

Syndrom náhlého arytmiického úmrtí

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Přehled

Co je syndrom náhlé arytmiické smrti?

Syndrom náhlé arytmiické smrti je onemocnění srdce, které zdědíte (ve většině případů), které ovlivňuje elektrický systém vašeho srdce a způsobuje abnormální srdeční rytmus. Bez léčby vás to vystavuje riziku náhlé smrti.

Typy podmínek SADS

- **Brugadův syndrom** : polymorfní ventrikulární tachykardie.
- **Katecholaminergní polymorfní ventrikulární tachykardie (CPVT): ventrikulární tachykardie**
- **Syndrom dlouhého QT (LQTS)**: způsobuje, že vaše srdce tráví příliš dlouho repolarizací nebo dobíjením na další srdeční tep. Interval QT (prostor mezi značkami Q a T ve vlnovém vzoru na EKG) ukazuje, jak dlouho trvá, než se vaše komory stáhnou a poté se dobijí a připraví na další úder. Můžete vyvinout ventrikulární tachykardii nebo fibrilaci komor.
- **Syndrom krátkého QT (SQTS)**: vaše komory netráví dostatek času nabíjením na další dobu. To může způsobit fibrilaci síní, komorovou tachykardii nebo ventrikulární fibrilaci.
- **Timothyho syndrom**: typ syndromu dlouhého QT intervalu (velmi vzácné).
- **Wolff-Parkinson-White syndrom (WPW)**: tachykardie způsobená elektrickým vedením, ke kterému dochází mezi horní a dolní komorou vašeho srdce abnormální cestou (kromě normální cesty).

Jak častý je syndrom náhlé arytmiické smrti?

Jeden z častějších typů syndromu náhlé arytmiické smrti, syndrom dlouhého QT intervalu, se vyskytuje u jednoho z 2 000 novorozenců. Jiný typ, Brugadaův syndrom, se vyskytuje u méně než 1 % populace. Odhaduje se, že jeden z 10 000 lidí má CPVT.

Každý rok zemře asi 200 000 Američanů na náhlou zástavu srdce. Asi 4000 dětí a mladých dospělých zemře každý rok na SADS.

Příznaky a příčiny

Jaké jsou příznaky syndromu náhlé arytmiické smrti?

Ačkoli se příznaky mezi různými typy syndromu náhlé arytmiické smrti poněkud liší, běžné příznaky zahrnují:

- Mdloby nebo záchvat při cvičení, vzrušení nebo strach.
- Bolest na hrudi během cvičení.
- Dušnost při cvičení.

Brugadaův syndrom

Ačkoli mnoho lidí nemá příznaky, jiní mohou zaznamenat:

- Srdeční zástava, často ve spánku.
- Mdloby .
- Záchvaty.
- Neobvyklé dýchání.

Katecholaminergní polymorfní komorová tachykardie (CPVT)

Před zástavou srdce nemusí být žádné příznaky, ale někteří lidé mohou během cvičení omdlít.

Syndrom dlouhého QT (LQTS)

Asi polovina lidí s LQTS nemá příznaky, ale někteří mohou mít:

- Srdeční zástava z arytmiie zvané Torsades de Pointes .
- Mdloby.
- Záchvaty .

Syndrom krátkého QT (SQTS)

Nearly 40% of people with this syndrome don't have symptoms, but others with it may have:

- Palpitations.
- Fainting.
- Cardiac arrest.

Timothy syndrome

Symptoms of Timothy syndrome include:

- Fainting.
- Cardiac arrest.
- Webbed fingers and/or toes in some cases.
- Developmental issues.

Wolff-Parkinson-White syndrome (WPW)

Symptoms of Wolff-Parkinson-White syndrome include:

- Palpitations.
- Lightheadedness.
- Shortness of breath.
- Fainting.
- Seizures.

What causes sudden arrhythmic death syndrome?

When a young person dies suddenly and their heart has a normal structure, healthcare providers may suspect the child had an undiagnosed arrhythmia. These conditions that cause arrhythmias, listed above, are usually passed down from parent to child. The chance of passing on these conditions is variable, depending on the exact gene and exact part of the heart involved.

Diagnosis and Tests

How is sudden arrhythmic death syndrome diagnosed?

A healthcare provider may suspect this condition in a young person who's having symptoms and has a relative who died of unexplained, sudden death before age 40. Diagnostic methods vary by the type of sudden arrhythmic death syndrome.

Diagnostic tools may include:

- Electrocardiogram (EKG).
- Genetic testing.
- Treadmill stress test.
- Catecholamine provocation test (checking your heart's reaction to a drug your provider administers).
- Holter monitor.
- Echocardiogram.
- Fetal ultrasound.
- Cardiac catheterization diagnostic test.
- Electrophysiologic study.

Management and Treatment

How is sudden arrhythmic death syndrome treated?

Treatments vary depending on the type of sudden arrhythmic death syndrome.

Brugada syndrome

Treatment for Brugada syndrome includes:

- Implantable cardioverter defibrillator (ICD).
- Quinidine.
- Not taking medicine that blocks your heart's sodium channels.

Catecholaminergic polymorphic ventricular tachycardia (CPVT)

- People with CPVT take beta-blockers.
- Implantable cardioverter defibrillator (ICD).

Long QT syndrome (LQTS)

Treatments for Long QT syndrome may include medicines or procedures, including:

- Beta-blockers.
- Implantable cardioverter defibrillator (ICD).
- Left cardiac sympathetic denervation (a minimally invasive procedure that cuts the autonomic nerves that go to your heart).
- Not taking certain medicines that make your QT interval on an EKG longer.

Short QT syndrome (SQTS)

Short QT syndrome treatments include medication and a procedure:

- Implantable cardioverter defibrillator (ICD).
- Quinidine.

Timothy syndrome

Treatments include medication and a device:

- Beta-blockers.
- Implantable cardioverter defibrillator (ICD).
- Not taking medications that lengthen your QT interval on an EKG.

Wolff-Parkinson-White syndrome (WPW)

Medication may prevent WPW symptoms. Your healthcare provider can treat — and possibly cure — WPW with ablation therapy that gets rid of the extra conduction pathway. They can also prescribe medications called antiarrhythmics to reduce the chance of abnormal heart rhythm.

What are the side effects of the treatments?

Some people can't tolerate beta blockers their healthcare provider prescribed.

Left cardiac sympathetic denervation may cause:

- Abnormal sweating.
- Dry hands.
- Inconsistent temperatures or colors in the face.

However, this procedure can improve your quality of life.

ICDs deliver an electrical shock to get your heart back to a normal rhythm, but they can cause:

- Infection.
- Problems with the device functioning properly.

Prevention

How can I reduce my risk?

Having family members who know how to do CPR and have an automatic external defibrillator (AED) at home can give your loved one with SADS peace of mind. Being prepared can allow you to respond if they need help.

How can I prevent sudden arrhythmic death syndrome?

If someone in your family has sudden arrhythmic death syndrome, check with your healthcare provider about having others in your family tested for the genetic mutation. They can help you figure out who should get tested. Once a loved one has a diagnosis, they can take preventive steps to protect themselves with medicine or a procedure.

Outlook / Prognosis

What can I expect if I have sudden arrhythmic death syndrome?

The outlook for sudden arrhythmic death syndrome depends on the type you have and whether you get an early diagnosis and prompt treatment. This is a lifelong condition that can be fatal.

Wolff-Parkinson-White syndrome (WPW) treatments offer the most hope, as an ablation can fix the problem.

People who are getting treatment for Long QT syndrome rarely die suddenly. However, many people who could do well with a treatment called left cardiac sympathetic denervation don't have that option available to them. With conditions like Short QT syndrome, research is scarce because there are so few cases (about 200).

More research is needed to find the best treatments for everyone with sudden arrhythmic death syndrome.

Placement of an ICD can prevent sudden cardiac death by treating abnormal heart rhythms. While this does not make the condition go away, this is an effective way to reduce the chance of death from these conditions.

Living With

How do I take care of myself?

If your healthcare provider prescribed medicine for you, be sure to take it without missing doses. The same is true for a child that has SADS. They need their medicine at certain times and shouldn't miss a dose.

Always check with a health care provider or pharmacist before starting a new medication. Many medications, including common antibiotics and anti-nausea medications, can interact with cardiac medications your provider prescribes and can impact the QT segment, which impacts the chance of sudden death in long QT syndrome.

You'll also want to understand how your (or your child's) implantable cardioverter-defibrillator (ICD) works and have your provider check it regularly. Some medical procedures and everyday electronics can interfere with how well ICDs work, so make sure you know what to avoid.

If your child has SADS, check with your provider to see if it's ok for them to play sports with their condition.

When should I see my healthcare provider?

If you have SADS, regular checkups are important to make sure you're taking the right amount of medicine. If your child has SADS, they'll need to take larger doses of medicine as they grow and gain weight. This is why frequent checkups are especially important for children. See your provider twice a year if you have an ICD.

When should I go to the ER?

Call 911 and do CPR if someone is in cardiac arrest.

Go to the ER or call 911 if you:

- Feel as though you're about to faint, have lightheadedness or experience heart palpitations.
- Have a sudden onset of chest pain or difficulty breathing.

What questions should I ask my doctor?

- What type of sudden arrhythmic death syndrome do I have?
- What treatment do you recommend for my specific situation?
- Can you help me figure out who to test in my family?
- Do I need a defibrillator?
- Are there medications I should avoid with my condition?

A note from Cleveland Clinic

Se syndromem náhlé arytmické smrti vám informovanost o vaší diagnóze pomůže porozumět vašim možnostem. Spolupracujte se svým poskytovatelem zdravotní péče, abyste se rozhodli, která léčba je pro vás nebo vašeho blízkého nejlepší. Pravidelné kontroly jsou důležité, zvláště pokud máte ICD. Můžete posílit sebe a svou rodinu tím, že budete mít automatický externí defibrilátor (AED) a zajistíte, aby každý věděl, jak jej používat a provádět KPR.

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