Corticobasal degeneration (CBD) typically presents with asymmetric dyspraxia, stiffness and dystonia (Corticobasal syndrome – CBS). Features of CBS occur in other diseases and there are no well validated clinical or biological markers that accurately predict its underlying pathology, making diagnosis difficult. We are conducting a multinational multicenter prospective longitudinal study of CBD/S funded by CBD solutions. Eligible patients undergo a standardized neurological assessment. We aim to recruit >150 patients. Of 12 cases recruited to date (7 female) 8 met Armstrong criteria for probable CBD and 4 for possible CBD. All patients exhibited limb apraxia; 91.6% akinesia; 83.3% rigidity; 75% dystonia; 58.3% oro-buccal apraxia; 50% cortico-sensory deficit; 41% alien limb phenomenon and 33.3% myoclonus. Most patients (83%) had asymmetric onset (Left: Right 1:1). The mean age at onset and at study entry was 63.9 (SD 5.83) and 70 years (SD 7.6) respectively. The average disease duration was 6.0 years (SD 3.85) and average time to diagnosis 4 years. This study will facilitate identification of specific clinical and biological markers for CBD and will ultimately contribute to the development of disease modifying therapies.
CORTICOBASAL SYNDROME AND CORTICOBASAL DEGENERATION

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