TTTS INFORMATION PACKET
What is TTTS?

Twin-twin transfusion syndrome (TTTS) occurs in twins that are sharing the same placenta (monochorionic twins or identical twins). Most identical twins share blood through vessels in their common placenta, but twins with TTTS have an unequal sharing of blood. The smaller twin (Donor) does not get enough blood while the larger twin (Recipient) becomes overloaded with too much blood. This patient education guide has been designed to inform you of options available to manage TTTS.

Twins may be either “fraternal” or “identical”. In fraternal twins, each baby has his or her own placenta and amniotic sac (bag of waters). Identical twins share a common placenta. TTTS occurs in identical twins when there is an uneven sharing of blood between two babies through blood vessels that are present in a common placenta. One baby (Recipient) has an excessive amount of amniotic fluid (polyhydramnios), while the other baby (Donor) appears to be stuck against the uterine wall due to very little or lack of amniotic fluid (oligohydramnios). TTTS occurs in 5-10% of identical twin pregnancies. If TTTS is left untreated, the pregnancy may be lost from cardiac overload in the larger twin, lack of enough blood getting to the smaller twin, or uterine contractions leading to miscarriage or premature delivery.

For more information on TTTS and a forum to communicate with other patients who have experienced TTTS, please visit [http://www.fetalhope.org](http://www.fetalhope.org) or [www.tttsfoundation.org](http://www.tttsfoundation.org)

How is TTTS Diagnosed?

The diagnosis of TTTS is made with an ultrasound evaluation, which shows polyhydramnios (maximum vertical pocket of amniotic fluid is 8 centimeters or more) in the Recipient twin, and oligohydramnios (maximum vertical pocket of 2 centimeters or less) in the Donor twin. Typically, a single placenta is present and the external genitalia are the same. A thin dividing membrane is also present in monochorionic/diamniotic twins (identical twins with one placenta and two sacs).

What are the Stages of TTTS?

TTTS does not behave in the same way in all patients; some cases are more severely affected than others. A classification of TTTS into five stages by severity has been developed by us.

- **Stage I** is characterized by a small amount of fluid (oligohydramnios) in the Donor (maximum vertical pocket of fluid is 2 centimeters or less) and a large amount of fluid (polyhydramnios) in the Recipient (maximum vertical pocket of fluid is 8 centimeters or more). The bladder of the Donor baby is visible.
- **Stage II**, along with poly / oligohydramnios (as defined above), there is no visible bladder in the Donor twin during the entire ultrasound evaluation.
- **Stage III** is characterized by one or both babies having ultrasound evidence of poor blood flow “Critically Abnormal Dopplers” (CADs). Critically Abnormal Dopplers are defined as the presence of at least one of the following:
  - Absent or reverse end-diastolic velocity in the umbilical artery (no blood flow or reversed blood flow through the umbilical artery in the latter part of the cardiac cycle).
Reverse flow in the ductus venosus (reversed blood flow in the vessel that goes through the liver to the heart).

- Pulsatile umbilical venous flow (blood is backed up from the heart into the umbilical vein).

In Stage III, if the Donor’s bladder is not seen, the TTTS is classified as “Classic”.

In Stage III, if the Donor’s bladder is seen, the TTTS is classified as “Atypical”.

- **Stage IV** is characterized by hydrops in one or both babies. This means there is fluid accumulation in any part of the baby such as: swelling of the head (scalp edema), abdomen (ascites), lungs (pleural effusions) or heart (pericardial effusions). This is evidence of heart failure.

In Stage IV, if the Donor’s bladder is not seen, the TTTS is classified as “Classic”.

In Stage IV, if the Donor’s bladder is seen, the TTTS is classified as “Atypical”.

- **Stage V** is when one or both babies have died.

### SUMMARY OF ULTRASOUND FINDINGS USED TO DETERMINE TTTS STAGE

<table>
<thead>
<tr>
<th>Stage</th>
<th>Poly/Oligo Hydramnios</th>
<th>Absent Bladder in Donor</th>
<th>Critically Abnormal Dopplers*</th>
<th>Hydrops</th>
<th>Demise</th>
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<tbody>
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<td>I</td>
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### What Is SLPCV?

The purpose of selective laser photocoagulation of communicating vessels (SLPCV) is to separate the circulation between the babies by using laser energy to seal off the vessels connecting the two. As a result, the babies no longer share blood and TTTS is cured. There are other blood vessels in the placenta that do not allow sharing of blood between the babies. These vessels feed areas of the placenta that belong to each twin. These vessels are not targeted during laser surgery.

### How is SLPCV Performed?

The procedure will preferably be performed under intravenous (IV) sedation and local anesthesia. Occasionally, with local or epidural anesthesia, the fetuses are still able to move. In that case, the recipient twin may need to be kept from moving in order to perform the surgery. Injecting a muscle-blocking drug into the thigh of the fetus with a very thin needle under ultrasound guidance does this. General anesthesia or epidural anesthesia may be used under certain circumstances.

**Procedure**

After adequate anesthesia, a small 2-3 mm (1/10") skin incision is made and a trocar (small metal tube) inserted into the amniotic sac of the recipient twin. An amniotic fluid sample may be obtained for genetic or microbiological studies. The pressure in the amniotic cavity may be measured with a special gauge attached to the trocar. An endoscope (medical telescope) is passed through the trocar to observe the blood vessels on the surface of the placenta. The blood vessels that link the circulations will be identified and interrupted (sealed off) using laser energy. A second trocar may need to be inserted in a small number of cases, especially if the placenta is anterior (front of the uterus). At the end of the procedure the sac of the recipient twin may need to have some of its fluid drained. You will be given antibiotics prior to and after surgery.

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One Baby or Two?

Occasionally, one baby dies after laser treatment. Babies may die because they are unhealthy due to TTTS or other conditions or possibly because of inadequate placenta share. At the time of surgery, it cannot be estimated whether a baby will have enough placenta or not. An estimate of how much placenta each baby has cannot be made either. Typically, if one of the babies dies, the other baby should not be affected since they are no longer sharing blood vessels. Death of one or both of the babies can occur anytime from the time of surgery until delivery.

What are the Criteria for Laser Treatment of TTTS

The criteria for laser surgery for TTTS includes:

1) Gestational age: 16 weeks, 0 days to 26 weeks, 0 days.
2) Diagnosis of TTTS:
   a) Single placenta with thin dividing membrane (shared placenta)
   b) Polyhydramnios: amniotic fluid maximum vertical pocket of 8 centimeters or more in the Recipient twin, prior to amniodrainage.
   c) Oligohydramnios: amniotic fluid maximum vertical pocket of 2 centimeters or less in the Donor twin, prior to amniodrainage.
   d) Same gender, if visible.

Exclusion criteria:
1) Abnormal Genetic studies.
2) A septostomy has been done. This is a hole in the dividing membrane (the membrane separating the babies) that has been made intentionally.
3) Ruptured membranes (leakage of amniotic fluid from the vagina).
4) Chorioamnionitis (infection in the uterus).
5) Ultrasound evidence of brain damage of either baby.
6) Placental abruption (separation of the placenta from the uterus)
7) Active labor

What Can Be Expected After Surgery?

Typically, you will remain in the hospital for 1-2 days. You will undergo an ultrasound the day after surgery to determine how the babies are doing.

You will then be discharged home to the care of your primary obstetrician and perinatologist. You will be asked to undergo a weekly ultrasound examination for a minimum of four weeks after the initial therapeutic procedure.

Possible Benefits of the Surgery

Laser surgery may benefit you directly, by allowing the pregnancy to continue and improve the odds of a successful outcome. Overall, current figures estimate that the outcomes of this procedure are approximately as follows: 85% chance of at least one living fetus (one or two), 50-60% chance of both fetuses surviving, and 15% chance of losing both fetuses. The main advantage to this method is that the surgery can cure TTTS. In addition, the incidence of cerebral palsy is approximately 5%. Death of one baby in utero typically is not associated with harm to the other baby.
What are the Complications of the Surgery?

Below is a list of some of the complications from this surgery.

1) Inability to perform the procedure due to intraamniotic bleeding or technical complications. For example, it is possible that the amniotic fluid could be blood tinged from previous procedures and not allow an adequate visualization.

2) Bleeding from the vessels being lasered (sealed off).

3) Death of one or both twins. This may be due to TTTS, complications of surgery or possible lack of enough placenta after surgery.

4) Cerebral palsy or other forms of brain damage. This complication may be present before surgery, after surgery or develop after birth.

5) Preterm labor, amniotic fluid leakage, gross premature rupture of membranes or detached membranes (the membranes are detached from the uterus). If an unintentional septotomy occurs (a hole in the membrane between the twins), there is an increased risk of fetal death from the umbilical cords of the babies becoming entangled. If any of these complications occur, you may need to remain in the hospital. Infection of the amniotic cavity may also occur in this setting and lead to these complications. If infection is diagnosed, you need to be delivered to prevent you from having further complications.

6) A significant drop in the heart beat of one or both twins during surgery. If this should occur, and the fetuses are viable (of an adequate age and condition to live outside the uterus), an emergency cesarean section could be performed.

7) Physical injury to you or the fetuses.

8) Bleeding from the uterine wall. This may not allow the surgery to be completed. There can be bleeding after removal of the trocar. This is usually controlled by placing external pressure on the surgical site. Occasionally, however, it may be necessary to perform a skin incision and place a suture on the wall of the uterus.

9) Placental abruption (separation of the placenta from the uterus). This is a rare but serious complication. If an abruption does occur, the babies would have to be delivered by an incision into your uterus (hysterotomy).

10) Need for blood transfusion. Bleeding of significant magnitude may require a blood transfusion (giving you blood).

11) Bleeding may be of such magnitude that it may be necessary to remove the uterus (hysterectomy). This would not allow you to have any further children. Severe bleeding may result in damage to many of your organs, brain damage, or even death.

12) It is possible that neither twin has any placental territory of its own (circular pattern). Under these rare circumstances, lasering of all communications would result in the death of both twins. It will not be known if your placenta has a circular pattern until surgery. If a circular pattern is observed, the surgeon may decide not to perform the laser surgery or only laser a few of the communications. If communications are not lasered, you may need treatment of TTTS with serial

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amniocenteses. The risks and benefits of laser therapy in which a circular placental pattern exists, are not fully understood or known at this time.

**Are There Other Treatment Alternatives?**

Possible management alternatives other than laser surgery for twin-to-twin transfusion syndrome include:

1) **Expectant management.** In this option, the pregnancy would be followed with ultrasound examinations. In addition, you may be offered a medication to assist the fetuses’ hearts, or a medication to reduce the recipient fetus’ urinary output to decrease the excess amniotic fluid. Unfortunately, these medical options have not been successful in treating fetuses with twin-twin transfusion, and are associated with virtually 95% pregnancy loss.

2) **Amniocentesis.** (drainage of amniotic fluid from the sac of the recipient twin). The goal of this procedure is to decrease the pressure in the uterus by removing the excess fluid in the Recipient’s sac. This procedure may need to be repeated as often as necessary. The success rate of this treatment approach is approximately 66%, with an incidence of 15% of brain damage.

3) **Umbilical cord occlusion.** This procedure interrupts the flow of blood through the umbilical cord of one of the fetuses. This fetus dies and remains inside the uterus for the rest of the pregnancy. The other twin continues the pregnancy and should not be affected by the death of the first twin. In the State of Florida, Umbilical-cord occlusion can be performed if the pregnancy is less than 24 weeks. This procedure is reserved for patient’s in whom one of the fetuses is deemed nonviable.

4) **Interruption of the pregnancy.** You may choose this option up to 24 weeks of gestation in the state of Florida.

5) **Fetal septostomy.** (Needling of the dividing membrane). This procedure consists of purposely making a hole in the dividing membrane with a needle. This results in equalization of the fluid volumes between the sacs. Septostomy does not change the course of TTTS and disruption of the membranes may lead to death of the fetuses from cord entanglement. For these reasons we strongly advise against this procedure. If you have had this procedure purposely done by your doctors, you will not be a candidate for surgery at our institution.

**Welcome to UM/Jackson Memorial Hospital**

Ultrasound evaluations and fetal surgery are performed at University of Miami/Jackson Memorial Hospital. The purpose of this document is to familiarize you with the procedure you will follow when you arrive at JMH for evaluation and possible surgery.

**Referral Assessment**

If you wish to have a referral for possible surgery please contact the Fetal Therapy Coordinator at 305-585-6636. She will fax an evaluation form to your doctor to complete and return to us. This information will be needed to determine the best treatment. Our physicians will review your records. Your doctor’s office will be contacted with our recommendation.

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All ultrasound reports, prenatal records and insurance information will be requested. Our office will begin the insurance process. Our insurance clerk will notify you as soon as the insurance company informs her that this is/is not a covered benefit.

The hospital policy, at this time, requires total payment prior to surgery if your insurance company does not cover the procedure. Additional charges will be incurred from the anesthesiologist and pathologist and billed separately. If your insurance reimburses at a later date, your money will be refunded. Approximately 90% of the insurance companies are covering the surgery.

### Ultrasound Assessment

Ultrasound evaluations are performed at Jackson Memorial Hospital. Jackson Memorial Hospital is the University of Miami’s primary teaching hospital. On the day of your appointment, a Jackson Concierge representative will meet you at the hospital near the Diagnostic Treatment Center (DTC). The representative will take you to the fourth floor of Women’s Hospital Center, where you will meet the Fetal Therapy Coordinator. The coordinator will take you to get ultrasound assessments.

During your appointment, you will undergo diagnostic ultrasound and fetal therapy consultation. In addition, if you are scheduled for surgery, you will also undergo pre-surgical testing and lab work. Make sure to eat before your appointment as it takes a significant amount of time. Feel free to bring drinks and snacks with you. Plan to be at the hospital for approximately four hours.

Please arrive 45 minutes prior to appt time. Make sure to eat before your appointments as it takes a significant amount of time. Feel free to bring drinks and snacks with you.

Following the ultrasound the doctor will discuss all findings and will review the treatment, surgery, prognosis, and recommended follow-up care. All questions and concerns will be answered at this time.

The Fetal Therapy Coordinator will explain the surgery, answer any questions, and help you with any special needs. You will receive a folder that contains information you need for surgery and additional information you will find helpful. A surgery consent form will be given to you to review. It explains the surgery in terms you can understand. This is in addition to the hospital consent form. Both consent forms need to be signed prior to surgery.

### Pre-Admission Testing

Following your ultrasound and consultation, the fetal therapy coordinator takes you to the Admitting Office (if necessary) and following that you go to Pre-admission testing. Upon arrival, you complete a medical history form. This form asks about your general physical status and any specific health problems you may have. The nurse draws blood for various tests that are necessary prior to surgery. She gives you instructions on time to check in and where to check in for your surgery. One of the anesthesiologists evaluates your physical condition to determine any specific needs you may have concerning your anesthesia. This completes your visit to pre-admission testing department. After this is finished you do not have to come back to the hospital until two hours prior to surgery.

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Preparing for surgery: NPO Status

Before surgery, you are not 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. In medical terms, this is known as "NPO" (nothing by mouth).

It is important to drink as much water as possible before you are “NPO” so you do not become dehydrated.

Before Surgery

On the day of surgery, an intravenous line (IV) will be inserted by needle stick to give fluids and medications during surgery. In most cases, this will be the only needle stick that is needed. You will be held in the pre-anesthesia area (fourth floor labor and delivery area of Women's Hospital Center). After you are prepared for surgery, your husband or significant other may join you until you are taken to the operating room (OR). An ultrasound is done prior to going to the operating room to confirm the babies’ heartbeats.

The specially trained fetal therapy nurse and an anesthesiologist will take you to the OR.

After you are prepared for surgery, your husband or significant other may join you until you are taken to the operating room. An ultrasound is done prior to going to the operating room to confirm the babies’ heartbeats. One of the specially trained nurses that are assisting in surgery and the anesthesiologist accompanies you to surgery.

During Surgery

Surgery is performed under local anesthesia, meaning you are awake but relaxed and your abdomen is numbed where the instrument is inserted. An anesthesiologist stays with you throughout the procedure. You are given additional medication as needed. Occasionally, general anesthesia, meaning you are put to sleep, is used.

A catheter will be put in your bladder to drain urine and will remain in place until later that evening. During surgery, one or two small incisions, approximately 1/10 inch long, are made on the abdomen. In most cases surgery lasts one to two hours, but this can vary depending on a number of factors.

After Surgery

Following surgery, you are taken to the Recovery Room where you are monitored for approximately one - two hours. Medication may be given after surgery to relax the uterus and stop any contractions. Pain or discomfort after surgery is usually minimal. If needed, pain relief medicine is available.

The catheter is removed from your bladder about four hours after you arrive in your room. Your husband or other support person may remain with you in your room.

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Following surgery, you may have food as tolerated. That night, activity is restricted to bathroom privileges only, but this depends upon your specific condition.

The day after surgery, a follow-up ultrasound will be done in the morning. Most patients are discharged following their ultrasound.

You may plan on being here approximately 3-4 days: one day for ultrasound and consultation and one day for surgery. You spend the night of surgery in the hospital. Most of our patients are discharged the following morning. We recommend spending one night at the hotel before traveling.

**Returning Home**

After surgery, your personal doctor resumes your care for the rest of the pregnancy and delivery. You and your physician 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 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 ultrasound reports and any other pertinent information to us.

**Transportation and Housing**

Patient rates are available at hotels near Jackson Memorial Hospital. Hotel rates and transportation information can be found at:

http://www.uhealthsystem.com/patient_services/international.asp

**References**


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