underwent cardiac transplantation from ECMO (Fig 2). Following the transplant, neurologic impairment with basal ganglia injury became apparent. Of the five BDG nonsurvivors, two were withdrawn from ECMO, one died during takedown of the BDG, and two died after decannulation but before discharge because of neurologic injury and multisystem organ dysfunction. Of the three BDG patients who had additional cannulae placed to augment ECMO flow, two were decannulated successfully and one survived to discharge.

Comment

The cavopulmonary connection is a challenging circulation to support with ECMO. Previous studies suggest that survival of patients with cavopulmonary connections supported with ECMO is poor; only ten survivors among a total of 34 patients (29%) described in the literature survived [9, 12–18]. These reports have been limited in patient numbers and have not focused on specific factors unique to the management of Fontan and BDG patients. Although our experience also demonstrates a low overall

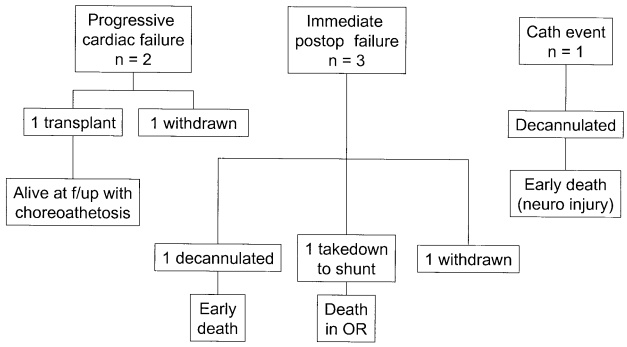

Fig 2. Outcomes of BDG patients supported with extracorporeal membrane oxygenation. (BDG = bidirectional Glenn; cath = *catheterization;* f/up = *follow-up;* neuro = *neurologic;* OR = *operating room;* postop = *postoperative.)*

survival rate of 30%, improved outcomes for this population may be possible with appropriate patient selection, a better understanding of optimal timing and technique for cannulation, and emphasis on end-organ protection on ECMO.

The Fontan circulation was supported more successfully with ECMO than the BDG circulation in our limited patient series. Subgroups of Fontan patients who were successfully supported with ECMO included patients with good myocardial function who had an acute, reversible event (air embolus at catheterization or RSV bronchiolitis) and those with low cardiac output after the Fontan operation who underwent Fontan takedown. Patients at risk for poor outcomes were adult-sized Fontan patients and patients with progressive failure of Fontan physiology with congestive heart failure who had deteriorated to cardiopulmonary arrest.

The BDG circulation was inadequately supported with ECMO in our patient series. While three of six BDG patients were successfully decannulated, all three of these patients had significant neurologic and other end-organ injury, and two of these patients died before hospital discharge. The only BDG survivor was transplanted from ECMO, but she suffered significant brain injury that became apparent after transplantation.

The poor outcome among our patients may be associated with the ongoing need for CPR at the time of ECMO cannulation. Patients with cavopulmonary connections and passive pulmonary blood flow are difficult to resuscitate effectively with conventional CPR. Patients with progressively failing Fontan and BDG physiology have significantly elevated systemic venous pressures, which are likely to decrease cerebral and other end-organ perfusion pressures and compromise oxygen delivery at baseline [20–23]. During CPR, the increase in intrathoracic pressure may restrict effective pulmonary blood flow and oxygenation in the Fontan and BDG circulation, as well as increase cerebral venous pressure that will further limit cerebral perfusion. Systemic output during CPR may also be limited by severe atrioventricular valve regurgitation (present in four of our patients who were cannulated during cardiac arrest). Once placed on ECMO, the development of end-organ dysfunction may be irreversible due to limitations of venous drainage and continued atrioventricular valve regurgitation. Therefore, these patients may be best managed by early consideration of ECMO support before deterioration in cardiopulmonary function occurs and end-organs have been severely compromised. Once end-organ dysfunction has occurred and myocardial recovery becomes unlikely, continuing ECMO support will not lead to a favorable outcome. Patients who received CPR for an acute arrest after a reversible event appeared to have better outcomes after CPR, likely due to preserved end-organ function before their acute event.

In our experience, the inability to maintain adequate venous drainage and systemic perfusion in cavopulmonary connections could be a contributing factor for neurologic injury and poor outcomes. This was most notable in the adult-sized Fontan patients. Using large venous cannulae from more than one site augmented venous return, but we often encountered limitations to cannulation of multiple central veins due to vessel occlusions from previous vascular injury. In the postoperative patient, cannulation via the open chest is an option for placement of larger venous cannulae, but this may prolong postoperative bleeding, as we observed in one of our adult Fontan patients. In the single, adult Fontan patient who achieved adequate ECMO flows (116 ml/kg/min) with the subsequent addition of another arterial and venous cannula, progressive end-organ dysfunction could not be reversed despite the improved ECMO flow.

The cannulation of the BDG patient is particularly challenging because of the separation of the systemic venous drainage (SVC to the pulmonary arteries and IVC to the heart). The two postoperative BDG patients supported with a venous cannula placed in the common atrium via an open chest, maintained full ECMO flows with the single venous cannula. However, both died due to lack of myocardial recovery. While the other three BDG patients supported with venoarterial ECMO eventually had a total of two venous cannulae placed (neck and femoral) to achieve full ECMO flow, two of these patients had the femoral cannula placed first, causing a delay in decompression of the SVC. We suspect that elevated venous pressure in the brain contributed to neurologic injury, which became apparent only after ECMO decannulation. In the BDG patient who had the first venous cannula placed percutaneously into the SVC, neurologic function appeared appropriate, but he died from ECMO circuit complications while awaiting cardiac transplantation.

Our experience suggests that significantly elevated SVC pressure, combined with a low systemic blood pressure in a BDG patient during cardiac arrest, places patients at high risk for neurologic injury due to inadequate cerebral perfusion pressure. Therefore, we recommend a cannulation strategy both for Fontan and BDG patients that includes initial decompression of the SVC and pulmonary arteries to maximize cerebral perfusion pressure during the acute resuscitation with a cannulation of head or neck vein, followed by IVC cannulation to incorporate additional lower body venous return to produce adequate ECMO flow. A single venous cannula is often insufficient for venous drainage in the Fontan and BDG circulation, even when the ventricle is ejecting well. After initial venous cannula placement, the presence of SVC syndrome or hepatic congestion suggests inadequate venous decompression, and the cannula may require repositioning or upsizing, if possible. Transthoracic or transesophageal echocardiography should also be considered to image the heart to assess decompression of the ventricle. Additionally, transcranial Doppler evaluation of cerebral blood flow velocity may also be considered to assess the adequacy of cerebral perfusion and venous decompression by the SVC cannula.

Future efforts to support the failing cavopulmonary circulation on ECMO should be focused on improvement of cerebral and other end-organ protection during the periods before and immediately after ECMO cannula-

tion. The incidence of severe neurologic injury among our patient population was 25%. The incidence of multisystem end-organ injury was 30%. Although we cooled the head with ice in an effort to decrease the cerebral metabolic rate during resuscitation, and continued mild cooling on ECMO after cannulation, we speculate there may be a benefit from deeper levels of hypothermia (32°C to 33°C) for cerebral protection [24]. A moderate degree of hypothermia can be achieved rapidly on ECMO and maintained for 12 to 24 hours with the benefit of full cardiovascular support. Moderate hypothermia, combined with maximizing ECMO flows and venous drainage to provide optimal perfusion, could be one strategy to minimize the neurologic and other end-organ injury that occurs during resuscitation of Fontan and BDG patients onto ECMO.

Although this is the largest ECMO experience reported to date in patients with a Fontan or BDG circulation, the relatively small number of patients with diverse problems limits the ability to make inferences or recommendations with certainty. Extracorporeal membrane oxygenation can be used successfully in patients with a Fontan circulation as a short-term resuscitative measure, or as a stabilization maneuver, pending surgical takedown of the Fontan. Extracorporeal membrane oxygenation support should be considered early before the development of end-organ dysfunction, which can be difficult to reverse on ECMO. Particular attention should be given to maximizing venous drainage and systemic perfusion using multiple venous cannulae, specifically with the initial cannula placed above, and the second cannula placed below, the diaphragm. While the survival of BDG patients treated with ECMO continues to be poor, early decompression of the SVC to maximize cerebral perfusion pressure during resuscitation may lead to better neurologic and overall outcomes in the future.

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