7.2 *The management of sGR*

sGR in monochorionic twins requires evaluation in a fetal medicine centre with expertise in the management of such pregnancies. ✓

In cases of early-onset sGR in association with poor fetal growth velocity and abnormal umbilical artery Doppler assessments, selective reduction may be considered an option. C

In sGR, surveillance of fetal growth should be undertaken at least every 2 weeks with fetal Doppler assessment (by umbilical artery and middle cerebral artery pulsatility index, and peak systolic velocity). If umbilical artery Doppler velocities are abnormal, the Doppler assessments should be undertaken in line with national guidance, measuring ductus venosus waveforms. D

Clinicians should be aware that there is a longer 'latency period' between diagnosis and delivery in monochorionic twins complicated by sGR compared with growth restriction in dichorionic twin pregnancy or singleton pregnancy. D

Abnormal ductus venosus Doppler waveforms (reversed flow during atrial contraction) or computerised CTG short-term variation should trigger consideration of delivery. B

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. ✓

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. ✓

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. ✓

Once there is a suspicion of sGR or the diagnosis has been prospectively made using ultrasound, the pregnancy should be managed in conjunction with a regional fetal medicine centre with specialist expertise in managing this condition. There is a need to measure amniotic sac liquor volumes and to assess Doppler velocities within the fetal arterial and venous circulations.[53,57,59] Evidence level 4

Umbilical artery Doppler evaluation in monochorionic twins with sGR allows the prospective definition of three subtypes (see Table 1)[53,57,59] and the consequences are dependent upon this evaluation and gestational age. Evidence level 3

When sGR is diagnosed such that the difference in EFW is greater than 20%, fetal and perinatal loss is increased.[19,56] Prior to viability (24 weeks), if the small twin has a significantly reduced fetal growth velocity (change in measured abdominal circumference of less than 1 SD over 14 days) in the presence of umbilical artery Doppler abnormalities, there is a significant risk of single fetal demise.[59] Evidence level 2+

In such circumstances, to protect the appropriately grown co-twin, selective termination of pregnancy using vaso-occlusive techniques, such as bipolar cord occlusion or radiofrequency ablation may be considered.[79,83] This should be assessed and performed in a tertiary centre with expertise. Again, informed but sensitive discussion with patients is mandatory.

Evidence level 2+

There is limited evidence to guide clinical care in these complex cases. However, there is international consensus that such monochorionic twin pregnancies require regular review with interval ultrasound biometry to monitor fetal growth velocity, and placental and fetal circulation assessment by umbilical artery, middle cerebral artery and ductus venosus Doppler waveform measurements.[53,57,59,84] The aim is to prolong pregnancy to at least viability and to achieve appropriate gestation for delivery (32–34 weeks), but to avoid the complication of single fetal death and the consequences for the surviving fetus.[53]

Evidence level 2–

Timing of delivery (less than 32 weeks of gestation) is dependent upon assessment by computerised CTG and/or ductus venosus waveform velocimetry. For monochorionic twin sGR where there is umbilical Doppler abnormality, delivery should be undertaken at 32 weeks (in line with singleton FGR with AREDV).[85]

Evidence level 2++

The placental anastomoses in monochorionic twins paradoxically may be beneficial for the smaller twin as transfusion from the larger twin may compensate for the placental insufficiency, thus interfering with the natural history in comparison with singleton and dichorionic twin pregnancies. This is also associated with the presence of artery–artery anastomoses. This prolongs survival in the growth-restricted fetus, resulting in a longer latency period to deterioration and delivery (up to 10 weeks versus 3–4 weeks from diagnosis of FGR).[8]

In monochorionic twins, where the placental vascular anastomoses remain intact, there is a risk of acute 'inter-twin' transfusional events causing fetal death and morbidity in the form of neurological morbidity. This is true of apparently uncomplicated monochorionic twins, but more prevalent in monochorionic twins complicated by sGR and even treated TTTS.[1,4]

Evidence level 3

7.3 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.

D

Fetoscopic laser ablation for the treatment of TTTS, using the Solomon technique, significantly reduces the risk of recurrent disease and TAPS.[15] The optimal management of TAPS, once it occurs, is not clear from the literature, but possible options include expectant management, delivery, intrauterine blood transfusion (intravenous and/or intraperitoneal, with or without partial exchange transfusion), selective feticide or fetoscopic laser surgery.[86] There is little evidence relating to the outcome and optimal management of TAPS. Fetoscopic laser surgery is the only treatment for the cause of this disease (in analogy with TTTS), but is technically challenging due to the absence of polyhydramnios and the presence of only minuscule anastomoses.[15] If detected prenatally, management options need to be individualised and uncertainty discussed with the parents.

Evidence level 3

Perinatal outcome in TAPS is not well described (with or without treatment) and appears to vary according to the severity. Outcome may range from double intrauterine fetal demise to the birth of two healthy neonates with a significant inter-twin haemoglobin discordance. Knowledge on the neonatal and long-term morbidity in TAPS is scarce and based on case reports and small series.[52,86] Neonatal morbidity in TAPS appears to be mainly limited to haematological problems at birth. Donor twins may be severely anaemic and require blood transfusions, whereas recipient twins may be severely polycythaemic and require partial exchange transfusion.[52] There have been cases of severe cerebral injury in TAPS described.[87]

Evidence level 3

7.4 The management of monochorionic twin pregnancies complicated by single twin demise

7.4.1 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. 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 (MRI) 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

In monochorionic twins, where the placental vascular anastomoses remain intact, there is a risk of acute 'inter-twin' transfusional events causing fetal death and morbidity in the form of neurological morbidity. This is true of apparently uncomplicated monochorionic twins, but more prevalent in monochorionic twins complicated by sGR and even treated TTTS.[1,4]

Damage to the surviving monochorionic twin after the death of its co-twin is believed to be caused by acute haemodynamic changes around the time of death, with the survivor losing part of its circulating volume into the circulation of the dying twin. This may cause transient or persistent hypotension and low perfusion, leading to the risk of ischaemic organ damage, notably but not exclusively, to the watershed areas of the brain.[88]

Evidence level 3

Systematic reviews[89,90] have identified 22 full manuscripts considered of high enough quality of evidence to include in the review and meta-analysis. Twenty manuscripts were used to calculate overall summary statistics for monochorionic and dichorionic twins showing rates of co-twin death after single fetal death (15% compared with 3%), rates of preterm delivery after single fetal death (68% compared with 54%), the rate of abnormal postnatal cranial imaging after single fetal death (34% compared with 16%) and the rate of neurodevelopmental impairment after single fetal death (26% compared with 2%). Odds ratios were calculated from 16 manuscripts. There was no significant difference reported between preterm delivery of monochorionic and dichorionic twins (OR 1.1, 95% CI 0.34–3.51; $P = 0.9$). After single fetal death, monochorionic twins had higher odds of abnormal cranial imaging after delivery, although, this was not significant (OR 3.25, 95% CI 0.66–16.1; $P = 0.12$). After single fetal death, monochorionic twins were 4.81 times more likely to have neurodevelopmental morbidity (95% CI 1.39–16.6; $P < 0.05$).

Evidence level 1–

Clinical management is complex and should be overseen by fetal medicine experts with the knowledge and experience to advise parents about the advantages and disadvantages of different approaches.[91] Rapid delivery is usually unwise, unless at term, as fetal brain injury of the surviving twin occurs at the time of demise of the co-twin. Therefore, immediate delivery only adds prematurity to the possible hypotensive cerebral injury the surviving twin may have already sustained. Serious compromise of the surviving fetus may be anticipated and this should be discussed with parents, including the significant risk of long-term morbidity. Evidence of fetal compromise (such as significant cardiotocographic abnormality and/or evidence of anaemia in the survivor if single fetal death occurs late in pregnancy) could represent continuing and/or established damage to the brain. Fetal anaemia in the surviving twin may be or have been associated with cerebral hypoperfusion and therefore, may be associated with future development of neurological morbidity.[89–91]

Evidence level 4

A conservative management policy is often appropriate, with serial fetal brain ultrasound imaging and a fetal cranial MRI scan planned, commonly 4 weeks after the 'sentinel event'.[90,91] The appearances of intracranial neurological morbidity on ultrasound are variable and may take up to 4 weeks to develop. Fetal MRI provides earlier and more detailed information about brain lesions (haemorrhagic or ischaemic) in the surviving fetus than ultrasound and its use should be considered.[92,93]

Evidence level 3

In cases of single intrauterine demise with MRI or ultrasound findings of neurological morbidity, late termination of pregnancy would be an option. The gestational age at the time of diagnosis is relevant and the views of the parents will be of paramount importance.

7.4.2 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

In a prospective series of 20 monochorionic pregnancies complicated by single fetal death, there was a strong correlation between fetal anaemia (assessed by cordocentesis) and fetal MCA PSV.[94]

Evidence level 3

In a small series (n = 26) of pregnancies complicated by TTTS and single fetal death, the prognosis was worse for donor twins following the death of the recipient twin than vice versa. This is in keeping with the concept of enhanced blood loss through a unidirectional anastomosis.[95] There are a few reports of intrauterine transfusion of anaemic surviving co-twins, but the value of this intervention is not established within the context of preventing perinatal and long-term neurological morbidity.[96,97]

Evidence level 3

The presence of an increased MCA PSV in the surviving twin would suggest fetal anaemia and therefore, a significant inter-twin transfusion. This would increase the risk of hypotensive neurological injury and thus, would be helpful information in the counselling of parents and timing of fetal brain MRI. Treatment by intrauterine transfusion is controversial, as this may improve fetal survival without reducing the long-term risks of neurological morbidity.

8. Timing and mode of delivery in uncomplicated monochorionic pregnancies

8.1 *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.

C

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

A

For uncomplicated monochorionic pregnancies there may be a higher risk of unexplained fetal demise despite intensive fetal surveillance.[98] The 2011 NICE guideline examined this topic.[1] Gestational age profile for spontaneous birth in twin and triplet pregnancies was assessed in one cross-sectional study which suggested that the majority (58%) of women with uncomplicated twin pregnancies give birth spontaneously before 37^{+0} weeks.[21]

Evidence level 2–

Furthermore, a 2013 systematic review concluded that even uncomplicated monochorionic twin pregnancies are at substantial risk of stillbirth throughout the third trimester, which is several-fold higher than in dichorionic twin pregnancies. Given the risk of fetal death to the co-twin, these data should inform decisions around timing of delivery in seemingly normal monochorionic twin pregnancies; women with monochorionic twins should be offered elective birth from 36^{+0} weeks with the administration of antenatal steroids.[1,98]

Evidence level 2+

The Twin Birth Study,[99] a multicentre, international randomised controlled trial of planned vaginal birth versus planned caesarean section for delivery of twins, included a subcohort of uncomplicated MCDA twins (600 of 1398 randomised). The study concluded that in twin pregnancies between 32^{+0} and 38^{+6} weeks of gestation (when the first twin is a cephalic presentation), planned caesarean section delivery did not significantly decrease (or increase) the risks of fetal or neonatal death, or serious newborn morbidity as compared with vaginal delivery. Furthermore, post hoc subgroup analysis demonstrated no significant interaction of chorionicity with the primary outcomes. It was concluded that there was no significant benefit from planned caesarean section for any subgroup, including monochorionic twins.

Evidence level 1+

