Dementia With Lewy Bodies and the Neurobehavioral Decline of Mervyn Peake

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Mervyn Peake (1911-1968) was an accomplished British artist, poet, novelist, and playwright. He was a prolific and talented illustrator and wrote hundreds of poems, 4 novels, and several plays. His exceptional career was prematurely ended by a neurodegenerative illness variously ascribed to Alzheimer disease, Parkinson disease, or postencephalitic parkinsonism. However, a detailed review of biographical accounts produces substantial evidence in support of a probable diagnosis of dementia with Lewy bodies, a clinical entity remaining undiagnosed outside specialty dementia clinics. Peake developed signs of parkinsonism and insidious cognitive decline during his fifth decade. A breakdown in his writing style has frequently been cited as reflecting his encroaching dementia. Visual hallucinations are portrayed in sketches, and together with paranoid delusions are apparent in poetry composed during his illness. His deterioration was progressive and punctuated by well-described episodes of confusion and psychosis. His occasional preservation of insight is poignantly captured in drawings of figures with dunce caps or pointed heads, often with expressions of fear and apprehension etched with an economy of strokes. Peake spent his final years in various psychiatric institutions but continued to exhibit lucid intervals even late into his illness. His tragic deterioration remained undiagnosed at the time, but in retrospect, his progressive dementia with parkinsonism, visual hallucinations, and marked cognitive fluctuations likely represents one of the earliest recognized historical cases of dementia with Lewy bodies.

Dementia with Lewy bodies (DLB) is estimated to account for 15% to 20% of cases of dementia, based on autopsy data1 and epidemiologic studies.2 Wider recognition of this disorder has emerged only recently, coinciding with descriptions published since the 1980s.3 To date, reports of prominent historical cases are lacking, although with increasing awareness, such cases are more likely to be recognized in retrospect. A probable diagnosis is apparent in the neurobehavioral deterioration of accomplished British artist, poet, novelist, and playwright, Mervyn Peake (1911-1968), whose celebrated Gormenghast novels were recently adapted into a critically acclaimed television miniseries by the British Broadcasting Corporation.4 Peake's persistent attempts at artistic expression in the face of a relentless, cognitively devastating illness provide us with a unique and intensely personal account of a historical case of DLB.

Mervyn Peake was born in Kuling (Lushan), China, in 1911. His father, a British missionary physician, brought his family back to England in 1922. As a young man, Peake entered art school and first exhibited his work in 1931. He soon established a reputation as a talented illustrator. He was called up for military service in 1940, and while serving in the army, started work on his Gormenghast novels. After the war, he was commissioned to document the aftermath of the conflict and completed a series of heartbreaking sketches of concentration camp survivors at Belsen. He continued to write and illustrate, producing many of his finest paintings, drawings, and poems during the late 1940s (Figure 1). He was much admired as an art instructor, and during this period also scripted several plays.3

Peake first showed signs of a mysterious neurodegenerative illness around 1956, developing signs of parkinsonism and symptoms of cognitive decline in his fifth
decade. Perplexed biographers have alternately speculated diagnoses of Alzheimer disease, Parkinson disease, or postencephalitic parkinsonism after questioning a history of viral encephalitis or encephalitis lethargica. The disintegration of Peake's writing style has frequently been regarded as reflective of his encroaching dementia. He also suffered from bouts of depression, which influenced his later work. After a stressful period that culminated with a number of disastrous play reviews, his behavior started to become erratic, and his wife reported that he experienced hallucinations. His deterioration was progressive, punctuated by well-described episodes of confusion and psychosis but relieved by intermittent lucid periods. He was ultimately diagnosed as having parkinsonism and "premature senility."

In 1960, neurosurgical approaches for Parkinson disease were being attempted, and Peake underwent one of these procedures in an effort to ameliorate his parkinsonian symptoms. The effects were disappointing. As a last resort, an appointment was arranged with the preeminent British neurologist of the day, Sir Russel Brain, but he was likewise unable to assist the ailing artist. Peake spent the next several years of his life in various institutions, treated with neuroleptics and electroconvulsive therapy, until his death in 1968 at the age of 57 years. His work has since drawn substantial critical acclaim and is presently the subject of increasing public interest. His influence on subsequent generations of artists and writers has been profound.

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Consensus Criteria for a Diagnosis of Dementia With Lewy Bodies (DLB)

1. The central feature is progressive cognitive decline of sufficient magnitude to interfere with normal social or occupational function. Prominent or persistent memory impairment may not necessarily occur in the early stages but is usually evident with progression. Deficits on tests of attention and of frontal-subcortical skills and visuospatial ability may be especially prominent.
2. Two of the following core features are essential for a probable, and 1 for a possible diagnosis of DLB:
   - Fluctuating cognition with pronounced variations in attention and alertness
   - Recurrent visual hallucinations that are typically well formed and detailed
   - Spontaneous motor features of parkinsonism
3. Features supportive of the diagnosis are:
   - Repeated falls
   - Syncope or transient loss of consciousness
   - Neuroleptic sensitivity
   - Systematized delusions
   - Hallucinations in other modalities
   - Depression
   - REM sleep行为 disorder
4. A diagnosis of DLB is less likely in the presence of:
   - Stroke disease, evident as focal neurologic signs or revealed on brain imaging
   - Evidence on physical examination and investigation of any physical illness or other brain disorder sufficient to account for the clinical picture

Abbreviation: REM, rapid eye movement.

In 1912, Friederich Lewy described the presence of intracytoplasmic inclusion bodies in the dopaminergic neurons of the substantia nigra in patients with Parkinson disease. Lewy bodies in the midbrain are spherical, eosinophilic inclusions, differing slightly in appearance from those described in the neocortex. Reports of cortical Lewy bodies were rare until new staining techniques for ubiquitin were introduced in the 1970s. The occurrence of cortical Lewy bodies in cases of dementia was firmly established by the 1980s. Antibodies to α-synuclein, which immunostain Lewy bodies, have since replaced the use of the ubiquitin stain, and DLB is now classified among the spectrum of diseases regarded as synucleinopathies.

Consensus criteria for the diagnosis of DLB were published in 1996 and refined in 1999 (Table). The prospective sensitivity and specificity of the consensus criteria for DLB are reported at 0.83 and 0.95, respectively. Previous retrospective studies yielded comparable specificities (0.87-1.00) but lower sensitivities (0.22-0.89). Hence, although the criteria might not identify every individual harboring the condition, there is high certainty...
of a diagnosis of DLB if the criteria are met. Age at onset generally ranges from 50 to 83 years,15 although autopsy-verified cases have been documented in patients with initial symptoms at an age as young as 27 and 33 years.16 The mean survival time is usually comparable with that of Alzheimer disease.15

**HISTORICAL EVIDENCE**

Peake’s progressive cognitive decline is apparent in his later work. His pronounced difficulties with attention and visuospatial ability are typical of DLB, in contrast with the more prominent decline in memory function observed in Alzheimer disease.11 Commencing with his later illustrations, one biographer observed that, “there is a coarseness of feeling, a lack of sympathy for the persons he portrays.”8 (p219) By 1958, his drawings were more caricature-like and later became “geometric and then almost abstract”8 (p219) in keeping with Peake’s inexorable deterioration of visuospatial function. In this respect, his case is comparable with those of Willem de Kooning17 and William Utermohlen,18 in whom similar changes were noted in artistic ability with advancement of probable Alzheimer disease, likely arising from involvement of posterior isocortical association areas, with resultant deterioration of visuospatial function.

Although problems with memory were apparent by 1960, Peake’s “attention span had become so short that he could no longer read a story and retain the idea for an illustration long enough to draw it.”8 (p223) Thus, both cognitive domains predominantly affected in DLB were especially involved. Conversely, a tenuous preservation of insight was captured in drawings of figures in dunce caps or with pointed heads, often with expressions of fear and apprehension etched with an economy of strokes (Figure 2A), distinguishing him from artists diagnosed as having probable Alzheimer disease.18

Peake’s parkinsonian signs and symptoms have been well documented in biographies.9 In his mid 40s, he developed shaking of the hands and was tentatively diagnosed as having Parkinson disease after evaluation at the National Hospital, Queen’s Square (London, England).3 The shaking “spread to his legs as well as his hands” and exhibited “remorseless progression” over the years.8 (p213) The beneficial effects of L-dopa in the management of Parkinson disease were not discovered until the 1960s, and so treatment in the preceding decade was limited to the use of anticholinergic medications and experimental surgical pallidotomy. Later photographs of Peake reveal the stooped posture and inscrutable expression characteristic of parkinsonism.

Peake was tormented by recurring visual hallucinations, the early development of which—soon after the onset of his other symptoms—is further suggestive of DLB. Many well-formed and detailed examples are evident in his sketches (Figure 2B). These hallucinations, together with the paranoid delusions he experienced while in the hospital, were incorporated into poetry composed during his institutionalization:

Heads float about me; come and go, absorb me; Terrify me that they deny the nightmare That they should be me, defy me; And all the secrecy; the horror Of truth, of this intrinsic truth Drifting, ah God, along the corridors Of the world; hearing the metal Clang; and the rolling wheels. Heads float about me haunted By solitary sorrows

Other poems, perhaps indicating auditory as well as visual hallucinations, and letters to his wife reflecting his frightening delusions, are further discussed in a recent biography.5

By the time he entered his sixth decade, Peake “was with us only in flashes, and those flashes were often over before we had grasped what he had said, or could reply. Most of the time he sat in silence, head bowed.”20 (p37) A remarkable example of the striking fluctuation in his mental status follows:

After dinner he was sitting humped on the sofa when suddenly he made a motion toward Maeve, and as she leaned over him he indicated that he wanted some paper and something to draw with. My wife produced a sheaf of quarto typing paper and placed a ball-point pen in his swollen hand. We went on talking while he sat with the sheaf of white paper on his knee. . . . Then I noticed that his hand had ceased its normal shaking, and that he was sitting upright with the paper held firmly, concentrating. For more...
than an hour, he seemed to lose touch with his illness altogether. He covered page after page with wonderful and preposterous beasts, leaping, snarling, laughing, cavorting. As soon as one was finished he turned to a fresh page and drew another. We self-consciously kept up our conversation, although all three of us were watching him in amazement. He appeared unaware of our presence. And then the spell was broken. He dropped the pen, and his hand began to shake again.21 (p209)

This is a compelling description of a dramatic fluctuation in cognition and alertness, which is typical of DLB and not other forms of dementia, such as Alzheimer disease. There are abundant examples of similar observations in accounts of his illness.5,7,8

Several features supportive of a diagnosis of DLB are also apparent in biographical material of Peake’s neurobehavioral decline. Repeated falls were observed, as “his sense of balance would suddenly desert him, and he would lurch and fall against objects and people.”21 (p209) Moreover, there is frequent reference to neuroleptic sensitivity, since “although the doctors gave him tranquillizers, the tremors in his hands and legs increased.”8 (p218) Thorazine and chlorpromazine were the most common neuroleptics used in the 1950s and 1960s. His wife often ironically observed that the medications doctors gave him only seemed to make him worse.8 One of his physicians also noted that some of the treatments “aggravated the parkinsonism, rather than improved it.”21 Worsening confusion and exacerbation of parkinsonism are common consequences of neuroleptic use in DLB.1,2,13

Peake’s symptoms developed at a younger age than most patients with DLB, although the duration of his illness was more in keeping with the diagnosis. Throughout his institutionalization, he was victim to systematized delusions and hallucinations. His occasional bouts of depression intensified over time. In addition, rapid eye movement sleep behavior disorder may have been present, since Peake was plagued by nightmares and “a restless nervous energy that prevented him from sleeping at night.”8 (p212) However, the published material does not allow for verification of this possibility.

Previous speculation has centered on the possibility that Peake suffered from postencephalitic parkinsonism,21 but such an explanation is dubious on several counts. The pandemic of von Economo encephalitis (encephalitis lethargica, or “sleeping sickness”), which was first described in 1916 and dwindled to obscurity by 1929, was associated with a syndrome of parkinsonism observed to manifest up to 4 or 5 years after the acute illness. There is no reliable record of acute encephalitis in Peake, and by the 1940s, cases of postencephalitic parkinsonism had become exceedingly rare.22 Moreover, no instances of oculogyric crises or convulsive movements, dramatic features typical of postencephalitic parkinsonism, were ever reported to occur in Peake. Finally, progressive dementia was not a prominent part of the clinical picture in the postencephalitic syndrome, which was generally limited to chronic parkinsonian phenomena.22

**COMMENT**

Mervyn Peake’s deterioration was a mystery at the time, but in retrospect, his progressive neurobehavioral decline associated with parkinsonism, visual hallucinations, and cognitive fluctuations likely represents one of the earliest recognizable, and possibly best-documented, historical cases of DLB. There is abundant biographical evidence that he exhibited all of the core features (only 2 of which are required for a probable diagnosis of DLB), as well as most of the ancillary features supportive of the diagnosis.

The burgeoning popularity of Peake’s work serves as a testament to his brilliant expressions of imagination. That such a gifted artist succumbed to a dementing illness with a predilection for attention and visuospatial function is particularly tragic. His later work stands as a unique and defiant record of his struggle with the disease. Recognition of the likely cause of his affliction as his popularity continues to grow may ultimately serve to heighten awareness of this distinct and devastating form of dementia.