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Original Research

PRIMARY OUTCOMES OF ONE - STAGE REPAIR FOR INTERRUPTED AORTIC ARCH AT VIETNAM NATIONAL CHILDREN'S HOSPITAL

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ABSTRACT

Objective: *This study focuses on primary outcomes of one-stage repair for interrupted aortic arch (IAA) from a single institution from Vietnam.*

Methods: *Between 2012 and September 2017, 27 patients underwent one-stage repair in Children Heart Center, National Hospital of Pediatrics. Kaplan - Meier survival analysis plotted survival and freedom from reoperation curves from follow-up data.*

Results: *Median age was 53 days (range, 9 days to 5 months) and weight was 3.5 kg (range, 2.3 to 6.5 kg). IAA type A in 16 cases (59.3%), IAA type B in 11 case (40.7%). Associated anomalies were ventricular septal defect in 20 (74.1%), aortopulmonary window in 4 (14.8%), truncus arteriosus in 3 (11.1%). Fourteen patients (51.9%) have left ventricular outflow tract obstruction (LVOTO). Median follow-up was 17 months (range, 1 day to 71 months). Survival rates at 30 days, 1 year and 5 years were 81.5%, 77,6% and 77,6% respectively. There were 5 early deaths in hospital, 4 deaths related to nosocomial infection with positive results in blood culture or endotracheal tube, the other was sudden death. One late death after 4 months was caused by severe pulmonary artery stenosis after total repair of interrupted aortic arch associated with truncus arteriosus. There were 5 late reoperations or balloon angioplasty, reoperation for LVOTO in 3, right ventricular outflow tract reconstruction in 1, the other required balloon angioplasty for recurrence of aortic arch stenosis.*

Conclusion: *Primary outcomes of one - stage repair for interrupted aortic arch are safe in our institution. Long - term outcomes evaluation is needed for this complex congenital heart disease.*

Keywords: *interrupted aortic arch, one-stage repair, selective cerebral perfusion, primary*

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I. INTRODUCTION

An interrupted aortic arch (IAA) is a rare congenital heart disease characterized by the discontinuity between the ascending aorta and the descending aorta or isthmus, described by Celoria and Patton [1]. The pathology accounts for about 1.5% of all congenital heart diseases. The mortality rate in the first year of life is about 90% if not treated surgically [2].

An interrupted aortic arch may be associated with other cardiac lesions such as ventricular septal defect, aortopulmonary window, truncus arteriosus, double outlet right ventricle, transposition of great arteriesand single ventricular physiology [3-8. Left ventricular outflow tract obstruction (LVOTO) is also common comorbidity in IAA [4,6,7]. Di George syndrome occurs in more than 25% of patients with IAA.

Many centersin the world have successfully performed one - stage surgical repair of IAA and associated lesions and have good long-term results [3-5]. LVOTO is considered one of the risk factors for mortality and reoperation [6-8]. In this study, we review our experience to assess the primary outcomes of one - stage total repair in patients with IAA at Children Heart Center - Vietnam National Children's Hospital.

II. PATIENTS AND METHODS

2.1. Patients

From January 2012 to September 2017, Children's HeartCenter - Vietnam National Children's Hospital performed the one - stage total repair in 27 patients with IAA. Information on clinical status, intracardiac and extracardiac lesions, surgical methods, and early mortality was retrospectively documented. Patients with univentricular physiology and transposition of great arteries were excluded. Patient information after hospital discharge is collected via regular follow - up.

The cardiac lesions were assessed before and after operation by echocardiography. Left ventricular outflow tract diameter less than the patient's weightplus 1.5 via the long - axis parasternal view on 2D ultrasonography was considered LVOTO.

Patients are follow - up regularly at our outpatient clinic at 1 month, 3 months, 6 months, 1 year and every year. The clinical examination, chest X-ray, echocardiogram and echocardiography were performed.

2.2. Operative techniques for aortic arch reconstruction

The operation was performed via the midsternal line sternotomy. The ascending aorta, the aortic arch with the brachiocephalic arteries, the pulmonary artery branch, the duc tusarteriosus, and the descending aorta were dissected to maximize the mobility of the aortic arch and decrease the risk of tracheal compression from the aorta. We used selective cerebral perfusion with moderate hypothermia (core temperature is 26 - 280C) via a Gore - tex shunt connected to the innominate artery or direct cannulation through the ascending aorta nearby the innominate artery. One patient required circulatory arrest to repair the aortic arch. Sometimes, a descending aorta arterial cannula was performed for lower body perfusion during cooling phase. Hypothermia was performed using cardiopulmonary bypass with aortic cannulation and bicaval cannulation. The mean lowest anal temperature is 28 ± 1Celsius. The pulmonary end of ductus arteriosus was ligated, and two branches of pulmonary arteries were encircled and occluded as soon as the CPB was initiated. When the temperature reached demand, cardioplegia was infused into the ascending aorta. The cannula placed via the ductus arteriosus was removed. The ductus tissues were mostly removed from the descending aorta. The aortic arch was opened longitudinal up to the ascending aorta. Theaorticarch and the descending aorta were anastomosed bydirect extended end - to - sideanastomosis or end - to - side anastomosis using continuous 7.0 - 8.0 polypropylene suture, sometimes added with an autologous patch from the pulmonary artery trunk. The total cardiopulmonary bypass was used after the aortic arch repair achievement, and the in tracardiac lesions were repaired subsequently.

Data were collected and analyzed using SPSS 20.0 software. Continuous variables were represented by mean, standard deviation, maximum and minimum values. Categorical variables were represented by percentage values. The study was approved by the Ethics committee of the Research Institute of Children's Health, National Children's Hospital. The patient inform consent has been waived due to retrospective study.

III. RESULTS

In 27 patients who underwent total repair, 14 were female, and 13 were male. The mean follow - up time was 24 ± 23 months. The mean age at surgery was 58 (median 53 days, 9 - 155 days), and the mean weight was 3.5 kg (2.3 - 6.5 kg). According to Celoria and Patton classification, there were 16 patients diagnosed with IAA type A (59.3%), and 9 patients have IAA type B (40.7%), no cases of type C. Associated lesions include ventricular septal defect (85.2%), LVOTO (22.2%), bicuspid aortic valve (25.9%), aortopulmonary window (14.8%), truncus arteriosus (11.1%), aberrant right subclavian artery (11.1%), right aortic arch (7.4%) **(Table 1)**.

Table 1: Associated lesions

ASSOCIATED INTRACARDIAC LESIONS	N (%)
IAA - TypeA - TypeB	16 (59.3%) 11 (40.7%)
Ventricular septal defect	23 (85.2%)
Left ventricular outflow tract obstruction	6 (22.2%)
Bicuspid aortic valve	7 (25.9%)
Aortopulmonary window	4 (14.8%)
Truncus arteriosus	3 (11.1%)
Aberrant right subclavian artery	3 (11.1%)
Right aortic arch	2 (7.4%)

The mean CPB and aortic cross-clamp time were 150 ± 38 minutes and 103 ± 25 minutes, respectively.

Selective cerebral perfusion was performed during the aortic arch repair, and the mean time of regional cerebral perfusion was 39 ± 12 minutes. One patient required circulatory arrest to repair the aortic arch **(Table 2)**.

Post - operation mean mechanical ventilation time was 77 hours (13 - 672 hours). Postoperative blood culture/endotracheal intubation was positive for pathogenic bacteria in 11 (40.7%) patients. Mild LVOTO or stenotic aortic valve after surgery was detected in 6 patients during follow - up period. Residual aortic arch stenosis was found in 19 patients: 18 with mild stenosis (< 20 mmHg) and 1 with moderate stenosis (< 30 mmHg).

Table 2: Perioperative characteristics

Perioperative characteristics	n (%) / X ± SD [min - max]
Bypass time	150 ± 38 [108 - 224]
Aortic cross clamp time	103 ± 25 [80 - 155]
Selective cerebral perfusion time	39 ± 12 [29 - 56]
Circulatory arrest	1
Aortic arch techniques	
Direct end - to - side anastomosis	26 (96.3)
Direct end - to - end anastomosis	1 (3.7)
Concomittant procedures	
Conal septum resection	4 (14.8)
Rastelli with valve conduit implantation	3 (11)
AP window closure	4 (14.8)
Complete AV block	1 (3.7)
Early death	6 (22.2)

Mortality:

The number of deaths was 6 (22.2%), with 5 patients (18.5%) dying early at the hospital or 30 days after discharge. In 5 early death, 4 patients had blood cultures or endotracheal cultures positive for bacteria before or after surgery, 1 patient died of an unknown cause. One patient, who underwent total repair IAA associated with truncus arteriosus, died 5 months after surgery; the mode of death was severe pulmonary arterial branches stenosis and pulmonary hypertension. The survival ratesat 30 days,1year, and 5 years were 81.5%, 77.8%, and 77.8%, respectively **(Figure 1).**

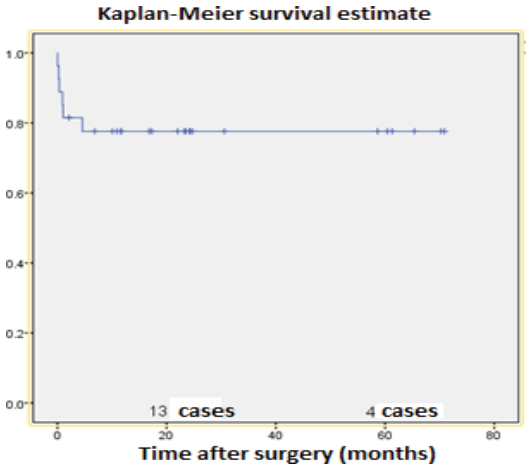

Figure 1: Survival estimated after one - stage repair for IAA

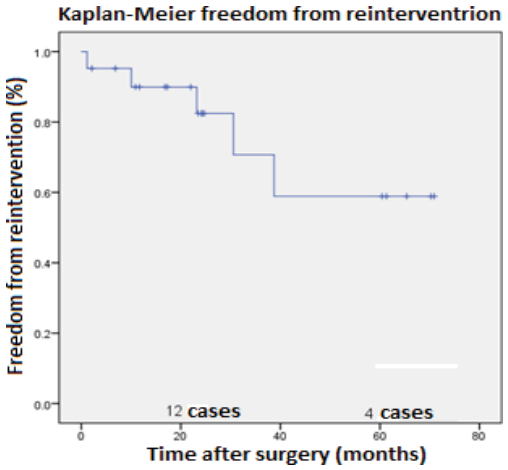

Figure 2: Freedom from reintervention after one - stage repair for IAA

In 21 survivors, 5 patients (23.8%) had reintervention. Three patients required reoperation due to LVOTO, 1 patient with recurrent aortic arch stenosis underwent balloon angioplasty, 1 patient underwent surgery for replacement of right ventricular - pulmonary conduit after total repair of IAA with truncus arteriosus. The rate of freedom from reintervention after surgery at 1 year, 2 years, 5 years, respectively 90%, 82.5%, and 59% **(Figure 2).**

IV. DISCUSSIONS

For many years, repairing IAA andintra - cardiac lesions concurrently was the method of choice Jonas et al. [9] reported a single - stage surgery with a satisfactory outcome (mortality at 5 years, 85%). Schreiberetal. [5] followed up on 94 patients who underwent one - stage surgery for 20 years, showing

a low early mortality rate (12%). The mortality of Children's Heart Center - National Hospital of Pediatrics is 22.2% higher than other centers; the main predictor for hospital death is the infection status before and after surgery, up to 11/27 (40.7%) patients have positive blood/endotracheal cultures for pathogenic bacteria. We have not ruled out infection due to prolonged mechanical ventilation or tracheal stenosis after aortic arch repair, especially in cases with the right aortic arch. However, we believe that the primary outcomes of this complex heart disease will be improved through the learning curve and the working condition (new facility, new equipment), especially with an early diagnosis for patients with duct - dependent congenital heart disease (including fetal diagnosis).

In the IAA cohort, the patients with diagnoses of truncus arteriosus and aortopulmonary window are considered the complex group owing to higher mortality and morbidity [8-10]. Considering the complexity of the surgical repair for IAA with truncus or AP window, some authors suggest staged repair with aortic arch reconstruction as the first stage, and intracardiac defect repair later when the patients get older and bigger [5,10]. However, the long - term mortality and morbidity are not different between the staged repair group and the one - stage repair group [7-9]. In our study, we aim to repair all intracardiac defects at the same time of operation because it provides easier postoperative management with normal circulation and predictive indicatorsfor perioperative care. This operative strategy is important for us due to lacking experience in palliative management at our center, especially with single ventricular physiology. We believe that after the learning curve, the primary outcomes of this complex congenital heart disease will be improved as the other centers in the world.

Although the results of primary repair surgery have improved in recent years, LVOTO is still a risk factor for reoperation [5]. LVOTO after IAA with ventricular septal defect is one of the most common reoperations during follow - up of patient's life [5-7]. The main cause of LVOTO is the posterior deviation of the infundibular septum. In our series, we try to resect the infundibular septum before VSD closure via tricuspid valve approach to diminish the possibility of LVOTO or prolongthe duration to reoperation, and avoid reoperation with complex operation (Ross - Konno operation, Yasui operation…) during infancy. In our study, 5 patients needed reintervention, including 3 patients due to stenosis of the left ventricular outflow tract. The surgical strategy which aims to alleviate the obstruction of LVOT by resection the conal septum in order to diminish the reoperation due to LVOTO in the future in patients with posterior deviation of the conal septum. However, a longer follow - up time with a biggernumber of patients is crucial to assess the safety of this strategy at our institution.

V. CONCLUSION

Primary outcomes of one - stage total repair for interrupted aortic arch at Children's Heart Center-National Hospital of Pediatrics are safeand results are encouraging. Longer follow - up with a bigger number of patients is essential.

REFERENCES

1. Celoria GC, Patton RB: Congenital absence of the aortic arch. Am Heart J 58:407, 1959
2. Van Praagh R, Bernhard WF, Rosenthal A, et al.: Interrupted aortic arch: Surgical treatment. Am J Cardiol 27:200-211, 1971
3. Hazekamp MG, Quagebeur JM, Singh S, et al. One stage repair of aortic arch anomalies and intracardiac defects. Eur J Cardiovasc Surg 1991;5:283-7.
4. Luciani GB, Ackermann RJ, Chang AC, Wells WJ, Starnes VA. One-stage repair of interrupted aortic arch, ventricular septal defect, and subaortic obstruction in the neonate: a novel approach. J Thorac Cardiovasc Surg 1996;111:348-58.
5. Christian Schreiber, Andreas Eicken, Manfred Vogt, Thomas Gunther. Repair of Interrupted Aortic Arch: Results After More Than 20 Years. Ann Thorac Surg 2000;70:1896-900.
6. Bove EL, Minich LL, Pridjian AK, et al. The management of severe subaortic stenosis, ventricular septal defect, and aortic arch obstruction in the neonate. J Cardiovasc Surg 1993;2: 289-96.
7. Fulton JO, Mas C, Brizard CPR, Cochrane AD, Karl TR. Does left ventricular outflow tract obstruction influence outcome of interrupted aortic arch repair? Ann Thorac Surg 1999;67: 177-81.
8. Brian W. McCrindle, Christo I. Tchervenkov, Igor E. Konstantinov, William G. Williams. Risk factors associated with mortality and interventions in 472 neonates with interrupted aortic arch: A Congenital Heart Surgeons Society study. J Thorac Cardiovasc Surg 2005; 129:343-50.
9. Jonas RA, Quaegebeur JM, Kirklin JW, et al.: Outcomes in patients with interrupted aortic arch and ventricular septal defect. J Thorac Cardiovasc Surg 107:1099-1113, 1994.
10. Yasutaka Hirata, MD, Jan M. Quaegebeur, MD, Ralph S. Mosca, et al.: Impact of Aortic Annular Size on Rate of Reoperation for Left Ventricular Outflow Tract Obstruction After Repair of Interrupted Aortic Arch and Ventricular Septal Defect. Ann Thorac Surg 2010;90:588-92.