Hello, it’s Andrew Schorr broadcasting live from Seattle as we continue our series today on Huntington’s disease, a fairly uncommon genetic condition; a scary one for families touched by it. Special thanks again to the Huntington’s Disease Society of America for helping us with these programs. We have talked in other days; we have given you an overview a couple of months ago with Dr. Jane Paulsen from the University of Iowa; be sure to hear that replay, it is very helpful for you on our website www.patientpower.info; but then earlier in the week we talked about it in children, and we had Dr. Martha Nance from Minneapolis and Dr. Sandra Kostyk from Ohio State who spoke about it there. Yesterday we were talking about in the prime of life, and we met Dr. Karen Anderson and her patient Alan Dogget in the Washington D.C., Maryland/Virginia area.

Well today we are going to go on, and we are going to introduce you to research. We’re also going to talk about Huntington’s much later in life, and how it affects people even looking at end of life when you are much older with Huntington’s, and I want to introduce you to someone who joins us who is beloved by the Huntington’s Disease Society of America, and he loves them, and that’s a Manhattan geriatrician internist, Dr. Anthony Lechich who joins us from Manhattan, which is my old stomping grounds where I grew up. Dr. Lechich thanks for joining us.

My pleasure, it’s a pleasure to be on.

Sir so you have been dealing with families touched by Huntington’s over so many years, the decades at your practice. I know you’ve been practicing about 30 years. Fortunately there are people who have it much later in life and so it’s something to be dealt with, but it doesn’t mean that they couldn’t live a long life, right?

That’s correct. Huntington’s as you probably heard in the earlier broadcasts can begin at all stages of life from children in the juvenile form as late as in the 80s, and the course of
Huntington’s can run generally around 25 years, with some variation depending upon certain other factors. The general age that we deal in our nursing home with though is onset somewhere in the third or fourth decades, and carrying that then forward for 20 years or so.

Andrew Schorr:
What we are going to do is talk with you later about how that course may go, how you can help people with both standard things and other things that you’ve explored or in research. We are going to be hearing from Dr. Jody Corey-Bloom in a second on tape. She’s flying to Europe today so she couldn’t be with us in person, but she is a leading researcher at the Huntington’s Disease Center of Excellence at the University of California San Diego Medical Center.

So we are going to talk about research, and we are also going to talk about managing it as people get older, and that’s sort of home base for Dr. Lechich. I’ll tell you what Dr. Lechich, if you don’t mind let’s go ahead and listen to that interview with Dr. Jody Corey-Bloom, get up on research, and give people guidance on participating in clinical trials because the good news is there are a lot of clinical trials going on to help families touched by that, and then we’ll come back to you.

All right let’s go ahead Ray and listen to that interview with Dr. Corey-Bloom, and then we’ll be back live right after that with Dr. Anthony Lechich from New York City.

Taped Interview with Dr. Jody Corey-Bloom on Clinical Trials

Andrew Schorr:
Joining us now is Dr. Jody Corey-Bloom who is Director of the University of California San Diego Huntington’s Disease Center of Excellence, and she is a neurologist there. She is very experienced and very well known in the treatment and the research of Huntington’s disease. Thank you so much for joining us doctor.

Dr. Corey-Bloom:
Oh it’s my pleasure Andrew.

Andrew Schorr:
So we are talking about really where research is headed. Tell us about some of the trials. Maybe let’s start with an overview. Should people be hopeful, and if so how come?

Dr. Corey-Bloom:
Well I think people should be really hopeful. I like to tell people that I started in the Alzheimer’s business actually doing quite a number of Alzheimer’s clinical trials, and recently I actually am doing more clinical trials for Huntington’s disease than Alzheimer’s disease, and that’s sort of often surprises people. They say, ‘Really, you know we didn’t
realize there was anything in clinical trials for Huntington’s disease.’ So it is a very exciting time, and I think people with Huntington’s disease have very good reason to be hopeful that we will have something in the not-too-distant future.

Andrew Schorr:
Well that would be great. Now give us a window into some of the trials that you have underway now; what they’re for, who might qualify, how people find out whether it’s right for them?

Dr. Corey-Bloom:
So you know in addition to clinical trials there actually are some observational trials going on, and I wanted to mention them first because they are really quite important. Everyone’s always interested about drugs, but the observational trials are also very important. These are sponsored by the Huntington’s study group, and they give us the opportunity to look at individuals who are affected by Huntington’s disease or part of an HD family, and to be able to follow them, to collect data over time looking at the natural history of Huntington’s disease, and really adding to our knowledge about the disease itself. I think with time we will likely turn these into clinical trials too, but I think it is actually very important to have the ability to be able to look at some of the earliest signs of Huntington’s disease and sensitive tests for clinicians to detect symptoms and factors that influence age and onset and those kinds of things. So those are ongoing and they are sponsored by the Huntington’s study group and actually quite exciting.

At our own site we are actually just beginning a functional MRI study, and this is actually sponsored by the HiQ Foundation, and we are excited about that too because we believe that we are going to actually be able to look at some of the earliest changes that occur in the brain, in the brains of people who are at risk for Huntington’s disease and who later go on to develop Huntington’s disease, but some of the clinical trials that are ongoing for treatment currently include the Memantine trial which is actually a three-site study. It’s what we call “investigator initiated.” So that means that several of us got together and kind of pitched the trial, if you will, to the drug company and asked them to support it, and we were able to design the study the way we wanted it to be and to design the outcome measures, and we have complete control over the data too. So that’s pretty exciting to us. So the three sites there are University of California San Diego where I am, but also Kansas, the University of Kansas, and Johns Hopkins, and so we are looking at memantine versus placebo over three months and then six months, and the ability of memantine to improve cognition, behavior and also function.

The UCSD site has actually completed its recruitment, but there are still some spots available at Kansas and also Johns Hopkins in case anybody is listening and they are interested.
Andrew Schorr: How would they go to the websites for those Huntington’s centers? Would that be the best?

Dr. Corey-Bloom: Yes, I think, or probably if they just went to www.clinicaltrials.gov, which lists all of the clinical trials currently ongoing for Huntington’s disease. Then there are also numbers; there are contact numbers for Hopkins and for Kansas for that study.

Andrew Schorr: Tell me about the FDA approval, for if I say this right, tetrabenazine. What does that mean? What’s the significance of that?

Dr. Corey-Bloom: Yes, so tetrabenazine is a medication, it’s an old medication that’s really been brought back, but tetrabenazine was actually really championed for many years by Dr. Joseph Jankovic in Texas, and Joe really felt that this drug was wonderful for chorea, but also many other what we call hyperkinetic movement disorders. So tics, for example, tremor, those kinds of things. So he really came up with the idea of using if for Huntington’s chorea, and the study was really quite positive with the respect to chorea.

Unfortunately I think the FDA is wrestling a little bit with it because I think they are questioning whether or not just improving the chorea of Huntington’s disease is a big enough intervention, and I think that they’re also concerned about depression and suicide as a result of the medications. So there are some issues, and I know it hasn’t been approved just yet, but we expect it to be any day.

Andrew Schorr: What’s your perspective on it?

Dr. Corey-Bloom: Well I really like the medication. I think it’s really important to use it wisely, and I think there is going to have to be significant training and information available with the medication, but it really works quite well for the chorea itself, and I think it’s just important to know how to use it carefully so that you don’t give a person too much of it.

Andrew Schorr: We spotted some articles just within the last week or two about, if I understand this right, intracellular antibody or something they are calling “intrabody.” So people come across this, and it was talking about a protein that could be used. Are you familiar with this?

Dr. Corey-Bloom: No, I’m not. So a protein that could be used to do what?
Andrew Schorr:
Well let’s see. “Scientists have created a tool for mopping up the clumps of mutant protein that drive neurodegeneration in Huntington’s disease.” This came out of Emory University, and they said that they had engineered a virus to make an intracellular antibody or intrabody against Huntington’s, the protein whose mutant form poisons the brain cells of people with Huntington’s.” I know this is early, but when we spot something like this we don’t know what to make of it, and maybe you could put that in a more global perspective for us?

Dr. Corey-Bloom:
Right. So it actually would be very exciting, and you know sometimes this work is done in animals to begin with.

Andrew Schorr:
It is in this case.

Dr. Corey-Bloom:
Yes, a transgenic, yes. You know the Huntington’s protein itself is thought to be somewhat toxic to the cells, and so if in fact they did have a way to sort of rid the cells of this Huntington’s protein, the hope would be that that would then translate into clinical improvement.

So one of the problems might be that the Huntington’s clump, if you will, itself is not the problem. So if it’s not and if in fact that there is just something wrong with the machinery of the cell, or some other processes upstream from the actual clump, then getting rid of the clumps may not actually make a huge difference. I think this interesting because it is something that we are also wondering about in Alzheimer’s disease for example. People talk about therapies to rid the brain of the amyloid plaque, and some people are questioning whether or not ridding the brain of the plaque will actually get rid of the disease, or is there something upstream of the formation of that plaque that is actually the real problem.

Andrew Schorr:
What is the frontier as you see it for Huntington’s research? So there are questions to be answered, but if you are optimistic what are we learning? I mean we’ve got the genetic test. Now we are observing people. There are some things to try to do for symptom management, but where do you think the breakthroughs might come? I know you might not know a single answer, but if it’s door number one, door number two or door number three, what are those doors?
Dr. Corey-Bloom:
Well I think that I and many would like to see the possibility of turning off the mutant gene because you know unlike many diseases, in Huntington’s disease we actually know what the gene is that is causing the problem. We actually can test for the size of the abnormal expansion, and so I think the hope would be that somehow we could find a way to really turn off that abnormal gene, and stop making the mutant Huntington’s. I think that would be very exciting, and that would probably be bringing about a cure quite honestly.

Andrew Schorr:
How do we help you get there?

Dr. Corey-Bloom:
Well I think a lot of progress is being made in terms of transgenic animals, and this has actually been done by a Japanese group in which the gene itself was turned off, and the transgenic mice themselves then actually made fewer clumps of the Huntington’s, had fewer cell losses in the brain and also showed better motor activity as a result. So this is a very positive thing because it actually in turning off the gene brought about clinical changes that were really relevant to Huntington’s disease.

You know I think now the big challenge will be, that’s a transgenic mouse, that’s a mouse in which you have control of the DNA. So this is before birth. So the challenge is going to be being able to do that in a timely fashion so that there hasn’t been significant disease, or finding a way to do it even perhaps once the gene, the abnormal gene has began to express itself.

Andrew Schorr:
Alright, so I guess bringing that back to our listeners, many families living with Huntington’s disease, how can we help you and your colleagues get to where you need to go to help our listeners?

Dr. Corey-Bloom:
Well I think right now for example we have many clinical trials going on for Huntington’s disease, some very exciting studies, DIMOND for example. I don’t know if you’ve heard about this. This is Dimebon. It’s actually a compound that was also studied in Alzheimer’s disease. We are actually doing a study of Dimebon in the Huntington’s study, and we are really hoping that this is actually going to improve cognition and also functioning in Huntington’s disease.

There are several other studies that are starting up, a study of creatine for example. There is going to be a coenzyme Q study, a large coenzyme Q study. So I think that there are many studies that are either underway or will soon be underway that are really quite
exciting, but we really need enough patients to join these studies so that we can fully enroll the studies and be able to look at the potential effects.

**Andrew Schorr:**
Here is a question we got from Connie in Ohio. She wonders, ‘Do scientists face greater challenges with Huntington’s disease since the onset is unpredictable?’

**Dr. Corey-Bloom:**
So that’s a very interesting question. Many of the neurodegenerative diseases actually have unpredictable onsets. For example although Alzheimer’s disease generally has it’s onset in the 70s and 80s, in fact there are many individuals who develop Alzheimer’s disease in their 50s, 40s and in some of the familial forms of Alzheimer’s disease even in the 30s.

In spinocerebellar degenerations for example there can also be significant variability in age of onset, but Connie is correct in that in Huntington’s disease more than any other neurodegenerative disease there can be great variation really depending on the size of the expanded mutation.

We know for example that the larger the mutation, the earlier the age of onset, and visa versa. The smaller the number of CAG repeats, usually the later the age of onset. That probably only explains 50 percent of the variance though, and there must be many other factors which we really don’t know at this point that impact on the age of onset.

**Andrew Schorr:**
Wow. Well there are a lot of questions to be answered. I’m sure you can count on people listening who want to team with you and researchers at the other Huntington’s Disease Centers of Excellence like the one you have at UC San Diego to try to help you get the answers for them and their offspring as well, their other family members. So I’m glad you started by saying you were optimistic, and Dr. Jody Corey-Bloom from UC San Diego, thank you for being with us. We wish you all the best with your work and thank you for the help you give current patients right now.

**Dr. Corey-Bloom:**
Thanks.

**Research Discussion with Dr. Lechich**

**Andrew Schorr:**
We are back live. Andrew Schorr in Seattle. I’m going to connect you with the man who is in the trenches in treating Huntington’s in New York City, Dr. Anthony Lechich. Dr. Lechich, so I don’t know if you are a fan of the TV show “CSI New York,” but I like to watch it because I used to be from where you are in New York, and they always sort of
solve the case at the end. When you listen to an interview like that with a leading researcher I guess we have the hope that they’ll sort of get their man. You know that they will solve the case. It sounds positive doesn’t it?

**Dr. Lechich:**
Absolutely positive. DNA is the answer to all of our prayers I guess. The genetic research that I’ve seen talked about in our own facility, and I have the high privilege of working with some of these great scientists like Dr. Corey-Bloom and Dr. Flint Beal and Dr. Jim Gusella in Boston and others, Nancy Wexler in Columbia and so on. These are superstars that are working in a very synergistic sort of a collaborative way on the cure and are getting more and more close as we come year after year to the annual meeting of HDSA. There is one tour de force, and I encourage anyone out there to attend, that they have every year. This year it’s in Pittsburgh this coming weekend actually, but the scientists line up on the stage, and they talk about Huntington’s and the research as it has progressed from year to year.

When I first stared going you know if was a much kind of less hopeful affair, but now things have really heated up, and there is a division of labor sort of like the Manhattan Project when they built the bomb where each expert in his area or her area works on their area. That’s sort of what they try to do with the Cure HD Project, and they have people at each center working on the effects of Huntington’s protein, the effects of the . . .

**Andrew Schorr:**
I want you to tell me more about that doctor. We have to take a commercial break right now. It’s our only one. We are going to hear a lot more from you as we continue, Dr. Anthony Lechich from New York City. We’ll be right back.

Welcome back to Patient Power, our third live webcast this week on the serious genetic condition Huntington’s disease. Families touched by it know it so well and worry about it. It affects about 30,000 in the United States. It usually begins, we heard on Monday, rare cases juveniles even as young as two but usually in young-to-mid adulthood with the slow destruction of brain cells that leads to involuntary movements, cognitive impairment and sometimes depression and paranoia.

We are going to talk about some of those effects, and it’s believed that there are about 150,000 people who have the mutations that cause the disease but have not begun to show clinical symptoms. I have a friend I mentioned yesterday I think, my friend who goes to the dog park. She confided in me that she has that. It is in her family, no symptoms and wondering is there another shoe that will drop and when. We’ve talked about that. Please listen to the replays on [www.patientpower.info](http://www.patientpower.info). There are already three hours there for you if we include the program with Dr. Paulsen a couple of months ago, but today we’ve been hearing about research.
We just hear from Dr. Jody Corey-Bloom who is winging her way to Europe as we speak, but we heard her on tape, and I think gave us a great background. I will just mention, now I’m a cancer survivor but I was in a clinical trial, and all these clinical trials typically or most of them are listed on www.clinicaltrials.gov, and she mentioned that. I think I’m here today because I was in a clinical trial that gave me a shot at tomorrow’s medicine today. It’s not always the way it works out, but you should inquire about that, and see if it could be helpful.

As Dr. Anthony Lechich was saying just a moment ago, big conference coming up this weekend, if you are listening live, so early in June in Pittsburgh. it’s the annual conference where patients, researchers and clinicians get together. You created this image for us Dr. Lechich of a real sign of hope it sounds like what you see coming.

Dr. Lechich:
Yes indeed. I think the team that has been assembled is superstar level, very dedicated. I think what’s wonderful about the conference is the scientists mingle with families and patients with disease, active symptoms, people actively choreic in the audience and so on. It’s really almost like a retreat for the nation to really exchange views and speak to each other and support each other, and that is so important in this process, which is a slow, evolving affair, which makes it particularly devastating. In other ways, unlike ALS for example, Lou Gehrig’s disease, which is sort of a short two years and death, there are positive things that are present with Huntington’s because some people who are coming into the pipeline with symptoms now absolutely could have their lives significantly altered in the course of the disease, significantly altered by therapies that we’ve heard about.

So this weekend it becomes a great source of information, and the HDSA website which we will give properly, will certainly have elaborate discussion about that. There are occupational therapists for swallowing, dietitians, social workers and so on, all great resources for the families to bring the disease out of the closet where it was and in many cases still is, into the public arena where the patients and the families can enjoy the support of other similar sufferers.

Listener Questions

Andrew Schorr:
Right. Well I’ve been talking, people probably think I’m a broken record, but one of my favorite shows, “House, MD” on the Fox Network, one of the young doctors on there has the HD gene. So in the big finale of the season she got her test result and I’m sure, and I’ve invited the producers to listen to this series, so Fox Network producers if you are listening take notes, but I’m sure it will be part of the plot line next season, and hopefully then millions of Americans will have greater understanding.
Let’s go to some questions Dr. Lechich, as people are sending them in, and many of them deal with a situation you handle all the time with people in the later stages, whatever their age is, the later stages of Huntington’s. This one is from Ann in Cadillac, Michigan. She writes, ‘I’m a social worker in a nursing home, and one of my residents who is 66 years old has HD. She becomes tearful easily and frequently screams for reasons as simple as wanting a drink of water. All staff attempt to anticipate her needs, but several of the nurse’s aides have come to me to say that they feel overwhelmed by her emotionality and behavior. She constantly apologizes for everything she does, and it really can be heart wrenching. She refuses to see a psychologist. Would counseling be a solution, or are we past that?’

**Dr. Lechich:**
Right. Well that is a typical type question. This is an example of an isolated case of Huntington’s in a nursing home. We’ve looked at that and try to be of help to people such as Ann who are dealing with the vagaries of Huntington’s. This individual patient is illustrating and demonstrating many of the typical challenges presented by Huntington’s patients at this stage.

She is having emotional lability or variability. That is something to expect. That is something to train to the staff to expect to happen without a known trigger frequently, although if we stay with the patient and have the same nursing assistance with the patient, they will frequently come up with behavioral plans and interventions that can minimize some of these emotional outbursts, but they can’t be eliminated. There is really no medication that can completely eliminate the variability.

Many of our patients at TCC, and just for the record we have 50 patients currently residing at Terence Cardinal Cooke Health Care Center here in New York with Huntington’s disease; some as long as 18, 19 years, and some just recently admitted. We’ve seen everything. We’ve seen this 100 thousand times with emotional lability and the aides having their own hearts wrenched by their helplessness to intervene successfully. So we need to reassure our staff and say this will pass, these are disinhibition-type symptoms.

If they become violent, if they become dangerous to staff or other residents they will require some medication, and we have virtually tried every known psychotropic medication. There is no particular magic bullet for these behaviors. You really rely on trial and error frequently. Also, again to redirect the patient sometimes simply arranging the room in a certain way, certain stimuli that may be aggravating or relieving can be used, but it will pass.

The other thing is there are stages of the disease which present tremendous challenges, but they don’t last. You know the patients will go through that, and whatever part of the brain is being affected and disinhibiting the behavior will eventually continue to be
destroyed really, and that behavior will cease generally. We’ve seen that and so the reassurance to the social worker and her staff is ‘Let’s do our best. Let the buck stop there.’

There is sometimes the requirement to go to a psych hospital for special interventions and protection and safety, but that is really the main reason for a psych hospitalization. If that happens it is incumbent upon us to educate the psych hospital because they may have limited experience. Here we are sort of blessed with a frequent flier reception at our neighboring hospitals, and we can educate them. I’ve given talks, and our psychiatrist has given talks. It’s a challenge, there is no question, and there have been many situations where nursing homes have chosen not to take HD patients, and we want to be out there to help any nursing home accept those patients, and not restrict or in any way prevent them from coming to a neighboring nursing home.

**Andrew Schorr:**
Dr. Lechich thank you for your devotion. Well here is the flip side of that from Sandra in Sacramento, California. She says, ‘My husband was diagnosed with Huntington’s about seven years ago. He is getting much, much worse, but refuses to go to a doctor. He is starting to get violent and out of control. I don’t know what to do. Being at this point in the disease is there any way of knowing how much longer he may live, or should he be hospitalized, and does research give us any lessons of what to do?’

**Dr. Lechich:**
Yes that’s an excellent scenario. Seven years ago means he is really in the middle. He may have in his particular manifestation of the disease a larger, violent phase, let’s say. She undoubtedly needs higher level of consultation even if he refuses. He needs to be seen because this violence could be self-injurious or injurious to her. This has been the scenario for us in many cases where the patient would need to be transferred to us.

I think in Sacramento we need to help Sandra look at resources in California. There are Centers of Excellence in California which will I’m sure be of help in identifying knowledgeable providers in the Sacramento area. If not, if she is really in a sense desperate, we need to help her find a willing participant physician or neurologist that we can educate and help with the management of the patient.

He is at that point which people do get to; not always, but some and that’s the challenge. The course of Huntington’s is variable as was previously mentioned by other speakers. It depends on a lot of things. It depends probably most on the extent of the repeats and how strong the gene is, if you will, but it also depends on the individual who gets it. Some people are literally unchanged personality-wise by it. We’ve seen that.

There is something to the idea that what you were before it hit you’ll become more of. You know if you are a rough and ready guy, a prize fighter, maybe you’ll become more
prone to that kind of course, but I’ve seen calm people turn into “Jekyll and Hydes” too with the disease. So it’s really a “fasten your seat belt and get ready,” but in the instance of Sandra, she is at the point where there will need to be some intervention, involuntary, for him.

**Andrew Schorr:**
Right, could be. Now we mentioned yesterday, and my friend Karen who has Huntington’s down in Orlando and had been on an earlier show with Dr. Paulsen, she runs a support group, and many of you may know her online. She called in, and we made the point that you are not alone through the Huntington’s Disease Society, online groups like Karen’s; you are not alone, and then again you can help bring education to your local neurologist for example who may not have experience with Huntington’s, or maybe you’ll be able to connect with a very experienced physician such as Dr. Lechich in Manhattan or the Huntington’s Disease Society Centers of Excellence; 21 of them I believe there are around the country. So there are places to go, and it is troubling and complex and tough on families, the whole genetic connection if you are related by blood and whether they get tested. All these issues we’ve talked about this week, but you are not alone.

Dr. Lechich here is another question. You know I wonder as a cancer survivor; I think anybody with any condition says, ‘Is this the disease at work, or is it me getting older or me having other heart problems?’ So here is a question from another Andrew, Andrew in Modesto, California. ‘Can severe medical problems trigger Huntington’s to develop earlier than normal? My partner’s having heart valve problems with kidney failure. His mother has Huntington’s, and since hospitalized my partner seems to be exhibiting symptoms. What can be made of this?’ So what’s connected and what isn’t?

**Dr. Lechich:**
Well that’s a very interesting question. I would only conjecture at the answer. Any predisposing condition theoretically would be exacerbated, especially one that has such a heavy emotional component. It’s not at all hard for me to conceive that these heart problems and kidney changes would accelerate the onset, or at least the appearance of the onset.

As Dr. Corey-Bloom mentioned there is a whole new wave of information coming by way of studying people who are gene positive; who have courageously subjected themselves to testing and about whom we are learning a great deal. Now these people who on the surface appear perfectly normal will on testing show signs of this and that.

Now that being said if that person undergoes a stress of huge proportion such as kidney failure and heart valves, it is not at all surprising to me that they would manifest symptoms of depression, of perhaps some motor changes. There are problems with kidney failure with weakness in nerve function and so on. So I would absolutely agree that he is being accelerated by this.
I also thought that there is a track that you are on with the normal CAG repeats that’s probably not going to be altered beyond perhaps the early manifestation that much, but that’s a complete conjecture at this point. I don’t think there is any research that looks at comorbidities with Huntington’s that I’m aware of, but that’s a fascinating question.

Andrew Schorr:
Okay, I have another question. Now you’ve been dealing with family so you have a number of people at the Terence Cardinal Cooke Health Care Center and so around those people are families where they either know or wonder about whether they, these other family members who you have contact with, have the HD gene. A couple of weeks ago President Bush signed the Genetic Information Nondiscrimination Act, as I’m sure you are aware. So New Yorkers usually have opinions sir. What is your opinion? What do you talk to people about, about whether they should be tested if they don’t know; mom, dad, brother, or sister are now in an institution?

Dr. Lechich:
Well I think that comes up a lot and we have the good benefit of Deborah Thorne at the Columbia University Center of Excellence as one of our genetic counseling resources. That is such an individualized question, and with so many nuanced turns and twists, testing really does carry, despite the new law, a lot of stigmas for certain individuals, and I think Jane Paulsen went through some of that in her discussion. I think our experience has been a fairly low-test rate.

There has been a major reluctance on some people’s family members to get tested, but in others for one very courageous woman, Katy Moser who has gone public with her positivity here, has made a point of showing the value and the positive aspects of having been tested, and she has gone on to become a spokesperson for that, but it’s not without its detractors. Some other family members may take issue with that. It has implications for them for example, and I think there is still law being written as we speak in cases of undiagnosed or untested individuals where people will contend that they refuse to test but they’re acting funny. So they could be labeled with or without the test as exhibiting some perhaps questionable behaviors for whatever reason the person saying that might have such as an employer or a spouse who is vying for custody of the kids and that sort of thing.

So it is an incredibly complicated question, and I think there are very strong arguments on all sides. I think probably the most important lesson though is to not make it so black and white. I think this was profoundly displayed by Nancy Wexler who is a great professor herself at risk, and who had not gone to be tested, at a national meeting mentioned that we have to think about HD as not an all or nothing phenomenon because there is such a spectrum, and it is such a humanly integrated thing in our patients and in our families.
I have to say, and I don’t know if I’m making that as clear as it should be made, but we must not consider “yes you’re dead; no you’re not” kind of thing. It’s like anything, including cancer genes and every other gene we all have. It is who we are, and it is absolutely essential for us to kind of accept that, and treat the patients and whatever we are treating with the same respect and dignity and concern and actually with the same rights as everybody has.

Andrew you’ve been through this yourself in a way with what you’ve said. You know there is a lot of strange turns that genetic diseases make us take, and I think the most important thing for me is that as medical director of this unit, we have to love our people as people to the last day and beyond, because their families are in need, and that’s it. I mean it’s not that complicated in a way, but this whole testing business is an ongoing saga. There is a lot of research and material on the websites on all of that.

Andrew Schorr:
Wow, Dr. Anthony Lechich you are so eloquent, and I think your 30 years of practice in medicine and helping families and the work you do at the Terence Cardinal Cooke Health Care Center there where you have many people who are... I bet, you know I know there are the trials and tribulations with the nurses and the aides and the staff, but it sounds like you all deliver care with a great deal of love, and I think that’s what we need.

In my own case with the leukemia that I have, there is a family connection. It’s one of the leukemias where there is somewhat of a pattern, and so we worry about that. Maybe not to the extent of people with the HD gene, but we worry about that.

Dr. Lechich I just want to ask, we got a question from Chloe from New Hampshire. Just briefly, do we know what percentage of HD patients are institutionalized? I know we were talking about 30,000 people with the actual diagnosis. Do you have any handle on people actually institutionalized?

Dr. Lechich:
Yes it is a question we’ve been grappling with for years. I’m going to go out on a limb and say less than 10 percent on that order of magnitude. That is a number though that is so impossible to drill into. We have sent questionnaires to families related to Centers of Excellence for example and have come up with, I can’t remember the exact number, but you know it is under 1,000 nationwide. So there are people with HD who were misdiagnosed who are still stashed away in mental institutions such as Woody Guthrie was, and in the earlier century practically all Huntington’s patients ended their lives in a back ward in a psych ward.

The number is difficult to come by, and I think what we seek to do with TCC is look not only at institutions like nursing homes, but look at the spectrum of support and where all
the 30,000 people in the pipeline are and how we can help them along the way and help them stay at the lowest level of care that they can live independently at.

Now there are a lot of reasons to want to do that, most of them where the patient should be obviously, but also financially. Obviously the patients who are in nursing homes cost the states a lot of money. So there is an incentive to keep people at the lower level of care, but the answer is a tough one, and I think families also do the yeoman’s work of keeping people in-house as long as possible, and some will end their lives within the family context, and they’ll never hit the radar screen. Jane I know alluded to the fact that our numbers probably are way low, and that we are really seeing not the tip of the iceberg, but only half of the iceberg you know. So that’s the general answer.

**Andrew Schorr:**
Right. Boy, every time I listen to you talk sir I hear the love and caring and devotion you have, and I know you have a whole team that works with you, and when you say TCC just to remind people that’s the Terence Cardinal Cooke Health Care Center there on Fifth Avenue in New York City, just a few blocks from where I grew up. I love hearing Dr. Lechich’s accent. I’m 3,000 miles away, but it makes me feel like I’m home sir. Thank you. Well I want to thank you for being with us.

**Dr. Lechich:**
Andrew I want to say just one quick thing. There was a question that had been posed earlier about educating doctors about HD. We are doing an education of doctors, and we are also doing this for Alzheimer’s and Parkinson’s as well. It’s a day in the life of a Huntington’s patient where medical students in their third year come through here, spend a whole day with us, lectures, most importantly one-on-one with our families and residents. It is a compelling experience. Every student that has been through it for the last five years has loved it. I just put it out there for anyone listening who is academically oriented. It’s great idea. I’d love to help anybody else with it if they need the help.

**Andrew Schorr:**
Thank you. We have to educate families, and we educate providers as well. Dr. Anthony Lechich, Medical Director and Senior Vice President for Medical Affairs at the Terence Cardinal Cooke Health Care Center in New York City, thank you sir.

Let me just recap for our listeners, remember we do these live but all the replays and the transcripts are there for you, and we keep adding the transcripts as quickly as we can type them on [www.patientpower.info](http://www.patientpower.info). So take a look at that. The replay is there from yesterday; we’ll get the one up hopefully later today. Thank you so much for listening in. I’m there with you as you deal with this if your family is affected with Huntington’s disease. Thanks so much to the Huntington’s Disease Society of America, website [www.hdsa.org](http://www.hdsa.org) if you are listening live. See if you can to Pittsburgh and be part of the community.