Doctors should carefully distinguish between Hemophagocytic lymphohistiocytosis (HLH) and enteroviral infection to treat each appropriately

childhood disability LINK

Summary

Though they have very different prognoses and treatment protocols, it is difficult to distinguish between enteroviral infection and hemophagocytic lymphohistiocytosis (HLH) because they share similar symptoms and laboratory findings. Treatment for HLH has high risks, especially in newborns, so doctors should exercise extreme caution when making the diagnosis for HLH and deciding to carry out its treatment. This study presents three newborns diagnosed with a severe enteroviral infection according to reverse-transcriptase PCR (rt-PCR) and one newborn diagnosed with HLH with no enteroviral infection. Of the three newborns with enteroviral infections, all presented some signs of HLH, but only one was ultimately diagnosed and treated for the disorder.

What families should know

Enteroviral infection is the most frequently diagnosed viral infection among newborns and is treated according to its severity. Hemophagocytic lymphohistiocytosis (HLH) is a rare but fatal disorder of an exaggerated immune response, treated with chemotherapy or immunosuppressive agents. HLH must be triggered by an initial infection, and in newborns, this most often comes from enterovirus, so newborns with HLH should always be evaluated for enterovirus. However, because HLH is so rare, it should only be diagnosed with great care in newborn patients displaying enteroviral infection.

What practitioners should know

Because enteroviral infection and HLH display such similar clinical and laboratory features, it is difficult to establish a foolproof set of criteria to diagnose HLH in newborn patients displaying enteroviral infection. This leaves doctors to exercise their good judgment in making the diagnosis. It has typically been thought that while enteroviral infection does affect the central nervous system (CNS) in newborns, HLH does not have this effect until later in life, providing a possible means for distinguishing between the two. This study, however, suggests that even newborns can display CNS effects of HLH. Though it is not currently common practice, this study of four newborns indicates that the degree of immune system attack on red blood cells in the bone marrow, or hemophagocytosis, may be a useful criterion to incorporate for making HLH diagnoses. Finally, this study suggests that immunoglobins administered early upon presentation of an inflammation response may provide a safer alternative to chemotherapy or immunosuppression in newborns, though further research is necessary to confirm this.

Reference

Lindamood, K., Fleck, P., Narla, A., Vergilio, J., Degar, B., Baldwid, M., et al. (2011). Neonatal enteroviral sepsis/meningoencephalitis and hemophagocytic lymphohistiocytosis: Diagnostic challenges. American Journal of Perinatology, 28, 337-346.