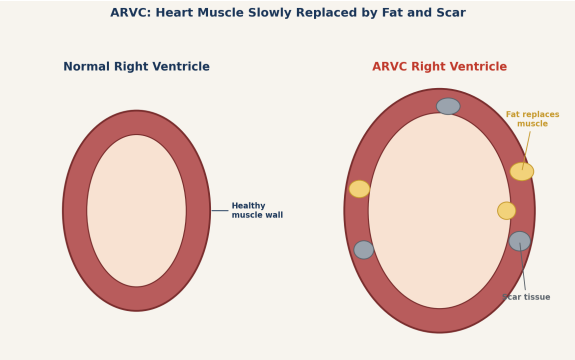
Understanding ARVC

Arrhythmogenic Right Ventricular Cardiomyopathy

An inherited heart-muscle disease that raises the risk of dangerous rhythms — and steps that protect you and your family.



Online: https://go.riasalimd.com/arvc-guide

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Names & Terms You Will Hear

Term	Meaning
ARVC	Arrhythmogenic right ventricular cardiomyopathy. An inherited disease where heart muscle is replaced by fat and scar.
ARVD	Arrhythmogenic right ventricular dysplasia. An older name for the same condition.
Arrhythmogenic cardiomyopathy (ACM)	A broader name. Sometimes the left ventricle is involved too, not just the right.
Right ventricle (RV)	The lower-right pumping chamber. It sends blood to the lungs. ARVC affects it the most.
Desmosome	The 'snap' that holds heart-muscle cells together. Faulty desmosome genes cause most ARVC.
Ventricular tachycardia (VT)	A dangerous fast rhythm that starts in the ventricles. ARVC scar can trigger it.
Ventricular fibrillation (VF)	A chaotic rhythm where the heart quivers and cannot pump. It causes cardiac arrest.
Sudden cardiac arrest (SCA)	The heart suddenly stops pumping. Without fast CPR and a shock, it is fatal in minutes.
Autosomal dominant	A way a gene is passed down. One changed copy is enough. Each child has a 50% chance of inheriting it.

If someone collapses and is not breathing normally — act fast:
1. Call 911 (or have someone call) and send for an AED.
2. Start Hands-Only CPR: push hard and fast on the center of the chest, 100-120 pushes per minute — about the beat of "Stayin' Alive." Do not stop.
3. Use an AED as soon as it arrives. Turn it on and follow the spoken steps. An AED will only shock a dangerous rhythm — it cannot shock a normal heart, so it is safe to use.

What Is ARVC?

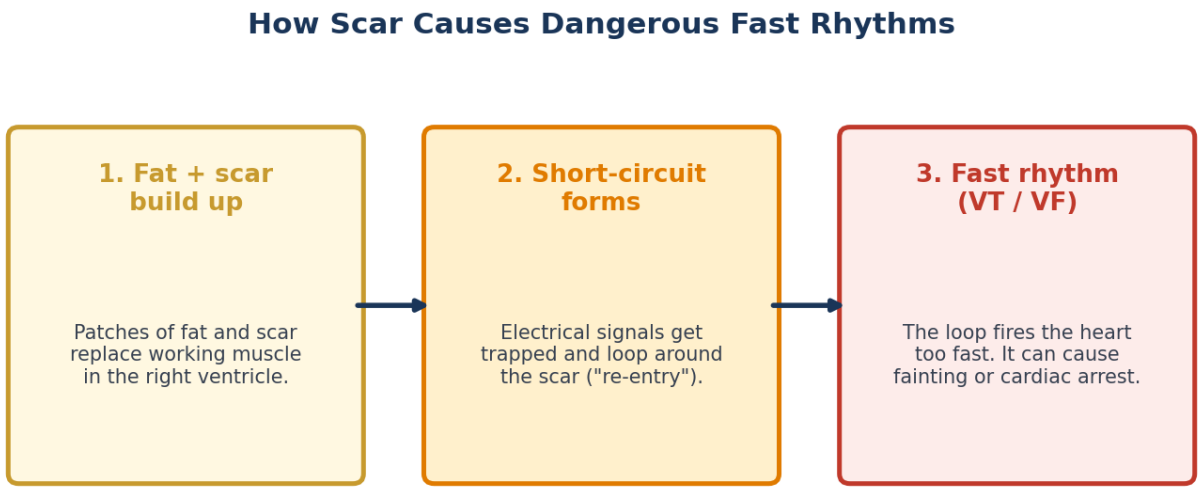
- ARVC is an **inherited (genetic) heart-muscle disease**. The muscle — mainly the right ventricle — is slowly replaced by **fat and scar tissue**.
- That scar creates electrical **short-circuits**. They can set off dangerous fast rhythms from the ventricles (VT or VF).
- ARVC is an **important cause of sudden cardiac arrest in young people and athletes**. Sometimes the arrest is the very first sign.
- It is uncommon — about **1 in 1,000 to 1 in 5,000 people**. But it is treatable once it is found, and prevention works.
- Symptoms often start between the **teens and age 40**. Many gene carriers feel fine for years before any sign appears.
- Over many years, the heart's pumping can also weaken, leading to **heart-failure symptoms** like breathlessness and swelling.
- ARVC is one of a family of inherited heart-muscle diseases. See our Dilated Cardiomyopathy guide for a related one.

How Scar Causes Dangerous Rhythms

- **The 'snaps' come loose.** Faulty desmosome genes weaken the connections between heart-muscle cells. Under stress, cells pull apart and die.
- **Fat and scar move in.** The body replaces the lost muscle with fat and scar. This is patchy, mostly in the right ventricle at first.
- **Scar blocks the signal.** Electricity flows smoothly through healthy muscle but gets stuck around scar. It can loop in a circle instead of moving straight through.
- **A short-circuit (re-entry) forms.** The looping signal fires the heart over and over, very fast. This is ventricular tachycardia (VT).
- **VT can turn into VF.** If the rhythm becomes chaotic (ventricular fibrillation), the heart quivers and stops pumping — sudden cardiac arrest.
- **Why an ICD helps:** It senses VT/VF and delivers a shock within seconds to reset the heart. See our VT/VF guide.

Why It Matters

- The biggest danger is a **dangerous fast rhythm** (VT/VF) that can cause fainting or sudden cardiac arrest — sometimes during or right after exercise.
- Because ARVC is **inherited**, your close relatives may carry the same gene. Screening them can catch the disease before it causes harm.
- ARVC is **one of the leading causes of sudden cardiac death in athletes and young adults** (about 10-15% of such deaths in some studies).
- Intense **endurance exercise can speed up the disease**. This makes ARVC different from most heart conditions — here, slowing down protects you.
- An **ICD (implantable defibrillator)** can stop a dangerous rhythm within seconds. For high-risk patients it is life-saving. See our Pacemakers and ICDs guide.
- With diagnosis, activity changes, medication, and the right device, **most people with ARVC live long lives**. The key is finding it and acting early.



This is why an ICD can be life-saving in ARVC: it stops a dangerous rhythm in seconds.

Three steps from scar to a dangerous rhythm: fat and scar build up, an electrical short-circuit forms, and the heart fires too fast (VT/VF).

Risk Factors

Some patients are more likely to develop this condition. Knowing your risk helps your care team take extra precautions.

Risk Factor	Why It Increases Risk
A desmosome gene change (PKP2, DSP, DSG2, DSC2, others)	These genes make the 'snaps' that hold heart cells together. A faulty copy lets cells pull apart and be replaced by fat and scar.
A parent, brother, sister, or child with ARVC	ARVC is usually autosomal dominant. Each first-degree relative has up to a 50% chance of carrying the same gene.
Sudden death in a young relative	Unexplained sudden death before age 40 in the family can be a sign of inherited ARVC. It deserves evaluation.
Intense endurance or competitive exercise	Long, hard exercise stresses the right ventricle. In gene carriers it speeds up scar and raises rhythm risk.
Fainting or palpitations with exertion	Fainting during or after exercise is a red flag in someone at risk. It needs a fast check.
Male sex	Men with ARVC tend to show the disease earlier and have a somewhat higher arrhythmia risk than women, though women are affected too.

Red-flag features and who should be screened. Red = highest urgency; amber = needs prompt attention.

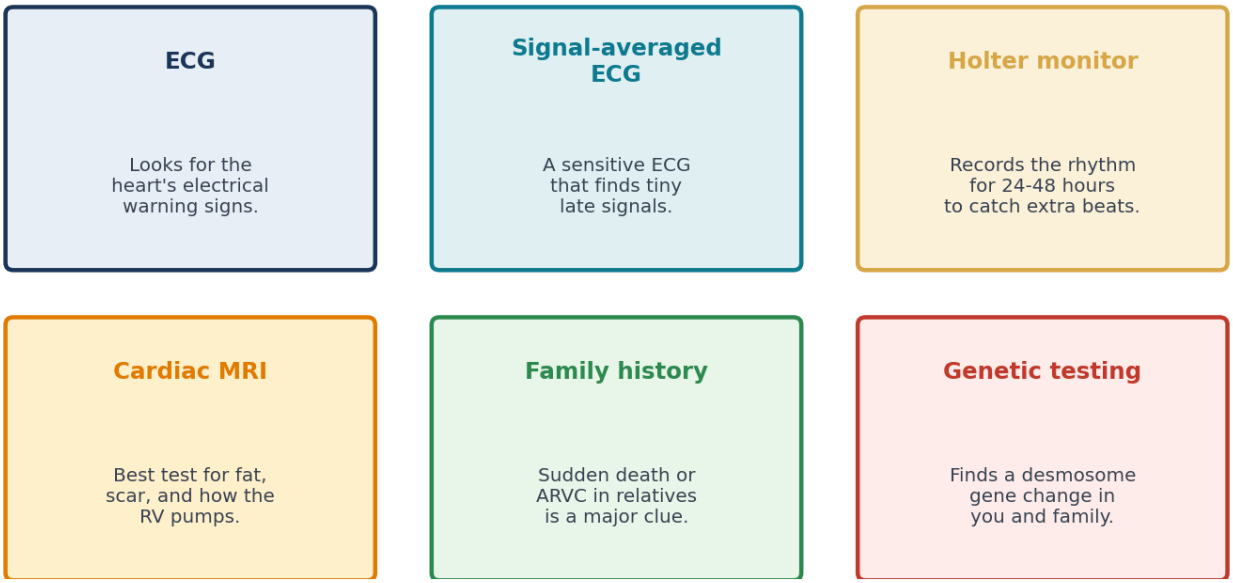
Red Flag	Why It Matters	What To Do
Fainting during or after exercise	A classic warning sign of a dangerous rhythm in ARVC	Stop activity; seek urgent heart evaluation
Racing or pounding heartbeat	May be VT starting from scar tissue	Sit or lie down; call us; consider 911 if it does not stop
A close relative with ARVC	Up to 50% chance you carry the same gene	Get heart screening and genetic counseling
Sudden death in a young relative	Can be the only sign of inherited disease	Tell your doctor; arrange family evaluation
Survived a cardiac arrest	Highest risk of another event	Almost always needs an ICD

Treatment Options

- **Avoid intense exercise:** This is a core part of treatment in ARVC. Cutting back on endurance and competitive sports lowers the chance of dangerous rhythms and slows the disease. Your cardiologist sets your safe limits.
- **Beta-blockers:** These slow the heart and calm extra beats. They are often the first medication used.
- **Antiarrhythmic drugs:** Medicines such as sotalol or amiodarone may be added to reduce VT episodes.
- **Catheter ablation:** A thin tube is threaded to the heart to burn or freeze the scar spots that cause VT. It is used for recurring VT, often alongside an ICD rather than instead of it.
- **ICD (implantable cardioverter-defibrillator):** A small device under the skin that watches the heart 24/7 and delivers a shock to stop VT/VF. It is recommended for those at high risk of sudden death. See our Pacemakers and ICDs guide.
- **Heart-failure medicines:** If the pumping weakens over time, standard heart-failure drugs help. Our Dilated Cardiomyopathy guide explains these.
- **Family screening and genetics:** Your relatives should be offered heart testing and, when a gene is known, genetic testing. See our Genetic Testing for Heart Disease guide.
- **Regular follow-up:** ARVC changes slowly over years. Repeat ECGs, monitors, and MRI scans track the disease and re-check your risk.

Putting the Pieces Together: How ARVC Is Diagnosed

No single test proves ARVC. Doctors add up clues from several tests ("Task Force Criteria").



ARVC is diagnosed by adding up clues from several tests — no single test proves it. ECG, signal-averaged ECG, Holter, cardiac MRI, family history, and genetics.

Why Exercise Restriction Matters (the Exception to "Exercise Is Good")

- **For most hearts, exercise is protective.** ARVC is different. Here, intense endurance exercise can make the disease worse.
- **Hard exercise stretches the right ventricle.** Over time this extra stress speeds up the loss of muscle and the build-up of fat and scar.
- **Gene carriers who train hard show the disease earlier.** Studies of family members found that endurance athletes developed ARVC sooner and had more dangerous rhythms.
- **Cutting back is proven to help.** Stepping away from competitive and endurance sport lowers arrhythmia risk and has saved lives in young athletes.
- **This does not mean 'do nothing.'** Light to moderate activity is usually safe and good for you. Your cardiologist will set your personal limits.
- **It applies to gene carriers too.** Even relatives who carry the gene but feel well are usually advised to avoid intense competitive sport.

Protecting Against Sudden Death: ICD and Family Screening

- **Know your risk.** Your team weighs your history (fainting, VT, a prior arrest), MRI findings, monitor results, and your specific gene to judge your risk.
- **ICD for high risk:** An implantable defibrillator is recommended when the risk of a dangerous rhythm is high. It stops VT/VF in seconds. See our Pacemakers and ICDs guide.
- **Survivors of cardiac arrest** almost always receive an ICD, because the risk of another event is high.
- **Screen your family.** First-degree relatives (parents, siblings, children) should be offered an ECG, an echo or MRI, and a monitor — even if they feel well.
- **Genetic testing guides the family.** If a gene change is found in you, relatives can be tested for that exact change. See our Genetic Testing guide.
- **Be CPR- and AED-ready.** Make sure the people around you know Hands-Only CPR and where the nearest AED is. See our Sudden Cardiac Arrest guide.

Comfort Measures at Home (No Medication Needed)

These simple steps support healing and ease symptoms. Use them alongside any medication your doctor prescribes.

- Follow your cardiologist's exercise limits. Light to moderate activity (like easy walking) is usually fine; intense endurance training is not.
- Avoid competitive sports unless your cardiologist has cleared you. This is one of the most powerful steps you can take.
- Limit alcohol and avoid stimulant drugs and high-dose caffeine, which can trigger extra beats.
- Learn your warning signs: a racing heartbeat, lightheadedness, or near-fainting. Sit or lie down right away and call us.
- Make sure close family members learn Hands-Only CPR and how to use an AED. They are your first line of help.
- Keep all follow-up appointments. ARVC is monitored over a lifetime, even when you feel completely well.

Risks, Benefits, and Alternatives

Every choice has trade-offs. Use this table to start the shared decision conversation.

Option	Risks	Benefits	Alternatives
Exercise restriction	Loss of competitive sport and intense training. Can be hard emotionally and socially.	Lowers the risk of dangerous rhythms and slows the disease. One of the few proven steps that changes the course of ARVC.	Supervised lighter activity. A frank talk with your cardiologist about what level is safe for you.
Beta-blocker	Tiredness, low heart rate, low blood pressure. Do not stop suddenly.	Calms extra beats and may reduce VT triggers. Well-tolerated by most people.	Antiarrhythmic drug if beta-blockers are not enough.
Antiarrhythmic drug (e.g., sotalol, amiodarone)	Can have side effects; amiodarone may affect the lungs, thyroid, or liver with long use.	Reduces how often VT happens. Helpful when extra beats break through a beta-blocker.	Catheter ablation. Adjusting the beta-blocker.
Catheter ablation for VT	Bruising, bleeding, rare heart injury. VT can come back because new scar keeps forming.	Targets the exact scar spots driving VT. Can greatly cut VT episodes and ICD shocks.	More medication. An ICD remains in place for protection.
ICD (defibrillator)	Surgery and infection risk (under 1-2%). Lead problems over time. Possible inappropriate shocks.	Stops VT/VF within seconds — directly prevents sudden cardiac death in high-risk patients.	Medications and ablation alone if risk is judged low (a shared decision with your team).

Common Misconceptions

Myth	Reality
"I feel fine, so my heart must be healthy."	ARVC can be silent for years. Many gene carriers feel completely well right up until a dangerous rhythm appears. Feeling fine does not rule it out.
"Exercise is always good for the heart, so more is better."	ARVC is the exception. In this disease, intense endurance exercise can speed up the damage and trigger dangerous rhythms. Here, easing back protects you.
"Only the person diagnosed needs to worry."	ARVC runs in families. Parents, siblings, and children should be offered heart screening — and genetic testing when a gene is known — even if they feel fine.
"If I get an ICD, I am cured."	An ICD is a safety net that stops a dangerous rhythm. It does not stop the disease. Exercise limits, medicine, and follow-up still matter.
"A normal ECG or one normal test means I do not have ARVC."	No single test proves or rules out ARVC. Doctors combine several tests — ECG, MRI, Holter, family history, and genetics — to reach a diagnosis.
"ARVC only affects the right side of the heart."	It usually starts on the right, but the left ventricle can be involved too. That is why it is sometimes called arrhythmogenic cardiomyopathy.

Possible Complications

Knowing what can go wrong helps you spot problems early. Most complications are uncommon, especially with treatment.

Where / What	What Can Happen
Dangerous fast rhythms (VT/VF)	Scar short-circuits can fire the heart far too fast, causing fainting or sudden cardiac arrest. See our VT/VF guide.
Sudden cardiac arrest	The heart suddenly stops pumping. Without immediate CPR and a shock it is fatal. It can be the first sign of ARVC.
Heart failure	After many years, the weakened muscle may not pump well, causing breathlessness, swelling, and fatigue.
Both ventricles affected	The disease can spread to the left ventricle, which raises heart-failure and rhythm risk.
ICD shocks and lead issues	A needed device can deliver shocks (sometimes when not needed) and may have lead problems over the years.
Anxiety and lifestyle impact	A diagnosis, exercise limits, and living with an ICD can be stressful. Support and counseling help.

Points to Know

If you remember nothing else, remember these key points.

- ARVC is an inherited disease where right-ventricle muscle is replaced by fat and scar.
- The scar creates short-circuits that can trigger dangerous fast rhythms (VT/VF) and sudden cardiac arrest.
- It is a leading cause of sudden cardiac death in young people and athletes — recognition saves lives.
- Intense endurance exercise can speed up ARVC. Activity restriction is part of treatment — the exception to 'exercise is good.'
- Diagnosis adds up clues from several tests: ECG, signal-averaged ECG, Holter, cardiac MRI, family history, and genetics.
- Treatment can include beta-blockers, antiarrhythmic drugs, catheter ablation for VT, and an ICD for high-risk patients.
- ARVC is inherited (usually autosomal dominant). First-degree relatives should be offered heart and genetic screening.
- Make sure your family knows Hands-Only CPR and how to use an AED — they are your first line of defense.

When to Call Us - and When to Call 911

If you are not sure, call. We would rather hear from you twice than miss a real problem.

- **Call 911** if someone collapses, is not responsive, and is not breathing normally — start CPR and send for an AED right away.
- **Call 911** for fainting, chest pain, or a fast pounding heartbeat that does not stop, especially during or after exercise.
- **Call 911** if you have an ICD and it delivers a shock and you feel unwell — or if it shocks more than once.
- **Call our office within 24 hours** if your ICD delivers a single shock and you feel fine afterward.
- **Call our office** for new or more frequent palpitations, lightheadedness, or near-fainting.
- **Call our office** for new breathlessness, leg swelling, or trouble keeping up with normal activity.
- **Call our office** if a close relative is newly diagnosed with ARVC or has a sudden, unexplained death — your family needs screening.
- **Call our office** before starting any new sport or exercise program so we can confirm what is safe for you.

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Trusted Resources

The links below are the trusted resources Dr. Ali used to write this guide and that he recommends you visit for more information. Each is a clickable link in the digital version and a Short.io tracked URL on the printed version.

- Cleveland Clinic — ARVC - Patient-friendly overview of ARVC causes, symptoms, and treatment.
- Mayo Clinic — ARVC - Symptoms, inherited basis, diagnosis, and management.
- Johns Hopkins ARVD/C Program - Overview from a leading ARVC specialty center and registry.
- MedlinePlus / NIH — ARVC Genetics - Plain-language NIH resource on how ARVC is inherited.
- AHA Hands-Only CPR - How to give Hands-Only CPR — push hard and fast on the center of the chest.
- PulsePoint AED Finder - App that helps you find the nearest AED and alerts CPR-trained bystanders.
- Our VT/VF Guide - Companion guide on ventricular tachycardia and fibrillation — the rhythms ARVC can cause.
- Our Sudden Cardiac Arrest Guide - Companion guide on cardiac arrest, CPR, and AEDs.
- Our Genetic Testing for Heart Disease Guide - How inherited heart conditions are tested and how families are screened.
- Our Pacemakers and ICDs Guide - What an ICD is, how it protects you, and what to expect.

Sources Used to Build This Guide

Every recommendation in this guide is grounded in one or more of the studies, guidelines, or trusted resources below. Short links are tracked so Dr. Ali can see which references patients are reading; nothing personally identifying is captured.

- Cleveland Clinic — Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) *[Patient Education]*
  Plain-language overview of fatty/fibrous replacement of the right ventricle, arrhythmia and sudden-death risk, exercise restriction, and ICD role.
- Mayo Clinic — Arrhythmogenic Right Ventricular Dysplasia (ARVC) *[Patient Education]*
  Patient-facing symptoms, inherited basis, diagnosis (MRI, ECG), and treatment including ICD and activity limits.
- American Heart Association — Cardiomyopathy Types (ARVC) *[Patient Education]*
  Frames ARVC among inherited cardiomyopathies and the link to ventricular arrhythmia and sudden cardiac death.
- 2019 HRS Expert Consensus Statement on Evaluation, Risk Stratification, and Management of Arrhythmogenic Cardiomyopathy *[Guideline]*
  Authoritative basis for diagnostic Task Force criteria, genetic/family screening, exercise counseling, and ICD indications.
- Marcus et al. — Diagnosis of ARVC/Dysplasia: Proposed Modification of the Task Force Criteria (2010) *[Guideline]*
  The 2010 modified Task Force Criteria — the multi-part diagnostic scoring system (imaging, ECG, arrhythmia, tissue, family history/genetics) referenced throughout this guide.
- Arrhythmogenic Right Ventricular Cardiomyopathy: A Comprehensive Review (PMC, 2025) *[Review]*
  Prevalence (~1 in 1,000 to 1 in 5,000), 10-15% of sudden cardiac death in the young and athletes, desmosome genetics (PKP2, DSP, DSG2, DSC2), and the exercise-progression link.
- James et al. — Exercise Increases Penetrance and Arrhythmic Risk in ARVD/C Desmosomal Mutation Carriers (JACC 2013) *[Study]*
  Key evidence that endurance and competitive exercise accelerate ARVC onset and raise ventricular-arrhythmia risk in gene carriers - the basis for activity restriction.
- Johns Hopkins Medicine — ARVD/C (Arrhythmogenic Right Ventricular Dysplasia) Overview *[Patient Education]*
  Specialist-program patient overview from a leading ARVC registry - diagnosis, family screening, and management framing.
- MedlinePlus / NIH — Arrhythmogenic Right Ventricular Cardiomyopathy (Genetics) *[Patient Education]*
  NIH plain-language genetics resource: autosomal dominant inheritance, incomplete penetrance, and first-degree relative screening.

About This Guide

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Reviewer note: Reviewed against AHA, Mayo Clinic, Cleveland Clinic, and Johns Hopkins ARVD/C patient pages. This guide adds: a plain-language scar-to-short-circuit mechanism, the exercise-restriction exception explained for patients, and a family-screening and sudden-death-prevention pathway with cross-links to our genetics and ICD guides.

Disclaimer

This guide is for educational purposes only and does not replace medical advice from your physician. Recommendations may not apply to every patient. Always follow the specific instructions of Dr. Ali and your care team. In an emergency call 911 or go to the nearest emergency department.

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