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Twin Anemia Polycythemia Sequence in a Monochorionic Twin Gestation: A Case Report

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Abstract

Twin-twin transfusion syndrome is a rare condition impacting monochorionic pregnancies, in which the placenta is shared by two fetuses. An even more rare condition, as a subtype of twin-twin transfusion syndrome, is twin anemia polycythemia sequence. Both conditions occur on the basis of arteriovenous anastomoses that form in the vessels radiating from the placenta and, therefore, provide opportunity for uneven placental blood flow to both fetuses. This uneven distribution of blood flow can cause different findings in the twins. Whereas the hallmark diagnostic finding of twin-twin transfusion syndrome is evidence of polyhydramnios in one fetus and oligohydramnios in the other fetus, in twin anemia polycythemia sequence the hallmark finding is, as the name suggests, anemia of one fetus and polycythemia of the other fetus. Early recognition of such findings is crucial. We present a case of a monochorionic diamniotic twin gestation in a young patient whose initial ultrasound findings hinted at a possible twin anemia polycythemia sequence.

Introduction

Twin-twin transfusion syndrome (TTTS) is a rare and serious condition that impacts about 10-15% of monochorionic twin gestations [1,2]. In monochorionic twin pregnancies, there is one shared placenta that supplies blood and nutrients to both fetuses. In such a case, it is possible that this shared blood flow is unevenly distributed between the twins, resulting in an increased flow to one twin and a decreased flow to the other twin. This is the basis of what TTTS is. A lesser-known subtype of TTTS, called twin anemia polycythemia sequence (TAPS), is another complication of monochorionic pregnancies and will be of more focus of our case report. Both conditions are progressive in nature, meaning they may worsen as the pregnancy moves forward, but in some cases they may be initially mild in severity and may actually resolve on their own.

Given that there is alteration in the amount of blood flow among the two fetuses in TTTS and TAPS, many complications can arise in either fetus. In both conditions, the twin that is receiving a greater proportion of the placental blood flow is often termed the 'recipient' twin, whereas the fetus that is receiving the lesser proportion of the blood flow is termed the 'donor' twin. We will often use these terms moving forward in our discussion. Therefore, the complications that can arise in either twin often occur in a manner than is quite contrastive.

As with most medical conditions, there are diagnostic criteria for both TTTS and TAPS. For TTTS, there must be a confirmed monochorionic twin gestation and the presence of polyhydramnios for one fetus and oligohydramnios for the other fetus, as assessed by the maximum vertical pocket (MVP) or the amniotic fluid index (AFI) [3,4]. As for TAPS, there also must be a confirmed

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monochorionic twin gestation and evidence of anemia in one fetus and polycythemia in the other fetus, as assessed by middle cerebral artery (MCA) peak systolic velocity using ultrasound doppler. Depending on the MCA measurements gotten, the severity of TAPS is determined by the Leiden staging system [5,6,7]. Unlike TTTS, in TAPS the AFI remains within normal limits.

Management of suspected TTTS or TAPS requires close monitoring of fetal growth and other metrics of fetal wellbeing, often assessed by performing nonstress testing (NST) and biophysical profiles (BPP). These tests are often completed outpatient and they look to assess fetal movement, tone, practice breathing, AFI, fetal heart rate, and other parameters [8,9]. MCA ultrasound doppler may also be utilized and monitored in the case of TAPS, since a rise in peak systolic velocity of the MCA is a sign of worsening of the condition. A fetal echocardiogram may also be warranted to assess for cardiac development, since fetal anemia may lead to increased work by the heart and, in worst case scenarios, lead to fetal heart failure [5,6].

The currently held treatment of TTTS is fetoscopic laser ablation surgery, a minimally invasive in utero surgical procedure [7]. This procedure identifies the site of the recipient twin, identifies the umbilical cord for that twin, and then localizes the smaller vessels coming from that cord on the placenta that anastomose, or connect to, the donor twin, and laser technology is used to coagulate these particular vessels until blood flow is halted [7]. As with most in utero procedures, this surgery possesses the risk of fetal loss and infection, amongst other things.

With regards to the treatment options for TAPS, it depends on the severity of the condition as determined by the Leiden staging system, as was mentioned prior. It is generally held that stage 1 and stage 2 TAPS be continuously monitored, and delivery prior to 37 weeks indicated if TAPS persists beyond 32 weeks [2]. Stage 3 and stage 4 TAPS are treated on a case-by-case basis, but it is generally held that fetoscopic laser ablation can be done at these stages [2, 7]. It is important to note, however, that laser ablation itself may result in TAPS, called post-laser ablation TAPS [4].

Case Presentation

Our case is about a 30-year-old G5P2112 who was about 18 weeks pregnant when she visited our maternal fetal medicine (MFM) clinic. The patient had a past medical history of asthma, and an obstetrical history of stillbirth at 29 weeks and one prior cesarean section delivery. Given the history of stillbirth, she was already taking a low-dose aspirin to help prevent against pre-eclampsia [8]. Additionally, because of the history of stillbirth, antiphospholipid antibody labs were drawn for the patient and were found to be normal. Alpha-fetoprotein (AFP) was also measured as a screening tool for spina bifida, and it was found to be within normal limits.

The patient was referred to our office because she had a confirmed twin pregnancy at her primary obstetrician's office, a type of pregnancy that inherently increases the risk of many adverse events and which, therefore, requires closer monitoring. Transabdominal ultrasound imaging at her first MFM visit with us confirmed the presence of a monochorionic-diamniotic twin pregnancy, as can be seen in Figure *1*. The MVP of fetus A was found to be 3.4 cm, and of fetus B was found to be 3.3 cm, both of which are within the normal limits, which is considered to be between 2-8 cm [6]. These measurements are shown in Figure *2A* (for fetus A) and Figure *2B* (for fetus B).

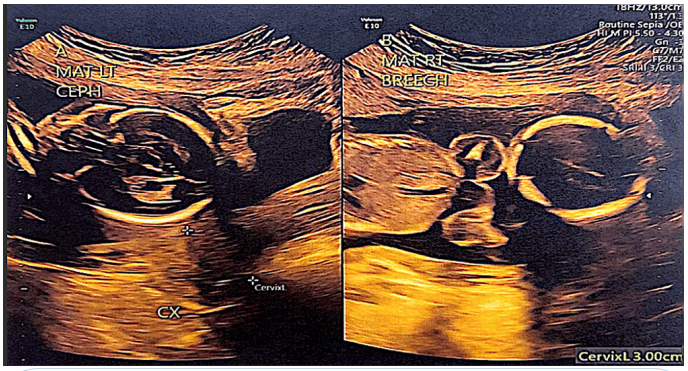

Figure 1: Ultrasonography at 16 Weeks
Showing a diamniotic twin pregnancy, fetus A on the left and fetus B on the right.

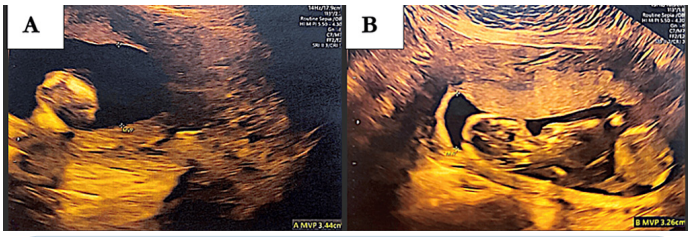

Figure 2: MCA dopplers had demonstrated a peak systolic velocity of 38.48 for fetus A, and 42.29 for fetus B, as shown in Figure *3A* and Figure *3B*. In this setting, it can be said that fetus A is the donor twin, fetus B is the recipient twin. This information is consistent with a possible diagnosis of TAPS. Patient recommendations included continued monitoring via ultrasound every two weeks, repeat fetal echocardiographs, continuation of low dose aspirin, and delivery of her twins at 34-37 weeks if the rest of the pregnancy proved uneventful.

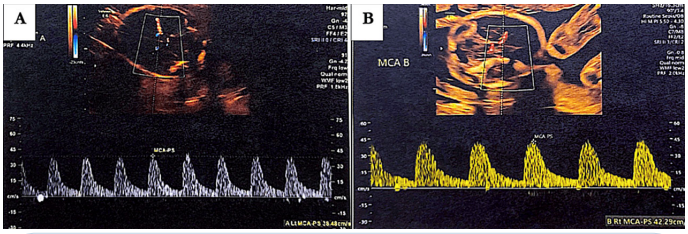

Figure 3: Middle Cerebral Artery Peak Systolic Velocity (MCA-PSV) at 21 Weeks
A: Left MCA measurements showing a peak systolic velocity of 38.48 cm/s for fetus A.
B: Right MCA measurements showing a peak systolic velocity of 42.29 cm/s for fetus B.

Subsequent measurements of the MCA-PSV are shown in Figure *4*. As shown, the MCA-PSV values for the recipient twin (fetus B) were elevated on two occasions, but on a most recent visit they had down trended and become within normal limits. This figure also shows that the MCA-PSV values for the donor twin (fetus A) were always within normal limits. Depending on the MCA measurements gotten, the severity of TAPS is determined by the Leiden staging system, as shown in Table *1* [5,6,7]. Following the red arrows that denote the MCA-PSV of the recipient twin, at the third visit with MFM it seems that the middle cerebral artery velocity of this twin has went below the 1.5 MoM threshold and was now within normal limits. Considering these findings, it was recommended to the patient that she continue on her current medication regimen, with the continued inclusion of a daily low-dose aspirin, as well as that she be scheduled for weekly NST and BPP testing with repeat MCA-PSV dopplers until delivery.

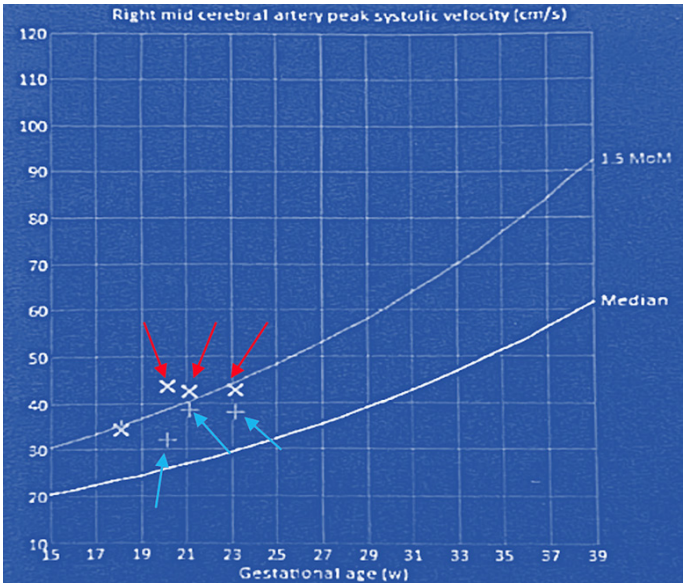

Figure 4: Middle Cerebral Artery Peak Systolic Velocity (MCA-PSV) at Approximately 20, 21, and 23 Weeks Gestation
MoM, multiples of the median
Fetus A: depicted by the blue arrows, all velocity values are within normal limits
Fetus B: depicted by the red arrows, the initial two velocity values are shown to be elevated above 1.5 MoM, whereas the third value is within normal limits

Table 1: Leiden Staging for Twin Anemia Polycythemia Sequence (TAPS)

TAPS Staging	Findings
Stage 1	>0.5 MoM difference in delta MCA-PSV
Stage 2	>0.7 MoM difference in delta MCA-PSV
Stage 3	Stage 1, or 2, but with cardiac compromise of the donor
Stage 4	The donor develops hydrops
Stage 5	One, or both babies die in utero

MoM, multiples of the median; MCA-PSV, middle cerebral artery-peak systolic velocity

Yale Medicine: Twin-to-Twin Transfusion Syndrome [5]

AJOG: Twin-Twin Transfusion Syndrome [6]

TAPS Support: Diagnosis [7]

Discussion

TTTS is a rare and serious condition that impacts about 1 in 10 monochorionic twin pregnancies [2]. In monochorionic twin pregnancies, it is possible that the shared placenta, which supplies blood and nutrients to both fetuses, may display unevenly distributed blood flow, resulting in an increased flow to one twin and a decreased flow to the other twin. This is the basis of TTTS, and it explains the predominant and diagnostic finding that is oligohydramnios of one fetus and polyhydramnios in the other fetus. TAPS is a subtype of TTTS and yet another complication of monochorionic pregnancies wherein the predominant finding, as the name suggests, is anemia of one fetus and polycythemia of the other fetus. These distinctions are depicted in the simple schematic in Figure *5* [4]. These findings also represent the main complications can arise in either fetus in such conditions. The complications that often ensue in pregnancies impacted by these progressive conditions can impact predominantly one or both twins and, in addition to the findings noted above, there may also be findings of fetal growth discordance due to the uneven placental blood flow distribution.

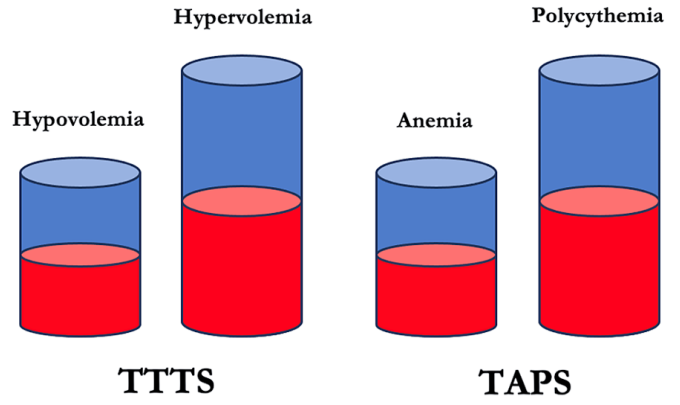

Figure 5: Twin-Twin Transfusion Syndrome (TTTS) vs. Twin Anemia-Polycythemia Sequence (TAPS).
Schematic showing the primary pathology in either condition. In TTTS, one fetus develops hypovolemia (oligohydramnios) whereas the other develops hypervolemia (polyhydramnios). In TAPS, one fetus develops anemia whereas the other develops polycythemia. Hopkins Medicine: TAPS-TTTS [4].

TTTS and TAPS can both be diagnosed prenatally. TTTS, there must be the presence of polyhydramnios for one fetus and oligohydramnios for the other fetus, as assessed by the MVP or the AFI [6]. As for TAPS, there also must be evidence of anemia in one fetus and polycythemia in the other fetus, as assessed by middle cerebral artery (MCA) peak systolic velocity using ultrasound doppler. Depending on the MCA measurements gotten, the severity of TAPS is determined by the Leiden staging system [7]. Unlike TTTS, in TAPS the AFI remains within normal limits.

Even without the presence of full diagnostic findings, if clinical suspicion is high for TTTS or TAPS then closer fetal monitoring should be initiated via a routine weekly NST, BPP, and doppler ultrasound if needed. A fetal echocardiogram may also be warranted to assess for heart development, since fetal anemia may lead to increased work by the heart and, in worst case scenarios, lead to fetal heart failure [10]. After delivery, measurement of fetal hemoglobin and further workup of placental pathology may help to definitively diagnose and gain more insight into the cause of TAPS for a pregnant [7]. The postnatal hemoglobin findings diagnostic of TAPS is shown in Table *2*.

Table 2: Postnatal Anemia Polycythemia (TAPS) Staging
TAPS Support: Diagnosis [7].

TAPS Staging	Findings
Stage 1	The hemoglobin difference between the twins is >8.0 g/dl
Stage 2	The hemoglobin difference between the twins is >11.0 g/dl
Stage 3	The hemoglobin difference between the twins is >14.0 g/dl
Stage 4	The hemoglobin difference between the twins is >17.0 g/dl
Stage 5	The hemoglobin difference between the twins is >20.0 g/dl

The management of TTTS depends on severity, as shown in Figure *6*, but the currently held treatment is that of fetoscopic laser ablation surgery, a minimally invasive in utero surgical procedure [4, 6]. This procedure identifies the site of the recipient twin, identifies the umbilical cord for that twin, and then localizes the smaller vessels coming from that cord that connect to the donor twin, and these particular vessels are coagulated us-

ing the laser until blood flow is stopped [4]. As with most in utero procedures, this surgery possesses the risk of fetal loss, infection, and other adverse outcomes. The treatment option for TAPS depends on the severity of the condition. It is generally held that stage 1 and stage 2 TAPS be continuously monitored, and delivery prior to 37 weeks indicated if TAPS persists beyond 32 weeks [2]. Stage 3 and stage 4 TAPS are treated on a case-by-case basis, but it is generally held that fetoscopic laser ablation can be done at these stages [2, 4].

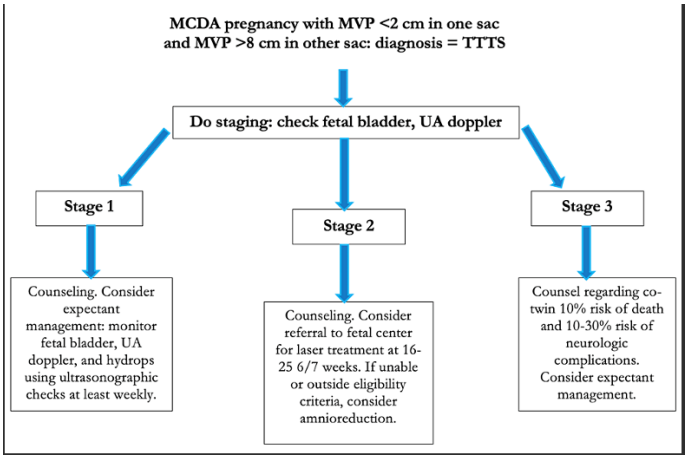

Figure 6: Management Algorithm for Twin-Twin Transfusion Syndrome (TTTS)
MCDA, monochorionic diamniotic; MVP, maximum vertical pocket; TTTS, twin-twin transfusion syndrome; UA, umbilical artery
Hopkins Medicine: TAPS-TTTS [4]
AJOG: Twin-Twin Transfusion Syndrome [6]

Conclusions

In patients with suspected or confirmed TTTS, further work-up in the form of additional ultrasound imaging to monitor fetal growth, as well as MCA ultrasound dopplers to determine if there is any change in the arterial flow between the two fetuses. Early diagnosis of TTTS, or of its subtype called TAPS, is vital in allowing for timely education and counseling of parents prior to delivery, and for preparation of a multidisciplinary approach to management of the babies upon delivery.

References

1. Twin Anemia Polycythemia Sequence. (2023). Accessed: 08/15/2023: https://women.texaschildrens.org/program/texas-childrens-fetal-center/conditions-we-treat/twin-anemia-polycythemia-se....
2. Twin Anemia-Polycythemia Sequence (TAPS). (2023). Accessed: 08/24/2023: https://www.uptodate.com/contents/twin-anemia-polycythemia-sequence-taps.
3. Twin-Twin Transfusion Syndrome (TTTS). (2021). Accessed: 08/28/2023: https://www.cincinnatichildrens.org/health/t/twin-twin-transfusion-syndrome.
4. Conditions We Treat: TAPS-TTTS. (2023). Accessed: 08/30/2023: https://www.hopkinsmedicine.org/gynecology-obstetrics/specialty-areas/fetal-therapy/conditions-we-treat/complications....
5. Twin-to-Twin Transfusion Syndrome. (2023). Accessed: 08/27/2023: https://www.yalemedicine.org/conditions/twin-to-twin-transfusion-syndrome.
6. Twin-Twin Transfusion Syndrome. (2023). Accessed: 08/30/2023: https://www.ajog.org/article/S0002-9378%2812%2901980-1/fulltext.
7. How Can I get a TAPS Diagnosis?. (2023). Accessed: 08/29/2023: https://www.tapssupport.com/diagnosis/.
8. Low-Dose Aspirin Use for the Prevention of Preeclampsia and Related Morbidity and Mortality. (2023). Accessed: 08/29/2023: https://www.acog.org/clinical/clinical-guidance/practice-advisory/articles/2021/12/low-dose-aspirin-use-for-the-preve....
9. Biophysical Profile. (2023). Accessed: 08/28/2023: https://www.chop.edu/conditions-diseases/biophysical-profile-bpp.
10. Fetal Anemia. (2023). Accessed: 08/27/2023: https://www.mercy.net/service/fetal-anemia/.