

Information for Parents When Twins Share One Placenta

Monochorionic Twins

Part 1

THE MULTIPLE BIRTHS FOUNDATION



Supporting Professionals Supporting Families

Compiled by

| | |
|----------------------------|--|
| Margie Davies | Midwifery Advisor Multiple Births Foundation |
| Jane Denton | Director Multiple Births Foundation |
| Helga Everard | Specialist Multiple Births Midwife. Queen Charlotte's & Chelsea Hospital |
| Mark Kilby | Professor of Fetal Medicine, Birmingham Women's Foundation Trust |
| Ruwan Wimalasundera | BS Consultant Obstetrician & Fetal Medicine Imperial College Healthcare NHS Trust |

Published by

**The Multiple Births Foundation
Level 4 Hammersmith House
Queen Charlotte's & Chelsea Hospital
Du Cane Road
London
W12 0HS**

Tel: 020 8383 3519

E-mail: mbf@imperial.nhs.uk

Website: www.multiplebirths.org.uk

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Monochorionic Multiple Pregnancies

Part 1

Introduction

Discovering you are pregnant with twins or triplets can come as quite a shock. The excitement of having twins may be combined with anxiety about the pregnancy particularly, if you are told that your babies are sharing one placenta and you start to read more about the risks.

At the Multiple Births Foundation we receive many queries from parents seeking accurate information about the management and care of a women with a monochorionic pregnancy. We hope this booklet helps to answer your questions with factual medical information to help you understand why monochorionic multiple pregnancies differ from other pregnancies and the reasons for increased obstetric/midwifery surveillance as well as suggested treatments.

It is important to remember that every case is different and as such the care and treatment YOU will receive will be tailored to your individual needs only.

You can contact the Multiple Births Foundation at any time if you have any queries e-mail: mbf@imperial.nhs.uk telephone: 020 8383 3519.

The booklet is in two sections: the first part explains about monochorionic twins generally and second part is about the complications which may arise and the possible treatments.

Types of placenta in a twin pregnancy

- a single (monochorionic) placenta.
- 2 separate (dichorionic) placenta .

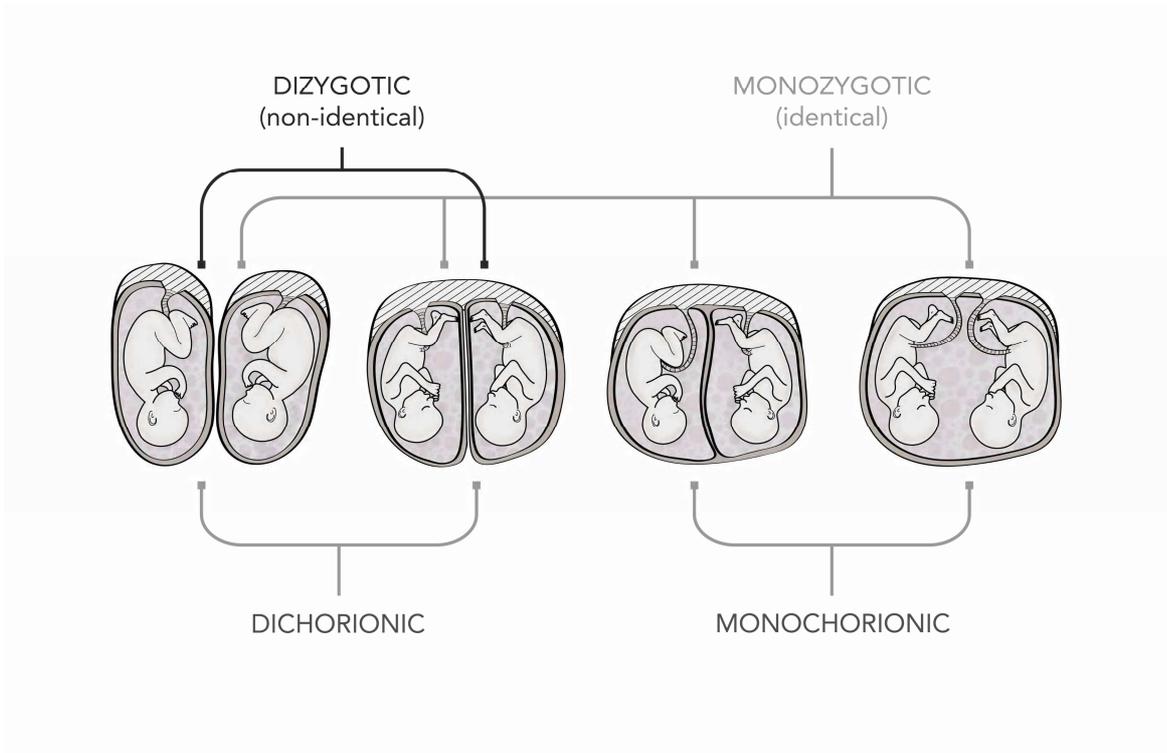
In **monochorionic** twins there is **one chorion** around both babies which means they share one placenta and the term **monochorionic** is used. Triplets and Quads can also be monochorionic. The inner membrane is known as the **amnion**. If the babies each have their own **amnion** the pregnancy will be referred to as **monochorionic diamniotic (MCDA)**. These babies are always identical.

In about 1% of **monochorionic** twin pregnancies, the babies will also share one inner membrane (**amnion**). These babies will be referred to as **monchorionic monamniotic (MCMA)**, and are always identical.

In **dichorionic** twins each baby will have its own placenta, with its own **amnion** and **chorion**. For these twins there will be **two chorions** and **two amnions** the term **dichorionic diamniotic (DCDA)** is used. If triplets each have their own placenta there will

be **three chorions** and **three chorions** they will be referred to as **trichorionic triamniotic (TCTA)**.

Approximately one third of monozygotic (identical) twins will each have their own placenta and the appearance will be exactly the same on ultrasound scans as the dichorionic DCDA dizygotic (non-identical) twins. See *diagrams*



The differences between chorionicity and zygosity

Chorionicity; is determined during pregnancy (ideally before 13 weeks) and is assessed by looking at the membranes as described above.

Zygosity; is determined usually after the babies are born, either because they are of different sex, by DNA testing, or is obvious as the babies look very different.

This booklet will look at Monochorionic twin pregnancies and their care.

Monochorionic pregnancies are classed as high risk because this type of 'twinning' is associated with increased risks of:

- **Fetal Growth Restriction**

Although in some monochorionic pregnancies the placenta is divided equally between the twins, in approximately 40% of cases one baby has a disproportionately smaller share of placenta which may result in that baby not growing as well. This is known as intrauterine growth restriction (IUGR). In most pregnancies complicated in this way one twin is small and the other grows 'normally,' this is known as discordant growth, but in some pregnancies both twins can be small.

- **Twin-to- Twin Transfusion Syndrome (TTTS)**

In 10-15% of monochorionic twin pregnancies a condition known as twin-to-twin transfusion syndrome (TTTS) develops. This is caused by abnormal blood vessels in the placenta connecting the two twin circulations. Instead of having a balanced flow of blood from one baby to the other, there is an unbalanced flow. So that one baby, the “donor”, receives too little blood and has a low blood pressure – and the other baby, the “recipient” receives too much blood and has a high blood pressure.

Management of a Monochorionic Pregnancy

The Royal College of Obstetricians & Gynaecologists (RCOG) has guidance on the management of monochorionic twins and your care is likely to be as follows:

1. Your first pregnancy scan is usually performed between 10 -14 weeks though in some cases it may be earlier (i.e. if you have had treatment for subfertility). If there are two or more babies, this is the time that it is easiest to assess the chorionicity and record the findings in your notes. If you choose to have the nuchal translucency measurement taken, (which is a screening test for Down’s syndrome), it will also be determined at this stage.

2. If your pregnancy is monochorionic, you will be monitored closely with regular ultrasound scans every 2 - 3 weeks from 16 weeks to 24 weeks gestation, to check the growth of the babies and for signs of TTTS. This is done by looking at amniotic fluid volumes and blood flow between the twins, you may be referred to your regional fetal medicine centre for this or can be seen at your local hospital if they have a protocol agreed with the regional centre. If there are any concerns these will be reported to the regional fetal medicine centre for the advice of the fetal medicine doctors. A detailed scan looking at the development of the babies (anomaly scan) is performed between 18 – 22 weeks. This includes a scan that looks at the structure of the babies’ hearts, as monochorionic twins are at greater risk of the heart abnormalities.

3. After 24 weeks of gestation, ultrasound scans will be performed to monitor fetal growth, know as umbilical artery Doppler studies. These will be at intervals of 2 to 4 weeks depending on your personal situation.

4. Between 32 and 34 weeks there should be a discussion with your consultant’s team regarding how the babies will be delivered. The decision is based on your individual circumstances; if the pregnancy has been uncomplicated and both babies are presenting head first a vaginal delivery may be an option. If there has been problems of any kind or the babies are presenting by the bottom (breech). Then a caesarean section is the usual choice of delivery.

5. If all is well, the delivery should be prior to 37 weeks gestation, by which ever method has been agreed.

6. If it looks as though your babies are going to be born prematurely, it is advisable to be seen by the paediatricians and have a visit to the neonatal unit (NNU). See *Bliss information*

What is Twin-to-Twin Transfusion Syndrome? (TTTS)

This most commonly affects monochorionic twins but is also known as fetto-fetal transfusion syndrome as it may affect triplets or more.

Although each baby has its own share of placenta, these two babies are “connected” by several types of blood vessels in the placenta. Where the blood vessels join together is known as an “anastomosis”. These links may be between two arteries (arterio-arterial) or two veins (veno-venous) so blood can flow equally in either direction to either baby. There can also be artery to vein (arterio-venous) anastomoses, which allows blood to flow only in one direction – from artery to vein. These are normally balanced by the other types of anastomoses so that usually an equal amount of blood flows from one baby to the other.

In 10-15% of monochorionic twin pregnancies a condition known as twin-to-twin transfusion (TTTS) develops. Instead of having a balanced flow of blood from one baby to the other, there is an unbalanced flow so that one baby receives too little blood (and is hypotensive) and is known as the **donor** twin. The other baby receives a greater amount of blood (and is hypertensive) and is known as the **recipient** twin. This causes the donor twin to have a low perfusion/blood pressure and on ultrasound scans there are features of low urine production (poorly visualised bladder and reduced amniotic fluid volume) as well as signs of poor blood flow within the placental circulation. In contrast, the recipient twin receives too much blood and is hypertensive, with a large blood flow through the baby’s kidneys causing the bladder to look enlarged on ultrasound and also the presence of significantly increased amniotic fluid volume (polyhydramnios) around this twin. In addition, this twin’s heart has to work harder to cope with the extra blood supply, causing the heart to enlarge.

What to look out for; The first sign that there may be a problem is when the mother complains that her abdomen (tummy) has suddenly (sometimes within 24 hours) become much larger and feels tight and uncomfortable all the time, she may also have noticed she has an increased thirst. This may be difficult for parents to be sure about as you are carrying twins and expect to be larger than in a singleton pregnancy! If you are not sure contact your hospital straight away for a check up. This is when the recipient twin has too much fluid around him/her and it is called polyhydramnios. The donor twin will have very little fluid, called oligohydramnios, and can also be referred to as a “stuck twin”; since this twin becomes stuck at the edge of the uterus. If TTTS goes undetected premature labour can occur. The woman should see a fetal medicine specialist as soon as possible if this situation should arise, so that specific treatment can be discussed and if appropriate started.

Monochorionic Monoamniotic Twins (MCMA)

This type of pregnancy is very rare, occurring in approx 1% of monochorionic twin pregnancies which is about one in 40,000 pregnancies. The complication here is that these twins not only share a placenta and a chorion but they are both in the same amniotic sac with no membrane dividing them so there is an added risk of the two cords becoming entangled.

Information for Parents When Twins Share One Placenta

Monochorionic Twins

**Twin-to-Twin Transfusion Syndrome (TTTS)
- explained in more detail**

Part 2

(This follows on from part 1 - please read that first)

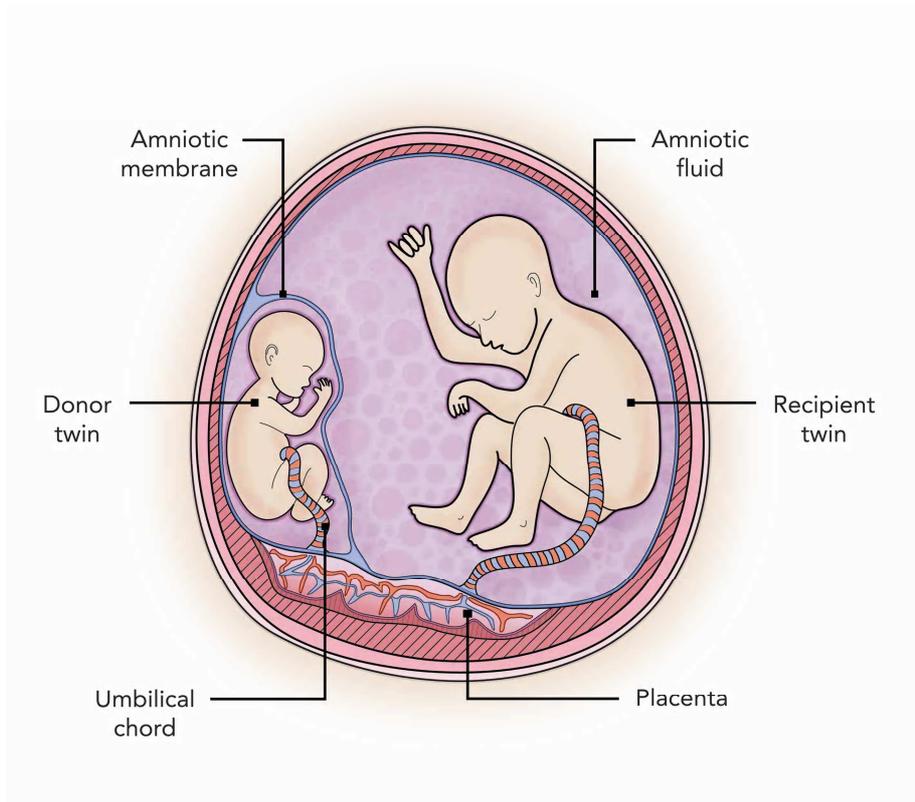
Monochorionic Multiple Pregnancies

Part 2

What is Twin-to-Twin Transfusion Syndrome?

Although each baby has its own share of placenta, these two babies are “connected” by several types of blood vessels in the placenta, so that there is a single placental circulation. Where the blood vessels join together is known as an “anastomosis”. These links may be between two arteries (arterio-arterial) or two veins (veno-venous) so blood can flow in either direction to either baby. There can also be artery to vein (arterio-venous) anastomoses, which allows blood to flow only in one direction – from artery to vein. These are normally balanced by the other types of anastomoses so that usually an equal amount of blood flows from one baby to the other.

In 10-15% of monochorionic twin pregnancies a condition known as twin-to-twin transfusion (TTTS) develops. Instead of having a balanced flow of blood from one baby to the other, there is an unbalanced flow so that one baby receives too little blood (and is hypotensive) and is known as the “**donor**”. The other baby receives a greater amount of blood (and is hypertensive) and is known as the “**recipient**”. This causes the donor twin to have a low perfusion/blood pressure and on ultrasound scans there are features of low urine production (poorly visualised bladder and reduced amniotic fluid volume) as well as signs of poor blood flow within the placental circulation as seen in Doppler scans. In contrast, the recipient twin receives too much blood and is hypertensive, with a large blood flow through the baby’s kidneys causing the bladder to look enlarged on ultrasound and also the presence of significantly increased amniotic fluid volume (polyhydramnios) around this twin. In addition, this twin’s heart has to work harder to cope with the extra blood supply and can become enlarged.



How will the babies be affected by Twin-to-Twin Transfusion Syndrome?

In milder cases the babies will be born healthy but there may be quite a difference in size and the donor twin may be anaemic and very pale. The recipient twin will be larger and may look very red at birth this is because they have too much blood (polycythaemia) which can lead to jaundice during the first week of life, because the baby's haemoglobin is high and the liver has difficulty in coping with the increased number of red blood cells. This is usually due to a 'transfusion' of blood between the twins either shortly before or during the delivery process.

In more severe cases of TTTS, the donor twin, who has a reduced blood flow, and usually a smaller proportion of the placenta supporting it. The blood flow to the baby's kidneys will also be down due to this less urine will be produced, less urine means reduced amniotic fluid (oligohydramnios). Eventually this donor baby will become wrapped within its own membranes with no fluid ("stuck twin").

The recipient baby, who has an increased blood flow, will produce large amounts of urine and therefore too much amniotic fluid (polyhydramnios). This also affects the way the fetal heart works as a 'pump' and eventual this baby may go into heart failure being unable to cope with the extra blood volume.

Overall, if severe twin-to-twin transfusion syndrome (TTTS) is left untreated the babies chances of surviving are poor – less than 10%. If TTTS is picked up early enough treatment can be given to help prolong the pregnancy until it reaches a viable gestation, giving the babies a chance of survival. However, in all but the mildest forms there is an increased risk of handicap in survivors.

The different stages of TTTS - the degree of severity.

It is important to realise that moving from one stage to the next can happen very quickly in some cases, though not all, and women should be aware if their abdomen (tummy) suddenly feels as though it has expanded and feels very tight all the time, (sometimes over night) then you should contact your hospital immediately. You may have an increased thirst as well.

- Stage I: The mildest form of TTTS with a significant difference in amniotic fluid volumes in the two sacs.
- Stage II: The donor now has no urine visible in its bladder.
- Stage III: Either the donor twin or recipient has abnormal blood flow patterns shown by the scans which assess the blood flow (Doppler scans)
- Stage IV: This is characterised by the development of heart failure or fetal hydrops in the recipient.
- Stage V: This is when one or both babies have died.

Although having TTTS can be a worrying and frightening experience, it is well known, and treated every day by specialist doctors who understand the condition. Once you are in the TTTS world, as it were, you will meet many other people whose lives are spent dealing with it and parents whose babies have had it. They will be there for you, giving you advice and support in difficult and dangerous situations, but there are solutions and you will be told exactly what your options are and what is best for you and your babies.

Just remember all cases are individual and what's right for one woman may not be for another, so the treatment for each of you will be different.

Treatment for TTTS

The treatment for TTTS depends to a large extent on the severity or stage of the TTTS, the treatment options available depend on this and the length of the pregnancy (gestation).

Less than 26 weeks of pregnancy

- **Mild TTTS**

With mild TTTS (stage I), in approximately a half to two thirds of cases the condition will remain stable or improve by itself. These pregnancies will need to be monitored very closely. If they remain stable but the amniotic fluid is excessive then taking off the excess fluid by a procedure known as amnio-reduction may be helpful. If the TTTS becomes progressive, laser ablation of the placental vessels would be recommended (see below).

In about one third of cases it will progress to severe TTTS.

Treatment is individualised in such cases.

- **Severe TTTS**

The following treatment should be considered:

In cases of severe TTTS, fetoscopic laser ablation is the treatment of choice (at gestational age of up to 26 completed weeks)(Cochrane review,2008).

Fetoscopic Laser Ablation Therapy.

This involves the insertion of a small telescope (2-3mm) know as a fetoscope into the uterus under ultrasound guidance which is usually carried out under local or regional anaesthetic. The blood vessels on the surface of the placenta joining the twins together can then be identified and occluded using the laser. By occluding all the anastomotic blood vessels the transfusion of blood from one baby to the other should be prevented.

This is the best treatment currently available and both babies survive in 60 – 70% of cases but only one baby may survive in 10 – 15% of cases. When TTTS is severe, both babies may die in approximately 20 – 25% of cases despite having treatment using the laser.

Some hospitals may coagulate all anastomoses where others only do selective ablation that means only the vessels where blood is going from the 'donor' twin to the 'recipient' twin are separated.

Selective Feticide

If the laser therapy has not cured the TTTS or the TTTS occurs at a very early stage of pregnancy (before 16 weeks) when a fetoscopic laser can not be performed, the fetal medicine specialist may suggest an option known as selective feticide. This would only be done as a last resort if it was felt that the risk of both babies dying without any further treatment was high. This procedure would involve occluding the blood flow in one baby's cord (which ever baby was most sick). This would mean that this baby dies in the womb, but would allow the other baby a good chance of survival. These procedures can only be performed in specialised centres. This will be discussed in detail with you if the need arises.

Later than 26 weeks of pregnancy

If TTTS occurs after 26 weeks of pregnancy and both babies have reached a viable weight then either amniodrainage or delivery would be an appropriate option. This would be discussed on an individual basis.

What is the long-term outcome for the babies with TTTS?

Most babies grow up to be perfectly healthy. Sometimes, although both children may be healthy with normal development, the donor twin remains smaller.

In severe cases the babies may not survive, because they are delivered too prematurely or because they go into severe heart failure before they are born.

In some treated (or untreated) cases the babies may have some long-term problems, either because they have suffered from complications as a result of being born prematurely, or because their heart, brain or kidneys have been damaged inside the womb.

Cerebral palsy is seen in about 7% of surviving babies treated by fetoscopic laser ablation.

All babies who have had TTTS will be carefully examined by a paediatrician after birth and will continue to be seen regularly until the doctors and parents are completely happy with their mental and physical development.

Monochorionic Monoamniotic Twins (MCMA)

This type of pregnancy is very rare, occurring in approx 1% of monochorionic twin pregnancies which is about one in 40,000 pregnancies. The complication here is that these twins not only share a placenta and chorion but they are both in the same amniotic sac with no membrane dividing them so there is an added risk of the two cords becoming entangled.

The highest risk of babies dying from cord entanglement is before 20 weeks gestation but it can happen at any stage of pregnancy. This is usually as a result of tightening of an already entangled umbilical cord occluding the baby's blood supply, known as a cord accident. If a cord accident does occur it tends to be a sudden phenomenon which cannot be predicted beforehand. Being admitted to hospital for regular monitoring of the babies does not seem to improve the outcome. However regular scans every two weeks to monitor growth and blood flow (Doppler scans) should be performed from 20 weeks gestation.

As cord accidents can happen at any stage and are unpredictable, delivery is recommended usually between 32-34 weeks gestation (after a course of maternal steroids). The exact timing of delivery is individual and should be discussed with your consultant team. Once the babies have reached this stage the advances in neonatal care give them a very good chance of survival. As cord accidents are highly likely to occur during vaginal delivery, it is usual to deliver the babies by caesarean section.

Because of the rarity of this condition, there is limited evidence about the best care which is why it is advisable for these pregnancies to be monitored closely by specialists with some experience of MCMA pregnancies.

Some centres give the mother a drug known as sulindac which reduces the amount of urine the babies produce, and so reduces the amniotic fluid, which then gives the babies less room to move. In theory this *reduces* the risks of cord accidents. However using sulindac is not universal practice and again its use should be discussed on an individual basis.

Glossary

Amnion: Inner lining of sac containing the developing fetus.

Anaemia: Low level of haemoglobin in the blood.

Anastomosis: Cross connection between arteries and veins.

Chorion: Outer lining of sac containing the fetus.

Dichorionic twins: Two babies who have developed in separate chorionic sacs.

Dizygotic: Formed from two separate zygotes.

Doppler: Blood flow scan

Fetofetal Transfusion Syndrome: (usually known as TTTS): Condition in which blood from one identical twin fetus transfuses into the other via blood vessels in the placenta.

Fraternal twins: See Dizygotic.

Hypertension: high blood pressure

Hypotension: low blood pressure

Hydrops: Severe heart failure and accumulation of fluid in the fetus.

Identical twins: See Monozygotic.

Monoamniotic twins: Two babies who have developed in the same amniotic sac.

Monochorionic twins: Twin babies who have developed in the same chorionic sac but not necessarily the same amniotic sac.

Monozygotic: Formed from one fertilised zygote.

Occlude: To stop or close something.

Oligohydramnios: reduced amniotic fluid.

Polycythaemia: Increased level of haemoglobin in the blood.

Selective fetocide: termination of affected fetus.

Stuck twin: When donor twin has no amniotic fluid in amniotic sac.

Trimester: Three months/one third of pregnancy:

| | |
|--------|-------|
| First | 0-14 |
| Second | 14-28 |
| Third | 29-40 |

Trizygotic: Formed from three separate zygotes.

Twin-to-Transfusion syndrome: See Fetofetal transfusion syndrome.

Zygoty: Describes the genetic make-up of children from a multiple birth.

Zygote: Fertilised ovum.