Chapter

Recent Updates in the Management of Monochorionic Twin Pregnancy

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Abstract

Monochorionic pregnancies are at high risk of developing severe complications leading to high perinatal morbidity and mortality. About 15% of these twins have unidirectional anastomosis of the placenta, which is responsible for the major complications specific to monochorionic pregnancies. An important first step in the management is the identification of the chorionicity. Once it is identified, a close follow-up every 2 weeks is vital to allow early detection of complications and their management. Approximately 1 in 10 monochorionic pregnancies develops twin-to-twin transfusion syndrome, congenital anomalies, anaemia polycythaemia sequence, selective intrauterine growth restriction and intrauterine death of a co-twin. Rare complications that can occur are twin reversed arterial perfusion syndrome. Timely screening and detection of all such complications can lead to appropriate intervention such as in utero foetoscopic laser treatment. These interventions can increase the survival rate of at least one or both twins with reduced neonatal morbidity. Besides, early detection can facilitate parents to have an informed choice to decide if the prognosis of the pregnancy is otherwise not good.

Keywords: monochorionic, updates, management, twin to twin transfusion, selective growth restriction, twin anaemia polycythaemia

1. Introduction

The incidence of twin pregnancy in the United States in recent times is approximately 3% [1]. With advancing age, different ethnic populations and advanced use of assisted reproduction technology, the incidence of dizygotic twins is far more common and accounts for 70% of all twin gestations. However, the incidence of monozygotic twins remains mostly constant worldwide and accounts for 3–5 per thousand births. The incidence of monochorionic twins is 1 in 300 pregnancies. In about 15% of these twins, there is an imbalance in foetal circulation. This results in conditions like twin-to-twin transfusion syndrome (TTTS), twin anaemia polycythaemia syndrome (TAPS), twin reversed arterial perfusion syndrome (TRAP), selective intrauterine growth restriction (sIUGR) and death of a co-twin.

In this chapter, we discuss the latest updates in the management of monochorionic twin pregnancy.

2. Chronicity and twinning

The most essential component of foetal well-being in twin pregnancy is the determination of the placental chronicity. Placental physiology has a vital impact on foetal and neonatal outcomes. Monozygotic twins develop when a single sperm fertilises with a single ovum during conception. Post conception, if the splitting of the egg occurs 2–3 days post fertilisation, it results in dichorionic, diamniotic twins. Approximately 30% of monozygotic twins are diamniotic and dichorionic. A splitting of egg 3–8 days post-conception results in monochorionic and diamniotic twins. About 70% of monozygotic twins are monochorionic diamniotic. If splitting occurs 9–12 days after fertilisation, it results in monoamniotic monochorionic twins. The

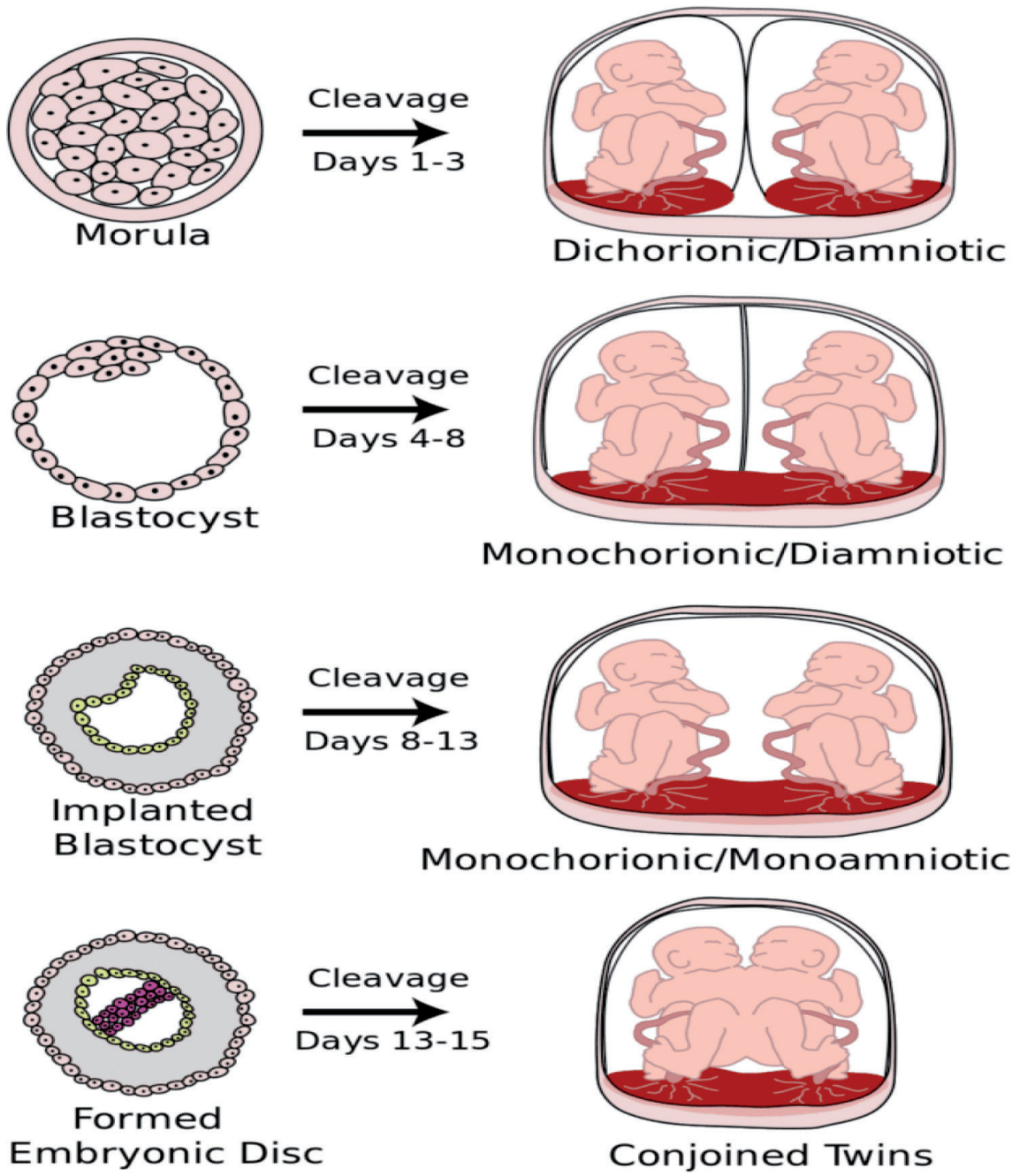

Figure 1.
Courtesy—Google images.

incidence of these twins is only 1%. There is a well-documented increased incidence of second-trimester loss, congenital anomalies, and prematurity in these twins. A splitting after 12 days fertilisation can result in conjoined twins (**Figure 1**).

As a result of a single placenta, monochorionic twins have substantial vascular communications between the two foetal circulations. In 80% of cases, the vascular anastomosis is bidirectional, which rarely leads to a haemodynamic imbalance between foetal circulations. However, it allows a direct vascular connection between the twins with an increased risk of foetal death [2, 3].

In 15% of monochorionic pregnancies, the placenta has have a predominance of unidirectional vascular anastomosis which results in twin-to-twin transfusion syndrome (TTTS) [2]. Other morbidities exclusive to monochorionic pregnancies are:

- Intrauterine growth restriction (sIUGR)
- Twin anaemia polycythaemia sequence (TAPS)
- Neurodevelopmental morbidity
- Trap reversal arterial perfusion syndrome (TRAP)
- The death of a single twin and its effects on the second twin

3. The role of ultrasound in determining chronicity and amnionicity

It is vital that all women with twin pregnancies should be offered an ultrasound examination between 11 + 0 and 13 + 6 weeks of gestation (crown-rump length 45–84 mm [2]. This is crucial to assess foetal viability, gestational age and chronicity. In monochorionic diamniotic pregnancies, the intertwin membrane becomes progressively thin after 9 weeks. A characteristic 'T' sign is seen on ultrasound with a 100% sensitivity and greater than 98% specificity for detecting monochorionic diamniotic gestation [4]. On the other hand, in dichorionic diamniotic pregnancies, a 'twin peak' or lambda sign is characteristic with a sensitivity greater than 97% and specificity of 100% in predicting chronicity [4]. It is a good practice to determine the amnionicity at the same time and document it as well (**Figure 2**).

Other sonographic signs to determine chronicity, especially when women present after 14 weeks of gestation, include number of placental masses, number of

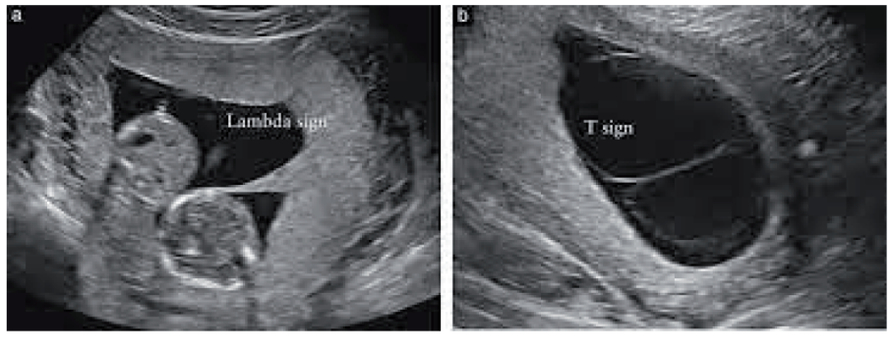

Figure 2.
Courtesy: (obgyn.onlinelibrary.wiley.com).

gestational sacs and concordant sex of the foetuses but the most valuable sign is the intertwin membrane using 2-dimensional ultrasound which is considered as highly accurate with very high sensitivity and specificity. The reliability of the number of placental masses may become arguable, as it is not unusual for dichorionic placentae to be commonly adjacent to each other and give appearance of a single mass. It is also noted that 3% of monochorionic twin pregnancies may have two placental masses on ultrasound, the presence of which does not prevent the presence of vascular anastomoses. It is likely that using a combination of ultrasound features, rather than one, would be more accurate [5].

4. The clinical implications of increased nuchal translucency in 11–13 weeks in twin pregnancy based on chronicity

Several studies have reported a comparison of increased nuchal translucency versus normal nuchal translucency in both dichorionic and monochorionic twins. However, a recent study has demonstrated that monochorionic twin pregnancies with increased nuchal translucency, but no chromosomal abnormalities had a higher incidence of structural anomalies and twin-specific complications. The most likely cause of increased nuchal translucency in these twins could be because of their unique vascular anastomoses which can lead to conditions like cardiac dysfunction, anaemia and possibly twin-to-twin transfusion syndrome. The studies indicated that detection of increased nuchal translucency is important in the early prediction of twin-specific complications although the appropriate intervention in such cases remains controversial [6]. Similarly, a discordance in nuchal translucency measurements of more than 20% is often seen in those cases of monochorionic diamniotic twin pregnancies that develop early twin to twin transfusion than those with normal outcomes [7]. The risk of TTTS and early intrauterine death in such cases is up to 30%. Such cases should be discussed with the foetal medicine expert and should be offered a detailed ultrasound and karyotyping [5].

As for the chromosomal screening, it is recommended in monochorionic twins when aneuploidy sceening is offered nuchal translucency should be used in conjunction with first-trimester serum markers (combined screening test) at 11 + 0 weeks to 13 + 6 weeks of gestation (crown–rump length 45–84 mm). Studies have shown that although non-invasive prenatal testing has a high screening efficiency in a singleton pregnancy, its performance in twin pregnancies remains unstable. Hence, it should be carefully used in the screening of chromosomal abnormalities in twins [8].

As it is well recognised that screening and diagnostic testing in twins are more complex than in singleton pregnancy, hence comprehensive counselling should be provided by health care experts before tests are taken. Health care providers should inform couples regarding the complex decisions which may be potentially required especially in cases of monochorionic twin pregnancy.

With regards to performing amniocentesis in monochorionic twins, if monochorionicity has been confirmed before 14 weeks of gestation and there is no gross discordant in growth of foetuses, it is acceptable to sample only one amniotic sac. Otherwise, both amniotic sacs need to be sampled as the rare possibility of discordant chromosomal anomalies in monochorionic pregnancy cannot be ruled out. Chorionic villus sampling (CVS) in monochorionic pregnancy will sample only the single placenta. This could miss the rare discordant chromosomal anomalies. Discordance for most of the common human aneuploidies (trisomy's 13, 18 and 21, Turner syndrome

and triploidy) has been reported in monochorionic twin pairs. In the event of such a case, a selective reduction by umbilical cord occlusion can be offered from 16 weeks onwards, with a survival rate of more than 80% for the healthy twin. It is extremely important to provide detailed counselling in such complex cases to the parents by foetal medicine experts [5].

5. Dating of the twins

It is recommended that the pregnancy should be dated according to the crown-rump length of the larger twin. Dating should take place when the crown-rump length is between 45 mm and 84 mm (equivalent to 11 + 0 to 13 + 6 weeks of gestation). Twins, if seen after 14 weeks, should be dated according to the head circumference of the larger twin [9].

6. Labelling of the twins

Labelling of twins should follow a reliable and steady strategy with options according to their site; for example right or left, upper or lower or according to the insertion of cord in relation to placental membrane insertion as recorded during first trimester scans. If there is a discordance in the twins, a description such as 'potential recipient' can be used for labelling. It is important to note that twins as seen on ultrasound may not be delivered in the same sequence especially during a caesarean section. Thus, it is strongly recommended to rescan the women just before performing a caesarean especially in cases of twin congenital abnormality like cardiac defects or diaphragmatic hernia and before any surgical intervention [5].

7. The antenatal care of women with monochorionic diamniotic twin pregnancy

It is recommended that monochorionic diamniotic pregnancy should have hospital-based care. If any complications arise, then tertiary care should be advised. Women with monochorionic diamniotic twin pregnancy require a good and strong emotional support throughout the pregnancy bearing in mind the foetal morbidity and mortality associated with the twins. All efforts should be made to reduce their anxiety, and comprehensive counselling should be done at the booking visit. The screening and diagnostic tests and their complexity should also be discussed at the initial visit.

General advice regarding diet and lifestyle should be given [10]. It should be emphasised that women with twin pregnancies are more prone to anaemia, pre-eclampsia, gestational diabetes, varicose veins as well as increased risk of venous thromboembolism, antepartum and postpartum haemorrhage. All usual antenatal screening blood tests should be advised as singleton pregnancy. If anaemia is detected, iron should be commenced in early pregnancy along with folic acid. Vitamin D should also be started from early pregnancy if a woman is deficient in vitamin D.

A prophylaxis dose of Aspirin 100–150 mg is recommended from 12 weeks to 36 weeks of pregnancy if other risk factors are present such as:

Previous history of hypertensive disorders in pregnancy; chronic hypertension in pregnancy; chronic kidney disease; history of Type 1 or type 2 diabetes; primigravida; history of autoimmune conditions such as antiphospholipid syndrome or systemic lupus erythematosus; age greater than 40 years; family history of pre-eclampsia or obesity greater than 35 at first visit [10].

The antenatal and ultrasound visit schedule for monochorionic diamniotic twins (**Figure 3**).

Most recent studies suggest a structured ultrasound schedule for the antenatal management of monochorionic twin pregnancy. It is now generally accepted that after the first visit and ultrasound at 12 weeks, monochorionic pregnancies should have antenatal visits every 2 weeks from 16 weeks onwards. At every visit, an ultrasound should be performed for the early detection of twin transfusion syndrome and twin anaemia polycythaemia syndrome, as early detection of these conditions results in a better perinatal outcome.

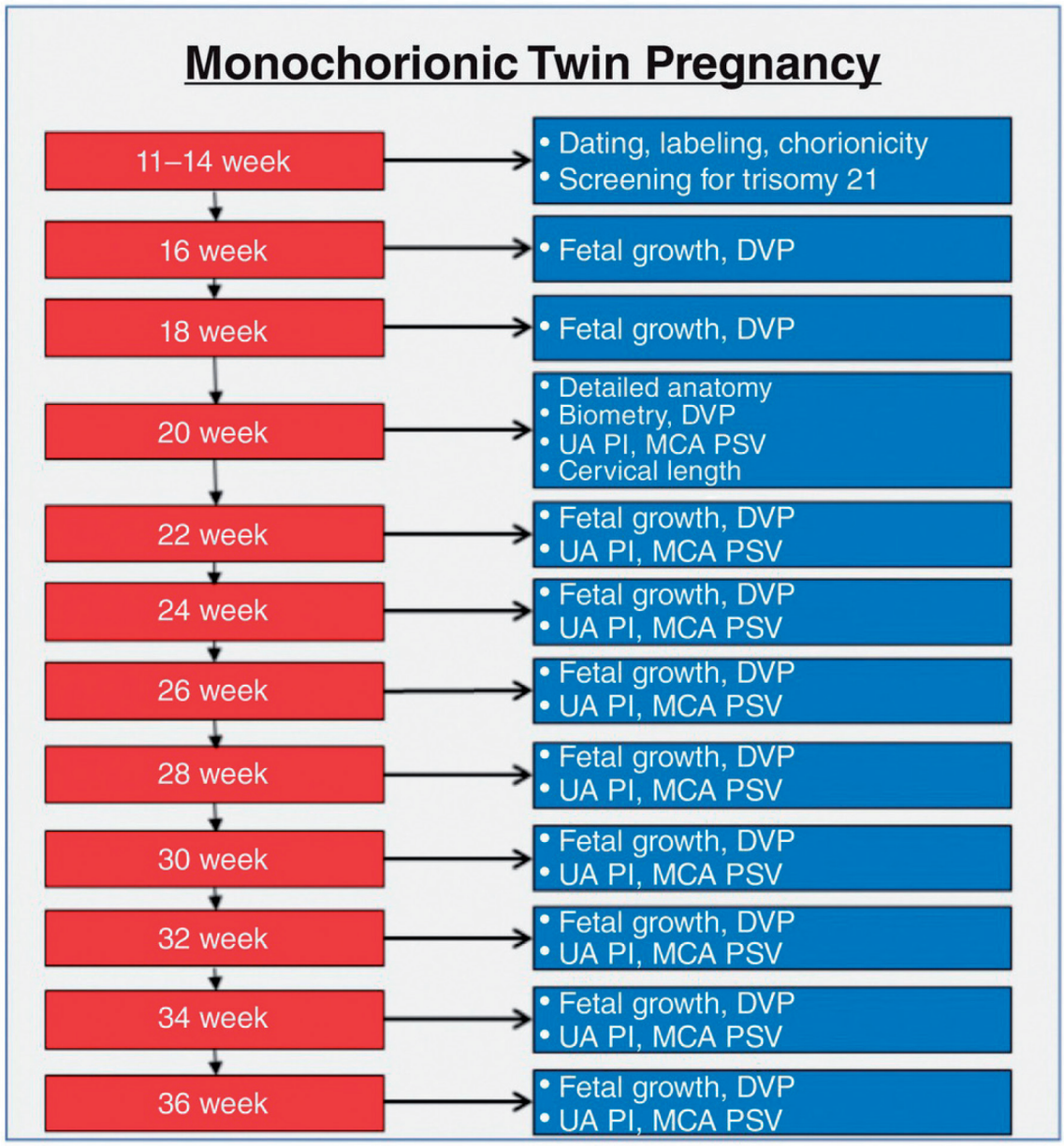

Figure 3.
Reproduced from Google image (Cambridge University Press).

At 16 and 18 weeks scan, it is recommended to measure foetal biometry and deepest vertical pocket (DVP) of amniotic fluid in each twin. At 20 weeks, a detailed anomaly scan should be performed along with biometry. The umbilical artery Doppler pulsatile index (UA PI) should also be recorded from this visit onwards along with the middle cerebral artery peak systolic velocity (MCA PSV). The MCA PSV helps in the early diagnosis of twin anaemia polycythaemia syndrome (TAPS) and is recommended to be done routinely from 20 weeks onwards [5]. Amniotic fluid measurements should be continued at this visit and a cervical length screening is also recommended at this stage. Antenatal visits should continue every 2 weeks until 36 weeks. At each visit, foetal biometry, umbilical artery Dopplers, middle cerebral artery peak systolic velocity and deepest vertical pocket of amniotic fluid in each sac should continue. A decision on delivery should be taken at 36 weeks.

8. Screening for structural anomalies

Twin foetuses should be assessed for the presence of any major anomalies at the first-trimester scan, and a routine second-trimester (anomaly) scan should be performed at around 20 (18–22) weeks of gestation as in singleton pregnancy.

Apart from this, a cardiac screening assessment should be performed in monochorionic twins. The cardiac scan should be performed according to standard ultrasound guidelines including laterality, situs and four-chamber, ventricular outflow tract and aortic arch. The abnormalities of the brain and cardiac abnormalities are more common in monochorionic twins. Other abnormalities associated with twins include neural tube defects, anterior abdominal wall defects, facial clefts, brain abnormalities, cardiac defects and gastrointestinal anomalies. Every 1 in 15 monochorionic pregnancies may have a risk of a major congenital anomaly. By doing a regular screening, the parents get a chance to prepare for the birth of a baby with a potential problem, offering them the option of termination. This also allows the transfer of the pregnancy to a tertiary care with special intervention facilities [5].

9. Screening for preterm birth

Cervical length assessment should be considered the optimal method of screening for preterm birth in a monochorionic pregnancy. A transvaginal scan visualises the cervix more objectively and the 2D modality is considered the most appropriate imaging modality.

The ratio of a curved/straight cervix decreases with a decrease in length, and this does not have important clinical implications [11]. Serial measurements should be done, and the shortest result should be taken. A short cervical length is a good predictor of preterm births even in later gestations. It is considered more accurate than digital exams and foetal fibronectin in the prediction of preterm birth (**Figure 4**) [12].

A cervical length of <20 mm at 20–24 weeks is the most accurate predictor of preterm labour with high sensitivity and specificity [12]. A higher rate of preterm birth is common in monochorionic twins and could be reflected and predicted by an increased rate of a short cervix.

The use of progesterone, bed rest, Arabin cervical pessary, antibiotics or oral tocolytics has not shown to reduce the risk of preterm labour in these cases. The use of vaginal progesterone in twin pregnancy with a TVS cervical length of <20–25 mm has

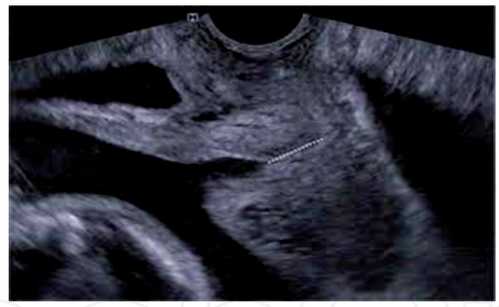

Figure 4.
Twin pregnancy with a short cervix. Courtesy: (mfmync.com).

been shown to reduce the incidence of preterm birth at <34 and <32 weeks in some studies [13]. However, the results were not conclusive. Progesterone may reduce the risk of neonatal morbidity and mortality [5].

The rates of preterm birth at <24 weeks, <28 weeks, <32 weeks and < 34 weeks have shown to be reduced by placement of a first-trimester cervical cerclage in twins only with a previous history of preterm birth [13].

A course of antenatal corticosteroids may reduce the risk of respiratory morbidity, necrotising enterocolitis and intraventricular haemorrhage but this should be timed and not given untargeted [14].

10. Screening and management of pathologies associated with monochorionic twins

10.1 Twin-to-twin transfusion syndrome

10.1.1 Aetiology

There is an increased number of arteriovenous (AV) anastomoses deep in the placenta in twin-to-twin transfusion syndrome. These are mainly capillary connections that happen in the cotyledon portion of the placenta. Unidirectional flow can occur in these AV anastomoses and result in shunting of blood towards one twin and away from the other, when the arteriovenous anastomoses are unbalanced. Bidirectional flow is usually maintained by arterioarterial (AA) and venovenous (VV) anastomosis. These are found more superficially on the placenta. AA anastomoses are thought to be protective against TTTS. There is apparently a reduction in AA anastomosis in monochorionic diamniotic twins and thus these twins are more susceptible to TTTS. On the other hand, monochorionic monoamniotic twins are thought to have more AA anastomoses, which is a theoretical reason why rates are lower in these twins than in MCDA twins [15, 16].

Due to the hypovolemia experienced by the donor twin, the renin-angiotensin-aldosterone system (RAAS) gets stimulated in that twin. This leads to oliguria and oligohydramnios. On the contrary, the other twin experiences hypervolemia which causes a cardiac stretch. This leads to an increase in atrial natriuretic peptide and brain natriuretic peptide release in the recipient twin. This inhibits the RAAS and leads to polyuria and polyhydramnios [17–19]. The consequences are atrioventricular valve insufficiency, diastolic dysfunction and pulmonary stenosis or atresia in the recipient twin.

Based upon data from referral centres, the Society for Maternal-Fetal Medicine (SMFM) estimates a prevalence of:

- Stage I: 11–15%
- Stage II: 20–40%
- Stage III: 38–60%
- Stage IV: 6–7%
- Stage V: 2% [20]

10.1.2 The diagnostic monitoring of TTTS

It is now well recognised that from 16 weeks onwards all women with monochorionic pregnancies should have a fortnightly ultrasound to diagnose TTTS. Literature suggests ultrasound-based signs as more reliable in the diagnosis of TTTS than physical examination or symptoms.

The ultrasound criteria recommended for the diagnosis of TTS are as follows:

Significant fluid discordance—this is very vital in the diagnosis of TTTS. An oligohydramnios with DVP less than 2 cm in the donor sac and polyhydramnios with a DVP greater than eight cm before 20 weeks and more than 20 cm after 20 weeks.

Discordant bladder appearances: No urine in the donor foetal bladder before 26 weeks of gestation. This should be used as a criterion of severe TTS.

Hemodynamic and cardiac comprise in both recipient and or donor twin [2].

Other studies suggest using hydrops fetalis (a condition associated with ascites, pleural, pericardial effusion and skin oedema) of the recipient twin as the criteria for ultrasound diagnostic features as well as growth restriction which can happen in 50% of donor twins as ultrasound criteria of TTTS along with the above standards. The foetal growth restriction is defined as an estimated foetal weight of <10% of normal or an abdominal circumference (AC) of <5% of normal in the setting of otherwise normal foetal growth [21].

10.1.3 Staging

The staging system is most utilised for TTTS is the Quintero Staging System, which is based upon two-dimensional ultrasound and Doppler study findings and is as follows:

- Stage I: oligohydramnios and polyhydramnios sequence, donor twin bladder is visible, Doppler studies of UA/UV/DV are normal in both twins.
- Stage II: oligohydramnios and polyhydramnios sequence, donor twin bladder is not visible, Doppler studies of UA/UV/DV are normal in both twins.
- Stage III: oligohydramnios and polyhydramnios sequence and abnormal Doppler study (only one of the following is required in either twin) [absent/reversed end-diastolic flow in UA, pulsatile flow in UV, or reversed a-wave flow in DV].

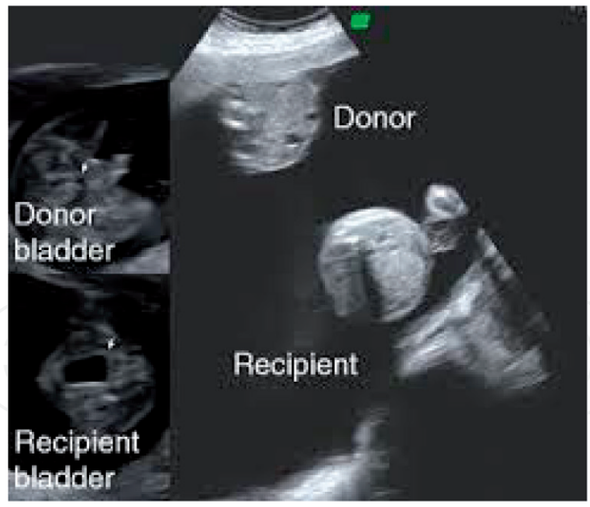

Figure 5.
Twin to twin transfusion syndrome. Courtesy (Springer open.com).

- Stage IV: oligohydramnios and polyhydramnios sequence, and one or both foetuses have hydrops.

- Stage V: oligohydramnios and polyhydramnios sequence, and one or both foetuses have died.

The above staging has been adopted from the Society of Maternal Fetal Medicine (**Figure 5**) [20].

10.1.4 Management options for TTTS

Management recommendations differ based on the stage of TTTS and gestational age and are outlined below [15].

- Stage I: The management of stage 1 is controversial. Expectant management is considered a recommended option. Similar outcomes comparing expectant management to amnioreduction and foetoscopic laser photocoagulation have been noted. Consider weekly ultrasounds for follow-up. Up to 25% of Stage I TTTS may progress to another stage. Expectant management is usually associated with a high survival chance of at least one twin in most pregnancies.

- Stage II, III, IV: The recommended treatment for these three stages is foetoscopic laser photocoagulation when the gestational age is <26 weeks. Laser photocoagulation has shown to have better outcomes than serial amnioreductions, including increased survival rates of one or both twins, delivery at greater gestational ages, and superior neurological outcomes. TTTS diagnosed before 26 weeks of gestation is best treated by laser ablation, as the evidence suggests that it leads to better outcomes compared with amnioreduction or septostomy. It is generally accepted that Quintero stages II and above will require treatment, and many centres will manage Quintero stage I conservatively. However, if laser ablation expertise is not available, amnioreduction is an acceptable alternative in pregnancies diagnosed after 26 weeks of gestation. There are some evidences that laser ablation is still the best form of treatment for TTTS, regardless of

whether it is diagnosed early (before 16 weeks) or late (after 26 weeks of gestation).

- Stage V: No interventions have been evaluated at this stage.

Foetoscopic laser photocoagulation is the most well-known procedure for the management of TTTS. The rate of twin survival increases significantly after treatment with foetoscopic laser therapy up to 88% for at least one twin and 62% for the survival of both twins [22]. It should be performed under ultrasound guidance typically between 15 and 26 weeks of gestation. The aim of the procedure is to create 2 chorions which will individually supply blood to each twin. Although the procedure can be performed even earlier or later, there are drawbacks when performed at different times than recommended. If done below 16 weeks, the risk of PPROM is high while the risk of difficulty in coagulation exists after 25 weeks. This is mainly due to the increase in the size of blood vessels after 25 weeks.

The following are the various kinds of foetoscopic laser procedures used [23]:

A) Selective laser

In this procedure, at first, abnormal vessels are mapped by following them from origin to termination. A vessel that starts from one foetus, incorporates into a cotyledon and then journeys through the other foetus. This is considered pathologic, and it is this vessel which is photocoagulated. On the contrary, the vessel that leaves the cord as an artery, enters a cotyledon and returns to the same foetus as a vein is not pathologic and not treated. This procedure is called selective photocoagulation [21, 24].

Laser coagulation of the anastomoses is done through bursts of energy over 3–4 seconds from 1 cm. The laser energy can be tailored to the working distance, type and size of the vessels. If larger vessels are involved, multiple shots may be required to coagulate the vessels. Higher laser energy can be more efficient to prevent perforation of foetal vessels, bleeding and may lead to less placental damage. Typically (Nd: YAG: wavelenth 1066 nm are used).

Caution should always be taken to avoid contact between the laser fibre and tissue. In case, the targeted vessel is behind the membrane, the laser energy can be fired through the membrane.

B) Sequential laser

The coagulation in this procedure is done in the following way order of type of connections [21].

- Donor artery—recipient vein
- Recipient artery—donor vein
- Artery-artery
- Vein-vein
- AAs and VVs are usually called superficial anastomosis, whereas arteriovenous (AV) anastomosis typically involves an anastomosis between the artery and

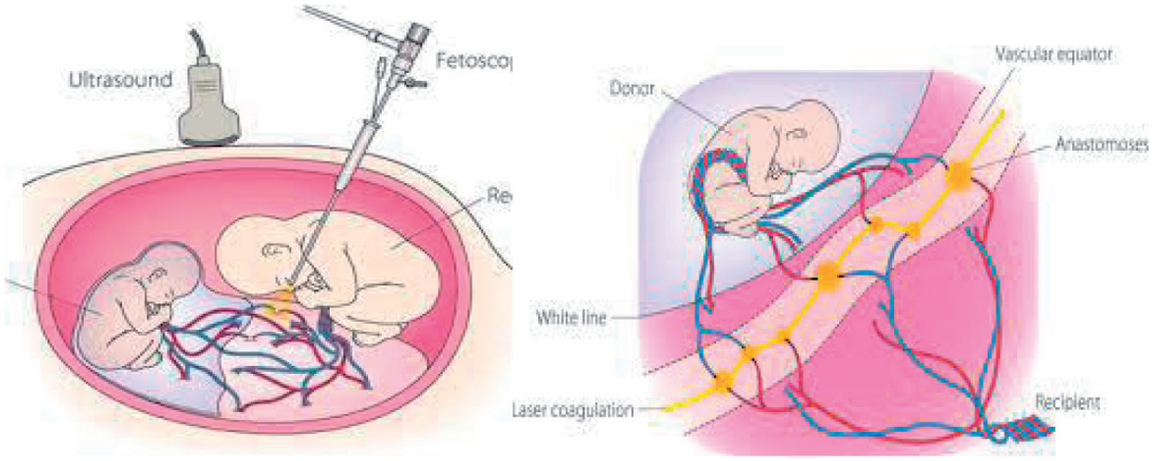

Figure 6.
Solomon technique: Courtesy—(google images-researchgate.net).

the vein and is called deep anastomosis. In an uncompensated case, the blood supply from the artery of one will twin will drain into the vein of the other twin. Sequential lasering involves grouped coagulation of AA, AV and VV anastomosis. In this technique, at first AV (donor to recipient) is coagulated, followed by VA (recipient to donor) [25]. Several studies (non-randomised) have shown better survival rates with this method. Technically, such a procedure reduces the risk of hypotension in the donor twin.

C) Solomon technique

The Solomon technique was developed as an advancement of coagulation techniques. The 'Solomon technique' involves initially completing coagulation of all visible anastomoses and then performing coagulation to connect the anastomoses ablation sites from one edge of the placenta to the other [22, 26]. This method enables the monochorionic placenta to be dichorionised by coagulating placental vessels and the surface of placenta. Although this technique results in fewer TTTS recurrences, decreased development of TAPS and increased perinatal survival, the risk of PPROM and placental abruption is high. This could be due to the increased exposure of the placental tissue to laser energy. This technique was found to be superior to the conventional technique in a randomised controlled study (**Figure 6**) [27].

10.1.5 Follow-up after laser coagulation

An initial follow-up a day after the procedure to rule out complications such as intrauterine death of one or both twins and cervical length.

The pregnancy can then be followed every two weeks as per the usual schedule. A weekly ultrasound is also recommended post-operatively [28]. The complications that could occur may be from procedure-related surgical complications to recurrences, brain, cardiac and limb abnormalities. As the procedure is associated with a risk of neurological damage, some centres perform a follow-up MRI, but this is still not a routine recommendation [27, 29].

Delivery is recommended in treated cases at around 35 weeks after giving steroids.

Illustrative diagrams of various laser techniques (**Figures 7** and **8**).

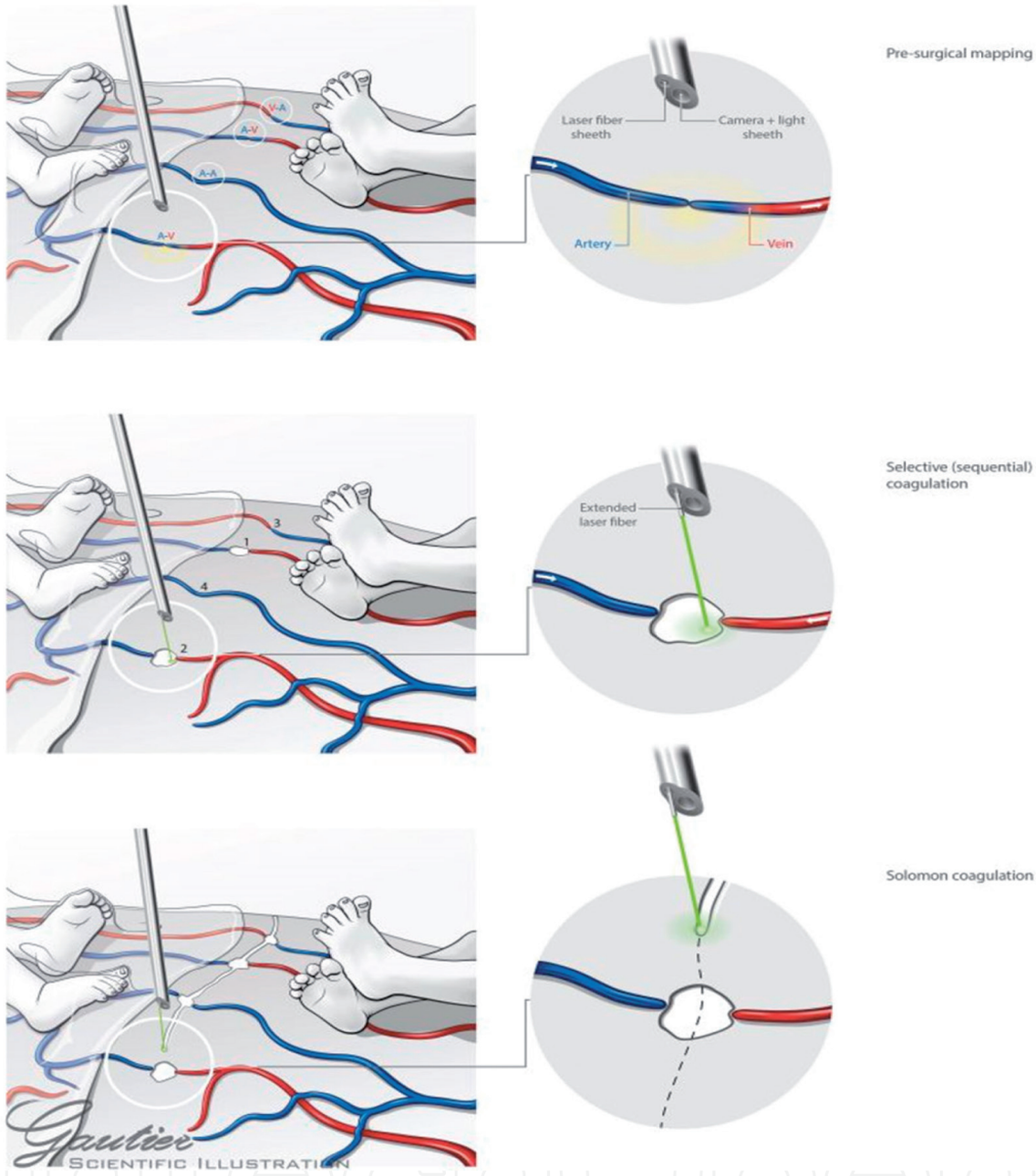

Figure 7.
Courtesy: Gautier Scientific Illustrations.

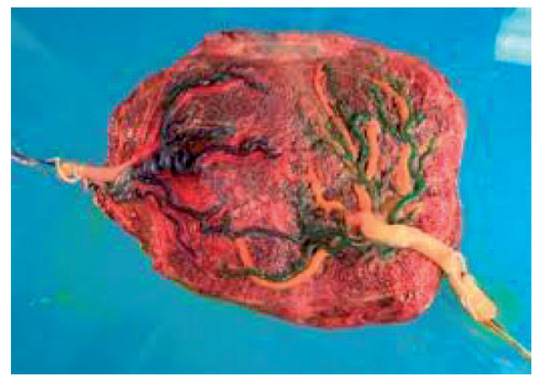

Figure 8.
Placental Anastomosis in TTTS (Courtesy: e-medicine.com).

10.1.6 Short- and long-term foetal outcomes following post laser treatment

Short-term early complications could include foetal demise of one or both twins. The risk is higher when the procedure is performed before 17 weeks. Similarly, the rates of preterm premature rupture of membranes (PPROM) may be higher when performed earlier than 17 weeks. TAPS may occur after missed small anastomosis in up to 3% of cases after dichoronisation.

Other complications could be a persisting or recurrent TTTS. This could happen in up to 1% of cases due to technical difficulties. A second laser could be a possibility but could be complicated by a haemorrhagic amniotic fluid due to a previous procedure.

Long-term foetal outcome shows that the survival of at least one or both twins is high. The survival rate of both twins is reported as 35–65%, whereas the survival of at least one twin is reported as between 70 and 88% [30]. Long-term neurodevelopment impairment was reported as 10% [31]. Neurological damage in TTTS may occur because of antenatal injury secondary to hemodynamic and haematological imbalance and/or from postnatal injury associated with prematurity [32] and low birth weight. Thermal injury damages may happen infrequently if unexpected foetal movements happen. A rare but serious complication could be vascular limb occlusion. This could vary from mild skin damage to total limb amputation and happens usually in the lower limbs.

10.1.7 Maternal complications of laser treatment

This could vary from mild peritoneal irritation due to leakage of fluid or blood in abdominal cavity, to other complications such as preterm labour, iatrogenic PPROM and delivery. Other complications could include infection, haemorrhage and placental abruption.

10.1.8 Amnioreduction

In the past, amnioreduction was the only treatment available for TTTS, but not anymore. Although foetal laser coagulation is now considered a gold standard for TTTS, a certain group of patients may still benefit from amnioreduction. Amnioreduction is also a more suitable option when TTTS is diagnosed after 26 weeks of gestation.

Amnioreduction is useful in the setting of TTTS when the criteria for laser surgeries are not met and when laser surgery is not technically possible in certain cases and in some cases of post-laser coagulation is a relatively simple treatment which does not require high-tech equipment [32].

Reduction of elevated amniotic fluid volume in polyhydramnios decreases the amniotic pressure which may lead to increased flow from the placenta to the foetus, as well as increases placental perfusion provided all other characteristics are unchanged. Controlled amnioreduction is considered a better option than random drainage of amniotic fluid [33].

Complications could include PPROM, infections, death of one or both twins (survival rates following this procedure range from 50 to 65%), need for serial amnioreductions, preterm labour, placental abruption, infection and decreased success of potential future foetoscopic laser photocoagulation. [15] The neurological impairments associated with amnioreduction are comparable with foetal laser therapy indicating that the predictor for neurological impairment is gestational age irrespective of management [32].

10.1.9 Septostomy

Septostomy is a procedure when an intentional rupture of the intertwin septum is done under ultrasound guidance. The aim of the procedure is to balance the amniotic fluid pressure in the two sacs. This may lead to a correction of the placental circulation, mainly in the donor twin's vessels. As the amniotic sac of the donor twin is filled, it reduces cord compression and improves foetal haemodynamics. As a result, the urine production of donor twin is improved [34].

Complications of septostomy are same as for serial amnioreduction, such as preterm labour and premature rupture of membranes. An additional risk is represented by the creation of a monoamniotic pregnancy that can lead to cord entanglement. The survival rates following septostomy for TTTS vary widely from 36 to 83%. Gestational age is increased in cases of septostomy compared with amnioreduction. Survival rates between septostomy and amnioreduction are comparable and septostomy offered the advantage of requiring a single procedure [34]. Another potential risk associated with septostomy is the presence of membrane flaps which may induce an amniotic band syndrome, a potentially dangerous complication. Experience with septostomy is limited and there is a need for further evaluation of this technique (**Figure 9**).

10.2 Twin anaemia polycythaemia sequence

Twin anaemia polycythaemia sequence (TAPS) is defined by significant intertwin haemoglobin discordance. It does not have the amniotic fluid discordance that characterises twin-twin transfusion syndrome (TTTS) in monochorionic twin pregnancies. This difference clearly distinguishes TAPS from TTTS.

TAPS is a rare disorder and can occur spontaneously (3–5%) or following foetoscopic laser ablation for TTTS (13–15%). This complication is thought to result from chronic transfusion through very small placental anastomoses. The pathogenesis of TAPS is not known.

A small number of usually very tiny and mostly unidirectional, arteriovenous placental anastomoses are seen in this condition. TAPS that follow laser surgery is associated with a smaller number of recurrent placental anastomoses than when it occurs spontaneously. The slow, and likely low volume, blood transfusion, which does not

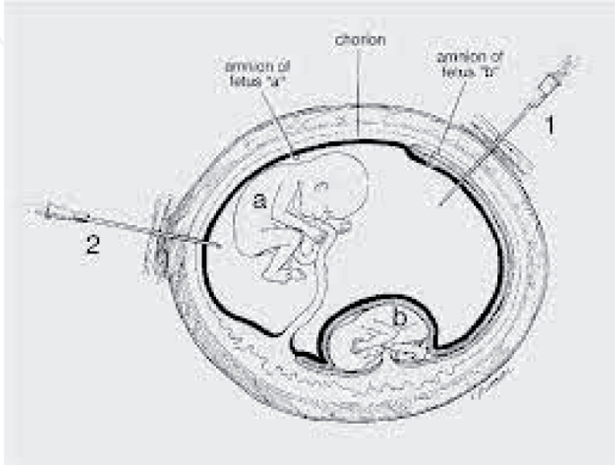

Figure 9.
Illustrative diagram of Septostomy (Courtesy: Semanticsscholar.org).

cause a great impact on the recipient's plasma volume (as the classic TTTS does), could be the possible explanation for there being no discordance in amniotic fluid volumes [35]. As a result of their extremely small size, these anastomoses can be missed during coagulation; thereby, Solomon technique is considered a better technique to prevent TAPS. The condition can be diagnosed both in antenatal and postnatal periods [36].

Middle cerebral artery peak systolic velocity (PSV) should be performed routinely following post-laser treatment to detect TAPS. The sensitivity of middle cerebral artery PSV to diagnose TAPS is more than 90%. Anaemia and polycythaemia both can be diagnosed using middle cerebral artery PSV antenatally. A middle cerebral PSV > 1.5 multiples of the median (MoM) for the donor twin and < 0.8 MoM in the recipient is proposed for antenatal diagnosis [37].

TAPS classification Antenatal stage Findings at Doppler ultrasound examination [37]:

- Stage 1—MCA-PSV donor >1.5 MoM and MCA-PSV recipient<1.0 MoM, without other signs of foetal compromise
- Stage 2—MCA-PSV donor >1.7 MoM and MCA-PSV recipient<0.8MoM, without other signs of foetal compromise
- Stage 3—as stage 1 or 2, with cardiac compromise of donor, defined as critically abnormal flow
- Stage 4—hydrops of donor
- Stage 5—intrauterine demise of one or both foetuses preceded by TAPS

Postnatal stage Intertwin Hb difference, g/dl

- Stage 1 > 8.0
- Stage 2 > 11.0
- Stage 3 > 14.0
- Stage 4 > 17.0
- Stage 5 > 20

Apart from the inter-twin haemoglobin difference of more >8 g/dl and one of the two following criteria should also be there; either a reticulocyte count ratio of >1.7 or the presence of only small vascular anastomoses (diameter 1 mm) on placental inspection (**Figure 10**) [37].

10.2.1 Treatment

Treatment options in utero include expectant management, intra-uterine transfusion both (intraperitoneal or intravascular) in the donor with or without partial exchange transfusion (PET) in the recipient, selective foeticide and laser therapy.

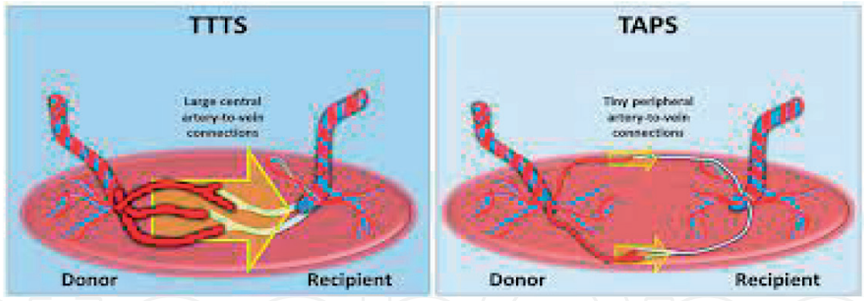

Figure 10.
The illustrative difference between TTTS and TAPS (courtesy—hopkinsmedicine.org).

Despite all options, laser remains the only treatment option that can resolve the possible causal mechanism. The preferred laser coagulation in TAPS should be the Solomon technique as this technique diminishes the risk of residual anastomoses and recurrent TAPS [38].

Intrauterine transfusions and partial exchange transfusions can temporarily stabilise the hemodynamic situation in severely affected foetuses. Overall, different management options may improve survival and perinatal outcomes like respiratory morbidities in affected foetuses [35, 39].

Some cases of intrauterine transfusions have been reported to be successful in prolonging the pregnancy. Even though the only pivotal treatment for TAPS is foetoscopic laser coagulation of the residue vascular anastomoses, this can be technically trickier than in TTTS. The reason for this is the absence of polyhydramnios and a stuck twin. This situation makes the visualisation of the vascular equator more difficult. The other reason is placental anastomoses in TAPS are known to be only few and minute. As a result, this may be missed during foetoscopy because of their size.

The precise perinatal mortality and morbidity frequency in TAPS is not known yet, probably because of the oddity of the condition. Spontaneous resolution of antenatal TAPS has also been described [40].

The results from recent studies suggest that spontaneous TAPS may have a better prognosis than post laser TAPS. As for mortality and morbidity, no differences were observed when comparing different management options for TAPS. Caution should be applied when interpreting these results due to scarcity of literature. A tailored antenatal management, considering the severity of TAPS and gestational age, is currently the recommended strategy [39].

10.2.2 Neonatal outcome

Haematological complications are commonly seen in TAPS donors and recipients. This may require postnatal blood transfusions or partial exchange transfusions. Another condition that the recipient can develop is polycythaemia hyper viscosity syndrome, which could possibly lead to necrosis of the skin and multiple limb ischemia. The recipient twin is also more at risk of thrombocytopenia due to impaired production because of tissue hypoxia [38].

Because of anaemia in donor twin and polycythaemia in recipient twin, cerebral injury can theoretically occur and cases of reported cerebral injuries have been reported in both twins. Recent studies show that long-term neurodevelopmental outcome in

Figure 11.
Twin Anaemia Polycythaemia Syndrome—Courtesy:(en.wikepedis.org).

post-laser TAPS, not indicate mild to moderate cognitive delay in 9% and 17% of TAPS survivors, respectively. No difference in neurological impairment was found between donors and recipients. The rate of neurological impairment in TAPS seems to be comparable to the rate of impairment in children with TTTS after laser surgery. Risk factors may include early gestational age at delivery and smaller babies (**Figure 11**) [38].

10.3 Selective intrauterine growth restriction: (sIUGR)

This condition affects approximately 10–15% of monochorionic (MC) twin pregnancies. Selective intrauterine growth restriction is diagnosed when the estimated foetal weight (EFW) in one twin of <3rd percentile or an intertwin EFW discordance ≥25% is observed on ultrasound.

The main complications are the possible risk of intrauterine death of one twin or neurological damage of both twins. Unequal sharing of the placenta is the main cause of this condition, and the clinical outcome is closely related to the placental vascular anastomosis [41].

10.3.1 Pregnancy outcome in sIUGR:

- The risks of foetal demise of one or both foetuses
- Preterm delivery
- Subsequent development of TTTS
- Increased risk for neurodevelopmental impairment, with poorer outcome of the smaller twin

10.3.2 The diagnosis of sIUGR

Diagnosis of sIUGR is typically made in the second trimester based on foetal biometric measurements, growth discordance and umbilical artery (UA) Doppler parameters. sIUGR is defined as [42]:

- Estimated foetal weight (EFW) <3rd percentile of one foetus [5] or

- At least two of the four following criteria [42]:
- EFW <10th percentile for one twin
- Abdominal circumference < 10th percentile for one twin
- Weight discordance ≥25%
- UA pulsatility index >95th percentile for the smaller twin

It is recommended that from 20 weeks of gestation (at 2-weekly intervals) onwards at each scan the estimated foetal weight discordance should be calculated using two or more biometric parameters. The percentage EFW discordance should be calculated using the following formula:

([larger twin EFW – smaller twin EFW]/larger twin EFW) × 100. Liquor volumes as DVP should be measured and recorded (to differentiate from TTTS) [2].

As the EFW discordance of greater than 20% is associated with an increase in perinatal risks, these pregnancies should be referred to the specialist centres for further evaluation and management. One parameter that best reflects the differences in intrauterine growth restriction (IUGR) in monochorionic pregnancy with respect to singletons or dichorionic twins is umbilical artery (UA) Doppler flow. The characteristics of UA Doppler flow may be strongly affected by the existence of intertwin vascular connections.

In MC twin pregnancies complicated by sIUGR, UA Doppler waveforms represent the combined effect of placental insufficiency and placental vascular anastomoses.

Three main wave form patterns of diastolic flow in umbilical artery of smaller twin have been recognised. Hence, sIUGR is classified as [43]:

- Type 1—It is characterised by persistently forward UA end-diastolic velocity without variation in the waveform with normal or elevated resistance. This type has the best prognosis and the mean gestational age at delivery was after 35 weeks. It has the lowest risk of intrauterine foetal death and survival rates are high. Usually, late-onset sIUGR are type 1 and their prognosis is good as well, although they are at increased risk of TAPS.
- Type 2—It is characterised by fixed absent or fixed reversed UA end-diastolic velocity without any alteration of the waveform in the smaller twin. Affected foetuses will have worsening conditions in mid-trimester and the delivery is usually required at an average gestational age of 30 plus weeks.

Although pregnancies with type 2 sIUGR are anticipated to have a predictable pattern of deterioration and a longer latency period between diagnosis and deterioration than type 3 sIUGR, in terms of risk of death of one twin and preterm delivery, their prognosis is worst among all 3 types of sIUGR. Interestingly, there is usually no neurological damage seen in majority of survivors.

- Type 3—It is characterised by a pathognomonic UA waveform that has a variable flow pattern that cycles between forward, absent, and reversed flow over a short interval, which is termed intermittent absent/reversed end-diastolic flow. This happens due to a large artery-to-artery anastomosis on the placental surface and

signifies the bidirectional volume flow across these vessels. It is more commonly observed in the UA of the smaller foetus since the interface of the two waveforms is shifted toward the smaller twin. An artery-to-artery anastomosis allows perfusion of oxygen and nutrients from the larger foetus to a portion of the smaller twin's placenta; consequently, type 3 sIUGR is associated with the largest degree of placental territory discordance (**Figure 12**) [44].

These pregnancies have unpredictable diagnosis, and foetal death can occur even shortly after a satisfactory ultrasound assessment. There is a high risk of neurologic morbidity as well, particularly of the larger twin. Survival rate has been reported up to 61%.

10.3.3 Diagnostic workup for suspected sIUGR should include [43]:

A detailed ultrasound anatomic survey of both twins to rule out structural anomalies.
Maternal viral serology or ultrasound markers to rule out foetal viral infections.
Evaluation of amniotic fluid of both twins to rule out coexisting TTTS and
Evaluation of middle cerebral artery peak systolic velocity to rule out coexisting TAPS.

10.3.4 Management based on the types [41, 43]:

- sIUGR type 1—Expectant management

Weekly ultrasound surveillance of (umbilical artery [UA], middle cerebral artery [MCA]).

Weekly biophysical profile scoring (BPP) from 28 to 32 weeks.

If the UA pulsatility index increases to >95th percentile or the MCA pulsatility index falls below the 5th percentile, a twice weekly surveillance is recommended with additional monitoring of abnormalities in the ductus venosus (DV) waveform.

If foetal status remains reassuring, as it usually does, it is recommended to deliver delivery at 34 + 0 to 35 + 6 weeks (after steroids) as in pregnancies with sIUGR the risk of unexpected foetal death is higher than uncomplicated monochorionic pregnancies. Earlier delivery is indicated if deemed necessary for any maternal or foetal indications.

- sIUGR types 2 and 3—Due to the complexity of the condition the approach to these cases is more complicated. Death of one twin is high in these cases and can result in acute foetal transfusion and volume shifts, which leads to the double foetal demise or neurologic damage in the surviving co-twin in up to 30% of cases.

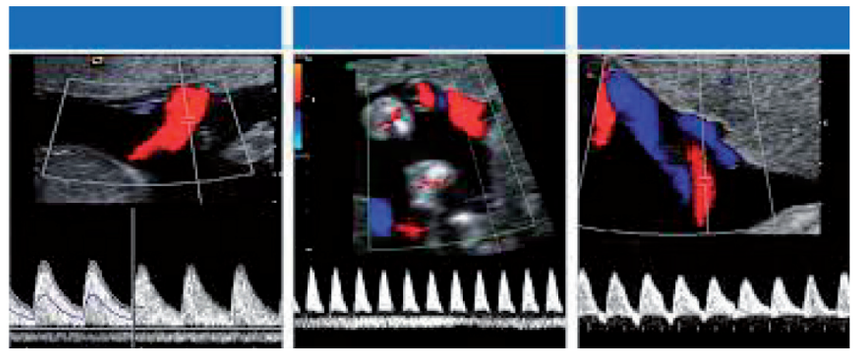

Figure 12.
The three types of selective intrauterine growth restriction (courtesy: isug.org).

The recommended approach may include either selective foetal reduction or foetoscopic laser ablation of intertwin placental vascular anastomoses (before the lower limit of viability), especially when there is foetal deterioration (progression from type 2 to type 3 sIUGR, venous Doppler abnormalities or oligohydramnios in the growth-restricted foetus.

Foetoscopic laser ablation can result in high mortality rate of the smaller twin and does not guarantee the survival of the normal twin but may protect the normally grown twin from the consequences of co-twin demise. The procedure may be more technically challenging compared with foetoscopic laser ablation for the treatment of twin-twin transfusion syndrome.

10.3.5 In patients not willing for above procedures

Weekly Doppler surveillance of UA, MCA and DV should commence from the diagnosis.

Perform weekly biophysical profile (BPPs) at 28 weeks.

If Doppler findings remain stable and BPPs are reassuring, they can be followed up as outpatients on a weekly basis.

If Doppler findings worsen increase surveillance with Dopplers/BPP to two to three times weekly and advise hospital admission for daily foetal monitoring with nonstress tests. Delivery should be considered between 32 and 34 week and earlier if indicated. Steroids should be given prior to deliver.

10.4 Selective feticide of one twin

Selective foeticide in monochorionic twins is performed by cord occlusion, intrafoetal laser ablation or radiofrequency ablation. The risk of miscarriage and or preterm birth may be influenced by the timing. A higher risk of these complications occurs when the procedure is performed in second trimester. When the diagnosis is made in the second trimester, a late selective termination in the third trimester can be offered to the woman, as the law permits. The risk of preterm birth in third trimester is less compared to the risk in second trimester. For selective foeticide of one twin of a monochorionic pair injection of intracardiac potassium chloride is not an option (unlike in dichorionic twin) due to the risk involved to the healthy co-twin. Instead, cord occlusion, intrafoetal laser ablation or radiofrequency ablation should be done. This procedure does not cause the healthy twin to lose its circulating blood volume in the terminating twin. The survival rate of the co-twin is approximately 80% in such cases [5].

Risks involved are

Premature rupture of the membranes and preterm birth in up to 20% of cases prior to 32 weeks.

Miscarriages in second trimester.

Increased risk of neurological damage in surviving twins.

10.5 Single intrauterine foetal demise. (Death of a co-twin).

Single intrauterine foetal demise (sIUFD) is a rare but exceptional perinatal problem in twin pregnancies. Monochorionicity and gestational age at the time of stillbirth seem to be decisive factors in terms of long-term neurologic outcome

prediction for the survivor [45]. Monochorionic pregnancies are at particular risk of sIUFD due to bidirectional inter-twin placental vascular anastomoses. The intertwin blood flow becomes unbalanced and can lead to acute and chronic inter-twin transfusion and profound anaemia secondary to foetal exsanguination into the low-pressure circulation of the dead foetus [42]. The co-twin is at increased risk of preterm delivery, long-term neurological complications, and death especially when the condition occurs after 14 weeks of gestation.

The increased risk of neurological damage in the surviving twin could be due is to the bidirectional inter-twin vascular anastomoses that are found in monochorionic placentation. This results in unbalanced inter twin blood flow and leading to acute and chronic inter-twin transfusion and profound anaemia, which are seen in conditions such as TTTS, twin-anaemia-polycythaemia sequence (TAPS) and twin-oligo-polyhydramnios sequence (TOPS) [42]. As a result of these conditions, a multi-organ injury may occur causing significant hypoperfusion of the surviving twin. This may have been initiated by acute foetal exsanguination leading to low-pressure circulation of the dead foetus. The end results are hypoxic-ischaemic injury to the central nervous system of the surviving twin (up to 36%) and subsequent brain injury, or intrauterine death of the surviving twin [42, 46]. The other proposed theory is that "thromboplastic materials" from the dead twin to the surviving twin through the placental anastomosis which in turn causes disseminated intravascular coagulation (DIC) in the surviving twin. This results in renal, pulmonary, hepatic, splenic and neurological infarcts in surviving twin. But there are doubts as to the fact that the DIC can occur so fast; hence, it may be unlikely to be a causative mechanism [47].

10.5.1 Diagnosis

The most accurate diagnosis can be made by Magnetic resonance imaging (MRI). Diffusion weighted imaging (DWI) has recently been shown to add to the accuracy of the diagnosis with a timelier diagnosis [45].

Ultrasound detection of brain damage is possible in later stages of foetal brain injury. Also, ultrasound lesions may identify lesions like atrophic and necrotic cystic lesions or ventriculomegaly but not those associated with hypoxic ischemic injuries. This is due to the technical difficulty of the acoustic bone shadowing of skull bones. An early diagnosis and multidisciplinary counselling must be provided to the parents to make an informed choice.

Three types of injuries have been identified:

1. Ischemic hypoxic lesions of white matter which is irrigated by middle cerebral artery.

   This leads to porencephaly, multicystic encephalomalacia, microcephaly and hydranecephaly.

2. Haemorrhagic lesions isolated or associated with ischemic lesions leading to post haemorrhagic hydrocephalus.

3. Anomalies secondary to vascular disruption leading to neural tubal defects and optic nerve hypoplasia.

The best time to identify these injuries on MRI is usually considered between 1 and 3 weeks. The use of DWI is unanimously accepted as superior with respect to the precocity of diagnosis. DWI signal changes occurring in cerebral ischemia may be detected early, within the first day of co-twin demise. Its usefulness is restricted to the

first week after the death of the co-twin, interval after which pseudo normalisation occurs [45]. Decision to terminate pregnancy should be reserved for foetuses with severe ischemic injury (**Figure 13**).

10.5.2 The ethical and clinical challenges

A multidisciplinary team with maternal foetal medicine specialist, neonatologist, paediatric neurologist and neurosurgeon should be involved in counselling. Parents should be given accurate information to make a final decision. Should parents opt for continuing the pregnancy the risks of cerebral palsy and iatrogenic preterm delivery should be explained. Foeticide is not legally accepted in all countries. Social, cultural, and religious beliefs also add up in deciding.

10.5.3 Management

Management of such cases will also depend upon any maternal or foetal infections or conditions that could impair the survival of the other twin. DIC could also happen in rare cases because of the release of tissue thromboplastin from dead foetus in maternal circulation activating extrinsic coagulopathy. However, this is not very common (25%). As it occurs usually 3–5 weeks following foetal demise, clotting profiles should be performed in week 1 and repeat in 2–3 weeks [48].

In the short term, the surviving twin should be assessed for evidence of ongoing foetal compromise using CTG or MCA Doppler to assess for foetal anaemia. If conservative management is chosen, foetal biometry and assessment of umbilical and MCA Doppler should be scheduled every 2–4 weeks. Delivery should be considered at 34–36 weeks, after a course of maternal steroids. If the MCA-PSV is normal in the first few days, foetal anaemia is unlikely to occur later [5].

10.6 Twin Reversed Arterial Perfusion (TRAP)

Twin reversed arterial perfusion (TRAP) sequence, also named as acardiac malformation, is an exclusive complication of monochorionic multiple pregnancy. In this condition, one of the twins has no cardiac structure (and so is called 'acardiac'), while a morphologically normal co-twin (called 'pump twin') supplies both circulations. Historically, the first case was described by Benedetti in 1533, and the first cases

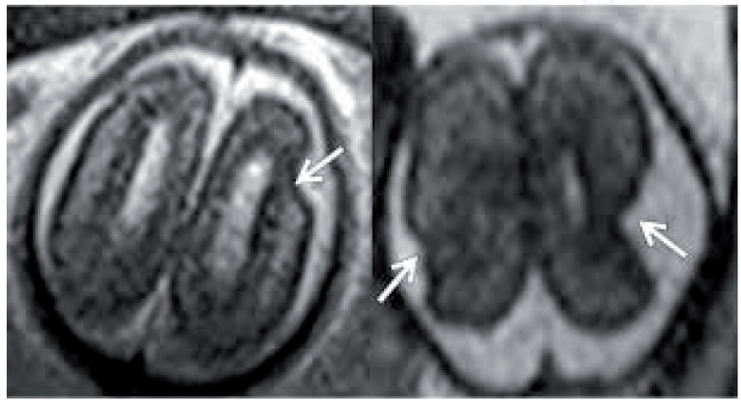

Figure 13.
MRI changes in the brain of the surviving twin after the death of co twin. Courtesy-(pubs.rsna.org).

had been reported in the international literature in the 1950s; the first description of prenatal diagnosis of an acardiac twin was reported by Lehr and Dire in 1978 [49].

10.6.1. Pathogenesis

Pathogenesis involves two pathways. First is that an unequal blood flow between the twins is noticed. The pump twin predominates due to its high-pressure flow, while the perfused twin receives a reversed deoxygenated blood flow. This leads to compromised morphogenesis. As there is no functioning heart developed the acardiac twin relies on the circulation of pump twin in a parasitic fashion.

The second pathogenesis proposed is that there is an embryogenic defect with a failure in heart formation, due to chromosome abnormality or environmental factors. Hence the single perfusion support for the acardiac foetus is received through anastomoses between the umbilical vessels. The acardiac twin is not viable but keeps getting vascular support from the pumped twin, which supplies deoxygenated blood to the acardiac twin. It has a well-developed body and upper extremities and a big size; hence, it remains a danger during the intrauterine period and is dangerous for the whole pregnancy.

The well-being of the pump twin can also be compromised through at least three mechanisms [49]:

1. Congestive heart failure (30%) and polyhydramnios of the pump twin (40%o, caused by a risen cardiac work due to the increased blood flow.

2. Preterm premature rupture of membranes (pPROM), preterm labour and preterm delivery (90%), caused by uterine overdistension, since the acardiac twin is often bigger than pump twin and it can reach a considerable size (acardiac twin to pump twin ratio > 70%).

3. Hypoxia and intrauterine growth restriction of the pump twin, caused by the deoxygenated blood that comes back to the pump twin through vascular anastomosis.

The perinatal mortality rate of this twin is 55%. The risk of demise of the pump twin in TRAP sequence if managed conservatively is up to 30% by 18 weeks.

10.6.2 Diagnosis

The diagnosis is made by ultrasound. Features noticed on ultrasound are:

a. Gross discrepancies in biometrical measurements of twins, regarding abdominal circumference.

b. Absence of a morphologically normal heart in one twin associated with several other malformations in head, trunk, upper and lower extremities: presence of subcutaneous oedema and fluid collections in the anomalous twin (**Figure 14**) [49].

Based on the morphology of the acardiac foetus, 4 different types have been described: acardiac acephalus; acardiac anceps; acardiac acormus; acardiac amorphous. However, they have no prognostic value and no difference in management options.

A classification based on prenatal ultrasound findings as acardius size and signs of impaired cardiac function of the pump twin have been proposed. This

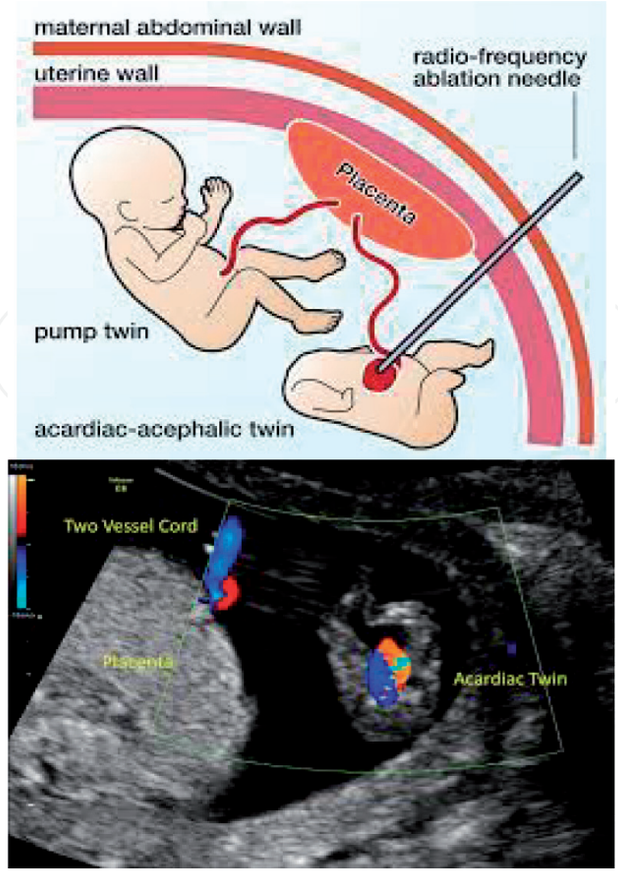

Figure 14.
Illustrative and ultrasound images of TRAP (courtesy—google image).

classification may help in identifying the most severe cases and those that need prenatal interventions [49].

Acardiac anomalies are divided into:

- Type I: small or medium-sized acardiac twins, identified by an abdominal circumference ratio < 50%.
- Type II: large acardiac twins, in which the abdominal circumference ratio is ≥50%.

Each type can be further divided into a 'subtype a and b', if pump-twin does not show signs of cardiovascular failure, or into a subtype.

10.6.3 Management

The main goals in the management of the TRAP sequence are preserving the survival of the pump twin and reaching the term for delivery. It is observed that the prognosis of the pump twin in Type Ia, acardiac foetus is quite reassuring. This allows a conservative management of pregnancy through periodic ultrasound. This approach is associated with a good outcome in 88% of cases. In the presence of an acardius Type Ib, it is reasonable to repeat ultrasound to identify a spontaneous resolution or a worsening that requires invasive treatment.

The Type IIa acardiac foetus can be large because of subcutaneous oedema or hydrops, and even if now of diagnosis, the pump foetus shows no signs of cardiac

failure, the large size could threat the whole pregnancy due to an increased risk of preterm labour. In this case, a prenatal treatment is required. The detection of a Type IIb acardius requires a prompt intervention [49].

The best timing of intervention is not clear. It should be performed preferably before 16 weeks [5]. A pump-twin loss rate of 33% in the time elapsed from the first-trimester diagnosis and the elective intervention at 16–18 weeks, highlights an important disadvantage of delayed intervention. A multicentre, open-label, randomised controlled trial currently ongoing (ClinicalTrails.gov: NCT02621645), named the TRAP Intervention Study (TRAPIST), comparing treatment at 13–15 weeks vs. treatment from 16 weeks, is expected to define the optimal timing of treatment.

Different minimally invasive techniques, such as cord coagulation, cord ligation and photocoagulation of the anastomoses, as well as intrafoetal methods, such as Radiofrequency Ablation and intrafoetal laser therapy, are performed as a means of preventing the demise of the pump twin. The survival rate of the pump twin using these treatment modalities is approximately 80%. TRAP sequence pregnancies should be monitored serially. The aim is to take intrauterine therapy as an option if cardiac strain becomes evident in the pump twin or there is increased perfusion (including the occurrence of polyhydramnios) and growth of the TRAP mass. Hence, these cases should be managed in a tertiary level centre [5].

11. Timing of delivery and Intrapartum management of uncomplicated monochorionic twins

Parents should be informed that usually a planned birth is recommended at 36 completed weeks and this does not increase the risk of any neonatal morbidity [10]. It is well documented that continuing pregnancy beyond 36 weeks is associated with increased risk of still births in monochorionic pregnancies. With an uncomplicated monochorionic twin pregnancy, vaginal birth and planned caesarean section are both safe choices for them, and vaginal delivery can be offered if the following criteria are met [10]:

- The pregnancy was uncomplicated throughout and has progressed beyond 32 weeks.
- There are no obstetric contraindications to labour.
- The first baby is in a cephalic (head-first) presentation.
- There is no significant size discordance between the twins.

If the first twin is not cephalic at the time of birth, a caesarean section should be offered. Corticosteroids should be offered prior to the planned delivery at 36 weeks.

During labour continuous, cardiotocography (CTG) should be commenced. A dual channel cardiotocography monitor should be used to allow simultaneous monitoring of both foetal hearts. As labour progresses, a foetal scalp electrode can be used for the first twin if no contraindications. If there is foetal dress and foetal blood sampling cannot be done, caesarean section should be performed after the birth of the first baby.

Once the first twin is delivered, continue to monitor the second baby using CTG. If the CTG shows a ‘suspicious’ or ‘pathological’ pattern, and vaginal birth is not

possible within 20 minute a caesarean section should be offered. Epidural should be offered for vaginal birth and regional anaesthesia for caesarean section.

Third stage should be managed actively with controlled cord traction and oxytocin (Active management). In a vaginal birth, active management consists of 10 IU of oxytocin by intramuscular injection immediately after the birth of the last baby and before the cord is clamped and cut. In a caesarean section, it consists of 5 IU of oxytocin by intravenous injection immediately after the birth of the last baby and before the cord is clamped and cut [10].

12. Conclusion

MCDA twins are associated with several well-known complications. Updated guidelines and basic standards should be adhered to for a better outcome of these complications. Challenging cases may need individual care, but basic principles of early screening, diagnosis, accurate follow-up, and timely intervention should be the best approach. Poor or suboptimal care may directly be related to lack of observance to updated guidelines and non-accessibility of advances in management. The advanced role of ultrasound and relatively newer technologies such as laser photocoagulation for the treatment of severe TTTS, radiofrequency ablation and cord occlusion for selective reduction have significantly enhanced the outcomes for many of the complications of MCDA twins. Screening for these conditions is of paramount significance for the early diagnosis with timely intervention to improve neonatal morbidity and mortality. A first-trimester ultrasound to evaluate the interface of the intertwin membrane with the placenta, timely detection of chorionicity and antenatal surveillance of these pregnancies is the key to improved outcomes in MCDA twins. Future researches are required to further improve the overall survival rate and reduce the incidence of neurological impairment associated with intervention procedures.

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