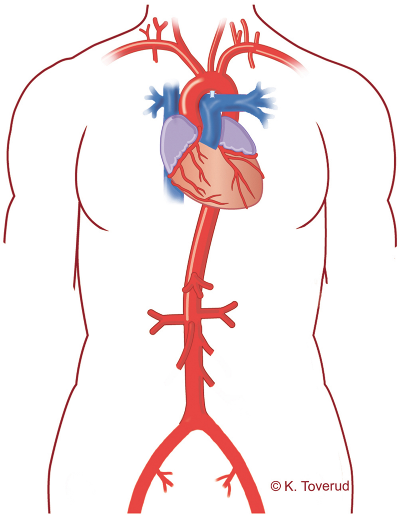
A PATIENT PERSPECTIVE ON AORTIC DISEASE

ECRD 15-16 MAY 2020 STOCKHOLM



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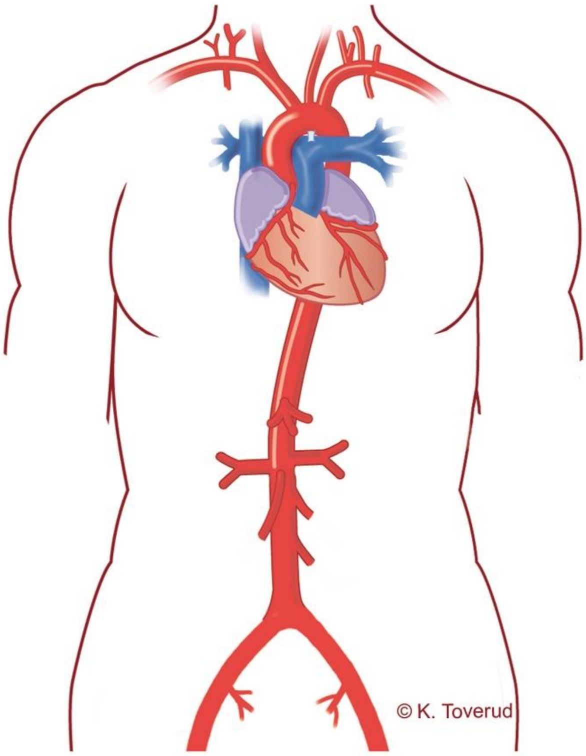

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A PATIENT PERSPECTIVE ON AORTIC DISEASE

Timo Söderlund, Sweden.

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ABSTRACT

Aortic diseases are often rare and deadly diseases. They are hard to detect and, they often don't cause any problems or pain before they are close to an acute phase and can turn deadly very fast.
Because of this, aortic diseases are frequently called "*silent killers*".

This paper covers one patient's personal experience of aortic diseases. "*This is what I learnt, from personal experience, from other patients, from medical articles available on the internet and from interaction with surgeons and aortic disease researchers, since I got ill in 2012*".

When doctors are not up-to-date on research and treatment of rare aortic disease, patients can be told that the aortic disease is probably caused by a stressful life and high blood pressure. This is not always accurate. There can well be an underlying familial disease, not known by that physician.

Recent years, research has taught us that many aortic diseases are hereditary or familial. It is important for us patients and families affected by aortic diseases, to take a stand if we want to help the researchers include our genome and disease history into their research and help them find out more about our diseases.

One of many challenges for us patients is to find good accurate new information as result of new research and findings. Language barriers makes this very hard for a majority of the global population.

There is a massive need to improve country-specific information, in local languages, for a majority of the population on our planet. A patient's future life and health can depend on self-taught knowledge on a rare disease.

The majority of the global population does not have access to acute aortic surgery when needed. Good, easy to understand information in local languages and feasible advice to promote prevention can possibly help many people to a healthier life, and prolong their life before their aortic disease progress.

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CONTENT

NOTE

This paper provides my personal views, experiences and thoughts. I live with Bicuspid Aortic Valve syndrome (BAV), Familial Thoracic Aortic Aneurysm and Dissection (FTAAD) and Atrioventricular Block (AV block) since 2012. I have NO medical education. The contents are written in layman's terms. I had lots of help, but there may still be faults in my spelling, grammar or text. NO medical advice should be taken from this paper. Always ask your own Medical Doctor for such advice.

THANK YOU FOR HELPING ME WITH THIS PAPER

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1. What are Aortic Diseases?

The aorta, is the largest blood vessel in the human body. It is the artery connected to the heart, bringing fresh oxygenated blood to the organs of the body.

The aorta is like a garden hose, with three layers in the aortic wall: an outer layer (adventitia), a middle layer (media) and an inner layer (intima).

A healthy aorta in an adult person has a diameter of approximately 2,5 cm (1 inch), and has a length of about 30 cm (1 foot). It starts at the heart and is separated in the belly into two large arteries that go to each leg (the iliac arteries).

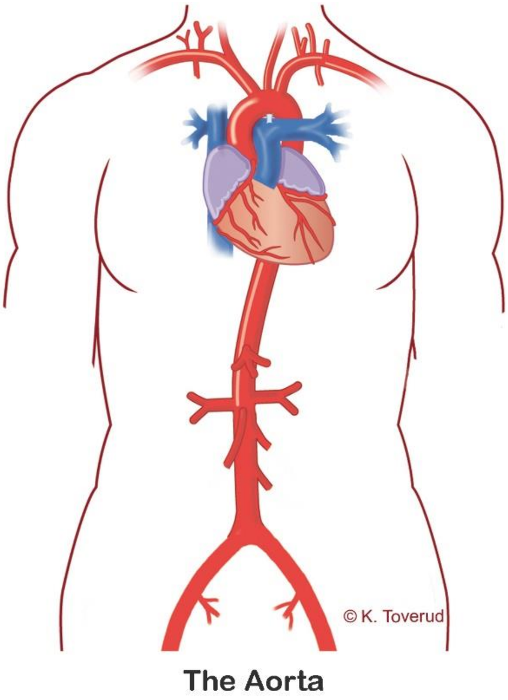

Figure 1: The Aorta. Image with permission by Kari C Toverud, Medical Illustrator www.karitoverud.com

The total volume of blood in an adult person is about 5 litres (1,3 gallon). This entire volume passes the aorta every minute at a normal heart rate ranging from 60 to 100 heartbeats per minute.

An aortic disease is a disease that affects the aorta and its ability to help transport blood to the organs of the body. If the aorta breaks down and the blood flow is interrupted, blocked, or if the aorta ruptures, causing internal bleeding and a loss of blood to the organs, the organs will not get enough oxygen to keep them alive. If the brain does not get enough oxygen this will cause a stroke and possible death.

Different diseases affect the aorta in different ways. Some of the problems that can occur are:

- a bulging of the aorta where the aorta swells like a balloon (***aortic aneurysm***). If the swelling gets too big, the aortic wall can rupture resulting in internal bleeding and death.

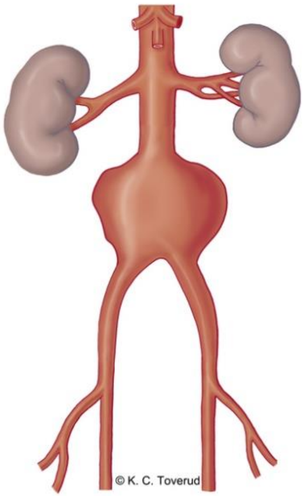

Figure 2: Aortic Aneurysm. Image with permission by Kari C Toverud, Medical Illustrator www.karitoverud.com

- a tear in the inner layer of the aorta (***aortic dissection***) resulting in the blood pushing in to the middle layer of the aortic wall. This creates a new second false blood flow (false lumen), causing a swelling of the aorta and result in problems with blood flow to the organs that are supplied farther down the aorta. The swelling can also lead to a rupture of the outer wall of the aorta which causes internal bleeding and death.

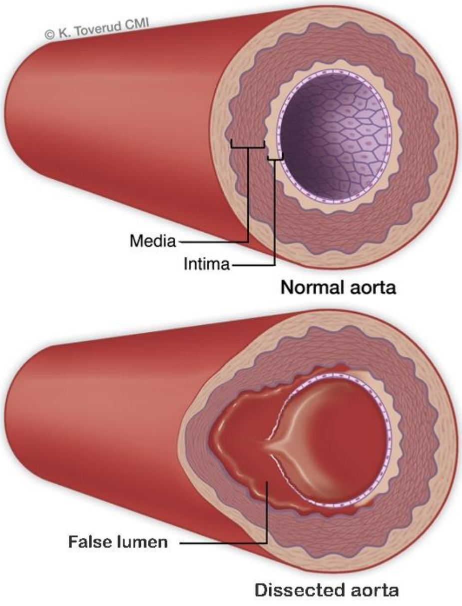

Figure 3: Aortic Dissection. Image with permission by Kari C Toverud, Medical Illustrator www.karitoverud.com

- a build-up of plaque inside the aorta (***aortic atherosclerosis***), hindering the path of the blood and causing a hardening and weakening of the aortic wall. These changes may result in the aortic wall stretching (aortic aneurysm) or tearing (aortic dissection). If the plaque breaks, it can create blood clots which then can lead to problems with the organs and/or the brain, leading to a stroke or death.

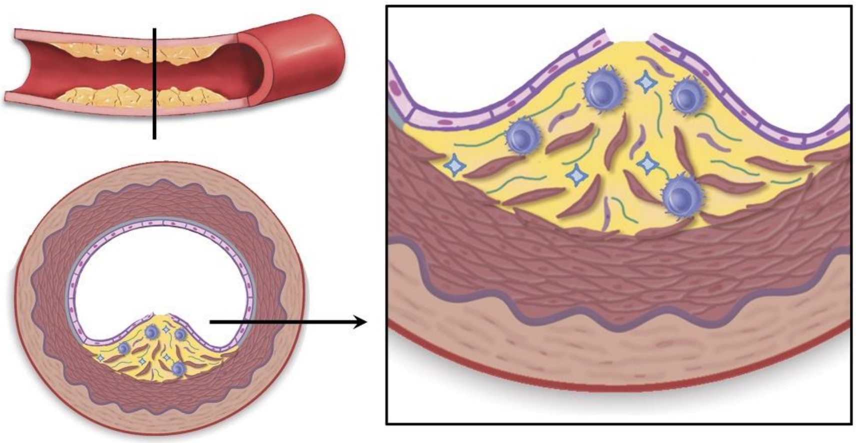

Figure 4: Aortic Atherosclerosis. Image with permission by Kari C Toverud, Medical Illustrator www.karitoverud.com

- an ***aortic intramural hematoma*** (IMH) is a rare condition that causes problems similar to an aortic dissection. It is caused by the formation of a hematoma in the middle layer of the aortic wall. The hematoma is believed to be caused by a bleeding from the tiny network of blood vessels inside the aortic wall (vasa vasorum), supplying the aortic walls with oxygen and nutrients. The progression of this disease can cause both aortic dissection and aortic rupture which can result in internal bleeding and death.

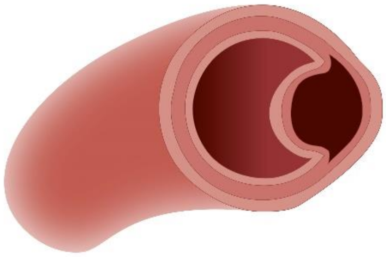

Figure 5: Aortic Intramural Hematoma. Image with permission by Deutsches Herzzentrum Berlin, Germany.

- ***penetrating aortic ulcer*** (PAU) affects the aortic wall. PAU may be found chronically in patients with significant aortic atherosclerosis. Together with aortic dissection (AD) and aortic intramural hematoma (IMH), PAU is potentially life-threatening. These three diseases (or conditions) are often called ***Acute Aortic Syndromes*** (AAS)

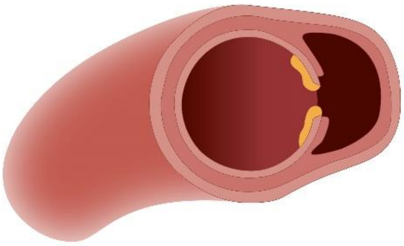

Figure 6: Penetrating Aortic Ulcer. Image with permission by Deutsches Herzzentrum Berlin, Germany.

- inflammations in the aortic wall (***aortitis***) causing a narrowing of the aorta, and later a bulging (aneurysm) or tear (scarring, rupture or dissection). Takayasu's Arteritis is a rare disease that causes aortitis.

- various rare ***malignant tumour diseases*** that affect the aorta causing problems related to the aortic wall and blood flow. Some are also metastatic diseases, causing death.

Bicuspid aortic valve syndrome (BAV) is when a person is born with the heart valve connecting the heart and the aorta (the aortic valve) with only two leaflets (or cusps) instead of three. BAV is not a rare disease. It affects 2-4% of the population. But in some cases, BAV (probably in combination with a rare disease) will affect the aorta. A part of the patients with BAV have *aortic aneurysms* in the aorta close to the heart, and some have a narrowing of the aorta a bit away from the heart (called *aortic coarctation*). The aortic aneurysm may eventually tear (aortic dissection) or rupture, resulting in internal bleeding and death.

There are many other diseases or syndromes that are associated to aortic disease. Some of them are connective tissue disorders like:

- ***Loeys Dietz Syndrome***
- ***Marfan Syndrome***
- ***Turner Syndrome***
- ***Vascular Ehlers-Danlos Syndrome***
- ***Williams Syndrome***

As you now understand, aortic diseases are often deadly and complex diseases. They are hard to detect and, in many cases, they don't cause any problems or pain before they are close to an acute phase (with an aortic dissection or aortic rupture) and can turn deadly very fast. Because of this, aortic diseases are frequently called "silent killers".

If we can find aortic diseases in time, we can treat the patient in various ways. This can include:

- follow-up and medicines to treat hypertension (high blood pressure)
- teach patients more about their disease so that they can adopt their lifestyle to minimize risk of progression of their disease at a to early age, like quit smoking, eating healthy, exercising daily and generally adopting to a healthy lifestyle.

If we are in a country with available aortic surgery for the general population, we can also offer the patient elective planned surgery. This is much safer for a patient than any emergency event and emergent surgery if and when the disease turns acute.

Note 1.1: *In addition to diseases that affect the aorta, the aorta can get damaged and stop functioning properly due to misuse of narcotics (like cocaine), accidents with heavy blunt trauma to the chest (like car accidents), and gunshot wounds or stabbings.*

2. Aortic Diseases. Are They Rare Diseases?

Yes. Many of the diseases that affect the aorta are categorized as so-called rare diseases.

A physician working in a small hospital meets patients who are affected by as many as up to 10 000 (well known) diseases, affecting a fair part of the population. These can range from pneumonia, stroke and heart attacks, to common forms of cancer.

In addition to these 10 000 well-known diseases, there are rare diseases. A rare disease, is a disease that affects only a small part of the population.

There are registries listing the diseases considered to be rare diseases. Various countries, list various amounts of diseases as rare diseases. Then there are international registries like Orpha.net. In 2019 it listed over 6 000 rare diseases. According to Orpha.net there were over 750 different rare disease-registries in Europe in May 2019.

Different countries have different ways in which they classify rare diseases. In some countries the criteria are that if a disease affects less than 1 person per 2 000, it is a rare disease. In other countries the criteria are less than 1 out of 10 000 (ten thousand). Some are even more rare. Like Takayasu’s Arteritis that only affect a handful (or less than a handful) of people out of a population of 1 000 000 (one million). Because there are many different rare diseases, recent estimates state that almost 350 million people on our planet are affected by a rare disease (4% of the global population).

Research on aortic diseases is done by researchers in many countries around our planet. It is very much needed, because many patients, even in 2020, still cannot get a proper diagnosis or reason for the underlying cause for their aortic disease. If we can find their disease, researchers can also develop appropriate treatment for that disease. Much more could be done in this field. The few successful researchers in the world need more support and more resources for their important work.

In many cases, when doctors are not up-to-date on the latest research and treatment of aortic disease, patients are told that the aortic disease is probably caused by a stressful life, and high blood pressure. This is not always accurate. There can well be an underlying disease, not known by that physician. Knowledge perhaps only known by researchers or medical doctors who are aortic disease experts.

Sharing information of recent findings by the researchers, is very important. We all need to help bring out the new results and new findings to our local hospitals and physicians. This is a simple way to faster give a basis for a better care for us patients.

Many researchers today know that stress, smoking, high blood pressure, and misuse of cocaine will make the underlying aortic disease progress faster. Recently scientists have found many genes and familial diseases that can be linked to a patient's aortic condition, and more are discovered every year. Most of these are rare, and classified as so-called rare diseases. It is likely many more are to be discovered the upcoming years.

There is a need for increased knowledge on rare aortic diseases for the medical community. Therefore it is important for us patients and families affected by aortic diseases, to take a stand if we want to help the researchers include our genome and disease history into their research and help them find out more about our diseases.

Increased awareness and good written information on the latest findings on aortic diseases can help doctors and physicians in smaller hospitals gain knowledge on the new findings on these rare diseases. Patient initiated awareness campaigns like "***Think Aorta***" and the "***Aortic Disease Awareness Day***" are important and successful ways to bring attention to news in the medical field. Both for medical professionals who are not experts on aortic diseases, as well as for the patients and their families.

Note 2.1: *Read more about THINK AORTA and AORTIC DISEASE AWARENESS DAY on page 21, in section 8. Ongoing Interesting Projects for Aortic Diseases*

3. Can an Aortic Disease be Hereditary?

Research over the last 20 years has taught us that many of the diseases that affect the aorta are hereditary or familial. Many of them are not well-known, some are categorised as rare diseases. Education to become a general physician does not include much knowledge about these, still little known, and often deadly diseases. Good knowledge is found mainly among medical doctors, aortic surgeons and researchers who spent more time for specialised studies and research around aortic diseases.

In cases for patients where there is no proper diagnosis of the underlying cause of the aortic disease, an investigation of the family disease-history can show that several people in the same family have suffered from aortic problems. This is a strong indication that the family has a hereditary aortic disease.

Researchers find new genes linked to aortic problems every year, and the list of known genes linked to aortic disease keeps growing.

A problem for some patients suffering from aortic diseases, and their families, is that many patients are sent to big university hospitals for acute aortic surgery when they become ill, and later they are returned to their smaller local hospital for continued treatment and follow up. These smaller hospitals do not perform advanced aortic surgery or genetic research on aortic diseases, so the local knowledge on these diseases, their diagnosis, follow-up and treatment are not up to date.

Even worse, the local small hospital believe they can treat these patients, but with a rare underlying disease, the patients can face medical complications that the local doctors don't anticipate.

People with aortic disease are encouraged to talk to a major teaching university hospital, or a specialized and established *Aortic Center,* affiliated to a known hospital that performs research on, or treats aortic diseases, when trying to figure out if it is a familial disease that caused their aorta to fail. They should get help to look for clues within their own families, because their siblings, children and grandchildren might be at risk.

In some cases, genetic testing can be helpful. However, sometimes there are no known genes. Still a family may have had multiple deaths caused by ruptured or dissected aortas. As important as genetic testing, is interviews and outlining a family's disease history. Screening of the aorta or aortic valve can give also answers when trying to figure out if a person is at risk of an aortic disease.

4. A Global Perspective on Aortic Disease from a Patient

4.1 First challenge is finding others with the same rare diagnosis.

In 2020, 8 years have passed since I survived an acute aortic dissection type A in 2012. I have seen a global community of aortic disease patients take form and grow over the past 5-6 years with help of social media. The main platform for patient interaction has been, and still is, Facebook.

It was originally mainly people from USA with aortic diseases who were active on social media, and who dominated the existing social media channels, using English language. For people, who do not live in the US, this can be a problem when trying to find good information. These big communities are mainly highlighting the situation and solutions available in the US.

But now this trend is growing in additional countries using national languages with regional or national platforms for patients. This enables patients to find discussions that are more relevant for them. People affected by rare aortic diseases are able to learn more about the actual healthcare situation, knowledge and available treatments in their own country. A national focus is important for patients in many countries, as the healthcare systems and solutions offered vary dramatically depending on the country.

Part of this development (and growth globally) with a patient centred on-line based community is initiated partly by the global **Aortic Disease Awareness Day**, or the September 19 movement, that has reached new countries every year during the last 4 years. The movement in the Awareness Day has ***promoted use of local languages*** to help reach people in many countries.

September 19 is day when big hospitals with good knowledge on diagnosis, treatment and follow-up of aortic diseases, invite their aortic disease patients, to a day filled with information. It is also a good way to allow patients and their families meet other patients who share their rare diagnosis.

The growth of the aortic disease awareness movement to new countries is also a direct result of the two multi lingual platforms for rare disease patients based on mapping systems, that have specific communities for aortic disease patients. One is called **RareConnect** and was founded by the rare disease umbrella organization, *Eurordis*, in Europe.

The other is called **DiseaseMaps**. DiseaseMaps was founded by a rare disease patient in Spain who was looking for others sharing his rare disease. He created DiseaseMaps as a new way to let rare disease patients find each other in real life.

Note 4.1: *Read more about AORTIC DISEASE AWARENESS DAY on page 21, in section 8. Ongoing Interesting Projects for Aortic Diseases. Read more about RARECONNECT and DISEASEMAPS on page 24 in section 9. If you have an aortic disease - you are not alone. Examples of ways to find others with your aortic disease.*

4.2 The most established patient organizations are in USA.

Many well-established organizations in the US are, and have been, front-runners in the way they create public awareness for aortic diseases. They could easily be said to be ***centres of excellence*** in their way to communicate in the US. Many countries can look, listen and learn from these organizations.

Aortic awareness organizations in USA are often ahead of the rest of the world. My understanding of the reason for this, is because the technique of aortic and cardiac surgery originally was developed in the US (together with Europe and Scandinavia). The hospitals in the US have a long history of advanced cardiac and aortic surgery compared to many other countries.

What differs USA from Europe and Scandinavia is the size of patient communities. The US are well ahead of the rest of the world, since they had many surviving patients talking the same language in one single country for a long time. This made it possible to start organizations and foundations attracting fairly good numbers of surviving patients over the years. In Europe we have more than twice the population, but we have surviving patients living in over 50 countries using 24 different official languages.

Many of the old organizations in the US share one obstacle that probably prevented them from growing outside USA. It is a partial lack of understanding that the US only has less than 5% of the global population, and that English is only understood by less than 15% of the global population.

For a movement to grow with people in the other 85% of the global population that do not understand English language, or with the 95% of the global population that does not share the medical and social systems of the US, one has to understand, and cope with these important facts when communicating with people outside USA, if we want to attract them to a joint patient community.

A way for a movement to grow in many places globally could be to adopt to national and regional circumstances locally, based on local settings and local language, and to only use a general value-base globally. Such a movement need to form locally on national basis depending on the current situation in a specific country, and be led by people who are engaged and who communicate in the language that the people understand in such a country. Such a movement may need to have different goals and visions depending on where in the world it is located.

In a country like USA or Sweden one goal can be to improve accuracy and speed when it comes to make a proper diagnosis in an emergency setting. In a country like Bangladesh, Mozambique or Indonesia, it is perhaps more about sharing good information about aortic diseases, and teaching people about what can prevent or progress such diseases, so that people affected by aortic diseases can make choices in their lives, based on this new knowledge.

4.3 Trying to overcome the language barrier – trying to reach out with help of aortic surgeons

A feasible way to follow the global knowledge on rare aortic diseases is to try to seek out the aortic surgeon perspective on aortic diseases. The aortic surgeons have chosen LinkedIn as their main communication platform on social media. Aortic surgeons often speak English, since part of their long education in medicine includes the use of foreign languages.

This make the surgeon community easier to access on a global level (with use of just one language) than in the patient community, where language skills are not as wide as in the highly educated surgeon community.

What I have learned from LinkedIn, and the surgeons I have gotten to know there, is that the skills in advanced aortic surgery are well developed in many countries over the world. Countries like Spain, Italy, Germany and France does not use English language among their patients. They instead use Spanish, Italian, German and French languages.

In these countries the medical and healthcare solutions and availability of medical assistance when a person suffers acute illness in an aortic trauma, can be as good, or sometimes even better than in the US for common citizens. Especially in regions around Paris in France (with the SOS AORTE concept) or in the Berlin region in Germany (with the AORTENTELEFON concept)

The patients with rare aortic diseases who do not speak English, were much more isolated a few years ago, and did not get as good information as the patients who also knew English language.

Today there are some good patient-administered aortic disease initiatives growing in a handful countries in Europe, using local languages. Examples are Germany, France, Sweden, Italy, The Netherlands and in the United Kingdom. But there is still a significant need for better local information in many countries, especially where English is not well known among the native population.

A challenge for the patients is to find good new information as result of new research and findings due to the language barrier. This will improve if the patients can find each other's and start patient-centred initiatives for the patient community in a country. The ***Aortic Disease Awareness Da***y movement and September 19 can be one way to help local initiatives to form in more countries.

4.4 Only a minority of the global population have access to advanced cardiac and aortic surgery.

Life is not fair. There are over 200 countries in the world. Only in a minority of all countries, the advanced medical solutions to save patients from acute aortic disease are available for the majority of the population.

In countries where such advanced medical care is not financed by taxes, the care is limited to those who can afford to pay for medical insurance. Acute open-heart surgery and open-repair of the aorta are very expensive treatments. As an example, the US insurance companies can be billed by hospitals up to over 500 000 USD for one acute open-heart surgery with repair of the aorta, with following post-operative treatment, for one single patient. With complications and longer time in intensive care after the surgery, the bill can easily go up to over 1 million USD. Money that the majority of the population on the planet will not be able to save in their full lifetime.

In some countries there is a limited resource available, and this is allocated for the political leadership of the country, for the military and for the few very rich people who can afford it.

In countries with very limited resources, or lack of resources, the vast majority of patients who suffers from acute aortic disease will die within hours or days from the moment when their aorta fails them. In many countries care is only possible for those few who can afford to go abroad for surgery.

4.5 There is a need for easy-to-understand accurate aortic disease information globally

The lack of comprehensive, accurate and easy-to-understand written information about aortic disease - its cause, how to diagnose it, possible measures to help prevent, or to delay it - in the vast majority of the languages used in the world is a major problem for the families of those who die.

The family lose a family member to a deadly aortic disease, but have no clue that this disease may be present in other family members because signs or markers ore often not apparent without medical testing or digital imaging of the aorta.

We could consider compiling some of the recent excellent information to useful brochures for patients and their families. If we then also translate them into more languages, more families could probably gain more healthy years alive by active life style choices, knowing there are steps and measures to take if a family is at risk for deadly hidden aortic disease.

Simple lifestyle decisions like avoiding smoking, avoiding obesity, promoting daily exercise, avoiding high blood pressure with help of monitoring and using available affordable medication – are all measures people all over the world, even those in poor countries, can choose. Choices that most probably will offer them more healthy years in their lives.

In summary, as I see the global patient-centred view on these diseases, we have a wide range of needs depending on what country we live in. The needs vary due to political and healthcare systems, available medical resources, financial ability and language barriers.

In the rich countries the needs of the patients can include;

- new treatments,
- developing patient centred post-operative care and follow-up,
- increased knowledge around familial disease and introduction of general screening of family members,
- faster response and more accurate diagnosis when a person turn acute ill,
- better, more effective and affordable medicines with less side effects.

In the poor countries, we probably need to focus on good, easy to understand information and feasible advice to promote prevention.

I notice very little activity in the area of global prevention of aortic diseases. In some cases, there is a little, but in most cases, there is nothing.

There is a massive need to improve country-specific information, in local native languages, for a majority of the population on our planet.

5. Available Written Information to patients globally

There are various amounts of written information for patients with aortic diseases. For a more common condition, like aortic aneurysms, there are more material in quite a lot of languages. For a much rarer condition, like acute aortic dissection - what causes it, how to diagnose it and how to treat it - there is a total lack of good written information for patients in most languages.

The manufacturers of stents and grafts (devices used by cardiac and vascular surgeons to repair the aorta) have produced product brochures for hospitals. Because these brochures are technical and quite promotional, they are not helpful or appropriate for patients. Most patients do not ever get to see these brochures.

The more well-known diseases, affecting the aorta, like Marfan Syndrome, have well established patient organizations in some parts of the world. These organizations have produced an array of good and useful written material in local languages for patients and their families.

For the still less known diseases affecting the aorta, there are only a few good patient-centred brochures available. In recent years some excellent brochures have been produced by hospitals that have good communication with their patient community, and who have listened to the needs for a patient perspective in such material.

Here is one of the best examples of a recent publication written for patients by medical experts:

"***Patient information Aorta***" – patient-centred brochure published September 2019 by Professor Volkmar Falk and Dr Stephan Kurz at ***Deutsches Herzzentrum Berlin*** (DHZB) Germany. The patient brochure is available on the internet in German and English language.

Standard values of the aortic cross section

	Men	Women
Aortic root	3.6-3.9 cm	3.5-3.7 cm
Aorta ascendens	2.9 cm	2.9 cm
Aorta descendens	2.4-2.9 cm	2.5-2.6 cm

Figure 7: Aortic Disease Awareness Day event in Berlin Germany on Sept 19 2019 with over 100 participants. ''Patient Information Aorta''. Brochure on aortic diseases for patients, presented in English edition by Deutsches Herzzentrum Berlin, Germany.

Note 5.1: *On LinkedIn, I have a shortlist in an article called ''**Basic info toolbox for patients with aortic disease**''. Here I collect downloadable information in 8 various languages on Aortic Diseases.*

Note 5.2*: For many languages there still are NO printed or downloadable good brochures explaining general and rare aortic diseases from a patient perspective. This is one of the challenges we need to address on a global level. Only through good, accurate and up-to-date information, can patients affected by aortic disease learn about their disease. Information that can help them make proper life-style choices to preserve their health.*

6. Challenges for Surviving Patients and Their Families

6.1 Surviving Acute Aortic Syndromes (AAS)

A patient who survived an acute aortic disease, like an acute aortic dissection type A, will be affected for the rest of their life. To survive, the patient requires acute cardiac surgery to replace the first part of the aorta with a vascular prothesis (called a graft). Many times, the course of time is very short from initial problems experienced by the patient, until open heart surgery is started in a big hospital with resources, expertise and experience of such life-saving surgery.

The hours between the initial acute symptoms from the aorta until the start of an acute surgery, are often dramatic and filled with emergency medicine providers trying to identify one of the most difficult medical diagnoses in a short amount of time.

If the patients get ill in a place with no available resources for acute surgery, an immediate emergency transfer is required to a larger hospital by helicopter or ambulance. Many times, the patient has lots of pain, and is given morphine, or other painkillers, to ease the pain.

Time is critical, and the patient often does not comprehend everything that is going on during these few very stressful hours. Sometimes the patients faint. Finally, when surgery is about to begin, the patient is sedated and given anaesthesia for the operation.

A day later, or a week later, of a month later the patient awakes after a very complex surgery which has taken from 6-12 hours. A surgery where the chest has been opened, the heart stopped, and the aorta repaired. Depending on the severity of the condition and potential complications, the time in intensive care after a successful surgery can take anywhere from a few days to weeks or even months. One of my closest friends woke up 6 weeks after his surgery, having been in a coma caused by stroke during surgery.

The lack of time to prepare mentally for such an invasive procedure, causes many patients to gradually experience emotional trauma, especially over time as they begin to understand what could have happened.

Some patients later suffer from post-traumatic stress disorder (PTSD), depressions and anxiety as a result of such a dramatic and traumatic event.

6.2 Cognitive skills may become impaired as result of the surgery that saves the patient’s life.

Some patients suffer from strokes, microbleeds or oxygen shortage as complications from acute open-heart surgery. Such complications, also in combination with post-op treatment with various medications to control blood pressure, and in combination the mental trauma some patients experience, can give cause to different problems.

Many acute aortic dissection type A patients I got to know over the years mention similar problems. Poor short-time memory, problems with spelling, problems in finding the appropriate word during speech, coordination of movement, delayed reaction time, problems with sight and vision – all in all - a mental and physical state where the patients experience problems in recognizing themselves as the same person they were before the acute surgery.

Some doctors in smaller hospitals do not have the experience to handle minor brain injuries, and more easily respond to patients with more visible problems, like partial paralysis or inability to walk or talk. If you are confined to a wheelchair a problem is easier to spot, than if you can walk, talk and smile.

I can just take myself as an example.

It took me two years after my surgery, to finally come to a brain damage unit in a larger hospital. The first two years after surgery I tried to explain that I did not recognize myself at all to doctors and nurses in my local hospital. Having brought up this issue many times, the hospital finally decided to performed a simple test on me. They showed me an image of a cat, and asked me what it was. I said "*it is a cat*". Then they showed me an image of a house. I said "*it is a house*". They asked me what day it was, and what month it was. I answered. They also asked about the weather outside. I told them about the weather. The test was done, and I was told I had no problem. They said that my mental state with some cognitive problems was normal and would go away with time. They were very clear that they had no idea how to help me, and that no help was to be found for what I experienced as my problems. I got a feeling they did not see any problem at all.

Finally, I got to hear about an expert centre treating and researching brain damages at an university teaching hospital, from other patients who had survived acute open-heart surgery. I contacted the hospital, and was asked to come for an interview.

One simple test when I was there, clearly revealed some of my problems. On my first visit, the professor who interviewed me asked me to count loudly from 100, backwards, in steps of 7. "*100*" and "*93*" was OK. But then I came no further. Trying to subtract 7 from 93 in my head was impossible. Trying to pass 90, I lost track of the figures. Another additional 20 tests confirmed and pointed out a direction of where my specific problems were to be found.

I was treated as a patient in this expert unit for almost 2 years, and got excellent help to recover part of "*my old self*". The feeling to meet people who understood my problems and a place where it was considered "normal" to experience such problems, made cry and feel relieved. Doctors who thought my problems were pretty normal, and also knew how they could be addressed.

6.3 Leaving the hospital, and facing the realities of the outside world can be a challenge

Many patients survive aortic disease to find themselves in a new mental state, that beside the physical situation can change their course of life totally. Many patients refer to this as "***the new normal***" A patient who goes from an ordinary-day normal life one day, to being stuck in a bed in a hospital, or at home, unable to work. Not really knowing how long this new situation will last.

This can affect patients and their families financially. They are often dealing with unexpected and unplanned medical procedures, test and surgeries. Patients and families have to contend with insurance companies, authorities as well as employers.

Different countries all have their own systems for insurances, healthcare costs, medical expenses, compensation during sick leave form an employment. Some countries have listed some aortic diseases and conditions in their insurance policies, others have not.

Not only is the person not able to work for some time after surgery, on top of that insurance companies may not be helpful since they do not have proper diagnoses listed, and employers often don't have any understanding of what the employee is suffering from.

If complications from surgery include minor or major brain damage, PTSD or depressions, insurance companies may have a problem understanding that they all in the perspective of the patient, relate to the aortic disease.

An insurance company or a doctor often are specialized in managing or treating specific diagnosis. The aortic disease patient may have a variety of different diagnosis. All of them as a result of the initial aortic problem. In the view of the patient – they are all part of the "***aortic disease***".

The patient needs to go to multiple hospitals or multiple doctors for treatment. Doctors and hospitals who often are not coordinated. This discrepancy in the view of one single patient, and the diagnoses, can be extremely frustrating for the patient.

The investigation of an insurance company may turn to doctors, educated some 20-25 years ago, who lack info on today's latest research on aortic diseases and how to treat them. The patients are then to be evaluated and decisions are made, without insurance companies or employers fully understanding all the aspects of an aortic disease, its treatment and how it can affect a person.

I have experienced this myself, and have also seen other people suffer after being denied their right to insurance benefits, due to lack of understanding of the disease or diagnosis that affects the patient.

Sometimes patients who can stand up, walk and talk are told by employers or insurance companies, "*you are fine now, your aorta is repaired and you are fine – you can now return to your old life*". The reality can well be that the patient still has;

- a weakened and dissected aorta beside the part that was mended,
- several dissected arteries,
- aneurysms on the aorta and in some of the arteries connected to the aorta,
- minor brain damage causing cognitive problems,
- mental problems with PTSD, depression or anxiety,
- problems with the heart as complication from life-saving open-heart surgery.

Note 6.3.1 *Many problems cannot be detected by looking at the face of a patient in a brief meeting, or just by evaluating a single diagnosis like Aortic Dissection Type A, or a medical code like ICD-10-CM I71.01. Aortic Disease patients with multiple diagnosis all caused by an initial aortic disease, may need a holistic assessment where all the diagnoses and problems are weighed in for a proper evaluation and decision.*

6.4 Post-operative care follow-up can vary a lot.

The post-operative care is very much dependent on a local hospital, as well as available resources and knowledge surrounding follow-up and rehabilitation of aortic disease patients. Some hospitals have advanced multi-skilled "**aortic centres**", with affiliated surgeons, cardiologists, physicians, physiotherapists, dietitians and psychologists – a full team covering most aspects of care needed by an aortic disease patient. Other hospitals have nothing to offer their patients, more than regular care, because they lack the skills, knowledge and understanding of this rare patient group.

One way for patients to try to overcome this obstacle, is to learn as much as possible about their disease. Both about the disease itself and available treatments.

One way to do this is to connect to other patients with the same diagnosis. When talking to patients in other countries, or even regions in the patient's own country, a patient can hear and learn about solutions and healthcare options provided in places on earth where knowledge is higher, and understanding and interest have formed more appropriate local healthcare solutions and treatments for these patients.

Sharing this new knowledge to interested doctors and physicians in the patient's local hospital, can help improve the situation for aortic disease patients. As the local hospital learns more, and understands more, about the needs and problems connected to aortic disease, this is a benefit to all the aortic disease patients in the hospital.

6.5 What about the first-degree relatives to the person who got ill?

The close family of all patients who are identified as having aortic disease should be evaluated. Whether it is an acute or accidental finding of an aortic disease, investigation in the parents, sisters, brothers, and children of the patient will often lead to finding aortic disease in ***these first-degree relatives***. All parents, brothers and sisters should get evaluated immediately, unless the sibling is much younger than the patient.

Note 6.5.1: *Children or siblings should be evaluated at an age 10 years before the age of the patient when he or she got acute ill or diagnosed. Or at age 50. Whichever is younger. (Ref Dr Benjamin A. Youdelman, USA)*

Note 6.5.2: *Read more about screening of first degree relatives in an article in the Journal of the American Heart Association from 31 July 2018 "Systematic Review of Studies That Have Evaluated Screening Tests in Relatives of Patients Affected by Nonsyndromic Thoracic Aortic Disease" by G Mariscalo, R Debiec, J A. Elefteriades, N J. Samani, and G J. Murphy*

6.6 Self-taught knowledge can help improve quality of life for aortic disease patients.

A patient's future life and health can very much depend on self-taught knowledge on the disease. Example can be found in use of blood pressure medication, blood thinning medication or painkillers. With accurate knowledge, a patient can gain the personal strength to dare say no to local physicians who does not understand the effect on certain medications, or the effect of badly combined medications, for a dissected aorta, or weakened, dilated aorta.

Several times I had to correct emergency medicine doctors who wanted to give me painkillers that are directly dangerous for patients with my diagnosis. I also had to resist doctors who wanted to cut some of medicines in local programs (run by hospitals to reduce the general use of medicines), having to explain to the doctor why I am on the specific medicine and what it does for my arteries or heart. Only genuine knowledge can help a patient find strength to discuss the local physician's proposal in such situations.

7. Challenges in Countries where there are no ways to treat Patients with Acute Aortic Disease. Can we do more by Prevention?

7.1 Aortic Diseases are often called “Silent Killers”. Most people will die when they get acute ill.

"*Three out of four people (75%) in the world does not have access to cardiac surgery when needed because of lack of infrastructure, human resources, and financial coverage.*" This is according to a medical paper from 2019 (ref 7.1.1.)

We all know from what we read in this paper, that acute problems with the aorta are life threatening, and in many cases acute cardiac or vascular surgery is the only option to save the life of a patient.

So how do we help families with deadly aortic disease in countries where the treatment is too expensive or not available at all? Do we just accept that appropriate diagnosis, treatment and care is for a few people in the rich countries on the planet, and close our eyes to those who live in countries or regions without advanced healthcare solutions?

For me, a life is worth the same, no matter if it is in London in United Kingdom, in the jungle or desert in a country on another continent, where languages other than English are spoken. For me, an effort to help a person or a family is as much worth, in any language, in any country on the planet.

We need a new systematic approach for families affected by deadly aortic disease, in the low- and middle-income countries. Countries without resources to treat them, when they are acutely ill.

In the short and medium term, we are not able to provide the majority of the global population with surgery or even proper diagnosis in time. To think that we even in a full life-time would be able to build up the most advanced cardiac surgery units in all countries in the world, and eliminate poverty in the world so that all people can access such advanced healthcare - is not a feasible way to help people. It is a dream, and such dreams do not help the poor people in the world here and now. They will continue to die, as we who have access to the best up-to-date information and expert care keep dreaming.

Reference 7.1.1: ***Global cardiac surgery: Access to cardiac surgical care around the world***, *2019 April 26, by Dr Dominique Vervoort, Dr Bart Meuris, Dr Bart Meyns and Dr Peter Verbrugghe*

7.2 A pragmatic solution can help people in the short- and medium-term perspective

However, we can choose a realistic, pragmatic approach to the problem for families affected by aortic diseases in the poor countries.

We just need to try to picture ourselves in such a country, living there with a still hidden undiagnosed deadly aortic disease.

How can I improve my health and possibly gain more healthy years if I had a notion of what disease was dwelling inside my body? Would I like to know what kind of life-style choices that possibly can offer me more healthy years before I eventually get acutely ill and die from my disease? Would I like to know? Would I like to have the opportunity to choose how I live my life, if I have the knowledge available?

For me this question is easy to answer. Having been close to death from acute aortic dissection type A, and then surviving – I would choose to answer YES to all the above questions. If I could gain more healthy years by knowledge, and by making positive life-style choices, I would do it.

What we in the rich countries can choose to do, is to help provide families affected by aortic diseases in less rich countries with good information about their diseases. Including what makes the disease progress, and how to make life-style choices to prevent or slow progression of their disease.

To do this, we need to identify a good, up-to-date, easy to understand, written information, and then secure appropriate funding and resources to translate it into many languages so it can be available over the internet for free.

Medical doctors can choose to hand over such easy-to-understand information to patients' families. This can be done when a patient either has died from a confirmed aortic disease, or where the diagnosis is made, but treatments are not available to the patient. The patient may be lost, but the information will be helpful to the rest of the family, helping children and siblings.

Such easy to understand and pragmatic knowledge can help the family learn about the disease, and help them make choices in their own lives to help them choose a healthier lifestyle that does not cause further progress of the disease. Familial aortic disease may be in their genes, still dormant and hidden.

If we accept this **pragmatic view** to address awareness in the countries with no possibility to offer treatment, we can use this approach everywhere. The knowledge of these diseases, the steps, actions and choices we all can chose to make, that possibly can give any person on the planet another 2-3 years of healthy life, (or more), independent of later access to thoracic surgery or super advanced care.

This is of course of benefit for all people. This includes people in the US or Europe, who has access to the best treatments in the world, but perhaps do not have time enough to make it to the hospital and get a proper diagnosis and treatment the day their aorta rupture or dissect, before they are dead.

Advice, such as "***stop smoking***", "***avoid obesity***", "***avoid hypertension***", and "***go to see your local nurse or medical doctor to check your health***" becomes understandable and meaningful in a new way. Eat healthy food. Move your body every day. Take care of your body. All this will probably help offer people a few more healthy years in their lives. Not only in Asia and Africa but also in America and Europe.

Figure 7: In 2015-2016 the Aortic Disease Awareness Day team had illustrations made to promote aortic disease awareness and prevention. A cartoon was invented for the September 19 movement called "Lady Aorta". All images were donated to The Marfan Foundation in the beginning of 2019.

8. Ongoing Interesting Projects for Aortic Diseases

There are many ongoing amazing projects about aortic diseases in the world. From a patient perspective I would like to highlight six such initiatives that need worldwide attention, so others learn from them. Solutions that perhaps can be used and shared by national or local hospitals in many languages, for many people to benefit from.

The first two solutions which I came across in my search for aortic diseases, their diagnosis, treatment, care and prevention, opened my eyes to two similar solutions in Europe. Two solutions made in two countries, but that resemble each other very much. Solutions that spurred two large hospitals in Paris (France) and Berlin (Germany) to increase the annual amount of acute aortic surgeries with 60-70% over night.

Both solutions help a smaller regional hospital make a faster and correct diagnosis for acute aortic syndromes when patients fall suddenly ill. Both **Aortentelefon** and **SOS-Aorte**, are concepts with regional information and communication between health-care providers, where aortic trauma is made visible by a concept with folders, posters, website and educations of healthcare providers.

In addition to highlighting acute aortic syndromes, both systems also provide guidance, including special telephone numbers, and a defined process on how to how to proceed from a smaller healthcare provider to get expert assistance immediately in the region, if an acute aortic syndrome is suspected.

8.1 Aortentelefon

"AORTENTELEFON" in the Berlin & Brandenburger area in Germany, created by ***Deutsches Herzzentrum Berlin (DHZB)*** with expert support over the phone and internet 24-7 to regional smaller healthcare providers, lead to an immediate increase in aortic emergency surgeries from 80 surgeries the year before the opening of the hot-line, to 138 surgeries the year following the new medical service was launched.

8.2 SOS Aorte

This French model to improve diagnose of acute aortic syndromes was introduced in the Paris area of France, with a specialized medical doctor providing 24-7 hot-line professional assistance to the regional hospitals, emergency medicine, and healthcare providers in their region.

It is done to help regional healthcare providers make a proper aortic diagnosis, and to help coordinate the needed care to save the life of the patient. ***SOS AORTE*** at ***Hôpital Européen Georges Pompidou***, lead to an immediate increase in aortic emergency surgeries from 174 surgeries (done the three years preceding the opening of the hot-line), to 287 surgeries (the three year period following the opening of the telephone line).

SOS AORTE has since been copy-pasted to other big hospitals in other regions of France with good results. One such example is ***Centre Aorte Timone*** in the Marseille region.

8.3 Vascupedia

You find VASCUPEDIA on this website: www.vascupedia.com

VASCUPEDIA is an open-access platform useable for medical professionals worldwide. It is founded and managed by a team of vascular surgeons in Germany. Vascular surgeons, thoracic surgeons, cardiologists, researchers, physicians, interventional radiologists, nurses and technicians are welcome to take part.

On Vascupedia medical professionals can **share experiences**, express opinions, participate in discussions and **learn from others**. Users can upload their own presentation (case reports, recorded video cases, lectures) or comment on posts of other participants. Active participation of the users of Vascupedia will help contribute to the education experience of vascular specialists and will most probably improve vascular care worldwide. The contents of Vascupedia is available also for patients and patient advocates who are passionate about learning more about vascular diseases.

8.4 Vascern

You find VASCERN on this website: www.vascern.eu

VASCERN is one out of 24 European Reference Networks (ERNs) for Rare and Low Prevalence Complex Diseases. VASCERN is a virtual network with focus on Vascular Diseases. VASCERN gathers 30 highly specialized multidisciplinary healthcare providers (HCPs) from 11 EU Member States in this area of expertise. VASCERN gather the best expertise to improve care in Vascular Rare Diseases in Europe. Networking, sharing and spreading expertise, promoting best practices, guidelines and clinical outcomes, patient empowerment, and improving knowledge through clinical and basic research are among VASCERN's objectives. There are specialised working groups focusing on aortic diseases within VASCERN. The European Commission and the Member States have shared responsibility for the establishment and development of ERNs, including VASCERN.

8.5 Think Aorta

You find the Think Aorta campaign on this website: www.thinkaorta.org

Statistics show that a diagnosis of acute Aortic Dissection is very difficult to make, even in an Emergency Department in a big hospital. It is often mistaken for a different, incorrect diagnosis such as a heart attack, gastritis, back or muscle pain.

The patient-led Think Aorta campaign was inspired by the tragic death of Mr. Tim Fleming from an Aortic Dissection in 2015, after he was sent home from a Dublin Hospital's Emergency Department with an incorrect diagnosis of gastritis.

Experience in the United Kingdom shows that if Emergency Departments educate staff about Aortic Dissection and lower the barriers to CT scanning (digital imaging), lives will be saved.

THINK AORTA is a patient led education and awareness campaign launched across the UK & Irelands Emergency Departments in 2018, with posters and podcast education material being sent to every single medical doctor in an emergency medicine setting.

The Royal College of Emergency Medicine in UK advise that this awareness campaign has received the highest levels of engagement of any campaign launched to date. The campaign has since been launched in the USA in January 2020 with printed THINK AORTA posters sent to hospital emergency departments to raise awareness for aortic disease. There are plans to extend the campaign to more countries across the globe.

Note 8.5.1: *The campaign THINK AORTA (United Kingdom and USA) reminds partly of AORTENTELEFON (Germany) and SOS AORTE (France) in its effort to put strong focus on the aorta for emergency medicine staff to help find the proper diagnose when the aorta is involved. The THINK AORTA differs from the AORTENTELEFON and SOS AORTE solutions in the following ways:*

- *THINK AORTA is very much a patient centred campaign. It is founded and run by patients in cooperation with medical doctors.*
- *THINK AORTA does not involve systematic solutions like SOS AORTE and AORTENTELEFON with procedures and routines for cooperation and communication between healthcare providers in a region. It is therefore faster to implement, but it is unclear if the results locally are as good as for the SOS AORTE or AORTENTELEFON concepts.*
- *THINK AORTA can easily be run as a national campaign in the emergency departments in various hospitals, in conferences and at various educations by use of printed posters and flyers, while SOS AORTE and AORTENTELEFON are strictly regional concepts.*
- *THINK AORTA can easily be translated and brought to new countries, independent of national political or financial solution for how the healthcare systems are set-up in the country.*
- *SOS AORTE and AORTETELEFON, requires adaptation to national healthcare rules and regulations, and especially to financing in a more complex way, since it is about cooperation between hospitals and healthcare providers. In tax financed healthcare systems, both systems will probably require pre-approval by healthcare politicians*

Note 8.5.2: *Perhaps the concepts of AORTENTELEFON, SOS AORTE and THINK AORTA will be even more similar in the future, as they learn from each other.*

8.6 Aortic Disease Awareness Day – September 19

Aortic Disease Awareness Day - September 19 - is a global awareness day for the rare diseases of the aorta. Events are hosted in big hospitals with good knowledge on aortic diseases, with participation of aortic disease survivors and their medical doctors, cardiac and vascular surgeons, researchers and cardiologists.

Such a meeting gives a possibility for Aortic Disease patients to meet other patients. Meeting another patient can give a very good impact on quality of life for the patients, because few people who do not have an aortic disease can fully understand what it is like to live this disease. It is also a great way to create awareness around these rare, deadly and many time familial diseases, to help us all keep the human aorta in mind when it comes to difficult diagnoses to make, and to bring light to the work done in the hospitals developing the diagnosis, treatment, research, prevention and care around aortic diseases.

The September 19 movement was founded by patients with a wish to use one day per year together to learn more about the latest findings around aortic diseases, and to meet other patients. Doing it always on the same date, allows the movement to grow globally and create a joined impact for increased awareness.

More info about September 19 and the AD Awareness Day can be found with its global steward, The Marfan Foundation.

Figure 8: Aortic Disease Awareness Day – September 19 – graphics by The Marfan Foundation

Note 8.6.1: *You can read the full story behind the Aortic Disease Awareness Day in two medical books for aortic surgeons:*

AORTIC DISSECTION, *publication date: Dec 1, 2016 by Professor Germano Melissano. Professor Roberto Chiesa. Edi.Ermes ISBN 978-88-7051-565-7, Chapter 1: Thoughts of an aortic dissection "survivor" (page 1-10)*

AORTIC COMPLEXITIES*, publication date: Dec 1, 2018 by Professor Germano Melissano. Professor Roberto Chiesa. Edi.Ermes ISBN 978-88-7051-663-0, Chapter 54: Growing a global movement of aortic disease awareness in real life. Turning theory to practice (page 491-501)*

9. If you have an aortic disease - you are not alone. Examples of ways to find others with your aortic disease.

9.1 Search the web randomly for info and for other patients

If you have a specific aortic disease, you can try to find communities with other patients by using the word of your own diagnose like "Vascular *Ehlers-Danlos Syndrome*", "*Marfan Syndrome*", "*Turner Syndrome*", or "*Loeys-Dietz Syndrome*".

You can also search with or more general words like "*aorta*", "*aortic aneurysm*", "*aortic dissection*" or "*aortic disease*". Also use such search words in your own native language.

On **Facebook** you can chose to search for **groups** or **pages** to find the patient communities, beside in the flow of posts. Try to search for hash-tags like **#aorta**, **#aorticawareness** and **#aorticdisease** on sites like:

- Google
- Facebook
- Twitter

9.2 DiseaseMaps

On www.diseasemaps.org you can find at least 9 different patient communities with aortic disease patients. Here you find others on a map. You can reach out to people close to where you live for a real-life meeting, you find information, forums and much more. You can read and post on DiseaseMaps in 16 various languages. Over 170 000 patients in the world uses DiseaseMaps for over 1 240 various rare diseases. It is free of charge to use DiseaseMaps. These are the communities with aortic disease patients up and running in January 2020 on DiseaseMaps:

- Aortic Dissection
- Aortic Aneurysm
- Marfan Syndrome
- Loeys-Dietz Syndrome
- Ehlers-Danlos Syndrome
- Turner Syndrome
- Shprintzen-Goldberg Syndrome
- Bicuspid Aortic Valve
- Takayasus Arteritis

9.3 RareConnect

On www.rareconnect.org you can find at least 3 different patient communities that have patients who have affected aortas. RareConnect also has a global map where patients can show their location to help you reach out to find people also close to where you live for a real-life meeting. It offers good information, forums and much more. You can read and post on RareConnect in 12 various languages. It is free of charge to use RareConnect. These are the communities that are up and running in January 2020 on RareConnect:

- Aortic Dissection
- Ehlers-Danlos Syndrome
- Turner Syndrome