

## Peroxisomes: A World Without Peroxisomes

### THE STORY

A newborn baby has been having frequent episodes of uncontrollable shaking. The baby is extremely weak and does not move around with any excitement or vigor. Its weakness prevents it from being able to effectively breastfeed. The infant does not seem to respond to any visual or auditory stimuli, which suggests that it's both blind and deaf. The infant has a number of unusual facial features including a high forehead, a very small jaw, abnormal looking eyes, and a very broad nose. Tragically, the infant dies within the next few months.

### Scientific Connection

This child died of Zellweger syndrome, a serious disorder that is lethal within the first few months of life. As you might guess from the description of this case, this child has numerous problems with multiple systems of the body. The infant is having seizures, it can't see or hear, and it is extremely physically weak. While the sequence of events that lead to these numerous abnormalities and characteristic facial features is currently unknown, the cause of it all is defective peroxisomes. Peroxisomes are organelles that have multiple functions within a cell. These include preventing damage caused by oxygen radicals, as well as playing a critical role in converting certain fats into biological energy. The peroxisome itself is like a little factory where both of these processes take place. A factory, however, is useless without workers inside to do the labor. A steel mill with no steelworkers produces no steel, and a shirt factory with no stitchers or cutters produces no shirts. The physical effects of Zellweger syndrome are caused by peroxisomes with no proteins to work inside of them. Zellweger syndrome is the most severe of a class of disorders known as peroxisomal biogenesis disorders: the cells contain peroxisomes, but the proteins necessary to staff them never make it to work. Instead, these proteins can be found wandering aimlessly around the cytoplasm. Zellweger syndrome is a consequence of having no or very few functioning peroxisomes and its severity is a sign of just how important peroxisomes are to the proper workings of the body.

### ↪ *Take Home Message* ◀

**Peroxisomes perform important functions related to the minimization of oxidative damage and the utilization of certain fats to make energy. The physiological effects of non-working peroxisomes are extremely severe and often fatal.**