

Early Outcome of Repair of Supra Cardiac Total Anomalous Pulmonary Venous Connection

Repair of Supra Cardiac Total Anomalous

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ABSTRACT

Objective: To discuss the results of supra cardiac TAPVC repair in our setup.

Study Design: Retrospective study

Place and Duration of Study: This study was conducted at the Pediatric Cardiac Surgery Department of Children's Hospital Lahore for a period of 03 years from January, 2019 to November, 2021.

Materials and Methods: Age, weight, presenting symptoms, operative details (like cross clamp time and cardiopulmonary bypass time), peri operative mortality, ICU stay, and complications were studied.

Results: From January 2019 to November 2021, 48 patients under went repair of supra cardiac TAPVC at children's hospital Lahore. Mean age was 13months, mean weight was 7kg, 9 (18%) patients died preoperatively. Ascending vertical vein was ligated in all cases, but in 2 patients we had to re-open the ascending vertical vein

Conclusion: With only a few centers in Pakistan doing infant cardiac surgery, early outcome after repair of supra cardiac TAPVC is reasonable in our set up, but still far worse than that of modern centers. Good patient selection, and post-operative management of pulmonary hypertension can result in better outcome. In this series we did not present the follow up of the patients, which is drawback of this article.

Key Words: Early Outcome, Supra Cardiac Total Anomalous, Pulmonary Venous Connection

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INTRODUCTION

The diagnosis of total anomalous pulmonary venous connection (TAPVC) is made when all four pulmonary veins drain anomalously to the right atrium or to a tributary of the systemic veins. It accounts for 1% to 1.5% of all congenital heart diseases. Supra cardiac TAPVC is the most common type of TAPVC that accounts for 45 to 55% of all TAPVCs.

Total anomalous pulmonary venous connection (TAPVC) is a cyanotic congenital heart disease in which all the pulmonary veins don't enter left atrium but rather into right atrium directly or indirectly through a tributary. It has 4 types (supracardiac, 45%; infracardiac, 25%; cardiac, 25%; mixed, 5%).¹To make life possible, right to left shunt through atrial septal defect is mandatory. Otherwise patient would die immediately after birth.²

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The most common type of TAPVC is supracardiac TAPVC³⁻⁶. The connection in supracardiac TAPVC is usually to a left vertical vein draining into the left brachiocephalic vein through which pulmonary venous blood reaches right atrium.

More than 75% of infants with supracardiac TAPVC would die in first year of life if left untreated⁷. Surgical repair should be performed at the time of diagnosis, as an elective procedure. Delaying the surgery leads to complication like right ventricle pressure and volume overload. In the last two decades, the improvement in surgical technique and improved postoperative management has resulted in improved survival of TAPVC patients,⁸ however, repair does remain a challenge with early mortality reported in the literature in the range of <10% to 50%. TAPVC in biventricular anatomy, with no concomitant intra cardiac anomaly has the best prognosis.^{9,10}

MATERIALS AND METHODS

It is a retrospective study. Files of all the patients who underwent supracardiac TAPVC repair at children's hospital Lahore in last 3 years (from January 2019 to November 2021) were reviewed. Age, weight, presenting symptoms, operative details (like cross clamp time and cardiopulmonary bypass time), peri operative mortality, ICU stay, and complications were studied.

Pre-operative evaluation: all the patients were admitted through OPD or emergency to cardiology/cardiac

surgery ward. They were discussed in the cath conference. Decisions to operate were made after reviewing the echo cardiogram. Where in doubt, CT Angio was performed to confirm the diagnosis. Patients with single ventricle were excluded from the study.

Surgical Strategy: Patients were taken to the operation room after taking written informed consent. Standard anesthesia was given. Under sterile conditions, median sternotomy was made. Patients were put on cardiopulmonary bypass with standard aorto bicaval cannulation. In most of the patients temperature was cooled to 28 degree centigrade. 2 patients required deep hypothermic circulatory arrest.

After clamping the aorta and DelNido cardioplegia in the aortic root, TAPVC was repaired with standard bi atrial approach. In 5 patients, suture less technique¹¹ was used. The sutureless technique involves anastomosing the pericardium to the pulmonary venous confluence and no direct suturing of pulmonary venous chamber to left atrium. In 3 patients superior approach¹² was used to repair TAPVC.

Post operatively patients were shifted to cardiac surgical ICU on mechanical ventilation. Epinephrine and milrinone infusions were started intra operatively and were continued in post-operative period. No patient received nitric oxide.

Operative mortality was defined as death within 30 days of an operation or within the primary hospitalization¹³.

RESULTS

48 patients under went repair of supra cardiac TAPVC at children's hospital Lahore.

Age: age of the patients ranged from 2 months to 12 years with the mean of 13 months.

Weight: weight ranged from 3.5 kg to 39 kg with the mean of 7 kg

Cyanosis, dyspnea, failure to thrive and multiple respiratory infections were the chief presenting complaints.

Table No.1: Demographical information

Total number	48
Age of the patients	Average 13 months (range 2 months – 12 years)
Weight of the patients	Average 7 kg (range 3.5kg - 39kg)
Obstruction present	5 patients
Mechanical ventilation	Mean 38hours (6 hours – 15 days)
ICU stay	Mean 70 hours (1- 30 days)

Cardiopulmonary bypass time: 45 minutes to 241 minutes, with the mean of 81 minutes.

Cross clamp time: 22 minutes to 184 minutes, with the mean of 62 minutes.

Concomitant surgical procedures: atrial septal defect (ASD) was present in all cases that was closed intra operatively. One patient had ventricular septal defect (VSD) that was closed concomitantly.

Ascending vertical vein was ligated in all cases but in 2 cases we had to re-open the vertical vein by removing the ligature. These 2 patients developed low cardiac output after coming off cardiopulmonary bypass but the hemodynamics improved when ascending vertical vein was re-opened.

Table No.2: Surgical Details

CPB time	Mean 81 minutes (45-241 minutes)
Aortic cross clamp time	Mean 61 minutes (22-184 minutes)
Sutureless technique	5
Superior approach	2
Standard bi atrial approach	41
Ascending vertical vein left open	2
Concomitant VSD closure	1
Concomitant PDA ligation	20
Deep hypothermic circulatory arrest	2

Mortality: 9 patients died (18.75%) . 8 out of 9 patients were having weight <5 kg. 3 patients died due to low cardiac output. 5 died because of persistent pulmonary hypertension, prolonged mechanical ventilation and multi organ failure. 1 died of post-operative bleeding.

Mechanical Ventilation: Patients remained on mechanical ventilation for 6 hours to 15 days with the mean of 38 hours.

ICU stay: Average ICU stay was 70 hours (1 – 30 days)

Complications: Table 3 gives the details of all the complications that occurred in post-operative period.

Table No.3: Complications

Mortality	9 (18.75%)
Post-operative bleeding	1
Pulmonary hypertensive crisis	5
Low cardiac output	3
Pneumothorax	1
Renal failure requiring dialysis	2
Stroke	1

DISCUSSION

Congenital heart disease (CHD) is the most common birth defect.¹⁴ It is estimated that every year 42000 babies are born with CHD in Pakistan.¹⁵

Children's hospital Lahore is one of the largest pediatric cardiac surgery centers in Pakistan. We are doing > 1000 pediatric cardiac surgeries per year. In last 3 years we did 48 supra cardiac TAPVC repairs.

Mortality: Although there are few series which had mortality of 3%^{16, 17} other centers reported that repair of

TAPVC carried a relatively high early mortality (10–20 %) ^{18,19}. Postoperative care for TAPVC is based mostly on preventing or treating pulmonary hypertension and maintaining systemic cardiac output. Ideally, the pulmonary artery pressure will decrease to less than half of the systemic pressure soon after separating from cardiopulmonary bypass. ²⁰ Nitric oxide and milrinone are used to treat pulmonary hypertension ²¹

Keeping the Vertical Vein Open: Whether the vertical vein should be ligated repair is controversial. Our strategy is to keep the vertical vein open if there is low blood pressure after turning off cardiopulmonary bypass. Chowdhury et al recommended some adjustable devices to close the vertical vein ²². Vertical vein ligation is advisable to close any left to right shunt, but in relatively small left ventricle, it can be left open ²³. Transcatheter vertical vein closure might render patients with high LAP suitable candidates for further hybrid approach.

Risk Factors: Weight less than 5 kg, pre-operative pulmonary venous obstruction, and persistent pulmonary hypertension were identified as risk factors.

CONCLUSION

With only a few centers in Pakistan doing infant cardiac surgery, early outcome after repair of supra cardiac TAPVC is reasonable in our set up, but still far worse than that of modern centers. Good patient selection, and post-operative management of pulmonary hypertension can result in better outcome. In this series we did not present the follow up of the patients, which is drawback of this article.

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Conflict of Interest: The study has no conflict of interest to declare by any author.

REFERENCES

- Kanter KR. Surgical repair of total anomalous pulmonary venous connection. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu* 2006; 40-4.
- Stein P. Total anomalous pulmonary venous connection. *AORN J* 2007;85:509–20.
- Shi G, Zhu Z, Chen J, Ou Y, Hong H, Nie Z, et al. Total anomalous pulmonary venous connection: the current management strategies in a pediatric cohort of 768 patients. *Circulation* 2017;135(1): 48–58.
- Harada T, Nakano T, Oda S, Kado H. Surgical results of total anomalous pulmonary venous connection repair in 256 patients. *Interact Cardiovasc Thorac Surg* 2018.
- Chowdhury UK, Airan B, Malhotra A, Bisoi AK, Saxena A, Kothari SS, et al. Mixed total anomalous pulmonary venous connection: anatomic variations, surgical approach, techniques, and results. *J Thorac Cardiovasc Surg* 2008;135(1):106–16.
- Sakamoto T, Nagashima M, Umezu K, Houki R, Ikarashi J, Katagiri J, et al. Long-term outcomes of total correction for isolated total anomalous pulmonary venous connection: lessons from 50-years' experience. *Interact Cardiovasc Thorac Surg* 2018;27(1):20–6.
- Burroughs JT, Edwards JE. Total anomalous pulmonary venous connection. *Am Heart J* 1960; 59:913-31.
- Zhao K, Wang H, Wang Z, Zhu H, Fang M, Zhu X, Zhang N, Song H. Early and intermediate-term results of surgical correction in 122 patients with total anomalous pulmonary venous connection and biventricular physiology. *J Cardiothorac Surg* 2015;10:172.
- Hancock Friesen CL, Zurakowski D, Thiagarajan RR, Forbess JM, del Nido PJ, Mayer JE, et al. Total anomalous pulmonary venous connection: an analysis of current management strategies in a single institution. *Ann Thorac Surg* 2005;79: 596-606.
- Morales DL, Braud BE, Booth JH, Graves DE, Heinle JS, McKenzie ED, et al. Heterotaxy patients with total anomalous pulmonary venous return: improving surgical results. *Ann Thorac Surg* 2006; 82:1621-7.
- Menon S, Mathew T, Karunakaran J, Dharan BS. Modified closed chamber sutureless technique for anomalous pulmonary venous connection. *Ann Pediatr Cardiol* 2017;10(1):58-60.
- Okonta KE, Agarwal V, Abubakar U. Superior repair: A useful approach for some anatomic variants of total anomalous pulmonary venous connection. *Afr J Paediatr Surg* 2013;10(2):131-4.
- Van Praagh R, Harken AH, Delisle G, Ando M, Gross RE. Total anomalous pulmonary venous drainage to the coronary sinus. A revised procedure for its correction. *J Thorac Cardiovasc Surg* 1972; 64:132-5.
- Pate N, Jawed S, Nigar N, Junaid F, Wadood AA, Abdullah F. Frequency and pattern of congenital heart defects in a tertiary care cardiac hospital of Karachi. *Pak J Med Sci* 2016;32:79-84.
- Christianson A HC, Modell B. *March of Dimes. Global report on birth defect. The hidden toll of dying and disabled children.* New York; 2006.

16. Jang SI, Song JY, Kim SJ, Choi EY, Shim WS, Lee C, Lim HG, Lee CH. The recent surgical result of total anomalous pulmonary venous return. *Korean Circ J* 2010;40(1):31-5.
17. Liufu R, Shi G, Zhu F, Guan Y, Lu Z, Chen W, Zhu Z, Chen H. Superior Approach for Supracardiac Total Anomalous Pulmonary Venous Connection. *Ann Thorac Surg* 2018;105(5):1429-1435.
18. Michielon G, Di Donato RM, Pasquini L, Giannico S, Brancaccio G, Mazzer E, et al. Total anomalous pulmonary venous connection: long-term appraisal with evolving technical solutions. *Eur J Cardiothorac Surg* 2002;22:184-91.
19. Kirshbom PM, Myung RJ, Gaynor JW, Ittenbach RF, Paridon SM, DeCampli WM, et al. Preoperative pulmonary venous obstruction affects long-term outcome for survivors of total anomalous pulmonary venous connection repair. *Ann Thorac Surg* 2002;74:1616-20.
20. Kaiser L, Kron I, Spray T. *Mastery of cardiothoracic surgery*. Lippincott Williams & Wilkins, Philadelphia; 2007.
21. Franco K, Thourani V. *Cardiothoracic surgery review*, 1 Har/Psc edn. Lippincott Williams & Wilkins, Philadelphia; 2011.
22. Chowdhury UK, Subramaniam KG, Joshi K, Varshney S, Kumar G, Singh R, et al. Rechanneling of total anomalous pulmonary venous connection with or without vertical vein ligation: results and guidelines for candidate selection. *J Thorac Cardiovasc Surg* 2007;133:1286-1294.
23. Cope JT, Banks D, McDaniel NL, Shockey KS, Nolan SP, Kron IL. Is vertical vein ligation necessary in repair of total anomalous pulmonary venous connection? *Ann Thorac Surg* 1997;64:23-8.