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Twin to Twin Transfusion Syndrome



Anatomy and Physiology of Twinning

The process of having twins occurs in many ways. Twins may be either “fraternal” or “identical”. In fraternal twins, each baby has his or her own placenta and amniotic sac (bag of waters). In identical twins, the two babies may share the same amniotic sac, placenta or both. Twin to Twin Transfusion Syndrome (TTTS) only occurs in identical twins who share a placenta. Most identical twins share blood vessels through the placenta, but twins with TTTS have unequal sharing of blood. One twin may not get enough blood while the other becomes overloaded with too much blood. This can lead to heart failure or growth problems.

Twin to Twin Transfusion Syndrome (TTTS)

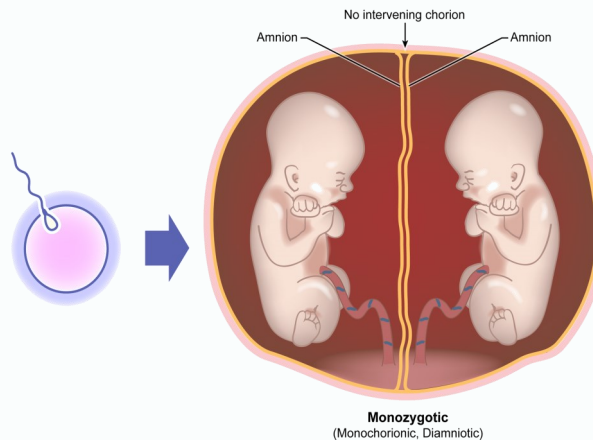
TTTS is a serious complication of an identical twin pregnancy in which there is only one placenta (whenua). It occurs in 10-15% of identical twin pregnancies. When TTTS occurs, there is an unequal sharing of blood between two babies due to blood vessels that communicate between the two babies in the single placenta. In the most serious cases, one baby (called the Recipient twin) is larger and surrounded by an excessive amount of amniotic fluid, while the other baby (known as the donor twin) is smaller and appears to be stuck against the uterine wall due to the reduced amount of amniotic fluid around it.

In the early stages of the disease, signs include a large amount of fluid around the larger Recipient twin and a smaller amount of fluid around the smaller Donor twin. The bladder of the Donor twin may still be seen during an ultrasound examination.

As the disease progresses, the Donor (smaller twin) may no longer have a visible bladder and a very small amount of amniotic fluid or no amniotic fluid. This is referred to as “stuck twin” syndrome, because the smaller twin actually appears to be stuck to the uterine wall or wrapped up in the amniotic membrane (“vacuum packed”).

Ultrasound Doppler studies provide information about the condition of the blood circulation for both of the twins. As the condition worsens, the Donor twin does not receive enough blood and the Recipient twin receives too much blood.

In the advanced stages, an excessive amount of fluid builds up within the Recipient twin due to increased kidney circulation. This is called “hydrops”. Related symptoms include swelling of the skin around the head, swelling of the abdomen (ascites), poor contractions of the heart, and heart failure. Either twin can develop hydrops, but more often it occurs in the Recipient twin.



An ultrasound will be done prior to going to surgery to confirm the babies’ heartbeats are present. When you arrive in the operating room you will be moved to the operating table. At this stage the anaesthetist will proceed with the anaesthetic. This will depend on the individual circumstances, but is likely to be either a local anaesthetic into the skin and sedation through an IV luer or a regional anaesthetic (epidural). You will be covered with a warm blanket to keep you comfortable during surgery.

During surgery, one or two small incisions, approximately 5mm long, are made on the abdomen. The incisions are so small that they only require band-aids to cover after surgery. In most cases surgery lasts one to two hours, but this can vary depending on a number of factors.

After Surgery

Following surgery, you will be taken to the Recovery Room until you are alert. After you are fully awake, you will be taken back to the ward.

Pain or discomfort after surgery seldom occurs. If present, it is usually very minimal. If needed, pain relief medication will be offered. The catheter will be removed from your bladder about four hours after surgery. Your partner or support person may remain with you in your room.

You will be given liquids to drink during the evening after surgery. Following this you may have regular food as tolerated. Activity is restricted to getting up to the toilet only, but this depends upon your specific condition.

The day after surgery, a follow-up ultrasound will usually be done. The reason for this is to allow time to see changes in fluid volume and blood flow in each baby. You will remain in the hospital for 1 to 2 days.

Follow-up Care

After surgery, your original doctor will resume your care for the rest of the pregnancy and delivery with input from the local Fetal Medicine team depending on local expertise. You and your midwife/doctor should feel free to contact us with any questions or concerns.

Weekly ultrasounds are recommended for the next month. After that time, if all is going well, ultrasounds are performed every two weeks or as directed by your doctor. Although you are returning home, we continue to follow your pregnancy closely. Please make arrangements with your doctor to forward all of your ultrasounds and any other information or test results to us. We carefully monitor the outcomes of patients and babies, so we will also follow up all surviving babies at 2 and 5 years of age.

Social Work and Pastoral Care

Representatives from Social Work and Pastoral Care are available for our patients and families for assistance with psychological, social and spiritual needs.

Staging of TTTS based on Sonographic and Doppler Findings

(TTTS) is staged in an effort to offer the most appropriate treatment for the level of disease present. Surgery may not be indicated in all cases.

In **Stage I** there is a small amount of fluid (known as oligohydramnios) in the Donor and a large amount of fluid (polyhydramnios) in the Recipient. At this stage, surgery may not be indicated but close observation for a change in condition is recommended.

In **Stage II**, along with poly/oligohydramnios, there is no visible bladder in the Donor. At this stage, laser surgery is considered.

Stage III is characterized by Critically Abnormal Dopplers (CADs), which means one of the babies has abnormal blood flow. Laser Surgery is offered for this stage. If laser surgery is not possible, umbilical cord ligation may be performed if absolutely necessary.

In **Stage IV**, all of the above findings are present and the baby is hydropic. This means there is swelling of the head and abdomen (ascites), the heart contracts poorly, and heart failure is present. The natural history of this disease has shown that at this stage a baby is not likely to survive.

Stage	Poly/Oligo Hydramnios	Absent Bladder in Donor	Critically Abnormal Dopplers	Hydrops	Demise
I	Yes	No	No	No	No
II	Yes	Yes	No	No	No
III	Yes	Yes	Yes	No	No
IV	Yes	Yes	Yes	Yes	No
V	Yes	Yes	Yes	Yes	Yes

Preparing for Surgery

Before surgery, you will not be allowed to eat or drink for a defined amount of time (usually 6-8 hours). This is to prevent the risk of vomiting during surgery. For a morning procedure you will be asked to fast from midnight the night before.

Before Surgery

When it is time to be admitted to the hospital for surgery, check in at Ward 96/98 at the time arranged.

Rectal suppositories will be given before and after surgery to relax the uterus and stop any contractions. A catheter will be put in your bladder to drain urine and will remain in place for about four hours after surgery.

The pregnancy may be lost from heart overload in the larger twin, lack of enough blood getting to the smaller twin, or preterm (early) labour because of the excess fluid causing the uterus to be 'overstretched'.

The purpose of this booklet is to provide information to you about TTTS, the treatment options, the service provided at National Women's Health and to answer any questions you may have.

Treatment

If not treated, severe TTTS has a mortality rate close to 100%. Current treatment methods include:

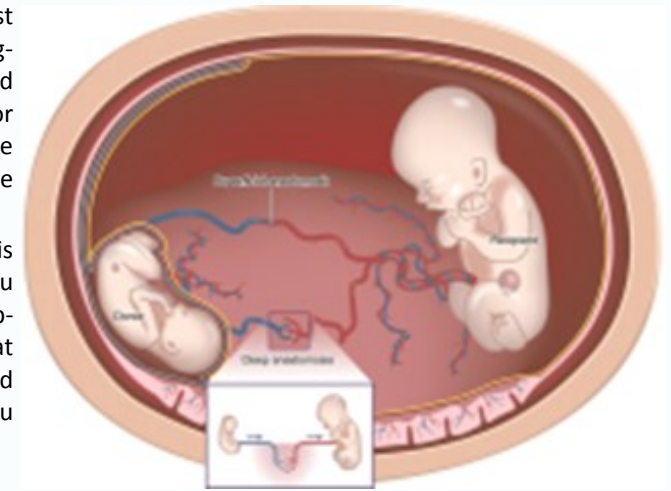
- Serial Amniocentesis
- Umbilical Cord Ligation
- Selective Laser photocoagulation of the communicating vessels
- Termination of Pregnancy

Serial Amniocentesis

In serial amniocentesis excess fluid is removed from the amniotic sac of the Recipient (larger) twin with a needle. Repeated removal of fluid may be needed. Each time fluid is removed, 1-4 litres may be obtained. How often fluid is withdrawn depends upon how severe the case. It may need to be done as often as every second day.

Pregnancies that develop signs of TTTS after 26 weeks gestation are usually managed by serial amniocentesis as the babies are now at a viable age i.e. there is a chance of survival. The success rate (at least one survivor) for this technique varies, but may be as high as 60-70%. The advantage of serial amniocentesis is that it may prevent pre-term labour and thereby extend the pregnancy for a few more weeks. The main disadvantages of this approach are that it does not fix the basic problem (unequal sharing of blood), therefore making repeat removal of fluid necessary, each with its own risk of trauma, infection, or pregnancy loss.

If one baby dies from the disease, the shared blood flow between the babies may allow for the second twin to bleed into the dead twin. This may cause death (up to 40%) or brain damage (15-25%). Serial amniocentesis may also cause the membranes to detach from the uterine wall. This may make any other form of invasive intrauterine therapy more difficult. Furthermore the success rate with this method is poor when the disease is severe (stages 3 & 4).



Umbilical Cord Ligation

In severe cases of TTTS, one of the babies may be extremely sick at the time of presentation. Attempts to save both babies in this case may be futile, as the sicker baby may die despite all efforts. This may also occur at a time before either twin is viable. Spontaneous death of the unhealthy baby may in turn cause death or neurological damage to the co-twin. An umbilical cord ligation may be performed to save the co-twin from harm. The chance of miscarriage following this procedure is about 10%. Umbilical cord ligation is offered only in severe cases and the intent is to save the life of the other twin.

Selective Laser Photocoagulation of Communicating Vessels (SLPCV)

Laser surgery may be performed to stop the sharing of blood between the two babies. The object of this method is to treat TTTS by laser coagulating the specific vessels that cause the problem of blood sharing in this syndrome and spare the remainder of the vessels. The laser is used to seal the vessels to stop the blood flow between the babies.

Generally there is an 80% survival rate for at least one baby and 65% for both, with an incidence of neurological complications of 2-5%. The surgery is done under regional or local anaesthesia. A small incision (5mm) is made in the mother's belly to insert an endoscope and laser fibre, under combined ultrasound and endoscopic guidance. This is a type of key hole surgery. The endoscope is a long, narrow telescope with a light and camera on the end. The blood vessels are identified and sealed through the endoscope.

The main advantage to this method is that the disease is corrected in the majority of cases with a single treatment even for the more severe stages of the disease (stage 3 & 4). In addition, cerebral palsy and other complications caused by the death of one of the twins are less likely to occur (approx 5%) than with serial amniocentesis. If one baby dies, the other twin is not likely to be affected. A recent European study has shown better outcomes for babies after laser therapy than repeated amnioreduction. This procedure is the gold standard treatment for Stage II, III & IV TTTS.

One or Two Babies?

The purpose of surgery is to separate the circulation between the babies by using the laser to seal the vessels connecting the two. In essence, these babies are now like fraternal twins. They are no longer sharing blood and each has its own portion of the placenta. The question many couples raise is why, in some cases, only one baby survives? This depends on how much placenta each baby receives when the placenta forms.

At the time of surgery, we can't give more placenta to either baby, nor can we tell how much placenta each baby has. According to research on placentas inspected after birth, each baby needs approximately 30% of the placenta to survive. If there is a loss of one of the babies after surgery, the other baby will be less likely to be affected since they are no longer sharing blood vessels. Loss can occur anytime from the time of surgery. If both babies are doing well after one month, both are likely to survive.

Termination of Pregnancy

After reviewing all information and options, you may decide you do not want to continue this pregnancy.

Criteria for Surgery

Patients are considered for surgery when the pregnancy is between 17-26 weeks gestation. Further criteria include the following:

- Same sex
- Single placenta
- Minimal fluid in donor (less than 2cm pocket)
- Thin dividing fetal membrane
- Prominent bladder in recipient
- Absence of other structural problems in either twin

A patient may not be offered surgery if the following conditions exist:

- Abnormal genetic studies
- Rupture or detached membranes
- Short or dilated cervix
- Pre-term labour
- Technically not possible to perform

Risks and Complications

There are potential complications associated with the surgery:

- There is the possibility of bleeding in the mother and/or babies, which could prevent the completion of the procedure
- Rarely, bleeding may be of such magnitude that we may need to make an abdominal incision (laparotomy) and place a suture on the uterus
- In extreme circumstances it may be necessary to remove the uterus to control bleeding. This would not allow the mother to have any further children. Severe bleeding could result in damage to many organs, brain damage, or even death
- Preterm labour, amniotic fluid leakage or premature rupture of membranes could occur. If any of these complications occur, we may need to keep the mother in the hospital. Infection of the amniotic cavity may also occur and lead to these complications. If infection is diagnosed, delivery is required to prevent further complications
- Placental abruption or separation has been rarely reported
- If it looks as if one baby is dying during the procedure, we may tie the cord to that twin to try to save the other twin, but only if your consent for this procedure is given in advance

There is a low incidence of these complications as precautions are taken to minimise these risks. You will be placed on antibiotics to prevent infection. Very small instrumentation is used to help prevent bleeding and preterm labour. All patients are evaluated thoroughly prior to surgery.