

Challenges in the Management of Pulmonary Atresia and Intact Ventricular Septum (PA IVS)

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OBJECTIVE

With mortality reaching 22% by 9 years of life, pulmonary atresia with intact ventricular septum (PA IVS) remains one of the most severe single ventricle conditions of early childhood. We reviewed our center's experience of patients with PA IVS to identify best management strategies for these patients.

METHODS

A total of 66 records of patients with PA IVS were reviewed. The distribution of initial right ventricle (RV) sizes was collected from echocardiographic reports and was characterized as follows: severely hypoplastic (45), normal (9), severely dilated (1), and not recorded (11). The median tricuspid valve (TV) Z-scores of patients with severe RV hypoplasia and normal RV size were -3.1 (Range: -0.7 to -5.9) and -2.1 (Range -0.1 to -6), respectively. As defined by catheterization report, coronary sinusoids were present in 31 patients (47%) and right ventricular dependent coronary circulation (RVDCC) in 20 patients (30%).

RESULTS

Patients were divided into 5 end statuses: death (17), Fontan (20), biventricular repair (18), awaiting Fontan (6), and heart transplant (5). Six of the 17 deaths had RVDCC ($p = 0.6$) and 10 had sinusoids ($p = 0.3$). Only one patient achieved a biventricular repair with a Z-score of < -3 .

Patients Characterized by End Status

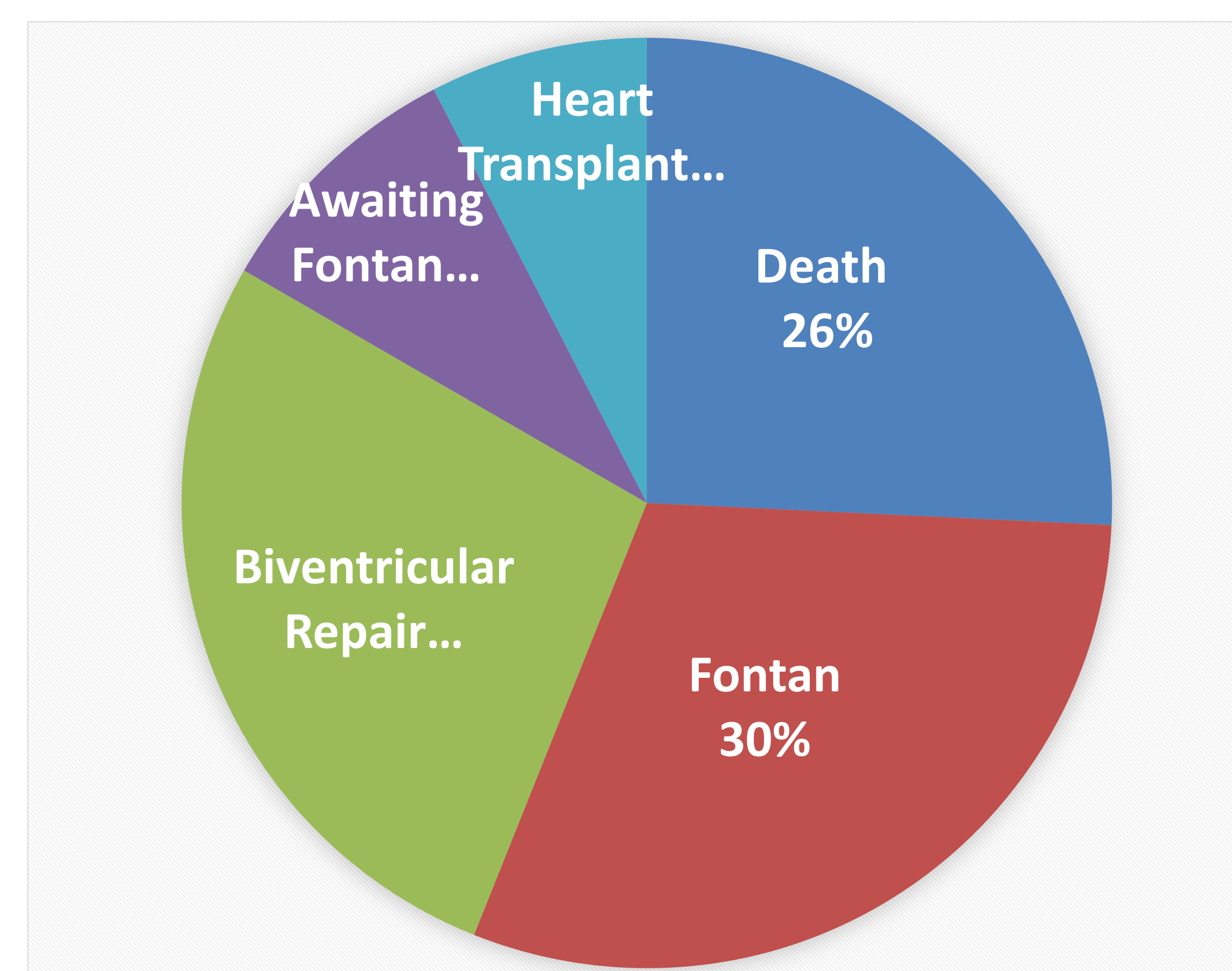


Figure 1. Breakdown of PA IVS patients in our center by end status.

Distribution of Patient Survival Following End Status

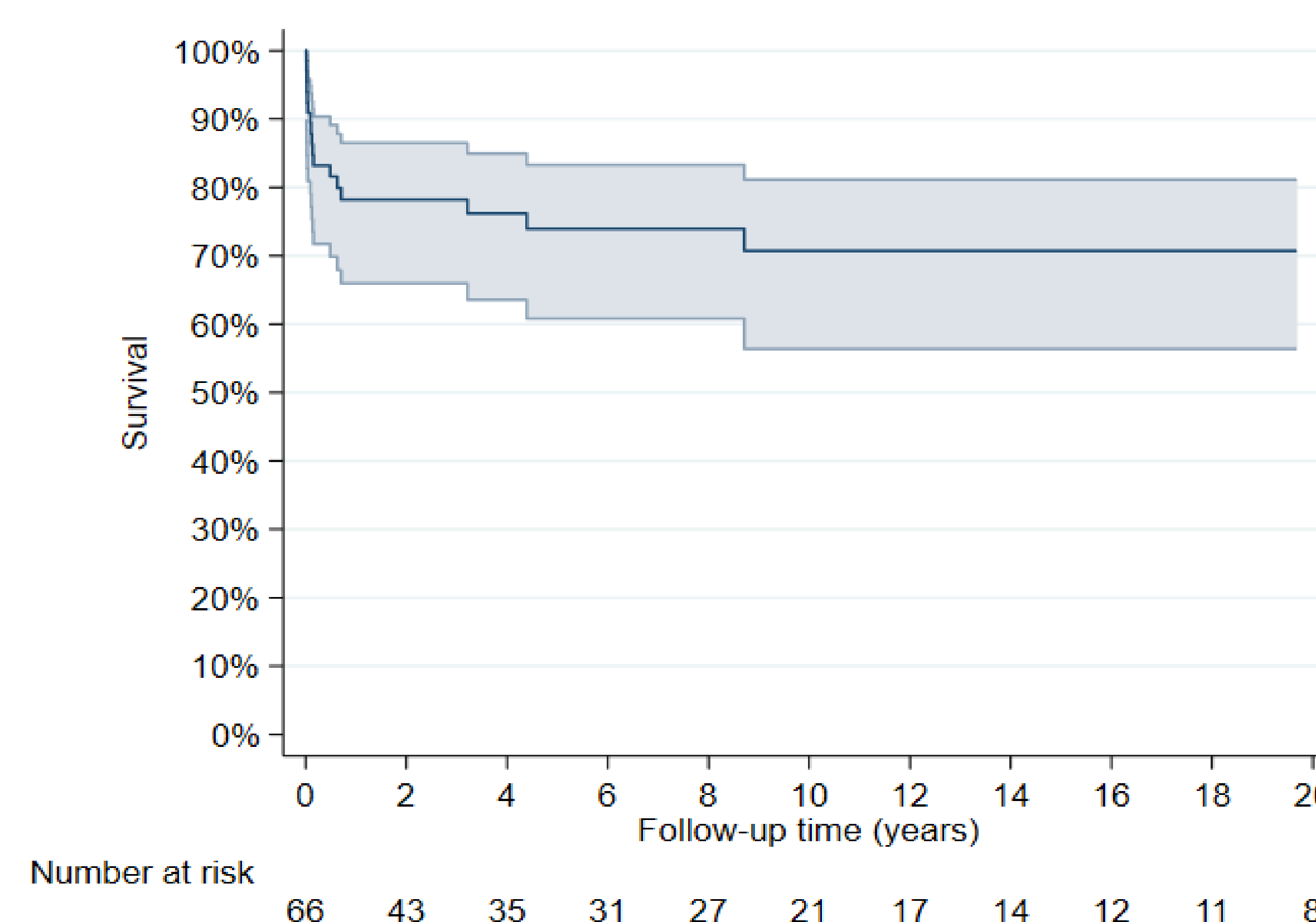


Figure 2. Kaplan Meier curve illustrating nearly 70% survival rate after 8 years follow up.

Distribution of Patient Survival Following End Status

Reason for Death	Number of Deaths (%)
Total Mortality	17 (100%)
Heart Failure	4 (23%)
Withdrawal of Treatment	4 (23%)
Sudden Cardiac Death	3 (18%)
Respiratory Failure	1 (6%)
Endocarditis	1 (6%)
Chronic Transplant Rejection	1 (6%)
Fontan failure	1 (6%)
BT Shunt Thrombosis	1 (6%)
Failure of Bidirectional Glenn Procedure	1 (6%)

Figure 3. Breakdown of PA IVS patients in our center by mortality.

RESULTS

Four of the 6 patients who had radio frequency perforation of the pulmonary valve followed by a surgical shunt procedure died when the two procedures were performed within the same hospital stay. Three of the 12 patients who had TV insufficiency died at 0, 5, and 7 months. No patients had any procedure to address TV insufficiency. Four of the 6 patients who had a severely hypoplastic RV and underwent a primary interventional catheterization died. Kaplan Meier shows a dramatic decrease in survival to 80% within 1st year of follow-up.

CONCLUSION

Our data shows that infants born with PA IVS continue to have lower survival than those with other single ventricle conditions and new approaches are needed to change this. Addressing TV regurgitation early in life and avoiding the opening of the right ventricular outflow tract (RVOT) of patients with a TV Z-score inferior to -3 might decrease mortality.

It is possible that some patients who need the creation of a systemic-pulmonary shunt would be better served by a gentle surgical opening of the RVOT at the time of the shunt procedure rather than a preceding catheter intervention.