

The Obstetric Nightmare of Grey Zones

*M. El-leithy, MD**

**Registrar, Department of Obstetrics and Gynecology, Ain Shams University*

Day 1 at 10:45 Ain Shams University-Maternity Hospital (ASUMH)-Reception Room(RR).

A 23 years old patient is referred to the RR from a private clinic with the following provisional diagnosis: “PG, twin pregnancy at 30 weeks gestation with death of a co-twin”.

She has an irrelevant family & past history. She states that she has had no antenatal care but a more or less uneventful pregnancy. 2 hours before reaching the RR, she sought medical advice in a private clinic because she felt a vague lower abdominal pain. During that visit, sonographic examination revealed a twin pregnancy with death of co-twin. The patient has no history of vaginal bleeding or rupture of the membranes (ROM).

On examination, her blood pressure is 120/70 mmHg, pulse is 90 bpm, temperature is 37.1C with no albuminuria in urine dipsticks.

Abdominal examination reveals that the uterine fundus is at a level midway between the xiphisternum & umbilicus. The twins were in longitudinal lie, the first one in cephalic presentation with fetal heart rate of 140 bpm and the second one in breech presentation with undetectable fetal heart sounds (FHS). The amount of liquor is below average. Uterine contractions are absent.

The cervix is soft, posterior, not effaced & closed. The presenting part is cephalic at -3 station and with intact membranes. The patient is admitted for further management.

What are the different causes of death of a co-twin?

Day 1 at 11:00 ASU-MH - labor and delivery rooms (LDR).

The senior registrar reviews the patient and orders an ultrasonographic scan which reveals: Twin pregnancy, placenta fundal anterior grade II, 1st twin alive, longitudinal lie, cephalic presentation, liquor above average and clear, fetal biometry corresponding to 30 weeks with scalp edema and cardiomegaly with no evidence of congenital anomalies. The 2nd twin is dead, in longitudinal lie, breech presentation, with liquor volume below average and fetal biometry corresponding to 25 weeks. Both twins were females.

What type of chorionicity is expected in this case?

What are the further complications expected in this case?

Day 1 at 11:15 ASU-MH - LDR

The assistant lecturer on-duty is summoned to review the patient and advises to withdraw a venous blood sample to test for RBS, CBC, PT, PTT, FDPs & fibrinogen which all come back within normal ranges.

What is the best management for this patient?

Day 1 at 11:45 ASU-MH - LDR.

The lecturer on-duty is notified and decides on conservative management in the form of a course of intramuscular corticosteroids to enhance fetal maturity, follow up of the fibrinogen level weekly, the fetal wellbeing tests every other day and an ultrasound scan every 2 weeks for detection of any evidence of fetal brain damage and for follow up of the fetal growth pattern.

When would you consider delivering this lady?

When do you expect any signs for fetal brain damage to appear?

Day 26 at 12:00 pm ASU-MH - Antenatal ward.

At 33 weeks and 5 days, spontaneous onset of labor occurs with spontaneous ROM, both twins are delivered vaginally. The 1st twin is a living female 2200 grams with APGAR score of 4/8 having signs of respiratory distress. The neonatologist advises its admission to the NICU for further assessment. The 2nd twin is a macerated stillbirth weighing 700 grams.

How would you counsel the family about the neonatal outcome?

Comment by the Editorial Board

Death of one twin occurring after 20 weeks of gestation would be defined as a late pregnancy loss and may occur in between 0.5-6.5% of all twin pregnancies.

The current literature does not clearly indicate why one of a pair of twins may die. To elucidate this uncertainty requires knowledge of the chorionicity of the pregnancy.

Single deaths in twins occur more commonly in monochorionic pregnancies and, although it would seem likely that some will have resulted from **twin-twin transfusion syndrome**, consistent evidence for this in the form of other characteristics of that syndrome (namely hydrops of one twin and severe growth restriction of the other) is not obvious from the literature.

Other etiological factors, which include **growth restriction** due to placental insufficiency affecting only one twin, can be a feature of either monochorionic or dichorionic pregnancies.

Congenital anomalies are more common in twins and, in one series; 25% of stillborn co-twins were congenitally malformed. Interestingly, none of these pregnancies was monochorionic.

Severe maternal diseases, such as preeclampsia, may affect both fetuses but often only one of them fatally.

A common complication following the intrauterine death of one twin is the onset of labor which seems to intervene within about three weeks of the event in the majority of cases.

In monochorionic pregnancies, the death of one twin confers a risk of cerebral damage in the co-twin of about 25%. Undoubtedly, the major cause of the damage comes from the cardiovascular consequences that follow the death of one twin, resulting in sudden hypotension within the system. This may be severe and long enough to kill the remaining fetus or at least to cause vital organ damage. The damage is probably virtually immediate, occurring in minutes or hours rather than days. The pattern of damage usually involves the territory of specific vessels such as the middle cerebral artery. There may be different levels of cerebral damage, including (progressively) multicystic encephalomalacia, porencephaly, hydranencephaly and eventually microcephaly. The appearance of porencephaly in ultrasound indicates that the brain has been severely damaged and that the outcome for this baby would be very poor. The early changes involve the white matter and appear as leucomalacia. The time course for these changes is uncertain but may occur between two to five weeks after the event. Renal cortical damage is another severe insult which may affect the surviving twin.

It is important to monitor the maternal coagulation profile for early detection of any evidence of maternal DIC.

As regards the time of delivery, the problem is that the death of one twin would, in about 25-50% of cases, lead rapidly to death or damage of the remaining twin fetus. The difficulty of decision making lies in the fact that expediting delivery puts the surviving fetus in the unavoidable risks of prematurity while delaying delivery puts the fetus and the mother in the uncertain but potentially dangerous risks of morbidity and mortality.

The main damage is to the survivor's brain and this may take up to five weeks to become apparent. Since this process occurs acutely but is not detectable by imaging techniques until 2-5

weeks later, most patients would present at a stage where the damage process (if it would occur) has already started. Thus, for the monochorionic pregnancy, the best plan is to delay delivery of the live fetus for as long as possible so that any intracranial features have time to become apparent. The presence of these changes should be determined by performing a weekly ultrasound examination of the fetal brain. The use of magnetic resonance imaging (MRI) should certainly be considered but the value of this modality has yet to be established in this circumstance. In addition to evaluating the fetal condition, the importance of monitoring the maternal clotting screen should not be missed. Evidence of profound anemia in the survivor when blood sampling was undertaken by cordocentesis within 24 hours of death of the co-twin did appear to have prognostic value the eventual outcome in this group being usually poor. Conversely, the absence of anemia appeared to be associated with a good outcome.

Severe maternal diseases, such as preeclampsia or antepartum hemorrhage, are obviously indications to expedited delivery for the sake of both the mother and the surviving twin who may be at risk from the same cause which killed its co-twin.

In the remaining cases where death of a co-twin is thought to be the result of an isolated problem (e.g congenital anomaly after exclusion of similar problem in the other twin), the probably safest mode of management is close observation of the mother and the fetus until planned delivery at around 34 weeks.

Generally, the neonatal outcome when one of a pair of twins dies is not good. In one study, the intrauterine death of one of the twins resulted in 70% being admitted to special care units compared with 5.6% of twins from pregnancies in which both babies were born alive.

In an Australian series, there was a 15 fold increase in cerebral palsy in twin pregnancies in which one died, compared with pregnancies where both were live born (96/1000 versus 6.4/1000).

In a large cohort study in the UK of twins in which one twin died, cerebral palsy in the survivors had an incidence of 106/1000 in like-sex twins compared with 29/1000 when the sex between twins was different.

There is nothing more difficult for an obstetrician than having to decide between the maternal and fetal benefits, especially in conditions where grey zones shade the risk-benefit ratio.