Delusional Disorders in Dermatology

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Definitions

Delusional disorder (DD) is defined by the presence of one or more delusions that are false beliefs of different content. The latter may include persecutory, erotomanic, grandiose, jealous, and somatically focused erroneous ideas. In contrast to schizophrenic delusions, there are no other characteristic symptoms of schizophrenia (e.g. persistent auditory hallucinations, disorganized thinking, negative symptoms). Various forms of perceptual disturbances (e.g. hallucinations, illusions, misidentifications, etc.) thematically related to the delusion may be present. Apart from actions and attitudes directly related to the delusion or delusional system, other mental functions (affect, cognition, speech, behaviour, etc.) are typically unaffected.

Classification In ICD-10, DD is coded as F22, attributed to *Persistent delusional disorders,* and placed in F2 class *Schizophrenia, schizotypal and delusional disorders.* In DSM-5, DD is coded as 297.1 and attributed to *Schizophrenia Spectrum and Other*

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Psychotic Disorders. The similar conceptualization is provided in ICD-11. DD is coded as 6A24 and placed among *Schizophrenia or other primary psychotic disorders.*

In both ICD-10 and DSM-5, DD is subdivided into several types according to delusional content.

DD types:

Type	The central theme of the delusion
Erotomanic	The individual's belief that they are the object of another person's love or sexual desire
Grandiose	The individual's belief that they possess a great talent or have made an important discovery which is underestimated and associated with an inflated sense of worth, power, knowledge, or identity.
Jealous	The individual's belief in their spouse or lover's infidelity.
Persecutory	The individual's belief that they are being intentionally harmed or is going to be intentionally harmed by other(s) who may conspire against, cheat, spy on, follow, poison, maliciously malign, harass, or obstruct etc.
Somatic	The individual's belief that something is wrong with bodily functions and/or appearance, or there are pathological sensations.

There are also mixed or unspecified types of DD, when there are several delusional themes but no one predominates or the dominant delusional belief cannot be clearly determined, respectively

Some delusions in DD of somatic type may have content related to skin, and as DD patients lack awareness or insight, i.e. strongly believe in an erroneous idea and deny mental illness, they present to a dermatologist rather than to a psychiatrist.

There are several somatic DD subtypes that dermatologists may encounter:

- Delusional infestation—DI (see Chap. 12): the belief that there is an infestation of insects or other parasites (or even inanimate objects) on or in the skin
- Delusions of internal infestation: the belief that there is an internal parasite that may migrate outside from the viscera through the skin (see Chap. 12)
- Delusional body dysmorphic disorder—DBDD (delusional dysmorphism): the belief that certain parts of the body, e.g. skin itself or its appendages, are abnormal, misshapen, or ugly
- Delusional olfactory reference syndrome—DORS: the belief that the individual emits a foul odour (cacosmia), e.g. perceiving skin as an origin of such a smell

2. *Historical names for DD:*
   (a) "Paranoia" by Heinroth [1818], Kahlbaum [1863] and Kraepelin [1899, 1921]
   (b) "Intellectual monomania" by Esquirol [1838]
   (c) "Chronic mania" by Griesinger [1845]
   (d) "Paranoid disorder" in DSM-III
   (e) "Monosymptomatic hypochondrical psychosis" by Munro (for DD of somatic type)

- Illness delusion or delusion of disease (hypochondriacal delusion) (AIDS-delusions, syphilis delusions, etc): the belief that the individual has an undiagnosed somatic illness or medical condition, e.g. sexually transmitted infection

1. *Aetiology and Pathology: see Chapter about delusional infestation*

Clinical Presentation

General considerations DD is characterized by a delusional system or a patient's specific explanatory model. The system includes a set of well organized and persistent ideas around a particular theme. Such delusional ideas are interconnected with each other in a "logical" structure with a set of "evidences" and explanations ("logical reasoning"). As there is no insight or awareness, such delusional ideas may be vigorously defended. Delusions occur in clear consciousness and not due to an underlying physical illness or to a psychiatric disorder other than delusional disorder.

Typically, delusional ideas lead to corresponding delusional behaviour. However, delusions in DD are relatively isolated (encapsulated) and coincide with otherwise intact non-delusional mental functioning, behaviour outside the delusional topic often remains normal and may mask episodes of delusional behaviour; or there are just some delusional acts incorporated into otherwise non-psychotic behaviours (partial psychosis). Esquirol (1818) stated that aside from these areas relevant to their "delusional systems", patients "think, reason, and act, like other men".

Delusional Disorders in Dermatology (Hypochondriacal or Somatic Delusions Related to Skin)

In *Delusional Body dysmorphic disorder* (DBDD), there is a delusional belief in an imagined defect in one's appearance. Patients also may have a minor or non-discernible defect that is real but rigidly and inflexibly believe they are deformed. It should be noted that there is also a non-delusional body dysmorphic disorder (BDD, see Chap. 14 about body dysmorphic disorder) not classified as a psychotic condition but classed as a somatoform disorder in ICD-10 or as an obsessive-compulsive (OCD) and related disorder in DSM-5 and ICD-11. However, this just means that appearance preoccupation may be a key feature of anxious, obsessive, overvalued, or delusional syndromes with a continuum of insight from full to absent. Here we discuss the latter.

The important feature of DBDD to distinguish from non-psychotic BDD is a lack of insight. However, sometimes it is hard to establish, as there are also OCD disorders with low insight and overvalued ideas. Thus, a dimensional approach may be recommended as there is a spectrum of dysmorphic concerns with a range of severity.

The Delusional system in dermatological DBDD includes a central belief about skin ugliness visible to others. Patients usually complain of a variety of skin blemishes that they may call "acne", "pimples", "nodules", "vascular spiders", etc. Cosmetic complaints may also include inappropriate skin colour (dyschromia) or irregular texture. The key psychotic features of the delusional system in DBDD are fixed delusional beliefs about appearance and delusional ideas of reference (referential thinking). Patients believe that other people take special notice of the supposed defect, and do not just pay attention to it but "react" and "behave" correspondingly: stare at it, talk about it behind patient's back, laugh at or even mock them.

Delusional behaviour in dermatological DBDD comprises seeking care from cosmetic dermatologists and surgeons to get rid of a defect. Due to lack of insight, patients believe that only a cosmetic procedure or an operation may change the situation. As a result, the patients' medical history may be full of clinically unnecessary dermatological treatments, cosmetic and surgical interventions or even self-mutilations. The paradox is that the results of such procedures may lead to complications that objectively are disfiguring. However, often those are not perceived by patients as a cosmetic problem or are perceived as less problematic than the primary complaint. Due to referential thinking, delusional behaviour may also lead to social isolation (avoidance of situations that may cause others to pay attention to their appearance) and camouflaging of the perceived defect (make-up, special clothes, glasses, etc.).

In *Delusional Olfactory reference syndrome (DORS),* there is a delusional belief that one emits a malodorous smell. As in DBDD (see above), DORS also has its non-delusional equivalent not classified as a psychotic condition but attributed to obsessive-compulsive or related disorders in DSM-5 and ICD-11. Thus, preoccupation with offensive odour may also encompass a range of insight.

The delusional system in dermatological DORS includes central beliefs about the individual's own skin as a definite source of an unpleasant smell. The entire skin may be "involved" or there could be some specific sites, mainly the groin, armpits, or feet. Patients may complain that their sweat has changed its physiological properties and produces such an odour or that the entire body metabolism has altered to produce the smell of the skin. Delusional beliefs about mucosa, i.e. oral (delusional halitosis) or vaginal odour, may also occur.

As in DBDD, there are ideas of reference corresponding to offensive body odour. In DORS, they focus on the impact of an "odour" on others (i.e. the conviction that people are taking notice, judging, or talking about the odour, especially in confined spaces, e.g. in trains, buses, elevators etc). Patients may perceive others' behaviours as caused by odour impact (delusional misinterpretation), including "evidence" of others' desire to stop any social interaction as soon as possible. Patients "notice" others' gestures indicating reaction to the odour, and insist others try to escape and avoid them in situations of any social interaction (e.g. those close to a patient "wrinkle their noses and move away", "touch their nose", "open a window", etc.).

Delusional behaviour includes attempts to camouflage, alter, or prevent the perceived odour.

Patients may wash excessively, change clothes with more than usual frequency, or overuse perfumes and deodorants. Referential thinking leads to avoidance of social situations, sitting at a distance from others, minimizing movement in an attempt "not to spread the odour" etc.

In *Illness delusions or delusions of disease*, there is a delusional belief that one has a specific illness. Delusions of disease typically include transmitted infections, especially venereal diseases (syphilis, gonorrhoea, in last decades especially HIV, etc.).

The Delusional system may comprise the conviction that one is infected. Typically, patients describe in detail how and when they caught the precise pathogen. Usually, they are at low risk of STDs and may cite a single sexual contact, they believe is the cause, that took place decades ago or describe some exotic ways of disease transmission. They appeal to medical literature describing signs and symptoms of the infection to "prove" the diagnosis. Patients often provide delusional explanations of negative medical and laboratory findings that fail to prove infection, e.g. "the smears were taken in periods of latent infection", "the procedure of blood sampling was performed improperly", etc.

Delusional behaviour includes "medical odyssey" as a search for a doctor able to confirm their delusional beliefs. As a result, they demand and often get multiple retests. Another option is a struggle against the infection. They repeatedly take courses of antibiotics or antiviral medications with no or minimal effect. Patients also try to prevent others from becoming infected. They may not just avoid sexual contacts but develop a system of preventive measures including disinfection of household articles and evading any physical contacts with relatives. Patients may even abandon their family/house and move to a separate apartment ("delusional migration").

Dermatological symptoms

Patients with DD of somatic type in dermatology usually complain of changes in their skin. However, it is important to distinguish subjective complaints (skin irregularity, skin odour etc.) and objective self-inflicted lesions as a result of delusional behaviour or underlying minimal dermatological conditions (e.g. mild acne). In DBDD, patients' skin may exhibit a picture of the consequences of multiple cosmetic procedures (scars, pigmentation, etc) and self-inflicted lesions performed in an attempt to improve skin appearance but in fact making it worse. In DORS, the skin is usually intact but in some cases may exhibit consequences of excessive cleansing in attempts to get rid of an odour (skin dryness, irritation, or contact dermatitis), similar to that seen in excessive washing in OCD. In illness delusions or delusions of disease, skin is typically unaffected but in cases with delusional behaviour, focused on genitalia disinfection procedures, corresponding local changes may occur (e.g. dermatitis due to overuse of local antiseptics). Thus, at first glance, skin lesions in DDs of somatic type in dermatology may look very similar to those observed in dermatitis artefacta and skin picking disorder (see corresponding chapters), but the underlying precise psychiatric pattern of self-mutilation is quite different and should be recognized.

Systemic symptoms
Patients with DBDD typically do not have general symptoms and complaints are limited to the perceived deformity. Patients with DORS and illness delusions or delusions of disease may complain of fatigue, dizziness, or malaise depending on the exact delusional system. In DORS, they may think that the skin odour is a result of a general metabolic disturbance of the whole body and as a result other systemic symptoms occur. In illness delusions or delusions of disease, they may focus on the idea of a total organism involvement in the infectious process and express any somatic complaints depending on "damage" to the preferred organ (heart, brain, intestines, etc).

Environmental Presentations
In DBDD and DORS, environmental presentations are rare. But patients with infectious illness delusions or delusions of disease may complain of infections of their immediate environment. In such cases, the delusional behaviour may be very similar to delusional infestation (see Chap. 12). They regularly change bedding, clothes, or furniture, disinfect their apartment and repetitively ask relevant services for decontamination. They may believe that relatives are also infected and make them take courses of antibiotics and antiviral medications or even some toxic non-conventional substances (e.g. veterinarian or industrial).

Practice Point
Pay attention to any delusional behaviours involving self-management as the later may have toxic systemic effects. Activities driven by delusional beliefs and focused on others may also be potentially dangerous for close relatives (be aware of safeguarding issues if it involves children in their care) or medical staff.

Why DD of Somatic Type Is so Debilitating

Self-referential thinking Ideas of reference may lead to severe interpersonal problems and social isolation. DBDD and DORS patients may give up their studies or jobs due to perceived "negative attitudes" to imagined disfigurement or odour "expressed by" classmates or colleagues. Some patients with infectious illness delusions or delusions of disease may exclude any personal relations being convinced that a pathogen is able to transmit easily during routine domestic activities.

Self-damage Some DBDD patients, being disappointed with the results of cosmetic procedures applied by professionals, may try to "fix" their defect or disfigurement themselves with tweezers or chemicals. As a result, real skin disfigurement appears or complications develop (scarring, secondary infection, bleeding). Some DORS patients overuse cleansers to release their skin from the odour, that then causes further skin irritations. DORS patients may ingest inappropriate substances to get rid of the odour if they believe the source is inside the body due to metabolic changes. Patients with illness delusions may overdose antibiotics or other medications to "cure" their infection.

Threat to those around Patients with DBDD may be a threat to cosmetologists, being dissatisfied with the results of their defect or disfigurement correction. As a result, they may not just threaten revenge but also commit physical violence. Patients with illness delusions, who become convinced that their relatives are also infected, may make them go in for inappropriate treatments with a high chance of adverse effects and negative health or life consequences. Some DD patients may also have litigious ideas related to inappropriate cosmetic or medical procedures done. They can place a heavy burden on medicolegal services with multiple claims to the authorities. It may be necessary to consider safeguarding procedures where patients who have care of children apply potentially damaging 'treatments' to their children because of their delusional beliefs that the child is similarly affected.

High rate of psychiatric comorbidity Many patients with DD (nearly 50%) also have depressive symptoms, and comorbid anxiety is also frequent. This may lead to a higher risk of suicidality.

Practice Point
If depressive symptoms are evident, do not hesitate to ask about suicidal ideas and intent.

Epidemiology and Aetiology

Prevalence Epidemiological data about DD are very sparse, as individuals with isolated delusions are often able to function unnoticed in the community and they rarely present themselves to mental health services. The DD lifetime prevalence is considered to be about 0.2%. In the community, point prevalence is 0.03%. Among psychiatric inpatients, it is estimated to be about 2–4%. In some special settings, DD of different types may be accumulated (DBDD in cosmetology, DI in parasitology and dermatology, etc). In prison, point prevalence of DD is reported to be as high as 0.24%, and the most common type of DD content there was persecutory (63.6% of cases), followed by mixed (18.2%), grandiose (14.5%), and somatic (3.6%). In psychiatric settings, the main delusion topic in DD is also persecutory (60.5%) followed by somatic (27.9%), delusional jealousy (7%), and erotomania

(4.7%). There is a lack of data about the prevalence of DD in dermatological settings, except DD of somatic type presented as delusional infestation (see Chap. 12).

Epidemiology In DD, the sex ratio is almost equal (1:1). In general, men are younger than women. The mean age at onset is between 35 and 55 years, 33.8 years in males and 46.4 years in females. DD of jealous type is considered to be more common in males than in females whereas DD of somatic type with delusional infestation and DBDD are more frequent in females (see also DI chapter).

Course and duration of illness DD onset may be acute or subacute, but is typically insidious. Although the severity of the delusion may fluctuate in the disease course, in most cases it is unremitting and chronic. Accordingly, DD is called *persistent delusional disorder* in ICD-10. The mean duration of illness, as published, ranges from a minimum of 3 months to a maximum of 37 years.

Diagnostic Process

General considerations As in other delusional conditions, in DD it is futile to persuade or get into an argument with patients. The thing that is of real value and importance is to establish a trusting relationship with a patient and his/her relatives (if available); however, this may be time-consuming and requires some patience and flexibility. It is beneficial to communicate in a non-confrontational manner with an engaged and interested expression trying to elucidate as much detail of the delusional system and behaviour as possible. Some closed questions about particular thoughts and behaviours typical for DD (see above) may improve this trust. This helps to show that a doctor is aware of the problem and does not consider it as "fake".

There is also an ethical and in some countries the legislative problem of feedback to patients about their condition. It seems that there is no universal approach. There is always a dilemma in deciding whether to announce the DD diagnosis, as this may lead to a loss of trust in the therapeutic relationship, lack of adherence to treatment and potentially subsequently a worse prognosis. Alternatively, a gradual approach may be taken, e.g. referring to the necessity of additional examinations, including psychiatric evaluation. Some general consideration that "an involvement of the nervous system is possible and it should be excluded" may be sufficient at the initial stage of patients' management. Anyway, the approach provided by a dermatologist (or other medical doctors) and a psychiatrist may differ in those circumstances. For example, a consultation-liaison psychiatrist may focus on comorbid psychiatric conditions (e.g. depression, anxiety, insomnia, etc.) related to delusional beliefs.

Diagnostic process The diagnosis of DD is made clinically. No relevant biological markers are available in routine clinical practice. Thus, first of all, it is important to exclude other organic causes for the patient's complaints (e.g. underlying skin disease, e.g. acne, in DBDD; metabolic diseases in DORs or infections in illness delu-

sions). However, even if such causes do really exist, this does not fully exclude DD. There are many DD cases triggered and maintained by some minor medical conditions or disfigurement. This may be illustrated in the joke by Joseph Heller: "Just because you are paranoid doesn't mean they aren't after you". Thus, the reality or falsity of the belief is not a main diagnostic criteria. It is more important to evaluate the patient's explanatory model that may reveal a delusional system. So, this is not a question of true or false, but how patients selectively choose and interpret facts which support their beliefs and interpret away facts that do not.

Differential diagnosis It includes other neuropsychiatric conditions and somatic illnesses. The symptoms are not due to another disorder or disease attributed to mental, behavioural, or neurodevelopmental disorders that could present with delusions, like schizophrenia, schizoaffective disorder, delirium, dementia, psychotic disorder due to another medical condition and substance/medication-induced psychotic disorder, depressive and bipolar disorders, obsessive-compulsive and related disorders (Table 26.1).

Table 26.1 Differential diagnosis for DD and other psychiatric conditions

Psychiatric diagnoses to be excluded	Features that differ from DD
Schizophrenia	Can be distinguished from DD due to other characteristic symptoms of schizophrenia, including disorders of thinking (e.g. disorganization), perception (e.g. hallucinations), self-experience (e.g. the experience that one's feelings, impulses, thoughts, or behaviour are under the control of an external force), cognition (e.g. impaired attention, verbal memory, and social cognition), volition (e.g. loss of motivation), affect (e.g. blunted emotional expression), and behaviour (e.g. behaviour that appears bizarre or purposeless, unpredictable, or inappropriate emotional responses that interfere with the organization of behaviour).
Depression, bipolar disorders, and schizoaffective disorder	Can be distinguished from DD by the temporal relationship between affective symptoms and delusions that may occur exclusively during mood episodes, e.g. in depressive or bipolar disorder with psychotic features, i.e. if the psychotic symptoms are better explained by an affective episode (depression, mania, or mixed episode).
Delirium or psychotic disorder due to another medical condition	Can be distinguished from DD due to medical history, sudden onset, physical examination, paraclinical or laboratory tests that reveal underlying medical condition with potential physiological influence on the brain. There is a temporal relationship between the decompensation of medical condition and psychosis (they begin and end at the same time), and effective management of the medical condition often reduces the severity of psychotic symptoms.
Substance/ medication-induced psychotic disorder	Can be distinguished from DD by the chronological relationship of substance use to the onset and remission of the delusional beliefs; often transient and single delusional ideas with no signs of developed delusional system; often associated with hallucinations. History of substance use and the nature of the substance being used, supplied with laboratory tests, such as a urine drug screen or a blood alcohol level, may distinguish condition.

(continued)

Table 26.1 (continued)

Psychiatric diagnoses to be excluded	Features that differ from DD
Dementia	Can be distinguished from DD due to other characteristic symptoms of dementia, including gradual, significant cognitive decline from a previous level of performance (complex attention, executive function, learning and memory, language, perceptual-motor, or social cognition)
Obsessive-compulsive disorder	Can be distinguished from DD due to preserved insight into the excessiveness and unreasonableness of obsessions and/or compulsions (except cases with poor or no insight) that are intrusive, experienced as distressing and "ego-dystonic" with no signs of delusional system development. In OCD, the patient has intrusive thoughts or imagery, identifies them as senseless, resists them, but resistance causes anxiety.

To finish the discussion of differential diagnoses of other DD of somatic type in dermatology, this is a list of medical conditions to be excluded in DORS:

- Dermatological (hyperhidrosis)
- Genito-urinary (rectal abscess, fistulae)
- Metabolic (trimethylaminuria)
- Otolaryngeal (halitosis)
- Dental (abscess)
- Neurological (temporal lobe epilepsy, arteriovenous malformations, Parkinson's disease)

Treatment

General considerations According to most data, the medications of choice in DD, as in other disorders with prominent delusions, are antipsychotics. The key problem of management in DD is compliance as there is no insight or awareness of a psychiatric origin of symptoms. As a result, it is difficult both to initiate appropriate treatment and maintain long-term adherence. However, there is data that DD may have a good prognosis if appropriate adherence can be achieved.

Pharmacological treatment

The only available RCT meta-analysis in the Cochrane Database (2015) states there is a paucity of high-quality randomized trials on DD, and there is currently insufficient evidence to make evidence-based recommendations for treatments of any type for people with DD. The limited data that were found could not be generalized to the population of people with DD. Thus, it is suggested that until further evidence is found, it seems reasonable to offer treatments which have efficacy in other psychotic disorders. This means that antipsychotics are medications of choice. Olanzapine, risperidone, quetiapine, aripiprazole, ziprasidone, amisulpride, sulpiride and haloperidol are medications most frequently used for

DD. However, there could be a need for augmentation with other psychotropic medications: antidepressants (if depressive symptoms are prominent) and hypnotics (in insomnia cases).

Regarding problems with adherence, long-acting injectable antipsychotics (depot antipsychotics) may be beneficial. Recent studies show that DD patients treated with such long-lasting medications show higher attendance rates for outpatient appointments and a lower rate of prescriptions for other psychotropic drugs. Thus, these drugs are considered as a promising option in the treatment of DD.

The general approach to the pharmacological treatment in DD does not differ greatly from that followed in patients with delusional infestation (see Chap. 12).

Non-pharmacological interventions Data about the efficacy of psychosocial interventions in DD are limited. There is a single RCT, considered to be of a high quality, that compared cognitive-behavioural therapy (CBT) and attention placebo control (APC). It showed a significant posttreatment change in several belief dimensions for both APC and CBT, e.g. the posttreatment decrease in the "strength of belief" parameters was 40% in CBT and 28% in APC. The most significant change in both groups lay in an increased ability to control actions and communications related to the belief. CBT also improved outcomes on depression and self-esteem. However, neither CBT nor ACP succeeded overall in reducing the strength of conviction to zero. Thus, from a practice point of view, psychotherapy, particularly CBT, may be considered as a good adjunct to antipsychotic treatment. Such an approach has been shown to be beneficial in several less good quality trials, most of them compared CBT with standard psychiatric care or treatment as usual. Regarding targets of CBT in delusional beliefs, there is a suggestion that it should be focused not primarily on belief reassurance, but on "allowing the patient a free rein to talk, gradually making the link between external stressors, emotion, and beliefs, leading to a joint exploration of alternative explanations for certain beliefs", i.e. trying to modify the valence of beliefs (that is, the emotion attached to the beliefs) rather than their content.

Discussing the diagnosis and management with a patient Patients with DD are convinced that their beliefs are true. So, as mentioned above, there is no value in arguing with them. However, very often they are open to discussion of their delusional ideas if they feel they have encountered an interested listener. Thus, a non-confrontational and empathic approach is a key not just for making the diagnosis, but also for discussing the diagnosis with the patient. Regarding the patient's ability to understand the diagnosis, it may be primarily announced in general as a "disorder of the nervous system", "generally underinvestigated", "rather rare but empirically proved to be effectively treated by neuroleptics" (see Chap. 12).

Practice Point
As engagement and adherence are the challenging issues in DD management, psychosocial interventions, performed in a non-confrontational manner, should be primarily focused on establishing a positive therapeutic relationship.

Practice Point
As DD is attributed to a primary psychotic disorder and high-quality treatment evidence is limited, it is suggested to use antipsychotics as first-line medications.

Prognosis

Existing prognosis data Primarily, DD has been considered difficult to manage due to high rates of treatment resistance and relatively poor prognosis. A lack of adherence to medication is considered as one of the most common factors associated with a poor response. However, recent retrospective analyses, case series and several trials suggest that DD has a "moderate", "acceptable", and even "good prognosis" if treated adequately. There are even some benign cases with a fast response and full remissions. Among predictors of better response are non-prominent hallucinations consistent with the content of delusions.

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