As with all pregnancies, parental views will also be important in reaching a conclusion about the best, individualised method of monochorionic twin delivery, including opting for caesarean section.

In addition to the complexities of twin delivery (i.e. malpresentation) there is a small risk of acute feto–fetal transfusional events during labour and this is one of the reasons why continuous electronic fetal monitoring during labour is recommended.

9. MCMA pregnancies

9.1 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. D

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

MCMA twins have classically been thought to be at risk from cord entanglement (almost always visualised) and fetal demise. A retrospective study of 30 MCMA twin pairs reported a total survival of 60%.[7] Two pairs died after 32 weeks. Of the ten twin pairs that died in utero, cord entanglement was documented in eight. The authors recommended elective delivery at 32 weeks of gestation. Evidence level 3

However, a study of 32 MCMA twin pregnancies has suggested that cord entanglement is a feature of all MCMA twin pregnancies and most deaths occur prior to 20 weeks due to other causes (TRAP or discordant fetal anomaly).[100] MCMA twins are, therefore, probably not as dangerous as previously thought, although, surveillance and management should always be individualised.[101] Management by using sulindac to reduce amniotic fluid volumes has been advocated, but evidence for this treatment is scanty. The authors, therefore, recommended re-evaluating the very early timing of delivery for MCMA twins.[100,101]

A retrospective multicentre cohort study of 193 MCMA twin sets found that fetal deaths occurred in 18.1% of fetuses. The prospective risk of a nonrespiratory neonatal complication was significantly lower than the prospective risk of fetal death after 32^{+4} weeks. This consortium's recommendation was to deliver MCMA twins at approximately 33 weeks of gestation. All such cases should be managed in fetal medicine centres with specialist expertise and decisions on delivery made on an individual basis.[102] Evidence level 2–

10. Higher order multiple pregnancies with reference to monochorionicity

10.1 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. ✓

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

A retrospective study of 88 naturally conceived triplet pregnancies managed in three tertiary referral units in the UK found a 5.5-fold increased risk of perinatal death in dichorionic triamniotic pregnancies (that is, containing monochorionic twins) than trichorionic triamniotic pregnancies (OR 5.5, 95% CI 2.5–12.2).[103] Referral bias may have influenced the findings. A similar retrospective study from two tertiary centres in Germany described 84% survival in fetuses of monochorionic and dichorionic triplet pregnancies combined compared with 92% in fetuses of trichorionic triplet pregnancies.[104] This difference did not reach statistical significance.

Evidence level 3

The consensus views arising from the 50th RCOG Study Group[4] recommend that selective reduction should be discussed in all higher order pregnancies. A systematic review provides information on the risks of this procedure in trichorionic triplets and on the alternative option of conservative management (from six cohort studies).[105] In the reduction group (n = 482 pregnancies) compared with the expectantly managed group (n = 411), the rate of miscarriage (before 24 weeks) was higher (8.1% versus 4.4%; relative risk [RR] 1.83, 95% CI 1.08–3.16; P = 0.036) and the rate of early preterm delivery was lower (10.4% versus 26.7%; RR 0.37, 95% CI 0.27–0.51; $P < 0.0001$). It was calculated that seven (95% CI 5–9) reductions needed to be performed to prevent one early preterm delivery, while the number of reductions that would cause one miscarriage was 26 (95% CI 14–193).[105]

Evidence level 2+

In monochorionic or dichorionic triamniotic triplets (because of shared placental vasculature), this would mean either a procedure to reduce the fetal numbers to one or to consider intrafetal ablative therapy to reduce dichorionic triplets to dichorionic twins.[106] Such options are associated with an increase in total pregnancy loss.

If TTTS does occur, then it is most appropriately treated by laser ablation and the overall prognosis is better for dichorionic versus monochorionic triamniotic triplet pregnancies.[76]

Evidence level 3

11. Discordant abnormalities in monochorionic pregnancies

11.1 *What is the incidence of and the therapeutic options for discordant abnormalities in monochorionic pregnancies, including TRAP 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. ✓

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. ✓

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. ✓

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. ✓

Monitoring for disseminated intravascular coagulopathy is not indicated in monochorionic twin pregnancies undergoing selective reduction. ✓

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. ✓

Documentation and discussion of heterokaryotypic monozygotic karyotypic abnormalities should take place. ✓

A higher rate of structural anomalies is observed in twins compared with singletons.[26,39] Approximately 1–2% of twin pregnancies face the dilemma of expectant management versus selective termination following diagnosis of an anomaly affecting only one fetus.

In a structurally or size discordant monochorionic pair, discordant aneuploidy is exceedingly rare, although, not impossible. Structural anomalies in monochorionic pregnancies are twice that expected in dichorionic pairs, given the monozygosity.[39] Detailed ultrasound assessment, fetal karyotyping and a discussion of prognosis is required with reference both to the abnormal and normal twin.

Selective termination in a monochorionic pregnancy is an option,[78,79] but as the fetal circulations are not independent, it cannot be performed with injection of medical therapeutics because of the effect on the co-twin. More invasive and higher risk procedures, such as cord coagulation, and intrafetal ablative procedures, such as radiofrequency ablation, are necessary to induce termination of one twin without causing morbidity or death in its co-twin.[79] Evidence level 4

It is essential that at the time of prenatal diagnostic tests, the pregnancy is mapped carefully, noting the position and site of the fetuses in relation to the placenta and amniotic sac, and documented. This is ideally performed by the operator who would perform the technique of selective termination of pregnancy if required. It is mandatory to discuss selective termination of pregnancy and complicating factors in monochorionic twins, including the potential risk to the normal twin.[1,34]

Twin and triplet pregnancies in which the abnormal fetus underwent umbilical cord coagulation by bipolar diathermy or intrafetal laser ablation for indications that included severe discordant abnormalities or TRAP sequence have been described.[107,108] Overall, up to 82% of co-twins survived. Preterm rupture of membranes (10–15%) and chorioamnionitis remain significant complications. Fetal loss rates are 15–18% and some series have recorded transfusional neurological sequelae in up to 15%. Similar experience is reported using radiofrequency ablation.[83,109]

Monochorionic twins complicated with an acardiac twin and TRAP sequence do not always require invasive treatment. Selection for treatment appears to be dependent on:

Evidence level 3

- the relative size of the 'acardiac' twin to the 'pump' twin (the larger the acardiac twin, the greater the risk and need for therapy) and
- the presence of any cardiovascular impairment in the 'pump' twin.[110]

If treatment is considered, then there is some evidence that treatment should take place before 16 weeks of gestation (but evidence is not strong) and it should be performed in centres with expertise in such treatment modalities. Careful monitoring and ultrasound surveillance are required.[83]

Rarely, monozygous twins can have different chromosome make-ups. This is known as heterokaryotypic monozygous twinning. When anomalies are identified in the first or early second trimester in one of a monochorionic twin pair which may be markers of aneuploidy, a discussion should take place as to the merits and risks of chorionic villus sampling versus waiting for a double amniocentesis at 15–16 weeks when both sacs are sampled and the individual karyotype of each twin can be determined with certainty.[34]

Evidence level 4

12. Conjoined twins

12.1 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.

✓

Conjoined twins are very rare and by definition are MCMA twin pregnancies. The prevalence is one in 90 000 to 100 000 pregnancies. The underlying pathogenic mechanism remains uncertain. Such MCMA twins are complex and require careful detailed expert ultrasound imaging (usually including MRI) and multidisciplinary discussion. In one series of 14 cases of prenatally diagnosed conjoined twins at a single referral centre, 20% of parents opted for termination of pregnancy, 10% of fetuses died in utero and the overall individual survival rate to discharge of those attempting pregnancy continuation was about 25%, the majority of whom have significant morbidity.[111] Most cases are now prenatally diagnosed and delivered by elective caesarean section, but vaginal deliveries of conjoined twins are reported.[112] Risk of dystocia and uterine rupture has been reported in association with cases undiagnosed prenatally.

Evidence level 2–

Prenatal diagnosis of conjoined twins with ultrasound is now well reported from the first trimester, with detailed assessment of cardiovascular anatomy important for determining prognosis and planning management.

13. 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

Fetal medicine centres should follow the NHS England Specialised Services Clinical Reference Group for Fetal Medicine recommendations for experience.[68] Evidence level 4

14. Recommendations for future research

- The use of serial MCA PSV in screening for TAPS in women with monochorionic twins and its evaluation in a diagnostic accuracy study, with relevance to pregnancy outcomes.
- Research evaluating early versus late treatment for monochorionic twins complicated by TRAP sequence and its assessment in terms of pregnancy outcomes.

15. Auditable topics

- Prospective outcome (primary outcomes: perinatal mortality and long-term paediatric morbidity) after fetoscopic laser ablation for TTTS corrected for stage, experience of operators and severity of disease at presentation. At least one survivor in 85% of twins.
- Offer women who present in the first trimester with monochorionic twins screening for trisomy 21 (100%).
- The proportion of neurological morbidity post laser ablation for the treatment of TTTS in each fetus (less than 10%).
- Labelling of twins undertaken at first scan and followed consistently with serial scans (100%).
- The proportion of monochorionic twins who have extended fetal heart views undertaken at the midtrimester anomaly scan (more than 85%).
- The proportion of monochorionic twins who have 2-weekly ultrasound from 16 weeks of gestation (more than 95%).

16. Useful links and support groups

- National Institute for Health and Clinical Excellence. *Multiple pregnancy. The management of twin and triplet pregnancies in the antenatal period.* NICE clinical guideline 129. Manchester: NICE; 2011 [https://www.nice.org.uk/guidance/cg129].
- Royal College of Obstetricians and Gynaecologists. *Multiple pregnancy: having more than one baby. Information for you*. London: RCOG; 2016 [https://www.rcog.org.uk/en/patients/patient-leaflets/multiple-pregnancy-having-more-than-one-baby/].
- The Multiple Births Foundation [http://www.multiplebirths.org.uk/].
- Twins And Multiple Births Association [https://www.tamba.org.uk/].

