Intestinal Microsporidiosis

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KEYWORDS

• Microsporidium • Enteritis • AIDS • HIV • Immunosuppression • Warthin-Starry • Diarrhea

KEY POINTS

- Microsporidia are ubiquitous obligate intracellular parasites most closely related to fungi that are found in a wide range of hosts and have very long environmental survival times.
- Humans become symptomatically infected with 1 of the 14 species causing human disease in the setting of a depressed or failing immune system.
- The diagnosis of microsporidiosis should be attempted by stool examination with special stains and/or tissue biopsy with confirmation by electron microscopy or molecular methods when available.

EPIDEMIOLOGY AND CLINICAL SPECTRUM OF MICROSPORIDIOSIS

Microsporidia are widespread obligate intracellular parasites. By molecular phylogenetic analysis, they are currently classified as fungi with which they share a common ancestor, but they behave similarly to intestinal protozoa. There are innumerable species of Microsporidium, with only an estimated 2% having proper speciation (~1500). Microsporidia infect a broad range of invertebrates, including fish, mosquitos, and flatworms, as well as vertebrates, and rarely infect humans unless they have an immunocompromised state due to human immunodeficiency virus (HIV) infection, solid organ transplantation,[1,2] chemotherapy, or immune suppression related to chronic autoimmune diseases. There are 14 species infecting humans from 8 genera, with 2 representing the vast majority of infections (**Table 1**). Although the self-limited and almost always asymptomatic infection of immunocompetent hosts is thought to be common, microsporidial diarrheal episodes in patients with AIDS can range from 30% to 70% depending on the series.[3–7] The organisms are most commonly associated with either contaminated unfiltered drinking water or swimming

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Table 1
Microsporidium species, diseases, and some reported reservoirs

Species	Diseases	Environmental Reservoirs
Anncaliia algerae, Anncaliia connori, Anncaliia vesicularum	Keratitis, myositis, systemic infection	Unknown
Enterocytozoon bieneusi	Diarrhea, wasting syndrome, cholangiopathy, cholangitis, acalculous cholecystitis, sinusitis, bronchitis, pneumonitis	Pigs, cattle, chickens, cats, monkeys, dogs
Encephalitozoon cuniculi	Systemic infection, keratoconjunctivitis, sinusitis, pneumonitis, urinary tract infection, nephritis, hepatitis, peritonitis, seizures, encephalitis	Carnivores, ruminants, primates, rabbits, rodents
Encephalitozoon hellem	Systemic infection, keratoconjunctivitis, sinusitis, bronchitis, pneumonia, nephritis, ureteritis, cystitis, prostatitis, urethritis	Ostrich, chickens, hummingbirds
Encephalitozoon intestinalis	Diarrhea, cholangiopathy, cholangitis, acalculous cholecystitis, sinusitis, bronchitis, pneumonitis, urinary tract infection, nephritis, bone lesions; nodular cutaneous lesions	Donkeys, cows, dogs, goats
Microsporidium spp	Corneal ulcer	Unknown
Nosema ocularum	Keratitis	Unknown
Pleistophora spp	Myositis	Fish, crustaceans
Trachipleistophora hominis, Trachipleistophora anthropothera	Systemic infection, including brain, heart, kidney, eye, myositis, keratoconjunctivitis, sinusitis	Unknown
Vittaforma cornea	Keratitis, urinary tract infection	Unknown

E bieneusi and *E intestinalis* account for the vast majority of human infections.
Data from Refs.[7,26,27]

in lakes, ponds, and rivers in proximity to animal reservoirs.[8] Epidemic outbreaks, such as those seen with *Cryptsporidium*, are rare with none reported in the United States and 1 reported in France with no causal source.[9] The spores of Microsporidium can persist in the environment for 1 to 12 months (or more) depending on the species.

Opportunistic infections by the species *Enterocytozoon bieneusi* and *Encephalitozoon intestinalis* have been reported in patients with heart-lung,[10] renal, and bone marrow transplantation; HIV-positive patients with CD4 counts less than 100 per microliter[11]; and occasionally self-limited transient "traveler's diarrhea" in immune-competent patients.[12] Both species present clinically most commonly with chronic watery diarrhea and malabsorption.[3] Less common presentations of the infection include cholangitis and acalculous cholecystitis with organisms in the biliary and pancreatic tracts, as well as nonspecific sinusitis and rhinitis with organisms in the tracheal, bronchial and nasal epithelium (on biopsy or in washings).[11] In addition, *E intestinalis* can be seen in colonic enterocytes[11,13] and as a disseminated infection