Practice Variation in Single-Ventricle Patients Undergoing Elective Cardiac Catheterization: A Report from the Congenital Cardiac Catheterization Project on Outcomes (C3PO)

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ABSTRACT

Objective. The objective of this study was to investigate variation in practice surrounding elective cardiac catheterization in patients with single-ventricle (SV) congenital heart disease.

Design. Patient and procedural characteristics and outcomes during SV catheterization were collected prospectively from eight centers using a web-based registry (Congenital Cardiac Catheterization Project on Outcomes). We attempted to identify a population of elective procedures by limiting the cohort in case type and timing. Cases were then stratified by stage of SV palliation (pre-bidirectional cavopulmonary anastomosis [pre-BCPA], pre-Fontan and post-Fontan) and limited by age. Subcohort analysis was performed by mode of airway management (assisted vs. spontaneous ventilation). Institutional variation was assessed.

Results. Between 2/2007 and 6/2010, 1459 (10.1%) of 14 467 cases in the registry met the inclusion and exclusion criteria, including 326 pre-BCPA, 571 pre-Fontan and 562 post-Fontan procedures. Median patient age was 0.4 (interquartile range 0.3, 0.5), 2.6 (1.0, 3.4) and 9.6 (5.2, 15.4) years and weight was 5.6 (4.8, 6.4), 12.2 (10.5, 14), and 26.3 (16.6, 51.8) kg in the pre-BCPA, pre-Fontan and post-Fontan cohorts, respectively. Cases were more commonly diagnostic in the pre-BCPA cohort (57%) whereas they were more commonly interventional in the pre-Fontan (69%) and post-Fontan (77%) cohorts. At least one adverse event (AE) occurred in 210 cases (14.4%) overall, including 20% of pre-BCPA, 11% of pre-Fontan and 14% of post-Fontan catheterizations. Mode of airway management was associated with statistically significant, but clinically small differences in hemodynamic measures in the pre- and post-Fontan cohorts, but not in the pre-BCPA group. Considerable practice variation exists across centers with variability in airway management, AE rate, case type, interventions performed and fluoroscopy time, in all SV cohorts.

Conclusions. Elective catheterization in SV patients, frequently performed with intervention, is common among US congenital cardiac centers. However, important variation in practice exists during these procedures in SV patients, at all stages of palliation.

Key Words. Congenital Heart Disease; Pediatric; Single Ventricle; Cardiac Catheterization; Practice Variation; Quality Improvement

Introduction

Despite major advances in perioperative care, management of congenital heart disease (CHD) with univentricular physiology remains highly complex and is associated with high rates of morbidity and mortality.[1,2] Most patients require staged surgical palliation beginning in the newborn period, followed by additional operations at approximately 3–6 months and 2–4 years of

age.[3] Given the substantial perioperative and "interstage" risks inherent to this population, detailed preoperative risk stratification is necessary in all cases. Most centers routinely employ the use of cardiac catheterization for invasive hemodynamic and anatomic assessment prior to subsequent staged surgical palliation, although the use of cardiac magnetic resonance (CMR) has recently emerged as an alternative strategy for noninvasive assessment.[4,5] Because of the small cohort of univentricular CHD followed at any one center, and lack of evidence-based guidelines surrounding the use and performance of cardiac catheterization in this population, there is potential for important inter- and even intra-institutional variability in care delivery.

Detailing variation in care across CHD centers is necessary to inform development of future interventional studies, quality improvement (QI) efforts and evidence-based guidelines. Although variation in perioperative management of single-ventricle patients surrounding stage I surgery and the subsequent interstage has recently been described,[6–9] variation in practice surrounding cardiac catheterization in this population has yet to be examined rigorously. To date, descriptions of catheterization-related practice in the univentricular CHD population have largely been confined to single-center reports detailing transcatheter interventions and associated outcomes.[10,11] Variation in intervention rates for post-Norwood recoarctation has been investigated in a recent report from the Pediatric Heart Network (PHN) Single-ventricle Reconstruction trial, which found wide variability across participating centers (0–50% intervention rates).[12] Important institutional variation in patient and procedural characteristics and outcomes associated with elective catheterization in single-ventricle patients, such as the age at pre–stage II catheterization, mode of airway management, rate of intervention and risk of adverse events (AEs) have not been rigorously described. A better understanding of the nature and degree of practice variation that exists in this high-risk population may allow for development and implementation of multicenter QI efforts aimed at reducing variation and improving clinical outcomes. Such QI initiatives are already underway in the congenital cardiac catheterization laboratory, with an early single-center effort related to assessment and management of congenital aortic valve stenosis having yielded positive results.[13]

Therefore, using the existing Congenital Cardiac Catheterization Project on Outcomes (C3PO) dataset,[14] we aimed to (1) describe patient and procedural characteristics, including interventions performed; (2) evaluate procedural outcomes including AEs; and (3) describe the institutional variation in elective catheterization practice in single-ventricle patients at three key time points: pre-bidirectional cavopulmonary anastomosis (pre-BCPA), pre-Fontan, and following Fontan completion (post-Fontan), outside of the immediate postoperative period.

Methods

Patient and procedural data were prospectively collected in the C3PO registry using a web-based data entry tool at eight participating institutions between February 1, 2007 and June 31, 2010 (the duration of data collection during which time cases were audited). Data collection, validation and auditing methods have previously been reported.[15] Institutional Review Board approval was obtained and maintained at all participating centers throughout the data collection and analysis period. In accordance with participant agreements, all interventional cardiologists who contributed to the dataset reviewed and approved the article prior to peer review submission.

Population and Characteristics

While the C3PO registry does not categorize procedures as "pre-Glenn," "pre-Fontan," or "post-Fontan," we attempted to create these cohort populations for the analysis by applying a series of inclusion and exclusion criteria. Figure 1 demonstrates the flow of patient selection. First, we identified all patients coded as single-ventricle physiology in the database (identified by a distinct diagnosis data element recorded in all cases). Cases were excluded if not classified as diagnostic or interventional case type (n = 73, e.g., surgical hybrid). To eliminate urgent or emergent procedures, cases were further excluded if performed within 1 month of a surgical intervention (n = 367), at less than 1 month of age (n = 12), or not performed electively (n = 500). The cohort was then stratified by subdiagnosis data elements and limited by age according to the following: (1) single-ventricle s/p shunt or balanced circulation and less than 12 months of age (pre-BCPA); (2) single-ventricle s/p Glenn and less than 6.5 years of age (pre-Fontan); and (3) single-ventricle s/p Fontan (post-Fontan). Age cutoffs were deter-

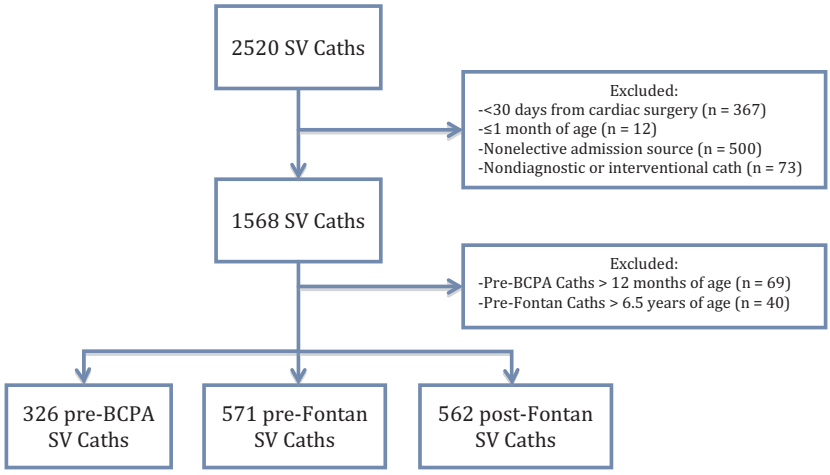

Figure 1. Flow diagram of patient selection for the study.

mined based upon the age-distribution curves for the pre-BCPA and pre-Fontan groups, in order to limit inclusion of "outlier" cases, as our objective was to characterize "routine" center practices.

Patient characteristics recorded include gender, age, weight, presence of a genetic syndrome, and/or noncardiac diagnosis. Procedural characteristics include admission source, airway management (spontaneous ventilation or assisted ventilation), case type (diagnostic or interventional), intervention(s) performed, use of inotrope, case duration, fluoroscopy time, contrast dose and packed red blood cell (PRBC) transfusion. Interventions were defined by interventional target and limited in count to one per target per case. For example, a patient undergoing both right and left branch pulmonary artery (PA) angioplasty would count once for each therapy, while a patient undergoing angioplasty of multiple right PA lobar segments would count just once. Hemodynamic variables in this report include preprocedural hemoglobin, cardiac index, ventricular end diastolic pressure, mixed venous saturation (MV_{sat}), systemic arterial saturation (SA_{sat}) and PA mean pressure.

An AE in the C3PO registry has been defined as any anticipated or unanticipated event, for which avoidable injury could have occurred, or did occur, as a potential or definite consequence of performance of the catheterization.[15,16] AEs were recorded at the time of identification either during the procedure or following the procedure. Information recorded included event name, a brief description, and attribution. All AEs underwent independent review by two interventional cardiologists for consistent severity classification using established standardized nomenclature on a five-level severity scale, based on clinical impact (1—*none*; 2—*minor*; 3—*moderate*; 4—*major*; 5—*death*).[15–17] High severity AEs (HSAE) were defined as level 3, 4, or 5 events.

Statistic Analysis

Analyses were performed separately for each of the three single-ventricle cohorts: (1) pre-BCPA; (2) pre-Fontan; and (3) post-Fontan. Categorical variables are summarized as frequency and percentage; continuous variables are summarized as median and interquartile range (IQR). Chi-square analysis was used to test for differences in categorical variables and Wilcoxon rank-sum for continuous variables. A P value ≤.05 was considered statistically significant. To assess for potential differences in hemodynamic measures by mode of airway management, each group was further stratified into subgroups by mode of airway management: spontaneous ventilation vs. assisted ventilation (positive pressure). Additionally, to assess for institutional variation in practice, patient/procedural characteristics and outcomes were evaluated by single-ventricle cohort, stratified by the contributing institution.

Results

Study Population

Between February 1, 2007 and June 31, 2010, 14 467 cases were entered into the C3PO registry. Of the 2520 (17%) cases that were performed in

Table 1. Patient and Procedural Characteristics by Single-Ventricle Group

Characteristic	Pre-BCPA *N* = 326	Pre-Fontan *N* = 571	Post-Fontan *N* = 562
Age (year)	0.4 (0.3, 0.5)	2.6 (1.9, 3.4)	9.6 (5.2, 15.4)
Weight (kg)	5.6 (4.8, 6.4)	12.2 (10.5, 14.0)	26.3 (16.6, 51.8)
Male gender	174 (53%)	350 (61%)	332 (59%)
Presence of genetic syndrome	30 (9%)	28 (6%)	30 (5%)
Presence of noncardiac diagnosis	83 (25%)	123 (22%)	164 (29%)
Spontaneous ventilation	64 (20%)	106 (19%)	213 (38%)
Interventional case type	141 (43%)	395 (69%)	432 (77%)
Use of inotrope	18 (6%)	18 (3%)	42 (7%)
Case duration (min)	93 (71, 124)	111 (85, 142)	127 (96, 162)
Fluoroscopy time (min)	27 (18, 41)	29 (19, 43)	37 (24, 55)
Contrast dose (mL/kg)	5.2 (3.9, 6.7)	4.8 (3.8, 6.2)	3.9 (2.6, 5.4)
Transfusion of PRBC	50 (15%)	17 (3%)	11 (2%)

Values are displayed as N (%) or median (interquartile range). PRBC, packed red blood cells; pre-BCPA, pre-bidirectional cavopulmonary anastomosis.

patients coded as having single-ventricle physiology, 1459 (10%) met the inclusion and exclusion criteria and were included in the analysis (Figure 1).

Pre-BCPA

Three hundred twenty-six cases meeting criteria performed in 309 patients were classified as pre-BCPA elective cardiac catheterizations. Patient and procedural characteristics are summarized in Table 1. Pre-BCPA cases were performed at a median age of 0.4 (0.3, 0.5) years and weight of 5.6 (4.8, 6.4) kg. A genetic syndrome was present in 9% of patients. Median catheterization case duration was 93 (71, 124) minutes, with a median contrast dose of 5.2 (3.9, 6.7) mL/kg and fluoroscopy time of 27 (18, 41) minutes. PRBC transfusion was most common in the pre-BCPA population (15%). The airway was managed with assisted ventilation in 80% of cases. Catheterization procedures were more commonly diagnostic than interventional in the pre-BCPA group (57% diagnostic). Interventional procedures performed are summarized in Table 2. The most common interventions included balloon angioplasty of the aorta (n = 29, 8.8%), PA (n = 21, 6.4%) and pulmonary vein (n = 11, 3.4%), as well as occlusion of a systemic to pulmonary collateral (n = 46, 14.1%) and veno-venous collateral (n = 21, 6.4%). At least 1 AE occurred in 64 (20%) pre-BCPA cases; 28 (9%) were classified as HSAEs with no deaths (Table 3). There were 6 major events in the pre-BPCA group, including complete heart block requiring cardiopulmonary resuscitation (CPR; n = 3), atrial tachyarrhythmia requiring electric cardioversion (n = 1), postextubation laryngospasm requiring CPR (n = 1) and bradycardia/hypotension requiring CPR (n = 1).

Pre-Fontan

Five hundred seventy-one cases performed in 491 patients were classified as pre-Fontan elective cardiac catheterizations. Pre-Fontan cases were performed at a median age of 2.6 (1.9, 3.4) years and weight of 12.2 (10.5, 14) kg. Median case duration was 111 (85, 142) min, with a median contrast dose of 4.8 (3.8, 6.2) mL/kg and fluoroscopy time of 29 (19, 43) min. The airway was managed with assisted ventilation in 81% of cases. Interventional case type predominated in the pre-Fontan population (69%). Occlusion of a systemic to pulmonary collateral (n = 274, 48%) and veno-venous collateral (n = 95, 16.6%) were performed frequently. Other common interventions included balloon angioplasty of the PA (n = 61, 10.7%) or stent implantation (n = 34, 6%) and balloon dilation of the aorta (n = 25, 4.4%). At least 1 AE occurred in 65 (11%) pre-Fontan cases, with 19 (3%) classified as HSAEs. There were four (<1%) major events in the pre-Fontan group, including complete heart block requiring CPR (n = 2), hypotension requiring CPR (n = 1) and SVC perforation (during IJ sheath placement) with resultant hemothorax requiring surgical repair (n = 1), and no deaths.

Post-Fontan

Five hundred sixty-two cases performed in 490 patients were classified as post-Fontan elective cardiac catheterizations. Post-Fontan procedures were performed at a median age of 9.6 (5.3, 15.4) years and weight of 26.3 (16.6, 51.8) kg. The post-Fontan group was characterized by a broad age range, consistent with the long duration of the post-Fontan period and variable utilization of elective catheterization in clinical practice throughout the post-Fontan course. Median case

Table 2. Interventional Procedures by Single-Ventricle Group

Procedure*	Pre-BCPA *N* = 326	Pre-Fontan *N* = 571	Post-Fontan *N* = 562
Balloon angioplasty	72 (22%)	105 (18%)	100 (18%)
Right PA (branch and lobar segments)	7	16	19
Left PA (branch and lobar segments)	14	45	28
Systemic to pulmonary shunt	8	5	—
Aorta	29	25	6
Systemic vein	4	13	14
Pulmonary vein	11	12	7
Intracardiac	9	4	33
Other	2	8	4
Stent placement	22 (7%)	43 (8%)	96 (17%)
Right PA (branch and lobar segments)	1	3	4
Left PA (branch and lobar segments)	1	31	33
RV-PA conduit	8	—	—
Systemic to pulmonary shunt	4	1	—
Aorta	5	3	4
Intracardiac	2	2	44
Other	4	3	19
Stent redilation	10 (3%)	32 (6%)	48 (9%)
Right PA (branch and lobar segments)	2	3	3
Left PA (branch and lobar segments)	1	20	25
Aorta	1	5	4
Intracardiac	—	—	10
Other	6	5	9
Device occlusion	62 (19%)	330 (58%)	347 (62%)
Fenestration or baffle leak	—	1	195
Venous collateral or LSVC	21	95	118
Systemic to pulmonary collateral	46	274	123
Other	—	3	10
Creation of interchamber connection	1 (<1%)	9 (2%)	14 (2%)

*Cases can include more than one intervention but no patient can be counted more than once for any specific (i.e., unique row) intervention. Values are displayed as N (%). Intracardiac, Fontan baffle, fenestration, or atrial septum; LSVC, left superior vena cava; PA, pulmonary artery; Pre-BCPA, pre-bidirectional cavopulmonary anastomosis; RV, right ventricle.

Table 3. Occurrence of Adverse Events by Single-ventricle Group

	Pre-BCPA *N* = 326	Pre-Fontan *N* = 571	Post-Fontan *N* = 562
Any adverse event	64 (20%)	65 (11%)	81 (14%)
Highest severity adverse event			
1. None	4 (1.2%)	7 (1.2%)	10 (1.8%)
2. Minor	32 (9.8%)	39 (6.8%)	50 (8.9%)
3. Moderate	22 (6.7%)	15 (2.6%)	15 (2.7%)
4. Major	6 (1.8%)	4 (0.7%)	5 (0.9%)
5. Death	0 (0%)	0 (0%)	1 (0.2%)

Values are displayed as N (%). Pre-BCPA, pre-bidirectional cavopulmonary anastomosis.

duration was 127 (96, 162) min, with a median contrast dose of 3.9 (2.6, 5.4) mL/kg and fluoroscopy time of 37 (24, 55) minutes. The airway was managed with assisted ventilation in 62% of cases. Interventional case type predominated in this cohort (77%) with Fontan fenestration closure and/or baffle leak closure (n = 195, 34.7%), systemic to pulmonary collateral occlusion (n = 123, 21.9%) and veno-venous collateral occlusion (n = 118, 21%) the most frequent. At least 1 AE occurred in 81 (14%) post-Fontan cases, with 21 (4%) classified as HSAEs including five major events and one death. Major events included atrial tachyarrhythmia requiring CPR and electric cardioversion, asystole requiring CPR, device embolization requiring surgical removal and stent-related symptomatic bronchial compression. One patient developed acute renal failure following fenestration device occlusion and underwent urgent surgical Fontan conversion with fenestration creation and subsequent recovery. There was one death following the development of massive abdominal hemorrhage requiring exploratory laparotomy after collateral vessel tear during attempted percutaneous collateral occlusion.

Differences Between the Cohorts

Apart from expected differences in age and size of the patients, characteristics of the three single-ventricle groups were otherwise similar with respect to gender, frequency of genetic syndrome, and presence of noncardiac diagnoses. More patients underwent multiple procedures in the pre-Fontan (70 patients; 150 cases) and post-Fontan (56 patients; 128 cases) groups than in the pre-BCPA group (16 patients; 33 cases).

Interventions were common in the population overall, but generally differed by stage of single-ventricle palliation. Notably, of the 202 branch PA interventions performed across the entirety of the single-ventricle cohort, PA angioplasty and stent implantation were disproportionately targeted to the *left* PA in 67% and 89% of cases, respectively. AEs and high severity AE rates were higher in the pre-BCPA cohort (AE 20%; HSAE 9%) but also prevalent in the pre-Fontan (AE 11%; HSAE 3%) and post-Fontan (AE 14%; HSAE 4%) cohorts.

Airway Management

A subgroup analysis stratified by mode of airway management evaluated for differences in patient and procedural characteristics and hemodynamic measures, demonstrated in Table 4. Patients in the pre-BCPA cohort that underwent catheterization with assisted ventilation were smaller and more likely to have a noncardiac diagnosis, when compared with those managed with spontaneous ventilation. Implementation of an airway management strategy that utilized spontaneous ventilation was more common in the post-Fontan population, compared with the earlier cohorts. Patients in the post-Fontan cohort that underwent catheterization with spontaneous ventilation were older, larger, and less likely to be undergoing an interventional procedure. In univariate analysis, although there were some statistic differences in reported hemodynamic measures between the subgroups, these differences were subtle and may not be clinically meaningful.

Institutional Variation

Significant practice variation was identified across the eight participating centers in all cohorts. There were differences in age and weight across centers at the pre-Fontan and post-Fontan catheterizations (Figure 2). Procedural characteristics were also variable between centers within each of the populations (Figures 3–6). Across institutions, interventional cases comprised 16–66% of the pre-BCPA cohort, 50–84% of the pre-Fontan cohort and 25–91% of the post-Fontan cohort, including differences in rates of collateral vessel occlusion at all stages. Utilization of an assisted ventilation strategy for airway management was quite variable across institutions. Further, unadjusted-for-risk overall AE rates and HSAE rates varied substantially, ranging from 0% to 50% (0–20%), 0% to 22% (0–5%), and 0% to 19% (0–8%) in the pre-BCPA, pre-Fontan and post-Fontan cohorts, respectively.

Table 4. Patient Characteristics Stratified by Mode of Airway Management and Single-ventricle Group

	Pre-BCPA			Pre-Fontan			Post-Fontan		
	Spontaneous *N* = 64	Assisted *N* = 262	*P*	Spontaneous *N* = 106	Assisted *N* = 465	*P*	Spontaneous *N* = 213	Assisted *N* = 349	*P*
Age (year)	0.4 (0.3, 0.5)	0.4 (0.3, 0.5)	.11	2.6 (2, 3.3)	2.6 (1.8, 3.4)	.69	11.3 (5.5, 18.5)	8.6 (4.8, 14.3)	<.001
Weight (kg)	6 (5.2, 6.8)	5.5 (4.7, 6.3)	.006	12.3 (11, 13.8)	12.2 (10.4, 14.1)	.98	29.6 (18, 55.9)	24.9 (16, 47.4)	.007
Male gender	38 (59%)	136 (52%)	.33	68 (64%)	282 (61%)	.58	124 (58%)	208 (60%)	.79
Presence of genetic syndrome	4 (6%)	26 (10%)	.47	8 (8%)	20 (4%)	.21	12 (6%)	18 (5%)	.85
Presence of noncardiac diagnosis	8 (13%)	75 (29%)	.007	16 (15%)	107 (23%)	.09	62 (29%)	102 (29%)	1.0
Elective admission source			.24			.05			.21
Outpatient	14 (22%)	72 (27%)		28 (26%)	181 (39%)		48 (23%)	95 (27%)	
Same day admit	38 (59%)	124 (47%)		74 (70%)	266 (57%)		146 (69%)	234 (67%)	
Inpatient	12 (19%)	66 (25%)		4 (4%)	18 (4%)		19 (9%)	20 (6%)	
Arterial and venous access	58 (92%)	252 (96%)	.22	104 (98%)	450 (97%)	1.0	205 (96%)	343 (98%)	.19
Interventional case type	27 (42%)	114 (44%)	.89	70 (66%)	325 (70%)	.48	152 (71%)	280 (80%)	.02
Hemodynamic variables									
Cardiac index (l/min/m^2)	3.6 (2.9, 4.2)	3.4 (2.7, 4.6)	.59	3.7 (3.2, 4.2)	4.0 (3.3, 4.9)	.01	3.1 (2.6, 3.7)	3.3 (2.7, 4.0)	.005
Ventricular EDP (mmHg)	7 (6, 10)	8 (6, 10)	.07	8 (5, 10)	8 (6, 10)	.06	10 (7, 12)	8 (6, 10)	<.001
MV_{sat} (%)	49 (46, 52)	51 (45, 58)	.05	61 (57, 65)	65 (61, 69)	<.001	65 (60, 70)	67 (62, 71)	.02
SA_{sat} (%)	73 (70, 78)	75 (70, 79)	.21	83 (80, 86)	84 (81, 87)	.08	90 (85, 92)	89 (84, 93)	.54
PA mean (mmHg)	15 (12, 18)	15 (12, 21)	.52	10 (8, 13)	12 (10, 14)	.001	14 (11, 16)	14 (11, 16)	.50

Values are displayed as N (%) or median (IQR). Assisted, assisted ventilation; EDP, end diastolic pressure; MV_{sat}, mixed venous oxygen saturation; PA, pulmonary artery; Pre-BCPA, pre-bidirectional cavopulmonary anastomosis; SA_{sat}, systemic arterial oxygen saturation; Spontaneous, spontaneous ventilation.

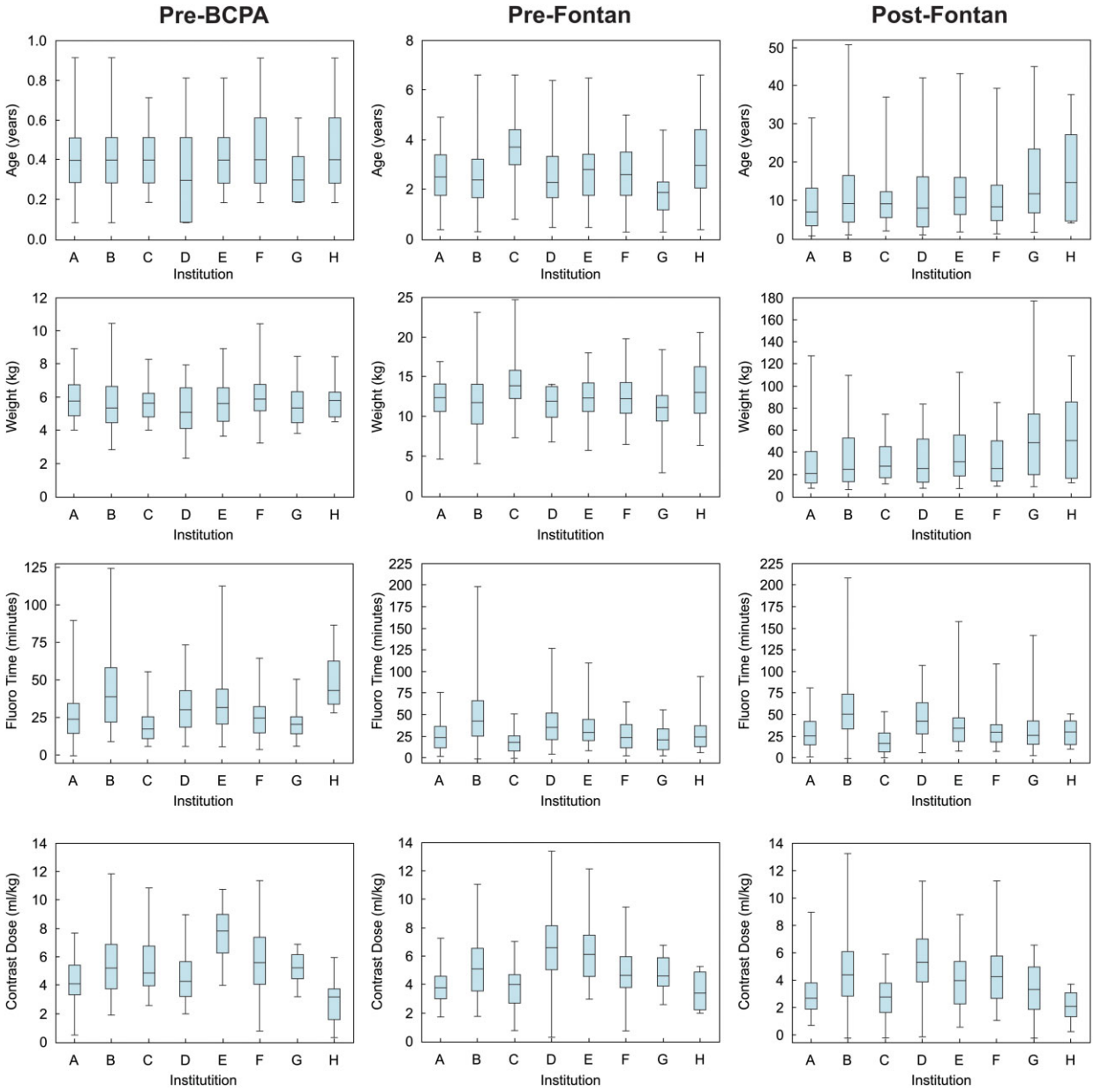

Figure 2. Matrix of box and whisker plots demonstrating institutional variation in patient and procedural characteristics during performance of pre-BCPA, pre-Fontan, and post-Fontan cardiac catheterization procedures.

Discussion

To date, work that has emerged from multicenter collaborations and registries in pediatric interventional cardiology has largely concentrated on defining outcomes of specific procedures (e.g., aortic valvuloplasty) or the development of risk-adjustment tools.[16,18–21] Efforts to explore outcomes in specific populations, such as in adults undergoing catheterization at pediatric centers, have been more limited.[22] In this report, we add to the body of population-specific analyses by focusing on single-ventricle patients undergoing elective catheterization.

Patients and Procedures

Efforts to retrospectively define a population of single-ventricle patients undergoing elective cardiac catheterization, and further stratify the population by stage of palliation, yielded groups with relative homogeneity. The asymmetry in

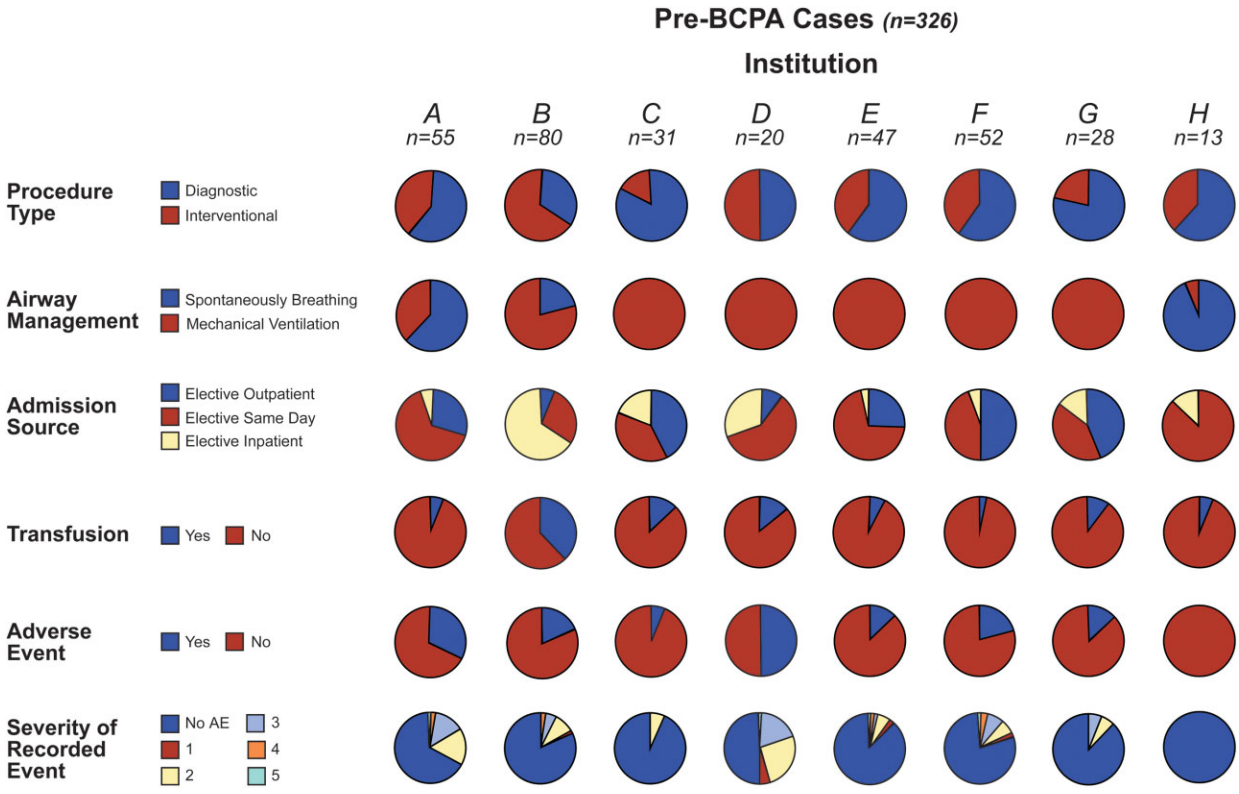

Figure 3. Matrix of pie charts demonstrating institutional variation in key procedural characteristics during performance of pre-bidirectional cavopulmonary anastomosis (pre-BCPA) cardiac catheterization procedures.

group size, found when comparing the pre-BCPA group to the others, probably reflects several factors, including the use of nonelective pre-BCPA catheterization for preoperative assessment (which would exclude the case from this elective cohort), use of pre-BCPA CMR in lieu of catheterization, inclusion of patients not undergoing preoperative evaluation (e.g., a pre-Fontan patient with extensive collateralization previously deemed not to be a Fontan candidate), and the short duration of the first interstage. Moreover, compared with the pre- and post-Fontan groups, patients in the pre-BCPA group were less likely to undergo multiple catheterizations. Without access to the total denominator of single-ventricle patients undergoing the BCPA or Fontan procedure at participating centers, we were unable to determine the relative use of elective catheterization vs. alternate tools (e.g., CMR) in the preoperative assessment of single-ventricle patients.

The goals of elective single-ventricle catheterization vary widely, with the greatest likelihood of a noninterventional invasive assessment occurring in the pre-BCPA group. In all groups, the most common type of intervention was occlusion of a collateral vessel. The high rate of systemic to pulmonary collateral occlusion is an important finding given the relative paucity of rigorous outcome data associated with collateral occlusion.[23–26] Practice variation with regards to systemic to pulmonary collateral occlusion has been investigated by others with wide variability in practice again demonstrated in this cohort.[26] More work is necessary to understand the short- and long-term clinical impact of this commonly performed intervention in order to develop evidence-based recommendations surrounding therapy.[23] Also noteworthy is the finding that the majority of PA interventions targeted the left PA (within each cohort, albeit more pronounced following BCPA). It is likely that this reflects several factors, including those influenced by anatomic diagnosis (e.g., reconstructed neo-aorta in hypoplastic left heart syndrome), surgical technique (e.g., right-sided shunt placement preferentially supplying the right PA) and surgical planning (e.g., proximity of right PA vs. left PA lesion to anticipated surgical field during next stage palliation).[10,27,28] Disproportionate left PA interventions would not have been predicted based upon the early results from the PHN

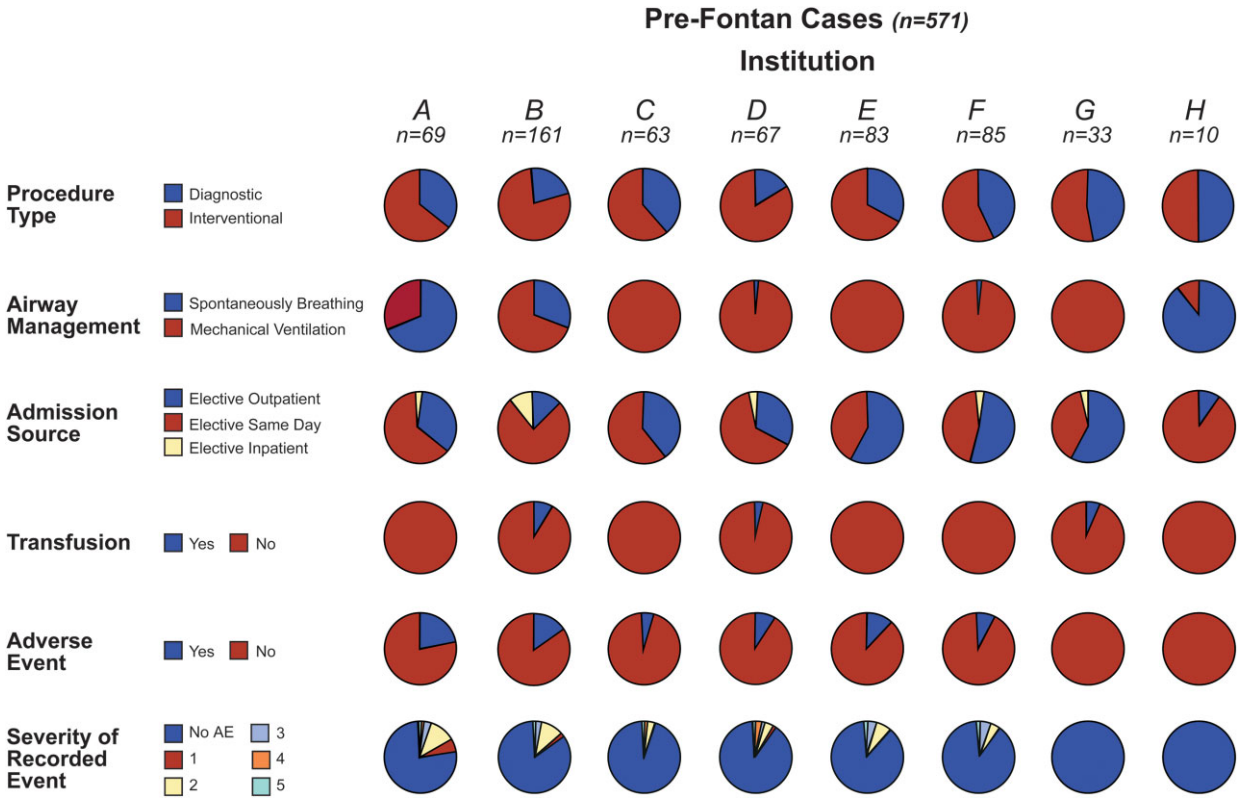

Figure 4. Matrix of pie charts demonstrating institutional variation in key procedural characteristics during performance of pre-Fontan cardiac catheterization procedures.

Single-ventricle Reconstruction trial, which demonstrated a greater burden of right PA stenosis at the pre-BCPA catheterization.[29]

Use of spontaneous ventilation was most common in the post-Fontan cohort, likely reflecting the increased age, size, and overall stability of this more mature cohort. Although the impact of assisted ventilation on acute postoperative hemodynamics has been assessed previously in stage II and Fontan palliated patients,[30–32] no prior reports have examined the relation of airway management to invasive hemodynamics in the setting of elective catheterization. In this setting, where a primary procedural goal is to evaluate candidacy for next stage surgical palliation, the accuracy of hemodynamic data are critical. While a rigorous evaluation of the impact of airway management on invasive hemodynamics would require a prospective randomized approach, this nonrandomized large multicenter sample of single-ventricle patients can provide some insight. Importantly, we found few significant differences in hemodynamic measures—and none clinically large—between subgroups when stratified by mode of airway management and evaluated at the *population* level. This analysis does not exclude the possibility that the impact of airway management strategy on hemodynamics may be substantial in a given patient. Given the large volume of cases, relative homogeneity of the population, considerable practice variation present and potential implications of airway management on hemodynamic measures (and possible AE rate), the single-ventricle population may be an ideal group to target for future multicenter prospective randomized study of spontaneous vs. assisted ventilation during elective catheterization.

AEs

Overall unadjusted AE and HSAE rates ranged from 11% to 20% and 3% to 9%, respectively, in the three single-ventricle groups investigated in this report. To put these data into context, previously published AE and (HSAE) rates from the C3PO registry for specific procedures include: ASD closure, 12% (5%); branch PA intervention, 22% (10%) and balloon pulmonary valvuloplasty, 9% (3%).[19,33,34] The overall AE and (HSAE) rates in the 2061 adult cases and 11 422 pediatric cases in the C3PO dataset were 10% (4%) and 13%

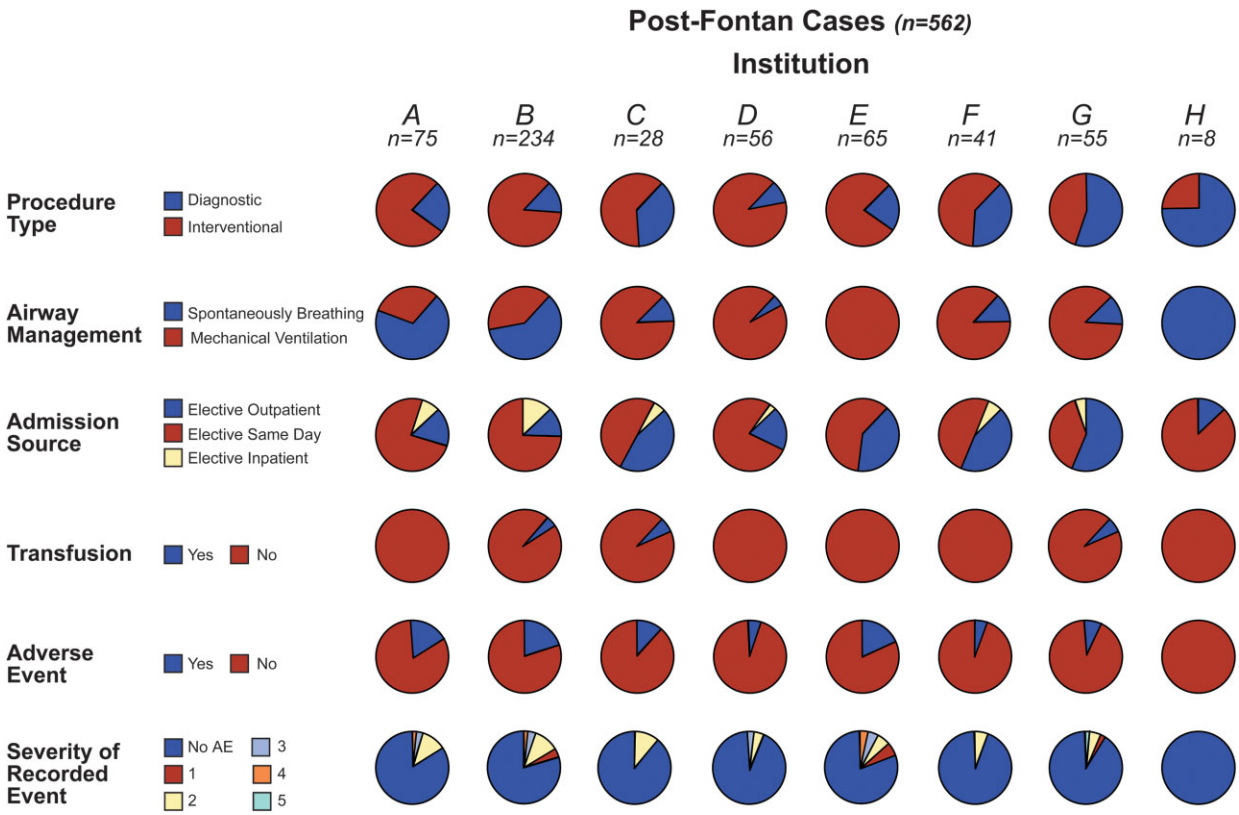

Figure 5. Matrix of pie charts demonstrating institutional variation in key procedural characteristics during performance of post-Fontan cardiac catheterization procedures.

(5%), respectively.[22] In this report, we did not further explore potential factors associated with AEs, nor develop risk-adjustment tools specific to this population. Not surprisingly, the rates of AEs (20%), HSAEs (9%), and PRBC transfusions (15%) were highest in the pre-BCPA group. The finding that this group is more vulnerable is supported by prior work from the C3PO collaboration, which demonstrated that age (less than 1 year) and reduced SA_{sat} are independent risk factors for HSAEs.[16] Additional hemodynamic factors associated with AEs, including those specific for the single-ventricle population, have been defined.[16] Low weight, a common finding in the pre-BCPA group, has also been found to be an independent risk factor for AEs.[35,36] Fortunately, HSAEs were uncommon overall and only 1 procedural death (0.07%) occurred in this large cohort, which is commensurate with the overall death rate in the entire C3PO cohort (0.29%).[15]

Institutional Variation

A wide degree of practice variation was identified in this report, including variability in patient and procedural characteristics, interventions, and outcomes. An example characteristic with broad variation found in this analysis was age at pre-Fontan catheterization, with at least two institutions (centers C and G) demonstrating no overlap in interquartile range of their respective populations, despite a median age of less than 4 years in both groups. This variation likely reflects surgeon/center preference related to patient factors (e.g., age and weight) and surgical techniques (e.g., lateral tunnel vs. extracardiac conduit Fontan completion). Airway management strategy also varied substantially, with some centers exclusively employing assisted ventilation across the entirety of the population (centers C-G for the pre-BCPA and pre-Fontan patients), while others (centers A and B) employed a mix of assisted and spontaneous ventilation strategies, probably allowing patient factors to influence that decision. The finding that low weight and presence of a noncardiac diagnosis were associated with the use of assisted ventilation in the pre-BCPA cohort supports the hypothesis that patient factors influenced airway management decision making.

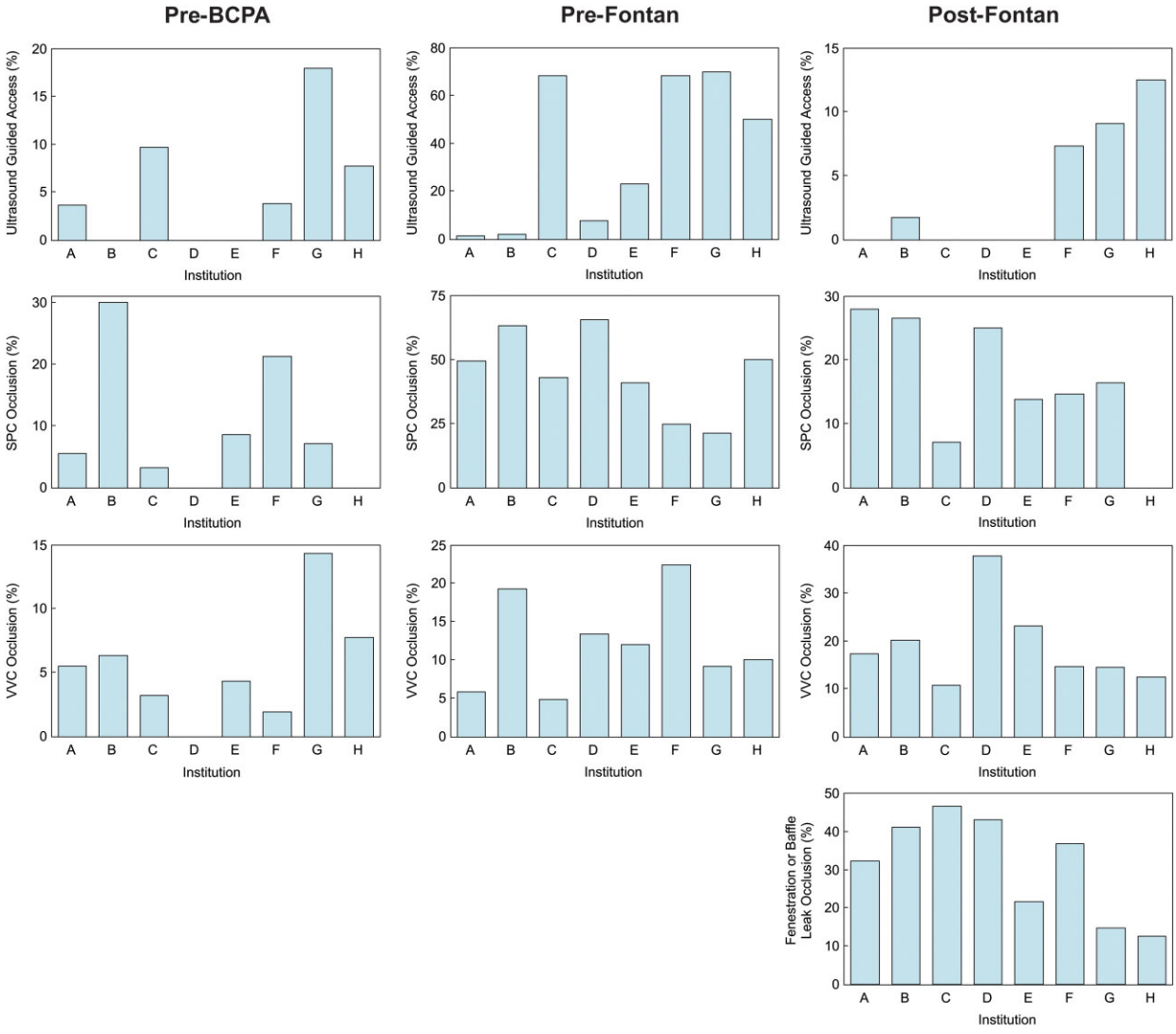

Figure 6. Matrix of bar graphs demonstrating institutional variation in key interventional procedures performed during pre-bidirectional cavopulmonary anastomosis (pre-BCPA), pre-Fontan, and post-Fontan cardiac catheterization procedures.

Wide variation in care delivered in the catheterization laboratory is consistent with prior studies of single-ventricle patients outside of this setting, which have demonstrated significant variation in perioperative, nutrition and interstage management.[7,9,37] Variation in catheterization outcomes, as measured by AE rate in the present report, has been demonstrated previously in this collaborative.[16] Inter-center variation in AE rates was significant in the single-ventricle population, present within each of the three single-ventricle groups studied, and bore no obvious relation to case volume. However, a more detailed analysis of factors associated with AE rate was not feasible in this study, because of limitations inherent to the small sample sizes encountered once the cohort was divided into three subgroups and then examined at the center level. To illustrate this problem further, it is worth noting that the two centers at the extreme ends of the AE rate spectrum in the pre-BCPA group (0% and 50%, respectively) represented the two lowest volume centers. The potential for one or two cases with an AE to skew a center's overall AE rate substantially at low volume centers limits the ability to draw broad conclusions regarding AE rate variability from this dataset. AE rate variability appeared to extend to both low- and high-severity AEs. Variability in HSAEs is of particular concern, both because of their clinical impact and the reliability of their capture, suggesting that variation in AE rate is not a manifestation of differential reporting.[15,16] To the

extent that AE rate directly relates to clinical status and patient outcomes—unlike, for example, fluoroscopy time which is a marker of radiation exposure and potential future risk, but is not typically related to short-term adverse outcomes—it would serve as an ideal metric for future QI work in this domain. The challenge is to ensure unbiased and near-complete capture of AEs, especially HSAEs, as these data are not accrued and recorded independent of provider input. Even in the dataset presented herein, which underwent independent auditing of a 10% data sample (which demonstrated 91% capture of HSAEs), variation in AE reporting is an important potential source of confounding.

The analysis of practice variation included herein is a necessary first step to engaging in QI efforts aimed at identifying "best practices" and reducing variation in catheterization-related care processes. Pediatric cardiologists and intensivists managing stage I single-ventricle patients in the perioperative and outpatient settings have engaged in QI efforts,[38] which have already resulted in reduced variation in interstage growth parameters.[39] Early successes in this population should motivate interventional cardiologists to begin QI work aimed at identifying "best practices" associated with the acquisition of highly accurate clinical data, improved clinic efficiency, and reduced AE rates. However, to the extent that much of the practice variation identified herein reflects institutional policy (e.g., aggressive vs. conservative approach to AP collateral occlusion, or timing of stage II surgery), and not decisions taken in isolation within the catheterization laboratory, these QI efforts must be undertaken at the broader programmatic level. Single-ventricle QI work in the catheterization laboratory must be but one domain of a linked multidomain and probably multicenter effort to improve outcomes for these high-risk infants, children and young adults. The next phase of the C3PO collaborative, C3PO-QI (comprised of a larger number of participating centers), along with the larger American College of Cardiology IMPACT (Improving Pediatric and Adult Congenital Treatments) registry, ought both serve as working groups from which the catheterization-related process improvement can be managed and outcomes tracked. To date, early efforts undertaken by these groups are focused on radiation reduction.[40,41] Improvement efforts focused on the single-ventricle population that include the catheterization domain could potentially be spearheaded by an umbrella organization with interest specific to this group, such as the National Pediatric Cardiology Quality Improvement Collaborative.

There are a number of important limitations to this study. First, despite the national multicenter design, participation was limited to eight large cardiac programs associated with a children's hospital and considered a referral base for congenital heart surgery. Second, despite efforts to narrow case selection, the retrospective nature of this work made identification of entirely homogenous subgroups difficult. Third, there are important biases introduced by institutional practice and case selection. For example, centers that perform CMR for routine pre-BCPA evaluation in low-risk candidates will necessarily refer only their high-risk candidates to catheterization, thereby altering their population's risk profile. Fourth, despite the large size of the cohort overall, individual center sample sizes were small, limiting the ability to perform rigorous tests of comparison, and magnifying the impact of individual cases and outcomes. Fifth, the comparison of groups by mode of airway management incorporates a number of biases, including those at the patient, operator and institutional levels, which serve to limit the interpretation and generalizability of these data. Sixth, the difficulties inherent to accurate event capture and data entry, both specific to the C3PO registry and to multi-institutional registries in general, are substantial and have been detailed in prior reports from this collaborative.[15] Lastly, in order to utilize an audited dataset for this study, we analyzed data that are now 5 years old, and it is likely that some shifts in practice have occurred since data collection, although we believe this analysis is still highly applicable to current practice.

In conclusion, we describe current approaches to, and practice variation within, single-ventricle patients undergoing elective cardiac catheterization at all stages of palliation. This description ought to serve as a baseline for future QI efforts in this population, aimed at reducing variation in care delivery, identifying "best practices" and ultimately improving patient outcomes.

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