Insular Jewish sect embraces technology to save its own

AP PHOTO / JOE KOHEN

Ultra-Orthodox Jews have developed a genetic screening system *that has greatly reduced the rate of Tay-Sachs births.*

BY DEBORAH PARDO-KAPLAN

Fifty years ago, children with Tay-Sachs disease filled a 16-bed unit at Kingsbrook Jewish Medical Center in Brooklyn, N.Y.

Tay-Sachs disease is a fatal disorder of the central nervous system commonly associated with the ultra-Orthodox Ashkenazi Jews from Central and Eastern Europe. Transmitted only through heredity, Tay-Sachs disease has no cure, and no treatment will prevent it from running its course.

The hospital became a hub for these young, debilitated patients who progressively lost their muscle control, vision and lives by the age of 5.

Those who worked at the hospital over the decades recall the overflowing ward. "I would have families with children at home waiting for bed space," said Fran Berkwits, a genetic counselor who worked at the hospital in 1957. Located within an ultra-Orthodox Jewish area of Brooklyn, Kingsbrook received many children from the local religious community. But, as preventative measures for the disease became widespread throughout the decades, doctors at the hospital saw fewer children suffering from Tay-Sachs. In 1996, the ward was empty for the first time.

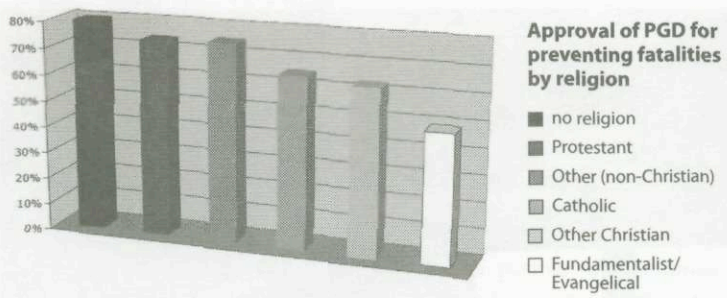
Ultra-Orthodox Jews, or Hassidim, of the 1960s and 1970s had yet to be educated about Tay-Sachs disease. At that time, the secular Jewish community spearheaded genetic testing and prevention, while religious Jews lagged far behind. Now, the two ends of the spectrum have switched. The younger generation of secular Jews has become less vigilant in prevention of the disease, despite the March of Dimes statistic that nearly one in 30 American Jews carries the Tay-Sachs gene. And the ultra-Orthodox have developed a genetic screening system so successful that it has greatly reduced the rate of Tay-Sachs births in its communities.

Ultra-Orthodox Jews have become extremely sophisticated in understanding genetic disorders, said Dr. David Rubin, a gastroenterologist involved with the Chicago Center for Jewish Genetic Disorders. "They probably lead the way in terms of genetic prevention," he said.

Tay-Sachs facts

- Tay-Sachs disease was first named after Warren Tay and Bernard Sachs, two 19th -century physicians who first described some of its symptoms.
- In the late 1960s, Dr. Shintaro Okada and Dr.John O'Brien at the University of California recognized the absence of the enzyme hexosaminidase A in Tay-Sachs children.
- The lack of the enzyme leads to a toxic build-up of a fatty substance in the brain's nerve cells. The child becomes progressively debilitated and dies between the ages of 5 and 8.
- Children with Tay-Sachs first show signs of the disease between 6 and 9 months. Symptoms include unending crying sprees, slow development and a loss of head control and vision.
- Tay-Sachs also has a late-onset form, often misdiagnosed as multiple sclerosis, which is not fatal.
- About one out of every 30 American Jews carries Tay-Sachs.

Controversial technology offers hope



Couples who have discovered both parents are carriers prior to conception can choose from several options: adoption, assisted reproductive technologies or preimplantation genetic diagnosis. Preimplantation examines embryos outside the womb. After testing for genetic abnormalities, such as the Tay-Sachs gene, healthy ones are reimplanted through in vitro fertilization. This option can cost $10,000, according to Jayne Gershkowitz of the Tay-Sachs and Allied Diseases Association. More than 1,000 babies have been born using preimplantion since its development more than a decade ago, according to a recent Genetics and Public Policy Center report.

Jewish rabbinic authorities have authorized preimplantation. The method is more controversial among other religious groups, such as fundamentalist and evangelical Christians, who often share common ethical views with Orthodox Jews. In a survey taken by the Genetics and Public Policy Center, these Christians rank lowest in support of preimplantation for fatal diseases, more than 20 percentage points below other Protestants. Fundamentalists and evangelicals also scored the highest among religious groups in wanting it completely banned.

— Deborah Pardo-Kaplan

Screening through Chevra Dor Yeshorim

Through a voluntary, confidential screening program called Chevra Dor Yeshorim, or "Association of an Upright Generation," unmarried Orthodox Jewish adults worldwide can be tested to find out if they carry the gene for Tay-Sachs. Each person tested receives a blood test and an identification number. Before dating, both members of the potential couple call Chevra Dor Yeshorim's automated hotline and enter their ID numbers. If both test positive for the Tay-Sachs gene, they are told they are considered unsuitable marriage partners because of the one-in-four chance their children will develop the disease.

Rabbi Joseph Eckstein, who lost four children to Tay-Sachs, founded Chevra Dor Yeshorim in 1983 in Brooklyn, six miles from Kingsbrook hospital. With laboratories worldwide, the organization has screened more than 184,000 people since its founding, according to Sarah Fekete, who works at the New York branch. To date, 646 prospective couples have been deemed incompatible. The confidential and anonymous nature of the screening works to avoid shameful situations, Fekete said, particularly the stigma of being a carrier.

"The option of not having children is not very acceptable from a Jewish perspective because of the obligation to be fruitful and multiply," said Dr. Daniel Eisenberg, a Jewish medical ethicist at Albert Einstein Medical Center in Philadelphia.

Counselors at Chevra Dor Yeshorim, which has offices in New York and Jerusalem, advise the two carriers of the risks, but they make their own decision whether to continue dating. "We don't know whether they follow the recommendations," said Berkwits, who now counsels part time for Chevra Dor Yeshorim. She knows whether her advice was heeded only when the organization receives another call from the same identification number for another potential match.

Chevra Dor Yeshorim succeeds in communities in which families arrange dating. In the mainstream Jewish community, though, "most people don't date that way," said Rabbi Michael Broyde, academic director of the Law and Religion Program at Emory University. "The broader Jewish community gets tested, finds out the results, and asks people they're dating," he added.

Tay-Sachs carriers

German-lineage Ashkenazi Jews, French-Canadians and the Cajun population of New Orleans have the highest rates of Tay-Sachs carriers. The disease affects one in 2,500 Ashkenazi Jewish babies. The carrier rate among these communities is one in 27, versus the general population rate of one in 250. Spanish-lineage Sephardic Jews have the same rate as the general population. If one parent carries the disease, there is a 50 percent chance the child will become a carrier without developing the disorder.

Since 1971, the Ashkenazi Jewish population in North America has reduced Tay-Sachs rates by 90 to 95 percent through genetic screening. Rates in Israel are also very low, as the government requires that couples get tested at its expense. In the last three decades, "more babies are being born with Tay-Sachs in the non-Jewish population than in the Jewish population," said Dania D'Achille, a genetic counselor at the Chicago Center for Jewish Genetic Disorders.

Tay-Sachs was first identified within the Jewish population. The American Jewish community raised funds in the late 1950s for research. As a result, doctors in California discovered the disease's enzymatic cause in the 1960s. The Jewish community then helped develop preventative genetic testing in the early 1970s.

The medical community attributes Tay-Sachs' origins to intramarriage in close communities, which increases the predominance of the disease. "It's based on arranged marriages in small communities that were very secluded in Europe," said Rubin, the gastroenterologist. "When you start mixing up the gene pool in a small number of people, you amplify recessive disorders."

Accustomed to low rates of Tay-Sachs throughout the 1970s and 1980s, younger Jews are now becoming slightly more lax about genetic screening. They remain less concerned because of the decreasing birth incidence, or they think intermarriage will prevent it, D'Achille said. They don't realize that the carrier status remains high.

"We have plenty of Tay-Sachs children born to interfaith couples," said Jayne Gershkowitz, executive director of the Boston branch of the Tay-Sachs and Allied Diseases Association. Founded in 1957 by five Jewish families in New York, the association developed genetic screening and continues to head research in genetic diseases. At her office, Gershkowitz sees not only Jewish but also Japanese, African-American, Hispanic, and Middle Eastern families. Gershkowitz receives about 20 new cases of Tay-Sachs per year, and estimates one-third of them are from the Jewish population.

She said she respects the work of Chevra Dor Yeshorim, but points out that it limits itself to one target group. "It's a very appropriate model for the observant Orthodox community," she said. She finds it challenging to reach the ultra-Orthodox communities who see her — and anyone who does not share their faith and lifestyle — as an outsider. "That's why Dor Yeshorim is so important," she said. "Because Dor Yeshorim is part of that community, it's a more accepting message. It's a peer-to-peer relationship."

Yet Chevra Dor Yeshorim has also raised awareness outside of the Orthodox communities, Gershkowitz said. She has received a number of calls from secular Jews mistakenly thinking they can receive free screening from Chevra Dor Yeshorim. She recommends they participate in a public genetic testing program. Chevra Dor Yeshorim absorbs much of the cost of testing for a panel of genetic disorders, but tests range from $150 to $350.

Jewish law and medical treatment

Most religious Jews are not opposed to genetic screening. They don't see the process as treading on divine territory.

"The Jewish tradition views all these activities as proper because we don't believe that the world was created as perfect," said Broyde. "We think our job is to perfect God's world."

Most clinical researchers and scientists think a combination of gene and stem cell therapies will prolong the lives of Tay-Sachs children. For now, though, genetic screening offers the most hope. "Every single child in the ultra-Orthodox communities is tested," said Berkwits, the genetic counselor, about how Chevra Dor Yeshorim makes the rounds at high schools in New York. "I couldn't believe it, but they do." ✖

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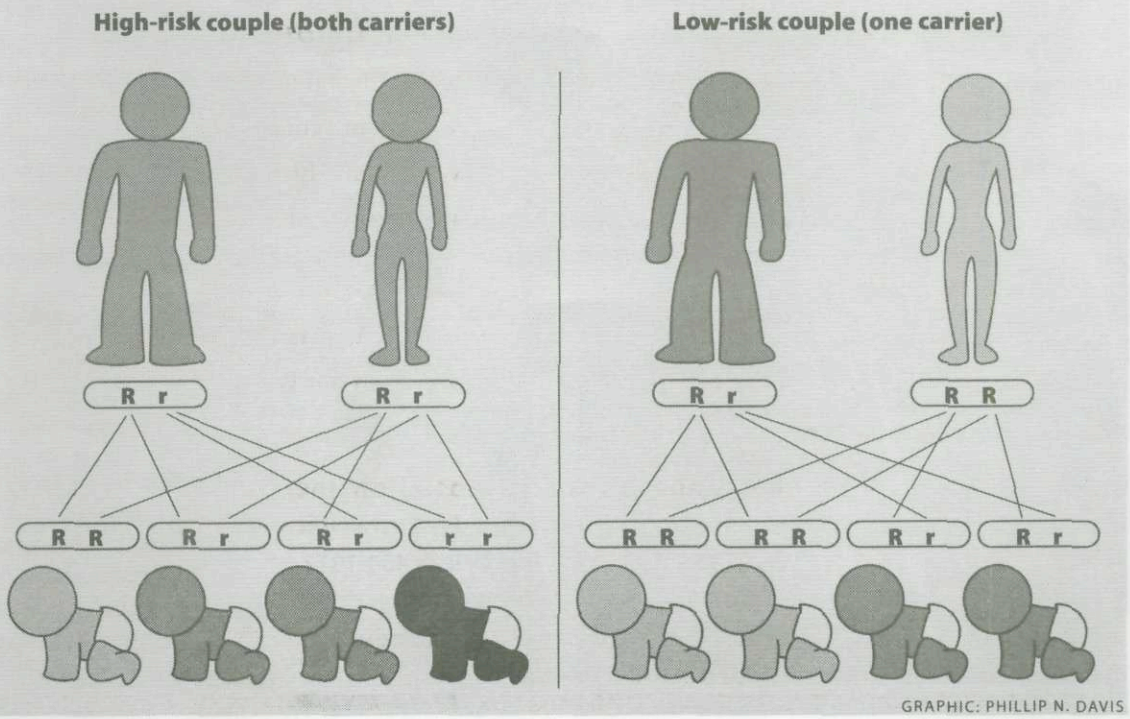
How recessive conditions are inherited

Almost all of our genes come in pairs, one inherited from each of our parents. A carrier of a genetic condition is a person who has one mutated or recessive copy of a gene and one normal or dominant copy of the same gene. A recessive condition like Tay-Sachs results when a child inherits two copies of an altered gene. Carriers don't have the disease, but can pass their mutated gene to their children.

With each pregnancy, high-risk couples (left) — in which both the man and woman are carriers — have a 25 percent chance of conceiving a child with that condition, a 50 percent chance the child will be a carrier and a 25 percent chance that the child will neither be a carrier nor afflicted with the disease.

Couples in which only one parent is a carrier (right) will never produce a child with a recessive disease. There is only a 50 percent chance in each pregnancy that the child will be a carrier. However, this is how the mutated gene stays in the population unnoticed.

Information courtesy of the National Tay-Sachs and Allied Diseases Association, Inc.



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