

## MP3-19

### Outcome of symptomatic partial atrioventricular septal defect necessitating repair during infancy.

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Background: Patients with partial atrioventricular septal defect are typically asymptomatic and electively repaired at pre-school age.

We sought to analyse the mortality, re-operation rate and risk factors with adverse outcome of symptomatic infants with partial atrioventricular septal defect necessitating repair during infancy in one UK centre in recent era (2000-15).

Methods: The inclusion criteria were as follows: 1. partial atrioventricular septal defect, 2. heart failure unresponsive to medical treatment, and 3. bi-ventricular repair during the first year of life. Thirteen patients (6 males), who had undergone corrective surgery at a median age of 197 (16-346) days, were included. Seven patients had an associated syndrome: Down (n=3), trisomy 47XXX (n=1), Noonan (n=1) and VACTERL (n=1). Five patients required either surgery (4 coarctation repair) or transcatheter intervention (1 balloon angioplasty of pulmonary artery) before primary repair. One patient with tracheobronchomalacia required cardiopulmonary resuscitation before repair.

Results: Median weight at primary repair was 5.1 kg (3.3-7.5 kg), median hospital stay 19 days (4-128 days). Five patients had unfavorable anatomy of the left atrioventricular valve: the leaflets were severely dysplastic in 2 cases, mildly dysplastic in 1 case, 1 patient had unusual orientation of the zone of apposition and 1 patient small left atrioventricular valvar annulus.

One patient died 68 days after primary repair from a non-cardiac cause.

Five patients (42%) were reoperated at a median interval of 2.8 years (33 days–8.3 years), including left atrioventricular valvar repair for severe regurgitation in 3 cases, relief of subaortic (n=1) and pulmonary vein stenosis (n=1). One patient required repetitive left atrioventricular valve repair, followed by mechanical valve replacement and pacemaker implantation.

The probability of developing severe left atrioventricular valvar regurgitation 10 years after primary repair in patients with unfavorable anatomy was 100% and 0% in patients with favorable anatomy ( $p < 0.01$ ).

After a median follow-up of 3.8 years, all patients were asymptomatic from the cardiac point of view and no patient had a severe atrioventricular valvar regurgitation.

Conclusion: Although the in-hospital mortality is not negligible in patients necessitating repair of partial atrioventricular septal defect in infancy, the long term outcome is favorable with no reported mortality and good functional status. The rate of re-operation mainly due to severe left atrioventricular valvar regurgitation is high in patients with dysplastic valve.