

Diagnosis and Treatment of Monochorionic-Diamniotic Twin Complications

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Learning Objectives

- 1 Review the evaluation and current staging criteria in the diagnosis of TTTS/TAPS
- 2 Understand current treatments available for complications of mono-di twins
- 3 Interpret the evaluation and care options for selective fetal growth restriction
- 4 Summarize current recommendations for surveillance in monochorionic twin gestations

Updates in Twin-to-Twin Transfusion Syndrome

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Society for Maternal-Fetal Medicine Consult Series #72: Twin-twin transfusion syndrome and twin anemia-polycythemia sequence

SMFM, R Miller, J Miller, S Obican, L Simpson, SMFM Publications Committee
2024

twins) with this strategy.^{24,25} **We recommend that ultrasound surveillance for TTTS begin at 16 weeks of gestation for all MCDA twin pregnancies and continue at least every 2 weeks until delivery, with more frequent monitoring indicated for clinical concern (GRADE 1C).**^{16,17,26–29} Concerns that should prompt

We recommend that routine sonographic surveillance for TTTS minimally include assessment of amniotic fluid volumes on both sides of the intertwin membrane and evaluation for the presence or absence of urine-filled fetal bladders, and ideally incorporate Doppler study of the umbilical arteries (GRADE 1C). Although middle cerebral artery (MCA) Doppler studies do not have a role in screening for TTTS per se, they are used to screen for TAPS (discussed later in this document), and should therefore be considered with routine surveillance sonograms for MC twins.^{16,22,30–33} Fetal weight assessments are also recommended at least every 4 weeks to evaluate for FGR or intertwin growth discordance.

both sides of the intertwin membrane and evaluation for the presence or absence of urine-filled fetal bladders, and **ideally incorporate Doppler study of the umbilical arteries (GRADE 1C);** (4) we recommend

Twin Monitoring

Dichorionic-Diamniotic

First trimester ultrasound

Genetic Screening as desired

Anatomy Ultrasound

Growth ultrasounds at least once a month

Consider antenatal testing

Delivery at 38 weeks

Monochorionic-Diamniotic

First trimester ultrasound

Genetic Screening as desired

TTTS/TAPS evaluations (fluid, bladder, and MCA Dopplers, UA Dopplers) starting at 16 weeks and then every 2 weeks

Anatomy Ultrasound

Growth ultrasounds at least once a month

Antenatal testing

Delivery at 37 weeks

Monochorionic-Monoamniotic

First trimester ultrasound

Genetic Screening as desired

Ultrasound evaluations starting at 16 weeks and then at least every 2 weeks (MCA Dopplers, UA Dopplers)

Anatomy Ultrasound

Hospital admission at "viability"

Growth ultrasounds at least once a month

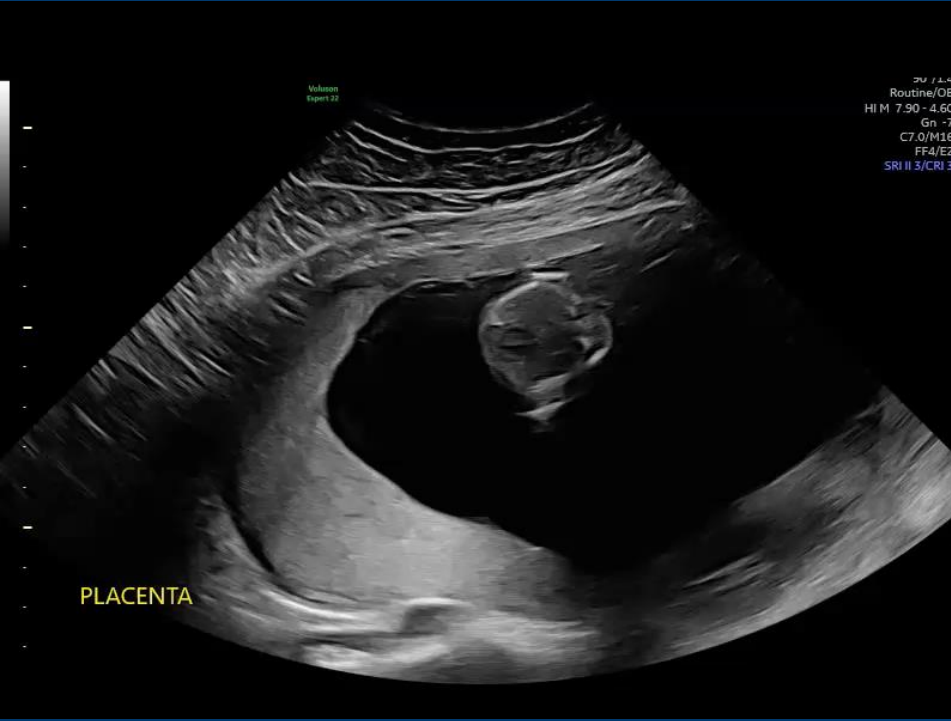
Delivery at 32-34 weeks

Screening in Monochorionic Twins

- First Trimester Ultrasound
 - Consider increased surveillance if NT discrepancy
- 16 Weeks until Delivery:
 - Growth ultrasounds every 4 weeks
 - TTTS/TAPS Evaluations at least every 2 weeks
 - Amniotic fluid level maximum vertical pockets
 - MCA Dopplers
 - Recent recommendation for Umbilical Artery Dopplers



- All advocate for Umbilical Artery Doppler and MCA Doppler monitoring
- Some qualify starting at 20 weeks while others start at 16 weeks
- Lack of clear evidence for monitoring of UAs
 - Based on small studies and expert opinion



Be suspicious if a fetus is defying gravity and doesn't leave the anterior uterine wall



TWB

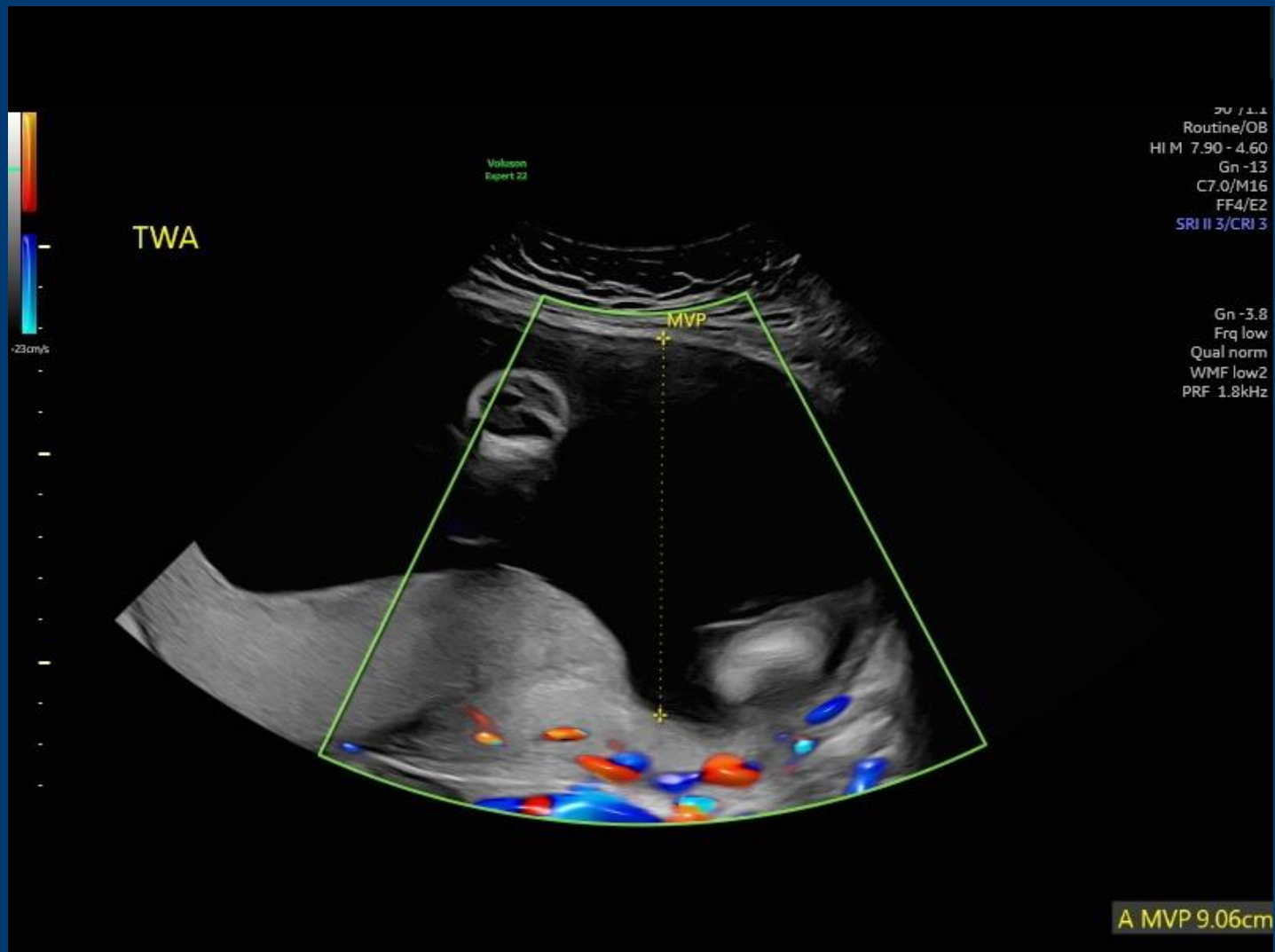
Voluson
Expert 22

80 7.1.17
Routine/OB
HI M 7.90 - 4.60
Gn -5
C7.0/M16
FF4/E2
SRI II 3/CRI 3

TWB

Voluson
Expert 22

80 7.1.17
Routine/OB
HI M 7.90 - 4.60
Gn -1
C7.0/M16
FF4/E2
SRI II 3/CRI 3



Twin-Twin Transfusion Syndrome

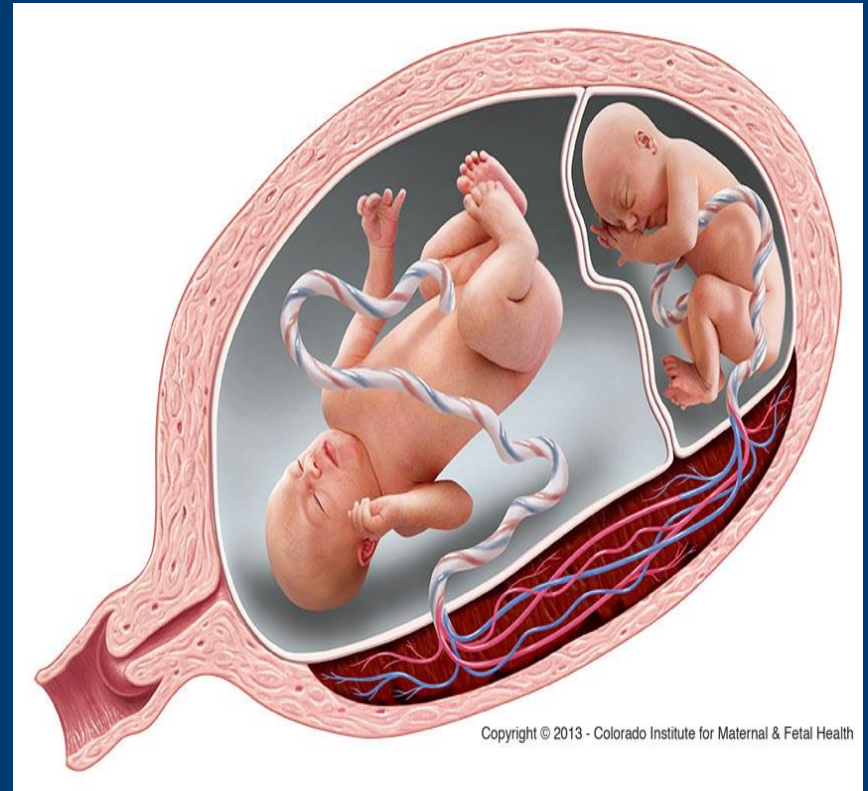
Disease in which monochorionic twins (shared placenta) have communicating vessels that manifest a compromise in the pregnancy

The name is a misnomer – more likely a manifestation of a volume discrepancy between fetuses

- Can cause overload and cardiac compromise in one twin

Occurs in 15% of monochorionic twin pregnancies

Associated with at least an 80% risk of mortality if left untreated



Twin-Twin Transfusion Syndrome

Quintero et al. Journal of Perinatology (1999); 19: 550-555

Stage	Recipient	Donor
1	MVP >8 cm	MVP <2 cm
2	Visible bladder	No bladder filling
3	UA A/REDV, DV absent/reversed a-wave, UV pulsations in either twin	
4	Hydrops of either twin	
5	Single or double fetal demise	

A/REDV, absent/reversed end diastolic velocity; DV, ductus venosus; MVP, maximum vertical pocket; UA, umbilical artery; UV, umbilical vein.



Low Suspicion for TTTS:

- Clear intertwin membrane seen
- Subjectively normal fluid for each fetus



TWIN B / D

Voluson
E10

bu7/1b
Routine 2 Trim./OB
HI H PI 6.70 - 3.30
Gn 2
C6/M5
FF3/E2
SRI II 3/CRI 1

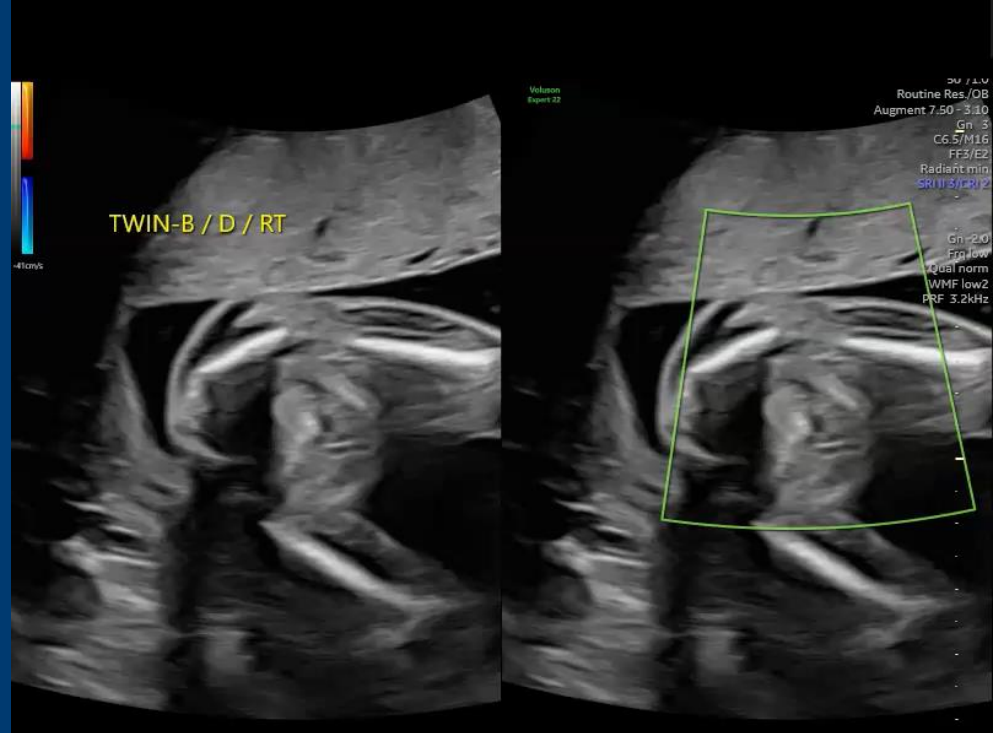


Quintero Criteria: MVP < 2 cm

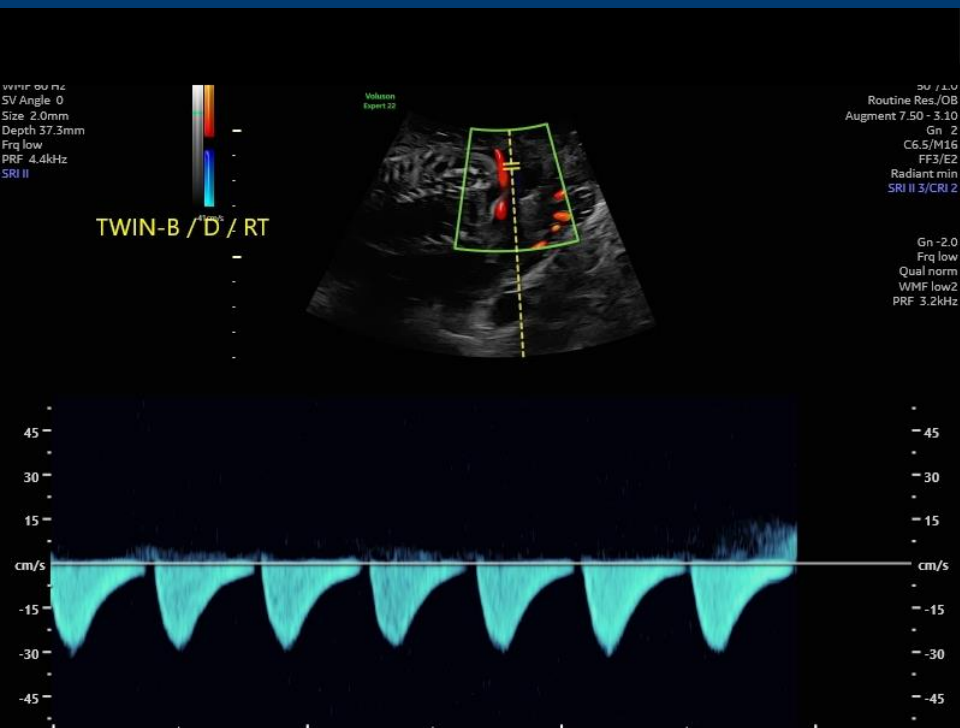


Sometimes we see a fetus
with obvious membrane but
no fluid:
“lollipop” appearance

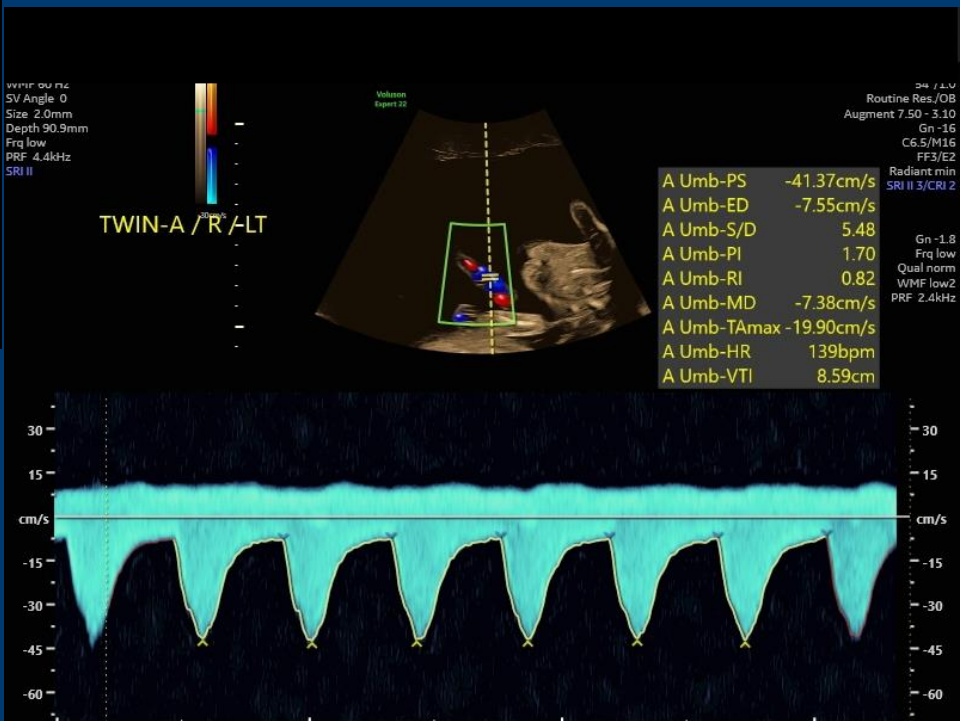
Evaluate Bladder with color
Doppler
"Absent bladder" is Quintero
Criteria



Recipient Bladder size is not
Quintero Criteria but it often
appears enlarged or
prominent as below



Umbilical Artery Dopplers showing Absent End Diastolic Flow in the Donor and increased resistance in the Recipient



/R



Evaluate for fetal hydrops

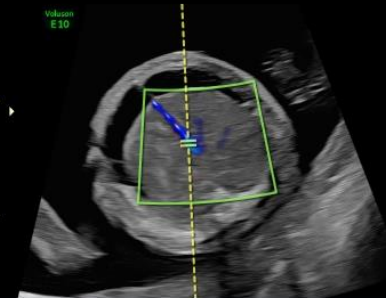
TWIN A/R



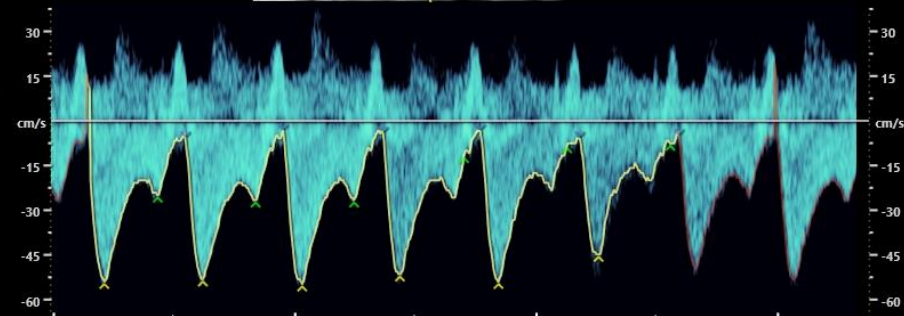
Hydrops and Heart Dysfunction are Key Parts of Evaluation



TWIN A/R



A DV-D	-18.20cm/s
A DV-a	-3.47cm/s
A DV-TAmax	-25.11cm/s
A DV-S/a	15.27
A DV-a/S	0.07
A DV-PI	1.97
A DV-PLI	0.93
A DV-PVIV	2.72



TWIN A/R



Cardiomyopathy:

- Heart appears less efficient
- Less “squeeze” than expected
- Abnormal Myocardial Performance Index

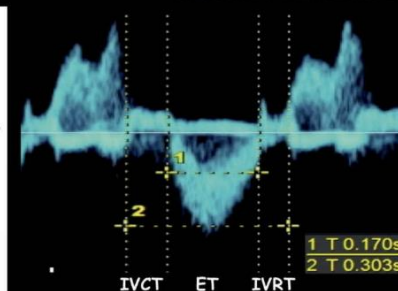
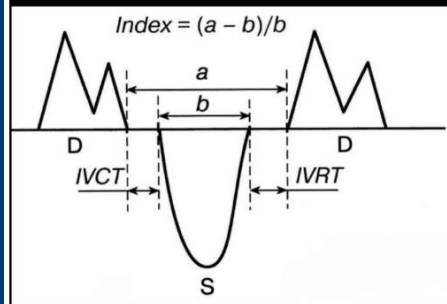
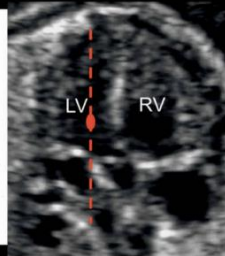
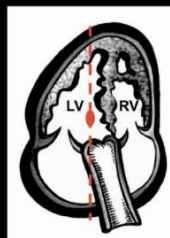
$$MPI = (IVCT + IVRT) / ET$$

ET - ejection time

IVRT - isovolumetric relaxation time

IVCT - isovolumetric contraction time

Normal value: ~ 0.36





Poor Prognosis:

- Abnormal umbilical artery Dopplers due to poor heart function
- Pulsatile umbilical vein due to poor right heart filling



Twin-Twin Transfusion Syndrome

Cardiac Evaluation

Stage	Donor	Recipient	Recipient cardiomyopathy
Cincinnati staging system			
I	Oligohydramnios (DVP <2 cm)	Polyhydramnios (DVP >8 cm)	No
II	Absent bladder	Bladder seen	No
III	Abnormal Doppler findings	Abnormal Doppler findings	None
IIIa			Mild
IIIb			Moderate
IIIc			Severe
IV	Hydrops	Hydrops	
V	Death	Death	
Classifications of recipient cardiomyopathy			
Variables/cardiomyopathy	Mild	Moderate	Severe
Atrioventricular valve regurgitation	Mild	Moderate	Severe
Right/left ventricular hypertrophy	Mild	Moderate	Severe
Myocardial performance index ^a	> +2 Z-score	> +3 Z-score	> +4 Z-score or severe biventricular dysfunction
Left	>0.43	>0.48	>0.53
Right	>0.48	>0.56	>0.64

DVP, deepest vertical pocket.

While not part of staging criteria,
cervix length matters in TTTS
Evaluation

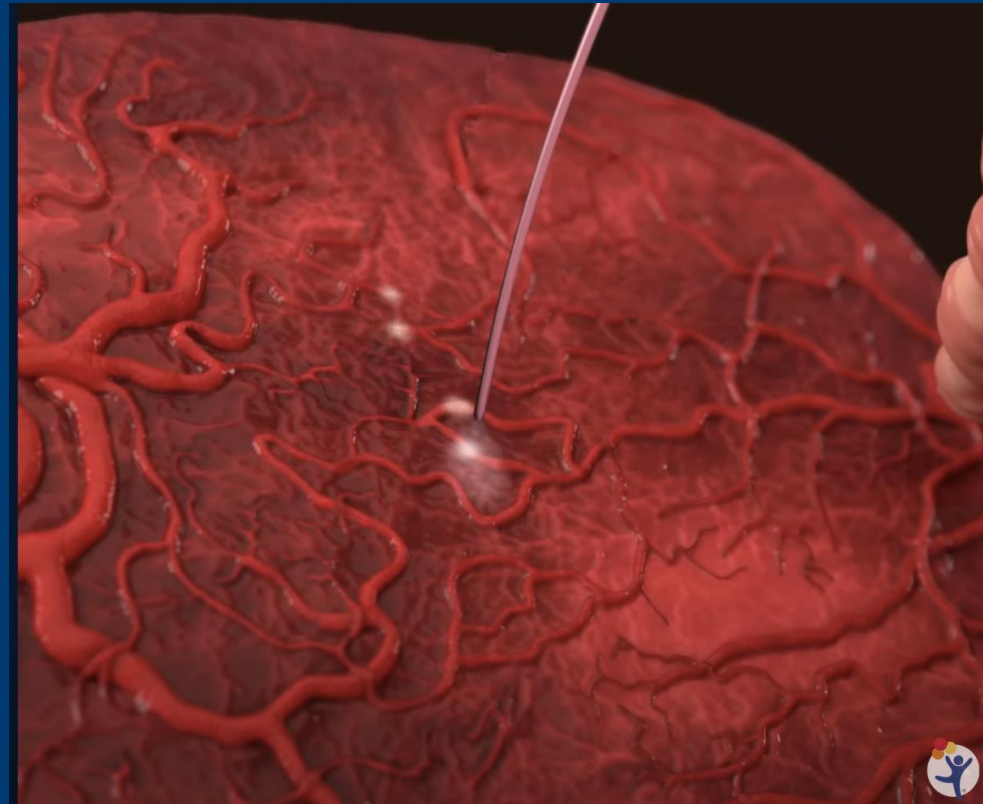
- Evaluate transvaginal ultrasound
- Sometimes amnioreduction with cerclage is recommended
- We always evaluate cervix length before and after laser procedure



Twin-Twin Transfusion Syndrome

Selective Fetoscopic Laser Photocoagulation

- Available from 16w to 26w6d
- Fetoscopic Laser Photocoagulation has become the standard treatment in advanced TTTS
- A small camera is inserted through the skin, through the uterus and into the amniotic sac
- Vascular “mapping” is done in order to only treat pathologic connections
- Yag laser from a 650 nm sheath treats the connections



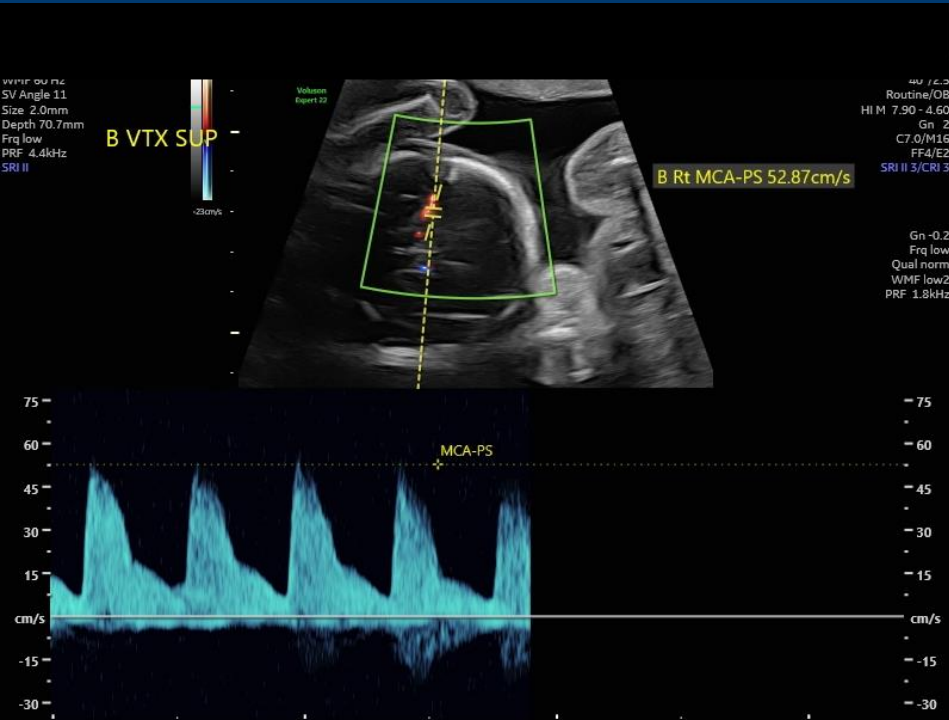
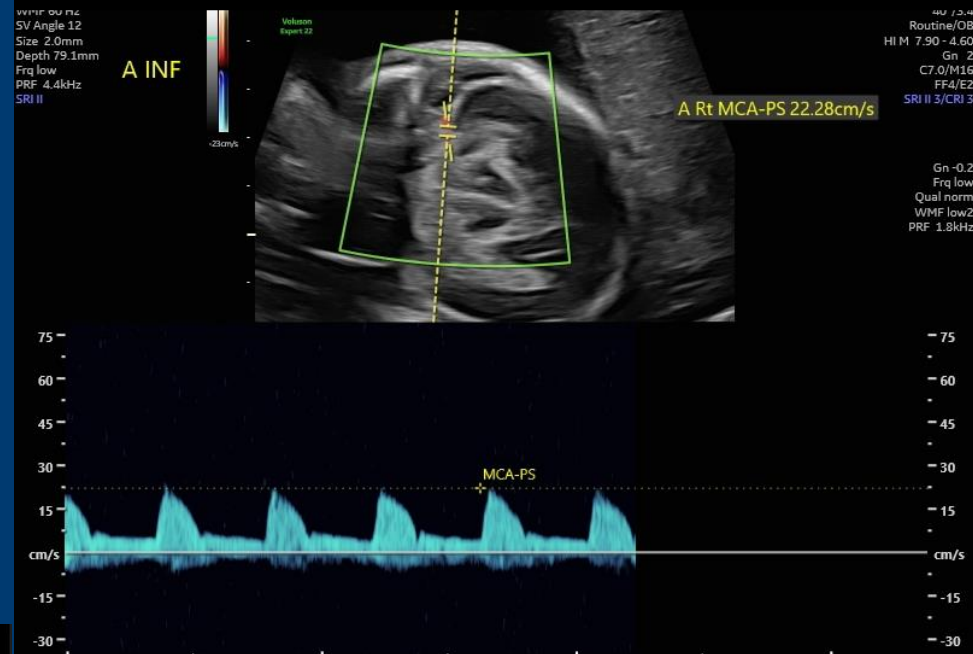
Twin Anemia Polycythemia Sequence (TAPS)

Society for Maternal-Fetal Medicine Consult Series #72: Twin-twin transfusion syndrome and twin anemia-polycythemia sequence

SMFM, R Miller, J Miller, S Obican, L Simpson, SMFM Publications Committee
2024

we recommend that providers consider incorporating middle cerebral artery Doppler peak systolic velocity determinations into all monochorionic twin ultrasound surveillance beginning at 16 weeks of gestation (GRADE 1C); and (14) consultation with a specialized fetal care center is recommended when twin anemia-polycythemia sequence progresses to a more advanced disease stage (stage $\geq$ II) before 32 weeks of gestation or when concern arises for coexisting complications such as twin-twin transfusion syndrome (Best Practice).

TAPS



Same technique as regular MCA Dopplers

- Ideal to be down the barrel with minimal angle correction (< 15 degrees)
- Take Peak Systolic Velocity without auto measurement
- Calculate the PSV MoM



May see a congested liver with a “starry appearance” as a sign of heart dysfunction or TAPS

Current Staging Criteria for TAPS

Stage	Slaghekke, 2010 [21]	Tollenaar, 2019 [36]	Delphi ^a [37]	Postnatal staging
MCA discordance		0.375 MoM	0.5 MoM	
1	MCA-PSV >1.5 MoM in donor and <1.0 MoM in recipient	MCA-PSV discordance of >0.5 MoM	MCA-PSV discordance of >1 MoM; or MCA-PSV >1.5 MoM in donor and <0.8 MoM in recipient	Hemoglobin difference >8 g/dL
2	MCA-PSV >1.7 MoM in donor and <0.8 MoM in recipient	MCA-PSV discordance of >0.7 MoM		Hemoglobin difference >11 g/dL
3	MCA findings as in stage 1 or 2, AREDF UA, increased PI or reversed a-wave DV, pulsatile UV in either twin	Same as Slaghekke, 2010		Hemoglobin difference >14 g/dL
4	MCA findings as in stage 1 or 2 and hydrops in the donor	Same as Slaghekke, 2010		Hemoglobin difference >17 g/dL
5	Demise in either twin preceded by TAPS	Same as Slaghekke, 2010		Hemoglobin difference >20 g/dL

TAPS Treatment Options



Observation with increased surveillance frequency



Laser treatment



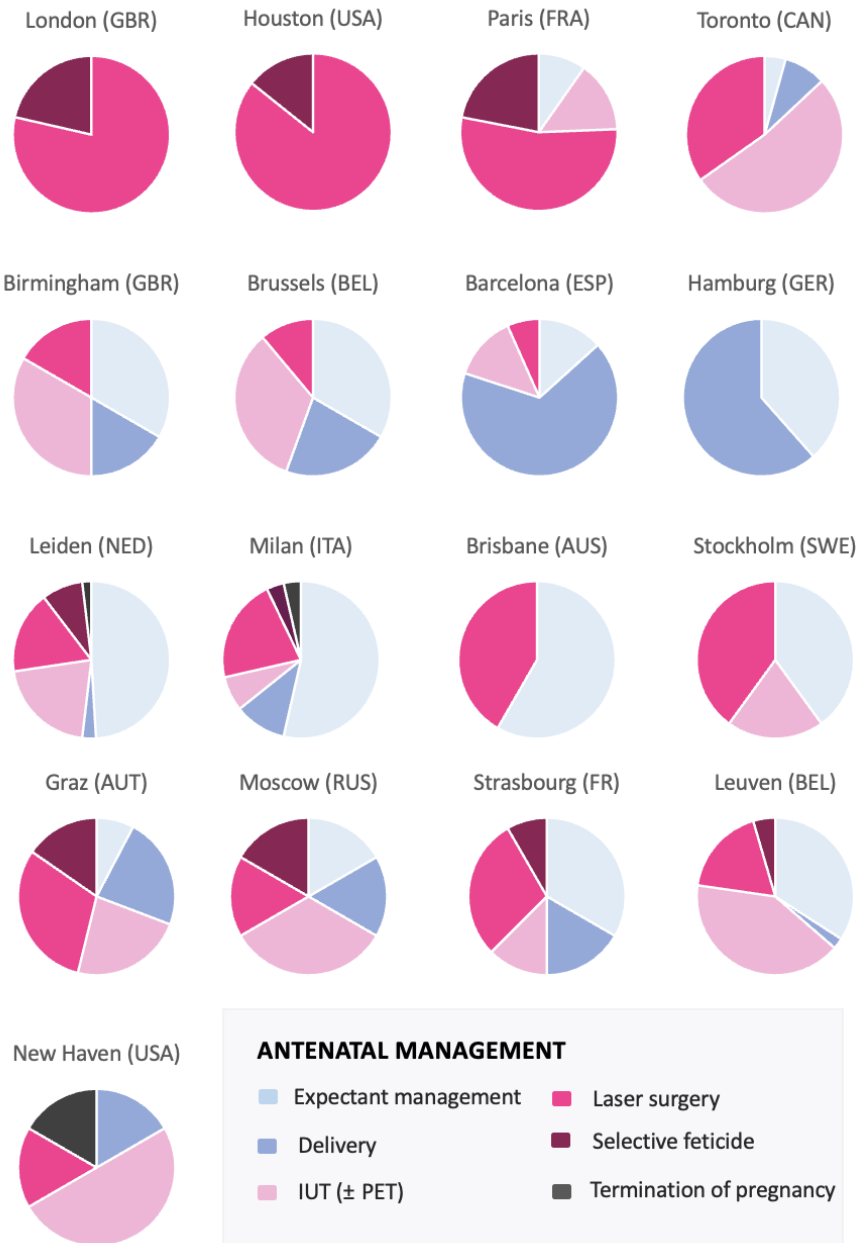
Intrauterine transfusion +/- exchange transfusion



Selective reduction



Delivery



- Significant differences in management approach
 - Most have significant amount of expectant management
 - Not all centers offer intrauterine transfusion

Selective Intrauterine Growth Restriction

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Selective Intrauterine Growth Restriction Delphi Criteria

- Monochorionic Twin Pregnancy with at least 2 of the following:
 - EFW of one twin $< 10^{\text{th}}$ percentile
 - Abdominal circumference of one twin $< 10^{\text{th}}$ percentile
 - EFW discordance $\geq 25\%$
 - Umbilical artery pulsatility index in smaller twin $> 95^{\text{th}}$ percentile
- Incidence: Up to 15% of monochorionic twins

Voluson
Expert 22

09.11.08
Routine Res./OB
HI H PI 9.60 - 3.90
Gn 2
C6.5/M7
FF3/E2
Radiant min
SRI II 2/CRI 2

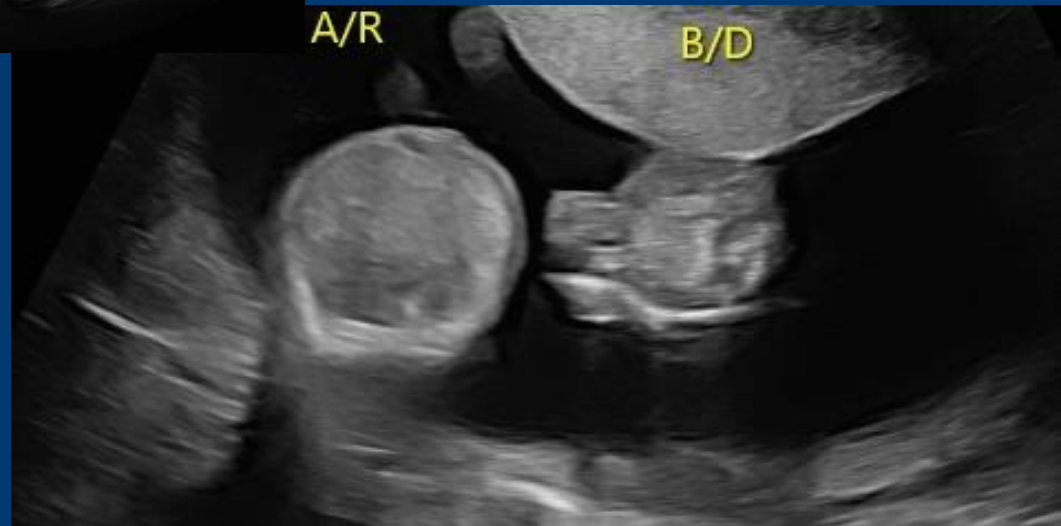
A

B

A/R

B/D

Subjective
discrepancy in
abdominal
circumference size



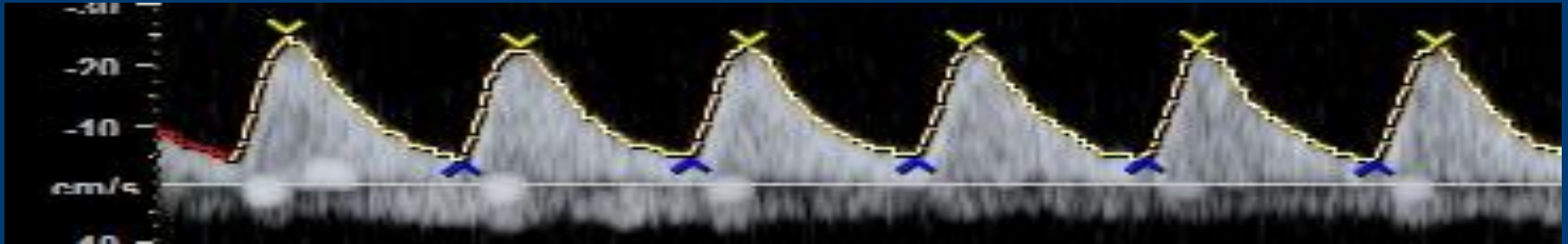
Selective Intrauterine Growth Restriction

- Associated with:
 - Preterm birth
 - Neonatal morbidity
 - Intrauterine fetal death
- **The death of one twin leads up to an estimated 50% incidence of neurologic injury or death of the cotwin**

Classification of Selective Intrauterine Growth Restriction

- Described by Gratacos et al separated into 3 types based on umbilical artery Doppler patterns
- Generally accepted criteria used in describing sIUGR and associated expectations
- Outcomes described with each pattern

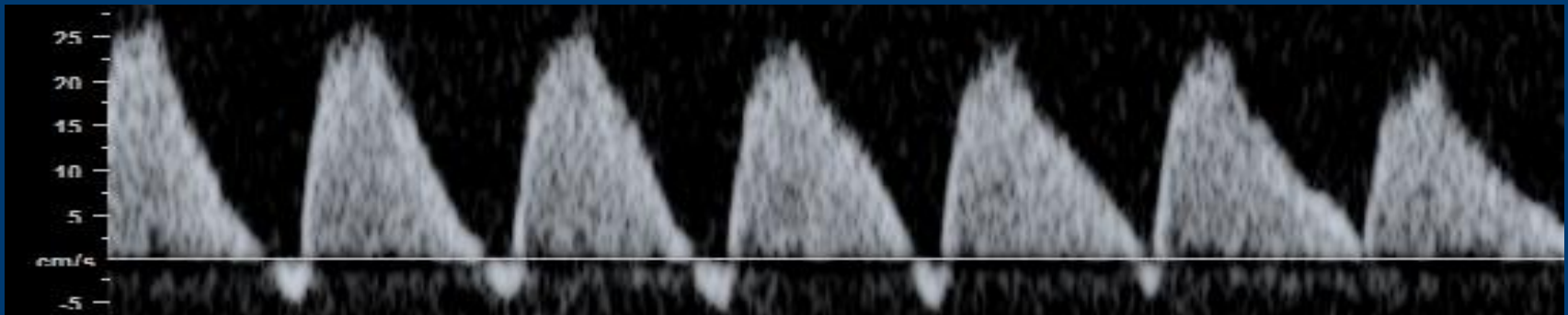
Type 1: Positive end diastolic flow in UA



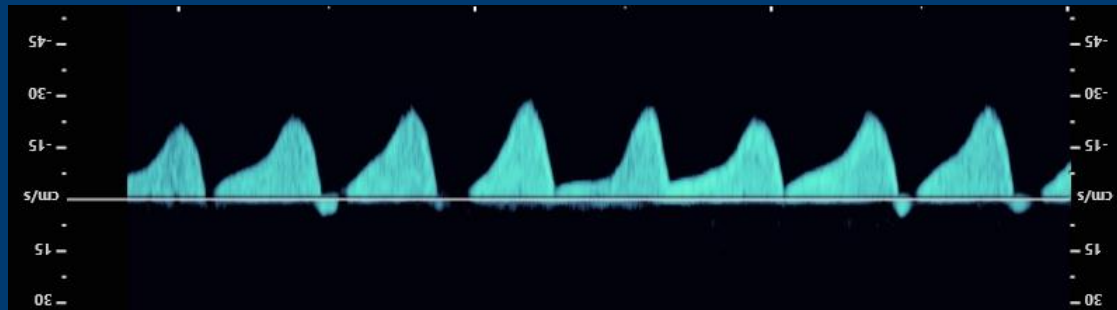
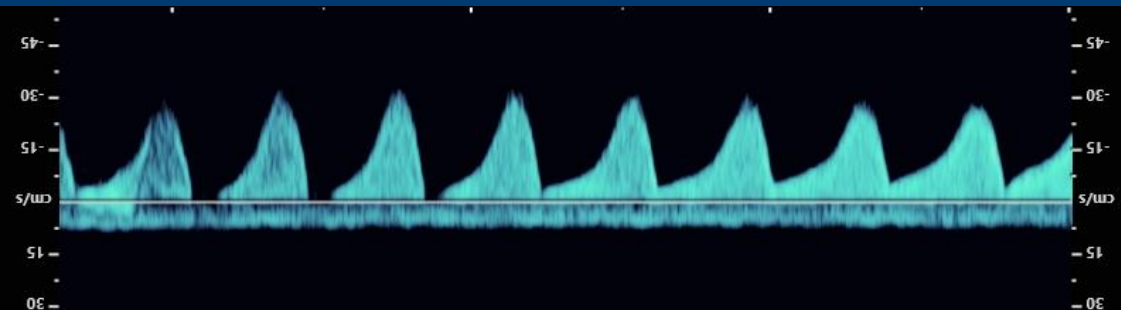
Type 2: Absent or reversed end diastolic flow throughout entire examination



Type 3: All three patterns seen through the examination (forward flow, absent flow, and reversed flow)

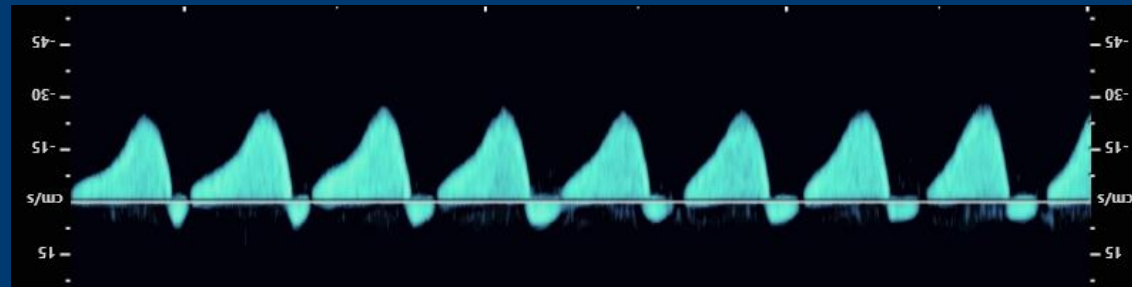


Technique

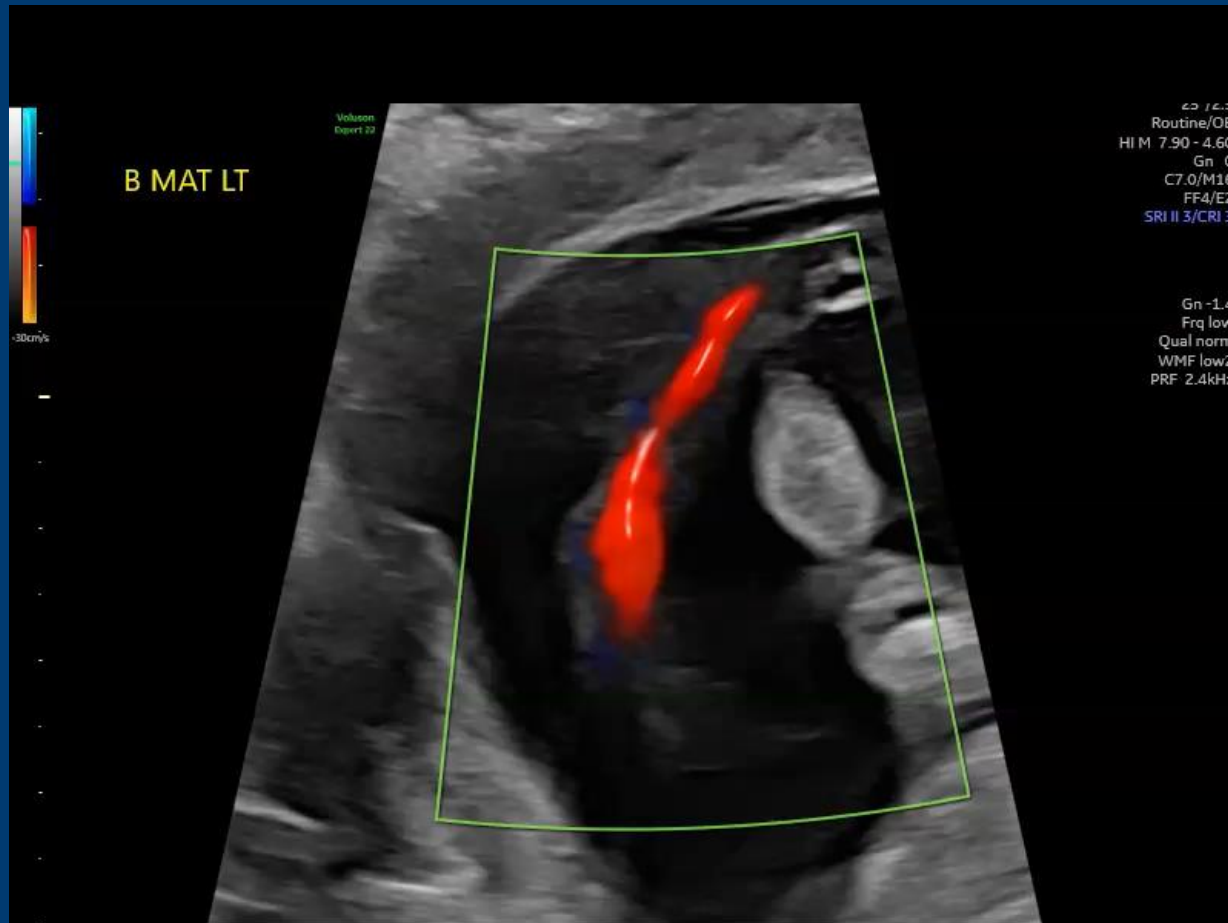


UA Doppler Recommendations:

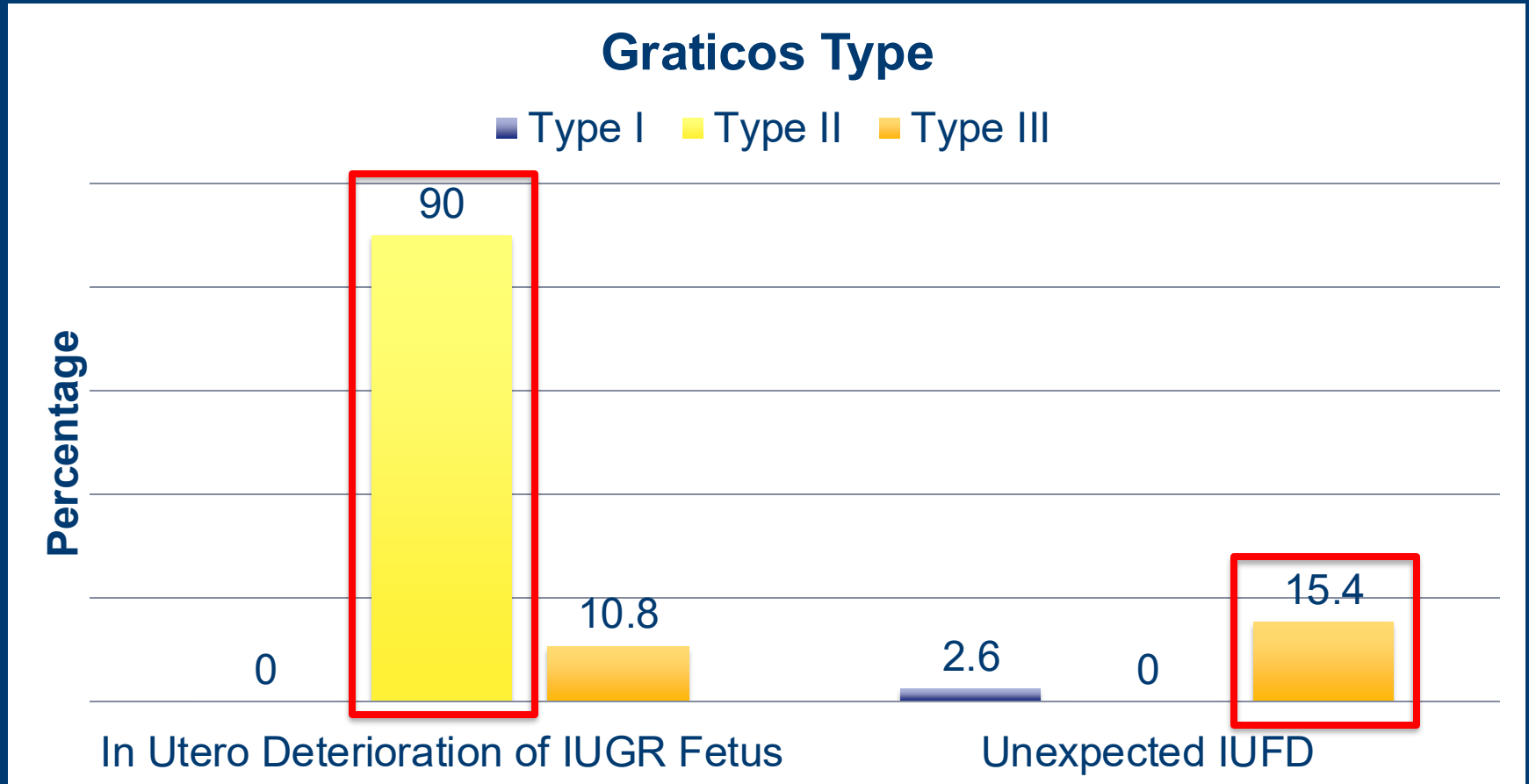
- Color Doppler
- Zoom up so that your gate is within the vessel completely
- Watch for at least 3 full cycles across the screen



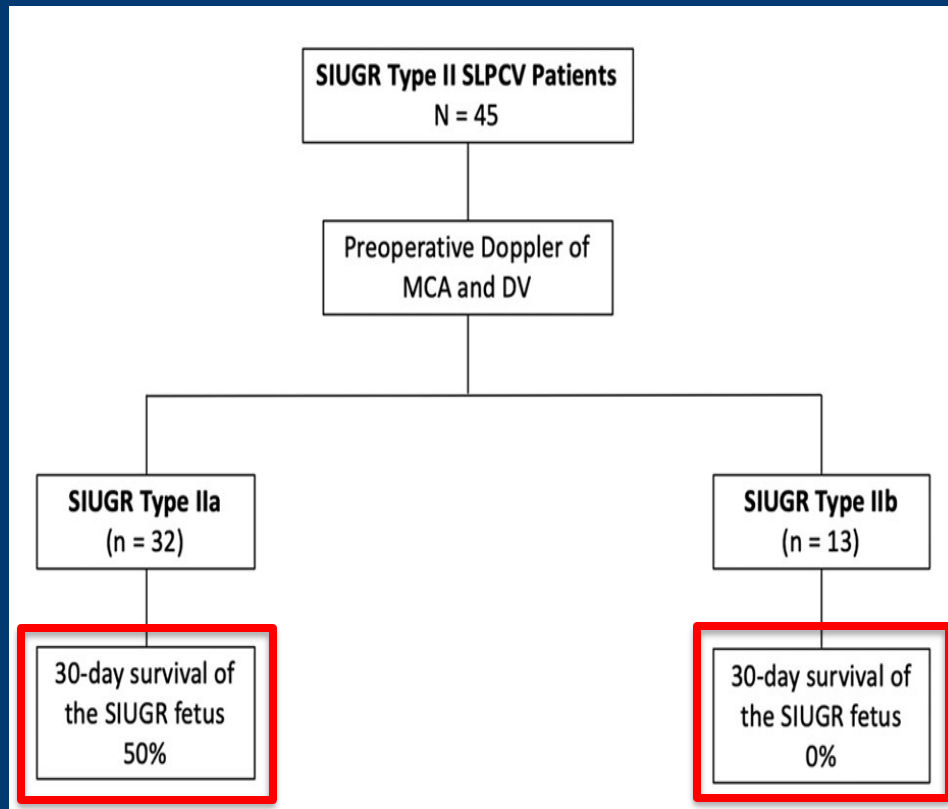
You can often see the “flash” of abnormal Doppler flow



Morbidity/Mortality by sIUGR Type



Subclassification of Gratacos Type II sIUGR



Type IIa:

- MCA PSV < 1.5
- DV with forward a-wave

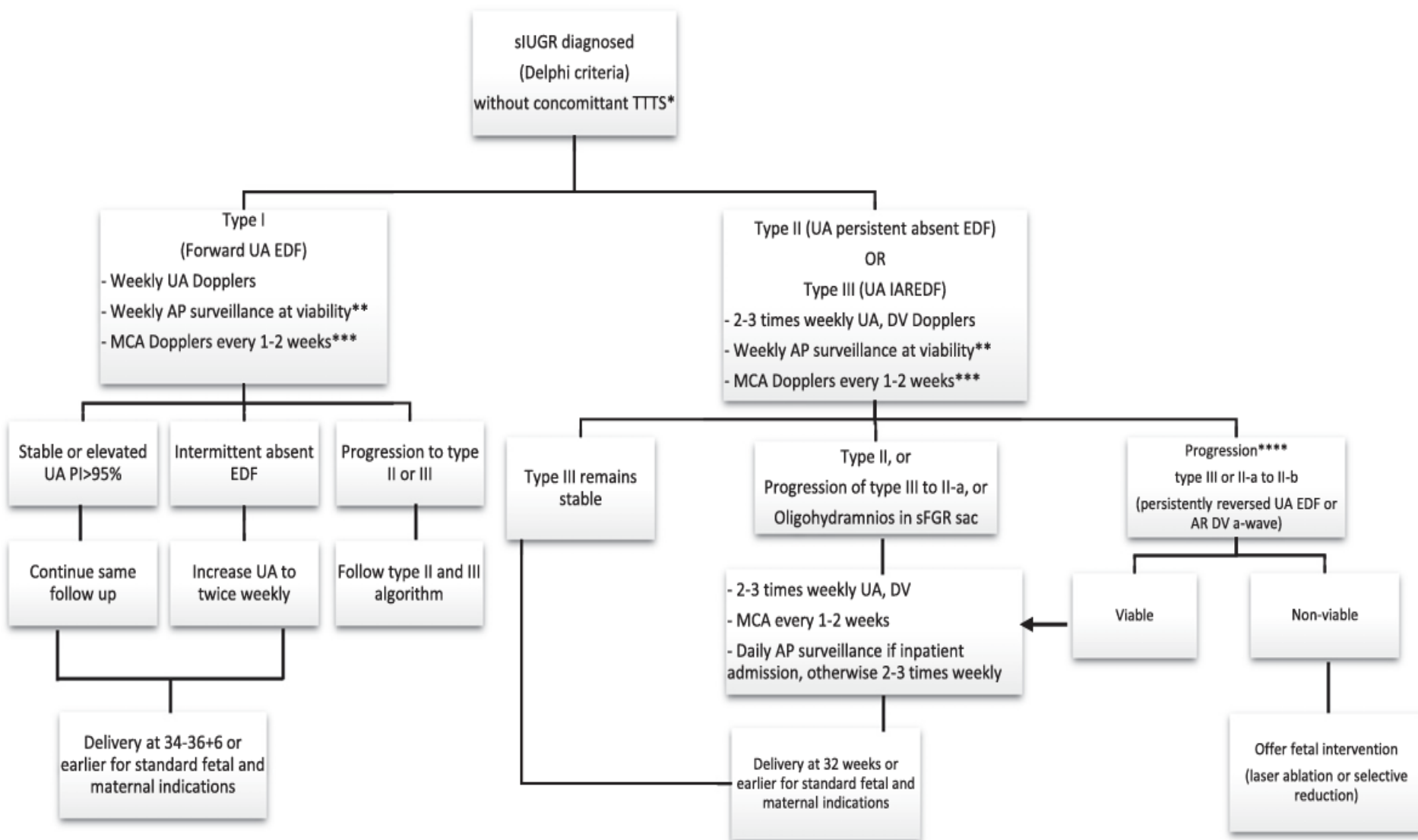
Type IIb:

- MCA PSV ≥ 1.5 AND/OR
- DV with intermittent or persistent absent or reversed a-wave

Selective Intrauterine Growth Restriction Management



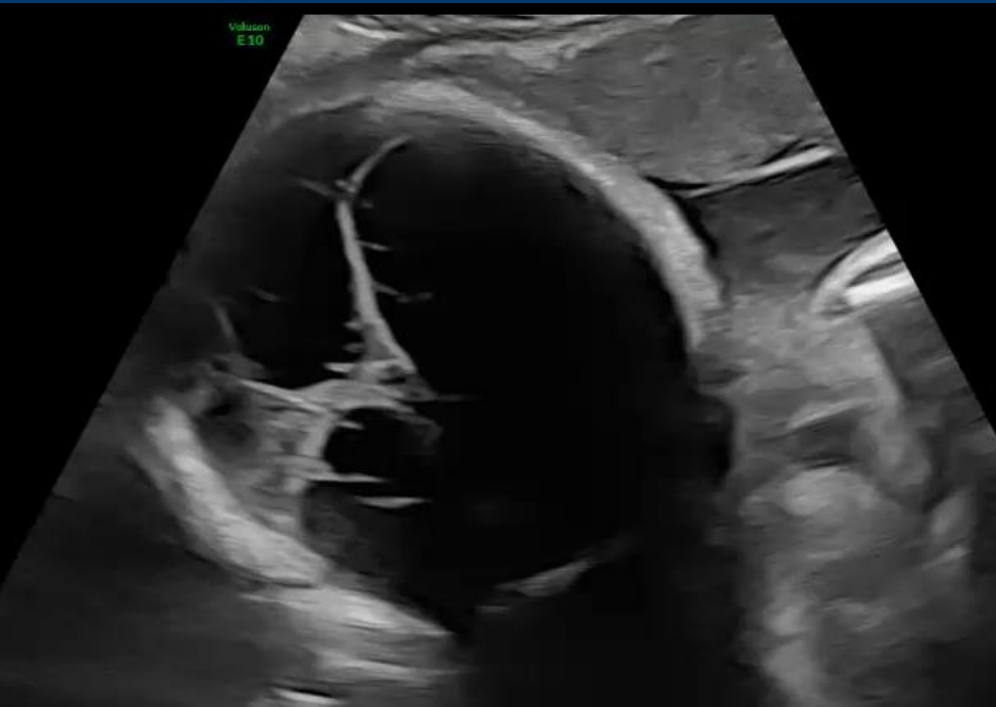
Algorithm for Management of sIUGR



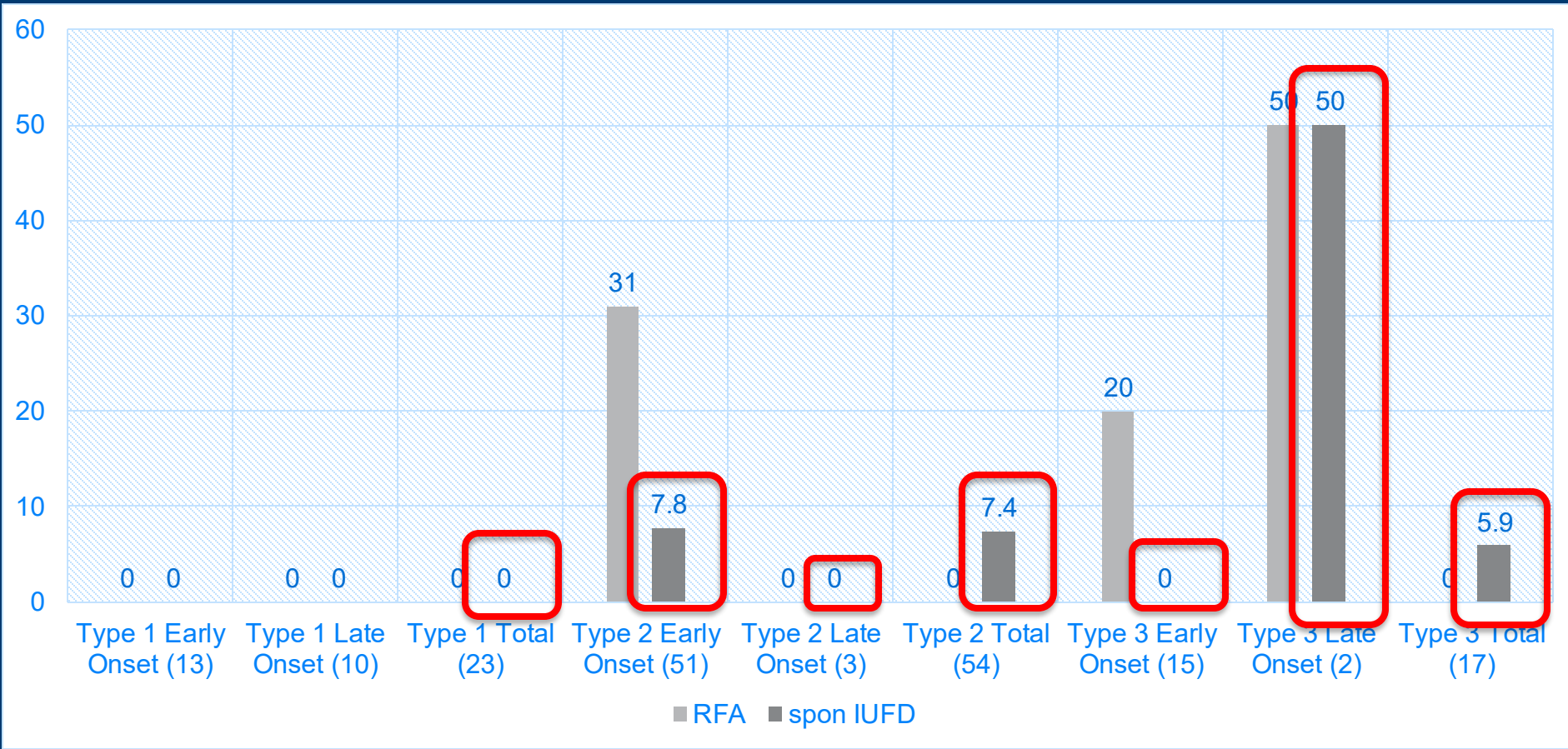
Why does Demise of one Fetus Matter?

- Various studies show different statistics but with most settling on:
 - At least 25 % chance of demise of the cotwin around time of the first loss
 - At least 25 % chance of neurodevelopmental injury of surviving fetus
 - We quote “Up to 50% chance of loss or neurodevelopmental injury to the cotwin”
- After Diagnosis of Twin Demise:
 - Weekly ultrasounds of surviving fetus
 - Consider fetal brain MRI 3-4 weeks post diagnosis
 - Injury usually not seen for at least 1-2 weeks post demise

- 2 weeks post cotwin demise in
Selective Fetal Growth Restriction:
- Severe ventriculomegaly suspected
 - Brain volume loss due to nonperfusion



Gratacos Type sFGR and Spontaneous IUFD Rate (%)



Cynthia K. Wautlet, MD, MPH¹, Emma Peek, BS¹, Lori Silveira, MS, PhD², Nicholas Behrendt, MD^{1,3}, Henry L. Galan, MD^{1,3}, Michael Zaretsky, MD^{1,3}

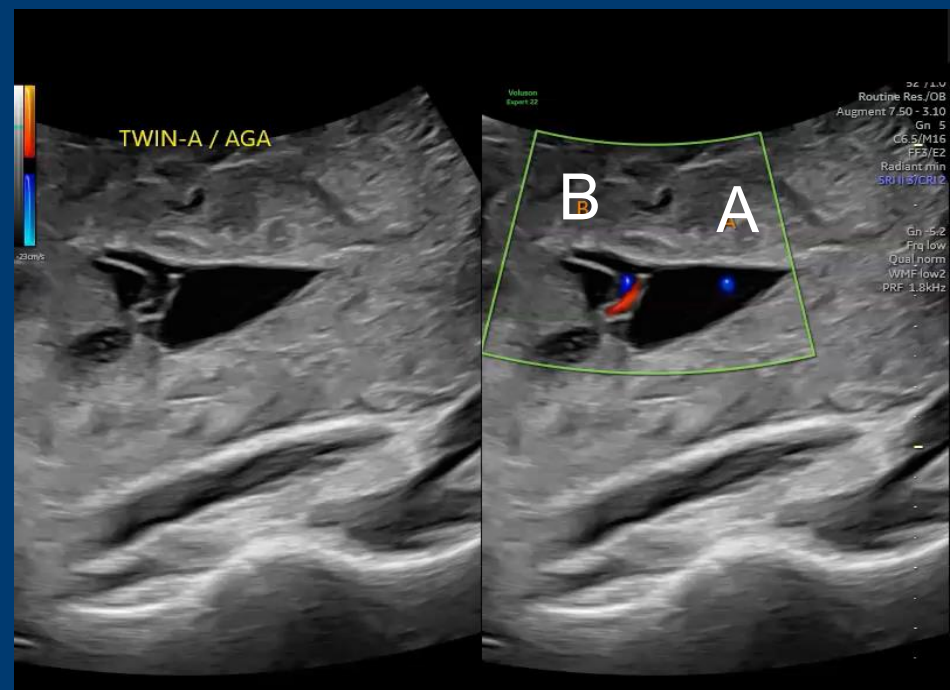
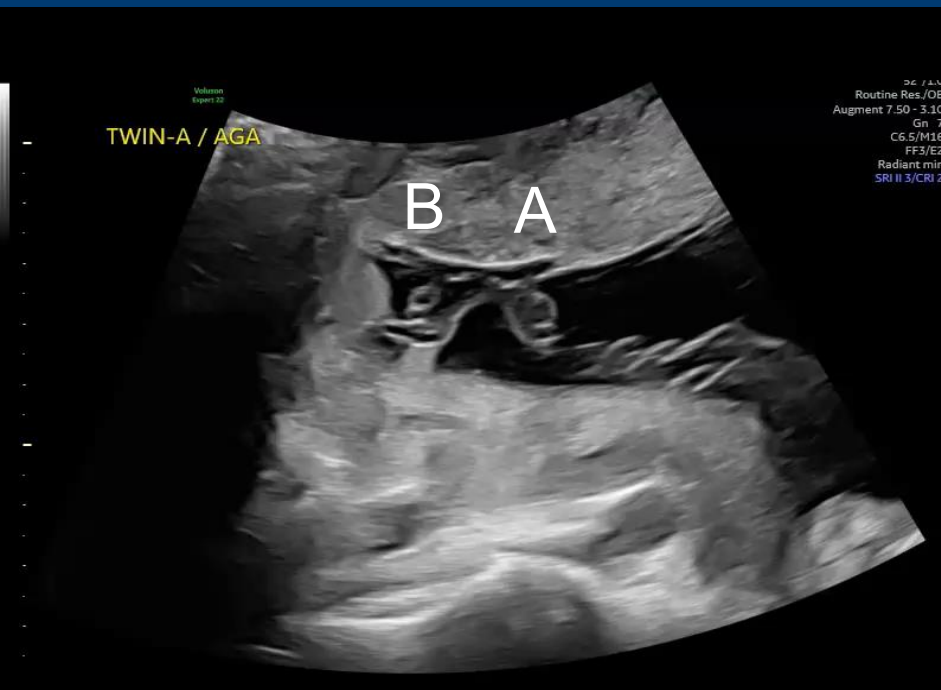
Selective Intrauterine Growth Restriction

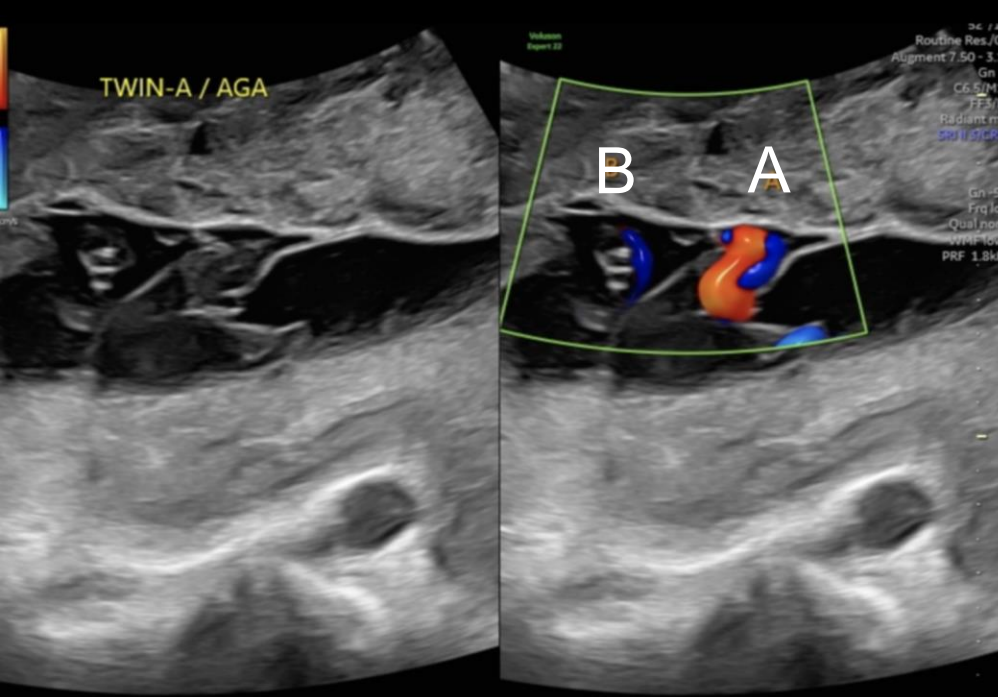
Surgical Management:

- Radiofrequency Ablation or Cord Coagulation
 - ? Improved outcomes of surviving fetus due to controlled death of smaller twin
 - 80-90% survival in remaining fetus (CFCC Experience)
- Laser treatment to functionally “dichorionize” the placenta
 - 90% survival in larger fetus
 - Up to 70% death of smaller twin
 - More invasive therapy

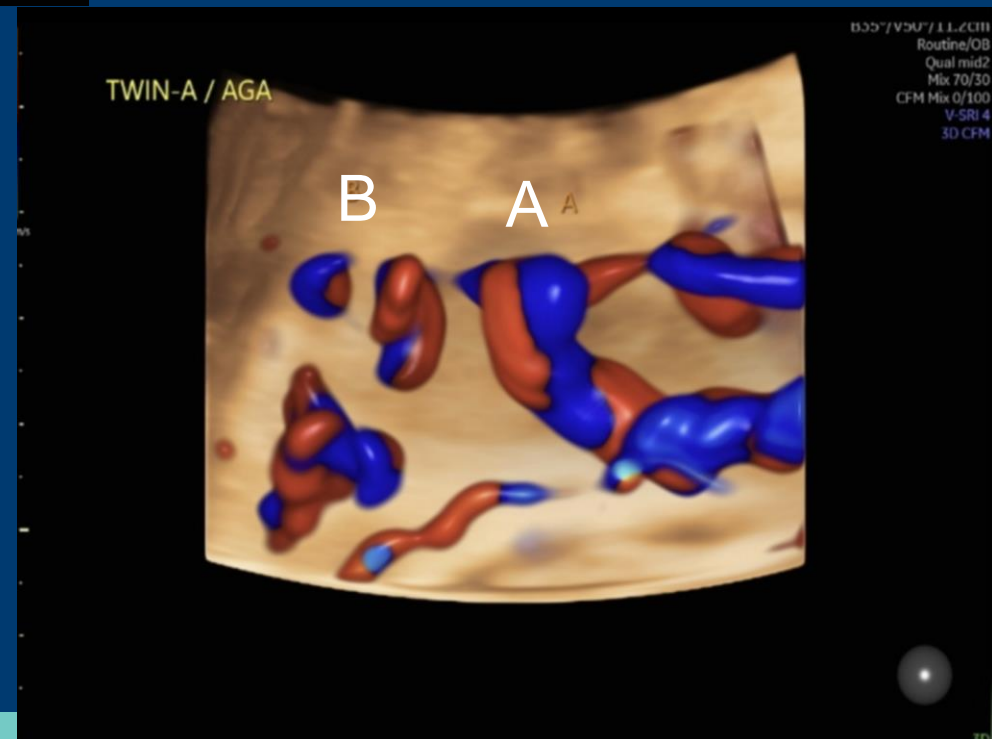
Close Proximity Placental Cord Insertions

- AKA "Double Barrel Cord Insertion"
- Variable definition but in general:
 - Both placental cord insertions are within 2 cm of each
- May impact ability to perform laser procedure for TTTS:
 - No vascular equator able to be established
 - Challenging to treat the direct connections between the cords





3D Doppler shows
the close proximity
cord insertions



Summary

- TTTS Staging criteria should include evaluation of fetal cardiac function
 - UA and MCA Dopplers recommended to be added
- sIUGR staging should dictate care due to significant morbidity/mortality
- TAPS can be independent from TTTS and is associated with morbidity/mortality
- TAPS treatment shows potential improved outcomes but still being investigated
- INDIVIDUALIZED EVALUATION AND TREATMENT PLAN IS THE KEY

Thank You

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Recent Recommendation to Evaluate Umbilical Artery Dopplers in all Monochorionic Twins

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Royal College of
Obstetricians &
Gynaecologists



ACOG

The American College of
Obstetricians and Gynecologists



Society for
Maternal-Fetal
Medicine



- All advocate for Umbilical Artery Doppler and MCA Doppler monitoring
- Some qualify starting at 20 weeks while others start at 16 weeks
- Lack of clear evidence for monitoring of UAs
 - Based on small studies and expert opinion

Why Umbilical Artery Dopplers?

Table 4. The ultrasound findings before the diagnosis of TTTS in stage I–II and III–IV. Data are expressed as median (range) and proportion (%)

Scan in study center ≤14 days before TTTS diagnosis N = 55	Diagnosis in stage I–II N = 27	Diagnosis stage III–IV N = 23	p value
GA at first diagnosis of TTTS	19+5 (16+4–30+4) wks	17+0 (13+6–28+3) wks	<0.01
Interval between scan and TTTS diagnosis	3 (1–14) days	4 (1–14) days	0.25
Abnormal Dopplers on pre-TTTS scan [†]	1/27 (4%)	16/19 (84%) [‡]	<0.0001
Discordant growth on pre-TTTS scan [‡]	8/27 (30%)	12/23 (52%)	0.10
Discordant amniotic fluid on pre-TTTS scan	26/27 (96%)	22/23 (96%)	1.00

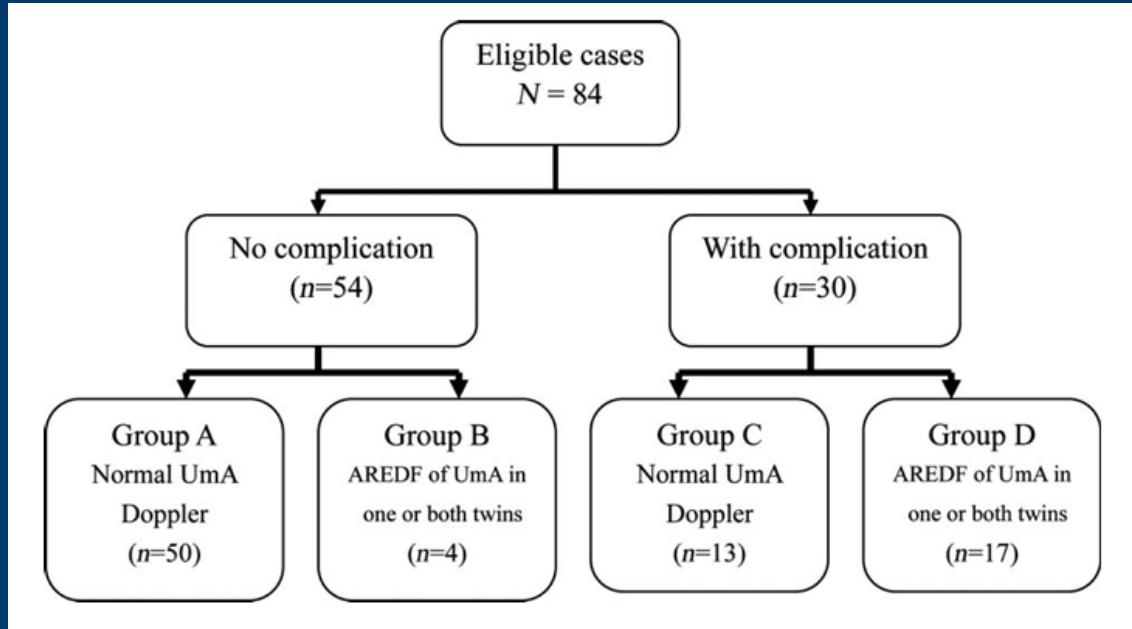
- Abnormal Doppler: Absent or reversed end diastolic flow in the umbilical artery or absent or reversed a-wave in the ductus venosus of one or both twins
 - 6 future donors with abnormal UA
 - 3 both future donor and recipient with abnormal Dopplers
 - 6 future recipients with abnormal DV
 - 1 future donor with abnormal DV

Why Umbilical Artery Dopplers?

- Dopplers can be abnormal preceding the diagnosis of TTTS
- Does not mean earlier stage diagnosis is made
 - Majority of cases were Stage III-IV at time of TTTS diagnosis
- May allow for increased surveillance to avoid Stage V TTTS (fetal demise)

What if we find abnormal UAs?

- 84 Monochorionic-Diamniotic twin pairs (16-20 weeks)
 - 30 with at least one complication (TTTS, sIUGR, discordant growth*)
 - 54 without a complication (AGA Fetuses)



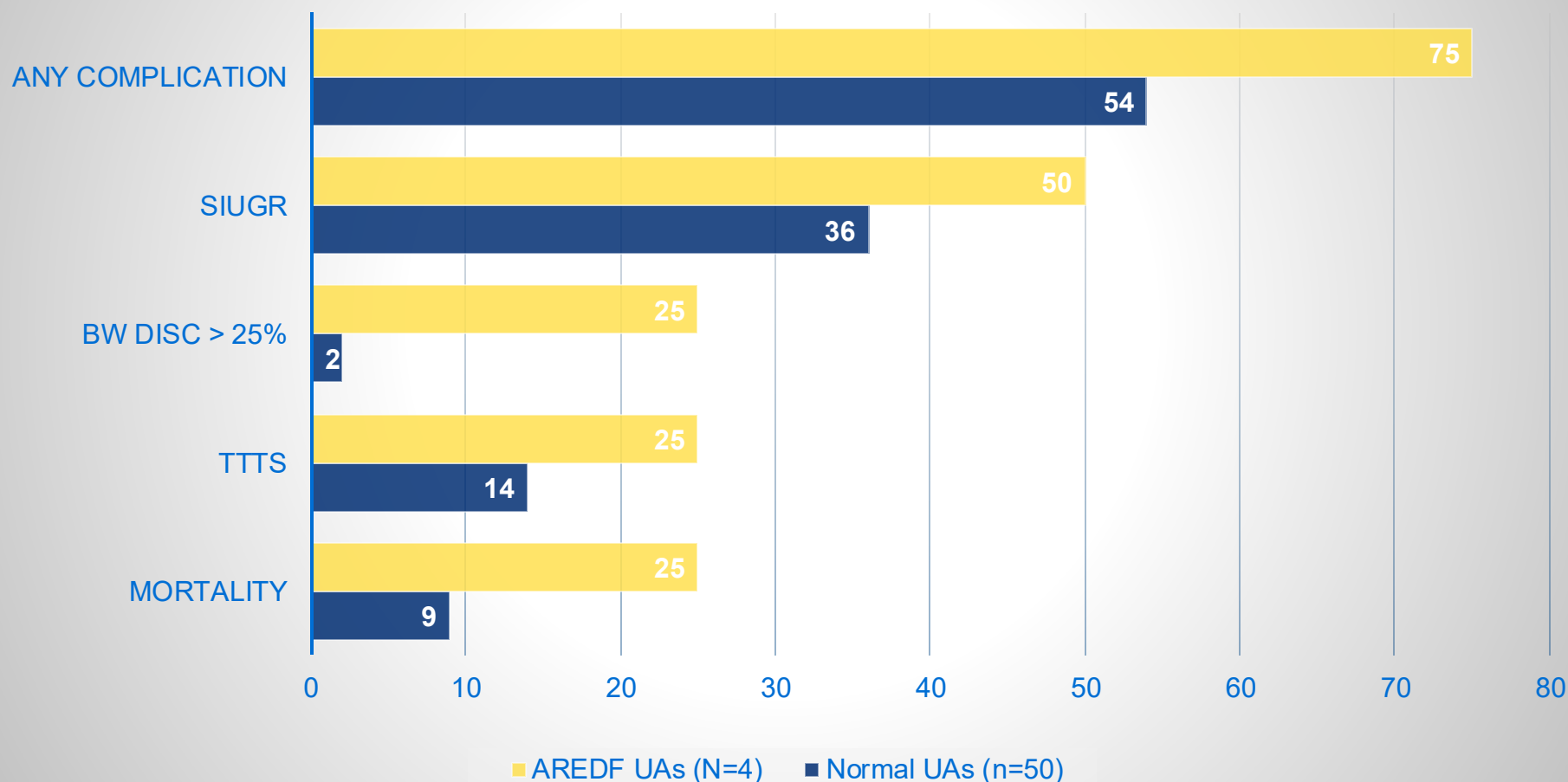
7.4% of no complications had AREDF

56.7% of complications had AREDF

*25% EFW discordance

Min Pan et al. (2012) Outcome of monochorionic twin pregnancies with abnormal umbilical artery Doppler between 16 and 20 weeks of gestation, The Journal of Maternal-Fetal & Neonatal Medicine, 25:3, 277-280

AGA Fetuses Normal vs. AEDF Umbilical Artery Dopplers (%)



Min Pan et al. (2012) Outcome of monochorionic twin pregnancies with abnormal umbilical artery Doppler between 16 and 20 weeks of gestation, The Journal of Maternal-Fetal & Neonatal Medicine, 25:3, 277-280

Incidence of Abnormal Umbilical Artery Dopplers in Monochorionic-Diamniotic Twins

<i>Parameter</i>	<i>Group 1: controls</i>	<i>Group 2: sIUGR</i>	<i>Group 3: TTS</i>
Gestational age at diagnosis (weeks)	32.2 [29–36]	24.7 [20–28]	26
Incidence of intermittent Doppler flow	5 (4/80)	45 (18/40)*	2 (1/50)
iAEDF alone	75 (3/4)	38.9 (7/18)	100 (1/1)
iREDF alone	0	16.7 (3/18)	0
Both iAEDF and iREDF	25 (1/4)	44.4 (8/18)	0
Persistence until delivery	75 (3/4)	88.9 (16/18)	100 (1/1)

5% incidence of abnormal UAs in AGA fetuses

Gratacos et al. Incidence and characteristics of umbilical artery intermittent absent and/or reversed end-diastolic flow in complicated and uncomplicated monochorionic twin pregnancies. *Ultrasound Obstet Gynecol* 2004; 23: 456–460

OR

Voluson
E8

Routine
Har-

Gn

C5 /

FF4 /

SRI II 5 / CF

