

## Cellular Organelle Disorders - Readings

Researchers identified the cellular defect behind cystic fibrosis in 1989. In this disorder, lung and pancreas cells trap salt within cells. The salty cellular interiors draw moisture in from surrounding tissue, drying out the mucus until it is so sticky that it clogs organs. Several new treatments, including a healthy gene introduced into the lungs in a nasal spray, target the illness at the cellular source. Compare the inside of cystic fibrosis affected cells to normal, surrounding tissue. Are these cells hypertonic, hypotonic, or isotonic to their surroundings? What organelle or organelles may be impacted by cystic fibrosis? Explain your answer.

Michael was a pleasant, happy infant who seemed to be developing normally until about six months of age. Able to roll over and sit for a few seconds, he suddenly seemed to lose those abilities. Soon, he no longer turned and smiled at his mother's voice, as he had before, and he did not seem as interested in his mobile as he once was. Concerned about Michael's reversals in development, his anxious parents took him to the doctor. It took exams by several specialists to diagnose Michael's Tay-Sachs disease, because, thanks to screening programs in the population groups known to have this inherited illness, fewer than ten new cases appear each year. Tay-Sachs patients are not able to break down fatty waste material that builds up on nerve cells. His nervous system would continue to fail, and he would be paralyzed and unable to see or hear by the time he died, before the age of four. What organelle or organelles are likely responsible for Tay-Sachs disease? Explain your answer.

Pompe disease is a rare (estimated at 1 in every 40,000 births), inherited and often fatal disorder that disables the heart and skeletal muscles. Patients with Pompe disorder are not able to break down large macromolecules such as glycogen into smaller molecules like glucose. This leads to increased muscle weakness and a lack of the monomers needed for cellular respiration. What organelle or organelles are likely responsible for Pompe disease? Explain your answer.