Optimal Use of Cardiology Referrals and Cardiac Studies

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This continuing medical education program includes:

- Statement of learning objectives
- Pretest questions and answers
- Audio recording and printed transcript of panel discussion
- Post-test questions
- Post-test response form
- List of recommended supplementary reading materials
Learning Objectives

This enduring material is designed for pediatricians, nurse practitioners and other allied health professionals who care for children. The intended result of this activity is to achieve a change in competence, performance, and patient care. After completing this activity, the participant should be better able to:

1. Discuss the indications for referral of a patient to a pediatric cardiologist.
2. List those cardiac studies which can be performed independently of the input of a pediatric cardiologist.
3. List those cardiac studies which should be ordered by a pediatric cardiologist in conjunction with the cardiologist’s evaluation.
4. Describe the indications for electrocardiography.
5. List the limitations associated with electrocardiography.
6. List the cardiac diagnoses typically managed by general pediatricians.

Pretest Questions

Before listening to the audio CD or examining the transcript of the panel discussion, you will find it useful to test your general understanding of the subject of this program with this brief introductory quiz. (The correct answers can be found on page 9 of this booklet.)

1. Patients diagnosed with cardiomyopathy also routinely need which of the following evaluations performed?
   - [ ] a. Tilt-table testing
   - [ ] b. Evaluation of first-degree relatives for evidence of cardiomyopathy
   - [ ] c. Nuclear medicine stress testing
   - [ ] d. Cardiac catheterization

2. Clinical signs of endocarditis include fever, a new or changing murmur, and splenomegaly.
   - [ ] True
   - [ ] False

3. Channelopathies, or cardiac conditions that include abnormal electrical depolarization and repolarization, DO NOT include:
   - [ ] a. Wolff-Parkinson-White syndrome
   - [ ] b. Long QT syndrome
   - [ ] c. catecholaminergic polymorphic ventricular tachycardia
   - [ ] d. Brugada syndrome

4. The cutoff limit for a normal QTc is a duration of 0.44 seconds.
   - [ ] True
   - [ ] False

5. Echocardiography can confirm the diagnosis of congestive heart failure.
   - [ ] True
   - [ ] False

6. A diagnosis often referred to cardiologists that can typically be evaluated and managed by general pediatricians is:
   - [ ] a. chest pain
   - [ ] b. Kawasaki disease
   - [ ] c. hypertrophic cardiomyopathy
   - [ ] d. rheumatic heart disease

This course can be completed by listening to the audio CD and by reading the transcript. A multiple-choice test must be completed to earn CME credits. In order to receive credit, a participant must have a designated passing grade of 75%. The estimated time to complete this activity is 3 hours.
Transcript of the Panel Discussion
(As recorded in the audio CD)

● DR. JEFFREY R. BORIS*: I'm Jeffrey Boris. I am a Clinical Professor of Pediatrics at the Perelman School of Medicine at the University of Pennsylvania and the Children's Hospital of Philadelphia. I'm an Ambulatory Pediatric Cardiologist, and I'm the Director of the Postural Orthostatic Tachycardia Syndrome Program.

● DR. ROBERT M. CAMPBELL:* I'm Robert Campbell, Professor of Pediatrics at Emory University School of Medicine and a Pediatric Cardiologist at Children’s Healthcare of Atlanta at Shiloh Heart Center.

● DR. LEO LOPEZ: I'm Leo Lopez, Professor of Clinical Pediatrics at the Herbert Wertheim College of Medicine at Florida International University and Associate Chief of Cardiovascular Medicine and Director of Noninvasive Cardiac Imaging at Nicklaus Children’s Hospital in Miami.

● DR. BORIS: Today, we're going to discuss “Optimal Use of Cardiology Referrals and Cardiac Studies.” What does this mean that we're talking about? We are going to specifically concentrate on patients who are seen in the outpatient clinic and in the emergency department. We're going to talk about why patients should be referred to a cardiologist, what tests the cardiologist can use; as well as reasons to get those tests and to not get those tests.

The first thing that we'd like to discuss are some issues where primary care providers, we would hope, would manage these issues by themselves. So, Leo, what kind of cardiac diagnoses do we think that primary care providers routinely manage?

● DR. LOPEZ: I'm assuming you're talking about signs and symptoms that a primary care provider might encounter that possibly warrant a cardiac evaluation. These would include a murmur, chest pain, syncope, or dizziness, palpitations. There are also other reasons why a primary care provider might consider a cardiology evaluation. For example, if an athlete needs a preparticipation evaluation, if they're using stimulant therapy for attention-deficit/hyperactivity disorder, if there are lipid issues, if the patient has hypertension.

● DR. BORIS: Robert, can you comment specifically on the referral of patients for stimulants? Because I know some data came out that talked about that within the last few years.

● DR. CAMPBELL: The stimulant issue has really raised a lot of confusion out in the general pediatric population. They're widely used medications, and there was a report out of Canada many years ago. Their equivalent of the FDA [Food and Drug Administration] issued a “Black box warning” that raised the specter of these drugs potentially contributing to sudden death in children that were receiving these medications. The concern was raised that all these patients should have pretreatment screening ECGs [electrocardiograms] and then follow-up ECGs to manage them while they're on the medications. The real problem is that the ECG doesn't show any sensitive or specific marker for risk or lack thereof, and therefore, it's not really a very good screening tool. There really wasn't any data to support that final conclusion.

● DR. BORIS: I know that there was an article that came out in the New England Journal of Medicine several years ago that was a very large population-based study that demonstrated that there was no increased risk of sudden cardiac death associated with methylphenidate. The American Academy of Pediatrics did come out with a statement saying that prestimulant ECGs and continued ECG monitoring was not indicated for patients on stimulants who didn't have any kind of specific history or other cardiac findings.

I do want to talk about hyperlipidemia and hypertension a little bit. Before I came to the Children's Hospital of Philadelphia, I was in private practice in western North Carolina. The primary care providers, the pediatricians and family practitioners, would do their best to talk with the patients and families about things like obesity and overweight and the parents got very indignant about being told that their child had this problem. So, what ended up happening was the primary care providers started referring these patients to us as cardiologists. Robert, do you have any comments on it?

● DR. CAMPBELL: One of the important parts is family history. If there's a predisposition to hyperlipidemia or to hypertension, that patient needs very close follow up and management. The most important reason for a hypertensive pediatric patient to be sent to a cardiologist, from my standpoint, is to make certain that we're ruling out coarctation. The treatment for that is a completely different treatment than for isolated essential hypertension. I think it's very rare that we actually see patients with more malignant hypertension that might be secondary to some other cause.

● DR. BORIS: It was just interesting that these families being sent to the cardiologist, all of a sudden, they really paid attention. I'm not recommending that this be done. It is interesting. There's a difference between adult internal medicine providers and pediatricians where a lot of adult primary care providers will manage the hypertension and hyperlipidemia. I think this is an evolving trend for pediatrics.

● DR. LOPEZ: I do think it's important that a primary care provider recognizes their limitations in terms of dealing with some of these issues like hyperlipidemia and hypertension. So, there are times when I think those referrals are appropriate, particularly if there's some discomfort with managing the problem itself.

● DR. BORIS: certainty, if things are going beyond diet and exercise interventions and they need to start thinking about using medications or if there's a strong familial history of very elevated lipids, I think that makes a lot of sense.

● DR. LOPEZ: Yes, I agree.

● DR. BORIS: Let's talk about other reasons why patients should be referred to a cardiologist. I think the most typical thing that we would expect is evaluation of congenital or acquired cardiac disease. Leo, do you want to talk about some other things that you would think would be appropriate for referral?

● DR. LOPEZ: Sure. Certainly, patients that have been diagnosed with Kawasaki disease warrant cardiology follow-up.

1. See Cooper et al (2) in List of Supplementary Reading Materials.
2. See Perlin et al (5) in List of Supplementary Reading Materials.
3. No financial interest or other relationship to disclose.
4. No financial interest or other relationship to disclose.
5. No financial interest or other relationship to disclose.
up because of the cardiovascular manifestations of Kawasaki disease. Anybody who has been diagnosed with pulmonary hypertension would warrant follow-up. Anybody with known cardiomyopathy or some problem with the heart muscle should have cardiology evaluation. One of the things to consider for these patients in particular is that family members also need to be seen by the cardiologist because of the genetic component to this disease.

DR. BORIS: Especially first-degree relatives. There’s a much higher incidence of these amongst first-degree relatives. Sometimes the pediatrician will have something that does not make sense to them or that they’re not used to. I think it goes back to Leo’s comment about comfort level. So, if you have a murmur or chest pain, dizziness, those kind of things, that should be some other factors associated with them, that are concerning or worrisome or “don’t meet criteria for normality,” then that should be considered. So I guess that means that the primary care providers need to know what “normal” is. Robert, do you want to stand up because of the cardiovascular manifestations of Kawasaki disease. Anybody who has been diagnosed with pulmonary hypertension would warrant follow-up. Anybody with known cardiomyopathy or some problem with the heart muscle should have cardiology evaluation. One of the things to consider for these patients in particular is that family members also need to be seen by the cardiologist because of the genetic component to this disease.

DR. BORIS: Yes. Myocarditis is fairly frequently missed, even by us cardiologists. Certainly, if the patient has a history of a congenital heart defect that’s been lost to follow-up, especially after having had surgical repair, those patients need to be seen. Back in the day, both surgeons and cardiologists would say to these patients, “You’re fixed.” What we have found is over the long haul there are so many different features, both from a neurocognitive standpoint, from an arrhythmia standpoint, from a cardiac function standpoint, from a

DR. CAMPBELL: Those are really worrisome diagnoses. Pericarditis and myocarditis can be very subtle onset, child who has very minor flu-like illness but doesn’t seem to really get better. There may be other affected family members who are now recovering and yet this child is not or may actually be getting worse. There should be a very high level of suspicion for that patient. The presence of chest pain, new onset chest pain or persistent chest pain, sometimes positional, worse lying down than sitting up, and the presence of fever should raise the question of peri-carditis. There’s a very scary diagnosis because those patients can be well at 8:00 in the morning, sick at 10:00, and dying by 1:00. So it’s really important to know how to sort out and how to sort out those kind of things. Frequently, patients will come into the emergency room or into the clinic with a history of chest pain plus a friction rub or those kind of things, that might suggest heart disease?

DR. BORIS: Absolutely. Now, if a patient has known heart disease, there are certain clinical features that would be a signal that patients should be evaluated by their cardiologist. If the patient has a fever with a new and/or changing murmur and with or without splenomegaly, that would be potentially suggestive of endocarditis. A patient may have a fever and/or respiratory illness and/or a GI [gastrointestinal] illness and the chest pain plus a friction rub or those same kind of things, fever, respiratory illness, GI illness, persistent chest pain, and then they have a gallop or a new murmur or an arrhythmia. Those are suggestive of pericarditis or myocarditis. Robert, do you want to talk about some of those aspects of disease?

DR. CAMPBELL: Right. An adult, if they’re working and had exercise-related chest pain, we’d immediately think of heart attack or angina. A newborn baby or a child several months old, their work really is around eating. So, if they’re spending a lot of energy, they’re working hard to eat, that may be a time when they might suffer myocardial ischemia or angina-type symptoms, and that might manifest as crying. Just this child who’s very colicky during feeds and then may also look very sweaty or gray or poorly perfused. That should always raise suspicion.

DR. BORIS: The classic history that I would always hear would be that the baby would be really hungry, want to eat, start eating, cry like crazy, and really seem to be in pain. As soon as the baby pulled off the nipple and calmed down, the crying went away, but the baby was hungry and wanted to eat again; right? Leo, do you want to talk about some other clinical features that might suggest heart disease?

DR. LOPEZ: Sure. If a patient presents with tachycardia or, more specifically, central cyanosis, where the parents might see a bluish tinge to the lips or to the mucous membranes certainly would warrant a cardiology evaluation. Patients with exercise intolerance would warrant a cardiology evaluation. Chest pain is one of the symptoms that we see fairly frequently, but usually it’s chest pain that’s associated with exercise or that’s associated with diaphoresis and radiation of the pain to the arm. It’s chest pain that’s so severe that the patient starts to cry. Then, of course, syncope is another symptom that will often warrant evaluation, particularly if it’s associated with exercise.

DR. BORIS: Yes. Myocarditis is fairly frequently missed, even by us cardiologists. Certainly, if the patient has a history of a congenital heart defect that’s been lost to follow-up, especially after having had surgical repair, those patients need to be seen. Back in the day, both surgeons and cardiologists would say to these patients, “You’re fixed.” What we have found is over the long haul there are so many different features, both from a neurocognitive standpoint, from an arrhythmia standpoint, from a cardiac function standpoint, from a
coronary artery disease standpoint that say to us that fixed is not fixed. So these patients definitely need to show back up and be seen.

**DR. BORIS:** It’s actually not an uncommon scenario in the emergency room for a patient to show up with a scar on their chest and have the family not mention the fact that heart surgery was performed in the past because they’re not aware of the fact that there may be potential problems with their heart in the future.

**DR. BORIS:** Some clinical physical findings that would suggest heart disease would include a harsh murmur that’s not vibratory. So when we talk about a vibratory murmur, it sounds sort of like this [makes buzzing sound]. That’s classic for a Still’s murmur. If you don’t have that but you have a really harsh-sounding murmur, that would be something that would suggest that the patient has a cardiac defect. Other things include a diastolic murmur; if the patient has an active precordium or a thrill, the patient has a click, which can be really hard to diagnose, their femoral pulses are decreased, or they have lag between their right radial and their femoral pulse; if the second heart sound is single or widely split; if there’s a gallop, a whoop or a friction rub. Robert, do you want to talk about some of your experience in having some of these patients with varied clinical findings show up?

**DR. CAMPBELL:** Again, I think the pediatricians are actually much better, much more competent with these diagnoses than they give themselves credit for. I think that they should trust their judgment. When they think it’s not normal or when they’re suspicious that it’s not normal, that’s a good indication for referral. I always recommend that they carry their differential diagnosis to the highest level of resolution that they can at that point in time and then use the conversation back from the consultants to help expand that area of normal or to confirm that this was an abnormal finding. It’s continuing education for them to have that referral.

**DR. BORIS:** Which means that they need to know, again, the definition of what normal is. Sometimes you have to go back and practice. I’ve had people ask me if they could hang out in clinic with me just to repeat their exposure and experience in learning physical exams. So, for example, when I’m teaching providers about innocent murmurs, I tell them, “You don’t need to tell me that this patient has a VSD [ventricular septal defect] murmur. What I need you to do is tell me that the patient has a normal or an abnormal exam, and if the patient’s abnormal, are they in heart failure?” So to know normal means, for example, for murmurs you need to know the six innocent murmurs cold and those definitions. But beyond that, you’re good.

**DR. CAMPBELL:** I think one of the important things to remember is that most family members don’t know what a murmur is. They don’t understand that the murmur is simply the noise of turbulence. What many of them will believe is murmur indicates the presence of a heart problem, it means there’s something wrong, there’s something dangerous going on. Therefore, there’s an incredible level of anxiety. One of the most important parts of a murmur evaluation in our cardiology office would be to sit down and explain what a murmur is, the noise of turbulence; and what a murmur isn’t, an automatic indication of heart disease. Families can leave very confused if you don’t take the time to explain what a murmur is. Is it that they come in believing that murmur means there’s something wrong, and you listen and say it’s normal, everything’s okay, when they leave the office, they will be incredibly confused.

**DR. BORIS:** Correct. I take that same approach as well when I’m counseling patients and families. It’s interesting what families will hear because, despite the fact that you counsel them, they’re still going to walk out and say, if the child has a ventricular septal defect, they may say the kid has a VSD. But they might just as easily walk out and say, “My child has a murmur” versus patients who have an innocent murmur, they may still say, “He’s got a murmur, but I don’t know what that means.” I think there’s definitely still a disconnect. I think that comes from fear and uncertainty.

**DR. LOPEZ:** Whenever I encounter a patient with an innocent murmur, I always tell them that I have a murmur that comes out when I have a fever or when I exercise. I think understanding that people who have normal cardiac anatomy can actually have that and actually speaking to somebody with the same thing helps.

**DR. BORIS:** It’s funny, I do the same thing when patients have the sharp, brief chest pain that’s worsened with inspiration, especially adolescents, which is typical chest wall pain. I tell them, “I used to have that when I was a teenager, and it went away when I got to be an adult. So it’s a big nuisance. Don’t worry about it.” Leo, do you want to talk about some other signs that suggest heart-related disorders?

**DR. LOPEZ:** Absolutely. Also, rheumatic fever is still with us, despite the fact that we live in 2017. It used to be a lot more common, but we still see it. What are the signs of rheumatic fever?

**DR. CAMPBELL:** Rheumatic fever, as the name says, requires the presence of a fever. The cardiology manifestations may be a new murmur, one that is innocent. Characteristics of mitral insufficiency or aortic insufficiency, a diastolic murmur. Then there are the other problems with neurologic findings, migratory arthralgia, rash. It’s uncommon enough now that I think many of us are uncomfortable or maybe incompetent with the diagnosis, honestly. You just have to have a high level of suspicion.

**DR. BORIS:** I think that rheumatic fever can diagnose itself as well. Just a couple of months ago, I had a patient who was truly diagnosed with Sydenham’s chorea but had nothing else. I got an ASO [antistreptolysin O] level on her. It was elevated. With a little bit of salbutamol therapy—I couldn’t use penicillin, she was allergic—her chorea came down. So you do have to, again, know the concepts of normal but know also the criteria for these various disease processes.

**DR. BORIS:** I think it is uncommon in the United States, but it’s certainly very common in other parts of the world. So, in areas where there are people from other
parts of the world, it’s important to be aware that you may see that in patients.

**DR. CAMPBELL:** Remembering that Kawasaki disease is a childhood disease, but there are many patients who have had that missed diagnosis or rheumatic cardiac and now have the chronic manifestations with significant valvular disease.

**DR. BORIS:** Absolutely. Another heart-related disorder that we do see a fair bit is Kawasaki disease. So Kawasaki disease, to remind our colleagues, includes a fever of 102.2°F or 39°C for 5 or more days; a rash that typically includes the groin; erythema of the hands and feet; and also some edema. Then, at least in my experience, I would say actually over 90% of the patients whom I have seen with Kawasaki disease have an apical 54, if you listen closely enough. To remind our colleagues, the concern that we have with Kawasaki disease would be what kind of things.

**DR. LOPEZ:** The most worrisome component of Kawasaki disease is the development of coronary artery abnormalities and specifically the development of coronary artery aneurysms and ectasia, which then sets the patient up for future problems with thrombus formation in the coronary arteries and coronary stenoses. That’s that’s the most significant cardiovascular manifestation. Other things that you need to think about include myocarditis, which certainly occurs in many of these patients during the acute period. Of course the long-term complications that we need to worry about include coronary insufficiency.

**DR. BORIS:** The good news—if you can say there’s good news with Kawasaki disease—is since the 1980s we have really had the efficacy to bring that risk of coronary artery aneurysms from 20% to 25% down to 2% to 5% in patients. So, the large majority of patients, if caught before day 10, ostensibly, of their illness, do well.

**DR. CAMPBELL:** I think one of the other concerns is that Kawasaki disease, whatever causes it, and we still don’t know, is basically a manifestation of arteritis, of medium-sized vessels that are inflamed. That’s why you have red hands and feet, red eyes. That’s why you have coronary artery changes. My concern about this long term is do those arteries heal back completely to baseline normal or is there some microscopic level of myocardial and then coronary artery dysfunction. So long term I think some of the concerns would be a patient who is active with new-onset exercise-related complaints—exercise chest pain, exercise intolerance, exercise shortness of breath or syncope—that’s a very alarming new symptom that needs very aggressive management to make sure that it’s not a coronary cause. The other is that these patients should be very aggressively counseled and followed in terms of potential concomitant hyperlipidemia. Probably this is a group that should be managed with drug treatment or dietary therapy more aggressively than, say, someone who was not affected with Kawasaki disease.

**DR. BORIS:** There’s been a fair bit of long-term follow-up, especially in Japan, in patients who have Kawasaki disease who have been treated with IVIG [intravenous immunoglobulin] and high-dose aspirin, which is the initial therapeutic approach for these patients. These patients have gotten into their 40s and 50s, and so far, with appropriate therapy, there is not an increased incidence of coronary artery disease.

**DR. CAMPBELL:** However, there have been some very interesting studies with intravascular ultrasound and also in the cardiac catheterization laboratory with vasodilators that demonstrate that there is minimal and mild abnormalities of the coronary arteries such that they don’t dilate appropriately and they are thickened; but, we don’t know if that actually still leads to premature atherosclerosis. I’d also like to have Robert discuss issues of lethal arrhythmias, including familial arrhythmias.

**DR. CAMPBELL:** One of our newest concerns in the office is a patient who presents with potential warning signs and a family history of sudden unexplained and unexpected death at young age, and by that we mean less than 50. It would be a patient who has unusual or unexpected or very intense exercise-related chest pain or shortness of breath. Others would be a patient with unexplained seizures or syncope or if there’s a family history of anybody with coronary disease, cardiomyopathy, or early-onset pacemaker or ICD [implantable cardioverter-defibrillator] implantation. In this day and age, I think you really have to know what those genetic cardiac conditions are. They include cardiomyopathies, channelopathies, channelopathies, and arrhythmias, and those are conditions where the repolarization and repolarization of the heart, aortic diseases, pulmonary arterial hypertension. So our world has gotten much more complex, and the attention to detail of family history, especially multigenerational, is now more important than ever.

**DR. BORIS:** With the channelopathies, to expand on these a little bit more, that includes long QT syndrome, short QT syndrome, catecholaminergic polymorphic ventricular tachycardia (CPVT), and Brugada syndrome. Some other family history I think that would be appropriate to look out for is if those entities that Robert just discussed occur in first-degree relatives and, we discussed earlier in our program, if there have been some bizarre deaths—that the patient was a good swimmer and drowned; or the patient “fell asleep” while driving, or the alarm clock went off, and the patient died. One of the things that I was always taught was, if the patient had syncope, these patients drop. So unlike typical syncope where they are dizzy and they may be able to let themselves down or maybe not, these patients face plant.

**DR. LOPEZ:** There are some other arrhythmias I think we need to keep in mind as well. Leo, do you want to talk about those?

**DR. BORIS:** Absolutely. Another heart-related disorder that we do see a fair bit is catecholaminergic polymorphic ventricular tachycardia. Along with heart rates less than 35 would warrant a cardiology evaluation, particularly if the patient is symptomatic.

**DR. CAMPBELL:** We’re going to switch gears a little bit now and talk about some of the cardiac tests that can help with diagnosis. These are tests that we use on a routine basis. They include electrocardiograms, chest radiograms, echocardiograms, Holter monitors, stress test, exercise stress testing. But besides electrocardiography, the three of us have discussed this prior to this discussion, and we feel that it’s probably not appropriate for primary care providers to be ordering these other tests in the absence of cardiac involve. I think that internal medicine providers and family practitioners order echocardiograms on their adult patients on a routine basis. Robert, I’d like you to talk about your thoughts on this, too.
DR. CAMPBELL: I think the important part is to understand the indication for the test and also how you're going to explain the results to the family. Many times, a normal echocardiogram and the family history of heart disease, which is nonspecific, the echocardiogram may be the least meaningful test in terms of trying to sort if whether the child is likewise affected. So I think that understanding the details around the indication for the test and then owning the patient discussion, the patient management after the test results are available, is all part of the process. I think another is that in my experience, pediatricians are really very good with murmur evaluation, and ordering an echocardiogram for murmur screening is probably a very cost- and procedure-ineffective approach. If a pediatrician has a patient who has a hyperactive precordium, who's cyanotic, who has femoral pulses that are decreased and is having exercise syncope, the first thing they do is refer to cardiology, which is an appropriate response.

DR. BORIS: So let's talk about the first test that we mentioned, and that's electrocardiogram. I think it's probably the most commonly ordered test by nonpediatric cardiologists. Leo, do you want to talk about ECGs a little bit?

DR. LOPEZ: Sure. An electrocardiogram, just to review for our colleagues, involves a 12- or a 15-lead study that's obtained over 2.5 seconds, usually includes a rhythm strip that's preferably the lead II rhythm strip. The electrocardiogram is good at evaluating heart rate, evaluating the rhythm of the heart. It looks for abnormalities such as premature beats, either premature atrial or premature ventricular beats. You're able to look at abnormalities like prolongation of the QT interval. It's also good at giving us a warning of chamber enlargement in terms of rhythm abnormalities. Things like sinus node dysfunction will be seen on an electrocardiogram. It also allows us to determine where the sinus node is or where the pacemaker of the heart is. One of the things that's important when we're talking about electrocardiograms is that, in order for us to know that there is an abnormality, we have to compare it to normal standards. There are age-related normal values that people can use in order to determine whether an electrocardiogram is normal or abnormal.

DR. CAMPBELL: I think the most important thing I can say is don't trust the computer interpretation of the QTc. The other is the pretest concern. If you're obtaining an EKG because of the concern of chest pain, then my level of suspicion about a borderline QT would be lower than if they've had a history of syncope or unexplained seizure. So again, it sounds redundant, but the clinical correlation is probably one of the most important things. I would also say that it's a critically important measurement on the EKG. It's the hardest to get correct, and it's important at that point to be using, I think, your pediatric cardiologist for any confirmation of that measurement and also the interpretation. The concern really is around two things. One, does this patient have a diagnosis of long QT syndrome, which can be congenital or genetic and run through families? So obviously, you want to make that determination, but most patients may be at risk for sudden cardiac arrest or sudden death. The other would be managing dosing when you're using drugs that are known to prolong the QT interval or polypharmacy, which heightens the risk for drug-associated or drug-induced QT prolongation. So it's a very important number to get correct.

DR. BORIS: There's at least one website that lists medications that can prolong QT intervals. So where is your threshold for referral for a corrected QT? Back when I was training almost 20 years ago, we used 0.45 seconds or 450 milliseconds, but I think that's changed over time.

DR. CAMPBELL: Yes. There's not a single number. It's age-dependent. It's also gender-dependent. In the adolescent population, a QTc greater than, say, 0.47 seconds or 470 milliseconds in a male should be of concern. Greater than or equal to 480 milliseconds in an adolescent female, I think you should be concerned too. The other thing that's important to note is that you can have genetic and gene mutation-positive long QT syndrome and have a normal-looking QT. I think of long QT syndrome, like anything that's called "syndrome," as being a little bit like a jigsaw puzzle. There are multiple pieces that you try to put together to derive the final picture. So there is not only QT prolongation, but you can also have T-wave morphology changes, which can make you very suspicious for an underlying repolarization abnormality. So when you're starting to get into this area, I think you really want to use not even just a general pediatric cardiologist but a pediatric cardiac electrophysiologist to take your hand and help shepherd your way through this family and this patient.

DR. BORIS: Absolutely. Some other reasons to get an electrocardiogram would be in the evaluation of chest pain, dizziness, and syncope; palpitations certainly, assuming those palpitations are happening frequently. Even if they're not happening frequently, sometimes it can show up things like Wolff-Parkinson-White syndrome, which would suggest that the patient has the risk for an underlying arrhythmia. Obviously, if there is congenital heart disease, either suspected, looking for chamber enlargement or abnormal location of the sinus node, we use it a lot in our clinics for follow-up of known congenital heart disease. Arrhythmia—and we've touched on that a little bit—if there's a suspicion of an arrhythmia because those patients may be at risk for sudden cardiac arrest or sudden death. The other would be managing dosing when you're using drugs that are known to prolong the QT interval or polypharmacy, which heightens the risk for drug-associated or drug-induced QT prolongation. So it's a very important number to get correct.

DR. CAMPBELL: Yes. There's not a single number. It's age-dependent. It's also gender-dependent. In the adolescent population, a QTc greater than, say, 0.47 seconds or 470 milliseconds in a male should be of concern. Greater than or equal to 480 milliseconds in an adolescent female, I think you should be concerned too. The other thing that's important to note is that you can have genetic and gene mutation-positive long QT syndrome and have a normal-looking QT. I think of long QT syndrome, like anything that's called "syndrome," as being a little bit like a jigsaw puzzle. There are multiple pieces that you try to put together to derive the final picture. So there is not only QT prolongation, but you can also have T-wave morphology changes, which can make you very suspicious for an underlying repolarization abnormality. So when you're starting to get into this area, I think you really want to use not even just a general pediatric cardiologist but a pediatric cardiac electrophysiologist to take your hand and help shepherd your way through this family and this patient.

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Leo, do you want to talk a little bit about the electrocardiogram? It's fairly easy to get an electrocardiogram; it's relatively inexpensive; and it's available in most clinical centers. The things that you need to know about the electrocardiogram is that it can be fairly insensitive and nonspecific. We spoke about the normal values that we use for the electrocardiogram, and we all just have to recognize some of the limitations associated with these normal values. These normal values were obtained in a relatively small population of Caucasians back in the 1970s, and we have not updated these normal values recently. There is a study going on right now to try and obtain a bigger and more robust data set of normal patients, looking at their electrocardiograms, and hopefully we will have those data sometime in the next year or so. The other limitation of the electrocardiogram includes the fact that we're only looking at 2.5 seconds of time for that particular patient. Some problems with arrhythmias may not become apparent during those 2.5 seconds.

DR. CAMPBELL: One of the other concerns is that a normal ECG may not exclude the presence of really serious heart disease. I was talking with Jeff earlier today and there was an ECG that was ordered last week for a family history of heart disease, a very nonspecific, very broad collection of potential disorders. The ECG was normal; so the most appropriate response for me was to pick up the phone and call the pediatrician saying, “What do we know about this family history?” What are the details, and would a normal ECG help us to exclude that in that patient?” Again, the pretest probability or the pretest suspicion before ordering any of these tests is critically important.

DR. BORIS: Absolutely. Let's talk now about chest radiograms or chest X-rays. Obviously, they come in a couple of flavors: PA [posteroanterior] or AP [anteroposterior], as well as a lateral. It does assume that there has been an adequate inspiration and adequate alignment. I can't tell you the number of times that I've looked at chest X-rays, and they've either been rotated or the lung fields are not inflated enough, and so you can't tell anything about the vasculature, you can't tell anything about heart size. Chest X-rays do evaluate some things, though. They can tell you about heart and chamber size, especially if you have a moderate-to-large pericardial effusion or a pleural effusion as well. It can demonstrate the position of the heart, and it can also help you to look at the apex of the heart. It can sometimes help with demonstrating the size, position, and appearance of the great arteries. That includes looking at things like coarctation of the aorta. You can look at something called a “3-sign” with coarctation of the aorta. You can see in patients who have had a homograft placed in the pulmonary or the aortic position. Sometimes you can see calcifications of those as the immune system does their work. Tumors, calcification, and masses also can show up on these. You can also look at—as I mentioned before—the lung fields, including looking for pleural effusions, assessment of the pulmonary vasculature, and function of the diaphragm. Something that we sometimes see postoperatively is if the phrenic nerve has been damaged; one of the hemidiaphragms may not come down all the way. Leo, do you want to talk about some other things that chest X-rays can be helpful for?

DR. LOPEZ: Sure. Chest X-rays are also useful for other extracardiac evaluations like looking for a thymus. It's often present in the neonate and can often be confused for the cardiac silhouette if somebody's not careful. You also have to know that, after cardiac surgery, the thymus is usually taken out. It shouldn’t be there if the patient's already had heart surgery. Bony findings, things like pectus excavatum, things like rib notching associated with aortic coarctation, tumors associated with the bone, hemivertebrae as is often found in VATER syndrome. Of course, the chest X-ray is useful for patients that have undergone some type of intervention, in terms of looking at the placement of devices to close atrial septal defects or patent ductus arteriosus, to look at the function of those devices to see whether or not they're still in the appropriate place. It's useful for looking at stents and whether or not they're still open or whether there are fractures that have appeared on the stents. Patients that have undergone heart surgery will have sternal wires, and looking at the sternal wires for fractures and displacement can sometimes help you in terms of taking care of those patients.

DR. BORIS: One of the other things that the presence of the thymus can do or conversely the absence of a thymus, it can suggest that the patient has 22q11 deletion or DiGeorge syndrome, which can be associated with very specific cardiac defects as well as other multisystem problems. What are some reasons then, Robert, we would not want to get a chest X-ray?

DR. CAMPBELL: Number one would be unnecessary radiation exposure. A chest X-ray is probably most helpful clinically for interpretation of lung findings, pneumonias or atelectasis. A large heart may indicate the presence of a large pericardial effusion, but it may also indicate the presence of a cardiomyopathy. So the distinction on X-ray may be hard. If you're going to be ordering an echocardiogram to assess intracardiac or cardiovascular abnormalities, I'm not sure that the chest X-ray adds that much more.

DR. BORIS: Now let's talk about echocardiograms. Echocardiograms are a transthoracic ultrasound of the heart as opposed to a transesophageal study, which is more invasive and more invasive. It combines two-dimensional color Dopplers, spectral Doppler, M-mode, and tissue Doppler to make several different types of assessments. It looks at the anatomy, looking at the two- and three-dimensional structure of the heart. It can look at the normal and the abnormal structures, including their spatial relationships, connections, and sizes, the chambers, the valves, the arteries, the veins, and even shunts that have been placed there or that have developed abnormally. It also demonstrates physiology. So with normal and abnormal ultrasound, it's able to help us estimate gradients of pressure and estimate systolic and diastolic function. We can estimate the gradient or the amount of obstruction across a stenotic valve. We can get a visual estimate, in terms of estimate of the severity of the degree of leak of a valve. We can also estimate the pressure in a chamber or across shunts.

Leo, what are some reasons that we as cardiologists would consider getting an echocardiogram?
and symptoms that may make one think of a congenital heart defect. Echocardiograms are also useful if a patient has a known congenital heart disease that needs to be followed up, either preoperatively or postoperatively. Echocardiography also is useful in patients where you suspect a cardiomyopathy. In patients who have known cardiomyopathy, it’s a useful surveillance tool. It’s also used in first-degree family members of patients with known cardiomyopathy. Other uses for echocardiography would be in the setting of a pericardial effusion. If you have other testing that has been done to make you worry about a congenital heart lesion, for example, if the electrocardiogram or the chest X-ray has some abnormality, if a pulse oximetry test reveals some abnormality that makes you think there is a congenital lesion, an echocardiogram would be useful. Lastly, if a newborn had a fetal echocardiogram that was suspicious for some type of heart lesion, then a postnatal echocardiogram would be warranted as well.

DR. BORIS: There are a number of systemic diseases that are also associated with cardiovascular disease, and they are numerous, but we do need to screen these patients: so, patients who’ve had cancer, especially when they’ve gotten anthracycline therapy or external beam radiation therapy; patients who’ve had hemoglobinopathies; patients who’ve had myeloproliferative disorders; patients who’ve had connective tissue disorders. We’ve already talked a little bit about inflammatory disorders like Kawasaki and rheumatic fever; also Takayasu arteritis and systemic lupus erythematosus. Patients with muscular dystrophy and renal failure, stroke, all those things are indications for echocardiography. Hypertension, a lot of people don’t realize that echocardiography is actually part of the initial evaluation in patients who truly do have a diagnosis of hypertension. Then there’s some storage diseases and abnormal situs of the heart. Also a history of maternal phenylketonuria.

Let’s talk about why we shouldn’t get an echocardiogram. I want to preface this by saying that all three of us were involved in a document in 2014 about appropriate use criteria. Robert was one of the lead authors on this, so we’ll let Robert talk about this bit.

DR. CAMPBELL: The 2014 statement from the American College of Cardiology dealt with appropriateness of initial transthoracic echocardiography in the outpatient setting for that initial patient evaluation and broke it down into three different classifications. One was “Appropriate,” one was “Rarely appropriate,” and the final was “May be appropriate.” It’s an important reference for you if you’re going to be ordering echocardiography. There are many types of clinical symptoms or clinical scenarios for which we may think to order an echo but the pretest probability of an abnormality is incredibly low. The other is that a negative echocardiogram sometimes does not exclude the presence of certain diseases. A negative echocardiogram does not exclude Kawasaki disease. A negative echo does not necessarily exclude endocarditis or rheumatic fever. Again, if there’s a concern about ordering an echocardiogram, hopefully in this day and age there’s a connection, a close communication, with a pediatric cardiologist to discuss whether this test is appropriate or not, and how to interpret the results.

DR. LOPEZ: It’s really important for people to have a familiarity with the common indications for echocardiography in the outpatient setting. I think this document is very good about giving some guidelines to not just primary care providers but also pediatric cardiologists on when an echocardiogram is warranted.

DR. BORIS: I want to clarify: On echocardiograms, there’s some really good data in pediatrics. If you have a positive blood culture and you have a new or changing murmur, echocardiogram is absolutely helpful, but it can’t rule out endocarditis, it cannot rule out a congenital coronary artery anomaly, it can’t rule out tamponade, so it can’t diagnose tamponade even if you have a pericardial effusion. Even if a patient has cardiomyopathy, it doesn’t mean that it diagnoses heart failure. So I think we need to remember that the physical examination and the history are so important to understanding what the diagnosis is and to not rely on the testing quite so much.

DR. CAMPBELL: We talked earlier about some of the cardiomyopathies and channelopathies, these disorders that may be genetic and run through families predictably will have a negative or a normal echocardiogram; therefore, if that’s the suspected disorder, the indication for an echocardiogram is probably low on the list. On the other hand, a concern about hypertrophic cardiomyopathy or dilated cardiomyopathy, some of which may be genetic, a negative echocardiogram early in life does not mean that you will not manifest a phenotype of hypertrophic or dilated cardiomyopathy later if you aren’t genetically affected.

DR. LOPEZ: I want to highlight one of the things that Jeff said in that the echocardiogram does not replace the physical examination, and it does not replace a good family history.

DR. BORIS: Robert, let’s talk about Holter monitors. Can you discuss what they are and how they’re used?

DR. CAMPBELL: The way I explain a Holter monitor to a family is it’s basically a 24-hour EKG tape recorder. It will record the EKG signal that it sees from that patient for 24 hours. The advantage of a 24-hour Holter monitor is that, during that 24 hours, we will be able to see sleep heart rates and rhythms; I ask our patients to exercise hard during that 24 hours to see the upper end of rates and rhythms. Then, of course, everything in between. It will give you a benchmark maybe before treatment of arrhythmias before using certain medications. The indication to use that to chase down palpitations would be very much dependent upon the frequency or the inducability or the provocability, if you will of the arrhythmia during that 24 hours. A 24-hour Holter monitor is what’s called an ambulatory EKG event recorder, which can be used over a course of 2 weeks or 4 weeks for symptoms that are less frequent, maybe not as predictable or provocative.

DR. BORIS: That’s one of the things that we see fairly frequently is people will automatically order a 24-hour Holter monitor for events, palpitations that don’t occur all that frequently. They’ll occur once a week or once a month. So a Holter monitor is not going to show you a lot. But a Holter can be helpful in patients who have frequent ventricular or atrial ectopy, in patients who have pacemakers, and in screening for occult arrhythmias in
patients who either have repaired congenital heart defects or in patients who have long QT.

So, Leo, what would we not want to get a Holter monitor for?

DR. BORIS: As Robert mentioned, there are rhythm events that don’t occur daily. In that setting you need something that will be able to capture that event. As Robert indicated, a transtelephonic monitor or a long-term monitor would be more appropriate in that setting.

DR. BORIS: Let’s talk about transtelephonic monitors or event recorders a little bit, Leo.

DR. LOPEZ: Sure. It’s a patient-activated or event-activated monitor. It’s not constant. What happens is the patient keeps this device for up to 30 days and will record any of the events that the patient chooses into memory, and then it’s off-loaded by a landline phone to some reading station, which is then transmitted to the cardiologist for interpretation.

DR. BORIS: So, the evaluation of infrequent events would be best served with one of these monitors.

DR. CAMPBELL: One of the new technological advancements has been what’s called an implantable loop recorder. Most patients would rather have heart surgery than to wear a 30-day event recorder in terms of the cosmetic concerns or the pride. They don’t want to be seen wearing this monitor at school, and the leads are quite corrosive to the skin. But for patients who have infrequent and unpredictable and nonprovocable symptoms but of concerning history, recurrent exercise syncope, where you’re trying to find the rate and rhythm during this event, an implantable loop recorder can be considered now.

It is an invasive procedure but relatively simply performed as an outpatient in most cath labs and is an alternative to long-term skin-lead electrode event recorders.

DR. BORIS: There are a couple of different kinds of these event recorders. One of them is called a loop recorder, another one I refer to as an event recorder. The loop recorder stays on with electrodes that you change every day. The event recorder is something that the parent may keep in a purse or a backpack or in a pocket or the patient can, and they bring it out and put it on the chest to activate it at the time of an arrhythmia.

Let’s switch gears a little bit and talk about exercise stress testing. Leo, do you want to talk about how those work?

DR. LOPEZ: Sure. Basically, a patient will go through a graded progressive exercise protocol either using a treadmill or a bicycle. During that period, the patient’s heart rate, blood pressure, oxygen saturation, electrocardiogram, and various other things are monitored. With this testing, we can assess things like exercise capability and tolerance. We can do VO₂ and determine the transition from aerobic to anaerobic metabolism.

DR. BORIS: This helps us evaluate things, as Leo mentioned, like exercise tolerance both before and after cardiac surgical intervention; if there are exercise-induced or genetic arrhythmias, which we cannot always deny but at least confirm the diagnosis. If the patient has a resting arrhythmia, especially PVCs [premature ventricular contractions], if you have suppression of arrhythmia with exercise, that can be reassuring, versus if you have worsening of your arrhythmia with exercise, that can be concerning. It can evaluate dysfunction of the sinus node. It can also be used for evaluation of exercise-associated chest pain, exercise-associated asthma.

What are some reasons that we would not want to get an exercise stress test, Robert?

DR. CAMPBELL: One of the important concerns would be the age of the patient and their ability to perform a maximum exercise test. We have been able to successfully coach and coerce young patients, sometimes even 4, 5, 6 years of age, and especially with bicycle testing. The other would be that if there’s not a good pretest probability. Again, a negative exercise test for certain things doesn’t exclude the presence of disease.

DR. BORIS: I think also—and Leo mention this before—when talking about obtaining VO₂ measurements, that’s done by putting a fairly tight-fitting mask over the nose and mouth of the patient or at least the nose of the patient. If a patient has claustrophobia, that would be a contraindication for trying to do that; certainly if the patient can’t physically accomplish the test from a musculoskeletal standpoint or from a balance standpoint. If there’s a high risk of lethal arrhythmia, I think that’s probably a relative contraindication. I think you need to be prepped for dealing with that arrhythmia in a pinch.

DR. CAMPBELL: That may be the single most important test to order, but it’s also the test that most needs the emergency action plan preparedness during the testing.

DR. BORIS: Absolutely. I had a patient who had exercise-associated ventricular tachycardia, she felt it but didn’t pass out. When I did the exercise stress test for her, I made sure that I had a syringe of lidocaine ready to give her. Lo and behold, at peak exercise, she popped into VT. I said, “How do you doing?” She said, “My chest hurts a little bit.” I’m like, “Okay. Let’s fix that,” gave her some lidocaine, and that was it.

DR. BORIS: Let’s talk about some other tests that we sometimes see but are less frequently used. We have a 24-hour ambulatory blood pressure monitor. Leo, do you want to talk about how those are used?

DR. LOPEZ: The problem with obtaining blood pressures in the office is that they can sometimes be falsely elevated. The 24-hour ambulatory blood pressure monitor allows us to look not only at the baseline blood pressure for the patient when they’re not faced with people wearing white coats, but it also gives us an idea of the variability of blood pressure during the course of the day because there are some normal standards for that.

DR. CAMPBELL: Actually, it demonstrates diurnal variation as well, so you can look at what’s going on when the patient’s alone. I know the adults are using this a lot more, and I think that’s increasing amongst pediatric providers as well. Can you talk about stress echocardiography?

DR. LOPEZ: Stress echocardiography in pediatrics is not as common as it is in adult cardiology partly because the faster heart rates make the sensitivity and specificity of the test not as good. There are people that will use stress echocardiography to see whether or not there are changes in myocardial function, particularly in patients that may have coronary insufficiency.

DR. BORIS: We’re certainly seeing those in our center with the coronary artery anomaly program. What about nuclear medicine studies? Those are studies that are done in adults. What kind of use do we have for pediatric patients?

DR. CAMPBELL: For adults, nuclear medicine studies probably would be most helpful in the setting of coronary disease. Fortunately, in our patient population, coronary disease associated with atherosclerosis is relatively rare. However, we do
have a patient population of postop or Kawasaki coronary abnormalities where that may be helpful. Alternatively, you can lay in bed in a fashion, such as magnetic resonance imaging now, which can give you good detail of proximal coronary arteries and also look at adenosine perfusion during the testing.

**DR. CAMPBELL:** Of the things that we’ll sometimes do with the nuclear medicine perfusion test we’ll look at differential pulmonary perfusion, especially in patients who have small branch pulmonary arteries. We can see how much is going to the left side versus to the right side, what the degree of obstruction is, and if these patients need to have intervention or not.

1. Of the tests that’s being used less frequently now, but I have patients come to me specifically in my postural orthostatic tachycardia syndrome program with an expectation for the tilt table test. For those that have not seen a tilt table test or are not familiar with it, the way a tilt table test works is the patient is brought into a relatively dark room in a postabsorptive state. They lay down on a table, they have an IV (intravenous) placed, and EKG leads, blood pressure, pulse ox and whatnot. The table has a footboard on it, and they are strapped down to the table. The patients are typically observed for anywhere from 10 to 20 minutes, depending on the protocol. Then the table is mechanically tilted up to 65 with 10 degrees head up, and then the patients are observed for anywhere from 10 to 40 minutes, again, depending on the protocol.

2. This used to be used a lot more frequently in the evaluation of things like syncope. The problem is that it’s probably a way too provocative test. If we were to go to the local high school and grab 100 asymptomatic patients and put them on a tilt table, 40 of them would tilt positive. So it’s very provocative. In my POTS program I don’t even use tilt table testing at all. I will do a 10-minute standing test, which is much more physiologic, and the patients appreciate that.

**DR. CAMPBELL:** The test is only helpful if it’s positive. There are only two positive results. One would be to reproduce the clinical symptoms of syncope and demonstrate that there is a causative hypotension and/or bradycardia or asystole behind that, which would then help confirm the diagnosis of vasovagal or neurocardiogenic syncope. The other test that’s positive would be the patient who has a “syncopal event” with normal heart rate and blood pressure, which raises the specter of stress or psychiatric syncope.

**DR. BORIS:** We’ve discussed the tests that we as cardiologists would use, but I think I would like to reiterate that, for my primary care colleagues, these tests should be ordered only in conjunction with a pediatric cardiologist except for maybe an electrocardiogram. It would be more cost-effective, more helpful, and more informative to the patients and families if the patient ends up having a pediatric cardiologist referral if there is doubt or it is unclear whether the patient has a congenital cardiac defect or some other acquired concern.

I’d like to thank my colleagues, Robert Campbell and Leo Lopez, and I’d also like to thank Washington University School of Medicine and St. Louis Children’s Hospital for giving us the opportunity to discuss “Optimal Use of Cardiology Referrals and Cardiac Studies.”

List of Supplementary Reading Materials

To help you obtain more information on the subject of this program, the following supplementary reading materials are recommended:


Answers to Pretest Questions

1. b
2. True
3. a
4. False
5. False
6. a
Post-test Questions

After listening to the audio CD and reading the transcript of this panel discussion, measure your expanded knowledge of the subject by answering the multiple-choice questions below or online at www.pedupdate.com.

1. Check only one answer for each of the 12 questions.
2. When you have completed the test, transfer all 12 responses to the Post-test Response Form.
3. Return your answers in the preaddressed envelope marked “Post-test Response.”
4. Your confidential score will be sent to you on a quarterly basis.
5. If any questions are answered incorrectly, the cumulative score card will indicate the pages and paragraph numbers where the correct answers will be found.

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Release date: April 1, 2017. Test invalid for credit after April 1, 2020.

1. An electrocardiogram should NOT be routinely obtained in the evaluation of
   ![Image](image1.png)
2. A history in an infant of crying in pain with eating that resolves when the baby stops eating is strongly suggestive of
   ![Image](image2.png)
3. A Still’s murmur is characterized by being a
   ![Image](image3.png)
4. Rheumatic fever includes which one of the following?
   ![Image](image4.png)
5. An electrocardiogram (ECG) measures the cardiac electrical signal for
   ![Image](image5.png)
6. An electrocardiogram should NOT be used to rule out
   ![Image](image6.png)
7. Coarctation of the aorta can manifest on a chest radiogram by
   ![Image](image7.png)
8. Echocardiography should NOT routinely be used in the evaluation of patients with
   ![Image](image8.png)
9. An echocardiogram can be used to
   ![Image](image9.png)
10. A Holter monitor should be used for recording and assessment of events occurring as frequently as
    ![Image](image10.png)
11. The ambulatory monitor that is patient-activated or event-activated is the
    ![Image](image11.png)
12. A cardiac test that can be considered reasonable to order by a primary care provider, independently of a pediatric cardiologist, is
    ![Image](image12.png)