Θρομβωτική

Θρομβοπενική

Πορφύρα

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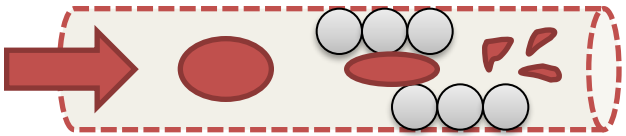
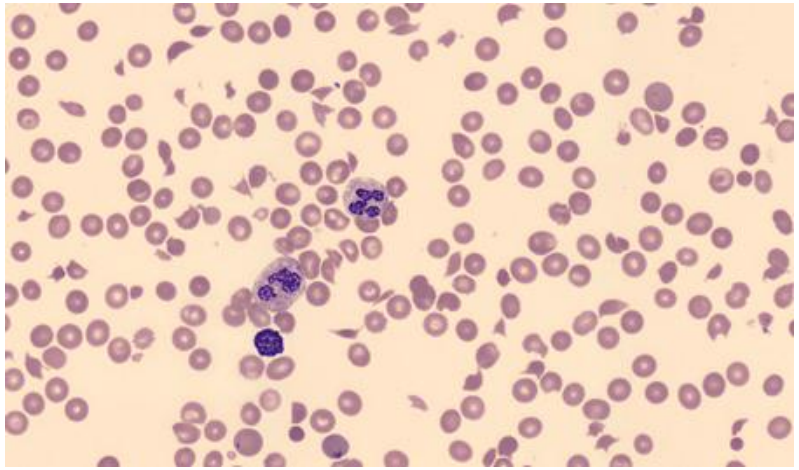
Επίκουρη Καθηγήτρια Αιματολόγιας,

Β Προπαιδευτική Παθολογική Κλινική, ΑΠΘ

Θρομβωτική Μικροαγγειοπάθεια

κλασσική κλινική τριάδα





1. Μικροαγγειοπαθητική (σχιστοκύτταρα)
Αιμολυτική (↑ LDH)
Αναιμία

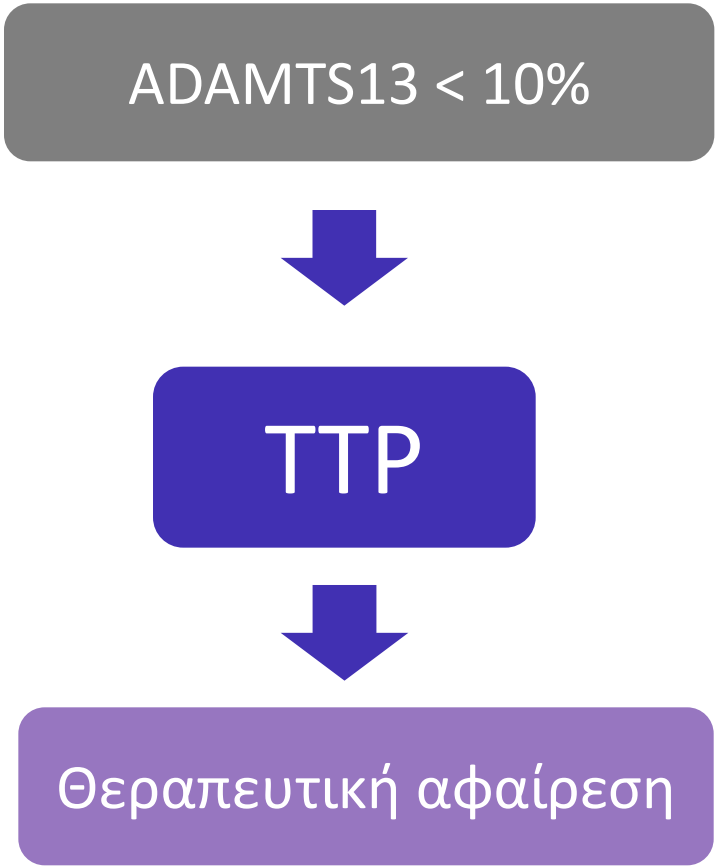
2. Θρομβοπενία
3. Οξεία βλάβη οργάνων (νεφρός, ΚΝΣ, κ.ά.)

Θρομβωτικές μικροαγγειοπάθειες

TTP

Θρομβωτική μικροαγγειοπάθεια

διαφορική διάγνωση βάσει παθοφυσιολογίας



Θρομβωτική θρομβοπενική πορφύρα

ιστορικά στοιχεία

- **1946:** 12 ασθενείς (+ ασθενής Moschkowitz από 1924)

Singer K, et al. Blood 1946

- **1960-80:** αναφορές διάσωσης με συνεδρίες πλασμαφαίρεσης (ασθενείς με κληρονομική διαταραχή)

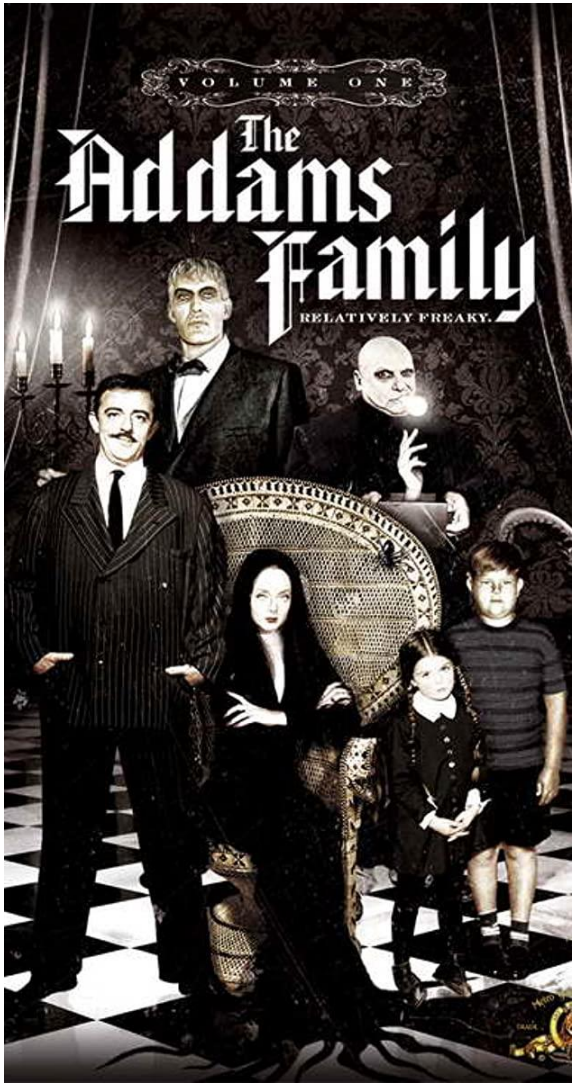
Upshaw JD. NEJM 1978
Schulman I, et al. Blood 1960;

- **1982:** ασυνήθιστα μεγάλα πολυμερή von Willebrand

Moake JL, et al. NEJM. 1982

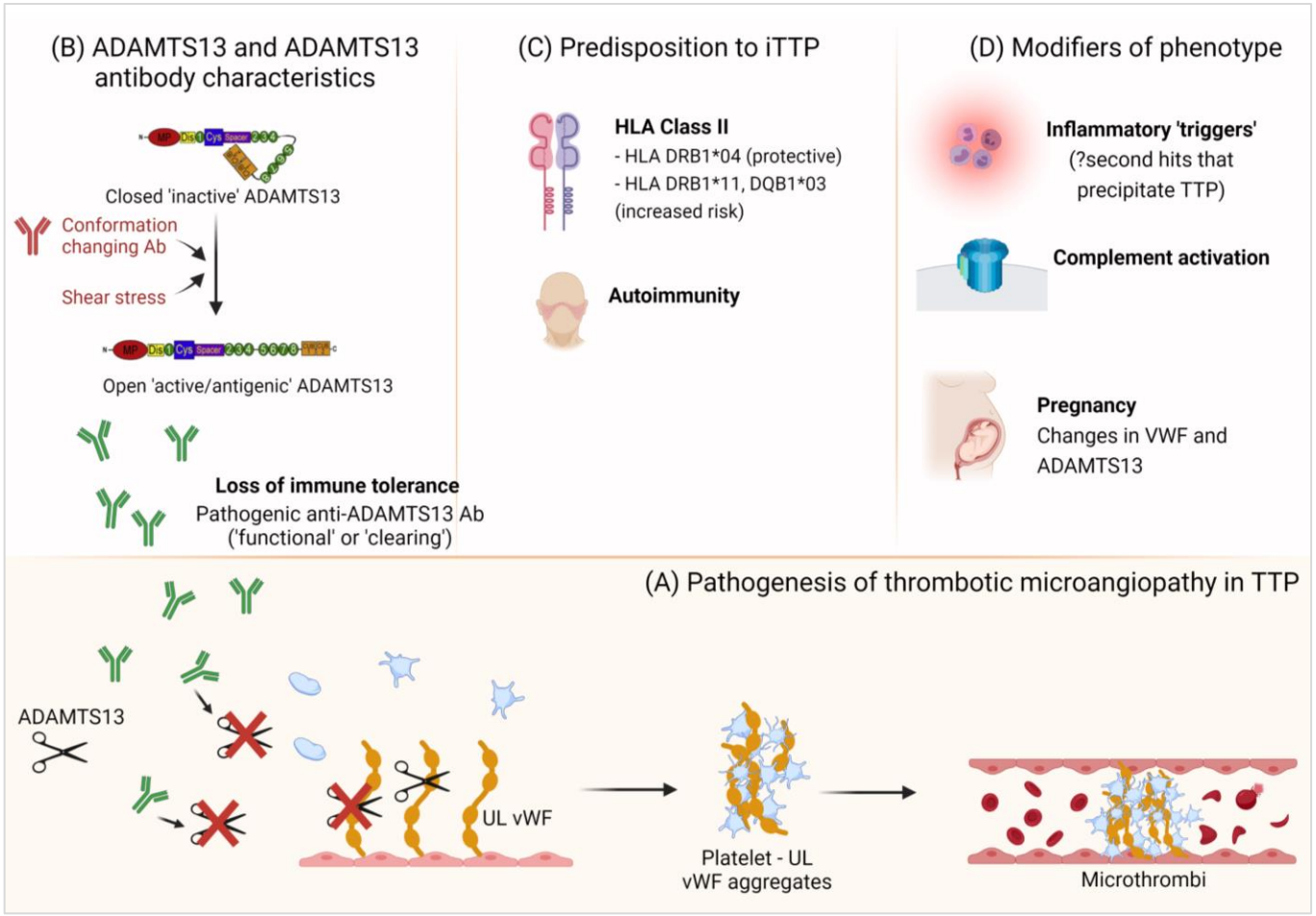
- **1996:** υπεύθυνη μεταλλοπρωτεάση = ADAMTS13

Levy GG, et al. Nature. 2001


VOLUME ONE
The Addams Family
RELATIVELY FREAKY.

Θρομβωτική θρομβοπενική πορφύρα

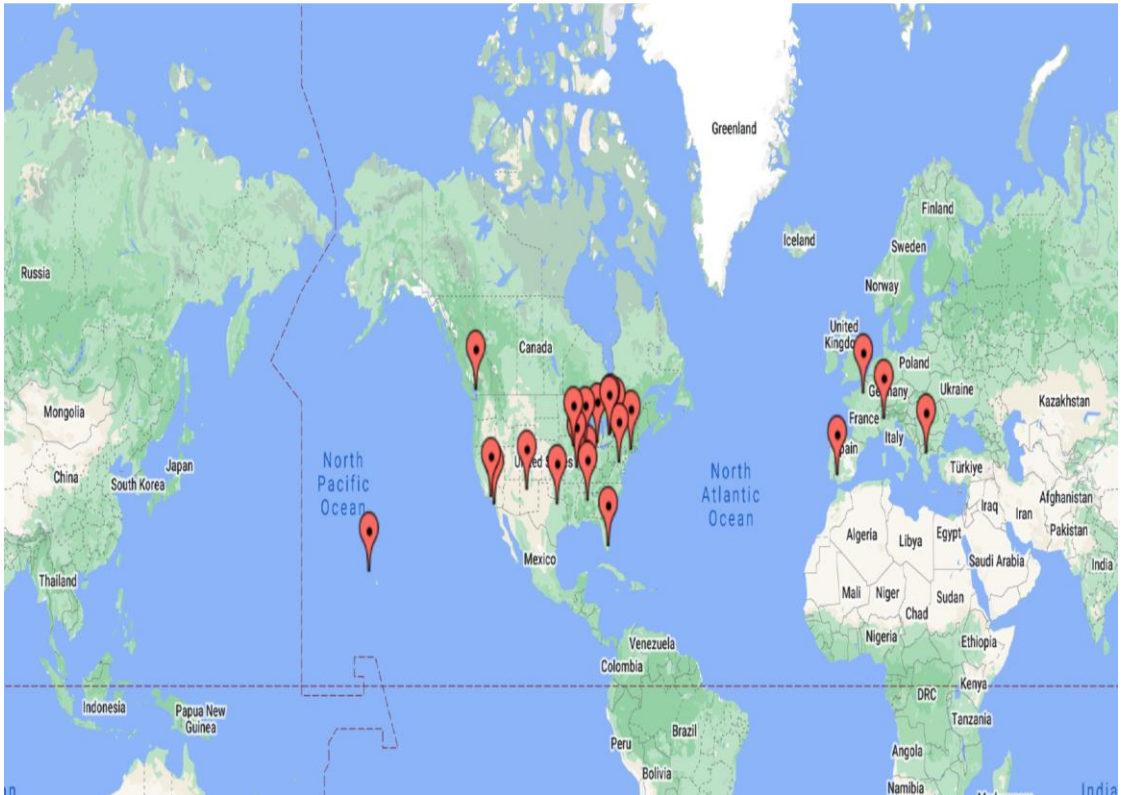
νεότερα δεδομένα στην παθοφυσιολογία

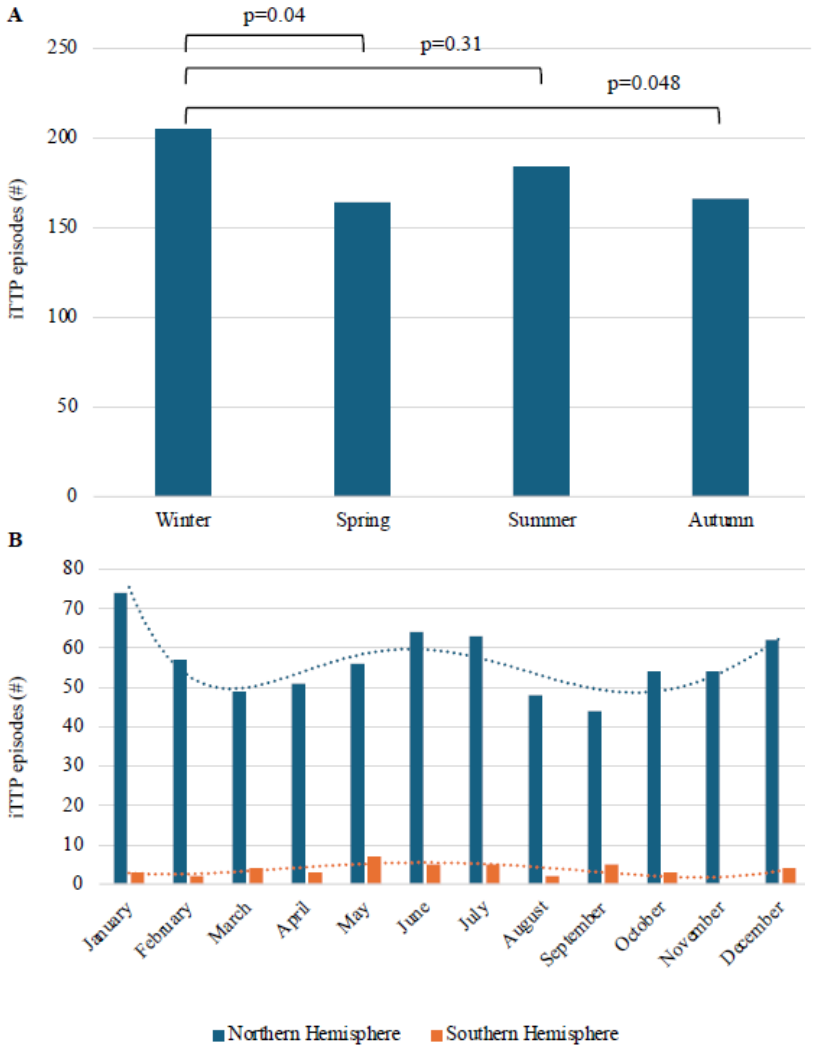


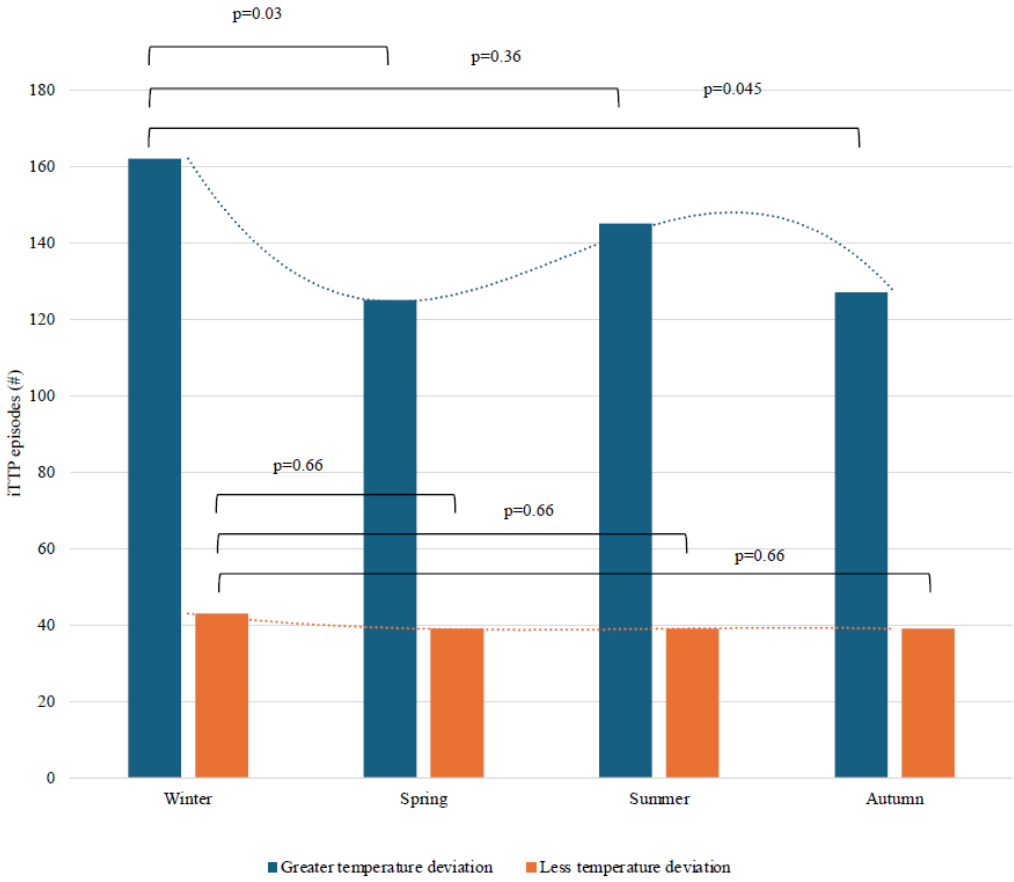
Sukumar S, Gavriilaki E, Chaturvedi S. Thrombosis Update 2021

Immune-Mediated Thrombotic Thrombocytopenic Purpura Seasonality: An International Analysis

Authors: Jeremy W. Jacobs, MD, MHS,[1] Caroline G. Stanek, MD,[2] Garrett S. Booth, MD, MS,[3] Argiris Symeonidis, MD, PhD,[4,5] Andrew W. Shih, MD, MSc,[6,7] Elizabeth S. Allen, MD,[8] Eleni Gavriilaki, MD, PhD,[9] Brenda J. Grossman, MD,[10] Katerina Pavenski, MD,[11,12] Amy Moorehead, MS,[11] Flora Peyvandi, MD, PhD,[13,14] Pasquale Agosti, MD, PhD,[13,14], Ilaria Mancini, PhD,[13] Laura D. Stephens, MD,[8] Jay S. Raval, MD,[15,16] Maria-Eva Mingot-Castellano, MD, PhD,[17] Elizabeth P. Crowe, MD, PhD,[18] Laetitia Daou, MD,[18] Menaka Pai, MD, MSc,[19,20] Donald M. Arnold, MD, MSc,[20] Marisa B. Marques, MD,[2] Ryan Henrie, MD,[21] Tyler W. Smith, MD, MSc,[6,7] Gayatri Sreenivasan, MD,[21] Rance C. Siniard, MD,[2] Lisa R. Wallace, DO,[10] Chisa Yamada, MD,[22] Miriam Andrea Duque, MD,[23] Yanyun Wu, MD, PhD,[23] Thomas J. Harrington, MD,[24] Diana M. Byrnes, MD,[24] Aikaterini Bitsani, MD, PhD, MSc,[25] Amanda K. Davis, MD,[26] Danielle H. Robinson, MD,[26] Quentin Eichbaum, MD, PhD, MPH,[3] Cristina A. Figueroa Villalba, MD,[27] Justin E. Juskewitch, MD, PhD,[1] Georgia Kaiafa, MD, PhD,[28] Eleni Kapsali, MD, PhD,[29] Ellen Klapper, MD,[30] Ingrid Perez-Alvarez, MD,[30] Monica S. Klein, MD,[1] Nikolaos Kotsiou, MD,[31] Chrysavgi Lalayanni, MD,[32] Evdokia Mandala, MD,[33] Fatima Aldarweesh, MD,[34] Rahaf Alkhateb, MD,[34] Zois Mellios, MD, MS,[35] Apostolia Papalexandri, MD,[32] Meredith G. Parsons, MD,[36] Annette J. Schlueter, MD,[36] Christopher A. Tormey, MD,[27] Cameron Wellard, PhD,[37], Erica M. Wood, MBBS,[37,38] Shiyang Jia, PhD,[37] Allison P. Wheeler, MD, MSCI,[3,39] Amy A. Powers, MD,[40] Christopher B. Webb, MD,[41] Sean G. Yates, MD,[41] Raïda Bouzid,[42] Paul Coppo, MD, PhD,[42,43] Evan M. Bloch, MBChB, MS,[18] Brian D. Adkins, MD[41]




A
p=0.04
p=0.31
p=0.048
iTTP episodes (#)
250
200
150
100
50
0
Winter
Spring
Summer
Autumn
B
iTTP episodes (#)
80
70
60
50
40
30
20
10
0
January
February
March
April
May
June
July
August
September
October
November
December
Northern Hemisphere
Southern Hemisphere


p=0.03
p=0.36
p=0.045
p=0.66
p=0.66
p=0.66
iTTP episodes (#)
180
160
140
120
100
80
60
40
20
0
Winter
Spring
Summer
Autumn
Greater temperature deviation
Less temperature deviation

ADAMTS13

Πότε τη μετράμε

- **Κλινική τριάδα θρομβωτικής μικροαγγειοπάθειας**
- Διάγνωση
- Τέλος θεραπείας
- Υποτροπή
- Κατά την ύφεση? ISTH Guidelines

ISTH Guidelines 2020

ADAMTS13

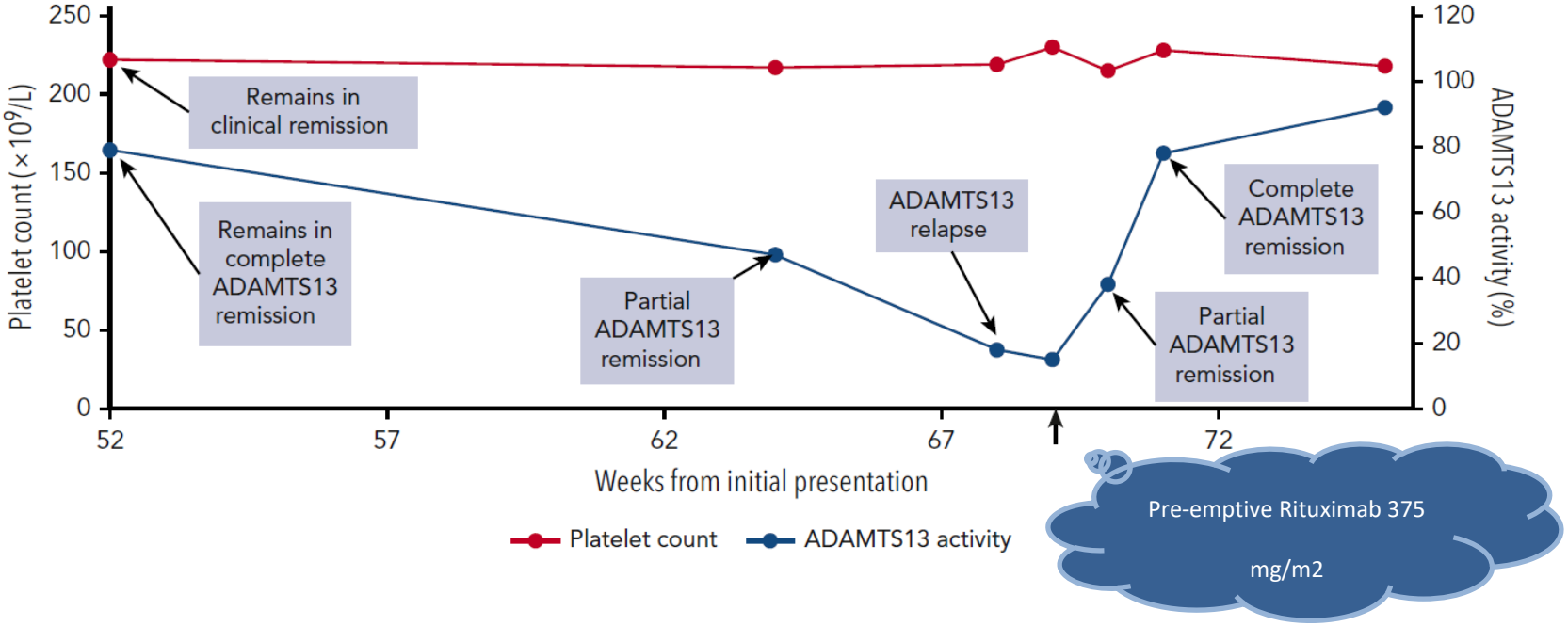
Αναθεώρηση ορισμών για την TTP

Category	Outcome	Definition	Management implications
Response	Clinical response	Sustained platelet count ≥150 × 10^9/L and LDH <1.5 times ULN and no clinical evidence of new or progressive ischemic organ injury.	Preremission implications In general, TPE may be discontinued and patients may be discharged from the hospital soon after they achieve a clinical response. Persistent severe ADAMTS13 deficiency after a clinical response is associated with an increased risk of clinical exacerbation. Immunosuppression (e.g., corticosteroids, rituximab) may be used to induce an ADAMTS13 remission. Use of anti-VWF therapy (e.g., caplacizumab) until attainment of ADAMTS13 remission is protective against clinical exacerbation.
Exacerbation	Clinical exacerbation	After a clinical response and before a clinical remission, platelet count decreases to <150 × 10^9/L (with other causes of thrombocytopenia excluded), with or without clinical evidence of new or progressive ischemic organ injury, within 30 d of stopping TPE or anti-VWF therapy	
Remission	Clinical remission	Sustained clinical response with either no TPE and no anti-VWF therapy for ≥30 d or with attainment of ADAMTS13 remission (partial or complete), whichever occurs first.	Postremission implications ADAMTS13 remission (partial or complete) is always accompanied by clinical remission. However, clinical remission may occur with or without an ADAMTS13 remission. Patients in clinical remission who do not achieve an ADAMTS13 remission or who experience an ADAMTS13 relapse are at increased risk of clinical relapse. In such patients, preemptive immunosuppression (e.g., rituximab) may be used to attain an ADAMTS13 remission, thereby reducing the risk of clinical relapse.
	Partial ADAMTS13 remission	ADAMTS13 activity ≥20% to <LLN.*,†	
	Complete ADAMTS13 remission	ADAMTS13 activity ≥LLN.	
Relapse	Clinical relapse	After a clinical remission, platelet count decreases to <150 × 10^9/L (with other causes of thrombocytopenia ruled out), with or without clinical evidence of new ischemic organ injury. A clinical relapse must be confirmed by documentation of severe ADAMTS13 deficiency.	
	ADAMTS13 relapse	After an ADAMTS13 remission (partial or complete), the ADAMTS13 level decreases to <20%.*,†	

Cuker A, et al. Consensus report. Blood 2021

ADAMTS13

Αναθεώρηση ορισμών για την TTP



Cuker A, et al. Consensus report. Blood 2021

ADAMTS13

Τι μετράμε

- **Δραστικότητα (activity)**
- Αντισώματα (χρήσιμα για τη διάγνωση επίκτητης TTP) ➔ όχι πάντα αυξημένα
- Αντιγόνο (έρευνα, άλλες ασθένειες)
- Γενετικός έλεγχος / Next-generation sequencing (επιβεβαιωτική εξέταση, έρευνα)

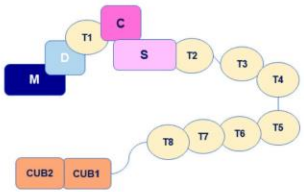
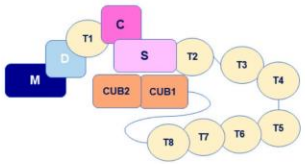
Scullie M, et al. BJH 2012

ADAMTS13

3η «μορφή» TTP ???

Acquired TTP

ADAMTS13 activity < 10 IU/dL

Most frequent characteristics	**Immune-mediated TTP : iTTP** *Positive anti-ADAMTS13 IgGs*	**TTP of unidentified mechanism : uTTP** *Negative anti-ADAMTS13 IgG*
Inaugural presentation	**Idiopathic**	**Non-idiopathic** Clinical contexts: - autoimmune disease - infection - cancer - transplantation - liver insufficiency - drug - pregnancy
ADAMTS13 conformation	 **open**	 **closed**
Mechanisms for ADAMTS13 deficiency	 **anti-ADAMTS13 IgG autoantibodies**	 **Synthesis/secretion defect** **Proteolytic degradation** ***(elastase, thrombin, plasmin)*** **Catalytic inhibition** ***(free hemoglobin, interleukines)***

Jolie BS, et al. Haematologica 2023

Θρομβωτική θρομβοπενική πορφύρα

παθοφυσιολογία, διάγνωση και θεραπεία

- **Κλινική τριάδα θρομβωτικής μικροαγγειοπάθειας**
- **Κληρονομική (Upshaw-Schulman) ή Επίκτητη**
- **Παθοφυσιολογία:**

Ανεπάρκεια ADAMTS13 ➔

Δε διασπώνται πολυμερή von Willebrand ➔

Θρομβωτική Μικροαγγειοπάθεια

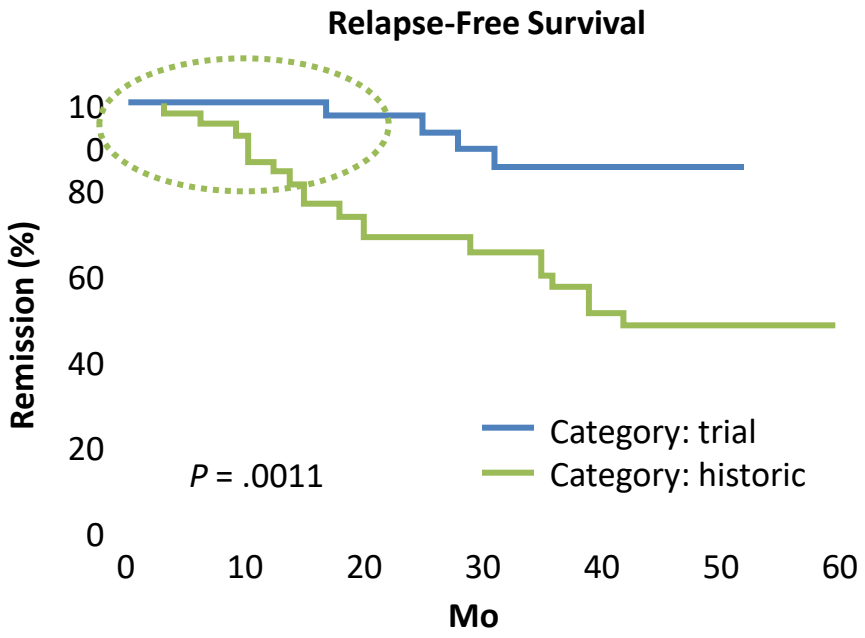
- **Διάγνωση: ADAMTS13 activity < 5 - 10%**
- **Θεραπεία:**

Επείγουσα έναρξη κορτικοειδών / πλασμαφαιρέσεων

Scullie M, et al. BJH 2012

Rituximab in aTTP: For the Best and (Not) for Worse

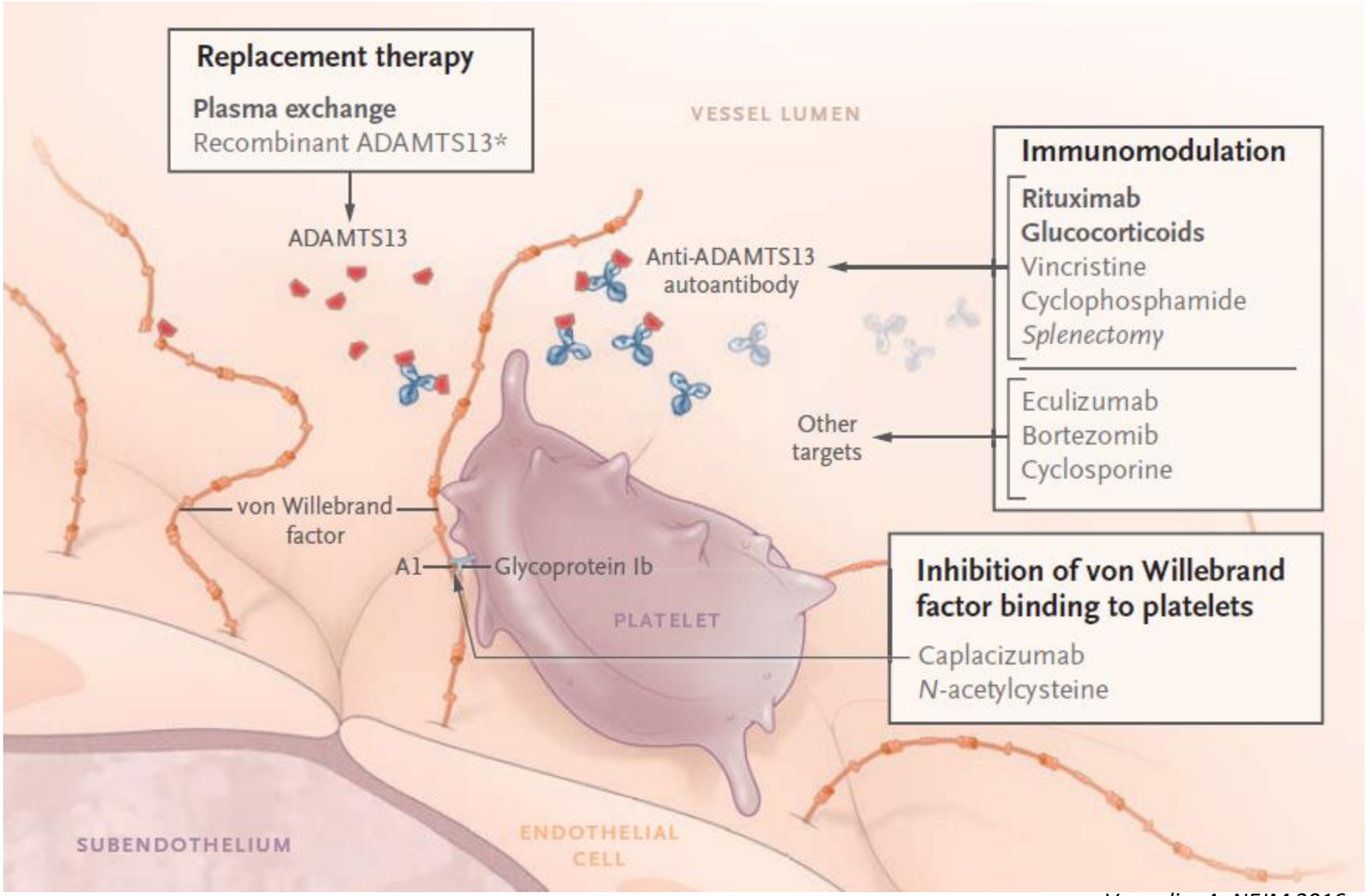
- Should all patients receive frontline rituximab?
  - Risk of overtreatment for significant number of patients in the acute phase
  - Treated patients are protected from relapse for 12-18 mo
- Patients without rituximab
  - 40% have undetectable (<10%) ADAMTS13 activity after the acute phase
  - 40% retain decreased (10%-50%) ADAMTS13 activity and are prone to relapse



Scully. Blood. 2011;118:1746.

Θρομβωτική θρομβοπενική πορφύρα

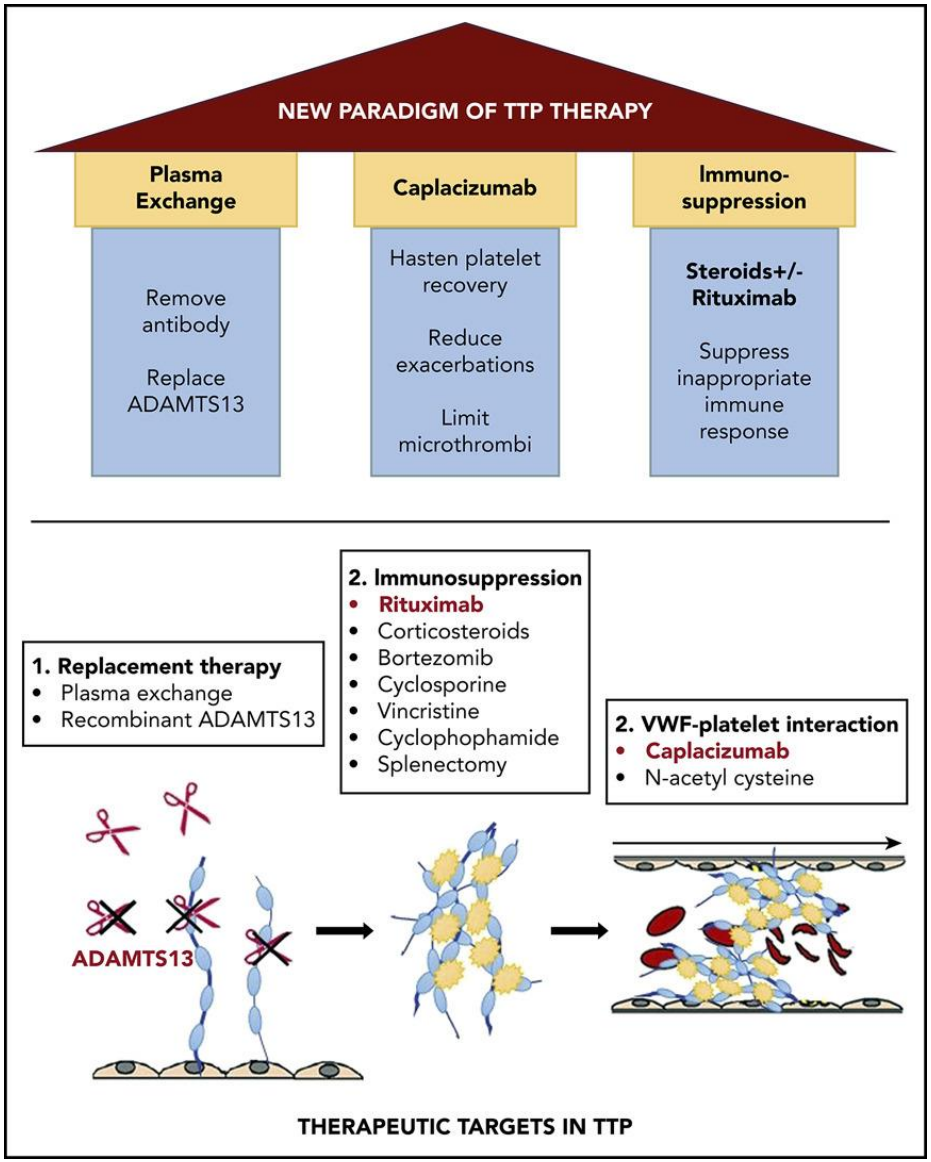
θεραπευτικές επιλογές



Veyradier A. NEJM 2016

Θρομβωτική θρομβοπενική πορφύρα

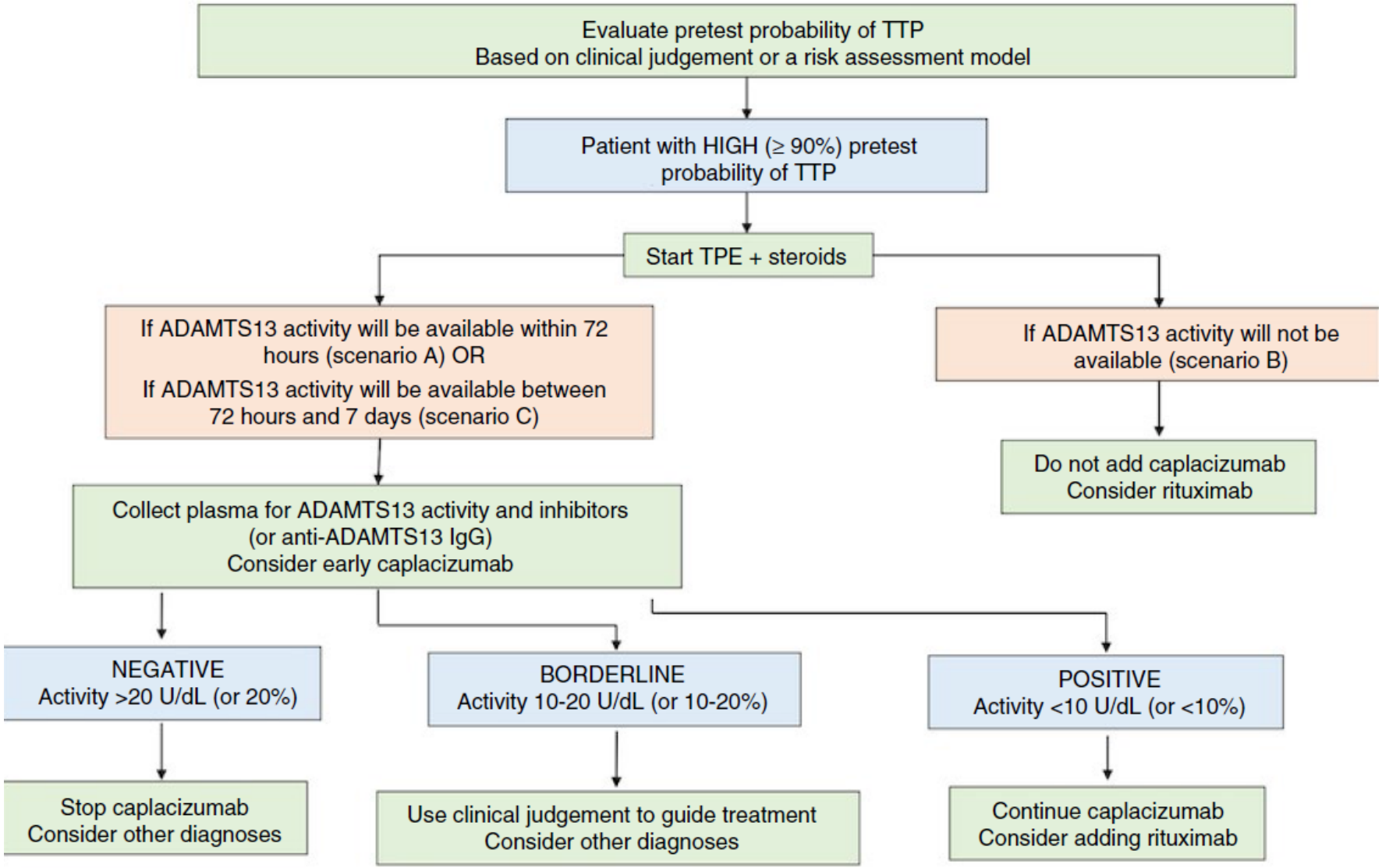
θεραπευτικές επιλογές



Mazepa M, et al Blood 2019

Θρομβωτική θρομβοπενική πορφύρα

θεραπευτικός αλγόριθμος



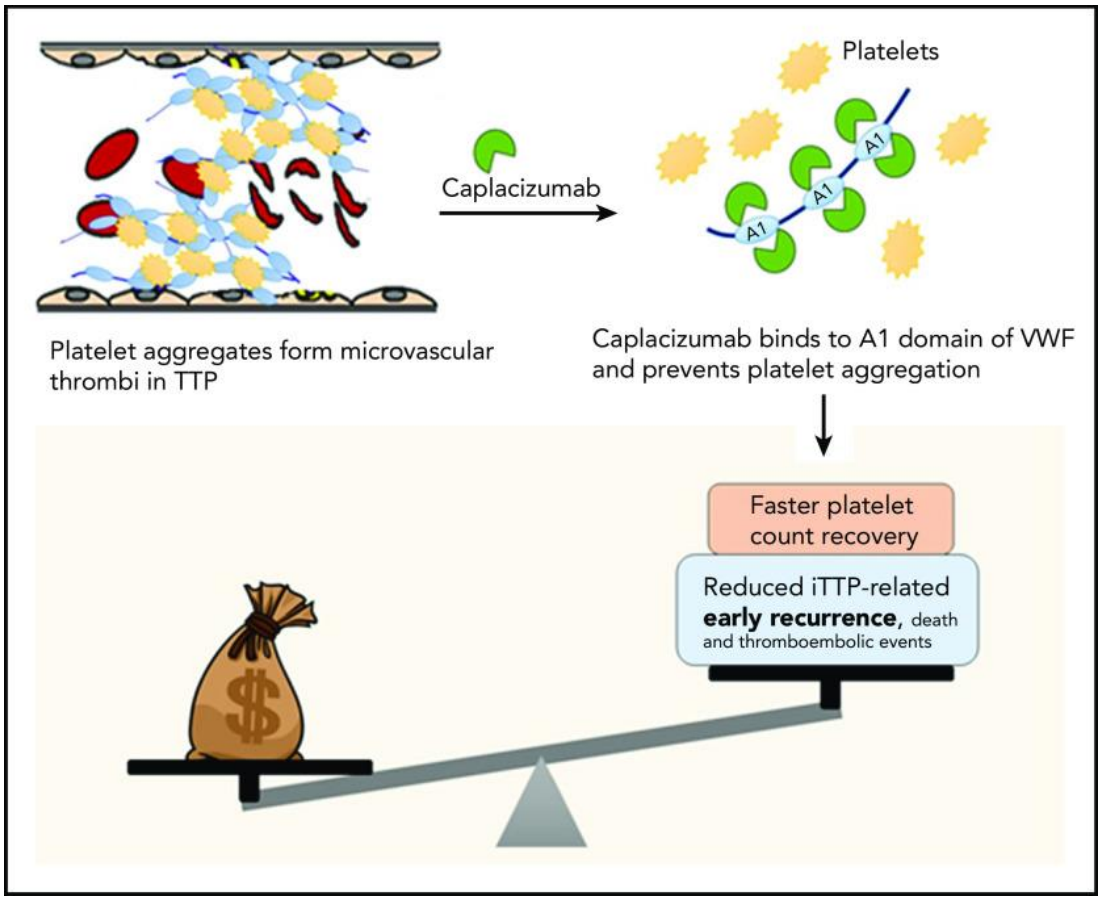
ISTH guidelines 2020

Θρομβωτική θρομβοπενική πορφύρα

νέα εγκεκριμένη θεραπεία

Caplacizumab:

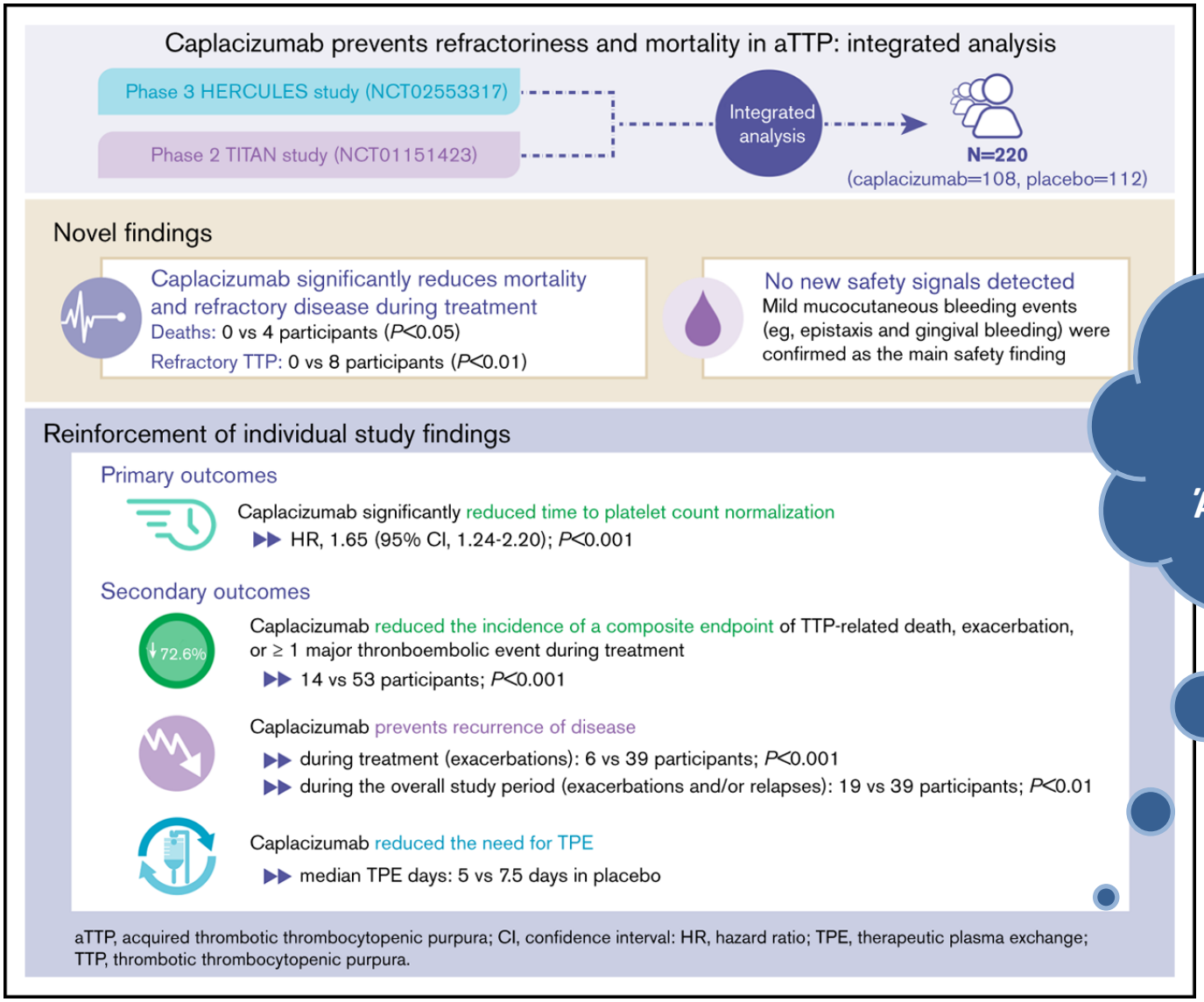
- Νανοαντίσωμα - αναστολέας von Willebrand
- **TITAN** (φάσης II) / **HERCULES** (φάσης III)



Chaturvedi S. Blood 2021

Θρομβωτική θρομβοπενική πορφύρα

caplacizumab



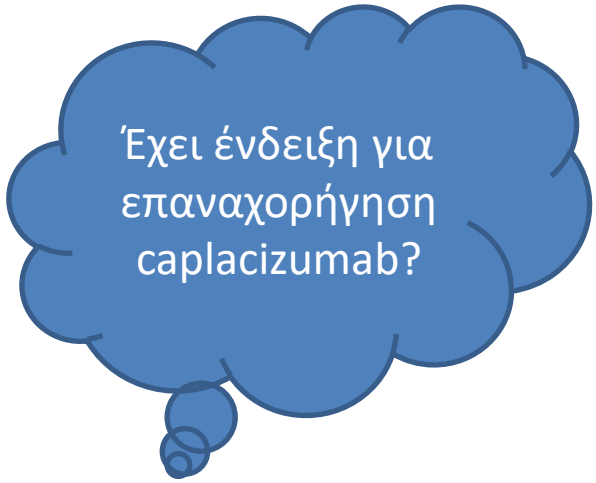
Κόστος?
Δοσολογικο σχήμα?
Άμεση έναρξη?

Peyvaldi F, et al Blood Adv 2021

Ασθενής

Πολλοστή υποτροπή TTP

- Άρρεν, 44 ετών
- 4η υποτροπή: πλασμαφαιρέσεις x 8 / caplacizumab x 30 ➔ γρήγορη άνοδος αιμοπεταλίων / ADAMTS13%
- 16/2/2021: Κλινικο-εργαστηριακή εικόνα ΤΜΑ (5η Υποτροπή)
- Ώσεις Rituximab (4-6): στις προ-τελευταίες 2 υποτροπές (όχι στην τελευταία λόγω COVID-19 – γρήγορης ανάρρωσης υπό caplacizumab)



Θρομβωτική θρομβοπενική πορφύρα

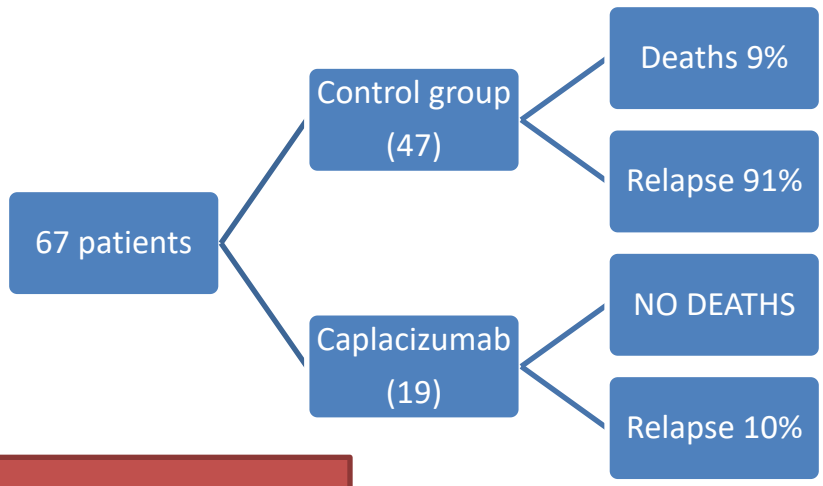
caplacizumab

Country (Year)	Centers	Control group	Treatment	Patient number	% presenting with recurrent TTP	% with ADAMTS13 <10%
France (2021)	Multicenter	Historical	Capla-cizumab+SOC	90	13	**100**
			SOC	180	12	**100**
UK (2021)	Multicenter	Historical	Capla- cizumab+ SOC	85	NA	**99**
			SOC	39	NA	**NA**
Spain (2022)	Multicenter	Concurrent	Capla- cizumab + SOC	77	4.5	**100**
			SOC	78	20.5	**100**
Germany/Austria (2023)	**Multicenter**	**Historical**	Capla-cizumab+SOC	113	2	**97**
			SOC	**119**	**4**	**92**

Gavriilaki E, et al. Frontiers in Medicine (Hematology) 2023

Θρομβωτική θρομβοπενική πορφύρα
caplacizumab

A



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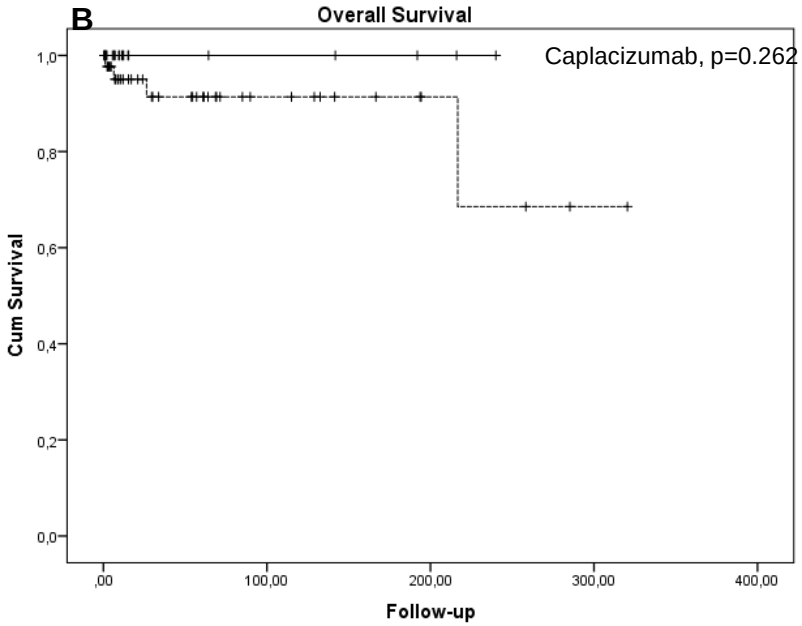

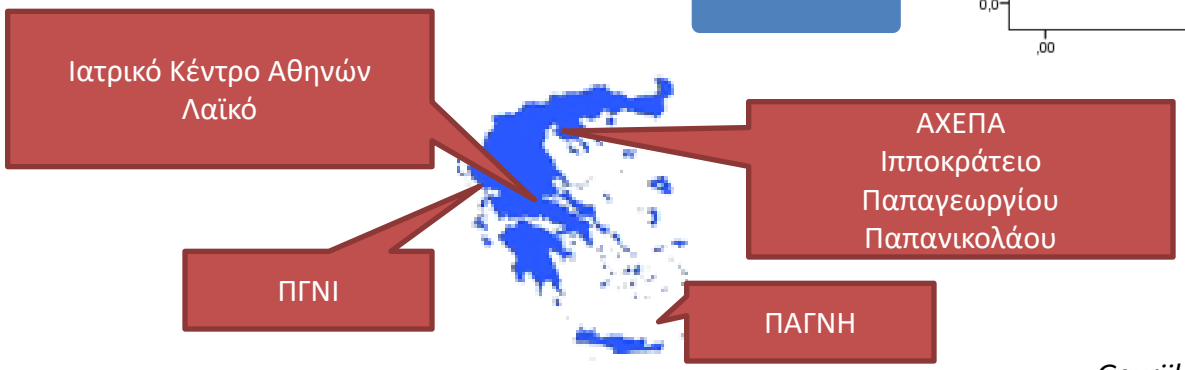

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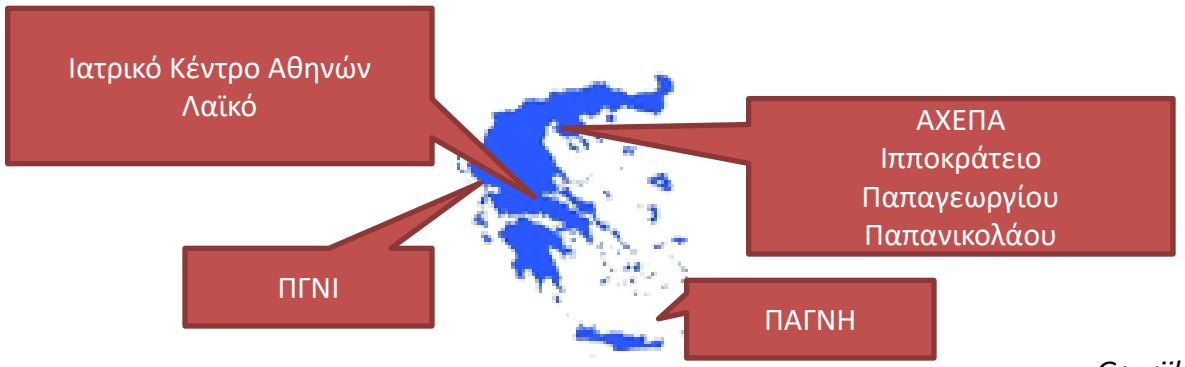
Θρομβωτική θρομβοπενική πορφύρα
caplacizumab

- Επιτυχής η επαναχορήγηση
- Επιτυχής η χορήγηση χωρίς πλασμαφαίρεση



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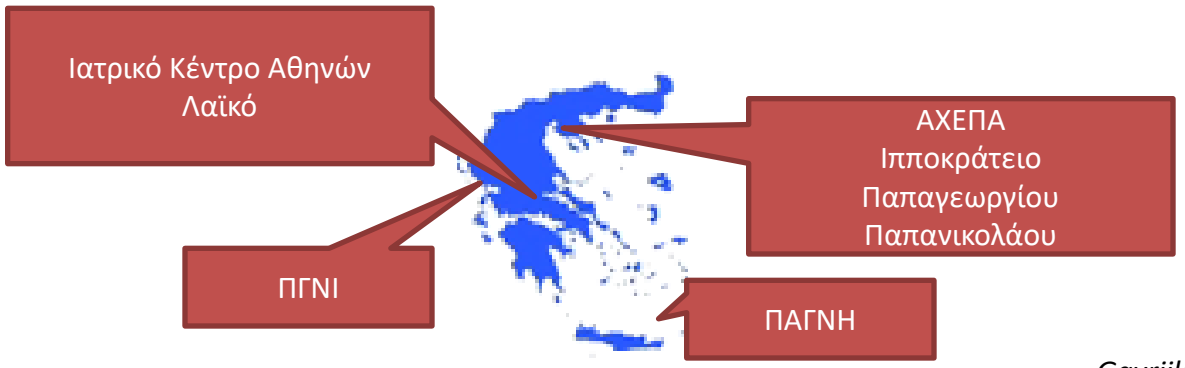
Θρομβωτική θρομβοπενική πορφύρα

caplacizumab – για πόσο ?

- Σύμφωνα με spc
- Συνέχιση χορήγησης σε exacerbation
- Μελετάται η διακοπή μόλις ανέβουν επίπεδα ADAMTS13 > 10%



Gavriilaki E, et al. Frontiers in Medicine (Hematology) 2023

ADAMTS13 recovery in acute thrombotic thrombocytopenic purpura after caplacizumab therapy

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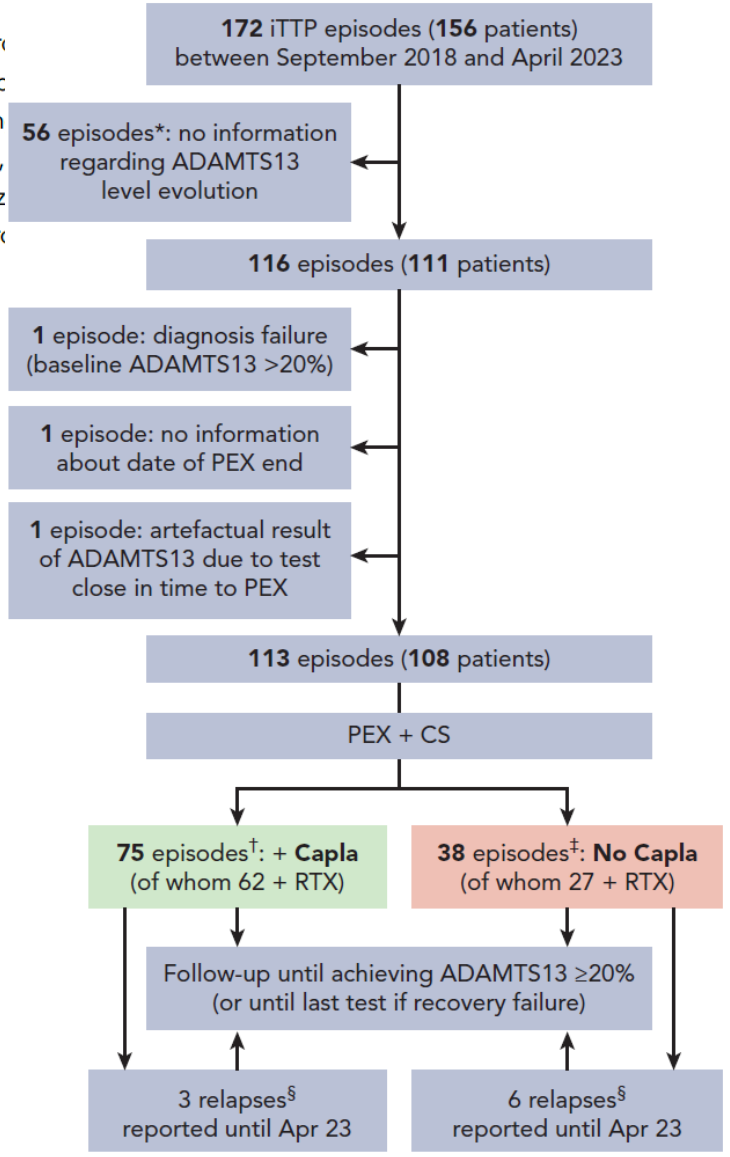
Inés Gómez-Seguí,[6] María-Liz Paciello-Coronel,[7] David Valcárcel-Ferreiras,[8] Moraima Jiménez,[8] Jc
José-María García-Gala,[10] Sonia Angós-Vazquez,[11] Miriam Vara-Pampliega,[12] Luisa Guerra-Domín
Ana Oliva-Hernandez,[15] Saioa Zalba-Marcos,[16] Inmaculada Tallón-Ruiz,[17] Sandra Ortega-Sánchez,
Gemma Moreno-Jiménez,[20] Lourdes Domínguez-Acosta,[21] María Araiz-Ramírez,[22] Luis Hernández
Julio del Río-Garma,[25] and Cristina Pascual-Izquierdo,[26,27] on behalf of the Spanish Apheresis Gr
Thrombocytopenic Purpura Registry



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KEY POINTS

- **There is no delay in ADAMTS13 recovery after PEX start in caplacizumab–treated patients with iTTP from the Spanish registry.**
- **Caplacizumab allows suspending PEX earlier, thus creating the impression that there is a delay in ADAMTS13 recovery after PEX end.**

blood VTH
vessels, thrombosis & hemostasis

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bloodVTH@hematology.org

Real-World Insights on the Management of Immune-Mediated Thrombotic Thrombocytopenic Purpura (iTTP) With Caplacizumab

Tracking no: VTH-2024-000151R2

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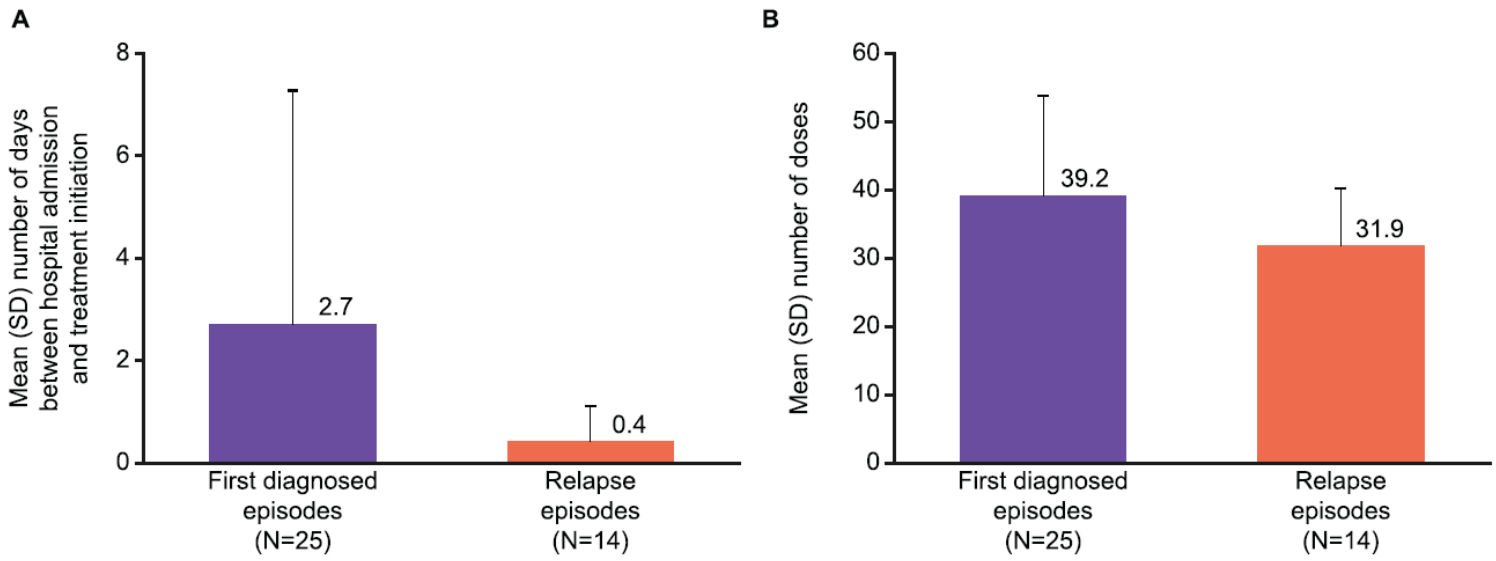
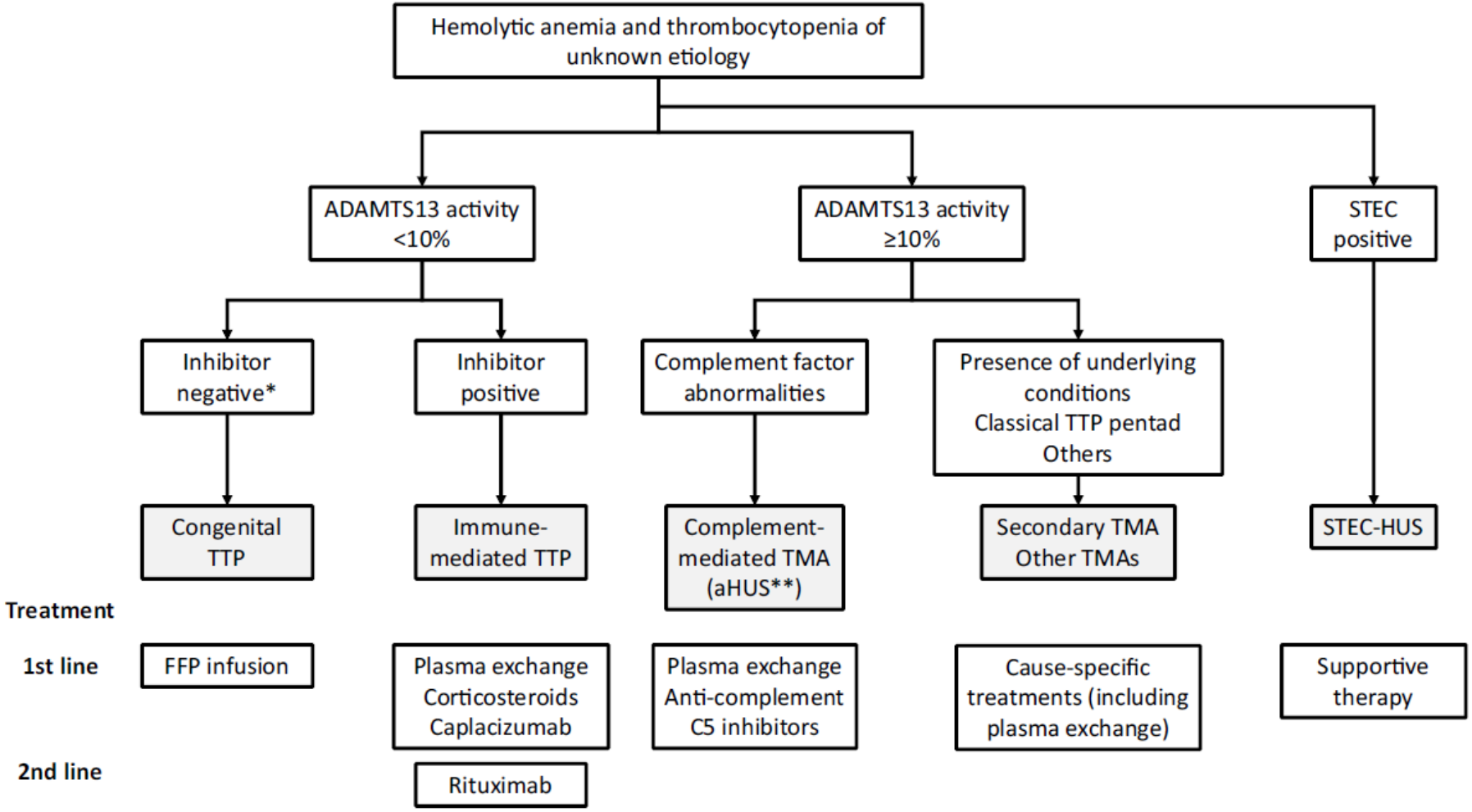


Άλλες οδηγίες



Japanese guidelines 2023

Άλλες οδηγίες

Remission period

CQ3: Is rituximab recommended if a patient with iTTP shows a marked reduction in ADAMTS13 activity during remission?
Answer: If ADAMTS13 activity decreases to < 10% during remission in a patient with iTTP, rituximab may be considered for preventing clinical relapse (off-label use in Japan, grade of recommendation: 2B).

Japanese guidelines 2023

Άλλες οδηγίες

Initial management of suspected TTP

Aim: urgent plasma exchange in a safe clinical environment

- Admit patient under the experienced TTP team to agreed location for acute TTP management
- Defrost AB OctaplasLG during patient transfer to ensure PEX starts ASAP
- Arrange wide bore intravenous catheter (or peripheral access to avoid delay) to enable PEX
- Initiate 1.5 volume plasma exchange (PEX) as soon as possible (target 4-8 hrs).
- Avoid platelet transfusion
- Initiate steroids post PEX
- Initiate caplacizumab on confirmation of TTP

Urgent investigations to confirm suspected diagnosis of TTP

Aim: *Investigations required for a new TTP referral*

- As Table III
- *ADAMTS13* samples pre PEX

Ongoing management

Aim: *continued daily plasma exchange +- immunosuppression +- Caplacizumab*

- Ongoing multidisciplinary team management of patient
- Follow protocol for TTP management. Continue PEX until platelets > 150×10^9/L.
- Initial rituximab.
- Continue caplacizumab.
- Enroll in clinical trial if available

British guidelines 2023

Caplacizumab Added to Plasma Exchange and Immunosuppression Hastens Recovery and Improves Survival in Immune TTP: an International Real-World Study of the TTP-IWG (The Capla 1000+ Project)

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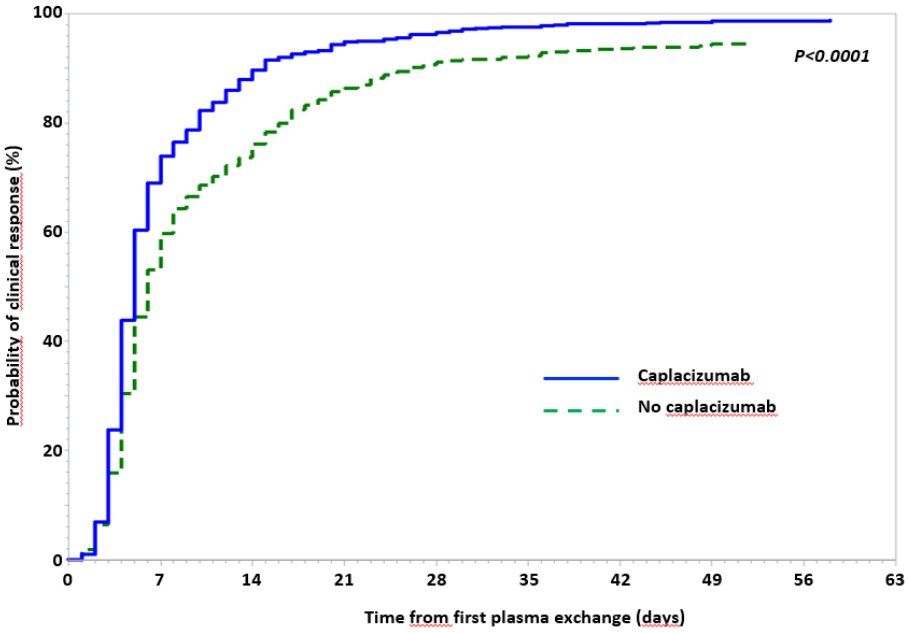

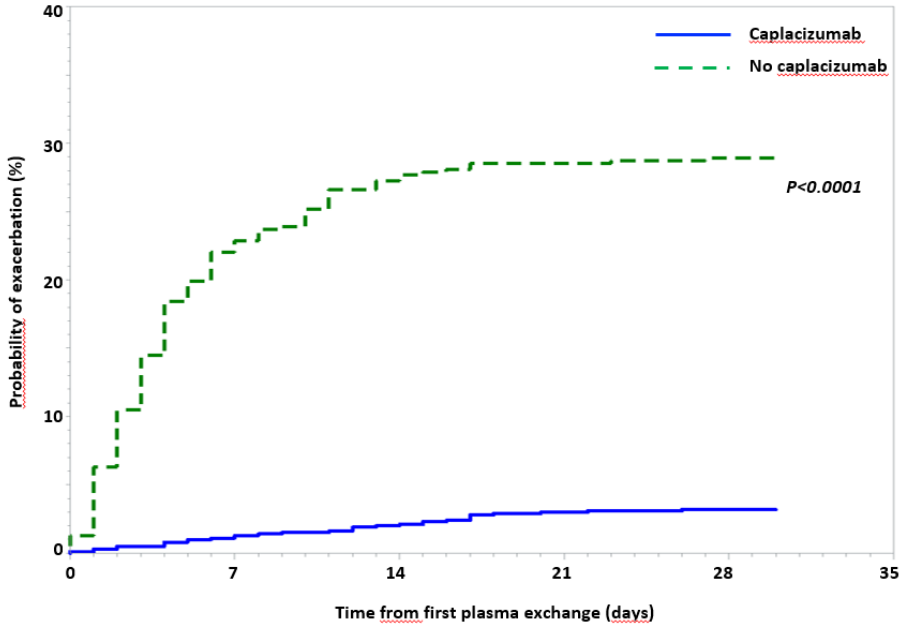

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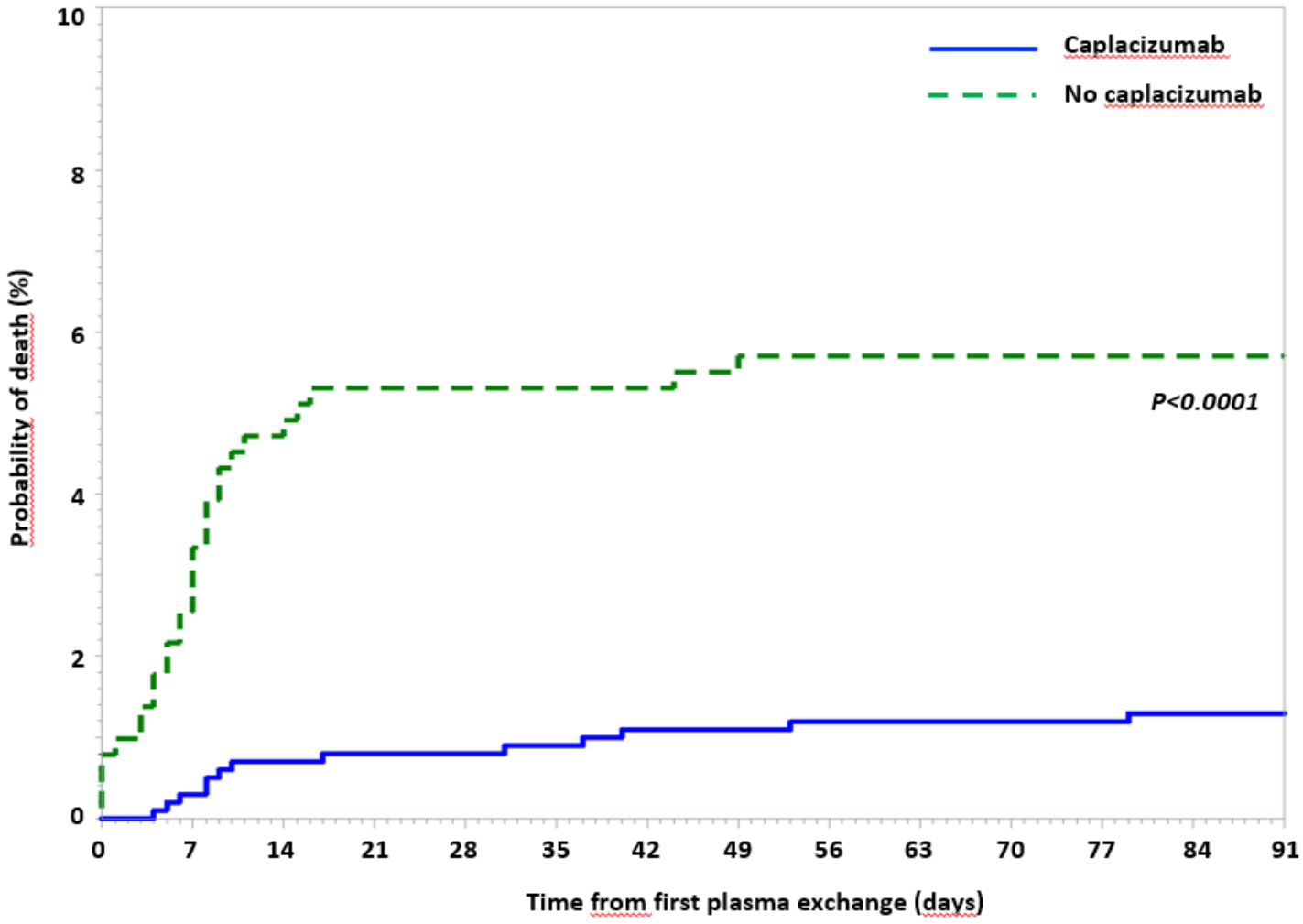
Caplacizumab Added to Plasma Exchange and Immunosuppression Hastens Recovery and Improves Survival in Immune TTP: an International Real-World Study of the TTP-IWG (The Capla 1000+ Project)



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Θρομβωτική θρομβοπενική πορφύρα

το μέλλον

Ανασυνδυασμένη ADAMTS13

- Κληρονομική
- Μεταφορά μέσω αιμοπεταλίων στην επίκτητη TTP

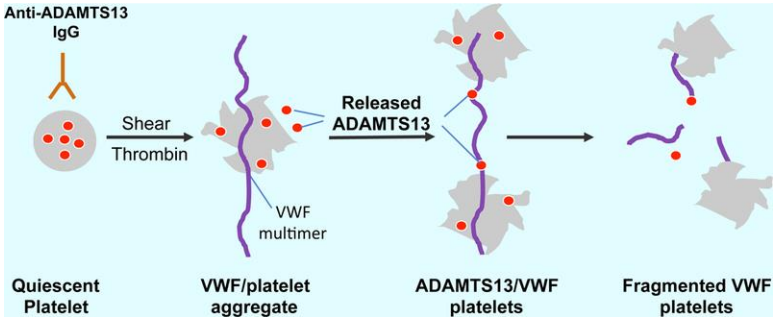
Plenary Paper

blood

CLINICAL TRIALS AND OBSERVATIONS

Recombinant ADAMTS-13: first-in-human pharmacokinetics and safety in congenital thrombotic thrombocytopenic purpura

Marie Scully,[1] Paul Knöbl,[2] Karim Kentouche,[3] Lawrence Rice,[4] Jerzy Windyga,[5] Reinhard Schneppenheim,[6] Johanna A. Kremer Hovinga,[7] Michiko Kajiwara,[8] Yoshihiro Fujimura,[9] Caterina Maggiore,[10] Jennifer Doralt,[11] Christopher Hibbard,[12] Leah Martell,[12] and Bruce Ewenstein[12]



- Προστασία από αυτοαντισώματα
- Πειραματική μελέτη
- Υγιείς εθελοντές

Abdelgawwad MS, et al. ATVB 2019

Ανασυνδυασμένη ADAMTS13

κληρονομική TTP

The NEW ENGLAND JOURNAL of MEDICINE

ORIGINAL ARTICLE

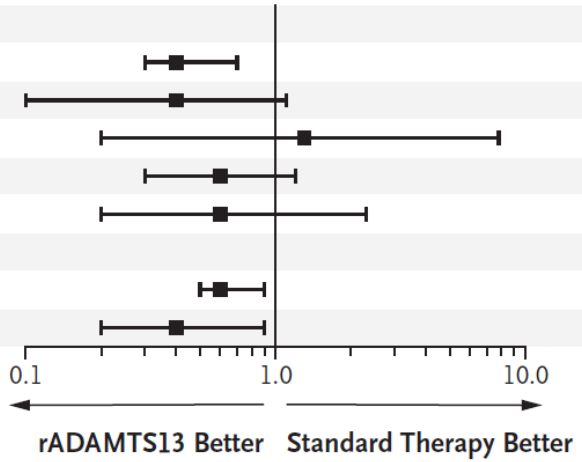
Recombinant ADAMTS13 in Congenital Thrombotic Thrombocytopenic Purpura

Marie Scully, M.D., Ana Antun, M.D., Spero R. Cataland, M.D., Paul Coppo, M.D., Ph.D., Claire Dossier, M.D., Nathalie Biebuyck, M.D., Wolf-Achim Hassenpflug, M.D., Karim Kentouche, M.D., Paul Knöbl, M.D., Johanna A. Kremer Hovinga, M.D., M. Fernanda López-Fernández, M.D., Ph.D., Masanori Matsumoto, M.D., Ph.D., Thomas L. Ortel, M.D., Ph.D., Jerzy Windyga, M.D., Ph.D., Indranil Bhattacharya, Ph.D., Michael Cronin, Pharm.D., Hong Li, Ph.D., Björn Mellgård, M.D., Ph.D., Munjal Patel, Ph.D., Parth Patwari, M.D., Sc.D., Shan Xiao, Ph.D., Pinghai Zhang, M.D., Ph.D., and Linda T. Wang, M.D., for the cTTP Phase 3 Study Investigators*

Ανασυνδυασμένη ADAMTS13

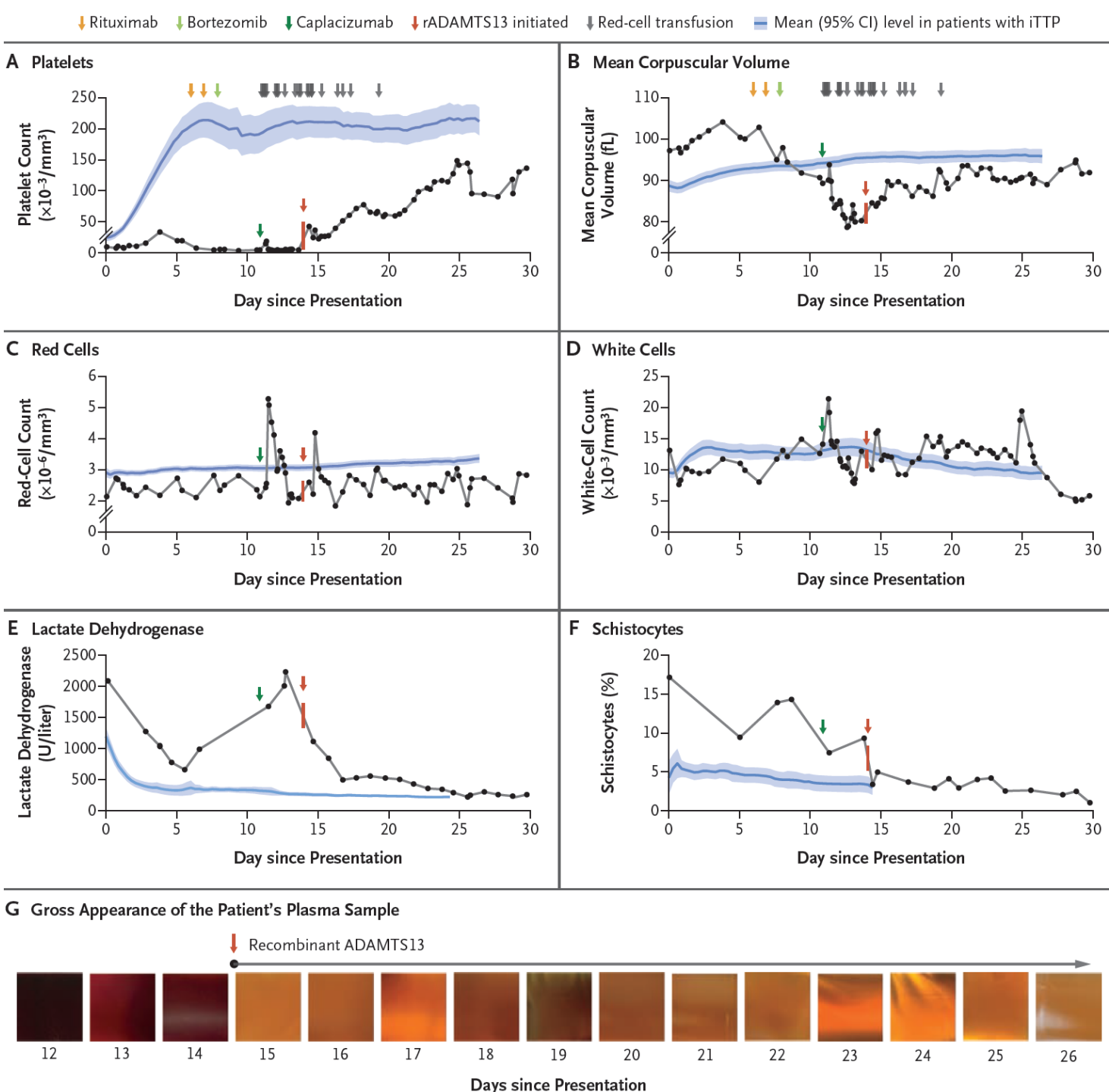
κληρονομική TTP

Outcome	rADAMTS13	Standard Therapy	Estimated Ratio (95% CI)
	least-squares mean annualized incidence rate (95% CI)		
Prespecified TTP manifestations			
Thrombocytopenia	0.74 (0.37–1.50)	1.73 (0.92–3.23)	0.4 (0.3–0.7)
Elevated LDH level	0.26 (0.10–0.72)	0.74 (0.38–1.44)	0.4 (0.1–1.1)
Increased creatinine level	0.10 (0.02–0.47)	0.08 (0.02–0.36)	1.3 (0.2–7.8)
Neurologic symptoms	0.18 (0.06–0.51)	0.29 (0.11–0.78)	0.6 (0.3–1.2)
Abdominal pain	0.11 (0.03–0.39)	0.17 (0.06–0.52)	0.6 (0.2–2.3)
Exploratory outcomes			
Composite TTP manifestations	2.28 (1.35–3.88)	3.68 (2.23–6.08)	0.6 (0.5–0.9)
Other TTP manifestations	0.32 (0.14–0.72)	0.81 (0.44–1.49)	0.4 (0.2–0.9)



Ανασυνδυασμένη ADAMTS 13

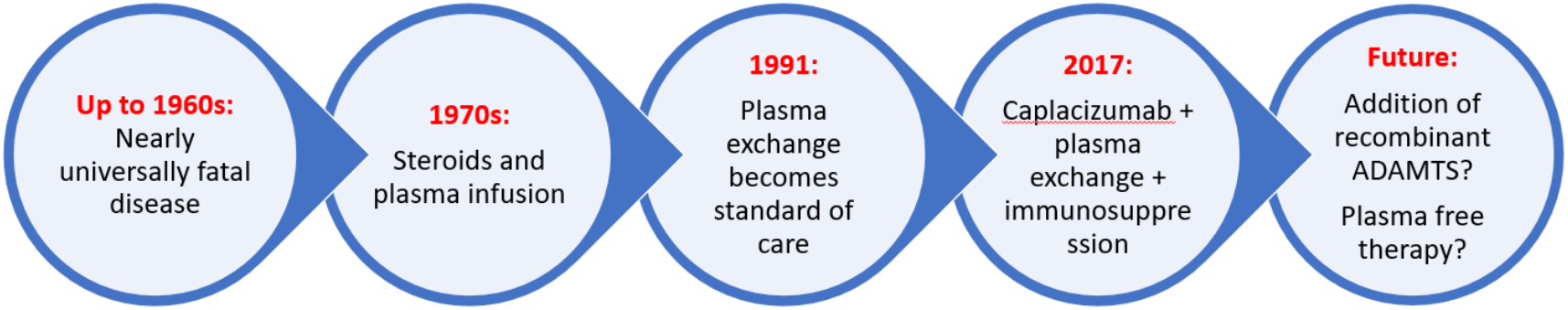
Επίκτητη TTP



Θρομβωτική θρομβοπενική πορφύρα

Εξέλιξη των θεραπευτικών επιλογών

100 χρόνια μετά



Sukumar S, Gavriilaki E, Chaturvedi S. Thrombosis Update 2021

Is plasma exchange history in TTP ?

- Μάρτυρες Ιεχωβά
- Μη διαθέσιμη πλασμαφαίρεση λόγω COVID-19
- Αναφυλαξία σε Octaplas
- Απόφαση ασθενούς

Θρομβωτική θρομβοπενική πορφύρα

αλλαγή της φυσικής πορείας νόσου?

Survivorship aspect	Outcome
Overall survival	Shorter survival than age and sex matched general population controls; iTTP relapse and cardiovascular disease are leading causes of death [94]
Lupus	Prevalence of SLE 37-fold greater than expected among age, race, and sex-matched reference population; SLE is the most common additional autoimmune disorder occurring in iTTP patients[92]
Hypertension	Lifetime prevalence in some cohorts is 45%, which is significantly greater than expected (23%) for the general population. The prevalence of HTN in this cohort prior to a diagnosis of TTP did not differ from the general population.[92]
Stroke	In addition to acute phase cerebral events, patients are at 5-fold increased risk for stroke during remission if ADAMTS13 activity <70%[95]
Depression and post-traumatic stress disorder	Rates for depression and PTSD are >80% and 35% respectively in some cohorts; Previous diagnosis of depression and unemployment attributed to TTP were associated with depression whereas younger age, pre-existing anxiety, and unemployment due to TTP were associated with PTSD[91]
Quality of life	Patients in remission consistently score lower across all domains of HRQoL surveys; these results do not improve over time and have no correlation with severity of the index TTP episode[110]
Cognitive impairment	Often develop impairment, particularly in certain cognitive domains: complex attention and sequencing, manual dexterity, rapid language generation, and list learning; can lead to disability and unemployment.[111]
Pregnancy outcomes	When index iTTP triggered by pregnancy, subsequent pregnancy is associated with recurrence. ADAMTS13 activity monitoring before and during pregnancy, with pre-emptive treatment, may prevent recurrence in this setting[77]

Sukumar S, Gavriilaki E, Chaturvedi S. Thrombosis Update 2021

Θρομβωτική θρομβοπενική πορφύρα

επιπλοκές

OPEN ACCESS

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SPECIALTY SECTION

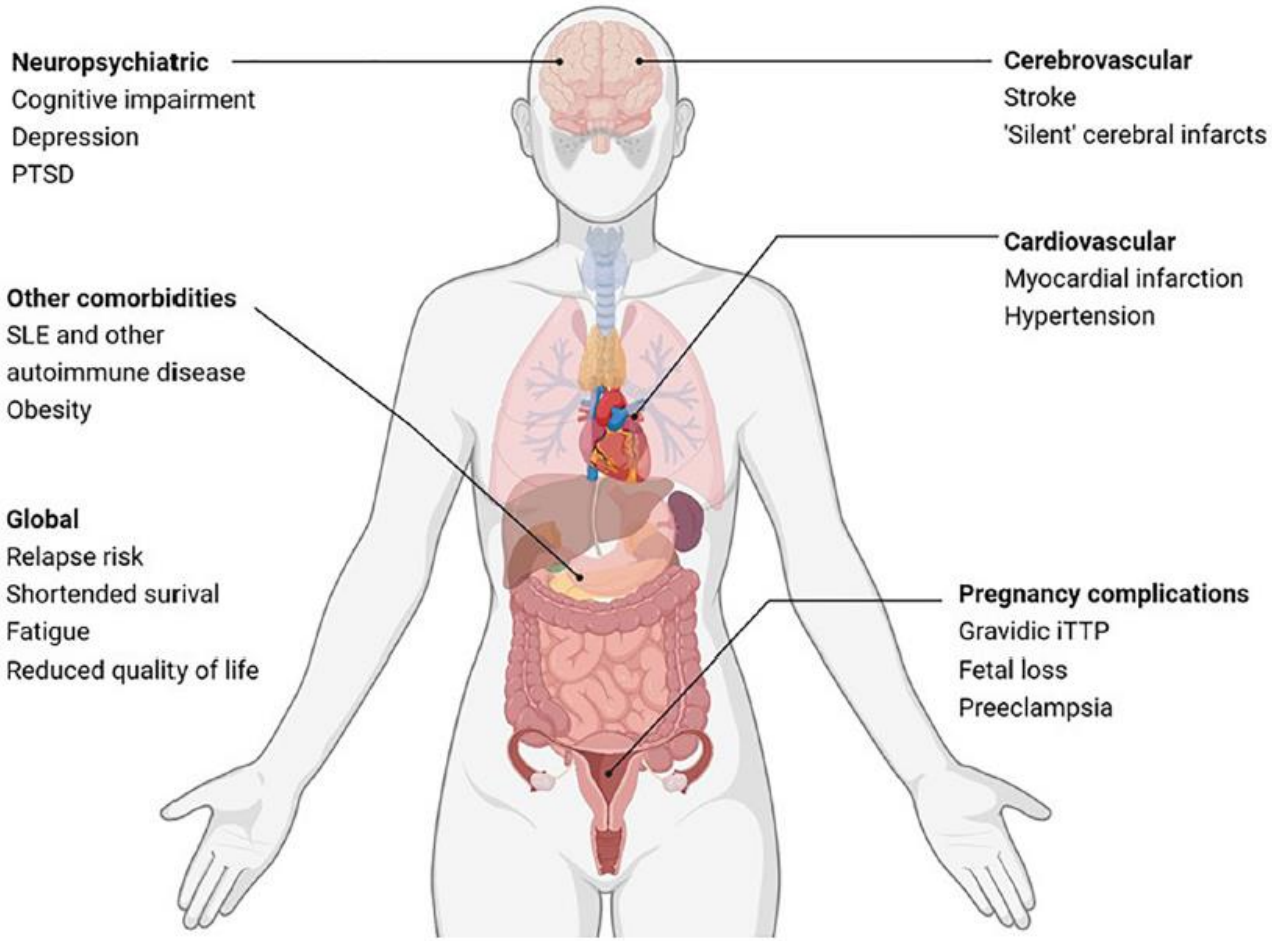
Immune thrombotic thrombocytopenic purpura: Spotlight on long-term outcomes and survivorship

Sruthi Selvakumar[1], Angela Liu[2] and Shruti Chaturvedi[3]*

[1]Dr. Kiran C. Patel College of Allopathic Medicine, Nova Southeastern University, Fort Lauderdale, FL, United States, [2]Division of Hematology and Oncology, Mount Sinai School of Medicine, New York, NY, United States, [3]Division of Hematology, Department of Medicine, Johns Hopkins University School of Medicine, Baltimore, MD, United States

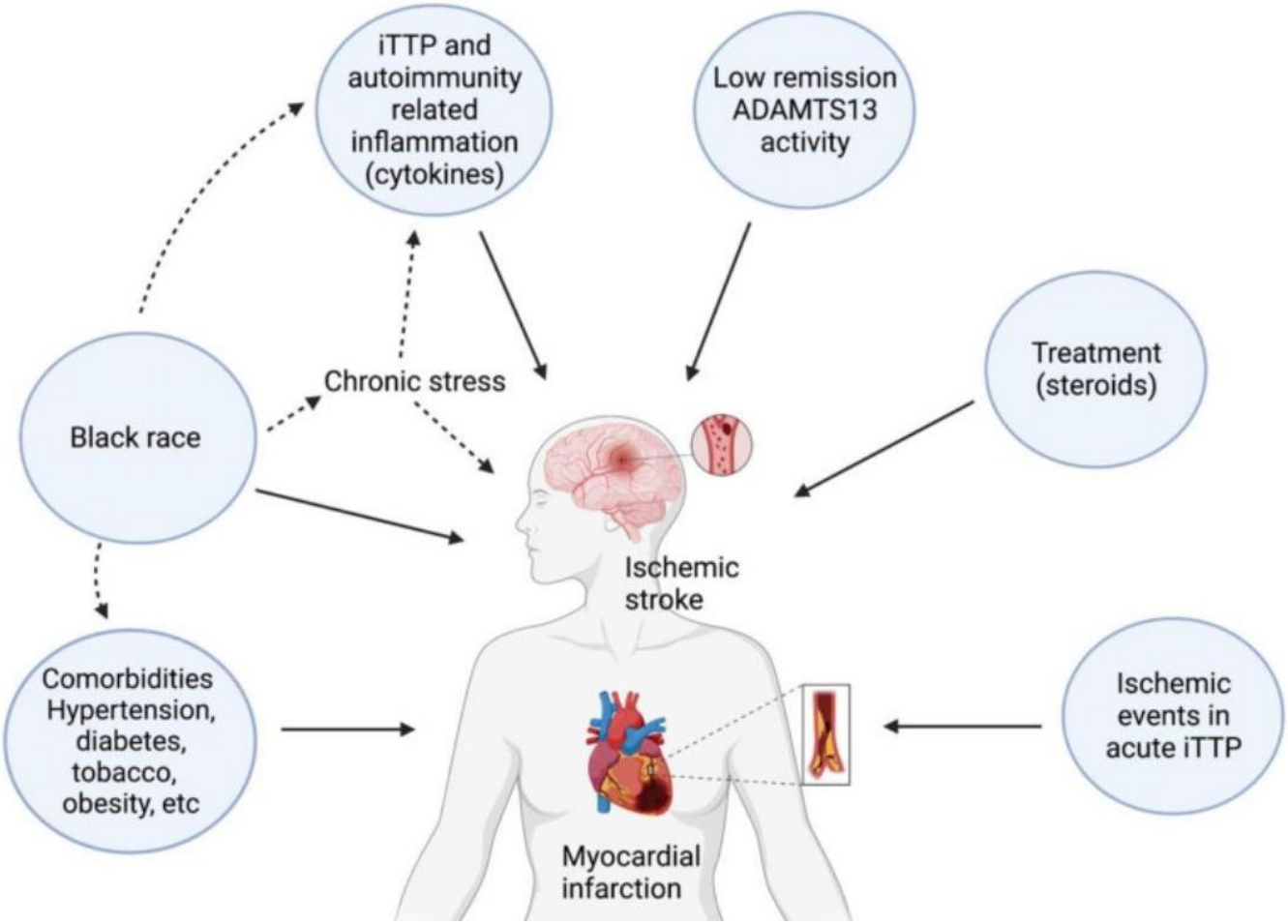
Θρομβωτική θρομβοπενική πορφύρα

επιπλοκές



Selvakumar S, et al. Front Med 2023

Καρδιαγγειακή νόσος



Sukumar C, et al. J Clin Med 2023

Θρομβωτική θρομβοπενική πορφύρα

αλλαγή της φυσικής πορείας νόσου?

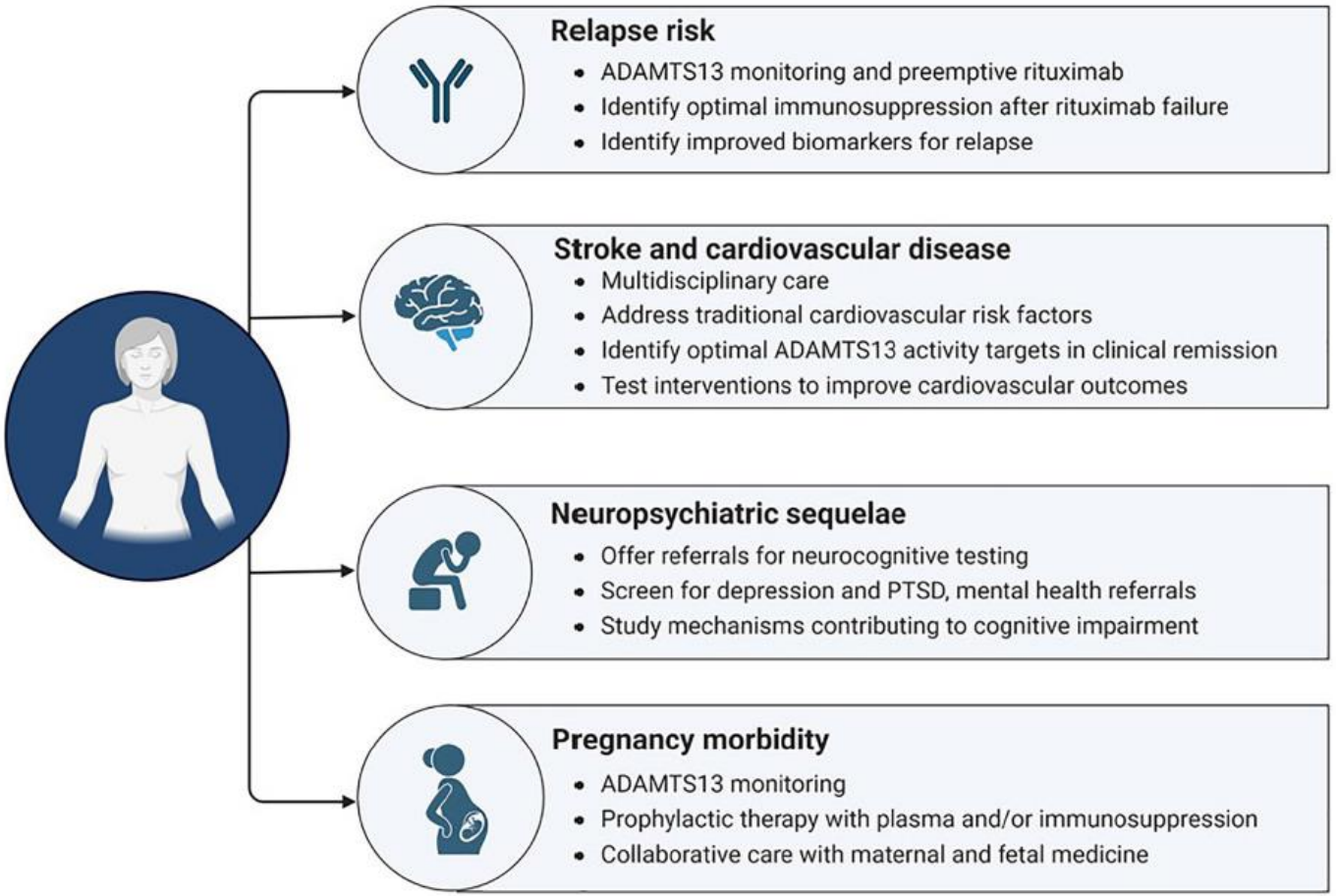


FIGURE 4
Opportunities to mitigate risk and improve long term outcomes in iTTP.

Selvakumar S, et al. Front Med 2023

Θρομβωτική θρομβοπενική πορφύρα

και κύηση ?

ΘΡΟΜΒΩΤΙΚΗ ΜΙΚΡΟΑΓΓΕΙΟΠΑΘΕΙΑ ΚΑΤΑ ΤΗΝ ΚΥΗΣΗ: Ο ΡΟΛΟΣ ΤΗΣ ΔΡΑΣΤΙΚΟΤΗΤΑΣ ADAMTS13

Ελένη Γαβριηλάκη[1], Δέσποινα Μπαρμπαρούση[2], Εμμανουήλ Παπαδάκης[3], Ευδοξία-Ευαγγελία Κοράβου[4], Λαμπρινή Βαχτσετζή[4], Τασούλα Τουλουμενίδου[4], Αποστολία Παπαλεξανδρή[4], Κατερίνα Μπαλάσκα[1], Νικόλαος Κώτσιου[1], Σοφία Χισσάν[1], Αναστασία Μπαντή[5], Χαρά Ματσούκα[2], Νίκη Ρουγκάλα[6], Θεώνη Λεωνιδοπούλου[6], Ευδοκία Μανδαλά[7], Μιχάλης Δούμας[1], Ιωάννα Σακελλάρη[2], Σοφία Βακαλοπούλου[1]

Θρομβωτική θρομβοπενική πορφύρα

και κύηση ?

RESEARCH LETTER

blood advances

TO THE EDITOR:

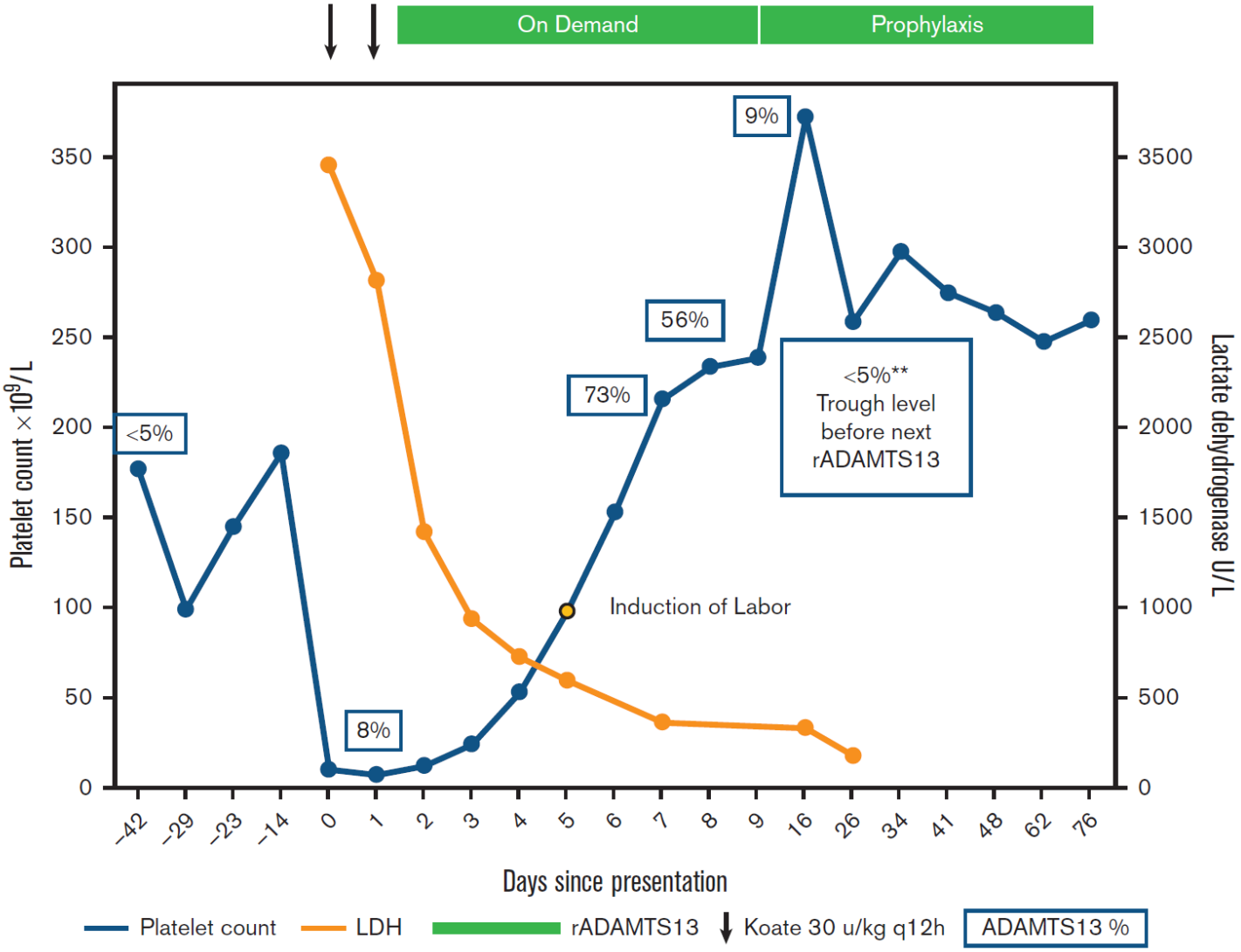
Recombinant ADAMTS13: an effective rescue therapy for acute cTTP during pregnancy

Simon Dadoun,[1] Karolina Adam,[2] Lisa Hensch,[3] Theonia K. Boyd,[4] Sami Ibrahimi,[5] James N. George,[5] Marie Scully,[6] and Senthil Sukumar[7]

[1]Department of Obstetrics & Gynecology, Baylor College of Medicine, Houston, TX; [2]Department of Obstetrics & Gynecology, Texas Children's Hospital, Houston, TX; [3]Division of Transfusion Medicine & Coagulation, Department of Pathology & Immunology, Baylor College of Medicine, Houston, TX; [4]Division of Anatomic Pathology, Department of Pathology & Immunology, Baylor College of Medicine, Houston, TX; [5]Hematology-Oncology Section, Department of Medicine, University of Oklahoma Health Sciences Center, Oklahoma City, OK; [6]Department of Haematology, University College London Hospital, Haematology Theme-NIHR UCLH/UCL Biomedical Research Centre, London, United Kingdom; and [7]Section of Hematology-Oncology, Department of Medicine, Baylor College of Medicine, Houston, TX

Θρομβωτική θρομβοπενική πορφύρα

και κύηση ?

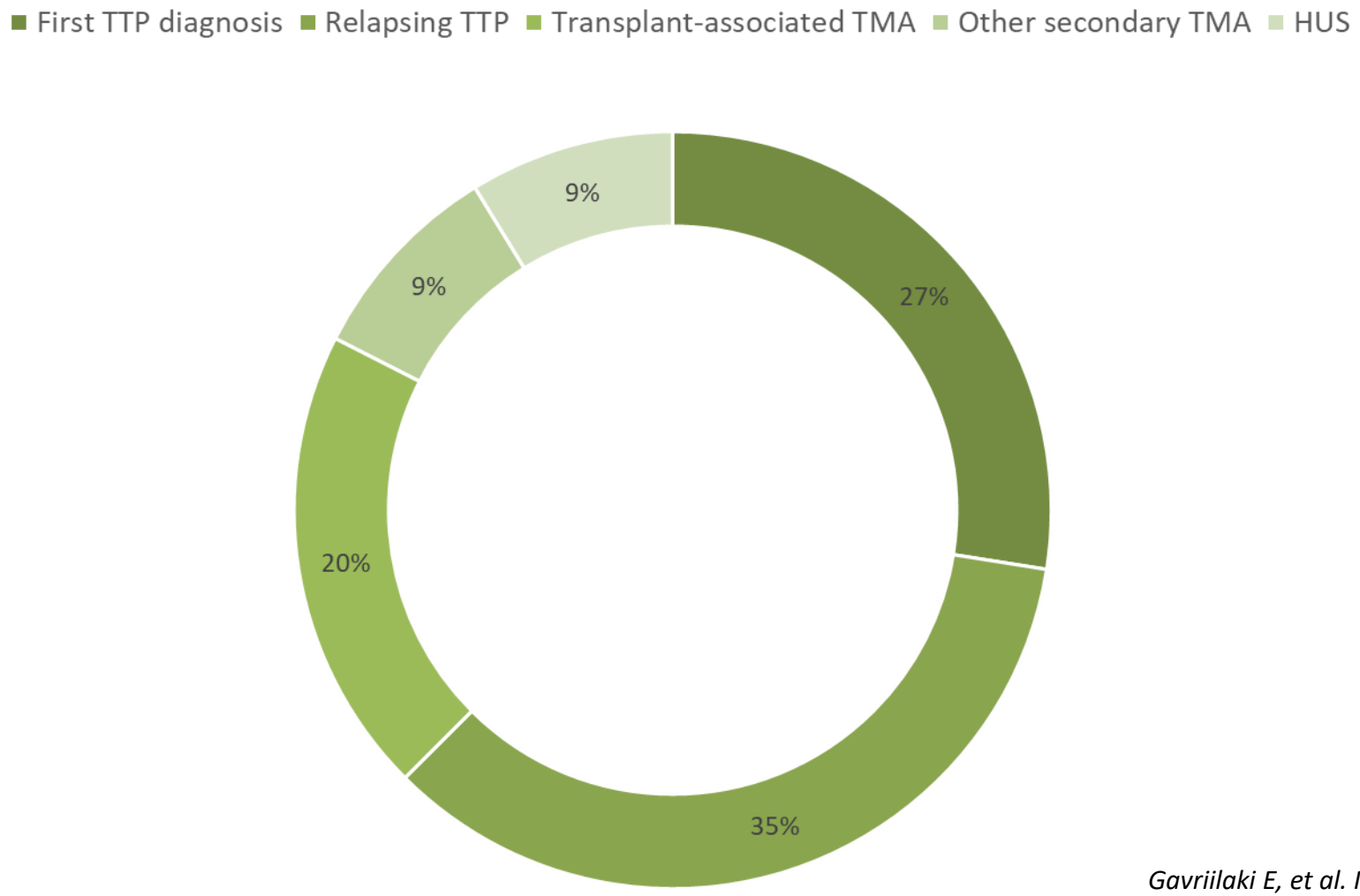


Θρομβωτικές μικροαγγειοπάθειες

HUS / CM-TMA

Θρομβωτική μικροαγγειοπάθεια

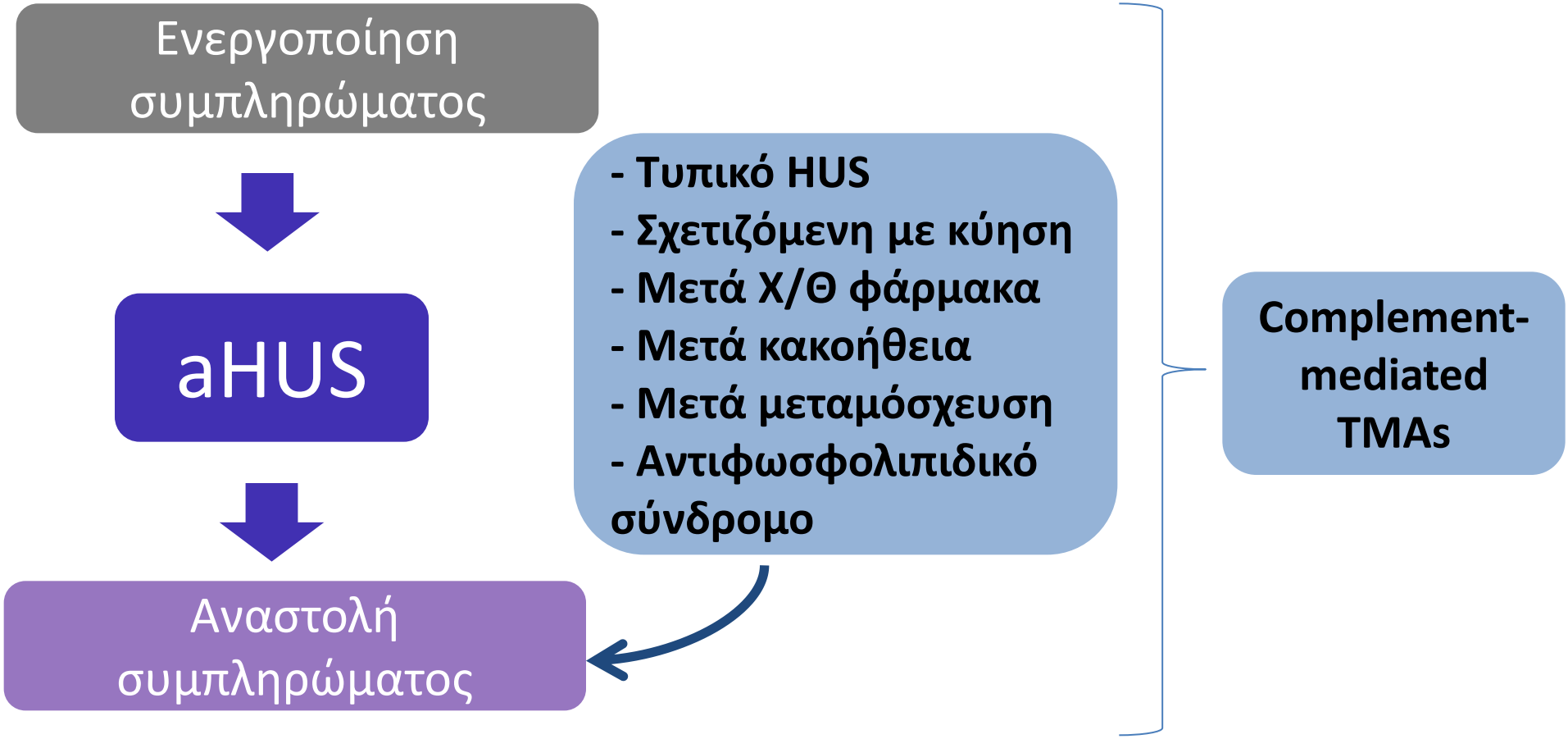
διαφορική διάγνωση



Gavriilaki E, et al. ISTH 2020

Θρομβωτική μικροαγγειοπάθεια

ονοματολογία που σχετίζεται με την παθοφυσιολογία



Gavriilaki E, et al. Fronitiers in Immunology 2018

Θρομβωτική μικροαγγειοπάθεια

διάγνωση στην πράξη

EUROPEAN HEMATOLOGY ASSOCIATION

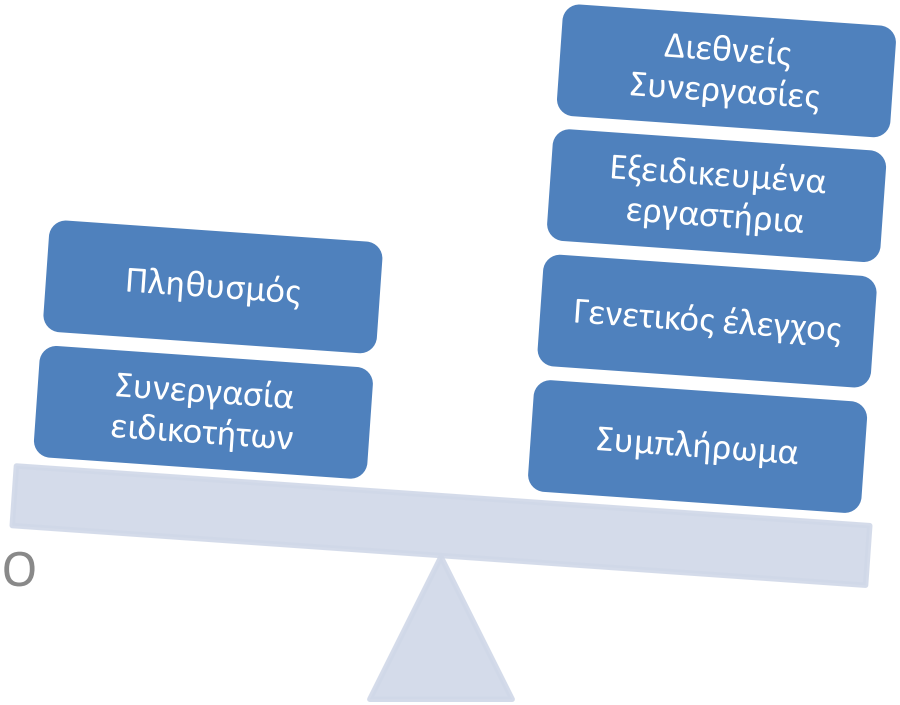
- Μικροβιολογικός έλεγχος (διαρροϊκό σύνδρομο, influenza, πνευμονιόκοκκος)
- Δραστικότητα ADAMTS13 / ανασταλτές
- Ορολογικοί δείκτες ενεργοποίησης συμπληρώματος (C5b-9, modified Ham test, CFH autoantibodies)
- Γενετικός έλεγχος (NGS-μελέτες)

ΕΘΝΙΚΟ ΜΗΤΡΩΟ

ΘΡΟΜΒΩΤΙΚΗ ΜΙΚΡΟΑΓΓΕΙΟΠΑΘΕΙΑ

Κατά

Υπέρ



ΕΘΝΙΚΟ ΜΗΤΡΩΟ

https://hshtma.med.auth.gr/Patient/Index

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Αποσύνδεση

Ασθενείς

Νέος Ασθενής | Εξαγωγή όλων

1. Δημιουργήστε Νέο Ασθενή αν δεν υπάρχει στην παρακάτω λίστα.

2. Αν εντοπίσετε τον ασθενή στη λίστα επιλέξτε Επεξεργασία για να τροποποιήσετε τα στοιχεία του ή να προσθέσετε επίσκεψη.

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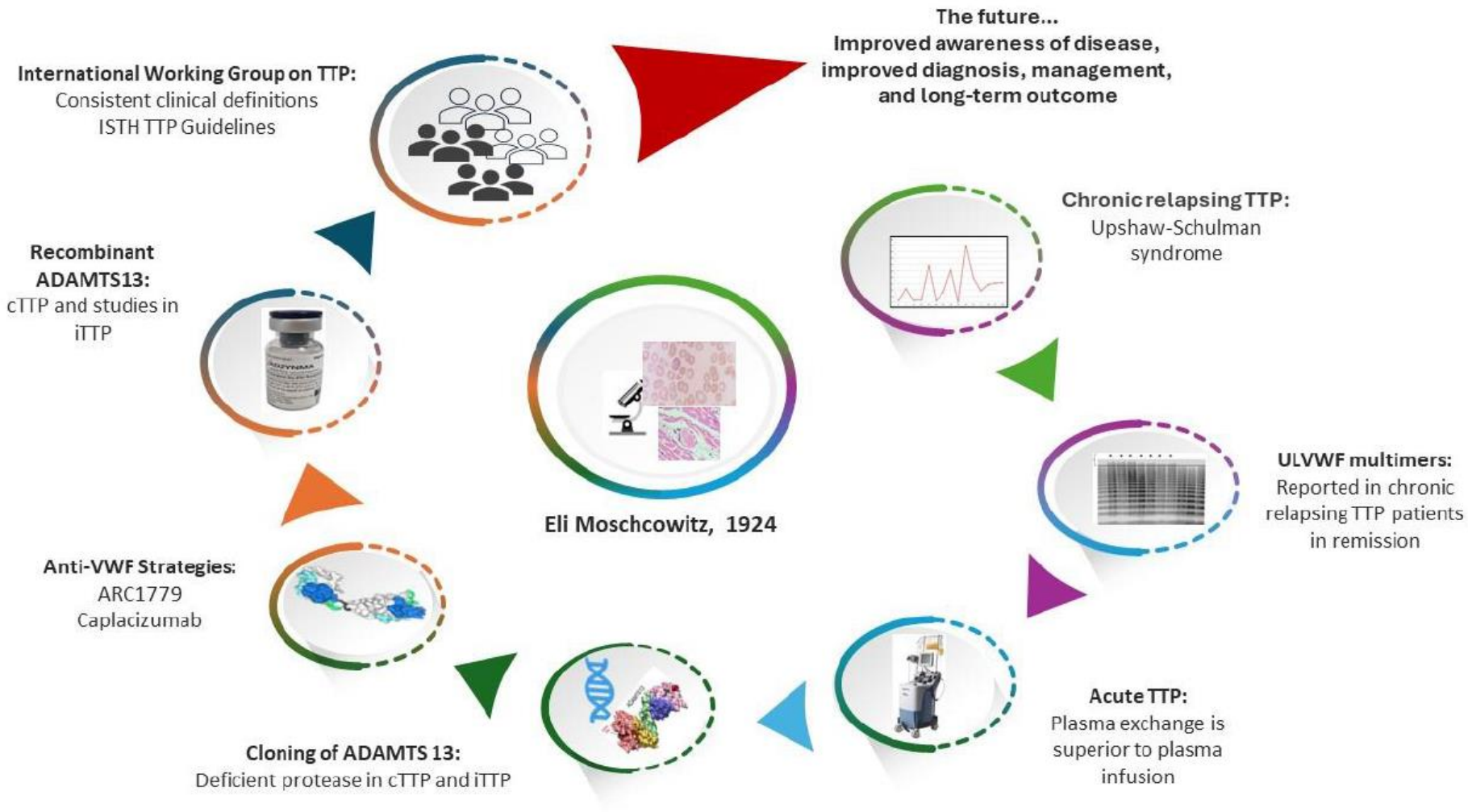
Κλινικές μελέτες στην Ελλάδα

- APPELHUS - CLNP023F12301/ Efficacy and Safety of **Iptacopan** (LNP023) in Adult Patients With Atypical Hemolytic Uremic Syndrome Naive to Complement Inhibitor Therapy (APPELHUS)

- A Phase 2b, multicenter, randomized, double-blind study of safety and efficacy of **TAK-755 (rADAMTS13)** with minimal to no plasma exchange (PEX) in the treatment of immune-mediated thrombotic thrombocytopenic purpura (iTTP)

- **Caplacizumab** and immunosuppressive therapy without first-line therapeutic plasma exchange in adults with immune-mediated thrombotic thrombocytopenic purpura (MAYARI)

Thrombotic Thrombocytopenic Purpura (TTP): 100 Years of Research on Moschcowitz's Syndrome



Cataland S, et al. Blood 2024

Ευχαριστίες

EUROPEAN HEMATOLOGY ASSOCIATION

Μητρώο εθνικής καταγραφής
των θρομβωτικών μικροαγγειοπαθειών