Curriculum Vitae

Riccardo Montioli, PhD

Assistant professor

Biological Chemistry Laboratory, Dep. of Neurosciences, Biomedicine and Movement sciences.

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Education & Qualifications:

2004. Graduate in Agro-Industrial Biotechnologies, 107/110, at the university of Verona, faculty of Sciences MM. FF. NN.

June 2008. Internship at the Dep. of Biochemical Sciences "A. Rossi Fanelli" of University of Rome for the acquisition of bioinformatics techniques.

23 april 2009 : Ph.D. in Biosciences *Curriculum* Biochemistry awarded at the University of Verona, Department of Morphological-Biomedical Sciences, Section of Biological Chemistry. Project title: "kinetic, spectroscopic and mutational analysis of three PLP-dependent enzymes belonging to the Fold type I. Catalytic mechanisms, folding process and dimerization"

Scientific activity

January 2010 to November 2018: Post-doc researcher at the Dep. of Neurosciences, Biomedicine and Movements, University of Verona.

December 2018 to date: Assistant Professor at the Dep. of Neurosciences, Biomedicine and Movements Sciences of University of Verona.

The scientific interests of Dr. Montioli are the structural, functional studies of enzymes involved in rare human diseases, by means of both biochemical and computational techniques. In particular, the main objectives of these studies are:

- Define the structural/functional relationships of the analyzed proteins
- Analyze the molecular defects of the pathogenic variants and associate to each patient genotype a specific enzymatic phenotype
- Clarify the molecular basis of the responsiveness to the available therapies, suggest genotype-specific treatments and new therapeutic strategies.

Technical skills and competences:

-Extensive practical laboratory skills in: (i) recombinant enzymes production and characterization by spectroscopic, chromatographic techniques; (ii) molecular biology approaches in recombinant protein engineering; (iii) enzyme kinetic and inhibition studies.

-Experienced in bioinformatics structural analysis of proteins by molecular modeling and MD simulation.

Teaching experiences

- Chemistry (Project: *Saperi Minimi*, University of Verona) degree in Physical Education and Sport Sciences, University of Verona
- Chemistry (Project: *Tandem* University of Verona)
- Biochemistry of Movement, degree in Physical Education and Sport Sciences, University of Verona
- Biochemistry, degree in Radiology Techniques for Imaging and Radiotherapy, University of Verona
- Biochemistry, degree in Nursing (VI), University of Verona

Full list of publications

1. Cellini, M. Bertoldi, **R. Montioli**, C. Borri Voltattorni "Probing the role of Tyr 64 of *Treponema denticola* cystalysin by site-directed mutagenesis and kinetic studies" *Biochemistry* (2005) 44, 13970-13980. IF 2.86
2. B. Cellini, **R. Montioli**, A. Bossi, M. Bertoldi, D. V. Laurents, C. Borri Voltattorni " Holo- and apo-cystalysin from *Treponema denticola*: two different conformations" *Arch. Biochem. Biophys.* (2006) 455(1):31-9 IF 3.31
3. B. Cellini, M. Bertoldi, **R. Montioli**, D. V. Laurents, A. Paiardini, C. Borri Voltattorni " Dimerization and folding processes of *Treponema denticola* cystalysin: the role of pyridoxal 5'-phosphate" *Biochemistry* (2006) 45(47):14140-54. IF 2.86
4. Cellini B., Bertoldi M., **Montioli R**., Paiardini A., C. Borri Voltattorni" Human wild-type alanine: glyoxylate aminotransferase and its naturally occurring G82E variant: functional properties and physiological implications" *Biochem J***.** 2007 Nov 15;408(1):39-50 IF 4.22
5. Cellini B, **Montioli R**., Bianconi S., Jorge P. Lopez-Alonso, C. Borri Voltattorni "Construction, purification and characterization of untagged human liver alanine-glyoxylate aminotransferase expressed in Escherichia coli" *Protein Pept Lett***.** 2008;15(2):153-9. IF 1.13
6. M. Bertoldi, B. Cellini, A. Paiardini, **R. Montioli**, C. Borri Voltattorni "Reaction of human liver peroxisomal alanine:glyoxylate aminotransferase with β-chloro L-alanine and L-cysteine: spectroscopic and kinetic analysis" *Biochim Biophys Acta-proteins and proteomics***.** 2008 Sep;1784(9):1356-62 IF 2.37
7. M. Bertoldi, B. Cellini, **R. Montioli**, C. Borri Voltattorni "Insights into the mechanism of the oxidative deamination catalyzed by DOPA decarboxylase" *Biochemistry.* 2008 Jul 8;47(27):7187-95 IF 2.86

8. **Montioli R**., B. Cellini, M. Bertoldi, A. Paiardini, C. Borri Voltattorni " An engineered folded PLP-bound monomer of *Treponema denticola* cystalysin reveals the effect of the dimeric structure on the catalytic properties of the enzyme**"** *Proteins.* 2009 Feb 1;74(2):304-17 IF 2.44
9. Cellini B, **Montioli R**, Paiardini A, Lorenzetto A, Voltattorni CB. "Molecular Insight into the Synergism between the Minor Allele of Human Liver Peroxisomal Alanine:Glyoxylate Aminotransferase and the F152I Mutation" *J Biol Chem***.** 2009 Mar 27;284(13) IF 3.96
10. Cellini B*, **Montioli R**, Paiardini A, Lorenzetto A, Maset F.,Bellini T., Oppici E.,Voltattorni CB. "Molecular defect of the glycine 41 variants of alanine glyoxylate aminotransferase associated with primary hyperoxaluria type I" *Proceedings of National Academy of sciences (PNAS)* U S A. 2010 Feb 16;107(7):2896-901 IF 9.35
11. Cellini B, Lorenzetto A, **Montioli R**, Oppici E and Carla Borri Voltattorni. "Human Liver Peroxysomal Alanine:glyoxylate Aminotransferase: Different Stability under Chemial Stress of the Major Allele, the Minor Allele, and its Pathogenic G170R Variant". *Biochimie* 2010 Dec;92(12):1801-11. IF 3.35
12. Cellini B, **Montioli R**, Voltattorni CB. "Human liver peroxisomal alanine:glyoxylate aminotransferase: Characterization of the two allelic forms and their pathogenic variants" *Biochim Biophys Acta-proteins and proteomics*. 2011 Nov;1814(11):1577-84. Epub 2010 Dec 20. IF 2.37
13. **Montioli R**, Cellini B, Borri Voltattorni C. "Molecular insights into the pathogenicity of variants associated with the aromatic amino acid decarboxylase deficiency." *J Inherit Metab Dis*. 2011 May 4. IF 4.04
14. Oppici E, **Montioli R**, Lorenzetto A, Bianconi S, Borri Voltattorni C, Cellini B. "Biochemical analyses are instrumental in identifying the impact of mutations on holo and/or apo-forms and on the region(s) of alanine:glyoxylate aminotransferase variants associated with Primary Hyperoxaluria Type I." *Mol Genet Metab*. 2011 Oct IF 3.37
15. Giardina G., **Montioli R**., Gianni S., Cellini B., Paiardini A., Voltattorni CB and Cutruzzolà F. "The open conformation of human DOPA decarboxylase reveals the mechanism of PLP addition to Group II decarboxylases. *Proceedings of National Academy of sciences (PNAS)* 2011 Dec 20;108(51):20514-9 IF 9.35
16. **Montioli R,** Fargue S, Lewin J, Zamparelli C, Danpure CJ, Voltattorni CB and Cellini B. "The N-terminal extension is essential for the formation of the active dimeric structure of liver peroxisomal alanine:glyoxylate aminotransferase" *Int J Biochem Cell Biol***.** 2012 Mar;44(3):536-46. IF 3.67
17. Cellini B, Oppici E, Paiardini A, **Montioli R**. "Molecular insights into primary hyperoxaluria type 1 pathogenesis" *Front Biosci* (Landmark Ed). 2012 Jan 1;17:621-34. Review. IF 2.75
18. Daidone F, **Montioli R**, Paiardini A, Cellini B, Macchiarulo A, Giardina G, Bossa F, Borri Voltattorni C. "Identification by virtual screening and in vitro testing of human DOPA decarboxylase inhibitors" *PLoS One*. 2012;7(2):e31610. IF 2.87
19. Cellini B, **Montioli R**, Oppici E, Voltattorni CB. "Biochemical and computational approaches to improve the clinical treatment of dopa decarboxylase-related diseases: an overview" *Open Biochem J*. 2012;6:131-8. IF 0.3
20. **Montioli R**, Oppici E, Cellini B, Roncador A, Dindo M, Voltattorni CB. "S250F variant associated with aromatic amino acid decarboxylase deficiency: molecular defects and intracellular rescue by pyridoxine." *Hum Mol Genet.* 2013 Apr 15;22(8):1615-24. IF 4.48
21. **Montioli R**, Cellini B, Dindo M, Oppici E, Voltattorni CB. "Interaction of human Dopa decarboxylase with L-Dopa: spectroscopic and kinetic studies as a function of pH" *Biomed Res Int***.** 2013;2013:161456. IF 2.14
22. Oppici E, Roncador A, **Montioli R**, Bianconi S, Cellini B. "Gly161 mutations associated with Primary Hyperoxaluria Type I induce the cytosolic aggregation and the intracellular degradation of the apo-form of alanine:glyoxylate aminotransferase." *Biochim Biophys Acta-molecular basis of diseases*. 2013 Sep 17. IF 4.35
23. A Roncador , E Oppici, **R Montioli**, F Maset, B Cellini "TAT-Mediated Delivery of Human Alanine:Glyoxylate Aminotransferase in a Cellular Model of Primary Hyperoxaluria Type I" *Int J Pept Res Ther* (2013) 19:175–184 IF 1.08
24. Hanau S, d'Empaire LP, Capone I, Alberighi S, **Montioli R**, Dallocchio F. "Evidence for dimer/tetramer equilibrium in Trypanosoma brucei 6-phosphogluconate dehydrogenase." *Biochim Biophys Acta-Proteins and proteomics*. 2013 Oct 1. doi:pii: S1570-9639(13)00352-X. IF 2.37

25. Cellini B, **Montioli R**, Oppici E, Astegno A, Voltattorni CB. "The chaperone role of the pyridoxal 5'-phosphate and its implications for rare diseases involving B6-dependent enzymes". *Clin Biochem*. 2014 Feb;47(3):158-65 IF 2.40
26. **Montioli R**, Dindo M, Giorgetti A, Piccoli S, Cellini B, Voltattorni CB. "A comprehensive picture of the mutations associated with aromatic amino acid decarboxylase deficiency: from molecular mechanisms to therapy implications". *Hum Mol Genet*. 2014 23(20) 5429–5440 IF 4.48
27. **Montioli R**, Roncador A, Oppici E, Mandrile G, Giachino DF, Cellini B, Voltattorni CB. S81L and G170R mutations causing Primary Hyperoxaluria type I in homozygosis and heterozygosis: an example of positive interallelic complementation *Hum Mol Genet*. 2014, 23(22) 5998–6007 IF 4.48
28. Oppici E, **Montioli R**, Cellini B. Liver peroxisomal alanine:glyoxylate aminotransferase and the effects of mutations associated with Primary Hyperoxaluria Type I: An overview. *Biochim Biophys Acta-Proteins and proteomics*. 2015 Sep;1854(9):1212-9 IF 2.37
29. **Montioli R**, Oppici E, Dindo M, Roncador A, Gotte G, Cellini B, Borri Voltattorni C. Misfolding caused by the pathogenic mutation G47R on the minor allele of alanine:glyoxylate aminotransferase and chaperoning activity of pyridoxine. *Biochim Biophys Acta-Proteins and proteomics*. 2015 Oct;1854(10 Pt A):1280-9. IF 2.37
30. Oppici E, **Montioli R**, Dindo M, Maccari L, Porcari V, Lorenzetto A, Chellini S, Voltattorni CB, Cellini B. The Chaperoning Activity of Amino-oxyacetic Acid on Folding-Defective Variants of Human Alanine:Glyoxylate Aminotransferase Causing Primary Hyperoxaluria Type I. *ACS Chem Biol*. 2015 Oct 16;10(10):2227-36 IF 4.43
31. Oppici E, **Montioli R**, Dindo M, Cellini B. Natural and Unnatural Compounds Rescue Folding Defects of Human Alanine: Glyoxylate Aminotransferase Leading to Primary Hyperoxaluria Type I. *Curr Drug Targets*. 2016;17(13):1482-91. Review. IF 2.29
32. **Montioli R**, Paiardini A, Kurian MA, Dindo M, Rossignoli G, Heales SJ, Pope S, Voltattorni CB, Bertoldi M. The novel R347G pathogenic mutation of aromatic amino acid decarboxylase provides additional molecular insights into enzyme catalysis and deficiency. *Biochim Biophys Acta-Proteins and proteomics*. 2016 Jun;1864(6):676-82. IF 2.37
33. **Montioli R**, Voltattorni CB, Bertoldi M. Parkinson's Disease: Recent Updates in the Identification of Human Dopa Decarboxylase Inhibitors. *Curr Drug Metab*. 2016;17(5):513-8. IF 2.96
34. Mesa-Torres N, Calvo AC, Oppici E, Titelbaum N, **Montioli R**, Miranda-Vizuete A, Cellini B, Salido E, Pey AL. Caenorhabditis elegans AGXT-1 is a mitochondrial and temperature-adapted ortholog of peroxisomal human AGT1: New insights into between-species divergence in glyoxylate metabolism. *Biochim Biophys Acta-Proteins and proteomics*. 2016 Sep;1864(9):1195-205. IF 2.37
35. Dindo M., **Montioli R**., Busato M., Giogetti A., Cellini B.,Voltattorni CB. Effects of interface mutations on the dimerization of alanine glyoxylate aminotransferase and implication in the mistargeting of the pathogenic variants F152I and I244T. *Biochimie* 2016 accepted for publication. IF 3.35
36. Fagagnini A, **Montioli R**, Caloiu A, Ribó M, Laurents DV, Gotte G. Extensive deamidation of RNase A inhibits its oligomerization through 3D domain swapping. *Biochim Biophys Acta-Proteins and proteomics.* 2017 Jan;1865(1):76-87 IF 2.37
37. **Montioli R***, Zamparelli C, Borri Voltattorni C, Cellini B. Oligomeric State and Thermal Stability of Apo- and Holo- Human Ornithine δ-Aminotransferase. *Protein J*. 2017 Jun;36(3):174-185. IF 1.32 ***Corresponding Author**
38. Giardina G, Paiardini A, **Montioli R**, Cellini B, Voltattorni CB, Cutruzzolà F. Radiation damage at the active site of human alanine:glyoxylate aminotransferase reveals that the cofactor position is finely tuned during catalysis. *Sci Rep*. 2017 Sep 15;7(1):11704. IF 4.12
39. Fagagnini A, Pica A, Fasoli S, **Montioli R**, Donadelli M, Cordani M, Butturini E, Acquasaliente L, Picone D, Gotte G. Onconase dimerization through 3D domain swapping: structural investigations and increase in the apoptotic effect in cancer cells. *Biochem J*. 2017 Nov 6;474(22):3767-3781. IF 4.22
40. **Montioli R**, Janson G, Paiardini A, Bertoldi M, Borri Voltattorni C. Heterozygosis in aromatic amino acid decarboxylase deficiency: Evidence for a positive interallelic complementation between R347Q and R358H mutations. *IUBMB Life*. 2018 Mar;70(3):215-223. IF 3.09
41. **Montioli R**, Desbats MA, Grottelli S, Doimo M, Bellezza I, Borri Voltattorni C, Salviati L, Cellini B. Molecular and cellular basis of ornithine δ-aminotransferase deficiency caused by the V332M

mutation associated with gyrate atrophy of the choroid and retina. *Biochim Biophys Acta Mol Basis Dis*. 2018 Nov;1864(11):3629-3638. IF 4.35

42. Rossignoli G, Grottesi A, Bisello G, **Montioli R**, Borri Voltattorni C, Paiardini A, Bertoldi M. Cysteine 180 Is a Redox Sensor Modulating the Activity of Human Pyridoxal 5'-Phosphate Histidine Decarboxylase. *Biochemistry*. 2018 Nov 6;57(44):6336-6348. IF 2.86
43. Himmelreich N*, **Montioli R**, Bertoldi M, Carducci C, Leuzzi V, Gemperle C, Berner T, Hyland K, Thöny B, Hoffmann GF, Voltattorni CB, Blau N. Aromatic amino acid decarboxylase deficiency: Molecular and metabolic basis and therapeutic outlook* *Mol Genet Metab*. 2019 May;127(1):12-22 IF 3.37**.**
44. **Montioli R**, Paiardini A, Giardina G, Zanzoni S, Cutruzzola F, Cellini B, Borri Voltattorni C. R180T variant of δ-ornithine aminotransferase associated with gyrate atrophy: biochemical, computational, X-ray and NMR studies provide insight into its catalytic features. *FEBS J* 2019 Jul;286(14):2787-2798 IF 4.75
45. **Montioli R**, Battini R, Paiardini A, Tolve M, Bertoldi M, Carducci C, Leuzzi V, Borri Voltattorni C. A novel compound heterozygous genotype associated with aromatic amino acid decarboxylase deficiency: Clinical aspects and biochemical studies. *Mol Gen Met* 2019 Jun;127(2):132-137 IF 3.37
46. **Montioli R**, Bisello G, Dindo M, Rossignoli G, Voltattorni CB, Bertoldi M. New variants of AADC deficiency expand the knowledge of enzymatic phenotypes. *Arch Biochem Biophys* 2020 Mar 30;682:108263 IF 3.31
47. Bianchetto-Aguilera F, Tamassia N, Gasperini S, Calzetti F, Finotti G, Gardiman E, **Montioli R**, Bresciani D, Vermi W, Cassatella MA. Deciphering the fate of slan + -monocytes in human tonsils by gene expression profiling. FASEB J 2020 Jul;34(7):9269-9284 IF 4.17
48. **Montioli R**, Bellezza I, Desbats MA, Voltattorni CB, Salviati L, Cellini B. Deficit of human ornithine aminotransferase in gyrate atrophy: Molecular, cellular, and clinical aspects. Biochim Biophys Acta Proteins Proteom 2020 Oct 14;140555 IF 2.37
49. **Montioli R**, Campagnari R, Fasoli S, Fagagnini A, Caloiu A, Smania M, Menegazzi M, Gotte G. RNase A Domain-Swapped Dimers Produced Through Different Methods: Structure-Catalytic Properties and Antitumor Activity. Life (Basel) 2021 Feb 21;11(2):168. doi: 10.3390/life11020168
50. **Montioli R**, Borri Voltattorni C. Aromatic Amino Acid Decarboxylase Deficiency: The Added Value of Biochemistry. Int J Mol Sci. 2021 Mar 19;22(6):3146. doi: 10.3390/ijms22063146.
51. **Montioli R**, Sgaravizzi G, Desbats MA, Grottelli S, Voltattorni CB, Salviati L, Cellini B. Molecular and Cellular Studies Reveal Folding Defects of Human Ornithine Aminotransferase Variants Associated With Gyrate Atrophy of the Choroid and Retina. Front Mol Biosci 2021 Jul 30;8:695205. doi: 10.3389/fmolb.2021.695205.
52. Longo C, **Montioli R**, Bisello G, Palazzi L, Mastrangelo M, Brennenstuhl H, de Laureto PP, Opladen T, Leuzzi V, Bertoldi M. Compound heterozygosis in AADC deficiency: A complex phenotype dissected through comparison among heterodimeric and homodimeric AADC proteins. Mol Genet Metab Sep-Oct 2021;134(1-2):147-155.
53. Fasoli S, Bettin I, **Montioli R**, Fagagnini A, Peterle D, Laurents DV, Gotte G. Dimerization of Human Angiogenin and of Variants Involved in Neurodegenerative Diseases. Int J Mol Sci. 2021 Sep 17;22(18):10068. doi: 10.3390/ijms221810068.

Book chapters

Barbara Cellini, Riccardo Montioli and Carla Borri Voltattorni "CYSTALYSIN: AN EXAMPLE OF THE CATALYTIC VERSATILITY OF PYRIDOXAL 5'-PHOSPHATE DEPENDENT ENZYMES" Vitamin B Research Advances, Nova Science Publishers, Hauppauge, NY, USA.