Case Report

MICS – SupraCardiac TAPVC Repair, via Vertical Midaxillary Right Thoracotomy: First Case Reporting from Bangladesh

Mohammad Eliyas Patwary

Department of Pediatric Cardiac Surgery, National Heart Foundation Hospital & Research Institute, Dhaka, Bangladesh.

Abstract

A 3 year 5months old girl, diagnosed case of supracardiac total anomalous pulmonary venous connection (TAPVC) with large atrial septal defect (ASD) admitted for surgical repair. Minimal invasive cardiac surgery (MICS) via Vertical Midaxillary Right Thoracotomy (VMRT) was done on 20th March 2025 and discharged on 5th post operative period with stable hemodynamics. At six month follow up echocardiogram and check Computed tomography (CT) angiography showed unobstructed pulmonary venous flow to left atrium.This procedure provides good early recovery and highly parent satisfaction.

Key words: MICS, complex congenital, TAPVC (supra-cardiac)

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Introduction

Minimal invasive cardiac surgery (MICS) in pediatric cardiac surgery is the topmost demanding option especially from parents of girl kids [1]. In our experience almost all type of atrial septal defect (ASD), ventricular septal defect (VSD), partial anomalous pulmonary venous drainage (PAPVD), some selected tetralogy of Fallot's (TOF) operated with excellent short-term outcome. The first case report pertaining minimally invasive approach was described for the supracardiac TAPVC by Amirghofran and Mohammad Shakiba [2]. For the first time we did this case for the request from parents and get confidence from the reported case video[2]. This case report describes the supracardiac total anomalous pulmonary venous connection (TAPVC) via MICS. To the best our knowledge this is the first report to the particular type of operation in Bangladesh.

Case presentation

A 3 years 5 months mildly cyanotic girl of 11.2kg body weight with 94 cm height. At admission her room air SPO2 was 90%, pulse 92beats/min, noninvasive blood pressure was 104/89(96) mmHg, no organomegaly within normal all other systemic examinations. Echo and computed tomography (CT) angiography revealed non-obstructed supracardiac TAPVC with large ASD with mild pulmonary hypertension.

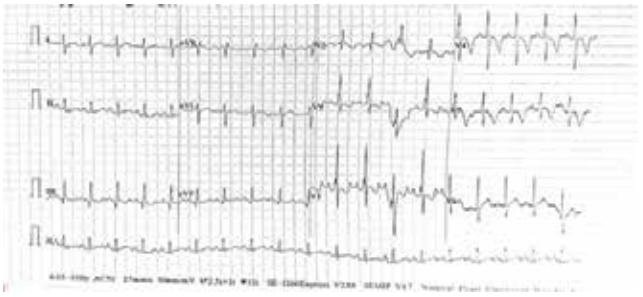


Figure 1: Preoperative electrocardiography



Figure 2: Preoperative chest X-ray- posterior/anterior view

Corresponding Author
Dr. Mohammad Eliyas Patwary,
Senior Consultant and Associate Professor, Departmentof Pediatric Cardiac Surgery, National Heart Foundation Hospital &Research Institute, Dhaka, Bangladesh,
Email"patwarye@yahoo.com

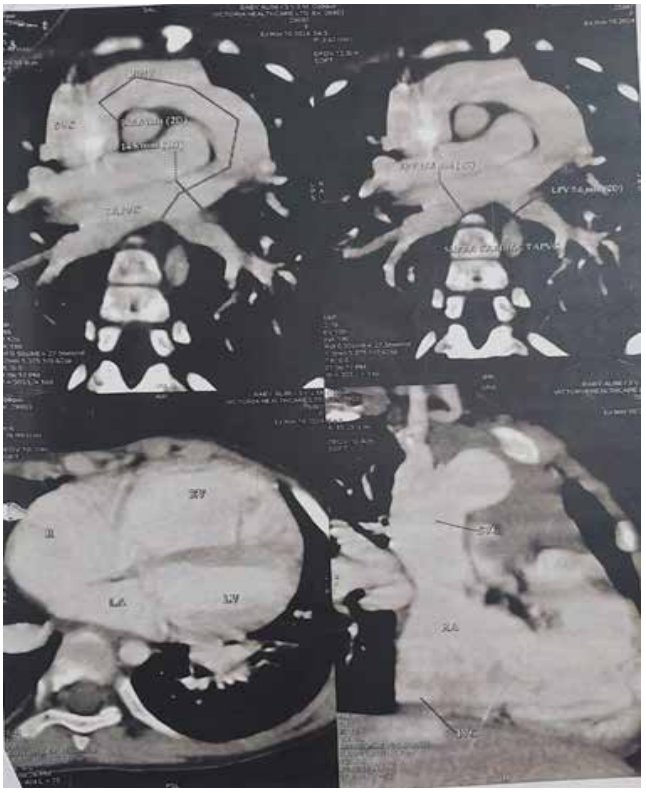


Figure 3: Preoperative computed tomography (CT) angiography

The procedure was performed though a 5cm vertical midaxillary right thoracotomy incision, which is the routine approach for minimal invasive cardiac surgeries in the pediatric setup of this tertiary care hospital of Bangladesh. After muscle sparing thoracotomy right lung was retracted with wet gauze, pericardium was opened parallel to right phrenic nerve and stay suture applied. After full heparinization central aortic and bi-caval canulation established CPB on mild hypothermia. At this stage Vertical veins are gentle dissection and taking control with silk snare (Figure 4). The left sided pleura and phrenic nerve was meticulously handled to make it intact, del Nido cardioplegia used for arrest the heart and right side of atrium was dissected to make well visualization of posterior common chamber behind the atriums. A horizontal incision on the rt atrium extended to left atrium via ASD, a large opening was formed in the posterior wall of left atrium. The opening site on the common chamber was stay suture applied and cut. A wide anastomosis was achieved between posterior wall of left atrium and the common chamber using 7-0 proline suture. This Vertical right midaxillary thoracotomy approach makes the anastomosis easier and less time consuming.

The ASD was closed with Dacron patch keeping a small 4mm fenestration at its center for overcoming postoperative pulmonary hypertensive crisis. Right atrium closed. Aortic cross clamp removed, weaned off Cardiopulmonary bypass. Vertical vein that was previously snared ligated with silk. Chest was closed and shifted to intensive care unit, extubated on first postoperative day and discharge on 5th post operative day with stable hemodynamics.7 days, 1 month, 3 months post operative follow up with ECG (Sinus rhythm), Chest X-ray (normal findings), Echocardiography showed normal pulmonary venous drainage to left atrium, a PFO with left to right shunt, normal pulmonary artery pressure.

After six months we did a CT angiography of heart and great vessels (Figure 5), showed three dimensional views of all pulmonary veins with its drainage to left atrium with excellent orientation.



Figure 4: Operative views though vertical right midaxillary thoracotomy approach

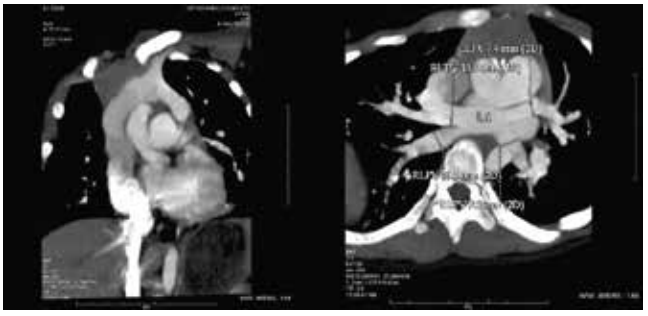


Figure 5: Computed tomography (CT) angiography after 6 months.

Discussion

Total anomalous pulmonary venous connection (TAPVC), also known as total anomalous pulmonary venous return, represents 1%-5% of cardiovascular congenital anomalies [3]. It is a cyanotic congenital heart defect in which all pulmonary veins, carrying oxygenated blood from the lungs, incorrectly drain directly or indirectly into the right atrium instead of the left. This results in oxygenated blood bypassing the left atrium, returning to the right side of the heart [4]. The malformation usually presents itself accompanied by neonatal cyanosis and tachypnea [5] and if a systemic-pulmonary shunt (right to left heart shunt) is not performed, it may be a cause of considerably mortality [3]; typical shunts occur by an atrial septal defect (patent foramen ovale) or patent ductus arteriosus (less common), with these communications being almost obligatory for survival [3]. TAPVC is a consequence of the persistence of the primitive connections of the pulmonary veins with the cardinal systemic veins, which results in a failure of common pulmonary veins to connect to the left atrium [6,7].

Craig et al. [8] described 4 types of TAPVC based on the site of pulmonary venous connection: supracardiac (type I), cardiac (type II), infracardiac (type III), and mixed connection (type IV). The supracardiac type is the most common, and it accounts for 44% of cases, followed by the infracardiac (26%), cardiac (21%), and mixed type (9%) [3].

In the supracardiac type, the pulmonary veins connect abnormally to the left innominate vein, superior vena cava (SVC), or azygos vein via an ascending vertical vein [4]. In the cardiac type, the pulmonary venous confluence drains into the coronary sinus, or, in rare cases, individual pulmonary veins drain directly into the right atrium [9]. In the infracardiac type, pulmonary veins form a descending vertical vein that drains below the diaphragm into the portal vein, hepatic vein, or inferior vena cava (IVC). The mixed type involves combinations of connections at different anatomical levels [10].

Supracardiac TAPVC is typically diagnosed in infancy due to severe symptoms like cyanosis, respiratory distress, recurrent chest infections, and failure to thrive [11]. The clinical presentation is different if the pulmonary venous connection is unobstructed (heart failure, mild cyanosis) or obstructed (respiratiory failure, severe heart failure) [12]. Echocardiography plays a pivotal role in diagnosing TAPVC, offering detailed visualization of abnormal pulmonary vein connections and their hemodynamic consequences [4]. The enlargement of the right atrium and ventricle along with right ventricular hypertrophy, suggested chronic volume overload and pulmonary hypertension [13]. CT angiography and MRI provided complementary anatomical and functional data crucial for surgical planning [4]. Three-dimensional reconstructions from CT were particularly useful in mapping the vertical vein's course and its connection to the systemic venous system. MRI can provide valuable in- sights into ventricular function and the extent of pulmonary over circulation [14,15].

The first series of operations performed for TAPVC in 1960s reported an extremely high mortality ranging from 65 to 88% [16]. Results of correction of TAPVC in infancy have improved markedly in recent years, with an operative mortality of 5% or less by same institution [16]. Operative and late mortality rates were 19.3% and 4.3% among 57 consecutive patients aged 15 days to 18 years (median 6 months), underwent rechanneling of mixed totally anomalous pulmonary venous connection in All India Institute of Medical Sciences, India [17]. Alteration of preoperative stabilization, early operation, progressive refinement of surgical technique and improved quality of postoperative management may have all contributed to this decline [16]. The timing of surgical intervention in TAPVC is critical; delays can lead to irreversible pulmonary hypertension and right heart failure [4].

The minimal invasive pediatric cardiac surgery has been used to correct various congenital cardiac anomalies like, ASD, VSD, partial AVSD, complete AVSD, Subaortic membrane (SAM), PAPVD) with excellent patient satisfaction [1]. Now MICS for TAPVC is an emerging, advanced approach primarily used for supracardiac and some other types of repairs in children.

Minimal invasive pediatric cardiac surgery especially for supracardiac TAPVC repair is a highly challenging and doable with favorable anatomy. To the best of the author knowledge, this is the first successfully done by minimal approach of such complexity in Bangladesh.

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