How do you get Wolman's Disease?

We have an important enzyme referred to as Lysosomal acid lipase (LIPA or LAL) which is required to metabolize certain lipids in the body. Without the LIPA enzyme, certain fats may accumulate in the tissues and organs of the body. Wolman disease is a rare, genetic disease and is found in infants in the first few days of childbirth. This disease is related to the lysosomal storage disorders and defective lysosomal enzymes associated with wolman’s disease. This results in build up of certain fatty acids and also complex carbohydrates within cells of the body.

Normal Function of Lysosomes

Lysosomes are an important organelle which, to make it easy, you can think of as a recycling centre. Lysosomes contain a powerful hydrolytic enzyme that is used break down macromolecules and digest worn out cell components which helps maintain and repair cell structure.
TREATMENTS AND HELPFUL RESOURCES

- Specialist can make a specialized nutrition plan for those affected.
- If adrenal glands are not functioning properly, medications can be used to supplement the hormones normally.
- Genetic counseling is available for affected individuals and their families.

SYMPTOMS AND EFFECTS

- In the first few weeks of life is when Wolman disease is first present. Infants that are affected can develop bloating and/or swelling in the stomach area (abdominal distention) and also can have enlargement of the liver or spleen. In some cases, there may be a fluid build up in the abdominal cavity.

- Infants who suffer from Wolman disease tend to have serious digestive abnormalities such as malabsorption, which results in the intestines failing to absorb nutrients and calories to form food.

- Infants with Wolman disease can suffer loss of coordination of muscle and motor skills.
- This often gets progressively worse which leads to life threatening complications during infancy including very low levels of circulating red blood cells, liver dysfunction or failure and severe weakness.

MLD - Early presentation
- Galactosemia
- Tyrosinemia
- Alpha-1-AT defic.
- Niemann Pick C
- Cystic fibrosis
- NN Hemochromatosis
- PTC
- Bile acid defects
- Fructose
- Peroxisomal diseases
- Wolman's disease
- Mitochondrial disorders