Objective: To establish the adenosine deaminase (ADA) activity in women who had been pregnant with a central nervous system (CNS) anomaly child. Methods: The study group was a group of 68 women who had been pregnant with a CNS anomaly child. The study also included 68 control subjects who were matched for age, gestational age and body mass index. Maternal venous blood was collected for measurement of ADA levels. We defined the diagnostic sensitivity, specificity and area under receiver-operating characteristic curve for ADA. Results: Plasma ADA activity was significantly higher in study group at 12.3 (1.7-33.3) U/L compared to median values of 3.3 (1.1-34.4) U/L in normal pregnancy (p < 0.05). The cut-off point of ADA 5.9 > U/L was associated with the highest combination of specificity (58.8%) and sensitivity (86.8%). Conclusion: ADA activity in women who have conceived fetus with CNS malformations was significantly higher than that in normal pregnant women.