Batista et al recently reported success using partial left ventriculectomy (PLV) to treat end-stage dilated cardiomyopathy (DCM), but this procedure has not been widely accepted as a therapeutic option, and PLV is rarely performed in children. Several factors have limited the acceptance of PLV for children with DCM, including, (1) pediatric cardiologists’ lack of confidence in an unproven procedure, (2) candidates for PLV are extremely ill and the surgical mortality rate is expected to be high, (3) functional improvement after PLV is inferior to that of heart transplantation, and (4) the operation does not treat the underlying disease, so it is questionable whether the patient will thrive even if the operation is successful. Perhaps the most important factor, however, is that the indications for PLV in children with DCM have not been discussed objectively.

From 1998 to 2000, we performed PLV in 4 children with end-stage DCM. The decision to perform this procedure was based on the patient’s clinical condition and social situation, as determined by their primary physician. We present here the role, indications and recommendations for PLV in children with DCM based on a retrospective comparison of children who underwent PLV and children who were treated medically.

**Methods**

Subjects

Between 1997 and 2000, we treated 9 children with DCM ranging in age from 8 months to 15 years. Four patients underwent PLV (PLV group) and 5 patients were managed medically (non-PLV group). The intermediate-term outcome of the PLV group has been reported previously.

Methods

Data from the PLV group and the non-PLV group were obtained by retrospective chart review. We analyzed several factors affecting outcome, including, (1) pediatric cardiologists’ lack of confidence in an unproven procedure, (2) candidates for PLV are extremely ill and the surgical mortality rate is expected to be high, (3) functional improvement after PLV is inferior to that of heart transplantation, and (4) the operation does not treat the underlying disease, so it is questionable whether the patient will thrive even if the operation is successful. Perhaps the most important factor, however, is that the indications for PLV in children with DCM have not been discussed objectively.

From 1998 to 2000, we performed PLV in 4 children with end-stage DCM. The decision to perform this procedure was based on the patient’s clinical condition and social situation, as determined by their primary physician. We present here the role, indications and recommendations for PLV in children with DCM based on a retrospective comparison of

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**Key Words:** Batista’s operation; Children; Dilated cardiomyopathy; Partial left ventriculectomy
Table 1 Preoperative Clinical Characteristics of the Children With Dilated Cardiomyopathy Who Underwent Partial Left Ventriculectomy

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age/sex</th>
<th>NYHA</th>
<th>CTR (%)</th>
<th>LVDD (%)</th>
<th>LVEF (%)</th>
<th>LVEDP/PCWP (mmHg)</th>
<th>BNP (pg/ml)</th>
<th>MR/TR</th>
<th>Inotrops &amp; ventilator</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>8m/F</td>
<td>IV</td>
<td>73</td>
<td>218</td>
<td>10</td>
<td>12</td>
<td>1,400</td>
<td>Moderate/trivial</td>
<td>DOA 5 µg, DOB 5 µg</td>
</tr>
<tr>
<td>2</td>
<td>3y5m/M</td>
<td>III</td>
<td>75</td>
<td>184</td>
<td>17</td>
<td>13</td>
<td>1,290</td>
<td>Moderate/trivial</td>
<td>(–)</td>
</tr>
<tr>
<td>3</td>
<td>2y2m/F</td>
<td>IV</td>
<td>76</td>
<td>215</td>
<td>12</td>
<td>20</td>
<td>1,300</td>
<td>Moderate/trivial</td>
<td>DOA 3 µg, DOB 3 µg, Preop. 5 days</td>
</tr>
<tr>
<td>4</td>
<td>2y5m/F</td>
<td>III</td>
<td>72</td>
<td>198</td>
<td>22</td>
<td>30</td>
<td>4,000&lt;</td>
<td>Severe/moderate</td>
<td>(–)</td>
</tr>
</tbody>
</table>

NYHA, New York Heart Association functional class; CTR, cardiothoracic ratio on chest roentgenogram; LVDD (%), % of predicted normal left ventricular end diastolic dimension on echocardiography; LVEF, left ventricular ejection fraction; LVEDP/PCWP, left ventricular end diastolic pressure or pulmonary capillary wedge pressure; BNP, serum brain natriuretic peptide concentration; MR, mitral regurgitation; TR, tricuspid regurgitation; DOA, dopamine; DOB, dobutamine.

Table 2 Clinical Characteristics of the Children With Dilated Cardiomyopathy Who Did Not Undergo Partial Left Ventriculectomy

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age/sex</th>
<th>NYHA</th>
<th>CTR (%)</th>
<th>LVDD (%)</th>
<th>LVEF (%)</th>
<th>LVEDP/PCWP (mmHg)</th>
<th>BNP (pg/ml)</th>
<th>MR/TR</th>
<th>Inotrops &amp; ventilator</th>
</tr>
</thead>
<tbody>
<tr>
<td>5</td>
<td>10m/F</td>
<td>II</td>
<td>60</td>
<td>133</td>
<td>55</td>
<td>18/14</td>
<td>124</td>
<td>Severe/trivial</td>
<td>(–)</td>
</tr>
<tr>
<td>6</td>
<td>12y/M</td>
<td>I</td>
<td>45</td>
<td>114</td>
<td>36</td>
<td>/</td>
<td>7.8</td>
<td>(–) / (–)</td>
<td>(–)</td>
</tr>
<tr>
<td>7</td>
<td>2y9m/M</td>
<td>II</td>
<td>59</td>
<td>163</td>
<td>37</td>
<td>12/15</td>
<td>168</td>
<td>Moderate (–)</td>
<td>(–)</td>
</tr>
<tr>
<td>8</td>
<td>15y/M</td>
<td>II</td>
<td>46</td>
<td>/</td>
<td>56</td>
<td>/</td>
<td>6.9</td>
<td>(–) / (–)</td>
<td>(–)</td>
</tr>
<tr>
<td>9</td>
<td>14y/M</td>
<td>II</td>
<td>49</td>
<td>109</td>
<td>39</td>
<td>/</td>
<td>26.5</td>
<td>(–) / (–)</td>
<td>(–)</td>
</tr>
</tbody>
</table>

NYHA, New York Heart Association functional class; CTR, cardiothoracic ratio on chest roentgenogram; LVDD (%), % of predicted normal left ventricular end diastolic dimension on echocardiography; LVEF, left ventricular ejection fraction; LVEDP/PCWP, left ventricular end diastolic pressure or pulmonary capillary wedge pressure; BNP, serum brain natriuretic peptide concentration; MR, mitral regurgitation; TR, tricuspid regurgitation.

Table 3 Intermediate Outcome and Current Status of the Children With Dilated Cardiomyopathy Who Underwent Partial Left Ventriculectomy

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age/sex</th>
<th>Intermediate outcome</th>
<th>Current status</th>
<th>CTR (%)</th>
<th>LVDD (%)</th>
<th>LVEF (%)</th>
<th>LVEDP/PCWP (mmHg)</th>
<th>BNP (pg/ml)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>8m/F</td>
<td>6 months later, underwent transplantation</td>
<td>NYHA I</td>
<td>57</td>
<td>160</td>
<td>35</td>
<td>15/21</td>
<td>43</td>
</tr>
<tr>
<td>2</td>
<td>3y5m/M</td>
<td>NYHA II</td>
<td>35 months NYHA I</td>
<td>57</td>
<td>160</td>
<td>35</td>
<td>15/21</td>
<td>43</td>
</tr>
<tr>
<td>3</td>
<td>2y2m/F</td>
<td>Weaned after bronchial stenting, died of hemoptysis 16 POD</td>
<td>NYHA II</td>
<td>69</td>
<td>114</td>
<td>46</td>
<td>24/34</td>
<td>390</td>
</tr>
<tr>
<td>4</td>
<td>2y5m/F</td>
<td>NYHA III</td>
<td>18 months NYHA II</td>
<td>69</td>
<td>114</td>
<td>46</td>
<td>24/34</td>
<td>390</td>
</tr>
</tbody>
</table>

CTR, cardiothoracic ratio on chest roentgenogram; LVDD (%), % of predicted normal left ventricular end diastolic dimension on echocardiography; LVEF, left ventricular ejection fraction; LVEDP/PCWP, left ventricular end diastolic pressure or pulmonary capillary wedge pressure; BNP, serum brain natriuretic peptide concentration; NYHA, New York Heart Association functional class; POD, postoperative day.

Results

The clinical characteristics of the PLV group are summarized in Table 1. Patients in the PLV group ranged in age from 8 months to 3 years and 5 months. Two were rated NYHA class III, and 2 were class IV. The CTR in the PLV group was 74±1.8%, ranging from 72% to 76%, the LVDD (%) was 204±16%, ranging from 184% to 218%, and the LVEF was 15.3±5.4% ranging from 10% to 22%. All 4 patients underwent cardiac catheterization preoperatively, and their left ventricular end diastolic pressure (LVEDP) or pulmonary capillary wedge pressure (PCWP) were between 12 and 30 mmHg. The BNP was greater than 1,200 pg/ml in each case. All had moderate to severe mitral regurgitation.

One patient (case 1) was maintained on moderate doses of catecholamines to manage borderline cardiogenic shock and underwent semi-emergency PLV. She recovered well from severe heart failure and remained well for 3 months after the surgery. However, her body weight subsequently increased as her heart failure gradually worsened, and she underwent emergency heart transplantation in the USA 6 months after PLV. The patient is now well 3 years after surgical repair.
transplantation. Another patient (case 3) also was maintained on catecholamines but required additional mechanical ventilation for 5 days preoperatively to manage cardiogenic shock. This patient underwent emergency PLV and did well for 2 weeks postoperatively but after bronchial stenting for left main bronchial stenosis, she developed hemoptyis and died suddenly. The 2 patients rated in NYHA class III received continuous intravenous infusions of diuretics and underwent PLV electively. These children are alive and well 18 and 35 months postoperatively. The families of these 2 patients rejected the option of heart transplantation because of social and economic considerations. In all 4 cases, the postoperative condition correlated with the preoperative status. The intermediate and current outcomes in the PLV group are summarized in Table 3.

The clinical characteristics of the non-PLV group are summarized in Table 2. Patients were between 10 months and 15 years of age, and the DCM was well-controlled by medical therapy alone in 4 of the 5 cases. One patient had severe mitral regurgitation and required mitral valve replacement when she was 1 year old.8

The CTR in the non-PLV group was 51.8±7.2%, ranging from 45% to 60%, the LVDd (%) was 137±23%, ranging from 109% to 163%, and the LVEF was 44.6±10.0%, ranging from 36% to 56%. Two patients had undergone cardiac catheterization, and their LVEDP or PCWP ranged from 12 to 18 mmHg. The BNP were less than 168 pg/ml in all cases. In addition to the patient who underwent mitral valve replacement for severe mitral regurgitation, only one other patient had moderate mitral regurgitation.

Differences in the CTR, LVDd (%), LVEF, and BNP between the PLV and non-PLV groups are shown in Fig 1. Every parameter was statistically different in both groups. The dark areas are the zones that we previously identified as tentative indications for PLV in end-stage pediatric DCM.3,5 As can be seen, these parameters are different in the 2 groups, so the criteria appear to be realistic.

The changes with PLV in CTR, LVDd (%), LVEF, and BNP among the patients in the PLV group are shown in Fig 2. Each parameter fell into the range of values that were managed medically. The 1 patient who underwent heart transplantation initially achieved a satisfactory reduction in the BNP, but it subsequently rose as her condition deteriorated.

**Discussion**

A heart transplantation program was recently started
in Japan, but legal problems preclude transplantation in infants. Therefore, PLV may be a reasonable treatment option for children with end-stage DCM who have no access to transplantation. There are now 6 case reports of PLV performed in children,1–4 of whom had DCM,1–3 including a 3-day-old boy. However, the appropriateness of PLV in this neonate has been questioned,15 which highlights the need to develop objective criteria for performing PLV in pediatric patients.

Based on our experience of performing PLV in 4 patients, we identified several factors that determine its appropriateness as therapy for pediatric DCM. We feel most strongly that patients in NYHA class III or IV who are hospitalized for heart failure and require heroic doses of catecholamines should undergo PLV before mechanical ventilation is needed. Less absolute criteria include a CTR ≥70%, LVEDd (%) ≥170%, LVEF ≤20%, and BNP ≥1,000 pg/ml.

These factors were generated from our retrospective analysis of the data from the PLV group. When we retrospectively applied these criteria to patients who were managed successfully by medical therapy during the same period, none of them would have been surgical candidates. Thus, these criteria provide an objective foundation that supports our surgical philosophy; that is, PLV should be reserved for patients whose clinical situation is deteriorating despite maximum medical therapy, but surgery should not be withheld so long that the patient becomes desperately ill.

We believe that PLV may be the definitive treatment in some cases, a bridge to heart transplantation in others, or a way to improve the quality of life and prolong survival. PLV also has been performed in patients with congenital massive dilatation of the left ventricle with systolic dysfunction13 and anomalous left coronary artery from the pulmonary artery with endocardial fibroelastosis.14

Considering the number of children suffering from DCM who have little chance of undergoing heart transplantation abroad because of economical constraints or the legal constraints in Japan, we recommend PLV as a reasonable treatment option when medical therapy is not effective.

References