Klasifikace vaskulitid

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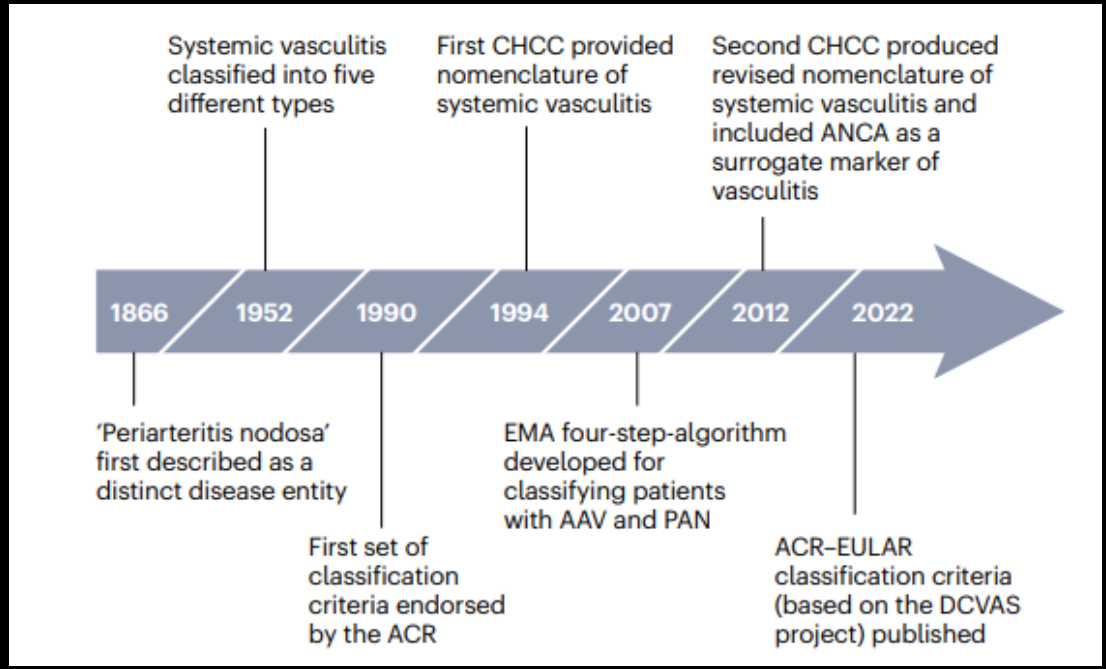
Úskalí klasifikace

- Klinické příznaky: heterogenní skupina x společná symptomatologie (konstitutivní projevy, orgánová postižení)
- Biopsie x zobrazovací metody x laboratorní nálezy
- Klasifikační ≠ diagnostická kritéria
  - Klasifikační -> homogenní, dobře definovaná skupina pacientů
  - Nízká senzitivita ke stanovení diagnózy [1)]

1) Limitations of the 1990 American College of Rheumatology classification criteria in the diagnosis of vasculitis, Roy 1998

Vývoj klasifikace

- ACR 1990 kritéria
  (GCA, TAK, GPA, EGPA, PAN, IgA, hypersensitivity)
- CHCC 1994 klasifikace
  (large x medium x small vessel)
- EMA 2007 algoritmus
  (AAV+PAN)
- CHCC 2012 klasifikace
- ACR/EULAR 2022 kritéria
  (AAV, GCA, TAK)



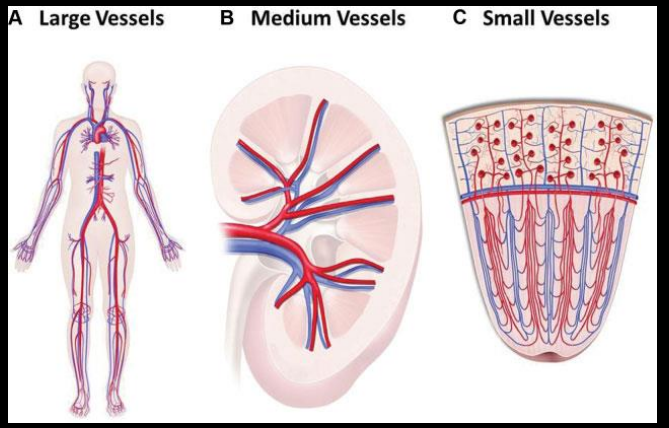
2) The new look of classification criteria for systemic vasculitis, Emmi, Vaglio, 2023

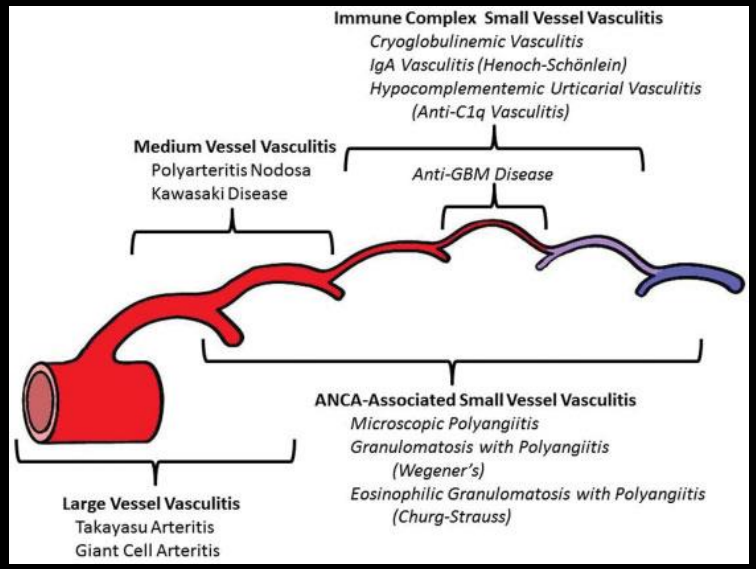
Chapel Hill Consensus Conference 2012

- Multioborová konference, 50% konsensus na návrh, >80% konsensus ke schválení
- Nosologie, kritéria (definice) nejsou validovaná
- Opouštění eponym

- Základ - kalibr cév

+ nové skupiny oproti 1994





3) Overview of the 2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides, Jenette 2012

CHCC 2012

Large vessel vasculitis	Single-organ vasculitis
Takayasu arteritis	Cutaneous leukocytoclastic angiitis
Giant cell arteritis	Cutaneous arteritis
Medium vessel vasculitis	Primary central nervous system vasculitis
Polyarteritis nodosa (PAN)	Isolated aortitis
Kawasaki disease	Others
Small vessel vasculitis	**Vasculitis associated with systemic disease**
ANCA associated vasculitis (AAV)	Lupus vasculitis
Microscopic polyangiitis (MPA)	Rheumatoid vasculitis
Granulomatosis with polyangiitis (GPA)	Sarcoid vasculitis
Eosinophilic granulomatosis with polyangiitis (EGPA)	Others
Immune complex vasculitis	**Vasculitis associated with probable etiology**
Anti-glomerular basement membrane (anti-GBM) disease	Hepatitis C virus-associated cryoglobulinemic vasculitis
Cryoglobulinemic vasculitis	Hepatitis B virus-associated vasculitis
IgA vasculitis (Henoch-Schönlein)	Syphilis-associated aortitis
Hypocomplementemic urticarial vasculitis (anti-C1q vasculitis)	Drug-associated immune complex vasculitis
Variable vessel vasculitis	Drug-associated ANCA-associated vasculitis
Behçet's disease	Cancer-associated vasculitis
Cogan's syndrome	Others

4) https://dx.doi.org/10.26044/ecr2020/C-14405

Příklady definice CHCC 2012

- **Large vessel vasculitis (LVV):** Vasculitis affecting large arteries more often than other vasculitides. Large arteries are the aorta and its major branches. Any size artery may be affected.
  - **Takayasu arteritis (TAK)**
    - Arteritis, often granulomatous, predominantly affecting the aorta and/or its major branches. Onset usually in patients younger than 50 years.
  - **Giant cell arteritis (GCA)**
    - Arteritis, often granulomatous, usually affecting the aorta and/or its major branches, with a predilection for the branches of the carotid and vertebral arteries. Often involves the temporal artery. Onset usually in patients older than 50 years and often associated with polymyalgia rheumatica.

ACR/EULAR 2022
- nová klasifikační kritéria

- Projekt DCVAS (Diagnostic and Classification criteria in VASculitis)
  - 136 pracovišť ve 32 zemích - 2011-2017
  - 6991 pacientů:

    ~ 5000 případů systémové vaskulitidy

    ~ 2000 komparátorů (tj. klinických imitátorů vaskulitidy)

  - klinické a histologické rysy
  - sérologie (ANCA)
  - nálezy zobrazovacích metod (cévní ultrasonografie, angiografie, CT, MRI a PET)

ACR/EULAR 2022 - principy

- Vážený přístup (bodování) – ve smyslu +/-
- Oblasti: klinické, laboratorní, zobrazovací, histologie
- Cut-off skóre ke stanovení dg.

- vypracována na základě případů, které byly prezentovány prospektivně na počátku jejich onemocnění
- reprezentativní případy vybrány panelem expertů – pouze při vysoké/střední míře jistoty diagnózy
- na širším souboru pacientů - zachovaná specificita, ale výrazně snížená senzitivita (ale výrazně lépe než ACR 1990)

ACR/EULAR 2022 - AAV

GPA, ≥ 5 bodů

CLINICAL CRITERIA	
Nasal involvement: bloody discharge, ulcers, crusting, congestion, blockage, or septal defect / perforation	+3
Cartilaginous involvement (inflammation of ear or nose cartilage, hoarse voice or stridor, endobronchial involvement, or saddle nose deformity)	+2
Conductive or sensorineural hearing loss	+1
LABORATORY, IMAGING, AND BIOPSY CRITERIA	
Positive test for cytoplasmic antineutrophil cytoplasmic antibodies (cANCA) or antiproteinase 3 (anti-PR3) antibodies	+5
Pulmonary nodules, mass, or cavitation on chest imaging	+2
Granuloma, extravascular granulomatous inflammation, or giant cells on biopsy	+2
Inflammation, consolidation, or effusion of the nasal/paranasal sinuses, or mastoiditis on imaging	+1
Pauci-immune glomerulonephritis on biopsy	+1
Positive test for perinuclear antineutrophil cytoplasmic antibodies (pANCA) or antimyeloperoxidase (anti-MPO) antibodies	-1
Blood eosinophil count ≥ 1 x10^9/liter	-4

5) 2022 American College of Rheumatology/European Alliance of Associations for Rheumatology classification criteria for granulomatosis with polyangiitis, Robson 2022)

MPA, ≥ 5 bodů

CLINICAL CRITERIA	
Nasal involvement: bloody discharge, ulcers, crusting, congestion, blockage or septal defect / perforation	-3
LABORATORY, IMAGING, AND BIOPSY CRITERIA	
Positive test for perinuclear antineutrophil cytoplasmic antibodies (pANCA) or antimyeloperoxidase (anti-MPO) antibodies ANCA positive	+6
Fibrosis or interstitial lung disease on chest imaging	+3
Pauci-immune glomerulonephritis on biopsy	+3
Positive test for cytoplasmic antineutrophil cytoplasmic antibodies (cANCA) or antiproteinase 3 (anti-PR3) antibodies	-1
Blood eosinophil count ≥ 1 x10^9/liter	-4

6) 2022 American College of Rheumatology/European Alliance of Associations for Rheumatology classification criteria for microscopic polyangiitis, Suppiah 2022

EGPA, ≥ 6 bodů

CLINICAL CRITERIA	
Obstructive airway disease	+3
Nasal polyps	+3
Mononeuritis multiplex	+1
LABORATORY AND BIOPSY CRITERIA	
Blood eosinophil count ≥ 1 x10^9/liter	+5
Extravascular eosinophilic-predominant inflammation on biopsy	+2
Positive test for cytoplasmic antineutrophil cytoplasmic antibodies (cANCA) or antiproteinase 3 (anti-PR3) antibodies	-3
Hematuria	-1

7) 2022 American College of Rheumatology/European Alliance of Associations for Rheumatology Classification Criteria for Eosinophilic Granulomatosis with Polyangiitis, Grayson 2022

ACR/EULAR 2022 - LVV

GCA, ≥ 6 bodů

ABSOLUTE REQUIREMENT	
Age ≥ 50 years at time of diagnosis	
ADDITIONAL CLINICAL CRITERIA	
Morning stiffness in shoulders/neck	+2
Sudden visual loss	+3
Jaw or tongue claudication	+2
New temporal headache	+2
Scalp tenderness	+2
Abnormal examination of the temporal artery[1]	+2
LABORATORY, IMAGING, AND BIOPSY CRITERIA	
Maximum ESR ≥ 50 mm/hour or maximum CRP ≥ 10 mg/liter[2]	+3
Positive temporal artery biopsy or halo sign on temporal artery ultrasound[3]	+5
Bilateral axillary involvement[4]	+2
FDG-PET activity throughout aorta[5]	+2

8) 2022 American College of Rheumatology/EULAR classification criteria for giant cell arteritis, Ponte 2022

TAK, ≥ 5 bodů

ABSOLUTE REQUIREMENTS	
Age ≤ 60 years at time of diagnosis	
Evidence of vasculitis on imaging[1]	
ADDITIONAL CLINICAL CRITERIA	
Female sex	+1
Angina or ischemic cardiac pain	+2
Arm or leg claudication	+2
Vascular bruit[2]	+2
Reduced pulse in upper extremity[3]	+2
Carotid artery abnormality[4]	+2
Systolic blood pressure difference in arms ≥ 20 mm Hg	+1
ADDITIONAL IMAGING CRITERIA	
Number of affected arterial territories (select one)[5]	
One arterial territory	+1
Two arterial territories	+2
Three or more arterial territories	+3
Symmetric involvement of paired arteries[6]	+1
Abdominal aorta involvement with renal or mesenteric involvement[7]	+3

9) 2022 American College of Rheumatology/EULAR classification criteria for Takayasu arteritis, Grayson 2022

Shrnutí

- Nomenklatura vaskulitid na základě CHCC 2012
- Zlatý standard pro stanovení diagnózy = názor klinika
- Klasifikační kritéria by neměla sloužit ke stanovení diagnózy (významný pokles senzitivity na neselektovaných souborech pacientů)
- ACR/EULAR 2022 – detailně statisticky validovaná kritéria
  - primárně k odlišení AAV a LVV mezi sebou
- DCVAS projekt: diagnostická kritéria v budoucnu?