Successful surgical correction of total anomalous pulmonary venous connection
(T APVC Type -I) : A case report

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Summary
A twelve years old female girl presented to us with the complaints of exertional dyspnoea and palpitation, occasional fever and cough since her early childhood. These symptoms were exaggerated for the last one year. Her parents gave history of bluish discolouration of lips and tongue during crying and exposure to cold in her early childhood. Echocardiography and cardiac catheterization was done. Initially it was diagnosed as a case of atrial septal defect (ASD) secundum variety. Before operation re-evaluation by echocardiography showed that it was a case of total anomalous pulmonary venous connection (T APVC Type- I). She was operated under cardiopulmonary bypass with hypothermia and low flow perfusion with antegrade intermittent cold blood cardioplegic arrest. Anastomosis between the common venous sinus with the left atrium, closure of ASD with pericardial patch and ligation of the vertical vein was done. Pre-operatively her arterial oxygen saturation (SPO$_2$) was 90% Post operatively just before discharge it was 100%. She un-eventfully recovered from operation. This cyanotic congenital heart disease is an uncommon anomaly which frequently confuses with ASD (Secundum). It bears a relatively favorable prognosis after surgical repair.

Key words
TAPVC, surgical correction, case report

Introduction
The term TAPVC describes the anomaly in which the pulmonary veins have no direct communication with the left atrium. Instead, they connect to the right atrium or to one of the systemic veins. In 1957, Darling and associates divided the anomaly into four subtypes that describe the anatomic corrections of the pulmonary venous to the systemic venous circulation. Type I (supracardiac), type II (cardiac), type III (infracardiac), type IV (Mixed). Associated cardiac defects are: ASD, nearly in all cases, Patent ductus arteriosus in 25 to 50%, asplenia syndrome. Cooley and Ochsner (1957) reported the first open heart correction using cardiopulmonary by pass in a patient with a supracardiac anomaly.

Figure: Type I-TAPVC  
Figure: Type 11-TAPVC

Figure : Type III-TAPVC
Case Report

A 12 years old female child presented with the history of exertional dyspnoea and palpitation, occasional cough with fever since her early childhood. These symptoms were exaggerated for the last one year. Her parents give history of bluish discolouration of her lips, tongue and whole body during crying and after exposure to cold. Other members of the family were healthy. She was ill looking cachectic, mildly cyanotic (central & peripheral), her height- 144 cm, body weight-23 kg, with engorged neck vein, mild hepatomegaly. Left lower parasternal heave was present. There was a systolic murmur in the pulmonary area with loud second heart sound. Both the lung fields were clear on auscultation. All routine investigations except electrocardiography (ECG) & chest postero-anterior (CXR-PA) view were within normal limit. Her blood group was B+ve. Her arterial saturation of oxygen (SAO₂) determined by oxymetry probe (SPO₂) was 90% pre-operatively. Pre-operative echocardiographic diagnosis was atrial septal defect (ASD), secundum variety by two cardiologist. Cardiac catheterization was done which showed oxygen saturation at different chambers of the heart in favour of ASD (Secundum). ECG showed right ventricular hypertrophy (RVH) with right axis deviation. CXR PIA view showed heart was enlarged in transverse diameter with a supracardiac shadow merging with the cardiac shadow. Lung fields were plethoric. Echocardiography showed pulmonary hypertension. Suspicious x-ray and echo findings led us to re-evaluate the case by different cardiologist. This time the echocardiography report was different. They diagnosed it as a case of TAPVC with ASD and PH. Four pulmonary veins had opened into the common venous sinus posterior to left atrium (LA), which had drained into left innominate vein via a vertical vein. There was an ASD secundum with right to left shunt. They could not find one pulmonary vein.

With this echo findings we planned to operate her under hypothermic low flow cardiopulmonary bypass (CPB). Mid sternotomy was done. The superior vena cava (SVC) left innominate vein was found dilated. There was a thrill over the SVC. A common venous sinus was found posterior to the left atrium. CPB was established by selective SVC, inferior venacava (IVC)+Aortic cannulation. It was found that all four pulmonary veins opened into the common venous sinus and drained into the left innominate vein via a left vertical vein. There was an ASD secundum. So our per-operative diagnosis was total anomalous pulmonary venous connection (TAPVC) type I with ASD (secundum) with pulmonary hypertension (PH). Anastomosis between left atrium and common venous sinus was done with 7/0 proline suture. Atrial septal defect was closed by pericardial patch with continuous proline suture. Left vertical vein was ligated by no -2 silk. The whole operation was done under CPB with antegrade intermittent cold blood cardioplegic arrest of the heart.

In the intensive care unit (ICU) patient was ventilated overnight with dobutamine and glycercyl trinitrate support. Patient was weaned from ventilator and extubated in the following morning. She was discharged on 10th post operative day(POD) with an advice for further follow up 3 weeks later. On discharge her pulse rate was 96/min, BP 99/55 (69) mm Hg and SPO2-100%.
Case Report

Discussion
TAPVC is an uncommon congenital cardiac malformation, constituting <1% of all congenital heart disease. The majority of cases present at birth or in infancy, although survival into adulthood is reported. Uncorrected TAPVC carries >80% mortality in the first year of life from progressive cyanosis and congestive heart failure. However, recent improvements in surgical technique and peri-operative management of TAPVC have had a substantial impact on reducing the morbidity and mortality of even the most critically ill neonates. Pre-operative perfect diagnosis is an important issue. In this case we are able to diagnose the case almost precisely. So we could plan our operation and we were able to come out successfully.

Conclusion
TAPYC is seldom found at National institute of cardio vascular diseases (NICYD) for operation. They are commonly confused with ASD (secundum) in inexperienced hand. Except this case twelve other TAPYC cases were operated at NICYD, Dhaka in the past. But all of them were misdiagnosed pre-operatively as ASD (secundum), they were diagnosed on the operation table by the cardiac surgeon. This is for the first time at the NICYD that a r APYC was diagnosed preoperatively Simple closure of the ASD without surgical correction of the anomaly In a case of rAPYC is life threatening. Proper and meticulous pre-operative assessment of the patient can save us from misdiagnosis of this disease. So we must be very careful in diagnosing the case both pre-operatively and peroperatively to save the life of patients.

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References