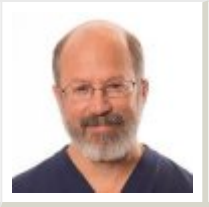
Twin to Twin Transfusion Syndrome: Algorithm for Diagnosing and Treating

Dr. Emanuel "Mike" Vlastos discusses the algorithm for diagnosing and trading twin to twin transfusion syndrome.



Featured Speaker:
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Learn more about Emanuel "Mike" Vlastos, MD, FACOG, FAAFP (https://www.childrensmercy.org/profiles/emanuel-j-vlastos/)

Transcription:

Melanie Cole (Host): Welcome to Transformational Pediatrics with Children's Mercy Kansas City. I'm Melanie Cole and today, we're discussing the algorithm for diagnosing and treating twin to twin transfusion syndrome. Joining me, is Dr. Emanuel "Mike" Vlastos. He's the Medical Director of Fetal Therapy at the Elizabeth J. Ferrell Fetal Health Center at Children's Mercy Kansas City. Dr. Vlastos, it's a pleasure to have you join us today. Let's start with a little definition. What is twin to twin transfusion syndrome? How often does it complicate monochorionic twins and tell us a little bit about its natural history.

Emanuel "Mike" Vlastos, MD, FACOG, FAAFP (Guest): So, twin to twin transfusion syndrome or more easily said TTTS happens in about ten to fifteen percent of monochorionic pregnancies. So, that's about five percent of pregnancies would fall into the twin category. The natural history of this is due to the twins having a shared placenta; they actually have connected blood vessels on the surface or within the placenta itself. So, each twin being identical, can pass blood from themselves to the co-twin and vice versa. Eight five to ninety percent of the time, there's a balance. However, ten to fifteen percent of the time, there's an imbalance where one twin is giving more blood than it receives in return. Given that story, one baby becomes low on blood and anemic. The other twin becomes very full of blood and plethoric. And the whole process of twin to twin gets started at that juncture.

The natural history is it moves from one to five stages. One being we have big fluid and little fluid and unfortunately, the fifth and last stages you can loose one or both babies.

Host: This is such an interesting situation Dr. Vlastos. Tell us a little bit about what we know about the underlying pathophysiology and do we know even what causes it?

Dr. Vlastos: At least the theories go that when a single egg and sperm get together and create a conception, that then tumbles down the fallopian tube and embeds into the lining of the uterus; and at some unknown timing, that particular pregnancy splits to create two identical pregnancies. At that same time, as they differentiate and begin to grow individually; they share a placenta. As the embryo becomes a fetus, as an umbilical cord forms and has unique arborizing vessels into the placenta like a tree root ball; they mate. The blood vessels as they are growing, don't realize they belong to individual

twins so they can literally fuse. Again, this may be balanced and there's not a problem throughout the pregnancy. However, if there's an imbalance with blood flow from one twin to the other; that's the incipience of the twin to twin transfusion syndrome.

Host: So, how and when is it diagnosed? How would you even know Dr. Vlastos and how is the diagnosis of this syndrome made? Please tell us a little bit about diagnosis and also about Quintero staging. How is it used for babies?

Dr. Vlastos: The only way we would know this is because of the modern technology of ultrasound. Primarily, it helps us know that there are twins, secondarily, that there is a single placenta and through several different techniques, the placenta can be said to be either shared or different between the twins. Of course, discovering the gender of the fetuses is helpful. Because if you have got a guy and a gal, they're fraternal twins. But same sex pregnancies, the next important differentiation is to find out if they are a one or two plus intations [00:03:59]. If they then share a placenta, the necessity of fetal surveillance steps in. That usually begins at 14 to 15 weeks of pregnancy. The twins are evaluated every two weeks. And this is to assure that twin to twin transfusion syndrome is not developing.

If that should happen, Dr. Quintero back in the late 90s created a staging for twin to twin transfusion syndrome. The first stage is big fluid and little fluid predicated by measuring the deepest pocket of fluid for each twin. The second stage is found when the small twin typically does not have a visible bladder, or the bladder doesn't cycle over an hour's period of time. That baby is very low on fluid and blood, so it tries to hold on to all of its fluids and does not create much urine.

The third stage finds that the blood moving through the cord, the baby's liver or the baby's brains are not flowing in a normal pattern. And that is very worrisome because it takes us sometimes to the fourth stage which literally is heart failure for one of the twins. Typically it's the larger twin whose heart is over working because of all the extra blood in that twin. And the baby can literally go into heart failure. And the fifth stage which nobody really likes to focus upon is the death of one or both twins. So, we wish to have early diagnosis of twin to twin to allow us to hopefully make things better.

Host: Dr. Vlastos, as you've told us about diagnosis and staging; is there an algorithm for screening and monitoring TTTS and tell us about the role of fetal echocardiography for the risk of cardiac anomalies. Tell us about this algorithm.

Dr. Vlastos: Yeah, the algorithm really was put forward by a combination of experts from American College of Obstetricians and Gynecologists and then the Society of Maternal Fetal Medicine. That algorithm has helped general practitioners of obstetrics whether that's family docs who have privileges to do obstetrics, whether that's OBGYNs in their office or maternal fetal medicine physicians. It's to start to see the twins starting at about 14 weeks to 15 weeks. Every fortnight or every two weeks, the second evaluation is done and then the third and the forth et cetera. Typically around 18 to 20 weeks, an echocardiographic evaluation is performed meaning a specific ultrasound focusing on the fetal hearts. This it to make sure there's normal structure of the hearts and there is normal function of the hearts. Continued interrogation goes to specific blood vessels within the twins, whether that's the ductus venosus, the middle cerebral artery or the umbilical artery and vein.

In that way, if there is abnormality of blood flow through these important vessels; this gives credence to

the third stage of twin to twin which by happenstance, is the most common presentation for this disorder. That algorithmic following every two weeks is until delivery with the majority of deliveries happening between 34 and 36 weeks without twin to twin transfusion syndrome or as early as 24 weeks should it be severe and not amenable to therapy.

Host: Well then let's talk about therapy. What is the first line treatment? How's it managed before birth and can it be treated, really before birth? Tell us a little bit about the management recommendations according to stage.

Dr. Vlastos: I appreciate that and indeed thank goodness, there are some therapeutic modalities today. The most common modality of treatment up until the modern times in the 90s, was actually amnioreduction. So, whether this was stage one through stage four, a needle was placed into the amniotic sac of the larger twin and that fluid was reduced down to a normal level. This was a temporizing measure and often had to be repeated. Although this was not a definitive therapy, it could buy time let's say to get a woman to a referral center to have definitive therapy.

In the late 1980s, a physician by the name of De Lia, was the first to put a laparoscope into a pregnant uterus specifically for twin to twin transfusion syndrome. Dr. De Lia's ah ha moment allowed for visualization of the placental surface. Finding the connected blood vessels between twin A and twin B and using laser to photocoagulate those vessels or stop the blood flow through those connections. With that, Dr. De Lia changed the mortality rate from 90 to 100% to 30 to 40%. Subsequent to his time, we have improved on his techniques using much smaller scopes that we put inside the uterus, approximately three millimeters. So, this intervention is done typically around 20 to 21 weeks on average. The average time of delivery then is between 33 and 34 weeks after laser therapy, now the standard of care for twin to twin transfusion syndrome.

Host: Then what about after birth? Is there treatment recommended?

Dr. Vlastos: Yes. So, typically, the treatment is prenatal, during pregnancy. Postnatally, these babies need to be followed. In particular the baby who is the recipient, the larger twin with the extra blood to make sure cardiac function is normal. On top of that, studies both here in the states as well as coming from Europe, have shown that between five to ten percent of all twins who have had twin to twin transfusion syndrome may develop delays in neurodevelopmental progression. The translation here is a neurologic examination certainly as a newborn but these patients formerly fetuses, now newborns, and then infants and then children really need to have annual examinations by their pediatricians to make sure they are hitting their milestones appropriately.

Host: Tell us about your outcomes Dr. Vlastos. How many TTTS surgeries have you done and what have been your outcomes? What have they looked like?

Dr. Vlastos: So, the experience here at Children's Mercy Hospital Kansas City has been about 70 cases over the last three years with the program beginning in the fall of 2017. At this juncture, we have a twin survival meaning both babies surviving twin to twin transfusion of approximately 70%. And we're hoping to improve upon that. That's certainly not 100%. So we actually have 15 to 20% of pregnancies where there is only a single twin that survives, and we have less than 5% of cases where there are no survivors for twin to twin transfusion syndrome. This is mirroring the national average and just a titch

above that.

Host: So, then tell us what you've seen as far as the major benefits of this algorithm since the Fetal Health Center has been using it and also while you're telling us that, how do parents use it?

Dr. Vlastos: So, going first with the parental side, certainly for moms and dads who know that they have identical twins, the algorithm can be viewed, and your gestational age can be viewed upon that algorithm to say well what kind of surveillance should be happening here. That is a template to know the kind of care that should be coming to you and your twins during pregnancy. The benefits of the algorithm at the Fetal Health Center was really to keep us on task and in particular, for our referring physicians. So, we have been fortunate to have referrals from seven different states surrounding the Missouri, Kansas area. So, when these moms have been through evaluation, whether therapy was needed or not and they return to their referring providers; the algorithm goes with them to make sure that those babies continue to have surveillance in utero. Should the findings be that there's the development of twin to twin or upstaging to stage two or greater; then they would need to return to the Fetal Health Center for definitive therapy.

Host: As we wrap up Dr. Vlastos, how does the Fetal Health Center team work with primary care providers involved in the mother's care and tell us about the focus that engaging a multidisciplinary approach looks like for mothers and babies with TTTS.

Dr. Vlastos: At the Fetal Health Center, we're very focused on the family care even during pregnancy. So, families that are referred will first get a phone call from our nurse coordinators. These nurses have all been on labor and delivery or within the NICU so they can speak from personal experience on care of women and newborns. With that an appointment is made. They have an outline for their day which will entail of course, an ultrasound evaluation, meeting with the maternal fetal medicine specialist. Depending upon what is found or known of the baby's cardiac status, they will meet with a pediatric cardiologist and finally, sit down to an integrative consultation to speak with these specialists face to face, discuss kind of the situation for their twins during pregnancy and the expectations of therapy and follow up from that point. That integrative care really allows for the broad picture of the pregnancy and what expectations surround care during the pregnancy, birth and the newborn period.

Host: Do you have any final thoughts for other providers on when you feel it's important that they refer and what you'd like them to know about the Fetal Health Center at Children's Mercy Kansas City?

Dr. Vlastos: Well the Fetal Health Center has evolved beginning in 2017 from a special delivery unit to allowing for fetal intervention from as early as 12 weeks to as late as 39 weeks of gestation for all types of fetal pathologies. In particular, for twin to twin transfusion syndrome, if there is any inkling that this might be at play; we are more than happy to evaluate your patient and say that there is indeed twin to twin or there's a similar entity at play or happily, that none of that is happening at this time and have the patient back to you for continued care by yourself and your community. My desire would be to see people early in the process. The twin to twin transfusion syndrome does not follow any of our logic which would be stage one goes to stage two then goes to stage three. The darndest order can jump from stage one to stage four and the fifth stage and there is fetal loss. So, if in doubt, please give us a call. We're open 24 hours a day, seven days a week. We're happy to speak with you and of course, happy to see and evaluate your patients.

Host: Thank you so much Dr. Vlastos. What a fascinating segment. Thank you for sharing your expertise with us today. This has been Transformational Pediatrics with Children's Mercy Kansas City. To refer your patient, or for more information please visit **www.childrensmercy.org/fetalhealthcenter (http://www.childrensmercy.org/fetalhealthcenter)** to get connected with one of our providers. Please also remember to subscribe, rate and review this podcast and all the other Children's Mercy podcasts. I'm Melanie Cole.