Aortic Disease

Maintenance of Certification Self-Assessment Module
2016

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Faculty Disclosure Summary

Listed below are individuals with control of the content of this program who have disclosed...

Relevant relationship(s) with industry:

None.

No other financial relationships with industry:

Joseph J. Malezewski, MD, FCAP, FACC (Presenter)
Julie C. Knudtson, MD (Planning Committee)
Babbi S. Pfilt, MD (Planning Committee)
Sherrie A. Preusse (Planning Committee)

References to off-label usage of pharmaceuticals or instruments in their presentation:

None.

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Target Audience

General pathologists, surgical pathologists, autopsy pathologists, cardiovascular pathologists, forensic pathologists, molecular pathologists, pathology residents, and pathology fellows.

Overview

Aortic specimens are commonly encountered in clinical practice, but formal training on how to handle them is lacking. Both the Society for Cardiovascular Pathology (SCVP) and the Association for European Cardiovascular Pathology (AECVP) have recognized the need for a uniform approach to the handling of these specimens. Pathologists and clinicians alike need to be aware of the latest advances in the field, focusing on the emerging technologies used in genetics and proteomics. This module aims to familiarize participants with the fundamentals of handling surgical aortic specimens. Specifically, the course will address information that is relevant and actionable for clinicians. It also seeks to inform participants about the emerging technologies available to help inform diagnoses and clinical practice.

Learning Objectives

The participant will:

- Describe the fundamental approaches to handling aortic specimens (both surgery- and autopsy-derived);
- Explain the impact of emerging ancillary tools such as molecular genetics on specimen handling and result reporting;
- Synthesize clinically actionable information that is essential to convey to care providers and, ultimately, the patient and/or the patient's family.
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Overview

- Aortic structure
- Primer on aneurysms
- Types of aortic aneurysms
  - acquired
  - heritable
- Tips for reporting
- Processing vascular specimens
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Aortic Structure
Elastic Artery

Four Basic Components:
- elastin fibers
- collagen fibers
- smooth muscle cells
- amorphous ground substance
Aortic Structure
Regional Anatomy

Overview
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Aortic Aneurysms

Definition:
Localized abnormal dilatation of an artery (beyond its borders).

Types:
True: contain all 3 layers of the vessel (intimal, media, adventitia)
False: contained rupture with wall formed by adventitia and thrombus
Dissecting: intra-medial hematoma
Aortic Aneurysms
Gross Pathology

Overview

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Types of Aortic Aneurysms
Acquired

- Hypertension
- Aortitis
- Cocaine-associated
- Exertional (weight-lifting)
Types of Aortic Aneurysms

Heritable
- Bicuspid aortic valve
- Marfan syndrome
- Loewe-Dietz syndrome
- Ehlers-Danlos syndrome, type IV
- Turner syndrome
- Noonan syndrome
- Homozygous familial hypercholesterolemia
- Arterial tortuosity syndrome
- Autosomal dominant polycystic kidney disease
- Non-transposition conotruncal anomalies
- Familial thoracic aortic aneurysm syndrome
- Autosomal recessive cutis-laxa
- Fibromuscular dysplasia
- Shprintzen-Goldberg syndrome

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Aortic Specimens
Examination & Processing of Specimens

Opened Specimen “Partial Resection”
Unopened Specimen “Segmental Resection”
Aortic Specimens
Examination & Processing of Specimens

**Gross Examination**

- **Features to note**
  - Dissection
  - Atherosclerosis (calcium?)
  - Intimal change (wrinkling)
- **Sections to submit**
  - 6-8 sections of aortic wall
  - Cut in different planes
  - Representative of entire specimen

- **Stains to order**
  - H&E
  - VVG (or other elastic stain)

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Aortic Specimens
Examination & Processing of Specimens

Representative Sections

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Aortic Specimens
Examination & Processing of Specimens

Representative Sections

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Aortic Specimens
Examination & Processing of Specimens

Histologic Examination

• Features to note
  • Medial degeneration
  • "cystic"-type
  • "diffuse"-type
  • Laminar medial necrosis
  • Medial disorganization
  • Aortitis
  • Dissection (acute vs. chronic)

Note:
• Histopathologic features non-specific
• Certain features have been associated with CTD
  • Med. degen (diffuse)
  • Med. disorganization

Medial Degeneration
Aortic Specimens
Examination & Processing of Specimens

Histologic Examination
• Features to note
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  • "diffuse"-type
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Medial Degeneration
Aortic Specimens
Examination & Processing of Specimens

**Histologic Examination**

- **Features to note**
  - Medial degeneration
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Laminar Medial Necrosis

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Aortic Specimens
Examination & Processing of Specimens

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Medial Disorganization

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Aortic Specimens
Examination & Processing of Specimens

**Histologic Examination**

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Medial Disorganization
Aortic Specimens
Examination & Processing of Specimens

Histologic Examination
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Aortitis

Aortic Specimens
Examination & Processing of Specimens

Histologic Examination
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    • Dissection (acute vs. chronic)

Aortitis

Aortic Specimens
Examination & Processing of Specimens

Histologic Examination
• Features to note
  • Medial degeneration
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    • Dissection (acute vs. chronic)

Acute Intramedial Dissection
Aortic Specimens
Examination & Processing of Specimens

**Histologic Examination**

- **Features to note**
  - Medial degeneration
  - "cystic"-type
  - "diffuse"-type
  - Laminar medial necrosis
  - Medial disorganization
  - Aortitis
  - Dissection (acute vs. chronic)

- Acute Intramedial Dissection

- Chronic Intamedial Dissection
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Aortic Specimens
Tips for Reporting

Template

According to the following tree descriptions:

Description: Resected or formalin labeled. 

(AA/0-1)

* in a segment of aorta,
measuring _______ cm in length, _______ cm in external diameter, and up to _______ cm in thickness. / with mural dissection / with old friable / with old soft material / with _____


Description: Resected or formalin labeled. 

(AA/1-

* in a previously opened segment of aorta, measuring _______ cm in length, _______ cm in external diameter, / with mural dissection
/ with old friable / with old soft material / with _____


Diagnosis: According aorta, aneurysm / dissection, focal / segmental / reaction: # (excluded with VVG stain).

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- Types of aortic aneurysms
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- Tips for reporting
- Processing vascular specimens
Vascular Specimens
Fibromuscular Dysplasia - General

- **Distribution**
  - renal, carotid, iliac, abdominal, cerebral
- **Consequences**
  - renovascular HTN
  - dissection, rupture
- **Angiography**
  - string-of-beads
- **Pathology**
  - ridges, aneurysms
Vascular Specimens
Fibromuscular Dysplasia - General

- **Distribution**
  - renal, carotid, iliac, abdominal, cerebral
- **Consequences**
  - renovascular HTN
  - dissection, rupture
- **Angiography**
  - string-of-beads
- **Pathology**
  - ridges, aneurysms

Vascular Specimens
Processing

**Gross Examination**
- **Features to note**
  - Aneurysm / Dissection
  - Intimal change (corrosion, athero, etc.)
- **Sections to submit**
  - 3-4 sections of vascular wall
  - Cross sections - temp art bx
  - Longitudinal sections - FMD

**Stains to order**
- H&E
- VVG (or other elastic stain)

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