A Simple Guide to Thoracic Aortic Surgery
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Dear Reader,

Many patients are uncomfortable with the recommendation to undergo aortic surgery. With this booklet, we would like to

- explain the most important facts about aortic diseases
- answer frequently asked questions
- reduce your anxiety
- improve your individual treatment

Approximately 84,000 heart operations are performed annually in Germany with the help of a heart lung machine. In 7,100 of these operations part of the thoracic (chest) aorta is replaced. There is a tendency for an increase in the number of aortic interventions. This is rooted mostly in the ageing population and increased life expectancy.

An abnormal enlargement of the aorta (aortic aneurysm) is the most common cause for its surgical replacement. Today, if well planned, this can often be done with minimally invasive techniques and generally, with reduced risk.

Often, aortic disease is unfortunately only recognised when serious complications arise. In particular this includes acute aortic dissection whereby the layers of the aortic wall tear. In the worst case, this may lead to a rupture of the aorta. In these cases it is necessary to undergo emergency surgery.

Acute aortic disorders are associated with a high death rate. Therefore, their prevention is of great importance. Regular check-ups play a large role in the treatment of patients with aortic diseases.

In the event that it is necessary to replace the thoracic aorta, careful preparation and planning of the surgery significantly reduces risk.

New aortic prostheses and hybrid approaches enable surgeons today to work on longer or hard to reach segments of the aorta during a single procedure.

Intra-operative management has significantly improved in the last ten years. This includes, for example, the protection of organs during aortic replacement surgery (so-called organ protection).
Also post-operative intensive care has continuously improved in recent years.

Since the introduction of aortic surgery in Germany, the Department of Cardio-thoracic, Transplantation and Vascular Surgery at the Hanover Medical School under the leadership of Prof. Hans Georg Borst and Prof. Axel Haverich has developed into a nationally and internationally renowned center for aortic surgery.

The continuous improvement in surgical techniques and reduction of surgical risk was and continues to be the principal research focus of the Hanover aortic surgeons.

As a result, revolutionary surgical techniques like the “Elephant Trunk” (1983) or the “Frozen Elephant Trunk” (2003) could be developed here. These have since become routine procedures throughout the world to treat patients with complicated diseases of the thoracic aorta.

We hope this booklet familiarises you with the topic of aortic disease.

Yours sincerely,

Malakh Shrestha

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The aorta

The anatomy of the aorta

The **aorta** is the main artery out of which all arteries making up the circulatory system originate.
The aorta begins at the aortic root (1) where the origins of the coronary arteries and the aortic valve are found.

The coronary arteries branch off at bulges in the aortic root called sinuses of Valsalva.

Above the aortic root, the ascending part of the aorta begins (ascending aorta) (2). This is followed by the curved aortic arch (3). The arteries that supply the head, neck and arms branch off from here.

The descending part of the aorta (descending aorta) (4) is found after the aortic arch. The inter-costal arteries and other arteries diverge from here to supply the spinal cord and other body parts with blood.

Together, the ascending aorta, aortic arch and descending aorta make up what is called the thoracic aorta (a).

The abdominal aorta (b) is found below the diaphragm. The vessels that supply the gastrointestinal tract, spleen, liver and kidneys split off from here. The lumbar arteries supply the vertebral column and spinal cord.

At the height of the forth lumbar vertebra, the aorta splits and becomes the pelvic arteries (iliac arteries) (5). These supply the pelvic organs and merge into the leg arteries.
Anatomy of the aorta

Normal sizes of the aorta

A healthy aorta is widest at the aortic root and narrows gradually until it divides into the pelvic arteries. A normal aortic diameter is $<40\text{mm}$. The aorta is typically larger in men than in women.

The aorta transports about 200 million liters of blood over the course of an average life span and is continuously subjected to arterial blood pressure. To endure this constant strain, the aorta, like all organs, undergoes a continual adaptation and repair process.

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The aortic wall has three layers

The innermost layer is the **intima**, followed by the **media** and last the outermost **adventitia**.

The **intima** is composed of the so-called endothelium, a cohesive single layer of flat cells. It controls, among other things, oxygen and gas exchanges between blood and the vessel wall. Injury to the endothelium enables clot formation (thrombus). An embolism occurs when a thrombus is swept up in the blood stream.

The **media** consists primarily of smooth muscle cells in the shape of rings. They regulate the vessel width and ensure a steady blood flow through their elasticity.

The **adventitia** is a mesh of connective tissue fibers that anchors the vessel into its surroundings. The so-called "vasa vasorum" are tiny vessels that run through the adventitia to supply it with blood.

The **lumen** is the vessel’s blood filled channel.
Diseases of the aorta

Overview of aortic diseases

Aortic diseases can be divided into **chronic** aortic diseases and **acute** aortic syndromes. Chronic aortic diseases can trigger an acute event. Acute aortic syndromes typically lead to chronic aortic diseases.

### Chronic aortic diseases:
- Aneurysm
- Chronic dissection

### Acute aortic syndromes:
- Aortic rupture
  - Open rupture
  - Closed rupture
- PAU (aortic ulcer)
- Aortic dissection
  - Type A
    - with malperfusion
    - without malperfusion
  - Type B
    - with malperfusion
    - without malperfusion

### Possible causes:
- Atherosclerosis
- Congenital connective tissue diseases
- Inflammation of the aorta
  - Infection
  - Autoimmune diseases
- Previous aortic dissection (chronic dissection)
Aortic aneurysms

An aortic aneurysm is the enlargement of the aorta to more than 1.5 its normal size. As a general rule, this is a diameter of about 40 mm.

The most common cause of an aortic aneurysm is atherosclerosis, which initiates the calcification and weakening of the aortic wall and is associated with chronically high blood pressure.

Other diseases that can lead to an aortic aneurysm are congenital connective tissue diseases, an inflammation of the aorta as a result of an infection or autoimmune disease or a previous aortic dissection.

Frequently, aortic aneurysms do not cause complaints. Often aortic aneurysms are detected by chance during routine tests like echocardiography (ultrasound of the heart) and x-ray.

An aortic aneurysm in the ascending aorta can also lead to leaking of the aortic valve. The resulting complaints (e.g. reduced stamina, heart failure) are sometimes the first symptoms for the patient.

Aortic aneurysms can develop in the chest and abdominal areas. Patients who have already been treated for an aortic aneurysm have an elevated risk of developing aneurysms in other untreated areas. Regular check-ups are, therefore, essential.

Only when the aortic diameter becomes very large will symptoms arise in neighboring organs (e.g. esophagus, wind pipe and vocal cord). This is rarely the case.
Aortic diseases

Examples of aortic aneurysms

- Aortic root aneurysm
- Ascending aortic aneurysm
- Aortic arch aneurysm
Aortic diseases

- Descending aortic aneurysm
- Thoracoabdominal aortic aneurysm (TAAA)
- Mega aorta syndrome
Acute aortic syndromes

The term acute aortic syndrome describes a sudden event concerning the aorta, which is typically coupled with extreme pain. It might be accompanied by collapse, shock or unconsciousness. Another possible symptom is a malperfusion syndrome (a disturbance in the blood supply to the organs). Acute aortic syndromes usually result from aortic aneurysms.

The larger the aorta, the thinner the aortic walls and the higher the blood pressure, the greater the risk for an acute aortic syndrome.

Aortic rupture

The term aortic rupture describes the bursting of the aortic wall. The result is the leakage of blood into the surrounding tissue. If large amounts of blood penetrate a cavity in the body, such as the pericardium or the chest cavity, this can lead to immediate death.
If the layers of tissue are able to stem the leakage, this is called a "**contained rupture**". The blood loss is slowed and those affected can sometimes reach the hospital in time.

An aortic rupture is an **acute emergency**, which must be **treated immediately**. The risk of death from an aortic rupture is extremely high. It is assumed that only 40 % of those affected reach the hospital alive. Of these, 20-30 % die despite emergency treatment.
Aortic diseases

PAU (Penetrating Atherosclerotic Ulcer)

An aortic ulcer is caused by atherosclerosis and causes localised damage to the inner aortic wall allowing blood to escape into the outer layers of the aortic wall. An ulcer is usually smaller than 2cm and looks like a bulge in imaging procedures. An untreated aortic ulcer can lead to an aortic dissection or rupture and should, therefore, be treated immediately.
Aortic dissection

An aortic dissection is a **cleavage in the aortic wall’s three layers**. In contrast to an aortic rupture, blood does not flow out of the aortic wall but instead swells up inside the wall layers, lifting the innermost layer, the intima, away from the aortic wall.

As the dissection advances, a new channel for blood flow is created. This is called a "**false lumen**". The original vascular channel is called the "**true lumen**". The "false lumen" usually develops in the direction of the blood flow.

An acute aortic dissection is expressed in a sudden event of characteristically **sharp or tearing pain in the chest, back or abdominal areas**. As the dissection advances, the pain can **travel** from, for example, the chest to the back to the sides and groin.

An aortic dissection can block the vessels leading out of the aorta causing a heart attack, stroke, paraplegia, acute malperfusion syndrome (disturbance in the blood supply) of the arms or legs, or insufficient blood supply to the abdominal organs.
A sudden discrepancy in the blood pressure of the extremities in connection with a painful event points towards an aortic dissection. After an aortic dissection, the blood is contained only by the thin outer wall layer (adventitia). A complete aortic rupture (see page 16) becomes a possible complication.

Categorising aortic dissections with the Stanford Classification
An acute aortic dissection type A is one of the most serious emergencies in vascular surgery. The term "Type A" means that the ascending aorta is affected. This is the case in about 65% of aortic dissections. The "Type A dissection" can be limited to the ascending aorta or can extend to the descending aorta, the abdominal aorta or even the pelvic and groin vessels.

A particular risk of a "Type A dissection" arises from the possible involvement of the aortic valve, coronary vessels and vessels to the head. This can lead to sudden death through a rupture (see page 16) and bleeding into the pericardium, the sack enclosing the heart (pericardial tamponade). Blood trapped inside the pericardium compresses the beating heart from the outside. This leads to heart failure.

As a result of the potential for complications, an untreated "Type A dissection" has a high mortality rate of about 40 – 60% in the first 48 hours (around 1% per hour!).
In contrast to a "Type A dissection" a "Type B dissection" is a dissection in the descending aorta and begins by definition behind the opening of the left arm artery (subclavian artery). This accounts for about 25% of patients with an aortic dissection. There is a lower risk for serious complications in comparison to a "Type A dissection" because the heart and vessels to the head aren’t affected. However, the abdominal and leg vessels can become obstructed or the aorta can tear causing bleeding.

If an aortic dissection also obstructs a vessel, preventing adequate blood supply from reaching the respective organ, this is called "malperfusion syndrome". Malperfusion syndrome is one of the most common causes of death from an aortic dissection. Even after successful emergency surgery, organ damage caused by malperfusion syndrome is often irreversible. If malperfusion syndrome is survived, there is still the threat of long-term damage such as chronic kidney failure or the effects of a stroke.
Congenital connective tissue diseases

Many connective tissue diseases that involve the aorta lead to the early development of aneurysms (see page 13). Patients with these diseases, in turn, are more likely to experience the complications associated with aneurysms (dissection and rupture) (see pages 16 and 19). As a result these patients require special preventive care. When aneurysms develop, a timely surgical treatment must take place to prevent complications.

The congenital connective tissue diseases affecting the aorta can be generally categorised into genetic diseases (e.g. Marfan syndrome, see page 23) and developmental defects that are congenital but not classically inherited (e.g. bicuspid aortic valve, see page 25). This is important for medical consultations with family members.

A portion of inherited aortic diseases cannot be explained through known genetic mutations. In recent years numerous mutations have been discovered that explain these inherited aortic diseases. However, they are unique in that aortic diseases inconsistently affect patients with these mutations. It is important that all family members are given continuous preventive care in the case of a positive family history.
**Marfan syndrome**

Marfan syndrome is the most common inheritable genetic connective tissue disease (about 1-2 in 10,000 people). It is inherited as "autosomal dominant", which means 50% of descendants are affected and it can occur in both men and women. A mutation in the FBN1 gene, which is responsible for the production of the protein "Fibrilin 1", causes Marfan syndrome. The defective protein leads to a "connective tissue weakness" that alongside joints, tendons and eyes, above all affects the aorta.

**Typical features of Marfan syndrome**

- Overlong limbs, a slender, taller body build (marfanoid habitus)
- Stretchy joints, soft skin
- Breastbone pointing outward (pigeon chest) or inward (funnel chest)
- Changes in the backbone (e.g. scoliosis)
- Eye lens dislocation (ectopia lentis)
- Nearsightedness
- Diseases of the heart valve, heart failure
- Aortic aneurysm, aortic rupture

Aortic diseases are of special importance to Marfan patients because they can lead to early death if left untreated, as a result of the risk of dissection or rupture.
Ehlers-Danlos syndrome

Ehlers-Danlos syndrome describes a group of inherited connective tissue diseases, which have in common stretchy skin (about 1-2 in 10,000 people). Ehlers-Danlos syndrome can be triggered by mutations in the genes that make the protein, collagen. This is the most important connective tissue protein in the human body.

Clinical consequences involve the aorta and great vessels, internal organs and above all the skin. Aortic dissections and ruptures (see pages 19 and 16) can occur without being preceded by the development of an aortic aneurysm.

Loeys-Dietz syndrome

The rare Loeys-Dietz syndrome (<1 in 1,000,000 people) was first described in 2005. Like Marfan syndrome, it is an autosomal dominant inheritable disease, which is triggered by mutations in two genes (TGF-beta receptor type I and II).

Both men and women can be affected. 50% of descendants inherit the disease. It manifests itself through connective tissue weaknesses, which involve the aorta and great arteries. Patients develop an aortic aneurysm (see page 13).

Clinically, Loeys-Dietz syndrome resembles Marfan syndrome (see page 23), which is why, before it was first identified, many patients were diagnosed as Marfan patients. A typical feature is a "split uvula", a congenital indentation in the uvula. As opposed to Marfan syndrome, patients with Loeys-Dietz syndrome experience almost no effect on their eyes.
Bicuspid aortic valve

Normally the aortic valve consists of three leaflets or cusps (tricuspid aortic valve). A bicuspid aortic valve has in contrast only two functional cusps. Most often patients are born with three cusps, two of which are fused together creating one large functioning cusp. The extent to which two cusps are fused varies and in mild cases may not be detected by an ultrasound of the heart.

The bicuspid aortic valve is the most common congenital heart defect and affects about 1% of people. It affects men more than women (about 2:1). It can be inherited. Blood relatives should be examined as well.

The flawed aortic valve leads to premature degeneration (thickening, calcification). A resulting increasing narrowness or leakiness of the aortic valve can develop in individuals as early as the 4th to 6th decade of life as opposed to in old age.

It is now known that this developmental defect is not limited to the aortic valve. In patients with a bicuspid aortic valve, a connective tissue dysfunction often affects the ascending aorta. The aorta usually has a thin wall and is prone to aneurysm development. The risk for aortic complications is raised.
Aortic diseases

Aortitis

Aortitis describes the **inflammation** of the aorta through an **infection** or an **autoimmune disease** (noninfectious aortitis). An aortitis is connected with a thickening of the aortic wall. The inflammation can be detected through several tests (e.g. PET-CT = positron emission tomography, MRI = magnetic resonance imaging).

**Infective aortitis**

In the past infective aortitis was often caused by **untreated syphilis** (a sexually transmitted disease caused by the treponema pallidum bacterium). The first patients to receive thoracic aortic surgery in the 40s and 50s of the last century were mostly young patients with syphilis. The possibility for antibiotic therapy has made syphilitic aortic aneurysms in central Europe uncommon. These days, patients who develop an aortic infection often have **serious underlying diseases** (e.g. immunodeficiency). Also, infections in neighboring body parts can spread into the aorta. This includes, for example, infections of the vertebral column (spondylitis) in older patients, which can compromise the descending aorta. Infective diseases of the aorta have a high risk of aortic rupture (see page 16). **Intensive treatment with antibiotics** is necessary.

**Surgery** to remove the infected tissue is complicated and risky, but in many cases, the only treatment option. There is a high risk of reinfection of the newly implanted material. The use of biological materials (e.g. donor vessels, so-called homografts) can reduce the risk of reinfection. Increased calcification of the donor vessel, however, can necessitate treatment at a later point.

Similar to other aortic diseases, symptoms of infective aortitis can emerge slowly or suddenly and can be accompanied with fever, chills or infection signs from laboratory tests. It is common for the disease to progress gradually with symptom-free intervals followed by recurring attacks of fever. As a result, diagnosis is difficult.
Giant Cell Arteritis

Giant cell arteritis is the most common rheumatic vessel inflammation in people over 50 years of age (1-15 in 100,000 people, frequency increases with age). Women are considerably more often affected than men (about 75%).

The inflammation leads to a thickening of the vascular wall. This can result in narrowing of the vessel. Mainly the large arteries of the head, especially the temporal arteries, are affected, which is why giant cell arteritis is also called temporal arteritis.

In 15% of patients, the aorta is affected. Here the inflammation leads to the development of aneurysms (see page 13). Patients are typically treated with immune suppression (e.g. cortisone) to reduce the inflammatory process.

Takayasu arteritis

Takayasu arteritis is related to giant cell arteritis. It is a rare autoimmune disease (< 1 in 1,000,000 people). It generally affects women under 40 years of age.

In Takayasu arteritis the inflammation of the great arteries leads primarily to a malperfusion syndrome (a disturbance in the blood supply to the organs).

Aside from the vessels to the head, the abdominal vessels can be affected. Bypass surgery may be necessary. However, as a result of the underlying disease, the bypasses have a high risk of becoming blocked over time.

Aneurysms (see page 13) can develop when the aorta is involved.
Techniques to evaluate the aorta

The diagnostic pathway begins with a doctor’s consultation and physical examination. If an aortic disease is suspected, follow-up is carried out for clarification purposes. The imaging procedure used depends on the anticipated disease.

Transthoracic Echocardiography

This is an ultrasound of the heart and its surrounding blood vessels. The examining doctor holds an ultrasonic transducer to the patient’s chest. This test delivers valuable information on the heart valves, pumping action of the heart, the aortic root and the ascending aorta.

The advantages of this test are its broad availability, the ease of use as well as the absence of radiation. However, echocardiography does not offer information on the aortic arch or the descending aorta.
Transesophageal Echocardiography

A transesophageal echocardiography is also an ultrasound of the heart. Here, an ultrasonic transducer is guided into the esophagus. Some find this procedure uncomfortable, so patients are often given temporary anesthesia.

Transesophageal echocardiography allows a more precise examination of the heart including the atria and part of the descending aorta. The aortic arch cannot generally be viewed.

Magnetic Resonance Imaging (MRI)

Magnetic resonance imaging (MRI), like CT examinations, produces numerous cross-sectional images of the examined body part. Indeed, MRI has no basis in x-rays but instead uses magnetic fields. Because the procedure takes longer, it is not used in emergency situations. MRI enjoys increasing popularity in clarifying diseases before surgery and in follow-up due to the lack of radiation exposure. However, it must be taken into account that the image quality and with it the information gained by the doctors usually cannot compare to a CT examination.

It is furthermore worth mentioning that this test cannot be used with patients with metal implants (e.g. pacemaker) because of the use of magnetic fields.
Computed Tomography (CT)

Computed tomography (CT) is the main technique for clarifying aortic diseases – in elective as well as in emergency situations. This test uses radiation waves, however not in one dimension like traditional x-ray images. The patient is wheeled on a table through a tunnel. A camera rotates around the body. This allows a multitude of x-ray images to be produced, which a computer converts into a series of cross-sectional images. In order to draw detailed conclusions about the aorta, the examination is combined with the administration of a dye or contrast agent (so-called CT angiography).

Computed tomography offers a high-resolution examination for every part of the body. Extremely precise conclusions about the aorta and the exiting blood vessels can be drawn. Today a CT examination can be completed within a matter of minutes.

Because radiation exposure can potentially cause genetic mutations, this test should only be used when the indication justifies it. Before contrast agent is used kidney function should be checked and the patient should be asked whether he has experienced allergic reactions against contrast agent in the past.
In general, the CT examination is a low-risk procedure. In examinations of aortic diseases the benefits greatly exceed the risks.

Through the high information value of the images of aortic diseases and its immediate accessibility, computed tomography is the most important test to depict the aorta before an operation as well as during follow-up.

If frequent follow-up examinations are expected and particularly with young patients, it is recommendable over the long term to use MRI (see page 29) as the imaging technique in order to avoid radiation.
Diagnostics

Conventional X-ray

The conventional chest x-ray is typically taken from two views (the back and the side) while the patient stands. The organs are projected one on top of the other. This is a suitable overview test to determine the sizes of the lungs and heart. The aorta is covered in part by the heart. Large aortic aneurysms are occasionally accidentally detected on an x-ray. However, they cannot be evaluated in detail with this test, which is why a CT or MRI typically has to follow.

Normal (left), aneurysms of the descending aorta (middle) and ascending aorta (right)
A PET is a **nuclear medicine examination**, which uses small amounts of a radioactive substance (tracer) to observe metabolic processes. A PET-CT combines the detection of metabolic processes with the cross-sectional images of a CT. The metabolic activities can be precisely matched to their location in the body.

A PET-CT can show the organs’ energy expenditure through radioactive labeled sugar. Because an inflammation uses a lot of energy through inflammatory cells in the tissue, a PET-CT can reveal inflammations in the aorta or aortic prosthesis.
Additional tests

If surgical treatment of an aortic aneurysm is indicated, pre-operative examinations will be planned. Among other things, important risk factors (e.g. a lung or kidney disorder) are screened. Moreover, associated heart diseases, which would need to be treated during surgery, are excluded.

Cardiac catheterisation

Cardiac catheterisation is performed by a cardiologist and is based on x-rays and the administration of a contrast agent (dye). General anesthesia is not necessary. The examination proceeds with a puncture in a groin vessel or an arm artery under local anesthesia. From here, very thin wires or catheters can be channeled into the heart. The procedure is painless. The right heart can be examined via a vein and the left heart, an artery.

The arterial "left heart catheterisation with coronary angiography" serves to exclude coronary heart disease (coronary artery calcification). A thin catheter is led to just before the openings for the right and left coronary arteries and a contrast agent is injected. Under x-ray fluoroscopy, the flow of the contrast agent in the coronary arteries can be directly observed.

Normalbefund einer linken und rechten Herzkrankzarterie
Narrowing in a blood vessel (stenosis) or anatomical anomalies can be detected. If major narrowing of blood vessels is discovered, this will usually be treated with bypasses during aortic surgery.

Echocardiography

If echocardiography was not performed in the preliminary stages to assess a part of the aorta and the aortic valve, it is performed before aortic surgery especially to assess the aortic valve and the pumping capability of the heart (see transthoracic echocardiography and transesophageal echocardiography, pages 28, 29).

Pulmonary function test

The pulmonary function test (spirometry) examines lung volume (the amount of air lungs can hold) as well as resistance in the air passage. Lung diseases like COPD and pulmonary fibrosis can be detected.

Lung function is important for the time spent on ventilation and during post-operative treatment. Reduced lung function hinders respiratory training and increases the risk of pneumonia or a lengthened stay on a ventilator in intensive care.
Indications

Some aortic diseases develop gradually over a longer period of time and can be conservatively monitored. Only by an escalation of the disease is surgery necessary. Conversely, other aortic diseases must be immediately treated with surgery.

Aortic dissection

The localisation of the section affected by an aortic dissection plays a decisive role.

A so-called "Type A dissection" (see page 20) must be surgically treated as soon as possible. This is an acute emergency situation, which, without surgical treatment, is associated with a high mortality rate.

A so-called "Type B dissection" (see page 21) can be "uncomplicated" or "complicated".

An "uncomplicated Type B dissection" is typically treated conservatively — in other words non-surgically.

Blood pressure, pulse rate and pain must be treated with medication. Patients with this disease must be regularly followed-up with imaging examinations to timely identify rapid enlargements in the aorta’s diameter.
A "complicated Type B dissection" is marked by, for instance, persistent pain, unmanageable blood pressure, a rapid enlargement of the aorta, poor perfusion of the organs or an aortic rupture.

For "complicated Type B dissections" a minimally invasive surgical treatment using "stents" (TEVAR) is recommended.

An open surgical therapy can, however, be considered when stenting is not possible.

Aneurysms of the ascending aorta or the aortic root

An aneurysm can appear in various areas of the thoracic aorta. If the ascending aorta is affected, surgery should be performed when the vessel diameter reaches 55 mm. If heart surgery is performed for other reasons, the ascending aorta should be treated alongside if its diameter has reached 45 mm.

There are various diseases, which have a high risk of an aortic complication. In these cases, the aorta should be operated on earlier.

It is recommended that patients with connective tissue diseases of the aorta (e.g. Marfan syndrome, see page 23) receive a surgical replacement of the ascending aorta as soon as the diameter reaches 50 mm.

If there are further special risk factors, like family history, a rapid growth, leaking of the aortic valve or the desire to start a family, surgery should be performed when the aorta’s diameter reaches 45 mm. Patients with a so-called bicuspid aortic valve (see page 25) are recommended to undergo surgery of an aneurysm of the ascending aorta when the diameter reaches 55 mm. When special risk factors are present, surgery should be performed when the diameter reaches 50 mm.
Aneurysm of the aortic arch

An aneurysm of the aortic arch should be operated on when the diameter of the aorta is **55 mm or larger**. If an operation on a bordering section of the aorta is planned, the aortic arch can sometimes also be replaced even if the diameter is **smaller** than 55mm.

Aneurysm of the descending aorta

A replacement of the descending aorta with **minimal invasive** techniques (stent, TEVAR) should occur when the vessel diameter reaches **55 mm**.

If a minimal invasive treatment is not possible, an **open surgical** treatment, however, should be only performed when the diameter reaches **60 mm**. An open surgical treatment is preferred for patients with connective tissue diseases.
Aortic aneurysms do not as a rule cause complaints.
Surgically treating an aortic aneurysm is a preventive measure to avoid aortic complications.

The generally formulated recommendation for surgical treatment of aortic aneurysms is based on a risk-benefit assessment:
The risk of an operation may not exceed the risk not to operate. The graphic to the left makes clear that the risk of an aortic complication grows disproportionately with increasing aortic diameter. Beginning with the diameter sizes given above, the rule holds true that the benefit of operating outweighs the risk. Because not all patients have the same surgical risks, the indication for surgical therapy of an aortic aneurysm must always be assessed individually.
Technical requirements for surgery

Heart-lung machine

Operations on the ascending aorta and the aortic arch are carried out with a heart-lung machine. It takes over the patient’s blood circulation and the supply of oxygen. It is connected to the heart’s right atrium and the aorta. Blood flows into the heart-lung machine, where it is enriched with oxygen, cooled or warmed and pumped back into the patient’s aorta. The lungs and heart are thereby circumvented. In this way, operations on "open", non-beating hearts can be carried out.

The blood is thinned with the help of heparin so that it does not clot in the heart-lung machine’s tubes. In aortic surgery using a heart-lung machine, the surgeon tries to seal off the segment being replaced with clamps (cross-clamping). After suturing in a vascular prosthesis, the blood flow is again released into the treated segment.
**Circulatory arrest**

Because of the aortic arch’s location and vessels that originate here, it cannot be cross-clamped for replacement. For an operation of the aortic arch, the heart-lung machine must be **turned off** for a short amount of time. Circulatory arrest then ensues. After removing the aneurysms and suturing in the vascular prosthesis, circulation is again started via the heart-lung machine.

To avoid damage to the organs due to lack of oxygen, various techniques are used to protect the organs (see the following techniques).

**Hypothermia**

**Lowering the body temperature** (hypothermia) reduces the cellular need for energy and, as a result, for oxygen. Cooling the body can therefore be used as a technique to protect organs from a lack of oxygen. This has been used since the 1960s in interventions in the aorta, in particular to **protect the brain** (so-called Neuroprotection), the least tolerant organ for oxygen shortage.

Cooling the entire body to temperatures **less than 20°C** is achieved indirectly with the heart-lung machine, which cools the circulating blood. After completing the aortic replacement under circulatory arrest and hypothermia, the body is again warmed with the heart-lung machine.

**Selective antegrade cerebral perfusion**

To support neuroprotection, the technique of selective antegrade cerebral perfusion was developed. By this is meant that during circulatory arrest only the brain vessels (=selective) in the normal direction of the blood flow (=antegrade) are supplied (=perfusion) with blood from the heart-lung machine.

In this way, a continual influx of nutrients and oxygen is maintained and an extension of the period of “safe circulatory arrest” is achieved. Because the abdominal organs react with less sensitivity than the brain to circulatory arrest, the lower body temperature under circulatory arrest can be raised (25-28°C) when cerebral perfusion is used.
Cerebrospinal fluid drain

If an operation of the **descending aorta** or the **thoracoabdominal aorta** is necessary, the arteries that supply the spinal cord with blood will be affected by the surgery. Many small arteries originate in these parts of the aorta - the intercostal arteries in the chest and lumbar arteries in the abdomen. The largest arteries are sutured into the aortic prosthesis. Not all branches, however, can be sutured in. This can lead to a malperfusion syndrome of the spinal cord. As a result the spinal cord swells up. However, it is trapped in the vertebral column’s spinal canal thus intensifying the malperfusion syndrome. There is the threat of paraplegia.

If the fluid suspending the spinal cord, is drained, the pressure on the spinal cord can be eased, blood flow improved and **damage to the spinal cord avoided**.

Before an aortic operation on the descending aorta, a cerebrospinal fluid drain is inserted under local anesthesia in order to drain the fluid and to measure the fluid pressure. This is a **thin plastic catheter**, which is connected to a pressure gauge and a reservoir bag.
Aortic prostheses

Aortic prostheses are made out of polyester. This is a high quality synthetic fibre that is woven into tubular shaped vascular prostheses. The prosthesis is designed with small pleats to provide flexibility to fit the patient’s anatomy. Because the woven material is not leak proof, it is sealed with collagen or gelatin.

There are many sizes and variations of aortic prostheses produced. The simplest prostheses are straight grafts. Complex prostheses have side branches for attaching vessels, a portal for the heart-lung machine or a section with a stent graft (so-called hybrid prostheses).

The surgical team in Hanover, Germany in collaboration with Vascutek, a producer of medical devices, has already developed an aortic prosthesis with all of these characteristics (Thoraflex Hybrid prosthesis, see aortic arch replacement, page 50).

Composite aortic prostheses are still produced today by hand. Sometimes several thousand stitches are necessary to suture an aortic prosthesis together. Production can take up to eight weeks.
Surgical techniques

Aortic valve reconstruction

By the end of the 1950s, techniques to reconstruct the aortic valve were already in use. The advantage over valve replacement using mechanical valves lies in the lack of a need for a long-term blood thinner (e.g. with warfarin) and in the reduced vulnerability of valve infection. An aortic valve reconstruction is only possible when the valve cusps show no serious structural changes (e.g. calcification).

Aortic valve reconstruction is suited for treating enlargements of the aortic root that are associated with leaking of the aortic valve. The most commonly applied technique for aortic valve reconstruction is the so-called David procedure or aortic valve reimplantation. It was first introduced in 1992 by Prof. Tirone David.

The entire ascending aorta is removed but for a small rim above the aortic valve. The coronary arteries are detached from the aorta as so-called buttons. Below the aortic valve stabilizing sutures are subsequently sewn through the aortic wall from inside to outside. With these sutures, an aortic prosthesis is pulled down over the outside of the aortic valve and secured deep into the aortic root. The aortic valve is now sutured into the aortic prosthesis. Finally, the two coronary arteries are re-implanted.

"David reconstruction"
For several decades various techniques for aortic valve reconstruction have been employed. In 1982 a technique was described by Sir Magdy Yacoub in which the ascending aorta is removed to just above the aortic valve and replaced with an aortic prosthesis that is cut to the appropriate size.

This so-called aortic valve *remodelling technique* does not involve a root stabilisation procedure. To prevent a later enlargement in the root, this technique can be combined with various stabilising techniques.
Therapy

Aortic valve replacement

Aortic valve replacement is necessary when structural changes in the valve (mostly calcification) lead to a severe narrowing (stenosis) or leaking (insufficiency) of the aortic valve. In many aortic operations of the ascending aorta the aortic valve is also affected and must be replaced when it cannot be reconstructed.

The original, no longer functional heart valve is cut out of the aortic valve annulus (a fibrous ring). Calcified material in the aortic valve annulus is removed with special forceps. The annulus is then measured and an appropriate prosthetic valve is chosen. Following this, sutures are placed in the aortic valve annulus. These threads are then sewn through the sewing ring of the prosthetic valve. The prosthetic valve is guided down into the annulus. The sutures are tied off and the prosthetic valve is checked for correct positioning.
**Mechanical prostheses**

Heart valve prostheses made out of synthetic materials are also called "mechanical valves". Most consist of a valve annulus in which two leaflets or cusps are attached to the inside. On the outside a sewing ring is affixed.

The advantage of mechanical valve prostheses is the virtually **unlimited durability**.

The disadvantage of this prosthesis is that the foreign surface of the prosthetic valve activates blood clotting. This drives clot formation on the prosthetic valve when clotting is not inhibited. This also leads to a risk of stroke. Therefore, patients who have a mechanical heart valve must take **blood-thinning medication for the rest of their lives**.

Most commonly warfarin is given. A side effect is that warfarin can cause unwanted bleeding (e.g. gastrointestinal hemorrhage). Therapy requires regular measurement of clotting parameters. This takes place in intervals of several weeks by a general practitioner or can be done by the patient himself with a measurement device.

**Bioprostheses**

Biological prosthetic valves consist mostly of bovine pericardial tissue or porcine heart valves that are fixed to a sewing ring.

The main advantage of these prostheses is that they do not activate blood clotting and so the patient only needs to follow a **blood-thinning therapy** for a short time after the surgery (**about 2-3 months**).

The disadvantage of bioprostheses is that they calcify and degenerate over time. In this way a new narrowing in the valve or leaking can develop. The valve must be replaced again. The **lifespan** of biological prosthetic valves is on average **10-15 years**.
Therapy

Valved conduit

If the aortic valve, aortic root and the ascending aorta all need to be replaced at once, composite grafts that consist of an aortic prosthesis and a prosthetic aortic valve are used.

So-called valved conduits are either sewn together at the factory or constructed by the surgeon during the operation.

Hand sewn biological valved conduit
Replacement of the ascending aorta

If only the ascending part of the aorta is affected by an aneurysm, this can be replaced as a rule with a simple, straight aortic prosthesis (so-called "tube graft").

For this, the ascending aorta is clamped before the aortic arch and, during a short period of cardiac arrest, subsequently replaced with a prosthesis that spans just above the openings of the coronary arteries to the aortic clamp. After removing the aortic clamp, the heart is again perfused with blood and begins to beat.

Partial aortic arch replacement

For many patients who undergo surgery due to an aneurysm of the ascending aorta, a small part of the aortic arch is also replaced under a short period of circulatory arrest. The replacement of the entire aortic arch is not necessary. It is often called, "hemiarch replacement", "partial aortic arch replacement" or more accurately "open anastomosis", because the aortic arch must be briefly opened, without it being completely replaced, for the suturing in of the ascending aortic prosthetic.

For a short period of circulatory arrest, patients must be cooled (25°C) to protect the organs. In addition, the vessels to the head are sometimes supplied with blood via a special catheter (antegrade cerebral perfusion), in particular when the suturing of the aortic prosthesis is expected to take longer than ten minutes.
Total aortic arch replacement

Because of its anatomical characteristics, replacing the entire aortic arch is one of the most complex interventions in heart surgery. The three arteries that supply the arms and head with blood begin at the aortic arch. After the aortic arch, the aorta transitions into the descending aorta, which supplies the lower part of the body with blood. The aortic arch can only be opened when the blood flow to these arteries is temporarily stopped. Thus, the operation takes place under circulatory arrest.

After opening the aortic arch during circulatory arrest, each individual vessel to the head is perfused (so-called selective antegrade cerebral perfusion) with a special catheter to supply the brain with oxygen. Initially the lower part of the body remains in circulatory arrest. At a later point, it will be again supplied with blood via a prosthetic aortic arch. Circulatory arrest is then ended.

The arteries that begin at the aortic arch are either sewn into the aortic prosthesis grouped as a combined "island" or separately.

"Four branch prosthesis" with "frozen elephant trunk" (Vascutek Thoraflex™ Hybrid)
There are special prostheses with four side arms (three for connecting the vessels and one for the heart-lung machine) available for the latter technique. It is casually called the "four finger prosthesis". In order to better suture it into the distal aortic arch (section of the aortic arch closest to the descending aorta), it has a "collar".

If the enlargement extends to the descending part of the aorta, this must usually be treated in a later, second operation via the left side of the chest. This can be facilitated by leaving a part of the prosthetic aortic arch "dangling" freely in the blood stream in the first operation, which will serve as a starting point for the second operation. This technique is called the "elephant trunk" technique. It was developed in Hanover, Germany by Professor Hans-Georg Borst and first introduced in 1983.

If the elephant trunk part of the prosthetic aortic arch is additionally equipped with a stent graft, it is called the "frozen elephant trunk" technique because the prosthesis no longer "dangles" freely but is fixed, or is "frozen". Because these prostheses are part "normal" aortic prosthesis and part "stented", they are called hybrid prostheses.

If only the beginning of the descending aorta is affected in addition to the aortic arch, a second operation can be skipped by using such hybrid prostheses. The first "frozen elephant trunk" prosthesis was developed in Hanover, Germany (Haverich-Chavan prosthesis) and has been continually improved since then. There now exists the Thoraflex™ Hybrid prosthesis by Vascutek, an aortic arch prosthesis that combines all of the above-mentioned features. It is, at the moment, the most complex aortic prosthesis available.

The main part of the operation when the surgeon is working on the aorta takes about one to two hours. The heart is isolated from blood flow from the aorta and must be protected during this time. Until recently this was achieved through cardiac arrest under cooled conditions (so-called cardioplegia). Because the risk for complications increases with the amount of time under cardiac arrest, a technique was developed in Hanover, Germany to supply the heart with blood during aortic arch surgery. Because the heart during this time usually beats normally, the technique is called the "beating heart" technique. This can considerably reduce the risk of aortic arch surgery.
Aortic arch aneurysm involving the descending aorta before and after surgery with the hybrid prosthesis
Valve sparing David procedure and complete aortic arch replacement with hybrid prosthesis with "beating heart" technique
Follow-up

Depending on the type of intervention, localisation and type of aortic disease, different follow-up appointments are required.

The first year of post-operative care serves to monitor the outcome of the operation and to allow for the review of complications that can arise from the surgery. Thereafter the follow-up serves to identify new aortic changes.

After surgery of the aortic root and ascending aorta

If surgery is limited to the aortic root and ascending aorta, annual echocardiography examinations of the heart will suffice. The first heart ultrasound takes place during hospital stay.

After heart valve surgery or a replacement of the ascending aorta, the patient should be referred to an out-patient cardiologist.

After surgery of the aortic arch and thoracoabdominal aorta

A CT angiography is the test of choice for the aortic arch and thoraco-abdominal aorta.

If there is a contra-indication for a computed tomography (CT) or the administration of a contrast agent (dye), magnetic resonance imaging (MRI) can be used to carry out the follow-up examination. To reduce radiation exposure in young patients, an MRI examination is recommended as long as an exact anatomical assessment is not necessary.
The first follow-up examination usually takes place before discharging the patient. Renewed imaging is recommended after 6 months and following that, annually. Under stable conditions and close consultation with the responsible physician, the intervals between the follow-up appointments can be lengthened.

When all affected parts are not replaced or the aortic disease is chronic

If not all affected sections of the aorta are replaced or there is a chronic aortic disease, post-operative care is not only needed to monitor the operation results but to identify new aortic changes.

Through continuous imaging and clinical examinations progression of the disease can be detected. In this way, the need for another operation can be discovered early. Complications can be avoided.

CT angiography is the most common examination technique to assess the aorta during follow-up. MRI examination can be used under certain conditions.

As well as aortic imaging, routine doctor visits should take place. In addition to the surgery, optimising cardiovascular risk factors and monitoring medication regime are essential.
After completing a rehabilitation plan, the patient should be able to return to daily life or work as usual. Because the healing process is different for every person, it is difficult to make binding statements.

In addition, some professions and hobbies should be stopped if patients have to take blood thinners (e.g. warfarin).

The breastbone should heal after about three months and patient can again take up exercise. Take care to exert yourself steadily. Endurance exercise (walking, bicycle riding, jogging, swimming) and moderate strength training are suitable.

The patient should refrain from driving an automobile for the first six weeks because glancing over the shoulder and turning the steering wheel put pressure on the chest that can cause pain.

As a passenger, take time getting in and out of an automobile to protect your chest. Using a seatbelt is still mandatory after an operation.

Next to surgery, medication plays an important role. Risk factors like too high blood sugar or high blood pressure must face targeted treatment.

After an aortic intervention, you should take a platelet aggregation inhibitor, like ASS 100 (once daily) for the rest of your life. An exception is when warfarin is necessary for blood thinning.
Travel and wellness

Long trips can be taken three months after surgery at the earliest.

Bring an adequate supply of medicine as well as a copy of the medical report.

Also, use caution when carrying heavy luggage.

Flying after being discharged from the hospital is possible. Stays at altitudes up to 2,000 meters are also safe.

Sauna visits should first be enjoyed no earlier than three months after the operation.
From left: Dr. T. Kaufeld, J. Umminger, H. Krueger, Dr. E. Beckmann, Prof. M. Shrestha, Dr. A. Martens
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This publication may be subject to modification.
In an emergency

Go directly to your closest hospital or call an ambulance.
Explain that you are a patient with aortic disease.