ΑΓΓΕΙΙΤΙΔΕΣ

ΝΕΩΤΕΡΑ ΔΕΔΟΜΕΝΑ - 2015

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OUTLINE

1. ANCA-associated Vasculitis:
   - Clinical features
   - Treatment
2. Behcet disease
3. Giant Cell Arteritis
4. Takayasu's Arteritis
5. Primary Central Nervous System Vasculitis

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CNS INVOLVEMENT OF GPA: CLINICAL -RADIOLOGICAL PRESENTATION DISTINGUISHES DIFFERENT OUTCOMES

DE LUNA G ET AL. RHEUMATOLOGY 2015;54

- **AIM:** CNS involvement presentation and outcomes in GPA pts
- **METHODS:**
  - **35** GPA pts from FVSG (retrospective) with CNS involvement (no PNS and CN)
  - Treatment responses → **modified Rankin scale** (mRS; 0-5)
  - **2 phenotypes:**
    - **Granulomatous** = pachymeningitis / isolated granuloma / hypophyseal involvement
    - **Vasculitic** = Ischaemic or hemorrhagic lesions

Characteristic	All patients	GPA phenotype		P-value[a]
		G-CNS	V-CNS	
Number	35[b]	20[c]	13[c]	–
Demography				
Age, median (range), years				
At GPA diagnosis	48 (2–78)	49 (18–77)	48 (2–78)	0.91
At CNS onset	51 (2–79)	53 (26–78)	48 (2–79)	0.45
Male, *n* (%)	26 (74)	16 (80)	9 (69)	0.68
Follow-up, median (range), months	55 (8–192)	55 (19–151)	41 (8–192)	0.99
GPA manifestations, *n* (%)				
ENT	28 (80)	17 (85)	10 (77)	0.66
Pulmonary	20 (57)	11 (55)	7 (54)	1.00
Nodules	14 (70)	8 (73)	5 (71)	1.00
Alveolar haemorrhage	6 (30)	3 (27)	2 (29)	
Peripheral neuropathy	17 (49)	11 (55)	5 (38)	0.48
Necrotizing glomerulonephritis	14 (40)	4 (20)	9 (69)	**0.01**
Ocular	11 (31)	7 (35)	4 (31)	1.00
Joint	11 (31)	8 (40)	3 (23)	0.46
Cutaneous	8 (23)	4 (20)	4 (31)	0.68
Gastrointestinal	4 (11)	2 (10)	2 (15)	1.00
Granulomatous vasculitis	25 (71)	12 (60)	11 (85)	0.25
Biological findings, *n* (%)				
Positive ANCA	31 (89)	19 (95)	11 (85)	0.55
Anti-PR3 specificity	26 (84)	14 (74)	11 (100)	0.13
Anti-MPO specificity	5 (16)	5 (26)	0	

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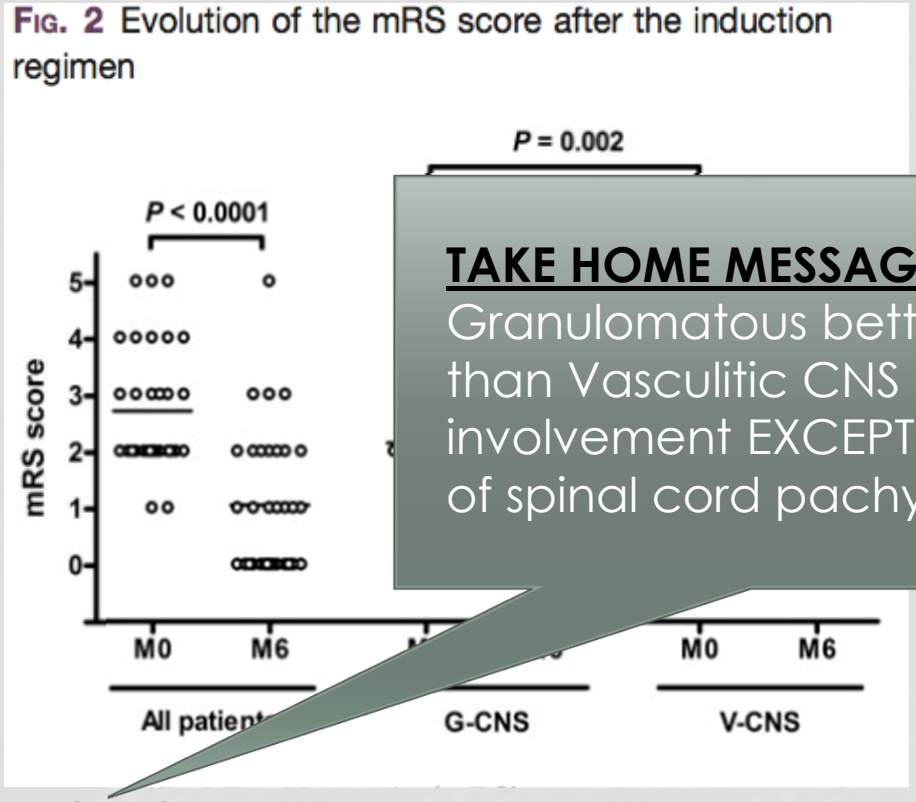
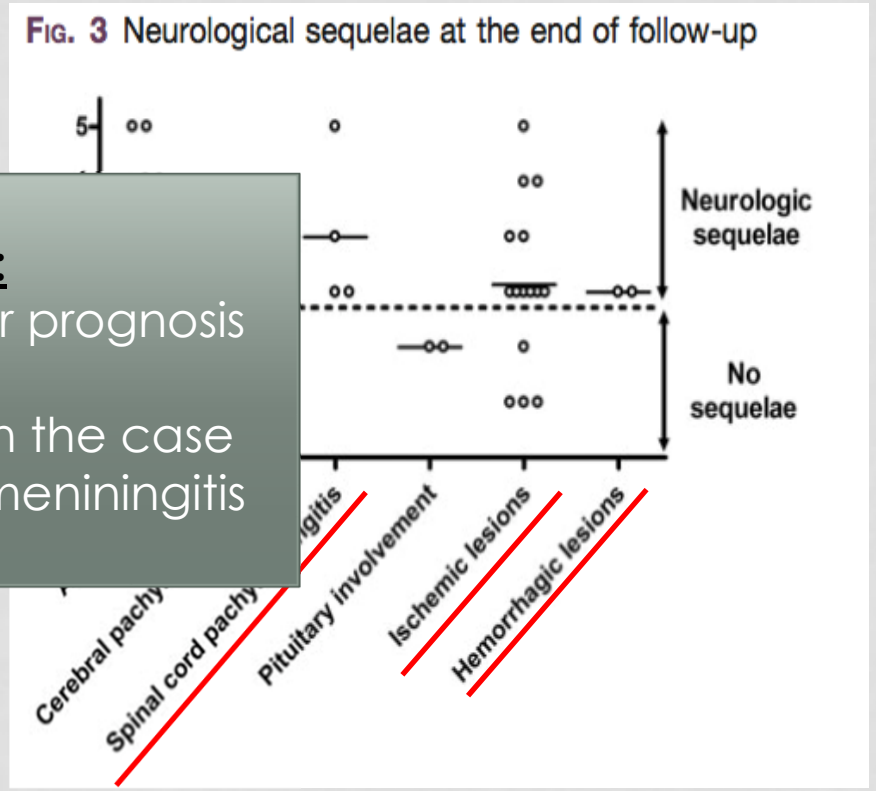
		G-CNS	V-CNS	
Number	35[b]	20[c]	13[c]	–
CNS symptoms, *n* (%)				
Headaches	23 (66)	19 (95)	4 (31)	**0.0002**
Sensory impairment	15 (43)	10 (50)	5 (38)	0.72
Motor impairment	11 (31)	1 (5)	9 (69)	**0.0002**
Vestibular syndrome	8 (23)	4 (20)	3 (23)	1
Hearing loss	8 (23)	7 (35)	1 (8)	0.11
Psychiatric/mood disorders	3 (9)	0	3 (23)	**0.05**
Diabetes insipidus	2 (6)	2 (10)	0	0.51
MRI findings, *n* (%)				
Cerebral pachymeningitis	16 (46)	16 (80)	0	–
Spinal cord pachymeningitis	4 (11)	3 (15)	0	–
Cerebral ischaemic lesions	15 (43)	0	13 (100)	–
Ischaemic stroke	9 (60)	0	8 (62)	
Extensive white matter lesions	6 (40)	0	5 (38)	
Cerebral haemorrhagic lesions	2 (6)	0	0	–
Brain and/or spinal cord vessel abnormalities	7 (20)	0	5 (38)	–
Pituitary gland enlargement with infundibular thickening	2 (6)	2 (10)	0	–
Granulomatous lesions, *n* (%)				
Brain	1 (3)	0	0	–
Spinal cord	1 (3)	0	0	–

CNS INVOLVEMENT OF GPA: CLINICAL -RADIOLOGICAL PRESENTATION DISTINGUISHES DIFFERENT OUTCOMES

DE LUNA G ET AL. RHEUMATOLOGY 2015;54





TAKE HOME MESSAGE:
Granulomatous better prognosis than Vasculitic CNS involvement EXCEPT in the case of spinal cord pachymeninigitis

- **RESULTS:**
  - Initial spinal pachymeningitis → ↑need for a new induction regimen for relapsing/refractory disease (P = 0.01)
  - **Long-term** neurological sequelae →**51**% pts =35% with G-CNS and 69% with V-CNS (P = 0.08)
  - Neurological sequelae → spinal pachymeningitis (100%) and ischaemic/haemorrhagic lesions (73%).

LONG-TERM OUTCOME OF AIRWAY STENOSIS IN GRANULOMATOSIS WITH POLYANGIITIS (WEGENER GRANULOMATOSIS)

AN OBSERVATIONAL STUDY

JAMA OTOLARYNGOL HEAD NECK SURG. 2014;140(11) MARTINEZ DEL PERO ET AL.

- **Aim:**
  - Frequency, lesion distribution and efficacy of intervention in airway stenosis in GPA pts
- **Methods:**
  - **44**/253 pts with airway stenosis
  - Main outcome measure = **airway patency for ≥12mo** + number of interventions required

Table 2. Characteristics of Patients With Airway Stenosis

Characteristic	Measured Value[a]
Women	32 (73)
Demography, median (IQR)	
Age at GPA diagnosis, y	30.1 (19.9-43.2)
Age at airway stenosis diagnosis, y	37.6 (26.6-46.8)
Disease duration, mo	146.1 (91.4-228.8)
Immunosuppressants tried, No.	4 (3-5)
CYC exposure, mg/kg	140 (76-199)
Treated with antibiotics	18 (41)
Distribution	
ENT system	43 (98)
Lungs	25 (73)
Kidneys	11 (34)
Other	16 (36)
Localized[b]	23 (52)
Early systemic[c]	10 (23)
Generalized[d]	10 (23)
Severe[e]	1 (2)
Serologic findings	
PR3-ANCA positive	27 (61)
MPO-ANCA positive	4 (9)
PR3 and MPO-ANCA positive	1 (2)
ANCA negative	11 (25)

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Figure 1. Distribution of Stenotic Lesions in the Airways of 44 Patients

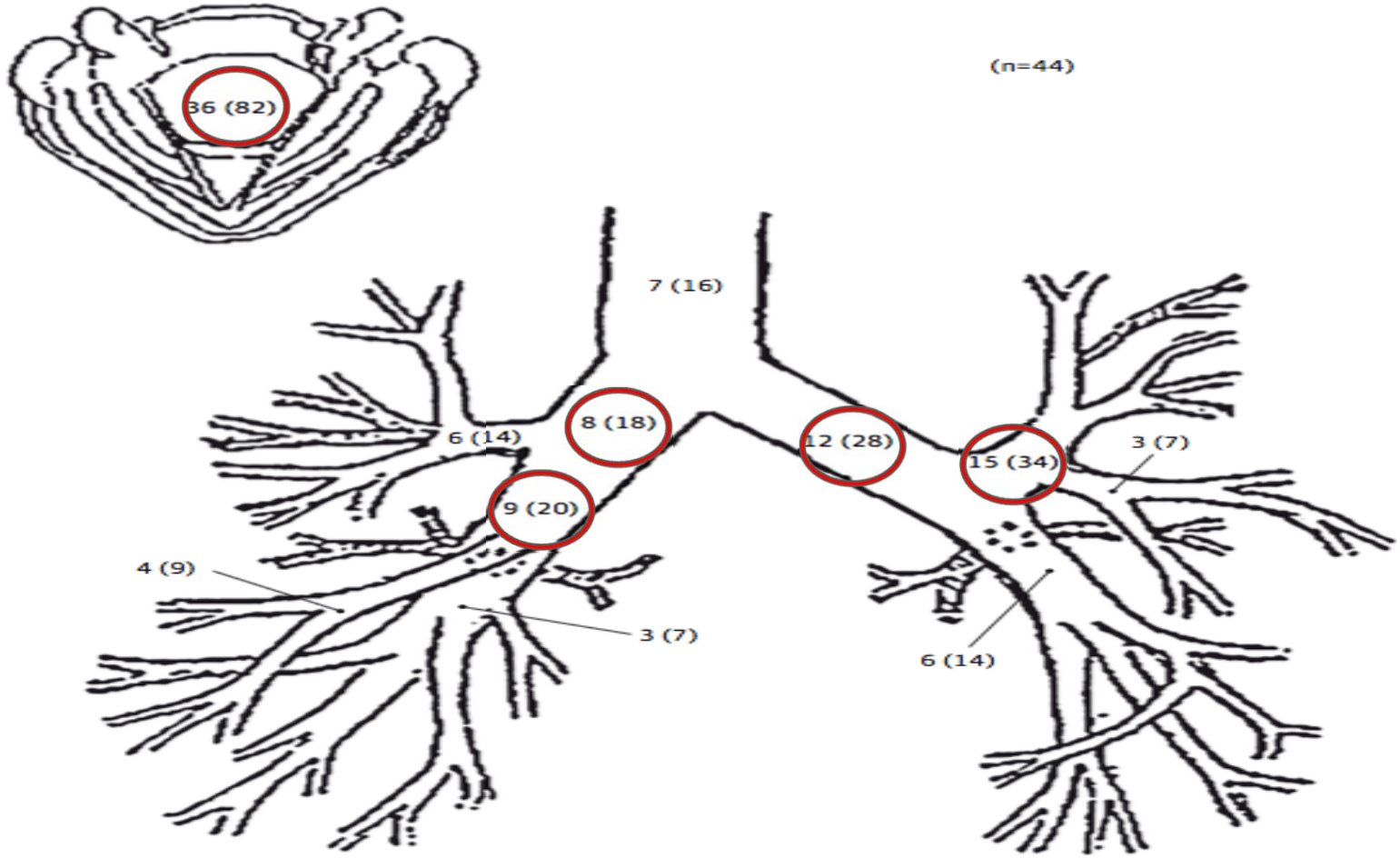

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Table 3. Characteristics of Procedures

Characteristic	Measured Value[a]
Continuous data, median No. (IQR)	
Lesions per patient	2 (1-4)
Interventions per patient	3 (1-8)
Interval between procedures (median-IQR), mo	4.9 (2.3-14.1)
Follow-up duration after last procedure, mo	27 (6.2-47.5)
Procedures during active endobronchial disease[b]	43 (20.1)
Procedures during lung infection[b]	66 (30.4)
Methods of dilatation (n = 213)[c,d]	
Balloon dilatation	130 (60.8)
Bougie dilatation	34 (15.9)
Laser dissection	24 (11.2)
Diathermy dissection	5 (2.34)
Argon-plasma coagulation	5 (2.34)
Cryotherapy	9 (4.21)
Adjuvant therapy	
Intralesional glucocorticoids	24 (9.3)
Topical mitomycin C	38 (14.7)
Intralesional alemtuzumab	9 (3.5)
None	142 (55.0)

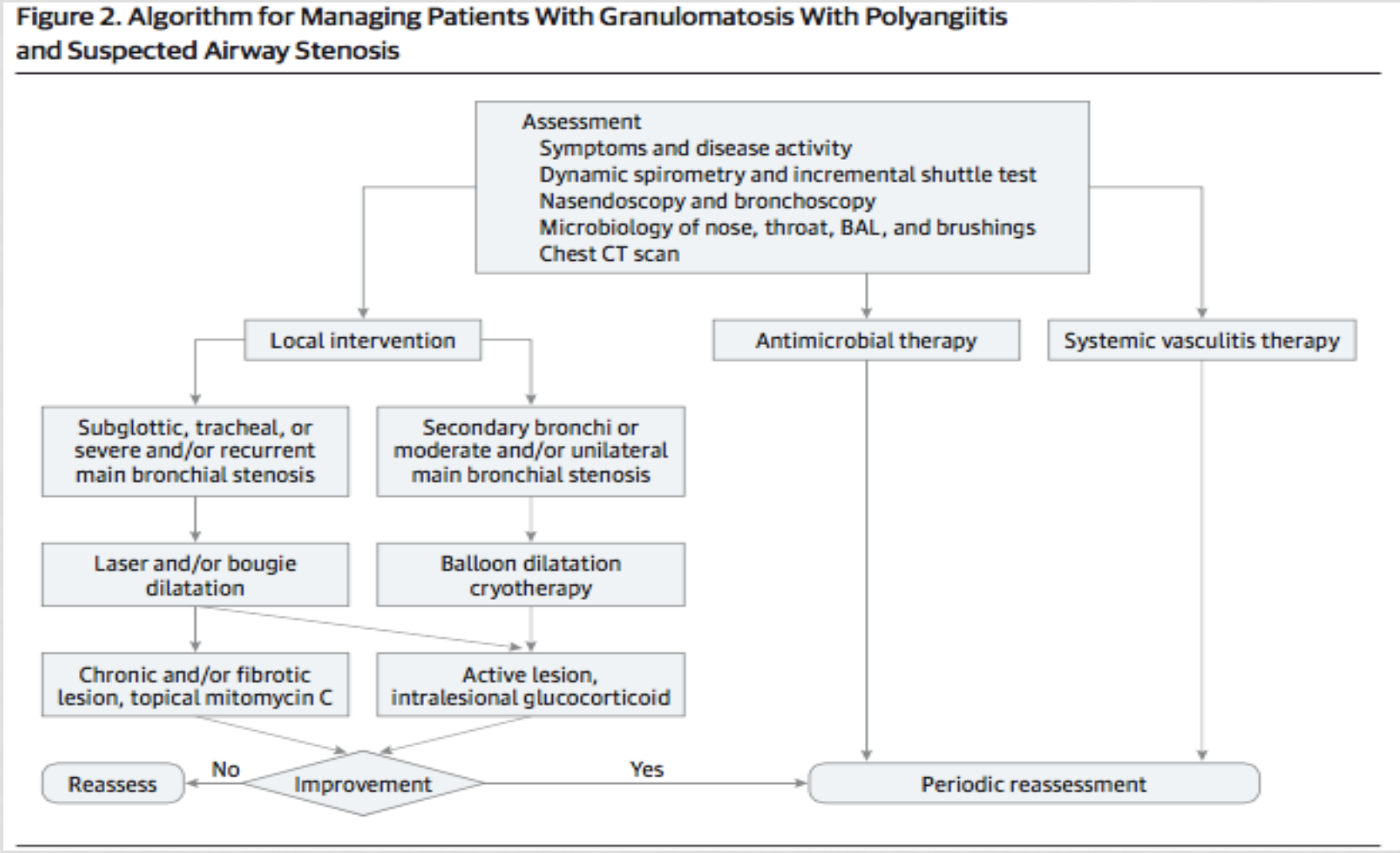
- ***97% (34/36 pts) achieved ≥ 12 mo airway stability period***
- ***Interval between procedures = 4.9 mo →After last intervention →≥27 mo of airway stability***
- ***Broncial distribution associated with more refractory course***
- ***Adverse events = 6.6%*** *(5x Infection, 2x bleeding, 2x perforation,2x stent problems,1x polyp, 1x death from active GPA)*

LONG-TERM OUTCOME OF AIRWAY STENOSIS IN GRANULOMATOSIS WITH POLYANGIITIS (WEGENER GRANULOMATOSIS)

AN OBSERVATIONAL STUDY

JAMA OTOLARYNGOL HEAD NECK SURG. 2014;140(11) MARTINEZ DEL PERO ET AL.

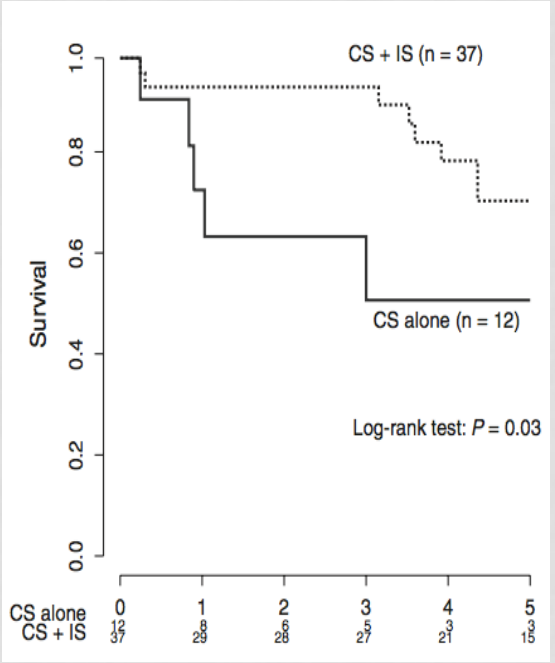
Figure 2. Algorithm for Managing Patients With Granulomatosis With Polyangiitis and Suspected Airway Stenosis



PULMONARY FIBROSIS IN AAV
A SERIES OF 49 PATIENTS AND REVIEW OF THE LITERATURE

COMARMOND CL ET AL — MEDICINE 2014;93: 340–349

- **AIM**: Pulmonary fibrosis in AAV → features and prognosis
- **METHODS:** 49 pts (men [61%]; median age at diagnosis of AAV, 68) with PF associated with AAV were identified.
- **RESULTS:**
- **81.6% = MPA** vs 18.4% = GPA
- anti-MPO = 88%
- Diagnosis of PF **preceded the onset** of vasculitis in 22 (**45%**) pts.
- Main radiologic pattern = **UIP** (43%).
- Px: GC + CY (73.5%) or RTX (2%)
- **Mortality** associated with:
  - **chronic respiratory insufficiency** (HR = 7.44; p = 0.003),
  - Induction with **GC alone** (HR = 2.94; p = 0.04), and
  - **initial weigh loss** (HR= 2.83; p = 0.041).
- **3-yr survival rate** in patients treated with GC alone = 64% vs combination with immunosuppressant **(CYC or RTX) = 94%** (p = 0.03).
- (**37%) pts died** (11 →respiratory insufficiency) after a median follow-up of 48 mo



- **CONCLUSION:** PF is a rare manifestation of AAV with a very poor prognosis. Induction therapy with CYC might improve the outcome.

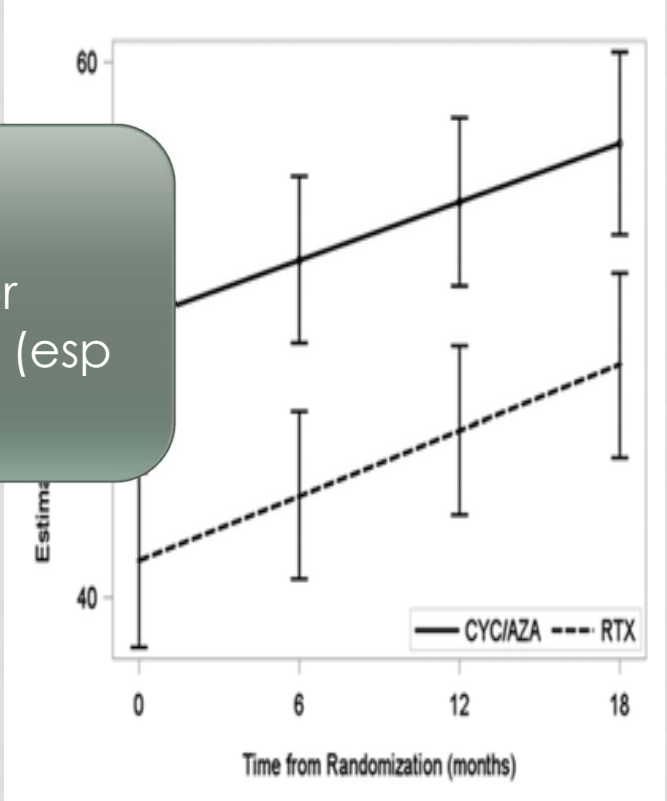
RITUXIMAB VERSUS CYCLOPHOSPHAMIDE FOR ANCA- ASSOCIATED VASCULITIS WITH RENAL INVOLVEMENT

GEETHA D ET AL — J AM SOC NEPHROL 2014; 26

- **AIM:** RTX equivalent to CYC also in renal disease?
- **METHODS:**
  - 102/197 (RA
    (Cr<4) at e
    51 to CYC/
  - Mean eGF
    ml/min pe
- **RESULTS:**
  - CR at 6mo →61% RTX vs 63% CYC/AZA
  - CR at 18mo → 75% RTX vs of 76% CYC/AZA
  - No differences in adverse events

Take home message:

RTX non inferior to CYC/AZA for induction in renal involvement (esp in pts with Cr<4)



PLASMA EXCHANGES FOR THE TREATMENT OF SEVERE SYSTEMIC NECROTIZING VASCULITIDES IN CLINICAL DAILY PRACTICE: DATA FROM THE FRENCH VASCULITIS STUDY GROUP

DE LUNA G ET AL. JOURNAL OF AUTOIMMUNITY XXX (2015) 1-7

- **AIM:** Indications, efficacy and safety of PLEX for the treatment of SNV (AAV & non-viral PAN)

- **METHODS:**
  - Retrospective study with 152 pts: GPA = 87, MPA = 56, EGPA =4 and PAN = 5

  - PLEX used for:
    - RPGN in 126 cases (86%),

    - Alveolar hemorrhage in 64 cases (42%), and

    - Severe mononeuritis multiplex in 23 cases (15%).

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DE LUNA G ET AL. JOURNAL OF AUTOIMMUNITY XXX (2015) 1-7

- **RESULTS:**

1. **Alveolar hemorrhage**: D/C of mechanical ventilation in all patients after a median time of 15 days.

2. **Mononeuritis multiplex** → showed improvement of severe ***motor weakness***(no effect on sensory symptoms): 52% →23% →19% →12.5% at baseline, 3, 6 and 12 mo after PLEX onset

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DE LUNA G ET AL. JOURNAL OF AUTOIMMUNITY XXX (2015) 1-7

- **RESULTS:**

3. **RPGN:**
   - significant improvement in renal function compared to baseline value ($P < 0.0001$),
   - plateau reached at month 3 after PLEX initiation,
   - eGFR improved especially as the number of PLEX increased (in contrast to Welsh's meta-analysis).

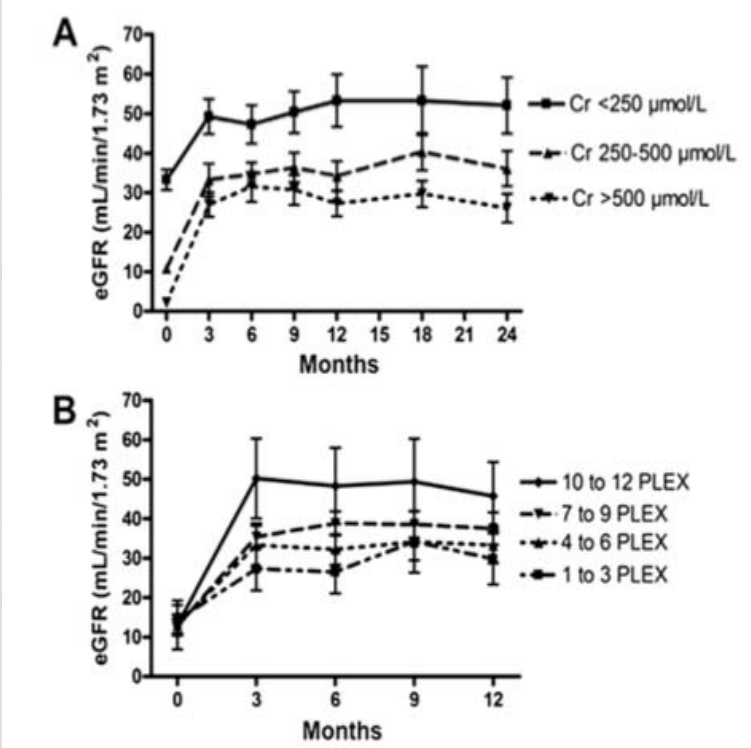

Fig. 1. Evolution of estimated glomerular filtration rate according to baseline serum creatinine level (**A**) and the number of plasma exchanges (**B**).

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DE LUNA G ET AL. JOURNAL OF AUTOIMMUNITY XXX (2015) 1-7

- **RESULTS:**
  - **Follow-** **(median) →18 deaths**
  - **Adverse events** attributable to PLEX→ 95 pts (**63%**) (incl. anemia in 66 (43%) infections in 20 (13%) etc).
  - No death during PLEX.

Take home messages:

- **Initial response in all indications**
- **More PLEX → more eGFR improvement**
- **eGFR improvement → plateau at 3mo**

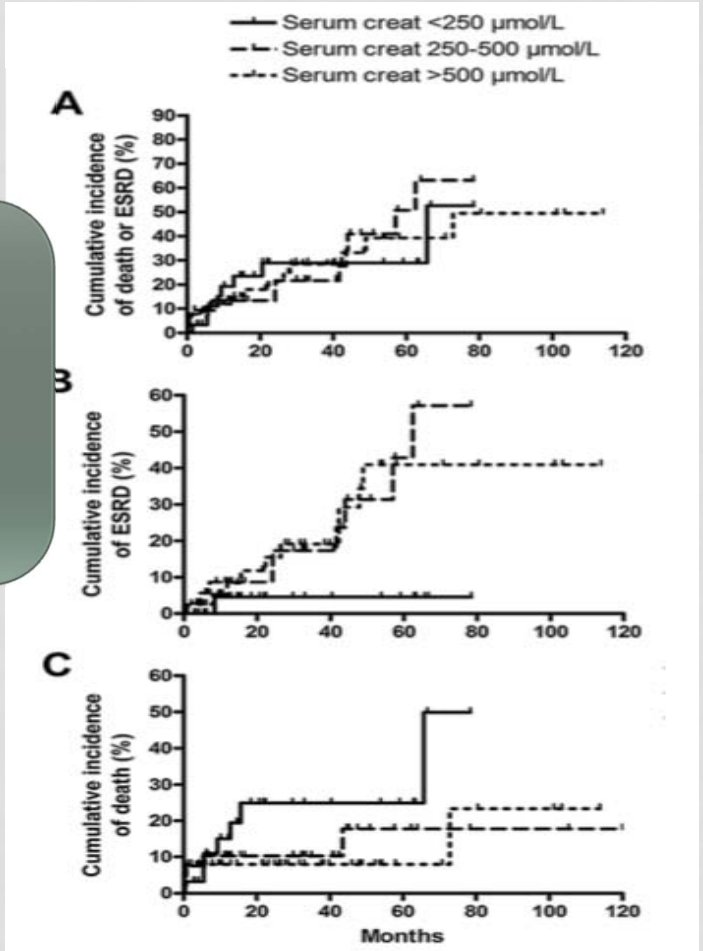

Fig. 2. Cumulative incidence of the composite outcome, i.e. death or ESRD (**A**), or ESRD (**B**) or death (**C**), in patients with rapidly progressive renal failure treated with plasma exchanges.

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BEHCET DISEASE WITH VASCULAR INVOLVEMENT
EFFECTS OF DIFFERENT THERAPEUTIC REGIMENS ON THE INCIDENCE OF NEW RELAPSES

ALIBAZ-ONER ET AL MEDICINE □ 2015;94

(6)

- **AIM:** Treatment modalities of Vascular Behcet disease and association with relapses
- **METHODS:**
  - Retrospective data of 936 pts with BD → 260 with vascular BD
  - Relapse rate and association with clinical features and therapeutic strategies

TABLE 1. Clinical Characteristics of Vascular Behçet Disease (n = 260)

Gender	
Male (n = 224)	86.2%
Age during first vascular event (years)	32.3 ± 9.5
Only venous disease (n = 220)	84.6%
Only arterial disease (n = 21)	8.1%
Both venous and arterial disease (n = 11)	4.2%
Cardiac involvement (n = 8)	3.1%
Rare vascular involvements	
Budd–Chiari syndrome (n = 3)	1.2%
Pulmonary aneurysm (n = 29)	11.2%
Pulmonary thrombosis (n = 7)	2.7%
Vena cava superior or inferior involvement (n = 22)	8.5%
Second vascular event (n = 86)	32.9%
Third vascular event (n = 17)	6.5%
Fourth vascular event (n = 3)	1.1%

BEHCET DISEASE WITH VASCULAR INVOLVEMENT
EFFECTS OF DIFFERENT THERAPEUTIC REGIMENS ON THE INCIDENCE OF NEW RELAP

ALIBAZ-ONER ET AL

15;94

Take home messages:
1) No added benefit of AC in VBD
2) In pts with VBD do NOT withdraw ISs early (<2 yrs)

1st

- *Presenting sign in **57.3%***
- Initial Px: IS = 88.8% vs AC =

2nd

- **32.9%** (n = 86)
- time interval between 1st and 2nd event = **25.5** (1–252) **mo**
- relapse **lower in pts taking ISs** (25.3% vs 85.7%, P<0.001) Only **50% -60% on IS** before relapses
- relapse rate **ISs = AC+ IS** (29.1% vs 22.4%, P = 0.28) BUT only ACs >>> only ISs (91.6% vs 29.1%, P<0.001).

3rd

- **6.5%**(n =17)
- relapse rate **ISs = AC + IS** (25.3% vs 20.8%, P = 0.93).

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EFFECTIVENESS AND SAFETY OF MEDIUM-DOSE PREDNISONE IN GIANT CELL ARTERITIS: A RETROSPECTIVE COHORT STUDY OF 103 PATIENTS

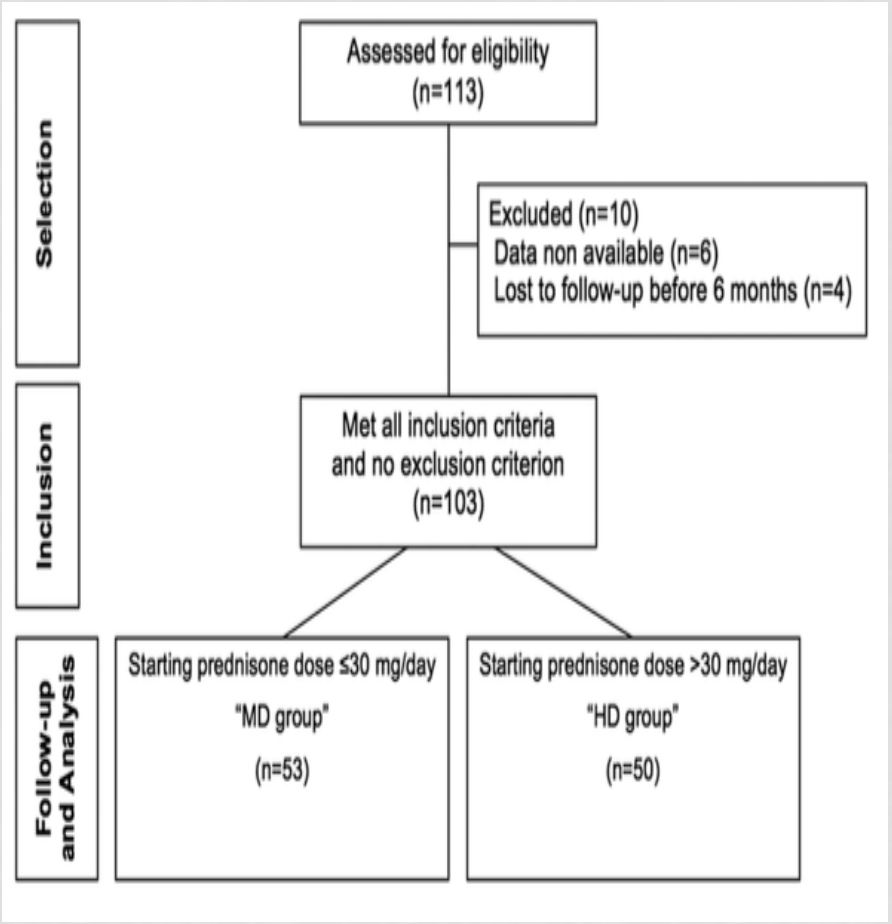
I.LES ET AL

CLIN EXP RHEUMATOL 2015; 33

- **AIM:** To compare the effectiveness and safety of medium-dose (MD) and high-dose (HD) prednisone regimens in patients with GCA

- **METHODS:**
  - Retrospective cohort study → 2 groups: a) Medium GC ≤30 mg (MD group) or b) >30 mg (HD group) (monotherapy or combined with methylprednisolone pulses and/or methotrexate) followed for 2.85 (2.57–3.52) yrs

  - Primary endpoint = time to clinical and biological remission receiving a prednisone maintenance dose ≤7.5 mg/day.



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I.LES ET AL

CLIN EXP RHEUMATOL 2015; 33

- **RESULTS:**

Take home messages:

1) MD valid alternative to HD + safer
2) Iv MP pulses has substantial impact on treatment efficacy

Therefore → iv MP + MD = better risk/benefit ?

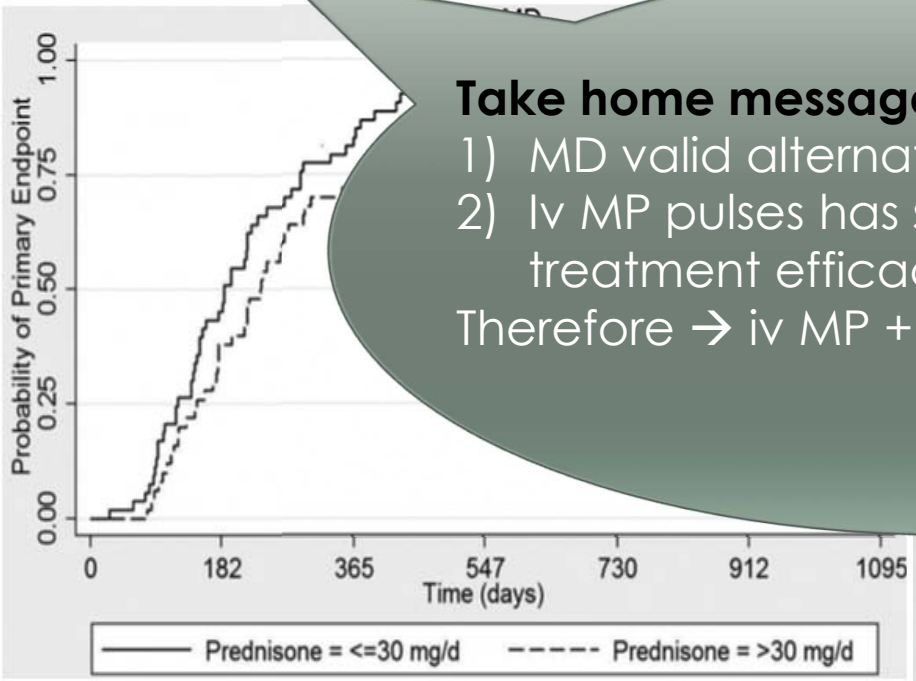

Fig. 2. Kaplan-Meier probability of achieving clinical and biological remission while receiving ≤7.5 mg/day of prednisone in MD and HD groups. The needed time to achieve the primary endpoint was shorter in MD group patients (MD=186 [147-223], HD=236 [177-276] days, log-rank test p=0.01). MD: medium-dose; HD: high-dose.

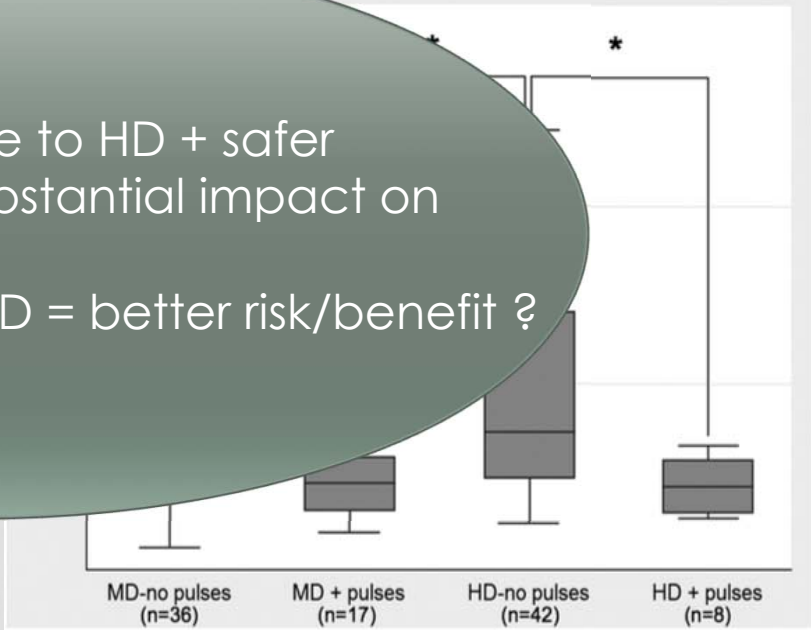

Fig. 3. Box plot comparing time to the primary endpoint between MD and HD groups with or without MP pulse therapy. The time to the primary endpoint in days for the MD without pulses subgroup was 262.5 (93-426); for the MD with pulses subgroup, 164.5 (103-213); for the HD without pulses subgroup, 263 (177-550); and for the HD with pulses subgroup, 176.5 (88-238).
*p=0.001 (Bonferroni-adjusted significance threshold p=0.008).
MD: medium-dose; HD: high-dose; MP: methylprednisolone.

Time to 1^{ary} Endpoint MD better than HD

Time to 1^{ary} Endpoint better with iv MP

Less Adverse events with MD vs HD (p=0.02)

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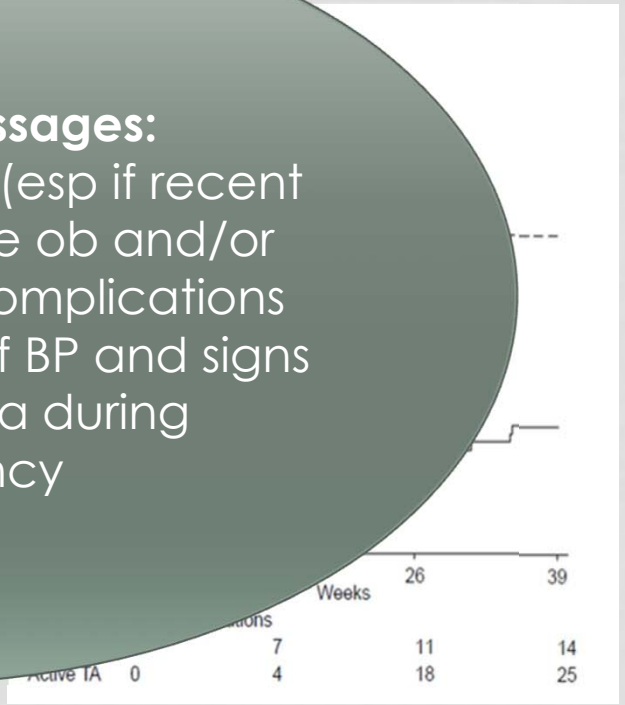
TAKAYASU'S ARTERITIS AND PREGNANCY

COMARMOND C ET AL — ARTHRITIS RHEUMATOL 2015 AUG

- **AIM:** Takayasu's arteritis (TA) and … outcomes?
- **METHODS:**
  - 240 pregnancies in …
  - 142 pregnancies in … pregnancies in 52 … diagnosis
- **RESULTS:**
  - Obstetrical compli… pregnancies **concom…** before diagnosis.
  - Obstetrical complications … **24%, prematurity (8%) and in… restriction or death (5%).**
  - Specific TA complications during pregnanc… and include **mainly new onset or worsening hypertension** [n=26, (26%)].
  - In multivariate analysis obstetrical and maternal complications associated with:
    - **Smoker** (OR=6.15) and
    - **Disease activity of TA** (i.e. NIH score >1) (OR=28.7)

Take home messages:

1. 40% of TA patients (esp if recent dx or active) have ob and/or disease-related complications
2. Close monitoring of BP and signs of preclampsia during pregnancy



EFFICACY OF BIOLOGICAL-TARGETED TREATMENTS IN TAKAYASU ARTERITIS: MULTICENTER RETROSPECTIVE STUDY OF 49 PATIENTS

MEKINIAN ET AL. CIRCULATION 2015 SEP

- **AIM:** Safety and efficacy of biologics in TA

- **METHODS:**
- 49 TA patie
- TNF-i (80%
- Active if
aging, systemic symptoms
- CR= NIH<2 +
n PRE)

Take home Message:

TNFi and TCZ equally effective and safe

- **RESULTS:**
- Overall response at 6 = 75% and 12 mo = 83% (+ ↓ PRE dose/CRP)
- 3-yr relapse-free = 91% with bDMARDS vs 59% for csDMARDs
- Similar efficacy between TNFi and TCZ
- In 2 yr f/u → AEs = 21% → discontinuation = 6.6%

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AN UPDATE OF THE MAYO CLINIC COHORT OF PATIENTS WITH ADULT PRIMARY CENTRAL NERVOUS SYSTEM VASCULITIS

DESCRIPTION OF 163 PATIENTS

SALVARANI ET AL, MEDICINE 94 (21); 2015

AIM: Epidemiology, clinical features, prognosis and treatment of PCNSV

Methods: 163 pts (1983-2011)→ 105 angiographic + 58 Bx

TABLE 1. Clinical Manifestations at Presentation in 163 Consecutive Patients With PCNSV

Findings	All Patients (n = 163), n (%)	Biopsy Confirmed (n = 58), n (%)	Angiogram Confirmed (n = 105), n (%)
Headache	97 (59.5)	31 (53.4)	66 (62.9)
Cognitive dysfunction	88 (54)	39 (67.2)	49 (46.7)*
Hemiparesis	66 (40.5)	10 (17.2)	56 (53.3)*
Persistent neurologic deficit or stroke	66 (40.5)	13 (22.4)	53 (50.5)*
Aphasia	40 (24.5)	15 (25.9)	25 (23.8)
Transient ischemic attack	42 (25.8)	9 (15.5)	33 (31.4)*
Ataxia	31 (19)	6 (10.3)	25 (23.8)
Seizures	33 (20.2)	16 (27.6)	17 (16.2)
Visual symptoms (any kind)	61 (37.4)	14 (24.1)	47 (44.8)
Visual field defect	30 (18.4)	5 (8.6)	25 (23.8)*
Diplopia (persistent or transient)	23 (14)	7 (12.1)	16 (15.2)
Blurred vision or decreased visual acuity	18 (11)	3 (5.2)	15 (14.3)
Monocular visual symptoms or amaurosis fugax	2 (1.2)	1 (1.7)	1 (1)
Papilledema	7 (4.3)	4 (6.9)	3 (2.9)
Intracranial hemorrhage	16 (9.8)	5 (8.6)	11 (10.5)
Amnestic syndrome	10 (6.1)	5 (8.6)	5 (4.8)
Paraparesis or quadriparesis	8 (4.9)	5 (8.6)	3 (2.9)
Parkinsonism or extrapyramidal signs	1 (0.6)	0	1 (1)
Constitutional symptoms†	15 (9.2)	6 (10.3)	9 (8.6)
Fever	16 (9.8)	8 (13.8)	8 (7.6)

AN UPDATE OF THE MAYO CLINIC COHORT OF PATIENTS WITH ADULT PRIMARY CENTRAL NERVOUS SYSTEM VASCULITIS

DESCRIPTION OF 163 PATIENTS

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TABLE 3. CSF Findings

	All Patients (n = 126), n (%)
Red blood cell count >0/mL, number of patients/total	96/116 (82.8)
Increased total protein concentration, leukocyte count, or red blood cell count, number of patient/total	114/123 (92.7)
Protein >45 mg/dL, or leukocyte >5 cells/mL, number of patients/total	100/123 (81.3)
Total protein concentration >70 mg/dL, number of patients/total	63/121 (52.1)
Protein >70 mg/dL, or leukocyte >10 cells/mL, number of patients/total	77/121 (63.6)

TABLE 4. Characteristics of 113 Positive Cerebral Angiograms

	All Patients* (N = 113), n (%)
Bilateral vasculitis	108 (95.6)
Large-vessel changes consistent with vasculitis	
Total	75 (66.4)
Unilateral	13 (11.5)
Bilateral	62 (54.9)
Small vessel changes consistent with vasculitis	
Total	103 (91.2)
Unilateral	10 (8.8)
Bilateral	93 (82.3)

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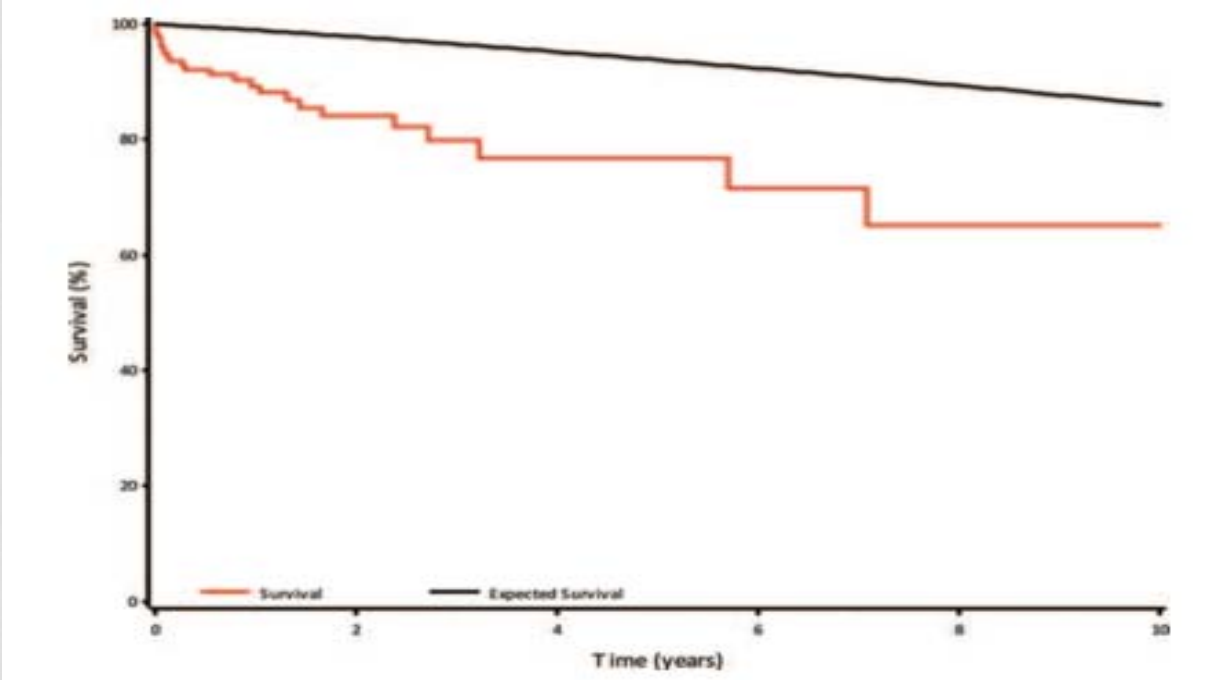
- **Annual Incidence rate = 2.4 cases per 10^6 pt/yrs**



- **Increased age and infarct on initial MRI → increased disability at follow-up**

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