Guidelines for Diagnosis and Treatment of Aortic Aneurysm and Aortic Dissection (JCS 2011)
– Digest Version –
JCS Joint Working Group

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Introduction of the Revised Guidelines

Since the “Guidelines for Diagnosis and Treatment of Aortic Aneurysm and Aortic Dissection (JCS 2006)” were issued by the Japanese Circulation Society (JCS) in 2006,1 we have seen advances in the treatment of aortic diseases, and partial revision of the Guidelines has thus been approved by the JCS Scientific Committee. Over the past 5 years, stent-grafting has rapidly become common in the treatment of aortic diseases in Japan, being employed in many institutions and becoming indispensable in the management of aortic diseases, particularly of the descending aorta. In addition, much about the relationship between aortic diseases and genetic abnormalities has become apparent, and new aspects of treatment have evolved. Chapters that address these new aspects of treatment in the present guidelines provide particularly detailed descriptions. These guidelines also indicate the difference between the interpretation of aortic dissection in Japan and that in Western countries. Accordingly, this update of the guidelines represents, in practical terms, a full-scale revision of the guidelines.

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Circulation Journal Vol.77, March 2013
The incidence of aortic diseases is high in Japan in comparison with other countries worldwide. In particular, the frequency of aortic dissection is ranked at the top, paralleling the situation in Italy. The cited underlying factors include a high prevalence of hypertension, a large proportion of elderly in the population, and high availability of computed tomography (CT) which facilitates the diagnosis of aortic diseases. Intramural hematoma (IMH) is a type of aortic dissection commonly referred to in Western countries. The pathological condition of this disease is, essentially, hemorrhage in the tunica media of the aortic wall, resulting in hematoma. There is currently discussion about the relationship between this condition and classic aortic dissection that originates from a tear in the tunica intima, with dissection progressing in the tunica media. Fundamentally, IMH is a pathological diagnosis. However, the current situation is that radiologists make a diagnosis of IMH only because there is no evidence of the presence of a tear on diagnostic imaging, or because the dissected lumen is not visualized. According to the American College of Cardiology Foundation/American Heart Association (ACCF/AHA) guidelines for the diagnosis and management of patients with Thoracic Aortic Disease issued in 2010, a theoretically incongruous concept of IMH with ulcer-like projection (ULP) has been presented. In the Western countries, a diagnosis of IMH is often made by a single CT session in clinical practice, and it is explained that a tear may form after a diagnosis of IMH and result in communicating aortic dissection. However, it is not certain that this explanation is valid. It is possible that the contrast agent does not enter the dissected lumen because of blood retention in the dissected lumen without reentry despite a tear being present from the beginning. In Western countries, cases with thrombotic occlusion of the false lumen of the ascending aorta in type III retrograde dissection have actually been referred to as type-A IMH. In addition, in Western countries, the efficacy of medical treatment for IMH is considered to be poor because IMH may proceed to classic aortic dissection in the future. In contrast, in Japan and South Korea, the frequent use of CT allows an accurate understanding of changes in the pathological features of this condition, and observation of the disease course upon medical treatment thus generally achieves favorable results.

Therefore, this set of guidelines provides the view that the term IMH, which may lead to an incorrect understanding of the pathological condition, should not be used clinically in Japan. We believe that non-communicating aortic dissection is the proper term that accurately expresses the pathological features of this condition, and is more useful for deciding an appropriate therapeutic strategy in clinical practice. Discussion should be continued through academic meetings in Western countries, concerning this discrepancy from ACCF/AHA guidelines.

We expect that these guidelines will serve as a good treatment guide for aortic dissection and aortic aneurysm. However, guidelines are only indices that are based on the currently available evidence, and are not obligatory rules that require complete adherence. It should be recognized that physicians who are particularly specialized in this field may choose a certain treatment that deviates from the guidelines, based on sufficient knowledge of the evidence from research and practical processes of new treatment as well as better medical services. However, it is important for young physicians to fully understand these guidelines in order to attain an up-to-date level of medical practice.

Japan has a high frequency of aortic diseases in comparison with other countries in the world, and is enjoying a high level of accuracy and much better results in medical practice which surpass those in Western countries. On behalf of all persons involved in the revision of these guidelines, we sincerely hope that these guidelines lead to further improvements in the level of healthcare in Japan and to saving more patients and achieving greater quality of life (QOL) for those affected by aortic diseases.

The following classifications are used for expressing the class of recommendations for particular methods of diagnosis and treatment and the level of evidence according to the relevant American College of Cardiology (ACC)/AHA Guidelines (http://circ.ahajournals.org/manual/manual_IIstep6.shtml; available in January 2011).

Classification of Recommendations
Class I: Conditions for which there is evidence and/or general agreement that a given procedure or treatment is useful and effective.
Class II: Conditions for which there is conflicting evidence and/or a divergence of opinion about the usefulness/efficacy of a procedure or treatment.
Class III: Conditions for which there is evidence and/or general agreement that the procedure/treatment is not useful/effective, and in some cases may be harmful.

Level of Evidence
Level of Evidence: A
Data derived from multiple randomized clinical trials
Level of Evidence: B
Data derived from a single randomized trial, or non-randomized studies
Level of Evidence: C
Consensus opinion of experts

Although the flap in aortic dissection usually has one to several tears, some cases have no clear tear or no communication between the true lumen and the false lumen. The former type is called communicating aortic dissection, and the latter non-communicating aortic dissection (synonymous with the conventional thrombosed type).

Recent progress in diagnostic imaging has allowed us to detect a condition in which the aortic media is detached due to hematoma, without an observable tear. This condition is called...
IMH or intramural hemorrhage, representing the distinct pathological concept termed "aortic dissection without tear". However, because IMH is based on a pathological diagnosis, we have decided not to use this term in the clinical setting. So-called IMH, devoid of tears on diagnostic imaging, is clinically regarded as non-communicating aortic dissection (synonymous with the conventional thrombosed type) and dealt with as a type of dissection.

Clinically, it is often difficult to clearly distinguish between IMH and aortic dissection that has an intimal defect and tear (ULP on diagnostic imaging), without evidence of blood flow in the false lumen (thrombosed false lumen with intimal defect, ULP-type). In addition, because the ability to detect ULP varies among different diagnostic imaging techniques, and ULP-type dissection includes some unstable cases regardless of the size of the ULP, this condition is clinically important. To bring clinical attention to the importance of ULP, it is recommended that ULP-type dissection be dealt with as communicating aortic dissection.

In addition, cases with partial thrombus in a false lumen and a thrombosed false lumen communicating with the true lumen should definitely be classified into the category of communicating aortic dissection.

Although penetrating atherosclerotic ulcer (PAU) was proposed as a concept that represents an ulcerated aortic atherosclerotic lesion that extends to the media, much remains unclear as to the relationship between PAU and aortic dissection. Various issues about IMH and PAU remain to be clarified, and due caution is therefore necessary when applying these terms.

2. Aortic Aneurysm

Aortic aneurysm is a circumferential or local enlargement (increased diameter) or protrusion of a part of the aortic wall.

When a part of the aortic wall is dilated and thereby forms a bump, or when the diameter is increased to a degree at least 1.5-fold greater than normal (exceeding 45 mm in the thoracic region and 30 mm in the abdominal region) in a fusiform manner, the condition is called aneurysm.

Aortic aneurysm is a localized dilatation (increased diameter) or protrusion of the aortic wall, and is called a fusiform type aortic aneurysm (Figure 1) or a saccular type aortic aneurysm (Figure 2) according to whether the shape is fusiform or saccular, respectively.

2. Glossary

Aortic dissection
- Dissecting aneurysm of the aorta: Aortic dissection forming an aneurysm.
- Classic aortic dissection: Dissection with a tear or flap, in contrast to IMH.
- True lumen: Original arterial lumen.
- False lumen: New lumen formed in the wall (dissected lumen is an inappropriate term).
- Flap: Septum (of the internal media). Although also referred to as detached intima, a flap actually consists of part of the intima and media.
- Tear: A tear involving parts of the intima and media and representing a communication between the true lumen and false lumen in dissection. Intimal tear is also idiomatically used as a synonym.
- Entry: An area where blood flow enters from the true lumen into the false lumen.
- Reentry: An area where blood flow enters from the false lumen into the true lumen.
- Communicating aortic dissection: The same as communicating aortic dissection in the classification by European Society of Cardiology (ESC), also known as classic dissection and double barreled aorta.
- Non-communicating aortic dissection: The same as non-communicating aortic dissection in the classification by ESC.
- Thrombosed type aortic dissection: Synonymous with non-communicating aortic dissection.
- Intramural hematoma (IMH): Used almost synonymously with dissection without a tear from the pathological viewpoint or non-communicating aortic dissection from the clinical viewpoint. This term is not, however, used clinically because it is based on a pathological diagnosis.
- Intramural hemorrhage: Synonymous with IMH.
- Ulcer-like projection (ULP): Small protrusion found in part of the false lumen on diagnostic imaging including arteriography. Although detection abilities vary among different diagnostic imaging techniques, the findings of ULP on imaging include various pathological conditions (e.g., tear, rupture of a branch, atherosclerotic ulceration). Clinically, ULP is an unstable condition regardless of the size, such that close observation is necessary. Therefore, to assure clinical atten-
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Table 1. Classification of Aortic Dissection

1. Classification based on the extent of dissection

<table>
<thead>
<tr>
<th>Table 1. Classification of Aortic Dissection</th>
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<tbody>
<tr>
<td>1. Classification based on the extent of dissection</td>
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<tr>
<td>Stanford classification</td>
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<tr>
<td>Type A: Dissection involving the ascending aorta</td>
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<tr>
<td>Type B: Dissection not involving the ascending aorta</td>
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<tr>
<td>DeBakey classification</td>
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<tr>
<td>Type I: A tear located in the ascending aorta and dissection extending to the aorta distal to the aortic arch</td>
</tr>
<tr>
<td>Type II: Dissection confined to the ascending aorta</td>
</tr>
<tr>
<td>Type III: A tear located in the descending aorta</td>
</tr>
<tr>
<td>Type IIIa: Dissection not involving the abdominal aorta</td>
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<tr>
<td>Type IIIb: Dissection involving the abdominal aorta</td>
</tr>
</tbody>
</table>

The following subcategories can be added to the DeBakey classification

| Arch type: A tear located in the aortic arch |
| Arch localized type: Dissection confined to the aortic arch |
| Arch extended type: Dissection extending to the ascending or descending aorta |
| Abdominal type: A tear located in the abdominal aorta |
| Abdominal localized type: Dissection confined to the abdominal aorta |
| Abdominal extended type: Dissection extending to the thoracic aorta |

(The term “retrograde type III dissection” is not used.)

2. Classification based on blood flow in the false lumen

| Communicating type: Presence of blood flow in the false lumen. Also included in this category are partially thrombosed false lumen or cases with blood flow in the long axis direction in the false lumen from the ULP, despite a large portion of the false lumen being thrombosed. |
| ULP-type: No blood flow in a large portion of the false lumen, but blood flow localized near the tear (ULP) |
| Non-communicating type: Crescent-shaped false lumen without either tear (including ULP) or blood flow in the false lumen |

3. Classification based on the disease phase

| Acute phase: Presentation within 2 weeks following the initial event. Very acute phase refers to those presenting within the first 48 hours. |
| Chronic phase: Presentation at 2 weeks or longer following the initial event |

ULP, ulcer-like projection.

- Circumferential enlargement of the aortic wall to a diameter 1.5-fold greater than normal is called fusiform type aneurysm, whereas knobby protrusion of part of the aortic wall is called saccular type aneurysm. If there is no clear differentiation, the case should be dealt with as a saccular type lesion.

- Thoracic aortic aneurysm (TAA): The name given to an aneurysm occurring in the aorta of the thorax. Ascending aorta refers to the part beginning from the aortic annulus to the bifurcation of the brachiocephalic artery, aortic arch refers to the origin of the brachiocephalic artery to the level of the 3rd to 4th thoracic vertebra (the sites of right and left bifurcations of the pulmonary artery) and descending aorta refers to the part from the level of the 3rd to 4th thoracic vertebra and below.

- Thoracoabdominal aortic aneurysm (TAAA): The name given to an aneurysm involving a continuous area from the thorax to the abdominal cavity. TAAA is divided into 4 types according to Crawford classification.

- Abdominal aortic aneurysm (AAA): The name given to an aneurysm occurring in the abdominal aorta.

- Inflammatory abdominal aortic aneurysm (IAAA).

- True aneurysm of the aorta: Synonymous with so-called aortic aneurysm. This term is used when clear distinction from pseudoaneurysm of the aorta is intended. Although the wall of the aneurysm consists of the original arterial wall, the media may not be histologically confirmed when the aneurysm is enlarged.

- Pseudoaneurysm (false aneurysm) of the aorta: Aneurysm having no aortic wall structure. This type of aneurysm often has a traumatic or infectious etiology.

3. Classifications and Pathological Conditions

1. Aortic Dissection

1) Classifications

Table 1 shows 1) classification by the extent of dissection, 2) classification by status of blood flow in the false lumen, and 3) classification by disease phase.

2) Pathological Conditions

Aortic dissection essentially characterized by dissection of the
aortic wall and blood flow into the dissection is subject to changes over time beginning from immediately after onset, and therefore presents as dynamic pathological conditions. These pathological conditions are also variable because the lesion extends to a broad zone of the blood vessel (Figure 3). The wide spectrum of pathological conditions is better understood if the status of the blood vessel is classified into 1) dilatation, 2) rupture, and 3) stenosis or occlusion, and combined with the site of dissection.

1) Dilatation
   (a) Aortic valve insufficiency
   (b) Aneurysm formation
2) Rupture
   (a) Cardiac tamponade
   (b) Bleeding into the thoracic cavity or other areas
3) Peripheral Circulatory Disturbance Due to Stenosis or Occlusion of the Aortic Branches
   (a) Angina, myocardial infarction
   (b) Cerebral ischemia
   (c) Upper limb ischemia
   (d) Paraplegia
   (e) Intestinal tract ischemia
   (f) Renal failure
   (g) Lower limb ischemia
4) Other Pathological Conditions
   (a) Disseminated intravascular coagulation syndrome (DIC), pre-DIC
   (b) Pleural effusion
   (c) Systemic inflammatory response syndrome

(3) Definition of Non-Communicating Aortic Dissection
Because it is difficult to clinically distinguish between dissection without a tear and dissection with a tear showing no blood flow in the false lumen, it is preferable to avoid the use of the term aortic IMH, which pathologically denotes hematoma in the aortic wall due to rupture of a blood vessel feeding the aorta.

Non-communicating aortic dissection is defined as follows:
1. It has a crescent-shaped false lumen.
2. There is no tear and blood flow entering from a tear. Namely, there is no communication between the false lumen and the true lumen.

If there is obvious blood flow in the long axis direction in the false lumen, the lesion should not be dealt with as non-communicating aortic dissection. In these guidelines, ULP-type dissection and non-communicating dissection are defined as different pathological conditions (Figure 4). On the other hand, cases with a thrombosed false lumen resulting from retrograde dissection that arose from a tear in the descending thoracic aorta or abdominal aorta should be classified as communicating dissection, although their imaging findings closely resemble with those of non-communicating dissection (Figure 5).

2. Aortic Aneurysm
(1) Classification
   1) Morphology of the Aneurysm Wall
      (a) True aneurysm of the aorta
      (b) Pseudoaneurysm of the aorta
      (c) Dissecting aneurysm of the aorta

   2) Location of the Aneurysm
      (a) Thoracic region
      (b) Thoracoabdominal region
      (c) Abdominal region

   3) Etiology
      (a) Atherosclerotic
      (b) Traumatic
      (c) Inflammatory
      (d) Infective
**4. Statistics and Epidemiology**

1. **Incidence Rate per Year**
   
   Limited regional investigation data indicate that the incidence rate per year seems to be about 3 out of 100,000 population.\(^{11}\)
   
   According to the statistics reported by the Japanese Association for Thoracic Surgery,\(^{12}\) there were 5,985 cases undergoing surgery for non-dissecting thoracic aortic aneurysm (TAA) or thoracoabdominal aortic aneurysm (TAAA) and 5,013 cases undergoing surgery for dissecting aortic aneurysm in 2008. The number of surgical cases tends to be increasing annually.\(^{12-16}\)

2. **Changes in the Incidence Rate According to Age**
   
   Based on estimations from autopsy cases, the onset of aortic dissection peaks in both men and women in their 70s,\(^{17}\) whereas the peak for non-dissecting aortic aneurysm is in men in their 70s and in women in their 80s.\(^{17}\)

3. **Changes in the Incidence Rate According to the Season, Time, and Day of the Week**
   
   The onset of aortic aneurysm tends to be more frequent in winter and less frequent in summer.\(^{18-20}\) In terms of time, aortic aneurysm occurs more frequently in the daytime, i.e., when people are active; the onset is reported to be particularly frequent during the period of 6:00 to 12:00. Conversely, the onset is rare from late at night until early in the morning.\(^{18,19,21}\)

4. **Aortic Dissection in Cases of Sudden Death**
   
   According to a report from the Tokyo Metropolitan Medical Examiner’s Office,\(^{20}\) pre-hospital mortality rate accounted for 61.4%, death within an hour after onset for 7.3%, 1 to 6 hours after onset for 12.4%, and 6 to 24 hours after onset for 11.7% of mortalities associated with aortic dissection. Combining pre-hospital with in-hospital mortality rates shows that 93% of deaths from aortic dissection occur within 24 hours after onset.
II Diagnosis

1. General Remarks

1. Acute Aortic Dissection
It is of utmost importance to maintain suspicion when diagnosing acute aortic dissection. Diagnosis should be made according to the flow chart shown in Figure 6.

2. True Aortic Aneurysm
Although many TAA cases are asymptomatic, symptoms such as hoarseness, difficulty swallowing, and vague back pain may occur. If TAA is detected, chest CT should first be performed to determine the most appropriate therapeutic strategy (Figure 7).

Abdominal aortic aneurysm (AAA) may present symptoms such as a feeling of abdominal distension, constipation, non-specific low back pain. A pulsatile mass in the abdomen is an objective finding that leads to suspicion of AAA in some cases. Abdominal ultrasonography is the simplest and non-invasive evaluation technique for initial diagnosis (Figure 8). The patient should be followed up by ultrasonography or CT to determine the most appropriate therapeutic strategy.

2. Radiological Diagnosis: Plain X-Ray Examination, CT, Angiography

1. Plain X-Ray Examination
(1) Aortic Aneurysm
TAA is often detected by plain chest X-ray examination at regular health check-ups in asymptomatic patients. Ascending aortic aneurysm usually presents a shadow protruding rightward continuing from the contour of the ascending aorta in the frontal view, whereas aortic arch aneurysm is often visualized as a tumorous shadow at the first left aortic arch in the frontal view, and descending aortic aneurysm as a fusiform or round shadow continuing from the contour of the aorta. In cases with AAA, calcification can sometimes be observed in the aneurysmal wall, indicating the presence of an aneurysm.

(2) Aortic Dissection
In cases with acute aortic dissection, mediastinal shadow en-

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Figure 6. Diagnosis and management algorithm for acute aortic dissection. ACS, acute coronary syndrome; CT, computed tomography; CRP, C-reactive protein; ECG, electrocardiograph; Hb, hemoglobin; ICU, intensive care unit; s/o, subjective symptoms/objective signs; TEE, transesophageal echocardiography; WBC, white blood cell.
When aortic aneurysm rupture is suspected, CT is useful if the patient’s condition allows time for such examination. It is important to carefully interpret CT images to avoid overlooking any minor bleeding.

**2. CT**

**(1) Method**

For CT of aortic aneurysm or aortic dissection, plain CT images and early contrast-enhanced images are indispensable, and late contrast-enhanced images should be added depending on the need in individual cases. Plain CT is useful for determining the degree of mural calcification, presence/absence of medial deviation, identifying hematoma in the false lumen in cases with non-communicating dissection, and evaluating the high-density area in the mural thrombus suggesting impending rupture of an aortic aneurysm.\(^2^2\)

**(2) CT of Aortic Aneurysm**

CT allows us to identify an aneurysm and provides data on the size and extent of the aneurysm, calcification and the status of the aneurysmal wall (e.g., inflammatory aortic aneurysm), the amount and status of the mural thrombus, the relationship between the aneurysm and surrounding organs, and the positional relationship between the aneurysm and major aortic branches. Although the diameter of the aneurysm is an important factor in determining whether surgery is indicated, "the maximum minor-axis diameter" should be used in principle for CT evaluation.\(^2^3\)

**(3) CT of Ruptured Aortic Aneurysm and Impending Rupture**

When aortic aneurysm rupture is suspected, CT is useful if the patient’s condition allows time for such examination. It is important to carefully interpret CT images to avoid overlooking any minor bleeding.

**(4) CT of Aortic Dissection**

CT is a highly reliable, non-invasive examination for diagnosing dissection. This examination is indispensable for diagnosing aortic dissection because it allows objective evaluation of the whole aorta within a short period of time in response to an urgent need.

On plain CT, deviation of intimal calcification is a critical point in diagnosis.

**1) Communicating Dissection**

In some cases with communicating dissection, blood flow in the false lumen is so slow that the false lumen cannot be visualized in the early-phase contrast, but inflow of the contrast agent is observed in the late-phase contrast. Therefore, it is necessary to obtain late contrast-enhanced images as well. The entry is recognized as a rupture of the flap.

**2) Non-Communicating Dissection**

CT images of non-communicating dissection are characterized by the presence of a false lumen filled with clotted blood or hematoma in the acute phase. This false lumen is visualized as a crescent-shaped or annular shadow resembling the mural
Body surface ultrasonography allows observation of the aortic root, ascending aorta, aortic arch, brachiocephalic artery, left common carotid artery, and left subclavian artery. In addition, arterial branches from the abdominal aorta, i.e., the celiac artery, superior mesenteric artery, renal artery, and common iliac artery, can also be observed. Transesophageal echocardiography can clearly visualize the area from the aortic root to the ascending aorta, aortic arch, and descending aorta.

3) Ulcer-Like Projection-Type Dissection
CT shows ULP as a localized luminal protrusion into the closed false lumen.

4) Diagnosis of Complications
Complications of aortic dissection often include serious conditions such as rupture, cardiac tamponade, and ischemia in organs or limbs. On CT evaluation, it is important to pay attention to the presence/absence of fluid accumulation around the heart, the relationship between arterial branches and the dissecting lumen, and the presence/absence of extension of the dissection into arterial branches.

3. Angiography
The role of angiography including digital subtraction angiography (DSA) in the diagnosis of aortic aneurysm or aortic dissection is diminishing.

3. Ultrasonography
Body surface ultrasonography and transesophageal echocardiography that are minimally invasive and provide large amounts of information are useful for visualizing the aorta. Body surface ultrasonography allows observation of the aortic root, ascending aorta, aortic arch, brachiocephalic artery, left common carotid artery, and left subclavian artery. In addition, arterial branches from the abdominal aorta, i.e., the celiac artery, superior mesenteric artery, renal artery, and common iliac artery, can also be observed. Transesophageal echocardiography can clearly visualize the area from the aortic root to the ascending aorta, aortic arch, and descending aorta.

1. Aortic Aneurysm
First, in imaging an aortic aneurysm, it is necessary to obtain the long axis and short axis views of the aorta on the body surface ultrasonography to observe the diameter of the aorta, shape of the aneurysm, positional relationship with branching of blood vessels, and the nature of the lumen and wall. Because there is a possibility that the aorta is flexed or deviated, the maximum minor-axis diameter should be measured when the short axis view is used.

2. Aortic Dissection
Ultrasonography is very useful for rapid diagnosis of aortic dissection, and can be used when it is difficult to use contrast agents because of issues such as renal dysfunction or allergy to contrast agents. In particular, body surface ultrasonography allows evaluation of branch dissection and complications of dissection, in addition to simple and non-invasive diagnosis of dissection. It is very important to evaluate cardiac tamponade (a complication of Stanford type A dissection), aortic valve regurgitation, progression of dissection into a branch, blood flow, and cardiac function.
4. MRI

1. Imaging Procedures25–28
(1) MRI
Magnetic resonance imaging (MRI) allows evaluation of any section of the vascular wall and lumen without the use of contrast agents. On the other hand, this procedure requires a long imaging time and may have artifacts due to turbulent flow or delayed flow orrespiration-related artifacts.

(2) Cine MRI
This imaging technique allows evaluation of hemodynamics without the use of contrast agents. However, a long imaging time is required, and information is basically limited to a single section.

(3) MRA
Magnetic resonance angiography (MRA) techniques are broadly classified into the time-of-flight (TOF) method, phase-contrast (PC) method, fresh blood imaging method in which a contrast agent is not used, and contrast-enhanced MRA in which a contrast agent is used. Among these techniques, the most common procedure for the aorta is contrast-enhanced MRA, which can provide good views of the blood flow in the lumen when evaluating the flexed part and the turbulent part. In comparison with the TOF or PC method, contrast-enhanced MRA is advantageous in that a shorter imaging time is required, there is high spatial resolution, and an arbitrary section can be set for imaging.

2. Clinical Application1,26,27,29–32
MRI is more advantageous than CT in that it requires no X-ray exposure, allows non-contrast imaging in patients with severe renal dysfunction, and enables the lumen to be evaluated in cases with severely calcified lesions. On the other hand, its disadvantages include lower spatial resolution, inability to visualize osseous structures due to the lack of information on calcification, and difficulty with responding to emergency cases because of the long imaging time.

MRI is not recommended for diagnosis of acute aortic aneurysm in patients in a poor general medical condition, because it is very time-consuming and has limitations in patient monitoring.

3. Safety of Implanted Devices and Metals
The safety of individual devices should be confirmed by references, the website (www.mrisafety.com), and the latest package insert.

5. Identification of Adamkiewicz Artery

To avoid paraplegia, one of the most serious complications of surgery involving the thoracic (abdominal) aorta, attempts have been made to identify the anatomical position of the Adamkiewicz artery by diagnostic imaging prior to surgery. Although CT and MRI have their own advantages and disadvantages, it has been reported that diagnostic capability reaches 90% for the Adamkiewicz artery if both CT and MRI can be performed in a particular patient.33 This data is approximately equivalent to that for angiography.34

1. CT
To visualize the Adamkiewicz artery by CT, the inside of the vertebral canal should be observed in an oblique coronal view by the multiplanar reformation (MPR) method using multi-detector-row CT (MDCT).33,35–37 Because the Adamkiewicz artery makes a distinguishable “hairpin turn” before confluence to the anterior spinal artery, it serves as a landmark for identification of the artery. The continuity of the route from the aorta to the intercostal (lumbar) artery, its posterior branch, root-medullary vein, Adamkiewicz artery, and anterior spinal artery should be observed as a traversable single blood vessel route by the curved planar reformation (CPR) method.33,35,36

2. MRA
Images should be obtained by contrast-enhanced MRA. There are two MRA techniques available for visualizing the Adamkiewicz artery.38 One is high spatial resolution MRA that focuses on spatial resolution,33,36 and the other is time-resolved MRA that focuses on time resolution.39,40 Time-resolved MRA is more common at present.

III Choice of Treatment Methods

1. Aortic Dissection (Recommendations for Choosing Treatment Methods for Acute Aortic Dissection: Tables 3, and 4)

In the treatment of aortic dissection, choosing between medical and surgical treatment is the most important decision affecting prognosis.

1. Treatment in the Acute Phase
(1) Stanford Type A Acute Aortic Dissection
Stanford type A aortic dissection in which dissection extends to the ascending aorta is a condition that has an extremely poor prognosis. Mortality is reported to be 1 to 2% per hour after the onset of symptoms.41 The main causes of death include rupture, cardiac tamponade, circulatory failure, cerebral infarction and intestinal tract ischemia.42,43 In general, surgical treatment, i.e., emergency surgery, is indicated for this condition, because the prognosis with medical treatment alone is extremely poor.

(2) Stanford Type B Acute Aortic Dissection
Because Stanford type B acute aortic dissection follows a better natural course than type A acute aortic dissection, medical treatment is generally chosen as the initial management strategy. On the other hand, surgical treatment is necessary for patients with complications such as rupture, refractory pain, or organ ischemia because the prognosis is extremely poor.44 However, because in-hospital mortality after surgical treatment in the acute phase is not low,44 a good alternative to the currently available surgical treatments is awaited. In recent years, endovascular treatment has been achieving favorable results as a therapeutic option for acute type B aortic dissection,45–47 and is now becoming a first-line therapy for patients with acute type B aortic dissection who have fatal complications.
Table 3. Recommendations for Acute-Phase Treatment for Stanford Type A Aortic Dissection

| Class I   | 1. Surgical treatment (emergency surgery) for type A communicating aortic dissection (type I, II, and retrograde type III) (Level of Evidence: C)  
2. Surgical treatment for aortic dissection in patients with severe complications* directly related to aortic dissection where surgery is expected to achieve improvement or stop progression (Level of Evidence: C)  
   *e.g., rupture of the false lumen, re-dissection, cardiac tamponade, disturbed cerebral circulation accompanied by disturbance of consciousness or paralysis, aortic valve insufficiency accompanied by heart failure, myocardial infarction, renal failure, intestinal circulatory disorder, thromboembolism of the limbs |
| Class IIA  | 1. Surgical treatment for aortic dissection where drug therapy has failed to control blood pressure and pain and for type A non-communicating aortic dissection (Level of Evidence: C)  
2. Medical treatment to be started under certain conditions (see Section 1.1.3 of Chapter III) for type A dissection without complications or persistent pain where the false lumen of the ascending aorta is thrombosed (Level of Evidence: C)  
3. Revascularization with surgical or endovascular procedures for impaired intestinal perfusion accompanied by acute aortic dissection where emergency aortic surgery is not indicated (Level of Evidence: C) |
| Class IIB  | 1. Aortic surgery for patients with serious brain damage (Level of Evidence: C) |
| Class III | 1. Revascularization for impaired organ perfusion where emergency aortic surgery is indicated (Level of Evidence: C) |

Table 4. Recommendations for Acute-Phase Treatment for Stanford Type B Aortic Dissection

| Class I   | 1. Medical treatment for uncomplicated type B communicating/ULP-type/non-communicating aortic dissection (Level of Evidence: C)  
2. Surgical treatment for aortic dissection in patients with severe complications* directly related to aortic dissection where surgery is expected to achieve improvement or stop progression (Level of Evidence: C)  
   *e.g., rupture of the false lumen, re-dissection, cardiac tamponade, disturbed cerebral circulation accompanied by loss of consciousness or paralysis, aortic valve insufficiency accompanied by heart failure, myocardial infarction, renal failure, intestinal circulatory disorder, thromboembolism of the limbs |
| Class IIA  | 1. Surgical treatment for aortic dissection where drug therapy has failed to control blood pressure and pain (Level of Evidence: C)  
2. Medical treatment for aortic dissection where drug therapy has failed to control blood pressure (Level of Evidence: C)  
3. Revascularization with surgical or endovascular procedures for impaired intestinal perfusion accompanied by acute aortic dissection where emergency surgery is not indicated (Level of Evidence: C) |
| Class IIB  | 1. Aortic surgery for patients with serious brain damage (Level of Evidence: C) |
| Class III | 1. Surgical treatment for uncomplicated type B aortic dissection (Level of Evidence: C)  
2. Revascularization for impaired organ perfusion where emergency aortic surgery is indicated (Level of Evidence: C) |

ULP, ulcer-like projection.

(3) Treatment for Special Dissection

1) Stanford Type A Non-Communicating Acute Aortic Dissection

There is a discrepancy in the view as to the treatment policy for Stanford type A non-communicating aortic dissection between Western countries and Japan or South Korea. It is also common within the country for surgeons and internists to have different opinions on this issue. Table 5 shows the therapeutic results described in previous reports.

At present, the treatment strategy for Stanford type A non-communicating acute aortic dissection is as follows.

First, emergency surgery should be considered for patients with complicated aortic valve insufficiency or cardiac tamponade. When there is ULP in the ascending aorta, surgery at an early stage should be considered because it is presumed that the already existing tear has developed into a ULP-type lesion.

Patients with an aortic diameter of 50 mm or greater or a hematoma thickness of 11 mm or greater are regarded as being at high risk, and surgery may be considered according to circumstances. In other cases, initial medical treatment seems to be possible. However, diagnostic imaging should be performed frequently during medical treatment, and it is advised that cases showing an increase in the thrombosed false lumen or a change to ULP or communicating type promptly undergo surgery.

2) Stanford Type A Retrograde Dissection With a Tear in the Descending Thoracic Aorta

In some cases, there is no tear in the ascending aorta, and retrograde dissection extends from a tear located in the descending thoracic aorta (or abdominal aorta in rare cases) to the ascending aorta. Such retrograde dissection can be managed with medical treatment by performing diagnostic imaging frequently during follow-up, while observing for possible increases in the thrombosed false lumen or new blood flow into the false lumen.

3) Stanford Type B Non-Communicating Acute Aortic Dissection

Stanford type B non-communicating acute aortic dissection shows a better prognosis than type A non-communicating or type B communicating acute aortic dissection. Caution is necessary for ULP occurring in the acute phase (i.e., transition to a ULP-type lesion), an aortic diameter of 40 mm or greater, and a false lumen thickness of 10 mm or greater, because...
(3) Results of Medical Treatment in the Chronic Phase
Table 5 shows the long-term prognosis with medical treatment. Acute-phase factors associated with a poor prognosis in chronic phase are listed below.

1. Stanford Type A Communicating Dissection
2. Stanford Type A Non-Communicating Dissection
3. Stanford Type B Communicating Dissection
4. Stanford Type B Non-Communicating Dissection

these are reported to be risk factors for disease progression such as increasing aortic diameter in the chronic phase.

2. Treatment in the Chronic Phase (Recommendations for Treatment of Chronic Aortic Dissection: Table 6)
The prognosis of aortic dissection in the chronic phase (i.e., at least 2 weeks after onset) is favorable. If the patient is stable, medical treatment is recommended, regardless of whether the dissection is of Stanford type A or type B. Invasive treatment should be considered if the patient has conditions such as rupture, impending rupture, an increased aortic diameter, aortic valve insufficiency, branch occlusion, extended dissection, recurrence.

(1) Results of Surgery in the Chronic Phase
According to the Japanese Association for Thoracic Surgery, the in-hospital mortality rate in 2008 was 6.5% for chronic type A, 8.7% for chronic type B (descending arch replacement: 15.0%, descending aortic replacement: 7.9%, thoracoabdominal aortic replacement: 13.9%).

(2) Results of Stent-Graft Insertion in the Chronic Phase
According to the Japanese Association for Thoracic Surgery, the in-hospital mortality rate was 22.2% for chronic type A and 4.2% for chronic type B in 2008.

(3) Results of Medical Treatment in the Chronic Phase
Table 6 shows the long-term prognosis with medical treatment. Acute-phase factors associated with a poor prognosis in chronic phase are listed below.

1. Stanford Type A Communicating Dissection
2. Stanford Type A Non-Communicating Dissection
3. Stanford Type B Communicating Dissection
4. Stanford Type B Non-Communicating Dissection

New appearance of ULP, age 70 years or older, acute-phase maximum aortic diameter, thickness of hematoma, ULP located in the distal arch or surrounding the diaphragm.
surgical indication is an aortic diameter of 50 to 59 mm, when the risk of rupture and aortic dissection during medical treatment are taken into consideration.

In cases with descending aortic aneurysm and TAAA, the risk of the complication of paraplegia is high. When the risk of rupture and aortic dissection during medical treatment is taken into consideration, an aortic diameter of about 60 mm seems to be a reasonable criterion for the surgical indication.

### 3. Abdominal Aortic Aneurysm (Recommendations for Choosing Treatment Methods for Abdominal Aortic Aneurysm: Table 9)

Treatment of AAA is aimed at preventing the following three risks: (1) rupture of the aneurysm, (2) peripheral embolism originating from the aneurysm, and (3) coagulopathy due to the aneurysm. If there are no signs of impending rupture, medical treatment should be chosen to avoid the risk of rupture. If rupture is highly likely, surgical treatment should basically be prioritized.

**1. Risk Assessment in Abdominal Aortic Aneurysm**

(1) **Risk of Aneurysm Rupture**

1. **Diagnosis and Rate of Enlargement**

As the maximum minor-axis diameter becomes greater, wall tension increases, and so does the risk of rupture. The early mortality rate of more than 5% for surgical cases of TAA or TAAA overall, even today with improved therapeutic efficacy, remains a major issue. If the risk of death after surgery for TAA is assumed to be 5%, a reasonable criterion for surgical indication is an aortic diameter of 50 to 59 mm, when the risk of rupture and aortic dissection during medical treatment are taken into consideration. In cases with descending aortic aneurysm and TAAA, the risk of the complication of paraplegia is high. When the risk of rupture and aortic dissection during medical treatment is taken into consideration, an aortic diameter of about 60 mm seems to be a reasonable criterion for the surgical indication.
29% of patients with AAA.

(2) Risk of Peripheral Embolism Due to Aortic Aneurysm
It has been reported that peripheral embolism occurs in 3 to

IV Medical Treatment

1. Aortic Dissection
1. Management in the Acute Phase
The most important aspects of treatment in the hyperacute phase are control of hypertension (goal, 100 to 120 mmHg), pulse rate control, pain relief, and rest. β-blockers should be used simultaneously with antihypertensive therapy to achieve aggressive pulse rate control, and analgesic or sedative therapy should be used to manage persistent pain.

2. Management in the Chronic Phase
The major goals of patient management in the chronic phase are to prevent re-dissection and rupture and to determine the timing and technique of (any subsequent) surgery.

(1) Blood Pressure Control
The most important aspect of medical management is to control blood pressure. β-blockers are reported to reduce dissection-related events such as hospitalization, and inhibit aneurysm enlargement.

(2) Bed Rest Level and Exercise
Although there may be few or minimal restrictions on routine ADL, evidence on exercise is limited.
(3) Follow-up by Imaging Tests
It is necessary to follow patients at regular intervals performing imaging modalities such as CT and MRI until 2 years after onset, the period associated with frequent dissection-related events. If the aortic diameter approaches that of the surgical indication, CT should be performed at shorter intervals. Considering the radiological exposure and contrast-induced nephropathy, prolongation of the interval between CT tests may be warranted in patients who have a small aortic diameter and a thrombosed false lumen in the absence of ULP.

<table>
<thead>
<tr>
<th>Table 11. Patients Indicated for a Course of Standard Rehabilitation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Selection criteria: Stanford type A non-communicating and Stanford type B aortic dissection</td>
</tr>
<tr>
<td>- Maximum aortic diameter of less than 50mm</td>
</tr>
<tr>
<td>- No organ ischemia</td>
</tr>
<tr>
<td>- Not complicated with DIC (FDP ≥40)</td>
</tr>
<tr>
<td>Exclusion criteria (where this approach should not be used)</td>
</tr>
<tr>
<td>1. Type of dissection for which this approach is not indicated</td>
</tr>
<tr>
<td>2. Type of dissection for which this approach is indicated but where there is a serious complication</td>
</tr>
<tr>
<td>3. Restlessness</td>
</tr>
<tr>
<td>4. Re-dissection</td>
</tr>
<tr>
<td>5. Mediastinal hematoma</td>
</tr>
<tr>
<td>6. Cardiac tamponade, predominantly right-sided pleural effusion</td>
</tr>
<tr>
<td>Setting the goal (discharge criteria)</td>
</tr>
<tr>
<td>1. Blood pressure controlled throughout the day with systolic blood pressure less than 130mmHg</td>
</tr>
<tr>
<td>2. Stable general status with no development of complications</td>
</tr>
<tr>
<td>3. Rehabilitation in bath completed or ADL recovered to the prehospital level</td>
</tr>
<tr>
<td>4. Understanding of what care needs to be taken in daily life (e.g., medication, diet, exercise, visiting doctors)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Table 12. Patients Indicated for a Course of Short-Term Rehabilitation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Selection criteria: Stanford type B aortic dissection</td>
</tr>
<tr>
<td>- Maximum minor-axis diameter ≤40mm</td>
</tr>
<tr>
<td>- No ULP in non-communicating dissection</td>
</tr>
<tr>
<td>- True lumen occupying one quarter or greater in communicating dissection</td>
</tr>
<tr>
<td>- Not complicated with DIC (FDP ≥40)</td>
</tr>
<tr>
<td>Exclusion criteria (where this approach should not be used)</td>
</tr>
<tr>
<td>1. Type of dissection for which this approach is not indicated</td>
</tr>
<tr>
<td>2. Type of dissection for which this approach is indicated but where there is a serious complication</td>
</tr>
<tr>
<td>3. Re-dissection</td>
</tr>
<tr>
<td>Setting the goal (discharge criteria)</td>
</tr>
<tr>
<td>1. Blood pressure controlled throughout the day with systolic blood pressure less than 130mmHg</td>
</tr>
<tr>
<td>2. Stable general status with no development of complications</td>
</tr>
<tr>
<td>3. Rehabilitation in bath completed or ADL recovered to the prehospital level</td>
</tr>
<tr>
<td>4. Understanding of what care needs to be taken in daily life (e.g., medication, diet, exercise, visiting the doctor)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Table 13. In-Hospital Rehabilitation Program</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stage</td>
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<tr>
<td>---</td>
</tr>
<tr>
<td>1</td>
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<tr>
<td>2</td>
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<tr>
<td>3</td>
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<tr>
<td>4</td>
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<tr>
<td>5</td>
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<tr>
<td>6</td>
</tr>
<tr>
<td>7</td>
</tr>
<tr>
<td>8</td>
</tr>
<tr>
<td>To be discharged</td>
</tr>
</tbody>
</table>

ADL, activity of daily living; DIC, disseminated intravascular coagulation syndrome; FDP, fibrin degradation products; ULP, ulcer-like projection.
(4) Cautions in Chronic-Phase Management of Surgical Cases
Remaining dissection and postoperative long-term complications are important issues, regardless of whether the dissection is Stanford type A or type B.

1) Long-Term Postoperative Complications
Major problems are aortic valve insufficiency, anastomotic pseudoaneurysm, re-dissection, and enlargement of the remaining false lumen.

2) Aneurysm Formation via the Remaining Dissection
It is not rare to find enlargement of the remaining distal false lumen in patients after surgery for aortic dissection.

3) Frequency of Reoperation (The Initial Surgery May Be in Either the Chronic Phase or the Acute Phase)
The reoperation rate after surgery for aortic dissection is reported to be 8 to 10%. It has also been reported that reoperation-free rate was 95%, 64%, and 35% of patients at 5, 10, and 15 years, respectively, after the initial surgery.

3. Rehabilitation
The rehabilitation program for cardiovascular diseases is designed separately for Phase I (acute phase and during hospitalization), Phase II (early after discharge and 1 to 2 months after onset), and Phase III (more than 2 months after onset) (Tables 11 to 14).

2. Thoracic Aortic Aneurysm

1. Basic Considerations for Medical Treatment
(1) Management of Atherosclerotic Risk Factors
It is important to provide treatment and management of atherosclerotic risk factors such as hypertension, dyslipidemia, diabetes mellitus, hyperuricemia, obesity, and smoking.

(2) Management of Atherosclerotic Complications
It is common for patients to have atherosclerotic diseases such as cerebrovascular disease, carotid artery disease, coronary artery disease, nephrosclerosis (renal arteriosclerosis), lower limb artery disease, and aortic aneurysms involving other sites. In particular, the frequency of concomitant coronary artery disease is high, indicating the importance of searching for the presence of major arterial lesions throughout the body.

2. Medical Treatment in Non-Surgical Cases
(1) Signs and Symptoms
Although most TAA cases are basically asymptomatic, an increase in the aneurysmal diameter may lead to the manifestation of (1) aortic valve insufficiency due to enlargement of the aortic root or ascending aorta; (2) coughing, shortness of breath, wheezing, recurrent pneumonia originating from compression of the trachea or the main bronchus; (3) dysphagia due to compression of the esophagus; (4) hoarseness due to compression on the recurrent laryngeal nerve; and (5) chest pain or back pain due to compression on surrounding organs or erosion into the ribs.

Because a progressive enlargement in aneurysmal diameter is suggested at the onset of symptoms, CT or MRI (MRA) examinations should be performed promptly.

(2) Blood Pressure Control During Follow-up Observation
The antihypertensive goal in non-surgical TAA cases should be 105 to 120 mmHg for systolic blood pressure, a level lower than in most hypertensive patients. β-blockers are considered to be the first-line treatment. If β-blockers at a maximum dose cannot achieve a sufficient hypotensive response, it is necessary to add other antihypertensive drugs, as needed, to reach the antihypertensive goal.

(3) Restriction of Physical Activity During Follow-up Observation
Patients should be instructed to avoid smoking, excessive drinking and eating, overwork, poor sleep, and mental stress. Patients should also be instructed to be careful when lifting or dragging heavy materials/objects that may result in a rapid blood pressure increase, straining during bowel movements, and persistent coughing.

(4) Imaging Evaluation During Follow-up Observation
Although it is difficult to predict the timing of rupture or the enlargement rate of aneurysmal diameter in TAA cases, the annual incidence of cardiovascular accidents is reported to be 6.5% when the TAA diameter is 50 to 60 mm and 15.6% when the diameter exceeds 60 mm.

It is necessary to evaluate changes in the diameter and morphology by CT or MRI on a regular basis, depending on the aneurysmal diameter and the tendency for enlargement.

3. Medical Treatment in Surgical Cases
(1) Clinical Signs and Symptoms
Because enlargement of the aorta at the non-replacement site or pseudoaneurysm or rupture of an anastomosed artificial graft is suspected when clinical symptoms are observed in patients after surgery, these manifestations should be treated in the same manner as in non-surgical patients.

(2) Blood Pressure Control
The antihypertensive goal should also be under 130 mmHg for systolic blood pressure even in surgical patients.

---

Table 14. Evidence of Acute-Phase Rehabilitation Therapy in Aortic Dissection

<table>
<thead>
<tr>
<th>Class IIa</th>
<th>Standard rehabilitation course for Stanford type B acute aortic dissection (maximum minor-axis diameter &lt;50mm without organ ischemia and FDP &lt;40) (Level of Evidence: B)</th>
</tr>
</thead>
</table>
| Class IIb | 1. Standard rehabilitation course for Stanford type A non-communicating acute aortic dissection (maximum minor-axis diameter <50mm without evidence of ULP in the ascending aorta, without organ ischemia and FDP <40) (Level of Evidence: C)  
2. Short-term rehabilitation course for Stanford type B acute aortic dissection (maximum minor-axis diameter <40mm without organ ischemia). The minimum true lumen exceeds one quarter of the whole lumen in the case of communicating dissection, and no ULP with FDP <40 in the case of non-communicating dissection. (Level of Evidence: C) |

FDP, fibrin degradation products; ULP, ulcer-like projection.
3. Abdominal Aortic Aneurysm

Any aneurysm with a diameter of 50 mm or greater carries a risk of rupture, and priority should be given to surgical treatment except in high-risk surgical patients. In regard to suppression of an extending aortic aneurysm with a diameter of 30 to 50 mm, no clearly effective therapeutic agents have yet been developed (Table 15).

Table 15. Medical Treatment for Abdominal Aortic Aneurysms

<table>
<thead>
<tr>
<th>Class I</th>
<th>Class Ila</th>
<th>Class IIb</th>
<th>Class III</th>
</tr>
</thead>
<tbody>
<tr>
<td>Smoking cessation</td>
<td>Doxycycline (Level of Evidence: B)</td>
<td>Statins (Level of Evidence: B)</td>
<td>Propranolol (Level of Evidence: A)</td>
</tr>
<tr>
<td>(Level of Evidence: B)</td>
<td>Roxithromycin (Level of Evidence: B)</td>
<td>ACE inhibitors (Level of Evidence: B)</td>
<td></td>
</tr>
</tbody>
</table>

ACE, angiotensin converting enzyme.

Table 16. Classification of Evidence Level: AHCPR (1993)

<table>
<thead>
<tr>
<th>Classification</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ia</td>
<td>Systematic reviews/meta-analyses</td>
</tr>
<tr>
<td>Ib</td>
<td>Randomized controlled trials</td>
</tr>
<tr>
<td>Ila</td>
<td>Non-randomized controlled trials</td>
</tr>
<tr>
<td>IIb</td>
<td>Other quasi-experimental studies</td>
</tr>
<tr>
<td>IIa</td>
<td>Non-experimental descriptive studies (e.g., comparative studies, correlation studies, case control studies)</td>
</tr>
<tr>
<td>IV</td>
<td>Opinions of expert committees and respected authorities</td>
</tr>
</tbody>
</table>

AHCPR, Agency for Health Care Policy and Research (presently, Agency for Healthcare Research and Quality [AHRQ]).

3. Abdominal Aortic Aneurysm

(3) Restriction of Physical Activity

Although artificial grafts are considered to have sufficient strength at the site of replacement, mild restriction of physical activity seems to be necessary, essentially as in non-surgical patients, for the purpose of avoiding aneurysm formation in the area without aortic replacement or in the anastomosed part.

(4) Imaging Evaluation

It is preferable that patients be evaluated for postoperative conditions by CT or MRI examinations at 3 to 6 months after surgery, and then annually thereafter.

V Surgical Treatment

1. Thoracic Aorta

1. Overview of Surgical Treatment of Thoracic Aortic Aneurysms

Surgical resection and replacement of the thoracic aorta is the gold standard for treatment of TAA and Stanford type A aortic dissection.

2. Fundamental Surgical Techniques and Surgical Adjuvant Treatments for Thoracic Aortic Surgery

(1) Aortic Root or Ascending Aorta Replacement

1) Standard Surgical Techniques

Surgical techniques for the aortic root are broadly classified into those based on replacement of the valve, such as valved graft (Bentall operation), homologous aorta (homograft), heterologous aorta, autologous pulmonary valve (Ross operation), and those preserving the autologous valve (aortic valve sparing surgery).

2) Reconstruction of Coronary Artery

The button-Bentall operation has become common in recent years. When mobilization of the coronary artery involves risks or is not possible because of reoperation, inflammation, among others, Piehler operation by interposing an artificial graft is effective.\(^\text{101,102}\)

3) Aortic Valve Sparing Surgery

This technique is broadly classified into the remodeling method\(^\text{103}\) and the reimplantation method.\(^\text{104}\) Aortic valve sparing surgery is more advantageous than the Bentall operation in that there is no need for anticoagulant therapy, leading to a decrease in prosthesis-related complications. Issues of concern include increased surgical risk due to prolonged aortic cross-clamping, and unclear long-term durability of the autologous aortic valve. To date, evidence strongly supporting recommendation of either Bentall operation or aortic valve sparing surgery is lacking. The Bentall operation is currently the standard procedure\(^\text{105}\) (Class IIb, level of evidence by Agency for Health Care Policy and Research [AHCPR], Table 16, the same as below).

(2) Aortic Arch Replacement

1) Standard Surgical Techniques

Access to aortic aneurysms in the arch or distal arch is generally achieved by a median sternotomy.\(^\text{106}\) On the other hand, left thoracotomy is used for distal aortic arch aneurysms mainly extending to the peripheral side. In cases with an extensive aortic aneurysm, an elephant trunk\(^\text{107}\) should be inserted prior to the second-stage treatment (surgery or stent-grafting).\(^\text{108}\)

2) Brain Protection

Although hypothermic circulatory arrest (HCA)\(^\text{109}\) is the basic adjuvant procedure during arch reconstruction, antegrade selective cerebral perfusion (SCP)\(^\text{110}\) or retrograde cerebral perfusion (RCP)\(^\text{111}\) has been added for the purpose of achieving longer and safer brain protection, resulting in better outcomes (Class IIb).\(^\text{112}\) For left thoracotomy, HCA or HCA + RCP/SCP...
should be used, and the arch should be reconstructed by the open proximal technique.113

(3) Descending Thoracic or Thoracoabdominal Aortic Replacement

1) Standard Surgical Techniques

In cases of descending aortic replacement, the descending aortic aneurysm is usually reached via a left thoracotomy through the 5th to 6th intercostal space. The thoracotomy through the 4th to 5th intercostal space may be used in cases with a proximal descending aortic aneurysm, and the thoracotomy through the 7th to 8th intercostal space may be used in cases with a distal descending aortic aneurysm near the diaphragm. In addition, TAAA should be reached via a thoracotomy through the 5th to 6th intercostal space and a spiral incision extending to the abdominal region. The intercostal artery and abdominal branch should be reconstructed individually, using small-caliber artificial grafts measuring 8 or 10 mm in diameter, or reconstructed in an island shape by an en bloc technique. Individual reconstruction is performed for Marfan syndrome.114

2) Adjuvant Procedures

In general, the distal perfusion method by partial cardiopulmonary bypass (femoro-femoral [F-F] bypass)115 or left heart bypass116,117 is used for protection of the spinal cord and abdominal organs, although reconstruction can be performed under simple cross-clamping.118 For patients in whom proximal cross-clamping near the arch is difficult or in whom lysis of adhesion for prior surgery is difficult, the HCA method under total cardiopulmonary bypass is used.119,120

3) Spinal Cord Protection

The Adamkiewicz artery should be identified by MRI or CT prior to surgery, as the findings can facilitate determining re-

4) Abdominal Organ Protection

Selective continuous perfusion of abdominal branches should be performed using a balloon-tipped cannula via partial cardiopulmonary bypass or collateral of the left heart bypass circuit.

3. Aortic Dissection

(1) Surgical Strategies for Acute Aortic Dissection (Figure 10)

Tables 3 and 4 show the current, generally accepted, surgical indications for aortic dissection.

In the acute phase, emergency surgery is basically indicated for Stanford type A dissection, whereas medical antihypertensive therapy is indicated for Stanford type B dissection. Prompt treatment is essential for complications of dissection. Cases complicated by extensive brain damage including coma are often excluded from surgical indications because irreversible brain damage is commonly present.126 However, whether the brain damage is reversible or irreversible should be determined with due caution.127

(2) Practical Aspects of Surgery for Acute Aortic Dissection

1) Principles of Surgery

Artificial graft replacement of the aorta including entry should be performed. Techniques are described below.

(1) Ascending aortic replacement: Extracorporeal circulation by blood perfusion from the femoral artery,128 the true lumen of the ascending aorta,129 or the axillary artery130 is used as an adjuvant procedure. Multiple routes of blood perfusion may be used when there is concomitant organ malperfusion. The current standard adjuvant procedure is deep HCA, by which the core temperature is reduced to 20 degrees C or lower.131 RCP may also be used concomitantly. For peripheral anastomosis, an open distal anastomosis is used under deep HCA without aortic cross-clamping.

(2) Total arch replacement: When the tear is present in the aortic arch, ascending and total arch replacement is preferable in principle, from the viewpoint of tear resection.132 In Marfan syndrome cases with Stanford type A aortic dissection, total arch replacement is indicated because hemi-arch replacement may result in enlargement of the remaining aortic arch.133,134 The elephant trunk method is used,135 with the aim of adding strength from the intimal side, preventing anastomosis leakage, and closing the peripheral

![Figure 10. Surgical strategies for acute aortic dissection. AR, aortic regurgitation; Asc, ascending aorta; CT, computed tomography; TEVAR, thoracic endovascular aneurysm repair; TTE, transthoracic echocardiography.](Image 60x484 to 273x726)
false lumen. In addition, in recent years, hybrid surgery combining total arch replacement with insertion of the frozen elephant trunk into the descending aorta has been increasing.\textsuperscript{136–138}

(3) Stanford type B dissection: Most acute type B dissections indicated for surgery are accompanied by rupture or seriously impaired organ perfusion. In such cases, surgical results are inevitably poor. The mainstay of treatment for complicated lesions of acute type B dissection is shifting to an endovascular approach.\textsuperscript{139–141}

2) Aortic Valve Regurgitation

(1) Aortic valve resuspension: Except in cases with annuloaortic ectasia (AAE) or organic aortic valve disease, resuspension of the aortic commissure is effective.\textsuperscript{142,143}

(2) Aortic root replacement: When the tear extends deep into the Valsalva sinus or when there is already AAE, the Bentall operation has conventionally been indicated.\textsuperscript{144} Although this operation is still the standard procedure, aortic valve sparing surgery has also recently been attempted.\textsuperscript{145,146}

3) Branch Malperfusion

Branch malperfusion is the main cause of increased complexity and severity of the disease state in cases with acute aortic dissection; this condition manifests with various symptoms in 20 to 40% of patients.\textsuperscript{147} As a rule, if aortic dissection shows an unstable course, aortic repair is the first priority, and intervention for peripheral vascular disease should be secondary.\textsuperscript{148} The results of aortic dissection repair in patients who have a concomitant branch malperfusion are poor, and the early mortality rate is reported to be 30 to 50%.

(3) Chronic Aortic Dissection

There are certain points to keep in mind when dealing with chronic dissection cases, and strategies tailored to individual patients are necessary.

1) Stanford Type A Dissection

The site of blood perfusion should be chosen chiefly from among the axillary artery,\textsuperscript{149,150} femoral artery and the cardiac apex,\textsuperscript{151} and the choice should be appropriate to each case.

(1) Repair of Aortic Valve Regurgitation

i) Aortic Valve Replacement

In cases with chronic aneurysms accompanied by aortic valve regurgitation, aortic valve sparing surgery involving aortic valvuloplasty should be performed. If this procedure is difficult, aortic valve replacement should be performed instead.

ii) Aortic Root Replacement With a Valved Artificial Graft

When aortic valve sparing surgery is difficult in patients with AAE complicated by dissection, aortic root replacement with a valved artificial graft should be performed.

iii) Aortic Valve Sparing Surgery

Among patients with AAE or dissection extending to the aortic root and requiring aortic root replacement, aortic valve sparing surgery is performed in patients with normal aortic valve leaflets or indicated for valvuloplasty.

(2) Aortic Reconstruction With an Artificial Graft

For ascending aortic replacement, “open distal anastomosis”, by which peripheral anastomosis is performed under circulatory arrest, is a common procedure. Although SCP is generally a safe method of brain protection, the most appropriate protective method is chosen according to each case. Although anastomosis to the true lumen is preferable for avoiding reoperation of the distal aorta in the remote phase, the possibility of organ ischemia should also be taken into consideration.

Total arch replacement is indicated for retrograde dissection with a tear in the descending aorta, rupture or enlargement of the aortic arch, occlusion of an arch branch, Marfan syndrome, among others. In general, open distal anastomosis assisted by SCP is performed. It is common to insert an elephant trunk into the descending aorta after resecting the flap and to perform the double-barreled anastomosis.

When staged surgery is not possible in extensive aneurysm cases, ascending, arch and descending aortic replacement should be performed via bilateral anterior thoracotomy or median sternotomy (+ left thoracotomy) as a single-stage procedure.\textsuperscript{152,153}

2) Stanford Type B Dissection

i) Descending Aortic Replacement

The enlarged descending aorta should be replaced with an artificial graft via left thoracotomy and distal aortic perfusion (e.g., F-F bypass, left heart bypass). To avoid retrograde dissection associated with proximal cross-clamping, not a few surgeons choose the technique of open proximal anastomosis for descending replacement. The technique of true lumen anastomosis or double-barreled anastomosis should be chosen as the distal anastomosis technique, depending on the individual case, taking into account peripheral blood flow.\textsuperscript{154} If the Adamkiewicz artery is fed by the false lumen, double-barreled anastomosis or reconstruction of the same artery is preferable.

ii) Partial Arch or Descending Aortic Replacement by Open Proximal Anastomosis

This procedure is indicated for patients in whom the intimal tear is located in the aortic arch and those in whom extension has reached the arch, as well as those in whom proximal cross-clamping is not possible. Although this surgery can be implemented with retrograde perfusion from the femoral artery alone, the route of blood perfusion to the proximal aorta is also often secured. After circulatory arrest is induced at 18 degrees C or lower, the proximal aorta is transected while the aortic arch is opened, and anastomosed to the artificial graft.

iii) Thoracoabdominal Aortic Replacement

To prevent spinal cord ischemia, following procedures are usually undertaken: (1) distal aortic perfusion and maintenance of blood pressure at a relatively high level; (2) implementation of CSF drainage; (3) prevention of steal phenomenon from the intercostal artery and lumbar artery while the aorta is opened; (4) identification of the preoperative spinal cord feeding artery and its reconstruction; (5) staged segmental aortic clamping;\textsuperscript{155} (6) pharmacologic spinal cord protection; (7) moderate hypothermia. Surgery must be planned with special attention to items (3) and (5). For reconstruction of the abdominal major arterial branches, selective perfusion of the celiac artery, the superior mesenteric artery, and the renal arteries should be performed, and cold Ringer’s lactate solution can be applied for renal arteries reconstruction only.\textsuperscript{156}

4. Results of Surgical Treatment for Aortic Dissection and True Aortic Aneurysm

(1) Current Status and Efficacy of Surgical Treatment for Aortic Dissection

Advances in diagnostic imaging have allowed early diagnosis of aortic dissection immediately after onset, and better thera-
2. Abdominal Aorta

1. Unruptured Abdominal Aortic Aneurysm
   (1) Surgical Indications (Table 9)

(2) Preoperative Evaluation

   1) Evaluation of the Aortic Aneurysm and Concomitant Aneurysms at Other Sites

   2) Assessment of Surgical Risk

   3) Evaluation of Ischemic Heart Disease

(3) Surgical Techniques

1) Approach

Transperitoneal approach or retroperitoneal approach is chosen according to each patient’s condition.

2) Aortic Cross-Clamping

3) Choice of Artificial Grafts

4) Maintenance of Pelvic Blood Flow

Surgery for AAA may be associated with impaired inferior mesenteric artery and internal iliac artery blood flows, leading to ischemia in the sigmoid colon or rectum, gluteal claudication, sexual dysfunction, and spinal cord ischemia. The occurrence of these conditions, however, is influenced by a number of factors. There is no distinct evidence concerning the presence/absence of reconstruction of the inferior mesenteric artery and internal iliac artery.

Table 17. Results of Surgery for Non-Dissecting Thoracic or Thoracoabdominal Aneurysms: The Japanese Association for Thoracic Surgery (2008)

<table>
<thead>
<tr>
<th>Replaced site</th>
<th>No. of cases</th>
<th>Hospital mortality</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ascending aorta</td>
<td>836</td>
<td>20 (2.4)</td>
</tr>
<tr>
<td>Aortic root</td>
<td>677</td>
<td>22 (3.2)</td>
</tr>
<tr>
<td>Ascending aorta + aortic arch</td>
<td>1,790</td>
<td>113 (6.3)</td>
</tr>
<tr>
<td>Aortic arch + descending aorta</td>
<td>165</td>
<td>13 (7.9)</td>
</tr>
<tr>
<td>Aortic root + ascending aorta + aortic arch</td>
<td>86</td>
<td>6 (7.0)</td>
</tr>
<tr>
<td>Descending aorta</td>
<td>452</td>
<td>22 (4.9)</td>
</tr>
<tr>
<td>Thoracoabdominal aorta</td>
<td>348</td>
<td>35 (10.1)</td>
</tr>
<tr>
<td>Bypass grafting</td>
<td>11</td>
<td>0 (0.0)</td>
</tr>
<tr>
<td>Stent-grafting</td>
<td>952</td>
<td>33 (3.5)</td>
</tr>
<tr>
<td>1) Transcatheter approach</td>
<td>730</td>
<td>20 (2.7)</td>
</tr>
<tr>
<td>2) Open stent-grafting</td>
<td></td>
<td></td>
</tr>
<tr>
<td>a) With arch replacement</td>
<td>74</td>
<td>0 (0.0)</td>
</tr>
<tr>
<td>b) Without arch replacement</td>
<td>142</td>
<td>13 (9.2)</td>
</tr>
<tr>
<td>3) Unspecified</td>
<td>6</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>5,317</td>
<td>264 (5.0)</td>
</tr>
</tbody>
</table>

Adapted from Gen Thorac Cardiovasc Surg 2010; 58: 356–383.

2. Ruptured Abdominal Aortic Aneurysm

(1) Diagnosis

Discussed in Chapter II.

(2) Treatment

The patient should be transferred to the operating room as soon as possible, and bleeding should be controlled by cross-clamping of the aorta either above the celiac artery or below the renal artery depending on the status of the hematoma.

(3) Therapeutic Results

The mortality rate is 40 to 70% among patients who arrive at the hospital. Circulatory failure may cause the complications of multiple organ failure, respiratory failure, renal failure, and colonic ischemia.
VI Endovascular Treatment

1. Aortic Dissection (Recommendations for Endovascular Treatment of Aortic Dissection: Table 18)

Endovascular treatment of aortic dissection includes transcatheter fenestration, stent placement, and stent-graft insertion. Although closure of the entry site with stent-grafting is currently the mainstay of treatment, its indications, the device to be used, and the placement technique still vary among institutions.

1. Transcatheter Fenestration, Stent Placement for Stenotic or Occluded True Lumen or Branch Vessels

(1) Indications
These procedures are performed mainly for the treatment of malperfusion syndrome associated with acute dissection.

(2) Methods
1) Transcatheter Fenestration
A guide wire should be passed from the true lumen into the false lumen, and the flap is then split by inflating the balloon to create a reentry site.

2) Stent Placement
A stent should be inserted into the lumen of the ischemic branch artery, and placed therein.

2. Entry Closure by Stent-Graft Insertion

(1) Indications
Although basically this technique is indicated for patients with chronic type B dissection in whom conventional open thoracotomy is considered to be a high-risk procedure, or those with acute type B dissection accompanied by rupture, organ ischemia, among others. Because surgical results are poor in type A aortic dissection with the primary entry site in the descending aorta, ever more specialists are taking the view that this condition is treatable, depending on the patient’s condition, even in the acute phase.

(2) Methods
See the section on treatment of true aortic aneurysm (Section 2.3 of Chapter VI).

(3) Results
It has been reported that the initial success rate was 70.8 to 94.4%, the incidence of endoleak was 2.8 to 19%, and the early mortality rate was 2.7 to 13%. Reports of long-term results are as yet limited, however.

2. Thoracic Aortic Aneurysm (Recommendations for Stent-Graft Treatment of Thoracic Aortic Aneurysm: Table 19)

1. Introduction
Thoracic stent-grafts provided by manufacturers were approved in Japan in March 2008, and their use is now spreading rapidly. Evidence of the efficacy of this treatment in TAA is still limited in Japan, and therefore this revision of the guidelines reflects mainly evidence obtained in Western countries.

2. Indications
Indications in various diseases are described below. Due caution is necessary when using stent-grafts provided by manufacturers, because each device has its own anatomical criteria.

Table 18. Endovascular Treatment for Aortic Dissection

<table>
<thead>
<tr>
<th>Class</th>
<th>Indications</th>
</tr>
</thead>
</table>
| I     | 1. Chronic follow-up after endovascular treatment (including imaging diagnosis) (Level of Evidence: C)  
       | 2. Support from a surgical team (Level of Evidence: C)  
       | 3. Entry closure with stent-grafting for complicated acute type B aortic dissection (Level of Evidence: C) |
| IIa    | 1. Stent placement for ischemic branch vessels resulting from compression of the true lumen due to aortic dissection (Level of Evidence: B)  
       | *Early treatment after the onset of dissection is important in acute cases.*  
       | 2. Transcatheter fenestration early after the onset of acute type B aortic dissection with blocked true lumen (Level of Evidence: B)  
       | 3. Entry closure with stent-grafting for chronic type B aortic dissection where surgery is indicated (Level of Evidence: B)  
       | 4. Entry closure with stent-grafting for acute type A aortic dissection caused by retrograde dissection (Level of Evidence: B) |
| IIb    | 1. Entry closure with stent-grafting for patients with chronic type B aortic dissection having high surgical risk (Level of Evidence: B)  
       | 2. Stenting for a narrowed segment of the true lumen in acute aortic dissection (Level of Evidence: C)  
       | 3. Entry closure with stent-grafting for acute type B aortic dissection to prevent formation of future aneurysms (Level of Evidence: C) |
| III    | 1. Use in patients who do not meet anatomical criteria (Level of Evidence: B)  
       | 2. Entry closure with stent-grafting for acute type B aortic dissection where branch vessels are obviously ischemic due to static compression (Level of Evidence: C)  
       | 3. Entry closure with stent-grafting for chronic type B aortic dissection where main branch vessels are perfused from the false lumen (Level of Evidence: C)  
       | *When transcatheter fenestration is performed simultaneously with or prior to stent-grafting, Class II b and Level of Evidence: C.* |

It is a prerequisite that the above procedures are conducted in hospitals skilled in endovascular treatment.
Table 19. Stent-Graft Treatment for Thoracic Aortic Aneurysms and Aortic Dissection

| Class I | 1. Support from a surgical team (Level of Evidence: C)  
| Class IIA | 1. Descending aortic aneurysm with high surgical risk* (Level of Evidence: B)  
| Class IIB | 1. Descending aortic aneurysm with low surgical risk* (Level of Evidence: B)  
| Class III | 1. Intervention for asymptomatic thoracic aortic aneurysm 55 mm or less in diameter (Level of Evidence: C)  

*Only when there is an anatomical indication.

for the procedure.

(1) Ascending Aortic Aneurysm
At present, endovascular treatment is not indicated for ascending aortic aneurysm.

(2) Aortic Arch Aneurysm
Endovascular treatment for aortic arch aneurysm should be performed only in patients of advanced age or with high surgical risk. Currently, the use of stent-grafts in this lesion is centered on hybrid approaches combining extra-anatomical bypass surgery for arch branches. 

The mortality rate and incidence of complications after open stent-graft placement (frozen elephant trunk technique, stented elephant trunk technique) are equivalent or superior to those after routine surgery. Therefore, this procedure should be accepted as a treatment option in this lesion. In particular, this procedure is useful for treating extensive aortic arch aneurysm or type A aortic dissection.

(3) Descending Aortic Aneurysm
Stent-graft treatment for descending aortic aneurysm is associated with reduced acute-phase mortality and a lower incidence of adverse events than surgical treatment, and shows favorably maintained conditions in the intermediate phase so long as anatomical situations agree with the indications for the device. Therefore, this treatment should be considered as the first-line therapy in high-risk surgical patients (Class Iia, Level of Evidence: B). In addition, because the spinal cord injury incidence is also lower, and QOL is thus more likely to be maintained than with surgical treatment in low-risk surgical patients, many are of the opinion that this treatment should be regarded as the first-line therapy (Class Iib, Level of Evidence: C).

(4) Thoracoabdominal Aortic Aneurysm
Endovascular treatment for TAAA requiring reconstruction of abdominal major branches is indicated for high-risk patients or those in whom surgery is difficult. However, stent-graft treatment for TAAA that secures enough landing zone by placing it just above the branch of the celiac artery or placing it to cover the celiac artery with good collateral circulation should be regarded as equivalent to surgery, as same as in cases with a descending aortic aneurysm.

(5) Traumatic Aortic Injury
Stent-graft treatment is the first-line therapy for traumatic aortic injuries, particularly those involving the arterial ligament or the descending aorta (Class I, Level of Evidence: B).

(6) Ruptured Thoracic Aortic Aneurysm
The results of thoracic endovascular aneurysm repair (TEVAR) for ruptured descending TAA are more favorable than those of surgical treatment. Therefore, TEVAR is recommended for ruptured descending TAA if the lesion is located at a site appropriate for this treatment (Class IIIa, Level of Evidence: B).

3. Methods of Stent-Graft Treatment
(1) TEVAR
TEVAR is focused on two main issues: “how to safely deliver the stent-graft to the target site and place it there” and “how to set a favorable landing zone”.

1) Access
Because the catheter sheath used for TEVAR is thick (20 to 27 Fr), it is more likely for iliac injury to occur during the approach from the femoral artery. Because the spinal cord injury incidence is also lower, and QOL is thus more likely to be maintained than with surgical treatment in low-risk surgical patients, many are of the opinion that this treatment should be regarded as the first-line therapy (Class I, Level of Evidence: B).

2) Landing Zone for Stent-Graft Placement
The most important aspect of stent-graft treatment is to obtain the landing zone for the stent on the proximal and distal sides of the aneurysm.

i) Left Subclavian Artery Coverage, Celiac Artery Coverage
When covering the left subclavian artery, ischemic symptoms of the left upper limb and the brain do not appear in most cases, but it has been pointed out that cerebral infarction and spinal cord injury are frequent complications. In cases requiring left subclavian artery coverage, the patency of the right subclavian artery and right vertebral artery and communication between the right and left vertebral arteries at the basilar artery level should be confirmed. When the patency and communication of these vessels cannot be confirmed, bypass to the left subclavian artery is considered to be indispensable.

When covering the celiac artery, it is rare to have complications or problems involving the blood flow to the abdominal organs or intestinal tract because there is perfusion from the
graft migration when it drifts out of its original location due to blood flow. Therefore, balloon dilatation for touch-up of the stent-graft is often performed under some form of blood flow control. These blood flow control measures include (1) pharmacological hypotension, (2) pharmacological cardiac arrest using adenosine triphosphate (ATP), among others, (3) rapid pacing, (4) inferior (or superior) vena cava occlusion. Measures (2) to (4) are theoretically justified because the lower the blood flow through the balloon-dilated portion, the lower the resistance to the balloon itself.

collateral circulation via the superior mesenteric artery in most cases. However, an increased incidence of spinal cord injury has frequently been reported, similar to cases of left subclavian artery coverage.

ii) Extensive Coverage of the Intercostal Artery
This is closely related to spinal cord injury and will be described in the section on complications.

3) Touch-up of the Stent-Graft
Although it is common for touch-up with a balloon to be performed after stent-graft deployment, the balloon may cause

Figure 11. Hybrid procedures with thoracic stent-grafting. (A) Left common carotid artery-to-left subclavian artery bypass grafting + TEVAR (debranching of one vessel + TEVAR). (B) Right axillary artery-to-left common carotid/left axillary artery bypass grafting + TEVAR (debranching of two vessels + TEVAR). (C) Right common carotid artery-to-left common carotid/left subclavian artery bypass grafting + TEVAR (debranching of two vessels + TEVAR). (D) Ascending aorta-to-left common carotid/left axillary artery bypass grafting + TEVAR (debranching of two vessels + TEVAR). (E) Ascending aorta-to-brachiocephalic/left common carotid/left axillary artery bypass grafting + TEVAR (debranching of three vessels + TEVAR). (F) Iliac artery-to-iliac/superior mesenteric/left and right renal artery bypass grafting + TEVAR. (G) Open stent-grafting (with surgery on ascending aorta and aortic arch). The ascending aorta and branches of the aortic arch are reconstructed using four-branch artificial graft. A stent-graft is inserted into the descending aorta with only the distal end fixed in the vessel, and the artificial graft of the aortic arch and the stent-graft inserted in the descending aorta are sutured at the aortic arch. TEVAR, thoracic endovascular aneurysm repair.
### Table 20. Acute and Mid-Term Results of Endovascular Treatment for Thoracic Aortic Aneurysm

<table>
<thead>
<tr>
<th>Thoracic aortic aneurysm</th>
<th>Acute results</th>
<th>Mid-term results</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Mortality</td>
<td>Complications</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Elective</td>
<td>5% (1.5 to 10.4%)</td>
<td>Brain damage: 3 to 5%</td>
</tr>
<tr>
<td>Emergent</td>
<td>12% (3.8 to 40.9%)</td>
<td>Spinal cord injury: 0 to 5%</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Respiratory failure: 3 to 8%</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Renal failure: 2 to 5%</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Arterial injury, bleeding, complications: 2 to 6%</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Endoleak: 4 to 15%</td>
</tr>
</tbody>
</table>

### Table 21. Summary of Follow-up Results of Patients With Thoracic Aortic Aneurysm and Dissection Produced by the Japanese Committee for Stentgraft Management (As of December 2010)

<table>
<thead>
<tr>
<th>Patient characteristics (n)</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Patients</td>
<td>688</td>
</tr>
<tr>
<td>Male/female</td>
<td>534/154</td>
</tr>
<tr>
<td>Age-year</td>
<td>72.8±9.1 (18 to 96)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Thoracic aortic aneurysm</td>
<td>586</td>
</tr>
<tr>
<td>True aneurysm</td>
<td>523 (49 in the arch, 464 in the descending aorta, 10 in the thoracoabdominal aorta)</td>
</tr>
<tr>
<td>Pseudoaneurysm</td>
<td>63 (4 in the arch, 58 in the descending aorta, 1 in the thoracoabdominal aorta)</td>
</tr>
<tr>
<td>Aortic dissection</td>
<td>102</td>
</tr>
<tr>
<td>Type A dissection</td>
<td>15</td>
</tr>
<tr>
<td>Type B dissection</td>
<td>87</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Anesthesia and the site of sheath insertion</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Anesthesia</td>
<td>General 642 (93.3%)</td>
</tr>
<tr>
<td></td>
<td>Epidural 20 (2.9%)</td>
</tr>
<tr>
<td></td>
<td>Local 22 (3.2%)</td>
</tr>
<tr>
<td></td>
<td>Others 4 (0.6%)</td>
</tr>
<tr>
<td>Sheath insertion</td>
<td>Thoracic aorta¹ 7 (1.0%)</td>
</tr>
<tr>
<td></td>
<td>Abdominal aorta/iliac artery² 154 (22.4%)</td>
</tr>
<tr>
<td></td>
<td>Femoral artery 527 (76.6%)</td>
</tr>
<tr>
<td>Guide wire</td>
<td>Pull-through technique 137 (19.9%)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Results immediately after the procedure</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Duration of fluoroscopic examination³</td>
<td>31.8±19.6 minutes</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Adverse events</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Graft migration</td>
<td>6 (0.9%)</td>
</tr>
<tr>
<td>Bleeding (requiring transfusion)</td>
<td>57 (8.3%)</td>
</tr>
<tr>
<td>Blood vessel injury</td>
<td>41 (6.0%)</td>
</tr>
<tr>
<td>Thoracic aorta</td>
<td>4</td>
</tr>
<tr>
<td>Brachial artery</td>
<td>2</td>
</tr>
<tr>
<td>Right subclavian artery</td>
<td>1</td>
</tr>
<tr>
<td>Iliac artery</td>
<td>30</td>
</tr>
<tr>
<td>Femoral artery</td>
<td>4</td>
</tr>
<tr>
<td>Arterial stenosis or occlusion</td>
<td>16 (2.3%)</td>
</tr>
<tr>
<td>Branches of the aortic arch</td>
<td>5</td>
</tr>
<tr>
<td>Renal artery</td>
<td>3</td>
</tr>
<tr>
<td>Iliac-femoral artery</td>
<td>7</td>
</tr>
<tr>
<td>Bypass graft</td>
<td>1</td>
</tr>
<tr>
<td>Ruptured aneurysm</td>
<td>0 (0.0%)</td>
</tr>
<tr>
<td>Trouble with device</td>
<td>0 (0.0%)</td>
</tr>
<tr>
<td>Death (intraoperative)³</td>
<td>1 (0.2%)</td>
</tr>
</tbody>
</table>

¹Use of artificial graft in 4 patients. ²Use of artificial graft in 13 patients. ³Number of patients analyzed: 473. ⁴Oclusion of the superior mesenteric artery (aortic dissection at the time of removing the sheath under thoracotomy).
4) Devices
As of December 2010 in Japan, two types of devices for the treatment of TAA were approved and in use. There were three more devices to treat TAA that were under clinical trial or were in the application approval process. The latter three include a device that has fenestrations for coping with arch branches and another designed for the open stent-graft procedure.

(2) Hybrid Procedure
Concerning the major branches of the aortic arch and abdominal aorta that must be covered to obtain the landing zone, there are reports describing methods for stent-graft insertion after bypass revascularization through an extra-anatomical route.\(^215\)–\(^219\),\(^237\)–\(^240\) (Figure 11).

Stent-graft treatment combined with bypass is not associated with marked differences in the mortality rate or the incidence of cerebral complications in cases with lesions involving the arch, in comparison with routine surgery. On the other hand, in the thoracoabdominal lesion, it has often been reported that the incidence of paraplegia, the foremost concern, is low.\(^261\)–\(^263\)

(3) Open Stent-Grafting
Open stent-grafting (Figure 11G), by which the descending aortic suture (distal anastomosis) is substituted by fixation with the stent-graft in aortic arch surgery, requires extracorporeal circulation, and is thus not a minimally invasive treatment. However, simplifying the anastomosis of the descending aorta does allow left thoracotomy to be avoided and reduces the time required for extracorporeal circulation. In addition, it is advantageous in that the scope of artificial graft implantation can be extensive. It has been reported that the outcomes in terms of the remaining false lumen are favorable when this technique is applied to aortic dissection.\(^225\)–\(^227\) On the other hand, a number of reports have indicated that the spinal cord injury incidence to be higher than with routine surgery.\(^136\),\(^264\)

4. Results and Complications of Stent-Graft Treatment and Measures Against These Complications

(1) Results

1) Initial Therapeutic Results
Table 20 shows the therapeutic results of stent-graft treatment for TAA in the initial and intermediate phases. As to the results of the initial treatment in Japan, Table 21 presents data on TAA cases just after stenting and during follow-up, based on statistics provided by the Japanese Committee for Stent-graft Management.

2) Mid-Term Results
The survival rate after stent-graft treatment for TAA is 40 to 87% at 5 years, which is comparable to that for surgical treatment.\(^265\)–\(^267\)

(2) Complications

1) Endoleak
Endoleak is the major problem in stent-graft treatment for aortic aneurysm. Endoleak is classified into types I to V according to its cause (Figure 12). Type I and III have a clearly poor prognosis. These types of endoleak require appropriate treatment.

2) Spinal Cord Paralysis
The risk factors for the occurrence of spinal cord injury related to stent-graft treatment are (1) extensive intercostal artery occlusion, (2) past history of surgery for AAA (and coverage of the internal iliac artery), and (3) left subclavian artery coverage. In patients who have more than one of these risk factors, preventive measures including spinal drainage are likely to be indispensable.

3. Abdominal Aortic Aneurysm (Recommendations for Stent-Graft Treatment for Abdominal Aortic Aneurysm: Table 22)

1. Introduction
Stent-grafts provided by manufacturers began to be covered by National Health Insurance (NHI) in April 2007 in Japan, and their use is rapidly becoming widespread.

2. Surgical Indications for Endovascular Aneurysm Repair (EVAR)

1) Indications for Treatment
According to the Society for Vascular Surgery (SVS) practice guidelines (The care of patients with an abdominal aortic aneurysm),\(^268\) this treatment is indicated for fusiform type or asymptomatic aneurysms measuring at least 55 mm in the maximum minor-axis diameter. In the ACC/AHA 2005 Practice Guidelines for the management of patients with peripheral arterial disease,\(^269\) the corresponding criterion is 55 mm or greater in cases with asymptomatic infrarenal AAA; cases with smaller AAA are subject to follow-up observation. Currently,
Although EVAR also shows long-term efficacy, data indicate (2) Long-Term Results demonstrated favorable results of EVAR in the initial and intermediate phases.

Randomized Endovascular Aneurysm Management) trial 275 resulted in underestimation of the value of EVAR. However, the majority of cases requiring re-intervention after EVAR are suitable candidates for minimally invasive treatment with catheterization. Therefore, future improvement in the device may reduce the re-intervention rate for EVAR.

(3) EVAR in High-Risk Patients
The EVAR trial 276 examined whether EVAR reduces the mortality rate in patients with poor general status in whom conventional OSR cannot be performed. There were no significant differences in all-cause mortality or aneurysm-related mortality between patients treated by EVAR and those who were followed without EVAR. However, the results would have been different if this study had been performed more recently with improved devices or techniques. In addition, many of the patients assigned to the non-intervention group later underwent EVAR or OSR, and yet were dealt with as the non-intervention group performing intention-to-treat (ITT) analysis. This resulted in underestimation of the value of EVAR.

Later analyses demonstrated the safety and usefulness of this procedure in patients with a number of complications. 78,276–282

4. Present Status and Results of EVAR in Japan
After approval of devices provided by manufacturers in 2006, EVAR became increasingly common with each passing year. According to the statistics of the Japanese Society for Vascular Surgery, EVAR accounted for only about 2.7% of all treatments for AAA in 2006, but this rate had increased to 10.7% in 2007 and 24.1% in 2008.

According to the follow-up study by the Japanese Committee for Stentgraft Management, there were 1,743 registered cases treated by EVAR during the 2 years between July 1, 2006, and June 30, 2008. Data for the immediate postoperative period included an operative death rate of 0%, and the most frequent adverse event was bleeding requiring blood transfusion, occurring in 3.2% of cases. In the initial postoperative phase, the overall in-hospital mortality rate was 0.5%. The results at 6 months after EVAR were similar, and adverse events were rare, with an aneurysm-related mortality rate of 0.2% (http://stentgraft.jp/pro/result/).

5. Practical Aspects of EVAR
(1) Preoperative Evaluation
Close examination of the heart is not necessary before EVAR.

### Table 22. Stent-Graft Treatment for Abdominal Aortic Aneurysms

<table>
<thead>
<tr>
<th>Class I</th>
<th>Indicated where anatomical criteria of the proximal neck are met (Level of Evidence: A)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Class Ia</td>
<td>Support from a surgical team (Level of Evidence: C)</td>
</tr>
<tr>
<td>Class Ib</td>
<td>Performance of the procedure in a room equipped with a fluoroscope which can perform DSA (Level of Evidence: C)</td>
</tr>
<tr>
<td>Class IIa</td>
<td>Life-long follow-up for patients in the chronic phase after stent-graft insertion (Level of Evidence: C)</td>
</tr>
<tr>
<td>Class IIb</td>
<td>Indicated for abdominal aortic aneurysm where the maximum minor-axis diameter is ≥55 mm in men and ≥50 mm in women (Level of Evidence: A)</td>
</tr>
</tbody>
</table>

**Note:**

*DSA, digital subtraction angiography.*

Despite the minimally invasive nature of EVAR, this treatment is not applicable to small AAA. EVAR is indicated for lesions of the same size as those subjected to open surgical repair (OSR) (Class IIa, Level of Evidence: B).

(2) Anatomical Indications
To perform EVAR, certain anatomical conditions must be met. These conditions are summarized as follows:

1. The proximal neck is long, relatively straight, and the diameter is not more than 28 to 32 mm;
2. The iliac artery as an access route is large and devoid of extreme flexion/meandering and/or calcification(s);
3. The distal neck is 10 mm or greater.

In particular, criteria for the length and properties of the proximal neck should be strictly followed because they exert major influences on post-placement results. 276–278 (Class I, Level of Evidence: A).

(3) Systemic Indications
EVAR was found to be superior to OSR in patients at low to intermediate risk. The efficacy of EVAR in high-risk patients remains unclear. However, at present in Japan, the first-line therapy for AAA is OSR, and NHI coverage for EVAR is restricted to high-risk patients in whom OSR is difficult.

(4) Other Indications
In general, emergency surgery for including rupture is excluded from the indications for this procedure.

3. Results of EVAR
(1) Results in the Initial and Intermediate Phases
The 2002 report from the EUROSTAR (EUROpean collaborators on Stent-graft Techniques for abdominal aortic Aneurysm Repair) database suggested the usefulness and safety of EVAR. 273 In addition, the EVAR trial 274 and DREAM (Dutch Randomized Endovascular Aneurysm Management) trial 275 demonstrated favorable results of EVAR in the initial and intermediate phases.

(2) Long-Term Results
Although EVAR also shows long-term efficacy, data indicate that OSR has achieved more stable results in terms of the re-intervention rate. 276,277 However, the majority of cases requiring re-intervention after EVAR are suitable candidates for minimally invasive treatment with catheterization. Therefore, future improvement in the device may reduce the re-intervention rate for EVAR.
if the patient has no symptoms with daily living activities of 4 metabolic equivalents (Mets) or higher. However, it is recommended that close examination be performed in patients who have one or more risk factors for cardiac disease, depending on individual cases.

(2) Preparation and Setting
EVAR should be performed in an operating room equipped with a radioscopic device (fixed mount type or portable type) or in an angiographic laboratory that maintains high standards of cleanliness and allows general anesthesia.

(3) Anesthesia
EVAR can be performed under epidural anesthesia, or even under local anesthesia in some cases. However, the use of epidural anesthesia or local anesthesia is not recommended as the first choice. Epidural anesthesia and local anesthesia should, nonetheless, be considered in high-risk patients in whom general anesthesia is difficult (Class IIa, Level of Evidence: B).

(4) Surgical Techniques
It should be angiographically confirmed after stent-graft deployment and touch-up that the stent-graft has been correctly placed at the target site, and that there is no endoleak. If there is type I or III endoleak, additional touch-up and extension should be applied to eliminate type I or III endoleak as thoroughly as possible.

(5) Intraoperative Treatment
Concomitant renal artery stenosis or iliac artery stenosis, if present, may be treated at the same time. Although gluteal claudication may occur when coil embolization of the internal iliac artery is performed, it subsides over time in most cases. Because there is a risk of ischemic enteritis, SVS practice guidelines recommend that antegrade blood flow in the unilaterally intact internal iliac artery be preserved (Class IIa, Level of Evidence: B).

6. Complications After EVAR
Specific complications after EVAR include endoleak, migration, graft occlusion, and rupture.

(1) Endoleak
See Section 2.4.2 of Chapter VI, and Figure 12.

(2) Graft Occlusion
With recent advances in stent-grafts, this has now become a relatively rare complication. If occlusion occurs, thrombus ablation, thrombolysis, extra-anatomical bypass, and other procedures are available treatment options.

(3) Enlargement of the Neck
It has been reported that enlargement of the proximal neck occurs in 1.5 to 16% of patients after EVAR. Thus, long-term follow-up observation is necessary.

(4) Device Migration
The presence of type I endoleak is said to lead to migration of the device. Various anatomical factors such as proximal neck length, flared neck, neck angle, mural thrombus at the proximal neck, and excessive oversizing are involved in the causes of migration.

(5) Follow-up After EVAR
Patients should be followed up for the remainder of their lives after EVAR.

(6) Open Conversion
Intraoperative open conversion has decreased markedly due to advances in devices. The incidence is 0 to 1.6%, according to recent data. On the other hand, most cases requiring open conversion in the late stage involve an endoleak. Although most endoleak cases can be managed with an endovascular approach, open conversion is indicated for enlargement of the aneurysmal diameter after failure of endovascular treatment.

7. Specific Aneurysms
Package inserts of stent-grafts specify that the following diseases are contraindications, as a rule, because the safety and efficacy of stent-grafts have not been evaluated for these diseases. However, in actuality, cases with these diseases may have good indications for stent-grafts if they meet the anatomical criteria. Such conditions are described in detail below.

(1) Ruptured Abdominal Aortic Aneurysm
As for rupture, EVAR may be considered in cases of rupture with stable hemodynamics or in those with impending rupture, if there are anatomical indications (Class IIa, Level of Evidence: B).

(2) Pararenal Abdominal Aortic Aneurysm, Thoracoabdominal Aortic Aneurysm
The primary factor affecting the results of EVAR is the properties of the proximal neck. From this perspective, EVAR is not indicated for pararenal AAA or TAAA. At present, a possible solution is hybrid surgery consisting of extra-anatomical bypass (debranching) to the abdominal branch followed by EVAR.

(3) Inflammatory Abdominal Aortic Aneurysm
In cases of inflammatory AAA, EVAR is useful because it is free of the disadvantages of OSR, such as thickening of the retroperitoneum and ureteral adhesion. Thickening of the retroperitoneum and ureteral obstruction are reported to show improvement after EVAR.

(4) Abdominal Aortic Aneurysm Accompanied by Horseshoe Kidney
About 25% of horseshoe kidneys have a main renal artery which branches from the normal site. In these cases, EVAR may be indicated if the necks are present on the proximal and distal sides.

(5) Abdominal Aortic Aneurysm Accompanied by Aortoenteric Fistula
This condition is not an indication for EVAR because stent-graft infection is induced, leading to persistent infection and sepsis. However, EVAR may be useful if performed for the purpose of improving the patient’s general state, at least until implementation of radical surgery is feasible.

(6) Infective Abdominal Aortic Aneurysm
EVAR is not indicated for this condition because it may cause persistent infection and sepsis. However, EVAR can serve as a bridge to radical surgery. In addition, radical cure may be achieved when EVAR is performed after infection control.
(7) **Congenital Connective Tissue Disorder**

EVAR is likewise not indicated for cases with AAA accompanied by congenital connective tissue disorders such as Marfan syndrome and Ehlers-Danlos syndrome, because of blood vessel fragility in these cases.299

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**VII Specific Pathological Conditions**

### 1. Marfan Syndrome

#### 1. Concept, Pathology, and Etiology

Marfan syndrome is an autosomal dominant inherited disorder reportedly occurring at a frequency of 1 out of 15,000 to 20,000. A genetic etiology is, however, unclear in about 20 to 30% of cases.

At the molecular level, there is an abnormality of fibrillin-1, 300,304,305 a main microfibril component, and various mutations of the fibrillin-1 gene have been found.302 There is a view that excessive fibrillin-1 signals to activate transforming growth factor (TGF)-β is the cause of this syndrome.303 Pathological changes such as cystic medial necrosis and disordered architecture of elastic fibers have been documented.304,305

#### 2. Clinical Condition

Characteristic features of Marfan syndrome are shown in Table 23.

#### 3. Diagnostic Method

Marfan syndrome is diagnosed according to the Ghent diagnostic criteria306 (Table 24).

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**Table 23. Characteristic Features of Marfan Syndrome**

| Skeletal: | Tall stature, long extremities, arachnodactyly, scoliosis, pectus excavatum, pectus carinatum, joint hyperextension |
| Cardiovascular: | Mitral valve prolapse, aortic valve insufficiency, aortic aneurysm, aortic dissection |
| Ocular: | Myopia, ectopia lentis, lens subluxation, retinal detachment |
| Others: | Dural ectasia, spontaneous pneumothorax |

**Table 24. Revised Ghent Criteria for Diagnosis of Marfan Syndrome and Marfan-Like Disorders**

In the absence of family history:

- (1) Aortic root lesions1 (Z ≥ 2) and ectopia lentis → “Marfan syndrome”*
- (2) Aortic root lesions (Z ≥ 2) and an FBN1 mutation2 → “Marfan syndrome”
- (3) Aortic root lesions (Z ≥ 2) and systemic features (≥7 points) → “Marfan syndrome”*
- (4) Ectopia lentis and an FBN1 mutation associated with aortic lesions3 → “Marfan syndrome”
  - When there is no FBN1 mutation associated with aortic lesions even in the presence of ectopia lentis, it is diagnosed as “ectopia lentis syndrome” regardless of systemic features.
  - When aortic root lesions are mild (Z < 2 at the Valsalva sinus) with systemic features (≥5 points plus skeletal features) but without ectopia lentis, it is diagnosed as “MASS”.4
  - When aortic root lesions are mild (Z < 2 at the Valsalva sinus) with mitral valve prolapse but without either systemic features (<5 points) or ectopia lentis, it is diagnosed as “mitral valve prolapse syndrome”.

In the presence of family history:

- (5) Ectopia lentis and family history → “Marfan syndrome”
- (6) Systemic features (≥7 points) and family history → “Marfan syndrome”*
- (7) Aortic root lesion (Z ≥ 2 in patients aged 20 or older and Z ≥ 3 in patients aged less than 20) and family history → “Marfan syndrome”* (Table 23).

*Diagnosis in these cases requires differentiation from Shprintzen-Goldberg syndrome, Loeys-Dietz syndrome, and vascular Ehlers-Danlos syndrome, which resemble Marfan syndrome, and after TGFBR1/TGFBR 2, COL3A1 testing, collagen biochemistry, if indicated. Other conditions or genes will emerge with time.

1Aortic root lesions: Enlargement of the aortic diameter at the Valsalva sinus (determined by Z scores) or aortic root dissection

2FBN1 mutation: Defined in a separate table (details not included)

3FBN1 mutation associated with aortic lesions: FBN1 mutation that has been identified in patients with aortic lesions

4MASS: A combination of myopia, mitral valve prolapse, borderline aortic root dilatation (Z < 2 at the Valsalva sinus), skin striae, and skeletal phenotypes

5Family history: Positive family history with a proband independently diagnosed using criteria (1) to (4) above

Scores of systemic features (Maximum 20 points, positive when points are 7 or more.)

- Wrist and thumb sign: 3 points (wrist or thumb sign: 1 point)
- Pectus carinatum: 2 points (pectus excavatum or chest asymmetry: 1 point)
- Hindfoot deformity: 2 points (pes planus only: 1 point)
- Pneumothorax: 2 points
- Dural ectasia: 2 point
- Protrusio acetabuli: 2 points
- Reduced upper segment/lower segment ratio and increased arm span/height ratio without severe scoliosis: 1 point
- Scoliosis or thoracolumbar kyphosis: 1 point
- Reduced elbow extension: 1 point
- Facial features (3/5) (dolichocephaly, enopthalmos, downslanding palpebral fissures, malar hypoplasia, retrognathia): 1 point
- Skin striae: 1 point
- Myopia (>3 diopters): 1 point
- Mitral valve prolapse: 1 point

Adapted from J Med Genet 2010; 47: 476–485.306 with permission from BMJ Publishing Group Ltd.
Inflammatory abdominal aortic aneurysm (IAAA) is a type of aortic aneurysm that shows aneurysmal dilatation of the abdominal aorta, prominent thickening of the wall, extensive fibrosis around the aortic aneurysm and retroperitoneum, and adhesion to surrounding abdominal organs. Histopathologically, there is extensive fibrosis accompanied by hyalinization in the adventitia and outer surrounding tissue. There is stratified infiltration of non-specific chronic inflammatory cells accompanied by formation of lymph follicles. Although much remains unclear as to the relation between IAAA and AAA, IAAA is generally considered to be a type of AAA that exhibits more prominent inflammation in response to smoking, viral infection, or other factors. On the other hand, along with the recent finding that immunoglobulin G (IgG) 4-related sclerosing disease is distributed throughout the body, it has been proposed that IAAA should be classified into two subtypes, i.e., IgG4-related IAAA and non-IgG4-related IAAA. IgG4-related IAAA is reported to be characterized by more marked inflammation and has features of systemic autoimmune disease, whereas non-IgG4-related IAAA showed more severe atherosclerosis and tends to have features resembling the more common AAA.

2. Frequency
IAAA is said to account for 3 to 15% of all AAA.

3. Clinical Symptoms
Inflammatory symptoms including abdominal pain, abdominal discomfort, low back pain, slight fever, and an elevated erythrocyte sedimentation rate (ESR) are observed. Hydronephrosis also occurs occasionally.

4. Diagnosis
Blood examination frequently reveals elevated ESR and positive C-reactive protein (CRP). However, there are no findings suggestive of bacterial infection (e.g., leukocytosis, positive culture).

Ultrasoundography shows thickening around the abdominal aneurysm (Mantle sign: hypoechoic areas anterior or anterolateral to the aneurysm) that is considered to be specific to this disease. Although surroundings of the aneurysm are viewed as low-density areas on plain CT, contrast-enhanced CT allows clear distinction of the aneurysm from its surroundings based on its hypervascularity.
on enhancement of the surroundings.

5. Treatment (Table 26)
In regard to the therapeutic strategy for the aneurysm itself, artificial graft replacement is indicated according to the criteria for surgery in cases with fusiform atherosclerotic AAA not complicated with IAAA. However, treatment of complications is the first priority in patients who have serious systemic complications.

6. Outcome and Prognosis
Although the natural course of this disease is unclear, the prognosis of surgically treated patients is generally favorable. The inflammation also tends to subside postoperatively.

VIII Aortic Disease and Genetics

1. Aortic Disease and Genetics
Advances in molecular biology and molecular genetics have identified some of the genes related to the development of aortic diseases including aortic aneurysm and dissection.

1. Thoracic Aortic Aneurysm and Genetics
Most TAA cases are non-syndromic and show no symptoms other than the presence of an aortic aneurysm. On the other hand, even among non-syndromic cases, about 20% have more than one affected family member, suggesting that genetic factors are involved in the development of this disease.

2. Site of Thoracic Aortic Aneurysm and Genetics
It has been suggested that aneurysms of the ascending aorta and descending thoracic aorta have different etiologic mechanisms.

3. Abdominal Aortic Aneurysm and Genetics
Etiologically responsible gene loci have been identified in some families having a history of AAA. However, in most cases, AAA is considered to be a multifactorial genetic disease in which environmental factors are also involved in the onset. Among arterial lesions recognized in AAA, atherosclerotic degeneration due to atherosclerosis is found in more than 90% of patients, presenting a feature different from that of medial degeneration due to hypertension or connective tissue involvement as seen in TAA cases. Thus, TAA and AAA are now considered to have different mechanisms of occurrence.

4. Pathological Features and Genetics
The main cause of TAA appears to be medial degeneration represented by medial cystic necrosis. On the other hand, atherosclerotic lesions are often found in AAA cases. Actually, most polymorphisms of genes for which an association was found by SNP (single-nucleotide polymorphism) association analysis of an AAA, are those related to inflammation.

2. Genetic Test
1. Genetic Test Considerations: Adherence to “Guidelines for Genetic Tests and Diagnoses in Medical Practice”
The Japanese Association of Medical Sciences gave notice to its sectional committees on the topic of “appropriate implementation of genetic test”, insisting that “Guidelines for Genetic Tests and Diagnoses in Medical Practice” be referred to when implementing genetic test.

2. Genetic Test Considerations: Use in Children
The “Guidelines for Genetic Tests and Diagnoses in Medical Practice” recommend the following as regards genetic test in minors: careful attention must be given to the rights of subjects who are minors; the test is to be performed only when effective treatment and preventive procedures are available; and efforts must be made to obtain assent to undergo the test through easy-to-understand explanations suited to the ages and developmental stages of all subjects to facilitate their full understanding to the greatest extent possible, even if they are children.

3. Testing Methods
Testing methods are roughly divided into genomic DNA analysis and analysis of messenger RNA extracted from tissue (e.g., blood vessels, skin). Because neither method is perfect, information including the limitations of these methods should be given to the subject prior to testing. If possible, applying a combination of these two methods is preferable.

3. Disease Specifics
1. Genetic Syndromes Associated With Aortic Aneurysms (Table 28)
(1) Marfan Syndrome
This is an autosomal dominantly inherited disorder for which the FBN1 gene on chromosome 15q21 is responsible.

The Ghent criteria, diagnostic criteria for Marfan syndrome, were revised in 2010 (Table 24).
(2) **Loeys-Dietz Syndrome**
Tortuous lesions of systemic arteries including the brain arteries, as well as aneurysms and dissection of large and small arteries, are frequently observed. In particular, aortic aneurysm is found in 98% of patients. In comparison with other similar connective tissue diseases such as Marfan syndrome, the onset is at a younger age, and arterial dissection tends to occur with smaller vascular diameters. At present, the definitive diagnosis is made by gene analysis.

(3) **Vascular Ehlers-Danlos Syndrome**
It is rather rare to find hyperextension of the skin and large joints, which are characteristic of other types of this syndrome. Cardinal symptoms are easy bleeding, dissection, or rupture of large and small arteries, gastrointestinal perforation, organ rupture, and delayed wound healing. Arterial rupture may be secondary to an aneurysm, dissection, or arteriovenous fistula, or may occur without warning. Because of severe tissue fragility, it is necessary to avoid invasive tests, such as catheterization, in cases with vascular complications.

(4) **Turner Syndrome**
This is a relatively common disorder occurring in 1 out of 2,000 to 3,000 female children. In the cardiovascular system, bicuspid aortic valve (10 to 25%) and aortic coarctation (8%) are common. Dilatation of the aortic root is reported to be found in up to 40% of females with Turner syndrome, but it develops into dissection less frequently than in Marfan syndrome or Loeys-Dietz syndrome.

(5) **Other Genetic Syndromes That May Be Associated With Aortic Aneurysm and Dissection**

1. **Congenital Contractural Arachnodactyly (Beals Syndrome)**
This is an autosomal dominantly inherited disorder characterized by a marfanoid habitus and long, slender fingers and toes.
Although slight dilatation of the aortic root is often seen, it is reported that such dilatation very rarely results in dissection. FBN2 is the responsible gene.339

2) Arterial Tortuosity Syndrome

Although tortuosity of arteries is the cardinal feature of arterial tortuosity syndrome, concomitant aortic aneurysm has also been reported.

3) Others

Concomitant aortic aneurysm and dissection have also been found in patients with polycystic kidney disease (PKD1, PKD2 genes), Noonan syndrome (e.g., PTPN11, KRAS, SOS1, RAF1 genes), Alagille syndrome (JAG1, NOTCH2 genes), some types of cutis laxa (ELN, FBXL4 genes), and osteogenesis imperfecta (e.g., COL1A1, COL1A2 genes). Most of these disorders exhibit an autosomal dominant pattern of inheritance. Diagnosis can be made based on clinical findings and family history.

2. Non-Syndromic Aortic Aneurysm

(1) Non-Syndromic Familial Thoracic Aortic Aneurysm

About 20% of patients with TAA or thoracic aortic dissection excluding those with known syndromic diseases such as Marfan syndrome have more than one patient in their families, suggesting the involvement of genetic factors in this disease. Familial analyses of patients with this form of TAA have been identified 6 responsible gene loci or genes (AAT1 to AAT6),340-343 all showing an autosomal dominant pattern.

(2) Non-Syndromic Abdominal Aortic Aneurysm

About 11 to 20% of patients with non-syndromic AAA have more than one patient in their families, suggesting the involvement of genetic factors. No genes responsible for this disease, consistent with a single-gene disorder, have yet been identified, and multiple gene polymorphisms are reported to be involved in the disease onset.

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