

# Pediatric cerebral venous sinus thrombosis – report of 10 cases and literature review

## Zakrzepica zatok żylnych mózgu u dzieci – opis 10 przypadków i przegląd literatury

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### KEYWORDS

acute otitis media, sinusitis, acute mastoiditis, cerebral venous sinus thrombosis, internal jugular vein thrombosis

### SUMMARY

Cerebral venous sinus thrombosis (CVT) is a life-threatening condition defined as partial or complete occlusion of a sinus or a cerebral vein. In pediatric population CVT is mainly caused as a complication of ongoing otitis or sinusitis. Retrospective review was conducted based on medical records of patients treated for venous sinus thrombosis in Pediatric Otolaryngology Department in Silesian Health Centre for Child in Katowice. Medical records were reviewed from January 2021 to July 2023. The study evaluated such parameters as: age, gender, source of infection, type of complications, radiological data, cultures examination, surgical procedures, pharmacological treatment and duration of hospitalization. The 10 children were included in this study. The most common sinus thrombosis concerned sigmoid sinus – 70% (7/10), 60% was diagnosed with internal jugular vein thrombosis (Lemierre's syndrome), 40% with epidural abscess, 30% had traversed sinus thrombosis, 10% had cavernous sinus thrombosis, 10% upper sagittal thrombosis, 20% had orbital abscess, 10% had subdural abscess. Cerebral venous sinus thrombosis is serious and life-threatening complication of acute otitis media and sinusitis. The management of this condition is always challenging and requires interdisciplinary approach. Early identification and prompt clinical and surgical intervention is essential for better outcome. Broad – spectrum antibioticotherapy and surgical treatment must be performed due to patients best recovery.

### SŁOWA KLUCZOWE

ostre zapalenie ucha środkowego, zapalenie zatok, ostre zapalenie wyrostka sutkowatego, zakrzepica zatok żylnych mózgu, zakrzepica żyły szyjnej wewnętrznej

### STRESZCZENIE

Zakrzepica zatok żylnych mózgu jest stanem zagrażającym życiu, definiowanym jako częściowe lub całkowite zamknięcie zatoki lub żyły mózgowej. W populacji pediatrycznej występuje głównie jako powikłanie zapalenia ucha środkowego lub zatok. Retrospektywny przegląd przeprowadzono na podstawie dokumentacji medycznej pacjentów leczonych z powodu zakrzepicy zatok żylnych na Oddziale Otolaryngologii Dziecięcej Śląskiego Centrum Zdrowia Dziecka w Katowicach. Analizie poddano dokumentację medyczną z okresu od stycznia 2021 do lipca 2023 roku. W badaniu oceniano takie parametry, jak: wiek, płeć, źródło zakażenia, rodzaj powikłań, dane radiologiczne, posiewy, zabiegi chirurgiczne, leczenie farmakologiczne oraz czas hospitalizacji. Do badania włączono 10 dzieci. Najczęściej zakrzepica zatok dotyczyła zatoki esowatej – 70% (7/10), u 60% rozpoznano zakrzepicę żyły szyjnej wewnętrznej (zespół Lemierre'a), u 40% – ropień nadtwardówkowy, u 30% – zakrzepicę zatoki klinowej, u 10% – zakrzepicę zatoki jamistej, u 10% – zakrzepicę zatoki strzałkowej górnej, u 20% – ropień oczodołu, u 10% – ropień podtwardówkowy. Zakrzepica zatok żylnych mózgu jest poważnym i zagrażającym życiu powikłaniem ostrego zapalenia ucha środkowego i ostrego zapalenia zatok. Leczenie tych schorzeń jest zawsze trudne i wymaga interdyscyplinarnego podejścia. Wczesna diagnostyka oraz szybka interwencja chirurgiczna są niezbędne do uzyskania lepszych wyników leczenia.

## INTRODUCTION

Cerebral venous sinus thrombosis (CVT) is a life-threatening condition defined as partial or complete occlusion of a sinus or a cerebral vein. In pediatric population CVT is mainly caused as a complication of ongoing otitis or sinusitis. Anatomy of bony structures in temporal bone or sinuses of children is predisposing factor to spreading the inflammation process from primary location to adjacent structures (1, 2). Venous sinuses of dura mater collect blood from brain, meninges, ears, eyeballs and orbits. The CVT can be caused by infection or other reasons (1). The infectious process can be located in sinuses, ear, meninges or any other organ that is the cause of spreading the bacteria to the blood (such as endocarditis). Non-infectious reasons include hematology disorders, cancer, surgery, multi-organ trauma, dehydration, hormonal problems, brain tumors, head trauma or systemic connective tissue diseases (3). Anatomy of the mastoid and the sigmoid sinus makes possible for otitic inflammation to activate platelets and fibrin resulting in an intrasinus thrombosis. Thrombus of one vein or one venous sinus can extend to adjacent veins or sinuses. The most common complaint is headache present in 88.8%. Other symptoms are seizures (39.3%), paresis (37.2%), focal neurologic deficits and altered mental status (4). Mainly thrombosis is diagnosed in the transverse sinuses (44-73%), the superior sagittal sinus (39-62%), sigmoid sinus (40-47%), deep venous system (10.9%) and cortical veins (3.7-17.1%) (5). About 15% of patients with CVT not survive or become disabled (6, 7).

Retrospective review was conducted based on medical records of patients treated for venous sinus thrombosis in Pediatric Otolaryngology Department in Silesian Health Centre for Child in Katowice. Medical records were reviewed from January 2021 to July 2023. The study evaluated such parameters as: age, gender, source of infection, type of complications, radiological data, cultures examination, surgical procedures, pharmacological treatment and duration of hospitalization. The 10 children were included in this study. 8 of them were diagnosed with otogenic complications and 2 with sinogenic complications. All otogenic intracranial complications were caused by acute otitis media. Group consisted of 7 boys and 3 girls. Average age was 6.1 (2 to 15-year-old). In 3 cases cultures examinations were negative due to ongoing antibiotic therapy. *Staphylococcus epidermidis* was positive in 3 patients and *Streptococcus pneumoniae* was the cause in 2 cases. All the patients had CT and MRI.

## RESULTS

The most common sinus thrombosis concerned sigmoid sinus – 70% (7/10), 60% was diagnosed with internal jugular vein thrombosis (Lemierre's syndrome), 40% with epidural abscess, 30% had transverse sinus thrombosis, 10% had cavernous sinus thrombosis, 10% upper sagittal thrombosis, 20% had orbital abscess, 10% had subdural

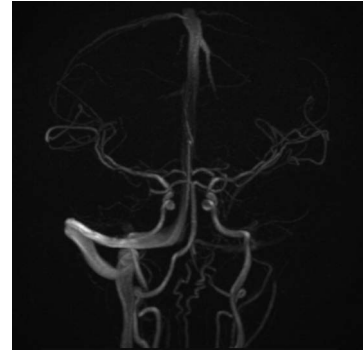


Fig. 1. Angio-MRI – thrombosis of left transverse and left sigmoid sinus

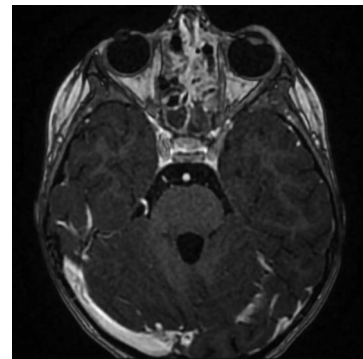


Fig. 2. MRI of the head – thrombosis of left transverse sinus

abscess (fig. 1-2). All patients were treated with surgery. Children with sinus complications (20%) underwent functional endoscopic sinus surgery. All complications of acute otitis (80%) required mastoidectomy with skeletonization of the cortical bone of the thrombotic sinus and myringotomy with or without placement of a ventilation tube (5/8). In two patients drainage of epidural abscess was performed. Antibiotic therapy was provided according to culture examination. 60% of all patients were treated with low molecular weight heparin (LMWH). Intravenous steroids were administered in 3/10 patients. All patients survived and were discharged home. Mean hospitalization was 19.6 days (11-42 days) (tab. 1).

## DISCUSSION

Upper respiratory infections and acute otitis media (AOM) are the most common infectious diseases in the pediatric population. Despite numerous medical advances, better diagnostics and common availability of antibiotic therapy the complications are still present and carry a risk of mortality. Risk factors for otogenic or sinogenic complications include hematologic disorders, cancer, multi-organ trauma, dehydration, hormonal disorders, brain tumors, head trauma, diabetes, immunodeficiency, nephrotic syndrome, cardiac disease or systemic autoimmune disorders (systemic lupus erythematosus, Behçet disease, Sjögren's syndrome,

**Tab. 1.** Patients included in the study (sex, age, origin of the infection, complications, culture examination, pharmacological treatment, surgical approach)

Pa-tient	Sex	Age	Origin of infection	Complications	Culture examination	Farmacological treatment	Surgical approach
1	M	15	Sinusitis	– cavernous sinus thrombosis	Negative	ceftriaxon, metro-nidazol, nadro-parinum	FESS
2	F	4	Left otitis	– mastoiditis – internal jugular vein thrombosis – inflammation of knee and hip joint – pneumoniae – sepsis	<i>Staphylococcus epidermidis</i>	linezolid, nadroparinum, vancomycin	mastoidecto-my, medial ear drainage
3	M	3	Left otitis	– mastoiditis – epidural abscess – transverse sinus thrombosis – sigmoid sinus thrombosis – internal jugular vein thrombosis – orbital abscess – abscess zygomatical	Negative	nadroparinum, vancomycin, ceftriaxon	mastoidectomy, drainage of intracranial epi-dural abscess
4	M	8	Left otitis	– epidural abscess – sigmoid sinus thrombosis – internal jugular vein thrombosis – soft tissue abscess of the face – orbital abscess	<i>Streptococcus intermedius</i>	vancomycin, nadro-parinum	mastoidecto-my, drainage of intracranial epidural abscess and orbital abscess
5	M	2	Right otitis	– mastoiditis – sigmoid sinus thrombosis – internal jugular vein thrombosis	<i>Staphylococcus epidermidis</i>	vancomycin ceftria-xon metronidazol	mastoidecto-my, medial ear drainage
6	M	15	Left otitis	– mastoiditis – left sigmoid sinus thrombosis – internal jugular vein thrombosis	<i>Streptococcus pneumoniae</i>	ceftriaxon, amika-cinum, vancomycin dexamethasone	mastoidecto-my, medial ear drainage
7	F	2	Left otitis	– mastoiditis – left sigmoid sinus thrombosis	<i>Enterococcus faecalis</i> <i>Staphylococcus aureus</i>	vancomycin ceftri-axon	mastoidec-tomy, medial ear dreinage
8	M	2	Left otitis	– mastoiditis with perinatal abscess – left sigmoid sinus thrombosis	<i>Streptococcus pneumoniae</i>	ceftriaxon dexamet-hasone	mastoidectomy
9	F	7	Sinusitis	– frontal osteitis – epidural abscess – subdural abscess – encephalitis – bilateral-transverse sinus thrombosis – upper sagittal sinus thrombosis	<i>Staphylococ-cus aureus</i> , <i>Streptococcus constellatus</i> , <i>Staphylococcus epidermidis</i>	meropenemum, van-comycin, ceftriaxon, dexamethasone, nadroparinum	FESS
10	F	3	Left otitis	– internal jugular vein thrombosis – epidural abscess – sigmoid sinus thrombosis – transverse sinus thrombosis	Negative	meropenemum, van-comycin ceftriaxon, nadroparinum	mastoidectomy, medial ear dra-inage

Wegener’s granulomatosis, sarcoidosis, and inflammatory bowel disease) (1, 3, 8, 9). Sometimes the otorhinological diagnostic process is delayed because neurologic, rather than otologic or sinogenic symptoms may dominate the presentation of the cerebral sinus thrombosis (10). Complications of sinusitis include frontal bone osteomyelitis, meningitis, subperiosteal abscess, epidural, subdural or intracerebral empyema, cerebral venous thrombosis and

orbital cellulitis and abscess (11-13). Thrombotic complications of sinusitis can concern all the dural venous sinuses but cavernous sinus is most often affected. Especially if the primary infection is located in ethmoid or sphenoid sinus. It is caused by direct linkage between paranasal sinuses and cavernous sinuses by ophthalmic veins. Cavernous sinus thrombosis presents painful ophthalmoplegia, chemosis, proptosis, retroorbital pressure, cranial nerve palsy, ocular

motility deficit and vision disorder (14-16). Complications of AOM are acute mastoiditis, facial nerve dysfunction, meningitis, subperiosteal abscess, epidural, subdural or intracerebral empyema and cerebral venous thrombosis (2, 17-19). Disseminated infection of acute sinusitis or otitis can also affect structures of orbit and soft tissue of face. Two children of this study had orbital abscess and zygomatic soft tissue abscesses due to otogenic extending of inflammation process. AOM can also lead to sepsis and its further consequences. In our study one 4-year-old child had acute otitis with spreading the bacteria to the blood causing sepsis. This resulted in pneumonia, urinary tract infection and inflammation of right hip and knee joint. Similar cases of otogenic septic arthritis are described in literature (19). Persistent fever, headache, otalgia and purulent otorrhea are usually the main symptoms of acute otitis. Sometimes the fever is not present (20). Nausea and vomiting heralds developing complications (10, 12). The clinical features of childhood CVT are dominated by seizures, symptoms of raised intracranial pressure (ICP) and abnormal consciousness (21). Chan et al. study showed visual disturbances such as diplopia and visual field deficits present in 18% children with CVT (8). Nearly half (46/105) of patients with CVT examined by Bevan et al. was reported with raised intracranial pressure. 8.7% (4/46) of these patients with raised ICP went on to have a ventriculo-peritoneal shunts. Indication for implanting ventriculo-peritoneal shunt was based on deteriorating vision in patients not responding to serial lumbar punctures (22). CVT in children can be also asymptomatic (1). Anatomical proximity of the mastoid and the sigmoid sinus makes possible for otitic inflammation to activate platelets and fibrin resulting in a intrasinus thrombosis. The pathophysiology of CVT is also based on raised cerebral venous pressure. Sinus occlusion due to thrombotic clot leads to backflow of blood into venules and capillaries, resulting in increased local intravessel pressure with the risk of massive cerebral edema. This mechanism leads to decreased cerebrospinal fluid absorption and ultimately causes secondary increased intracranial pressure (23-25). In deteriorating patients with suspicion of developing complications the neuroimaging of head must be performed – computer tomography (CT) or magnetic resonance imaging (MRI) (26-30). Once otogenic CVT is diagnosed, empiric broad – spectrum antibiotic therapy should be initiated. If a specific pathogen is later identified, more specific antimicrobial drug according to culture examination should replace the initial treatment. Castellazzi et al. recommend treatment with meropenem and vancomycin for a total of 6 weeks (2). Systemic steroids should be administered in case of neurological disorders, intracranial oedema and vision disorder (10). Functional endoscopic sinus surgery should be performed for sinogenic complications. As for otogenic CVS urgent mastoidectomy with skeletonization of the

cortical bone of thrombotic sinus and myringotomy with or without placement of a ventilation tube is necessary (2, 10). Low molecular weight heparin (LMWH) is recommended (2, 10, 31-34). The pediatric patients in the study by Castellazzi et al. underwent prolonged anticoagulation therapy for 6 months after discharged from hospital (2). Connor et al. assessed the use of oral anticoagulants in 117 children with cerebral venous thrombosis. The comparison of 2 group divided into therapy of rivaroxaban or warfarin demonstrated similar effect on clot resolution (35). Although majority of patients with otogenic or sinogenic CVT have a complete recovery there is still a 5.6-9% risk of death (17, 36). Pediatric mortality rate is lower comparing to mortality in adults with CVT. Up to 10% are found to have permanent neurological deficits – the most commonly remains changes in vision, seizures and hemiparesis (36, 37). In our study all children had full recovery. There are multiple studies describing higher death rates in patients with CVT and COVID-19 compared with those without COVID-19 (38-40). The mechanism that explains post-Covid-19 state of hypercoagulation is the coronavirus affinity to the angiotensin converting enzyme-2 receptor expressed in vascular endothelial cells. Linkage of the virus and the receptor leads to direct endothelial damage. All components of the Virchow's triad (endothelial dysfunction, altered flow dynamic and hypercoagulable state) may be present and responsible for higher risk of thromboembolic complication (41). Other studies report new finding of intravenous thrombosis after vaccination against SARS-CoV-2. The new syndrome has been termed "vaccine-induced immune thrombotic thrombocytopenia (VITT)". It is observed after administration of the adenovirus vector-based vaccines vaccine (AstraZeneca/Oxford) and (Janssen/Johnson & Johnson). The VITT syndrome presents with thrombosis at unusual sites, such as for example CVT associated with thrombocytopenia and high levels of D-Dimer (42-45). Our study is based on patients with CVS admitted to our Department in 2021-2023 years. Comparing to previous years (2018-2020) numbers of children with otogenic or sinogenic CVT in pandemic years (2021-2023) is five times higher. Due to often asymptomatic demonstration of COVID-19 in pediatric population and lack of data of suffered in the past from COVID-19 and COVID-19 vaccination history we can't refer and compare our study to after COVID-19 hypercoagulation phenomena.

## CONCLUSIONS

Cerebral venous sinus thrombosis is serious and life-threatening complication of acute otitis media and sinusitis. The management of this condition is always challenging and requires interdisciplinary approach. Early identification and prompt clinical and surgical intervention is essential for better outcome. Broad – spectrum antibiotic therapy and surgical treatment must be performed due to patients best recovery.

**CONFLICT OF INTEREST**  
**KONFLIKT INTERESÓW**

None  
Brak konfliktu interesów

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