Journal of the American Heart Association

AHA SCIENTIFIC STATEMENT

Falling Through the Cracks: The Current Gap in the Health Care Transition of Patients With Kawasaki Disease

A Scientific Statement From the American Heart Association

Nagib Dahdah, MD, MBA; Samuel C. Kung, MD, MAS; Kevin G. Friedman, MD; Ariane Marelli, MD, MPH, FAHA; John B. Gordon, MD; Ermias D. Belay, MD; Annette L. Baker, RN, MSN, CPNP, FAHA; Dhruv S. Kazi, MD, MSc, MS, FAHA; Patience H. White, MD, MA; Adriana H. Tremoulet, MD, MAS, FAHA; on behalf of the American Heart Association Rheumatic Fever, Endocarditis, Kawasaki Disease Committee of the Council on Lifelong Congenital Heart Disease and Heart Health in the Young, and the Council on Arteriosclerosis, Thrombosis and Vascular Biology

BACKGROUND: Health care transition (HCT) is a period of high vulnerability for patients with chronic childhood diseases, particularly when patients shift from a pediatric to an adult care setting. An increasing number of patients with Kawasaki disease (KD) who develop medium and large coronary artery aneurysms (classified by the American Heart Association according to maximal internal coronary artery diameter Z-scores ≥5 and ≥10, respectively) are becoming adults and thus undergoing an HCT. However, a poor transition to an adult provider represents a risk of loss to follow-up, which can result in increasing morbidity and mortality.

METHODS AND RESULTS: This scientific statement provides a summary of available literature and expert opinion pertaining to KD and HCT of children as they reach adulthood. The statement reviews the existing life-long risks for patients with KD, explains current guidelines for long-term care of patients with KD, and offers guidance on assessment and preparation of patients with KD for HCT. The key element to a successful HCT, enabling successful transition outcomes, is having a structured intervention that incorporates the components of planning, transfer, and integration into adult care. This structured intervention can be accomplished by using the Six Core Elements approach that is recommended by the American Academy of Pediatrics, the American Academy of Family Physicians, and the American College of Physicians.

CONCLUSIONS: Formal HCT programs for patients with KD who develop aneurysms should be established to ensure a smooth transition with uninterrupted medical care as these youths become adults.

Key Words: AHA Scientific Statements ■ coronary artery aneurysms ■ health care transition ■ Kawasaki disease ■ transition of care

Over the past several decades, there has been a significant increase in the number of children and adolescents with chronic conditions surviving into adulthood, with important implications for transition from pediatric to adult health care systems. As a result, there has been a sizeable increase in young adults with childhood-onset conditions entering adult health care facilities. Health care transition (HCT) is the process of moving from a child to an adult model of health care and includes changing from pediatric, parent-supervised health care to more independent, patient-centered adult health care. Without a planned HCT process, barriers to HCT are experienced by children, young adults, and families, and may result in increased morbidity, mortality, and health care costs.[1] The field of HCT is a relatively new field. It was only in 2011 that the American Academy of Pediatrics published on the importance of HCT programs in pediatrics.[2] That same year, the Transition Readiness Assessment Questionnaire, a measure of readiness for transition from pediatric to adult health care for youth with special health care needs, was internally validated.[3] The 2016 to 2017 National Survey

JAHA is available at: www.ahajournals.org/journal/jaha

Opening Vignette

While growing up, my understanding of Kawasaki disease (KD) was limited. I was diagnosed with KD when I was 17 months old. As a child, I remember numerous visits to see cardiologists, while as a teenager I underwent stress tests and imaging studies. By then, I had a vague idea of what KD was, but I did not fully realize the potential complications of my coronary artery aneurysms. When it was my time for transition to adult-based care, it was unfortunately not well coordinated. Follow-up visits became more sporadic throughout college and beyond. I equated a lack of symptoms with a clean bill of health, not fully comprehending the thrombotic risks associated with my aneurysms. I did not revisit KD again until several years later in medical school, when I was drawn to work on KD research and learned about anticoagulation therapy. I was fortunate to not have experienced a heart attack, but it is evident that lapses in my health care transition could have ultimately led to disastrous outcomes. My story is a call to improve the transition of care for patients with KD worldwide, lest we fall through the cracks.

—Samuel C. Kung, MD, MAS

of Children's Health reported that only 17% of children with special needs and 14% of children without special needs received transition-planning guidance from health care professionals.[4]

Young adults with a history of Kawasaki disease (KD), the most common cause of acquired heart disease in children, are faced with similar HCT barriers. A survey in Japan, published in 2018, found that >40% of patients with KD and coronary artery lesion had been lost to follow-up.[5] Given the undeniable risk of life-threatening coronary artery complications following KD, specific emphasis for HCT is even more relevant.[6] The key steps to successful HCT in patients with KD and with coronary artery disease, including an emphasis on patient education and self-management skills, are just starting to be recognized.[7] Having a guide to the elements needed in an HCT program for children with coronary artery damage attributable to KD is critical to ensure their safety as they move into adulthood. This document reviews the existing life-long risks and current guidelines for long-term care of patients with KD and offers guidance on assessment and preparation of patients with KD for HCT as well as key elements for a successful HCT program.

EPIDEMIOLOGY AND LIFE-LONG RISKS FOR PATIENTS WITH KD

The incidence of KD, the most common cause of acquired heart disease in children, is highest in Japan, where by 10 years of age, >1 in 100 children have a history of KD, with 1 in every 65 boys and 1 in every 82 girls diagnosed with KD.[8,9] In the United States, despite some seasonal variations in KD, incidence rates are 20 to 25 per 100 000 children <5 years of age, >10 times lower than that in Japan. Despite a lower incidence, it is estimated that 1 in 1600 Americans will have a history of childhood KD by 2030.[10] KD is reported in >60 countries, with increasing incidence in many parts of the world.[11–14] The mortality rate in KD is generally <1%, likely an underestimation given missed cases and that this is not a reportable disease in many parts of the world.[13,15] However, some patients experience significant morbidity related to the development of coronary artery aneurysms, defined by the American Heart Association as Z-score (internal diameter adjusted for body surface area) ≥2.5.[16] Coronary artery aneurysms are classified by the patient's maximal coronary Z-score as small (Z-score ≥2.5 and <5.0), medium (Z-score ≥5.0 and <10), and large or giant (Z-score ≥10).[17] About 20% of patients with KD develop transient coronary artery dilatation (Z-score ≥2 and <2.5), and up to 5% develop medium or large coronary artery aneurysms.[18] In a comparative analysis of coronary artery measurements from American and Japanese patients with KD, nearly 30% of patients in the United States and 44% of patients in Japan met the cut point of a coronary artery Z-score ≥2.5%, and 5% of patients from both countries developed coronary artery aneurysms with a maximum Z-score ≥5.[19] Extremes of age (infants and children >8 years), male sex, delayed diagnosis and treatment, and higher markers of inflammation at the time of diagnosis have been associated with increased risk of cardiovascular complications,[20–23] specifically coronary artery aneurysms.

Although coronary artery aneurysm regression is reported in 75% of patients, long-term effects in the inflamed arterial wall remain a concern, especially in patients with KD with giant aneurysms that have not regressed. With aneurysmal remodeling or regression, there is the risk that myofibroblast proliferation can result in abnormal vascular reactivity.[24,25] Thrombosis and progressive stenosis of a giant aneurysm can cause ischemic heart conditions, myocardial infarction, and sudden cardiac death in up to 48% of such patients.[16,26]

Additional lines of study suggest that long-term follow-up is necessary for patients with KD and with coronary artery issues. Optical coherence tomography imaging of patients with KD with various degrees of coronary sequelae has shed new light on the vascular damage in these patients. In a series of 33 patients aged 12.0±5.4 years, lesions, such as parietal fibrosis, cellular infiltrates, intimal hyperplasia, or neovascularization, were detected even in some coronary artery segments without previous dilation by echocardiogram. Similar lesions were present in significantly higher proportions in coronary segments

with persistent aneurysms, followed by segments with angiographically regressed aneurysms and segments with coronary artery dilatation but no previous evidence of aneurysms.[27] A recent review of mid-term complications after acute KD in patients with documented coronary artery aneurysms was completed by the International KD Registry.[28] In this cohort of 1651 patients with KD, patients with coronary artery aneurysms with Z-scores <10 were primarily risk free, with only 1 patient with an aneurysm Z-score of 8.5 showing chronic cardiac ischemia, resulting in heart failure and death. The risk of luminal narrowing and coronary artery thrombosis in this cohort was otherwise restricted to patients with large or giant aneurysms (Z-score ≥10). The strongest predictor of cardiac risk continues to be the extent of damage to the coronary arteries. The persistence of large coronary artery aneurysms has a high risk of complications, which does not totally exclude those with remodeled aneurysms.[6,29] Beyond the coronary complications, myocarditis is seen in the acute phase of KD, with myocardial function generally thought to be normal beyond the acute phase in patients who do not develop ischemic heart disease.[30] Nevertheless, increased left ventricle volume, decreased strain, altered diastolic parameters up to 12 months after onset of KD, and increased myocardial collagen distribution and content irrespective of the presence or the absence of coronary aneurysms suggest a possible long-term effect on myocardial structure and function.[31]

CURRENT AMERICAN HEART ASSOCIATION KD LONG-TERM GUIDELINES

According to the 2017 American Heart Association KD guidelines, patients with KD with no history of coronary aneurysms do not appear to be at increased risk of cardiac events in adulthood and typically do not require long-term cardiology follow-up.[16,24,32] Patients with KD and with aneurysms, either persistent or those that have been remodeled and have decreased to a normal internal luminal dimension, should have long-term follow-up, with their care transitioned to an adult cardiologist once they reach adulthood, generally between the ages of 18 and 21 years.[16] Most important, patients with documented and persistent moderate or large aneurysms require life-long surveillance with yearly or biyearly assessment.[16] Therefore, there remains a significant number of patients with KD who should have long-term cardiology follow-up into adulthood.

Transition from pediatric to adult care for these patients needs to be a process involving a deliberate and coordinated series to ensure uninterrupted care. Adult patients with KD would benefit from follow-up with an adult cardiologist who understands both coronary artery disease as well as unique issues related to KD.

As KD typically occurs in young children, the 2017 KD guidelines laid out several educational goals that are important to discuss with parents as well as patients with KD starting at approximately age 12 years. These "knowledge goals" include: (1) review of details of their illness, including sustained complications, cardiac sequelae, any cardiac events, and related procedures; (2) the names and rationale for any diagnostic tests performed; (3) precise information on medications that are required; (4) importance and rationale behind restrictions on physical activity if any; and (5) specific symptoms or signs that warrant immediate medical attention. This information must be understood and retained by the patient, who should also understand the importance of uninterrupted life-long cardiology care. Considerations on contraception and pregnancy of female patients warrant clarification. Later on, the small but real risk of the possibility of KD in offspring should be understood. Finally, long-term prognosis and associated health issues should be discussed as well as preventive strategies to achieve healthy lifestyle behaviors.

"Self-management skills" should be made apparent to the young adult with KD at the time of transfer to increase the chance of success in the HCT. Those skills include the ability to contact health care professionals, to schedule and attend appointments and tests, as well as to know when and how to access emergency care or mental health services. Other self-caring goals are more specific, such as creating and using a portable health summary and maintaining health records and adhering to a medication regimen, including requesting prescription refills and being able to communicate independently and effectively with health care professionals.

However, despite these established objectives, there is currently no clear path for HCT in many centers that care for patients with KD. Some centers transition patients with KD to adult cardiologists, others consider adult congenital heart specialists, independently or in conjunction with KD specialists, whereas other centers delay HCT indefinitely.

HCT PLANNING: THE NEED FOR READINESS

In 2018, the American Academy of Pediatrics, the American Academy of Family Physicians, and the American College of Physicians jointly published the "Clinical Report: Supporting the Health Care Transition From Adolescence to Adulthood in the Medical Home."[1] This report builds on an earlier 2011 clinical report and represents professional organizations' consensus on practice-based implementation of HCT for all children and youth beginning in early

adolescence and continuing into young adulthood.[2] The American Academy of Pediatrics, the American Academy of Family Physicians, and the American College of Physicians recommend a structured transition that is supported by 2 systematic reviews.[33–35] These systematic reviews show statistically significant positive transition outcomes for youth with special needs in the areas of: (1) population health (adherence to care, self-care skills, quality of life, and self-reported health), (2) experience of care (increased satisfaction and reduction in barriers to care), and (3) utilization (decrease in time between last pediatric and first adult visit, increase in adult ambulatory visits, and decrease in hospital admissions and length of stay) with use of this approach. This evidence-based structured transition incorporates the following components: planning, transfer, and integration into adult care. To accomplish these 3 components of HCT, the 2018 American Academy of Pediatrics, American Academy of Family Physicians, and American College of Physicians clinical report recommends using an approach called the Six Core Elements of HCT (Figure 1). This approach was developed by Got Transition, the federally funded national HCT center, and has been tested in a series of quality improvement learning collaboratives that demonstrated the effectiveness of an organized transition process in pediatric, family medicine, and internal medicine practices. The Six Core Elements (policy, tracking, readiness, planning, transfer of care, and transfer completion), with sample customizable tools and measurement and implementation resources (https://www.gottransition.org/six-core-elements/), have been widely used in primary and specialty care systems[36] (Figure 1). This report reiterated that HCT is not just transfer alone but that all 3 components are required and that the HCT process should start early, between the ages of 12 and 14 years.

PREPARATION OF YOUTH WITH KD FOR HCT: A STARTING POINT

A key starting point for improving transition is preparation of youth with KD as well as parents/caregivers for an adult model of care. This preparation includes the following elements, outlined in the Six Core Elements approach for the pediatric clinician (Figure 1).

The first core element is policy, which includes preparing an office transition guide for families, including discussion of the practice's approach to an adult model of care around privacy and consent. The second core element is tracking, which ensures that a patient is offered a transition readiness/self-care skill assessment conducted periodically. The third core element is transition readiness. The pediatric clinician should have a standardized way of assessing the youth's self-care skills. The fourth core element is planning, which involves creating a transition plan jointly developed with youth and families that includes preparing for decision-making support, assembling a medical summary and emergency care plan shared with youth and families, identifying an adult cardiologist and primary care clinician, and preparing transfer information for and communication with the new clinician. The fifth core element is transfer of care, which requires engagement[37] during this vulnerable transition time of both the adult clinician and the pediatric subspecialist to help the young adult schedule a follow-up appointment with the adult health care clinician within 3 to 6 months of leaving the pediatric clinician. The sixth core element is transition completion, during which the pediatric clinician confirms that the young adult has

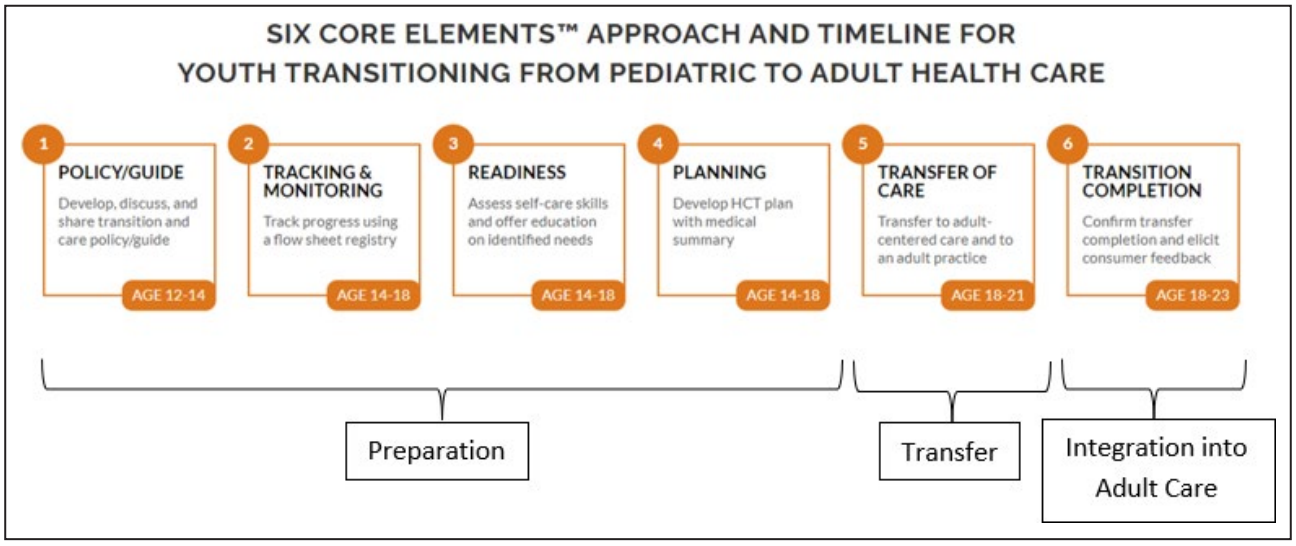

Figure 1. Six Core Elements Approach and Timeline for Youth Transitioning From Pediatric to Adult Health Care.
Age is presented in years. Adapted from steps of the Six Core Elements of Health care Transition (HCT). © 2020 Got Transition (www.GotTransition.org).[36]

been seen by the adult clinician and facilitates obtaining patient feedback about the HCT process.

HCT preparation involving all 6 core elements is a process that evolves over several years and includes working with the adolescent's developmental stages to achieve maturity, education, and self-management. Integrating the age of transition with the required HCT steps and elements results in an uninterrupted, longitudinal process; this process leads to the highest probability of success (Figure 2).[37] Just as there is a Six Core Elements package for pediatric primary care and pediatric cardiologists, there is also a Six Core Elements package for adult primary care and adult cardiologists to assist them in integrating and engaging the young adult with KD into their practice (https://www.gottransition.org/six-core-elements/integrating-young-adults/).[36]

TRANSITION READINESS OF THE PATIENT WITH KD FOR HCT

One of these preparation elements involves conducting a readiness or self-care skill assessment to evaluate the young adults' understanding about their own health and how to navigate adult health care. There are several readiness assessment tools available, but none to date has been externally validated in any disease, including KD.[38–45] This means that a high score on any of the readiness assessments does not necessarily predict a successful outcome. Use of a readiness/self-care assessment tool is primarily to start the conversation on what the young adult can do to better prepare for an adult approach to care and to an adult health care system. One aspect of transition readiness is to understand the youth's and families' perception of transition and their willingness to engage in the transition process.

Got Transition's readiness assessment tool assesses the youth's overall approach to HCT by asking 2 motivational interviewing questions: (1) How important is it to you to transition to an adult provider or to manage your own health care? (2) How confident are you in transitioning to an adult clinician or managing your own health care? These questions have a scale answer from 1 to 10, where 0 is not important or confident and 10 is very important or confident.[46] These 2 questions can assist the clinician gauge where the youth is in his or her overall readiness, letting the clinician know if the youth does not feel transition is important and whether the individual is confident in his or her ability to move forward. To further explore the importance or confidence of transitioning, more probing questions can be asked (Table 1).[47] Many adult and pediatric specialty clinicians in collaboration

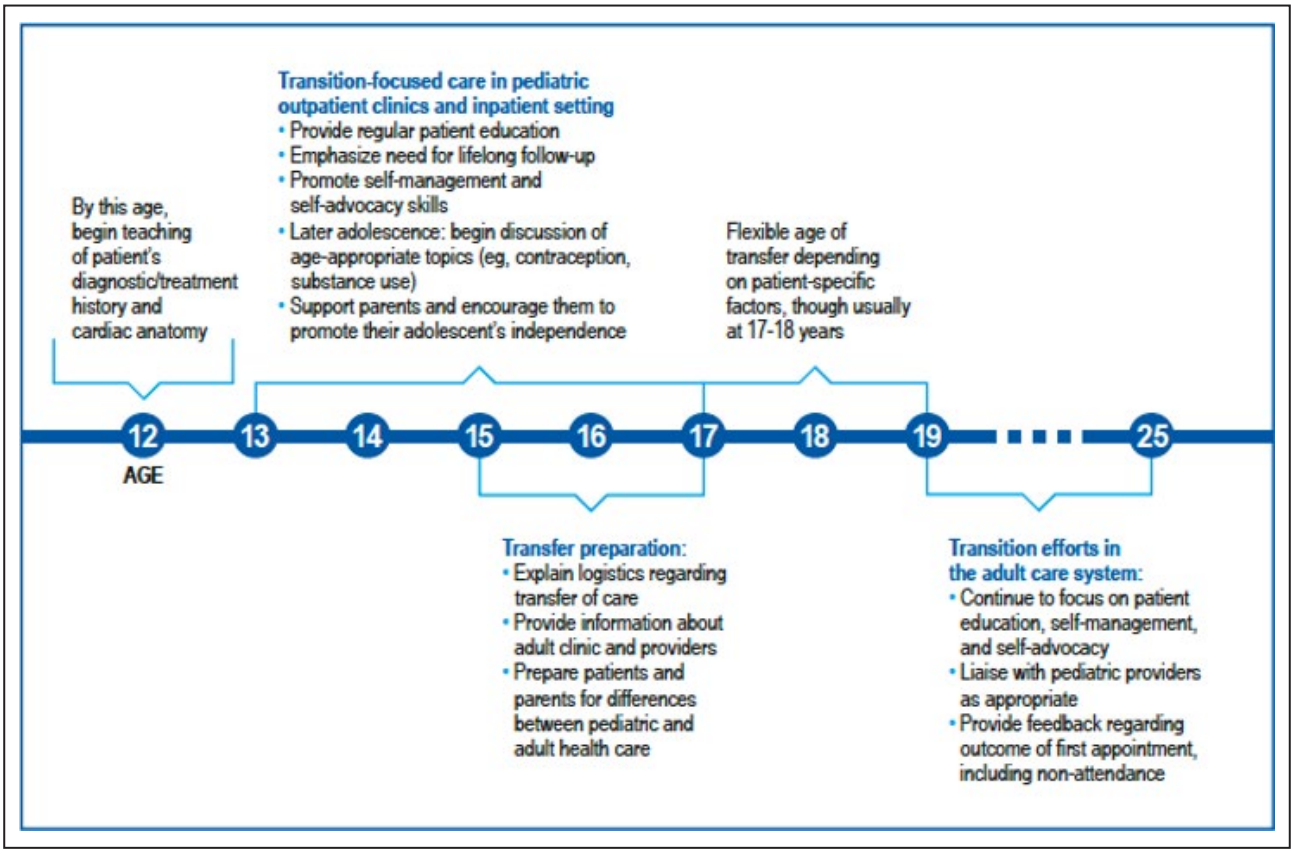

Figure 2. Suggested timeline for transition and transfer of care.
Age is presented in years. Reprinted with permission from Mackie et al. © 2019 Canadian Cardiovascular Society.[37]

with patient representatives and pediatric and adult stakeholders have customized the transition readiness assessment tool and other tools from the Six Core Elements for particular diseases. Such examples include the American College of Cardiology's congenital heart disease HCT information and the American Heart Association Congenital Heart Defects Committee.[48,49] Similarly, a KD-specific transition readiness assessment tool can be created for patients with KD.

FINDING THE RIGHT ACCEPTING PHYSICIAN

HCT to an adult cardiologist is potentially difficult because of lack of exposure to KD for most adult cardiologists in training and clinical practice. The clinical issues to be managed revolve around the risk for thrombosis and stenosis of the coronary and peripheral vessels. Prevention and management of coronary thrombosis or stenosis associated with coronary aneurysms represents challenges that may differ from those seen with atherosclerotic disease. The use of statins in children with complicated KD is becoming more common but for reasons other than decreasing cholesterol.[50,51] However, the literature on the utility of statins in the adult with complicated KD is lacking. Because coronary pathology following KD differs from that caused by atherosclerosis, the in situ anti-inflammatory effects of statins may be taken into consideration for adult patients with KD and with coronary artery complications.[52–54] When complications do arise, percutaneous interventions can be challenging because of a large clot burden (that can obscure vessel size and result in undersized stenting) or dense calcification of lesions (requiring atherectomy before dilatation).[55] Surgical bypass in the setting of a moderate clot burden and good flow can lead to graft failure because of competitive flow in the native vessel. An adult cardiologist cognizant of the natural history of KD coronary aneurysms who can partner with experienced interventional cardiologists or cardiothoracic surgeons would be well suited to manage these more affected patients.

Table 1. Questions to further explore the importance and confidence in transitioning

Exploring importance
• What would have to happen to make transferring to an adult clinician and managing your own health feel more important to you? • Why have you given yourself such a high or low score on importance to transferring to an adult clinician? • What would need to happen for your importance score to move up from x to y? • What worries you about moving up from x to y? • What are the good things about preparing for transition to an adult clinician? • What concerns do you have about transferring to an adult clinician? • If you were to transfer to an adult clinician, what would it be like?
Exploring confidence
• What would make you more confident about taking charge of your health/transferring to an adult clinician? • Why have you given yourself such a high or low score on confidence? • How could you feel more confident about managing your health/transferring to an adult clinician, so your score goes from x to y? • How can I help you succeed in taking charge of your own health? • What are some of the practical things you need to do to feel more confident in managing your health/transferring to an adult clinician? Do any of them sound achievable? • Is there anything you can think of that would help you feel more confident?

NEEDS OF THE HCT KD PROGRAM

Although many adult health care centers may only see a handful of adults with KD, developing and adopting standardized practice workflows based on evidence and published guidelines can help ensure that patients with coronary artery damage from KD in childhood receive high-quality care. With a plethora of clinical guideline publications, quality of care delivery has been closely linked to standardized practice based on evidence.[1,56] Over the past decade, the focus of quality has begun to shift from standardization to personalization. Delivering proactive, rather than reactive, care in a manner that is personalized and patient centered is emerging as a cornerstone to quality care.[57,58] For patients with KD, where genetics, environment, and behavior converge to determine outcomes, quality of care despite low patient volumes is particularly important.

Quality determinants of HCT programs can be assessed using an adapted Donabedian approach, characterizing structure, process, and outcomes related measures of patients with KD during HCT.[59] The structural components of HCT delivery need to reflect the required interdisciplinary expertise (notably, adult and pediatric care providers with on-site interventional cardiac catheterization capabilities as well as the capacity to perform coronary artery bypass grafting by expert surgeons). This combination of expertise can most often be found in tertiary or quaternary care institutions. Process measures of quality require the ability of an adult cardiologist to prevent complications of coronary disease, anticipate the risk of coronary complications with noninvasive and invasive imaging modalities, and treat acute coronary syndromes when they occur. This hybrid expertise is vital for optimizing care delivery for the growing number of pediatric patients with KD reaching adulthood. Ultimately, quality outcomes in HCT means minimizing the care gaps that can lead to avoidable complications and adverse comorbidity related to myocardial dysfunction, including the prevention of the delayed presentation of acute myocardial infarction and sudden cardiac death. Resource allocation for HCT programs for KD, as with other conditions, depends on the critical mass of patients, the institutional priorities, competing demands on the health care system, and the ability to deliver cost-effective health management solutions.

MODELS OF HCT TO DELIVER THE BEST CARE

Like other youth with pediatric conditions that extend across the lifespan, the course of patients with KD and a history of coronary artery aneurysms has long periods of stability variably punctuated by acute but reversible deterioration.[6] In light of the variation in the patterns of disease expression, care delivery needs to be nimble and highly responsive to patients and families as needs wax and wane throughout the longevity that survival permits. In general, HCT programs can be established using one of several models: as part of broader HCT interdisciplinary programs aimed at supporting patients with chronic conditions, as part of a single specialty care program depending on the organ affected, or as dedicated stand-alone structures. There may certainly be barriers encountered to establishing an HCT program given the limitation of resources available in certain settings. In the absence of the ability to create a full interdisciplinary program, an alternative may be to assess a patient's readiness for transfer by evaluating the patient's understanding of the illness, medications, and rationale for certain testing starting at age 12 years, identifying an adult provider in the community who understands the complexities of KD and is willing to follow these patients, and providing that adult provider and patient with KD a medical summary that delineates key clinical issues, including the worst Z score, most recent cardiac imaging, and medications.

RESEARCH NEEDS

Although much progress has been made in designing and implementing HCTs over the past decade, there are critical gaps in knowledge about their optimal implementation and effectiveness that must be addressed in future studies. Specifically for patients with KD, areas of future study include the following: (1) evaluating the impact of HCTs from pediatric to adult health care; (2) studying transition outcomes in terms of population health (eg, adherence to care and self-care skill development), experience of youth, young adults, families, and clinicians, and use (eg, time between last pediatric and first adult visit, adherence to adult clinician appointments, and decreased emergency department and urgent care use) and cost savings; and

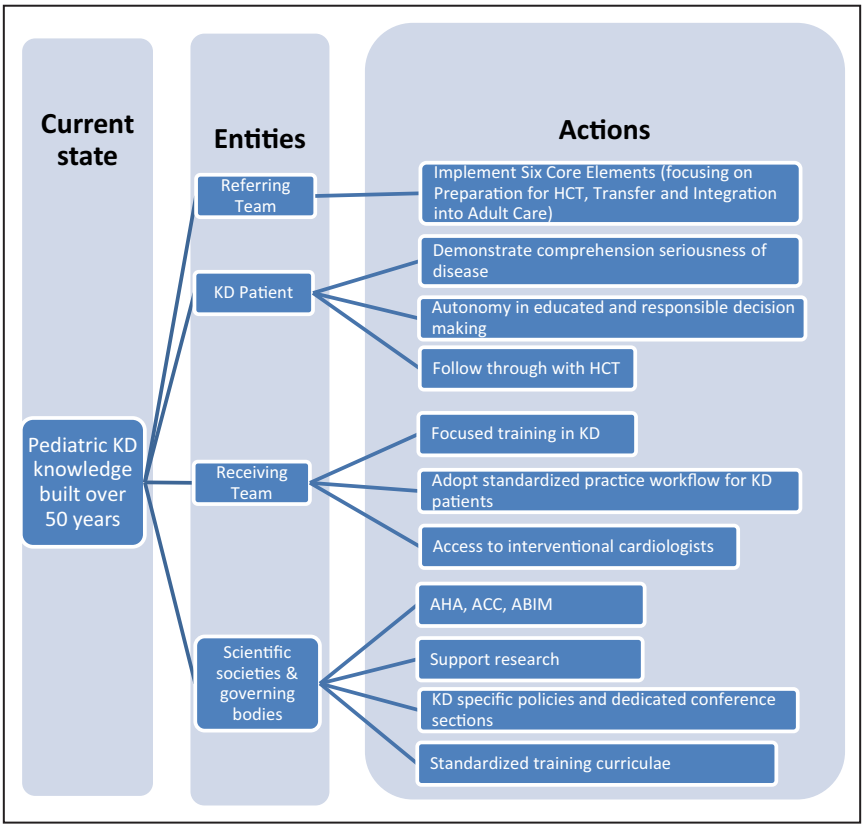

Figure 3. A comprehensive approach for proper transfer of care focuses on preparedness of the patient's team that is referring the patients, the patient with Kawasaki disease (KD), the receiving team, and scientific societies and governing bodies.

ABIM indicates American Board of Internal Medicine; ACC, American College of Cardiology; AHA, American Heart Association; and HCT, health care transition.

(3) conducting surveys of young adults about their experience with transferring and integrating into adult care.

Digital technologies can be incorporated into a variety of disease models to direct patients and families to appropriate resources in a timely manner. Their effectiveness should be the subject of future studies.[60,61] The life-course health development framework was created to shift the emphasis away from disease and toward health, with the knowledge that health is a consequence of genetic, biological, and social determinants, and with the understanding that health development is an adaptive process.[62] This is particularly relevant to patients with KD who will have largely normal lives, potentially punctuated with episodes of disease attributable to cardiac damage in childhood. Thus, the maintenance of health as an adaptive process that requires an understanding of risk management over long periods of time will lead to a more personalized approach to surveillance recommendations not only during transition of care but across the life course.

CONCLUSIONS

In summary, a successful HCT program for patients with KD is a multipronged approach that focuses on preparedness of the KD patient's team that is referring the patients, the patient with KD, the receiving team, and scientific societies and governing bodies (Figure 3).

ARTICLE INFORMATION

The American Heart Association makes every effort to avoid any actual or potential conflicts of interest that may arise as a result of an outside relationship or a personal, professional, or business interest of a member of the writing panel. Specifically, all members of the writing group are required to complete and submit a Disclosure Questionnaire, showing all such relationships that might be perceived as real or potential conflicts of interest.

This statement was approved by the American Heart Association Science Advisory and Coordinating Committee on April 16, 2021, and the American Heart Association Executive Committee on May 24, 2021. A copy of the document is available at https://professional.heart.org/statements by using either "Search for Guidelines & Statements" or the "Browse by Topic" area.

The American Heart Association requests that this document be cited as follows: Dahdah N, Kung SC, Friedman KG, Marelli A, Gordon JB, Belay ED, Baker AL, Kazi DS, White P, Tremoulet A; on behalf of the American Heart Association Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee of the Council on Lifelong Congenital Heart Disease and Heart Health in the Young; and Council on Arteriosclerosis, Thrombosis and Vascular Biology. Falling through the cracks: the current gap in the health care transition of patients with Kawasaki disease: a scientific statement from the American Heart Association. *J Am Heart Assoc*. 2021;10:e023310. DOI: 10.1161/JAHA.121.023310

The expert peer review of American Heart Association (AHA)–commissioned documents (eg, scientific statements, clinical practice guidelines, systematic reviews) is conducted by the AHA Office of Science Operations. For more on AHA statements and guidelines development, visit https://professional.heart.org/statements. Select the "Guidelines & Statements" drop-down menu, then click "Publication Development."

Disclosures

Writing Group Disclosures

Writing group member	Employment	Research grant	Other research support	Speakers' bureau/ honoraria	Expert witness	Ownership interest	Consultant/ advisory board	Other
Nagib Dahdah	CHU Sainte-Justine (Canada)	None	None	None	None	None	None	None
Adriana Tremoulet	University of California, San Diego	None	None	None	None	None	None	None
Annette L. Baker	Boston Children's Hospital	None	None	None	None	None	None	None
Ermias D. Belay	Centers for Disease Control and Prevention	None	None	None	None	None	None	None
Kevin G. Friedman	Children's Hospital Boston	None	None	None	None	None	None	None
John B. Gordon	San Diego Cardiac Center and Sharp Memorial Hospital	None	None	None	None	None	None	None
Dhruv S. Kazi	Richard A. and Susan F. Smith Center for Outcomes Research	None	None	None	None	None	None	None
Samuel C. Kung	UCSD School of Medicine	NIH (awarded for 6/2018–6/2019 to support training toward obtaining MAS in Clinical Research [field of study: Kawasaki disease])*	None	None	None	None	None	None
Ariane Marelli	McGill University (Canada)	None	None	None	None	None	None	None
Patience White	The National Alliance	None	None	None	None	None	None	None

This table represents the relationships of writing group members that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all members of the writing group are required to complete and submit. A relationship is considered to be "significant" if (1) the person receives ≥$10 000 during any 12-month period, or ≥5% of the person's gross income; or (2) the person owns ≥5% of the voting stock or share of the entity, or owns ≥$10 000 of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.* Modest.

Reviewer Disclosures

Reviewer	Employment	Research grant	Other research support	Speakers' bureau/ honoraria	Expert witness	Ownership interest	Consultant/ advisory board	Other
Moshe Arditi	Cedars-Sinai Medical Center	None	None	None	None	None	None	None
Georgi Christov	Great Ormond Street Hospital (United Kingdom)	None	None	None	None	None	None	None
Michael H. Gewitz	New York Medical College	None	None	None	None	None	None	None
Silvana Molossi	Texas Children's Hospital/Baylor College of Medicine	None	None	None	None	None	None	None
Anne H. Rowley	Northwestern University	National Institutes of Health (research grant to identify antigenic targets of Kawasaki disease)*; Dr Ralph and Marian Falk Catalyst Award (research grant to investigate a new human hepacivirus as a causative agent of Kawasaki disease)*	None	None	None	None	None	None

This table represents the relationships of reviewers that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all reviewers are required to complete and submit. A relationship is considered to be "significant" if (1) the person receives ≥$10 000 during any 12-month period, or ≥5% of the person's gross income; or (2) the person owns ≥5% of the voting stock or share of the entity, or owns ≥$10 000 of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.* Significant.

REFERENCES

1. White PH, Cooley WC; Transitions Clinical Report Authoring Group, American Academy of Pediatrics, American Academy of Family Physicians and American College of Physicians. Supporting the health care transition from adolescence to adulthood in the medical home. *Pediatrics*. 2018;142:e20182587. doi: 10.1542/peds.2018-2587
2. American Academy of Pediatrics, American Academy of Family Physicians, American College of Physicians, Transitions Clinical Report Authoring Group, Cooley WC, Sagerman PJ. Supporting the health care transition from adolescence to adulthood in the medical home. *Pediatrics*. 2011;128:182–200. doi: 10.1542/peds.2011-0969
3. Sawicki GS, Lukens-Bull K, Yin X, Demars N, Huang IC, Livingood W, Reiss J, Wood D. Measuring the transition readiness of youth with special healthcare needs: validation of the TRAQ–Transition Readiness Assessment Questionnaire. *J Pediatr Psychol*. 2011;36:160–171. doi: 10.1093/jpepsy/jsp128
4. Lebrun-Harris LA, McManus MA, Ilango SM, Cyr M, McLellan SB, Mann MY, White PH. Transition planning among US youth with and without special health care needs. *Pediatrics*. 2018;142:e20180194. doi: 10.1542/peds.2018-0194
5. Kamiyama H, Ayusawa M, Ogawa S, Saji T, Hamaoka K. Health-care transition after Kawasaki disease in patients with coronary artery lesion. *Pediatr Int*. 2018;60:232–239. doi: 10.1111/ped.13500
6. Tsuda E, Tsujii N, Hayama Y. Stenotic lesions and the maximum diameter of coronary artery aneurysms in Kawasaki disease. *J Pediatr*. 2018;194:165–170.e2. doi: 10.1016/j.jpeds.2017.09.077
7. Kovacs AH, McCrindle BW. So hard to say goodbye: transition from paediatric to adult cardiology care. *Nat Rev Cardiol*. 2014;11:51–62. doi: 10.1038/nrcardio.2013.172
8. Makino N, Nakamura Y, Yashiro M, Kosami K, Matsubara Y, Ae R, Aoyama Y, Yanagawa H. Nationwide epidemiologic survey of Kawasaki disease in Japan, 2015–2016. *Pediatr Int*. 2019;61:397–403. doi: 10.1111/ped.13809
9. Nakamura Y, Yashiro M, Yamashita M, Aoyama N, Otaki U, Ozeki Y, Sano T, Kojo T, Ae R, Aoyama Y, et al. Cumulative incidence of Kawasaki disease in Japan. *Pediatr Int*. 2018;60:19–22. doi: 10.1111/ped.13450
10. Huang SK, Lin MT, Chen HC, Huang SC, Wu MH. Epidemiology of Kawasaki disease: prevalence from national database and future trends projection by system dynamics modeling. *J Pediatr*. 2013;163:126–131.e1. doi: 10.1016/j.jpeds.2012.12.011
11. Lin YT, Manlhiot C, Ching JC, Han RK, Nield LE, Dillenburg R, Pepelassis D, Lai LS, Smythe JF, Chahal N, et al. Repeated systematic surveillance of Kawasaki disease in Ontario from 1995 to 2006. *Pediatr Int*. 2010;52:699–706. doi: 10.1111/j.1442-200X.2010.03092.x
12. Holman RC, Curns AT, Belay ED, Steiner CA, Schonberger LB. Kawasaki syndrome hospitalizations in the United States, 1997 and 2000. *Pediatrics*. 2003;112:495–501. doi: 10.1542/peds.112.3.495
13. Singh S, Vignesh P, Burgner D. The epidemiology of Kawasaki disease: a global update. *Arch Dis Child*. 2015;100:1084–1088. doi: 10.1136/archdischild-2014-307536
14. Lin MT, Wu MH. The global epidemiology of Kawasaki disease: review and future perspectives. *Glob Cardiol Sci Pract*. 2017;2017:e201720. doi: 10.21542/gcsp.2017.20
15. Chang RK. Hospitalizations for Kawasaki disease among children in the United States, 1988–1997. *Pediatrics*. 2002;109:e87. doi: 10.1542/peds.109.6.e87
16. McCrindle BW, Rowley AH, Newburger JW, Burns JC, Bolger AF, Gewitz M, Baker AL, Jackson MA, Takahashi M, Shah PB, et al. Diagnosis, treatment, and long-term management of Kawasaki disease: a scientific statement for health professionals from the American Heart Association. *Circulation*. 2017;135:e927–e999. doi: 10.1161/CIR.0000000000000484
17. Dallaire F, Dahdah N. New equations and a critical appraisal of coronary artery Z scores in healthy children. *J Am Soc Echocardiogr*. 2011;24:60–74. doi: 10.1016/j.echo.2010.10.004
18. Son MB, Gauvreau K, Ma L, Baker AL, Sundel RP, Fulton DR, Newburger JW. Treatment of Kawasaki disease: analysis of 27 US pediatric hospitals from 2001 to 2006. *Pediatrics*. 2009;124:1–8. doi: 10.1542/peds.2008-0730
19. Ogata S, Tremoulet AH, Sato Y, Ueda K, Shimizu C, Sun X, Jain S, Silverstein L, Baker AL, Tanaka N, et al. Coronary artery outcomes among children with Kawasaki disease in the United States and Japan. *Int J Cardiol*. 2013;168:3825–3828. doi: 10.1016/j.ijcard.2013.06.027
20. Muta H, Ishii M, Sakaue T, Egami K, Furui J, Sugahara Y, Akagi T, Nakamura Y, Yanagawa H, Matsuishi T. Older age is a risk factor for the development of cardiovascular sequelae in Kawasaki disease. *Pediatrics*. 2004;114:751–754. doi: 10.1542/peds.2003-0118-F
21. Song D, Yeo Y, Ha K, Jang G, Lee J, Lee K, Son C, Lee J. Risk factors for Kawasaki disease-associated coronary abnormalities differ depending on age. *Eur J Pediatr*. 2009;168:1315–1321. doi: 10.1007/s00431-009-0925-0
22. Minich LL, Sleeper LA, Atz AM, McCrindle BW, Lu M, Colan SD, Printz BF, Klein GL, Sundel RP, Takahashi M, et al; Pediatric Heart Network Investigators. Delayed diagnosis of Kawasaki disease: what are the risk factors? *Pediatrics*. 2007;120:e1434–e1440. doi: 10.1542/peds.2007-0815

23. Salgado AP, Ashouri N, Berry EK, Sun X, Jain S, Burns JC, Tremoulet AH. High risk of coronary artery aneurysms in infants younger than 6 months of age with Kawasaki disease. *J Pediatr*. 2017;185:112–116.e1. doi: 10.1016/j.jpeds.2017.03.025
24. Newburger JW, Takahashi M, Burns JC. Kawasaki disease. *J Am Coll Cardiol*. 2016;67:1738–1749. doi: 10.1016/j.jacc.2015.12.073
25. Shimizu C, Oharaseki T, Takahashi K, Kottek A, Franco A, Burns JC. The role of TGF-beta and myofibroblasts in the arteritis of Kawasaki disease. *Hum Pathol*. 2013;44:189–198. doi: 10.1016/j.humpath.2012.05.004
26. Kato H, Sugimura T, Akagi T, Sato N, Hashino K, Maeno Y, Kazue T, Eto G, Yamakawa R. Long-term consequences of Kawasaki disease: a 10- to 21-year follow-up study of 594 patients. *Circulation*. 1996;94:1379–1385. doi: 10.1161/01.CIR.94.6.1379
27. Dionne A, Ibrahim R, Gebhard C, Benovoy M, Leye M, Dery J, Lapierre C, Girard P, Fournier A, Dahdah N. Difference between persistent aneurysm, regressed aneurysm, and coronary dilation in Kawasaki disease: an optical coherence tomography study. *Can J Cardiol*. 2018;34:1120–1128. doi: 10.1016/j.cjca.2018.05.021
28. McCrindle BW, Manlhiot C, Newburger JW, Harahsheh AS, Giglia TM, Dallaire F, Friedman K, Low T, Runeckles K, Mathew M, et al; International Kawasaki Disease Registry. Medium-term complications associated with coronary artery aneurysms after Kawasaki disease: a study from the International Kawasaki Disease Registry. *J Am Heart Assoc*. 2020;9:e016440. doi: 10.1161/JAHA.119.016440
29. Daniels LB, Gordon JB, Burns JC. Kawasaki disease: late cardiovascular sequelae. *Curr Opin Cardiol*. 2012;27:572–577. doi: 10.1097/HCO.0b013e3283588f06
30. Tacke CE, Romeih S, Kuipers IM, Spijkerboer AM, Groenink M, Kuijpers TW. Evaluation of cardiac function by magnetic resonance imaging during the follow-up of patients with Kawasaki disease. *Circ Cardiovasc Imaging*. 2013;6:67–73. doi: 10.1161/CIRCIMAGING.112.976969
31. Dionne A, Dahdah N. Myocarditis and Kawasaki disease. *Int J Rheum Dis*. 2018;21:45–49. doi: 10.1111/1756-185X.13219
32. Akagi T, Rose V, Benson LN, Newman A, Freedom RM. Outcome of coronary artery aneurysms after Kawasaki disease. *J Pediatr*. 1992;121:689–694. doi: 10.1016/S0022-3476(05)81894-3
33. Prior M, McManus M, White P, Davidson L. Measuring the "triple aim" in transition care: a systematic review. *Pediatrics*. 2014;134:e1648–e1661. doi: 10.1542/peds.2014-1704
34. Gabriel P, McManus M, Rogers K, White P. Outcome evidence for structured pediatric to adult health care transition interventions: a systematic review. *J Pediatr*. 2017;188:263–269.e15. doi: 10.1016/j.jpeds.2017.05.066
35. Schmidt A, Ilango SM, McManus MA, Rogers KK, White PH. Outcomes of pediatric to adult health care transition interventions: an updated systematic review. *J Pediatr Nurs*. 2020;51:92–107. doi: 10.1016/j.pedn.2020.01.002
36. Got Transition. Six Core Elements of Health Care Transition. 2014. Accessed September 7, 2020. https://www.gottransition.org/six-core-elements/.
37. Mackie AS, Fournier A, Swan L, Marelli AJ, Kovacs AH. Transition and transfer from pediatric to adult congenital heart disease care in Canada: call for strategic implementation. *Can J Cardiol*. 2019;35:1640–1651. doi: 10.1016/j.cjca.2019.08.014
38. Wood DL, Sawicki GS, Miller MD, Smotherman C, Lukens-Bull K, Livingood WC, Ferris M, Kraemer DF. The Transition Readiness Assessment Questionnaire (TRAQ): its factor structure, reliability, and validity. *Acad Pediatr*. 2014;14:415–422. doi: 10.1016/j.acap.2014.03.008
39. Moynihan M, Saewyc E, Whitehouse S, Paone M, McPherson G. Assessing readiness for transition from paediatric to adult health care: revision and psychometric evaluation of the Am I ON TRAC for Adult Care questionnaire. *J Adv Nurs*. 2015;71:1324–1335. doi: 10.1111/jan.12617
40. Ferris ME, Harward DH, Bickford K, Layton JB, Ferris MT, Hogan SL, Gipson DS, McCoy LP, Hooper SR. A clinical tool to measure the components of health-care transition from pediatric care to adult care: the UNC TR(x)ANSITION scale. *Ren Fail*. 2012;34:744–753. doi: 10.3109/0886022X.2012.678171
41. Cohen SE, Hooper SR, Javalkar K, Haberman C, Fenton N, Lai H, Mahan JD, Massengill S, Kelly M, Cantu G, et al. Self-management and transition readiness assessment: concurrent, predictive and discriminant validation of the STARx questionnaire. *J Pediatr Nurs*. 2015;30:668–676. doi: 10.1016/j.pedn.2015.05.006
42. Nazareth M, Hart L, Ferris M, Rak E, Hooper S, van Tilburg MAL. A parental report of youth transition readiness: the parent STARx questionnaire (STARx-P) and re-evaluation of the STARx child report. *J Pediatr Nurs*. 2018;38:122–126. doi: 10.1016/j.pedn.2017.08.033
43. Klassen AF, Grant C, Barr R, Brill H, Kraus de Camargo O, Ronen GM, Samaan MC, Mondal T, Cano SJ, Schlatman A, et al. Development and validation of a generic scale for use in transition programmes to measure self-management skills in adolescents with chronic health conditions: the TRANSITION-Q. *Child Care Health Dev*. 2015;41:547–558. doi: 10.1111/cch.12207
44. Wiemann CM, Hergenroeder AC, Bartley KA, Sanchez-Fournier B, Hilliard ME, Warren LJ, Graham SC. Integrating an EMR-based transition planning tool for CYSHCN at a children's hospital: a quality improvement project to increase provider use and satisfaction. *J Pediatr Nurs*. 2015;30:776–787. doi: 10.1016/j.pedn.2015.05.024
45. Betz CL. California healthy and ready to work transition health care guide: developmental guidelines for teaching health care self-care skills to children. *Issues Compr Pediatr Nurs*. 2000;23:203–244. doi: 10.1080/014608600300029867
46. Got Transition. Transitioning Youth to an Adult Health Care Clinician. 2014. Accessed September 7, 2020. https://www.gottransition.org/six-core-elements/transitioning-youth-to-adult/transition-readiness.cfm.
47. Got Transition. Incorporating Health Care Transition Services Into Preventive Care for Adolescents and Young Adults: A Tool Kit for Clinicians. 2018. Accessed September 7, 2020. https://www.gottransition.org/resource/?clinician-toolkit-preventive-care.
48. American College of Cardiology Releases CHD Transition Tools (ACC website). 2016. Accessed September 29, 2020. https://www.acc.org/membership/sections-and-councils/adult-congenital-and-pediatric-cardiology-section/section-updates/2016/07/21/16/34/acc-releases-chd.
49. Sable CF, Uzark E, Bjornsen K, Canobbio K, Connolly MM, Graham HM, Gurvitz TP, Kovacs MZ, Meadows A, Reid AK, et al; On behalf of the American Heart Association Congenital Heart Defects Committee of the Council on Cardiovascular Disease in the Young, Council on Cardiovascular Nursing, Council on Clinical Cardiology, and Council on Peripheral Vascular Disease. Best practices in managing transition to adulthood for adolescents with congenital heart disease: the transition process and medical and psychosocial issues: a scientific statement from the American Heart Association. *Circulation*. 2011;123:1454–1485. doi: 10.1161/CIR.0b013e3182107c56
50. Niedra E, Chahal N, Manlhiot C, Yeung RS, McCrindle BW. Atorvastatin safety in Kawasaki disease patients with coronary artery aneurysms. *Pediatr Cardiol*. 2014;35:89–92. doi: 10.1007/s00246-013-0746-9
51. Tremoulet AH, Jain S, Jone PN, Best BM, Duxbury EH, Franco A, Printz B, Dominguez SR, Heizer H, Anderson MS, et al. Phase I/IIa trial of atorvastatin in patients with acute Kawasaki disease with coronary artery aneurysm. *J Pediatr*. 2019;215:107–117.e12. doi: 10.1016/j.jpeds.2019.07.064
52. Huang SM, Weng KP, Chang JS, Lee WY, Huang SH, Hsieh KS. Effects of statin therapy in children complicated with coronary arterial abnormality late after Kawasaki disease: a pilot study. *Circ J*. 2008;72:1583–1587. doi: 10.1253/circj.CJ-08-0121
53. Geovanini GR, Libby P. Atherosclerosis and inflammation: overview and updates. *Clin Sci (Lond)*. 2018;132:1243–1252. doi: 10.1042/CS20180306
54. Denby KJ, Clark DE, Markham LW. Management of Kawasaki disease in adults. *Heart*. 2017;103:1760–1769. doi: 10.1136/heartjnl-2017-311774
55. Gordon JB, Daniels LB, Kahn AM, Jimenez-Fernandez S, Vejar M, Numano F, Burns JC. The spectrum of cardiovascular lesions requiring intervention in adults after Kawasaki disease. *JACC Cardiovasc Interv*. 2016;9:687–696. doi: 10.1016/j.jcin.2015.12.011
56. Blumenthal D. Part 1: quality of care–what is it? *N Engl J Med*. 1996;335:891–894. doi: 10.1056/NEJM199609193351213
57. Institute of Medicine (US), Committee on Quality of Health Care in America. *Crossing the Quality Chasm: A New Health System for the 21st Century*. Washington, DC: National Academies Press; 2001.
58. Marelli A. The future of adult congenital heart disease research: precision health services delivery for the next decade. *Can J Cardiol*. 2019;35:1609–1619. doi: 10.1016/j.cjca.2019.09.015
59. Gurvitz M, Marelli A, Mangione-Smith R, Jenkins K. Building quality indicators to improve care for adults with congenital heart disease. *J Am Coll Cardiol*. 2013;62:2244–2253. doi: 10.1016/j.jacc.2013.07.099
60. Bhavnani SP, Parakh K, Atreja A, Druz R, Graham GN, Hayek SS, Krumholz HM, Maddox TM, Majmudar MD, Rumsfeld JS, et al. 2017 Roadmap for innovation-ACC health policy statement on healthcare transformation in the era of digital health, big data, and precision

health: a report of the American College of Cardiology Task Force on Health Policy Statements and Systems of Care. *J Am Coll Cardiol*. 2017;70:2696–2718. doi: 10.1016/j.jacc.2017.10.018

61. Huang JS, Terrones L, Tompane T, Dillon L, Pian M, Gottschalk M, Norman GJ, Bartholomew LK. Preparing adolescents with chronic disease for transition to adult care: a technology program. *Pediatrics*. 2014;133:e1639–e1646. doi: 10.1542/peds.2013-2830
62. Marelli A. Trajectories of care in congenital heart disease—the long arm of disease in the womb. *J Intern Med*. 2020;288:390–399. doi: 10.1111/joim.13048