Valeria Guglielmi

Curriculum vitae

Personal information

First name/Last name: Valeria Guglielmi
Date and place of birth: 4th October 1984, Verona, Italy.
Citizenship: Italian
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Education

- PhD in Neuroscience, Department of Neurological and Movement Sciences, University of Verona, Italy. 7th May 2013. Thesis title: "Biochemical features of SERCA1 in Brody disease and identification of candidate genes in Brody syndrome".
- Master's degree in Molecular and Industrial Biotechnology, with honors, University of Verona, Italy. 19th March 2009. Thesis title: "The role of programmed cell death in mitochondrial diseases".
- Degree in Agro-Industrial Biotechnology, with honors, University of Verona, Italy. 27th October 2006. Thesis title: "Lipid Transfer Proteins and their defense role from pathogens in plants".

Research experiences

- March 2016 to present: Postdoctoral Fellow, Department of Neurosciences, Biomedicine and Movement Sciences, Section of Anatomy and Histology, University of Verona, Italy. Advisors: Dr. Gaetano Vattemi, Prof. Manuela Malatesta.
- July 2014 - June 2015: Postdoctoral Fellow, Department of Neurology, Houston Methodist Research Institute, Houston, TX, USA. Advisor: Prof. Stanley H. Appel.
- January 2013 - June 2014: Postdoctoral Fellow, Department of Neurological and Movement Sciences, Section of Clinical Neurology, Laboratory of Neuropathology, University of Verona, Italy. Advisor: Dr. Gaetano Vattemi.
- January 2010 - December 2012: PhD student in Neuroscience, Department of Neurological and Movement Sciences, Section of Clinical Neurology, Laboratory of Neuropathology, University of Verona, Italy. Advisor: Dr. Gaetano Vattemi.
- May 2009 - December 2009: Research fellow, Department of Neurological and Movement Sciences, Section of Clinical Neurology, Laboratory of Neuropathology, University of Verona. Advisor: Dr. Giuliano Tomelleri.
- April 2008 - March 2009: Undergraduate student, Department of Neurological and Movement Sciences, Section of Clinical Neurology, Laboratory of Neuropathology, University of Verona, Italy. Advisors: Dr. Roberto Chignola, Dr. Gaetano Vattemi

Other research activities

- Visiting postdoctoral fellow, University of Brescia, Department of Molecular and Translational Medicine, Section of Oncology and Experimental Immunology Division. July 2017 - May2018 (not on daily basis).
- Visiting postdoctoral fellow within the 7PR21/BASTION/WP1 (Twinning-WP1.9) at the Department of Immunology, Center for Biostructure Research, Medical University of Warsaw, Warsaw, Poland. 26th October 2013- 23rd November 2013.
- Visiting PhD student, Department of Biotechnology, Laboratory of Genetic Biotechnologies, University of Verona, Italy. November 2011 - June 2012.

Scientific Activities and Interests

Mitochondrial diseases

- Characterization of the cell death program occurring in *ragged red fibers* (RRFs) and identification of the repairing event ongoing in skeletal muscle of patients with mitochondrial disorders.
- Identification of nitrated proteins and individuation of vessel wall as target of oxidative and nitric oxide stress in skeletal muscle of patients with mitochondrial respiratory chain dysfunction.
- Study of the expression and localization of TNF-α, and its receptors TNFR1 and TNFR2, in skeletal muscles of patients suffering from mitochondrial diseases and investigation of their functional role in the pathophysiology of mitochondria function/dysfunction.

Brody myopathy

- Study of the SERCA1 expression in skeletal muscle of patients affected by Brody myopathy and characterization of some biochemical features of this protein that can be useful in the diagnosis of the disease.
- Identification of putative SERCA1-binding proteins and evaluation their expression and functional activity in patients' muscle to provide new candidate proteins involved the pathogenesis of Brody myopathy, especially in those cases with unknown causative gene (Brody syndrome).

Myofibrillar myopathy

- Characterization of the expression and subcellular localization of RNA polymerase II associated proteins (RPAPs) in muscle of patients with myofibrillar myopathies (MFMs).
- Study of the effects of mutations in human myotilin on the structure and function of skeletal muscle in zebrafish.

Myotonic dystrophy

- Study of the expression, fiber distribution, and function of SERCA1 and SERCA2 in muscle of patients with myotonic dystrophy type1, type2 and hypothyroid myopathy.
- Study of poly(lactic-co-glycolic acid) (PLGA) nanoparticles as a delivery system for pentamidine for the treatment of myotonic dystrophy type 1 using primary myoblasts obtained from patients' muscle as an *in vitro* model of the disease.

Nanoparticles as drug delivery system in skeletal muscle disorders.

Evaluation of the biocompatibility, mechanisms of internalization, intracellular distribution, permanence and degradation of different types of nanoparticles i.e. polymeric poly(lactic-co-glycolic acid) (PLGA) NPs, mesoporous silica nanoparticles (MSN) and liposomes on primary human myoblasts and myotubes.

Bortezomib muscle toxicity

Identification of skeletal muscle toxicity of bortezomib, a proteasome inhibitor approved for the treatment of multiple myeloma patients in first lane, and study of the cytotoxicity and the biological effects of this drug on primary human myoblasts.

Inflammatory myopathies

Identification of putative target antigens specific of dermatomyositis and of polymyositis and definition of serological profiles in patients with idiopathic inflammatory myopathies.

Amyotrophic Lateral Sclerosis

- Evaluation of the efficacy of cells belonging to the monocyte/macrophage lineage as a therapeutic agent for ALS using the transgenic mSOD1^{G93A} mice.
- Investigation of the influence of some protein factors on the suppression activity of human regulatory T cells.

Technical skills

Molecular biology: Isolation and quantification of proteins and nucleic acids. SDS-PAGE, 2D-PAGE and native electrophoresis. Western blot. Immunoprecipitation. PCR and molecular cloning. Agarose gel electrophoresis. Transformation of bacteria. Real-time PCR.

Cell Biology: Cell cultures techniques. Isolation and culture of primary human myoblasts. PBMC isolation from human blood. Human monocytes, T cells and regulatory T cells isolation. Culture of human T lymphocytes including regulatory T lymphocytes. Differentiation of primary murine bone marrow-derived macrophages. Cell viability assays. *In vitro* Treg suppression assay. Preparation of samples for flow cytometry. Use of flow cytometer.

Histology: Immunohistochemistry and immunofluorescence. Use of optical microscope (bright field and fluorescence). Freezing tissues for cryosectioning and histological/immunohistochemical analysis. Laser capture microdissection. Preparation of sample for transmission electron microscopy (TEM) and immunoelectron microscopy. Whole-mount immunofluorescence and whole-mount in situ hybridization on zebrafish embryos.

Animal models: Mouse handling techniques. Mouse transcardial perfusion and tissues collection. Intraperitoneal injection. Blood collection from tail vein. Basic experience on zebrafish maintenance and breeding. Microinjection in one-cell stage zebrafish embryos.

Language skills

- Native language: Italian.
- English: good in reading, speaking and writing.

Computer skills

The knowledge of Windows, Office applications (Word, Excel and Power Point), Adobe Photoshop, GIMP 2, Adobe Illustrator, Image J, Axion Vision, Image Pro Plus, Leica Application Suite Advanced Fluorescence (LAS AF), Vector NTI, Image Master and BD FACSDiva software. Ability to use the main databases for DNA and protein sequence analysis.

Congress Attendance

- XXVII AINI (Italian Association of Neuroimmunology) Congress. 07 - 10 May 2018, Trieste, Italy.
- 13th MCM (Multinational Congress on Microscopy). 24 - 29 September 2017, Rovinj, Croatia.

- XXVI AINI (Italian Association of Neuroimmunology) Congress and 16th ESNI (European School of Neuroimmunology) Course. 26 - 30 June 2017, Venezia, Italy.
- 10° AIM (Italian Association of Myology) Congress. 03 - 05 June 2010. Milano, Italy.
- XIX AINI (Italian Association of Neuroimmunology) Congress. 01 - 04 October 2009. Porto Cervo (Arzachena, OT), Italy.
- 9° AIM (Italian Association of Myology) Congress. 11-13 June 2009. Verona, Italy.

Abstracts and oral communications

- **Guglielmi V**., Marini M., Tomelleri G., Vattemi G. Cancer-associated immune-mediated myofibrillar myopathy with anti-carbonic anhydrase III antibodies. (Poster) XXVII AINI Congress. 07 - 10 May 2018, Trieste, Italy.
- **Guglielmi V.**, Carton F., Stella B., Arpicco S., Berlier G., Marengo A., Boschi F., Cardani R., Renna L. V., Meola G., Malatesta M. Nanoparticles as drug delivery system in skeletal muscle disorders: in vitro studies of PLGA nanoparticles as carriers for pentamidine for the treatment of myotonic dystrophy type 1. (Oral communication). 13th MCM. 24 - 29 September 2017, Rovinj, Croatia.
- **Guglielmi V.** Marini M., Tomelleri G., Vattemi G. ATP synthase subunit alpha is specifically recognized by serum antibodies of a patient with selective loss of thick filaments. (Poster) XXVI AINI Congress and 16th ESNI Course. 26 - 30 June 2017, Venezia, Italy.
- **Guglielmi V.** "The role of Programmed Cell Death in mitochondrial diseases". (Oral communication). 10° AIM Congress. 03 - 05 June 2010, Milano, Italy.
- **Guglielmi V.** "Identification and characterization of muscle-specific antigens in patients affected with idiopathic inflammatory myopathies". (Oral communication). XIX AINI Congress. 01 - 04 October 2009, Porto Cervo (Arzachena, OT), Italy.
- **Guglielmi V**., Marini M., Vattemi G., Tonin P., Palmucci L., Tomelleri G. "Proteomic analysis in SERCA1 and calsequestrin storage myopathy". (Oral communication). 9° AIM Congress. 11 - 13 June 2009, Verona, Italy.

Other congress contributions

- A novel Emerin mutation in a patient with early onset Emery-Dreifuss muscular dystrophy. Bozzetti S., Vattemi G., Rimessi P., Macchione F., Pancheri E., Guglielmi V., Ferlini A., Tonin P. XVIII AIM (Italian Association of Myology) Congress. 06 - 09 June 2018, Genoa, Italy.
- **Guglielmi V.,** Gualandri F., Vattemi G., Pegoraro E., Voermans N., Marini M., Neri M., van Engelen B., Ferlini A., Tomelleri G. "SERCA expression in Brody disease". 11° AIM (Italian Association of Myology) Congress. 26 - 28 May 2011, Santa Margherita di Pula, Cagliari, Italy.
- Marini M., Vattemi G., Gualandri F., Neri M., **Guglielmi V.**, Ferlini A., Tomelleri G. 2D immunoblot: a novel diagnostic tool for myofibrillar myopathies. 11° AIM (Italian Association of Myology) Congress. 26 - 28 May 2011, Santa Margherita di Pula, Cagliari, Italy.
- Russignan A., Gualandi F., Oosterhof A., Vattemi G., Marini M., Tonin P., Rimessi P., Neri M., **Guglielmi V.**, Poli C., van Kuppevelt T.H., Ferlini A., Tomelleri G. "Pathological, biochemical and genetic features of Brody disease in a new family". 9° AIM Congress. 11 - 13 June 2009, Verona, Italy.

- Poli C., Fisher D., Vattemi G., Tonin P., Marini M., **Guglielmi V.**, Scarpelli M., Russignan A., Filosto M., Tomelleri G. "Identification and characterization of two cases of immune-mediated myofibrillar myopathy". 9° AIM Congress. 11 - 13 June 2009, Verona, Italy.
- Zappini F., Vattemi G., Tonin P., Marin M., Russignan A., Scarpelli M., **Guglielmi V.**, Filosto M., Tomelleri G. "Clinical and pathological features of a complement-mediated necrotizing myopathy". 9° AIM Congress. 11 - 13 June 2009, Verona, Italy.

Awards

SISM (Società Italiana Scienze Microscopiche) award for the participation to the 13th Multinational Congress on Microscopy. 24 - 29 September 2017. Rovinj, Croatia.

Invited talks

"Get lost in the pathogenesis of muscle diseases". Medical University of Warsaw, Warsaw, Poland. 14th November 2013.

Editorial activity

Editorial board member, Annals of Behavioral Neuroscience. From January 2018.

Professional associations

- Associazione Italiana di Neuroimmunologia (AINI). From 2009.
- Società Italiana di Scienze Microscopiche (SISM). From 2017.

Scientific publications in peer-reviewed journals

1. **Guglielmi V.**, Tomelleri G., Vattemi G. Myofibrillar myopathies through the microscope: From diagnosis to molecular pathogenesis. Microscopie 2018, 29. DOI: 10.4081/microscopie.2018.7325
2. **Guglielmi V.**, Marini M., Tomelleri G. and Vattemi G. Over Expression of NOS2 in Ragged-red Fibers from Patients with Mitochondrial Disorders due to Mutations in mtDNA. Annal Behav Neurosci, 1(1): 07-13 (2018)
3. **Guglielmi V**, Voermans NC, Oosterhof A, Nowis D, van Engelen BG, Tomelleri G, Vattemi G. Evidence of ER stress and UPR activation in patients with Brody disease and Brody syndrome. Neuropathol Appl Neurobiol. 2017 Aug 12.
4. **Guglielmi V**, Nowis D, Tinelli M, Malatesta M, Paoli L, Marini M, Manganotti P, Sadowski R, Wilczynski GM, Meneghini V, Tomelleri G, Vattemi G. Bortezomib-Induced Muscle Toxicity in Multiple Myeloma. J Neuropathol Exp Neurol. 2017 Jul 1;76(7):620-630.
5. Dalle Vedove F, Fava C, Jiang H, Zanconato G, Quilley J, Brunelli M, **Guglielmi V**, Vattemi G, Minuz P. Increased epoxyeicosatrienoic acids and reduced soluble epoxide hydrolase expression in the preeclamptic placenta. J Hypertens. 2016 Jul;34(7):1364-70.
6. **Guglielmi V**, Oosterhof A, Voermans NC et al. Characterization of sarcoplasmic reticulum Ca2+ATPase pumps in muscle of patients with myotonic dystrophy and with hypothyroid myopathy. Neuromuscul Disord. 2016 Jun;26(6):378-85.
7. **Guglielmi V**, Vattemi G, Chignola R et al. Evidence for caspase-dependent programmed cell death along with repair processes in affected skeletal muscle fibres in patients with mitochondrial disorders. Clin Sci (Lond). 2015 Nov 2.

8. Marini M, **Guglielmi V**, Faulkner G et al. Immunoblot as a potential diagnostic tool for myofibrillar myopathies. Electrophoresis. 2015 Dec;36(24):3097-100.
9. Vattemi G, Mirabella M, **Guglielmi V** et al. Muscle biopsy features of idiopathic inflammatory myopathies and differential diagnosis. Auto Immun Highlights. 2014 Sep 10;5(3):77-85.
10. **Guglielmi V**, Marini M, Masson ÉF et al. Abnormal expression of RNA polymerase II-associated proteins in muscle of patients with myofibrillar myopathies. Histopathology. 2015 Dec;67(6):859-65.
11. Cappelletti C, Galbardi B, Kapetis D, Vattemi G, **Guglielmi V**, Tonin P, Salerno F, Morandi L, Tomelleri G, Mantegazza R, Bernasconi P. Autophagy, inflammation and innate immunity in inflammatory myopathies. PLoS One. 2014 Nov 3;9(11): e111490.
12. Vattemi G, Marini M, Di Chio M, Colpani M, **Guglielmi V**, Tomelleri G. Polymyositis in solid organ transplant recipients receiving tacrolimus. J Neurol Sci. 2014 Oct 15;345(1-2):239-43.
13. **Guglielmi V**, Vattemi G, Gualandi F et al. SERCA1 protein expression in muscle of patients with Brody disease and Brody syndrome and in cultured human muscle fibers. Mol Genet Metab. 2013 Sep-Oct;110(1-2):162-9.
14. **Guglielmi V.**, Voermans N.C., Gualandi F. et al. Fourty-Four Years of Brody Disease: It is Time to Review. J Genet Syndr Gene Ther 2013, 4:181.
15. Vattemi G, Marini M, Ferreri NR, Hao S, Malatesta M, Meneguzzi A, **Guglielmi V**, Fava C, Minuz P, Tomelleri G. Overexpression of TNF-α in mitochondrial diseases caused by mutations in mtDNA: evidence for signaling through its receptors on mitochondria. Free Radic Biol Med. 2013 Apr 22;63C:108-114.
16. Vattemi G, Neri M, Marini M, Gualandi F, Tonin P, Bertolasi L, **Guglielmi V**, Catalli C, Novelli G, Ferlini A, Tomelleri G. Selective pseudohypertrophy of vastus medialis muscles associated with calpain 3 deficiency. Neurologist. 2012 Sep;18(5):306-9.
17. Voermans NC, Laan AE, Oosterhof A, van Kuppevelt TH, Drost G, Lammens M, Kamsteeg EJ, Scotton C, Gualandi F, **Guglielmi V**, van den Heuvel L, Vattemi G, van Engelen BG. Brody syndrome: A clinically heterogeneous entity distinct from Brody disease: A review of literature and a cross-sectional clinical study in 17 patients. Neuromuscul Disord. 2012 Nov;22(11):944-54.
18. Minuz P, Fava C, Vattemi G, Arcaro G, Riccadonna M, Tonin P, Meneguzzi A, Degan M, **Guglielmi V**, Lechi A, Tomelleri G. Endothelial dysfunction and increased oxidative stress in mitochondrial diseases. Clin Sci (Lond). 2012 Mar;122(6):289-97.
19. Vattemi G, Neri M, Piffer S, Vicart P, Gualandi F, Marini M, **Guglielmi V**, Filosto M, Tonin P, Ferlini A, Tomelleri G. Clinical, morphological and genetic studies in a cohort of 21 patients with myofibrillar myopathy. Acta Myol. 2011 Oct;30(2):121-6.
20. Vattemi G, Mechref Y, Marini M, Tonin P, Minuz P, Grigoli L, **Guglielmi V**, Klouckova I, Chiamulera C, Meneguzzi A, Di Chio M, Tedesco V, Lovato L, Degan M, Arcaro G, Lechi A, Novotny MV, Tomelleri G. Increased protein nitration in mitochondrial diseases: evidence for vessel wall involvement. Mol Cell Proteomics. 2011 Apr;10(4):M110.002964.
21. Vattemi G, Gualandi F, Oosterhof A, Marini M, Tonin P, Rimessi P, Neri M, **Guglielmi V**, Russignan A, Poli C, van Kuppevelt TH, Ferlini A, Tomelleri G. Brody disease: insights into biochemical features of SERCA1 and identification of a novel mutation. J Neuropathol Exp Neurol. 2010 Mar;69(3):246-52.
22. Vattemi G, Tonin P, Neri M, Marini M, Gualandi F, **Guglielmi V**, Ferlini A, Tomelleri G. Calpain 3 deficiency presenting as fibre type disproportion. Neuropathol Appl Neurobiol. 2009 Dec;35(6):614-7.

Verona, 11th July 2018