Obsessional Slowness in Down Syndrome: Severe Variant of OCD or Separate Disorder?

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In a recent report by Charlot, Fox and Friedlander, a series of eleven patients with Down syndrome and slowness was presented. These individuals spent several hours each day performing routine tasks such as bathing, dressing, and eating. Slowness was considered a significant problem and interfered with normal functioning. These authors reviewed the literature and found other reports of people with “obsessional slowness.” Most of these case reports described individuals who were diagnosed with Obsessive Compulsive Disorder (OCD). Although checking rituals were observed, low levels of anxiety and perfectionistic tendencies were commonly described. Some investigators have suggested that obsessional slowness can be seen as a primary clinical condition, while others have argued that it represents a severe variant of OCD. Only two other reports were found about individuals with a developmental disability having slowness as a clinical problem. In both of these papers, individuals with Down syndrome were described.

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In a recent retrospective case series, Charlot et al.⁶ described 11 individuals with Down syndrome and obsessional slowness. All of these individuals referred for treatment of slowness spent hours each day performing routine daily functions which interfered with the completion of daily routines, including missed vocational and other programs. These individuals had some symptoms of an obsessional compulsive disorder, but the authors suggest that the slowness was not always a product of repetitive behavior. They reviewed literature regarding the phenomenon of obsessional slowness in individuals without developmental disabilities.

Obsessional slowness was first described by Rachman¹⁶ in 1974. His case series included ten patients diagnosed with Obsessive Compulsive Disorder (OCD) who spent hours performing routine daily functions such as eating, dressing, and bathing. The extreme slowness often resulted in social isolation and unemployment. However, it was not always clear that slowness was caused by ritualistic behavior. Unlike most patients with OCD, Rachman’s patients reported low levels of anxiety. Also, they were described as being meticulous and perfectionistic. The low anxiety levels, extreme slowness, and obsessive compulsive personality traits seemed to suggest that this subgroup was qualitatively different from other patients with OCD.

Not long after the Rachman paper, three more case reports of individuals with obsessional slowness symptoms were published.³⁴⁹ A debate gradually emerged over whether or not obsessional slowness should be viewed as a severe form of OCD or if it would be better characterized as a separate disorder. Of interest is the fact that the patients with obsessional slowness were not given the usual treatment for OCD (exposure and response prevention) but were treated with behavioral shaping techniques (prompting, pacing and shaping). Most responded positively to an intense daily treatment regimen, but many relapsed as soon as the behavioral therapy was faded.³⁴⁹¹⁰¹⁶ Psychopharmacologic interventions were not described.

Later reports of this unusual disorder included a more extensive and systematic investigation of 17 inpatients with OCD and significant slowness in which the investigators constructed a survey to establish criteria for identification of obsessional slowness cases.¹² Hymas and colleagues assessed and compared a neurologically impaired and a normal control group with patients identified as having obsessional slowness. The obsessional slowness group presented with rituals including checking, touching, ordering, counting, repeating, washing, and mental rehearsing. Twenty-three percent of the sample with obsessional slowness had a
Very little has been written about the subject of obsessional slowness in individuals with developmental disabilities. Including the Charlot et al. case series, only three reports were identified in a Medline search of this topic. Pary described slowness as a primary feature in a small number of his patients with Down syndrome. He also noted that his patients with slowness manifested little anxiety. Pary cited investigations of regional cerebral blood flow (rCBF) and other imaging studies of individuals with Down syndrome in which a hypometabolism in the orbital cortex is found. This would contrast with the usual findings for subjects with OCD and obsessional slowness, who typically present with a hypermetabolism in the same cerebral region. Since obsessional slowness might be a severe form of OCD, Pary speculated as to whether or not these patients might benefit from therapy with SSRIs. In the only other published report of obsessional slowness in an individual with developmental disabilities, the successful treatment of a 26-year-old woman with Down syndrome using pacing and shaping techniques to treat slowness was described. These authors reported improvement was maintained at a 15 month follow up, unlike the research in other patient groups, which showed relapse to be common after intensive therapy was faded.

A number of studies have described OCD in samples of individuals with a cognitive disability, including responses to SSRIs, but slowness is rarely addressed. Although no data are available, it has been speculated that obsessional slowness is very rare in people with developmental disabilities, but may occur more often in individuals with Down syndrome.

In the literature addressing obsessional slowness in people without developmental disabilities, there appeared to be more male cases. However, none of these studies used random sampling techniques, and varied methods were employed to identify cases of slowness. The Charlot et al. cases were not systematically selected, but there were almost equal numbers of male and female patients. All of the patients in the Charlot et al. series were referred to outpatient psychiatric clinics because their slowness interfered with routine daily functions. Slowness was described at times as resulting directly from interfering ritualistic behaviors including ordering the environment, touching, checking, and hoarding. However,
patients were observed to have periods of freezing, and apparent problems with the initiation of movements (including possibly, the initiation of speech in two cases), similar to the neuromotor problems described by Hymans and colleagues. The relationship between possible early signs of idiopathic Parkinson Disease and slowness were not explored by Charlot et al., but would be of interest in any future investigations.

Five of the patients with Down syndrome and obsessional slowness described by Charlot et al. had comorbid tics. Low levels of anxiety as well as perfectionist tendencies were described. Charlot et al. cited Rasmussen and Eisen, who suggested that OCD may occur along a continuum. On one end of the continuum, there are individuals with low levels of anxiety, hoarding and ordering rituals, tics, slowness and features of an anankastic personality. On the opposite end of the continuum, one finds individuals with high levels of anxiety, pathologic doubt or fears of contamination. Some additional support for this suggestion of phenomenologically distinct OCD subtypes comes from literature regarding treatment outcomes. Patients with tics, ordering, hoarding, and slowness appear to have less positive responses to the usual anti-OCD therapies (SSRIs and exposure and response prevention). Rasmussen and Eisen hypothesized that this subgroup of OCD patients, who seem to have a more treatment resistant form of the disorder, may suffer from a greater degree of dysfunction in dopaminergic pathways in the brain.

Six of the patients in the Charlot et al. case series had hypothyroidism, a problem which occurs at inflated rates in individuals with Down syndrome. The authors noted that their patients with hypothyroidism had hormone replacement therapy, and they did not feel that this condition accounted for the slowness problem. Six of the Charlot et al. patients had depressive episodes before the onset of compulsive symptoms and slowness. The authors argued that slowness seemed to persist even after the depression was resolved, and they did not feel that slowness could be explained on the basis of a psychomotor retardation caused by depressive illness. This is consistent with the Hymans et al. findings that slowness was not correlated with measures of depression.

In addition to depression and hypothyroidism, Charlot et al. suggested that attention deficit-hyperactivity disorder inattentive subtype, passive aggressive personality traits, and psychotic disorders presenting with catatonia should be considered when patients with Down syndrome present with significant slowness. All of the Charlot et al. patients were reported to have onset of slowness symptoms late in adolescence or early adulthood. This would suggest that disorders that typically emerge in childhood are less likely alternative diagnoses.

One further diagnostic consideration discussed by Charlot et al. was Alzheimer’s disease. Alzheimer’s disease has been reported to occur at elevated rates in individuals with Down syndrome. None of the patients in the Charlot et al. series were diagnosed with Alzheimer’s disease. However, in future investigations, patients with Down syndrome and obsessional slowness could be followed longitudinally to determine if any of these individuals develop dementia, and if so, at what rates. A more comprehensive investigation would follow individuals with Down syndrome, and examine who develops depression, OCD, slowness or dementia and to what extent these conditions overlap.

There were anecdotal reports of partial response to treatment with SSRIs for a majority of the patients in the Charlot et al. series. The authors speculated as to whether or not neuroleptic augmentation might be helpful for patients with comorbid autistic disorder or tics. Other augmentation strategies suggested included the addition of clomipramine, beta blockers (i.e., pindolol), or dopamine agonists (i.e., Dextedrine) which might be considered in cases responding poorly to other therapies. Behavioral treatment strategies have been described as beneficial to patients with obsessional slowness. Shaping and pacing were reportedly of some benefit to the individuals described in the Charlot et al. report. It was further suggested by these authors that educating caregivers was a useful and important intervention. Some of the patients had been seen by caregivers as “stubborn” or “lazy.” Individuals with Down syndrome have often been described as having a temperament characterized by “stubbornness.” This tendency might even be considered as a possible risk factor for slowness.
To establish the validity of a separate obsessional slowness syndrome, it would first have to be demonstrated that this disorder can reliably be differentiated from other disorders, including OCD. The type of structured approach used by Hymas et al. would be helpful, one in which clinically significant slowness is operationally defined in clear terms using a technique that could be replicated. A road block to systematic investigation of the problem in individuals with and without developmental disabilities is the fact that obsessional slowness appears to be relatively rare. However, prospective controlled investigations would be most helpful to improve our understanding of this unusual clinical phenomenon.

REFERENCES


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