



DEUTSCHES HERZZENTRUM BERLIN
STIFTUNG DES BÜRGERLICHEN RECHTS

Patient Information Aorta

Diseases, Diagnosis, and Therapy

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Prof. Dr. Volkmar Falk, Medical Director of the German Heart Center Berlin

Foreword

Dear patients,

Every year about 23,000 surgeries are performed on the aorta, on the main artery directly at the heart. The numbers are rising, because many diseases of the aorta do not occur until an older age and life expectancy is rising.

The German Heart Center Berlin is one of the internationally leading hospitals for aortic surgery. Continuous research and the resulting enhancement of diagnosis and therapy are key priorities of our range of services.

Nowadays, many aortic surgeries can be done in a minimally invasive procedure. Acute life-threatening illnesses of the aorta still mostly require complex open heart surgeries.

For these types of emergencies we have around the clock interdisciplinary teams of experienced and specialized surgeons, cardio anesthetists, cardio technicians, and nurses. Despite all medical advancements: the diagnosis of an illness of the aorta is connected to great fears and worries for many people.

With this patient guide we would like to provide patients, their families and people who are interested in the topic with comprehensive and extensive information: about the aorta, its illnesses and methods of preventions, diagnosis and therapy.

For any other questions please do not hesitate to contact us.

With the best wishes for your health,

Prof. Dr. Volkmar Falk
Medical Director

Dr. Stephan Kurz, MPH
Head of the research group aorta

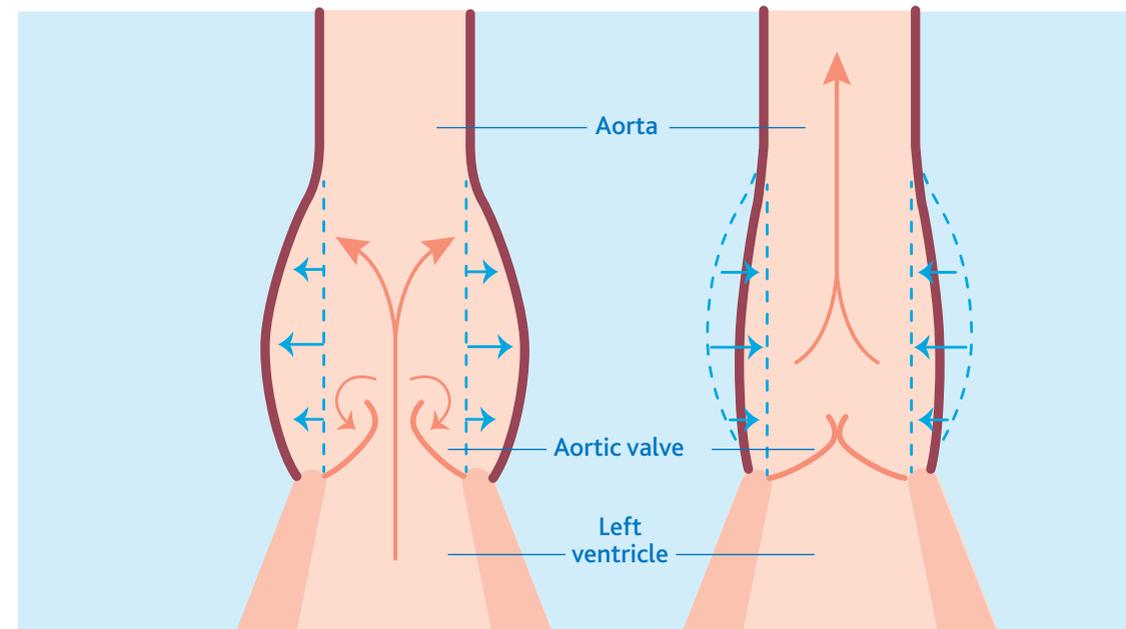
1 The aorta

1.1 Definition and function

The aorta, also called main artery, transports over a system of aortic branches oxygenated blood from the heart into the whole body. This transport is vital to provide all the cells in the body with oxygen and energy, and then carry off degradation products and carbon dioxide. Therefore almost five liters of blood are flowing through the aorta every minute.

The blood inside the body is transported by the pressure created by the heart (blood pressure). Blood flow is divided into two phases. During the ejection phase of the heart (systole) the blood volume is pumped from the heart into the body's circulation system.

During the subsequent filling phase (diastole) of the heart, the heart muscle relaxes and the so-called bagpipe function of the aorta comes into action. During the systole the elastic aorta and the blood vessels close to the heart widen, whereby their volume increases, as well as part of the blood that is ejected from the heart is being saved for a short moment. Through elastic resetting forces of the blood vessel wall, the volume of the aorta reduces again and the blood flow evens out.



Bagpipe function of the aorta

1.2 Anatomy

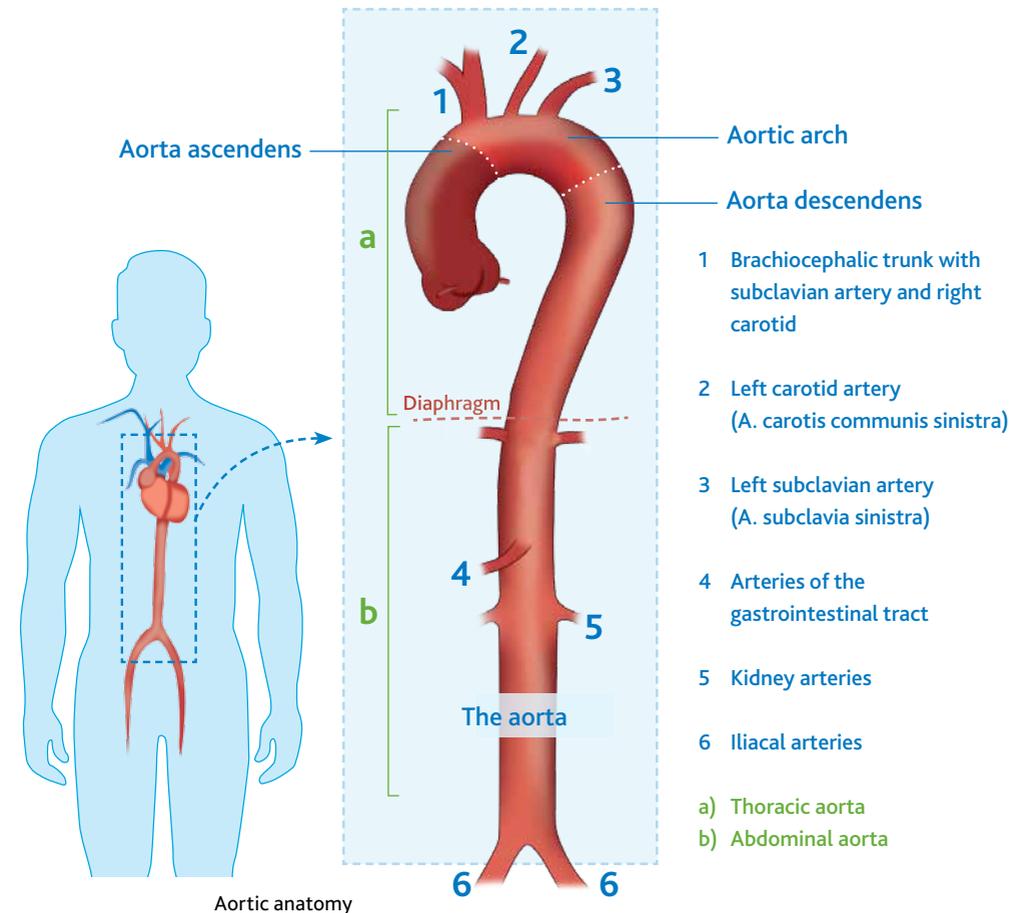
The aorta begins with the aortic root, which is separated by the aortic valve from the heart. At the heart the coronary arteries directly supply the heart muscle with oxygenated blood. Afterwards the aorta runs straight up for about five to six cm as aorta ascendens.

Afterwards it bends to the left towards the spine and creates the aortic arch. Arising from this section are the arteries that supply the head (carotids), the neck and the arms. Subsequently the aorta runs parallel to the spine as the aorta descendens until the height of the fourth lumbar vertebrae, where it splits into the two arteries of the pelvic space, the iliac arteries, and down to the legs.

The first section of the aorta – from the root to the passing through the diaphragm – is called thoracic aorta. In this area of the aorta multiple blood vessels branch off to supply organs of the chest, the head, and the arms. The subjacent part of the aorta, inside the abdominal cavity up to the partition into the iliac arteries, is called abdominal aorta. Here, all blood vessels take root that supply the abdominal organs, for example gastrointestinal tract, liver, kidneys, and spleen. Finally, the iliac arteries supply the legs with oxygenated blood.

Diameter of the aorta

The diameter of the aorta reduces throughout its length and is dependent on many factors like gender, age or BMI. The adjacent table gives an overview of standard values.

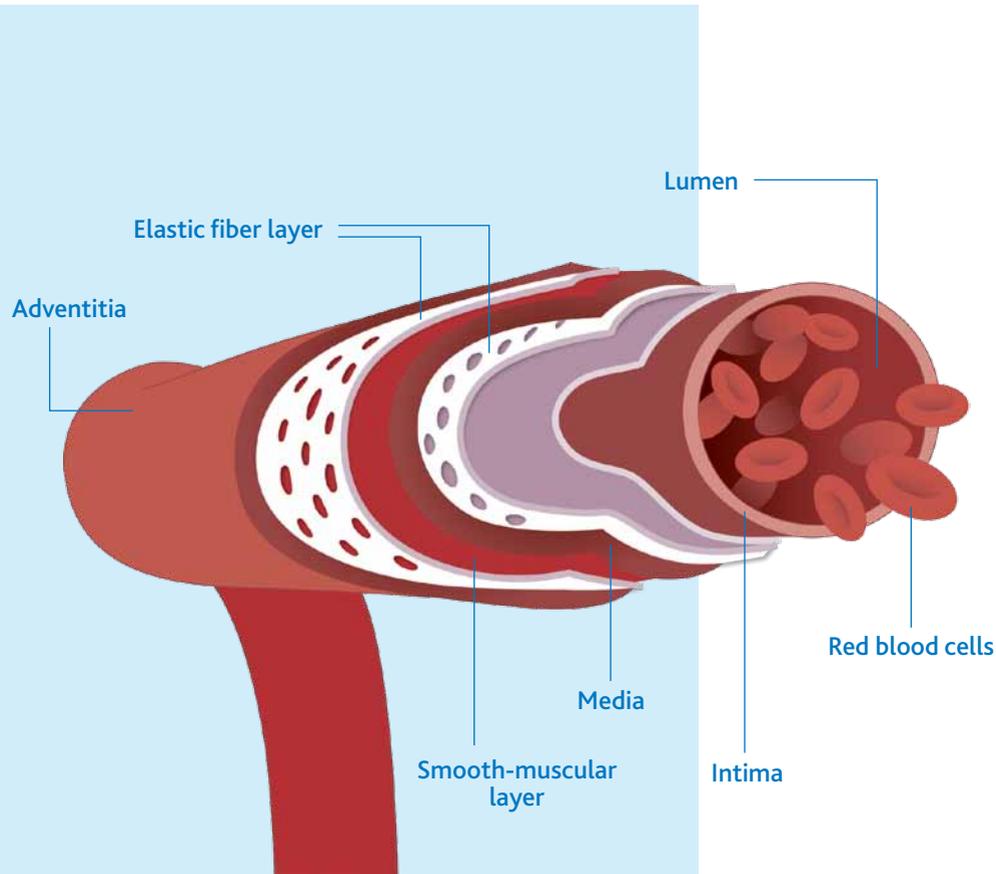


Aortic anatomy

Standard values of the aortic cross section

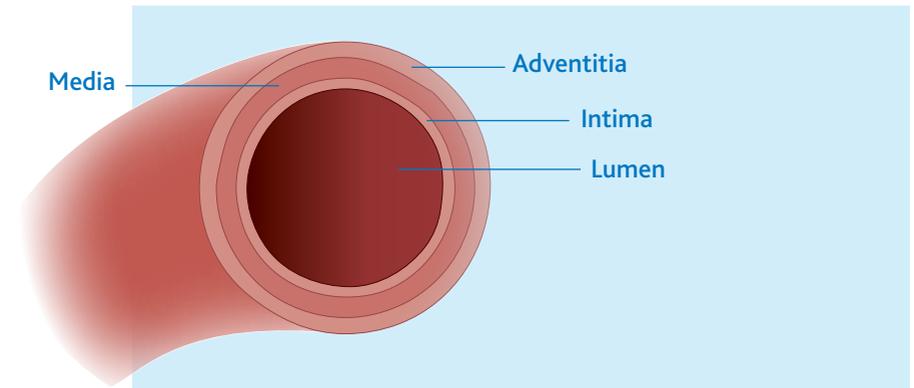
	Men	Women
Aortic root	3.6-3.9 cm	3.5-3.7 cm
Aorta ascendens	2.9 cm	2.9 cm
Aorta descendens	2.4-2.9 cm	2.5-2.6 cm

1.3 Aortic wall layers



Structure of an artery

The wall of blood vessels basically has three layers, the intima, media, and adventitia. Every layer fulfills a different function.



Wall layers of the aorta (cross section of a healthy aorta)

Intima

The inner layer is the border to the inside of the blood vessel, the so-called lumen. It consists of a single layer of endothelial cells. They control the entry of blood components into the surrounding tissue and are necessary for the exchange of oxygen and nutrition. Damage to the endothelial enhances the creation of blood clots, in order to stop a bleeding, for example.

Media

The middle and by far widest layer consists of circular arranged smooth muscle cells. Through contraction or relaxation of these muscle cells, the diameter of the single blood vessel can be changed and thereby supply organs individually according to demand. In addition this layer contains a lot of elastic and collagen fibers.

Adventitia

The adventitia, the outer layer of the blood vessel wall, consists of connective tissue and is necessary to embed the blood vessels into the surrounding tissue. Additionally, the adventitia contains tiny blood vessels, nerves, and lymph vessels, to supply itself.

2 Diseases of the aorta

There are acute (sudden onset) and chronic (slowly developing) diseases of the aorta. In the following pages, we are trying to explain the different diseases in detail.

Diseases of the aorta

Acute

Acute dissection of the aorta

Penetrating aortic ulcer (PAU)

Intramural hematoma (IMH)

Traumatic or iatrogenic aortic rupture

Acute inflammation of the aorta

Chronic

Aortic aneurysm

Chronic dissection of the aorta

Chronic inflammation of the aorta

2.1 Chronic diseases of the aorta

Arteriosclerosis

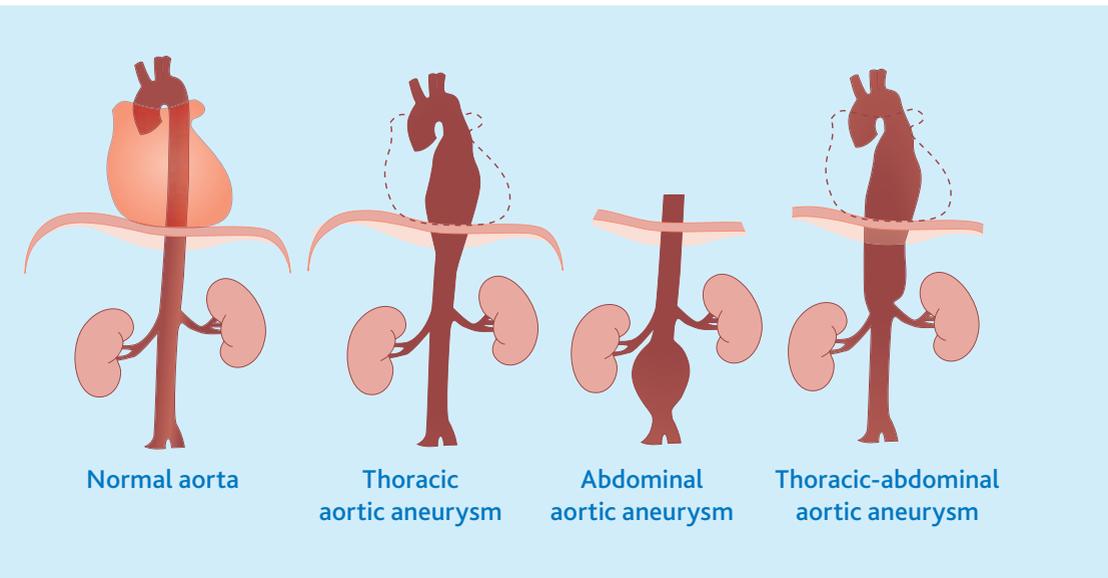
Cardiovascular diseases caused by arteriosclerosis are the main cause of death in Germany. To put it simply, arteriosclerosis describes the pathological stenosis of an artery. This constriction happens, because blood fats, blood clots, connective tissue, and calcium oxide collect on the blood vessel walls. Other clinical pictures of the aorta, like the aortic aneurysm or dissection, benefit a great deal from arteriosclerosis.

Aortic aneurysm

An aortic aneurysm leads to an enlargement of all three layers of the blood vessel walls and enlarges the diameter of the blood vessel itself. Widens the aorta within the chest, it is called a thoracic aortic aneurysm. Is the widening inside the abdominal cavity, we are talking about an abdominal aortic aneurysm. Abdominal aortic aneurysms are predominantly found under the renal arteries. If the aneurysm stretches out across the chest as well as the abdominal area then we call it a thoracic abdominal aortic aneurysm.

Different risk factors are associated with aortic aneurysm for example hypertension, smoking, and diabetes mellitus (see chapter 3). These facilitate the thickening of the aortic walls (arteriosclerosis) and thereby the development of an aneurysm.

Aortic aneurysms lead to no specific symptoms, which mean affected patients usually do not notice the aneurysm. Should it cause any symptoms, they display as back, abdominal or flank pains. In most cases, aortic aneurysms are being discovered by accident during ultrasound examinations or computed tomography (CT).



Types of aortic aneurysms

2.2 Acute diseases of the aorta

If an aortic aneurysm stays undiscovered for too long, it may lead to a rupture of the aorta at this place or bleeding between the layers of the walls (aortic dissection). To avoid complications with patients that have a known aortic aneurysm, they are advised to have it checked every six to twelve month.

If an abdominal aortic aneurysm creates back, abdominal or flank pains, it has to be reevaluated, because surgery may be needed. Even if the aneurysm does not cause any symptoms but shows a large diameter or grows fast (within one year), surgery has to take place.

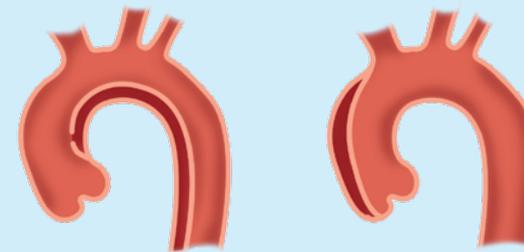
With aneurysms in the chest area (thoracic aortic aneurysm) it is similar. If such an aneurysm causes symptoms, surgery should be weighed against the risk of rupture. Even without symptoms, an aneurysm at this position should be operated upon, if its diameter succeeds 5.5 to 6 cm or its diameter increases fast.

Exact aortic diameters at which surgery is advised can be found in chapter 5.1.

Chronic aortic dissection

Like the aortic aneurysm, the chronic aortic dissection also counts to the slowly progressing diseases of the main artery. Here it is very important to control the progress of the disease on a regular basis and to see a physician immediately if new symptoms occur.

Acute diseases of the aorta, like the acute aortic syndrome, include a variety of sudden onset clinical pictures. These often occur with heavy pains and are an emergency situation. The acute aortic syndrome is sorted into five groups, which are explained in the following chapters.



Class 1

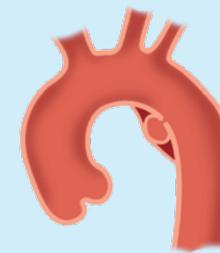
Class 2

Class 1

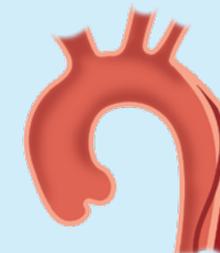
Classic aortic dissection

Class 2

Intramural hematoma
(10–25%)



Class 3



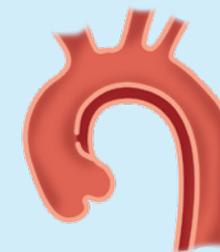
Class 4

Class 3

Implied aortic dissection
with cavity of the aortic wall

Class 4

Penetrating aortic ulcer (2–7%)



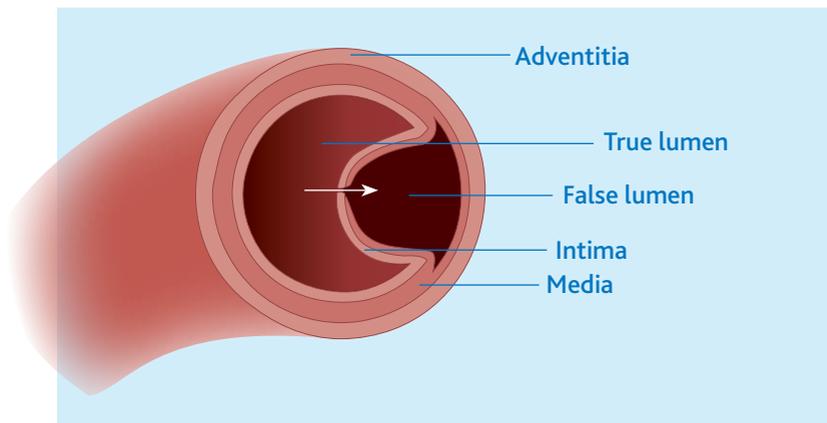
Class 5

Class 5

latrogenic or traumatic
aortic dissection

2.2.1 Acute aortic dissection

In the classic aortic dissection the inner layer of the wall (intima) of the aorta rips apart, so that blood flows into the layers underneath and "scrambles forward". Therefore the wall layers separate; next to the normal inside of the blood vessel (so-called true lumen), a new, blood-filled blood vessel is created (so-called false lumen).



Cross-section of the aorta with acute aortic dissection

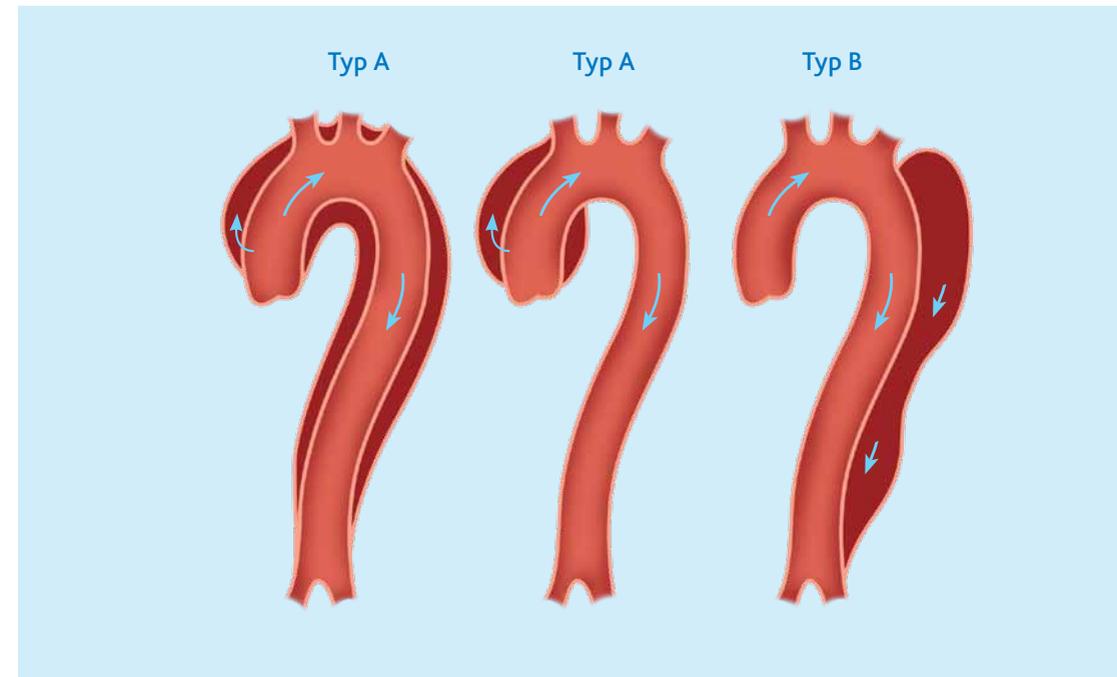
Blood vessels originating from the aorta, for example towards the head or the coronary arteries, can be constricted or no longer receive enough blood to supply their organs. This insufficient blood circulation can cause, depending on the blood vessels concerned, a multitude of symptoms.

Most of the patients report sudden acute pain, that has a ripping or piercing character. If the dissection enhances along the aorta, the pain (annihilative pain) feels like it is wandering. A multitude of factors raise the risk for the occurrence of an aortic dissection, for example, untreated high blood pressure, smoking and different connective tissue diseases (see chapter 3).

Types of aortic dissection

The aortic dissection is being classified by the location of the "scramble bleeding". A common one is the Stanford classification. It differs between type A and type B dissection.

In type A dissection the ascending part of the aorta (aorta ascendens) is affected by the "scramble bleeding". Independent of this situation, the aortic arch, the descending aorta (aorta descendens) or the pelvis and groin blood vessels may be involved. It is called a type B dissection, if the "scramble bleeding" starts after the left subclavian artery (arteria subclavia sinister) branches off.



Stanford-classification of aortic dissection

Type A-Aortic dissection

The acute type A dissection is a life-threatening situation that needs immediate surgical intervention at a cardio surgical center. The mortality rate within the first 48h is high (1-2% per hour). Depending on the localization of the dissection, heart attacks, strokes, paralyses or insufficient blood supply of the inner organs can be the consequence.

The symptoms can resemble other, more often occurring diseases, like a heart attack. Though common risk factors, like previous illnesses or surgeries on the aorta, but also current results, like a difference in blood pressure between the arms in combination with the typical pain characteristics lead to the diagnosis of an acute aortic dissection.

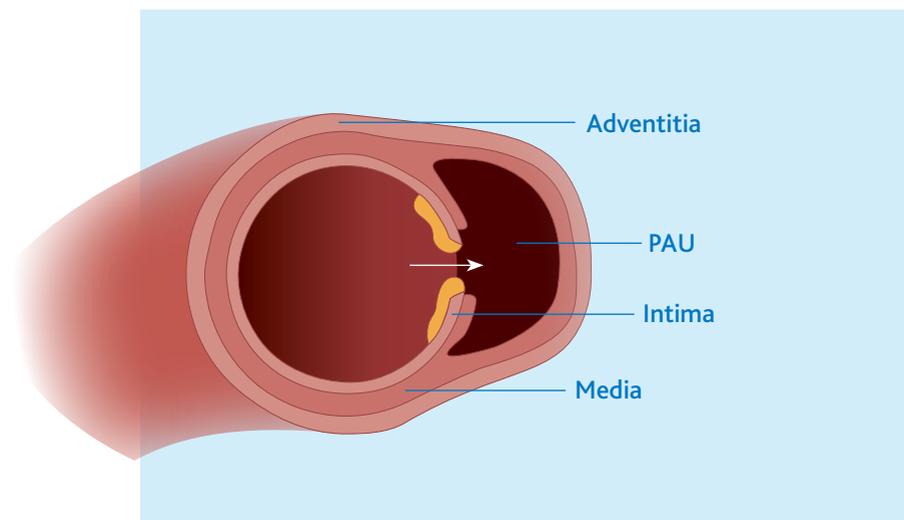
If the outer layer of the aorta cannot withstand the pressure of the "scramble bleeding" and rips as well, an aortic rupture occurs. This leads to death in most cases. Also, bleeding into the heart sac is a feared complication, because it constricts the heart and prevents it from beating effectively (a so-called pericardial tamponade).

Type B-Aortic dissection

Type B dissection is rarer than type A dissection. The risk of a sudden rupture is less; therefore you have more time to do a specific diagnosis. The surgical intervention usually is by stent implantation.

2.2.2 Penetrating aortic ulcer (PAU)

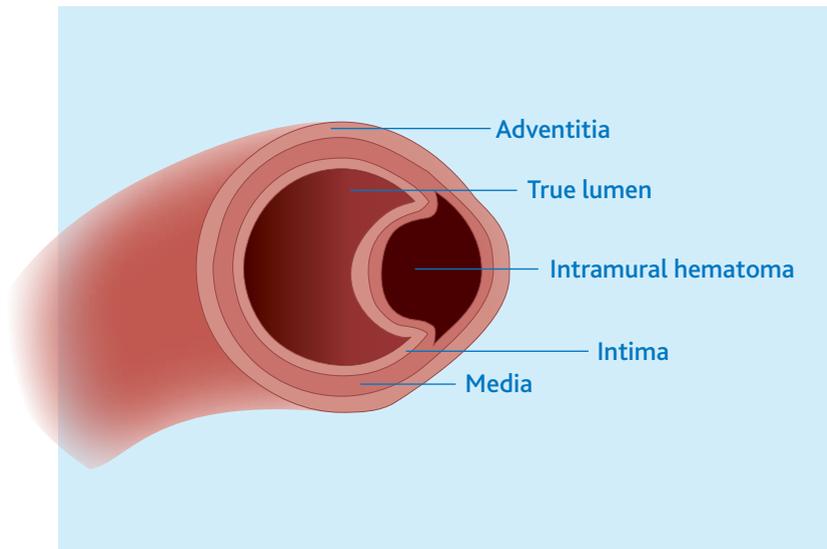
The penetrating aortic ulcer (PAU) is a type of ulceration of the aorta on the base of an arteriosclerotic blood vessel wall. The ulcer penetrates the single layers of the aortic walls and leads to a localized, about 2-centimeter big protrusion of the aorta. Untreated a PAU can lead to an aortic dissection, an aortic aneurysm or an aortic rupture.



Cross-section of the aorta with a penetrating aortic ulcer (PAU)

2.2.3 Intramural hematoma (IMH)

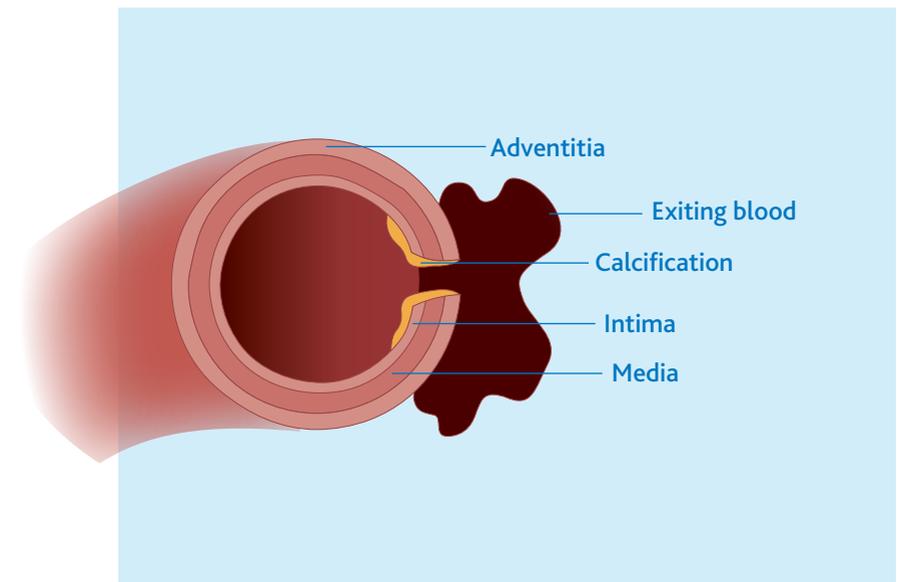
It is called an intramural hematoma (IMH) if there is a bleeding into the blood vessel wall without a rupture or a false lumen (see chapter 2.2.1). The causes of an IMH are not clearly known. It is assumed that small blood vessels that supply the aorta (so-called vasa vasorum) rip apart and cause the bleeding into the blood vessel wall. The intramural hematoma is also seen as a complication of a PAU (see chapter 2.2.2) or discussed as a traumatic event. It is possible that an IMH recedes back spontaneously. Although in some cases it can lead to an aneurysm or a ruptured blood vessel wall.



Cross-section of the aorta with intramural hematoma (IMH)

2.2.4 Traumatic aortic rupture

A traumatic aortic rupture is often caused by a sudden and forceful, indirect external force. Common accident mechanisms, for example, are a frontal impact during a car crash or a fall from a great height. Thereby single layers of the blood vessel walls or the whole aorta wall can rupture and blood can leak into the surrounding tissue.



Cross-section of the aorta with traumatic aortic rupture

3 Risk factors for aortic diseases

Personal factors and habits, anatomical or heretical particularities and other accompanying diseases can elevate the risk to develop an illness of the aorta.

3.1 General risk factors

People between the ages of sixty and ninety are generally most affected. The risk of an aortic dissection is double as high in men as in woman. An untreated high blood pressure (hypertension) over a longer period of time raises the possibility for the appearance of an aortic aneurysm or an aortic dissection. This is also true for factors that in a long term change the appearance of a blood vessel and facilitate the development of arteriosclerosis. Arteriosclerosis hardens the walls of a blood vessel, which makes it rigid and brittle. Supporting factors are for example dyslipidemia, diabetes mellitus, and smoking.

3.2 Special living situations

Under certain circumstances an aortic dissection can occur during pregnancy. The risk is higher if there is a primary disease. In individual cases healthy women can be affected. Demanding strength and endurance training, especially weight lifting with high intensity, can cause a fast rise in blood pressure under strain which can lead to an aortic dissection. Also, the consumption of cocaine is linked to the risk of an aortic dissection.

3.3 Anatomical anomalies

Anatomical anomalies of the heart or the blood vessels can increase the risk of an aortic disease or complication.

Bicuspid aortic valve

About 1% of all newborns are born with a bicuspid aortic valve. This anatomical anomaly has a familial predisposition. Men are two times more affected than women. The aortic valve normally consists of three parts. Two of the three folds melted into each other and therefore people with a bicuspid aortic valve have two differently sized folds. This change in structure leads to early calcification, thickening, and tightening of the aortic valve. The ascending aorta that follows after this aortic valve is often thin-walled. All these factors raise the risk for the occurrence of aortic aneurysms and dissections.

Aortic stenosis

Congenital stenosis of the aorta can show a great bandwidth of symptoms. The narrowing can have different degrees. The aortic isthmus stenosis has a congenital narrowing of the aortic arch or rather the end of the aortic arch. Within 7% of all congenital heart diseases and up to 20% of all patients with Turner-syndrome this anomaly occurs. The mid-aortic-syndrome is a rare disease, which is described with genetic diseases (Alagille-syndrome, Williams-Beuren-syndrome). In 60% of the cases, it occurs without a palpable reason (idiopathic). The mid-aortic-syndrome is also described as abdominal aortic isthmus stenosis, because the abdominal aorta and its main branches are narrowed. Before and after the stenosis, an expansion and weakening of the aortic wall (aortic aneurysm) can occur. In relation with high blood pressure, the risk for an aortic dissection rises.

3.4 Diseases with genetic predisposition

Diseases of the connective tissue that are hereditary produce a structural changed collagen and elastin, which weakens the three-layer wall construction of the aorta. The consequences are a stretched and thinned aortic wall (aneurysm), tears (dissection) or a complete rupture of the blood vessel wall.

Marfan-syndrome

The Marfan-syndrome affects men and women. Every year one in 3,000 children is born with this hereditary connective tissue disease. The genome mutation concerns the FBN1-genome, which causes a structural weak version of the protein fibrillin-1 to be produced. The connective tissue, which is formed by this protein, is essentially weaker as in humans without this genetic predisposition. This connective tissue weakness primarily affects joints, tendons, the eye, and aortic walls. Over time 95% of affected people develop diseases of the heart valves (aortic valve insufficiency), thoracic aorta aneurysm or aortic dissection. Not just the change of the aorta itself raises the risk of aortic complications. Also anatomical change of the ribcage (funnel chest, chicken breast) and of the spine (kyphoscoliosis) can lead to an aortic dissection by a change of compression force and traction.

Turner-syndrome

The Turner-syndrome is a hereditary disease, which leads to a loss of function or total loss of one X chromosome. It only affects women. Yearly one in 3,000 born girls carries this genetic predisposition. The manifestation of symptoms can differ depending on the genetic results, and may start with stunted growth, deformities of the kidneys and absent puberty. The risk for aortic illnesses (aortic aneurysm, dissection, ruptures) is raised because of the genetically malformed blood vessels. Patients with Turner-syndrome more often have bicuspid aortic valves (see chapter 3.3).

Ehlers-Danlos-syndrome

The Ehlers-Danlos-syndrome describes a group of hereditary connective tissue diseases that can affect men and women. About one in 50,000 children are born every year on earth with the genetic predisposition. It is about a sum of genetic defects, which concern the collagen metabolism. Collagen is a structural giving protein that as a faulty version leads to a typical hypermobility and hyperextensibility of the joints. Skin, as well as blood vessels, are extremely vulnerable. 4% of these patients experience aortic dissection or rupture, which can occur spontaneously without prior appearance of an aortic aneurysm.

Loeys-Dietz-syndrome

One in 100,000 children is born yearly with this genetic predisposition. The Loeys-Dietz-syndrome occurs through mutation in two genomes. The clinical symptoms are similar to the Marfan-syndrome, without the eyes being affected. The skin is as vulnerable as in Ehlers-Danlos-syndrome patients. Characteristic is a cleft palate or rather a cleft uvula (uvula bifida). The deformities within the vascular system are extensive. In the whole body, dilated or distended arteries run in parts, in which aneurysm can occur. Typically twined arteries appear, especially around the neck. 95% have an aneurysm close to the aortic root.

3.5 Inflammation

Vascular inflammation (vacuities) of the aorta (aortitis) can be caused by autoimmune processes of rheumatoid diseases (noninfectious aortitis). Rarely do they occur because of a bacterial infection (infectious aortitis). Because of the inflammation, the walls of the aorta thicken and aneurysms can occur, which raise the risk of an aortic dissection.

Aortitis caused by rheumatoid diseases

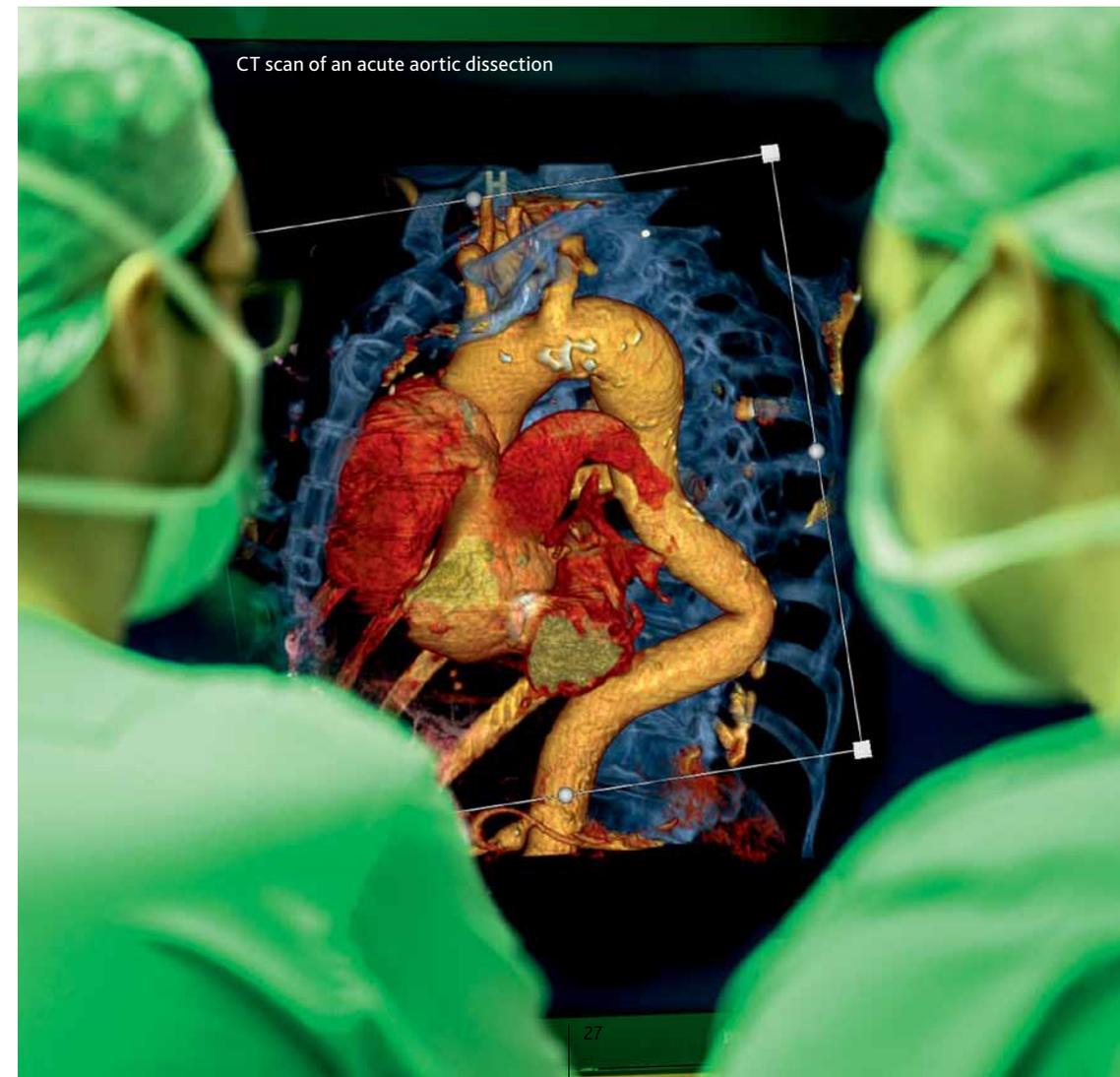
Giant-cell arthritis appears mostly in the blood vessels of the head, especially within the temporal arteries. With 15% of the patients, the aorta is also affected. Yearly about three people in 100,000 are taken ill with it. Therefore the giant-cell arthritis is the most common vacuities with humans above the age of fifty. Women are three times more likely to be affected than men. The Takayasu arthritis is a very rare autoimmune vacuity, which mostly affects the aortic arch with its branching great arteries. In Germany, one person of one million people is affected by this disease, mostly women under the age of forty.

Bacterial aortitis

Infectious vacuities of the aorta mostly occurred in the past as a complication of syphilis infection (*treponema pallidum*), before the discovery of an effective antibiotic therapy made this disease a rarity. People with a healthy aorta in general do not fall ill with bacterial aortitis. It is special circumstances that raise the risk of an infectious inflammation of the aorta:

- **Patients with an acquired immunodeficiency disease or a congenital immunodeficiency**
- **Spreading of an infection that started outside of the aorta (endocarditis, spondylitis, tuberculosis)**
- **Infection of the aortic prosthesis**

The diagnosis of a bacterial aortitis can be a challenge. Physical complaints can start slowly or suddenly. Besides a general feeling of illness, bouts of fever and chills can alternate with symptom-free intervals.



CT scan of an acute aortic dissection

3.6 Reducing risk factors

Which risk factors impact to what extent, differs individually. The better you know them, the better you are able to control them. The most important strategies as an overview:

Blood pressure

If you have been diagnosed with high blood pressure (hypertension), trust yourself to change the necessary habits. Stay in contact with your physician. Use your appointments to ask questions. If you have concerns or think that you are not tolerating any of your medications, please bring that up.

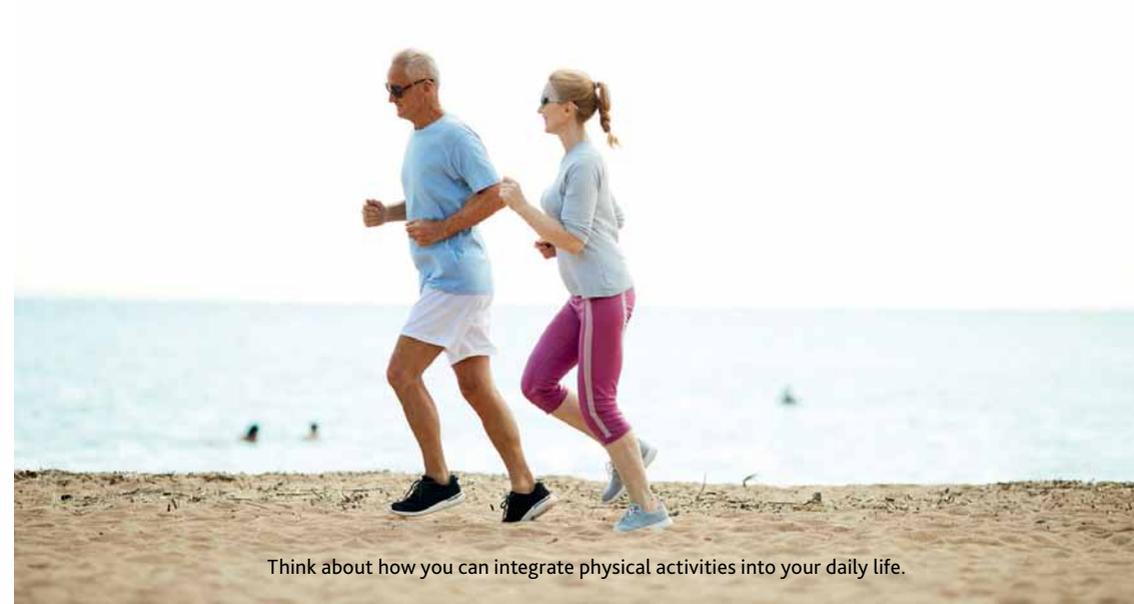
Nutrition and exercise

You make life easier for your blood vessels if you keep your weight within the norm and increase your physical fitness. It is less important how you start, but that you start at all. Find out how you can change your habits so that you can be one step closer to a balanced nutrition and how to integrate physical activity into your regular schedule. In order for the changes to your lifestyle to be a challenge you can manage, pick small, short term, and controllable tasks.

Nicotine

If you smoke, find out how much significance it has in your life:

- **How did you start smoking?**
- **What does nicotine do for you?**
- **What made you take breaks from smoking?**
- **How do you feel about the recommendation to quit smoking?**
- **How do you see your chances of being a non-smoker?**



Think about how you can integrate physical activities into your daily life.

Change is a field of tension, in which not only your motivations to stay a smoker and our motivations to advise you to quit smoking have an effect. There is always the other side, in which often lies the key for change. You smoke because it gives you an advantage in your living situation. Maybe it reduces stress? Or is a part of spending time with friends? When you give up nicotine, you will have to confront challenges that may be difficult or unpleasant. There are a lot of assistant programs, which can support you.

Safety

Buckle up when you drive a car. The seat belt reduces the risk of massive trauma to your chest.

Teamwork

Discuss the interval between appointments and the possible necessity of drug therapy or surgical intervention together with your physician. Dare to ask your questions. Please let attending physicians of other medical fields or your dentist know, if you or anyone in your family has had an aortic dissection, connective tissue disease or anatomical anomalies of the heart. It may happen that you will be advised to have direct relatives examined.

4 Diagnosis of aortic diseases

There are different possibilities to examine the aorta and display it visually. Depending on the aortic disease and therapeutic treatment, additional examinations might be necessary. Subsequently, we are going to explain some of these examinations.

Echocardiography

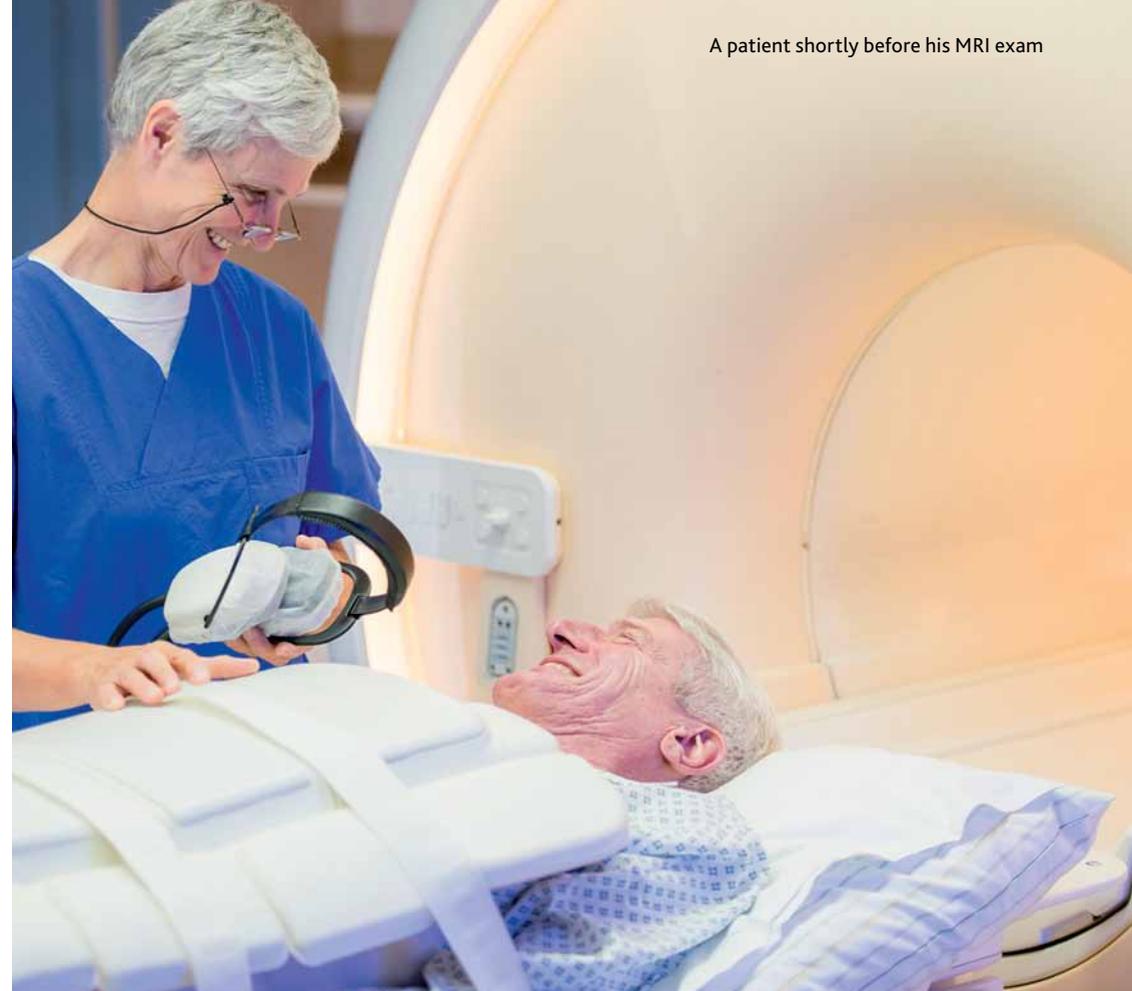
Echocardiography is an ultrasound examination of the heart. Hereby multiple information about the heart, for example about the pump function, heart valves or ascending aorta, can be found out.

During the transthoracic echocardiography (TTE) the sonic head is placed from the outside onto the chest. This method is often available and poses no risk for the patient. During the transesophageal echocardiography (TEE), the heart is being examined from the inside through the esophagus. Therefore parts of the hearts, like the atria, can be assessed better. Concerning aortic surgery, the TEE is the tool of choice to control certain parts and functions of the heart and aorta before and during surgery.

Senior consultant Dr. Jan Knierim during an ultrasound of the heart



A patient shortly before his MRI exam

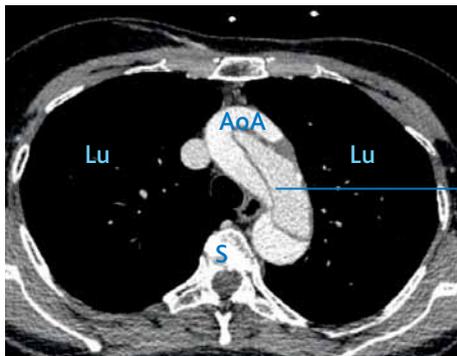


Magnet resonance imaging (MRI)

During magnet resonance imaging (MRI) cross-sectional images are created with the help of a strong magnetic field. You can assess soft and nerve tissue especially well. In contrast to a CT scan the patient is not exposed to any radiation. Because the examination is relatively long, it is not usable in an emergency situation. Also, patients with metal in their bodies, like prostheses or pacemakers, cannot undergo the procedure because of the strong magnetic field.

Computed tomography (CT)

Computed tomography is based on the use of x-rays and produces many cross-sectional images like an MRI. The patient is lying inside a rotating x-ray tube. The computer calculates from many two-dimensional x-rays a three-dimensional complete image. CT is the choice when it comes to diagnosing aortic diseases. It is much faster than an MRI and therefore can be used in emergency situations. In order to better visualize blood vessels (e.g. the aorta), a contrast agent can often be applied. This examination is called a CT angiography.



Dissection membrane
(split wall)

Aortic dissection at the aortic arch (layered x-ray image through the chest; in order to see the blood vessels/aorta better, a contrast agent was used; AoA: aortic arch; Lu: Lung; S: Spine)

X-ray

The classic x-ray examination of the chest offers a good overview of the bony structure, the lung tissue, and heart size. If possible, the examination takes place standing up and from two sides. The aortic arch can be viewed to a limited extent; big aortic aneurysm can often be discovered. Otherwise, this examination puts emphasis on the assessment of the lung tissue.

Positron emissions tomography (PET-CT)

The positron emission tomography uses radioactive markers to measure metabolic activity within the tissue. It is also a cross-sectional examination and can be fused with the CT examination. In cardiology, a PET is used to find out about the vitality of the heart muscle. Aortic surgery uses radioactive sugar markers to create an image of an infection of the aorta or prosthesis. Inflamed tissue has a higher metabolism and uses more sugar, which leads to more radioactive materials to collect at those sites.

Heart catheter examination

During heart catheter examination the heart and its coronary arteries can be inspected. Therefore a tube (catheter) is mostly inserted into the groin and carefully advanced towards the heart. Using x-rays and contrast agents, single blood vessels and possible obstructions can be seen. During catheterization a stent can be implanted to reopen an obstructed blood vessel, a so-called heart catheter intervention. The examination plays a main role in the diagnosis and therapy of coronary heart disease (CHD) and acute coronary syndrome (heart attack/ infarction). Often patients with an aortic disease also suffer under CHD, this is the reason why this examination is done frequently.

Lung function examination

The lung function examination (spirometry or LuFu) judges the lung volume as well as air flow speed. Different lung diseases can be evaluated, for example, COPD (chronic obstructive pulmonary disease). Concerning aortic surgery, the lung function examination plays a part in assessing possible long term ventilation during an extended intensive care stay.



5 Therapy of aortic diseases

In many cases of diagnosing an acute aortic disease, immediate surgical intervention is necessary. But it is not uncommon that aortic diseases develop slowly and sometimes even without symptoms over a long period of time, and therefore only need – after they have been discovered – strict supervision and avoidance of the risk factors. Whereas the ascending part of the aorta will only be “conventional” operated upon (“open-surgery”), treatment of the descending aorta with “interventional” methods (TEVAR; thoracic endovascular aortic repair) is the standard.

5.1 Aortic aneurysm

When an aortic aneurysm is diagnosed, normal blood pressure levels should be tried to be reached (if necessary with medication). Is the diameter of the aorta under the suggested range for surgery, regular checkups become as important as the optimal blood pressure adjustment. Also reducing risk factors (e.g. stop smoking) can be advised, because it slows down the growth of the aneurysm.

Aorta ascendens

An important role for a decision regarding the treatment plays the diameter of the aneurysm. In case of an aneurysm of the ascending aorta, surgical intervention is advised in case of a diameter of 55cm or a rapid increase in aortic diameter (over 10mm per year). If risk factors, for example a genetic predisposition, a rapid increase in size, a weakness of the aortic valve or the wish for pregnancy are the case, then surgical intervention is advised at an aortic diameter of 45mm.

Patients with connective tissue diseases (e.g. Marfan-syndrome) are advised to have the ascending aorta replaced at a diameter of 50cm. In an open chest surgery (open-surgery), a blood vessel prosthesis (stent), partly with aortic valve prostheses, is implanted.

Aortic Arch

An aneurysm of the aortic arch should be operated upon if the diameter exceeds 55mm. If adjoining sections of the aorta are also affected, the aortic arch can be replaced by a stent at a smaller diameter.

Descending aorta

In the case of aneurysms of the descending aorta that require immediate treatment (diameter 55cm), a stent can be placed by means of minimally invasive technique (TEVAR; thoracic endovascular aortic repair). One condition is that the aneurysm has enough distance to the branching kidney and pelvic vessels. Big aneurysms under these circumstances may not be suitable for this kind of procedure. In this case, an open-surgery has to take place. Also in cases of connective tissue diseases, a conventional surgical intervention is necessary. Thereby a tube graft or y-graft, depending on the location of the aneurysm is going to be implanted.

5.2 Aortic dissection

Concerning aortic dissection, the location of the dissection plays a major part in the decision of the treatment. With type A dissection, a fast surgical intervention is advised. It is an acute emergency situation that without a surgical intervention has a high rate of mortality. The combination of blood pressure therapy, pain management, and immediate surgical intervention is the key to the patient's survival.

In case of a type B dissection, we differentiate into two groups: an "uncomplicated" and a "complicated" form.

An "uncomplicated" type B dissection does normally not need surgical intervention. Instead, the blood pressure, pain, and heart rate are treated by medication. Also by reducing the risk factors, the advancement of the aneurysm is tried to be countered. Important are regular examinations by a physician (including imaging), in order to quickly intervene in cases of relevant change of the aorta.

With the "complicated" type B dissection, the pain is ongoing. The blood pressure is also hard to manage. Link to this is a fast growth in diameter or even a rupture of the aorta. Adding to reducing the blood pressure, a minimally invasive treatment with a stent implantation (TEVAR) is advised. If this treatment is not possible, open-surgical treatment is necessary.

5.3 Requirements for aortic surgery

Heart-Lung-Machine

For surgery on the ascending aorta or the aortic arch, the blood flow needs to be redirected during surgery. A heart-lung machine is used for this situation. Two important facts:

- 1. Vital organs, mainly the brain, have to be supplied with oxygen-rich blood during the surgery.**
- 2. The field of surgery has to be empty of blood.**

Cardiac arrest

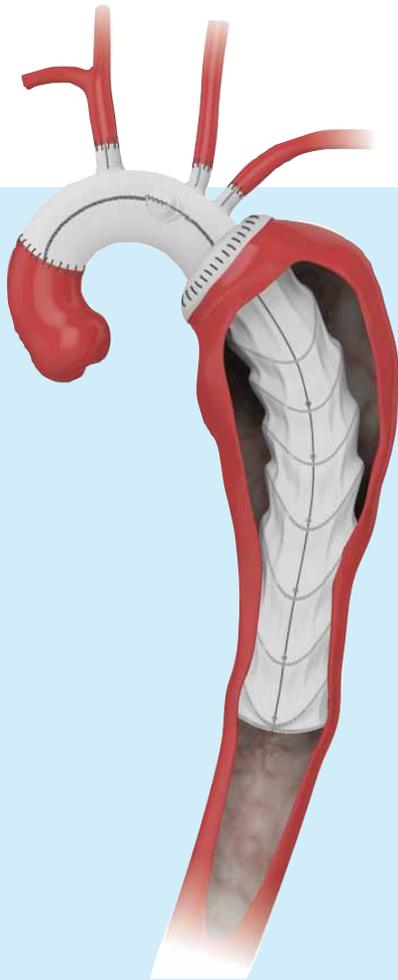
During the surgery, the heart stops to beat under controlled circumstances. An "inoperative" heart uses less oxygen for the heart muscle cells. Therefore the surgeon has a time window for the treatment of the aortic disease. The pump function of the heart during this time is replaced by the heart-lung machine.

Hypothermia

A second method to reduce the need for oxygen of the cells is a so-called hypothermia, the cooling of the body temperature down to 28°C. Not only do the heart muscle cells benefit from this, but also the brain cells, that react with a lower oxygen consumption to the reduced oxygen supply.

Selective antegrade brain perfusion

In order to better protect the brain cells from being damaged by oxygen deprivation (neuroprotection), the procedure of selective antegrade brain perfusion exists. The extracranial arteries are being supplied by oxygenated blood from the heart-lung machine.



Aortic arch prosthesis
made of polyester



Biological aortic valve replacements made from animal tissue (left), mechanical prosthesis made of metal and plastic

5.4 Surgical techniques

Aortic stent/ prostheses

The affected area of the aortic aneurysm is being replaced by a stent. Depending on location, this may happen interventional (TEVAR) or open-surgical. The stent is an elastic pipe made of synthetic fibers.

Aortic valve reconstruction and replacement

The complexity of aortic dissection is measured to what degree the aortic valve and the coronary arteries are involved. It is always important to keep as many endogenous structures as possible. Therefore reconstruction of the aortic valve is preferred. If the function of the valve cannot be restored by the body's own structures, it has to be replaced. Hereby mechanical or biological heart valves can be used.

Mechanical heart valves last very long, but the patient will have to take blood thinners for the rest of their lives. Biological heart valves are mostly made from cattle pericardium tissue or pork heart valves. With this type of valve prostheses, the patient only has to take blood thinners for the first few weeks after surgery. But this valves do not last indefinitely and have to be replaced within ten to fifteen years.

6 Aftercare

The aftercare is of big importance following an aortic surgery and depends on the type of aortic disease. Subsequently you will read what you need to look out for after having surgery on the aorta.

6.1 Rehabilitation

The rehabilitation after aortic surgery is typically divided into three phases:

- **Early rehabilitation starts in the hospital and includes early mobilization and physiotherapy.**
- **The subsequent cure (in German called Anschlussheilbehandlung = AHB) in case of aortic and heart surgeries takes place as inpatient treatment, but (if required) can also happen as an outpatient treatment. Movement therapy, physiotherapy, psychosomatic guidance (e.g. reducing fears), and health education (e.g. controlling risk factors) are part of the program.**
- **Afterwards reintegration (work and non-work) follows. This relates to a slow rise in workload and strategies to reduce and avoid stress.**

6.2 Regular checkups

Patients with an aortic disease need lifelong care independent of how the disease is treated (medication, intervention or open surgery). The frequency and extent of these examinations depend on the type of intervention and the aortic disease. The aftercare during the first year serves the purpose of confirming the success of the therapy as well as quickly discovering and removing possible complications, connected to the intervention.

For the aftercare, clinical examinations and imaging procedures (e.g. echocardiography, CT-angiography, etc.) are necessary. Also, regular visits with a family practitioner are advised.

6.3 Life after aortic surgery

Driving a car

Within the first six weeks after the surgery, you should completely forego driving a car, bike or any other vehicle yourself. Because the surgery happens on the chest, special attention should be paid to the breastbone (sternum), to treat it gently. Even as a passenger you should always wear a safety belt.

Medication

One of the most important pillars in the therapy of aortic aneurysm and dissection next to surgical intervention is medication. Risk factors for the heart and cardiovascular system, like high blood pressure (hypertension) or high blood sugar levels should be treated and closely monitored (see chapter 2.3.6). Depending on the type of aortic disease and its treatment, a lifelong therapy with antiplatelet drugs might be necessary. A single dose of ASS 100mg daily is recommended. In special cases an anticoagulant like Marcumar or Falithrom might be indicated.

Lifestyle

Because aortic aneurysms and dissections are occurring as a result of arteriosclerotic connective tissue changes, all risk factors need to be reduced or avoided after surgical intervention of the aorta (see chapter 2.3.6). Smoking and high blood pressure are the most important factors.

Physical activities and sports can be restarted about three months after surgery or rather after the bony healing of the sternum is done. To avoid blood pressure peaks, a steady workout should be preferred. Activities with constant strain are for example swimming, biking, jogging, and walking. Weight lifting should only be done with moderation.

After an aortic intervention, the sexual activity is not impaired. Although excessive physical strain should be avoided in order to prevent high blood pressure.

Traveling and Wellness

In face of the size of the intervention and the following aftercare bigger journeys should be planned the earliest three months after the surgery took place. Make sure you have enough of your medication as well as carry copies of your documents with you. These include physician's letter, prostheses pass, and so on. Be very careful, especially within the first months after surgery, that you avoid heavy lifting and carrying (e.g. suitcases). Here, too, avoid physical strain. Stays at high altitudes up to 2,000 meters above sea level are commonly not a problem. Because of wound care and healing, you should wait two to three months before you visit a sauna or swimming pool.



Work life

After successful therapy, an inability to work usually for three months or rather until the bony healing process is finished, is in place. In the course of time, depending on the practiced profession and degree of recovery, employability will be assessed. Was an aortic disease completely cured by surgery, life within the community is possible almost without any impairment; therefore no disability is caused by the aortic surgery. Big, still existing aortic aneurysms and chronic aortic dissections can lead to impairment in (work) life and can constitute as a disability.

Genetic examination

Aortic aneurysms and dissections can be connected to genetically caused, congenital connective tissue diseases or rather syndromes. They can differ greatly in manifestation. If a human genetic examination is indicated, must be decided individually. They are recommended for relatively young patients with aortic disease, because consequences can occur for their aftercare as well as for close relatives.

The Marfan Center at the Charité and German Heart Center Berlin (DHZB) arose from twenty years of specialized Marfan consultation hours of the human genetics at the Charité and the heart surgery at the DHZB. A new legal regulation (paragraph 116b SGB V) for the outpatient treatment of rare diseases, like the Marfan-syndrome, allows, starting 2009, to offer a combination of this complex care and treatment under one roof. Experiences and knowledge of the physicians are bundled and ideally, patients can take care of all necessary appointments at the Marfan Center with minimal time effort.



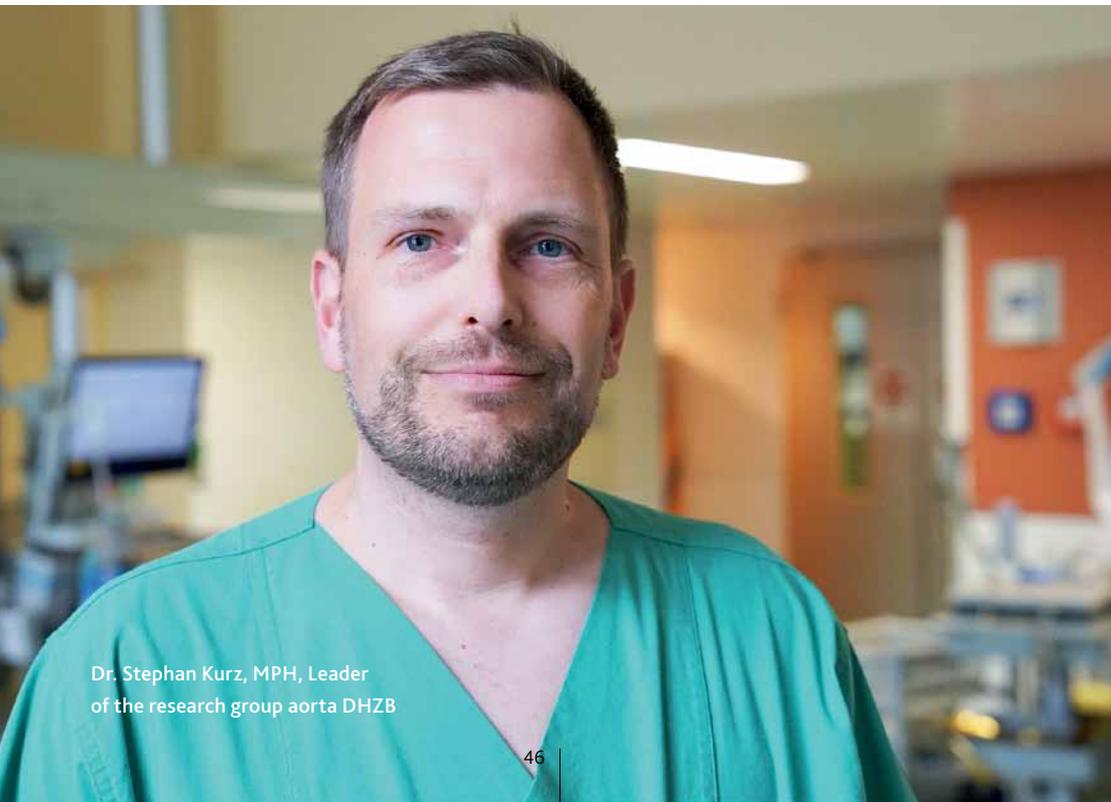
Prof. Dr. med. Christoph Starck, Leading Senior Physician with an aortic prosthesis



The team of the surgical outpatient clinic at the DHZB

7 The aortic telephone at DHZB

The German Heart Center Berlin is the center with the highest level of experience and numbers of yearly interventions in the Berlin/ Brandenburg region when it comes to the treatment of acute aortic dissection. With a catchment area of six million citizens, this is a huge responsibility. The pre-clinical management of aortic emergencies is an important component in the care of the affected patients. There is a lot to be organized until the patient arrives at the German Heart Center Berlin. Especially for these situations, a centralized hotline for aortic emergencies has been set up. The team of the aortic telephone is always occupied by experienced specialists and they give important information to the assigning physician when it comes to treating the aortic emergency and organize the process until surgery can take place.



Dr. Stephan Kurz, MPH, Leader
of the research group aorta DHZB

8 Outpatient clinic and consultations

Aortic-consultation of the cardio surgical outpatient clinic at the DHZB

Phone +49 30 4593-2002

Marfan Center of the Charité and the DHZB

– Outpatient consultation for patients with genetic aortic diseases.

Phone +49 30 450 665 356

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Thank you for your support:

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