Iranian Heart Journal; 2022; 23 (4)

Original Article

Outcomes of the Warden Procedure for Partial Anomalous Pulmonary Venous Connection: A New Idea From a Single-Center Experience

Gholamreza Omrani[1], MD; Yaser Tolouei tabar[1*], MD; Maziar Gholampour Dehaki[1], MD; Sanaz Asadian[1], MD; Nahid Rezaeian[1], MD

ABSTRACT

Background: Partial anomalous pulmonary venous connection (PAPVC) is prevalently right-sided. The Warden procedure (WP) is performed for the repair of PAPVC when the anomalous right-sided pulmonary veins (PVs) connect to the superior vena cava (SVC) far from the SVC-right atrium (RA) junction. We aimed to describe the mid-term outcomes in the WP. Moreover, we compared the outcomes in double-SVC cases with those without left-sided superior vena cava (LSVC).

Methods: In this retrospective study, the medical records of 25 (52% female) patients who underwent the WP between 2009 and 2019 were evaluated. Baseline, perioperative, and follow-up data, including mortality, SVC and PV obstruction, the presence of single right-sided or double SVC, and sinoatrial (SA) node dysfunction, were recorded.

Results: The mean (± SD) follow-up time was 5.08 years (±2.59 y). No mortality, SVC or PV obstruction, and SA node dysfunction were noted. SVC-RA anastomotic site mild stenosis occurred in 2 patients. Fourteen of the 25 patients (56%) had double SVC. Subgroup analysis of 2 groups of LSVC positive and LSVC negative revealed mild SVC anastomosis site stenosis in 1/14 (7%) of LSVC-positive and 1/11 (9%) of LSVC-negative patients (*P*=1.00). SVC anastomotic site patch augmentation was necessary in 3 of the 14 (21%) cases of the LSVC-positive group and none of the LSVC-negative patients (*P*=0.23).

Conclusions: The WP was associated with satisfactory outcomes. This method is excellent in patients who have concurrent LSVC. Coincident double SVC anatomy could be a new indication for performing the WP in relatively high PAPVC. ***(Iranian Heart Journal 2022; 23(4): 6-12)***

KEYWORDS: Warden procedure, Partial anomalous pulmonary venous connection, Superior vena cava, Double superior vena cava, Left-sided superior vena cava

[1] Rajaie Cardiovascular Medical and Research Center, Iran University of Medical Sciences, Tehran, IR Iran.

***Corresponding Author:** Yaser Tolouei tabar, MD; Rajaie Cardiovascular Medical and Research Center, Iran University of Medical Sciences, Tehran, IR Iran.
Email: yasertolouei@yahoo.com **Tel:** +989122751094

Received: April 2, 2021 **Accepted:** June 12, 2021

The anomalous pulmonary venous connection is the condition of the attachment of the pulmonary veins (PVs) to the systemic circulation. It is classified into 2 categories: total anomalous pulmonary venous connection and partial anomalous pulmonary venous connection (PAPVC). In the former, all PVs connect to the right side venous circulation, whereas some of the PVs drain into the right-side venous circuit in PAPVC.

Alsoufi et al[1] introduced a classification for PAPVC based on the connection site of the anomalous PVs. They proposed 5 types: right PAPVC to the superior vena cava (SVC), right PAPVC to the right atrium (RA), right PAPVC to the inferior vena cava (scimitar syndrome), left PAPVC to the innominate vein, and bilateral PAPVC. Mixed or dual type PAPVC is not defined in this classification.

PAPVC is found in 0.4% to 0.7% of autopsy studies. Prevalently, the right PVs connection to the RA or SVC occurs, and 90% of the cases have an associated atrial septal defect (ASD).[2,3] Thus, there is a left-to-right shunt, and a spectrum of manifestations may appear based on the shunt severity. In the presence of significant left-to-right shunting, pulmonary arterial hypertension will develop. Considering clinical presentations and shunt severity, different policies can be adopted by the clinician. The options include conservative management, surgical repair, and medical treatment with pulmonary artery vasodilators.[4]

There are several techniques for repairing the PAPVC of the right upper PVs to the SVC.[1] The goal of repair is to close the ASD and redirect the anomalous PVs into the LA. Dividing the SVC into 2 separate tracks and manipulating the SVC-RA junction during surgery increases the risk of the obstruction of the SVC pathway and sinoatrial (SA) node damage. Single-patch and double-patch repair methods could be selected when the junction of ectopic veins is close to the distal SVC.[5] The cavoatrial anastomosis technique, the Warden procedure (WP), is utilized when the PV connection site is far from the SVC-RA junction. In this method, the risk of SA node dysfunction is reported to be decreased.[6] In patients with double SVC and, thus, a narrower right-sided SVC, the concern regarding SVC-RA anastomotic site stenosis would be increased. Moreover, difficulties may be encountered utilizing single or double-patch techniques. We suppose that the WP might be more feasible in such circumstances.

In the present investigation, we report the mid-term outcomes of the patients who underwent the WP in our center. Furthermore, we compare the outcomes between a group of patients with double SVC and those without left-sided superior vena cava (LSVC).

METHODS

Study Population

This retrospective study included 25 patients (52% female) with a mean (±SD) age of 9.13 years (±13.66 y) at surgery who underwent the WP for PAPVC repair between 2009 and 2019 in Rajaie Cardiovascular Medical and Research Center. Ethical approval was obtained from the institutional review board committee. The requirement for written informed consent was waived due to the retrospective design of the study.

Surgical Procedure

Following a median sternotomy and pericardiotomy, the azygos vein was ligated and cut with the goal of the maximum mobilization of the SVC. In the next step, heparin injection, followed by the cannulation of the aorta, the SVC, and the inferior vena cava, was carried out. Then, a cardiopulmonary bypass was set. In the

presence of double SVC, the cannulation of the LSVC or the placement of a coronary sinus vent was performed. Mild hypothermia was applied, and the aorta was clamped. Next, a cardioplegic solution was injected into the aortic root. The SVC was obliquely transected above the anomalous vein connection site and sewed in its distal end. The lateral RA wall was opened, and the SVC was directed toward the sinus venosus ASD utilizing a patch (either pericardial or GORE-TEX). Creating a properly sized ASD is mandatory. The tip of the RA appendage was amputated, and the RA trabeculae were removed. Afterward, SVC-RA appendage anastomosis was performed. If required, the augmentation of the SVC-RA appendage anastomosis utilizing a pericardial patch was done to prevent future stenosis. All SVC-to-RA anastomoses were done without the use of any conduit (direct anastomosis). The autologous pericardium was used as an intra-atrial baffle in all the cases. Concurrent secundum ASD or patent foramen ovale was repaired.

Data Collection

The medical records of the study population were precisely evaluated. According to the institutional protocols, regular clinical, electrocardiographic (ECG), and echocardiographic examinations are carried out for individuals undergoing congenital cardiac surgeries. In selected patients, additional examinations, including computed tomography angiography, may be asked. Clinical, ECG, and imaging findings during the postoperative follow-up period (5.08 y [±2.59]) were registered. In the case of missing follow-up information, telephone calls were made, and the patients were recruited for additional examinations whenever needed.

SVC and PV Stenosis

Stenosis was defined as a pressure gradient exceeding 2 mm Hg in the SVC or PV pathway on serial echocardiography examinations. Additionally, a gradient of more than 5 mm Hg was considered severe stenosis. On computed tomography angiography, the reduction in diameter was regarded as stenosis. Further, significant stenosis was determined as a diameter reduction exceeding 50%. Early stenosis was defined as the presence of stenosis in the first postoperative month on echocardiography or computed tomography angiography, and late stenosis was defined as the occurrence of narrowing after the 30th postoperative day.

SA Node Dysfunction

SA node dysfunction was determined as persistent sinus bradycardia or junctional/nodal rhythm, ectopic atrial rhythm or a wandering pacemaker, or pauses exceeding 3 seconds. Two categories of early and late SA nodal dysfunction were defined. Early dysfunction referred to the detection of the mentioned ECG abnormalities in the first postoperative month, whereas the late dysfunction pointed to the ECG abnormalities later in the postoperative period.

Statistical Analysis

Categorical variables were represented as frequencies and percentages, while the continuous variables were reported as the mean ± standard deviation (SD). The differences between the groups of patients with and without LSVC were analyzed using the χ^2 test for the categorical variables. The mean values were compared using a 2-sample *t* test. All the statistical analyses were performed with the SPSS software, version 22.0 (IBM Corp), and a *P* value of less than 0.05 was considered statistically significant.

RESULTS

Twenty-five cases, including 13 females (52%), were enrolled in our study. The mean ± SD for age at the time of surgery was

9.13±13.66 years. The youngest patient was 8 months old at the time of surgery, and the oldest was 62 years old. The mean (± SD) follow-up time was 5.08 years (± 2.59 y) (range =1–10 y).

Fourteen of the 25 patients (56%) had double SVC. Ninety-six percent of the study population (24/25) had a sinus venosus ASD. Concurrent patent ductus arteriosus, patent foramen ovale, and secundum type ASD were noted in 4/25 (16%), 8/25 (32%), and 6/25 (24%) of our subjects, respectively. In 3 out of the 25 patients (12%), the SVC connected to the LA. In preoperative evaluations, 18/25 (72%) of the cases suffered mild right ventricular (RV) dysfunction. Moderate RV failure was detected in 5/25 (20%) of the patients, and 1/25 (4%) cases revealed severe RV dysfunction. RV function was normal in 1/25 (4%) patients in preoperative assessments.

Pericardial patch augmentation of the SVC-RA anastomotic site was performed in 3 (12%) cases. All these cases had LSVC. In 5 of the 25 patients (20%), enlargement of the sinus venosus ASD was necessitated. None of the patients represented signs or symptoms of SVC or PV stenosis during the follow-up. Ten patients (40%) revealed mild RV dysfunction during the follow-up. In the rest of the study subjects (15/25), no RV dysfunction was detected. Echocardiographic criteria for mild SVC-to-RA anastomotic site stenosis were present in 2 (8%) cases in the early and late postoperative periods. No SVC or PV stenosis was detected in the study subjects on follow-up echocardiography examinations. None of the cases developed SA node dysfunction after surgery. Moreover, no mortality was reported during the follow-up. Table 1 demonstrates the demographic and follow-up descriptive data of the study population.

Table 1: Demographic and follow-up descriptive data of the study population

Variables	Frequency (n)*	Percent %
Sex(female)	13/25	52
Double SVC anatomy	14/25	56
Associated PDA	4/25	16
Associated 2^0 ASD	6/25	24
Associated PFO	8/25	32
Sinus venosus ASD	24/25	96
SVC to LA connection	3/25	12
SVC anastomosis patch augmentation	3/26	12
ASD enlargement	5/25	20
Postoperative SVC stenosis	2/25	8
Postoperative PV stenosis	0/25	0
Postoperative SAN dysfunction	0/25	0
Postoperative mortality	0/25	0
Variables	Mean	±SD
AGE at surgery (y)	9.13	13.66
Pump time (min)	107.08	47.13
Cross-clamp time	65.08	40.36
ICU stay time (d)	2.96	2.24
Hospital stay time	8.52	3.83
Follow-up time	5.08	2.59

SVC, superior vena cava; PDA, patent ductus arteriosus; 2^0 ASD, secundum atrial septal defect; PFO, patent foramen ovale; LA, left atrium; PV, pulmonary vein; SAN, sinoatrial node; RV, right ventricle; SD, standard deviation

A subgroup analysis was done by splitting the patients into 2 groups according to the presence of LSVC. Mild SVC anastomosis site stenosis was present in the follow-up period in 1/14 (7%) of LSVC-positive and 1/11 (9%) of LSVC-negative patients, which was not statistically meaningful (P=1.00). SVC anastomotic site patch augmentation

was needed in 3 out of the 14 (21%) cases of the LSVC-positive group and none of the LSVC-negative patients. The difference was not statistically significant (*P*=0.23). Sinus venosus ASD enlargement was needed in 3 out of the 14 (21%) cases with LSVC and 2 out of the 11 (18%) cases without LSVC, which was not statistically meaningful (*P*=1.00).

DISCUSSION

PAPVC is defined as the connection of some, but not all, PVs to the RA or its venous tributaries. It commonly occurs on the right side, and it is associated with ASD. Three common surgical techniques are described for the repair of right-sided PAPVC and ASD: the single-patch technique, the double-patch method, and the WP.[7,8] The single-patch technique is utilized in selected patients when the drainage sites of the anomalous PVs and the SVC-RA junction are close to each other. SVC stenosis is not prevalent in this technique; however, a limited number of patients could be managed in this way. When the drainage site of the PVs and the SVC-RA junction are not close enough to perform the single-patch technique, either the double-patch method or the WP could be performed. For the prevention of SVC opening stenosis, the SVC is opened laterally, and another patch is inserted. This procedure is called “the double-patch technique”, and its major complication is a temporary or permanent SA node dysfunction. In the WP, the SVC is transected above the drainage site of the anomalous PVs, and its distal stump is closed. The inferior part of the SVC is directed toward the LA with a patch. Following RA appendage amputation, the superior end of the SVC is anastomosed to the RA appendage. In this procedure, the stenosis of the SVC-RA anastomosis site may occur.[6,8,9] This procedure is particularly carried out when the drainage sites of the anomalous PVs and the SVC-RA junction are distant from each other.

In the present study, we investigated the follow-up findings of the patients for whom the WP was carried out between 2009 and 2019 in our center. Warden and his colleagues[6] proposed the cavoatrial anastomosis technique as an alternative surgical method of PAPVC to SVC repair for the first time. Multiple studies have evaluated the outcome of patients who undergo the WP for PAPVC to SVC repair since its introduction in 1984. The initial results of the procedure were promising; nonetheless, only a few studies have included acceptable sample size and follow-up periods. Moreover, the majority of these investigations were conducted on adult populations.[1,5,6,10,11] In our investigation, we evaluated all the referred cases who underwent the WP in a time range of 10 years. Therefore, we accomplished an acceptable follow-up duration. Moreover, the inclusion of the pediatric population strengthened our investigation.

In our investigation, we encountered a notable frequency of double SVC in the study population (56%). We postulated that our surgeons may prefer performing the WP in these patients due to the smaller size of the right SVC to avoid SVC stenosis. On the other hand, we thought the same reason might result in SVC-RA anastomotic site stenosis in the WP. In a study by Lin et al,[12] 67 patients who underwent the WP were followed up for SVC stenosis. They found that an SVC diameter of less than 10 mm was a predictor of SVC stenosis or obstruction after the WP. Therefore, we conducted a subgroup analysis, and to our knowledge, this investigation is the first of its kind to compare the outcomes of the WP between LSVC-positive patients and LSVC-negative individuals. Remarkably, we found no difference in the occurrence of SVC anastomotic site stenosis in the 2 groups of

patients, despite a slightly more need for SVC anastomotic site patch augmentation in the LSVC-positive group. We concluded that the WP in the presence of LSVC not only precluded following SVC stenosis but also caused no significant SVC-RA anastomotic site narrowing. Accordingly, it could even be recommended in such circumstances. A point that should be mentioned is that the surgeon should be ready to perform SVC anastomotic site patch augmentation during the procedure.

In an investigation published by Shahriari et al[13] in 2006, 54 patients with sinus venosus ASD and PAPVC who underwent surgical repair were followed up for 13 years. In that retrospective study, the results of the WP and internal patch repair were compared. Just 1 patient in the Warden group died after 9 years because of acquired pulmonary hypertension due to a long-standing left-to-right shunt. Moreover, 1 patient of the Warden group developed symptomatic PV obstruction after 5 years. The authors demonstrated that the caval division for the repair of high PAPVC seemed to be competent with a low incidence of morbidity and mortality. They also mentioned that the WP was an efficient option for patients with high PAPVC.

In a recent study conducted on 40 patients who underwent the WP and were followed up for 22.5 months (range =12–39 mon), no mortality was detected. Moreover, no evidence of SVC or PV stenosis was observed, and all the patients had normal sinus rhythm. The authors of the study also compared their findings with those from a group of patients who underwent double-patch repair and revealed that both the double-patch technique and the WP had remarkable early and mid-term outcomes and depicted that the WP caused less sinus node dysfunction in the early postoperative period.[14]

In another study on 42 subjects who were followed up for a median time of 6 years (range =1 mon to 21 y), no mortality or permanent pacemaker requirement was reported. The investigators emphasized that the mid-term outcomes were excellent, with a low occurrence of SVC stenosis.[5] Similarly in our research, we encountered no mortality, nor did we detect any SVC or PV obstruction. SA node dysfunction occurred neither in the early nor in the late postoperative period in our patients.

Despite our promising results, we still believe that further investigations should be performed to convince surgeons to apply the WP conveniently for high PAPVC. It is particularly true in the group of patients with LSVC. Large-scale multicentric studies including different PAPVC repair procedure groups with detailed preoperative, postoperative, and follow-up data are warranted. Accordingly, we should compare the groups so as to choose the best option for patients.

CONCLUSIONS

The WP was associated with no mortality, low incidence of SVC stenosis, no PV obstruction, and no SA node dysfunction in the mid-term follow-up of the patients in the present study. Moreover, the WP appears to be a reasonable choice for patients who have concurrent LSVC. We suppose that the presence of double SVC in association with high PAPVC could be a new indication for WP.

REFERENCES

1. Alsoufi B, Cai S, Van Arsdell GS, Williams WG, Caldarone CA, Coles JG. Outcomes after surgical treatment of children with partial anomalous pulmonary venous connection. *The Annals of thoracic surgery*. 2007; 84(6):2020-2026.

2. Ahmed O, Maraj R, Rerkpattanapipat P, Yazdanfar S. An unusual case of bilateral partial anomalous pulmonary venous return. *Chest*. 1999;116(4):398S-398S.

3. Zwetsch B, Wicky S, Meuli R, Schnyder P. Three-dimensional image reconstruction of partial anomalous pulmonary venous return to the superior vena cava. *Chest*. 1995;108(6):1743-1745.

4. El-Kersh K, Homsy E, Daniels CJ, Smith JS. Partial anomalous pulmonary venous return: a case series with management approach. *Respiratory medicine case reports*. 2019;27:100833.

5. Yong MS, Griffiths S, Robertson T, et al. Outcomes of the Warden procedure for partial anomalous pulmonary venous drainage in children. *Interactive CardioVascular and Thoracic Surgery*. 2018;27(3):422-426.

6. Warden HE, Gustafson RA, Tarnay TJ, Neal WA. An Alternative Method for Repair of Partial Anomalous Pulmonary Venous Connection to the Superior Vena Cava. *The Annals of thoracic surgery*. 1984/12/01/ 1984;38(6):601-605. doi:https://doi.org/10.1016/S0003-4975(10)62317-X

7. Lawrance K, Grimshaw V, Hoyle G, Hicks J, Nixon P, Wooler G. Surgical treatment of atrial septal defect and partial anomalous pulmonary venous drainage. *The Journal of thoracic and cardiovascular surgery*. 1962;43(5):622-633.

8. Said SM, Burkhart HM, Schaff HV, et al. Single-patch, 2-patch, and caval division techniques for repair of partial anomalous pulmonary venous connections: does it matter? *The Journal of thoracic and cardiovascular surgery*. 2012;143(4):896-903.

9. Hijii T, Fukushige J, Hara T. Diagnosis and management of partial anomalous pulmonary venous connection. *Cardiology*. 1998;89(2):148-151.

10. DiBardino DJ, McKenzie ED, Heinle JS, Su JT, Fraser Jr CD. The Warden procedure for partially anomalous pulmonary venous connection to the superior caval vein. *Cardiology in the Young*. 2004;14(1):64.

11. Gustafson RA, Warden HE, Murray GF. Partial anomalous pulmonary venous connection to the superior vena cava. *The Annals of thoracic surgery*. 1995;60:S614-S617.

12. Lin H, Yan J, Wang Q, et al. Outcomes of the warden procedure for partial anomalous pulmonary venous drainage. *Pediatric cardiology*. 2020;41(1):134-140.

13. Shahriari A, Rodefeld MD, Turrentine MW, Brown JW. Caval division technique for sinus venosus atrial septal defect with partial anomalous pulmonary venous connection. *The Annals of thoracic surgery*. 2006;81(1):224-230.

14. Zubritskiy A, Naberukhin Y, Arkhipov A, et al. Outcomes of double-patch and Warden techniques in patients with supracardiac partial anomalous pulmonary venous connection. *Heart, Lung and Circulation*. 2020;29(1):156-161.