Case Report

Atypical Findings of Suspect Twin to Twin Transfusion Syndrome Quintero V

Bahar Sangkur Gusasih, Setyorini Irianti, Muhammad Alamsyah Aziz, Andi Kurniadi

Department of Obstetrics and Gynecology
Faculty of Medicine Universitas Padjadjaran
Dr. Hasan Sadikin General Hospital
Bandung

Abstract

Objective: To report atypical findings in a case of suspected twin-to-twin transfusion syndrome Quintero V.

Methods: Case report

Case: A-38-year old woman, G3P2AO presented at 34–35 weeks of gestation with a diagnosis of a diamniotic monochorionic twin pregnancy. The first fetus was in breech position with polyhydramnios and intrauterine fetal demise (IUFD) in the second fetus, raising suspicion for TTTS Quintero stage V. However, this case presented an atypical finding: the single deepest pocket (SDP) of the donor fetus was normal (without oligohydramnios), a rarity in TTTS cases that should be noted. The diagnosis of TTTS Quintero V was based on maternal-fetal ultrasound findings. No additional complications were observed in this case upon further examination.

Conclusions: Several therapies are available for TTTS including amnioreuction, laser ablation of the vascular placental anastomosis, selective feticide, and septostomy. Timing of delivery after management of singleton fetal death in the late second or early third trimester is still debatable. Delivery method is determined based on obstetric indications.

Keywords: atypical, case report, TTTS, Twins.

Correspondence author. Bahar S. Gusasih. Department of Obstetrics and Gynecology.
Faculty of Medicine Universitas Padjadjaran. Dr. Hasan Sadikin General Hospital. Bandung.
Email; bahar21001@mail.unpad.ac.id

INTRODUCTION

A multiple pregnancy is defined as a pregnancy with more than one fetus, a pregnancy with two fetuses is called multiplet.[1] A pregnancy with three fetuses is called a triplet, four are quadruplets and five are quintets.[2] The multiple birth rate has increased from 18.9 to 32 .1 per 1000 live births in the United States from 1980 to 2005. This increase occurred due to fertility therapy and the application of assisted reproductive techniques (ART) as well as the increasing number of women who gave birth over the age of 35 years.[1] In Taiwan, between 1983-1995, only 12% of 34 triplet pregnancies resulted from natural conception, with the remaining 88% were the result of ovulation induction (including in vitro fertilization or IVF). Simirlarly, in Japan, 73.2% of multifetal pregnancies with more than two fetuses result from IVF, 22.1% by ovulation induction, and only 4.3% by spontaneous pregnancy. In Indonesia, as many as 30% of ART produce multifetal pregnancies.[1]

The prevalence of dizygotic (fraternal) twins is higher than that of monozygotic twins, with rates of 70% and 30%, respectively. Population-specific variations exist in the prevalence of dizygotic twins, whereas the global prevalence of monozygotic twins remains relatively constant at 3-5 per 1000 births. Several factors influence the occurrence of multiple births such as race, heredity, parity, age, and maternal BMI. The timing of delivery, position, and presentation of the fetus during delivery, along with its chorionicity and amniocity, need to be taken into account when managing multiple pregnancy

labor.[3,4] However, there is also evidence that multiple pregnancies increases the likelihood of pregnancy complications, particularly in cases of monochorionic multiple pregnancies.[5]

In monochorionic multiple pregnancies, twin-to-twin transfusion syndrome (TTTS) is a serious complication. The diagnosis of TTTS is made using antenatal sonography and is characterized by the presence of oligohydramnios (deepest vertical pocket <2 cm) in the donor with a small or empty bladder and polyhydramnios (single deepest pocket (SDP) >8 cm) with bladder distension in the recipient. [5-7] TTTS affects 10–15% of monochorionic multiple pregnancies and typically manifests between gestation weeks 16 and 26. For optimal perinatal outcomes, TTTS must be identified early and accurately. A notable variation in amniotic fluid (polyhydramnios in recipient babies, oligo or an hydramnios in donor babies) is the primary finding of prenatal ultrasound. Multiple pregnancies of the same sex, monochorionic, weight differences between twins of more than 20%, hydramnios in large fetuses, oligohydramnios and stuck twins in small fetuses, and hemoglobin differences of more than 5 g/dL are the typical findings and diagnostic criteria for TTTS. [1,5-7]

The placentas of the multiple fetuses in a monochorionic pregnancy are shared. The two fetal umbilical circulations are connected by a vascular anastomosis in the chorion in nearly all instances. Deep arteriovenous anastomoses that allow unidirectional blood flow are the foundation of the TTTS mechanism.[5] Following identification, the Quintero staging system typically classifies TTTS. As in this instance, Quintero V TTTS is TTTS with the death of one or both fetuses.[6,8] There are several ways to treat TTTS, ranging from close observation and termination to intervention techniques like selective and non-selective laser fetoscope coagulation.[6,9] Based on the stage of TTTS, a treatment plan can be selected.

However, atypical findings of TTTS are rarely found and should be taken as special notes, asin this case. We will report a rare finding of TTTS in which the SDP level of the donor fetus was normal (not oligohydramnios). Therefore, this paper will also discuss the clinical aspects of multiple pregnancies (gemelli) with suspected TTTS Quintero V, which is characterized by intrauterine fetal death (IUFD) in one of the fetuses. The goal of this study is to examine and assess the diagnostic approach, pathogenesis, pathophysiology, and treatment of TTTS to determine whether they align with theoretical expectations.

CASE

A-38-year-old woman, G3P2AO presented at 34–35 weeks of gestation with a diagnosis of a diamniotic monochorionic twin pregnancy. The first fetus was in breech position with polyhydramnios and intrauterine fetal demise (IUFD) in the second fetus, raising suspicion for TTTS Quintero stage V. She reported no complaints of increasing labor pain or profuse vaginal discharge. She first learned of her twin pregnancy at her 2-month follow-up with an obstetrician. She denied any family history of multiple pregnancies or congenital abnormalities, as well as any history of medication use, herbal intake, industrial area exposure, or chronic illnesses such as hypertension, diabetes, asthma, or heart Due to her symptoms, she initially sought care at a local hospital and was then referred to a tertiary Maternal-Fetal Medicine Polyclinic. This is her third pregnancy, and she has been married once. Her antenatal care (ANC) included six visits, split between a midwife (three visits) and an obstetrician (three visits).

Her physical examination results were within normal limits. On obstetric examination, two fetuses were found with the first fetus in 5/5 breech position and the second fetus in 5/5 head position. Fetal heart rate (FHR) were found in the first fetus and no FHR was found in the second fetus. The first fetus was estimated to weigh 2759 grams and the second fetus was 512 grams. Speculo examination and internal examination were not performed on this patient (Figure 1).

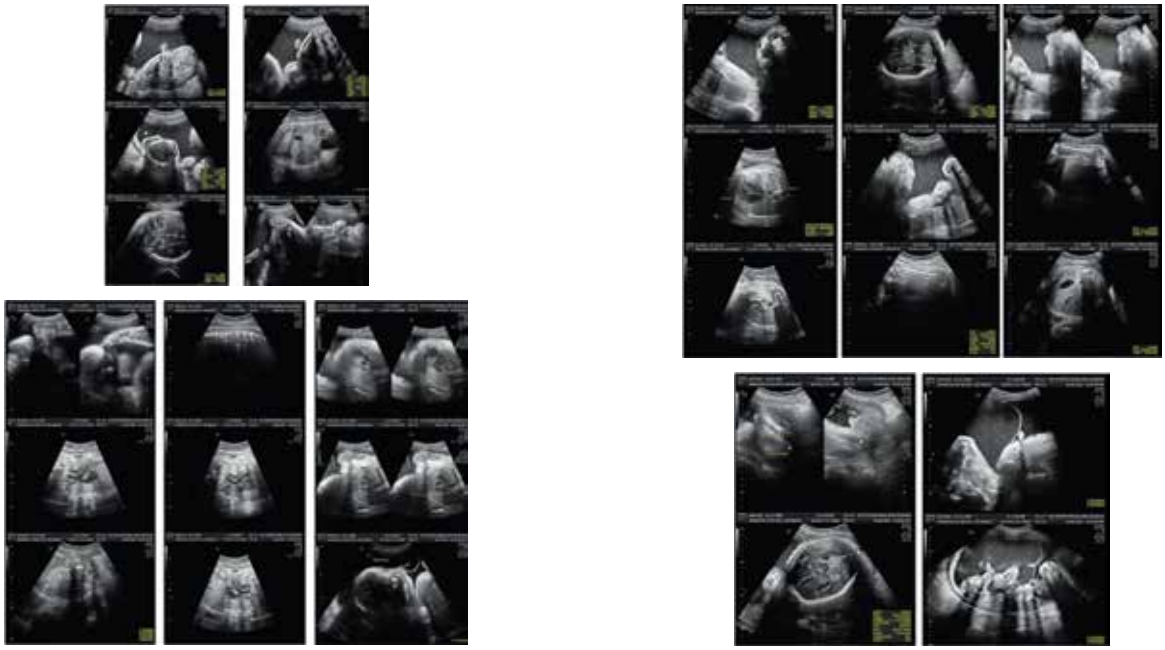

Figure 1. Maternal-Fetal Ultrasound of This Patient's Babies

Laboratory tests showed elevated quantitative D-Dimer levels. Cardiotocography (CTG) for the first twin indicated a category I status. Maternal-fetal ultrasound revealed a 35–36 weeks' gestation with twins, with the first twin in breech position, and the second twin showing polyhydramnios and intrauterine fetal death (IUFD), consistent with Twin-to-Twin Transfusion Syndrome (TTTS) Quintero stage V. Fetal MRI of the head showed no evidence of periventricular leukomalacia or congenital anomalies (Figure 2).

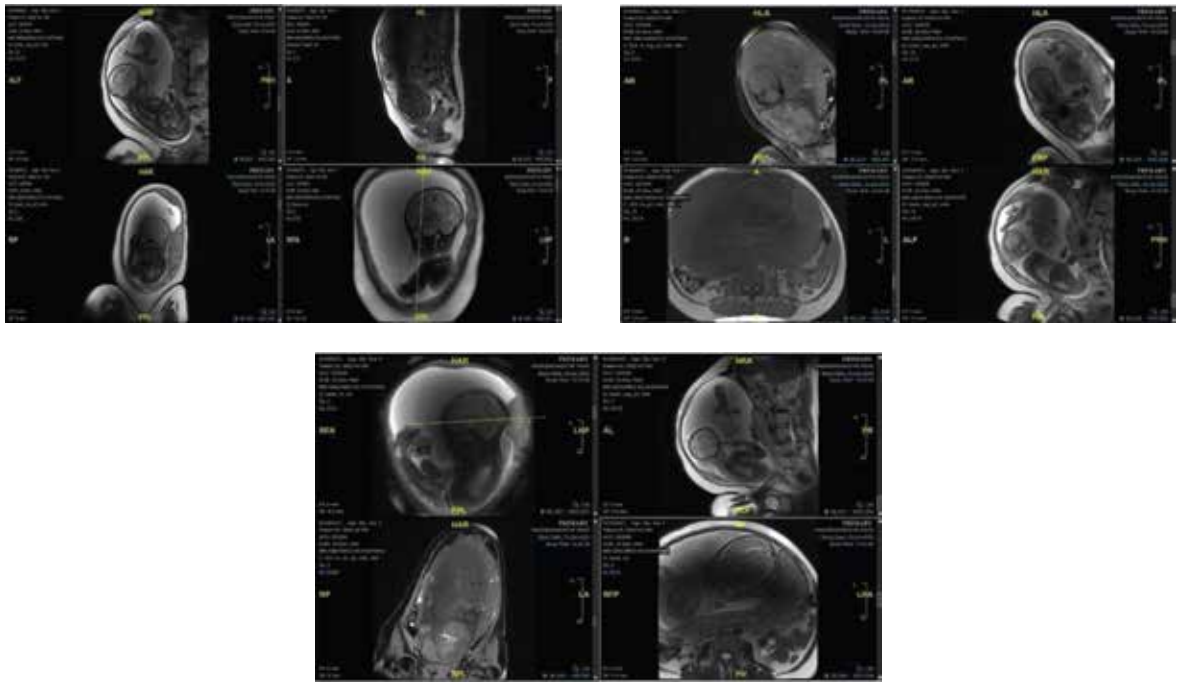

Figure 2. Maternal Abdominal MRI for The First Baby

The initial working diagnosis was G3P2A0 35-36 weeks of gestational age; Gemelli (Multiple Pregnancy); Baby I was in breech position; Baby II was in head position; Polyhydramnios, IUFD; Suspected Twin-to-Twin Transfusion Syndrome (TTTS Quintero V). Conservative treatment was

initially planned. At the following weeks, the patient was admitted to the Obstetrics emergency ward with a diagnosis of G3P2A0 gravida 35-36 weeks; Gemelli; Baby I was in breech position; Baby II was in head position, polyhydramnios, IUFD; suspected Twin to Twin Transfusion Syndrome (TTTS Quintero V). Then conservative management was carried out, until the two following days, an irregular FHR was found in the first baby with a CTG impression of category III. Intrauterine resuscitation was attempted, but as fetal distress in Baby I persisted, an emergency caesarean section was decided. Informed consent and close monitoring of patients were performed. The first baby boy was born weighing 2610 grams and APGAR Score 1'/5' was 7/9. The second baby was born without signs of life, weighing 510 grams, already experiencing grade III maceration. The placenta was born after active management in the third stage with a size of 500 grams (monochorionic diamniotic) (Figure 3).

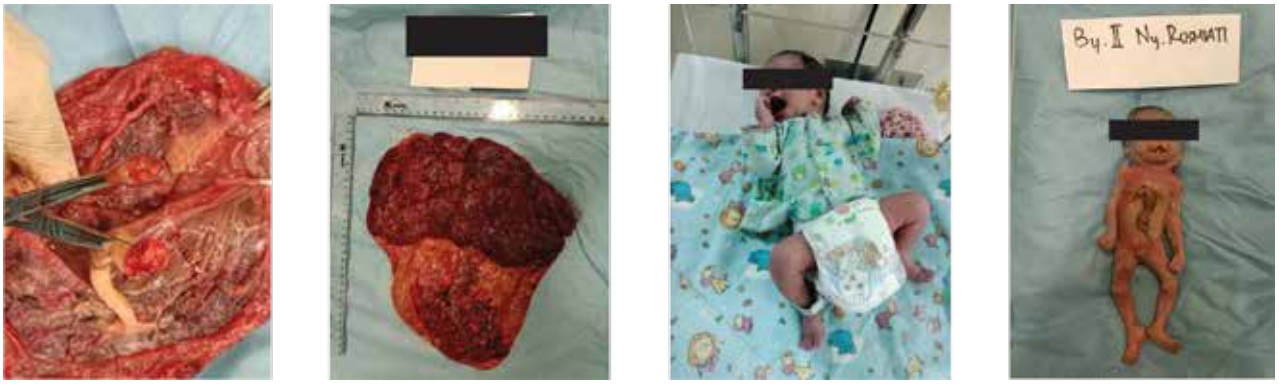

Figure 3. Babies and Placenta

The histopathological examination of the placenta revealed chorangiosis in the larger placenta. The smaller placenta was noted to be within normal limits, as were both umbilical cords.

DISCUSSIONS

In this case, the patient was a G3P2A0 gravida 35-36 weeks; Gemelli; Baby I was in breech position; Baby II was in head position, polyhydramnios, IUFD; suspected Twin-to-Twin Transfusion Syndrome (TTTS Quintero V). Multiple pregnancies are associated with an increased risk of complications during pregnancy, such as hypertension, spontaneous abortion, placenta previa, and fetal malformations. They are also associated with an increased risk of preterm labor, premature rupture of membranes, low birth weight of the baby, and low APGAR scores, which in turn It is also associated with an increased risk of perinatal death in multiple pregnancies. [1-4]

Twin-to-twin transfusion syndrome (TTTS) is a significant issue in many monochorionic pregnancies. In these pregnancies, the fetuses share a single placenta, and nearly all cases involve vascular anastomoses within the chorion that connect the two fetal umbilical circulations. The TTTS mechanism is based on unidirectional blood flow through deep arteriovenous anastomoses. Deoxygenated blood from the placental arteries of both the recipient and donor twins is pumped toward their respective cotyledons. Once oxygen exchange occurs in the chorionic villi, the oxygenated blood is transported out of the cotyledons via the placental vein of the recipient twin. [1,2,5-7.]

Multiple anastomotic connections are probably present in all monochorionic placentas. Monochorionic twins' placentas form a special anastomosis. Three types of anastomoses are possible; arterio-arterial, arteriovenous, and veno-venous (25%). Anastomoses between the veno-venous and arterio-arterial veins are "superficial" and "bidirectional." Anastomosing blood vessel connections, which enable connections between arteries and veins, are referred to as superficial connections because they show up on the chorion's surface. According to the pressure differential between the twins, the flow in this region is bidirectional. Veno-venous anastomosis can be fatal and is linked to lower rates of perinatal survival. Similar to arteriovenous anastomoses, arterio-arterial anastomoses are flexible and can compensate for imbalances that arise in unidirectional flow. There is typically only one type of arterio-arterial

anastomosis in monochorionic placentas. Veno-venous anastomoses in twins with monochorionic placentas cause the venous drainage, which carries oxygen-rich blood, to become rigid and divided. [5,7]

Arteriovenous anastomoses, on the other hand, are "deep" and "unidirectional." While "unidirectional" anastomosis only permits flow in one direction, "deep" anastomosis takes place in the placental lobule's deeper capillary layers. One twin can receive an arterial supply from the other child through an arteriovenal anastomosis, while the other twin receives venous (oxygen-rich) drainage. The monochorionic placenta in arteriovenous anastomosis is actually composed of three parts: two that belong to each twin separately and a third that is supplied by the arteriovenous anastomosis and belongs to the twins collectively.[5,7] Because arteriovenous anastomoses are unidirectional, transfusion imbalance may result unless other deep anastomoses or superficial anastomoses provide sufficient compensation through oppositely directed transfusions. Multiple arteriovenous and venoarterial anastomoses, in addition to arterio-arterial and/or veno-venous anastomoses, are present in about 90% of monochorionic placentas. [4,6] Arterioarterial anastomoses are more effective at compensating for flow imbalances than opposite-direction arteriovenous anastomoses, according to mathematical computer models that simulate TTTS. This is because the arterio-arterial anastomosis, which takes place at the capillary level, has substantially less resistance than the arteriovenous anastomosis.[4,6]

Despite the fact that vascular anastomosis is a necessary anatomical prerequisite for TTTS pathogenesis, the actual mechanism is more intricate than a simple net transfer of red blood cells because hemoglobin levels are typically similar in both twins. As previously stated, TTTS is not a hemoglobin incompatibility issue with polycythemia recipients or anemic donors; rather, it is an amniotic fluid incompatibility issue with volume-overloaded recipients and volume-deficient donors. As a result, hormones associated with maintaining fluid and pressure homeostasis may also be at play. Keep in mind that when two twins exchange blood, one of them naturally becomes exposed to the other's hormonal environment. Consequently, the recipient's hypertensive cardiomyopathy and volume overload may be partially explained by the transfer of renin-angiotensin-aldosterone effectors from the donor. [1,2,5-7]

Acute hypotension may cause cerebral pathology in the victim if one of the affected pregnancy's twins passes away. An embolism of thromboplastic material derived from a deceased fetus is a less common cause. When one twin dies (fetal demise/IUFD), brain damage and rapid antenatal hypovolemia and ischemia are caused by transfusion of the twin anastomosis from the surviving twin's high-pressure blood vessels to the dead twin's low-resistance blood vessels. According to one piece of research, there is a 26% higher chance of neurodevelopmental morbidity and complications from single fetal death in monochorionic twins than in dichorionic twins. The gestational age at the time of the infant's death is correlated with this morbidity. Monochorionic twins have nearly eight times the risk of neurodevelopmental morbidity compared to dichorionic twins at the same gestational age if death occurs between 28 and 33 weeks of gestation (OR=1.48).[5]

Data showed that in this instance, the first baby in a monochorionic diamniotic pregnancy had a different amnion size (normal), while the second baby had polyhydramnios and intrauterine fetal death (single fetal demise). As a result, Quintero V's finding was deemed atypical as it did not fit the criteria for Twin-to-Twin Transfusion Syndrome (TTTS). The Cincinnati Modification of Quintero Staging System could not be applied in this instance since fetal cardiomyopathy was not evaluated. Due to a normal middle cerebral artery-peak systolic velocity (MCA-PSV), there were no Twin Anemic-Polycythemic Syndrome (TAPS) sequelae. According to the mother's abdominal MRI results, the recipient child did not have any neurological complications (which typically occur in 18% of cases of single fetal demise in the cotwin). No congenital abnormalities were found in the recipient, such as acardiac twins and there were no sequelae of Twin Reversed Arterial Perfusion Sequence (TRAP). However, a finding in this case did not align with the underlying theory: the SDP level of the donor fetus (the smaller fetus) was normal (no oligohydramnios), and the larger fetus did not exhibit polyhydramnios; in fact, the opposite was observed. This finding should be noted carefully, as it raises the question of whether atypical presentations of TTTS are common or if this finding may result from other conditions that warrant further investigation.

For TTTS, several treatment options are available, including selective feticide, amnioreduction, laser

ablation of the vascular placental anastomosis, and septostomy. Amnioreduction involves needle drainage to remove excess amniotic fluid. Although septostomy, which creates an intentional hole in the dividing amniotic membrane, was once used, it has largely been discontinued as a medical intervention (Society for Maternal-Fetal Medicine, 2013). Randomized trials indicate that laser therapy offers better neonatal outcomes compared to selective amnioreduction. A trial involving 42 women found comparable 30-day survival rates of one or both twins treated with either selective fetoscopic laser ablation or amnioreduction (75% vs. 65%, respectively). Further assessment of twins from the Eurofetus trial up to six years of age showed no significant improvement in neurologic outcomes or additional survival benefit after six months. For severe TTTS (stages II–IV), laser ablation of the anastomosis is currently the recommended treatment. However, the optimal approach for stage I TTTS remains debated. Close and continuous monitoring is essential after laser therapy.[6,7-10]

If growth disturbances and excessive amniotic fluid occur before 20 weeks, fetal therapy has typically been considered. In most cases, no action is necessary, and both fetuses will die. Because the twins' circulatory systems are shared, any drug injected into one can have an impact on the other. Feticide techniques (also known as fetal therapy) for fetuses that are chosen for reduction thus comprise techniques such as radiofrequency ablation, fetoscopic ligation, or coagulation using laser or monopolar energy to obstruct the umbilical vein or cord. The remaining fetus is still at significant risk, though, even after this surgery.[1,5-9]

Management decisions should be based on the surviving fetus's risk, cause of death, and gestational age. For this specific indication, further monitoring is not necessary in the case of fetal loss occurring in the first trimester. However, if fetal loss occurs after the first trimester in monochorionic twin pregnancies, there is a significant risk of morbidity or mortality for the surviving twin. This is often due to vascular anastomoses, which can cause one twin's death and lead to sudden hypotension in the other. In cases where one twin in a monochorionic pregnancy dies after the first trimester but before viability, pregnancy termination may be considered due to the increased risks associated with continued pregnancy. Occasionally, maternal complications may result in the death of one fetus but not all of them. Since delivery usually takes place three weeks after fetal death is diagnosed. The goal in managing these cases is to prolong the pregnancy while minimizing risks, regardless of whether the intrauterine environment is compromised.[4-6,11,12]

It is controversial to schedule an elective delivery for the late second or early third trimester following conservative management of a singleton fetal death. Twin pregnancies that are monochorionic are more challenging to handle and typically end between 34 and 37 weeks of gestation. Most choose delivery over gravid management when there is a single fetal death at term, particularly if the cause is unknown. In cases like these, the American College of Obstetricians and Gynecologists, or ACOG (2016), advocates for a customized approach.4-6 In this instance, fetal lung maturation was the goal of the patient's conservative treatment plan. Later, while the patient was being observed, it was found that baby I was experiencing fetal distress. This led to the decision to perform a cesarean section to end the patient's pregnancy.

CONCLUSIONS

The existence of "unidirectional blood flow from deep arteriovenous anastomoses" could cause of suspected TTTS Quintero V. Based on ultrasonography results, TTTS is diagnosed. A diamniotic monochorionic gemelli pregnancy with IUFD in the second fetus II led to the clinical suspicion of TTTS Quintero V in this instance. In this instance, there were aberrant results, though, as the donor fetus's SDP level was normal (not oligohydramnios). In this instance, no additional follow-up issues were discovered. For TTTS, a number of treatments are available, such as selective feticide, amnioreduction, laser ablation of the vascular placental anastomosis, and septostomy. It is controversial to deliver a baby in the late second or early third trimester following management of a singleton fetal death. The mode of delivery is chosen based on obstetrics indications.

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