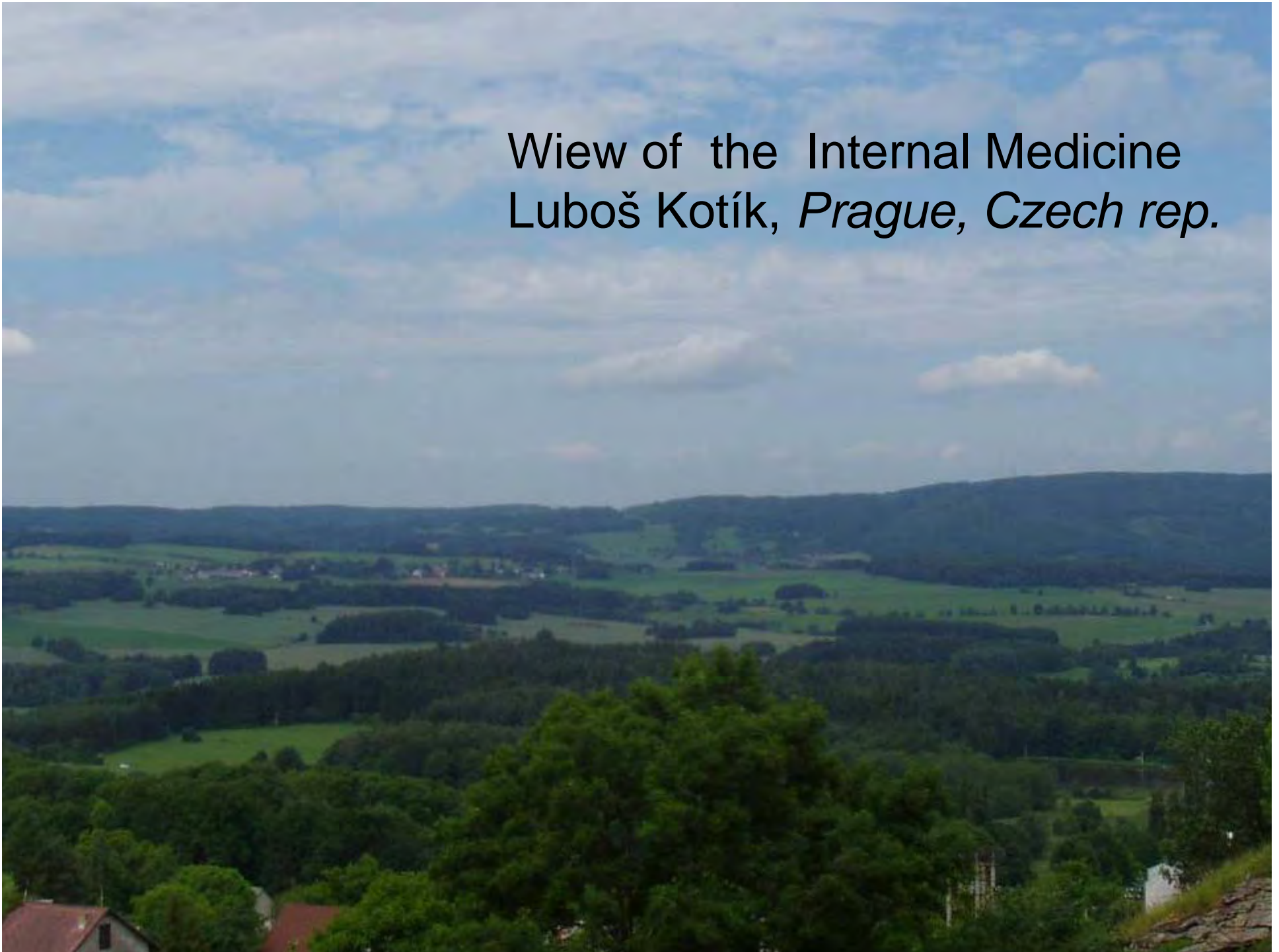


The background of the slide is a soft-focus landscape photograph. It shows a line of trees with sparse, light-colored foliage, possibly in late autumn or winter, set against a pale, hazy sky. The foreground is a field of grass and fallen leaves, also rendered in a soft, painterly style. The overall color palette is muted, with greens, browns, and greys.

# **Polymyalgia rheumatica and Giant cell arteritis**

**Problem not only for rheumatologist.**

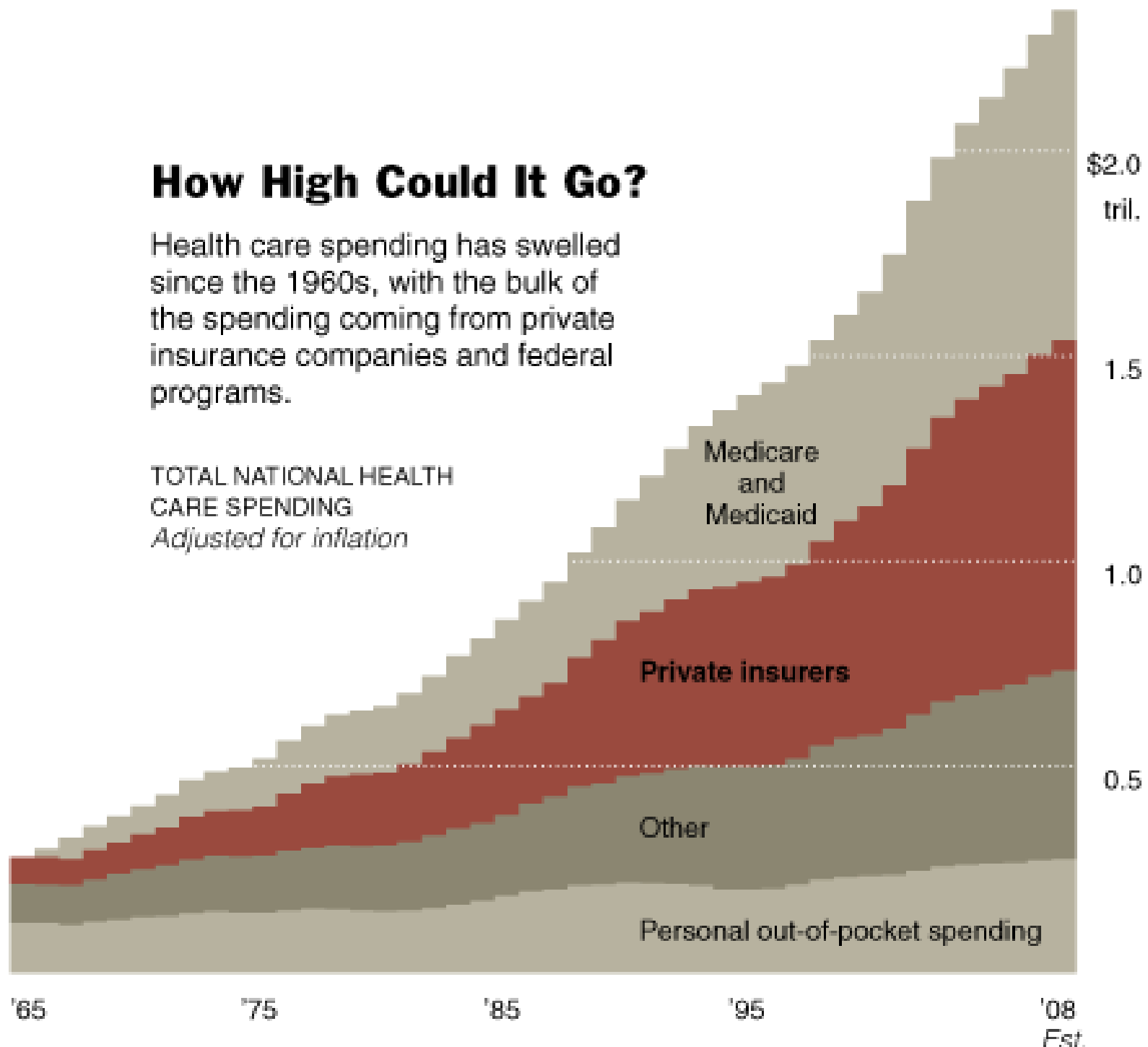
View of the Internal Medicine  
Luboš Kotík, *Prague, Czech rep.*



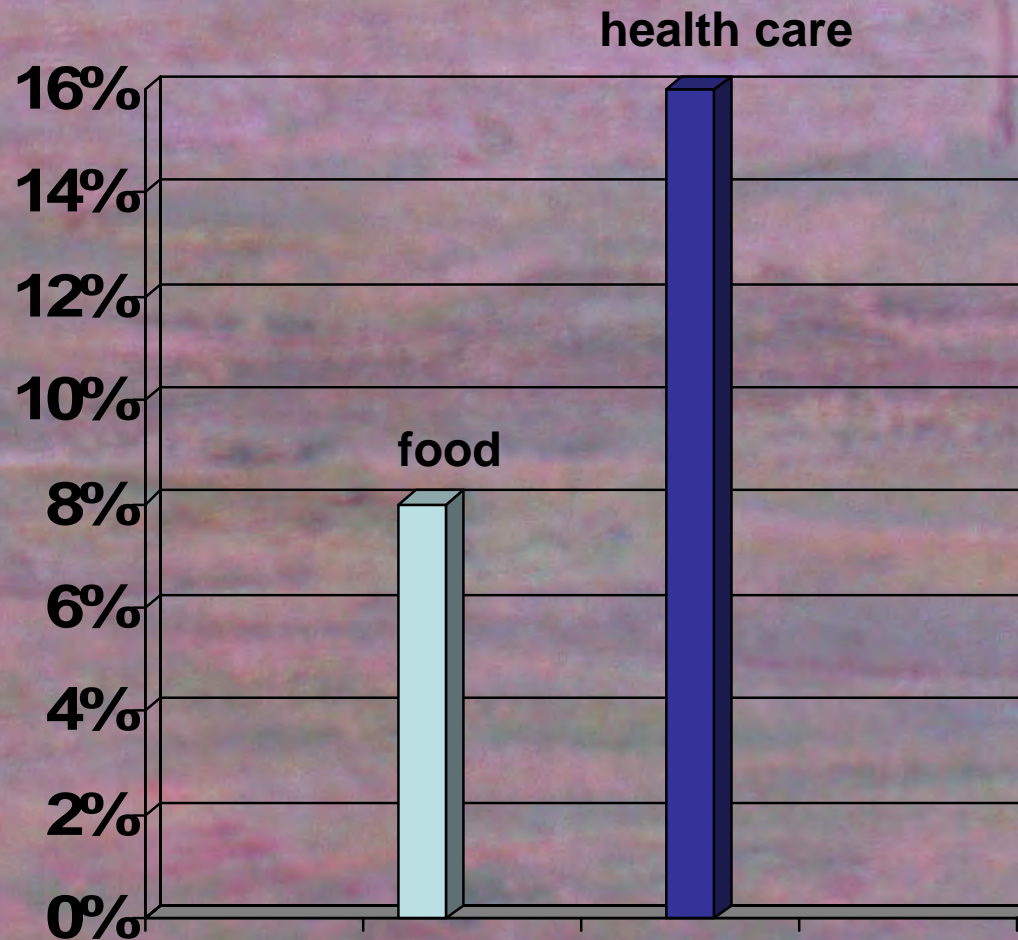
## How High Could It Go?

Health care spending has swelled since the 1960s, with the bulk of the spending coming from private insurance companies and federal programs.

TOTAL NATIONAL HEALTH CARE SPENDING  
*Adjusted for inflation*



# Expenses for food and health care in USA 2008 v % GNP



# Useless expences in technological era

- expensive technology replaced knowledge and ordinary medical skills
- fear of possible mistakes imposed by overdemanding society
- defensive diagnostic process
- defensive therapy
- pressure to use inovative methods, that are expensive and often not proven effective

# Diagnostic failure

- Based on autopsy data- cancer,( breast, colon), pulmonary embolism, aortic dissection, pneumonia-

- Diseases atypically presented
- The symptoms are variable
- There is no routinely used screening test
- Necessary to take diagnosis into consideration
- Examples:
  - Polymyalgia rheumatica
  - Still's disease
  - Acute intermittent porphyria
  - Antiphospholipid syndrom

# History

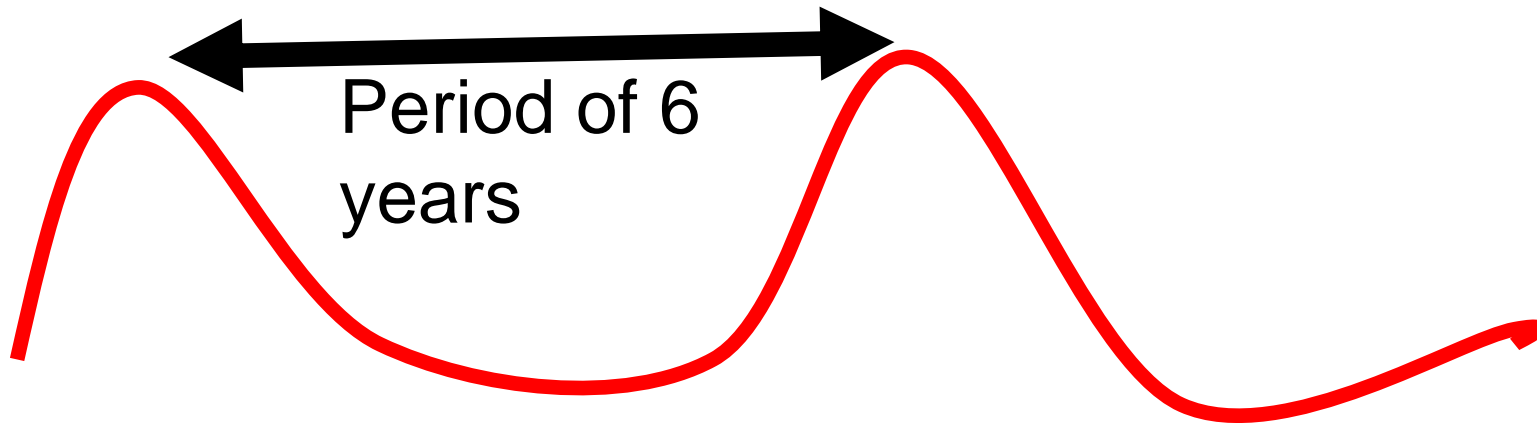
- **1890 – Hutchinson –clinique of temporal arteritis**
- **1898 - Bruce senil revmatic gout**
- **1930 – Horton – histology of TA**
- **1957 – Barber - polymyalgia rheumatica**
- **1960 - Hughes – relationship TA + PMR**
- **1984 – Healey clasification criteria PMR**

# Epidemiology

- ***Salvarani – Minesota 1971-1991, PLM- prevalence –***
- ***1/ 133 subjects over 50***
- ***maximum of incidence 70-80 years,***
- ***Incidence higher to the north***
  
- ***Salvarani –Minesota 1995 - prevalence TA***
- ***-1 / 5000 subjects over 50, higher frequency found in autopsy***
  
- ***sometimes seen in clusters***

# Etiology a pathogenesis?

- Cytokins – interleukin 6,interferon gama
- HLA bound



Parvovirus B 19 DNA  
Cluster occurrence

# Manifestation of PMR

- Symetric pain and morning stiffness – shoulders, neck, hip
- Distal involvement possible
- Fatigue
- Unintentional weight loss
- Weakness or a general feeling of being unwell
- Sometimes, a slight fever or FUO

## Involvement of structures in PMR

- ***synovitis –more involved extraarticular structures***
- ***subdeltoid and subacromial bursitis (NMR, US)***
- ***?? % of cases nonerosiv synovitis of knees and wrists, carp. tunel syndrom and swelling of hands and feets***

# 7 CORE CRITERIA 2008

- age  $\geq$  50 years,
- duration of symptoms  $\geq$  2 weeks
- bilateral shoulder and/or pelvic girdle aching
- duration of morning stiffness  $>$  45 min
- elevated erythrocyte sedimentation rate
- elevated C-reactive protein
- rapid steroid response ( $>$  75% global response within 1 wk to prednisolone/prednisone 15 -20 mg daily)

# Symptoms of Giant cell arteritis

- *Headache, dry cough, claudication of the jaw, swallowing pain, hoarseness*
- *20% visus problems (40% amaurosis fugax as warning sign)*
- *Symptoms from any branches of arterial tree*
- *15% as FUO*
- *Mono a polyneuropatic symptoms on extremities*
- *Claudication syndrom*
- *Aneurysm of desc. aorta 17 times more frequent as a late complication, rupture is possible*
- *Pericarditis*

**Diagnostic criteria of temporal arteritis  
( Am.Col.of Rev. 1990)  
at least 3 criteria**

- **Age > 50 let**
- **New localised head pain**
- **Tenderness or depressed pulses on temporal arteries**
- **SR > 50/ hod.**
- **Histology of temporal arteries**

# Lab results

- 10-20% has normal SR
- 1/3 has abnormal liver enzymes
- CRP more sensitive for diagnosis and follow up than SR
- Interleukin 6 - the best marker, not always available
- Anemia of chronic illness

# Asymptomatic Giant Cell Arteritis in PMR

- 35 patients with isolated symptoms of PMR
- 11- positiv (31%) positron emission tomography in arteries- mostly subclavian
- intensity of FDG uptake in the large vessels did not correlate with the risk of relapse.

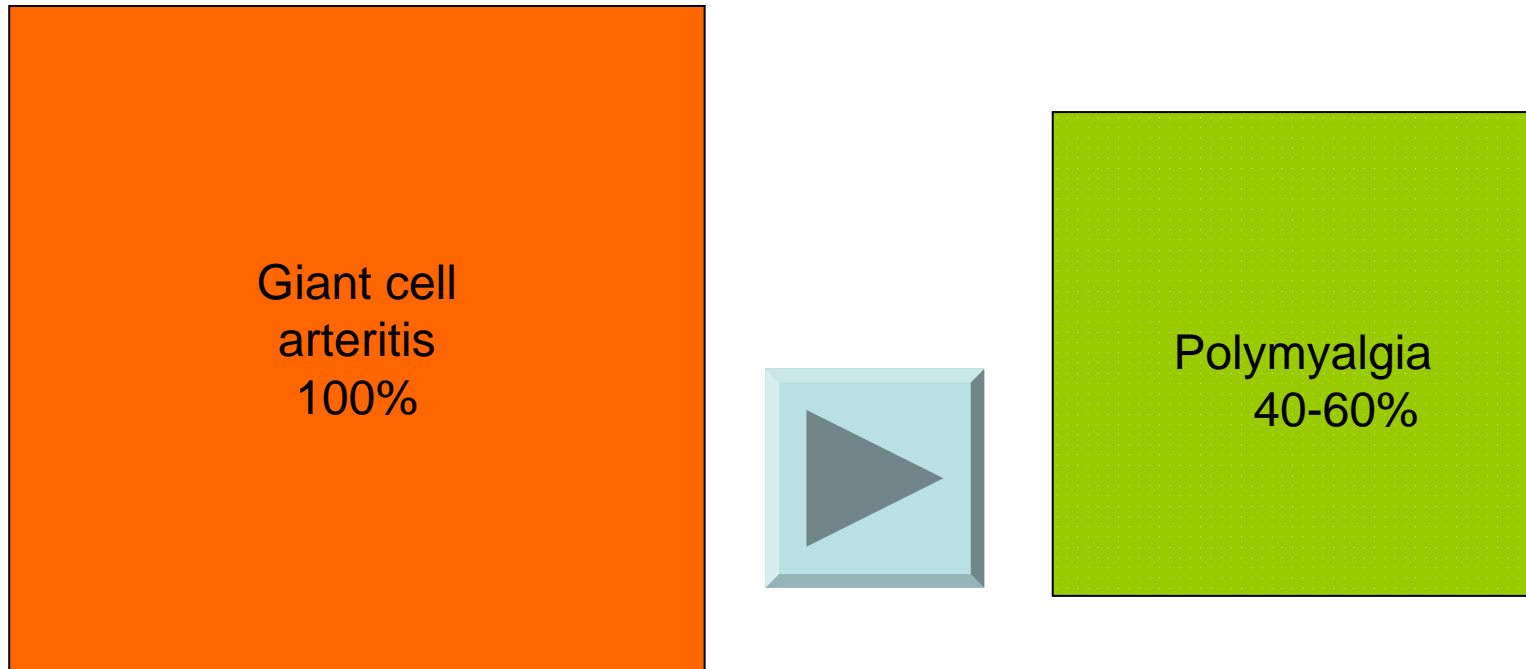
# Relationship of PMR and GCA

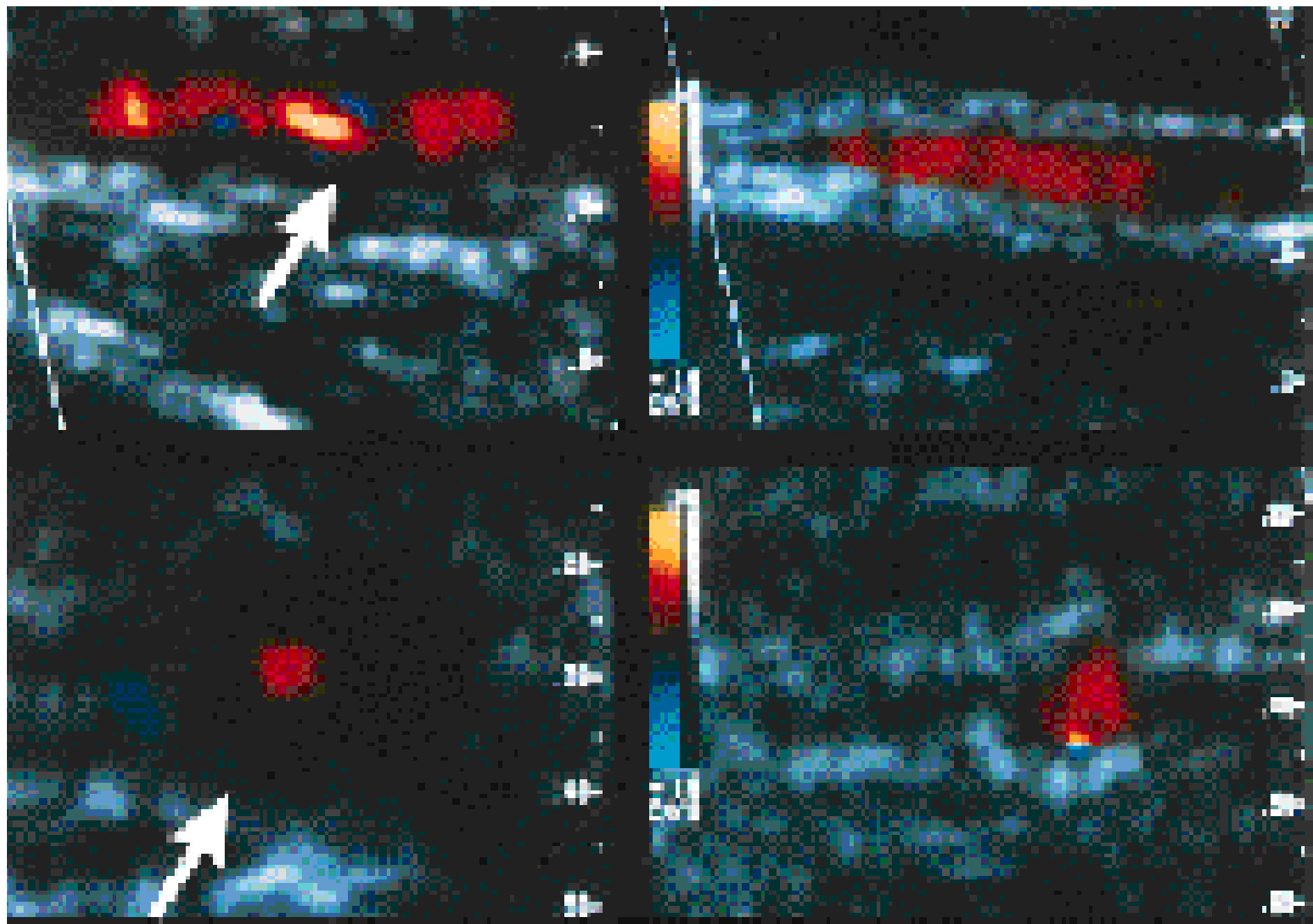
Polymyalgia  
revmatica

+

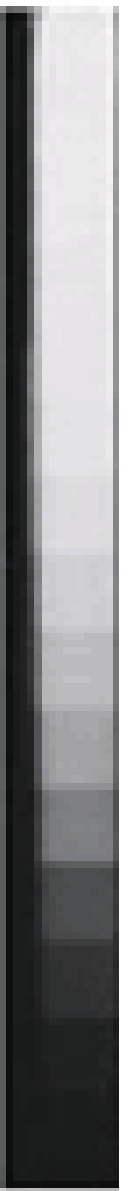
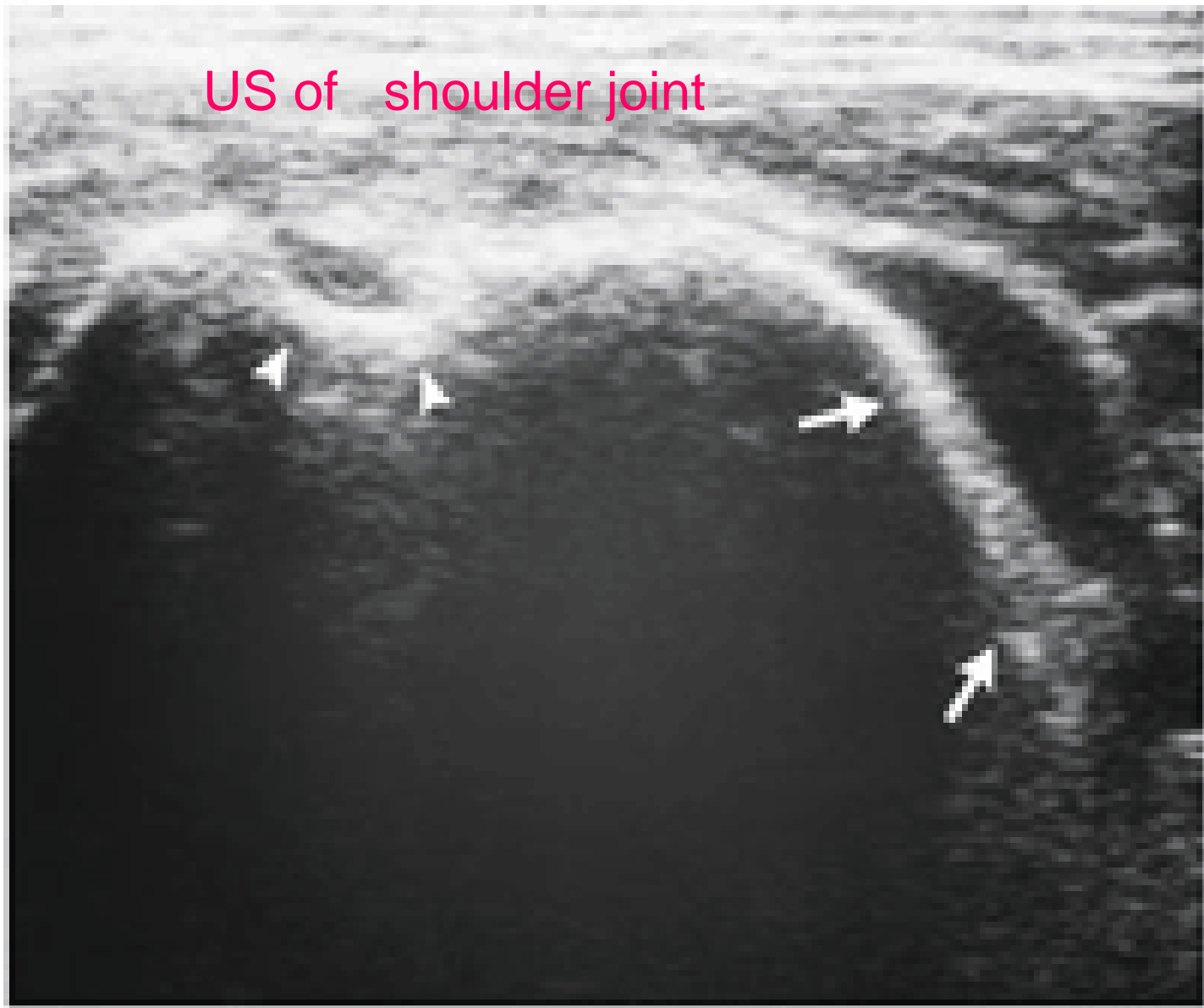
Giant Cell  
arteritis  
Temp  
Art.

# When manifests as Giant Cell arteritis

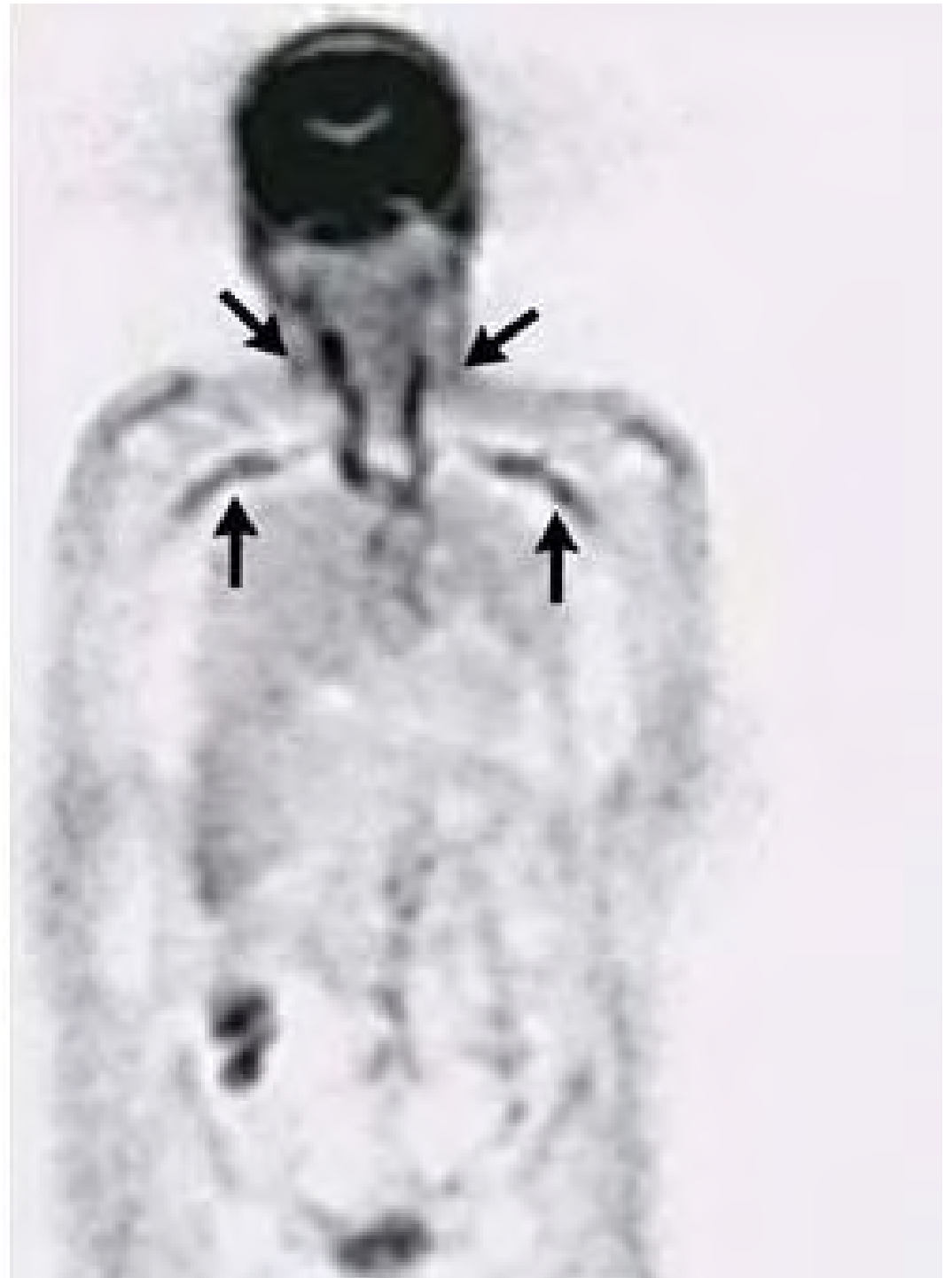




US of shoulder joint



A



# Diferencial diagnosis

- Fibromyalgia syndrom
- Rheumatoid arthritis
- Takayashu arteritis
- Osteomalacia or simple deficit of vitamin D
- Depression
- Asymetric polyarthritits as paraneoplastic syndrom
- PMR paraneoplastic sign ???

# Therapy

- **Starting dose 20 mg Prednison for PMR  
60 mg for AT with vision defects**
- **Corticosteroid therapy usually at least 2 years**
- **Risk factor for longer therapy:  
increasing age at diagnosis, female sex, higher baseline erythrocyte sedimentation rate, and lower daily CS dose**
- **Mean daily maintenance dose of prednisolone during the first and second year:  
5.7 mg and 4.3 mg for PMR  
6.6 mg and 4.1 mg for TA  
8.3 mg and 4.7 mg for PMR with TA**
- **biological ( infliximab), immunosupresive (metotrexat) therapy- no benefit**

[Narváez J](#), J Rheumatol. 1999 Sep;26(9):1945-52.

[Myklebust G](#) Scand J Rheumatol. 2001;30(5):260-7.

Last 8 years in internal  
departement  
23 patients

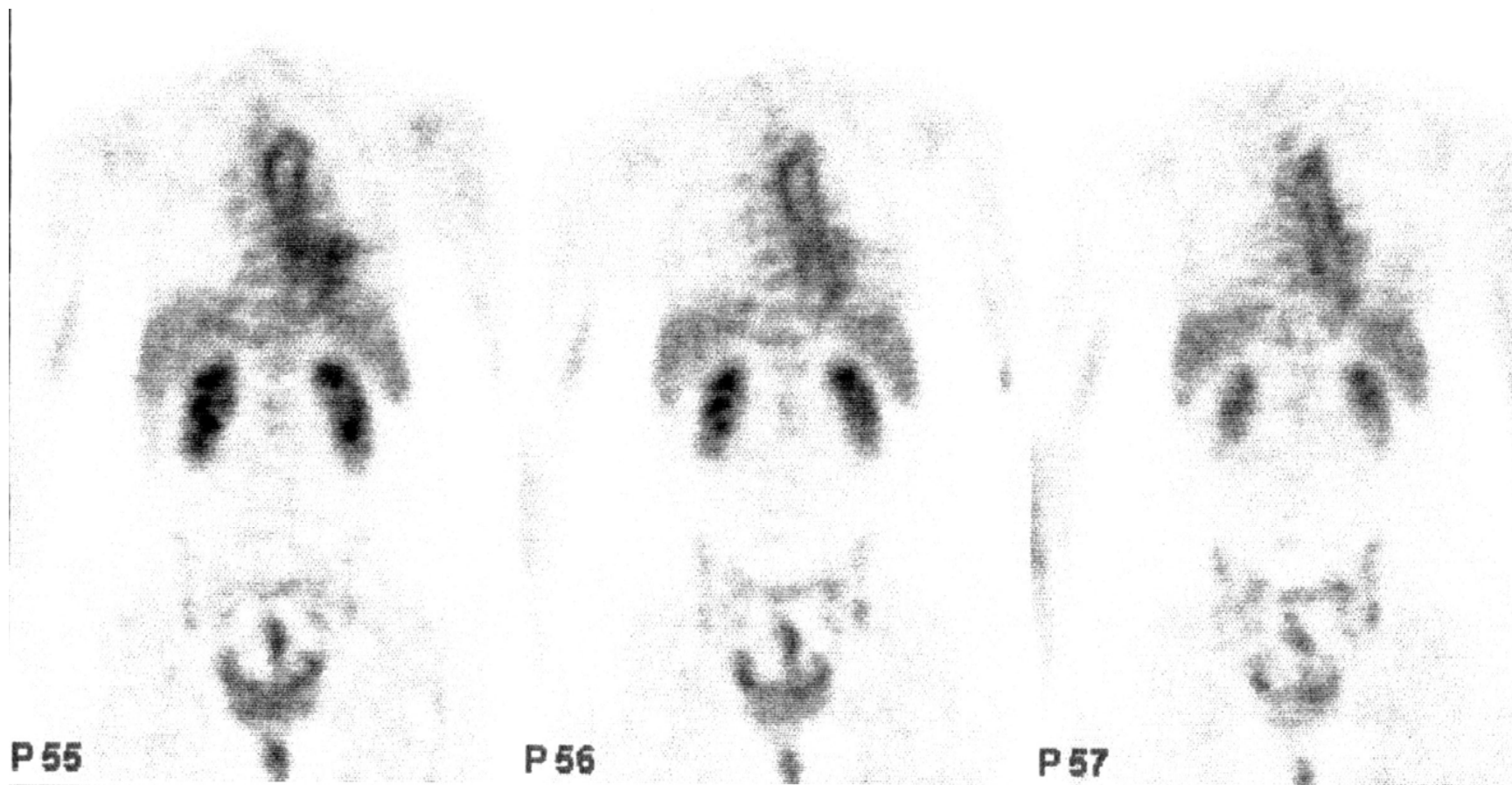
- ***19 women + 4 men***
- ***9 patients with clasical manifestation***
- ***14 patients with atypical manifestation***

# Leading signs of PMR and GCA with atypical presentation

N = 14

- ***Fewer of unknown origin– 6***
- ***Fatigue, anemia, 4***
- ***Irritable cough, 2***
- ***Abdominal pain, anemia, 1***
- ***Pericardial effusion, + FUO 1***

# Žena 58 let – PET scan



# What to remember

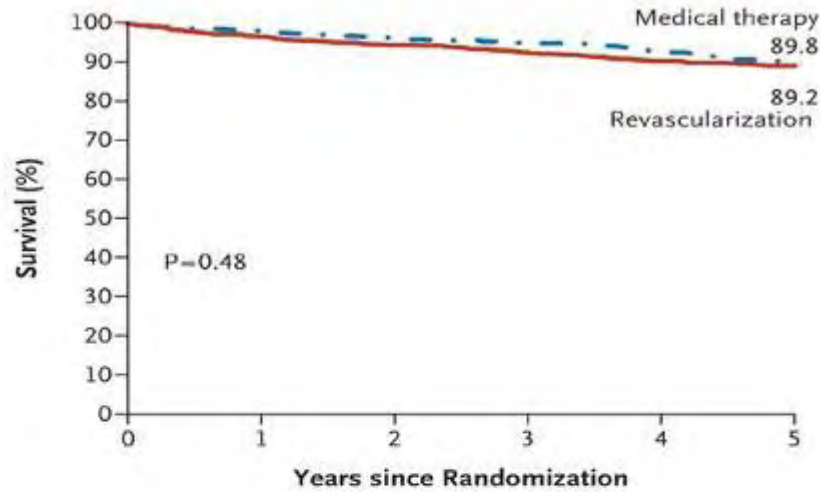
- Consider PML or/and GCA in patients with elevated SR a CRP and symptoms, that were not explained by first logical diagnostic steps.
- Try ultrasound on TA or shoulder joints
- Try PET with fluorodeoxyglucose
- Try Prednison test when you are still in doubt

What is the difference between these two ?



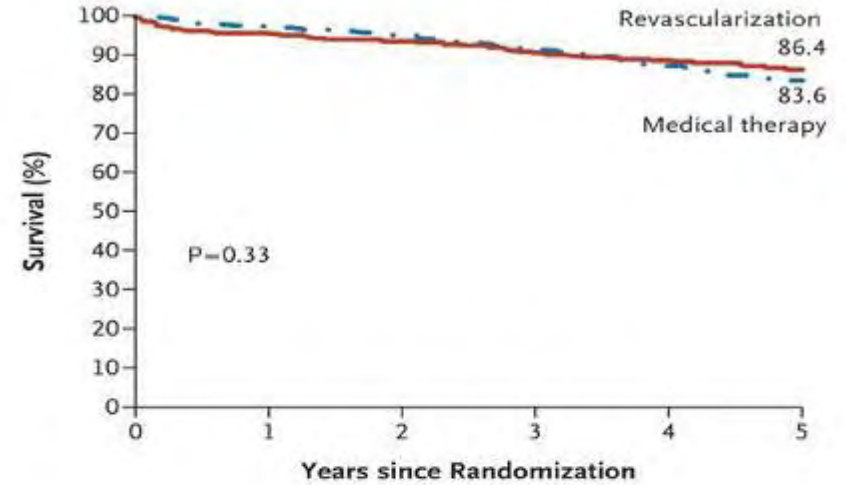


**A Survival in PCI Stratum**



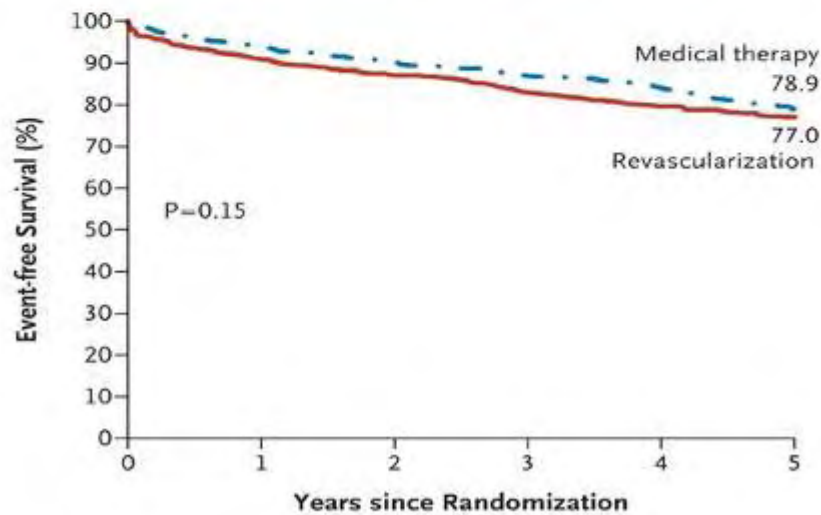
No. at Risk 1605 1562 1529 1505 1306 863

**B Survival in CABG Stratum**

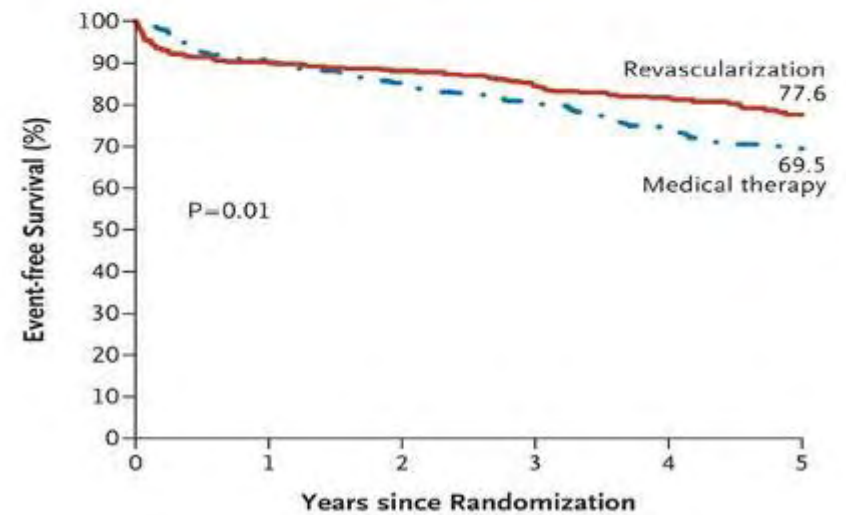


No. at Risk 763 734 718 692 586 333

**C Freedom from Major Cardiovascular Events in PCI Stratum**



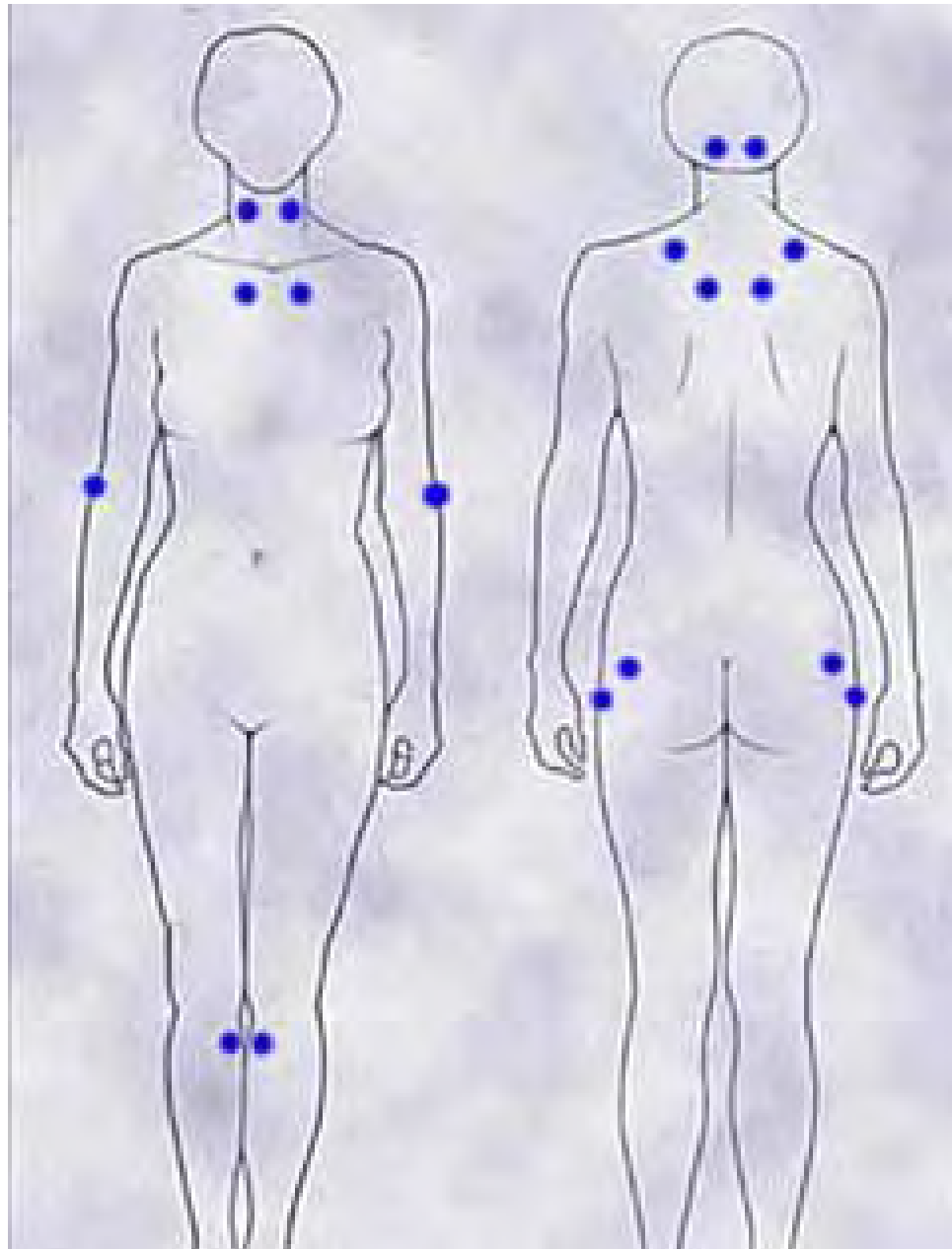
**D Freedom from Major Cardiovascular Events in CABG Stratum**





# Diferential diagnosis with fibromyalgia

- The pain is described as a constant dull ache, typically arising from muscles.
- additional pain when firm pressure is applied to specific areas of body, called tender points
- awaken tired, even though they seem to get plenty of sleep.
- include headaches, sensitivity to light, dizziness, memory problems, and numbness and tingling in arms and legs
- irritable bowel syndrome, bladder control problems and mood disorders, such as depression and anxiety.
- Duloxetine (Cymbalta), Gabapentin (Neurontin)



# Diagnostic criteria of PMR, Healey 1984

All must be present

- ***age > 50 let, SR > 40/ hour***
- ***Pain >1 month and morning stiffness >1 hour either in neck or shoulders or hips***
- ***Quick relief after prednison maxim . dose 20 mg a day***
- ***All other diagnoses with similar clinical picture ruled out***

# Mechanism of misdiagnosis

**Anchoring** refers to the tendency to seize on the first impression

**Availability** refers to the tendency to assume that an easily remembered prior experience explains the new situation and vice versa

**Attribution** refers to the tendency to invoke stereotypes in our minds and “attribute” symptoms and findings to the stereotype

# Méně obvyklé příznaky

- ***Vysoké teploty***
- ***Klaudikace horních končetin, žvýkacího svalstva, chrapot***
- ***Mozkové cévní příhody***
- ***Perikarditis***
- ***17 x vyšší pravděpodobnost aneurysmatu hrudní nebo břišní aorty***
- ***Postižení periferních kloubů jako u RA***

**Celkem za dobu 15 ti let na  
int.odd. zachyceno s polymyalgií  
nebo velkobuněčnou arteritidou  
15 pacientů**

- 13 žen + 2 muži**
- 10 pacientů s klasickými příznaky**
- 5 pacientek s atypickými příznaky**
- 3 pacienti s relapsy**
- Ve spádové oblasti by mělo být 10 x více**

# Vedoucí příznaky pacientů přijatých na interní oddělení.

N = 5

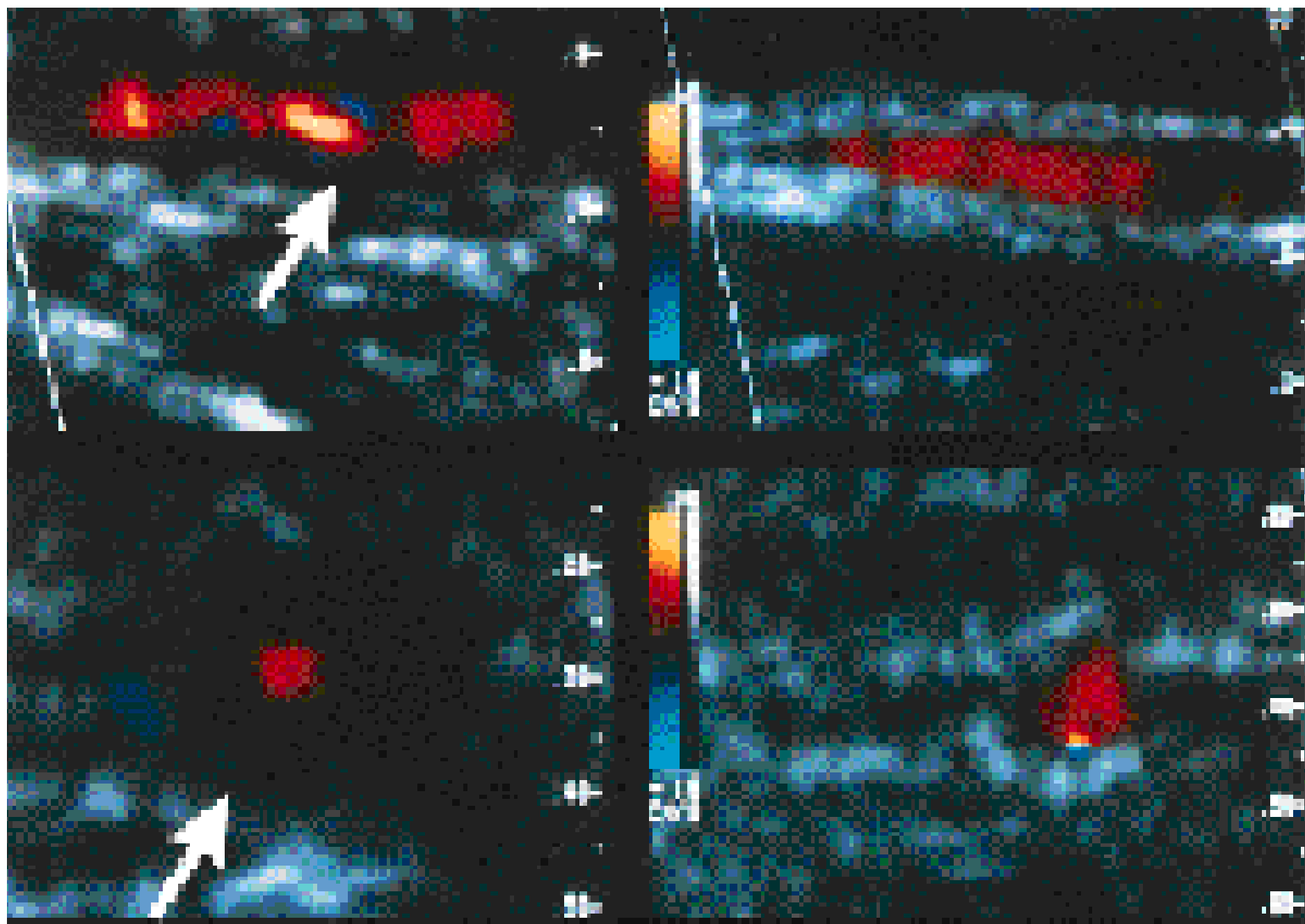
- ***Teploty nejasného původu – 2***
- ***Perikardiální výpotek nejasné etiologie***
- ***Dráždivý kašel, únava, anemie, vysoké CRP***
- ***Bolesti břicha, anemie, vysoká FW***

# Méně obvyklé příznaky

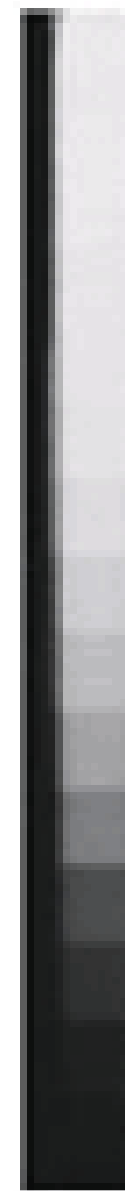
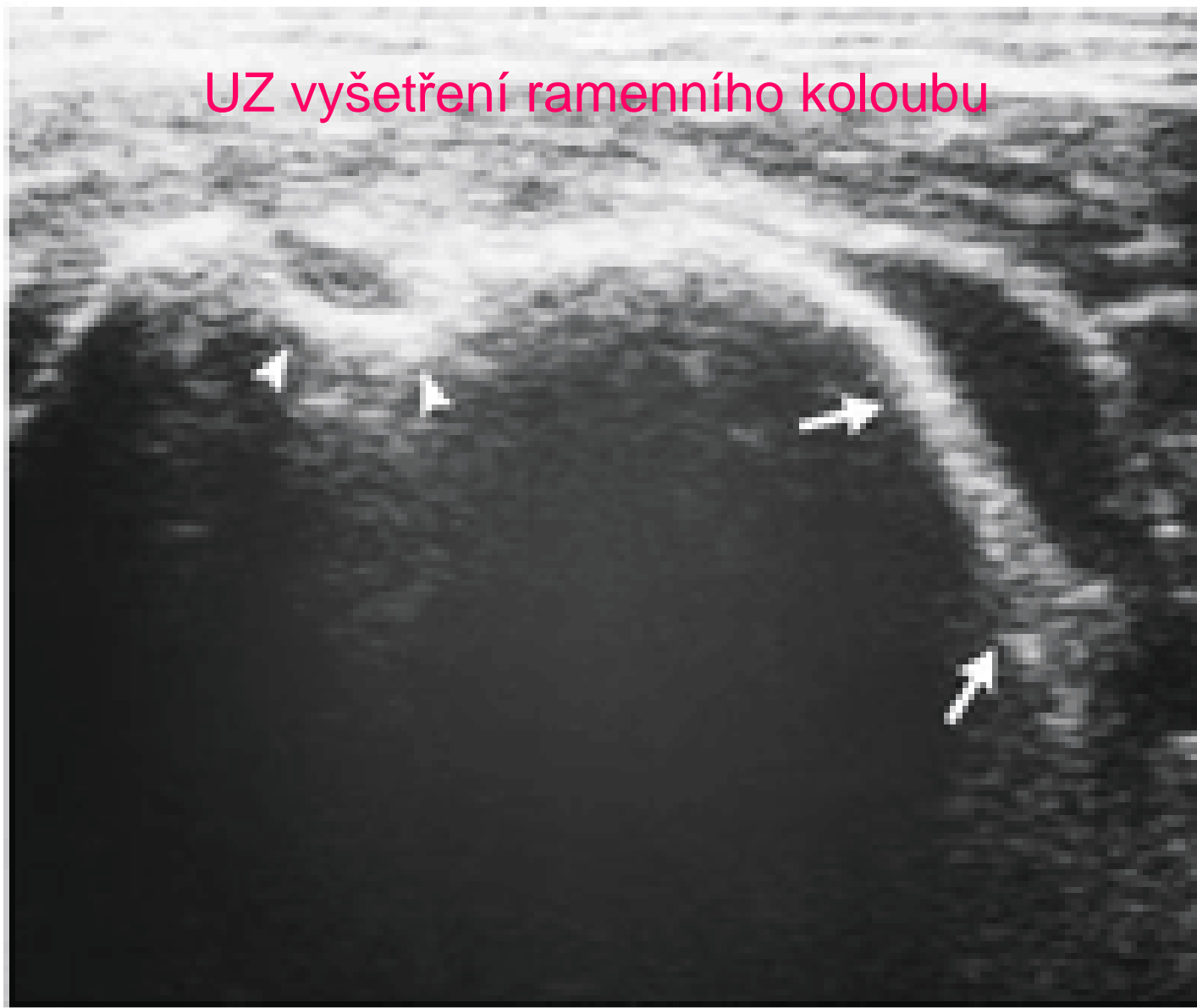
- ***Vysoké teploty***
- ***Klaudikace horních končetin, žvýkacího svalstva, chrapot***
- ***Mozkové cévní příhody***
- ***Perikarditis***
- ***17 x vyšší pravděpodobnost aneurysmatu hrudní nebo břišní aorty***
- ***Postižení periferních kloubů jako u RA***

# Laboratorní vyšetření

- 10-20%emocných s PLM i teporální arteritidou mají normální FW
- Třetina má abnormální jaterní testy
- C reaktivn protein citlivější než FW při dg. i relapsu
- Interleukin 6 - nejlepší, zřídka k dispozici



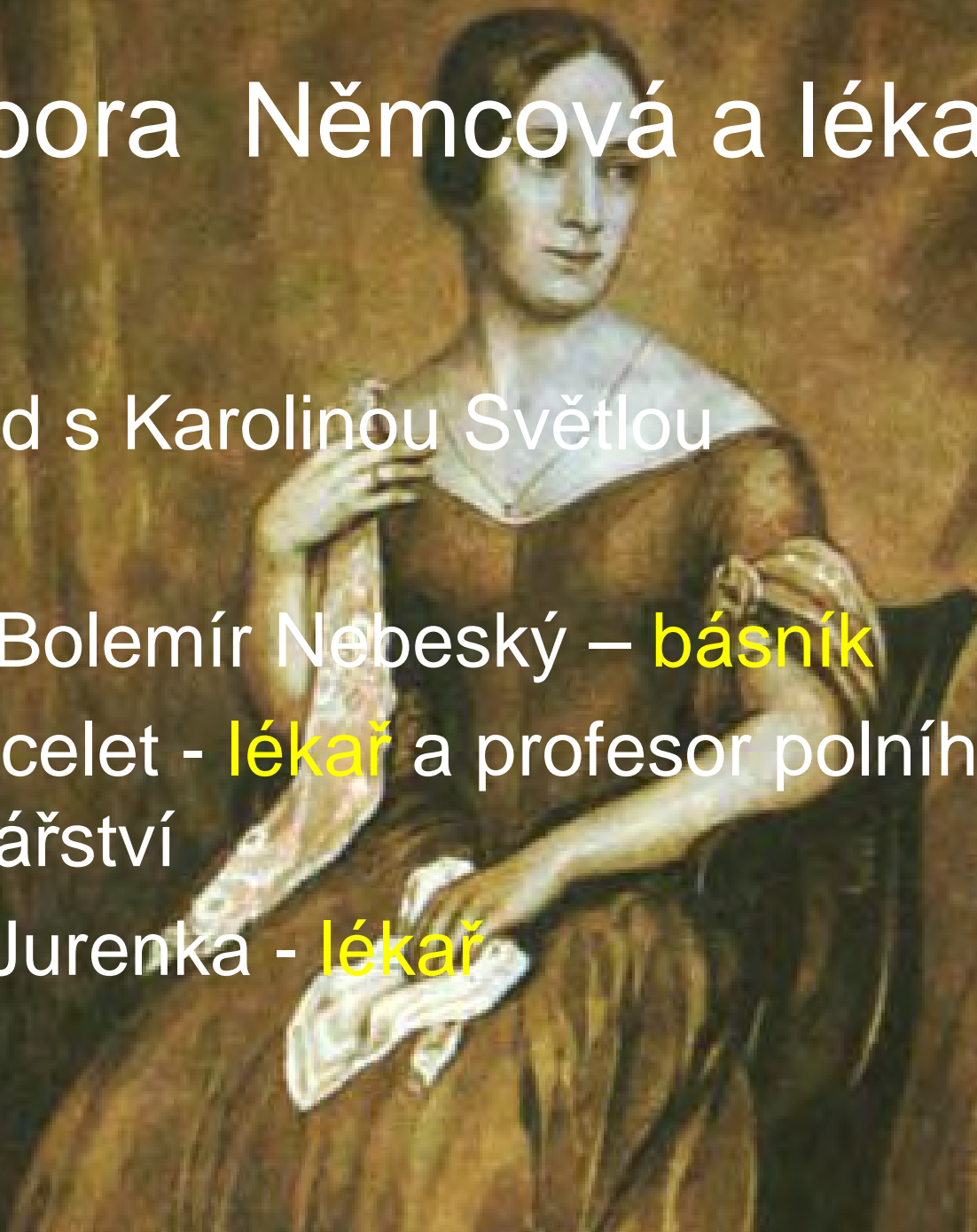
# UZ vyšetření ramenního koloubu



A

# Barbora Němcová a lékaři

- Rozchod s Karolinou Světlou
- Václav Bolemír Nebeský – básník
- Jan Helcelet - lékař a profesor polního hospodářství
- Hanuš Jurenka - lékař





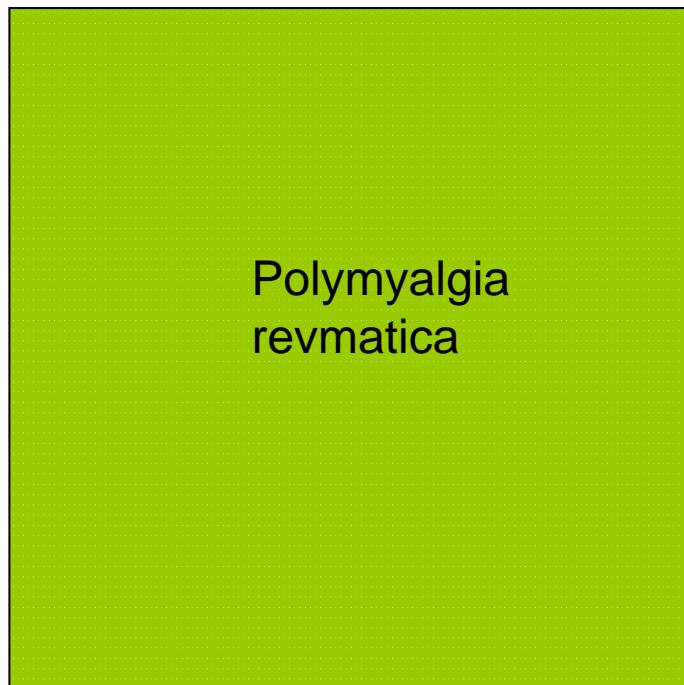
# PLM stejná etiologická záhada jako původ Boženy Němcové ?



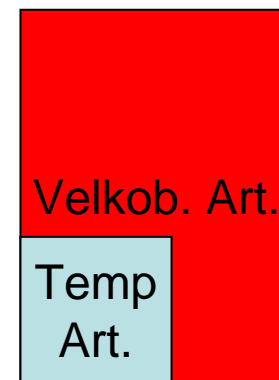
- V.Zaháňská (nebo její schovanka) – Metternich, car Alexander I.
- sestra Dorothea Talleyrandová – Clam Martinic
- Francisco de Goya – Francisca Garcia

# Jak je to časté onemocnění

- Populace nad 50 roků
- 58 / 100 000 / 1 rok



+

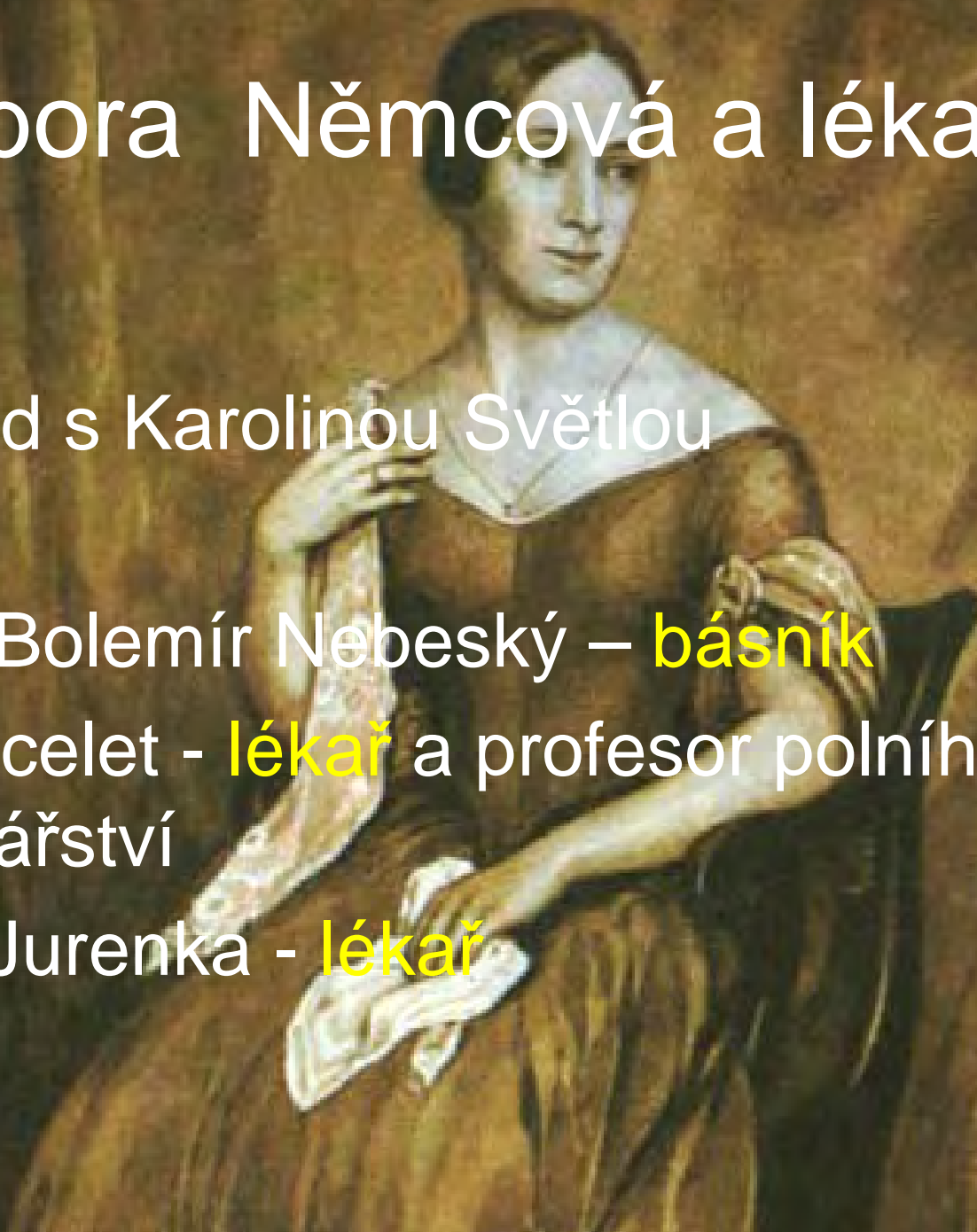


# Vzájemné vztahy obou projevů



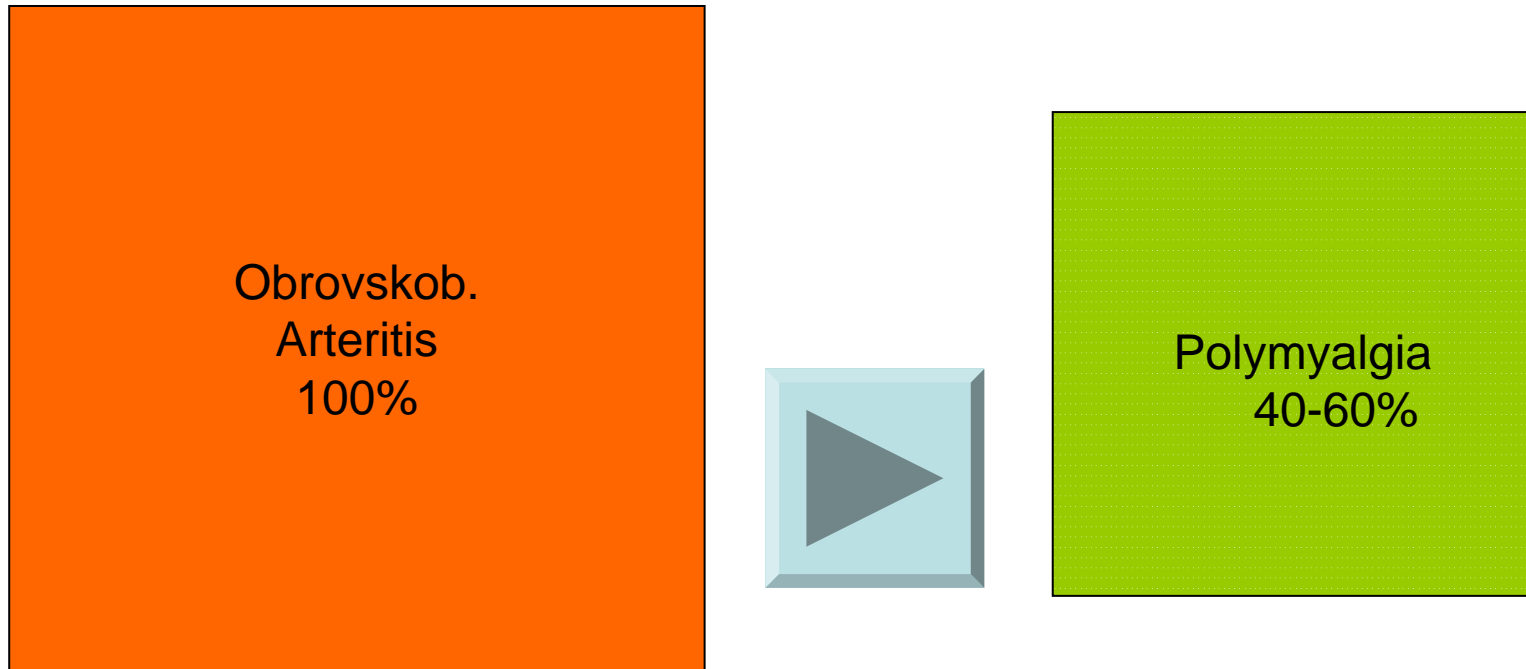
# Barbora Němcová a lékaři

- Rozchod s Karolinou Světlou
- Václav Bolemír Nebeský – básník
- Jan Helcelet - lékař a profesor polního hospodářství
- Hanuš Jurenka - lékař






# Vzájemné vztahy obou projevů



# When only manifestation of PMR



Polymyalgia  
rheumatica  
100%

Giant cell  
arteritis  
20%  
100%??

# Developing classification criteria for polymyalgia rheumatica 2008.

- **1 step:** 27 international experts who anonymously rated the importance of 68 potential criteria.
- **2. step:** meeting of the experts, who were provided with the results of the first round of ratings and were then asked to re-rate the criteria.
- **3.step:** 43 criteria that received > 50% support at round 2 was evaluated using an extended mailed survey of 111 rheumatologists and 53 nonrheumatologists in the United States, Canada, and Northern and Western Europe.
- **over 70% of survey respondents agreed on the importance of 7 core criteria**

[J Rheumatol. 2008; 35\(2\):270-7](#)

Dasgupta B; Salvarani C;