



## Annabelle's Challenge

### Who We Are

A UK registered charity providing support to patients and families touched by vascular Ehlers-Danlos Syndrome (vascular EDS) across the UK and globally. We are also proud recipients of The Queen's Award for Voluntary Service and work closely with the EDS National Diagnostic Service.

At the heart of the charity is Annabelle, the inspiration behind our patient organisation which was established in January 2013. We now have over 400 members being supported by the charity.

### Our Objectives

**Education:** To advance the education of the general public and medical profession in all areas relating to vascular EDS.

**Research:** The relief of sickness and preservation and protection of good health by the provision of funding for the development of research and early diagnosis of vascular EDS.

**Support:** To promote and protect the physical and good mental health of sufferers of vascular EDS through the provision of financial assistance, support, education and practical advice.

### VEDS Steering Group

The VEDS Steering Group will help to shape the future of the charity as it continues to grow providing support, guidance and overseeing the progress of all community-based projects including events and research in the UK.

The VEDS Steering Group is made up of volunteers who are directly touched by vEDS including patients, parents and carers, they are supported by genetic counsellors from the EDS National Diagnostic Service with representation from both clinics based in Sheffield and London.

### Steering group aims:

- Provide input based on their direct experience of vEDS.
- Give strategic direction.
- Support the charity.
- Enable more people diagnosed with vEDS to fulfil their potential by working to address issues at the earliest possible stage through the VEDS Support Programme.

The steering group usually meets at key stages during the course of a project and influences strategic decisions including reviewing and advising on vEDS research proposals.





## Meet Annabelle



Annabelle was born on 25th February 2009 weighing 6lb 5oz. We noticed as a baby Annabelle bruised very easily and without explanation, this would occur just from simply tickling her on her chest to the upsetting sight of her having finger marks and bruises on her legs when holding them whilst changing her nappy.

For any family it would be very difficult trying to keep yourself together knowing your baby has some sort of medical condition especially an unknown condition yet to be diagnosed.

The bruising on Annabelle was now a daily occurrence and most often unexplained, we also had the added pressure and upset witnessing people pre-judging us, even within the A&E department, we knew we had to keep fighting for answers!

We knew this was not normal because our two eldest sons never had issues like this which made us more determined to push our GP to have Annabelle referred for tests. The search for a diagnosis continued over three years with numerous hospital visits and appointments.

More blood tests were carried out all of which came back clear, however on one occasion the nurse was horrified that she had left her hand print on Annabelle and agreed it wasn't normal to bruise so easily and referred us to the haematology team for more tests!

It was at this stage Annabelle was diagnosed with the bleeding disorder, von Willebrand. The haematologist decided it was a good idea to refer her to a dermatologist because of previous injuries and the fact her skin was so thin.

The consultant Dermatologist suggested Annabelle could have a form of Ehlers-Danlos and referred her to genetics in Manchester and then to the EDS National Diagnostic Service at Sheffield.

On the 14th December 2012 we received a call from the genetics team to inform us the results had arrived and could we go back to Sheffield for an 'end of clinic' appointment a week before Christmas, at this point we knew it must be a positive diagnosis.

Just five days later we attended clinic and received the news that confirmed Annabelle has vascular EDS. The journey back home was surreal, but we had an answer, no more searching for a diagnosis.

Our lives had changed forever, Annabelle's Challenge had begun!





## Meet Jared



Jared is Annabelle's dad and the Founder & CEO of Annabelle's Challenge (AC) and is responsible for the day to day running of the charity and works closely with the EDS National Diagnostic Service.

He is also actively involved with the VEDS Support Programme, a volunteer NHS responder with the Royal Voluntary Service and a Mental Health First Aider accredited by MHFA England.

Jared established Annabelle's Challenge in January 2013 shortly after Annabelle's diagnosis in December 2012 and the organisation quickly started to gain momentum and in May 2014 became a UK registered charity.

*"Thank you for joining and welcome to Annabelle's Challenge, the team and I look forward to supporting you and your family on your journey with vascular EDS."*

## Our journey so far

Our journey with vascular EDS began in January 2013 and has grown to become a leading organisation for information and advice on vascular EDS, liaising with health professionals to raise awareness and increase knowledge and management of the condition.

Annabelle's Challenge has been officially recognised by Her Majesty The Queen in June 2018, winning the Queen's Award for Voluntary Service, an MBE equivalent and the highest award that can be made to a voluntary group.

In August 2021 the charity was awarded £177,415 in National Lottery funding to support its work with individuals living with vascular EDS. The charity will use the funds to deliver the VEDS Support Programme this includes helpline support, delivering talks to schools, hosting regional support groups, visiting families in their home, and facilitating ambulance markers.



## Celebrating 10 years!

## Ehlers Danlos Syndrome National Diagnostic Service

The Ehlers-Danlos Syndrome National Diagnostic Service is a highly specialised service commissioned by NHS England for individuals and families who are suspected to have rare forms of Ehlers-Danlos Syndrome (EDS) such as vascular EDS.

Established in 2009 the service runs two specialist clinics for patients at Sheffield Northern General Hospital and the Northwick Park & St Mark's Hospitals in London.

The aims of the service

- Correctly investigate and diagnose cases of suspected vascular EDS.
- Develop guidelines and pathways of care for vascular EDS.
- Provide information about vascular EDS for patients and their carers.
- Research regarding vascular EDS.
- Provide genetic counselling.
- Make relevant referrals.

### What is a genetic counsellor?

Genetic counsellors can give people information about genetic conditions and how they are also inherited, they work very closely with clinical geneticists. Clinical geneticists are qualified to diagnose genetic conditions.

The role of a genetic counsellor is to help a family to understand better the implications of a diagnosis of a genetic condition in a family. This includes helping the family to understand who else in the family might be at risk and how the condition is inherited. They may also help the family to make decisions about management of the condition.

For vascular EDS, there are tests available during pregnancy, and some people may wish to discuss these with their genetic counsellor. By exploring available options, families can make their own informed decisions about pregnancy and other matters.

### What are the possible benefits of genetic counselling?

Families who have had genetic counselling report that it can help them to:

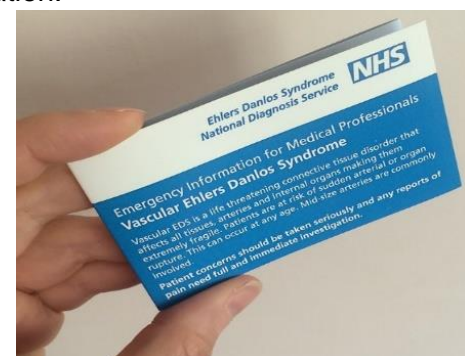
- Gain a better understanding of the condition in the family.
- Make decisions about the condition in the family.
- Talk about the condition to other people in the family who might be at risk.
- Take action to better manage the family condition.
- Communicate with health professionals, teachers, social workers etc. about their needs.

Different people may handle genetic information in different ways, depending on their own personal beliefs and their past experiences. Genetic counsellors can help individuals to make their own decisions in the context of their own unique medical, moral and social situation.

### Emergency Information 'Blue Wallet Card'

The EDS service have created an Emergency Information 'Blue Wallet Card' to be presented in an emergency at hospital. If you are a patient of the EDS service and require additional cards i.e., to have a copy at a Grandparents house or at school, please speak to one of our patient coordinators.

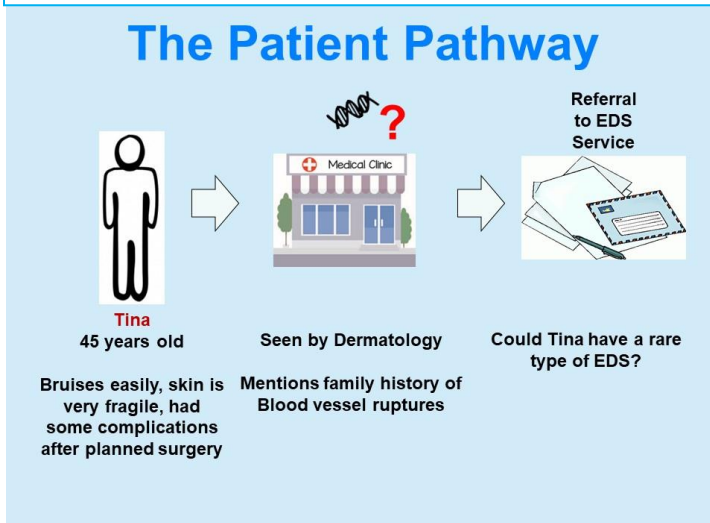
If you are not a patient of the EDS service please contact Annabelle's Challenge on 0800 917 8495 to discuss your referral options.



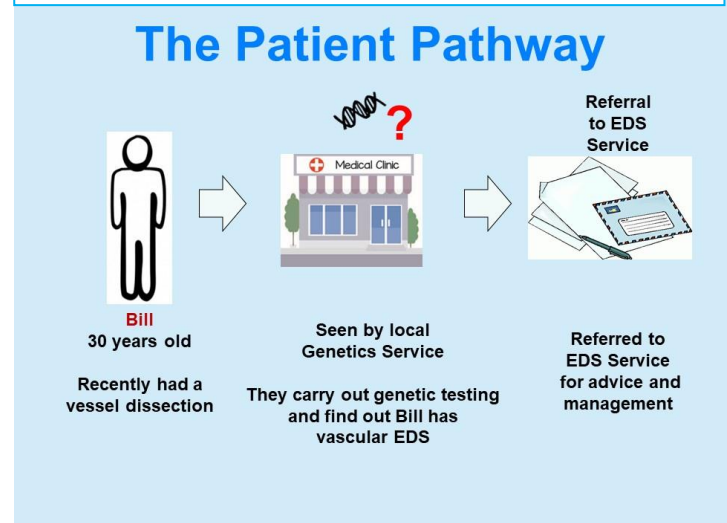
### The Patient Pathway

What happens when you or a family member are referred to the EDS service? There are two main groups of referrals that we get:

The most common is when no diagnosis has been made, but the referring doctor is suspicious the patient has vascular EDS, an example is Tina:



The second group of patients who are referred to the EDS service already knowing their diagnosis through their local genetics service, such as Bill:



There is also a third group of patients who are referred to the EDS service because they have a family member with vascular EDS and wish to know if they may also have it.

### When a referral is accepted there are four main steps:

1. Our EDS coordinators will usually be in touch to let you know the referral has been accepted, gather some personal details and create your case file.
2. The next step is usually a phone call with a genetic counsellor. This is an opportunity for us to gain an understanding into the background of the referral. We also gather information about the family and medical history.
3. There is usually a few weeks between the pre clinic call and the appointment, to allow us to gather any further information, request notes from other hospitals and discuss the case with the wider team. We then meet you in clinic, usually this is face to face, but we now offer virtual appointments. The clinic appointment usually involves a detailed examination, discussion of the condition, genetics and opportunity to ask questions. If any genetic testing is suggested, this usually happens here.
4. The last step is follow up: This can involve getting results (if not known already) and often the long term management and yearly check-ins with families with a known diagnosis. For example, attending an annual cardiology clinic.

### What happens at a Genetics Clinic appointment?

The clinic appointment will vary depending on whether we already know the diagnosis or whether we are meeting the person to hopefully make a diagnosis.

All appointments are usually 30 to 60 minutes long with a Consultant and Genetic Counsellor. It may involve one or more of the following:



- Discussing your particular concerns.
- Discussing your medical history.
- Drawing your family tree, which can give valuable information that can help make a definite diagnosis and identify who else may be at risk in the family.
- Detailed discussion.
- Information on Genetics.
- Questions and Answers.

It is therefore useful to find out as much as you can from your relatives prior to your appointment, and that you bring this information with you when you come for your appointment.

If you have been referred because you or your child is suspected of having vascular EDS, then your appointment will involve a physical examination. A consultant will examine your skin looking for scars and determine how stretchy and soft your skin is. They will also assess your joints, and how bendy they are. The consultant may wish to take some photographs of your skin and joints, with your permission. These will form part of your medical record, and if necessary, will enable the consultant to discuss your case with colleagues in the service.

The consultant will then assess how likely it is that vascular EDS may be present in your family. It may not be possible to determine this for certain at your first appointment. There are many types of EDS, and sometimes further tests are required before a diagnosis can be made. So it is possible that you may need to come to the Genetics Clinic more than once.

If and when a definite diagnosis is made, the Genetic Counsellor/ Consultant can then explain things in more detail, and try to help you understand the facts, in everyday language. This can take place by either appointment in clinic or by telephone or video.

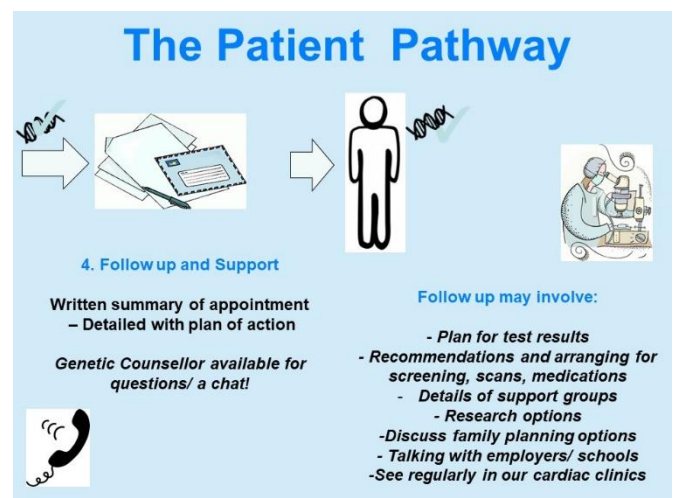
The final step of the patient pathway and usually the most important is what happens after the genetic clinic.

We always follow up with a written summary of the appointment so everyone knows the next steps. The genetic counsellor also checks in with the family to make sure the family are ok and if there is anything else we can help with. The clinics are busy and there are often questions raised after the appointment and we are here to answer them!

### Follow up and support for vascular EDS

- Yearly Check-ins (in person/ telephone/video).
- Advice on lifestyle and management.
- Cardiology appointments.
- How to talk to family/ school/ employer/ health professional about the condition.
- Planned operations/ A&E advice.\*\*
- Options for pregnancy.
- Any general questions.

**\*\* Please note the EDS service is only open weekdays**



Meet our Sheffield team



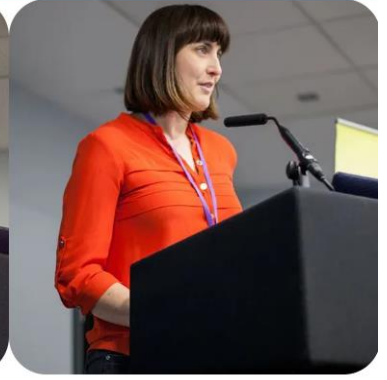
**Yvonne Bedford**  
EDS Coordinator



**Dawn Keyworth**  
EDS Secretary



**Claire Green**  
Genetic Counsellor



**Jessica Bowen**  
Genetic Counsellor



**Tammy Kammin**  
Genetic Counsellor



**Dr Glenda Sobey**  
Lead Consultant



**Dr Diana Johnson**  
Consultant



**Sheffield Clinic**

The EDS National Diagnostic Service  
Northern General Hospital  
Herries Road  
Sheffield  
S5 7AU

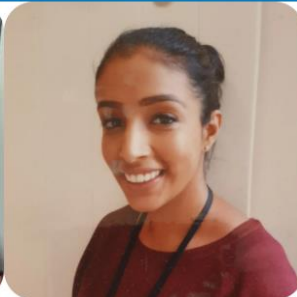
Tel: 0114 271 7764  
Email: eds.sheffield@nhs.net



Meet our London team



**Julie Leary**  
EDS Coordinator



**Ravinder Sehra**  
Genetic Counsellor



**Juliette Harris**  
Genetic Counsellor



**Dr Fleur van Dijk**  
Lead Consultant



**Dr Neeti Ghali**  
Consultant

**London Clinic**

The EDS National Diagnostic Service  
Northwick Park & St Mark's Hospitals  
Watford Road  
Harrow  
HA1 3UJ

Tel: 020 8869 3166  
Email: LNWH-tr.EDSLONDONOFFICE@nhs.net





## VEDS Support Programme

The VEDS Support Programme is available to you and your family, we offer advocacy, emotional and practical support. As a member of Annabelle's Challenge, you now have free access to all our resources and advice:

- VEDS management planning.
- Talks and support in educational settings.
- Counselling Service.
- Peer support.
- Regional support groups.
- Private closed Facebook group.
- Home visits.
- Virtual online support and 1-2-1 calls.
- VEDS Retreat Weekends, Conferences and Events.
- Patient Co-ordinator support / Freephone helpline 0800 917 8495.
- Free MedicAlert UK subscription & exclusive discounts on bracelets.
- Free Sunflower lanyard and EDS awareness card.
- Ambulance medical marker requests.
- Access to research studies and clinical trials.

### Home Visits

We provide support and advice through our freephone helpline and virtual appointments, however on a more practical level we can visit you in your own home or preferred location to talk through the management of this condition and how to prepare for the future.

### School Support

It is likely your child's school will not understand the severity of a diagnosis of vascular EDS and the care and safe management involved. We can help you and your child's school to put in place an Individual Healthcare Plan. Our team will also visit the school to talk to the teachers, staff and SENCo about vEDS and the plans that will need to be put in place to help keep your child safe.

### Peer Support

Feelings of isolation often accompany the everyday effects of living with vascular EDS. Frequently, those feelings stem from the perception that no one understands what you are going through. Sometimes you simply need to talk freely about how you're feeling.

Speak with someone who understands. Our peer support community is comprised of patients, family members and carers of all ages affected by vEDS. They are ready to listen to your cares and concerns; offer emotional support; and will share their experiences and offer their understanding.

How do I get involved? It's easy... just contact Annabelle's Challenge on 0800 917 8495.

An Annabelle's Challenge staff member will contact you and get to know you and your needs and make arrangements for a peer support volunteer to contact you by telephone, text message or e-mail, however you prefer. Remember - you are not alone. Annabelle's Challenge is here to help.







## VEDS Support Groups

### Regional Support Groups

Our support groups are an important and helpful way to share information, listen to each other's experiences and together find ways of navigating the complexities of living with or caring for someone with vascular EDS.

The meetings are for those affected by vascular EDS i.e. parents, carers, friends, children, and partners. Children are more than welcome to attend and refreshments are provided by your regional area coordinator.

It is free to attend any regional support group, simply select the region nearest to your location at the date of the planned event.

To join your local support group:

- Email - [info@annabelleschallenge.org](mailto:info@annabelleschallenge.org)
- Or complete the registration form on our website - <https://www.annabelleschallenge.org/support-groups>



### Private Facebook Group

You are welcome to join our private Facebook closed group which is for vascular EDS members only. If you would like to join the group search '**vascular ehlers danlos syndrome uk**' and connect with others in a safe and secure online space. This invitation is also extended to your family members.





## Patient Coordinator Support

### Meet Danika



Danika joined Annabelle's Challenge in April 2022 taking on the role of patient coordinator.

"Before joining the charity I was a childcare professional with 13 years' experience of working with children and their families.

Recently I completed my studies of Mental Health at college and gained a qualification in Mental Health Awareness Level 1 and Level 2 in Social Care and Children's And Young Peoples Mental Health."

Alongside her role as Patient Coordinator, Danika is also a Mental Health First Aider accredited by MHFA England.



### Meet Scarlett



Scarlett joined Annabelle's Challenge in October 2023 taking on the role of patient coordinator.

"I am so pleased to be given this amazing opportunity to take on the role as a patient coordinator for the charity to help support all our members both existing and newly diagnosed. I have previous experience in childcare, and I love to travel especially to Ireland".

Alongside her role as Patient Coordinator, Scarlett is also a Mental Health First Aider accredited by MHFA England.



To speak to a patient coordinator call us on 0800 917 8495 or email: [info@annabelleschallenge.org](mailto:info@annabelleschallenge.org)



## Get Involved

Our volunteer fundraisers are vital to our charity. Not only do they raise significant amounts of funds for Annabelle's Challenge they also raise vital awareness of who we are and what we do for the vascular EDS community across the UK and globally, helping us to reach more people who need us.

You could make a donation by monthly direct debit, a single donation via our website or just simply support our drive to help those who are affected by vascular EDS.

### How can I support Annabelle's Challenge?

You could help organise fundraising events in your local community, with family and friends or with other groups or work colleagues. You can also get involved with coordinating fundraising collections for Annabelle's Challenge.

Our volunteer fundraisers help organise:

- Coffee mornings
- Quiz nights
- Race nights
- Charity Balls
- Hair shaving challenge
- Marathons, Races & Walks
- Corporate events
- School events
- Walks and bike rides
- In-store bag packing
- Christmas jumper days at work
- Abseils
- Wine tasting & food events
- Skydiving
- Dog Walks
- Cake sales
- REDS4VEDS Day
- *And much more!*



### What skills do I need?

We are looking for enthusiastic volunteers to support Annabelle's Challenge.

### How much time do I need to volunteer?

It is up to you how much time you want to give. We will work with your fundraising event ideas with awareness leaflets, posters, promotions, and support.

To discuss ways to help the charity or to create a fundraiser please get in touch with us at the office on 0161 797 4746.





## MedicAlert

Annabelle's Challenge is an official partner of MedicAlert UK & Ireland.

### Exclusive MedicAlert UK discount codes for vEDS members:

- New member of Annabelle's Challenge
  - o Code **AC35**: Providing free of charge 1<sup>st</sup> year membership + £5 towards jewellery
- Existing member of Annabelle's Challenge **AND** anyone wanting an additional piece of jewellery
  - o Code **AC15**: Providing 15% off an additional piece of jewellery

### MedicAlert information:

- MedicAlert is useful in emergency situations when a person cannot speak for themselves (e.g. unconscious).
- MedicAlert IDs enable emergency access to details of your medical conditions, allergies, medications, implants, doctors and consultants, next-of-kin, documents and more, when it is needed most.
- Suggestions of what documents to attach on your MedicAlert record are, but not limited to: Diagnostic letter, Emergency Information for Medical Professionals sheet (*find In case of emergency section*) health management plans, pacemaker ID cards, DNR forms.

For further information and to order your MedicAlert visit: [www.medicalert.org.uk](http://www.medicalert.org.uk)



"MedicAlert keeps me  
safe every day."

- Jacob Clark



## Travel Advice & Insurance

### Flying

Air travel is not considered to increase the risks of rupture, however concern should be given to long haul flights due to the time taken to get medical assistance in case of an emergency.

### 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.

### Travel Plans

We recommend planning for your vacation, making travel plans will not only help to ensure your safety 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 & emergency information blue wallet card with you
- Wear your Medic Alert bracelet
- Purchase travel insurance

Most importantly, make memories which will last a lifetime – take your vacations and relax!

### 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 such as *Money Super Market* to get the best package at the right premium.

Consider your bank as most bank accounts offer a reward package that includes travel insurance.

### Sunflower Lanyards

Annabelle's Challenge has partnered with the Hidden Disabilities Sunflower Scheme to provide Sunflower lanyards and EDS awareness cards free of charge to all vEDS members through our online shop.

#### A discreet way to make the invisible visible.

Wearing the Sunflower discreetly indicates to people around the wearer including staff, colleagues and health professionals that they need additional support, help or a little more time.



Since the Sunflower was established in the UK in 2016, it has been adopted globally by organisations to support both their colleagues and customers.

The Sunflower is now recognised across a broad range of sectors - ranging from retail, financial services, transport including over 130 airports, travel and tourism as well as over 450 universities, schools and colleges, all of the UK railway network, theme parks, supermarkets, leisure facilities, healthcare, financial services, emergency services, theatres and over 350 charities.

The Sunflower has been launched in Australia, Belgium, Canada, Denmark, Ireland, the Netherlands, New Zealand, the UK and the USA.



The Queen's Award  
for Voluntary Service

# Vascular EDS Management





## VEDS Management Plan

### Diagnosed with vEDS, what now?

Firstly, stay positive - you are not alone! Support is available to you and your family through Annabelle's Challenge, our members, and your genetic counsellor.

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 colonoscopy (camera) please contact your consultant immediately and alert them of your diagnosis.
- Discuss informing your close family members of the diagnosis and consider which friends you are going to tell and when. Give thought to how much information you want to share and who with.
- Once your GP has received a copy of the confirmed diagnosis you may find it helpful to book an extended appointment with the GP to alert them to the condition and discuss any needs you may have.
- Once the cardiologist has received the referral letter about your diagnosis (which you will get a copy of) they will contact you separately to make an appointment for a review.
- Research your local hospital, can it perform vascular surgery? Ideally a hospital with either a trauma centre or has a cardiac/vascular team with MRI/CT and PICU/ICU would be advantageous in an emergency.
- If it is a child that has been diagnosed you need to arrange a meeting with school and include their SENDCo, an Individual Healthcare Plan (IHP) needs to be put in place.
- 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 focus on what alternative activity can be taken part in instead.
- Pregnancy presents a particular risk in female patients with vascular EDS. Discuss any planned pregnancy with the medical and obstetrics referral team prior to conception. There are no formal recommendations regarding the best method of delivery for patients of vascular EDS. A caesarean section between 35 and 37 weeks of gestation is the approach by the reference centre for rare vascular diseases. If you do become pregnant please let the EDS service and cardiology team know as soon as possible.
- There is concern that a group of antibiotics known as **fluoroquinolones or quinolones** may have a role in causing or worsening aortic aneurysms. In the UK, the drugs used in this group include: ciprofloxacin, levofloxacin, moxifloxacin, norfloxacin, ofloxacin and nalidixic acid.
- Tranexamic acid (sometimes shortened to txa) is a prescription medicine that controls bleeding. It helps your blood to clot. If you're having a tooth taken out, using tranexamic acid mouthwash can help stop bleeding. If you bleed more than usual, you may need to take tranexamic acid which should be discussed with your doctor.  
<https://www.nhs.uk/medicines/tranexamic-acid/>

We will help you with implementing your management plan, we can work on this together over time and at your pace to help reduce any potential stress or anxiety.



## My Checklist

Join Annabelle's Challenge Charity	✓
Join the private UK Facebook group	
Join my local vEDS Regional Support Group	
Join MedicAlert UK and order a bracelet using discount codes provided	
Order a Sunflower lanyard from Annabelle's Challenge <a href="#">online shop</a>	
Request an Ambulance Medical Marker/ Flag (AC)	
Create Emergency Preparedness Kit (Blue wallet card, emergency information sheet and TWIMC letter) details found in this pack	
Create a Care Team	
Referral to the EDS service, if you are not known to them (AC)	
Can I claim benefits? <a href="https://www.annabelleschallenge.org/benefits">https://www.annabelleschallenge.org/benefits</a>	
Download resources: <a href="https://www.annabelleschallenge.org/resources">https://www.annabelleschallenge.org/resources</a>	
Consider applying for Lasting Power of Attorney (LPA), health and welfare: <a href="https://www.gov.uk/power-of-attorney">https://www.gov.uk/power-of-attorney</a>	
Order additional free vEDS awareness leaflets and distribute to your local health care providers, GP etc. Available to order on the website.	
<b>IF MEMBER IS A CHILD:</b>	
Inform nursery / school / college	
Create an Individual Healthcare Plan (IHP) with SENCo team (AC)	
Book Annabelle's Challenge to visit school for a staff presentation (AC)	
Complete a PE school activity risk assessment	
Plan how/when/what to tell your child about vascular EDS (AC)	

**(AC) Contact Annabelle's Challenge on 0800 917 8495 for help & support.**

Please note Annabelle's Challenge is a patient organisation and does not provide medical advice, you should contact your local doctor or GP with any medically related concerns or questions. The information provided in this pack and on our website is obtained from various reliable sources and should not be construed to represent opinion or advice by Annabelle's Challenge and/or its staff, volunteers and trustees.

## Ambulance Medical Marker



We strongly recommend that people with vEDS have an Ambulance medical marker set up at their address.

In the event of an emergency, the presence of this marker will alert an ambulance crew that there is a person at that address with a diagnosis of vEDS. This can result in a more specific urgent treatment approach.

An ambulance marker can be placed on a home address of a person with vEDS (and other addresses such as work/school/university).

Please contact us for more information about how to arrange this by calling 0800 917 8495.

### VERY IMPORTANT EXTRA INFORMATION

- Confirmation must be received from the ambulance trust that the ambulance medical marker is in place and the person informed.
- **In an emergency you should CALL 999.** If the patient is at the address with the ambulance medical marker in place, the operator/emergency team should be alerted about the ambulance marker and the vEDS diagnosis.
- The ambulance medical marker does not replace the need for having a vEDS emergency blue wallet card (available for patients of the EDS National Diagnostic Service, please contact us if you need a replacement or extra copies) and/or medic alert bracelet ([www.medicalert.org.uk](http://www.medicalert.org.uk)) accessible at all times which should also be presented in case of an emergency - these can provide important extra information.
- Most ambulance markers are only in place for 12 months and need renewing by the GP by emailing the ambulance trust with the relevant details. The Ambulance trust must be informed in the event of an address change.

Medical Emergency? DO NOT CALL 111 - **YOU MUST CALL 999**



*Information provided by National Diagnostic Ehlers Danlos Service. Accurate to best of our knowledge on 17/11/2020.*

Annabelle's Challenge and the EDS Service cannot be held liable in the event of an emergency and the appropriate procedures not being followed and/or the response from the attending ambulance crew. Most medical markers/flags are only in place for 12 months and the doctor who organised it must be asked by the patient or their family/carer to renew it.





## 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 very 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.

Medical professionals may also wish to contact the person that made your diagnosis or the EDS National Diagnostic Service.

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.

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 their diagnostic letter, *Emergency Information Sheet for Medical Professionals* and Emergency Blue Wallet Card to be carried and provided to the Emergency Department clinicians at the time of consultation. Copies of such documents are available to UK members when joining the charity once the diagnosis has been established by genetic testing.

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 may include the creation of an informed care team, depending on your personal health and circumstances these could include:

- Vascular Surgery
- General Surgery
- Cardiology
- Gastro
- Physiotherapy
- Neurology
- Dentistry
- Haematology
- Dermatology
- Genetic Counselling

**For help with creating your care team please contact the Annabelle's Challenge helpline on 0800 917 8495 or speak to your GP.**

## **MENTAL HEALTH SUPPORT FOR PEOPLE WITH RARE FORMS OF EDS**

Receiving a diagnosis or living with a rare form of EDS can be a very 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. 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.*

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### **NHS SUPPORT**

If you would like to talk to somebody about mental health or wellbeing, in the first instance you should talk to your GP. You can also read information about accessing mental health services and support:

Mental health services for young people: [www.nhs.uk/using-the-nhs/nhs-services/mental-health-services/](http://www.nhs.uk/using-the-nhs/nhs-services/mental-health-services/)

Find a local NHS **urgent** mental health helpline in England: [www.nhs.uk/service-search/mental-health/find-an-urgent-mental-health-helpline](http://www.nhs.uk/service-search/mental-health/find-an-urgent-mental-health-helpline)

You can also **self-refer** directly to a psychological therapies service (IAPT) without seeing your GP as long as you are registered with a GP at:

[www.nhs.uk/service-search/find-a-psychological-therapies-service/](http://www.nhs.uk/service-search/find-a-psychological-therapies-service/)

These services offer therapies, such as cognitive behavioural therapy (CBT), for common problems including stress and anxiety.

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### **PRIVATE COUNSELLORS AND THERAPISTS**

British Association for Counselling and Psychotherapy approved therapists can be found at the following link: [www.bacp.co.uk/search/Therapists](http://www.bacp.co.uk/search/Therapists)

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### **GENERAL MENTAL HEALTH CHARITIES**

#### **NHS recommended charities**

[www.nhs.uk/conditions/stress-anxiety-depression/mental-health-helplines/](http://www.nhs.uk/conditions/stress-anxiety-depression/mental-health-helplines/)

**Samaritans** - offers free, confidential, 24-hour emotional support over the phone (**116 123**) or by email ([jo@samaritans.org](mailto:jo@samaritans.org)), or face-to-face at your local branch: <https://www.samaritans.org/branches>

**Mind** - the mental health charity - offers information about mental health and accessing support on their website [www.mind.org.uk](http://www.mind.org.uk) They also provide information over the phone (**0300 123 3393**), by email ([info@mind.org.uk](mailto:info@mind.org.uk)) or by text (86463). Mind has also launched a 'Find the Words' Campaign. This aims to support individuals in talking about mental health with their GP: [www.mind.org.uk/findthewords](http://www.mind.org.uk/findthewords)

**Carers UK** - offers a free helpline (**0808 808 7777**) that offers advice on financial and practical matters related to caring, as well as a listening service: [www.carersuk.org/](http://www.carersuk.org/)

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#### **Annabelle's Challenge Vascular EDS Charity**

[www.annabelleschallenge.org/](http://www.annabelleschallenge.org/)

Freephone helpline 0800 917 8495

[www.annabelleschallenge.org/get-support](http://www.annabelleschallenge.org/get-support)



## Mental Health Helplines

Don't suffer in silence – there are helplines out there that can offer support.

### Support Line - 0208 554 9004

A confidential helpline providing emotional support to individuals of any age on any issue. Support line also has a database of local services. Ring for helpline opening hours. You can also email: [info@supportline.org.uk](mailto:info@supportline.org.uk)

### Saneline - 0845 767 8000

Offers practical care and support to anybody affected by mental health problems. Open noon to 11pm Monday to Friday, noon to 6pm Saturday & Sunday.

### Nightline (for University students)

National organisation of NightLine student helplines in Universities across the UK. Visit the website to find if your Uni has one. [www.nightline.ac.uk](http://www.nightline.ac.uk)

### Shout - Text 85258

24/7 text service, free on all major mobile networks, for anyone in crisis anytime, anywhere. It's a place to go if you're struggling to cope and you need immediate help.

### Kooth

Your online mental wellbeing community. Free, safe and anonymous support. Website includes articles, discussion boards, chat with the team and daily journal. [www.kooth.com](http://www.kooth.com)

### Childline - 0800 1111

Is a free, private and confidential service that you can access online and on the phone. They can provide help and support for people up to their 19th birthday. [www.childline.org.uk](http://www.childline.org.uk)

### Winston's Wish - 08088 020 021

Provides specialist child bereavement support services across the UK, including in-depth therapeutic help in individual, group and residential settings. They support children and young people after the death of a parent or sibling. [www.winstonswish.org.uk](http://www.winstonswish.org.uk)

**Discover  
simple steps  
to look after  
your mental  
health**

**NHS**

**every mind  
matters**

**MHFA England**





**The Queen's Award  
for Voluntary Service**

# School Information



## VEDS School Support

If your child has already started school and has recently been diagnosed with vascular EDS you should arrange a meeting with the school or college SENCo as soon as possible. You will need to discuss the diagnosis and the changes that will need to be implemented.

Your local authority is responsible for identifying and meeting their special educational needs and disabilities (SEND) in a mainstream school.

Children vary in how they cope with vascular EDS, and some may need extra help to manage the condition that another child is able to handle by themselves.

We provide vEDS school support to every educational setting across the UK:

- Advice and a presentation on vascular EDS in school to all staff.
- Support for SENCo/ALNCo/Teaching Assistants/Wellbeing Officer.
- Help with implementing an Individual Healthcare Plan (IHP) or an EHC Plan.
- Help with transition to High School/College.
- Support for University students.
- Risk Assessments including PE.
- Medical emergency information.
- Ambulance medical marker placed on the school.

### SENCo

SENCo stands for special educational needs and disabilities coordinator, this is the teacher who is responsible for assessing, planning and monitoring the progress of children with special needs/SEN.

The SENCo will be the person who liaises with you, your child and the teaching staff about your child's needs within school, any adaptations needed, and implementing a healthcare plan. In Wales the SENCo is also known as an additional learning needs coordinator (ALNCo), or an additional support coordinator.

### School Talk, Advice & Support

It is highly likely your school has never come across a pupil with vascular EDS.

We are here to provide direct support and advice to schools across the UK including a visit by a representative from the charity to present to the staff and SENCo team about vascular EDS, this usually takes place within 1-2 weeks of requesting a school talk.

### School Attendance

A pupil with vascular EDS will need frequent medical appointments, their attendance record must not be penalised because of these appointments. The school's medical conditions policy should be clear on how these appointments are entered into the register.

### University Students

If you are a university student diagnosed with vascular EDS we can help. We can talk to your wellbeing officer to ensure suitable arrangements are put in place such as an ambulance medical marker, health care plan and reasonable adjustment plan.

Some students with vascular EDS suffer from fatigue or may have to attend clinic appointments which could impact on your assignments, in these exceptional circumstances you can apply for an extension to an assignment deadline through personal mitigating circumstance (PMC).



### Individual Healthcare (IHP) Plan

Individual Healthcare Plans (IHP) are documents drawn up involving people who might be required to contribute to a child's care while at school. This could include the head teacher, the class teacher, SENCo, care or support staff, other staff members who might need to provide medical or emergency care, you and your child.

An IHP plan is intended to set out what sort of support your child needs to participate in school life just like other children. It's a written document that specifies what sort of help the school can provide for your child – for example, what to do in a medical emergency.

### An IHP is essential with vascular EDS.

When developing an IHP the following should be considered:

- The medical condition: Vascular Ehlers-Danlos Syndrome, it's signs and symptoms.
- The child's resulting needs, including medication.
- The level of support needed.
- Who will provide this support and their training needs.
- Arrangements for school trips or other school activities.
- What to do in the event of an emergency, including who to contact, and contingency arrangements.

Schools in Wales use an Individual Development Plan (IDP).

### Education, Health and Care (EHC) Plan

An education, health and care (EHC) plan is for children and young people aged up to 25 who need more support than is available through special educational needs support.

EHC plans identify educational, health and social needs and set out the additional support to meet those needs. In most cases children with vascular EDS do not need an EHC plan.

### Exercise

Most of the usual aspects of daily living and recreational activities are not restricted in vEDS. The major concerns associated with high level sports activities are the risk of vascular rupture with the trauma of collisions and the change in blood pressure during these activities.

### Book a school visit

Contact our patient coordinators on 0800 917 8495 to discuss and book a school visit from Annabelle's Challenge available to any educational setting across the UK.

We will work with you and the school team to implement plans to help keep your child safe in school. Please allow 15 minutes for the presentation to take place and 30 minutes for questions and answers. We recommend the child, and their parent/s or caregiver attend the presentation, this is something we will discuss with you in advance of attending the school.





## Template for SENCo: Individual Healthcare Plan for a pupil with Vascular EDS

Name of school/setting	
Child's name	
Group/class/form	
Date of birth	
Child's address	
Medical diagnosis or condition	Vascular Ehlers-Danlos Syndrome
Date	
Review date	

### Family Contact Information

Name	
Relationship to child	
Phone no. (work)	
(home)	
(mobile)	
Name	
Relationship to child	
Phone no. (work)	
(home)	
(mobile)	

### Clinic/Hospital Contact

Name	
Phone no.	

### G.P.

Name	
Phone no.	

Who is responsible for providing support in school

SSA  
SENCO

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.

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.

Fragile blood vessels (arteries) which can lead to major complications, including ruptures, dissections or aneurysms. These can happen spontaneously and with very 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.

Implement a lift pass and allow <Pupil> to leave class and end of school day 5 minutes early

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 SSA. 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.

Medic Alert Number: <GB >, contact number for medic alert 0207 407 2818.

Ambulance marker should be applied for the school address.

#### 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 999 then call family.

Sudden intense pain – call 999 then family.

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

SSA  
SENCO

#### Plan developed with

Parent, SSA, SENCO, Head Teacher & 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

This template is available to download at  
[www.annabelleschallenge.org/resources](http://www.annabelleschallenge.org/resources)



Insert  
Photo  
Here

# Medical Emergency Plan (Template)

Name: < > Class/year: < >

Condition: Vascular Ehlers-Danlos Syndrome <https://www.annabelleschallenge.org/>

**Brief descriptions:** 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.

**Keyworker:** < > TA

**Action to be taken:** avoid crowds, impact sports, high risk injury activities.

**Temperatures-** <Pupil> does not always feel the temperature changes. If **hot**, allow to remove blazer, sit near an open window or go outside to cool down. If he/she remains warm ring home. If **cold**- needs to ensure that he/she warms up- he/she will need to warm her hands if they are becoming purple. This can be done by placing in arm pit area

**Bruises and Injuries** <Pupil> can get bruises without an impact- we need to monitor these and inform home of any we notice via keyworker or SENCo email. If there has been an impact injury inform home. For bruising monitor- <Pupil> is very good at letting us know how common the situation is for him/her. If necessary, ring home and ask for advice- this may require a video call so injuries can be shown. It may be that <Pupil> needs to be given a dose of their medication (currently not kept in school). <Pupil> bruises easily so even the slightest bump can result in swelling and bruising. **Cold compress to be immediately applied** to any knock or bump no matter how slight it may seem. If <Pupil> pulls a muscle inform home of this. <Pupil> does not feel pain like everyone else, so this also needs to be monitored.

**Breathing-** risk of hypoxia- this can then lead to more severe complications- if running ensure that <Pupil> is not out of breath and still able to hold a conversation

**Buzzing in ears-** this can be the sign of a rupture- Ring 999, say Ambulance is for <Pupil> and he/she has Vascular Ehlers-Danlos Syndrome (VEDS)- ensure that they understand severity of VEDS. Inform parents. Do not move <Pupil> if possible

**Headaches-** <Pupil> will let you know if it a normal headache- do not move <Pupil> if possible- Ring 999 if severe, say Ambulance is for <Pupil> and he/she has Vascular Ehlers Danlos Syndrome (VEDS)- ensure that they understand severity of VEDS. Inform parents. Phone home for all headaches so they are aware, face time may be required. <Pupil> may also mention blurred vision and her speech may be slurred, this is a medical emergency and requires an ambulance, the onset may be very sudden.

**Risk of collapse-** Due to their condition, collapse is possible and may happen suddenly with little or no warning. Ring 999, say Ambulance is for <Pupil> and he/she has Vascular Ehlers-Danlos Syndrome (VEDS)- ensure that they understand severity of VEDS. Inform parents. Do not move <Pupil>, but monitor.

<Pupil> wants to be as independent as possible and would like to be treated like everyone else. <Pupil> knows his/her illness and his/her limitations. <Pupil> needs to be included in as much as possible. <Pupil> should not be left alone.

This template is available to download at  
[www.annabelleschallenge.org/resources](https://www.annabelleschallenge.org/resources)

## PE & School Activity Risk Assessment Template

Activity	Specific activities	Exact activities	Risk Factors	Low Risk	Med Risk	High Risk	Could student do warm up exercises then Referee? Or participate with adjustments?
Archery			Injury to muscles and tendons in upper arm		*		
Athletics	Shot Put		Lifting and stretching			*	Y
	Javelin		Stretching			*	Y
	Long Jump		Going from nothing to full pelt			*	
	Tripple Jump		Stretching & elevating			*	
	High Jump		Jumping & Stretching - Landing			*	
	Sprinting		Going from nothing to full pelt			*	
	Distance running	*Steady pace*	Distance running puts lots of strain on the whole body.		*		
Badminton			Injuries to the lower extremities, sprains and fractures.		*		
Basketball			Ball injury, pushed or banged			*	Y
Bowls				*			
Boxing						*	
Cricket			Ball injury		*		Y
Cycling			Falling off		*		
Dance			People knocking into child, over stretching	*			
Dodgeball			Ball injury			*	
Fitness	Cardiovascular	jogging, skipping, shuttle runs, relays, ladders.		*			
		Circuit training	Depends how fast these are conducted, must avoid strenuous activities		*		
	Free Weights	Dumb bells, Kettlebells, barbell, plates, medicine ball	Should not lift more than 10kg weights, Med risk if low weights are used 1-2 kg		*		Y
	Machine Weights	Leg Press, leg extension, leg curl, vertical traction, shoulder press, chest press, lateral pull down	Should not lift more than 10kg weights. Med risk if low weights are used 1-2 kg		*		Y
	Body Weights	Sit ups, lunges, step ups, squats, wall sits, calf raises, jumping jacks, air punches	Should not lift more than 10kg weights. Med risk if low weights are used 1-2 kg		*		Y
Football		Use softball, non-competitive	Contact sport, tackling, ball injury			*	Y
Gymnastics			Overstretching already hypermobile joints		*		More, if falls
Handball			Ankle / knee sprains / collision			*	
Hockey			Hard Ball injury, Hockey Stick injury, collision			*	Y
King ball			Soft Ball - lots of people running around			*	
Kwik Cricket			Ball or bat injury		*		
Lacrosse			Contact sport, injury			*	
Netball			Ball injury, pushed or banged		*		
Orienteering			Running injuries, impact from falling on slippery surface	*			
Physio				*			
Rounders			Ball or bat injury		*		
Rowing		Leisurely only - non competitive	Vascular rupture			*	
Rugby			Contact sport, tackling, severe injury			*	
Softball		Use softball, non-competitive	Ankle sprain, ball or bat injury		*		
Swimming			People swimming into him/her, over crowded pool, back stroke, pool temp too cold	*			
Table Tennis				*			
Tag-Rugby			Sprains and strains, falling, collision		*		
Tennis		Non-competitive	Sprains and strains, ball injury		*		
Trampolining			Vascular rupture			*	
Ultimate Frisbee			Knee / Ankle / Shoulder injury or hit by the Frisbee			*	
Volleyball			Finger / Ankle sprain, bruising, ACL injury			*	Y
Yoga			Overstretching already hypermobile joints	*			
Zumba			Need regular breaks	*			

	Safe for student to participate
	Student can participate under close supervision/ adjustments in place
	Student should not participate

Y= Unless adjustments are put in place i.e warm up exercises / referee / use of soft ball / rest breaks / outside temp / small groups of students 1:1 etc

The above activities are an **example for guidance only** and a risk assessment should be carried out specific to the needs/ adjustments of the vEDS student concerned.

This template is available in Excel to download at  
[www.annabelleschallenge.org/resources](http://www.annabelleschallenge.org/resources)



# IN CASE OF EMERGENCY

Scan QR Code for Emergency Information for Medical Professionals







## Emergency Information

### Signs and symptoms to look out for:

- Severe or continuous pain in the head, neck, back or abdomen which may be diffuse or localised.
- Sudden onset of bleeding
- Unexplained swelling of a limb.
- Symptoms of a stroke (drooping of the face, arm weakness, slurred or garbled speech).
- Weakness of the limbs.
- Dizziness or loss of consciousness.
- Shortness of breath and/or difficulty breathing or speaking.
- Vision disturbances.
- Redness, pain and prominence of one or both eyes.
- The sound of pulsations in the head.
- A large bruise increasing in size.

### In the event of any of these presenting signs or symptoms:

1. **Call 999 for an ambulance / attend the Emergency Department**
2. Inform the emergency services and any medical professionals of the diagnosis of Vascular Ehlers Danlos syndrome
3. Show your '*Emergency Information for Medical Professionals*' sheet / blue wallet card
4. Explain the increased risk of arterial or organ rupture.

## Patient Self-Advocacy

It's a must for anyone who's managing a serious or rare disease such as vascular EDS and for those who are caregivers. You'll feel more prepared to self-advocate if you start learning all you can about vascular EDS, especially about how to get access to the medical care you need, and any scans that may be required such as a CT or MRI.

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, they might suggest they know about Ehlers Danlos syndrome (EDS) when in fact they probably know absolutely nothing about the vascular type.

You and/or your caregiver will need to become patient advocates. Advocacy is important **because it may reduce the chances of errors, incorrect discharge and harm to a vEDS patient**. Primarily, nurses may need to speak on behalf of you and/or your caregiver and collaborate with the healthcare team.

Being a self-advocate means learning how to be direct about what you think you may need in terms of your treatment plan. It also means listening and learning from your health care providers, all of whom are experts in their fields. From these discussions, you and your medical team can develop a plan that works for you. Being a self-advocate also means asking for help when you need it.

**Be confident, voice your opinion, be firm and be positive.**

## Emergency Preparedness

# DO NOT CALL 111 – YOU MUST CALL 999

Ideally go to a hospital with either a Trauma centre or has a cardiac/vascular team with MRI/CT and PICU/ICU would be advantageous. For all these reasons should an ambulance be needed, there would be considerable advantages in the patient being taken to the nearest Emergency Department with this expertise.

It is advisable to have the following resources to hand and show them to the ambulance crew and hospital staff:

- Emergency Information for Medical Professionals sheet and/or blue wallet card
- Your care team list of consultants
- Covering letter/ To Whom It May Concern letter
- Medic Alert ID

You should keep copies of your emergency information in your home as a 'grab & go pack', in your car and at school, university or the workplace.

Ask the nurse or doctor to contact a consultant on your care list for additional support, ideally this will be a consultant that understands your diagnosis or the EDS service.

Vascular EDS patients in severe pain **must not** be discharged from the Emergency Department even if investigations indicate that there is no major vascular event occurring because presentation of complications in vascular EDS can be subtle and unusual (Senior/ Specialist advice must be sought).



Please note the 'Emergency Information for Medical Professionals' is the same content as the blue wallet card and is available free of charge. Replacement/ additional cards can be requested through our patient coordinators by calling 0800 917 8495.

The resources provided (emergency information, wallet card, ambulance marker, Medic Alert etc) are intended to provide important information about vascular EDS to aid care in an emergency situation. However, this does not necessarily mean you will be dispatched an ambulance any quicker or treated in hospital any sooner than any other patients waiting in the emergency department.

[GP headed paper]

## TEMPLATE EXAMPLE ONLY

<Date>

### TO WHOM IT MAY CONCERN

Dear Sir/Madam,

**RE: <NAME, DOB, ADDRESS, NHS NO>**

<Name> has Vascular Ehlers Danlos Syndrome (VEDS) with a history of [*conditions and dates*].

- 
- 

He/she is currently taking [*list of medications and doses*]

- 
- 

He/she is under the care of [*names, hospitals, contact numbers*] who are happy to be contacted.

Ideally a hospital with either a Trauma centre or has a cardiac/vascular team with MRI/CT and PICU/ICU would be advantageous for this patient.

For all these reasons there would be considerable advantages in him/her being taken to the Emergency Department at < > hospital.

<Name> is an expert patient and is aware that vEDS has the potential to cause spontaneous fatal bleeding, arterial dissection or organ rupture, often with subtle presentation. He/she is also aware that any interventional procedures carry an increased risk in vEDS patients.

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.

Vascular Ehlers Danlos Syndrome is entirely different from the more benign, non-vascular forms of Ehlers Danlos Syndrome and should be treated as such. Please see attached Emergency Information for Medical Professionals document for further information.

[*any further information*]

Yours faithfully,

<GP name>

This template is available to download at  
[www.annabelschallenge.org/resources](http://www.annabelschallenge.org/resources)



## Ehlers Danlos Syndrome National Diagnostic Service

### 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 ultra sound guidance to avoid inadvertent arterial injuries.
- Fluoroquinolones/quinolones are a group of antibiotics that should be avoided if possible.

Continued...

## **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.

## **Signs and symptoms that require medical attention**

- Severe pain in the head, neck or abdomen which may be diffuse or localised.
- Sudden onset of bleeding.
- Unexplained swelling of a limb.
- Symptoms of a stroke (drooping of the face, arm weakness, slurred or unclear speech).
- Weakness of the limbs.
- Dizziness or loss of consciousness.
- Shortness of breath and/or difficulty breathing or speaking.
- Visual disturbance.
- Redness, pain and prominence of one or both eyes.
- The sound of pulsations in the head.
- A large bruise increasing in size.

**Vascular EDS is a serious condition that is associated with a risk of sudden arterial or organ rupture at any age.**

### **Sheffield Clinic**

The EDS National Diagnostic Service  
Northern General Hospital  
Herries Road  
Sheffield  
S5 7AU

Tel: 0114 271 7764

Email: [eds.sheffield@nhs.net](mailto:eds.sheffield@nhs.net)

### **London Clinic**

The EDS National Diagnostic Service  
Northwick Park & St Mark's Hospitals  
Watford Road  
Harrow  
HA1 3UJ

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## Care Team

The general approach to medical management may include the creation of an informed care team, depending on your personal health and circumstances these could include the following listed in the care team template. Once you have established a care team specific to your needs, please complete the sections of care relevant to you and add to your file.

### CARE TEAM TEMPLATE

What Services are involved?				
Name	Service	Contact Details	Telephone	Dates of involvement
	Genetic Counselling/ EDS Service			
	Paediatrician			
	Cardiology			
	Audiology			
	Dental			
	Dermatology			
	Haematology			
	Gastro			
	Physiotherapy			

Download an Excel version of this template at: [www.annabelleschallenge.org/resources](http://www.annabelleschallenge.org/resources)



## Ambulance Medical Markers frequently asked questions

**Will having a medical marker in place result in an urgent response to my call to the ambulance service?** No. However, it will help the ambulance service prioritise your needs, and may result in a more specific approach to treatment.

**Do I need to set up an ambulance marker for each individual with vEDS if they all live at the same address?** Yes. However, an ambulance marker is flagged up in the system when it is called to a specific address and so it would be important to tell the 999 call handler that there is an ambulance marker in place and the name of the person in question.

**How long does a medical marker stay on record?** This will depend on your local ambulance trust. Therefore, we recommend that you find this out when the marker is first set up. Most markers expire after 12 months and need renewing, but some are life-long.

**What do I do when my medical marker runs out?** The ambulance trust usually contacts the HCP who placed the marker e.g., GP/ EDS service to say that it is running out and whether a renewal is required. However, we would advise you keep a note of when the marker is due to expire and the details of the ambulance trust that you are under and then around the date of the expiry to check it is being/ has been renewed. Any problems with this process contact Annabelle's Challenge for ambulance marker contact details.

**Do I need to tell the 999 call handler that there is an ambulance marker in place?** Yes. Although the address should trigger the ambulance maker. We recommend that you tell the 999 call handler that there is one in place.

**If I need to call 999 for an ambulance, but am not at 'home', can I trigger the ambulance marker by referencing the 'home' address linked to the marker?** No. The trigger for the ambulance marker is the address that the ambulance is going to, rather than the individual patient or 'home' address. Therefore, if you are not at the address linked to the medical marker you will be unable to trigger it by providing your 'home' address or personal details.

**Can I set up a medical marker for more than one address?** Yes, if you have two addresses where you spend a considerable amount of time (e.g., home and work/ school/ college) and the addresses are under the same ambulance trust, then you can submit the two addresses when applying for the ambulance marker. If the addresses fall under two different ambulance trusts (i.e., they are in two different parts of the country) you will need a separate ambulance marker application for each trust.

**Can I attach medical information to the ambulance marker to help the ambulance service triage my call?** Each ambulance trust has different forms to complete when applying for an ambulance marker, and these ask for different amounts of information about the condition and/or your health which is provided by your GP during the set up process.

**Will the attending paramedic be aware of the medical marker and the information about vEDS attached to the marker?** The fact you have a medical marker in place should be known to the attending ambulance crew as the dispatch team should tell them, however this is not always the case and we recommend that you have the *Emergency Information for Medical Professionals* A4 sheet and/or wallet card, letter from EDS service / and medical history available for the paramedics when they arrive (refer to documents in this pack).