



SYNDROME CATASTROPHIQUE DES ANTIPHOSPHOLIPIDES (CAPS) : ÉTUDE RÉTROSPECTIVE MULTICENTRIQUE DESCRIPTIVE

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INTRODUCTION

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- Complication rare d'une maladie rare ... (1% des SAPL)
- Complication grave ! (mortalité 37%)
- Peu de données

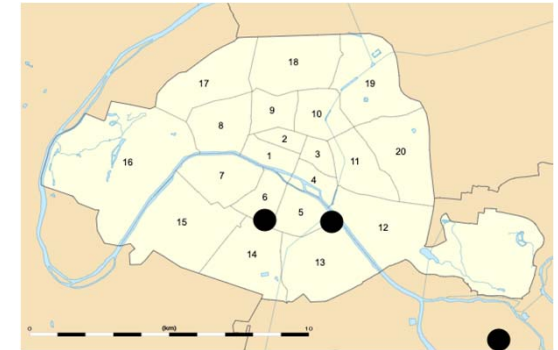
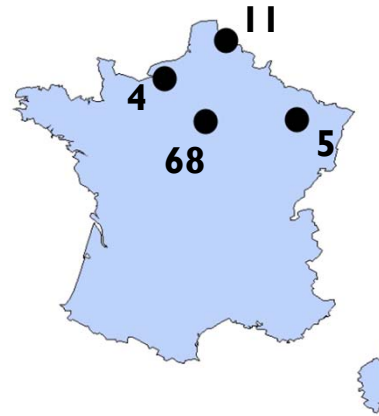


PATIENTS ET MÉTHODES

Recrutement

- Multicentrique

(Lille, Rouen, Nancy, Cochin, Pitié, Mondor)



- Recueil rétrospectif (base SAPL-LUPUS Cochin / Lille)

- Accord CPP, CNIL, ...



Critères CAPS

- 1 Evidence of involvement of three or more organs, systems and/or tissues^a
- 2 Development of manifestations simultaneously or in less than a week
- 3 Confirmation by histopathology of small vessel occlusion in at least one organ or tissue^b
- 4 Laboratory confirmation of the presence of antiphospholipid antibodies (lupus anticoagulant and/or anticardiolipin antibodies)^c + **Anti β 2 GPI**

Definite catastrophic APS

- All 4 criteria

Probable catastrophic APS

- All 4 criteria, except for only two organs, systems and/or tissues involvement
- All 4 criteria, except for the absence of laboratory confirmation at least **X** weeks apart due to the early death of a patient never previously tested for aPL prior to the catastrophic APS event
- Criteria 1, 2 and 4
- Criteria 1, 3 and 4 and the development of a third event in more than a week but less than a month, despite anticoagulation

12

Asherson RA et al, Lupus, 2003

Erkan D et al, Autoimmun Rev, 2010

RÉSULTATS

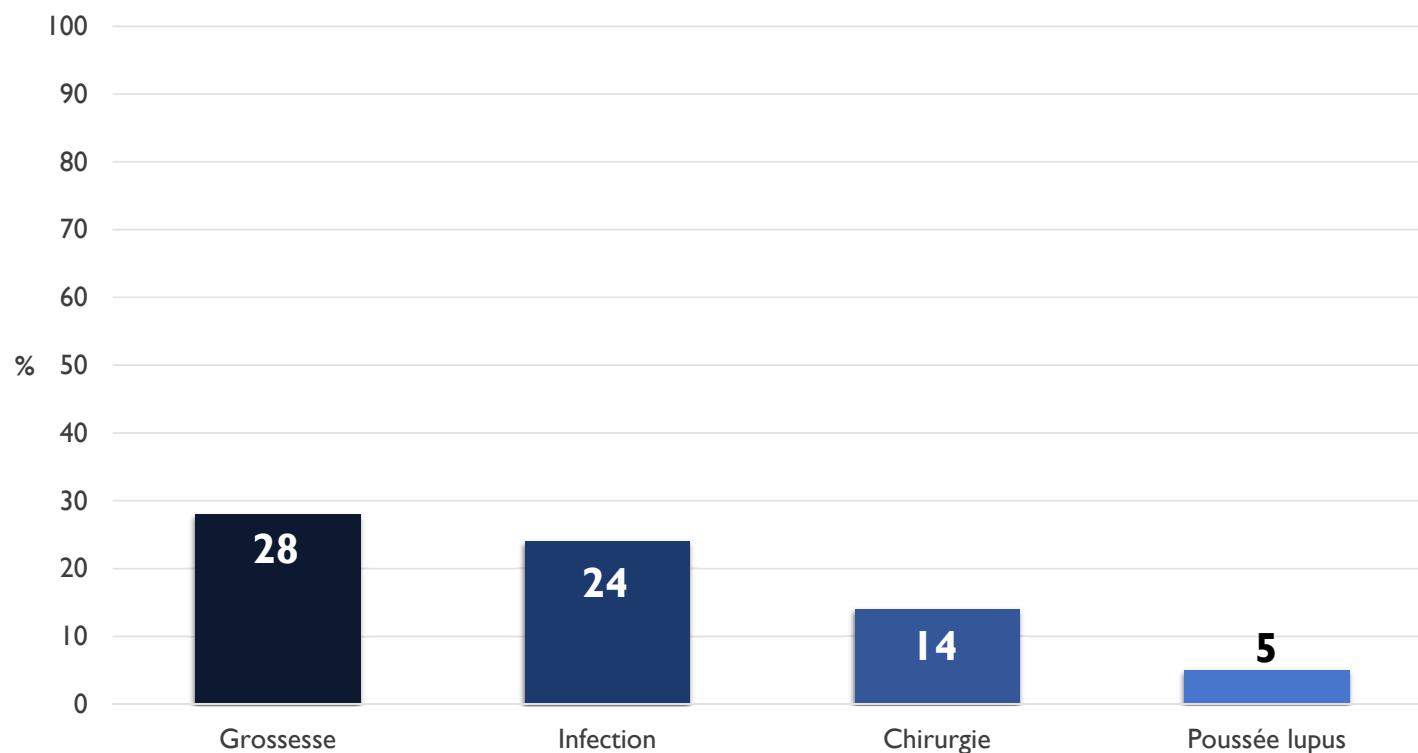
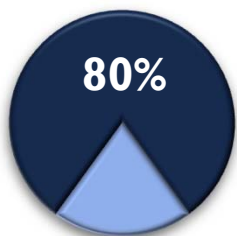
Démographie

CAPS certain / probable
17 (19%) / 71 (81%)

Caractéristiques	n = 88	Anticorps	n = 87
Sexe (%)	59 F (67)	ACC (%)	78 (90)
Age médian (ans)	39 [16-78]	ACL (%)	79 (91)
CAPS inaugural (%)	30 (34)	Anti β 2 GPI (%)	58 (67)
Lupus (%)	32 (36)	Triple + (%)	51 (59)

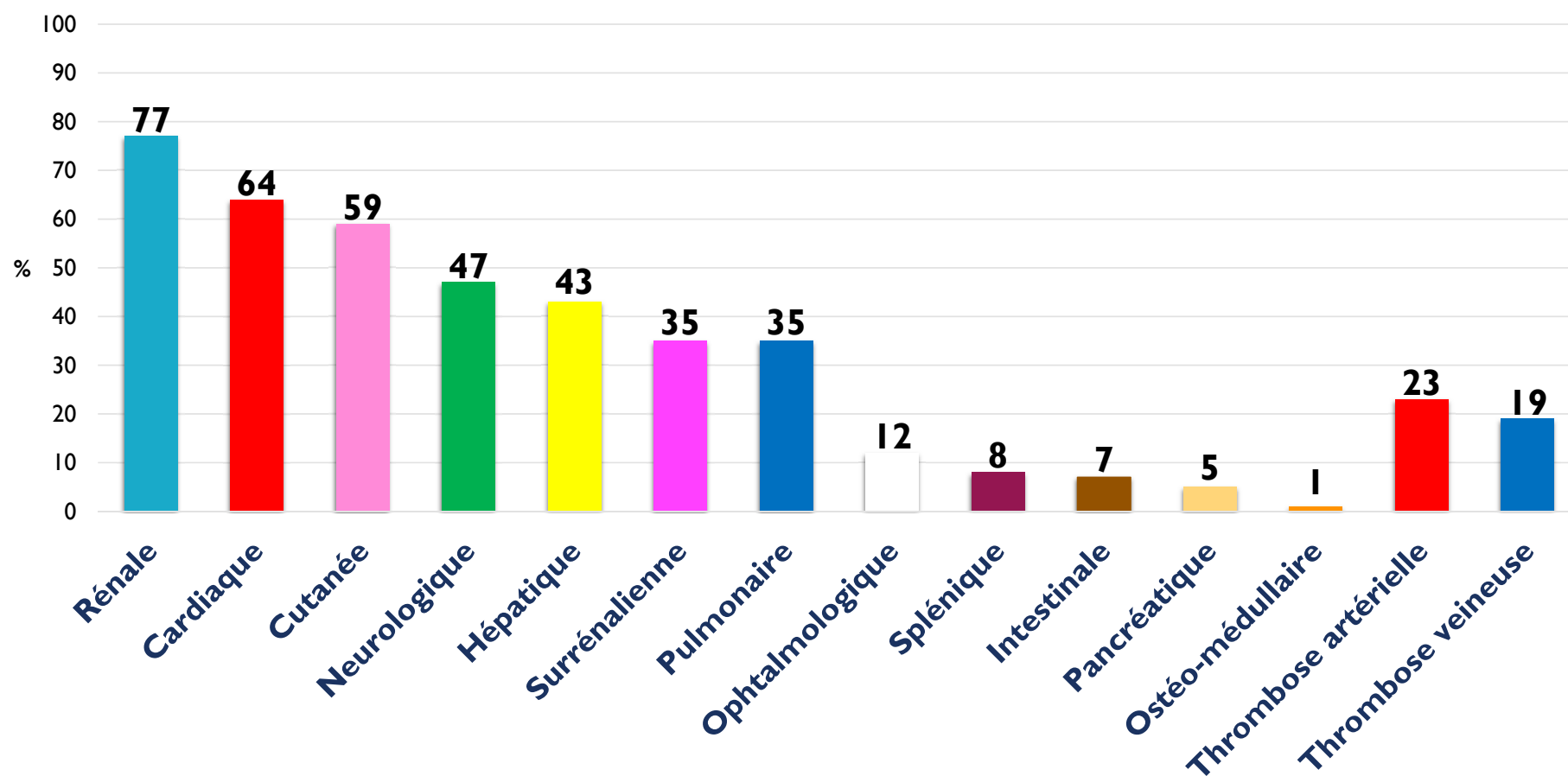
Facteur déclenchant (n=74)

Au moins 1 facteur déclenchant

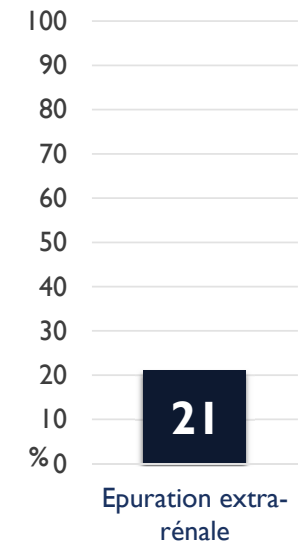
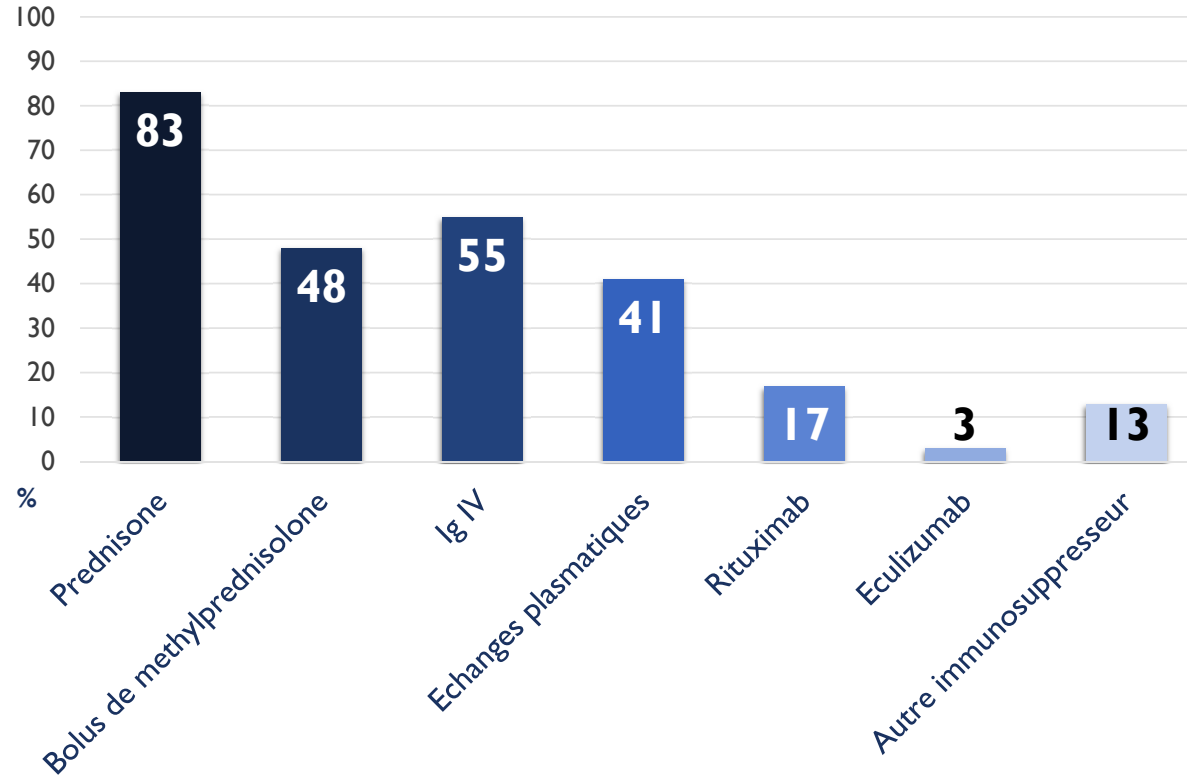
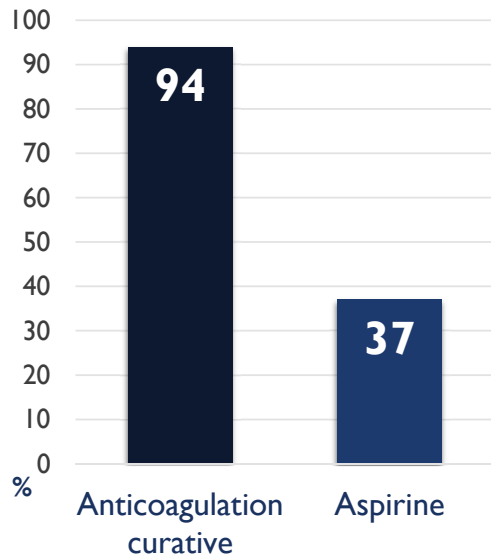


54 Patients SAPL thrombotique → 22 arrêt ou relais AVK (41%) + 10 sous dosage AVK (18%)

Atteintes d'organe (n=88)

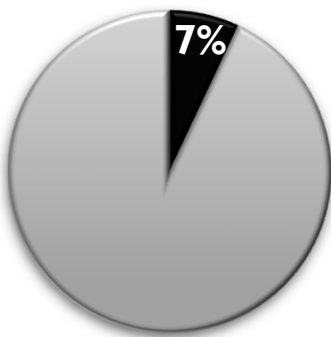


Traitement CAPS (n=87)

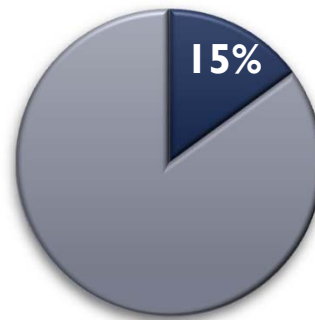


Pronostic

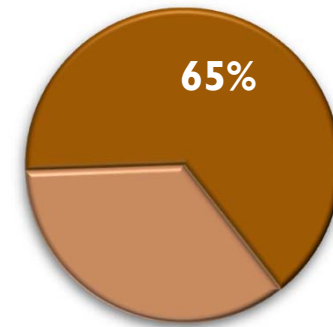
Décès 1er épisode



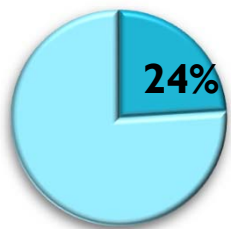
Récidive CAPS



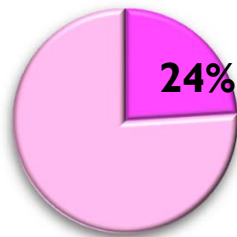
Séquelles



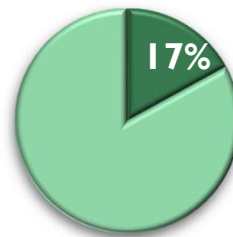
Insuffisance rénale chronique



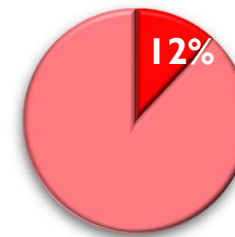
Insuffisance surrénalienne



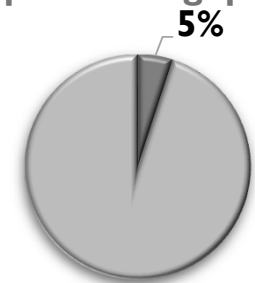
Séquelles neurologiques



Insuffisance cardiaque



Séquelles ophtalmologiques





DISCUSSION

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- Données préliminaires certes ...
- Mais déjà une centaine de cas !
- Recrutement qui ne demande qu'à augmenter



Merci de votre attention !