



Communicable Disease Case Reporting and Investigation Protocol
PRIMARY AMEBIC MENINGOENCEPHALITIS (PAM)
(Caused by *Naegleria fowleri*)

I. IDENTIFICATION AND DEFINITION OF CASES

A. Clinical Description: *Naegleria fowleri* (*N. fowleri*) is a free-living ameboflagellate that invades the brain and meninges via the nasal mucosa and olfactory nerve to cause acute, fulminant hemorrhagic meningoencephalitis (primary amebic meningoencephalitis – PAM), primarily in healthy children and young adults with a recent history of exposure to warm fresh water. Rarely, infections have been reported when people flush their sinuses with tap water contaminated with *N. fowleri*. Initial signs and symptoms of PAM begin 1-14 days after infection and include sudden onset of headache, fever, nausea, vomiting, and stiff neck accompanied by positive Kernig’s and Brudzinski’s signs. In some cases, abnormalities in taste or smell, nasal obstruction and nasal discharge might be seen. Other symptoms might include photophobia, mental-state abnormalities, lethargy, dizziness, loss of balance, other visual disturbances, hallucinations, delirium, seizures, and coma. After the onset of symptoms, the disease progresses rapidly and usually results in death within 3-7 days.

PAM can be treated with [miltefosine](#), which is now commercially available and can be shipped overnight or sooner. Although treatment may be successful if initiated early in the course of disease, most infections have been fatal. Fortunately, infections are very rare. There is no person-to-person communicability of *N. fowleri* infections.

Clinicians and laboratorians: For 24/7 diagnostic assistance, specimen collection guidance, shipping instructions, and treatment recommendations, please contact the Centers for Disease Control and Prevention’s (CDC) Emergency Operations Center at 770-488-7100. More detailed guidance can be found in CDC’s [Information for Public Health & Medical Professionals](#).

B. Laboratory Criteria:

- **Confirmatory laboratory evidence:** Detection of *N. fowleri* antigen or nucleic acid in a clinical specimen (for example, immunohistochemistry or polymerase chain reaction [PCR]).
- **Supportive laboratory evidence:**
 - Visualization of motile amebae in a wet mount of cerebrospinal fluid (CSF).
 - Isolation of *N. fowleri* in culture from a clinical specimen.

Notes: *N. fowleri* can cause clinically similar illness to bacterial meningitis, particularly in its early stages. Unlike other free-living amebae species, *N. fowleri* is commonly found in the CSF. Microscopic examination of a CSF wet mount may detect motile trophozoites, and a Giemsa-stained smear will show trophozoites with typical morphology. Definitive diagnosis by a reference laboratory may be required. Photos to aid laboratory diagnosis of *N. fowleri* infection can be found on the [CDC website](#). If suspected free-living ameba trophozoites are visualized in a clinical specimen, tediagnosis can be arranged at CDC by emailing photos through [DPDx](#), CDC’s Division of Parasitic Diseases and Malaria tediagnosis tool. Instructions for submitting photos through DPDx are available at the [DPDx website](#).

Positive clinical specimens should be forwarded to CDC for laboratory confirmation and ameba identification via the Wisconsin State Laboratory of Hygiene (WSLH). Contact the Bureau of Communicable Diseases (BCD), Communicable Diseases Epidemiology Section (CDES), to obtain approval and instructions prior to submitting specimens to WSLH (see “Contacts for Consultation” on page 4).

C. Wisconsin Surveillance Case Definition:

- **Probable:** a clinically compatible illness that meets the supportive laboratory criteria for infection.
- **Confirmed:** a clinically compatible illness that meets the confirmatory laboratory criteria for infection.

II. REPORTING

- A. **Wisconsin Notifiable Disease Category I – Methods for Reporting:** This disease shall be reported **IMMEDIATELY, BY TELEPHONE**, to the patient’s local health officer or to the local health officer’s designee upon identification of a case or suspected case, per Wis. Admin. Code § [DHS 145.04 \(3\) \(a\)](#). In addition to the immediate report, complete and fax, mail, or electronically report an Acute and Communicable Disease Case Report (DHS [F-44151](#)) to the address on the form, or enter the data into the Wisconsin Electronic Disease Surveillance System ([WEDSS](#)) as an incident of Primary Amebic Meningoencephalitis (PAM) / *Naegleria fowleri* Infection, within 24 hours.

In addition, the local health officer should **immediately** notify CDES. If PAM is suspected, urgent confirmatory testing and treatment with miltefosine should be initiated immediately. Clinicians should be directed to contact the CDC 24/7 Emergency Operations Center at 770-488-7100 for diagnostic assistance and treatment recommendations.

- B. **Responsibility for Reporting:** According to Wis. Admin. Code § [DHS 145.04\(1\)](#), persons licensed under Wis. Stat. ch. [441](#) or [448](#), laboratories, health care facilities, teachers, principals, or nurses serving a school or day care center, and any person who knows or suspects that a person has a communicable disease identified in [Appendix A](#).
- C. **Clinical Criteria for Reporting:** An infection presenting as acute meningoencephalitis or encephalitis with confirmatory or supportive laboratory evidence of *N. fowleri* infection. The clinical presentation of PAM is like that of acute meningitis caused by other pathogens and symptoms include headache, nausea, vomiting, anorexia, fever, lethargy, and stiff neck. Disorientation, mental status changes, seizure activity, loss of consciousness, and ataxia may occur within hours of initial presentation.

Patients presenting with clinically compatible illness and found to have a history of recreational freshwater exposure in the two weeks prior to presentation or known to have performed nasal irrigation (for example, use of a neti pot for treatment of sinus conditions, or practice of ritual ablution including nasal rinsing), in the absence of another explanation for their condition, should raise the index of suspicion for PAM.

- D. **Laboratory Criteria for Reporting:** Laboratory evidence of infection by detection of *N. fowleri* nucleic acid (for example, polymerase chain reaction) or *N. fowleri* antigen (for example, direct fluorescent antibody) in CSF, biopsy, or tissue specimens, visualization of motile amoebae in a wet mount of CSF, or isolation of *N. fowleri* in culture from a clinical specimen. When available, molecular characterization should be reported (for example, genotype).

III. CASE INVESTIGATION

- A. **Responsibility for case investigation:** It is the responsibility of the local health department (LHD) to investigate or arrange for investigation of suspected or confirmed cases as soon as is reasonably possible. A case investigation may include information collected by phone, in person, in writing, or through review of medical records or communicable disease report forms, as necessary and appropriate.

Begin the investigation as soon as possible after receiving the report. Contact and work with CDES staff on the investigation. Investigation responsibilities are outlined below in section III C.

B. Required Documentation:

1. Complete the WEDSS disease incident investigation report (Primary Amebic Meningoencephalitis (PAM) / *Naegleria fowleri* Infection), including appropriate, disease-specific tabs.
2. Obtain copies of **all** laboratory test results and electronically attach or upload them to the disease incident filing cabinet.
3. Complete CDC Free-Living Ameba Infection Case Report Form (will be uploaded to WEDSS disease incident filing cabinet by CDES). Request patient medical records, laboratory reports, and autopsy reports as needed to complete the case report. Upload the completed form to the disease incident filing cabinet.
4. Upon completion of investigation, set WEDSS disease incident process status to “Sent to State” and notify CDES.

C. Additional Investigation Responsibilities:

1. Enter all available initial case report information into WEDSS.
2. Review the case definition, background on the disease, and any laboratory testing results.
3. Contact the patient's health care provider to confirm the PAM diagnosis. Determine if patient exhibited illness signs or symptoms consistent with meningitis or encephalitis, obtain current patient status, onset date of illness, and determine if patient is immunocompromised. Ask what information has been shared with the patient and family.
4. Assess patient risk factors by interviewing the patient or a proxy (that is, parent, guardian, or next of kin) if patient is deceased or too ill to be interviewed. Complete the interview using the appropriate sections of the Free-Living Ameba Infection Interview Worksheet and enter information into WEDSS.
 - Ascertain all recreational swimming and home water exposures in the two weeks before symptom onset and record details of locations and dates.
 - Inquire about use of neti pots for sinus rinsing, water sources used, and how practice was conducted.
 - Identify other possibly exposed contacts and/or family members who may be at risk.
 - Provide education on pathogen-specific prevention and address case-patient family's questions or concerns.
5. Follow up on special situations, including environmental assessments of suspect bodies of water or other agent-specific exposures elicited during interviews. Consult CDES.
6. Coordinate submission of the positive patient specimen to CDC for confirmation and ameba identification. Contact CDES to obtain prior approval and submission instructions prior to submission. A completed CDC form 50-34 (specimen submission form) citing WSLH as the state public health laboratory must accompany the specimen to CDC. Definitive laboratory confirmation and diagnosis by the CDC is required.

IV. PUBLIC HEALTH INTERVENTIONS AND PREVENTION MEASURES

- A. In accordance with Wis. Admin. Code § [DHS 145.05](#), local public health agencies should follow the methods of control recommended in the current editions of *Control of Communicable Diseases Manual*, edited by David L. Heymann, published by the American Public Health Association, and the American Academy of Pediatrics' *Red Book: Report of the Committee on Infectious Diseases*, unless otherwise specified by the state epidemiologist.
- B. In the event of a locally acquired case, it may be appropriate to conduct an assessment of the environment where the case was possibly exposed, and notify the public to prevent additional exposures. Consult CDES for guidance on public messaging and how to conduct the assessment and collect samples.
- C. Provide education on how to prevent *N. fowleri* infections from recreational fresh water:
 - Avoid water-related activities in bodies of warm freshwater, hot springs, and thermally polluted water such as water around power plants.
 - Avoid water-related activities in warm freshwater during periods of high water temperature and low water levels.
 - Hold the nose shut or use nose clips when taking part in water-related activities in bodies of warm freshwater such as lakes, rivers, or hot springs, to prevent contaminated water from going up the nose.
 - Avoid digging in or stirring up the sediment while taking part in water-related activities in shallow, warm freshwater areas.
 - You cannot get *N. fowleri* infection from drinking contaminated water. You can only get it when the water goes up the nose.
- D. Provide education on how to prevent *N. fowleri* infections during sinus rinsing. Very rarely, *N. fowleri* infections have been reported when people use contaminated tap water for nasal rinsing during showering, irrigating, or cleansing their sinuses (nose) for health or religious reasons (neti pot use). *N. fowleri* can grow in public and private water tanks, tanks, and pipes, especially where little or no disinfectant (like chlorine or chloramine) is present.

When preparing a saline or other type of solution for irrigating, flushing, or rinsing your sinuses with a neti pot, sinus rinse bottle, or other irrigation device:

- Use labeled distilled or sterile water purchased from a store.

- If distilled or sterile water is not available, use water that has been previously boiled for 1 minute and left to cool. At elevations above 6,500 feet, boil for 3 minutes.

E. Educate other potentially exposed persons. It is unusual to have multiple cases with the same exposure. However, other persons potentially exposed to the same source as the case should be educated about symptoms of PAM and told to seek immediate medical attention if they develop characteristic symptoms.

V. CONTACTS FOR CONSULTATION

A. Local health departments and tribal health agencies: <https://www.dhs.wisconsin.gov/lh-depts/index.htm>

B. CDC Emergency Operations Center: 770-488-7100

C. BCD, Communicable Diseases Epidemiology Section: 608-267-9003

D. Wisconsin State Laboratory of Hygiene: 1-800-862-1013; after hours emergency number: 608-263-3280

VI. RELATED REFERENCES

Heymann D.L., ed. Infections with Free-Living Ameba. In: *Control of Communicable Diseases Manual*. 20th ed. Washington, DC: American Public Health Association, 2015: 6-9.

Pickering L.K., ed. Amebic Meningoencephalitis and Keratitis. In: *Red Book: 2015 Report of the Committee on Infectious Diseases*. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics, 2015: 231-234.

Cope J.R., Conrad D.A., Cohen N., *et al.* Use of the Novel Therapeutic Agent Miltefosine for the Treatment of Primary Amebic Meningoencephalitis: Report of 1 Fatal and 1 Surviving Case. *Clin Infect Dis*. 2016; 62(6):774-776.

CDC PAM Frequently Asked Questions: <https://www.cdc.gov/parasites/naegleria/general.html>

CDC PAM Information for Public Health & Medical Professionals:
https://www.cdc.gov/parasites/naegleria/health_professionals.html

CDC PAM Diagnostic Tests: <https://www.cdc.gov/parasites/naegleria/diagnosis-hcp.html>

CDC DPDx Telediagnosis: <https://www.cdc.gov/dpdx/contact.html>

DPH *Naegleria fowleri* Primary Amebic Meningoencephalitis Fact Sheet:
<https://www.dhs.wisconsin.gov/publications/p01085.pdf>

CDC PAM Sinus Rinsing for Health or Religious Practice – best practices:
<https://www.cdc.gov/parasites/naegleria/sinus-rinsing.html>