BIOGRAPHICAL SKETCH
Instituto Paulo Gontijo ALS Award

NAME	**POSITION TITLE**
Edor Kabashi	Principal Investigator, Institute of Brain and Spinal Cord (ICM), Paris, FRANCE

EDUCATION/TRAINING

INSTITUTION AND LOCATION	DEGREE	YEAR(s)	FIELD OF STUDY
Department of Biology, Mount Saint Mary's College, Emmitsburg, MD, USA		1995-1997	Biology and Psychology
Department of Biology, Faculty of Science, McGill University, Montreal, QC, Canada	BSc (Hon.)	1998-2000	Molecular Neurobiology
Montreal Neurological Institute, McGill University, Montreal, QC, Canada	PhD	2001-2006	Role of Ubiquitin-Proteasome Pathways in ALS
Centre of Excellence in Neuromics, Universite de Montreal, Montreal, QC, Canada		2006-2009	Molecular Biology and Genetics of ALS
University Pierre and Marie Curie, Paris France	HdR Habilitation	2014	Genetics of neurodegeneration

A. POSITIONS AND HONORS

CURRENT POSITIONS

2013 Permanent Scientist (CR1) and Atip/Avenir Researcher, Institut National de la Santé et de la Recherche Médicale (Inserm) Website: www.inserm.fr

2011 Principal Investigator, Institute of Brain and Spinal Cord (ICM) at CHU Pitié-Salpêtrière, Paris, FRANCE Website: www.icm-institute.org Lab website: https://sites.google.com/site/kabashilab/

PREVIOUS POSITIONS

2009-2014 Associate Professor, Dept of Pathology and Cell Biology, University of Montreal, CANADA
2006 – 2009 Post-doctoral Fellowship, Centre of Excellence at University of Montreal, Montreal, CANADA
2001– 2006 PhD studentship, Montreal Neurological Institute and McGill University, Montreal, CANADA

Selected Honors and Awards (** One fellowship awarded per year*)

2016 ERC Consolidator Grant, European Research Council
2016 Academic Award of Excellence, Albanian-Canadian Excellence (ACE), Canada
2015 *Young Investigator Award, European Network for the Cure of ALS, Ireland
2015 Prime d'excellence, Inserm, Comission CSS8, Paris, France
2014 Laureat des Sciences Medicales, Association France Alzheimer, France
2014 Habilitation to supervise PhD students (HDR), UPMC, Paris, France
2013 1st place in National Concours as Permanent Researcher CR1; Inserm, France
2011 – 2014 Equipe ATIP/AVENIR, INSERM/CNRS, Paris, France
2011 – 2013 Contrat de Durée Determinée, INSERM, Paris, France
2009 – 2012 Development Grant, Muscular Dystrophy Association, Tucson, USA
2009 and 2011 Brain Star Awards, CIHR Institute of Neuroscience, Canada
2009 – 2011 *Herbert Jasper Fellowship, GRSNC/University of Montreal.
2006 – 2009 *Tim Noel Fellowship, CIHR/ALS Canada Partnership, Canada
2008 *Grand Prix d'Excellence, Jornees Genetiques, RGMA, Quebec City
2004 – 2006 *Doctoral Studentship, CIHR/ALS Canada Partnership
2004 – 2005 Studentship Research Award, Faculty of Medicine, McGill University
2001 BSc. with Honours, McGill University, Major in Biology
1999 Award for work in the Kosovar Refugee Camp, Gov't of Canada.
1998 Mention bien, Universite de Sorbonne (Paris V)
1997 – 1998 Dean Miles Honor Society, Mount Saint Mary's College, USA
1996 – 1998 Academic Achievement Scholarship, Mount Saint Mary's College
1996 Presidential Award, Rogers High School, Newport, USA

B. SELECTED PEER-REVIEWED PUBLICATIONS

* Corresponding Author; H-index=23; i-10 = 32; 2600+ citations; 23 published articles as main author.

1. Ciura S, Sellier C, Campanari M-L, Charlet-Berguerand N, **Kabashi E.** The most prevalent genetic cause of ALS-FTD, C9orf72 synergizes the toxicity of ATXN2 intermediate polyglutamine repeats through the autophagy pathway. **Autophagy** *in press.* http://dx.doi.org/10.1080/15548627.2016.1189070 (***IF 11.8***)
2. **Lesage S, …, Kabashi E,** Singleton A, Corti O, Brice A. Loss of VPS13C Function in Autosomal-Recessive Parkinsonism Causes Mitochondrial Dysfunction and Increases PINK1/Parkin-Dependent Mitophagy. **Am J Hum Genet.** 2016 Mar 3;98(3):500-13. (**Citations 1**; ***IF 11.2***)
3. Pradat PF, **Kabashi E,** Desnuelle C. Deciphering spreading mechanisms in amyotrophic lateral sclerosis: clinical evidence and potential molecular processes. **Curr Opin Neurol.** 2015 Oct;28(5):455-61. (**Citations 1**; ***IF 5.3***)
4. Sellier C, Campanari ML, Julie Corbier C, Gaucherot A, Kolb-Cheynel I, Oulad-Abdelghani M, Ruffenach F, Page A, Ciura S, **Kabashi E**, Charlet-Berguerand N. Loss of C9ORF72 impairs autophagy and synergizes with polyQ Ataxin-2 to induce motor neuron dysfunction and cell death. **EMBO J.** 2016 Apr 21. pii: e201593350. (**Citations 1**; ***IF 10.2***) *News and views.*
5. Ramanoudjame L, Rocancourt C, Lainé J, Klein A, Joassard L, Gartioux C, Fleury M, Lyphout L, **Kabashi E**, Ciura S, Cousin X, Allamand V. Two novel COLVI long chains in zebrafish that are essential for muscle development. **Hum Mol Genet.** 2015 Dec 1;24(23):6624-39. (**Citations 1**; ***IF 8.1***)
6. Rau F, Lainé J, Ramanoudjame L, Ferry A, Arandel L, Delalande O, Jollet A, Dingli F, Lee KY, Peccate C, Lorain S, **Kabashi E**, Athanasopoulos T, Koo T, Loew D, Swanson MS, Le Rumeur E, Dickson G, Allamand V, Marie J, Furling D. Abnormal splicing switch of DMD's penultimate exon compromises muscle fibre maintenance in myotonic dystrophy. Nat Commun. 2015 May 28;6:7205 (**Citations 4;** ***IF 10.7***)
7. Lattante S, Ciura S, Rouleau GA, **Kabashi E***. Defining the genetic connection linking ALS with FTD. **Trends in Genetics** 2015 May;31(5):263-273. (**Citations 17;** ***IF 11.6***) *Cover image,*
8. Lattante S, de Calbiac H, Le Ber I, Brice A, Ciura S, **Kabashi E***. Sqstm1 knock-down causes a locomotor phenotype ameliorated by rapamycin in a zebrafish model of ALS/FTLD. **Hum Mol Genet.** 2015 Mar 15;24(6):1682-90. (**Citations 5**; ***IF 8.1***)
9. Lattante S, Millecamps S, Stevanin G, Rivaud-Péchoux S, Moigneu C, Camuzat A, Da Barroca S, Mundwiller E, Couarch P, Salachas F, Hannequin D, Meininger V, Pasquier F, Seilhean D, Couratier P, Danel-Brunaud V, Bonnet AM, Tranchant C, LeGuern E, Brice A, Le Ber I, **Kabashi E***. Contribution of ATXN2 intermediary polyQ expansions in a spectrum of neurodegenerative disorders. **Neurology.** *2014 Sep 9;83(11):990-5.*(**Citations 17;** ***IF 8.0***) *News and Views.*
10. Lattante S, Le Ber I, Galimberti D, Serpente M, Rivaud S, Camuzat A, Clot F, Fenoglio C, Scarpini E, Brice A, **Kabashi E***. Defining the association of TMEM106B variants among frontotemporal lobar degeneration patients with GRN mutations and C9orf72 repeat expansions. **Neurobiol Aging.** 2014 Jun 28. pii: S0197-4580(14)00461-8. (**Citations 3***; IF 6.6*).
11. Patten SA, Armstrong GA, Lissouba A, Kabashi E, Parker JA, Drapeau P. Fishing for causes and cures of motor neuron disorders. **Dis Model Mech.** 2014 Jul;7(7):799-809. (**Citations 17;** ***IF 5.0***).
12. Le Ber I, De Septenville A, Guerreiro R, Bras J, Camuzat A, Caroppo P, Lattante S, Couarch P, **Kabashi E**, Bouya-Ahmed K, Dubois B, Brice A. Homozygous TREM2 mutation in a family with atypical frontotemporal dementia. **Neurobiol Aging**. 2014 Oct;35(10):2419.e23-5. (**Citations 15***; IF 6.6*).
13. Le Ber I, Van Bortel I, Nicolas G, Bouya-Ahmed K, Camuzat A, Wallon D, De Septenville A, Latouche M, Lattante S, Kabashi E, Jornea L, Hannequin D, Brice A; French research Network hnRNPA2B1 and hnRNPA1 mutations are rare in patients with "multisystem proteinopathy" and frontotemporal lobar degeneration phenotypes. **Neurobiol Aging.** 2014 Apr;35(4):934.e5-6. (**Citations 12***; IF 6.6).*
14. Ciura S, Lattante S, Le Ber I, Latouche M, Tostivint H, Brice A, **Kabashi E***. Loss of function of C9orf72 causes motor deficits in a zebrafish model of ALS. **Ann Neurol** 74, 180-187 (**Citations 102***; IF 11.1). Featured in Wall Street Journal, MSN, Yahoo, La Presse, Metro, CBC, etc.*
15. Lattante S, Le Ber I, Camuzat A, Ciura S, Brice A, **Kabashi E***. TREM2 Mutations Are Rare in a French Cohort of Patients with Frontotemporal Dementia. **Neurobio Aging** (**Citations 2***; IF 6.6).*
16. Lattante S, Le Ber I, Camuzat A, Pariente J, Brice A, **Kabashi E***. Screening UBQLN-2 in French frontotemporal lobar degeneration and frontotemporal lobar degeneration-amyotrophic lateral sclerosis patients. **Neurobiol Aging**. 2013 Aug;34(8):2078.e5-6. ***(Citations 21; IF 6.6)***.
17. Vaccaro A, Patten SA, Aggad D, Julien C, Maios C, **Kabashi E**, Drapeau P, Parker JA. Pharmacological reduction of ER stress protects against TDP-43 neuronal toxicity in vivo. **Neurobiol Dis.** 2013 Jul;55:64-75 ***(*Citations 49**; ***IF 5.3)***.

18. Lattante S, Rouleau GA, **Kabashi E***.TARDBP and FUS Mutations Associated with ALS: Summary and Update. **Hum Mutat.** 2013 Jun;34(6):812-26. (**Citations 74**; ***IF 5.7)***
19. Le Ber I, Camuzat A, Guerreiro R, Bouya-Ahmed K, Nicolas G, Gabelle A, Didic M, Millecamps S, Lenglet T, Latouche M, **Kabashi E**, Campion D, Hannequin D, Hardy J, Brice A.*SQSTM1* mutations in French FTD and FTD-ALS patients. **JAMA Neurol***. (***Citations 15; *IF 7.6)***.
20. Kabashi E*, El Oussini H, Bercier V, Gros-Louis F, Valdmanis PN, McDearmid J, Mejier IA, Dion PA, Dupre N, Hollinger D, Sinniger J, Dirrig-Grosch S, Camu W, Meininger V, Loeffler JP, René F, Drapeau P, Rouleau GA, Dupuis L. Investigating the contribution of VAPB/ALS8 loss of function in amyotrophic lateral sclerosis. **Hum Mol Genet***.* 2013 Mar 3. (**Citations 19**; ***IF 8.1***)
21. Martin E, Schüle R, Smets K, Rastetter A, Boukhris A, Loureiro JL, Gonzalez MA, Mundwiller E, Deconinck T, Wessner M, Jornea L, Oteyza AC, Durr A, Martin JJ, Schöls L, Mhiri C, Lamari F, Züchner S, De Jonghe P, **Kabashi E**, Brice A, Stevanin G. Loss of function of glucocerebrosidase GBA2 is responsible for motor neuron defects in hereditary spastic paraplegia. **Am J Hum Genet**. 2013 Feb 7;92(2):238-244. (**Citations 66**; ***IF 10.6***). *Selected by F1000.*
22. Lattante S, Le Ber I, Camuzat A, Brice A, **Kabashi E***. Mutations in the PFN1 gene are not a common cause in patients with amyotrophic lateral sclerosis and frontotemporal lobar degeneration in France. **Neurobiol Aging**. 2012 Nov 24. (**Citations 14**; ***IF 6.6***).
23. Vaccaro A, Patten SA, Ciura S, Maios C, Therrien M, Drapeau P, Parker JA, **Kabashi E***. Methylene blue protects against TDP-43 and FUS neuronal toxicity in C. elegans and D. rerio **PLoS One**. 2012;7(7):e42117. (**Citations 39**; ***IF 4.4***)
24. Martin E, Yanicostas C, Rastetter A, Naini SM, Maouedj A, **Kabashi E**, Rivaud S, Brice A, Stevanin G, Soussi-Yanicostas N. Spatacsin and spastizin act in the same pathway required for proper spinal motor neuron axon outgrowth in zebrafish. **Neurobiol Dis**. 2012 Dec;48(3):299-308. (**Citations 15**; ***IF 5.3***)
25. Kabashi E*, Bercier V, Lissouba A, Brustein E, Liao M, Rouleau GA, Drapeau P Genetic interactions between *FUS* and *TARDBP* but not *SOD1* in an *in vivo* ALS model. **PLoS Genetics** 2011 Aug;7(8):e1002214. Epub 2011 Aug 4 **(Citations 54; *IF 9.5***) ***** *Brain Star Award and Selected by F1000.*
26. Kabashi E, Champagne N, Brustein E, Drapeau P. In the swim of things: recent insights to neurogenetic disorders from zebrafish. ***Trends Genet.*** 2010 Aug;26(8):373-81. (**Citations 26; *IF 11.6***)
27. Kabashi E, Brustein E, Champagne N, Drapeau P. Zebrafish models for the functional genomics of neurogenetic disorders. **Biochim Biophys Acta**. 2010 Sep 28. (**Citations 44; *IF 5.2***)
28. Kabashi E, Lin L, Tradewell M, Dion P, Bourgouin P, Rochefort D, Durham H, Rouleau GA, Drapeau P. Gain and loss of function of ALS-related mutations of TARDBP (TDP-43) cause motor deficits in vivo. *Hum Mol Genet* 2010 Feb 15;19(4):671-83. (**Citations 221; *IF 8.1***)
29. Belzil VV, Valdmanis PN, Dion PA, Daoud H, **Kabashi E**, Noreau A, Gauthier J, Hince P, Desjarlais A, Bouchard JP, Lacomblez L, Salachas F, Pradat PF, Camu W, Meininger V, Dupre N, and Rouleau GA. Mutations in FUS cause FALS and SALS in French and French Canadian populations. **Neurology**. 2009 Oct;73:1176-9. (**Citations 105; *IF 8.0***)
30. Kabashi E, Daoud H, Rivière JB, Bourgouin P, Provencher P, Pourcher E, Dion P, Dupré N, Rouleau GA. No *TARDBP* mutations in a French-Canadian population of patients with Parkinson's disease. **Arch Neurol**. 2009 Feb;66(2):281-2. (**Citations 8; *IF 7.1***)
31. Daoud H, Valdmanis PN, **Kabashi E**, Dion P, Dupre N, Camu W, Meininger V, Rouleau GA. Contribution of TARDBP mutations to sporadic amyotrophic lateral sclerosis. **J Med Genet.** 2008 Oct 17. 46(2):112-4. (**Citations 123; *IF 7.0***)
32. Kabashi E, Valdmanis PN, Dion P, Rouleau GA. Reply to: Oxidized/misfolded SOD-1: the cause of all amyotrophic lateral sclerosis? **Ann Neurol.** 2008 Sep 28;64(3):358. (***IF 10.8***)
33. Valdmanis PN, **Kabashi E**, Dyck A, Hince P, Lee J, Dion P, D'Amour M, Souchon F, Bouchard JP, Salachas F, Meininger V, Andersen PM, Camu W, Dupré N, Rouleau GA. Association of paraoxonase gene cluster polymorphisms with ALS in France, Quebec, and Sweden. **Neurology.** 2008 Aug 12;71(7):514-20. (**Citations 34; *IF 8.0***)
34. Gros-Louis F, Kriz J, **Kabashi E**, McDearmid J, Millecamps S, Urushitani M, Lin L, Dion P, Zhu Q, Drapeau P, Julien JP, Rouleau GA. Als2 mRNA splicing variants detected in KO mice rescue severe motor dysfunction phenotype in Als2 knock-down zebrafish. **Hum Mol Genet**. 2008 Sep 1;17(17):2691-702. (**Citations 29; *IF 8.1***)
35. Kabashi E, Valdmanis PN, Dion P, Spiegelman D, McConkey BJ, Vande Velde C, Bouchard JP, Lacomblez L, Pochigaeva K, Salachas F, Pradat PF, Camu W, Meininger V, Dupre N, Rouleau GA. TARDBP mutations in individuals with sporadic and familial amyotrophic lateral sclerosis. **Nat Genet.** 2008 May;40(5):572-4. (**Citations 936; *IF 36.4***) *Brain Star Award and Featured in Presse Canada.*

36. Kabashi E, Agar JN, Hong Y, Taylor DM, Minotti S, Figlewicz DA, Durham HD. Proteasomes remain intact, but show early focal alteration in their composition in a mouse model of ALS. **J Neurochem**. 2008 Apr;105(6):**2353-2366**. (**Citations 22; *IF 4.3***)

37. Valdmanis PN, Meijier I, **Kabashi E** et al. Functional characterization of strumpellin, mutated in hereditary spastic paraplegia. ***Neuromuscular Disorders*** 2007 17(9-10) 893. (**Citations 0; *IF 2.8***)

38. Kabashi E, Valdmanis PN, Dion P, Rouleau GA. Oxidized/misfolded superoxide dismutase-1: the cause of all amyotrophic lateral sclerosis? **Ann Neurol.** 2007 Dec;62(6):553-9. (**Citations 84; *IF 10.8***)

39. Valdmanis PN, **Kabashi E**, Dion PA, Rouleau GA. ALS predisposition modifiers: knock NOX, who's there? SOD1 mice still are***. Eur J Hum Genet.*** 2008 Feb;16:140-2. (**Citations 4; *IF 3.6***)

40. Taylor DM, Gibbs BF, **Kabashi E**, Minotti S, Durham HD, Agar JN. Tryptophan 32 potentiates aggregation and cytotoxicity of a copper/zinc superoxide dismutase mutant associated with familial amyotrophic lateral sclerosis. **J Biol Chem**. 2007 Jun 1;282(22)16329-35. (**Citations 26; *IF 5.3***)

41. Kabashi E, Durham HD. Failure of protein quality control in amyotrophic lateral sclerosis. **Biochim Biophys Acta**. 2006 Nov-Dec;1762(11-12):1038-50. (**Citations 65; *IF 5.2***)

42. Taylor DM, **Kabashi E**, Agar JN, Minotti S, Durham HD. Proteasome activity or expression is not altered by activation of the heat shock transcription factor Hsf1 in cultured fibroblasts or myoblasts. **Cell Stress Chaperones**. 2005 Autumn;10(3):230-41. (**Citations 4; *IF 3.2***)

43. Kabashi E, Agar JN, Taylor DM, Minotti SM, Durham HD. (2004) Focal Dysfunction of the Proteasome: A Pathogenic Factor in a Mouse Model of ALS. **J Neurochem.** 89:1325-35. (**Citations 107; *IF 4.3***)

Co-author of 15 additional papers from the ALS-Untangled Consortium, two book chapters and numerous meeting abstracts.

Supervision

Period 2011-2015 as a group leader at the ICM: *In italics-current lab members*, *-common publications.
Post-Doctoral Fellows: **Seven,** three of which are current lab members: *Sorana Ciura * (2011- Embo and AFM-Telethon fellow)*, *Doris-Lou Demy (2015- ERA-NET fellow)*, *Maria-Letizia Campanari (2015- ICM fellow)*, Serena Lattante * (2012-2015 ENP fellow; currently Researcher at Università Cattolica del Sacro Cuore, Rome Italy), Claire Pujol * (2013-2014) and Ara Petcu (2012-2013).
PhD Students: Three: *Hortense de Calbiac * (2014-* FRM fellow), Adriana Dabacan (2014- co-supervised with Raul Muresan, Romania). Alexandra Lissouba * (2010- co-supervised with Pierre Drapeau, Canada)
MSc Students: Nine, with two current lab members: *Gabrielle Devienne (2015-), Raphael Munoz (2015-),* Hortense de Calbiac * (2012-2013), Alison Dominguez (2014), Valerie Bercier * (2013), Sarah Dayan (2013) *, Shauna Katz (2012), Remi Lebigre (2011), Brianne Chereau (2011).
Also supervised and trained one MD student; nine research assistants and three undergraduate students.

Teaching (2010-2014):

Lecturer : Pathologies & Maladies Neurodégénératives, Université Pierre et Marie Curie, 2012-2014
Lecturer: Vieillissement & Maladies Neurodégénératives, Université Paris Diderot, 2014.
Lecturer : Neurobiology of Disease, Dept of Pathology and Cell Biology, University of Montreal, 2010

Selected Platform presentations

Kabashi E (2016) C9orf72 synergizes the toxicity of ATXN2 intermediate repeats. ENCALS meeting, Milan, Italy
Kabashi E (2015) Genetic models to study ALS. ENCALS meeting, Dublin, Ireland
Kabashi E (2015) Identifying functional partners of mutant TDP-43/FUS using genetic models in zebrafish. Annual.Meeting of the Robert Packard Foundation for ALS Research.
Kabashi E (2014) Understanding the Function of Mutant TDP-43. Strasbourg France, *Breakthroughs in MND*.

Major on-going Grants

Project Title	*Funding source*	*Amount (Euros)*	*Period*	*Role of the PI*
Defining the pathogenic network in ALS	European Research Council, EU	**2,000,000**	**2016-2021**	Project supervisor and leader
Molecular partners of mutant TDP-43/FUS	Robert Packard Center for ALS; John Hopkins Univ. USA	**90,000**	**2012-2014** (renewable each year)	Project supervisor and leader
Dysregulation of RNA in the pathogenesis of ALS	ANR France, ERA-NET Research Program for rare diseases, EU	**291,000**	**2014-2017**	Project leader for WP2 and WP3; Zfish validation