### **Sherri and Jessica**

## Mother of Rachel who lives with Canavan Disease, age 16 and Jessica, age 17





#### **SIBLINGS**

Being their mom is a joyful thing.

https://youtu.be/vxYumSuJGgk

**Sherri:** The greatest joy of having my girls is their -- I think joy is too simple of word -- I'm in awe of both my kids that they've taught me so many things I never would have been envisioned life could. I never thought my legacy would be being a good mom, but they make it easy to do. With Rachel even though she can't talk and walk and even though she has 24 hour a day needs of care, the things she has taught me and the joy she has brought not only to her sister and me but to so many other people is amazing. In the sense that you know when you first look at her they say, "What beautiful red hair she has."

And when she was younger she would laugh and smile all the time, and that always brightened people's day. I feel she has mastered what we all strive for in life and that is to be loved and to enjoy life to laugh and smile. And when she was younger she did that all the time. Now that she is 16, Jessica reminds me that she is a teenager and perhaps that's why she is not so smiley and happy at home, because most teenagers aren't, but when she is at school I understand that she is laughing all day long and she has a great time at school.

So the pride I have when I hear that she enjoys school and that she brightens up all the other classmates up when she is there. And if she can't make it to school for a day, she is truly missed, you know that makes me proud. And with Jessica, you know, she is just an extraordinary kid, she has gone through so much -- she has learned about life that no child should ponder upon, and that is, she knew about Rachel's disability from the get go. They were only, 18 months apart. And so having Rachel's need to care for so much of her time is all that Jessica knows.

She has the attitude of "it is what it is." But she has learned to take the best of it and go and run with it. She is a senior in high school and she wants to be a neuroscientist. She wants to understand the brain and to help cure diseases, not only Canavan diseases, but other neurological diseases so that other families don't have to go through what we've gone through. And the ambition that she has -- you want me to talk about you, we're not done talking about you? It's -- I'm talking about Jessica now. Okay. I'll get back to you in a sec, okay. So, you know, she -- and Jessica always have such compassion, for so many situations. She is so mature compared to kids her age I'm just in awe of her, she is truly an

amazing kid and the joy she brings, all the things that she says and does and desires, you know a mother couldn't be more proud of her. So the joy I mean is -- yeah, being a mom is joyful thing, being their mom is a joyful thing.

## If the child is asking the question, it's because they need to know. https://youtu.be/Gz5LFwMK9RM

**Sherri** - So, we always made sure that Jessica was fully aware it was always open and honest. We answered Jessica's questions as she asked them and we kept it as simple and as age appropriate we could be, but we were counseled to just always speak the truth, and if the child is asking the question, it's because they need to know. So Jessica always has been fascinated with science and how things work and when she was 4 years old she asked why does Rachel have Canavan disease and I don't if it's inherited? And so I tried to simplify the answer, but that wasn't good enough for her.

And so we explained about how the father and mother -- each have two genes and we give one of our genes to the baby and one of our genes has a mutation that causes Canavan disease and the other gene that we have does not have that mutation, so it's a typical gene. If daddy and I gave you each of our genes that do not have the mutation, then you don't have the disease and you're not a carrier. If I gave you a mutated gene, but daddy gave you one of his that's not mutated, you are a carrier and vice versa.

If -- and in Rachel's case I gave Rachel one of my mutated genes and her father gave Rachel one of his mutated gene that's why she has Canavan disease, and she says, "Oh I get it," and she got a pencil and paper or a crayon and paper and drew out a male figure -- a stick figure and a female stick figure and drew lines and wrote two genes under each individual and had one indicate that it was mutated gene and one not -- and then four children underneath them and was able to draw -- and she totally put what all the books say -- how it looks like in a picture form, and that's, you know, one of confirmations that I knew that she was a special kid, that she just, you know, she always asks until she gets.

And so I knew a simple answer wouldn't work. And she understood that she is not going to have that disease and that she could be a carrier, or may not be, but she knows she doesn't have the disease.

#### She told me how important it is to know the sad stuff too.

https://youtu.be/L8OzcbLcTxc

**Sherri:** So, with having such a rare disease; it's important to be in touch with other families going through this. And thankfully through social media we're able to get in touch with families and we speak daily about the ins and outs of what life is like and the things we're dealing with and you know -- fortunately some funny things that happened we post just to

make light of it. And sadly, this past week we had lost one of the children with Canavan disease, and it was a shock because it was just -- he died in his sleep and he was healthy and happy, and the mom just posted a picture of him and her on the website just the day before and -- of him being happy and smiling. And so it was a big shock and I felt the need to tell Jessica. I tried to hide sad stuff like that from her in the past and she had spoken to me about how important it was to know the sad stuff too, because it is reality and she doesn't want to find out, you know, a year later -- 6 months later that someone who she is thinking of had passed already and wasn't able to think at that time of the person. So, she felt it was important to know also the real world and what's going on. So, I had to tell her that Vinnie had passed and Vinnie was a special -- more special than -- to us, even though all the children are so special, because he was a redhead also. And so we always said that they could be brother and sister because they had so many similarities. We talked about how it was just a fluke thing that happened and that he just died out of the blue. We talk about it, we talk about how scary it is that we don't know what tomorrow will be like and we know that as long as we're doing the best we can and making sure that we're happy then you know that's what life is for, is to be happy and respectful and love and we're doing all the things that we're supposed to do. And so if, god forbid, tomorrow doesn't come for Rachel, we can say that she had a good life. We of course don't want that to happen. I'm planning on retiring with her and she we're going to share the same nursing home and go from there, but we can't plan any other way, but go from there.

#### She wanted to have a normal typical world

https://youtu.be/JKm1CUv8Kl4

**Sherri:** So Jessica's first day of kindergarten, we're all excited and we brought Jessica in and there were tables in the classroom with name tags -- name plates. And so we found Jessica's nameplate. We were one of the first people to arrive, so not everyone was there, and Jessica saw who she was sitting next to, a girl I never knew, but apparently Jessica knew from preschool or whatever and she said, "I don't want to sit next to her." And that totally threw me, because Jessica has always been friendly and welcoming and I could not believe I heard this, that she did not want to sit next to someone. So I said, "Well, maybe you should talk to teacher and see what the teacher says." So the teacher said, "Fine, no problem," and she switched her. And then when she came home from school I said, "Why is it that you didn't want to sit next to this person?" And she said, "Because she has a funny arm and she walks funny." And I was shocked, because here at home she has such a disabled sister. I would think that she would be the most welcoming of someone who has a disability, and instead she didn't want anything to with her, she did not want to sit next to her. And I was heartbroken to hear that.

And so I had spoken with her teacher later on and I had spoken also with a child physiologist, only in passing I had mentioned it. And they both explained to me that at home Rachel is part of Jessica's world where being disabled is a constant, and that at school its Jessica's world, and that she wants to have a normal, typical world where there is no disability and no medical issues. So she was taking control of the situation and making it happen because she knew this

girl had a disability and she didn't want to be part of that. Actually as heartbroken as I was that she felt that way and didn't want to sit next to this girl, it also kind of made me proud in the fact that she was able to acknowledge it and figure out how to remedy it. And so it kind of reassured me that she is going to be okay. A few years later she ended up being very good friends with this girl. And she actually said to me, you know she is not so bad, she says, I don't know what my problem was when I was in kindergarten. So she has learned to look past disabilities growing up, but at that time in kindergarten that was an eye opening experience to realize.

**Sherri:** So one day we were picking Jessica up from school and I had Rachel with me. So I pulled into the handicap spot and wheeled her out in the wheelchair to go and to get Jessica. And as I'm wheeling into the school the kids were already assembled in the lobby area, and one of the kid says to Jessica, is -- eww, is that your sister, and Jessica stood up proud as a peacock and said, "Yes it is," and bumped me aside and took Rachel's wheelchair and started wheeling her around. And you know it's another proud mom moment that she -- she wasn't going to let this kid intimidate her or make her feel sorry, and she was in fact going to show that she was proud of her sister and it's not so bad.

## We postponed Make-a-Wish and Disney World until Jessica was old enough to remember it

https://youtu.be/YQZ5WwOFZDI

Sherri: When we first learned about the diagnosis, it was confirmed when Rachel was 8 months old, which meant Jessica was just a little over 2. And we were told she wouldn't live past the age of 4. And so my first thinking was, how do we tell Jessica, how do we handle having this 2 year old learn of her sister dying? And when you first get the diagnosis all you hear is your child is going to die, and that's -- your every thinking is, when is she going to die? And you go through mourning the life that you thought you would have with this child, because obviously it's not going to be how you planned. And it's just your every waking movement is she going die, when is she going to die, how old will Jessica be when she dies. We were granted a make-a-wish and Rachel was 2 years old when we got the make-a-wish grant and I thought, that's great, let's go to Disney World, but Jessica would have only been 3 and we thought -- I wanted it to be so that she remembers what it -- this trip is about. And so I asked if we could postpone it until Jessica was 6 years old. And they of course said yes. And during those years I just kept praying that Rachel would live long enough so that we could go to Disney World with Jessica and have Jessica and Rachel with us for this family outing and to remember it. And I just had to have faith that it will happen. And thankfully we were able to go to Disney World. It rained every day, but it was a great memory, and you remember it?

Jessica: Yeah.

**Sherri**: Okay, good.

#### Jessica has asked, "Will I have to take care of Rachel?"

https://youtu.be/ZHcz0EsCwkl

**Sherri**: When we were driving by a nursing home Jessica asked me, "When you and daddy are old and you die, will I have to take care of Rachel?" and I pull car over and I parked and I turned to her and I said, "You'll never *have* to take care of Rachel. If you want to, you're welcome to. But your job is to live your life and we will never make you *have* to take care of Rachel." And she is like, "Okay," and was able to keep going. She never asked me that question again, now she plans -- I don't know if it's still true, but she plans on being a successful scientist and being able to buy a bread and breakfast for Rachel and I to work and live in, and therefore Rachel can stay with us and I can cook breakfast and take care of her. So she is always planning on what -- how to include us in her life, so I don't know if that's a typical teenager plan, but it's my kid's plan. It's still true?

Jessica: Yeah

Sherri: Okay, whew.

#### We often talk about what things would be like if Rachel were typical. https://youtu.be/4sDxqCAKXyA

**Sherri:** There are times, having a teenager, a typical teenager in the house, I often give Rachel a big kiss and say, "Thank you for not talking back to me." At this age I many times say Rachel is my easy child. You know -- having a teenager has its challenges if you're a regular typical family and then ten folds of the emotions that are going on in this family, it's definitely something I ponder. Wondering if -- what life would be like having Rachel, having -- not have Canavan disease -- we had planned on the kids being close in age. And I have a brother who is 20 months older than me, -- their dad has a brother who is 15 months younger than him. And we -- although growing up, we admit that we had our sibling arguments, as adults it was awesome. So I wanted -- I had planned on Jessica having someone close in age to play with. And there are many times that I wish that she had someone to go bike riding with or to play kickball in the yard, do all the things that I did growing up. And being robbed of that is -- makes me mad. I have a bumper sticker on my fridge that says, "I plan, God laughs."

And you know, I've learned to deal with fact that, you know, my plan isn't necessarily the one that's going to happen, but make the best of it. I see Rachel -- I see -- Rachel's personality seems quite similar to my own. And so I think that Rachel would have been -- I was a tomboy growing up, and I think Rachel would have been more like that, whereas Jessica enjoys shopping and the girly stuff that I can't stand. And thinking about it I wonder if there would be a lot of sibling rivalry because -- they would be so different from each other. But knowing Jessica's personality and the compassion she has I would think that if Rachel was like me she also would be easy to get along with, that it wouldn't be so bad. But we often talk about if Rachel was typical-- you know, what things would be like. And then we laugh and giggle and I go change her diaper or do whatever of the special needs things needs to be done.

#### What would Rachel say to Jessica if she could speak...

https://youtu.be/ndqvv8vtlwc

So one of the questions was, what would Rachel say to Jessica if she could speak? I think she would say how proud she is to have Jessica. She really enjoys when -- there are times when Rachel will whine and I can't figure out why and comfort her, I'll always ask Jessica why, and Jessica always comes up with an answer that soothes Rachel. So I think they have a bond with themselves that they can really be there for each other and help each other.

# SPIRITUALITY God gives me the strength to keep carrying on. https://youtu.be/TjpNQpY-cDg

Sherri: So one of the fundraisers we did for gene therapy, raising money for gene therapy, my sorority did a walkathon, and it was one of the first fundraisers that we've done. So we're at the walkathon walking around and my sorority sisters, who I've graduated with, you know of course participated in it. And I was the only Jewish one in that sorority. And so they felt that I was religious because I could answer their questions about Judaism. I wouldn't say I'm that religious, but I at least knew the answers. So walking around during the walkathon one of my sorority sisters said, "So do you still believe in God after this happened?" And at that time -- with Canavan disease the children cry inconsolably for the first year and a half. And at that time you get the diagnosis they're not going to live past the age of four, and you sit there while this screaming child is happening at 3 in the morning for the 20th hour. You say to yourself, "Why do I put up with this? Why not just throw her out the window?" And that's when I have to believe in God, that God gives me the strength not to throw her out of the window. So I don't think God caused the disease. I think God gives me the strength to keep carrying on.

## You may not appreciate it now, but when the whole thing happens, there is a plan.

https://youtu.be/\_OrusDdAtDg

Jessica: So in Judaism, when a girl is 12 or 13 they have a Bat Mitzvah, which means you declare that you will choose Judaism as your religion. At the time I was still really mad at God, why would he afflict -- why would he have my sister-- have this disease which is so painful for her, where she has muscle cramps and can't say what is bothering her, why would she do that to such a beautiful child. So I didn't have my Bat Mitzvah because of that. But a year later when Rachel -- she actually had her Bat Mitzvah and my mom wanted me to be a part of it and I'm dragging my heels on it. So I ended up writing a Dvar Torah, which is a commentary on the Para shah or a chapter from the Bible that week. And that week the chapter was the 10 Commandments and the Promises-that God made to Moses to say to the Israelites, if you do this, you'll get this and if you don't do

this, you'll do that -- this will happen. So I made a commentary on that. I don't believe -- I don't like formed religion per se because I think that it uses fear to manipulate people into doing the right thing. But I do believe that there is like something higher than ourselves, so.

Sherri: It was an amazing Dvar Torah in the sense that she incorporated why bad things happen and is it that Rachel has Canavan disease because I did something bad in my childhood or her father did something bad or even Jessica did something bad, and she was able to tell of a anecdote of this man who had a horse that ran away and the village came to console him. And he said, "I don't know if it's a good thing or a bad thing". And then a few days later the horse came back with a herd of wild horses. And so now this farmer has this herd of horses and the village people came to congratulate him and say, congratulations on such a great thing that happened, and he says, "I don't know if it's a good thing or a bad thing". And then one day his son was riding one of the wild horses and was thrown and broke his leg. And the village people came, once again, to console him that the son has a broken leg. Once again, "I don't know if it's a good thing or a bad thing". And then the government declared that there was a war and they came by to collect all able-bodied men to fight in the war, and because the son had a broken leg, he wasn't carted away. And the farmer said, "Now I know, it was a good thing that my horse ran away." And Jessica went on to explain that although right in front of us it's happening, we don't know whether the ultimate plan is going to be a better outcome for why this happened to Rachel.

And thinking that Jessica is working in the lab of the people who are working to cure Canavan disease, and that never would have happened -- in fact the lab working on Canavan disease may not have happened. When the doctor started at UMass he didn't have any plan of working on Canavan disease, but because it's local, I introduced him to Rachel, and he immediately fell in love with her and said, "I'm going to devote my first project to curing Canavan disease." So if that didn't -- if Rachel wasn't around to prompt him, then that wouldn't have happened. So this isn't a road I would choose to have been on, but the fact that Rachel is here and we're just making the best that we can of the situation and with her Dvar Torah, you know teaching the contrivance about -- you may not appreciate it now, but when the whole picture happens, there is a plan.



#### **DECISION-MAKING AND PALLIATIVE CARE**

It's tough to talk about end-of-life preparedness, but it's part of what we have to go through.

https://youtu.be/3lmQHDJxJvM

**Sherri:** I think that's the scary part, I don't want to ever feel that I could have done something more. And so it's scary that I don't know. When she was first diagnosed we were counseled to have a DNR for her. And reluctantly, I did it and I put it in a drawer and never told anybody about it, and it sat there for 11 years. And then this past doctor's visit her neurologist said - that because her scoliosis is so bad that if they were to do chest compressions, it would cause more harm for her and more suffering and that I should consider dusting off the DNR and taking it out. And I

said, "Okay, I'll think about it." And then at her pediatrician appointment, she also brought it up. So, reluctantly I did dust it off and I have it now it her room, should I need to show someone it. I waited some time before I told Jessica. Did I tell you yet?

Jessica: Yeah.

**Sherri:** I waited some time before I told her, I didn't want to make Jessica sad. And it doesn't mean that I am ready to give up on Rachel, but it means that I am prepared to not allow Rachel to suffer. And, you know -- as Jessica mentioned we have a fabulous relationship where we are open and honest with each other and are able to share our feelings and go with that. So, it's tough to talk about end of life issues and preparedness, but it's part of what we have to go through.

#### It has taken the specialists 11 years to get through to me.

https://youtu.be/FCDFJcukCIU

**Sherri:** We've also have been told about intubation, that because her muscles are so weak that if we were to intubate her we may not be able to take her off intubation once we do it, and that would perhaps prolong her life, but not the quality of life. And her father and I have always felt very strongly that we may not have quantity of life, but as long as she has quality of life, that's the most important thing. And so I'm still not sure what I'll do if that time arrives. But, having some sort of discussion and some kind of plan to choose from, I think, is very important to help during that time.

Interviewer: And who is helping you to have those conversations or think about those things?

**Sherri:** Her pediatrician, her neurologist, our home nurses. You know they each try in their own way to get through. Like I said it has taken 11 years for them to actually get through to me, for me to acknowledge that, okay, I'll now listen and do. But so, you know, I think having them realize it's not falling on deaf ears, but it's not -- you know, it doesn't mean I'm going to do it. So, you know, mention it, I'll keep it in the back of my mind. And when I'm ready I'll do what needs to be done.