Double Trouble: There is NO diagnosis of twins

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Learning Objectives

To recognize and report the different types of twin pregnancies on early ultrasound scans, and understand the clinical necessity for accurately evaluating the different types of twinning. To recognize the common complications of twin pregnancies.
Background

Professor Kypros Nicolaides said "There is NO diagnosis of twins. There are only monochorionic twins or dichorionic twins.\textsuperscript{1}. We frequently encounter obstetric ultrasound reports that do not clearly determine chorionicity and amnionicity in the first trimester. Monochorionic twin pregnancies in particular are "double trouble" - not only can any of the complications of singleton pregnancies occur, but up to 40% of monochorionic twin pregnancies are affected by twin-twin transfusion syndrome (TTTS), discordant fetal growth, intra-uterine or perinatal death\textsuperscript{2}. The incidence of twinning is increasing in almost all industrialised countries, including Australia\textsuperscript{1}, largely due to the use of assisted reproductive technology.
Twinning can occur as the result of fertilization of 2 ova by 2 sperm (dizygotic twins), or the dividing and separating of a single fertilized ovum (monozygotic twins). While dizygotic twins are virtually always dichorionic diamniotic, monozygotic twins can be dichorionic or monochorionic. The chorionicity and amnionicity depends on the timing of the division of the zygote (Fig. 1 on page 9). If separation occurs within 4 days of fertilization, the pregnancy will be dichorionic and diamniotic (DCDA). If separation occurs between days 4 and 7 then the pregnancy will be monochorionic and diamniotic (MCDA). Separation occurring after day 7 will result in a monochorionic monoamniotic pregnancy (MCMA). Separation occurring after day 13 is usually incomplete, resulting in conjoined twins.

Why is the type of twinning so important?

The high complication rates compared with dichorionic pregnancies result in monochorionic pregnancies needing much closer clinical and ultrasound surveillance. The Royal Australian and New Zealand College of Obstetricians and Gynaecologists recommend ultrasound surveillance for TTTS and IUGR (intra-uterine growth restriction) every 2 - 3 weeks following the nuchal translucency scan, and 2 weekly from 18 weeks gestation. In their College statement on the Management of Monochorionic Twin Pregnancy, it is also stated "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." Up to one third of all twin pregnancies are monochorionic, and one third of these will have IUGR affecting at least one twin, 10 - 15% will be complicated by severe TTTS, and the miscarriage risk (with both fetuses alive at early scan) is 12%, compared with 2% in dichorionic pregnancies.

Determining Chorionicity in the first trimester.

Chorionicity can be correctly determined in > 99% of pregnancies in the first trimester. The most reliable ultrasound feature of dichorionicity is the presence of 2 separate placentas and the presence of the lambda or twin peak sign. The lambda sign represents the projection of chorionic tissue between the 2 layers of amnion (Fig. 2 on page 9). The absence of this sign indicates the pregnancy in monochorionic (Fig. 3 on page 10). Notice also that the membrane is quite thick in the DCDA pregnancy, but thin in the MCDA pregnancy.

Determining Amnionicity in the Monochorionic Pregnancy.
The determination of amnionicity is less accurate than the determination of chorionicity. A monoamniotic pregnancy will not have a dividing membrane, but the membrane can be difficult to see and the absence of a membrane in the first trimester does not give a definitive diagnosis of an MCMA gestation. The only reliable indicator of a monoamniotic pregnancy is cord entanglement, which can only occur in the absence of an intertwin membrane (Fig. 4 on page 11). The number of yolk sacs has been used as an indicator in the past, but there are now several reports of MCMA pregnancies with more than one yolk sac, and MCDA pregnancies with only one yolk sac.

Complications of Twinning.

The risk of fetal or neonatal death in 5 times higher for twin pregnancies than singletons, and the risk of IUGR is 10 times higher. These are problems that affect both dichorionic and monochorionic pregnancies. There are several entities that are unique to monochorionic pregnancies, largely as a result of placental sharing. These include:

- twin-to-twin transfusion syndrome (TTTS)
- twin reversed arterial perfusion (TRAP) sequence
- cord entanglement in MCMA pregnancies
- conjoined twinning

The evaluation of well-being in monochorionic pregnancies relies heavily on Doppler assessment of the umbilical arteries, fetal middle cerebral arteries and ductus venosus.

The **umbilical artery** dopplers measure the degree of placental resistance, which should slowly decrease over the pregnancy as the placenta develops (Fig. 5 on page 12). A high SD (systolic-diastolic) ratio, absent or reversed flow in diastole is associated with an increase in perinatal mortality.

**Middle cerebral artery** (MCA) dopplers give a very sensitive indication of fetal brain oxygenation (Fig. 6 on page 13). Small changes in blood flow and oxygenation cause an increase in blood flow in both systole and diastole, the so-called "head-sparing" effect, which protects the fetal brain from hypoxic damage. A severely compromised fetus may "normalize" blood flow, signalling decompensation, which may thus be a trigger for delivery.

The **ductus venosus** is the main vessel carrying oxygenated blood from the placenta to the fetal heart and then fetal circulation (Fig. 7 on page 14). As fetal hypoxia increases, or fetal intravascular fluid overload occurs (such as in a TTTS recipient), the heart works
harder to maintain oxygenation, eventually leading to increased right atrial pressure. As this occurs, the flow in the ductus venosus may show deepening, or even reversal of the a wave. Subsequently, the umbilical vein may become pulsatile.

**Discordant Twins.**

There is no universal definition of twin discordance, but the most commonly reported values are a difference in fetal weights of 20 - 25%\(^\text{10}\). Discordance occurs in around 15% of twin pregnancies (Fig. 8 on page 15), and in monochorionic gestations, it is usually due to unequal placental sharing. Abnormal cord insertions are also more common in twin pregnancies than singletons, and is associated with IUGR. Velamentous or marginal cord insertions are seen in 43% of monochorionic pregnancies, and often in the donor twin in TTTS\(^\text{11}\). The ultrasound findings include:

- one twin 20% or more smaller than the other
- the amniotic fluid volume is commonly reduced around the small twin (MCDA)
- abnormal umbilical artery Doppler in the small twin: initially increased SD ratios, then absent end diastolic flow and finally reversal of flow in diastole
- abnormal middle cerebral artery dopplers: increased flow in both systole and diastole, the "head-sparing" effect
- abnormal ductus venosus dopplers

**Twin-to-Twin Transfusion Syndrome.**

Severe TTTS has a mortality of 80 - 100% if left untreated\(^\text{12}\). This haemodynamic derangement produces a donor twin, characterized by oligohydramnios, growth restriction and abnormal umbilical artery dopplers, and a recipient twin characterized by polyhydramnios, cardiac failure, hydrops and abnormal venous dopplers. The following findings suggest the diagnosis (not all need to be present):

- amniotic fluid discrepancy between sacs (Fig. 9 on page 16)
- discrepancy in size of the umbilical cords
- growth discrepancy between twins, often > 20% (Fig. 10 on page 17)
- abnormal umbilical artery and/or ductus venosus dopplers (Fig. 11 on page 18)
- inability to visualize bladder of donor twin (Fig. 9 on page 16)
- signs of cardiac dysfunction in the recipient twin

A significant discrepancy in the nuchal translucency measurements between MC twins in the first trimester is associated with an increased risk of TTTS during the pregnancy\(^\text{13}\).
**Twin Reversed Arterial Perfusion Sequence.**

TRAP affects around 1% of monochorionic pregnancies, and is more common in MCMA pregnancies than MCDA pregnancies\(^\text{14}\). The exact pathogenesis is uncertain, but the development of an artery-to-artery anastomosis in the placenta leads to reversed blood perfusion from the donor or pump twin to the acardiac twin\(^\text{15}\). The acardiac twin has a variable appearance, but typically without cranial or thoracic structures (Fig. 12 on page 19), and the upper limbs are absent or malformed (Fig. 13 on page 20, Fig. 14 on page 21). The pump twin has a high risk of cardiac failure, intrauterine death, polyhydramnios and premature delivery (Fig. 15 on page 22), with a mortality rate of around 50%\(^\text{16}\).

**Cord Entanglement in MCMA Pregnancies.**

Cord entanglement is to be expected when both fetuses share the same sac. The rate of fetal demise of MCMA twins after 20 weeks is around 15%, largely attributed to cord accidents\(^\text{14}\) (Fig. 16 on page 23).

**Conjoined Twinning.**

Conjoined twins are rare, occurring in about 1 in 50 000 pregnancies\(^\text{17}\). The majority of pregnancies succumb or are terminated, giving an incidence of less than 1 in 250 000 live births\(^\text{17}\). For unknown reasons there is a female predominance with a ratio of 3:1. Conjoined twins are classified by the site of fusion (Fig. 17 on page 24), with thoracopagus (joined at the chest) being most common. The ultrasound features include variable degrees and sites of fetal fusion (Fig. 18 on page 25), single placenta (MCMA pregnancy) and polyhydramnios. The degree of fusion can be further assessed with fetal MRI (Fig. 19 on page 26, Fig. 20 on page 27), to assist with parental counselling, delivery planning and potential for separation.

**Co-existent Twin and Molar Pregnancies.**

This rare situation has a relatively poor prognosis, with the probability of a live birth at about 40%\(^\text{18}\). Almost all reported cases are DCDA gestations\(^\text{19}\). The ultrasound findings include a normal fetus in one sac, molar change in the other sac, and theca lutein cysts in the ovaries (Fig. 21 on page 28).
Fig. 1: The development of the different types of twins.

Fig. 2: Early dichorionic diamniotic pregnancy. There are 2 separate placentas, and the gestational sacs are separated by a thick membrane showing a "twin peak" sign.

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**Fig. 3:** Early monochorionic diamniotic pregnancy. The intertwin membrane is very thin and there is no "twin peak" sign.

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Fig. 4: Video clip of a monochorionic, monoamniotic pregnancy. The 2 umbilical cords are entangled, confirming the monoamnionic status.
Fig. 5: Normal umbilical artery doppler trace.

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Fig. 6: Normal middle cerebral artery doppler trace.

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**Fig. 7:** Normal ductus venosus doppler trace.

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Fig. 8: DCDA twin pregnancy at morphology scan. There is already a significant difference in fetal size. The umbilical artery dopplers are normal for twin A, but show reversed flow for twin B.
Fig. 9: Twin-twin transfusion syndrome. Top row: the membrane is stuck to the donor twin. There is oligohydramnios in the donor twin's sac, and polyhydramnios in the recipient twin's sac. Bottom row: a bladder is not seen in the donor twin, but is present in the recipient twin.

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Fig. 10: Twin-twin transfusion syndrome. Comparative images between 2 affected fetuses, showing the degree of growth discordance. This is also reflected in the growth curves.

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Fig. 11: Twin-twin transfusion syndrome. Dopplers from an affected pregnancy. The umbilical artery dopplers show elevated SD ratios. The ductus venosus traces show deepening A waves, with reversal in twin A.

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Fig. 12: TRAP sequence. Video clip showing a scan through the uterus, to demonstrate both the normal "pump" twin and the acardiac twin.

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Fig. 13: TRAP sequence. Photograph of the acardiac twin post delivery.

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**Fig. 14:** TRAP sequence. X-ray of the acardiac twin.

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**Fig. 15:** TRAP sequence. Post-mortem photograph of an early pregnancy that miscarried.

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**Fig. 16:** Post delivery photograph of an MCMA placenta, showing a large cord knot.

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Fig. 17: Conjoined twins. Video clip of an 18 week pregnancy, joined at the face, with separate bodies.

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Fig. 18: Conjoined twins. Ultrasound images of fetuses joined at the pelvis and chest, with separate heads.

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Fig. 19: Conjoined twins. Postmortem MRI showing the spinal fusion.

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**Fig. 20:** Conjoined twins. Postmortem x-ray showing the degree of skeletal fusion.

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Fig. 21: Ultrasound images of a normal fetus and placenta, as well as a complete hydatidiform mole.

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Conclusion

After viewing this presentation, the radiologist should be able to correctly describe twin pregnancies on early ultrasound, and recognize the complications that can occur, especially in monochorionic pregnancies.
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References


