Atrioventricular Septal Defects

The atrioventricular septal defect (AVSD) is a spectrum of disorders, ranging from partial AVSDs, transitional AVSDs to complete AVSDs as well as AVSD associated with heterotaxy syndrome and also Tetralogy of Fallot. All of these defects comprising the spectrum are characterized by the existence of a common AV junction.[1]

Operations for atrioventricular septal defects carry substantial risks, and these are one of the most challenging groups of children for cardiac surgeons and anesthetists, taking their care. Children with AVSDs, especially the complete AVSD have a significant morbidity and mortality resulting from postoperative left atrioventricular valve regurgitation, residual intracardiac shunts, postoperative pulmonary hypertension, and various life-threatening cardiac arrhythmias.

The first successful surgical repair of a complete AVSD was done by Lillehei *et al*. in 1955. Using the technique of controlled cross circulation.[2] Long-term results from earlier studies[3,4] had demonstrated a 15 years survival of 86%–89%, however despite improved survival, several single-center studies have reported that up to 10% of patients after complete AVSD repair will require a reoperation within 10–15 years, for left A-V valve regurgitation and left ventricular outflow tract obstruction. Ginde, *et al*.[5] operated on 198 children with complete AVSD between 1974 and 2000. Their overall perioperative mortality was 10.1% with a significant decrease to 2.9% in the late surgical era 1991–2000. The estimated overall survival for the entire cohort was 85% at 10 years, 82% at 20 years and 71% at 30 years. The indications for reoperation included Left AV valve (LAVV) regurgitation 7.1% and left ventricular outflow tract obstruction 3.5%. Need for reoperation was a significant risk factor for late mortality. Most of the late deaths were cardiac related, attributable to complications from residual cardiac lesions, or from postoperative complications after reoperations for residual cardiac lesions. The improvements in mortality rates in later years was attributed to improvement in perioperative management including techniques of cardiopulmonary bypass (CPB), myocardial protection, anesthetic and postoperative management. Pulmonary hypertension may have contributed to risk for late mortality; children repaired in the early era were older at the time of surgery and more likely to have the pulmonary vascular occlusive disease.

Surgery for LAVV regurgitation is the most common indication for reoperation occurring very early at a median period of 1.1 years after the first surgery. Reoperation can be better achieved with valve repair rather than valve replacement. Left ventricular outflow tract obstruction is frequently due to an acquired discrete subaortic membrane. In addition, the classic I patch technique of surgical repair is a risk factor for reoperation, largely due to residual ventricular septal defects, and patch dehiscence. The classic I patch technique involves dividing the common AV valve leaflets and suspending them from a single patch resulting in more distortion of the valve tissue and AV valve regurgitation. The risk appears lower with the two patch repair technique.

LAVV valvuloplasty is superior to left LAVV replacement because of preserved somatic growth, and absence of hemorrhagic, thromboembolic, and infectious complications, however durability of repair is it's weakest link, especially in children with complex LAV valve pathology such as LAVV dysplasia, double outlet LAVV, and pseudoparachute LAVV. Symptomatic infants, particularly those with complete AVSD, should be operated at 1–3 months of age to prevent the development of LAVV dysplasia and annular dilatation. Routine LAVV cleft closure and commissural and/or annular plication should be routinely performed at primary repair.[6]

Pulmonary artery (PA) banding is no longer used in the elective routine repair of complete AVSD in favor of a 1 stage repair. However, 5%–10% of patients coming for AVSD surgery will require PA debanding.[7]

Common AV canal defects (CAVSD) can be divided into[1] complete form characterized by a common AV valve and an unrestricted ventricular septal communication and[2] transitional or intermediate form charcterized by two separate AV valve orifices and a restrictive ventricular septal defect. Both forms in addition, have an ostium primum atrial septal communication.

Repair of CAVSD in early infancy conventionally carried greater morbidity and mortality than repair performed later in life however improved surgical techniques myocardial protection, and postoperative care has led to a progressive reduction in operative mortality to <3%.

The historical reason for avoiding early repair was linked to the difficulties in the repair of flimsy AV valve leaflets in small children, however, due to large left to right shunting (after 4–6 weeks of life), such infants experience cardiac symptoms which are not always controlled with anti-congestive treatment. When associated with Down's syndrome they have repeated chest infections, which further worsens abnormal pulmonary vascular resistance. Variables associated with increased hospital mortality include longer CPB and aortic cross-clamp time, the presence of LAVV dysplasia is linked to worse preoperative function; more often needing additional measures for repair of LAVV and a higher incidence of later reoperation for

LAVV dysfunction. Avoidance of LAVV cleft closure and partial cleft closure is significantly associated with higher LAV reoperation rate. Partial closure is needed only if the posterior/mural LAVV leaflet is hypoplastic to avoid stenosis of LAVV.

Unbalanced CAVSD defined as a CAVSD with an AV valve override of more than 60% over either ventricle or the presence of hypoplastic nonapex forming ventricles or in case of a right dominant CAVSD a left ventricle with indexed volumes at least 2 SD smaller than normal, represent 10%–15% of all AVSD. This defect is characterized by underdevelopment of one of the ventricles and varying degrees of malalignment of the common AV valve, over the hypoplastic ventricle and associated hypoplasia of the outflow valve related to decreased flow.[8] Management strategy includes single ventricle (SV) palliation, and primary or staged biventricular SV palliation, and primary or staged biventricular repair. More recently, biventricular conversion from SV palliation has been advocated, particularly in patients with trisomy 21 and heterotaxy, who tolerate SV palliation poorly. Staged biventricular recruitment has also been advocated for complete biventricular repair.

Deciding how to manage this complex group of patients requires assessment of clinical imaging and hemodynamic data.[9] Ventricular volumes determined by three dimensional echocardiography[10] and magnetic resonance imaging scans may guide therapy with the ability to recruit ventricles with volume as low as 15–30 ml/m^2.

In a study by St. Louis *et al*.[11] using the Society Thoracic Surgeons (STS), congenital heart surgery database analyzing the results of surgical correction of 2399 children from 101 centers from 2008 to 2011. Of these children, 78% had Down's syndrome. Median age at surgery was 4.6 months with 11.8% aged <2.5 months. PA band removal was performed in 4.6% of patients at surgical repair.

Major complications occurred in 9.8% including permanent pacemaker implantation in 2.7%. Overall mortality was 3%. Weight <3.5 kg and age <2.5 months were associated with higher mortality, longer postoperative length of stay in the Intensive Care Unit (ICU) and higher incidence of major complications. Surprisingly, children with Down's syndrome had lower morbidity and mortality than other children, and their duration of ICU stay was similar.

The natural history of complete AVSD includes premature death due to complications of congestive heart failure and or PA hypertension. Repair during infancy is recommended for all children. The age at elective repair has steadily declined from 1 year a few decades ago to 3–6 months at most centers today. Early repair minimizes the risk of premature death or pulmonary vascular obstructive disease. CAVSD is frequently associated with Down's syndrome. Infants with Down's syndrome and L-R shunts have long been believed to be susceptible to pulmonary arterial hypertension and respiratory complications, however analysis of STS-congenital heart surgery database, multi-institutional data revealed similar mortality rates for children with or without Down's syndrome, across various spectrum of pediatric cardiac surgical procedures, length of ICU stay was prolonged in some specific procedures but not in Down's with CAVSD repair.[12]

Xie *et al*. from the Royal children's Hospital Melbourne[13] presented data from 2000 to 2011 including 138 patients with CAVSD using the 2 patch technique in 92% and one patch in 2.2% their operative mortality was 1.4% and overall mortality was 5.8%. Freedom from reoperation was 84.3% at 8 years. Age >6 months at repair was associated with higher rates of reoperation, however, operating at age <6 months was associated with longer ICU stay. Moderate AVV regurgitation at discharge was a risk factor for reoperation. Down's syndrome present in 75% of patients was not a risk of a factor for higher mortality, morbidity, reoperations, ICU stay or duration of ventilation.

Pilchard *et al*.[14] in their excellent review on the perioperative and intensive care management of children with complete AVSD have highlighted some key topics. (a) Down's syndrome: up to 50% to 70% of children with AVSD may have Down's syndrome, also having a high incidence of extracardiac anomalies including abnormalities of the CNS, hypotonia, seizures, gut abnormalities, thyroid disorders, and airway abnormalities such as tracheal stenosis and laryngotracheomalacia, also there may be sedation issues, pulmonary arterial hypertension, and vascular access concerns, higher right-sided cardiac pressures and pulmonary vascular resistance. (b) Arrhythmias: There is a 72% incidence of postoperative arrhythmias most commonly junctional ectopic tachycardia, followed by complete AV block, ventricular tachycardia and reentrant SVT and nonsustained SVT. At present, intravenous amiodarone is the recommended pharmacological treatment for most postoperative tachyarrhythmias to control the rate and restore AV synchrony. (c) Pulmonary arterial hypertension: occurring in approximately 2% of children with hypertensive crisis occurring in 0.75%. This results in prolonged mechanical ventilation and increased the length of ICU stay and mortality of 20%. The incidence has reduced after the introduction of nitric oxide and earlier age at surgery. PA catheters may be required in children with PA pressures 60% of systemic or if nitric oxide was required for separation from the bypass. Management would include pulmonary vasodilators (nitric oxide) sedation and analgesia, judicious use of PEEP to prevent atelectasis, and sodium bicarbonate to buffer acidosis. Rebound pulmonary hypertension after stopping nitric oxide requires sildenafil; (d) Thyroid hormone: hypothyroidism is common in children with congenital heart disease and in Down's is as common as 7% to 50% requiring routine screening. CPB also suppresses thyroid hormone production, causing

decreased myocardial contractility and cardiac output. (e) Sedation and analgesia: especially in Down's syndrome due to abnormal opioid receptor concentration in cerebral cortex along with differences in nociception, anxiety levels, and neurotransmission. Dexmedetomidine and caudal analgesia offer promising therapy, with dexmedetomidine infusion, having additional antiarrhythmic effects.

This issue of the annals has an excellent article from Germany on the perioperative management of this difficult subset of children with complete AVSD. The authors Janai *et al*.[15] and their team retrospectively studied 157 children with complete AVSD who underwent surgical correction from 1999 to 2013 with AVSD repair. Mean age at surgery was 125 ± 56.9 days. Nearly 63.6% of these children had Down's syndrome. Although the authors have followed routine anesthetic, CPB and postoperative management principles as would be followed today across the world in most pediatric cardiac centers, their series is astonishing because of the 0% mortality reported by them compared to the 3%–5% reported in most of the other series[5-7,11] the main reasons probably are the early age at surgery, overall team experience, protocolized management, use of transesophageal echocardiography both intra- and postoperatively for management. No doubt these are results the rest of the world should aspire for.

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