Complications of Monochorionic Twins

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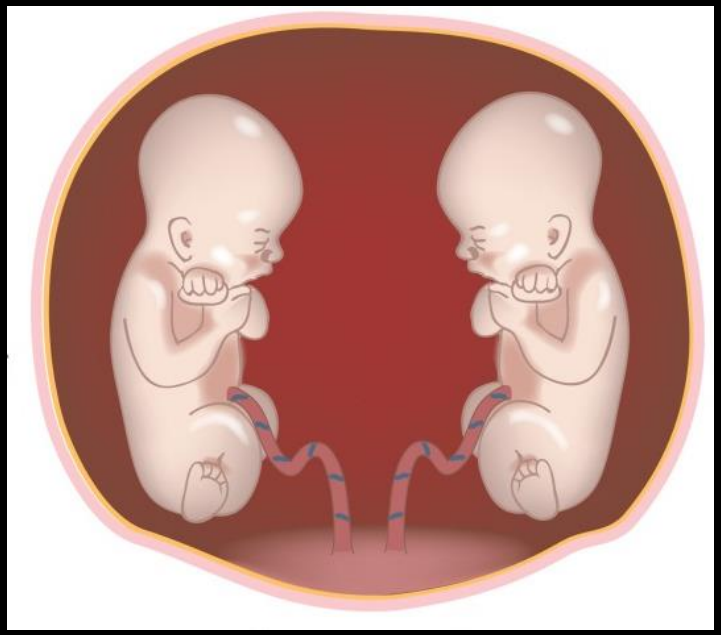
Potential Complications of Monochorionic Twins

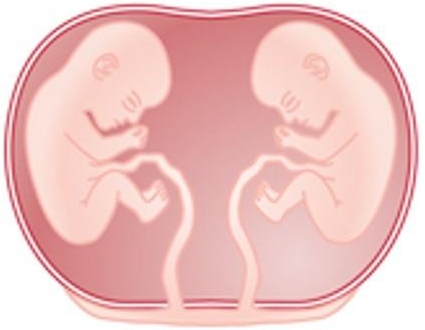
- Monoamniotic twins
- Conjoined twins
- Twin reversed arterial perfusion (TRAP) sequence
- Twin-twin transfusion syndrome (TTTS)
- Unequal placental sharing (UPS)
  - Discordant twin growth
  - Selective intrauterine growth restriction (sIUGR)
- Twin anemia-polycythemia sequence (TAPS)
- Single twin demise in the second or third trimester

Monoamniotic Twins

- 1% of all monozygotic twins
- Results from cleavage at 8-13 days

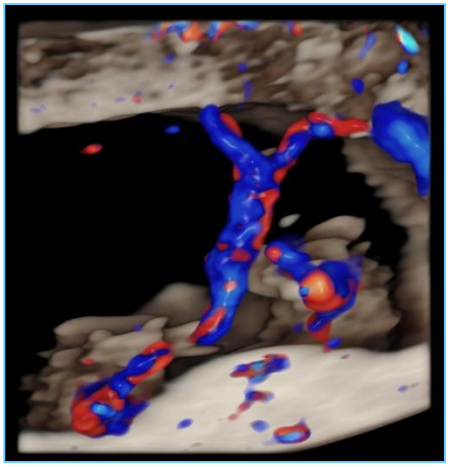


Monoamniotic Twins

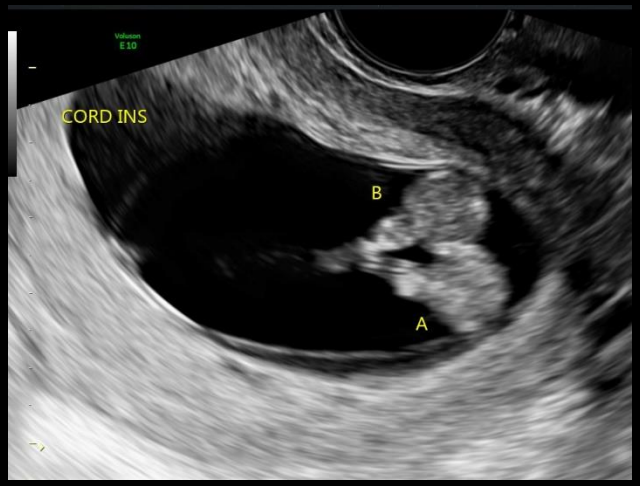


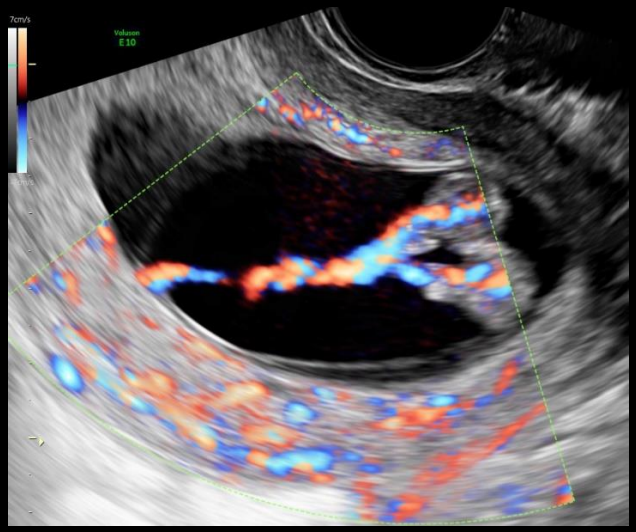
How do you make the diagnosis?

- Lack of separating membrane on serial exams
- Cord entanglement
  - Utilize color Doppler
  - Present in >80% of cases
- Single placenta with two cord insertions
  - Often in very close proximity
- Associated congenital anomalies
  - Present in 10% of cases



Monoamniotic Twins

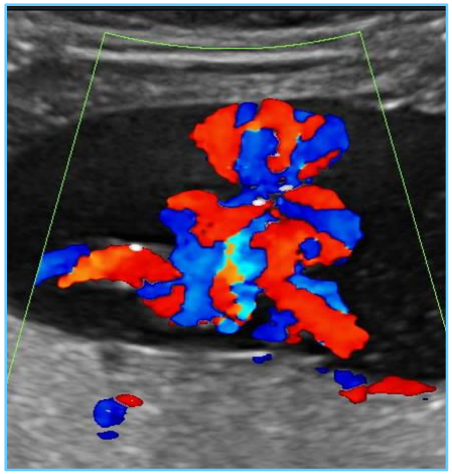




- Forked cord
- Abnormal vessel number

Monoamniotic Twins

What are your management recommendations?

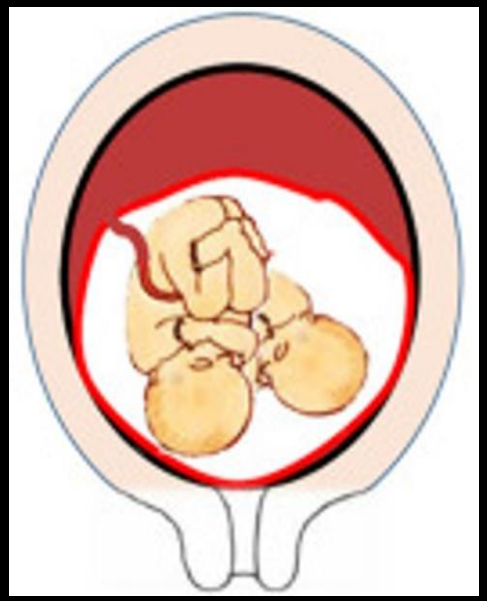


- Monthly growth scans
- Hospital admission at 24-28 weeks
- Serial surveillance
  - BPP
  - NST / continuous EFM
  - Doppler
- CD at 32-34 weeks

Contemporary management has increased survival from 50-60% to over 90%

Conjoined Twins

- Rare event
- Results from cleavage at 13-15 days



Cephalopagus

Thoracopagus

Conjoined Twins

How do you make the diagnosis?

- Monoamniotic placentation
- Same relative positions of twins to each other in all views
- Direct opposition of the twins
- Extreme extension of the fetal spines
- Shared organs, vascular connections, associated anomalies

Management recommendations?

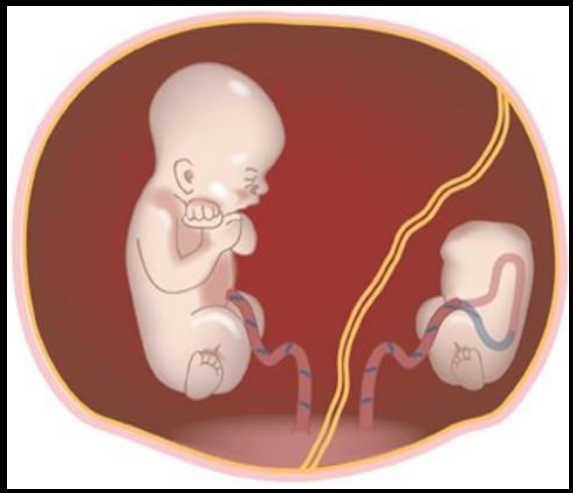
Dicephalic Parapagus

Thoracopagus

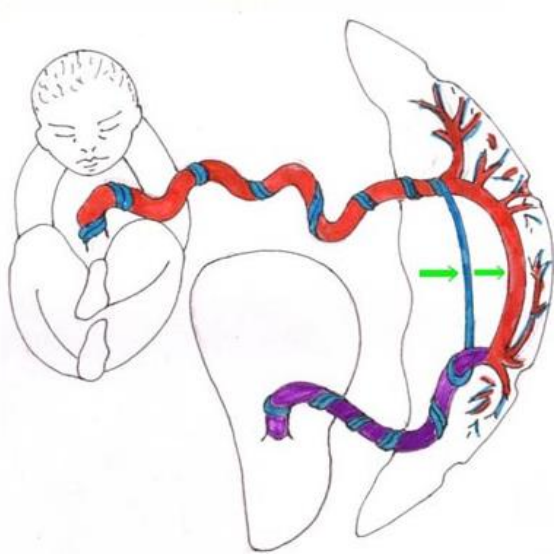
TRAP Sequence

What is twin reversed arterial perfusion sequence?

- Complication of monochorionic twins
  - Prevalence: 1:100 monochorionic twins
  - 75% diamniotic, 25% monoamniotic
- Vascular disruption during early embryogenesis
  - Results in aberrant arterioarterial anastomoses between twins
  - Acardiac twin lacks direct placental perfusion



TRAP Sequence



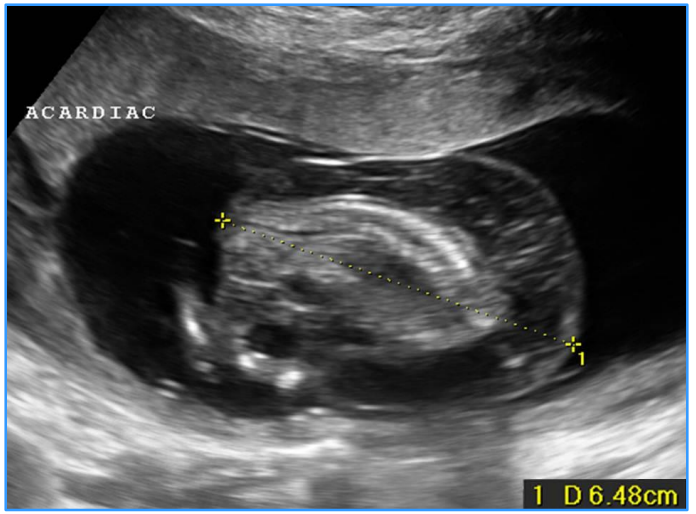
Acardiac twin is dependent on retrograde arterial supply of relatively deoxygenated blood from its co-pump twin

- Asymmetric circulation leads to abnormal development of acardiac twin, favors growth of caudal aspects
- Differential diagnosis
  - Demise of one twin
  - Discordant twin growth
  - Cystic hygroma
  - Acrania / anencephaly
  - Placental teratoma

TRAP Sequence

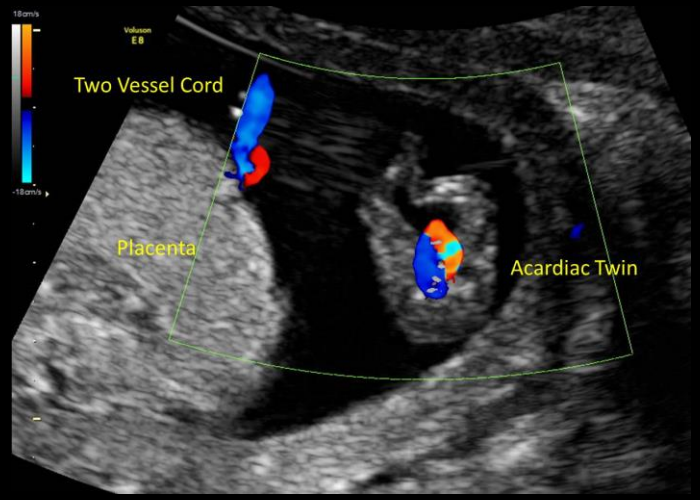
How it is diagnosed?

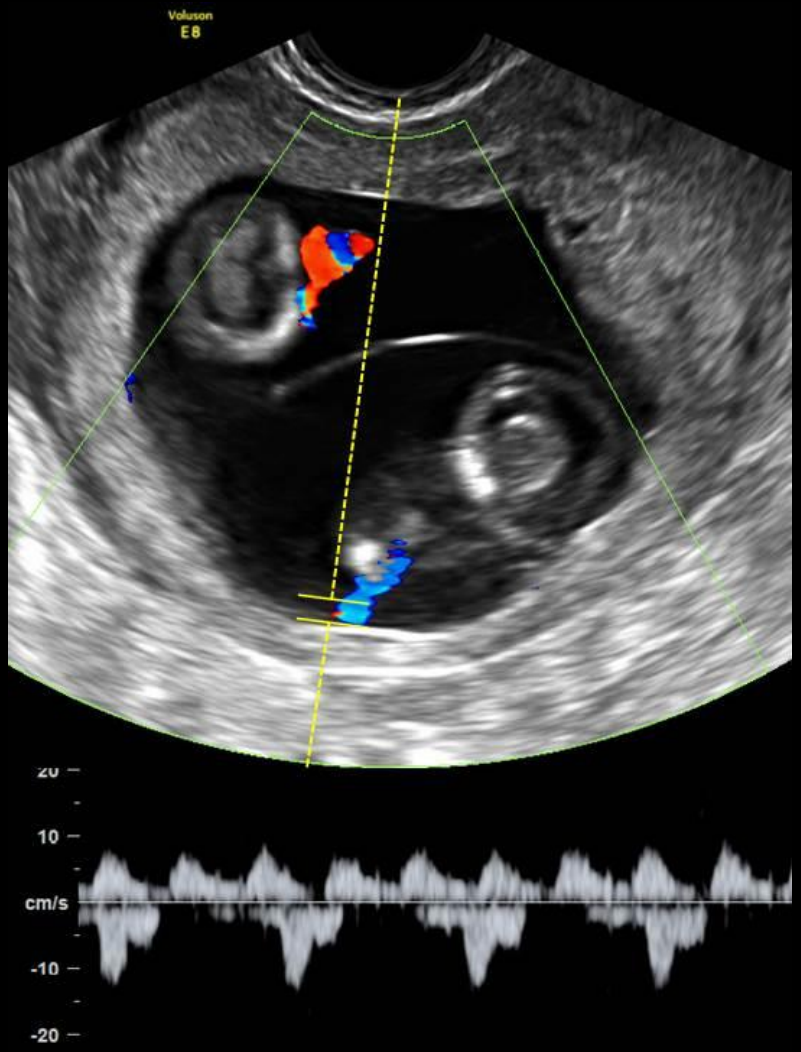
- Abnormal early development of one twin of monochorionic pair
- Acardius acephalus most common
  - Well-formed fetal pelvis and lower limbs
  - Absent head/skull and usually without thoracic organs and arms



TRAP Sequence

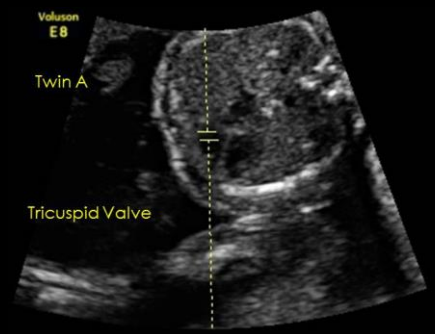
- Acardiac twin lacks a normal heart but may have a rudimentary structure with pulsations
- Paradoxical arterial flow towards the acardiac twin from pump twin

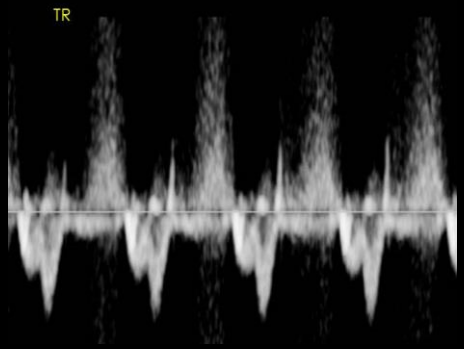


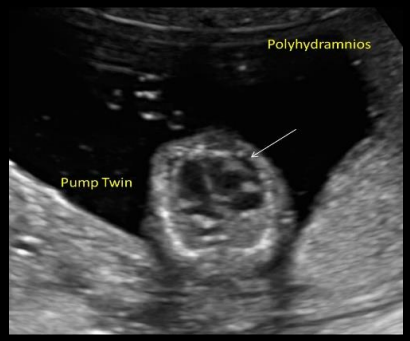


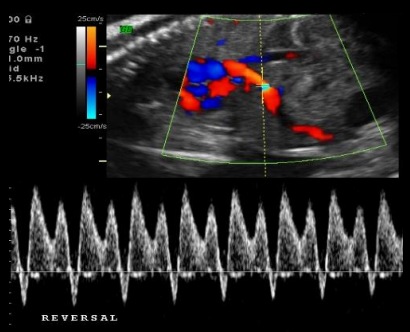
TRAP Sequence

What do you recommend?









Pump twin is at risk for:

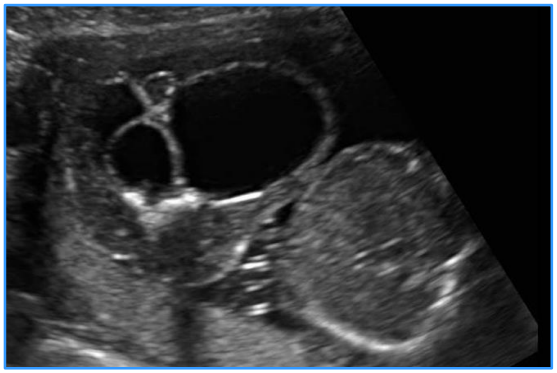
- anomalies (5-10%)
- aneuploidy (10%)
- two vessel cord (65%)
- hemodynamic compromise (30%)
  - cardiac dysfunction, cardiomegaly, hydrops

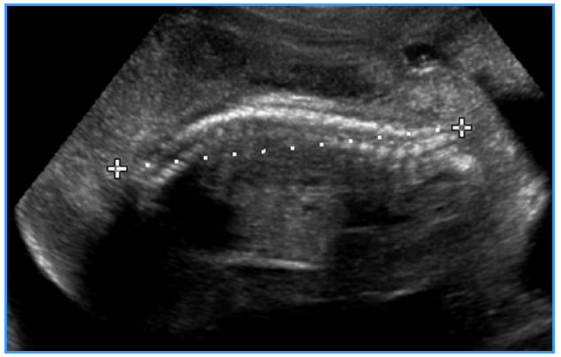
Simpson, Decision Support in Medicine, 2014

TRAP Sequence

Size matters!

- Ratio of acardiac/pump twin >05-0.7 increases risk for:
  - cardiac failure (30%)
  - polyhydramnios (50%)
  - preterm delivery (90%)





Estimate size of acardiac twin:

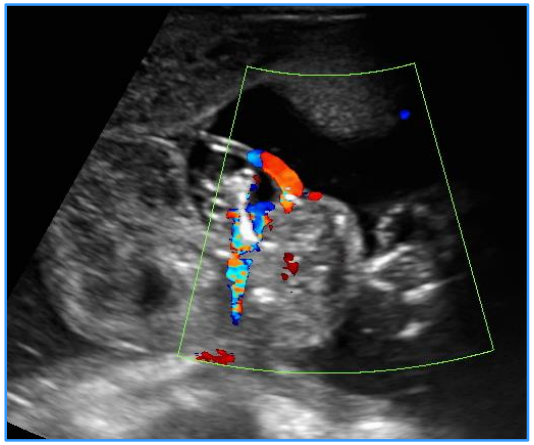
- EFW (g) = length x width x height x 0.52
- EFW (g) = (-1.66 x length) + (1.21 x length^2)

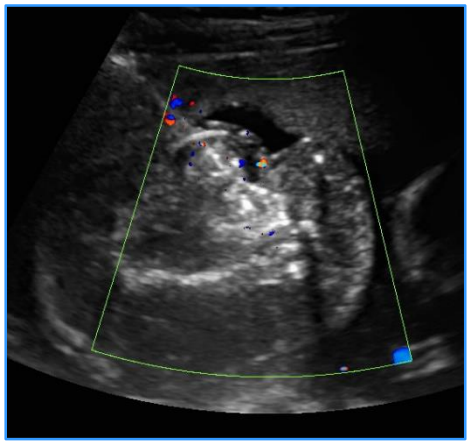
Lee et al (NAFTNet), 2013;
Jellin et al, 2010;
Oliver et al, 2013

TRAP Sequence

Consider invasive cord occlusion therapy when . . .

- High acardiac-to-pump ratio (≥50%)
- Rapid growth of acardiac twin
- Hemodynamic compromise of pump twin

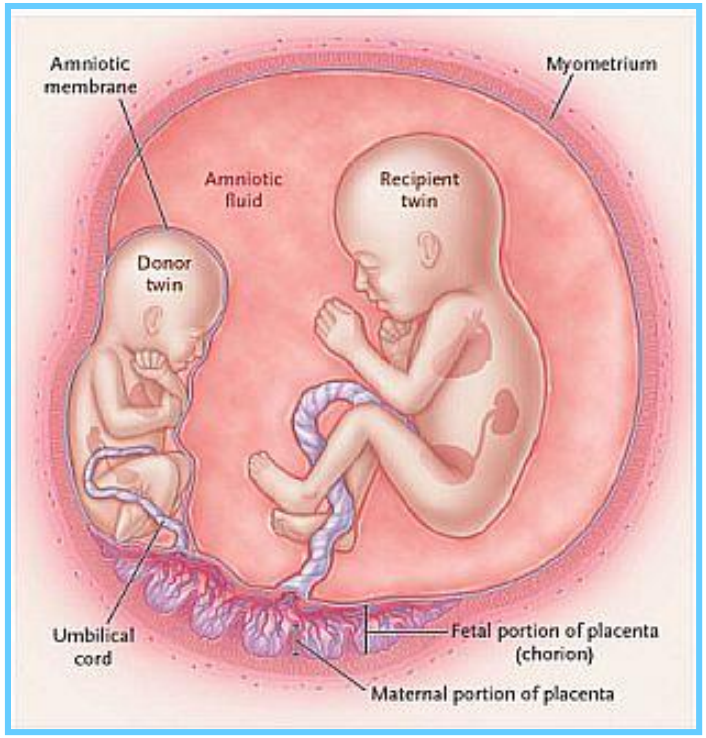




Lee et al (NAFTNet), 2013
Aitken et al, 2014

- Variety of techniques available
  - bipolar coagulation, radiofrequency ablation (RFA) most popular
  - RFA targets intrafetal cord insertion within acardiac twin
  - survival 80-90%, mean GA at delivery 34-36 weeks

Twin-Twin Transfusion Syndrome

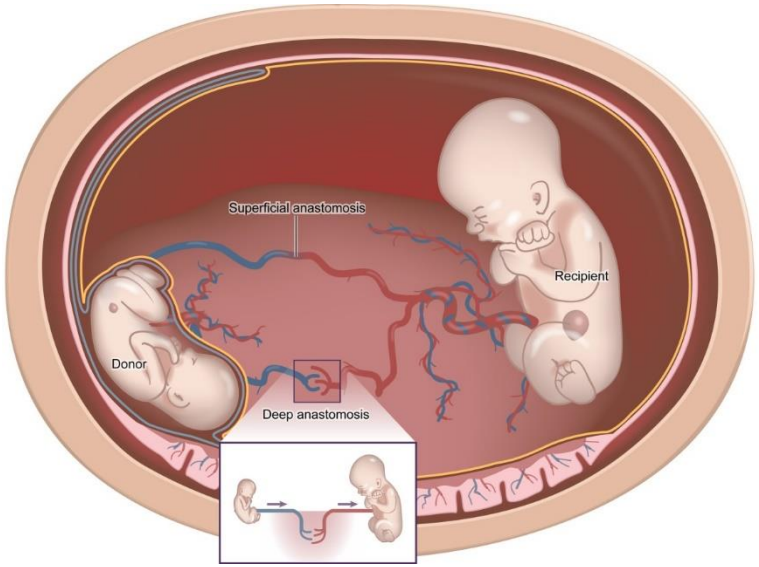


- Complicates 8-10% of monochorionic diamniotic twin gestations
- Untreated TTTS developing before the third trimester has a perinatal mortality rate of >70%
  - 15-50% risk of handicap in survivors

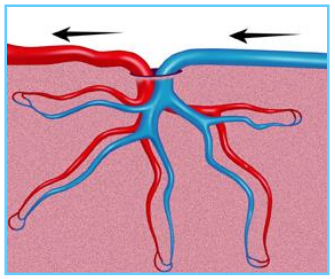
Simpson et al. ACOG 2011:204:145

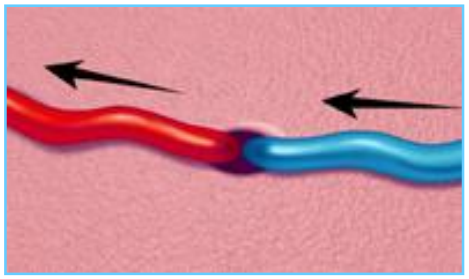
Twin-Twin Transfusion Syndrome

What it is?



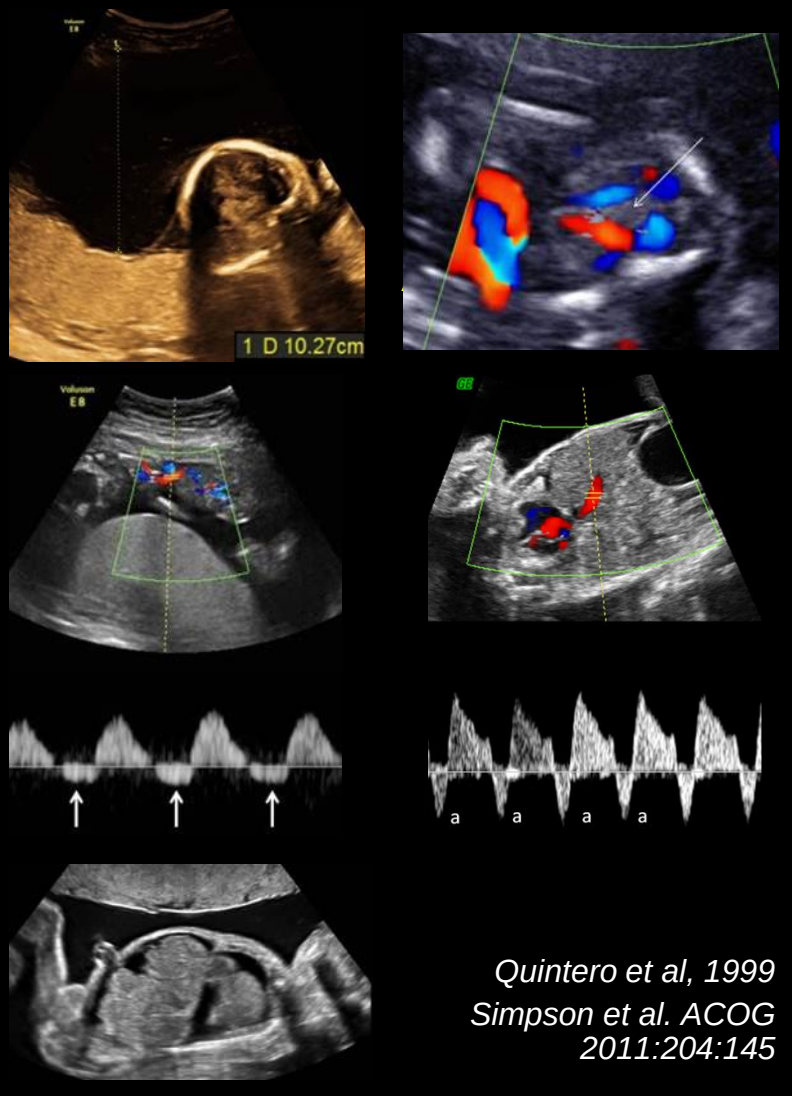
- Intertwin transfusion
  - Unequal sharing of blood
  - Changes in regional blood flow
  - Alterations in cardiac function
- Due to presence of vascular anastomoses in single placenta
  - 80-100% have intertwin anastomoses
  - Superficial bidirectional AA and VV
  - Deep unidirectional AV





Twin-Twin Transfusion Syndrome

Stage	Ultrasound Assessment	Criteria
I	Amniotic fluid	MVP <2 cm in donor sac; MVP >8 cm in recipient sac
II	Fetal bladder	Nonvisualization of fetal bladder in donor twin over 60 minutes of observation
III	Doppler studies	Absent or reversed umbilical artery diastolic flow, reversed ductus venosus a-wave flow, pulsatile umbilical vein flow
IV	Fetal hydrops	Hydrops in one or both twins
V	Fetal cardiac activity	Fetal demise in one or both twins

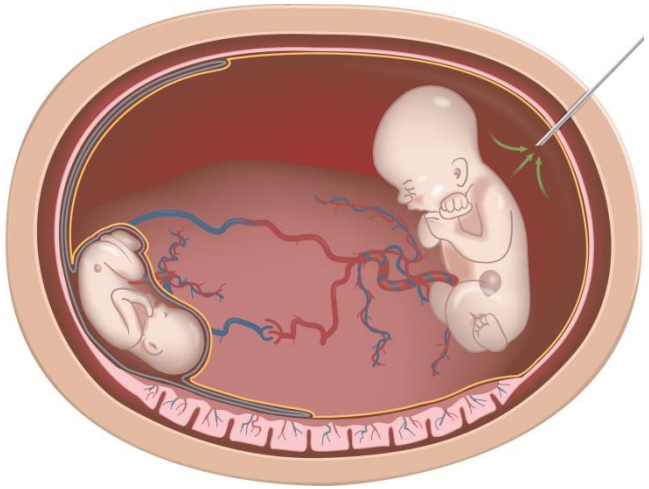


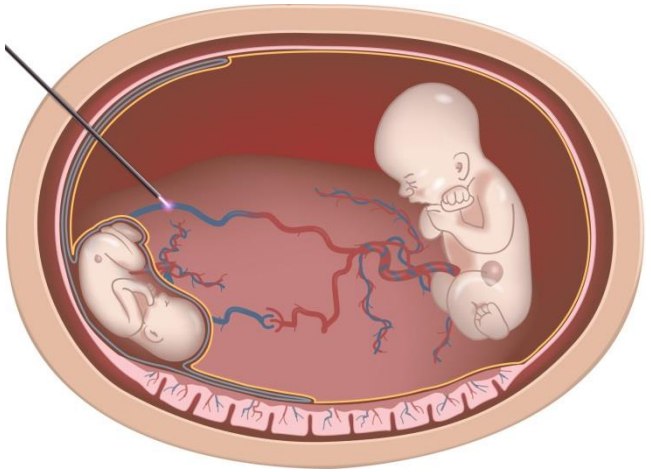
Quintero et al, 1999
Simpson et al. ACOG 2011:204:145

Twin-Twin Transfusion Syndrome

What do you recommend?

- Pregnancy termination
  - Early, advanced stage TTTS
- Amnioreduction
  - Beyond 26 weeks
  - Declines fetoscopic laser therapy
  - Fetoscopic laser therapy unavailable
- Laser photocoagulation of communicating vessels
  - 18-26 weeks
  - Advanced stage TTTS
- Delivery
  - Late presentation





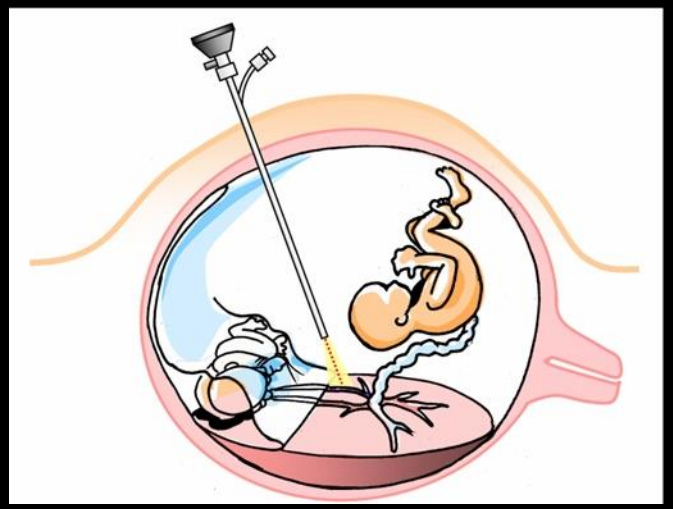
TTTS Outcomes	Stage I	Stage II	Stage III	Stage IV	Normal Neurologic Outcome at 6 months and 6 years
Resolution	27%	10%	1%	–	
Amnioreduction					31% 70%
At least one survivor	90%	80%	48%	26%	
Two survivors	50%	48%	20%	6%	
Laser					52% 82%
At least one survivor	86%	84%	78%	62%	
Two survivors	92%	82%	66%	52%	

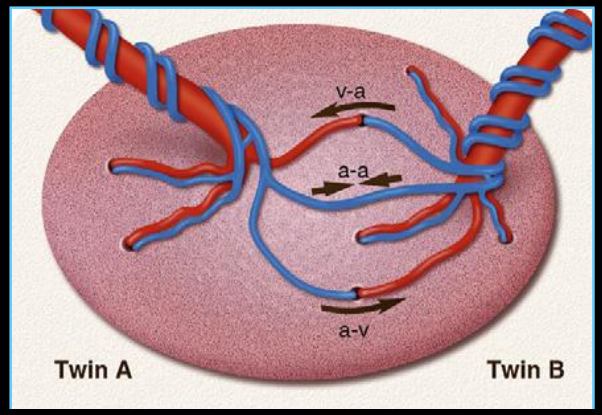
Mari et al, 2001; Hecher et al, 2000; Quintero et al, 2003; Senat et al, 2005; Galen et al, 2005; Salomon et al, 2010

- In over 1000 published cases of laser performed for TTTS from six different centers, there were:
  - Two survivors: 50%
  - Single survivor: 30%
  - No survivors: 20%
- Normal neurologic development at 2 years of age: 80-90%

Simpson et al. ACOG 2011:204:145

Fetoscopic Laser Therapy





Unequal Placental Sharing

What it is?

Pathologic discordance in territorial share of the common placenta in monochorionic twins

- Discordant twin growth
  - 20-25% discordance significant
- Selective IUGR
  - EFW ≤10th percentile of one twin

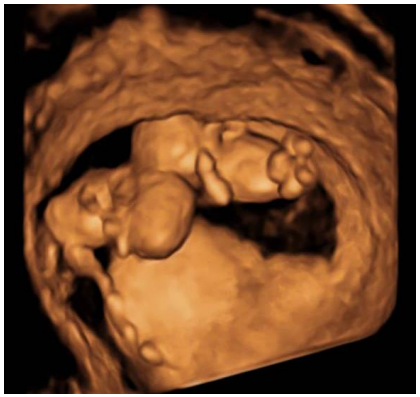
Unequal Placental Sharing

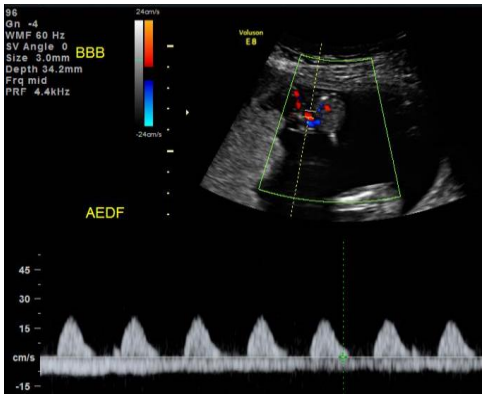
When should you be suspicious?

Disparate CRL's and AC's early sonographic signs of subsequent discordant growth and/or sIUGR in MC twins

Unequal Placental Sharing

- Development of sIUGR in monochorionic twins may be function of
  - Unequal placental sharing / placental insufficiency
  - Intertwin anastomoses and intertwin flow patterns
- Abnormal umbilical artery waveforms of sIUGR twin may represent effects of
  - Placental resistance
  - Type and size of intertwin anastomoses
- Leads to substantial clinical differences in apparently similar cases





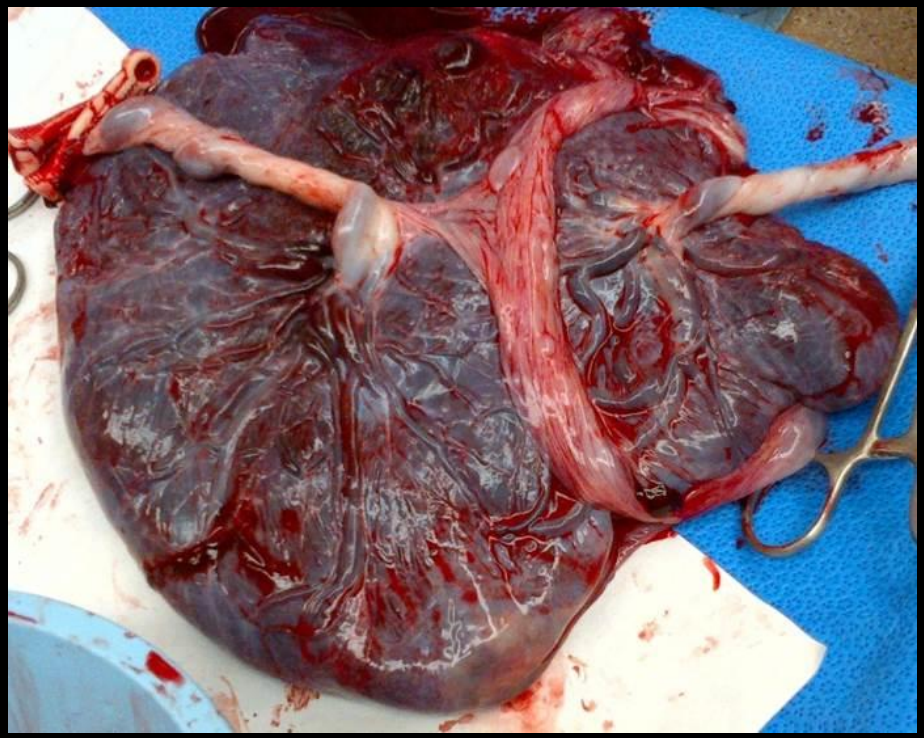
Unequal Placental Sharing

How it is diagnosed?

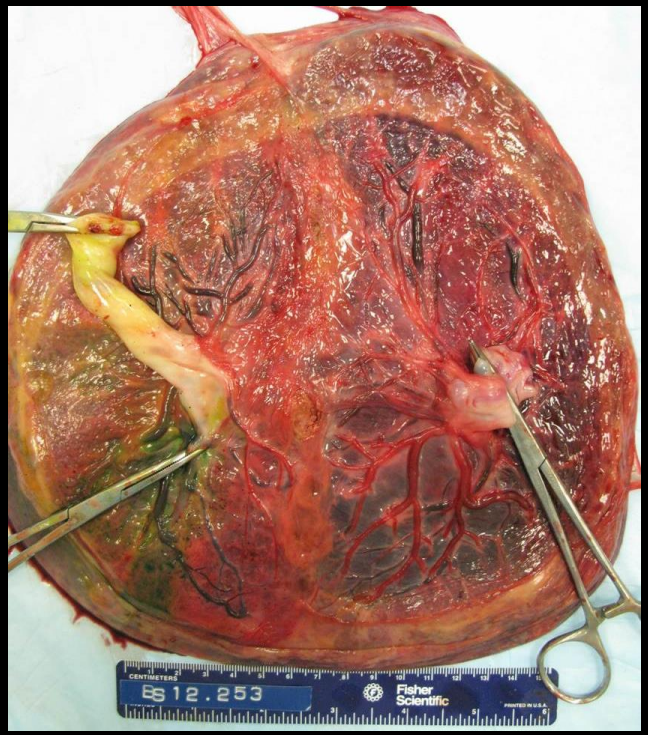
Feature	UPS	TTTS
Growth discordance ≥20%	++++	++
EFW ≤10th %ile	++++	++
Cardiac dysfunction	Smaller sIUGR twin	Larger recipient twin
Amniotic fluid discrepancy	Oligohydramnios in IUGR twin Normal fluid in AGA twin	Oligohydramnios in one sac (MVP ≤2 cm) Polyhydramnios in other sac (MVP ≥8 cm)

- Must have oligohydramnios-polyhydramnios sequence for diagnosis of TTTS
- Must have growth discordance +/- sIUGR for diagnosis of UPS

UPS versus TTTS



UPS with sIUGR



TTTS laser case

UPS with sIUGR Staging

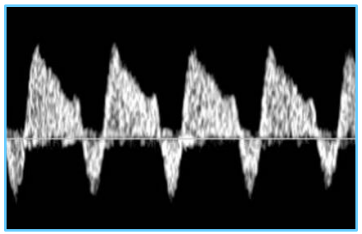
Type	UA Dopplers	Placenta	Intertwin Flow Via Anastomoses	IUFD Risk
I	Positive DF	Small placental territory, many anastomoses	Compensates for small placental share	2-4% (unpredictable)
II	Persistent AREDF	Smaller placental territory, many anastomoses	Attenuates severity of sIUGR	0-30% (predictable) Decrease risk with serial surveillance/ early delivery
III	Intermittent AREDF	Tiny placental territory, close PCIs, large AAAs	Enables survival of sIUGR twin but potential for acute, massive transfusion	10-20% (unpredictable) Unstable hemodynamics due to large AAAs

Gratacos et al, 2007
Valsky et al, 2010

UPS with sIUGR

What are your management considerations?

- Spontaneous demise of MCDA twin carries 10% risk of death of co-twin, 20% risk of neurologic injury
  - Acute anemia due to massive blood transfer from surviving twin into dead twin/placenta
- Presence of placental anastomoses may be protective for sIUGR twin – compensatory flow from its co-twin
- Management strategy remains a challenge!
  - Influenced by severity of sIUGR, gestational age, parental decisions, technical issues



UPS with sIUGR

Role for cord occlusion?

- Option in select cases to prevent death/injury of co-twin
  - Early detection of type II or III
  - High risk of spontaneous demise
    - *Cardiac dysfunction*
    - *Abnormal venous Dopplers*
    - *Associated anomalies*
- Prognosis related to GA at reduction – lower survival rate if performed <18 weeks
- Survival >80%, normal neurologic outcome in >90%

Role for laser?

- Higher obstetric risks
- Technical challenges
  - Lack of polyhydramnios
  - PCIs in close proximity (type III)
  - Large, diameter AAAs (type III)
- Substantial risk for demise of sIUGR twin anyway (65-75%)
- May be reasonable when selective feticide not an option
  - Gestational age, availability, parental decision

Parra-Cordero et al, 2015; Chalouhi et al, 2013; Valsky et al, 2010; Gratacos et al, 2008; Quintero et al, 2001

UPS with Type III sIUGR

	No Laser N=31	Laser N=18	P
GA diagnosis	22.0 wk	22.0 wk	0.21
GA delivery	31.0 wk	32.6 wk	0.32
Discordance	33%	38%	0.22
IUFD overall	14.5%	36.1%	0.02
IUFD smaller twin	19.4%	66.7%	0.001
Both alive	80.6%	27.8%	<0.001

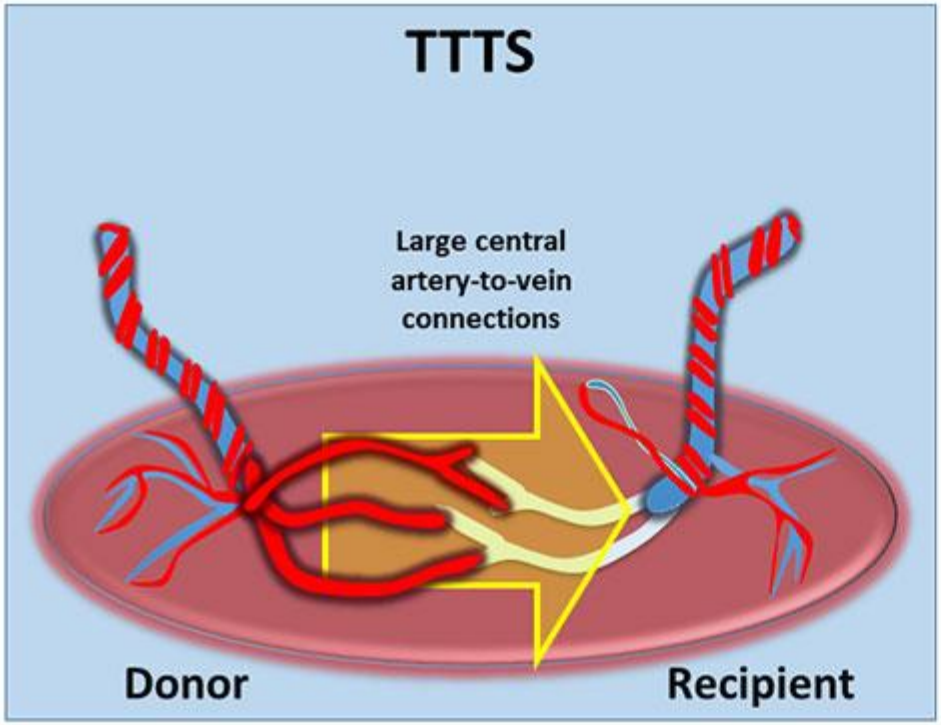
Gratacos et al, 2008

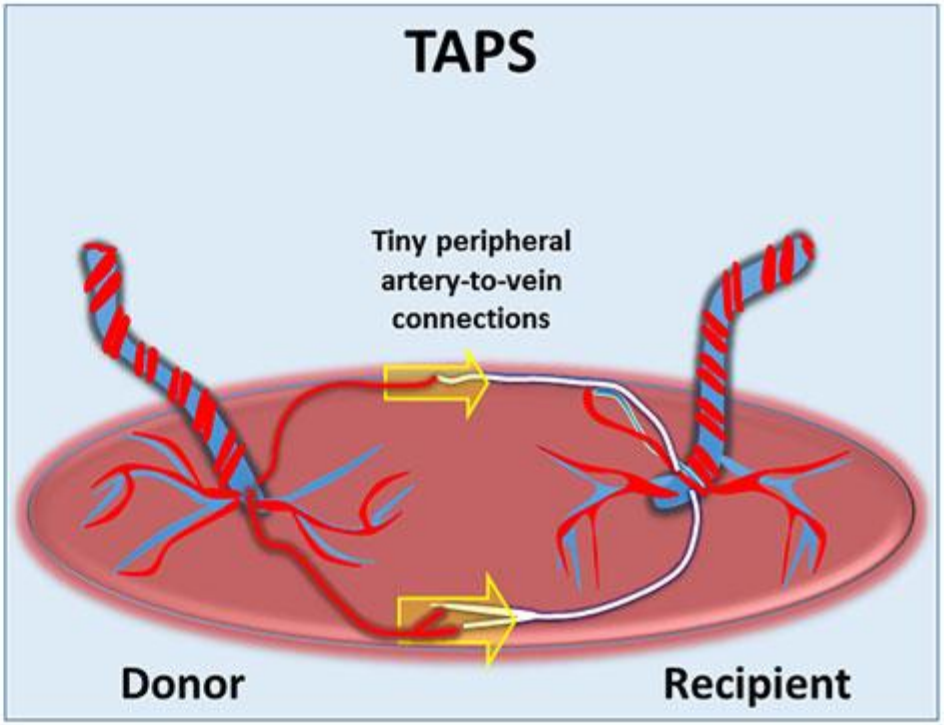
Laser therapy for sIUGR needs further investigation

TAPS

What it is?

- Twin anemia polycythemia sequence
- Chronic form of fetofetal transfusion



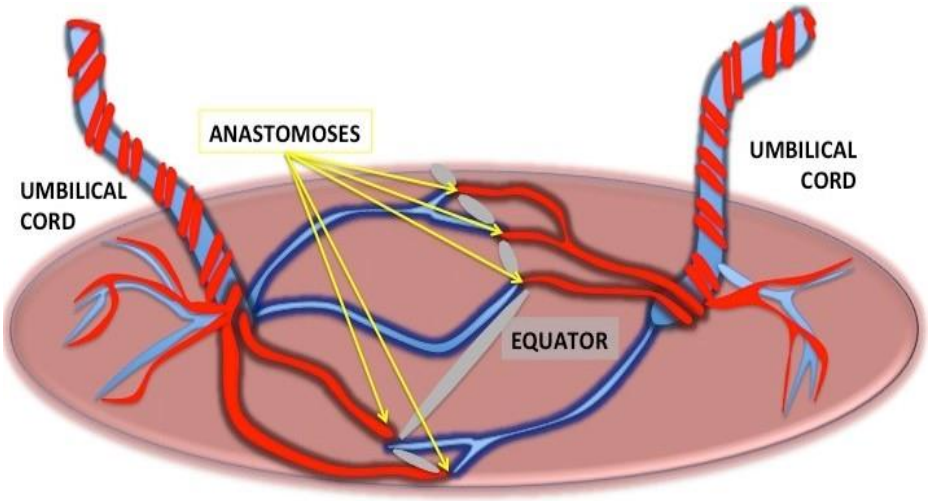


TAPS

Spontaneous (3-5% of MC twins)

Iatrogenic (10-15% post-laser)

- Residual anastomoses in 5-30% of laser cases
- Solomon technique reduces risk

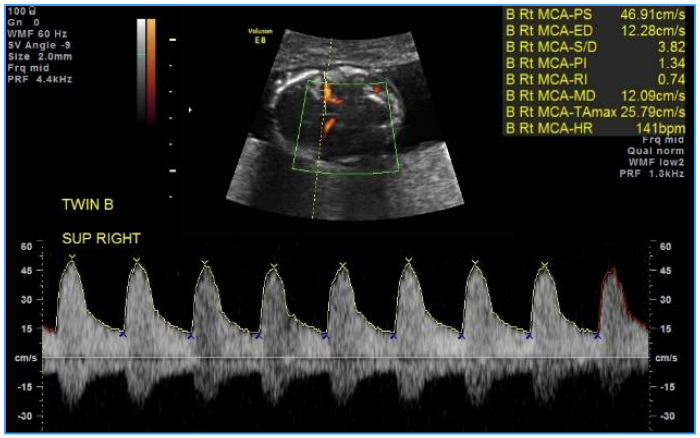


Slaghekke et al, 2010, 2014
Lopriore et al, 2009, 2010

TAPS

How is it diagnosed?

- Elevated PSV-MCA in one twin = anemia
- Decreased PSV-MCA in co-twin = polycythemia



Stage	Criteria
1	PSV-MCA >1.5 MoM in donor PSV-MCA <1.0 MoM in recipient
2	PSV-MCA >1.7 MoM in donor PSV-MCA <0.8 MoM in recipient
3	Stage 1 or 2 with cardiac compromise of donor
4	Hydrops of donor
5	Single or double IUFD

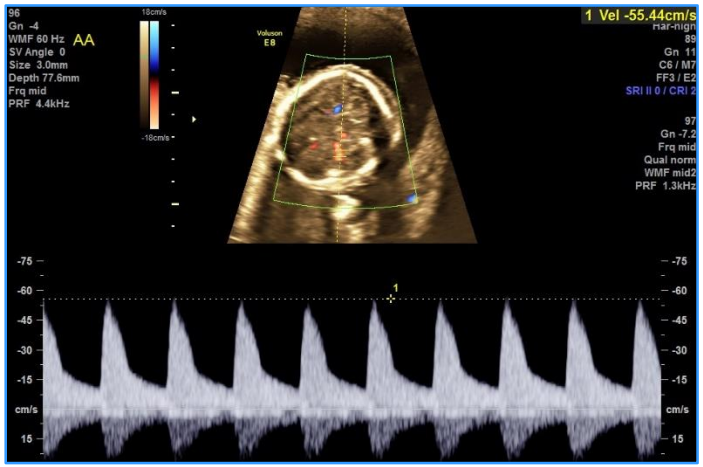
Slaghekke et al, 2010, 2014
Lopriore et al, 2009, 2010

TAPS

What are your management recommendations?

- *Depends on gestational age, technical considerations, disease severity*

Options

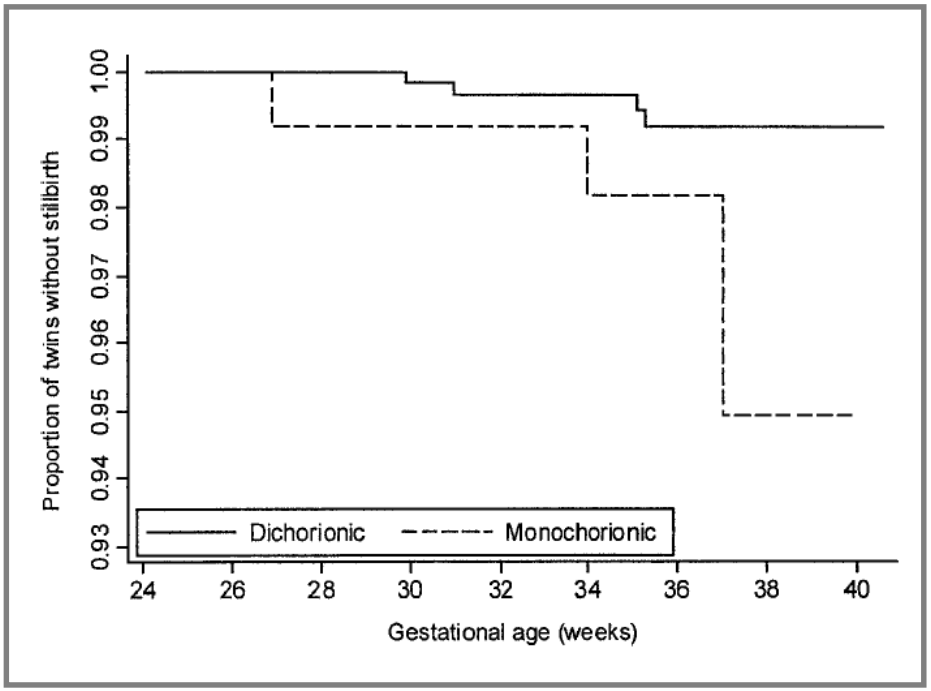


- Repeat laser: technical difficulties
- Fetal transfusion: not curative
- Cord coagulation
- Expectant management
- Early delivery

Single MC Twin Demise

Retrospective cohort analysis of 1000 consecutive twins ≥24 weeks

- 804 DCDA − 1.1% stillbirth
- 198 MCDA − 3.6% stillbirth



Analysis of 151 normal MCDA twins ≥24 weeks

All demises within 2 weeks of a normal scan

Overall risk of late fetal death in uncomplicated MCDA twins:

- 4.6% per pregnancy
- 3.3% per fetus

Lee et al, 2008
Barigye et al, 2005

Single MC Twin Demise: Acquired CNS Injury

Chorionicity	Co-twin death after single demise	Abnormal CNS imaging in surviving co-twin	Neurodevelopmental impairment in surviving co-twin
Dichorionic	3%	16%	2%
Monochorionic	15%	34%	26%

- For single death in MC pair, 5 times more likely to have neurologic morbidity

Griffiths et al, 2015; Hillman et al, 2011

Take Home Message

Never take your eyes off MC twins