CASE REPORT

Role of Perioperative Echocardiographic in Surgical Correction of the Pulmonary Venous Anomalies

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ABSTRACT

Anomalous pulmonary venous drainage is the anomalous drainage of one or more than one pulmonary vein draining into right atrium, superior vena cava, inferior vena cava, coronary sinus, or innominate vein. Right-sided pulmonary vein anomalous drainage is usually associated with sinus venosus type atrial septal defect. Pericardial patch repair and rerouting of the right pulmonary vein are commonly performed operations for such cases. These operations involve a risk of obstruction to the flow of superior vena cava or rerouted pulmonary vein in the postoperative period. The defects are well visualized on the transesophageal echocardiography (TEE). We are reporting three cases operated for anomalous drainage of right-sided pulmonary vein, highlighting the perioperative echocardiographic features.

Keywords: Anomalous pulmonary vein drainage, Sinus venosus defect, Transesophageal echocardiography.

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INTRODUCTION

The sinus venosus defects are well visualized on the transesophageal echocardiography (TEE) because of the proximity of the TEE probe to these structures. We report three cases of superior sinus venosus atrial septal defect (SVASD) with anomalous drainage of right-sided pulmonary veins operated for the pericardial patch repair with rerouting of the anomalous pulmonary vein. We will also describe the perioperative echocardiographic features before and after the surgery.

CASE REPORTS

Case 1

A 6-year-old male child presented with a history of recurrent lower respiratory tract infection since birth.

There was no history of cyanosis or pedal swelling. On examination, the child had a heart rate of 80 per minute. On auscultation, he had bilateral normal vesicular breath sounds and ejection systolic murmur grade III. Child was in NYHA III. Electrocardiogram showed normal sinus rhythm and chest X-ray showed cardiomegaly. The 2D echocardiography revealed intracardiac type total anomalous pulmonary venous connection (TAPVC) wherein all pulmonary veins were draining into the common chamber and the common chamber into the right atrium (RA). There was 16 mm ostium secundum atrial septal defect (OS ASD) with right to left shunt. There was no right ventricular outflow tract obstruction (RVOTO), left ventricular outflow tract obstruction (LVOTO), or associated patent ductus arteriosus (PDA). Right atrium and right ventricle (RV) were dilated. The patient was listed for intracardiac repair.

After induction and monitoring (invasive blood pressure, central venous pressure) in place, a TEE probe was inserted and TEE examination was accomplished using 2D TEE probe (6VT-D probe of the GE vivid E9 echocardiography system (GE Medical Systems, Horten, Norway)).

A superior type of SVASD, shunting in left-to-right direction, was seen. The defect was seen at zero sector angle by withdrawing the probe from mid-esophageal (ME) 5-chamber view and slightly rotating it clockwise until it reached the junction between the superior vena cava and RA. On turning the navigation scan to (ME) bicaval view and withdrawing the probe from this position to the level of right pulmonary artery (RPA), it enabled us to visualize the sinus venosus defects situated inferior to the RPA. On advancing the sector between 110 and 120° and rotating the shaft a bit clockwise, we could identify the right pulmonary vein opening at the SVC-RA junction (Figs 1A and B). Left-sided pulmonary veins were found draining into the LA.

After performing median sternotomy, pericardium was harvested. Left superior vena cava (LSVC) was seen draining into the left atrium (LA) and the right superior pulmonary vein (RSPV) was draining into RA. All other pulmonary veins were draining into LA. Cardiopulmonary bypass (CPB) was established with aortobicaval cannulation and LSVC cannulated. Heart was arrested with antegrade aortic cardioplegia in

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Figs 1A and B: Superior type of SVASD with unroofing of RUPV seen in modified ME bicaval view

Fig. 2: Postoperative SVC obstruction seen in ME bicaval view

Fig. 3: Right Inferior pulmonary vein draining into right atrium seen in ME modified four-chamber view; SVC: Superior vena cava

moderate hypothermia. Right atrium was opened; pericardial patch buffering of opening of LSVC into RA and RSPV into LA was done. Closure of ASD was done with prolene 5.0 continuous sutures.

Postoperative TEE examination ruled out the presence of a residual ASD but revealed turbulent flow at SVC-RA junction indicated SVC obstruction (Fig. 2). The pressure gradient measured in transgastric view with the Doppler aligned parallel to the blood flow in the SVC and SVC-RA junction was 8 mm Hg. The obstruction at SVC-RA junction was corrected on going again on bypass.

Case 2

A 6-year-old female child that presented with a history of recurrent lower respiratory tract infection since birth was diagnosed with 24 mm large OS ASD with left to right shunt. There was associated partial anomalous pulmonary vein (PAPVC) with the right upper pulmonary vein (RUPV) draining into RA. Right atrium and RV were dilated. The patient was listed for ASD closure.

After induction and monitoring (IBP, CVP) in place, TEE examination was accomplished using 2D TEE probe (6VT-D probe of the GE vivid E9 echocardiography system (GE Medical Systems, Horten, Norway)). Preoperative TEE revealed OS ASD with PAPVC wherein the RUPV was draining into SVC, the right inferior pulmonary vein (RIPV) adjacent to it in RA (Fig. 3), and the left pulmonary veins into LA. There was 24 mm OS ASD with left to right shunt. There was no RVOTO, LVOTO, or associated PDA. RA and RV were dilated.

After performing median sternotomy, pericardium was harvested. RSPV was seen draining into SVC and RIPV adjacent to it in RA (Fig. 4). All left pulmonary veins were draining into LA. Cardiopulmonary bypass was established and RA was opened, Polytetrafluoroethylene (PTFE) patch closure of ASD by excising foramen ovale tissue, thereby enlarging LA along with pericardial patch enlargement of opening of SVC into RA, was accomplished. While RSPV was rerouted to LA, RIPV was left to drain into RA. Postoperative TEE examination ruled out the presence of a residual ASD and SVC or

Fig. 4: Right Inferior pulmonary vein and SVC draining separately into right atrium along with OS ASD; SVC: Superior vena cava, OS ASD: Ostium secundum atrial septal defect

Fig. 5: Right superior and middle pulmonary veins draining into SVC – RA junction seen in modified ME bicaval view; SVC: Superior vena cava, RA: Right atrium, RPA: Right pulmonary artery

pulmonary venous flow obstruction. The pericardial patch had rerouted the RUPV to the LA. The RUPV-LA pressure gradient was less than 5 mm Hg.

Case 3

A 2-year-old female child weighing 10 kg presented with poor weight gain since birth. She was diagnosed with 9 mm SVASD + 7 mm OS ASD with left to right shunt. There was associated anomalous pulmonary vein drainage with right superior and middle pulmonary veins (RSPV and RMPV) draining into SVC-RA junction (Fig. 5). Left veins were draining into LA. Right atrium and RV were dilated. Intraoperative preCPB TEE confirmed similar findings. After performing median sternotomy, pericardium was harvested and findings were confirmed. Pericardial patch closure of SV and OS ASD with pericardial patch baffling of RSPV and RMPV into LA was done (Fig. 6).

Postoperative TEE examination ruled out the presence of a residual ASD and SVC or pulmonary venous flow obstruction. The pericardial patch had rerouted the RPV to the LA.

All patients were successfully weaned from the ventilator within 24 hours and discharged after an uneventful recovery.

DISCUSSION

Pulmonary venous anomalies include many anatomic variations, with a wide range of clinical presentations and outcomes.

The anomalies may be grouped into four categories:

1. Abnormal numbers of pulmonary veins
2. Normal pulmonary venous connections with anomalous drainage
3. Stenotic connections
4. Anomalous connections.

Fig. 6: ME four-chamber view showing pericardial patch baffling of the right pulmonary vein to the LA; SVC: Superior vena cava, RA: Right atrium, RV: Right ventricle, LA: Left atrium, LV: Left ventricle, RUPV: Right superior pulmonary vein

The incidence of pulmonary venous anomalies varies widely according to the particular anomaly observed. An abnormal number of pulmonary veins are commonly encountered; anatomic studies show a single right or single left pulmonary vein in nearly 25% of the population, and a third pulmonary vein is present on either side in 1.6 to 2%.¹

Normal pulmonary venous connections with anomalous drainage occur when the common pulmonary vein incorporates normally into the LA, resulting in normally positioned pulmonary venous connections, but due to interatrial anatomy the pulmonary venous inflow is directed into the morphologic RA. Most commonly, this is due to anatomically leftward malposition or malattachment of septum primum, which can result in partial or total anomalous pulmonary venous drainage, depending upon the degree of malposition and the number of veins affected.

The sinus venosus defects are located either superiorly at the right atrial level and the superior caval vein or inferiorly at the level of the inferior caval vein. Sinus venosus defect is a type of anomalous pulmonary venous drainage with normal connections that occurs due to “unroofing” or absence of the sinus venosus tissue between the right pulmonary veins and the superior vena cava or RA.^{2,3} A great number of variations are observed in the anatomical drainage patterns of the pulmonary veins.⁴

Diagnosis of the SVASD may sometimes be difficult on TTE because the atrial septum appears intact in most views. Multiplane TEE is very useful in imaging SVASD and is superior to the TTE in the diagnosis of this lesion.^{2,5-7} The proximity of the septum to the esophagus permits the entire structure to be adequately visualized in virtually every patient. The superior SVASD is best visualized in the ME bicaval view and in the transverse imaging plane at the SVC-RA junction. This required a slight withdrawal of the transducer shaft upward from the aortic valve level to the junction of the RA and SVC.⁸ The defect appeared as an absence of the atrial septum just beneath the orifice of the superior vena cava. In an ME bicaval view, the RPA is usually seen in the superior aspect of the defect. The inferior aspect of the defect is formed by the atrial septum. Right pulmonary artery is seen superior to SVC.⁹ According to Van Praagh et al,³ the unroofing of RUPV is invariably present in the SVASD near the SVC. A normally draining RUPV can be visualized in a transverse plane by turning the probe clockwise from a 5-chamber view and in a longitudinal image plane developed from the bicaval view either by turning the probe to the right or by rotating the transducer forward 10 to 20° from the standard position.¹⁰ All these features were present in our patients. The examiner should identify all four pulmonary veins entering the LA, as variable anatomy may change the course of the surgery. The view of RIPV is obtained by rotating the probe clockwise from the view of RUPV at either the modified bicaval view or at the 30 to 40° sector angle from the ME four-chamber view and then rotating the probe slightly clockwise, which was obtained in our patient.¹¹

Persistent left SVC is the most common congenital thoracic venous anomaly with a prevalence of 0.3 to 0.5% in general population. The most common subtype of PLSVC results in the presence of both left and right SVCs. A bridging innominate vein may or may not be present. Variations have also been reported in the insertion of left SVC. In 80 to 90% of individuals, the persistent LSVC drains into the RA via the coronary sinus and is of no hemodynamic consequence. It is evident by the presence of a dilated coronary sinus on echocardiography further

confirmed by saline contrast echocardiography. In the remaining cases, it may drain in the LA resulting in a right- to left-sided shunt as in our case. Almost 40% of patients with PLSVC can have a variety of associated cardiac anomalies,^{12,13} such as atrial septal defect, bicuspid aortic valve, coarctation of aorta, coronary sinus ostial atresia, and cor triatriatum.

The basic principle of surgical repair for the SVASD with unroofed RUPV is to redirect the flow of RUPV into the LA without creating a pulmonary venous or SVC obstruction or injuring the SA node or its supplying artery. In single-patch repair technique, the defect is closed by creating a pericardial tunnel that directs the flow from RUPV to the LA.

However, as the pericardial tunnel is created within the SVC lumen, SVC flow obstruction may be sequel in the postoperative period. Following repair of sinus venosus defect, obstruction of both the pulmonary venous pathway and the SVC has been described.¹⁴ In repaired anomalous pulmonary venous drainage, echocardiographic follow-up evaluation should include assessment for any stenosis of the venous baffle to the LA, residual leaks, and presence of any stenosis of the systemic vein, particularly SVC. The rerouted pulmonary vein is seen on turning the probe to the right from the SVC-RAA anastomosis, which may not be visualized simultaneously with the pulmonary vein.

It is difficult to assess the absolute size of the surgical anastomotic site after termination of the CPB by direct visualization in one plane with TEE. However, TEE provides important anatomical information in the intraoperative evaluation of residual anastomotic obstruction by the presence of turbulent flow. It strongly indicates residual or inadequate repair of the anastomosis.¹⁵ Therefore, TEE can provide early detection of inadequate repair during surgery and allow immediate revision without delay.

In summary, with the help of TEE, we could recognize echocardiographic features associated with a superior type of SVASD and anomalous drainage of the right-sided pulmonary vein in the SVC or RA. TEE is a useful diagnostic and monitoring tool during the surgeries involving a superior SVASD, anomalous pulmonary vein drainage, and postoperative sequel if any.

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Role of Perioperative Echocardiographic in Surgical Correction of the Pulmonary Venous Anomalies

Sinus venosus atrial septal defect (SVASD) is an extraseptal defect, which occurs due to deficiency in the common wall between the left atrium (LA) and the superior vena cava (SVC). Sinus venosus atrial septal defect constitutes approximately 4 to 11% of all atrial septal defects. Approximately, 90% of cases are associated with partial anomalous pulmonary venous connections (PAPVCs), causing additional left-to-right shunting. Sinus venosus atrial septal defects are of two types. Superior SVASDs (the more common type) are located in the most superior and posterior region of the atrial septum in close proximity to the right-sided pulmonary veins (PVs) and the entrance of the SVC into the right atrium (RA). Inferior SVASDs (rare) are located posterior and inferior to the fossa ovalis and adjacent to the inferior vena cava (IVC). Both types are associated with PAPVCs, which is characterized by failure of one or more PVs to connect with the LA. Superiorly located SVASD is associated with unroofing of right upper pulmonary vein (RUPV). A deficiency in the wall, which separates SVC from LA, results in the superior type of sinus venosus defect associated with an unroofed RUPV, which drains into SVC or RA. The aim for the repair of SVASD with unroofed RUPV is to close the interatrial defect and to redirect the anomalous pulmonary venous drainage into the LA without creating a pulmonary venous or SVC obstruction, or injuring SA node or its supplying artery lest sinus node dysfunction (SND) occur.

There are three types of surgical repairs that can be done for SVASDs:

1. Single-patch closure of the atrial septal defect with anomalous PV baffled to LA. In this technique, a pericardial tunnel is created within the SVC. This tunnel not only closes the defect but also directs flow of blood from RUPV to LA. Obstruction of flow in SVC is a complication that may occur in the postoperative period. Pulmonary venous obstruction is another known complication after the internal patch closure of a SVASD. Unobstructed blood flow in SVC and PVs should be confirmed using transesophageal echocardiography (TEE) after surgery.
2. Two-patch technique – One patch to baffle the PV and one to augment the SVC. The SVC–RA junction is augmented with a pericardial patch to avoid SVC obstruction. This procedure carries the risk of permanent sino-atrial node dysfunction caused by injury to sinus node artery by the incision made across SVC–RA junction.
3. When there is difficulty in making an unobstructed baffle or when PVs drain into high SVC (high above the junction between the SVC and RA), a Warden procedure is performed wherein the SVC is transected above the level of uppermost opening of the PAPVC. The cardiac end of the SVC containing the anomalous PV is baffle closed to the LA using a pericardial patch. The cut end of lower part of SVC is suture closed. The right atrial appendage (RAA) is constructed into a tubular structure (prepared as a pedicle flap) and anastomosed to the upper stump of the SVC. In the Warden procedure, there is no cavoatrial incision and therefore no risk of damage to sinus node artery or the sinus node. Incidence of arrhythmias and SND remains low with Warden repair, although it carries the risk of SVC–RAA anastomotic site obstruction and PV obstruction.

Transesophageal echocardiography has superior ability to visualize the anatomy of interatrial septum, SVASDs, and drainage of PVs, compared with transthoracic echocardiography (TTE) due to the proximity of the probe to the RA and SVC. Best TEE views for SVASD: Mid-esophageal four chamber (ME4C), mid-esophageal two chamber (ME2C), mid-esophageal (ME) bicaval, deep transgastric (TG), and bicaval equivalent.

Two-dimensional (2D) imaging: Location and size of atrial septal defect (ASD) must be noted. Superior defect is located above foramen ovale near SVC. Inferior defect is located in inferior part of interatrial septum (IAS) below foramen ovale near IVC. Size of ASD must be measured in multiple planes. Atrial septal defect size is quantified as – Small: 3 to 5 mm; moderate: 5 to 8 mm; and large: 8 mm or larger. Interatrial septum must be examined for multiple defects. Echocardiographic features associated with a superior type of SVASD with unroofing of the RUPV include the following:

- Overriding of the rim of fossa ovalis by SVC;
- Unroofing of RUPV, which drains into SVC or RA;
- Deficiency of tissue in the wall that normally separates LA from SVC, resulting in inter-atrial shunt; and
- Tear drop appearance of SVC due to drainage of RUPV seen in transverse plane above SVC–RA junction.

Structure and function of mitral valve (MV) and tricuspid valve (TV) is examined.

Annulus of MV and TV are measured. The pulmonary valve is examined for restricted motion, doming leaflets, and commissural fusion. Anatomy and size of the right sided SVC is noted. The oval lumen of the persistent left SVC

(PLSVC) is seen between the LA appendage and left upper pulmonary vein (LUPV) in the ME two-chamber view. It is seen as an echo-free space between the left atrial appendage and LUPV, with probe turned slightly counterclockwise. It is possible to trace the course of PLSVC in the mid-esophageal views up to the site at which it enters the coronary sinus. Injection of agitated saline through a left-hand peripheral vein fills the LSVC lumen with small bubbles, which exit through the coronary sinus into the RA in most patients. Mid-esophageal modified bicaval view or deep four-chamber view may be used for this visualization. In the presence of PLSVC, the coronary sinus is often enlarged to more than 1 cm (diameter). In the presence of a well formed PLSVC, the cardiac end of Right SVC (RSVC) may be small in size, which has a surgical implication. Precardiopulmonary bypass (CPB) TEE examination of the cardiac end of RSVC may help predict the necessity for the Warden repair.

Right atrial enlargement and RV enlargement must be looked for. In hemodynamically significant shunts, volume overload may result in right atrial enlargement, RV dilation, paradoxical septal motion, main and branch pulmonary artery enlargement, and left atrial enlargement. The interventricular septum may appear flat during diastole. In long-standing cases, diastolic dysfunction of left ventricle may occur due to under-filling, advanced age, and associated coronary artery disease. Therefore, it is necessary to assess biventricular function. Pulmonary hypertension, RV hypertrophy, right to left shunt and systolic flattening of interventricular septum in systole may indicate Eisenmenger syndrome.

Color flow Doppler and spectral Doppler: Color flow Doppler (CFD) and spectral Doppler are used to identify size and direction of flow through ASD. In most of the cases, shunting is left to right, of low velocity, and biphasic (varying during systole and diastole). Direction of the shunt is determined primarily by color Doppler. The degree of shunting is determined by the ventricular compliance. Left to right shunting across an ASD increases with age. It is often minimal during infancy as the RV is initially less compliant. Measuring velocity of flow using spectral Doppler is not usually helpful because a pressure gradient between left and right atria greater than 2 to 3 mm Hg is rare. Moreover, to use spectral Doppler, the ultrasound beam needs to be aligned in a parallel fashion. Left superior vena cava can be distinguished from LA appendage and LUPV using spectral Doppler.

Anomalous pulmonary venous drainage if any must be located using CFD during pre-CPB TEE. For this, it is necessary to assess drainage of all PVs. To look for the left PV, ME 4C, and ME 2C views are used. Slightly withdrawing the probe from ME 4C view, and rotating the probe to the right, reveals the SVASD and anomalously draining RUPV into the SVC. Sinus venosus atrial septal defect is seen with the SVC overriding the fossa ovalis in high bicaval view. The right PVs can be visualized by increasing the sector angle to 120° to 130° after withdrawing the probe to the level of right pulmonary artery. The right upper lobe and middle lobe PVs may join to form a common RUPV, which directly open in the RA. In some cases, both lobar veins may separately drain into RA. Surgical decision is aided by identification of anatomy using TEE. Using pulse wave Doppler, SVC/IVC is examined for evidence of high normal velocity (aliasing) flow suggestive of anomalous PV drainage. It is necessary to advance and withdraw the probe to locate upper and lower veins. The number of anomalous PVs draining directly in the SVC and distance between the proximal (highest) opening of PV and the SVC-RA junction can be visualized by rotating the sector angle between 90° and 130° and withdrawing the probe from SVC-RA junction. Utilizing M mode in the bicaval view, the SVC size can be measured. A dilated coronary sinus is an indication that the blood flow through SVC is high. The SVC anatomy influences the choice of the surgery. Warden procedure is preferable when the PAPVCs drain into SVC at a site more than 2 cm away from SVC-RA junction and also when size of RSVC is smaller than LSVC in the absence of a connecting vein. Intraoperative TEE is of importance in the pre-CPB period of Warden repair for guiding the surgeon in planning the operation. Length of the patch which is used to direct blood flow from the PVs to the LA can be determined by identifying the highest opening of PV in SVC. The chances of obstruction of SVC lumen are higher when the patch is longer.

It is necessary to identify and grade any existing mitral regurgitation and tricuspid regurgitation using CFD and continuous wave Doppler (CWD).

If TR is present, CWD is used to measure RV systolic pressure. Right ventricular systolic pressure = pulmonary artery systolic pressure = 4 (square of peak velocity of TR jet) + right atrial pressure. Color flow Doppler and CWD/PWD are used to assess any pulmonary stenosis in upper esophageal aortic arch short-axis view or ME ascending aortic short-axis views. Atrial septal defect flow itself may result in velocity at the pulmonary valve of up to 2.5 m/sec.

Postrepair TEE assessment: The atrial septum is evaluated for any residual shunting. In the post-CPB period following Warden repair, it is necessary to look for vascular obstructions. There should be no obstruction or narrowing of SVC-RA appendage anastomosis (Warden anastomosis), SVC baffle, SVC, and PV. The intra-atrial baffle is evaluated for leaks and stenosis. The patch of baffle should appear well placed without dehiscence or

impingement of surrounding structures. Mid-esophageal four-chamber and bicaval views are used to confirm that the patch is diverting pulmonary venous blood to the LA. The warden anastomosis is inspected in ME bicaval view with injection of agitated saline and also on color Doppler. When agitated saline is injected via right internal jugular vein, ME bicaval view with slight rightward rotation of the probe demonstrates flow of agitated saline through the Warden anastomosis. Flow obstruction at this site should be ruled out on TEE examination in the immediate post-CPB period. Using ME windows, it is not possible to align the Doppler beam in a parallel fashion to blood flow across SVC–RAA anastomosis.

Modified (by gentle sector angle adjustment and probe manipulation) TG RV inflow view and TG RV inflow-outflow views are utilized to visualize the Warden anastomosis using color Doppler. The modified TG bicaval view is another useful view to visualize the Warden anastomosis. Pressure gradient across the Warden anastomosis is interrogated on spectral Doppler. If SVC–RAA anastomosis is narrow, increase in PWD gradients are observed at this site in TG RV inflow view and TG RV inflow outflow view. Usually, SVC–RAA gradient of more than 5 mm Hg is unacceptable. Tacy et al have mentioned that loss of biphasic pattern in SVC is more indicative of obstruction rather than absolute value of gradient as it varies with the loading conditions.

The obstruction of rerouted PVs is detected in ME-modified bicaval view, which is obtained by extending the sector angle to 110° to 120° and probe rotation to right. Ultrasound beam can be easily aligned with the RUPV. Pulmonary venous obstruction is ruled out if the pulsed wave Doppler gradient is less than 2 mm Hg. Superior vena cava and PV flow should be laminar, low velocity, and phasic. The presence of flow acceleration in any of the rerouted PVs indicates that there may be obstruction at the level of the baffle.

Color Doppler imaging of SVC–RA junction in the ME bicaval view with the probe slightly rotated to the right will show no color flow from the native SVC to the RA, as the blood is rerouted to LA. The original SVC–RA junction is now a part of the pericardial baffle.

Intraoperative TEE can delineate the anatomy of lesion, which assists the surgeon in making choice of operative procedure. After repair of lesion, TEE imaging has potential to diagnose anastomotic site obstruction and may alarm the surgeon to undertake corrective measures.

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