



Case report

Twin reversed arterial perfusion sequence managed by bipolar cord coagulation and amniopatch: Case report

Dudy Aldiansyah^{a,b,*}, Muara P. Lubis^{a,b}, Desy Handayani^a, Edwin M. Asroel^{a,b}, Melvin N. G. Barus^{a,b}, Bugis Mardina Lubis^{c,d}

^a Department of Obstetrics and Gynecology, Faculty of Medicine, Universitas Sumatera Utara, Medan, 20155 Indonesia

^b Fetal Therapy Unit, Division of Maternal Fetal Medicine, Department of Obstetrics and Gynecology, H. Adam Malik General Hospital, Medan, Sumatera Utara, 20136 Indonesia

^c Department of Pediatric, Faculty of Medicine, Universitas Sumatera Utara, Medan, 20155 Indonesia

^d Division of Neonatology, Department of Pediatric, H. Adam Malik General Hospital, Medan, Sumatera Utara, 20136 Indonesia

ARTICLE INFO

Keywords:

TRAP sequence
Bipolar cord coagulation
Amniopatch
Amnioinfusion
Case report

ABSTRACT

Introduction and importance: Twin Reversed Arterial Perfusion (TRAP) Sequence is a rare condition that occurs in monochorionic twin pregnancies, resulting in the coexistence of a normal “pump” twin and an acardiac twin. Indonesia is implementing fetal therapy centers that are starting to treat fetal cases such as TRAP Sequence.

Case presentation: An 18 years old pregnant woman with monochorionic diamniotic pregnancy was detected by ultrasonographic examination. A live fetus with normal fetal heart rate estimated fetal weight was 661 g, and consistent with 25 w gestational age. Additionally, an acardiac twin with polyhydramnios and EFW of 1829 g. Bipolar cord coagulation, amniopatch, and amnioinfusion were performed. The patient's condition was stable and managed closely.

Clinical discussion: This is the first procedure reported in Indonesia for TRAP sequence case. It reduces cardiac strain on the pump twin and increases the chance of survival.

Conclusion: The patient was diagnosed with TRAP Sequence and bipolar cord coagulation to interrupt blood supply to the non-viable twin, amniopatch with autologous platelet concentrate followed by cryoprecipitate amnioinfusion were reported for the first time in Indonesia.

1. Introduction

In Twin Reversed Arterial Perfusion (TRAP) Sequence, the twin in charge of pumping blood to both fetal circulations is at risk of heart failure that may threaten its life [1]. The incidence of this syndrome, which is caused by TRAP sequence, is 1/35,000 in all pregnancies and 1/100 in monochorionic twin pregnancies [2]. A recent study estimated the incidence as 2.6% of monochorionic twin pregnancies and 1/9500–11,000 pregnancies [3]. Hypo-oxygenized blood coming via umbilical arteries feeds, although not sufficient, the acardiac twins' lower extremities and part of the abdomen through the common iliac artery and abdominal aorta. When the blood reaches the upper extremities and cranium of the acardiac fetus, the oxygen saturation which is already low decreases even more and causes insufficient tissue and organ development [2]. In TRAP syndrome, there is mostly arterioarterial anastomosis in the placenta and one of the twins is acardiac and obtains

its blood from the other pump twin. Mortality is 100% in the acardiac twin and almost 50% in the pump twin due to heart failure and prematurity [3]. In the TRAP sequence, the high risk of death for the normal pump twin has led to the development of a wide range of intrauterine interventions to improve outcomes [4].

2. Case presentation

A healthy pregnant woman, 18 y.o, G1P0A0, complained of shortness of breath for the past 3 days. It was her first pregnancy and the patient had no history of medication during the first trimester, no genetic history of the family about the congenital anomaly, and no psychosocial disorders history. History of uterine contraction, amniotic fluid leakage, and blood slime was not found. The patient came to our Fetal Therapy Unit with TRAP Sequence, Mono Chorionic Diamniotic (MCDA), intrauterine pregnancy (25–26) w gestational age. Obstetric

* Corresponding author.

E-mail address: dudy.aldiansyah@usu.ac.id (D. Aldiansyah).

<https://doi.org/10.1016/j.ijscr.2022.106893>

Received 11 January 2022; Received in revised form 24 February 2022; Accepted 27 February 2022

Available online 1 March 2022

2210-2612/© 2022 The Authors. Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license

(<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

physical examination found a fundal height of 4 fingers below the xiphoid process. Fetal heart rate was 134 bpm, and uterine contraction was not found. Complete blood count showed Hb (9.7 g/dL) and Ht (29%), leucocyte (9.960/ μ L), and thrombocyte (220.000/ μ L). From ultrasonography examination, a live fetus with a fetal heart rate was 144 bpm and fetal weight 661 g MVP of pump twin was 5.65 cm. Abnormal ductus venous waveform of the pump twin indicated a sign of impending cardiac failure. The EFW of the acardiac twin was 1829 g, which was calculated from the formula ($V = \pi/6 * \text{length} * \text{anterior-posterior diameter} * \text{width}$ in cm^3 or g). The acardiac pump twin ratio was 0.96, calculated from the ratio of the abdominal circumference of the acardiac twin and pump twin. MVP of the acardiac twin was 6.15 cm. Cervical length was 35 mm. Therapies for the patient were O_2 4 lpm, IVFD RL and MgSO_4 40% (30 mL) for tocolytic, 12 dpm, ketoprofen supp 2×4 , folic acid 1×1 , ferrous sulfate 1×1 , dexamethasone 6 mg/12 h for 48 h, and transfusion of PRC 2 bags. The patient was offered bipolar cord coagulation, amniopatch with blood products (thrombocyte concentrate and cryoprecipitate) after the bipolar cord coagulation did. Bipolar cord coagulation, amniopatch, and amnioinfusion were performed. Amnioinfusion was performed because the placenta was almost completely anterior so we make a pocket for a window to reach the acardiac's twin umbilical cord.

The operation was performed by the obstetrician's team-leading by dr. Dudy Aldiansyah, Sp.OG(K), M.Ked(OG). The procedure was carried out with skin disinfected prepared with 10% iodopovidone, and short-term antibiotic prophylaxis was administered (ampicillin 1 g intramuscularly). 20 mL of 1% xylocaine without adrenaline were administered locally down to the myometrium and the procedure was ultrasound (GE Voluson® P6 BT 16, Seongnam, GE, Ultrasound Korea, LTD) guided. A small skin incision was loaded in a performed before the percutaneous introduction of a nine French sharp trocar 10 French catheters (Cordis Avanti® + Sheath Introducer, 402-605×, Johnson & Johnson). This was a 10 French disposable catheter originally designed for vascular access for central vein catheterization with a silicone valve system for a leak-proof seal. We did the Saldinger technique to insert 10 French disposable catheters (Cordis Avanti® + Sheath Introducer, 402-605×, Johnson & Johnson). Amnioinfusion of 300 mL was carried out because the placenta was posterior, hence we created a window to reach the acardiac twin umbilical cord [5].

The second insertion of 10 French disposable catheters was inserted in a different place. Bipolar cord coagulation was performed. Increased 3-mm disposable bipolar coagulation forceps (Karl-Storz Take-Apart Forceps, 2.4 mm) were inserted under ultrasound control. The procedure was aimed at coagulating the cord at the placental or abdominal insertion. The easier access point was chosen. The insertion of the trocar was at least 10 cm of acardiac twin distant from the expected point of coagulation to let the forceps open and grasp the cord. A longitudinal portion of the cord was exposed to the bipolar forceps, which was introduced with its jaws perpendicular to the cord. The correct grasp of the cord was checked by pulling the bipolar forceps and checking the passive movements of the cord and the fetus when the abdominal insertion was grasped. After grasping the cord initial coagulation was performed using 20 W for 10 s. The procedure was deemed successful when echogenic bubbles were seen coming from the cord and the cord itself appeared hyperechogenic. In the absence of bubbles, the power was increased in 5 or 10 W increments to a maximum of 50 W. When the appropriate power was obtained, the coagulation was determined using 1 or 2 min, without releasing the cord. The cord was coagulated for an average of 10 min, even when the flow stopped earlier. Confirmation of occlusion was also provided by the absence of detectable color Doppler flow in the distal part of the cord, with at least 2 min of persistent asystole. The cardiac activity of the co-twin was checked by ultrasound during the procedure, immediately after the procedure, 2 and 12 h after the procedure, peak systolic velocity measurements in the middle cerebral artery (MCA-PSV) were also recorded to detect fetal anemia MCA was 20.02 cm/s with 0.65 MoM [6].

After that, an amniopatch was performed. The procedure was performed under local anesthesia and with the use of a 20 or 22 G needle to access a safe location under ultrasound guidance. The protocol was used thrombocyte concentrate and cryoprecipitate. Infusions of 40 mL of thrombocyte concentrate and 30 mL of cryoprecipitate were injected into the amniotic sac. During and after the procedure, fetal heart rate and the accumulation of amniotic fluid were monitored by ultrasound. The cervical length after the procedure was 28 mm. Further, follow-up consisted of ultrasound evaluation from 24, 48 to 72 h post-procedure, and weekly or fortnightly thereafter. A 3-day after the procedure, ascites were found, and the patient was followed closely. A 2-week later ascites were decreased [7].

The patient was hospitalized and observed regularly in the ward for 2 weeks. The patient agreed to do intervention after extensive counseling and informed consent. The patient's condition was stable, then we performed serial ultrasonography examination every week and the result was improvement and reduction of hydrops fetalis. A 3 weeks later, estimated 28 weeks of gestational age, the patient came to the hospital within labor. The patient was delivered to the delivery room with cervical dilatation of 8 cm. Cesarean section was performed, and the twin baby was born with a pump twin who was male, body weight was 1300 g, body length was 38 cm, and Apgar score was 6/7, while the monster baby was IUFD. Mother and pump twin condition were stable after surgery. Blood flow in the Doppler central blood flow to the acardiac twin fetus and hydrops in the pump fetus disappeared after 2 weeks of follow-up. All the steps of the operation procedure are in accordance to [8], and the pictures during the procedure are shown in Figs. 1, 2, 3, and 4.

The surviving infant was monitored for up to 6 months after birth with increased weight and height as shown in Table 1. In Echocardiogram examination, no abnormalities were shown in the infant.

3. Clinical discussion

This is the first case of bipolar cord coagulation and amniopatch in the TRAP sequence reported in Indonesia which still developing a few fetal therapy centers, one in Jakarta, and one in Medan. We still lack cases because of a lack of referral system, and some patients came late to the hospital. Before, we had treated TRAP sequence cases with fetoscopy laser coagulation, but it was not successful. This is the first case report in Indonesia that informs the success in the management of TRAP Sequence. The incidence has been quoted as 2.6% of monozygotic twins and between 1 in 9500 and 11.000 pregnancies in a recent study [9]. TRAP should be suspected in monozygotic twin pregnancies when one fetus appears anatomically normal and the other lacks apparent cardiac structures and activity [10]. A recent study demonstrated that TRAP sequenced diagnosed in the first trimester had a fatal outcome after expectant management, with losses occurring at ≤ 16 weeks. The number of referred cases was significantly lower and the gestational age at detection was higher. Therefore, a subset of cases might have been missed or died at the local institution without being referred to the tertiary center [11].

Organogenesis defect due to anastomosis in the placenta in the early period of gestation is suggested in pathogenesis. Blood circulation between twins is provided either via artery-artery or vein-vein anastomosis on the placental surface [3]. Acardiac acephalus, acardius acornus, acardius amorphous, and acardius myelacephalus types are reported. The most common anomaly is acardiac acephalus, in which the heart and head are absent, like in the present case [3]. In acardius acephalus (60%–75% of cases), the cranial and thoracic structures are missing but there is the preservation of the lower limbs, as seen in this case. In acardius amorphous (20% of cases), there is an amorphous mass of tissue with no recognizable features. Acardius anceps (10% of cases) is the most differentiated form, with a partly formed head, thorax, and abdomen. Acardius acornus (5% of cases) demonstrates only cephalic structures [12].

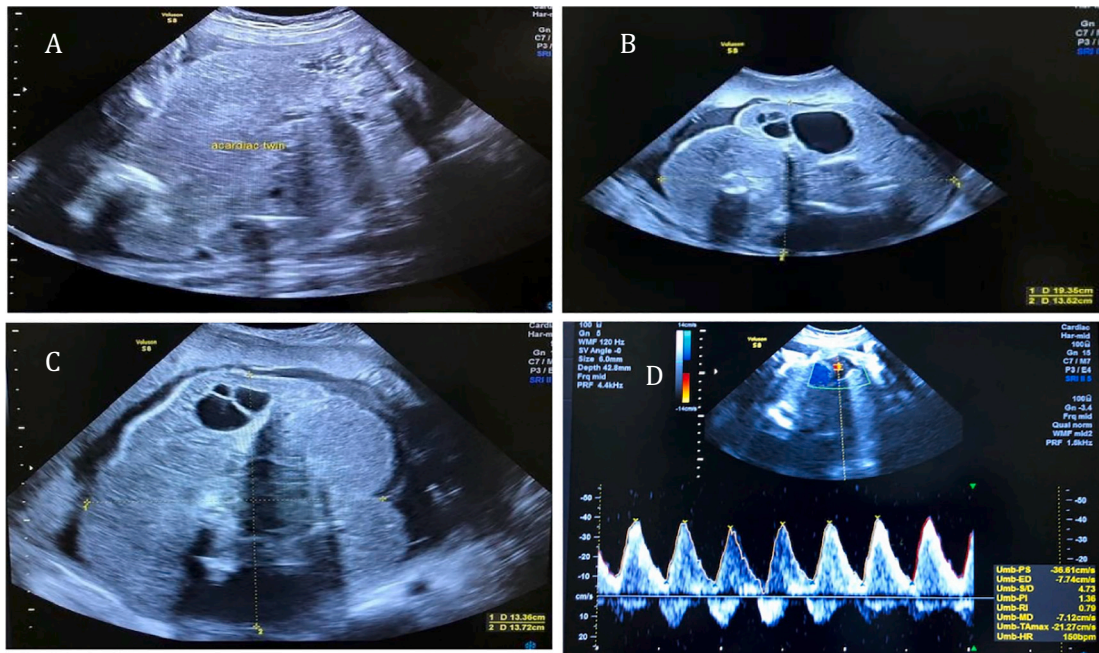


Fig. 1. A, B, and C are acardiac twins, D is the Doppler of ductus of the acardiac twin.

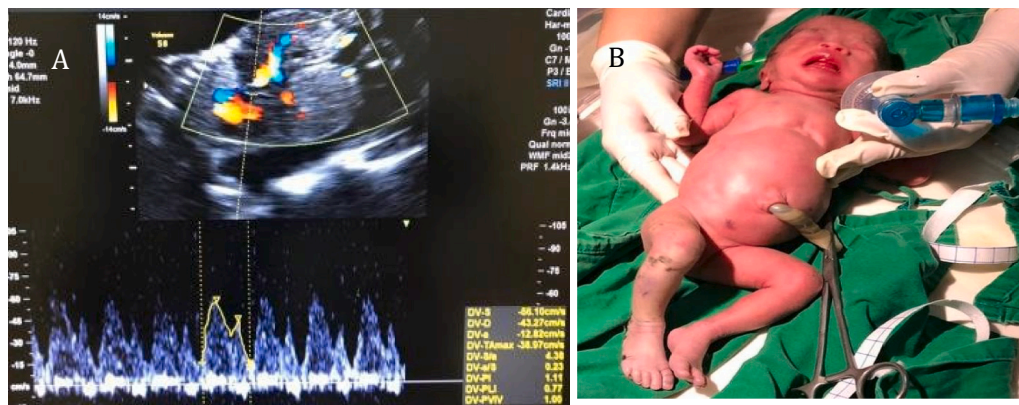


Fig. 2. Pump Twin (A) ductus venosus of the pump twin and (B) the pump twin when delivered.

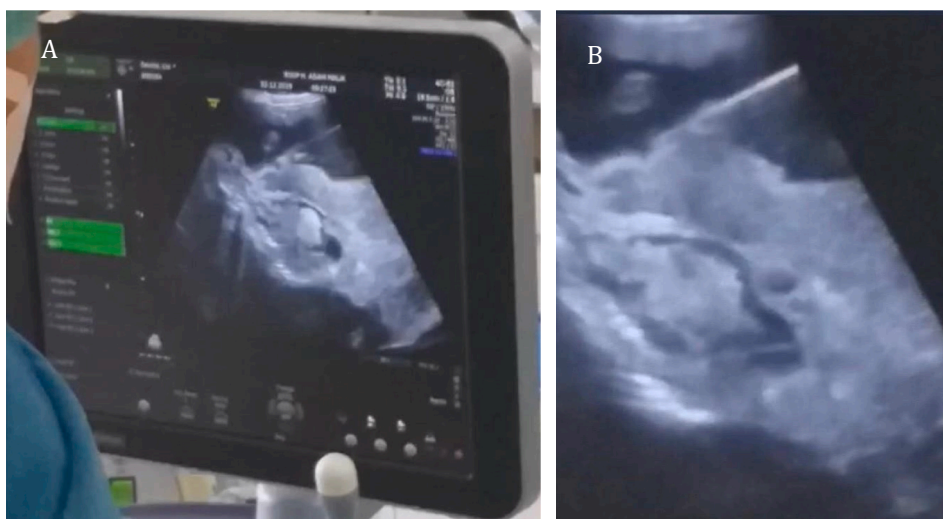


Fig. 3. Bipolar cord coagulation procedure in the operation theatre room.

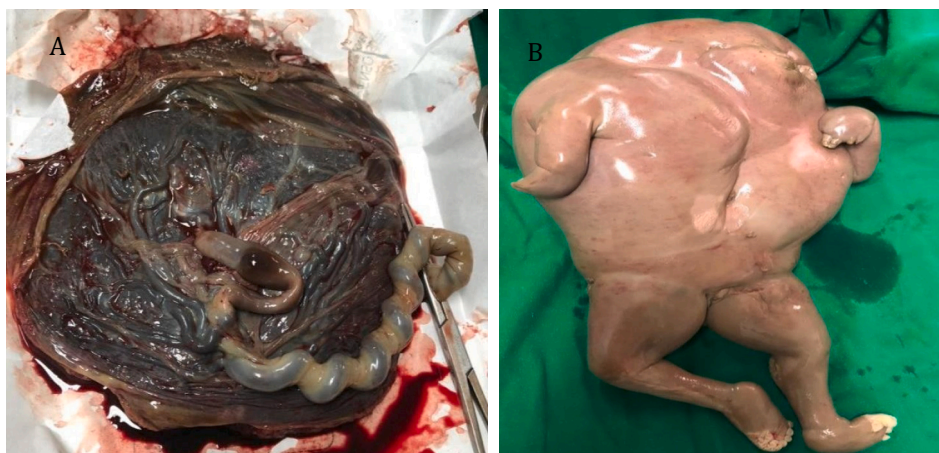


Fig. 4. Acardiac twin and the placenta: (A) placenta of twin and (B) pump twin delivered by C-Section.

Table 1

The measurement of weight and height of the surviving infant in 6 months.

	1st month	2nd month	3rd month	4th month	5th month	6th month
Weight	2400 g	3200 g	4100 g	5300 g	6300 g	7100 g
Length	45 cm	50 cm	54 cm	57 cm	62 cm	68 cm

Umbilical cord anomalies are seen in up to 97% of cases. The most common cord anomaly is a single umbilical artery of the acardiac twin; however, a single umbilical artery of the pump twin, marginal or velamentous placental insertion, cord entanglement, and juxtaposed cord insertions are also present in some cases [7]. Arterial flow, which is expected to return to the placenta, passes through the pump fetus directly to the receiver fetus. The lower extremities can develop when the hypoxic fetus receives even limited oxygen and food resources. However, as in this case, if hypoxia is deep and starts in the early weeks of gestation, organ development is not likely to occur [2]. While the chromosome is generally normal in the pump fetus, chromosomal anomalies might be detected in the acardiac fetus. Mortality of the pump fetus increases depending on heart failure and prematurity caused by polyhydramnios [2].

More recently, with the widespread introduction of an ultrasound scan at 11–13 weeks' gestation, as part of early screening for fetal aneuploidies, many cases of TRAP sequence are now diagnosed during the first trimester of pregnancy. Despite this earlier diagnosis, intrauterine therapeutic interventions were delayed after 15 weeks gestation, because the risks of amniorrhexis and miscarriage were considered to be high if carried out before obliteration of the celomic cavity [4]. Usually, if the ratio of the weight of the acardiac to the pump twin is greater than 0.7, the risk of preterm birth is higher [9]. Doppler study reveals the presence of blood flow in it and flow of blood will be towards the acardiac twin. The acardiac phenotype ranges from well-developed lower extremities, pelvis, and abdomen to a tissue mass-like teratoma that is not readily recognizable as fetal parts. After the diagnosis of the TRAP sequence, a ratio of the weight of the acardiac twin to pump twin should be considered to predict fetal prognosis [10].

Conservative or symptomatic treatments cannot resolve the underlying problems and always lead to poor results. Poor outcomes have also been proved when such pregnancies are treated conservatively by serial amniocentesis, with less than 30% survival rate of at least one twin and a high incidence of neurologic sequelae as well as possible relevance to postnatal heart diseases. Therefore, management targeted at the etiology should be employed to improve the outcomes. Cord occlusion has been reported effective in treating TRAP sequence [1].

Pregnancies with TRAP Sequence have the options of expectant

management, antenatal intervention, and delivery. At present, there is a controversy regarding elective versus treatment of TRAP sequence. Spontaneous flow arrest in 21% of cases during 16–18 weeks gives hope of conservative management in these cases. In one series of TRAP sequences, the perinatal mortality rate for conservatively managed pump twins was very high due to the development of heart failure and preterm delivery. However, these survival rates relate to pregnancies with TRAP sequence diagnosed at or after 16 weeks gestation [10].

Subsequently, a series of other less invasive intrauterine interventions aiming to arrest the circulation of the acardiac twin was introduced. Intravital Radiofrequency Ablation (RFA) in recent years has become the method of choice than interstitial laser [13]. RFA has been done for TRAP sequence cases and is superior to minimally invasive umbilical cord coagulation modalities. Livingston et al 2007 showed that RFA had a high success rate with a survival rate of 80% [14]. Other methods are insertion of cord coils, ligation with or without transection of the umbilical cord, endoscopic laser coagulation of placental anastomoses between the pump and acardiac twins, endoscopic laser coagulation, and endoscopic or ultrasound-guided monopolar or bipolar diathermy of vessels within the cord supplying the acardiac twin and, more recently, ultrasound-guided ablation of intra-vital vessels by injection of alcohol, monopolar diathermy, laser or radiofrequency. These interventions are usually carried out at or after 16 weeks of gestation. The survival rate of about 80% following interruption of the blood supply to the acardiac twin is higher than the reported rate of 45% with expectant management [4]. Since there is extensive experience in bipolar cord coagulation, it is the most popular cord occlusion technique. Bipolar cord coagulation is the most frequently used technique for selective termination in monozygotic twin pregnancies with a survival rate of 71–85% and a live birth rate of 80–83% [9]. There are studies that however report survival of 80 to 90% of pump twins in pregnancies that underwent in utero coagulation and ablation procedures [9].

Another technique for the management of TRAP sequence is microwave ablation. This technique uses energy that causes dipole molecules (water) within the near field to create heat, leading to thermocoagulation of the surrounding tissue. A focused burn with more energy can be applied to the target tissue, thus allowing for high tissue temperatures and faster ablation times. Microwave ablation does not require grounding pads, reducing the risk of injury [15].

High Intensity of Focused Ultrasonography (HIFU) was also found to give benefit because it is less invasive and no need for uterine puncture. From Seo K et al. 2019, the total fetal survival was 67% and the efficiency rate was 83% after HIFU therapy. It has been suggested that HIFU is beneficial because it reduces the cardiac load of the pump fetus. HIFU has three benefits: few fetal complications, treatment is not limited by

the position of the placenta, and treatment can be performed before 16 weeks of gestation [16].

Iatrogenic preterm prelabour rupture of membrane (iPPROM) remains one of the most complications after a fetoscopic procedure with the risk of fluid leakage being 1–2%. It depends on such as trocar size and surgical duration. Several authors have reported their experience after a fetoscopic procedure with amniopatch. An overall live birth rate of 65% after fetoscopy-related PPROM and 66% after needle procedure-related PPROM after amniopatch procedure [6]. In addition to these unique problems, severe selective intrauterine growth restriction (IUGR) and discordant structural anomalies are the other challenging issues in monochorionic pregnancies, which might necessitate selective termination of the affected fetus to preserve the healthy co-twin [12].

4. Conclusion

A patient is diagnosed with TRAP Sequence and bipolar cord coagulation to interrupt blood supply to the non-viable acardiac twin, amniopatch is performed to make some pocket window to reach cord insertion on acardiac twin because the placenta is mostly anterior. This reduces cardiac strain on the pump twin and increases the chance of survival. This is the first procedure and reported Indonesia for TRAP sequence case.

Funding sources

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Ethical approval

This case report was conducted with the approval of the Ethics Committee of the Faculty of Medicine, Universitas Sumatera Utara, Indonesia, No.1315/KEP/USU/2021.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Dudy Aldiansyah M.D.; Muara P Lubis M.D.; Edwin M Asroel M.D.; Melvin N.G Barus M.D.; and Bugis Mardina Lubis M.D., contribute to patient care, compilation, and revising manuscripts critically.

Research registration

None.

Guarantor

Dr. Dudy Aldiansyah, Sp.OG(K), M.Ked(OG).

Declaration of competing interest

There is no conflict of interest.

Acknowledgment

The authors thank the supervisors and the Department of Obstetrics and Gynecology residents for their guidance and assistance in conducting this case report. In addition, the authors also thank all parties involved in this case especially Desy Handayani, Hariadi Supanto, Dizalia Ananda, Hayatun N Namira, and Reni Hayati. And also, best kind and regard to Jose L Peiro, MD, PhD, MBA, Professor of surgery-University of Cincinnati Medical College for the advice in this manuscript.

References

- [1] H. Zhi-ming, F. Qun, Y. Yong-zhing, L. Yan-min, C. Jun-hong, C. Yong-zhen, Z. Yi, C. Min-ling, Fetal reduction by bipolar cord coagulation in managing complicated monochorionic multiple pregnancies: preliminary experience in China, *Chin. Med. J.* 123 (5) (2010) 549–554.
- [2] E. Yildirim, Spontaneous triplet pregnancy and trap sequence, case report, *BMC Pregnancy Childbirth* 19 (2019) 328.
- [3] H.Y. Yildizdas, E. Umit, S. Mete, Y. Gulseren, S. Huseyin, O. Ferda, Twin reversed arterial perfusion syndrome in a monochorionic monoamniotic twin pregnancy, *Turk. J. Pediatr.* 59 (2017) 724–727.
- [4] P. Chaveeva, C.P. Leona, S. Alexandros, K. Przemek, H.N. Kypros, Optimal method and timing of intrauterine intervention in twin reversed arterial perfusion sequence: case study and meta-analysis, *Fetal Diagn. Ther.* 35 (2014) 267–279.
- [5] R. Robyr, Y. Masami, Y. Ville, Selective fetocide in complicated monochorionic twin pregnancies using ultrasound-guided bipolar cord coagulation, *BJOG* 112 (2005) 1344–1348.
- [6] J. Richter, A. Henry, G. Ryan, P. DeKonick, L. Lewi, J. Depres, Amniopatch procedure after previable iatrogenic rupture of membrane : a two center review, *Prenat. Diagn.* 33 (2013) 391–396.
- [7] M.M. Lanna, M.A. Rustico, M. Dell'avano, V. Schena, S. Faiola, D. Consonni, A. Righini, B. Scelsa, E.M. Ferrazzi, Bipolar cord coagulation for selective fetocide in complicated monochorionic twin pregnancies: 118 consecutive cases at a single center, *Ultrasound Obstet. Gynecol.* 39 (2012) 407–413.
- [8] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, for the SCARE Group, The SCARE 2020 guideline: updating consensus Surgical Case Report (SCARE) guidelines, *Int. J. Surg.* 84 (2020) 226–230.
- [9] E.M. Ziki, Z.M. Chirenje, M.G. Madzlyire, There are studies that however report survival of 80 to 90% of pump twins in pregnancies that underwent in utero coagulation and ablation procedures, *Pan Afr. Med. J.* 32 (2019) 54.
- [10] S. Dubey, M. Verma, P. Goel, R. Punia, Twin reversed arterial perfusion: to treat or not? *J. Clin. Diagn. Res.* 11 (1) (2017) QD05–QD07.
- [11] C. Berg, D. Holst, M.R. Mallmann, I. Gottschalk, U. Gembruch, Early vs late intervention in twin reversed arterial perfusion sequence, *Ultrasound Obstet. Gynecol.* 43 (2014) 60–64.
- [12] R. Has, I. Kelolioglu, A.C. Esmer, H. Ermis, O. Durai, Y. Dogan, C. Yasa, O. Demir, H. Yumru, A. Yildirim, L. Ibrahimoglu, A. Yuksel, Bipolar cord coagulation in the management of complicated monochorionic twin pregnancies, *Fetal Diagn. Ther.* 36 (2014) 190–195.
- [13] J.L. Peiro, T. Crombleholme, Errors in fetal surgery, *Semin. Pediatr. Surg.* 28 (3) (2019) 143–150.
- [14] J.C. Livingston, F. Yen-Lim, W. Polzin, J. Mason, T.M. Crombleholmes, Intrafetal radiofrequency ablation for twin reversed arterial perfusion (TRAP): a single-center experience, *AJOG* 197 (4) (2007) 399.e1–399.e3.
- [15] C.D. Stephenson, L.A. Temming, R. Pollack, D.A. Iannitti, Microwave ablation for twin reverse arterial perfusion sequence: a novel application of technology, *Fetal Diagn. Ther.* 38 (2015) 35–40.
- [16] K. Seo, K. Ichizuka, T. Okai, S. Dohi, M. Nakamura, J. Hasegawa, R. Matsuoka, S. Yoshizawa, S.I. Umemura, M. Nagatsuka, A. Sekizawa, Twin reversed arterial perfusion sequence using high intensity focused ultrasound, *Ultrasound Obstet. Gynecol.* 54 (2019) 128–134.