Abulia : No Will, No Way
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Abstract
Abulia refers to impaired ability to perform voluntary actions, show initiative, make decisions along with decrease in movements, speech, thought and emotional reactions. We describe here two patients who developed this condition following bilateral insult to different sites in the centromedial core of the brain, the first following the cerebral venous thrombosis and the second after the right ACA and MCA infarct. Both these patients improved following treatment with Bromocriptine. These cases are described for proper identification and management by the clinicians. ©

INTRODUCTION
Abulia is commonly defined as loss or impairment in the ability to perform voluntary actions, show initiative or make decisions with decrease in movement, speech, thought and emotional reactions. It is the predominant behavioral disturbance with bilateral lesions of the basal ganglia, frontal lobes and cingulate gyrus. Historical analysis suggests that disorders such as abulia and impulsiveness were captured as early as the 19th century by psychiatrists in the clinical category of disorder of the will. Even today clinicians (neurologists, speech and therapists, psychologists psychiatrists) often underdiagnose this condition Abulia or fail to differentially diagnose it from post-stroke depression. We attempt to highlight the importance of identification of Abulia in the following twin case studies.

CASE REPORT
The first case VG, a 35 year old right handed lady presented with history of altered consciousness fifteen days prior to hospitalisation which lasted for three days. On admission, she was alert lying in awkward uncomfortable position with vacant facial expressions and flattened affect. Her eye movements were normal but would continue to look away from the examiner. Her cranial nerve examination, power, tone, reflexes as well as plantars were normal. Symptoms such as paucity in speech, difficulty in swallowing, incontinence and loss of appetite persisted for over a month thereafter.

On baseline assessment by a Speech-Language pathologist one month post stroke, patient was conscious and well-oriented to time, place and person. We used a language assessment which included subsections to test verbal skills such as naming, repetition, responses to routine what, where, when..., semantic and reading skills, writing abilities as well as oro-motor skills. Patient responded to all sections of the test appropriately, meaningfully and intelligibly. However, she required repeated and coerced questioning and her verbal output was limited to single words and simple phrases. Also her voice lacked normal volume and animation. Her reading and writing skills in her first language were also intact though slow to elicit. Aphasia and Dysarthria were thus ruled out based on the results of language assessment. Her relatively intact language observable through the written modality in the face of her limited verbal output highlighted the discrepancy between patients potential and performance. An objective measure of slowness was obtained by timing the performance through sequentially eliciting the numbers in forward and reverse order. Our patient required several prompts to complete the test. On oral-peripheral mechanism examination, no abnormality was detected. Gag reflex was observable, swallowing was normal. Accurate imitation of oro-motor movements was possible. Apraxia was thus ruled out. Hence patient was counseled and coerced to start oral feeds gradually. However she was not so inclined and would not eat voluntarily. Interestingly, “Miller Fisher’s” observation of telephone effect was positive and she would talk at length fluently and animatedly on phone. There was no history of emotional outbursts and she denied worry, tension, nervousness or depression. In the absence of aphasia, dysarthria, apraxia and significant depression, patients limited verbal output and lack of other spontaneous, goal directed behaviours such as feeding self, and indicating toilet needs led to the clinical diagnosis of Abulia which correlated well
Fig. 1: MR Venography TOF showing Hypoplastic Right Transverse Sinus

Fig. 2: Sagittal MIP Reconstruction Image Venography showing Interrupted Flow Signals in the Straight Sinus

Fig. 3: Mid Sagittal T2W Images

Fig. 4: Left Para Sagittal T2W Images showing Abnormal Bright Signals in the Basal Ganglia

with MRI findings of Brain suggestive of deep cerebral veins thrombosis causing venous infarct in bilateral thalami, left caudate and left lentiform nuclei. Patient was started on bromocriptine. Within a period of ten days, patient improved. She was able to imitate some speech limited to her needs, she started responding to others in complete sentences, eating a full diet orally, participating in conversations and jokes and was discharged from the hospital. At two months post stroke, patient had recovered well. However she could not resume complete household responsibilities. She showed mild memory deficits and goal directed behavior was limited to smaller tasks. Larger tasks involving several steps were daunting e.g. cooking an elaborate meal required several external prompts. This memory disturbance can be attributed to thalamic involvement.

The second, Case JB, a 48 year old female was a poorly controlled diabetic admitted with severe giddiness and blurring of vision. On examination, she would respond by starting blankly at people seeming indifferent. She did not indicate basic needs nor react to caregivers. Detailed language testing was not possible due to lack of patient co-operation. She had left hemiparesis with weakness greater in the upper limb than the lower limb. MR-angiogram of the Brain showed deep non-hemorrhagic infarcts in right anterior cerebral artery and right middle cerebral artery territory in the gangliocapsular and right frontal regions with mild effacement of the frontal horn of the right lateral ventricle.
Patient continued to be non-verbal when discharged from hospital one and a half month post stroke. Two months post stroke on follow-up to the hospital patient was able to support a verbal conversation comfortably though not initiate it. Family members reported that she started speaking incidentally when angry with them one day and gradually over the next few days verbal output increased considerably. Thus both patients had different clinico-anatomical correlation.4

Patient was started on bromocriptine. Within a week patient started recovery, became alert and oriented, recognising family, responding to simple verbal commands, rejecting treatment in the form of injections or IV thus responding to pain. A language assessment was conducted which demonstrated her intact language through reading and writing skills and her general lack of volition to cooperate for speech-language therapy and physiotherapy despite potential, drew us to the conclusion of Abulia. Her site of lesion had a strong...
symptoms, different lesions, different types of stroke and different recovery period, yet the underlying pathology of will or abulia drew a common substrate between them.

**DISCUSSION**

The term abulia refers to the psychomotor retardation that characterise the incomplete or partial forms of akinetic mutism and is caused because of bilateral lesion of the centromedial core of the brain from anteromedial frontal lobes down to the upper brain stem. Fisher in 1984 analysed 37 cases which comprised 10 cases of cingulate gyrus infarction bilaterally, 10 cases of mesodiencephalic infarction, 8 cases of hydrocephalus, 3 cases of third-ventricle cyst and 1 case of post-operative craniopharyngioma. It was cyst and one case of post-operative craniopharyngioma. It was hypothesised of hydrocephalus causing abulia through pressure on the frontal periventricular white matter bilaterally. This patient improved on removal of CSF and reduction of the intracranial pressure. Similarly, Cairns et al studied a case of third ventricle cyst causing lateral pressure on the medial thalami and thus producing the disorder. Miller Fisher has observed the involvement of the centromedial brain in his study of wide variety of pathological processes leading to the various shades of akinetic mutism.

Namekawa M et al have discussed of a case of Abulia without memory disturbance due to infarction of bilateral capsular genua without involvement of the inferior thalamic peduncles with SPECT showing remarkable hypoperfusion in the bilateral frontal cortex. This 68 year old right handed gentleman had presented with acute onset somnolence without hemiparesis, dysarthria or sensory disturbance. He gradually became wakeful but continued to be abulic. It was hypothesised that it was the disconnection at the thalamofrontal projection at the genua of internal capsule which caused the somnolence, apathy and abulia. Lesions involving both the inferior thalamic peduncle and nearby mamillothalamic tracts are known to cause the memory disturbance. This patient was spared of the inferior thalamic peduncle and hence no memory disorder.

Another patient described by Samson et al demonstrates the importance of the prompt diagnosis of the condition and proper treatment. This 62 year lady with bilateral infarcts of the caudate nuclei was unsuccessfully treated for depression for two years before the realisation of the condition being Abulia and treated with bromocriptine with daily dose of 10 mg.

Based on the discovery in animals of dopamine terminals in the neo cortical areas, Lindvall et al in 1974 identified the dopaminergic fibers arising from the substantia nigra, ascending in the medial fore brain bundle and reaching the frontal and anterior cingulate regions through the septal area. However the details of mesencephalic dopaminergic pathways in humans have not been clearly elucidated the medial fore brain bundle not been pin pointed. Experimental lesions produced in this pathway have been reversed by dopaminergic agents and in humans, abulia patients have improved on bromocriptine or if combined with Ephedrine.

Abulia is also known to be caused by closed head injury, frontal lobe disease, frontal lobotomy, tumour of the corpus callosum, brain tumour involving the walls of third ventricles and the thalamus and many other conditions. Different rates and types are summarised in Table 1.

**Table 1**

<table>
<thead>
<tr>
<th>Condition</th>
<th>Abulia</th>
<th>Aphasia</th>
<th>Post stroke depression</th>
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<tr>
<td>1. Site of Lesion</td>
<td>Bilaterally centromedial core of the brain (reduced dopaminergic activity)</td>
<td>Dominant frontal, parietal and temporal lobe.</td>
<td>Bilateral functional lesions in the antero frontal and temporal lobes and caudate nuclei.</td>
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<td>2. Nature of disorder</td>
<td>Disorder of behavior and initiative</td>
<td>Disorder of language</td>
<td>Disorder of mood</td>
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<td>3. Clinical picture</td>
<td>Looks away, stares emptily, appears to be in a daze, Miller fishers telephone effect is present.</td>
<td>Smiles at others and attempts to communicate.</td>
<td>Patient appears persistently sad, anxious, often tearful and conveys feelings of hopelessness and pessimism.</td>
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<td>4. Patients performance</td>
<td>Is PARADOXICAL - may suddenly express topic of concern in complete grammatical language animately and then revert back to quiet state and surprising everyone for few seconds. However, overall does not perform despite potential</td>
<td>Performance improves with stimulation, worsens with factors such as fatigue and noise. Patient improvement is gradual and usually performs to his utmost potential.</td>
<td>Patient usually responds with latency and all movements are slow and halfhearted.</td>
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<td>5. Patient co-operation</td>
<td>Patient refuses to Cooperate for formal testing despite potential. Patient would usually turn away from the clinician ignoring him completely.</td>
<td>Patient usually co-operates well and attempts all forms of objective testing to the best of his potential.</td>
<td>Patients co-operation may vary with degree of depression, current mood and clinicians choice of activity. Patient appears disinterested and unable to concentrate.</td>
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<td>6. Indication of needs</td>
<td>Doesn’t indicate even basic needs such as hunger, toilet and doesn’t even respond to pain.</td>
<td>Indicates basic and other needs with use of whichever modality possible, socializes appropriately.</td>
<td>Indicates basic and personal needs depending on mood.</td>
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lesions demonstrating the fact that it is the structural lesion of the described pathways which is responsible for the clinical syndrome.

Abulia is also known in bilateral destruction of the anterior cingulate gyrus through infarction in anterior cerebral artery territory. Bilateral mesodienccephalon infarct occurs from embolism in subthalamic-thalamic penetrating artery of Percheron damaging the medial thalamus and upper part of mesencephalon. These patients are initially drowsy or stuporous with fluctuation in responses and later develop abulia.

Abulia is the relatively uncommon yet debilitating lack of spontaneous, goal-directed behaviour that is seen predominantly with lesions of the basal ganglia, frontal lobes and cingulate gyrus. Difficulty in initiating and sustaining spontaneous movements and reduction in emotional responsiveness, spontaneous speech & social interaction are identified as being characteristic of Abulia. Often expressed as inability to make decisions or set goals, this condition is commonly confused with post-stroke depression, aphasia and other neuropsychiatric conditions such as schizophrenia, dementia. All of these clinical conditions have specific lines of treatment hence it is imperative that abulia be identified clinically - by correlation with site of lesion radiologically on MRI, neuropsychological evaluation and speech appraisal. This differential diagnosis at hospital admission will ensure faster and complete patient recovery with better understanding of patient behavior and thus improved family support. (Table 1).

In conclusion, brain injured patients with site of lesion in basal ganglia, thalamus, frontal lobes, cingulate gyrus should alert the clinician to differentially diagnose abulia. Treatment could include bromocriptine trials and their efficacy needs to be researched over larger population. Clinicians need to research and develop a working classification for disorders of diminished drive and motivation and instruments for clinical assessment and decision making.

REFERENCES