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The causes, diagnostics, and treatment of psychogenic blindness – a systematic review.

Przyczyny, diagnostyka i leczenie ślepoty psychogennej – przegląd systematyczny.

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Abstract

Introduction: Conversion disorders are characterized by the presence of motor or sensory dysfunction, resulting in significant discomfort or disability, the occurrence of which is not justified by any somatic disease. Psychogenic blindness, a specific manifestation of conversion disorders, refers to the loss or impairment of vision that cannot be attributed to any organic abnormalities in the eyes or visual system. The exact mechanisms underlying psychogenic blindness are not fully understood, but it is believed to involve complex interactions between the brain, visual pathways, and psychological processes. This systematic review aims to summarize reported cases of blindness in conversion disorders.

Material and methods: A comprehensive search of the MEDLINE and Embase databases identified 13 relevant articles reporting a total of 17 patients with psychogenic blindness.

Results: The risk and triggering factors for psychogenic blindness included acute stress (for example acute physical injury), chronic stressful situations (such as chronic health problems, disruptions in family relationships), and multiple factors including coexisting psychiatric conditions (like anxiety or other conversion disorders). The course of blindness varied among patients, with duration ranging from hours to years. Diagnostic methods involved thorough ophthalmic and neurological examinations, brain imaging, and psychiatric evaluations.

Conclusions: This review provides valuable insights into managing patients with psychogenic blindness. The findings highlight

the need for a multidisciplinary approach involving ophthalmologists, neurologists, and mental health specialists. Further research is required to elucidate the underlying mechanisms and develop effective treatment strategies for individuals with psychogenic blindness.

Keywords: psychogenic blindness, conversion disorders, causes

Streszczenie

Wstęp: Zaburzenia konwersyjne charakteryzują się występowaniem dysfunkcji ruchowych lub czuciowych, powodujących znaczny dyskomfort lub niepełnosprawność, których występowania nie uzasadnia żadna choroba somatyczna. Ślepota psychogenna, będąca specyficzną manifestacją zaburzeń konwersyjnych, odnosi się do utraty lub upośledzenia wzroku, którego nie można przypisać żadnym organicznym nieprawidłowościom w układzie wzrokowym. Dokładne mechanizmy leżące u podstaw ślepoty psychogennej nie są w pełni poznane, ale uważa się, że obejmują złożone interakcje między mózgiem, drogami wzrokowymi a procesami psychologicznymi. Niniejszy przegląd systematyczny ma na celu podsumowanie opisanych dotychczas przypadków ślepoty w zaburzeniach konwersyjnych.

Materiał i metody: Przez kompleksowy przegląd baz danych MEDLINE i Embase zidentyfikowano 13 stosownych artykułów opisujących łącznie 17 pacjentów ze ślepotą psychogenną.

Wyniki: Do czynników ryzyka wyzwalających ślepotę psychogenną należą zarówno ostre i przewlekłe stany stresowe (ostry uraz fizyczny, długotrwałe problemy zdrowotne, konflikty rodzinne) jak i współistniejące zaburzenia psychiatryczne (stany lękowe lub inne zaburzenia konwersyjne). Przebieg ślepoty u pacjentów był zróżnicowany, trwający od godzin do lat. Metody diagnostyczne obejmowały dokładne badania okulistyczne i neurologiczne, obrazowanie mózgu oraz ocenę psychiatryczną.

Wnioski: Niniejszy przegląd systematyczny dostarcza cennych informacji na temat prowadzenia pacjentów ze ślepotą psychogenną. Praca ta podkreśla potrzebę multidyscyplinarnego podejścia obejmującego wiedzę z zakresu okulistyki, neurologii i psychiatrii. Konieczne są dalsze badania w celu wyjaśnienia mechanizmów leżących u podstaw i opracowania skutecznych strategii leczenia osób ze ślepotą psychogenną.

Słowa kluczowe: ślepota psychogenna, zaburzenia konwersyjne, przyczyny

Introduction

Conversion disorders, also known as functional neurological disorders, are characterized by symptoms of disrupted motor or sensory function, that result in substantial discomfort or disability. However, the symptoms are not due to any confirmed somatic disease and are usually incoherent with the results of a medical examination [1]. Even though not fully understood, conversion disorders have been described broadly, with a prominent, valuable source of knowledge being the work of Prusiński et al. [2]. In the International Classification of Diseases 11 (ICD-11), conversion disorders are listed among "dissociative disorders", which relate to a distinct pathophysiological process, described as partial or total disturbance in the integration of identity, perceptions, sensations, thoughts, affects, memories, and control over physical motions, or behavior [3]. The clinical symptoms range widely and include sensorial impairment, irregular movements, non-epileptic seizures, gait abnormalities, and even sensory deficits like blindness or deafness [1]. The Diagnostic and Statistical Manual of Mental Disorders, 5th edition (DSM-5), classifies conversion disorders as "somatic symptom and related disorders" and presents the following criteria of diagnosis: (i) one or more signs of the disturbance of voluntary motor or sensory function, (ii) symptoms are incompatible with known neurological or medical diseases, (iii) the symptoms or deficits cannot be better explained by a different mental or medical issue, and (iv) the symptoms or deficits cause distress, impairment within the patient's areas of functioning, or necessitate a medical evaluation [4]. These symptoms are functional, of a psychogenic nature – that means that they are not caused by an organic disorder, not intentionally produced, and not under the individual's conscious control. However, conversion disorders should always be differentiated from factitious disorder and malingering [5].

The exact mechanisms underlying conversion disorders are not fully understood. It is believed that psychological factors, such as acute stress or chronic unsolvable problems, can lead to the unconscious expression of distress through physical symptoms, disrupting the brain-body communication pathways. The proposed pathomechanisms of conversion disorders include symptom modeling with favoring of "feedforward" signaling, the limbic system's overactivity, and malfunctioning of the brain regions that give movement the sense of deliberation [6]. The known risk factors for

conversion disorders include trauma, stress, history of abuse in childhood, poor adaptation skills, coexisting psychiatric illnesses, multiple somatic disorders, and low socioeconomic status. The onset of symptoms can be triggered by physical injuries, stroke, or migraines [7]. The diagnosis of conversion disorders involves thorough medical evaluations to rule out any organic causes of the symptoms. Psychological assessments are also conducted to identify any underlying psychological stressors or conflicts. Treatment approaches usually involve a combination of psychotherapy (including cognitive-behavioral therapy and psychodynamic therapy), and sometimes medications to manage associated symptoms, such as anxiety or depression [8, 9].

A specific manifestation of a conversion disorder is psychogenic blindness, also known as functional blindness or hysterical blindness. ICD-11 classifies psychogenic blindness as a dissociative neurological symptom disorder, with visual disturbance (code 6B60.0) [3]. It is a condition, in which an individual experiences a loss or impairment of vision that cannot be attributed to any organic or structural abnormalities in the eyes or visual system. The patients do experience, but are unable to control their visual symptoms, which are caused by a disruption of higher cortical regions involved in visual awareness [10]. It is estimated, that medically unexplained vision loss constitutes 1-5% of cases presenting to an ophthalmology clinic, and among school-aged children, the incidence of non-organic vision loss has been estimated at 1.75% [11, 12]. However, the exact incidence of these disorders has not yet been fully established. The exact mechanisms underlying psychogenic blindness are not fully understood, however, studies indicate that in this group of patients, cognitive disorders typical of organic brain damage occur significantly more often [13]. It is believed that pathomechanisms involve complex interactions between the brain, the visual pathways, and psychological processes. It is hypothesized that there is a functional impairment in the cortico-striatal-thalamic circuits, which are responsible for regulating sensorimotor activity and voluntary movement [14]. A comprehensive evaluation by healthcare professionals, including ophthalmologists, neurologists, and psychiatrists, is necessary to diagnose and differentiate psychogenic blindness from organic visual impairments. If a non-organic cause of vision loss is suspected, there are specific tests that can be useful in uncovering the psychogenic nature of the disease. One of the examples is the use of plano lenses or artificial eye drops and hoping for a placebo effect while reading the Snellen chart (the "magic lens" and "magic drops" tests). Other techniques, which involve fogging, the assessment of stereoscopic vision, the use of red-green glasses, and more, have been described in detail by Moore et al. [12].

Even though known for centuries as "hysteria" [15], modern medicine still understands very little about conversion disorders, especially psychogenic blindness, and a lot of physicians struggle with the diagnosis and treatment of non-organic vision loss. This is most probably due to the insufficient knowledge of medical professionals about the significant impact of psychological welfare on the visual system [16].

Aim

This systematic review aims to summarize the reported cases of blindness in conversion disorders based on cases of patients with a known or suspected cause of blindness.

Material and methods

This review was performed with a systematic approach by searching the MEDLINE and Embase databases with search queries ("conversion disorder" AND "blindness" AND "cause") and ("psychogenic blindness" AND "cause"). A total number of 75 articles were identified with an initial search of the two databases performed from May 22nd, 2023, through May 29th, 2023. After removing duplicates, 63 articles were left for screening. The inclusion criteria were as follows: original articles written in English or Polish describing patients with a conversion disorder manifesting as blindness, with a known or suspected cause. Excluded were review articles and articles, that did not meet the inclusion criteria. After excluding unsuitable papers, 13 articles were included in the systematic review. The PRISMA flow diagram is presented in Figure 1. The 13 reviewed articles reported on a total of 17 patients with psychogenic blindness, which was a manifestation of a conversion disorder.

Results

Based on the reports of 17 patients, we summarized the triggering factors, course of blindness, possible risk factors, diagnostics, and treatment methods of psychogenic blindness.

Triggering and risk factors

To summarize the triggering and risk factors that initiated the onset of psychogenic blindness, we grouped them into categories of psychogenic blindness precipitated by: (i) acute stress, (ii) chronic stress, and (iii) multiple factors.

- Acute stress

Acute stress has been a triggering factor for the onset of psychogenic blindness in 7 out of 17 patients. Two patients, a 21-year-old female, and a 22-year-old male, experienced vision loss with onset before upcoming exams in college or university [11, 17]. Another patient suffered

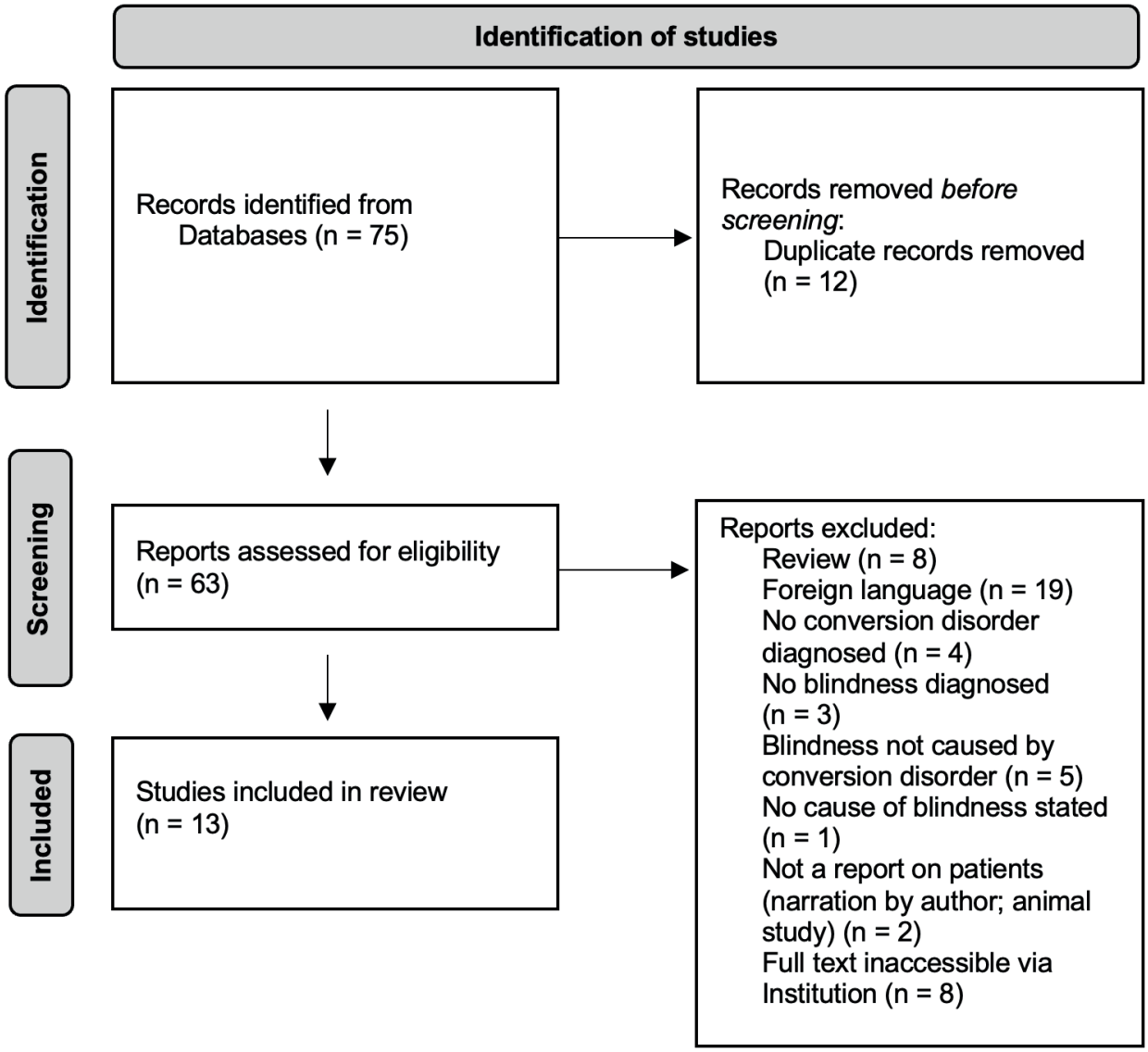

Figure 1. PRISMA flow diagram presenting the method of choosing research included in the systematic review.

from vision loss after a traumatic visual experience: a 48-year-old man looked at his broken finger after a traffic accident, which caused him to faint and lose vision [18]. Cutler et al. described a case of a 25-year-old female, who suffered from multiple personality disorder. Transient, temporary blindness lasting for a couple of hours, along with personality changes, were sometimes precipitated by stress, for example of a newly diagnosed pregnancy or coming back home from the hospital to an unhappy home [19]. Another woman, aged 33, experienced blindness for 13 years, following a non-specified accident (with an initial diagnosis of cortical blindness), and also suffered from multiple personality disorder. However, after 4 years of psychotherapy, the patient started to regain limited, partial vision. What is interesting, is that the regained sight was restricted only to a few of the multiple personalities presented by the patient. The patient went from sight to blindness with the alteration of personality [20].

Acute stress can also be caused by undergoing medical procedures, which precipitated the onset of psychogenic blindness in 2 patients. An 11-year-old girl suffered a 6-hour episode of vision loss caused by the misplacement of an intravenous cannula into the brachial artery in the emergency department, which caused pain, numbness, and pallor of the arm. These symptoms led to agitation and anxiety, prompting the onset of psychogenic, bilateral blindness [21]. Another woman, aged 29, presented with complete blindness after a surgical removal of a cervical spinal cord stimulator under general anesthesia. The blindness appeared on the first postoperative day after waking up from sleep and dissolved spontaneously on the 7th day after surgery [22].

- Chronic stress

In the collected articles, 8 out of 17 patients suffered from conversion disorders manifesting with blindness in response to chronic stress. Werring et al. studied the cases of 5 patients, 4 of whom (females aged 32, 16, 51,

35) underwent chronic stress caused by losing custody over children, losing a job, bad physical condition, and mourning; however, no specific details of the patients' history were given [11]. Rota et al. described a 23-year-old patient with anxiety stemming from complicated family relationships, especially with the mother. The patient also suffered from migraines [23]. Another patient, a 47-year-old woman, presented with bilateral blindness, which developed gradually after brain surgery due to a ruptured aneurysm (which was the presumed cause of vision loss). However, the patient also had marital problems – her husband was a criminal who later left. Psychotherapy and an experimental kind of cognitive therapy helped to relieve the symptoms of blindness, with recovery after 6 months [24].

Two of the reviewed cases described psychogenic vision loss appearing in response to the presumed expectations of the environment, which were a source of chronic stress for the patients. A 10-year-old girl came from a family with congenital glaucoma, where her siblings received more attention due to their developed blindness caused by the familial disease. The patient, even though not diagnosed with glaucoma, claimed to also be blind and persisted to keep her eyes shut [25]. In another case, a 55-year-old woman became blind in response to a false-positive test result assessing anti-retinal antibodies, which led to a false diagnosis of autoimmune retinopathy leading to vision loss [26]. This form of "adjustment" to the environment could have also been caused by stress related to believing in the development of inevitable blindness in the future.

- Multiple factors

Two patients had a history too complex to distinguish a specific triggering factor, and the blindness was most likely prompted by a constellation of factors. A 9-year-old girl was reported to have progressive vision loss, associated with a constellation of psychiatric symptoms, such as bulimia, anxiety, and low self-esteem [27]. Shell et al., on the other hand, presented a case of a 39-year-old woman with multiple co-existing psychiatric and organic health issues. Acute bilateral blindness probably developed in response to pain and stress associated with the diagnostics and treatment of a sterile abscess at the tip of the catheter of an epidural morphine pump [28].

Other concomitant risk factors, which were present in the aforementioned cases, included: acute physical injury [21, 26], chronic health problems [24, 28], coexisting conversion disorders (multiple personality disorder [19, 20], or mutism [17]), migraine headaches [17, 23], family problems [17, 19, 23, 24], unfulfilling or unhappy personal life [19, 25], stress in school or at university [17, 21].

The course of blindness in conversion disorders

Psychogenic blindness can last from a couple of hours up to many years, can be intermittent or continuous, with a presentation unchanged over time or varied. The characteristics of blindness presented by the 17 patients varied. Many of the discussed patients presented with full bilateral vision loss [17, 19-24, 28], but some have also suffered from partial loss of visual field [11, 26, 27] or hemianopsia [18]. The course of blindness can also vary: the symptoms can be stable [17, 21-23, 28], change over time (either deepen [24, 26, 27] or change, for example from binasal hemianopsia to left homonymous hemianopsia [18]) or even be intermittent [19, 20]. The duration of blindness can be very diverse, in the presented cases the symptoms lasted for a couple of hours [19, 21, 28], days [22, 23], weeks [17], months [18, 25, 27], or even years [11, 20, 24, 26].

Differential diagnosis

The differential diagnosis of blindness in the presented cases of patients included vascular, ocular, and neurological causes. Medical conditions considered as potential causes of unexplained blindness have been summarized in Table 1.

Diagnostic methods

Patients presenting with blindness undergo many thorough tests, which aim to uncover an organic, potentially treatable cause of vision loss. Based on the most commonly used tests in the presented cases of patients, the standard diagnostic tests for patients with unexplained blindness should include a detailed

Table 1. Potential causes of unexplained blindness, used to perform a differential diagnosis in the cases of presented patients.

Differential diagnosis of unexplained blindness		
Vascular causes:	Ocular causes:	Neurological causes:
- cerebrovascular accidents [18], such as bilateral occipital infarcts [17], - bilateral internal carotid artery aneurysm [14], - central retinal artery occlusion [17, 21], - ischemic optic neuropathy [18].	- optic neuritis [11], - posterior uveitis [11], - toxic amblyopia [11], - ocular paraneoplastic syndrome (in the case of false-positive anti-retinal antibody detection) [22].	- hydrocephalus [14], - cortical blindness [18], - migraine [11, 17].

ophthalmic and neurological examination, magnetic resonance imaging (MRI) of the brain, visual evoked potentials (VEP), and, if all examinations were negative or inconsistent, a psychiatric or psychological evaluation [11, 18, 20-24, 26-28]. However, some authors performed additional testing to reach a diagnosis: an assessment of antibody levels for autoimmune diseases, an electroretinogram (ERG) [26, 27], a cranial computed tomography (CT) [22, 28], an electroencephalogram (EEG) [24, 28], a single photon emission computed tomography (SPECT) of the brain [23], and more.

Traditionally, in psychogenic illnesses, most test results, especially those that assess the symptoms objectively, are expected to be negative, and a diagnosis of psychogenic blindness could be considered an exclusion diagnosis. However, with the development of diagnostic methods, descriptions of specific findings in conversion disorders are reported in the medical literature. From the reviewed material, Rota et al. described changes in the SPECT examination of the brain – during the period of vision loss, a focal hyperperfusion in the left thalamus and posterior cingulate cortex was revealed. On a follow-up examination performed 2 months after recovery, these changes were not visible. The authors hypothesized that these regions of the brain are responsible for perception, but no conscious awareness of vision in psychogenic blindness [23]. Werring et al. performed functional magnetic resonance imaging (fMRI) on the 5 described patients, who also presented changes in the activity of the brain tissue. Upon examination, patients with psychogenic vision loss had lower levels of activity in the visual cortex compared to a control group, but higher levels of activity in the left inferior frontal cortex, left insula, left claustrum, left limbic system, left posterior cingulate cortex, bilateral striatum, and bilateral thalami. The authors postulate that this unusual activation can suggest suppression of the primary visual cortex or a shift toward unconscious perception [11]. Similar findings associated with altered activity of the brain during vision and blindness were reported by Strasburger et al., who described the case of a 33-year-old female with multiple personality disorder, where some personalities were blind, and some were sighted. Along with the personality changes, changes in the VEP examination were seen – amplitudes and latency values were within normal limits in personalities with vision, and abnormal results were seen in the blind state. These results also indicate that the visual stimulus is modulated within the brain, likely in the lateral geniculate nucleus of the thalamus, and influences the signal transduced from the retina to the visual cortex [20].

Another factor that can be taken under consideration while assessing a patient with unexplained vision loss or a suspected psychogenic blindness, is the overall behavior of the patient. In the reviewed literature, some authors described specific symptoms, which retrospectively were consistent with the diagnosis of blindness in the course of a conversion disorder. Rota et al. described that the patient did not bump into objects despite bilateral blindness [23]. Similarly, Cai et al. reported a patient with severe constriction of the visual field, who ambulated very well as for the extent of vision loss seen on ophthalmic examination. The authors also noted inconsistencies in test results with previous visits and a lack of a physiological visual field expansion at a distance [26]. Chung et al. reported a bizarre change of symptoms in a patient (binasal hemianopsia changing to left homonymous hemianopsia) and inconsistency of the visual field examination with the results of VEP [18]. Khan et al. described a 10-year-old girl, who, despite full bilateral blindness, was able to perform simple tasks using vision subconsciously - picking up objects from the floor, or taking candy from another person [25]. A 21-year-old, supposedly fully blind patient reported by Mulugeta et al., was able to draw objects in correspondence with a narrow field of vision [17]. Parry-Jones et al. described an interesting symptom accompanying psychogenic vision loss – the 47-year-old female patient had longstanding difficulties in reading Braille. This was hypothesized to be caused by the lack of increased tactile acuity, which should accompany the decrease in visual stimuli [24]. Lastly, Shell et al. reported that their patient did not show any concern about the newly developed bilateral blindness, despite the severity of the condition [28].

Treatment

The treatment of psychogenic blindness in the presented cases consisted of psychotherapy and, if needed, pharmacotherapy. General psychotherapy was implemented by many authors [19, 20, 22, 24, 27], with some using specifically cognitive behavioral therapy [18, 21], hypnotherapy [20], family therapy [27], behavioral and social skill therapy [17], or teaching relaxation techniques [17]. Additionally, providing a supportive and understanding environment can help promote recovery and restore vision [17]. As for pharmacotherapy, mostly benzodiazepines taken on an ad hoc basis were prescribed [17, 19, 27], and in two cases fluoxetine [27] or amitriptyline [17].

Conclusions

This systematic review aimed to summarize the reported cases of psychogenic blindness in order to provide insight into the possible triggering and risk factors, course of disease, and diagnostic approaches for blindness in conversion disorders. The findings highlight the need for a multidisciplinary approach

involving ophthalmologists, neurologists, and mental health specialists to accurately diagnose and differentiate psychogenic blindness from organic visual impairments. Further research is required to elucidate the underlying mechanisms and develop effective treatment strategies for individuals with psychogenic blindness. For more details regarding the described cases, we refer the readers to the cited literature, as discussing every patient's full history exceeded the scope of this review.

Ethical considerations

The process of preparing this review did not involve any use of human or animal subjects.

Conflict of interest

The authors have declared no conflict of interest.

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