Sarcoïdose: *introduction*

Epidémiologie-diagnostic

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COI disclosure

- Pas de COI avec la présentation
- Membres de conseils scientifiques français et internationaux sur la FPI (Bohringer-Ingelheim, Roche)
- PI et Membre de *steering committee* sur le traitement de la FPI
- Lecture aux « avancées de pneumologie » (Astra)

Introduction

- Epidémiologie/mortalité
- Diagnostic souvent/parfois difficile

Epidémiologie diverse

	prévalence	incidence	F/M	origine
US	141/10^5 AA 49/10^5 Cauc		F>>M	
UK		5/10^5		
France	32/10^5 10/10^5 Cauc	4.9/10^5	F>M	Afrique>Maghreb>Europe
Suède	160/10^5		M>F	
Japon		1/10^5		

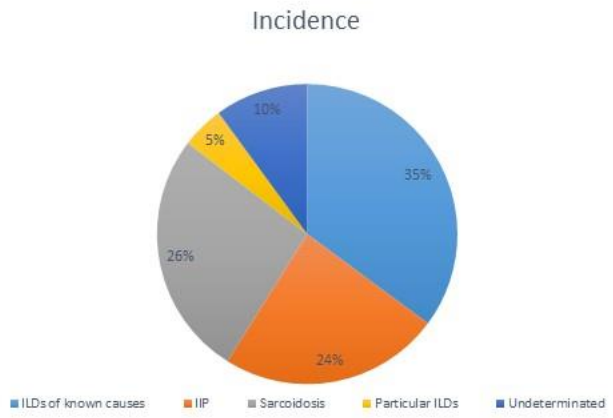
Formes familiales: instructives pour mieux comprendre la pathogénie; expression semblable aux formes sporadiques

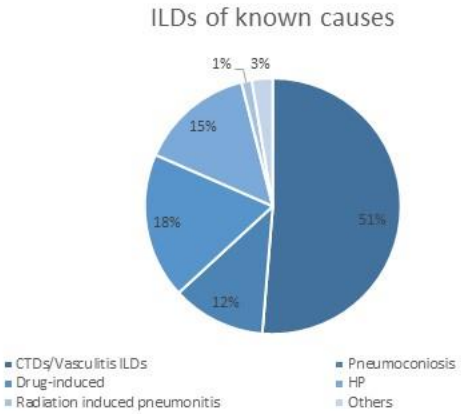
Arkema ERJ 2016; Baughman Ann ATS 2016; Gribbin Thorax 2006
Morimoto ERJ 2008; Duchemann ERJ 2017; Pacheco OJRD 2016

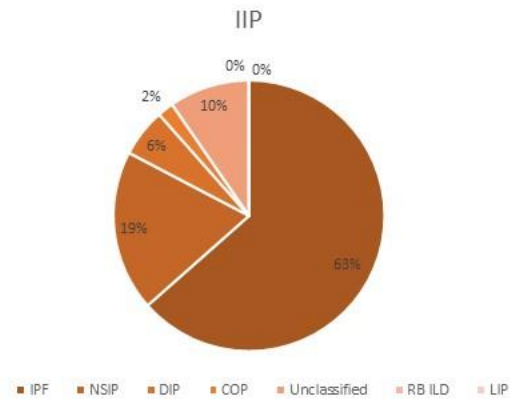
ILD cases	Seine Saint Denis population > 15 years old (1 194601 inhabitants)			
	N	Prevalence (/10^5)	n	Incidence (/10^5/year)
All identified cases	1170	97.9	232	19.4
Reviewed cases	848	71.0	219	18.3
ILDs of known cause	260	21.8	77	6.5
CTDs/vasculitis	145	12.1	39	3.3
Pneumoconioses	42	3.5	9	0.8
Drug-induced ILD	31	2.6	14	1.2
Hypersensitivity pneumonitis	28	2.3	11	0.9
Radiation induced pneumonitis	7	0.6	1	0.1
Others *	7	0.6	2	0.3§
Idiopathic interstitial pneumonias	145	12.14	52	4.4
IPF	98	8.2	33	2.8
NSIP	20	1.7	10	0.8
Desquamative interstitial pneumonia	10	0.8	3	0.3
Organizing pneumonia	9	0.8	1	0.1
Unclassified (despite SLB)	6	0.5	5	0.4
Respiratory bronchiolitis with ILD	2	0.2	0	0.0
Lymphoid interstitial pneumonia	0	0.0	0	0.0
Sarcoidosis	361	30.2	58	4.9
Particular ILDs	22	1.8	10	0.8
Lymphangioleiomyomatosis	9	0.8	4	0.3
Chronic idiopathic eosinophilic pneumonia	5	0.4	1	0.1
Pulmonary Langerhans cell histiocytosis	4	0.3	2	0.2
Pulmonary alveolar proteinosis	2	0.2	1	0.1
Others †	2	0.2	2	0.1
Undetermined diagnosis	60	5.0	22	1.8
Differential diagnosis between IPF and NSIP	34	2.9	13	1.1

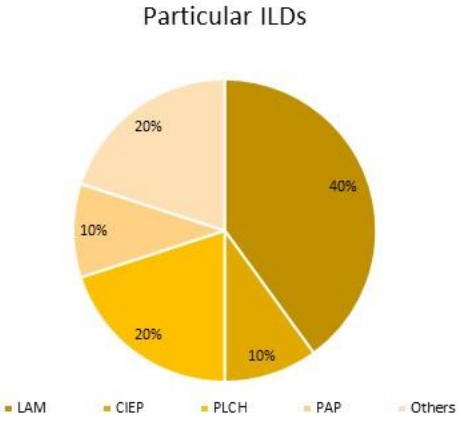
B Duchemann Eur Respir J 2017

Incidence des PID









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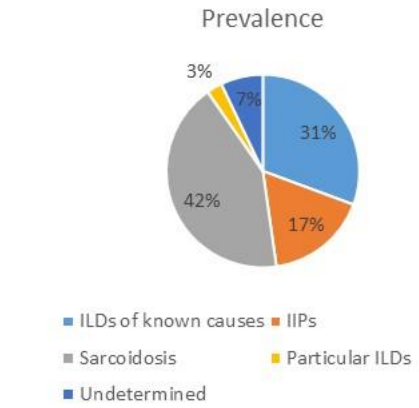
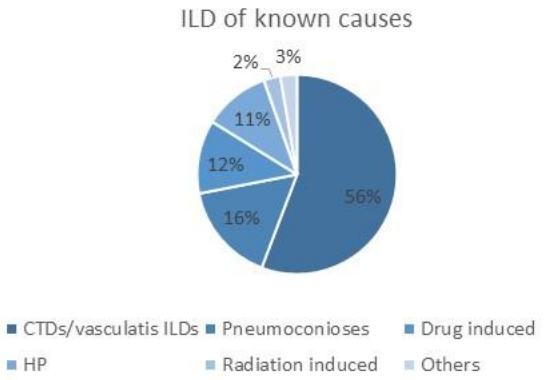
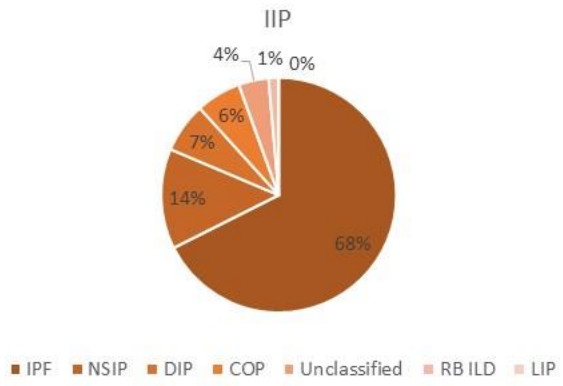
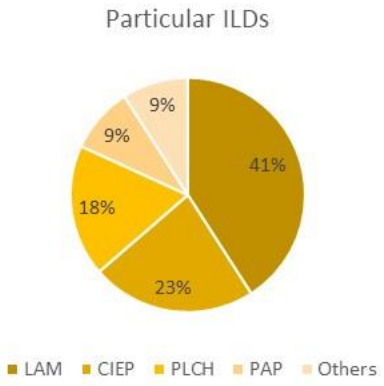
Prévalence des PID









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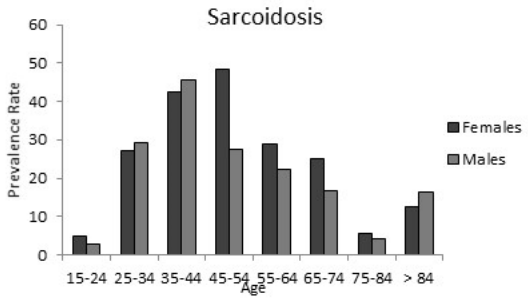
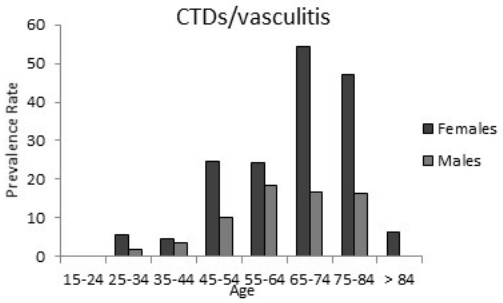
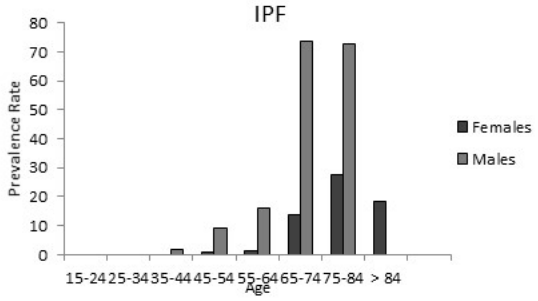
Prévalence des PID selon l'âge







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Nathan Thorax 2014

Table 3: Odd ratio by geographical origin for the main etiological diagnoses (multinomial logistic regression analysis, reviewed cases)

Diagnosis	Europeans	Afro-Caribbeans	Maghrebi	Others
Sarcoidosis **OR (95% CI)** **p-value**	1 NA	2.972 (1.615-5.468) < 0.001	1.822 (1.118-2.970) 0.016	1.727 (0.792-3.769) 0.170
IPF **OR (95% CI)** **p-value**	1 NA	0.604 (0.163-2.241) 0.451	1.418 (0.824-2.440) 0.208	0.288 (0.036-2.303) 0.240
CTDs/vasculitis ILDs **OR (95% CI)** **p-value**	1 NA	4.383 (2.210-8.693) < 0.001	1.767 (1.032-3.027) 0.038	2.191 (0.931-5.157) 0.073

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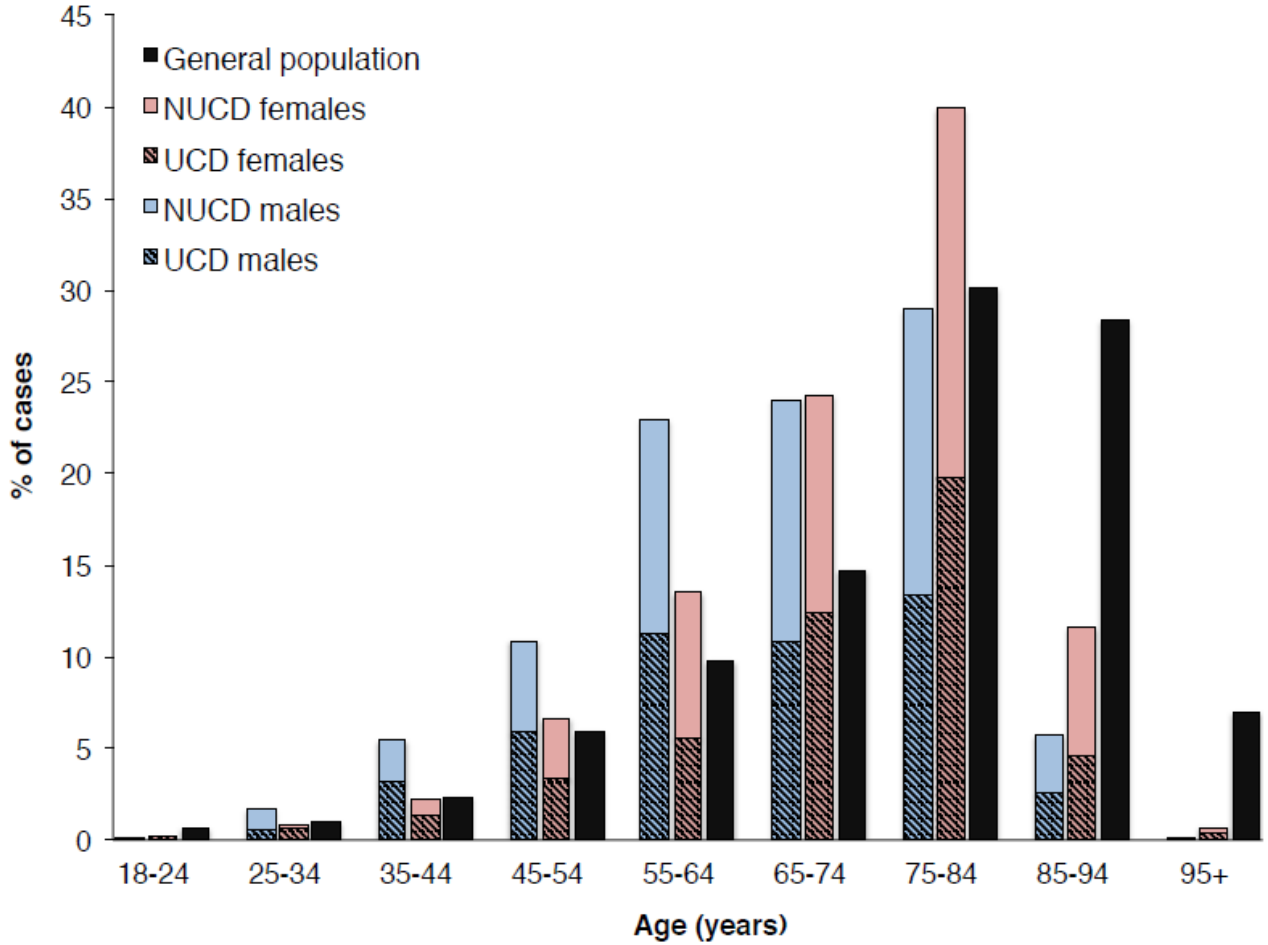
Mortalité par sarcoïdose

- « suivi des patients » (jusqu'à 7% DC)
  - Dépend du mode de recrutement (primaire ou tertiaire) et de la durée de suivi
- Études autopsiques
  - USA: poumon > cœur > SNC
  - Japon: cœur (70% des DC par sarcoïdose)
- Certificats de DC (OMS)
- « Mécanismes » conduisant au DC

Mortalité: sarcoidose *vs* population générale



Swigris AJRCCM 2011; Jamilloux Eur Respir J 2016

Sarcoidosis mortality in Sweden: a population-based cohort study

Marios Rossides[1], Susanna Kullberg[2], Johan Askling[1,3], Anders Eklund[2], Johan Grunewald[2], and Elizabeth V. Arkema[1]

[1] Clinical Epidemiology Unit, Dept. of Medicine, Solna, [2] Respiratory Medicine Unit, Dept. of Medicine, Solna & Center for Molecular Medicine, [3] Rheumatology Unit, Dept. of Medicine, Solna,
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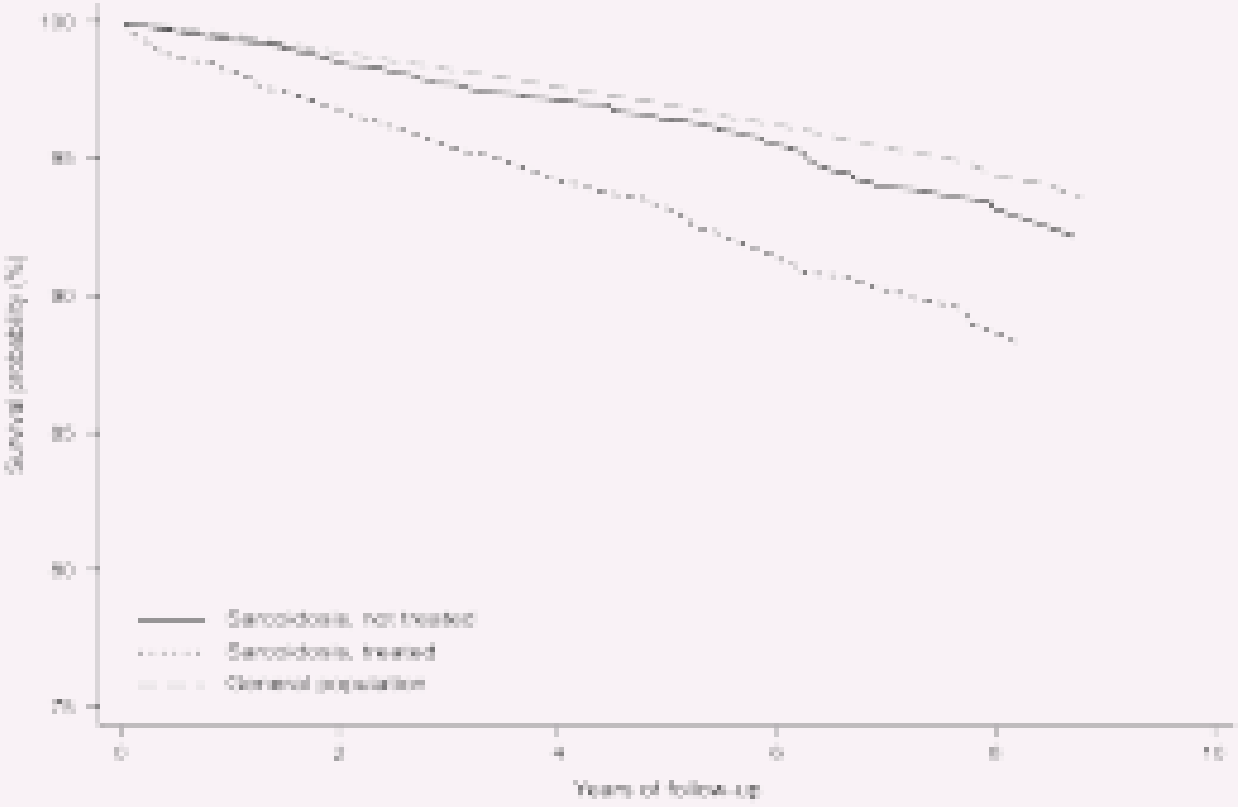
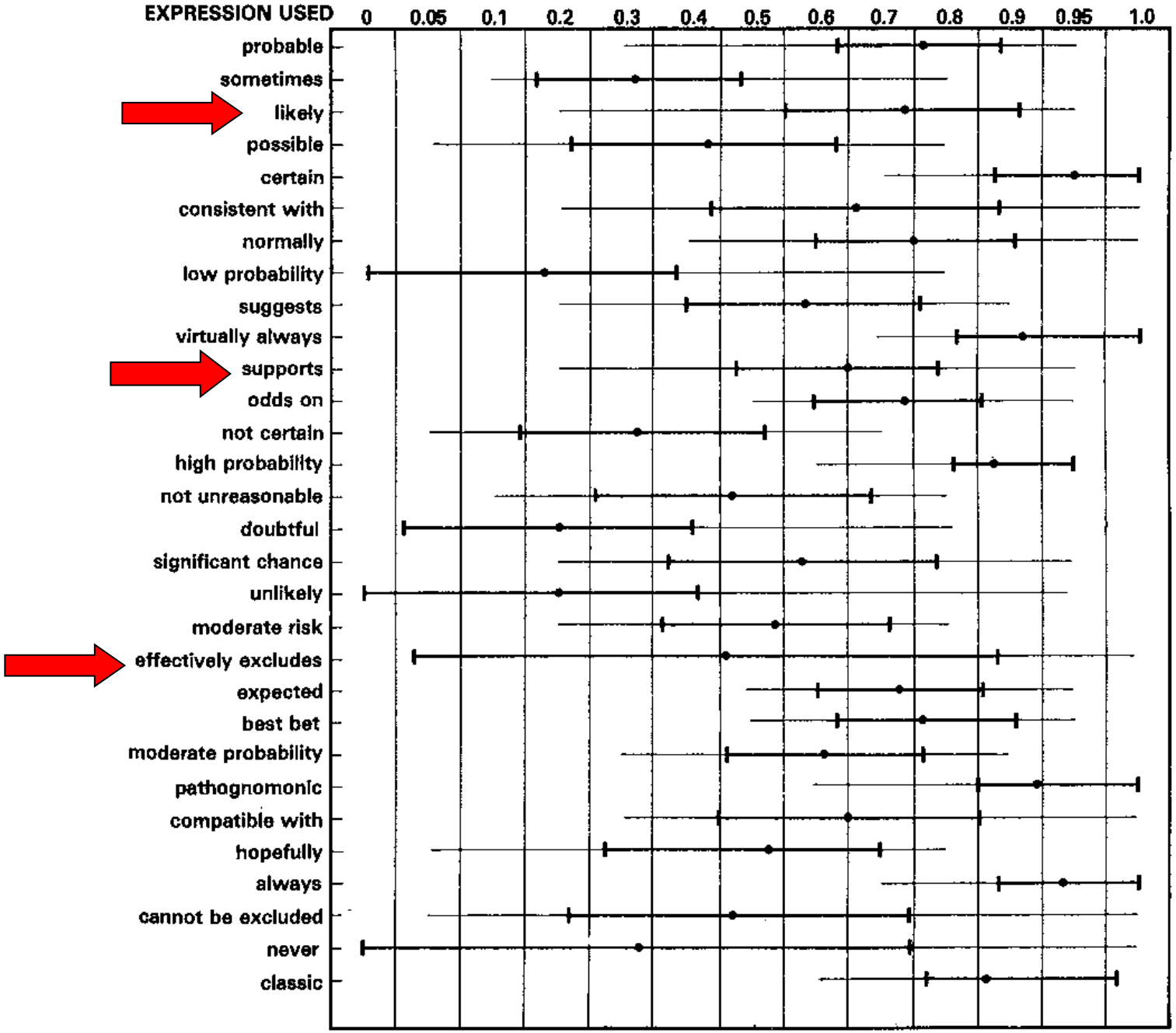


Figure 2. Adjusted survival probability functions stratified by treatment status around diagnosis.

Diagnosis of sarcoidosis *per se*

- The diagnosis of sarcoidosis is based on the following*:
  - A compatible clinical and/or radiological picture
  - Histological demonstration of noncaseating granulomas
  - Exclusion of other diseases capable of producing a similar histological or clinical picture
- The diagnosis rests on**:
  - the correct clinical setting, typical chest radiograpic or CT appearances and
  - a biopsy showing non-caseationg granulomas

**Statement, AJRCCM 1999; **BTS guidelines, Thorax 2008*


EXPRESSION USED
0
0.05
0.1
0.2
0.3
0.4
0.5
0.6
0.7
0.8
0.9
0.95
1.0
probable
sometimes
likely
possible
certain
consistent with
normally
low probability
suggests
virtually always
supports
odds on
not certain
high probability
not unreasonable
doubtful
significant chance
unlikely
moderate risk
effectively excludes
expected
best bet
moderate probability
pathognomonic
compatible with
hopefully
always
cannot be excluded
never
classic

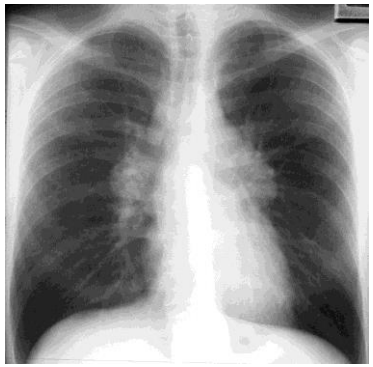
NEJM 1980

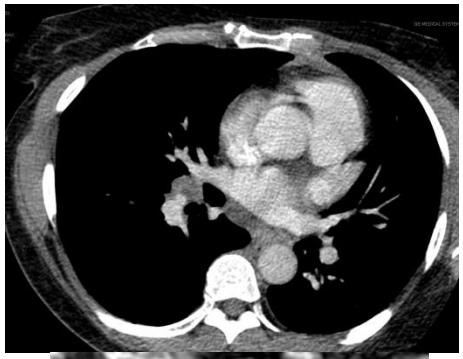
Diagnostic

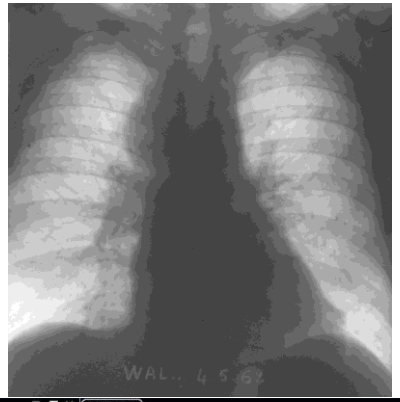
- 3 critères mais leur poids respectif dépend du scenario au Dc
- Retard Dc et recours à x Médecins avant un Dc
- « working diagnosis » +++: probabilité plus précise avec le temps
- Sites de prélèvements
  - Algorithme
  - EBUS (↕ médiastinoscopie)
  - Biopsie de GG périphériques sous échographie (PYB, SVDLD 2016)
  - Biopsie de nodule conjonctival
  - Site orienté par TEP

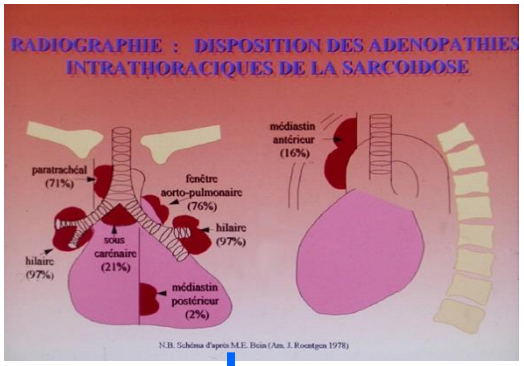
Judson Chest 2004; Baughman AJRCCM 2010; Culver Curr Opin Pulm Med 2015

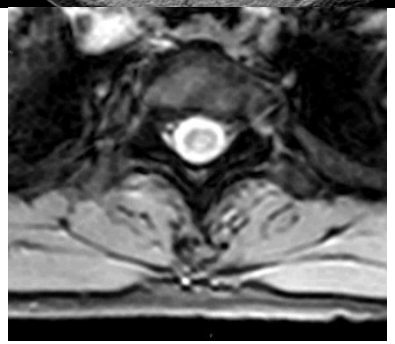
Thoracic lymphadenopathy





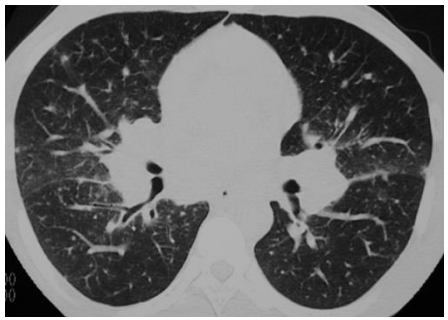


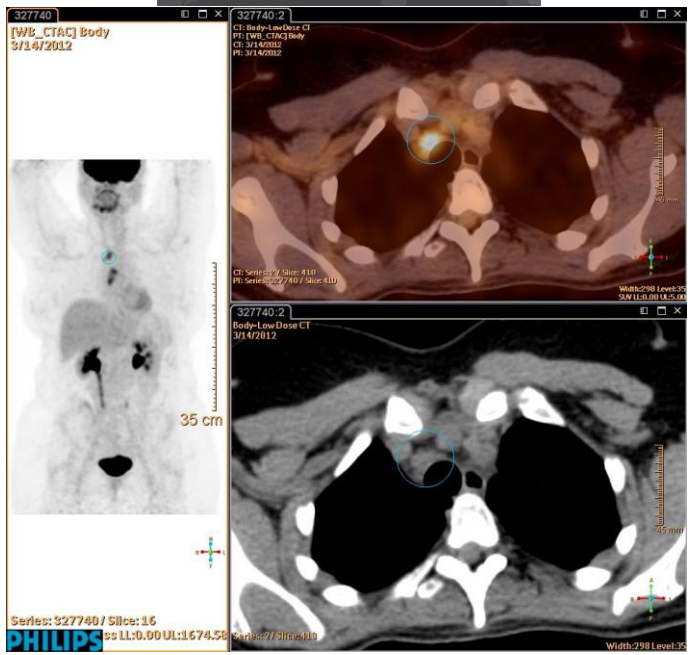




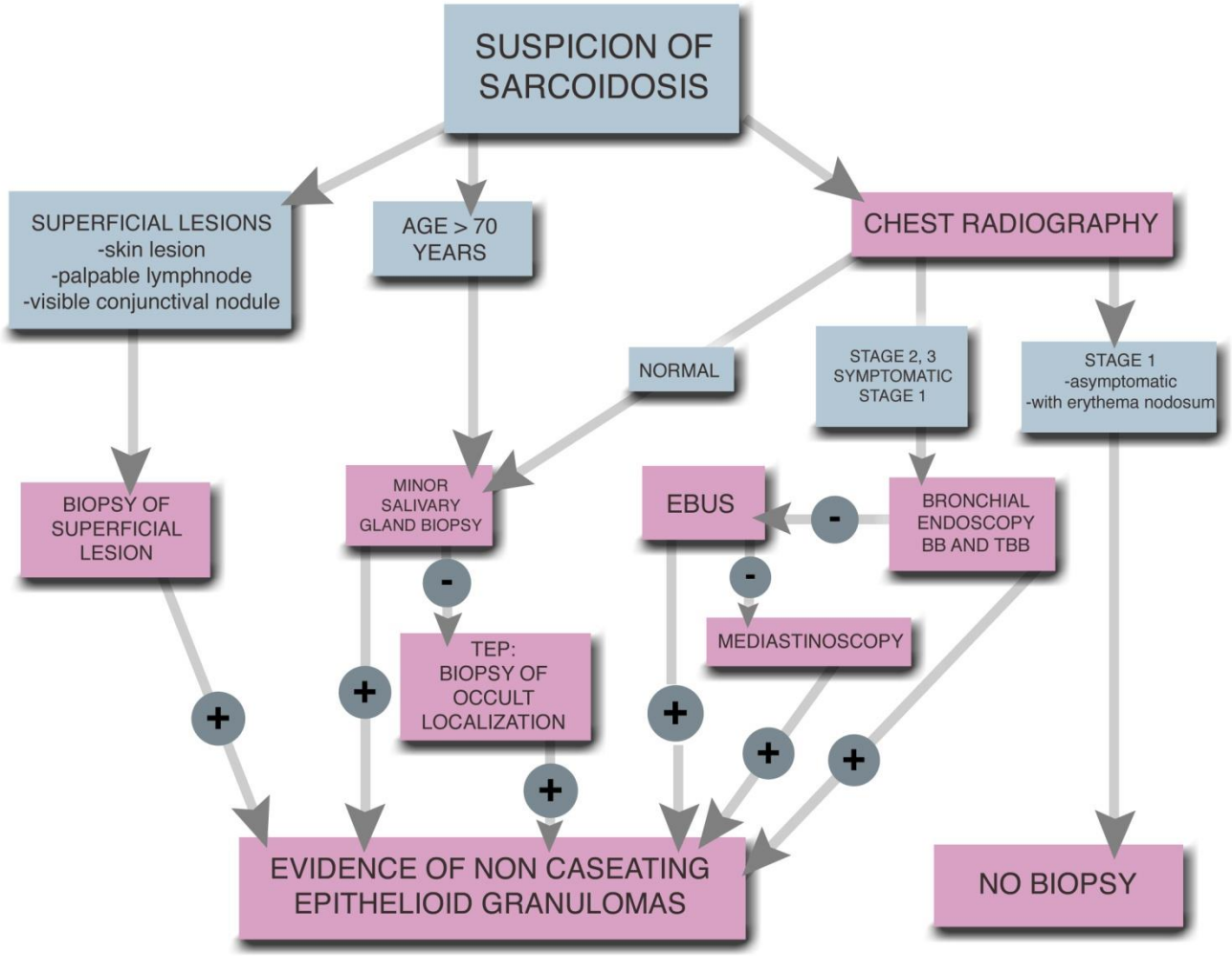
EN, uveitis, isolated

+ -





Bein AJR 1978: Neville OJM 1983: Butt SVDLD 2011



Valeyre Lancet 2014; Benouar SVDLD 2016

Diagnostic différentiel

	Main diseases	Etiological agent	Diagnosis criteria
Infections	Tuberculosis, histoplasmosis, leprosy, Whipple's disease	Infectious agents	Epidemiologic factors, microbiology
Environmental agents	Chronic beryllium disease	Beryllium	Exposure to beryllium; beryllium hypersensitivity
Drugs	Drug-induced granulomatosis	IFN α and β; anti-TNFα; intravesical BCG-therapy; natalizumab; alemtuzumab; inhibiteurs CTLA4; inhibiteurs PD1 et PDL1	Anamnesis
Immuno-deficiency	Common variable immuno-deficiency; Chronic granulomatous disease	-	Recurrent infections ; hypogammaglobulinemia Recurrent pyogenic infections ; defective respiratory burst in phagocytes
Genetic disease	Blau's syndrome	-	Presentation; familial history; genetic investigation
Proliferations	Lymphomas Solid neoplasias	-	Pathology; molecular biology
Unknown origin	Wegener's granulomatosis Crohn's disease Primiray biliary cirrhosis	-	Presentation; ANCA Presentation Presentation ; anti-mitochondrial antibodies ; pathology

Remerciements

- Avicenne + P13 (EA2363)
  - H Nunes; Y Uzunhan; D Bouvry; O Freynet; F Jeny
  - C Planès; PY Brillet; M Brauner; D Piver; M Kambouchner; JF Bernaudin
- Equipes de génétique (A Calender; Y Pacheco); immunologie (M Miyara; G Gorochov); épidémiologie et environnement (I Annesi-Maesano; PA Rosental); modèles (C Planès; V Besnard; F Jeny)
- Groupe GSF, groupe de travail officiel de la SPLF
  - F Cohen-Aubart, M Mahévas, B Wallaert, M Miyara, JJ Boffa
  - P Sève, Y Jamilloux