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Does timing of surgery predict long term respiratory outcomes in infants with Absent Pulmonary Valve Syndrome?

Torok K. (1), Brettle E. (2), Seale A. (1), Miller P. (1) ,Stickley J. (1), Nagakumar P. (2), Desai T. (1) Department of Cardiology (1), Department of Respiratory Medicine (2) Birmingham Children`s Hospital, United Kingdom

Background: Absent pulmonary valve syndrome (APV) is a rare congenital heart defect, usually associated with Tetralogy of Fallot (TOF), characterized by pulmonary insufficiency and marked pulmonary artery dilatation. Some patients develop respiratory failure due to bronchial compression and may require prolonged respiratory support post-surgical repair. We aimed to investigate outcomes of patients with APV and the need for prolonged respiratory support in relation to the timing of the surgery.

Methods: We retrospectively reviewed data of patients diagnosed with APV born or with estimated due date between January 2007 and January 2017. Prolonged ventilation was defined as greater than 3 months of non-invasive or invasive respiratory support.

Results: Thirty patients were identified of which 22 (73%) were prenatally diagnosed. Pregnancy was discontinued in one, in-utero death in three. One lost to follow-up.

Of the remaining 25 live born, 21 had classical TOF/APV and four were found to be “variants”. Variants all had patent ductus arteriosus along with disconnected left pulmonary artery (n□1), no aortic override with ventricular septal defect (n□1) and intact ventricular septum (n□2).

One baby died immediately after birth and two patients had palliative care due to severe airway compression and unable to wean ventilation support. Surgical repair was performed in 20/25 patients (80%), two are awaiting surgery.

Of those undergoing surgery, two patients died - one during surgery and a further patient died following worsening bronchomalacia five months post-surgery. Survival from birth of the surgical group at one and five years was 89% (CI 75 to 100%).

Six patients who underwent surgery (30%) required prolonged respiratory support post-operatively (Table).

Timing of surgery	Number of patients	Number alive	Number with prolonged respiratory support
< 1month	5	3	4
1 month – 6 months	7	7	2
6 months +	8	8	None

Conclusion: The majority of patients with APV are diagnosed antenatally. There is an important pre-operative mortality and 25% of babies undergoing surgery require prolonged ventilation. Babies having repair in the first month of life are particularly at risk. Further studies are needed to identify prenatal factors that predict postnatal cardio respiratory outcomes so parents can be counseled appropriately.