

## Native Pulmonary Vein Stenosis: Encouraging Mid-term Outcome

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**Purpose:** Native (congenital) pulmonary vein stenosis (PVS) is a rare entity with limited outcome research. The obstruction may affect one or multiple veins and is often lethal. Multiple interventional approaches have evolved which include surgical and catheterization techniques. The objective of this study is to report midterm outcomes in related to these therapeutic modalities.

**Methods:** Retrospective review of the cardiac database identified 23 patients with native PVS over the last 13 years. Patients were divided into three groups based on type of initial intervention (balloon angioplasty, surgical angioplasty and surgical marsupialization). Patients with acquired PVS were excluded from the study. Mortality, and number of re-interventions were the primary outcomes. One-way analysis of variance models (ANOVA) and Fisher's exact tests were used to compare the subject characteristics between the interventions. Log rank tests and survival curves using the Kaplan-Meier product limit method were used to compare outcomes among the three interventions.

**Results:** The mean age at diagnosis was 13.4 months. Average age at initial intervention was 14.3 months with a mean follow up of 62 months. Gestational ages ranged from 23 to 40 weeks, and birth weights ranged from 440 to 3570 grams. The majority of subjects (61%) are pre-term infants. Of the 23 patients, 10 (43.5%) had balloon angioplasty, 3 (13.0%) had surgical angioplasty, and 10 (43.5%) had surgical marsupialization. The mean PVS gradient was reduced from 9.2 mmHg at time of diagnosis to 3.4 mmHg post intervention. No statistical significance was found between the interventions with respect to type of intervention in regards to survival time ( $p=0.5207$ ), re-intervention-free time ( $p=0.7823$ ) or re-intervention-free survival time ( $p=0.6122$ ). 18 of the 23 patients (78%) were alive at mean follow up of 62 months post intervention. No statistical significance between right side vs left side disease was noted. Residual post initial intervention gradient was significantly associated with time to re-intervention ( $p=0.0115$ ). Both higher pre and post initial intervention gradients were significantly associated with less survival time ( $p=0.012$  and  $p=0.0331$  respectively).

**Conclusion:** Outcome data on native pulmonary vein stenosis are limited in the current literature. Our study is unique as it is the first to compare outcomes between surgical and catheterization techniques purely in native PVS. In spite of the long follow up of 62 months (the longest so far reported in the current literature), survival of patients was an encouraging 78%. While earlier studies suggested superiority of surgical techniques we found no statistically significant difference between the types of intervention.