

Delusional Disorder and Shared Psychotic Disorder

Once viewed as too rare to warrant a separate classification, delusional disorder has emerged in recent years as a focus of clinical research and treatment innovation. Better definition and a growing literature have revitalized the efforts to characterize, understand, and treat these conditions. Limited but growing evidence supports not only its occurrence, but its distinctiveness from schizophrenia and mood disorder as well as its treatability. Delusional disorder refers to a group of disorders, the chief feature of which is the presence of a nonbizarre delusion. It is the delusion and the relative absence of other psychopathology that unifies these disorders in terms of natural history and impact on functioning. Once called paranoia, this condition as defined in the fourth edition of Diagnostic and Statistical Manual of Mental Disorders (DSM-IV) and the 10th revision of International Statistical Classification of Diseases and Related Health Problems (ICD-10) is easier to recognize and less subject to misdiagnosis.

Despite such advances, clinicians are relatively ill-informed about delusional disorders and many have only seen an occasional example. There are several possible reasons why this is so. Persons with this condition do not regard themselves as mentally ill and actively oppose psychiatric referral. Because they may experience little impairment, they generally remain outside hospital settings, appearing reclusive, eccentric, or odd, rather than ill. If they do have contact with professionals, it is more likely to be with lawyers regarding litigious concerns; with medical specialists regarding health concerns; or with the police regarding complaints of trespass, persecution, or threat, rather than with psychiatric clinicians regarding complaints of emotional disorder. A hallmark of these disorders is that the patient does not believe that he or she is deluded or in need of psychiatric assistance. In the infrequent psychiatric encounter, clinicians tend to diagnose these disorders as other conditions, often as schizophrenia or mood disorders.

Although delusional disorders are uncommon, they are probably not as rare as previously thought. While many individuals with such disorders seek assistance from other medical specialists, judges, or the police, they are increasingly being recognized as psychiatrically ill. The relationship of these disorders to other psychoses remains unclear, and much about them is puzzling. The DSM-IV requirement of excluding other conditions is prudent given the special importance of differential diagnosis. Armed with newer and better criteria, clinical research is ongoing in areas such as natural history, pathogenesis, neuropsychology, neuroimaging, treatment, and even genetics. Although the DSM-IV criteria are not definitive, they have provided a sound basis for clinical and research investigation. Systematic studies based on larger samples of these disorders are needed to anchor classification with sound information; however, such studies may be difficult to conduct because of the large numbers of patients required and their reluctance to participate in research. A biological basis for these disorders is proposed on many grounds, but its definition remains elusive. The study of misidentification syndromes (e.g., the Capgras's syndrome) has led to interesting hypotheses and methods that draw on neuropsychological and clinical insights that may inspire progress in delusional disorders. Treatment remains an obstacle, although recent reports suggest that favorable responses to psychopharmacologic and psychotherapeutic interventions are more common than previously thought.

DEFINITION

Delusional disorder is the current classification for a group of disorders of unknown cause, the chief feature of which is the delusion (Table 13.2–1). Although the specific content of the delusion may vary from one case to the next, it is the occurrence of the delusion, its persistence, its impact on behavior, and its prognosis that unifies these seemingly different disorders. In considerable agreement with Emil Kraepelin's concept of paranoia, the revised third edition of DSM-III-R provides reliable criteria for identifying cases and collecting systematic information about these conditions. This development in classification helped to reestablish the clinical importance of this group of disorders and may have reversed a trend of infrequent diagnosis. The criteria use the term delusional to avoid the ambiguity of the term paranoid used earlier in the third edition of DSM (DSM-III) classification, paranoid disorders, as well as to emphasize that the category includes disorders in which delusions other than those of the persecutory or jealous type are present. Although these changes were initially confusing, especially in terms of comparisons to diagnostic approaches elsewhere, they have gained acceptance and have created a more level playing field for further empirical contributions.

Table 13.2-1. DSM-IV Definition of Delusion and Certain Common Types Associated With Delusional Disorders

Delusion A false belief based on incorrect inference about external reality that is firmly sustained despite what almost everyone else believes and despite what constitutes incontrovertible and obvious proof of evidence to the contrary. The belief is not one ordinarily accepted by other members of the person's culture or subculture (e.g., it is not an article of religious faith). When a false belief involves a value judgment, it is regarded as a delusion only when the judgment is so extreme as to defy credibility. Delusional conviction occurs on a continuum and can sometimes be inferred from an individual's behavior. It is often difficult to distinguish between a delusion and an overvalued idea (in which case the individual has an unreasonable belief or idea but does not hold it as firmly as is the case with a delusion).

Delusions are subdivided according to their content. Some of the more common types are listed below:

Bizarre—A delusion that involves a phenomenon that the person's culture would regard as totally implausible.

Delusional jealousy—The delusion that one's sexual partner is unfaithful.

Erotomaniac—A delusion that another person, usually of higher status, is in love with the individual.

Grandiose—A delusion of inflated worth, power, knowledge, identity, or special relationship to a deity or famous person.

Mood-congruent—See mood-congruent psychotic features.

Mood-incongruent—See mood-incongruent psychotic features.

Of being controlled—A delusion in which feelings, impulses, thoughts, or actions are experienced as being under the control of some external force rather than being under one's own control.

Of reference—A delusion whose theme is that events, objects, or other persons in one's immediate environment have a particular and unusual significance. These delusions are usually of a negative or pejorative nature, but also may be grandiose in content. This differs from an idea of reference, in which the false belief is not as firmly held nor as fully organized into a true belief.

Persecutory—A delusion in which the central theme is that one (or someone to whom one is close) is being attacked, harassed, cheated, persecuted, or conspired against.

Somatic—A delusion whose main content pertains to the appearance or functioning of one's body.

Thought broadcasting—The delusion that one's thoughts are being broadcast out loud so that they can be perceived by others.

Thought insertion—The delusion that certain of one's thoughts are not one's own, but rather are inserted into one's mind.

Mood-congruent psychotic features—Delusions or hallucinations whose content is entirely consistent with the typical themes of a depressed or manic mood. If the mood is depressed, the content of the delusions or hallucinations would involve themes of personal inadequacy, guilt, disease, death, nihilism, or deserved punishment. The content of the delusion may include themes of persecution if these are based on self-derogatory concepts such as deserved punishment. If the mood is manic, the content of the delusions or hallucinations would involve themes of inflated worth, power, knowledge, or identity, or a special relationship to a deity or a famous person. The content of the delusion may include themes of persecution if these are based on concepts such as inflated worth or deserved punishment.

Mood-incongruent psychotic features—Delusions or hallucinations whose content is not consistent with the typical themes of a depressed or manic mood. In the case of depression, the delusions or hallucinations would not involve themes of personal inadequacy, guilt, disease, death, nihilism, or deserved punishment. In the case of mania, the delusions or hallucinations would not involve themes of inflated worth, power, knowledge, or identity, or a special relationship to a deity or a famous person. Examples of mood-incongruent psychotic features include persecutory delusions (without self-derogatory or grandiose content), thought insertion, thought broadcasting, and delusions of being controlled whose content has no apparent relationship to any of the themes listed above.

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In 1994 DSM-IV refined the definition and the boundaries with other disorders, including substance-induced disorders, mental disorders due to general medical conditions, mood disorders, and schizophrenia. No laboratory test exists to assist in diagnosis. The DSM-IV definition, like its predecessors, hinges on the presence of a nonbizarre delusion. DSM-IV acknowledges the difficulty of judging whether a delusion is bizarre, meaning clearly implausible, not understandable, and not derived from ordinary life experiences. In contrast, the nonbizarre delusion involves situations or circumstances that can occur in real life (e.g., being followed, infected, or deceived by a lover). DSM-IV also emphasizes the differential diagnoses of schizophrenia, mood disorders, substance-induced disorders, and mental disorders due to a general medical condition before the diagnosis of delusional disorders can be made. These conceptual refinements and demarcations from other conditions have increased the usefulness of the criteria for delusional disorder.

Delusional Disorder

According to DSM-IV, the diagnosis of delusional disorder can be made when a person exhibits nonbizarre delusions of at least 1 month's duration that cannot be attributed to other psychiatric disorders. Definitions of the term delusion and types relevant to delusional disorders are presented in Table 13.2–1. Nonbizarre means that the delusions must be about situations that can occur in real life, such as being followed, infected, loved at a distance, and so on; that is, they usually have to do with

phenomena that, although not real, are nonetheless possible. There are several types of delusions, and the predominant type is specified when the diagnosis is made. In general, the patient's delusions are well systematized and have been logically developed. The person may experience auditory or visual hallucinations, but these are not prominent features. Tactile or olfactory hallucinations may be present and prominent if they are related to the delusional content or theme, examples are the sensation of being infested by bugs or parasites, associated with delusions of infestation, and the belief that one's body odor is foul, associated with somatic delusions. The person's behavioral and emotional responses to the delusion appear to be appropriate. Impairment of functioning is not marked and personality deterioration is minimal, if it occurs at all. General behavior is neither obviously odd nor bizarre.

Shared Psychotic Disorder

This unusual condition has also been called *folie à deux* and induced or shared psychotic disorder. It develops in an individual in the context of a close relationship with another person who has an established delusion that he or she also believes, and requires an absence of psychotic disorder prior to the onset of the induced delusion; it is usually classified with paranoid disorders.

HISTORY

Nineteenth-century psychiatry devoted much attention to the description of paranoid disorders, in which delusions are a cardinal feature. Karl Ludwig Kahlbaum's description of paranoia in 1863 was the first in a series of contributions that culminated in the classification of paranoia, and inspired that of *folie à deux*, morbid jealousy, the better-known schizophrenias, and mania. His work also led to a recognition that paranoid features are nonspecific characteristics of many medical diseases. Subsequent work has led to refined criteria for paranoid and related disorders and has reestablished awareness of less common paranoid presentations such as delusional disorder.

Many clinicians remember being taught that paranoia is so rare that most would not see a single such patient during an entire career. This widespread belief has compromised interest in paranoid disorders. The fact that most persons with delusional disorder live in the community and do not generally seek psychiatric care has made it difficult to carry out systematic case studies. Indeed, knowledge of these conditions has grown slowly. However, case series such as those of Alistair Munro (for delusional disorder, somatic type) or those of Nils Retterstol have been influential in shaping understanding and awareness. What they reveal is that there are persons with these disorders, that the disorders are complex forms of psychiatric illness, and that much remains to be learned.

A major change in the classification of delusional disorders in DSM-III-R and DSM-IV has been to emphasize the central role of delusions in those disorders and to steer away from the vague label of paranoid, which has become synonymous with suspicious and has come to apply largely to a personality disorder. Indeed, suspiciousness occurs in only some of these disorders. The history of the concept of paranoia indicates that lack of clarity in its use is not new. The word paranoia was coined by the ancient Greeks from roots meaning beside and self. Hippocrates applied this term to delirium associated with high fever, but other writers used it to describe demented conditions and madness. It sometimes meant thinking amiss, folly, and the like; hence, its meaning was unclear. For centuries the term fell into disuse until a revival of interest in the nineteenth century.

In 1863 Karl Kahlbaum classified paranoia as a separate mental illness: "a form of partial insanity, which throughout the course of the disease, principally affected the

sphere of the intellect." Influenced by the new scientific methods of empirical medicine, Kahlbaum emphasized the importance of natural history in mental illness and restricted the use of the term paranoia to a persistent delusional illness that remained largely unchanged throughout its course. He noted that delusions could occur in other medical and psychiatric conditions.

Emil Kraepelin found the paranoid concept troublesome and altered his thinking on it with each edition of his influential textbook. His final view advocated three types of paranoid disorder. Like Kahlbaum, Kraepelin based his conclusions on an analysis of the natural history of mental disorders, particularly on outcome, because etiology was obscure. He restricted the definition of paranoia to an uncommon, insidious, chronic illness (he saw 19 cases) characterized by a fixed delusional system, an absence of hallucinations, and a lack of deterioration of the personality. The types of delusions included persecutory, grandiose, jealous, and possibly hypochondriacal. He considered this illness to derive from defects in judgment, a disorder of personality caused by constitutional factors and environmental stress. Paraphrenia was a second paranoid disorder that developed later than dementia precox and was milder.

Hallucinations (auditory in particular) occurred, but there was no mental deterioration (dementia). Finally, there was dementia paranoides, an illness that initially resembled paranoia but had an earlier onset and showed a deteriorating course. Because of this latter feature, Kraepelin considered dementia paranoides a form of dementia precox that arose from disorders of thought, cognition, and emotion. Kurt Mayer's follow-up of Kraepelin's 78 paraphrenia cases challenged the validity of this category because the vast majority of patients showed an outcome indistinguishable from that of dementia precox, casting doubt on the separability of this group. Karl Kolle's follow-up of Kraepelin's paranoia cases indicated some overlap with dementia precox. Kraepelin also emphasized that isolated paranoid symptoms occurred in a variety of psychiatric and medical illnesses.

Eugen Bleuler also recognized paranoia; he broadened its definition to include cases with hallucinations—a paranoid form of dementia precox for which he coined the term schizophrenia—and an intermediate group. However, he thought that the paranoia described by Kraepelin was so rare that it did not warrant a separate classification. Further, he argued that schizophrenic symptoms must be suspected and carefully sought even in those cases. He believed that paraphrenia and intermediate conditions were forms of schizophrenia linked by "much that was identical," and particularly by a common disturbance in associative processes. He also emphasized that paranoid symptoms occurred in other conditions and that to label the symptoms schizophrenic required at least one of the fundamental symptoms: loosened associations, ambivalence, inappropriate affect, and autism. Bleuler's contributions reinforced a trend toward the diagnosis of paranoid illness as a form of schizophrenia. Sigmund Freud used the autobiographical writings of Judge Daniel Schreber to illustrate the role of psychological defense mechanisms in the development of paranoid symptoms. He proposed that Schreber's illness involved a process of denial or contradiction of repressed homosexual impulses toward his father. Persecutory and other delusions result from projecting these denied yearnings onto the environment. Freud did not differentiate subtypes of paranoid disorder, and confused the issue somewhat by proposing that the term paraphrenia be substituted for the term dementia precox or schizophrenia. The major impact of Freud's work was to suggest hypotheses that indicated the relationship between certain delusions and personality.

Ernst Kretschmer's work on the theory of paranoia emphasized that certain sensitive personalities, characterized by depressive, pessimistic, and narcissistic traits,

developed paranoid features acutely when key or precipitating experiences occurred at critical moments in their lives. He observed that these individuals did not develop schizophrenia and had a favorable prognosis. A number of other observations, predominantly but not exclusively emanating from European clinicians (e.g., the American concept of hysterical psychosis), proposed connections between personality and delusion development. Those efforts, based on various theories of the cause of paranoid disturbance, have persisted despite modest empirical support. Out of such work have come terms, such as reactive and psychogenic psychosis, which have figured in various classification schemes, undermining the effort to bring international consistency in definition.

Many barriers remain to international agreement on definition. For example, the term paraphrenia, unlike paranoia, has slipped into near obscurity in North America. In the United Kingdom, however, the diagnosis of late paraphrenia is often used and it is occasionally used in the United States. This term refers to cases of late-onset paranoid symptomatology not characterized by the presence of dementia, confusion, or mood disorder. Interestingly, Kraepelin did not identify a late age of onset in his cases. The potential overlap with late-onset cases of schizophrenia has been a focus of investigation and controversy. With the removal of the DSM-III age criterion for schizophrenia (upper limit of age of onset at 45) in DSM-III-R, cases of late-onset symptoms have tended to be diagnosed as schizophrenia in the United States. Nevertheless, clinical research continues to address the puzzle of whether late-onset cases, despite considerable overlap in clinical features, arise from a variety of causes. Current controversy is based on these historical antecedents and contemporary practices. DSM-III introduced greater rigor in the assessment by requiring clearer criteria boundaries among the varied disorders with delusions. Increased awareness that delusions result from numerous conditions has had a positive influence on the diagnostic process. Yet much of current clinical and research writing on paranoid conditions has characteristically avoided defining the terms paranoid and delusion, apparently because everyone was assumed to know what these terms mean. In popular and literary usage the term paranoid has come to mean insane, angrily suspicious, distrustful, or irrationally irritable. However vague the concept may be, it continues to be used in clinical work. Because it is necessary to differentiate conditions with paranoid features, a useful concept of the term is fundamental. However, the nature and definition of delusions upon which modern psychopathology and psychiatry are built remain unclear.

Shared Psychotic Disorder

Jules Baillarger first described the syndrome in 1860, calling it *folie à communiquer communiquée*, although the first description is commonly attributed to Ernest Charles Lasègue and Jules Falret, who described the condition in 1877 and gave it the name of *folie à deux*. The syndrome has also been called communicated insanity, contagious insanity, infectious insanity, psychosis of association, and double insanity. Marandon de Montyel divided *folie à deux* into three groups (*folie imposee imposée*, *folie simultanee simultanée*, and *folie communiquee communiquée*), and Heinz Lehmann added a fourth group, *folie induite*.

PARANOID CONCEPT

Paranoid signs and symptoms are among the most dramatic and serious disturbances in psychiatry and medicine but the term paranoid refers to a variety of behaviors that may not be psychopathological nor indicative of schizophrenia; hence, the meaning of the term has become obscure. Some clinicians label ordinary suspiciousness paranoid; others restrict use of the term to persecutory delusions; still others apply the term only

to grandiose, litigious, hostile, and jealous behavior, despite the fact that those behaviors may be within the normal spectrum. To make the paranoid concept useful and less vague requires consideration of several points:

1. The term paranoid is a clinical construct used to interpret observations, and in order to apply this construct effectively, the clinician must know its meaning and be able to make accurate observations of potentially paranoid behavior.
2. Use of the term paranoid means the clinician has judged that the person's behavior is psychopathological. This judgment is usually based on the discovery that the person who displays such features is either disturbed or disturbing to others.
3. Although many contributions to understanding paranoid phenomena have focused on conditions in which paranoid features are central (e.g., schizophrenia for Bleuler, paranoia and dementia paranoides for Kraepelin), those features are not necessarily associated with schizophrenia and can appear in other psychiatric and medical disorders. Hence, paranoid features indicate psychopathology, but no specific cause or outcome (Table 13.2–2).

Table 13.2-2. Conditions and Agents Associated With Delusions and Other Paranoid Features

Neurological disorders

Adrenoleukodystrophy
Arteriosclerotic psychoses
Blunt head trauma
Brain tumors
Cerebrovascular disease
Cerebral anoxia
Complex partial seizure disorder
Delirium
Dementia
Fat embolism
Hearing loss
Huntington's disease
Hydrocephalus
Hypertensive encephalopathy
Idiopathic basal ganglia calcification
Idiopathic Parkinson's disease
Intracranial hemorrhage
Marchiafava-Bignami disease
Menzel-type ataxia
Metachromatic leukodystrophy
Migraine
Motor-neuron disease
Multiple sclerosis
Muscular dystrophy
Narcolepsy
Postencephalitic parkinsonism
Presenile dementia (Alzheimer's and Pick's diseases)
Roussy-Levy syndrome
Senile psychoses
Spinocerebellar degeneration
Subarachnoid hemorrhage
Subdural hematoma

Sydenham's chorea
Temporal arteritis
Metabolic and endocrine disorders
Acute intermittent porphyria
Addison's disease
Complication of surgical portacaval anastomosis for cirrhosis
Cushing's syndrome
Folate deficiency
Hemodialysis
Hypercalcemia
Hypoglycemia
Hyponatremia
Hypopituitarism
Liver failure
Malnutrition
Niacin deficiency
Pancreatic encephalopathy
Parathyroid disorders
Pellagra
Pernicious anemia
Phenylketonuria
Systemic lupus erythematosus
Thiamine deficiency
Thyroid disorders
Uremia
Vitamin B12 deficiency
Wilson's disease
Sex chromosome disorders
47 XXY
Klinefelter's syndrome
Turner's syndrome
Infections
Acquired immune deficiency syndrome
Encephalitis lethargica
Creutzfeldt-Jakob disease
Malaria
Syphilis
Toxic shock syndrome
Trypanosomiasis
Typhus
Viral encephalitides
Psychiatric disorders
Brief psychotic disorder
Delusional disorder (including classic paranoia)
Shared psychotic disorder
Mood disorders
Psychotic disorders not otherwise specified
Schizoaffective disorder
Schizophrenia (all types)
Schizophreniform disorder

Alcohol and other substances
 Alcohol withdrawal
 Amphetamine
 Anesthetic nitrous oxide
 Atropine toxicity
 Barbiturate
 Chronic alcohol hallucinosis
 Chronic bromide intoxication
 Cocaine
 Ephedrine
 Marijuana
 Mescaline and other hallucinogens
 Perbitine
 Withdrawal from minor tranquilizers and hypnotic medications
 Toxic agents
 Arsenic
 Carbon monoxide
 Manganese
 Mercury
 Thallium
 Pharmacological agents
 Adrenocorticotrophic hormone
 Amphetamine and related compounds
 Antiparkinson agents
 Anabolic steroids
 Antiarrhythmic drugs
 Antibiotics (cephalosporin, penicillin)
 Anticholinergic drugs
 Antihypertensive agents
 Antimalarials
 Antitubercular drugs
 Bromocriptine
 Bupropion
 Chemotherapeutic agents (asparaginase)
 Cimetidine
 Corticosteroids
 Diphenylhydantoin
 Disulfiram
 Imipramine and other tricyclic drugs
 Levodopa
 Mephentermine
 Methyldopa and imipramine (combination)
 Pentazocine
 Phenylpropanolamine and sympathomimetic agents
 Propylhexedrine

4. The observations that form the basis for judging behavior to be paranoid are of two kinds: subjective (part of the private mental experience of the patient, e.g., a delusion) and objective (observable as a manifest form of behavior, such as litigiousness, guardedness, and grandiosity). Table 13.2–3 is a list of the subjective and objective

features that have traditionally been labeled paranoid and that are frequently found in association; some of these features can be manifestations of normal behavior. The judgment that such features are paranoid may rest on how extreme or inappropriate they are, their presence in combination or association with other behaviors on the list, and the presence of delusions.

Table 13.2-3. Paranoid Features

Objective features

Anger

Critical, accusatory behavior

Defensiveness

Grandiosity or excessive self-importance

Guardedness, evasiveness

Hate

Hostility

Humorlessness

Hypersensitivity

Inordinate attention to small details

Irritability, quick annoyance

Litigiousness (letter writing, complaints, legal action)

Obstinacy

Resentment

Seclusiveness

Self-righteousness

Sullenness

Suspiciousness

Violence, aggressiveness

Subjective features*

Delusions of self-reference, persecution, grandeur, infidelity, love, jealousy, imposture, infestation, disfigurement

Overvalued ideas

* Part of private mental experience. The patient often discloses those features during the clinical interview, but may not do so, even with specific questioning.

5. The term paranoid delusion has traditionally referred to a wide variety of delusions, not simply those of grandeur, persecution, or jealousy. Because of recent confusion that term probably should not be used. The term paranoid and related terms are defined in Table 13.2-4.

Table 13.2-4. Terminology Connected with Paranoia

Delusional disorders DSM-III-R category emphasized that the cardinal feature of these conditions is delusions; DSM-IV criterion is one or more nonbizarre delusions lasting for more than 1 month

Paranoia Old term for an insidiously developed disorder in which persons suffer from an unshakable delusional system but have no disturbance in the clarity or form of their thinking; also known as paranoia vera, simple delusional disorder, delusional monomania

Paranoic or paranoiac Old adjectives used to describe persons with paranoia

Paranoid Broad term meaning suspicious to most people. In psychiatry it is a clinical construct used to describe various objective and subjective features of behavior deemed to be psychopathological; refers to no specific condition (e.g., to be paranoid does not mean that schizophrenia is present)

Paranoid delusion Older term used to refer to persecutory and grandiose delusions because of their occurrence in the paranoid subtype of schizophrenia; this term has suffered from the confusion associated with the paranoid concept; DSM-III-R recommended that it no longer be used

Paranoid disorders DSM-III term for an idiopathic group of conditions including paranoia, acute paranoid disorder, shared paranoid disorder, and atypical paranoid disorder; no longer used

Paranoid personality Enduring traits of paranoid behavior not due to schizophrenia or other mental disorder; generally, there is no evidence of delusions or other features of psychosis

Paranoid syndrome Term applied to constellations of paranoid features that occur together and can arise from multiple sources including depression, general medical conditions, substance-induced disorders, and schizophrenia

Paraphrenia Old term for conditions lying theoretically between schizophrenia and paranoia and sharing features of both (hallucinations but no deterioration). It, too remains controversial and probably should not be used until research validates its meaning. In use in U.K. Designate patients with late-onset paranoid features without confusion, dementia, or mood disorder

Delusions

When Karl Jaspers formulated the concept of delusion widely used today, he suggested three criteria: (1) subjective certainty, (2) incorrigibility, and (3) falsity of content. He viewed these criteria as tentative, preferring to consider them as approximations to a definition in that they provided practical suggestions for detecting delusions rather than actually defining them. This and later contributions emphasized a certain humility about the delusion concept that has not been sustained in contemporary formulations of this psychopathological feature. Numerous, often ignored, problems compromise the clinical research utility of the delusion concept. For example, according to DSM-III-R and DSM-IV, delusion is "an incorrect inference about external reality." This definition has certain implicit and complicating features: (1) there is a process of inference separable from the belief that the process produces, (2) this same process is used by normal persons to generate beliefs about the world, and (3) this process is impaired in delusional patients. As pointed out later in this chapter, the validity of the latter two assertions is questionable. Also, central to the concept of delusional disorder is the distinction between bizarre (impossible) and nonbizarre (possible) delusions. This distinction has been difficult to apply reliably in clinical assessment, yet on it rests considerable weight in making the diagnosis of delusional disorder. Further examples of difficulties concerning the definition of delusion have been discussed by Manfred Spitzer, who has traced the movement from philosophy to empirical science in the evolution of the definition of delusion. Awareness of the vagaries and imprecise nature of the definition of delusion is essential to clinical and research efforts.

Since the early nineteenth century delusions have been classified by content or theme. Other descriptive dimensions have gained acceptance through clinical use and some empirical research: understandability, degree of certainty, systemization, complexity, relevance to patient's life, plausibility, onset, associated psychopathology, and time course. These features are used to grasp the nature of the delusional experience, translate clinical observations into diagnostic and treatment interventions, and design research.

In clinical encounters delusions are usually easy to detect. Certain features (Table 13.2–3) of the patient's behavior may suggest the presence of delusions or help confirm the impression that the beliefs are delusional. In subtle cases, however, this task is more challenging. The clinician must make a judgment, based on the behavior and reported private mental experience of the patient, of whether or not delusional beliefs are present. Attempts to present counterevidence and argument may be useful to determine whether the patient's views can be influenced by evidence that is usually sufficient to alter the belief of a normal person. This judgment often depends on deciding whether a threshold indicative of psychopathological disorder has been passed, possibly reflected in the inappropriateness or extreme nature of the patient's behavior, rather than the simple truth or falsity of the belief. In practice, the only effective approach to assessing delusions is to put together as comprehensive a picture as possible regarding the nature of the patient's condition. Lacking laboratory tests for delusions, clinical judgment will be required to some degree in virtually all cases. At the theoretical level, the definition of delusions is moving gradually away from its roots in philosophy and phenomenological description toward a more empirically derived set of features. This process will take considerable time to achieve a satisfactory resolution of the many issues plaguing this aspect of psychopathology.

COMPARATIVE NOSOLOGY

Certain advances have been made in the nosology of delusional disorders, but the variety of current definitions illustrates that consensus has not yet been achieved. The reasons for such differences are multiple: the principal reason is the lack of relevant data—delusional disorders occur infrequently. Typically, patients continue to function and live in the community without ever seeking clinical intervention. When they do, the condition is easily misdiagnosed because patients may have minimal overt identifying characteristics. Limited knowledge, based largely on case reports, exists; systematic, larger-scale studies are uncommon. Most of these studies are European and have employed varied classifications. Also, the fundamental concept that these disorders are distinct from schizophrenia and mood disorders has until recently been unrecognized by many psychiatrists.

Kahlbaum was the first to use the term paranoia to designate a diagnostically separate group of disorders. Kraepelin developed this diagnostic concept further by emphasizing the chronic and unremitting nature of paranoia and the lack of other features such as hallucinations that distinguished it from schizophrenia. In 1952 the first edition of DSM (DSM-I) defined paranoid reactions as conditions in which there are persecutory or grandiose delusions, with emotional responses and behavior consistent with the delusions, but generally lacking hallucinations. The subtypes were paranoia (a chronic disorder with systematized delusions) and paranoid state (a more acute, less persistent condition with less systematized delusions). In 1968 DSM-II largely preserved these concepts.

DSM-III

Although new definitions were established in DSM-III in 1980, earlier concepts are still evident. The essential features of paranoid disorders according to DSM-III were persistent persecutory delusions or delusional jealousy not due to any other mental disorder. Included in the group of paranoid disorders were paranoia, shared paranoid disorder, acute paranoid disorder, and a residual category called atypical paranoid disorder. The boundaries between these conditions and other disorders, such as paranoid personality disorder or paranoid schizophrenia, were noted to be vague. Different types of paranoid disorders were classified on the basis of chronicity. The criteria narrowed the bounds of previous classifications by not including cases with

marked hallucinations or certain delusions (e.g., hypochondriacal, erotomanic, and others).

DSM-III-R

In 1987 DSM-III-R simplified the DSM-III definition, attempted to minimize the confusion associated with the term paranoid, and highlighted the view that the formation of delusions in the absence of schizophrenia, mood disorder, or organic disorder is the essential feature of these conditions. In contrast to DSM-III, diagnosis in DSM-III-R and DSM IV requires a month-long duration of symptoms. Subtyping is based on the predominant type of delusion, which is specified (such as jealous, erotomanic, or somatic). This latter feature broadens the category to include a variety of unusual delusions as well as the more common persecutory type. In many respects these criteria are virtually identical to Kraepelin's definition of paranoia. The two exceptions were Kraepelin's reluctance to endorse a subtype of somatic or hypochondriacal paranoia or to permit cases with hallucinations to fall within this diagnosis. Kraepelin believed that cases with hypochondriacal delusions rarely occurred in this pattern.

Shared paranoid disorder was renamed induced psychotic disorder in DSM-III-R and was placed in the category of psychotic disorders not elsewhere classified, along with schizophreniform, schizoaffective disorders, and brief reactive psychosis. This represents a fundamental departure from DSM-III, which classified this disorder among the paranoid disorders. The delusional content of patients with this disorder may concern not only persecution or jealousy but virtually any form of delusion, hence the change in terminology. The term induced may better describe the nature of the condition, but hardly resolves the puzzle of causation.

DSM-IV

In 1994 a revised classification made modest changes in the DSM-III-R criteria in an attempt to refine the definition of delusional disorders. In DSM-III-R the distinction between schizophrenia and delusional disorders had been unclear and controversial. In DSM-III-R this boundary was defined by the nonbizarre qualities of delusions in delusional disorder and the absence of other active-phase symptoms of schizophrenia. Also important was the required absence of other odd or bizarre behavior apart from the delusion. Because the distinction between bizarre and nonbizarre is difficult to define and therefore to apply reliably, other terms such as systematized and prominent were suggested. In practice, however, those terms also have limitations. This problem has helped to promote the case for modifying the criteria in another way: specifically, to use the level of impaired functioning as a means of characterizing the distinction between schizophrenia (considerable impairment) and delusional disorders (relatively less impairment). However, the variability of outcomes in both disorders undermines this strategy somewhat. DSM-IV suggests that when poor functioning occurs in delusional disorder, it is the result of the delusional beliefs themselves. For example, a person quits a job because he or she believes that the fumes in the workplace are causing a cancerous growth. That person's financial situation worsens and preoccupation with repeated medical consultations enhances a downward spiral. In contrast, poor functioning in schizophrenia usually results from cognitive compromise and positive and negative symptoms, especially avolition. The resolution of how to make modifications, however, depends on the effectiveness of the criteria in defining homogeneous and valid subsets of psychotic disordered patients. For this purpose, field trials and data analyses have been used to inform the decision scientifically. Although the DSM-IV criteria reflect progress, their validity remains only partly established.

Another unsettled issue that DSM-IV attempts to resolve is the classification of delusional variants of somatoform disorder, specifically body dysmorphic disorder. In this condition the patient suffers from preoccupation with imagined or slight defects in appearance (such as skin blemishes, the size of one or more body parts); it is accompanied by impairment in social and occupational functioning, shame, and repetitive, often ritualistic behaviors. These may include skin picking, mirror checking, requests for reassurance, and attempts to camouflage the supposed deformity. In some cases, the preoccupation appears to be delusional. However, the relationship between nondelusional and delusional variants is unclear; whether the disorders are distinct or overlapping remains unknown. DSM-IV permits dual diagnosis of body dysmorphic disorder and delusional disorder when a delusional belief is present in the former condition. This resolution, in which the same symptoms are given two diagnoses, accurately reflects the available research data on the relationship of these two disorders and also underlines the need for further research to clarify these distinctions. A similar problem arises with respect to delusional variants of hypochondriasis and of obsessive-compulsive disorder, and a similar solution is applied: obsessive-compulsive disorder patients may also be diagnosed as delusional disorder.

Shared Psychotic Disorder

DSM-IV renamed the DSM-III-R category induced psychotic disorder, calling it shared psychotic disorder. This change reflects the attempt to avoid the term paranoid and to identify the condition without reference to any presumed cause or mechanism. The goal is to define the boundaries between this condition and more common ones, such as other psychotic disorders, mood disorders with psychotic features, substance-induced psychotic disorders, and psychotic disorders due to a general medical condition.

ICD

The ninth revision of International Statistical Classification of Diseases (ICD-9) contained more categories for paranoid disorder than the American schemes. Most paranoid disorders fall under the rubric paranoid state, including simple paranoid state, paranoia, paraphrenia, and induced psychosis. Additional subcategories include other and unspecified paranoid states. Acute paranoid reactions and psychogenic paranoid psychosis are classified separately. DSM-III, DSM-III-R, and DSM-IV generally reflect an atheoretical position with respect to the causes of these disorders whereas ICD-9 was less neutral. For example, psychogenic paranoid psychosis implies a kind of causal mechanism. The categories of paranoid disorder according to these classifications are summarized in Tables 13.2–5.

Scroll right to see more columns.

Table 13.2-5. Comparative Nosology of Delusional Disorder

ICD-9 (1979)	DSM-III (1980)	DSM-III-R (1987)	ICD-10 (1993)	DSM-IV (1994)
Paranoid state, simple		Delusional disorder		
Paranoia	Paranoia disorder	Delusional (paranoid) disorder	Delusional disorder	Delusional disorder
Paraphrenia (involuntary paranoid state, late paraphrenia)			Delusional disorder	
Induced psychosis	Shared psychotic (folie à deux, induced)	Shared paranoid disorder	Induced psychotic disorder	Induced delusional disorder

paranoid disorder)		
Other specified states (paranoia querulans, Sensitiver Beziehungswahn)	Delusional disorder	
Unspecified paranoid states disorder	Atypical paranoid disorder, unspecified	Persistent delusional
Acute paranoid reaction (bouffée bouffée delirante délirante)	Acute paranoid disorder	Paranoid reaction
Psychogenic paranoid psychosis (protracted reactive paranoid psychosis)		

ICD-10 pays more attention to creating classifications similar to DSM-III-R and DSM-IV. Paraphrenia, for example, is subsumed under persistent delusional disorder but delusions must be present for about 3 months in order to diagnose delusional disorder. The subtypes of the disorder overlap with DSM-IV subtypes. For those conditions of less duration, acute and transient psychotic disorder is diagnosed. Induced (shared) delusional disorder is considered a separate designation with a phenomenology similar to persistent delusional disorder.

EPIDEMIOLOGY

Delusional disorder has been considered an uncommon if not rare condition from its earliest descriptions even though epidemiological information is meager. Recent demographic evidence covering a period from 1912 to the 1970s provides an estimate of incidence, prevalence, and related statistics (Table 13.2–6). However, this evidence was assembled using definitions that are not the same as those of DSM-III, DSM-III-R, or DSM-IV. Subsequent data will in all likelihood be somewhat different using the newer criteria. It is clear that the estimates are merely indications, but can be useful guidelines to future appraisals.

Table 13.2-6. Epidemiological Features of Delusional Disorder

Incidence*	0.7–3.0
Prevalence*	24–30
Age at onset (range)	18–80 (mean 34–45 years)
Type of onset	Acute or gradual
Sex ratio	Somewhat more frequently female
Prognosis	Best with early, acute onset
Associated features	Widowhood, celibacy often present; history of substance abuse, head injury not infrequent

Portions of the table adapted from K S Kendler: Demography of paranoid psychosis (delusional disorder). Arch Gen Psychiatry 39: 890, 1982.

* Incidence and prevalence figures represent cases per 100,000 population.

However, certain features of the data are remarkable. For example, the stability of estimated incidence has been striking over extended periods of time in this century. The prevalence of these disorders substantiates the widely held clinical impression that they are uncommon conditions (compared with mood disorders and schizophrenia) but are not rare. Some studies indicate that delusional disorder accounts for a surprising 2 to 8 percent of inpatient psychiatric admission for

"functional psychosis." Patients with delusional disorders are somewhat more likely to be women (but this is an inconsistent feature), and to be more socially and educationally disadvantaged as compared to patients with mood disorders. Women tend to be older than men at the time of diagnosis. While the onset age range is wide (18 to 80), most patients are middle-aged. There is suggestive evidence that immigrant status, celibacy among men, and widowhood among women are associated with delusional disorder but all such observations need to be unambiguously replicated.

ETIOLOGY

The cause of delusional disorder is unknown. The epidemiological and clinical literature suggests that certain risk factors may be relevant to etiology and deserve further research elaboration. These risk factors are found in Table 13.2–7. Whether they are risk predictors or simply characteristics or markers of the disorder is unknown. Familial psychiatric disorder, including delusional disorder, is the best documented risk factor at present.

Table 13.2-7. Risk Factors Associated With Delusional Disorder

Advanced age

Sensory impairment/isolation

Family history

Social isolation

Personality features (e.g., unusual interpersonal sensitivity)

Recent immigration

Genetic or family studies that have begun to appear in the literature indicate the possible specific family transmission of delusional disorder. A recent study of genetic variation in deoxyribonucleic acid (DNA) sequence coding for dopamine type 4 (D4) receptor proteins strongly suggests the involvement of the relevant gene in conferring susceptibility to delusional disorder. The comparison subjects either had schizophrenia or were normal controls.

Paranoid features, including the types of delusions encountered in these disorders, occur in a large and growing number of conditions (Table 13.2–2). Differences in classifying idiopathic delusional disorder add to the problems of understanding causation. Theories and explanations of delusions abound in the literature but empirical evidence to support those theories is limited. With so many uncertainties, any conclusions concerning the cause of delusional disorder must be made cautiously. Delusional disorder is an uncommon, probably heterogeneous, group of illnesses whose validity has been questioned since Kahlbaum published his views. The major phenomenological feature of these conditions is the formation and persistence of delusions. It is well known that delusions occur in a variety of psychiatric and medical conditions, and that the pathogenesis of delusions is not fully understood. Hence, discussion of etiology in the delusional disorders can proceed along two lines: (1) determining the distinctiveness of the category itself, (2) examining the theories proposed to account for the pathogenesis of delusion formation per se, and (3) integrating the available evidence into testable proposals.

Distinctiveness of Delusional Disorder

An issue that is central to attributing causation is whether delusional disorder represents a separate group of conditions or is an atypical form of schizophrenic and mood disorders. The relevant data come from a limited number of studies and is inconclusive. Epidemiological data suggest that delusional disorder is a separate condition; it is far less prevalent than schizophrenic or mood disorders; age of onset is

later than in schizophrenia although men tend to experience the illness at earlier ages than women; and the sex ratio is different from that of mood disorder, which occurs disproportionately among women. Findings from family or genetic studies also support the theory that delusional disorder is a distinct entity. If delusional disorder is simply an unusual form of schizophrenic or mood disorders, the incidence of these latter conditions in family studies of delusional disorder patient probands should be higher than that of the general population. However, this has not been a consistent finding. A recent study concluded that patients with delusional disorder are more likely to have family members who show suspiciousness, jealousy, secretiveness, even paranoid illness, than families of controls. Other investigative efforts have found paranoid personality disorder and avoidant personality disorder to be more common in the relatives of patients with delusional disorder than in the relatives of controls or of schizophrenic patients. A recent study documented modest evidence for an increased risk of alcoholism among the relatives of patients with delusional disorder as compared to probands with schizophrenia, probands with psychotic disorder not otherwise specified, and probands with schizophreniform disorder.

Investigations into patient's natural history also lend support to the suggestion that delusional disorder is a distinct category: age of onset appears to be later than in schizophrenia and outcome generally is better for delusional disorder patients than for schizophrenia patients. Although fraught with methodological shortcomings, premorbid personality data indicate that schizophrenia patients and patients with delusional disorder differ early in life. The former are more likely to be introverted, schizoid, and submissive; the latter extroverted, dominant, and hypersensitive. Delusional disorder patients may have below-average intelligence. Precipitating factors, especially related to social isolation, conflicts of conscience, and immigration, are more closely associated to delusional disorder than schizophrenia. These characteristics support Kraepelin's view that environmental factors may play an important etiological role. Clinical characteristics such as greater intensity of delusions, uncommon occurrence of negative symptoms, and possible association with cerebrovascular disorder in late-onset cases also suggest differences from late-onset schizophrenia. Recent observations of successful treatment with pimozide (Orap) in several subtypes of delusional disorders suggest the possibility of a common pathogenetic mechanism in these disorders. Follow-up studies indicate that the diagnosis of delusional disorder remains fairly stable: only a small proportion of cases (3 to 22 percent) are diagnosed later as having schizophrenia, and even fewer (6 percent) are diagnosed later as having a mood disorder. Outcome in terms of hospitalization and occupational adjustment is markedly more favorable for delusional disorder than for schizophrenia. When social or occupational functioning is poor in delusional disorder, it generally occurs as the result of the delusional beliefs themselves, not because of cognitive impairment or negative symptoms.

The evidence argues in favor of the distinctiveness of delusional disorder, but it is likely that at least some patients diagnosed as having delusional disorder will develop schizophrenia or mood disorders. Hence, current clinical criteria have limitations and need improvement, which may be possible with the use of laboratory techniques or more specified clinical definitions. Furthermore, the data suggest that delusional disorder is relatively chronic and is probably biologically distinct from other psychotic disorders.

PATHOGENESIS

Although a clear understanding of the pathogenesis of delusions remains an unfulfilled hope, several major theories have been advanced. Any adequate hypothesis

for delusion formation must deal with certain facts: (1) delusions occur in a variety of medical and psychiatric diseases; (2) not all persons with such conditions develop delusions; (3) the types of delusions are relatively few and strikingly repetitious despite the variety of diseases; (4) delusions can clear rapidly with treatment of the underlying condition or its termination; (5) delusions can persist, and even become systematized; (6) delusions often accompany perceptual changes such as hallucinations or impaired sensory input; (7) delusions may be highly encapsulated features in persons such that their functioning may not be compromised socially, intellectually, or emotionally. Also, any adequate hypothesis must respond to two questions. First, why does the patient have a delusion? This is a question concerning the form of the psychopathology. Second, why does the patient have this particular delusion? This is a question concerning the content of the psychopathology.

There are three categories of theory in delusion formation.

1. Delusions arise in an otherwise intact cognitive system because a deviant pattern of motivational interest is present (psychodynamic mechanism, social attribution theory).
2. Delusions arise as the result of a fundamental cognitive defect that impairs the patient's capacity to draw valid conclusions from evidence (disorder of reasoning).
3. Delusions arise from normal cognitive processes directed at explaining abnormal perceptual experiences (psychobiological mechanism, anomalous experience hypothesis).

These theories need not be mutually exclusive. Delusional beliefs probably are the result of different processes involving one or more of the proposed mechanisms.

Psychodynamic Mechanism In 1911 Freud published "Psychoanalytic Notes Upon an Autobiographical Account of a Case of Paranoia (Dementia Paranoides)." His interpretation of this case, which became the foundation of the psychodynamic theory of paranoia, was based on his reading of the memoirs of the presiding judge of a Dresden appeals court, Daniel Paul Schreber, who had suffered episodes of psychiatric illness in 1884, 1885, and 1893. The second episode led to two prolonged hospitalizations from which the patient obtained discharge in 1902 following legal action, although he was still delusional. Freud asserted that Schreber's 1903 account, *Memoirs of My Nervous Illness*, offered a legitimate basis for theory, as "paranoiacs cannot be compelled to overcome their internal resistances, and $\frac{1}{4}$ in any case they only say what they choose to say $\frac{1}{4}$." Freud argued that the written case report can take the place of personal acquaintance, and in the case of Schreber, Freud never saw the patient. Freud asserted that Schreber's case illustrated a general mechanism of delusion formation involving denial or contradiction and projection of repressed homosexual impulses that break out from the unconscious. The forms of delusion in paranoia can be represented as contradictions of the proposition "I (a man) love him (a man)." The following examples illustrate the forms of illogic.

1. Delusion of persecution. In the contradiction "I do not love him, I hate him," a hatred that persons deem unacceptable at the conscious level is transformed and becomes instead: "He hates (elaborated to "persecutes") me." Patients can then rationalize their anger by consciously hating those persons whom they perceive to hate them.
2. Delusion of erotomania. The proposition "I do not love him—I love her" is transformed through projection to "She loves me—and so I love her."
3. Delusional jealousy. To protect against unwarranted, threatening impulses the patient transforms the proposition in this manner: "I do not love him—she (a wife, lover) loves him." Hence, jealous delusions represent the transformed attractions of the deluded for the lover.

4. Delusion of grandiosity (megalomania). Here the contradiction made is, "I do not love him, I love myself."

The essence of the theory is that delusions represent attempts to manage the stirrings of unconscious homosexuality. According to the classic theory, the dynamics of unconscious homosexuality are similar for female as well as male patients.

Comment

Many theorists have added to the psychodynamic lore on delusion formation from the standpoint of understanding personality factors. For example, some of the vulnerability to delusion formation may be related to deficiently developed trust, narcissistic dynamics, or exaggerated traits such as hypersensitivity.

Critique

Freud's mechanism of delusions sidesteps the distinction between form and content in psychopathology. He proposes an inferential process to account for the particular delusion but does not clearly address the issue of why a delusion is formed rather than another symptom, such as hallucination. Verification of the hypothesized mechanism clearly rests on finding evidence that delusions are associated with indications of homosexual tendencies. The theory has been perpetuated in part because an absence of homosexuality can never be proved, and such tendencies can be used as a pillar, even if not a scientifically or empirically demonstrable pillar, in the psychodynamic argument. The few experimental attempts made to test the hypothesis have been inconclusive or equivocal. Although homosexual concerns have been found among some delusional patients, the variety of conditions with similar delusions argues against a common mechanism of unconscious homosexuality in all. Persons who delusional patients say are persecuting them are not always known by them, nor is the persistence of such delusions adequately accounted for in that formulation.

Nevertheless, the classic approach has had immense influence and has provided important psychoanalytical concepts, such as projection, and an awareness that developmental experiences may operate to influence the content of delusional thinking. Systematic empirical study would be valuable.

Disordered Reasoning Because the definition of delusion (Table 13.2–1) emphasizes the operation of reasoning processes that have gone haywire, it is not surprising that a number of attempts have been made to establish that disorder of reasoning is related to delusion formation and that such disorders can be observed among deluded patients. Related to the psychodynamic formulation is the proposal that delusions arise on the basis of defects in formal logical reasoning. Popular in the 1950s and 1960s, this view, promulgated by Eilhard von Domarus among others, suggested that errors in logic such as the principle of identity (two subjects are identical on the grounds of identical predicates) have an etiological role. For example, "Charles Manson used drugs; I use drugs, therefore I am Charles Manson." The empirical assessment of that proposal has failed to establish that deluded patients exhibit more defects in reasoning; rather it appears that normal and deluded persons both make similar errors of reasoning with comparable frequencies.

Two other proposals involving disturbance in reasoning have been studied recently. The first portrays the difficulty underlying delusion formation as a failure in the application of Bayesian reasoning. According to this model of developing beliefs, making choices, and drawing conclusions, delusional patients accept conclusions at levels of probability too low for acceptance by nondelusional persons. However, attempts to demonstrate that failure have had equivocal results. The second proposal suggests that the reasoning processes of delusional patients are influenced by the subject's tendency to assign meaning in a biased manner. The bias arises in making

judgments about one's own behavior and that of another person by assigning motives and characteristics to the person involved. Application of this model reflecting motivational and reasoning difficulties (based on social attribution theory) has been tested, but the results do not provide sound support for the formulation.

Other Psychological Mechanisms In Manic Depressive Insanity and Paranoia, Kraepelin considered the delusions of paranoia to be the "morbidly transformed expression of the natural emotions of the human heart" and, more specifically, "a kind of psychological compensation for the disappointments of life." He dismissed the Freudian psychodynamic mechanism on the grounds that it did not refer to a clear concept of paranoia and that it was not supported by evidence. He also emphasized constitutional factors, especially disturbances of judgment, in his formulation. Other authors have made similar suggestions about the role of need fulfillment in the development of paranoia. For example, delusions of persecution might serve to maintain the self-esteem of the deluded person, according to a social attribution view about delusion formation in which a normal bias—that of assigning blame for negative outcomes to other persons or circumstances—is exaggerated.

Critique

These contributions do not address the issue of pathogenesis rigorously. They explain the content of the delusion but not its form. The commonness of the risk factors or antecedent features cited repeatedly as central to delusion formation contrasts dramatically with the uncommonness of delusional disorder.

Psychobiological Mechanisms The French neurologist Ga'tan G. de Clerambault proposed in 1942 that chronic delusions resulted from abnormal neurological events. Infections, lesions, intoxication, and other forms of damage produce automatisms that puzzle or distress the patient initially and eventually demand explanation. The explanations take the form of delusions. Automatisms include hallucinations, a constant parade of memories, feelings of familiarity, false recognition, arresting of thought, disturbances in attention, bizarre tactile sensations, and even kinesthetic sensation. The view that delusions are an explanation for hallucinations is an old concept in psychiatry that has not been well formulated. The fact that hallucinations have been introduced into and retracted from the definition of paranoia over the years also reflects a lack of clarity regarding a possible connection between the two forms of psychopathology.

In 1974 Brendan Maher proposed a similar hypothesis that conceptualized delusions as explanations of anomalous experiences that arise in the environment, the peripheral sensory system, or the central nervous system. A central tenet of his view is that the processes whereby delusional beliefs are formed are similar in their essential nature to those that operate in the formation of normal beliefs and even of scientific hypotheses. Integral to the hypothesis is the assumption that components of this normal operational sequence have a neural substrate that may be activated either by sensory input (as in the hallucinatory effects of drugs) or by the effects of brain damage (as in alcoholism). The activation of any part of the sequence demands explanation and may thus give rise to delusions. The sequence, activated by disturbances in sensory experience, emotional incongruity, or central nervous system abnormalities, has the following stages: (1) anomalous experience, (2) feelings of significance, (3) testing for reality of experience, (4) developing tentative hypotheses, (5) additional observation, (6) exploring insights, and (7) confirmation of the insight by selective observation. In Maher's explanation, the patient is delusional because he or she actually experiences anomalies that demand explanation. The particular content of the delusion is drawn from the past or current circumstances, experience, and the personal

and cultural background of the patient. The explanation answers questions such as the following: What is happening? Why? Why do other people deny it is happening? Why is it only happening to me? Who is responsible for it? The delusional explanation offers relief from puzzlement, and that relief works against abandonment of the explanation.

Critique

The psychobiological formulation has gone largely unstudied, but there is supporting evidence in the form of studies of altered perception among patients and healthy controls experiencing sensory impairment or sensory deprivation, and among persons taking various drugs of abuse. These studies have demonstrated a high incidence of delusion formation. The failure to detect a fundamental defect in the cognitive process of delusional patients or to identify basic differences in belief formation between persons with delusions and normal controls provides indirect support as well. Indeed, delusions are formed in persons with a range of levels of intelligence and education, further supporting the view that a disturbance in the cognitive processes is not the source of the problem. Also a number of medical conditions show evidence of delusions but no history of cognitive impairment. Clearly, this hypothesis warrants further examination, and it remains to be seen how applicable it is to conditions, such as delusional disorder, in which the occurrence of hallucinations is debated. Sensory impairment and central nervous dysfunction, although apparently likely, have not been firmly established for the disorder. The anomalous experience hypothesis focuses on the psychological mechanisms underlying delusion formation, but a complementary proposal concerns the anatomic loci associated with delusional thinking. Jeffrey Cummings and others have used the growing data on the psychopathological consequences of neurological disease to suggest that delusions occur in diseases involving the limbic system—in particular, temporal lobe structures and caudate nuclei. Diseases characterized by excessive dopaminergic activity or reduced cholinergic activity also carry a heightened risk of delusion formation. Cummings further hypothesizes that the common locus of delusion formation is limbic dysfunction that predisposes the individual to misinterpretation of the environment accompanied by inappropriate perception of threat. Both disease- and patient-related factors influence the content, complexity, and timing of the delusion.

PATHOGENESIS

Although limited by the sparseness of research in the area, observations of pharmacological treatment provide complementary insights into the pathogenetic puzzle. Data from treatment reports on delusional disorder suggest that pimozide (Orap) a highly specific dopamine-blocking agent, has greater effectiveness than typical antipsychotic drugs in this condition; some data even suggest that it has a unique role. There are several pharmacological effects of pimozide, in addition to dopamine receptor blockade, that may help explain its effectiveness: (1) relative lack of nonadrenergic blocking action (2) calcium channel antagonism, and (3) opioid receptor blockade. The effect of opiate receptor blockade has been proposed as relevant to reported specific effectiveness in delusional infestation partly based on observations of opiate receptor blocking interventions in delusional disorder somatic type, with delusions of infestation. Intravenous administration of the opioid agonist fentanyl (sublimaze) led to intensified cutaneous sensations whereas administration of naloxone (Narcan) an opioid antagonist, resulted in complete remission of the patient's cutaneous sensation. That pimozide is especially effective in delusional disorder, somatic type, supports the notion that its opiate receptor antagonism blocks

central recognition of abnormal peripheral sensation; such a view is consistent with the anomalous experience hypothesis.

Other Relevant Factors Delusions have been linked to a variety of additional factors such as social and sensory isolation, socioeconomic deprivation, and personality disturbance. The deaf, the visually impaired, and possibly immigrants with limited ability in a new language may be more vulnerable to delusion formation than the normal population. Vulnerability is heightened with advanced age. Delusional disturbance and other paranoid features are common in the elderly. In short, multiple factors are associated with the formation of delusions, and the source and pathogenesis of delusional disorders per se have yet to be specified.

Integration The pathogenesis of delusions in general and delusional disorder in particular remains a field of hypotheses with little firm grounding. A variety of theories exist, but empirical support for any theory is markedly limited. Of those available, however, the anomalous experience hypothesis appears to be the best supported and certainly is the most consistent with research findings from other domains. Given the research explosion in neuroscience and psychopathology, this hypothesis should be explored as fully as possible. In delusional disorder, for example, the anomalous experience hypothesis needs to be further specified, for example, on what kinds of anomalous experience could lead to the jealousy delusion, the erotomanic delusion, and so forth. Studies now under way in the misidentification delusions, such as the Capgras's syndrome, provide a model for how such research might be directed. Progress may result from further studies of the neurobiology underlying successful treatment strategies in delusional disorder as well.

DIAGNOSIS AND CLINICAL FEATURES

Delusional Disorder

Diagnosing delusional disorders requires that the clinician match the features of the case to the appropriate criteria (Table 13.2–8). When the clinician has successfully ruled out other disorders, certain features of the case can help to substantiate the diagnosis of delusional disorder. The ICD-10 criteria for delusional disorder are listed in Table 13.2–9.

Table 13.2-8. DSM-IV Diagnostic Criteria for Delusional Disorder

- A. Nonbizarre delusions (i.e., involving situations that occur in real life, such as being followed, poisoned, infected, loved at a distance, or deceived by spouse or lover, or having a disease) of at least 1 month's duration.
- B. Criterion A for schizophrenia has never been met. Note: Tactile and olfactory hallucinations may be present in delusional disorder if they are related to the delusional theme.
- C. Apart from the impact of the delusion(s), or its ramifications, functioning is not markedly impaired and behavior is not obviously odd or bizarre.
- D. If mood episodes have occurred concurrently with delusions, their total duration has been brief relative to the duration of the delusional periods.
- E. The disturbance is not due to the direct physiological effects of a substance (e.g., a drug of abuse, a medication) or a general medical condition.

Specify type (the following types are assigned based on the predominant delusional theme):

Erotomanic type: delusions that another person, usually of higher status, is in love with the individual

Grandiose type: delusions of inflated worth, power, knowledge, identity, or special relationship to a deity or famous person

Jealous type: delusions that the individual's sexual partner is unfaithful

Persecutory type: delusions that the person (or someone to whom the person is close) is being malevolently treated in some way

Somatic type: delusions that the person has some physical defect or general medical condition

Mixed type: delusions characteristic of more than one of the above types but no one theme predominates

Unspecified type

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Table 13.2-9. ICD-10 Diagnostic Criteria for Delusional Disorders

Delusional disorder

A. A delusion or a set of related delusions, other than those listed as typically schizophrenic in criterion G1(1)b or d for paranoid, hebephrenic, or catatonic schizophrenia (i.e., other than completely impossible or culturally inappropriate), must be present. The commonest examples are persecutory, grandiose, hypochondriacal, jealous (zelotypic), or erotic delusions.

B. The delusion(s) in criterion A must be present for at least 3 months.

C. The general criteria for schizophrenia are not fulfilled.

D. There must be no persistent hallucinations in any modality (but there may be transitory or occasional auditory hallucinations that are not in the third person or giving a running commentary).

E. Depressive symptoms (or even a depressive episode) may be present intermittently, provided that the delusions persist at times when there is no disturbance of mood.

F. Most commonly used exclusion clause. There must be no evidence of primary or secondary organic mental disorder as listed under organic, including symptomatic, mental disorders, or of a psychotic disorder due to psychoactive substance use.

Specification for possible subtypes

The following types may be specified if desired: persecutory; litigious; self-referential; grandiose; hypochondriacal (somatic); jealous; erotomanic.

Other persistent delusional disorders

This is a residual category for persistent delusional disorders that do not meet the criteria for delusional disorder. Disorders in which delusions are accompanied by persistent hallucinatory voices or by schizophrenic symptoms that are insufficient to meet criteria for schizophrenia should be coded here. Delusional disorders that have lasted for less than 3 months should, however, be coded, at least temporarily, under acute and transient psychotic disorders.

Persistent delusional disorder, unspecified

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DSM-IV defines the core psychopathological feature of delusional disorder as persistent, nonbizarre delusions not explained by other psychotic disorders. Onset can be sudden, following a precipitating event that the patient often reports, or the disorder may emerge gradually and may become chronic. Behavioral and emotional responses are generally appropriate: neither a mood disorder nor the volitional, thinking, and emotional disturbances of schizophrenia are present (including hallucinations, which are quite restricted in delusional disorder). In general, patients

with delusional disorder show little disorganization or impairment in their behavior or in the clarity of their thinking.

The delusions are unusual yet they refer to aspects of life that might occur, such as being conspired against, cheated on, physically ill, in love, jealous, and so forth. They are, as George Winokur has suggested, "possible," rather than totally incredible and bizarre as are many of the delusions of schizophrenia. Delusions are categorized according to their content; the most common are characterized by persecution, disease, and jealousy. The delusions are fixed (persistent) and unarguable. Patients interpret facts to fit the delusion rather than modifying the delusion to fit the facts. There is systematization in the delusional thinking, meaning that a single theme or series of connected themes is present with links to the predominant delusion. Normal life and functioning gradually give way to the dominance of delusional concerns. Many have proposed that there is a descriptive continuum between paranoid personality disorder, delusional disorder, and the paranoid subtype of schizophrenia in terms of degrees of disorganization and impairment. However, there is little evidence to support the concept that these disorders share more than overlapping psychopathology.

The presence of hallucinations in delusional disorder has been debated, with some theorists arguing that schizophrenia is a more likely diagnosis in such cases and others not, so long as the hallucinations are not marked and persistent. The resolution of this issue remains distant, but it is reasonable to consider infrequent, poorly organized, and simple hallucinations that are not a prominent part of the psychopathology to be a feature of delusional disorder. The hallucinations are usually auditory but may be visual and tend to be more common in acute cases. Other types of hallucinations may occur; however, tactile or olfactory hallucinations may be present and may even be prominent if they are related to the delusional theme.

The person's emotional contact and behavior are generally intact. The emotional response is usually consistent with the delusional concern, and the mood is often appropriately depressed, frustrated, or even intensely angry or elated. Restlessness and agitation may be present. Loquaciousness and circumstantiality, usually accompanying descriptions of the delusions, are found in some patients, but formal thought disorder as sometimes found in schizophrenia is absent. Persons with delusional disorder may behave in a remarkably normal way much of the time; they become strikingly different when the delusion is focused on, at which time thinking, attitude, and mood may change direction abruptly. A. Munro has called this shift in response a characteristic, possibly unique feature of delusional disorder. In the delusional mode the patient is hyperalert, preoccupied, and driven by the delusional concern. In the normal mode, the patient's mood becomes calm, the conversation neutral, and the patient finds it easier to focus on other issues. The shift between modes can be difficult for lay persons to comprehend. Social and marital functioning are more likely to be compromised than intellectual and occupational functioning. Associated features in delusional disorder include those of the paranoid syndrome (Table 13.2–3). The degree of hostility and suspiciousness may be such that violent or aggressive behavior results. Litigious behavior is common among such patients. However, some patients, notably those with somatic delusions, may not display hostility, anger, or even suspiciousness to any considerable degree.

MENTAL STATUS EXAMINATION The patient's complaints are brought to the attention of the clinician by the patient or a third party, such as police, family, neighbors, or a consulted physician or attorney. The patient may have attracted attention by asking for protection, quarreling with neighbors, visiting too many

clinics, or similar behavior. The complaint focuses on the distressing behavior and possibly on incidental symptoms. The patient will not complain of a psychiatric condition; in fact, he or she will deny that or the presence of any psychiatric symptoms. Often to the surprise of those expecting to observe a range of mental deviances, examination leads to the discovery that thinking, orientation, affect, attention, memory, perception, and personality are intact. The patient's thinking is so clear and the delusional features are so central to his or her concerns that the clinician begins to anticipate the interview responses of the patient to the point that accurate predictions of specific actions and reactions are possible. Such predictability may distinguish the behavior of the patient with delusional disorder from behavior associated with other psychotic conditions. The patient's behavior and responses to the interview are consistent with the range of features in other paranoid conditions. There may be hostility, anger, lack of cooperation, and a sarcastic or challenging quality in most of what the patient says.

The capacity to act in response to delusions is an important dimension of the evaluation. Level of impulsiveness should be assessed and related to any potential for violent or suicidal behavior. The patient's self-righteousness, the intensity of the delusional experience, and its emotional impact on the patient may be clues to possible violent behavior; any plans for harming others, including homicide, should be inquired about. Suicidal behavior is an equally important concern. Impulses for self-harm arise in settings of frustration, demoralization, and even depression. If such thoughts exist, the patient should be asked how they were handled in the past. Jealousy and erotomania are perhaps especially important concerns in the assessment of possible aggression and violence. Stalking, history of abuse, and arrest records should be inquired about. Careful judgment and diplomatic interviewing are especially important in such presentations.

ASSESSMENT OF DELUSIONS The detection of delusions solidifies the judgment that a paranoid condition is present. Delusions are usually easy to detect. Features of behavior (Table 13.2–3) may suggest their presence. Associated psychopathological symptoms such as hallucinations, disturbed form of thought, and mood disorder may also indicate that delusions are part of the clinical picture.

The clinical challenge is clear in subtle cases. Fundamentally, the clinician must make a judgment based on available observations and the reported private mental experience of the patient. Attempts to dissuade the patient with counterevidence and counterarguments may be useful in determining whether the patient's beliefs can be influenced in ways that are usually sufficient to change a nondelusional person's mind. Spending time in discussion with the patient to grasp the nature of delusional thinking in terms of its themes, impact on the patient's life, complexity, systematization, and related features may be crucial in making the judgment. The most sensible guideline for all cases of suspected delusional thinking is to establish as comprehensive a picture as possible concerning the condition of the patient, including the patient's subjective private experience and evidence of psychopathological symptoms. Such information should reduce much of the uncertainty of the evaluative process.

PERSECUTORY TYPE The delusion of persecution is a classic symptom of delusional disorder; persecutory type and jealousy type delusions are probably the forms seen most frequently by psychiatrists. In contrast to persecutory delusions in schizophrenia, the clarity, logic, and systematic elaboration of the persecutory theme in delusional disorder leave a remarkable stamp on this condition. The absence of

other psychopathology, of deterioration in personality, or of deterioration in most areas of functioning also contrasts with the typical manifestations of schizophrenia. A 56-year old woman, X-ray technician who had emigrated as an adult from Europe, and married late in life, presented to the emergency room. Her complaints were that her husband's business partner of many years intended to get her husband to resign from the business and to destroy their home. Over a number of months she had become gradually aware that a variety of apparently inconsequential incidents (such as unusual cars parked on her isolated residential street, seeing individuals she knew at restaurants, and feeling as if she were being followed each time she drove her car) pointed to a conspiracy to disrupt and ultimately destroy their lives. Her delusion of persecution was remarkably systematized and detailed; her mood in describing this was tense and irritable. There was no evidence of hallucinations, confusion, thought disorder, or mood disorder. Cognition was intact. The patient was quite intelligent and saw the clinical consultation as a means of assisting her husband to deal with the distress of being targeted in such a manner. (The husband had accompanied his wife on these consultations. He also had experienced some delusional thinking in accord with hers.)

The patient showed no evidence that suggested suicidality or potential for violence toward others. She initially refused all medication but gradually over several months of therapy and parallel frequent legal consultations agreed reluctantly to take risperidone (Risperda) and later, for postpsychotic depression, paroxetine (Paxil). She responded within weeks to 0.5 to 1 mg of risperidone administered daily or on alternate days; she refused to take the medication continuously. Within a year, she began to focus on other issues and the emotional intensity of the delusional concerns diminished although they could be aroused with modest stimulation in conversation or from happenings in her home or neighborhood.

JEALOUS TYPE Delusional disorder with delusions of infidelity has been called conjugal paranoia when it is limited to the delusion that a spouse has been unfaithful. The eponym Othello syndrome has been used to describe morbid jealousy that can arise from multiple concerns. The delusion usually afflicts men, often those with no prior psychiatric illness. It may appear suddenly and serve to explain a host of present and past events involving the spouse's behavior. The condition is difficult to treat and may diminish only on separation, divorce, or death of the spouse.

Richard Krafft-Ebing described the symptom of delusional jealousy in alcoholics in 1891 and believed that extreme jealousy was pathognomonic for alcoholism. Other disorders with this symptom were later described. A recent retrospective analysis of 8134 psychiatric inpatients disclosed a prevalence of delusional jealousy of 1.1 percent among the major diagnostic groups. Among ICD-9 paranoid disorders, a 6.7 percent lifetime point prevalence was determined. Delusional disorder with alcohol dependence frequently shows the single delusion of jealousy, a persistent feature that sometimes remits if alcohol abuse is brought under control. In personality disorders the symptom may be confused with extreme jealousy, but other psychotic features should be absent. The prevalence of delusional jealousy among hospitalized patients with mood disorder was a surprisingly low 0.1 percent. A study of 26,000 psychiatric inpatients using DSM-III-R criteria yielded a 0.17 percent rate of delusional disorder, jealous type. Jealous delusions occur much more frequently in other disorders than in delusional disorder.

Marked jealousy (usually termed pathological or morbid jealousy) is thus a symptom of many disorders including schizophrenia (where female patients more commonly display this feature), epilepsy, mood disorders, drug abuse, and alcoholism—for

which treatment is directed at the primary disorder. Jealousy is a powerful emotion; when it occurs in delusional disorder or as part of another condition it can be potentially dangerous and has been associated with violence, notably both suicide and homicide. The forensic aspects of the symptom have been noted repeatedly, especially its role as a motive for murder. However, physical and verbal abuse occur more frequently than extreme actions among individuals with this symptom. Caution and care in deciding how to deal with such presentations are essential not only for diagnosis, but also from the point of view of safety.

A 47-year-old carpenter was brought for psychiatric examination following complaints by neighbors about his loud yelling and verbal abuse of his girlfriend. The patient resented the psychiatric referral, but was willing to give an account of his concerns. His girlfriend, he complained, was having an affair with someone, but he was not sure who the interloper was. On his own, however, he had begun gathering evidence: strands of hair found in the apartment, photographs of soiled sheets, and suspicious items from the trash—all of which he claimed proved that an affair was ongoing. He revealed plans to tape-record, possibly videotape, his girlfriend's activities while he was on the job. Upon admitting that he had told his girlfriend that he would kill her if the affair persisted, he was admitted to the hospital. He was treated with a serotonin–dopamine antagonist in low dosages and responded with a reduction in the intensity of his rage and preoccupation. Eventually, he left the hospital, but only after his girlfriend had moved away. He still harbored suspicions but accepted the termination of the relationship without voluble opposition.

EROTOMANIC TYPE Patients with erotomania have delusions of secret lovers. Most frequently the patient is a woman, but men are also susceptible to the delusion. The patient believes that a suitor, usually more socially prominent than herself, is in love with her. The delusion becomes the central focus of the patient's existence and the onset can be sudden.

Erotomania, the *psychose passionelle*, is also referred to as de Clerambault's Clérambault's syndrome to emphasize its occurrence in different disorders. Besides being the key symptom in some cases of delusional disorder, it is known to occur in schizophrenia, mood disorder, and other organic disorders. There is no mention of erotomania in DSM-III: the condition was termed atypical psychosis. DSM-III-R reinstated the condition, and it remains in DSM-IV.

Patients with erotomania frequently show certain characteristics: they are generally but not exclusively women, unattractive in appearance, in low-level jobs, and they lead withdrawn, lonely lives being single and having few sexual contacts. They select secret lovers who are substantially different from themselves. They exhibit what has been called paradoxical conduct, the delusional phenomenon of interpreting all denials of love, no matter how clear, as secret affirmations of love. The course may be chronic, recurrent, or brief. Separation from the love object may be the only satisfactory means of intervention. Although men are less commonly afflicted by this condition than women, they may be more aggressive and possibly violent in their pursuit of love. Hence, in forensic populations men with this condition predominate. The object of aggression may not be the loved individual but companions or protectors of the love object who are viewed as trying to come between the lovers. The tendency toward violence among men with erotomania may lead initially to police rather than psychiatric contact. In certain cases resentment and rage in response to an absence of reaction from all forms of love communication may escalate to a point that the love object is in danger.

A 29-year-old male financial analyst, while having lunch in a downtown restaurant observed the arrival of a well-known local media personality, an attractive woman about his age. He experienced several moments of eye contact with the woman and became convinced that she had fallen in love with him. There ensued a barrage of flowers, letters, phone calls, and even several attempts to meet with her at her workplace. The woman rebuffed all such efforts and eventually called the police. The man was arrested on a stalking charge after he was observed following the woman to her residence. He was angry and threatening to the police, finally admitting that he had purchased a handgun but refusing to give a reason for the purchase. He was remanded to a forensic psychiatric unit, treated with pimozide, and eventually discharged on a court-supervised probation.

SOMATIC TYPE Delusional disorder with somatic delusions has been called monosymptomatic hypochondriacal psychosis. The condition differs from other conditions with hypochondriacal symptoms in degree of reality impairment. In delusional disorder the delusion is fixed, unarguable, and presented intensely, because the patient is totally convinced of the physical nature of the disorder. In contrast, persons with hypochondriasis often admit that their fear of illness is largely groundless. The content of the somatic delusion may vary widely from case to case. Munro has described the largest series of cases and has used the content of delusions to define three main types: (1) delusions of infestation (including parasitosis); (2) delusions of dysmorphophobia, such as of misshapeness, personal ugliness, or exaggerated size of body parts (this category seems closest to that of body dysmorphic disorder); and (3) delusions of foul body odors or halitosis. This latter category, sometimes referred to as olfactory reference syndrome, appears somewhat different from the category of delusions of infestation in that patients with the former have an earlier age of onset (mean 25 years), male predominance, single status, and absence of past psychiatric treatment. Otherwise the three groups, although individually low in prevalence, appear to overlap.

The frequency of these conditions is low, but they may be underdiagnosed because patients present to dermatologists, plastic surgeons, and infectious disease specialists more often than to psychiatrists in the unremitting search for curative treatment. This may partially account for Kraepelin's skepticism about the occurrence of this form of paranoia. Several recent reports indicate that pimozide (a diphenylbutylperidine and highly specific dopamine blocker) and certain serotonin-specific reuptake inhibitors may be effective in treatment of such disorders, even in cases with a variety of delusional themes. There may be a heightened association of shared psychotic disorder involving primary cases of hypochondriacal delusion; one series reported a quarter of cases with such an association.

Patients with this condition have a poor prognosis without treatment. It affects both sexes roughly equally. A previous history or family history of psychotic disorder is uncommon. In younger patients, a history of substance abuse or head injury is frequent. Although anger and hostility are commonplace, shame, depression, and avoidant behavior are even more characteristic. Suicide, apparently motivated by anguish, is not uncommon.

A 40-year-old single unemployed man is referred by his primary care physician because of repeated consultations related to his complaint of hair loss. A dermatologist evaluated the patient, found no pathology, and told the patient that the minimal hair loss was normal. The patient refused to accept this judgment and demanded a further consultation. Because of managed-care restrictions, the patient consulted two additional specialists with his own (meager) funds with similar results.

He had quit his job because of embarrassment about the hair loss and had become increasingly indebted financially. The psychiatric consultation infuriated him but he cooperated because he thought that the hair loss had begun with some "pills" he had been prescribed several years previously for anxiety and insomnia and that a psychiatrist might have something to add to understanding his case, including perhaps an antidote that might relieve the loss of hair. Treatment with an antidepressant agent proved unsatisfactory and the patient was started on an atypical antipsychotic drug with modest success. He complained less frequently about the hair loss and eventually began to express concern about his loneliness and his fear of being a burden to his aging parents, whom he lived with for financial reasons. His insight, however, remained limited and he intermittently voiced his concerns about his appearance and hair loss to his psychiatrist.

GRANDIOSE TYPE Delusions of grandeur (megalomania) have been noted for years. They were described in Kraepelin's paranoia and have been associated with conditions fitting the description of delusional disorder. Whether this subtype occurs in clinical practice sufficiently enough to warrant a classification is debatable.

A 51-year-old man was arrested for disturbing the peace. Police had been called to a local park to stop him from carving his initials and those of a recently formed religious cult into various trees surrounding a pond in the park. When confronted, he had scornfully argued that, having been chosen to begin a new townwide religious revival, it was necessary for him to publicize his intent in a permanent fashion. The police were unsuccessful at preventing the man from cutting another tree and made the arrest. Psychiatric examination was ordered at the state hospital, and the patient was observed there for several weeks. He denied any emotional difficulty and had never received psychiatric treatment. There was no history of euphoria or mood swings. The patient was angry about being hospitalized and only gradually permitted the doctor to interview him. In a few days, however, he was busy preaching to his fellow patients and letting them know that he had been given a special mandate from God to bring in new converts through his ability to heal. Eventually, his preoccupation with special powers diminished and no other evidence of psychopathology was observed. The patient was discharged, having received no medication at all. Two months later he was arrested at a local theater, this time for disrupting the showing of a film that depicted subjects he believed to be satanic.

MIXED TYPE The category of mixed type applies to patients with two or more delusional themes. However, this diagnosis should be reserved for cases in which no single delusional type predominates.

UNSPECIFIED TYPE The category of unspecified type is reserved for cases in which the predominant delusion cannot be subtyped within the previous categories. A possible example is certain delusions of misidentification, for example, Capgras's syndrome, named after the French psychiatrist who described the *illusion des sosies* or the illusion of doubles. The delusion in Capgras's syndrome is the belief that a familiar person has been replaced by an impostor or persons. Others have described variants of the Capgras's syndrome, namely the delusion that persecutors or familiar persons can assume the guise of strangers (Fregoli Fregoli's *Frégoli's phenomenon*) and the very rare delusion that familiar persons could change themselves into other persons at will (*intermetamorphosis*). Each disorder is not only rare but is highly associated with schizophrenia, dementia, epilepsy, and other organic disorders. Reported cases have been predominantly in women, have had associated paranoid features, and have included feelings of depersonalization or derealization. The delusion may be shortlived, recurrent, or persistent. It is unclear whether delusional

disorder can appear with such a delusion. Certainly, the Fregoli Frégoli and intermetamorphosis delusions have bizarre content and are unlikely, but the delusion in Capgras's syndrome is a possible candidate for delusional disorder. The role of hallucination or perceptual disturbance in this condition needs to be explicated.

Shared Psychotic Disorder

Shared psychotic disorder (also referred to over the years as shared paranoid disorder, induced psychotic disorder, folie à deux, and double insanity) was first described by Lasegue and Falret in 1877. It is probably rare, but incidence and prevalence figures are lacking and the literature consists almost entirely of case reports. The disorder is characterized by the transfer of delusions from one person to another. Both persons are closely associated for a long time and typically live together in relative social isolation. In its most common form, folie imposée imposée (which is covered by the DSM-IV criteria in Table 13.2–10), the individual who first has the delusion (the primary case) is often chronically ill and typically is the influential member of a close relationship with a more suggestible person (the secondary case) who also develops the delusion. The secondary case is frequently less intelligent, more gullible, more passive, or more lacking in self-esteem than the primary case. If the pair separates, the secondary case may abandon the delusion, but this outcome is not uniformly seen. The occurrence of the delusion is attributed to the strong influence of the more dominant member. Old age, low intelligence, sensory impairment, cerebrovascular disease, and alcohol abuse are among the factors associated with this peculiar form of psychotic disorder. A genetic predisposition to idiopathic psychoses has also been suggested as a possible risk factor. The ICD-10 criteria for induced delusional disorder are given in Table 13.2–11.

Table 13.2-10. DSM-IV Diagnostic Criteria for Shared Psychotic Disorder

- A. A delusion develops in an individual in the context of a close relationship with another person(s), who has an already-established delusion.
- B. The delusion is similar in content to that of the person who already has the established delusion.
- C. The disturbance is not better accounted for by another psychotic disorder (e.g., schizophrenia) or a mood disorder with psychotic features and is not due to the direct physiological effects of a substance (e.g., a drug of abuse, a medication) or a general medical condition.

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Table 13.2-11. ICD-10 Diagnostic Criteria for Induced Delusional Disorder

- A. The individual(s) must develop a delusion or delusional system originally held by someone else with a disorder classified in schizophrenia, schizotypal disorder, persistent delusional disorder, or acute and transient psychotic disorders.
- B. The individuals concerned must have an unusually close relationship with one another, and be relatively isolated from other people.
- C. The individual(s) must not have held the belief in question before contact with the other person, and must not have suffered from any other disorder classified in schizophrenia, schizotypal disorder, persistent delusional disorder, or acute and transient psychotic disorders in the past.

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Other special forms have been reported, such as folie simultanee simultanée, where two people become psychotic simultaneously and share the same delusion. Occasionally, more than two individuals are involved (e.g., folie a à trois, quatre, cinq; also folie a à famille), but such cases are especially rare. The most common relationships in folie a à deux are sister-sister, husband-wife, and mother-child, but other combinations have also been described. Almost all cases involve members of a single family.

There is some question whether patients with such conditions are truly delusional rather than highly impressionable, as frequently there is merely passive acceptance of the delusional beliefs of the more dominant person in the relationship until they are separated, at which point the unusual belief may remit spontaneously. In the DSM-IV criteria the requirement that the secondary case not have a psychotic disorder prior to onset of the induced delusion illustrates the relevance of this question. Also, the psychopathology of secondary cases varies. In DSM-III such patients were required to meet the criteria for paranoid disorder (i.e., show evidence of disturbed personality and perhaps evidence of other psychiatric disorder, mental subnormality, or dementia); some cases may fit the definition of delusional disorder.

A 40-year-old woman consulted physicians to help cure her problem of disagreeable body odor. The physicians failed to satisfy the woman's hopes of diagnosis and treatment, because they found nothing wrong with her. They did occasionally recommend psychiatric consultation, which she refused. Her husband, a quiet, retiring man of 35, accompanied his wife to all medical specialist consultations. When questioned, he shared his wife's concerns about body odor and provided many examples of how distressing this problem had become. When he was told that there really was nothing wrong with his wife, he objected repeatedly and proclaimed that the doctors were incompetent. A psychiatrist was called to the clinic to see the couple and found consistent stories from both. The woman accepted a recommendation for hospitalization on the psychiatry-medical unit, and the husband returned home. After weeks of evaluation and treatment, the woman was discharged. The husband had stopped visiting her, and when informed that his wife would be coming home he said that he thought she had been cured of her problem. However, 3 months later the couple was once again visiting different specialists.

A 52-year-old man was referred by the court for inpatient psychiatric examination after being charged with disturbing the peace. He had been arrested for disrupting a trial, complaining of harassment by various judges. He had walked into a courtroom, marched to the bench, and begun to berate the probate judge. While in the hospital, he told in detail of conspiratorial goings-on in the local judiciary. A target of certain judges, he claimed he had been singled out for a variety of reasons for many years: he knew what was going on; he had kept records of wrongdoings; and he understood the significance of the whole matter. He refused to elaborate on the specific nature of the conspiracy. He had responded to it with frequent letters to newspapers, the local Bar association, and even to a Congressional subcommittee. His mental state, apart from his story and a mildly depressed mood, was entirely normal. A family interview revealed that his wife and several grown children had shared the belief in a judicial conspiracy directed against the patient for a number of years. There was no change in delusional thinking in the patient or the family after 10 days of observation and the patient refused follow-up treatment.

The intensity of conviction is governed by the presence of the primary case in the life of the secondary case. Protection is provided by others who share the delusion and

believe that the response is reasonable. Munro has found that shared psychotic disorder is frequently associated with delusional disorder, somatic type. In the second case described for persecutory delusional disorder, the husband, a somewhat passive and isolated man, shared his wife's convictions. With her treatment, he also became less concerned about a conspiracy and began to share his doubts about the whole matter with his therapist.

A recent summary of the Japanese literature indicated that in 97 cases of *folie à deux*, the phenomenology and epidemiology were similar to those in western reports.

PATHOLOGY AND LABORATORY EXAMINATION

Pathology

As in most psychiatric conditions, there is no evidence of localized brain pathology to correlate with clinical psychopathology in patients with delusional disorder. These patients seldom die early and show no consistent abnormalities on neurological examination. Delusions can complicate many disorders and virtually all brain disorders. Certain disorders produce delusions at rates greater than that expected in the general population: for example, epilepsy (especially of the temporal lobe), degenerative dementias (dementia of the Alzheimer's type and vascular dementia), cerebrovascular disease, extrapyramidal disorders, and traumatic brain injury. Although many types of delusions have been reported in patients with brain disorders, there appear to be particular connections between delusion phenomenology and certain kinds of brain dysfunction. For example, patients with more severe cortical impairment tend to experience more simple, transient, persecutory delusions. This type of delusional experience is characteristic of conditions such as Alzheimer's disease, dementia and metabolic encephalopathy that are also associated with significant cognitive disturbance. More complex (i.e., elaborate and systematic) delusional experiences tend to be more chronic, intensely held, resistant to treatment, and associated with neurological conditions producing less intellectual impairment and strong affective components. Those features occur in patients with neurological lesions involving the limbic system or subcortical nuclei rather than cortical areas. That, coupled with the observation of response of some patients to drug treatment, such as pimozide and other medications, provides a rational basis on which to hypothesize the presence of subcortical pathology, possibly involving systems subserving temporolimbic areas. Available evidence suggests that if there is a lesion, it will be subtle.

Imaging studies have begun to yield subtle findings about delusional disorder. In one study using quantitative volumetry in magnetic resonance imaging, 16 patients with delusional disorder showed lateral ventricle enlargement greater than in subjects with schizophrenia (N = 31) and almost twice that of age-matched healthy controls (N = 35). Although this study showed no evidence of cortical infarcts (cerebrovascular injury), other studies have suggested that unsuspected cerebral infarction may occur in a high proportion of late-onset cases with delusional disorder. A further examination of these 16 delusional disorder subjects revealed that the degree of physiological right-left asymmetry was significantly greater in the temporal lobes.

Another study has tentatively concluded that eye-tracking dysfunction in the saccadic system is present in delusional disorder, possibly reflecting some attentional impairment related to voluntary saccadic eye movement areas. Despite the subtle nature of such findings, future empirical studies, guided by etiological hypotheses, could lead to breakthroughs. Given the low incidence of delusional disorder, intensive studies of specific cases and of conditions with delusions from known causes (and with identifiable neuropathologies) offer useful beginning points. Recent studies of

misidentification syndromes (e.g., Capgras's syndrome) offer the prospect of developing more refined models of neuropathological mechanisms for delusional disorder.

Laboratory Examination

A range of assessments is often necessary, but several have a high likelihood of detecting key factors in the case. The use of drug-screening measures is particularly valuable given the marked delusional responses induced by a number of substances, especially alcohol, amphetamines, cocaine, and other central nervous system stimulants.

Neuropsychological assessment may help to disclose evidence of impaired intellectual functioning suggestive of brain abnormalities. The assessment of intelligence may show discrepancies between verbal and performance scores as well as scatter in overall performance. Limited data on delusional disorder (especially the more chronic forms) suggest that average or marginally low intelligence is characteristic of patients with this condition. A preliminary comparison of patients with late-onset delusional disorder and schizophrenia has indicated neuropsychological impairment to be somewhat less for the former group. Projective testing such as the Rorschach test has limited value in making the diagnosis but may confirm features consistent with it. Deviation on the paranoia scale of the Minnesota Multiphasic Personality Inventory-2 (MMPI-2) has strong correlations to paranoid features and may help substantiate the diagnosis or raise it as a possibility.

DIFFERENTIAL DIAGNOSIS

Delusional Disorder

Because delusional disorders are uncommon, idiopathic, and possess features characteristic of the full range of paranoid illnesses, differential diagnosis has a clearcut logic: delusional disorder is a diagnosis of exclusion. There are many conditions to consider (Table 13.2–4), especially the more common disorders associated with paranoid features (Table 13.2–12). To avoid premature diagnosis, careful evaluation is required.

Table 13.2-12. More Common Disorders Associated With Paranoid (Delusional) Features

Alcohol abuse

Drug abuse (especially CNS stimulants)

Anticholinergic toxicity

Sedative-hypnotic withdrawal

Delirium

Dementia

HIV infection

Brain tumor

Epileptic disorder

Mood disorders

Schizophrenia/schizoaffective disorders

Clinical assessment of paranoid features requires three steps. Initially, the clinician must recognize, characterize, and judge as pathological the presence of paranoid features. Next, the clinician should determine whether they form a part of a syndrome or are isolated. Finally, a differential diagnosis should be developed. The first of the three steps must be pursued thoroughly. The clinician must be aware that a range of objective traits or behaviors (Table 13.2–3) is often found in paranoid illness and may constitute the only clue that a paranoid illness is present. Patients with paranoid

symptoms are frequently unwilling to reveal their subjective experiences to examiners or to cooperate in the clinical investigation. Careful interviewing of the patient and other informants may disclose evidence that the behavior is clearly psychopathological; in other cases, however, that conclusion must await further observations. Sometimes investigation is required to determine whether the belief is indeed delusional or not. Premature acceptance that the patient has delusional disorder has at times been an embarrassment to some clinicians who learn that the patient was not deluded. If the judgment that the patient is delusional seems unassailable, then careful elaboration of the nature of the delusion is called for. The delusional thinking should be examined for its fixity, logic, encapsulation, degree of systematization and elaboration, and its effect on planning and action.

Having determined that a paranoid condition is present, the clinician should attend to premorbid characteristics (personality, adjustment, symptom development, medical problems, and so forth), the course, and associated symptoms to detect patterns of syndromic psychopathology or isolated symptom presentations—this is step two. The discovery of clouded consciousness, perceptual disturbance, other psychopathology, physical signs, or confusing symptoms may suggest different causes for paranoid features. Isolated acute paranoid symptoms, on the other hand, often appear early in medical illness.

Finally, the clinician should resist the temptation to make the diagnosis of schizophrenia or delusional disorder prematurely in cases where paranoid features are present because these features occur regularly in a variety of psychiatric and medical illnesses. Consequently, awareness of the multiple causes of paranoid features (step one) is essential to completing the differential diagnosis (step three).

Certain principles should guide effective assessment. First, it is important to have knowledge of the paranoid features and patterns of the clinical disorders in which they occur. For example, a small percentage (perhaps 10 percent) of schizophrenia cases have their onset after age 40, and most idiopathic psychiatric conditions do not begin after age 50. Second, the premorbid status of the patient should be determined. Generally, a normal premorbid state suggests that acute paranoid features are the consequence of medical disease. Third, an abrupt change in personality, mood, ability to function, and mental state should be noted because this may indicate complications resulting from medical disease. Fourth, in those cases in which there is evidence that the patient has been refractory to psychotropic medication or psychotherapy, the continuing presence of paranoid features should alert the clinician to consider alternative diagnoses.

The final diagnosis in cases where paranoid features are prominent should be made only following: (1) a complete medical and psychiatric history with special attention (because of their high prevalence) to alcohol and other drug substance use (including drugs of abuse, prescribed drugs, and over-the-counter medication use history); (2) a thorough physical examination, including neurological and mental status examinations; (3) appropriate laboratory studies, particularly serological, toxicological, endocrine, microbiological, radiological, and electroencephalographic studies.

There are certain delusional conditions that, because of their frequency and seriousness, should be routinely considered in the differential diagnosis, as among the most likely sources of delusions (Table 13.2–13). For example, delirium, dementia, psychotic disorder due to a general medical condition, and substance-induced psychotic disorder should receive special attention. Awareness of the potential for patients with each of these disorders to present with delusions in a state of clear

consciousness prior to the elaboration of the defining syndromal symptoms should be kept in mind.

Scroll right to see more columns.

Table 13.2-13. Differential Diagnosis of Delusional Disorder

Disorder	Delusions	Hallucinations	Awareness	Other Features
Delusional disorder	+	Occasionally	Alert	Relatively free of psychopathology
Psychotic disorder due to a general medical condition, with delusion	+	+ frequent	May be impaired	Cognitive changes; perceptual changes; substance abuse history; impairment of functioning
Substance-induced psychotic disorder likely	+(can be bizarre)	+	Acute: impaired Chronic: may be	History of impaired functioning
Schizophrenia	+(bizarre) alert	+ Alert	Emotional changes, pervasive thought disorder; role impairment	
Major depressive episode	+(mood congruent)	± mood and neurovegetative features	Alert	Concerted changes in
Manic Episode	+(mood congruent)	± need for sleep activity, energy, lack of inhibition	Alert	Concerted changes in
Obsessive-compulsive disorder	-	- Alert	Alert	Not psychotic; impaired functioning likely
Personality disorder	-	- Alert	Alert	Not psychotic
Somatoform disorder	-	- Alert	Alert	Not psychotic
Shared psychotic disorder	+	- Alert	Alert	Close associate has same delusions

Psychotic Disorder Due to a General Medical Condition, With Delusions Delusions arise in a number of organic diseases and syndromes, many of which are listed in Table 13.2–2. What they frequently have in common is a disturbance of perception, particularly of visual and auditory functioning. Physical, neurological, and mental status studies as well as laboratory examinations will usually detect the organic causes of delusions. Each evaluation should focus on perceptual disturbance. Medical conditions associated with delusions should be searched for according to the guidelines outlined concerning differential diagnosis.

Substance-Induced Psychotic Disorder, With Delusions Drug intoxications are particularly relevant to this disorder. Substances of abuse, such as amphetamines, hallucinogens, phencyclidine, and cocaine; over-the-counter drugs, such as sympathomimetics; and prescribed drugs, such as steroids, methyl dopa (Aldomet) and levodopa (Dopar, Larodopa) can cause psychotic disorder, with delusions, sometimes without prominent cognitive impairment. In acute states, confusion, disorientation,

and clouding of consciousness may be evident; in chronic cases the picture may be more difficult to distinguish from delusional disorder because cognitive changes are less pronounced. A careful drug history and screen may establish the diagnosis. A history of alcohol abuse or dependence is so common that it should always be considered; alcoholism is often associated with jealousy, persecutory ideas, and poor impulse control.

Cognitive Disorders Dementia should be considered when paranoid features occur, particularly in older persons. Mental status examination should uncover characteristic cognitive changes absent in delusional disorder. Delirium, with its fluctuating course, confusion, memory impairment, and transient delusions, contrasts with the clarity of mental functioning and the persistence of delusions in delusional disorder and should be considered in acute cases with paranoid features.

Schizophrenia Delusions may be the presenting feature of schizophrenia and this diagnosis should be considered when the delusions are implausible or bizarre, affect is blunted or incongruous with thinking, auditory and possibly visual hallucinations are prominent, thought disorder is pervasive, or role functioning is impaired. Patients with paranoid schizophrenia may have somewhat less bizarre delusions, but role functioning is impaired; also prominent auditory hallucinations are often present, unlike in delusional disorder.

Shared Psychotic Disorder The delusions and symptoms of shared psychotic disorder may resemble those of delusional disorder; however, the delusions arise in the context of a close relationship with a delusional person, are identical in content to the delusions of that person, and diminish or disappear when secondary and primary cases are separated.

Mood Disorders With Psychotic Features The persistent and profound dysphoric mood of patients with depression often points to the proper diagnosis; in delusional disorder, affect may be intense, but is not itself an overwhelming or preoccupying experience to the patient. Delusions in depression, if present, are frequently related to mood (mood-congruent delusions) and usually indicate severe depression. For example, patients with feelings of worthlessness or guilt may consider that persecution against them is justified as a punishment for their evil ways. Somatic delusions may be puzzling to differentiate if the clinician fails to consider associated psychopathological features. If delusions occur exclusively during mood episodes, the diagnosis is mood disorder with psychotic features. Depression refers to a host of signs and symptoms, and usually has a constellation of neurovegetative features (affecting appetite, sleep, libido, energy, and so forth) that are not part of delusional disorder. Moreover, depression is frequently cyclical and is often associated with a positive family history of mood disorder. Patients with delusional disorder, in contrast, are remarkably free of symptoms other than the delusion. Chronic demoralization may result from repeated failure to obtain the kind of response desired in delusional disorder. Not infrequently, mood symptoms that meet the criteria for a mood episode are present in a delusional condition. Delusional disorder is diagnosed only if the total duration of all mood episodes remains brief relative to the total duration of the delusional disturbance. There is some evidence to suggest that depression is the most common comorbid condition in delusional disorder.

Manic Episode Manic delusions, often grandiose and therefore mood congruent, occur in the severest stages of this illness. This could mislead the diagnostician, but the cyclical nature, the marked change in mood (often euphoric or irritable at a very intense level), the reduced need for sleep, increased energy, easy distractibility, lack of focused concentration ability, lack of social inhibition, and increased activity level

of manic episodes should be decisive in distinguishing that condition from delusional disorder.

Obsessive-Compulsive Disorder Severe forms of this disorder should be considered in the differential diagnosis, especially obsessive-compulsive disorder with poor insight. Preoccupation with fear, unusual rituals, and obsessional beliefs may be puzzling, yet the pervasive effects of the condition on functioning differ from the experience of delusional disorder. Moreover, delusions and hallucinations should be absent. In practice, this differential diagnosis may be difficult to determine without a period of observation. In some cases it may be necessary to make the diagnosis and that of delusional disorder.

Somatoform Disorders Severe forms of body dysmorphic disorder may be difficult to distinguish from delusional disorder. The degree of conviction about imagined physical disfigurement may be the only guide for differential diagnosis. Lack of other features of psychopathology, often present in such cases, may also help to make the distinction.

Hypochondriasis may also be distinguished on the basis of absence of delusions, although many of the behaviors associated with delusional disorders, somatic type, may occur. Usually such patients reveal some doubt or uncertainty about the validity of their health preoccupations. Their overvalued beliefs about disease or affliction may clearly resemble delusional disorder, somatic type; severe cases may require considerable diagnostic effort, and as in obsessive compulsive disorder, also require a second diagnosis of delusional disorder.

Paranoid Personality Disorder Individuals with paranoid personality disorder by definition have abundant paranoid features. They are persistently oversensitive, ready to take offense, suspicious, resentful, rigid, and frequently self-centered. Rather than delusions, such persons tend to report strongly held ideas (overvalued ideas); generally, however, they are believed to be free of delusions, which is the most useful differential feature. There is some evidence that this personality pattern occurs often enough in families of probands with delusional disorder to suggest a possible genetic connection between the two.

Schizoid Personality Disorder and Schizotypal Personality Disorder Paranoid features may occur in patients with these personality disorders as well. The pervasive disturbance in personality functioning and the absence of delusions and other psychotic features are usually definitive distinguishing characteristics. Delusional disorder has generally not been associated with this type of premorbid pattern of personality.

Disorders of Aging Any discussion of differential diagnosis of paranoid features is incomplete unless consideration is given to the occurrence of paranoid features in the elderly. Paranoid features develop frequently in the elderly, and assessment in such cases should be particularly thorough because information about paranoid features among the aged is limited. There are several facts worth knowing: (1) the association of depressive illness with paranoid features is high enough to warrant suspicion of mood disorder in all cases with paranoid features; (2) there appears to be a late-onset syndrome sometimes labeled late paraphrenia or late-onset schizophrenia in which paranoid characteristics and hallucinosis frequently occur (this diagnosis, however, is warranted only when no other disorder can be diagnosed); (3) the sudden onset of acute paranoid features in the elderly can be a sign of cerebrovascular injury or other medical illness; (4) many of the medical conditions associated with delusions have increased incidence in the elderly population; for example, delusions can arise in the early course of presenile dementia and senile dementia conditions when deficits in

clinical examination probes or neuropsychological performance may be inconspicuous; (5) perhaps most important for the general clinician is to recognize sources of increased risk of paranoid disorder among older individuals. It is now known that many factors contribute to the incidence of paranoid features in the aged, including lack of stimulating company, isolation, physical illness, the aging process itself, loss of hearing, and loss of visual acuity, each of which should be carefully assessed. Delusional disorder may be present in the elderly, may even have its onset in the elderly, but the frequency of other causes of paranoid features calls for a prudent, systematic search.

Shared Psychotic Disorder

Malingering, factitious disorder with predominantly psychological signs and symptoms, psychotic disorder due to a general medical condition, and substance-induced psychotic disorder need to be considered in the differential diagnosis of shared psychotic disorder. The boundary between shared psychotic disorder and generic group madness, such as among the victims of the Jonestown massacre in Guyana, is unclear.

COURSE AND PROGNOSIS

Delusional Disorder

Onset can begin in adolescence but generally occurs from middle to late adulthood with variable patterns of course, including lifelong disorder in some cases. Studies generally indicate that delusional disorder does not lead to severe impairment or change in personality, but rather to a gradual, progressive involvement with the delusional concern. Suicide has been associated with such disorders, although most patients live a normal life span. The base rate of spontaneous recovery may not be as low as previously thought, especially because only the more severely afflicted patients are referred for psychiatric treatment. Retterstol's personal follow-up investigation of a large series of cases has provided much of the viewpoint on the natural history of the disorder.

The more chronic forms of the illness (patients presenting with features for more than 6 months) tend to have their onset early in the fifth decade. Onset is acute in nearly two-thirds of the cases, and gradual in the remainder. In 53 percent the delusion has disappeared at follow-up, is improved in 10 percent, and is unchanged in 31 percent. In more acute forms of the illness the age of onset is in the fourth decade, a lasting remission occurs in over half of patients, and a pattern of chronicity develops in only 10 percent; a relapsing course has been observed in 37 percent.

Thus the more acute and earlier the onset of the illness, the more favorable the prognosis. The presence of precipitating factors signifies a positive outcome, as does female sex and being married. In terms of prognosis, the persistence of delusional thinking is most favorable for cases with persecutory delusions, and somewhat less favorable for delusions of grandeur and jealousy. However, outcome in terms of overall functioning appears somewhat more favorable for the jealousy subtype. Such patients may experience fewer hospitalizations and are less likely to have severe psychotic or schizophrenic deteriorations. Work status at follow-up has indicated that the majority of patients are employed. These observations, although limited to few cases, provide some basis for optimism: perhaps half of cases with delusional disorders may remit, but relapse and chronicity are common.

Comorbidity Depression can be diagnosed as a coexistent disorder in the course of delusional disorder. Evidence indicates that depression is an independent disorder in such cases, that is, the disorders appear to be coincidental in their combination rather than related etiologically. This judgment must be regarded as somewhat tentative, but

the clinical value of recognizing comorbid (and often treatable) conditions is straightforward.

Shared Psychotic Disorder

The nature of the disorder suggests that separation of the submissive person who has shared psychotic disorder (the secondary case) from the dominant person (the primary case) should result in the resolution and disappearance of the psychotic symptoms in the submissive person. Often, the submissive person requires treatment with antipsychotic drugs, just as the dominant person needs antipsychotic drugs for the psychotic disorder. Because the persons are almost always from the same family, they usually live together after being released from hospital. If separated, the patient will experience a possible remission; if not separated, the patient may have a similar prognosis as the primary case.

TREATMENT

Delusional Disorder

Delusional disorder has generally been regarded as resistant to treatment and interventions have often focused on managing the morbidity of the disorder by reducing the impact of the delusion on the patient's (and family's) life. However, in recent years the outlook has become less pessimistic or restricted in planning effective treatment for these conditions. The goals of treatment are to establish the diagnosis, to decide on appropriate interventions, and to manage complications (Table 13.2–14). Fundamental to the success of these goals is an effective and therapeutic doctor-patient relationship, which is far from easy to establish. The patients do not complain about psychiatric symptoms and often enter treatment against their will; even the psychiatrist may be drawn into their delusional nets.

Table 13.2-14. Diagnosis and Management of Delusional Disorder

Rule out other causes of paranoid features

Confirm the absence of other psychopathology

Assess consequences of delusion-related behavior

Demoralization

Despondency

Anger, fear

Depression

Impact of search for "medical diagnosis," "legal solution," "proof of infidelity," etc. (i.e., financial, legal, personal, occupational, etc.)

Assess anxiety and agitation

Assess potential for violence, suicide

Assess need for hospitalization

Institute pharmacological and psychological therapies

Maintain connection through recovery

Psychosocial Treatments There is not enough evidence to substantiate the claims for any particular school or approach in talking with the patient. Insight-oriented therapy is usually contraindicated, but a combination of supportive psychotherapeutic approaches and possibly cognitive-behavioral interventions is sensible. It is unlikely that there is any psychiatric condition that requires greater diplomacy, openness, and reliability from the therapist. Considerable skill is required to deal with the profound and intense feelings that accompany these disorders.

Awareness of the fragile self-esteem and unusual sensitivity of these patients is essential for general management and somatic treatment. Clinical experience indicates that direct questioning about the veracity of the delusion, apart from carefully

establishing its nature and the evidence to support it during clinical evaluation, is seldom helpful. Although forging an alliance may be especially difficult, responding to the patient's distress rather than to the delusion itself may be effective.

Understanding that fear and anxiety serve to stimulate hostility may be the key to adopting a flexible approach that promotes empathy but maintains physical and emotional distance. Patients with the disorder suffer; they often feel demoralized, miserable, isolated, and abandoned. They may face rejection at home, from police or medical specialists, or on the job. However, they can be approached, and their treatment can focus on these experiences.

The goals of supportive therapy are to allay anxiety and initiate discussion of troubling experiences and consequences of the delusion, thereby gradually to develop a collaboration with the patient. In some patients this strategy allows the psychiatrist to suggest means of coping more successfully with the delusional thinking. For example, psychiatrists might encourage patients to keep their delusions to themselves because others might feel surprised, dismayed, or amazed, all at considerable cost to the patient. It may be possible to provide educational intervention to help amenable patients to understand how factors such as sensory impairment, social and physical isolation, and stress contribute to making matters worse. In all such approaches, the overriding aim is to assist in a more satisfying general adjustment.

Cognitive approaches have attempted to reduce delusional thinking through modification of the belief itself, focusing on the associated reasoning or the reality testing of the deluded patient. Unlike noncognitive behavioral approaches that center attention on reduction of verbal behavior (talking about the delusion), this strategy seeks a more lasting and clinically meaningful intervention through multiple techniques that keep the relationship with the patient collaborative. These techniques include distancing, homework, and exploration of emotions associated with various delusions. The effectiveness of cognitive and behavioral therapies has not been studied enough to justify recommendation. Additionally, it is important to determine the long-term as well as the short-term impact of these treatments; nevertheless, they are promising enough to justify continued assessment.

Somatic Treatment Delusional disorder is a psychotic disorder by definition, and the natural presumption has been that the condition would respond to antipsychotic medication. Because controlled studies are limited and the disorder is uncommon, the results required to support this practice empirically have been only partially obtained. The disparate findings in the recent literature on delusional disorder treatment have been summarized recently, with several qualifications. Of approximately 1000 articles published since 1961, the majority since 1980, 257 cases of delusional disorder (consistent with DSM-IV criteria) of which 209 provided sufficient treatment detail to make comparison, were assessed. Overall treatment results indicated that 80.8 percent of cases either recovered fully or partially. Pimozide (the most frequently reported treatment) produced full recovery in 68.5 percent and partial recovery in 22.4 percent of cases (N = 143) treated whereas there was full recovery in 22.6 percent and partial recovery in 45.3 percent of cases (N = 53) treated with typical neuroleptic agents [e.g., thioridazine (Mellaril), haloperidol (Haldol), chlorpromazine loxapine (Thorazine), perphenazine (Trilafon), and others]. The remaining cases (N = 13) were noncompliant with any treatment, a finding the authors regard as an underestimation (6.2 percent). There were no specific conclusions drawn regarding treatment with selective serotonin reuptake inhibitors, (SSRIs), although a number of such reports have been published. While treatment of the somatic subtype generated the largest number of reports, these authors' meta-analysis indicated that the patterns of response

were similar across all subtypes of delusional disorder. Follow-up data and personal experience indicated that long-term, possibly permanent, administration of medication is necessary to maintain remission.

The results of treatment with the serotonin-dopamine antagonists (i.e., clozapine [Clozaril], risperidone olanzapine [Zyprexa], and others) is preliminary. Two known cases of the persecutory subtype have been treated successfully with risperidone and there are published reports of clozapine effectiveness in the persecutory subtype (N = 2) and the somatic subtype (N = 2), and of risperidone effectiveness in the somatic subtype (N = 1). Unfortunately, systematic case series will develop slowly, but these preliminary results suggest that the atypical neuroleptic agents may add to the available treatment options.

Given the limited samples available, case reports are especially valuable; although many authors recommend multisite trials (to augment the small numbers of cases available at any one site), it would be beneficial for further single case reports to be published in the meanwhile. The existing literature could be improved with more attention paid to diagnosis, prior treatments, outcome, and level of compliance, as well as dosage schedules, adverse effects, length of treatment, as well as the reasons for selecting or changing particular agents. Use of (N = 1) single case research design strategies might also enhance the generalizability of findings.

The impression is growing that antipsychotic drugs are effective, and a trial, especially with pimozide or a serotonin-dopamine antagonist is warranted. Certainly, trials of antipsychotic medication make sense when the agitation, apprehension, and anxiety that accompany delusions are prominent.

Delusional disorders respond less well generally to electroconvulsive treatment than do major mood disorders with psychotic features. Some cases may respond to SSRIs, especially cases of body dysmorphic disorder with delusional concerns (Table 13.2–15). Where differential diagnosis is unclear between delusional disorder and psychotic depression, a trial of combined therapy with antipsychotic and antidepressant medications therapy may be worthwhile. When standard strategies are unsuccessful, trials of lithium (Eskalith) or of anticonvulsant medication such as carbamazepine (Tegretol) probably should be considered. However, no systematic information to support such approaches is yet available.

Table 13.2-15. Pharmacological Agents With Reports of Successful Use in Delusional Disorder

Dopamine receptor antagonists (particularly pimozide)

Serotonin-dopamine antagonists

Selective serotonin reuptake inhibitors

Somatic treatment is difficult to implement on two levels, The patients' insistence on lack of psychiatric problems may be an insurmountable barrier to initiating treatment, and their sensitivity to all adverse effects may constitute an additional frustrating factor in their care. Noncompliance continues to be a frequent observation in published clinical studies. An open and clear approach to warn patients about and to assist them through possible unpleasant experiences is essential, but the intrinsic nature of active resistance to psychiatric intervention also requires attention. In general, some patients, especially younger patients with delusional disorder, respond to supportive management and somatic treatment. Unfortunately, others, especially the elderly, may be refractory to attempts to reduce their delusional thinking. In all cases goals that are realistic and modest are the most sensible. As most of the

difficulty of this disorder results from the effects of the patient's actions concerning the delusions, any preventive approach in that domain has considerable value. Hospitalization Most delusional disorder patients can be treated effectively in outpatient settings; hospitalization may be necessary when there is potentially dangerous behavior or unmanageable aggressiveness. The patient may show signs of poor impulse control, excessive motor and psychic tension, unremitting anger, brooding, suicidal tendencies, and even threats of self-harm or aggression toward others. Suicidal ideation and planning are also potential grounds for hospitalization. Follow-up studies report suicide above the population base rate; patients with erotomania, jealousy, and persecutory delusions are particularly at risk. Once the psychiatrist decides to hospitalize the patient, it is preferable to inform the patient tactfully that voluntary hospitalization is necessary. If this strategy fails, legal means must be undertaken to commit the patient to a hospital.

Shared Psychotic Disorder

The initial step in treatment is minimally the temporary separation of the affected person from the source of the delusions, the dominant partner. This step may not only be therapeutic but diagnostic when evidence of reduced delusional thinking and preoccupation accrue. The patient may need significant support to compensate for the loss of that person. The patient with shared psychotic disorder should therefore be carefully observed for the remission of the delusional symptoms. Antipsychotic drugs can be used if the delusional symptoms have not abated in 1 or 2 weeks.

Psychotherapy with nondelusional members of the patient's family should be undertaken, and psychotherapy with both the patient with shared psychotic disorder and the dominant partner may be indicated later in the course of treatment. In addition, the mental disorder of the dominant partner should be treated. The clinician might use family therapy and social support to modify the family dynamics and to prevent the recurrence of the syndrome. It is often useful to make sure that the family unit is exposed to input from outside sources to decrease the family's isolation. In short, a comprehensive approach emphasizing support and, when necessary, medication is useful.