CASE REPORT

Anesthesia in A Patient with Pulmonary Atresia with Intact Ventricular Septum (PA IVS) Underwent Bidirectional Cavo-Pulmonary Shunt (BCPS)

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ABSTRACT

Background: Pulmonary atresia with intact ventricular septum (PA-IVS) is a rare condition, encompassing approximately 1%–3% of congenital heart diseases. Patients with PA-IVS have functional single-ventricle physiology. The ultimate possible outcomes for patients with PA-IVS are biventricular circulation, 1.5-ventricle or single-ventricle palliation, or cardiac transplantation. The bidirectional cavopulmonary shunt (BCPS) procedure directs flow from the superior vena cava into both the right and left pulmonary arteries, permitting flow to both lungs. The shunt is considered to be the second stage of palliation and is generally preparative for the third-stage Fontan procedure. The BCPS improves systemic arterial oxygen saturation without increasing ventricular work or pulmonary vascular resistance.

Case: A 10-month-old, 25-day-old boy, weight 9.93 kg, body length 72 cm, diagnosed with PA-IVS, restricted persistent foramen ovale (PFO), right ventricle hypoplastic, and turtuous patent ductus arteriosus (PDA), underwent BCPS, atrial septectomy, and PDA stent evacuation surgery. The patient underwent a PDA stenting and ballooning atrial septectomy (BAS) at 17 days of age at the cathlab. The patient was cyanotic with stable hemodynamics and a saturation of 72% preoperatively.

Discussion: Preoperative fasting must be observed to maintain the patient's hydration state. Pulmonary blood flow and systemic blood flow must be balanced. An adequate analgetic can prevent pain stimuli that increase pulmonary vascular resistance. Drugs to reduce the afterload, such as milrinone, are needed. Mechanical ventilation was set to get PaCO2 between 40 and 45 mmHg. Maintain the normal heart beat, preload, and contractility to maintain cardiac output (CO) with saturation 80–85%. Wean from mechanical ventilation as soon as possible.

Conclusion: Anesthetic management for BCPS in patients with single ventricles from the preoperative period, intraoperative period, and postoperative period. Understanding single ventricle phsiology is important in order to treat the patient.

Keywords: anesthetic management; bidirectional cavopulmonary shunt (BCPS); intact ventricular septum; pulmonary atresia; single ventricle

INTRODUCTION

Pulmonary atresia with intact ventricular septum (PA-IVS) is a rare condition, encompassing approximately 1–3% of congenital heart diseases. Patients with PA-IVS have functional single-ventricle (SV) physiology. PA-IVS is one of the SVs that can undergo biventricular repair.[1]

Patients with functional SV have pulmonary and systemic circulations that are supplied in parallel, creating significant cyanosis and ventricular volume overload. The goal of palliative surgery is to convert SV circulation from a parallel to a series arrangement.[2]

Abnormalities in SV are characterized by undeveloped both ventricles, one of which is rudimentary or hypoplastic, with only one dominant ventricle supplying the blood to the lungs and systemic. Understanding the anatomy difference and the effects of anesthesia on cardiovascular physiology and respiration are important for perioperative care. The treatment of SV consists of 3 stages, that is: (1) Stage 1: Norwood, BT shunt, PA banding or hybrid (PDA stent, atrial septectomy); (2) Stage 2: bidirectional cavopulmonary shunt (BCPS); (3) Stage 3: Fontan or total cavopulmonary connection.

An optimal communication between anesthiologist, surgeon, and cardiologist is very important for patient treatment. Understanding the physiology of SV will make anesthetic management easier, as in this case report.

CASE

A 10-month-old, 25-day-old boy, with the chief complaint of cyanosis at the finger and lip, mainly when he cries. He breathes faster when he plays or crawls. There was no current complaint when the examination was carried out. There is currently no cough or shortness of breath. The defects have been known since the patient was 2 days old, where the baby turns cyanotic when crying and seizures occur. Born by caesarean section, she immediately cried without cyanosis. The patient underwent a PDA stenting and ballooning atrial septectomy (BAS) at 17 days of age at the cathlab. The patient received therapy with captopril 4 mg three times daily, bisoprolol 0.313 mg three times daily, and aspilet 50 mg once daily (which had been stopped 7 days before the operation).

On physical examination, the airway was free, respiration rate 28 times per minute, saturation 72%, without retraction, wheezing, or rales. Blood pressure 139/83 mmHg, heart rate 101 times per minute, regullar, the heart sound S1S2, there is murmur. As you are aware, there is no swelling, and the finger nails and lips are cyanotic. There are laboratory examinations (Table 1) and chest x-rays (Figure 1). From the chest x-ray, there is enlargement of the heart, with a cardio-thoracic ratio (CTR) of 65%, the heart waist disappearing, and the apex upward. There is no infiltration at both lungs.

Table 1. Laboratories

Hb	15.9	PT	11.6	Billi D	0.16	Na	137	Ur	17.6
Ht	49.5	aPTT	31.7	Billi I	0.18	K	4.5	Cr	0.33
Leukocyte	11850	INR	1.13	Billi T	0.34	Cl	104	TCM	Heg
Thrombocyte	339000	SGOT	36	Alb	4.3	Ca	2.52	HbSAg	NR
Blood Sugar	86	SGPT	24	Glob	1.9	Mg	2.4	Anti HCV	NR

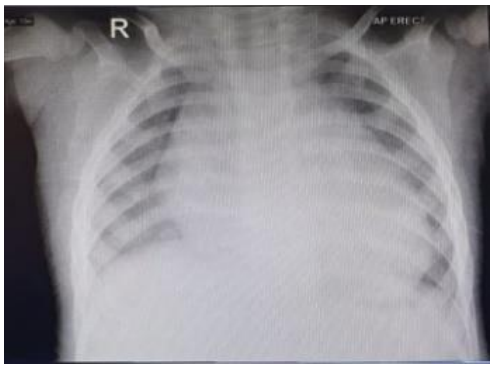


Figure 1. Chest x-ray

The catheterization results were: PA-IVS, PFO-restrictive R-L shunt, RV hypoplastic, and PDA-turtous L-R shunt with distal stenosis. PDA dilation is carried out with Ryujin baloon 2.5 x 15 mm; continue with PDA stenting using stent xience prime 4 x 15 mm; good results. BAS is carried out with BAS balloon number Z-5 at 2 cc twice, with good results.

Multi slice computed tomography (MSCT) results: innominate vein (+), situs solitus, atrioventricular concordance, all pulmonal vein boils down to the left atrium, intact IVS, pulmonal artery is confluence, right proximal 11.7 mm, distal 11.1 mm, left proximal 7.34 mm, distal 6.39 mm, PDA stent (+), descendent aorta 7.34 mm, aorta arch on the left, coroner anomaly (-), collateral (-), thymus is hyperthropy. Echocardiography results: situs solitus, atrioventricular (AV) concordance, pulmonary atresia, atrial septal defect.

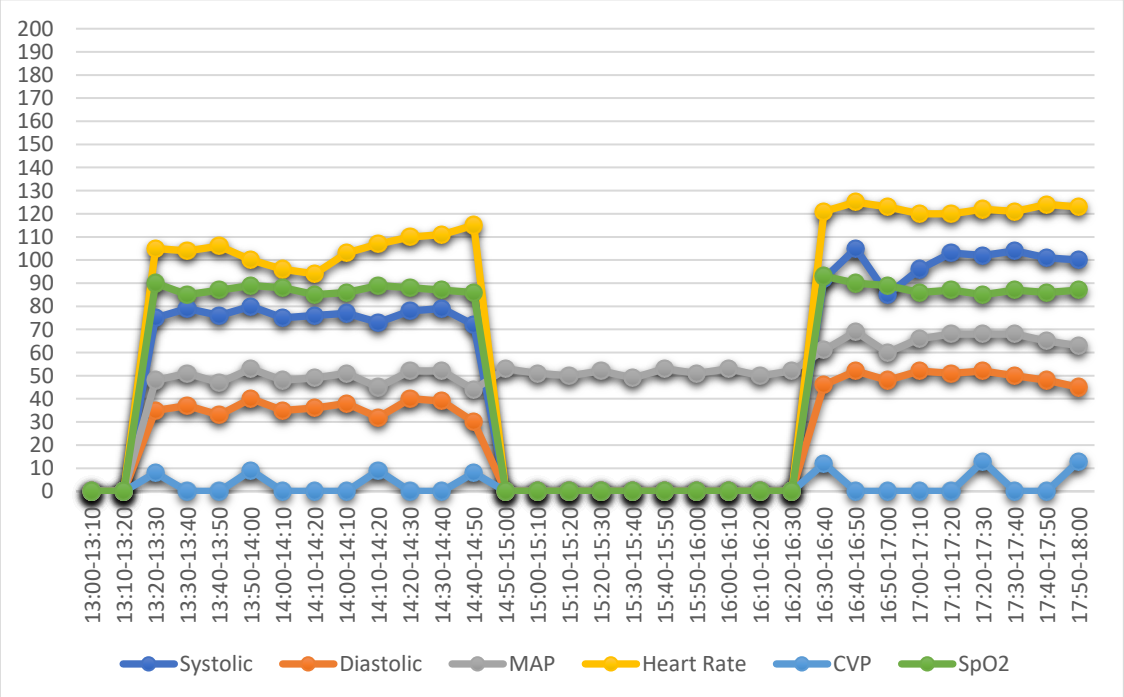
Patient assessed with physical status American Society of Anesthesiologists (ASA) 4. Anesthesia preparation consists of informed consent about general anethesia and its steps, fasting for 6 hours for solids and 2 hours for clear water before the operation, blood transfusion, postoperative analgesia, and postoperative care at the intensive care unit (ICU).

On the day of surgery, the patient enters the operating room and receives inhalation anesthesia with sevoflurane. Installation of monitoring devices such as electrocardiography (ECG) and oxygen saturation simultaneously. After the patient is sedated, we continue with the installation of an intravenous (IV) line on the right site and a radial artery catheter on the left site. Injection of midazolam 0.5 mg, fentanyl 20 mcg, vecuronium 1 mg, and inhalation with sevoflurane 1 vol%. Hemodynamics during induction: blood pressure 78/38 mmHg, heart rate 107 beats per minute, oxygen saturation (SpO2) 85%. Intubation is performed after the drug onset is achieved with endotracheal tube (ETT) number 4.5 (cuff), depth of 12 cm, and evaluation of breathing sound by auscultation. Mechanical ventilation with pressure control with settings: pressure 12 mmHg, respiratory rate 25 times per minute, positive end expiratory pressure (PEEP) 4, inspiration: expiration 1:2, and oxygen fraction 50%, obtained tidal volume 90–100 mL, end tidal CO2 maintained 40–45 mL. Installation of the central venous catheter (CVC) at the right femoral vein with the number 5 Fr cannula was continued with the insertion of a trans esophageal echocardiography (TEE) probe and a temperature probe in the right nose. Echocardiography was performed with TEE.

Mid esophageal (ME) 4 chamber: trivial mitral regurgitation (MR), intact VSD, ASD R-L shunt, small right ventricle size, ME aorta valve long axis: trivial MR, trace aorta regurgitation (AR) trace, ME inflow outflow: pulmonal atresia, mild TR.

Patient in the supine position, preparation for operation field disinfection. Operation starts with a mid-sternal incision. Injection of fentanyl 40 mcg, vecuronium 1 mg, and heparin 300 IU/kg body weight shortly before incision. When the incision and retractor installations were performed, changes in hemodynamic parameters were less than 20% (Graphic 1). Aorta and superior vena cava cannulations were performed. Cardioplegia cannula installed antegrade, aorta cross clamp, cardioplegia streamed, and the heart stopped beating.

Maintenance of anesthesia with O2: air 50%, with oxygen flow 1 liter per minute, sevoflurane 1%, and periodic administration of vecuronium. The hemodynamics during the weaning to post-bypass period are stable, without significant changes, and maintained with milrinone at 0.5 mcg/kg BW/minute. Bleeding 200 ml was replaced with crystaloid 200 ml, fresh frozen plasma (FFP) 179 ml, and trombocyte concentrate (TC) 51 ml. Modified ultrafiltration (MUF) was performed for 10 minutes. Oxygen saturation is 87%, with an oxygen fraction of 50%. Blood pressure: 100/45 mmHg; heart rate: 122 beats per minute. The operation takes 201 minutes, the bypass time is 95 minutes, and the aorta cross clamp takes 5 minutes. Total urine production is 100 ml.



Graphic 1. Hemodynamics during operation

Table 2. Blood gas analysis

Pre bypass				During bypass I				Post bypass			
PH	7.40	Lac	1.1	PH	7.34	Lac	0.9	PH	7.294	Lac	2.3
PCO_2	28	Hb	14.2	PCO_2	44	Hb	9.1	PCO_2	45.4	Hb	10.6
PO_2	58	Hct	42	PO_2	228	Hct	27	PO_2	82.3	Hct	32
HCO_3	17.9	Na	138	HCO_3	23.5	Na	138	HCO_3	22.2	Na	138.5
BE	- 7	K	4.02	BE	-2.5	K	4.02	BE	-3.6	K	3.6
SpO_2	89	Cl	107	SpO_2	99	Cl	108	SpO_2	82.3	Cl	102
GDS	89	Ca	1.28	GDS	141	Ca	1.29	GDS	234	Ca	1.22
ACT	158	Mg	0.59	ACT	759	Mg	0.92	ACT	119	Mg	0.61

After surgery, the patient is transferred to the intensive care unit. Blood pressure 123/52 mmHg, heart rate 129 beats per minute, and milrinone 0.5 mcg/kgBW/minute. Breathing support with mechanical ventilation pressure control mode: pressure 12, respiration rate 25 times per minute, inspiration: expiration (I:E) 1:2, PEEP 3, oxygen fraction 40%, obtained tidal volume 90–100 mL, saturation 87%.

In the first 6 hours, hemodynamics monitoring, mechanical ventilation, oxygen saturation, and laboratory examinations (blood gas analysis, Hb, electrolyte) are performed. From the laboratory results obtained, Hb was 8.7 g/dL, so a transfusion of 100 mL packed red cells (PRC) was carried out, and after Hb evaluation, Hb was 13 g/dL. The inotropics used milrinone (0.75 mcg/kgBB/minute). Analgetics with morphine (10 mcg/kgBW/h). gradually weaning from the mechanical ventilator, and at 5 hours in the ICU, the patient has been extubated.

Echocardiography evaluation in the ICU was obtained: situs solitus, AV concordance, aorta from left ventricle, PA dibuntukan, VSD (-), LV contractility is good, EF 58.5%, RV contractility reduce (tricuspid annular plane systolic excursion (TAPSE) 5.9 mm), mild MR, mild – moderate TR TVG 72 – 75 mmHg, mild AR, ASD (+) 5 mm, bidirectional shunt, torrent pulmonary vein (PV) flow, PDA (-), aorta arch on the left, superiorvena cava (SVC) – right pulmonary artery (RPA) connection is good, laminar flow (+), reversal flow (-).

The patient was admitted to the ICU for 2 days; on the 3rd day, the patient was transferred to the intermediate room for 1 day and moved to the ward. Pain management with morphin 10 mcg/h for 3 days is continued with paracetamol 150 mg 3 times daily, without additional dose for acute pain, and it is considered adequate.

DISCUSSION

Pulmonary atresia with intact ventricular septum (PA-IVS) was first described in 1783. PA-IVS is an obstructive abnormality of the right heart in the presence of pulmonary valve atresia and the absence of a ventricular septal defect (VSD). PA-IVS is a rare condition (1–3% of congenital heart defects).[3-6]

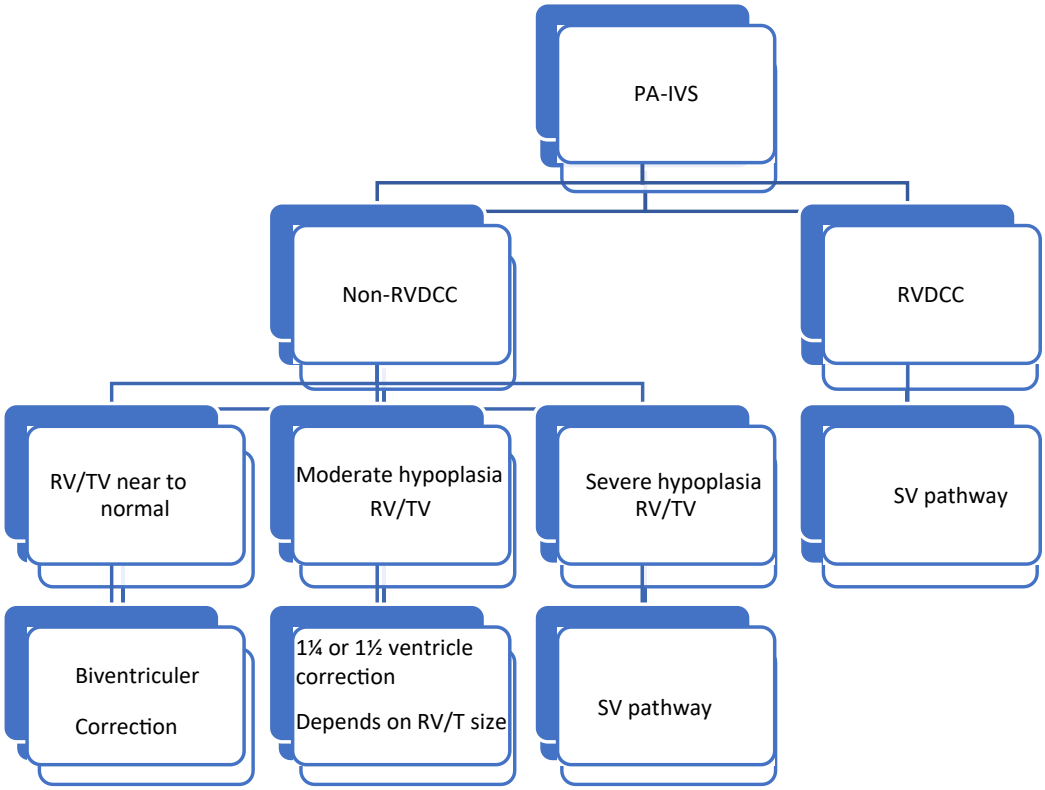
The anatomical and physiological spectrum of PA-IVS is broad, with varying degrees of hypoplasia of the tricuspid valve and right ventricle, ranging from simple pulmonary valve membrane atresia with a formed right ventricle to severe hypoplasticity of the right ventricle with ventriculocoronary fistula.

Coronary artery abnormalities are common in patients with PA-IVS and have a serious effect on the procedure and outcome of surgery. Identification of coronary artery fistulas is not sufficient using echocardiography. MSCT is a diagnostic alternative, but cardiac catheterization is the definitive method for describing coronary artery anatomy and determining the presence or absence of right ventricle-dependent coronary circulation (RVDCC). Therefore, all patients with a diagnosis of PA-IVS must undergo a cardiac catheterization examination.[1,7]

Ventriculocoronary fistula in the presence of coronary artery stenosis will cause RVDCC, where most of the myocardium depends on backflow from the right ventricle. The percentage of PA-IVS cases with RVDCC is around 10 – 20%.[8,9]

RVDCC is defined as the presence of myocardial perfusion dependent on fistulas connected to ventricles, caused by significant obstruction in the proximal coronary arteries and inadequate backflow to the coronary.[10]

When coronary circulation is established, algorithms such as Figure 1 can be used for interventions with catheterization or surgery. The aim of the surgery is to provide enough pulmonary blood flow to replace the PDA and eliminate right ventricular outflow obstruction if possible.[1]



Picture 2. Management of patient with PA-IVS

A good understanding of anatomy and physiology is required for appropriate anesthesia and perioperative management. Specific perioperative considerations are required in the presence of RVDCC because these patients have a high risk of heart failure related to ischemia. Communication with the operator and perfusionist is very important in this case. When stage 2 (BCPS) and stage 3 (fontan) are performed with cardiopulmonary bypass (CPB), a second arterial cannula through the right atrium is used to maintain pressure in the right ventricle to ensure adequate coronary perfusion.[5]

Neonates with PA-IVS have the physiological characteristics of the presence of atresia of the pulmonary valve, which prevents blood from flowing from the right ventricle. With right ventricle outflow tract (RVOT) obstruction, the right ventricle becomes small and hypertrophied. As a result, pressure at the right ventricle increases, making a right-left shunt through the interatrial connection and mixing blood in the left heart. Desaturated blood flows into the aorta to provide systemic and pulmonary blood flow through the patent ductus arteriosus (PDA). These patients have a functional single ventricle.[11,12]

Peroperative management

Preoperative

Neonates with PA-IVS depend on the PDA for adequate pulmonary blood flow and also require prostaglandin E1 (PGE1). Admiministration of PGE1 requires intravenous access. There should be physiological monitoring that should be performed in the intensive care unit to identify physiological changes and side effects. The incidence of apneumonia in neonates who get PGE1 is around 15-20%, and they need to be intubated and require mechanical ventilation. Patients with severe cyanotis require oxygen supplements.[5,13]

Special attention is needed to maintain equilibrium between pulmonary and systemic flow (Qp:Qs), especially if mechanical ventilation is required. Hyperoxygen or hypocarbia may cause excess pulmonary blood flow. Standard evaluations such as chest x-rays, electrocardiograms, and catheterization are used for evaluating the coronary and the presence or absence of RVDCC.

Intraoperative

Stage 1 (neonatal palliative) can be done by creating a systemic-pulmonary artery shunt and decompression of the right ventricle. Things that need to be considered in the systemic-pulmonary artery shunt are maintaining an adequate balance of Qp:Qs and cardiac output, avoiding low diastolic pressure, and monitoring the echocardiogram for the presence or absence of myocardial ischemic signs. Low diastolic pressure causing ischemic of the myocardium.

At the right ventricular decompression procedure (valvotomy or transmural patch), preload or filling pressure and ventricular function must be maintained, and inotropic administration that can reduce afterload (dobutamine, milrinone, and nirtic oxide (NO)) is required. Inadequate preload will cause fatal myocardial ischemic events in patients with right ventricular coronary fistula, or RVDCC. Inadequate pressure in the right ventricle during CPB or assited by veno-arterial (VA) extracorporeal membrane oxygenation (ECMO) can cause the occurrence of myocardial infarction almost all over myocardium.[5]

Stage 2 (BCPS)
Patients with PA-IVS who cannot perform biventricular repair will undergo BCPS surgery at the age of 3-6 months. Installation of standard, non-invasive monitoring. Arterial line installed on the contralateral side of the systemic-pulmonary artery shunt. TEE is used to ensure myocardial function and the outcome of surgery.

Anesthesia induction and maintenance are achieved through a balance between opioids, inhalation gas, and muscle relaxants. To maintain adequate flow through the new BCPS, it must be normocapnia. If hypoxemia occurs after BCPS, mild hypercarbia with a normal pH is more beneficial because cerebral vasodilation increases cavopulmonary blood flow, thereby increasing oxygen delivery to the system. Maintain low pulmonary vascular resistance. Prevent atelectasis, excessive lung distention, and metabolic acidosis. The head-up position (30º) increases venous drainage. Spontaneous ventilation and early extubation provide advantages for patients with BCPS.[5]

Stage 3 (Fontan)
Intravascular volume must be maintained. Make sure the AV is synchronous (sinus rhythm or pacing). Special consideration is given if there is an RVDCC, where one arterial cannula passes through the RA to deliver pressure on the RV while in CPB. Partial CPB flow with mechanical ventilation.[5]

Postoperative
Extubation should be done as soon as possible because intrathoracic negative pressure reduces the afterload of the right ventricle, thereby increasing blood flow to the lungs. Inotropic administration is needed at the postoperative stage.[5]

Single Ventricle (SV)
SV is classified as a complex congenital heart disease, where there is a mixture between pulmonary venous blood and systemic veins and one ventricle that functions to pump blood to the lungs and systemic.

Morphologically, there are some anatomical variations of SV (Table 1). Hypoplastic left heart syndrome (HLHS) is the most common variation (about 25% of single ventricle cases). Other abnormalities include double inlet ventricle (right or left ventricular dominance), pulmonary atresia with intact ventricular septum (left ventricular dominance), tricuspid atresia (right or left ventricular dominance), and an unbalanced atrioventricular canal defect (right or left ventricular dominance).[14]

Table 6. Classification of SV (pathologically)

Pathology	Pulmonary blood flow	Reasons for not being able to do biventricular repair
SV		
LV (DILV)	↓ more often than ↑ depends on pulmonary or aortic obstruction	RV hypoplastic
RV (DORV)	↓ more often than ↑ depends on pulmonary or aortic obstruction	LV hypoplastic or inlet VSD straddling TV
Unbalanced atrioventricular canal defect	↑ more often than ↓	inlet VSD straddling TV
Heterotaxy syndrome	↓ more often than ↑ depends on pulmonary or aortic obstruction	Common AV valve, common atrium
Tricuspid Atresia		
Type I – GA NR	↓ most often	
Type II – D TGA	↑ in general	
Type III – L TGA	↓ or ↑	
HLHS	↑	Hypoplastic LV and ascending aorta

Captions: DILV, double-inlet left ventricle; DORV, double-outlet right ventricle; LV, left ventricle; RV, right ventricle; AVC, atrioventricular canal; GA, great arteries; NR, normally related; TGA, transposition of great arteries; HLHS, hypoplastic left-heart syndrome; VSD, ventricular septal defect; TV, tricuspid valve. *Heterotaxy syndrome characterized by a failure to distinguish the right and left, causing ambiguity of visceral or atrial sites and often perceived as SV right

Pathophysiology of SV

Functional pathophysiology of systemic SV: mixing of systemic and pulmonary venous blood occurs in the atria or ventricles. Optimal oxygen delivery involves balancing systemic vascular resistance (SVR) and pulmonary vascular resistance (PVR), as well as maintaining adequate cardiac output.[16]

If PVR decreases and SVR increases, then blood flow to the lungs will be excessive. It can be seen from saturation > 85% that lung compatibility decreases, hepatomegaly, ventricular dilation, and atrioventricular valve regurgitation occur, so that CO is low. If blood flow to the lungs is inadequate, the patient will experience desaturation (SpO2 < 75%), cyanosis, and CO decreasing (Diagram 1).

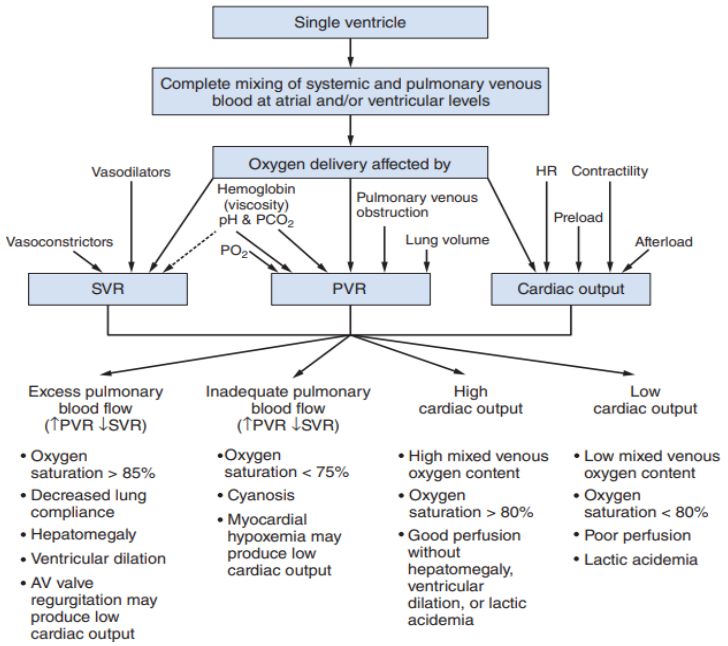


Diagram 1. Patophysiology of SV

SV surgical procedure

Surgical procedures for patients with SV are palliative rather than corrective. The surgical procedure consists of three stages (Figure 2). On each stage, there are three physiological goals that must be achieved: (1) There must be a source with an adequate amount of pulmonary blood flow, sufficient to prevent hypoxemia but not too large that can cause pulmonary arterial hypertension (PAH); (2) There is no obstruction to drain blood from the pulmonary veins to the systemic ventricles; (3) There is no obstruction from the systemic to any part of the aorta.[1]

The preoperative evaluation of patients with SV includes a chest x-ray, pulse oxymetry, hemoglobin, electrolytes, echocardiography, and electrocardiogram. Patients with SV require comprehensive monitoring during surgery. The monitoring tools are: pulse oxymetry (SpO2), end tidal CO2 (EtCO2), near infrared spectrometry (NIRS), electrocardiogram, temperature, arterial line, central venous pressure (CVP), urinary catheter, and TEE.

Stage 1

At this stage, the goal is to suffice blood flow to the lungs to prevent severe cyanosis and heart failure, to provide systemic blood flow to which there is no obstruction, as well as to maintain blood flow to the lungs: blood flow to systemic (Qp:Qs) ~ 1.

Patients with restrictive blood flow will experience cyanosis because blood flow to the lungs is reduced; therefore, a blalock taussig (BT) shunt is performed. Patients with excessive blood flow to the lungs will easily experience heart failure. In these patients, PA banding is performed to reduce high pulmonary pressure. Patients with HLHS will have noorwood for reducing blood flow obstruction to systemic.

The hybrid aims to provide pulmonary and systemic blood flow to minimize the stress of surgery and neurological injury. This is a combination of: (1) percutaneous stenting of the PDA to prevent PDA closure, and it may stop the use of prostaglandin; (2) off-cardiopulmonary bypass (CPB) surgery controls pulmonary blood flow with bands surrounding the right and left pulmonary arteries, mechanically restricting pulmonary blood flow but creating an unobstructed flow from the right ventricle through the PDA to the aorta; (3) if necessary, a percutaneous atrial septostomy is made, either at the same time or separately, to ensure complete intracardiac mixing.

Early surgical injuries (due to inflammation and stress response) are minimized, and the use of CPB is avoided until the child is stronger both physiologically and neurologically. Some institutions use the hybrid procedure at stage 1 as the standard procedure, while others use this approach only for children with high risk (children with a low body weight < 2.5 kg or premature).[17]

Pulmonary artery banding (PA banding)

The important principle that must be understood when the PA banding is performed is to know the basic condition of the patient, thereby ensuring that the tightness is appropriate for the patient. FiO2 and PaCO2 should also be close to the basic values. PA banding that is too tight will cause severe cyanosis and bradycardia, while if it is too loose, it will cause a slight change from the basic condition.

Methods for assessing the tightness of the bands: (1) changes in oxygen saturation and PaO2. Target SpO2 75%-85%, PaO2 40-45 mmHg; (2) direct measurement of pulmonary artery pressure in the distal of the banding PA. Target 1/3 systemic arterial pressure (20%-25%).[10]

PA banding is performed through thoracotomy or median sternotomy, without CPB. When it is too tight, it will significantly decrease lung flow and cause a high ventricular afterload. TEE has a role in assessing ventricular function during PA banding. If ventricular distention occurs, the band needs to be loosened or with inotropic administration.[1]

BT Shunt

The preparation of fasting must be observed to maintain the hydration state of the patient and avoid delays in operating time. Administration of heparin before shunt installation to prevent thrombosis.

Something to be concerned about is that when the shunt opens, the blood pressure decreases because some systemic blood flow will be taken to the lungs. Therefore, fluid administration is needed when opening the shunt. O2 saturation improves; therefore, the oxygen fraction titrated with target SaO2 is 75–85%. Give inotropics if needed when opening the shunt.

Ensure oxygen delivery is adequate by maintaining a hematocrit of 40–45% and sufficient intravascular volume. Installation of the artery line on the contralateral side of the BT shunt. Lung retraction or side clamp PA can cause atelectation during the shunt in tide; therefore, intermittent lung reinflation is needed.

The expected objectives at this stage are: physiological Qp; Qs = 0.7-1.5: 1, oxygen saturation 75–80%, mix vein saturation (Sa-vO2) 25-30%, and blood pressure > 60/30 mmHg.

Problems that happen in stage 1 are: [1]

a. Increased blood flow to the lungs (Qp: Qs > 2-3; 1).
   Increased saturation (SaO2 > 85%, Sa-vO2 35–45%), blood pressure drops (blood pressure < 60/30 mmHg, diastolic < 15–25 mmHg). These are caused by low PVR, the BT shunt being too big, residual arch obstruction, and high SVR.

   Increased blood flow to the lungs can be overcome by increasing PVR (controlled hypoventilation, mild acidosis, low FiO2), increasing oxygen delivery (reducing afterload, inotropics, hematocrit > 40%), or surgical intervention (BT shunt tied, arch reconstruction).

b. Decreased blood flow to the lungs (Qp:Qs < 0.7:1)
   Saturation decreases to < 65-75%, SvO2 < 30%, blood pressure > 70/40 mmHg, and diastolic > 40 mmHg. Caused by a high PVR and a small shunt.

   Overcome this with controlled hyperventilation to lower PVR. Administration of sedation or paralysis, aggressive treatment of atelectasis, consideration of giving nitric oxide, increasing oxygen delivery with inotropics, and if needed, surgery to correct the shunt.

c. Low cardiac output
   Desaturation (SaO2 < 70%, Sa-vO2 35-40%, SvO2 < 30%), low blood pressure (< 60/30 mmHg). It is caused by ventricular dysfunction (myocardial ischemic, reduced

contractility, afterload mismatch, regurgitation of the atrioventricular valves).

Overcome by reducing O2 consumption (sedation / paralysis, inotropic / afterload reduction), surgical intervention (valve repair, arch reevaluation).

Stage 2

The bidirectional cavopulmonary shunt (BCPS), also called the cavopulmonary (CP) shunt, is a modification of the Glenn shunt and the Hemi-Fontan shunt. This is the second stage of palliative procedures for pediatric patients with SV. It is performed at 3–6 months of age, where SVC is anastomosed to PA and inferior vena cava (IVC) remains connected to RA.[18] At that age, the pulmonary vascular pressure has decreased significantly.

The implementation of the second stage differs in some institutions,[19,20] It is generally performed if the following conditions are achieved: (1) appropriate body weight; (2) age 3-6 months; (3) normal atrioventricular valve function; (4) mean pulmonary artery pressure (MPAP) < 18 mm Hg; (5) transpulmonary gradient (TPG) < 9 mm Hg; (6) normal pulmonary blood vessels; (7) no venovenous collaterals; (8) ventricular end-diastolic pressure < 12 mm Hg.[21]

The goal of the second stage is to maintain pulmonary blood flow and simultaneously reduce the systemic load of the ventricles. Volume reduction triggers the remodeling needed for the preparation of the fontan procedure. This prevents a sudden decrease in ventricular diastolic dimensions and a decrease in muscle mass and ventricular compliance from returning to normal before the Fontan procedure.[22] The second stage of the procedure is to balance pulmonary and systemic blood.

The important physiological principle after this second stage is the presence of cavopulmonary-cerebral circulation, that is, the connection of the superior vena cava, which drains the cerebral circulation to the pulmonary artery. PaCO2 increases blood flow to the brain and has a direct effect by increasing pulmonary artery blood flow. With an increase in blood flow to the pulmonary artery, there is more oxygenated blood in the lungs, and the blood flow to the left atrium through the pulmonary vein is also getting bigger, thereby increasing the amount of well oxygenated blood that flows to the systemic is increase.[16]

In the second-stage procedure (BCPS), the expected arterial saturation is 80–85%. After the BCPS procedure, SVC pressure is equal to PAP (14–18 mmHg), whereas TPG (difference between pulmonary artery pressure (PAP) and left atrial pressure (LAP)) wants to be achieved at 8–10 mmHg. If needed, afterload reduction (a pulmonary vasodilator) such as milrinone is given.

Mechanical ventilation with minimal mean airway pressure and a PaCO2 target of 40-45%, prevents atelectasis and weaning as soon as possible. Prevent sympathetic responses that can cause tachycardia.

Pulmonary blood flow must be as low as possible so that it can receive blood flow from the superior vena cava, which will then flow to the left heart. Ensure that the airway is wide open and that airway secretion, suction, and lung distention are optimally performed at the end of CPB.[15]

Superior cavopulmonary circulation cannot be sustained long-term, there are important sequelae that need to be considered, such as severe cyanosis, the occurrence of pulmonary arteriovenous malformations (PAVMs), or the formation of decompressing venovenous collaterals.[23]

As the child grows, the circulation becomes less effective in maintaining systemic oxygen saturation, especially when the child can walk. When the child gets bigger, blood flow to the SVC is less than to the IVC.[23]

Sufficient preparation for fasting, maintaining the hydration state of the patient, and avoiding delaying the operation schedule. Installation of large intravenous lines and, if necessary, the installation of external pads.

Administration of O2 supplements to these patients is safer. Mechanical ventilation management should maintain adequate systemic reverse blood flow and minimal PVR. Extubation is carried out as soon as possible.

Adequate oxygen delivery is achieved by maintaining a hematocrit of 40–45%, and the intravascular volume should be sufficient. Head elevation creates optimal cerebral drainage, thereby increasing blood flow to the SVC and lungs.

Problems that can arise after these procedures in the second stage:

a. Low cardiac output is caused by ventricular dysfunction, hypovolemi, atrioventricular valve dysfunction, and arrhythmia.
b. Desaturation: Due to decompression of the vein (collateral), the anastomosis undergoes stenosis.
c. Superior vena cava syndrome: with an increase in PVR, it will cause an increase in pressure in the superior vena cava.
d. Systemic hypertension, with increased PVR → increased SVC pressure → increased intracranial pressure (ICP) → MPAP will increase to maintain cerebral perfusion pressure (CPP).

Stage 3: Fontan or total cavopulmonary connection

It was first described in 1971 by Fontan and Baudet as palliative therapy for tricuspid atresia. Fontan is the final stage of palliative procedures for patients with SV.

After this procedure, blood flow becomes serial through the arterial systemic circulation, venosus systemic circulation, and pulmonary circulation, then returns to functional SV. Cyanosis that is still present up to the second stage will decrease.

The patient was ready to perform this procedure in terms of physiology when the ventricular myocardium was remodeling at optimal index conditions for space dimensions and contractility for the volume and workload of the heart. Some centers use a minimum range limit after the second stage, which is 6 months. Combined with catheterization and serial echocardiography results to assess preparedness for the final stage. These data should show adequate function of the ventricles and atrioventricular valves, as well as atrial pressure and central venous pressure that are appropriate, showing a small transpulmonary pressure gradient and low ventricular diastolic end pressure. There are some reasons for not delaying Fontan's procedure; one of them is the

risk of stroke related to venous embolism.[24]

In addition, arterial systemic hypoxemia has a negative impact on all organ systems, and it is best to eliminate it as soon as possible. Some centers perform fontan procedures between the ages of 12 – 24 months.[25] There are two approaches at the fontan stage: intracardiac and extracardiac.

Criteria to performed Fontan:
1. Age > 4 years
2. Normal PAP (mPAP < 15 mmHg)
3. PARi < 4 woods units/m2
4. PA confluence and normal size
5. Sinus rhythm (using pacing/ablation)
6. Competent AV valve
7. Normal ventricular function

The expected things after this procedure is TPG < 10 mmHg and low PVR. A high PVR will decreasing the CO if there is no fenestration. CO is maintained by keeping a normal pulse rate, preload and contractility. Administration of inotropic, pulmonary dilator and afterload reduction if required or by installation of an atrial line (transthoracic by the surgeon).

Patients who underwent fontan have undergone several procedures and therefore need drugs to reduce anxiety. Sufficient fasting is necessary to maintain the hydration state of the patient. The installation of a large intravenous line is due to the risk of bleeding because the patient has undergone several procedures, as well as the installation of external pads.

Mechanical ventilation is set up with PaCO2 targets of 40–45 mmHg, SaO2 > 95%, low mean airway pressure, and atelectasis prevention. Adequate oxygen delivery by maintaining HCt 40–45% and sufficient preload (adequate fluid administration). For these patients, the administration of O2 supplements is safe.

Things that worth noting at inhalational induction in these patients: (1) the presence of venous congestion in the head and tongue, so that SVC pressure is relatively high; (2) coughing, or anything else that causes high intrathoracic pressure can stop blood flow to the lungs, and arterial hypoxemia happens quickly; (3) with Qp:Qs 0.5–0.7:1 inhalation induction will be slower.[1]

Problems that can arise after a Fontan procedure:

1. Pumonal blood flow is low, and atrial preload is inadequate. TPG > 10 mmHg.
   Caused by obstruction of the fenestration, obstruction of the pulmonary artery or vein, or early closure of the fenestration. Treatment: volume replacement, lowering PVR, inotropics, reducing afterload, acidosis correction, or interventions to open or create the fenestration.

2. Systemic ventricular dysfunction (TPG 5 – 10 mmHg)
   Caused by systolic or diastolic dysfunction, regurgitation or stenosis of the AV valve, AV dissync, afterload missmatch, or contractility. Treatment: volume replacement, acidosis correction, careful use of PEEP in cases of pulmonary edema, inotropics, afterload reduction, pacing, surgical intervention (takedown to Bidirectional Glenn Shunt).[17]

In these patients, PA-IVS, restriched PFO, and severe hypoplastic RV were obtained without RVDCC (known from the MSCT's result and the latest catheterization), so the procedure was performed as in SV treatment, which is through 3 stages. Although the results of MSCT and catheterization examinations did not obtain RVDCC or coronary ventriculo fistels, the evaluation of ischemia was carried out through ECG monitoring during surgery.

The first stage, or palliative neonate, has been done: the installation of stents in PDA and BAS in the cath lab at the age of 17 days. This intervention is carried out as soon as possible to improve blood flow to the lungs.

The patient is 10 months old, and his pulmonary vascular resistance pressure has decreased so that it is sufficient for pulmonary vascular perfusion with non-pulsatile venous flow from the superior vena cava so that the BCPS procedure can be performed.

Shunt closure (PDA ligation) creates circulation to the lungs through the superior vena cava. The aorta only drains blood to the systemic, thus reducing the ventricular load that originally provided blood flow to the systemic and pulmonary, now it only provides blood flow to the systemic.

Preoperative examinations in these patients include laboratory (Hb, electrolytes), chest x-ray, oxygen saturation, echocardiography, and electrocardiogram (ECG). Monitoring during surgery includes oxygen saturation, end tidal CO2 (EtCO2), ECG, temperature, artery line, central venous catheter installation, urine production, and TEE.

The patient is fasting for 6 hours for solids and 3 hours for clear water before surgery to maintain the hydration state of the patient and avoid patients with hypovolemia.

Induction of anesthesia using a combination of inhaled anesthesia, opioids, and muscle relaxants in general, a total opioid fentanyl of 5–10 mcg/kg body weight is used, with the aim of adequate emergence, so that extubation can be carried out as soon as possible. 16 In these patients, induction with sevoflurane and vecuronium as the muscle relaxant was done with a total dose of fentanyl of 110 mcg (11 mcg/kgBW).

Correct use an intraoperative ventilator can provide adequate systemic venous return and minimize pulmonary vascular resistance. Using excess peak inflating pressure not only increases intathoracic pressure and inhibits venous return, but also increases pulmonary vascular resistance through direct transmission to pulmonary blood vessels.[2]

The degree of hypercarbia that suits can be achieved by administering tidal volumes of 8–12 ml/kgBW, PEEP of 3–5 cm H2O, and a respiration rate of 4–8 times per minute. Additional PEEP and large tidal volumes can be used to limit excessive pulmonary blood flow.[1]

In this patient, the pressure control was 12, and the tidal volume achieved was 100 mL. Using PEEP 4 cm H2O, respiration rate 25 times per minute with EtCO2 maintained 40–45 and saturation ranging from 85–90%, then the O2 fraction lowered.

Mechanical ventilation settings in the ICU are pressure control mode, respiration rate 12 times per minute, O2 fraction 40%, PEEP 3, and tidal volume 88–100 mL, with saturation acquired at 80–90%. FiO2 adjusted to the saturation target of 80–85%. PaCO2 is maintained at 40–45 mmHg. Within 5 hours in the ICU, the patient has been weaned from mechanical ventilation and extubated. The aim of this is to reduce intrathoracic pressure and improve pulmonary blood flow.

Patients who undergo the BCPS procedure will experience cavopulmonary cerebral circulation; PaCO2 will be maintained at 40–45, thereby increasing cerebral blood flow and venous return to the SVC. But high PaCO2 will increase PVR; therefore, milrinone is given as a pulmonary dilator. Milrinone was subsequently lowered gradually, until it stopped, and continued with sildenafil 3.125 mg three times daily.

In cases of ventricular dysfunction caused by excess fluid, CPB needs inotropic administration. Milrinone (0.5–1.0 μg/kg/min) can be used for patients with high SVR and hypertension. 1 An adequate analgetic such as morphine (10 mcg/kgBW) can prevent pain stimuli that can increase pulmonary vascular resistance.

One of the problems in the perioperative period is the presence of systemic hypertension. This is a response to maintain cerebral perfusion pressure (CPP) due to an increase in venous pressure in the brain.[16]

In these patients, systemic hypertension occurs, so captopril 1.25 mg once daily and spironolactone 12.5 mg once daily are given. This patient was also given concor 0.3125 mg once daily peroral as a rate control.

CONCLUSION

Patient with a heart defect PA IVS has functional SV. The operation procedure consists of three stages. Patients with PA IVS require a more thorough examination for the presence or absence of RVDCC.

Management of anesthesia in patients with SV who underwent the second stage of the procedure (BCPS) starts with preoperative preparation, monitoring during the operation, and postoperative care.

Understanding single ventricle phsiology is important in order to treat the patient. Good teamwork is needed in handling patients with SV who will undergo several stages of surgery.

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