Unusual, Rare Case of Ascending Aortic Atresia and Right Ventricle Dependent Coronary Circulation in a Newborn

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Abstract: We describe a newborn with complex, congenital cardiac anomalies, most notably single ventricle physiology, interruption/atresia of the ascending aorta and a coronary circulation that was totally Right Ventricle (RV) dependent. This arrangement left the coronary supply in a very precarious situation, theoretically much worse than most patients with Pulmonary Atresia/Intact Ventricular Septum (PA/IVS), since the entire coronary circulation was RV dependent. The patient underwent a palliative surgical correction, but expired the following day. We believe this is the first reported case in the literature with this constellation of lesions.

Keywords: RV Dependent, Pulmonary Atresia/Intact Ventricular Septum, Complete Atrioventricular Canal.

INTRODUCTION

We describe a newborn with complex, congenital cardiac anomalies, most notably single ventricle physiology, interruption/atresia of the ascending aorta and a coronary circulation that was totally Right Ventricle (RV) dependent. We believe this is the first reported case in the literature with this constellation of lesions.

CASE REPORT

The patient was a thirty five week gestational age, 2.44 kg baby born with a rather unusual and rare cardiac anatomy. This included Complete Atrioventricular Canal (CAVC), common atrioventricular valve (AVV) attachments to a single left ventricle papillary muscle, mild common AVV regurgitation, nearly common atrium with a moderate secundum atrial septal defect (ASD) and a large primum ASD, Double Outlet Right Ventricle (DORV) with a large sub-pulmonary ventricular septal defect (VSD), large patent ductus arteriosus (PDA) with bidirectional flow, persistent left superior vena cava (LSVC) draining to the coronary sinus, dysplastic pulmonary valve (PV) with mild pulmonary insufficiency (PI), absent aortic valve and atresia/interruption of the ascending aorta proximal to the innominate artery. There also was a 5 millimeter (mm) "stump", the vestigial ascending aorta, arising from the right ventricle (RV), giving rise to the right and left coronary arteries with dysplastic, non-functioning valve leaflets present. Blood would flow freely into this stump during systole and reverse course during diastole. The distal arch vessels branched off the PDA and supplied the right and left subclavians as well as the right and left carotids (Figure **1**). Additional findings included dysmorphism, cleft lip and palate, microcephaly and micropenis. A more "typical" DORV/ CAVC anatomic diagram is appended (Figure **2**)

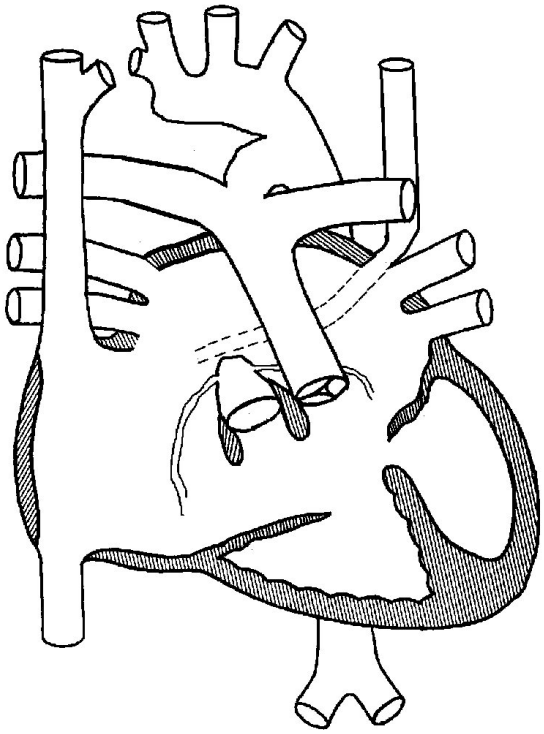

Figure 1: Anatomical drawing of patient's anatomy.

The patient was intubated on admission to the Neonatal Intensive Care Unit (NICU) for respiratory distress and placed on Prostin (alprostadil also know as prostaglandin E_1) at 0.05 micrograms/kilogram/ minute (mcg/kg/min) to maintain ductal patency. The baby was ventilated on room air with pulse oximeter saturations (SpO_2) in the low 90s. On day two of life the patient was started on a calcium gluconate infusion at

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E-ISSN: 2310-9394/14

20 milograms/kilogram/hour (mg/kg/hr) for persistent hypocalcemia and a milrinone infusion at 1 mcg/kg/min for high lactates, due to sytemic hypoperfusion that peaked at 10 millimoles/liter (mmol/L). On day three of life an epinephrine infusion was added at 0.03 mcg/kg/min for hypotension to augment cardiac contractility. Also nitrogen was titrated into the ventilator at a rate of 0.75 liters per minute (LPM) to achieve an inspired oxygen concentration (FiO_2) of 15% and the partial pressure of carbon dioxide (PCO_2) was permitted to passively rise to around 50 millimeters of mercury (mm Hg) in an attempt to increase pulmonary vascular resistance (PVR) and ensure adequate coronary perfusion. Positive end expiratory pressure (PEEP) of seven was also added. Despite these efforts the patient remained hemodynamically labile.

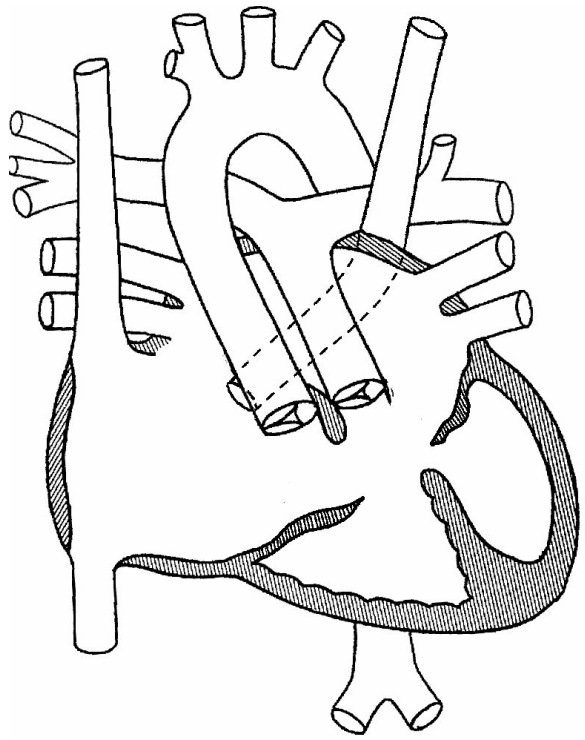

Figure 2: "Typical" Double outlet right ventricle/Complete Atrioventricular canal anatomy.

On day six of life the patient presented to the operating room for surgical repair. This included a Norwood-type palliation for aortic arch atresia, a 3.5 mm Impra central shunt, main pulmonary artery (MPA) transection and MPA plasty with pericardium. The patient was transported with a self inflating bag to maintain a low FiO_2; a PEEP valve was added to the bag because the patient did not tolerate manual ventilation without it. Hypotension and bradycardia resulted without it.

On arrival to the operating room, there existed in situ a double lumen umbilical vein line (UVL), a femoral arterial line (A-line) and two peripheral intravenous (IV) lines. Monitoring consisted of an A-line, central venous pressure (CVP), electrocardiogram (ECG), noninvasive blood pressure (NIBP), capnography with complete gas bench, airway and circuit manometry, esophageal and rectal temperatures, and Near Infrared Spectroscopy (NIRS) with bilateral, frontal cerebral probes, right kidney probe and right thigh probe. The 3.0 uncuffed endotracheal tube (ETT) was changed to 3.0 cuffed ETT due to the presence of large air leak. The baby had been receiving rocuronium, midazolam and fentanyl via infusion in the Cardiac Intensive Care Unit (CICU), but these were all discontinued for transport. The patient remained on Prostin at 0.01 mcg/kg/min and Milrinone at 1 mcg/kg/min. The patient received an additional dose of 25 mcg Fentanyl along with glycopyrolate 0.1 mg, dexamethasone 3 mg and rocuronium 10 mg on induction. The prebypass period was uneventful without any periods of hemodynamic instability or evidence of coronary ischemia. The initial arterial blood gas (ABG), before surgical incision, was pH 7.30, PCO_2 61.9 mm Hg, PO_2 74 mm Hg, HCO_3 30.4 mmol/L, Base excess +2 mmol/L, SaO_2 92%, hemoglobin (Hg) 17.7 gm/dL, Hematocrit (Hct) 52%, lactate 4.51 mmol/L; remaining electrolytes were unremarkable.

After surgical incision and median sternotomy, cannulation was achieved via the main pulmonary artery (MPA) and right atrial appendage. A perfusion cannula was also placed into the "stump" to ensure there was adequate coronary blood flow during cooling; this was also used to administer cardioplegia every 20 minutes and to perfuse the coronaries during the rewarming phase. Once on cardiopulmonary bypass (CPB), the patient was cooled to 18° Celsius (C). A cannula was inserted into the takeoff of the right common carotid artery to ensure perfusion of the brain during the arch reconstruction. Low flow regional CPB was established through this cannula. Then the PDA was divided followed by the division of the MPA. The distal end of the MPA was oversewn with a pericardial patch. Then a 12 mm Contegra, sans the valve tissue, was used to reconstruct the ascending aorta to reestablish continuity between the proximal stump of the MPA and the distal arch vessels. A central shunt was completed next with a 3.5 mm Impra graft. A surgical decision was made not to reimplant the coronaries from the "stump" into the neo-aorta due to the technical difficulty this would present. After

adequate warming and restitution of normal sinus rhythm (NSR), separation from CPB was achieved. The patient was placed on 1 mcg/kg/min of milrinone and 0.1 mcg/kg/min of epinephrine during this time. ABP was 55/20 on termination of CPB. The pressure in the aortic "stump" was 55/2 at the same time; SaO_2 was 90. The arch gradient was checked via manometry (there was none) and then a common atrial line was inserted demonstrating a pressure of 6-10 mm Hg. The patient was returned to the CICU with the chest open, covered by a silastic shield.

Twenty minutes after arrival to the CICU, the patient required a brief period of chest compressions and resuscitation with three epinephrine boluses due to hypotension and bradycardia. The patient responded adequately and remained in critical condition throughout the remainder of the day and night. The patient received transfusions with packed red blood cells (PRBC) and other component therapy and activated Factor VII for continued bleeding. Lactates peaked at 13 mmol/L during the night, but had decreased to 3 mmol/L by the next morning. The next day a mediastinal exploration and delayed sternal closure were performed. The patient initially tolerated this procedure well but after 40 minutes became hypotensive and bradycardic. Resuscitation was commenced with chest compressions, epinephrine and calcium. The chest was reopened and direct cardiac compressions continued. An attempt was made to institute cardiopulmonary support (CPS), but this too was unsuccessful. The patient expired. The family refused an autopsy.

DISCUSSION

Typical Hypoplastic Left Heart Syndrome (HLHS) patients presenting to the operating room for a Stage 1 Norwood are some of the most challenging cases done in pediatric congenital heart programs. Anatomic and physiologic variables make these cases a challenge for the surgeon and anesthesiologist. Careful control of respiratory and circulatory variables must be maintained to ensure adequate coronary perfusion. Typically blood flows retrograde down the ascending aorta to supply the right and left coronary arteries. In this circumstance it is imperative that diastolic blood pressure (DBP) be maintained to provide for adequate coronary perfusion pressure. Maneuvers typically employed include maintaining a normal pH of 7.40, an arterial partial pressure of oxygen (PaO_2) as close to 40 mm Hg as possible by reducing the FiO_2 to 21%, and a partial pressure of carbon dioxide (PCO_2) as close to 40 mm Hg by restricting minute ventilation (MV) or adding exogenous carbon dioxide (CO_2). These maneuvers maintain pulmonary vascular resistance (PVR) and reduce the systemic runoff of blood to the pulmonary circulation, thereby increasing systemic blood flow and coronary blood flow [1].

Typically aortic interruption is classified according to Celoria and Patton [2]. Type A interruption occurs distal to the left subclavian artery (LSCA), Type B occurs distal to the left common carotid and Type C occurs distal to the innominate artery. The incidence is Type A 42%, Type B 53% and Type C 4% [3]. In the preceding case the anatomy was quite unusual given the interruption occurred proximal to the inominate artery. In fact the authors could only find one reported case of this variant in the literature [4]. In that case a Stage 1 Norwood was attempted, but the patient died on post-operative day (POD) seven after requiring Extracorporeal Membrane oxygenation (ECMO) and suffering an intracranial hemorrhage. Jux, Kaulitz, *et al.* reported two cases of antegrade flow in the ascending aorta despite the presence of aortic atresia due to retrograde coronary perfusion through coronary fistulas and sinusoids [5]. In addition, although not exactly like this case, eleven patients with double aortic arch with aortic atresia and left-sided type B interruption have been reported in the literature [6]. This lesion is almost always fatal.

In our case (Figure **1**), the coronary circulation was totally right ventricle (RV) dependent, typically seen with pulmonary atresia (PA) and intact ventricular septum (IVS), leading to RV "steal", run off from the coronaries into the RV during diastole [7]. In normal patients the right ventricle (RV) receives blood supply during both systole and diastole, while the left ventricle (LV) receives most of its blood supply only during diastole; only 10%-20% of LV blood flow occurs during systole and mostly to the subepicardial region. During systole, intracavitary LV pressure equals the systolic coronary artery pressure and flow is reduced or ceases, particularly to the subendocardium [8] (Figure **3**). This makes the sub-endocardium particularly sensitive to ischemia when coronary occlusion occurs as during adult coronary artery disease.

In this case, both coronaries were supplied by the aortic "stump" from the RV which was experiencing systemic blood pressure. Systolic blood pressure in the "stump" equaled systolic blood pressure in the RV cavity. DBP at the "stump" equaled right ventricular end diastolic pressure (RVEDP). This is what we would

expect since there was free, unrestrictive communication (no competent valve) between the RV cavity and the "stump". This arrangement left the coronary supply in a very precarious situation, theoretically much worse than most patients with PA/IVS, since the entire coronary circulation was RV dependent Moreover, since the coronary circulation was not interfacing with the systemic or pulmonary circulations as in more typical single ventricle physiology patients, maneuvers employed to increase PVR to mitigate coronary "steal" were rendered ineffective. This type of anatomy has been previously described, but resulted in death after an attempted systemic to pulmonary artery shunt [9].

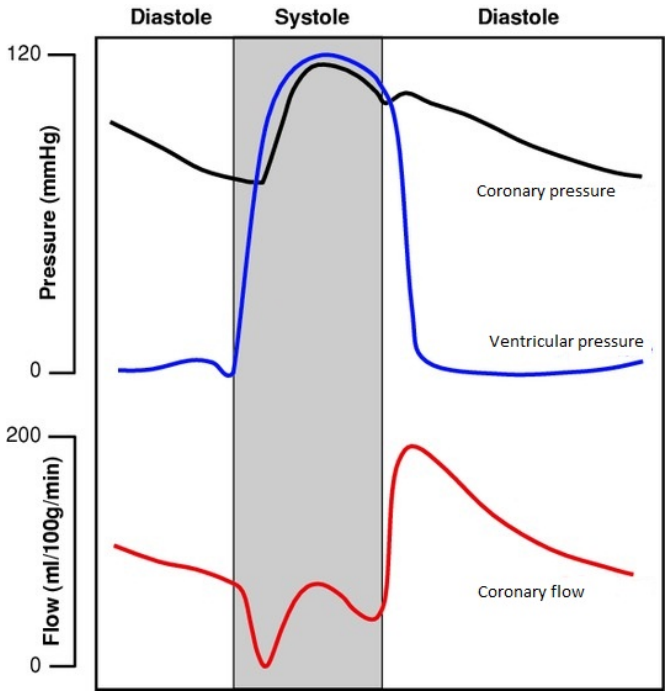

Figure 3: Normal coronary perfusion pressure and flow.

In theory there should be very little coronary perfusion pressure in this patient. During diastole the "stump" pressure was equal to the RVEDP and during systole, the stump pressure was equal to the right ventricle end systolic pressure (RVESP) (Figure **4**). In the OR and then in the CICU, it was imperative to keep the ABP at least 50 mm Hg systolic. During the brief periods when the systolic pressure decreased to below this level, the patient would decompensate quickly requiring active resuscitation, indicating that a substantial portion of the coronary blood flow occurred during systole, not diastole. This decrease in RV pressure is somewhat analogous to what occurs in PA/IVS when the RV is "decompressed" by balloon valvuloplasty of the pulmonary valve. Since this patient maintained hemodynamic stability without evidence of ischemia on electrocardiogram (ECG) as long as ABP was at least 50 mm Hg, the patient's demise probably resulted from a brief period of hypotension, decreasing coronary perfusion and leading to ischemia and death.

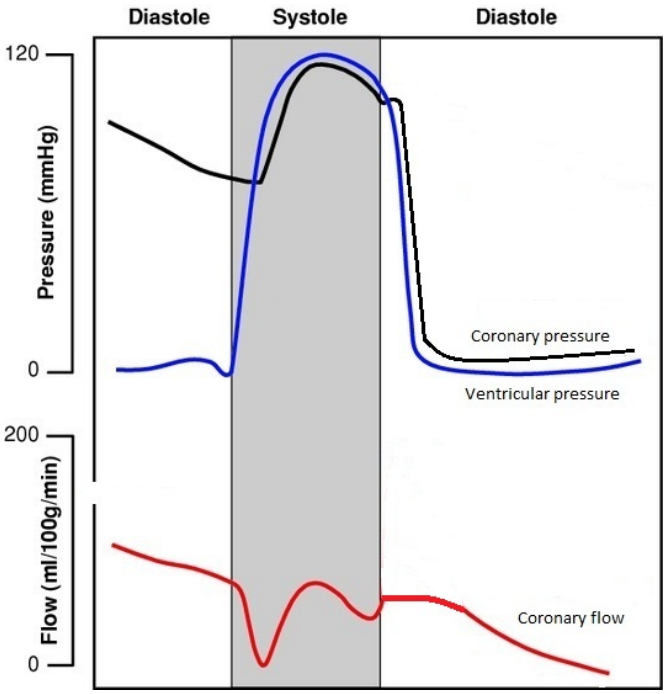

Figure 4: Coronary perfusion pressure and flow in this patient.

ABBREVIATIONS

ABG	=	Arterial blood gas
A-line	=	Arterial line
ASD	=	Atrial septal defect
AVV	=	Atrioventricular valve
CICU	=	Cardiac Intensive Care Unit
CPB	=	Cardiopulmonary bypass
CPS	=	Cardiopulmonary support
C	=	Celsius
CVP	=	Central venous pressure
CAVC	=	Complete Atrioventricular Canal
DORV	=	Double Outlet Right Ventricle
ECG	=	Electrocardiogram

ETT = Endotracheal tube

Hct = Hematocrit

Hg = Hemoglobin

FiO_2 = Inspired oxygen concentration

ICU = Intensive care unit

IV = Intravenous

LSVC = Left superior vena cava

LPM = Liters per minute

MPA = Main pulmonary artery

mcg/kg/min = Micrograms/kilogram/minute

mm = Millimeter

mmol/L = Millimoles/liter

mg/kg/hr = Milograms/kilogram/hour

NIRS = Near Infrared Spectroscopy

NICU = Neonatal Intensive Care Unit

NIBP = Noninvasive blood pressure

NSR = Normal sinus rhythm

PRBC = Packed red blood cells

PCO_2 = Partial pressure of carbon dioxide

PDA = Patent ductus arteriosus

PEEP = Positive end expiratory pressure

PA/IVS = Pulmonary Atresia/Intact Ventricular Septum

PI = Pulmonary insufficiency

PV = Pulmonary valve

PVR = Pulmonary vascular resistance

SpO_2 = Pulse oximeter saturations

rSO_2 = Regional oxygen saturation inderx

RV = Right Ventricle

RVEDP = Right ventricular end diastolic pressure

RVESP = Right ventricle end systolic pressure

UVL = Umbilical vein line

VSD = Ventricular septal defect

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Received on 28-03-2014 Accepted on 08-05-2014 Published on 05-09-2014

DOI: http://dx.doi.org/10.14205/2310-9394.2014.02.01.1