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Huntington's disease in the news and entertainment media - Part I: Stigma and genetic testing

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At Risk for Huntington's Disease

HD is a genetically caused brain disorder that causes uncontrollable bodily movements and robs people's ability to walk, talk, eat, and think. The final result is a slow, ugly death. Children of parents with HD have a 50-50 chance of inheriting the disease. There is no cure or treatment.

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Despite its status as an orphan disease unknown to many, Huntington’s disease occasionally comes into focus in the mainstream news media and the entertainment industry.

HD’s biggest exposure came March 18, 2007, when the Sunday edition of *The New York Times* featured a long page-one story on 23-year-old Katie Moser, who had tested positive for this devastating, deadly brain disorder and was confronting her fate.

On September 15, 2011, HD entered the discussion again in a *Dear Prudence* column titled “Deadly Family Secret.” Emily Yoffe, the author of the advice column for the online newsmagazine *Slate*, responded to a young mother who had just learned that her newborn baby boy was at risk for HD. (Click here to read more.)

The writer, who signed her letter to Prudence “So Devastated,” recounted how, a week after the birth, she learned that her mother-in-law had HD. Thus the young mother’s untested husband has a 50-50 chance of inheriting the disease, and, if he indeed has the abnormal gene, the baby would face the same risk.

That letter once again highlighted how stigma, denial, and ignorance plague the HD community (click here to read more).

Some sound advice

Yoffe provided some excellent commentary and advice, though, as noted below, I found other aspects quite frustrating.

As an HD-positive person, support group member, and 2011 Person of the Year of the Huntington’s Disease Society of America (HDSA), I completely agree with Yoffe that the mother-in-law should have informed her son about his at-risk status, especially given the fact that her own mother had died of HD.

The young mother and her husband are now racing to educate themselves about the disease and its consequences – an anxiety-ridden situation they could have avoided, or at least planned for, had his mother revealed the family history of HD in a timely manner. They could have arranged for PGD, or preimplantation genetic diagnosis, to assure that their baby would be born without HD.

From my own extended family’s experience with HD and my observation of many others in my 13 years as an HD advocate, I believe that full disclosure is always the best policy.

Yoffe recommended that the couple tap into the resources provided by HDSA and its network of support groups and Centers of Excellence for

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International Huntington Association
Huntington's Disease Drug Works
Huntington's Disease Lighthouse
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HDSA Orange County (CA) Affiliate
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HD Blogs and Individuals

Chris Furbee: Huntingtons Dance
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Family Services and Research. She also advised that they consult a genetic counselor to discuss genetic testing for the husband, adding that the wife should inform an at-risk, pregnant cousin about her aunt's diagnosis. With that knowledge, the cousin can test the fetus for HD.

A deliberate decision

Also, I was deeply relieved to read Yoffe's advice to the young father that "there is no rush about making that choice" about his own genetic testing.

My own experience with testing supports this approach.

After learning of my mother's diagnosis the day after Christmas 1995, I wanted to get tested immediately. But after speaking with my mother's geneticist and becoming involved in the HDSA-San Diego support group, I learned that I should not rush into testing. Testing presented enormous risks involving job security, insurability, and my psychological health. Ultimately, I waited until 1999, when my wife became pregnant, to learn my fate. In 2000 our daughter tested negative for HD in the womb.

The omission of research

However, I disagree with other aspects of Yoffe's response.

She omitted, or perhaps didn't know, that a man often passes on a worse form of HD, sometimes causing a gene-positive child to develop juvenile Huntington's disease. Thus the mother's failure to inform "So Devastated" and her husband presents potentially even more devastating implications for their new family.

While Yoffe properly described HD as a "particularly cruel" condition, she failed to mention the huge strides made in research to combat the disease. That research provides immense hope for people such as "So Devastated" and her extended family (click here to read more).

A skewed view of testing

As a result of this omission, Yoffe presented a skewed view of genetic testing.

In referencing HDSA guidelines, she wrote that "there is almost never a reason to test a young person for the disease, which tends to strike in middle age."

Despite the highly appropriate caution about not rushing into testing, genetic testing does play an increasingly important part in the solution to Huntington's disease. Simply put, scientists, physicians, and drug companies need at-risk, gene-positive, and affected HD people to participate in research studies and clinical trials in order to understand the disease more fully and to test the safety and efficacy of potential drugs. (Click here to read more.)

Testing for HD directly benefits the effort to find treatments to make HD manageable like diabetes or perhaps even to bring about a cure.

By getting tested, at-risk people provide hope – for themselves and for the tens of thousands of people around the world affected by HD.

Treatments 'on the horizon'

Although many people in the HD community, as well as journalists, still have not perceived this message, it has now existed in the public domain for a number of years.

A prominent example came in September 2005, when Dr. Martha Nance,

the director of the HDSA Center of Excellence at the Hennepin County Medical Center in Minneapolis, responded to my own deep fears about HD by demonstrating in a *Washington Post* article that "treatments for neurodegeneration [brain diseases] are on the horizon."

Similarly, as I have illustrated with my own experience, at-risk and gene-positive HD people have given testimony regarding the importance of research studies and clinical trials.

Testing no longer just a personal matter

Yoffe's *Dear Prudence* column on HD effectively personalized how a "deadly family secret" can devastate successive generations. It reminds us of the urgent need to end the stigma associated with HD (and, by extension, with other neurological diseases).

Rightly so, Yoffe points out the great importance of sharing the truth about a genetic test with other family members and taking advantage of the information and services provided by HDSA.

However, I believe that HD and genetic testing are no longer just personal matters to be interpreted and confronted alone. They involve and impact the entire HD community – including the mutually beneficial, inextricable ties between the researchers and the patients (and potential patients like me and that letter-writer's husband and son).

To defeat HD, we can never forget the big picture of our quest.

(In Part II, I will reflect on a recent episode about HD and suicide on the TV show *Private Practice*.)

Posted by Gene Veritas at 10:29 AM

Labels: advocate , at risk , Center of Excellence , clinical trials , denial , gene , gene-positive , genetic , genetic counseling , genetic testing , get tested , HD-positive , HDSA , Huntington's , PGD , stigma , support group

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