

DATE	01/09/2022
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Part A. PERSONAL INFORMATION

Surname, forename	Goicoechea de Jorge, Elena		
ID Number/passport	14305533Q	Age	44
Researcher identification numbers	Researcher ID	L-4580-2016	
	Código ORCID	http://orcid.org/0000-0002-4978-2483	

A.1. Current position

Institution	Universidad Complutense de Madrid (UCM)		
Department / Center	Immunology, Ophthalmology and ENT/ Faculty of Medicine		
Full address	Av. Complutense S/N		
Phone	+34 665534625	email	egoicoec@ucm.es
Post/Professional Category	Lecturer in Immunology	Start date	2020
UNESCO code	240000		
Key words	Complement, genetics, factor H, FHRs, aHUS, C3G		

A.2. Education (*title, institution, date*)

Degree	Institution	Completion date
Bachelor in Biochemistry	University Autónoma de Madrid	2000
PhD in Molecular Biology	University Autónoma de Madrid	2007

A.3. Previous positions and Employments

2015-2020 Ramón y Cajal Researcher, UCM, Spain
 2011-2015 Junior Research Fellow, Imperial College London, UK
 2008-2011 Research Associate, Imperial College London, UK
 2002-2007 PhD student, Centro de Investigaciones Biológicas (CSIC), Madrid, Spain

A.4. Indicators of Quality in Scientific Production

Research "Sexenios": 3 Last: 2020
 PhD Thesis supervised: 2 (+2 in progress)
 Total Citations (ISI Web Sci): 3.336
 Total indexed Publications (ISI Web Sci): 42
 Total publications first quartile: 35 (83%)
 Average citation/year (ISI Web Sci): 201
 h-index (ISI Web Sci): 24

Part B. Free Summary of CV

Dr. Goicoechea de Jorge is a biochemist who has been working in the complement field for more than 20 years. Throughout her scientific career, she has mainly focused on the study of the complement system and its association with disease. Her interest in complement began with her doctoral training in Prof. Santiago Rodríguez de Córdoba's group, where she studied the molecular basis of atypical haemolytic uremic syndrome (aHUS), a paradigm of complement-mediated renal disease. She was the first one to demonstrate that gain-of-function mutations in complement activators are associated with aHUS (**Goicoechea de Jorge** et al. PNAS, 2007) and she developed the first animal model of aHUS in collaboration with Prof. Matthew Pickering (Pickering, **Goicoechea de Jorge** et al. J. Exp. Med. 2007).

Once completed her PhD training and motivated by the idea of studying complement dysregulation *in vivo*, she moved to Imperial College London. During this time, she completed a program of work demonstrating that complement C5 activation is critical for renal injury to develop in the aHUS model (**Goicoechea de Jorge** et al. JASN, 2011), a timely work as it paved the way to the implementation of complement therapeutics with the use of eculizumab, a

monoclonal antibody that blocks C5 activation and that has changed the natural history of aHUS. She also expanded her expertise to the study of other complement-mediated renal diseases such as C3 glomerulopathies (C3G) and identified the first mutation in complement factor H-related 5 protein (FHR-5) associated with familial C3G, leading to a publication in Lancet (Gale, **Goicoechea de Jorge**, et al., 2010). This finding opened a new research avenue in the complement field and gave her the opportunity to outline a program of work and secure a Junior Research Fellowship from Imperial College. Since then, she focused her research in understanding the biology of the FH protein family and its association with disease. She discovered the dimerization status for FHR-1, FHR-2 and FHR-5 proteins and the dramatic consequences that the duplication of the dimerization domains has in pathology (**Goicoechea de Jorge** et al. PNAS, 2013).

After seven years of a fruitful scientific period in the UK, she was awarded a Ramón y Cajal contract, and she settled as an independent investigator at the Complutense University Madrid (UCM) in 2015. Since then, she has continued working on deciphering the biological role of the FHR protein family and its association with renal diseases (i.e. atypical haemolytic uremic syndrome, C3 glomerulopathy and IgA nephropathy) representing one of the main research lines in her lab. Her main contributions in this field have provided insight into the genotype-phenotype correlations of FHR mutations associated with aHUS and C3G, and into the understanding of how disease penetrance is modulated in such complex diseases (Márquez-Tirado et al. 2022. JASN; Martín-Merineró et al. 2021. Blood; Arjona et al. Blood. 2020; **Goicoechea de Jorge** et al. 2018, JASN). During these years, she also became interested in exploring new scientific avenues and she opened a new research line in the laboratory to investigate the role of complement dysregulation in the pathogenesis of ANCA-associated vasculitis (AAV), a systemic autoimmune disorder causing inflammation of small vessels. This work has led to the identification of the first common genetic variants in genes of the complement alternative pathway associated increased susceptibility and/or severity in AAV and identified the crucial role of the FH and FHR-1 plasma levels in determining disease severity (Lucientes Continente et al. JASN, submitted).

Since her position at UCM, she combines her research with teaching, getting a permanent position as a Lecturer in the Department of Immunology at the Faculty of Medicine in 2020. Her teaching activities are directed to both undergraduate and postgraduate students. She participates in several master's degrees, where she has also taken the advantage to develop a new subject "Complement Physiopathology", and she has supervised several PhD, MSc and Bsc students. Since 2019, Dr. Goicoechea de Jorge is a member of the board of the European Complement Network.

Parte C. Accomplishments (Last 10 years)

C.1. Publications

- 1 Márquez-Tirado B; Gutiérrez-Tenorio J; et al; Goicoechea de Jorge E. 2022. Factor H-related protein 1 drives disease susceptibility and prognosis in C3 glomerulopathy. J Am Soc Nephrol. 33(6):pp.1137-1153.
- 2 Poppelaars F; Goicoechea de Jorge E; Jongerius I; et al; SciFiMed Consortium. 2021. A Family Affair: Addressing the Challenges of Factor H and the Related Proteins. Frontiers in Immunology. PMID: 33868311, pp.DOI: 10.3389/fimmu.2021.660194.
- 3 Caravaca-Fontán F; Trijillo F; Alonso M; et al; Praga M. 2021. Validation of a Histologic Scoring Index for C3 Glomerulopathy. American Journal of Kidney Diseases. Elsevier. 77-5, pp.684-695.
- 4 Gyapon-Quast; Goicoechea de Jorge E; Malik T; et al; Pickering MC. 2021. Defining the Glycosaminoglycan Interactions of Complement Factor H-Related Protein 5. Journal of Immunology. PMID: 34193601, pp.DOI: 10.4049/jimmunol.2000072.
- 5 Martín -Merineró H; Subías M; Pereda A; et al; Rodríguez de Córdoba S. 2021. Molecular bases for the association of FHR-1 with atypical hemolytic uremic syndrome and other diseases. Blood. 137, pp.3484-3494.
- 6 Caravaca-Fontán F; Díaz-Encarnación M; et al; Praga M. 2021. Longitudinal change in proteinuria and kidney outcomes in C3 glomerulopathy. Nephrol Dial Transplant. Mar 29;gfab075. doi: 10.1093/ndt/gfab075. Online ahead of print.
- 7 Mycophenolate Mofetil in C3 Glomerulopathy and Pathogenic Drivers of the Disease. 2020. Clinical Journal of the American Society of Nephrology. 15, pp.1-12.

- 8 Arjona E; Huertas A; Goicoechea de Jorge; Rodríguez de Córdoba S. 2020. The familial risk of developing atypical Hemolytic Uremic Syndrome Blood. doi: 10.1182/blood.2.
- 9 Gimenez-Moyano S; Villacorta J; Lucientes L; et al; Fernandez-Juarez G. 2020. Urinary soluble CD163 as a biomarker of disease activity and relapse in antineutrophil cytoplasm antibody-associated glomerulonephritis Clinical Kidney Journal. 1, pp.1-8.
- 10 Elena Goicoechea de Jorge; Agustín Tortajada; Sheila Pinto García; et al; Santiago Rodríguez de Córdoba. 2018. Factor H Competitor Generated by Gene Conversion Events Associates with Atypical Hemolytic Uremic Syndrome. Journal of the American Society of Nephrology. 29-1, pp.240-249.
- 11 Goicoechea de Jorge E; Lopez Lera A; Bayarri-Olmos R; Yebenes H; Lopez-Trascasa M; Rodríguez de Córdoba S. (1/6). 2018. Common and Rare Genetic Variants of Complement Components In Human Disease Molecular Immunology. 102, pp.42-57.
- 12 Goicoechea de Jorge E; Yebenes H; Serna M; Tortajada A; Llorca O; Rodríguez de Córdoba S. (1/6). 2018. How novel structures inform understanding of complement function. Seminars in Immunopathology. 40-1, pp.3-14.
- 13 Agustín Tortajada; Eduardo Gutiérrez; Elena Goicoechea de Jorge; et al; Santiago Rodríguez de Córdoba. 2017. Elevated factor H-related protein 1 and factor H pathogenic variants decrease complement regulation in IgA nephropathy. Kidney International. 92-4, pp.953-963.
- 14 Csicsi, A.I.; Kopp, A.; Zoldi, M.; et al; Józsi, M.2015. Factor H-related protein 5 (CFHR5) interacts with pentraxin 3 and the extracellular matrix and modulates complement activation. Journal of Immunology. 194-10, pp.4963-4973.
- 15 Joseph Caesar; Hayley Lavender; Philip Ward; et al;. 2014. Competition between antagonistic complement factors for a single protein on N. meningitidis rules disease susceptibility. eLife. 3-e04008. 2
- 16 MC Pickering; VD D'Agati; CM Nester; et al;. 2013. C3 glomerulopathy: consensus report. Kidney Int. 84-6, pp.1079-1089.
- 17 Goicoechea de Jorge E; Caesar JJ; Malik TH; et al; Lea S. 2013. Dimerization of complement factor H-related proteins modulates complement activation in vivo. Proc Natl Acad Sci U S A. 110-12, pp.4685-4690.
- 18 TH Malik; PJ Lavin; Elena Goicoechea de Jorge; et al; Pickering MC. 2012. A hybrid CFHR3-1 gene causes familial C3 glomerulopathy. J Am Soc Nephrol. 23-7, pp.1155-1160.
- 19 Vernon KA; Goicoechea de Jorge E; Hall AE; et al; Pickering MC. 2012. Acute presentation and persistent glomerulonephritis following streptococcal infection in a patient with heterozygous complement factor H-related protein 5 deficiency. Am J Kidney Dis. 60-1, pp.121-125.
- 20 S Johnson; L Tan; S van der Veen; et al; Tang C. 2012. Design and evaluation of meningococcal vaccines through structure-based modification of host and pathogen molecules. PLoS Pathog. 8-10, pp.e1002981.

C.2. Research Projects and Grants

- 1 Screening of inFLammation to enable personalized Medicine; SciFiMed (ID 899163). Unión Europea (Horizon 2020). Principal investigator: Elena Goicoechea de Jorge. UCM. 01/01/2021-31/12/2024. 334.875 €.
- 2 Deciphering the molecular basis of complement-mediated renal diseases. (RTI2018-095955-B-I00). Ministerio de Ciencia, Innovación y Universidades. Principal Investigator: Elena Goicoechea de Jorge. UCM. 01/01/2019- 31/12/2021. 121.000 €.
- 3 The role of microRNAs in complement-mediated renal disease. Ministerio de Economía y Competitividad. Principal Investigator: Elena Goicoechea de Jorge. UCM. 2015-2018. 157.300 €.
- 4 Deciphering the molecular basis of atypical hemolytic syndrome and C3 glomerulopathy. Fundación Inocente Inocente. Universidad Complutense de Madrid. Principal Investigator: Elena Goicoechea de Jorge. UCM. 2017-2019. 27.960 €.
- 5 The complement system in health and disease. Ayudas para la realización de Programas de Actividades de I+D entre Grupos de Investigación de la Comunidad de Madrid. Comunidad de Madrid. UCM. Team member. 2018-2021. 828.091,87 €.
- 6 Complement in health and disease. Redes de Excelencia. MINECO/AEI. Universidad Complutense de Madrid. Collaborator. 2017-2019. 20.000 €.

C3. Patents

Susan Lea; Matthew Pickering; Elena Goicoechea de Jorge. PCT/GB2014/050258. Patent in Complement System Modulators Reino Unido. 30/01/2014. Imperial College London (Imperial Innovations).

C4. Education, translation and transfer activities.

During the last years, due to my positions at the Faculty of Medicine at UCM, I combine my research activity with teaching. I actively participate in both undergraduate and postgraduate courses in Immunology. Postgraduate courses include Master's Degree in Translational Medicine (UCM), Master's Degree in Molecular Biomedicine (UAM) and Master's Degree in Immunology Research (UCM). In the latter, I co-direct the course "Complement Pathology", which gives the opportunity to teach the students with cutting-edge knowledge. In addition, I also participate in specialized courses in renal diseases given in different Spanish Hospitals. Finally, I have trained several PhD, BSc and MSc students. PhD thesis projects:

1. *Title*: "Bases Moleculares del Síndrome Hemolítico Urémico Atípico y Glomerulopatías de C3: Implicación de las proteínas relacionadas con Factor H en la patogénesis y estudio de microARNs como biomarcadores"

Institution: Universidad Complutense de Madrid

Student: Josué Gutiérrez Tenorio

Calification: "Sobresaliente Cum Laude por unanimidad"

Date of defense: 02/10/2020 *Link*: <https://eprints.ucm.es/id/eprint/49549/>

2. *Title*: "Trastornos moleculares y genéticos en las glomerulopatías causadas por desregulación del complemento. Correlaciones genotipo-fenotipo e implicaciones terapéuticas"

Thesis co-directors: Elena Goicoechea de Jorge; Manuel Praga Terente

Institution: Universidad Complutense de Madrid

Student: Fernando Caravaca Fontán

Calification: "Sobresaliente Cum Laude por unanimidad"

Date of defense: 11/11/2020 *Link*: <https://eprints.ucm.es/id/eprint/67010/>

C5. Awards and Honors

2022-Onwards. Elected secretary of the "European Complement Network".

(<http://ecomplement.org/>).

2019-2022. Board member of the "European Complement Network".

2019- Onwards. Advisory board member for the european proyect "CORVOS". H2020, Marie Skłodowska-Curie actions.

2019. Member of the LSC of the "17th European Meeting on Complement in Human Disease" (<http://www.emchd2019.com/>).

2019. Certification "I3 program".

2015-2020. Ramón y Cajal Researcher Contract (UCM).

2011-2015. Junior Research Fellow (Imperial College London).