

# Retrospective Study of Complete Atrioventricular Canal Defects: Anesthetic and Perioperative Challenges

## Abstract

**Objective:** The objective of this study was to highlight anesthetic and perioperative management and the outcomes of infants with complete atrioventricular (AV) canal defects. **Design:** This retrospective descriptive study included children who underwent staged and primary biventricular repair for complete AV canal defects from 1999 to 2013. **Setting:** A single-center study at a university affiliated heart center. **Participants:** One hundred and fifty-seven patients with a mean age at surgery of  $125 \pm 56.9$  days were included in the study. About 63.6% of them were diagnosed as Down syndrome. Mean body weight at surgery was  $5.6 \pm 6.3$  kg. **Methods:** Primary and staged biventricular repair of complete AV canal defects. Measurements and main results: A predefined protocol including timing of surgery, management of induction and maintenance of anesthesia, cardiopulmonary bypass, and perioperative intensive care treatment was used throughout the study. Demographic data as well as intraoperative and perioperative Intensive Care Unit (ICU) data, such as length of stay in ICU, total duration of ventilation including reintubations, and total length of stay in hospital and in hospital mortality, were collected from the clinical information system. Pulmonary hypertension was noted in 60% of patients from which 30% needed nitric oxide therapy. Nearly 2.5% of patients needed permanent pacemaker implantation. Thorax was closed secondarily in 7% of patients. In 3.8% of patients, reoperations due to residual defects were undertaken. Duration of hospital stay was  $14.5 \pm 4.7$  days. The in-hospital mortality was 0%. **Conclusion:** Protocolized perioperative management leads to excellent outcome in AV canal defect repair surgery.

**Keywords:** Atrioventricular canal defect, congenital cardiac anesthesia, congenital cardiac surgery, mortality, pulmonary hypertension

## Introduction

The prevalence of atrioventricular (AV) canal defects is 2.9% of all congenital heart defects in Germany.<sup>[1]</sup> Complete AV canal defects account for 77% of them.<sup>[2]</sup> Perioperative mortality for correction of complete AV canal defects is 3.4%.<sup>[3]</sup> The aim of this retrospective study was to assess perioperative morbidity and mortality in patients undergoing repair of complete AV canal defects.

## Materials and Methods

After approval by local ethic committee (Reference Number: 133-16-25042016), patients undergoing biventricular repair with double-patch technique only for complete AV canal defects from 1999 to 2013 in our center were included in the study. The ethic committee waived written informed consent of patients. A predefined protocol determining

timing of surgery, anesthesia management, cardiopulmonary bypass (CPB), and Intensive Care Unit (ICU) treatment was used throughout the study. Demographic and perioperative data were collected from clinical information system, and retrospective chart review was performed.

Perioperative management protocol included pre-, intra- and post-operative management [Figure 1].

### Preoperative management

All infants were treated with digoxin (5 µg/kg/day - therapeutic level of 0.5–1 ng/ml), furosemide (1–2 mg/kg/day in 3 divided doses), and spironolactone (1–2 mg/kg/day). Serum digoxin levels were measured only in patients with signs of toxicity. Captopril (1–2 mg/kg/day in 3 divided doses) was added with severe AV valve regurgitation. Hypothyroidism (thyroid-stimulating hormone [TSH] >6 mIU/L) was treated with oral thyroxine (beginning with 12.5 µg/day)

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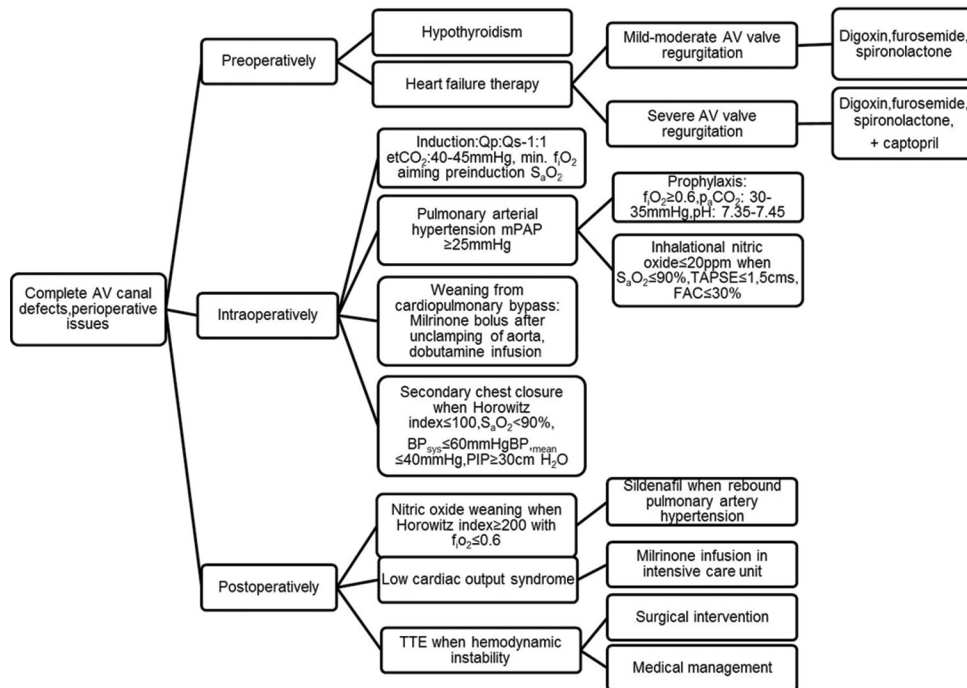
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**Figure 1: Algorithm for management of patients with complete atrioventricular canal defects.**  $\text{FiO}_2$ : Fractional inspiratory oxygen concentration, Min.: Minimum, Qp: Qs-pulmonary to systemic blood flow,  $\text{BP}_{\text{sys}}$ : Systolic arterial pressure,  $\text{BP}_{\text{mean}}$ : Mean arterial pressure, TTE: Transthoracic echocardiography, FAC: Fractional area change, TAPSE: Tricuspid annular plane systolic excursion, mPAP: Mean pulmonary artery pressure, PIP: Peak inspiratory pressure

and was titrated till blood levels of TSH and free thyroxine were normalized. Target weight for surgery was  $\geq 5$  kg.

## Intraoperative management

### Anesthetic management

Neither premedication nor preemptive analgesia was used. Noninvasive blood pressure, oxygen saturation, and five-lead electrocardiogram were monitored during induction. Total intravenous anesthesia (TIVA) was given with etomidate (0.4 mg/kg), midazolam (0.1 mg/kg), and fentanyl (3–4  $\mu\text{g}/\text{kg}$ ). Sevoflurane (2–4 MAC) induction was done only in patients coming to induction room without venous access. After venous access was obtained in these patients, sevoflurane was stopped. Muscle relaxation was achieved with pancuronium bromide (0.1 mg/kg); one-fourth of the induction dose was repeated every 45 min. Remifentanyl (0.3–0.5  $\mu\text{g}/\text{kg}/\text{min}$ ) and midazolam (0.2–0.4 mg/kg/h) infusions were used for maintenance of anesthesia. Cefazolin (30 mg/kg) was given as antibiotic prophylaxis during induction and after CPB. A mean arterial pressure  $>40$  mmHg was targeted. Minute volume ventilation was adjusted to achieve end-tidal  $\text{CO}_2$  40–45 mmHg using pressure-controlled ventilation (Primus, Draeger, Luebeck, Germany).  $\text{FiO}_2$  was adjusted to maintain preinduction  $\text{SaO}_2$  value. Methylprednisolone was administered (20 mg/kg during induction and same dose was added to priming solution). After intubation, central venous access was achieved with triple-lumen catheter (internal jugular  $\gg$  subclavian  $\gg$

femoral vein). For invasive blood pressure measurement, 24-gauge arterial cannula was inserted (radial  $\gg$  brachial  $\gg$  femoral artery). Nasal, rectal, and foot skin temperature was monitored. Blood gas analysis (arterial and central venous) was performed after induction and every 30 min. Near infrared spectroscopy with optodes placed on the forehead and in the region of lumbar vertebra was used (NIRO 200<sup>®</sup>, NIRO 200 NX<sup>®</sup>, Hamamatsu Photonics, Germany).

### Cardiopulmonary bypass priming solution and additives

Priming volume constituted of mannitol 15% (4 ml/kg), 80 ml of albumin 5%, 5 ml of sodium bicarbonate 8.4%, heparin (100 IU/kg), and tranexamic acid (10 mg/kg). Since 2007, the priming volume was reduced from 320 ml to 140–180 ml (Kids D100<sup>®</sup> Sorin, Italy, CAPIOX FX-05<sup>®</sup>, Terumo Corporation, Japan). Additional doses of tranexamic acid (10 mg/kg) were given before skin incision and post-CPB after reversal of heparin. Nearly 300 IU/kg heparin was given and CPB was initiated when activated clotting time ACT was  $\geq 400$ . Target blood heparin concentration was 3 IU/ml (HEPCON, HMS PLUS<sup>®</sup>, Medtronic, Minneapolis). Eighty milliliter of erythrocyte concentrate was added (hematocrit on CPB 28%–30%). CPB flow was maintained at 2.5 L/m<sup>2</sup>/min and reduced to 70%–80% when rectal temperature of 28°C–30°C was achieved. Alpha-stat management was used throughout CPB.

Antegrade cardioplegia was administered in dose of 35 ml/kg (St. Thomas Hospital I Solution<sup>®</sup>, Abbott

Laboratories, Chicago, USA). About 30 ml/kg was repeated when cross-clamp was  $\geq 90$  min.

#### *Rewarming and cardiopulmonary bypass weaning*

With convective warming (underbody Bair Hugger, Arizona), nitroglycerine was used (10–15  $\mu\text{g}/\text{kg}/\text{min}$ ) during rewarming.<sup>[4]</sup> Mean arterial blood pressure of  $\geq 40$  mm Hg was targeted using maximum pump flows maintaining cerebral oximetry parameters within 20% of baseline limit. Ultrafiltration at rate of 100–150 ml/kg/h was performed during rewarming. Fresh frozen plasma (100–150 ml for patients  $< 10$  kg body weight and 200–250 ml for patients  $> 10$  kg body weight) was administered as volume substitution during ultrafiltration.

After unclamping aorta, a bolus of 50  $\mu\text{g}/\text{kg}$  milrinone was given and continuous dobutamine (5  $\mu\text{g}/\text{kg}/\text{min}$ ) infusion was started. Focused transesophageal echocardiography was performed. AV blocks were managed with sequential AV pacing. Junctional ectopic tachycardia (JET) was treated with hypothermia (35°C), deepening of analgesia and sedation, amiodarone bolus (3 mg/kg) and infusion, and R-wave synchronized ventricular sensing with atrial pacing.<sup>[5]</sup>

#### *Weaning from cardiopulmonary bypass*

Pressure-controlled ventilation (Servo 300 Siemens, Germany, Servo-I, Maquet) was performed with:  $\text{FiO}_2$  0.8–1.0, positive end expiratory pressure: 3–5  $\text{cmH}_2\text{O}$ , I:E ratio of 1:2, maintaining  $\text{SaO}_2 > 95\%$  and Horowitz index  $\geq 200$ , moderate hyperventilation ( $\text{paCO}_2$  between 30 and 35 mmHg), peak inspiratory pressure (PIP)  $\leq 30$   $\text{cmH}_2\text{O}$ .

- Blood pH levels of (7.35–7.40) were maintained
- Weaning from CPB was started at minimum hematocrit of 30%, calcium  $\geq 1.2$  mmol/l, and temperature  $\geq 36^\circ\text{C}$ . After weaning, systolic blood pressure was maintained at 70–80 mmHg,  $\text{ScvO}_2 \geq 60\%$ , and Horowitz index  $\geq 200$ .

Nitric oxide  $\leq 20$  ppm was introduced when fractional area change  $< 30\%$ , tricuspid annular plane systolic excursion (TAPSE)  $< 1.5$  cm, interventricular septal shift, elevated central venous pressure, and subnormal oxygenation ( $\text{SaO}_2 < 90\%$ ). Pulmonary arterial hypertension (PAH) was defined as mean pulmonary artery pressure (PAP)  $> 25$  mmHg using Doppler echocardiography. In the 1<sup>st</sup> year of study period, PAP was measured invasively through surgically placed pressure line in pulmonary artery. Following two re-sternotomies due to bleeding after withdrawal of the PAP line, this method was abandoned and echocardiography was used instead.

Heparin was antagonized with protamine (Hepcon, HMS PLUS®, Medtronic, Minneapolis).

Chest was closed secondarily, in case of cardiorespiratory instability (i.e., Horowitz index  $\leq 100$ ,  $\text{ScvO}_2 < 60\%$ ,

$\text{S}_a\text{O}_2 < 90\%$ , PIP  $\geq 30$   $\text{cm H}_2\text{O}$ , systolic blood pressure  $< 60$  mmHg, and mean arterial pressure  $\leq 40$  mmHg) with maximum inotropic support (dobutamine 10  $\mu\text{g}/\text{kg}/\text{min}$  + adrenaline  $\geq 0.05$   $\mu\text{g}/\text{kg}/\text{min}$ ).

#### **Postoperative management**

Transthoracic echocardiography (TTE) was performed in ICU with episodes of hemodynamic instability.

Infusion of remifentanyl (0.3–0.5  $\mu\text{g}/\text{kg}/\text{min}$ ) and midazolam (0.2–0.4 mg/kg/h) was changed to fentanyl (6–12  $\mu\text{g}/\text{kg}/\text{h}$ ) and midazolam (0.2–0.4 mg/kg/h) infusion in patients requiring  $\geq 24$  h of ventilation.

Below 1 ml/kg/h urine output, infusion of loop diuretics was started after excluding hypovolemia and low cardiac output (LCO). Acute renal failure was defined as persistent oliguria below 1 ml/kg/h for  $> 4$  h, serum creatinine  $> 75$   $\mu\text{mol}/\text{l}$ , metabolic acidosis, and serum potassium  $> 5.5$  mmol/l despite diuretic therapy and inotropic support.<sup>[6]</sup>

Milrinone infusion (0.3–0.5  $\mu\text{g}/\text{kg}/\text{min}$ ) was started postoperatively only in patients with signs of LCO. LCO was defined by  $\text{ScvO}_2 \leq 60\%$ , arteriovenous oxygen saturation difference  $\geq 30\%$ , urine output  $< 1$  ml/kg/h, metabolic acidosis with base excess  $\geq -4$  mmol/l, serial elevation of serum lactate levels  $\geq 2$  mmol/l in two consecutive blood gas analyses, and echocardiographic evidence of ventricular dysfunction.<sup>[7]</sup>

Captopril therapy was started (1–2 mg/kg/day in 3 divided doses) when residual mitral regurgitation was  $\geq$  moderate and systolic blood pressure was  $\geq 90$  mmHg.

Ventilatory weaning started with reduction of  $\text{FiO}_2$  to 0.6 maintaining Horowitz index  $\geq 200$ . This was followed by rapid tapering of nitric oxide 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,  $\text{FiO}_2$  was increased to  $> 0.6$  and nitric oxide to 20 ppm and oral sildenafil was 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.

Patients with open chest received cefotaxime (150–200 mg/kg/day in 3 divided doses) in combination with piperacillin (200–240 mg/kg/day in 3–4 divided doses). Sepsis was diagnosed with petechial hemorrhages, band neutrophils in blood  $\geq 15\%$ , rise in blood procalcitonin levels, elevation of interleukin-6, systemic inflammatory response syndrome, and positive blood culture. Therapy was directed with antibiotics based on culture and sensitivity.<sup>[8]</sup>

Pain was assessed daily with KUSS<sup>[9]</sup> scoring system in awake patients and COMFORT B<sup>[10]</sup> system in intubated patients. Satisfactory pain therapy was defined as KUSS<sup>[9]</sup> score  $\leq 4$  or a COMFORT-B<sup>[10]</sup> score 11–22.

During weaning, remifentanyl/fentanyl infusions were bridged with piritramid infusion (0.8 mg/kg/day in infants and 0.4 mg/kg/day in neonates), as synthetic opioid. Piritramid infusion was weaned after the removal of intercostal drains adding nonopioid analgesic. In newborns and infants, paracetamol was given in dose of 7.5 mg/kg orally or intravenously every 8 h (maximum daily dose of 30 mg/kg/day). Metamizol infusion was used in patients >12 months of age in dose of 15 mg/kg/dose every 6 h.

Extubation was performed when Horowitz index was >200, pCO<sub>2</sub> between 35 and 40 mmHg, and ScvO<sub>2</sub> >70%, with minimum catecholamine therapy (monotherapy with dobutamine ≤5 µg/kg/min or milrinone 0.3–0.5 µg/kg/min).

In symptomatic patients, transfusion trigger was set as hematocrit <32% in hypovolemic and <30% in normovolemic patients.<sup>[11]</sup>

### Statistics

All results were calculated using GraphPad Prism trial version software for Windows seven (© 2016 GraphPad software, CA, USA) and expressed as mean ± standard deviation.

## Results

### Demographic data

157 infants (49% males and 51% females) were included in study.

One hundred and forty-six (93%) underwent corrective surgery during infancy with mean age of 125 ± 56.9 days. Remaining 11 patients (7%) underwent late correction due to associated congenital heart defects. Secundum atrial septal defect (ASD - 25.5%) and patent foramen ovale (PFO - 14.01%) were the most common associated acyanotic congenital heart defects. Tetralogy of Fallot (TOF - 4.5%) was the most commonly associated cyanotic lesion [Table 1]. Nearly 63.6% of patients were diagnosed with Down syndrome. Incidence of PAH was 60% (*n* = 94) without significant difference between Down and non-Down patients. Mean body weight at surgery was 5.61 ± 6.3 kg.

Preoperative AV valve insufficiency was mild in 52.8%, moderate in 28.6%, and severe in 18.4% of patients.

Captopril was added to preoperative heart failure therapy in 18.4% of patients.

### Surgical intervention

Primary repair was performed in 144 (94.72%) and staged repairs in 13 (8.3%) of patients. Staged procedures included modified Blalock–Taussig shunt (*n* = 4), coarctation repair (*n* = 4), and pulmonary artery banding (*n* = 5).

**Table 1: Incidence of complete atrioventricular canal defects and associated congenital heart defects**

Diagnosis	n (%)
Isolated CAV canal defects	48 (31)
ASD II	40 (25)
PFO	22 (14)
PDA	9 (6)
ASD II + PDA	7 (4.5)
PFO + PDA	7 (4.5)
TOF	4 (3)
TOF + LPA stenosis	2 (1)
PFO + LSVC-CS	2 (1)
ASD II + LSVC	2 (1)
Neonatal coarctation of aorta	4 (3)
Muscular VSD + PFO	1 (0.6)
ASD II + PDA + TOF	1 (0.6)
DORV	1 (0.6)
Coronary fistula	1 (0.6)
ASD II + PS	1 (0.6)
(TAPVC) + LSVC-LA	1 (0.6)
LSVC + unroofed CS	1 (0.6)
Tricuspid straddling	1 (0.6)
Ventricular imbalance	1 (0.6)
IAA	1 (0.6)

CAV: Complete atrioventricular, ASD II: Atrial secundum septal defect, PFO: Patent foramen ovale, PDA: Patent ductus arteriosus, TOF: Tetralogy of Fallot, LPA: Left pulmonary artery, LSVC: Left superior vena cava, CS: Coronary sinus, VSD: Ventricular septal defect, DORV: Double outlet right ventricle, PS: Pulmonary stenosis, TAPVC: Total anomalous pulmonary venous connection, LA: Left atrium, IAA: Interrupted aortic arch

### Cardiopulmonary bypass data

Mean duration of CPB was 125.9 ± 41.6 min with cross-clamp time of 73.9 ± 24 min.

### Postoperative data

Patients were ventilated for mean of 3.3 ± 2.5 days. Length of stay in ICU was 5.9 ± 2 days. Total length of stay in hospital was 14.5 ± 4.7 days.

### Complications

PAH crisis (30%) and arrhythmias (19.7%) were the most common. Chest was closed secondarily in 7% [Table 2].

### Echocardiographic findings at discharge

Mitral valve insufficiency was trivial in 59.1%, mild in 36.3%, moderate in 22.2%, and severe in 0.6% of patients. Tricuspid valve insufficiency was trivial in 51%, mild in 40%, and moderate in 8.9%. Mild mitral valve stenosis and mild tricuspid valve stenosis were reported in one patient each.

The in-hospital mortality was 0%.

**Table 2: Perioperative complications**

Complication	n (%)
Reintubations	14 (8.9)
Preoperative mechanical ventilation	2 (1.2)
LCO syndrome	9 (5.7)
Arrhythmias	31 (19.7)
Permanent pacemaker implantation	4 (2.5)
Diaphragmatic palsy	1 (0.6)
Pulmonary hypertension needing nitric oxide	47 (30)
Respiratory tract infections	14 (8.9)
Pleural effusion	8 (5)
Pneumothorax	2 (1.2)
Chylothorax	8 (5)
Acute respiratory distress syndrome	4 (2.5)
Sepsis	10 (6.3)
Seizures (postoperative new onset)	3 (1.9)
Reoperations due to significant rest defects	6 (3.8)
Resternotomy	5 (3.2)
Renal insufficiency requiring temporary peritoneal dialysis	4 (2.5)
Secondary thorax closure	11 (7)
ECMO	1 (0.6)
Cardiorespiratory arrest postoperative	1 (0.6)

LCO: Low cardiac output, ECMO: Extracorporeal membrane oxygenation

## Discussion

In this single-center study in patients undergoing surgical repair for complete AV canal defects, excellent in-hospital mortality of 0% was achieved. Reported mortality in literature ranges between 2.0%<sup>[12]</sup> and 3.4%.<sup>[3]</sup> Every aspect of institutional management strategy will be discussed subsequently.

### Timing of surgery

In our study population, 93% of patients were operated in infancy with mean age of  $125 \pm 56.9$  days. This was in accordance with age at surgery of 4.6–6 months reported by other authors.<sup>[12,13]</sup> Remaining 7% of patients underwent corrective surgery at later age due to associated congenital heart defects.

### Special anesthetic considerations in Down syndrome patients

Association of AV canal defects and Down syndrome is seen with an incidence between 61.1%<sup>[12]</sup> and 78.4%.<sup>[13]</sup> Hypothyroidism was diagnosed in 19.7%. Of these, 76% were Down syndrome. We reported stridor, atelectasis, and pneumonia in 8.9%. In accordance to findings of Ihringer *et al.*,<sup>[14]</sup> these respiratory complications were more often in Down syndrome patients (69%). PAH occurred in 60% without significant difference between Down and non-Down patients.

## Management of preoperative heart failure

Heart failure management using digoxin, furosemide, and spironolactone is consistent with guidelines of the German Society of Pediatric Cardiology.<sup>[15]</sup> Captopril was added only in patients with severe AV valve regurgitation. Captopril decreases pulmonary-to-systemic blood flow ratio in infants with large left to right shunts having increased systemic vascular resistance.<sup>[16]</sup>

### Anesthetic management

TIVA was used in all patients. There are no studies in literature showing superiority of intravenous or inhalational type of anesthesia.

### Ventilatory management

It was aimed at avoiding pulmonary hyperperfusion ( $Q_p/Q_s - 1:1$ ) before CPB. This was achieved with normoventilation (paCO<sub>2</sub> 40–45 mmHg), minimum FiO<sub>2</sub> to achieve preinduction SaO<sub>2</sub>. In patients with the left to right shunts, permissive hypercapnia and SaO<sub>2</sub> around 90% prevent excessive pulmonary blood flow maintaining adequate tissue oxygenation.<sup>[17]</sup>

Pulmonary vascular resistance increases with hypoxia, acidosis, and hypercarbia (paO<sub>2</sub> <60 mmHg, pH <7.35, and paCO<sub>2</sub> >40 mmHg).<sup>[18]</sup> Post-CPB, moderate hyperventilation was used to reduce pulmonary vascular resistance and to prevent PAH crisis.<sup>[19]</sup>

### Methylprednisolone therapy

The use of methylprednisolone is discussed controversially in literature. Clarizia *et al.*<sup>[20]</sup> described reduction in length of ICU and hospital stay and reduction in ventilation time using perioperative corticosteroids. In contrast, Pasquali *et al.*<sup>[21]</sup> concluded that corticosteroid use increases morbidity in low-risk pediatric cardiac procedures. In our study, blood sugar levels increased nonsignificantly on CPB in all patients, which subsequently normalized spontaneously in ICU. It was not analyzed if this hyperglycemia resulted secondary to physiological stress (CPB, hypothermia, and surgery), methylprednisolone, or combination of both. Nearly 6.3% of our patients developed sepsis. Atz *et al.* also described similar incidence of sepsis in 6 out of 103 patients undergoing AV canal defect repairs.<sup>[22]</sup> Our mean CPB time and aortic cross-clamp time were comparable to database of Jacobs *et al.*<sup>[12]</sup>

### Pulmonary arterial hypertension management

PAH is associated with increased mortality and morbidity.<sup>[23]</sup> It was noted in 60% of patients from which 30% needed inhaled nitric oxide therapy and ventilation  $\geq 24$  h. Milrinone was used as bolus dose.<sup>[7]</sup> Dobutamine infusion was started when more inotropic support was needed.<sup>[24]</sup> Milrinone infusion was used postoperatively only in patients with signs of LCO (5.7%). About 77.8% of European hospitals used milrinone in 70.7% of all drug

regimens as combination therapy to prevent LCO.<sup>[25]</sup> We combined milrinone and dobutamine due to their different modes of action. No vasopressors were required. PAH crises were managed with nitric oxide therapy ( $\leq 20$  ppm). Inhalational nitric oxide improves oxygenation and reduces need for extracorporeal membrane oxygenation (ECMO), so also in hospital mortality and morbidity.<sup>[26]</sup> Initially, pulmonary arterial (PA) pressure was measured using an invasive PA line. Two redo sternotomies after the removal of PA line lead to cessation of its use and more reliance on echocardiography for assessing severity of PAH.

### Management of arrhythmias

Temporary pacing was needed in 12.1% of patients operated for complete AV canal defects.<sup>[13]</sup> Reported incidence of permanent pacemaker implantation ranges from 1.5% to 2.7%.<sup>[12,13]</sup> In our study, rhythm disturbances occurred in 19.7%. Most common were AV-block needing temporary pacemaker (9.4%), JET (7%), nodal rhythm (2%), and supraventricular tachycardia (1.3%). Permanent pacemakers were implanted in 2.5% of patients.<sup>[12,13]</sup>

### Secondary chest closure

Jacobs *et al.* reported secondary chest closure in 3%.<sup>[12]</sup> This is less than in our study population, where chest was secondarily closed in 7%. This might have helped in lowering the incidence of LCO<sup>[27]</sup> in our patients but prolonging the duration of ventilation ( $>24$  h).

### Postoperative Intensive Care Unit management

Negative fluid balance was achieved with institution of diuretic therapy in all patients. Peritoneal dialysis was initiated with signs of acute renal failure<sup>[6]</sup> (2.5%). Risk factors for peritoneal dialysis included prolonged CPB, LCO, and PAH.

### Sildenafil therapy as bridging for nitric oxide

Nemoto *et al.*<sup>[28]</sup> reported beneficial effects of sildenafil as bridging therapy for nitric oxide in patients after congenital heart surgeries. In our study, all patients with persistent PAH received sildenafil and were successfully weaned off from nitric oxide and ventilator.

### Fast tracking

We never attempted to extubate patients in operating room due to complexity of repair, concomitant presence of PAH, and Trisomy 21. All these are known risk factors for fast-track failure.<sup>[29]</sup>

### Low cardiac output syndrome

LCO occurs with an incidence of 25% after congenital heart surgery.<sup>[30]</sup> Prophylactic milrinone reduces incidence of LCO after congenital cardiac surgeries.<sup>[25]</sup> We reported LCO syndrome in 5.7%. This could be explained by our management strategies.

### Reoperations

Incidence of reoperations needing CPB ranges in different studies from 3%<sup>[13]</sup> to 3.9%.<sup>[12]</sup> Resternotomy was needed in 1.3% of patients with bleeding, in 8% of patients not needing CPB,<sup>[13]</sup> and in 3.8% of babies needing CPB. Causes included cleft dehiscence, mitral valve replacement (2 patients each), residual ventricular septal defect (1 patient), and secondary chordae causing subaortic obstruction (1 patient). About 9.8% of patients experienced  $\geq 1$  of the 6 major complications.<sup>[13]</sup> Emergency resternotomy was done in 3.2% (PA catheter removal, tamponade).

### Other less frequent complications

Resuscitation was performed in one patient after surgical correction due to cardiac tamponade. High-frequency jet ventilation and ECMO were needed in one patient 6 days postoperatively due to acute respiratory distress syndrome. Phrenic nerve was injured in one patient.

### Total length of hospital stay

Mean postoperative length of stay varied from 8<sup>[13]</sup> to 13.1<sup>[12]</sup> days. We reported postoperative length of stay in hospital of  $14.5 \pm 4.7$  days.

### Limitations

This is a single-center retrospective study, which does not compare different treatment strategies. The achieved mortality of 0% does not allow us to conclude that every single strategy in our study is optimal. It may demonstrate that an existing management protocol is helpful to achieve excellent results in these high-risk patients. The management protocol has been adopted over the study period. Ultrasound-guided central venous and arterial punctures have been introduced. Heart–lung machine was miniaturized reducing priming volume and consequent hemodilution. PA pressure is measured indirectly using Doppler over tricuspid or pulmonary regurgitation jet compared to invasive measurement in the early period.

### Conclusion

Excellent outcome in patients with complete AV canal defects can be achieved with a well-defined interdisciplinary pre-, intra-, and post-operative management strategies.

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Nil.

### Conflicts of interest

There are no conflicts of interest.

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