Received: 11 May 2017 | Accepted: 20 May 2017
DOI: 10.1111/chd.12500

SPECIAL SECTION ON CORONARY ANOMALIES

WILEY Congenital Heart Disease

Catheter-based anatomic and functional assessment of coronary arteries in anomalous aortic origin of a coronary artery, myocardial bridges and Kawasaki disease

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Abstract

Most diagnostic testing in patients with anomalous aortic origins of coronary arteries, myocardial bridges, and coronary artery changes after Kawasaki disease are performed with the use of noninvasive techniques. In some cases, however, further diagnostic information is needed to guide the clinician in treating these patients. In such instances, cardiac catheterization with invasive anatomic and functional testing is an invaluable tool. Moreover, interventional treatment in the cardiac catheterization laboratory may be performed in a small subset of these patients. As the diagnosis of these conditions is now becoming more common, it is important for pediatric interventional cardiologists to be familiar with these techniques. In this article, the role of angiography, intravascular ultrasound, fractional flow reserve, and optical coherence tomography in these patients is reviewed.

KEYWORDS
anomalous aortic origin of a coronary artery, cardiac catheterization, Kawasaki disease, myocardial bridges

1 | INTRODUCTION

Though many decisions regarding the management of patients with anomalous aortic origin of a coronary artery (AAOCA), myocardial bridges, and Kawasaki disease are made based on symptoms and noninvasive modalities discussed elsewhere in this article, cardiac catheterization plays a vital role in cases where there is a need for more information to guide clinicians in their decision making. Additionally, some patients can potentially be treated in the catheterization laboratory as well based on the specifics of the lesion. In this review, we will discuss some of the basic principles involved in the anatomic and functional assessment of these lesions in the cardiac catheterization laboratory.

2 | GENERAL PRINCIPLES AND CORONARY ANGIOGRAPHY

In small children, coronary angiography is performed with general anesthesia. However conscious sedation is used in older children and adults. Patients are anticoagulated adequately throughout the procedure. It is generally advisable to perform an aortic root angiogram before selective coronary angiography is to be performed in these patients. The reason for this is twofold. First, an aortic root angiogram will define the origin of the coronary arteries and guide the interventional cardiologist in appropriate catheter selection. Second, in the event of severe ostial compromise, it may not be recommended to engage the ostium of the coronary artery due to hemodynamic compromise that can be encountered. When advancing coronary wires or intravascular ultrasound (IVUS), it is prudent to leave the guiding catheter outside the coronary ostium in patients with severe ostial compromise. We start with using standard coronary catheters (3.3 Fr-5 Fr) to perform coronary angiograms but for other anatomic and functional assessment described below, the use of 5–6 Fr guiding catheters is needed. Prior to advancing coronary wires or IVUS catheters inside coronary arteries, 1 μg/kg of intracoronary nitroglycerin is administered (adult dose 100–200 μg) to prevent coronary artery spasm. In children, we generally have used 5–6 Fr sheaths and catheters to facilitate these procedures in patients weighing 25 kg or more and using these modalities, routine surveillance in children more than 7 years of age can safely be performed.[1] However, a number of factors dictate the use of 5–6 Fr sheaths and catheters in children. The use of 5–6 Fr sheaths may not be possible in the presence of significant femoral artery stenoses (from prior catheterizations or arterial line placements). Similarly, the use of these guiding catheters may not be recommended if the coronary arteries are small. Conversely, if clinically indicated, the use of 5–6 Fr sheaths and catheters in children less than 25 kg can be used to facilitate anatomic and functional assessment of coronary arteries.

In the case of AAOCA, the catheter curve required is dictated by the sinus from which the anomalous coronary artery arises from, for example, in the setting of an anomalous left coronary artery from the right coronary cusp, Judkins Right or Amplatz Right coronary catheters or guiding catheters will most likely engage the anomalous coronary artery. Angiograms in the respective aortic cusp prove helpful in providing detail of the ostium in anomalous coronary arteries with ostial narrowing. In many patients, an interarterial course between the aorta and pulmonary artery can be readily seen with angiograms.[2] If standard catheter curves are not successful in engaging the coronary artery, catheters can be shaped with heat to provide an appropriate curve. The classic angiographic "milking effect" sign is seen in patients with myocardial bridges when the luminal diameter of the effected segment is reduced by 70% or more in systole and there is a persistence of 35% or more narrowing in mid to late diastole.[3] This effect is more prominent with the administration of nitroglycerin which vasodilates the noninvolved coronary artery segments.[3] Angiography in patients with Kawasaki disease with large or complex coronary artery aneurysms can be beneficial and is generally recommended 6–12 months after the acute illness, unless clinically indicated sooner.[4] Long-term follow-up angiography is useful as guided by symptoms, noninvasive imaging and if a catheter-based intervention is to be performed.[4] These studies help identify the extent of aneurysms, angiographic narrowing (Figure 1) around aneurysms and calcified areas.

3 | INTRAVASCULAR ULTRASOUND

The role of IVUS in the assessment of AAOCA is controversial. In our experience, IVUS may be useful in cases where there is a need to clearly define intralumenal or intramyocardial compromise in the absence of clear cardiac symptoms. Angelini and colleagues have shown that IVUS can evaluate stenosis of the intramural segment in right AAOCA in detail, which correlated with symptoms in their experience.[5] In the pediatric population in patients with right AAOCA, this poses clear challenges where symptoms are often vague and ill defined (an operation is indicated in left AAOCA). Nevertheless, it is plausible that in a certain small subset of patients, this modality may be useful in risk stratifying patients and also useful in some patients postoperatively, if clinically indicated. In patients with myocardial bridges, IVUS can show the "half moon phenomenon," which is seen between the epicardial tissue and segment of coronary bridging, but spares the normal coronary artery proximal and distal to the lesion.[6] In addition, IVUS demonstrates that bridging coronary segments are often spared from atherosclerotic lesions presumably due to altered flow characteristics, with lower flow in the effected segments resulting in less endothelial injury.[6] Long-term vascular changes with IVUS continue to be observed in patients with Kawasaki disease and regressed coronary artery aneurysms.[7] Virtual histology IVUS (VH-IVUS) has recently been employed in patients with a history of Kawasaki disease and has helped define atherosclerotic changes in effected coronary artery segments.[8] Additional experience suggests that there is a role for IVUS in the follow-up of these patients and identification of patients in need for intervention.[9]

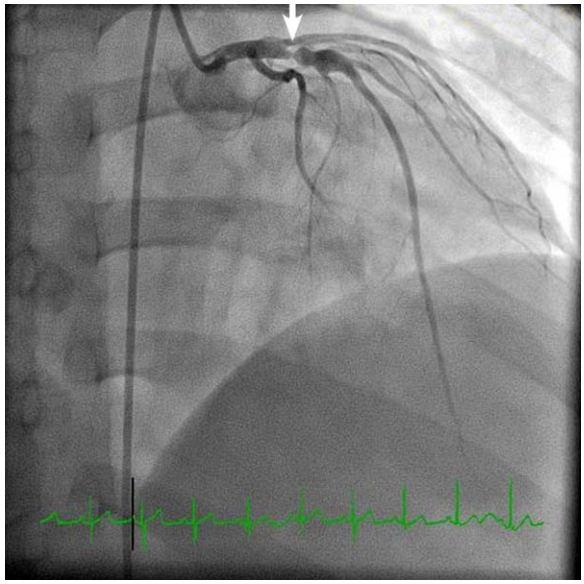

FIGURE 1 Left coronary artery selective angiogram in a 9-year-old with a history of Kawasaki disease showing severe stenosis (arrow) in the left anterior descending coronary artery (LAD) proximal to an aneurysm

4 | FRACTIONAL FLOW RESERVE

Pressure flow wires can estimate the fraction of normal flow in the presence of significant coronary artery stenosis. This is done by using the ratio of the coronary pressure distal to the stenosis to the aortic pressure, and is performed at maximal hyperemia (eg, with adenosine administration), averaged over the cardiac cycle.[10,11] Based on discussions from the Symposium on Coronary Artery Anomalies, 2014, we will also be using dobutamine when performing FFR, in order to mimic exercise physiology. A normal fractional flow reserve (FFR) is 1.0 and significant stenosis is indicated by an FFR of $<$0.75–0.80.[10,11] As of December 2014, we have used FFR thus far in one patent with an anomalous coronary artery origin and a long intramyocardial course, when the decision making was difficult. In patients with myocardial bridges, clinical ischemia is suggested by an FFR of less than 0.75.[3] We and others[12] have used FFR now in our decision making regarding catheter-based intervention and surgery in children with Kawasaki disease (Figures 1 and 2) Similar to the adult experience, an FFR of $<$0.75 has been found to be clinically indicative of myocardial ischemia.[12]

5 | OPTICAL COHERENCE TOMOGRAPHY

Optical coherence tomography (OCT) is a relatively new technology with significantly more resolution than IVUS at the expense of

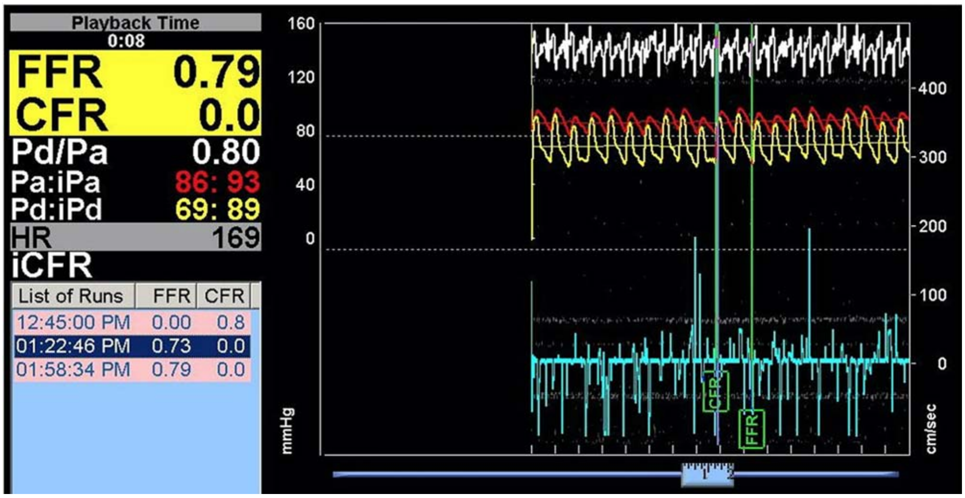

FIGURE 2 Fractional flow reserve (FFR) tracing in the same patient with adenosine infusion, showing an FFR indicative of significant stenosis (0.79 in this case) of the LAD

providing less depth.[11] It does have certain limitations requiring clearing/displacing blood from the area of interest. This may create challenges in the case of AAOCA due to the ostial nature of the lesion which is often severe.[5] Like IVUS, OCT may demonstrate important characteristics of myocardial bridges that are not readily identified by standard coronary artery angiography.[13] Similarly, OCT has recently been show to identify coronary artery changes in children with Kawasaki disease not apparent by standard coronary artery angiography.[14] Importantly, in children, this new OCT technology has been shown to be feasible in 5 French coronary artery guide catheter systems.[14]

6 | COMPLICATIONS

Complications of the catheter-based diagnostic modalities discussed above are rare.[15–17] Coronary dissection is rare using IVUS, occurring in less than 0.5% procedures,[15] and our experience in children is similar. Coronary spasm may occur but can be treated with intracoronary nitroglycerin.

7 | CONCLUSIONS

With the increasing diagnosis and treatment of coronary lesions discussed here, pediatric/congenital interventional cardiologists will be faced with more and more situations in the future requiring the use of these specialized modalities. These procedures should only be performed by interventional cardiologists familiar with these modalities. Partnering with our adult interventional cardiology colleagues is important to catering to the needs of this unique patient population. Opportunities still exist for further miniaturization of the technologies discussed above for small children.

CONFLICT OF INTEREST

None.

DISCLOSURES

None.

AUTHOR CONTRIBUTIONS

The author contributed in the concept/design, drafting, critical revision and approval of the article.

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How to cite this article: Qureshi AM, Agrawal H. Catheter-based anatomic and functional assessment of coronary arteries in anomalous aortic origin of a coronary artery, myocardial bridges and Kawasaki disease. *Congenital Heart Disease*. 2017;12:615–618. https://doi.org/10.1111/chd.12500