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

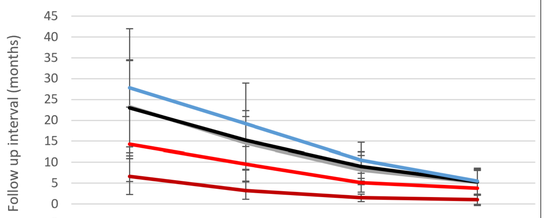
- March-April 2020
- Members of the American Academy of Pediatrics Section on Cardiology and Cardiovascular Surgery
- Pediheart online community
- **Email survey**
  - Preferred **interval of follow up**
    - Five age groups
    - Degrees aortic stenosis, insufficiency, & dilation
  - Indications for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

DEFINITIONS

severe AS	severe AI
>4 m/s	LV z-score 4

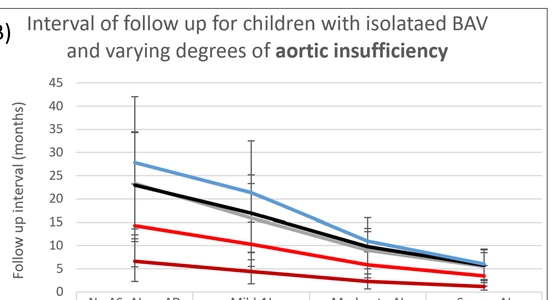
mild AD	moderate AD	severe AD
z-score 2-4	z-score 4-6	z-score >6

A) Interval of follow up for children with isolataed BAV and varying degrees of **aortic stenosis**



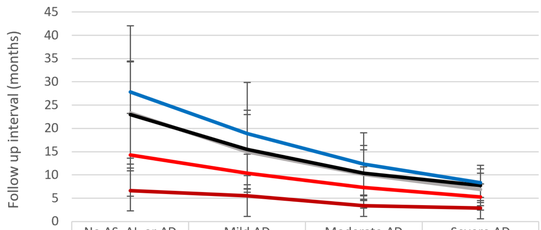
	No AS, AI, or AD	Mild AS	Moderate AS	Severe AS
newborn	6.6	3.2	1.5	1
1 year	14.3	9.6	5.1	3.8
6 years	23.3	14.6	8.1	5.2
12 years	23	15.2	9	5.3
18 years	27.8	19.2	10.5	5.5

B) Interval of follow up for children with isolataed BAV and varying degrees of **aortic insufficiency**



	No AS, AI, or AD	Mild AI	Moderate AI	Severe AI
newborn	6.6	4.4	2.3	1.2
1 year	14.3	10.3	5.9	3.5
6 years	23.3	15.9	9	5.5
12 years	23	16.9	9.8	5.8
18 years	27.8	21.4	11	6

C) Interval of follow up for children with isolataed BAV and varying degrees of **aortic dilation**



	No AS, AI, or AD	Mild AD	Moderate AD	Severe AD
newborn	6.6	5.5	3.4	2.9
1 year	14.3	10.4	7.3	5.3
6 years	23.3	15	10.1	6.9
12 years	23	15.5	10.4	7.7
18 years	27.8	18.9	12.3	8.3

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.

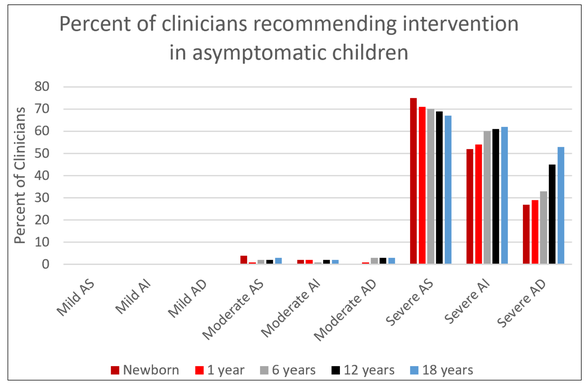


Figure 2: Percent of clinicians recommending surgical or catheter-based **intervention** at different ages & variable degrees of aortic stenosis (AS), aortic insufficiency (AI) , or aortic dilation (AD) in the setting of isolated BAV.

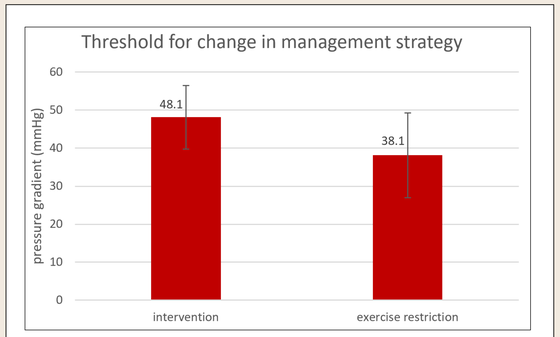
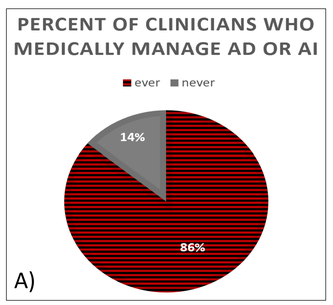


Figure 3: Aortic pressure gradient treatment triggers.



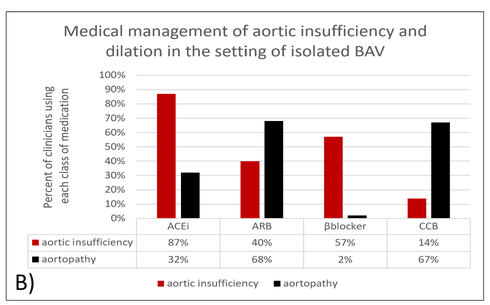


Figure 4. Medical management of isolated BAV. A) Percent of clinicians who indicated they ever medically managed insufficiency or dilation associated with isolated BAV. B) Medications preferred for treating those two conditions.

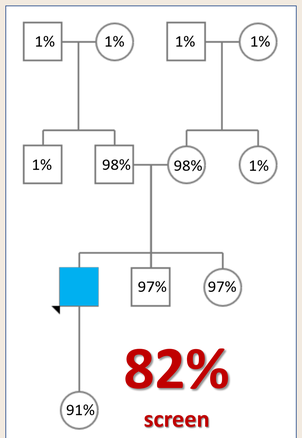


Figure 5. Of the 82% of clinicians who advise screening, most focus on first degree relatives.

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.

ADDITIONAL KEY INFORMATION

Thanks: AAP SOCCS and Pediaheart community
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*Yamauchi et al (2018) Disease progression and variation in clinical practice for isolated bicuspid aortic valve in children. Congenit Heart Dis 13 (3):432-439