Aortic
Dissection
Awareness

The Patient Charity

Bereavement Due To Aortic Dissection

aorticdissectionawareness.org

Acknowledgements

A treasured photo of Janet Duggan with her daughters Helen and Natasha. Sadly, Janet died from aortic dissection in 1988 due to Marfan's Syndrome.

Aortic Dissection UK & Ireland would like to thank all of the bereaved family members, partners, friends, patients, health and legal professionals who contributed to this booklet.

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Bereavement Due To Aortic Dissection

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Foreword

Losing a loved one to aortic dissection is devastating. My heart goes out to you if you find yourself in that position. After losing not one but two of my dear sisters to what was initially referred to as spontaneous aortic rupture, I was looking for accurate information and for others who had heard of this condition. In 2020 I found the national patient charity Aortic Dissection Awareness UK & Ireland through their website and online "AD Buddies" support group. I had so many unanswered questions. I found myself wondering if timely diagnosis, awareness of treatment options in emergency settings or access to up-to-date information for GPs would have made any difference. I really needed somewhere to express these thoughts to people who would understand.

In the charity, I immediately found myself among of a group of survivors, families, friends and carers, all touched in some way by aortic dissection, who welcomed me with open arms. Their lived experience, empathy and support was something I found very helpful and comforting.

Having an interest in genetics, I soon got involved in the team writing the handbook "**Aortic Dissection: The Patient Guide**" and I felt empowered to volunteer alongside other AD buddies, healthcare professionals and academics to help with research into aortic disease. The opportunity to influence and input from a patient and family perspective into research has helped me come to terms with my own loss and its wider implications.

I always felt that there was a need for a publication for those bereaved by aortic dissection. With the help of Tim Deeming from Tees Law and Sharon Hassan, who lost her husband Jacob to aortic dissection, the idea of this booklet was born.

I'd like to thank the whole team of patients, family members, legal and medical advisors involved in putting this booklet together, from the early discussions about what information should be included, to writing, illustrating and proof-reading the final publication.

I hope you find this booklet helpful in your situation and as an introduction to the support that Aortic Dissection Awareness UK & Ireland can offer.

You are not alone.

Lisa Skinner

Bereavement Support Lead

Aortic Dissection Awareness UK & Ireland

Lisa Skinner

Bereavement Support Lead
Aortic Dissection Awareness UK & Ireland

Introduction

Firstly, may we offer our sincere condolences on your recent loss. This guide is written by families who have lost a loved one to aortic dissection, together with aortic patients, carers and medical professionals. It covers all the things we wish we had known at the time of our bereavement and signposts you to other things that may help you in the days to come.

You may have been given a copy of this guide following the death of a loved one from aortic disease or aortic dissection, or you may have found it online. Whether you had never heard of aortic disease or aortic dissection before and your loved ones' death was completely unexpected, or whether they had been living with an aortic condition, we hope that the information here will help you at this difficult time.

We have left some space at the end of this booklet for you to jot down any questions you have as you read through it. This will help you to prepare for discussions with medical staff and others.

> It's important to understand that, at a time like this, no question is a 'silly' one. If you feel you need clarification, don't be afraid to ask

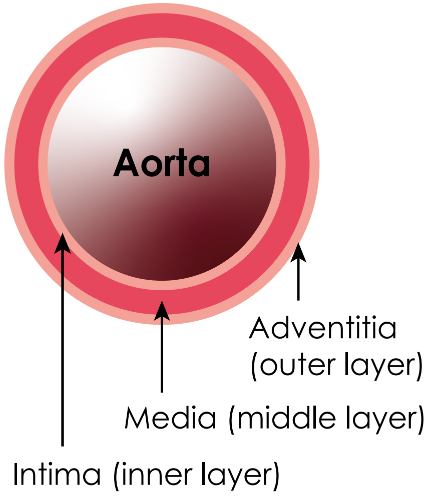
Spreading the message at a celebration of life

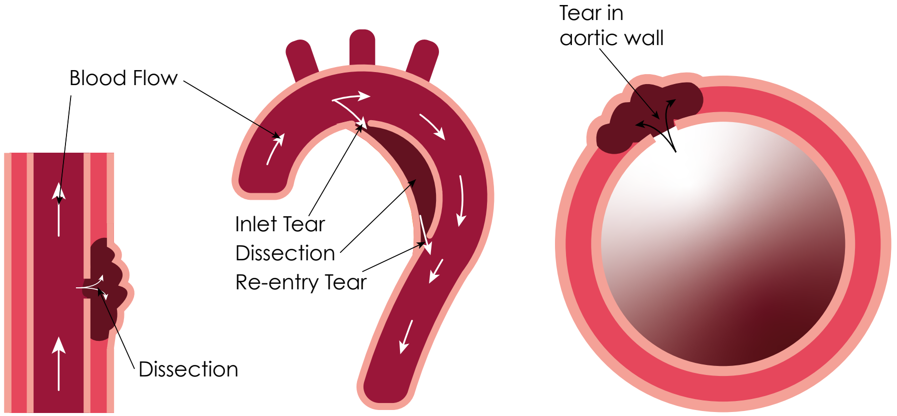
Chapter 1

About aortic dissection

What is the aorta and what is aortic dissection?

The aorta is the main artery from the heart, carrying blood to every part of the body including the heart itself, the head and brain, the arms, legs and vital organs in the chest and belly. The aorta is made up of three layers. The inner layer provides a seal and a smooth surface for blood to flow past. The middle layer is made up of elastic muscle fibres and is thicker to provide strength to withstand the pressure of the blood as it flows. The thinner outer layer provides structure and support for the aorta.





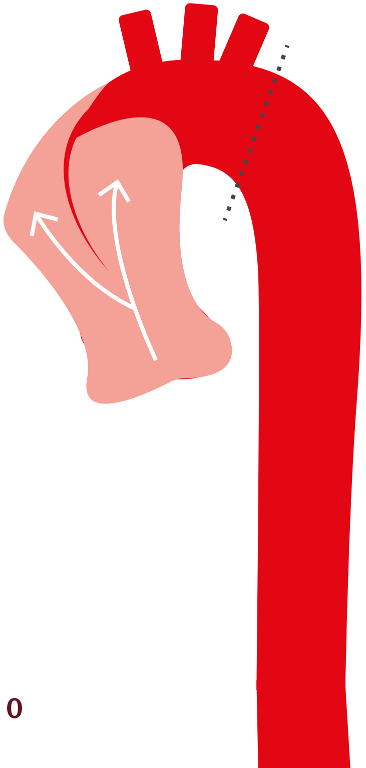
Aortic dissection

You may have been told that your loved one had an aortic dissection or an aortic rupture. An aortic dissection is where a tear occurs in the inner layer of the aorta, allowing blood to circulate in between the inner and middle layers. The dissection may disrupt blood flow to the heart, brain, limbs and vital organs in the chest and belly, which will also then be affected. Complications of this include heart attack and stroke.

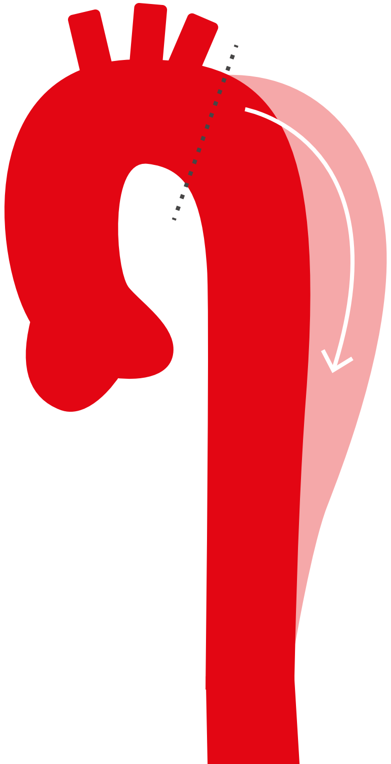
Depending on the position and progression of the tear, it can lead to swelling and stretching of the aorta and to further tears. You may also hear the terms Type A and Type B dissection. This classification is used by medical staff to describe where the initial tear in the lining of the aorta occurred. Type A refers to a tear in the first part of the aorta near where it exits the heart. Type B dissection occurs further down in the descending part of the aorta.

If all the layers within the wall of the aorta tear, blood leaks out and is no longer pumped around the body. This is known as aortic rupture.

Type A



Type B



Why did this happen?

Not all underlying causes of aortic disease, dissection or rupture are fully understood, but a common feature is that the aortic wall has become weakened in some way and is unable to withstand the pressure of the blood or the movement of the aorta created by the heart beating and the body moving.

There are several general health risk factors and genetic risk factors that increase the likelihood of suffering from aortic disease, and rupture.

General risk factors

- Aneurysm (a bulge in the aorta that could be due to a genetic condition or high blood pressure. They are more common with advancing age).
- High blood pressure (This exerts excessive force on the aorta, causing it to stretch).
- Bicuspid aortic valve - BAV (The aortic valve usually has 3 flaps, but people with BAV have only two, which can change the blood flow and have consequences for the aorta.
- Trauma (e.g., road traffic accident).
- Atherosclerosis (the build-up of fatty deposits on the inside of the aorta, often associated with smoking, diabetes, high cholesterol and high blood pressure)
- Pregnancy (which increases stress on the cardiovascular system). The charity has a free leaflet on planning a pregnancy with Aortic Disease. **See useful links.**
- Weightlifting and other strenuous sports.
- The use of illicit drugs such as cocaine.

Genetic Risk Factors

As well as the risk factors mentioned above, aortic disease can be caused by small changes in our genes. These small gene changes are known as mutations or variants and can be passed on, so that aortic disease can run in families. You may hear this described as “familial, heritable or inherited aortopathy”.

Genetic variations affecting the aorta can be classed as either syndromic or non-syndromic.

Syndromic

A syndrome affects multiple body systems, not just the aorta. Someone with syndromic aortic disease will often (but not always) have several typical features. These might be tall stature, long limbs, curvature of the spine, a particular facial appearance, or tortuous blood vessels (visible on a CT or MRI scan) for example. The most common syndromes associated with aortic disease are Marfans, Loeys Dietz and Vascular Ehlers Danlos Syndromes. Another is Turner’s Syndrome, which only affects women and is associated with bicuspid aortic valve and a shorter stature. These syndromes typically weaken the connective tissues of the body and are sometimes referred to as connective tissue disorders (CTDs).

Weakening of the aortic wall tissue due to a syndrome means that a person is at increased risk of aortic dissection. We have included some websites in the **useful links section** for further information on these syndromes. When someone has one of these syndromes, the faulty gene can usually be identified by genetic testing.

Non-syndromic

In non-syndromic inherited aortic disease, the condition runs in the family, but there are none of the physical features seen in the syndromes described above. Though these cases have a genetic cause, presently the genetic fault cannot always be found through genetic testing. However, the number of genetic causes that can be tested for is increasing all the time, as more is understood about them.

In both syndromic and non-syndromic types of inherited aortic disease, not all members of the family will be affected, and some may be affected more or less severely than others. A parent with a faulty gene causing aortic disease usually has a 50:50 chance of passing it on to each child. Occasionally, someone with aortic disease can be the first person in a family to have a particular genetic mutation, which might be the start of a family trait.

> “5-20% of aortic dissections are associated with connective tissue disorders.”

The Hill family lost their beloved Linda (left) to Aortic Dissection

Chapter 2

What to do after a loved one has died

What do I need to do after someone dies?

When someone dies it is an extremely emotional time for family and friends. Despite this, there are several statutory and other formalities that must be addressed. It is a time when these actions are not at the forefront of peoples' minds but must be carried out.

The important actions include registering the death, notifying national and local government departments, banks, building societies, insurance companies, employers or former employers, pension providers, etc.

Some of these organisations, such as the regional deaths registration service, banks, hospitals and funeral directors, provide booklets and leaflets on the subject containing a useful checklist.

Registering a death

When someone passes away, their death must be registered.

In England, Wales, and Northern Ireland this should normally be done within 5 days at a local Registrar Office and in Scotland, within 8 days at the Registrar's office in the region where the death occurred.

When registering a death, certain documents need to be produced, such the Medical Certificate of Cause of Death (MCCD) from the deceased's doctor, their marriage and birth certificates and proof of address. The MCCD is usually sent electronically by the doctor who certified the death to the Registrar's Office.

In Ireland you should register the death within 3 months of the date the person died but have up to 12 months to register the death.

Also, in Ireland, the Death Notification Form is issued by a doctor. In those cases where an inquest or postmortem is required, an Interim Certificate of the Fact of Death is obtainable from the Coroner Service to enable the formal arrangements to be made.

In some instances, the death will be referred to the Coroner or the Procurator Fiscal in Scotland, **hereafter both are referred to as "the Coroner"**, for investigation, determination and a report on the cause of death. In these cases, the Medical Certificate of Cause of Death (MCCD) will be issued to the Registrar by the Coroner.

The Registrar's Office charges a fee for the registration and will provide a copy of the Death Certificate. You may need additional copies to send as evidence to other organisations such as banks; these may be obtained for a fee.

In Ireland, there is no charge for registering a death, but there may a be charge for a copy of the death certificate.

Notifying organisations

When notifying organisations such as Department of Work and Pensions (DWP), Department of Social Protection (in Ireland), banks, insurance companies, pension providers, etc., they may wish to see the Death Certificate. This may be a scanned copy, but in some instances, an original copy might be requested.

On some occasions, you may be asked to provide an original or certified copy of a grant of probate, Letters of Administration or other documents.

In some local authority areas, the Registrar will provide access to the "Tell Us Once" service on the government website. This allows you to enter information about the deceased person and this will inform all appropriate national and local governmental departments, thus reducing the need to write letters and make telephone calls.

Other organisations, such as banks and building societies, might have signed up to the "Death Notification Service" which notifies its members of the death.

Probate

Probate is a legal process that concerns disposal of the deceased's assets (money, property, etc). Guidance on probate can be obtained from the government website.

There may be a need to apply for a Grant of Probate over the deceased's estate. In some cases, such as where the assets of the deceased person are held jointly with another person (such as their husband or wife), they will pass automatically to the joint owner, so there will be no need to apply for probate.

All the probate forms, including on-line application, can be found on the government website or the Probate and Inheritance Tax Helpdesk Tel. 0300 123 1072

In Scotland, the application, or "Confirmation" is obtained via the Sheriff's Court.

For Ireland, a Grant of Representation is required.

If the estate is complex or you are not confident in attending to the application yourself, solicitors and specialist legal services can help with probate applications; fees are charged for this work.

Details of the relevant websites can be found in the **useful links section** at the end of this booklet.

I have been told there will be an inquest. Why is this needed and what does it involve?

An inquest is an investigation by a Coroner into how the deceased came about their death. A doctor or the police will have referred the death to the Coroner.

In Scotland, a legal enquiry into the cause and circumstances of a death is known as a Fatal Accident Inquiry. It is requested by the Procurator Fiscal rather than a Coroner.

The Coroner (or Procurator Fiscal), **hereafter both are referred to as "the Coroner"**, has a duty to investigate a death if they have reason to suspect that one of the following circumstances applies:

a. The deceased died a violent or unnatural death
b. The cause of death is unknown
c. The deceased died while in custody (e.g. in prison) or in state detention (e.g. in a mental health hospital run by the NHS)

If the Coroner is not sure whether the duty to investigate the death arises, they can make preliminary enquiries to satisfy themselves whether their duty to investigate has been triggered.

Retired GP Jacob Hassan sadly lost his life to a misdiagnosed aortic dissection

For example, the Coroner can ask for a post-mortem examination and/or can obtain documents that they believe will help them identify whether the duty to investigate the death arises.

A post-mortem report may give the Coroner new information to help decide whether a fuller investigation is required.

Following their preliminary enquiries, the Coroner may decide that they are satisfied that the duty to investigate does arise and an inquest hearing is required.

If the post-mortem shows that the deceased died as a result of natural causes and there is no other information to suggest that the death was violent or unnatural, then the Coroner must discontinue the investigation and inquest.

Alternatively, the Coroner may also call a pre-inquest review hearing (PIRH), although they are not obliged to.

The Coroner (or Procurator Fiscal) is not allowed or expected to make any finding relating to liability or negligence. The allocation of blame is for the civil courts not the coroner's court.

Should I have a lawyer representing me?

You are not required to have to have a lawyer representing you at an inquest or pre inquest hearing as the coroner will explain what is happening and ensure there are opportunities for you to ask questions. However, at some inquest hearings other interested parties (such as public bodies) may be represented by a lawyer and you may feel you need legal help to prepare for the inquest due to the circumstances around your loved ones' death.

If you feel you need legal support for an inquest or subsequent representation for a civil claim it is important to seek advice as early as possible. It is important to use a good clinical negligence lawyer of sufficient ranking and who has experience of an aortic dissection case. There are also time limits applied to raising a negligence claim.

Aortic Dissection Awareness UK & Ireland has a network of legal experts with experience of aortic dissection cases who are willing to have an initial no-obligation chat with bereaved relatives considering seeking legal advice, so that you understand your options. Please contact us if you would like to know more.

Further guidance on choosing a lawyer is also available from The Law Society and Action against Medical Accidents (AvMA) **see useful links** at the end of this document.

See also chapter 4 Raising Your Concerns and Complaints.

More detailed information about Coroners, The Procurator Fiscal and inquests or their equivalent in each nation can be found in the **useful links section** at the end of this document.

What is a Post-mortem and why is it needed?

A post-mortem (also called an autopsy) is an examination of the body after death by a qualified pathologist to assist in determining the cause of death.

The Coroner or Procurator Fiscal, **hereafter both are referred to as "the Coroner"**, will appoint a pathologist to conduct a post-mortem. The pathologist should be independent, therefore where a death has occurred in hospital, the pathologist should be from another hospital or area.

Most post-mortems require detailed physical examination. Whilst some post-mortems can be completed non-invasively such as by MRI or CT scanning, there are some difficulties with this method:

a. Scanning techniques for post-mortem are not available in every area of the country.

b. Scanning techniques are not always appropriate, for example, if a toxicology and/or histology report is required.

c. Where scanning techniques are used, the family of the deceased or next of kin will be expected to pay a fee.

d. Scanning techniques may not avoid the need for a more invasive post-mortem. Following receipt of the scan results, the Coroner may decide that the more invasive type of post-mortem is required after all.

Ultimately, the Coroner will decide whether a post-mortem, by way of scanning, is appropriate.

The Coroner has authority to remove from the body, and preserve, material that has a bearing upon the cause of death. This could include organs or tissue. The Coroner cannot retain these after the inquest has concluded without the consent of the relatives.

What options do families have as to whether to agree to a post-mortem or not (and a second post-mortem)

The Coroner has complete discretion about whether a post-mortem should be performed. You cannot object to a Coroner's post-mortem. That said, the decision should be made with sensitivity, considering the relatives' views and any religious and cultural sensitivity, as far as possible.

The Coroner must give notice of the date, time, and place of the post-mortem to the next of kin, personal representative and any other interested person.

What happens after the post-mortem?

After the pathologist has made their assessment, the Coroner will usually authorise the release of the body and then funeral arrangements can be made. The pathologist will then provide a post-mortem report to the Coroner. The report should contain a detailed analysis of the deceased's body and then provide conclusions about the cause of death. The post-mortem can include other medical factors which contributed to the person's death.

Once the post-mortem process has concluded, the Coroner is required to release the body for cremation or burial as soon as possible. The Coroner can only authorise burial or cremation once they are satisfied the body is no longer needed for the purposes of their investigation.

Second post-mortem examinations

A second post-mortem examination is an examination by a different pathologist, who considers the original report and carries out work to agree or challenge its conclusions. They are most commonly conducted by forensic pathologists.

The vast majority of second post-mortem examinations are requested in cases of potential homicide, mostly by a defendant's solicitors. However, sometimes bereaved families may want a second post-mortem examination when they are concerned to verify the cause of death.

There is no absolute right or entitlement for a suspect, defendant, the police, or any other interested person to have a second post-mortem examination. Whilst the body remains under the Coroner's legal control, the decision whether to arrange a second post-mortem examination (and in what form) remains a judicial decision for the Coroner to make, taking account of the reasons in support of a request and any competing considerations.

Further information about post-mortems can be found in the **useful links section.**

What are Medical Examiners?

Medical examiners are part of a national network of specially trained independent senior doctors from any specialty. Overseen by the National Medical Examiner, they scrutinise all deaths that do not fall under the coroner's jurisdiction. Medical examiners are a core part of the process of investigating patient deaths across the NHS in England, Wales and Northern Ireland. Further information about the national medical examiner system can be found in the **useful links section.**


bereavement
Questions
??
ANGER
future
uncertainty
help
SUPPORT
EXPLANATION
understanding
SADNESS
Connection
PURPOSE
FEAR
Closure
SHOCK
help
LOSS
GRIEF
Care
COPING

Aortic Dissection Buddies meet-up

Chapter 3

Bereavement Support

What support is available to help me after losing someone to aortic dissection?

Losing a loved one suddenly can be life altering and catastrophic to those left behind. A sudden and unexpected death presents unique challenges and may not allow a chance to prepare for the loss or to say goodbye. It can be difficult to predict how you may react to a loss, but you may experience numbness, denial, shock, anger or sadness and become overwhelmed. In addition, a sudden death with a suspected genetic cause can complicate the grief process further and present additional challenges for those left behind. We recognise that grief is a very personal thing. This booklet has been created to offer support and help you navigate your personal, unique and complex journey of grief after losing a loved one suddenly to Aortic Dissection.

The experience of losing a loved one is often referred to as bereavement, grief and mourning. Grief can affect us all in different ways. It can affect our emotional and physical health and wellbeing. We all have unique responses to loss, and this can be influenced by many additional factors, including the sudden nature of a death.

Elisabeth Kübler-Ross, a Swiss-American psychiatrist and pioneer of near-death studies, first suggested that there are five stages of grief: denial, anger, bargaining, depression and acceptance (known as the “Kübler-Ross Model”) in her book “On Death and Dying”, published in 1969. This is not necessarily a sequence that your grief will follow. You may experience different stages of grief at different times and in no particular order. The five stages of grief are however useful to understand the different reactions that you may experience after your loss.

For those left behind, life can change forever, and it is important to understand that each individual journey of grief is unique, with no right or wrong way to feel, and no set time limits to grieve.

For further reading on how bereavement impacts on people, psychotherapist Sasha Bates wrote an excellent book, "Languages of Loss: A psychotherapist's journey through grief", which was published in 2020, about her journey with grief after losing her husband, the actor, director and playwright Bill Cashmore, at the age of 56 to Aortic Dissection.

There are several charities and other organisations, including not-for-profit organisations, who offer help and support for the recently bereaved. This is a list of the ones our buddy group members have found particularly helpful.

- Citizen's advice
- Samaritans
- Cruse Bereavement Care
- Cruse (for young people)
- Winston's Wish
- National Bereavement Service
- Child Bereavement UK
- The Silver Line
- Dying Matters
- Widowed and Young
- The Good Grief Trust
- Compassionate friends (loss of a child, whatever age)

We have included their website details in our **useful links section.**

> "As Aortic Nurse specialists, we feel the loss of any of our patients hugely. The journey we share and the support we provide is something we are very proud of. We want to continue and extend this support as much as we can to the family and friends."
>
> ***Roxanne Noces***

Chapter 4

What to do if you have any concerns or complaints

After experiencing the death of someone close, you may want to understand more about the reasons they died, or you might have some comments, questions or concerns about the care or treatment they received. You should ask to speak to someone in the team who looked after your loved one in the first instance. If you would prefer to speak to someone else who was not directly involved in their care, you can ask the relevant independent patient advocacy organisation, such as PALs, at the hospital if they can help.

Who are PALS?

In England and Wales (see also below) The Patient Advice Liaison Service (PALS) has been part of every NHS trust since 2002. PALS are a point of contact for patients, their families and their carers to raise concerns. PALS can give advice about the formal complaint process, but its goal is to resolve patient concerns quickly and informally where possible.

Where there are concerns, you may ask PALS to investigate these to try to help you resolve issues informally. They can also provide support and advice about making a formal complaint and how to access further advocacy services, if required.

Llais

In April 2023, the Welsh government established an independent statutory body called Llais Wales to give the people of Wales much more say in the planning and delivery of their health and social care services. Llais can also provide free independent and confidential support with raising complaints and help explain your options in terms of the outcome.

PASS

In Scotland, a similar role is carried out by The Patient Advice and Support Service. They can support anyone who uses the NHS in Scotland to understand their rights and responsibilities, raise concerns, give feedback or comments, or make a complaint about NHS treatment.

PCC

In Northern Ireland, The Patient and Client Council will support you if you have a concern about any part of Health and Social Care in Northern Ireland.

Patient Advocacy Service

In Ireland, the Patient Advocacy Service can provide information and support to assist with formal complaints about care in a Public Acute Hospital. They can also support people in the aftermath of a Patient Safety Incident. Following a response to any formal complaint, they can support you to explore your options. PAS is an independent, free and confidential service.

How should organisations respond to Patient Safety Incidents?

NHS England developed The Patient Safety Incident Response Framework (PSIRF) to respond to patient safety incidents and improve patient safety. This replaces the previous Serious Incident Framework (2015) and is mandatory for acute, ambulance, mental health, and community healthcare providers (all of these services are provided under NHS standard contracts). The new framework places emphasis on understanding the factors contributing to incidents and learning from them. One of the primary aims is to involve those affected by patient safety incidents and apply various approaches to learning from incidents. Responses need to be tailored and proportionate to the severity of the incidents.

Similarly in NHS Wales, patient safety incidents are managed through the Patient Safety Incidents Policy which builds on existing processes from the Putting Things Right (PTR) framework. This applies to local health boards, NHS trusts, independent providers, and primary care practitioners.

The Scottish Patient Safety Programme (SPSP) is the current approach used to address patient safety incidents in NHS Scotland. SPSP was established in response to evidence that nearly 1 in 10 patients admitted to hospitals experienced unintentional harm, with many incidents preventable. This builds on Scotland's Safer Patients Initiative and extends its work to all NHS Boards in Scotland. Healthcare professionals can report adverse incidents and near-misses involving medical devices, diagnostic equipment, facilities, and more through the National Services Scotland platform.

Ireland and Northern Ireland currently manage patient safety incidents through a variety of approaches. The complexity of current arrangements is outside the scope of this document and the advice subject to change.

In Ireland, the Patient Safety (Notifiable Incidents and Open Disclosure) Act 2023 is currently in the process of being implemented and will provide a framework for mandatory open disclosure of specified patient safety incidents. The emphasis will be on transparency and communication with patients. Healthcare professionals can report incidents to allow learning and to prevent a recurrence. This reporting is an opportunity to review practices and share insights and is not about allocating blame.

Northern Ireland Adverse Incident Centre (NIAIC) investigates reported adverse incidents involving medical devices, non-medical equipment, and buildings within the healthcare environment. It issues guidance to prevent recurrence and protect patients, staff, and users. Community pharmacists can also anonymously report incidents, including near misses, via an online system. The focus is on learning and sharing good practices without concerns about identifying the reporting pharmacist.

For further information on the relevant websites and documents discussed please **see useful links section.**

Derek Fogarty and his daughters. Sadly Kirsty (far left) and Angela (far right) both died of Aortic Rupture.

Chapter 5

Screening for risk of aortic dissection - are other family members at risk?

Are other family members at risk?

If your loved one died of an Aortic Dissection aged 60 or younger, it is generally recommended that close blood relatives should be screened in case there is a genetic risk of aortic disease. The hospital team who cared for your loved one, or your GP, can refer you to an NHS Regional Genetics service or Inherited Cardiac Conditions (ICC) service to take a family history and, if necessary, arrange for screening.

For information, we have included an example of a letter that the hospital could give to you or other family members who might be at risk of aortic dissection. This is to enable family members to have a discussion with their own GP to arrange screening. See appendix 1.

If screening for you or other blood relatives is not mentioned, you can ask a member of the medical team or your GP to investigate this.
For more information, see our THINK AORTA, THINK FAMILY campaign website: www.thinkaorta.net/family

> “My sister died suddenly from aortic rupture. She was here one minute and gone the next. A specialist told me there was no point worrying about whether it could be hereditary as nothing could be done to prevent it. After my second sister died from the same condition, I discovered that there is preventive surgery and medical intervention that might have reduced the chances of them dying. If only our family had been offered imaging and surveillance.”
>
> ***Lisa Skinner***

What does screening involve?

Screening will involve imaging (such as Echocardiogram, MRI or CT scans) to look closely at the aorta and genetic testing for family members.

The combination of imaging results and genetic testing can help doctors work out someone's risk of an aortic event and whether early medical or surgical intervention will help manage that risk and potentially prevent Aortic Dissection.

Imaging

CT or MRI scans are the best way of accurately viewing the whole aorta to assess its condition and take measurements. An echocardiogram can view the first part of the aorta, where it exits the heart, which can be a useful initial scan.

Genetic Testing

Genetic testing can lead to significant discoveries for the family's health, so it needs careful consideration and thought. Genetic counsellors are trained and experienced in helping people to understand the pros and cons of testing. They can also help families navigate the psychological and practical decisions around testing. Ultimately the decision is down to each individual and the genetics specialist will respect your decision on the matter. The genetics team will also ensure permission is obtained for any personal information that might be shared between medical experts and family members as a result of testing.

If a geneticist recommends testing, and you agree to go ahead with this, you will be asked to sign a consent form. A sample of blood will be taken and sent away for the DNA to be analysed. Results take up to six months to be returned and you should receive an appointment with your genetics team to discuss the results.

NHS criteria for genetic testing for aortic conditions in England and the equivalent in other nations can be found in the **useful links section.**

Genetic tests can lead to three types of results: -

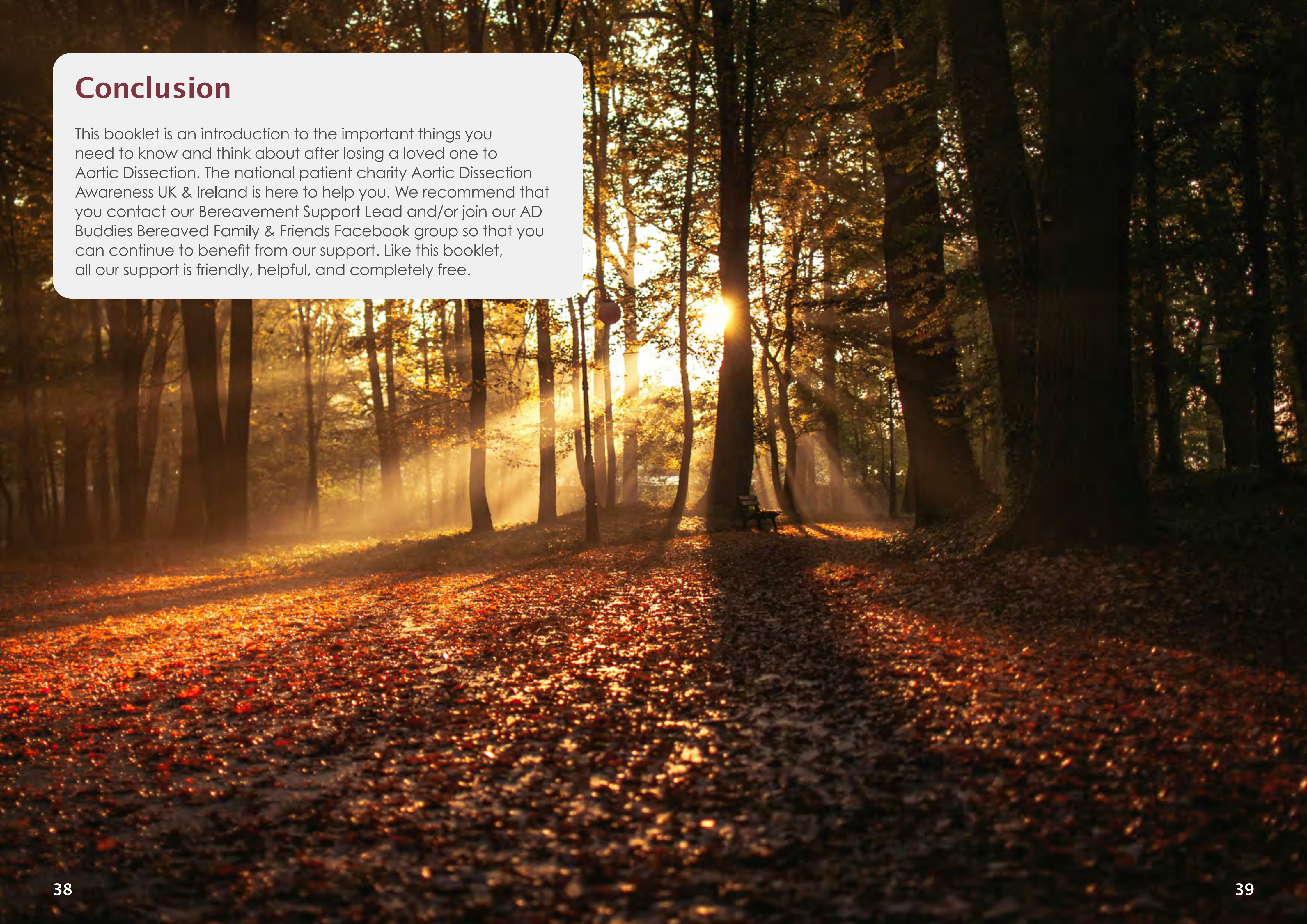
1. A positive test result where a genetic change known to be associated with an aortic issue is found in your DNA. Other members of your family may be invited for screening and testing – known as cascade testing.
2. A negative result where no significant change is found. This means that your relative's aortic condition was not caused by a genetic change or that it was not caused by a gene change known at this time. As new genetic variations are constantly being discovered, you may be advised to have further testing in the future.
3. A variant of unknown significance (VUS), where a genetic change is found in your DNA, but its significance is not clear. It might be one of many harmless genetic variations that make each one of us unique, or it might be the cause of your family's aortic condition. Additional testing may be required, and other family members may also be invited to have genetic testing to look for this variant in their DNA.

It is important that any test result is explained properly and that you have the opportunity to ask questions, in order to understand the meaning of the findings.

What if screening shows that I or a family member are at risk?

The combination of Imaging and genetic testing provides information to help doctors work out if you, or your family members, are at risk of an aortic dissection or rupture and whether early medical or surgical intervention could help manage this risk. This might mean you have to take medication to protect the aorta or stop it deteriorating as quickly, as well as having regular surveillance scans. Sometimes, doctors may recommend surgery to replace part of the aorta before a dissection occurs.

> “Timely diagnosis and surgical intervention can save lives.”



Conclusion

This booklet is an introduction to the important things you need to know and think about after losing a loved one to Aortic Dissection. The national patient charity Aortic Dissection Awareness UK & Ireland is here to help you. We recommend that you contact our Bereavement Support Lead and/or join our AD Buddies Bereaved Family & Friends Facebook group so that you can continue to benefit from our support. Like this booklet, all our support is friendly, helpful, and completely free.

Glossary

- **Aorta.** The aorta is the main artery from the heart, carrying blood to every part of the body including the heart itself, the head and brain, the arms, legs and vital organs in the chest and belly. The aorta is made up of three layers. The inner layer provides a seal and a smooth surface for blood to flow past. The middle layer is made up of elastic muscle fibres and is thicker to provide strength to withstand the pressure of the blood as it flows. The thinner outer layer provides structure and support for the aorta.
- **Aortopathy.** Aortopathy means any disease of the aorta, which is the main artery of the body.
- **Artery.** A vessel through which the blood passes away from the heart to various parts of the body. The wall of an artery consists typically of an outer coat (tunica adventitia), a middle coat (tunica media), and an inner coat (tunica intima).
- **Connective tissue.** Your connective tissue supports many different parts of your body, such as your skin, eyes, and heart. It is like a "cellular glue" that gives your body parts their shape and helps keep them strong. It also helps some of your tissues do their work. It is made of many kinds of proteins. Cartilage and fat are types of connective tissue.
- **Connective tissue disease.** A connective tissue disease is any disease that affects the parts of the body that connect the structures of the body together.
- **Coroner.** Coroners are independent judicial officers, appointed by the local authority, and are either doctors or lawyers responsible for investigating the cause of deaths.
- **CT scan.** A Computerised Tomography (CT) scan is a test that uses X-rays to take detailed pictures of the inside of your body. It's usually used to diagnose conditions or check how well treatment is working.
- **DNA.** Deoxyribonucleic acid (DNA) is a molecule that contains the biological instructions that make each species unique. DNA, along with the instructions it contains, is passed from adult organisms to their offspring during reproduction.
- **Familial Aortopathy.** A disease of the aorta that tends to occur more often in family members than is expected by chance alone.
- **Heritable or inherited Aortopathy.** A disease of the aorta that tends to be passed from parent to offspring, i.e. it has a specific genetic component.
- **MRI scan.** Magnetic Resonance Imaging (MRI) is a type of scan that uses strong magnetic fields and radio waves to produce detailed images of the inside of the body.
- **Pathologist.** Is a medical expert, often a doctor, in the study of diseases, and especially someone who carries out a detailed examination of a dead person's body to find out why someone has died.
- **Procurator Fiscal.** A legally qualified public prosecutor within the Crown Office and Procurator Fiscal Service.

  All suspicious, sudden, and accidental deaths must be reported to the Procurator Fiscal, and they have a responsibility to identify if any criminal action has occurred and, where appropriate, prosecute.
- **Tortuous Blood Vessels.** People with Tortuous blood vessels have arteries and veins that show significant twisting, bending, and turning on MRI or CT images.

  Tortuosity arises from abnormal elongation of the arteries; since the end points of the arteries are fixed, the extra length twists and curves. Other blood vessel abnormalities that may occur in this disorder include constriction (stenosis) and abnormal bulging (aneurysm) of vessels, as well as small clusters of enlarged blood vessels just under the skin (telangiectasia).

  It can be associated with inherited connective tissue disorders. (See section on “Syndromic” and “Non-syndromic” genetic risk factors.

Useful Links

Disclaimer:

The links listed on this page were correct at the time of publication. The ones on our website may be more up-to-date. The easiest way to access these links is to go to the webpage

tinyurl.com/ADBereavement

and click the link that you want from there.

Pg No.	Subject	URL
	Aortic Dissection Awareness UK & Ireland	https://aorticdissectionawareness.org/
	THINK AORTA	https://www.thinkaorta.net/
	Aortic Dissection: The Patient Guide	https://aorticdissectionawareness.org/patients-and-families/the-patient-guide https://aorticdissectionawareness.org/resources/aortic-dissection-the-patient-guide
11	Leaflet: "Planning Pregnancy with Aortic Disease"	https://aorticdissectionawareness.org/resources/planning-pregnancy-with-aortic-disease
12	Genetic Risk Factors – Syndromes For further information see NHS A-Z index	https://www.nhs.uk/conditions/
	Or Health Direct. gov.au	https://www.healthdirect.gov.au/genetic-disorders
16	Notifying organisations	www.gov.uk/tell-us-once
16	Notifying organisations	www.deathnotificationservice.co.uk
17	Probate - UK	www.gov.uk/applying-for-probate
17	Probate – Bereavement Guide - Ireland	https://www.citizensinformationboard.ie/downloads/guides/Bereavement_Guide_2023.pdf
20	Find a solicitor	https://solicitors.lawsociety.org.uk/
20	Action against Medical Accidents – a charity registered in England & Wales and in Scotland	https://www.avma.org.uk/
20	Guide to Coroner Services for Bereaved People – England and Wales	https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/859076/guide-to-coroner-services-bereaved-people-jan-2020.pdf
20	Coroner Service publications - Ireland	https://www.gov.ie/en/campaigns/coroner-service/
20	Coroners Service for Northern Ireland	https://www.justice-ni.gov.uk/articles/coroners-service-northern-ireland
20	Procurator Fiscal role in investigating deaths in Scotland	https://www.copfs.gov.uk/about-copfs/our-role-in-investigating-deaths/
20	Procurator Fiscal Guide for Bereaved family members in Scotland	https://www.copfs.gov.uk/services/bereavement-support/guide-for-bereaved-family-members/
23	Information about post-mortems - UK	https://www.nhs.uk/conditions/post-mortem/
23	Information about post-mortems - Ireland	https://www.citizensinformation.ie/en/death/sudden-or-unexplained-death/postmortems/
23	National Medical Examiner system – England and Wales	https://www.england.nhs.uk/patient-safety/patient-safety-insight/national-medical-examiner-system/
29	Charities and other organisations, including not-for-profit organisations, that offer help and support for the recently bereaved	A sample list of organisations supporting the recently bereaved:
		Citizen's advice – www.citizensadvice.org.uk
		Samaritans – www.samaritans.org
		Cruse Bereavement Care – https://www.cruse.org.uk/
		or www.crusescotland.org.uk
		Cruse (for young people) – www.hopeagain.org.uk
		Winston's Wish - www.winstonswish.org
		National Bereavement Service – www.thenbs.org
		Child Bereavement UK – www.childbereavementuk.org
		The Silver Line – www.thesilverline.org.uk
		Dying Matters – www.dyingmatters.org

	Charities and other organisations, including not-for-profit organisations, that offer help and support for the recently bereaved	Widowed and Young - https://www.widowedandyoung.org.uk/
		The Good Grief Trust - https://www.thegoodgrieftrust.org/
		Compassionate friends (loss of a child, whatever age) – www.tcf.org.uk
		Bereavement Support and Services in Ireland – Health Service Executive - https://www2.hse.ie/mental-health/services-support/bereavement/
		Irish Hospices Foundation - https://hospicefoundation.ie/our-supports-services/bereavement-loss-hub/
		Family Support Northern Ireland - https://www.familysupportni.gov.uk/Support/19/help-you-and-your-loved-ones-cope-with-bereavement The NI Bereavement Network: https://bereaved.hscni.net/
31	PALS - England	https://www.nhs.uk/nhs-services/hospitals/what-is-pals-patient-advice-and-liaison-service/
31	Llais – Wales – Home page	https://www.llaiswales.org/
31	Llais – Wales – Advocacy Guide	https://www.llaiswales.org/have-your-say/raising-concern-about-health-and-social-care-services/llais-complaints-advocacy-guide
31	PASS - Scotland	https://pass-scotland.org.uk/
31	PCC – Northern Ireland	https://pcc-ni.net/about-us/
31	PCC – Northern Ireland - Connect	https://pcc-ni.net/pcc-connect/
32	Patient Advocacy Service - Ireland	https://www.patientadvocacyservice.ie/about-us/overview-and-remit/
32	Serious Incident Response Framework - England	https://www.england.nhs.uk/patient-safety/patient-safety-insight/incident-response-framework/
32	NHS Wales Executive	https://executive.nhs.wales/
32	NHS Wales – Patient Safety Incident Policy	https://du.nhs.wales/files/incidents/patient-safety-incident-policy-2021-05-10-pdf2-pdf/
32	NHS Scotland – The Scottish Patient Safety Programme	https://www.gov.scot/publications/scottish-improvement-journey-nationwide-approach-improvement-compiled-2016-17/pages/4/
32	NHS Scotland – Report an incident	https://www.nss.nhs.scot/health-facilities/incidents-and-alerts/report-an-incident/
33	Northern Ireland Department of Health home page	https://www.health-ni.gov.uk/
33	Health Service Executive, Ireland, home page	https://www.hse.ie/
35	THINK AORTA, THINK FAMILY	https://www.thinkaorta.net/family
36	NHS England National Genomic Test Directory – Testing for Rare and Inherited Diseases	https://www.england.nhs.uk/wp-content/uploads/2018/08/rare-and-inherited-disease-eligibility-criteria-v2.pdf
36	The National Strategy for Accelerating Genetic and Genomic Testing in Ireland	https://www.hse.ie/eng/about/who/strategic-programmes-office-overview/national-strategy-for-accelerating-genetic-and-genomic-medicine-in-ireland/national-strategy-for-accelerating-genetic-and-genomic-medicine-in-ireland.pdf
36	The National Strategy for Accelerating Genetic and Genomic Testing in Ireland – Strategic Implementation Plan for 2023	https://www.hse.ie/eng/about/who/strategic-programmes-office-overview/national-strategy-for-accelerating-genetic-and-genomic-medicine-in-ireland/genetic-and-genomic-implementation-plan.pdf
36	The Scottish Genomic Test Directory for Rare and Inherited Disease	https://www.genomics.nhs.scot/test-directories/rare-and-inherited-disease/
36	Access to genetic testing in Northern Ireland - website	https://belfasttrust.hscni.net/service/laboratory-services/clinical-genetics/
36	All Wales Medical Genetics Service - website	https://cavuhb.nhs.wales/our-services/all-wales-medical-genomics-service-awmgs/

Appendix 1

Example of letter from Clinical Genetics Unit / Genetic Counsellor:

To whom it may concern

Dear Relative

Aortopathy in the Family

Your relative had an aortic dissection (a tear in the major blood vessel leading from their heart). Although the exact cause of this is not certain yet, there is a possibility that there may be an underlying inherited component to this, which we are currently investigating.

Thus, there may also be implications for other relatives, specifically any first-degree relatives (children, parents, siblings). For this reason, we would suggest that such relatives have a baseline echocardiogram to check their main aorta. Ideally, MRI of the entire aorta should be carried out as well.

Please take this letter to your GP to be referred to the cardiologists. Please note that at this point in time, there is no specific diagnosis in your relative. We are just trying to clarify things by gathering information in the wider family, as well as checking your own health.

Yours sincerely

Principal Genetic counsellor.

Space for your own notes

Space for your own notes

Space for your own notes

Aortic
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