Case Report

Twin Reversed Arterial Perfusion (TRAP) Sequence: A Case Report and Review of Treatment

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Acardiac twin or twin reversed arterial perfusion (TRAP) sequence is a rare unique complication of monozygotic multiple pregnancy. In this disorder, there is a normally formed donor (the pump twin) who has features of congestive heart failure (CHF) as well as a recipient (the acardiac twin) who lacks a well-defined a heart structure. Also evident are other structures, namely the TRAP sequence from pump to acardiac fetus via single artery-to-artery and vein-to-vein anastomoses directly between the two cords or indirectly on the chorionic plate. Overall, the perinatal mortality rate for the pump twin is 35-55%. Prenatal diagnosis and prognosis factors can be examined through ultrasound. The optimal management of acardiac twin pregnancies is controversial. The expected treatment of acardiac anomaly presently relies on maximizing the chance of term delivery and preventing CHF in the healthy pump twin or interrupting vascularization between the two twins. This article reported the experience of acardiac twin management in Thammasat University Hospital and reviewed the current knowledge of the condition, prenatal diagnosis, prognosis factor and management options focusing on conservative management compared to invasive treatment.

Keywords: *Acardiac twin, TRAP sequence*

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A rare and unique medical complication of monozygotic twins is known as "Acardiac Twin". Acardiac twin has many names that have been derived according to the main anatomical abnormality (holoacardius or pseudoacardius), the apparent pathophysiological basis (Twin reversed arterial perfusion sequence or TRAP sequence) or the total dependence on the normal co-twin (chorioangiopagus parasiticus). Overall incidence is 1:35,000 births; 1/100 of monochorionic twin[1] and 1/30 of monochorionic triplets[2]. One third has a monochorionic monoamniotic placenta and two-thirds are associated with a monochorionic-diamniotic placenta[3].

In this disorder, a normally formed donor (the pump twin) who has features of congestive heart failure (CHF) as well as a recipient (the acardiac twin) who is deficient in a well defined heart structure and other structures. The etiology and pathophysiology underlying of acardiac anomaly are not absolutely understood. The two main hypotheses attempting to explain the origin of acardiac anomaly are:

1) Abnormal placental vasculature leading to circulatory reversal with subsequent alteration in cardiac development;

2) Abnormal cardiac embryogenesis occurring as a primary event.

These hypotheses were supported by the publication of Van Allen et al[4], which reported 14 cases of acardiac twin and clarified the underlying pathophysiology of this condition, namely the twin reversed arterial perfusion (TRAP) from pump to acardiac fetus via single artery-to-artery and vein-to-vein anastomoses directly between the two cords or indirectly on the chorionic plate. Arterial blood flows in a retrograde way from pump twin toward, rather than away from, the affected acardiac fetus. Moreover, the umbilical vein of the parasitic acardiac fetus returns blood into the placenta and back to the pump twin. Within the arterial perfusion pressure of the pump twin exceeds that in the acardiac fetus, which thus receives reverse blood flow of deoxygenated arterial blood from its co-twin and nutrient-poor. Blood bypasses the placenta, enters the circulation of the acardiac fetus and passively follows a course through the iliac arteries

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at subnormal pulse pressure to perfuse preferentially and predominantly to the lower part of the body. Thus, only the lower body was perfused, and disrupted growth and development of the upper body results. This circulatory event explains why the lower limbs of an acardiac fetus are better formed than its more cephalad structures. Because of this vascular connection, the normal donor twin must not only support its own circulation but also pump its blood through the underdeveloped acardiac recipient. This may lead to cardiomegaly and high-output CHF in the normal twin[5].

Concurrent with technological advancements in diagnostic and therapeutic capabilities in the field of obstetrics, numerous reports have been published to describe acardiac anomaly in different clinical settings, possible prognostic factors that may influence its management, including techniques and outcomes of various modes of treatment. The purpose of this article is to review the current knowledge of the condition, prenatal diagnosis and management options focusing on conservative management compared to invasive treatment.

Case Report

A 25-years-old Thai gravida 1, para 0 was referred from antenatal care unit to fetal diagnosis and therapy center, Thammasat University Hospital at 20 weeks' gestation. A twin pregnancy with single intrauterine fetal demise was suspected. Detailed ultrasonogram revealed that the surviving twin was structurally normal with a fetal biometry of 20 weeks' gestation. After completion of ultrasonogram, monochorionic twin pregnancy was diagnosed.

Twin A (pump twin) was in cephalic presentation and had normal structure compatible with 20 weeks gestation. Twin B (acadiac twin) was grossly malformed at cephalic part. There was moderate size (20x10 cm) of septate cystic hygroma at the thoracic area. Cardiac structure or organ above thorax could not be identified as presented in Fig. 1. This acadiac twin showed both well-developed lower extremities with club feet. On Doppler study, the direction of blood flow in umbilical arteries of twin B was in the opposite direction while twin A had a normal pattern.

The intervention of blood supply occlusion to acardiac twin was deliberated. This case was sent to Faculty of Medicine Siriraj Hospital, Mahidol University for intervention. After well informed consent to couples, needle guided with color Doppler ultrasound was inserted to pregnant abdomen at 3 days after first being diagnosed. Amniotic fluid was collected for karyotype study then radiofrequency ablation was done at iliac artery of acardiac twin. This operation was successful. This pregnant women was sent back to follow-up at Thammasat University Hospital one week after operation. Normal karyotype (46XX) was reported. Elective cesarean section was performed at 36 weeks and 3 days gestation with female newborn 3,500 grams Apgar 9, 10 and acardiac twin mass 25 grams (Fig. 2).

Prenatal diagnosis

Obviously, the acardiac fetus had no chance of survival. The survival of pump twin is the aim of prenatal diagnosis. Early diagnosis of this condition plays an important role in case-management and aids preparing treatment, and can improve the chance of the pump twin's.

Ultrasonography is the safe and noninvasive procedure for malformation detection. It can be used to predict the fetal prognosis and assist maternal counseling.

Acardiac twin diagnosis by ultrasonography was first introduced by Lehr and DiRe[6]. Presently, the diagnosis of acardiac twin is solely based on using ultrasound. Ultrasonographic finding showed absent of identifiable cardiac pulsation in one twin, poor definition of the head, trunk and upper extremities, deformed lower extremities, marked and diffuse subcutaneous edema and abnormal cystic areas in the upper body part of the affected twin.

In addition, Smith et al[7] reported that the acardiac fetus was associated with severe anomalies and anencephalic condition. Club feet, absent of upper limbs and toe were often seen. Limb movements are

Fig. 1 Ultrasonography showed monochorionic diamniotic twin and cystic hygroma of acardiac twin.

Fig. 2 The normal pump twin with acardiac fetus after RFA (radiofrequency ablation).

Fig. 3 Acardiac fetus after RFA (radiofrequency ablation).

sometimes visualized in the acardiac fetus by a result of the pump twin movement. Furthermore, more than two-thirds of cases have two-vessel umbilical cord. Lower aortic branch of these vessels supplied remnant structures.

Even though the meaning of acardiac twin was a no heart twin, to the contrary, the presence of cardiac motion does not exclude the diagnosis, as it may result from a rudimentary heart or transmitted pulsation from the cardiac function of the pump twin[8]. The pump twin is typically structurally normal. Nonetheless, hydropic anomalies or malformation occurred in 9% of cases. Congestive heart failure (CHF) was usually developed due to increased cardiovascular demand, resulting in preterm delivery[1]. In addition, Pinet et al[9] reported approximately 10 percent of major malformations; including cardiogenic defects gastroschisis and skeletal abnormalities. The pump twin may develop CHF with cardiomegaly, hepatosplenomegaly, pericardial and pleural effusions, ascites and polyhydramnios. In diamniotic, acardiac twin cases, polyhydramnios and oligohydramnios were found in the pump and acardiac twin, respectively. In year 1994, Healey[10] reported that 70 percent of acardiac fetus sac with a functional urinary tract developed

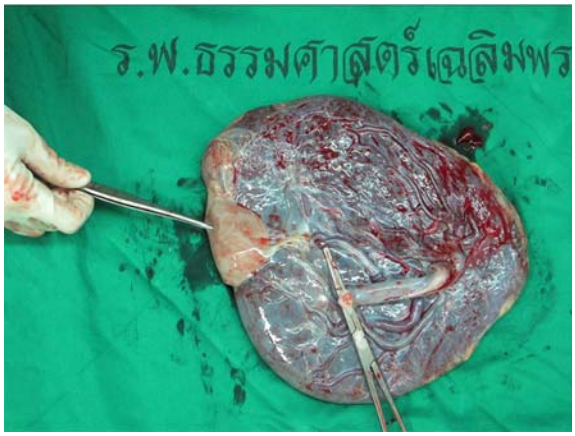

Fig. 4 Monochorionic diamniotic twin with acardiac twin.

polyhydramnios.

Nowadays, Doppler ultrasound remains the standard method of an acardiac twin diagnosis. The first application of color Doppler ultrasound in the prenatal diagnosis as well as documentation of the reversed arterial perfusion pattern was reported by Pretorius et al in 1988. The definitive diagnosis of acardiac twin can be easily established with color flow imaging at presentation by simply demonstrating the

presence of blood flow within the abnormal fetus. More detailed examination of the blood flow pattern in these cases will reveal a paradoxical direction of arterial blood flow towards, rather than away from the acardiac fetus, and from caudal to cranial in this twin's abdominal aorta. In addition, Ishimatsu et al[11] presented that reverse flow in the umbilical artery from Doppler ultrasonography was an acardiac twin diagnostic finding. It helped to determine the site of vascular connection and assess circulatory health of the pump twin. Moreover, Shbire et al[8] reported that Doppler velocimetry of the umbilical cord in acardiac twin showing a markedly abnormal peak systolic-to-end diastolic velocity ratio. Small resistive index differences are associated with poor outcome.

Recently, three dimension ultrasonography (3D U/S) and magnetic resonance imaging (MRI) were used to confirm the diagnosis and precisely establish the extent of fetal malformations. Size and the atypical, grotesque appearance of the acardiac twin were the limitation of a study by 3D U/S and MRI[12,13].

After exclusion of chromosomal aberrations and malformations in the pump twin, close sonographic follow-up may be initiated. Serial sonography with color Doppler sonography should be performed every 1-2 weeks to assess growth status in each twin and CHF indicators in the pump twin. The most important differential diagnosis of acardiac fetus is single intrauterine death of one grossly abnormal monochorionic twin. Before the color Doppler era, the TRAP sequence diagnosis was made early in the second trimester of pregnancy with fetal demise occurrence. Similarly, a vanishing twin was the consequence of the cardiac activity absence in the first trimester[14]. Serial ultrasonography was used to evaluate the continued growth of the presumed dead fetus in twin pregnancies. The subsequent scans are often the first clue to making the correct diagnosis. Other less common differential diagnosis included umbilical cord and placental teratoma. These conditions were easily differentiated. An acardiac twin is always separate with the absence of spinal development, some evidence of body organization and attached umbilical cord, whereas the teratoma was completely disorganized[3,15].

Prenatal prognosis factors

Overall, the perinatal mortality rate of the pump twin was 35 and 55 percent based on the 184 and 49 cases of acardiac twin reviews by Healey and Moore et al[1,10], respectively. Perinatal mortality came from preterm labor because the hydramnios and CHF were strongly associated conditions in the pump twin. Moreover, the TRAP-sequence was complicated by polyhydramnios, preterm labor, in utero CHF and intrauterine demise of both twins at percentage of 51, 75, 28 and 25, respectively[10].

The acardiac twin's growth evaluation was the best management in antenatal period. The occurrence of morbidity and mortality rate of pump twin were strongly related to the ratio of the acardiac and pump-twin's weight (APTW). If the APTW ratio was above 70 percent, the incidence of preterm delivery, hydramnios and pump-twin CHF was 90, 40 and 30 percent, respectively. Compared to the APTW ratio less than 70 percent group, the complications were 75, 30 and 10 percent, respectively. If APTW ratio was above 50 percent, the predictive value of preterm delivery and the pump-twin death were 71 and 45 percent, respectively[1]. Furthermore, recent a study by Minakshi et al[16] showed if the APTW ratio was less than 25 percent, the prognosis was excellent without further therapy. In contrast, risk of CHF increased to 94 percent as the APTW ratio was above 50 percent. CHF developed in all pump twins when the APTW ratio exceed than 70 percent. However, it was impossible to make an accurate estimate of the acardiac fetal weight. The biparietal diameter of the acardiac twin was not available andits femur was shorter than normal. A possible estimation was the comparison of the abdominal perimeter ratio of the acardiac to pump twins. The standard prorate ellipsoid formula (i.e. length of acardiac x abdominal diameter divided by 2) was frequently used. Data from the literatures suggested that estimation of the relative weights in acardiac fetus provided prognostic information regarding outcomes, assisted in maternal counseling and determined optimal management.

Treatment

The optimal management of acardiac twin pregnancies is controversial. The treatment of acardiac anomaly presently relied on maximizing the term delivery's chance, prevention, CHF treatment in the healthy pump twin and interruption of the twin vascularization. Currently, management options included observation with close antepartum surveillance (conservative treatment) and surgical interventions. Alternatively, a variety of therapeutic options had been proposed. Early therapeutic effort was initiated to prevent and treat the complications. Expectant management was the alleviation of

polyhydramnios to relieve maternal discomfort and prolong pregnancy. The repeated amniocentesis was used to control the amniotic fluid volume[17]. Maternal indomethacin therapy used to alleviate polyhydramnios as well as to prevent premature labour[18]. Maternal digoxin administration was used to treat fetal CHF in the pump twin[19]. Before starting the medical or intervention treatment, the underlying vascular causes should be investigated and managed first. Treatment without the underlying vascular problem correction may postpone the definitive treatment.

Conservative treatment

Many previous reports of conservative treatment before 2003 showed that the neonatal mortality rate of the pump twin was extremely high 50-75%[1,4,20-22]. In contrast, Sullivan et al[23] suggested that the neonatal mortality of pump twins may be considerably less than previous reports based on their reported survival rate of 90 percent. They believed that the best initial treatment is expectant management; particularly, as cessation of blood flow to the acardiac fetus may occur spontaneously, or the size of the acardiac twin may remain small and not pose any complication. However, 40 percent of cases (4/10) reported in their series were associated with a very small acardiac twin with a postpartum APTW ratio less than 3 percent.

Although the authors did not provide estimates of prenatal APTW ratio that affect prognosis, Jelin et al[24] confirmed the importance of acardiac twin size in 76 pregnancies complicated by TRAP sequence. Their study included 18 cases with the APTW ratio less than 50 percent and underwent conservative management and radiofrequency ablation (RFA) treatment at percentage of 61 and 39, respectively. Survival to delivery rate was 100 and 91 percent in RFA and conservative group, respectively. None of conservative pump twins had hydrops fetalis. In another series of five expectantly managed cases with APTW ratio ranging from 81-171 percent, only one pump twin survived[22]. Correspondingly, two cases reported by Minakshi et al[16] showed the conservative treatment was the best suited for salvation of the pump twin when the APTW ratio less than 25 percent. There were no signs of impending CHF in this study. Therefore, Wong and Sepulveda[25] suggest that expectant management was the only appropriate treatment in cases that had no poor prognostic factors (large acardiac twin). Treatment was indicated when serial ultrasonography and colour Doppler velocimetry revealed the presence of adverse factors such as signs of CHF and abnormal Doppler velocimetry observations. Prenatal treatment was important for acardiac twin pregnancy[25].

Lewi et al[26] studied the outcome of acardiac twins diagnosed in the first trimester. They found that 33 percent of the pump twins spontaneously died between diagnosis and planned intervention. Spontaneous flow arrest and persistent flow toward the acardiac twin were found at percentage of 21 and 46, respectively, during 16-18 weeks of pregnancy. As mentioned above, some authors believed that early prophylactic treatment of all cardiac twins should be started at 16 weeks in reverse blood flow cases[27].

Surgical interventions

In 1983, Platt et al[17] reported the first cord occlusion procedure to clamp the umbilical cord. The procedure was performed via either hysterotomy or endoscopy. In the same way, Van Allen et al[4] reported 14 cases and clarified the underlying pathophysiology. The first successful of extreme invasive intervention for acardiac twinning was reported in 1989 by Robie et al[28]. A 710-gram acardiac twin was selectively delivered by hysterotomy (sectio parva) at 22 weeks with subsequent delivery of a 2,130-gram healthy pump twin at 33 weeks later. Many aggressive interventions were subsequently reported. These procedures also produced significant maternal complications including pulmonary edema due to aggressive tocolytic agent usage and abruptio placenta secondary to procedure itself. Consequently, many minimally invasive techniques had been originated. These procedures were percutaneous cord occlusion by coil embolization of the umbilical cord through a hysterotomy incision with ultrasound-guided, endoscopic neodymium yttrium aluminium garnet (Nd: YAG) laser coagulation of the umbilical vessels and the umbilical cord ligation via fetoscope and infrafetal ablation. Currently, the surgical interventions were divided to cord occlusion and infrafetal technique as presented in Table 1. Both minimally invasive procedures provided the satisfied maternal and fetal outcomes.The surgical intervention of acardiac twin was applied in widespread range due to it uncommonness. However, there was no single optimal technique due to its rarity and heterogeneity presentation. Obviously, the advantages and disparity of cord occlusion and infrafetal ablation had been reported by many authors[28-34].

Cord occlusion

Table 1. Minimal invasive treatment modalities for vascular occlusion of acardiac twin[25]

Cord occlusion	
Ultrasound guided	Coil
	Alcohol-suture material
	Ligation
	Monopolar diathermy
	Bipolar diathermy
Fetoscopic guided	Ligation
	Laser
Infrafetal ablation	
Ultrasound guided	Absolute alcohol
	Monopolar diathermy
	Laser
	Radiofrequency
Fetoscopic guided	Currently not available

Mainly, cord occlusion can be achieved using either ultrasound or fetoscope for guidance. Van Allen et al[4] described this technique firstly. Ultrasonographic guide can use with much technique for cordocclusion for example coil, alcohol-suture material, ligation, monopolar diathermy and bipolar diathermy. However, one potential disadvantage of this technique includes the use of instruments with larger diameters than needle-based procedures may cause preterm rupture of membranes. In addition, this technique has limited use inedematouscords and requires disruption of the dividing membrane if the access is through the amniotic sac of the pump twin[25]. Moreover, the identification and access to the umbilical cord is usually difficult, particularly in cases with anterior placenta, occasionally requiring additional procedures such as amnioinfusion and amniotomy to gain access to the acardiac twin's umbilical cord. This results in prolongation of operative time and a more invasive procedure, together with the potential risk of pseudomonoamnionicity due to disruption of the intertwin membrane and entanglement of the cord. Another procedure, fetoscopic-guided ligation of the umbilical cord was first attempted by McCurdy et al[29]. Their procedures were performed on pregnancies after 24 weeks of gestation who failed to interrupt blood flow. This suggests that advancing gestational age may be associated with a hydropic cord or large umbilical vessels that are more difficult to coagulate. From the experience reported in the literature, advancing gestational age seems to make the umbilical vessels grow to a large diameter. The effectiveness of laser coagulation is reduced in a large umbilical cord. In comparison, Arias et al[30] presented 22 cases of acardiac twin treated by invasive and expectant management. Pump twin mortality with surgery and expectant management were 13.6 and 50 percent, respectively. In addition, fetoscopic laser coagulation (placental vascular anastomoses or the umbilical cord) of the acardiac twin, with the possibility of applying bipolar forceps as an additional minimally invasive surgical technique, offered an effective treatment option in the management of the TRAP sequence. Survival rate of this treatment was 80 percent. Sixty-seven percent of pregnancies with surviving pump twins go beyond 36 weeks of gestation without further complications[31]. Tan and Sepulveda[32] reported the fetoscopic guided umbilical cord ligation results of 16 cases. Perinatal mortality and severe preterm (less than 32 weeks) delivery rates were 38 and 70 percent, respectively. Nowadays, the topic of cord occlusion is inconclusive. New occlusive procedures to the umbilical cord, which are less invasive than fetoscopic procedures, have been developed. Thus, ultrasonographic guided needling techniques can be performed in facilities and low-cost materials. It is less traumatic to the uterus and membranes. Fetoscopic approach requires expensive equipment and skill operator.

Infrafetal ablation

Now, Infrafetal ablation has been accomplished with several techniques, in addition to ultrasound guide with absolute alcohol, monopolar diathermy, laser and radiofrequency. Radiofrequency ablation (RFA) is the most recent technique[33]. This method uses radiofrequency energy that is progressively increased until impedance of the device indicates satisfactory coagulation or when colour Doppler ultrasound confirms obliteration of blood flow to the acardiac fetus. Because the energy from the radiofrequency device is applied only to the tip after deployment into tissue, it avoids injury to the co-twin and surrounding tissues. Consequently, occlusion techniques disadvantages the cord of acardiac twin usually short, edematous and sometimes located near the pump twin. It can cause damage to the pump twin's cord, difficulty to complete occlusion and bleeding from accidental cord rupture.

In contrast, an intrafetal approach targets the abdominal aorta or pelvic vessels of the acardiac twin. The vessels are easily identified regardless of cord condition, placental location, amniotic fluid volume and position of the acardiac twin. Correspondingly, the use of colour Doppler ultrasound has greatly facilitated by allowing the clear visualization of the acardiac twin's

feeding vessel and its main intra-abdominal branches[25]. Moreover, Tan and Sepulveda[32] review 71 cases of acardiac twin treated by minimally invasive techniques. Overall survival rate of pump twin was 76 percent. Infrafetal ablation was associated with later median gestational age at delivery and higher median treatment-delivery interval than cord occlusion techniques. It was also associated with a lower technical failure rate, lower rate of premature delivery or rupture of membranes before 32 weeks and higher rate of clinical success than cord occlusion techniques.

More than 90 percent of survival rate was achieved in monochorionic diamniotic pregnancies complicated by TRAP sequence who treated by RFA[34]. Above all, infrafetal ablation is the treatment of choice. It is simpler, safer and more effective than cord occlusion techniques[16].

Conclusion

An acardiac twin is a rare condition. Precise and early diagnosis was important. Treatment techniques and optimal timing could provide an excellent outcome. Perinatal mortality of the untreated pump twin was rather high (50-70%). Differential diagnosis from intrauterine fetal death was recommended. Prognostic factor based on APTW ratio. Hydrop fetalis pump twin anomalies, CHF and polyhydramnios were common complications. If the APTW ratio is less than 50 percent without complication then serial ultrasound observation is the treatment of choice. Optimized timing to intervention was considered to treat any deterioration condition. If APTW ratio was between 50-70 percent with some problem then treatment should be initiated. Infrafetal ablation is the treatment of choice for acardiac twins. It is simpler, safer and more effective than the cord occlusion techniques.

What is already known on this topic?

Acardiac twin or twin reversed arterial perfusion (TRAP) sequence is a rare unique complication of monozygotic multiple pregnancy. In this disorder, there is a normally formed donor (the pump twin) who has features of congestive heart failure (CHF) as well as a recipient (the acardiac twin) who lacks a well-defined heart structure. Overall, the perinatal mortality rate for the pump twin is high. The optimal management of acardiac twin pregnancies is controversial. The expected treatment of acardiac anomaly presently relies on maximizing the chance of term delivery and preventing CHF in the healthy pump twin or interrupting vascularization between the two twins.

What this study adds?

Precise and early diagnosis of acardiac twin was important. Treatment techniques and optimal timing could provide an excellent outcome. Prognostic factor based on APTW ratio. Hydrop fetalis pump twin anomalies, CHF and polyhydramnios were common complications. Treatment should be started if the APTW ratio is more than 50 percent. Infrafetal ablation by RFA was easy and safe to performed. It gave an excellent outcome.

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Potential conflicts of interest

None.

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ทารกแฝดที่มีการเชื่อมต่อและไหลย้อนกลับของหลอดเลือดสายสะดือ: รายงานผู้ป่วยและทบทวนการดูแลรักษา

ต้องตา นันทโกมล, อธิตา จันทเสนานนท์, จรินทร์ทิพย์ สมประสิทธิ์, สกล มนูสุข, เด่นศักดิ์ พงศ์โรจน์เผ่า, คมสันติ์ สุวรรณฤกษ์

ทารกแฝดที่มีการเชื่อมต่อและไหลย้อนกลับของหลอดเลือดสายสะดือหรือแฝดที่ไม่มีหัวใจ เป็นภาวะที่พบได้น้อยมากของทารกแฝดชนิดไข่ใบเดียวกัน ภาวะนี้เกิดจากการพัฒนาที่ผิดปกติของการไหลเวียนเลือดของแฝดคนรับ โดยมีการเชื่อมต่อระหว่างหลอดเลือดแดงกับหลอดเลือดแดงของแฝดคนให้แฝดคนที่มีลักษณะโครงสร้างที่ปกติเรียกว่าแฝดคนให้ ซึ่งอาจพบภาวะหัวใจล้มเหลวและแฝดคนรับที่ไม่มีระบบไหลเวียนเลือดที่ติดต่อกับรกโดยตรง ทำให้การพัฒนาของร่างกายผิดรูปไปที่มักมีความผิดปกติในส่วนศีรษะและขาดการพัฒนาของหัวใจ สาเหตุเนื่องจากมีการเชื่อมต่อกันของหลอดเลือดแดงกับหลอดเลือดแดง และหลอดเลือดดำกับหลอดเลือดดำที่ผิดปกติโดยเลือดจะไหลเวียนจากแฝดคนให้ผ่านไปยังแฝดคนรับ แล้วกลับเข้าหลอดเลือดแดงสายสะดือของแฝดคนให้อีกครั้ง อัตราการตายของแฝดคนให้พบร้อยละ 35-55 ในกรณีที่ไม่ได้รับการรักษา การวินิจฉัยและการพยากรณ์โรคสามารถทำได้โดยการตรวจคลื่นเสียงความถี่สูง การรักษาที่เหมาะสมที่สุดยังมีข้อขัดแย้งกันอยู่แต่หลักการคือการอุดกั้นหลอดเลือดที่เชื่อมต่อกันระหว่างแฝดทั้งสอง รายงานนี้นำเสนอตัวอย่างผู้ป่วยและทบทวนสมมติฐาน การเกิดโรค การวินิจฉัยก่อนคลอด การพยากรณ์โรค และการรักษา