

Spread of segmental/multifocal idiopathic adult-onset dystonia to a third body site

*Tommaso Ercoli*¹, R. Erro², G. Fabbrini^{3,4}, R. Pellicciari⁵, P. Girlanda⁶, C. Terranova⁶, L. Avanzino^{7,8}, F. Di Biasio⁷, P. Barone², M. Esposito⁹, G. De Joanna⁹, R. Eleopra¹⁰, F. Bono¹¹, L. Manzo¹¹, A.R. Bentivoglio^{12,13}, M. Petracca¹², M.M. Mascia¹, A. Albanese¹⁴, A. Castagna¹⁵, R. Ceravolo¹⁶, M.C. Altavista¹⁷, C. Scaglione¹⁸, L. Magistrelli^{19,20}, M. Zibetti²¹, L. Bertolasi²², M. Coletti Moja²³, M.S. Cotelli²⁴, G. Cossu²⁵, B. Minafra²⁶, A. Pisani^{26,27}, S. Misceo²⁸, N. Modugno⁴, M. Romano²⁹, D. Cassano³⁰, A. Berardelli^{3,4}, G. Defazio¹

¹Department of Medical Science and Public Health, Institute of Neurology, University of Cagliari, Cagliari, Italy

²Department of Medicine, Surgery and Dentistry "Scuola Medica Salernitana", University of Salerno, Baronissi (SA), Italy

³Department of Human Neurosciences, Sapienza University of Rome, Rome, Italy

⁴IRCCS Neuromed, Pozzilli (IS), Italy

⁵Department of Basic Science, Neuroscience and Sense Organs, Aldo Moro University of Bari, Bari, Italy.

⁶Department of Clinical and Experimental Medicine, University of Messina, Messina, Italy

⁷Ospedale Policlinico San Martino - IRCCS, Genoa, Italy

⁸Department of Experimental Medicine, Section of Human Physiology, University of Genoa, Genoa, Italy

⁹Clinical Neurophysiology Unit, Cardarelli Hospital, Naples, Italy

¹⁰SOC Neurologia, AOU S. Maria della Misericordia; Neurological Unit 1, Fondazione IRCCS. Istituto Neurologico "Carlo Besta", Milan, Italy

¹¹Center for Botulinum Toxin Therapy Neurologic Unit. A.O.U. Mater Domini, Catanzaro, Italy

¹²Fondazione Policlinico Universitario Agostino Gemelli - IRCCS, Rome, Italy

¹³Institute of Neurology, Università Cattolica del Sacro Cuore, Rome, Italy

¹⁴Department of Neurology, IRCCS Istituto Clinico Humanitas, Rozzano, Milan, Italy

¹⁵IRCCS Fondazione Don Carlo Gnocchi, Milan, Italy

¹⁶Neurology Unit, Department of Clinical and Experimental Medicine, University of Pisa, Pisa, Italy

¹⁷Neurology Unit, San Filippo Neri Hospital ASL Roma 1, Roma, Italy

¹⁸IRCCS Institute of Neurological Sciences, Bologna, Italy

¹⁹Movement Disorders Centre, Neurology Unit, Department of Translational Medicine, University of Piemonte Orientale, Novara, Italy

²⁰University of Insubria, Varese, Italy

²¹Department of Neuroscience Rita Levi Montalcini, University of Turin, Turin, Italy

²²Neurologic Unit, University Hospital, Verona, Italy

²³Mauriziano Hospital Umberto I, Torino, Italy

²⁴Neurology Unit, ASST Valcamonica, Esine, Italy

²⁵Neurology Service and Stroke Unit, Department of Neuroscience, A.O. Brotzu, Cagliari, Italy

²⁶IRCCS Mondino Foundation, Pavia, Italy

²⁷Department of Brain and Behavioral Sciences, University of Pavia, Pavia, Italy

²⁸Neurologic Unit, San Paolo Hospital, Bari, Italy

²⁹Neurology Unit, AOOR Villa Sofia Cervello, Palermo, Italy

³⁰Unit of Neurology, Ospedale Maria Vittoria, Turin, Italy

Introduction: Adult-onset focal dystonia can spread to involve one, or less frequently, two additional body regions [1]. Spread of focal dystonia to a third body site is not fully characterized.

Objective: The aim of the study is to fully characterize the spread of segmental/multifocal dystonia to a third body site in idiopathic adult-onset patients.

Materials and Methods: We retrospectively analyzed data from the Italian Dystonia Registry [2], enrolling patients with segmental/multifocal dystonia involving at least two parts of the body or more. Survival analysis estimated the relationship between dystonia features and spread to a third body part.

Results: We identified 340 patients with segmental/multifocal dystonia involving at least two body parts. Spread of dystonia to a third body site occurred in 42/241 patients (17.4%) with focal onset and 10/99 patients (10.1%) with segmental/multifocal dystonia at onset. The former had a greater tendency to spread than patients with segmental/multifocal dystonia at onset. Gender, years of schooling, comorbidity, family history of dystonia/tremor, age at dystonia onset, and disease duration could not predict spread to a third body site. Among patients with focal onset in different body parts (cranial, cervical, and upper limb regions), there was no association between site of focal dystonia onset and risk of spread to a third body site.

Conclusion: Spread to a third body site occurs in a relative low percentage of patients with idiopathic adult-onset dystonia affecting two body parts. Regardless of the site of dystonia onset and of other demographic/clinical variables, focal onset seems to confer a greater risk of spread to a third body site in comparison to patients with segmental/multifocal dystonia at onset.

References:

- [1] BD Berman, CL Groth, SH Sillau, S Pirio Richardson, SA Norris, J Junker, N Brüggemann, P Agarwal, RL Barbano, AJ Espay, JA Vizcarra, C Klein, T Bäumer, S Loens, SG Reich, M Vidailhet, C Bonnet, E Rose, HA Jinnah, JS Perlmutter, Risk of spread in adult-onset isolated focal dystonia: A prospective international cohort study, *J. Neurol. Neurosurg. Psychiatry.* (2019) 1–7.
- [2] G Defazio, M Esposito, G Abbruzzese, CL Scaglione, G Fabbrini, G Ferrazzano, S Peluso, R Pellicciari, AF Gigante, G Cossu, R Arca, L Avanzino, F Bono, MR Mazza, L Bertolasi, R Bacchin, R Eleopra, C Lettieri, F Morgante, MC Altavista, L Polidori, R Liguori, S Misceo, G Squintani, M Tinazzi, R Ceravolo, E Unti, L Magistrelli, M Coletti Moja, N Modugno, M Petracca, N Tambasco, MS Cotelli, M Aguggia, A Pisani, M Romano, M Zibetti, AR Bentivoglio, A Albanese, P Girlanda, A Berardelli, The Italian Dystonia Registry: rationale, design and preliminary findings, *Neurol. Sci.* 38 (2017) 819–825.