

X-linked hyper IgM syndrome is estimated to affect about 2 in 1,000,000 newborn boys. The autosomal recessive forms of hyper IgM syndrome are extremely rare. Hyper IgM syndrome type 2 is estimated to affect fewer than 1 in 1,000,000 people in the general population. The other forms of hyper IgM syndrome have only been described in the medical literature in a very small number of people. Because rare disorders often go undiagnosed, determining their true frequency in the general population is difficult. The X-linked form predominately affects males; the autosomal recessive forms affect both males and females. X-linked hyper IgM syndrome accounts for about 70% of people with this disorder.