Histidinemia is one of the most common inborn errors of metabolism. Based upon newborn screening of more than 20 million infants in several countries, histidinemia is estimated to occur in about one in 11,500 births overall. The disorder appears to be most prevalent among people of French Canadian or Japanese descent. Based upon newborn screening reports, approximately one in 8,600 infants in Quebec and one in 9,500 infants in Japan are affected by the disorder. The abnormality begins at birth and affects males and females in equal numbers. Histidinemia is now thought to be a primarily benign disorder. In some states in the United States (e.g., New York and Massachusetts) routine screening of newborns for histidinemia was conducted through blood or urine tests. However, newborn screening for histidinemia has been discontinued. A diagnosis of histidinemia may be made based upon increased levels of histidine in the blood or urine.