

Focus on the renin-angiotensin, complement, coagulation and kallikrein-kinin system in inflammation: from rare diseases to specific proteins as therapeutic targets.

Rationale

The renin-angiotensin system with its pro and anti-inflammatory effects, the complement system with C1 inhibitor that inhibits components of the kallikrein-kinin system (KKS), such as activated FXII, FXI and plasma kallikrein, as well plasmin affecting the fibrinolytic system; the coagulation and the KKS, tightly interlinked with the contact system, which is integral to the intrinsic pathway of coagulation (FXI) and thrombus formation; each of them consist of a large number of distinct plasma and membrane-bound proteins and receptors that can be activated through proteolytic cascades. Notably, all systems are excessively activated during inflammation, they have potent pro-inflammatory and prothrombotic effects and they increase vascular permeability, leading to edema.

Although the dysregulation of each protein system is involved in the pathogenesis of common disorders, only a few rare diseases such as hereditary angioedema, paroxysmal nocturnal hemoglobinuria, atypical uremic hemolytic syndrome, C3 glomerulopathies, rare coagulopathies, are approved for clinical treatment targeting renin-angiotensin, complement coagulation and KKS. The diagnosis and the treatment of these rare diseases are a challenge for health system because it is necessary to improve basic and clinical knowledge and management skills in order to offer the best response to patient's needs. In our workshop we would love to discuss the role of these complex plasma protein systems in disease pathogenesis and then share future pharmacological strategies to treat rare and common diseases with plasma contact factors and complement targeted therapies.

In order to achieve the best results within our workshop, it is of fundamental importance that everyone is an active and responsible part in our job project and in the sharing of final common choices.

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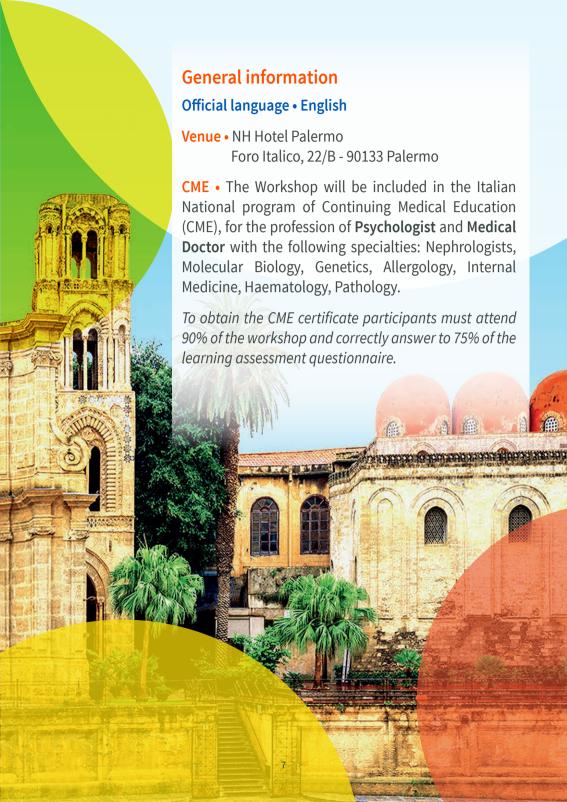
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	Frida	y, October 13 th	
	12.30	Registration and Light lunch	
	13.20	Introduction Francesco Arcoleo	
		1° WORKSHOP Chairman: Francesco Arcoleo	
	13.30	The complement system: the overture Pietro Accardo	
	14.00	Q&A	
		2° WORKSHOP Chairman: Maurizio Margaglione	(3)
	14.15	Complement genetics Santiago Rodriguez de Cordoba	1111111
700	14.45	The kallikrein-kinin system: genetics and biology Davide Firinu	17
	15.15	Q&A	
4	15.30	Coffee break	
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Saturday, October 14 th			
	4° WORKSHOP Chairman: Santiago Rodriguez de Cordoba		
09.00	Hereditary angioedema Riccardo Senter		
09.30	ITACA: a network of Italian centers involved in diagnosis and treatment of rare diseases of kallikrein-kinin system <i>Mauro Cancian</i>		
10.00	Atypical uremic syndrome Gianluigi Ardissino		
10.30	Paroxysmal nocturnal hemoglobinuria Rosario Notaro		
11.00	C3 glomerulopathy Vincenzo Montinaro		
11.30	Q&A		
11.45	Coffee break		
	5° WORKSHOP Chairman: <i>Paul Morgan</i>		
12.00	Unmet needs of rare diseases: from patient view to basic and clinical research network Massimo Triggiani		
12.30	Conformational diseases: deficit AAT model Pierpaolo Coni		
13.00	Images of rare diseases: from hereditary angioedema to acquired haemophilia Pietro Accardo		
13.30	Q&A Results of all lectures and workshops		
13.45	Round table Pietro Accardo, Francesco Arcoleo, Gianluigi Ardissino, Roberta Bulla, Mauro Cancian, Pierpaolo Coni, Davide Firinu, Claire Harris, Maurizio Margaglione, Vincenzo Montinaro, Paul Morgan, Santiago Rodriguez de Cordoba, Riccardo Senter, Massimo Triggiani		

14.00

Light lunch





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