Guidance Addressing the Use of Pediatric Heart Exceptions

OPTN Heart Transplantation Committee
Purpose of Guidance Document

- Increase equal access to transplant for medically urgent, unique pediatric candidates

- Provide consistent guidance for review of following diagnoses:
  - Dilated cardiomyopathy
  - Hypertrophic or restrictive cardiomyopathy
  - Single ventricle heart disease
  - Coronary allograft vasculopathy and retransplantation

- Assist in activities of the National Heart Review Board for pediatric candidates

- Increase efficiency of review process by providing guidance on specific information to include in an exception request
Pediatric Heart Transplants at All Statuses by Diagnosis

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Pre-Policy (3/22/15 - 3/21/16)</th>
<th>Post-Policy (10/1/16 - 12/31/17)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cardiomyopathy</td>
<td>205</td>
<td>224</td>
</tr>
<tr>
<td>Congenital Heart Defect (CHD)</td>
<td>195</td>
<td>258</td>
</tr>
<tr>
<td>Other</td>
<td>43</td>
<td>47</td>
</tr>
</tbody>
</table>
Pediatric Heart Transplants at Status 1A by Exception and Diagnosis

Cardiomyopathy

- Pre-Policy (3/22/15 - 3/21/16): 7
- Post-Policy (10/1/16 - 12/31/17): 38

CHD

- Pre-Policy (3/22/15 - 3/21/16): 11
- Post-Policy (10/1/16 - 12/31/17): 9

Other

- Pre-Policy (3/22/15 - 3/21/16): 9
- Post-Policy (10/1/16 - 12/31/17): 15

OPTN ORGAN PROCUREMENT AND TRANSPLANTATION NETWORK
Dilated Cardiomyopathy Guidance

- Pediatric candidates may be eligible for Status 1A exception if:
  - Under 5 kg and:
    - Admitted to hospital of listing
    - Supported by a continuous infusion of at least one high-dose intravenous inotrope
    - OR a continuous infusion of at least two intravenous inotropes
  - Under 10 kg and:
    - Meets conditions above **AND** has evidence of poor systemic perfusion
  - Over 10 kg:
    - Presence of contraindications to mechanical circulatory support
Pediatric candidates may be eligible for Status 1A exception if

- Admitted to hospital of listing and meets *any* of the following criteria:
  - Supported by a continuous infusion of at least one high-dose intravenous inotrope **OR** a continuous infusion of at least two intravenous inotropes
  - Has had an episode of sudden death or recurrent prolonged runs of hemodynamically significant arrhythmia that are not controlled by medical therapy
  - Has had syncopal episodes felt to be related to restricted ventricular filling
  - Has evidence of increased pulmonary vascular resistance (exceeding 6 WU*m2)
Single Ventricle Heart Disease Guidance

- Pediatric candidates with this CHD may be eligible for Status 1A exception if:
  - Admitted to hospital of listing **AND**
  - Experiencing complications related to their congenital heart disease **AND**
  - Is actively receiving therapy for said complication, without regard for change in the candidate’s cardiac support

- Pediatric candidates may be eligible for Status 1B exception if palliated through a Fontan procedure
Coronary Allograft Vasculopathy (CAV) and Retransplantation Guidance

- Pediatric candidate may be eligible for Status 1A exception if:
  - Prior heart transplant and evidence of chronic rejection or significant CAV AND
  - History of recent cardiac arrest, or signs or symptoms placing patients at high-risk for sudden cardiac death

- Pediatric candidates may be eligible for Status 1B exception if
  - Has a history of revascularization (either surgical or transcatheter) for CAV
Standard Information for Pediatric Heart Exception Requests

- Requests specific description of candidate’s current diagnoses and methods of support, inclusive of inotropes and mechanical circulatory support

- Specifically describe how:
  - The candidate meets the exception criteria, or
  - Why standard therapies may not be ideal

- Describe why the current policy does not adequately account for the candidate’s particular situation and high risk of waitlist mortality
Feedback Requested

- Are there other contraindications to the use of a Ventricular Assist Device (VAD) that should be considered?

- What, if any, measure of sensitization could be included to assist in determining whether a Coronary Allograft Vasculopathy candidate should be considered for Status 1A listing by exception?

- What other considerations would make this guidance more useful?

*Please introduce yourself when you speak*
Guidance Addressing the Use of Pediatric Heart Exceptions

- Guidance is provided to determine status exception requests for candidates with the diagnoses of:
  - Dilated cardiomyopathy
  - Hypertrophic or restrictive cardiomyopathy
  - Single ventricle heart disease
  - Coronary allograft vasculopathy and retransplantation

- Outlines standard information to include when submitting pediatric heart exception requests

*Feedback is summarized and shared as public comment*