

Clinico-pathological Study of Early-Onset Cases of Behavioral Frontotemporal Dementia

Araki Kimura ⁽¹⁾⁽³⁾⁽⁴⁾, Kenji Ikeda ⁽¹⁾⁽⁴⁾, Aki Shimosawa ⁽²⁾, Akito Nagakura ⁽³⁾⁽⁴⁾, Kenichi Oshima ⁽⁴⁾,
Daisuke Taniguchi ⁽⁵⁾, Airi Tarutani ⁽¹⁾, Takashi Nonaka ⁽¹⁾, Tadafumi Kato ⁽³⁾, Masato Hasegawa ⁽¹⁾

(1) Tokyo Metropolitan Institute of Medical Science, Dementia Research Project, (2) Tokyo Metropolitan Institute of Medical Science, Division of Molecular Pathology and Histology, (3) Juntendo University, Department of Psychiatry and Behavioral Science, (4) Tokyo Metropolitan Matsuzawa Hospital, Department of Psychiatry, (5) Juntendo University, Department of Neurology

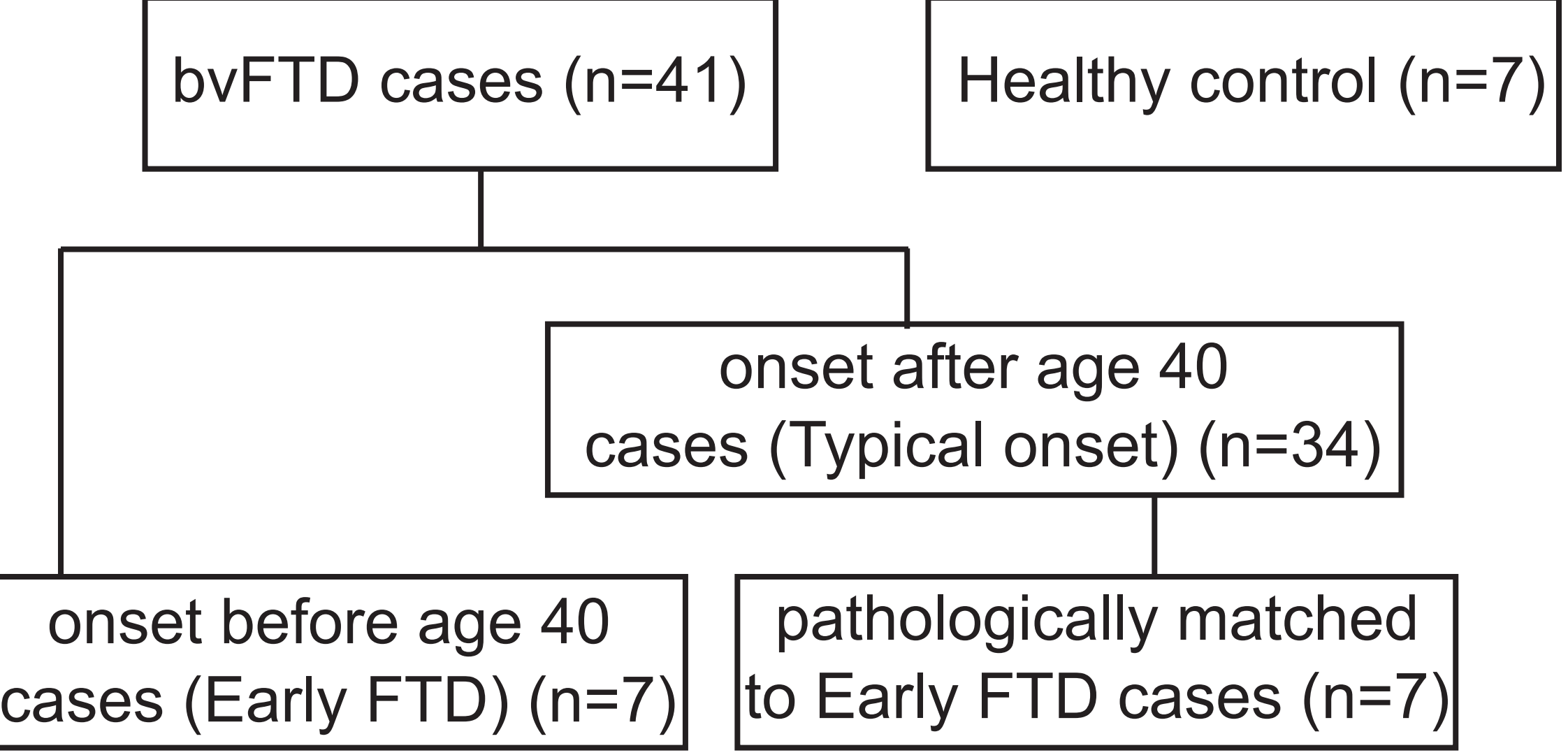


BACKGROUND

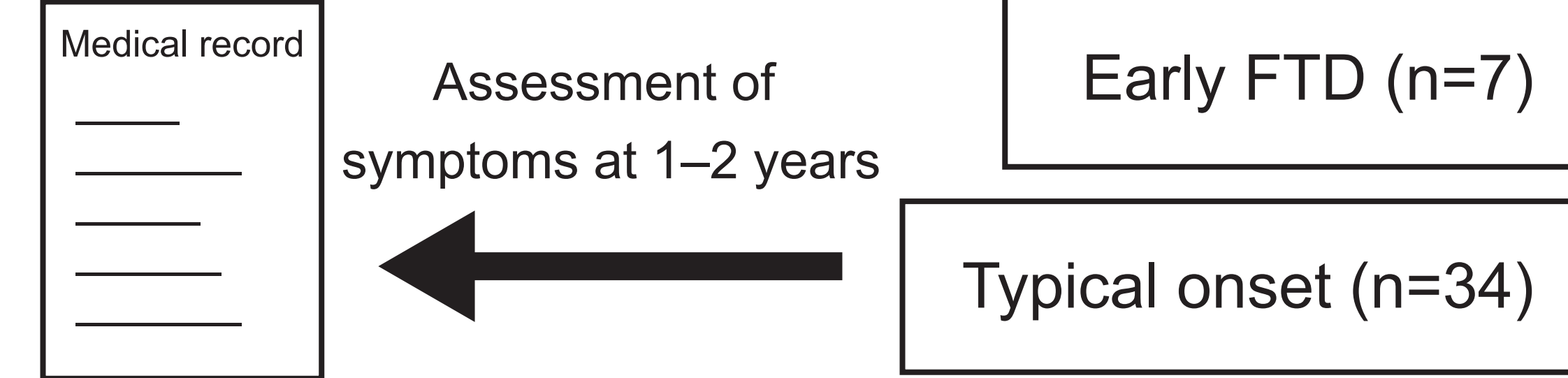
Behavioral variant frontotemporal dementia (bvFTD) cases with onset before age 40 are rare and frequently misdiagnosed as primary psychiatric disorders due to symptom overlap⁽¹⁾. This study aims to elucidate the clinicopathological characteristics of early-onset bvFTD cases.

METHODS

Participants enrollment process



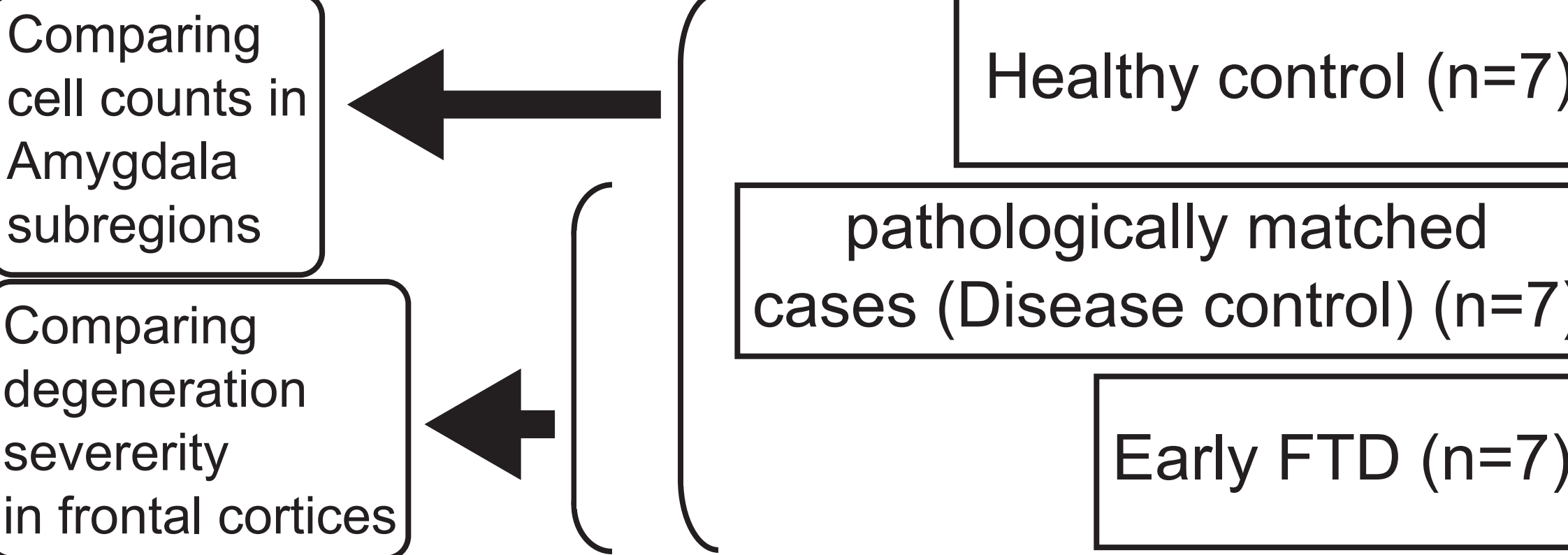
Comparison of initial symptoms



Neuropathological assessment



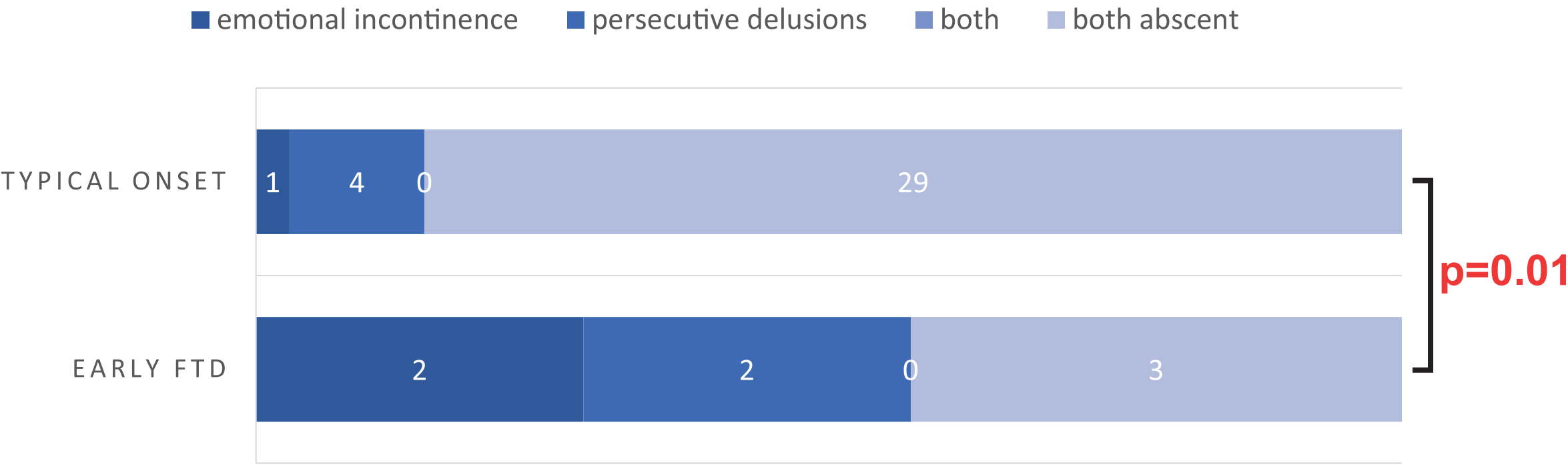
Semiquantitative analysis of specific regions



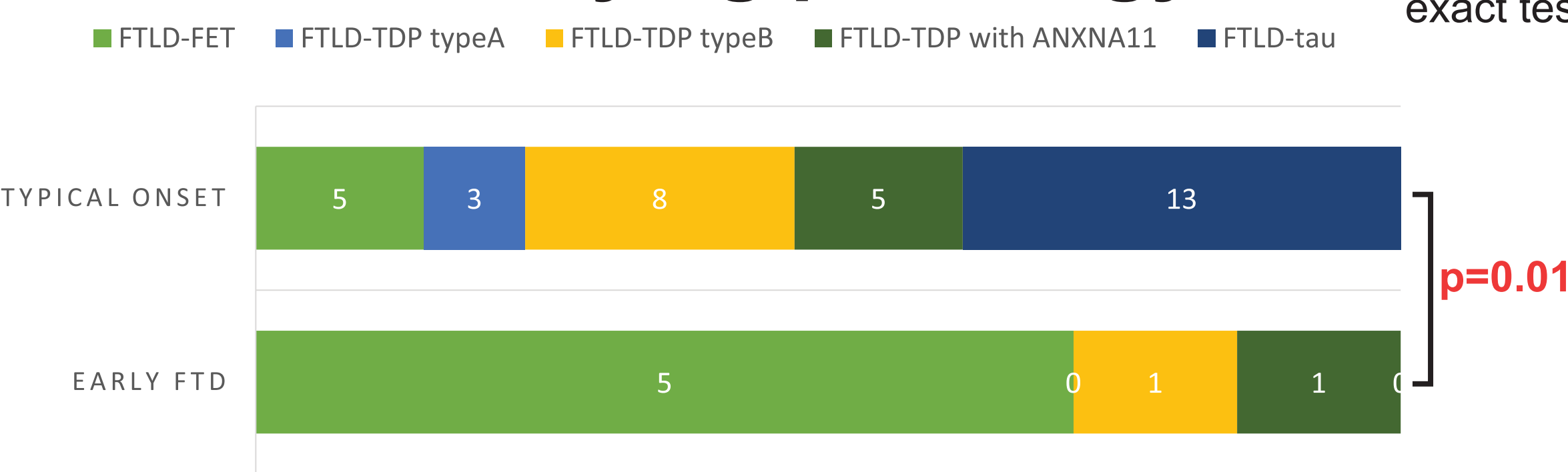
RESULTS

	Early FTD	Typical onset	Disease control	Healthy control
Number of cases	7	34	7	7
Mean age of onset (yr)	33.71	55.65	53.43	N/A
Mean age at death (yr)	44.71	62.18	58.43	48.86
Mean duration of illness (yr)	9.71	6.43	4.97	N/A
Mean brain weight (g)	967.8	1055.48	1110	1324.3
FTLD-tau	0	13	0	N/A
FTLD-TDP type A	0	3	0	N/A
FTLD-TDP type B	1	8	1	N/A
FTLD-TDP with ANXA11	1	5	2	N/A
FTLD-FET	5	5	4	N/A
TAF15 positive	5	4	4	N/A

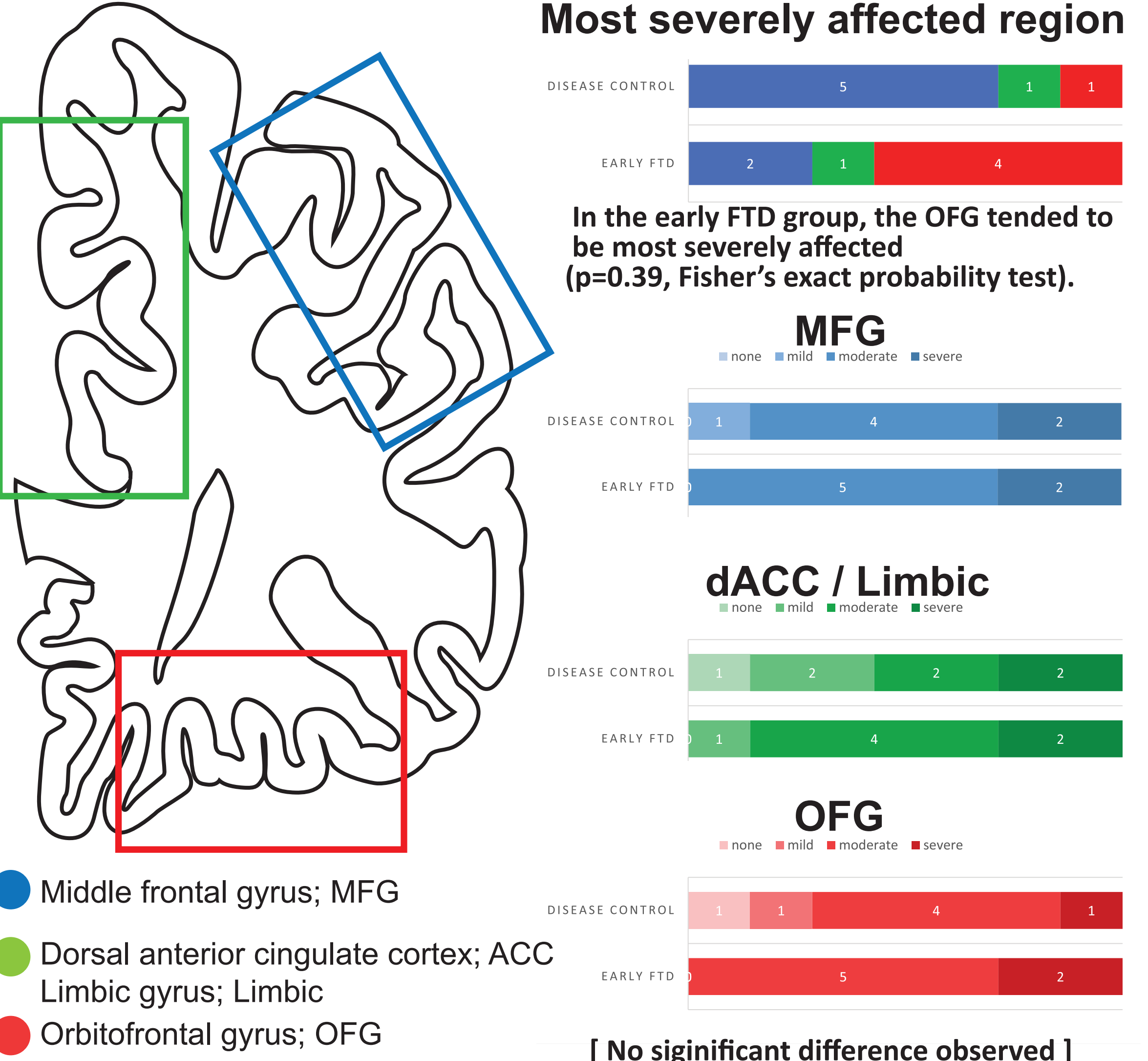
Initial symptoms



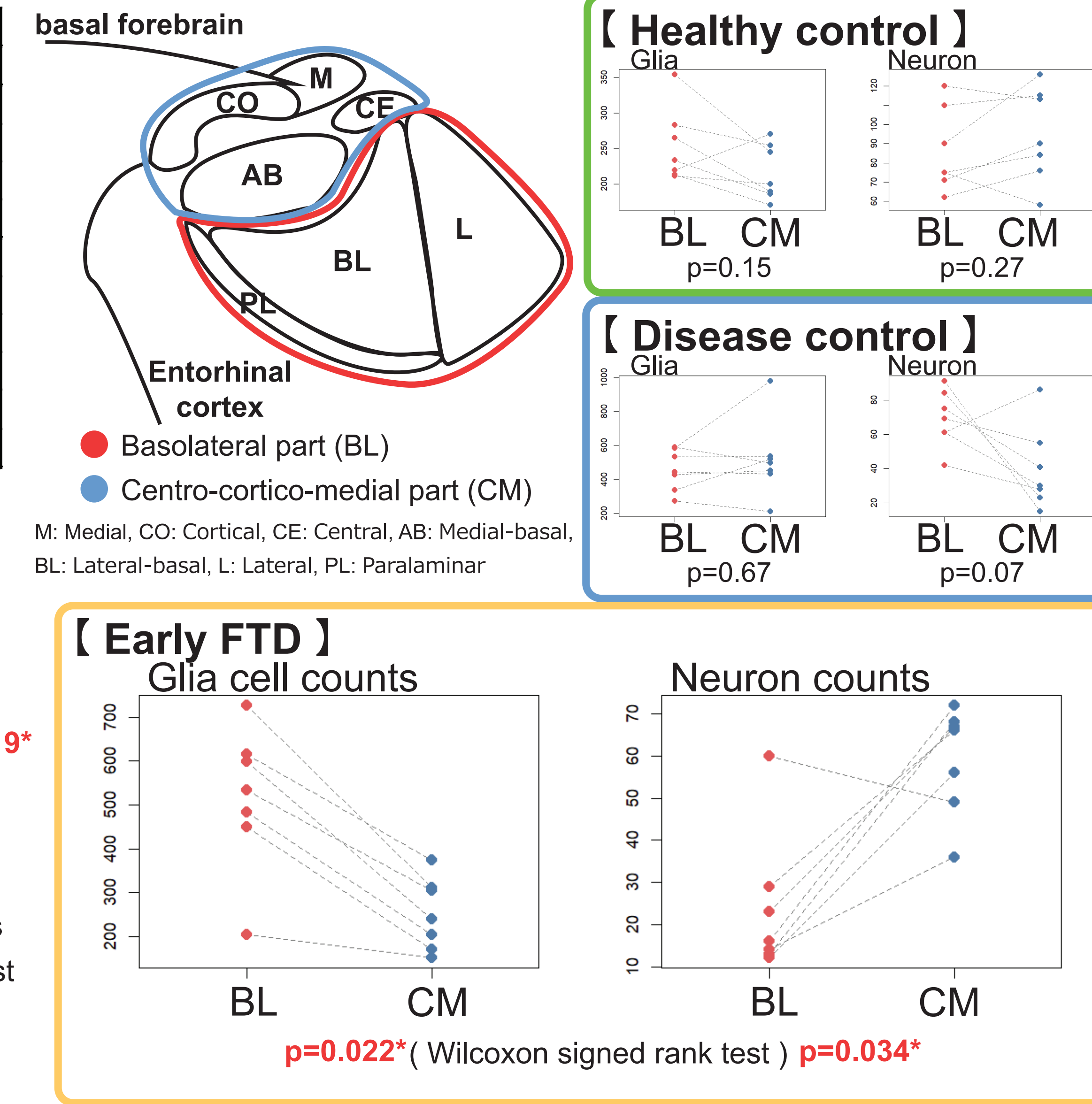
Underlying pathology



Comparison of frontal cortices



Comparison of amygdala pathology



CONCLUSION

The amygdala BL region has reciprocal connections with frontal lobes^(2, 3), and is considered to play a pivotal role in emotional regulation^(2, 4). The emotion-related symptoms observed as initial manifestations in early-onset bvFTD are consistent with the tendency towards predominant degeneration in the amygdala BL region and the OFG.

Acknowledgements

We thank the patient and family member who gave permission for research use. We are grateful to TMiMS Division of Molecular Pathology and Histology, and Matsuzawa hospital Department of Laboratory for their expert histopathological technical support.

Reference

1. Z. Chemali, et al. *Am J Alzheimers Dis other Dement.* (2010), 2. W. Gao, et al. *Neuroimage.* (2021), 3. H. Barbes, et al. *BMC Neurosci.* (2003), 4. R.D. Ray, et al. *Neurosci Biobehav Rev.* (2012)