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Abstract: This article presents an interview with Josef Ekstein, director of the non-profit organization Committee for the Prevention of Jewish Genetic Diseases. Marriage is a minefield if your community is prone to lethal genetic diseases, your religion forbids abortion and prenatal screening is out of the question. For Josef Ekstein, an orthodox Jew living in Brooklyn, this was personal. He was forced to watch helplessly as four of his children fell prey to an inherited disease. Ekstein's child was 2 years old when he was diagnosed with the genetic disease Tay-Sachs. He went knocking on the doors of community leaders telling them that this was a problem and we had to do something about it. He wanted to encourage conventional testing, but community leaders were sceptical. So Ekstein developed an alternative, confidential testing method.

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The rabbi's dilemma

Marriage is a minefield if your community is prone to lethal genetic diseases, your religion forbids abortion and prenatal screening is out of the question. For rabbi Josef Ekstein, an orthodox Jew living in Brooklyn, this was personal: he was forced to watch helplessly as four of his children fell prey to an inherited disease. As he told Alison George, the only choice was between giving up or fighting back and finding a solution

How did you get involved?

My son was born in 1965. For the first six months of his life he seemed perfect. Then I noticed he was losing some of his capabilities. He got all floppy and couldn't support himself. I started to run around asking doctors what was wrong and they said: "He's a lazy child, don't worry about it." As the days passed, it got worse. He started having seizures, he couldn't move, he couldn't swallow and eventually he went blind. My child was 2 years old when he was diagnosed with the genetic disease Tay-Sachs. This was the first time I'd heard of it. There is no cure. My son suffered terribly and died at the age of 4.

Did you or your wife realise that Tay-Sachs was carried in your families?

No. Although this disease is relatively common among Ashkenazi Jews from eastern Europe like me, the families probably didn't know it. In times past, without antibiotics, children died

all the time. A child with Tay-Sachs has a weakened immune system, so would pick up an infection more easily. If a child died this way, parents would not have known about the underlying genetic disease.

What happened once you and your wife realised you were carriers?

It was a horrible waiting game. I can't tell you the pain of carrying a baby for nine months then having to wait for six or seven months to see if this child is going to be healthy or going to die. Babies with Tay-Sachs are usually very beautiful, with big blue eyes, and this makes the pain even greater. My second child, a girl, also had Tay-Sachs. I don't know how I got through it. My strategy to overcome this tragedy was my faith, and to look forward, not backwards. I am not very eager to look back.

What causes this disease?

Tay-Sachs is a lysosomal storage disease. Children with these diseases are missing an enzyme which clears waste products from the body. Without this enzyme, the waste products accumulate: in the case of Tay-Sachs, in the brain and nervous system. As waste builds up, there is increased pressure on the nervous system, and eventually the child dies.

Weren't there tests for Tay-Sachs?

A test was developed in the 1970s. But it was not used by the orthodox Jewish community because abortion is forbidden, so prenatal screening was out of the question.

So you were in a bind. What happened next?

In 1983, yet another of my children was diagnosed with Tay-Sachs, the fourth one. When you get one shock, then another, then another, either you give up and die or you fight back. I thought: "I don't want another person or another family to go through that. No matter what, this has to stop." I knew there was a way around this problem and that the transmission of Tay-Sachs was preventable. But first I had to get our community to admit the existence of genetic diseases. Because of the stigma, everyone was in denial.

Why was that?

A disease that runs in the family was a very taboo subject in our community. Families who had children with diseases felt stigmatised and didn't talk about it for fear that their healthy children would not be able to marry. I know of two brothers who had children with Tay-Sachs, but neither told the other. I heard of a doctor and his wife who had a Tay-Sachs child and would hardly ever go out. There was only one person they trusted with their secret, so he was the only the person who could baby-sit. Many ill children were sent away to care homes. I did this myself. I was one of those people who had tried to deny the problem.

How did you go public?

I went knocking on the doors of community leaders, rabbis, anyone who was ready to listen to me and some of those who weren't, telling them that this was a problem and we had to do something about it. The point I made was that this was a problem for the entire community, not just for me.

How did the community react?

At first they didn't like it. Parents of sick children were afraid of their dirty laundry coming out in public. The rest of the people did not know what I was talking about. But I knew that this disease was preventable, and the only way it could be prevented was if someone spoke out. Soon things started to change. People became more open about their problems. I was one of the first to admit my problems, and I would like to think that I helped to start this new wave of openness.

Did you have a plan?

At first I wanted to encourage conventional testing, but rabbis and community leaders were sceptical. They feared that if we identified carriers we would do more harm than good, so we developed an alternative, confidential testing method. I had to learn about genetics the hard way, by teaching myself. It was difficult, but if there is a will, there is a way.

How does your system work?

We call our screening system Dor Yeshorim, and it is based on the genetic compatibility of couples. DNA from both partners is screened for genes that can cause serious recessive

diseases. On average, 1 in 4 children born to a couple with the same recessive gene will have such a disease. Before a couple decide to get engaged we can tell them if they are genetically compatible ? whether or not they carry a mutation for the same disease. Unfortunately our system can only deal with recessive diseases. There are many more ethical and practical questions with dominant traits.

How do you do this test?

We test young people in schools and colleges, long before they consider serious relationships. The screening is carried out by accredited laboratories, and we insist on confidentiality. At the time of the test, every person is given a coded number, and Dor Yeshorim keeps a record of this number, the participant's date of birth, and telephone number. We do not ask for the person's name and the results are not revealed at this time. Before two people become serious about each other, they contact Dor Yeshorim with both code numbers, their phone number and dates of birth and we inform them whether they are compatible or not. Even if one person in the couple is a carrier, the pair are still compatible and so we do not reveal that one of them has the gene. Incompatible couples are informed of their risks and are given as much counselling as they need to deal with the implications.

Why don't you give individuals the full information? What about the right to know?

People also have the right not to know, and using the Dor Yeshorim system means they have opted not to know. So we do not reveal the status of an individual. After all, if a person is a carrier, it has no bearing on their health and it never will. Research shows that this knowledge has a negative emotional impact on young people.

How did you get the tests off the ground?

At the beginning of Dor Yeshorim, we had much opposition. Only 45 people came forward to be tested and most of them did it out of pity for me. They felt they were doing me a favour. But the idea caught on and the next year we tested 175 people and the year after that 750, and it gained support from other rabbis. Now testing has become part of Jewish culture. There still is a small amount of opposition, but that is to be expected. The only people who attract no criticism are those who do nothing.

Has Dor Yeshorim worked?

Yes. We have the joy of seeing it succeed. Today, Tay-Sachs is almost non-existent in New York's orthodox community, and in Israel too. In the 1970s, the 16-bed Tay-Sachs ward in New York's Kingsbrook Jewish Medical Center was filled to capacity. Since 1996, the ward has had no Tay-Sachs patients.

Are there other diseases prevalent in the Ashkenazi community?

Yes. Cystic fibrosis is common, and diseases such as Gaucher's Fanconi anaemia, and familial dysautonomia (FD) ? a debilitating condition that impairs the nervous system.

Do you test for these?

In total, we now test for nine different diseases, with the option of testing for Gaucher's disease, a condition that is not always severe and one that can be treated, but only at great expense. As medical science advances, and tests become available for other recessive diseases, we will add them if there is a need. But we don't add the test just because it is available. We have to think of the needs of the community.

How many people has Dor Yeshorim tested?

So far we have tested close to 170,000 people, and more than 1300 of them were found to be incompatible. We have saved many families from the agony of having children with devastating, genetically carried diseases. The statistics are not complete but so far we estimate that, with the implementation of all 10 tests, approximately 1 in 100 couples are not compatible. We give these people many hours of counselling. Except for some rare exceptions, most of them do not proceed with the relationship. Since Dor Yeshorim began, no genetically diseased children were born to the parents who used our tests.

Have you got involved in medical research?

Yes. I realised I could help make scientific discoveries. For example, through connections within the Ashkenazi community and with the consent of those affected, I was able to make many samples available to researchers working on FD. There were over 80 families involved,

and these samples would have taken years for the researchers to collect. A group had been hunting for 10 years for the gene that causes FD. We teamed up with Berish Rubin of Fordham University in New York City and discovered the gene: it was identified soon after Dor Yeshorim got involved, and we have already included this in our tests. Dor Yeshorim also commissioned research that showed that an over-the-counter vitamin can help treat FD.

How do you feel about patenting tests?

Patenting can threaten the entire genetic prevention system. Yes, a small royalty should be paid to companies or individuals who make the discovery. However, patenting can destroy all the advantages made from a discovery, and prevent developments from benefiting mankind. Companies sometimes get greedy and charge way too much, which can prevent people from taking a test.

How good are we at dealing with the information emerging from the human genome project?

Unfortunately a lot less money is spent on ways to impart genetic information than on the discovery itself. The most important aspect is how the information about someone's genetic status is handled. If you don't do it right, then the information could do more harm than good. Dor Yeshorim takes into account many issues ? psychological, emotional and societal ? when it introduces a new test.

"Since Dor Yeshorim began, no genetically diseased children were born to the parents who used our tests"

PHOTO (BLACK & WHITE): Rabbi Josef Ekstein, an orthodox Ashkenazi Jew, was born in Hungary in 1944. His family left for Israel four years later, then moved to Argentina. Educated at the United Talmudical Academy in Brooklyn, New York, he became a rabbi in 1963 and supervised the production of kosher meat. He is now director of the not-for-profit organisation he founded, Dor Yeshorim ? the Committee for the Prevention of Jewish Genetic Diseases, and has co-written nine research papers, among them one for *Advances in Genetics* (vol 22 p 297)

PHOTO (BLACK & WHITE)

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As told to Alison George; Photographed by Marc Asnin

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