

## MP4-19

### Outcomes of congenitally disconnected and non confluent branch pulmonary arteries - a single centre experience over 24 years

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#### Aim:

To describe our experience of all children presenting with congenitally disconnected non-confluent branch pulmonary arteries. We studied the pattern of presentation, diagnostic modalities, intervention and outcomes.

#### Methods:

Retrospective review of data of children with diagnosis of disconnected pulmonary arteries between 1989 and 2013. We included all children with diagnosis of non-confluent branch pulmonary arteries with or without alternative pulmonary blood flow including pulmonary atresia. We excluded acquired disconnection of pulmonary arteries, and anomalous origin of branch pulmonary artery from aorta.

#### Results:

35 patients were identified. 85% of our population presented during infancy (age range from 4 days to 17years) with slight female preponderance (females 20 vs males 15). The commonest mode of presentation was in association with congenital cardiac condition (cyanotic in 30, non-cyanotic in 4) except for one patient with isolated disconnection with coexisting lung hypoplasia. The most common congenital cardiac lesion was Tetralogy of Fallot. The disconnected branch pulmonary artery was on right-sided in 6(17%) vs left-sided in 22(83%). The supply to the disconnected segments was from ductus arteriosus in 22 patients, the aortic arch was right-sided in 10(28%) vs left-sided 20(56%) and unknown in 5(12%). 8 patients had associated genetic syndrome.

Diagnosis was confirmed by cardiac catheterisation in 21 patients (60%). Median age at first intervention was 10 months (1 to 254 months). Primary intervention was trans-catheter in 6(18%) and surgery 28(82%). Average number of procedures after initial intervention was 5 (range 1-9). One patient had unsuccessful recruitment. At last follow up, 24 patients with full correction were doing clinically well and 1 had developed with pulmonary hypertension. 2 remains cyanotic and awaiting further intervention. . Mortality in this series was three patients (8%).

#### Conclusion:

Congenitally disconnected branch pulmonary arteries is a rare abnormality. Most patients will need staged palliation approach before complete repair with good long term outcome.