When The Heart Refuses To Run

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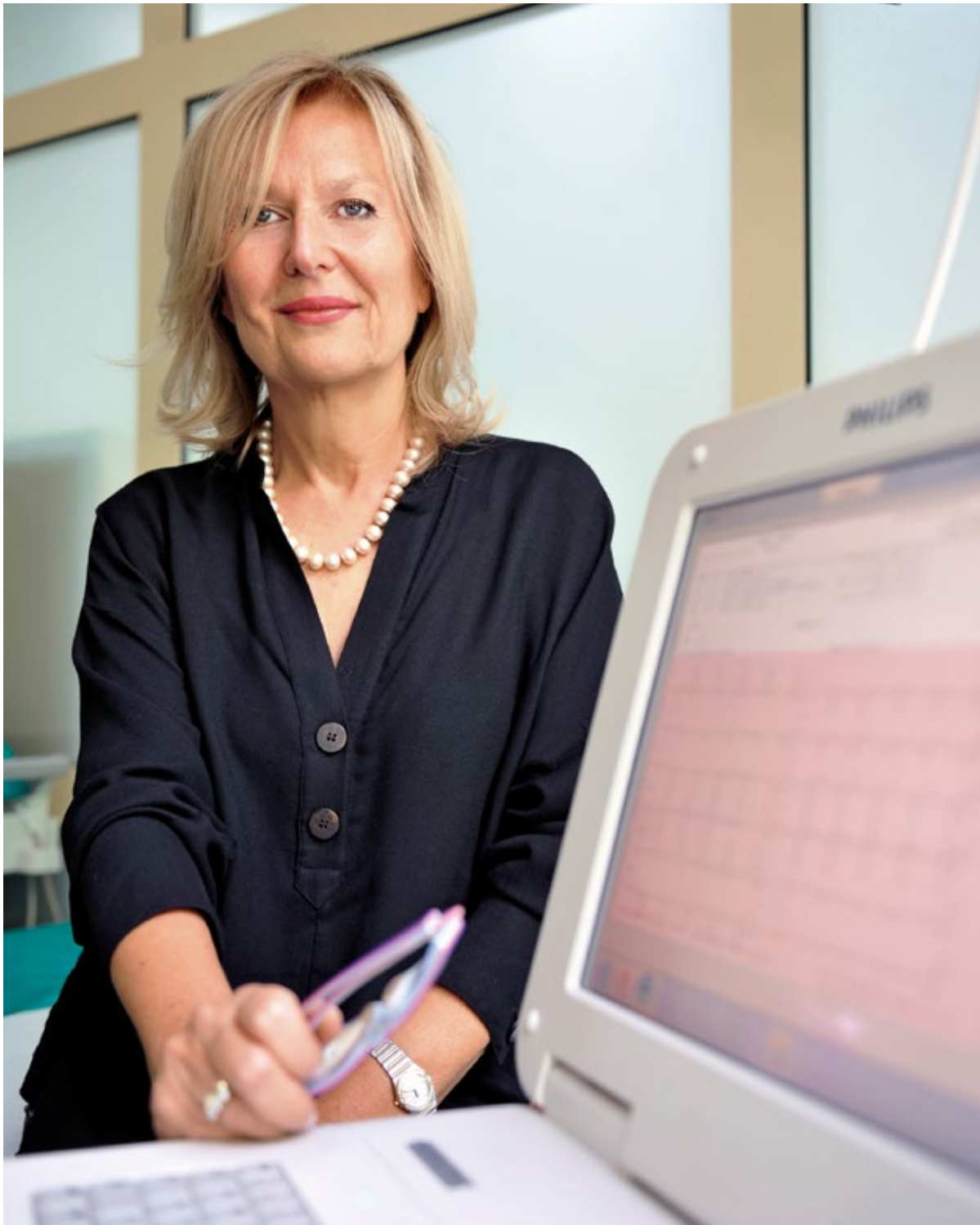
Prof. Katarzyna Biernacka from the Warsaw Institute of Cardiology explains sudden cardiac arrest and how to protect against it.



ACADEMIA: Last year three people died while running marathons in Poznań and Warsaw. Does this often occur during running events?

KATARZYNA BIERNACKA: Approximately one in 40,000 marathon runners will experience sudden cardiac arrest. A similar number of people run the Boston marathon every year, so technically we should expect one death each year. These are not alarming numbers. More people would die in car accidents at this time.

But here we are talking about the sudden deaths of healthy, young people, so is exercise really good for your health?

Italian, French and American studies have shown that sudden death occurs four times more often in young, apparently healthy, active people than among their peers who are not active in sports. So the question of whether sports are harmful to health seems entirely justified in this context. Between 1980 and 2000, Italian researchers worked on a project that revolutionized our perspective on this issue. They achieved absolutely incredible, even spectacular results, because after 20 years of observations they noted a lower risk of sudden death among athletes than among the normal population of young, healthy people.

How did they manage to do that?

They conducted much more thorough preliminary tests than is normally done when assessing the health of athletes. In addition to taking medical history and performing a routine examination, athletes underwent an ECG test, which helped detect many conditions that can cause arrhythmia during exercise. Some who were diagnosed with this condition were able to train again after receiving proper treatment, others had to give it up, but thanks to the diagnosis and subsequent treatment they did not die on the field or while running.

This groundbreaking study provided an answer to the question of whether practicing sports is harmful to the health. It turned out that it can only harm those who are not aware they have a heart disease predisposing them to dangerous arrhythmia, which may lead to sudden cardiac arrest.

So in order to avoid this you just need to do an ECG?

Yes. After the Italian discovery it was recommended that sports clubs introduce mandatory ECG screenings. But there is still a debate on this subject between Europe and the United States.

Why?

Americans believe that an ECG test is not needed because it does not help detect all heart diseases. Sports clubs are also resistant because they are convinced it may unnecessarily disqualify healthy athletes from competing. In a sense they are right, because in many cases an abnormal electrocardiogram may simply be due to overtraining and may not necessarily indicate a harmful condition at all. Of course these days cardiologists know how to distinguish between a healthy person's electrocardiogram and that of someone who has a health condition, but this interpretation is extremely difficult. There is always a margin of error, so there is a real risk of harming a healthy person if he is forbidden from playing sports. On the other hand, allowing someone who is at risk of sudden death to play sports is unacceptable.

Is there anything else that can be done besides an ECG test?

Of course, there are many other preventive methods, ranging from very simple test, such as a heart echo or the 24-hour Holter monitor, to much more complex ones like an MRI, an electrophysiology study, or a heart biopsy.

Who is required to undergo these tests?

Each country has its own rules when it comes to that. In Poland, only national team athletes and all athletes under the age of 21 are required to undergo such screening. If they are deemed to be at risk, they are not allowed to continue playing sports. Of course this is a much more complex issue than a regular patient-doctor relationship, because in this case it's not just a problem for an individual patient but an entire sports club, which wants a good player to continue competing at all costs. On the other hand, obviously no one wants an athlete to die on the field or on the track.

What types of health conditions are we talking about here?

It is usually the elderly or the sick who die suddenly, as the main cause of sudden death is coronary heart disease and heart failure. Coronary artery disease occurs much less frequently in young people, although in older athletes (those over 35) coronary heart disease is the main reason for sudden death. The main causes of sudden death and heart failure among young people are congenital heart defects and cardiomyopathies. These are conditions that generally prevent people from playing sports because they simply cause them to tire quickly. Limiting their exercise tolerance prevents them from playing competitive sports. Certain genetic diseases, which do not impair myocardial contractility, increase the risk of sudden cardiac death in sports. What types of diseases are they? They are genetic diseases that cause electrical instability. These include channelopathies and some cardiomyopathies. In essence, the heart remains structurally unchanged, appearing and contracting normally in the echo and other imaging tests, but under certain conditions there is a serious risk of arrhythmia.

How difficult is it to diagnose these conditions?
In the case of cardiomyopathy, especially hypertrophic cardiomyopathy, an ECG test should be sufficient to diagnose the problem. A heart echo is usually enough to confirm the diagnosis. Arrhythmogenic right ventricular cardiomyopathy is much more difficult to recognize. In this case, the left ventricle may not show any damage for many years, and it is the one responsible for exercise tolerance. So young people with severe right ventricular cardiomyopathy can function extremely well and have a very high exercise tolerance. They can play sports without knowing they are ill and they are the ones most at risk of sudden cardiac arrest.

In turn, channelopathies the strictly electrical diseases, pose a lower risk to athletes. These involve the disruption of ionic conduction in the myocardial cell membrane. Under certain circumstances, especially during adrenergic stimulation, which always occurs during physical exercise, ventricular fibrillation and sudden cardiac arrest may occur. These are primarily the long QT syndromes. There are many genetic types of long QT syndromes, but the three main ones, types 1, 2 and 3, are the most common. In Long QT syndrome type 1 arrhythmias can be triggered by exercise, and

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because the repolarization period, which is reflected on the ECG as the QT interval, is longer in cold conditions, people with this disorder are most at risk of sudden cardiac arrest while swimming. Another group of athletes at risk of dying while swimming are those who suffer from catecholaminergic polymorphic ventricular tachycardia. This condition can not be detected by a regular ECG. It requires additional testing. There are other channelopathies, such as the Short QT syndrome or Brugada syndrome. These can be easily detected by a regular ECG test.

I have to stress the importance of medical histories. If there is a history of sudden deaths among young people in the family, then additional tests should be performed on the athlete.

So are genetic factors involved here?
Yes, caused by a gene mutation. The arrhythmogenic right ventricular cardiomyopathy I mentioned before is a particular threat to athletes and for many reasons it should be mentioned, especially since it may not cause any symptoms for many years.

We are talking about cases of young people with good exercise tolerance (so there is no reason for them not to play sports), and then suddenly they pass out while playing football, which is usually preceded by heart palpitations. The ECG shows changes that can be misleading because healthy athletic people may have very similar changes, but once you know what to look for, you need to send this person for additional tests. Arrhythmogenic right ventricular cardiomyopathy is a huge threat to athletes because intense physical activity not only causes arrhythmia, but also worsens the condition. The harder they train, the faster the disease manifests itself, the arrhythmias become more dangerous and symptoms of heart failure appear earlier.

Is it possible to keep the heart functioning normally? Do all athletes suffering from cardiac diseases have to give up their sporting careers?
The Americans believe that if for three months an athlete with a Long QT syndrome does not present a syncope, he can return to the game with proper pharmacological treatment. I consider that very risky and in Europe we have a much more cautious approach, but following the right treatment and even providing the athlete with a cardioverter defibrillator, allows him to train normally. If the athlete has experienced a defibrillator discharge, it is up to him whether he wants to continue experiencing more such discharges, which are very unpleasant, if playing sports is so important to him that he will endure any inconvenience and wants to continue training.

A cardioverter defibrillator is a device that delivers a shock to stabilize the heart. Basically it saves a person's life during a dangerous episode of arrhythmia. Defibrillator discharges are so painful that patients usually don't want to experience one again, which is usually why those athletes who have experienced such a discharge don't return to playing sports. But there are also some who still run despite the discharges (they just aren't allowed to play contact sports, as that can damage the device).

Let's get back to arrhythmogenic right ventricular cardiomyopathy. Can medicine help people with this condition return to playing sports?
When it comes to arrhythmogenic right ventricular cardiomyopathy, we can not allow the athlete to continue training, as each hour of training worsens the condition of his right ventricle. It brings him closer to heart failure and severe arrhythmias. And we can not fit him with a defibrillator just so he can play sports. Implanting a defibrillator for medical reasons will not allow him to return to the field, because that is precisely

what causes this condition to worsen. Genetic studies done in athletes with arrhythmogenic right ventricular cardiomyopathy have shown a much less frequent occurrence of mutations responsible for its formation than in non-athletes who also have this condition. So the question arose, why does that happen?

It turned out that athletes do not have to carry a mutation to present this condition. This raises the question of whether there is an acquired (and therefore not genetically conditioned) form of arrhythmogenic right ventricular cardiomyopathy where physical exercise itself can cause damage to the right ventricle. To determine this, numerous experimental studies on animals were carried out. It turned out that it was easier to induce ventricular tachycardia in rats subjected to prolonged physical exertion than in rats that were not subjected to such exercise. Therefore, it was concluded that a form of acquired arrhythmogenic right ventricular cardiomyopathy most likely exists. It is not known why some people can play sports and never suffer damage to the right ventricle, while in others this damage is caused by prolonged physical exertion. Most likely this is due to some genetic structure still unknown to us.

Let's talk about what researchers do know.

We know that this is a disorder of intercellular connections, called desmosomes, which are found only in the heart and in the skin. These intercell connections are what give the skin and heart exceptional tensile strength. The skin must protect the entire interior of our body, while the heart must behave like a single cell. Conduction between cells must be very smooth and coordinated, so that the heart can contract properly and send blood to the large arteries. Therefore, cardiac cells (cardiomyocytes) must be very closely linked, and the desmosomes are responsible for this connection, so when they are damaged they cause arrhythmogenic right ventricular cardiomyopathy.

Mutations responsible for this disorder affect various desmosomal proteins. Damage to any of these proteins causes the desmosomes to weaken and when the heart pumps more blood during physical exercise, it stretches the ventricles of the heart and the desmosomes. If these are defective, they are more easily damaged and thus each time someone with arrhythmogenic right ventricular cardiomyopathy exercises the disease is exacerbated.

Separating the cardiac cells from each other disrupts ionic conduction, which are responsible for stimulating the cells, which can lead to dangerous arrhythmia. But that's not all. A cell separated from other cells is considered by the body to be superfluous. When this happens, one of the desmosomal proteins called plakophilin, travels to the cell nucleus with information that the cell has been separated from the cellular syncytium and the nucleus begins the process of apoptosis, killing the cell. Next, fibrous and fat tissues replace the dead cells forming a sort of scar, which is a substrate for reentrant ventricular tachycardia. If there is extensive damage, fibrosis occurs over a large area of the ventricle, causing severe contractility impairment in the later stages of the disease. So the only way to treat arrhythmogenic right ventricular cardiomyopathy in its advanced stages is a heart transplantation.

In athletes with no genetic mutations, however, it is still unclear what causes this serious illness. We simply don't know why some respond one way to exercise while others respond differently. We still need to find an answer to that. In any case, if any abnormalities in the right ventricle are detected during an ECG or echocardiogram, the athlete should not be allowed to practice sports for at least six months. If after this time everything returns to normal, these abnormalities were most likely caused by exercise.

So it is the athletes that are no longer allowed to train who end up coming to you?

I see athletes with abnormalities that sports cardiologists have deemed potentially dangerous to them, so they come to me for additional tests. Of course I also deal with non-athletes, but I must say that among all my patients with arrhythmogenic right ventricular cardiomyopathy, athletes are clearly predominant. The worst thing is when I have to tell an athlete that he is not allowed to continue training or compete. Sometimes they don't want to accept the fact that their career is over. I once saw a young boy who didn't want to accept that he was sick and, unfortunately, he died on the field during a football match.

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But there are also happy moments in my work, both for me and my patients, when I can tell them that it was a false alarm, that they are healthy, that the abnormalities in the ECG are harmless or that the arrhythmias we found can be cured. We have treated Olympic champions here for cardiac arrhythmia who were able to return to competing and continued to be very successful.

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