
Design: A retrospective observational case series.

Methodology: Pediatric patients who underwent Warden repair between October 2011-September 2015 were recruited. Their preoperative clinical details, anesthetic techniques, intraoperative TEE findings and postoperative events were recorded from the medical records. The categorical variables and the continuous variables were expressed as number (percentages) and mean ± SD respectively.

Results: A total of 35 patients were operated for Warden repair during the study period. Anesthesia was induced with the aim to prevent any fall in pulmonary vascular resistance. The right internal jugular vein was cannulated under ultrasound guidance using a short length cannula to monitor right superior vena cava pressure. Intraoperative TEE revealed the drainage of PAPVC high into RSVC in 22 patients. Persistent LSVC was found in 9 patients. After repair, TEE imaging detected a high gradient at Warden anastomotic site in 5 patients and 3 of them required revision of surgery. Rerouted pulmonary veins required surgical correction in 2 patients in view of obstruction. None of them had pulmonary venous and SVC obstruction in the postoperative period.

Conclusion: The primary aim of anesthesia is to avoid any fall in PVR. Right IJV cannulation can be beneficial. The intraoperative TEE can help in delineating the anatomy of lesion and detecting anastomotic site obstruction.

Key words: Sinus venosus atrial septal defect; Superior vena cava obstruction; Transesophageal echocardiography; Warden repair

INTRODUCTION

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). Approximately, 90% of cases are associated with partial anomalous pulmonary venous connections (PAPVCs). The basic concept behind the repair of SVASD is to close the interatrial defect and to redirect the anomalous pulmonary venous drainage into the LA. The surgical correction of SVASD is

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more complex in comparison with the repair of ostium secundum atrial septal defect (ASD) and carries the risk of stenosis of the SVC or the rerouted pulmonary veins (PVs) and sinus node dysfunction (SND).[3,4] Different surgical techniques have been described in the literature in order to avoid these complications, which are determined by the anatomy of the lesion.[2,5] Based upon the size of the SVC and the site of drainage of the PVs into the SVC, we perform patch repair of the defect (single or double) or Warden procedure at our institute. When the PVs drain high above the junction between the SVC and right atrium (RA), Warden procedure is preferred over the patch repair. Principles of anesthesia management in these cases are driven by the anatomic complexity of the lesion, degree of intracardiac shunting, severity of pulmonary hypertension, and function of the right heart. Intraoperative transesophageal echocardiography (TEE) plays a pivotal role in assessing the anatomy of the defect and has a potential to alter the course of the surgery. We retrospectively analyzed patients with SVASD and PAPVCs operated for Warden procedure at our institute, focusing on intraoperative anesthetic challenges and the role of the TEE.

MATERIALS AND METHODS

After obtaining approval from the Institutional Ethics Committee, we collected data from the medical records of 35 pediatric patients, who were subjected to the Warden procedure from October 2011 to September 2015. Patient consent for the retrospective study was waived off by the Ethics Committee as per our Institutional Protocol. The data were categorized into different sections such as demography, clinical features, investigations, anesthesia induction techniques, invasive cannulation procedures, ventilatory strategies, and monitoring modalities. The TEE details during the period before the establishment of cardiopulmonary bypass (CPB) were retrieved from the TEE data charts, and echocardiographic images were collected from storage hard disks. Information was obtained on the anatomy and size of the right-sided SVC (RSVC), presence of left SVC (LSVC), number of anomalously draining PVs, distance between the proximal pulmonary venous opening in the SVC to SVC-RA junction, and the location and size of the ASD. Other findings noted were the right ventricular systolic pressure derived from tricuspid regurgitation (TR) jet, grade of volume overloading of the RV, and biventricular function. Patient case files were searched for documented incidents of post-CPB complications such as SVC and pulmonary venous obstruction, presence of residual shunt across the ASD patch, SND, and requirements for inotropes and cardiac pacing. Duration of mechanical ventilation and any adverse events in the postoperative period were recorded. The categorical variables and the continuous variables were expressed as the number (percentages) and mean ± standard deviation, respectively.

Standard protocols were followed for anesthesia and intraoperative TEE examination. The medical records revealed that all patients were kept nil per os in compliance with the ASA Guidelines and premedicated with syrup triclofos 50–70 mg/kg 1 h prior to the surgery. Inside the operation theatre, after application of standard monitors (5 lead electrocardiography (ECG), pulse oximetry and noninvasive blood pressure), anesthesia was induced with intravenous fentanyl 5–10 mcg/kg, midazolam 0.05–0.1 mg/kg, and propofol 2–3 mg/kg when the intravenous access was available. In patients with no intravenous access, inhaled sevoflurane was administered in increasing concentration up to 8% along with oxygen/air mixture (FiO₂ of 0.3–0.5). Pancuronium 0.1–0.15 mg/kg was injected to facilitate tracheal intubation. Mechanical ventilatory strategies were directed toward maintaining the pulmonary vascular resistance (PVR), which included ventilation with a high tidal volume (8–12 ml/kg), positive end-expiratory pressure (PEEP) of 5 cmH₂O, FiO₂ of 0.3–0.4% and maintaining mild hypercarbia (PaCO₂ 40–50 mmHg). Invasive arterial cannulas were secured in femoral or radial artery and triple lumen central venous catheters were inserted via femoral vein. A short cannula was placed in the right-sided internal jugular vein (IJV) in all patients under ultrasound guidance to monitor the RSVC pressure. Intraoperative TEE examination was performed in all the patients before and after the repair of SVASD. The TEE images were acquired from midesophageal (ME) and transgastric (TG) windows. The SVASD and PAPVCs were inspected in high ME 4-chamber view and high bicaval view [Video 1 and Figure 1]. The SVC-RA junction was also imaged in TG RV inflow view and RV inflow-outflow view.

Warden repair[6,7] is explained in Figure 2. The SVC was divided above the uppermost insertion of the anomalous PVs and its cardiac end was oversewn. A bovine pericardial patch or patient’s own pericardial patch depending upon surgeon’s preference was used to reroute the PVs across the ASD into LA. Right atrial appendage (RAA) was prepared as a pedicle flap and anastomosed to the proximal end of SVC. In post-CPB period, the warden anastomosis was inspected in ME bicaval view with
injection of agitated saline  [Video 2] and also on color Doppler  [Video 3]. The anastomosis was also inspected using color Doppler in TG RV inflow-outflow view and RV inflow view  [Video 4]. Pressure gradient across the Warden anastomosis was interrogated on spectral Doppler  [Figure 3]. The obstruction of rerouted PVs was detected in ME modified bicaval view, which was obtained by extending the sector angle to 110–120° and probe rotation to right. Pulmonary venous obstruction was ruled out if the pulsed wave Doppler gradient was <2 mmHg.

**RESULTS**

A total of 35 pediatric patients underwent Warden repair during the study period, 19 (55%) of whom were males. The demographic details are mentioned in Table 1. Common symptoms were recurrent respiratory tract infections and dyspnea on exertion. Chest auscultation predominantly revealed the presence of wide fixed split of second heart sound in all patients and ejection systolic murmur in pulmonary area in 57% of them. ECG and chest X-ray showed the presence of right heart enlargement and features of increased pulmonary blood flow. Transthoracic echocardiography (TTE) confirmed the diagnosis of SVASD associated with PAPVCs. All patients had mild-to-moderate TR, and mild pulmonary hypertension was found in 25 (71%) patients. Other findings were additional OSASD and LSVC in 10 (28%) and 9 (26%) of patients, respectively. The findings on TTE are summarized in Figure 4.

Intraoperative TEE findings confirmed the presence of SVASD in all patients.

The PAPVCs were present exclusively with the RSVC. One anomalously draining PVs was present in 22 (63%) of patients. Similarly, 2 and 3 anomalously draining PVs were detected in 12 (34%) and 1 (3%) of patients,
respectively. In 22 (63%) patients, uppermost opening of the PAPVCs was high up in the SVC, more than 2 cm away from the SVC-RA junction [Table 1]. Persistent LSVC was detected in 26% of patients. Decision to snare the LSVC or vent it through coronary sinus during conduct of the CPB was based on the size of the LSVC and presence and size of the bridging vein between the LSVC and the RSVC. When bridging vein was large in size, the LSVC could be snared, whereas in the presence of small bridging vein, the LSVC was vented through coronary sinus.

The mean duration of CPB and aortic cross-clamp time were 86.94 ± 21.65 min and 26.06 ± 14.55 min, respectively. After weaning the patients from CPB, dobutamine was required in 8 (23%) patients, whereas cardiac pacing was not required in any of them. TEE inspection was done to evaluate the results of the surgical repair. The Warden anastomosis was considered adequate in size when the RSVC pressure and SVC-RAA Doppler gradient were <15 and 5 mmHg, respectively. A high gradient of ≥8 mmHg at SVC-RAA junction was noticed in 5 (14%) patients; 3 of them underwent the SVC augmentation with additional pericardial patch during the second run of CPB [Figure 3]. In the fourth patient, the gradient was 10 mmHg in the immediate post-CPB period, which reduced to <5 mmHg after discontinuation of dobutamine infusion. In the remaining one, a high gradient of 7 mmHg was accepted due to the presence of a large bridging vein between the RSVC and the LSVC, which subsequently reduced to 3 mmHg in the postoperative period. Two (6%) patients had obstruction of one of the anomalously draining PVs, which required surgical revision with reinstition of CPB. The mean duration of ventilation and Intensive Care Unit stay were 4.88 ± 4.70 h and 1.5 ± 1.00 days, respectively. One patient had an episode of supraventricular arrhythmia 4 h after the surgery and required temporary pacing for 12 h. In the remaining patients, no SND was observed. Doppler flow evaluation at 6 months follow-up period showed SVC-RAA gradient <5 mmHg in all patients.

**DISCUSSION**

Sinus venosus ASD constitutes approximately 4–11% of all ASDs. The development of this interatrial communication is attributed to the deficiency in the common wall between the SVC and the right-sided PVs. Therefore, it is commonly associated with the anomalous drainage of one or more PVs into the RA or the SVC causing additional left-to-right shunting. The surgical options to deal with this defect include single or double patch repair. In single patch repair, a patch is used to reroute anomalously draining pulmonary vein across the interatrial defect into the LA, whereas in double patch repair, an additional second patch is employed to augment the SVC lumen. During Warden repair, the SVC is transected above the highest point of insertion of PAPVCs, and its cardiac end is closed. The unroofed PVs are redirected into the LA using a pericardial patch, which is succeeded by the anastomosis of the proximal end of SVC to the RAA.
Choice of the surgical procedure depends upon anatomy of the anomalously draining PVs and size of the RSVC. When the anomalous PVs open high, more than 2 cm, in the small-sized RSVC lumen away from the cavoatrial junction, a long patch is required to reroute the PVs across the ASD. It may occupy a significant portion of the lumen, potentially leading to the SVC obstruction. Augmenting the SVC-RA junction with the second patch as during double-patch technique, can injure SA node. Warden repair is designed in a way to avoid cavoatrial incision and damage to sinus node and its artery.[9,10] Incidence of arrhythmias and SND remains low with Warden repair,[11] although it carries the risk of SVC-RAA anastomotic site obstruction[10] and PVs obstruction.[12]

Patients with SVASD typically present with signs and symptoms of right ventricular volume overload and pressure overload as the pulmonary hypertension advances.[8] Excessive pulmonary blood flow in children with left-to-right shunt causes the development of hyperkinetic pulmonary hypertension and reversible increase in the PVR. Any reduction in the PVR associated with the administration of high FiO\textsubscript{2}, hyperventilation, and induction of respiratory alkalosis may result in further increase in the pulmonary blood flow and aggravation in the overloading of the RV volume. Hence, preventing fall in the PVR is an important strategy to maintain hemodynamic stability in the pre-CPB period. High tidal volumes and PEEP are well-tolerated by the patients. Site of central venous cannulation is another area of concern in the patients operated for the superior SVASD. Tip of the long central venous cannula introduced via right IJV may reach the site of anastomosis and impede the surgical anastomosis. Cannulation of left-sided IJV may be preferable to prevent this problem; however, it may be at a disadvantage in the presence of the LSVC as surgeon invariably applies snare around the vessel during the surgery. Moreover, coronary sinus damage may be a potential risk as the LSVC drains into the coronary sinus in majority of patients. Hence, it would be a logical choice to introduce the long central venous cannula via femoral vein. Inserting a short cannula via right IJV serves the purpose of RSVC pressure monitoring. Sometimes, dilemma may arise in identifying the IJV during venous puncture as aspirated blood may be bright red due to high opening of anomalous PVs in the SVC. This problem can be overcome by performing cannulation under the ultrasound guidance.

Role of the intraoperative TEE is of paramount importance in the pre-CPB period of Warden repair for guiding the surgeon in planning the operation. The TEE is more sensitive than TTE in visualizing the interatrial septum and pulmonary venous drainage[13,14] due to the proximity of the probe to the RA and SVC. In many cases, the right upper lobe and middle lobe PVs join to form a common right upper pulmonary vein (RUPV),[15] which directly open in the RA. However, both lobar veins draining separately into the RA are not uncommon. The highest opening of anomalous pulmonary vein in the SVC determines the length of patch required to reroute the PVs to the LA. The longer the patch more would be the probability of the patch obstructing the SVC lumen. The SVC anatomy also influences the choice of the surgery. In normal situation, the right and left innominate veins join to form the SVC. When the L SVC opens separately into the coronary sinus, the RSVC may be connected to it by an intervening bridging vein or both may be disconnected from each other in the absence of the bridging vein. The RSVC may remain smaller in the presence of a large LSVC, which partly shares the venous drainage from head and neck region. A comprehensive TEE report of anatomy facilitates the surgical decision. Warden procedure is preferable when the PAPVCs drains into SVC at a site more than 2 cm away from SVC-RA junction.[5,16] when size of RSVC is smaller than LSVC in the absence of a connecting vein.[17]

The characteristic echocardiographic features of SVASD include SVC overriding the rim of fossa ovalis, unroofed RUPV, and tear-drop appearance of the SVC in a horizontal plane above SVC-RA junction. Pre-CPB TEE helps identifying the anatomy of all PVs, especially those of the right-sided ones. Withdrawing the probe from bicaval view up to the level of right pulmonary artery and increasing the sector angle to 120–130° brings the right upper PVs in the ultrasound scan. Number of PVs draining directly in the SVC and distance between the highest opening of pulmonary vein 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. Size of the SVC can be measured in the bicaval view using M mode as the ultrasound beam can be angled perpendicular to its long axis. The oval lumen of the LSVC is seen between the LA appendage and left upper pulmonary vein in the LA appendage view, which can be easily distinguished from both the structures using spectral Doppler. 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 majority of patients. Although it may be difficult to measure the size of the LSVC using intraoperative TEE due to the
limitations in obtaining section of the LSVC in short axis, dilatation of the coronary sinus provides indirect clue regarding the degree of flow through the LSVC. The TEE evaluation of Warden repair in the post-CPB period is focused predominantly on detecting the residual shunts and vascular obstructions. Integrity of the patch in diverting pulmonary venous blood to the LA is confirmed in ME 4-chamber view and bicaval view. The native SVC-RA junction no longer empties into the RA as it becomes the part of the pericardial baffle. Flow acceleration in the rerouted PVs suggests obstruction at the level of their opening in the baffle. Pulmonary venous views have been incorporated in the newer 28 standard intraoperative TEE views, wherein the ultrasound line of interrogation can be aligned with the RUPV with minimum angulation. Usually, a pressure gradient <2 mmHg is considered unobstructed with the RUPV with minimum angulation. Usually, a pressure gradient <2 mmHg is considered unobstructed pulmonary venous drainage. Free flow across the Warden anastomosis site is evaluated in ME high 4 chamber view and bicaval view, using color Doppler and agitated saline. The major limitation of the ME windows is the inability to align the Doppler beam parallel to the flow across the SVC-RAA anastomosis. This problem can be addressed by visualizing the anastomosis in TG views. Although the anastomosis is not seen in standard TG RV inflow view and RV inflow-outflow view, off-axis visualization is possible with gentle probe manipulation and sector angle adjustment. The modified TG bicaval view, which has been described by the author, may be another useful view to visualize the Warden anastomosis. Usually, a SVC-RAA gradient of more than 5 mmHg is unacceptable. Tacy 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. In the current study, even though five patients had a high SVC-RAA gradient, only three required patch augmentation of anastomotic site as the biphasic pattern was preserved in remaining two. In the fourth patient, a high gradient was associated with tachycardia and hypertension and RSVC pressure of 10 mmHg. Discontinuation of dobutamine lowered the gradient. A high gradient of 7 mmHg at SVC-RAA anastomotic site was accepted in the fifth patient due to the presence of a large connecting vein and LSVC. The bridging vein functions as a pop-off mechanism for the decompression of the RSVC. We admit that the limitations of our study are retrospective, single-centered case series analysis constituting a small sample size. The results of our series cannot be compared with other techniques as it is just an observational study without any control group.

CONCLUSION

Anesthetic technique aims to maintain PVR as in all cases of left to right shunts. It may be beneficial to insert a short length cannula in right IJV to monitor SVC pressure in addition to central venous access through femoral vein. Intraoperative TEE may delineate the anatomy of lesion which assists the surgeon making choice of procedure. The Warden procedure may be a safe approach when SVASD is associated with PAPVCs draining high into SVC with low incidence of complications. Intraoperative TEE imaging has potential to diagnose anastomotic site obstruction and may alarm the surgeon to undertake corrective measures.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

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