AVSD
Postoperative Management & Complications

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Complex AVSD

- **AVSD in heterotaxy syndrome**
- **Unbalanced AVSD**
- **AVSD with cono-truncal anomalies**
  - AVSD with TOF
  - AVSD with DORV + Pulmonary Stenosis
  - AVSD with Truncus Arteriosus (TA)
  - AVSD with TGA
- **AVSD with LVOTO**
- **AVV abnormalities**

Postoperative Management

Generally similar to that in all patients undergoing corrective repair of CHDs

Postoperative Management
Possible postoperative problems

- **Mortality**
- **Low CO syndrome**
- **Arrhythmia: m/c JET, followed by complete AVB, VT, and re-entrant SVT (Delaney JW, 2006)**
- **PH**
- **Residual or recurrent lesions**
  - Left AVV regurgitation/stenosis, residual ASD/VSD, LVOTO
- **Coronary artery problem, rare & mainly LCX**

Echocardiography to evaluate ventricle function, VSD/ASD leakage, AVVR, AVV stenosis, LVOTO if any hemodynamic problem

Postoperative Management
PH

- PH was noticed in 60% of patients who underwent C-AVSD repair during infancy and 30% needed NO therapy. (Janai AR, 2018)
- Especially, in patients who underwent AVSD repair at a later age (>6-12 M) & have Down syndromes
- When PH is identified postoperatively, it is important to rule out secondary causes of PH such as severe MR, MS, or residual VSD.
- For children with Down syndrome, the development of PH is multifactorial and other non-cardiac issues likely contribute to the risk (eg, obstructive sleep apnea, hypoventilation, recurrent aspiration in addition to genetic predisposition).

Complications During FU

Complications During FU

- **Late mortality**
- **PH, persistent or recurrent**
  - Surgical repair in early infancy significantly reduces the risk of developing PVD; however, it does not eliminate it.
- **Arrhythmia including AV block**
  - Reported incidence of PPM implantation ranges from 1.5% to 2.7%. (Jacobs JP, 2010; Janai AR, 2018; St. Louis JD, 2014)
- **AVVR, usually left AVVR (MR)**
- **Left AVV stenosis (MS)**
- **LVOTO**
- **Residual VSD or ASD**

Mortality

- **Operative mortality**
  - Improved surgical techniques and postoperative care has led to a progressive reduction in operative mortality to <3%. (Chauhan S, 2018)
- **Overall mortality: depends on FU duration**
  - less than 10% at 10 years (Stulak JM, 2010; Pontailler M, 2014; Ginde S, 2015)
- **For children with partial and transitional AVSDs, surgical outcomes are excellent, with minimal perioperative mortality (<1%) and with long-term survival that is similar to that of the general population. (Minich LL, 2010; Devlin PJ, 2016; Buratto E, 2015)**

Mortality & Risk Factors

- **Major risk factors for early mortality include**
  - young age at time of repair (age <2.5 months, weight <3.5 kg)
  - presence of associated cardiac defects
  - STS database showed children with Down's syndrome had lower morbidity and mortality than other children (St Louis JD, 2014). But in other reports, there was no difference in mortality between Down and non-Down syndrome patients (Masuda M, 2005; Lange R, 2007).

Reoperations after AVSD Repair

- In several large series, reoperation after AVSD repair was required in 15-20% of patients at 10 to 20 years. (Birim O, 2009; Stulak JM, 2010; Pontailler M, 2014; Ginde S, 2015)

Complications During FU
Left AVVR

- **Left AVVR is the leading reason for reoperation and was necessary in 10-15% of patients (Minich LL, 2010; Bové T, 2018; Ten Harkel AD, 2005).**
- **Mechanism of left AVVR: non- or partial cleft closure, degeneration of valve tissue, annular dilatation, and suture dehiscence between the leaflets and the patch (Prifti E, 2013)**

Complications During FU
Risk Factors for Reoperation - LAVVR

- **Age at the operation <3 months**
- **Unfavorable morphology of the LAVV**
- **Associated cardiac malformations**
- **Non-Down syndrome**
- **Absence of cleft closure**
- **Moderate or more LAVVR on postop echo**
  - One third of patients with significant late LAVVR had no significant early postoperative regurgitation (Ho DY, 2020)
- **Particular AVSD type did not appear to be a risk factor for mortality or LAVVP failure.**

Formigari R, 2004; Hoohenkerk GJF, 2010; Pontailler M, 2014; Sojak V, 2016; Krupickova S, 2018; Schleiger A, 2019; Fong LS, 2021

Complications During FU
Redo for LAVVR

- **Re-intervention rates for the 2nd LAVV repairs after primary AVSD surgery remain high (approximately 1/3 of the patients over a median FU of 12 months), particularly for patients with LAVV stenosis gradient ≥5 mm Hg and mild or greater LAVV regurgitation on postoperative TEE. (Gellis L, 2023)**
- **LAVV valvuloplasty is superior to LAVV replacement.**
  - preserved somatic growth, and absence of hemorrhagic, thromboembolic, and infectious complications
  - But durability of repair is weakest point, especially in children with complex LAVV pathology. (Chauhan S, 2018)

Complications During FU
Left AVV stenosis (MS)

- Left AVV stenosis can result from excessive suture closure of cleft, congenital anomaly of AVV (small LAVV orifice or subvalvar abnormality) or inappropriate surgical division of common AVV between two ventricles.
- Careful assessment of valve morphology prior to surgery and avoiding aggressive cleft closure in cases with small left mural leaflet decreases the incidence of postoperative MS.
- Reoperation for AVV stenosis or inflow obstruction usually requires valve replacement. (Robinson JD, 2009)

Complications During FU
LVOTO - Incidence

- Relief of LVOTO is the second most common reason for reoperation after initial repair of partial and complete AVSD, occurring in 2-7% of patients (Pontailler M, 2015; Mery CM, 2019; Bové T, 2018; IJsselhof R, 2017).

Complications During FU
LVOTO - Mechanism

- Mechanism: LVOTO may develop from combination of anatomic characteristics of AVSD, including elongation and narrowing of the LVOT and crowding of the LVOT by AVV tissue that may be accentuated over time by patient growth or development of subaortic membrane, septal hypertrophy and prominent anterolateral muscle bundle.
- Tight adherence of SBL to septal crest causes LVOT to be longer and narrower. LVOTO is more common in P-AVSD and Rastelli type A of C-AVSD.
- Myers et al analyzed the characteristics at initial repair (Down syndrome, age at repair, AVSD type, and repair techniques) and at reoperation for LVOTO (age at reoperation, mechanisms of LVOT obstruction, and reoperation repair technique) in 56 patients but only modified single-patch technique was identified as a predictive factor for reoperation for LVOTO (P= 0.04) (Myers PO, 2012).

SMC Data
Demographic data

- **216 op in 182 AVSD patients (M/F 63/119), 2000-2023**
- **Down 34**
  - C-AVSD 27/61, Intermediate & Partial AVSD 7/121
- **C-AVSD 62**
  - Age at total repair median 138 ± 537 (37*-138) days
  - MPA banding w/wo PDA ligation in 10 (complete)
- **Partial or intermediate AVSD 120**
  - Age at total repair median 4.2 ± 21.9 Y (10 days** – 81Y)

*** Rastelli A, severe MR ⇒ redo-MV repair 3Y later**

**** P-AVSD with RPA interruption**

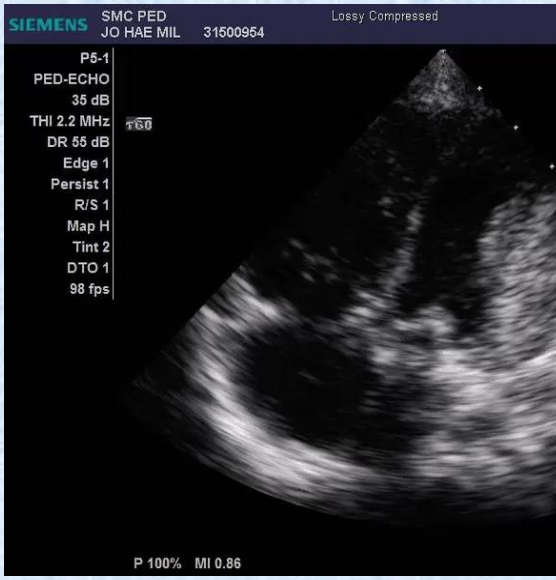
SMC data
Mortality

- **3 op mortality (3/182)**
  - 2 low CO & V-dysfunction, on ECMO (complete)
  - 1 massive bleeding (partial)
- **2 late deaths**
  - 1 brain hemorrhage - MVR on warfarin, h/o acute myocarditis
  - 1 sudden death - LV dysfunction d/t LCx damage, PPM d/t SSS in Lt isomerism

SMC Data
Reoperations

- **21 reoperations in 18 patients after total repair**
  - Twice for MR in 1, three times for MR in 1
  - Partial 11/120, Complete 7/62
- **9 additional reoperations after total repair at outside hospitals (partial 4, complete 5)**

SMC Data
Indications for reoperation (N= 27)



- **LAVV dysfunction 17**
  - MR 7 + 5
  - MR & TR 1 + 0
  - MSR 1 + 0
  - MS 2 + 1
- **VSD, MR, TR 1* + 0 ⇒ redo-CAVSD**
- **ASD, MR, TR 0 + 1 ⇒ redo-PAVSD**
- **ASD 1 + 0 ⇒ ASD closure**
- **VSD 1** + 0 ⇒ VSD closure**
- **LVOTO 2 (both PAVSD) + 2 (both CAVSD of unknown type) ⇒ relieve LVOTO**
- **Complete AVB 1 (& + MR repair 2) ⇒ PPM insertion**
- **LV hematoma 1 ⇒ hematoma evacuation**

*** modified one patch technique, ** two patch technique**

SMC Data
Left AVV problems (N= 19)

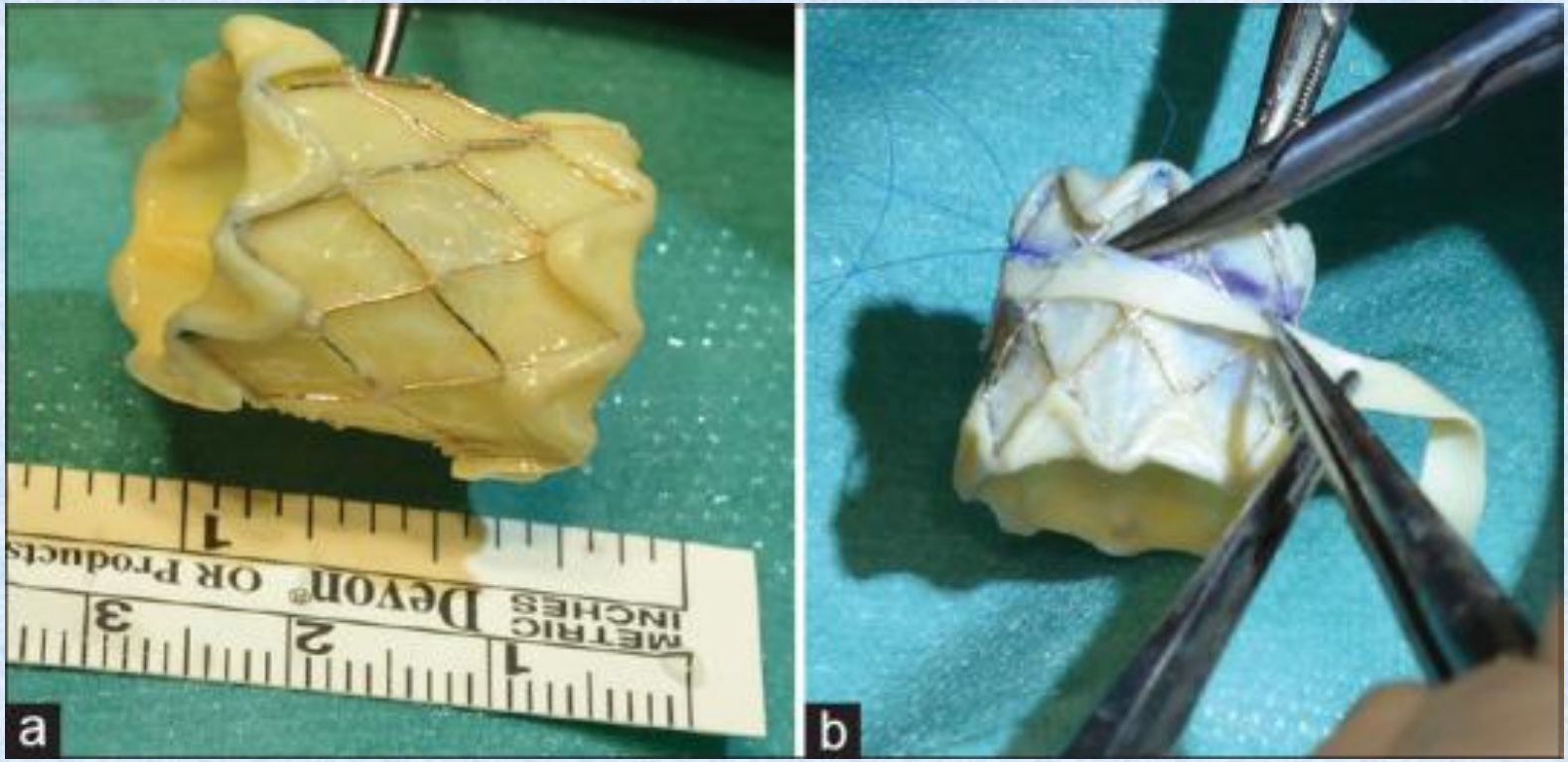
▪ MR (& TR1)	13	⇒ MVr 9, MVR 4*
▪ MSR & TR	1	⇒ MVR & TVR 1
▪ MS	3	⇒ MVR in all
▪ VSD, MR, TR	1	⇒ redo-CAVSD
▪ ASD, MR, TR	1	⇒ redo-PAVSD

- 3 times of reoperations for LAVVR

Repeated reoperation case

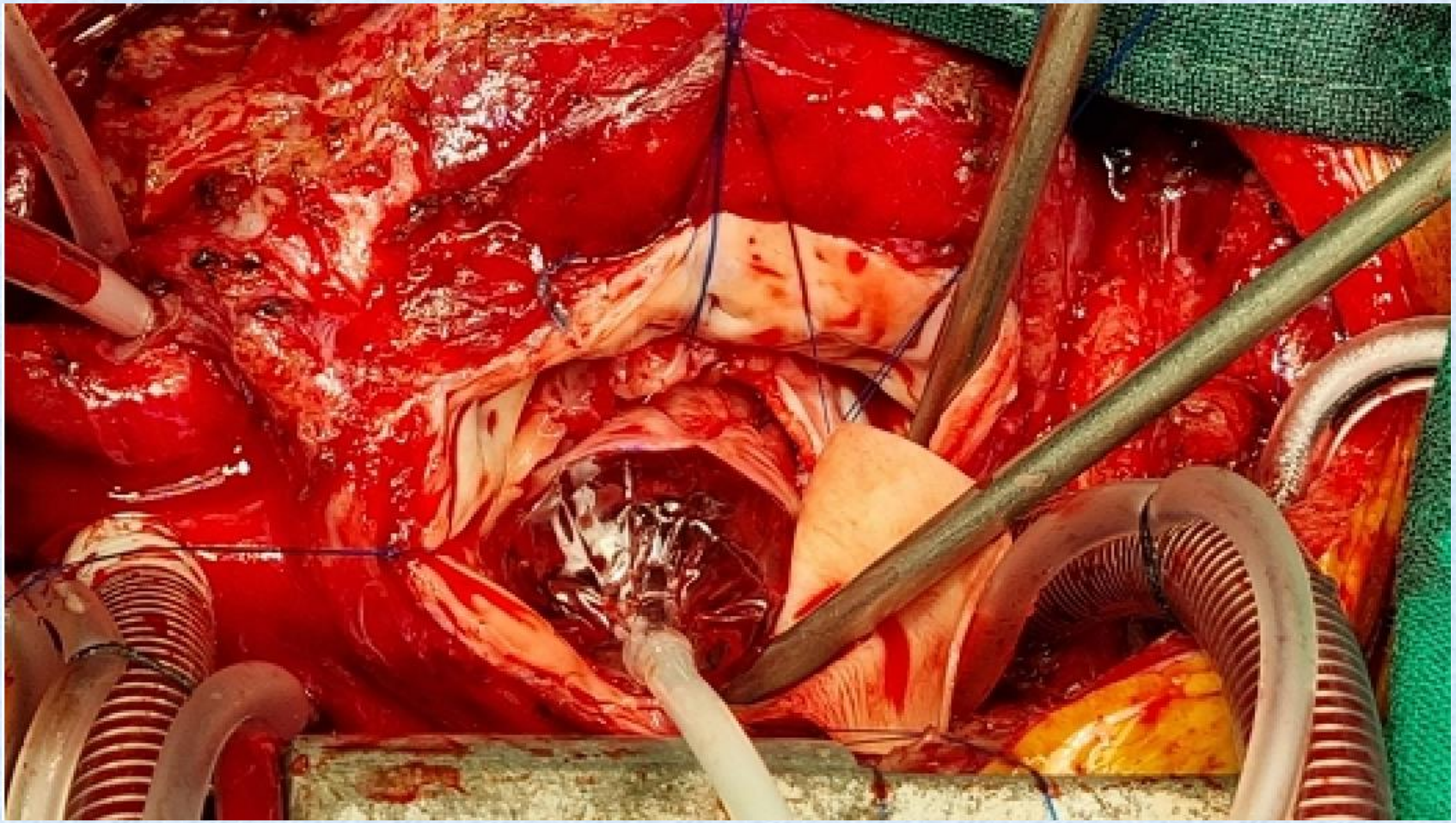
- 17 Mo/F now
- Presented with dyspnea, feeding difficulty at 2Mo of age
- s/p C-AVSD at 3Mo of age (modified one patch)
- 3 weeks later LAVV repair d/t severe LAVVR
- MVR with SJM AV 19 mm at 4 Mo of age (supra-annular position)
- Recurrent brain hemorrhage at usual PT INR (2.0-2.5)
- Mechanical MV malfunction during dose reduction
- MVR with Melody TPV 18 mm at 10 Mo of age

Repeated reoperation case
Melody Valve in Mitral Position

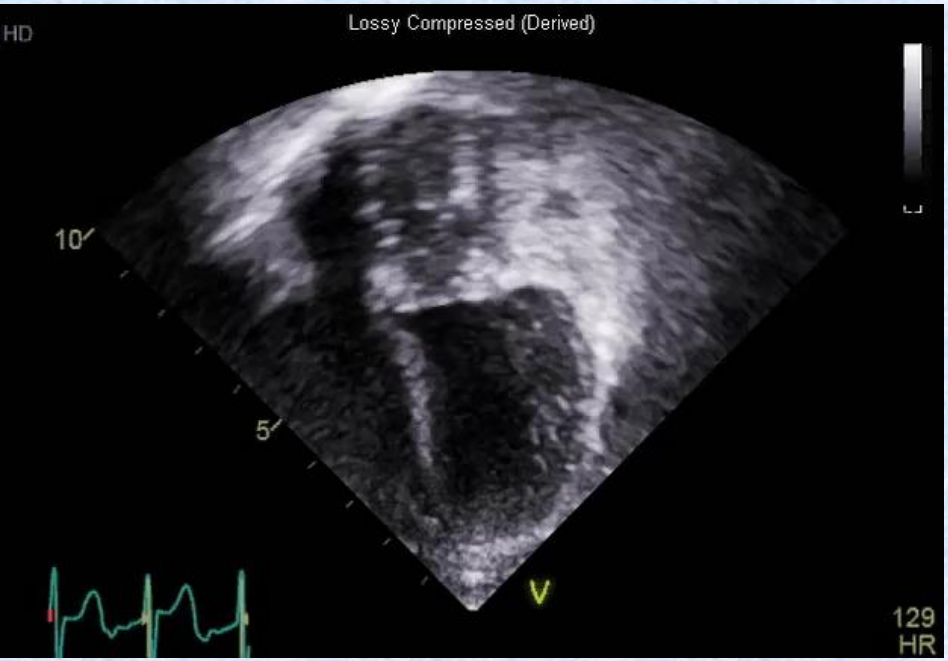


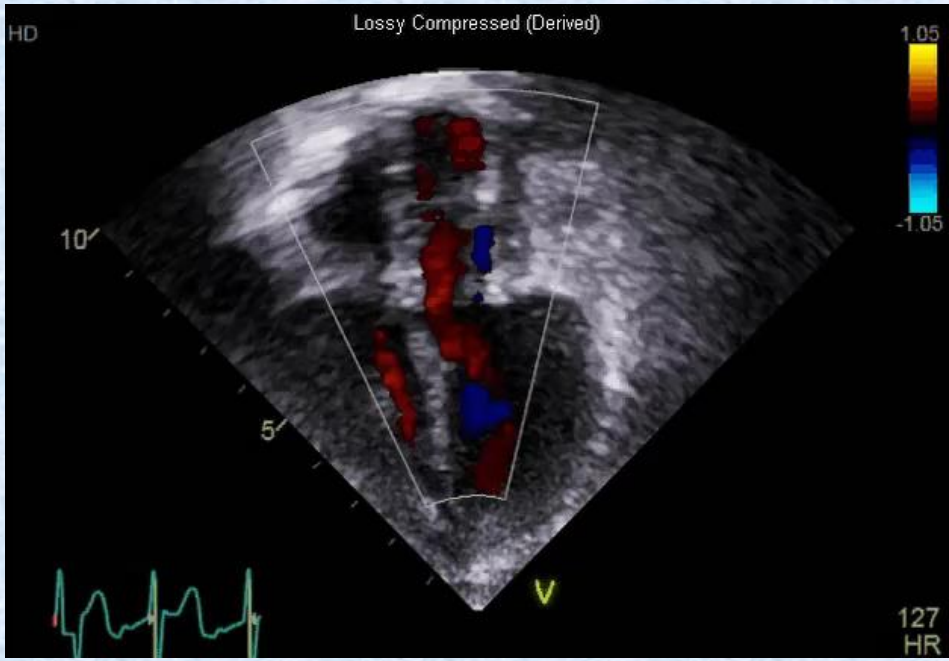
(a) Foreshortened Melody valve by turning back the struts. (b) Creation of a neo-sewing ring at the junction of proximal 2/3rd and distal 1/3rd of Melody® valve. (Dranseika V, 2021)

Repeated reoperation case
Melody Valve in Mitral Position

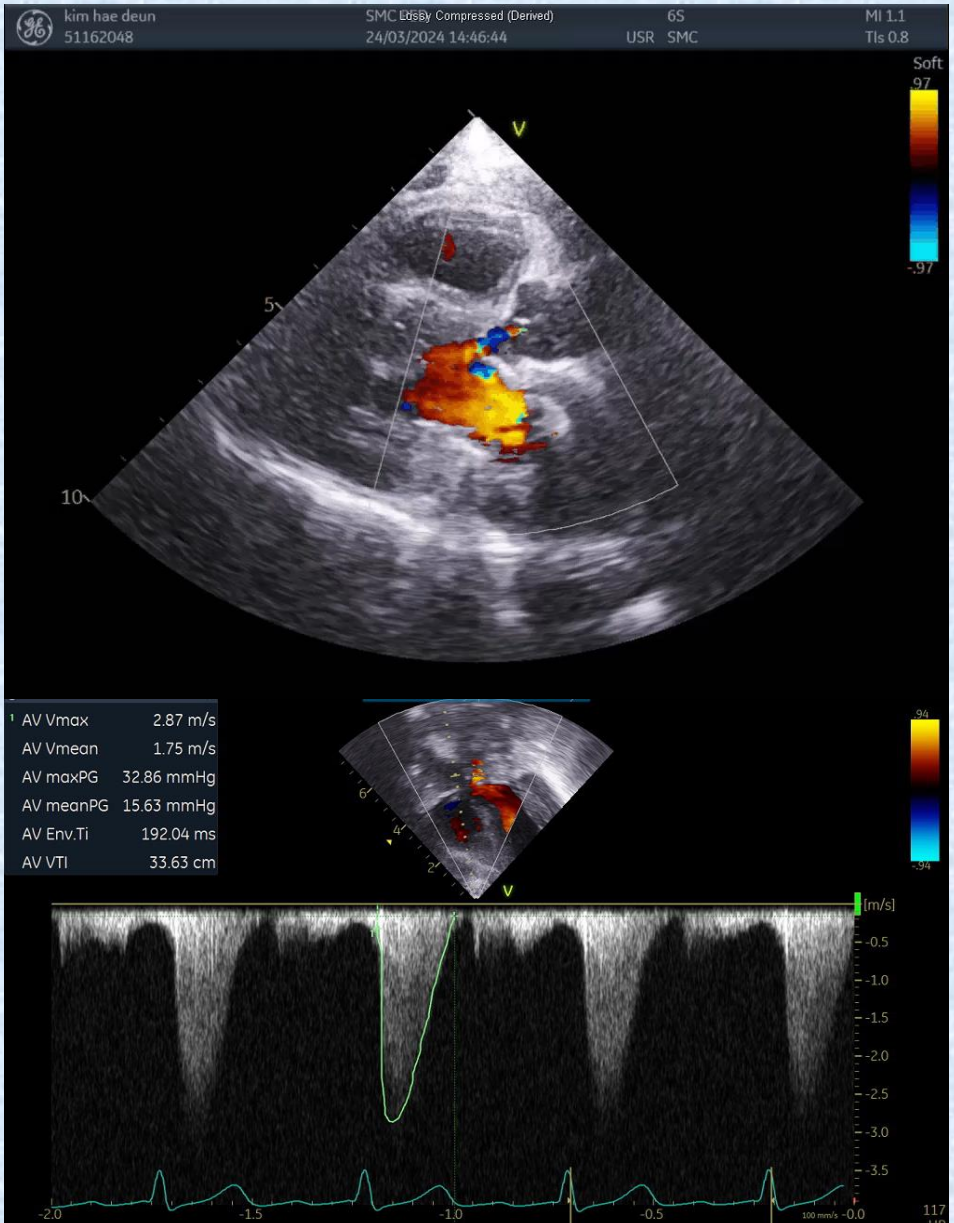


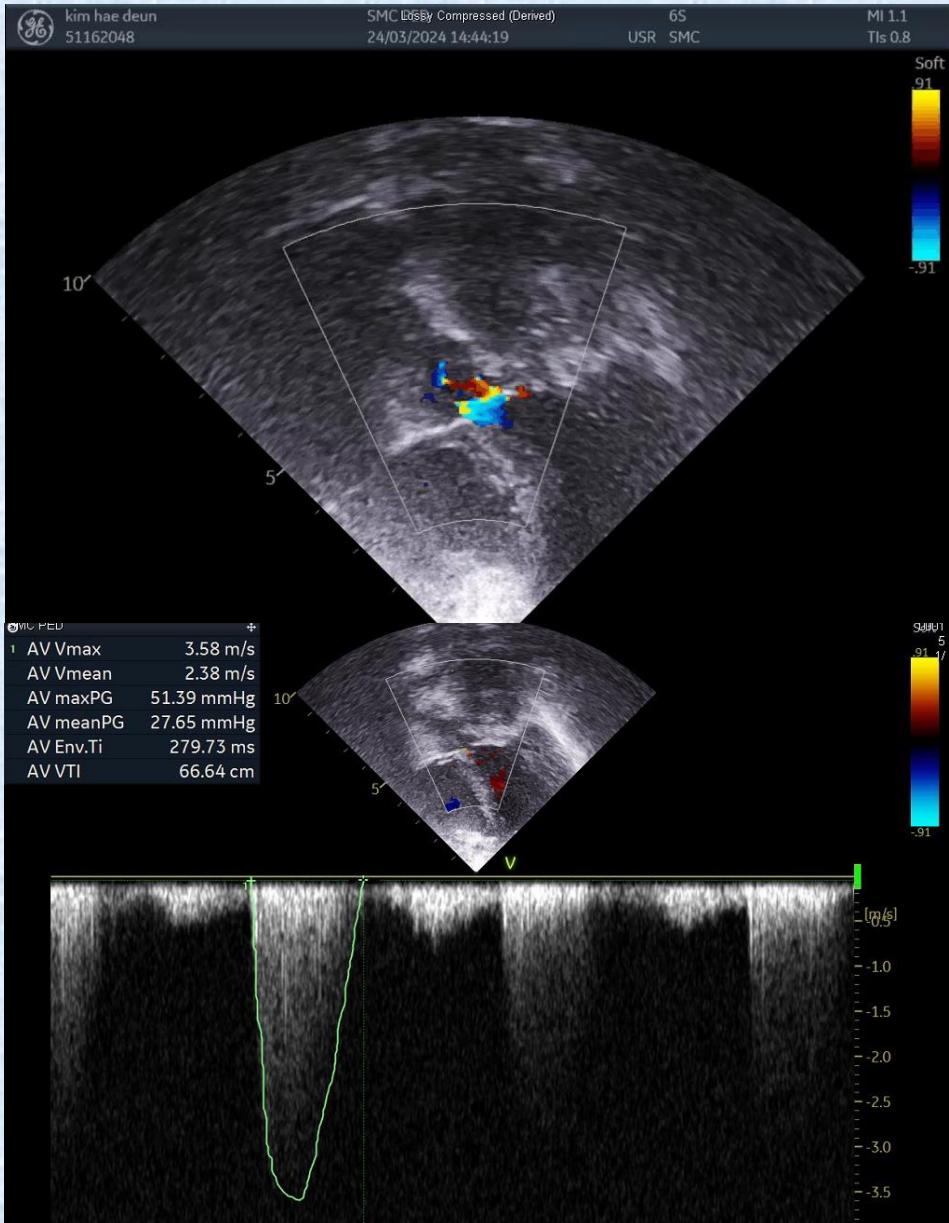
Repeated reoperation case
Melody Valve in Mitral Position



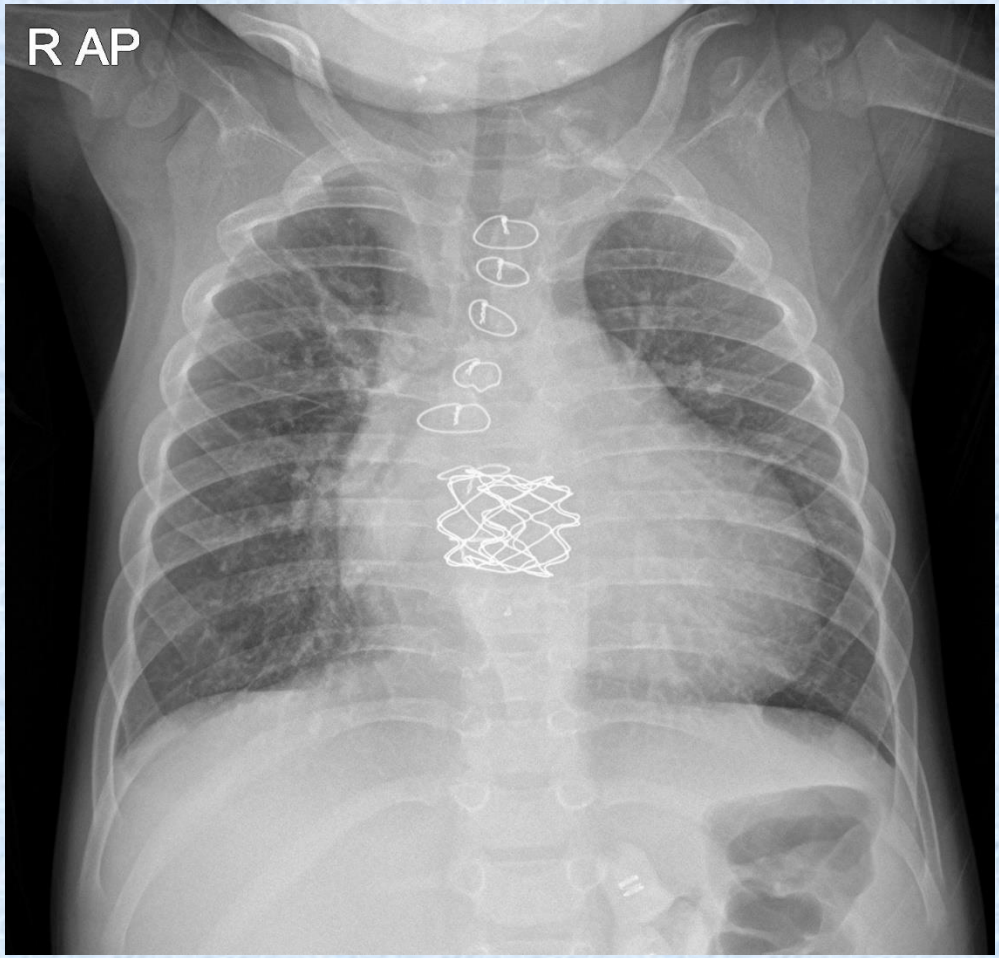


Repeated reoperation case
Melody Valve in Mitral Position





Repeated reoperation case
Melody Valve in Mitral Position



Summary

- Recently AVSD repair can be done with low mortality but reoperation is not uncommon.
- LAVVR is the most common issue during FU. Although LAVV repair is ideal, LAVV replacement is inevitable in some cases. In these cases, Melody valve can be an option in infants or small children.
- Reoperation for LAVV stenosis usually requires valve replacement.
- All AVSD patients should be followed after total repair to monitor for AVVR, arrhythmias, LVOTO, PH, and ventricular dysfunction.


Thank you
for your attention

SMC Data
Associated cardiac problems

- Arrhythmia: A-fib in 3, complete AVB 2 (all in P-AVSD)
- Severe PH 5: VSD/ASD partial closure 4 in C-AVSD,
  ASD partial closure 1 in P-AVSD
- Unroofed CS 9 (2 complete, 7 partial/intermediate)
- CS osteal atresia 2 (P-AVSD)
- PS/PPS 4 (1 in complete, 3 in partial)
- Situs inversus 1 (partial)
- Left isomerism with IVC interruption 3 (partial)
- PAPVR 1 (partial)
- Vascular ring 1
- CABG 1 (paritial)
- Infective endocarditis with vegetation 1 (paritial)

Postoperative Management
PH

- **Avoid precipitating or aggravating factors**
  - Avoid hypoxia, acidosis, and hypercarbia (paO2 <60 mmHg, pH <7.35, and paCO2 >40 mmHg). (Rudolph AM, 1966)
  - Use analgesics, sedatives (dexmedetomidine) & paralyze patients in the immediate postoperative period
  - Ventilator maneuvers include judicious use of PEEP to prevent atelectasis and V/Q mismatch, increase of FiO2, lowering of PCO2 (25-30 mm Hg) but not too much hyperventilation.
  - The use of supplemental sodium bicarbonate to buffer acidosis, as opposed to hyperventilation, is preferred to avoid decreased cerebral blood flow in the post-bypass patient.

Outcome from STS CHS Database

- Society Thoracic Surgeons (STS), congenital heart surgery database analyzing the results of surgical correction of 2399 children from 101 centers from 2008 to 2011 (St Louis JD, 2014)
- Median age at surgery was 4.6 months with 11.8% aged <2.5 months. PAB removal was performed in 4.6% of patients at surgical repair.
- Overall mortality of more than 3%.
- Major complications occurred in 9.8% including permanent pacemaker implantation in 2.7%.
- Weight <3.5 kg and age <2.5 months were associated with higher mortality, longer postoperative length of stay in the ICU and higher incidence of major complications. Children with Down's syndrome had lower morbidity and mortality than other children, and their duration of ICU stay was similar.

Complications During FU
Arrhythmia

- Late postoperative arrhythmias were less common, occurring in 2-3% of patients. Bradyarrhythmias, particularly complete heart block, are the most common long-term rhythm problems following surgical repair of AVSDs, with approximately 3-4% of patients requiring permanent pacemaker implantation (Kharbanda RK, 2018; Backer CL, 1995; St Louis JD, 2014; Di Mambro C, 2018).
- Some authors hypothesize a difference in the need for pacemaker implantation between single-patch and double-patch techniques; however, this difference could not be significantly proven. (Li D, 2017; Pan G, 2014; Wu Y, 2020)

Staged repair for C-AVSD in patients weighing less than 4.0 kg

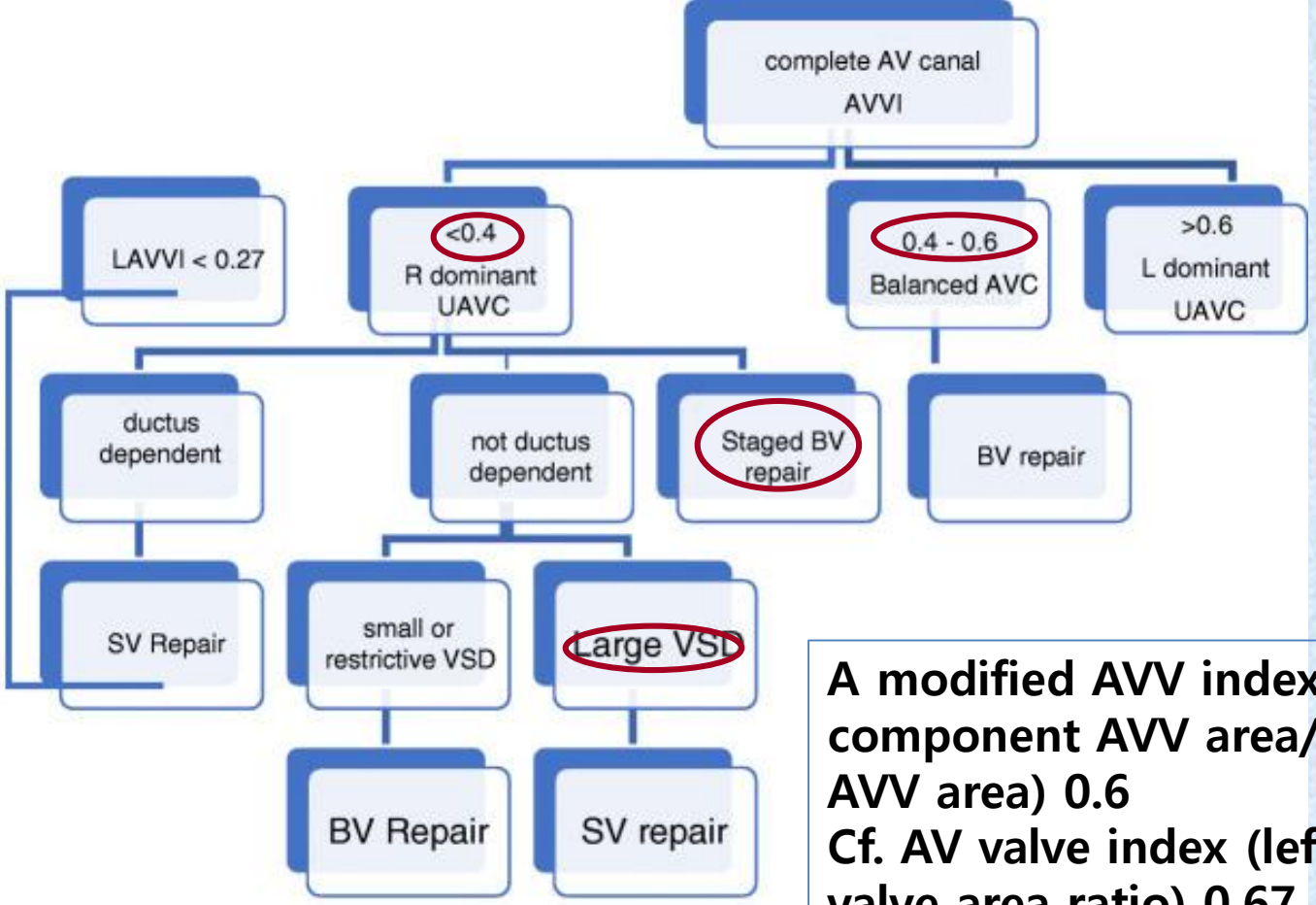
- Kobayashi Y, et al. The Journal of Thoracic and Cardiovascular Surgery 2024
- https://doi.org/10.1016/j.jtcvs.2023.07.003
- This study compared the mortality, left AVV–related reoperation, and left AVV competence in symptomatic neonates and small infants who underwent staged repair incorporating PAB or primary repair for C-AVSD.
- Methods: Patients weighing less than 4.0 kg at the time of undergoing staged (n = 37) or primary (n = 23) repair for balanced C-AVSD between 1999 and 2022 were reviewed. The mean FU period was 9.1 years. Freedom from moderate or greater left AVVR was estimated with the Kaplan–Meier method.
- Results: The staged group included smaller children (median weight, 2.9 vs 3.7 kg) and a higher proportion of neonates (41% vs 4%). All patients in the staged group survived PAB and underwent intracardiac repair (median weight, 6.8 kg). After PAB, the severity of left AVVR improved in 10 of 12 patients (83%) without left AVV anomaly who had mild or greater left AVVR and a left AVV Z score greater than 0. Although survival and freedom from left AVV–related reoperation at 15 years (P = .195 and .602, respectively) were comparable between the groups, freedom from moderate or greater left AVVR at 15 years was higher in the staged group (P = .026).
- Conclusions: Compared with primary repair, staged repair for C-AVSD in children weighing less than 4.0 kg resulted in comparable survival and reoperation rates and better left AVV competence. PAB may mitigate secondary left AVVR unless a structural valve abnormality exists. Selective deferred intracardiac repair beyond the neonatal and small-infancy period may still play an important role in low-weight patients.

Complications During FU
Redo for LAVVR

- Prosthetic valve implantation in a severely hypoplastic annulus, as seen in small children, poses a surgical challenge. High profile tissue prostheses have a potential for LVOTO, poor hemodynamic performance in small sizes, and rapid degeneration. Low profile mechanical prostheses represent the best option. In cases with severe annular hypoplasia, a supraannular placement may be the only option (Kadoba K, 1990). However, in the patient with AVSD, the annulus may be enlarged without injuring fibromuscular tissue separating the right and left AV valves by augmenting the VSD patch. A drawback associated with this procedure may be the potential development of tricuspid insufficiency because of concomitant annular enlargement. (Ando M, 2001)

Unbalanced AVSD

- However, in a multicenter study of 257 infants with CAVC, these echocardiographic measures of unbalance correlated poorly with one another, suggesting that they likely assess distinct morphologic and functional parameters (Meza JM, 2019).



A modified AVV index (left component AVV area/total AVV area) 0.6
Cf. AV valve index (left/right valve area ratio) 0.67

Staged repair consists of
Shifting the patch towards the Right, borrowing from the tricuspid
Partitioning of the Tricuspid to create second MV
Repair of any anomalies in MV
Leave behind a Restrictive ASD
Restricting a large ASD leaving VSD open
Leave behind a restrictive VSD

The approach for unbalanced AVCD with small LV. Staged repair consists of shifting the patch towards the right, borrowing from the tricuspid, partitioning of the tricuspid to create second MV, repair of any anomalies in MV, leave behind a restrictive ASD, restricting a large ASD leaving VSD open, and leave behind a restrictive VSD. (Jagannath BR, 2021)

FU after AVSD Repair
Endocarditis prophylaxis

- **For patients undergoing procedures likely to result in bacteremia (eg, dental work) within six months of complete repair, prophylactic antibiotics are recommended for prevention of endocarditis.**
- **After six months, antibiotics are recommended at the time of such procedures**
  - if the patient had a repair involving use of prosthetic material for valve repair,
  - if there is a residual shunt or valvular regurgitation at the site or adjacent to the site of the prosthetic patch or prosthetic device,
  - if the patient has had a prior episode of endocarditis.

Postoperative Management
Low cardiac output syndrome

- LCO was defined by ScvO2 ≤60%, arteriovenous oxygen saturation difference ≥30%, urine output <1 mL/kg/h, metabolic acidosis with base excess ≥-4 mmol/L, serial elevation of serum lactate levels ≥2 mmol/L in two consecutive blood gas analyses, and echocardiographic evidence of ventricular dysfunction. (Bailey JM, 2004)
- Prophylactic milrinone (0.3-0.5 μg/kg/min) with/without dobutamine to reduce incidence of LCO after CHD surgeries. (Vogt W, 2011)
- Delayed chest closure might have helped in lowering the incidence of LCO (Shalabi RI, 2002) but prolonging the duration of ventilation (>24 h). (Janai AR, 2018)

Postoperative Management
Ventilatory Management

- **Preoperatively it is aiming at avoiding pulmonary overflow.**
  - This can be achieved with normo-ventilation (paCO2 40-45 mmHg) with minimum FiO2. In patients with the L-to-R shunts, permissive hypercapnia and SaO2 around 90% prevent excessive PBF maintaining adequate tissue oxygenation. (Shekerdemian L, 1999)
- **After total repair, ventilator management is focused to reduce PVR and to prevent PAH crisis.**
  - Avoid hypoxia, acidosis, and hypercarbia (PaO2 <60 mmHg, pH <7.35, and paCO2 >40 mmHg). (Rudolph AM, 1966)
- **Ventilatory weaning can be started with reduction of FiO2 to 0.6 maintaining Horowitz index ≥200. This is followed by rapid tapering of NO from 20 to 10 ppm, slower tapering to 5 ppm, and thereafter at rate of 1 ppm every 2 h to avoid rebound PAH. With signs of rebound PAH, FiO2 can be increased to >0.6 and NO to 20 ppm and oral sildenafil is started (0.5–1 mg/kg every 6 h). Sildenafil was given intravenously (0.2–0.3 mg/kg as continuous infusion over 20 min every 6 h) where oral absorption could not be guaranteed. (Janai AR, 2018)**
- **Extubation can be done when Horowitz index (PaO2/FiO2) is >200, pCO2 between 35 and 40 mmHg, and ScvO2 >70%, with minimum catecholamine therapy (monotherapy with dobutamine ≤5 µg/kg/min or milrinone 0.3–0.5 µg/kg/min). (Janai AR, 2018)**

AVSD & Down syndrome

- **Up to 50% to 70% of children with AVSD may have Down's syndrome. (Pilchard J, 2010)**
- **Hypothyroidism, GI obstruction**
- **Risk of PH**
  - In addition to hemodynamic effect of AVSD & genetic predisposition, airway abnormalities (such as tracheal stenosis, laryngotracheomalacia), obstructive sleep apnea, hypoventilation, recurrent aspiration
- **Respiratory complications such as stridor, atelectasis, pneumonia were more often in Down syndrome patients (69%). (Ihringer K, 2013; Janai AR, 2018)**