Objective: To examine the structural integrity of the superior longitudinal fasciculi and its relationship to working memory in patients with Cerebral Autosomal Dominant Arteriopathy with Subcortical Infracts and Leukoencephalopathy (CADASIL).

Participants and Methods: Twelve patients with CADASIL and ten healthy controls underwent MRI including diffusion-tensor imaging (DTI) and cognitive assessment including a verbal n-back test. Grey matter, white matter, intracranial, and subcortical hyperintensity (SH) volumes were measured. Quantitative DTI tractography was used to assess the structural integrity of the superior and inferior longitudinal fasciculi (SLF, ILF) bilaterally and the genu and splenium of the corpus callosum.

Results: The groups did not differ significantly by age or education. The CADASIL group had higher SH volume (p=.012) and reduced structural integrity (p<.026) in the SLF bilaterally but not the left ILF, and in the splenium but not the genu of the corpus callosum. The CADASIL group had poorer performance at higher (2- and 3-back, p<.022) but not lower (0- or 1-back) working memory loads. The groups did not differ on task with minimal working memory demands (confrontational naming). In the whole sample, 2-back performance was correlated with white matter integrity of the right SLF (r=.461, p=.031) and left SLF (r=.513, p=.015) but the right ILF or splenium.

Conclusions: White matter integrity and working memory are reduced in CADASIL. The results support a role for the SLF in working memory and provide support for the utility of quantitative tractography for exploring the cognitive correlates of specific white matter tracts in healthy and clinical populations.