

## A Real World Example of Managing Genetic Information— The Case of Tay-Sachs

Most Americans recoil when they hear the word “eugenics,” and conjure images of Nazi scientists weeding out non-Aryan stock from the gene pool. Nevertheless, powerful gene tests soon will provide us with information that will challenge our ability to interpret and confidently make choices. Genetic science has already produced a list of tests to detect diseases caused by single, detectable genes. Some of these diseases are heartbreakingly painful and often fatal: Tay-Sachs syndrome, sickle-cell anemia, Huntington’s chorea (the disease that killed folksinger Woody Guthrie), and cystic fibrosis. In these diseases, the parents each have one good gene and one bad gene.

Although they themselves are usually perfectly healthy. Parents with recessive genes like these risk a 1 in 4 chance of having an affected child. But society may attach a stigma to being a carrier of a genetic disease, as was the case in the early days of testing for sickle cell anemia (most common among African-Americans). The trait became seen as identical to the disease. Healthy carriers were discriminated against by employers, including the U.S. Air Force, which did not allow carriers to become pilots.

Tay-Sachs disease occurs more frequently in the Ashkenazi Jewish population (Jews descended from ancestors in Eastern and Central Europe)

than in the general population. After Rabbi Josef Ekstein lost five children to Tay-Sachs, he decided in 1985 to take action to spare others this tragedy. Since traditional Jewish communities allow neither birth control nor abortion, Rabbi Ekstein saw preventing “at-risk” marriages as the key to preventing Tay-Sachs.

According to Rabbi Ekstein, people avoided testing for fear of being labeled Tay-Sachs carriers. So he went to work on a testing system the community would accept. The Dor Yeshorim Program in New York offers testing at schools and other routine sites to reach a large percentage of the community. The results are not revealed, even to the individual. Each person gets a coded ID number. Program leaders say that a couple, long before even considering marriage, can contact Dor Yeshorim and check their compatibility. By providing their two ID numbers, a couple can find out, anonymously, if they are at risk for having children. By withholding individual results, the carrier stigma is avoided but the information needed to prevent the births and deaths of Tay-Sachs children is available.

Some geneticists and medical ethicists question a program that attempts to arrange marriages according to genetic test results, while concealing information

about carrier status if only one partner is a carrier. However, the rate of Tay-Sachs disease in the Ashkenazi Jewish population has fallen below the rate of the general population. So, every year, representatives of the Committee For Prevention of Jewish Genetic Diseases go to Orthodox high schools and offer students a blood test. Jewish communities around the world have adopted the Dor Yeshorim model with its emphasis on incorporating community values into decisions about the uses of genetic information. This type of understanding will be needed as researchers devise new genetic tests.

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