# Not Your Mom's Genes

You just learned about a case in which mitochondrial DNA was inherited from both parents. Researchers now know that this might be much more common than once thought: As many as 1 in 5,000 people might inherit their mitochondrial DNA from *both* parents, creating a mixture of mitochondria inside of cells. How does this happen?

Here's how the process *normally* works. Just like most other cells in the human body, sperm have their own mitochondria, which help power the movements of their tails as they swim towards an egg (**Figure 1**). But when it comes to fertilization, the sperm's main contribution is the DNA in its nucleus, which is contained in the sperm's head region. When egg and sperm come together, most of the sperm's midpiece, or neck, and tail are lost. If any mitochondria make it through, they are quickly destroyed. Under most circumstances, this leaves only mom's mitochondria in the mix. Clearly something unusual was happening in the family you read about.

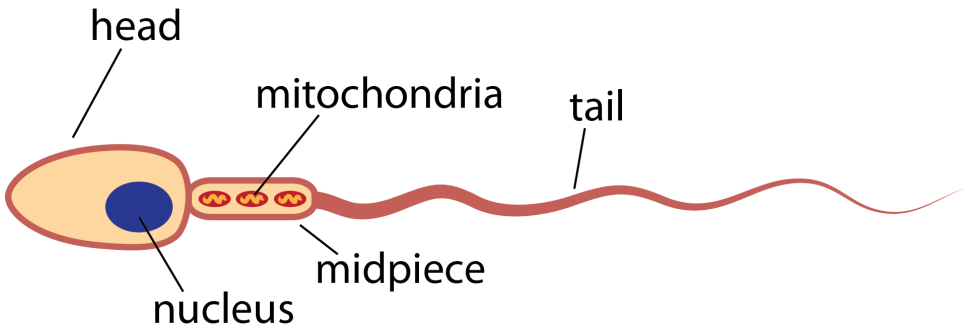


**Figure 1. Anatomy of Sperm.** Sperm contain all their nuclear DNA in their head. The mitochondria in their midpiece, a neck-like section below the head, produce ATP to power the tail, which helps the sperm swim toward the egg. The mitochondria do not usually enter the egg, and if they do, they are usually destroyed.

Take a look at the pedigree in **Figure 2** that matches the family you just read about. Each color and symbol represents a set of mitochondrial DNA. Stripes indicate that the individual has more than one set of mitochondrial DNA in their cells.

There are five nuclear families (meaning two parents and one or more biological offspring) in this pedigree. The nuclear families are shaded and labeled A, B, C, D, and E. Notice that three of these families, B, D, and E, follow the typical rules of mitochondrial inheritance: all the kids get their mitochondrial DNA from mom. The other two cases, families A and C, are strange: both parents passed their mitochondrial genes on to their kids.

When the researchers looked at the mitochondrial DNA of all these individuals, they didn't find any mutations that could explain this strange pattern. They concluded that it had to be a mutation in the *nuclear* DNA that was causing the unusual inheritance of *mitochondrial* DNA. Further analysis led the researchers to conclude it was a mutation that affected males.

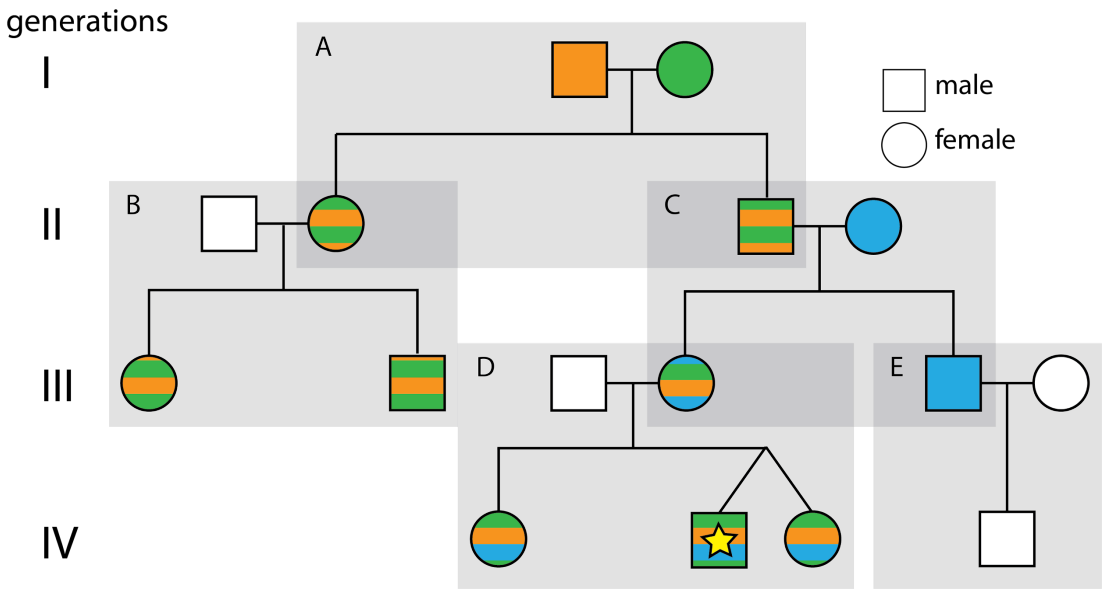


**Figure 2. An Unusual mtDNA Family Pedigree.** The four-year-old patient (starred) whose genetic tests started the whole story is marked with a star. He and his siblings (generation IV) all had mixed mitochondrial genomes, which they inherited from their mom (generation III) (family D). The boy's mom was a mix of her parents (the boy's grandparents, generation II) (family C). The boy's grandfather and his sister were mixes of their parents (the boy's great-grandparents, generation I) (family A). *Source:* Figure modified from Luo *et al.* 2018.

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Sperm mitochondria can be destroyed at multiple steps. The researchers are still figuring out exactly how the mutation they have discovered enables sperm mitochondrial DNA to remain intact within the fertilized egg. For now, it remains a mystery!

### Reference

Luo *et al.* Biparental Inheritance of Mitochondrial DNA in Humans. *PNAS,* 2018.

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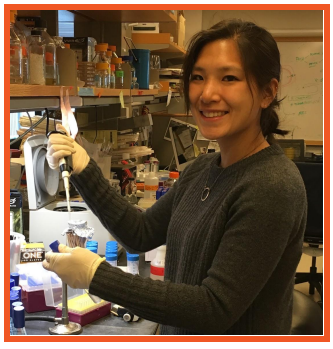

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