



OTITIS MEDIA

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Otitis media (OM) is the medical term for an infection of the middle ear. OM is a common problem for healthy children, and it is one of the more stubborn problems for children with MPS or ML. Children with MPS or ML tend to have many ear infections which can be very difficult to treat. This is complicated by the fact that ear infections occur most frequently in the toddler and preschool years, many times before a diagnosis of MPS or ML or a related disease has been made.

There are two types of OM, acute OM and OM with effusion (OME). Acute OM occurs when fluid is present in the middle ear accompanied by signs or symptoms of ear infection such as bulging eardrum (tympanic membrane) often with pain, or perforated eardrum, often with drainage of purulent material (pus). Symptoms can also include ear tugging, fever, irritability, diminished appetite, vomiting, and diarrhea. Complications, although rare, can include tympanic membrane perforation, acute mastoiditis (inflammation of the mastoid bone), cholesteatoma (mass of cells and cholesterol in the middle ear), meningitis and epidural abscess. Language development can also be affected by repeated ear infections. In normal children, the underlying causes of ear infections appear to be eustachian tube dysfunction resulting from either obstruction by large adenoidal tissue or from abnormalities that impair the normal drainage of fluid from the middle ear. In children with MPS or ML, this is also complicated by the accumulation of glycosaminoglycans in the middle ear and upper respiratory structures.

OME is diagnosed when there is fluid in the middle ear without signs or symptoms of middle ear infection. The child may not have any symptoms but hearing can be affected. Any child who has fluid in the middle ears for a total of three months should undergo a hearing evaluation. OME may be present in a child with MPS or ML for years.

A careful examination of the ear may be difficult for a child with MPS or ML but is essential for proper diagnosis. Ear, nose and throat (ENT) specialists, also called otolaryngologists, can be instrumental in uncovering the diagnosis of MPS or ML in children due to recurrent infections and abnormalities seen under examination. Once a diagnosis of MPS or ML has been made, the ENT specialist can be very helpful with many of the issues regarding the ears, nose and throat.

Persistent ear infections may necessitate culturing the fluid in the middle ear to identify the offending organism. It might be viral, bacterial, or occasionally fungal in nature. Proper identification of the organism enables the physician to prescribe the appropriate medication. Viral infections frequently clear up in 24 to 72 hours on their own. If the organism is fungal in nature, frequent antibiotic use will only worsen the situation. Children can receive a vaccine for streptococcus pneumoniae, which is one of the more

common bacteria causing problems. Some studies have shown that this can possibly help reduce the number of future infections. Occasionally older children may have infections caused by *Pseudomonas aeruginosa*, or *Staphylococcus aureus* which can be more difficult to treat. If the child has tympanostomy tubes as described in the following paragraph, topical agents may be used to treat the infection. Corticosteroids may also be administered and can be helpful. There is a wide array of antibiotics available for treatment; however, some require refrigeration or frequent dosing. Antibiotic shots can be considered for a child who is difficult to medicate.

In most cases of repeated ear infections, insertion of tubes into the eardrum is recommended to allow the fluid to drain. Tympanostomy (ear drum) tubes insertion is a 10 to 15-minute procedure performed under general anesthesia. This surgery is called myringotomy, and the tubes may be referred to as myringotomy tubes. This procedure should ALWAYS be done at a properly equipped hospital and only after consultation with the anesthesiologist due to anesthesia concerns in MPS OR ML children. After the procedure the drained fluid should be cultured to identify the offending organism. The tubes assist the child by maintaining ventilation to the middle ear by artificial means. There are several different types of ventilation tubes. The more permanent ones, such as T-tubes, are generally considered the best choice for MPS OR ML individuals. Removal of the adenoids and tonsils might also be recommended for MPS OR ML children with recurrent acute OM. If the child is to undergo general anesthesia for the placement of ear tubes, adenoidectomy and tonsillectomy should also be considered. This avoids some of the anesthesia risks of having more frequent procedures requiring anesthesia.

Ear infections seem to be a persistent problem in children with MPS OR ML and related diseases, and anything that can help alleviate the symptoms may be warranted. Each child may respond differently to different treatment options, so every option should be tried if needed.