VASCULITIDES

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WVES | Winnipeg Vascular & Endovascular Symposium

PRESENTER DISCLOSURE

Presenter: Naomi Wedel

I have no current relationships with commercial entities

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RAYNAUD'S

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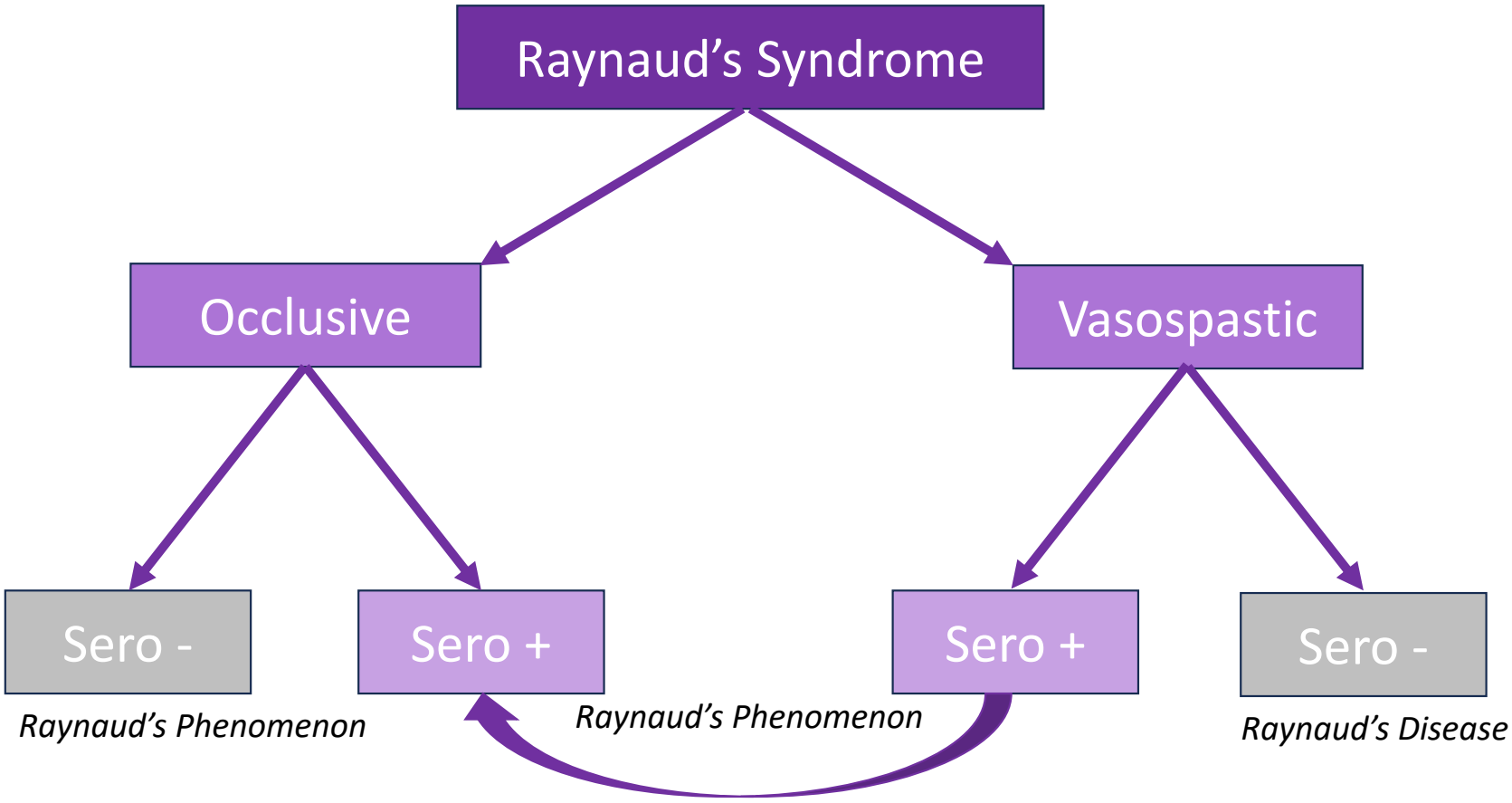
NOMENCLATURE

- Raynaud's Syndrome
  - Overarching term
- Raynaud's Phenomenon
  - Overarching term for primary and secondary RP
  - Secondary Raynaud's
- Raynaud's Disease
  - Primary Raynaud's

ALGORITHM


Raynaud's Syndrome
Occlusive
Vasospastic
Sero -
Sero +
Sero +
Sero -
Raynaud's Phenomenon
Raynaud's Phenomenon
Raynaud's Disease

Secondary Raynaud's Phenomenon

Sero +

Connective Tissue Disorder
- Scleroderma (CREST)
- Systemic lupus erythematous
- Rheumatoid arthritis
- Sjogren syndrome
- Mixed CTD
- Dermatomyositis/polymyositis
- Vasculitis
  - Small vessel
  - Medium vessel
  - **Large vessel**

Sero -

Occupational/trauma
- **Hypothenar hammer syndrome**
- **Vibration induced**
- Frostbite

Occlusive
- **Atherosclerosis**
- **Thromboangiitis obliterans**
- **Arterial emboli**
- **Thoracic outlet syndrome**

Malignancy
- MM, leukemia, adenocarcinoma

Hematologic
- PCV, cryoglobulinemia, cold agglutinins

Infection
- hepatitis B/C, parvovirus, purpura fulminans

Drug-induced vasospasm
- Vasopressors, ergot, cocaine, amphetamines, bleomycin

MANAGEMENT

- Clinical = Vasospastic attacks of pallor precipitated by cold or stress
  - Tissue loss = Secondary RP
- Serological = ANA, ENA, C3/C4, ANCA, RF, dsDNA, anti-ssa/Ro, anti-Scl-70
- Imaging = CTA, vascular US, cold challenge, nail-fold capillary microscopy
- Conservative = education, avoid triggers, temperature biofeedback
- Pharmacologic = vasodilators reduce frequency and severity attacks
  - CCB (nifedipine, amlodipine) = First line
    - Topical nitroglycerin, Alpha 1 Rc antagonists (prazosin)*, ACEi, ARB, Serotonin reuptake inhibitors
  - PDEi (sildenafil)* = improved blood flow and ulcer healing
  - Prostaglandins (iloprost IV)* = ulcer healing
- Surgical = sympathectomy, botox, nerve stimulation

*Secondary RP = treat underlying cause

VASCULITIS

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VASCULITIS

- Classification and nomenclature can be unnecessarily confusing
- Most important first step: consider the possibility of 'some sort of vasculitis' → then narrow down the specific type
- Two main classification systems:
  - American College of Rheumatology (ACR)
  - Chapel Hill Consensus Conference
- Simplest way to sort out the vasculitides is to list them according the size of the artery

CLASSIFICATION

Vascular Surgery

Internal Medicine

Vascular Surgery

TABLE 138.1 International Chapel Hill Consensus Conference Nomenclature[3]

Nomenclature		Vasculitis
Large-vessel vasculitis		Giant cell arteritis (also known as temporal arteritis)
		Takayasu arteritis
Medium-vessel vasculitis		Polyarteritis nodosa
		Kawasaki disease
Small-vessel vasculitis	Immune complex-mediated	Antiglomerular basement membrane (anti-GBM) disease
		Cryoglobulinemic vasculitis
		IgA vasculitis (Henoch–Schönlein purpura)
		Hypocomplementemic urticarial vasculitis (anti-C1q vasculitis)
	Antineutrophil cytoplasmic antibody-mediated (ANCA-associated pauci-immune)	Granulomatosis with polyangiitis (previously known as Wegener granulomatosis)
		Microscopic polyangiitis
		Eosinophilic granulomatosis with polyangiitis (previously known as Churg–Strauss syndrome)
Variable-vessel vasculitis		Behçet disease
		Cogan syndrome
		Thromboangiitis obliterans (Buerger

IDENTIFY THE POSSIBILITY

- No single presentation
- Clinical
  - Large = claudication/CLTI, asymmetric BP, absence of pulses
  - Medium = cutaneous changes, digital gangrene, renovascular HTN
  - Small = cutaneous changes, pulmonary-renal syndromes
- Characteristic histology -> Bx not practical for large vessel
- Imaging
  - Irregular/asymmetric tapering and narrowing
  - Concentric thickening
  - “Beading” with segmental stenosis
  - Aneurysm formation
  - Wall enhancement (nuclear medicine)

Large-Vessel

Type of Vasculitis	Thoracic Aortic Disease	Abdominal Aortic Disease	Carotid/Vertebrall Arterial Disease	Stroke due to Small- or Medium-Artery Disease	Upper- and Lower-Extremity Arterial Stenosis	Renal Arterial Disease	Coronary Artery Disease
GCA	++*	+†			+	+	Rare‡
TA	+++§	++			+++	++	+
Behçet's disease	+	++	Rare	Rare	+	Rare	Rare
Other large-vessel diseases (RPC, CS, RPF, IA)	++	+	+	Rare	+	+	Rare

VARIABLE-VESSEL

Behcet's

- Young adult, <40 yo
- Mediterranean/Middle and Far East
- Mucocutaneous manifestations
  - Genital and oral ulcers, epididymitis, arthritis, GI lesions, uveitis
- 30% variable- vessel vasculitis
  - Aorta and major branches, pulmonary arteries
  - Venous thrombosis

Cogan's Syndrome

- Young adults, <40 yo
- Interstitial keratitis (corneal scarring)
- Sensorineural hearing loss and vestibular dysfunction
- Large vessel vasculitis in up to 15%
  - Similar to Takayasu's

Thromboangiitis Obliterans

- Young adults, <40 yo
- 98% associated with smoking
- Mediterranean, Middle East, Asia, Eastern Europe
- Medium/small-vessel vasculitis
  - Infrapopliteal and infrabrachial
- Superficial migrating thrombophlebitis

Takayasu Arteritis

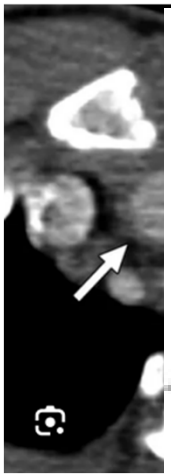
- Population = female, 30's from East Asia
- Pathology = panarteritis with skip areas
- Diagnosis = if 3 of 6 criteria are met
  1. **Age criteria**: <40 at disease onset
  2. **Claudication**: especially of upper extremities
  3. **Pulse:** decreased pulsation of one or both brachial arteries
  4. **Blood pressure**: difference > 10 mmHg systolic between arms
  5. **Bruit**: over one or both subclavian arteries or abdominal aorta
  6. **Angiography**: narrowing/occlusion of the aorta, its primary branches or large arteries in proximal upper/lower extremities
     - Not due to atherosclerosis, FMD

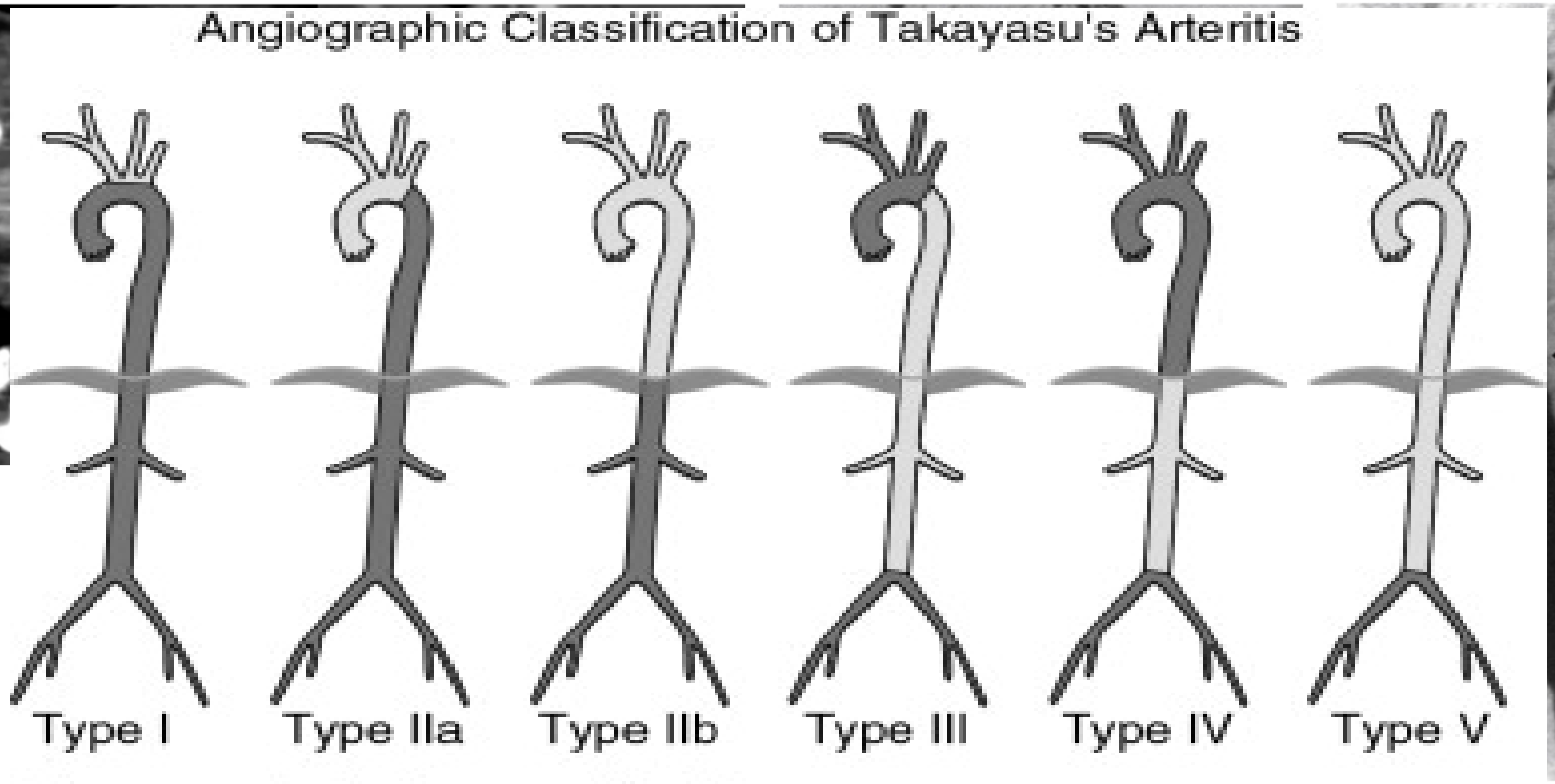
TA- Clinical Manifestations

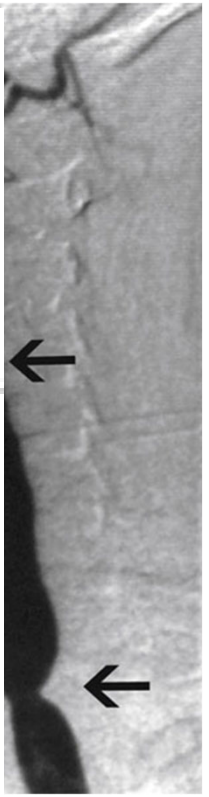
- Prepulseless systemic phase (less than half of patients):
  - Fever, Weight loss, Malaise, Generalized arthralgias and myalgias
- Occlusive Phase = “pulseless” disease
  - 90% predilection to aortic arch and its branches (symmetric)
  - Burned out = vessel fibrosis, aneurysmal degeneration

Artery	%	Manifestations
Subclavian	93	Arm claudication, RP, diminished/absent pulses, asymmetric BP
Carotid	58	Carotidynia, headache, vision changes, syncope
Abdominal Aorta	47	Mesenteric angina
Renal	38	HTN, Renal failure
Aortic Arch	35	Aortic insufficiency
Vertebral	35	Visual changes
Mesenteric	18	Mesenteric angina
Pulmonary	20	Chest pain, dyspnea

TA- IMAGING







Types can be further modified by:
C: involved coronary artery
P: involved pulmonary artery

TA- Treatment

- Treatment depends on disease activity
- Mainstay of treatment = **corticosteroids**
  - Prednisone/prednisolone 1 mg/kg daily for 1-3 months then taper
  - If fail to respond in 3 months or show a flare of the disease during tapering add cytotoxic agents = Methotrexate, Azathioprine, Cyclophosphamide
- Surgery = delayed until active phase over (significant risk of re-occlusion)
  - No vessel wall thickening/edema on imaging, ESR normal, no constitutional sx
  - Revascularization = bypass (best long-term prognosis), patch angioplasty angioplasty/stenting
- 10-year survival rate is about 90%
  - Reduced in patients with major complications = 40%

Giant Cell Arteritis

- Population = female, > 50 years, Northern European descent
- Diagnosis
  - No pathognomonic signs
  - Temporal Bx
    - Lymphocytic infiltrate, non granulomatous, IEL fragmentation
    - False negative
      - Sampling errors
      - Steroids initiated prior to dx = intimal fibrosis, medial scarring, asymmetric destruction IEL

Criterion	Definition
1. Age at disease onset ≥50 years	Development of symptoms or findings beginning at age 50 or older
2. New headache	New onset of or new type of localized pain in the head
3. Temporal artery abnormality	Temporal artery tenderness to palpation or decreased pulsation, unrelated to arteriosclerosis of cervical arteries
4. Elevated erythrocyte sedimentation rate	Erythrocyte sedimentation rate ≥50 mm/hour by the Westergren method
5. Abnormal artery biopsy	Biopsy specimen with artery showing vasculitis characterized by a predominance of mononuclear cell infiltration or granulomatous inflammation, usually with multinucleated giant cells

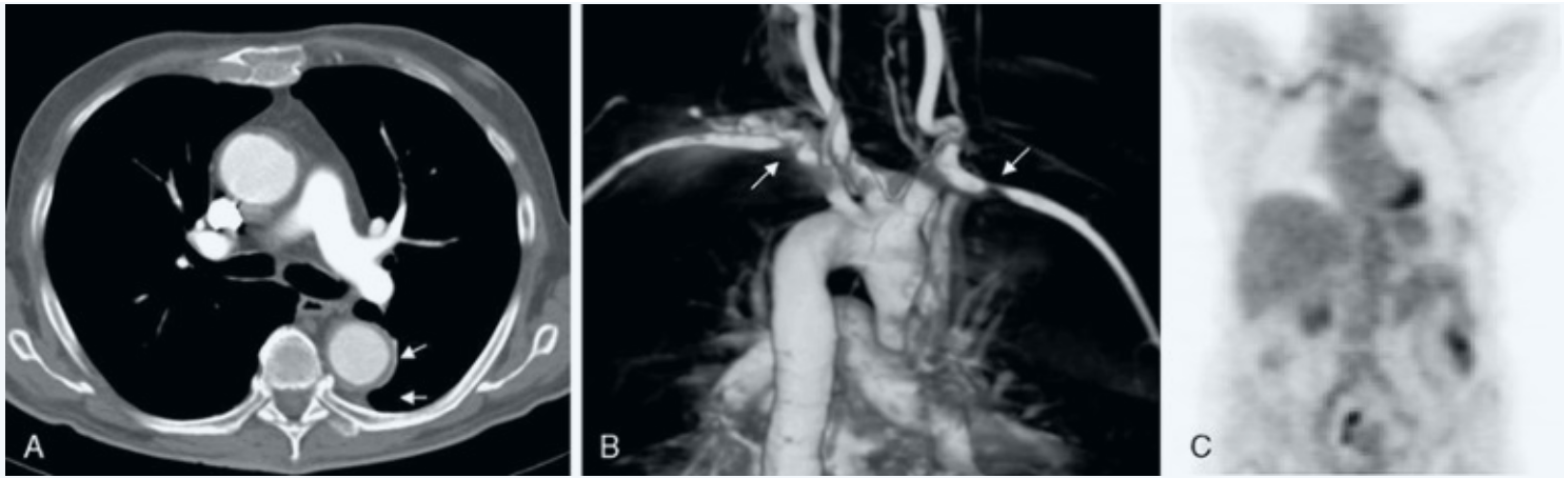
GCA- Clinical Manifestations

- Classic = transient diplopia, headache, jaw claudication, temporal artery changes
- Vascular = increased ratio of TAA:AAA
  - Subclavian arteries involved in 70%, axillary 40%, iliacs 37%, femorals 37%
- PMR = May precede GCA symptoms or be the only clinical manifestation

General Features	
Age (mean, range)	75 (50-94)
Sex (female/male)	178/72
Weight loss	61%
Fever	47%
Ophthalmic Events	**22%**
Blindness (permanent)	14%
Amaurosis fugax	10%
Transient diplopia	4%
Cerebrovascular Accident	**2%**
Symptomatic Large Vessel Involvement (Claudication and/or Bruit)	**5%**
Polymyalgia Rheumatica (PMR)	**48%**

Cranial Symptoms	**86%**
Headache	77%
Temporal artery abnormality (swollen, tender, weak/absent pulse)	74%
Jaw claudication	44%
Scalp tenderness	39%
Facial pain	18%
Earache	18%
Odynophagia	12%
Ocular pain	8%
Tongue pain	5%
Carotodynia	5%
Toothache	5%
Trismus	1%

GCA- Imaging



GCA- Treatment

- Glucocorticoid therapy is treatment of choice
  - Induces dramatic improvement in symptoms within days
  - Prednisone at 40-60 mg/day initial dose x 2-4 weeks
- Methotrexate helps prevent relapses and reduces glucocorticoid use
- Presence of any ocular symptoms is a medical emergency

- Surgical intervention as needed
  - Wait until no longer active
  - Aneurysmal degeneration
  - Revascularization rarely required due to collateral networks

THANKS!

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