

## PROF. PAOLO BERNARDI

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Place and date of birth	Cividale del Friuli (Italy) November 21 <sup>st</sup> , 1953
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Citizenship	Italian

### STUDIES

High School Diploma	Liceo Classico "Paolo Diacono", Cividale del Friuli (Italy) 1971
M.D. Degree	University of Padova (Italy), cum laude 1978

### APPOINTMENTS

1979 – 1987:	<b>Assistant Professor</b> , University of Padova Medical School
1988 – 1999:	<b>Associate Professor</b> , University of Padova Medical School
2000 – present:	<b>Full Professor</b> , University of Padova Medical School
2001 – 2004:	<b>Deputy Dean</b> of the Medical Faculty, University of Padova
2003 – 2009:	<b>Chairman</b> , Department of Biomedical Sciences, University of Padova
2012 – 2018:	<b>Director</b> , Postgraduate School of Clinical Pathology, University of Padova
2012 – present:	<b>Coordinator</b> , Ph.D. Program in Biomedical Sciences, University of Padova

### HONORS

1984:	<b>EMBO Fellow</b> , University of Helsinki, Finland
1985 – 87:	<b>Fogarty Fellow</b> , Whitehead Institute for Biomedical Research, Cambridge Massachusetts, USA
2006 – 2010 and 2014-2016:	<b>Council Member</b> , Bioenergetics Subgroup, Biophysical Society USA
2006 – 2014:	<b>Socio Corrispondente</b> , Istituto Veneto di Scienze, Lettere ed Arti, Venice
2008:	<b>Visiting Professor</b> , Oregon Clinical and Translational Research Institute, Portland, Oregon, USA
2009 – 2011:	<b>Council Member</b> , Associazione di Biologia Cellulare e del Differenziamento
2010 – 2013 and 2016-present:	<b>President</b> , Italian Group of Biomembranes and Bioenergetics
2010 – present:	<b>Member</b> , Accademia Europaea
2012:	<b>Prize for Physiology and Pathology</b> , Ministro Beni e Attività Culturali, Accademia Nazionale dei Lincei, Rome
From 2015:	<b>Socio Effettivo</b> , Istituto Veneto di Scienze, Lettere ed Arti, Venice
From 2016:	<b>Scientific Advisory Board Member</b> , Institute Cochin, Paris
From 2016:	<b>Scientific Committee Member</b> , Fondazione Fibrosi Cistica, Verona
From 2016:	<b>Membro</b> , Consiglio direttivo della Scuola Galileiana di Studi Superiori, Padova
From 2019:	<b>Socio Corrispondente</b> , Istituto Lombardo - Accademia di Scienze e Lettere, Milano

### SCIENTIFIC INTERESTS AND CAREER HIGHLIGHTS

Paolo Bernardi began his studies on mitochondrial physiology and ion transport under the guidance of Giovanni Felice Azzone, one of the founding Fathers of Bioenergetics. His education in Cellular and Molecular Biology was completed with a long-term stay at the Whitehead Institute for Biomedical Research - Massachusetts Institute of Technology, where he worked under the supervision of Harvey F. Lodish. He pioneered the field of mitochondrial channels and their role in cellular pathophysiology. In particular, he focused on the permeability transition pore (PTP), a high conductance channel that is increasingly recognized as a key player in cell death. During the early 1990s he defined key points of regulation of the PTP in isolated mitochondria (membrane potential, matrix pH, Me<sup>2+</sup>-binding sites, specific redox-sensitive sites). He then developed tools to reliably monitor mitochondrial function *in situ*, and addressed mechanistic questions on the PTP as a target in degenerative diseases and cancer. His studies have been extended to *in vivo* models, and led to the demonstration that early mitochondrial adaptation plays a key role in hepatocarcinogenesis [Klöhn et al. (2003)

*Proc Natl Acad Sci USA* 100, 10014-10019] and in onset of the Warburg effect [Sciacovelli et al. (2013) *Cell Metab* 17, 988-999]; and that mitochondrial dysfunction mediated by the PTP unexpectedly causes muscular dystrophy in collagen VI deficiency [Irwin et al. (2003) *Nat Genet* 35, 367-371; Angelin et al. (2007) *Proc Natl Acad Sci USA* 104, 991-996; Merlini et al. (2008) *Proc Natl Acad Sci USA* 105, 5225-5229]. These studies paved the way to a potential therapy of Ullrich Congenital Muscular Dystrophy and Bethlem Myopathy with NIM811, a non immunosuppressive analog of cyclosporin A [Zulian et al. (2014) *Hum Mol Genet* 23, 5353-5363]. The recent identification of the PTP, which appears to originate from dimers of the FoF<sub>1</sub> ATP synthase [Giorgio et al. (2013) Dimers of mitochondrial ATP synthase form the permeability transition pore, *Proc Natl Acad Sci USA* 110, 5887-5892; Urbani et al. (2019) Purified F-ATP synthase forms a Ca<sup>2+</sup>-dependent high-conductance channel matching the mitochondrial permeability transition pore, *Nat. Commun.* (2019) 10, 4341] and the dissection of its key regulatory sites [Giorgio et al. (2017) Ca<sup>2+</sup> binding to F-ATP synthase  $\beta$  subunit triggers the mitochondrial permeability transition, *EMBO Rep.* 18, 1065-1076; Antoniel et al. (2018) The unique histidine in OSCP subunit of F-ATP synthase mediates inhibition of the permeability transition pore at acidic pH, *EMBO Rep* 19, 257-268] offer great promise for further molecular definition of the pore and of its function in health and disease. Development of novel chemical inhibitors of the PTP with potential use in degenerative diseases developed with NIH funding is currently one of the most successful research programs of the Bernardi lab.

Paolo Bernardi was a major actor in the Mitochondrial Renaissance of the 1990s. As early as 1992 he was one of the few to realise the importance of mitochondria in cell death well before the role of cytochrome *c* release was shown to be a key event in apoptosis. He pioneered the field rapidly reaching international recognition, as testified by 214 invited lectures at meetings and 133 seminars at prestigious Institutions worldwide. He has organized or coorganized key meetings on mitochondrial pathophysiology that significantly contributed to the continuing success of the field (see below). His achievements were possible through the work and training of junior personnel (from 1991 Prof. Bernardi has supervised the work of 27 Graduate Students from the University of Padova, 4 visiting Graduate Students from other Universities and 20 postdoctoral Fellows).

## MEETINGS AS ORGANIZER

1. Conference *New Perspectives in Mitochondrial Research*, Padova, Italy 1993
2. Colloquium *Mitochondria in Cell Death*, 8<sup>th</sup> European Bioenergetics Conference, Valencia, Spain 1994
3. Colloquium *The Mitochondrial Permeability Transition in Accidental and Programmed Cell Death*, 9<sup>th</sup> European Bioenergetics Conference, Louvain-la-Neuve, Belgium 1996
4. First Conference *Frontiers in Mitochondrial Research*, Albany, NY 1996
5. Conference *New Perspectives in Mitochondrial Research*, Padova, Italy 1997
6. Second Conference *Frontiers in Mitochondrial Research*, Albany, NY 1998
7. Third Conference *Frontiers in Mitochondrial Research*, Albany, NY 2000
8. Symposium *New Frontiers in Mitochondrial Research – from Bioenergetics to Dynamics*, Bertinoro, Italy 2007
9. 52<sup>nd</sup> Annual Meeting of the Biophysical Society USA, Bioenergetics Subgroup Meeting *Mitochondria Bioenergetics in Disease and Therapeutics*, Long Beach, CA 2008
10. 54<sup>th</sup> Annual Meeting of the Biophysical Society USA, Bioenergetics Subgroup Meeting *Mitochondria in Disease*, San Francisco, CA 2010
11. 58<sup>th</sup> Annual Meeting of the Biophysical Society USA, Bioenergetics Subgroup Meeting *Ion Channels in the Inner Mitochondrial Membrane*, San Francisco, CA 2014
12. Cold Spring Harbor Asia Conference *Mitochondria*, Suzhou, China 2015
13. 19<sup>th</sup> European Bioenergetics Conference, Riva del Garda, Italy 2016
14. Cold Spring Harbor Asia Conference *Mitochondria*, Suzhou, China 2017
15. Cold Spring Harbor Asia Conference *Mitochondria*, Suzhou, China 2019

## INVITED PRESENTATIONS to Internationally Established Conferences (selected from 214)

- **European Bioenergetics Conferences:** 8<sup>th</sup> (Valencia, Spain 1994), 9<sup>th</sup> (Louvain-la-Neuve, Belgium 1996), 10<sup>th</sup> (Göteborg, Sweden 1998), 12<sup>th</sup> (Arcachon, France 2002), 14<sup>th</sup> (Moscow, Russian Federation 2006), 15<sup>th</sup> (Dublin, Ireland 2008), 16<sup>th</sup> (Warsaw, Poland 2010), 17<sup>th</sup> (Freiburg, Germany 2012), 20<sup>th</sup> **Opening Lecture** (Budapest, Hungary 2018), 21<sup>st</sup> (Aix-en-Provence, France 2020).

- **Gordon Research Conferences:** *Bioenergetics* Andover, New Hampshire 1997; *Macromolecular Organization and Cell Function* Oxford, UK 2000; *Mitochondria and Chloroplasts* Lucca, Italy 2010; Lucca, Italy 2014; *Intracellular Ion Channels* Waltham, Massachusetts 2015; *Bioenergetics* Andover, New Hampshire 2015; *Mitochondria in Health and Disease* (Ventura Beach, California 2019)
- **International Society for Heart Research Conferences:** Rhodes, Greece 1998; Brisbane, Australia 2004; Manchester, UK 2006; San Diego, California 2013; Buenos Aires, Argentina 2016; New Orleans, Louisiana 2017; Osaka, Japan 2017; Beijing, China 2019
- **Keystone Symposia:** *Mitochondrial Dysfunction in Pathogenesis* Santa Fe, New Mexico 2000; *Mitochondria and Pathogenesis, Keynote lecture* Copper Mountain, Colorado 2002; *Metabolomics: From Bioenergetics to Apoptosis* Snowbird, Utah 2006; *Mitochondrial Dynamics and Function* Banff, Alberta 2012; *Mitochondria, Metabolism, and Heart Failure/Diabetes* Santa Fe, New Mexico 2015
- **Biophysical Society USA, Bioenergetics Subgroup Meeting:** *Mitochondrial Ion Channels: Their Physiological and Pathological Relevance* Baltimore, Maryland 2004; *Mitochondria Bioenergetics in Disease and Therapeutics* Long Beach, California 2008; *Ion Channels in the Inner Mitochondrial Membrane* San Francisco, California 2014; *Symposium on Mitochondrial Calcium Fluxes* (San Diego, California 2020)
- **Cold Spring Harbor,** Meeting *The Evolving Concept of Mitochondria: From Physics to Biology to Medicine* (Cold Spring Harbor, New York 2018)
- **Cold Spring Harbor Asia** Conferences on *Mitochondria* Suzhou, China 2015, 2017, 2019
- **Philippe Laudat Conference** *The Mitochondrion: Critical for Health, Death and Disease* Aix-les-Bains, France 1998
- **Fernström Symposium** *Role of Mitochondria in Apoptosis and Neurodegeneration* Lund, Sweden 2000
- **Katzir Conference** *Cellular Implications of Redox Signaling* – Weizmann Institute of Science in Abano Terme, Italy 2001
- **Meetings of FEBS (Federation of the European Biochemical Societies):** 27<sup>th</sup>, Lisbon, Portugal 2001; 29<sup>th</sup>, Warsaw, Poland 2004; 35<sup>th</sup>, Göteborg, Sweden 2010, Krakow, 2019)
- **European Society of Cardiology (ESC):** Barcelona, Spain 2009; Roma, Italy 2016; Barcelona, Spain 2017
- **Euromit V,** The Fifth European Meeting on Mitochondrial Pathology Venezia, Italy 2001
- 41<sup>st</sup> Annual Meeting of the **Society of Toxicology USA** Nashville, Tennessee 2002
- 35<sup>th</sup> Annual Meeting of the **American Society of Nephrology** Philadelphia, Pennsylvania 2002
- **Seventh Wiggers-Bernard Conference** *Mitochondrial Dysfunction in Shock, Sepsis and Organ Failure* Vienna, Austria 2003
- **Cold Spring Harbor Meeting** *Mitochondria in Neurological Disease and Aging* Cold Spring Harbor, New York 2005
- **Novartis Foundation** Symposium 287 *New Perspectives on Mitochondrial Biology* London, UK 2006
- **MiP – 64<sup>th</sup> Harden Conference** *Mitochondrial Physiology* Ambleside, UK 2007
- **New York Academy of Sciences** *Mitochondria and Oxidative Stress in Neurodegenerative Disorders* New York, 2007
- **World Muscle Society** Geneva, Switzerland 2009
- **American Society for Investigative Pathology** *Mitochondrial Medicine* Anaheim, California 2010
- 65<sup>th</sup> Annual Meeting of the **Society of General Physiologists** *Mitochondrial Physiology and Medicine* Woods Hole, Massachusetts 2011
- 9<sup>th</sup> Conference of the **Asian Society of Mitochondrial Research and Medicine** Beijing, China 2012
- **Cell Symposium** *Mitochondria: from Signalling to Disease* Lisbon, Portugal 2013
- 4<sup>th</sup> **European Calcium Society Workshop** *Ca<sup>2+</sup> and Cell Death* Leuven, Belgium 2013
- **British Society for Cardiovascular Research** Meeting *Cardiac Metabolic Disorders and Mitochondrial Dysfunction* Oxford, UK 2017
- **Weizmann Meeting on Mitochondria** (Rehovot, Israel 2018)
- **FASEB Conference** *Mitochondrial Biogenesis and Dynamics in Health and Disease, Keynote Lecture* (Palm Springs, California 2019)

## EDITORIAL ACTIVITIES

1. **Chief Specialty Editor,** *Frontiers in Mitochondrial Research* (from 2012).
2. **Editorial Board Member (current):** *Biochimica et Biophysica Acta* (from 1999), *Pharmacological Research* (from 2015), *Current Opinion in Physiology* (from 2017).
3. **Editorial Board Member (past):** *Journal of Biological Chemistry* (1997-2002, 2003-2008, 2010-2015), *Archives of Biochemistry and Biophysics* (1998-2003), *IUBMB Life* (2002-2007).
4. **Guest Editor:** *Biochimica et Biophysica Acta* “Mitochondria in Cell Death” (1998); *BioFactors* “New Perspectives in Mitochondrial Research” (1998); *IUBMB Life* Special Issue “Perspectives in Mitochondrial Research” (2001); *Biochimica et Biophysica Acta* EBEC Special Issue (2016).
5. **Section Editor,** *Pathobiology of Human Disease*, Elsevier (2014).

6. Member of the **Education Committee**, *International Union of Biochemistry and Molecular Biology* (1998-2003).

## FUNDING

Ministry for the University and Scientific Research – Italy; University of Padova; Telethon – Italy; National Institutes of Health - Public Health Service (USA); AIRC (Italian Association on Cancer Research); Fondazione Cassa di Risparmio di Padova e Rovigo; Leducq Foundation.

## BIBLIOMETRIC INDICATIONS

As of October, 2019 Prof. Bernardi has published 254 peer-reviewed articles that have received 32,012 citations with an H index of 91 (Google Scholar) or 79 (Scopus). His most quoted paper [Bernardi P (1999) Mitochondrial transport of cations: Channels, exchangers and permeability transition. *Physiol Rev* 79, 1127-1155] has 1,623 citations; his most quoted experimental paper [Basso, E., Fante, L., Fowlkes, J., Petronilli, V. Forte, M.A. and Bernardi, P. (2005) Properties of the Permeability Transition Pore in Mitochondria Devoid of Cyclophilin D, *J Biol Chem* 280, 18558-18561] ranks 4<sup>th</sup> with 798 quotes. His “top ten” list gathers over 8,200 quotes.

## PUBLICATIONS, LAST 5 YEARS (2014-2019)

1. **Bernardi, P.** and Rasola, A. (2014) Inner Membrane Permeabilization – The Permeability Transition, *Pathobiology of Human Disease* (McManus, Linda M. and Mitchell, Richard N., Eds.) Academic Press, San Diego pp. 162-169
2. Ciscato, F., Sciacovelli, M., Villano, G., Turato, C., **Bernardi, P.**, Rasola, A. and Pontisso, P. (2014) SERPINB3 protects from oxidative damage by chemotherapeutics through inhibition of mitochondrial respiratory Complex I, *Oncotarget* 5, 2418-2427
3. Da-Rè, C., Franzolin, E., Biscontin, A., Piazzesi, A., Pacchioni, B., Gagliani, C., Mazzotta, G., Tacchetti, C., Zordan, M.A., Zeviani, M., **Bernardi, P.**, Bianchi, V., De Pittà, C. and Costa, R. (2014) Functional characterization of drim2, the *Drosophila melanogaster* homolog of the yeast mitochondrial deoxynucleotide transporter, *J. Biol. Chem.* 289, 7448-7459
4. Nowikovsky, K. and **Bernardi, P.** (2014) LETM1 in mitochondrial cation transport, *Front. Physiol.*, 5:83
5. Da-Rè, C., De Pittà, C., Zordan, M.A., Zeviani, M., Teza, G., Nestola, F., Costa, R., and **Bernardi, P.** (2014) UCP4C mediates uncoupled respiration in larvae of *Drosophila melanogaster*, *EMBO Rep.* 15, 586–591
6. Villano, G., Turato, C., Quarta, S., Ruvoletto, M., Ciscato, F., Terrin, L., Semeraro, R., Paternostro, C., Parola, M., Alvaro, D., **Bernardi, P.**, Gatta, A. and Pontisso, P. (2014) Hepatic progenitor cells express SerpinB3. *BMC Cell Biol.* 15, 5
7. Šileikytė, J., Blachly-Dyson, E., Sewell, R., Ricchelli, F., **Bernardi, P.\*** and Forte, M.\* (2014) Regulation of the Mitochondrial Permeability Transition Pore by the Outer Membrane does not Involve the Peripheral Benzodiazepine Receptor (TSPO), \*Corresponding Authors, *J. Biol. Chem.* 289, 13769-13781
8. Carraro, M, Giorgio, V., Šileikytė, J., Sartori, G., Forte, M., Lippe, G., Zoratti, M, Szabò, I. and **Bernardi, P.** (2014) Channel Formation by Yeast F-ATP Synthase and the Role of Dimerization in the Mitochondrial Permeability Transition, *J. Biol. Chem.* 289, 15980-15985 – **paper of the year in Bioenergetics**
9. Zulian, A., Rizzo, E., Schiavone, M., Palma, E., Tagliavini, F., Blaauw, B., Merlini, L., Maraldi, N.M., Sabatelli, P., Braghetta, P., Bonaldo, P., Argenton, F. and **Bernardi, P.** (2014) NIM811, a cyclophilin inhibitor without immunosuppressive activity, is beneficial in collagen VI congenital muscular dystrophy models, *Hum. Mol. Genet.* 23, 5353-5363
10. Antoniel, M., Giorgio, V., Fogolari, F., Glick, G.D., **Bernardi, P.** and Lippe, G. (2014) The oligomycin-sensitivity conferring protein of mitochondrial ATP synthase: Emerging new roles in mitochondrial pathophysiology, *Int. J. Mol. Sci.* 15, 7513-7536
11. Fancelli, D., Abate, A., Amici, R., **Bernardi, P.**, Ballarini, M., Cappa, A., Carezzi, G., Colombo, A., Contursi, C., Di Lisa, F., Dondio, G., Gagliardi, S., Milanese, E., Minucci, S., Pain, G., Pelicci, P.G., Saccani, A., Storto, M., Thaler, F., Varasi, M., Villa, M., Plyte, S. (2014) Cinnamic Anilides as New Mitochondrial Permeability Transition Pore Inhibitors Endowed with Ischemia-Reperfusion Injury Protective Effect in Vivo, *J. Med. Chem.* 57, 5333-5347
12. Sorato, E., Menazza, S., Zulian, A., Sabatelli, P., Gualandi, F., Merlini, L., Bonaldo, P., Canton, M., **Bernardi, P.** and Di Lisa, F. (2014) Monoamine oxidase inhibition prevents mitochondrial dysfunction and apoptosis in myoblasts from patients with collagen VI myopathies, *Free Radic. Biol. Med.* 75C, 40-47
13. Gibellini, L., Pinti, M., Boraldi, F., Giorgio, V., **Bernardi, P.**, Bartolomeo, R., Nasi, M., De Biasi, S., Missiroli, S., Carnevale, G., Losi, L., Tesei, A., Pinton, P., Quaglino, D., and Cossarizza, A. (2014) Silencing of mitochondrial Lon protease deeply impairs mitochondrial proteome and function in colon cancer cells, *FASEB J.* 28, 5122-5135

14. Da-Rè, C., von Stockum, S., Biscontin, A., Millino, C., Cisotto, P., Zordan, M.A., Zeviani, M., **Bernardi, P.**, De Pittà, C., and Costa, R. (2014) Leigh Syndrome in *Drosophila melanogaster*: Morphological and Biochemical Characterization of *Surf1* Post-transcriptional Silencing, *J. Biol. Chem.* **289**, 29235-29246
15. Guzzo, G., Sciacovelli, M., **Bernardi, P.** and Rasola, A. (2014) Inhibition of succinate dehydrogenase by the mitochondrial chaperone TRAP1 has anti-oxidant and anti-apoptotic effects on tumor cells, *Oncotarget* **5**, 11897-11908
16. Zulian A., Tagliavini, F., Rizzo, E. Pellegrini, C., Sardone, F., Zini, N., Maraldi, N.M., Faldini, C., Merlini, L., **Bernardi, P.\*** and Sabatelli, P.\* (2014) Melanocytes from patients affected by Ullrich congenital muscular dystrophy and Bethlem myopathy have dysfunctional mitochondria that can be rescued with cyclophilin inhibitors, \*Corresponding Authors, *Front. Aging Neurosci.* **6**, 324
17. Rasola, A. and **Bernardi, P.** (2014) The mitochondrial permeability transition pore and its adaptive responses in tumor cells, *Cell Calcium* **56**, 437-445
18. **Bernardi, P.** and Di Lisa, F. (2015) The mitochondrial permeability transition pore: Molecular nature and role as a target in cardioprotection, *J. Mol. Cell. Cardiol.* **78**, 100-106
19. von Stockum, S., Giorgio, V., Trevisan, E., Lippe, G., Glick, G.D., Forte, M., Da-Rè, C., Checchetto, V., Mazzotta, G., Costa, R., Szabò, I., and **Bernardi, P.** (2015) F-ATPase of *D. melanogaster* Forms 53-Picosiemen (53-pS) Channels Responsible for Mitochondrial Ca<sup>2+</sup>-induced Ca<sup>2+</sup> Release, *J. Biol. Chem.* **290**, 4537-4544
20. Di Lisa, F. and **Bernardi, P.** (2015) Modulation of Mitochondrial Permeability Transition Prevents Energetic Failure in Ischemia-Reperfusion Injury of The Heart. Advantages and Limitations, *Curr. Med. Chem.* **22**, 2480-2487
21. **Bernardi, P.**, Di Lisa, F., Fogolari, F. and Lippe, G. (2015) From ATP to PTP and back. A dual function for the mitochondrial ATP synthase, *Circ. Res.* **116**, 1850-1862
22. **Bernardi, P.**, Rasola, A., Forte, M. and Lippe, G. (2015) The Mitochondrial Permeability Transition Pore: Channel Formation by F-ATP Synthase, Integration in Signal Transduction and Role in Pathophysiology, *Physiol. Rev.* **95**, 1111-1155
23. Roy, S., Šileikytė, J., Schiavone, M., Neuenswander, B., Argenton, F., Aubé, J., Hedrick, M.P., Chung, T.D.Y., Forte, M.A.\*, **Bernardi, P.\*** and Schoenen F.J.\* (2015) Discovery, Synthesis, and Optimization of Diarylisoxazole-3-carboxamides as Potent Inhibitors of the Mitochondrial Permeability Transition Pore, \*corresponding Authors, *ChemMedChem* **10**, 1655-1671
24. Šileikytė, J., Roy, S., Porubsky, P., Neuenswander, B., Wang, J., Hedrick, M., Pinkerton, A.B., Salaniwal, S., Kung, P., Mangravita-Novo, A., Smith, L.H., Bourdette, D.N., Jackson, M.R., Aubé, J., Chung, T.D.Y., Schoenen, F.J., Forte, M.A. and **Bernardi, P.** (2015) Small Molecules Targeting the Mitochondrial Permeability Transition. Updated 2015 Jan 16, In: *Probe Reports from the NIH Molecular Libraries Program [Internet]*. Bethesda (MD): National Center for Biotechnology Information (US); 2010-
25. **Bernardi, P.** and Forte, M. (2015) Commentary: SPG7 is an essential and conserved component of the mitochondrial permeability transition pore. *Front. Physiol.* **6**, 320
26. Granatiero, V., Giorgio, V., Cali, T., Patron, M., Brini, M., **Bernardi, P.**, Tiranti, V., Zeviani, M., Pallafacchina, G., De Stefani, D. and Rizzuto, R. (2016) Reduced mitochondrial Ca<sup>2+</sup> transients stimulate autophagy in human fibroblasts carrying the 13514A>G mutation of the ND5 subunit of NADH dehydrogenase, *Cell Death Diff.* **23**, 231-241
27. Roy, S., Šileikytė, J., Neuenswander, B., Hedrick, M.P., Chung, T.D.Y., Aubé, J., Schoenen, F.J., Forte, M.A. and **Bernardi, P.** (2016) N-Phenylbenzamides as Potent Inhibitors of the Mitochondrial Permeability Transition Pore, *ChemMedChem* **11**, 283-288
28. **Bernardi, P.** and Di Lisa, F. (2016) Correspondence on article "Cyclosporine before PCI in Patients with Acute Myocardial Infarction" *N. Engl. J. Med.* **374**, 88-90
29. Fontaine, E. and **Bernardi, P.** (2016) Lethal and non-lethal functions of the permeability transition pore. *Mitochondria and Cell Death* (D.M. Hockenbery ed.) Springer, New York 2016, pp. 1-15
30. Carraro M. and **Bernardi, P.** (2016) Calcium and reactive oxygen species in regulation of the mitochondrial permeability transition and of programmed cell death in yeast, *Cell Calcium* **60**, 102-107
31. Scotton, C., Bovolenta, M., Schwartz, E., Falzarano, M.S., Martoni, E., Passarelli, C., Armaroli, A., Osman, H., Rodolico, C., Messina, S., Pegoraro, E., D'Amico, A., Bertini, E., Gualandi, F., Neri, M., Selvatici, R., Boffi, P., Maioli, M.A., Lochmuller, H., Straub, V., Bushby, K., Castrignanò, T., Pesole, G., Sabatelli, P., Merlini, L., Braghetta, P., Bonaldo, P., **Bernardi, P.**, Reghan Foley, A., Cirak, S., Zaharieva, I., Muntoni, F., Capitanio, D., Gelfi, C., Kotelnikova, E., Yuryev, A., Lebowitz, M., Zhang, X., Hodge, B.A., Esser K. and Ferlini, A. (2016) Deep RNA profiling identified Clock and molecular clock genes as pathophysiological signatures in collagen VI myopathy *J. Cell Sci.* **129**, 1671-1684
32. **Bernardi, P.** (2016) 19<sup>th</sup> European Bioenergetics Conference–Preface, *Biochim. Biophys. Acta* **1857**, 1023-1026
33. Zulian, A., Schiavone, M., Giorgio, V. and **Bernardi, P.** (2016) Forty years later: mitochondria as therapeutic targets in muscle diseases, *Pharmacol. Res.* **113**, 563-573

34. **Bernardi, P.** and Forte, M (2016) Commentary: The *m*-AAA protease associated with neurodegeneration limits MCU activity in mitochondria, *Front. Physiol.* **7**, 583
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