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| CV Date | 12/09/2024 |
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Part A. PERSONAL INFORMATION

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|--|-----------------------------|---------------|--|
| First Name | Cristina | | |
| Family Name | Ugalde Bilbao | | |
| Sex | Female | Date of Birth | |
| ID number Social Security, Passport | | | |
| URL Web | | | |
| Email Address | cristina.ugalde@cib.csic.es | | |
| Open Researcher and Contributor ID (ORCID) | 0000-0002-9742-1877 | | |

A.1. Current position

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|---------------------|--|--------------|--|
| Job Title | Tenured Scientist | | |
| Starting date | 2023 | | |
| Institution | Spanish National Research Council (CSIC) | | |
| Department / Centre | Molecular Biomedicine / Centro de Investigaciones Biológicas Margarita Salas (CIB) | | |
| Country | | Phone Number | |
| Keywords | Cell physiology; Molecular mechanism of disease; Molecular, cellular and genetic biology | | |

A.2. Previous positions (Research Career breaks included)

| Period | Job Title / Name of Employer / Country |
|-------------|---|
| 2011 - 2023 | I3 Researcher of the National Health System / ISCIII / FUNDACIÓN DE INVESTIGACIÓN BIOMÉDICA HOSPITAL 12 DE OCTUBRE |
| 2018 - 2018 | Guest Researcher / MONASH UNIVERSITY (Melbourne, Australia) |
| 2005 - 2011 | Miguel Servet Researcher of the National Health System / ISCIII / FUNDACIÓN DE INVESTIGACIÓN BIOMÉDICA DEL HOSPITAL 12 DE OCTUBRE |

A.3. Education

| Degree/Master/PhD | University / Country | Year |
|--|--------------------------------|------|
| Doctorate in Biochemistry | Universidad Autónoma de Madrid | 1998 |
| Licenced in Biology - Biochemistry and Molecular Biology Section | Universidad Autónoma de Madrid | 1993 |

Part B. CV SUMMARY

Since December 2023 I work as **Tenured Researcher** at the **Spanish National Research Council (CSIC)**. My previous scientific track as an independent senior researcher started in 2005, when I obtained a Miguel Servet Research Contract (CP04/00011) and became Principal Investigator in the Laboratory of Rare, Mitochondrial and Neuromuscular Diseases (LERMN) at the 'Hospital 12 de Octubre' Research Institute (i+12, Madrid). In 2011 I became an I3 Researcher of the National Health System (BOE-A-2011-4033), position that I maintained until my incorporation at CSIC. In addition, I am linked to the CIBER for Rare Diseases (CIBERER) since its establishment in 2006. Since my incorporation at the LERMN lab I have obtained **numerous funding sources** for scientific projects from both national and international public agencies, as well as human resources. As a PI, I have concatenated seven individual research projects granted by ISCIII (PI05/0379, PI08/0021, PI11/00182, PI14/00209, PI17/00048, PI20/00057 and PI23/00169), to i) study the pathophysiological consequences of mitochondrial disorders, and ii) identify/characterize new protein biomarkers to improve their diagnosis. I have been co-PI of two interdisciplinary coordinated projects

from Comunidad Autónoma de Madrid (CAM, S2010/BMD-2361 and P2018/BAA-4403), to develop new technologies for the study of mitochondrial disorders. I have also been co-PI of two international projects granted by the National Institutes of Health (NIGMS-NIH, USA; grants 1R01GM105781-01 and R35GM118141), in collaboration with Prof. A. Barrientos (University of Miami). One of them is currently ongoing through a stable consortium, and is focused on the biogenetic regulation of mitochondrial supercomplexes using genetically-edited human cellular models of mitochondrial disease. Finally, in 2018 I was recipient of a mobility grant (ISCIII BA17/00006) for a 1-yr sabbatical stay as a Guest Researcher at Prof. M. Ryan's lab (Monash University, Australia).

My **research interests** focus on the molecular mechanisms that regulate the biogenesis and function of the human mitochondrial OXPHOS system in health and disease, from both basic and translational perspectives. I have extensive experience in the application of genetic-molecular and biochemical-proteomic techniques, as well as in mitochondrial biogenesis, genetics, metabolism and bioenergetics, in the context of both basic research and applied to the direct diagnosis of mitochondrial pathologies in patients. During my scientific career I have published a total of **56 articles and 4 book chapters**. It is worth highlighting my most **recent contributions as corresponding author in first-decile scientific journals**, among them: three articles published in Cell Metabolism (FI 2022: 31,373), one in Trends in Biochemical Sciences (FI 2022: 14,264), two in EMBO Journal (FI 2022: 14,012), three in Cell Reports (FI 2022: 9,995), one in Free Radical Biology & Medicine (FI 2022: 8,101) and one in Semin Cell Dev Biol (FI 2022: 7,727). I have an H index of 32 and more than 3800 citations. Likewise, I have been an invited speaker at numerous (inter)national conferences, as well as at institutional seminars of various institutions in Europe, USA and Australia.

Over the last 10 years, my group has consisted on average of 1 postdoc, 2 predocs and a technician. **Academically**, I have supervised **10 doctoral theses**, 2 of them currently ongoing, as well as several University Master projects. Since 2005 I am a **regular grant reviewer** for national agencies including ANECA, ISCIII, DEVA or BIOEF, as well as for international research agencies, such as Agence Nationale de la Recherche (ANR, France), Czech Science Foundation, Knut and Alice Wallenberg Foundation (Suecia) o Princess Beatrix Fonds (Netherlands). Since 2020 I belong to the **Editorial Board** of the journals Frontiers in Physiology and Frontiers in Cell and Developmental Biology, and I am also regular reviewer of high-ranking scientific journals, including Cell Metabolism, Science or Nature. Finally, I am a Founder/Executive Board Member of the European Society for Mitochondrial Research and Medicine (E-mit, www.emit.org).

Part C. RELEVANT ACCOMPLISHMENTS

C.1. Most important publications in national or international peer-reviewed journals, books and conferences

AC: corresponding author. (n° x / n° y): position / total authors. If applicable, indicate the number of citations

- 1 Scientific paper.** Fernández-Vizarra E.; López-Calcerrada S.; Sierra-Magro A.; et al; (19/19) Ugalde C. (AC). 2022. Two independent respiratory chains adapt OXPHOS performance to glycolytic switch. Cell Metabolism. 34-11, pp.1792-1808. Google Scholar (16) <https://doi.org/10.1016/j.cmet.2022.09.005>
- 2 Scientific paper.** Rafael Pérez Pérez; Teresa Lobo Jarne; Dusanka Milenkovic; et al; (14/14) Cristina Ugalde (AC). 2016. COX7A2L Is a Mitochondrial Complex III Binding Protein that Stabilizes the III2+IV Supercomplex without Affecting Respirasome Formation. Cell Reports. 16-9, pp.2387-2398. Google Scholar (107) <https://doi.org/10.1016/j.celrep.2016.07.081>
- 3 Scientific paper.** Baden P; Perez M; Kalb S; et al; Deleidi M; (14/15) Ugalde C. 2023. Glucocerebrosidase, a Parkinson's disease-associated protein, is imported into mitochondria and regulates complex I assembly and function. Nature Communications. Accepted. <https://doi.org/10.1038/s41467-023-37454-4>

- 4 **Scientific paper.** Erika Fernandez-Vizarra; (2/2) Cristina Ugalde (AC). 2022. Cooperative assembly of the mitochondrial respiratory chain. Trends in Biochemical Sciences. 47-12, pp.999-1008. Google Scholar (23) <https://doi.org/10.1016/j.tibs.2022.07.005>
- 5 **Scientific paper.** María Illescas; Ana Peñas; Joaquín Arenas; Miguel A Martín; (5/5) Cristina Ugalde (AC). 2021. Regulation of Mitochondrial Function by the Actin Cytoskeleton. Frontiers in Cell and Developmental Biology. 9-(12), pp.795838. Google Scholar (34) <https://doi.org/10.3389/fcell.2021.795838>
- 6 **Scientific paper.** Ana Peñas; Miguel Fernández de la Torre; Sara Laine-Menéndez; et al; (12/12) Cristina Ugalde (AC). 2021. Plasma Gelsolin Reinforces the Diagnostic Value of FGF-21 and GDF-15 for Mitochondrial Disorders. International Journal of Molecular Sciences. 22-(12), pp.6396. Google Scholar (8) <https://doi.org/10.3390/ijms22126396>
- 7 **Scientific paper.** Erika Fernandez-Vizarra; Sandra López-Calcerrada; Luke E. Formosa; et al; (11/11) Cristina Ugalde (AC). 2021. SILAC-based complexome profiling dissects the structural organization of the human respiratory supercomplexes in SCAFI-KO cells. BBA - Bioenergetics. 1862-7, pp.148414. Google Scholar (13) <https://doi.org/10.1016/j.bbabi.2021.148414>
- 8 **Scientific paper.** Alberto García-Bartolomé; Ana Peñas; María Illescas; et al; (11/11) Cristina Ugalde (AC). 2020. Altered Expression Ratio of Actin-Binding Gelsolin Isoforms Is a Novel Hallmark of Mitochondrial OXPHOS Dysfunction. Cells. 9(9):1922.. Google Scholar (10) <https://doi.org/10.3390/cells9091922>
- 9 **Scientific paper.** Alba Timón-Gómez; Joshua Garlich; Rosemary A Stuart; (4/5) Cristina Ugalde; Antoni Barrientos. 2020. Distinct Roles of Mitochondrial HIGD1A and HIGD2A in Respiratory Complex and Supercomplex Biogenesis. Cell Reports. 31(5):107607. ISSN 1460-2075. Google Scholar (46) <https://doi.org/10.1016/j.celrep.2020.107607>
- 10 **Scientific paper.** Teresa Lobo-Jarne; Rafael Pérez-Pérez; Flavia Fontanesi; et al; (12/12) Cristina Ugalde (AC). 2020. Multiple pathways coordinate human complex IV assembly and respiratory supercomplexes stabilization. EMBO Journal. e103912. ISSN 1460-2075. Google Scholar (64) <https://doi.org/10.15252/emboj.2019103912>
- 11 **Scientific paper.** Alba Timón-Gómez; Rafael Pérez-Pérez; Eva Nyvltova; Flavia Fontanesi; (5/6) Cristina Ugalde; Antoni Barrientos. 2020. Protocol for the Analysis of Yeast and Human Mitochondrial Respiratory Chain Complexes and Supercomplexes by Blue Native Electrophoresis. STAR Protocols. 1(2):100089.. ISSN 1460-2075. Google Scholar (17) <https://doi.org/10.1016/j.xpro.2020.100089>
- 12 **Scientific paper.** Marguerita Protasoni; Rafael Pérez-Pérez; Teresa Lobo-Jarne; et al; Erika Fernández-Vizarra; (11/12) Cristina Ugalde (AC). 2020. Respiratory supercomplexes act as a platform for complex III-mediated maturation of human mitochondrial complexes I and IV. EMBO Journal. e102817. ISSN 1460-2075. Google Scholar (122) <https://doi.org/10.15252/emboj.2019102817>
- 13 **Scientific paper.** Susana Peralta; Adrián González-Quintana; Marta Ybarra; et al; Miguel A. Martín; (9/10) Cristina Ugalde (AC). 2019. Novel ATAD3A recessive mutation associated to fatal cerebellar hypoplasia with multiorgan involvement and mitochondrial structural abnormalities. MOL GENET METAB. 128-4, pp.452-462. Google Scholar (22) <https://doi.org/10.1016/j.ymgme.2019.10.012>
- 14 **Scientific paper.** Teresa Lobo Jarne; Eva Nývltová; Rafael Pérez Pérez; et al; Antoni Barrientos; (9/10) Cristina Ugalde (AC). 2018. Human COX7A2L Regulates Complex III Biogenesis and Promotes Supercomplex Organization Remodeling without Affecting Mitochondrial Bioenergetics. Cell Reports. 25-7, pp.1786-1799. ISSN 2211-1247. Google Scholar (58) <https://doi.org/10.1016/j.celrep.2018.10.058>
- 15 **Scientific paper.** Lobo-Jarne T.; (2/2) Ugalde C. (AC). 2018. Respiratory Chain Supercomplexes: Structures, Function and Biogenesis. Semin Cell Dev Biol. 76, pp.179-190. Google Scholar (160) <https://doi.org/10.1016/j.semcdb.2017.07.021>
- 16 **Scientific paper.** Alberto García-Bartolomé; Ana Peñas; Lorena Marín-Buera; et al; (9/9) Cristina Ugalde (AC). 2017. Respiratory chain enzyme deficiency induces mitochondrial location of actin-binding Gelsolin to modulate the oligomerization of VDAC complexes and cell survival. Hum Mol Genet. 26-13, pp.2493-2506. Google Scholar (9) <https://doi.org/10.1093/hmg/ddx144>

- 17 **Scientific paper.** Marín-Buera L.; García-Bartolomé A.; Morán M.; et al; Ugalde C. 2015. Differential proteomic profiling unveils new molecular mechanisms associated with mitochondrial complex III deficiency. *J Proteomics*. 113, pp.38-56. Google Scholar (21)
- 18 **Scientific paper.** Barrientos A; (2/2) Ugalde C (AC). 2013. I function, therefore I am: overcoming skepticism about mitochondrial supercomplexes. *Cell Metab*. 18-2, pp.147-149. Google Scholar (41)
- 19 **Scientific paper.** Moreno-Lastres D; Fontanesi F; García-Consuegra I; Martín MA; Arenas J; Barrientos A; (7/7) Ugalde C (AC). 2012. Mitochondrial complex I plays an essential role in human respirasome assembly. *Cell Metab*. 15-3, pp.324-335. Google Scholar (303)
- 20 **Scientific paper.** Morán M; Moreno-Lastres D; Marín-Buera L; Arenas J; Martín MA; Ugalde C. 2012. Mitochondrial respiratory chain dysfunction: implications in neurodegeneration. *Free Radic. Biol. Med*. 53-3, pp.595-609. Google Scholar (162)
- 21 **Book chapter.** Blázquez A; Marín-Buera L; Morán M; García-Bartolomé A; Arenas J; Martín MA; Ugalde C. 2013. Mitochondrial complex III deficiency of nuclear origin: molecular basis, pathophysiological mechanisms and animal models. In *Mitochondrial disorders caused by nuclear genes* (Lee-Jun Wong, editor). Springer Science+Business media, LLC. New York, USA.. XII, pp.219-238.

C.3. Research projects and contracts

- 1 **Project.** PI23/00169 - ENCEFALOMIOPATÍA MITOCONDRIAL PEDIÁTRICA: APROXIMACIONES DIAGNÓSTICO-TERAPEUTICAS. Instituto de Salud Carlos III. (Instituto de Investigación Hospital 12 de Octubre (i+12)). 01/01/2024-31/12/2026. 252.500 €.
- 2 **Project.** 1R35 GM118141 - Mitochondrial Biogenesis in Health and Disease. National Institutes of Health (NIH), EEUU. Cristina Ugalde Bilbao. (FIB Hospital 12 de Octubre (Madrid) / Miami University (USA)). 01/06/2021-31/05/2026. 912.879 €.
- 3 **Project.** PI20/00057 - New tools for the diagnosis and treatment of mitochondrial OXPHOS disorders. Instituto de Salud Carlos III. (Instituto de Investigación Hospital 12 de Octubre (i+12)). 01/01/2021-31/12/2023. 147.620 €.
- 4 **Project.** S2018/BAA- 4403-SINOXPHOS: Soluciones interdisciplinares con control de edición génica al déficit bioenergético OXPHOS. Consejería de Educación de la Comunidad Autónoma de Madrid. Iván López Montero. (Instituto de Investigación Hospital 12 de Octubre (i+12)). 01/01/2019-31/12/2022. 719.900 €.
- 5 **Project.** PI17/00048 - Desarrollo de una plataforma de diagnóstico no invasivo para las enfermedades OXPHOS basada en la detección de nuevos biomarcadores proteicos implicados en la disfunción mitocondrial. Instituto de Salud Carlos III. Cristina Ugalde Bilbao. (Instituto de Investigación Hospital 12 de Octubre (i+12)). 01/01/2018-31/12/2020. 212.650 €.
- 6 **Project.** BA17/00006 - Identificación de nuevos genes asociados a patologías mitocondriales OXPHOS mediante el desarrollo combinado de tecnologías CRISPR/Cas9 y espectrometría de masas cuantitativa. Instituto de Salud Carlos III. Cristina Ugalde Bilbao. (Monash University). 01/01/2018-31/12/2018. 41.970 €. Principal investigator.
- 7 **Project.** PI14/00209 - Búsqueda y caracterización de biomarcadores de enfermedades mitocondriales asociadas a déficits enzimáticos del sistema OXPHOS. Instituto de Salud Carlos III. Cristina Ugalde Bilbao. (FIB Hospital 12 de Octubre). 01/01/2015-31/12/2017. 224.150 €.
- 8 **Project.** 1R01GM105781-01 - The Biosynthetic Pathway of Mitochondrial Respirasomes (1R01GM105781-01). National Institutes of Health (NIH), EEUU. Cristina Ugalde Bilbao. (FIB Hospital 12 de Octubre (Madrid) / Miami University(USA)). 01/05/2014-31/12/2017. 1.180.630 €.