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# Susac syndrome in pregnant women – literature review

## Zespół Susaca u ciężarnych – przegląd literatury

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## Abstract

Susac syndrome is an extremely rare disease that may present with ophthalmological, audiological, and neurological manifestations, typically affecting young women. Among the approximately 400 cases described in the literature, 5% involve women during pregnancy or the postpartum period. The classic triad of symptoms includes encephalopathy, visual disturbances caused by occlusion of the retinal artery branches, and sensorineural hearing loss, typically bilateral. The diagnostic process involves evaluation of the clinical picture, magnetic resonance imaging, lumbar puncture, and audiological tests. Treatment should be tailored to pregnant individuals; typically, it is based on intravenous steroids, intravenous immunoglobulins, oral steroids, or plasma exchange. It is vital to take into consideration the teratogenicity of drugs and their effect on the foetus. Due to the varied clinical presentations of Susac syndrome, patients are often misdiagnosed or underdiagnosed, and late implementation of treatment may lead to irreversible hearing loss or blindness. The literature shows that most pregnancies affected by Susac syndrome lead to safe delivery of healthy newborns. However, due to the lack of standardised recommendation regarding this condition, further research is needed to establish standards of treatment. The primary limitation for expanding research is the rarity of the disease. The main aim of the study is to summarise recent knowledge about Susac syndrome in pregnant women.

**Keywords:** pregnancy, neurology, Susac syndrome

## Streszczenie

Zespół Susaca jest bardzo rzadką chorobą, która może objawiać się manifestacjami okulistycznymi, audiologicznymi i neurologicznymi. Zazwyczaj chorują na nią młode kobiety. Spośród około 400 przypadków opisanych w literaturze 5% dotyczy kobiet w ciąży lub w okresie poporodowym. Klasyczna triada objawów obejmuje encefalopatię, problemy ze wzrokiem spowodowane zamknięciem gałęzi tętnicy siatkówki oraz czuciowo-nerwową utratę słuchu, zwykle obustronną. W procesie diagnostycznym zespołu Susaca przeprowadza się ocenę obrazu klinicznego pacjenta, rezonans magnetyczny, punkcję lędźwiową z oceną płynu mózgowo-rdzeniowego i badania audiologiczne. Leczenie powinno być dostosowane do ciężarnych pacjentek i zazwyczaj obejmuje dożylne podawanie sterydów, dożylne podawanie immunoglobulin, doustne podanie sterydów lub wymianę osocza. Niezwykle ważne jest uwzględnienie teratogenności leków i ich wpływu na płód i ciężarną. Ze względu na zróżnicowany obraz kliniczny u pacjentów z zespołem Susaca choroba jest często błędnie diagnozowana lub niedodiagnozowana, a późne wdrożenie leczenia może prowadzić do nieodwracalnej utraty słuchu lub ślepoty. Literatura wskazuje, że większość ciąż dotkniętych zespołem Susaca prowadzi do bezpiecznego porodu zdrowych noworodków. Z powodu braku standardowych zaleceń dotyczących diagnostyki i leczenia tego schorzenia konieczne są dalsze badania w celu ustalenia uniwersalnych wytycznych. Głównym ograniczeniem w rozszerzaniu badań jest rzadkość choroby. Główny cel pracy stanowi podsumowanie aktualnej dostępnej wiedzy na temat zespołu Susaca u kobiet w ciąży.

**Słowa kluczowe:** ciąża, neurologia, zespół Susaca

## INTRODUCTION

Susac syndrome (SuS) is a rare autoimmune endotheliopathy of unknown origin, with the majority of patients being young women, aged 20–40 years, which corresponds to the potential childbearing age (Kapica-Topczewska et al., 2017; Pereira et al., 2020). The condition was first described by Susac in 1979 (Al-Hasan et al., 2020; Gross et al., 2019; Pereira et al., 2020; Wilf-Yarkoni et al., 2024). Female patients are affected three times more frequently than males, and the syndrome is most common among the Caucasian population (Al-Hasan et al., 2020; Pereira et al., 2020). To date, approximately 450 cases of SuS have been reported (Cviková et al., 2024). The occurrence of SuS in pregnancy and the postpartum period is estimated to account for 5% of cases (Al-Hasan et al., 2020; Feresiadou et al., 2014). The disease leads to the disruption of microvessels, typically the precapillary segments of arteries, within the brain, inner ear, and retina (Cviková et al., 2024; Pereira et al., 2020). Histopathological examination reveals focal microangiopathies of small to medium vessels in these locations (Gross et al., 2019). In 2007, Susac and colleagues suggested the potential role of anti-endothelial cell antibodies (AECA) as the cause of the syndrome, but it is important to note that these antibodies are not specific to this disease, so screening tests based on their presence are not recommended (Pereira et al., 2020; Susac et al., 2007). Other studies point to the possibility of cytotoxic T-lymphocyte adhesion to the endothelium of microvessels located in the central nervous system (CNS), which was confirmed in animal models (Gross et al., 2019). Literature reports indicate that SuS relapses in pregnancy may be caused by hormonal and immunological changes (Al-Hasan et al., 2020).

The typical presentation includes the classic triad of symptoms, which is as follows:

- encephalopathy;
- visual disturbances caused by occlusion of the retinal artery branches;
- sensorineural hearing loss (Gross et al., 2019).

It is important to note that the full triad is rarely present at the onset of the disease – it has been estimated that approximately 13% of patients initially present with all three symptoms (Cviková et al., 2024; Pereira et al., 2020).

| Subset 1 | Subset 2 |
|---|---|
| Severe neurological symptoms | Ophthalmological symptoms<br>Recurrent episodes of branch retinal artery occlusions<br>Absent or mild neurological symptoms |

*Tab. 1. Common clinical subsets of Susac syndrome*

The presentation during pregnancy may be varied and atypical (Al-Hasan et al., 2020). However, the review conducted in 2024 examined 311 cases and estimated that the full triad was present in 60% of the cases reviewed (Wilf-Yarkoni et al., 2024). Neurological symptoms are the most frequent, with an estimated occurrence of 67–98% (Cviková et al., 2024; David et al., 2022; Pereira et al., 2020). Symptoms from the central nervous system may include cognitive dysfunction, behavioural changes, confusion, delusions, ataxia, aphasia, sensory loss, and headaches (Cviková et al., 2024). Headaches typically start approximately six months before other symptoms (Gomez-Figueroa et al., 2017). Ocular dysfunction, which occurs in approximately 50% of cases, usually involves partial vision loss, typically central, paracentral, or altitudinal (Cviková et al., 2024; David et al., 2022). Acoustic deficits may be unilateral or bilateral and can occur overnight (Cviková et al., 2024; Gomez-Figueroa et al., 2017; Patel et al., 2018). Patients often describe a sensation of pressure or fullness in the affected ear (David et al., 2022). It is believed that an asymmetric hearing deficit, particularly with higher frequency involvement, is associated with a more severe presentation of the disease (Bose et al., 2023). Other symptoms may include nausea, vomiting, movement impairment, urinary system malfunction, myalgia, or arthralgia (Cviková et al., 2024).

Clinically, the disease presentation can be divided into two subsets, which are presented in Tab. 1 (Pereira et al., 2020). Another classification may be based on clinical courses, as suggested by Rennebohm, which are presented in Tab. 2 (Marrodan et al., 2022; Pereira et al., 2020; Wilf-Yarkoni et al., 2024). A self-limiting course is considered the most common among SuS patients (Pereira et al., 2020).

Diagnostic methods for SuS include magnetic resonance imaging (MRI) of the brain, fluorescein angiography (FA) of the retina, and audiometry (Pereira et al., 2020). During pregnancy, MRI without gadolinium is preferred (Willekens and Kleffner, 2021). Also, the use of fluorescein dye is not recommended (Marcos-Figueiredo et al., 2018). Making a definitive diagnosis can be challenging due to the rarity of the full triad of symptoms. Major diseases taken under consideration in the differential diagnosis include multiple sclerosis (MS) and acute disseminated encephalomyelitis (Pereira et al., 2020). The disease primarily affects the white matter of the brain, but other structures, including grey matter, the thalamus, cerebellum, and meninges may be also involved. Pathognomonic signs in the MRI for SuS include "snowball" lesions located in the corpus callosum, caused by multifocal microinfarcts (Pereira et al., 2020).

Currently, there is a lack of standardised guidelines regarding the treatment of pregnant patients (Cviková et al., 2024;

| Monocyclic | Polycyclic | Chronic |
|---|---|---|
| Fluctuating course with self-limitation, lasting up to 2 years | Recurrent course, lasting more than 2 years | Chronic, continuously active disease course |

*Tab. 2. Clinical courses of Susac syndrome*

Pereira et al., 2020). Most management strategies are based on clinical experience and case reports. The treatment of choice is immunosuppressive therapy, preferably aggressive and early implemented (Pereira et al., 2020). Prompt implementation of treatment may lead to favourable outcomes, even in severe cases (Pereira et al., 2020). The main concern regarding this type of treatment during pregnancy is the potential toxicity and harm to the foetus (Antulov et al., 2014; Ioannides et al., 2013). Despite the fact that treatment is considered effective, due to the tendency for recurrence, SuS patients need lifelong observation and monitoring (Pereira et al., 2020).

## MATERIALS AND METHODS

To conduct this review, we searched the PubMed database using the keywords "Susac syndrome pregnant" and "Susac syndrome pregnancy". Eighteen articles were identified, of which nine were included in the review. The main inclusion criterion was that the articles described cases of SuS in pregnant women. Papers were excluded if they were inaccessible, written in languages other than English, or lacked a description of SuS cases. Articles discussing other conditions or SuS in the postpartum period were also excluded.

## Susac syndrome in pregnant women

The literature includes descriptions of SuS cases occurring during pregnancy. Nine cases were included in this review. A brief summary of identified cases is presented in Tab. 3 (Al-Hasan et al., 2020; Antulov et al., 2014; Engeholm et al., 2013; Feresiadou et al., 2014; Gomez-Figueroa et al., 2017; Gordon et al., 1991; Hua et al., 2014; Ioannides et al., 2013; MacFadyen et al., 1987). The cases describe nine pregnant women aged 21 to 35 years, with gestational ages ranging from 13 to 37 weeks. In most cases, SuS followed a monocyclic or probably monocyclic course (six out of nine cases), and in one case the course was chronic continuous. In three women, the symptoms of onset were related to audiological dysfunction, another three presented primarily with neurological changes, while two experienced visual deficits. Of these nine patients, seven delivered healthy babies. In two cases, therapeutic abortion was performed. The main treatment strategy was intravenous steroids administration. Other methods included oral

| Study | Patient's data (age, GA) | Symptoms | Treatment | Pregnancy outcome | Course/symptoms of onset |
|---|---|---|---|---|---|
| Gordon et al., 1991 | 28 years, 28 weeks GA | Weakness, dysarthria, apathy, visual deficit, bilateral sensorineural hearing loss | Heparin with partial response; warfarin with almost total recovery | Delivered healthy baby | Not stated/ocular |
| MacFadyen et al., 1987 | 31 years, GA not mentioned | Ataxia, dysarthria, visual deficit, bilateral sensorineural hearing loss | Oral prednisone with partial remission | Delivered healthy baby | Monocyclic/ocular |
| Ioannides et al., 2013 | 28 years, 13 weeks GA | Weakness, dysarthria, unilateral hearing loss (right-sided), loss of visual acuity | IVMP with no response; PLEX and IVIg with no response | Therapeutic abortion at 15 weeks GA | Chronic continuous/neurological |
| Engelholm et al., 2013 | 32 years, 32 weeks GA | Encephalopathic syndrome, weakness, visual deficit, sensorineural hearing loss | IVMP with partial response, oral prednisone; MTX and MMF was implemented in chronic treatment | Delivered healthy baby | Probably monocyclic/neurological |
| Hua et al., 2014 | 25 years, 14 weeks GA | Amnestic symptoms, gait dysfunction, loss of visual acuity | IVMP with partial response, oral prednisone with partial response | Delivered healthy baby | Monocyclic/audiological |
| Antulov et al., 2014 | 21 years, 35 weeks GA | Weakness, cognitive dysfunction, unilateral hearing loss (left-sided) | IVIg with complete response, AZA | Delivered healthy baby | Probably monocyclic/neurological |
| Feresiadou et al., 2014 | 35 years, 37 weeks GA | Loss of visual acuity, unilateral hearing loss (left-sided) | IVMP with partial response, oral prednisone with almost total recovery | Delivered healthy baby | Monocyclic/audiological |
| Gomez-Figueroa et al., 2017 | 34 years, 15 weeks GA | Behavioural changes, apathy, abulia, disorientation, gait deterioration | IVIg and IVMP pulses without improvement | Therapeutic abortion at 17 weeks GA | Probably monocyclic/audiological |
| Al-Hasan et al., 2020 | 31 years, 36 weeks GA | Vertigo, nausea, visual deficit, unilateral hearing loss (left-sided) | IVMP, oral prednisone, IVIg with an improvement | Delivered healthy baby | Not stated/not stated |

**AZA** – azathioprine; **GA** – gestational age; **IVIg** – intravenous immunoglobulins; **IVMP** – intravenous methylprednisolone; **MMF** – mycophenolate mofetil; **MTX** – methotrexate; **PLEX** – plasma exchange.

*Tab. 3. Summary of Susac syndrome cases in pregnant women*



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steroids, intravenous immunoglobulins, plasma exchange, and anticoagulants.

## DISCUSSION

SuS typically presents with neurological, ocular, and audiological symptoms. However, there are also cases of SuS involving dermatological findings, such as livedo reticularis or racemosa (Srichawla, 2022). Because of its varied clinical picture, the disease is often underdiagnosed or misdiagnosed (Marrodan et al., 2022). Literature indicates that there are cases of disease recurrence connected with pregnancy and the postpartum period (Pereira et al., 2020). It is important to remember that autoimmune diseases, such as SuS, often affect young women, and the possible course of the disease during pregnancy is hard to predict (Ioannides et al., 2013). However, data on the impact of pregnancy on SuS is limited (Ioannides et al., 2013).

Despite the fact that Susac et al. (2007) described AECA as a potential pathogenic factor, further studies revealed that the AECA were present in only 30% of the individuals tested. There is a need for wider research to determine whether AECA are responsible for the development of the syndrome or are they a byproduct of endothelial damage, but – as stated above – it is not recommended to perform routine screening based on these antibodies. Gross and colleagues revealed that blocking CD8+ T-cell-mediated pathways using anti-α4-integrin-intervention led to the mitigation of disease symptoms in preclinical models. They also found that four patients obtained improvement in their clinical picture after implementing therapy with natalizumab, which is a humanised monoclonal antibody targeting α4-integrin (Gross et al., 2019).

MRI imaging can be helpful in establishing a definitive diagnosis of Susac syndrome and can be performed in pregnant women. A characteristic feature of the disease is hyperintensity of the white matter on T2-weighted or FLAIR images (Cviková et al., 2024). As mentioned before, the most typical sign of SuS is the presence of a snowball lesion, but vertical "spokes" and triangular "icicles" can also be found within the corpus callosum (Cviková et al., 2024; David et al., 2022; Pereira et al., 2020). The evolution of these lesions leads to the development of larger defects, usually referred to as "punched-out" abnormalities (Pereira et al., 2020). Other MRI signs that may be associated with this syndrome include enhancement of leptomeningeal, cortical or deep grey matter involvement, and microinfarcts within the internal capsule, which are responsible for signs collective known as the "string of pearls" (Cviková et al., 2024; Pereira et al., 2020). When differentiating SuS from MS, it is important to note that callosal lesions in MS are typically ovoid and located peripherally, in the septal area or adjacent to the walls of the lateral ventricles (Cviková et al., 2024). Also, spinal cord involvement in SuS is rare, in contrast to its frequency in MS (David et al., 2022). In some cases, brain findings are revealed post-mortem or via stereotactic brain biopsy (Bose et al., 2023). MRI is generally considered safe in pregnant women, for both diagnostic and monitoring purposes (Willekens and Kleffner, 2021). However, gadolinium should not be used during pregnancy, as it may increase the risk of neonatal death (Willekens and Kleffner, 2021).

FA examination typically reveals arterial wall hyperfluorescence, which is caused by leakage from damaged microvessels (Pereira et al., 2020). This sign is evidence for the involvement of the retina. Data on the use of fluorescein in pregnant women is restricted. It is vital to note that fluorescein dye crosses the placenta and may be present in breast milk for more than 76 hours following administration (Marcos-Figueiredo et al., 2018). Due to insufficient information regarding its teratogenic effects or other risks, it is not recommended to implement this agent in women during pregnancy or the postpartum period (Marcos-Figueiredo et al., 2018).

Pure-tone audiometry may reveal sensorineural hearing deficits, typically unilateral or bilateral, affecting medium and low frequencies ranging from 500 to 1,000 Hz (Cviková et al., 2024; Patel et al., 2018; Pereira et al., 2020; Sikorska et al., 2023). Tympanometry usually reveals a Type A pattern, which is typical for normal middle ear function (Patel et al., 2018). Initially, about 37% of patients present with hearing loss, and up to 96% develop this impairment over the course of the disease (Cviková et al., 2024). Differential diagnosis of otological conditions should include Ménière's syndrome, Cogan syndrome, and sudden sensorineural hearing loss (Patel et al., 2018). These diagnostic examinations may be performed during pregnancy (Lyu et al., 2020).

Another method used in the diagnostic process is lumbar puncture, which can be performed during pregnancy. Examination of the aspirated fluid may show elevated protein levels and lymphocytic pleocytosis (David et al., 2022; Pereira et al., 2020; Sikorska et al., 2023). A key distinguishing feature between SuS and MS is that in SuS oligoclonal bands are typically absent, while they are present in MS (Pereira et al., 2020). Patients may be also evaluated by vestibular examination or videonystagmography (Cviková et al., 2024).

The choice of the immunosuppressive agent may be a great challenge when managing a pregnant patient, as there is a risk of harm to the foetus (Antulov et al., 2014; Ioannides et al., 2013). Treatment is initially based on the intravenous administration of high doses of corticosteroids, followed by a switch to high-dose oral corticosteroids with gradual tapering (Pereira et al., 2020; Pérez-Lombardo et al., 2019). Another method may be intravitreal administration of corticosteroids, reported to yield good outcomes in patients with ophthalmological involvement (Pereira et al., 2020).

Other agents that can be used in SuS management include intravenous immunoglobulins, rituximab, mycophenolate mofetil (MMF), cyclophosphamide, and azathioprine. Drugs may be combined to provide the best possible effects (Pereira et al., 2020). Adjuvant therapies include

anticoagulants, antiplatelets, and plasma exchange (Bose et al., 2023; Pereira et al., 2020). Patients with ophthalmologic occlusions can be also treated with hyperbaric oxygen (Pereira et al., 2020).

Hearing loss is, in most cases, irreversible (Sikorska et al., 2023). Among in the treatment options for patients with hearing loss are intratympanic steroid injections and cochlear implants (Bose et al., 2023; Patel et al., 2018; Pereira et al., 2020). Peyre et al. (2024) suggested that severe hearing loss may be linked to the lack of immunosuppressive treatment at the moment of SuS diagnosis. The use of cochlear implants should be taken into account individually. It is important to consider the risks associated with anaesthesia in pregnant women. Intratympanic steroids, such as dexamethasone, appear to be safe during pregnancy, as they are administered locally, do not cause systemic adverse effects, and do not cross the placenta (Fu et al., 2019). Fu et al. (2019) revealed that intratympanic steroids do not lead to any abnormalities in newborns and babies, who present normal height and weight parameters.

The length and dosage of administered drugs depend on the severity and duration of the disease. Patients with neurological involvement usually require longer and more aggressive therapy than those with ophthalmological symptoms (Pereira et al., 2020). Sadly, in many patients treatment is initiated late, leading to irreversible damage, such as blindness, hearing loss, or dementia (Kapica-Topczewska et al., 2017; Marrodan et al., 2022). Physicians must tailor the treatment to the pregnant patient, keeping in mind which methods and agents are permissible during pregnancy.

Currently, there are no established guidelines for the treatment of pregnant women with SuS. It is vital to remember that potential toxicity to the foetus must be taken into consideration (Willekens and Kleffner, 2021). In most reported cases, pregnant women were treated with steroids, intravenous immunoglobulins, and plasma exchange, while cyclophosphamide and rituximab were assigned to severe cases and administered in the postpartum period (Willekens and Kleffner, 2021). Both MMF and methotrexate are contraindicated during pregnancy and breastfeeding (Willekens and Kleffner, 2021). MMF can be responsible for teratogenic impact and pregnancy loss (Marrodan et al., 2022). Azathioprine is not considered teratogenic, but it may cause premature delivery and low birth weight, so it is not recommended as a first-line agent in pregnant patients (Willekens and Kleffner, 2021). Also, natalizumab may be considered in pregnant SuS patients (Willekens and Kleffner, 2021). Therapy with cyclosporin A and tacrolimus may be continued during pregnancy, if already in use, but should not be initiated during this period (Willekens and Kleffner, 2021). Acetyl salicylic acid can be implemented for vascular occlusion prevention (Willekens and Kleffner, 2021). To sum up, researchers suggest that first-line treatment should be based on oral steroids (prednisone or methylprednisolone) in low doses and monthly administration of intravenous immunoglobulins (Willekens and Kleffner, 2021). In some patients, therapeutic abortion may be considered – Aubart-Cohen described a case when this procedure was implemented to allow the use of cyclophosphamide (Aubart-Cohen et al., 2007; Ioannides et al., 2013). Also, labour induction may be an option to allow earlier treatment initiation, depending on gestational age (Antulov et al., 2014).

The optimal time to conceive is during a period of no symptoms of the disease activity. It is advised to attempt pregnancy after approximately six months of remission, but it is important to note that a relapse-free pregnancy is not guaranteed (Willekens and Kleffner, 2021). Patients should be in stable condition, either not receiving any treatment or with the management that is suitable for pregnant individuals (Willekens and Kleffner, 2021).

The rarity of the disease means that there is currently a lack of large randomised clinical trials and management guidelines. The small number of described cases of SuS in pregnancy is a primary limitation regarding this topic. Collaboration between neurologists, gynaecologists, and obstetricians is important to ensure the management of Susac syndrome that is safe for both the mother and her foetus.

## CONCLUSION

SuS is a rare disease that may present with ocular, audiological, and neurological manifestations. Approximately 5% of the cases involve women during pregnancy or the postpartum period. The diagnostic process includes evaluating the clinical picture and magnetic resonance imaging. Treatment should be tailored to pregnant individuals and typically involves intravenous steroids, intravenous immunoglobulins, or oral steroids. The literature shows that most pregnancies affected by SuS lead to safe delivery of a healthy newborn. Due to the lack of standardised recommendations regarding this condition, further research is needed to establish standards of treatment. The main limitation is the rarity of the disease.

**Conflict of interest**

*The authors do not report any financial or personal connections with other persons or organisations which might negatively affect the content of this publication and/or claim authorship rights to this publication.*

**Author contribution**

*Original concept of study; collection, recording and/or compilation of data; analysis and interpretation of data; writing of manuscript; critical review of manuscript; final approval of manuscript: KK, RK, AK.*

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