Akinetic mutism

Term describing patients tending neither to move (akinesia) nor speak (mutism). Akinetic mutism was first described in 1941 as a mental state where patients lack the ability to move or speak.

However, their eyes may follow their observer or be diverted by sound.

Patients lack most motor functions such as speech, facial expressions, and gestures, but demonstrate apparent alertness.

They exhibit reduced activity and slowness, and can speak in whispered monosyllables.

Patients often show visual fixation on their examiner, move their eyes in response to an auditory stimulus, or move after often repeated commands.

Patients with akinetic mutism are not paralyzed, but lack the will to move.

Many patients describe that as soon as they 'will' or attempt a movement, a 'counter-will' or 'resistance' rises up to meet them.

seen in Bilateral frontal lobe dysfunction, (due to bilateral anterior cerebral artery infarct after vasospasm of anterior communicating artery aneurysm rupture or with large bilateral frontal lesions; may actually be abulia) or with bilateral cingulate gyrus lesions.

Akinetic mutism is a key diagnostic feature of prion diseases, however, their rapidly progressive nature makes detailed investigation of the language disorder in a large cohort extremely challenging.

A systematic, prospective investigation of language disorders in a large cohort of patients diagnosed with prion diseases in 568 patients of the National Prion Monitoring Cohort. All patients had at least one assessment with the MRC Scale, a milestone-based functional scale with language and non-language components. Forty patients, with early symptoms and able to travel to the study site, were also administered a comprehensive battery of language tests (spontaneous speech, semantics, syntax, repetition, naming, comprehension and lexical retrieval under different conditions).

5/568 (0.9%) patients presented with leading language symptoms. Those with repeated measurements deteriorated at a slower rate in language compared to non-language milestones. Amongst the subgroup of 40 patients who underwent detailed language testing, only three tasks-semantic and phonemic fluency and sentence comprehension-were particularly vulnerable early in the disease. These tasks were highly correlated with performance on non-verbal executive tests. Patients were also impaired on a test of dynamic aphasia.

These results provide evidence that the language disorder in prion disease is rarely an isolated clinical or cognitive feature. The language abnormality is indicative of a dynamic aphasia in the context of a prominent dysexecutive syndrome, similar to that seen in patients with the degenerative movement disorder progressive supranuclear palsy (PSP) [1)].

A 72-year-old female had suffered from head trauma resulting from falling down the stairs. She was

diagnosed as subdural hematoma on the right frontal lobe and subarachnoid haemorrhage. At 5 weeks after head trauma, when starting rehabilitation, she showed no spontaneous movement or speech. She participated in a comprehensive rehabilitative management programme, including movement therapy and dopaminergic drugs, for improvement of AM. During 5 week's intensive rehabilitation, she showed gradual improvement of AM: she became able to perform some daily activities by herself including eating, dressing and walking.

On 5-week DTT, the neural connectivity of the caudate nucleus (CN) to the medial prefrontal cortex (PFC; Broadmann area [BA]: 10 and 12) and the orbitofrontal cortex (BA: 11 and 13) was decreased in both hemispheres; in contrast, the neural connectivity of the CN to the medial PFC was increased on the left side on 10-week and 6-month DTT.

Recovery of an injured prefronto-caudate tract concurrent with the improvement of AM was demonstrated in a patient with TBI, using follow-up DTTs [2].

1)
Caine D, Nihat A, Crabb P, Rudge P, Cipolotti L, Collinge J, Mead S. The language disorder of prion disease is characteristic of a dynamic aphasia and is rarely an isolated clinical feature. PLoS One. 2018 Jan 5;13(1):e0190818. doi: 10.1371/journal.pone.0190818. eCollection 2018. PubMed PMID: 29304167.
2)
Jang SH, Kim SH, Lee HD. Recovery of an injured prefronto-caudate tract in a patient with traumatic brain injury: A diffusion tensor tractography study. Brain Inj. 2017;31(11):1548-1551. doi: 10.1080/02699052.2017.1376761. Epub 2017 Sep 28. PubMed PMID: 28956643.