Congenital Cardiovascular Interventional Study Consortium

Brief Report – Pulmonary Flow Restrictor (PFR) Registry

Date: November 4, 2025

PFR Registry Organizers

Daisuke Kobayashi, MD: St. Louis Children's Hospital / Washington University in St. Louis
David Balzer, MD: St. Louis Children's Hospital / Washington University in St. Louis
Thomas Forbes, MD: Joe DiMaggio Children's Hospital /Memorial Health System

---

Executive Summary

This brief report presents the results of a preliminary analysis from the **Pulmonary Flow Restrictor (PFR) Registry**. The registry was launched in mid-2024 and has rapidly evolved as a collaborative, multicenter effort. As of this report, **148 cases** have been enrolled from **17 participating centers**, with **132 patients** included in the current analysis.

Upon request from the **PDA Stenting Symposium 2026** organizers, early registry data were presented at the conference. This document is intended to provide an overview of the collected data, highlighting patient demographics, procedural characteristics, and early outcomes.

This report is **not a formal publication** but rather an internal summary designed to illustrate the registry's current progress and share initial observations of interest with collaborators. The findings reflect where the registry stands today and provide insight into the evolving practice of transcatheter PFR implantation.

The registry continues to expand its participating sites and accumulate procedural experience through collaborative data collection. The registry organizers are committed to providing timely feedback, promoting data sharing, and advancing best practices in the care of patients undergoing PFR implantation.

Document Content

This brief report provides an early update and insight from the ongoing multicenter PFR Registry, presenting:

- **Registry progress** and current enrollment status
- **Patient demographics and baseline characteristics**
- **Procedural data** of PFR implantation
- **Preliminary one-year outcomes** based on ongoing data collection

1. Registry Progress

- **Total enrollment:** 148 cases
- **Participating centers:** 17
- **Study cohort analyzed:** 132 patients (BL branch PAs n=126, Single PA branch n=6)
- **Annual enrollment:** steadily rising; **2025 marks the highest year-to-date volume**
- **Unique implantation locations (n = 7):** modified BTS (n=2), stented PDA (n=2), Sano conduit (n=2), and central shunt (n=1).
- **Top enrolling centers:**
  Phoenix Children's (n = 27), St. Louis Children's (n = 23), Children's Hospital of Alabama (n = 22), Columbia University / New York-Presbyterian (n = 15), Children's Hospital Los Angeles (n = 13).

Figure. Bar graph showing number of site participation and cumulative case enrollment from 2024Q2 to 2025Q3.

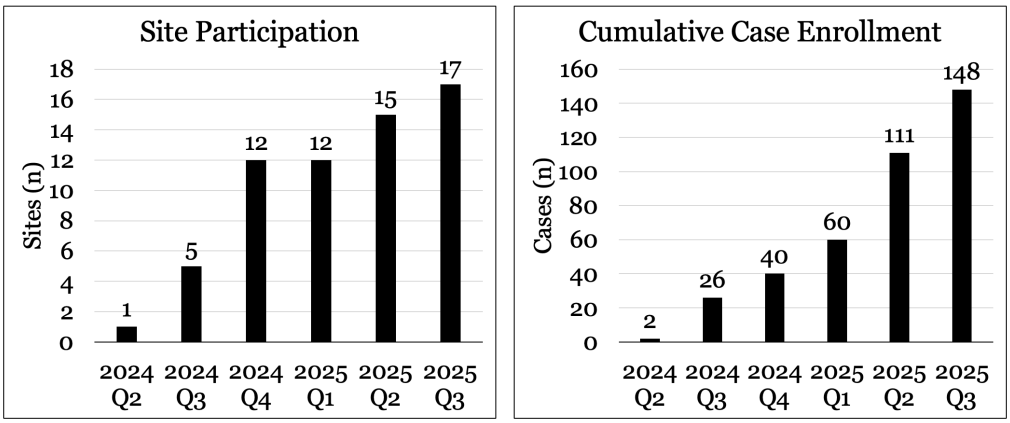


Figure. Number of enrolled cases per year. Year 2025 has the largest number of enrollments.

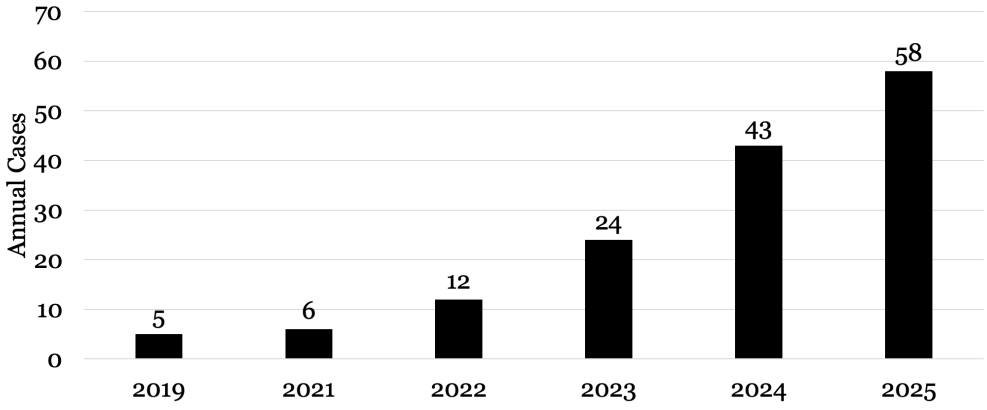


Figure. Number of case enrollment per center.

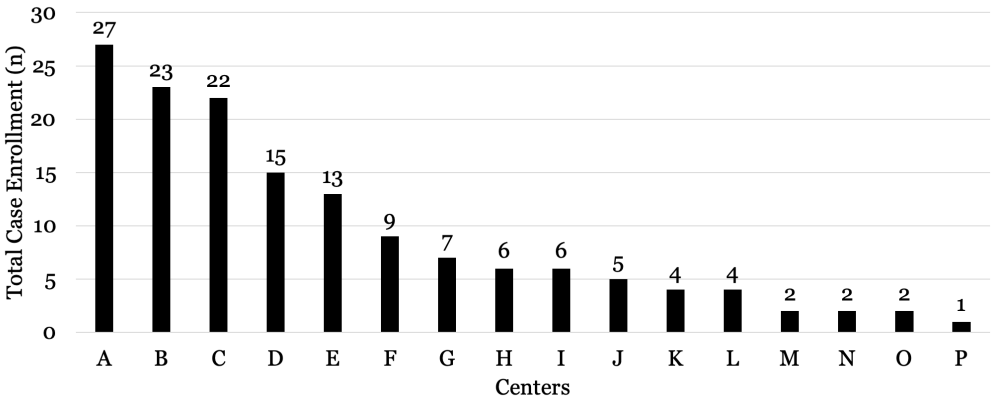
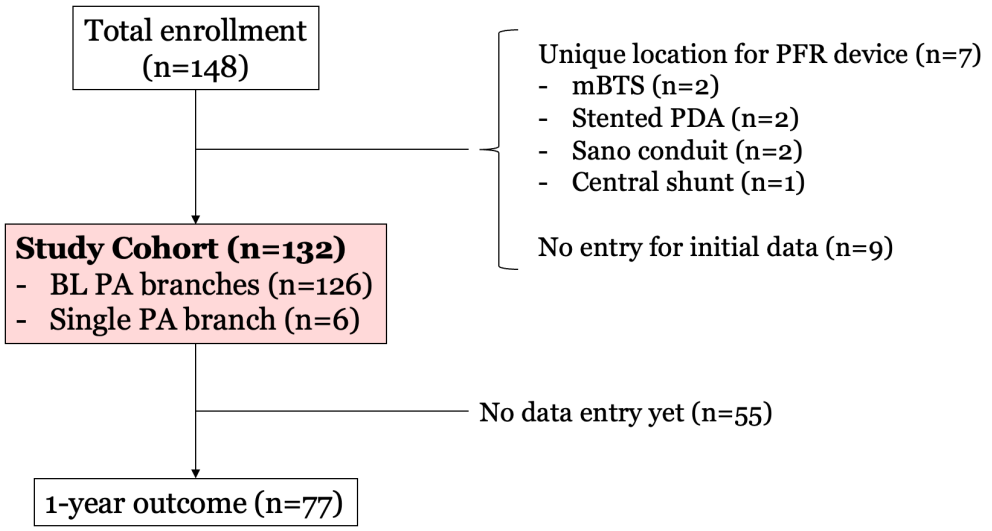


Figure. Study cohort tree



2. Patient Demographics and Baseline Characteristics (n = 132)

Characteristic	Findings
Male	n= 70 (53%)
Gestational age	Median 37 weeks (range 28–40)
Premature (<37 weeks)	n= 46 (35%)
Birth weight	Median 2.7 kg (range 0.91–4.12)
Low birth weight (<2.5 kg)	n= 50 (38%)
Ventricular physiology	SV n=55 (42%), BV n=61 (46%), Undetermined n=16 (12%)
Genetic syndrome	n= 51 (39%) — frequent 22q11 deletion, Trisomy 18/21, Heterotaxy, and others
Comorbidities	n= 68 (52%) — IUGR, chronic lung disease, renal insufficiency, brain malformation, airway issues, and others
Moderate/severe ventricular dysfunction and/or AV valve regurgitation	n= 34 (26%)

Cardiac diagnoses based on ventricle morphology status

1. **Single ventricle (n=55):** HLHS (n=36), other SV CHD (n=10), Unbalanced AVSD (n=6), Shone's complex (n=1), Aortic stenosis (n=1)
2. **Biventricle (n=61):** Other BV CHD (n=34), VSD (n=14), AVSD (n=6), Shone's complex (n=6), aortic stenosis (n=1)
3. **Undetermined (n=16):** unbalanced AVSD (n=4), VSD (n=3), HLHS (n=3), aortic stenosis (n=1), other SV CHD (n=3), other BV CHD (n=1), Shone's complex (n=1)

Genetic Syndrome

A **confirmed genetic syndrome** was identified in **51 patients (39%)**, highlighting the strong association between congenital heart disease requiring PFR and underlying genetic abnormalities.

1) **Frequent syndromes included:**
   - **22q11 deletion (n = 11)**
   - **Trisomy 18 (n = 5)**
   - **Trisomy 21 (n = 5)**
   - **Heterotaxy (n = 4)**
2) **Other reported syndromes and pathogenic variants** encompassed a wide spectrum of chromosomal and single-gene disorders, such as:
   Apert syndrome, Jacobsen syndrome, Kabuki syndrome, Phelan-McDermid syndrome, Pompe disease, Sotos syndrome, Turner syndrome, Noonan syndrome, Ehlers-Danlos syndrome, VACTERL association, X-linked adrenoleukodystrophy, Cri du Chat syndrome with 16p duplication, cystic fibrosis, **SMAD** mutation, **pathogenic**

TTN variant, **15q11.2 microdeletion**, **1q21.1 interstitial loss**, **chromosome 4q, 5p/5q, 6q, 8, 10q, 13, and 21/11** structural abnormalities, and multiple dysmorphic features of uncertain etiology.

Pre-PFR Echocardiography Findings (n=132)

1) **Systemic ventricular systolic function:**

- Normal — 98 (74%)
- Mildly reduced — 13 (10%)
- Moderately reduced — 10 (8%)
- Severely reduced — 11 (8%)

2) **Atrioventricular (AV) valve regurgitation:**

- None / trivial — 94 (71%)
- Mild — 18 (14%)
- Moderate — 16 (12%)
- Severe — 4 (3%)

Overall, **34 infants (26%)** demonstrated **moderate or severe ventricular dysfunction and/or significant AV valve regurgitation**, indicating a substantial proportion of patients with compromised pre-procedural cardiac function.

Comorbidities

Comorbid conditions were common, present in **68 patients (52%)**, underscoring the medically fragile nature of infants undergoing PFR implantation.

1) **Frequent comorbidities** included:

- IUGR (n = 26)
- Chronic lung disease (n = 10)
- Renal insufficiency (n = 10)
- Brain malformation (n = 8)
- Airway abnormality (n = 6)
- SGA (n = 7)
- Hepatic disease (n = 5)
- Neurological disorder (n = 5)
- Hypotonia (n = 2)
- Infection (n = 3)
- NEC (n = 1)
- Fetal hydrops (n = 1)

2) **Other reported significant comorbidities** included agenesis of the right lung with absent RPA and LPA sling, transient abnormal myelopoiesis after chemotherapy, congenital syphilis with maternal substance exposure, giant omphalocele with micrognathia, gastric perforation, obstructed TAPVR with renal hypoplasia, cardiogenic shock, and esophageal atresia.

Summary:
More than half the infants were **high-risk**, frequently premature, low birth weight, or with genetic syndromes and complex comorbidities.

3. Initial indications for PFR Implantation (n=132)

- **Bridge to single ventricle palliation: n=50, 38%**
- **Bridge to biventricular repair: n=65, 49%**
- **Bridge to transplant: n=10, 8%**
- **Palliative purpose: n=7, 5%**

Bridge to Transplant (n=10)

	Case details
1	HLHS with **severe** TR and RV dysfunction
2	HLHS with **significant** pulmonary valve abnormality
3	HLHS with **severe** ventricular dysfunction and severe AVVR
4	HLHS with **obstructed** TAPVR and intact atrial septum
5	HLHS with **Coronary atresia**
6	IAA/VSD, **Severe** ventricular dysfunction
7	Critically ill on VA **ECMO**
8	Unbalanced AVCD, CoA, cystic fibrosis, **severe** ventricular dysfunction
9	Heterotaxy, unbalanced AVCD, **aortic atresia** with dysplastic pulmonary valve
10	Truncus arteriosus with **severe** truncal valve insufficiency

Palliative Purpose (n=7)

	Case details
1	**Trisomy 18**, VSD, Heart Block
2	**Trisomy 18**, 30-wk GA, VSD, ventricular dysfunction
3	**Trisomy 18**, 2.1 kg, VSD, PH, giant omphalocele
4	**Apert syndrome**, **HLHS**, severe extracardiac anomaly
5	1.7 kg, **4p deletion, 10q duplication**, DORV, CoA, Respiratory failure, stage 4 CKD
6	DIRV, arch hypoplasia, **moderate AVVR**
7	**1.1 kg, 32-week GA, HLHS**, twin-twin transfusion Sx, moderately reduced function

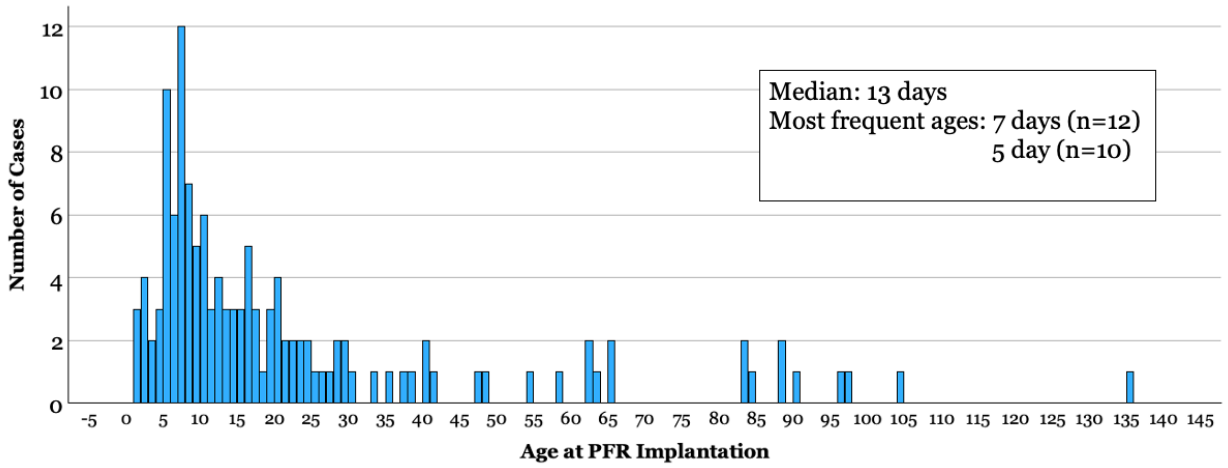
***Interventions Prior to PFR Implant (n=15)**

- Atrial septal intervention (n=6)
- PDA closure/closure attempt (n=2)
- Balloon aortic valvuloplasty (n=2)
- Vertical vein stenting (n=2)
- Surgical CoA repair (n=1)
- Aborted Norwood (n=1)
- ECMO (n=1)

4. Procedural Characteristics

Among 132 patients included in the analysis, the **median age at the time of PFR implantation** was **13 days**, with the most frequent implantation occurring at **5 and 7 days of life.** The **median body weight** was **3.0 kg**, and **36 infants (27%)** weighed **<2.5 kg** at the time of the procedure.

Figure. Histogram showing the distribution of age at the PFR implantation



Pre-procedural hemodynamic instability and respiratory compromise were common. **Inotropic support** was required in **48 patients (36%)**, while **endotracheal intubation** was present in **78 (59%)**, and **mechanical ventilation** was used in **77 (58%)**. **Ductal-dependent systemic or pulmonary circulation** was present in **93 patients (71%)**. The **median pre-procedural oxygen saturation** by pulse oximetry was **91%**.

PFR implantation procedure: Echocardiographic guidance was utilized in **54 procedures (41%)**, and the **overall procedural success rate** was **92% (122/132)**. The **Microvascular Plug (MVP)** was placed in **bilateral pulmonary artery (PA) branches (n = 126)**, while **single-branch placement** occurred in **6 patients**.

PFR device position: For the **right pulmonary artery**, the device was positioned proximally in **44% (n=58)**, straddling with non-compromised flow in **22% (29)**, and distally in **27% (n=35)**. For the **left pulmonary artery**, the device was positioned proximally in **61% (n=81)**, straddling with non-compromised flow in **27% (n=27)** and with compromised flow in **2% (n=2)**, and distally in **9% (n=12)**. The PFR device in the RPA tended to be positioned more distally than in the LPA.

The **systemic oxygen saturation** decreased slightly but significantly after device implantation (**pre: 88 ± 13%; post: 84 ± 9%; p = 0.004**), consistent with the desired physiologic effect of pulmonary flow restriction.

Table. MVP size selection and Proximal branch pulmonary artery measurement

RPA	N (%)	Prox RPA measure (mm) *mean ± SD	Prox RPA measure (mm) *median (IQR)
3Q	1 (1%)	NA	NA
5Q	9 (7%)	3.8 ± 1.0	3.6 (3.0 – 4.9)
7Q	**82 (62%)**	5.0 ± 1.2	5.1 (4.6 – 5.5)
9Q	40 (30%)	6.9 ± 1.0	6.9 (6.0 – 7.7)
LPA	**N (%)**	**Prox LPA measure (mm) *mean ± SD**	**Prox LPA measure (mm) *median (IQR)**
3Q	1 (1%)	NA	NA
5Q	9 (7%)	3.9 ± 0.8	3.9 (3.2 – 4.5)
7Q	**82 (62%)**	5.1 ± 0.7	5.0 (4.6 – 5.6)
9Q	40 (30%)	7.0 ± 1.4	6.9 (6.0 – 7.9)

Fenestration Technique

Fenestration technique of the MVP varied among centers. The most common method was **Bovie cautery (n = 70)**, followed by **dilator-based fenestration (n = 42)** and **scalpel puncture (n = 19)**. Among dilator techniques, **5F dilators** were most frequent (**n = 25**), followed by **4F dilators (n = 9)**.

Most devices had **one fenestration (n = 104)**, while **two (n = 25)** and **three (n = 2)** fenestrations were less common. The **estimated fenestration size** was typically **1–2 mm (n = 108)**, with **larger fenestrations (≥ 3 mm)** in **18 patients (14%)**.

Concurrent and Additional Procedures

Concurrent transcatheter interventions were performed in **38 patients (29%)**, including **PDA stenting (n = 21)**, **balloon atrial septostomy or ASD dilation (n = 22)**, **atrial septal stent placement (n = 1)**, **PDA occlusion (n = 1)**, and **balloon aortic valvuloplasty (n = 1)**.

PFR retrieval after initial release occurred in **14 patients (11%)**.

Radiation and Procedural Metrics

Median **procedure time** was **97 minutes (IQR 66–148)**, **fluoroscopy time** was **28 minutes (IQR 18–51)**, and **dose–area product per kilogram (DAP/kg)** was **61 µGy·m²/kg (IQR 25–116).**

5. Adverse Events

Adverse events occurred in **17 of 132 patients (13%)**, classified as follows:

- **Catastrophic (Level 5):** 2
- **Major (Level 4):** 4
- **Minor & Moderate (Level 2&3):** 11

Catastrophic Events (Level 5)

- Intra-procedural cardiac arrest during PDA stenting → death
- Severe post-implant hypoxemia with bradycardia → death

Major Events (Level 4)

- Severe hypoxia requiring device removal and surgical bilateral PA banding
- Cardiovascular decompensation after sheath removal requiring ECMO
- SVT after ASD creation, with hypoxia and CPR, necessitating ECMO
- Pulmonary valve perforation requiring surgical repair

Minor & Moderate Events (Level 3)

- Arrhythmias compromising hemodynamics (**n = 3**)
- Pulmonary valve injury (**n = 1**)

*Acute PFR thrombosis (**n = 1**, successfully managed)

6. Post-Procedural Anticoagulation and Antiplatelet Therapy

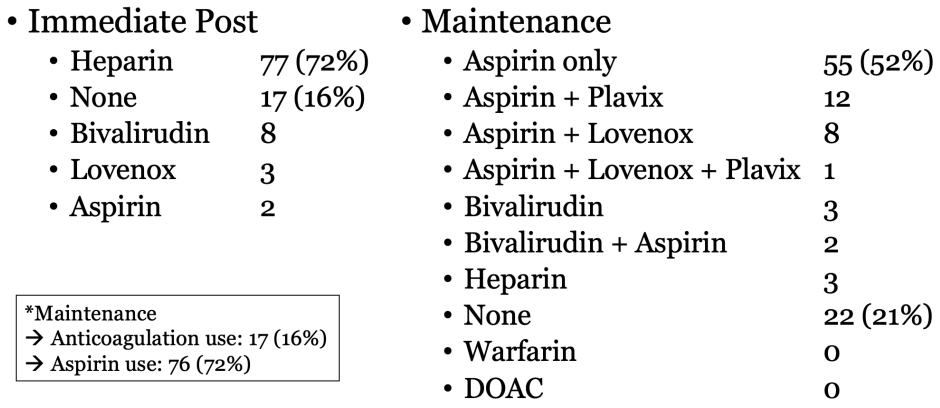
Post-procedural anticoagulation practices were heterogeneous. Among **107 patients** with available data, **immediate anticoagulation** following the procedure included:

- **Heparin** in **77 (72%)**,
- **Bivalirudin** in **8 (7%)**,
- **Lovenox** in **3**, and
- **No immediate anticoagulation** in **17 (16%)**.

Maintenance therapy commonly included **Aspirin monotherapy (55 patients, 52%)**, followed by **Aspirin + Plavix (12, 11%)**, **Aspirin + Lovenox (8, 8%)**, and **triple therapy (1 patient)**.

Aspirin use at discharge was documented in **72%** of cases. **No patients** received DOACs or warfarin during the early maintenance phase.

Figure: Immediate and maintenance anti-coagulation and Anti-platelet therapies after PFR implantation

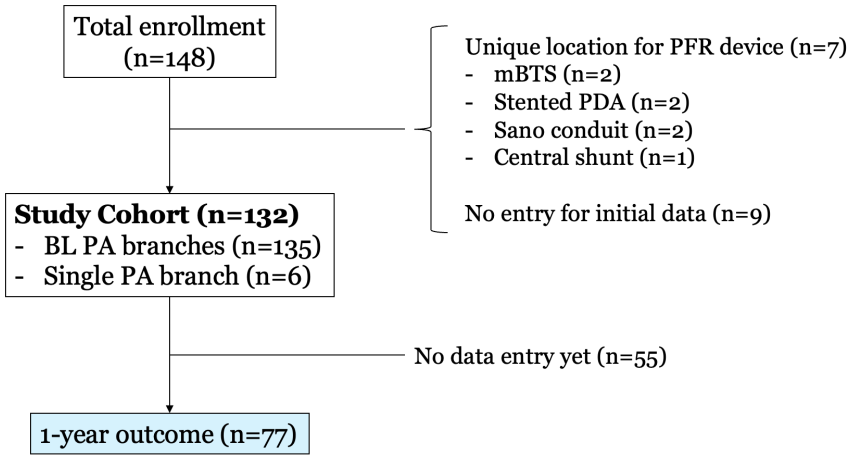


7. PA intervention at and after surgical PA removal (surgical data available in 67 cases)

At and/or after device removal, **pulmonary artery intervention** was performed in **16 of 67 patients (24%)**, most commonly angioplasty or stenting for residual stenosis or deformity at the former device site.

	At surgery	Post-surgery	After discharge before 1 yo
1	Intraoperative LPA stenting		LPA stenting
2	Intraoperative LPA stenting		
3	Intraoperative LPA balloon angioplasty		BL PA balloon angioplasty
4	Surgical angioplasty for BL PAs	LPA balloon angioplasty	
5	Surgical angioplasty for LPA	LPA balloon angioplasty	
6	Surgical angioplasty for LPA		
7	Surgical angioplasty for LPA		
8	Surgical angioplasty for BL PAs		
9	2nd bypass run to remove MVP Gore-Tex		
10		LPA stenting	
11		LPA stenting	
12		LPA stenting and RPA balloon angioplasty	LPA stent re-dilation
13		RPA and LPA balloon angioplasty	LPA stenting
14		RPA balloon angioplasty	
15			PA balloon angioplasty
16			RPA surgical plasty and LPA stenting

8. One-Year Outcomes



At the time of analysis, **77 patients** had reached one-year follow-up. The **overall one-year survival** was **57% (44/77)**, including **two survivors following cardiac transplantation**.

Survival outcomes varied significantly according to ventricular morphology:

- **Single-ventricle physiology:** 43% (15/35)
- **Biventricular physiology:** 68% (25/37)
- **Borderline ventricle:** 75% (3/4)

When patients with **palliative or bridge-to-transplant intent (n = 10)** were excluded, the **adjusted one-year survival** was **60% (40/67)**.

9. Limitations and Future Directions

This analysis reflects early multicenter experience from **17 centers** and **148 cases**, the largest dataset to date on transcatheter PFR implantation. Uneven case distribution across centers may introduce selection bias.

The 6-year dataset (2019–2025) reflects evolving clinical practice, and earlier cases may not fully represent contemporary PFR techniques or procedural strategies.

Future goals include expanding participation to **>50–60 centers**, improving data completeness, and conducting analyses to identify predictors of **procedural success, adverse events, and outcomes**. Ongoing **collaboration** will be key to refining procedural strategy and improving results in this high-risk population.