Presented by Capgras Syndrome in anti-NMDA receptor encephalitis: a Neuro-Rehabilitation approach

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SUMMARY
A middle-aged gentleman with multiple sclerosis was diagnosed with anti-NMDA receptor encephalitis. He suffered from the typical sequelae including seizures, dysautonomia, anxiety, agitation, and memory impairment but unusually developed a rare delusional misidentification disorder whereby he believed that his wife and children were imposters—Capgras syndrome.

BACKGROUND
Anti-NMDA receptor encephalitis is known to present with psychiatric symptoms like agitation, hallucination, delusional misidentification disorder, and can be difficult to diagnose. It is considered a rare but possibly common autoimmune encephalitis.

INITIAL PRESENTATION
This middle-aged patient with known relapsing-remitting multiple sclerosis (diagnosed in 2010, preceding relapse of 2019) presented with seizures, headaches, and an inter-ictal facial palsy. Despite initial treatment for infectious/viral meningoencephalitis, he deteriorated and developed status epilepticus requiring Intensive Care (ICU) management.

Diagnosis with anti-NMDA receptor encephalitis (see Table 1), he was started on immunosuppressive therapies (high dose steroids, and plasma exchange). Due to the progression of his symptoms he ultimately received a second course of plasma exchange, one cycle of rituximab, intravenous immunoglobulins, further steroid therapy, and six cycles of bortezomib.

CAPGRAS PRESENTATION
After ICU discharge, the patient was noted to have worsening confusion and psychotic features (disturbing hallucinations, paranoia, low mood and frustration). The patient’s delusional misidentification disorder (Capgras syndrome) also became apparent; he described his partner as looking the same as his partner but not being ‘his’ partner. The patient felt his partner had assimilated into his family and believed that his family did not realise that he was an imposter. He believed hospital staff were not providing correctly photos, changing them to make his imposter partner look more like the partner of old. The patient felt that there was a discrepancy between the eyes that allowed him to distinguish between the true partner and impostor. At this point the patient was diagnosed with Capgras syndrome.

DISCUSSION
There are no diagnostic criteria for Capgras syndrome; it is typically described as a delusional misidentification disorder with the belief that a close relative or family member has been replaced by an identical or near-identical impostor. Capgras syndromes usually occur in elderly patients and are associated with transient ischemic attacks, transient global amnesia, and recently in acquired neurodegenerative diseases, traumatic head injury and more frequently in neurodegenerative diseases (especially Lewy body disease and Alzheimer disease) [1,2]. Capgras syndrome is a rare condition, despite the well-known phenomenon of psychiatric manifestations in autoimmune encephalitis.

A widely accepted theory was put forth by Ellis and Young, who described Capgras syndrome as a ‘mirror image’ of prosopagnosia. This suggests that two components are required in recognising familiarity in faces: facial identity recognition requiring recall of associated semantic information (inferior temporal lobe, and the limbic-mediated emotional activity (particularly the amygdala). Prosopagnosia is when there is an inability to consciously recognise the face but are still able to elicit an emotional response subconsciously – as evidenced by a strong skin conductance response despite subjects reporting that a face is not familiar. In contrast, while conscious recognition is intact in patients with Capgras, its connection to the limbic system is disrupted leading to memory mis-management, rendering the recognised face devoid of emotional association [3]. Hinton and Flamachradian further build on this theory, adding that a second lesion in the right frontal lobe can cause a change of ‘effortless’ face recognition and that this may affect the function in order to allow the left hemisphere to compensate unhealthily, thus leading to the generation of the following explanation: ‘face signals that normally tell us “I know you” when they no longer feel familiar’. This is known as the ‘the hit hypothesis’: the first hit disrupts the connection between the vertical stream processing in the temporal lobe and the limbic complex, the second affecting the right frontal cortex.

REHABILITATION
The patient was transferred to the neuro-rehabilitation unit (NPU) after an eight-month stay in the ICU and a three-week stay on a general neurology ward. The management of this patient was multi-faceted as there were several goals set as part of his stay within the rehabilitation unit. One goal was regarding agitation, another was to improve and difficulty with swallowing, and a third was memory. The patient’s memory was poor, he was disengaged, struggling to make simple decisions and initiate sequence processes.

Psychiatry
Following medication to the NRU the patient was receiving 1mg bid risperidone and while this did not sedate the patient, it did not resolve the patient’s psychotic symptoms. On increasing risperidone to 1.5mg bid the patient described his clarity as improving to ‘75%’. A subsequent increase to 2mg bid was largely well tolerated, and the patient described being able to ‘think’ more easily (i.e., having less fragmented and disjointed thoughts). Theory A/B strategy was used as a method of comparing the patient’s views to the objective reality. This allowed the patient to try rationalise through the hallucination and orientate himself accordingly. Although this was difficult due to the poor recall and episodic memory from the existing brain injury.

OCCUPATIONAL THERAPY
Due to his Capgras syndrome, the patient’s partner was advised not to act as the primary care giver and was not involved in any discussions with patient, nor any feedback with the NPU treatment team. The patient was started on 10mg escitalopram and eventually up-titrated on risperidone to 15mg. This regime was less sedating and resulted in the delusion partially improving. It did not completely resolve however, potentially reflecting the resistant nature of Capgras syndrome.

Psychology
Psychological assessment of other NRU patients, there were no partner-based goals set for this patient. Therefore, sessions regarding the patient’s brain injury and cognitive function occurred with the patient and their children, directly, led by doctors or therapists. The patient was keen for information to his loved, and thus his brain injury to be supported. By the time it was deemed safe to the patient, the patient partner deliver the information accurately. When the risperidone was weaned down and replaced with escitalopram, the patient appeared to be more stable in terms of having less frequent to reflective emotions. Theory A/B strategy was useful as a method of comparing the patient’s views to the objective reality. This allowed the patient to try rationalise through the hallucination and orientate himself accordingly. Although this was difficult due to the poor recall and episodic memory from the existing brain injury.

Speech and Language Therapy (SLT)
Usually communication work is performed with the ‘main communication partner’ which in most cases is a partner in a same-sex relationship. In this case, the patient’s partner was also an imposter, and was advised not to act as the primary care giver and was not involved in any discussions with patient, nor any feedback with the NPU treatment team.

NURSING
On a day-to-day basis the patient’s Capgras syndrome did not interfere with tolerating, personal care and medication. The nursing staff noted that the patient had very fixed views and was not keen to compromise. For example, if the patient was asked to help him with his incontinence, the patient flatly denied that this was a problem and refused to engage. Through prompting and suggestions, the nursing staff were able to eventually encourage the patient to use toilet facilities even though this was one of the main difficulties, despite delusions of uniqueness. In this case, the patient’s behaviour may have been more reflective of his change in cognition rather than Capgras syndrome.

Perspective from the patient’s partner
On discussion with the patient’s partner, she noted that towards the end of the ICU admission his behaviour towards her appeared to change and he would no longer make eye contact with her. He would ask her to leave when other family were present and even ask ‘Who are you?’. The patient’s partner subsequently tried to engage with the patient by bringing in cake and food which the patient enjoyed. Though the patient’s partner noted that physically his walking had improved and mobilised with more confidence. Emotionally the patient’s partner noted that he became more disordered and distant regarding his own urinary incontinence. He appeared more expressive, more alert and ‘more him’. His empathy appeared to improve and he demonstrated more natural and appropriate behaviour, more typical for him pre-morbidly. With more engagement from the patient, his partner also highlighted more disordered speech and activity, while being less physically affectionate (especially towards her) than before.

Overall, by the end of their NRU stay the patient was ‘warmer’ in his behaviour to his partner. Differences remained however the partner feels that she will be able to be a ‘force of good’ for the patient. The patient underwent a graded discharge with intended plans for a 24-hour self-funded package of care.

LEARNING POINTS
• From a rehabilitation perspective, when managing a patient suffering from Capgras syndrome adaptation of existing practices may be required in order to ensure the patient is not antagonised by being encouraged to engage with the patient’s delusion.
• Managing complicated patients like this involves not only pharmacological options but also psychological/psychiatric intervention and employment of non-confrontational techniques to help manage his delusional state better.

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Table 1: Diagnostic criteria for anti-NMDA receptor encephalitis

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<tr>
<th>Criteria</th>
<th>Score</th>
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<tbody>
<tr>
<td>All three criteria must be met</td>
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<tr>
<td>1. Rapid onset (within 1 month) of one of the following major groups of symptoms</td>
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<tr>
<td>a. Disturbances in consciousness or behaviour disturbance</td>
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<td>b. Seizures</td>
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<td>c. Movement disorder; akathisia; or bilateral chorea</td>
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<td>d. Autonomic dysfunction or ataxia</td>
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<tr>
<td>e. Delusional misidentification of persons, animals, or odours</td>
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<tr>
<td>2. Temporal lobe, thalamus or limbic structures</td>
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<tr>
<td>3. No other diagnosis</td>
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Image 1:2 (Left side) showing non-patients’ exclusion of p.