REPAIR OF MIXED TYPE TOTAL ANOMALUS PULMONARY CONNECTION (TAPVC) IN AN INFANT: A Case Report

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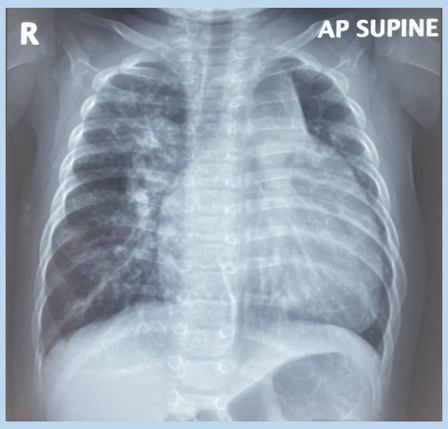
Background

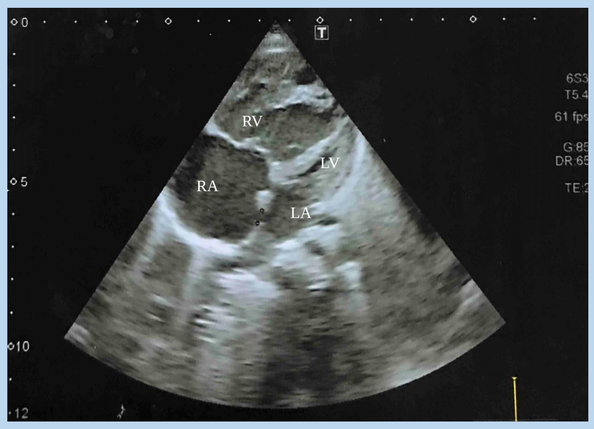
Total anomalous pulmonary venous connection (TAPVC) is referred to a specific condition in which there is no direct connection between any pulmonary veins and the left atrium. TAPVC is divided into supracardiac, cardiac, infracardiac and mixed types. With the accumulation of experience in surgical treatment and intensive postoperative care, the outcomes of TAPVC have improved over time, with reported mortality rates consistently < 10%. However, mixed-type TAPVC (5%), in which more than one level of pulmonary venous drainage exists, seems to be the most problematic subgroup.

Case Report

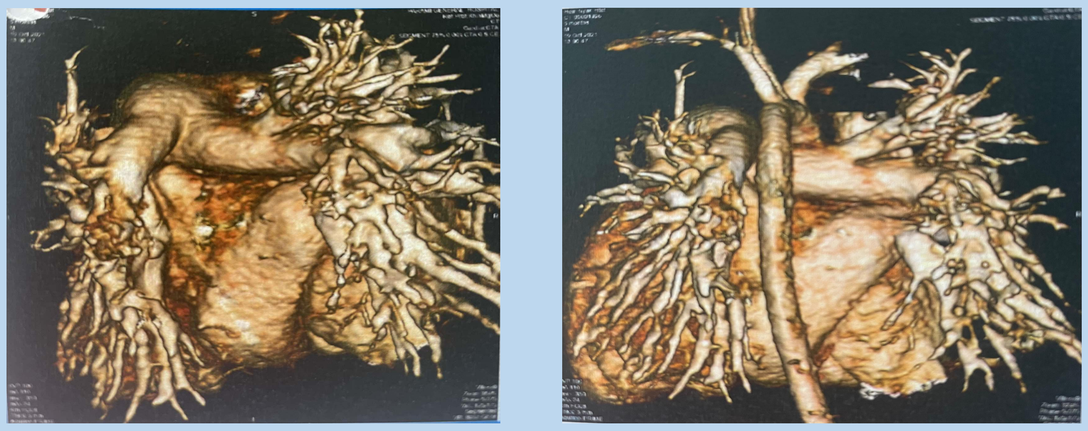
A 4 month old boy was referred to CVSW (YKCH) who was diagnosed with TAPVC. He was 4.5 kg with tachypnea, central cyanosis and respiratory distress. Aloud P2 was heard. ECG showed normal sinus rhythm with right axis deviation. Echography revealed enlarged both right atrium and right ventricle with relative small left chambers. Secundum Atrial septal defect(ASD) was small. Right Upper Pulmonary Vein (RUPV) entered into coronary sinus, the rest three PVs united to form a vertical vein and entered into innominate vein and SVC. Therefore, we proceeded to CXR and CT (Angiogram) for surgery.



CXR showed huge cardiomegaly with very plethoric lungs



Echo showed small left chambers and restrictive ASD



CT (Angio) revealed RUPV and RLPV joined to form a common channel and drained into coronary sinus into RA. LUPV and LLPV joined to form a vertical vein and drained into SVC and then to RA

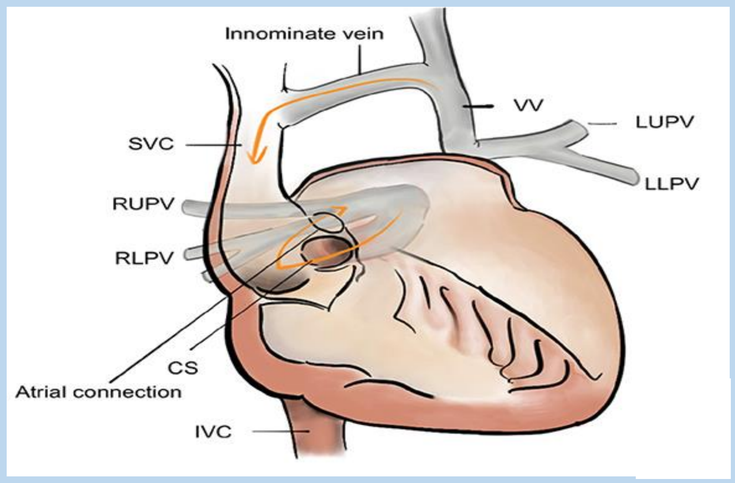
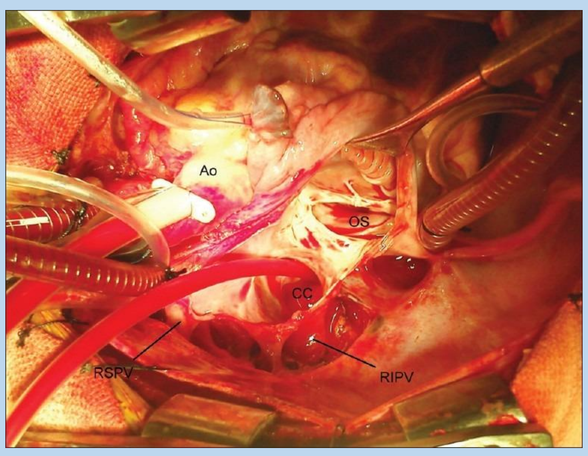


Illustration for Mixed type of TAPVC (Type IVd 2+2 (intra+supra) according to Darling's classification)

Procedure

The surgical correction of anomaly was carried out by complete unroofing of the CS wall into the LA and patch closure of the atrial septal defect (ASD) (similar to treatment of the cardiac type); then, the confluent was anastomosed to the LA.



ASD was closed by using pericardium patch with directing the coronary sinus drain into LA

Preoperative stabilization and postoperative course were challenging because of pulmonary venous congestion and pre-existing heart failure. The child needed mechanical ventilator support for 5 days and had an uneventful recovery and was discharged on day 23.

Discussion

The correct diagnosis and accurate anatomical description of TAPVC is necessary for planning the surgery. Accomplishing the detection in the mixed type of anomalous is difficult without CT (Angio). Surgical correction for TAPVC can be beneficial, but complications including severe preoperative cardiopulmonary instability in infants with obstruction, postoperative paroxysmal pulmonary hypertension, and delayed development and progression of pulmonary vein stenosis are still expected.

Comment

Our cardiac surgical team has operated 20 cases of TAPVR since 2012. Among them, mixed type (Type IV) is rare condition but usually presents with uncontrolled heart failure and severe pulmonary venous congestion during infancy. Along with mixed type, every variant requires an appropriately specific approach regarding operative technique.

Futher information

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