MATTHEW HAZLE: I have my support team over here.

[LAUGHTER]

Well, thank you for having me. It's great to be here.

So I think, if I'm correct, I'm probably the only pediatric-oriented to talk today. Hopefully this will be useful for you guys and interesting. I'm going to talk about sudden death in children, which is, of course, a horrible topic but interesting to those of us who do this for a living. And just to start, we going to just go over the goals of my presentation.

First of all, I want to review the common conditions that are associated with sudden death in children and teenagers. I want to give some recommendations on how to screen for these conditions that are often previously unrecognized. Hopefully, there are some folks in the audience who will be doing this.

I want to, along the way, provide a framework for when to refer to someone like me for the quote "work up." And then lastly, I want to touch upon what to do if you actually get that phone call as a provider, either a patient of yours or a patient's family member who has an arrest or a sudden death and how to navigate that. Because I think oftentimes, especially primary care providers are that first contact.

So just some terminology, real quick. What is sudden cardiac death? Well, it's the sudden cessation of cardiac activity with hemodynamic collapse, usually due to a malignant arrhythmia where we're talking about VT or VF. It's by and large in previously healthy children.

If circulation is restored, the-- either spontaneously or via defibrillation, then it's classified as a sudden cardiac arrest or an aborted cardiac arrest. Some people use the term aborted cardiac death. I find that a little confusing, so I like those two. If there is not a successful intervention, then it is classified as a sudden cardiac death. And you might see in the literature or in the terminology, if a patient who has a sudden death has a negative autopsy and there's no structural component identified, then often this Sudden Arrhythmic Death Syndrome is used, or SADS.

So when we're talking about sudden death, obviously athletes are, come to mind, even though what I do want to stress is this can affect anybody. The athletes, I think, may be more prominent in the news and social media, so it definitely grabs everyone's attention. So there is a lot of literature, fortunately, on athletes, and I'll go through some of that.

But just thinking of numbers, OK, what is the scope of this problem here? US athletes under 35 years of age. There's about 10 million people a year who participate in some level of sport. The sudden death estimate-- there's a big range. And I'll go through that a little bit, but if you have to take all the numbers and just give you guys a number to hang your hat on, I think one in 200,000 participants a year is a reasonable number.

The range, of course, is very extensive, so as mentioned here, one in 25,000 in some studies, up to one in 350,000 in others. So if you're looking at the US as a whole, you're dealing with about 50 to 100 cases a year. In an area the size of Forsyth County, 350,000 or so, you're looking at about one per year on average.

What's interesting is we're, as more and more literature comes out, we're finding that there's probably a differential risk based on a lot of factors, age, gender, race, even the type of sport. If you look at this study, it just kind of highlights that. They looked at sudden death in NCAA athletes over a four year period, which was, involved almost 45,000 students a year.

And they found quite a discrepancy in the different sports and the different athletes, ranging from, for example, football players, one in 40,000. But then if you look down in basketball players, it was kind of impressive, one in 11,000. But then if you dive into that a little more, male, NCAA, Division I, African-Americans, one in 5,000, and that's pretty striking.

The one criticism of this study is that it didn't-- not everybody had full autopsies, et cetera, so there could have been some sudden deaths that were not even cardiac. But I think it definitely, maybe gives us a framework to think about, OK, who deserves more extensive screening because, as I'll touch on later, we can't, unfortunately, screen everyone. It's just too expensive.

So I thought it would be helpful to go through some of the causes of sudden death in children, and we break it down into two main categories. We think of structural or functional abnormalities of the heart. We think of electrical abnormalities. The heart is usually normal in that case, by autopsy, and then a kind of grab bag others. I'll focus mostly on the structural, functional, and electrical causes, but cardiomyopathy is obviously a big one.

Hypertrophic cardiomyopathy, by far the most common cause of sudden death in children and teens. Other cardiomyopathies are important, though. Dilated cardiomyopathies, a cardiomyopathy called ARVC, which stands for arrhythmogenic right ventricular cardiomyopathy, and that's more of a recently appreciated condition. Coronary abnormalities are also high on the list. When we throw-- when I say coronaries, coronary artery disease, of course, in an adult-oriented audience, that means something very different than to me.

I think of an anomalous origin of a coronary, and I'll describe what that means. But if you look at studies that involve especially the older end of the spectrum up into the 30s, definitely coronary artery, classic coronary artery disease is on that list. And a prior history of Kawasaki is definitely a risk factor for ischemic heart disease and sudden death.

Congenital heart disease, both undiagnosed and postoperative, are on that list, as well. Aortopathies and specifically Marfan syndrome, is something that we manage and get asked to evaluate quite frequently. And then lastly, more of an inflammatory condition, would be myocardities.

The electrical conditions which I primarily deal with would be Long QT syndrome, Wolf-Parkinson-White syndrome, a condition called Catecholaminergic Polymorphic Ventricular Tachycardia, which is a mouthful. We usually use the acronym CPVT, and Brugada syndrome.

The others, of course, drugs, stimulants, prescription, medications. Pulmonary hypertension is definitely on that list. Fortunately, we don't see a lot of that but something to consider. And commotio cordis is a rare condition when an athlete suffers an impact to the chest at a critical timing on the T-wave in the vulnerable period, and they can have VF because of that.

So this is the-- if you look at any, if you go to any talk in the pediatric world, sports talk, electrical talk, this is the classic slide from Barry Maron. He's done a lot of work with hypertrophic cardiomyopathy, but he looked at about 1,500 competitive athletes in Minnesota over a 25-year period. And this is the graph, the distribution he got in sudden deaths, and you can see that a lot of them are structural. So you have hypertrophic cardiomyopathy, over a third. There's some there, that intermediate LVH possible, so that puts it even above 40%.

Coronary artery abnormalities, a big piece. Myocarditis, ARVC, so going around clockwise, you have a lot of structural. And then up in the top, you have a very small component of electrical, and I think we're understanding more that electrical is definitely more prevalent. But even back in 2007, some of those were probably not as well understood.

I wanted to just highlight this one because even though it's an older study, it's a good one just based on the sheer number of people involved. Six million military recruits studied, and they have an incidence of one in 50,000 who had sudden deaths. And if you look at the cardiac causes up there, which were about half, you again see the common themes. You see the anomalous left coronary artery, 33%, so in this case it was a third of all these sudden deaths.

Myocarditis, and that's confirmed. When we see myocarditis, that's usually identified by autopsy. Hypertrophic cardiomyopathy and, I think because it involved up to 35 years of age, even classic coronary artery disease.

What I wanted to highlight here, and we'll talk about this as relates to screening, is half of these young adults had exertional symptoms before they died, and the vast majority of these sudden deaths were exercise-related. If you look at the unknown causes, there was an impressive number who had a family history of sudden deaths. So this is important, of course, thinking about screening. Family history is going to be really important.

So just to go through some of these in detail. Hypertrophic cardiomyopathy, the most common cause of sudden death in the US. It's, the prevalence of this condition is about one in 500 people, which is kind of scary when you about it.

It's, this condition, in addition to most of these that I'm going to talk about, are autosomal dominant. So you have a 50% chance of passing this on to your offspring. So it makes it kind of easy to sort out a family tree. But these mutations are involving, by and large, the myocyte structural proteins, and then you get pathologic hypertrophy, scarring, fibrosis, ischemia, and that's a setup for VT/VF.

The-- interestingly, kids who have this condition most often have events when they're not exerting themselves. So rest or mild exertion is a common, is a common scenario. Even though we focus on the athlete with hypertrophic cardiomyopathy, what I wanted to kind of just impress upon everyone is kids can also have these events at rest. And a lot of them are asymptomatic or only mildly symptomatic before their first event.

So how do we pick this up in the clinic? Well, history is big. History of palpitations, excessive dyspnea, chest pain, or syncope during exertion. You're going to see this theme throughout all these conditions, but that dyspnea is important because these kids with thick hearts have diastolic dysfunction.

So when they exert themselves, the heart can't relax, and they get increased left atrial pressure. They get respiratory symptoms. They get dyspneic. So we get a lot of referrals for exertional dyspnea, and that's one thing we do think about a lot.

Family history is important with these, sudden death of an unknown or unexplained, less than age 50. On exam, you're going to have a systolic murmur, which is, of course, not that helpful. But the classic is when you have them stand up or Valsalva, the murmur gets louder. And that's the classic exam for a dynamic left ventricular outflow tract obstruction because of that thick septum.

Here's a classic EKG. When you're thinking of hypertrophic cardiomyopathy, you have a teenage athlete who fainted on the basketball court. Automatic cardiac work-up, right? So you get this EKG, and you're definitely worried. LVH, a lot of ST, T-wave abnormalities, inferior and laterally. So this is kind of the hallmark EKG that you're going to be worried about.

This is a condition that, again, us electrical people get all excited about, but it's very rare. But I wanted to just present it so you guys have heard of it. You've seen it. The prevalence is estimated at one to 2,000-- 1 in 1,000 to 2,000, but in Europe, it's actually quite a common cause of sudden death. And that's probably gene related in that group, but in Italy, it's a big deal.

Autosomal dominant, again. We're talking about mutations and structural proteins, the components that hold the cells together. Cell-cell adhesion, proteins called desmosomes, and you get this abnormal fibrofatty infiltration of the RV. You can see that histologic section. The right ventricle has pretty much been replaced by this fibrofatty abnormal tissue, which is a set up for VT.

And the kids who are going to be symptomatic are going to have palpitations from their VT and presyncope/syncope during exertion from the VT. Chest pain, of course, is going to be on that list, although not super specific for this condition. But there will be a handful of these, as with all of these conditions, that will present with sudden death as their initial presentation.

Here's an EKG. , Again, I'm not here to go over a bunch of EKGs, although I'd love to do that. The epsilon wave comes up. You guys may have heard that term. This is an epsilon wave, and the epsilon wave is terminal depolarization abnormalities from the abnormal RV myocardium.

So at the right precordial leads, you're going to see that kind of slurring, that delayed depolarization, widened QRS with a little bump on it. That is an epsilon wave. We get, some people get really excited when they see bumps different places on the end of the QRS or the T-wave, but I just wanted you guys to see what a real epsilon wave looks like.

This is one that we do a lot of echoing for in the clinic. When the kid has exertional chest pain, this is one of the reasons that buys them an echo. We're looking for an anomalous origin of the coronary artery.

If you look at the normal, on the left of course, you can see the course of the coronaries. An anomalous left coronary from the right sinus of Valsalva has an abnormal take-off, usually slit-like orifice in an abnormal course through the great arteries to get back to the left side. And all those factors, especially if there's a, what we call an intramural course of that artery through the wall of the aorta, that's a set up for demand ischemia. So anomalous left is a biggie on the list for us when we have exertional chest pain or syncope.

You can have anomalous right from the left sinus, not as commonly associated with sudden death, but definitely described. We don't really know the prevalence because there's people running around with this at this time. But in that-- in some studies, it is an important cause of sudden death in young people, usually under age 30, and often sudden death is the initial presentation.

The scary thing about this one is exam, resting EKG, all normal. But echo will pick it up. And if you can't identify the coronary origins by echo, which we usually can in healthy kids because they're small and make good pictures, we will CT some of these kids if [INAUDIBLE]. But you-- that's, that's the-- we definitely got to nail down the coronaries when we see these kids for a work-up.

I wanted to touch on Marfan syndrome because we see a lot of rule-out Marfan. And I will say that if a kid is tall and goofy looking, doesn't mean they have Marfan syndrome. That's the majority of kids that we see. Family history gets me more interested. An uncle who was tall and goofy looking, who died suddenly. OK, now I'm interested.

Because again, these are autosomal dominant mutations, although it can be sporadic. Family history should be 50% likelihood of passing it down. So it's quite-- it's, prevalence is one in five to one in 10,000. So it's not super common, but that's the classic phenotype, right?

You've got very tall, very lanky, arachnodactylly, which is the skinny fingers and toes. A pectus deformity, scoliosis, eye problems, skin problems, meaning stretch marks and stuff like that, so a lot of musculoskeletal symptomatology. The phenotype that we worry about, of course, is the dilated aortic root, which is shown there on the CT. And of course that is a risk factor for dissection and sudden death.

Myocarditis is one that can strike suddenly. I think a lot of myocarditis is subclinical, but if you look at ED data, about one in a hundred thousand ED visits will have a diagnosis of myocarditis. If you look at the retrospective studies that did autopsies, histologically present and up to 20% of cases, so it is important.

There-- it can happen at any age, but we definitely see a peak incidence in infants and in teenagers. Infants are more likely to present with heart failure. I think teens are probably more likely to present with the sudden death issue.

Usually infectious etiologies, so we have a previously healthy kid, and they get a virus. And it's just the perfect storm of the right host and the right virus, and they get myocarditis. And you have to have a suspicion for it, of course.

When a kid comes in after an arrest, that's high on the list. A kid in the clinic with exertional chest pain, et cetera, who is not having fever, who is not having viral symptoms, I think less likely. But the classic EKG is going to show you diffuse ST abnormalities, and this is an example of an infant with myocarditis. And often associated with pericardial irritation, or pericarditis, so you'll see this is a good example of pericarditis.

WPW is an important one. The incidence of the pattern, not the syndrome, is one in a thousand. So quite common actually, but in order to have the syndrome, you have to have the SVT that goes along with it.

So a kid with symptomatic WPW would be presenting with palpitations, most likely. Fortunately not a lot of syncope, not a lot of sudden death that we see, but there is a handful of sudden deaths that could be this condition because autopsy will be normal, right? And if you have no preceding EKG, you would never know. Most of-- a lot of these kids are asymptomatic. So we have a whole work-up algorithm on what you do with the asymptomatic kid who's identified to have pre-excitation, but there's a very small portion who will have a lethal, potentially lethal event as their presentation.

It's hard to know what is the actual risk or incidence of sudden death in these patients, but in symptomatic patients, we think it's not zero, for sure. 0.25% a year in some studies, so you have a kid-- and this is what I have to navigate with parents. The yearly risk is very low, but you take a cumulative risk over their lifetime and it's not zero. 3% to 4%, maybe.

And so that's why we often get, steer us towards ablation in these kids, especially if they're symptomatic, I think the adult EP lab is, they get kind of bitter because we take all the WPW these days. But they-- a lot of them get ablated before they become adults. So they don't see pathways as much as they used to back in the day.

Here's a good example of WPW, so pre-excitation, short PR interval and widened QRS with the delta wave.

This is the mechanism for SVT, so you're going down the normal conduction system normally. This is the most typical kind. Down the normal conduction system, up the pathway in a reentrant fashion.

So narrow complex SVT is the vast majority of what we see. This is not the cause of sudden death. So WPW, the reason WPW is associated with sudden death is not the SVT, which is 99% of what we deal with.

This is the cause of sudden death in people with WPW. If you're unlucky enough to have atrial fibrillation and you have a rapidly conducting pathway, you can get a pre-excited atrial fibrillation, which is a bizarre wide complex, fast rhythm that can degenerate to VF. So that's the cause of sudden death in WPW.

And fortunately, it's not common, but the reason we think symptomatic patients are at higher risk than the asymptomatic ones is oftentimes I think SVT will degenerate to atrial fibrillation. And we see that in the lab, too. We get a kid really fired up with SVT or pacing, and they'll go into atrial fibrillation.

Long QT syndrome is an important one. One in 2,500. These are genetic defects in the channels that regulate the heartbeat. So sodium and potassium channels are the biggies.

Autosomal dominate, again. Family history is critical with this. Unexplained death, drowning, car accidents, stuff like that that's-- Uncle Bob was in a car accident, and nobody knew why, or something like that. Especially drowning is a very-- we hear drowning, we get really interested in the possibility of long QT.

The risk of sudden death is very much dependent on where your mutation is, the host, the gender, the age, et cetera, but untreated, very malignant condition. 75% of-- up to 75% of people will have malignant arrhythmia by adulthood. Fortunately, a lot of Long QT is very effectively managed with beta blockers. So if we can identify these patients, we can really save a lot of them.

There's been some studies recently that even suggest that Long QT channelopathies are an important cause of SIDS deaths, and that's been confirmed by genetic analysis of SIDS deaths. And again, that's kind of more the power of genetic testing that has come online in the last decade or so.

When you think of Long QT syndrome, I want you guys to remember the triad of syncope, seizures, sudden death. Long QT does not present with palpitations, chest pain, unlike these other ones. Syncope, especially recurrent syncope, secondly that doesn't fit the classic vasovagal model, think about Long QT, for sure.

Traditionally, QT interval over 440 was, I think, classified as abnormal. I think we've gone a little more-- we gotten a little more lenient as we have more and more data on this, such that I would say in a child, I get interested if it's above 460. And then after puberty, you have a little shift because of the effects of testosterone, estrogen.

In boys, after puberty, 450 is an important cutoff, girls 470. And-- but of course, if the story is concerning, the QT almost doesn't matter, because up to a quarter of Long QT syndrome patients will have stone cold normal QT intervals. So that is something that you always want to remember. If the story is concerning, still could be on the list, for sure.

Here's an example of a Long QT patient, a four-year-old boy who got worked up because of a family history of sudden death. QTC 490, definitely-- I mean this is pretty much diagnostic with a good family history and an EKG like this.

This is another example. It's a poor quality, but I just put it up there so you guys can appreciate. This is what we get. We get the faxed EKG. Newborn, what do we do?

This is a newborn who has a very abnormal QT. Newborns can have abnormal QTs. It doesn't mean they have Long QT syndrome, but this gets your attention for sure.

And then the backstory is that dad has Long QT syndrome. Dad's mom passed away when dad was a newborn, suddenly. So this is a slam dunk, and that's why they got the EKG. But this is a good example. Broad-based t-wave, very long QT interval.

I threw in Brugada because I think it's out there. People understand it more. There's more data on it. It's a rare condition, but again, people throw that term out there. It-- Is it-- it is characterized by this ST elevation the right precordium. The A is the classic, Type 1, and then these what we call coved or saddleback morphologies are the important findings in the Brugada syndrome.

VT, sudden death are the hallmarks of Brugada, especially during setting a fever. So if you have a family with Brugada, which is very rare, you have a child with Brugada, you get-- you definitely get nervous with fever. They'll come into the ER with fever, and they'll have that pattern, whereas without fever, they might not be present.

Just to contrast Brugada versus a normal finding in an athlete, especially, we see early repolarization all the time. And one thing that we used to-- at least I do when I get asked, could this be Brugada, is you look at the ST interval. And you look at is it down-sloping or up-sloping, and that can kind of help you decide, and I thinking this early repolarization or Type 1 Brugada?

This one is an interesting one. It's rare fortunately, one in 10,000. But this is one that I definitely think about, I worry about if a kid just drops on the court, drops when they're doing their mile without much prodrome.

It has to do with the ryanodine receptor, which is calcium processing in the myocyte. And so there's autosomal dominant, typically, mutations in the ryanodine receptor. And so at high-- it's heart rate dependent. At higher heart rates, you get calcium overload ventricular arrhythmias. And patients will present with recurrent syncope because it kind of gets missed in childhood with physical activity, of course, but the one thing that is commonly overlooked is emotion. So anything with catecholamines.

So, I'll never forget. I had a case in residency where a girl, a little girl, I think she was around five or six, had recurrent syncope when she was flying her kite. And it kind of just got, I don't know, blown off, but she had an arrest. And it was all during excitement, not necessarily hardcore exertion.

We saw another girl in my fellowship who was having anxiety attacks, and the anxiety we found out was basically being provoked because she was having VD at school and passing out. And it was like this horrible, you know, vicious cycle. So think of emotion-- if you have syncope with emotional stress, that's an important consideration.

Normal resting EKG, but when you exercise them, you're gonna, you're gonna-- it's going to come out, if they've got the real deal. So you put them on the treadmill and as the heart rate goes up, they start to get ectopy and then a bi-directional ectopy and VT. And so even though the echo's normal, EKG is normal, we can usually diagnose it by an exercise test.

So we kind of went through the conditions. What the challenge-- how do we pick these up? How do we screen for these? Very low prevalence, right? I mean, even though like we touched on before, it's in the news. It's in social media. Parents come to clinic, and they want to make sure their kid doesn't have what their classmate had, or some kid at the neighboring high school had. It seems like it's very common, but it's very low prevalence, as far as the thinking of how are we as medical people going to screen for this.

This one recent study I thought was interesting. Barry Maron again, looked at teenage athletes in the state of Minnesota and found that the risk of sudden death was equal to a fatal lightning strike in that state over the same period. So that's kind of the concept that I want to impose.

They're very difficult to detect clinically, so how are we going to pick these up? Warning symptoms can be nonspecific, of course. A lot of times parents, rightfully so, will just blow them off. Oh, you're fine. And that's not necessarily wrong, but it can be disregarded. It can be kind of pushed to the side as far as-- especially if the symptoms aren't that impressive, like the exertional dyspnea issue with hypertrophic cardiomyopathy.

And in some cases, you can have a normal physical exam, a normal EKG. The challenge is, the work-up is very expensive, right? There's no-- it's kind of hard to have a middle ground.

You're either committing to echo, exercise test, a lot of work up, and restricting a child sometimes. In the fall, we get a lot, a big bolus of these "rule-out badness" referrals, and the kid has been restricted and they missed the tryouts, and so then their season is over. And so that's of course, not ideal.

So the pre-participation evaluations that we refer to, the concept of that is based on the fact that sudden death is exertional-related in a lot of cases, and in a lot of cases, you'll get a family history and some symptoms that you can really hang your hat on. The problem is even the AHA, the form that we all kind of go by, and I'll show it to you, none of them have been validated in a prospective way. So we don't really know how effective these are, but we use them.

So this is the AHA guidelines. It's a 12-step questionnaire, if you will, that is used. And it's back from 2007, but we still use it. You-- the three areas that we really focus on are personal medical history, family medical history, and the physical exam. So you can see there's a common theme that we've been talking about, exertional chest pain, unexplained syncope, excessive exertional unexplained dyspnea or fatigue with exercise, prior recognition of a heart murmur, elevated blood pressure, family history, premature death sudden or unexpected before age 50, especially if they know it was due to heart disease.

Disability from heart disease in a close relative under age 50, and those are important because some people may have no clue what Uncle Bob had. But they go, oh yeah, he had a heart condition for sure. Maybe he had a, you know, dilated cardiomyopathy or something. So you have to kind of use nonspecific terms, in some cases. And then of course, if you have a family history of a specific condition, which is all the ones we just went through, that would be very helpful.

Physical exam, heart murmur. Now murmurs are very common in kids, so we don't recommend referring every kid with a murmur on their pre-participation physical to cardiology. But the murmur, especially on an abnormal grade III or higher diastolic murmur, a harsh murmur, or the murmur that gets louder with Valsalva or standing, that one, that one would be a reasonable referral.

Femoral pulses are important. Lower extremity blood pressures are important in a kid who has hypertension because coarctation can be easily diagnosed that way. Marfan syndrome, the physical findings of Marfan syndrome would be something to definitely refer for. And the last one on the list is blood pressure, which not so much in my mind related to sudden death, but just an important kind of general well-being.

If you look at the literature, there's kind of a trend towards even more specific questions. This group just recently used those AHA 12 elements and an EKG, which is controversial, to screen 1,600 high school kids, collegiate, and professional athletes, and they found that 25% had a positive response on the personal or family history. And they found that based on various criteria to read an EKG, there was up to 27% percent had abnormal EKGs, so now you're looking at 25% of all these kids are going to be flagged for additional screening. I guess you-- I don't know. You could argue either way on that, but that seems pretty high.

So they developed, this is the Stanford 12 elements which is a little more specific. And I won't go through all these in detail, but you kind of get the flavor. So under history, you have, have you been diagnosed with these conditions? And they go through the list, all the ones we care about.

"Has there ever been a time where you had discomfort, chest pain, tightness, or pressure during exercise?" So that's similar. "Have you ever passed out or had a seizure--" that's an important thing-- "during or after exercise?" Because sometimes seizures is misinterpreted as seizure, epilepsy, but it actually was anoxia from your VT/VF event.

The exercise, shortness of breath is on there. Heart racing, palpitations is on there. Prior restriction for any reason, which they found was that was not uncommon, actually, and history of heart murmur. The family history, again, more specific, and they actually went down to 40 years and younger. Probably to rule out some of those, just to kind of eliminate some of those garden variety, coronary artery disease patients. So they really wanted to know about under 40.

And they talked a lot, they specifically talk about-- again, parents may not know the diagnosis, but they remember that a family member had a defibrillator placed. If you had a defibrillator placed at age 30, OK. I want to know more about that for sure.

So in my mind, and this was actually listed in this pediatrics article, the big four. This is kind of something you can hang your hat on. The things you want to ask a kid or a family-- have you ever fainted, passed out, or had a seizure suddenly, without warning, during exercise or a response to auditory triggers, such as doorbells, alarm clocks, and ringing telephones? That auditory trigger is very specific for Long QT. Long QT 2, especially Type 2, will have VF events with an auditory stimulus.

Have you ever had exercise induced chest pain or shortness of breath? OK, that's easy to remember. Are you or anyone in your-- are you related to anyone with a sudden unexplained death before age 50? That's easy to remember.

And then more specifically, are you related to anyone who's been diagnosed with the biggies? Hypertrophic cardiomyopathy, Long QT. Even if you just ask those two, I think that would capture a lot.

Just a little plug, here. The majority of syncope is benign. So if you can take the extra couple minutes to really convince yourself we're dealing with vasovagal syncope, we can save a lot of referrals actually. The majority of what we see is also vasovagal, but you're looking for the classic triggers, right?

Standing. I was standing or just stood up. Personal pain, the sight of blood, urination is a common one in kids. And combing-- what's called hair grooming syncope. So the classic story is the girl who's doing her hair, or mom's doing her hair under the hot lights in the bathroom, and she goes down.

The vasovagal events, you'll get a prodrome, usually dizziness, lightheadedness, nausea, feeling hot, leg weakness. Impaired vision and hearing is a common one. Muffled hearing, tunnel vision. You get a lot of tachycardia palpitations along with that because of the catechols that you kind of get a rash right before you go down.

And then post event, they're going to look pale. They're going to look kind of sweaty, diaphoretic, but they always should have a pretty quick recovery. They can be altered for a while, but they should be breathing. They should be pretty much, essentially, normal-ish vital signs. And a quick recovery to me is really important.

So the big question when you're screening, these days, is do these, do all athletes deserve an EKG? And there's a lot of-- we go to the electrical meetings every year. There's a lot of talks on this stuff.

You can find a lot of data for and against it. Other countries do it. Japan, Italy have had data that suggests it does improve the ability to detect these conditions before an event.

Of course, the argument being you can detect these before they have symptoms. A lot of them will have the EKG abnormalities as we've kind of gone through. So it makes sense, right?

The problem is it's very expensive. So you're going to screen everyone. You're going to get positives. That's positive equal work ups

And then one study that was recently done suggested that if you're going to do it for all US athletes, you're looking at $10 million per life saved. That's a big number, and it just makes it prohibitive to do it that way. So it's currently not endorsed by the AHA to do screening pre-participation EKGs on everybody as a blanket statement.

So this was a recent article done by-- the Sports Medicine Group often gets involved with this EKG screening, and the American Medical Society Sports Medicine put out a position statement which I kind of, I like the way they worded everything. So they acknowledged that there's not conclusive evidence to say universal screening should be done. But they also recognize that there's limitations in the EKG, and that the thing that I liked about it is they kind of said, well, it shouldn't be a choice between mandating national screening and no screening at all. How about a middle ground? How about provider autonomy to be able to do it if you feel like it makes sense for your practice?

And of course you have to have the infrastructure to do it, right? You have to have people who can interpret the EKG correctly. That's huge if you're going to commit to that. And maybe we should be screening certain populations, as opposed to everybody.

So I'm thinking, this is my proposal. Consider an EKG in any patient, not just an athlete. We talked about athletes, of course, in this talk, but anybody who has any of those four red flags.

And remember that this EKG has to be interpreted correctly. So if you're going to do it, make sure you don't make the decision to refer or restrict based on the computer read that has all this craziness on it. Because a lot of EKG abnormalities are normal athletic training findings.

So, this is a good example. Bradycardia, first degree AV block, right bundle branch block, early repolarization are very common in the athletes. We've gotten to the point where we don't even, and literature supports this, you don't even have to work up an athlete who has isolated LVH criteria on their EKG if they don't have any symptoms. That's a tough one to stare at that EKG and be like, yep, you're OK. But it's pretty well documented.

So this is just one bit of good news. We've talked about how a lot of these things are tough to pick up, and EKGs are really expensive to do on everyone. But this big European study looked at 15,000 athletes who had cardiac symptoms. They got EKGs on everyone.

When they were read it appropriately, the-- no kids with a normal EKG, despite their cardiac symptoms, had actual pathology when they got the full work up. So I think it is a nice tool to offer to parents if you have the ability to do that in your clinic. In the right circumstance, of course.

So I wanted to also touch upon what do you do if you get this call that one of your patients or a family member has had an arrest or a sudden death. You've got to think about the living relatives. So when I see patients, it's often a parent who has passed or, more rarely, fortunately, a sibling. And you got to think about, immediately, OK. How do I protect the rest of this family and kind of help them?

So one thing you want to ask is, was an EKG ever done on this patient while they were living, because that could be very helpful. EKGs for the primary relatives, so siblings, parents, and kids. Primary relatives, very helpful. We're in the era where molecular autopsy is very feasible. The problem is it's not universal.

So if a kid has a sudden arrest and they go to one county or another, they might get a completely different autopsy, medical evaluation. And they may not even have DNA that's stored appropriately for analysis. The paraffin-embedded tissue is not good for DNA. So you have to have somebody who knows how to collect it in a stable fashion, and that makes the world of death.

OK. Take home points. Many of the dangerous and potentially lethal cardiac sudden death conditions can be diagnosed by a careful history, exam, and EKG. I would argue that these standardized questionnaires are good because they give you a framework to be consistent, and they do basically touch on all the important things we think about. The key is screen everybody, not just athletes. Screen more than once, even.

It's tough in a primary care office, you can imagine, going through that whole thing. And it's easy for me, because that's all I'm thinking about, but I do understand that it is very difficult to do that in a primary care setting. Chest pain and excessive dyspnea, palpitations or syncope during exertion, or emotional stress should always be investigated further.

It's appropriate not to perform the million-dollar work up on a kid who has a history consistent with vasovagal syncope. Even if it happened on the football field, August, double days, standing there dizzy, went down, came right back, probably OK. It's hard to ignore those, but they're usually OK. So it's OK not to do-- pull the trigger on the million-dollar work up if you really are confident in a vasovagal mechanism, if their exam and their EKG and their family history is all reassuring.

And then lastly, if you have a patient or a family member who suffers a sudden death, advocate for an autopsy by a medical examiner who has a knowledge in these conditions and can get an adequate-- even if data is not there, make sure, if you get that phone call, make sure they collect DNA tissue sample for DNA analysis that can be stored long term, because that's huge.