

Fontan Outcomes Network Case Review Conference

Advanced Therapies For the Failing Fontan Circulation

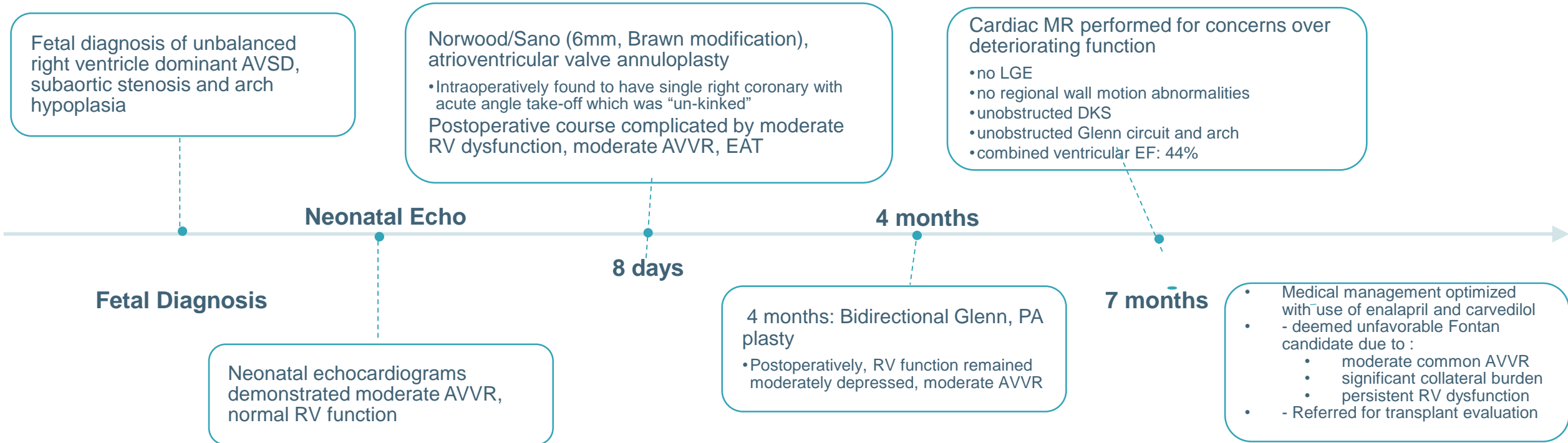
January 18, 2022

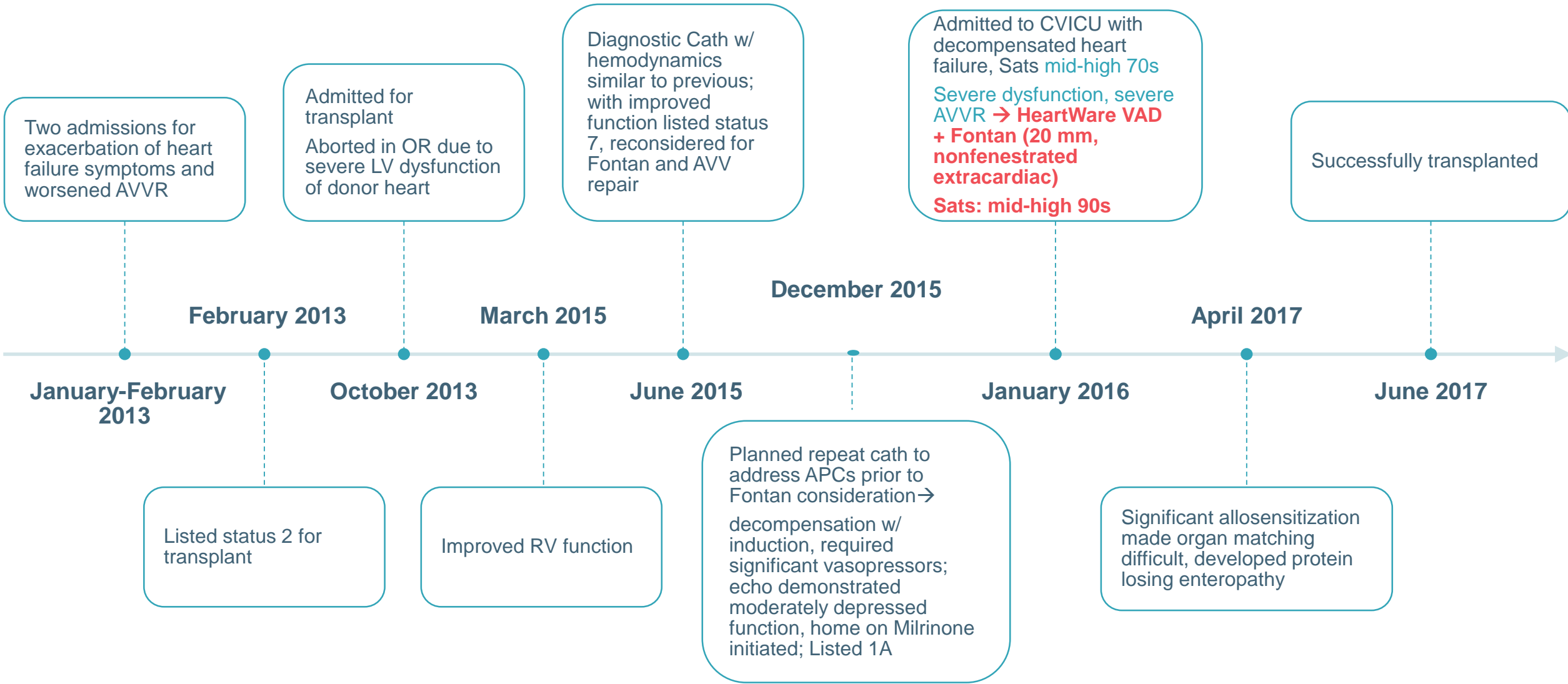
Texas Children's Hospital

Summary Slides



Case 1





Case 1 Key Points

- Case of chronic moderate-severe AVV failure, chronic systolic dysfunction, and some degree of lymphatic failure spanning from birth through Glenn stage.
 - Glenn circulation is less favorable for VAD support, in part due to dichotomous systemic venous return complicating decompressing benefits of VAD in this physiology.
- Concurrent construction of Fontan circulation with placement of a ventricular assist device, an approach thought to optimize candidacy for and clinical status going into eventual heart transplantation.
 - This strategy enacts not only improvement in oxygen delivery through employment of the VAD but also improvement in oxygen content when completing the Fontan circulation.
 - TCH experience: 5 such cases of Glenn + VAD converted to Fontan (median age: 4.7 years, median weight: 16 kg)
- This strategy supported growth/rehabilitation, maintained end organs, and ultimately allow for a ~ 1.5 year bridge to transplantation.

Case 2

Young Adult with Fontan failure:

- **Severe AVVR**
- Mild-mod dysfunction
- Ascites and peripheral oedema
- Multiple recurrent admissions

Anatomy:

Heterotaxy - asplenia, RAI
Dextrocardia
HLHS variant (mitral atresia, VSD)
DORV with pulmonary atresia
Morphologic RV
Right-sided IVC
Bilateral SVCs
Midline liver

Intervention:

Pulmonary valvuloplasty (1 week)
Bilateral bidirectional Glenn (1 year)
Fenestrated extra-cardiac Fontan (3 years)
AP collateral coil occlusion (2015)

Fontan Circulation complications

FALD

- coarse liver compatible with cirrhosis (MRI)
- heterogeneous and nodular cirrhosis (CT)

Bilateral hydrocoeles

Abdominal ascites

Iron-deficiency anemia

Current diagnosis
 Fontan failure
 Severe AVVR
 Mild-moderate RV dysfunction

Baseline anatomy
 Right atrial isomerism
 Heterotaxy
 Dextrocardia
 Single morphologic RV
 AVSD with severe AVVR

Previous interventions
 Extra-cardiac Fontan

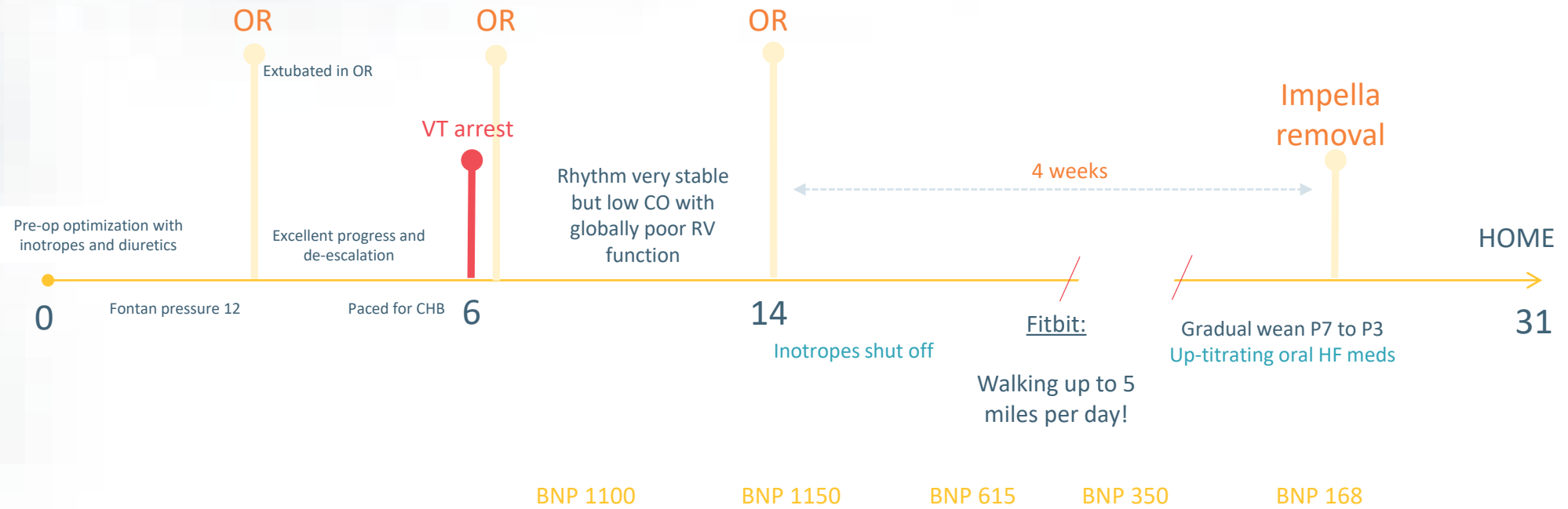
Threats
 Recurrent admissions with decompensated heart failure
 Ascites
 Borderline renal function

Procedures:

Redo sternotomy
 Mechanical AVVR

Permanent PPM Generator

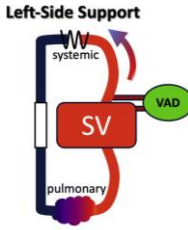
Axillary Impella 5.5
 10 mm Chimney graft



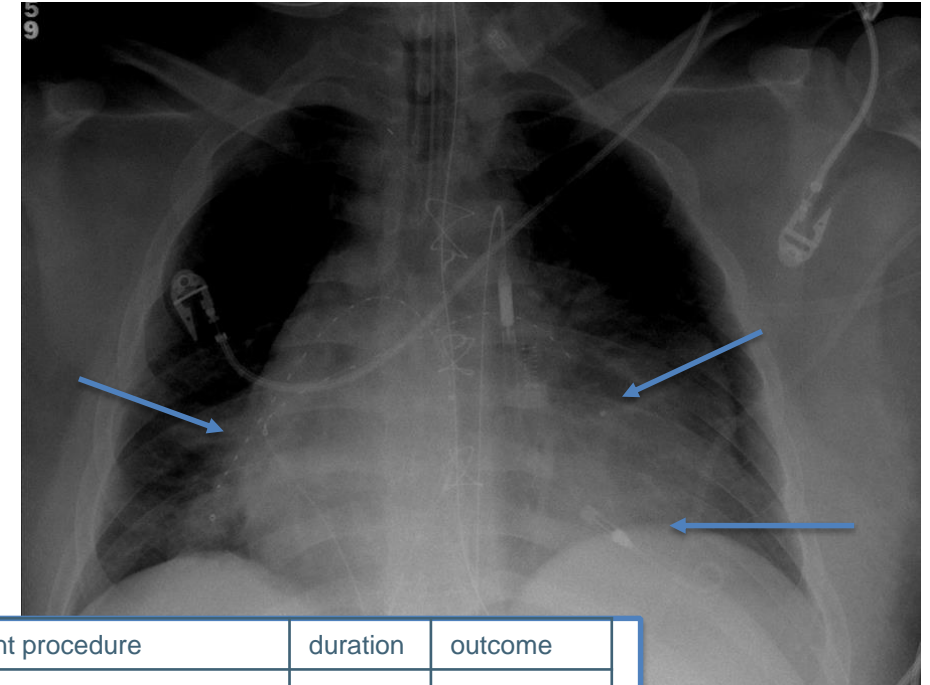
Axillary Impella 5.5 via 10 mm graft

Advantages	Complications
Axillary implant permits extubation, mobilization and rehabilitation	Ventricular ectopy and tachycardia
Fewer end-organ complications versus ECMO	Inadequate support (malposition, size)
Fewer bleeding and thrombo-embolic complications versus ECMO	Aortic regurgitation
Longer-term support (versus ECMO); up to ~5 weeks	Systemic AVV regurgitation
Effective cardiac decompression and enhanced cardiopulmonary recovery	Bleeding / pectoral hematoma
Permits weaning all inotropes and instead introduction of oral heart failure goal-directed therapy and heart rate control	Brachial plexus injury
	Hemolysis
	Thromboembolic complications

Percutaneous ventricular assist device use in failing Fontan circulation



- Percutaneous VAD (pVAD) can be used for stabilization after cardiac intervention¹
 - 2012-2015; n=10
 - Median age 26 yrs, range 4-38 yrs, 9/10 required inotropes prior to pVAD
 - Impella: 2.5 x3, CP x6, 5.0 x1; femoral implant 6/10
 - Duration of support between 3 hrs to 11 days and median 4 days;
 - 2 device removed due to hemolysis; 1 patient aortic insufficiency mild -> mod; 2/10 patients died prior to discharge



	age	sex	anatomy		Comorbids	Device	Concomitant procedure	duration	outcome
A	37	F	DILV, TGA	SV-Fontan	Cyanosis, CKD, FALD, heart block	Impella CP	None	4	Alive
B	32	M	DILV, TGA	SV-Fontan	Acute PE, Heart block, renal failure	Impella CP	Placement of acoustic pulse thrombolysis catheters	6	Alive
D	20	M	HS - dextrocardia, venous anomaly, AVSD, DORV, PS	SV-Fontan	Afib, VT, AVVR	Impella 5.5	left axillary graft	31	Alive

TCH/BSLMC data, accepted with revision

Case 2 | Key points:



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Adult Congenital
Heart Program

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- In-hospital optimization can dramatically improve baseline status before intervention
- AVV repair/replacement is a high-risk alternative to advanced therapies...*if peri-operative period can be navigated*
- Key to observe for declining function/reduced reserve especially when introducing afterload reduction and awareness for substrate for arrhythmias / dyskinesia
- Impella offered major advantages:
 - Immediately shut off inotropes
 - Immediate mobilization and rehabilitation
 - Sub-acute remodeling ~5 weeks
 - Up-titration of oral HF medications
 - Slow wean of support