

Segawa syndrome affects girls and women more often than boys and men. In sporadic cases (i.e., new mutations), women are affected four times more often than men. Women are also more likely to have severe symptoms than men are. The exact incidence of Segawa syndrome in the general population is unknown. Researchers believe that the disorder is often misdiagnosed or goes undiagnosed, making it difficult to determine its true frequency in the general population. Segawa syndrome and tyrosine hydroxylase deficiency, which is also known as autosomal recessive dopa-responsive dystonia, account for approximately 5-10 percent of all cases of primary dystonia in childhood. Segawa syndrome was first described in the medical literature in 1971. It was originally called hereditary progressive dystonia with marked diurnal fluctuation.