Management of Pediatric Isolated Bicuspid Aortic Valve: Current Practice Survey

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BACKGROUND

• Incidence of isolated bicuspid aortic valve: 0.5-2%
• Risks
  • Progressive dilation, stenosis & insufficiency
  • Dissection
  • Slower progression & lower risk in childhood
• Follow up:
  • Adult guidelines recently changed (2008→2014)
  • Previously recommended every 2 years
  • Now, a function of severity and progression
  • Yearly if aortic root >4.5 cm
• No pediatric guidelines
• Retrospective study
  • Intervals shorter & more variable for children
  • Than recommended for adults at the time
  • Shorter intervals if diagnosed younger or with
  • Aortic root or ascending aorta dilation; earlier
  • Era of diagnosis, or some AS/AR at follow up

OBJECTIVE

Ascertain current practice in management of isolated bicuspid aortic valve in pediatric patients

DESIGN/METHODS

• March-April 2020
• Members of the American Academy of Pediatrics Section on Cardiology and Cardiovascular Surgery
• Pediheart online community
• Email survey
  • Preferred interval of follow up
  • Five age groups
  • Degrees aortic stenosis, insufficiency, & dilation
  • Indications for intervention
  • Medical management strategies
  • Echocardiographic screening of relatives

RESULTS

• 106 responses with usable data; 97% pediatric cardiology; 17.7 +/- 12 years in practice; from all sizes of practice
• Shorter intervals of follow up for younger patients & those with more severe disease (figure 1)
• Intervention recommended for severe disease (figures 2 & 3)
• Medical management is widespread, with different medications preferred for AI and AD (figure 4).
• Echocardiographic screening usually recommended for first degree relatives (Figure 5).

CONCLUSIONS

This study describes current practices regarding management of isolated BAV in children.

DEFINITIONS

<table>
<thead>
<tr>
<th>Degree of Stenosis</th>
<th>AS</th>
<th>AI</th>
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<tbody>
<tr>
<td><strong>Severe</strong></td>
<td>&gt;4 m/s</td>
<td>LV z-score 4</td>
</tr>
<tr>
<td><strong>Moderate</strong></td>
<td>2-4</td>
<td>2-4</td>
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<tr>
<td><strong>Mild</strong></td>
<td>2-4</td>
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Figure 1: Recommended interval for follow up (months), for varying degrees of A) aortic stenosis (AS), B) aortic insufficiency (AI), and C) aortic dilation (AD) in the setting of isolated BAV.

Figure 2: Percent of clinicians recommending surgical or catheter-based intervention at different ages & variable degrees of aortic stenosis (AS), aortic insufficiency (AI), or aortic dilation (AD) in the setting of isolated BAV.

Figure 3: Aortic pressure gradient treatment triggers.

Figure 4: Medical management of isolated BAV. A) Percent of clinicians who indicated they ever medically managed insufficiency or dilation associated with isolated BAV. B) Medications preferred for treating those two conditions.

Figure 5: Of the 82% of clinicians who advise screening, most focus on first degree relatives.

CONTACTS

Thanks: AAP SOCCS and Pediheart community
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