Tyrosinemia type I is a rare autosomal recessive genetic metabolic disorder characterized by lack of the enzyme fumarylacetoacetate hydrolase (FAH), which is needed for the final break down of the amino acid tyrosine. Failure to properly break down tyrosine leads to abnormal accumulation of tyrosine and its metabolites in the liver, potentially resulting in severe liver disease. Tyrosine may also accumulate in the kidneys and central nervous system. Tyrosinemia type I affects males and females in equal numbers. The prevalence has been estimated to be 1 in 100,000 to 120,000 births worldwide. In Quebec, Canada, the birth prevalence is estimated to be 1/16,000. The estimated prevalence in the Saguenay-Lac Saint-Jean region of Quebec is one in 1,850 births. In Norway, the birth prevalence is estimated to be 1 in 60,000 births.