Table 1. Complications associated with inter-twin vascular anastomoses

TTTS (Quintero staging) Associated with 15% of monochorionic twins	I	**A significant discordance in amniotic fluid volumes**. This is defined as oligohydramnios with deepest vertical pocket (DVP) < 2 cm in donor sac and polyhydramnios in the recipient sac (DVP > 8 cm before 20 weeks of gestation and > 10 cm after 20 weeks of gestation). Donor bladder visible and Doppler normal.
	II	Bladder of the donor twin not visible and severe oligohydramnios due to anuria. Doppler studies are not critically abnormal.
	III	Doppler studies are critically abnormal in either the donor or recipient, with typically abnormal umbilical arterial Doppler velocities in the donor and/or abnormal venous Doppler velocities in the recipient (reversed flow during atrial contraction within the ductus venosus and/or pulsatile umbilical vein velocities).
	IV	Ascites, pericardial or pleural effusion, scalp oedema or overt hydrops present usually in the recipient.
	V	One or both babies have died (not amenable to therapy).
TAPS 2% of uncomplicated monochorionic diamniotic (MCDA) and up to 13% of monochorionic twins post laser ablation		Signs of fetal anaemia in the donor and polycythaemia in the recipient **without** significant oligohydramnios/polyhydramnios being present. Donor has increased middle cerebral artery peak systolic velocity (MCA PSV) (> 1.5 multiples of the normal median) and recipient has decreased MCA PSV (< 1.0 multiples of the normal median).
sGR (growth discordance of > 20%) Approximately 10–15% of monochorionic twins	I	Growth discordance but positive diastolic velocities in both fetal umbilical arteries.
	II	Growth discordance with absent or reversed end-diastolic velocities (AREDV) in one or both fetuses.
	III	Growth discordance with cyclical umbilical artery diastolic waveforms (positive followed by absent then reversed end-diastolic flow in a cyclical pattern over several minutes [intermittent AREDV; iAREDV]).
TRAP sequence Approximately 1% of monochorionic twins		Acardiac twin (with usually no cardiac tissue) being perfused by the anatomically 'normal' pump twin through a large artery–artery anastomosis on the placental surface.

3. Identification and assessment of evidence

This guideline was developed in accordance with standard methodology for producing Royal College of Obstetricians and Gynaecologists (RCOG) Green-top Guidelines. The Cochrane Library (including the Cochrane Database of Systematic Reviews and the Database of Abstracts of Reviews of Effects [DARE]), EMBASE, Trip, MEDLINE and PubMed (electronic databases) were searched for relevant randomised controlled trials, systematic reviews and meta-analyses. The search was restricted to articles published between 1966 and 2015. The databases were searched using the relevant Medical Subject Headings (MeSH) terms, including all subheadings, and this was combined with a keyword search. Search words included 'monochorionic twin', 'TTTS', 'twin twin transfusion syndrome', 'TRAP syndrome', 'amnioreduction', 'laser ablation', 'septostomy' and 'cord occlusion' and the search was limited to humans and the English language. The National Library for Health and the National Guideline Clearinghouse were also searched for relevant guidelines and reviews. The

most important of these is the 2011 National Institute for Health and Care Excellence (NICE) clinical guideline 129,[1,5] which was based upon an extensive review of the evidence for the antenatal management of twin and triplet pregnancies. An evidence update to this guideline was published in 2013.[6] The proceedings of the 50th RCOG Study Group on Multiple Pregnancy also gave important expert opinion used in this document.[4]

In addition, qualitative information and patient representation has been provided by Mr Keith Reed on behalf of the Twins And Multiple Births Association and Ms Jane Denton on behalf of The Multiple Births Foundation.

Where possible, recommendations are based on available evidence. In the absence of published evidence, these have been annotated as 'good practice points'. Further information about the assessment of evidence and the grading of recommendations may be found in Appendix I.

4. Diagnosis of monochorionic twin pregnancy

4.1 *How is monochorionicity diagnosed prenatally and what is the accuracy of prenatal ultrasound chorionicity allocation?*

All women with a twin pregnancy should be offered an ultrasound examination between 11^{+0} weeks and 13^{+6} weeks of gestation (crown–rump length 45–84 mm) to assess fetal viability, gestational age and chorionicity, and to exclude major congenital malformations. B

Chorionicity should be determined at the time the twin pregnancy is detected by ultrasound based upon the number of placental masses, the appearance of the membrane attachment to the placenta and the membrane thickness. This scan is best performed before 14 weeks of gestation. D

A photographic (thermal copy) record should be taken and placed in the patient's notes documenting the ultrasound appearance of the membrane attachment to the placenta and an electronic copy stored (Appendix II). ✓

If there is uncertainty about the diagnosis of chorionicity, a photographic record of the ultrasound appearance of the membrane attachment to the placenta should be retained and a second opinion should be sought. ✓

If there is still doubt in the diagnosis of chorionicity, the woman should be referred to a specialist without delay, as chorionicity is best determined before 14 weeks of gestation. D

On ultrasound, the fetuses in twin pregnancies should be assigned nomenclature (i.e. upper and lower, or left and right) and this should be clearly documented in the woman's case notes to ensure consistency throughout pregnancy. C

First trimester ultrasound scanning is important in multiple pregnancies as it confirms viability, determines gestational age, defines chorionicity (by denoting placental mass numbers and membrane thickness), determines the number of amniotic sacs and may identify fetal anomalies (i.e. large cystic hygroma, anencephaly).[1,6] In monochorionic twins, it is also important to exclude 'acardiac twinning', which is associated with TRAP. At this ultrasound examination, screening for Down syndrome may also be offered to couples (see below).

Evidence level 2++

In spontaneously conceived twins, gestational age can be determined at the first trimester scan by using the crown–rump length of the larger fetus to avoid the risk of estimating it from a baby with early growth pathology.

The accurate assessment of chorionicity is more sensitive and specific in the first trimester and it is important to assess and document chorionicity clearly at this gestational age. NICE guidance[1,5,6] indicates that this should be performed at the time of the first trimester scan to screen for Down syndrome. There is significant benefit in identifying women with monochorionic pregnancies early as these pregnancies will require additional fetal surveillance. It also allows women to be fully counselled on the risks and appropriate management of monochorionicity to be implemented. Differentiating between monochorionic and dichorionic pregnancies will result in a saving of time and emotional stress by avoiding unnecessary additional interventions in dichorionic pregnancies.

Evidence level 4

Clinicians should be aware that, although ultrasound assessment of chorionicity is very accurate, it does not have 100% sensitivity. The NICE guideline identified 14 studies investigating diagnostic accuracy of the following characteristics (as determined by an ultrasound scan) for determining chorionicity:[1,5,6,21]

- Number of placental sites and the characteristics of the inter-twin membrane placental insertion (so-called lambda/T-sign) in the first trimester. Approximately 3% of monochorionic placentas have two placental masses (bilobed placenta), so these are not necessarily dichorionic.[22]
- Inter-twin membrane thickness using two-dimensional ultrasound: i) with thickness usually subjectively defined, but in the literature, this is less than 1.8 mm for monochorionic twins; and ii) 1.5–2.0 mm can be inconclusive for monochorionic twins, but when used with other factors, chorionicity can usually be defined in 99% of cases.[23,24]
- The number of membrane layers (subjectively noted).[25]
- Composite measures based on the above characteristics and others (number of placental masses, number of gestational sacs, concordant fetal sex [in monochorionic twins] and number of fetal poles).[1,4–6]

Evidence level 1+

The strongest likelihood ratios were reported for a composite method involving presence of a lambda or T-sign, and noting the number of placental masses identified using ultrasound in the first trimester. Using such criteria, sensitivity and specificity were greater than 95%.[24]

Evidence level 2+

Monochorionic twin pregnancies have a single placental mass and a thin inter-twin membrane that inserts into the placenta at a perpendicular plane (T-sign). In contrast, dichorionic twin pregnancies have two placental masses (or adjacent placental masses forming a ‘lambda sign’ as placental tissue is present where the thick inter-twin membrane inserts onto the placenta [Appendix II]).[1,5,6]

Evidence level 2++

This assessment should be followed up with noting concordant fetal sex at the midtrimester ultrasound scan.[4]

Evidence level 4

It is sometimes necessary to review the assigned chorionicity of multiple pregnancies in the second or even third trimester. A retained photographic record (either in the patient’s handheld notes or stored in the hospital notes) will allow this to be easily reviewed. An archived image should also be stored.

The sensitivity and specificity of ultrasound to define chorionicity is highest before 14 weeks of gestation.[1,23,24] If a trained sonographer, as part of routine first trimester screening, cannot define with confidence the chorionicity of the pregnancy, then a second opinion must be sought from a senior sonographer or the woman should be offered referral to a healthcare professional who is competent in determining chorionicity by ultrasound scan as soon as possible.[1]

Evidence level 3

If it is difficult to determine chorionicity, even after referral (for example, because the woman has booked late in pregnancy), then the pregnancy should be managed as monochorionic until proved otherwise.[1]

Accuracy in labelling monochorionic twins is important so that serial growth scan measurements can be consistently applied to the same twin throughout pregnancy and that any intrauterine or neonatal management can be directed towards the correct twin. A retrospective study[24] found that ascribing labels based on lateral or vertical orientation was a reliable method of twin labelling. This study, based on 416 twin pregnancies, labelled the twin whose gestational sac was closest to the cervix at the 11–14-week scan as 'twin 1' and recorded the twins' orientation as lateral or vertical given their relative positions to each other (left/right or top/bottom, respectively). Approximately 90% of twins were oriented laterally and 10% vertically. The presenting order of 8.5% of the laterally oriented twins changed between the first and last scans; there were no such changes in the vertically oriented pregnancies. Based on a subset of mixed-sex twins (n = 108), the presenting order in a third trimester scan was different to the subsequent birth order in 5.9% of pregnancies delivered vaginally and 20.3% of pregnancies delivered by caesarean section.

Evidence level 2+

The authors concluded that recording the twins' relative position to each other was a reliable method of labelling twin pregnancies compared with designating the babies as 'twin 1' and 'twin 2', and was conceptually more open to the possibility that the anticipated birth order of the twins might change, especially where delivery is by caesarean section.[24]

5. Outcome of monochorionic twin pregnancy

5.1 What is the outcome of monochorionic compared with dichorionic twin pregnancies?

Clinicians and women should be aware that monochorionic twin pregnancies have higher fetal loss rates than dichorionic twin pregnancies, mainly due to second trimester loss and, overall, may have a higher risk of associated neurodevelopmental morbidity. This should form part of the parental counselling.

✓

A prospective Scandinavian study[26] of 495 pregnancies diagnosed before 15 weeks found fetal loss at less than 24 weeks of gestation to be 14.2% in monochorionic compared with 2.6% in dichorionic pregnancies ($P < 0.05$). The number of babies alive at 8 months of age (percentage of fetuses at inclusion) was 82.4% monochorionic compared with 96.3% dichorionic ($P < 0.01$). Mean birthweight (both twins) and mean gestational age at delivery was lower in monochorionic twins ($P < 0.01$). A retrospective study of 1407 twin pregnancies in the Netherlands over a 10-year period found the perinatal mortality rate to be 11.6% in monochorionic twins compared with 5.0% in dichorionic twins. Monochorionic twins continued to be at greater risk even after 32^{+0} weeks.[27] A retrospective study of all twin pregnancies of known chorionicity from a large regional cohort of nine hospitals over a 10-year period in south London has indicated an

Evidence level 2+

increased fetal loss rate prior to 24^{+0} weeks in monochorionic twins (60.3/1000 fetuses) compared with dichorionic twins (6.6/1000 fetuses) and that this risk continues through pregnancy as risk of stillbirth after 26^{+0} weeks.[28,29] This has been confirmed by several studies in different parts of the world.[30,31]

In addition, neurological morbidity may be up to seven-fold higher in preterm monochorionic infants compared with dichorionic infants due to associated complications of TTTS, sGR and single twin death in utero.[32,33]

Evidence level 2+

6. Optimal screening for chromosomal abnormalities, structural abnormalities and other fetal complications in monochorionic twin pregnancies

6.1 *What is the optimum method of screening for chromosomal abnormalities in monochorionic twin pregnancies?*

Women with monochorionic twins who wish to have aneuploidy screening should be offered nuchal translucency measurements 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). C

In women with monochorionic twin pregnancies who 'miss' or who have unsuccessful first trimester screening for aneuploidy, second trimester screening by the quadruple test should be offered. D

Early data with noninvasive prenatal testing (NIPT) are encouraging, but results should be interpreted with caution until larger studies have been carried out. C

Monochorionic twins are not at increased risk of chromosomal abnormalities over dichorionic twins, but the prevalence of aneuploidy is slightly increased overall in multiple pregnancies because of an indirect association with increased maternal age.[1,4]

Evidence level 3

First trimester combined screening using nuchal translucency and the serum analytes of free beta-human chorionic gonadotrophin and pregnancy-associated plasma protein A (combined test screening) should be offered to pregnant women with monochorionic twins at a crown–rump length of between 45 and 84 mm.[1] This screening test has good sensitivity for detecting aneuploidy (90%), but the false-positive rate (up to 10%) is higher than in singletons (2.5%) and dichorionic twins (5%).[1]

When screening is performed by nuchal translucency, first trimester serum screening and maternal age, a pregnancy-specific risk is calculated in monochorionic twins. This is in contrast to dichorionic twins in whom a fetus-specific risk is calculated.[1,34]

Evidence level 2+

Approximately 10% of pregnant women do not have their first antenatal booking appointment in the first trimester. If the gestation at first visit is after 14 weeks, then second trimester serum screening should be offered using the quadruple screen test (with an 80% detection rate and a false-positive rate of 3%) so as to not disadvantage these women. This screening test in monochorionic twins provides a pregnancy-specific risk.[1,35]

Evidence level 3

