Cardiology in the Young

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Variation in the management and treatment of children with giant coronary artery aneurysm following Kawasaki disease

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Abstract

Objectives: Giant coronary artery aneurysms are rare but potentially fatal complications of Kawasaki disease. The lack of evidence-based recommendations on their management and treatment cause guidelines and practices to differ. We aimed to assess these variations. *Methods:* An anonymous online survey regarding surveillance, imaging, pharmacological management, and interventional practices was distributed among 134 physicians attending to Kawasaki disease patients worldwide. A p-value of <0.05 was deemed significant. *Results:* The majority (60%) of respondents were general paediatric cardiologists, and 29% interventional specialists. The average years in practice was 15 ± 9.6. Physicians from Asia had the most experience with giant coronary artery aneurysms. American practitioners preferred combining anticoagulants with aspirin. Beta-blockers and statins were more likely used in teenagers versus younger children. Cardiac catheterisation was most (52%) chosen for coronary surveillance in patients with echocardiogram anomalies, followed by Coronary CT-angiography. The indications for coronary intervention were split among respondents, regardless of geographic region or experience. The preferred treatment of coronary stenosis was percutaneous intervention (69%) versus bypass surgery. For thrombosis, thrombolytics (50%) were preferred over percutaneous (39%) and surgical (11%) interventions. Most (92%) preferred intervening in young children in a paediatric facility but were split between a paediatric and adult facility for older children. Most chose combined management by adult and paediatric specialists for either age-scenarios (70, 82%). *Conclusion:* As identified by our study, the lack of large studies and evidence-based recommendations cause uncertainty and ambivalence towards certain treatments. International collaborative efforts are needed to provide more robust evidence in the management of these patients.

Background

Coronary artery aneurysm is a serious complication of Kawasaki disease. Patients with coronary artery aneurysm are at risk of thrombosis and stenosis, which can lead to ischaemic heart disease, myocardial infarction, and sudden cardiac death.[1,2] The risk is higher in patients with giant coronary artery aneurysms, with up to 48% suffering myocardial infarction or death on long-term follow-up.[3] A single-centre retrospective study from Japan showed cumulative rates of coronary artery intervention of 28, 43, and 59% at 5, 15, and 25 years in patients with giant coronary artery aneurysms (Z-Score≥10 or absolute dimension ≥8 mm).[4] An international multicentre collaboration further consolidated knowledge on Z-score-based giant coronary artery aneurysms, identifying an incremental risk as the Z-score increases above the value of 10.[5]

Due to the risk for coronary artery stenosis or thrombosis, careful surveillance is a key component in the management of Kawasaki disease patients with giant coronary artery aneurysms. However, there continue to be variations among different guidelines in the timing, frequency and mode of surveillance.[6] The 2017 American Heart Association statement recommends greater frequency of cardiac surveillance in those with giant coronary artery aneurysms.[1] In contrast, the Japanese Circulation Society (JCS/JSCS) guidelines indicate electrocardiography, echocardiography, and assessment for inducible ischaemia at a lower frequency of 1 to 5 years.[5] The guidelines set by a United Kingdom lead group also differ slightly by recommending the consideration of annual stress imaging for inducible myocardial ischaemia.[6] Advanced coronary imaging modalities, such as CT, MRI, or other invasive tests, are similarly recommended in both JCS/JSCS and American Heart Association guidelines [1] which is

less strict in frequency compared to the United Kingdom management of giant coronary artery aneurysms.[6]

Thromboprophylaxis is an important component to minimise the risk of coronary artery thrombosis in Kawasaki disease patients with giant coronary artery aneurysms.[1] The indication for long-term aspirin (Amino Salicylic Acic (ASA)) is well-established amongst expert recommendations.[1,5,6] In patients with giant coronary artery aneurysms, anticoagulation with warfarin or low-molecular-weight heparin is "reasonably indicated" according to the American Heart Association.[1] It is similarly deemed "likely to be useful and effective" in the JCS.[5] The United Kingdom expert group states that long-term warfarin was associated with better outcomes in patients with giant coronary artery aneurysms, but recognises the lack of randomised controlled trials.[6] The use of dual antiplatelet therapy (ASA and clopidogrel) remains a class IIa (likely to be useful and effective) according to the JCS, but only "may be considered in addition to anticoagulation" (class IIb) in the American Heart Association.[1,5]

Notwithstanding the fact that atherosclerosis is not caused by Kawasaki disease, empirical use of statin has been advocated in the long-term management of Kawasaki disease.[6] Its use in atherosclerotic disease in adults is based on reducing chronic inflammation and lowering low-density lipoprotein cholesterol.[1] Similarly, in small short-term studies involving Kawasaki disease patients with aneurysms, statin treatment showed reductions in high-sensitivity C-reactive protein and improved endothelial function.[7] A safety study in children with acute Kawasaki disease and coronary artery (CA) involvement, age 2 years or older, has not reported any significant side effects.[7] Statin empirical long-term therapy can thus be considered in patients with giant coronary artery aneurysms, but clinical trials on acute Kawasaki disease are yet to be performed.[1]

Experience with coronary artery intervention after Kawasaki disease in children is limited. Decisions regarding the need for revascularisation are often difficult and based on experience in adults with atherosclerotic heart disease, rather than evidence-based data in children or adults with Kawasaki disease.[6,8] Although surgical and percutaneous interventions have the same goal of relieving ischaemic symptoms and recovering myocardial viability, the underlying pathophysiology behind Kawasaki disease and atherosclerotic heart disease remains different. Based on adult guidelines, mechanical revascularisation should be considered in patients with >50% stenosis of the left main coronary artery (or equivalent) in patients with symptoms and documented ischaemia, a multi-vessel coronary disease with a reduction in left ventricular function, or the presence of high-risk findings on non-invasive ischaemia testing, notably early inducible myocardial ischaemia, exercise-induced arrhythmias, or poor exercise tolerance ($\leq$ 3 MET) due to symptoms such as angina or dyspnoea.[2] Moreover, patients with symptoms refractory to maximal medical management and those with silent ischaemia involving >10% of the myocardium could benefit from revascularisation. Prior recommendations also included stenosis >70% on angiography, but recent studies have shown no benefit for revascularisation based on lesion severity alone.[9] Current practice in adults is based on fractional flow reserve testing, as patients with a ratio <0.8 benefit from intervention.[10]

Experience with coronary artery bypass grafting after Kawasaki disease has mostly been described in Japan. Long-term follow-up of these patients showed a 25-year survival rate of 95% (95% confidence interval 88–98) in 114 patients, with a 60% rate of cardiac event freedom (95% confidence interval 46–72).[5] The most frequent cardiac events were surgical re-interventions and other interventional procedures, in which the prevalence remained generally low: 3.47 and 3.97%, respectively.[11] The early Japanese series reported lower patency rates on long-term follow-up of 93, 73, and 65% at 1, 5, and 15 years in patients younger than 12 years of age compared to 95, 91, and 91% in older children.[5] More recently, a systematic review and meta-analysis of surgical myocardial revascularisation outcomes in 1,191 patients from 32 studies showed that coronary artery bypass grafting is a safe procedure, with an early mortality rate of only 0.28% (95% confidence interval: 0.00–0.73%).[11] Small series have reported the use of percutaneous coronary interventions in patients with Kawasaki disease.[12–15] Limited experience favours percutaneous coronary intervention in patients with single-vessel disease, a multi-vessel disease with focal, easily treatable lesions, and normal left ventricular function. Percutaneous approach can also be favoured in patients with significant comorbidities although still challenging due to the heavily fibrotic and calcific lesions.[15] No randomised or prospective trials have compared the outcomes of these approaches.

Overall, most of the available literature on coronary revascularisation after Kawasaki disease is based on small single-centre series. A multicentre data registry of patients with coronary artery aneurysms after Kawasaki disease could provide useful information in this challenging patient population, including a better definition for indications of revascularisation and the choice of procedure. In the perspective of better determining the objectives of future registries, we designed the present study to describe physicians' current practices in the management of patients with giant coronary artery aneurysms following Kawasaki disease, including surveillance practices, medical management, indications for intervention, and the type of coronary intervention.

Method

Study design

This cross-sectional study was conducted through the collection of data via a standardised online survey. The survey questions regarding the management and treatment of patients with giant coronary artery aneurysms after Kawasaki disease were written and validated by the principal investigators, who are experienced in Kawasaki disease patient care and questionnaire design. The survey includes vignettes distinguishing between age ranges or symptomatic versus asymptomatic patients (Appendix). The survey included multiple choice questions, as well as ranking questions. Respondents also had the option to provide comments on personal experience with Kawasaki disease patients. To increase outreach and accessibility, a Spanish version was conceived by a fellow paediatric cardiologist fluent in the language. The survey was hosted by the online platform Microsoft Office Forms® and then sent to identified physicians via e-mail. Survey responses were completely anonymous.

Population and eligibility

Physicians involved in the care of Kawasaki disease patients with coronary artery aneurysm were identified using cardiology associations' distribution lists (American College of Cardiology, American Academy of Pediatrics, American Heart Association, Canadian Pediatric Cardiology Association, International Kawasaki Disease Registry, Pediatric Heart Network, National Quality Collaborative, International Kawasaki Disease Genetics

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Table 1. Respondents' characteristics

	Overall ($n = 134$)	North America ($n = 46$)	South America ($n = 50$)	Asia ($n = 20$)	Europe ($n = 11$)	Middle-East-North-Africa ($n = 7$)	*P*-value
Specialty training							<0.001
General pediatric cardiologist	80 (60)	38 (83)	24 (48)	12 (60)	4 (36)	2 (29)	
General adult cardiologist	1 (0.7)	0	0	0	1 (9)	0	
Interventional paediatric cardiologist	39 (29)	5 (11)	24 (48)	6 (30)	2 (18)	2 (29)	
Other	14 (10)	3 (6)	2 (4)	2 (10)	4 (36)	3 (42)	
Years in practice	15 ± 9.6	18 ± 10.4	12 ± 8.8	17 ± 9.8	11 ± 6.5	18 ± 4.2	0.023
Number of KD patients per year							<0.001
Less than 10	67 (50)	25 (54)	31 (62)	2 (10)	5 (45)	4 (57)	
10 to 30	45 (34)	14 (30)	14 (28)	8 (40)	6 (55)	3 (43)	
More than 30	22 (16)	7 (15)	5 (10)	10 (50)	0	0	
Practice setting							0.002
Group practice	16 (12)	5 (11)	10 (20)	0	0	1 (14)	
Hospital affiliated with university	100 (75)	41 (89)	28 (56)	15 (75)	10 (91)	6 (86)	
Hospital not affiliated with university	18 (13)	0	12 (24)	5 (25)	1 (9)	0	

Consortium, Cardiac Society of ANZ), the Kawasaki disease Arab initiative (Kawarabi) and the Pan Arab Congenital Heart Disease Association, and personal contacts and corresponding authors of publications on Kawasaki disease. Inclusion criteria required that the participant be involved in the cardiac care and management of Kawasaki disease. Physicians who could not read and understand written English or Spanish were excluded from the study.

Data analysis

The results of the survey were described using frequencies and percentages. The Chi-square test was used for the comparison of practices between providers' different characteristics. The expected value of the null hypothesis (no variation in practice) was calculated according to the participants' region/continent of practice, years of practice, and patient presentation. A two-tailed p-value of < 0.05 was deemed significant.

Results

Participant characteristics

A total of 134 physicians completed the survey, with representation from North America (46, 35%), South America (50, 37%), Asia (20, 15%), Europe (11, 8%), and the Middle East and North Africa regions (7, 5%). The majority (60%) of respondents were practicing general paediatric cardiologists, whereas 29% were interventional paediatric cardiologists. A minority of 14 respondents (10%) were not specialised in cardiology but identified as Kawasaki disease patient caregivers. Finally, one participant was an adult cardiologist. Physicians' time in practice averaged 11.4 years overall (standard deviation = 9.6). Physicians from North America had a mean length of practice of 18 (stansard deviation = 10.4) years, whereas the physicians from the rest of the world averaged to 13.4 (standard deviation = 8.8) years of practice. Majority of the respondents (67, 50%) saw less than ten Kawasaki disease patients per year. A third (45, 33.6%) of respondents saw between 10 and 30 patients per year. Finally, a minority (22, 16.4%) saw 30 Kawasaki disease patients or more per year, among whom 27% were physicians from the US. Finally, most (100, 74.5%) respondents practiced in an academic hospital setting (Table 1).

Surveillance and imaging practices

Beyond the first year from diagnosis, 71 (53%) physicians reported that following up on their patients with giant coronary artery aneurysms every 6 months was an appropriate interval of time, 48 (35%) reported follow-up every 3 months, 12 (9%) every 12 months or longer, and 3 (2.2%) every 9 months. Physicians from North America were more likely to report follow-up every 6 months versus other regions (80% vs 39%, $p < 0.001$) (Figure 1). The imaging modality of choice in an asymptomatic Kawasaki disease patient with no abnormalities on electrocardiography nor transthoracic echocardiogram (besides the presence of coronary aneurysms), was coronary computerized tomography angiogrpahy (CTA). Stress testing for inducible ischaemia (stress cardiac MRI, stress echo, positron emission tomography/single photon emission computed tomography (PET/SPECT) nuclear stress test) was not significantly used more often in older patients (42, 31.3% for the 12 year old vs 19, 14.2% for the 2 year old), ($p = 0.28$). Most physicians (83, 61.9%) do not use pharmacological stress testing. Pharmacologic testing was reserved for young patients or those unable to exercise, particularly in North America (20, 51%). Respondents from the rest of the world significantly preferred to avoid pharmacological stress testing (61, 45.5% vs 22, 16.4% in North America) ($p < 0.01$). Physicians who cared for the least Kawasaki disease patients (less than 10 per year) significantly chose to avoid pharmacological stress testing as well ($p < 0.01$).

The use of cardiac catheterisation as the first-line imaging modality stayed relatively stable regardless of patient age (26,

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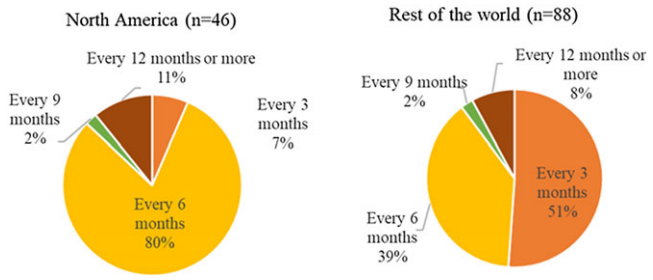

Figure 1. Frequency of cardiac surveillance according to region of practice.

19.4% in the 2 year old vs 22, 16.4% in the 12 year old). However, the use of cardiac catheterisation varied depending on symptoms and transthoracic echocardiogram results. In a patient with isolated chest pain and no abnormalities on echocardiogram, 23 (17.2%) physicians chose to use cardiac catheterisation as their investigation of choice. Its use doubled to 52 (38.8%) with depressed ventricular function on echocardiogram. In general, physicians in North America tend to use cardiac catheterisation less frequently for surveillance compared to other parts of the world regardless of age, clinical presentation, or echocardiographic findings (Figure 2).

Pharmacological management

Antiplatelet and anticoagulation

Aspirin was the most popular choice of medication (ranging from 56.7 to 76.9%) in the antiplatelet management of Kawasaki disease patients with a history of giant coronary artery aneurysms. It was often used in combination with other anticoagulants (warfarin, low molecular weight heparin, or direct oral anticoagulant (48.5–60.4%) regardless of the patient's age (2 year old vs. 12 year old) or presence of coronary artery stenosis on angiography. The combined use of aspirin and anticoagulation was significantly more prevalent in North America compared to other regions, regardless of patient age or state of stenosis (Figure 3). Dual antiplatelet therapy was less used than combined aspirin and anticoagulant therapy, with a range of 11.9–32.8% of respondents choosing the former. This regimen was most chosen in the case of 12-year-old patients with documented stenosis (44, 32.8%).

Beta-blockers

Beta-blockers were the third most chosen medication by respondents. Its use was more frequent in older patients and those with coronary stenosis. Fewer (19, 14.2%) respondents chose to use beta-blockers in a 2-year-old patient with no stenosis compared to 25 (18.7%) in a 12 year old. The presence of stenosis increased its use, with 45 (33.6%) respondents choosing beta-blockers in a 2-year-old patient, increasing once again to 70 (52.5%) respondents in a 12 year old with documented coronary artery stenosis (Figure 4).

Statins

The use of statins was significantly higher in an older patient with documented stenosis. Very few physicians, 8 (6%), recommended statins in a 2-year-old patient with no stenosis, compared to 44 (32.8%) in a 12 year old. Although the acceptability of the use of statins was higher in case of stenosis, its use remained lower in a 2 year old (16, 11.9%) compared to its use in an older patient (71, 53%) (Figure 4). The use of statins did not vary significantly among regions of practice ($p = 0.831$).

Interventional management

When assessing the need of intervention in patients at risk of acute coronary events, most physicians (101, 75.4%) reported basing their decisions on a combination of symptoms, cardiac enzyme elevation, and electrocardiography changes rather than one element or another. The responses were divided almost equally between myocardial perfusion or viability studies (36, 26.9%), coronary artery imaging (32, 23.9%), and ischaemic changes on exercise electrocardiography (24, 17.9%), with no significant difference between regions ($p = 0.961$) (Figure 5).

In the case of coronary artery stenosis, physicians preferred percutaneous coronary intervention (PCI) to coronary artery bypass grafting (93, 69.4% vs 41, 30.6%). There were no significant regional differences ($p = 0.778$). In the case of coronary artery thrombosis, the most popular method of intervention was also percutaneously (53, 39.5%), followed by intra-coronary thrombolysis (41, 30.6%), systemic thrombolysis (25, 18.7%), and coronary artery bypass grafting (15, 11.2%). There was no statistically significant regional difference ($p = 0.53$) in the management of coronary artery stenosis/thrombosis or in the choice of intervention based on institutional experience in coronary artery intervention.

When prompted about the appropriate setting and team for intervention, most physicians (123, 91.8%) recommended a PCI in a paediatric cardiology centre in younger patients (6 year old) while responses were equally split (68, 50.7%) between paediatric and an adult cardiology centre in older patients (16 year old). A combined team approach (of paediatric and adult cardiac interventionalists) was favoured by most responders regardless of patient's age (108, 80.6% for a 6 year old, and 109, 81% for a 16 year old). From this perspective, there were no differences among regions of practice. In the case of a coronary artery bypass grafting in younger children (6 year old), most respondents recommended a paediatric centre (119, 88.8%), with a combined surgical team of paediatric and adult cardiac surgeon (97, 72.4%). For older children (16 year old) undergoing CABG, the responses were also split (74, 55.2% vs 60, 44.8%) between an adult and paediatric cardiac centre. The preference remains for a combined surgical team for the older patients as well, regardless of the setting of intervention (Figure 6). North American respondents were significantly more likely to recommend a paediatric centre ($p = 0.002$) and a paediatric intervention team ($p < 0.0001$) compared to the rest of the world.

Discussion

While the acute treatment of Kawasaki disease has been well established, the management of its grave complication, coronary artery aneurysms, varies due to the lack of evidence-based data. Giant coronary artery aneurysms are rare but can be particularly fatal, thus warranting careful surveillance, appropriate pharmacological therapy, and well-established interventional measures—all of which have discrepancies between institutions and guidelines. This survey study collected and analysed the current management practices in giant coronary artery aneurysms after Kawasaki disease from physicians globally.

There are various guidelines for the management of Kawasaki disease, most concentrate on the acute and the subacute phase of the disease.[1,5,16–27] However, three main professional societies

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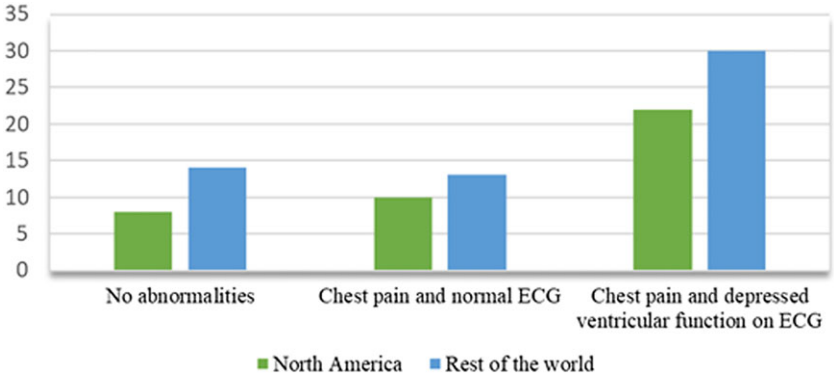

Figure 2. Use of cardiac catheterisation according to parts of the world.

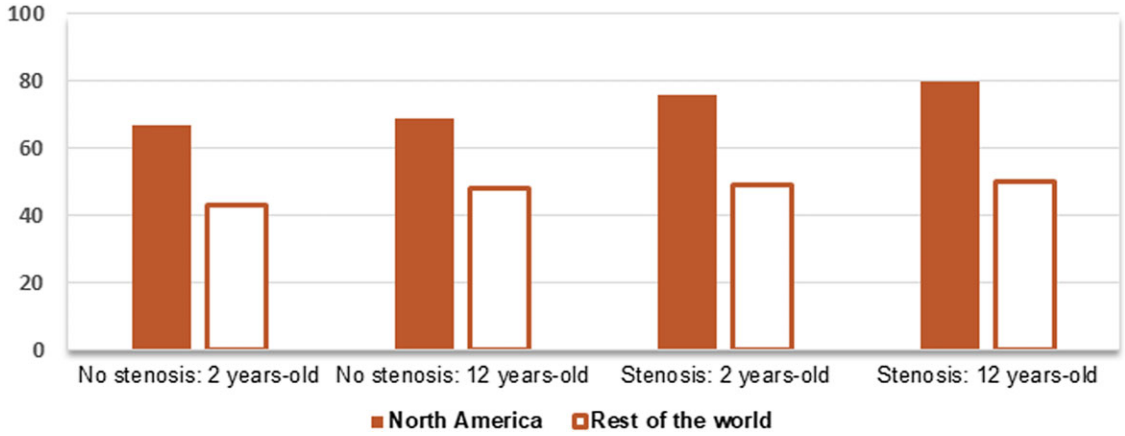

Figure 3. Percentage of respondents who use an association of aspirin and anticoagulant therapy according to parts of the world and age vignette.

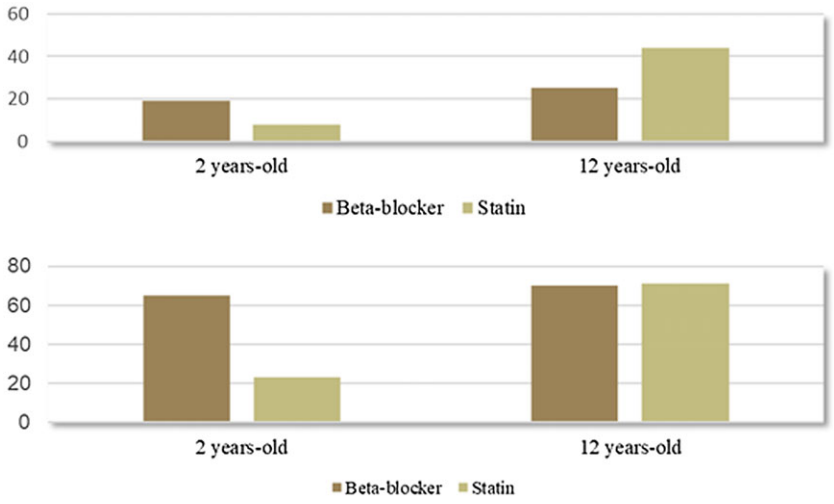

Figure 4. Use of beta-blockers and statins use based on age vignette and whether they were with (top) or without (bottom) coronary artery stenosis.

detail recommendations for long-term management of Kawasaki disease patients with CA aneurysms.[1,5,6] A certain degree of variations among these guidelines is evident. The 2017 American Heart Association statement tends to be the most conservative in the frequency of cardiology surveillance in those with giant coronary artery aneurysms (Z Score ≥10 or absolute dimension ≥8 mm) recommending echocardiography and electrocardiography at 1, 2, 3, 6, 9, and 12 months in the first year after an episode of acute Kawasaki disease and every 3 to 6 months thereafter.[1] Assessment for inducible myocardial ischaemia is recommended every 6–12 months. The Japanese Cardiovascular Society (JCS) guideline indicate electrocardiography and echocardiography assessment every 6–12 months,[5] and assessment for inducible ischaemia every 1–5 years.[1,5] The guidelines set by a United Kingdom lead group also differ slightly in that they recommend the consideration of stress imaging for inducible myocardial ischaemia annually.[6] Advanced coronary imaging modalities, such as CT, MRI, or other invasive tests, are similarly recommended in both JCS and American Heart Association guidelines—considered for periodic surveillance every 1–5 years after the first year.[1,5]

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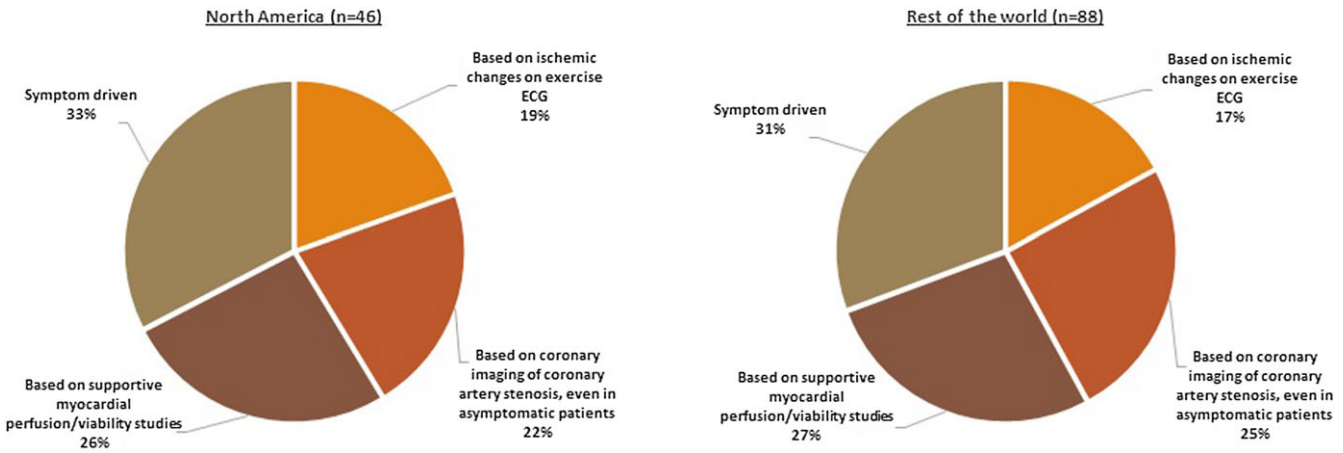

Figure 5. The indication for coronary intervention (surgical or percutaneous) was the most controversial topic among respondents, irrespective of geographic practice setting.

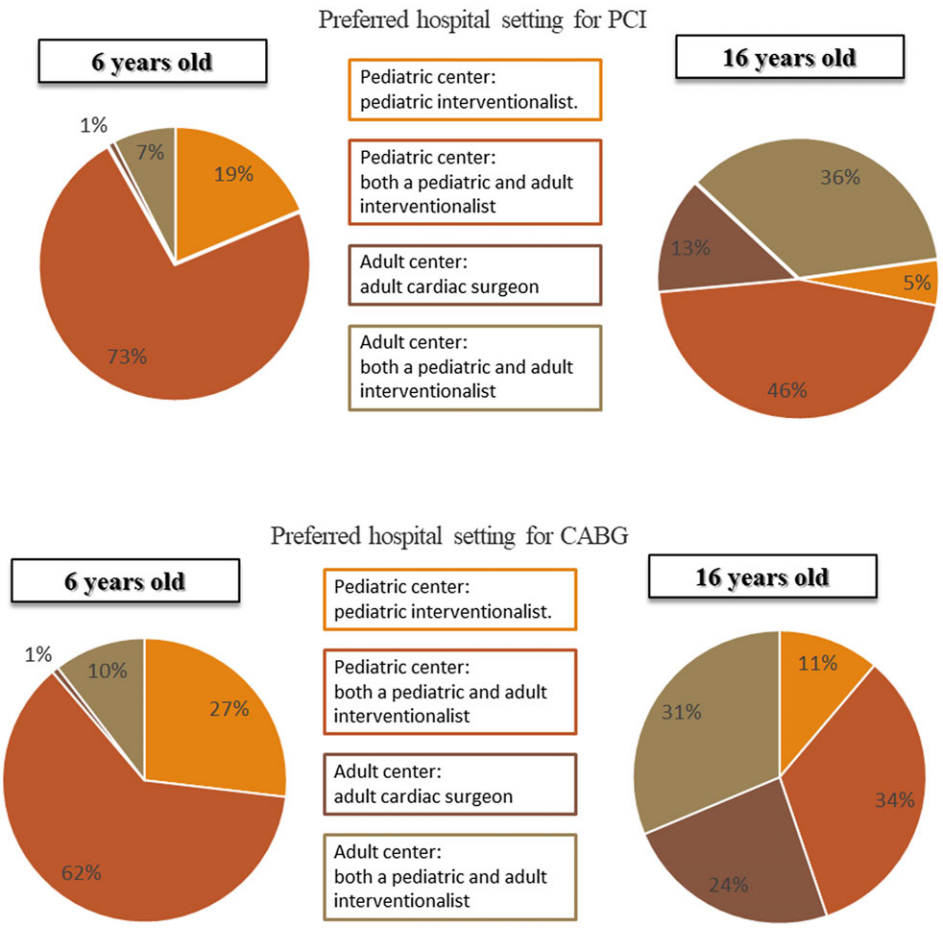

Figure 6. Preferred hospital setting for PCI (top) and CABG (bottom) according to patient age vignette.

However, CT is required at 6–12 months as a baseline, according to the United Kingdom management of giant aneurysms.[6] Among the 134 physicians who completed the survey, 50% were seeing less than ten Kawasaki disease patients per year, limiting experience and exposure. Time to follow-up beyond one year of diagnosis varied between responses, with about 53% of participants choosing an interval of 6 months - a middle-ground between the different guidelines (American Heart Association recommending 3–6 months and JCS recommending 6–12 months).

There was a clear variation in the first-choice imaging modalities in all cases of patients, as well as discrepancies between choices made in the present survey and available guidelines. Particularly, an asymptomatic patient with no ECG abnormalities was most likely to be investigated with a CTA, being a non-invasive but still sensitive method of detecting abnormalities in distal vessels and the presence of thrombus.[1] The 2020 JCS guidelines recommend cardiac catheterisation only in patients with evidence of inducible myocardial ischaemia on testing or documented

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significant coronary stenosis on imaging.[5] Cardiac catheterisation was still chosen by 17% of physicians in patients showing no signs of ischaemic events. The use of cardiac catheterisation varied among regions, with North American responders choosing it significantly less than their counterparts. Stress testing was appropriately chosen more frequently in patients with signs of an ischaemic event, as well as older patients. The specific stress test to be used remains vague, with responses being divided between stress echocardiography, stress MRI, and PET/SPECT nuclear stress test. Stress testing for inducible ischaemia remains a challenge in younger children, thus limiting their use and the occasion for data collection.

Combined aspirin and anticoagulant therapy is the most frequently chosen regimen in the present survey, which is concordant with available studies showing a 91% 10-year freedom from cardiac events under such a regimen.[28] The same study showed the reduction of myocardial infarction in patients under additional anticoagulation—from 16/49 patients treated with aspirin alone to 1/19 patients under combined aspirin and warfarin.[28] However, our study shows that practice region remained a factor of variation, with physicians from North America choosing said therapy more often. The use of beta-blockers and statins seemed to remain controversial, with consistently low votes from physicians, which increased with patient age and state of stenosis. Studies have shown that beta-blockers may reduce the risk of death, especially in the long-term management of giant coronary artery aneurysms.[1] Similarly, the empirical use of statins has been shown in small Kawasaki disease series to reduce C-reactive protein (CRP) and improve endothelial function, with little effect on child growth.[1] As such, the 2017 American Heart Association guidelines suggest that these adjunctive therapies could readily be used in Kawasaki disease patients with giant coronary artery aneurysms.[1] However, the modest size of these studies could be the cause for hesitation—multiple comments collected from respondents highlight the lack of evidence in the use of statins and, to a lesser extent, of beta-blockers.

In our study, the most chosen interventional treatment for coronary artery stenosis and thrombosis was PCI. However, current guidelines tend to favour coronary artery bypass grafting because of its greater likelihood of complete revascularisation.[1] In fact, the American Heart Association only recommends PCI for single-vessel disease or, in the case of significant comorbidities that make coronary artery bypass grafting too high-risk.[1] Moreover, PCI techniques such as balloon angioplasty have been shown to be a poor technique in the treatment of stenotic lesions due to their strong calcification. The optional comment section of the survey collected respondents' personal experiences with nine patients who had undergone coronary artery bypass grafting in their respective centre, with two having died from subsequent dilated cardiomyopathy. On the other hand, more than 40 reports of PCI cases were collected, which have been successful. Several responders highlight that the choice of undergoing PCI is due to lack of evidence and experience in performing coronary artery bypass grafting in children. Our survey could not address all nonsurgical methods used to alleviate CA obstruction. The particularity of Kawasaki disease giant coronary artery aneurysms anatomy adds another dimension to stenosis as a large thrombus could develop in the absence of (as well as in presence of stenosis), which may be addressed by in situ or systemic thrombolysis.[8] Current guidelines do not showcase recommendations regarding the appropriate choice of intervention. Our study points towards a clear preference for combined efforts by a paediatric interventionalist/surgeon and an adult interventionalist/surgeon. In this perspective, our findings are in line with the 2017 American Heart Association Scientific Statement addressing a major gap for a smooth and efficient transition of care between late childhood (adolescence) and the industrious adult life.[29] As pointed out in this survey, there is no magical formula in the care of giant coronary artery aneurysms patients as who is the best team to address the follow-up and the acute care of coronary Kawasaki disease complications. The need for close collaboration between paediatric and adult cardiologists is evident. The transmission of knowledge between these two disciplines must not be left to the odds of occurrence. Instead, as concluded in the above reference, "*a successful Health Care Transition program for patients with Kawasaki disease is a multipronged approach*" that must not only implicate "*the receiving team*" but the "*scientific societies and governing bodies*" as well.

Limitations

As with any survey, poor response rates is a challenging and a limiting factor in gaining an adequate and representative study population. Although we consider our results representative of general practice, higher participation rate from non-traditional Kawasaki disease expert countries could have brought different perspectives. The relatively small sample size of respondents, recruited from known associations and memberships, may not be representative of the specialties, geographical distribution, and breadth of management practices of those caring for patients with Kawasaki disease and large coronary artery aneurysm worldwide. Yet, to our knowledge, the survey provides Kawasaki disease practitioners' opinion from the largest survey on the management of giant coronary artery aneurysms from different parts of the world. It is conceivable, however, that physicians, who do not consider their opinion strong enough to matter, have elected to recuse themselves to fill out the survey.

Conclusion

In this international survey, we identified several areas of practice variation in the management of patients with giant coronary artery aneurysms after Kawasaki disease. Some practices showed discrepancies from the currently available guidelines, such as the 2017 American Heart Association and the 2020 JCS. Other practices differed significantly between different geographical regions notably between North America and the rest of the world. The lack of larger studies and evidence-based recommendations has certainly impacted management practices across the world as expressed in our results reflected in wide variations towards certain treatments. Improved international collaborative efforts are needed to provide robust evidence for the management and evolution of these patients. The conceptualisation of an international patient case database would certainly be a first step towards the advancement of research and treatment of Kawasaki disease patients with giant coronary artery aneurysms.

Supplementary material. The supplementary material for this article can be found at https://doi.org/10.1017/S1047951124026829.

Competing interests. None.

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