"Tay-Sachs disease is a rare, neurodegenerative disorder in which deficiency of an enzyme (hexosaminidase A) results in excessive accumulation of certain fats (lipids) known as gangliosides in the brain and nerve cells. This abnormal accumulation of gangliosides leads to progressive dysfunction of the central nervous system. This disorder is categorized as a lysosomal storage disease. Lysosomes are the major digestive units in cells. Enzymes within lysosomes break down or ""digest"" nutrients, including certain complex carbohydrates and fats. When an enzyme like hexosaminidase A, which are needed to breakdown certain substances like fats, are missing or ineffective, they build up in the lysosomal. This is called abnormal “storage”. When too much fatty material builds up in the lysosome, it becomes toxic destroying the cell and damaging surrounding tissue."