An Orange Socks Story: Kierra- 22Q and Tracheomalacia

Interview by: Gerald Nebeker, President of Orange Socks

Gerald: I was privileged to have an Orange Socks interview with Kierra by phone about her daughter Evanna, who has 22Q, also known as 22 deletion 11 or DiGeorge syndrome. Kierra is very active in reaching out to others through her blog, the irvinefamilyblog.com, and her Instagram page, "@evannasspecialheart." I was uplifted by her story, and I'm sure you will be as well.

Gerald: Kierra, thank you so much for taking the time to speak with me. When did you find out that your daughter, Evanna, had 22Q?

Kierra: Everything kind of first started when we went for that 20-week detailed ultrasound. Most people know it is when the ultrasound waves find out the gender, whether you're going to have a boy or girl, but it's actually technically called a detailed ultrasound, where they go over all the body systems. During that ultrasound, they noticed quite a major heart defect in the beginning. Then, when we came back the following week, they gave us one diagnosis and casually mentioned the DiGeorge syndrome or 22 Deletion 11. As things got further into the pregnancy, we had another major echocardiogram at a different hospital, when they gave us an even more specific heart diagnosis. With this particular heart defect, there is a really high risk of also having 22Q. So technically, we kind of knew about it when she was born, but we sort of just chose to not do an amniocentesis as it was not going to change our decision on proceeding with the pregnancy. I didn't want to add more risk to the situation, so we just opted to find out after she was born. We saw that 22Q does have some light physical features, sometimes lower set ears, a small mouth, a broader nose, and longer fingers and longer toes. Evanna's were hard to see; her facial features are very subtle, so we've been told. We still noticed them, so we sent away for genetic testing right after she was born. Two months later, those results came back, and they were positive, but we weren't surprised at all. If anything, we would have actually been more surprised if the results had come back negative.

Gerald: Interesting. What have been some of the challenges? She is three years old now, correct?

Kierra: Yes.

Gerald: Okay, and so what have been some of the challenges that you've faced with her over the last three years?

Kierra: There actually have been a lot, and definitely a lot of the challenges or complications are because of 22Q. She was born with a major heart defect, and required open-heart surgery. Fortunately, we endured the very bumpy recovery. She developed another diagnosis called tracheomalacia, which is something associated with DiGeorge syndrome. It's one of those

things that some 22Q kids have, and it means a floppy trachea, so after surgery, we could not extubate her off the hospital ventilator. We tried for five months and did different surgeries, and after five months in the ICU, we finally proceeded with the tracheostomy so she could be ventilated better. Again because of additional complications and just one thing after another, we were in the hospital for a total of 400 days, and 322 of those days, almost 11 months, were exclusively in ICU. We had quite the long experience, going in for open-heart surgery and expecting to only be in for a month or two, and having that turned into this whole 13-month admission. And back and forth between two different children's hospitals, and just complication after complication. Definitely the 22Q had a role in that rougher recovery, and not bouncing back as quick enough.

Gerald: What a challenge.

Kierra: Yeah, it was quite the year.

Gerald: How is she now?

Kierra: She's doing okay. We were discharged after 400 days on November 9, 2015. It was quite the going home, our whole medical team was nervous. They definitely said when we were discharged, that she was one of the most complicated cases coming through their door, and for us to go home because not only does she have this very, very complicated heart condition, maybe she also had this critical airway, and those are two major double whammies. Kids like her often bounce back, they said. They often wind up being readmitted. Thankfully, while we've had a curve ball here and there, we were discharged at the end of 2015. We did have a couple of readmissions last year, but only two of them were unplanned. We had a few planned, like for a G-tube surgery and for her bronchoscopy, but we only had two unplanned admissions, and they both went relatively quickly. We were only in for three weeks, which is a drop in the bucket for us. In 2017 so far, things are going really well. She has been progressing and stabilizing, and we're actually just starting to wean her off the ventilator. We have just started doing sprints a little while ago. We'll go off the vent and work towards decannulation, towards the process of taking the trach out.

Gerald: That's great. So, tell me, what have been the joys in having her?

Kierra: We just love her to pieces. She has been both our greatest blessing, and our greatest trial. We've gained so much knowledge from her. Throughout this journey, the strength my husband and I have gathered has helped our relationship. I'm sure things like this can easily break a family apart, and they talk about divorce rates going up. If anything, I feel like this has brought us closer together. My husband and I worked to become a really good team, and we coordinate. He has his strength, and I have my strength, and we work together and gel really well. It has just been interesting and lovely all at the same time.

Gerald: That's super. What has been her impact on your extended family, friends and neighbors?

Kierra: One of the things that has not been normal is not having relationships built. Unfortunately, a lot of her life has been in a hospital, so relationships between grandparents or aunts and uncles have not been able to be built. Even now that she's home, she just can't go to grandma and grandpa's for the afternoon. That's just not a possibility right now. She requires being looked after by someone 24/7. Either my husband or me, because we've been properly trained on how to care for her, or by a nurse. My parents, and particularly my mom, talks about how she has a really awesome relationship with my nieces and nephews, but she doesn't have that same relationship with my daughter. Unfortunately, that's because of the medical complications. I'd say that's the biggest impact. Thankfully, it hasn't really impacted relationships for my husband and me. For the most part, most of our family and friends have stuck around and have been really great support for us, and have been there for us. I know a lot of times this can end a lot of relationships with people, but thankfully it hasn't made that impact, it has been more positive.

Gerald: That's interesting. I'm curious, with having a child who has had so many complications, has that had any impact on potentially your plans to have future children?

Kierra: It did for a little while. Originally, I was hoping to have one kid after the other with only one year in between, but having Evanna in the hospital definitely delayed our plan. My husband was very adamant that we would wait until at least she got home and was somewhat stable and in routine before we would talk about more kids. Last year, we came home and once things started to settle down, we decided to start trying. We did get pregnant last year, but I ended up having a miscarriage at 12 weeks. We were upset about it, but not too upset, because we knew about the pregnancy not being viable pretty early at six weeks. Unfortunately, when you go through a situation like we did with our daughter, your perspective changes greatly, and a miscarriage isn't as devastating. We took the summer off, and then we started trying this fall, and it took a little while with this one, but we are pregnant right now and due later this year at the end of December.

Gerald: Congratulations.

Kierra: With baby number two, yeah, we are really excited.

Gerald: That's awesome. If I were to come to you, just having received the diagnosis that I had a daughter who had 22 Deletion 11, what advice would you give me?

Kierra: I belong to various support groups, and there are people who join the group and say, "I just got this diagnosis, what advice can you give me?" Usually, the advice I give is take it one day at a time. It's one of those diagnoses where you just have to wait and see. There are common things that are associated with 22Q, like heart defects and/or cleft palates, but until the child is born, most of the time you really aren't going to know what issues the child will have. As one presents itself, another one might present itself later in life, so you have to take it one day at a time and take it one issue at a time, I guess.

Gerald: Oh, good advice. With everything that you've gone through, with all the medical issues, is it worth it?

Kierra: Oh, absolutely. There's not one day that we regret proceeding with this pregnancy whatsoever. We love her so much, and we just can't imagine our lives without her. Of course, it's a little more complicated, but everybody has their complications or difficulties, and right now, that's her and that's okay. We love her. Like I said, she has been our greatest blessing. She has just taught us so much, and if anything, we are so proud of her. She has this very calm nature about her. When we are admitted, we are always told how content and happy she is, and for a toddler, she is so well behaved when getting an assessment. At first, we had lots of appointments and lots of follow-ups and assessments on a monthly basis, and she is just a well-behaved girl who takes everything in stride. I am so proud of her, that I just beam. We still currently have echocardiograms every three months, and I beam that the echo techs fight over her because they love doing their exam on her. She is interested in anatomy and is so well-behaved as a toddler. I'm that proud mommy because she is a well-behaved toddler who the echo techs fight over.

Gerald: Oh, that's great. I think it's wonderful that you are expecting again and you have such a blessing in your life with Evanna. I think that's awesome. What are some future things that you're looking for in relation to Evanna?

Kierra: Well, she's got some stuff going on right now with her heart. She's currently considered terminal. She was born with a very severe heart defect. People always ask if she has a hole in her heart, and I explain that it's very complicated because of her anatomy. Because of how complicated and intertwined the condition is, she is not eligible for a transplant of any sort. It's just not possible anatomically, so unfortunately, some options are no longer available to us. We are in the middle of a pretty big referral right now. We're from Canada, but there is a surgeon in California whose name is Frank Hanley, and he happens to specialize in our daughter's particular diagnosis. He pioneered unifocalization surgery and is an expert in that field. We are having a cast done for his view, and if he feels like there's anything that he can do and accepts our case, then Canada will fly our daughter to him for another open-heart surgery. However, this surgery is not going to be the almighty, miracle, life-saving surgery. At the end of the day, Evanna is still considered terminal. Our goal with him is to improve her current prognosis and improve her overall quality of life. That's what we're hoping to gain if he accepts our case. We've been highly warned and prepared for the potential that he'll say no. He doesn't like saying no, he likes a challenge, but sometimes there are cases where there's nothing that even he can do. We are well-warned that Evanna is on that cut, on the very severe end. That's where we are now with things, which is kind of interesting. We'll hopefully have an answer around July because she's stable, and Doctor Hanley usually books around two months in advance. That puts us around October or November when I'll be eight months pregnant. If she's stable and if he can't do her surgery in a timely manner, then we might have to push it off until the new year after Christmas. That's where we are right now with things, so even though things are stable,

and even though lots of things have improved, there are still a lot of underlying things we are dealing with.

Gerald: I'll keep my fingers crossed for you.

Kierra: Thank you.

Gerald: Anything else that you'd like to say, any concluding remarks?

Kierra: I have a little saying. One thing I've learned and one thing I try to emphasize out there is that we have good years and bad years, and life is about surviving the bad years while learning and growing through them, and then enjoying the good years. That's what we're doing. 2015 was not a good year for us at all; 2016 was a little better, but we still had curve balls. So far, this year has been an awesome year. We've had a lot of good things happen, so we are just trying to enjoy it while at the same time waiting for the other shoe to drop. People tell me that you don't want to live life waiting for the other shoe to drop, but unfortunately that's just not something you can do when you have a special needs child. It's always a reality, especially to new parents. A lot of us choose to prepare for the moments when life decides to get a little chaotic.

Gerald: Yeah, super. Well, thank you again. Awesome.

Kierra: No problem.