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Impact of Down syndrome in children with complete atrioventricular septal defect (cAVSD)

Background: Complete atrioventricular septal defect (CAVSD) is frequently (60-80%) associated with Down's syndrome. In the current era of early diagnosis, better preoperative care and early intervention, the postoperative outcomes following complete atrio-ventricular septal defect (CAVSD) repair have improved significantly. Trisomy 21 was shown to reduce risk of re-operation due to lower incidence of dysplastic atrio-ventricular valves in CAVSD. Little is known about the effect of trisomy 21 or atrioventricular valve dysplasia on early postoperative outcome following CAVSD repair.

Methods: Over a 6-year period (2004-2009) we evaluated the early postoperative outcomes following complete AVSD repair and looked for any difference between infants with (DS) and without (NDS) Down's syndrome with special reference to pre-operative echocardiogram findings, atrioventricular valve (AV valve) morphology, immediate and early post-operative clinical course.

Results: In our study, we found no difference in the prevalence of AV valve dysplasia between DS and NDS groups. Following complete AVSD repair, we found no significant difference in cardiac complications but prevalence of non-cardiac complications was significantly higher in DS group. There was no significant difference in lengths of ventilation and PICU stay between two groups. The overall incidence of early re-operation was 4.5%. The incidence of permanent pacemaker insertion was <2%. There was no early mortality.

Conclusions: In our study, we found no difference in the prevalence of AV valve dysplasia between children with and without DS undergoing CAVSD repair. This finding contrasts with previously published data, and further confirmatory studies are required. Although clinical outcomes were similar, children with DS had a significantly higher prevalence of non-cardiac complications in the early postoperative period than children without DS.

Biography

Ajay Desai is a Consultant in paediatric intensive care medicine at Royal Brompton Hospital in London, United Kingdom. He is the Lead for the paediatric extracorporeal membrane oxygenation (ECMO) programme, with a special interest in extracorporeal cardiopulmonary resuscitation (ECPR).

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