

REVIEW OF TWIN-TO-TWIN TRANSFUSION SYNDROME: A COMPREHENSIVE ANALYSIS

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ABSTRACT

Twin to twin transfusion syndrome (TTTS) is a frequent complication that typically occurs during the second trimester of pregnancy in identical twins who share the same placenta. It happens when one twin transfers fluids and hormones to the other through blood vessel connections on the placenta. If TTTS is not recognized and treated, it is the primary cause of fetal loss before they reach viability. The most reliable method to determine if twins share a placenta is through ultrasound in the first trimester, which guides the monitoring of these twins. TTTS is diagnosed using ultrasound, where one twin experiences excessive amniotic fluid (polyhydramnios) due to an overload of fluids and frequent urination (polyuria), while the other twin has insufficient

amniotic fluid (oligohydramnios) due to decreased urination (oliguria). The severity of the condition is assessed by evaluating bladder filling and the blood flow in the arteries and veins using Doppler patterns. Examining the cardiac function of the fetuses provides insights into the cardiovascular impact of TTTS and helps identify fetuses that may need postnatal follow-up. The standard treatment for TTTS is fetoscopic laser ablation, which involves sealing the blood vessel connections between the twins to make their circulations independent of each other and cure the condition. To improve overall outcomes and understand the long-term effects of TTTS, strategies to reduce preterm birth after treatment and standardized reporting by laser treatment centers are crucial considerations.

Keywords: Twin to twin Transfusion Syndrome, Placenta, Pregnancy, Birth.

INTRODUCTION

Over the past three decades, the occurrence of twin pregnancies has significantly risen in high-income and middle-income countries. This increase can be attributed to factors such as women waiting until later in life to conceive, a decline in fertility rates, and more effective utilization of assisted reproductive techniques. It's widely accepted that twin pregnancies are associated with elevated risks for both mothers and their infants. Maternal adaptation to a twin pregnancy often leads to several complications. Notably, the risk of maternal mortality in twin pregnancies is 2.5 times greater than that in singleton pregnancies. Additionally, the perinatal mortality rate in twins is two to three times higher compared to singleton infants, primarily due to factors like low birth weight, premature birth, fetal growth limitations, and oxygen deprivation during childbirth. Twin to twin transfusion syndrome (TTTS) is a condition that affects about 10-15% of monochorionic twins due to an imbalance in fluid exchange across the blood vessels connecting them. It stands as the primary cause of pregnancy loss before viability in this type of twin pregnancy. Identifying monochorionic during the first trimester and adhering to international guidelines for regular monitoring, at least every two weeks after 16 weeks of pregnancy, offers the best chance for early diagnosis and effective treatment through fetoscopic laser surgery. The current surgical approach results in a survival rate of over 70% for at least one of the twins, although preterm birth is a common outcome of the intervention. This article reviews the unique characteristics of the monochorionic placenta that contribute to TTTS, as well as the methods of diagnosis, treatment, and expected outcomes.

FORMS OF TWIN PREGNANCY

Monozygotic Twins

Monozygotic twins originate from the division of a single fertilized egg. The specific types of monozygotic multiple pregnancies that occur depend on when the egg division takes place.

Dizygotic Twins

When two or more eggs are individually fertilized by sperm, it leads to the formation of dizygotic twins, also known as nonidentical or fraternal twins. Because they are fertilized by separate sperm cells, these twins can be of different genders. Additionally, in the case of dizygotic twins, each embryo has its own placenta, and there is no connection between the circulatory systems of the two embryos.

Diamniotic Dichorionic Monozygotic Twin Pregnancy

This type of monozygotic twin pregnancy occurs when the embryo splits at or before the third day of gestation. This division results in the development of two separate chorions and two amniotic sacs. It can lead to the formation of either two distinct placentae or a single fused placenta. Approximately 8% of all twin pregnancies fall into this category.

Diamniotic Monochorionic Monozygotic Twin Pregnancy

The embryo division in this scenario occurs after the formation of the inner cell mass, typically taking place between the 4th and 7th day of gestation. This results in the development of a single chorion but two separate amnions. Approximately 20% of all twin pregnancies fall into this category.

Monoamniotic Monochorionic Monozygotic Twin Pregnancy

In this case, the embryo division occurs between the 8th and 12th days of gestation, leading to the development of a single chorion and a single amnion. Such instances of monozygotic twins are quite rare, accounting for less than 1% of all twin pregnancies.

Conjoined or Siamese Monozygotic Twin Pregnancy

When the embryo division takes place at or after the 13th day of gestation, it results in the development of conjoined twins who share a specific part of their bodies. The occurrence of this type of monozygotic twins is exceedingly uncommon. The connection between the twins can initiate at various points and may be either dorsal, ventral, or lateral. Among the different types of conjoined twins, parapagus twins (laterally joined) are the most prevalent.

MECHANISMS OF TWIN-TO-TWIN TRANSFUSION SYNDROME

TTTS occurs in approximately 10-15% of cases involving monochorionic twins when there is an imbalance in the transfer of fluids and vasoactive substances across arteriovenous vascular connections. Comparing the placentas of uncomplicated monochorionic twins to those with TTTS reveals that the presence of imbalanced arteriovenous connections is a crucial factor in the development of this condition. Surface-to-surface connections between arteries or veins are considered protective against TTTS, as they enable more efficient redistribution of fluids across various vessel diameters compared to reciprocal artery-to-vein connections. This continual net transfer of fluids and vasoactive substances from one twin to the other results in abnormal intravascular volume status and triggers compensatory responses in both twins. Observable ultrasound findings include disparities in amniotic fluid levels, bladder filling, and cardiovascular effects. The recipient twin experiences increased blood flow to the umbilical

vein, indicating higher preload. This increased stretching of the cardiac chambers leads to the release of atrial natriuretic peptide and brain natriuretic peptide, which stimulate diuresis, causing polyhydramnios. Additionally, there is an elevation of the potent vasoconstrictor endothelin, contributing to hypertension in the recipient twin, and subsequently leading to cardiac hypertrophy and valvular regurgitation. On the other hand, the donor twin experiences hypovolemia, resulting in decreased urine production, which leads to oligohydramnios and minimal or no visible bladder filling. As a response, the renin-angiotensin system becomes more active, and this effect is transmitted to the hypervolemic recipient twin through their shared circulatory system. This amplifies hypertension and cardiomyopathy in the recipient twin, and these effects cannot be solely attributed to changes in volume status. The hemodynamic consequences of TTTS can contribute to both functional and, ultimately, structural heart issues for each fetus. Changes in the cardiac function of the recipient twin may be observed before TTTS fully develops and can include cardiac enlargement, biventricular hypertrophy, valvular regurgitation, and impaired contractility. In some cases, right ventricular hypertrophy and hypertension, along with tricuspid regurgitation, may lead to reduced flow through the pulmonary valve and the right outflow tract. This essentially creates a functional obstruction just below the pulmonary valve in up to 9% of recipients. While these findings may improve after treatment, there may be persistent pulmonary stenosis or functional atresia, which would necessitate postnatal intervention. Structural heart problems and cardiac dysfunction are less likely to be diagnosed in donor fetuses. Nevertheless, decreased blood flow through the aortic isthmus due to hypovolemia, reduced venous return, and increased placental resistance may lead to the development of coarctation.

MANAGEMENT OF TWIN TO TWIN TRANSFUSION SYNDROME

As TTTS tends to have a more favorable outcome when diagnosed at earlier stages, in cases of severe TTTS occurring at a very early gestational age (before 16 weeks), termination of the pregnancy may be considered as an option. Various therapies currently in use focus on achieving a balance in fluid levels between the two amniotic sacs and disrupting the connection of blood vessels between the twins.

Anniotic Fluid Reduction through Amniocentesis:

Serial amniocentesis is a method that entails the extraction of excess amniotic fluid from the recipient twin's amniotic sac through the amniocentesis procedure. This approach may be

applicable for less severe cases of TTTS that occur later in the pregnancy. However, it is generally not considered effective for more advanced stages of TTTS (specifically stages III and IV). Typically, no more than 5 liters of amniotic fluid are removed during a single session, which usually lasts around 30 minutes or less. It's important to note that this procedure may only temporarily restore amniotic fluid balance in both twins' sacs since fluid levels may return to their previous state within a few days. Therefore, the procedure may need to be repeated every few days. Repeated amniocenteses as a treatment for TTTS can result in various complications, including premature labor, premature membrane rupture, and, in rare cases, infection or placental abruption. Pregnancies managed with serial reduction amniocentesis typically lead to deliveries around the 29-30 week mark of gestation.

Septostomy

Septostomy involves the creation of an opening in the membrane that separates the amniotic sacs of the fetuses using a needle. This opening facilitates the transfer of fluid from the recipient twin's amniotic sac to the sac of the donor twin, which has either minimal or no fluid. Although the risk of complications such as infection, premature labor, and premature rupture of membranes is rare, membrane perforation comes with an additional potential risk—the possibility of the hole between the two sacs becoming too large. In some cases, this can lead to the complete disruption of the separating membrane, allowing the babies to share the same amniotic space. In the worst-case scenario, this could result in the entanglement of the umbilical cords of the two twins, potentially leading to the death of one or both fetuses. However, the advantage of membrane perforation over amnioreduction is that patients undergoing this procedure typically require fewer interventions compared to those treated with amnioreduction.

Selective Cord Coagulation

In this medical intervention, performed under the guidance of ultrasound, one of the twins is intentionally sacrificed to save the life of the other twin. This procedure is employed when it is either impossible to conduct laser ablation of the connecting vessels or when one of the twins is in such critical condition that laser ablation is unlikely to succeed. By halting the blood flow in the umbilical cord of the twin facing a life-threatening situation, the other twin can be shielded from the adverse consequences of their sibling's demise. In this process, the umbilical cord is held, and an electrical current is applied to coagulate the blood vessels within the cord to obstruct the blood flow through them. Complications associated with this procedure may include

premature delivery and the premature rupture of the fetal membranes.

ROLE OF NURSE IN MANAGING TTTS

Given the challenging outlook for fetal well-being in Twin to Twin Transfusion Syndrome (TTTS), healthcare providers play a crucial role in addressing this condition. It is imperative for them to have a comprehensive understanding of the underlying pathophysiology of TTTS. This includes recognizing how imbalances in blood and fluid exchange between the twins via shared placental connections can lead to severe complications. Early diagnosis of TTTS in pregnancy is of paramount importance. Identifying this condition as soon as possible allows for prompt intervention, increasing the chances of a positive outcome. Early detection enables healthcare providers to assess the severity of TTTS and formulate an appropriate treatment plan tailored to the specific needs of the expectant mother and her twins. Effective management strategies are vital in addressing TTTS. These strategies may encompass a range of medical interventions, from amnioreduction to fetoscopy laser ablation or selective cord coagulation, depending on the stage and severity of the condition. Coordinating these interventions and closely monitoring the pregnancy ensures that the most suitable treatment options are employed to maximize the chances of a successful outcome for both twins. In addition to the medical aspect, healthcare providers must also play a role in patient education and support. Patients and their families need to be informed about the condition, the treatment options, potential complications, and expected outcomes. Offering emotional and psychological support is equally important, as TTTS can be an emotionally taxing experience for the pregnant woman and her family.

CONCLUSION

To improve overall outcomes and understand the long-term effects of TTTS, strategies to reduce preterm birth after treatment and standardized reporting by laser treatment centers are crucial considerations. To enhance perinatal results, it is crucial to achieve precise diagnosis and appropriately direct TTTS cases to specialized laser treatment centers. Furthermore, effective interventions that aim to minimize preterm birth, especially in cases involving a shortened cervix, are essential advancements. The adoption of standardized reporting of key outcome measures is a vital initiative for treatment centers to implement.

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