In type I sGR, planned delivery should be considered by 34–36 weeks of gestation if there is satisfactory fetal growth velocity and normal umbilical artery Doppler waveforms. [*New 2016*] ✓

In type II and III sGR, delivery should be planned by 32 weeks of gestation, unless fetal growth velocity is significantly abnormal or there is worsening of the fetal Doppler assessment. [*New 2016*] ✓

It is important to prospectively inform parents that in sGR and TTTS (even after apparently successful treatment) there can be acute transfusional events (which are neither predictable nor preventable) and therefore, despite regular monitoring, there may still be adverse perinatal outcomes. [*New 2016*] ✓

Management of TAPS

Clinicians should be aware that the natural history, fetal and neonatal implications, and optimal treatment and/or surveillance of monochorionic pregnancies diagnosed with TAPS are poorly established. [*New 2016*] D

The management of monochorionic twin pregnancies complicated by single twin demise

What are the consequences for the surviving twin after fetal death of the co-twin in a monochorionic pregnancy and what is optimal clinical management?

Clinicians should be aware that monochorionic pregnancies not complicated by TTTS, sGR or TAPS are still at risk of fetal death and neurological abnormality. D

After a single fetal death in a monochorionic pregnancy, clinicians should be aware that the risks to the surviving twin of death or neurological abnormality are of the order of 15% and 26%, respectively. [*New 2016*] B

Single fetal death in a monochorionic pregnancy should be referred and assessed in a fetal medicine centre, with multidisciplinary expertise to manage these cases. ✓

Fetal magnetic resonance imaging of the brain may be performed 4 weeks after co-twin demise to detect neurological morbidity if this information would be of value in planning management. D

How should fetal anaemia be monitored after single twin intrauterine death?

Fetal anaemia may be assessed by measurement of the fetal MCA PSV using Doppler ultrasonography. D

Timing and mode of delivery in uncomplicated monochorionic pregnancies

What is the optimal timing and method of delivery for otherwise uncomplicated monochorionic pregnancies (without TTTS, sGR or TAPS)?

Women with monochorionic twins should have timing of birth discussed and be offered elective delivery from 36^{+0} weeks with the administration of antenatal steroids, unless there is an indication to deliver earlier. [*New 2016*] C

It is appropriate to aim for vaginal birth of monochorionic diamniotic twins unless there are other specific clinical indications for caesarean section. A

Monochorionic monoamniotic (MCMA) pregnancies

What are the specific problems of MCMA pregnancies and how should they be managed?

MCMA twins almost always have umbilical cord entanglement when visualised using colour flow Doppler. Such a finding has not consistently been demonstrated to contribute to overall morbidity and mortality. [*New 2016*] D

MCMA twins have a high risk of fetal death and should be delivered by caesarean section between 32^{+0} and 34^{+0} weeks. [*New 2016*] D

Higher order multiple pregnancies with reference to monochorionicity

What is the outcome of monochorionic and dichorionic compared with trichorionic triplet pregnancies?

Clinicians should be aware that monochorionic/dichorionic triplet pregnancies have higher fetal loss rates than trichorionic triplet pregnancies and may be complicated by feto–fetal transfusion syndrome, sGR and TAPS. C

Selective reduction should be discussed in all higher order pregnancies including triplets. [*New 2016*] ✓

Increased ultrasound surveillance is warranted in a fetal medicine centre with expertise to manage such cases. ✓

Discordant abnormalities in monochorionic pregnancies

What is the incidence of and the therapeutic options for discordant abnormalities in monochorionic pregnancies, including twin reversed arterial perfusion sequence?

Monochorionic twins that are discordant for fetal anomaly must be referred promptly for assessment and counselling in a fetal medicine centre with consideration for treatment. ✓

Karyotyping of monochorionic twins should be managed in a fetal medicine centre. [*New 2016*] ✓

Meticulous mapping of the position of the twins within the uterus should be performed both at the time of prenatal diagnostic tests and invasive treatments. [*New 2016*] ✓

During amniocentesis, both amniotic sacs should be sampled in monochorionic twin pregnancies, unless monochorionicity is confirmed before 14 weeks and the fetuses appear concordant for growth and anatomy. [*New 2016*] ✓

Prior to invasive testing or in the context of twins discordant for an abnormality, selective reduction should be discussed and made available to those requesting the procedure after appropriate counselling. [*New 2016*] ✓

Monitoring for disseminated intravascular coagulopathy is not indicated in monochorionic twin pregnancies undergoing selective reduction. [*New 2016*] ✓

Selective feticide by intravascular injection of an abortifacient is not an option in monochorionic pregnancies because of the presence of placental anastomoses. The potential risks of intrafetal/umbilical cord ablative procedures should be discussed prospectively, including the risk of co-twin loss and neurological morbidity. [*New 2016*] ✓

Documentation and discussion of heterokaryotypic monozygotic karyotypic abnormalities should take place. [*New 2016*] ✓

Conjoined twins

How are conjoined twins diagnosed and what are the outcomes?

Conjoined twins are exceedingly rare and prenatal assessment is required in a tertiary fetal medicine centre so that diagnosis can be confirmed and prognosis discussed in conjunction with a multidisciplinary team. [*New 2016*] ✓

What are the training competencies required for managing monochorionic pregnancies?

All sonographers who undertake routine ultrasound scans during pregnancy must be trained to establish chorionicity and the correct labelling of twins. ✓

All sonographers who undertake midtrimester (18^{+0}–20^{+6} weeks) and fetal growth scans of monochorionic twins should be made aware of the appearances of TTTS, sGR and TAPS, and the need to refer patients on to specialist centres if such features present. ✓

Fetal medicine centres undertaking fetal therapy for relatively rare complications of monochorionic twins should have a minimum of two experienced operators and more than 15 cases per year (rolling 3-year average) to maximise perinatal outcomes and minimise long-term morbidity. D

1. Purpose and scope

The purpose of this guideline is to evaluate and provide recommendations on best practice for the management of problems associated with monochorionic placentation and their effects upon multiple pregnancies. The use of ultrasound to determine chorionicity and amnionicity is key to the management of multiple pregnancies and the potential risks to the fetuses. This guideline will outline the best evidence to guide clinical care, including fetal surveillance and treatment of complications for monochorionic multiple pregnancy. It is important to emphasise that this guideline is focused upon the management of monochorionic multiple pregnancies rather than all multiple pregnancies.

It is also recognised that women carrying a monochorionic pregnancy (most commonly twins) may have concerns and anxieties surrounding their pregnancy. This requires accurate information given in a sensitive manner by healthcare professionals and support by a multidisciplinary team, ideally within a multiple pregnancy clinic.[1] Support is also often given in conjunction with the Twins And Multiple Births Association and The Multiple Births Foundation within the UK.

2. Introduction and background epidemiology

A monochorionic pregnancy is a multiple pregnancy, most commonly a twin pregnancy, in which babies are dependent on a single, shared placenta. Approximately 30% of twin pregnancies in the UK are monochorionic. Monochorionic placentation can also occur in rarer, higher order multiples, especially triplets (i.e. dichorionic or monochorionic triplets).

There has been an increase in all types of multiple pregnancies with the increasing use of assisted reproductive technology, sociodemographic changes in our population associated with migration and deferment of pregnancy to a later maternal age. Assisted reproductive technology increases the prevalence of both dichorionic and monochorionic twinning. However, using day 5 blastocyst transfers seems to have a significantly higher rate of monozygotic twinning (adjusted OR 2.04, 95% CI 1.29–4.48) compared with cleavage stage day 3 transfers.[2,3]

Monochorionic and dichorionic twin pregnancies have increased risks of preterm birth, fetal growth restriction (FGR), pre-eclampsia, postpartum haemorrhage and postnatal complications, such as infant feeding difficulties and adverse puerperal mood change.[1,4]

The particular challenges of monochorionic pregnancies arise from the shared placenta and vascular placental anastomoses that are almost universal and connect the fetal circulations of both twins.

Specific complications associated with inter-twin vascular anastomoses are listed in Table 1. Note that there may occasionally be some challenging diagnostic overlap among these definitions (e.g. twin-to-twin transfusion syndrome [TTTS] versus selective growth restriction [sGR] with reduced liquor around the smaller twin – see section 6.4.3):

- TTTS
- sGR
- twin anaemia-polycythaemia sequence (TAPS)
- twin reversed arterial perfusion (TRAP) sequence
- single intrauterine death; although not exclusive to monochorionic twin pregnancy, it is more common and has global effects on the co-twin.

In addition, the consequences for single fetal death and the management of discordant fetal anomalies (i.e. structural and chromosomal anomalies) in monochorionic twins is important.

Monochorionic diamniotic (MCDA) twin pregnancies carry a higher risk of overall fetal and perinatal loss compared with dichorionic pregnancies. Furthermore, monochorionic monoamniotic (MCMA) pregnancies, where both twins are in a single amniotic sac (1% of monochorionic twins), carry a very high risk of perinatal loss, most commonly before 24 weeks.[1,4–7]

Almost all monochorionic placentas contain vascular anastomoses running between the two fetal umbilical cords within and on the surface of the placenta. These connect the fetal circulations. In 80% of cases, these are bidirectional vascular anastomoses which rarely lead to haemodynamic imbalance between the fetal circulations, but allow a direct vascular connection between the twins with an increased risk of fetal death.[8,9]

In TTTS, which complicates up to 15% of monochorionic pregnancies,[4] the placentas have a predominance of unidirectional artery–vein anastomoses. This may lead to a haemodynamic imbalance within the circulations of the twins, directly adversely affecting fetal cardiac function, fetoplacental perfusion, but also adversely affecting fetal renal function by discordant activation of the renin–angiotensin axis.[4,10]

Postnatal perfusion studies have noted unequal placental 'territory' shared by the fetuses with associated marginal or 'velamentous' cord insertions. Such findings are common both in TTTS and sGR (which is often associated with TTTS).[9,11–13]

Very rarely, TTTS complicates MCMA twin pregnancies, as well as both dichorionic and monochorionic triplet pregnancies.[11,14]

TAPS is an important association in complicated monochorionic pregnancies, especially TTTS, occurring in up to 13% of cases post laser ablation.[15] It may relatively rarely be associated with apparently uncomplicated MCDA twins. The pathogenesis of TAPS is evidenced by postnatal placental injection studies demonstrating 'miniscule' artery–vein anastomoses (less than 1 mm) allowing the slow transfusion of blood from the donor to the recipient. This may be associated with highly discordant haemoglobin levels at birth (80 g/l or greater).[15–18]

Significant intrauterine fetal size discordance (difference in estimated fetal weight [EFW] of greater than 20%), termed 'selective growth restriction' (sGR), occurs in up to 15% of monochorionic twins in the absence of TTTS and in over 50% of monochorionic twins complicated by TTTS.[19] sGR is recognised as a specific monochorionic twin pathological entity associated with a significant differing placental territory between each fetus, inter-fetal placental anastomoses and abnormal fetoplacental blood flow.[13] The incidence of size discordance is as great in dichorionic pregnancies in some series,[20] but management of discordant growth is more complex in monochorionic pregnancies due to the associated placental anastomoses conjoining the fetal circulations.

It is recognised that in monochorionic twin pregnancies, because of these potential and specific complications, there may be significant anxiety and concern in parents, even if such abnormalities are not present. Accurate information presented in a sensitive manner during discussions is important so as to allay unnecessary fears, while imparting to couples the importance of appropriate increased prenatal surveillance.[1,5,6]

