Objective: To describe a rare inner ear malformation that required multiple surgical interventions in order to lower the risk of repetitive ear infections, CSF leakage, and meningitis.

Case report: A 17 month-old girl was admitted with high fever, eyes deviation, tonic-clonic seizures and nuchal rigidity, left otitis media with a nasal clear watery discharge. Patient’s past history was remarkable for a left ear profound hearing loss. Brain imaging revealed a meningoencephalitis with fluid occupying the left middle ear, mastoid cavity and Eustachian tube (ET). The HRCT scan defined a cystic cochleovestibular malformation (IP-I, Sennaroglu, 2002) [Fig. 1].

A left exploratory tympanoscopy confirmed a CSF leak through oval and round window fistulas. [Fig. 2]

Subtotal petrosectomy was performed: both anomalous pathways were surgically sealed, the external auditory canal was closed as a blind sac, Eustachian tube obliterated, with peristomeum reinforced with muscle and cemented in place by bone wax. Autologous fat harvested from the abdomen was used to obliterate the cavity, treated with an antibiotic agent and skin closed in layers. [Fig. 3]

Because of high frequency relapses, in presence of a wider caliber of the Eustachian Tube in the involved ear [Fig. 5] a closure of nasal opening of the ET was performed, using left lower turbinate mucosa as sealing graft. [Fig. 6]

Conclusion: The close relationship between upper airways and middle ear can spread infection to the brain, using ET and inner ear malformation as a favorite route. Accurate diagnosis and closing off all connection with the external environment are mandatory in order to avoid life-threatening consequences.