A Case of Pervasive Refusal Syndrome related to Covid-19 pandemic

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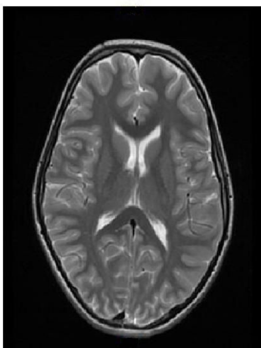
Background

Pervasive Refusal Syndrome (PRS)is a rare and potentially life threatening condition, typically affecting children aged 7-15 and girls three times as frequently as boys. [1] It is described by Nunn and Thompson[2] as a profound psychological response to uncontrollable events, and has been previously been linked to events such as grief, abuse, parental conflict and migration. [3] This is a case where the threat posed by the global covid-19 pandemic appears to have triggered this emotional response.

Although not recognised by DSM-5 or ICD-11, cases of pervasive refusal syndrome have emerged throughout medical literature. Diagnostic criteria were identified by Jaspers et al. [4] as in the table below. Patients present with extreme behavioural disturbance and resistance to all attempts at engaging children in normal daily activities, including total refusal of diet and neglect of self-care. Early identification of this disorder is essential to ensure appropriate management, which often requires a cross-discipline approach between medical and psychiatric teams.

Diagnostic Criteria for Pervasive Refusal Syndrome
A – partial or complete refusal in three or more of the following domains: 1. Eating 2. Mobilisation 3. Speech 4. Attention to personal care B - active and angry resistance to acts of help and encouragement C – social withdrawal and school refusal D – No organic condition accounts for the severity of the degree of symptoms E - No other psychiatric disorder could better account for the symptoms F – The endangered state of the patient requires hospitalisation
Core criteria are A, B and C. Additional criteria are D, E and F.
Jaspers et al, 2009



Case Report

Michael was always described as a happy, calm child, 10 years old. He enjoyed school and loved playing outdoors with his younger sister, Rose. He had been progressing well with his life and neither his parents nor his school had any concerns for him. Like all of us, following the covid-19 pandemic and school closures, Michael began washing his hands and became more conscious of daily hygiene safety advice. However, for Michael things escalated to a very difficult level. Initially, he manifested extreme levels of anxiety with heightened levels of distress. He ran away from open doors or windows for fear he would catch the virus, he insisted on changing his clothes several times per day, he would become extremely emotionally distressed if anyone touched him accidentally while he was outside on his scooter and could spend hours afterwards crying and screaming.

Things reached a crisis in June, when Michael began to show a profound refusal to engage in basic care tasks and a dramatic social withdrawal. Things deteriorated to the point where he ultimately required admission to a paediatric hospital. He refused to eat and drink, resisted attempts at washing and toileting, lay in bed with the covers over this head, became non-verbal and refused to engage with any conversation or games. He showed very prolonged periods of screaming. Medical work-ups could not identify any cause for this acute change in behaviour
Ultimately the situation deteriorated to the point of requiring TPN and PEG feeding. Michael was given a diagnosis of pervasive refusal syndrome, secondary to severe covid-19 related anxiety.

Efforts to manage Michael's distress using medication were initially unsuccessful. He was trialled on promethazine and olanzapine to manage his severe agitation, with very limited results. A trial of haloperidol caused an acute dystonic reaction, for which Michael had to be intubated. A trial of sertraline showed no benefit.

Michael's condition began to improve following the implementation of a behavioural plan in a low stimulation environment. This initially consisted of the use of specific scripted, consistent phrases that were predictable and reduced stimulation and distress in all communications towards Michael. The addition of risperidone and, later, fluoxetine led to a gradual improvement in symptoms. Michael and his family engaged in a number of rehabilitative and therapeutic interventions as an in-patient.

Michael has shown significant improvements and, happily, has now returned home to his family. He does however continue to experience anxiety, particularly in social situations, and has not yet returned to school. He remains vulnerable but has been working hard, with both the community CAMHS team and the support of his parents, to return to his previous activities.

Discussion

Pervasive Refusal Syndromehas previously been described in the context of sexual abuse, neglect, among asylum seekers and in the context of neurodevelopmental conditions such as autism.[3] In Michael's case, the 'uncontrollable event' which appears to have triggered his condition is the threat posed by covid-19 and lockdown, both generally and to his family more tangibly as his father was a front-line worker. We have not identified any other cases, to date, of PRS linked to covid-19. However, as lockdown measures continue globally there is the possibility of further emerging cases.

Through treatment in low stimulation environments, with consistent communication, rehabilitation and medication, followed by individual and family therapies when patients are more able, patients with pervasive refusal syndrome show a slow, but generally complete, recovery. A systemic review of case reports shows a mean in-patient stay of 9.69 months, (range 1 – 36 months).[5] Of the 76 patients included, 62 made a full recovery, 13 made a partial recovery and 1 patient died.

A challenge in managing children with PRS is the cross-discipline approach needed. Michael's acute management required input from paediatricians, anaesthetists, medical nursing staff, and liaison psychiatry, before stabilisation to a point where he could be transferred to an acute psychiatric unit for intensive psychiatric support. A challenge to his care was the fact that in-patient units in Ireland are designed to cater for children aged 12+, with no specialist under 12 unit available as there is in other countries. Ultimately Michael was cared for in a high dependency bed, separate from other, older adolescent patients in the psychiatric in-patient unit.

Conclusion

Michael's case highlights the importance of appropriate diagnosis and management of severe mental illness in young children. A cross-discipline approach, involving both medical and psychiatry teams, is vital both to the diagnosis and management of Pervasive Refusal Syndrome. Awareness of rare diagnoses such as this will reduce the delay to treatment. A challenge in Ireland is accessing psychiatric in-patient treatment for very young children, with specialist units in Ireland designed to better cater for young people aged 12+.

References

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Consent

Michael (not his real name) and his parents have kindly agreed to allow us to tell his story, in the hope of teaching current and future psychiatrists about this rare condition. We send them our thanks and appreciation.