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Arteriovenous fistula creation for hypoxia after single ventricle palliation: A single-institution experience and literature review

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Abstract

Background: Hypoxia is a common and sometimes severe morbidity of single ventricle congenital heart disease (CHD). Creation of an arteriovenous fistula (AVF) is occasionally performed for patients after superior or total cavopulmonary connection (SCPC or TCPC) in an attempt to improve oxygen saturations. Despite previous reports, AVF creation is a rare palliation with inadequately defined benefits and risks. We sought to determine changes in peripheral oxygen saturation (SpO_2) and risk of adverse event after AVF creation in children with single ventricle CHD at our institution.

Methods: We conducted a retrospective chart review of patients with a history of single ventricle palliation and history of surgical AVF creation who were seen at our tertiary care center from 1996 to 2017.

Results: A total of seven patients were included in our study. SpO_2 for the overall cohort did not significantly increase after AVF creation (pre-AVF 79.1 ± 6.9%, post-AVF 82.7 ± 6.0% [P = .23]). SpO_2 trended up for large shunts (>5 mm) (pre-AVF 75.0 ± 7.6%, post-AVF 84.0 ± 5.3% [P = .25]). SpO_2 did not improve for small shunts (≤5 mm) (pre-AVF 82.3 ± 6.5%, post-AVF 81.0 ± 8.5% [P = .50]). The 12-month overall and transplant-free survival were 85.7% and 71.4%, respectively. Freedom from AVF-related complication (cephalic edema, thrombotic occlusion) was 51.4% at 12 months.

Conclusion: Palliative AVF creation for patients with single ventricle CHD and hypoxia does not universally improve SpO_2 and is prone to early complications. Despite a lack of durable benefit and known risks, AVF creation remains a reasonable palliation for a subset of patients after SCPC who are not candidates for TCPC, or potentially as a bridge to heart transplantation.

KEYWORDS

arteriovenous fistula, cavopulmonary connection, hepatic factor, hypoxia, pulmonary arteriovenous malformations, single ventricle

Abbreviations: AVF, arteriovenous fistula; BDG, bidirectional Glenn; BT, Blalock-Taussig; CHD, congenital heart disease; Hgb, hemoglobin; OHT, orthotopic heart transplant; PAVMs, pulmonary arteriovenous malformations; PBF, pulmonary blood flow; SCPC, superior cavopulmonary connection; SpO_2, peripheral oxygen saturation; TCPC, total cavopulmonary connection.

1 | INTRODUCTION

Due to advances in medical and surgical care, the majority of individuals with single ventricle congenital heart disease (CHD) are surviving to adulthood.[1-4] Despite improved survival, patients are at risk for multiple long-term complications.[3] A common morbidity, which occasionally becomes severe and debilitating, is chronic hypoxia.

Hypoxia is universal after superior cavopulmonary connection (SCPC), or bidirectional Glenn (BDG), due to obligate mixing of hepatic vein and/or inferior vena cava blood flow into the systemic circulation, but the degree of hypoxia is variable. Pulmonary arteriovenous malformations (PAVMs) frequently develop after SCPC and may worsen hypoxia over time. PAVMs likely form in patients after SCPC from lack of exposure of the pulmonary circulation to a putative hepatic factor present in hepatic venous blood flow. This hypothesis is supported by the finding that patients with total cavopulmonary connection (TCPC), or modified Fontan operation, and unilateral hepatic vein streaming (typically in the setting of heterotaxy) develop worsening of PAVMs in the contralateral lung (i.e., the lung lacking hepatic factor perfusion) and increasing hypoxia.

Arteriovenous fistula (AVF) creation is occasionally performed in an attempt to improve oxygen saturations for patients after SCPC or for patients after TCPC with hypoxia due to PAVMs. The rationale for AVF creation is to increase effective pulmonary blood flow and increase hepatic factor circulation to the pulmonary vasculature. A few studies previously reported their experiences with AVFs in patients with single ventricle CHD and hypoxia. Results are mixed regarding a clear benefit in oxygen saturation. Postoperative complications, including edema, headache, and spontaneous fistula occlusion, are also variable with rates ranging from 0% to 48%.[5-12] Thus, AVF creation continues to have inadequately defined benefits and risks. The primary aim of this study was to determine change in peripheral oxygen saturation (SpO_2) after AVF creation in patients with single ventricle CHD and prior SCPC or TCPC palliation seen at our institution. Secondarily, we sought to assess the risk of adverse events after AVF creation. Finally, we reviewed the available published literature on AVF creation in this patient population.

2 | METHODS

2.1 | Patient Selection

We conducted a retrospective chart review of patients seen at the Children's Hospital of Wisconsin from 1996 to 2017. Inclusion criteria included a history of single ventricle CHD, surgical palliation with SCPC (classic Glenn or bidirectional Glenn palliation) or TCPC, and a history of surgical AVF creation. Patients were not excluded if SCPC, TCPC, or AVF creation were performed at an outside institution prior to establishing medical care at Children's Hospital of Wisconsin. This study was approved by the local institutional review board (IRB).

2.2 | Oxygen saturation

Peripheral oxygen saturation and hemoglobin (Hgb) levels were recorded from the medical record. For inpatient records, we reported the average SpO_2 value (as calculated by the electronic medical record for the average of SpO_2 values recorded for that 24-hour period) when available. If an average SpO_2 value was not available, we reported the SpO_2 recorded in the vital signs of the daily progress note. Baseline (pre-AVF) SpO_2 and Hgb were determined by the last recorded values prior to entering the operating room for AVF creation. Final (post-AVF) SpO_2 and Hgb were determined by the last recorded values prior to takedown, spontaneous occlusion, death, or at the end of the follow-up period. Percent SpO_2 change was calculated using the pre-AVF SpO_2 and all subsequently recorded SpO_2 values. To minimize confounding Hgb levels due to acute variability (bleeding and transfusion), only pre-AVF and post-AVF Hgb levels were recorded.

2.3 | Survival analyses

Data regarding patient diagnosis, clinical variables, details of AVF surgery, and postoperative outcomes were obtained. To investigate survival following AVF creation, we analyzed overall survival, transplant-free survival, and freedom from AVF-related complication. Freedom from AVF-related complication was defined as survival without AVF takedown, AVF re-intervention (ie, thrombectomy), spontaneous AVF occlusion, or symptomatic side effects (ie, chest edema). Orthotopic heart transplantation (OHT) was excluded as an AVF-related complication, and patients were censored from this analysis at time of OHT.

2.4 | Statistical analysis

Cohort data are expressed as mean ± standard deviation (SD). Due to the small and heterogeneous composition of this cohort, a matched control group was not created for comparison. We performed a Wilcoxon matched pairs signed rank test to compare the pre- and post-AVF variables (SpO_2 and Hgb) for the entire cohort and subgroups. We generated Kaplan-Meier survival curves to report overall survival, transplant-free survival, and freedom from AVF-related complication. All analyses were performed using GraphPad Prism 8 (GraphPad Software, San Diego, CA). Statistical significance was accepted at the level of $P < .05$.

3 | RESULTS

A total of seven patients with a history of single ventricle palliation underwent AVF creation during the study period (Table 1). All patients underwent AVF creation due to hypoxia and diagnosed or suspected PAVMs. Of the seven patients, six had a form of SCPC, and one had a TCPC. Four of the patients with SCPC had standard bidirectional connection with end-to-side anastomosis of the superior

TABLE 1 Patient demographics, clinical data, and fistula outcomes

Patient	Gender	Primary Diagnosis	Heterotaxy	Physiology	AVF type	Age at AVF (years)	Pre-AVF SpO_2 (%)	Post-AVF SpO_2 (%)	AVF duration (years)	AVF outcome
1	F	DILV	N	Bidirectional SCPC	Subclavian	3.3	82	84	19.9	Patent
2	M	DORV	N	Unidirectional SCPC	Axillary (9 mm side-to-side)	26.7	76	82	4.2	Takedown (cephalic edema)
3	M	HLHS	N	Unidirectional SCPC	Brachial-basilic (7 mm side-to-side)	8.8	82	90	1.1	Family relocated
4	F	Unbalanced AVSD	Y	Bidirectional SCPC	Brachial-basilic (15 mm side-to-side)	7.1	67	80	0.3	Died (sepsis)
5	M	Unbalanced AVSD	Y	Bidirectional SCPC	Carotid-jugular (5 mm PTFE shunt)	19.6	89	90	0.9	Thrombotic occlusion
6	M	DILV	Y	Bidirectional SCPC	Brachial-basilic (4 mm PTFE shunt)	6.7	76	73	0.1	Takedown (OHT)
7	M	Unbalanced AVSD	Y	TCPC	Brachial-basilic (4 mm PTFE shunt)	12.6	82	80	1.3	Takedown (OHT)

Abbreviations: AVF, arteriovenous fistula; AVSD, atrioventricular septal defect; DILV, double inlet left ventricle; DORV, double outlet right ventricle; HLHS, hypoplastic left heart syndrome; OHT, orthotopic heart transplant; PTFE, polytetrafluoroethylene; SCPC, superior cavopulmonary connection (Glenn); SpO_2, peripheral oxygen saturation; TCPC, total cavopulmonary connection (Fontan).

vena cava to the right pulmonary artery. Two of the patients with SCPC had a "classic Glenn" connection (end-to-end anastomosis of the superior vena cava to the right pulmonary artery). Patient #2 underwent classic Glenn palliation at age 6 years due to a failed pulmonary artery band on the left pulmonary artery and subsequently acquired left pulmonary vascular disease. Patient #3 had BDG palliation at 4 months old but was later revised at 11 months old to a "classic Glenn" connection due to left pulmonary artery thrombosis and failed attempts at left-sided Blalock-Taussig (BT) shunt placement.

All patients with SCPC who underwent AVF creation were not candidates for TCPC due to severe co-morbidities (patient #1), pulmonary vascular disease (patients #2 and 3), history of failed TCPC (patients #4 and 6), and complex anatomy (patient #5). Patient #5 did not initially have TCPC due to his underlying anatomy (dextrocardia, midline hepatic veins, and interrupted inferior vena cava) and mild hypoxia. He later underwent TCPC after spontaneous occlusion of his AVF and continued worsening hypoxia. One patient (patient #7) with TCPC had history of an extracardiac non-fenestrated Fontan procedure, and subsequently had AVF creation for hypoxia and angiographic evidence of unilateral streaming of hepatic vein flow to one lung and PAVMs in the contralateral lung.

Of the seven patients, four (patients #1-4) had AVF creation during the "early" era (1996-2014) and three (patients #5-7) had AVF creation during the "recent" era (2015-2017). With the exception of patient #1, whose fistula size is unknown, the "early" era utilized a large AVF size (defined as >5 mm), whereas the "recent" era utilized small AVF size (≤5 mm).

3.1 | AVF creation

The type and location of AVF varied based on underlying anatomy and surgeon preference (Table 1). One patient (patient #1) had a subclavian artery-to-vein AVF creation at an outside institution prior to transitioning her medical care to our institution, and operative reports were unavailable to determine the details of the technique and size of the AVF. The remaining six AVFs were all performed at our institution, including four that involved the brachial artery and basilic vein, one that involved the axillary artery and vein, and one that involved the carotid artery and jugular vein. With the exception of patient #5 (carotid-jugular fistula), the vein was ligated distally after creation of the AVF. The size of the AVF varied from 4 to 15 mm, and four utilized polytetrafluoroethylene (PTFE) shunts for the artery-vein connection. After AVF creation, six patients were only prescribed aspirin for long-term anti-platelet therapy. One patient (patient #4) was hospitalized at the time of AVF creation and was maintained on a heparin drip.

Of the seven patients with AVF creation, one (patient #4) was critically ill with severe hypoxia. She had acute thrombosis of her AVF on postoperative day #5 and returned to the operating room for thrombectomy. She remained critically ill after thrombectomy and ultimately died prior to hospital discharge on postoperative day #98 due to sepsis, likely unrelated to AVF creation. Of the remaining six patients that were discharged home after AVF creation, one

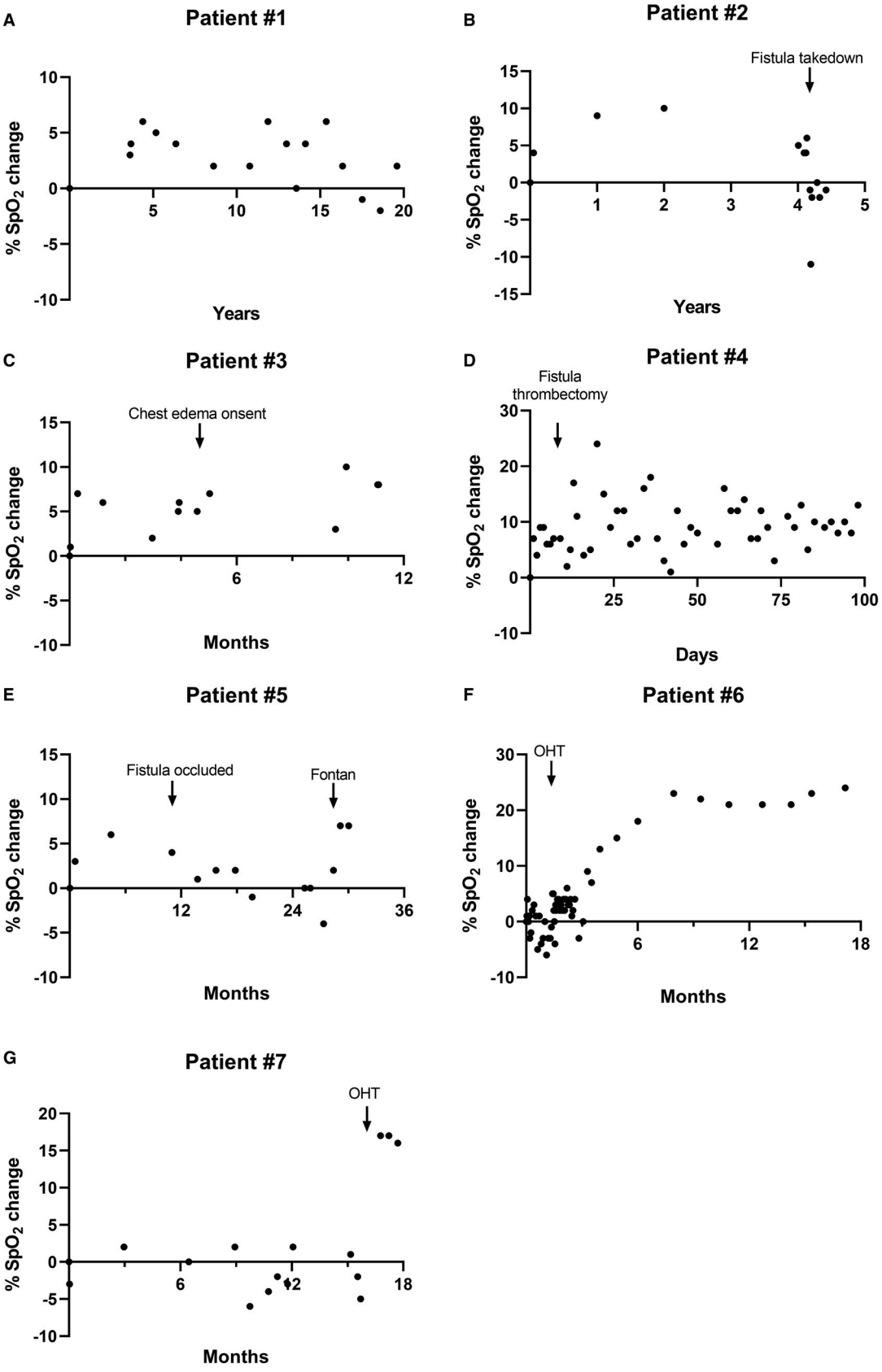

FIGURE 1 Patient-specific oxygen saturation trajectories after AVF creation. A-G, SpO_2 changes for each individual patient after AVF creation. Pre-AVF SpO_2 represents patient's baseline SpO_2 prior to AVF creation at time 0. B-E, AVF-related complications for patients #2-5 are indicated on their respective graphs. AVF, arteriovenous fistula; SpO_2, peripheral oxygen saturation

AVF remained patent at 19.9 years follow-up without AVF-related complication. Two patients underwent AVF as a successful bridge to OHT at 0.1 and 1.3 years follow-up. One patient (patient #2) had takedown of his AVF at 4.2 years due to intractable cephalic edema and headaches. One patient (patient #5) had spontaneous occlusion at 0.9 years. Another patient (patient #3) developed worsening chest edema 4.6 months after AVF creation but was ultimately lost to follow-up at 1.1 years with a patent AVF.

3.2 | Oxygen saturation

Pre- and post-AVF SpO_2 for each individual patient are listed in Table 1. Additionally, patient-specific SpO_2 trajectories after AVF creation are displayed in Figure 1. Notably, one patient (patient #1) was seen for consultation prior to AVF creation at an outside institution but did not re-establish follow-up at our institution until 3.6 years post-AVF.

There was no significant difference in pre- and post-AVF SpO_2 for the overall cohort (pre-AVF 79.1 ± 6.9%, post-AVF 82.7 ± 6.0% [P = .23]). When comparing change in SpO_2 based on AVF size, there was a non-significant trend for improved SpO_2 for large AVF (defined as >5 mm) (pre-AVF 75.0 ± 7.6%, post-AVF 84.0 ± 5.3% [P = .25]) compared to small AVF (≤5 mm) (pre-AVF 82.3 ± 6.5%, post-AVF 81.0 ± 8.5% [P = .50]). Similarly, patients in the "early" era (1996-2014) trended toward improved SpO_2 (pre-AVF 76.8 ± 7.1%, post-AVF 84.0 ± 4.3% [P = .12]).

There was no significant difference in pre- and post-AVF Hgb for the overall cohort (pre-AVF Hgb 17.5 ± 1.5 g/dL, post-AVF Hgb 16.1 ± 2.6 g/dL [P = .09]). One patient (patient #1) was excluded from Hgb comparison because we lacked report of a pre-AVF Hgb contemporaneous to AVF creation.

3.3 | Survival

Kaplan-Meier survival curves after AVF creation are shown in Figure 2. The 12-month overall survival, transplant-free survival, and freedom from AVF-related complication were 85.7%, 71.4%, and 51.4%, respectively. The median overall survival, transplant-free survival, and freedom from AVF-related complication were 53.3, 53.3, and 49 months, respectively. AVF-related complications occurred in 3/4 (75%) patients in the "early" era and 1/3 (33%) patients in the "recent" era. AVF outcomes and complications are reported in Table 1 and Figure 1, respectively.

4 | DISCUSSION

Our study summarizes our institutional experience with palliative AVF creation for patients with single ventricle CHD who have progressive hypoxia after SCPC or TCPC. We found that changes in SpO_2 after AVF creation were highly variable and, in general, did not significantly improve during follow-up. Overall survival after AVF creation, when performed either as long-term palliation or bridge-to-OHT, was good; however, AVF-related complications, such as edema and AVF thrombosis, were not uncommon. These are important findings that can be used for comparison to other published case series.

AVF creation variably impacted oxygen saturation in our cohort. A larger fistula (>5 mm) appeared to increase SpO_2 (pre-AVF 75.0 ± 7.6%, post-AVF 84.0 ± 5.3% [P = .25]) but the power to detect a statistically significant difference was limited by patient number (n = 3). Our equivocal findings mirror the mixed results of previous studies (Table 2). Early small studies consistently reported improved PaO_2, symptoms, and SpO_2 after AVF, though no statistical analyses were included.[5-8] A more recent study by Hickey et al reported that SpO_2 increased from 74% to 83% after AVF creation in a mixed cohort of patients after SCPC and TCPC (P < .01; n = 21).[10] Conversely, Quiñonez et al reported no statistically significant improvement in clinical symptoms, oxygen saturation, or hemoglobin in patients after SCPC and TCPC (n = 11).[11] Most recently, Chanana et al reported a statistically significant increase in SpO_2 (pre: 71 ± 10%, follow-up: 81 ± 5%; P < .05) in 23 patients after SCPC only, which remains the largest single cohort to date.[12]

Although studies show mixed results with regards to improvement in SpO_2 following AVF creation, examining the data more closely suggests that there may be a subset of patients following SCPC, but not TCPC, who may benefit from AVF. McElhinney et al found that PAVMs angiographically resolved in 4/5 surviving

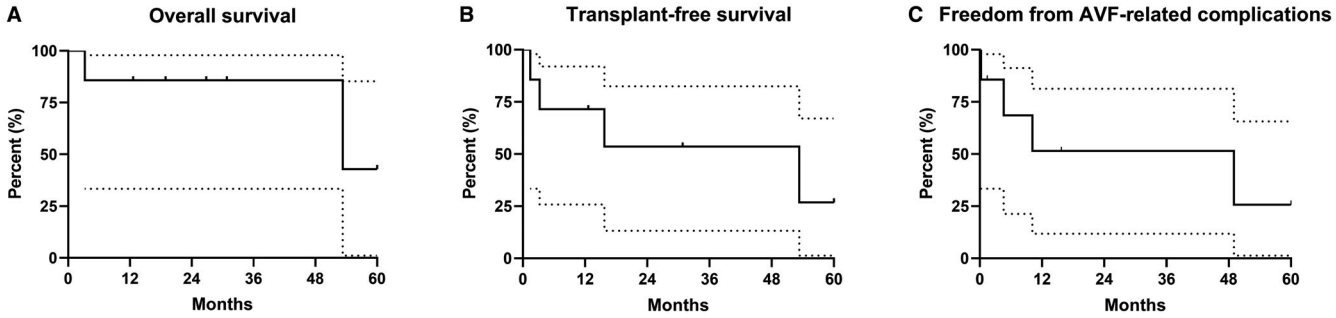

FIGURE 2 Kaplan-Meier survival curves after palliative AVF creation. A, Overall survival, B, transplant-free survival, and C, freedom from AVF-related complication after palliative AVF creation. AVF-related complications include AVF takedown, AVF re-intervention, spontaneous AVF occlusion, and symptomatic side effects. Orthotopic heart transplantation (OHT) was excluded as an AVF-related complication and patients were censored at time of OHT. Solid line represents mean survival and dashed lines represent 95% confidence intervals

TABLE 2 Review of previously published AVF cohorts

Study	Number of patients (SCPC/TCPC)	Age at AVF (years)	Fistula type (size)	Distal vein ligated	Follow-up (years)	Results	Complications
Glenn et al[5]	1 (1/0)	13	Axillary (8 mm)	N	1.3	Pre: PaO_2: 57 mmHg	1/1 (100%)
						Post: PaO_2: 110 mmHg	Immediate postoperative hematoma
Mitchell et al[6]	5 (5/0)	Mean 17.1	Brachial-Basilic (10 mm)	Y	Mean 2.6	Subjective improvement in symptoms (4/5; 80%)	1/5 (20%)
		Range 7.2-27.5			Range 1-4.1		UE claudication & coldness
Kopf et al[7]	8 (8/0)	Not reported	Axillary (not reported)	Unknown	Not reported	SpO_2	2/8 (25%)
						Pre: 70.8 ± 2.9%,	UE edema
						Post: 82.5 ± 2.1%	
Magee et al[8]	10 (10/0)	12.9 ± 5.6	Axillary (6-7 mm)	Y	Mean 7.4	SpO_2	3/10 (30%)
		Range 4-20			Range 0.1-15.5	Pre: 80 ± 2%,	UE edema (n = 1; re-intervention at 19 days to ligate distal vein)
						Post: 85 ± 2%	Spontaneous occlusion (n = 2)
						Follow-up: 84 ± 3%	
						Angiographic resolution of PAVMs in 7/8 (F/U > 1 year)	
McElhinney et al[9]	11 (8/3)	Median 14.0	Brachial-Basilic (4 mm)	N	Median 4.3	PAVMs angiographically resolved in 4/5 surviving patients with unidirectional SCPC flow	2/11 (18%)
		Range 5-32			Range 1.5-7.1	PAVMs did not resolve (0/3) in TCPC patients	Spontaneous occlusion (n = 2)
Hickey et al[10]	21 (16/5)	Median 13.5	Axillary (6-7 mm)	Y	Mean 11	Overall SpO_2	2/21 (10%)
		Range 0.4-36				Pre: 74% (63-90%)	Spontaneous occlusion (n = 1); unspecified surgical takedown (n = 1)
						Post: 81% (75-95%)*	
						Most recent: 83% (75-90%)*	
Quiñonez et al[11]	11 (10/1)	22.6 ± 11.4	Axillary (median 5 mm, range 3-6 mm)	N	Median 2.85	SpO_2	4/11 (36%)
		Range 7.9-41.8			Range 0.01-7.22	Pre: 84% (80-86%)	UE edema (n = 2);
						Follow-up: 82% (76-88%)^{NS}	UE numbness (n = 1);
							cephalic edema & headaches (n = 1)
Chanana et al[12]	23 (23/0)	Median 4.5	Axillary (median 10 mm, range 6-15 mm)	Y	Median 3.92	Overall SpO_2	11/23 (48%)
		Range 0.3-23.2			Range 0.01-14.5	Pre: 71 ± 10%, F/U: 81 ± 5%*	Decreased UE pulse (n = 4); chest edema (n = 1); increased EDP prompting takedown (n = 2); spontaneous occlusion (n = 4)

Complete data are reported as mean ± SD when available. Incomplete data are reported as mean, median, and/or range.

Abbreviations: AVF, arteriovenous fistula; EDP, end-diastolic pressure; NS, not statistically significant; PaO_2, arterial partial pressure of oxygen; PAVMs, pulmonary arteriovenous malformations; SCPC, superior cavopulmonary connection; SpO_2, peripheral oxygen saturation; TCPC, total cavopulmonary connection; UE, upper extremity.

*$P < .05$.

patients with unidirectional SCPC physiology, but PAVMs did not resolve in patients (0/3) with TCPC physiology and unilateral hepatic vein streaming (with contralateral PAVMs). For those with resolved PAVMs, SpO_2 increased from 76% to 92%, whereas SpO_2 modestly increased from 80% to 84% in those patients with persistent PAVMs (ie, TCPC physiology).

Chanana et al subdivided their cohort into three groups: (a) urgent AVF performed <1 month after SCPC palliation due to severe cyanosis and inadequate pulmonary blood flow (n = 6); (b) elective AVF in patients not considered TCPC candidates (n = 10); and (c) elective AVF in patients considered potential TCPC candidates (n = 7). With re-analysis, SpO_2 statistically increased in group 1 (pre: 57 ± 5%, follow-up: 84 ± 3%), whereas there was no statistical difference in groups 2 and 3, indicating that their positive results were driven solely by patients in group 1 with inadequate pulmonary blood flow.

We speculate that to prevent PAVMs, or at least limit clinically significant PAVMs, hepatic factor must be introduced early to mitigate changes to the pulmonary vasculature. For patients after SCPC who are not candidates for TCPC or OHT, we advocate for early creation of a small AVF before the onset of severe hypoxia. This strategy may allow a small "dose" of hepatic factor to perfuse the lungs and prevent development of clinically significant PAVMs, as well as decrease risk of adverse side effects (ie, cephalic edema and venous hypertension). The apparent lack of benefit of AVF in patients with TCPC physiology and maldistribution of hepatic vein blood flow may be that hepatic factor is metabolized in the ipsilateral lung and "unavailable" to recirculate to the contralateral lung via AVF flow. In contrast, hepatic factor may be "available" for delivery to the lungs via an AVF in patients with SCPC physiology because hepatic venous effluent is transported directly to the heart and systemic arterial vasculature without first traversing the pulmonary capillary bed.

Our reported AVF-related complication rate of (4/7, 57%) is similar to previously published case series where the prevalence of complications ranged from 0% to 48% (Table 2).[5-12] AVF-related complications in our study included head, neck, and chest edema, as well as occlusive thrombosis. Magee et al stated that upper extremity edema is prevented by distal vein ligation during AVF creation.[8] However, our patients who experienced head, neck, and chest edema (but not upper extremity edema) underwent distal vein ligation. Interestingly, cohorts reported by McElhinney and Quiñonez did not have distal vein ligation.[9,11] McElhinney et al reported zero complications of edema (0/11, 0%), whereas Quiñonez et al reported both upper extremity edema (2/11, 18%) and cephalic edema with headaches (1/11, 9%). Quiñonez et al speculated that distal vein ligation may increase venous pressures and subsequently cause cephalic edema and headaches.

Multiple technical factors, in addition to distal vein ligation, may account for differences in complication rates. Potential technical factors include fistula size, creation of a direct side-to-side anastomosis vs placement of a synthetic shunt material (ie, PTFE shunt), and use of postoperative anti-coagulation and/or anti-platelet therapy. For our cohort, similar to previously published cohorts, fistula size was determined case-by-case by the surgeon based on patient size and clinical status. None of the patients in our cohort had a change in their anti-coagulation or anti-platelet regimen prior to or following AVF creation. As previously stated, all patients who survived to hospital discharge were continued on aspirin monotherapy. This strategy is consistent with the available data in previous reports.

AVF creation remains a rare palliative procedure at our institution. In general, we consider AVF creation for patients after SCPC who are not candidates for TCPC, or potentially for patients with severe hypoxia after SCPC or TCPC as a bridge to OHT. As similarly described by Quiñonez et al, our current practice is to place a 4-5 mm PTFE shunt to construct the AVF because the PTFE shunt allows greater control over fistula size and reduces risk of future growth potential compared to direct side-to-side anastomosis.[11]

Our retrospective study has several limitations. This small, heterogenous cohort limits robust statistical analysis, generalizability, and valid comparison to a matched cohort. In addition to a heterogenous cohort, AVF size, location, and surgical technique were also variable because AVFs were performed by multiple surgeons over an extended time period. These important differences could impact interpretation of AVF outcomes. Hgb may also impact systemic oxygen saturations in this population. Unfortunately, multiple Hgb values were not available for the majority of patients during long-term outpatient follow-up for meaningful analysis. Finally, an important outcome after AVF creation is exercise tolerance. Our available data limited objective comparison of pre- and post-AVF exercise capacity (6-minute walk test and/or cardiopulmonary exercise test). Inclusion of objective exercise measurements pre- and post-AVF could be helpful for future study.

5 | CONCLUSION

Palliative AVF creation for patients with single ventricle CHD and hypoxia does not universally improve oxygen saturation and is prone to complications. Despite lack of durable benefit and complication risks, AVF creation remains a reasonable palliation for a subset of patients after SCPC who are not candidates for TCPC, or potentially as a bridge to OHT.

CONFLICT OF INTEREST

The authors declare that they have no conflict of interest with the contents of this article.

AUTHOR CONTRIBUTIONS

ADS: Responsible for concept/design, data collection, data analysis/interpretation, statistics, draft preparation, critical revision of article, and approval of final article. SJK: Responsible for data analysis/interpretation, critical revision of article, and approval of final article. RKW: Responsible for critical revision of article and approval of final article. SG: Responsible for concept/design, data analysis/

interpretation, statistics, critical revision of article, and approval of final article.

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