



# *Loeys-Dietz Syndrome Emergency Preparedness*



© 2024 LOEYS-DIETZ SYNDROME FOUNDATION CANADA  
ALL RIGHTS RESERVED.

FOR MORE INFORMATION ABOUT LDS VISIT

[WWW.LOEYSDIETZCANADA.ORG](http://WWW.LOEYSDIETZCANADA.ORG)

# What is this booklet for?

When you are living with a family member with Loeys-Dietz Syndrome, or having Loeys-Dietz syndrome, an emergency can arise at any moment. This booklet aims to provide you and your loved ones with the tools you need to feel prepared for a possible emergency.

## Medical Disclaimer

The information provided here is meant to empower individuals to make informed decisions – it does not, however, replace a reader's relationship with their doctor.

The information outlined in this booklet is for general use only. Please speak with a qualified healthcare professional before making medical decisions.

This resource has been researched, reviewed, and vetted by subject experts who have worked hard to ensure that the information provided is accurate and reliable; however, we cannot guarantee that it is error-free or complete.

Research for LDS is ongoing.

The Loeys-Dietz Syndrome Foundation Canada is not responsible for the quality of the information or services provided by organizations mentioned on [loeysdietzcanada.org](http://loeysdietzcanada.org), nor do we endorse any service, product, treatment, or therapy.

# Table of Contents

1. Emergency signs and symptoms -----	4
a. Hollow Organ Rupture -----	4
b. Ocular Detachment -----	5
c. Pneumothorax -----	5
d. Dissection -----	6
e. Differences Between Sexes -----	7
f. Recurrent aneurysms -----	7
2. Other Urgent Situations -----	7
a. Fractures -----	7
3. Important Information for Medical Practitioners -----	8
a. Medications that should not be prescribed -----	9
b. Diagnosis -----	9
c. Cervical Spine Instability -----	10
d. Patient Flagging -----	10
4. Emergency Preparedness -----	11
a. Calling an ambulance -----	11
b. Steps for emergency preparedness -----	12
5. Emergency Preparedness for Parents with School-Aged Children -----	15
6. Emergency Preparedness for Parents of Adult Children Attending Post- Secondary Education -----	16
7. Wearable Medical Alert Devices -----	18
a. Information to include -----	18
b. Universal ID -----	19
c. QR codes -----	20
d. Electronic Alert Systems -----	20
8. Advanced Directives -----	21
9. Raising Awareness -----	21
a. Zebra Lanyard -----	21
b. Emergency ID Card -----	22
10. Tips from Doctors, Nurses, Patients and First Responders -----	23
11. Resources -----	24

# Emergency Signs and Symptoms

The following 4 situations are the most common reasons why people with LDS might need emergency care. They can be remembered with the acronym HOP-D.

**H:** Hollow Organ Rupture  
**O:** Ocular Detachment  
**P:** Pneumothorax  
**D:** Dissection

If any of the symptoms described here occur, it's important to seek medical help right away.

## HOLLOW ORGAN RUPTURE

People with LDS are at higher risk for ruptures in organs like the intestines, uterus, and spleen. Here's what to look for:

### Intestinal rupture

Signs you may notice if intestines rupture:

- Severe abdominal cramping
- Bloating or swollen abdomen
- Fever or chills
- Nausea or vomiting
- Pain or tenderness when you touch your abdomen

## Uterine Rupture

Uterine rupture tends to happen during pregnancy. If you are pregnant, or planning to be, talk to your doctor about how to reduce the risks. For more information, please refer to the [Family Planning with LDS](#) guide.

## Spleen rupture

Signs of a spleen rupture may include:

- Pain in the upper left side of the abdomen
- Tenderness when touching the upper left abdomen
- Pain in the left shoulder
- Confusion
- Feeling light-headed or dizzy

## OCULAR DETACHMENT

In people with LDS, the connective tissue supporting the retina can be weaker, which may lead to a detachment. This can cause sudden changes in vision, such as:

- Dots or lines floating in your vision
- Flashes of light
- A dark curtain or shadow moving across your field of vision
- Blurry vision

## PNEUMOTHORAX (COLLAPSED LUNG)

A spontaneous lung collapse, or pneumothorax, may result in:

- Sharp chest or shoulder pain
  - The pain might worsen with a deep breath or cough
- Shortness of breath, possibly leading to nasal flaring (widening of the nostrils)
- A blueish tint to the skin (from lack of oxygen)
- Feeling light-headed
- Shock or sudden collapse

## DISSECTION

This is the most dangerous complication for people living with LDS. It occurs when the innermost layer of the aorta (the body's main artery) tears, causing blood to pool and create a weak area that could eventually rupture.

**Dissection symptoms may resemble those of a heart attack, but there are specific signs to watch for:**

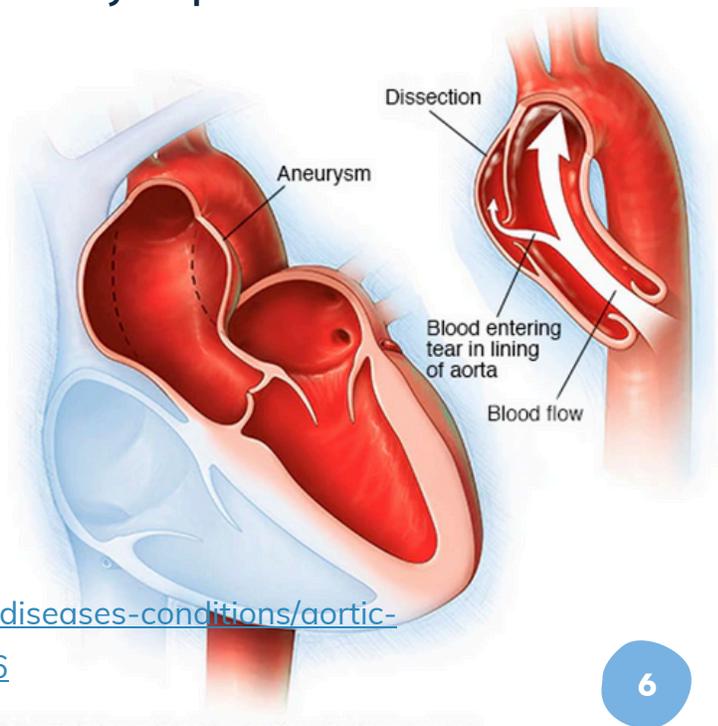
- Inconsistent blood pressure: one arm may have a much higher or lower blood pressure than the other (a difference of more than 20mmHg)
- Pulse deficit (peripheral pulse): you may notice a weaker pulse in some areas, like your wrists or ankles.
- Chest pain with a “tearing” quality radiating to the back between the shoulder blades
- Paresthesia: a prickling or tingling sensation in the limbs.
- Radiating pain: the pain will spread towards the location of the dissection

**Other heart attack-like symptoms that may be present include:**

- Chest, abdominal and back pain
- Limb weakness
- Fainting (Syncope)
- Shortness of breath

If these symptoms occur, it's a medical emergency and requires immediate attention.

Picture Source: <https://www.mayoclinic.org/diseases-conditions/aortic-dissection/symptoms-causes/syc-20369496>



## DIFFERENCES BETWEEN SEXES

In biologically female individuals, symptoms of dissection tend to develop more slowly. This can make diagnosis challenging because the symptoms may be mild, short-lived, or not obvious enough to meet the criteria for a clear diagnosis. As a result, the risk of missing a dissection diagnosis can be higher, even through lab tests or routine checks.

## RECURRENT DISSECTIONS

A dissection can happen again, either in the same location or in a new part of the body. Keeping detailed records of previous dissections is important. This information should be readily accessible, both in your home (on inner home signage) and in a medical binder, to help medical professionals quickly understand your history and provide appropriate care.

More information about [inner home signage](#) can be found on page 23, and [medical binder](#) can be found at this link: [https://ontariocaregiver.ca/wp-content/uploads/2024/02/Healthcare-Journey-Binder-ENGLISH\\_FINAL-es.pdf](https://ontariocaregiver.ca/wp-content/uploads/2024/02/Healthcare-Journey-Binder-ENGLISH_FINAL-es.pdf)

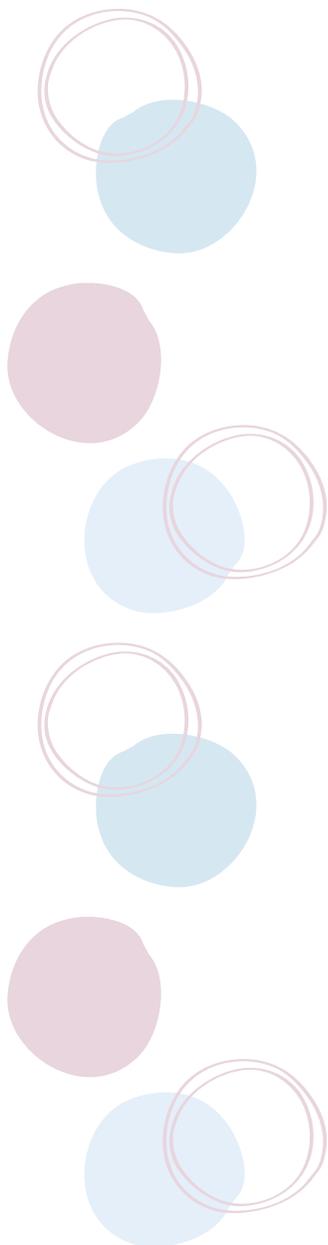
# Other Urgent Situations

## FRACTURES

Fractures might require immediate medical attention or could be addressed in a non-emergency setting, depending on the severity: Individuals with LDS often have lower bone density, making their bones more fragile and prone to fractures, similar to people with osteoporosis. On average, people with LDS are at a 20% higher risk of having a fracture by the age of 14. Because of this, after any fall or accident, it's important to be cautious. Seeking medical attention promptly will help prevent the injury from worsening and ensure proper care is given.

# Important Information for your Medical Practitioners

The John Ritter Foundation has a good overview of important information, referred to as the Ritter Rules: <https://johnritterfoundation.org/your-aortic-health/ritter-rules/>. These rules outline essential steps for preventing, diagnosing, and treating thoracic aortic dissection.



## Ritter Rules

**Urgency**

+

**Pain**

+

**Misdiagnosis**

+

**Imaging**

+

**Risk Factors**

+

**Triggers**

+

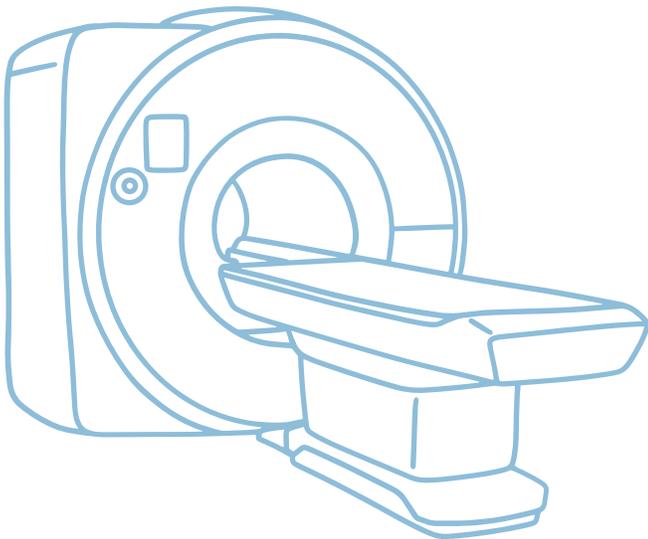
**Prevention**

+

## MEDICATIONS THAT SHOULD NOT BE PRESCRIBED

- People with LDS should avoid the following medications due to their potential to increase the risk of aortic dissection or complications:
- Decongestants (for regular use)
- Triptans (used for managing headaches or migraines)
- Fluoroquinolone antibiotics (associated with increased risk of dissection), including:
  - Ciprofloxacin
  - Moxifloxacin
  - Levofloxacin
  - Norfloxacin
  - Ofloxacin

## DIAGNOSIS OF AORTIC DISSECTION



### Aortic dissection **CAN** be ruled out using:

- Computed Tomography (CT)
- Magnetic Resonance Imaging (MRI)
- Transesophageal echocardiogram

### Diagnosis of aortic dissection **CANNOT** be ruled out through:

- Chest X-ray
- EKG (Electrocardiogram)

\*Important note for biologically female patients: Symptoms of dissection may develop more slowly in women, which means longer periods of observation might be necessary before ruling out the possibility of dissection. Refer to the section on the [Differences Between Sexes](#) for more details.

While the symptoms of a heart attack and aortic dissection can be similar, there are important differences to help distinguish between the two. For details, refer to the [Dissection](#) section.

## CERVICAL SPINE INSTABILITY

- Approximately 76% of individuals with Loeys-Dietz syndrome exhibit cervical spine instability, particularly between C1-C3 vertebrae.
- When intubating patients with LDS, extra precautions should be taken to avoid dislocation or further injury to the spine.

# Patient Flagging Systems

Given the wide variety of electronic medical record (EMR) systems in use across different medical institutions, this section provides general guidance on creating patient flags, with specific resources for the EPIC EMR system. Custom flags help ensure that critical health information is visible to the care team. Staff are encouraged to learn how to create these warning labels within their own institutions' EMR systems. A patient can visit their local medical institutions to have flags added to the EMR in preparation for a potential emergency.

### 1. Acute Care Plan Flag

- This patient flag is used to alert the care team to important health information in emergency situations, where time is critical, and key details might be missed. This flag can include steps for managing a patient's condition, such as inserting a warning to use a CT scan to rule out aortic dissection before performing other diagnostics. Given the high risk of death from dissection if not promptly treated, this flag helps prioritize the correct procedures.

\*Note\*: Procedures for flagging a patient and evaluating the need for a flag are specific to each medical institution. Always follow your medical facility's protocols for flagging high-risk patients and communicating this to the care team.

## 2. FYI Flag

- In the EPIC Electronic Medical Record (EMR) system, an FYI flag can be added to a patient record, to communicate key health information to the care team. This flag is especially useful for teams made up of multiple healthcare providers, ensuring that vital information isn't overlooked. This guide can assist medical professionals in setting up customized flags for important patient information: [link to PDF guide.](#)

# Emergency Preparedness

Emergencies can be stressful, but being prepared with a plan can help you and your family respond effectively. Here's a guide to help you be ready:

## WHEN TO CALL AN AMBULANCE

Consider calling an ambulance in these situations:

- Loss of consciousness
- Sudden confusion, trouble speaking, or dizziness
- Severe, persistent chest pain
- Sudden shortness of breath
- Fractured or broken bones
- Uncontrollable bleeding
- Sudden vision problems
- Sudden weakness or numbness
- Severe pain
- Severe burns or allergic reactions

**\*This is not an exhaustive list; use your discretion based on the situation.\***

## STEPS FOR EMERGENCY PREPAREDNESS

### 1. Discuss Emergency Scenarios

- a. Talk with your family about different emergencies, like an aortic dissection, versus a fracture
- b. Decide when you would call an ambulance, or when it would be safe to drive to the hospital yourself

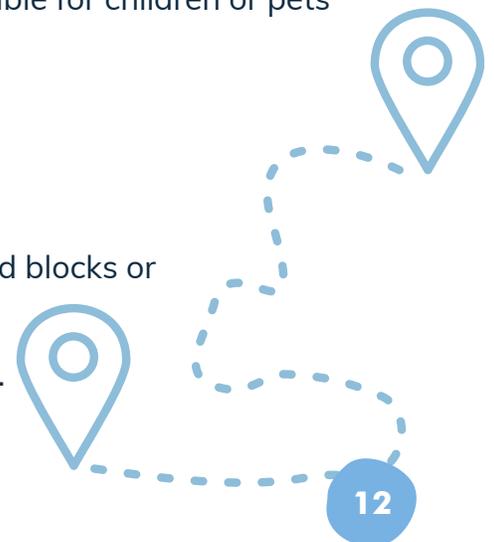
### 2. Assign Family Roles

Based on your personal circumstances, family members or other loved ones can fulfill predetermined roles in the event of an emergency, such as:

- a. **Leader:** Takes control, calls 911 or decides whether to drive to the hospital: Delegate tasks to others
- b. **Caregiver:** Provides basic care, including medications and monitoring health changes; Shares the patient's medical history with arriving Emergency Medical Services (EMS); Accompanies the patient to the hospital
- c. **Driver:** Drives the patient to the nearest emergency room, ensuring the fastest route; May handle dropping off children or pets as needed
- d. **Liaison:** Ensures the emergency bag is ready and gives it to the caregiver travelling to the hospital or to the EMS; Communicates updates to family, primary doctor and others; Communicates with parties responsible for children or pets care

### 3. Plan Emergency Routes

- a. Plan more than one route to the hospital in case of road blocks or accidents
- b. Have more than one emergency room option available. You may opt to choose a hospital with greater expertise in handling aortic dissection cases



## 4. Plan Care for Dependents

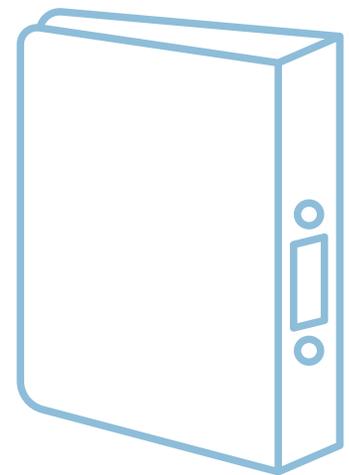
- a. Arrange for someone to care for children or pets during an emergency. Have contact information available.
- b. Assign the Liaison to communicate with the dependants' caregivers

## 5. Create an emergency kit

Prepare a kit with these essentials:

- a. Medical Binder or Folder. Ensure this binder or folder is updated after any imaging, medication change, or change in treatment. Store the medical binder in a visible place like above the fridge or in the medicine cabinet, where first responders are trained to check. You may also consider keeping a copy in your hospital bag. Include key information and documents like:
  - i. Basic information, name, date of birth, sex, etc.
  - ii. Copies of ID (health care, driver's license)
  - iii. Current medications and allergies
  - iv. Names of physicians
  - v. Medical history, highlighting major events
  - vi. Copies of recent tests and/or procedures
  - vii. Advanced care directives

Find a binder template here: [Medical Binder Template](#)



- b. Personal Belongings
  - i. Clothing that is comfortable and loose fitting (not a tripping hazard)
  - ii. Eyeglasses, dentures, their cases and their cleaning solutions
  - iii. Toiletries (soap, shampoo, toothbrush, toothpaste, deodorant, hairbrush, shaver)
  - iv. Cell phone charger
  - v. Items that may provide comfort
  - vi. Blanket (it can be quite cold in the ER)
  - vii. Comfortable footwear (easy to remove and re-wear, along with preventing slipping)
  - viii. Medications

c. Sustenance

- i. Water and non-perishable snacks (granola bars, dry fruit, cereal, chips, crackers, etc.)

*\*\*Depending on the emergency, these may be shared with the patient. However in some cases it is crucial to remain fasted. Please confer with EMS, an emergency room nurse or doctor for further instructions\*\**

6. Speak with the local first responders

- a. Inform your local fire department and paramedics about your rare condition (e.g., risk of aortic dissection), so they are better prepared if they need to respond. Explain your living situation (e.g., living alone or with family) to help them know how to assist. Ask where they will look for medical information, such as a binder with medical records, when they enter your home to ensure you have your information in the right place.
- b. Contact your local hospital to ensure they are aware of your medical condition and ask that they flag it in their system, particularly the risk of aortic dissection. Refer to [Patient Flagging Systems](#) for more information.

# Emergency Preparedness for Parents of School-Aged Children

As children grow and start school, or join extracurricular activities, it's important to ensure that school staff are aware of how to care for a child with Loeys-Dietz syndrome (LDS). To support this, the Loeys-Dietz Syndrome Foundation Canada provides a detailed [School Packet](#) which includes:

1. Sample Letter to School Administration and Staff. Use this letter to explain LDS and how it affects your child, outlining any accommodations or emergency plans needed.
2. Medical Information and Emergency Contacts. Ensure the school has up-to-date medical information and contact numbers for emergencies.
3. General Facts on LDS. Share basic information about the syndrome so that teachers and staff can understand the unique challenges your child faces.
4. Signs and Symptoms Requiring Immediate Emergency Care. Provide a list of symptoms to watch for that may indicate a medical emergency.
5. School Accommodations. List accommodations your child may need to succeed at school, such as physical activity limitations or classroom adjustments.

Although the [School Packet](#) provides a solid foundation, it's important to customize it to fit your child's individual needs. Consider setting up a meeting with key staff members, such as the homeroom teacher, school nurse, and administration, to go over your child's health concerns and emergency plans.



**During the meeting:**

Educate staff about LDS and provide an opportunity for them to ask questions.

Walk them through the emergency response plan, similar to the approach described in the [Emergency Preparedness](#) section.

This proactive communication helps ensure the school team is equipped and confident in handling any emergencies your child may face, allowing them to thrive in a safe and supportive environment.



## Emergency Preparedness for Parents of Adult Children Attending Post- Secondary Education

When your child transitions to post-secondary education, it can be both exciting and challenging, especially if they have Loeys-Dietz syndrome. To ensure their safety and health are prioritized, here are some tips to help you feel more secure about their well-being while they're away from home.

### 1. Speak with Your Child's Residence Advisor (RA)

- RA's Role: Residence Advisors (RAs) are responsible for the welfare of students living in dorms and can be key partners in managing crises.
- What to Discuss: During your conversation with the RA, explain your child's specific medical needs, such as how to identify signs of emergencies (e.g., aortic dissection symptoms or respiratory distress) and the importance of immediate response. Share any essential medications your child may need.

- **Emergency Contact:** Ask how you can reach the RA if you're unable to contact your child during a potential emergency.

## 2. Campus Health Center

- **Services Offered:** Visit the campus health center to familiarize yourself with the medical services offered, including wellness and mental health support, or obtaining a new primary physician.
- **Local Physician:** If your child is studying far from home, finding a local physician familiar with LDS can help expedite medical care when needed. There may be a physician in the Campus Health Center who can attend to your child's care. You may also consider contacting a specialist with experience treating LDS.
- **Insurance Policies:** Investigate any student insurance plans that may provide coverage for medical and alternative healthcare needs, offering financial support in case of emergencies or ongoing treatments.

## 3. Download the Campus Security App

- **Security App:** Many universities have apps that allow students to quickly contact campus security or emergency services. Make sure your child has the app installed, and understands how to use it in case of emergencies.
- **Proactive Monitoring:** Speak with security personnel about your child's condition, and explore if it's possible for them to monitor emergency calls from your child to ensure a swift response.

## 4. Visit Local Hospitals and First Responders

- **Local Hospital Visit:** Visit the nearest hospital to your child's school and speak with staff about their capabilities in handling LDS-related emergencies like aortic dissection. Ask if they have the necessary equipment and expertise to treat your child's specific condition.
- **Establishing Health Records:** Try to set up a referral through your child's general practitioner for a specialist at the local hospital, ensuring your child's medical records are established before an emergency occurs.

- First Responders: Reach out to the local paramedics and firefighters to alert them about your child's presence in the area and their health risks. This preparation can help ensure faster and more informed responses in a crisis.
- Health Flags: Refer to the [Patient Flagging Systems](#) section for information on how to set up specific health protocols within the hospital's records, ensuring the best possible care during an emergency.

By taking these steps, you can help ensure your adult child has the right resources and support system while attending post-secondary education, giving you peace of mind that they'll receive proper care even when away from home.

## Wearable Medical Alert Systems

Wearing a medical alert system can be lifesaving for people with Loeys-Dietz syndrome, especially during emergencies when medical personnel need critical health information quickly. These devices provide vital information such as your name, medical conditions, allergies, and instructions for specific treatments. Medical alert systems are often quite versatile and can be placed on bracelets, watchbands, and various other accessories that will help them fit in with your lifestyle.

## INFORMATION TO INCLUDE ON YOUR MEDICAL ID

When customizing your wearable medical ID, it's essential to consider which information will help first responders or medical staff understand and act on your health needs quickly. Here are some suggestions:

1. Condition: Loeys- Dietz syndrome
2. Risk of dissection must be ruled out with a CT scan
3. Cervical spine instability
4. Allergies to medications etc.

Customize further based on any other significant health conditions or risks. By wearing an ID with this information, you ensure that medical responders have immediate access to critical data, improving your safety in an emergency situation.

## UNIVERSAL MEDICAL ID

<https://canada.universalmedicalid.com/>

Types of IDs: This site offers various options for medical ID jewelry like necklaces, bracelets, and even tags that can be attached to phone cases. All of these can be engraved with essential medical details.

Interactive Health Record (IHR): For an additional fee, you can opt for an interactive health record. This feature includes a medical card with a QR code that links to your detailed health records, making it easily accessible for medical professionals.

## MEDICAL ID QR CODES

If you would like to provide a more detailed medical profile for Emergency First Responders, you can consider created a shareable, digital document that can be accessed with a QR code. One way to achieve this would be to:

1. Create a Google Doc with your medical information
2. Provide access to anyone who has the link
3. Use the link to create a QR code
4. Print and post your QR code on your fridge, in your wallet, and in your medicine cabinet.



[MyID](#) is a company who offers a solution for anyone who does not feel comfortable creating their own profile and QR code. You can visit their site to find products, including wristband sleeves and QR code stickers, here: <https://shop.getmyid.com>

Picture Source: <https://shop.getmyid.com/collections/myid-products/products/myid-sleeve-medical-id>

## ELECTRONIC MEDICAL ALERT DEVICES

Electronic medical alert devices are ideal for connecting individuals with trained professionals who can quickly assess emergencies and notify both emergency services and personal contacts. Below are some options to consider:

1. [Life Assure](#)
2. [LivingWell Companion](#)
3. [Caretrak](#)

Another option would be devices like the iPhone, Apple Watch, and newer Google devices, in which you can have your medical information added to an emergency section for first responders and health crews to look at. Some of these devices even include fall detection and auto calling to emergency services in the event that you cannot respond after a fall.

# Advanced Directives

Planning for the future, especially regarding healthcare decisions, can be challenging, but advanced care directives ensure that your wishes are respected even if you're unable to communicate them. These directives are vital for preserving your autonomy and ensuring your end-of-life care reflects your values, beliefs, and preferences.

Advanced directives can cover:

- **Living Will:** A document outlining specific medical treatments you do or do not want if you become incapacitated.
- **Healthcare Proxy:** A person you appoint to make medical decisions on your behalf.
- **Do Not Resuscitate (DNR) Order:** Instructions not to perform CPR if your heart stops.
- **Power of Attorney:** A legal document granting someone the authority to make decisions about your healthcare and finances.

For more information on advanced directive options, legal guidelines, and resources specific to your province or territory, visit the [End of Life Law and Policy in Canada](#). This resource provides a comprehensive guide to help you navigate your choices in Canada.

# Raising Awareness

## ZEBRA PRINT LANYARD OR RIBBON

*“When you hear hoofbeats, think horses, not zebras”*

The zebra print lanyard is a symbol for rare diseases and is used to remind medical professionals to think beyond common diagnoses when they encounter patients with common symptoms. The saying, “When you hear hoofbeats, think horses, not zebras,” emphasizes looking for the usual cause first, but the zebra print reminds healthcare providers that sometimes it's a rare condition, like an aortic dissection, that's causing the issue.

Wearing this lanyard is a simple way to raise awareness about rare diseases. Though it may not assist directly during emergencies, it helps to make rare conditions, such as Loeys-Dietz Syndrome (LDS), more visible and understood. It's a reminder that individuals with rare conditions can live fulfilling, dignified lives beyond their diagnosis.

## IDENTIFICATION CARD

The Loeys-Dietz Syndrome Foundation Canada has developed a [medical emergency card](#) tailored for individuals with Loeys-Dietz syndrome. It includes quick information on the [4 most severe emergencies](#) as listed in the first section of this booklet. It also includes an area to indicate current medications, surgeries and other important medical information. This card is a quick reference tool that can be invaluable during emergencies, offering first responders a clear snapshot of the patient's medical history and key concerns.

<p><b>EMERGENCY ALERT CARD</b> <b>LOEYS-DIETZ SYNDROME</b> <b>(Heritable Connective Tissue Disorder)</b></p> <p>This person is <b>AT RISK</b> for:</p> <ul style="list-style-type: none"> <li><b>AORTIC &amp; ARTERIAL DISSECTION (head to pelvis)</b></li> <li><b>PNEUMOTHORAX</b></li> <li><b>HOLLOW ORGAN RUPTURE</b></li> <li><b>RETINAL DETACHMENT</b></li> </ul> <p><b>UNFOLD FOR MORE INFORMATION</b></p>		<p><b>MORE MEDICAL INFORMATION</b></p> <ul style="list-style-type: none"> <li>This person may have <b>FRAGILE TISSUES</b>: extreme care should be taken in transport to and at the hospital. However, tissues are not too fragile for surgery.</li> <li><b>OTHER EMERGENCY complications</b>: cardiac tamponade, hemorrhagic shock, ectopia lentis, hemoptysis, and during pregnancy: high risk of vascular events, uterine rupture.</li> <li>Information for <b>MEDICAL PROFESSIONALS</b> on Loeys-Dietz syndrome:</li> </ul> 																																				
<p><b>CONTACT US</b> 1-888-LDS-FCAN www.loeysdietzcanada.org</p> 																																						
<p><b>CURRENT MEDICATIONS</b></p> <table border="1"> <thead> <tr> <th></th> <th>DOSE</th> <th>FREQUENCY</th> </tr> </thead> <tbody> <tr><td> </td><td> </td><td> </td></tr> </tbody> </table>			DOSE	FREQUENCY																									<p><b>PATIENT INFORMATION</b></p> <table border="0"> <tr> <td><input type="checkbox"/> MECHANICAL VALVE</td> <td><input type="checkbox"/> DIFFICULT AIRWAY</td> </tr> <tr> <td><input type="checkbox"/> PACEMAKER</td> <td><input type="checkbox"/> PRIOR AORTIC REPAIR</td> </tr> <tr> <td><input type="checkbox"/> CONTACT LENSES</td> <td><input type="checkbox"/> CERVICAL SPINE INSTABILITY</td> </tr> <tr> <td><input type="checkbox"/> BLOOD THINNER(S)</td> <td>If C-spine is unstable, use caution during intubation to avoid dislocation.</td> </tr> </table> <p><b>CURRENT ANEURYSMS:</b></p> <p>EMERGENCY CONTACT NAME (RELATIONSHIP):</p> <p>PHONE NUMBER 1:                      PHONE NUMBER 2:</p>		<input type="checkbox"/> MECHANICAL VALVE	<input type="checkbox"/> DIFFICULT AIRWAY	<input type="checkbox"/> PACEMAKER	<input type="checkbox"/> PRIOR AORTIC REPAIR	<input type="checkbox"/> CONTACT LENSES	<input type="checkbox"/> CERVICAL SPINE INSTABILITY	<input type="checkbox"/> BLOOD THINNER(S)	If C-spine is unstable, use caution during intubation to avoid dislocation.
	DOSE	FREQUENCY																																				
<input type="checkbox"/> MECHANICAL VALVE	<input type="checkbox"/> DIFFICULT AIRWAY																																					
<input type="checkbox"/> PACEMAKER	<input type="checkbox"/> PRIOR AORTIC REPAIR																																					
<input type="checkbox"/> CONTACT LENSES	<input type="checkbox"/> CERVICAL SPINE INSTABILITY																																					
<input type="checkbox"/> BLOOD THINNER(S)	If C-spine is unstable, use caution during intubation to avoid dislocation.																																					
<p><b>SURGERIES: (PREVIOUS &amp; UPCOMING)</b></p> <table border="1"> <thead> <tr> <th>NAME OF SURGERY</th> <th>DATE</th> </tr> </thead> <tbody> <tr><td> </td><td> </td></tr> </tbody> </table>		NAME OF SURGERY	DATE																	<p>FULL NAME:</p> <p>DATE OF BIRTH:                      BLOOD TYPE:</p> <p>HEALTH CARD NUMBER:</p> <p>ALLERGIES (REACTIONS):</p> <p>To treat allergic reactions: Use antihistamine first. Because of its effect on the circulatory system, only use epinephrine injector in life-threatening cases.</p> <p><b>UNFOLD FOR MORE INFORMATION</b></p>																		
NAME OF SURGERY	DATE																																					

# Tips from Doctors, Nurses, Patients and First Responders

## 1. Know where your **phone** is always

- a. If you live alone or are alone during a crisis this is your main method of alerting emergency services

## 2. Wear your **medical alert system**

- a. Make it a routine to wear your medical alert system, as it is a simple method of mobilizing first responders during a crises

## 3. Visit your **local first responders** (fire station, emergency department)

- a. Establish a relationship with your local first responders. Introduce yourself, inform them about your condition, and share key medical concerns. This helps them become familiar with your needs, so in a crisis, they can act swiftly and appropriately. If hospital staff are unfamiliar with your condition, this rapport allows time for preparation.

## 4. Use a **Door Sign** to Indicate an LDS patient

- a. A sign on your door signaling that you have Loeys-Dietz syndrome can be vital, especially if you're unconscious.
- b. Near the door, hang a sign that shows where your [emergency binder](#) is located. The binder should provide an overview of your medical history, including allergies, past dissections, and any current medications. This is especially helpful for first responders or even a friend unfamiliar with your condition.
- c. Consider including any quick info like allergies, previous dissections, and medication on the signage to avoid first responders searching for pertinent medical information

## 5. **Medical Binder** and **Medications**

- a. Place a [medical emergency binder](#) on the top of the fridge as medical professionals are trained to search this common area first for medical items
- b. Keep medications together, ideally in the medicine cabinet, in the event they need to be administered, they are readily available.

These proactive steps can help ensure that in the event of an emergency, you and those around you can act quickly and effectively.

# Resources

Family Planning with LDS: [https://loeysdietzcanada.org/wp-content/uploads/2023/12/FINAL-EN-COPY-LDS-Family-Planning\\_12-2023.pdf](https://loeysdietzcanada.org/wp-content/uploads/2023/12/FINAL-EN-COPY-LDS-Family-Planning_12-2023.pdf)

Ritter Rules: <https://johnritterfoundation.org/your-aortic-health/ritter-rules/>

FYI Patient Flag: [https://hit.healthsystem.virginia.edu/index.cfm/\\_api/render/file?fileID=E2F57A31-17A4-77A0-3EC55AE780DAB3CC&fileEXT=.pdf](https://hit.healthsystem.virginia.edu/index.cfm/_api/render/file?fileID=E2F57A31-17A4-77A0-3EC55AE780DAB3CC&fileEXT=.pdf)

My Health Care Journey Binder: <https://ontariocaregiver.ca/my-health-care-journey-binder/>

LDSFC School Packet: [https://loeysdietzcanada.org/wp-content/uploads/2022/10/LDSFC\\_Loeys-Dietz-Syndrome-School-Packet.pdf](https://loeysdietzcanada.org/wp-content/uploads/2022/10/LDSFC_Loeys-Dietz-Syndrome-School-Packet.pdf)

Universal Medical ID: <https://canada.universalmedicalid.com/>

MyID QR Code Products: <https://shop.getmyid.com>

Life Assure: <https://www.lifeassure.com/>

LivingWell Companion: [https://www.telus.com/en/personal-health/livingwell-companion/livingwelideal?  
&cmp=KNC\\_HS\\_Google\\_&SEM\\_CID=11908595029&SEM\\_AG=112379211541&SEM\\_KW=telus%20medical%20alert&SEM\\_MT=b&gclid=Cj0KCQiAg\\_KbBhDLARIsANx7wAy7TRMIJlg\\_DBaJISBfV\\_\\_QJhtPQ7hjr16pJ00ulGUu\\_JM1\\_PZdBFAaAttgEALw\\_wcB&gclsrc=a](https://www.telus.com/en/personal-health/livingwell-companion/livingwelideal?&cmp=KNC_HS_Google_&SEM_CID=11908595029&SEM_AG=112379211541&SEM_KW=telus%20medical%20alert&SEM_MT=b&gclid=Cj0KCQiAg_KbBhDLARIsANx7wAy7TRMIJlg_DBaJISBfV__QJhtPQ7hjr16pJ00ulGUu_JM1_PZdBFAaAttgEALw_wcB&gclsrc=a)  
w.ds

Caretrak: [https://www.caretrak.ca/?  
gclid=Cj0KCQiAg\\_KbBhDLARIsANx7wAxQvXE7NOwJAKMOoFhS\\_F0ACTmpcGOYGnn84wssykpD8A8KA2MaLUaAvtVEALw\\_wcB](https://www.caretrak.ca/?gclid=Cj0KCQiAg_KbBhDLARIsANx7wAxQvXE7NOwJAKMOoFhS_F0ACTmpcGOYGnn84wssykpD8A8KA2MaLUaAvtVEALw_wcB)

End of Life Law and Policy in Canada: [http://eol.law.dal.ca/?page\\_id=231](http://eol.law.dal.ca/?page_id=231)

Emergency Alert Card: [https://loeysdietzcanada.org/wp-content/uploads/2024/09/FINAL\\_Fillable\\_LDSFC-Emergency-Alert-Card\\_May-16-2023.pdf](https://loeysdietzcanada.org/wp-content/uploads/2024/09/FINAL_Fillable_LDSFC-Emergency-Alert-Card_May-16-2023.pdf)

# References

Bevans, M., & Sternberg, E. M. (2012). Caregiving Burden, Stress, and Health Effects Among Family Caregivers of Adult Cancer Patients. *JAMA*, 307(4), 398–403. <https://doi.org/10.1001/JAMA.2012.29>

Bhattacharyya, R., Swergold, N., & Bethel, C. (2018). Congenital pericardioperitoneal hernia in a patient with Loeys-Dietz syndrome. *Journal of Pediatric Surgery Case Reports*, 32, 41–42. <https://doi.org/10.1016/J.EPSC.2018.01.013>

Boone, P. M., Scott, R. M., Marciniak, S. J., Henske, E. P., & Raby, B. A. (2019). The genetics of pneumothorax. *American Journal of Respiratory and Critical Care Medicine*, 199(11), 1344–1357. [https://doi.org/10.1164/RCCM.201807-1212CI/SUPPL\\_FILE/DISCLOSURES.PDF](https://doi.org/10.1164/RCCM.201807-1212CI/SUPPL_FILE/DISCLOSURES.PDF)

Borke, J. (2021, January 12). Collapsed Lung (Pneumothorax) . PennMedicine. <https://www.pennmedicine.org/for-patients-and-visitors/patient-information/conditions-treated-a-to-z/collapsed-lung-pneumothorax>

Caza, T. N., Mercedes, A., Stoppacher, R., & Catanese, C. A. (2016). A Complex Case of Loeys-Dietz Syndrome: A Case Report and Review of the Literature. *Academic Forensic Pathology*, 6(4), 731. <https://doi.org/10.23907/2016.069>

Christian, H. (2019). Stress Fractures in Loeys-Dietz Syndrome. *International Archives of Orthopaedic Surgery*, 2(1). <https://doi.org/10.23937/2643-4016/1710008>

Detached retina (retinal detachment) - NHS. (2020, December 16). National Health System. <https://www.nhs.uk/conditions/detached-retina-retinal-detachment/>

Erlinger, C. (2017, March 29). Heart Attack: Men vs. Women. The Heart Foundation. <https://theheartfoundation.org/2017/03/29/heart-attack-men-vs-women/>

Fuhrhop, S. K., McElroy, M. J., Dietz, H. C., MacCarrick, G. L., & Sponseller, P. D. (2015). High Prevalence of Cervical Deformity and Instability Requires Surveillance in Loeys-Dietz Syndrome. *The Journal of Bone and Joint Surgery. American Volume*, 97(5), 411. <https://doi.org/10.2106/JBJS.N.00680>

# References

- Gastrointestinal Perforation. (2022, November 7). Cleveland Clinic.  
<https://my.clevelandclinic.org/health/diseases/23478-gastrointestinal-perforation>
- Guerrero, A. L., Mateja, A., Rasooly, M., Levin, S., Magnani, A., Dempsey, C., MacCarrick, G., Dietz, H. C., Brittain, E., Boyce, A. M., & Frischmeyer-Guerrero, P. A. (2022). Predictors of low bone density and fracture risk in Loeys–Dietz syndrome. *Genetics in Medicine*, 24(2), 419–429. <https://doi.org/10.1016/j.gim.2021.10.002>
- Heart Attack Symptoms in Women. (2015, July 31). American Heart Association.  
<https://www.heart.org/en/health-topics/heart-attack/warning-signs-of-a-heart-attack/heart-attack-symptoms-in-women>
- How to prepare for an emergency room visit. (2022, May 17). HCA Florida Healthcare.  
<https://www.hcafloridahealthcare.com/healthy-living/blog/how-to-prepare-for-an-emergency-room-visit>
- How to use FYI flags. (n.d.). Retrieved 18 December 2023, from  
[https://hit.healthsystem.virginia.edu/index.cfm/\\_api/render/file/?fileID=E2F57A31-17A4-77A0-3EC55AE780DAB3CC&fileEXT=.pdf](https://hit.healthsystem.virginia.edu/index.cfm/_api/render/file/?fileID=E2F57A31-17A4-77A0-3EC55AE780DAB3CC&fileEXT=.pdf)
- Isselbacher, E. M., Bonaca, M. P., Di Eusanio, M., Froehlich, J., Bassone, E., Sechtem, U., Pyeritz, R., Patel, H., Khojenezhad, A., Eckstein, H. H., Jondeau, G., Ramponi, F., Abbasi, M., Montgomery, D., Nienaber, C. A., Eagle, K., & Lindsay, M. E. (2016). Recurrent Aortic Dissection: Observations from the International Registry of Aortic Dissection. *Circulation*, 134(14), 1013–1024. <https://doi.org/10.1161/CIRCULATIONAHA.115.019359>
- Levy, D., Goyal, A., Grigorova, Y., Farci, F., & Le, J. K. (2022). Aortic Dissection. *StatPearls*.  
<https://www.ncbi.nlm.nih.gov/books/NBK441963/>
- Lobaton, G. O., Chen, Y. J., Jelin, E., & Garcia, A. V. (2021). Unusual case of delayed congenital diaphragmatic hernia in Loeys-Dietz syndrome: a case report. *Journal of Surgical Case Reports*, 2021(1), 1–3. <https://doi.org/10.1093/JSCR/RJAA604>
- Loeys, B. L., & Dietz, H. C. (2018). Loeys-Dietz Syndrome. *GeneReviews*®.  
<https://www.ncbi.nlm.nih.gov/books/NBK1133/>

# References

Loeys-Dietz Syndrome - Symptoms and Causes | Penn Medicine. (n.d.). PennMedicine. Retrieved 26 October 2022, from <https://www.pennmedicine.org/for-patients-and-visitors/patient-information/conditions-treated-a-to-z/loeys-dietz-syndrome>

Loeys-Dietz Syndrome Foundation Canada School Packet. (2022, October). Retrieved 31 October 2023, from [https://loeysdietzcanada.org/wp-content/uploads/2022/10/LDSFC\\_Loeys-Dietz-Syndrome-School-Packet.pdf](https://loeysdietzcanada.org/wp-content/uploads/2022/10/LDSFC_Loeys-Dietz-Syndrome-School-Packet.pdf)

Loeys-Dietz Syndrome Foundation Canada Family Planning with LDS. (2023, December). Retrieved 30 December 2024, from [https://loeysdietzcanada.org/wp-content/uploads/2023/12/FINAL-EN-COPY-LDS-Family-Planning\\_12-2023.pdf](https://loeysdietzcanada.org/wp-content/uploads/2023/12/FINAL-EN-COPY-LDS-Family-Planning_12-2023.pdf)

Lu, L., Liu, M., Sun, R. R., Zheng, Y., & Zhang, P. (2015). Myocardial Infarction: Symptoms and Treatments. *Cell Biochemistry and Biophysics* 2015 72:3, 72(3), 865–867. <https://doi.org/10.1007/S12013-015-0553-4>

Mayo Foundation for Medical Education and Research. (2021, August 6). Aortic dissection. Mayo Clinic. <https://www.mayoclinic.org/diseases-conditions/aortic-dissection/symptoms-causes/syc-20369496>

MacCarrick, G., Black, J. H., Bowdin, S., El-Hamamsy, I., Frischmeyer-Guerrerio, P. A., Guerrerio, A. L., Sponseller, P. D., Loeys, B., & Dietz, H. C. (2014). Loeys–Dietz syndrome: a primer for diagnosis and management. *Genetics in Medicine*, 16(8), 576. <https://doi.org/10.1038/GIM.2014.11>

Nienaber, C. A., Fattori, R., Mehta, R. H., Richartz, B. M., Evangelista, A., Petzsch, M., Cooper, J. V., Januzzi, J. L., Ince, H., Sechtem, U., Bossone, E., Fang, J., Smith, D. E., Isselbacher, E. M., Pape, L. A., & Eagle, K. A. (2004). Gender-Related Differences in Acute Aortic Dissection. *Circulation*, 109(24), 3014–3021. <https://doi.org/10.1161/01.CIR.0000130644.78677.2C>

Peicius, E., Blazeviciene, A., & Kaminskas, R. (2017). Are advance directives helpful for good end of life decision making: a cross sectional survey of health professionals. *BMC Medical Ethics*, 18(1), 1–7. <https://doi.org/10.1186/S12910-017-0197-6/TABLES/4>

# References

Preparing for an Emergency Room Visit. (n.d.). Memorial Care. Retrieved 2 November 2022, from <https://www.memorialcare.org/services/emergency-care/preparing-emergency-room-visit>

Research, C. for D. E. and. (2019). FDA warns about the increased risk of ruptures or tears in the aorta blood vessel with fluoroquinolone antibiotics in certain patients. FDA. <https://www.fda.gov/drugs/drug-safety-and-availability/fda-warns-about-increased-risk-ruptures-or-tears-aorta-blood-vessel-fluoroquinolone-antibiotics>

Ritter Rules. (n.d.). The John Ritter Foundtion for Aortic Health. Retrieved 22 November 2022, from <https://johnritterfoundation.org/your-aortic-health/ritter-rules/>

Ruptured spleen. (n.d.). Mayo Clinic. Retrieved 23 November 2022, from <https://www.mayoclinic.org/diseases-conditions/ruptured-spleen/symptoms-causes/syc-20352317>

Zebra Stripes Awareness Ribbon Meaning for Rare Disease and Gifts. (2017, October 4). Awareness Gallery Art. <https://awarenessgallery.com/blog/2017/10/04/zebra-stripes-awareness-ribbon-meaning-for-rare-disease-and-gifts/>