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Introduction
Monoamniotic twin pregnancies present the highest risk of mortality among all twins. Optimal prenatal care (inpatient vs outpatient) for monoamniotic twin pregnancy remains controversial. In this paper we analyzed our experience in three cases of monoamniotic twin pregnancies with different clinical settings.

Material and Methods
From January 2015 to December 2016, a total of 3 cases were managed as per hospital protocol (Figure 1).

Case I
A 31- yo woman at term (38$^{2/7}$ weeks), complaining water breaking with no labour pain. US showed twin fetuses, placental chorionicity and amnionicity were difficult to evaluate. Fetal well-being was assured and c-section was decided. No neonatal morbidity. Monoamniotic pregnancy was diagnosed after delivery (Figure 2).

Case II
A 37-years-old woman with preterm (31$^{1/7}$ weeks) mono-mono pregnancy. US found no dividing membrane with adequate amniotic fluid.& cord entanglement (Figure 2). On the 7$^{th}$ day of treatment, she went to active phase of labour with fetal bradycardia; emergency CS was decided. Vigorous male babies were born weighing 1650 and 1600 gram without subsequent fetal morbidity.

Case III
A 31-years-old woman at 30$^{5/7}$ weeks of GA. US showed cord entanglement (Figure 4). On the 6$^{th}$ day she went to labour with water breaking. C-section was performed. 1400 gram and 1250 gram female were born (apgar score 6-7). Surprisingly, tight umbilical cord knotting was found during operation.

Discussion
Prenatal diagnosis is important to determine the optimal management. Cord entanglement may be undetectable as we can learn from Case III. Termination at 32-34 weeks of GA reduces perinatal mortality rate to 2.4%. In-patient monitoring with continuous monitoring at 26-28 weeks of GA has been proven effective in preventing complications. Cord accidents usually unpredictable, thus, intermittent monitoring sometimes inadequate. Despite little experiences, our protocol was proven effective in preventing fatal neonatal morbidity and mortality.

Conclusions
We recommend in-patient care starting at 28 weeks of gestational age. Termination of pregnancy is considered at 32-34 weeks of gestational age by c-section. Cases must be treated at the highest healthcare facility with available monitoring & emergency CS.

References
UMBILICAL CORD ENTANGLEMENT IN MONOCHORIONIC-MONOAMNIOTIC TWINS PREGNANCY

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INTRODUCTION

Monochorionic-monoamniotic twin is a type of monozygotic twin with potentially high morbidity and mortality. The incidence of this condition varies considerably from 1/1,650-93,734 pregnancies.

Chorionicity greatly affects the outcomes of twin pregnancies, if monochorionic twin is accompanied by monoaomi, complications of umbilical cord entanglement may increase fetal mortality.

Previous literatures showed that perinatal mortality of monochorionic twin pregnancies reaches 70%. But after the development of electronic fetal monitoring and neonatal care, recent data indicates that perinatal mortality decreases to 10-20%. Entanglement and knotting of the umbilical cord are the cause of death in monochorionic twins for > 50%. Ultrasonography (USG) plays an important role in assessing such complications. In patient management with close fetal monitoring, continued with elective cesarean delivery may be chosen.

CASE

A 37-year-old female, multiparous (G3P2A0) referred from a local hospital with complaints of labour pain and rupture of membrane (ROM) an hour prior to admission. Twin pregnancy was confirmed during routine obstetrician visit at 13-14 weeks gestation, with suspicion of monochorionic-monoamniotic pregnancy. Her two previous pregnancies were delivered in the midwife at term without any complications. No family history of twins or use of assisted reproductive methods was noted.

The patient's general condition was good. Obstetric examination revealed a twin pregnancy with head-breech presentation, regular uterine contractions, and 2 cm cervical dilatation accompanied by rupture of the membrane. USG examination at the emergency room (ER) showed both fetuses in good condition, with estimated fetal weight of 1812 grams and 1745 grams. Unfortunately, monochorionic pregnancies have not yet been excluded. Laboratory tests showed no signs of infection. Patient was diagnosed with G3P2A0 31-32 weeks twin with threatened preterm labour (TTL) + ROM.

We decided to take conservative treatment to maintain pregnancy by tocolytic administration (oral nifedipine according to the South Australian protocol), corticosteroid injection for lung maturation (dexamethasone 12 mg every 24 hours for 2 days, intramuscular), and diuretics.

5th day: USG evaluation was repeated, and monoamniotic pregnancy was confirmed with a suspicion of cord entanglement. Doppler velocimetry study was within normal limits. Team decided to continue conservative care with close monitoring of the ER, as well as the preparation of the neonatal intensive care unit (NICU). After 24 hours, repeat ultrasound evaluation was performed, and cord entanglement without a true knot was confirmed (Figure 1). Close monitoring was continued by using daily cardiotocography (CTG) and Doppler study.

7th day: bradycardia in both fetuses, not improve with intrauterine resuscitation. Green code cesarean section was then undertaken. Both infants were born with good condition (Apgar scores 8-9 and 7-8), with birth weight of 1650 grams and 1600 grams. Gross placental examination showed monoamniotic placenta with extensive cord entanglement without true knot (Figure 2). Patient was discharge in the 1st postnatal day after 24 hours of observation and monitoring in the NICU.

DISCUSSION

Monoamniotic twins can be diagnosed in the first trimester using USG assessment. In our case, monochorionic-monoamniotic pregnancy has been detected since 13-14 weeks of gestation. No medical record presence and patient was referred in 31-32 weeks gestation. Our patient has no TTT, TAPS, TRAP, conjoint twin and growth abnormality. Cord entanglement complicates 71% of all monoamniotic twins and is responsible for 50% of fetal deaths in one or both fetuses. Fetal weight discordance is more likely in monochorionic pregnancy compared to diamniotic pregnancy (1 in 5 pregnancies versus 1 in 15 pregnancies). In our case, this growth discordance was not noted.

Sudden deaths in monochorionic pregnancies are often caused by the umbilical cord entanglement but can be prevented by proper monitoring and premature delivery planning. This is consistent with our case, where fetal distress was unexpectedly present and abrupt. However, close in-hospital monitoring may prevent fetal mortality.

The existence of umbilical cord entanglement between the two new fetuses was noted during inpatient treatment in Sanglah Hospital from USG examination as shown in Figure 1. After complete examination, it was decided to continue conservative treatment until 34 weeks gestation with daily doppler study and CTG monitoring and type planned delivery is delivery with cesarean section at 34 weeks gestation.

In general, the principle management of monoamniotic pregnancy is elective cesarean delivery at 32 weeks of gestation after corticosteroid treatment for infant lung maturation. In our case, the multidisciplinary team decided to conserve the pregnancy until 34 weeks; due to limited neonatal care resources. Nevertheless, in the course of treatment, fetal distress was noted and green code cesarean section was performed. No adverse maternal and neonatal outcomes were experienced.

SUMMARY

Monochorionic monamniotic twin pregnancy is a rare type of monogygotic twin pregnancy which poses greatest risk for infant morbidity and mortality. USG is a major diagnostic modality for detecting umbilical cord abnormalities in monochorionic-monoamniotic pregnancies. Continuous observation in hospital setting using daily USG and CTG will provide better outcomes than out-patient approach. The preferred method of delivery is cesarean section at gestational age > 32 weeks after corticosteroid administration for fetal lung maturation. Early diagnosis and appropriate management will reduce infant morbidity and mortality in this population.

Reference

Twin heart(s): A case report of thoracoomphalopagus twins with conjoined heart.

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- We present a rare case of conjoined twins thoracoomphalopagus type diagnosed prenatally by ultrasound. We report echocardiographic features of a rare anatomy and physiology of two hearts with common atrial chamber and fused ventricle. Our case adds to a pool of reported cases of thoracopagus twins in the local setting. The study reports 15 cases and outcomes of thoracopagus conjoined twins from 1974-2016 in the Philippines. Cases of thoracopagus conjoined twins were reviewed and thoracopagus with shared heart were deemed inseparable and incompatible with survival. The management of thoracoomphalopagus twins with shared heart is difficult and challenging in poor resource setting.

- Keywords: conjoined twins, fetal echocardiography, surgical separation, thoracoomphalopagus, ultrasound
TRAPS cases in dr. Soetomo hospital: Expectant vs Active management

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INTRODUCTION

Congenital anomaly is the third leading perinatal mortality after prematurity and malnutrition. In Indonesia, congenital anomaly occurs 2071 in dr. Soetomo hospital it is 2.7% from total deliveries. The accuracy of prenatal diagnosis (around 71%) using sonography was contributed to the increasing diagnosis of congenital anomaly. We classify cases by the possibility of correction: 43% correctable (Anencephaly); 36% postnatal correction (Cleft palate); 21% intrauterine correction (PVTT, TTTS, TRAPS) 4% multiple pregnancy cases complicated with TTTS or TRAPS. This study aims to report cases of TRAPS managed expectantly and actively managed.

Case Report

Case-1: Female, G5 came on 24/25 weeks. One baby was normal with pericardia effusion and one baby with acardiac features. No abnormality on Doppler study. On 27 weeks, we performed photocoeagulation at Harapan Kita Hospital. We identify the equator line and did coagulation on six anastomosis points on the equator line. On the seventh attempt of coagulation, bleeding occurred and the procedure was ceased. We irrigate with normal saline 5500 cc until the amniotic fluid is clear. On the second fetus, we performed amnioreduction around 500 cc.

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<th>Second fetus</th>
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<tr>
<td>BPD</td>
<td>27/59 mm</td>
</tr>
<tr>
<td>AC</td>
<td>23/50 mm</td>
</tr>
<tr>
<td>FL</td>
<td>3.20 mm</td>
</tr>
<tr>
<td>EFW</td>
<td>846 g</td>
</tr>
<tr>
<td>Pericardial effusion, bladder outline</td>
<td>No</td>
</tr>
<tr>
<td>DVT</td>
<td>0.8 cm</td>
</tr>
<tr>
<td>PI</td>
<td>1.71</td>
</tr>
<tr>
<td>SDS</td>
<td>0.8</td>
</tr>
<tr>
<td>PVS</td>
<td>40.8</td>
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Table 1 The prenatal sonographic evaluation

We performed weekly surveillance and found remarkable condition to the donor fetus. The first baby developed well with no pericardia effusion. We decided to continue the pregnancy until term.

On the 32 weeks the patient complained of watery discharge and then had cesarean delivery (CD). The baby weighed 1600 g, no asphyxia and the second baby weighed 650 g with remnant of cranium features, micromelia.

The mother was discharged three days postoperative, and during third month follow up, the living baby weighed 2000 g with no complication.

Case-2: Female, G1 came on 37/38 weeks with no history of proper antenatal care before. She only had once sonography on 20 weeks stated with intrauterine cyst. The first baby had normal feature and no hydrops, the other was acardiac mass with remnant of umbilical cord inside.

The incidence of multiple gestation continues to increase accounting for more than 3% of all live births in the United States. The perinatal mortality rate for twins is significantly higher than for singletons at all gestational ages. Twin reversed arterial perfusion (TRAP) sequence or acardiac twinning is a monochorionic multiple gestations in which one twin has an absent, rudimentary, or nonfunctioning heart. The incidence is estimated for 1% of monochorionic twin pregnancies or 1 in 35,000 births.

The criteria that may suggest the need for intervention include the abdominal circumference measurement in the acardiac twin greater than or equal to that of the pump twin, polyhydramnios with maximum vertical fluid pocket greater than 8 cm, abnormal Doppler indices, hydrops in the pump twin or the rapid enlargement of the acardiac twin. If impending cardiac failure are present at an early gestational age, consideration for an invasive procedure to interrupt the vascular supply to the acardiac mass. The survival rate post-coagulation 70-80% and increase until 90% if the intervention occurred in 16-18 weeks.

In the absence of such poor prognostic features, we can do expectant management with serial sonographic evaluation may be reasonable.

Conclusions

The goal of antepartum management of a pregnancy complicated by the TRAP sequence is to maximize outcome for the structurally normal twin. In case 1 we had pericardia effusion and then required photocoeagulation of the anastomoses immediately. In case 2 we had no effort on detection and therapy but the acardiac twin could grow bigger without disrupting the normal twin. We concluded in this report that in the absence of poor prognosis (fetal hydrops, cardiac failure, polyhydramnios) the management of TRAPS could be expectant rather than active.

References

A CASE SERIES OF MONOCHORIONIC, MONOAMNIOTIC TWINS

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& ANALYN T. NGO-YU, MD, FPOGS, FPSMFM (Consultant Adviser)

INTRODUCTION: Monochorionic, monoamniotic pregnancy is a rare type of placentation occurring in 1-5% of all monozygotic pregnancies. The objective of presenting this case series is to compare the antenatal surveillance and management done on each patient including timing of delivery and demonstrate which among them has the best outcome.

CASE I:
E.C., 25yo G1P0 at 29 2/7 weeks AOG who was referred due to an ultrasound result of twin pregnancy. She was diagnosed with monochorionic, monoamniotic placentation during a congenital anomaly scan done in the institution. She advised early admission for close fetal monitoring, but opted follow-up as outpatient basis. Weekly BPS with Doppler, Biometry every 2 weeks and daily fetal movement counting were advised. She was admitted due to labor pains on her 31 6/7 weeks AOG. However, on auscultation only one fetal heart note was noted. An ultrasound was done and confirmed an intrauterine demise of one twin. Hence, she underwent emergency cesarean section delivering to Twin A, stillborn girl weighing 1620 grams, and Twin B alive girl weighing 1675 grams, with birth length of 40 cm, Apgar score: 3, 6, 7 and Ballard’s score 30-31 weeks. Inspection of the placenta showed monochorionic, monoaamniotic placenta with three-vessel cord with multiple cord entanglement. Twin B was admitted at NICU with the diagnosis of Birth Depression with Respiratory Distress Syndrome but was discharged recovered on the 35th hospital stay.

CASE II:
This is a case of J.D., 20 yo, Gravida 2 Para 1 (1001) at 29 5/7 weeks AOG who was referred due to an ultrasound result of twin pregnancy. She was diagnosed with monochorionic, monoaamniotic placentation during a congenital anomaly scan done in our institution. She was advised early admission for close fetal monitoring, but opted follow-up on outpatient basis. Weekly BPS with Doppler, Biometry every 2 weeks and daily fetal movement counting were advised. She was admitted due to labor pains on her 30 weeks AOG. However, on auscultation only one fetal heart note was noted. An ultrasound was done and confirmed an intrauterine demise of one twin. Hence, she underwent emergency cesarean section delivering to Twin A-alive preterm girl, cephalic, with birth weight of 1530 grams, birth length of 40 cm, Apgar score 6, 8, Ballard’s score 30 weeks and Twin B-stillborn girl with birth weight of 1205 grams. Inspection of the placenta showed monochorionic, monoaamniotic placenta with three-vessel cord with triple tight nuchal cord coil on Twin B. Twin A was admitted at the NICU for Respiratory Distress Syndrome, Prematurity and Low Birth Weight but eventually died on the 14th day of life.

CASE III:
This is a case of G.D., 30yo G3P2 (1101) at 24 4/7 weeks AOG referred due to an ultrasound result of twin pregnancy. She was diagnosed with monochorionic, monoamniotic placentation during a congenital anomaly scan done in the institution. Patient was counseled on the possible complications of her condition and was subsequently admitted at the Maternal High Risk ward. Part of the serial maternal surveillance includes BPS with Doppler studies every 3 days, Biometry every 2 weeks and NST twice daily. Referral to the newborn medicine was done and the plan was for possible termination of pregnancy anytime a non-reassuring fetal status develops. The plan of an elective cesarean section at 34 weeks age of gestation was discussed with the patient and consultation with the neonatologist was done in preparation for delivery. On her 39th hospital stay, patient underwent an elective cesarean section at 34 1/7 weeks AOG delivering to alive preterm twins, girl A- 34 weeks by BS (1855g, 31 cm), AS 8.9 and twin B- 34 weeks by BS (2110g, 31 cm) AS 8.9. Inspection of the placenta showed monochorionic, monoamniotic placentation, with three-vessel cord with beginning cord entanglement (3X). Both twins were admitted at the NICU with the diagnosis of Prematurity, Twin B- to consider Respiratory Distress Syndrome. Both twins were discharged improved.

CONCLUSION: Monoamniotic pregnancies should ideally be managed as in-patients in a tertiary hospital to provide better and increased frequency of antenatal surveillance with a multidisciplinary team of experts. Decision on the appropriate timing of delivery must be made as a team, weighing the benefits and the risks of preterm delivery to both fetuses. Proper counselling of both parents must be done to elucidate them, not only in the prognosis and survival of these fetuses but also in the occurrence of unexpected intrauterine demise. Elective cesarean section may be performed at 32 to 34 weeks as long as the fetal conditions are reassuring. Further and larger studies on monochorionic, monoamniotic pregnancies can be done to arrive at a standardized protocol in managing these patients.
Perinatal Outcomes for Monochorionic Monoamniotic Twins Complicated with Twin Reversed Arterial Perfusion Sequence

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OBJECTIVE

To discuss the perinatal management and outcomes for monochorionic monoamniotic (MCMA) twins complicated with twin reversed arterial perfusion sequence (TRAPS).

METHOD

A retrospectively study was conducted to evaluate perinatal management and outcomes of MCMA twin pregnancy complicated with TRAPS at West China Second University Hospital from Jan. 2012 to Jul. 2017.

RESULTS

Six case of MCMA twins complicated with TRAPS were identified at 13+2-23 gestational weeks. Two cases underwent induce labor for intrauterine death of pump twin. Three cases underwent conservative management and delivered a healthy pump twin at 31+3, 37+5, 39+5 gestational weeks. One case was treated with bipolar cord coagulation of acardiac twin at 23+5 weeks and delivered a pump twin at 31 gestational weeks and the survival baby died 3+ months after birth.

CONCLUSION

MCMA twins complicated with TRAPS is very rare. To get a better perinatal outcome, early diagnosis, regular antenatal examination, close fetal surveillance are all critical, and selective termination should be done if necessary.
Perinatal Outcomes and managements for monochorionic diamniotic triplet pregnancies
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OBJECTIVE
To study perinatal management and outcomes for monochorionic diamniotic (MCDA) triplet pregnancies.

METHOD
A retrospective study was conducted to evaluate perinatal management and outcomes of MCDA triplet pregnancies at West China Second University Hospital from January 2012 to September 2017 including maternal age, pregnancy type, prenatal examination, delivery method, perinatal outcomes and mortality. Meanwhile, placental and umbilical cord examination was given after delivery to identify diagnosis of MCDA triple pregnancy and umbilical cord entanglement.

RESULTS
A total of 4 cases of MCDA triplet pregnancies included in our study were primipara and spontaneously conceived with age between 20 and 28 years. Two cases had the regular prenatal examination in our hospital and were identified diagnosis at 13+4, 18+5 gestational weeks, respectively. They had caesarean section at 32+3, 32+5 gestational weeks and gave birth to six health and survival babies. Meanwhile, No.3 and No.4 didn’t have regular prenatal examination. The diagnosis of MCDA triplet pregnancies were made after delivery. Both of them were transferred to our hospital due to preterm premature rupture of membranes at 28 weeks, 21+3 weeks respectively. No.3 had caesarea section due to maternal factor at 28+3 gestational weeks and newborns were died in 6 days after birth (1 newborn), 23 days after birth (2 newborns). No.4 gave birth to three dead fetus at 21+3 gestational weeks for inevitable abortion and were complicated with malformations (one fetus had cleft lip and palate, the other one were acardiac).

CONCLUSION
MCDA triplet pregnancies are very rare. To get a better perinatal outcome, we recommend that MCDA triplet pregnant should have early diagnosis, regular antenatal care, close prenatal monitoring, and communication.
Chromosomal abnormalities and chromosomal microarray analysis in twins with structural anomalies.

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OBJECTIVES: To evaluate the type and detection rate of chromosomal abnormalities in twins with structural anomalies and compare the incremental yield of CMA with conventional karyotyping.

STUDY DESIGN: This was a retrospective analysis of 596 twin pregnancies in which one or two fetuses had congenital structural anomalies in a center over an 11-year period. Twin pregnancies were categorized by chorionicity and amnionicity as monochorionic monoamnionic (MCMA), monochorionic diamnionic (MCDA) and dichorionic diamnionic (DCDA). Chromosomal abnormalities detected by both G-banding karyotyping and CMA were analyzed by chorionicity and types of structural anomalies.

RESULTS: Of 617 fetuses (596 pairs) with structural anomalies, 465 (75.4%, 465/617) fetuses performed invasive prenatal testing and cytogenetic results were obtained. The detection rate of overall chromosomal abnormalities in DCDA fetuses (25.2%, 77/306) was higher than that in MCMA (2.4%, 1/41) and MCDA (14.4%, 17/118) fetuses (P < 0.05). Monosomy X (5.9%, 7/118) was the most frequent chromosomal abnormality diagnosed in MCDA group, while trisomy 21 (7.2%, 22/306) was the most frequent in DCDA group. CMA revealed pathogenic CNVs in 2.3% (5/222) of structurally abnormal twins who had normal karyotypes. Both in DCDA (56.6%, 30/53) and MCDA (50.0%, 7/14) fetuses with ultrasound anomalies, hydrops fetalis had highest detection rates of chromosomal abnormalities, of which 67.6% (25/37) were Turner syndrome (45, X). Discordant karyotypes were identified in 11 out of the 101 MCDA pairs in which invasive cytogenetic results of both twins were obtained.

CONCLUSION: Dichorionic twins with structural anomalies are at higher risk of chromosomal abnormalities than monochorionic twins; this may be associated with increased maternal age and the use of assisted reproductive technologies in dichorionic twins. CMA increase the detection of genomic imbalances in these twins but the incremental yield of CMA in twins was lower than the previously reported rate in singletons with structural anomalies.
Background
This scenario that our patient present was a serious and rare complication of monochorionic twin pregnancy, the incidence is 1/35000 in all pregnancy. The diagnosis could be made around the end of first trimester from sonography.

Case
This 39-year-old female is pregnant with monochorionic diamniotic twin via artificial reproductive method. This pregnancy was diagnosed "Twin-twin transfusion syndrome (TTTS)" at 8th gestational age with one of the embryo acardia. Due to persistent enlargement of the acardiac fetus, fetal scope ablation was performed. The level II sonography and amniocentesis showed normal results of the “pump twin”. The patient underwent amniocentesis two more times on 2017/08/10 and 2017/08/30 due to polyhydromnio induced frequent uterine contraction. By this time, there was no signs of fetal hydrops noted but mild cardiomegaly was noted on the "pump twin". Due to preterm premature rupture of membranes, preterm labor and small for gestational age of the pump twin, Cesarean section was performed on 2017/09/26(GA32+4) after discussing with the family, Rinderon administered and with neonatologists’ standby. The pump twin was born with APGAR score 4(1')→8(5'), birth body weight:1080g and in stable clinical status after delivered.

Discussion
The earliest definition was made in 1983 by Van Allen et.al, which clarified this scenario as reversed blood flow resulted from anastomosis of vessels. The twins shared a placenta and the recipient twin received the hypoxia blood flow from the pump twin. The blood flow of the recipient twin run through the umbilical arteries which tend to flow to lower limbs and resulted in poorly developed upper trunk. A third of the acardia fetuses have an abnormal karyotype. Around 9% of the pump twins have trisomy. [1]

The classification of conditions of acardia:

- **Acardius anceps**
  The head is poorly formed, but trunk and limbs are fairly well developed
- **Acardius acephalus**
  The head is absent but the trunk and limbs are more or less well developed. The most frequent variety, responsible for 60% to 75% of cases
- **Acardius acormus**
  Rare type of acardia in which there is development of the fetal head only. The head is usually directly attached to the placenta via a cord arising in the cervical region.
- **Acardius amorphous**
  The defect consists of an irregular, skin-covered mass without a proper form of a fetus.

Weisz et al. present that cases with the following factors suggest aggressive treatment: abnormal Doppler pattern or the recipient twin’s abdominal circumstance is equal or larger than the donor[3]. The most common causes of death of the donor twin are anemia, heart failure or preterm labor. Treatment could be started since GA13, including, fetoscopic laser coagulation, radiofrequency ablation, amniocentesis, or high-intensity focused ultrasound(HIFU)[4]. Lewi et al., found spontaneous flow arrest in 21% of cases during 16-18 weeks, which gives the hope of conservative management in these cases

Overall, early diagnosis could help attending physician to follow up closely and decided whether intervention needed.

Fig. 1 The acardiac fetus and the placenta it shared with the pump twin.