DOI: https://doi.org/10.17816/nb634695



Schizophrenia spectrum disorders: does the non-psychotic schizophrenia exist?

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ABSTRACT

Current understanding of schizophrenia comes from Kraepelin's dementia praecox doctrine, a disease that combined Kalbaum's catatonia, Hecker's hebephrenia and chronic delusional psychoses and later renamed to schizophrenia thanks to Bleuler. Since the beginning of the XX century, the systematics of schizophrenia has undergone a serious evolution and narrowing of diagnostic boundaries, which is reflected in various revisions of the ICD and editions of the DSM. Currently, the existence of non-psychotic forms of schizophrenia, mainly manifested by negative symptoms, is being questioned. In addition, the need for diagnostic differentiation of schizophrenia from autism spectrum disorder, which have only external similarities, but require fundamentally different therapeutic interventions, is emphasized.

Keywords: *dementia praecox*; schizophrenia; schizophrenia spectrum disorders; schizotypal personality disorder; ICD-10; ICD-11; DSM-5; antipsychotics.

To cite this article:

Sivolap YP, Portnova AA. Schizophrenia spectrum disorders: does the non-psychotic schizophrenia exist? // Neurology Bulletin. 2024;56(3):240–246. DOI: https://doi.org/10.17816/nb634695



COOVECTOR

Accepted: 08.08.2024

Published online: 17.09.2024

240

DOI: https://doi.org/10.17816/nb634695

Расстройства шизофренического спектра: существует ли непсихотическая шизофрения?

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АННОТАЦИЯ

Современные представления о шизофрении начинаются учением Крепелина о dementia praecox — болезни, объединившей кататонию Кальбаума, гебефрению Геккера и хронические бредовые психозы и в дальнейшем получившей наименование шизофрении благодаря Блейлеру. С начала XX века систематика шизофрении претерпела серьёзную эволюцию и сужение диагностических границ, что нашло отражение в различных пересмотрах Международной классификации болезней (МКБ) и редакциях Диагностического и статистического руководства по психическим расстройствам (Diagnostic and Statistical Manual of Mental Disorders, DSM). В настоящее время ставится под сомнение существование непсихотических форм шизофрении, преимущественно проявляющихся негативными симптомами. Кроме того, подчёркивается необходимость диагностического отграничения шизофрении от расстройства аутистического спектра, обладающих лишь внешним сходством, но предполагающих принципиально разные терапевтические вмешательства.

Ключевые слова: dementia praecox; шизофрения; расстройства шизофренического спектра; шизотипическое расстройство личности; МКБ-10; МКБ-11; DSM-5; антипсихотики.

Как цитировать:

Сиволап Ю.П., Портнова А.А. Расстройства шизофренического спектра: существует ли непсихотическая шизофрения? // Неврологический вестник. 2024. Т. 56, № 3. С. 240–246. DOI: https://doi.org/10.17816/nb634695

Рукопись получена: 30.07.2024

Рукопись одобрена: 08.08.2024

Опубликована online: 17.09.2024

241

DOI: https://doi.org/10.17816/nb634695

Шизофрения спектрының тайпылышлары: психотик булмаган шизофрения бармы?

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АННОТАЦИЯ

Шизофрения турындагы заманча күзаллаулар Крепелинның dementia praecox — Кальбаум кататониясен, Хеккер гебефрениясен hәм хроник саташу психозаларын берләштергән авыру турындагы тәгълиматы белән башланып, соңрак Блейлер тәкьдиме белән шизофрения исемен ала. ХХ гасыр башыннан шизофрения систематикасының житди эволюциягә дучар булып, диагностика чикләренең тараюы Авыруларның халыкара классификацияләрендә (МКБ) hәм Психик тайпылышлар буенча диагностика hәм статистика кулланмасы (DSM) басмаларында чагылыш таба. Хәзерге вакытта шизофрениянең күбесенчә тискәре симптомнарда чагыла торган психотик булмаган формалары булу шик астына алына. Моннан тыш, шизофренияне фәкать тышкы билгеләре белән белән генә шуңа охшаган, әмма бөтенләй икенче төрле дәвалау алымнары куллану тиешле аутизм спектры тайпылышларыннан аерып карау кирәклегенә аерым игьтибар ителә.

Төп төшенчәләр: dementia praecox; шизофрения; шизофрения спектры тайпылышлары; шәхеснең шизотипик тайпылышы; МКБ-10; МКБ-11; DSM-5; антипсикотиклар.

Өземтәләр ясау өчен:

Сиволап Ю.П., Портнова А.А. Шизофрения спектрының тайпылышлары: психотик булмаган шизофрения бармы? // Неврология хәбәрләре. 2024. Т. 56, Чыг. 3. Р. 240–246. DOI: https://doi.org/10.17816/nb634695

Кулъязма алынган: 30.07.2024



Кулъязма хупланган: 08.08.2024

Бастырылган online: 17.09.2024

The history of modern ideas on schizophrenia dates back to the end of the XIX century, which, to a certain extent, coincides with the emergence of a nosological trend in psychiatry, which is primarily associated with the names of Karl Kahlbaum and Emil Kraepelin. Karl Kahlbaum presented a description of catatonia, which he regarded as an different disease and was later named Kahlbaum's disease. A few years later, Ewald Hecker, a junior colleague and student of Kahlbaum, identified hebephrenia as a special kind of insanity in young patients with an unfavorable prognosis¹. In 1898, Emil Kraepelin, at the XXIX Congress of psychiatrists of Southeastern Germany in Heidelberg and a year later in the famous sixth edition of his textbook of psychiatrycombined Kahlbaum's catatonia, Hecker's hebephrenia, and chronic delusional psychoses with systematic evolution into what he thought was a single disease-dementia praecox, or early (premature) dementia².

At the beginning of the 20th century, the Swiss psychiatrist Otto Diem proposed adding another form to the list of conditions collectively termed *dementia praecox–dementia simplex*, or simple dementia that does not manifest psychotic symptoms³.

In 1911, another, and much more famous, Swiss psychiatrist, Eugen Bleuler, in the monograph "dementia praecox oder Gruppe der Schizophrenien" ("dementia praecox, or a group of schizophrenias")⁴ coined the word "schizophrenia," which quickly replaced the phrase "early dementia" in professional terminology. Bleuler, with his characteristic modesty, emphasized that his concept of schizophrenia was nothing more than an application of the ideas of Sigmund Freud to Emil Kraepelin's teaching on dementia praecox.

Emil Kraepelin's *dementia praecox* and Eugene Bleiler's schizophrenia can hardly be considered identical clinical phenomena: Kraepelin described mainly psychotic disorders with an unfavorable course and outcome, whereas Bleiler's descriptions included relatively benign disorders, including non-psychotic, cases. Eugen Bleiler identified the primary, or obligate, symptoms of schizophrenia, without which diagnosis is impossible and which later became known as the "four A's": autism, associative disorders, affective flattening, and ambivalence. Bleuler regarded psychotic symptoms as delusions and hallucinations as secondary and optional. Additionally, he admitted and emphasized the possibility of the existence of non-psychotic forms of schizophrenia, including its latent variants, which served as one of the involuntary reasons for the emergence of a very ambiguous concept of sluggish (mild) schizophrenia in the domestic psychiatry of the Soviet period.

Throughout the 20th century up to the first quarter of the 21st century, national and international classifications distinguished, with some variations and changes, such forms of schizophrenia as catatonic, hebephrenic, paranoid, and simple, as well as residual schizophrenia and postschizophrenic (postpsychotic) depression.

The International Classification of Diseases of the 10th revision (ICD-10) traditionally includes paranoid, hebephrenic, and catatonic forms of schizophrenia, as well as simple schizophrenia, manifesting as a gradual increase in negative symptoms, oddities in behavior, and non-compliance with the requirements of society without obvious psychotic symptoms⁵ [1].

The ICD-11 does not recognize any of the listed syndromic forms of schizophrenia. The first episode of schizophrenia, schizophrenia with multiple episodes, and continuous schizophrenia are highlighted here⁶, and catatonia is removed from the schizophrenia or other primary psychotic disorders section into an independent section [2]. In our opinion, this indicates significant progress in the 11th revision relative to the 10th one.

The diagnosis of schizophrenia in the Diagnostic and Statistical Manual of Mental Disorders, 5th edition (DSM-5) is based on six formalized criteria (A, B, C, D, E, and F). The first basic criterion (A) includes five signs:

2) hallucinations;

3) disorganized⁷ speech (for example, frequent derailment⁸ or incoherence);

4) significantly impaired or catatonic behavior, and

5) negative symptoms.

One is diagnosed with schizophrenia if he or she exhibits at least two of the five signs listed above, but at least one of them should relate to the first three [3].

¹⁾ delire;

¹ Additionally, Ewald Hecker introduced the concept of cyclothymia into the psychiatric nomenclature (note by the authors, as well as subsequent notes).

² The Latin construction *dementia praecox* used by Emil Kraepelin, which was firmly established first in the German and later in the international psychiatric lexicon, was first proposed by the Czech psychiatrist Arnold Pick, founder of the Prague neuropathological school, although Benedict Morel used it long before both of them, but as the French equivalent—démence précoce.

³ Later, the term *dementia praecox* would be replaced by the term schizophrenia simplex, or simple schizophrenia, characterized exclusively by negative symptoms without positive (psychotic) manifestations.

⁴ As indicated by the very title of the monograph, Bleuler regarded schizophrenia as a set of diseases.

⁵ Apart from the listed forms, undifferentiated and residual schizophrenia and postpsychotic depression are distinguished in ICD-10; "other schizophrenia" (other schizophrenia) and unspecified schizophrenia are also mentioned.

⁶ The above systematics of schizophrenia, as well as several other mental disorders, was borrowed from the DSM-5.

⁷ In the original, disorganized.

⁸ In the original, derailment.

243

Schizophrenia in the DSM-5 is included in the section "schizophrenia Spectrum and Other Psychotic Disorders". Apart from schizophrenia, this section presents delusional disorder, short-term psychotic disorder, schizophrenic-like disorder, schizoaffective disorder, psychotic disorder caused by a psychoactive substance or drug, and psychotic disorder associated with another medical cause.

Additionally, the list of disorders similar to schizophrenia includes schizotypal disorder, and its affiliation to personality disorders is specified in parentheses as follows: schizotypal (personality) disorder. A detailed description of schizotypal disorder is given in cluster A of the section "Personality disorders" together with paranoid and schizoid personality disorders, and, in this cluster, the term is used already without brackets as follows: schizotypal personality disorder [3].

Owing to the similarity of the clinical manifestations of schizotypal personality disorder (as well as schizoid and paranoid personality disorders) with the symptoms of schizophrenia, there is not a single formalized diagnostic criterion in the DSM-5 that is common to two or more of the three mental disorders. Thus, from the point of view of modern psychiatric systematics given in the DSM-5, nonpsychotic mental disorders similar to schizophrenia and belonging to the category of personality disorders cannot serve as a basis for the diagnosis of the latter.

The same rule applies to the largely outdated, but for a number of non-medical reasons, still relevant ICD-10, in which schizophrenia and schizotypal disorder⁹ appear in neighboring, but separate sections — F20 and F21.

There is a notable difference between the original (Englishlanguage) version of ICD-10 (International Classification of Diseases), posted on the website of the World Health Organization (WHO), and the version adopted in the Russian Federation. In the original version, under section F21 (Schizotypal disorder) non-psychotic conditions were included in this category, which, in the previous 9th version of the ICD, were regarded as varieties of schizophrenia, including pseudoneurotic and pseudopsychopathic forms of schizophrenia. There is no equal sign between schizophrenia and schizotypal disorder; there is no pseudoneurotic and pseudopsychopathic schizophrenia in the ICD-10. In the Russian version of ICD-10, pseudoneurotic (neurosislike) schizophrenia and pseudopsychopathic (psychopathylike) schizophrenia are¹⁰ regarded as variants of schizotypal disorder and are assigned the codes F21.3 and F21.4, respectively, which are absent in the original official version of ICD-10.

Thus, the Russian version of ICD-10, to a certain extent, equates schizotypal disorder and non-psychotic forms

of schizophrenia, which causes an unjustified and inconsistent expansion of the diagnostic boundaries of the disease.

An unjustified diagnosis of schizophrenia (primarily its nonpsychotic forms) in patients with autism spectrum disorder is common in Russian psychiatric practice. Apparently, one of the main reasons for mixing two fundamentally different types of mental disorders is that Eugen Bleuler, the author of the terms "schizophrenia" and "autism," attributed the latter to the obligatory manifestations of the former, as mentioned above. The external similarity between special behavioral disorders in children with a certain type of developmental disorder of the nervous system and Bleuler's autism led Leo Kanner to propose the concept of early childhood autism in 1943¹¹ [4].

The inadmissibility of diagnosing schizophrenia in patients with autistic manifestations is due not only to differences in the biological nature and formal psychopathological affiliation of these different conditions, but also to a fundamental difference in therapeutic approaches. Antipsychotics are the main and sometimes the only class of pharmacological agents in the treatment of schizophrenia, whereas no drugs superior to placebo in reducing symptoms of autism have been found so far. Psychological and corrective interventions play the main role in helping patients with autistic disorder, and antipsychotics (exclusively risperidone and aripiprazole) are prescribed only for comorbid behavioral disorders. Antipsychotics prescribed for autism spectrum disorder without comorbid behavioral disorders are ineffective, exacerbate the cognitive deficit inherent in many patients, and, in many cases, worsen their physical health.

The German psychiatrist Kurt Schneider made a major contribution in the diagnosis of schizophrenia by identifying key signs of this disease, which the author called symptoms of the first rank. The symptoms of schizophrenia of the first rank, according to Kurt Schneider, are the basis for the diagnosis of this disease according to the formalized criteria of ICD-10, but are regarded in ICD-11 as insufficiently specific for schizophrenia, in line with the opinion of the expert community. Kurt Schneider, who, apparently, was not inferior to Eugene Bleiler in scientific modesty, insisted that, in the diagnosis of schizophrenia, it is much more important to use the above-mentioned famous Bleiler "four A's" as obligatory signs of the disease, not the symptoms of the first rank described by himself.

The authors of this article pay tribute to the impressive and, in many ways, still relevant scientific legacy of Kurt Schneider, but they find it difficult to accept his last thesis as evidence of his modesty. In our opinion, and in accordance

⁹ In the 2019 version of ICD-10 posted on the WHO website, the word "personality" is not used in relation to schizotypal disorder.

¹⁰ In the traditional Russian systematics of mental disorders, neurosis-like schizophrenia and psychopathic schizophrenia are regarded as clinical variants of sluggish (mild) schizophrenia.

¹¹ Modern psychiatric taxonomies (ICD-10 and DSM-5) do not use the adjectives "early" and "childhood" and only refer to autism spectrum disorder, although ICD-10 still uses the term "childhood autism".

EDITORIAL S

with modern ideas about the boundaries of schizophrenia, it is precisely such psychotic manifestations as auditory hallucinations, the sound of thoughts, delusional perception, and other symptoms of the first rank described by him that allow distinguishing this disease from similar non-psychotic mental disorders, including schizotypal personality disorder.

The consequences of unnecessarily expanding the diagnostic boundaries of schizophrenia include avoidable stigmatization, serious reputational damage to the mental health professional community, and reluctance of patients to seek mental health care.

Unjustified prescription of antipsychotics to patients with personality disorders or autism (including those associated with misdiagnosis) can lead to poor physical health and a high risk of premature death, primarily due to cardiovascular (coronary heart disease and myocardial infarction) and metabolic (obesity, type 2 diabetes, and atherosclerosis) causes [5].

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* Anna A. Portnova, MD, Dr. Sci. (Med.); Chief Researcher; address: 74 Michurinsky Prospect, 119602 Moscow, Russia; ORCID: 0000-0001-5938-0202; eLibrary SPIN: 5899-0069; e-mail: aapserbsky@mail.ru Thus, the cost of the traditional approach in psychiatryfailure to recognize new disease forms, such as autism spectrum disorder and pushing the boundaries of schizophrenia, — may lead to loss of human lives. The purpose of this publication was not so much to renew a long discussion on one of the difficult topics of psychopathology (in which, as the fundamental divergence of opinions of the discussing parties shows, one cannot count on professional consensus), but rather to raise ethical questions in clinical psychiatry.

ADDITIONAL INFORMATION

Funding source. This publication was not supported by any external sources of funding.

Competing interests. The authors declare no conflicts of interests. **Authors' contribution.** The equal part of both authors.

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