



Personal data

Name Surname

Andrea Brancaccio

Degree

PhD in Biochemistry

Birthdate

15/06/1966

Place of birth-Nationality

Rome-Italy

Office/Lab Telephone

39-6-30155135

Office/Lab Fax

39-6-30154309

Office/Lab E-mail

andrea.brancaccio@cnr.it

web/E-mail

brancaccio66@yahoo.it

Curriculum Vitae

Education and training

Timeline

1994-1997: Post-doctoral position in Prof. Jurgen Engel's laboratory, Department of Biophysical Chemistry, Biozentrum, University of Basel, Basel (CH)

1993: two months in Prof. John S. Olson's laboratory, Department of Cell Biology, Rice University, Houston (USA)

1990-1993: PhD in Biochemistry with a thesis entitled "Reactivity control in myoglobin and in active site mutants", Dipartimento di Scienze Biochimiche, Università di Roma La Sapienza, Rome (Italy).

1987-1989: Degree in Biology with a thesis entitled Human hemoglobin modification with 3-5 dibromo-salicyl-fumarate (dBrSF): thermodynamics, kinetics and biotechnology. University of Rome La Sapienza, Rome (Italy).

Profile

In 1989, Andrea Brancaccio received a degree in Biology at the University of Rome La Sapienza, in the Department of Biochemistry, under the supervision of Prof. Maurizio Brunori. He studied the cross-linking of hemoglobin with a diaspirinic compound which increases its thermostability. Afterwards, he started his graduate training. At the beginning, he studied the effect of increasing amounts of sucrose on the allosteric behaviour of hemoglobin. His research interests afterwards focused on the structural-functional relationships in recombinant myoglobins. He investigated the role of several mutations in the active site (Brancaccio et al., Journal of Biological Chemistry, 1994). He obtained his PhD degree in Biochemistry in 1994.

He then moved to the Biozentrum of the University of Basel with a post-doctoral position in the laboratory of Prof. Jürgen Engel where his research interest focused on the extracellular matrix and in particular on dystroglycan. During this period, he collected the very first structural data on alpha-dystroglycan using electron microscopy on the native molecule as well as on some recombinant fragments, thus discovering an N-terminal autonomous module whose structure he subsequently contributed to solve by X-ray crystallography (Bozic et al., Journal of Biological Chemistry, 2004). During his post-doc he also investigated structurally and functionally the interaction between agrin and alpha-dystroglycan. In 1998 he moved back to Italy to join CNR at the Institute of Chemistry of Molecular Recognition (now Institute of Chemical Sciences and Technologies), where his main research interest is still the structural-functional relationships of dystroglycan.

Over the last 10 years, he has spent long periods as a visiting scientist at the School of Biochemistry of the University of Bristol (UK), where he was initially involved on a project on Archaeal chaperonins while he kept following his main interest in dystroglycan and other extracellular matrix proteins. During these years, he was senior author of a series of structural papers on the N-terminal domain of alpha-dystroglycan (WT and mutated) which underlined the importance of its highly flexible structure in the intracellular maturation of the dystroglycan precursor. Following on this line of work, the recently characterized and ever-growing group of

primary dystroglycanopathies became central to its research focus. In parallel with this experimental work, he has performed bioinformatic analyses investigating a series of evolutionary aspects of the dystroglycan protein and gene (Adams & Brancaccio, *Biology Open*, 2015) and of the enzymes responsible for its post-translational modification (Bigotti & Brancaccio, *Open Biology*, 2021).

His work on the transmembrane beta-subunit of dystroglycan recently resulted in the characterization of a site-directed mutant of its ectodomain, C667F. This mutation was shown to cause a severe Muscle-Eye-Brain disease phenotype, and using a combined approach based on cellular biochemistry and super-resolution microscopy, Dr. Brancaccio and collaborators demonstrated that the mutated polypeptide is engulfed in the ER/Golgi (Signorino et al., *Human Mutation*, 2018). The murine knockin model of this mutation has been established and is under analysis in collaboration with the University of Bonn. He has been the recipient of a bilateral CNR-CINVESTAV grant (2019-2020) for analyzing the interactors of beta-dystroglycan in the nucleus and has meanwhile started a new project (funded 2024-2026) within the Medical School of Bristol University on the potential involvement of the agrin-dystroglycan axis in cardiomyocytes regeneration (Bigotti et al., *Front Bioeng Biotechnol.*, 2020). Currently, in Bristol he is also investigating the anti-viral properties of the alpha-dystroglycan N-terminus (with 2 grants and 1 patent awarded so far to this project).

Employment and research experience

Timeline

2014-today: Visiting Fellow at the School of Biochemistry of Bristol University, Bristol (UK).

Since Jan. 2020: Research Manager (equivalent to Full Professor) at Istituto di Scienze e Tecnologie Chimiche "Giulio Natta" - SCITEC (CNR), Rome (Italy).

Oct. 2001 to Dec. 2019: Primo Ricercatore (equivalent to Assistant Professor) at Istituto di Scienze e Tecnologie Chimiche "Giulio Natta" - SCITEC (CNR), Rome (Italy).

2014-2025: Habilitated as Full Professor of Biochemistry and Clinical Biochemistry in the 2012 Scientific National Habilitation (ASN).

2001-2018: Contract Professor (teaching Molecular Biology) at the Catholic University of Rome (Italy). At the end of 2010, elected within the Faculty Council as Representative of Contract Professors of the Faculty of Medicine of Catholic University for 2011-2012. Re-elected for 2013-2014.

1997-2001: Staff Researcher at the Molecular Recognition Chemistry Institute, ICRM (CNR), Rome (Italy)

Grants (as Principal Investigator)

2019-2020 (2 years) Bilateral program CNR-CINVESTAV (Italy-Mexico) **"Identifying dystroglycan binding partners in the cellular nucleus: proteomic analysis of β -DG nuclear interactors"**

2016-2018 (2 years) French AFM-Telethon Pr. 20009 **"Establishing new models for primary dystroglycanopathies"**

2006-2009 (3 years) Italian Telethon Pr. GGP06225 **"Analysis at the molecular level of the interaction between α - and β -dystroglycan and assessment of its implication for skeletal muscle physiopathology"**

2003-2005 (2 years) Italian Telethon Pr. GGP030332 **"Analysis at the molecular level of the interaction between α -dystroglycan and β -dystroglycan"**

2003-2004 (2 years) Antarctic National Scientific Commission (Grant 2003/1.01) **"Molecular analysis of dystroglycan in antarctic fish"**

2001-2003 (3 years) **"Role of dystroglycan in tumorigenesis"** (RBNE014BML_007)

Chief Investigator of a Research Unit in the Project "Transgenic models for complex etiology diseases", FIRB 2001

2000-2002 (2 years) Italian Telethon Pr. 1267 **"Analysis at the molecular level of the interaction between α - and β -dystroglycan"**

Grants (as Co-PI or Collaborator)

"Investigating the extracellular matrix protein agrin and its potential of inducing cardiac repair"

Project Grant from the British Heart Foundation, Co-PI - in collaboration with Maria Giulia Bigotti (University of Bristol, UK). **2024-2026 (3 years)**

"Peptides hydrogels incorporating graphene oxide modulate intra-articular hyaluronic acid delivery: advanced treatment for knee osteoarthritis"

Progetti PRIN PNRR 2022 del MUR - in collaboration with Cleofe Palocci (Università La Sapienza) and Francesca Sciandra (CNR). **2023-2025 (2 years)**

_"Dystrovir, a broad-spectrum antiviral"

University of Bristol - Wellcome Trust Institutional Translation Partnership Award (iTPA), Co-Pi - in collaboration with Maria Giulia Bigotti (University of Bristol, UK). [2023 \(1 year\)](#)

_"Broad spectrum antiviral therapeutic"

Commercialisation Development Fund of University of Bristol, Co-PI - in collaboration with Maria Giulia Bigotti (University of Bristol, UK). [2022-2023 \(2 years\)](#)

_"Design, synthesis and biological evaluation of novel chalcone-based fibrates as novel PPAR α agonists"

Bilateral program CNR/NRC (Italy-Egypt) - in collaboration with Francesca Sciandra (CNR) and Iman Y.A. Ghannam (NRC). [2018-2020 \(2 years\)](#)

_"Dissecting the molecular mechanism of a group II chaperonin: protein folding in the thermosome and role of prefoldin"

Wellcome Trust Research Career re-entry Fellowship assigned to Dr. Maria Giulia Bigotti - in collaboration with Maria Giulia Bigotti (University of Bristol, UK). [2013-2017 \(4 years\)](#)

_"Neurodegenerazione e neuroprotezione: ruolo dell'espressione della neuroglobina indotta dagli ormoni estrogeni"

Progetti PRIN 2010-2011 del MIUR - in collaboration with Paolo Ascenzi (Università di Roma 3) and Bruno Giardina (UCSC). [2012-2015 \(3 years\)](#)

_"Generation and characterization of antibodies to α -dystroglycan"

Grant awarded by Limb Girdle Muscular Dystrophy 2I Research Fund (LGMD2I Fund) in collaboration with Glenn Morris (RJAH Orthopaedic Hospital Trust, Oswestry, UK) and Susan Brown (Royal Veterinary College, London, UK). [2012 \(1 year\)](#)

_"Characterization of a mouse model to evaluate the functional role of the α/β - dystroglycan interface"

Post-doctoral fellowship awarded by the French Muscular Dystrophy Association (AFM) to Francesca Sciandra to support her research activities in my laboratory for 12 months. [2010-2011 \(1 year\)](#)

_"A model of dystroglycan's domains by molecular dynamics simulations"

Standard HOPC Grant 2010 (40000 SPTE) - CASPUR (Consorzio Interuniversitario per le Applicazioni di Supercalcolo per Università e Ricerca) in collaboration with Maria Cristina De Rosa (ICRM, CNR). [2010 \(1 year\)](#)

_"Congenital muscular dystrophies: protocols for clinical and molecular diagnosis, and advances in pathogenesis", Progetti Finalizzati del Ministero della Salute - in collaboration with Enrico Bertini (Bambin Gesù Hospital, Roma) and Eugenio Mercuri (Neuropsychiatry, UCSC). [2008-2009 \(2 years\)](#)

_"Role of the dystrophin-associated glycoprotein complex in limb-girdle and congenital muscular dystrophies: from molecular pathophysiology to potential therapy". Project funded in the frame of the bilateral Italy (ISS) - USA (NIH, Office for Rare Diseases) agreement (2007-2009) - in collaboration with Tamara Petrucci (ISS) and Enzo Ricci (Neurology, UCSC). [2007-2008 \(2 years\)](#)

_"Functional basis and structural aspects of the β -dystroglycan extracellular domain, a natively unfolded protein involved in neuromuscular disorders", Cofin Projects 2004, MIUR - in collaboration with Maurizio Paci (Tor Vergata University). [2004-2005 \(2 years\)](#)

Patents

_Patent GB 2315095.6 "Anti-Viral Agents", filing date: 2 October 2023.

Contracts (PI)

_MicroRNA in human blood leukocytes and coronary angioplasty: towards a new generation stent displaying a lower propensity to restenosis. Abbott Laboratories, Abbott Park, Illinois, USA. [2009 \(1 year\)](#)

Publications: [Google Scholar Page](#) [ORCID](#) [SCOPUS](#) [ResearcherID](#) [ResearchGate](#)
[Mendeley](#)

2023

A missense mutation (C667F) in β -dystroglycan results in reduced dystroglycan protein levels leading to myopathy and destabilization of the blood-brain and blood-retinal barrier protein network. Tan, R.L., Sciandra, F., Hübner, W., Bozzi, M., Reimann, J., Schoch, S., **Brancaccio, A.**, Blaess, S. preprint on BioRxiv (2023) <https://doi.org/10.1101/2023.11.10.566559>

The α -dystroglycan N-terminus is a broad-spectrum antiviral agent against SARS-CoV-2 and enveloped viruses.

Bigotti, M.G., Klein, K., Gan, E.S., Anastasina, M., Andersson, S., Vapalahti, O., Katajisto, P., Erdmann, M., Davidson, A.D., Butcher, S.J., Collinson, I., Ooi, E.E., Balistreri, G., **Brancaccio, A.** and Yamauchi, Y. preprint on BioRxiv (2023) <https://doi.org/10.1101/2023.11.06.565781>

Verbascoside elicits its beneficial effects by enhancing mitochondrial spare respiratory capacity and the Nrf2/HO-1 mediated antioxidant system in a murine skeletal muscle cell line.

Sciandra, F., Bottoni, P., De Leo, M., Braca, A., **Brancaccio, A.** and Bozzi, M. (2023) *Int J Mol Sci* 24(20):15276.

Cryo-electron tomography of *C. elegans* mitochondria reveals how the ATP synthase dimer interface shapes crista membranes.

Buzzard, E., McLaren, M., Bragoszewski, P., **Brancaccio, A.**, Daum, B., Kuwabara, P., Collinson, I. and Gold, V.A.M. preprint on BioRxiv (2023) <https://doi.org/10.1101/2023.02.02.526626>

_2022

Determination of agrin and related proteins levels as a function of age in human hearts.

Skeffington, K.L., Jones, F.P., Saadeh Suleiman, M., Caputo, M., **Brancaccio, A.** and Bigotti, M.G. (2022) *Front Cardiovasc Med* 9:813904.

_2021

High degree of conservation of the enzymes synthesizing the laminin-binding glycoepitope of α -dystroglycan.

Bigotti, M.G. and **Brancaccio A.** (2021) *Open Biology* 11:210104.

_2020

The molecular basis and biologic significance of the β -dystroglycan-emerin interaction.

Gómez-Monsiváis, W.L., Monterrubio-Ledezma, F., Huerta-Cantillo, J., Mondragon-Gonzalez, R., Alamillo-Iniesta, A., García-Aguirre, I., Azuara-Medina, P.M., Arguello-García, R., Rivera-Monroy, J.E., Holaska, J.M., Hernández-Méndez, J.M.E., Garrido, E., Magaña, J.J., Winder, S.J., **Brancaccio, A.**, Martínez-Vieyra, I., Navarro-García, F. and Cisneros, B. (2020) *Int J Mol Sci.* 21(17):594.

Loss of dystroglycan drives cellular senescence via defective mitosis-mediated genomic instability.

Jiménez-Gutiérrez, G.E., Mondragon-Gonzalez, R., Soto-Ponce L.A., Gómez-Monsiváis, W.L., García-Aguirre, I., Pacheco-Rivera, R.A., Suárez-Sánchez, R., **Brancaccio, A.**, Magaña, J.J., Perlingeiro, R.C.R. and Cisneros, B. (2020) *Int J Mol Sci.* 21(14):E4961.

Aggrin-mediated cardiac regeneration: some open questions.

Bigotti, M.G., Skeffington, K.L., Jones, F.P., Caputo, M. and **Brancaccio, A.** (2020) *Front Bioeng Biotechnol.* 8:594.

Identification and modeling of a GT-A fold in the α -dystroglycan glycosylating enzyme LARGE1.

Righino B, Bozzi M, Pirolli D, Sciandra F, Bigotti MG, **Brancaccio A**, De Rosa MC. (2020) *J Chem Inf Model.* 60(6):3145-3156.

_2019

Analysis of α -dystroglycan/LG domain binding modes: investigating protein motifs that regulate the affinity of isolated LG domains.

Dempsey, C.E., Bigotti, M.G., Adams, J.C. and **Brancaccio, A.** (2019) *Front Mol Biosci.* 6:18.

A molecular overview of the primary dystroglycanopathies.

Brancaccio, A. (2019) *J Cell Mol Med.* 23, 3058-3062.

_2018

Intra-ring allostery controls the function and assembly of a hetero-oligomeric class II chaperonin.

Shoemark, D.K., Sessions, R.B., **Brancaccio, A.** and Bigotti, M.G. (2018) *FASEB J.* 32, 2223-2234.

The enzymatic processing of α -dystroglycan by MMP-2 is controlled by two anchoring sites distinct from the active site.

Gioia, M., Fasciglione, G.F., Sbardella, D., Sciandra, F., Casella, M.L., Camerini, S., Crescenzi, M., Gori, A., Tarantino, U., Cozza, P., **Brancaccio, A.**, Coletta, M. and Bozzi, M. (2018) *PLoS One* 13(2):e0192651.

A dystroglycan mutation (p.Cys667Phe) associated to Muscle-Eye-Brain disease with multicystic leucodystrophy results in ER-retention of the mutant protein.

Signorino, G., Covaceuszach, S., Bozzi, M., Hübner, W., Mönkemöller, V., Konarev, P.V., Cassetta, A., Brancaccio, A. and Sciandra, F. (2018) Hum Mutat. 39, 266-280.

_2017

Evaluation of the effect of a floxed Neo cassette within the dystroglycan (*Dag1*) gene.

Sciandra, F., Scicchitano, B.M., Signorino, G., Bigotti, M.G., Tavazzi, B., Lombardi, F., Bozzi, M., Sica, G., Giardina, B., Blaess, S. and Brancaccio, A. (2017) BMC Res Notes 10(601).

The effect of the pathological V72I, D109N and T190M missense mutations on the molecular structure of α -dystroglycan.

Covaceuszach, S., Bozzi, M., Bigotti, M.G., Sciandra, F., Konarev, P.V., Brancaccio, A. and Cassetta, A. (2017) PLoS One. 12(10):e0186110.

Structural flexibility of human α -dystroglycan.

Covaceuszach, S., Bozzi, M., Bigotti, M.G., Sciandra, F., Konarev, P.V., Brancaccio, A. and Cassetta, A. (2017) FEBS Open Bio 7, 1064-1077.

α -Dystroglycan hypoglycosylation affects cell migration by influencing β -dystroglycan membrane clustering and filopodia length: a multiscale confocal microscopy analysis.

Palmieri, V., Bozzi, M., Signorino, G., Papi, M., De Spirito, M., Brancaccio, A., Maulucci, G. and Sciandra, F. Biochim Biophys Acta. (2017) Biochim Biophys Acta. 1863, 2182–2191.

An evaluation of the evolution of the gene structure of dystroglycan.

Brancaccio, A. and Adams, J.C. (2017) BMC Res Notes. 10(1):19.

_2016

Internal (His)₆-tagging delivers a fully functional hetero-oligomeric class II chaperonin in high yield.

Paul, D.M., Beuron, F., Sessions, R.B., Brancaccio, A. and Bigotti, M.G. (2016) Sci. Rep. 6, 20696.

Quantification, 2DE analysis and identification of enriched glycosylated proteins from mouse muscles: difficulties and alternatives.

Menegoci Eugênio, P.F., Assunção, N.A., Sciandra, F., Aquino, A., Brancaccio, A. and Carrilho, E. (2016) Electrophoresis 37, 321-334.

_2015

The evolution of the dystroglycan complex, a major mediator of muscle integrity.

Adams, J.C. and Brancaccio, A. (2015) Biol. Open 4, 1163-1179.

Proteasome activity is affected by fluctuations in insulin-degrading enzyme distribution.

Sbardella, D., Tundo, G.R., Sciandra, F., Bozzi, M., Gioia, M., Ciaccio, C., Tarantino, U., Brancaccio, A., Coletta, M. and Marini, S. (2015) PLoS One 10(7):e0132455.

Genetic engineering of dystroglycan in animal models of muscular dystrophy.

Sciandra, F., Bigotti, M.G., Giardina, B., Bozzi, M. and Brancaccio, A. (2015) Biomed Res. Int. 2015, 635792.

The structure of the T190M mutant of murine α -dystroglycan at high resolution: insight into the molecular basis of a primary dystroglycanopathy.

Bozzi, M., Cassetta, A., Covaceuszach, S., Bigotti, M.G., Bannister, S., Hübner, W., Sciandra, F., Lamba, D. and Brancaccio, A. (2015) PLoS One 10(5):e0124277.

Role of gelatinases in pathological and physiological processes involving the dystrophin-glycoprotein complex.

Bozzi, M., Sciandra, F. and Brancaccio, A. (2015) Matrix Biol. 44-46, 130-137.

α -Dystroglycan is a potential target of matrix metalloproteinase MMP-2.

Sbardella, D., Sciandra, F., Gioia, M., Marini, S., Gori, A., Giardina, B., Tarantino, U., Coletta, M., Brancaccio, A. and Bozzi, M. (2015) Matrix Biol. 41, 2-7.

A new monoclonal antibody against human α -dystroglycan reveals reduced core protein in some, but not all, dystroglycanopathy patients.

Humphrey, E.L., Lacey, E., Le, L.T., Feng, L., Sciandra, F., Morris, C.R., Hewitt, J.E., Holt, I., Brancaccio, A., Barresi, R., Sewry, C.A., Brown, S.C. and Morris, G.E. (2015) Neuromuscul. Disord. 25, 32-42.

2014

Insights from molecular dynamics simulations: structural basis for the V567D mutation-induced instability of zebrafish α -dystroglycan and comparison with the murine model.

Pirolli, D., Bozzi, M., Sciandra, F., Giardina, B., Brancaccio, A. and De Rosa, M.C. (2014) PLoS One 9(7):e103866.

2013

Probing the stability of the “naked” mucin-like domain of human α -dystroglycan.

Bozzi, M., Di Stasio, E., Scaglione, G.L., Desiderio, C., Martelli, C., Giardina, B., Sciandra, F. and Brancaccio A. (2013) BMC Biochem. 14, 15.

Increased levels of expression of dystroglycan may protect the heart.

Brancaccio, A. (2013) Neuromuscul. Disord. 23, 867-870

The multiple affinities of α -dystroglycan.

Sciandra, F., Bozzi, M., Bigotti, M.G. and Brancaccio, A. (2013) Curr. Protein Pept. Sci. 14, 626-634.

2012

Enzymatic processing by MMP-2 and MMP-9 of wild-type and mutated mouse β -dystroglycan.

Sbardella, D., Inzitari, R., Iavarone, F., Gioia, M., Marini, S., Sciandra, F., Castagnola, M., Van Den Steen, P., Opdenakker, G., Giardina, B., Brancaccio, A., Coletta, M. and Bozzi, M. (2012) IUBMB Life 64, 988-994.

Insertion of a myc-tag within α -dystroglycan domains improves its biochemical and microscopic detection.

Morlacchi, S., Sciandra, F., Bigotti, M.G., Bozzi, M., Hübner, W., Galtieri, A., Giardina, B. and Brancaccio, A. (2012) BMC Biochem. 13, 14.

Dystroglycan is associated to the disulfide isomerase ERp57.

Sciandra, F., Angelucci, E., Altieri, F., Ricci, D., Hübner, W., Petrucci, T.C., Giardina, B., Brancaccio, A. and Bozzi, M. (2012) Exp. Cell. Res. 318, 2460-2469.

DAG1, no gene for RNA regulation?

Brancaccio, A. (2012) Gene 497, 79-82.

2011

An immunological analysis of dystroglycan subunits: lessons learned from a small cohort of non-congenital dystrophic patients.

Pavoni, E., Sciandra, F., Tasca, G., Tittarelli, R., Bozzi, M., Giardina, B., Ricci, E. and Brancaccio, A. (2011) Open Neurol. J. 5, 68-74.

Insertion of 16 aminoacids in the BAR domain of the oligophrenin 1 protein causes mental retardation and cerebellar hypoplasia in an Italian family.

Pirozzi, F., Di Raimo, F.R., Zanni, G., Bertini, E., Billuart, P., Tartaglione, T., Tabolacci, E., Brancaccio, A., Neri, G. and Chiurazzi, P. (2011) Hum. Mutat. 32, E2294-307.

A gain-of-glycosylation mutation associated with Myoclonus Dystonia Syndrome affects trafficking and processing of ϵ -sarcoglycan in the late secretory pathway.

Waite, A., De Rosa, M.C., Brancaccio, A. and Blake, D. (2011) Hum. Mutat. 32, 1246-1258.

A second Ig-like domain identified in dystroglycan by molecular modelling and dynamics.

De Rosa, M.C., Pirolli, D., Bozzi, M., Sciandra, F., Giardina, B. and Brancaccio, A. (2011) J. Mol. Graph. Model. 29, 1015-1024.

2010

Corneal deposit of ciprofloxacin after laser subepithelial keratomileusis procedure: a case report.

De Benedetti, G. and Brancaccio, A. (2010) J. Ophthalmol. 2010, 296034.

2009

Enzymatic processing of β -dystroglycan recombinant ectodomain by MMP-9: identification of the main cleavage site.

Bozzi, M., Inzitari, R., Sbardella, D., Monaco, S., Pavoni, E., Gioia, M., Marini, S., Morlacchi, S., Sciandra, F., Castagnola, M., Giardina, B., Brancaccio, A. and Coletta, M. (2009) IUBMB Life 61, 1143-1152.

Mutagenesis at the α - β interface impairs the cleavage of the dystroglycan precursor.

Sciandra, F., Bozzi, M., Morlacchi, S., Galtieri, A., Giardina, B. and Brancaccio, A. (2009) FEBS J. 276, 4933-4945.

Identification of a mutation in agrin that causes congenital myasthenia and affects synapse function.

Huzé, C., Bauché, S., Richard, P., Chevessier, F., Goillot, E., Gaudon, K., Ben Ammar, A., Chaboud, A., Bernard, V., Rouche, A., Alexandri, N., Kuntzer, T., Fardeau, M., Fournier, E., Brancaccio, A., Ruegg, M.A., Koenig, J., Eymard, B., Schaeffer, L. and Hantai, D. (2009) Amer. J. Hum. Genet. 85, 155-167.

Functional diversity of dystroglycan.

Bozzi, M., Morlacchi, S., Bigotti, M.G., Sciandra, F. and Brancaccio, A. (2009) Matrix Biol. 28, 179-187.

2008

Cib2 binds integrin α 7Bb1D and is reduced in laminin α 2 chain-deficient muscular dystrophy.

Häger, M., Bigotti, MG, Meszaros, R, Carmignac, V, Holmberg, J, Allamand, V, Akerlund, M, Kalamajski, S, Brancaccio, A, Mayer, U, Durbeej, M. (2008) J. Biol. Chem. 283, 24760-24769.

Expression of multiple AQP4 pools in the plasma membrane and their association with the dystrophin complex.

Nicchia, G.P., Cogotzi, L., Rossi, A., Basco, D., Brancaccio, A., Svelto, M. and Frigeri, A. (2008) J. Neurochem. 105, 2156-2165.

First molecular characterization and immunolocalization of keratopithelin in adult human skeletal muscle.

Sciandra, F., Morlacchi, S., Allamand, V., De Benedetti, G., Macchia, G., Petrucci, T.C., Bozzi, M. and Brancaccio, A. (2008) Matrix Biol. 27, 360-370.

2007

Duplication of the dystroglycan gene in most branches of teleost fish.

Pavoni, E., Cacchiarelli, D., Tittarelli, R., Orsini, M., Galtieri, A., Giardina, B. and Brancaccio, A. (2007) BMC Mol. Biol. 8, 34.

Dystroglycan, a possible mediator for reducing congenital muscular dystrophy?

Sciandra, F., Gawlik, K., Brancaccio, A. and Durbeej, M. (2007) Trends Biotechnol. 25, 262-268.

Genetic analysis of the dystroglycan gene in bronchopulmonary dysplasia affected premature newborns.

Concolino, P., Capoluongo, E., Santonocito, C., Vento, G., Romagnoli, C., Zuppi, C., Ameglio, F., Brancaccio, A. and Sciandra, F. (2007) Clin. Chim. Acta 378, 164-167.

2006

Purification and characterization of an antifungal thaumatin-like protein from *Cassia didymobotrya* cell suspension culture.

Vitali, A., Pacini, L., Brancaccio, A., Botta, B., Pucillo, L., Bordi, E., De Mori, P., Maras, B. and Giardina, B. (2006) Plant Physiol. Biochem. 44, 604-610.

Concerted mutation of Phe residues belonging to the β -dystroglycan ectodomain strongly inhibits the interaction with α -dystroglycan in vitro.

Bozzi, M., Sciandra, F., Ferri, L., Torreri, P., Pavoni, E., Petrucci, T.C., Giardina, B. and Brancaccio, A. (2006) FEBS J. 273, 4929-4943.

Activation of MuSK and binding to dystroglycan is regulated by alternative mRNA splicing of agrin.

Scotton, P., Bleckmann, D., Stebler, M., Sciandra, F., Brancaccio, A., Meier, T., Stetefeld, J. and Ruegg M.A. (2006) J. Biol. Chem. 281, 36835-36845.

Altered expression of α -dystroglycan subunit in human gliomas.

Calogero, A., Pavoni, E., Gramaglia, T., D'Amati, G., Ragona, G., Brancaccio, A. and Petrucci, T.C. (2006) Cancer Biol. Ther. 5, 441-448.

2005

α -Dystroglycan, the usual suspect?

Brancaccio, A. (2005) Neuromuscul. Disord. 15, 825-828.

O-mannosylation of α -dystroglycan is essential for lymphocytic choriomeningitis virus receptor function.

Imperiali, M., Thoma, C., Pavoni, E., Brancaccio, A., Callewaert, N. and Oxenius, A. (2005) J. Virol. 79, 14297-14308.

α -Dystroglycan does not play a major pathogenic role in autosomal recessive hereditary inclusion-body myopathy.

Broccolini, A., Gliubizzi, C., Pavoni, E., Gidaro, T., Morosetti, R., Sciandra, F., Giardina, B., Tonali, P., Ricci, E., Brancaccio, A. and Mirabella, M. (2005) Neuromuscul. Disord. 15, 177-184.

Immunodetection of partially glycosylated isoforms of α -dystroglycan by a new monoclonal antibody against its β -dystroglycan-binding epitope.

Pavoni, E., Sciandra, F., Barca, S., Giardina, B., Petrucci, T.C. and Brancaccio, A. (2005) FEBS Lett. 579, 493-499.

First genetic analysis of lattice corneal dystrophy type I in a family from Bulgaria.

Capoluongo, E., De Benedetti, G., Concolino, P., Sepe, M., Ambu, R., Faa, G., Sciandra, F., Santonocito, C., D'Alberto, A., Caselli, M. and Brancaccio, A. (2005) Eur. J. Ophthalmol. 15, 804-808.

The dystroglycan complex: from biology to cancer.

Sgambato, A. and Brancaccio, A. (2005) J. Cell. Physiol. 205, 163-169.

2004

The structure of the N-terminal region of murine skeletal muscle α -dystroglycan discloses a modular architecture.

Bozic, D., Sciandra, F., Lamba, D. and Brancaccio, A. (2004) J. Biol. Chem. 279, 44812-44816.

The effect of an ionic detergent on the natively unfolded β -dystroglycan ectodomain and on its interaction with α -dystroglycan.

Bozzi, M., Di Stasio, E., Cicero O.D., Giardina, B., Paci, M. and Brancaccio, A. (2004) Protein Sci. 13, 2437-2445.

A fast and accurate procedure to collect and analyze unfolding fluorescence signal: the case of dystroglycan domains.

Di Stasio, E., Bizzarri, P., Misiti, F., Pavoni, E. and Brancaccio, A. (2004) Biophys. Chem. 107, 197-211.

Increased expression of dystroglycan inhibits the growth and tumorigenicity of human mammary epithelial cells.

Sgambato, A., Camerini, A., Faraglia, B., Pavoni, E., Montanari, M., Spada, D., Losasso, C., Brancaccio, A. and Cittadini, A. (2004) Cancer Biol. Ther. 3, 967-975.

2003

Structural characterization by NMR of the natively unfolded extracellular domain of β -dystroglycan.

Bozzi, M., Bianchi, M., Sciandra, F., Paci, M., Giardina, B., Brancaccio, A. and Cicero, D.O. (2003) Biochemistry 42, 13717-13724.

Localization of phospho- β -dystroglycan (pY892) to an intracellular vesicular compartment in cultured cells and skeletal muscle fibers in vivo.

Sotgia, F., Bonuccelli, G., Bedford, M., Brancaccio, A., Mayer, U., Wilson, M.T., Campos-Gonzalez, R., Brooks, J.W., Sudol, M. and Lisanti, M.P. (2003) Biochemistry 42, 7110-7123.

Dystroglycan expression is frequently reduced in human breast and colon cancers and is associated with tumor progression.

Sgambato, A., Migaldi, M., Montanari, M., Camerini, A., Brancaccio, A., Rossi, G., Cangiano, R., Losasso, C., Capelli, G., Trentini, G.P. and Cittadini, A. (2003) Am. J. Pathol. 162, 849-860.

Recombinant expression of *Mus musculus* myoglobin.

Bianchi, M., Clementi, M.E., Maras, B., Schininà, E., Bozzi, M., Giardina, B. and Brancaccio, A. (2003) Protein Expr. Purif. 29, 265-271.

An adhesion molecule involved in muscular dystrophies: structural and functional analysis of dystroglycan domains.

Brancaccio, A. (2003) Ital. J. Biochem. 52, 51-54.

Dystroglycan and muscular dystrophies related to the dystrophin-glycoprotein complex.

Sciandra, F., Bozzi, M., Bianchi, M., Pavoni, E., Giardina, B. and Brancaccio, A. (2003) Ann. Ist. Super. Sanità 39, 173-181.

The origin of dystrophin-glycoprotein complex(DGC)-related muscular dystrophies: the need for protection against an ancestral pathogen?

Brancaccio, A. (2003) Ital. J. Biochem. 52, 68-71.

2002

The effect of anions on azide binding to myoglobin: an unusual functional modulation.

De Rosa, M.C., Bertonati, C., Giardina, B., Di Stasio, E. and Brancaccio, A. (2002) *Biochim. Biophys. Acta – Proteins and Proteomics* 1594, 341-352.

Multiple functions of the adhesion molecule dystroglycan.

Brancaccio, A., Sciandra, F. and Petrucci, T.C. (2002) in *Recent Res. Devel. in Biophys. Biochem.* 2, 271-28.

2001

Identification of the β -dystroglycan binding epitope within the C-terminal region of α -dystroglycan.

Sciandra, F., Schneider, M., Giardina, B., Baumgartner, S., Petrucci, T.C. and Brancaccio, A. (2001) *Eur. J. Biochem.* 268, 4590-4597.

Plasticity of secondary structure in the N-terminal region of β -dystroglycan.

Boffi, A., Bozzi, M., Sciandra, F., Woellner, C., Bigotti, M.G., Ilari, A. and Brancaccio, A. (2001) *Biochim. Biophys. Acta -Proteins and Proteomics* 1546, 114-21.

Dystroglycan distribution in adult mouse brain: a light and electron microscopy study.

Zaccaria, M.L., Di Tommaso, F., Brancaccio, A., Paggi, P. and Petrucci, T.C. (2001) *Neuroscience* 104, 311-324.

A synthetic peptide corresponding to the 550-585 region of α -dystroglycan binds β -dystroglycan as revealed by NMR spectroscopy.

Bozzi, M., Veglia, G., Paci, M., Sciandra, F., Giardina, B. and Brancaccio, A. (2001) *FEBS Lett.* 499, 210-214.

Functional flexibility of dystroglycan, a transmembrane linker between the extracellular matrix and the cytoskeleton.

Petrucci, T.C., Macchia, G., Macioce, P., Brancaccio, A., Paggi, P. and Ceccarini, M. (2001) *Cell Mol. Biol. Lett.* 6, 226.

2000

Characterization of the β -dystroglycan-growth factor receptor 2 (Grb2) interaction.

Russo, K., Di Stasio, E., Macchia, G., Rosa, G., Brancaccio, A. and Petrucci T.C. (2000) *Biochem. Biophys. Res. Commun.* 274, 93-98.

Anomalous dystroglycan in carcinoma cell lines.

Losasso, C., Di Tommaso, F., Sgambato, A., Ardito, R., Cittadini, A., Giardina, B., Petrucci, T.C. and Brancaccio, A. (2000) *FEBS Lett.* 484, 194-198.

1999

Analysis of heparin, α -dystroglycan and sulfatide binding to the G domain of laminin α 1 chain by site-directed mutagenesis.

Andac, Z., Sasaki, T., Mann, K., Brancaccio, A., Deutzmann, R. and Timpl, R. (1999) *J. Mol. Biol.* 287, 253-264.

Structural basis of glycosaminoglycan modification and of heterotypic interactions of perlecan domain V.

Friedrich, M.V.K., Göhring, W., Mörgelin, M., Brancaccio, A., David, G. and Timpl, R. (1999) *J. Mol. Biol.* 294, 259-270.

A new crystal structure, Ca^{2+} dependence and mutational analysis reveal molecular details of E-cadherin homoassociation.

Pertz, O., Bozic, D., Koch, A.W., Fauser, C., Brancaccio, A. and Engel, J. (1999) *EMBO J.* 18, 1738-1747.

Binding of the G domains of laminin α 1 and α 2 chain and perlecan for heparin, sulfatides, α -dystroglycan and several extracellular matrix proteins.

Talts, J.F., Andac, Z., Göhring, W., Brancaccio, A. and Timpl, R. (1999) *EMBO J.* 18, 863-870.

Structural and functional analysis of the N-terminal extracellular region of β -dystroglycan.

Di Stasio, E., Sciandra, F., Maras, B., Di Tommaso, F., Petrucci, T.C., Giardina, B. and Brancaccio, A. (1999) *Biochem. Biophys. Res. Commun.* 266, 274-278.

1998

Agrin is a high affinity ligand of dystroglycan in non-muscle tissues.

Gesemann, M., Brancaccio, A., Schumacher, B. and Ruegg, M.A. (1998) *J. Biol. Chem.* 273, 600-605.

Crystallization and preliminary X-ray study of saporin, a ribosome inactivating protein from *Saponaria officinalis*.

Savino, C., Federici, L., Brancaccio, A., Ippoliti, R., Lendaro, E. and Tsernoglou, D. (1998) Acta Crystallogr. D – Biological Crystallography 54, 636-8.

A single disulfide bridge (Cys¹⁸²-Cys²⁶⁴) is crucial for α -dystroglycan N-terminal domain stability.

Brancaccio, A., Jenö, P. and Engel, J. (1998) Ann. N. Y. Acad. Sci. 857, 228-231.

Sequence analysis suggests the presence of an IG-like domain in the N-terminal region of α -dystroglycan which was crystallized after mutation of a protease susceptible site (Arg¹⁶⁸ → His).

Bozic, D., Engel, J. and Brancaccio, A. (1998) Matrix Biol. 17, 495-500.

Dystroglycan: a receptor for extracellular matrix proteins with a wide tissue distribution: structural and functional aspects.

Brancaccio, A. (1998) Biochimica in Italia 8-9, 9-21.

1997

The N-terminal region of α -dystroglycan is an autonomous globular domain.

Brancaccio, A., Schulthess, T., Gesemann, M. and Engel, J. (1997) Eur. J. Biochem. 246, 166-172.

1996

***Aplysia limacina* myoglobin cDNA cloning: an alternative mechanism of oxygen stabilization as studied by active site mutagenesis.**

Cutruzzolà, F., Travaglini Allocatelli, C., Brancaccio, A. and Brunori, M. (1996) Biochem. J. 314, 83-90.

Alternative splicing of agrin alters its binding to heparin, dystroglycan and the putative agrin receptor.

Gesemann, M., Cavalli, V., Denzer, J.A., Brancaccio, A., Schumacher, B. and Ruegg, M.A. (1996) Neuron 16, 1-20.

1995

Electron microscopic evidences for a mucin-like region in chick muscle α -dystroglycan.

Brancaccio, A., Schulthess, T., Gesemann, M. and Engel, J. (1995) FEBS Lett. 368, 139-142.

Cloning and sequencing of skeletal muscle α -dystroglycan.

Brancaccio, A., Ruegg, M.A. and Engel, J. (1995) Matrix Biol. 14, 681-685.

Formate binding to ferric wild type and mutant myoglobins. Thermodynamic and X-ray crystallographic study.

Leci, E., Brancaccio, A., Cutruzzolà, F., Travaglini Allocatelli, C., Tarricone, C., Bolognesi, M., Desideri, A. and Ascenzi, P. (1995) FEBS Lett. 357, 227-229.

Interactions among residues CD3, E7, E10 and E11 in myoglobins: attempts to simulate the O₂ and CO binding properties of *Aplysia* myoglobin.

Smerdon, S.J., Krzywda, S., Brzozowsky, A.M., Davies, G.J., Wilkinson, A.J., Brancaccio, A., Cutruzzolà, F., Travaglini Allocatelli, C., Brunori, M., Li, T., Brantley, R.E. Jr, Carver, T., Eich, R., Singleton, E. and Olson, J.S. (1995) Biochemistry 34, 8715-8725.

Conformational states of hemoproteins by XANES: the mutant VR myoglobin.

Della Longa, S., Bianconi, A., Brancaccio, A., Brunori, M., Congiu Castellano, A., Cutruzzolà, F., Hazemann, J.L., Missori, M. and Travaglini Allocatelli, C. (1995) Physica B – Condensed Matter 208 & 209, 743-745.

1994

Structural factors governing azide and cyanide binding to mammalian metmyoglobins.

Brancaccio, A., Cutruzzolà, F., Travaglini Allocatelli, C., Brunori, M., Smerdon, S.J., Wilkinson, A.J., Dou, Y., Keenan, D., Ikeda-Saito, M., Brantley, R.E. Jr and Olson, J.S. (1994) J. Biol. Chem. 269, 13843-13853.

Engineering *Ascaris* hemoglobin oxygen affinity in sperm whale myoglobin: role of tyrosine B10.

Travaglini Allocatelli, C., Cutruzzolà, F., Brancaccio, A., Vallone, B. and Brunori, M. (1994) FEBS Lett. 352, 63-66.

1993

Hydration and allosteric transitions in hemoglobin.

Bellèlli, A., Brancaccio, A. and Brunori, M., (1993) J. Biol. Chem. 268, 4742-4744.

Crystal structure of a distal site double mutant of sperm whale myoglobin at 1.6 Å resolution.

Rizzi, M., Bolognesi, M., Coda, A., Cutruzzolà, F., Travaglini Allocatelli, C., Brancaccio, A. and Brunori, M. (1993) FEBS Lett. 320, 13-16.

Structural and functional characterization of sperm whale myoglobin mutants: role of arginine (E10) in ligand stabilization.

Travaglini Allocatelli, C., Cutruzzolà, F., Brancaccio, A., Brunori, M., Qin, J. and La Mar, G. (1993) Biochemistry 32, 6041-6049.

Solution ¹H-NMR determination of the distal pocket structure of cyanomet complexes of genetically engineered sperm whale myoglobin His64(E7) →Val, Thr67(E10) →Arg. The role of distal hydrogen bonding by Arg67(E10) in modulating ligand tilt.

Qin, J., La Mar, G., Cutruzzolà, F., Travaglini Allocatelli, C., Brancaccio, A. and Brunori, M. (1993) Biophys. J. 65, 2178-2190.

1992

The amino acid sequence and oxygen-binding properties of the single hemoglobin of the cold-adapted Antarctic teleost *Gymnodraco acuticeps*.

Tamburrini, M., Brancaccio, A., Ippoliti, R. and Di Prisco, G. (1992) Arch. Biochem. Biophys. 1, 295-302.

Hemoglobin Dallas (α97(G4)Asn → Lys): functional characterization of a high oxygen affinity natural mutant.

Lendaro, E., Ippoliti, R., Brancaccio, A., Bellelli, A., Vallone, B., Ivaldi, G., Sciaratta, G.V., Castello, C. Tomova, S., Brunori, M. and Amiconi, G. (1992) Biochim. Biophys. Acta – Molecular Basis of Disease 1180, 15-20.

Biochemical and rheodynamic properties of red blood cells crosslinked with glutaraldehyde.

Arevalo, F., Bellelli, A., Brancaccio, A., Ippoliti, R., Lendaro, E. and Brunori, M. (1992) Biotechnol. Appl. Biochem. 16, 195-200.

1990

Cooperative ligand binding of crosslinked hemoglobins at very high temperatures.

Bellelli, A., Ippoliti, R., Brancaccio, A., Lendaro, E. and Brunori, M. (1990) J. Mol. Biol. 213, 571-574.