SOCIAL COGNITION AND BEHAVIOUR IN NEUROPSYCHIATRIC DISORDERS

Elizabeth Finger, MD
Schulich School of Medicine, Western University
London, ON, CANADA

General Approach: Altered social cognition and behavior is a feature of several neurologic and neuropsychiatric disorders. Social behavioural changes develop as a function of the neuroanatomy and neurophysiology of the underlying disease, and may manifest differently if triggered by a focal lesion (i.e. stroke) vs. a disease affecting multiple regions or networks (i.e. frontotemporal dementia). Assessing a patient who presents which changes in social cognition and behavior requires a systematic approach and consideration of a range of disorders in the differential diagnosis as detailed below.

History: As in the approach to any neurologic complaint, changes in social cognition and behavior require ascertainment of the time course of symptoms. As patients typically do not have insight into their own behavioural changes, information from a reliable collateral source is required. Specifically, the neurologist should consider the length of time the informant has known the patient, to determine whether their perspective will be sufficient to understand the patient's baseline cognitive and social functioning, as well as sensitive to acute or gradual changes over recent years. If the informant is a spouse or partner, gauging whether marital/relationship conflict may bias the judgements of the informant and the severity and frequency of the behaviours is difficult. Determining whether the behaviours in question are also demonstrated with others, apart from the closest partner, is critical. This is particularly critical as the context and timing of clinical assessments may not demonstrate behaviours which occur in more familiar settings. A thorough developmental history including cognitive, motor and social milestones, educational and occupational attainments, criminal and relationship history will inform understanding of the patient's baseline abilities and serve as a comparison to the present concern. Once the time course of change and the main symptoms are identified, more detailed inquiry into relevant symptoms and functional levels can be guided by the use of disease specific tools to assist in diagnosis and guide management, as described below.

Exam and Ancillary Testing: In majority of patients presenting to the neurologist for assessment of changes in social cognition or behaviour change, a subacute or gradual time course is identified. Routine neurologic examination should include testing of vertical and horizontal saccades, axial tone, frontal release signs (grasp, snout reflexes) glabellar tap, and assessment of parkinsonism. Cognitive testing is also indicated to identify deficits in other cognitive domains including memory, language, executive and visuospatial function. In many disorders, patients may perform well on standard cognitive testing, but careful observation of their behavior during testing and the nature of errors (e.g. errors due to impulsivity, perseveration, or inattention) made may reveal significant abnormalities. Brain imaging, preferably MRI including susceptibility sequences, is required to rule out structural lesions such as slow growing brain tumors, chronic subdural hematomas, stroke, and to evauate patterns of atrophy. In the absence of a structural lesion, in an adult patient presenting with gradual changes in social behavior, the differential diagnosis is typically focused on neurodegenerative etiologies, psychiatric disorders, or the interaction of developmental disorders with aging, metabolic disturbances, or life stressors, as described below.

Frontotemporal Dementia: bvFTD is defined by the gradual onset and progression of changes in behavior, including disinhibition, loss of empathy, apathy, and may include hyperorality and perseverative or compulsive behaviours. (1) By the time a patient presents to a neurologist, careful history typically reveals several years of such behavioural changes. Disinhibition in FTD may manifest in a variety of ways, including increased disclosure of personal information to strangers or acquaintances, hypersexual comments or behaviours, decline in manners (shoveling in food, burping in public), rude comments to acquaintances or strangers, and impulsivity. Apathy is a common and manifests as a loss of interest in usual social and non-social activities. A subset of patients may demonstrate compulsions, such as touching items in a room, counting figures, or picking up trash in public places. Hyperorality typically involves increased consumption, particularly of sweets, and in the extreme, can include consumption of spoiled foods and inedible objects. Some patients will begin to use tobacco or alcohol for the first

time or increase their use of such substances. Signs of abnormal behaviour may be observed during the course of the exam. Patients with bvFTD may show evidence of poor grooming and hygiene, and loss of manners, such as frequently interrupting or belching during the examination. A flat affect, or conversely, a silly, child-like affect may be seen. Patients with bvFTD may be restless, even attempting to leave the room mid-examination. Patients with a flat affect may appear apathetic and lack of spontaneous speech, giving only brief one- to two word responses to questions despite preserved language abilities. Although such abnormal behaviours typically increase over the course of the disease, at early stages patient's conduct may be generally appropriate for the limited duration of the examination. Positive snout or grasp reflex may be present, although these frontal release signs are not sensitive or specific for FTD. Several symptom inventories are available which can aid in the identification and quantification of FTD related behaviours including the Frontal Behavioural Inventory (2), the Cambridge Behavioural Inventory (3), and the Neuropsychiatric Inventory(4).

FTD phenocopy: Patients who present with such behavioural changes reported by a partner or family, but who do not demonstrate objective deficits on exam or focal frontal or temporal atrophy on imaging and who do not progress have been described as having "FTD phenocopy" syndrome. Patients meeting criteria for FTD phenocopy syndrome have higher rates of psychiatric disorders including depression and bipolar disorder, substance abuse, and may include individuals with relative developmental deficits in aspects of social cognition and behavior (i.e. Autism Spectrum Disorder) that appear to increase due to aging, relationship conflict, substance abuse, or cluster C personality traits (obsessive compulsive).(5)

Antisocial Personality Disorder and Psychopathy: Individuals with antisocial personality disorder may share some behavioural traits with FTD, including impulsivity, frequent agitation/irritability, aggression, diminished or absent regard for others, financial irresponsibility and lack of remorse for harmful actions (DSM-V). Antisocial Personality Disorder can be distinguished from FTD by the developmental nature of behaviours. Patients meeting criteria for antisocial personality disorder demonstrate criminal or harmful behaviours beginning in youth (before age 15), meeting criteria for conduct disorder. Careful history from a reliable source will often reveal a long list of prior criminal charges or incarcerations, as well as substance abuse. A subset of individuals with antisocial personality disorder will also meet criteria for psychopathy and demonstrate the personality traits and behaviours of antisocial personality disorder, but also typically lack the capacity for empathy from youth. While acts of reactive aggression (such as assault or sexual offences which reflect acting out due to frustration or failure to obtain a desired goal) are seen in antisocial personality disorder and psychopathy, instrumental aggression (premeditated, calculated acts such as armed robbery) are hallmark of psychopathy. Lying to manipulate is common in antisocial personality disorder and psychopathy, while confabulation may be observed in FTD. Antisocial and psychopathic traits can be systematically assessed using the Psychopathy Checklist-Revised (Hare, R. D. Legal and Criminological Psychology, 1998) or the Psychopathic Personality Inventory (Lilinfeld Lilienfeld, S. O., & Andrews, B. P., Journal of Personality Assessment ,1996).

Acquired Sociopathy and Traumatic Brain Injury: Traumatic brain injury damaging the orbitofrontal, ventromedial prefrontal cortex, or amygdala can lead to symptoms of anti-social personality disorder in individuals who did not demonstrate antisocial behavior in youth, as in the classic case of Phineas Gage. Changes in personality and behavior following such injuries can include impulsivity, irresponsibility, hypersexual behaviours, diminished empathy, apathy, poor insight, and reactive aggression.(6) While the acute onset following a traumatic head injury and static course of symptoms can distinguish such behavioural changes due to acquired sociopathy from FTD, patients with acquired sociopathy may present with increasing behavioural problems suggestive of a progressive course, which reflect decompensation of the old injury due to aging or additional insults (i.e. frontal stroke).

Autism Spectrum Disorders: Individuals with autism spectrum disorders demonstrate deficits that can overlap with those observed in patients with FTD. Specifically, autism spectrum disorders feature deficits in verbal and nonverbal social communication, including difficulty processing facial expressions, tone or other gestural cues, difficulty with the normal give and take of conversation, reduced empathy, reduced facial expressions, poor eye contact, stereotyped, repetitive motor behaviours, and rigid, inflexible adherence to routines. In contrast to FTD, autistic spectrum disorders feature the presence of such traits and behaviours beginning in the early developmental period. Additional traits not commonly observed in FTD include early life fixation on a specific item or highly restricted narrow interests, lifelong difficulties with humor and non-literal inferences, and early deficits in imaginative play. As discussed above in the FTD phenocopy disorder, patients with mild autism spectrum disorders may present in adulthood when these symptoms or behaviours become manifest due to a change in life

circumstances (new relationship, stressors) or interactions with aging. Ascertainment of the developmental history including social functioning in childhood and early adulthood from a long time family member or friend is typically required to identify latent autism spectrum disorder as the etiology of social behavioural impairments presenting later in life. While clinically validated screening tools for diagnosis of autism spectrum disorder in adults are lacking, (7) self-report inventories show utility in research studies include the Autism Spectrum Quotient (Dr. Simon Baron-Cohen- http://www.autismresearchcentre.com/) and the Ritvo Autism and Asperger Diagnostic Scale-Revised (Dr. Susan Bejerot; http://psychcentral.com/quizzes/autism-quiz.htm).

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