Sinus node dysfunction after partial anomalous pulmonary venous connection repair

Carlo Pace Napoleone, MD,a Elisabetta Mariucci, MD,b Emanuela Angeli, MD,a Guido Oppido, MD,a and Gaetano D. Gargiulo, MDa

Objective: Repair of partial anomalous pulmonary venous connection to superior vena cava using an internal patch has been described as a potential cause of obstruction at the systemic or pulmonary vein level and of sinus node dysfunction. Our experience with this operation was reviewed.

Methods: From 1991 to 2011, 59 patients with a diagnosis of partial anomalous pulmonary venous connection to superior vena cava underwent surgical repair with intracardiac patch rerouting alone (45 patients) or with associated superior vena cava patch enlargement (14 patients). Follow-up evaluation was performed, including electrocardiogram, echocardiogram, electrocardiogram Holter monitor recording, and exercise stress test.

Results: There were no early or late deaths and no reoperations at a mean follow-up of 46 ± 45 months. All patients were asymptomatic in New York Heart Association class I. Echocardiographic evaluation excluded any obstruction at the pulmonary or systemic vein level. At follow-up, 55 patients (93%) presented sinus rhythm and were free from antiarrhythmic medications, 2 patients (3%) presented atrial fibrillation, 1 patient (2%) presented atrial fibrillation and asymptomatic sinus node dysfunction, and 1 patient (2%) presented ectopic atrial rhythm. Electrocardiogram Holter recording demonstrated sinus node dysfunction in 6 of 34 patients (18%). Exercise stress test showed chronotropic incompetence in 8 of 27 patients (30%): All except 1 patient presented sinus rhythm at basal electrocardiogram, and only 4 patients had some evidence of sinus node dysfunction on electrocardiogram Holter recording.

Conclusions: Intracardiac repair of partial anomalous pulmonary venous connection can be performed with good results at medium-term follow-up. The rate of sinus node dysfunction or other arrhythmias and obstruction at pulmonary or systemic vein level is comparable to other techniques. Exercise stress test evaluation is the best way to detect asymptomatic sinus node dysfunction. (J Thorac Cardiovasc Surg 2014;147:1594-8)

Sinus venosus atrial septal defects (SV-ASDs), commonly located at the superior vena cava to right atrial (SVC-RA) junction, represent 10% of all atrial septal defects (ASDs) and are associated in approximately 90% of cases with partial anomalous pulmonary venous connections (PAPVCs).1,2 Numerous surgical procedures have been proposed to correct this anomaly.3 The most common operation consists of closing the ASD with a single patch encircling the PAPVC to baffle the pulmonary venous drainage to the left atrium.2 When the PAPVC is committed to the superior vena cava (SVC), a postoperative obstruction of the pulmonary veins, the SVC, or both has been described.5 A second patch across the SVC-RA junction has been proposed to minimize the risk of SVC obstruction but may result in sinoatrial node dysfunction (SND).5

The “Warden procedure” was proposed in 1984 as the solution to these problems.7 In this technique, the SVC is sectioned above the PAPVC and anastomosed to the right atrial (RA) appendage. SV-ASD closure including the entire SVC orifice is then obtained with a patch. Since then, many reports have stated the superiority of the “Warden procedure” over the single- or 2-patch technique in terms of SND or SVC stenosis at follow-up.8,9 At the Bologna Medical School, we have used the single- or 2-patch correction according to the surgeon’s preference, usually depending on the distance between the anomalous pulmonary vein connection and the SVC-RA junction or on the residual SVC caliber after PAPVC baffling. To compare our results with the literature, we retrospectively reviewed our experience with particular attention to SND and SVC or pulmonary vein stenosis at follow-up.

MATERIALS AND METHODS
Patient Population
From 1991 to 2010, 59 consecutive patients underwent surgical repair of PAPVC to SVC at the Pediatric Cardiac Surgery Unit of Bologna Medical School, University of Bologna, Italy. The median age at operation was 14 years (range, 0.6-75 years); 33 patients (57%) were children aged...
Abbreviations and Acronyms

AF = atrial fibrillation
ASD = atrial septal defect
ECG = electrocardiogram
PAPVC = partial anomalous pulmonary venous connection
RA = right atrial
SN = sinoatrial node
SND = sinoatrial node dysfunction
SV- = sinus venosus atrial septal defect
ASD
SVC = superior vena cava
SVC- = superior vena cava to right atrial
RA

Surgical Technique

All patients underwent surgical correction via median sternotomy. The SVC was cannulated close to the innominate vein, and moderately hypothermic (28°C-34°C) cardiopulmonary bypass was started. Cardiac arrest was achieved with cold crystalloid cardioplegia and topical cooling with slush saline. A right atriotomy parallel to the interatrial septum was performed with subsequent reconstruction of the endocardial layer. None of the patients required a permanent pacemaker for bradyarrhythmias.

At follow-up, all patients were asymptomatic and none required reoperation. Echocardiographic evaluation excluded the presence of any residual obstruction or stenosis of the SVC or pulmonary veins and of any shunt. There were no early or late deaths.

RESULTS

At the end of the operation, all patients were free of any residual pressure gradient at the SVC or anomalous pulmonary veins level as shown by intracardiac pressure evaluation. There were no early or late deaths.

At follow-up, all patients were asymptomatic and none required reoperation. Echocardiographic evaluation excluded the presence of any residual obstruction or stenosis of the SVC or pulmonary veins and of any shunt at the atrial septal level. None of the patients required a permanent pacemaker for bradyarrhythmias.

Four adult patients (4/59, 7%; mean age at operation, 62 ± 14 years) presented persistent atrial fibrillation (AF) postoperatively. One of them, a 75-year-old man with persistent AF preoperatively, presented AF with a slow ventricular rate postoperatively and was discharged without antiarrhythmic medications. The other 3 patients were successfully treated with antiarrhythmic medications or electrical cardioversion postoperatively and discharged in sinus rhythm.

At follow-up, 2 patients, aged 70 and 72 years at operation, developed a first episode of persistent AF and were treated with rate-control medications. The 75-year-old patient presented persistent AF with asymptomatic SND (sick sinus syndrome with prevalent bradycardia) at follow-up, but he did not require any antiarrhythmic medications or pacemaker implantation.

One patient, aged 62 years at operation and with postoperatively treated AF, developed an atypical atrial flutter 1 year after operation. He underwent successful transcatheter ablation and was free of antiarrhythmic medications at follow-up.

One young patient, aged 6 years, developed an ectopic atrial tachycardia 6 weeks after surgical operation and was treated successfully with flecainide plus beta-blocker for 6 months with no further recurrence at follow-up.

Briefly, at follow-up with basal 12-lead ECG, 55 patients (93%) were in sinus rhythm and free of antiarrhythmic stress test evaluation because ages less than 10 years and more than 65 years were considered a bias in obtaining a reliable result.

SND was defined as sinus bradycardia, sinus arrest, sinoatrial block, and paroxysmal supraventricular tachyarhythmias alternating with periods of bradycardia or even asystole recorded with standard 12-lead ECG or 24-hour ECG-Holter recording. A minimum heart rate of less than 40 beats/min was considered sinus bradycardia in patients aged more than 16 years, whereas a minimum heart rate of less than 50 beats/min was considered sinus bradycardia in younger patients. A pause more than 3 seconds in adults and more than 3 times the basic cycle length in younger patients was considered pathologic.

To avoid ECG-Holter overdiagnosis of SND in young patients with sinus bradycardia or sinus pauses due to increased vagal tone, we considered ECG-Holter findings consistent with pathologic SND only if even exercise stress test documented chronotropic incompetence in the same patient. Exercise stress test was considered consistent with SND if chronotropic incompetence was documented, such as if the patient was unable to reach a heart rate 85% or greater of theoretic maximal value for age with maximal effort.

RESULTS

At the end of the operation, all patients were free of any residual pressure gradient at the SVC or anomalous pulmonary veins level as shown by intracardiac pressure evaluation. There were no early or late deaths.

At follow-up, all patients were asymptomatic and none required reoperation. Echocardiographic evaluation excluded the presence of any residual obstruction or stenosis of the SVC or pulmonary veins and of any shunt at the atrial septal level. None of the patients required a permanent pacemaker for bradyarrhythmias.

Four adult patients (4/59, 7%; mean age at operation, 62 ± 14 years) presented persistent atrial fibrillation (AF) postoperatively. One of them, a 75-year-old man with persistent AF preoperatively, presented AF with a slow ventricular rate postoperatively and was discharged without antiarrhythmic medications. The other 3 patients were successfully treated with antiarrhythmic medications or electrical cardioversion postoperatively and discharged in sinus rhythm.

At follow-up, 2 patients, aged 70 and 72 years at operation, developed a first episode of persistent AF and were treated with rate-control medications. The 75-year-old patient presented persistent AF with asymptomatic SND (sick sinus syndrome with prevalent bradycardia) at follow-up, but he did not require any antiarrhythmic medications or pacemaker implantation.

One patient, aged 62 years at operation and with postoperatively treated AF, developed an atypical atrial flutter 1 year after operation. He underwent successful transcatheter ablation and was free of antiarrhythmic medications at follow-up.

One young patient, aged 6 years, developed an ectopic atrial tachycardia 6 weeks after surgical operation and was treated successfully with flecainide plus beta-blocker for 6 months with no further recurrence at follow-up.

Briefly, at follow-up with basal 12-lead ECG, 55 patients (93%) were in sinus rhythm and free of antiarrhythmic stress test evaluation because ages less than 10 years and more than 65 years were considered a bias in obtaining a reliable result.

SND was defined as sinus bradycardia, sinus arrest, sinoatrial block, and paroxysmal supraventricular tachyarhythmias alternating with periods of bradycardia or even asystole recorded with standard 12-lead ECG or 24-hour ECG-Holter recording. A minimum heart rate of less than 40 beats/min was considered sinus bradycardia in patients aged more than 16 years, whereas a minimum heart rate of less than 50 beats/min was considered sinus bradycardia in younger patients. A pause more than 3 seconds in adults and more than 3 times the basic cycle length in younger patients was considered pathologic.

To avoid ECG-Holter overdiagnosis of SND in young patients with sinus bradycardia or sinus pauses due to increased vagal tone, we considered ECG-Holter findings consistent with pathologic SND only if even exercise stress test documented chronotropic incompetence in the same patient. Exercise stress test was considered consistent with SND if chronotropic incompetence was documented, such as if the patient was unable to reach a heart rate 85% or greater of theoretic maximal value for age with maximal effort.
medications, 2 patients (3%) were taking rate-control medications for persistent AF, 1 patient (2%) presented AF and asymptomatic sick sinus syndrome with prevalent bradycardia, and 1 patient (2%) presented ectopic atrial rhythm.

A 24-hour ECG-Holter monitor recording was obtained in 34 patients, and an exercise stress test was performed in 27 patients at a mean time from operation of 10±5 years. Twenty-four–hour ECG-Holter monitor recording demonstrated SND in 6 patients (6/34, 18%); mean age, 40±23 years). The results are summarized in Table 1, in which the patient population is divided into 2 groups according to their age, because different cutoff values are considered for patients aged less than or more than 16 years.

The exercise stress test showed chronotropic incompetence in 8 patients (8/27, 30%); mean age, 23±13 years). All patients except 1 presented sinus rhythm with normal heart rate at basal ECG, and only 4 of them have some evidence of SND at ECG-Holter recording (Table 2).

No correlation was evident between the surgical variables and the incidence of SND at follow-up (Table 3). In particular, age at operation was not statistically significant in affecting SND incidence and surgical strategy (Table 4).

**DISCUSSION**

Although surgical repair of PAPVC has been performed with excellent early outcomes for multiple decades, there have been numerous concerns about postoperative problems related to the obstruction of the SVC or pulmonary veins after baffle redirection of pulmonary venous blood into the left atrium or to anastomotic strictures after implantation of the anomalous pulmonary veins directly on the left atrium. An additional concern is related to postoperative dysrhythmias that have been reported in multiple series after surgical repair of PAPVC.

Historically, the most common operation for SV-ASD with PAPVC to the RA or RA-SVC junction consists of closing the ASD with a single atrial patch that includes the PAPVC, thereby baffling the pulmonary venous drainage to the left atrium. However, if the PAPVC is committed to the SVC, baffling by a single patch can result in obstruction to the pulmonary veins or the SVC, or both. In this case, the necessity to carry the atrial incision into the SVC up to the anomalous pulmonary vein entrance may result in SND by disrupting the sinoatrial node (SN) or the SN artery. Moreover, in cases of long incision or small residual SVC caliber, a second patch across the SVC-RA

**TABLE 1. Results of 24-hour electrocardiogram Holter monitor recording**

<table>
<thead>
<tr>
<th>Age &lt;16 y</th>
<th>Age ≥16 y</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patients</td>
<td>8</td>
</tr>
<tr>
<td>Age (y)</td>
<td>13 ± 3 (9-16)</td>
</tr>
<tr>
<td>Medications*</td>
<td>0</td>
</tr>
<tr>
<td>SND-related symptoms</td>
<td>0</td>
</tr>
<tr>
<td>Mean HR: 24 h (beats/min)</td>
<td>90 ± 11 (79-108)</td>
</tr>
<tr>
<td>Minimum HR (beats/min)</td>
<td>59 ± 20 (27-97)</td>
</tr>
<tr>
<td>Minimum HR &lt;50 beats/min, patients (%)</td>
<td>1 (12.5%)</td>
</tr>
<tr>
<td>Minimum HR &lt;40 beats/min, patients (%)</td>
<td>—</td>
</tr>
<tr>
<td>Sinus arrest or sinoatrial block, patients (%)</td>
<td>0</td>
</tr>
<tr>
<td>AF/AFL/AT episodes or frequent SVEB, patients (%)</td>
<td>0</td>
</tr>
</tbody>
</table>

*Antiarrhythmic or antihypertensive medications with bradycardic effects.

**TABLE 2. Exercise stress test results**

<table>
<thead>
<tr>
<th>Mean value ± SD (range) or %</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patients</td>
</tr>
<tr>
<td>Age (y)</td>
</tr>
<tr>
<td>Medications*</td>
</tr>
<tr>
<td>Peak effort HR (% of theoretic value)</td>
</tr>
<tr>
<td>Chronotropic incompetence</td>
</tr>
</tbody>
</table>

*Antiarrhythmic or antihypertensive medications with bradycardic effects. Unable to reach HR ≥85% of theoretic value at peak effort, excluded patients with medications.
A more recent report compared the single-patch, double-patch, and Warden technique in 54 patients. The incidence of low atrial or junctional rhythm was significantly higher with the double-patch repair (55%) compared with the single-patch (24%) and Warden (0%) repairs. No differences were evidenced in terms of SVC stenosis among the 3 techniques.8

Many reports in the literature seem to corroborate the finding that the patch technique, and above all the double-patch technique, is associated with a high incidence of SND on short- and mid-term follow-ups, whereas the Warden procedure essentially eliminates SN injury and thus SND.

It is important to underline that in all these reports the SND diagnosis was based on symptoms and 12-lead ECG. As demonstrated by our experience, the actual SND prevalence can be largely underestimated. Limiting analysis to symptoms and ECG evaluation would have disclosed only 2 patients with SND in our experience, with a prevalence of 3% (2/59 patients) at a mean term follow-up, a result that can be easily compared with that of Warden technique reported in the literature.4,6-8,9,11-13

SND diagnosis was based on 24-hour Holter evaluation at follow-up by Agarwal and colleagues,14 who reported no incidence of SND among 58 patients undergoing the Warden operation. In our experience, this value was greater because 24-hour Holter disclosed 6 of 34 patients (18%) who reached the criteria for SND.

Although many experienced clinicians claim to recognize SND in individual patients, no single metric has been established as a diagnostic standard.10 Regardless, we think that the most accurate way is to perform a more comprehensive evaluation with ECG, ECG-Holter, and exercise stress test because of the intermittent nature of the SND episodes. In this way, we observed a different incidence of SND (8/27 patients, 30%), which we considered the real incidence of rhythm problems after single- or double-patch repair of this heart disease.

To the best of our knowledge, our report is the first to apply a global evaluation of SND in patients undergoing operation for PAPVC. Without this approach, all other reports may underestimate the incidence of SND and, for this reason, are not comparable to our results. We hope that our work will be shared by other researchers to disclose the correct incidence of SND after PAPVC surgical correction.

**Study Limitations**

This report presents the well-known limitations of a retrospective and nonrandomized study. We are aware

---

**TABLE 3. Correlation between surgical and clinical variables and postoperative sinus node dysfunction**

<table>
<thead>
<tr>
<th></th>
<th>SND</th>
<th>SR</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patients</td>
<td>8</td>
<td>51</td>
<td></td>
</tr>
<tr>
<td>Age (y)</td>
<td>13.1 ± 12.1</td>
<td>25.7 ± 22.7</td>
<td>NS</td>
</tr>
<tr>
<td>Surgically created ASD</td>
<td>1/8 (12.5%)</td>
<td>9/51 (17.6%)</td>
<td>NS</td>
</tr>
<tr>
<td>2-patch technique</td>
<td>1/8 (12.5%)</td>
<td>13/51 (25.4%)</td>
<td>NS</td>
</tr>
<tr>
<td>Baffle material</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pericardial</td>
<td>5/8 (62.5%)</td>
<td>43/51 (84.3%)</td>
<td>NS</td>
</tr>
<tr>
<td>Teflon</td>
<td>3/8 (37.5%)</td>
<td>8/51 (15.7%)</td>
<td>NS</td>
</tr>
<tr>
<td>Postoperative rhythm problem</td>
<td>0/8</td>
<td>8/51 (15.7%)</td>
<td>NS</td>
</tr>
</tbody>
</table>

ASD, Atrial septal defect; NS, not significant; SND, sinus node dysfunction; SR, sinus rhythm.

---

**TABLE 4. Correlation between age and surgical variables**

<table>
<thead>
<tr>
<th></th>
<th>Yes</th>
<th></th>
<th>No</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No.</td>
<td>Mean age</td>
<td>No.</td>
<td>Mean age</td>
</tr>
<tr>
<td>Surgically created ASD</td>
<td>10</td>
<td>20.9 y</td>
<td>49</td>
<td>24.7 y</td>
</tr>
<tr>
<td>2-patch technique</td>
<td>14</td>
<td>18.6 y</td>
<td>45</td>
<td>25.7 y</td>
</tr>
</tbody>
</table>

ASD, Atrial septal defect; NS, not significant.
that the incidence of chronotropic incompetence can be overestimated if the test is not driven to maximal effort, and even ECG-Holter can lead to overdiagnosis of SND in young patients with sinus bradycardia or sinus pauses due to increased vagal tone. Regardless, we believe that SND can remain undetected without these tests. We did our best to perform maximal effort and symptom-driven exercise tests and tried to avoid ECG-Holter SND overdiagnosis by performing a complete evaluation of all clinical data (eg, considering ECG-Holter findings consistent with pathologic SND only if even exercise stress test documented chronotropic incompetence in the same patient).

CONCLUSIONS

Our data report a short-term incidence of SND with the single- or double-patch technique almost comparable to the reported experiences with the Warden procedure. Nevertheless, the long-term incidence of SND after PAPVC repair can be approximately 20% to 30% and may warrant long-term follow-up with ECG-Holter monitor recordings and exercise stress test even if the patients are asymptomatic, because SND can be demonstrated only with an extensive and accurate evaluation. To define the true late effects on rhythm and to better compare the 2 techniques, further studies reporting long-term follow-up of patients receiving the Warden technique with ECG-Holter monitor recordings and exercise stress test are necessary.

References