CARDIOVASCULAR EVENTS IN RHEUMATOLOGIC DISORDERS

M. Saghafi

Professor of Rheumatology

For nearly half a century, excess rates of cardiovascular disease (CVD) have been reported among patients with inflammatory rheumatic diseases.

Cardiovascular (CV) mortality and morbidity, in particular, ischemic heart disease and heart failure, are significantly higher among persons with rheumatoid arthritis (RA) and/or systemic lupus erythematosus (SLE) and likely other autoimmune disorders compared with persons in the general population of the same age.

Frequencies of Various Manifestations of Systemic Lupus Erythematosus*

Manifestation	Frequency
Constitutional symptoms (fatigue, fever, weight loss)	90%-95%
Mucocutaneous involvement (malar rash, alopecia, mucosal ulcers, discoid lesions, etc.)	80%-90%
Musculoskeletal involvement (arthritis/arthralgia, avascular necrosis, myositis, etc.)	80%-90%
Serositis (pleuritis, pericarditis, peritonitis)	50%-70%
Glomerulonephritis	40%-60%
Neuropsychiatric involvement (cognitive impairment, depression, psychosis, seizures, stroke, demyelinating syndromes, peripheral neuropathy, etc.)	40%-60%
Autoimmune cytopenia (anemia, thrombocytopenia)	20%-30%

Extra-articular manifestations in spondyloarthritis

Extra-articular manifestations	Clinical manifestations
Cardiac	Aortitis; aortic insufficiency; conduction disorders; bundle-branch and atrioventricular blocks
Renal	Secondary amyloidosis; IgA nephropathy
Pulmonary	Fibrosis of the upper lobe and pleural thickening
Neurological	Cauda equina syndrome

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Low prevalence of renal, cardiac, pulmonary, and neurological extra-articular clinical manifestations in spondyloarthritis: analysis of the Brazilian Registry of Spondyloarthritis Carlos Ewerton Maia Rodrigues[I]

Prevalence of the extra-articular manifestations in spondylo-arthritis according to the clinical diagnosis

Clinical diagnosis of spondyloarthritis (n = 1,472)	Extra-articular manifestations			
	Cardiac (n = 44)	Renal (n = 17)	Pulmonary (n = 19)	Neurological (n = 13)
Primary AS	29 (2%)	5 (0.3%)	14 (1.0%)	8 (0.5%)
AS + psoriasis	1 (0.1%)	1 (0.1%)	0	0
PsA	11 (0.7%)	6 (0.4%)	1 (0.1%)	3 (0.2%)
ReA	1 (0.1%)	1 (0.1%)	1 (0.1%)	0
uSpA	1 (0.1%)	0	1 (0.1%)	0
AS + IBD	0	0	1 (0.1%)	0
Arthritis + IBD	0	1 (0.1%)	0	1 (0.1%)
Juvenile SpA	1 (0.1%)	3 (0.2%)	1 (0.1%)	1 (0.1%)

AS: ankylosing spondylitis; PsA: psoriatic arthritis; ReA: reactive arthritis; uSpA: undifferentiated spondyloarthritis; IBD: inflammatory bowel disease; juvenile SpA: juvenile spondyloarthritis.

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Prevalence of the extra-articular manifestations of spondylo-arthritis according to the clinical form

Extra-articular manifestations	Clinical form of spondyloarthritis (n = 1,472)			
	Axial	Peripheral	Mixed	Enthesitic
Cardiac (n = 44)	12 (0.8%)	6 (0.4%)	25 (1.7%)	1 (0.1%)
Renal (n = 17)	2 (0.1%)	2 (0.1%)	13 (0.9%)	0
Pulmonary (n = 19)	9 (0.7%)	0	10 (0.7%)	0
Neurological (n = 13)	2 (0.1%)	2 (0.1%)	9 (0.5%)	0

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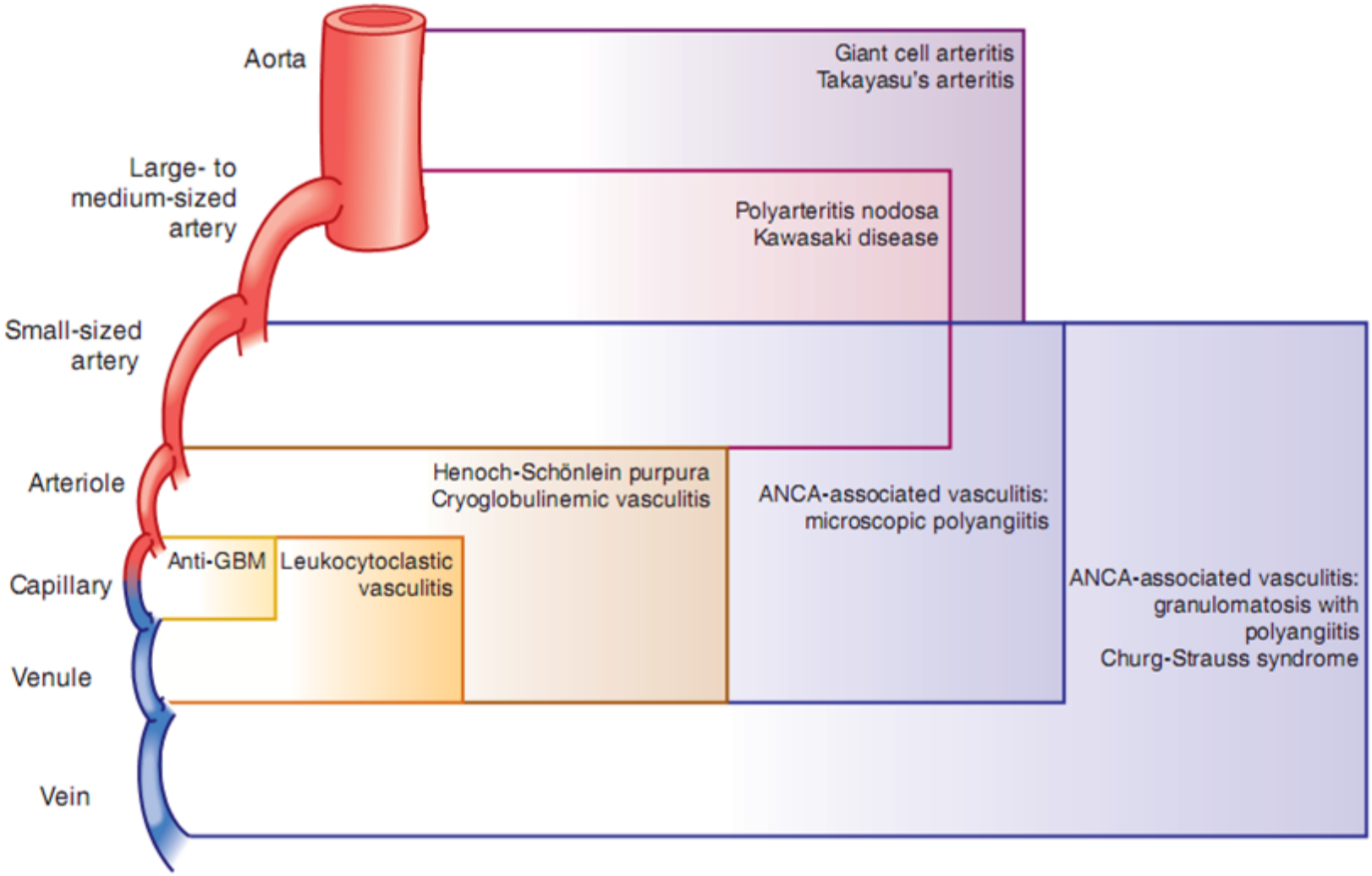
Medsger Systemic Sclerosis Severity Scale*

Organ System	0 (Normal)	1 (Mild)	2 (Moderate)	3 (Severe)	4 (End Stage)
General	Wt loss <5%; Ht 37%+; Hb 12.3+ g/dL	Wt loss 5%-10%; Ht 33%-37%; Hb 11.0-12.2 g/dL	Wt loss 10%-15%; Ht 29%-33%; Hb 9.7-10.9 g/dL	Wt loss 15%-20%; Ht 25%-29%; Hb 8.3-9.6 g/dL	Wt loss 20+%; Ht 25%; Hb <8.3 g/dL
Peripheral vascular	No Raynaud's; Raynaud's not requiring vasodilators	Raynaud's requiring vasodilators	Digital pitting scars	Digital tip ulcerations	Digital gangrene
Skin	TSS 0	TSS 1-14	TSS 15-29	TSS 30-39	TSS 40+
Joint/tendon	FTP 0-0.9 cm	FTP 1.0-1.9 cm	FTP 2.0-3.9 cm	FTP 4.0-4.9 cm	FTP 5.0+ cm
Muscle	Normal proximal muscle strength	Proximal weakness, mild	Proximal weakness, moderate	Proximal weakness, severe	Ambulation aids required
Gastrointestinal tract	Normal esophagogram; normal small bowel series	Distal esophageal hypoperistalsis; small bowel series abnormal	Antibiotics required for bacterial overgrowth	Malabsorption syndrome; episodes of pseudo-obstruction	Hyperalimentation required
Lung	DLCO 80+%; FVC 80+%; no fibrosis on radiograph; sPAP <35 mm Hg	DLCO 70%-79%; FVC 70%-79%; basilar rales; fibrosis on radiograph; sPAP 35-49 mm Hg	DLCO 50%-69%; FVC 50%-69%; sPAP 50-64 mm Hg	DLCO <50%; FVC <50%; sPAP 65+ mm Hg	Oxygen required
Heart	ECG normal; LVEF 50+%	ECG conduction defect; LVEF 45%-49%	ECG arrhythmia; LVEF 40%-44%	ECG arrhythmia requiring therapy; LVEF 30%-40%	CHF; LVEF <30%
Kidney	No history of SRC with serum creatinine <1.3 mg/dL	History of SRC with serum creatinine <1.5 mg/dL	History of SRC with serum creatinine 1.5-2.4 mg/dL	History of SRC with serum creatinine 2.5-5.0 mg/dL	History of SRC with serum creatinine >5.0 mg/dL or dialysis required

*If two items are included for a severity grade, only one is required for the patient to be scored as having disease of that severity level.

CHF, congestive heart failure; DLCO, diffusing capacity for carbon monoxide, % predicted; ECG, electrocardiogram; FTP, fingertip-to-palm distance in flexion; FVC, forced vital capacity, % predicted; Hb, hemoglobin; Ht, hematocrit; LVEF, left ventricular ejection fraction; sPAP, estimated pulmonary artery pressure by Doppler echo; SRC, scleroderma renal crisis; TSS, total skin score; Wt, weight.

Adapted from Medsger TA Jr, Bombardieri S, Czirjak L, et al: Assessment of disease severity and prognosis, *Clin Exp Rheumatol* 21(3 Suppl 29):S51, 2003.



Classification by blood vessel size. ANCA, antineutrophil cytoplasmic antibody; GBM, glomerular basement membrane.

Typical Clinical Manifestations of Large, Medium, Involvement by Vasculitis

Large	Medium
Limb claudication	Cutaneous nodules
Asymmetric blood pressures	Ulcers
Absence of pulses	Livedo reticularis
Bruits	Digital gangrene
Aortic dilation	Mononeuritis multiplex
Renovascular hypertension	Microaneurysms
	Renovascular hypertension

Revised Jones Criteria for Diagnosis of Acute Rheumatic Fever*

Major Manifestations
Polyarthritis Chorea Subcutaneous nodules Erythema marginatum Carditis
Minor Manifestations
Arthralgia Fever Previous rheumatic fever or rheumatic heart disease
Supporting Evidence of Preceding Streptococcal Infection
Positive throat culture for group A beta-hemolytic streptococci Increased antistreptolysin O or other streptococcal antibodies Recent scarlet fever
Other Findings
Elevated acute phase reactants (C-reactive protein or erythrocyte sedimentation rate) Prolonged P-R interval

Summary of 2002 World Health Organization Criteria for the Diagnosis of Rheumatic Fever (RF) and Rheumatic Heart Disease (RHD)

Diagnostic Categories	Criteria
Primary episode of RF	Two major or one major and two minor manifestations plus evidence of a preceding group A streptococcal infection
Recurrent attack of RF in patients without established RHD	Two major or one major and two minor manifestations plus evidence of a preceding group A streptococcal infection
Recurrent attack of RF in patients with established RHD	Two minor manifestations plus evidence of a preceding group A streptococcal infection
Rheumatic chorea, insidious onset of rheumatic carditis	One major manifestation or evidence of a preceding group A streptococcal infection
Chronic valve lesions of RHD (i.e., patients presenting for the first time with pure mitral stenosis, mixed mitral valve disease, and aortic valve disease)	No other criteria required for diagnosis of RHD

Cardiovascular Events in:

- Relapsing polychondraitis
- Gout
- Sarcoidosis
- Amyloidosis
- Marfan Syndrom
- Ehler-Danlos Syndrom
- Anti rheumatic medications

Physicians who care for patients with rheumatic diseases should manage these individuals as a high cardiovascular risk subgroup, regardless of their traditional risk factor parameters.