Echocardiogram Unmasked Hemodynamic Advantage of Atrial Pacing in Securing Ventricular Preload in a Fontan Patient with Junctional Rhythm

Clinical Implications to Preserve Supraventricular Rhythm

Hirofumi Saiki,1,2 MD, Kohei Kawada,1 MD, Seiko Kuwata,1 MD, Manabu Takanashi,1 MD, Takuma Fukunishi,1 MD, Kagami Miyaji,3 MD and Hideaki Senzaki,1,4 MD

Summary

While the advancement of perioperative management has expanded Fontan candidacy, not all patients have a successful postoperative course. Our case was a right isomerism patient who could not leave the ICU due to high central venous pressure and low output syndrome. Initial observation of the monitor ECG showed his rhythm to be supraventricular, however, an echocardiogram indicated simultaneous contraction of the atrium and ventricle, implying a junctional rhythm. While neither central venous pressure nor blood pressure improved with temporary pacing, better central venous and pulmonary venous blood flow patterns during pacing unraveled its positive impact. The patient successfully left the ICU after permanent pacing implantation. Hemodynamic study revealed a beneficial impact of atrial pacing in securing cardiac output and ventricular preload, lowering central venous pressure, and shortening blood transit time, which is partly attributed to the optimization of the fenestration function in reservation of the preload. Our case emphasizes the significant advantage of atrial pacing in a failing Fontan patient with junctional rhythm by reducing venous congestion and maximizing the benefit of fenestration.

(Key word: Fontan circulation, Hemodynamics, Fenestration, Blood transition time)

Case Report

The case was a one-year old boy diagnosed with right isomerism, a single atrium, and a single ventricle with pulmonary atresia. The fenestrated Fontan procedure using an extracardiac conduit cavopulmonary anastomosis was performed. Cardiac catheterization before surgery revealed a pulmonary vascular resistance of 1.16 U · m² and mean pulmonary pressure of 11 mmHg without significant atrioventricular valve regurgitation. After his arrival in the ICU, he went under ECMO support due to newly developed common atrioventricular valve regurgitation (CAVVR). After a week, the CAVVR was surgically controlled and ECMO was discontinued. Despite a tiny CAVVR with an optimal ventricular ejection fraction of more than 65%, the central venous pressure (CVP) was consistently as high as 22 to 25 mmHg. No fenestration signal was detected. While his blood pressure was optimal, it was dependent on CVP and the systemic edema could not be resolved. Although we attempted to dehy-
drate him using diuretics (furosemide 3 mg/kg, traseamide 0.2 mg/kg, trichloromethiazide 0.5 mg/kg and tolvaptan 0.4 mg/kg) with preservation of ventricular preload using pulmonary vasodilators (sildenafil 2 mg/kg, macitentan 0.4 mg/kg and selexipag 0.02 mg/kg), his condition barely improved for more than a week. While we had assumed his rhythm was a supraventricular rhythm, as it appeared to have a tiny p wave with marked heart rate variability, we became skeptical of this following examination of the echocardiogram, which revealed simultaneous contraction of the atrium and ventricle (Figure 1, Figure 2 ECG original rhythm). Based on these findings, temporary atrial pacing with the minimal possible rate was attempted, however, improvement of the hemodynamic index including blood pressure (BP), CVP and central venous oxygen saturation (SvO2) was imperceptible. To verify the advantage of pacing, we conducted an echocardiogram during temporary pacing (Figure 2). As expected, the echocardiogram showed amelioration of the inferior venous flow pattern and suppressed pulmonary venous reverse flow. As the venous flow pattern during temporal atrial pacing suggested possible benefits on the venous decongestion and blood pressure fluctuation after urination appeared to be suppressed for more than a day, a permanent pacemaker was implanted when he underwent reconstruction of the fenestration. As expected, atrial pacing stabilized his hemodynamics with a lower CVP than before and he was able to leave the ICU. Cardiac catheterization was performed after 2 weeks of PMI. Under AAI mode, the cardiac index was as high as 3.7 L/minute/m², which decreased by 30% when off pacing (Table). With a similar heart rate (HR), ventricular stroke volume was markedly high with pacing, suggesting there was an advantage in preserving ventricular preload. The mean blood transition time (MTT) by indocyanine green (DDG-3300, Nihon Koden) markedly shortened with atrial pacing, which confirmed the advantages in smooth venous rheology. Simultaneous blood pressure measurements of the LV and central venous or pulmonary arterial wedge (PAW) pressure also revealed benefits in increasing systolic pressure and suppressing CVP and estimated left atrial pressure in atrial pacing (Figure 3). Since termination of the atrial pacing also changed the fenestration flow pattern from right-left shunting to bidirectional shunting, amelioration of the preload reserve was attributed to both the suppression of atrial pressure and enhanced function of the fenestration. At this point, he was free of edema and left the hospital with decreased diuretics (furosemide 2 mg/kg/day). We set his pacemaker to AAI mode with 130 bpm and continued the same combination of pulmonary vasodilators (sildenafil 2 mg/kg, macitentan 0.4 mg/kg, and selexipag 0.02 mg/kg) to minimize venous congestion. Six months after discharge, cardiac catheterization to reconfirm the hemodynamic advantage was performed. The CVP, PA wedge pressure, systemic BP and SvO2 on the pacing were 6 mmHg, 3 mmHg, 89/42 (62) mmHg and 62.1%, respectively, which was almost unchanged compared to under off pacing (7 mmHg, 4 mmHg, 90/41 (61) mmHg, and 65.9%, respectively) and was apparently a supraventricular rhythm (Figure 4).

**Discussion**

Of the original 10 commandments for the Fontan procedure by Choussat, sinus rhythm is the second. While some of the 10 commandments can be weighted less owing to advanced perioperative care, clinical difficulties demand us to recall them. In the present case, the acute advantage of atrial pacing was unremarkable in CVP and arterial pressure, whereas the echocardiogram identified a marked improvement in venous flow and suppressed pulmonary venous reverse flow. The echocardiogram findings in Figures 2, 3 suggested simultaneous contraction of the atrium and ventricle during junctional rhythm, which augmented an atrial pressure increase during ventricular contraction, impeding pulmonary venous return. Atrial pacing might have generated the physiological reciprocal contractions of the atrium and ventricle, and lowered the atrial...
pressure during the ventricular systolic phase. The catheter exam clearly indicated such an advantage of supraventricular rhythm in the Fontan hemodynamics, which was also a significant benefit to maximize the impact of fenestration and improve venous congestion assessed by MTT. As an acute pacing trial could not clearly indicate any hemodynamic advantage in lowering CVP or increasing blood pressure, atrial pacing alone might be insufficient to completely improve hemodynamics. Meanwhile, as fenestration should have improved the patient’s hemodynamics during their stay in ICU, a combination of atrial pacing and fenestration should work synergistically to maximize the impact of fenestration, as implied by the improvement in hemodynamics seen during cardiac catheterization.

The unfavorable impact of junctional rhythm on the Fontan circulation has been emphasized for years. Hasselman, et al. measured ventricular and central venous pressure waveforms simultaneously in fenestrated Fontan patients and found reversal of fenestration flow in the Fontan circulation during non-sinus rhythm.\(^5\) We also observed reversal of fenestration during junctional rhythm which might have been attributed to the natural closure of fenestration. Indeed, the stiff ventricular property identified by the pressure-volume analyses was associated with natural closure of the fenestration in Fontan patients.\(^4\) Based on these reports and the fact that fenestration was naturally closed despite a high CVP of more than 22 mmHg, we were convinced of the necessity for permanent

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**Table. Hemodynamic Impact of the Atrial Pacing**

<table>
<thead>
<tr>
<th></th>
<th>AAI 130 bpm</th>
<th>AAI 100 bpm</th>
<th>off 95 bpm</th>
</tr>
</thead>
<tbody>
<tr>
<td>CI (L/minute/m²)</td>
<td>3.03</td>
<td>2.74</td>
<td>2.21</td>
</tr>
<tr>
<td>SV index (mL/m²)</td>
<td>23.3</td>
<td>27.4</td>
<td>23.2</td>
</tr>
<tr>
<td>LVEDVI (mL/m²)</td>
<td>40.2</td>
<td>45.6</td>
<td>38.7</td>
</tr>
<tr>
<td>LVP (mmHg)</td>
<td>77/EDP 6</td>
<td>80/EDP 9</td>
<td>72/EDP 7</td>
</tr>
<tr>
<td>Wedge (mmHg)</td>
<td>4</td>
<td>6</td>
<td>7</td>
</tr>
<tr>
<td>CVP (mmHg)</td>
<td>8</td>
<td>8</td>
<td>10</td>
</tr>
<tr>
<td>MTT (seconds)</td>
<td>20.1</td>
<td>17.2</td>
<td>32.8</td>
</tr>
</tbody>
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CI indicates cardiac index; CVP, central venous pressure; LVEDVI, left ventricular end diastolic volume index; LVP, left ventricular systolic pressure; and MTT, mean transit time using ICG.

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**Figure 2.** Different flow patterns of the pulmonary vein and inferior vena cava by atrial pacing. The finding of test pacing is shown. The p wave in the original heart rate image appeared to preceede the QRS wave, but the atrium and ventricle appeared to contract simultaneously, as indicated in Figure 1. As compared with atrial pacing, the PV flow pattern under original rhythm had a large inflow waveform to the atrium with marked enlarged atrial reverse flow. Such a flow pattern appeared to have contributed to alternation of the flow pattern in the IVC.
atrial pacing for this case. Importantly, maintaining the fenestration flow might be key to avoid end-organ dysfunction in Fontan patients.\textsuperscript{7} While Ceresnak, \textit{et al.} defined a responder of atrial pacing as an increase of systolic blood pressure by 10 mmHg and improvement of urine output or blood gas,\textsuperscript{8} an echocardiogram was a useful tool for identifying the advantages of pacing in our case. Indeed, CVP or blood pressure was almost the same in the acute reaction by atrial pacing.

Sinus node dysfunction is not rare, considering 21-59\% of postoperative Fontan patients developed it later in their life.\textsuperscript{7,8} Ceresnak, \textit{et al.} reported that atrial pacing had a hemodynamic advantage in 20\% of Fontan patients early after operation,\textsuperscript{8} and Cohen, \textit{et al.} reported a positive impact of atrial pacing in the treatment for protein losing enteropathy, one of the manifestations of Fontan related to end organ dysfunction.\textsuperscript{9} These reports suggest it has a therapeutic or prophylactic role for Fontan failure. While the dependence of atrial pacing disappeared in the subsequent assessments in our case, we decided to continue atrial pacing with pharmacological HR suppression so that the patient would avoid developing heart failure or end organ dysfunction in the future via the ventricular energetic disadvantage or possible activation of cardiovascular fibrogenesis.\textsuperscript{10,11} As an increase in HR might counter the advantages of having fenestration in the preload reserve,\textsuperscript{4} an indication for pacing therapy to ameliorate the hemodynamics requires careful consideration to weigh the advantages of securing the preload reserve and disadvantage of increasing HR.

Conclusions

Determining blood flow patterns by echocardiography may provide a clue to identifying the hemodynamic advantages of atrial pacing. In Fontan patients with junctional rhythm, atrial pacing may improve circulation, both by increasing the ventricular preload and improving venous return.
Simultaneous measurement of ascending aorta and central venous pressure with and without atrial pacing. The pacing dependence of systolic pressure and central venous pressure observed in Figure 3 disappeared 1 year after Fontan completion. The supraventricular rhythm was recovered at this point.

Disclosure

Conflicts of interest: No conflicts of interest to declare.

References

5. Oka N, Miyamoto T, Tomoyasu T, Hayashi H, Miyaji K. Risk Factors for Mid-Term Liver Disease After the Fontan Procedure.