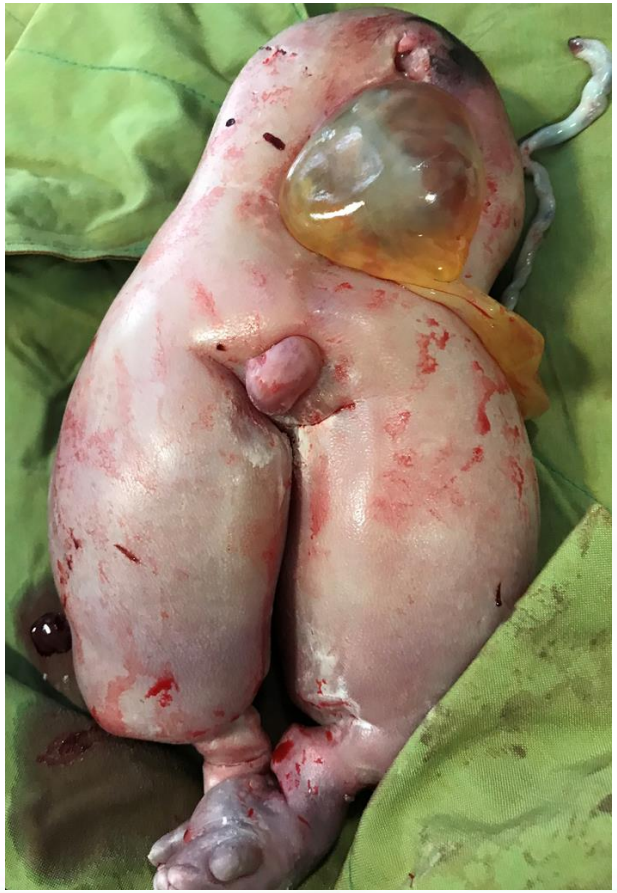
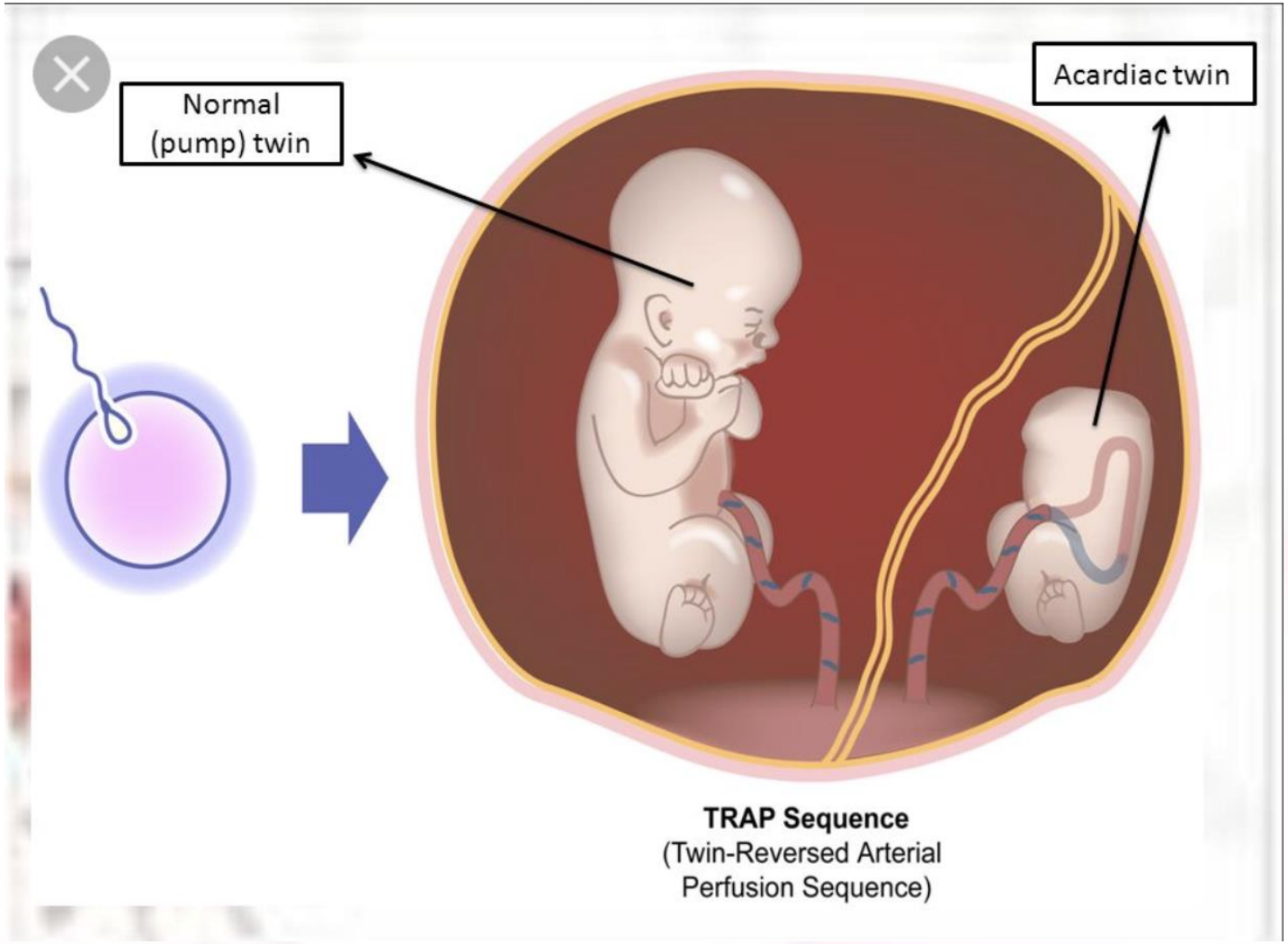
Quran Gate-Shiraz-Iran

Twin Reversed Arterial Perfusion (TRAP) Sequence



Introduction

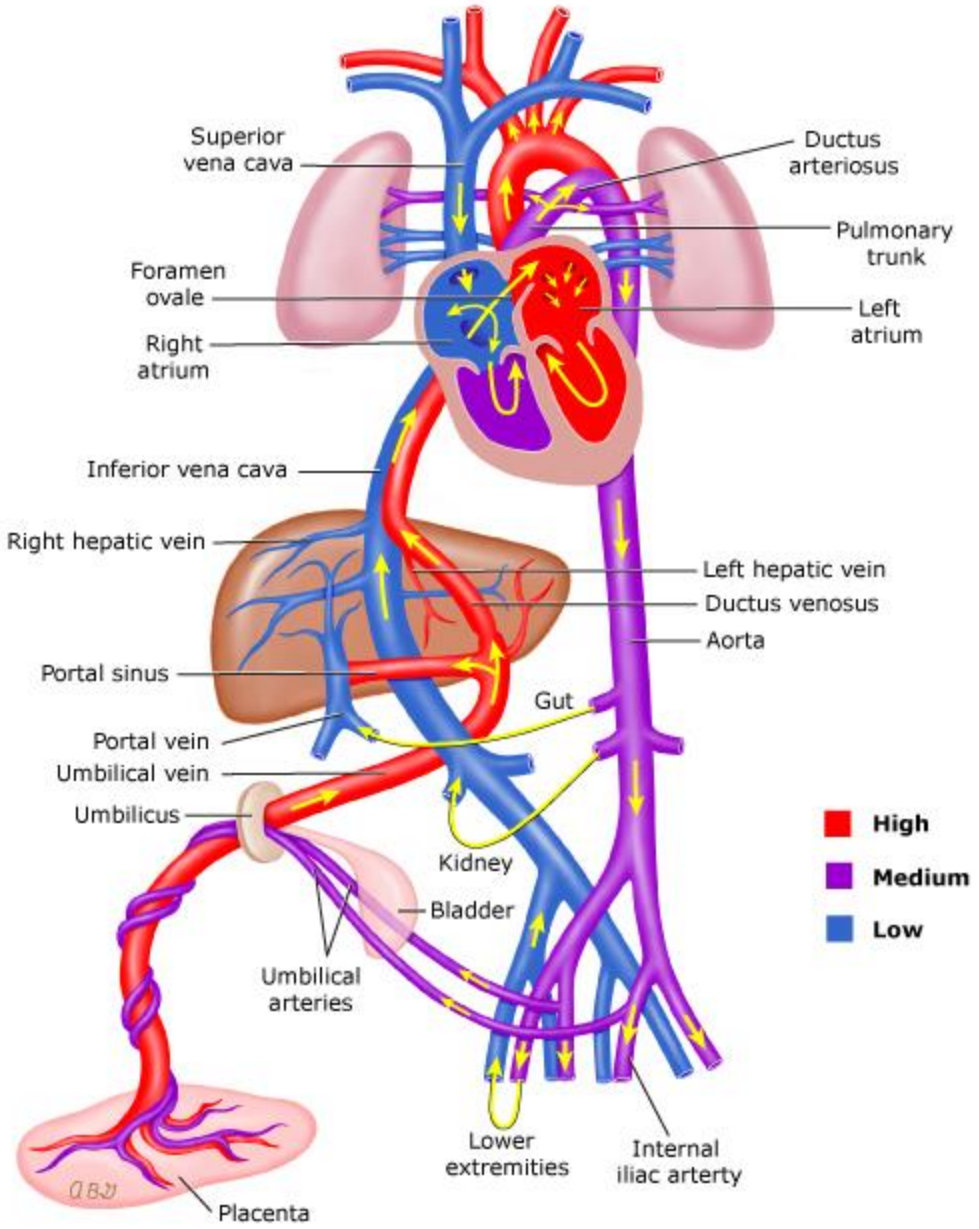
- a rare**, unique complication of MC twin** pregnancy in which a twin with an absent or a nonfunctioning heart ("acardiac twin") is perfused by its co-twin ("pump twin") via placental arterial anastomoses.
- The acardiac twin usually has a poorly developed heart, upper body, and head.
- The pump twin is at risk of heart failure and problems related to preterm birth.
- **Incidence** : 1% of MC twin pregnancies and 1 in 35,000 pregnancies
- A 2015 study estimated the incidence of acardiac twins is 2.6 percent of MC twin pregnancies and 1 in 9500 to 11,000 pregnancies



TRAP Sequence
(Twin-Reversed Arterial
Perfusion Sequence)

Pathophysiology

- In TRAP sequence, the pump twin maintains this normal pattern of fetal circulation.
- A portion of its cardiac output travels through **placental arterial-arterial** anastomoses to the umbilical artery and eventually to the systemic circulation of the recipient co-twin, thus creating "**reversed" circulation** in this twin.
- **Veno-venous** and **arterio-venous anastomoses** also occur.



Fetal circulation

- The acardiac twin has been classified according to the degree of abnormal development .
- The most common anomaly is acardiac acephalus : the fetal thoracic organs and head are absent.
- Other rarer types are acardius acormus(only the fetal head develops); acardius amorphous(consisting of a shapeless mass of tissue with no recognizable human parts); and acardius myelacephalus(the head and one or more extremities are partially developed) .

- Chromosomal abnormalities in up to 10% of cases
- **An increased risk of congenital anomalies (10%) in pump twin : anencephaly, gastrochisis, renal & skeletal abnormalities & cardiac anomalies**
- **Genetic counseling & comprehensive evaluation of pump twin are recommended.**

Prenatal diagnosis

- TRAP is diagnosed prenatally by ultrasound findings .
- The differential diagnosis of TRAP is :
  - **a single intrauterine demise of a twin**
  - **an anomalous second twin.**
- The diagnosis is confirmed by **assessing the flow pattern to the acardiac twin in a MC pregnancy and supported by observation of continued growth of the acardiac twin on serial ultrasound examinations**

Ultrasound findings

- **Acardiac twin :**
- The degree of abnormality in the acardiac twin varies widely, ranging from a fetus **with well-developed lower extremities, pelvis, and abdomen to a tissue mass that is not readily recognizable as fetal parts**.
- **The cranium may be absent or present with cranial defects, such as anencephaly or holoprosencephaly**.
- There can be **limb defects, anterior abdominal wall defects, and absence of lungs, kidney, spleen, and/or liver.**
- **The umbilical cord contains two vessels in up to 70 percent of acardiac fetuses**
- **Hallmarks of the acardiac twin include massive edema of the head, trunk, and upper extremities.**

- **Pump twin** :
- Signs of high-output cardiac failure: polyhydramnios, cardiomegaly, pericardial and pleural effusions, ascites, and tricuspid regurgitation.
- The development of cardiac failure in TRAP sequence is related to the **ratio of the size of the acardiac twin to that of the pump twin.**
- When this ratio exceeds 0.70 (ie, **the calculated weight of the acardiac twin is 70 percent or greater compared to the weight of the pump twin**), the risk of congestive heart failure in the pump twin is approximately 30 percent, compared to a risk of 10 percent when the ratio is less than 0.70 .

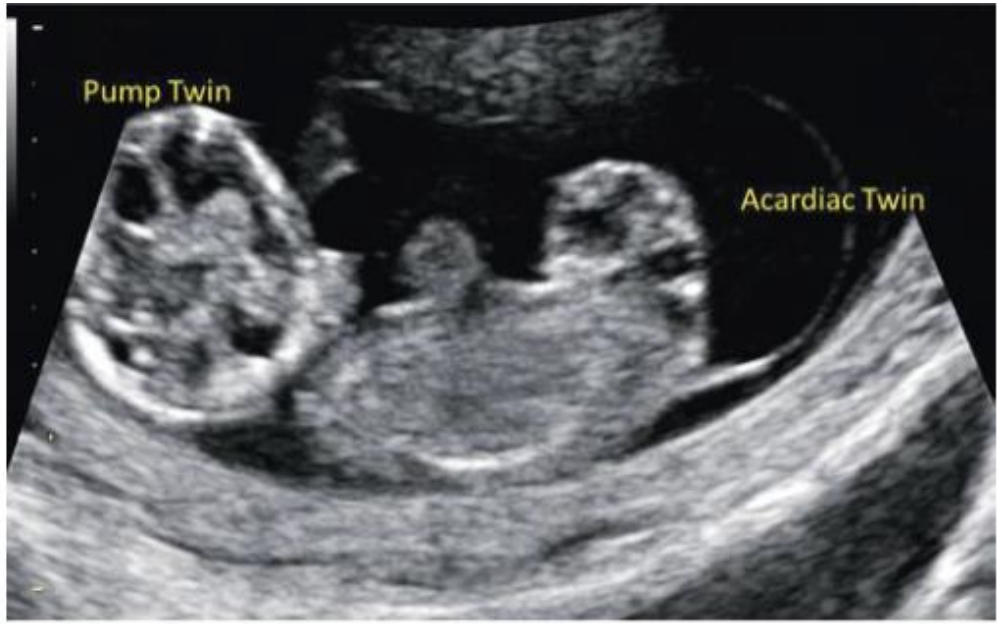


FIG 7-36 Longitudinal ultrasound image shows abnormal upper body development of the anomalous acardiac twin, which lacked demonstrable cardiac activity despite continued growth. The image includes an axial view through the brain of the morphologically normal pump twin.

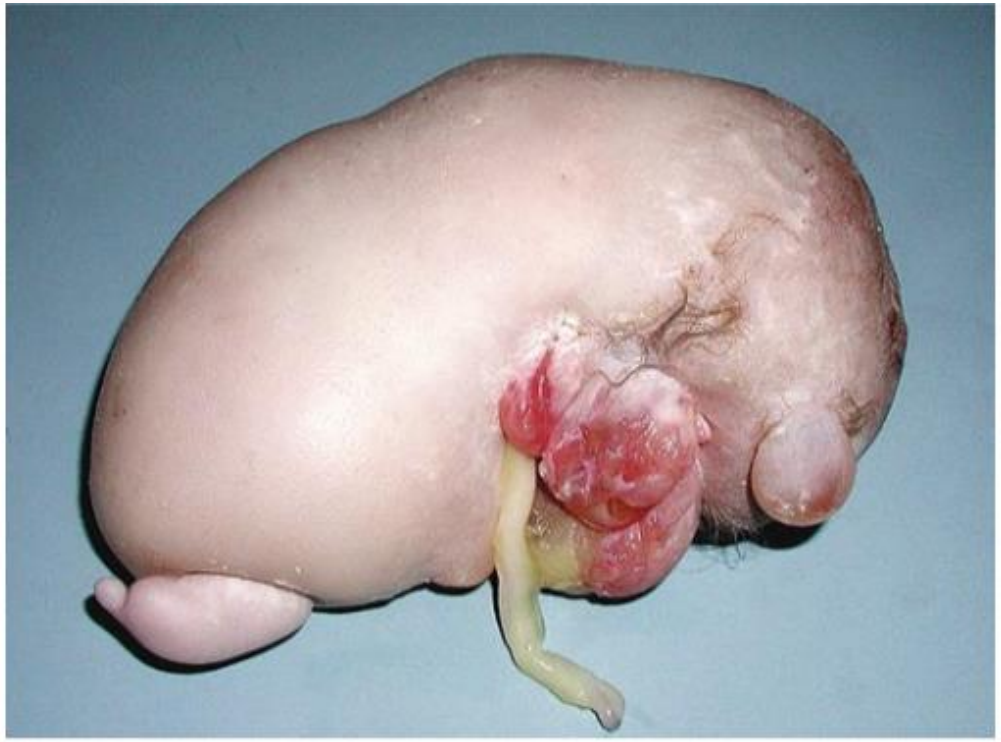
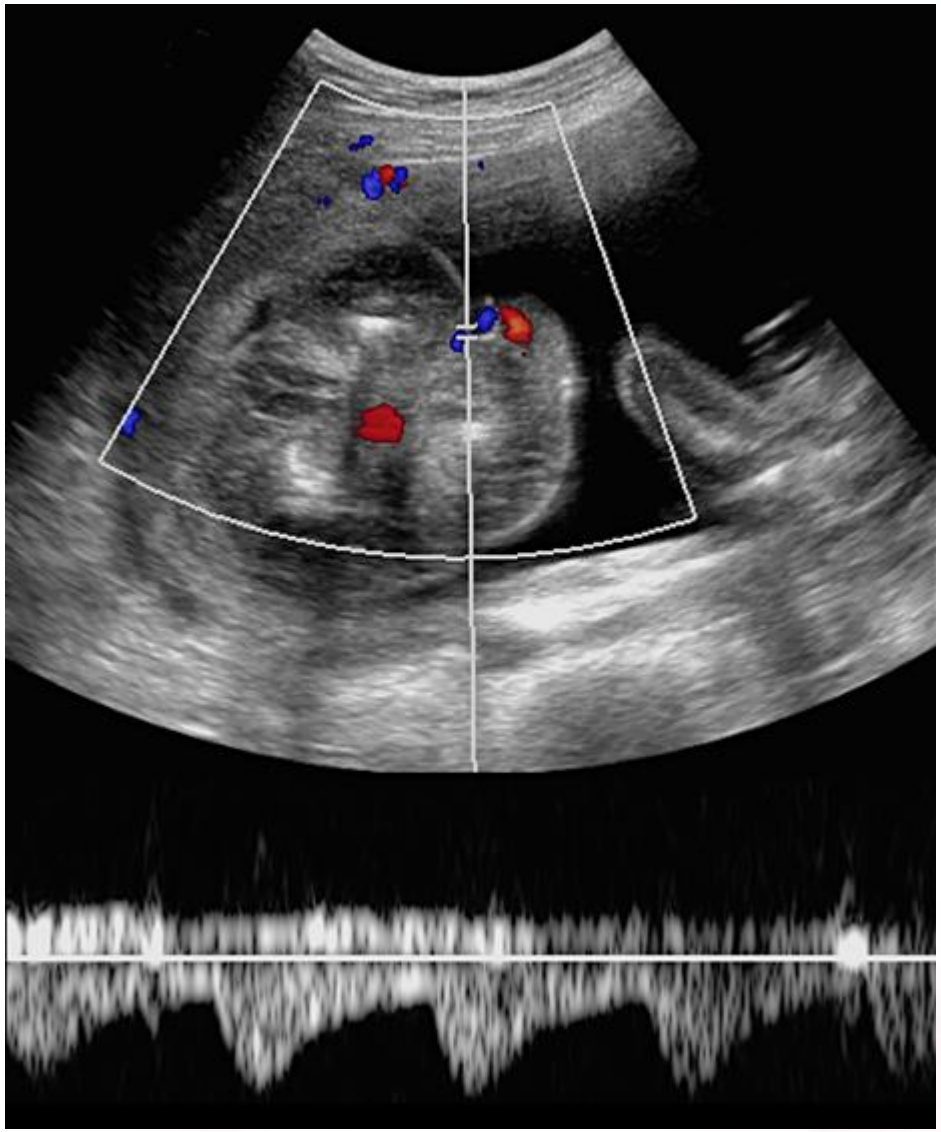


FIG 7-37 Acardiac twin demonstrating the amorphous, vestigial structures cephalad and caudad as the distance from the umbilical cord arterial circulation increases. (Pathologic image courtesy Melinda Sanders, MD, and Erika Walz, PA, University of Connecticut Health

- **Doppler ultrasound** : Doppler velocimetry is used to definitively diagnose the sequence.
- **CDS is helpful in demonstrating the presence of blood vessels inside the TRAP fetus (despite the absence of heartbeat), often located alongside a bladder-like structure in the pelvis & continuing with aorta toward the upper part of body, if present.**



Note that the arterial flow is going towards the fetal body, indicating perfusion from the normal co-twin.

Prognosis

- **Mortality rate for pump twins was 55 percent** without treatment .
- The principal reasons for this high mortality were development of **heart failure and preterm birth and its sequelae**.

Indications of poor prognosis

- ***Ratio of weight of the acardiac twin/weight of the pump twin greater than 0.70.***
- ***Polyhydramnios*** (maximum vertical pocket ≥8 cm)
- ***Cardiac failure in the pump twin***
- ***Increase in relative size of the acardiac twin : An acardiac pump twin ratio ≥1.0 is significant*** (ie, AC of the acardiac twin equal to or greater than that of the pump twin).
- ***Hydrops in the pump twin***
- ***Monoamniotic pregnancy*** (cord entanglement risk)

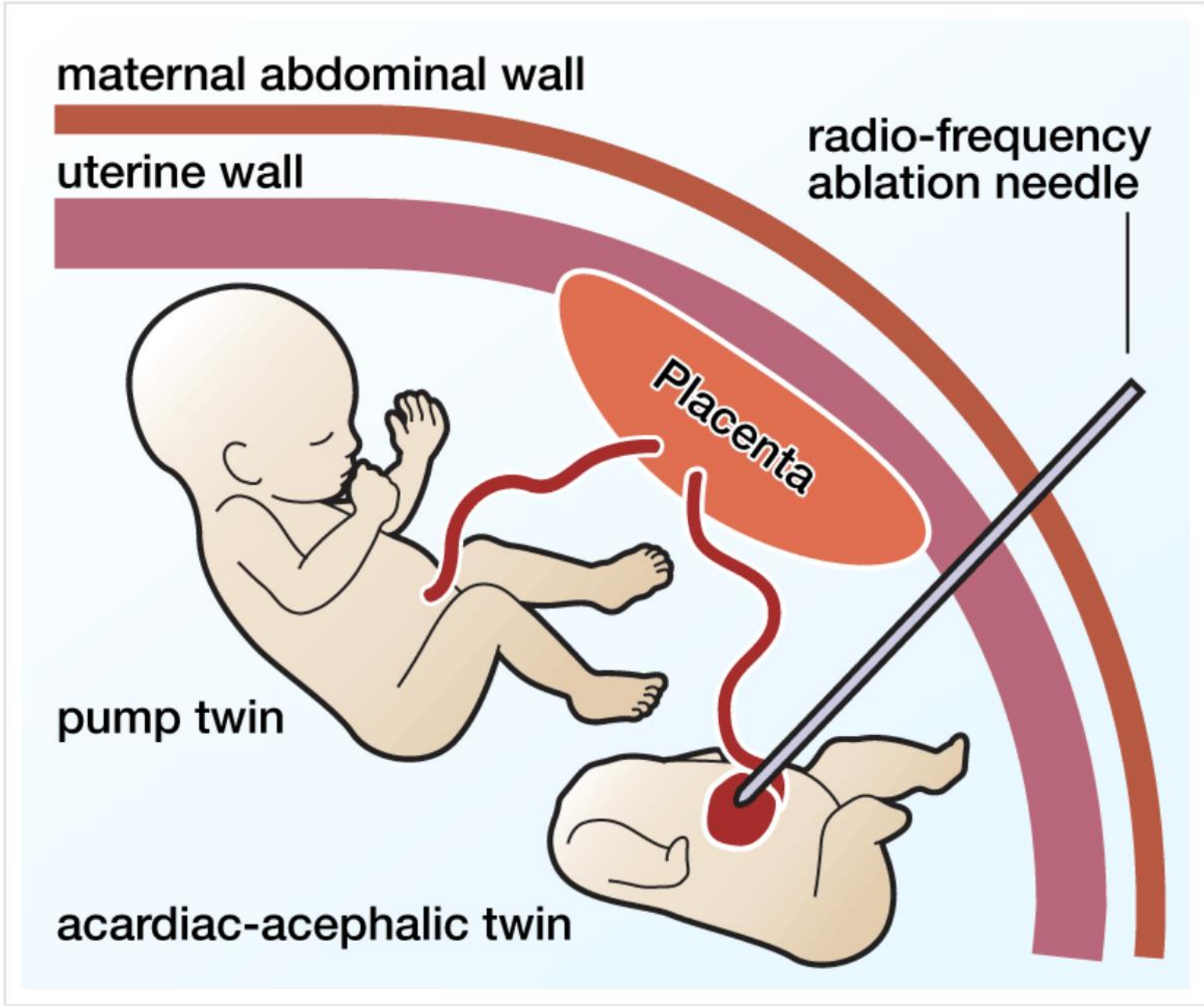
Obstetrical management

- **Weekly ultrasound surveillance**
- Ultrasound surveillance is increased to **two times a week** if there is evidence of pre-hydrops (ie, fluid in only one cavity-ascites, pleural effusion).
- **Fetal echocardiography**
- **Antenatal corticosteroid between 23 and 34 weeks of gestation**

In utero therapy

- Laser coagulation
- Bipolar cord coagulation
- Radiofrequency ablation (RFA)

Before any intervention, the karyotype of the pump twin should be determined.



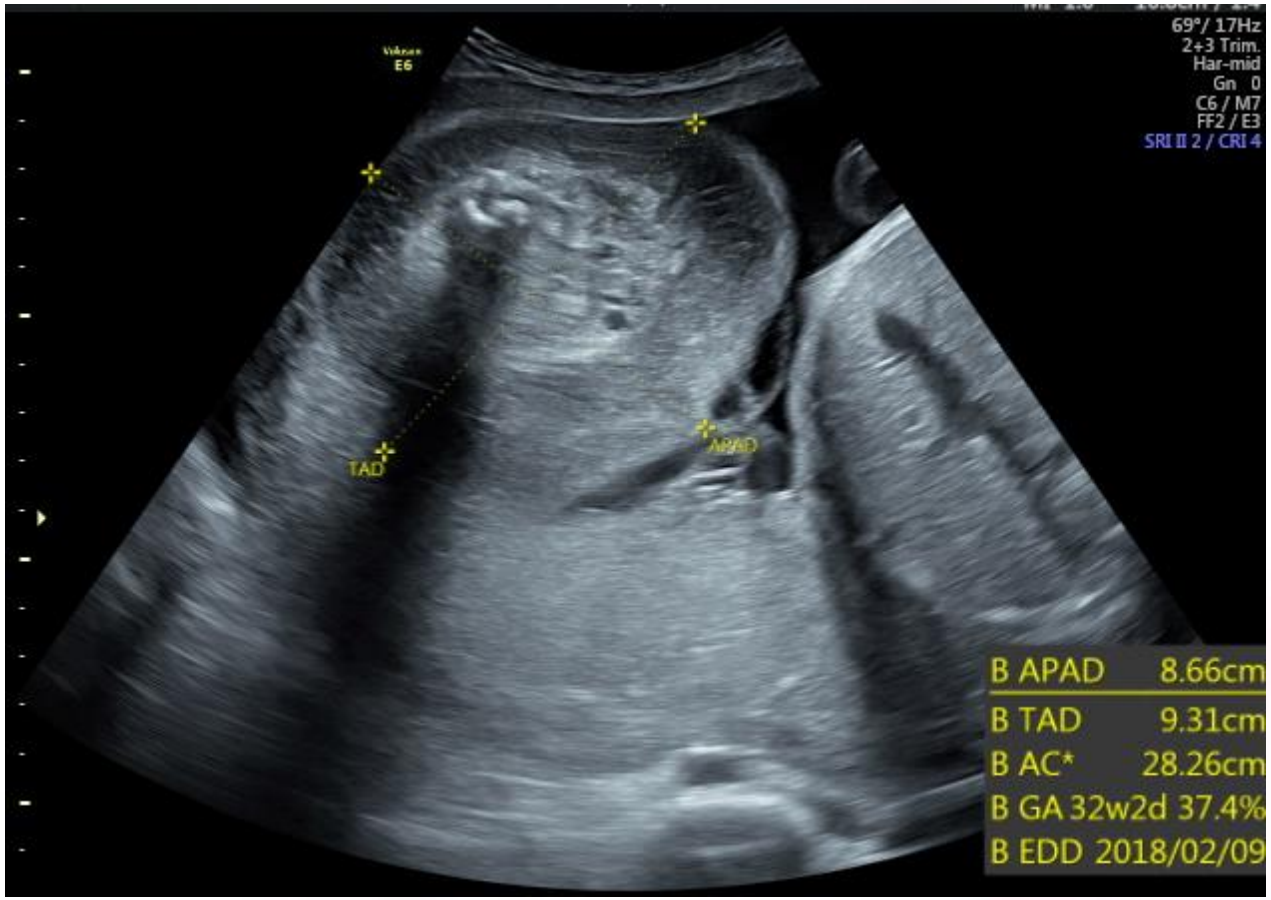
Outcome after in utero therapy

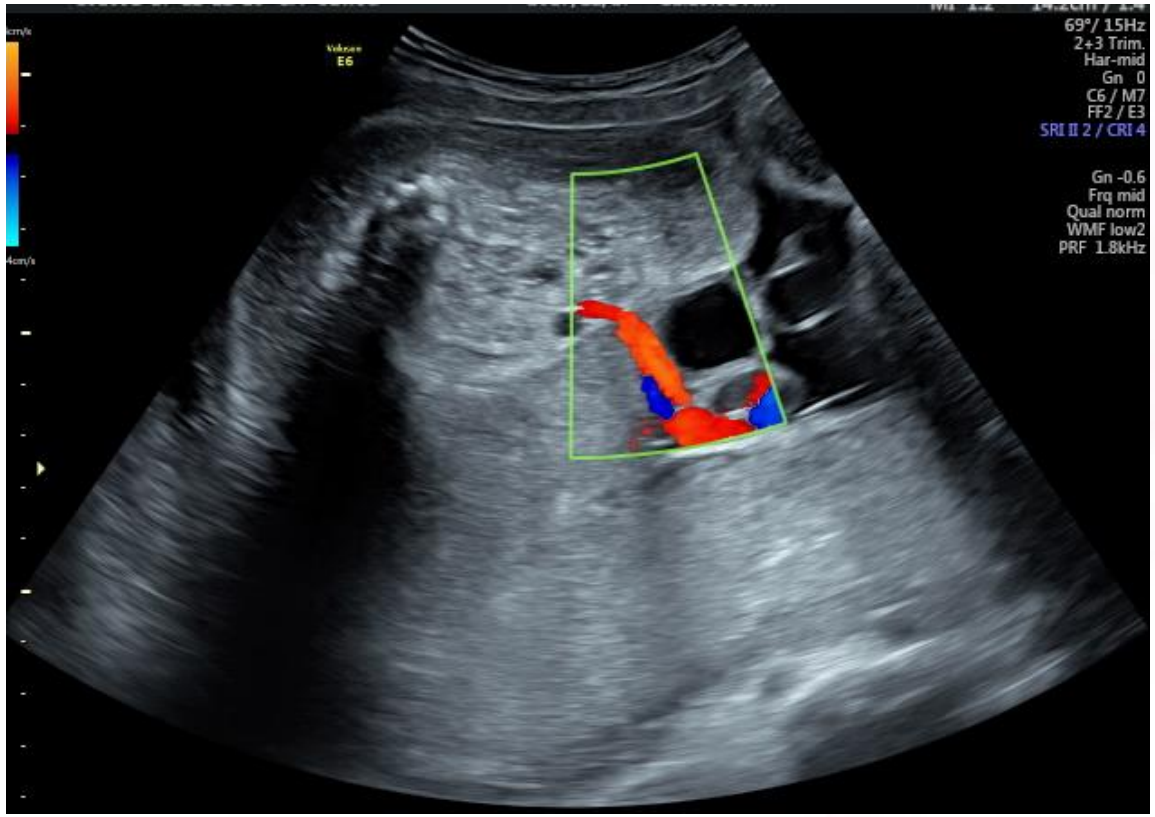
- Survival of **80 to 90 percent of pump twins**
- Maternal complications are uncommon, and include **bleeding, need for laparotomy to complete the procedure, thermal injury, chorioamnionitis leading to maternal sepsis, and disseminated intravascular coagulation**

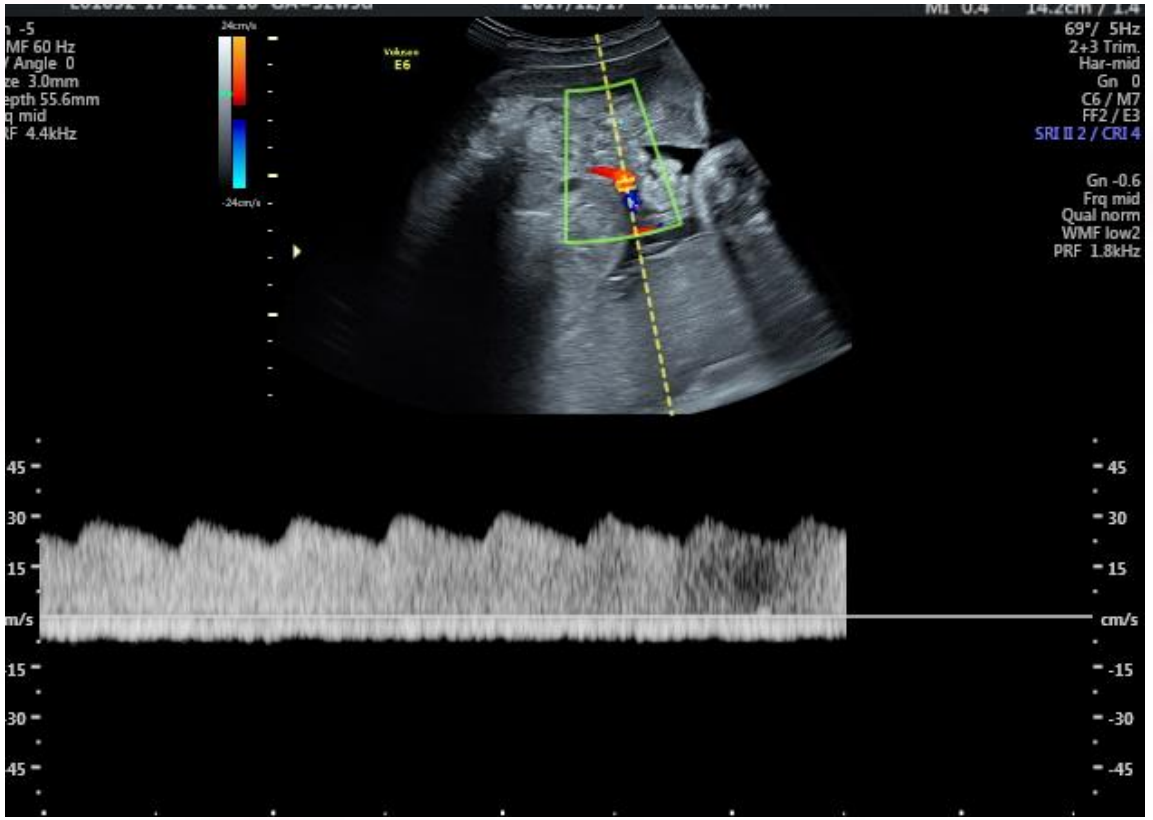
Delivery

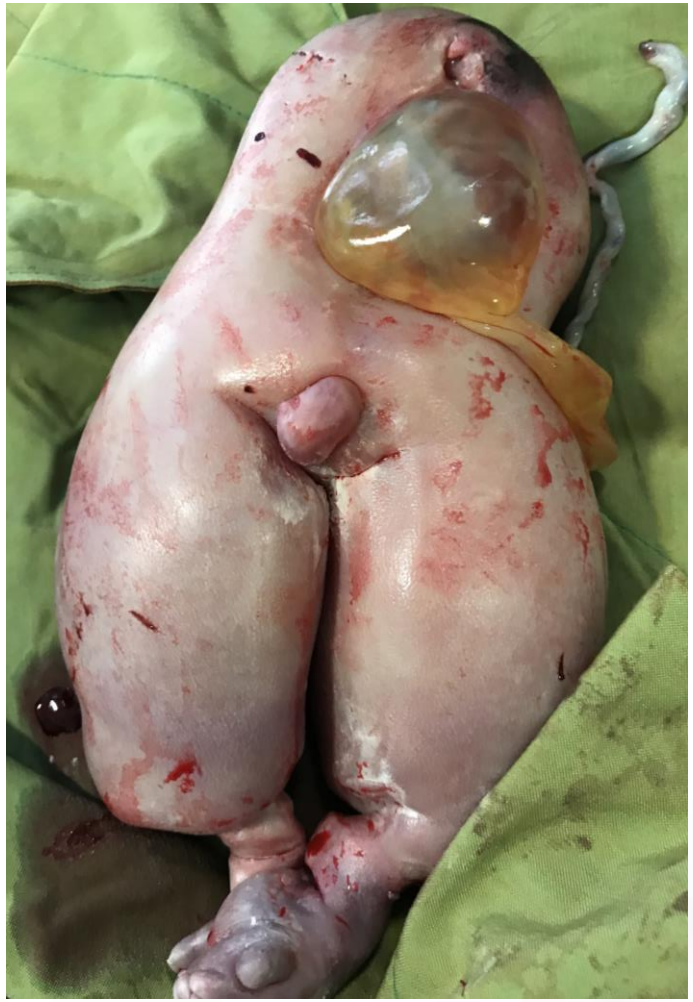
- ***We deliver TRAP pregnancies at 34 to 36 weeks of gestation***.
- **Cesarean delivery is indicated if there is a malpresentation, nonreassuring fetal heart rate pattern or low biophysical profile score for the pump twin, monoamniotic twins, or other contraindications to vaginal birth (eg, placenta previa)**.

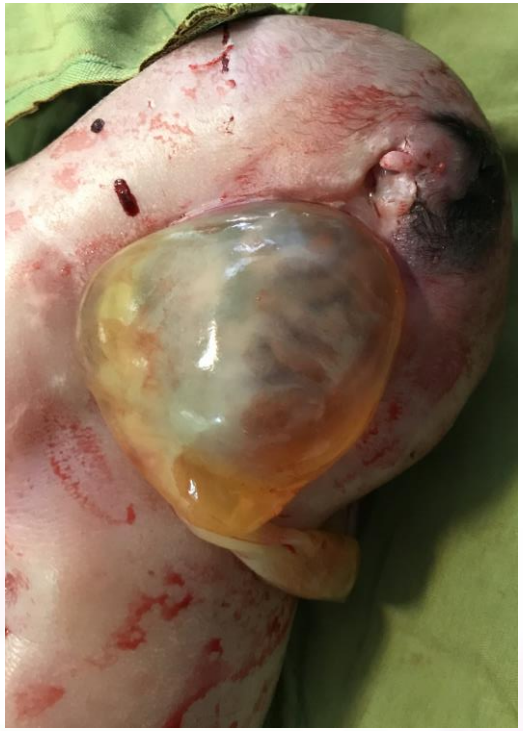
Case 1 : 32 wks


69°/ 17Hz
2+3 Trim.
Har-mid
Gn 0
C6 / M7
FF2 / E3
SRI II 2 / CRI 4
TAD
APAD
B APAD 8.66cm
B TAD 9.31cm
B AC* 28.26cm
B GA 32w2d 37.4%
B EDD 2018/02/09

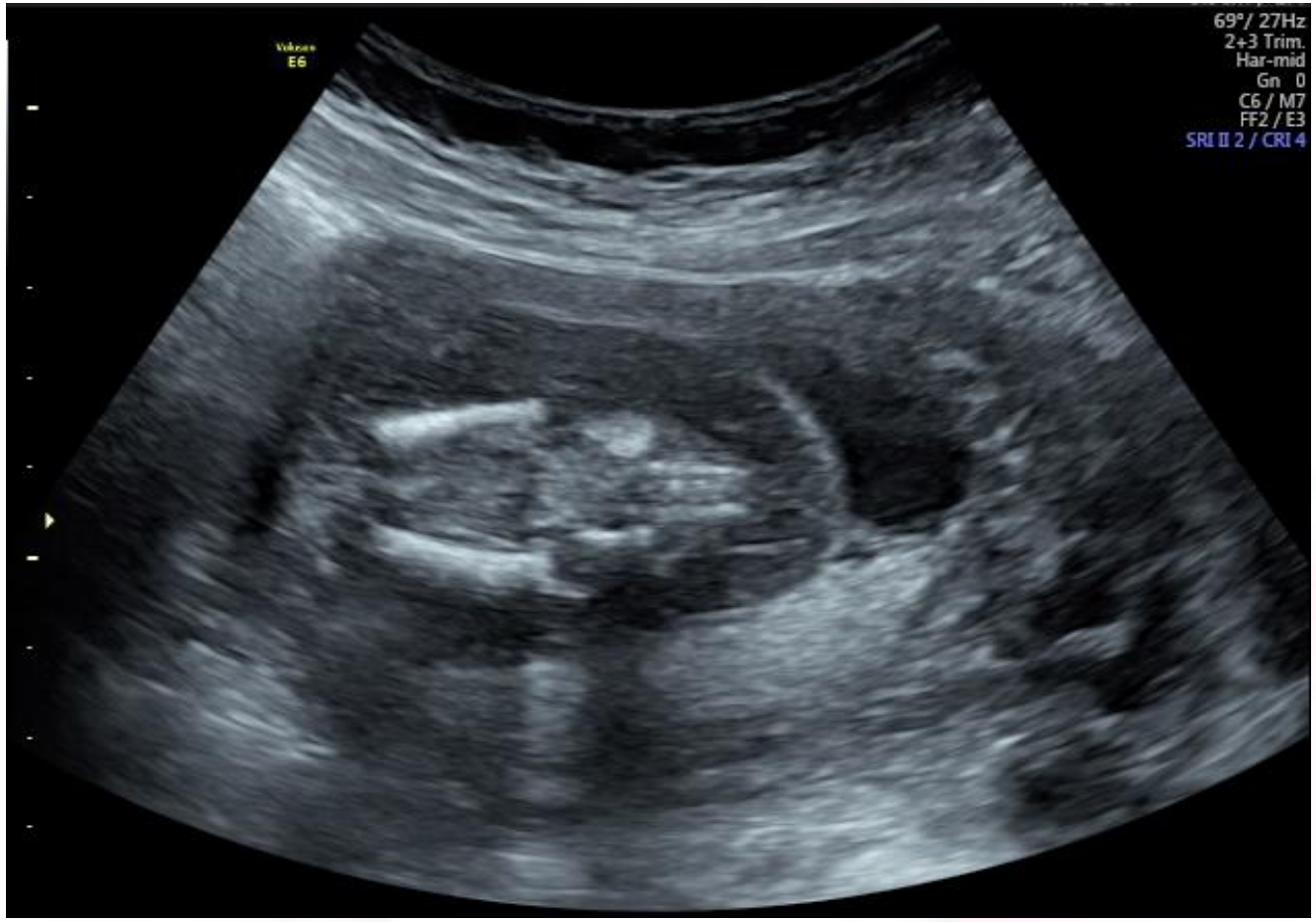


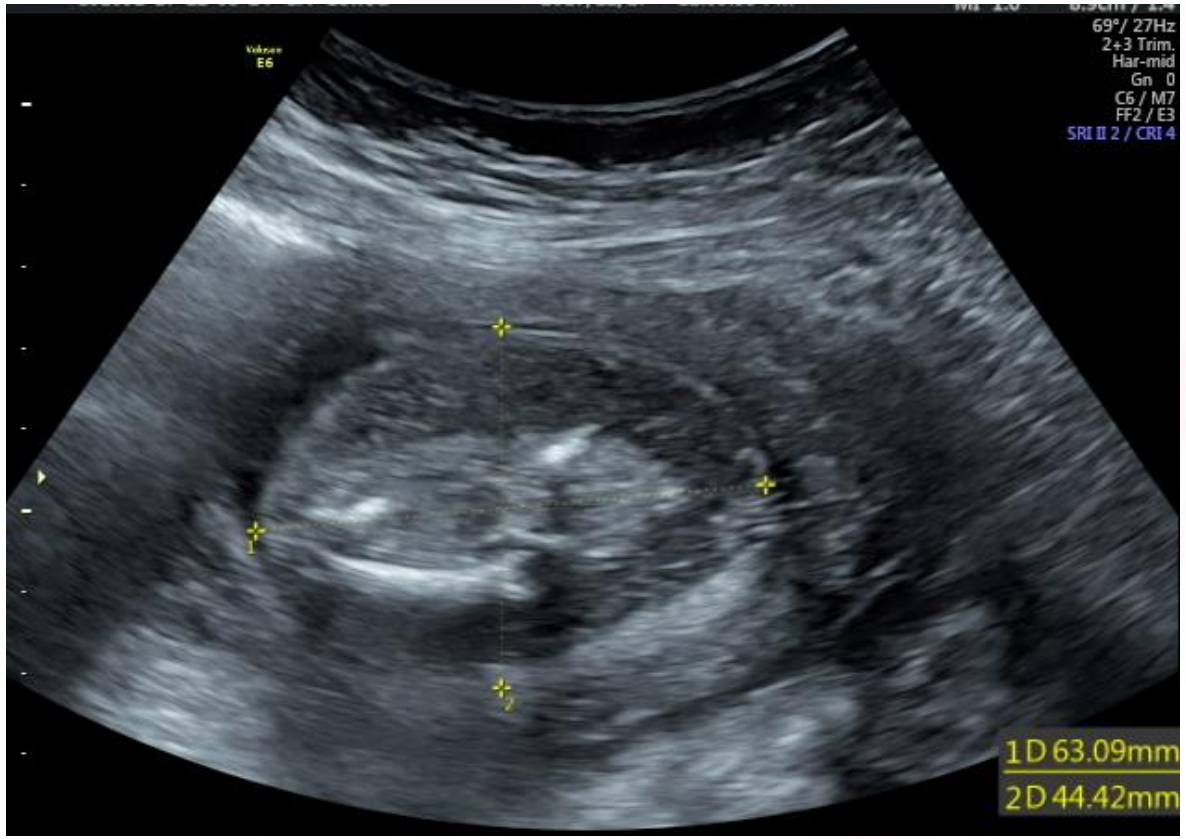


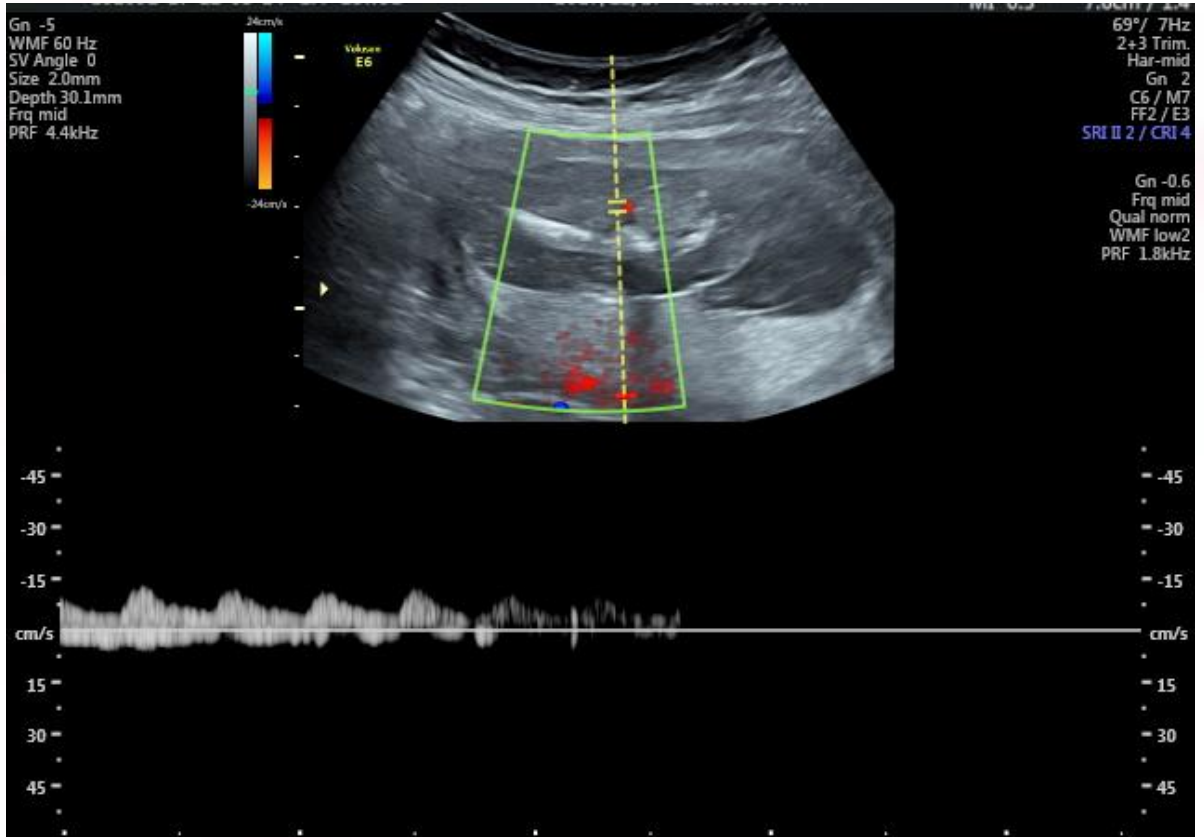




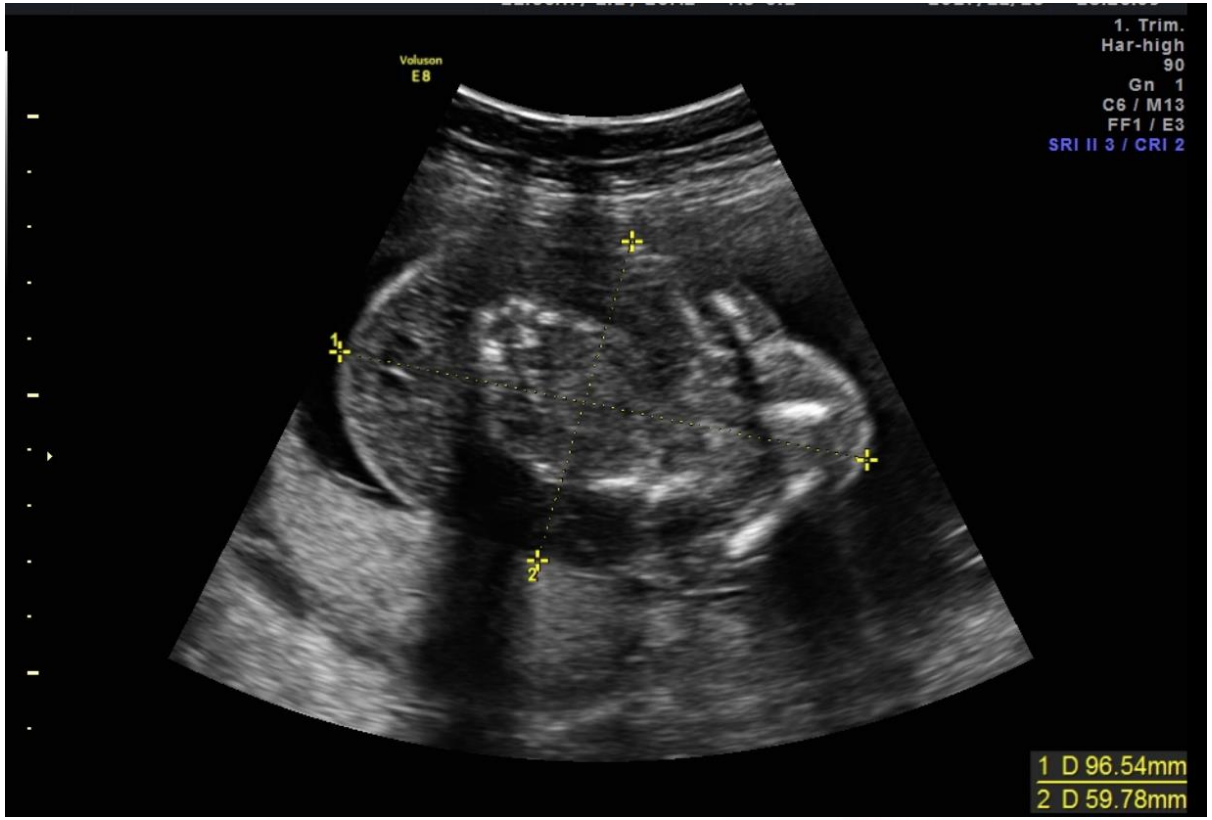
Case 2 : 19 wks

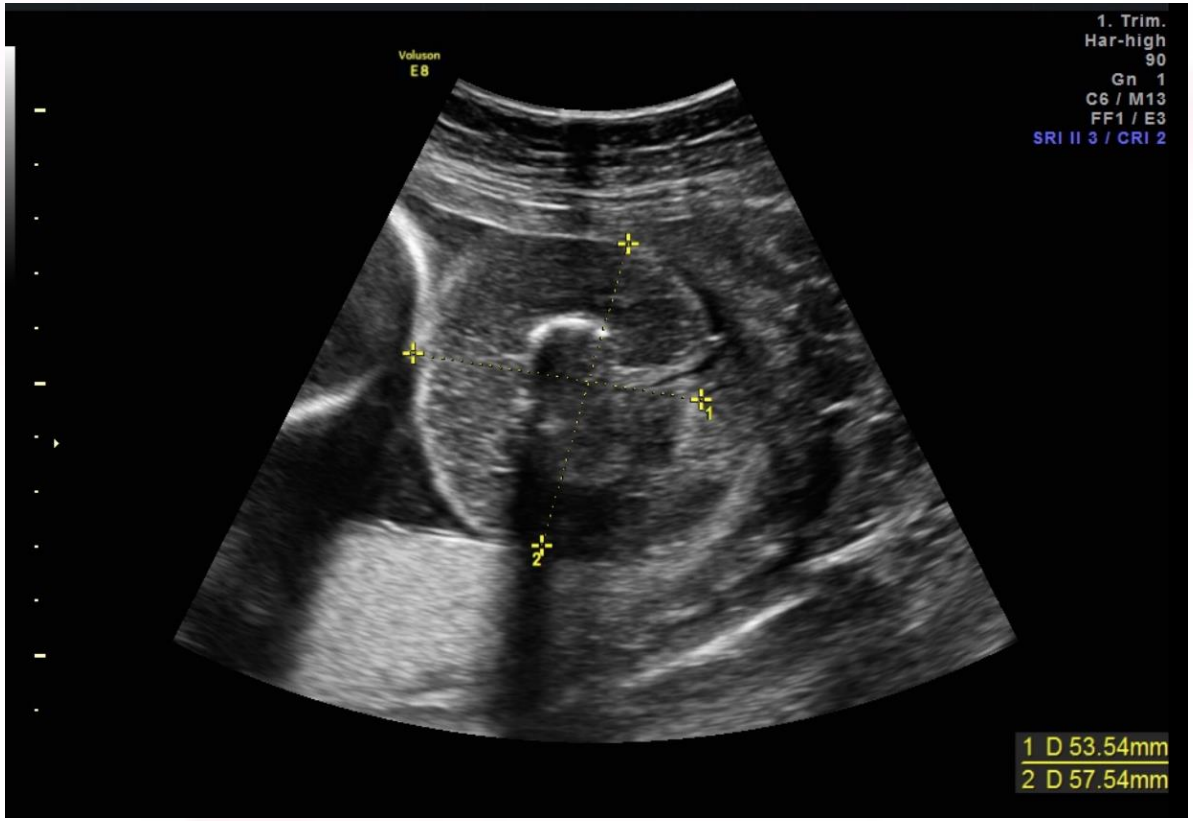


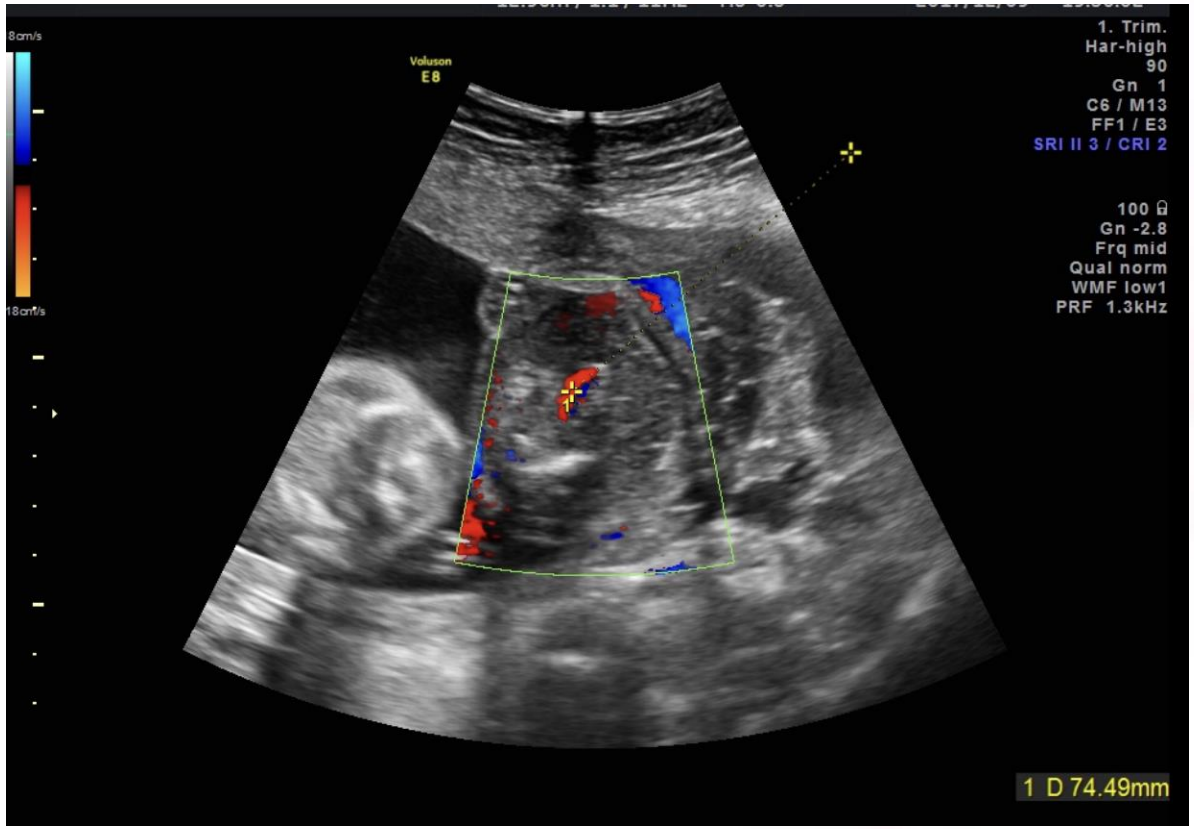

69°/ 27Hz
2+3 Trim.
Har-mid
Gn 0
C6 / M7
FF2 / E3
SRI II 2 / CRI 4
1D 63.09mm
2D 44.42mm

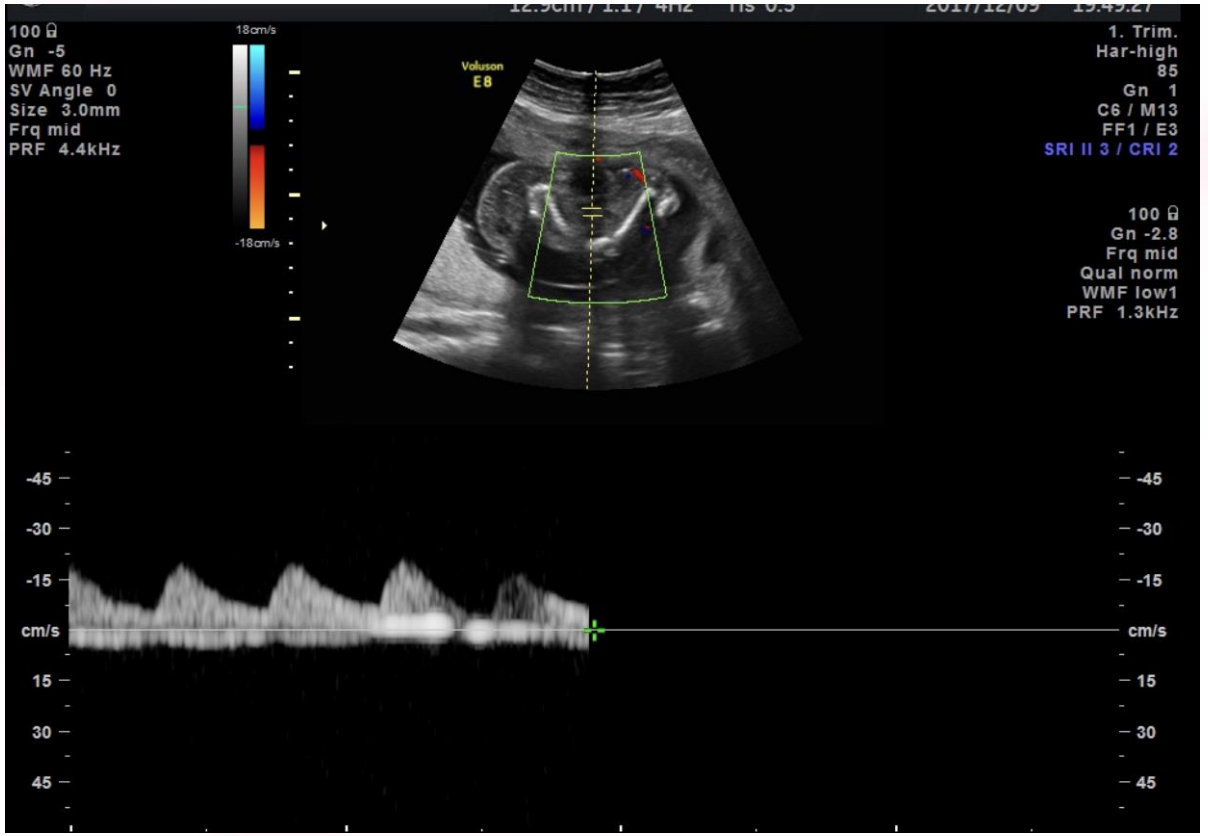

Gn -5
WMF 60 Hz
SV Angle 0
Size 2.0mm
Depth 30.1mm
Frq mid
PRF 4.4kHz
69°/ 7Hz
2+3 Trim.
Har-mid
Gn 2
C6 / M7
FF2 / E3
SRI II 2 / CRI 4
Gn -0.6
Frq mid
Qual norm
WMF low2
PRF 1.8kHz
-45
-30
-15
cm/s
15
30
45

Case 3 : 18 wks

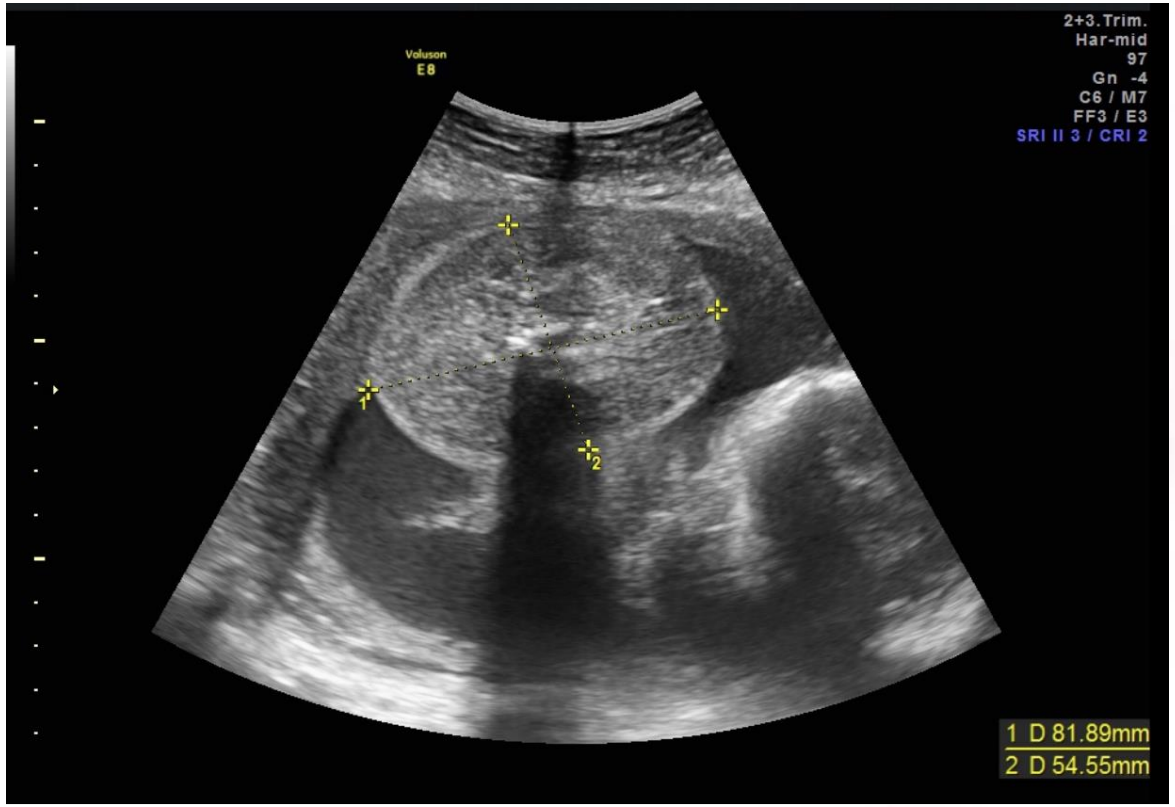


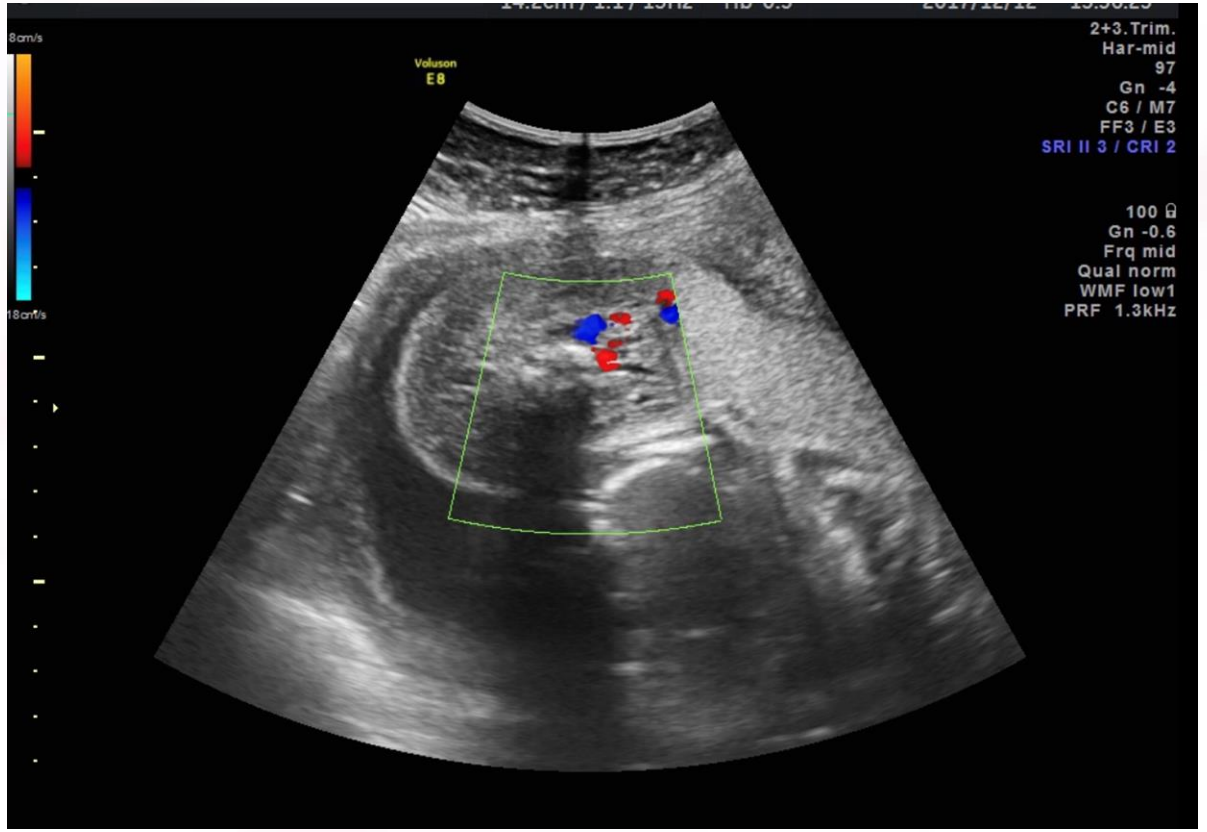


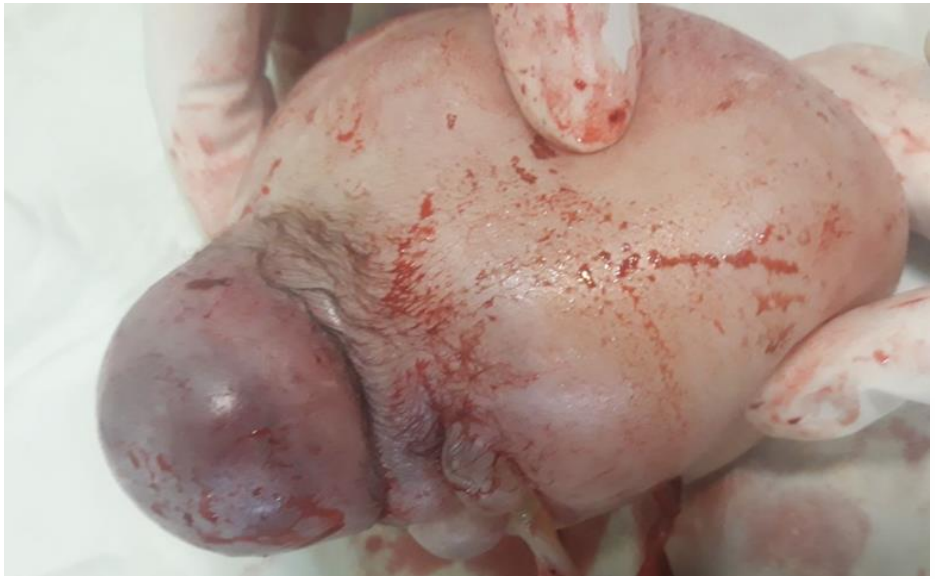


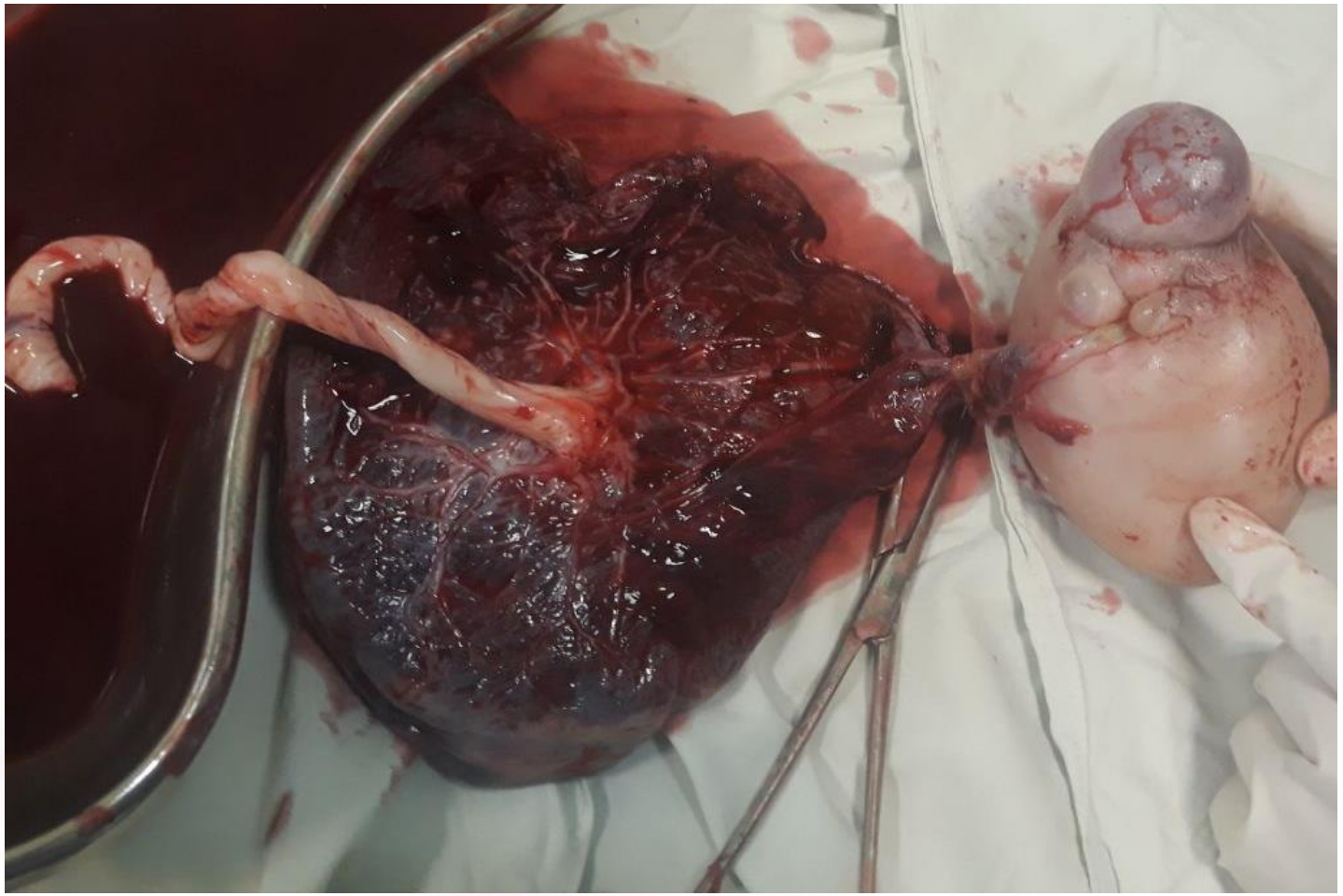


Case 4: 26 wks







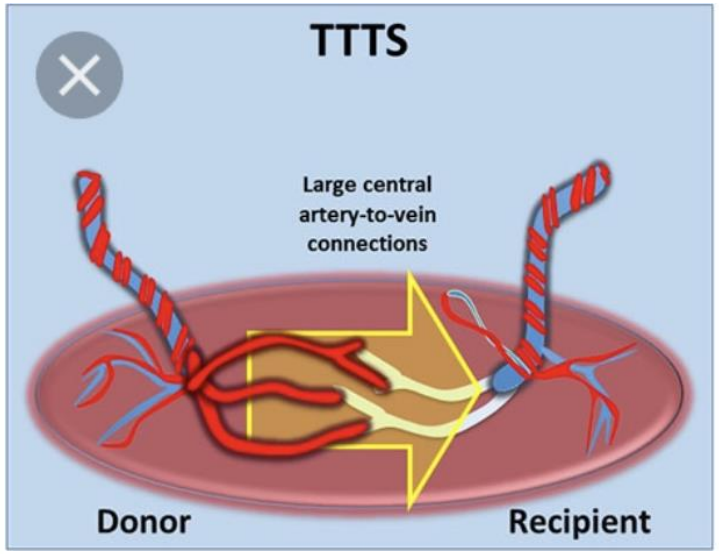


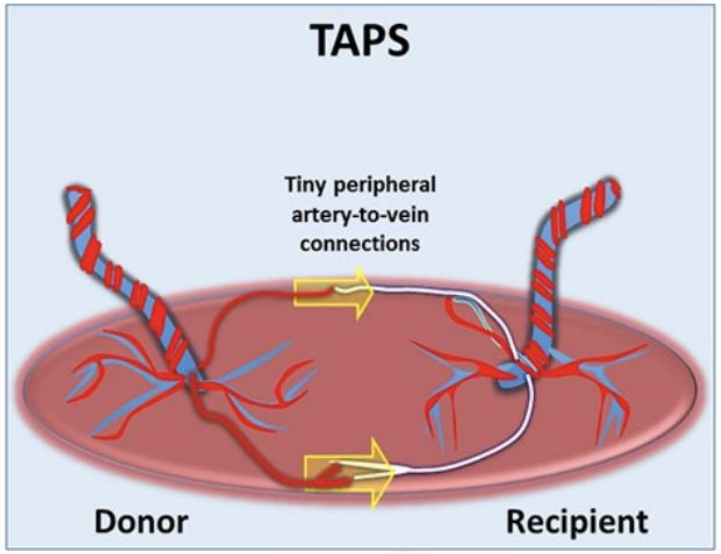
Twin Anemia Polycythemia Sequence (TAPS)

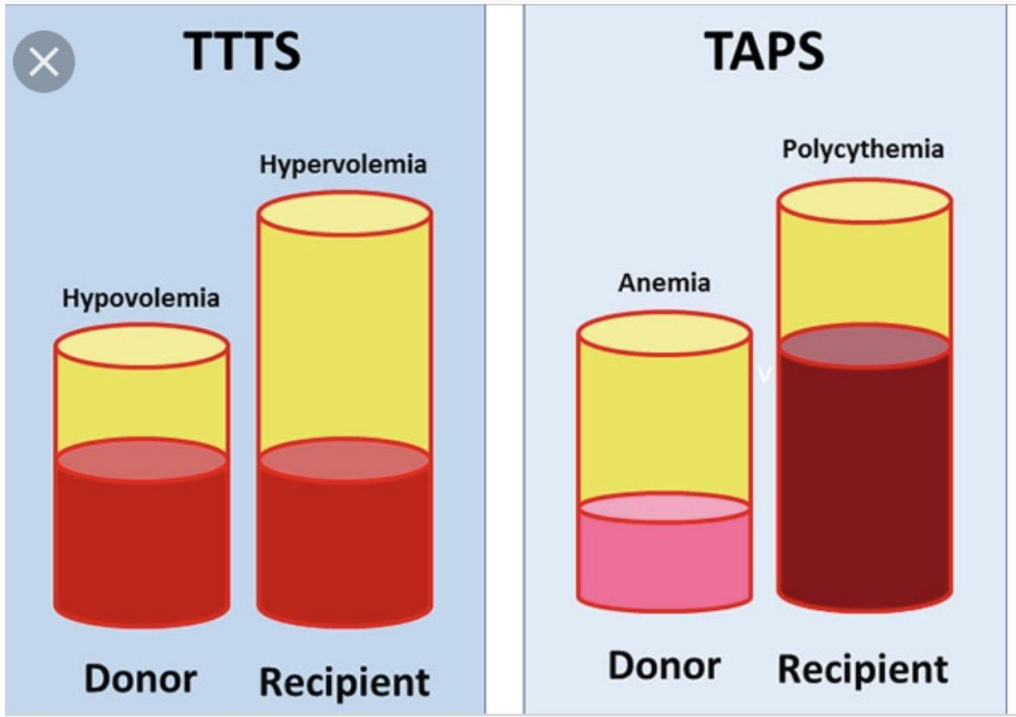
- **Definition** : A form of TTTS in which there is a large intertwin hemoglobin difference, without oligohydramnios-polyhydramnios sequence .
- ***Spontaneous*** : 3 to 6% of previously uncomplicated third trimester MCDA twins . Most cases are diagnosed in the late second or third trimester.
- ***Post-laser*** : TAPS occurs in 2 to 13% of TTTS pregnancies treated with laser ablation up to **six weeks after the procedure** .

Pathogenesis

- In pregnancies treated with laser , the presence of residual anastomoses later developed TAPS.
- In the majority of cases, the residual anastomoses were very small (<1 mm) unidirectional arterio-venous (AV) anastomoses without accompanying arterio-arterial anastomoses.
- severe polycythemia can lead to **fetal and placental thrombosis**, while severe anemia can lead to **hydrops fetalis** .

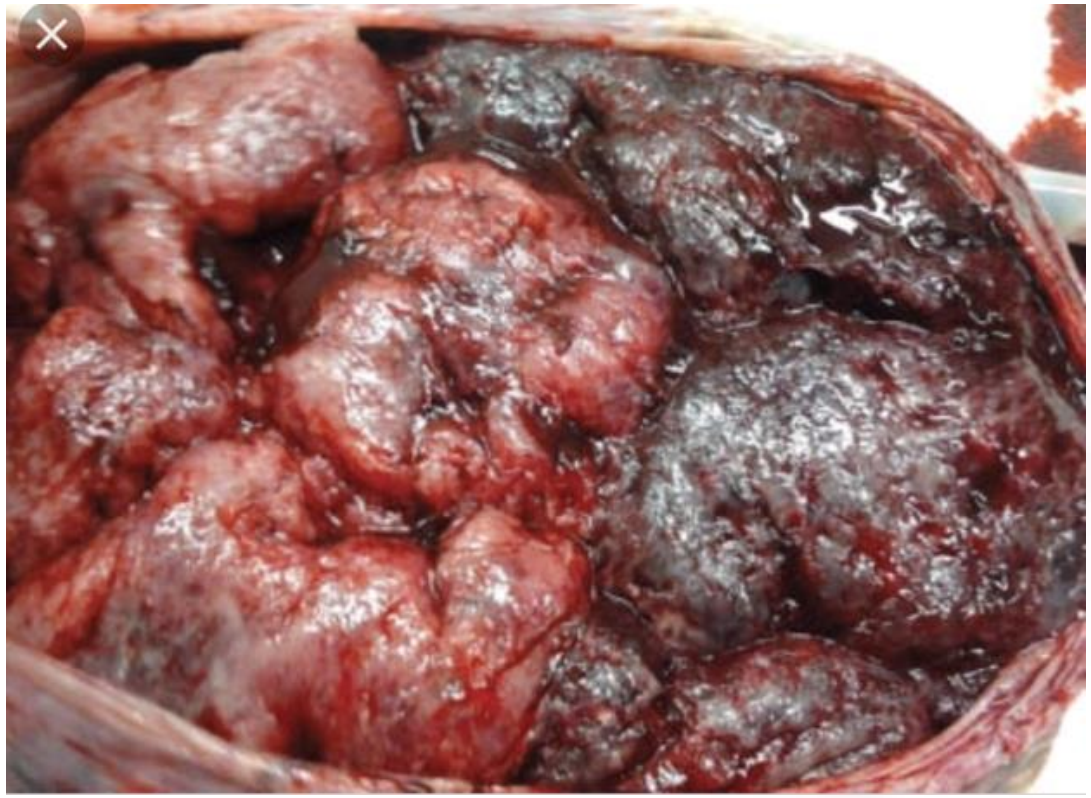


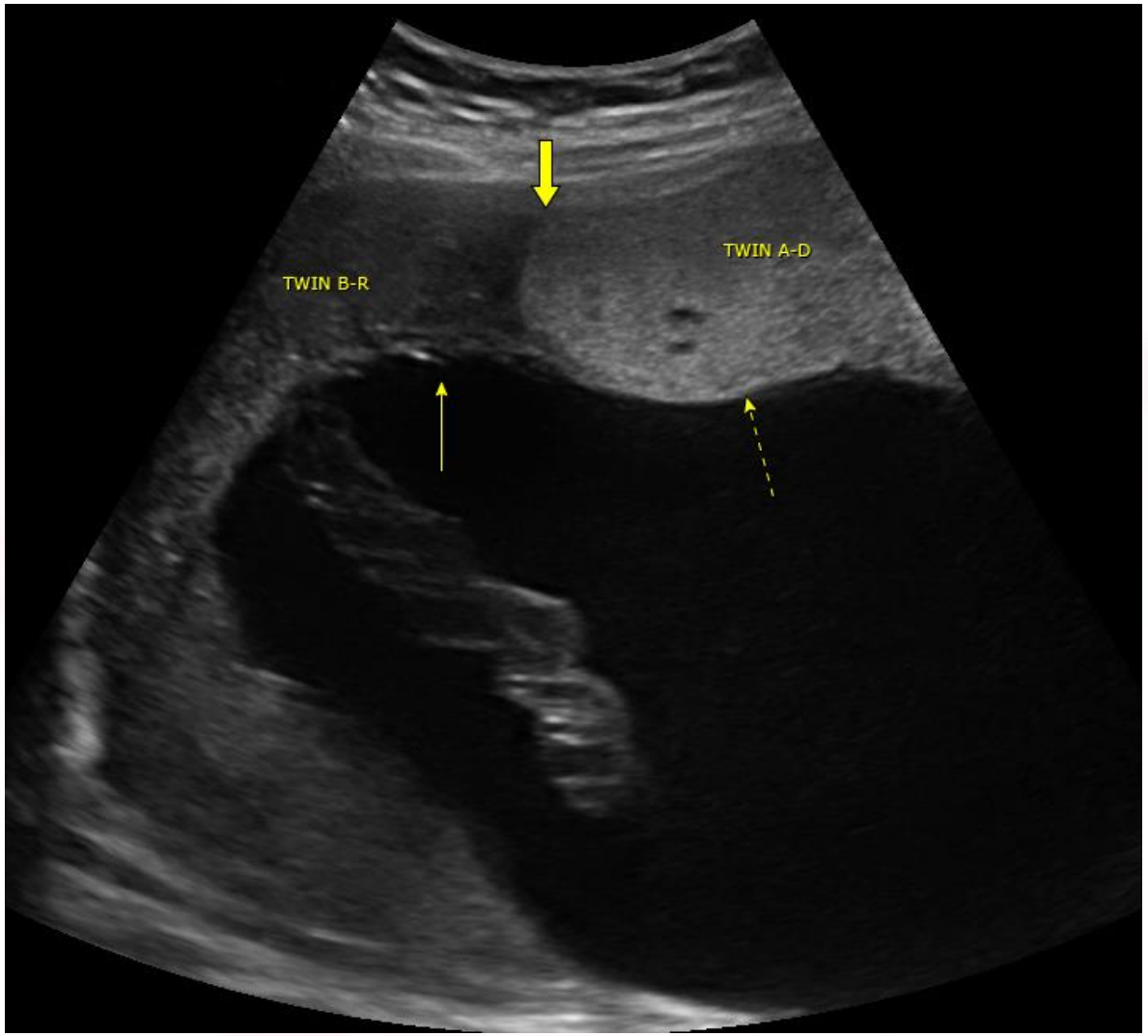


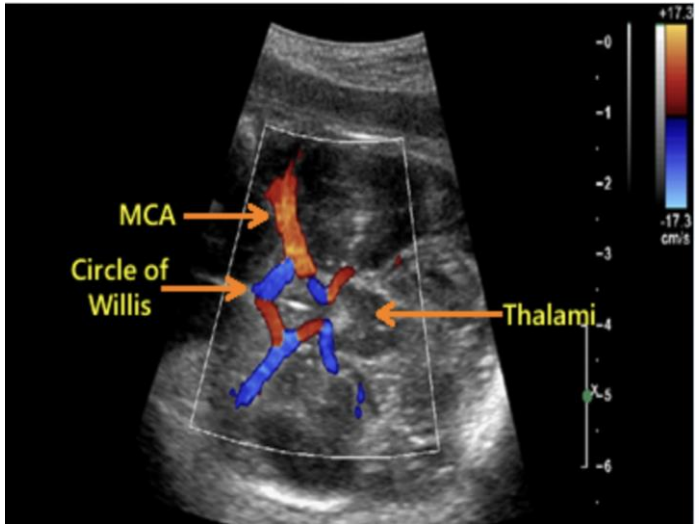


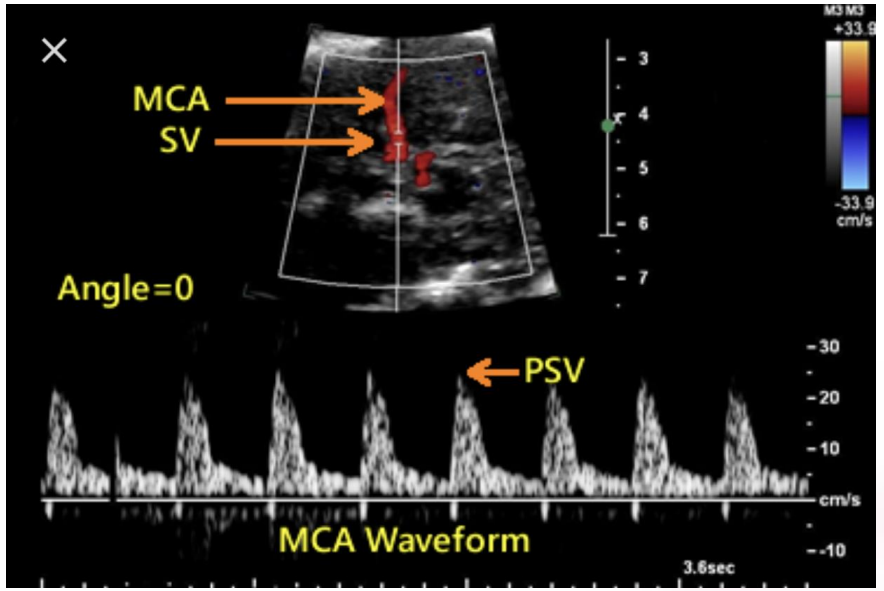
Diagnosis

- TAPS is diagnosed when ***MCA-PSV is >1.5 MoM*** in one twin and ***<0.8 MoM*** in the other twin .
- Placental discordance is noted on ultrasound: **the anemic donor has a thickened hyperechoic placenta and the plethoric recipient has a thinner hypoechoic placenta, with clear demarcation between the donor and recipient territories** .
- There may also be growth discordancy.
- Postnatal diagnosis is made by an ***intertwin hemoglobin difference ≥8.0 g/dL*** in conjunction with ***an intertwin reticulocyte ratio of >1.7*** (reticulocyte count of the donor twin divided by the reticulocyte count of the recipient twin) and a placental injection examination showing very small superficial AV anastomoses .




TWIN B-R
TWIN A-D





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Expected Peak Velocity of Systolic Blood Flow in the MCA as a Function of Gestational Age

The proximal middle cerebral artery is enlarged to to occupy more than 50% of the image and is sampled 2 mm after its origin from the internal carotid artery. The angle of the ultrasound beam and the direction of blood flow should be zero degrees. The risk of anemia is highest in fetuses with a pre-transfusion peak systolic velocity of 1.5 times the median or higher.

ENTER:

Gestational age (weeks) [ ]

Observed MCA Peak Systolic Velocity (cm/sec) [ ]

Calculate Clear Form

Calculations:

The Median Peak Systolic Velocity for this age is [ ]

Your measurement is [ ] **Multiples of Median**

Classification

Table 2 Antenatal and postnatal staging of twin anemia–polycythemia sequence (TAPS)[109,110]

Stage	*Antenatal staging*	*Postnatal staging: intertwin Hb diff (g/dL)*
1	Donor MCA-PSV > 1.5 MoM and recipient MCA-PSV < 1.0 MoM, without other signs of fetal compromise	> 8.0
2	Donor MCA-PSV > 1.7 MoM and recipient MCA-PSV < 0.8 MoM, without other signs of fetal compromise	> 11.0
3	Stage 1 or 2 and cardiac compromise in donor (UA-AREDF, UV pulsatile flow, or DV increased or reversed flow)	> 14.0
4	Hydrops of donor twin	> 17.0
5	Death of one or both fetuses, preceded by TAPS	> 20.0

AREDF, absent or reversed end-diastolic flow; DV, ductus venosus; Hb, hemoglobin; MCA, middle cerebral artery; MoM, multiples of median; PI, pulsatility index; PSV, peak systolic velocity; UA, umbilical artery; UV, umbilical vein.

Screening

- **In order to screen for TAPS, the MCA-PSV should be measured from 20 weeks onwards in both fetuses, and during the follow-up of cases treated for TTTS and cases of sFGR .**
- Outcome may range from double intrauterine fetal demise to the birth of two healthy neonates with a significant inter-twin haemoglobin discordance .

Treatment

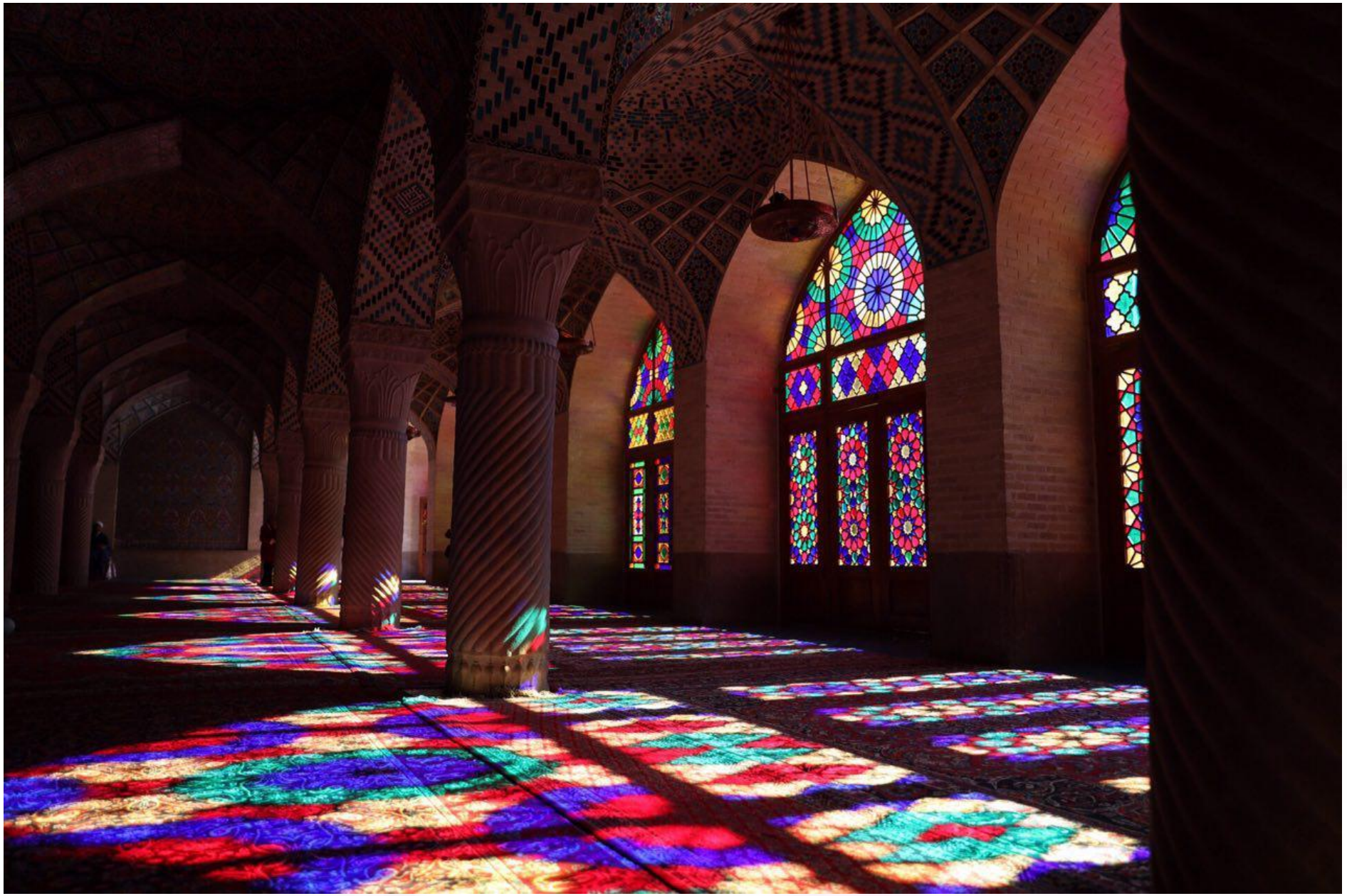
- We reserve treatment for pregnancies with **stage II TAPS (ie, MCA-PSV of >1.7 MoM in one fetus and <0.8 MoM in the other fetus).**
- TAPS after laser ablation has been treated with **repeat laser therapy, in utero fetal transfusion , selective feticide, expectant management, and early delivery.**
- **The management options depend on the gestational age at diagnosis, parental choice, severity of the disease and technical feasibility of intrauterine therapy.**

- In more chronic cases of TAPS, **intrauterine transfusion of red cells** to the anemic fetus can be undertaken.
- **An intraperitoneal approach** is preferred due to allow for the slow absorption of red cells.
- Some centers will undertake **a partial exchange** of the plethoric twin at the same setting to potentially reduce the complications associated with hyperviscosity. In these cases, aliquots of blood are removed and replaced with equal volumes of sterile saline.
- Repeat procedures are based on **subsequent MCA-PSVs**.
- These patients are delivered **at 32 weeks** of gestation, in the absence of complications necessitating earlier delivery

- It appears that the main neonatal morbidity is anemia (requiring transfusion) and polycythemia (requiring partial exchange transfusion) .
- The risk of neurodevelopmental delay in MC twins complicated by TAPS is increased (20%) .
- Therefore, brain imaging during **the third trimester** and neurodevelopmental assessment at the **age of 2 years** are recommended .(isuog)

References

- Up-to-date 2018
- Callen’s ultrasonography in obstetrics and gynecology 2017
- Ultrasound of congenital fetal anomalies of Paladini 2014
- ISUOG 2017
- RCOG guideline 2016



Nasir Al-Mulk Mosque-Shiraz-Iran