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Table 1 Effects of Parenteral Prostacyclin Therapy on Echocardiographic Variables

Echocardiogram Variable	n	Unit of Measurement	Echocardiogram prior to starting Prostacyclin Therapy	Echocardiogram after starting Prostacyclin Therapy	Difference between studies	95% Confidence Intervals of Difference between Studies	P value (Wilcoxon signed rank test)
RV Systolic Pressure	103	mmHg	82.9 ± 23.1	68.8 ± 22.0	14.0 ± 21.0	9.92 - 18.15	<0.001
RV Diastolic Diameter	92	cm	4.8 ± 0.8	4.6 ± 0.9	0.2 ± 0.7	0.05 - 0.36	0.005
LV Diastolic Diameter	99	cm	3.8 ± 0.8	4.2 ± 0.7	-0.4 ± 0.6	-0.57 - -0.32	<0.001
RV:LV End Diastolic Diameter Ratio	87		1.3 ± 0.	1.1 ± 0.3	0.2 ± 0.3	0.14 - 0.28	<0.001
LV Ejection Fraction	113	%	65.9 ± 11.8	67.0 ± 9.2	-1.1 ± 11.0	-3.17 - 0.94	0.280
RA Size	107	Normal/mild/mod/severely dilated (%)	11/13/33/43	11/19/33/37			0.260
LA Size	108	Normal/mild/mod/severely dilated (%)	74/18/6/2	64/23/8/5			0.024
RV Function	108	Normal/mild/mod/severely decreased (%)	9/19/32/40	24/23/30/23			<0.001
RV Size	111	Normal/mild/mod/severely dilated (%)	2/9/34/55	24/23/29/23			<0.001
Tricuspid							

Regurgitation 114 Trace or none /mild/mod/severe regurgitation (%) 2/34/37/272/47/34/170.004 Pericardial Effusion 106 Absent/present (%) 66/3456/440.047 Inferior Vena Cava 67 Normal/dilated but collapsible/dilated and failed to collapse 46/31/2254/24/220.689

1174**Donor Bicuspid Aortic Valve: Double Trouble or No Problem?**

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Introduction: Valvular disease in pediatric donor hearts may be a relative contraindication to graft use. Outcomes following use of donor hearts with milder forms of valvular disease have not been previously reported. We describe 4 cases of pediatric heart transplantation (HTx) in which the donor heart had a bicuspid aortic valve (BAV).

Case Report: Of the 469 HTx performed at our center since 1985, 4 donor hearts had a BAV. All recipients were female; median age 11 years (range 0.3 to 19 years). In all cases, the BAV was not discovered until after HTx. All donors were less than 30 years old. The patients were followed for a median of 6 years (range 2 to 9 years) with all patients alive at last follow up. Two patients transferred to adult care and 2 patients are followed by our clinic. In follow up, no patient required an aortic valve intervention or had infective endocarditis. At last review, no patient had greater than mild aortic insufficiency or more than mild aortic stenosis. Three patients developed mild to moderate left ventricular hypertrophy in the first year post-transplant that improved over time. One patient experienced a peri-operative embolic stroke at time of transplant unrelated to the bicuspid aortic valve.

Summary: On short- and intermediate-term follow up, donor hearts with bicuspid aortic valve demonstrated acceptable graft longevity and valvular function. A functionally normal bicuspid aortic valve in a pediatric heart transplant donor should not be a contraindication to organ acceptance.

Patient #	Patient Data			
	1	2	3	4
Age at Transplant (years)	19	6	17	0.3
Time Followed (years)	4.8	9	7	2
Fused Cusps	Left and right	Left and right	Right and non-coronary	Left and right
Peak Gradient (mmHg) at Last Follow Up	No gradient	No gradient	13	16
Insufficiency at Last Follow Up	Mild	None	Mild	None
Left Ventricular Hypertrophy at Last Follow Up	Mild	None	None	None
Endocarditis	No	No	No	No
Stroke	No	No	No	Yes - perioperatively

1175**Extracorporeal Membrane Oxygenation Dependent COVID19 Hospital Transfers**

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Introduction: The World Health Organization has recorded over 8 Million cases of COVID19 as of October 2020. Despite receiving appropriate lung protective ventilation and medical treatment, some of these patients develop refractory hypoxemia and acute respiratory distress syndrome. Extracorporeal membrane oxygenation has been recognized as a lifesaving therapy for patients with ARDS secondary to COVID19. There are few centers in the United States equipped with the necessary staff and the experience to take care of such critically ill patients. Some patients are too ill to be transferred with conventional mechanical ventilation, and they require interhospital transport while on ECMO.

Case Report: We have developed a highly specialized ECMO Deployment Team dedicated to the cannulation and transport of COVID19 patients while on venous-venous (VV) ECMO or venous-arterial (VA) ECMO. We use routine bedside ECMO cannulation via bilateral femoral vessels configuration at the outside hospital. The patient is stabilized and transported by air or ground to one of our affiliated hospitals. Here we present a series of five patients who were cannulated by our team at an outside institution and transported while on ECMO support to one of our three system hospitals. Patient ages ranged between 49-64 years old. Four patients required VV ECMO for severe hypoxemia secondary to COVID19 ARDS. One patient required VA ECMO due to viral myocarditis secondary to COVID19. Time on ECMO ranged from 9-33 days. Three of the five patients recovered successfully and were discharged home, rehab or LTAC. One patient is still currently on ECMO and one patient is deceased. There were no reported or documented transmission of COVID19 to the members of the ECMO deployment team.

Summary: The potential for survival of the critically ill due to COVID19 often demands a higher level of care. However, stable transport to an appropriate institution presents a limiting factor. Our method of a dedicated ECMO Deployment Team appears to provide favorable outcome for these patients.

Patient	Age	Sex	BMI	PMH	Indication	Pre-ECMO support	Cannulation Sites (for Drainage & 2nd return)	Drainage cannula size	Return cannula size	Transport mode	Days on ECMO	ECMO outcome	Length of Stay
1	56	M	33.2	none	Pulmonary	Max Vent support, Prop, Vaso, Nimblek, Flet	L Fem V & R Fem V	26	21	Helicopter	9	recovered	19
2	60	M	31.7	HTN, Hypertrophic cardiomyopathy	Pulmonary	Prop, Nimblek, Prop, Flet, Vaso, Epi, Max Vent support	L Fem V & R Fem V	23	21	Ground	23	recovered	26
3	64	F	30.3	CAD, HLD	Cardiac (viral myocarditis)	SABP, Levo, Vaso, DBTX	R Fem V & R Fem A	21	16	Fixed wing	11	recovered	30
4	54	M	34.2	HTN, A-fib	Pulmonary	Max Vent, Prop, Nimblek, Prop, Vaso, Flet	L Fem V & R Fem V	25	21	Helicopter	33	deceased	33
5	49	M	29.4	DVT	Pulmonary	Max Vent support, prop, Nimblek, Prop, Vaso, Flet	L Fem V & R Fem V	25	19	Ground	16 (as of 10/26)	current treatment	16 (as of 10/26)

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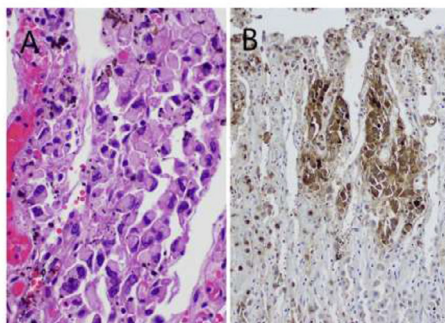
Metastatic Signet Ring Cell Carcinoma Masquerading as Acute on Chronic Thromboembolic Pulmonary Hypertension Requiring ECMO

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Introduction: Pulmonary thromboembolism can result from underlying hypercoagulability. Malignancy can cause hypercoagulability; however, gastrointestinal malignancy is rare in pediatric patients. We present the case of an adolescent female with severe pulmonary thromboemboli and subsequent right heart failure requiring venoarterial extracorporeal membrane oxygenation (VA ECMO) who was found on autopsy to have widely metastatic signet ring cell carcinoma (SRCC) of gastric origin.

Case Report: An obese 16-year-old female taking combination oral contraceptives for menorrhagia presented to an outside facility with chest pain, dyspnea, and vomiting. She had no recent history of travel, surgery, or trauma. Her father had been diagnosed with protein C/S deficiency, though she had tested negative for this. She had inverted T-waves with elevated troponin. Chest computed tomography angiogram revealed bilateral pulmonary arterial thrombi and evidence of right heart strain. After transfer to our institution, she developed cardiogenic shock from right ventricular failure. She was placed on peripheral VA ECMO. After failing to wean from ECMO, she was evaluated by our lung transplant committee but was declined for listing. She continued to deteriorate despite ECMO support and maximal medical therapy, developing refractory sepsis and coagulopathy. After one month on ECMO, decannulation was performed. She expired two days later due to multi-system organ failure. Autopsy revealed widely metastatic diffuse SRCC of gastric origin that was not seen on multiple rounds of axial imaging and endoscopies (Figure 1).

Summary: This report highlights an unusual cause of pediatric hypercoagulability resulting in pulmonary embolism and right heart failure requiring mechanical circulatory support. It is imperative to maintain a broad differential diagnosis for pediatric patients requiring mechanical circulatory support.



A. Multiple clusters of atypical signet ring cells were seen in the autolyzed gastric mucosa [H&E, 40X original magnification].
B. Clusters of atypical signet ring cells in autolyzed gastric mucosa highlighted by immunohistochemical staining for carcinoembryonic antigen (CEA) [20X original magnification].

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Hermansky-Pudlak Syndrome and Lung Transplantation: A Single Center Case Series

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Introduction: Hermansky-Pudlak Syndrome (HPS) is a rare, autosomal recessive disorder defined by oculocutaneous albinism, platelet storage disorder with resultant bleeding diathesis and pulmonary fibrosis leading to end-stage lung disease. Despite known bleeding diathesis, patients with HPS-related pulmonary fibrosis have successfully tolerated lung transplantation. We detail the lung transplant evaluation of 10 patients with HPS-fibrosis, as well as the outcomes of 6 patients transplanted at a single center in the United States.

Case Report: Between 2003 and 2019, 10 patients with HPS-related pulmonary fibrosis underwent lung transplantation evaluation at our center (table 1 & 2). Three patients were declined listing. Reasons included advanced multi-system disease, obesity, and severe protein malnutrition placing candidates at high risk for perioperative complications and morbidity. Seven patients were listed for lung transplant (table 3), with six undergoing transplantation. Three patients tolerated perioperative ECMO. The majority received intraoperative prophylaxis for their bleeding diathesis. Reported estimated blood loss was increased in those that did not receive intraoperative prophylaxis. One patient expired in the immediate post-operative period due to arterial ECMO cannula dislodgement. No allograft developed fibrosis. Two patients developed chronic rejection, but allograft lifespans were greater than 6 years. For surveillance biopsies, the majority received routine pre-treatment with desmopressin with no significant bleeding. One patient did not and sustained substantial pulmonary hemorrhage, requiring intubation.

Summary: Early lung transplant evaluation can be pursued in patients with HPS-pulmonary fibrosis. This case series illustrates that these patients tolerate extra-corporeal membrane oxygenation as a bridge to transplantation, and can safely undergo transplant surgery and surveillance transbronchial biopsies with the use of anti-hemorrhagic prophylaxis.

	All Evaluated N = 10	Listed for Transplant N = 7	Declined Listing N = 3	P value
Mean Age	47	55.5	53	
Female Sex	5	4	1	
HPS Subtype -1	8	6	2	
Puerto Rican Ancestry	8	5	3	
Former Smoker	4	2	2	
Comorbidities				
Chronic Kidney Disease	2	1	1	
Diabetes	3	2	1	
History of Skin Cancer	1	1	0	
Obesity (BMI > 30)	5	4	1	
Obstructive Sleep Apnea	3	2	1	
Prolonged QTc	3	3	0	
Pulmonary Hypertension	7	4	3	
Right Ventricular Dysfunction	3	1	2	
Radiographic Findings				
Non-specific Interstitial Pneumonia	4	3	1	
Usual Interstitial Pneumonia	3	2	1	
Mean Forced Vital Capacity (L)	2.14 (43%)	2.37 (41%)	1.43 (37%)	.265
Median DLCO (ml/min/mmq)	8.65 (32%)	11.8 (37%)	2.1 (6%)	.086
Median 6-minute walk test distance ft	1305	1417	500	.056

	All Evaluated N = 10	Listed for Transplant N = 7	Declined Listing N = 3
History of symptoms/signs bleeding diathesis	6	3	3
Prior blood product transfusion	5	3	2
Albuminuria	2	2	1
Prior surgical intervention - no complications	4	4	0
Platelet Aggregometry performed	5	5	0

	Transplanted N = 6	Prephylaxis with DDAVP/ACA N = 4	No prophylaxis N = 2	P value
Female Sex	3			
Single lung transplant	3			
Mean Ischemic time (min)	320			
Single lung Transplant Mean Ischemic time (min)	236			
Human Leukocyte Antigen antibodies	2			
Extra-corporeal membrane oxygenation	5			
Desmopressin (DDAVP)	4			
Anticoagulant (ACA)	1			
Mean estimated blood loss (ml)	320	231	500	.538
Mean blood products administered (ml)	975	647	363	
Transverse days	2.5	2	3.5	.345
Inhalation days	2.67	2.25	3.5	.396
ICU days	8.8	4.5	4	.973
Bleeding prophylaxis prior to transbronchial biopsies	5			

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Behcet's Disease Unmasked after Heart Transplantation

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Introduction: Behcet's disease (BD) is an autoinflammatory, multisystem vasculitis. Cardiac involvement is rare and clinical diagnosis is challenging. We present a case of Behcet's disease diagnosed after heart transplantation (HTx).