Folie a Deux Versus Genetically Driven Delusional Disorder: Case Reports and Nosological Considerations

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Folie a deux is thought to occur in persons who live close together, so that delusions of the primarily ill patient are adopted by the partner. Alternatively, if both patients are related, they may share the same genetically driven psychiatric illness. We present the cases of two psychotic couples (monozygotic twins and a mother and daughter pair, respectively), in which both subjects featured an almost identical psychopathological syndrome with the same delusional content and thus fulfilled the diagnostic criteria of folie a deux. When Leonhard’s diagnostic system was applied, all four patients were found to suffer from affect-laden paraphrenia, arguing against the presence of folie a deux in those dyads. Thus, thorough psychopathological examination in suspected folie a deux settings can help to obtain valid diagnoses.

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DELUSIONAL and paranoid disorders belong to the most fascinating diseases in psychiatry. The term “paranoia” originally was used to denote an isolated delusion. More commonly, but less precisely, it is referred to as the delusion of persecution by others; it then is opposite to the delusion of grandeur. Both delusions can occur in the same patient at the same time, while others show monosymptomatic delusions throughout their life.1 The absence of “dementia praecox” symptoms prompted Kraepelin to coin the term “pure paranoia” as a distinct nosological entity.2 Hallucinations may occur, albeit this is not necessary. However, paranoiac delusions as well as delusional ideas in general are not specific for a distinct psychiatric disorder, but rather symptoms of various nosologic entities; however, the group of schizophrenias is found most frequently in this respect. In the ICD-10 classification,3 delusions can be found in several subgroups of schizophrenia, but also as monosymptomatic delusional disorder or within schizoaffective disorders. The psychobiology of delusion up to now remains almost unknown. This might be due to the fact that the phenomenon “delusion” is not uniform and moncausal, but rather a symptom of varying pathophysiologic mechanisms.

Folie a deux (FD), or shared psychotic disorder, is a rare psychiatric condition in which an “inducer” (primary patient), who is the “originally” ill patient, transmits his delusional beliefs on another patient; both then share the same disordered ideation. FD is found in parent-offspring, sibling-sibling, or husband-wife constellations, albeit 50% of the psychotic dyads consist of mother-daughter or sister-sister pairs4; risk factors include longstanding social isolation and very close relationship of the psychotic couple, as well as mental retardation of the secondary patient.5,6 Interestingly, all kinds of rare delusional contents can be transmitted within the FD setting, eg, Cotard’s syndrome,7 Capgras syndrome,8,9 and Fregoli illusion,10 koro,11 de Clerambault’s,12 and Ekbom’s syndrome.13

The differentiated psychopathology of Leonhard and Beckmann14 subdivides schizophrenia in unsystematic and systematic schizophrenias, which can be divided in further subforms.15 One subgroup of the unsystematic schizophrenias shows predominantly delusional symptoms and is called affect-laden paraphrenia (AP).14 Often starting with a so-called irritated reference syndrome, AP thereafter progresses to its full syndrome which features irritation, suspicion, and delusions, which are triggered by pathological affect; delusions can be paranoid or grandiose, frequently both are present. Hallucinations often accompany the delusional ideation. Patients suffering from AP show unshakeable delusional beliefs and tend to get hostile and upset when being contradicted. The diagnostic criteria are summarized in Table 1. AP is substantially genetic in its origin16 and seems to follow autosomal-recessive heritability.17 However, the penetrance is variable, and anticipation might play an important role as do yet unknown biological factors; interestingly, women suffer more often from this disorder than men.18 Within ICD-10, AP has to be classified under various nosologic entities.

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diagnoses: paranoid schizophrenia, schizoaffective disorder, and delusional disorder.

We here present two patient dyads in which the psychotic couples featured almost identical psychiatric symptoms. The first couple, a pair of monozygotic twins, is presented in greater detail because of the extreme course of illness.

CASE HISTORIES

Case 1: B.N. and C.N.

B.N. and C.N. were born 1958 as monozygotic twins as evidenced by molecular genetics. Their mother had no psychotic illness. The father died in the 1970s due to alcohol intoxication. He was reported to be extremely suspicious and paranoid, and seemed to suffer from acoustic hallucinations. Based on the available information, a working diagnosis of paranoid schizophrenia can be assumed. The twins’ brother is mentally healthy.

Both twins completed high school without problems. During their early 20s, psychic alterations were noted for the first time. The twins, who were formerly known as pleasant and kind persons, became hostile and violent towards their mother and elicited strange behaviors, such as throwing away all of their clothing and disconnecting all fuses. Due to their aggressivity, they were committed to a psychiatric hospital, where their relationship with one another was described as symbiotic and pathological. Paranoid delusions, irritability, suspiciousness, and cenesthetic hallucinations were noted. A diagnosis of paranoid-hallucinatory schizophrenia was established for C.N.—the more dominant of the twins—and FD was suspected in the case of B.N. The latter was readmitted 2 months later, while her sister was referred to a state hospital.

B.N. again displayed ideas of reference, irritability, cenesthetic hallucinations, and delusions of persecution and grandiosity. She stated that she deserved a special treatment due to the fact that she claimed to be an “elite human being.” Consistently, irritability, increased drive, incoherence, and the delusion of being poisoned were noted. C.N. was also admitted to various psychiatric hospitals and displayed a psychopathology always similar to her sisters’. She thought she was an elite woman, and therefore claimed special treatment. Together with such grandiose ideas, persecutory delusions and fear of being poisoned were noted, as were irritability and suspiciousness. Meanwhile B.N. managed to rent an apartment and C.N. shortly thereafter moved in with her. However, the social and hygienic situation of the twins constantly deteriorated, with consecutive but fruitless and short commissions to various hospitals.

In the year 1995, C.N. was admitted to a state hospital, where she spent the following 4 years. As she did not cooperate with respect to neuroleptic treatment, she only made slight progress and was consistently delusional, e.g., she was convinced that doctors would experiment with her by conducting neurosurgery. She was also irritable and affect-laden. Later, she claimed to be in a concentration camp and subject to torture. A bathtub had been programmed to handicap her physically and mentally. Following treatment with perazine, her situation slowly improved. B.N. still lived in her apartment, which was described as extremely dirty;
she spent her days in the streets and subways and her psychopathology remained constant, with continuing delusions of persecution and poisoning, as well as corresponding hallucinations.

In January 2000, C.N. was dismissed and again moved in with her sister, which caused the rapid decompensation of B.N. The living situation now became more and more problematical. Six months later, B.N. unexpectedly came to our clinic. She was very upset, irritated, and in a deprived, cachectic state. Her speech was incoherent; neologisms were present, as were grandiose and persecutory ideas. She claimed that an apparatus was implanted in her head, by which electroshocks were delivered. Upon medication, she gradually improved, albeit only slowly. Her formal thought disorder soon disappeared, and she described her delusional beliefs: her sister had obstructed the toilet with parts of dead bodies, she should be poisoned since the hospital food had tasted like flesh of rotten corpses. The police had told her that a computer had to be implanted in her brain, and she had been programmed by NASA, which also attacked her with radiation and ABC weapons. By a surgical procedure, “farting glands” and animal parts had been implanted in her. She was convinced that mankind soon would leave earth in rockets, that elite persons like lawyers or businessmen had already left the planet, and only stupid people would remain here. Retrospectively she described the occurrence of Capgras’ syndrome with respect to her sister. She admitted to hearing imperative voices constantly throughout the last 5 years. Delusions were unshakable and accompanied by markedly irritable affect; apart from her delusional system, during treatment B.N. became friendly and approachable. Albeit insight could not be obtained, she learned to hide her delusions. We could monitor her regularly; her health state successively increases, so that she is able to live in a rehabilitation facility and to perform office work again. Her delusional ideas are still present, albeit under disclosure.

During B.N.’s stay in our department, neither B.N. nor her mother knew the whereabouts of C.N. Several months later she was transferred to our department at the instigation of her legal guardian: C.N. took a flight to a foreign country and was arrested at the airport due to psychiatric symptoms. Again, she was described as logorrheic, delusional, and irritated. Shortly thereafter she was transferred to a psychiatric hospital, where she stayed the next 9 months. When C.N. was admitted to our department, we were frankly surprised by the striking similarity between the twins, not only in physical, but also in psychopathological terms. C.N. displayed the same psychopathology as did her sister: she spoke in the same fast and irritated, logorrheic manner, with similar pronunciation, and was equally upset and hostile when talking about her delusions and the circumstances of her admission. She reported again a lot of delusional ideas; the evidently contradicting reality did not bother her. As a part of a misidentification syndrome involving her family, she also noted that people constantly vanished and new ones appeared. She denied that she had harmed her sister, but stated that B.N. had been shot by laser weapons in a psychiatric hospital. Her beliefs were absolutely unshakable and contradiction was answered with anger. In the beginning of therapy, she quite often was irritated and abusive; she talked to herself and to imaginary persons, which gave us the impression that she was hallucinating, although she denied this. In the same manner as her sister she complained about being operated by the use of medications causing “pain, disintegration, and seizures.” In the following weeks, C.N. constantly improved. Although she always discussed her medication dosage, she was quite friendly in everyday life; similar to her sister, she soon kept her delusional ideas to herself and calmed down, and her underlying charming and cheerful character became evident. At the time of writing, she is in her best condition ever since her disease started and close to discharge.

Case 2: T.U. and U.U.

T.U. was born 1961; her premorbid personality was described as shy, cautious, and slightly suspicious. While she was studying, she became ill for the first time at the age of 25. She was noted to be aggressive and hostile; she suddenly claimed to be anesthetized and raped, albeit this seemed not to be true. On admission to a state hospital, she had paranoid ideas, frightening auditory and cenesthetic hallucinations, as well as ideas of reference and poisoning. Following neuroleptic treatment, her delusional ideas cleared and she was dismissed recovered. However, she was admitted again 1 year later, with comparable symptomatology. Suspi-
sciousness as well as tension and slightly disordered thought were described. Paranoid ideas, which were not noted in detail, were present with respect to her parents and her husband. Furthermore, she had the delusion—as well as corresponding cenesthetic hallucinations—that she was pregnant due to a violation. She responded well to treatment and was diagnosed as suffering from paranoid schizophrenia.

In 1988, she was first brought to our department for treatment. T.U. showed paranoid ideas, and was extremely suspicious, irritated, anxious, and depressed. Within 2 weeks, she became logorrheic and reported some of her delusional beliefs: her parent’s house had been turned into a brothel, and she had heard voices of other people discussing her sexual life. During treatment, cenesthetic and olfactory hallucinations were seen, as well as the corresponding delusion of being pregnant. Six months thereafter she presented again with aggressivity and social retreat; she displayed an irritate reference syndrome and multimodal hallucinations including olfactory ones. Upon administration of neuroleptics, she gradually improved and could be discharged in partial remission. During 1992, 1994, and 1998, similar stays at our department followed; intermittently, she worsened, but her mother refused to take her to a psychiatric hospital. She reported in 1998 that she had been raped in her childhood. She wrote in a letter: “I notice varying signs […] like a swollen throat, my olfactory organs sense strange and alien odors like the smell of spermatic fluid in my clothes.”

Two years later, she was brought to our department due to increasing aggressivity and irritability. She claimed later on that other, “lesbian” patients had abused her sexually during the night, which was for sure delusional. Her delusions had become systematized; she also suspected her father and his friends of having raped her, probably by the use of narcotics. Signs thereof included photographs of her in which she could detect “clear signs” of abuse, which she showed repeatedly to evidence her delusion. She was sure that she did not suffer from any psychiatric disorder but from a “psychological deficit,” stemming from those incidences. The delusions were accompanied by marked irritation. Partial remission was again achieved by neuroleptic medication, albeit her delusions are still vividly present.

U.U., the mother of T.U., was seen during her daughter’s stay in 1998 in our department. While talking to her about T.U.’s disorder, she suddenly became very upset and hostile; she reported that the reason for the disease had been sexual abuse by T.U.’s uncle when she was 2 years old. She was irritated, affect-laden, and hostile, and any attempt to calm her down was futile; her belief was unshakable, and three examiners suspected a diagnosis of delusional disorder or affect-laden paraprenia, respectively.

During the stay of T.U. in the year 2000, one of the present authors (A.R.) saw U.U. for the first time. While talking to her about the disorder of her daughter, she became upset and very hostile. She was sure that her daughter’s disorder was caused by the aforementioned sexual abuse at the age of 2, albeit she never witnessed it directly. Upon slight contradiction, U.U. became aggressive and rushed outside the ward.

U.U. has been recently admitted to our department in October 2001; she had been referred by the department of dermatology. She had been there for the treatment of dermatitis, which was according to her due to ice cream. She was sure the ice was poisoned by a white powder, which she suspected to be anthrax. Similar things happened with another ice cream; she was sure she had been the victim of a terrorist anthrax attack, which also caused her dermatitis. During her stay in our department, one of the present authors (A.R.) examined her; she did not remember the encounter 2 years before. When being asked about her family history, she said that her daughter T.U. was said to suffer from schizophrenia, and that she was still very angry and upset about this diagnosis because it would be wrong—in fact, her daughter had been abused; she again reported the above-mentioned delusion in almost the same words, with unshakable belief and strong affective involvement.

**DISCUSSION**

FD is a rare psychiatric condition that might cause diagnostic problems, especially if the partners are consanguineous: does the supposed secondary patient suffer indeed from a “transmitted” delusional belief or from an independent, endogenous psychosis? In the case of unrelated patients, a genetic connection can be ruled out; we will thus focus on related subjects. Two exemplary cases of psychotic couples have been presented, in
which both participants shared similar psychopathology and delusions—in the case of the strikingly similar twins B.N. and C.N., persecutory delusions of various types, grandiose ideas that they were elite beings, the fear of being intoxicated or infected, the feeling of impairment by radiation weaponry and operations, cenesthetic hallucinations, and misidentification syndromes; in the case of the mother-and-daughter dyad T.U. and U.U., ideas of sexual abuse of T.U. which were shared and reinforced by U.U. and were without doubt of delusional character.

To our knowledge, this is the first case report in the English literature that involves the shared delusion of sexual abuse both in mother and child; there is, however, one report of this shared delusion in siblings.\(^{21}\) To be sure, the possibility that the abuse indeed happened must be ruled out in such cases. In our patients, there was not the slightest evidence for a real sexual molestation. Especially regarding the “victim” T.U., some similarities to the so-called false memory syndrome are present, albeit in that condition memories commonly “reappear” during hypnotherapy or psychotherapy\(^{22}\) and the ideas are not kept against all evidence to the contrary. In this respect, confabulations are a further differential diagnosis, but the monosymptomatic character of the ideas that were retained without substantial variation over time rules out this possibility.

In both cases, the diagnosis of FD could be applied formally, as the relationship was unusually intimate and there was a dominant partner (the mother in the second case and the more dominant sister in the first) in every couple. However, in FD, psychosis in the secondary patient should remit when the partners are separated, which did not occur in our examples (both dyads were separated for a significant time due to therapeutic interventions).\(^{4}\) We therefore reject the diagnosis of FD for our cases, but rather argue for the presence of an endogenous psychosis in all four participants, namely, AP according to Leonhard’s nosology, in which the delusional ideas were colored by the intimate relations in a pathoplastic manner.

All participants of the presented cases were independently diagnosed as suffering from AP, fulfilling all of the exactly defined criteria of this disorder (Table 1).\(^{14}\) In FD, one should expect that only the delusional content is shared by the secondary patient, but not the full syndrome of a distinct nosological entity. Interestingly, FD in most cases involves delusional beliefs; we are not aware of reports that other psychopathological symptoms, such as psychomotor disturbances, flattened affect, or formal thought disorder, are transmitted to the secondary patient. According to Leonhard’s nosology, the primary patient is most likely to suffer from AP with its prominent and longstanding delusions. In other forms of endogenous psychoses, delusions are only fluctuating and less prominent or—as in the case of some forms of systematic paraphrenia—stated without any affective involvement.

AP thus remains an important differential diagnosis in a FD constellation in consanguineous partners. It more often affects women and has a substantial genetic background.\(^{18}\) Similarly, FD affects predominantly women\(^{22}\); sister-sister and mother-child relationships account for about 50% of all reported cases.\(^{4,23}\) and FD occurs in about 70% in first-order relatives, arguing for at least some genetic basis.\(^{4}\) White\(^{24}\) found 16 cases of FD in mostly monozygotic twins in the English literature up to 1995. One further case was presented by Shiwach and Sobin in 1998.\(^{25}\) Interestingly, the latter could show that the primary diagnosis in monozygotic twins pairs suffering from FD is delusional disorder, which corresponds well to AP. As in our case of C.N. and B.N., the reported cases of FD in monozygotic twins might most likely represent cases of AP. Similarly, on critical consideration of the FD cases presented by Enoch and ball,\(^{4}\) one might suspect AP rather than FD in three of the five cases, albeit statements based on third-person accounts present difficulties.

Regarding the diagnosis of FD, we suggest that every participant be examined separately with emphasis on the psychopathology of the patient. If a full psychopathological syndrome is present, especially that of AP, the diagnosis of FD should be rejected. The shared psychotic beliefs should then be regarded as pathoplastic coloring of the disorder. Conversely, the term FD should only be applied in related patients when the following main criteria are met (modified from Enoch and Ball\(^{4}\)): (1) a high degree of commonality in the content of delusion; (2) absence of the same clear-cut psychopathological syndrome according to distinct disease entities.
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in the primary and the secondary patient; (3) significant improvement of the symptoms in the secondary patient after separation from the primary patient without substantial neuroleptic treatment; (4) intimate relationship and living in close proximity; and (5) dominance of the primary patient in some respect. Supporting criteria may include mental retardation or dependent personality structure in the secondary patient, living of the couple in poverty or in isolation, and the presence of some kind of advantage for the secondary patient in sharing the delusions. The concept of folie simultanée as proposed by Gralnick and modified by Munro should be used with caution, especially in siblings; in our hands, the term FD should be reserved for the folie imposée type, as folie simultanée could well be due an autochthonous disorder in both participants.

In conclusion, we suppose that a substantial portion of reported FD cases in relatives actually represent AP in both patients. A thorough distinction between FD and AP is crucial and pertinent for further treatment; furthermore, the application of differentiated psychopathology might empower molecular genetic studies of schizophrenic disorders, which have to date been obfuscated by the apparent heterogeneity of the samples studied. In particular, AP is an attractive candidate disorder for gene mapping due to the significant genetic background of the disorder, so that a clear distinction between this disorder and FD is a prerequisite to obtaining valid findings.