

Twin-to-Twin Transfusion Syndrome - TTTS

At an ultrasound examination of your twins, twin-to twin transfusion syndrome (TTTS) was discovered. This is a brief information about what TTTS is, what consequences it has on your pregnancy and what treatment options are available.

What is TTTS?

Monozygotic twins occur in approximately 3-4/1000 pregnancies. TTTS can occur in monozygotic twins (monochorionic twins), as they share a common placenta. The twins' blood circulations are connected through blood vessels (anastomoses) in the placenta. These anastomoses can cause several different complications. TTTS is a disease that affects the fetal blood circulation and it occurs in 10-15% of all monochorionic twin pregnancies. In all monochorionic pregnancies there is a blood flow (transfusion) across the anastomoses. However, in TTTS the distribution results in an unequal sharing of the blood volume between the twins. The reason for this is unclear, but it leads to an increased blood volume (hypervolemia) in one fetus and a decreased blood volume (hypovolemia) in the other fetus.

How is TTTS detected?

At the ultrasound examination the doctor evaluates the amount of amniotic fluid, the size of the fetuses, the size of the urinary bladders, the appearance of the placenta and the dividing membrane, as well as the fetal and umbilical blood circulations. Sometimes a more thorough examination of the fetal hearts is also performed. The fetus that suffers from a decreased blood volume (donor) often has a decreased growth and stops urinating. As the amniotic fluid largely consists of fetal urine from pregnancy week 15-16, this results in a decreased amount of amniotic fluid (oligohydramnios). There might also be an impaired blood flow in the fetal blood circulation, e.g. in the vessels in the umbilical cord. The fetus that suffers from an increased blood volume (recipient) urinates more in order to lose fluid and this results in an increased amount of amniotic fluid (polyhydramnios). The recipient could also develop heart failure due to the increased demands on the fetal heart. The increased amount of amniotic fluid could cause symptoms to the pregnant woman such as an enlarged and tense uterus and painful contractions.

When does TTTS occur?

TTTS can occur at any time in pregnancy but the most common is between 16-25 weeks of gestation. In order to timely identify TTTS, women with monochorionic twin pregnancies are offered ultrasound scans at least every second week.

What are the consequences of TTTS?

Untreated TTTS results in a severely affected fetal blood flow with oxygen deficit in the donor and heart failure in the recipient. Both conditions increase the risk of brain injury in the fetuses. The risk of extreme premature delivery and late miscarriage is increased due to the increased amount of amniotic fluid. Altogether this means that the risk of both fetuses dying is 80-90% and in those that survive the risk of brain damage is around 50%.

Is there any treatment for TTTS?

Between gestational weeks 15 and 28 there is a possibility to perform fetoscopic laser treatment of the vascular anastomoses in placenta. The aim of this procedure is to find all anastomoses using a fiberoptic camera and with laser coagulate them to stop the blood flow between the fetuses. This is the only procedure that treats the actual cause of TTTS and that gives both fetuses a chance of survival.

However, in some situations it is not possible to safely coagulate all anastomoses. The reason for this could be that the placenta is in a location that makes it difficult to reach all anastomoses or that there are complications such as bleeding into the uterine cavity or that the amniotic membrane detaches from the uterine wall. In some situations, one fetus might already be severely injured by the TTTS. In these circumstances one option could be to close off the umbilical cord to one of the fetuses (cord occlusion). The purpose of this procedure is to increase the chance of healthy survival of one fetus, but means that the other fetus dies.

- Fetoscopic laser treatment: Both fetuses survive in approximately 55-60%. In 25-30% of cases only one fetus survives and in 10% the pregnancy ends with miscarriage.
- Fetoscopic cord occlusion: Only one fetus survives in 80-90%. In 10-20% the pregnancy ends with miscarriage or demise of both fetuses.

A fetoscopic operation generally does not confer increased risks to the pregnant woman. The instrument used is approximately 3 mm in diameter and is inserted through the abdominal wall into the uterine cavity using local anesthesia. Consequently, the pregnant woman is not submitted to general anesthesia. During the operation the doctors examine the pregnancy with ultrasound to precisely determine where to exactly insert the instruments. The operation is conducted under sterile conditions. The woman receives extra anesthetic or sedative medication when needed. She also receives prophylactic antibiotics to decrease the risk of infection and medication to prevent contractions. The risk of bleeding and infection that require treatment is small. Serious complications are very rare. After the operation the woman stays at least one day in the hospital for observation.

The procedure increases the risk of premature rupture of membranes before 32 weeks of gestation. This occurs in 20-25% of cases. The consequences depend on in what gestational age this occurs. As long as there is no infection or bleeding, the pregnancy could still continue several weeks. It is important after the operation to avoid premature delivery before 32 weeks of gestation as this increases the risk of the babies being ill or dying.

Despite a successful operation, these babies are at increased risk of brain damage. The risk is equal in the donor and the recipient. In 5-10% there is a serious neurological handicap such as cerebral palsy. In another 5-10% there is milder neurological handicap with different manifestations. The reason for neurological handicap is both the TTTS and the increased occurrence of premature delivery. Some of the babies that suffer from serious cardiac disease during pregnancy might have remaining cardiac problems. The children need to have a follow-up with a pediatrician and/or pediatric cardiologist into school age.

If TTTS occurs after pregnancy week 28 it might be a better option to attempt to prolong the pregnancy through drainage of amniotic fluid and treat the babies after delivery. However, if possible, fetoscopic laser treatment is a better option until gestational week 28.

Depending on in what gestational age TTTS is discovered there is as well an option to terminate the pregnancy.

What happens after the operation?

The day after the operation the pregnancy is assessed using ultrasound. If the woman is well, she is allowed to go back home after this examination. Follow-up scans are initially performed weekly at her closest university hospital. If the TTTS regresses and there are no complications follow-ups can be scheduled at her local hospital after 2-3 weeks. In approximately 10% some form of twin-to-twin transfusion might reoccur. If this happens before 28 weeks of gestation there might be a possibility to perform a second operation and a new assessment should be performed at the fetal therapy center where the original operation was performed.

What happens if one fetus dies?

If one fetus in a monochorionic pregnancy dies before a fetoscopic operation has been performed, the risk of the other fetus dying is approximately 20-25%. This is due to the vascular anastomoses in the placenta that can cause a rapid decrease in blood pressure and anemia in the other fetus when one twin dies. If the fetus survives there is a risk of brain damage in 20-25% of cases. If the cause of fetal demise in a monochorionic twin pregnancy is TTTS, the risk of brain damage in the survivor is even higher.

If a fetoscopic laser operation has been performed the risk of death or damage in the surviving fetus is substantially less as the vascular anastomoses in the placenta have been closed.

If one fetus dies there is no advantage of early delivery, but the best option for the surviving fetus is to stay in the womb as long as possible. Preferably until 37 weeks of gestation, provided there are no other complications.

Delivery

If the TTTS treatment was successful and no other complications occur during the pregnancy, the recommendation is delivery in gestational week 36-37. Normal vaginal delivery is recommended if there are no other risk factors. If there are any remaining problems the aim is still to delay delivery until 32-34 weeks of gestation if possible and delivery may then have to be with cesarean section. It is important that delivery is planned together with an obstetrician, a fetal medicine specialist and pediatricians specialized in the care of sick newborns (neonatologists). The recommendation is that these children should be born in a university hospital with specific competence to take care of newborns that suffered from TTTS during fetal life. This is particularly important in cases with remaining problems and where delivery is necessary before 32-34 weeks of gestation.