

CV  
PAOLO SCUDIERI

Personal Data:

Date and place of birth: 06/04/1986, Genoa, Italy

Education:

2018: PhD, Pediatric Sciences - Genetics, at University of Genoa, ITALY

2016: Residency in Clinical Pathology (summa cum laude) at University of Pisa, ITALY.

2010: Master's degree in Biological Sciences (summa cum laude). University of Genoa, ITALY.

2008: Bachelor's degree in Biological Sciences (summa cum laude). University of Genoa, ITALY.

Positions:

2024-today: assistant professor (RtdB) - MEDS-01/A (Ex MED/03) - Medical Genetics, DiNOGMI, University of Genoa; Medical Genetics Unit, IRCCS Giannina Gaslini, Genoa, Italy.

2020-2024: non-tenured assistant professor (RTDa) Sector MED/03 - Medical Genetics, DiNOGMI, University of Genoa; Medical Genetics Unit, IRCCS Giannina Gaslini, Genoa, Italy.

2016-2020: researcher at the Telethon Institute of Genetics and Medicine, Pozzuoli (Italy).

2010-2016: post-graduate fellowship at IRCCS Giannina Gaslini, Genoa (Italy).

Professional Appointments:

2021-2023: European Cystic Fibrosis Society member (Membership number: 4812)

2020-today: Review Editor on the Editorial Board of Pediatric Neurology (specialty section of Frontiers in Neurology and Frontiers in Pediatrics)

2018-2021: *Ad hoc* reviewer for International Journal of Molecular Sciences, Journal of Applied Genetics, Life, Frontiers Pharmacology.

Teaching:

2021-today: Teaching position as non-tenured assistant professor. Topic: Medical Genetics, Degree Course in Nursing, 12 hours. Faculty of Medicine. University of Genoa, Italy.

2021-today: Teaching position as non-tenured assistant professor. Topic: Human Genetics and Laboratory, Degree Course in Medical-pharmaceutical biotechnology, 16 hours. Faculty of Medicine. University of Genoa, Italy.

2021-2024: Supervisor of 3 PhD students in Pediatric Sciences – Genetics, University of Genoa, Italy.

2023-2024: Supervisor of 3 undergraduate students in Medical-pharmaceutical biotechnology, University of Genoa, Italy.

2022-2023: Supervisor of 1 undergraduate student in Medical-pharmaceutical biotechnology, University of Genoa, Italy.

Awards and recognition

- 2023: Habilitation for Associate Professorship 06/A1 (MED/03 – Medical Genetics)
- 2020: Gerd Döring Award 2020, an initiative of the European Cystic Fibrosis Society given annually to honor an exceptional early career young European scientist.
- 2019: Habilitation for Associate Professorship 05/D1 (BIO/09 – Physiology)
- 2019/2018/2017/2015/2013: Italian Cystic Fibrosis Research Foundation Free Registration Award, European Cystic Fibrosis Society Conference

Management or participation in the activities of a research group characterized by collaborations at national or international level:

- PRIN PNRR 2022 (Edoardo Moretto, PI); "Investigating A $\beta$ -tau mediated axonal dysfunction in Alzheimer's disease". Role in the project: Partner (i.e. responsible of the 2nd unit). Duration: 2 years (2023-2025). Budget: 105.000 euro.
- Cariplo Telethon Alliance GJC2022 (Lorenzo Cingolani, PI); "Targeting Hippocalcin-like protein 4 (HPCAL4) provides new therapeutic opportunities for episodic ataxia type 2 and epileptic encephalopathy 42". Role in the project: Partner (i.e. responsible of the 2nd unit). Duration: 2 years (2023-2025). Budget: 70.000 euro.
- Ministero della Salute – Ricerca Finalizzata 2021 (Paolo Scudieri, PI); "Glut-1 deficiency: new therapeutic strategies to increase glucose transport across the blood brain barrier". Role in the project: PI. Duration 3 years (2023-2026). Budget: 210.000 euro.
- LICE Foundation grant 2022 (Emilia Ricci, PI); "Analisi del profilo di espressione genica, della caratterizzazione morfologica e del controllo del differenziamento degli interneuroni GABAergici nelle mutazioni del gene ZEB2. Un possibile valido modello per lo studio delle epilessie con coinvolgimento primario dei network cortico-sottocorticali". Role in the project: Partner (i.e. responsible of the 2<sup>nd</sup> unit). Duration 2 years (2022-2024). Budget: 46.500 euro.
- Vertex Cystic Fibrosis Research Innovation Award (Paolo Scudieri, PI); "Alternative targets for the treatment of cystic fibrosis basic defect". Role in the project: PI. Duration 3 years (2021-2023). Budget: 750.000 USD.
- Fondazione per la Ricerca sulla fibrosi cistica – FFC#11/2021 (Paolo Scudieri, PI); "Studio dei bersagli alternativi per la correzione del difetto di base della fibrosi cistica". Role in the project: PI. Duration: 2 years (2021-2023). Budget: 104.000 euro.
- Cystic Fibrosis Foundation (Pedemonte Nicoletta, PI); "Meeting unmet needs: theratyping orphan cystic fibrosis mutations". Role in the project: Partner (i.e. responsible of the 2<sup>nd</sup> unit). Duration: 2 years (2020-2021). Budget: 264.600 USD.
- Ministero della Salute – Ricerca Finalizzata 2018 (Sondo Elvira, PI); "Therapeutic approaches for cystic fibrosis patients with rare mutations". Role in the project: Partner Partner (i.e. responsible of the 2<sup>nd</sup> unit). Duration: 3 years (2020-2022). Budget: 420.000 euro.
- Fondazione per la Ricerca sulla fibrosi cistica – FFC#4/2018 (Barraja Paola, PI); "Towards the discovery of new correctors based on nitrogen heterocyclic systems". Role in the project: Partner (i.e. responsible of the 2<sup>nd</sup> unit). Duration: 2 years (2018-2020). Budget: 82.000 euro.
- Vertex innovation awards (Luis Galietta, PI); "ATP12A as a novel therapeutic target in cystic fibrosis lung disease". Role in the project: collaborator. 2018-2020.
- Mukoviszidose (Ulrich Martin, PI); "Evaluation of TMEM16A as a modifier of cystic fibrosis lung phenotype utilizing CF patient-specific human induced pluripotent stem cells". Role in the project: collaborator. 2018-2020
- Cystic Fibrosis Foundation (Luis Galietta, PI); "TMEM16A and ATP12A as therapeutic targets in cystic fibrosis lung disease". Role in the project: collaborator. 2017-2019.
- Fondazione Telethon (Luis Galietta, PI); "Pharmacological modulation of ion transport to treat the basic defect in cystic fibrosis and other genetic diseases". Role in the project: collaborator. 2016-2020.
- Fondazione per la Ricerca sulla fibrosi cistica (Luis Galietta, PI); "Task Force for Cystic Fibrosis". Role in the project: collaborator. 2014-2017.
- Fondazione Telethon (Luis Galietta, PI); "Role of ANO10 in Spinocerebellar Ataxia". Role in the project: collaborator. 2014-2015.

- Fondazione per la Ricerca sulla fibrosi cistica (Luis Galiotta, PI); “Development of small molecules to correct the defective chloride transport in cystic fibrosis”. *Role in the project: collaborator*. 2012-2014.
- Fondazione Telethon (Luis Galiotta, PI); “Identification of novel strategies to correct the chloride transport defect in cystic fibrosis”. *Role in the project: collaborator*. 2010-2013.

Speaker at scientific congresses in Italy or abroad

Dr. Paolo Scudieri participated in 18 national and international congresses: in 6 he was selected for an oral presentation, in 2 of these he was invited as a speaker and in 10 participated as a presenter of scientific posters.

- 2021: *Invited Speaker* for the 44<sup>th</sup> European Cystic Fibrosis Conference. Title: ATP12A as a novel therapeutic target in cystic fibrosis lung disease. ECFS digital 9-12 June 2021.
- 2018: *Oral presentation*. Scudieri P, Musante I, Caci E, Galiotta LJ. Title: ATP12A as an alternative therapeutic target in cystic fibrosis. 15th European Cystic Fibrosis Society Basic Science Conference. Loutraki, Greece.
- 2017: *Oral presentation*. Scudieri P, Musante I, Gianotti A, Galiotta LJ. Title: Role and structure-function relationship of the TMEM16A chloride channel. 14th European Cystic Fibrosis Society Basic Science Conference. Albufeira, Portugal.
- 2016: *Invited speaker* for Physiology 2016 – Joint Meeting of the American Physiological Society and The Physiological Society. Dublin, 2016. Title: Ion channel and lipid scramblase activity associated with expression of TMEM16F/ANO6 isoforms.
- 2013: *Oral presentation*. Scudieri P, Caci E, Gianotti A, Morelli P, Galiotta LJ. Title: TMEM16A/ANO1 expression in human bronchial epitheli.”10th European Cystic Fibrosis Society Basic Science Conference. Malaga, Spain.
- 2011: *Oral presentation*. Scudieri P, Ferrera L, Sondo E, Caci E, Galiotta LJ. Title: Role of TMEM16A protein as a calcium-activated chloride channel. 41th Course “Channels and transporters” of International School of Biophysics. Erice, Italy.
- 2011: *Oral presentation*. Ferrera L, Scudieri P, Sondo E, Pedemonte N, Caci E, Ubbi I, Pagani F, Galiotta LJ. Title: Native calcium activated chloride channels and their association with TMEM16A protein expression. 8th European Cystic Fibrosis Society Basic Science Conference. Tirrenia-Pisa, Italy.

Scientific contributions:

From 2011 to 2025 Paolo Scudieri is the author of 61 scientific contributions, of which 60 in international peer-reviewed scientific journals, and 1 in international books.

Total number of publications: 61

Citations= 1868; H-index = 22 (Source: Scopus)

Publications

Book chapter:

APS/Springer book – Ion Channels and Transporters of Epithelia in Health and Disease. Kirk L. Hamilton and Daniel C. Devor, Editors. Chapter # 17, Paolo Scudieri, Luis J.V. Galiotta - TMEM16 proteins (Anoctamins) in epithelia.

Journal publications:

1. Scala M, Tomati V, Ferla M, Lena M, Cohen JS, Fatemi A, Brokamp E, Bican A, Phillips JA 3rd, Koziura ME, Nicouleau M, Rio M, Siquier K, Boddaert N, Musante I, Tamburro S, Baldassari S,

- Iacomino M, Scudieri P; Undiagnosed Diseases Network; Rosenfeld JA, Bellus G, Reed S, Al Saif H, Russo RS, Walsh MB, Cantagrel V, Crunk A, Gustincich S, Ruggiero SM, Fitzgerald MP, Helbig I, Striano P, Severino M, Salpietro V, Pedemonte N, Zara F. De novo variants in DENND5B cause a neurodevelopmental disorder. *Am J Hum Genet.* 2024, 111(3):529-543. doi: 10.1016/j.ajhg.2024.02.001.
2. Gorrieri G, Tamburro S, Baldassari S, Guerrisi S, Zara F, Ricci E, Maria Cordelli D, Scudieri P\*, Musante I. Generation of two iPSC lines from Mowat-Wilson syndrome patients carrying heterozygous ZEB2 mutations. *Stem Cell Res.* 2024, 76:103333. doi: 10.1016/j.scr.2024.103333. \* Corresponding author
3. Dębczyński M, Gorrieri G, Mojsak D, Guida F, Zara F, Scudieri P. ATP12A Proton Pump as an Emerging Therapeutic Target in Cystic Fibrosis and Other Respiratory Diseases. *Biomolecules.* 2023, 13(10), 1455. Doi: 10.3390/biom13101455.
4. Dębczyński M, Mojsak D, Tamburro S, Baldassari S, Musante I, Casciaro R, Ciciriello F, Zara F, Scudieri P\*, Gorrieri G. Generation of an induced pluripotent stem cell line (IGGi002A) from nasal cells of a cystic fibrosis patient homozygous for the G542X-CFTR mutation. *Stem Cell Res.* 2023, 72:103232. doi: 10.1016/j.scr.2023.103232. \* Corresponding author
5. International League Against Epilepsy Consortium on Complex Epilepsies. GWAS meta-analysis of over 29,000 people with epilepsy identifies 26 risk loci and subtype-specific genetic architecture. *Nat Genet.* 2023, 55(9):1471-1482. doi: 10.1038/s41588-023-01485-w.
6. Renda M, Barreca M, Borrelli A, Spanò V, Montalbano A, Raimondi MV, Bivacqua R, Musante I, Scudieri P, Guidone D, Buccirosi M, Genovese M, Venturini A, Bandiera T, Barraja P, Galiotta LJV. Novel tricyclic pyrrolo-quinolines as pharmacological correctors of the mutant CFTR chloride channel. *Sci Rep.* 2023, 13(1):7604. doi: 10.1038/s41598-023-34440-0.
7. Yaldiz B, Kucuk E, Hampstead J, Hofste T, Pfundt R, Corominas Galbany J, Rinne T, Yntema HG, Hoischen A, Nelen M, Gilissen C; Solve-RD consortium. Twist exome capture allows for lower average sequence coverage in clinical exome sequencing. *Hum Genomics.* 2023, 17(1):39. doi: 10.1186/s40246-023-00485-5.
8. Denommé-Pichon AS, Matalonga L, de Boer E, Jackson A, Benetti E, Banka S, Bruel AL, Ciolfi A, Clayton-Smith J, Dallapiccola B, Duffourd Y, Ellwanger K, Fallerini C, Gilissen C, Graessner H, Haack TB, Havlovicova M, Hoischen A, Jean-Marçais N, Kleefstra T, López-Martín E, Macek M, Mencarelli MA, Moutton S, Pfundt R, Pizzi S, Posada M, Radio FC, Renieri A, Rooryck C, Ryba L, Safraou H, Schwarz M, Tartaglia M, Thauvin-Robinet C, Thevenon J, Tran Mau-Them F, Trimouille A, Votypka P, de Vries BBA, Willemsen MH, Zurek B, Verloes A, Philippe C; Solve-RD DITF-ITHACA; Solve-RD SNV-indel Working Group; Solve-RD Consortia; Orphanomix Group; Vitobello A, Vissers LELM, Faivre L. A Solve-RD ClinVar-based reanalysis of 1522 index cases from ERN-ITHACA reveals common pitfalls and misinterpretations in exome sequencing. *Genet Med.* 2023, 25(4):100018. doi: 10.1016/j.gim.2023.100018.
9. Chelleri C, Scala M, De Marco P, Guerriero V, Ognibene M, Madia F, Guerrisi S, Di Duca M, Torre M, Tamburro S, Scudieri P, Piccolo G, Mattioli G, Buffelli F, Uva P, Vozzi D, Fulcheri E, Striano P, Diana MC, Zara F. Somatic Double Inactivation of NF1 Associated with NF1-Related Pectus Excavatum Deformity. *Hum Mutat.* 2023, 2023: 3160653. Doi: 10.1155/2023/3160653.
10. Guidone D, Buccirosi M, Scudieri P, Genovese M, Sarnataro S, De Cegli R, Cresta F, Terlizzi V, Planelles G, Crambert G, Sermet-Gaudelus I, Galiotta LJ. Airway surface hyperviscosity and defective mucociliary transport by IL-17/TNF- $\alpha$  are corrected by beta-adrenergic stimulus. *JCI Insight.* 2022: e164944. doi: 10.1172/jci.insight.164944.
11. Scudieri P, Pusch M. Take a big sip and shrink it with ASOR. *Pflugers Arch.* 2022; 474(11):1121-1122. doi: 10.1007/s00424-022-02750-3.

12. Baldassari S, Cervetto C, Amato S, Fruscione F, Balagura G, Pelassa S, Musante I, Iacomino M, Traverso M, Corradi A, Scudieri P, Maura G, Marcoli M, Zara F. Vesicular Glutamate Release from Feeder-Free hiPSC-Derived Neurons. *Int J Mol Sci.* 2022, 23(18), 10545. doi: 10.3390/ijms231810545.
13. Borgia P, Baldassari S, Pedemonte N, Alkhunaizi E, D'Onofrio G, Tortora D, Calì E, Scudieri P, Balagura G, Musante I, Diana MC, Pedemonte M, Vari MS, Iacomino M, Riva A, Chimenz R, Mangano GD, Mohammadi MH, Toosi MB, Ashrafzadeh F, Imannezhad S, Karimiani EG, Accogli A, Schiaffino MC, Maghnie M, Soler MA, Echiverri K, Abrams CK, Striano P, Fortuna S, Maroofian R, Houlden H, Zara F, Fiorillo C, Salpietro V. Genotype-phenotype correlations and disease mechanisms in PEX13-related Zellweger spectrum disorders. *Orphanet J Rare Dis.* 2022, 17(1):286.
14. Golec A, Pranke I, Scudieri P, Hayes K, Dreano E, Dunlevy F, Hatton A, Downey DG, Galletta L, Sermet I. Isolation, cultivation, and application of primary respiratory epithelial cells obtained by nasal brushing, polyp samples, or lung explants. *STAR Protoc.* 2022, 3(2):101419. doi: 10.1016/j.xpro.2022.101419.
15. Philp AR, Miranda F, Gianotti A, Mansilla A, Scudieri P, Musante I, Vega G, Figueroa CD, Galletta LJV, Sarmiento JM, Flores CA. KCa3.1 differentially regulates trachea and bronchi epithelial gene expression in a chronic-asthma mouse model. *Physiol Genomics.* 2022. doi: 10.1152/physiolgenomics.00134.2021.
16. Balagura G, Xian J, Riva A, Marchese F, Ben Zeev B, Rios L, Sirsi D, Accorsi P, Amadori E, Astrea G, Baldassari S, Beccaria F, Boni A, Budetta M, Cantalupo G, Capovilla G, Cesaroni E, Chiesa V, Coppola A, Dilella R, Faggioli R, Ferrari A, Fiorini E, Madia F, Gennaro E, Giacomini T, Giordano L, Iacomino M, Lattanzi S, Marini C, Mancardi MM, Mastrangelo M, Messina T, Minetti C, Nobili L, Papa A, Parmeggiani A, Pisano T, Russo A, Salpietro V, Savasta S, Scala M, Accogli A, Scelsa B, Scudieri P, Spalice A, Specchio N, Trivisano M, Tzadok M, Valeriani M, Vari MS, Verrotti A, Vigeveno F, Vignoli A, Toonen R, Zara F, Helbig I, Striano P. Epilepsy Course and Developmental Trajectories in STXBP1-DEE. *Neurol Genet.* 2022, 8(3):e676. doi: 10.1212/NXG.0000000000000676.
17. Scala M, Drouot N, MacLennan SC, Wessels MW, Krygier M, Pavinato L, Telegrafi A, de Man SA, van Slegtenhorst M, Iacomino M, Madia F, Scudieri P, Uva P, Giacomini T, Nobile G, Mancardi MM, Balagura G, Galloni GB, Verrotti A, Umair M, Khan A, Liebelt J, Schmidts M, Langer T, Brusco A, Lipska-Ziętkiewicz BS, Saris JJ, Charlet-Berguerand N, Zara F, Striano P, Piton A. De novo truncating NOVA2 variants affect alternative splicing and lead to heterogeneous neurodevelopmental phenotypes. *Hum Mutat.* 2022. doi: 10.1002/humu.24414.
18. Riva A, Nobile G, Giacomini T, Ognibene M, Scala M, Balagura G, Madia F, Accogli A, Romano F, Tortora D, Severino M, Scudieri P, Baldassari S, Musante I, Uva P, Salpietro V, Torella A, Nigro V, Capra V, Nobili L, Striano P, Mancardi MM, Zara F, Iacomino M. A Phenotypic-Driven Approach for the Diagnosis of WOREE Syndrome. *Front Pediatr.* 2022, 10:847549. doi: 10.3389/fped.2022.847549.
19. Schwarz N, Seiffert S, Pendziwiat M, Rademacher AV, Brünger T, Hedrich UBS, Augustijn PB, Baier H, Bayat A, Bisulli F, Buono RJ, Bruria BZ, Doyle MG, Guerrini R, Heimer G, Iacomino M, Kearney H, Klein KM, Kousiappa I, Kunz WS, Lerche H, Licchetta L, Lohmann E, Minardi R, McDonald M, Montgomery S, Mulahasanovic L, Oegema R, Ortal B, Papacostas SS, Ragona F, Granata T, Reif PS, Rosenow F, Rothschild A, Scudieri P, Striano P, Tinuper P, Tanteles GA, Vetro A, Zahnert F, Goldberg EM, Zara F, Lal D, May P, Muhle H, Helbig I, Weber Y. Spectrum of Phenotypic, Genetic, and Functional Characteristics in Patients With Epilepsy With KCNC2

Pathogenic Variants. *Neurology*. 2022, 98(20): e2046-e2059. doi: 10.1212/WNL.000000000000200660.

20. Accogli A, Lu S, Musante I, Scudieri P, Rosenfeld JA, Severino M, Baldassari S, Iacomino M, Riva A, Balagura G, Piccolo G, Minetti C, Roberto D, Xia F, Razak R, Lawrence E, Hussein M, Chang EY, Holick M, Calì E, Aliberto E, De-Sarro R, Gambardella A, Network UD, Group SS, Emrick L, McCaffery PJA, Clagett-Dame M, Marcogliese PC, Bellen HJ, Lalani SR, Zara F, Striano P, Salpietro V. Loss of Neuron Navigator 2 Impairs Brain and Cerebellar Development. *Cerebellum*. 2022. doi: 10.1007/s12311-022-01379-3.
21. Gorrieri G, Zara F, Scudieri P. SLC26A9 as a Potential Modifier and Therapeutic Target in Cystic Fibrosis Lung Disease. *Biomolecules*. 2022, 12(2):202. doi: 10.3390/biom12020202.
22. Raffaghello L, Principi E, Baratto S, Panicucci C, Pintus S, Antonini F, Del Zotto G, Benzi A, Bruzzone S, Scudieri P, Minetti C, Gazzero E, Bruno C. P2X7 Receptor Antagonist Reduces Fibrosis and Inflammation in a Mouse Model of Alpha-Sarcoglycan Muscular Dystrophy. *Pharmaceuticals* (Basel). 2022, 15(1):89. doi: 10.3390/ph15010089.
23. Venturini A, Borrelli A, Musante I, Scudieri P, Capurro V, Renda M, Pedemonte N, Galiotta LJV. Comprehensive Analysis of Combinatorial Pharmacological Treatments to Correct Nonsense Mutations in the CFTR Gene. *Int J Mol Sci*. 2021, 22(21):11972. doi: 10.3390/ijms222111972
24. Scala M, Anijs M, Battini R, Madia F, Capra V, Scudieri P, Verrotti A, Zara F, Minetti C, Vernes SC, Striano P. Hyperkinetic stereotyped movements in a boy with biallelic CNTNAP2 variants. *Ital J Pediatr*. 2021, 47(1):208. doi: 10.1186/s13052-021-01162-w
25. COVID-19 Host Genetics Initiative. Mapping the human genetic architecture of COVID-19. *Nature*. 2021, 600(7889):472-477. doi: 10.1038/s41586-021-03767-x
26. Accogli A, Wiegand G, Scala M, Cerminara C, Iacomino M, Riva A, Carlini B, Camerota L, Belcastro V, Prontera P, Fernández-Jaén A, Bebek N, Scudieri P, Baldassari S, Salpietro V, Novelli G, De Luca C, von Stülpnagel C, Kluger F, Kluger GJ, Wohlrab GC, Ramantani G, Lewis-Smith D, Thomas RH, Lai M, Verrotti A, Striano S, Depienne C, Minetti C, Benfenati F, Brancati F, Zara F, Striano P. Clinical and Genetic Features in Patients With Reflex Bathing Epilepsy. *Neurology*. 2021, 97(6):e577-e586. doi: 10.1212/WNL.00000000000012298
27. Scala M, Schiavetti I, Madia F, Chelleri C, Piccolo G, Accogli A, Riva A, Salpietro V, Bocciardi R, Morcaldi G, Di Duca M, Caroli F, Verrico A, Milanaccio C, Viglizzo G, Traverso M, Baldassari S, Scudieri P, Iacomino M, Piatelli G, Minetti C, Striano P, Garrè ML, De Marco P, Diana MC, Capra V, Pavanello M, Zara F. Genotype-Phenotype Correlations in Neurofibromatosis Type 1: A Single-Center Cohort Study. *Cancers*. 2021, 13(8):1879. doi: 10.3390/cancers13081879
28. Baldassari S, Musante I, Iacomino M, Zara F, Salpietro V, Scudieri P. Brain organoids as model systems for genetic neurodevelopmental disorders. *Front. Cell Dev. Biol*. 2020, 8:590119. doi: 10.3389/fcell.2020.590119
29. Scudieri P, Musante I, Venturini A, Guidone D, Genovese M, Cresta F, Caci E, Palleschi A, Poeta M, Santamaria F, Ciciriello F, Lucidi V, Galiotta LJV. Ionocytes and CFTR Chloride Channel Expression in Normal and Cystic Fibrosis Nasal and Bronchial Epithelial Cells. *Cells*. 2020, 9(9):E2090. doi: 10.3390/cells9092090
30. Naso F, Intartaglia D, Falanga D, Soldati C, Polishchuk E, Giamundo G, Tiberi P, Marrocco E, Scudieri P, Di Malta C, Trapani I, Nusco E, Salierno FG, Surace EM, Galiotta L, Banfi S, Auricchio A, Ballabio A, Medina DL, Conte I. The light-responsive miR-211 regulates retinal cell clearance by modulating lysosome biogenesis via Ezrin targeting. *Embo J*. 2020, 10:e102468. doi: 10.15252/embj.2019102468
31. Pedemonte N, Bertozzi F, Caci E, Sorana F, di Fruscia P, Tomati V, Ferrera L, Rodríguez-Gimeno A, Berti F, Pesce E, Sondo E, Gianotti A, Scudieri P, Bandiera T, Galiotta L. Discovery of a

- picomolar potency pharmacological corrector of the mutant CFTR chloride channel. *Sci Adv.* 2020, 6:eaay9669. doi: 10.1126/sciadv.aay9669
32. Genovese M, Borrelli A, Venturini A, Guidone D, Caci E, Viscido G, Gambardella G, Di Bernardo D, Scudieri P, Galletta L. TRPV4 and purinergic receptor signaling pathways are separately linked in airway epithelia to CFTR and TMEM16A chloride channels. *J Physiol.* 2019, 597(24):5859-5878. doi: 10.1113/JP278784
  33. Maniero C, Scudieri P, Shaikh LH, Zhao W, Gurnell M, Galletta L, Brown M. ANO4 is a novel marker of zona glomerulosa that regulates stimulated aldosterone secretion. *Hypertension.* 2019, 74(5):1152-1159. doi: 10.1161/HYPERTENSIONAHA.119.13287
  34. Spanò V, Montalbano A, Carbone A, Scudieri P, Galletta LJ, Barraja P. An overview on chemical structures as  $\Delta F508$ -CFTR correctors. *Eur J Med Chem.* 2019, 180:430-448. doi: 10.1016/j.ejmech.2019.07.037
  35. Vega G, Guequén A, Johansson MEV, Arike L, Martínez-Abad B, Nyström EEL, Scudieri P, Pedemonte N, Millar-Büchner P, Philp AR, Galletta LJ, Hansson GC, Flores CA. Normal Calcium-Activated Anion Secretion in a Mouse Selectively Lacking TMEM16A in Intestinal Epithelium. *Front Physiol.* 2019, 10:1040. doi: 10.3389/fphys.2019.00694
  36. Musante I, Scudieri P, Venturini A, Guidone D, Caci E, Castellani S, Conese M, Galletta LJ. Peripheral localization of the epithelial sodium channel in the apical membrane of bronchial epithelial cells. *Exp Physiol.* 2019, 104(6):866-875. doi: 10.1113/EP087590
  37. Amato F, Scudieri P, Musante I, Tomati V, Caci E, Comegna M, Maietta S, Manzoni F, Di Lullo AM, De Wachter E, Vanderhelst E, Terlizzi V, Braggion C, Castaldo G, Galletta LJ. Two CFTR mutations within codon 970 differently impact on the chloride channel functionality. *Hum Mutat.* 2019, 40(6):742-748. doi: 10.1002/humu.23741
  38. Gazzero E, Baratto S, Assereto S, Baldassari S, Panicucci C, Raffaghello L, Scudieri P, De Battista D, Fiorillo C, Volpi S, Chaabane L, Malnati M, Messina G, Bruzzone S, Traggiai E, Grassi F, Minetti C, Bruno C. The danger signal extracellular ATP is involved in the immuno-mediated damage of alpha-sarcoglycan deficient muscular dystrophy. *Am J Pathol.* 2019, 189(2):354-369. doi: 10.1016/j.ajpath.2018.10.008
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