

Vascular Ehlers-Danlos Syndrome Information Pack

TABLE OF CONTENTS

Table of Contents	1
What is Vascular Ehlers-Danlos Syndrome (vEDS)?	2
Signs & Symptoms.....	2
Vascular EDS Diagnosis.....	4
VEDS Management Plan	5
Create a Care Team.....	7
Mental Health and Wellbeing:	8
Travel Advice and Insurance.....	10
Financial Assistance.....	11
Creating a Medical Emergency File.....	15
Care Team Template.....	18
School Information Example	19
My checklist	26
Resources.....	27
Contributors.....	27
Reference list	28

WHAT IS VASCULAR EHLERS-DANLOS SYNDROME (VEDS)?

Vascular Ehlers-Danlos syndrome is an inherited connective tissue disorder that is caused by defects in a protein called [collagen](#). It is generally considered the most severe form of [Ehlers-Danlos syndrome](#) (EDS). Common symptoms include thin, translucent skin; easy bruising; characteristic facial appearance; and fragile arteries, muscles and internal organs.^{[1][2]} Vascular EDS is usually caused by a change (mutation) in the [COL3A1](#) gene. Rarely, it may be caused by a mutation in the [COL1A1](#) gene.^[3] Inheritance is autosomal dominant.^[2] Treatment and management is focused on preventing serious complications and relieving signs and symptoms.^{[2][4]}

Source: <https://rarediseases.info.nih.gov/diseases/2082/vascular-ehlers-danlos-syndrome>

SIGNS & SYMPTOMS

We know that day to day, many people with vascular EDS have no problems and live full and enjoyable lives. However, people with vascular EDS do have fragile connective tissues, as the condition is caused by faulty type III collagen.

What are the main signs and symptoms of vascular EDS?

Individuals with vascular EDS may have the following features:

- Tendency to bruise very easily because blood vessels are more fragile.
- Thin skin which makes small blood vessels visible on the upper chest, and legs.
- Risk of hollow organ rupture, most commonly the large bowel. There is also a risk of rupture of the spleen, or the uterus (womb) in pregnancy.
- Fragile blood vessels (arteries) which can lead to major complications, including ruptures, dissections, or aneurysms.

Occasionally there may be other features present including:

- Hypermobility of small joints. (i.e. fingers and toes).
- Premature ageing of the skin on hands and feet.
- Facial features, including a thin nose and lips, large eyes, small earlobes, and fine hair.
- Joint contractures caused by shortening of the ligaments.
- Partial collapse of the lung called a pneumothorax.
- Problems, such as bleeding or receding gums.
- Varicose veins may occur in early adult life.
- Wounds may take longer to heal.
- Clubfoot.

Skin

Although thin skin with readily visible venous patterning is one of the typical features described in individuals with vascular EDS, it is often a subtle finding and bruising that is not explained by trauma is more common.

Acrogeria, is a cutaneous condition characterised by premature ageing, more especially in the form of unusually fragile, thin skin on the hands and feet, however this is uncommon in patients diagnosed with vascular EDS.

Bruising may increase with aspirin or non-steroidal anti-inflammatory medications and certain in the presence of anticoagulant treatment.

Musculoskeletal

Height varies through the normal range although small for family may be more common. Congenital hip dislocation is increased as is congenital talipes and limb reduction defects (perhaps secondary to amputations by amniotic bands) when compared to the average population but these alone are generally insufficient to warrant diagnostic testing.

Distal joint contractures occur in a small proportion of individuals, these are progressive and can be disabling. There also appears to be an increase in the relative frequency of muscle and tendon rupture, but these data have not been aggregated.

COL3A1 Haploinsufficiency

Patients with haploinsufficiency vascular EDS have a different underlying cause of their vascular EDS. Most vascular EDS patients have one 'normal' type III collagen gene (COL3A1) and one 'abnormal' (or mutated) type III collagen gene.

This leads to abnormal collagen fibres, with 7/8ths of the collagen produced being abnormal. Haploinsufficiency vascular EDS is caused by having one 'normal' type III collagen gene and then one gene that does not produce any collagen. This is known as a 'null mutation'. Therefore, these patients only produce normal collagen, but in less quantity compared to a person unaffected by vascular EDS.

As a result, haploinsufficiency vascular EDS tends to have later onset of complications by almost 15 years and extended life expectancy compared to other vascular EDS patients.

Compared with individuals with missense or exon-skipping mutations, it was found that life span was extended, the age of first complication was delayed by almost 15 years, and major complications were limited to vascular events.

VASCULAR EDS DIAGNOSIS

In many people without a family history of the condition a diagnosis of vascular EDS is not considered until they present with a medical emergency such as dissection or rupture of an artery, an organ rupture (for example bowel or womb) or after the discovery of one or more aneurysms (a swelling in an artery).

There are other medical conditions with some of the same symptoms such as Marfan syndrome, Loeys-Dietz syndrome and SCAD therefore it is especially important that a suspected diagnosis of vascular EDS is confirmed by a genetic test.

The gene associated with vascular EDS is called COL3A1 and an alteration in this gene is found in over 99% of people who have vascular EDS. In rare cases, genetic testing does not confirm the diagnosis and a skin biopsy to look at a piece of skin under the electron microscope can help clarify whether vascular EDS is the correct diagnosis.

You will need to see a genetic specialist in order to determine vEDS. This can be done by talking to your GP and/or specialist, who will then set up a referral to the closest genetic specialist to you.

VEDS MANAGEMENT PLAN

You've just been diagnosed, now what? Firstly, stay positive – you are not alone! Support is available to you.

To help you plan ahead we have listed some practical points to consider:

- If you have any upcoming surgery or investigations that will be invasive (such as a camera) please contact your consultant immediately and alert them of your diagnosis.
- Discuss informing your close family members of your diagnosis, they too should be tested for Vascular EDS. Discuss with your geneticist about getting a 'Family Group Number' which can be given to family members to help streamline the process of getting tested.
- Consider which friends you are going to tell about your diagnosis. Give thought to how much information you want to share and who with.
- Contact your GP and start the process of compiling a medical care team. Unfortunately, due to the rarity of this condition you and your family members will need to become your own advocates to inform consultants on Vascular EDS.
- Research your local hospital, can it perform Vascular surgery? Does it have any consultants who are aware of Vascular EDS?
- Prepare a file to keep your [Emergency Information Sheet](#), [Care Team List](#), and a copy of the letter of Diagnosis from your diagnosing doctor. A copy of your file will possibly need to be kept by your workplace in case any issues arise while at work.
- If it is a child who has been diagnosed, you will need to arrange a meeting with the school to discuss Vascular EDS with them. If possible, include the child's Principal, main Teacher and any Teacher Aids that work in your child's classroom. Make sure they have a copy of the [Emergency File](#) and [Care Plan](#) outlined below.
- Begin to think about sporting activities. You are advised to stop participating in any contact sports, lifestyle changes will also need to be considered. If you have to stop doing an activity such as soccer or rugby, focus on what alternative activity can be done instead. This also applies to children who have been diagnosed, encourage them away from contact sports.
- Look at joining the [Ehlers-Danlos NZ Society](#) to stay up to date with information on EDS in NZ, as well as joining our support group on [Facebook Loosely Speaking](#), and finding a [Support Hub](#) within your region so you can talk to other EDSers both on a national level as well as a local level.

- With a diagnosis of Vascular EDS we recommend getting a [MedicAlert bracelet](#). This will help medical professionals identify quickly that you have vEDS in the event you suffer a medical emergency and are unable to verbalise your condition. With vEDS, every second counts. Please discuss this with your GP or Specialist.
- If you are struggling financially, please have a chat with [Work and Income NZ](#), you may be entitled to benefits and extra financial assistance to cope with medical expenses and equipment you may need. You may also be entitled to financial support for travelling to-and-from medical appointments, this is called [National Travel Assistance](#) and you can find more information on the [health.govt](#) website.

CREATE A CARE TEAM

It is not uncommon for someone with Vascular EDS to attend their local Emergency Department only to find that the medical staff have never heard of the condition. This is understandable because Vascular EDS is a rare condition.

It is therefore important that you inform them of your Vascular EDS diagnosis. It is helpful to be able to show proof of your diagnosis and give some information about what the diagnosis means.

Patients with Vascular EDS have said that it can be hard to get medical staff to acknowledge their diagnosis, which is obviously frustrating. If any medical staff want further advice it may be a good idea to give them details of your main consultant, or cardiologist.

Each individual with Vascular EDS should have a primary clinician who acts as the care coordinator, and who is linked to a geneticist or other specialist with detailed knowledge of the disorder. The care team should include the primary care practitioner, a vascular surgeon, and a general surgeon. Your GP should also be included in your team, as most often your GP is the one who will be doing referrals and taking care of any prescription medications you may need.

This team exists to care for the affected individual in the case of major complications, such as bowel rupture, arterial dissection, or rupture. A clear protocol should be created, the local emergency department should have data about the individual and both the affected individual and relevant family members should know the protocol for contact.

A protocol for the emergency department evaluation needs to be established and the on-call clinicians must recognise the needs of the patient. Each affected individual should have a diagnostic letter and Emergency Information Sheet to be carried and provided to the Emergency Department clinicians at the time of consultation.

For younger women planning pregnancy a high-risk team should be assembled that includes experienced obstetricians and vascular surgeons.

The goals of medical management are to minimise the likelihood of adverse events and to assure that quality of life is minimally impaired.

The general approach to medical management includes the creation of an informed care team in the following areas of expertise:

- Vascular Surgery
- General Surgery
- Cardiology
- Gastro
- Physiotherapy
- Neurology
- Dentistry
- Genetic Counselling
- Rheumatology

MENTAL HEALTH AND WELLBEING:

Receiving a diagnosis or living with a rare form of EDS can be exceedingly difficult emotionally and can have a huge impact, including anxiety, stress, low mood and emotional exhaustion. This is a normal response, and it is common for people to feel they would like some more emotional support.

Finding out about the possibility of EDS in general can bring a huge variety of emotions and thought processes. Each variation of EDS can bring about their own feelings and thoughts unique to that particular prognosis, vEDS is no different. By the nature of vEDS, there are a number life altering impacts that can affect mental health. It's understandable and very normal to have your mental health affected, nothing at all to be judged for or beat yourself up about.

You may find you grieve all the different things you had thought about yourself and your future. The hopes you had for yourself, your career, travel plans, family, and children. All these things may still be achievable but may need to be done in a different way to what you had pictured. All these things may lead to times when you feel down or maybe even lead to depression.

Feeling down or experiencing depression is a very normal reaction. It's how we handle the down feelings or depression that can alter how they affect our day-to-day lives. There is no one single list of symptoms of depression, equally; there is now definitive list of causes of depression. It's what we do in these times of struggle that make an impact.

So what are some things that can help?

- Finding a supportive empathetic GP - Getting support for mental health and possibly referrals to other support services
- Talking to family and friends
- Finding and doing things you enjoy. for example: reading, walking, being in nature, listening to music etc. things that are manageable for you in that moment that might bring a little enjoyment and happiness to your world.
- Reaching out and asking for help - through loosely speaking, calling or texting helplines, family and friends.
- Making time to sit and feel what you are feeling - that's ok and healthy, sometimes its needed before you can identify what would be helpful in moving forward.

The following information may be of help in providing possible avenues of support and information. Please note that the following information is provided in good faith and as a guide only.

MINISTRY OF HEALTH New Zealand Government Ministry of Health

<https://www.health.govt.nz/>

PRIVATE COUNSELLORS AND THERAPISTS List of counsellors, psychotherapists and psychologists who are registered with a recognised New Zealand professional body.

<https://www.talkingworks.co.nz/>

GENERAL MENTAL HEALTH HELPLINES

Crisis helpline - 0508 826865 free, nationwide service available 24 hours a day, 7 days a week and is operated by highly trained and experienced telephone counsellors who have undergone advanced suicide prevention training.

<https://www.lifeline.org.nz/services/suicide-crisis-helpline>

Healthline - 0800 611 116 - for medical advice from trained nurses

<https://www.health.govt.nz/your-health/services-and-support/health-care-services/healthline>

Need to talk (free text or free call) - 1737 - to talk to trained and qualified counsellors

<https://1737.org.nz/>

Youthline - 0800 376 6333 - free text – 234 <https://www.youthline.co.nz/>

Anxiety helpline - 0800 269 4389 <https://www.anxiety.org.nz/>

Depression helpline - 0800 111 757 <https://www.depression.org.nz/>

Lifeline - 0800 543 354 - 24/7 to speak with trained support staff who can also help link you to ongoing services. <https://www.lifeline.org.nz/>

The Samaritans - offers free, confidential, 24-hour emotional support over the phone (0800 72 66 66). <https://www.samaritans.org.nz/>

Mental Health Foundation - The Mental Health Foundation of New Zealand is a charity that works towards creating a society free from discrimination, where all people enjoy positive mental health & wellbeing. <https://www.mentalhealth.org.nz/>

Changing Minds - Changing Minds is a national not-for-profit organisation operated entirely by those with personal experience of recovery from mental health and/or addiction issues, we work in the broad area of wellbeing services, advocacy, human rights, and health policy. <https://changingminds.org.nz/>

GROW - a Community for Mental Health and Personal Development <https://grow.org.nz/>

EDS CHARITIES AND SOCIETIES -

Ehlers-Danlos Syndromes New Zealand - A society to raise awareness, and support patients, within New Zealand of all Ehlers-Danlos Syndromes and Hypermobility Syndromes <http://ehlers-danlos.org.nz/>

The Ehlers-Danlos Society - The International Society for EDS, to spread awareness and make changes within the medical system for the support of those with EDS/HSD www.ehlers-danlos.com

Annabelle's Challenge Vascular EDS Charity www.annabelleschallenge.org

TRAVEL ADVICE AND INSURANCE

Flying

There is no indication against being on an aeroplane with a diagnosis of Vascular EDS, but it is worth considering the risk associated with long haul flights. If a medical emergency were to happen mid-flight there would be a considerable delay before any medical assistance would be available.

Assisted Travel

Book special assistance at the airport for your outbound and inbound journey. Most airports provide this service and a patient diagnosed with Vascular EDS is no exception. Special assistance is also provided by most train operators including wheelchair ramps.

Please note: It is advisable to check with your operator/airport and book in advance.

Airport Sunflower Lanyards

By wearing one of the airport's Sunflower Lanyards, it will discreetly indicate to staff that you have a hidden disability, and you would like additional support. Airport staff have been trained to recognise these lanyards, and to provide you with any help you may need at various stages at the airport.

The lanyard also "permits access to the family and priority lanes at security as well as the use of special assistance lanes on arrival at the airport." It will also allow you priority boarding on to the aircraft and special assistance at the onward destination airport.

Please note: Please contact the airport customer service team directly who will be able to provide you with information on their current services and a lanyard, if available. As of December 2020 this initiative is being launched in New Zealand, so may not currently be available at all airports.

Travel Plans

We recommend planning for your vacation, making travel plans will not only help to ensure your safety, but it will also ensure you maximise your time away so you can enjoy your break.

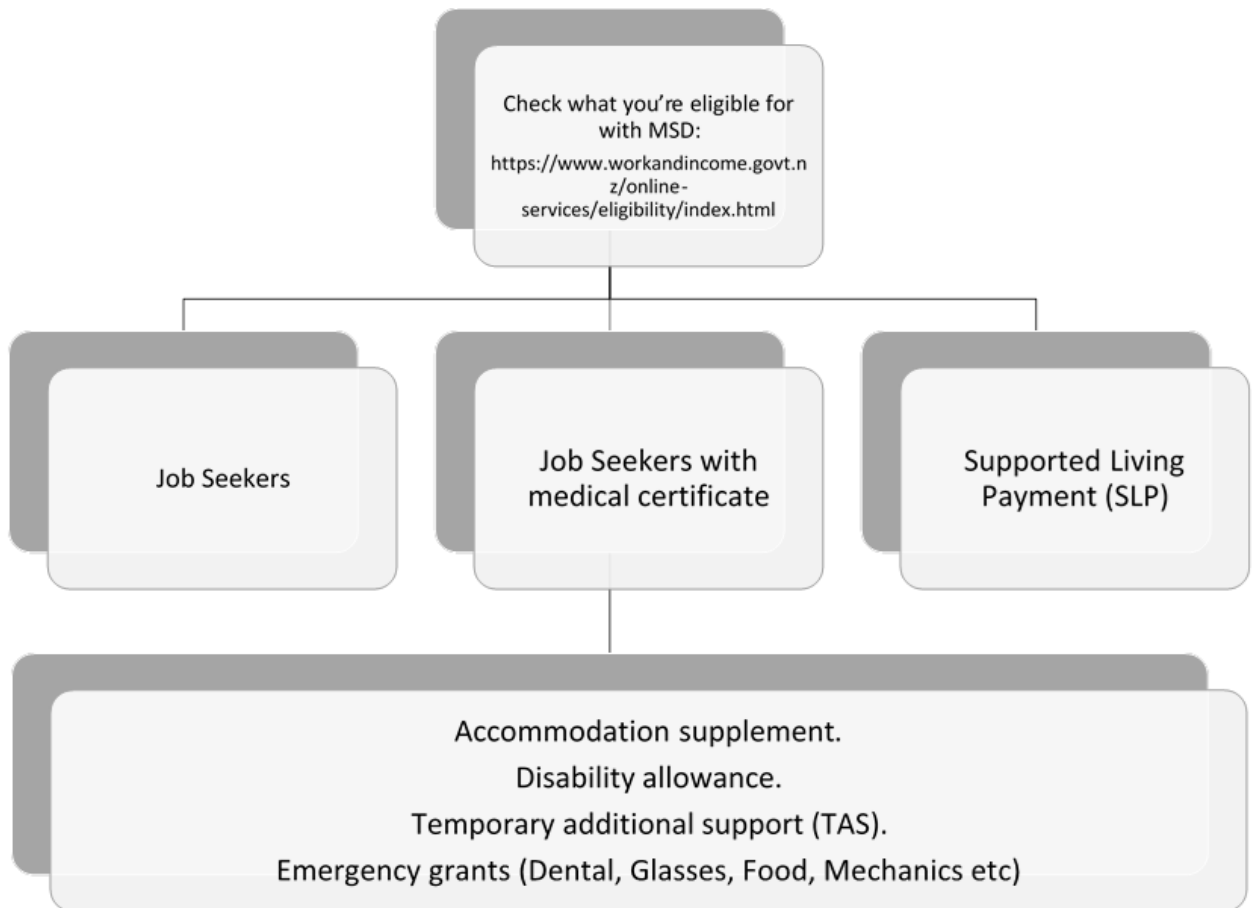
- Where is the nearest hospital with emergency care?
- Take your Vascular EDS Emergency File.
- Wear your MedicAlert bracelet if you have one.
- Purchase travel insurance.
- Most importantly, make memories which will last a lifetime!

Travel Insurance

Travel Insurance is essential and should not cost you significantly more because of a diagnosis of Vascular EDS. It is well worth shopping around and visiting comparison websites to get the best package at the right premium.

FINANCIAL ASSISTANCE

Living with a chronic condition like EDS, where every day can be different to the one before, it can be hard to continue working and maintain your physical health. Here we have a list of financial services that you may be entitled to.



In depth details of the above

[Jobseekers Support](#) – This payment is for people who are between jobs or returning to work after a period away.

To apply:

Fill in the online application form or pick up a paper copy from your local MSD office. This form will also tell you what you need to do next, documentation you need to give to MSD, how the process of receiving the Jobseeker support works and the terms and conditions that need to be met and abided by.

[Jobseekers Support with Medical certificate](#) – This is for people who are likely to be able to work a minimum of 20 hours per week within the next 2 years but are presently unable to work due health struggles.

To Apply:

- Fill in the online application form or pick up a paper copy from your local MSD office. This form will also tell you what you need to do next, documentation you need to give to MSD, how the process of receiving the Jobseeker support works and the terms and conditions that need to be met and abided by.
- You will also need to see your GP who fills in a digital medical certificate (they already have this in their systems). This certificate will state your medical conditions along with estimation of work availability. This then gets sent automatically to MSD which then uses your name and personal details to be linked to your application.

[Supported Living Payment \(SLP\)](#) – This payment is for people who have long term health challenges and are medically deemed unable to work for a minimum of 2 years.

To apply:

- You can either fill in the form online / download and print / pick up a copy of the form from your local MSD office.
- Once this form is filled in, you then need to see your GP who needs to give you an MSD medical certificate stating you will be unable to work for a minimum of 2 years and that you will be unable to work more than part time hours.
- It can also help to have a letter from your GP, specialist, physio etc reinforcing the diagnosis and that you won't be able to work more than 20 hours per week for a minimum of 2 years. This isn't necessary for the application but can help minimise possible questions.
- Your application then goes to a review board who will access your application. They can request you to see a Dr of their choosing (at their expense) for an impartial 2nd opinion. This isn't always required but if it is, I suggest you take the [diagnostic criteria](#) with you and any other documentation you have that may help clarify your situation. The report then gets sent back to MSD and added to your file to be assessed again then have the decision made.

Additional application information:

[Accommodation Supplement](#) - The amount of this payment is defined by the area you live and the type of accommodation (rent/ board or home mortgage with rates etc). Link:

To apply

- Download and print or pick up an application form from your local MSD office.
- You need proof of your accommodation costs (rent/ board/ mortgage payments/ rates etc) to include in the application.

- Take this to your appointment with MSD.

[Disability Allowance](#) - This is to help pay for ongoing medical expenses important to your health so can vary from person to person. Costs that can be covered are: GP visits, Prescriptions, Natural supplements, Alternative treatment (physio. osteopath, chiropractor etc), Clothing needed for your health, Bracing, Pool / Gym memberships, Dietary needs etc. The full list can be found [here](#).

To apply:

- The application can be found as part of the SLP form / download from website and print / pick up a copy from your local MSD office.
- Make a list of all your expenses related to your medical needs for a year (including costs that are yet to happen)
- You will need your GP to sign a medical certificate that is included in the application form endorsing these expenses.
- Take this application to your MSD appointment which will then be processed broken down into weekly amounts which are paid into your account as part of your overall payment.
- The disability allowance has a total sum of approximately \$64 per week.

[Temporary Additional Support \(TAS\)](#) - This is to help with costs that are uncovered by the different supplementary, for example: medical costs above the \$64 p/w for the disability allowance.

To apply:

- The application can be found online to download and print or pick up a copy from your local MSD office.
- You need to fill in the form including sharing what you are doing to try and reduce your costs.
- Take this form with you to your MSD appointment or drop into your local MSD office for them to process.
- The TAS is reviewed every 3 months and can be reapplied for online via your 'My MSD'.

[Emergency Grants](#) - These are there to help cover a variety of costs (emergency dental, car repairs, travel for medical needs or funerals or tangi's, medical appointments that are with people who aren't available to see through your local DHB or if you need to see someone else as a result of being declined from your DHB. Emergency costs that are unexpected but are necessary to supporting daily life).

To apply:

- Get a quote for the expense from the appropriate business.
- Phone 0800 559 009 and say "Emergency costs" then follow through with the voice prompts.

- These costs generally need to be paid back and are at the discretion of MSD as to if you are eligible to receive the funding.

[My MSD](#) - through this service, you can: see your weekly payment, apply for a food grant, check your personal detail among a variety of other things.

You may not always need to have an in-person appointment for these applications, there is now a facility online via 'My MSD' to upload documentation. This facility needs the person you speak with at MSD to make it available and they will talk you through how to use it.

If there is a decision made in relation to any of your applications that you disagree with, you can request a [Review of the Outcome](#).

You may also be entitled to financial support for traveling to-and-from medical appointments, this is called [National Travel Assistance](#) and you can find more information on the Government Health website.

CREATING A MEDICAL EMERGENCY FILE

There are several things that should be contained within your Medical Emergency File, and here we will go into more detail on this. We also have easy printable resources to help you create your Medical Emergency File. You will need several copies of your file - One for at home, one for your car or to be kept with you at all times, and one for the workplace, or if it is a child, at their school.

Your file should include:

- A copy of the letter of diagnosis from your specialist. Photocopy your letter and print a copy for each of your files.
- [Emergency Information Sheet and Care Team List](#)

For a child you will also want to include a [Care Plan](#) for the school as well as a [PE Risk Analysis Form](#), we have given samples below.

Please feel free to save these two packs to your computer and edit them to include the information that is relevant to you, and then print them to add to your Medical Emergency Files.

IN CASE OF EMERGENCY

The following resources should be presented to the attending ambulance crew and hospital staff in the event of an emergency or planned surgical procedure.

Contents:

- Emergency Information Sheet
- Copy of letter of diagnosis.
- My Care Team

We advise that you keep copies of your emergency information in the home as a 'grab and go', in your car, and at school or the workplace.

Emergency Information for Medical Professionals

Vascular Ehlers Danlos Syndrome

Vascular EDS is a life-threatening connective tissue disorder that affects all tissues, arteries and internal organs making them extremely fragile. Patients are at risk of sudden arterial or organ rupture. This can occur at any age. Mid-size arteries are commonly involved.

Patient concerns should be taken seriously, and any reports of pain need full and immediate investigation.

Presenting symptoms

- Arterial or intestinal rupture commonly presents as sudden acute abdominal, chest or pelvic pain, that can be diffuse or localised.
- Cerebral arterial rupture may present with altered mental status and be mistaken for drug or alcohol use.
- Redness, pain, and prominence of one or both eyes and the sound of pulsations in the head can be manifestations of a carotid-cavernous fistula.
- Coronary dissection may present with acute myocardial infarction. Carotid dissection may present with stroke. Limb arterial dissection can present with acute limb ischaemia or claudication.
- Colonic perforation can present with acute abdominal pain and/or signs of infection.
- Pneumothorax can present with pain and shortness of breath.

Management guidance

The fragility of all tissues means that invasive procedure should be avoided where possible. All members of the medical team should be aware of the potential risk for greater than usual harm.

- Vascular dissection and rupture or bleeding can be subtle in presentation, therefore a lower threshold for investigations and imaging is indicated.
- Immediate investigation by MRI or CT scan should be performed.
- Use non-invasive techniques only, avoiding stress and tension on skin, organs, or vessels during physical examination.
- Avoid angiography, enemas, and endoscopies.
- Avoid intramuscular or subcutaneous injections of heparin or heparin substitutes, as these can cause massive subcutaneous haematoma and bruising.
- Central lines should be placed only with ultrasound guidance to avoid inadvertent arterial injuries.
- Bleeding into the body wall or cavity should be managed conservatively with transfusion and support.

Emergency surgery

Surgical risks are higher for Vascular EDS patients. The threshold for intervention should be higher. All conservative management options should be carefully considered before surgery.

- The primary indication for surgical intervention is life threatening complications of arterial or organ rupture.
- A vascular surgeon should be present during surgery.
- The anaesthetist should be aware of fragile mucus membranes when intubating.
- Self-retaining retractors should be used carefully, excessive retraction leads to multiple tissue tears and haematomas.
- Tissues are fragile and do not hold sutures well.

TREATMENT WITH BLEEDING IN THE EMERGENCY

Should the patient have a cut or injury that requires surgery, then it is important that there are plastic surgeons who are involved as the surgical techniques required are not straightforward and require plastic surgery expertise. There may also be a place for intravenous or oral Tranexamic Acid (TXA) following on from the first line treatment. If an anaesthetist is required, they should be made aware that patients with vascular EDS can take longer to be anaesthetised and the performance of the anaesthetic should be closely monitored.

For more information on Surgical Intervention with vEDS please click [here](#).

Vascular EDS Medical Emergency?
YOU MUST CALL 111

CARE TEAM TEMPLATE

Once you have established a care team specific to your needs, please complete the sections of care that are relevant to you and add to the [Emergency Section](#) of this pack. Examples below:

What Services are involved?				
Doctor's Name	Service	Contact Details	Phone Number	Dates of Involvement
	Genetic Counselling			Reviewed annually unless there are any issues
	Paediatrician			Reviewed annually unless there are any issues
	Cardiology			Reviewed annually unless there are any issues
	Rheumatology			Reviewed annually unless there are any issues
	Dental			Every 6 Months

SCHOOL INFORMATION EXAMPLE

Template for school: Individual healthcare plan with Vascular EDS

Name of School/Setting	
Childs Name	
Group/Class/Year	
Date of Birth	
Child's Address	
Medical Diagnosis or Condition	Vascular Ehlers-Danlos Syndrome (Insert any other/comorbid conditions the school needs to be aware of)
Date	
Review date	

Family Contact Information

Primary Family Contact Name	
Relationship to Child	
Phone Number Home	
Phone Number Mobile	
Phone Number Work	

Secondary Family Contact Name	
Relationship to Child	
Phone Number Home	
Phone Number Mobile	
Phone Number Work	

Clinic/Hospital Contact

Name	
Contact Phone Number	

G.P.

Name	
Contact Phone Number	

Who is responsible for providing support in school	
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Describe Medical Needs and give details of child's symptoms, triggers, signs, treatments, facilities, equipment or devices, environmental issues, etc.

Vascular EDS is a life-threatening connective tissue disorder that affects all tissues, arteries and internal organs making them extremely fragile. Patients are at risk of sudden arterial or organ rupture. This can occur at any age. Mid-size arteries are commonly involved.

Tendency to bruise very easily because blood vessels are more fragile.

Thick skin which makes small blood vessels visible on the upper chest, and legs.

Risk of hollow organ rupture, most commonly the large bowel. There is also a risk of rupture of the spleen.

Fragile blood vessels (arteries) which can lead to major complications including ruptures, dissections, or aneurysms. These can happen spontaneously and with truly little warning or signs of pain.

Avoid contact sports, such as team games, football, rugby, tennis, boxing, or martial arts.

Can suffer from fatigue and allowances should be given for time out to recover.

Avoid sudden changes of load e.g. lifting very heavy weights, sudden changes of acceleration (sprinting), or isometric exercises such as weight training.

Brass and woodwind musical instruments also involve physical exertion, so alternative instruments may be more suitable.

All teachers and staff should be made aware that <Pupil> has Vascular EDS.

Flexible joints which may lead to dislocation.

Name of medication, dose, method of administration, when to be taken, side effects, contra-indications, administered by/self-administered with/without supervision.

Daily care requirements

<Pupil> should have support in class and around school in the event of a sudden change to health i.e. risk of spontaneous arterial or organ rupture.

Assisting <Pupil> whilst moving around school will significantly reduce injuries from bumping into other students and furniture. A fellow pupil 'buddy' should be considered to assist <Pupil> in and around school when needed.

Specific support for the pupil's educational, social, and emotional needs

School to adapt activities to allow full inclusion where at all possible or provide alternatives. Although there will be some limitations to physical activities, it is important that opportunities to maximise participation are provided. This would ensure the best learning environment so that the child is empowered to lead a full, rewarding and as normal a life as possible, without putting them at unnecessary risk during school years.

Arrangements for school visits/trips etc

<Pupil> should not be excluded from school visits/trips. They should be supported by either parent or Teacher's Aid (TA). Avoid activities that include contact sports.

Other information

Should <Pupil> be taken to hospital with any cuts, under no circumstances should it be stitched if required until a plastic surgeon has assessed him/her, due to the fragility of the skin.

MedicAlert Number: < >, contact number for MedicAlert 0800 840 111

Describe what constitutes an emergency, and the action to take if this occurs.

Falling, banging into someone or something, bruises appearing anywhere on the body. Extra care/vigilance/supervision when <Pupil> is accessing gross motor activities.

Tiredness, lethargy/passing out could be indications of a rupture – **Call 111** then call family.

Sudden intense pain – **Call 111** then call family.

Who is responsible in an emergency (state if different for off-site activities)?

Plan developed with

Parent, TA, Head Teacher, and any other relevant representatives

Staff training needed/undertaken – who, what, when

All staff to be made aware of the pupil's condition including support staff/ temp staff.

P.E Risk Analysis Template Example

Activity		Risk Factors	Low Risk	Med Risk	High Risk	Could <Pupil> Do warm up exercises then Referee. Yes/No
Swimming		People swimming into <Pupil>, overcrowded pool			*	
Baseball/Softball/T Ball		Ball or Bat injury			*	
Soccer		Contact Sport, tackling, ball injury, joint injury			*	
Hockey		Hard ball injury, Hockey stick injury, collision			*	Y

Dance		People knocking into <Pupil>, over stretching	*			
Gymnastics		Overstretching already hypermobile joints			*	More, if falls
Fitness	Cardiovascular		*			
	Weights	Should not lift more than 10kg weights			*	
Athletics						
	Shot Put	Lifting and stretching			*	Y
	Javelin	Stretching			*	Y
	Long Jump	Going from nothing to full pelt			*	Y
	Sprinting	Going from nothing to full pelt			*	Y
	Distance Running				*	
Ultimate Frisbee		Frisbee injury			*	Y
Dodgeball		Soft Ball – Lots of running around		*		
Cricket		Ball injury		*		

Rugby		Contact Sport, Collision, ball injury			*	Y
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Please note: The above are examples only.

[Emergency File Template Here.](#)

[School File Template Here.](#)

Please remove any irrelevant information and fill with all the information that is relevant to you. We recommend doing this in conjunction with your medical team.

MY CHECKLIST

Become a member of EDSNZ	
Join MedicAlert NZ - www.medicalert.co.nz	
Create my Care Team document	
Can I claim benefits? Benefits and payments - Work and Income https://www.workandincome.govt.nz/eligibility/	
Put together my Emergency Information File	
IF MEMBER IS A CHILD	
Inform School	
Create a health Care Plan with the school	
Put together an Emergency Information File	
Complete a PE Risk Analysis	
Consider 1:1 teaching aid	

Vascular EDS Medical Emergency?
YOU MUST CALL 111

RESOURCES

[vEDS Emergency Pack](#)

[Child's Education Emergency Pack](#)

[Nationwide and Local Support](#)

[Mental Health Support](#)

CONTRIBUTORS

[Annabelle's Challenge](#) - A big thank you to Annabelle's Challenge for providing a good majority of this document. While we, the Ehlers-Danlos Syndromes New Zealand, have changed some of it, and added in relevant links and information for New Zealand patients, the bones and a lot of the content contained herein are because of the amazing work of the volunteers at Annabelle's Challenge.

[Care Team Document](#)

[Emergency Information](#)

[School Healthcare Plan](#)

[School Emergency Plan](#)

[PE Risk Assessment](#)

[VEDS Diagnostic Criteria](#)

[VEDS Publications](#)

NIH [Vascular Ehlers-Danlos Syndrome](#)

[Ehlers-Danlos Syndromes New Zealand](#)

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