Original article

Clinical profile of patients with Total Anomalous Pulmonary Venous Return and their short term outcome in pediatric cardiac centre at Dhaka Shishu Hospital

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Background: Total anomalous pulmonary venous return (TAPVR) is an uncommon congenital cardiovascular anomaly with poor natural prognosis without proper intervention. It has been detected more frequently in recent year due to the advent of echocardiography. The aim of this study is to evaluate the clinical manifestations, age at diagnosis and short term outcomes in TAPVR patients. ***Methods:*** From 1st January 2013 to 31st December 2013, a total of 34 cases with TAPVR were admitted in pediatric cardiac centre at Dhaka Shishu Hospital, Dhaka, Bangladesh. All of them were evaluated with 2-dimensional (2-D) and color Doppler echocardiography examination. CXR and ECG were also done. Patient's sex, age at diagnosis, types of TAPVR, clinical manifestations, radiological finding, ECG findings and outcomes were compiled and analyzed. ***Results:*** In 34 patients with TAPVR, 23 (67.6%) were male and 11 (32.4%) were female with male to female ratio of 2.09:1. Most of the patients were diagnosed between 0-6 months of age that is 13 (38.2%) cases were in 0-2 month's age group, 14 (41.2%) cases were in more than 2 month's to 6 month's age group. Tachypnea and cyanosis were more common symptoms. The types of TAPVR was supra-cardiac 18 (52.9%), cardiac 11 (32.4%), infra-cardiac 3 (8.8%) and mixed in 2 (5.9%) cases. Pulmonary hypertension was present in 31 (91.2%) of 34 cases. Among them, 20 (58.8%) patients had severe pulmonary hypertension. The most common associated intra-cardiac lesions of TAPVR patients were ASD 13 (38.2%) and PFO 13 (38.2%). ECG findings of TAPVR, 18 (52.9%) patient had right axis deviation (RAD), right ventricular hypertrophy (RVH) and 14 (41.2%) had right axis deviation (RAD), right ventricular hypertrophy (RVH), right atrial enlargement (RAE). X-ray findings of TAPVR patients, 32 (94.1%) patients had Cardiomegaly and increased pulmonary vascularity. Among admitted patient, 3 (8.8%) patients died due to pneumonia and intractable heart failure, 31 (91.2%) patients referred to advanced cardiac centre for operative treatment. ***Conclusions:*** Tachypnea and Cyanosis were an obvious clinical symptom of TAPVR. 2-D and color Doppler echocardiography can provide quick and accurate diagnostic information of TAPVR. Death rate is high in TAPVR patient in spite of adequate medical management. So, early detection and definitive surgical treatment of TAPVR is much needed.

Keywords: total anomalous pulmonary venous return; supra-cardiac TAPVR; cardiac TAPVR; infra-cardiac TAPVR; mixed type TAPVR

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Introduction

Total anomalous pulmonary venous return (TAPVR) is a rare congenital anomaly, corresponding to approximately 2% of all congenital heart defects[1]. A recognized classification divides TAPVR into four groups according to the site of connection[2]. With type I, having a supracardiac connection (50%), the common pulmonary trunk joins the left vertical vein, the innominate vein, or the superior vena cava. With type II, having a cardiac connection (30%), the

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anomalous pulmonary venous drainage enters the coronary sinus or flows directly into the right atrium. With type III, the site being infracardiac (15%), connection occurs to the portal vein, venous duct, or inferior vena cava below the diaphragm. With type IV, a mixed site variety (5%), the anomalous venous return occurs at several levels [1]. Pathophysiologically, these four types are sub classified according to whether the pulmonary venous return is obstructed or non-obstructed. The clinical presentation and prognosis are different for the two later conditions, being poorer for the obstructed type. Although obstruction may occur with any anatomic type of TAPVR, the highest incidence is encountered with the infra-cardiac type[2]. Mortality is high in TAPVR patient and it is associated with the severity of obstruction in pulmonary venous drainage, the age at presentation (older the age less favorable the outcome), the presence of pulmonary hypertension and associated cardiac anomalies and in developing countries malnutrition and sepsis are associated with increased mortality and morbidity.[3-5] The diagnosis was suggested by echocardiography and confirmed by catheter-angiography [3] and magnetic resonance imaging [4] which allowed definition of the anatomy.

Materials and Methods: A prospective study was conducted in pediatric cardiac centre at Dhaka shishu hospital from 1st January 2013 to 31st December 2013. Dhaka Shishu Hospital which is a tertiary care hospital having specialized Pediatric cardiac centre and Color Doppler echocardiography facilities. Informed written consent was taken from all enrolled patient's parents or attendants. All the children who diagnosed as total anomalous pulmonary venous return after admitted in pediatric cardiac centre at Dhaka shishu hospital during these periods were included in this study. After admission of patient, detailed history, physical examination done and diagnosis confirmed by Chest x-ray, ECG and Color Doppler Echocardiography. During study period, total 34 TAPVR patients were included in this study. During echocardiography, Phenobarbitone (10mg/kg) administered intravenously for sedation. Suprasternal, parasternal, apical, and subcostal views were employed to explore the presence of TAPVR. According to the different drainage sites of the anomalous pulmonary veins to the heart, TAPVR is classified into 4 types: supra-cardiac type (anomalous pulmonary vein draining into upper part of heart), intra-cardiac type (anomalous pulmonary vein draining into middle part of heart), infra-cardiac type (anomalous pulmonary vein draining below the heart), and mixed type (which has two or more drainage sites). Pulmonary hypertension is defined as a mean pulmonary artery pressure (PAP) greater than 25 mmHg at rest or pulmonary artery systolic pressure (PASP) greater than 35 mmHg at rest. Interpretation of ECG that is RAD considered when downwards deflection of QRS complex at Lead 1 and upwards deflection of QRS complex at AVF, Tall R wave in V_1 greater than 7 considered RVH, tall P >2.5 mm in lead 11 considered RAE. Cardiomegaly is considered when cardiothoracic ratio >60% and plethoric lungs field considered when pulmonary vascularity visible >2/3 of lungs field. The patient's sex, age at diagnosis, types of TAPVR, clinical manifestations, associated intra-cardiac lesion, radiological, ECG findings and short term outcomes of these patients were entered into and descriptive statistical analysis done by SPSS statistical software (version 21).

Results:

In 34 patients with TAPVR , 23 (67.6%) were male and 11 (32.4%) were female with male to female ratio of 2.09 : 1. So, male patients outnumbered female patients.

Table -1 :Types of TAPVR (n=34)

Types of TAPVR	Frequency	Percentage
Supra cardiac	18	52.9
Cardiac	11	32.4
Infra cardiac	3	8.8
Mixed	2	5.9

32 (94.1%) patient had tachypnea, 15 (44.1%) had cyanosis, 13 (38.2%) had recurrent respiratory tract infection, 4 (11.8%) had congestive heart failure and

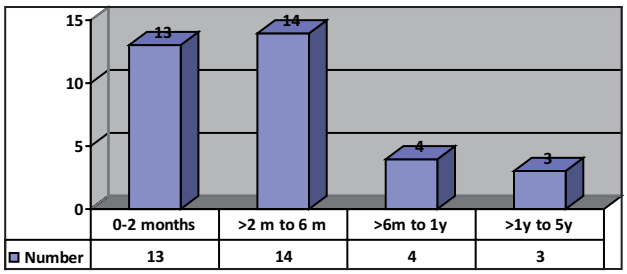

Figure-1:Age distribution of patients (n =34)

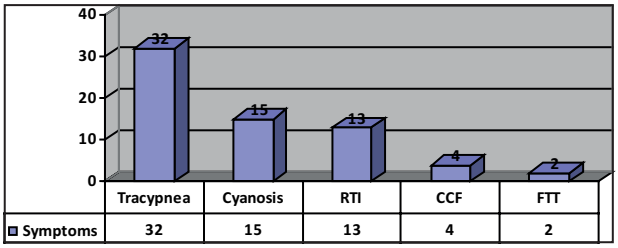

Figure – 2:Presenting complaints of TAPVR (n=34)

2 (5.9%) patient presented with failure to thrive.

Table – 2: Associated intracardiac lesions of TAPVR (n=34)

Intra-cardiac lesion	Frequency	Percentage
ASD	13	38.2
PFO	13	38.2
Complete AV canal defect	1	2.9
PDA and PFO	3	8.8
ASD and PS	2	5.9
ASD and PDA	1	2.9
PFO and PS	1	2.9

Table – 3 : Types of pulmonary hypertension of TAPVR (n=34)

Types of pulmonaryhypertension	Frequency	Percentage
Severe	20	58.8
Moderate	7	20.6
Mild	4	11.8
No	3	8.8

Table–4: ECG findings of TAPVR patient (n=34)

	Frequency	Percent
RAD, RVH	18	52.9%
RAD, RVH, RAE	14	41.2%
No change	2	5.9%

Table – 5: X-ray findings of TAPVR (n=34)

	Frequency	Percent
Cardiomegaly and increased pulmonary vascular marking	32	94.1%
No change	2	5.9%

Table – 6: Out come of patient (n=34)

	Number	Percentage
Died	3	8.8%
Referred	31	91.2%

Discussion: TAPVR is a rare congenital heart defect [5,6] if left untreated 80% of the patients die in first year of life.[7] However it is not unusual to find few patients surviving into adulthood especially in a developing country where antenatal screening and routine checkup is done infrequently. The factors favoring survival into adulthood are a large ASD and non obstructed drainage through a short route.[8] As the advent of echocardiography, TAPVR can be readily diagnosed without much difficulty. The sensitivity and specificity for diagnosis by echocardiography including cross-sectional and color Doppler flow mapping have been reported to be up to 97% and 99%, respectively. [9-12] The symptoms and signs of TAPVR are variable, usually depending on the pathological anatomy and the changes of hemodynamics. Tachypnea, difficult feeding and cyanosis are usually the initial symptoms.[13] Repeated respiratory tract infection, and failure to thrive are also present. More than 94% of our TAPVR patients had tachypnea initially, but 39% of them had frequent respiratory tract infection. This findings are similar with Ping-YW et al.[14] Regarding sex of patient in our study, male patient two times more than female. This findings are similar with Mohammad AK et al [15] and Meng LL et al [16] but Ping- YW et al [14] showed male female ratio almost equal. we found supra-cardiac type of TAPVR 53%, cardiac type of TAPVR 32%, infra-cardiac type of TAPVR 9% and mixed type of TAPVR (6%). These findings are similar with other study that is Jensen et al [17] showed, supra-cardiac type of TAPVR 52%, cardiac type of TAPVR 30%, infra-cardiac type of TAPVR 12%, mixed type of TAPVR (6%). and Delisle et al [18] found supra-cardiac type of TAPVR 45%, cardiac type of TAPVR 26%, infra-cardiac type of TAPVR 24%, mixed type of TAPVR (5%). Keith et al [19] found supra-cardiac type of TAPVR 45%, cardiac type of TAPVR 30%, infra-cardiac type of TAPVR 18% and mixed type of TAPVR (7%). The most frequently associated intra-cardiac anomalies of our study are ASD and PFO followed by PDA, Pulmonary stenosis and Complete AV canal defect but Jong LR et al [20] study showed PDA was the most frequently associated intra-cardiac anomaly followed by Pulmonary stenosis, Single atrium and Single ventricle. Among 34 patients with TAPVR, 31 patients had variable type of hypertension. Gathman and Nadas [13] had reviewed 75 pediatric patients with TAPVR and found that three-fourth patients with markedly elevated pulmonary artery pressure. Their findings were also confirmed by Delisle *et al.*[18] Chest roentgenographic findings were cardiomegaly and increased pulmonary vascularity in 32 cases (94.1%). Jong LR et al [20] findings were, Of the 25 cases with TAPVR, cardiomegaly were in 22 cases (88%) and increased pulmonary vascularity in 23 cases (92%). The ECG findings showed right axis deviation (RAD), right ventricular hypertrophy (RVH) in 18 (52.9%) cases; right axis deviation (RAD), right ventricular hypertrophy (RVH) and

right atrial enlargement (RAE) in 14 (41.2%) cases. Our findings are similar with Jong LR et al [20] were, Of the 25 cases with TAPVR right axis deviation (RAD) in 22 cases (88%), right atrial enlargement (RAE) in 13 cases (52%), right ventricular hypertrophy (RVH) in 24 cases (96%). Among admitted patients, 3(8.8%) patients died due to pneumonia and intractable heart failure. 31 (91.2%) patients referred to advanced cardiac centre for operative treatment. Ping YW et al [14] Study showed 3 cases (9.2%) died during medical treatment due to pneumonia and intractable heart failure. The natural course of TAPVR is unfavorable because of progressing pulmonary artery hypertension and heart failure. Open heart repair is necessary in most of the cases to resolve this lethal anomaly. The estimated first year survival rate in patients without treatment was only 25%; 50% death occurred before 3 months of age, and 80% death occurred before 1 year of age.[21] However, the operation mortality in patients under 1-year of age decreased significant from 50% in 1970s to 30% after 1970s.[22,23]

Conclusion: Total anomalous pulmonary venous return (TAPVR) is a rare congenital anomaly. Tachypnea and Cyanosis were an obvious clinical symptom of TAPVR. Death rate is high in TAPVR patient in spite of adequate medical management. So, early detection of TAPVR and referral for surgical treatment can reduce death rate of TAPVR patient.

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