

## Prise en charge des maladies systémiques autoimmunes



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## Maladies systémiques autoimmunes : Connective tissue diseases

- Systemic disorder
- Vascular involvement is frequent
- Connective tissue is frequently altered
- No infectious or tumoral cause is easily identifiable
- Biomarkers of autoimmunity are frequently positive

### Clinical entities

- SLE
- SS
- SSc
- PM / DM
- MCTD
- RA
- ...

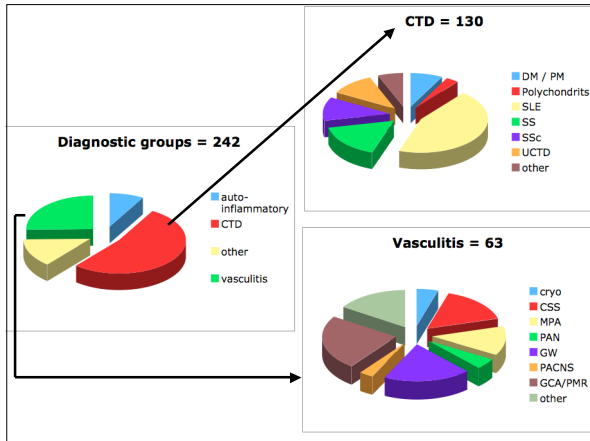
- Vasculitis
  - WG
  - MPA
  - GCA
  - Polyarteritis nodosa
  - Cryoglobulinemic
  - ...

- Anti-phospholipid syndrome
- ...

- Behçet disease
- AOSD
- ...

### Incidence and prevalence

			Incidence/10 <sup>5</sup>	Prevalence/10 <sup>5</sup>
• SLE	woman	Caucasian	0.4 - 7.2	50 - 150
• SLE	woman	Afro-Amer	10 - 40	40 - 408
• SLE	men	all	0.4 - 1.3	7 - 9
• SSc	woman	all	0.4 - 1.6	} 0.4 - 25.3
• SSc	men	all	0.1 - 0.2	
• GW			0.2 - 1.2	2.4 - 15.7
• GCA		(> 50 y)	9.4 - 76.6	135 - 223



### Problems associated with the management of collagen vascular diseases

- Identification of the precise clinical entity of which the patient suffers
- Defining the prognosis
- Establishing the level of disease activity
- Lack (paucity) of evidence-based therapeutic approaches
- Tailoring the therapy to the characteristics of the patient and disease activity

**EUSTAR**

- EUSTAR: European Scleroderma Trials And Research group ([www.eustar.org](http://www.eustar.org))
- SCTC, scleroderma clinical trial consortium
  - Until therapies for scleroderma are documented to be effective, protocol participation is the treatment of choice ([www.sctc-online.org](http://www.sctc-online.org))

Eular Scleroderma Trials And Research group

**ASTIStrial**  
 •Autologous•Stemcell•Transplantation•  
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patients with severe systemic sclerosis

Immunoablation + SCT =	Control treatment =
1. Mobilization (CYC 2x2 g/m <sup>2</sup> , G-CSF 10 mg/kg) 2. Leukapheresis/CD34-selection 3. Conditioning (CYC 200 mg/kg, rbATG 7.5 mg/kg) 4. Reinfusion CD34+ cells	12x monthly i.v. pulse CYC 750 mg/m <sup>2</sup>
For 1.80 m <sup>2</sup> and 70 kg body weight:	
CYC: 7.2 g mobilization 14 g conditioning	CYC tot: 16.2 g



ASTIS trial March 2001 – September 2009:  
151/155 patients randomized in 24 centers

