example of an often underrecognized neurological disorder with which neuropsychologists should be familiar, this case uniquely raises ethical questions relevant to care providers and current treatment guidelines regarding genetic testing among families carrying highly heritable neurological conditions. In particular, personal ethical challenges around deciding to pursue or forego pre-symptomatic testing, and implications for family planning, highlight the importance of genetic counseling for affected families.

Categories: Stroke/Cerebrovascular Injury & Disease (Adult)
Keyword 1: genetic disorders
Keyword 2: cerebrovascular disease
Keyword 3: neuropsychological assessment
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93 Impact of Cardiovascular Risk on Cognitive and Brain Aging in Autosomal Dominant Frontotemporal Dementia

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Objective: Poor cardiovascular health occurs with age and is associated with increased dementia risk, yet its impact on frontotemporal lobar degeneration (FTLD) and autosomal dominant neurodegenerative disease has not been well established. Examining cardiovascular risk in a population with high genetic vulnerability provides an opportunity to assess the impact of lifestyle factors on brain health outcomes. In the current study, we examined whether systemic vascular burden associates with accelerated cognitive and brain aging outcomes in genetic FTLD.

Participants and Methods: 166 adults with autosomal dominant FTLD (C9orf72 *n*= 97; GRN *n*= 34; MAPT *n*= 35; 54% female; M^{age} = 47.9; $M^{education}$ = 15.6 years) enrolled in the Advancing Research and Treatment for Frontotemporal Lobar Degeneration (ARTFL) and Longitudinal Evaluation of Familial Frontotemporal Dementia Longitudinal FTD study (ALLFTD) were included. Participants completed neuroimaging and were screened for cardiovascular risk and functional impairment during a comprehensive neurobehavioral and medical interview. A vascular burden score (VBS) was created by summing vascular risk factors (VRS) [diabetes, hypertension, hyperlipidemia, and sleep apnea] and vascular diseases (VDS) [cerebrovascular disease (e.g., TIA, CVA), cardiac arrhythmia (e.g., atrial fibrillation, pacemaker, defibrillator), coronary artery disease (e.g., myocardial infarction, cardiac bypass, stent), and congestive heart failure] following a previously developed composite (range 0 to 8). We examined the interaction between each vascular health metric (VBS, VDS, VRS) and age (vascular health*age) on clinical severity (CDR plus NACC FTLD-SB), and white matter hyperintensity (WMH) volume outcomes, adjusting for age and sex. Vascular risk, disease, and overall burden scores were examined in separate models.
Results: There was a statistically significant interaction between total VBS and age on both clinical severity (β=0.20, p=0.044) and WMH burden (β=0.20, p=0.032). Mutation carriers with higher vascular burden evidenced worse clinical and WMH outcomes for their age. When breaking down the vascular burden score into (separate) vascular risk (VRS) and vascular disease (VDS) scores, the interaction between age and VRS remained significant only for WMH (β=0.26, p=0.009), but not clinical severity (β=0.04, p=0.685). On the other hand, the interaction between VDS and age remained significant only for clinical severity (β=0.20, p=0.041) but not WMH (β=0.17, p=0.066).
Conclusions: Our results demonstrate that systemic vascular burden is associated with an "accelerated aging" pattern on clinical and white matter outcomes in autosomal dominant FTLD. Specifically, mutation carriers with greater vascular burden show poorer neurobehavioral outcomes for their chronological age. When separating vascular risk from disease, risk was associated with higher age-related WMH burden, whereas disease was associated with poorer age-related clinical severity of mutation

https://doi.org/10.1017/S1355617723002977 Published online by Cambridge University Press

carriers. This pattern suggests preferential brain-related effects of vascular risk factors, while the functional impact of such factors may be more closely aligned with fulminant vascular disease. Our results suggest cardiovascular health may be an important, potentially modifiable risk factor to help mitigate the cognitive and behavioral disturbances associated with having a pathogenic variant of autosomal dominant FTLD. Future studies should continue to examine the neuropathological processes underlying the impact of cardiovascular risk in FTLD to inform more precise recommendations, particularly as it relates to lifestyle interventions.

Categories: Stroke/Cerebrovascular Injury & Disease (Adult)
Keyword 1: aging disorders
Keyword 2: cardiovascular disease
Keyword 3: vascular cognitive impairment
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94 A Case Study Using Serial Neuropsychological Assessments to Understand the Effects of Recurrent Intracerebral Hemorrhages in an Individual with Cerebral Amyloid Angiopathy

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Objective: Cerebral amyloid angiopathy (CAA) is one of the most frequent causes of non-traumatic intracerebral hemorrhage (ICH). ICH recurrence risk is significantly higher in patients with CAA than for those without the condition, and CAA is a risk factor for the development of dementia, particularly Alzheimer's disease. There is a growing body of research describing neuropsychological impairment observed in patients with CAA. Among patients with a history of CAA-related ICH, the most commonly identified cognitive impairments include attention, processing speed, executive functioning, and episodic memory. However, little is known about potential additive or synergistic effects of each CAA-related lesion (such as recurrent ICHs) on cognitive functioning.
Participants and Methods: We present a case of a 74-year-old female with sporadic CAA, who had recurrent ICHs involving the left occipitoparietal lobe, left frontoparietal lobe, right occipital lobe, and left frontal lobe. She experienced residual visual impairment and probable Charles Bonnet Syndrome. Her clinical presentation and cognitive functioning were tracked with an inpatient neuropsychological evaluation completed after each ICH occurrence within the past year, as well as an outpatient neuropsychological evaluation completed approximately 3-months post-discharge from her most recent hospital admission. Record review, including clinical notes, lab tests, and imaging results supplement her performance on serial inpatient and outpatient neuropsychological evaluations.
Results: Data from three inpatient neuropsychological screenings and one lengthier outpatient evaluation are presented. With each inpatient evaluation, her profile demonstrated further cognitive decline involving visuospatial skills, semantic fluency, and episodic memory. In fact, results from her last inpatient screening raised concern for an underlying cortical degenerative process. In contrast, her follow-up outpatient evaluation, after three separate ICH events within one year, demonstrated an isolated set-shifting impairment, with intact performance across all other domains, which ruled out the prior suspicion of a cortical process.
Conclusions: While specific domains of cognition are more vulnerable in CAA, it is difficult to identify a specific and expected cognitive pattern given the extensive number of varied neurological insults patients typically develop throughout the disease course. This case demonstrates the wide range effects of repeated ICH, as well as the contrast between the acute effects of new lesions and the lasting effects of these lesions on cognitive ability after a period of recovery and stabilization. Given that our service was able to perform neuropsychological assessment in the acute phase of each ICH and in the subacute phase after a period of stabilization, this case adds to the literature by providing an example of the additive or synergistic effects of each CAA-related lesion over time.

Categories: Stroke/Cerebrovascular Injury & Disease (Adult)

https://doi.org/10.1017/S1355617723002977 Published online by Cambridge University Press