

# THE EDS MOVEMENT

*Charging forward. Saving lives.*

## EMERGENCY PREPAREDNESS KIT

**Patient Name:**

This kit is your tool to help you be prepared in the event of an emergency. We suggest putting the completed packet in a brightly colored envelope or folder so it is easy to find. At home, keep it near the door so it is handy for Emergency Medical Services (EMS) and perhaps tack it up on the wall at work. Also provide a copy of this packet to your Power of Attorney and Healthcare Proxy. Portable USB drives can hold all this information and can be carried on a key chain. Some medical alert services have these drives available with their logo or you can purchase them in any office supply store.

Many people put emergency contact information in their cell phone filed under ICE (In Case of Emergency). Use ICE1, ICE2 and so on. EMS personnel are trained to look for this on your cell phone.

If you need emergency care, it may be helpful for you to call your primary care provider to call ahead on your behalf and notify the doctor in charge of the ER of a possible arterial dissection, rupture, or organ rupture before you arrive. You or someone close to you may also want to call the ER before you arrive.

We recommend that you complete these documents so they are available in case of an emergency. Some of these documents may not apply to you (for instance, not everyone will want a Do Not Resuscitate Order). Remember to update your information regularly.

*Last Updated*

- Personal Information Form ..... \_\_\_\_\_
- Medical History ..... \_\_\_\_\_
- Genetic Testing Results ..... \_\_\_\_\_
- Medical note/summary from your last appointment with your doctor ..... \_\_\_\_\_
- Doctor(s) Information ..... \_\_\_\_\_
- Insurance Information ..... \_\_\_\_\_
- Family Medical History..... \_\_\_\_\_
- Legal Information..... \_\_\_\_\_
- Power of Attorney (sample included under Legal Information, below)..... \_\_\_\_\_
- Healthcare Proxy (sample included under Legal Information)..... \_\_\_\_\_
- Living Will (sample included under Legal Information) ..... \_\_\_\_\_
- Do Not Resuscitate Order (sample included under Legal Information) ..... \_\_\_\_\_

**Include a copy of the most recent version of each of the following from your doctor**

- Ultrasound/CD with Written Report ..... \_\_\_\_\_
- MRI/MRA Films with Written Report..... \_\_\_\_\_
- CT/CTA Films with Written Report..... \_\_\_\_\_
- Blood Work Results ..... \_\_\_\_\_

**Other resources**

- VEDS: Basic Facts..... \_\_\_\_\_
- Fact Sheet for Paramedics and Emergency Medical Technicians..... \_\_\_\_\_
- Letter to the Emergency Physician (Template) ..... \_\_\_\_\_
- VEDS Information Line for Emergency Physicians..... \_\_\_\_\_
- Medical Alert Bracelet Information..... \_\_\_\_\_
- Emergency Alert Card ..... \_\_\_\_\_

# PERSONAL INFORMATION

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First Name: \_\_\_\_\_ Last Name: \_\_\_\_\_  
Date of Birth: \_\_\_\_\_ Social Security Number: \_\_\_\_\_ - \_\_\_\_\_ - \_\_\_\_\_  
Gender: Male Female Marital Status: \_\_\_\_\_

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## Contact Information

Home Address: \_\_\_\_\_  
City: \_\_\_\_\_ State: \_\_\_\_\_ Zip: \_\_\_\_\_  
Home Phone: \_\_\_\_\_ Work Phone: \_\_\_\_\_  
Cell Phone: \_\_\_\_\_ Email: \_\_\_\_\_  
Occupation: \_\_\_\_\_ Employer: \_\_\_\_\_  
Work Address: \_\_\_\_\_  
City: \_\_\_\_\_ State: \_\_\_\_\_ Zip: \_\_\_\_\_

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## Health-related Information

Height: \_\_\_\_\_ Weight: \_\_\_\_\_ Blood Type: \_\_\_\_\_  
Normal Blood Pressure: \_\_\_\_\_ Resting Heart Rate: \_\_\_\_\_  
Alcohol Consumption (number of drinks consumed): \_\_\_\_\_ per day per week  
Smoking: Non Smoker 1 pack or less/week 2-3 packs/week 1 pack/day More than 1 pack/day  
Former Smoker / Date Quit: \_\_\_\_\_

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## Language Information

Language spoken at home: \_\_\_\_\_ Do you need an interpreter? Yes No  
*If you need an interpreter and the hospital is temporarily unable to provide one, who can they contact to provide assistance?*  
First Name: \_\_\_\_\_ Last Name: \_\_\_\_\_  
Home Phone: \_\_\_\_\_ Work Phone: \_\_\_\_\_ Cell Phone: \_\_\_\_\_

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## Emergency Contacts

**Contact 1** First Name: \_\_\_\_\_ Last Name: \_\_\_\_\_  
Address: \_\_\_\_\_ Relationship: \_\_\_\_\_  
City: \_\_\_\_\_ State: \_\_\_\_\_ Zip: \_\_\_\_\_  
Home Phone: \_\_\_\_\_ Work Phone: \_\_\_\_\_ Cell Phone: \_\_\_\_\_  
**Contact 2** First Name: \_\_\_\_\_ Last Name: \_\_\_\_\_  
Address: \_\_\_\_\_ Relationship: \_\_\_\_\_  
City: \_\_\_\_\_ State: \_\_\_\_\_ Zip: \_\_\_\_\_  
Home Phone: \_\_\_\_\_ Work Phone: \_\_\_\_\_ Cell Phone: \_\_\_\_\_

First Name: \_\_\_\_\_ Last Name: \_\_\_\_\_

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### Diagnosis

VEDS (Vascular Ehlers-Danlos Syndrome) Age at diagnosis: \_\_\_\_\_

Notes/comments: \_\_\_\_\_

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### Current Medications (include vitamins/supplements)

1. **Name:** \_\_\_\_\_ **Dosage:** \_\_\_\_\_ **Schedule:** \_\_\_\_\_

*Reason:* \_\_\_\_\_

2. **Name:** \_\_\_\_\_ **Dosage:** \_\_\_\_\_ **Schedule:** \_\_\_\_\_

*Reason:* \_\_\_\_\_

3. **Name:** \_\_\_\_\_ **Dosage:** \_\_\_\_\_ **Schedule:** \_\_\_\_\_

*Reason:* \_\_\_\_\_

4. **Name:** \_\_\_\_\_ **Dosage:** \_\_\_\_\_ **Schedule:** \_\_\_\_\_

*Reason:* \_\_\_\_\_

5. **Name:** \_\_\_\_\_ **Dosage:** \_\_\_\_\_ **Schedule:** \_\_\_\_\_

*Reason:* \_\_\_\_\_

6. **Name:** \_\_\_\_\_ **Dosage:** \_\_\_\_\_ **Schedule:** \_\_\_\_\_

*Reason:* \_\_\_\_\_

If you have additional medications, please list them on page 7 of this form.

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### Allergies

1. \_\_\_\_\_ 4. \_\_\_\_\_

2. \_\_\_\_\_ 5. \_\_\_\_\_

3. \_\_\_\_\_ 6. \_\_\_\_\_

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### Vascular History (Arteries, Veins, Aorta)

What is your vascular history of aneurysms, ruptures, dissections?

*Please attach most recent imaging studies.*

## **Bowel History (Intestines, Bladder, Stomach, etc.)**

What bowel issues do you have or have you previously had? (e.g., intestinal rupture, gastroparesis, bladder rupture, etc.)

*Please attach most recent imaging studies.*

## **Uterine History**

What uterine issues do you have or have you previously had? (e.g. uterine rupture, endometriosis, etc.)

*Please attach most recent imaging studies.*

## **Cardiac History (Heart)**

What cardiac issues do you have or have you previously had? (e.g., mitral valve prolapse, bicuspid aortic valve or aortic aneurysm/dissection, etc.)

*Please attach most recent imaging studies.*

## **Ocular History (Eyes)**

What ocular issues do you have or have you previously had? (e.g., carotid cavernous sinus fistula, retinal detachment, strabismus, cataracts, etc.)

*Please attach most recent imaging studies.*

## **Orthopedic History (Bones & Joints)**

What skeleton and joint issues do you have or have you previously had? (e.g., scoliosis, dislocations, etc.)

*Please attach most recent imaging studies.*

## **Pulmonary History (Lungs)**

What lung issues do you have or have you previously had? (e.g. asthma, pneumothorax, pulmonary blebs, hemothorax, etc.)

*Please attach most recent imaging studies.*

First Name: \_\_\_\_\_ Last Name: \_\_\_\_\_

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## Recent Surgeries /Procedures

What surgeries/procedures have you had (e.g., aortic repair, arterial repair, intestinal surgery, etc)?

**1. Surgery/Procedure:** \_\_\_\_\_

Location: \_\_\_\_\_ Date: \_\_\_\_\_

Doctor who performed surgery: \_\_\_\_\_ Doctor's phone: \_\_\_\_\_

**2. Surgery/Procedure:** \_\_\_\_\_

Location: \_\_\_\_\_ Date: \_\_\_\_\_

Doctor who performed surgery: \_\_\_\_\_ Doctor's phone: \_\_\_\_\_

**3. Surgery/Procedure:** \_\_\_\_\_

Location: \_\_\_\_\_ Date: \_\_\_\_\_

Doctor who performed surgery: \_\_\_\_\_ Doctor's phone: \_\_\_\_\_

**4. Surgery/Procedure:** \_\_\_\_\_

Location: \_\_\_\_\_ Date: \_\_\_\_\_

Doctor who performed surgery: \_\_\_\_\_ Doctor's phone: \_\_\_\_\_

**5. Surgery/Procedure:** \_\_\_\_\_

Location: \_\_\_\_\_ Date: \_\_\_\_\_

Doctor who performed surgery: \_\_\_\_\_ Doctor's phone: \_\_\_\_\_

**6. Surgery/Procedure:** \_\_\_\_\_

Location: \_\_\_\_\_ Date: \_\_\_\_\_

Doctor who performed surgery: \_\_\_\_\_ Doctor's phone: \_\_\_\_\_

**7. Surgery/Procedure:** \_\_\_\_\_

Location: \_\_\_\_\_ Date: \_\_\_\_\_

Doctor who performed surgery: \_\_\_\_\_ Doctor's phone: \_\_\_\_\_

**8. Surgery/Procedure:** \_\_\_\_\_

Location: \_\_\_\_\_ Date: \_\_\_\_\_

Doctor who performed surgery: \_\_\_\_\_ Doctor's phone: \_\_\_\_\_

*If you have additional surgeries/procedures, please list them on page 8 of this form.*

First Name: \_\_\_\_\_ Last Name: \_\_\_\_\_

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### Additional Current Medications (include vitamins/supplements)

7. **Name:** \_\_\_\_\_ Dosage: \_\_\_\_\_ Schedule: \_\_\_\_\_

*Reason:* \_\_\_\_\_

8. **Name:** \_\_\_\_\_ Dosage: \_\_\_\_\_ Schedule: \_\_\_\_\_

*Reason:* \_\_\_\_\_

9. **Name:** \_\_\_\_\_ Dosage: \_\_\_\_\_ Schedule: \_\_\_\_\_

*Reason:* \_\_\_\_\_

10. **Name:** \_\_\_\_\_ Dosage: \_\_\_\_\_ Schedule: \_\_\_\_\_

*Reason:* \_\_\_\_\_

11. **Name:** \_\_\_\_\_ Dosage: \_\_\_\_\_ Schedule: \_\_\_\_\_

*Reason:* \_\_\_\_\_

12. **Name:** \_\_\_\_\_ Dosage: \_\_\_\_\_ Schedule: \_\_\_\_\_

*Reason:* \_\_\_\_\_

13. **Name:** \_\_\_\_\_ Dosage: \_\_\_\_\_ Schedule: \_\_\_\_\_

*Reason:* \_\_\_\_\_

14. **Name:** \_\_\_\_\_ Dosage: \_\_\_\_\_ Schedule: \_\_\_\_\_

*Reason:* \_\_\_\_\_

15. **Name:** \_\_\_\_\_ Dosage: \_\_\_\_\_ Schedule: \_\_\_\_\_

*Reason:* \_\_\_\_\_

16. **Name:** \_\_\_\_\_ Dosage: \_\_\_\_\_ Schedule: \_\_\_\_\_

*Reason:* \_\_\_\_\_

17. **Name:** \_\_\_\_\_ Dosage: \_\_\_\_\_ Schedule: \_\_\_\_\_

*Reason:* \_\_\_\_\_

18. **Name:** \_\_\_\_\_ Dosage: \_\_\_\_\_ Schedule: \_\_\_\_\_

*Reason:* \_\_\_\_\_

19. **Name:** \_\_\_\_\_ Dosage: \_\_\_\_\_ Schedule: \_\_\_\_\_

*Reason:* \_\_\_\_\_

First Name: \_\_\_\_\_ Last Name: \_\_\_\_\_

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### **Additional Recent Surgeries /Procedures**

What surgeries/procedures have you had (e.g., aortic repair, arterial repair, intestinal surgery, etc)?

**9. Surgery/Procedure:** \_\_\_\_\_

Location: \_\_\_\_\_ Date: \_\_\_\_\_

Doctor who performed surgery: \_\_\_\_\_ Doctor's phone: \_\_\_\_\_

**10. Surgery/Procedure:** \_\_\_\_\_

Location: \_\_\_\_\_ Date: \_\_\_\_\_

Doctor who performed surgery: \_\_\_\_\_ Doctor's phone: \_\_\_\_\_

**11. Surgery/Procedure:** \_\_\_\_\_

Location: \_\_\_\_\_ Date: \_\_\_\_\_

Doctor who performed surgery: \_\_\_\_\_ Doctor's phone: \_\_\_\_\_

**12. Surgery/Procedure:** \_\_\_\_\_

Location: \_\_\_\_\_ Date: \_\_\_\_\_

Doctor who performed surgery: \_\_\_\_\_ Doctor's phone: \_\_\_\_\_

**13. Surgery/Procedure:** \_\_\_\_\_

Location: \_\_\_\_\_ Date: \_\_\_\_\_

Doctor who performed surgery: \_\_\_\_\_ Doctor's phone: \_\_\_\_\_

**14. Surgery/Procedure:** \_\_\_\_\_

Location: \_\_\_\_\_ Date: \_\_\_\_\_

Doctor who performed surgery: \_\_\_\_\_ Doctor's phone: \_\_\_\_\_

**15. Surgery/Procedure:** \_\_\_\_\_

Location: \_\_\_\_\_ Date: \_\_\_\_\_

Doctor who performed surgery: \_\_\_\_\_ Doctor's phone: \_\_\_\_\_

**16. Surgery/Procedure:** \_\_\_\_\_

Location: \_\_\_\_\_ Date: \_\_\_\_\_

Doctor who performed surgery: \_\_\_\_\_ Doctor's phone: \_\_\_\_\_



# DOCTOR(S) INFORMATION

First Name: \_\_\_\_\_ Last Name: \_\_\_\_\_

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## Primary Care Physician

First Name: \_\_\_\_\_ Last Name: \_\_\_\_\_

Address: \_\_\_\_\_

City: \_\_\_\_\_ State: \_\_\_\_\_ Zip: \_\_\_\_\_

Cell Phone: \_\_\_\_\_ Pager: \_\_\_\_\_ Fax: \_\_\_\_\_

---

## Vascular Surgeon

First Name: \_\_\_\_\_ Last Name: \_\_\_\_\_

Address: \_\_\_\_\_

City: \_\_\_\_\_ State: \_\_\_\_\_ Zip: \_\_\_\_\_

Cell Phone: \_\_\_\_\_ Pager: \_\_\_\_\_ Fax: \_\_\_\_\_

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## Geneticist

First Name: \_\_\_\_\_ Last Name: \_\_\_\_\_

Address: \_\_\_\_\_

City: \_\_\_\_\_ State: \_\_\_\_\_ Zip: \_\_\_\_\_

Cell Phone: \_\_\_\_\_ Pager: \_\_\_\_\_ Fax: \_\_\_\_\_

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## General Surgeon

First Name: \_\_\_\_\_ Last Name: \_\_\_\_\_

Address: \_\_\_\_\_

City: \_\_\_\_\_ State: \_\_\_\_\_ Zip: \_\_\_\_\_

Cell Phone: \_\_\_\_\_ Pager: \_\_\_\_\_ Fax: \_\_\_\_\_

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## Ophthalmologist

First Name: \_\_\_\_\_ Last Name: \_\_\_\_\_

Address: \_\_\_\_\_

City: \_\_\_\_\_ State: \_\_\_\_\_ Zip: \_\_\_\_\_

Cell Phone: \_\_\_\_\_ Pager: \_\_\_\_\_ Fax: \_\_\_\_\_

# DOCTOR(S) INFORMATION *(continued)*

## **Orthopedist**

First Name: \_\_\_\_\_ Last Name: \_\_\_\_\_

Address: \_\_\_\_\_

City: \_\_\_\_\_ State: \_\_\_\_\_ Zip: \_\_\_\_\_

Cell Phone: \_\_\_\_\_ Pager: \_\_\_\_\_ Fax: \_\_\_\_\_

---

## **Cardiologist**

First Name: \_\_\_\_\_ Last Name: \_\_\_\_\_

Address: \_\_\_\_\_

City: \_\_\_\_\_ State: \_\_\_\_\_ Zip: \_\_\_\_\_

Cell Phone: \_\_\_\_\_ Pager: \_\_\_\_\_ Fax: \_\_\_\_\_

---

## **Other Specialist**

First Name: \_\_\_\_\_ Last Name: \_\_\_\_\_

Medical Specialty: \_\_\_\_\_

Address: \_\_\_\_\_

City: \_\_\_\_\_ State: \_\_\_\_\_ Zip: \_\_\_\_\_

Cell Phone: \_\_\_\_\_ Pager: \_\_\_\_\_ Fax: \_\_\_\_\_

*If you have additional doctors, please list them on page 11 of this form.*

# DOCTOR(S) INFORMATION *(continued)*

First Name: \_\_\_\_\_ Last Name: \_\_\_\_\_

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## **Other Specialist**

First Name: \_\_\_\_\_ Last Name: \_\_\_\_\_

Medical Specialty: \_\_\_\_\_

Address: \_\_\_\_\_

City: \_\_\_\_\_ State: \_\_\_\_\_ Zip: \_\_\_\_\_

Cell Phone: \_\_\_\_\_ Pager: \_\_\_\_\_ Fax: \_\_\_\_\_

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## **Other Specialist**

First Name: \_\_\_\_\_ Last Name: \_\_\_\_\_

Medical Specialty: \_\_\_\_\_

Address: \_\_\_\_\_

City: \_\_\_\_\_ State: \_\_\_\_\_ Zip: \_\_\_\_\_

Cell Phone: \_\_\_\_\_ Pager: \_\_\_\_\_ Fax: \_\_\_\_\_

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## **Other Specialist**

First Name: \_\_\_\_\_ Last Name: \_\_\_\_\_

Medical Specialty: \_\_\_\_\_

Address: \_\_\_\_\_

City: \_\_\_\_\_ State: \_\_\_\_\_ Zip: \_\_\_\_\_

Cell Phone: \_\_\_\_\_ Pager: \_\_\_\_\_ Fax: \_\_\_\_\_

---

## **Other Specialist**

First Name: \_\_\_\_\_ Last Name: \_\_\_\_\_

Medical Specialty: \_\_\_\_\_

Address: \_\_\_\_\_

City: \_\_\_\_\_ State: \_\_\_\_\_ Zip: \_\_\_\_\_

Cell Phone: \_\_\_\_\_ Pager: \_\_\_\_\_ Fax: \_\_\_\_\_

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## **Other Specialist**

First Name: \_\_\_\_\_ Last Name: \_\_\_\_\_

Medical Specialty: \_\_\_\_\_

Address: \_\_\_\_\_

City: \_\_\_\_\_ State: \_\_\_\_\_ Zip: \_\_\_\_\_

Cell Phone: \_\_\_\_\_ Pager: \_\_\_\_\_ Fax: \_\_\_\_\_

# INSURANCE INFORMATION

First Name: \_\_\_\_\_ Last Name: \_\_\_\_\_

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## Primary Health Insurance

Type of Policy: EPO HMO PPO POS Other, please specify: \_\_\_\_\_

Authorization required: Yes No Authorization No.: \_\_\_\_\_ Effective Date: \_\_\_\_\_

Insurance Company: \_\_\_\_\_

Address: \_\_\_\_\_ Phone: \_\_\_\_\_

City: \_\_\_\_\_ State: \_\_\_\_\_ Zip: \_\_\_\_\_

Subscriber: Self Spouse Other, please specify: \_\_\_\_\_

Subscriber First Name: \_\_\_\_\_ Subscriber Last Name: \_\_\_\_\_

Subscriber ID Number: \_\_\_\_\_ Subscriber Date of Birth: \_\_\_\_\_

Subscriber Employer: \_\_\_\_\_

Employer Address: \_\_\_\_\_ Group number: \_\_\_\_\_

Address: \_\_\_\_\_ Phone: \_\_\_\_\_

City: \_\_\_\_\_ State: \_\_\_\_\_ Zip: \_\_\_\_\_

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## Secondary Health Insurance

Type of Policy: EPO HMO PPO POS Other, please specify: \_\_\_\_\_

Authorization required: Yes No Authorization No.: \_\_\_\_\_ Effective Date: \_\_\_\_\_

Insurance Company: \_\_\_\_\_

Address: \_\_\_\_\_ Phone: \_\_\_\_\_

City: \_\_\_\_\_ State: \_\_\_\_\_ Zip: \_\_\_\_\_

Subscriber: Self Spouse Other, please specify: \_\_\_\_\_

Subscriber First Name: \_\_\_\_\_ Subscriber Last Name: \_\_\_\_\_

Subscriber ID Number: \_\_\_\_\_ Subscriber Date of Birth: \_\_\_\_\_

Subscriber Employer: \_\_\_\_\_

Employer Address: \_\_\_\_\_ Group number: \_\_\_\_\_

Address: \_\_\_\_\_ Phone: \_\_\_\_\_

City: \_\_\_\_\_ State: \_\_\_\_\_ Zip: \_\_\_\_\_

First Name: \_\_\_\_\_

Last Name: \_\_\_\_\_

## Father

Aortic Dissection  
Bowel Perforation  
Aneurysm  
Stroke or TIA  
Asthma  
Blood Disorder (e.g., anemia)  
Congestive Heart Failure  
COPD (chronic obstructive pulmonary disease)  
Coronary Artery Disease  
Diabetes  
Malignancy  
Neuromuscular Weakness  
Obstructive Sleep Apnea  
Pancreatitis  
Peripheral Artery Disease  
Renal Dysfunction  
Seizures  
Thyroid Disease  
Other (please specify): \_\_\_\_\_

## Mother

Aortic Dissection  
Bowel Perforation  
Aneurysm  
Stroke or TIA  
Asthma  
Blood Disorder (e.g., anemia)  
Congestive Heart Failure  
COPD (chronic obstructive pulmonary disease)  
Coronary Artery Disease  
Diabetes  
Malignancy  
Neuromuscular Weakness  
Obstructive Sleep Apnea  
Pancreatitis  
Peripheral Artery Disease  
Renal Dysfunction  
Seizures  
Thyroid Disease  
Other (please specify): \_\_\_\_\_

## Grandparents

Aortic Dissection  
Bowel Perforation  
Aneurysm  
Stroke or TIA  
Asthma  
Blood Disorder (e.g., anemia)  
Congestive Heart Failure  
COPD (chronic obstructive pulmonary disease)  
Coronary Artery Disease  
Diabetes  
Malignancy  
Neuromuscular Weakness  
Obstructive Sleep Apnea  
Pancreatitis  
Peripheral Artery Disease  
Renal Dysfunction  
Seizures  
Thyroid Disease  
Other (please specify): \_\_\_\_\_

## Other Relatives

Aortic Dissection  
Bowel Perforation  
Aneurysm  
Stroke or TIA  
Asthma  
Blood Disorder (e.g., anemia)  
Congestive Heart Failure  
COPD (chronic obstructive pulmonary disease)  
Coronary Artery Disease  
Diabetes  
Malignancy  
Neuromuscular Weakness  
Obstructive Sleep Apnea  
Pancreatitis  
Peripheral Artery Disease  
Renal Dysfunction  
Seizures  
Thyroid Disease  
Other (please specify): \_\_\_\_\_

*PLEASE NOTE: This section and the sample forms included are NOT intended to be treated as legal advice. Laws pertaining to healthcare matters and patients' rights and wishes vary greatly from state to state. The Marfan Foundation strongly encourages you to contact a legal professional in your state for full and complete guidance and legal advice on these complicated and sensitive issues, both in general and particularly before completing any of the forms included in this packet.*

The legal, ethical, and psychological issues surrounding serious illness and death aren't easy to discuss. But it's far easier on everyone if you have a healthcare proxy, durable power of attorney, living will, and other advance directives in place before you're faced with a serious accident or illness. If you don't have these documents prepared in advance, you may find yourself in a situation in which you're unable to communicate your wishes regarding the extent of treatment efforts, such as resuscitation and life-support machines. The following pages are provided to help you communicate your wishes should you be unable to do so in the event of an emergency. If you have any other questions please feel free to contact us at (800) 8-MARFAN.

Lawyer's Name: \_\_\_\_\_

Law Firm: \_\_\_\_\_

Address: \_\_\_\_\_

City: \_\_\_\_\_ State: \_\_\_\_\_ Zip: \_\_\_\_\_

Phone: \_\_\_\_\_

## **What Is a Power of Attorney?**

A power of attorney is a document in which you state that you give someone else (usually a relative or friend) the authority to make certain decisions and act on your behalf. The person to whom you give these powers is called an "agent" or "attorney-in-fact." You are called the "principal." Just because the word attorney is used does not mean that the person you give authority to has to be a lawyer.

Executing a power of attorney does not mean that you can no longer make decisions; it just means that another person can act for you also. For example, you may be hospitalized for a brief period of time and need someone to deposit your checks in the bank or pay your bills. As long as you are capable of making decisions, the other person must follow your directions. You are simply sharing your power with someone else. You can revoke the agent's authority under the power of attorney at any time if you become dissatisfied with what they are doing. A power of attorney ends upon your death. Thereafter, your will, or the law of intestacy, governs the handling of your estate. A power of attorney document is not a substitute for a will. © Copyright, 2005 Legal Services for the Elderly

## **What Is a Healthcare Proxy?**

A healthcare proxy is a written document used by any competent person to authorize another person, usually a family member, to make healthcare decisions if the person who signs the proxy becomes unable to do so.

When you complete a healthcare proxy, you can either give complete authority to your "proxy" to make all decisions regarding your healthcare, or you can give specific instructions to the person

you designate regarding specific issues (such as your desire to have your breathing artificially maintained by medical equipment, etc.).

## ► **Why do I need a healthcare proxy?**

Without a healthcare proxy, your doctor may be required to provide you with medical treatment that you would have refused if you were able to do so. For example, your doctor may be required to provide you with artificial nutrition and hydration, a respirator, or CPR, even though you are in a coma with no hope of recovery, or are terminally ill.

## ► **When does it take effect?**

The healthcare proxy becomes effective only when you become unable to make decisions, as determined by a physician. Until then, you continue to be in charge of making your own healthcare decisions. It can be revoked orally, and you always have the right while competent to sign a new healthcare proxy.

## ► **How is a healthcare proxy different than a power of attorney?**

A healthcare proxy is different than a power of attorney. A power of attorney primarily authorizes the person you designate to make financial decisions for you. It cannot be used to make healthcare decisions. You must complete a healthcare proxy in order to have an agent make healthcare decisions when you are not able.

## ► **What is the difference between a healthcare proxy and a living will?**

A healthcare proxy is also different than a living will, although each serves the same purpose of allowing you to make decisions in advance about your healthcare. A living will is a document that you sign in advance in which you specifically set forth your decisions about healthcare treatment. Unlike the healthcare proxy, however, it does not authorize you to appoint an agent to make decisions that you did not anticipate when you completed the living will.

The healthcare proxy provides specific instructions and also designates an agent to make decisions when there are events you did not anticipate.

## **What Is a Living Will?**

Many people recognize that death is as much a part of the life cycle as birth, growth, maturity, and old age. Some states allow persons to manage their final illness through a “living will,” a legal document of healthcare instructions. In some states this document simply provides directions and instructions to your doctor. In other states, it also permits you to appoint a healthcare proxy—a person who can make decisions for you when you are not able to do so because of illness or incapacitation. Some states call this document a living will while others call it an “advance directive.” It may include a directive to physicians to withhold or withdraw life-sustaining procedures under certain circumstances.

A living will or advance directive is effective from the date it is executed until you die or until the directive is revoked. If more than one living will or advance directive has been executed, the last one to be executed will control.

Living wills vary between states.

**Healthcare Proxy (SAMPLE)**

**Durable Power of Attorney for Healthcare**

I, \_\_\_\_\_ (your name), hereby appoint \_\_\_\_\_

\_\_\_\_\_  
(Name, home address, telephone number) as my healthcare agent to make any and all healthcare decisions for me, except to the extent that I state otherwise. This proxy shall take effect when and if I become unable to make my own healthcare decisions.

Optional instructions: I direct my agent to make healthcare decisions in accordance with my wishes and limitations as stated below, or as he or she otherwise knows. I have discussed with my healthcare proxy my wishes regarding artificial hydration and nutrition. (Attach additional pages if necessary.)

(Unless your agent knows your wishes about artificial nutrition and hydration feeding tubes, your agent will not be allowed to make decisions about artificial nutrition and hydration.)

Name of substitute or fill-in agent if the person I appoint above is unable, unwilling or unavailable to act as my healthcare agent: (Name, home address, telephone number)

\_\_\_\_\_  
Unless I revoke it, this proxy shall remain in effect indefinitely, or until the date or conditions stated below.

This proxy shall expire (specify date or conditions, if desired): \_\_\_\_\_

Signature \_\_\_\_\_ Date \_\_\_\_\_

Address \_\_\_\_\_

Statement by Witness (must be 18 or older. Witness cannot be the person chosen as the healthcare proxy.): I declare that the person who signed this document is personally known to me and appears to be of sound mind and acting on his or her own free will. He or she signed (or asked another to sign for him or her) this document in my presence.

Witness 1 name (print) \_\_\_\_\_ Signature \_\_\_\_\_

Address \_\_\_\_\_

Witness 2 name (print) \_\_\_\_\_ Signature \_\_\_\_\_

Address \_\_\_\_\_

**COPIES OF THIS FORM SHOULD BE GIVEN TO YOUR HEALTH CARE PROXY, YOUR DOCTOR AND YOUR ATTORNEY. KEEP AN EXTRA COPY FOR YOUR RECORDS.**

*PLEASE NOTE: This form is provided as an example, it is not intended to provide legal advice and should not be completed without the advice and assistance of an attorney in your state who is generally knowledgeable in matters relating to healthcare and patients' rights.*



**Non-Hospital Order Not to Resuscitate (SAMPLE)**

Person's Name: \_\_\_\_\_

Date of Birth: \_\_\_\_\_

DO NOT RESUSCITATE THE PERSON NAMED ABOVE.

Physician's Name (please print): \_\_\_\_\_

Physician's Signature: \_\_\_\_\_

License Number: \_\_\_\_\_

Date: \_\_\_\_\_

It is the responsibility of the physician to determine, at least every 90 days, whether this order continues to be appropriate, and to indicate this by a note in the person's medical chart. The issuance of a new form is NOT required, and under the law this order should be considered valid unless it is known that it has been revoked. This order remains valid and must be followed, even if it has not been reviewed within the 90 day period.

Adapted from the New York State Department of Health

*PLEASE NOTE: This form is provided as an example, it is not intended to provide legal advice and should not be completed without the advice and assistance of an attorney in your state who is generally knowledgeable in matters relating to healthcare and patients' rights.*

# THE VEDS MOVEMENT

*Charging forward. Saving lives.*

## VEDS: THE BASIC FACTS

VEDS (Vascular Ehlers-Danlos Syndrome) is a life-threatening genetic condition of the body's connective tissue. Knowing the signs of VEDS, getting a proper diagnosis, and receiving the necessary monitoring and physician care can enable people living with VEDS to have the best chance at survival.



Our community of experts estimates that many people who have VEDS do not know it. Without proper diagnosis and treatment, they are at high risk for an early sudden death.

VEDS affects our connective tissue, which helps to hold the body's cells and tissues together.

There are also several disorders related to VEDS that cause people to struggle with the same or similar physical problems, and anyone affected by these disorders also needs an early and accurate diagnosis.

### ► What are the features of VEDS?

Every person's experience with VEDS is slightly different. Some may have every feature, some may have only a few features, and other people have different combinations of features. Some people may not have outward signs at all. Some features of VEDS are easier to see than others. These include:

- Thin, translucent skin with increased vein visibility
- Characteristic facial appearance (thin lips, small chin, thin nose, large or deep-set eyes)
- Premature aged appearance of the hands and feet (acrogeria)
- Hypermobility of small joints ("double-jointedness")
- Gum recession and fragility
- Born with a hip dislocation or clubfoot

# **VEDS: THE BASIC FACTS** *(continued)*

- Tendon and muscle rupture
- Keratoconus (cornea of eye bulges outward causing thinning)
- Early onset varicose veins
- Spontaneous or easy bruising that is not explained by other causes
- Sleeping with the eyes open or partially open

## **Complications of VEDS**

Complications of VEDS that may not be apparent from outward appearance include:

- aneurysms,
- artery dissections,
- and arterial tears (including the aorta),
- sudden lung collapse,
- bowel perforations,
- carotid-cavernous sinus fistula (CCSF)
- uterine rupture during pregnancy

Special tests are needed to detect these complications, and these complications may present as emergencies.

### ► **What causes VEDS?**

VEDS is caused by a change (mutation) in the gene that tells the body how to make collagen III, a protein that is an important part of connective tissue. This mutation creates different VEDS features and causes medical problems.

### ► **Who has VEDS?**

- It is estimated that 1 in 40,000-60,000 people have VEDS, but is often not recognized. This includes men and women of all races and ethnic groups.
- People can inherit VEDS; that is, they get the mutation from a parent who has it. This happens in about half of people with VEDS. Other people have a spontaneous mutation, meaning that they are the first in their family to have VEDS.
- People with VEDS have a 50 percent chance of passing the mutation on each time they have a child.
- People are born with VEDS, but many of the features present throughout early life may not be recognized until after a major medical event.

### ► **What is life like for someone with VEDS?**

Most people with VEDS work, go to school, and enjoy active hobbies, and may need to adapt their physical activity to reduce their risk of injury. With an early diagnosis, helpful medical monitoring and activity adjustments can begin early in life. In general, they should not play active team sports that have a risk of collision. In addition, they should not lift heavy objects when at work, home, or the gym. Someone with VEDS may have a reduced quality of life due to the nature of the disorder, unpredictable presentation of medical emergencies, lack of a cure or treatment, and reduced life expectancy.

# **VEDS: THE BASIC FACTS** *(continued)*

## ► **What should you do if you suspect VEDS?**

If you suspect that you or a loved one have VEDS, look for a doctor who knows about VEDS and make an appointment to be evaluated. Keep in mind that you can have VEDS features, but not meet the requirements for a firm diagnosis. Regardless of the diagnosis, it is important to get treatment for the features you have and follow-up with your doctor as they recommend. Genetic testing for an alteration in COL3A1 provides a definitive diagnosis when present and lack thereof can shift clinical care and concerns for other related conditions.

## ► **How is VEDS diagnosed?**

Doctors experienced with connective tissue disorders can often suspect a VEDS diagnosis after exams of several parts of the body, including:

- A detailed medical and family history, including information about any family member who may have the disorder or who had an early, unexplained, death.
- A complete physical examination and tests to identify VEDS features that are not visible during the physical exam.

Genetic testing is crucial to making an accurate diagnosis and differentiating between other conditions which may resemble VEDS.

## **Do you have questions? Would you like more information?**

- Call our help center, **800-862-7326, ext. 126** to speak with a nurse who can answer your questions and send you additional information.
- Visit our website at **TheVEDSMovement.org**. You can print information that interests you and ask questions online.

# THE VEDS MOVEMENT

*Charging forward. Saving lives.*

## FACT SHEET FOR PARAMEDICS AND EMERGENCY MEDICAL TECHNICIANS

### **VEDS (Vascular Ehlers-Danlos Syndrome)**

#### ► **What is VEDS?**

VEDS is an uncommon (perhaps 1/50,000 people) life-threatening genetic disorder, and an early, accurate diagnosis is essential. VEDS affects our connective tissue, which helps to hold the body's cells and tissues together. Knowing the signs of VEDS can save lives. Our community of experts estimates that nearly half of the people who have VEDS do not know it.

The most common presentation in the emergency situation include: aortic dissection, arterial dissection and rupture, spontaneous pneumo- or hemato/pneumothorax, bowel perforation, carotid cavernous fistula formation, and uterine rupture late in pregnancy.

Some clinical features of VEDS are easier to see than others, and may help in the clinical diagnosis, although not everyone with VEDS has these features. These include thin, translucent skin with increased vein visibility, characteristic facial appearance (thin lips, small chin, thin nose, large or deep-set eyes), premature aged appearance of the hands and feet, hypermobility of small joints, spontaneous or easy bruising, and sleeping with the eyes open or partially open.

### **VEDS and Aortic/Arterial Dissection**

#### ► **What is aortic or artery dissection?**

One of the primary features of VEDS is a fragile vascular system, which is prone to dissection. Dissection can occur in any artery in the body, including the aorta. A dissection is a tear involving the inner layer of the artery wall, which allows blood to enter and creates a separation of the inner and outer layers of the vessel. Dissection of the arteries can lead to a weakening of the outer wall of the affected artery, resulting in rupture or aneurysm formation. In an aortic dissection, occlusion of aortic branch vessels can cause myocardial infarction, pericardial tamponade, stroke, kidney failure, bowel ischemia, paraplegia or limb ischemia. Disruption of the aortic valve can result in valvular insufficiency and cardiac failure. Occlusion of an artery in the head or neck can cause a stroke.

# **FACT SHEET FOR PARAMEDICS AND EMERGENCY MEDICAL TECHNICIANS** *(continued)*

## ► **Why is emergency diagnosis and treatment of aortic or arterial dissection an important issue?**

An aortic dissection that remains untreated will ultimately lead to a fatal rupture. In the absence of urgent surgical intervention, the fatality rate associated with acute aortic dissection that originates near the heart is very high. This makes it essential to evaluate symptoms that could be related to a dissection. Any arterial dissection can be an emergency and evaluation is needed immediately.

## ► **What are the symptoms of aortic dissection?**

The patient with an aortic dissection usually complains of severe pain, most often in the chest (front, back, or both), and commonly between the shoulder blades. Occasionally, the pain may be reported as being in the upper abdomen (if the tear begins in that part of the aorta). The patient may describe the pain as ripping, tearing, or sharp like a knife. It may also be described as pleuritic.

Symptoms and signs of shock are ominous findings, and indicate that the dissection has progressed to the point at which tissue perfusion is compromised. However, dissections can also cause a variety of other symptoms in the extremities: pain, pallor, pulselessness, parasthesias; and paralysis (the 5 Ps). Rarely, if the dissection compromises blood flow to the spinal cord, there may be weakness in one or both legs or arms. In addition, neurologic events that would seem due to a stroke or transient ischemic attack (TIA) may be due to a dissection.

Important points of the physical examination, patient history, and assessment that raise the possibility of an aortic dissection:

- Take note if the patient tells you that he/she has an aneurysm, VEDS, or family history of VEDS or a related condition.

*NOTE: This should alert the EMS provider to consider rapid transport with treatment provided en route.*

- The patient may describe symptoms of shock.
- The patient may describe pain or paresthesias in extremities.
- The patient may describe the pain in the front or back of the chest or upper abdomen as ripping, tearing, or sharp like a knife. At times, it is described as pleuritic.

During the physical examination, the following findings may be noted:

- signs of shock
- pallor, pulselessness, paralysis in extremities
- aged appears of the hands
- thin, translucent skin with visible veins
- hypermobility of the small joints

# **FACT SHEET FOR PARAMEDICS AND EMERGENCY MEDICAL TECHNICIANS** *(continued)*

## ► **What are the symptoms of arterial dissection?**

A patient presenting with an arterial dissection may complain of sudden onset pain, which is often described as “severe,” “sharp,” “tearing,” or “ripping.” It may be located anywhere in the body, depending on which artery is affected. Sometimes the pain is less severe, but the patient still has a feeling that “something is very wrong.” The patient may complain of nausea, shortness of breath, fainting, loss of pulse, paresthesia, or paralysis.

The presentation of an arterial dissection may be very different depending on where the dissection is located. For example:

- If the dissection occurs in the arteries in the head or neck, pain may be accompanied by stroke symptoms, including a droopy eyelid with a small pupil on the same side. A room spinning sensation or dizziness may be present, as well as difficulty seeing.
- If the dissection occurs in the coronary artery, the symptoms may be similar to or cause a heart attack, or cause abnormalities in heart rhythm.
- If the dissection occurs in the aorta, severe pain usually in the chest (front, back or both) but occasionally in the abdomen if the tear begins there.

## **VEDS and Arterial Rupture**

### ► **What is arterial rupture?**

Arterial rupture is a full tear of all the layers of the artery wall. This can occur in any artery in the body and can be fatal if not treated immediately. Emergency care is needed immediately.

### ► **Why is emergency diagnosis and treatment of an arterial rupture an important issue?**

A patient with an arterial rupture is internally bleeding. This is an emergency. If this is not treated, this can be a fatal event. Spontaneous arterial rupture is the most common cause of death in people with VEDS.

### ► **What are the symptoms of an arterial rupture?**

The patient with an arterial rupture usually presents with sudden onset pain, which may be located anywhere in the body. Sometimes the pain is less severe, or if the injury is not sudden but was the result of an injury. Rapid swelling of the area may be present, as well as nausea, clammy skin, shortness of breath, fainting or lightheadedness, loss of pulse, paresthesia, and paralysis. Stroke symptoms may occur if the ruptured artery is in the brain.

## **VEDS and Bowel Perforation**

### ► **What is a bowel perforation?**

Bowel perforation occurs when the wall of the bowels (typically a hollow organ, such as the intestines) tears. In patients with VEDS, a bowel perforation commonly occurs in the sigmoid colon (lower left quadrant of the abdomen). Esophageal and gastric rupture, and small bowel rupture have also been reported.

# **FACT SHEET FOR PARAMEDICS AND EMERGENCY MEDICAL TECHNICIANS** *(continued)*

## ▶ **Why is emergency diagnosis and treatment of a bowel perforation an important issue?**

A bowel perforation allows fecal matter to enter the abdominal cavity, and can result in sepsis and death.

## ▶ **What are the symptoms of a bowel perforation?**

A patient presenting with a bowel perforation will generally complain of abdominal pain that gradually increases and becomes severe or debilitating. This is usually in the lower left quadrant of the abdomen, because the most common area affected is the sigmoid colon. Other symptoms of an intestinal perforation include nausea, vomiting, chills, rectal bleeding, and fever. Although the sigmoid colon is the most common area affected, some patients have experienced bladder or stomach rupture. The symptoms for bladder or stomach rupture may differ from the symptoms for an intestinal rupture.

## **VEDS and Uterine Rupture**

### ▶ **What is a uterine rupture?**

Uterine rupture is typically limited to the third trimester of pregnancy. A uterine rupture occurs when the wall of the uterus tears. The baby may move into the abdominal cavity outside of the uterus. This is a medical emergency for both the baby and the mother.

### ▶ **Why is emergency diagnosis and treatment of uterine rupture an important issue?**

A uterine rupture can cause intra-abdominal bleeding, which can lead to hypovolemic shock and death. The baby is also at risk of injury or death due to compromised oxygen supply.

### ▶ **What are the symptoms of a uterine rupture?**

A patient may present with discomfort in the lower area of the belly present with lower back pain that is intermittent (like uterine contractions). The pain may be acute and severe (can feel like ripping pain) and in some cases be accompanied by shoulder pain. They may be blood in the urine, blood vaginal discharge with dark brown or bright red bleeding. There may also be a change in the contour/shape of the belly where the baby bump has been located. Maternal pulse may be barely palpable and be accompanied by a fast heart rate, new low blood pressure, dizziness, and fainting. Upon examination, there may be abnormalities in the fetal heart rate pattern.

## **VEDS and Carotid Cavernous Sinus Fistula (CCSF)**

### ▶ **What is a CCSF?**

Carotid Cavernous Sinus Fistula formation (CCSF) is a medical emergency which may occur without injury or trauma. CCSF is an abnormal connection between an artery in the neck and the network of veins at the back of the eye. This formation happens as a result of a small tear that sometimes occurs in one of the carotid arteries.



# FACT SHEET FOR PARAMEDICS AND EMERGENCY MEDICAL TECHNICIANS *(continued)*

## ► Why is emergency diagnosis and treatment of CCSF an important issue?

If left untreated, a CCSF can lead to permanent vision loss.

## ► What are the symptoms of a CCSF?

Symptoms of a CCSF may gradually worsen and many initially mistake the CCSF for a sinus infection. The patient will present with redness and pain around the eye, as well as a sudden onset of a swooshing sound in the head. The eye may also bulge, and the patient may experience a slow onset of blurred vision.

## VEDS and Pneumothorax

### ► What is a pneumothorax?

Collapsed lung happens when air or gas collects in the space between the lungs and the chest and prevents the lung from inflating completely.

### ► Why is emergency diagnosis and treatment of pneumothorax an important issue?

Treatment of a pneumothorax varies based on the size of the pneumothorax and severity of the condition. A large pneumothorax left untreated can cause breathing problems and cyanosis.

### ► What are the symptoms of a pneumothorax?

The patient may complain of chest pain, which may be sudden onset, sharp, and lead to feelings of tightness in the chest. They may also experience shortness of breath, a rapid heart rate, rapid breathing, and a cough. They may be fatigued, and in severe cases the skin may develop a bluish color (cyanosis) due to decreased blood oxygen level.

## EMERGENCY IMAGING CONSIDERATIONS

### Imaging in the case of an emergency should include:

CT/CTA, MRA/MRI. Ultrasound, or Echocardiography to determine if the patient is experiencing an acute arterial, aortic, gastric, pulmonary, or obstetric emergency. **Take extreme caution with invasive imaging.**

### **Certain types of imaging can be life-threatening to an individual with VEDS. These include:**

#### Catheter Arteriography

Conventional arteriography involves the use of a catheter, x-ray imaging, and contrast material injection to produce detailed pictures of the blood vessels. A catheter is inserted into an artery and then guided to the area that needs to be examined. THIS TYPE OF ARTERIOGRAPHY SHOULD GENERALLY BE AVOIDED FOR INDIVIDUALS WITH VEDS UNLESS ABSOLUTELY NECESSARY, DUE TO THE RISK OF ARTERIAL RUPTURES AND DISSECTIONS ASSOCIATED WITH THE INVASIVE PROCEDURE. These should be performed with great caution and only to identify life-threatening sources of bleed prior to facilitating treatment.

# **FACT SHEET FOR PARAMEDICS AND EMERGENCY MEDICAL TECHNICIANS** *(continued)*

## **Endoscopy**

Endoscopy produces images of the digestive tract by using a flexible tube with a light and camera called an endoscope. The endoscope is guided down the throat and through the digestive tract, and the procedure typically involves sedation. Endoscopies are discouraged and should be used in individuals with VEDS only to identify life-threatening sources of bleeding prior to facilitating treatment and only if other non-invasive forms of imaging cannot be used.

## **Colonoscopy**

Colonoscopy is used to detect problems in the colon or rectum. During the procedure, a flexible tube with a camera called a colonoscope is inserted into the rectum and guided through the colon. Colonoscopies are routinely performed in the general population for cancer screening, however, in individuals with VEDS routine colonoscopy in the absence of concerning symptoms or a strong family history of colorectal cancers is strongly discouraged due to the risk of bowel rupture during the procedure.

## **EMERGENCY CARE FOR AORTIC DISSECTION**

Basic Life Support (BLS) Advanced Life Support (ALS)

Follow the local or regional treatment and transport standards for SHOCK.

### **For additional information, contact:**

#### **The Marfan Foundation**

22 Manhasset Avenue

Port Washington, NY 11050

516-883-8712 | 800-862-7326

**Marfan.org**

# THE VEDS MOVEMENT

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## LETTER TO THE EMERGENCY DEPARTMENT

The attached letter on Page 24 of this section is a sample letter to the Emergency Department. It is intended to be carried to the Emergency Department with the patient during an emergency, along with the Emergency Card included at the end of this Emergency Preparedness Kit. The information in this letter has been reviewed and approved by The Marfan Foundation Professional Advisory Board (PAB). The PAB is comprised of the leading medical experts on VEDS, Marfan Syndrome, Loeys-Dietz, and other related conditions.

You may wish to take this letter to your physician and have them prepare something similar. They may also use this letter, and attach additional information on another page with their signature.

The VEDS Movement  
 Division of The Marfan Foundation  
 22 Manhasset Avenue  
 Port Washington, NY 11050



*Charging forward. Saving lives.*

Date

To the Emergency Department Triage Nurse/Attending Physician,

\_\_\_\_\_ has VEDS (Vascular Ehlers-Danlos Syndrome), a genetic condition that places them at a high risk of arterial dissection and rupture (including the aorta), hollow organ rupture (including the intestines, gravid uterus and other hollow organs), carotid-cavernous sinus fistula, and pneumothorax.

If this patient presents with severe head, chest, abdominal, back, or limb pain, it should be considered a trauma situation and the patient should be triaged appropriately. The patient may be experiencing a life-threatening emergency.

**An X-Ray is not sufficient to rule out the possibility of an aortic dissection.** Spontaneous arterial rupture is the most common cause of sudden death in patients with this condition.

**This patient's complaint should be immediately investigated using MRA or CTA testing.**

**Certain types of imaging can be life-threatening to this individual** due to the fragility of the tissue. These include:

- Catheter Arteriography.
- Endoscopy
- Colonoscopy

Surgical Considerations:

- Surgical procedures are more likely to be successful when the treating physician is aware of the diagnosis of VEDS and its associated tissue fragility. The use of gentle retraction, pledgeted repairs, and padded clamps for vascular surgery may improve surgical outcomes.

#### The Medical Experts

Dr. Sherene Shalhub, MD, MPH, FACS- Vascular Surgeon (University of Washington)  
 Dr. Peter H. Byers, MD- Geneticist/Internist/Pathologist (University of Washington)  
 Dr. Harry C. Dietz, MD- Geneticist/Pediatrician/Cardiologist (Johns Hopkins Medicine)  
 Dr. Shaine Morris, MD- Pediatric Cardiologist (Texas Children's Hospital)  
 Dr. Mark Lindsay, MD, PhD- Pediatric Cardiologist (Massachusetts General Hospital)  
 Dr. James Black, MD- Vascular Surgeon (Johns Hopkins Medicine)

If you need to reach the medical experts for emergency consultation, contact can be made through their associated hospital's line. If you need more information about this condition, please contact The Marfan Foundation Help and Resource Center: 1-800-862-7326 EXT. 126.

*Sincerely,  
 The VEDS Movement, a division of The Marfan Foundation*

# THE VEDS MOVEMENT

*Charging forward. Saving lives.*

## VEDS INFORMATION LINE FOR EMERGENCY PHYSICIANS

This number contains pre-recorded, basic information about considerations for Vascular Ehlers-Danlos Syndrome (VEDS) for medical professionals to know in an emergency. This information is also available online at **TheVEDSMovement.org/Emergency** or **TheVEDSMovement.org/Emergencies**.

Individuals with VEDS are encouraged to give this information to the emergency department.

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470-563-VEDS (8337)

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*Note: This line is NOT monitored and will not help you decide if you are having an emergency. If you think you are having an emergency, call 911 or visit your nearest emergency room.*

# THE VEDS MOVEMENT

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## MEDICAL ALERT BRACELET

### Another Tool for Ensuring Correct Emergency Care

People with VEDS know they are at increased risk of aortic and arterial dissection, bowel perforation, uterine rupture, and pneumothorax. Often, they are more knowledgeable about their condition than the healthcare providers who treat them. This can be problematic and, in fact, life-threatening in the hospital emergency department when quick diagnosis and treatment of aortic dissection and other emergencies is critical to saving an individual's life.

We strongly urge people with VEDS and related conditions to wear a medical alert bracelet to safeguard their own health.

To ensure your medical ID is the most effective, include the following:

1. Your name
2. Include life-threatening complications. For example: "High genetic risk of arterial/aorta dissection, rupture and organ rupture, pneumothorax and carotid cavernous sinus fistula (CCSF)."
3. In Case of Emergency (ICE): Choose an emergency contact who is familiar with your medical history so they can advocate for you when you need them.
4. Allergies: Include your medication and food allergies. If you have room on your medical ID, you may also choose to include information about the FDA warning on Fluoroquinolones.
5. Put your medical condition last. TIP: Keep the VEDS diagnosis simple. "VEDS" will be more effective than "Vascular Ehlers-Danlos Syndrome" so you are not mistaken for having one of the other forms of Ehlers-Danlos Syndrome.

Although there are many companies that offer this service, one of the most well-known is MedicAlert®. For more information about MedicAlert, call **888-633-4298** or visit **MedicAlert.org**.

# THE VEDS MOVEMENT

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## EMERGENCY ALERT CARD

Print out card for every member of household. Fill in patient name and contact information. Cut along dotted line. Fold vertically and horizontally. Carry this card at all times.

FOLD  
↓



### EMERGENCY ALERT CARD

**DO NOT SEND THIS PERSON HOME UNTIL ARTERIAL/AORTIC DISSECTION OR RUPTURE, PERFORATED BOWEL, PNEUMOTHORAX or CAROTID CAVERNOUS SINUS FISTULA IS RULED OUT**

**This person has VEDS, a genetic condition, which puts him/her at a extremely high risk for:**

- ▶ arterial/aortic dissection/rupture
- ▶ bowel or uterine rupture
- ▶ pneumothorax
- ▶ carotid cavernous sinus fistula

A CTA or MRA are the most definitive tests to identify these complications. Choose one that is most readily available and expertly performed and interpreted. An X-ray does **NOT** rule out the possibility of dissection.

**PROCEED WITH CAUTION AND AVOID INVASIVE PROCEDURES**

Release contrast and fluids slowly. Skin bruises and tears very easily. Monitor blood pressure closely. Do not use strong adhesives.

Important information for medical professionals and contacts for consultants are available.



800-862-7326 | [TheVEDSMovement.org](http://TheVEDSMovement.org)

VEDS (Vascular Ehlers-Danlos Syndrome) is a rare condition that is characterized by a high risk for arterial and bowel rupture, rupture of the uterus toward the end of gestation, spontaneous pneumothorax, and carotid cavernous sinus fistula formation, which may be **life threatening**. Individuals with VEDS have mutations in COL3A1 which encodes the type III collagen chains. Type III collagen is a major component of the walls of vessels, the bowel and other tissues.

**PATIENT NAME:** \_\_\_\_\_

In an emergency situation, contact the following people:

\_\_\_\_\_  
*Name and contact info for primary care physician, geneticist or other specialist*

\_\_\_\_\_  
*Name and contact info for vascular surgeon or other specialist*

**The most common cause of death in a patient with VEDS is spontaneous arterial rupture. 50% of patients diagnosed with aortic dissections die within 48 hours.** Organ rupture can also occur and bowel rupture can cause sepsis quickly. **DO NOT** dismiss these potential events until they have been definitively ruled out.

**Unless absolutely required for diagnosis or care, use of arterial blood gas measurement, arteriography, and colonoscopy are to be avoided because of the fragility of the tissues, particularly the vasculature and bowel. Surgically placed central lines should be restricted to the internal jugular and femoral routines using ultrasound guidance if possible.** If absolutely necessary, endoscopy (colonoscopy, etc) should be done cautiously under low pressure, with consideration of use of a pediatric endoscope.

Signs of an arterial or aortic dissection can include pain, cardiovascular instability, pulselessness, paresthesia, paralysis, syncope, headache, stroke symptoms, or a sense that "something is terribly wrong."

Individuals with VEDS are at increased risk for rapid progression and poor outcome from aortic and arterial dissection. Specialized and aggressive medical and surgical practices that are tailored to this patient population may be needed. If diagnosed with aortic dissection, arterial dissection, or bowel perforation, this patient should be acutely stabilized and then immediately transferred to a tertiary care center with the capability of definitive surgical management if stabilization for transport is possible. If the diagnosis is known, then consultation with experts is advised.

This is the consensus of the Professional Advisory Board of The Marfan Foundation, and is in keeping with evidence-based guidelines for the diagnosis and management of patients with thoracic aortic disease established by the American College of Cardiology Foundation and the American Heart Association in collaboration with eight other professional organizations.

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