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Akinetic mutism and the story of David

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ABSTRACT

Following a description about the characteristics of akinetic mutism (AM) and how it differs from locked-in syndrome (LIS) and a disorder of consciousness (DOC), we present the case of David, a 71-year-old man with AM. David sustained a stroke following a middle cerebral artery (MCA) thrombus. A CT scan at the time detected old ischaemic infarcts affecting the right frontal lobe, the left occipital lobe and the left parietal lobe so David had bilateral brain damage. Initially thought to have a DOC, further neuropsychological assessments administered when David had improved a little, resulted in the diagnosis of AM. Although David spoke little, when he did speak, his words and phrases were well articulated, grammatical and with appropriate intonation. He was alert and visually aware and he was not paralysed. We discuss whether the diagnosis was correct and address the difficulties in assessing such patients.

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KEYWORDS Akinetic mutism; differential diagnosis; neuropsychological assessments

Introduction

Akinetic mutism (AM) is a rare syndrome characterised by reduced motor function including facial expressions, gestures and speech output but with some degree of alertness following damage to the frontal lobe and/or frontal subcortical circuits (Nagaratnam, Nagaratnam, Ng, & Diu, 2004). It is sometimes seen in patients who are recovering from a disorder of consciousness (DOC; Formisano et al., 2011). There are many causes of AM including infarction with frontal lobe or bilateral thalamus damage, hydrocephalus, anoxia, an anterior communicating artery aneurysm, traumatic brain injury, stroke, carbon monoxide poisoning or tumour (Herklots, Oldenbeuving, Beute, Roks, & Schoonman, 2016). The first documented case of AM was reported by Cairns, Oldfield, Pennybacker, and Whitteridge (1941). They described a 14-year-old girl who suffered from a cystic tumour of the third ventricle causing recurrent episodes of mutism and immobility. The most profound characteristics were:

Mutism, loss of feeling tone, loss of emotional expression, of spontaneous and of most other voluntary movements, and total incontinence of urine and faeces ... she was incapable of originating

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active manifestations of any kind, with the notable exception that ocular fixation and movement occurred in response to the movement of external objects and to sounds (Cairns et al., 1941, p. 279).

Since then, other cases of AM have been reported in the literature depicting a variety of pathologies and presentations.

It is common for AM to occur following bilateral lesions. Nagaratnam et al. (2004) reports on eight patients with AM following stroke and the accompanying anatomical sites affected. Their appraisal highlights the main areas associated with AM, namely the frontal lobes, basal ganglia and mesencephalothalamic regions. This concurs with the categorisation of AM into two forms: the "frontal" or "hyperpathic" form and the "mesencephalic" or "somnolent" form (Segarra, 1970). These categorisations are based on the anatomical location of the damage and clinical presentation, in particular the degree of alertness and presence of oculomotor or pupillary paralysis displayed by the individual. The frontal form results from bilateral cortical damage of the anterior cingulate, medial frontal cortex or orbitobasal cortex accompanied by an intact alertness, the presence of visual tracking of external stimuli and the fact that affected individuals can be easily roused when asleep. The mesencephalic form results from lesions of the mesencephalic region accompanied by a lower level of alertness whereby the individuals will open their eyes at times and keep them open to continued stimulation but will soon close them, returning to a state of lethargic inertia. Individuals may also display oculomotor paralysis, a lack of visual tracking or an absence of pupillary light reflexes (Nagaratnam, McNeil, & Gilhotra, 1999; Otto et al., 1998; Schiff & Plum, 2000; Segarra, 1970).

Due to the latter criteria of mesencephalic form and the clinical presentation similar to other disorders of consciousness and motivation, there is ambiguity as to how these presentations should be diagnosed. Ackermann and Ziegler (1995) suggest that the mesencephalic form does not follow the pure classification of AM and that the terms "apallic syndrome" or "vegetative state" may be more appropriate in these cases. Furthermore, there are discrepancies in the literature about the nature of AM; it has been described as a transitory state between coma and locked-in syndrome, between coma and the apallic syndrome, a separate disorder of consciousness, a subtype of minimally conscious state, a disorder of motivation or a combination of these (Kotchoubey, Schneck, Lang, & Birbaumer, 2003; Otto et al., 1998; Vachalova, Gindl, & Heckmann, 2014). This ambiguity has contributed towards the release of a paper by the American Congress of Rehabiliation Medicine (1995) to clarify the use of nomenclature when describing disorders of consciousness. They state that AM can be considered a subcategory of minimally responsive state because meaningful responses can be elicited following sensory or pharmacological stimulation, but the underlying impairment involves a severely diminished neurological drive or intention. Therefore, AM can be described as being on a continuum of disorders of motivation. Apathy is the least severe disorder on the scale (diminished motivation in the presence of intact consciousness, attention, cognitive capacity and mood), followed by abulia and then AM as the most severe disorder of motivation (almost total absence of spontaneous behaviour and speech with preserved visual tracking) (American Congress of Rehabilitation Medicine, 1995; Marin & Wilkosz, 2005).

To clarify the neurobehavioural criteria required for the diagnosis of AM, there is eye opening, which is well maintained and occurs with spontaneous visual tracking; little to no spontaneous speech or movement discernible; command following and verbalisations can be elicited but occur infrequently; and reduced movements and speech which cannot be attributed to neuromuscular disturbance or an arousal disorder (American Congress of Rehabilitation Medicine, 1995). The ambiguity aforementioned about the varying perceptions of AM as a disorder of consciousness and/or motivation alongside the differing clinical characteristics and rarity of the disorder further contribute towards errors and differential diagnosis. Rodriguez-Bailon, Trivino-Mosquera, Ruiz-Perez, and Arnedo-Montoro (2012) proposed a protocol for neuropsychological assessment and differential diagnosis of AM which involves three dimensions of assessment: attention (including orientation and alertness); language; and praxia. They describe the use of the assessment and apply it to a patient with AM.

It has been reported that some patients with a diagnosis of AM or abulia display the "telephone effect" whereby increased speech occurs whilst on the phone (Giacino, Fins, Laureys, & Schiff, 2014; Johnson-Greene, 1999). Fisher (1983) suggested that the occurrence of this phenomenon may be due to a combination of the sensory cues and overlearned behaviour of speaking on the phone, which causes a temporary reversal of the decrease in drive that arises from marked down regulation of the "mensencephalofrontal activating system" as with AM. The presence of the telephone effect can also help prevent erroneous diagnoses as it is not evident with minimally conscious patients, those with locked-in syndrome (LIS) or those with the diagnosis of depression. Paradoxical performance can also contribute towards the diagnosis as it has been reported that AM patients may inconsistently verbalise complex or correct ideas for a few seconds or minutes (Herklots et al., 2016).

Furthermore, the difficulty and uncertainty surrounding diagnosis of AM, particularly when confused with aphasias or other disorders of consciousness or mood, naturally leads to differences in investigated and applied interventions (Rodriguez-Bailon et al., 2012). In Cairns et al.'s (1941) account, puncturing the cyst allowed for the patient to recover from AM. Since then, other pharmacological treatments have been researched for more varied cases of AM. The dopaminergic system has received the most attention from the literature when investigating treatment for cases of abulia and AM because in many cases, the lesions responsible are in the mesencephalic-frontal activating system, which is a dopaminergic pathway (Nagaratanam, 2005). Parkinson's disease is caused by a loss of dopamine secreting neurones and is associated with AM (Formisano et al., 2011; Formisano & Zasler, 2014) It has been found that restoration of the dopaminergic function can lead to the amelioration of the clinical features of AM, particularly when the development of the disorder follows damage to the anterior cerebral artery (D'Souza, Kakoullis, Hegde, & Tadros, 2010; Yang et al., 2007). Therefore, it is natural that pharmacological treatments have focused on drugs that stimulate and can restore dopaminergic function: dopamine agonists, stimulants, activating antidepressants as well as other psychotropics such as modafinil and donepezil have been found to be effective in the treatment of disorders of diminished motivation including abulia and AM (D'Souza et al., 2010; Herklots et al., 2016; Marin & Wilkosz, 2005). Dopamine agonists are the most widely used intervention for AM but Devinsky, Morrell, and Vogt (1995) conclude that they can only improve AM when there are lesions of the ascending dopaminergic fibres and the limbic regions are intact, but not when the lesions include the areas where these fibres terminate. While amantadine has been shown to improve functioning in patients with a DOC (Giacino et al., 2012) and L-dopa can improve parkinsonism in vegetative patients with parkinson features (Jellinger, 2004; Matsuda, Matsumura, Komatsu, Yanaka, & Nose, 2003), it should be noted that there is not one treatment, pharmacological or non-pharmacological, that is approved for AM due to the diverse causes, lesions and presentations of the disorder (Nagaratanam, 2005).

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The present study focuses on a man who has been diagnosed with AM following a middle cerebral artery (MCA) thrombus. The assessment, diagnosis and presentation are discussed. Dopiminergic drugs have never been prescribed for him because he has only recently been described by us as having AM and the current medical team do not want to prescribe further medication until his high level of anticonvulsant medication has been reduced.

Case report

David (we are using David's real name with permission of his wife) is a 71-year-old man who gualified and spent most of his working life as an electrician. He is married with two children and five grandchildren. On 6 June 2013, he presented to his local hospital with sudden onset facial droop and left-sided weakness. His Glasgow Coma Scale score (GCS) at the time was 14/15. A CT scan showed a right MCA thrombus with old ischaemic infarcts affecting the right frontal lobe, left occipital lobe and the left parietal lobe. Four days later his GCS dropped to 7/15. David was diagnosed with a malignant infarct and sent to another hospital where he subsequently underwent emergency decompressive craniotomy due to herniation of the brain. He was transferred to the intensive care unit. Complications while David was in intensive care included resistant hypertension and aspiration pneumonia, which was treated with medication and ventilation support. He was extubated and returned to his local hospital on 27 June 2013. In November 2013 a cranioplasty was performed followed by complications requiring multiple procedures and revisions of the cranioplasty. The last operation was performed in March 2014. Sometimes cranioplasty can improve cognitive functioning; in the words of Schiffer, Gur, Nisim, and Pollak (1997, p. 231):

It is generally believed that the reason for repair of the skull defect is cosmetic or protective. There is evidence, however, that in selected cases neurologic deterioration can be reversed by cranioplasty. In the sinking scalp flap syndrome the deterioration has been thought to be related to the concavity of the skin flap and underlying brain tissue secondary to atmospheric pressure and also to the in-and-out displacement of the brain through the skull defect.

In July 2014, David was transferred to a specialist hospital for people with severe brain injury before being admitted to the Raphael Medical Centre in December 2014 for further rehabilitation including physiotherapy, occupational therapy, speech and language therapy, music therapy and neuropsychology A follow up CT scan in October 2014 showed an extensive infarction of the right cerebral hemisphere involving the entire carotid territory with enlargement of the underlying lateral ventricle and more limited ischaemic changes on the left. There was diffuse brain atrophy and ventriclar enlargement. The two CT scans were compared showing the ventricles were larger on the second scan. The cerebral sulci and basal cisterns were wide in both scans probably due to brain atrophy.

Assessment

Soon after admission, David was referred for a neuropsychological assessment. He was seen on four occasions and in different places: in bed, in his wheelchair twice, and in physiotherapy. This was to maximise the chances of eliciting a good response. However, David appeared to be asleep for most of the time, or it could have been

that he was blocking out the assessor. He was assessed with the Wessex Head Injury Matrix (WHIM: Shiel, Wilson, McLellan, Horn, & Watson, 2000). This is a behavioural observation tool consisting of 62 items in a roughly hierarchical order from least to most difficult. It provides a sequential framework of behaviour covering a patient's level of responsiveness and interaction with the environment. Behaviours observed and noted by the WHIM may occur either spontaneously or in response to stimulation. The WHIM can monitor small changes from coma through to emergence from post-traumatic amnesia in patients with traumatic brain injury (Shiel et al., 2000) but has also been used with stroke patients (Majerus, De Linden, & Shiel, 2000). David's highest score on the WHIM was 46 (brief phrases). He said "Yes" twice, appropriately. No other verbal response was seen at that time. The WHIM scores were very variable and it was not clear if David was in the minimally conscious state or was emerging from it. There was a suspicion that he was refusing to engage with the assessment.

David was reassessed in October 2015, over two years since his last stroke. He was seen three times and it was felt that he was more alert and easier to engage. He did not fall asleep, he smiled on each occasion and on 29 October he spoke several times in complete sentences. It was at this time that a diagnosis of akinetic mutism was made because, despite the fact that he could speak in fully formed and clearly understandable sentences, he rarely did so. There was no indication of a language disorder. For example, he once said, "That's all I keep hearing, Mr T" and, when a colleague came into the room and said, "Hello", he replied "I can't do it, I can't say hello", thus showing some awareness of his problem.

Meanwhile David was assessed in speech and language therapy, initially, in December 2014 and January 2015. Formal assessments proved challenging because of his reduced alertness and responsivity. He was also medically unstable. Dysphagia and low level communication assessments specially devised by the speech and language therapists were administered. An assessment of his swallowing showed he had a severe oropharyngeal dysphagia so was to remain nil by mouth and to receive all nutrition and hydration via a percutaneous endoscopic gastrostomy (PEG) tube. This was necessary because David had episodes of desaturation (low oxygen levels), sudden coughing fits, and vomiting; he was also being treated for aspiration pneumonia. All these conditions affected his lung capacity. Assessment of comprehension suggested that David could open his eyes, turn his head towards the therapist and maintain eye contact when addressed but this could mean he was responding to auditory stimuli rather than to the meaning of the request. Some members of staff reported hearing David answer questions using two to five word phrases or sentences. His typical response to questions was silence although he occasionally used full sentences. He did not respond to the reading and writing tasks of the communication assessment. The speech and language therapists recommended that David's swallowing and communication abilities should be reassessed when he was medically stable and more alert.

David was reassessed in speech and language therapy in August 2015. His oxygen saturation levels were stabilised, the episodes of vomiting had ceased and he was no longer suffering from aspiration pneumonia. Thus, there was nothing obvious to prevent him from swallowing. He received a detailed swallowing assessment, which indicated moderate dysphagia. A gradual process of re-introducing food by mouth began. By November 2015, David was able to tolerate up to 250 ml of normal fluids and three-quarters to one small plate of pureed food per day. David demonstrated

the ability to self-feed using a glass and a fork or spoon, with medium support of hand over hand assistance. During this time, David's communicative abilities had ranged from a head nod and shake for yes/no responses to providing step-by-step instructions on how to carry out wiring to his son. We were told that David could provide these instructions accurately and appropriately. We did not, however, observe this behaviour ourselves.

Unlike some patients with AM, David does not show hypomimia (reduced facial expression), sialorrhoea (drooling), seborrhoea of the face or naso-glabellar rigidity (permanent wrinkles between the nose and the eyebrows, i.e., when alert he is able to smile and raise his eyebrows appropriately, he manages his own secretions and does not drool, his facial skin is clear and he does not have wrinkles.

By February 2016, with improved alertness, engagement and medical stability David was able to tolerate food and drink of increased quantities and tougher consistencies. As far as communication skills went, he showed he was able to understand humour and sarcasm; he could smile and his intonation was appropriate. He still spoke very little and his reading and writing skills could not be formally assessed. As a West Ham football supporter, he sometimes sang the West Ham song ("I'm forever blowing bubbles") and at other times would complete the last word of each line when somebody else sang the song. Other words heard were "Hello" and "Hiya". Also "My name's David", "I play bat at cricket" and "a crow pecked me while I was singing". David was beginning to engage better during sessions particularly when the topic was of interest to him or was humorous. He showed preference for familiar people, talking more and being more responsive around such individuals. On occasion, he was able accurately to tell staff where he grew up and knew names of places he had worked as well as stating that "something's happened to my brain".

In July 2016, when seen by the neuropsychology department, he was very cross and kept banging his arm on the wheelchair. On this occasion, he spoke more than ever before. He said, "I'm a poor old soul who can't get better". Later he said, "Someone turned the thermostat down" and "I got my nose running" (his nose was not running). A few minutes after this he said, "Punch the glass out". When asked why, he responded, "Why wouldn't you do that?" and finally "I'm running out of power". This was a very hot day and he may have been feeling the heat.

These assessments suggest that David does not have a language disorder and *can* speak but rarely does so. He would also appear to have some understanding of his difficulties.

Discussion

We describe David, a man with the rare condition akinetic mutism. Patients with this condition lack most motor functions such as speech, facial expressions, and gestures, but demonstrate apparent alertness (Otto et al., 1998).

Perhaps the first question to address about David is whether the diagnosis of AM was correct. Evidence for this comes from the criteria laid down by the American Congress of Rehabilitation Medicine (1995). This states that there must be well-maintained eye opening with spontaneous visual tracking; little or no spontaneous speech; command following and verbalisations can be elicited but occur infrequently; and the reduced movements and speech cannot be attributed to neuromuscular disturbance or an arousal disorder. David exhibits all of these characteristics.

According to Segarra (1970), there are two kinds of AM, the frontal form and the "mesencephalic" or "somnolent" form. It is clear that David does not have the mesencephalic form: although he may have had a DOC when first assessed in 2014, he certainly did not have this at the time of his neuropsychological reassessment in October 2015. By then he was visually alert and looking at people. We know from his CT scan that he had damage to the right frontal lobe, the left occipital lobe and the left parietal lobe and later had an emergency decompressive craniotomy when part of the right frontal bone was removed. Thus his anatomical damage fits the diagnosis of the frontal form of AM. The bilateral cortical damage, his intact alertness, the fact that he is able to visually track external stimuli, and he can be easily roused when asleep all point towards the frontal form of AM. Most other cases of AM, for example those reported by Rodriguez-Bailon et al. (2012), have left frontal damage whereas David had primarily right frontal damage (he is right handed) but he had additional brain damage.

AM is sometimes confused with LIS and with a DOC (Herklots et al., 2016). Patients with LIS are fully conscious but unable to move or speak due to paralysis of nearly all voluntary muscles except the eyes (Smith & Delargy, 2005). David definitely does not have LIS as manifested by his, albeit infrequent, speech and the fact that he is not paralysed. Nor does he have a DOC as he can sing and he has a sense of humour. Once, when one of us was singing and dancing to the West Ham football song, he laughed and when asked jokingly if his name was Veronika, he smiled broadly. We therefore remain convinced of the diagnosis of AM.

Another question arising from this study is how best to assess people with AM? David's original neuropsychological assessment was with the WHIM, a test for people with a disorder of consciousness. This was employed because David was asleep for much of the time and it was hard to know his true level of cognitive ability. Even at that time though, it was thought that the WHIM was probably underestimating his true ability. The first neuropsychological report noted "the WHIM may have underestimated his true ability as it was possible he was refusing to engage with the assessment".

Giacino et al. (2014) mentioned that some people with AM speak better on the telephone but this was not true of David. It has been hypothesised that one of the reasons for the prevalence of the telephone effect in individuals with AM is because the ringing telephone is an unpredicted event that causes a release of dopamine which activates the damaged motivational circuit and results in brief verbalisations (Yarns & Quinn, 2013). However, Fisher (1983) suggested that it was in fact a mixture of the sensory cues, such as the ringing noise or the telephone itself, and the overlearned behaviour of speaking on the phone that causes a temporary reversal in the severe decrease in drive in people with AM. In the case of David, he did not display this behaviour, but he was observed to produce other potentially over-learned behaviours such as feeding himself or singing along to familiar songs. It is possible that these behaviours could be elicited in other people with AM and therefore, should be noted during assessments.

Other tests used for people with severe disabilities require some kind of response such as "Yes" and "No", or pointing with a finger or the eyes to a stimulus. David would not do any of these consistently so assessment was mostly through observation. The speech and language therapy report stated that "low level communication assessments specially devised by the speech and language therapists were administered". These low level communication assessments are based on categories including level of awareness in relation to visual and auditory stimuli as well as proprioceptive skills. Further elements of the assessment looked for comprehension skills: yes/no, personal, numerical and problem solving questions as well as command following beginning with one key word level and up to paragraph level comprehension. Assessment of expressive language skills include automatic speech, naming of pictures/objects, providing descriptions and conversational skills. Speech intelligibility and reading and writing tasks from word to sentence level complete the low level communication assessment used with David.

It was known that he liked football so video clips of football were shown to him. This did, at times, elicit singing. On some occasions he was more vocal than at others so we were able to ascertain that, on the rare occasions he spoke, his words and phrases were well articulated, they were grammatical and his intonation was appropriate with no signs of a language disorder. What David's case illustrates, however, is difficulty finding the right procedure for measuring his cognitive abilities.

Rodriguez-Bailon et al. (2012) recommend three areas that should be assessed in order to make a differential diagnosis of AM. The areas are attention, language and praxis. David's attention was adequate as he would sometimes engage with singing, he was usually visually alert, he could track the video screen and follow people with his eyes. As for language, we have already seen that he could nod and shake his head and provide his son with the steps required to carry out an electrical wiring task. This conversation was not observed by staff, however, the speech and language therapy report did not find evidence of a language disorder. With regard to praxis, a formal assessment of apraxia was not possible because of David's reduced movements and difficulty with engaging in any tests. We were unable to measure his ability to use fine motor skills as it was difficult for him to hold or use a pen or pencil because of contractures in his hands. When asked to write or draw or copy once a pen had been placed in his hand, he just sat with the pen in his hand without moving. David has been observed to hold a fork, a cup and a glass when attempting to self-feed. He is not independent in this, however, and requires assistance. One of the characteristics of AM is reduced movements, which is the akinetic part of akinetic mutism so it is not surprising that David's movements are affected. Rodriguez-Bailon et al.'s (2012) patient also had difficulty initiating movements.

Thus David shows the characteristic of AM: he is alert, makes few movements or facial gestures, does not have a language disorder and does not have LIS or a DOC. He fits the description provided by the American Congress of Rehabilitation Medicine (1995) by presenting with almost total absence of spontaneous behaviour and speech with preserved visual tracking.

Conclusions

We describe David, a man with AM, a rare disorder in which the individual is awake and alert and does not have a language disorder but rarely speaks. This condition is one of diminished motivation in people who are conscious without impairments of attention and dysphasia and who are not severely depressed. We consider whether the diagnosis is correct, we show that David does not have one of the other disorders sometimes mistaken for AM, and we discuss the difficulties in assessing the cognitive ability of patients with AM.

Disclosure statement

No potential conflict of interest was reported by the authors.

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