

BroadcastMed | When in Doubt Kick Them Out? Long QT Syndrome Sports Guidelines

DOUG: Welcome back to another in our series of Mayo Clinic heart.org programs on late-breaking clinical information. Today we're going to be talking about Long QT syndromes in athletes. And I'm glad to have with us Mike Ackerman, who is a Professor of Medicine here, and is the Director of our program.

MIKE Thanks, Doug.

ACKERMAN:

DOUG: Yeah. Good to be here. But I have to say, Michael, that I'm a little bit surprised by this one. This obviously is coming out of the *JAMA* article that was out last week. Whenever I think about Long QT syndromes, I worry about swimmers and athletes.

So my question for you is, you've kind of broken with the typical dogma. And I'm just wondering your experience, why you were willing to make that step?

MIKE Well, Doug, it's great to be here and join you. And you're right, we did break with the dogma and the status quo.

ACKERMAN: Which, per guidelines, whether it's the Bethesda Conference guidelines in North America, or the European Society guidelines, it really has been for this disease, and many other diseases that could potentially cause sudden death, a mantra of if in doubt, kick them out.

And about eight years ago, really, I was starting to take care of more and more patients, and seeing the consequences of the guideline-based recommendation for disqualification where I thought to myself, can't we do better than this? These patients were coming here, really being shackled and being told, why don't you go into an 8-by-8 seclusion chamber and come out in 30, 40 years. And don't get your heart rate up past 80, don't do this, don't do that.

And it became clear to us that we ought to be able to do better, that we really should move the goalposts. The goalpost shouldn't be, for this disease, just preventing death. But it should be, how do we help these individuals regardless of their desires to live life large? And we thought we could help these athletes, in this case, live larger than what they had been told by talking to them about sports participation, letting them and their parents be part of the decision-making process.

DOUG: But you've seen, in guidelines, and in fact, if you look at the *Bethesda Seven*, and you wrote that.

MIKE Right.

ACKERMAN:

DOUG: Those guidelines, if you look at a Long QT of say, 470 or something like that, it seems a little bit strict. I have to give you that. But on the other hand, if you're going to be leaving the decision up to the patients, or in this case, the athletes, aren't they going to choose to play every time?

MIKE Not necessarily. That's part of what we call a mythbuster. The myth is out there that if you let them be part of the decision-making process, of course they'll always stay an athlete. Who wouldn't? And actually, we found that that's just not true.

That when you let them be part of this process, and it's not just the athlete and their parents, making in isolation. You and I, as the physician, we're part of that team. And we're part of that process.

I had about 157 athletes who were athletes at the time of their Mayo Clinic evaluation. And after their evaluation and the discussion and the consultation and the counseling, 27 of them, almost 20% said, you know what? It's not worth it to me. Hearing what I'm hearing about my risk, it's not worth it. I'll hang up the shoes and I'll move on to something else.

So we showed that about one in five of the athletes say, no more. And so I think it just shows that these families are fully capable of self disqualification. Nobody knows the stakes are higher than the parents and the athletes. They know what they're potentially sentencing their athlete to. They know that sports, that adrenaline, the thrill of victory, the agony of defeat, could be a trigger to the Long QT heart.

I mean, that's been published for a long, long time, that adrenalin is the common trigger to potentially irritate the Long QT heart into a potentially life-threatening arrhythmia. The parents know that. The athletes know that. So they're not making these decisions lightly.

DOUG: But sometimes you have somebody who has a marker that seems way out of proportion. Let's say a QT interval of greater than 550, and they've already had an event. So that one may be a bit more straightforward. Or you may have somebody who is aligning on the other side.

But when they actually ask you, we can't make the decision, what do you use to make that decision, or in your side of the counseling?

**MIKE
ACKERMAN:** I use what do they desire? How do they want to live life? So for example, in our *JAMA* article, of the 130 athletes who chose to remain an athlete, we had one athlete in over five years of follow up. So over 650 athlete years of observation, one athlete had an event.

Was he high risk? You bet he was. This was an

LQT1 boy, presented at around age five, with cardiac arrest. Had a defibrillator, had a defibrillator shock him during warm ups for soccer, warm up for baseball. And the family was counseled, it is not if your child will have another event. It will likely be when, even if you're taking your beta blocker well.

And in his case, he was very non-compliant. So he was dancing close to the fire. And the defibrillator activated, worked. And he's still an athlete.

Why? Because for this family, having a ball was really important to this child and to his development, and to his structure in the family. And we felt that our treatment program, even if he should have an event, ought to work. I should say, that he's also had a defibrillator activation when he was at a county fair and some girl ran up and stole his candy cane from him.

Now, we didn't restrict that activity from him. And it was really in his case, and in every patient's case, how do they want to live their life? And do we have a robust treatment program to enable that?

So some activities, of course, may be off right now. Not going to be a Top Gun Air Force pilot with this label, no matter how well I think their disease is treated. But staying engaged in the sports of life that they love, maybe we can do it.

We did see that those who chose disqualification compared to those who did not, their risk profile was slightly higher than those who stayed in the game and chose to stay in the game. And maybe that means our counseling directed them to be more inclined to say eh, I think I'll be done with the sport of choice.

DOUG: What does this mean about AEDs in a sports facility? What does this mean about AED-type vests?

**MIKE
ACKERMAN:** Yeah I think the AED becomes part of the treatment program. It's part of their sports safety gear. So for our athletes who choose to stay in the game, they have to have an AED. Not in the facility, but in their sports bag.

They need to have coaches and trainers who are willing to say, yeah, I'll accept you onto the team with this part of your sports gear. I view it really, as no different than the athlete with asthma. That could be life threatening.

They have their inhaler, the athlete with a peanut allergy. That could be life threatening. They have their EpiPen. Our athletes have, as an additional safety precaution, an external defibrillator. Which, by the way, has never been used in any of my athletes, but as part of the whole treatment program and their safety gear.

DOUG: So is it time to rewrite *Bethesda*? See, *Bethesda Seven* didn't come out that long ago.

**MIKE
ACKERMAN:** Yeah. Well, *Bethesda Conference* was published in 2005, led by Dr. Maron and Dr. Zipes. And in fact, it's been authorized to have another *Bethesda*. Because there's new information that's coming out all the time.

We acknowledged, in *Bethesda Conference Guidelines* in 2005, that it currently is grounded in the quote, "art of medicine," that there is relative lack of evidence. And now, as evidence emerges, it's time to relook at things. But it should be viewed that we can't restrict on a one-size-fits-all.

Maybe what we're learning in Long QT syndrome, and what we're observing in our program, can begin to give confidence to loosen a guideline recommendation for this disease in a patient who's seen in the right center, evaluated the right way, given the right information to provide a well-informed decision. But it doesn't mean, necessarily, that this recommendation applies to other genetic heart rhythm diseases.

Say, for example, hypertrophic cardiomyopathy, where worst-case scenario, an athlete with Long QT syndrome goes down in the field. An external defibrillator will rescue that Long QT athlete. That might not happen in an athlete with hypertrophic cardiomyopathy and a 3-centimeter septum, for example.

Getting electricity to them in 30 seconds, 3 minutes, may not be fast enough. So it just can't be a one-size-fits-all recommendation.

DOUG: And does that go back to the dynamic versus the static type of exercise? Are you saying that really those aren't the predictors, that those aren't the guides that we once thought that they were?

**MIKE
ACKERMAN:** Yeah I don't know that the type of sport is that predictive of a likelihood of a Long QT trigger or a CPVT trigger, or a hypertroph trigger. We tried, in the guidelines, to put the sports in their dynamic static box. And we tried to keep that safehaven of the class 1A. You too can be kicked out of everything. But we'll let you do billiards, bowling, cricket, curling, golf, and riflery.

Ping pong.

MIKE Ping pong is out. Ping pong is outside of the class 1A safehaven. But that was our attempt in the guidelines to keep some sports in play, in contrast to the ESC guidelines, which kicked the athlete out of every sport. Zero sports were created as an exception.

DOUG: So it kind of sounds like there's a little bit more room here to take a look at athletes and Long QT syndrome. It doesn't mean that we're just ignoring them. In this particular case, you've done a wonderful job of examining and qualifying them, and actually letting them be part of the decision.

It suggests that there may be some room to take a look at the guidelines that were published in the past, and rethink them and perhaps loosen them up just a little bit so that patients, these student athletes, may be in a better position to participate than what we thought before.

So this has been great, Michael. Thank you very much for being with us. And we look forward to seeing you again sometime in one of the next of our series at The Mayo Clinic heart.org sessions. Thanks for joining us.