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A 33-day-old Infant with the Transposition of the Great Arteries; A Rare Case Report

*Seyyed-Mohsen Hosseininejad[1], Fateme-Sadat Hosseininejad[2], Hasan Esmaeili[3], Mehdi Aarabi[4], Fateme-Sadat Tabatabaei[5]

[1]Student Research Committee, Golestan University of Medical Sciences, Gorgan, Iran.
[2]Student Research Committee, School of Public Health, Shahroud University of Medical Sciences, Shahroud, Iran.
[3]Assistant Professor of Neonatal and Pediatric Cardiology, Golestan Pediatric and Neonatal Research Center, Golestan University of Medical Sciences, Gorgan, Iran.
[4] Department of Orthopedic Surgery, Toronto East General Hospital, 825 Coxwell Avenue, Toronto, ON M4C 3E7, Canada.
[5]Pediatric Resident, Golestan Pediatric and Neonatal Research Center, Golestan University of Medical Sciences, Gorgan, Iran.

Abstract

Dextro-transposition of the great arteries (d-TGA) is the one most common cyanotic congenital heart disease in neonates. The discordant ventriculoarterial arrangement results in parallel circulation, it so is vital to understand the management. We report a rare interesting but critical case of 33-day-old boy who developed cyanosis and had transposition of great arteries combined with interrupted aortic arch (IAA), diagnosed as d-TGA, and describe the entire interventional management. The patient underwent surgical correction of the transfection and defects.

Keywords: Infant, Cardiopulmonary Bypass, Transposition of Great Vessels.

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Corresponding Author:

Seyyed-Mohsen Hosseininejad: Student Research Committee, Golestan University of Medical Sciences, Shastkola, Gorgan, Iran. Email: Hosseininejad.s.mohsen@gmail.com

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1- INTRODUCTION

Complete transposition of great arteries (transposition of the great artery, TGA) is one of the most common cyanotic congenital heart diseases, with an estimated prevalence of 4.7 per 10 000 live births in the United States (1). This lesion is an entriculoarterial discordance that results in the right ventricle attaching to the aorta and the left ventricle in series with the pulmonary system. If untreated, this congenital anomaly has a mortality of 90% in the first year of life (2). Its anatomic features include a normal atrioventricular connection and an abnormal ventricular arterial connection: the right ventricle connects the aorta; the left ventricle connects the pulmonary artery. Transposition of the Great Arteries, d-Type (d-TGA) first introduced by Matthew Baillie in 1808 (3), is one of the most common congenital heart defects. The prenatal diagnosis of d-TGA remains difficult due to the fact that the four chamber view (4CV) is normal in most cases (4).

Pathophysiological feature of TGA patients is that the systemic circulation and pulmonary circulation systems are two separate parallel loops, and the patients can survive only due to the communication between the pulmonary circulation such as PDA, PFO, ASD, VSD, body pulmonary collateral and so on. The amount of mixing blood of the systemic circulation and pulmonary circulation determines the survival of the TGA patients. The direct causes of death include severe hypoxia, heart failure and cerebral vascular accident (5). General pediatric heart diseases are caused by genetic or environmental factors (6). It is believed the reason of TGA was that the aorticopulmonary septum failed to form helix and formed straight septum (7). The condition of the present child was complicated and the present article narrated the perioperative management.

2- CASE REPORT

A 33-day-old Turkmen boy, weighing 3,900 gr, presented with a hypoxia and extremities and lip cyanosis on clinic checkup visit by a medical intern in our academic hospital, and was referred to the attending physician with a high suspicion for a serious cardiac underlying problem. He was urgently underwent Color Doppler ultrasonography which mainly displayed transposition of great arteries so that the diagnosis revealed as congenital heart disease-TGA, PDA, PFO. After primary stabilizing measures besides intubation and mechanical ventilation, he was admitted at the hospital for emergent operation. Before operation, wet rales at the lower left lung, was auscultated, SPO2 showed 68% (without oxygenation), and alprostadil infusion started. The electrocardiogram showed sinus rhythm, electrical axis shifted to the right +110 degrees; the left atrium was large, and the right atrial and right ventricle, were also large.

Blood gas reported pH 7.38, PCO25.3 kPa, PO2 4.7 kPa, Na+ 130 mmol/L, K+ 4.5 mmol/L, Ca2+ 1.05 mmol/L, Glu 3.2 mmol/L, Lac 11.4 mmol/L, BE ¯1.4 mmol/L, SB 227 mmol/L, $HCO3^-$ 23.7 mmol/L. The infant was kept warm by blower and water blanket insulation, and was given ketamine 3.5 mg, fentanyl 0.03 mg, midazolam 1 mg, and vecuronium 2 mg, then a 3.0 catheter was inserted through the mouth; the right femoral artery was punctured, the right internal jugular vein was punctured, and alprostadil 5 ng/kg/minute were punctured, and dopamine 4 ng/kg/min was infused; breath pressure control was given after intubation, adjusting end-tidal CO2 to 25–30 mmHg, and the blood pressure was maintained at 50–60/30–40 mmHg, HR at 135–150 beats/min, and SPO2 was gradually increased from 60% to 85% thereafter. The sternum was dissected and the aorta and pulmonary artery were isolated. The distal aortic arch was found

interrupted (PDA conduit above); the pathophysiological changes of the patient was changed into following after an on spot discussion: General circulation: superior and inferior vena cava > "right atrial "> right ventricle > "aorta "> upper limb arteries > "upper extremity venous system "> Back to the superior and inferior vena cava. Pulmonary circulation: left pulmonary vein> "left atrium" > ventricular > "pulmonary" >back around pulmonary vein. Communication exists between the left atrium and the right atrium: PFO; Communication exists between the pulmonary artery and aorta: arterial catheter, supplying to the lower limb. In addition there might be pulmonary collateral circulation. After teamwork discussion, the following strategy was decided: conventional aorta, superior and inferior vena cava catheterization + pulmonary artery catheterization (after pulmonary artery occlusion).

Blood gas results before and after pulmonary artery catheterization showed that the oxygen supply to femoral artery was significantly improved. At the beginning of the bypass, SPO2 immediately rose to 100%; with slow cooling, a low-dose intermittent bolus injection of epinephrine given to avoid the early stop of heartbeat started; the blood pressure was maintained, cardioplegia perfusion was begun after the exploration was finished. After the descending aorta was cut off, the arterial catheter-sides were anastomosed to the posterior wall of the ascending aorta, and then Switch surgery was performed. Modified ultrafiltration was carried out after re-beat, supplementing fresh frozen plasma, red blood cells, platelets, followed by bio-gel coating, hemostasis with hemostatic drugs, infusion of dopamine, epinephrine, isoproterenol, Amrinone (INN), to keep HR 150 beats/min, blood pressure 66/49 mmHg, CVP 10–18 cmH2O, SPO2 100%, ETCO2 (End Tidal) 30–35 mmHg. ST section <1.5, with no significant arrhythmia indicate good coronary blood supply and better intraoperative myocardial protection. With delayed sternal closure, placing the atrial, ventricular pacing wire, the patient was transferred to Neonatal Intensive Care Unit (NICU). On the second day, the patient circulation was stable, inhaling NO, with heater warm, HR in 169 beats/min, SPO2 at 100%; BP at 68/53 mmHg; and CVP at 9 cm H2O. On the third day, the vasoactive agents only had dopamine, Amrinone maintenance; the circulating physiological state was significantly improved; chest was closed on 3 day after the operation, HR of 165 beats/min, with the SPO2 100%, BP 75/59 mmHg, and CVP 15 cm H2O were shown. Five days after the surgery, our child was sent back to the general wards, and discharged 15 days after correcting operation.

3- DISCUSSION

Complete TGA consists of 4 types: type 1, with intact ventricular septum; type 2, pulmonary stenosis with intact ventricular septum; type 3, ventricular septal defect, and type 4, ventricular septal defect with pulmonary stenosis. Type 1 was the most common one, followed by type 3. For type 1, blood mixing between the two circulation systems rely merely on the atrial septal defect (PFO), and non-closure of patent ductus arteriosus the same as our case, soon leading to cyanotic and breathing difficulty in infants; oxygenation cannot be used to improve the symptoms but guaranteed in our case; once TGA was diagnosed, patients should be immediately given prostaglandin E1 (0.05 to 0.1 μg/kg/min) to maintain a patent ductus arteriosus. Those had a poor response should be given an emergent balloon dilation of the atrial septal defect to increase blood mixing between the two circulation systems, so as to gain more time for arterial switch operation. At the same time, muscle relaxant drugs, sedative

drugs should be used to reduce the oxygen consumption and mechanical ventilation, and inotropic agents should be used to improve the cardiac function (2-6). Our present case suffered TGA type 1. For the type 3, ventricular septal defect type between the two circulation systems have more communication, therefore the blood was fully mixed and the symptoms were relatively late and lighter, but within a few months after birth the patients can have heart failure. Arterial switch operation (ASO) is the translocation of aorta and pulmonary artery transposition turned; meanwhile the coronary artery was grafted to the aorta to achieve a complete anatomical correction; this is currently an ideal treatment of complete transposition of great arteries translocation. Surgery should be performed within one month after birth so that the left ventricle can develop normally (8, 9, 13-15), however our case with the age of 33 days was actually a magic of life as he, with suffering d-TGA, was alive at such age.

4- CONCLUSION

By this present case, in spite of introducing a magic in an affected infant, we will have a better understanding of the measures for perioperative managements in the arterial switch operation: insulation, to maintain a certain pre and after load and pulmonary arterial pressure. Other key issues include vitro ultrafiltration, blood conservation, myocardial protection, pulmonary artery catheterization, lower body perfusion, deep hypothermic environment regulation, management of coronary artery bypass surgery, and hemostasis after bypass, maintaining blood volume, vasoactive agents use, delayed sternal closure, and pacemaker.

5- ABBREVIATION

PDA: Patent ductus arteriosus,

PFO: Patent foramen ovale,

ASD: Atrial septal defect,

VSD: Ventricular septal defect,

D-TGA: Dextro-transposition of the great arteries,

IAA: Interrupted aortic arch,

CVP: Central Vein Pressure,

ETCO2: End Tidal Carbone dioxide,

INN: Inamrinone trade name Inocor,

HR: Heart rate,

NICU: Neonatal intensive care unit,

SPO2: Oxygen saturation,

BP: Blood pressure,

H2O: Dihydrogen Monoxide (water),

4CV: Four chamber view.

6- CONFLICT OF INTEREST

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