To the Editor: Monochorionic triplet pregnancies are very rare, accounting for 3.5–4.0% of all triplet gestations and occurring in 0.001–0.004% of all deliveries. They are associated with higher risks of perinatal morbidity and mortality. Conjoined twins are a unique and rare complication of monochorionic pregnancies and occur when two identical individuals are fused through parts of their anatomy; this rare event occurs with an incidence of one in 100,000–200,000 live births. Although the presence of conjoined twins in a monochorionic triplet is exceptionally rare, it poses great challenges regarding both obstetric counseling and management.

In this study, we report an extremely rare case of conjoined twins in a spontaneous monochorionic triplet pregnancy. All available literature reporting conjoined twins in monochorionic triplets is reviewed here in order to present the impact of different management strategies on the prognosis of the nonconjoined fetus.

A 36-year-old woman, gravida 4, para 0, was referred to our Obstetrics and Gynaecology Department in a tertiary hospital with suspicion of conjoined twins in a monochorionic triplet pregnancy at 13 1/3 weeks of gestation. She had three histories of embryo damage during early pregnancy, and this pregnancy was a successful spontaneous pregnancy. After referral to our department, a transabdominal ultrasound examination was performed, and a monochorionic diamniotic triplet pregnancy with conjoined twins was observed. One amniotic sac contained a normal fetus [Figure 1a], while the other amniotic sac had a set of conjoined twins fused through the thorax and abdomen with two heads, two separate hearts, four arms, four legs, and a single umbilical cord [Figure 1b]. The patient chose selective feticide of the conjoined twins after extensive counseling, and informed consent was obtained. Microwave ablation was performed to halt umbilical cord blood flow of the conjoined twins at 16 weeks of gestation, and the procedure was technically successful. Ultrasound scan on postoperative day one confirmed the cardiac asystole of the conjoined twins and the survival of the normal fetus. The patient was then discharged, and the unexplained intrauterine demise of the normal fetus was observed 1 week postprocedure when the patient went for examinations at the local hospital. The pregnancy was terminated by induced abortion at the hospital, and we did not obtain a picture of the induced fetus. Informed consent for publication of this case report was obtained from the patient.

Conjoined twins are a rare and specific complication of monochorionic twinning. The occurrence of this phenomena may be due to the incomplete division of the inner cell mass in a single blastocyst or the reunion of two originally separate embryonic discs. Without intervention, half of the conjoined twins will die in utero with another 44% dying during the neonatal period. When monochorionic triplet pregnancies are complicated with conjoined twins, it is challenging for obstetricians to make counseling and management recommendations regarding these rare cases.

With the development and widespread use of ultrasound technologies, the accurate prenatal diagnosis of conjoined twins in monochorionic triplets is possible. Management options available to patients include the continuation of the pregnancy, termination of the pregnancy, and selective fetal reduction of the conjoined twins. When patients opt to continue without intervention, the nonconjoined fetus is at a higher risk of prematurity and intrauterine demise. Termination of the entire pregnancy, on the other hand, occurs at the expense of the normal fetus. Selective feticide aimed at terminating the conjoined twins seems to be an ideal option. However, it is associated with significant difficulties due to the existence of placental vascular anastomoses. Methods
that can occlude umbilical cord blood flow of the conjoined twins are required to accomplish selective termination. In our case of a monochorionic diamniotic triplet pregnancy complicated with conjoined twins, microwave ablation was performed to achieve cord occlusion of the target twins, and the procedure was technically successful. Unfortunately, the unexplained intrauterine demise of the nonconjoined fetus occurred 1 week postoperation.

To review the literature reporting conjoined twins in monochorionic triplets diagnosed prenatally and the impact of different management options on the prognosis of the normal fetus, a Medline search of “conjoined twins” and “triplet” was performed. To date, seven cases of monochorionic triplet pregnancies complicated with conjoined twins and a normal fetus have been reported previously. The relevant information of the seven cases and our case are presented in Table 1.

Among the recorded eight cases, 3 (37.5%) women were primigravida. One (12.5%) was achieved by intracytoplasmic sperm injection (ICSI) and the transfer of three frozen-thawed blastocysts, while the other 7 (87.5%) women had spontaneous triplet pregnancies. Chorionicity was confirmed prenatally and classified as monochorionic diamniotic in all cases. Prenatal ultrasound diagnosis of conjoined twins in a triplet pregnancy was made in the first trimester in 5 (62.5%) of the cases and in the second trimester in the other three. The types of conjoined twins included two cases of cephalopagus, two cases of thoraco-omphalopagus, and one case each of thoracopagus, xipho-omphalopagus, thoraco-omphalo-ischiopagus, and parapagus.

Regarding the management options, two patients chose to terminate the entire pregnancy. Two women opted to continue the pregnancy, and they proceeded well. In case 2, the patient gave birth to the conjoined twins and a normal female infant weighing 2041 g by cesarean section at 36 weeks of gestation. The conjoined female infants died 1 h apart at the age of 6 months. In case 7, the woman underwent elective cesarean section at 34 weeks of gestation due to premature rupture of membrane (PROM), delivering conjoined twins that died 32 min after birth, and a healthy female fetus weighing 3175 g. The remaining four women underwent selective fetal reduction with different techniques at approximately 16 weeks of gestation with poor outcomes in all cases. In case 1, 0.8 ml potassium chloride was injected into the heart of the conjoined twins, and unintentional death of the normal triplet was observed a few hours postprocedure. In case 3, endoscopic laser occlusion of the umbilical cord was performed successfully to achieve cardiac asystole of the conjoined twins, while the normal triplet died of cord entanglement at 28 weeks due to the inadvertent septostomy during the procedure. In case 6, the patient underwent radiofrequency ablation, and PROM occurred 5 h after the surgery with a spontaneous abortion 1 week later.

Adverse pregnancy outcome is higher in monochorionic than in dichorionic or trichorionic triplets. When triplet pregnancies are complicated with conjoined twins, accurate diagnosis of chorionicity is critical for management options and the prognosis of the normal triplet. Although expectant treatment is associated with a high incidence of conjoined twin demise, which leads to cerebral injuries or loss of the normal fetus due to the unidirectional blood transfusion through the placental

### Table 1: Conjoined twins in a monochorionic triplet pregnancy: Review of the literature

| Case number | Reference | Age (years) | Gravity | Parity | Conception | Placentaion | Gestational age at diagnosis (weeks) | Type of conjoining | Management | Outcome of the normal single triplet (birthweight) | Remark |
|-------------|-----------|-------------|---------|--------|------------|------------|-------------------------------------|-------------------|------------|-----------------------------------------------|---------|
| 1           | Lipitz et al., 1995[4] | 25          | 2       | 1      | Spontaneous MCDA | 16         | Thoracopagus | Selective fetal reduction at 16 weeks | IUD | Cesarean section at 36 weeks | Fetal death at 28 weeks due to cord entanglement |
| 2           | Gardeil et al., 1998[5] | 34          | 3       | 2      | Spontaneous MCDA | 13         | Thoraco-omphalo-ischiopagus | Expanscant | Live birth (2041 g) | Cesarean section at 36 weeks | |
| 3           | Sepulveda et al., 2003[6] | 41          | 3       | 2      | Spontaneous MCDA | 13         | Cephalopagus | Selective fetal reduction at 16 weeks | IUD | Cesarean section at 36 weeks | Fetal death at 28 weeks due to cord entanglement |
| 4           | Suzumori et al., 2006[7] | 33          | 4       | 2      | Spontaneous MCDA | 13         | Cephalopagus | TOP | |
| 5           | Sellami et al., 2013[8] | 20          | 1       | 0      | Spontaneous MCDA | 21         | Xipho-omphalopagus | TOP | |
| 6           | Talebian et al., 2015[9] | 38          | 1       | 0      | ICSI MCDA | 12² | Thoraco-omphalopagus | Selective fetal reduction at 16 weeks | Expanscant | Miscarriage | PROM 5 hours postprocedure |
| 7           | Mariona et al., 2017[10] | 26          | 2       | 1      | Spontaneous MCDA | 9² | Parapagus | Live birth (3175 g) | Cesarean section at 34 weeks | |
| 8           | Present case | 36          | 4       | 0      | Spontaneous MCDA | 13² | Thoraco-omphalopagus | Selective fetal reduction at 16 weeks | IUD | Cesarean section at 36 weeks | |

ICSI: Intracytoplasmic sperm injection; MCDA: Monochorionic-diamniotic; TOP: Termination of pregnancy; IUD: Intrauterine death; PROM: Premature rupture of membrane.
vascular anastomosis, the only cases of the normal triplet live birth were treated this way.[5,10] Selective termination is an ideal management in this situation which requires complete cord occlusion of the conjoined twins, and potassium chloride injection into the heart of the target fetus is not feasible. None of the normal triplets survived after this treatment as a result of different complications.[6,9] Accurate prenatal diagnosis and counseling, therefore, are very important in managing patients with this complication.

In conclusion, we report the case of conjoined twins in a spontaneous monochorionic triplet pregnancy undergoing microwave ablation for selective reduction. Review of the literature and our case suggest that selective reduction carries a high risk of poor pregnancy outcomes, and palliative care may be a reasonable management strategy. Counseling and management of conjoined twins in monochorionic triplet pregnancies present great challenges for obstetricians, and more research is required to improve the prognosis of the normal triplet.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initial will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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