

A Case of “Pfpopschizophrenia”: Kraepelin’s Bridge Between Neurodegenerative and Neurodevelopmental Conceptions of Schizophrenia

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The notion that schizophrenia, as conceived by Kraepelin, is a neurodegenerative disorder has been challenged by new research on the nature of the cognitive impairment in this disease; instead, schizophrenia is increasingly described as a neurodevelopmental disorder (1). For Kraepelin, the unifying observation among the catatonic, hebephrenic, and paranoid types of dementia praecox was a continued decline of intellectual function. However, empirical findings have demonstrated that cognitive function does not universally decline in patients with schizophrenia. Some authors have suggested that low intellectual functioning is a premorbid and fixed feature (2). Others have sought to separate patients according to different “trajectories” of lifetime intellectual function that include subsets of schizophrenic patients with intellectual functioning that was either compromised from a very early age or preserved at normal premorbid levels (3, 4). Finally, there have been attempts to determine if there are differences in change among cognitive functions (intellect, memory, executive, and attention) in schizophrenia (3). These data seem to be mutually exclusive of some of Kraepelin’s most important observations, and they generally support the “static encephalopathy” concept that asserts that schizophrenia is due to a stable neurodevelopmental brain lesion (5). The increasing dominance of the neurodevelopmental paradigm is illustrated by the American Academy of Child and Adolescent Psychiatry’s declaration that “schizophrenia is a neurodevelopmental disorder” (6) and also by Prusiner’s exclusion of schizophrenia (7) from the neurodegenerative disorders, all of which, in his perspective, “result from abnormalities in the processing of proteins.” One wonders whether this dichotomous landscape has room for only one perspective at a time.

However, neurodegeneration was not so monolithic in Kraepelinian thought. In fact, Kraepelin did recognize and appreciate cases in which low intellectual functioning existed long before the onset of psychotic thought and did not worsen with the onset of the psychosis. That is, Krae-

pelin described a type of schizophrenia that resulted from a disruption in neurodevelopment. Kraepelin’s “Pfpopschizophrenie” referred to the 3.5% (8) to 7.0% (9) of cases of dementia praecox that were “engrafted” (*pfpopf*, “to engraft” in German) upon, or the result of, “imbecility” during development. (The antique terms “imbecility” and “feeble-mindedness” refer to mild and moderate mental retardation; “idiocy” refers to severe and profound mental retardation.) This concept was a recurrent idea in Kraepelin’s textbooks and should not be seen as a historical anomaly. It was an active piece of his view of dementia praecox and was recognized and discussed by Bleuler and others in the field for many years.

This diagnostic entity is important because it suggests that at some level Kraepelin accepted a model of schizophrenia that was not simply neurodegenerative. His observations of pfpopschizophrenics, in fact, may be compatible with the ideas of the neurodevelopmental model. Of more importance, the modern reification of the concept of many schizophrenias, rather than just one, might rely on the demonstration of variations in cognitive impairment that reflect one of these paradigms and not the other. We suggest that this dichotomous relationship allows for coexistence, as Kraepelin did, and we propose that future study of

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this group in this manner would benefit both psychopathology as well as the clinical status of this vulnerable population.

In this article, we present a case of a patient who might have been diagnosed with pfpopschizophrenia. His case provides compelling evidence for the diagnosis of both mental retardation and schizophrenia. With the benefit of formal neuropsychological testing performed while the patient was both in the pre- and postmorbidity states, as well as some prenatal and childhood medical records, we review the difficult differential diagnosis of the case, the current literature on the relationship between intelligence and psychopathology, and the history of pfpopschizophrenia and its importance both to this case and to current thought about schizophrenia.

Case Presentation

Mr. A was a 29-year-old African American man who was transferred to the day hospital and shelter of our health center (the index admission) after his third lifetime inpatient psychiatric admission—a 7-month, court-

ordered forensic evaluation at a nearby state mental health center. On admission he stated, "I'm here to treat my paranoid schizophrenia."

Mr. A had been stable since his last inpatient admission 18 months before that admission. Although psychiatric care had been arranged for him at that discharge, he had not followed up. He had lived with his mother and brother without any psychiatric treatment at all. His mother described him as having been "his usual self, happy-go-lucky." His brother's social worker said that he had always been polite and interested in work during this period. He had had a job in a dry-cleaning facility, but he had quit because he believed the other workers were teasing him. Fifteen months before psychiatric admission, his mother had brought him to the emergency room when he would not open the door to his room—and he was discharged to his home.

As a result of an argument over cigarettes (his mother had refused to give him one of her own) 7 months before admission, Mr. A assaulted and battered his mother with the intent to kill her. He felt justified in his attack but understood that it was wrong. He was arrested and brought to court, where he was referred to the state inpatient forensic unit for evaluation. Test results were negative for substances of abuse in his urine and serum at the time of admission. Upon arrival on the inpatient unit, Mr. A was uncooperative and unwilling to participate in examinations. Because he was selectively mute, lying still, and unresponsive to attempts at engagement, he was described as "catatonic." His affect was without emotion, and he described his mood as "casual." When he did speak, he produced thoughts that were tangentially associated. At other times he produced meaningless word fragments and syllables. He complained of hearing "voices" from outside of his head telling him to "watch out." He was scared that his mother wanted to hurt or kill him. Mr. A reported no sadness, he had no periods of inflated self-esteem, and he did not have a lessened need for sleep. He reported no suicidal, homicidal, or violent ideation. He reported no feelings of guilt, no lack of concentration, no decreased libido, no change in appetite, no change in interest in pleasurable activities, and no fatigue in the weeks before the attack.

Once admitted to the forensic unit, Mr. A stayed in his room under the bed sheets for weeks. He was given risperidone, but it was discontinued because it caused a rash. Later he was given quetiapine, up to 700 mg/day. At that point there was little progress in terms of socialization and affect, but these aspects of his presentation improved with the additional administration of haloperidol on a daily basis. Later he was given propranolol for akathisia. He began to venture from his room and to socialize more. As he began to transition to the day hospital, he also began to visit his mother at her home, to which she did not object.

Mr. A's past psychiatric history began at age 16, 13 years before the current admission, when he was referred to the diagnostic clinic at a local hospital after an unarmed robbery. Results of an examination had discerned no thought disorder or perceptual disturbances. He received the diagnoses of conduct disorder and developmental learning disorder. Outpatient treatment was recommended, but he did not follow up. One year later he had a "breakdown," during which he punched his hand through a glass window, but he received no

care at the time. He may have been well for the 8 years between that incident and an episode 4 years before admission (at age 25) in which he cut his left wrist intentionally while cooking but sought no psychiatric care.

Mr. A's first psychiatric hospitalization was at age 27, 18 months before the current admission. He initially was brought to the emergency room because of bizarre behavior at home, including talking to, and swinging sticks at, persons not present. He was mute and curled into a fetal position on a stretcher when he was evaluated in the emergency room. He was referred to an inpatient unit, where he was also described as "catatonic." He was uncooperative and unresponsive, stayed in bed with the covers over his head, and had a clear sensorium. He responded to treatment with oral fluphenazine, 10 mg/day, and he eventually stated that stresses in his life had made him upset. He was discharged while taking fluphenazine. He did not take it after discharge, and he did not keep his outpatient psychopharmacology appointment. He was readmitted 5 days later when he was brought to the emergency department from a bank in which he had engaged in an altercation with a teller, later noting a wish to harm her. This second hospitalization lasted 3 months. During this time his psychosis was unresponsive to fluphenazine, risperidone, and haloperidol, but it dramatically responded to olanzapine. Results of an EEG at that time were normal. After the second discharge, he failed to follow up.

Mr. A occasionally used alcohol and had tried crack cocaine and marijuana one or two times each. His drug of choice was marijuana, although he may not have used it within 8 years of admission.

Mr. A's medical history was significant for intrauterine exposure to his mother's severe iron-deficient pancytopenia (her hematocrit measurement hovered around 29%), which was unsuccessfully treated with massive doses of iron. His mother also had hyperemesis gravidum, which was treated with chlorpromazine. She gained only 15 lb during his entire gestation; only 2 lb were gained in the third trimester.

Through much of his infancy and childhood, Mr. A suffered from the effects of malnutrition. At 2 months he was diagnosed with failure to thrive secondary to malnutrition, and at 10 months he was admitted to the hospital for viral meningitis, failure to thrive, iron-deficient anemia (with a hematocrit measurement of 26%), and pneumonia. He experienced a febrile seizure at 12 months. At 18 months his weight was less than the third percentile of the normal range, at which point he was treated for 5-minute generalized tonic-clonic convulsions with fecal incontinence. From ages 5 to 7 he had seizures that ceased without medication.

Mr. A's medical history also included asthma. He had a positive reaction to the purified protein derivative of tuberculosis and so was administered pyridoxine and isoniazid therapy in the months before the current admission. His HIV status was unknown, but he had few risk factors for HIV given his never having had sexual contact and his never having injected drugs of abuse.

Mr. A walked by age 1. He repeated the first grade. In the second grade, a teacher noted that he had "perceptual problems in the visual area, particularly visuomotor skills and possibly auditory processing problems combined with attention-getting behavior, [and] hyperactivity," and a psychologist observed that he did well with in-

dividual attention but had poor language skills and problems with visuomotor control. He had “no” friends, and peers did not allow him to play with them. He was seen as guarded and defiant, and he teased others for attention. In class he also repeatedly fell asleep, awaking “incoherent.” He was, however, good at telling stories. Neuropsychological studies were performed at ages 8 and 9, the results of which are noted in Appendix 1. The results showed his language skills to be “depressed.” Problems in visuomotor control were again noted, particularly in terms of fine motor skills. Gross motor skills, however, were normal. He also did not pass a hearing test at that time. He repeated seventh grade twice. He attended school until the ninth grade and then was expelled after “problems with aggression, including bullying younger kids,” and an attempted robbery.

Mr. A was the older of two sons of the same parents. His father was not present for almost all of his life. His mother had seven brothers and seven sisters; almost all of them and their respective families lived in the city housing project in which Mr. A, his mother, and his brother lived. Mr. A and his mother stated that they were “different” from the other family members and tried to keep an identity separate from them. All were, according to Mr. A’s mother, of normal height and mental capacity. Mr. A had never been sexually active, but he stated that he was heterosexual. He enjoyed playing games, such as basketball and ping-pong. The two brothers and their mother lived in an “extremely” impoverished manner, and until recently, they had had no telephone in the home. The mother supported all three of them with monthly funds that she received for the care of Mr. A’s brother.

Mr. A’s interactions with the law included an arrest as a teenager for possession of a gun. After involvement in an unarmed robbery, he was placed in the supervision of the state and put in a number of foster and group homes and in an institution for juvenile delinquents until age 17.

Mr. A’s family history was significant for low intellectual functioning in both his mother and brother. His mother had scored in the 50s on the WAIS-R intelligence test. Mr. A’s brother was born prematurely and at a low birth weight after a pregnancy that had also been complicated by pancytopenia and hyperemesis gravidarum. The brother’s gestation was also compromised by a varicella infection in the weeks before birth. At delivery the brother was a “floppy-blue immature male” with an Apgar score of 4 at 1 minute and 7 at 5 minutes. At 10 minutes he grunted and flared and was cyanotic while breathing room air. The results of Stanford-Binet testing at age 17 showed that his overall intelligence was far below average; he had a score equivalent to a mental age of 6 years and 6 months. His basic academic skills were at a first- and second-grade level. Significant delays had been noted in his receptive vocabulary and visuomotor integration. In the classroom, he was noted to be “severely hungry” for attention, but he was not noted to be aggressive. Neuropsychological testing at age 22 confirmed an intellectual age of 5–7 years.

Upon examination for the current admission, Mr. A was noted to be a thin, short, African American man who appeared to be younger than his stated age and who wore athletic clothing and berets or hats. He had a moustache that he allowed to grow long, but he had little other facial hair. His hair was braided; he usually

slouched in a chair. There were no appreciable extrapyramidal movements, no tremors, and no asterixis. He produced no abnormal movements. He was cooperative when he was not distracted. His speech was often slow; many words were poorly pronounced. His mood was “great” (which was incongruent with his life situation). Usually his affect was constricted and flat, but he had occasional, inappropriately large smiles. He produced loose associations, often making statements in groups that were entirely out of context. He described anxiety by saying, “Someone’s going to bite me,” when he was walking down a hallway; he reported no “anger” at that time. He did not report, and did not seem to be responding to, internal stimuli. He knew that he had a mental disorder but seemed unable to grasp its effect on his life. He reported no intent to harm himself or others, including his mother. His arousal level ranged from sleepy to awake; he was oriented to person, place, and time. He could spell “WORLD” but only backward: “D-L-W.” He recalled two of three objects immediately, which improved to three after prompting. According to Mr. A, the similarity between a bird and plane was that “They fly.” The results of tests performed at admission included normal thyroid levels. His hematocrit was in the anemic range (37.7%), and he had a mean corpuscular volume of 78 $\mu\text{m}^3/\text{cell}$.

Karyotyping of Mr. A, his brother, and his mother produced a sex-specific normal result for each. A fluorescent in situ hybridization study with use of the Tuple1 probe performed on region 22q11.2 of Mr. A’s chromosomes, the velocardiofacial syndrome area, found no abnormalities.

Neuropsychological tests performed while Mr. A was taking haloperidol and quetiapine showed overall intellectual functioning in the “extremely low” range. His functioning was borderline in terms of vocabulary and visuoconstruction on the WAIS-III and on measures of attention, verbal learning and memory, visual analysis, and fine motor speed, but his functioning was worse in the executive domains, such as problem solving, planning, reasoning, abstraction, and mental flexibility, and he was “slowed” in his thinking. His overall WAIS-III scores were full=63, verbal=65, and performance=66. Mr. A had poor spatial reasoning and scored lower than the first percentile for such tasks. He could not understand the Wisconsin Card Sorting Test (10). On the Trail Making Test (11), he took 50 seconds to perform the first trial (with no errors) and 158 seconds to perform the second (with two errors). Marked impulsivity and a low tolerance for frustration were observed. On the other hand, he showed relative strength in reading single words, naming categories, and recognizing on verbal learning tasks. His scores on the Wide-Range Achievement Test, 3rd ed. (12) placed him in the 14th percentile, consistent with “higher premorbid function” in the time before the current illness. It was felt that there was no evidence for a developmental learning disorder given the consistency of his skills. He demonstrated paranoid tendencies but no other thought disorders.

On the basis of his history and examination, Mr. A was admitted to the day hospital and continued taking the same medications. Results of random drug toxicology screens were negative. He remained in behavioral control. Very often his interactions with other patients consisted of annoying them “for attention,” he noted. He continued to complain of not having gotten enough

sleep, despite sleeping more than 10 hours per night, so his dose of quetiapine was reduced from 500 mg to 400 mg at bedtime.

A family meeting occurred on the unit that was attended by the authors, Mr. A, his mother, his brother, and his brother's social worker. During the meeting Mr. A's mother minimized the violence that Mr. A had inflicted upon her at the onset of the current episode. Strangely, during the meeting Mr. A was noted by his brother's social worker to be more vociferous and arrogant than ever before. He interrupted and dismissed his mother. Mr. A seemed to tease his brother, having the two shake hands repeatedly while the others spoke. His brother made some eye contact but produced no speech and often smiled on making eye contact.

After 4 months Mr. A left the day hospital and all care at our health center, although follow-up care had been recommended and established. At the time of this writing, he was living with his mother and brother, getting disability payments by means of a representative payee, and receiving no further care.

Discussion

Schizophrenia and Mental Retardation

Any psychiatric diagnosis in mild and moderately mentally retarded individuals is made with inherent difficulty and often relies on nonverbal communication. Mr. A received a diagnosis of schizophrenia because he met every criterion for the DSM-IV category: the characteristic symptoms of hallucinations, grossly disorganized or catatonic behavior, and negative symptoms had been present for more than 1 month; his social and occupational skills were very poor; his history failed to reveal evidence suggestive of schizoaffective disorder, bipolar disorder, a substance use disorder, or a general medical condition that contributed to his current state; the condition had lasted for more than 6 months; and no specific pervasive developmental disorder had been diagnosed in the past. The subtype was difficult to determine because his presentation included features of the catatonic, disorganized, and paranoid subtypes. We assigned the catatonic form because it superseded the others. DSM-IV criteria require several differential diagnoses. First, his impairments were not due to childhood disintegrative disorder, Asperger's disorder, Rett's disorder, or autistic disorder; none was diagnosed in childhood (nor was mental retardation, for that matter). The diagnoses of conduct disorder and learning disability, given at age 16, did not account for his current presentation. His illness was unlikely to be a mood disorder, given his lack of such symptoms. Attention deficit disorder was not borne out by neuropsychological testing. Mr. A's axis II diagnosis was mild mental retardation because his IQ was between 50–55 and 70 and because his low intelligence impaired his ability to adapt to the world according to the standards expected for his age. Thus, under the current system of diagnosis, Mr. A received two current diagnoses.

Given Mr. A's poor psychosocial adaptation at a young age, it is worth wondering whether his schizophrenia was simply present from his childhood. There is no "childhood

schizophrenia" in DSM-IV; theoretically, DSM-IV schizophrenia criteria apply to people of all ages. Schizophrenia that has its onset in childhood (before age 12) can be reliably diagnosed and differentiated from the pervasive developmental disorders (13). More recent research has found that childhood-onset schizophrenia is continuous with the adult-onset form in its clinical and neurobiological aspects (14). Nevertheless, Mr. A's presentation as a child was much more consistent with a deficit in cognition than in thought.

Genetic questions arise in considering Mr. A's family: Mr. A, his mother, and his brother all had intellectual deficits and iron-deficient pancytopenia. Mr. A, the only one with psychosis (his brother could not be evaluated for psychosis), was taller and less darkly pigmented than the other two. Thus, consideration of such a patient should include assessment of family genetics. There are many heritable conditions that produce disruptions in psychic life, producing either nonspecific psychosis or retardation. That schizophrenia is a familial condition was known, for example, by Kraepelin and Kety et al. (15), and the fact that patients with schizophrenia are more likely to have chromosomal anomalies was reported by Tsuang (16), documenting the higher prevalence of the XXX anomaly among female schizophrenic inpatients. We conducted genetic testing, hoping that, despite many shared phenotypic characteristics, the genotypic difference that was psychosis would manifest itself on a chromosomal level. We also used molecular genetic methods to test Mr. A for velocardiofacial syndrome, which is associated with schizophrenia and learning disabilities (17). As noted, the patient did not have velocardiofacial syndrome, and there was no common pattern nor any major genetic differences noted among Mr. A, his brother, and his mother.

Intelligence and Psychopathology

DSM-IV reifies the field's conception that mental retardation and schizophrenia are separate, independent conditions that are not mutually exclusive within the multiaxial system. Given the fact that Mr. A carried two diagnoses, it is important to review the current literature on their comorbidity. Both lines of modern research—i.e., the psychopathology of persons with mental retardation and the cognitive aspects of persons with schizophrenia—continue to establish the connection noted by Kraepelin.

In general, the types of psychopathology suffered by patients with mental retardation resembles those of normal individuals (18). However, many studies, notably the Isle of Wight Study (19), have documented that these occur together at a greater-than-normal incidence. Psychotic episodes among patients with mental retardation are not uncommon. Reid (20) found that 8% of retarded patients did have psychosis; Lund (21) documented schizophrenia in 1.3% of patients with mental retardation, as well as psychosis not otherwise specified in 5.0%. Thus, the current literature supports the idea that psychiatric disturbances are more common in patients with mental retardation than in persons of normal intelligence.

On the other hand, is schizophrenia necessarily associated with low intelligence? This has been the subject of many large studies that have relied on basic, yet standardized, examinations of intelligence. Lane and Albee (22) demonstrated in 1964 that future schizophrenic patients scored significantly lower on intelligence tests, compared both to age-matched peers and to their own siblings at the same ages. One study showed that there is a cross-sectional relationship between low IQ and psychosis (23). A cohort study of 50,000 Swedish 18-year-old army recruits (24) found a significant association between low intelligence at age 18 and subsequent schizophrenia over the entire range of intelligence scores. Of note, many studies have demonstrated that the premorbid deficits are greater in men than in women (22). These studies support the concept that the cognitive deficits noted in schizophrenia occur before the onset of psychosis.

Pfropfschizophrenia

Pfropfschizophrenia may reflect Kraepelin's appreciation for developmental, rather than degenerative, processes in schizophrenia. He estimated that from 3.5% (8) to 7.0% (9) of all cases of dementia praecox were due to "idiocy" or "imbecility," states of stark and readily apparent intellectual deficits. He defined this condition as "Pfropfschizophrenie"—schizophrenia engrafted upon mental retardation. Pfropfschizophrenia, since it included episodes of "schizophrenic" psychosis, differs from "oligophrenia," which described low psychic or mental functioning without psychopathology (Appendix 2 is a summary of the features of pfropfschizophrenia). Milici (25) described a patient with pfropfschizophrenia in terms of "some mental confusion, a more or less ill-defined symptomatology, and often a paucity of delusions which are evanescent and of a naive, childish type...the mental illness...usually has been precipitated by obvious forces [such as arrest or involuntary admission] in contrast to the generally trivial situations which often precipitate schizophrenia." Kraepelin initially included motor "mannerisms and stereotypies" within his definition, but, consistent with the objections of Bleuler (26) and Neustadt (27), he ultimately excluded disordered movements from the definition. Kraepelin specifically noted, however, that these patients were more likely to be hebephrenic (8, p. 260).

It is important to note that although Kraepelin recognized very-early-onset dementia praecox, pfropfschizophrenia was not equivalent to childhood schizophrenia (it was placed, in fact, in the sections of his textbook on retardation rather than in those on dementia praecox). It was a recurrent schizophrenic psychosis "engrafted" upon the pathologically developed brain of the feeble-minded. For Bleuler, who also recognized pfropfschizophrenia, the disorder was related to "a want of associations" that resulted from disordered development (26). It is likely that, for the same reasons that Bleuler discounted the importance of motoric signs in the diagnosis of schizophrenia in general, he also contested Kraepelin's inclusion of stereotypies and mannerisms in the definition of pfropfschizophrenia. This

disorder differed from "simple schizophrenia" (reproposed as "simple deteriorative disorder" in DSM-IV), which traditionally has described a type of schizophrenia characterized by poverty of content of thought and other negative signs in the absence of the hebephrenia or catatonia of pfropfschizophrenia.

The widespread interest in pfropfschizophrenia reflects a time in the history of psychiatry in which attention to intellectual functioning was great (it coincided with the development of standardized tests of intelligence). A former superintendent of our institution, E.E. Southard, proclaimed that "the slightest evidence of any kind or degree of mental deficiency appears to me to have extraordinary importance" (28), and he made extensive studies of the connection between low intelligence and schizophrenia (29). This diagnostic category was bolstered by the work of both Meyer (30) and Karl Menninger (31), both of whom agreed that, as in pfropfschizophrenia, schizophrenic psychoses could be "reversible." Clarence O. Cheney (32) accepted the notion of engraftment as Kraepelin had proposed. On the other hand, May (33) did not agree with the idea of engraftment. Of "feeble-minded patients" brought to the hospital with psychosis, he noted, "there is no logical reason for the assumption that this is a combination of two diseases, mental deficiency and dementia praecox, or that the latter psychosis has been engrafted upon the former condition. That the feeble-minded are subject to psychotic episodes has been known for centuries" (33).

It is unclear exactly how it happened, but the excitement over pfropfschizophrenia—and of intellectual deficits in schizophrenia—declined just before the second world war. It is possible that this reflects the influence of either psychodynamic thought in the United States, Schneiderian first-rank symptom profiles in Europe, or the separation of the classification of mental retardation from the purview of psychiatrists to specialists in mental retardation. A MEDLINE search for "propfschizophrenia," conducted in 2000, revealed no related citations, and a search for "oligophrenia" uncovered articles mostly from the states of the former Soviet Union. Of interest, the diagnosis was retained in Arieti's textbook (34) at least as late as its 1965 edition.

Thus, it seems contrary to current opinion in the field to propose pfropfschizophrenia as a diagnosis in this instance. However, it is a diagnosis that is unifying, parsimonious, and meaningful as cognitive deficits assume greater importance in the study of schizophrenia. On the basis of our distillation of Kraepelin and his followers' descriptions of pfropfschizophrenia (Appendix 2), Mr. A's illness seems to fit the picture of pfropfschizophrenia. When psychotic, he showed catatonic-like states as well as hebephrenia; at other times the psychosis was hardly detectable. He certainly had a low level of intelligence all of his life but not lifelong psychosis. His psychotic episodes were precipitated by significant (such as arrest or hospitalization), not trivial, events. And finally, although Kraepelin rescinded motor abnormalities as a part of his definition, the patient did manifest catatonic features,

adding to the picture of pfpopschizophrenia. An interesting modern study comparing the characteristics of those who are both mild to moderately retarded and schizophrenic versus healthy comparison subjects found that the patients had a greater frequency of premature birth, lower birth weight, and impaired hearing (35), all of which Mr. A had as well. Thus, we feel that Mr. A might have fit the diagnosis of pfpopschizophrenia.

Finally, the current episode began with commission of a violent crime, after which this patient was remanded to a forensic unit; it is important to address issues of legal responsibility and competency in someone like this patient. It seems that the combination, or the "engraftment," of schizophrenia on Mr. A's intelligence status should not necessarily confuse the legal picture. There are standards for assessing the competency of any individual with a mental disorder; suffering from two mental disorders should neither increase nor decrease the potential of being found incompetent. All mentally ill offenders, such as Mr. A, should be assessed on a case-by-case basis without prejudice.

Conclusions

The movement toward the static, neurodevelopmental model of schizophrenia is specifically supported by evidence regarding cognition (21) that contradicts Kraepelin's progressive, neurodegenerative model. The presence of propfschizophrenia in Kraepelin's textbooks suggests that, at some level of understanding, the originator of dementia praecox accepted that the disorder could be the result of a defect in normal brain development. This raises an issue that must be constantly reified in schizophrenia research: there may be many types of schizophrenia. To the extent that we have established the notion that Kraepelin accepted multiple psychopathic mechanisms, the field should continue its work in establishing the different types of this disorder through the study of symptom profiles as well as molecular and chromosomal genetics, degenerative, developmental, or environmental models or a combination of them all. Propfschizophrenia may have been Kraepelin's acceptance of all of these models, and we must not allow the neurodegenerative/neurodevelopmental dichotomy to impede any line of inquiry.

Mr. A's case is compelling because of his stark disorder in functioning in the community. Had he come to our institution 80 years ago, during a time of great interest in the relationship between cognition and psychosis, the "extraordinary importance" of the case would have been noted by E.E. Southard. We hope for a resurgence in the study of patients with both schizophrenia and mental retardation. There is clear evidence of the greater frequency of mental disorders in retarded persons. With pfpopschizophrenia and Kraepelin's observations as a model, we feel there may be much to gain by such study.

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The authors thank Daphne Dorce, M.D., Peter Tischler, M.D., Michael Murphy, M.D., Margaret Lenca, Ph.D., Larry Seidman, Ph.D., Gwen Zornberg, M.D., and Roberta Apfel, M.D., for their help with this article and Conny D. Huthsteiner, M.D., for translating the German reports.

APPENDIX 1. Results of Neuropsychological Tests Administered to a Patient With Possible Pfpopschizophrenia

Age 9

WISC-R: borderline score
Wide-Range Achievement Test, 3rd ed. (WRAT-3): word recognition score=1.6, spelling score=1.8, arithmetic score=1.8
Limited communication
Poor auditory discrimination

Age 10

WISC-R: low-average score, with mental age of 9.1, but serious persistent visuomotor deficits and problems with auditory sequencing
WRAT-3: word-recognition score=2.6, spelling score=2.6, arithmetic score=3.1
Slosson Intelligence Test: score higher than those in previous years
Peabody Individual Achievement Test: score=2.6
Wepman Speech Sound Perception Test: decreased powers of auditory discernment
Beery Test of Visual-Motor Integration: severe visuomotor problems; mental age closer to 5.6 years
Goodenough-Harris Drawing Test: immaturity

APPENDIX 2. Summary of the Features of Pfpopschizophrenia

Hebephrenic features

Paucity of delusions, which are evanescent and of a naive, childish type
Interepisode periods of childlike, easygoing behavior that is not psychotic
Mental retardation
Psychotic episodes precipitated by significant, nontrivial experiences
Stereotypies or other movement disorders (removed by Kraepelin later)

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