CATEGORY: BEST PRACTICE STATEMENT

Management of monochorionic twin pregnancy

This statement has been developed and reviewed by the Women's Health Committee and approved by the RANZCOG Board and Council.

A list of Women's Health Committee Members can be found in Appendix A.

Disclosure statements have been received from all members of this committee.

First endorsed by RANZCOG: March 2011
Current: Mar 2021
Review due: Mar 2024

Objectives: To provide advice on the management of monochorionic twin pregnancies.

Outcomes: Improved fetal and maternal outcomes from a monochorionic twin pregnancy.

Target audience: All health practitioners providing maternity care and patients.

Values: The evidence was reviewed by the Women's Health Committee (RANZCOG), and applied to local factors relating to Australia and New Zealand.

Background: This statement was first developed by Women's Health Committee in March 2011 and most recently reviewed in Mar 2021.

Funding: The development and review of this statement was funded by RANZCOG.

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1. Plain Language Summary

Monochorionic twins are monozygotic; that is, they arise from one fertilised ovum and commonly have a shared placenta with vascular anastomoses between the two fetal circulations. Monochorionic twins are usually diamniotic, with each twin in a separate amniotic sac. Rarely, the twins may be in a single sac (monoamniotic) or even conjoined. These configurations depend upon the stage of development at which the inner cell mass divided.

Monochorionic twins are at risk of specific complications, in addition to the increase in common pregnancy complications that occur in singleton pregnancies. They require careful surveillance by a centre with sufficient experience and expertise to recognise the onset of complications and referral for subspecialty care in the event that complications develop.

2. Summary of Recommendations

Recommendation 1	Grade
Chorionicity is a critical consideration in the management of twin pregnancies and should be determined by ultrasound and documented in all twin pregnancies prior to 14 weeks gestation[1].	Consensus-based recommendation
Recommendation 2	**Grade**
All women with monochorionic pregnancies should receive 2 weekly ultrasound surveillance for TTTS and IUGR from 16 week's gestation.[1] Ultrasound should be undertaken by a centre with sufficient experience to recognise these complications and refer appropriately if they occur.	Consensus-based recommendation
Recommendation 3	**Grade**
Ultrasound examination in monochorionic twins should include growth, amniotic fluid volume in each sac and bladder visibility. Umbilical arteryand middle cerebral artery Doppler wave forms are routine from 20 weeks but may be required at earlier scans if abnormalities are already apparent.[1, 2]	Consensus-based recommendation
Recommendation 4	**Grade**
Laser ablation of vascular connections is the recommended treatment for the majority of pregnancies with TTTS that require intervention, and referral to a laser surgery facility should be arranged - even where this may require interstate or inter country transfer.[3]	Consensus-based recommendation
Recommendation 5	**Grade**
Early referral is recommended to allow optimal treatment before the onset of severe disease and cervical shortening.	Consensus-based recommendation
Recommendation 6	**Grade**
Monochorionic twins, without IUGR or TTTS, appear to have a higher stillbirth rate than other twin pregnancies despite intensive surveillance.[4] This has led to the recommendation that these pregnancies should be delivered by 37 weeks' gestation.	Evidence Based recommendation Grade B

3. Discussion and recommendations

3.1 What are the specific complications of monochorionic twin pregnancies?

Monochorionic twin pregnancies exhibit the increased complication rates characteristic of (the more common) dichorionic twin pregnancies (such as risk of preterm birth, and increased maternal risks), but are also at higher risk of a number of specific monochorionic complications. These include:

- Twin to twin transfusion syndrome (TTTS) which will occur in approximately 15 per cent of monochorionic diamniotic (MCDA) twin pregnancies
- Selective intrauterine growth restriction (IUGR), commonly due to unequal placental sharing and velamentous cord insertion
- Death of one twin (see below)
- Twin reversed arterial perfusion (TRAP) sequence

All of these conditions contribute to an overall higher perinatal mortality and preterm birth rate for monochorionic, when compared to dichorionic twins.[4,5]

3.2 How is the chorionicity determined in multiple pregnancy?

Chorionicity is a critical consideration in the management of twin pregnancies and should be determined by ultrasound and documented in all twin pregnancies during the first trimester.[1, 6] Chorionicity is more difficult to determine accurately after chorion and amnion fusion (14 weeks gestation), with only gender discordance providing assurance of dizygosity (and therefore dichorionicity) in later pregnancy.

Recommendation 1	Grade
Chorionicity is a critical consideration in the management of twin pregnancies and should be determined by ultrasound and documented in all twin pregnancies prior to 14 weeks gestation[1].	Consensus-based recommendation

3.3 What are the management considerations for monochorionic gestations?

Women should be informed about the implications of a monochorionic pregnancy in early gestation, so that the parents can fully discuss options for managing the pregnancy and plan their future pregnancy care. In particular, they need to know the importance of notifying their obstetric care provider of acute increasing abdominal girth or breathlessness, as these may be signs of polyhydramnios due to TTTS.

Screening tests for aneuploidy have a lower detection rate in twin pregnancies than in singletons and in some centres providing cFTS, nuchal translucency alone will be used without the addition of biochemistry. Non-invasive prenatal testing has an established place for aneuploidy screening in twin pregnancies. A recent meta-analysis has confirmed a pooled sensitivity of 99% for trisomy 21 and 85% for trisomy 18 in twins, although the difference in detection between monozygotic and dizygotic twin pregnancies is a little less certain given the number of monozygotic twin pregnancies contributing to this meta-analysis was relatively low[7].

3.4 What are the recommendations in relation to surveillance for Twin-Twin Transfusion Syndrome (TTTS)?

Ultrasound signs such as discordant nuchal translucency or discordant crown-rump length (CRL) in the first trimester increase the likelihood of a later diagnosis of Twin-twin transfusion syndrome (TTTS) or IUGR but are not diagnostic, and have insufficient predictive value to be used as screening tests. These complications still occur in the presence of reassuring early scans.

Frequent ultrasound surveillance of monochorionic twins is recommended; early recognition of TTTS will facilitate referral to a tertiary centre for consideration of intervention in a timely manner. Recommended scanning schedules vary; ISUOG recommends 2 weekly scanning from 16 weeks' gestation[1]. Two-weekly ultrasound surveillance has been shown to reduce the incidence of 'late stage' TTTS at diagnosis.[8] Earlier stage diagnosis and earlier intervention is likely to improve outcomes.

For this reason, it is recommended that all women with monochorionic pregnancies should receive 2 weekly ultrasound surveillance for TTTS and IUGR following their first trimester scan (11-14 weeks) to confirm chorionicity, assess nuchal translucency and early anatomy. Ultrasound should be undertaken by a centre with sufficient experience to recognise these complications and refer appropriately if they occur. Outcomes with TTTS are optimised where there is timely diagnosis and referral to a tertiary centre for consideration of surgical therapy.

Recommendation 2	Grade
All women with monochorionic pregnancies should receive 2 weeklyultrasound surveillance for TTTS and IUGR from 16 weeks' gestation.[1] Ultrasound should be undertaken by a centre with sufficient experience to recognise these complications and refer appropriately if they occur.	Consensus-based recommendation

TTTS may take one of 2 forms:

TOPS (Twin Oligohydramnios/Polyhydramnios Sequence), affects approximately 10 per cent of monochorionic twins, and is most commonly seen in the midtrimester. This is recognised as 'classical' TTTS, with oligohydramnios, poor growth and abnormal umbilical artery Dopplers in the donor, and polyhydramnios progressing to cardiac dysfunction and cardiac failure in the recipient.

TAPS (Twin Anaemia/ Polycythaemia Sequence) affects up to 5 per cent of monochorionic twins, and 10 per cent of twins that have undergone laser therapy for TOPS. TAPS results in very slow transfusion (5-15ml/ 24 hours) from donor to recipient, so is not characterised by extreme amniotic fluid discordance and cardiac dysfunction, but by significantly discordant middle cerebral artery (MCA) peak systolic velocities, reflecting anaemia and polycythaemia in the donor and recipient, respectively. It is more common in later pregnancy, and is often recognised as 'neonatal TTTS' when very discordant haemoglobin levels are recognised at birth. Nevertheless, TAPS can also be associated with significant fetal anaemia and in utero compromise requiring treatment. For this reason, ultrasound examination in MC twins should include growth, amniotic fluid volume in each sac, bladder visibility and (after 20 weeks) umbilical artery and middle cerebral artery Doppler wave forms.[1]

Recommendation 3	Grade
Ultrasound examination in monochorionic twins should include growth,amniotic fluid volume in each sac and bladder volume. Umbilical arteryand middle cerebral artery Doppler wave forms are routine from 20 weeks, but may be required at earlier scans if abnormalities are alreadyapparent.[1, 2]	Consensus-based recommendation

3.5 How should Twin-Twin Transfusion Syndrome be managed?

Twin-Twin Transfusion Syndrome should be managed in a tertiary centre. Laser ablation of vascular connections is the recommended treatment for the majority of pregnancies with early onset, severe TTTS. Referral to a centre with facilities for laser surgery should be offered.[9] Early referral is recommended to allow optimal treatment before the onset of severe disease and cervical shortening. Amnioreduction prior to laser surgery may lead to increased membrane separation and make subsequent laser treatment more difficult. Mild TTTS (e.g. Stage 1), or late gestation disease (e.g. >26 weeks) may occasionally be managed expectantly or by amnioreduction,[10] with or without preterm delivery. Some severe cases may be managed by cord ligation of one twin, particularly if there is a fetal anomaly in one twin.[11] Ongoing surveillance post laser for TAPS (see above) is necessary post laser ablation. This applies even where the placenta has been completely dichorionised (using the Solomon technique, which has been reported to reduce, but not eliminate, the risk of post laser TAPS).[12]

Recommendation 4	Grade
Laser ablation of vascular connections is the recommended treatment for the majority of pregnancies with TTTS that require intervention, andreferral to a laser surgery facility should be arranged - even where thiswould mean interstate transfer[9].	Consensus-based recommendation

Recommendation 5	Grade
Early referral is recommended to allow optimal treatment before the onset of severe disease and cervical shortening.	Consensus-based recommendation

3.6 What are the recommendations for surveillance in a monochorionic twin pregnancy for selective fetal growth restriction?

Selective fetal growth discordance occurs in 10-15% of monochorionic twins and is diagnosed when one twin has an estimated fetal weight (EFW) <10th percentile for gestation and the EFW discordance between the twins is >25%.[1] It has been further subclassified according to the pattern of diastolic flow in the smaller twin.[13] This subclassification has important prognostic implications for the pregnancy outcome.[14] Typical gestation at recognition is 20-24 weeks, with Type II (absence or reversal of forward flow in the umbilical artery on Doppler) and Type III

(cyclical reversal and absence of forward flow in the umbilical artery Doppler) typically diagnosed earlier and with increased perinatal morbidity and mortality than Type 1 (forward flow in the umbilical artery Doppler). Management strategies are poorly defined and expert advice should be sought once selective IUGR is recognised in a monochorionic twin pregnancy, usually with referral to a tertiary centre.

3.7 What should be considered in the event of death of one of a monochorionic twin pair?

Death of one twin in a monochorionic pair may result in death or neurological disability in the survivor. These events occur around the time of the fetal death, postulated due to agonal hypotension as the blood volume of the survivor is transfused precipitously into the body of the co-twin through shared vascular communications, or possibly due to the release of thromboplastins from the deceased twin into the shared circulation. One of the advantages of laser therapy (or cord ligation) in TTTS is that it provides some neuroprotection for the surviving twin in the event of co-twin demise. Delivery of the survivor at a preterm gestation will not prevent further damage unless there is evidence of cardiotocography (CTG) abnormalities or significant fetal anaemia. Ongoing ultrasound or MRI assessment of the brain in the survivor to diagnose neurological damage secondary to hypovolaemia should be considered. MCA surveillance should be offered for the surviving twin, and intrauterine transfusion offered if the survivor has evidence of severe anaemia.

In the event of first trimester demise of one of monochorionic twins there is the potential for the development of twin reversed arterial perfusion (TRAP) sequence, where the surviving twin continues to perfuse the deceased twin's body via placental anastomoses. This rare complication is important to recognise as it carries a high risk of compromise of the surviving twin. Such cases should be managed by or in consultation with centres experienced in their management, both conservative and operative. Early referral is important as operative interventions have greater success and lower complications when performed at earlier gestations.

3.8 What is the recommended gestation at birth?

Monochorionic twins, without IUGR or TTTS, appear to have a higher stillbirth rate than other twin pregnancies despite intensive surveillance[4]. This has led to the recommendation that these pregnancies should be delivered by 37+0 weeks gestation. The mode of delivery for monochorionic twins should be individualised for each woman. Vaginal delivery is not contraindicated in monochorionic twin pregnancies[15]. As for all twins, vaginal delivery should only be undertaken where appropriate facilities exist for continuous intrapartum fetal surveillance and recourse to emergency caesarean section.

Recommendation 6	Grade
Monochorionic twins, without IUGR or TTTS, appear to have ahigher stillbirth rate than other twin pregnancies despite intensive surveillance[4]. This has led to the recommendation that these pregnancies should be delivered by 37 weeks' gestation. This has led tothe recommendation that these pregnancies should be delivered by 37+0 weeks' gestation.	Evidence based recommendation Grade B

3.9 Complex monochorionic twin pregnancies

Monochorionic twin pregnancies of higher complexity, such as monoamniotic twins, monochorionic twins with discordant anomalies or monochorionic twins within a triplet pregnancy, are even higher risk and early advice should be sought from units experienced in the management of these rare conditions.

4. References

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16. National Health and Medical Research Council. NHMRC additional levels of evidence and grades for recommendations for developers of guidelines. Canberra 2009.

5. Links to other College statements

Consent and provision of information to patients in Australia regarding proposed treatment (C-Gen 02)

Evidence-based medicine, obstetrics and gynaecology (C-Gen 15)

6. Patient information

A range of RANZCOG Patient Information Pamphlets can be ordered via the RANZCOG website

Appendices

Appendix A Women's Health Committee Membership

Name	Position on Committee
Professor Yee Leung	Chair and Board Member
Dr Gillian Gibson	Deputy Chair, Gynaecology
Dr Scott White	Deputy Chair, Obstetrics
Associate Professor Ian Pettigrew	Member and EAC Representative
Dr Kristy Milward	Member and Councillor
Dr Will Milford	Member and Councillor
Dr Frank O'Keeffe	Member and Councillor
Professor Sue Walker	Member
Dr Roy Watson	Member and Councillor
Dr Susan Fleming	Member and Councillor
Dr Sue Belgrave	Member and Councillor
Dr Marilyn Clarke	ATSI Representative
Associate Professor Kirsten Black	Member
Dr Thangeswaran Rudra	Member
Dr Nisha Khot	Member and SIMG Representative
Dr Judith Gardiner	Diplomate Representative
Dr Angela Brown	Midwifery Representative, Australia
Ms Adrienne Priday	Midwifery Representative, New Zealand
Ms Ann Jorgensen	Community Representative
Dr Rebecca Mackenzie-Proctor	Trainee Representative
Dr Leigh Duncan	Maori Representative
Prof Caroline De Costa	Co-opted member (ANZJOG member)
Dr Christine Sammartino	Observer

Appendix B Overview of the development and review process for this statement

i. Steps in developing and updating this statement

This statement was originally developed in March 2011 and was most recently reviewed in March 2021. The Women's Health Committee carried out the following steps in reviewing this statement:

- Declarations of interest were sought from all members prior to reviewing this statement.
- Structured clinical questions were developed and agreed upon.
- An updated literature search to answer the clinical questions was undertaken.
- At the February 2021 teleconference, the existing consensus-based recommendations were reviewed and updated (where appropriate) based on the available body of evidence and clinical expertise. Recommendations were graded as set out below in Appendix B part iii). This statement was approved by RANZCOG Board at their March 2021 meeting.

ii. Declaration of interest process and management

Declaring interests is essential in order to prevent any potential conflict between the private interests of members, and their duties as part of the Women's Health Committee.

A declaration of interest form specific to guidelines and statements was developed by RANZCOG and approved by the RANZCOG Board in September 2012. The Women's Health Committee members were required to declare their relevant interests in writing on this form prior to participating in the review of this statement.

Members were required to update their information as soon as they become aware of any changes to their interests and there was also a standing agenda item at each meeting where declarations of interest were called for and recorded as part of the meeting minutes.

There were no significant real or perceived conflicts of interest that required management during the process of updating this statement.

iii. Grading of recommendations

Each recommendation in this College statement is given an overall grade as per the table below, based on the National Health and Medical Research Council (NHMRC) Levels of Evidence and Grades of Recommendations for Developers of Guidelines. Where no robust evidence was available but there was sufficient consensus within the Women's Health Committee,consensus-based recommendations were developed or existing ones updated and are identifiableas such. Consensus-based recommendations were agreed to by the entire committee. Good Practice Notes are highlighted throughout and provide practical guidance to facilitate implementation. These were also developed through consensus of the entire committee.

Recommendation category		Description
Evidence-based	A	Body of evidence can be trusted to guide practice
	B	Body of evidence can be trusted to guide practice in most situations
	C	Body of evidence provides some support for recommendation(s) but care should be taken in its application
	D	The body of evidence is weak and the recommendation must be applied with caution
Consensus-based		Recommendation based on clinical opinion and expertise as insufficient evidence available
Good Practice Note		Practical advice and information based on clinical opinion and expertise

Appendix C Full Disclaimer

Purpose

This Guideline has been developed to provide general advice to practitioners about women's health issues concerning management of monochoroionic twin pregnancy and should not be relied on as a substitute for proper assessment with respect to the particular circumstances of each case and the needs of any person. It is the responsibility of each practitioner to have regard to the particular circumstances of each case.

Quality of information

The information available in the Management of Monochorionic Twin Pregnancy is intended as a guide and provided for information purposes only. The information is based on the Australian and New Zealand context using the best available evidence and information at the time of preparation. While the Royal Australian and New Zealand College of Obstetricians and Gynaecologists (RANZCOG) had endeavoured to ensure that information is accurate and current at the time of preparation, it takes no responsibility for matters arising from changed circumstances or information or material that may have become subsequently available. The use of this information is entirely at your own risk and responsibility.

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