

Small vessel diseases

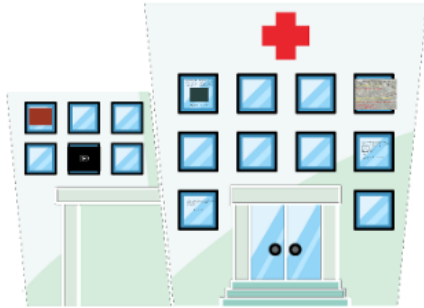
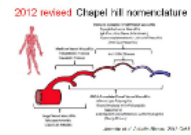
- Granulomatosis with polyangiitis (GPA)
- Microscopic polyangiitis (MPA)
- Churg-Strauss disease
- Henoch-Schönlein purpura
- Cryoglobulinemia

Medium Vessel diseases

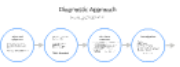
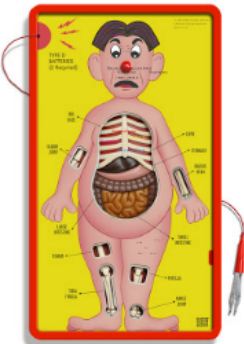
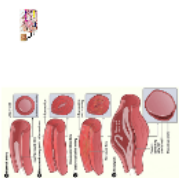
- Behçets
- Kawasaki
- Polyarteritis Nodosa (PAN)
- Rheumatoid Vasculitis

Large vessel diseases

- Giant cell arteritis
- Polymyalgia rheumatica
- Isolated Aortitis
- Takayasu's arteritis



Vasculitis: A Common Rare disease

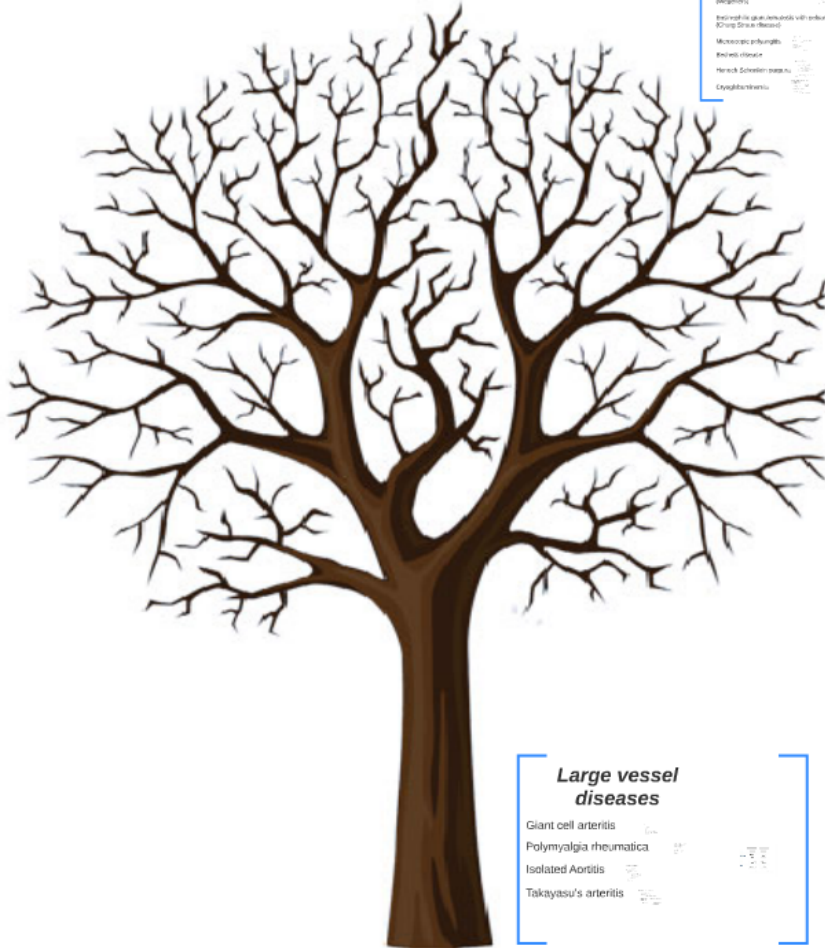


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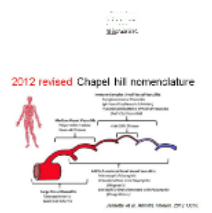
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- Cryoglobulinemia



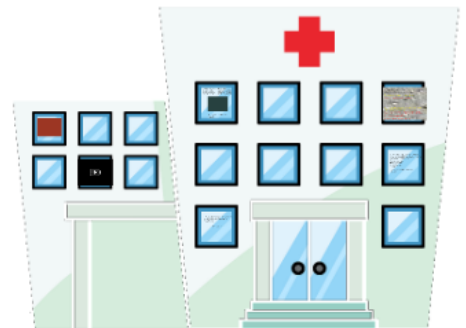
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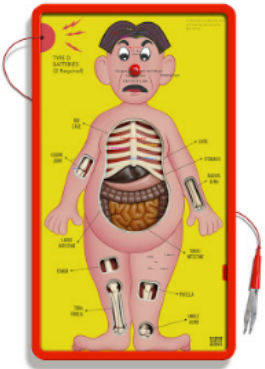
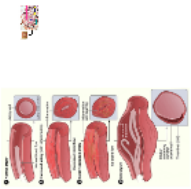


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Vasculitis: A Common Rare disease



Systemic symptoms

- Constitutional symptoms
- Joint symptoms
- Skin symptoms
- Organ symptoms

Small vessel diseases



Large vessel diseases

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Polymyalgia rheumatica

Isolated Aortitis

Takayasu's arteritis

Vasculitis: A Common Rare disease

Presenter disclosure:

Dr. Michael Fernando MPC

Honaria: AstraZeneca for a talk into exercise in diabetes.

Other: Shares in mutual funds, Weed MD, currently applying for ethics for an RCT looking at Exercise and diabetes.

This program has received in-kind support from the Vasculitis Foundation in the form of Logistical support and an introduction to Dr Christian Pagnoux a vasculitis researcher based in Toronto.

Potential for conflict of interest:

None.

Mitigating Potential bias:

No specific medications, treatments or brand names will be discussed.

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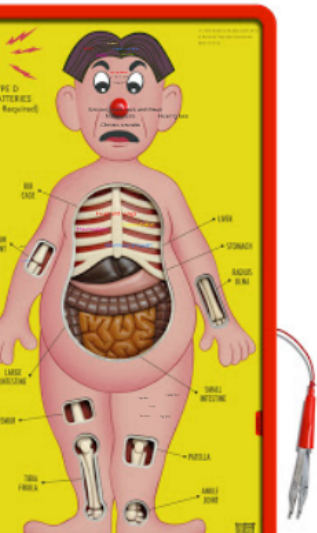
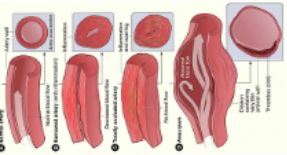
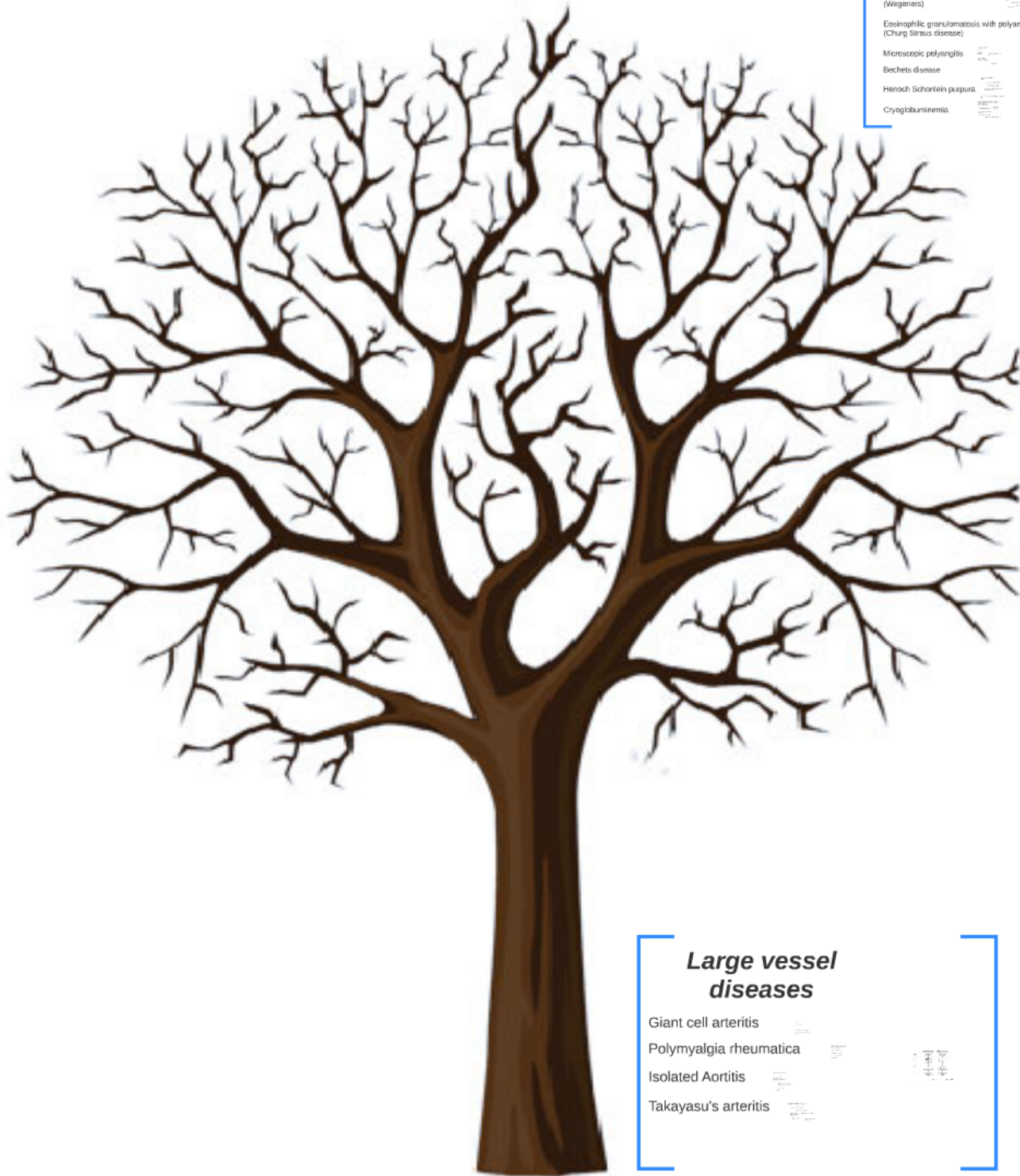
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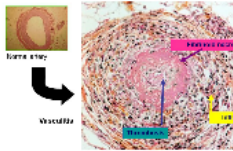
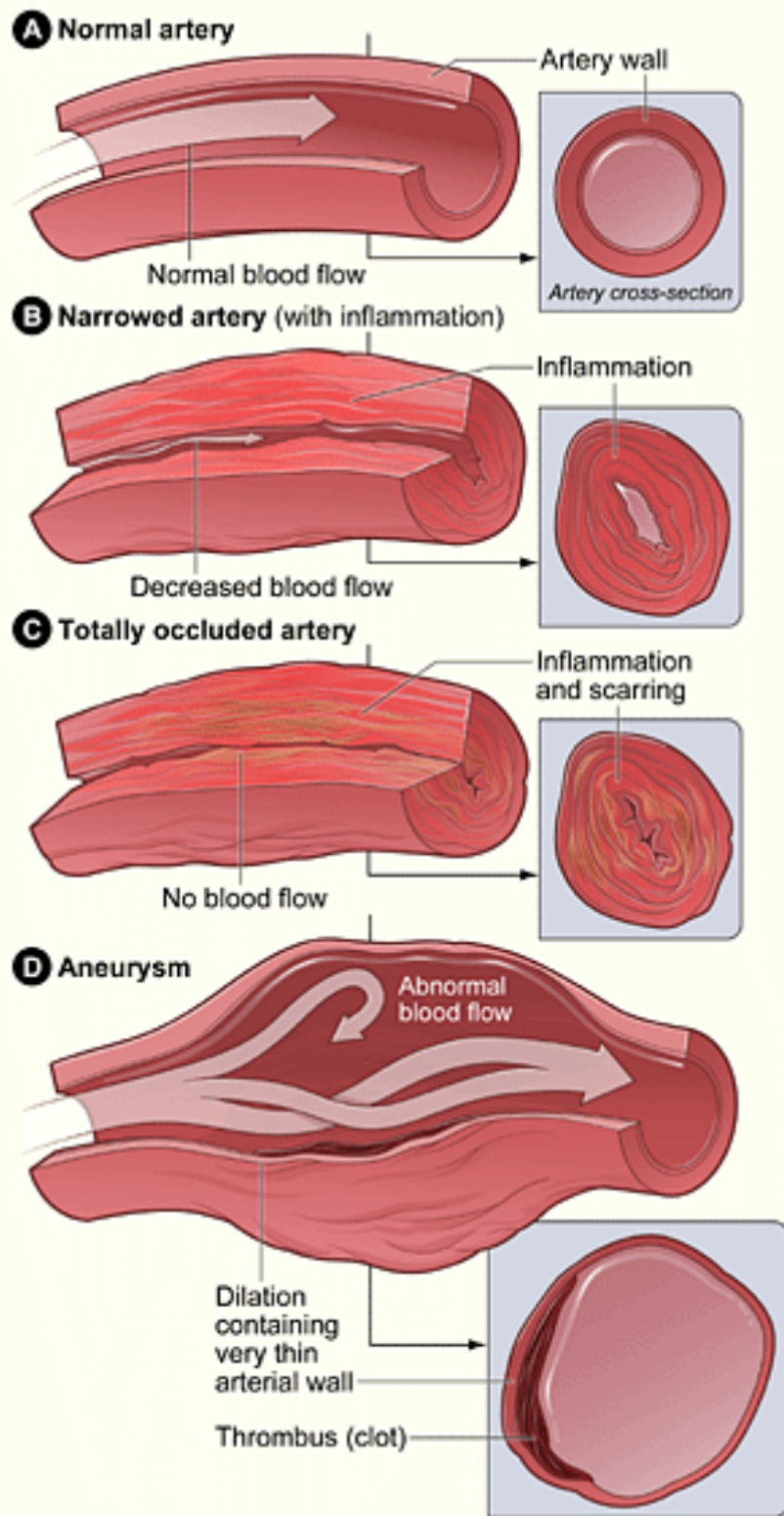
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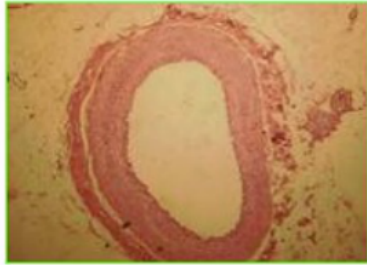
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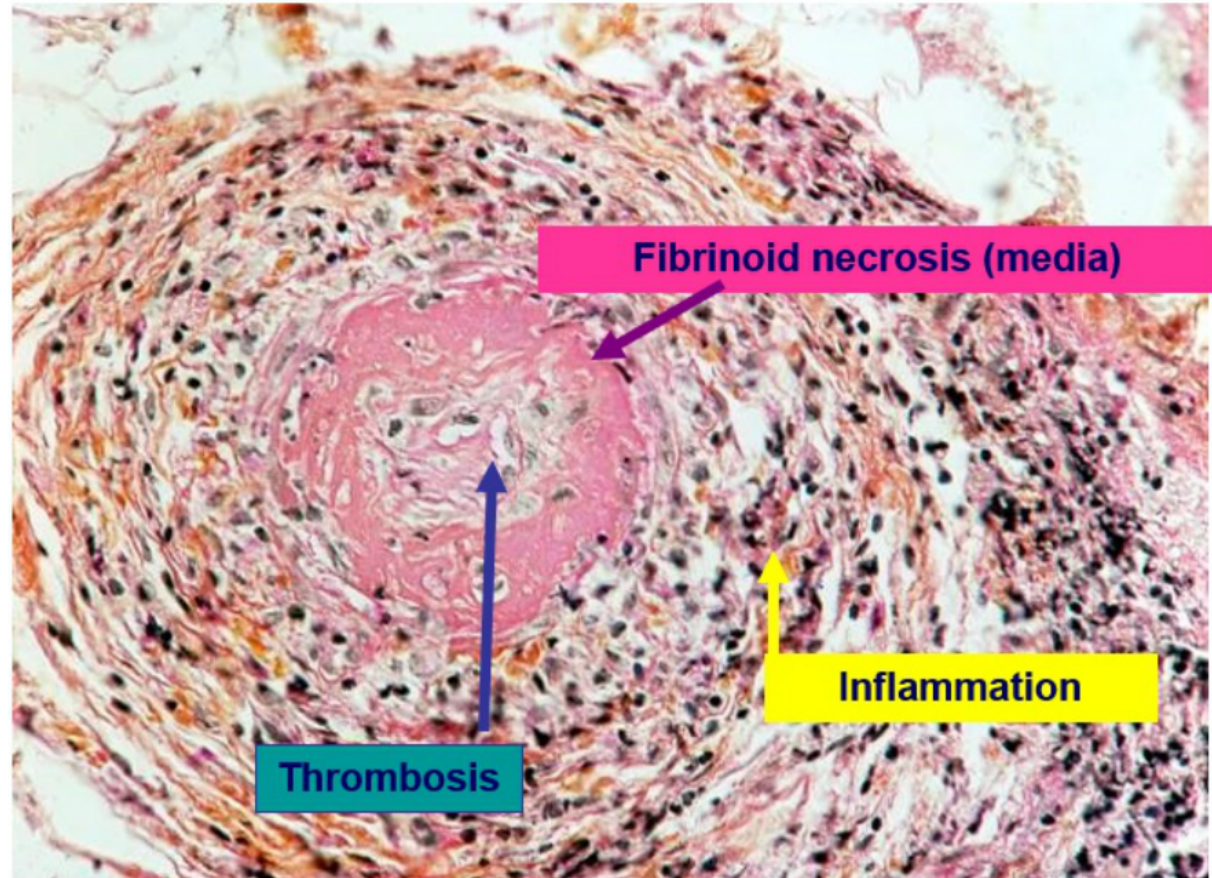




Normal artery



Vasculitis



Fibrinoid necrosis (media)

Thrombosis

Inflammation

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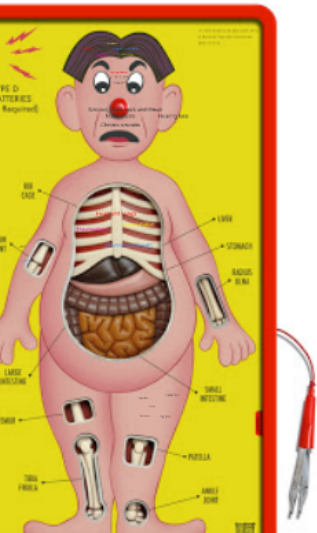
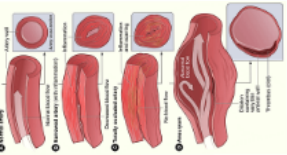
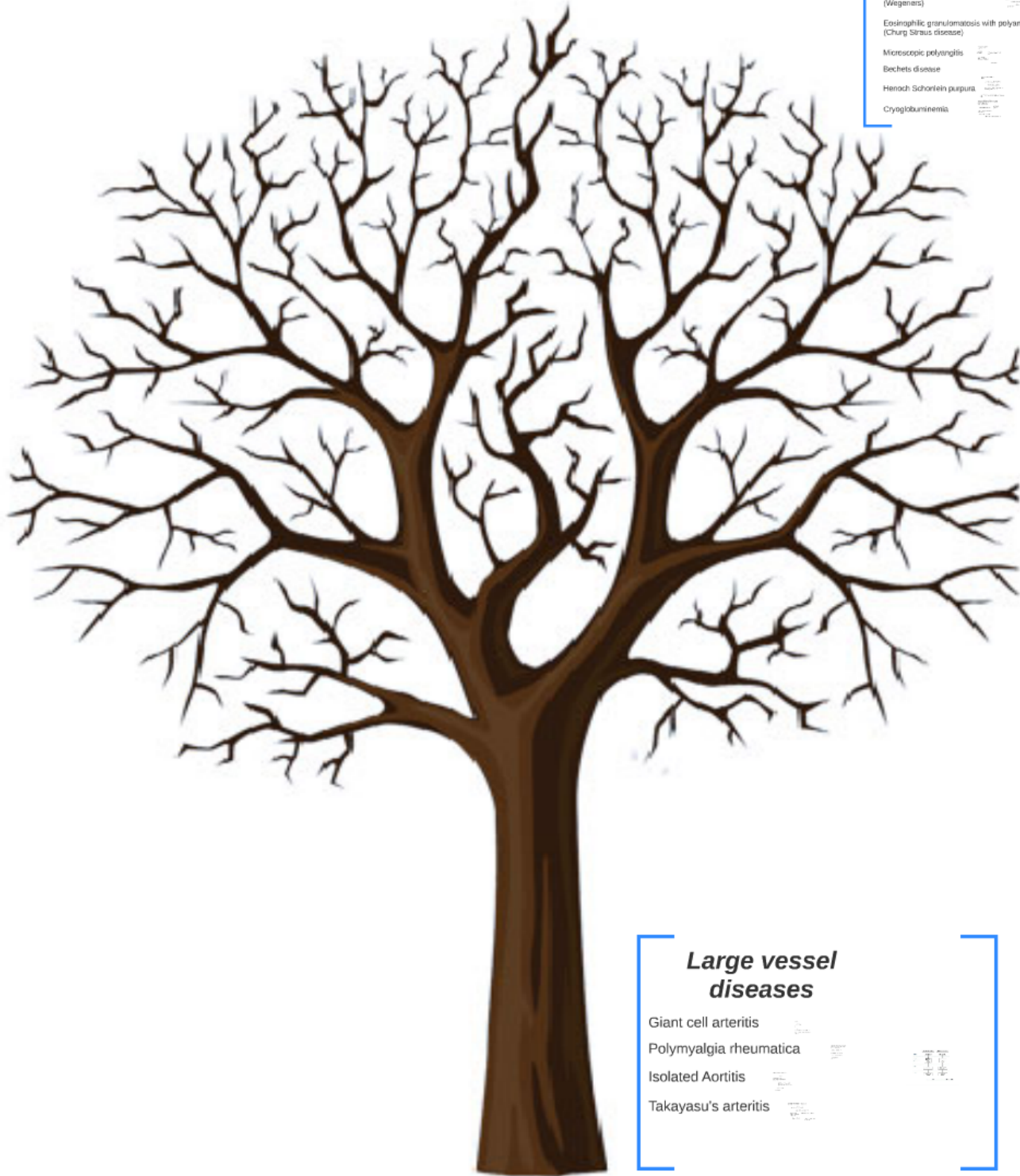
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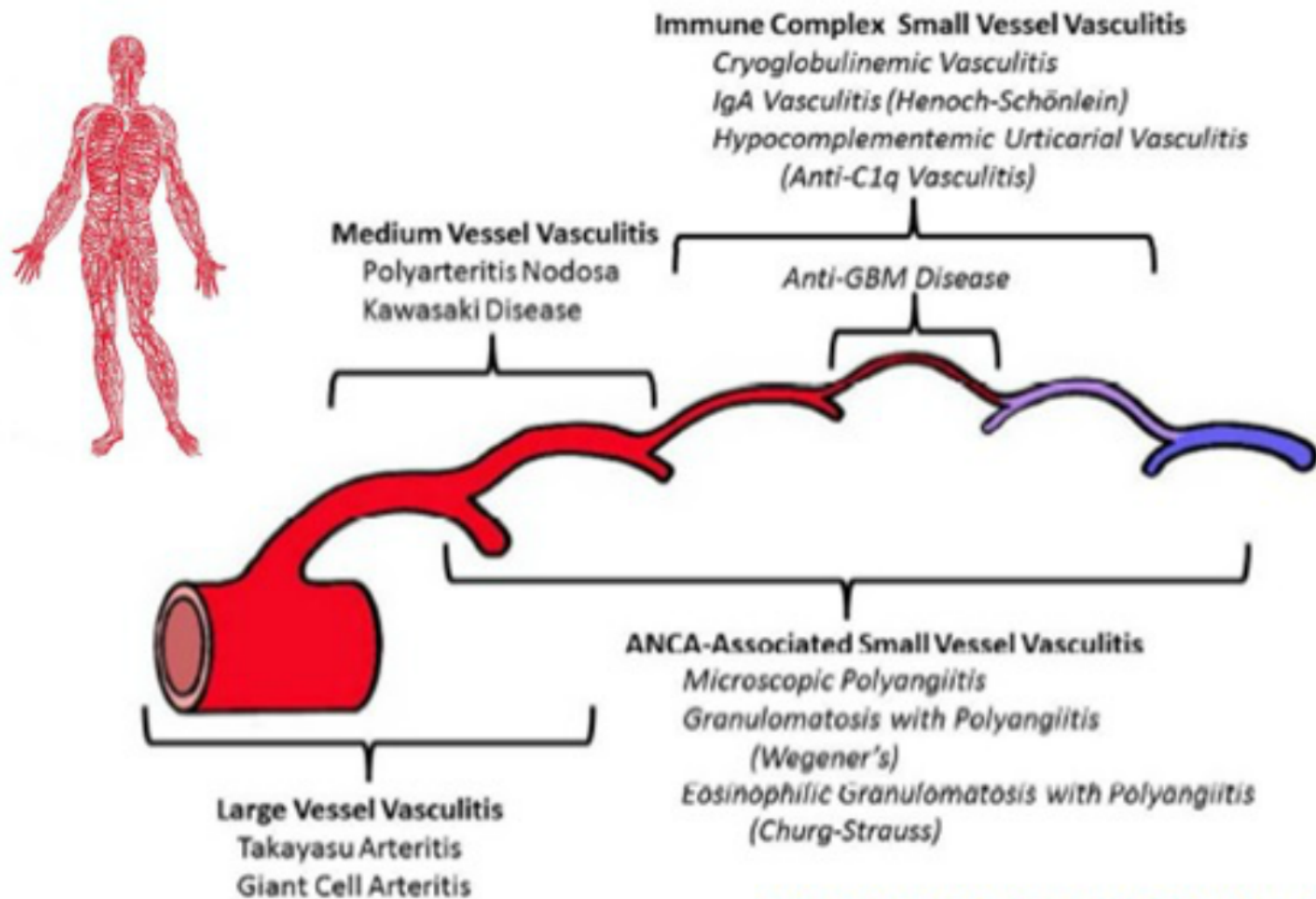
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2012 revised Chapel hill nomenclature



Jennette et al. *Arthritis Rheum.* 2012 Oct 8.

2012 revised Chapel hill nomenclature

- **Variable Vessel Vasculitis (VVV):** Behçet's Disease (BD) and Cogan's Syndrome (CS).
- **Single Organ Vasculitis (SOV):** Cutaneous Leukocytoclastic Angiitis, Cutaneous Arteritis, Primary CNS Vasculitis and Isolated Aortitis.
- **Vasculitis Associated with Systemic Disease:** Lupus Vasculitis, Rheumatoid Vasculitis and Sarcoid Vasculitis.
- **Vasculitis Associated with Probable Etiology:** Hepatitis C Virus-Associated Cryoglobulinemic Vasculitis, Hepatitis B Virus-Associated Vasculitis, Syphilis-Associated Aortitis, Serum Sickness-Associated Immune Complex Vasculitis, Drug-Associated Immune Complex Vasculitis, Drug-Associated ANCA-Associated Vasculitis and Cancer-Associated Vasculitis.

Large vessel diseases

Giant cell arteritis

ICA
 Age-related
 ESR
 50+
 50% of patients with fever
 Pain of scalp, limb girdle and joints
 Vision

Polymyalgia rheumatica

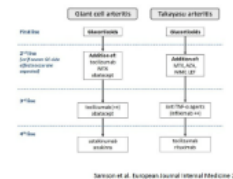
Gender and race-related: usually
 elderly, female, European ancestry,
 White and Chinese descent

Isolated Aortitis

Genetic and serological
 HLA-B*27
 ANCA
 ANCA-related disease
 ANCA-related disease or other
 vasculitis

Takayasu's arteritis

Maybe viral but unknown
 Super rare (2.5% EBV)
 Asian females under 40
 Age and sex predominate
 Female, weight loss
 CRP and ESR
 Transcatheter aortic
 stenosis



GCA

Autoimmune

F>M

>50s

Signs symptoms quite familiar

Risk of stroke, blindness and aortic aneurysm

Large vessel diseases

Giant cell arteritis

ICA
 Age-related
 ESR
 50+
 50% for women, 40% for men
 Pain of scalp, limb and joint
 Lockjaw

Polymyalgia rheumatica

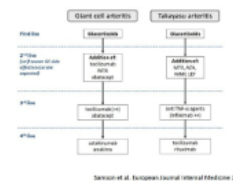
Gender and race-related: usually
 affects Caucasians
 60-70 years old
 Only very slight elevation of
 ESR and/or CRP
 Neck pain
 Morning stiffness

Isolated Aortitis

Genetic and infectious
 Myxoma
 High cholesterol
 Aortic aneurysm or dissection
 Dissecting aortic aneurysm of
 aorta and/or descending aorta
 Aortic root dilation
 ESR & CRP
 Tumor markers

Takayasu's arteritis

Maybe viral but unknown
 Super rare (2.5/100k)
 Asian females under 40
 Age and race specific
 Fatigue, fever, weight loss
 ESR and CRP
 Transcatheter aortic valve
 intervention



Shoulder and hips affected - usually stiffness. Can also get depression, fevers and low appetite

Second most common inflammatory disease in the elderly (>70s)

Only really effects europeans

Complications can include aneurysms

Very treatable with steroids

Large vessel diseases

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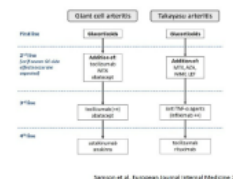
Gender and race-related: usually
 affects Caucasians
 60-70 years old
 Only very slight overlap with
 GCA
 ESR
 50+

Isolated Aortitis

Genetic and infectious
 Males
 High ESR
 High CRP
 High ferritin
 High IgG
 High IgA
 High IgM
 High IgG4
 High IgA1
 High IgM1
 High IgG2
 High IgA2
 High IgM2
 High IgG4
 High IgA1
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Takayasu's arteritis

Maybe viral but unknown
 Super rare (2.5-3%)
 Asian females under 40
 Age and sex-specific
 Female, weight loss
 CRP and ESR
 Transcatheter aortic valve
 replacement



Stenosis and aneurysms

Very rare

Mainly associated with inflammatory diseases and other vasculitides

Can present with shortness of breath and heart failure and chest pains (also fainting and

ESR is handy

Treat the cause

Large vessel diseases

Giant cell arteritis

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 Pain of scalp, limb girdle and joints
 Vision

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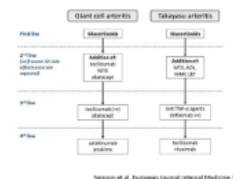
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 elderly, European descent,
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 Only very slight overlap with
 ICA
 ESR
 50+

Isolated Aortitis

Genetic and infectious
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 High cholesterol
 Aortic aneurysm or dissection
 Aortic regurgitation
 Concomitant with absence of
 fever and high ESR and more
 subtle clinical picture
 ESR < 10
 Tumor markers

Takayasu's arteritis

Maybe viral but unknown
 Super rare (2.5% ICA)
 Asian females under 40
 Age and race par-
 ticularly with Asian
 countries of birth
 Fatigue, fever, weight loss
 ESR and CRP
 Transcatheter aortic
 stenosis



Maybe Viral but unknown

Super rare (2-3/million)

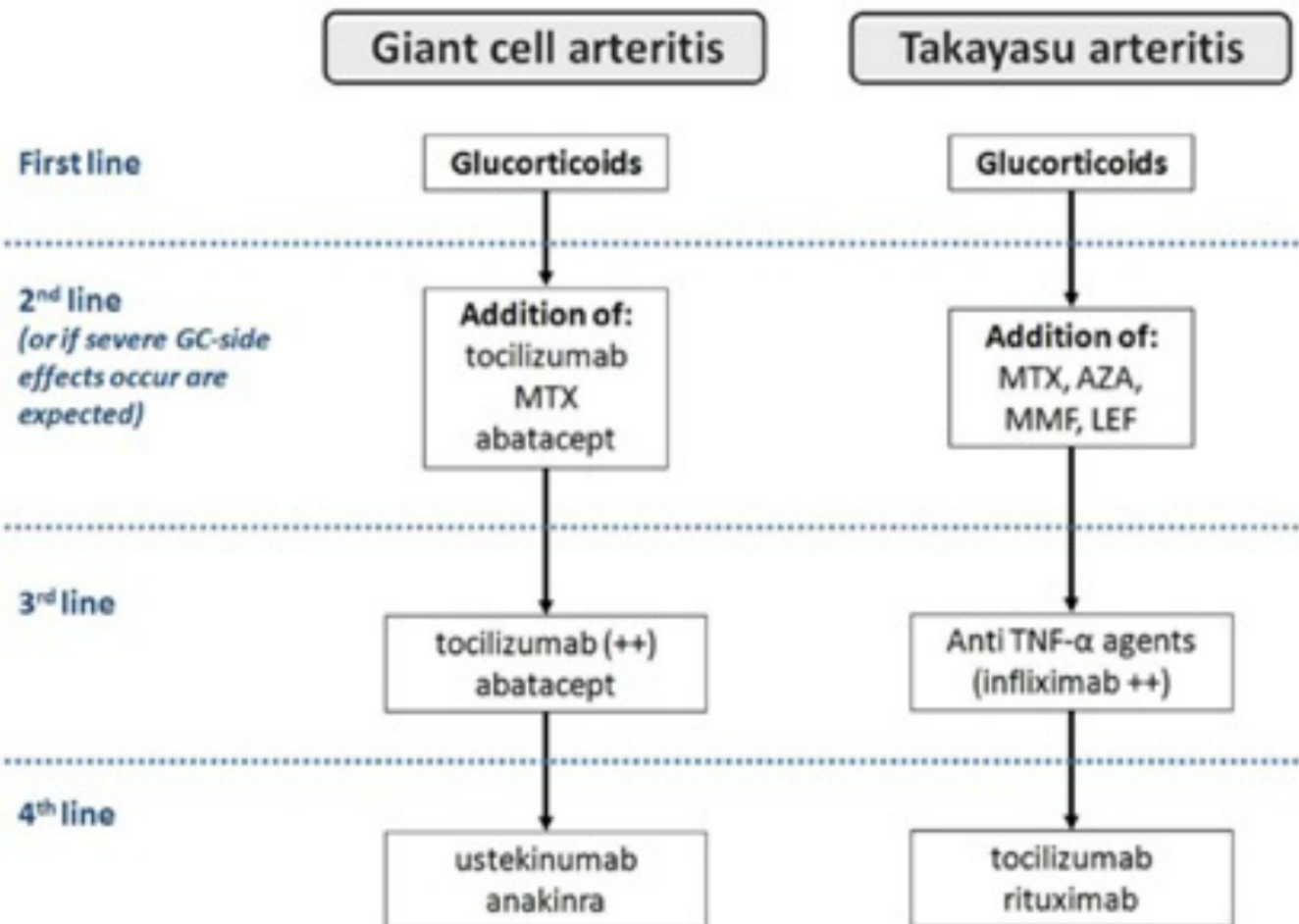
Asian females under 40

Arm and leg pain,
confusion, weak pulses,
shortness of breath

Fatigue, fever, weight loss

ESR and CRP

Treatable but need
to suspect it



Samson et al. European Journal Internal Medicine 2018

Medium Vessel diseases

Behets

More common in middle eastern and Mediterranean people

Oral and genital ulcers, eye inflammation and arthritis

Males affected worse than women

No diagnostic tests

Treatable with immunosuppressants

Main complication is vision loss

Kawasakis

Affects kids

Fever for 5 days

Redness in eyes and swelling in hands and feet

Main danger is to coronaries

Needs to get echo and ?angio

Polyarteritis Nodosa (PAN)

Affects ages 40-60

Identified with HEP B infection

No specific test

Immunosuppression for symptom control

Rheumatoid Vasculitis

Complication of RA

More common in male smokers with RA

Any organ affected

Potentially life threatening

Treatment of RA helps control RV

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Complication of RA

More common in male smokers with RA

Any organ affected

Potentially life threatening

Treatment of RA helps control RV

Affects ages 40-60

Symptoms include:

- fever
- feeling tired
- decreased appetite
- rapid and sudden weight loss
- skin rash or sores
- muscle aches
- joint pain
- abdominal pain
- blood in the stool
- flank pain
- testicular pain in men
- chest pain
- difficulty breathing
- difficult to treat hypertension (high blood pressure)
- numbness or tingling of the hands or feet
- sudden loss of strength in the hands or feet

Idiopathic but
associated with
Hep B infection

No specific test

Immunosuppression
for symptom control

Medium Vessel diseases

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Oral and genital ulcers, eye inflammation and arthritis

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More common
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Any organ
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Potentially life threatening

Treatment of RA helps control
RV

Small vessel diseases

Granulomatosis with polyangitis (Wegeners)

Affects any organ but often effects lungs, kidneys, sinuses

Inflamed areas may become granulomatous

90% have a c-ANCA that doesn't respond to URTI Tx

Symptoms depends on organs effected

Team approach

c-ANCA PR3

immunosuppression

Eosinophilic granulomatosis with polyangitis (Churg Straus disease)

Rhinitis, asthma, raised eosinophils

ANCA associated

Purpura and vasculitis visible

90% treated effectively with steroids, however asthma can persist

Cardiac complications are main issue

Mainly affects over 50s

Microscopic polyangitis

Effects almost any organ

Also ANCA associated

Myeloperoxidase antibody present

Similar to GPA - immunosuppressants for remission but not cure

Team approach

Bechets disease

Henoch Schonlein purpura

Most common vasculitis in children

Skin rash, abdo pain and arthritis

Rash is typically purpura

Associated with nephritic syndrome and occasionally intussusception

Self limited illness. Kidney problems can become chronic

Cryoglobulinemia

Deposition of immune complexes

Affects patients with HCV

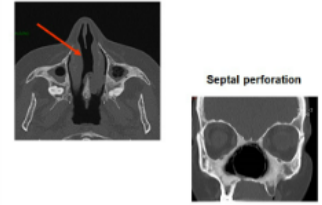
Skin, joints and nerves are involved

Liver, spleen, kidneys have malignancy nodules and ptx

If HCV neg = Anti viral

Obese - immunosuppressants

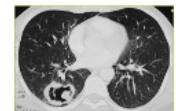
Affects any organ but often effects lungs, kidneys, sinuses



Inflamed areas may become granulomatous



90% have a cold that doesn't respond to URTI Tx

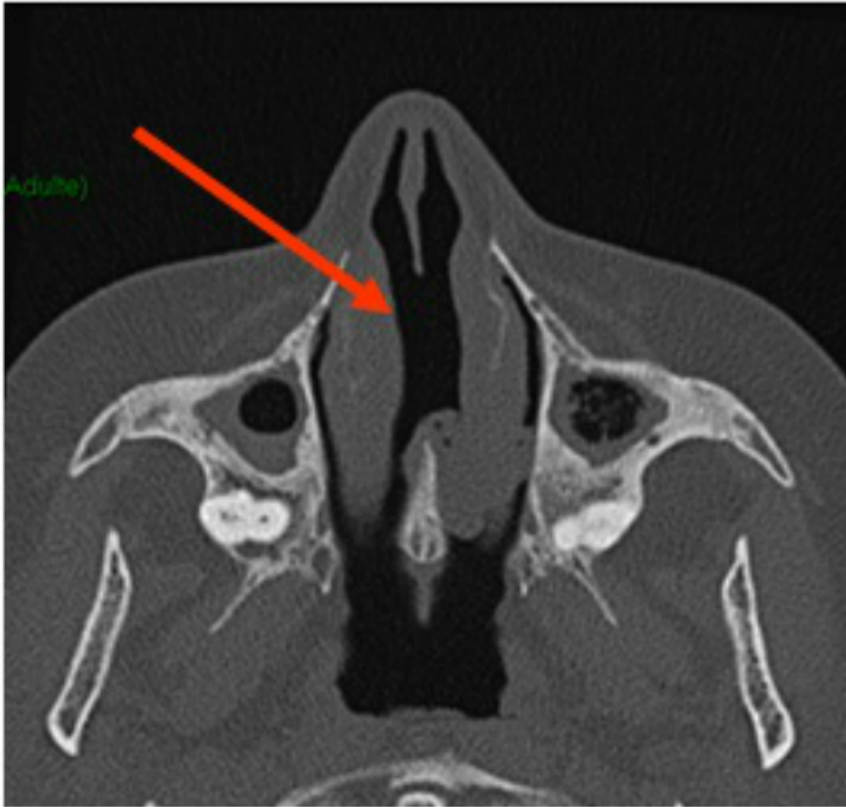


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cANCA PR3

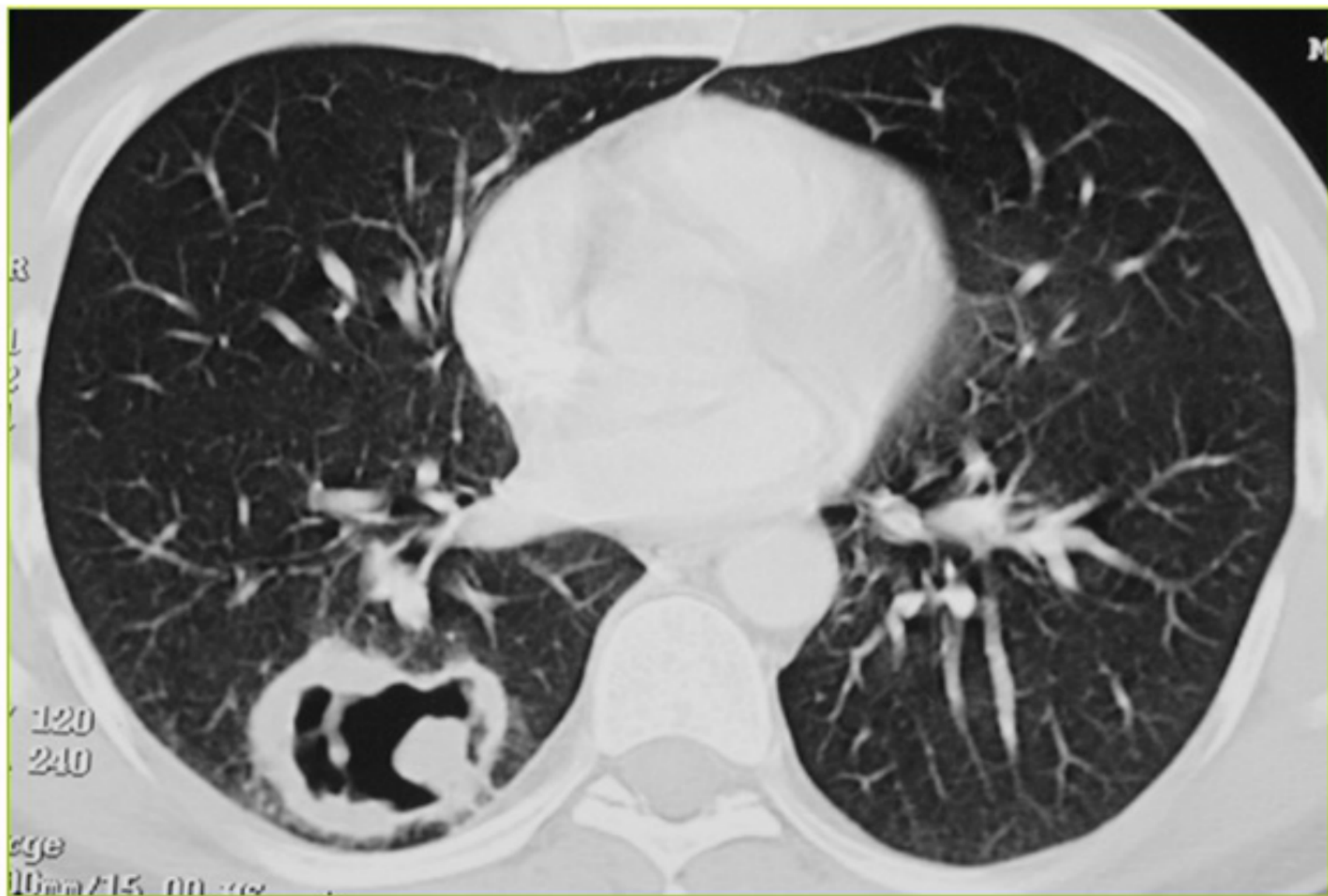
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Septal perforation







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Self limited illness. Kidney problems can become chronic

Cryoglobulinemia

Deposition of immune complexes
Affects patients with HCV
Skin, joints and nerves are involved
Lungs, kidneys, liver have malignancy associated with this
If HCV neg = Anti viral
Obese - immunosuppressants

Rhinitis, asthma, raised eosinophils

ANCA associated

Purpura and mononeuritis multiplex

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Affects patients with HCV

Skin, joints and nerves are involved

Lupus, dengue, HIV and many malignancy exclude this one

If HCV neg = Anti viral

Obese - immunosuppressants

Deposition of immune complexes

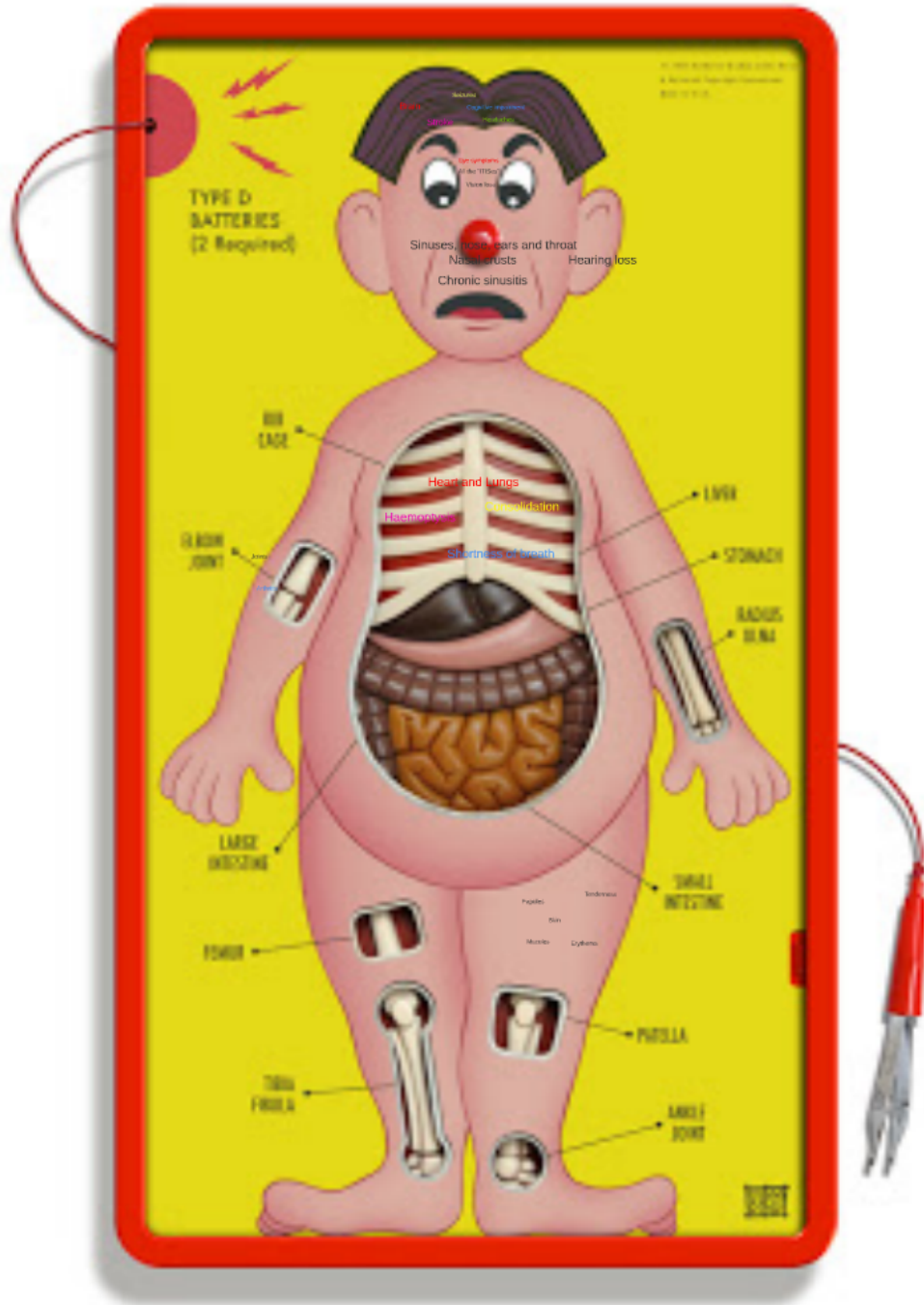
Affects patients with HCV

Skin, joints and nerves but sometimes kidneys

Lupus, sjorgens, RA or haem malignancy patients can get it

If HCV +ve = Anti virals

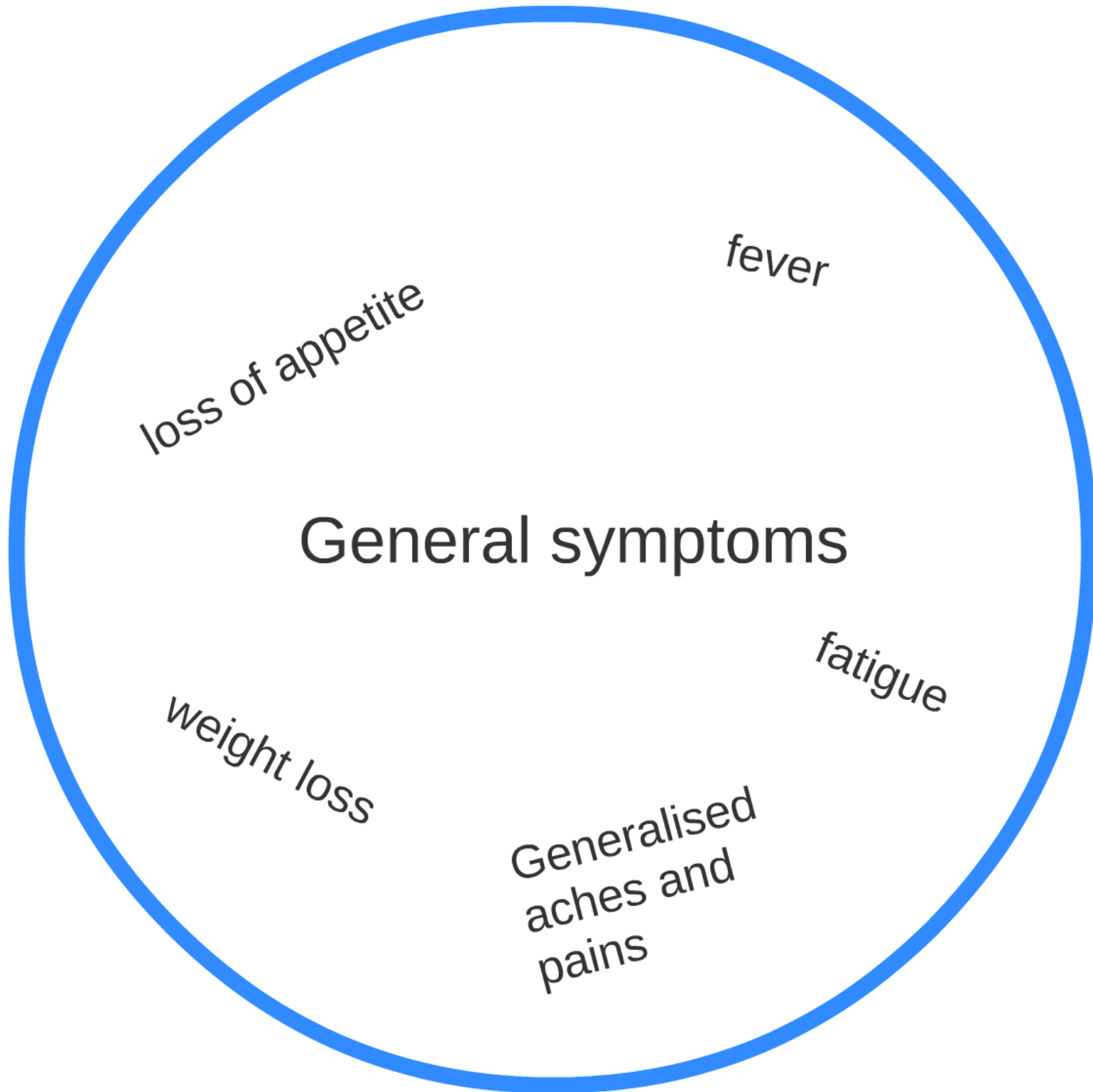
Otherwise - immunosuppresants

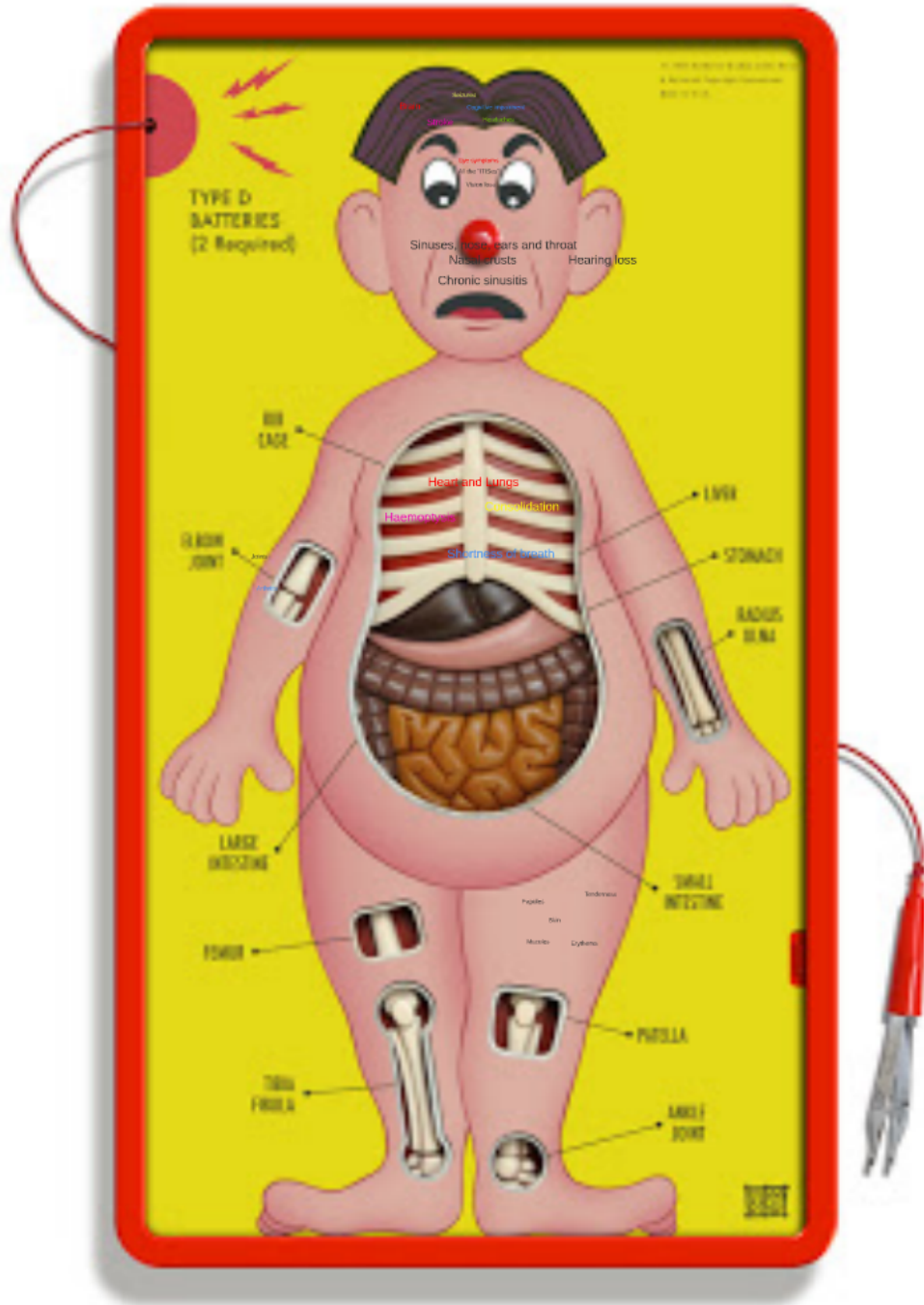
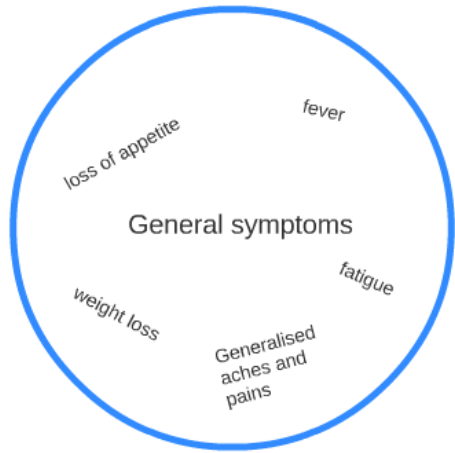


Diagnostic Approach

BMJ 2006; 333:409-416







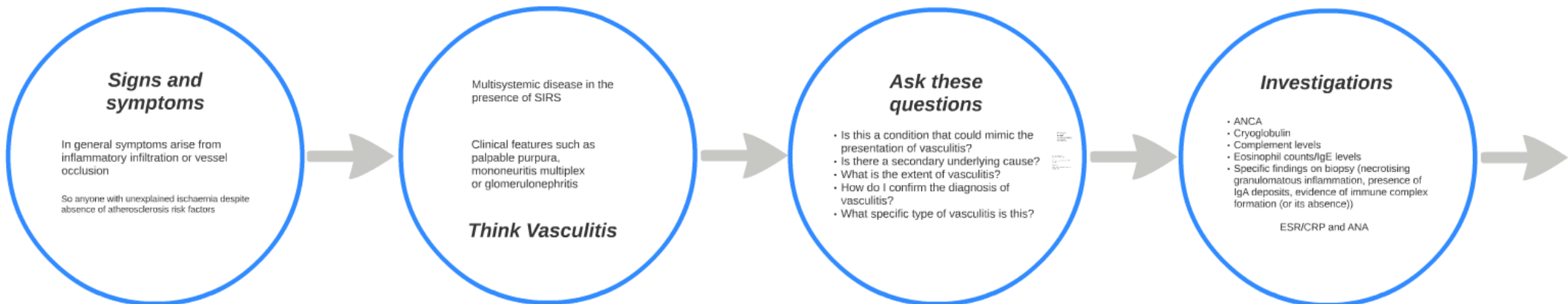
Diagnostic Approach

BMJ 2006;332:409-416
doi:10.1136/bmj.332.7804.409



Diagnostic Approach

Postgrad Med J. Aug 2006; 82(970): 483–488.
doi: 10.1136/pgmj.2005.042648

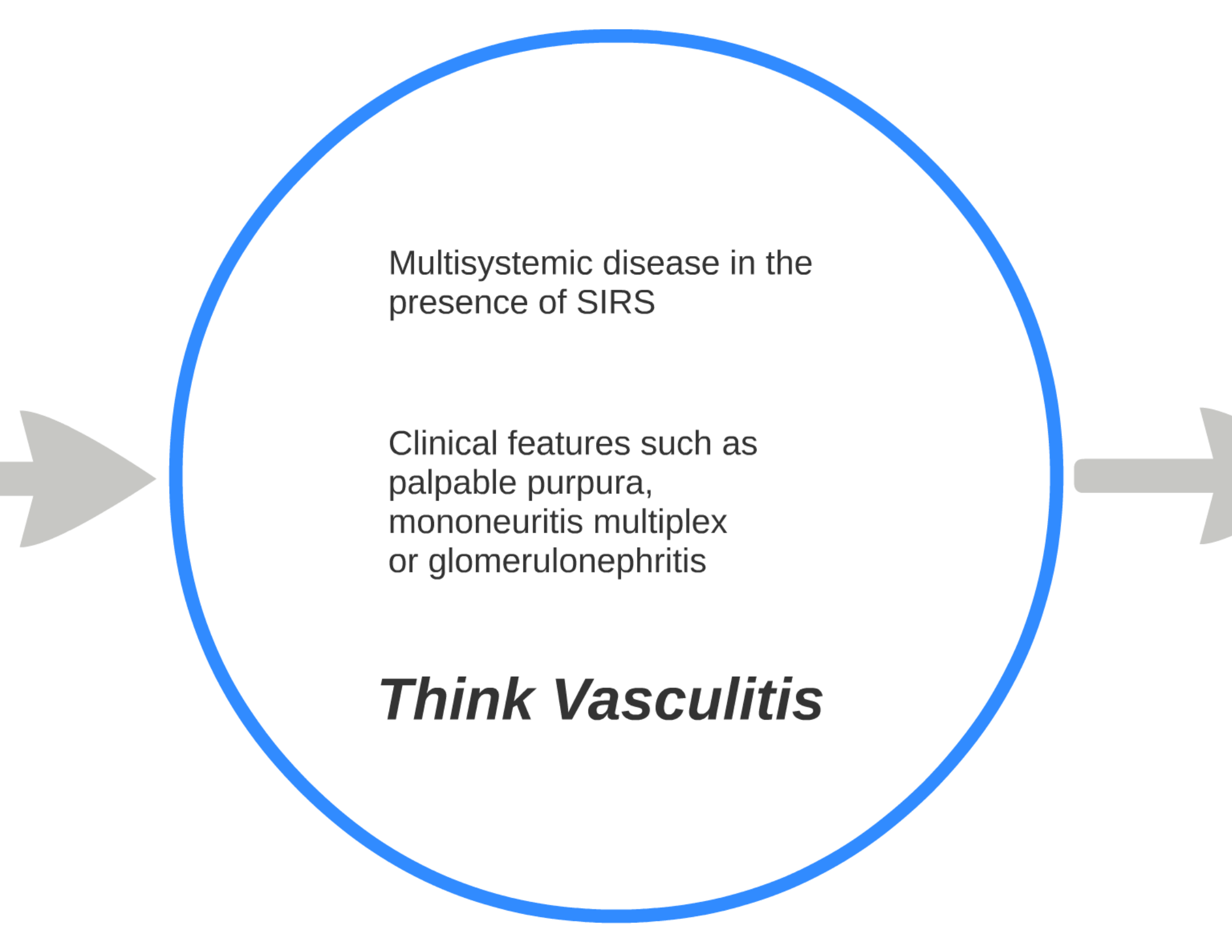


Signs and symptoms

In general symptoms arise from inflammatory infiltration or vessel occlusion

So anyone with unexplained ischaemia despite absence of atherosclerosis risk factors





Multisystemic disease in the
presence of SIRS

Clinical features such as
palpable purpura,
mononeuritis multiplex
or glomerulonephritis

Think Vasculitis

Ask these questions

- Is this a condition that could mimic the presentation of vasculitis?
- Is there a secondary underlying cause?
- What is the extent of vasculitis?
- How do I confirm the diagnosis of vasculitis?
- What specific type of vasculitis is this?

Infections,
emboli,
antiphospholipid
syndrome

Systemic disease
EBV, SLE, ANCA, SJS
Hypersensitivity vasculitis, C-ANCA
Sjögren's
Drug (granulomatous vasculitis
reaction)

Infections,
emboli,
antiphospholipid
syndrome

Ask these questions

- Is this a condition that could mimic the presentation of vasculitis?
- Is there a secondary underlying cause?
- What is the extent of vasculitis?
- How do I confirm the diagnosis of vasculitis?
- What specific type of vasculitis is this?

Infections,
emboli,
antiphospholipid
syndrome

Systemic disease

Medication (e.g. C. PPI,
Sulfonamides)

Smoking

Drug (granulomatous hepatitis,
hepatitis)

Inflammatory disease
(IBD, Sarcoidosis, SLE)

Infectious diseases (Hep B, C, HIV,
Syphilis)

Neoplasm

Drugs (propylthiouracil, montelukast,
hydralazine)

Ask these questions

- Is this a condition that could mimic the presentation of vasculitis?
- Is there a secondary underlying cause?
- What is the extent of vasculitis?
- How do I confirm the diagnosis of vasculitis?
- What specific type of vasculitis is this?

Infections,
emboli,
antiphospholipid
syndrome

Systemic disease

EBV, SLE, RA, etc.

Medications (e.g. C, H, S)

Drugs (e.g. methimazole, levamisole)

Drugs (e.g. methimazole, levamisole)

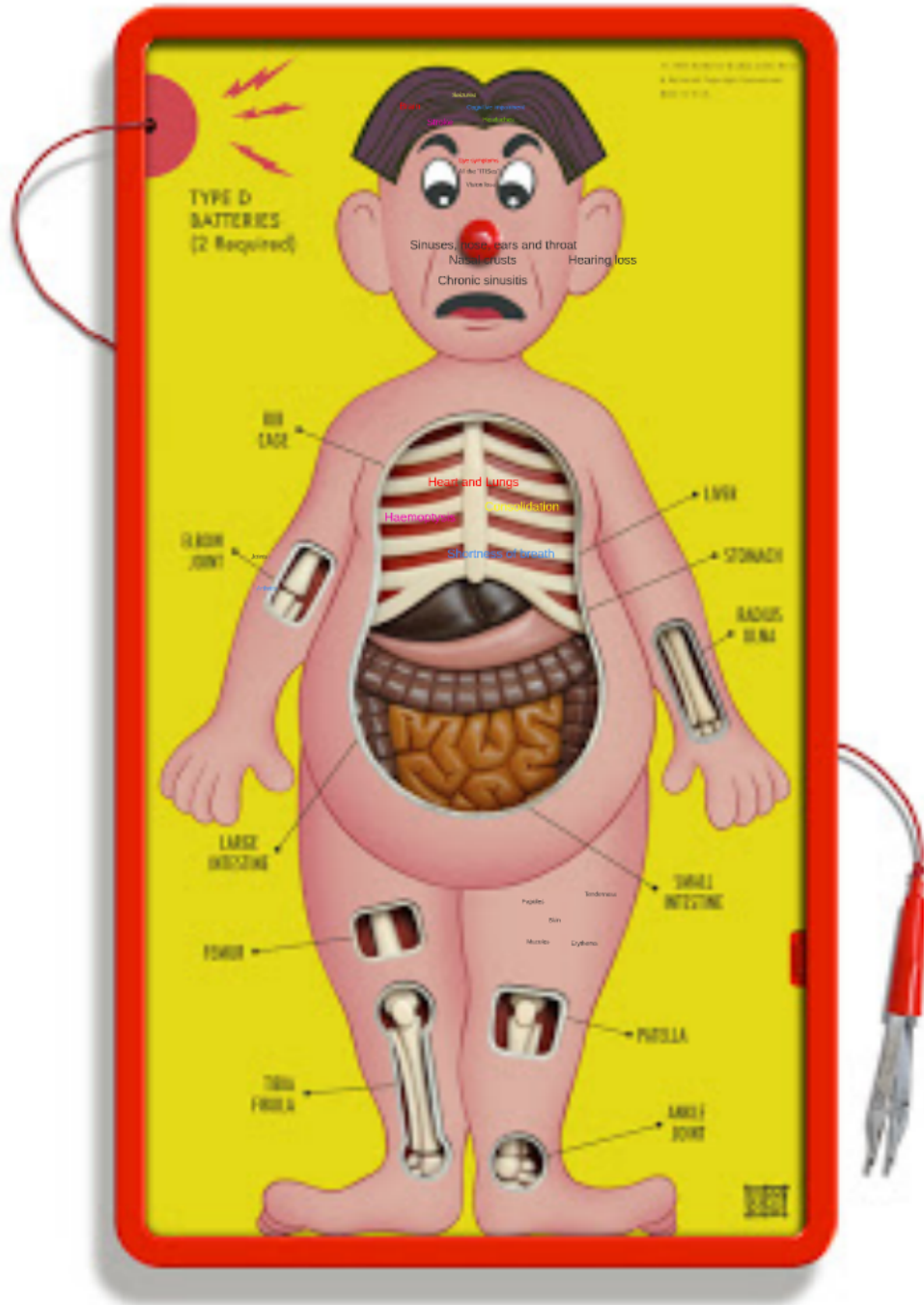
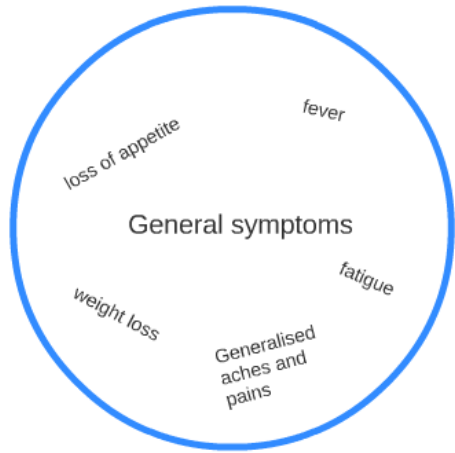
Drugs (e.g. methimazole, levamisole)



Investigations

- ANCA
- Cryoglobulin
- Complement levels
- Eosinophil counts/IgE levels
- Specific findings on biopsy (necrotising granulomatous inflammation, presence of IgA deposits, evidence of immune complex formation (or its absence))

ESR/CRP and ANA



Diagnostic Approach

BMJ 2006; 333:409-416





Eye symptoms

All the "ITISes"

Vision loss



Brain

Seizures

Cognitive impairment

Stroke

Headaches

Eye symptoms

All the "ITISes"

Vision loss

Sinuses, nose, ears and throat



Sinuses, nose, ears and throat

Nasal crusts

Hearing I

Chronic sinusitis

An anatomical diagram of the thoracic cavity, showing the rib cage and the internal organs. The heart is located in the center, and the lungs are on either side. The diagram is color-coded to highlight specific conditions: red for the heart and lungs, yellow for consolidation, magenta for haemoptysis, and blue for shortness of breath.

Heart and Lungs

Consolidation

Haemoptysis

Shortness of breath



Joints

Arthritis

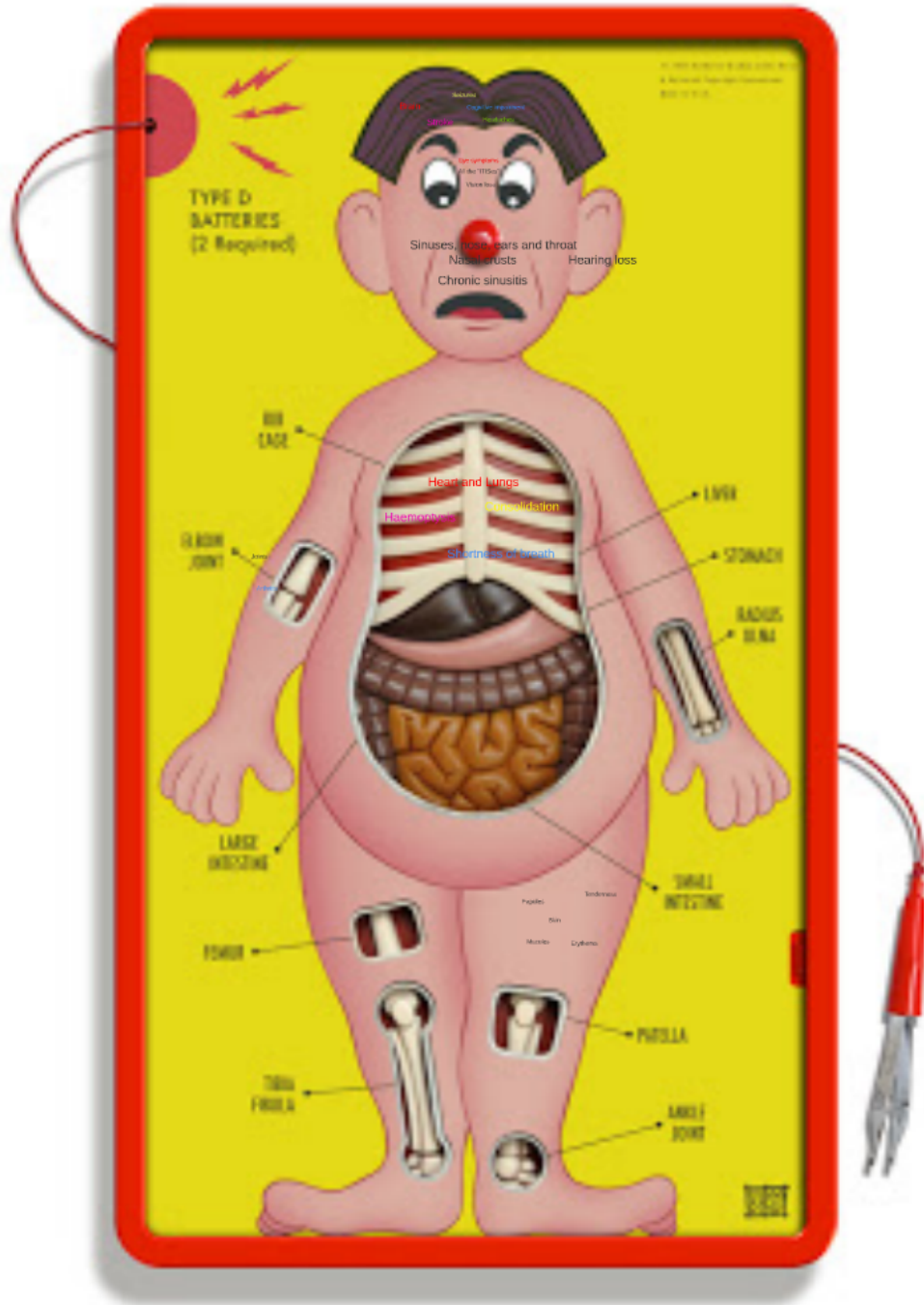
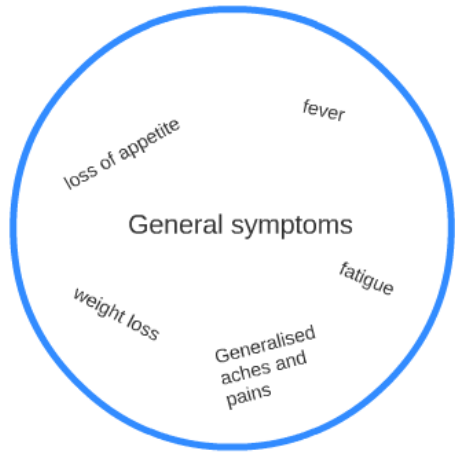
Tenderness

Papules

Skin

Macules

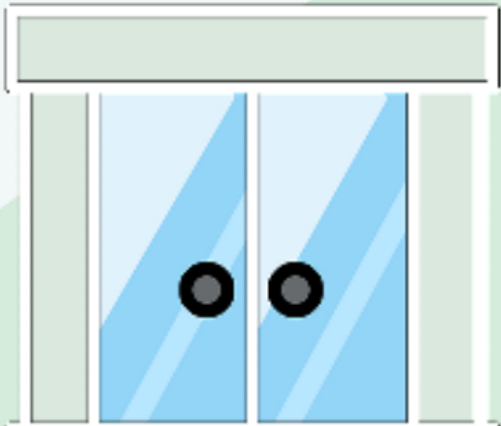
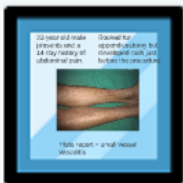
Erythema



Diagnostic Approach

BMJ 2006; 333:409-416





23 year old male presents and a 14 day history of abdominal pain.

Booked for appendicectomy but developed rash just before the procedure



Histo report = small vessel vasculitis

37 year old female with 5 hr history of upper and lower limb weakness on waking from sleep Flaccid tone

Pain in distal upper limbs on exertion relieved by rest

O/E

decreased radial and brachial pulses

30 year old male presents with bilateral
pyramidal signs and orogenital
ulcerations

He's Turkish!

15 year old female presents with persistent sinusitis symptoms with severe nasal crusting

Dismissed as allergies for several years

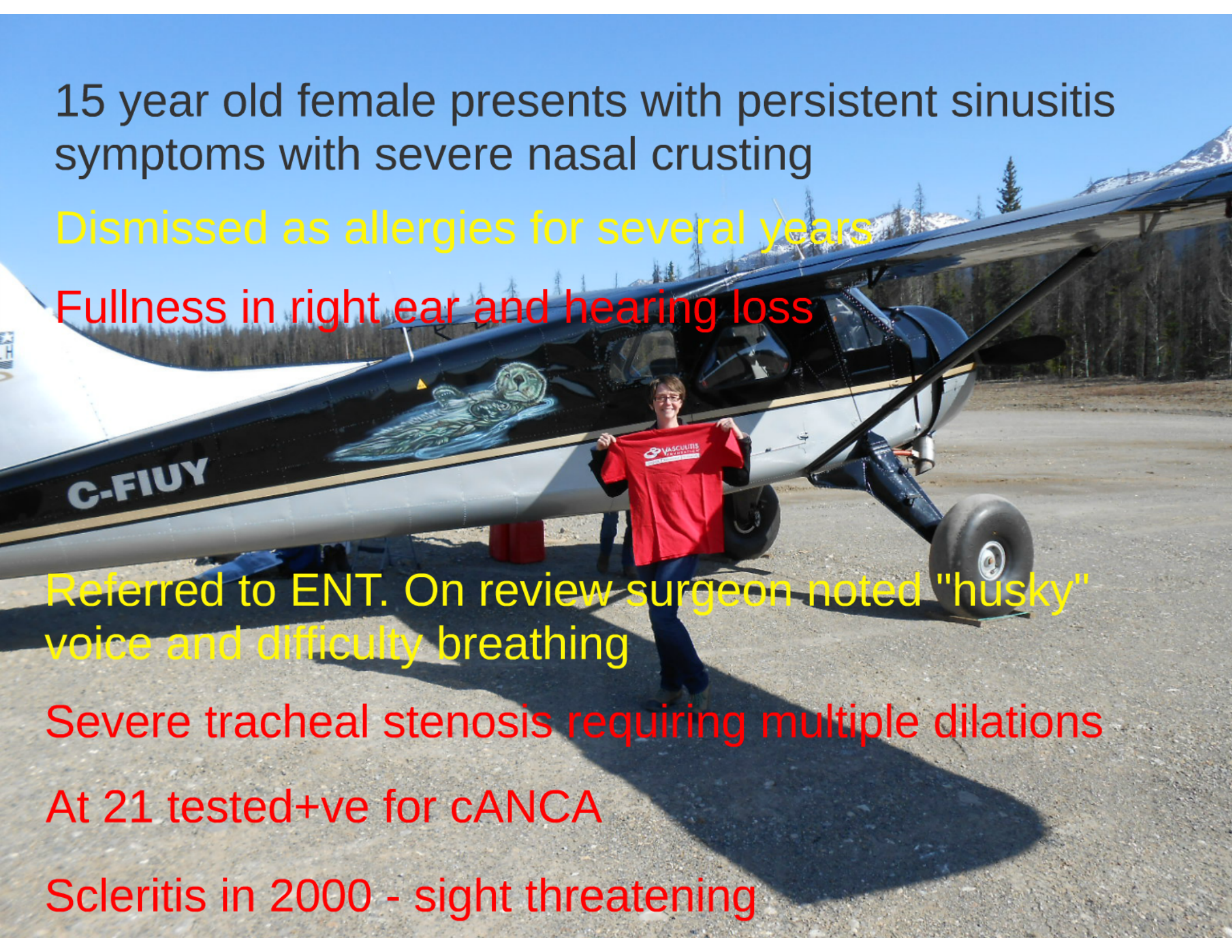
Fullness in right ear and hearing loss

Referred to ENT. On review surgeon noted "husky" voice and difficulty breathing

Severe tracheal stenosis requiring multiple dilations

At 21 tested +ve for cANCA

Scleritis in 2000 - sight threatening





Principles of treatment of severe, systemic GPA

CYCLOPHOSPHAMIDE

IV (pulse): 15 mg/kg at D1,15,29 then /3 wk



Oral (continuous): 2 mg/kg/day

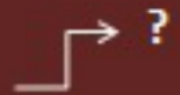


→ AZATHIOPRINE 2 mg/kg/day

→ METHOTREXATE 0.3 mg/kg/wk

→ LEFLUNOMIDE 20 mg/day

→ MYCOPHENOLATE MOFETIL 2 g/day



RITUXIMAB

375 mg/m²/wk (or 1 g at D1 & 15)



+ Glucocorticoids

R

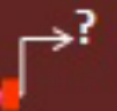
3 - 6 months

> 18 months

± Plasma exchange?

INDUCTION

MAINTENANCE





You **Tube**