REVIEW ARTICLE

Atypical Rare Intertwin Transfusions: Twin Anemia Polycythemia Sequence and Acute Peripartum Intertwin Transfusion

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Abstract

The twin anemia-polycythemia sequence (TAPS) and acute (peripartum) intertwin transfusion (AITT) are rare forms of transfusions between twins in monochorionic (MC) multiple pregnancy. These types are met less often than twin-to-twin transfusion syndrome (TTTS), the prevalence is 2–3% for TAPS after laser ablation and 3–6% spontaneously and 1.5–2.5% for AITT in comparison with 9–15% for TTTS in MC twins. The understanding of natural history and staging of the development of anemia-polycythemia helps explain some cases of unexpectedly unfavorable perinatal outcomes in newborn twins with the considerable hematological discordance without presence of remarkable prenatal signs of twin-to-twin transfusion. As opposed to the conventional wisdom, TAPS may be associated with the significant amniotic fluid discordance besides typical US signs such as discrepancy of twins' peak systolic velocity in middle cerebral artery (MCA PSV). The TAPS may not have the progredient antenatal pattern. In discordant values of PSV MCA, the extended monitoring of the fetal cardiovascular profile is required to estimate the degree of perinatal risk as well as select the optimal time for delivery. Postnatally, the differential diagnosis of AITT and TAPS is based on the logics of retrospective of fetal MCA PSV values. The AITT diagnosis may be assumed for the MC twins with normal antenatal MCA Doppler velocities and episodes of bradycardia of fetuses during the delivery. At the same time, TAPS that is being developed just before the delivery can hardly be certainly ruled out. This article focuses on the review of current views as for perinatal diagnosis of rare forms of intertwin transfusion as well as on particularities of some clinical cases with unexpected association of symptoms. It is shown that some unusual symptoms of TTTS and TAPS as well as their mixed forms influence the US images and complicate the classification, staging, and prognosis of the abnormalities.

Keywords: Acute intertwin transfusion, Monochorionic twins, Twin anemia-polycythemia sequence.

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Introduction

The population frequency of both natural multifetal pregnancy and that one induced by assisted reproductive technologies has been growing for the last decades.[1,2] Monochorionic (MC) twin pregnancies are characterized by a high complication rate.[3] The ultrasound (US) monitoring of multifetal pregnancy is mandatory and has the exclusive priority in the obstetric clinic.

The US features of some specific complications of MC multiple pregnancy are known and well distinguishable. The US patterns and typical alteration sequences of twin-to-twin transfusion syndrome (TTTS) in most cases possess the typical staging of signs such as twin's oligohydramnios/polyhydramnios with "classical" values of depth of vertical amniotic fluid (AF) pockets discordance equal to <20 mm/>60–80–100 mm (different according to gestational age); donor's decreased or not visualized bladder; and recipient's specific cardiovascular impairment.[4] They all are well studied at the present day. The circulatory imbalance in TTTS causes the discordance of hematological markers (hemoglobin, hematocrit) of the twins.[5]

At the same time, the newborn MC twins may have the hematological discordance that is not related to the prenatal development of the classical progressing TTTS.[5]

The twin anemia-polycythemia sequence (TAPS) and acute (peripartum) intertwin transfusion (AITT) are less common forms of intertwin transfusions in MC multiple pregnancy. These forms are met less often than TTTS.[6,7] The prevalence is 2–3% for TAPS after laser ablation and 3–6% spontaneously and 1.5–2.5% for AITT in comparison with 9–15% for TTTS in MC twins.[6,7] The understanding of staging of anemia-polycythemia development helps explain the cases of unexpectedly unfavorable perinatal outcomes in MC newborn twins with the considerable hematological discordance if there are no prenatal US signs of twin-to-twin transfusion.

The TAPS is a rare atypical form of chronic intertwin transfusion occurring in approximately 5% of MC twins.[6,7] The TAPS has no vivid prenatal US markers in some cases. The TAPS stages are described in studies with relatively small number of observations.[5] It is an abnormality of a high perinatal risk. The death of one or both twins may be the outcome of advanced forms of TAPS. The diagnosis, perinatal prognosis, and approach in TAPS cause more difficulties than those in TTTS.

Common clinical and pathogenetic features of TAPS are following:

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- Transfusion predominantly through small anastomoses.[5,8]
- May be developed both initially (spontaneous TAPS) or after the incomplete laser coagulation of anastomoses in TTTS.[8,9]
- As the laser therapy of TTTS is widely used, there is an increased reporting about TAPS development.[9]
- The hemoglobin and hematocrit levels are significantly discordant in twins.
- The discordance degree of hematological markers of newborns depends on the antenatal stage of TAPS (Table 1).[6]

The US features of TAPS are following:

- Usually there are no twins' oligohydramnios/polyhydramnios with "classical" AF discordance; the urinary bladder of the donor fetus is filled within normal.[8]
- The US image of the liver of the recipient fetus reminds the *starry sky* due to the echogenic walls of portal venules in the setting of hypoechoic hepatic parenchyma[6] (Fig. 1).
- Prenatal diagnosis of TAPS is based on the discordance of middle cerebral artery peak systolic velocity (MC8A PSV) of the twins: more than 1.5 MoM in the donor, less than 0.8 MoM in the recipient.[8,9]
- The thickness and US structure of twin's placental portions may be different[8] (Fig. 2).
- Twins may have isolated MCA PSV discordance at the first and second stages of TAPS; any other US signs of fetal compromise may be not observed at these stages.
- Thereafter, the abnormalities of Doppler cardiovascular profile of fetuses in the setting of hemodynamic recipient overload and donor's anemia are defined (Table 1).[6]

The actual guidelines dedicated to MC multiple pregnancy monitoring strictly recommend to evaluate twins' MCA PSV biweekly, beginning from 20th gestational week.[6]

In TAPS, the approach is individual and depends on the fetal gestational age, stage, and family decision (Figs 1 and 2). Approach options are conservative monitoring, early delivery, and intrauterine transfusion for the anemic fetus.

The AITT is an acute and temporary circulatory imbalance of twins developed in the setting of preexisting well-being. The diagnostic criteria for acute peripartum TTTS are not clear and most reports use a hemoglobin difference of more than 8 g/dL at birth in the absence of TTTS or TAPS.[5]

There are two types of AITT:

- Between live fetuses: developing before or in the course of delivery.
- During intrauterine fetal demise of one of MC fetuses (acute perimortem TTTS): it is accompanied by the embolization syndrome of the survived twin or acute perimortem TTTS.

Perimortem AITT features the specific natural history after the death of one of MC twins and is considered in the publications dedicated to the problems of the survived MC twins.

Features of peripartum AITT are following:

- It is met less often than the syndromes of chronic intertwin transfusions (with the frequency of about 1.5–2.5%).[5,10]
- It is developed before or in the course of delivery in the setting of antenatal well-being.[11]
- This transfusion is possible through large superficial anastomoses.
- The mechanism of this type of transfusion is not absolutely clear till now.
- The pressure gradient in the placental flow of twins with the blood overflowing to the lower pressure region appears during the uterine contractions[5] as well as episodes of recipient's bradycardia (more often during the delivery).[11]
- One more type of peripartum transfusion distinguished in some publications is the fetoplacental transfusion after the birth of the first fetus. In this case, the hematological alterations may occur only in one of the twins.[12]
- There are no any reliable antenatal US predictors of AITT;[13] therefore, this type of transfusion is not described in Society of Ultrasound in Obstetrics and Gynecology (ISUOG) guidelines dedicated to twins US monitoring.
- Some authors believe that it is impossible to certainly distinguish AITT and TAPS developed just before the delivery.[5]

The following peculiarities are representative for AITT cases (Fig. 3):

- The concordant or slightly discordant fetal growth
- Normal filling of urinary bladders of both fetuses
- Normal arterial and venous Doppler, no considerable discordance of fetal MCA
- Episodes of bradycardia in the recipient fetus during the delivery
- The perinatal complications such as Apgar scale; the period of stay in the ICU tends to be less severe in AITT than in cases of TAPS.[14,15]

Table 1: TAPS stages[6]

Stage	*US/Doppler*
I	Donor's MCA PSV >1.5 MoM
	Recipient's MCA PSV <1.0 MoM
	Without other signs of fetal compromise
II	Donor's MCA PSV >1.7 MoM
	Recipient's MCA PSV <0.8 MoM
	Without other signs of fetal compromise
III	Signs of stages I–II accompanied by with impaired placental Doppler and fetal cardiovascular profile (umbilical artery, ductus venosus, umbilical vein)
IV	Hydrops
V	Death of one or both fetuses

Discussion

Unfortunately, the antenatal detection of abnormal perinatal course of MC multiple pregnancy and prediction of intrapartum complications are difficult and even impossible in some cases.

According to the most general view, AF discrepancy with twins' oligohydramnios/polyhydramnios is a mandatory attribute of TTTS whereas the absence of this sign is pathognomonic for TAPS.[5] But in some cases TAPS may be associated with significant AF discrepancy besides typical US symptoms such as discordance of twins' MCA PSV.

Thereby, TAPS may have some antenatal US features similar to TTTS, as well as AITT may be hardly distinguished from TAPS that was developed just before the delivery. A common feature for both abnormalities is the absence of the typical US sequence that is peculiar to TTTS in the presence of hematological discrepancy between twins after birth.[14,15] The represented series of cases demonstrates limited capacities of routine monitoring of MC multifetal pregnancy for exact detection of the type of complication. The unusual symptoms of TTTS and TAPS as well

as their mixed forms influence the US images and complicate the classification, staging, and prognosis of the abnormality.

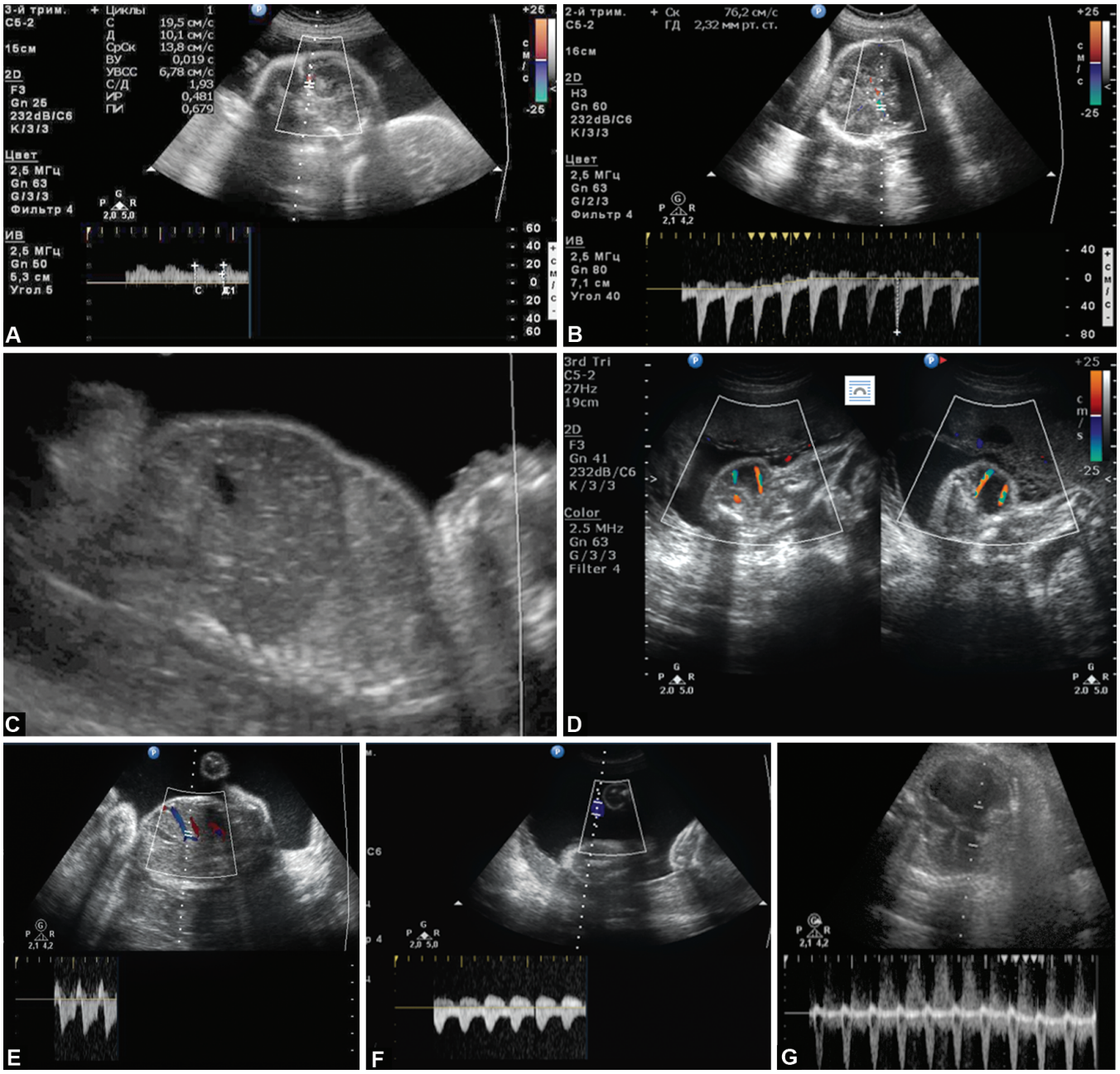
The perinatal complications of TAPS are related both to the preterm delivery and prematurity and antenatal impairment. The diagnostics, staging, and perinatal management in TAPS cause more questions than in classical progressing TTTS. The TAPS may not have the progredient antenatal pattern. If defining the considerable discordance of twins' MCA PSV, the extended monitoring of the fetal cardiovascular profile is required. If the cardiovascular score is reduced in one or both fetuses in TAPS, the early delivery is to be considered after family counseling about the two types of risks, both intrauterine fetal death and iatrogenic prematurity, depending of gestational age.

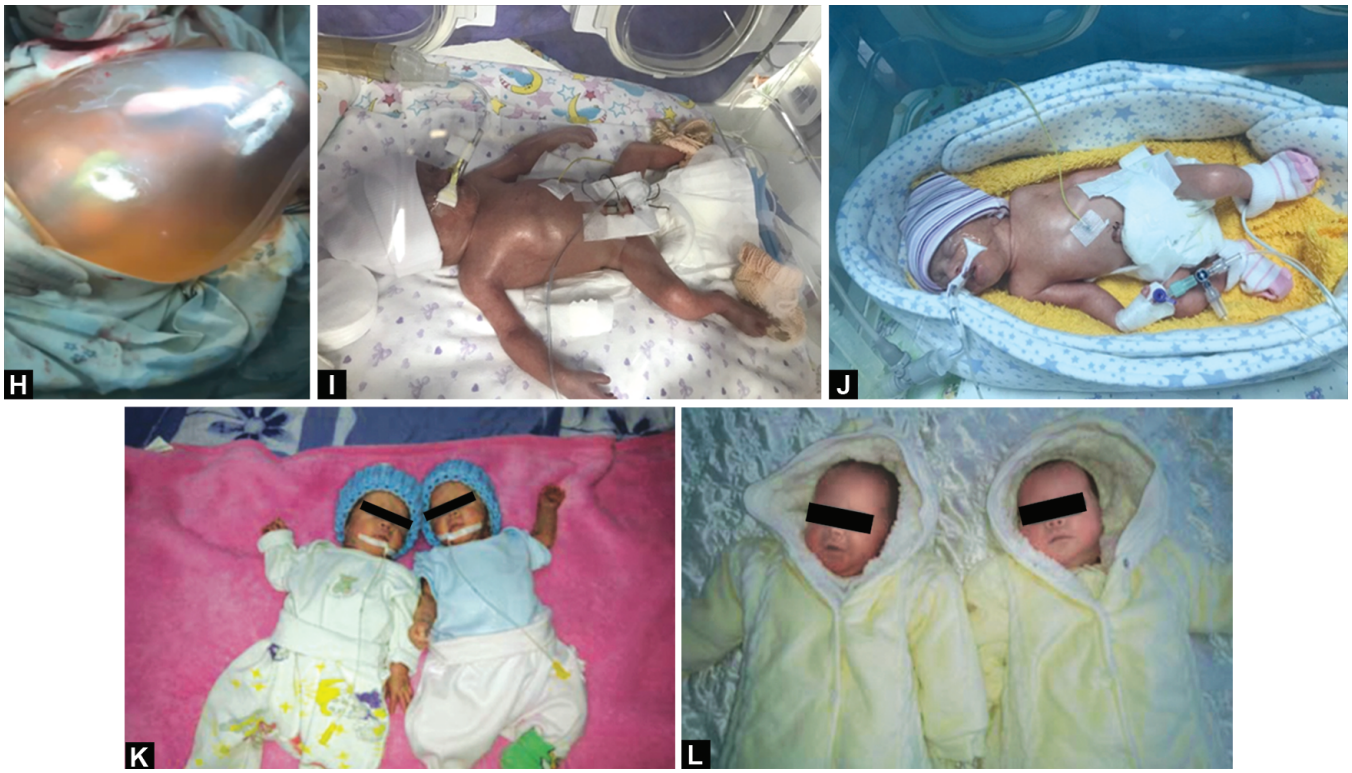
The AITT has no specific antenatal US predictors. The differential diagnosis of AITT and TAPS is based on the logics of retrospective of fetal MCA PSV values: AITT diagnosis may be assumed for the MC twins with the normal antenatal MCA Doppler velocities. It tends to be impossible to certainly exclude TAPS developed just before the delivery.

Conclusion

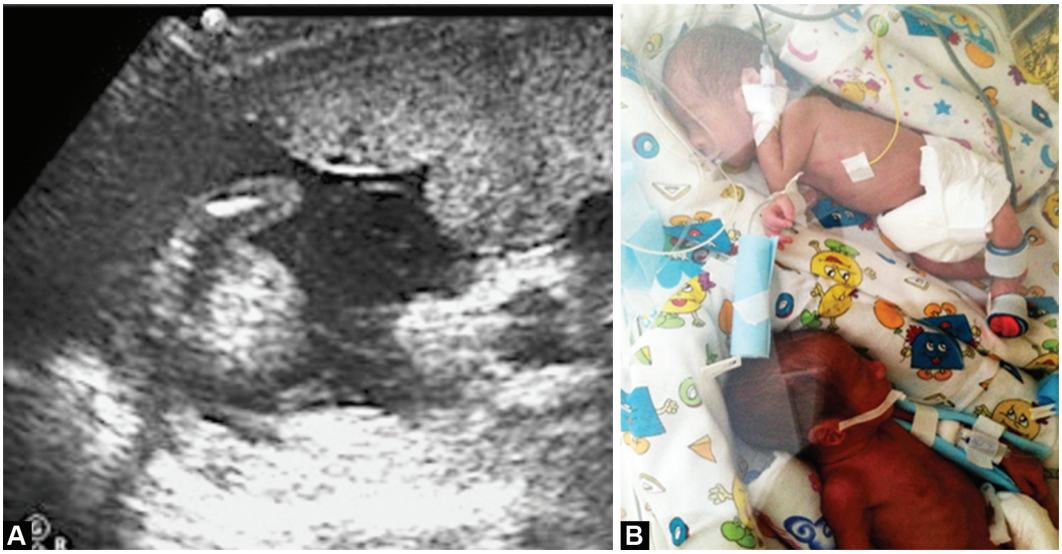
As opposed to the conventional wisdom, TAPS may be associated with the significant AF discordance besides typical US signs such as discrepancy of twins' MCA PSV.

The TAPS may not have the progredient antenatal pattern. In discordant values of PSV MCA, the extended monitoring of the





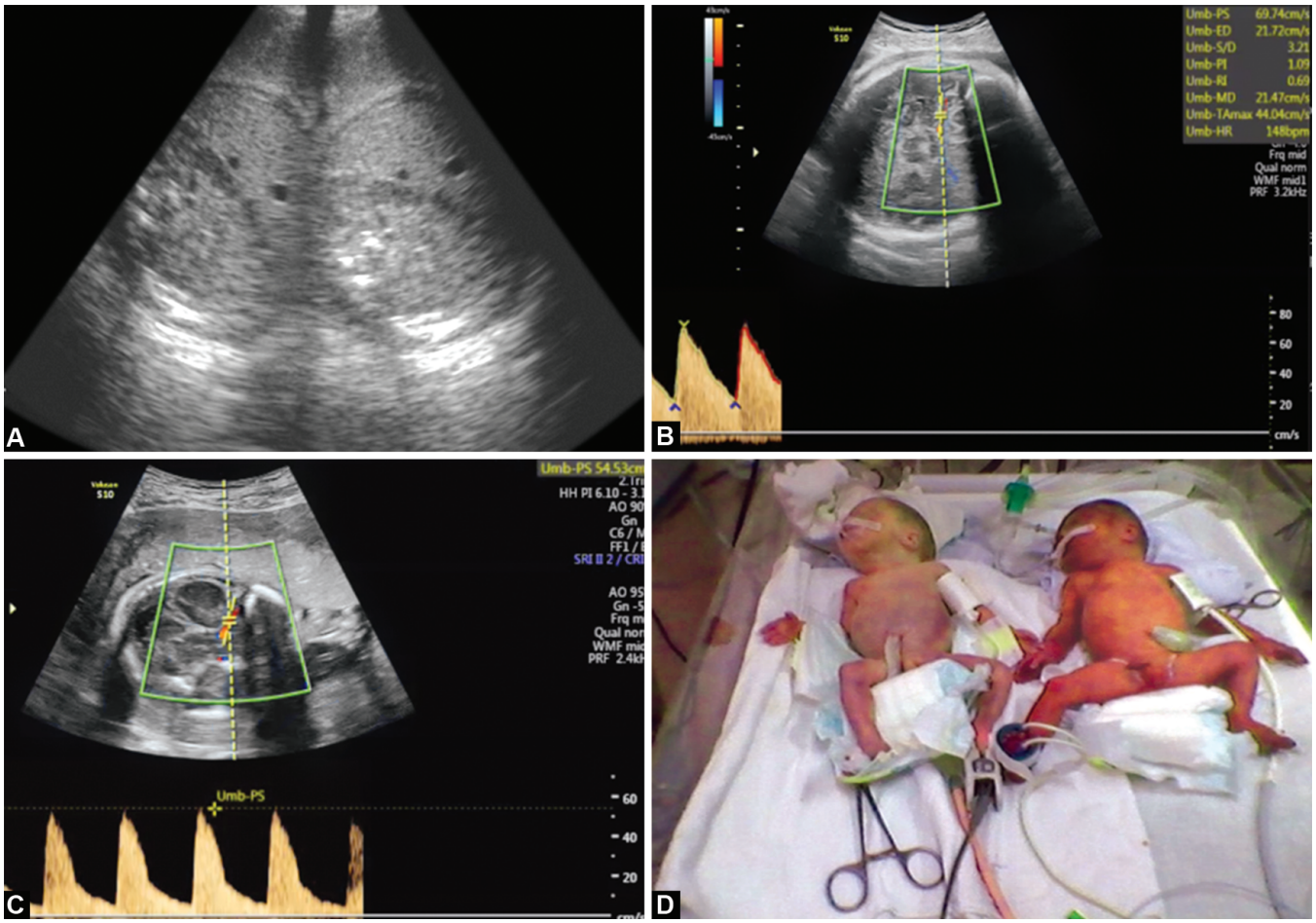
Figs 1A to L: Is it stage III TAPS accompanied with MCA PSV discordancy, twins' polyhydramnios/oligohydramnios 120/30 mm, and severe cardiovascular dysfunction of recipient fetus or not—Quintero TTTS? (A and B) Recipient's MCA PSV of 20 cm/seconds (0.55 MoM), donor's MCA PSV of 76 cm/seconds (2.11 MoM); (C) Recipient's liver with "starry sky" parenchyma; (D) Fetal bladders are filled normally; (E to G) Recipient's cardiac compromise, cardiovascular disorders (a-reverse wave in ductus venosus, umbilical vein pulsation, cardiomegaly, monophasic atrioventricular flow); (H) CS, severe polyhydramnios of recipient; (I and J) Newborns in the ICU; (K and L) Children at the age of 3 and 6 months



Figs 2A and B: TAPS, first stage not progressing till birth: (A) 35 GW. Different thickness of placental portions of fetuses. Twins had MCA PSV discordance with recipient's MCA PSV of 46 cm/seconds (0.9 MoM), donor's MCA PSV of 78 cm/seconds (1.55 MoM); (B) Photograph of newborns in ICU

fetal cardiovascular profile is necessary to estimate the degree of perinatal risk as well as select the optimal time for delivery.

Postnatally, the differential diagnosis of AITT and TAPS is based on the logics of retrospective of fetal MCA PSV values. The AITT diagnosis may be assumed for the MC twins with normal antenatal MCA Doppler velocities and episodes of fetal bradycardia during the delivery. But it is impossible to certainly rule out TAPS, which is being developed just before delivery.



Figs 3A to D: Is it a peripartum AITT or undetected early stage of twins TAPS? Clinical example: (A) 36–37 GW, MC twins, symmetrical growth of fetuses; (B and C) Concordant MCA PSV of twins before birth; (D) Delivery at 37–38 GW, episodes of fetal bradycardia during the delivery, newborns with unexpected anemia-polycythemia

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