The challenging diagnosis of SAPHO Syndrome: a report of five cases and review of literature

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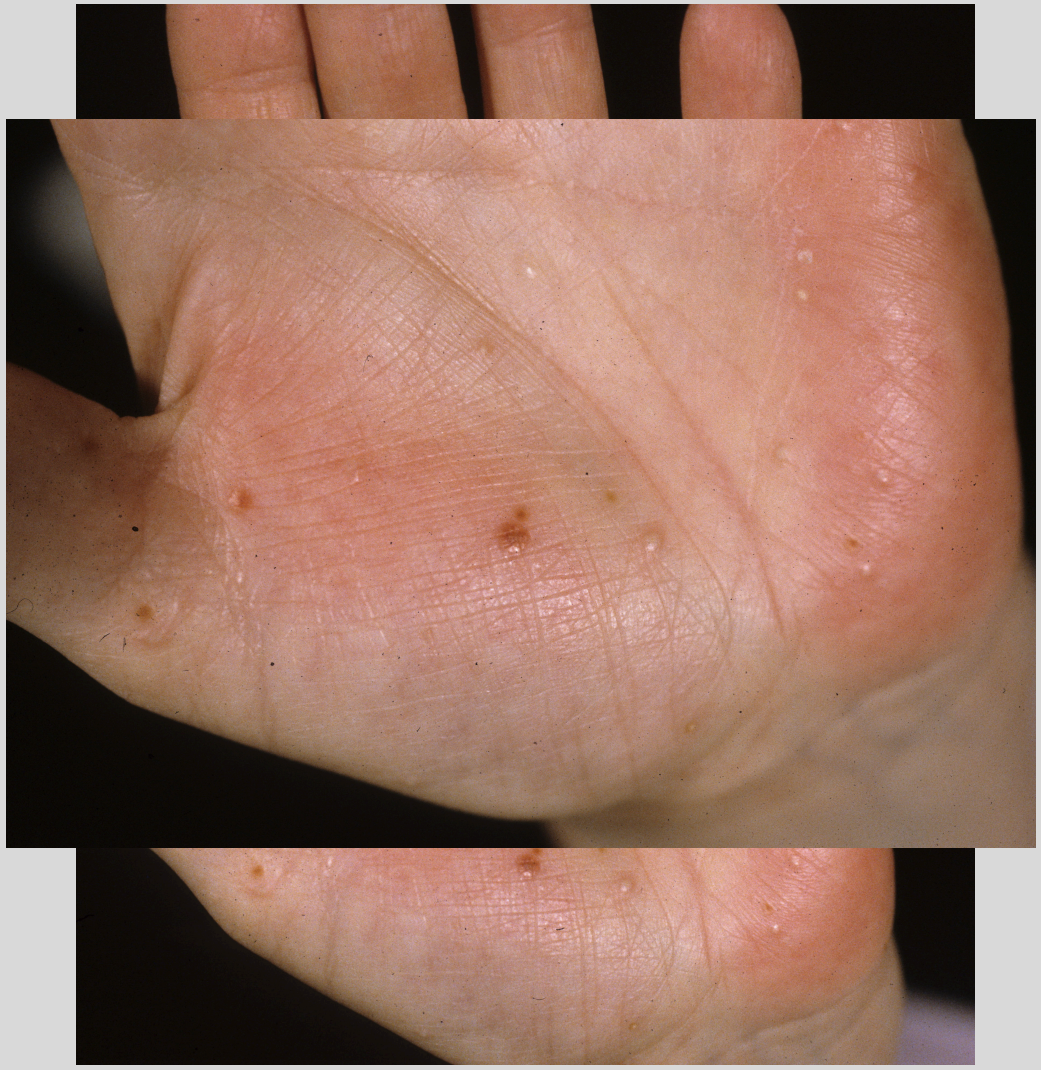
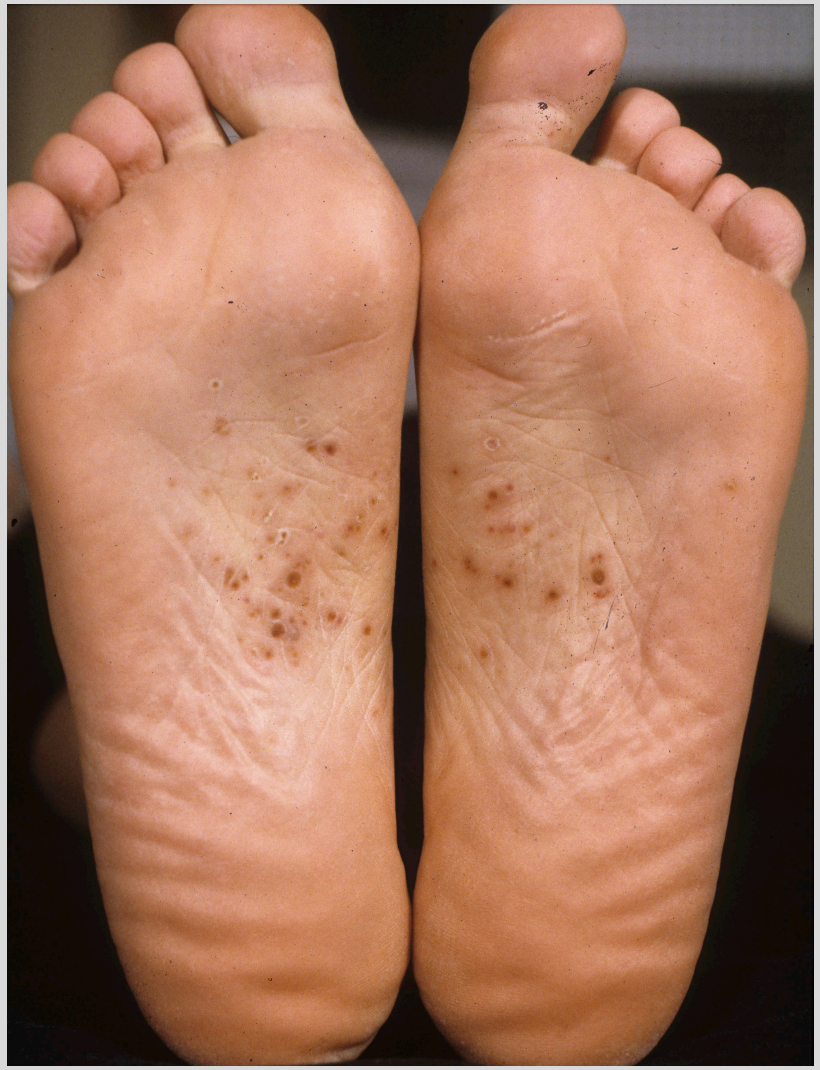
Riccardo CAVICCHIOLI

CLINICAL PRESENTATION

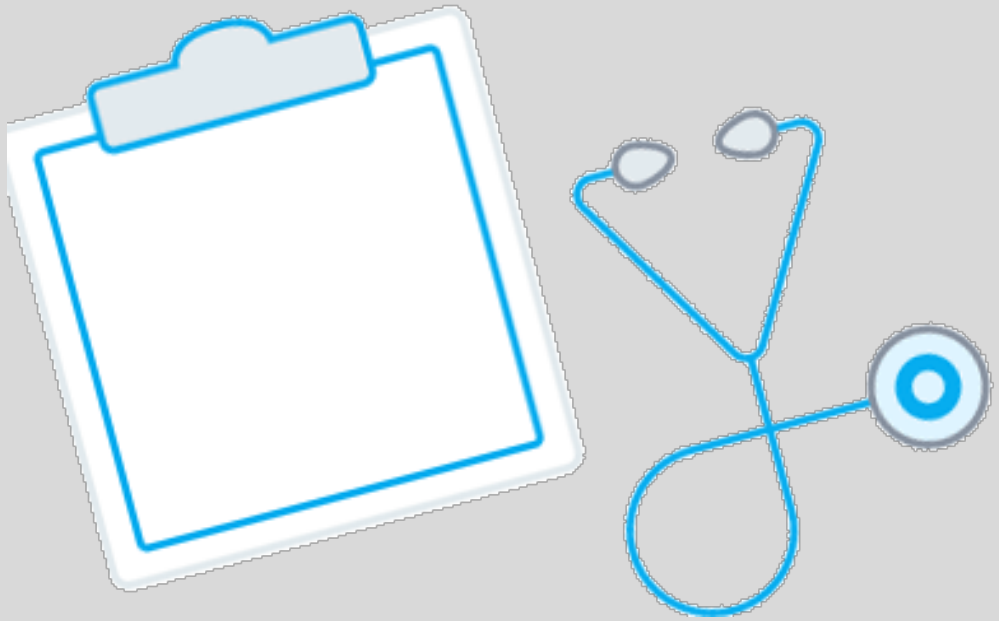




ANAMNESIS

~~Comorbidity~~

~~Drugs~~



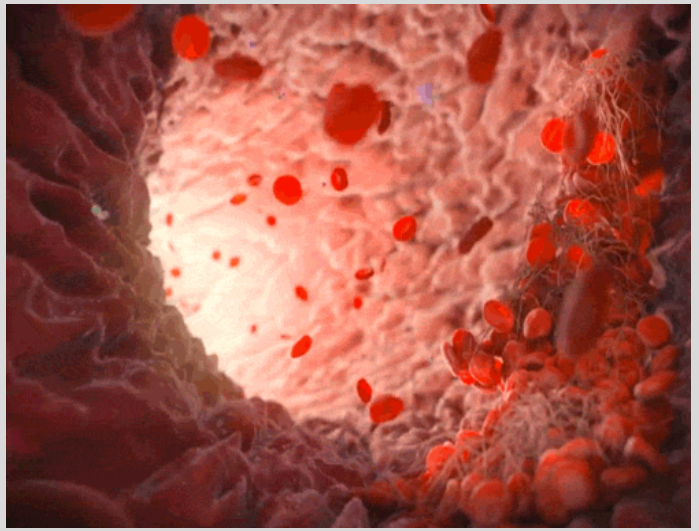
JOINT PAIN: sterno-clavicular junctions, knees

INVESTIGATION

HEMATO-CHEMICAL EXAMS

- Blood count + formula: in the standard, except:

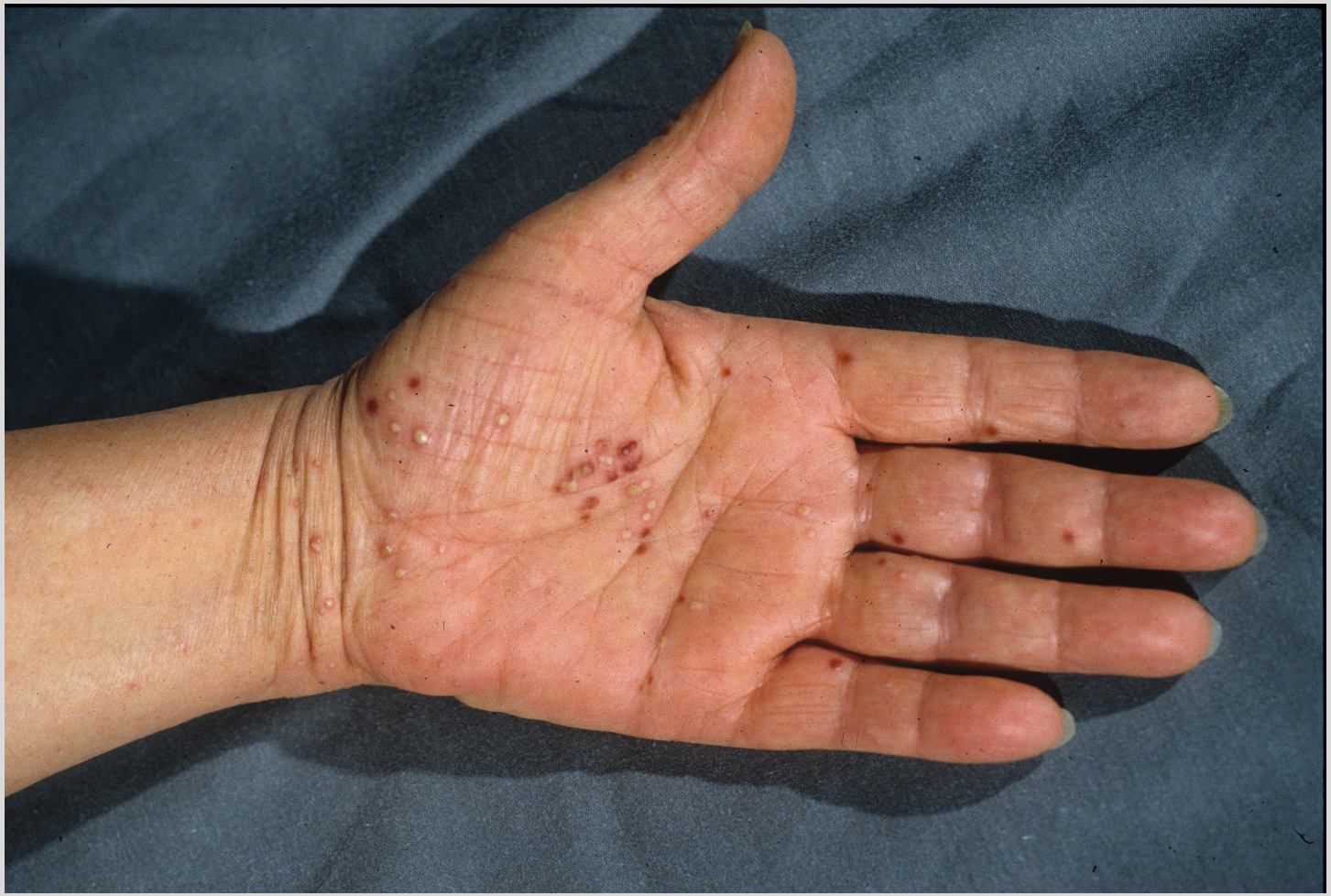
- **Neutrophil granulocytes**: 9.1 migl / ul
- **Hb**: 11.4 g / dl **V.E.S** .: 42 mm / h **PCR**: 14 mg/dl (Mosse)
- **ANA, anti-DNA, ENA, ANCA and anti-phospholipids**: negative
- **RF**: negative



- Liver and kidney function tests: normal

INVESTIGATION

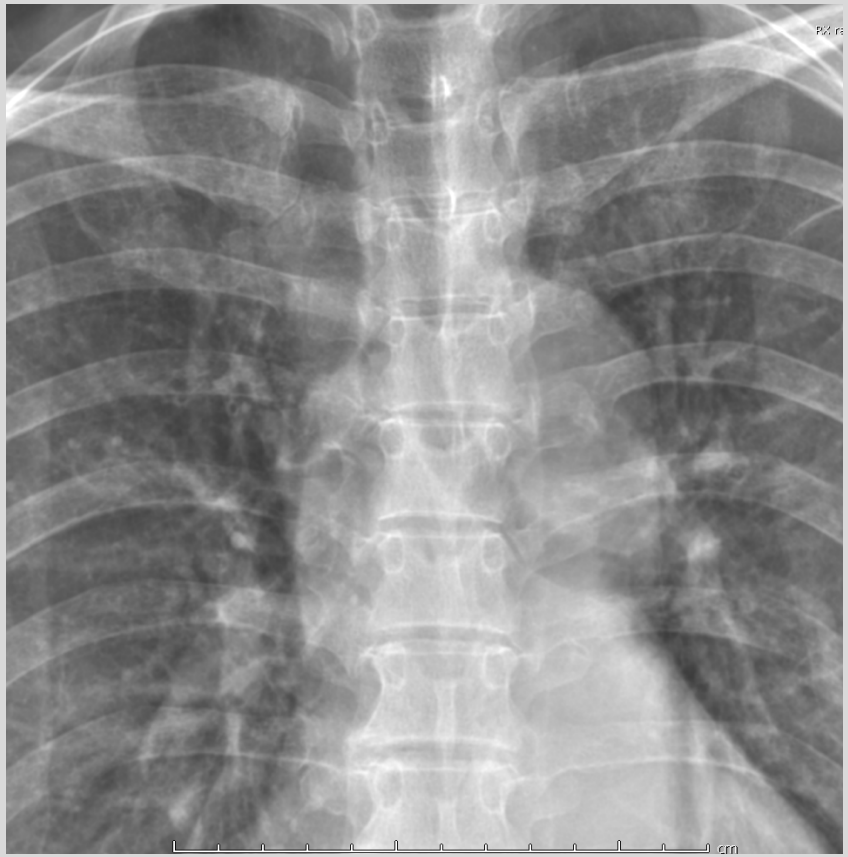
CUTANEOUS BIOPSY FOR ISTOLOGICAL EXAM

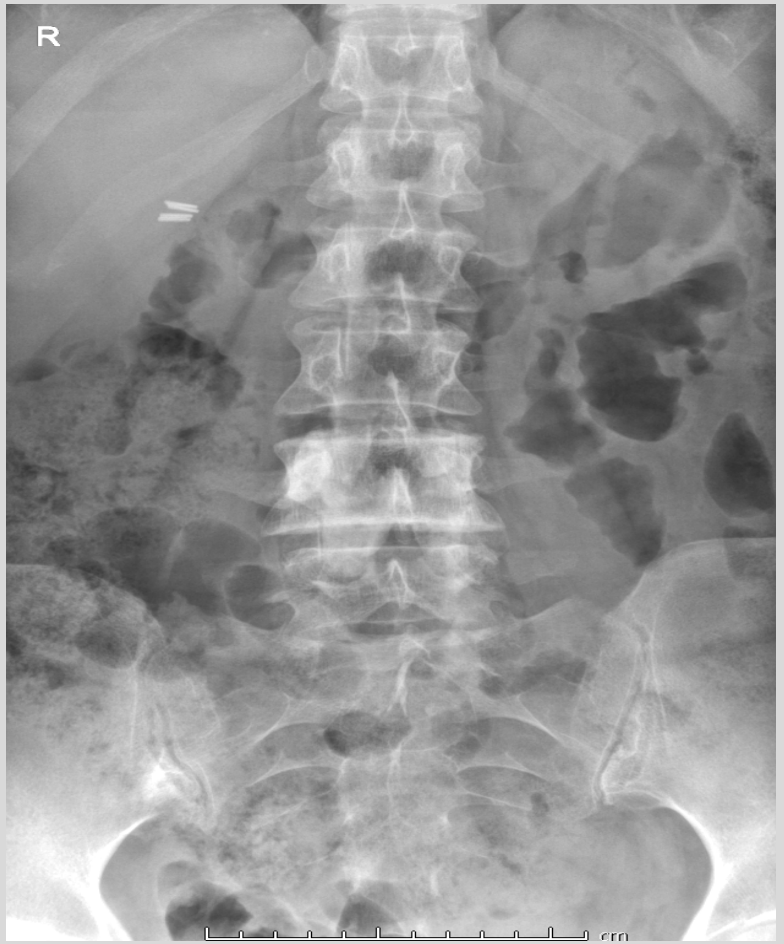


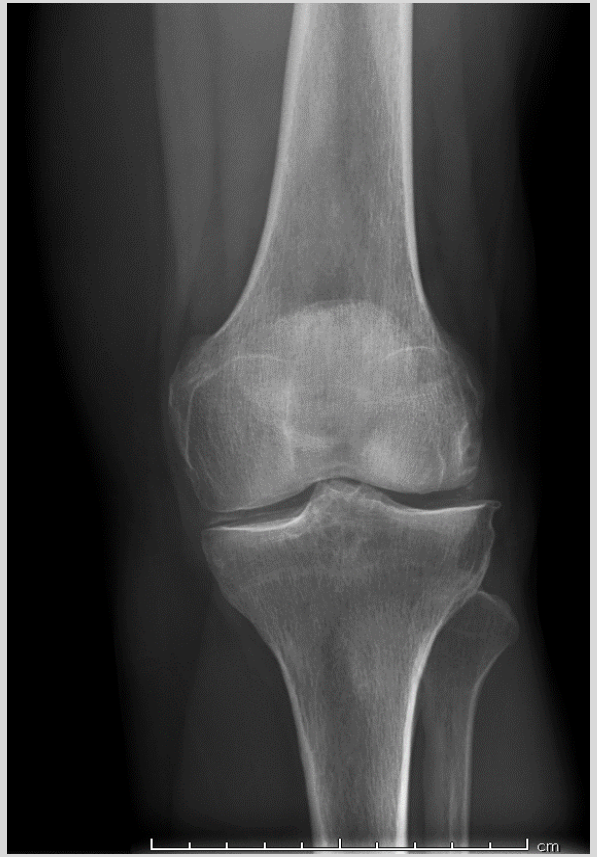
Infiltrato
Negatività

INVESTIGATION

RADIOLOGY



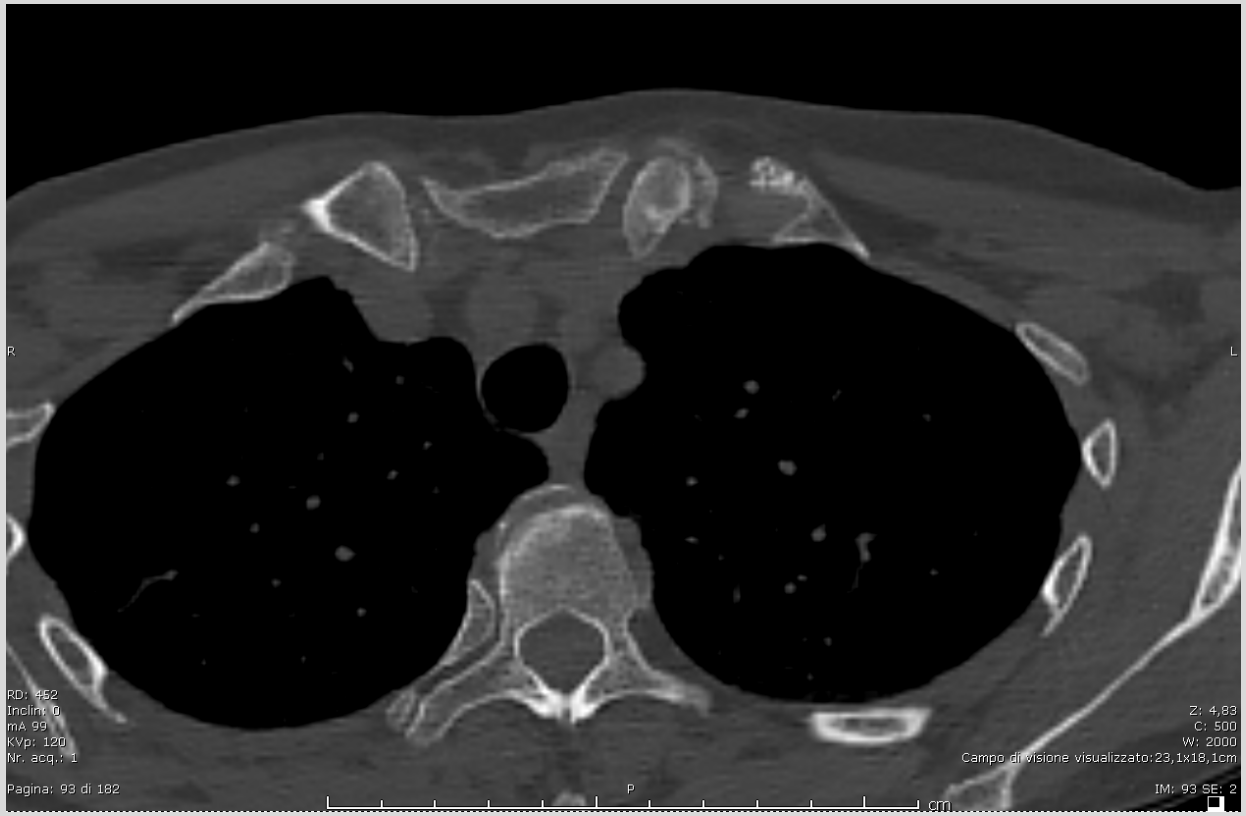


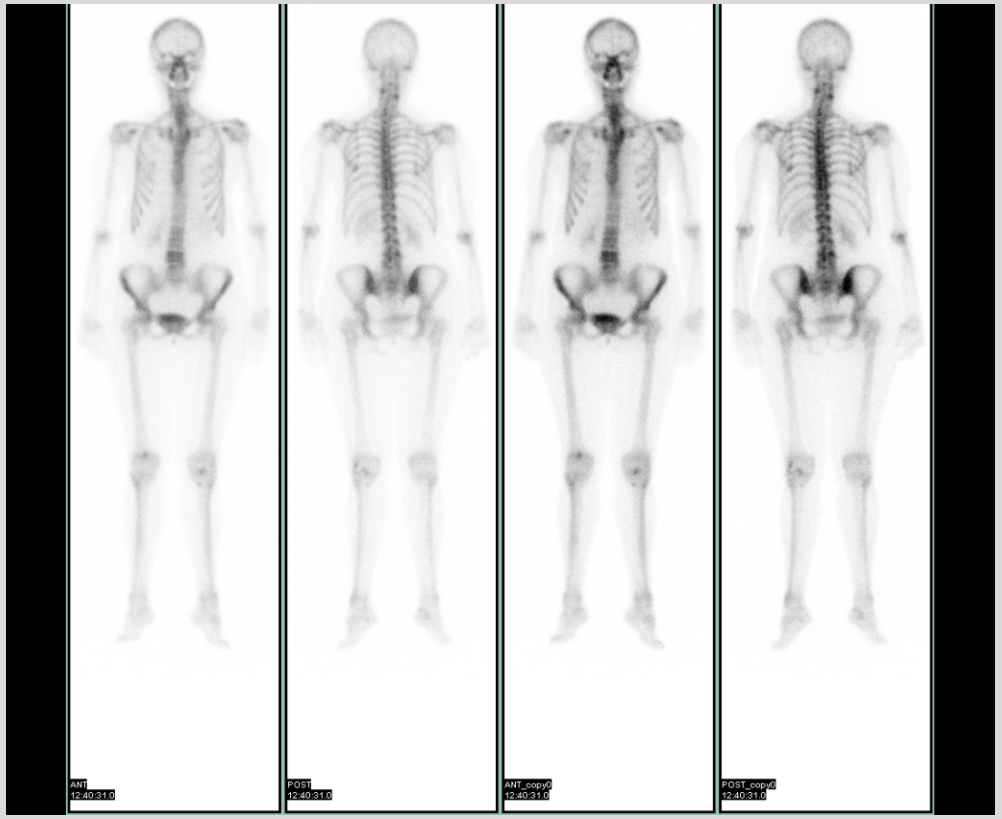


RX

INVESTIGATION

RADIOLOGY

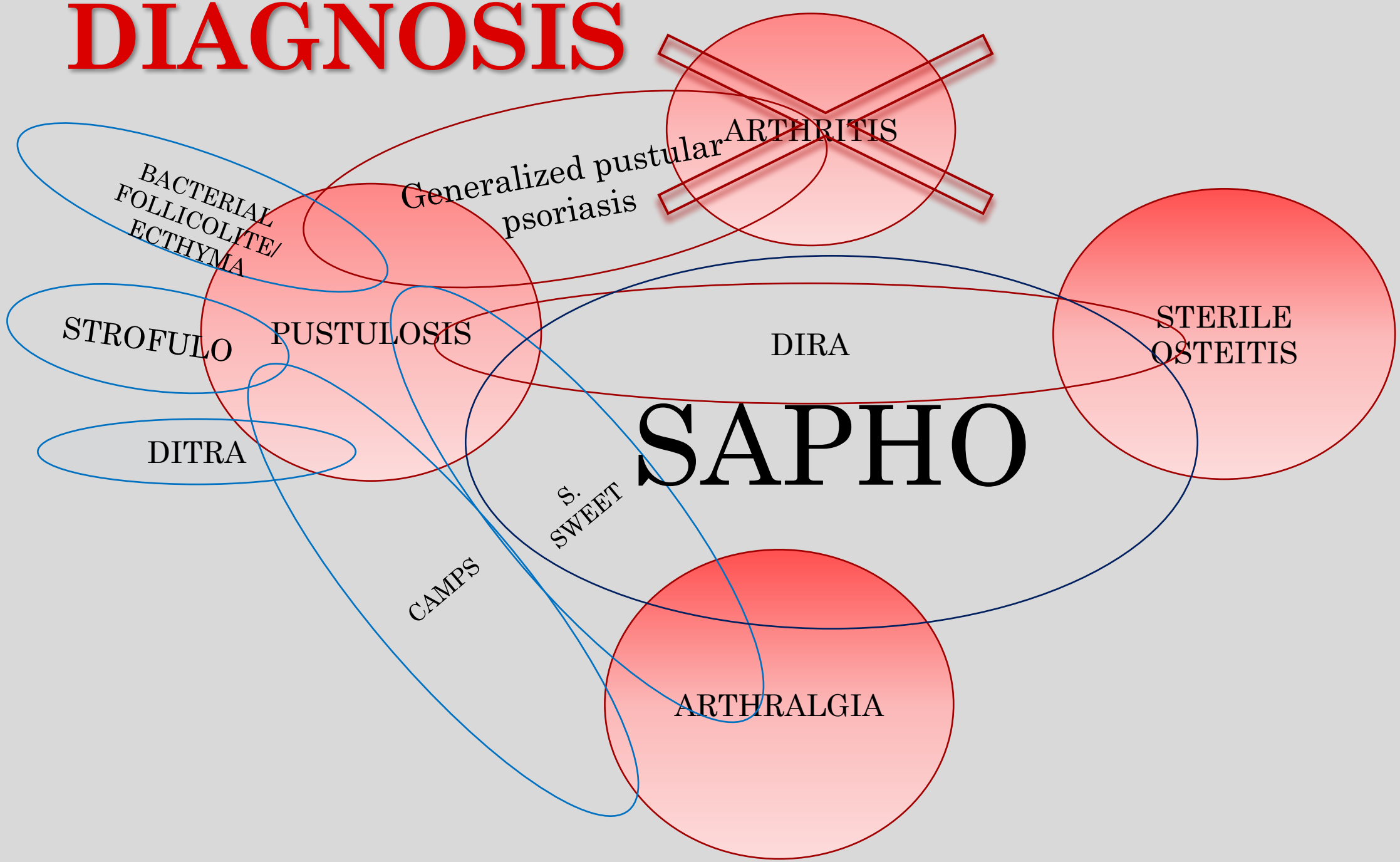




CT

SCINTIGRAPHY

DIAGNOSIS


ARTHRITIS
Generalized pustular psoriasis
BACTERIAL FOLLICOLITE/ ECTHYMA
STROFULO
PUSTULOSIS
DITRA
DIRA
STERILE OSTEITIS
SAPHO
S. SWEET
CAMPS
ARTHRALGIA

WHAT IS SAPHO SYNDROME ?

- **The acronym SAPHO stands for Synovitis, Acne, Pustulosis, Hyperostosis and Osteitis**
- **Estimated Prevalence 1:10000**
- **Etiology still unknown**

Burgemeister LT, Baeten DLP, Tas SW. Biologics for rare inflammatory diseases: tnf blockade in the SAPHO syndrome. The Netherlands Journal of Medicine 2012;70:444-449.

CLINICAL PRESENTATION

Clinical presentation is **HETEROGENEOUS**

Table 1 Frequency of Clinical Manifestations in SAPHO Patients

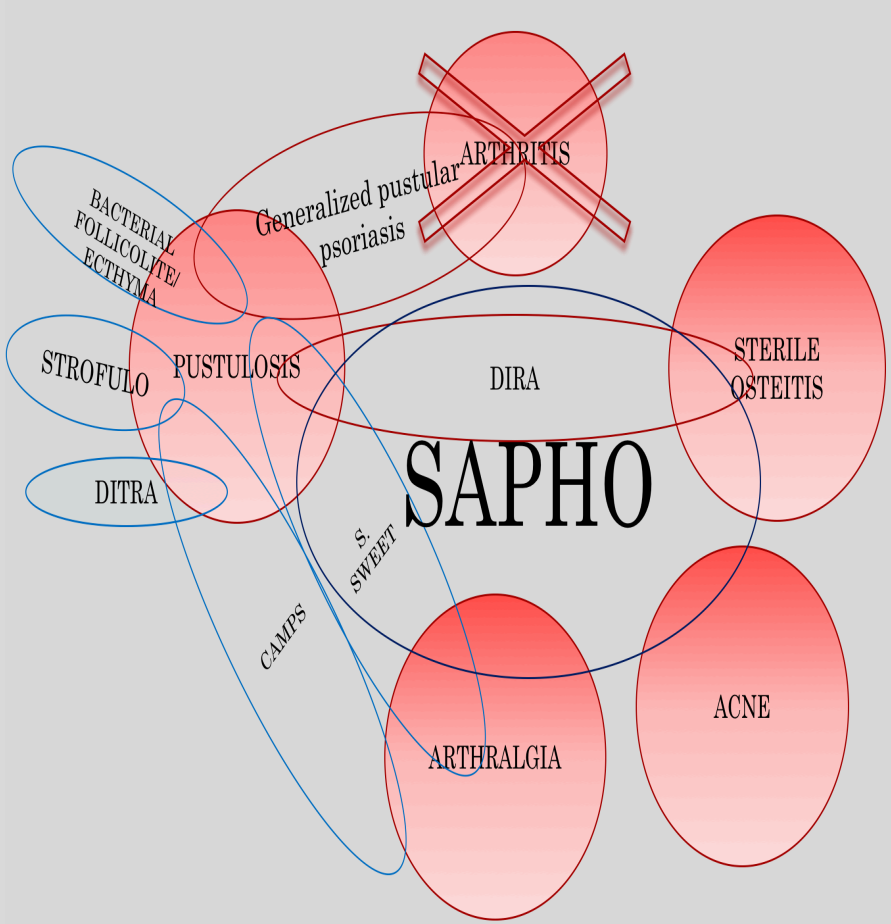
Reference	(18)	(24)	(19)
Country	France	France	Italy
n	120	52	71
Age at diagnosis (mean, range)	37.7 (5-67)	42 (15-73)	45.5 (35.7-54.0)[a]
Male/female	50/70	26/26	23/48
Years of study	1974-1997	1984-2007	1990-2008
Follow-up (yr)	Mean 4.9 (range 1-23)	NA	0
Anterior chest wall involvement (%)	63	73	70
Inflammatory back pain (%)	NA	13	24
Spinal (%)	33		34
Sacroiliitis (%)	40	27	
Long bones/peripheral (%)	5.8		
Mandible (%)	10.8		
Peripheral arthritis (%)	33	33	
Cutaneous manifestations (*N*/%)	110 (84)	63	40 (56)
Of those (in %)			
PPP overall	>58[b]	52	65
Severe acne overall	29.7	39	25
Psoriasis vulgaris overall	[c]	33	7.5
PPP, isolated	37.6		50
PPP + psoriasis vulgaris	20.8		0
PPP + severe acne	7.9[c]		12
PPP + hidradenitis suppurativa			2
Psoriasis vulgaris, isolated	11.9		8
Severe acne, isolated	21.8		23
Severe acne + hidradenitis suppurativa			5
Other manifestations			
Inflammatory bowel disease (*n*/%)	9 (7.5)	2 (4)	2 (3)
Of those Crohn's disease	6/9 (67%)	2/2 (100%)	2/2 (100%)
Of those ulcerative colitis	3/9 (33%)	0	0
HLA-B27 (%)	13	18	4

NA, not available; PPP, palmoplantar pustulosis.
[a]Median and interquartile range.
[b]Cannot be determined exactly from the data provided because some patients with SA had both PPP and PV, and only data for SA plus PPP and/or PV were reported.
[c]This figure is for the combination of severe acne with PPP and/or psoriasis vulgaris.

Nguyen MT, Borchers A, Selmi C, Naguwa SM, Cheema G, Gershwin ME. The SAPHO Syndrome. Semin Arthritis Rheum 2012;42(3):254-265.

DIAGNOSIS

DIFFERENTIAL DIAGNOSIS



DIAGNOSTIC CRITERIA

Box 1
Inclusion and exclusion criteria for SAPHO syndrome

Inclusion Criteria[a]

- Osteoarticular manifestations of acne conglobata, acne fulminans, or hidradenitis suppurativa
- Osteoarticular manifestations of PPP
- Hyperostosis (of the anterior chest wall, limbs, or spine) with or without dermatosis
- CRMO involving the axial or peripheral skeleton with or without dermatosis

Sometimes Reported

- Possible association with psoriasis vulgaris
- Possible association with inflammatory enterocolopathy
- Features of ankylosing spondylitis
- Presence of low-virulence bacterial infections

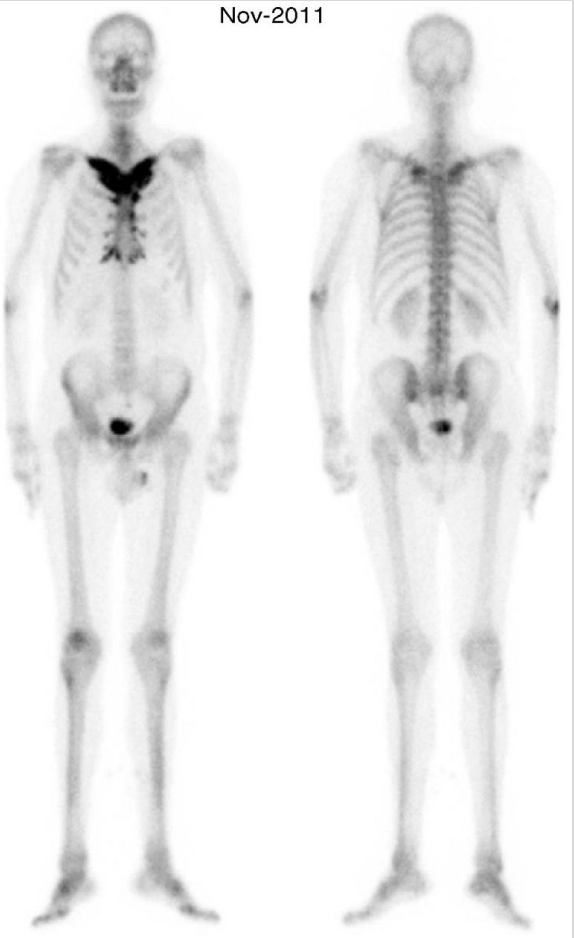
Exclusion Criteria

- Septic osteomyelitis
- Infectious chest wall arthritis
- Infectious PPP
- Palmoplantar keratoderma
- Diffuse idiopathic skeletal hyperostosis, except for fortuitous association
- Osteoarticular manifestations of retinoid therapy

[a] The presence of 1 of the 4 inclusion features is sufficient for a diagnosis of SAPHO syndrome.

Data from Benhamou CL, Chamot AM, Kahn MF. Synovitis-acne-pustulosis-hyperostosis-osteomyelitis syndrome (SAPHO): a new syndrome among the spondyloarthropathies? Clin Exp Rheumatol 1988;6(2):109–12.

PECULIAR SIGNS



Carneiro S, Sampaio-Barros PD. SAPHO Syndrome. Rheum Dis Cin North Am 2013;39(2):401-418.

Carranco-Medina TE, Hidalgo-Calleja C, Calero-Paniaguaa I, *et al.* Manifestaciones trombóticas en el síndrome SAPHO. Revisión de la literatura. Reumatol Clin 2015;11(2):108-111.

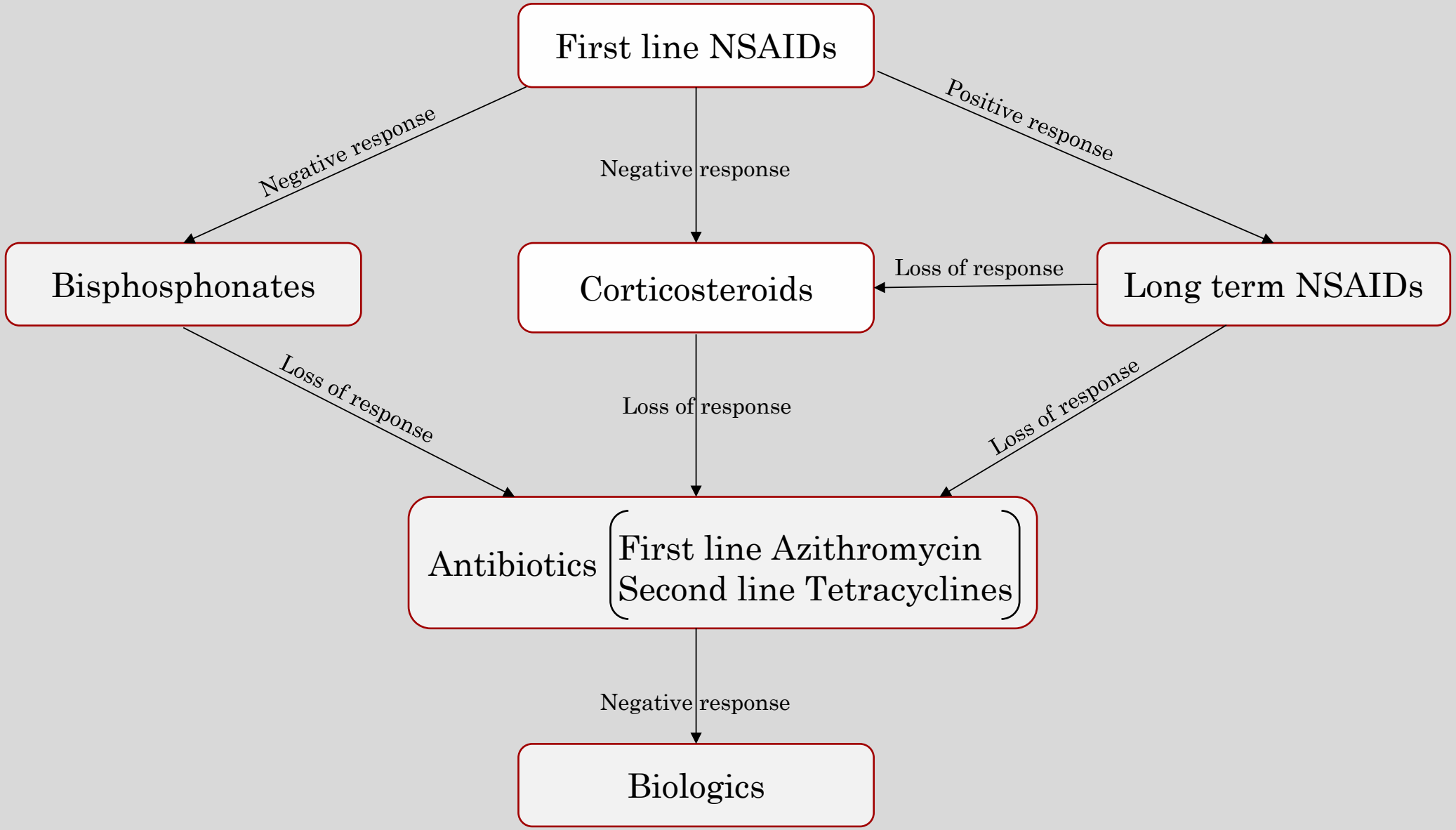
CASES

PZ	SEX	AGE	PPP	ACNE	ASEPTIC OSTEOMYELITIS	ARTHRALGY	EDEMA / SWELLING
1	F	60	+	-	-	+	-
2	F	36	+	-	-	+	+
3	F	55	+	-	+	+	-
4	F	42	+	-	-	+	+
5	F	55	+	-	-	+	+

THERAPY

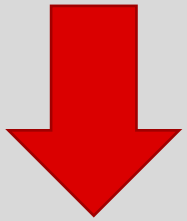
PZ	NSAIDs	CORTICOSTEROIDS	BISPHOSPHONATES	ANTIBIOTICS	BIOLOGICS
1	+	-	+	-	-
2	+	+	-	+	-
3	+	-	-	+	+
4	+	-	-	+	-
5	+	-	-	+	-

TREATMENT ALGORITHM



TAKE HOME MESSAGES

- **CHALLENGING DIAGNOSIS**
- **COLLABORATION Dermatologist-Rheumatologist**



- **EARLY DIAGNOSIS**
- **COST / EFFECTIVENESS**

THANK YOU FOR YOUR ATTENTION !

SPECIAL THANKS

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Prof.ssa Claudia LASAGNI

Dott.ssa Laura BIGI

Prof. Andrea COSSARIZZA