Cardiology in the Young

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Variation in management of paediatric isolated bicuspid aortic valve: current practice survey

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Original Article

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Abstract

Background: Prior to the recent release of appropriate use criteria for imaging valvulopathies in children, follow-up of valvular lesions, including isolated bicuspid aortic valve, was not standardised. We describe current follow up, treatment, and intervention strategies for isolated bicuspid aortic valve with varying degrees of stenosis, regurgitation, and dilation in children up to 18 years old and compare them with newly released appropriate use criteria. *Methods:* Online survey was sent to members of the American Academy of Pediatrics Section on Cardiology and Cardiac Surgery and PediHeartNet. *Results:* Totally, 106 responses with interpretable data were received. For asymptomatic patients with isolated BAV without stenosis, regurgitation, or dilation follow-up-intervals increased from 7+/−4 months in the newborn period to 28 +/− 14 months at 18 years of age. Respondents recommended more frequent follow-up for younger patients and those with greater disease severity. More than 80% of respondents treat aortic regurgitation or aortic dilation in the setting of bicuspid aortic valve medically. In general, intervention was recommended once stenosis or regurgitation became severe (stenosis of >4 m/s; regurgitation with LV Z score 4) regardless of age, but was not routinely recommended for younger children (newborn – age 6 years) with severe dilation. Exercise was restricted at 38+/−11 mmHg echocardiographic mean gradient. *Conclusions:* Current follow-up, treatment, and intervention strategies for isolated bicuspid aortic valve deviate from appropriate use criteria. Differences between the two highlight the need to better delineate the disease course, clarify recommendations for care, and encourage wider adoption of guidelines.

Isolated bicuspid aortic valve occurs in 0.5%–2% of the population.[1-3] The risk for progressive aortic stenosis, aortic regurgitation, aortic dilation, and aortic dissection underpins guidelines recommending close follow-up and imaging, medical management strategies, and activity restriction in adults.[4-6] Recent studies have shown a lower risk and slower rate of progression of aortic disease associated with bicuspid aortic valve in children as compared to adults.[7-9] Despite this research, new appropriate use criteria for imaging in children with bicuspid aortic valve recommend shorter intervals of follow-up than those recommended in earlier guidelines for adults.[5,6,10]

There are limited data describing management strategies in children up to age 18 years, and until this year, there were no formal guidelines for management of isolated bicuspid aortic valve in children. To correspond with the release of the first appropriate use criteria for imaging of this valvular heart disease,[10] this study describes current follow-up intervals and management strategies for isolated bicuspid aortic valve. Children up to and including 18 years of age were included, despite the presence of adult guidelines that would apply to the 18-year-old patients, as paediatric cardiologists often follow patients up to their mid-20s and recent appropriate use criteria were written to include 18-year-old patients. A better understanding of current clinical practice will allow for identification of areas of discordance between what is known about the course of this disease in children, follow-up recommendations, and how clinicians currently manage patients. Further exploration of these areas of discordance may guide future revisions to the current guidelines or interventions to encourage standardisation of practice.

Methods

Study design

This cross-sectional, online survey study of paediatric cardiac providers received an exemption from the University of Wisconsin Health Sciences Institutional Review Board and was administered in March and April of 2020. An initial invitation was sent on 3/11/2020, and two reminder emails were sent two and five weeks later.

Study population

Eligible participants were members of either the American Academy of Pediatrics Section on Cardiology and Cardiac Surgery or the PediHeartNet online community with active email addresses on file. Members of these organisations include a spectrum of professionals involved in the care of children with heart disease.

Survey development

The survey contained 17 questions. Responses included numeric input as well as binary, multiple choice, and five-point Likert scale selections. Survey domains included demographic data as well as interval of follow-up; indications for intervention or exercise restriction; use of medication to manage aortic dilation and aortic regurgitation; and echocardiographic screening of relatives of children of various ages with isolated bicuspid aortic valve and various degrees of aortic stenosis, aortic regurgitation, and aortic dilation (Table 1). In all clinical scenarios, the children in question were stated to be asymptomatic. We included patient scenarios in the age groups typically cared for by paediatric cardiologists (newborns and patients at ages 1, 6, 12, and 18 years). Following initial development, the survey was reviewed for content, design, and readability and was beta-tested to establish completion time.

Data management and statistical analysis

This survey was generated on the Qualtrics web-based survey platform (Qualtrics, Provo, UT). Eligible participants were emailed a secure, anonymous link to the study. Anonymised data extracted from Qualtrics was batch processed and analysed using descriptive statistical tools in Excel (Microsoft, Redmond, WA) for both continuous (mean, median, mode, range, and standard deviation) and categorical variables (percents and total counts). The significance of age-related differences in recommendations was evaluated using repeated measures ANOVA in Stata (StataCorp, College Station, TX). A p-vale of 0.05 was a priori defined as significant.

Results

Participants

The survey was sent to a total of 1777 paediatric providers through PediHeartNet and 865 through the American Academy of Pediatrics Section on Cardiology and Cardiac Surgery. Of 239 providers who started the survey, 106 responded to at least some of the study questions. All responses to each individual question were utilised. As there is considerable overlap between membership in the two organisations, and we could not assure that all e-mail addresses were active, we could not calculate a response rate.

Demographic characteristics are shown (Table 2). Respondents were primarily from the United States (97%) with most respondents from the Midwest (30%), South (27%), and Northeast (26%). Most respondents were physicians (92%) practicing in paediatric cardiology (97%) and, excluding trainees (5% of the sample), had been in practice for an average of 17.7 +/− 11.7 years. They hailed from programmes with surgical volumes ranging from zero cases per year (22%) to more than 300 per year (43%).

Interval for follow up

Interval for follow-up of asymptomatic children at five set ages with isolated bicuspid aortic valve and varying degrees of aortic stenosis, aortic regurgitation, and aortic dilation is shown (Fig 1). In general, closer follow-up was recommended for younger children and those with more severe disease.

Table 1. Definitions of severity of aortic disease

Aortic disease	Measurement
Aortic dilation	Aortic Z-score
Mild	2–4
Moderate	4–6
Severe	>6
Aortic stenosis	Peak velocity
Severe	> 4 m/s
Aortic regurgitation	Left ventricular z-score
Severe	4

Table 2. Participant characteristics

Characteristic	N = 106
Primary Field, n (%)	
Pediatric Cardiology	103 (97)
Cardiac Surgery	2 (2)
Not specified	1 (1)
Type of provider, n (%)	
Physician	97 (92)
Trainee	5 (5)
Advanced Practice Provider	4 (4)
Number of years in practice, $\bar{x} \pm sd$	18 ± 12
Surgical volume per year at home institution, n (%)	
Zero	23 (22)
1–50	2 (2)
51–100	11 (10)
101–200	15 (14)
201–300	9 (8)
>300	46 (43)
Geographic region, n (%)	
United States of America	97 (92)
Midwest	29 (30)
West Coast	10 (10)
Central Plains/Rocky Mountains	4 (4)
Northeast	25 (26)
South	26 (27)
Southwest	0 (0)
Canada	3 (3)
Europe	2 (2)
Middle East	1 (1)
South America	1 (1)
No response	2 (2)

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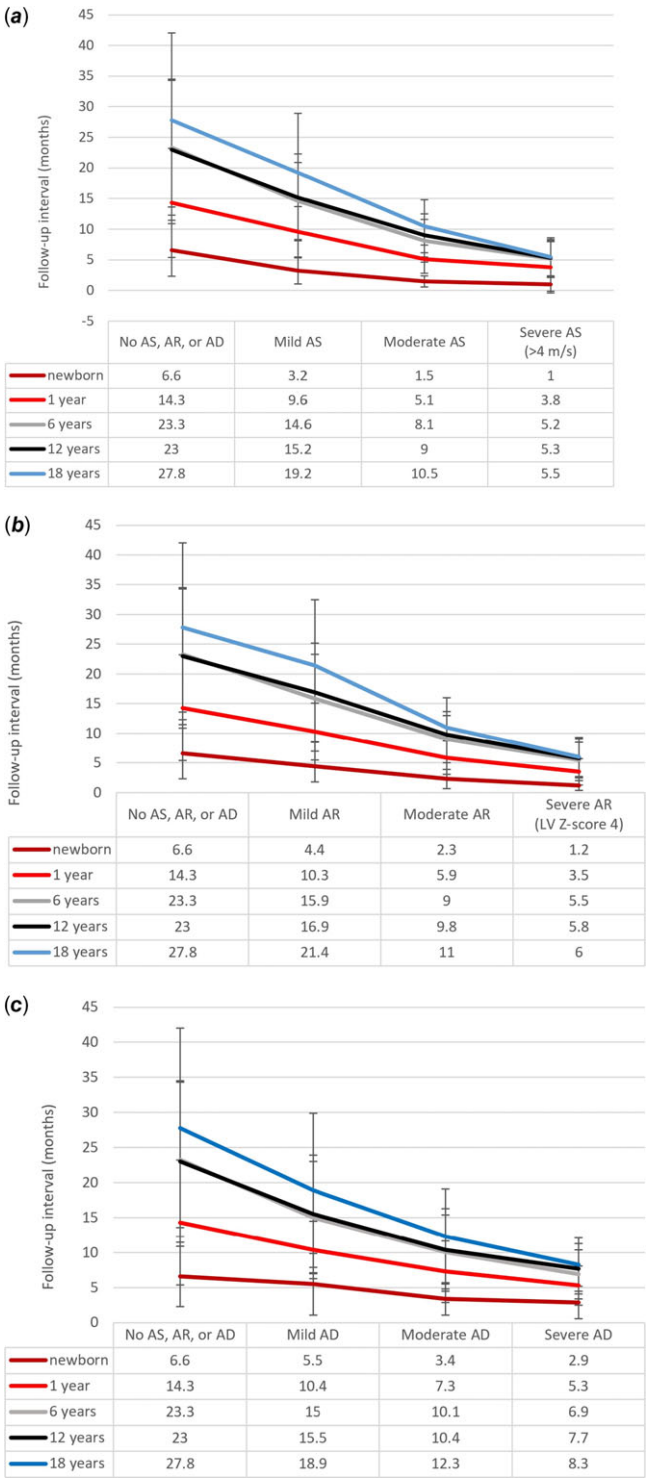

	No AS, AR, or AD	Mild AS	Moderate AS	Severe AS (>4 m/s)
newborn	6.6	3.2	1.5	1
1 year	14.3	9.6	5.1	3.8
6 years	23.3	14.6	8.1	5.2
12 years	23	15.2	9	5.3
18 years	27.8	19.2	10.5	5.5

	No AS, AR, or AD	Mild AR	Moderate AR	Severe AR (LV Z-score 4)
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

	No AS, AR, 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. Follow-up intervals. Recommended interval of follow-up in months for patients with isolated BAV and varying degrees of AS (panel A), AR (panel B) and AD (panel C). AD = aortic dilation, AR = aortic regurgitation, AS = aortic stenosis.

For asymptomatic younger children (newborn or 1 year) with absent or mild aortic stenosis, aortic regurgitation, or aortic dilation, follow-up was recommended between 3 and 14 months on average, while for those patients with moderate valvular disease, follow-up was recommended between 1 and 7 months.

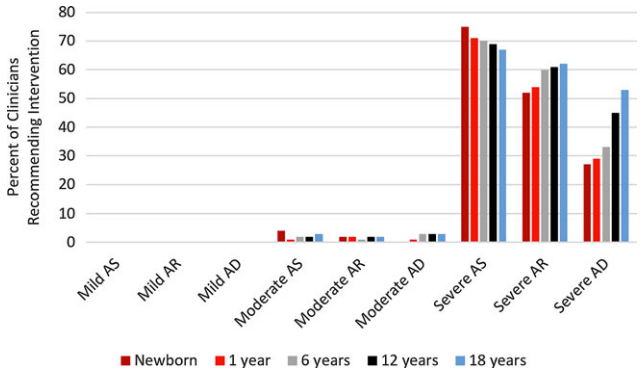

Figure 2. Indications for Intervention. Percent of clinicians recommending surgical or catheter-based intervention at different ages in patients with isolated BAV and varying degrees of AS, AR, and AD. AD = aortic dilation, AR = aortic regurgitation, AS = aortic stenosis.

For asymptomatic older children (ages 6, 12, or 18 years) with absent or mild aortic stenosis, aortic regurgitation, or aortic dilation, follow up was recommended every 1–3 years on average. For asymptomatic older children with moderate aortic stenosis, aortic regurgitation, or aortic dilation, follow up was recommended every 8–10 months.

While the majority (50%–75%) of providers indicated they would recommend intervention for asymptomatic patients with severe aortic stenosis or aortic regurgitation regardless of their age (Fig 2), those providers who chose not to intervene chose an average follow-up interval of 5–6 months for patients ages 6 years and up (Fig 1). A smaller percentage of providers recommended intervention for asymptomatic patients with severe aortic dilation (25%–55% depending on the age of the patient, Fig 2). Instead, providers opted for close follow-up, ranging from 3 months for newborns to 8 months for an 18-year-old patient (Fig 1).

Indications for intervention

Many providers do not recommend intervention for aortic stenosis or aortic regurgitation based on qualitative descriptions of valvular dysfunction in the absence of symptoms (Fig 2). No clinicians recommended intervention for asymptomatic patients with no or mild aortic stenosis, aortic regurgitation, or aortic dilation. Very few (<10%) recommended intervention for moderate aortic stenosis, aortic regurgitation, or aortic dilation. Many recommended intervention for severe aortic regurgitation (50%–60%) or aortic stenosis (65%–75%), with a greater percentage of providers recommending intervention at younger ages in aortic stenosis and a greater percentage recommending intervention at older ages in aortic regurgitation. While a slight majority of providers recommended intervention for 18-year-old patients with severe aortic dilation (55%), only a minority of providers (25%-45% depending on age) recommended intervention for younger patients with severe aortic dilation in the absence of symptoms.

The vast majority of providers recommended intervention for patients with aortic stenosis at set pressure gradients regardless of age or symptomatology (Table 3). Providers recommended intervention for any age patient at an average pressure gradient of 48+/−8 mmHg. Repeated measures ANOVA was used to explore the relationship between patient age and clinician recommendations. The same percentage of clinicians recommended intervention at some threshold even in asymptomatic patients regardless of the

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Table 3. Intervention or exercise restrictions in asymptomatic patients with aortic stenosis

Clinicians	Only with symptoms n (%)	Intervene at threshold n (%)	Echo mean gradient (mmHg) $\bar{x} \pm sd$
Intervention			
Newborn	8 (9)	86 (91)	46 ± 8
1 year	6 (6)	88 (94)	48 ± 9
6 years	8 (9)	86 (91)	49 ± 9
12 years	7 (7)	87 (93)	49 ± 8
18 years	6 (6)	88 (94)	49 ± 8
Exercise restrictions			
6 years	18 (20)	75 (80)	39 ± 10
12 years	7 (7)	87 (93)	38 ± 11
18 years	6 (6)	88 (94)	38 ± 12

Table 4. Medication recommended for management of aortic regurgitation and aortic dilation

Clinicians recommending medication	N = 104
Aortic Regurgitation, n (%)	
Never	14 (13)
Rarely	54 (52)
Occasionally	25 (24)
Frequently	11 (11)
Always	0 (0)
Aortic Dilation, n (%)	
Never	14 (13)
Rarely	38 (37)
Occasionally	32 (31)
Frequently	18 (17)
Always	2 (2)

patient's age ($p = 0.810$); however, the threshold chosen for intervention was lower in younger patients ($p < 0.001$).

Indications for activity restriction

Providers recommended exercise/activity restrictions for asymptomatic 6-, 12- and 18-year-old patients at an average pressure gradient of 38+/−11 mmHg. Repeated measures ANOVA was used to explore the relationship between patient age and clinician recommendations. Both the recommendation for activity restriction in an asymptomatic patient ($p < 0.001$) and the threshold at which that recommendation would be made to restrict activity ($p = 0.004$) varied by age, with more clinicians recommending restriction at a lower threshold in older patients.

Use of medication

In total, 87% of providers report having used medications to manage aortic regurgitation or dilation (Table 4). Angiotensin-converting enzyme inhibitors and angiotensin receptor blockers were the preferred medications for management of aortic regurgitation, while angiotensin receptor blockers and beta blockers were the preferred medications for management of aortic dilation (Fig 3).

Echocardiographic screening of relatives

In total, 82% of providers recommended echocardiographic screening of an affected patient's first-degree relatives.

Discussion

While previous guidelines for follow-up, imaging, and intervention of isolated bicuspid aortic valve targeted adult patients, this year, appropriate use criteria for imaging follow-up of children were released (Table 5).[4-6,10,11] Prior to the release of these guidelines, clinicians relied on individual clinical expertise and extrapolation of recommendations from adult guidelines to guide their care of patients with this lesion. While we now have a consensus statement on the use of imaging for follow-up of this lesion; paediatric-specific exercise,[12,13] medical management, and interventional recommendations are still lacking.

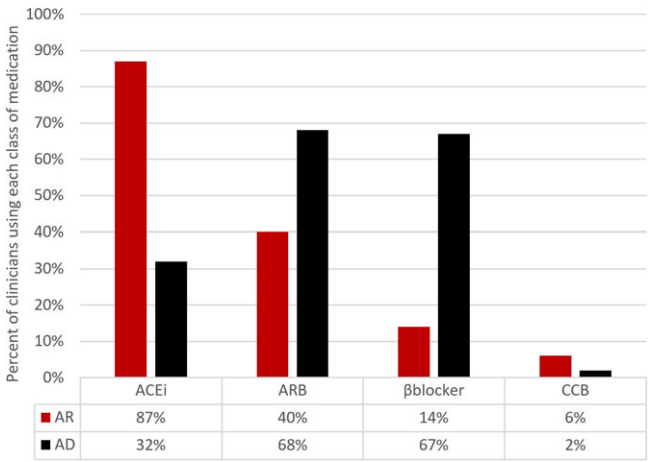

Figure 3. Medications used in BAV. Percent of those clinicians who ever use medications for the treatment of AR and AD who recommend each of four common antihypertensives: ACEi, ARB, beta blockers, CCB. ACEi = angiotensin converting enzyme inhibitor, AD = aortic dilation, AR = aortic regurgitation, ARB = angiotensin receptor blocker, AS = aortic stenosis, CCB = calcium channel blocker.

Follow-up intervals

In general, clinicians recommended a shorter interval of follow-up for the oldest children (age 18) with isolated bicuspid aortic valve than recommended by current clinical practice guidelines for adults regardless of the degree of valvulopathy (Figs 1 and 4, Table 5).[4]

Follow-up intervals recommended for asymptomatic children ages 6–12 years with isolated bicuspid aortic valve and mild or moderate aortic stenosis or aortic regurgitation were comparable to those recommended in the newest appropriate use criteria for children. Despite research showing a low risk and slow rate of progression aortic valve dysfunction in children compared to adults,[7,9] the intervals recommended in the newest appropriate use criteria and those reported by clinicians were shorter than those recommended for adults (Fig 4).[6,10]

By contrast, clinicians followed up newborns and infants with isolated bicuspid aortic valve and mild or moderate aortic stenosis

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Table 5. Timeline of recommended* frequency of imaging for management of aortic lesions

Lesion & severity	2006 CPG[11]	2014 CPG[4]	2017 AUC[6]	2020 AUC-pediatric[10]		
				<3 m	3–12 m	1–18 y
Aortic Stenosis						
Mild	3–5 y	3–5 y		1–4 w	3–6 m	1–2 y
Moderate	1–2 y	1–2 y	1–2 y	1–4 w	1–3 m	6–12 m
Severe	12 m	6–12 m	12 m		Intervention indicated	
Aortic Regurgitation						
Mild		3–5 y	3–5 y	1–4 w	3–6 m	1–2 y
Moderate		1–2 y	1–2 y	1–4 w	1–3 m	6–12 m
Severe	3–6 m	6–12 m			intervention indicated	
Aortic Dilation						
Surveillance (BAV)			3–5 y		3–5 y if no dilation	
Mild		>12 m†			2–3 y if stable z-scores	
Moderate		>12 m‡			6–12 m if incr. z-scores	
Severe	12 m§	12 m**	<12 mo††		intervention indicated	

*Clinical practice guidelines (CPG) and appropriate use criteria (AUC); m = months; y = years
†If dilation <4.5 cm, stable dilation, no family history of aortic dissection
‡i.b.i.d
§If dilation >4 cm
**If >4.5 cm, rapid rate of change in diameter, or family history of aortic dissection
††If >4 cm and one or more of the following: >4.5 cm, rapid rate of change in diameter, first-degree relative with aortic dissection

or aortic regurgitation at intervals that were longer than those recommended in the new paediatric appropriate use criteria (Fig 4).[10] For newborns with mild valvulopathy, follow-up was reported at 3–4 months (compared to the recommended 1–4 weeks), while infants with the same severity of disease were followed up at 10 months (compared to the recommended 3–6 months). For newborns with moderate valvulopathy, follow-up was reported at 6–8 weeks (compared to the recommended 1–4 weeks), while infants with the same severity of disease were followed up at 5–6 months (compared to the recommended 1–3 months). Formulating recommendations for follow-up of these young children can be challenging as there are few patients and a paucity of data at this age range; however, the longer-than-recommended follow-up intervals reported by clinicians suggest that clinical experience has shown that the overall progression of disease in infants is not as rapid as implied by the new appropriate use criteria.

For patients with isolated bicuspid aortic valve without aortic stenosis, aortic regurgitation, or aortic dilation, surveillance imaging was recommended at significantly shorter intervals (6 months for newborns, 14 months for infants, 2 years for children) than recommended by current paediatric or adult appropriate use criterion (3–5 years, Table 5).[6,10]

Comparison of survey data regarding aortic dilation to the new paediatric appropriate use criteria is challenging as the appropriate use criteria used different classifications than were used in this study. As with aortic stenosis and aortic regurgitation, paediatric providers followed up 18-year-olds with varying degrees of aortic dilation at approximately the same intervals as those recommended for adults, with shorter intervals recommended for younger children and those with more severe disease (Fig 1, Table 5).

Indications for intervention

Current appropriate use criteria do not offer recommendations for imaging follow-up of patients with isolated bicuspid aortic valve and severe aortic stenosis, aortic regurgitation, or aortic dilation as "it was assumed that these patients will undergo surgical or catheter-based interventions".[10] This aggressive approach toward intervention does not appear to correlate with contemporary practice as our data show that between 25% (newborn with severe aortic stenosis) and 70% (newborn with severe aortic dilation) of providers would not recommend intervention in an asymptomatic child with severe aortic stenosis, aortic regurgitation, or aortic dilation.

Indications for activity restriction

While guidelines for the participation of individuals with structural heart disease in competitive athletics exist for children ages 12 years and older, there are no guidelines addressing recreational activities or athletic participation for younger patients.[14] Clinicians in this study made recommendations for younger patients with isolated bicuspid aortic valve based on pressure gradients across the valve.

Use of medication

Clinicians were split on whether or not to medically manage aortic regurgitation and aortic dilation (Table 4). While there may be a theoretical benefit to medical therapy in aortic regurgitation and aortic dilation, there are no formal recommendations regarding medical management of either of these entities for adults or children in the absence of hypertension. The most recent clinical practice guidelines for adults suggest that calcium channel blockers,

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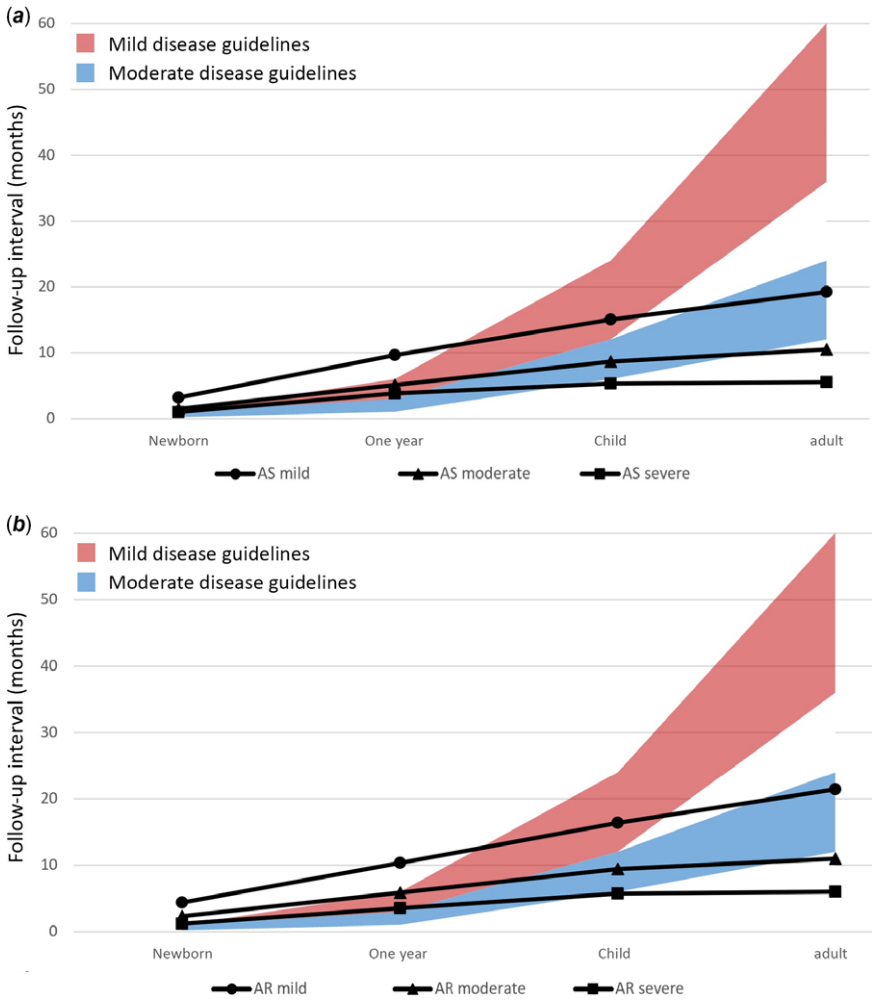

Figure 4. Clinician follow-up vs. AUC. Comparison of clinician recommendations with AUC recommendations for follow-up interval of patients with isolated BAV with AS (panel A) and AR (panel B). Survey data shown as a means for each age and degree of valvular dysfunction. AUC guidelines shown as shaded areas representing the range of intervals over which follow-up is recommended at each age for patients with mild (red) and moderate (blue) valvular dysfunction. AR = aortic regurgitation, AS = aortic stenosis, AUC = appropriate use criteria.

angiotensin-converting enzyme inhibitors, and angiotensin receptor blockers may have an advantage over beta blockers in the treatment of hypertension in the setting of aortic regurgitaiton; however, the theoretical advantage of beta blockers and angiotensin receptor blockers in the setting of aortic dilation has not been demonstrated clinical studies.[4] Medications preferred by clinicians in this study corresponded to these theoretical advantages (Fig 3).

Echocardiographic screening of relatives

In total, 82% of respondents recommended echocardiographic screening of first-degree relatives of patients with isolated bicuspid aortic valve. Despite the fact that 20%–30% of affected patients will have at least one family member with bicuspid aortic valve, there is not yet data on the cost-effectiveness of such screening or its impact on patient outcomes.[4]

Study limitations

Given likely crossover of membership in the American Academy of Pediatrics Section on Cardiology and Cardiac Surgery and PediHeartNet, it is impossible to calculate a response rate. While the survey was sent to a minimum of 1777 individuals through the American Academy of Pediatrics Section on Cardiology and Cardiac Surgery, not all individuals would have been eligible to complete the study as this organisation accepts medical students and residents as members. In addition, this survey was sent out during March and April of 2020, as communities and organisations around the United States were beginning to mount dramatic COVID-19 pandemic responses and email inboxes were being flooded with daily missives about new developments. While the 106 usable responses were representative of providers from a wide swath of the country practicing at a wide range of practices, it is impossible to determine if there are any characteristics that differentiate those providers who responded to this survey from those who did not. Although unlikely, as the survey was conducted using an anonymous link to the survey, we cannot exclude the possibility that the survey was completed by individuals outside the American Academy of Pediatrics Section on Cardiology and Cardiac Surgery and PediHeartNet or that respondents could have accessed the survey multiple times. Finally, our study was limited to isolated aortic valve disease and aortopathy and did not address mixed disease.

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Conclusions

While children have been shown to have generally slower progression of aortic valve dysfunction than do adults in the setting of isolated bicuspid aortic valve, most respondents evaluate children at shorter intervals than those recommended for adults. New appropriate use criteria for children recommend follow-up of the youngest patients (newborns and infants) at shorter intervals than those reported by the clinicians in our study, suggesting that disease progression in these young patients may be slower than implied by the appropriate use criteria. Additionally, current appropriate use criteria for imaging do not provide guidance on follow-up of patients with severe valve dysfunction, despite the reality that many providers report continued observation in the absence of symptoms. Further exploration of these areas of discordance may guide future revisions to the current guidelines or interventions to encourage standardisation of practice. Other areas of exploration include the relative efficacy of various medications in the treatment of aortic regurgitation and dilation in the setting of this disease, the risks and benefits of current patterns of activity restriction, and the cost-effectiveness and clinical utility of echocardiographic screening of relatives. As we learn more, we can adjust our practice and recommendations to better align with our ever-evolving knowledge of this clinical entity.

Supplementary material. To view supplementary material for this article, please visit https://doi.org/10.1017/S1047951121001499

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Conflicts of interest. None.

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