

# aPL+ /APS subset of SLE

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A CROSS-SECTIONAL ANALYSIS FROM A SWEDISH COHORT

aPL in SLE

Common (30-40%)

APS may develop in 50% over 20 year follow-up

*Pons-Estel GJ et al, J Autoimmun 2017*

# APS

2006 classification criteria ("Sydney criteria")

## Laboratory tests:

+ aCL IgG or IgM

- Medium/high titer
- > 99<sup>th</sup> percentile, or > 40 GPL units

+ a $\beta_2$ GP1 IgG or IgM

+ Lupus anticoagulant (LA)

**At least 2 positive tests (persistent positivity)**

**12 weeks apart**

## Clinical manifestations

### Thrombosis

- Arterial, venous, small vessels; confirmed by imaging or histopathology

### Pregnancy morbidity

- Miscarriages
  - 1 late (after week 10)
  - 3 early (before week 10)
- Premature delivery
  - Before week 34
  - Preeclampsia
  - Placenta insufficiency

*Miyakis et al. J Thrombosis and Haemostasis 2006*

# Antiphospholipid antibodies/syndrome & Damage in SLE

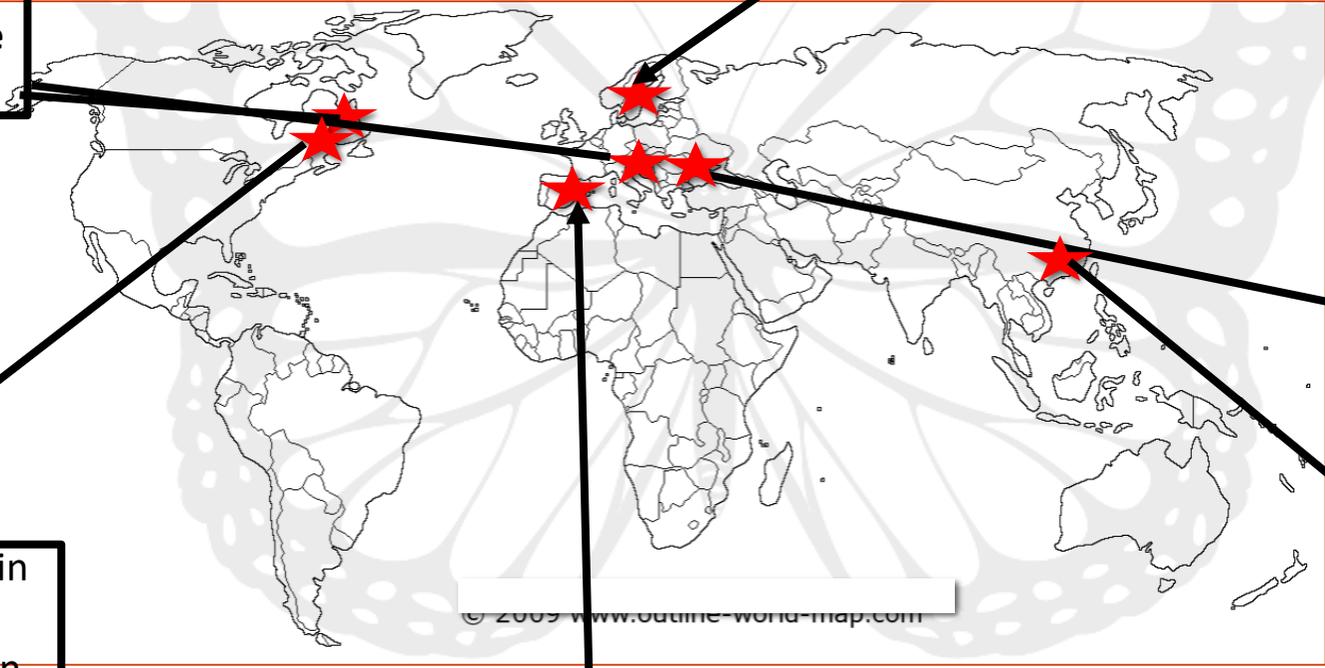
(SLICC-DI)

Significant **Damage increase** over 5-15 years assoc with **significant aPL profile**  
*Taraborelli et al Lupus 2016*

?

**aCL/LA Cluster** associated with overall SLICC damage, specifically with:  
**Arterial**  
**Venous**  
**Neuropsychiatric**  
*Arrtim-Esen J Rheumatol 2014*

**Damage increased faster in LA positive patients**  
LA stronger predictor than dsDNA for arterial events  
*Petri et al A & R 2012*



**Damage** accrual higher in patients with with **APS (11%)** at 5 and 15 years  
*Ruiz-Irastorza, Arch Inten Med 2012*

**aPL** associated with **Neuropsychiatric damage**  
*Mok et al Medicine 2006*

# Aim

Characteristics of the aPL+/APS subgroup of consecutive SLE patients seen at Karolinska University Hospital network 2004-2014, assessed cross-sectionally



503 SLE  
patients  
( $\geq 4$   
1982  
revised  
ACR  
Criteria)

**ACR items**

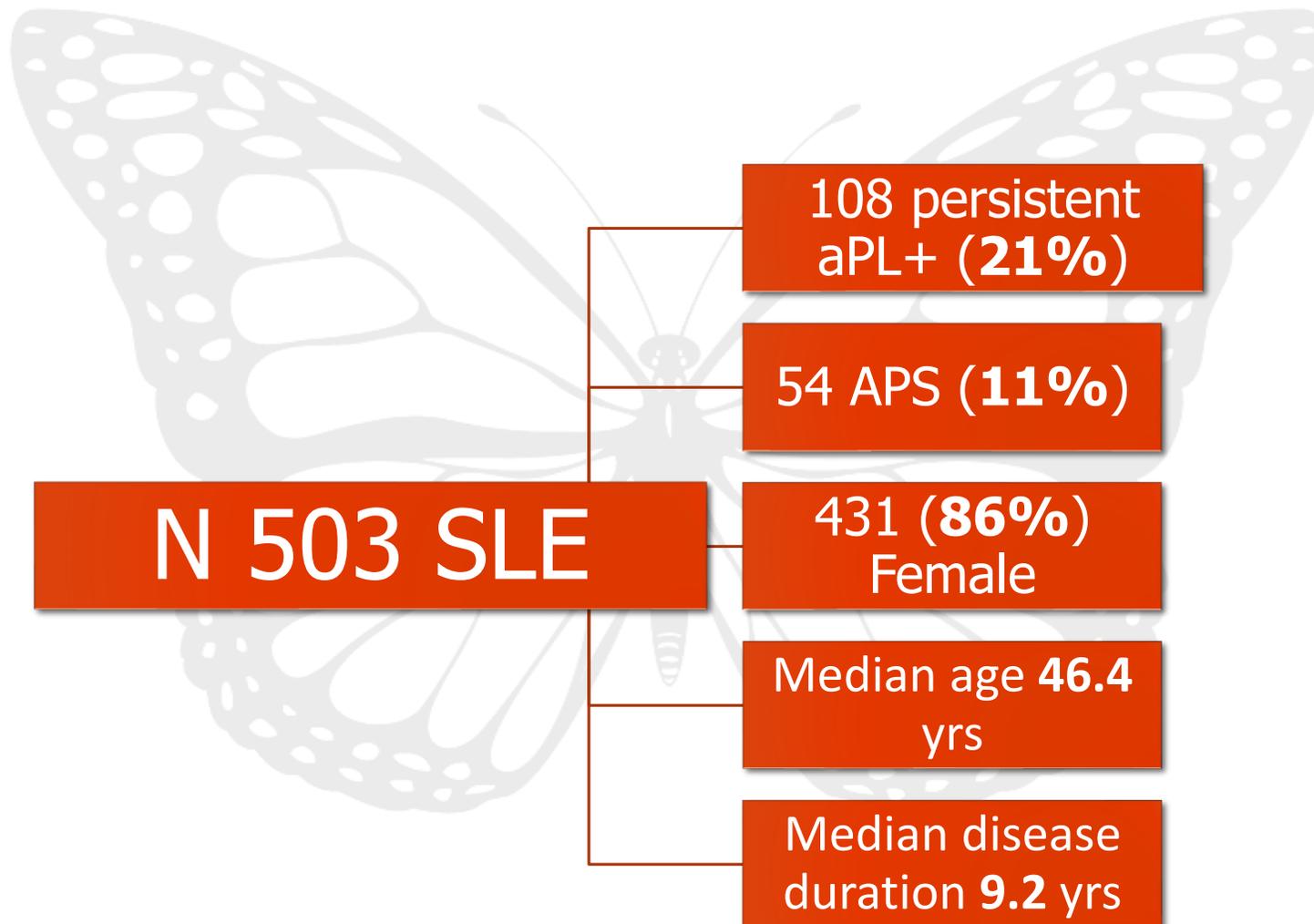
**APS and persistently positive aPL –  
Sydney 2006 Criteria**

Organ damage – **SLICC Damage Index (SDI)**

Disease activity – Systemic Lupus Activity Measure  
**SLAM**

# SLICC/ACR SLE Damage Index (SDI)

12 categories (40 items)	Ocular
	Neuropsychiatric
	Renal
	Pulmonary
	Cardiovascular
	Peripheral vascular
	Gastrointestinal
	Musculoskeletal
	Skin
	Premature gonadal failure
	Diabetes
	Malignancy



	<b>APS N=54 (11 %)</b>	<b>No APS N=449 (89%)</b>	<b>p</b>	<b>Persistent aPL+ N=108 (22%)</b>	<b>No persistent aPL+ N=395 (78%)</b>	<b>p</b>
<b>Age</b> (median + IQR)	47.7 (35-58)	46 (33-57)	ns	47 (33-57.6)	47 (34-57.6)	ns
<b>Gender</b> Female (n, %)	44 (81%)	387 (86%)	ns	90 (83%)	339 (86%)	ns
<b>Disease duration</b> (median + IQR)	6.7 (0.9-14)	9.5 (2-20)	0.05	9.8 (2.7-21.7)	8.8 (1.7-18.8)	ns
<b>SLAM&gt;6</b>	26 (48%)	220 (49%)	ns	49 (45%)	195 (49%)	ns

# Characteristics of aPL+/APS subsets in SLE vs. other SLE patients, $N_{\text{total}} = 503$

	<b>APS N=54 (11 %)</b>	<b>No APS N=449 (89%)</b>	<b>p</b>	<b>Persistent aPL+ N=108 (22%)</b>	<b>Not persistent aPL+ N=395 (78%)</b>	<b>p</b>
Butterfly erythema	39%	49%	0.04	31%	51%	0.01
Discoid skin lesion	9%	18%	0.09	9%	20%	0.008
Pericarditis	15%	18%	0.07	10%	20%	0.002
Hemolytic anemia	11%	5%	0.03	11%	4%	0.0001
Thrombocytopenia	32%	18%	0.03	34%	16 %	0.0001
Anti-dsDNA	79.6%	65.8%	0.04	78.7%	64%	0.004
C4	0,13	0.16	0.02	0,13	0.16	0.0004
C3	0.82	0.89	0.02	0.82	0.90	0.0007
<b>SDI <math>\geq 2</math> (n, %)</b>	30 (57%)	156 (35%)	0.004	52 (48%)	134 (34%)	0.007
<b>SDI <math>\geq 3</math> (n, %)</b>	24 (45%)	83 (18%)	<0.0001	33 (31%)	74 (19%)	0.009

# aPL+ alone

	aPL+APS- N=54	aPL- N=395	p
Disease duration (median + IQR)	13.2 (5.4 – 27.2)	9.2 (1.9- 18.7)	0.004
Butterfly erythema	30%	51%	0.01
Hemolytic anemia	12%	4%	0.03
Thrombocytopenia	32%	16%	0.002
Anti-dsDNA	77.6%	64%	0.04
C4	0.13	0.16	0.02
SDI: GFR < 50%	15%	4.5%	0.01

# SLE + aPL positivity

Complement  
consumption/  
activation

Thrombocytopenia

Anti-dsDNA

Hemolytic anemia

Damage (SDI)

APS-defining events +  
cognitive impairment  
or major psychosis

Discoid skin lesions

Pericarditis

Butterfly erythema





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 *Konung Gustaf V:s 80 års fond*

 Stockholms läns landsting

