Maternal Fetal Medicine and Perinatology Association Türkiye
23-06-1998
IX. National Congress
24 - 27 September 2014 Harbiye Military Museum

Management of Monozygotic twins

Isaac Blickstein, MD

Level	Type of twining	Sharing	Frequency
	Dizygotic	Uterus	
	Dichorionic-monozygotic	Genome	
	Monochorionic-biamniotic	Chorion, circulation	
	Monochorionic-monoamniotic	Amniotic cavity	
	Conjoined	Organs	

Review

THE TWIN-TWIN TRANSFUSION SYNDROME

Isaac Blickstein, MD

Twin-twin transfusion syndrome is a complication of monozygotic-monochorionic twinning with serious perinatal implications. An extensive literature review revealed that

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Table 1. Composite Definition of the Twin-Twin Transfusion Syndrome*

Criteria	Suggested cutoff values
I. Minor	
1) Sonographic	Inter-twin abdominal circumference difference >18 mm, poly/oligohydramnios, signs of monozygosity
2) Doppler velocimetry (umbilical arteries)	Inter-twin difference in S/D ratios >0.4
II. Major	
1) Transplacental shunt	
2) Birth weight	Inter-twin difference ≥15% (heavier twin = 100%)
3) Hemoglobin†	Inter-twin difference ≥5 g/dL

S/D = systolic/diastolic.

* Two major criteria or one minor and one major criterion are needed.

† Criteria may be found by antenatal funipuncture or postnatally.

the twin-twin transfusion syndrome. Wittmann et al

It’s all about

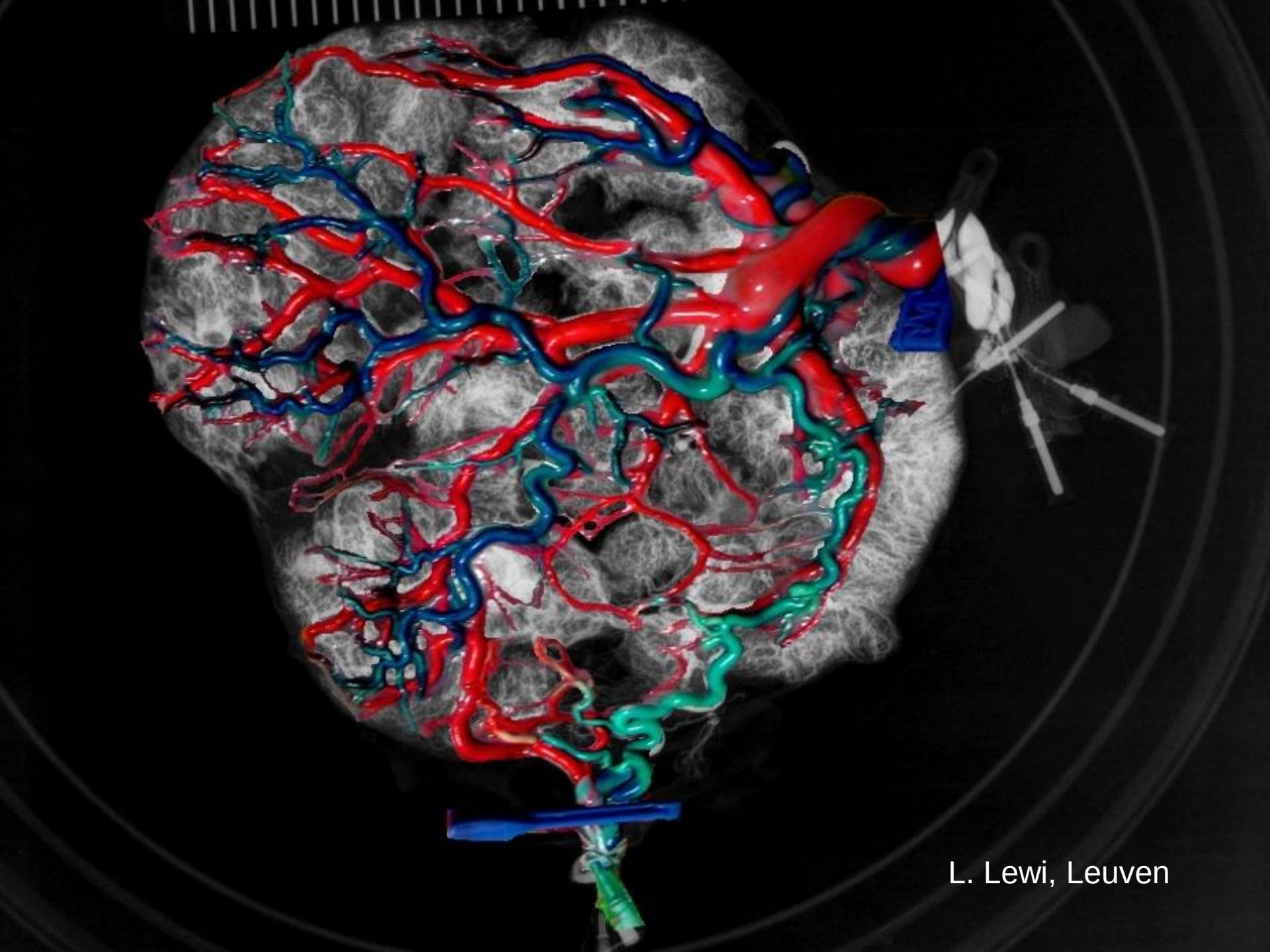
Territory

It’s all about

Territory

Pipelines


PEINTRES DE LA VIE MODERNE
MAGELLAN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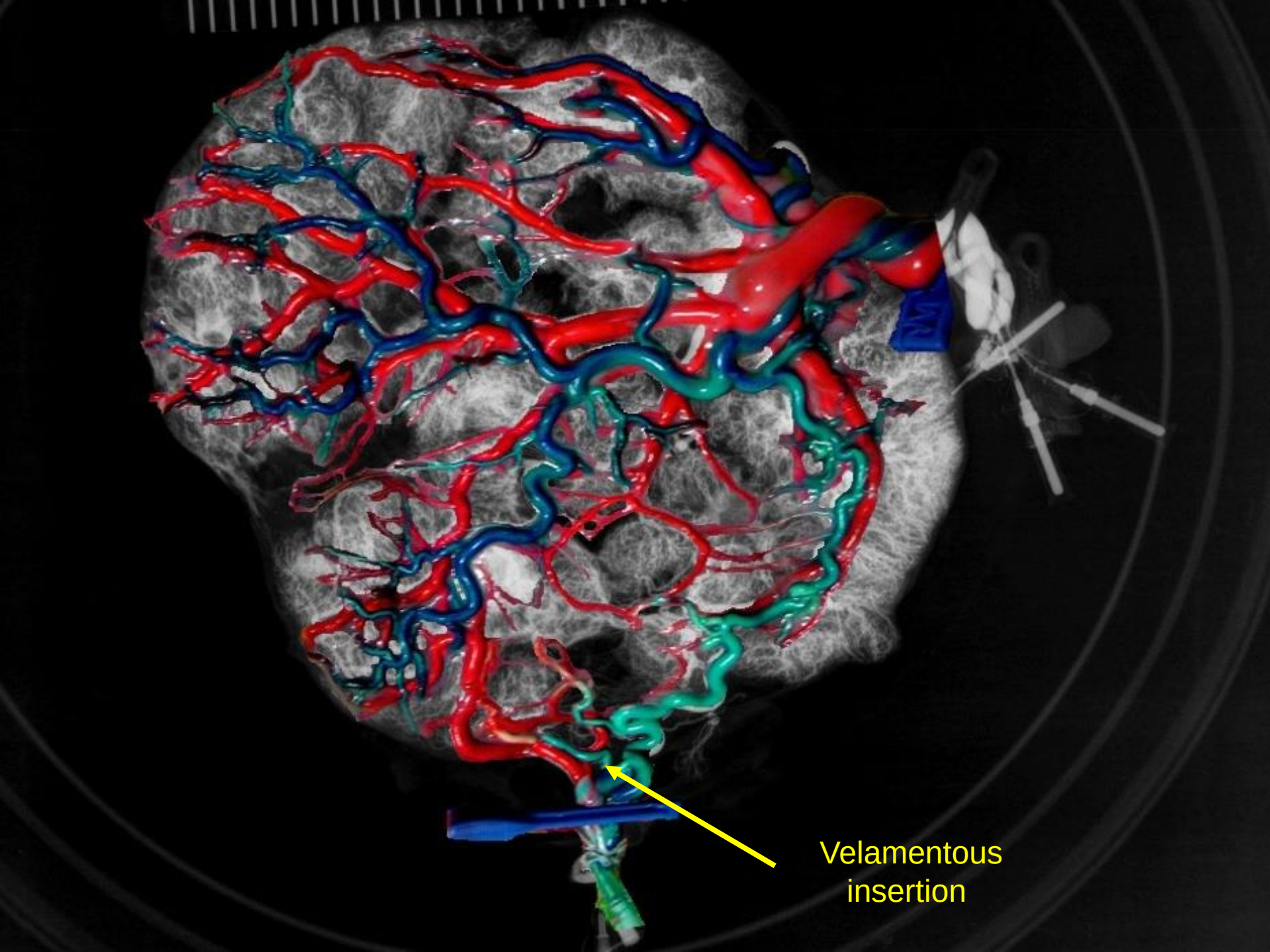
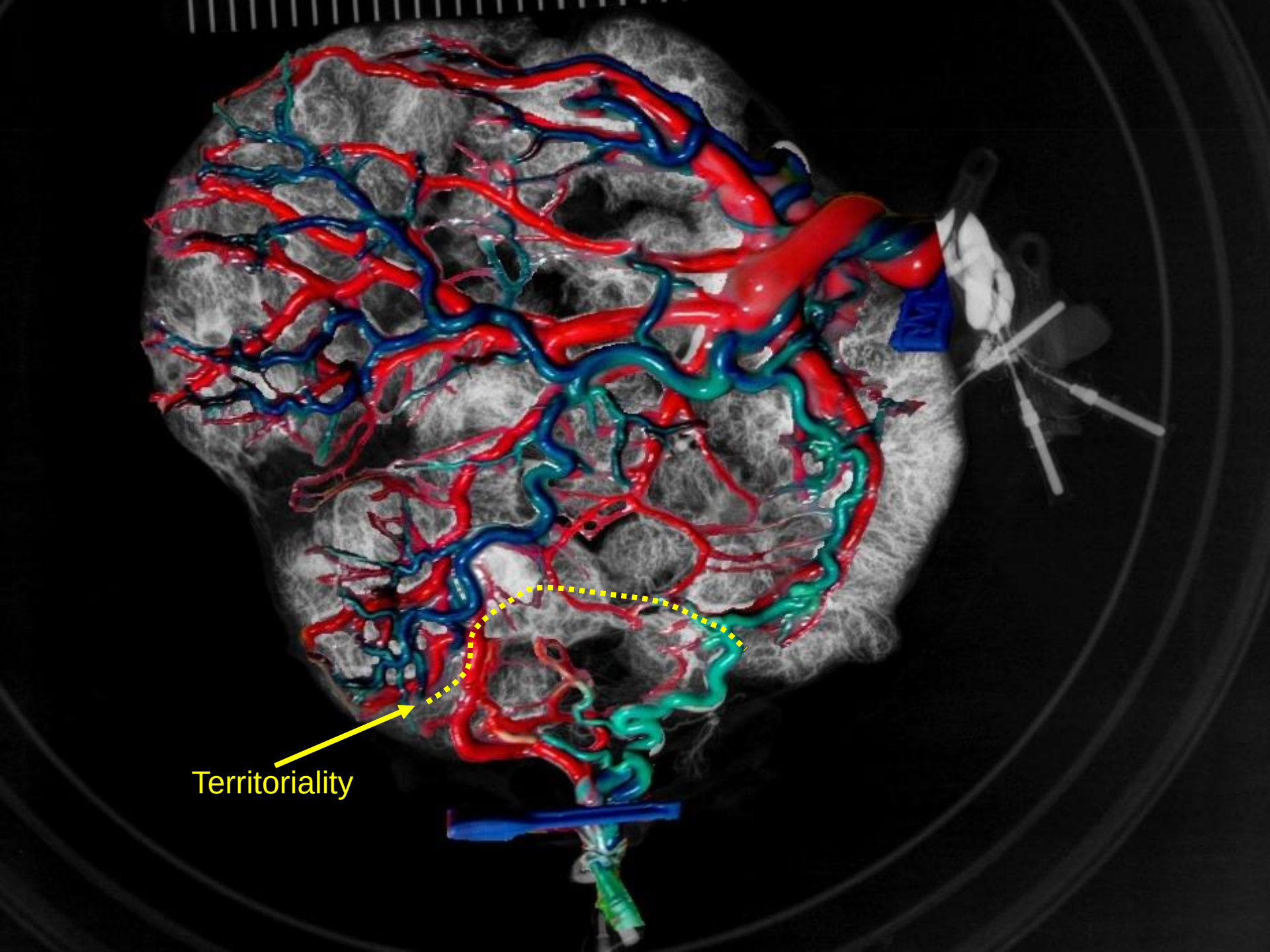
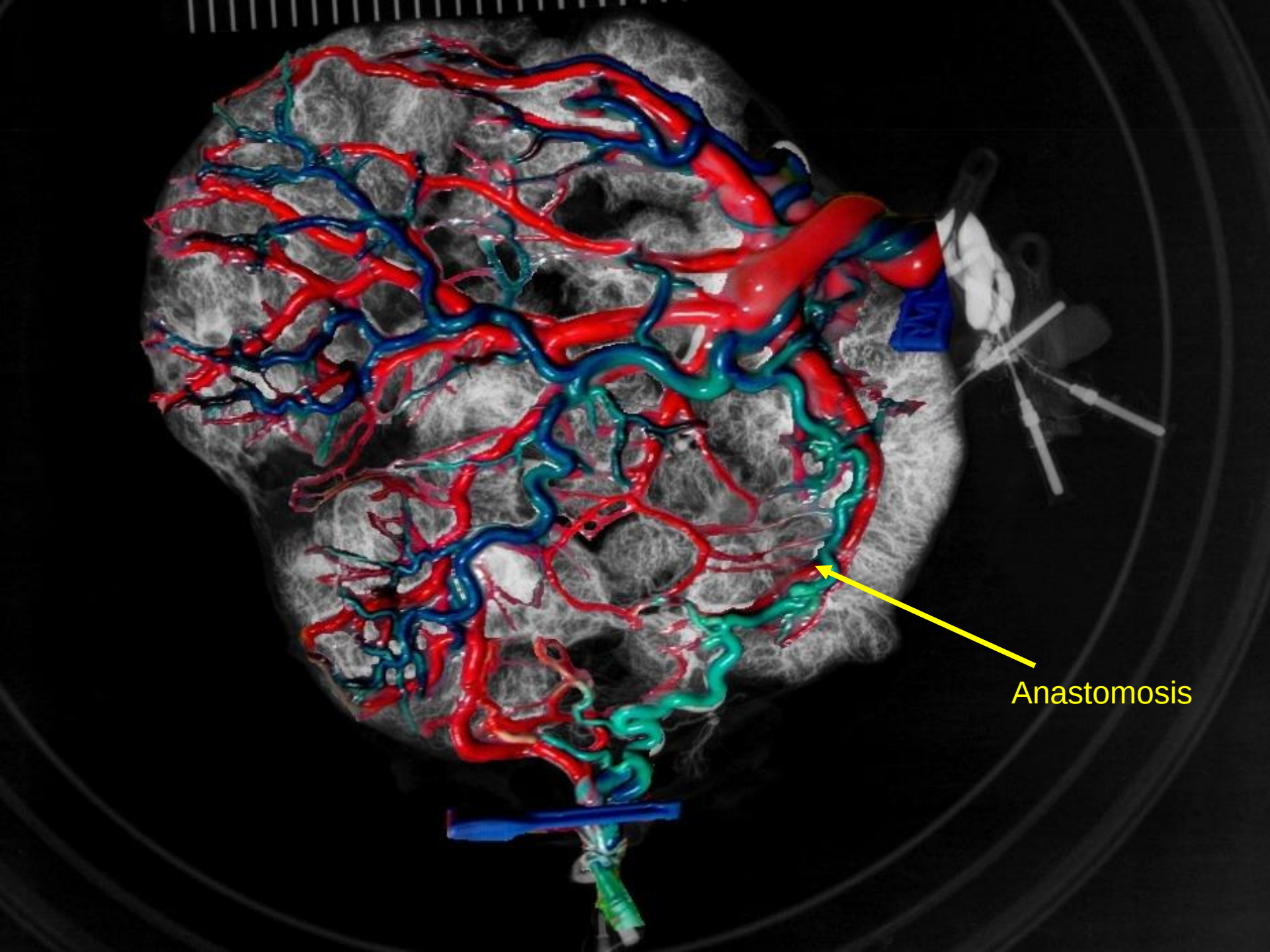
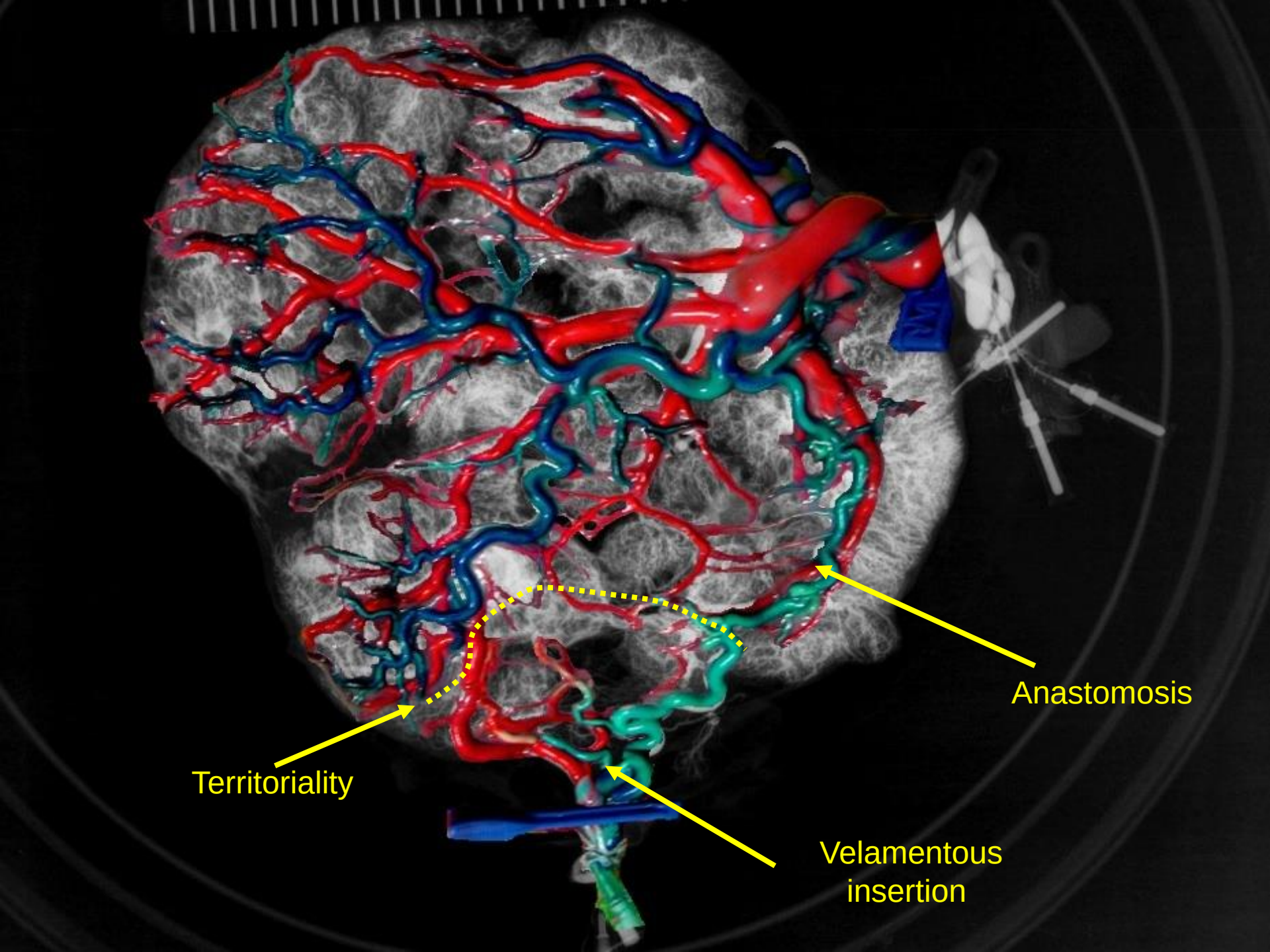
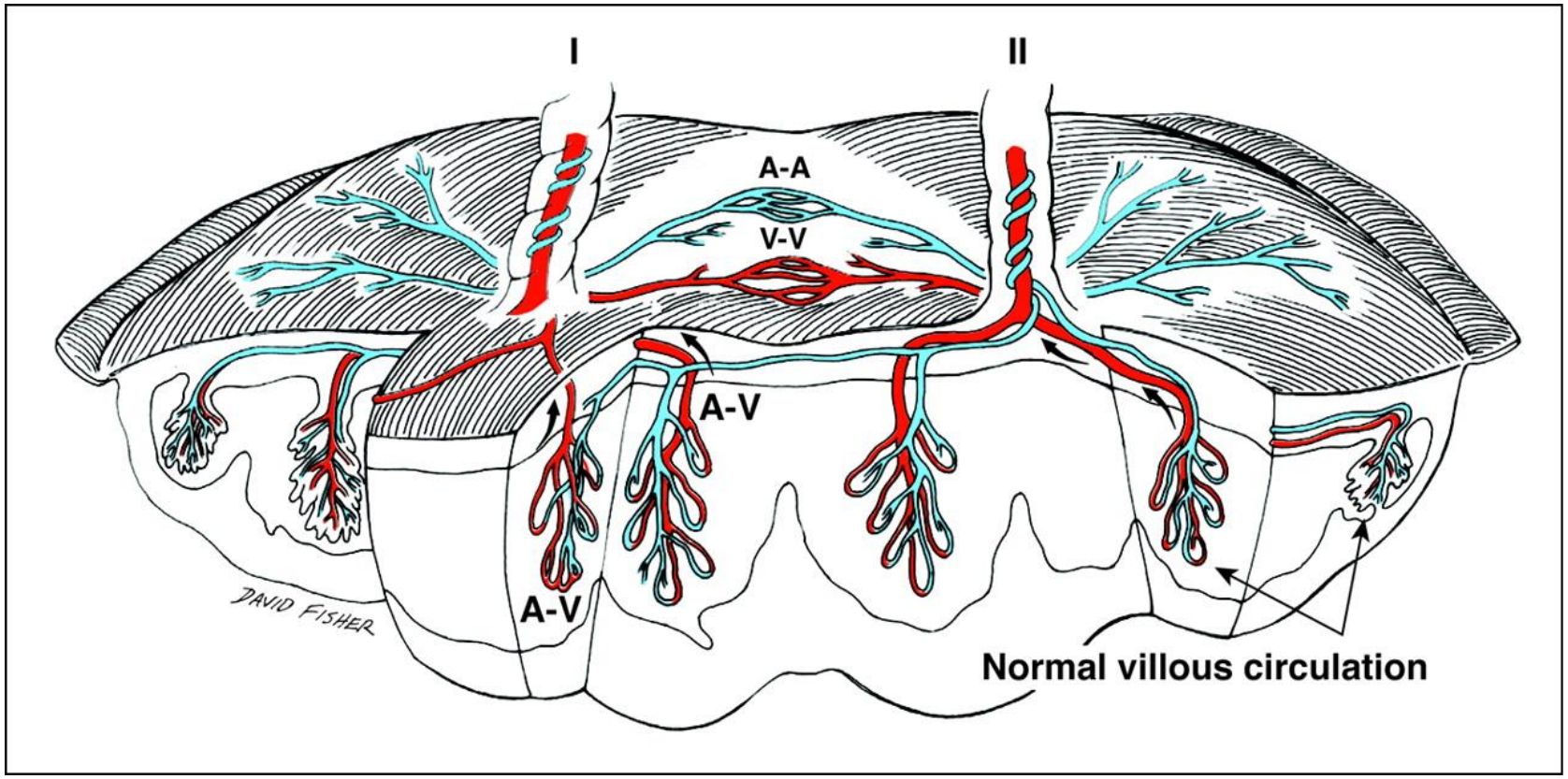

Velamentous
insertion

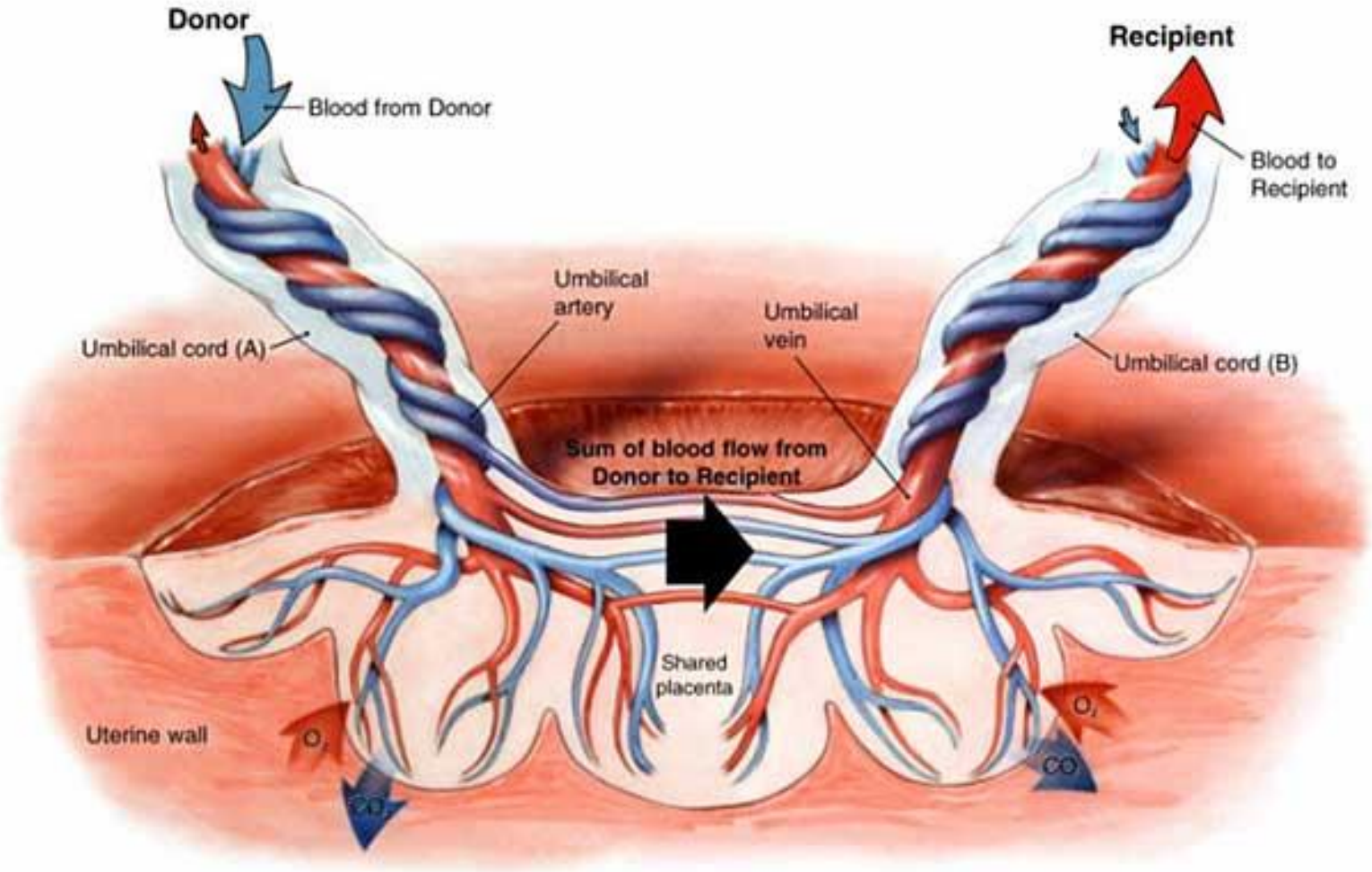

Territoriality

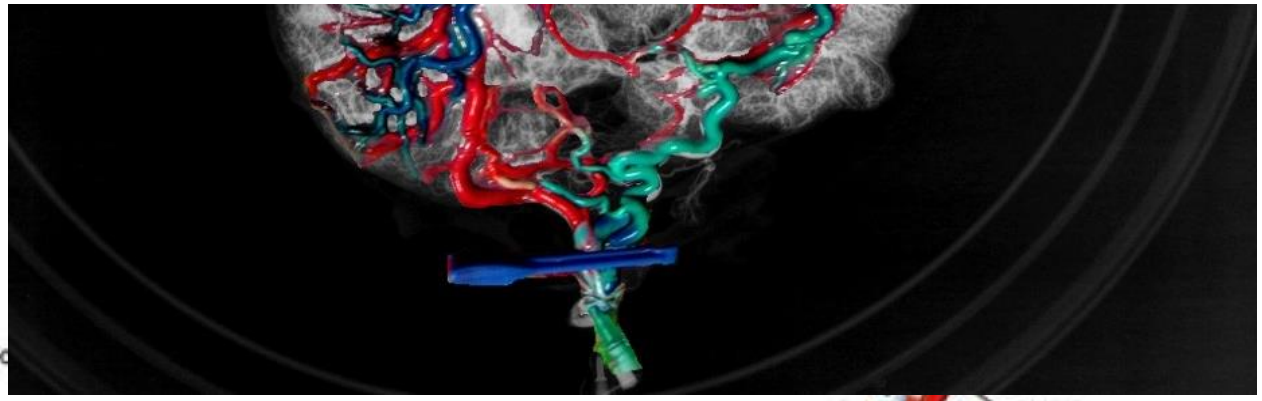

Anastomosis

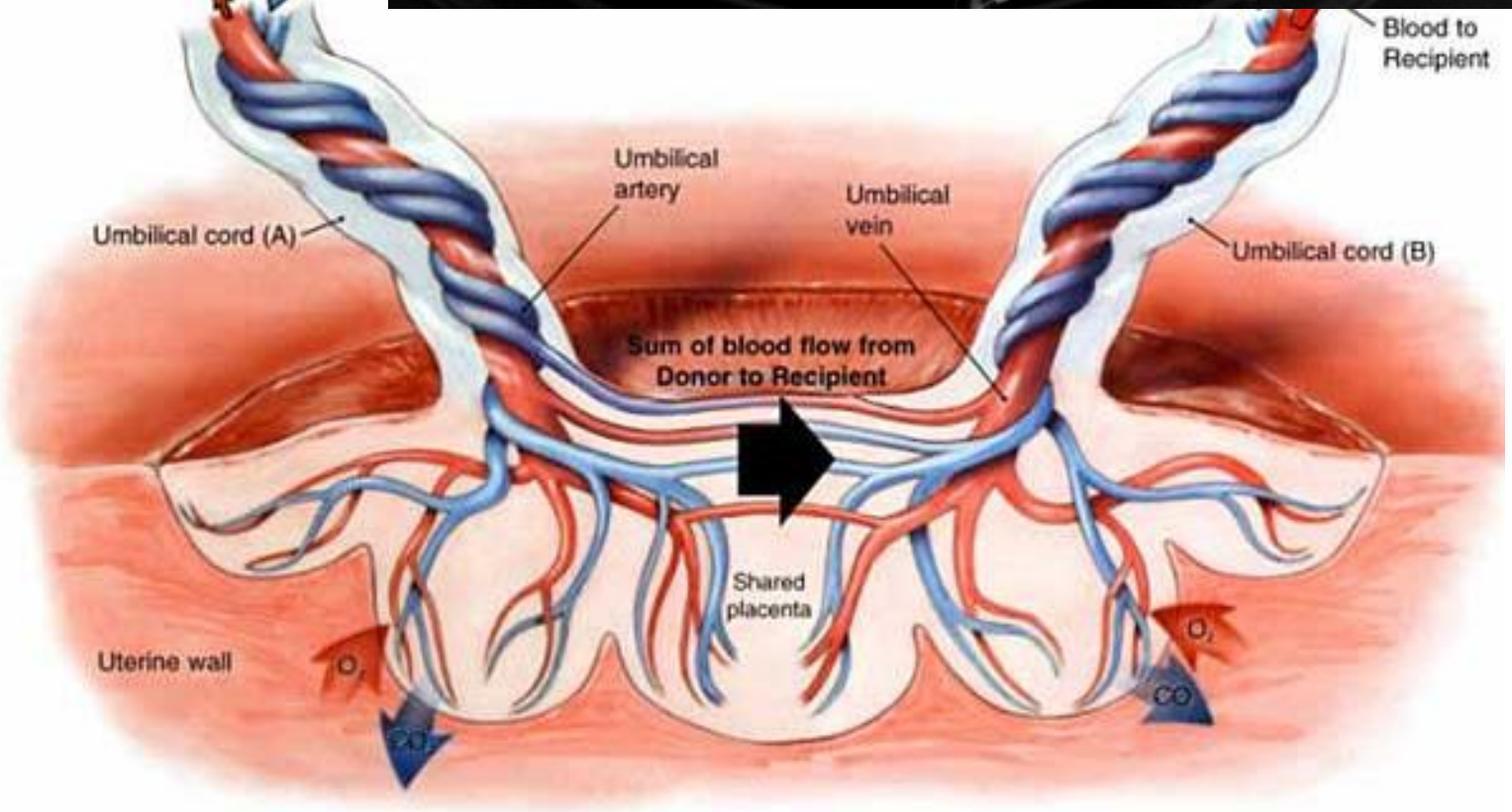

Anastomosis
Territoriality
Velamentous
insertion


I
II
A-A
V-V
A-V
A-V
DAVID FISHER
Normal villous circulation

Twin / Twin Transfusion Syndrome






Donor
Blood to Recipient
Umbilical artery
Umbilical vein
Umbilical cord (A)
Umbilical cord (B)
Sum of blood flow from Donor to Recipient
Shared placenta
Uterine wall
O_2
CO_2
O_2
CO

Shunt(s)

Cardiac overload (r)

Polyhydramnios

[Hormonal output (r)]

Oligohydramnios (d)

Twin Oligo Polyhydramnios Sequence = TOPS

Shunt(s)

Cardiac overload (r)

Polyhydramnios

Hormonal output (r)

Oligohydramnios (d)

~~Twin Oligo Polyhydramnios Sequence = TOPS~~

Shunt(s)

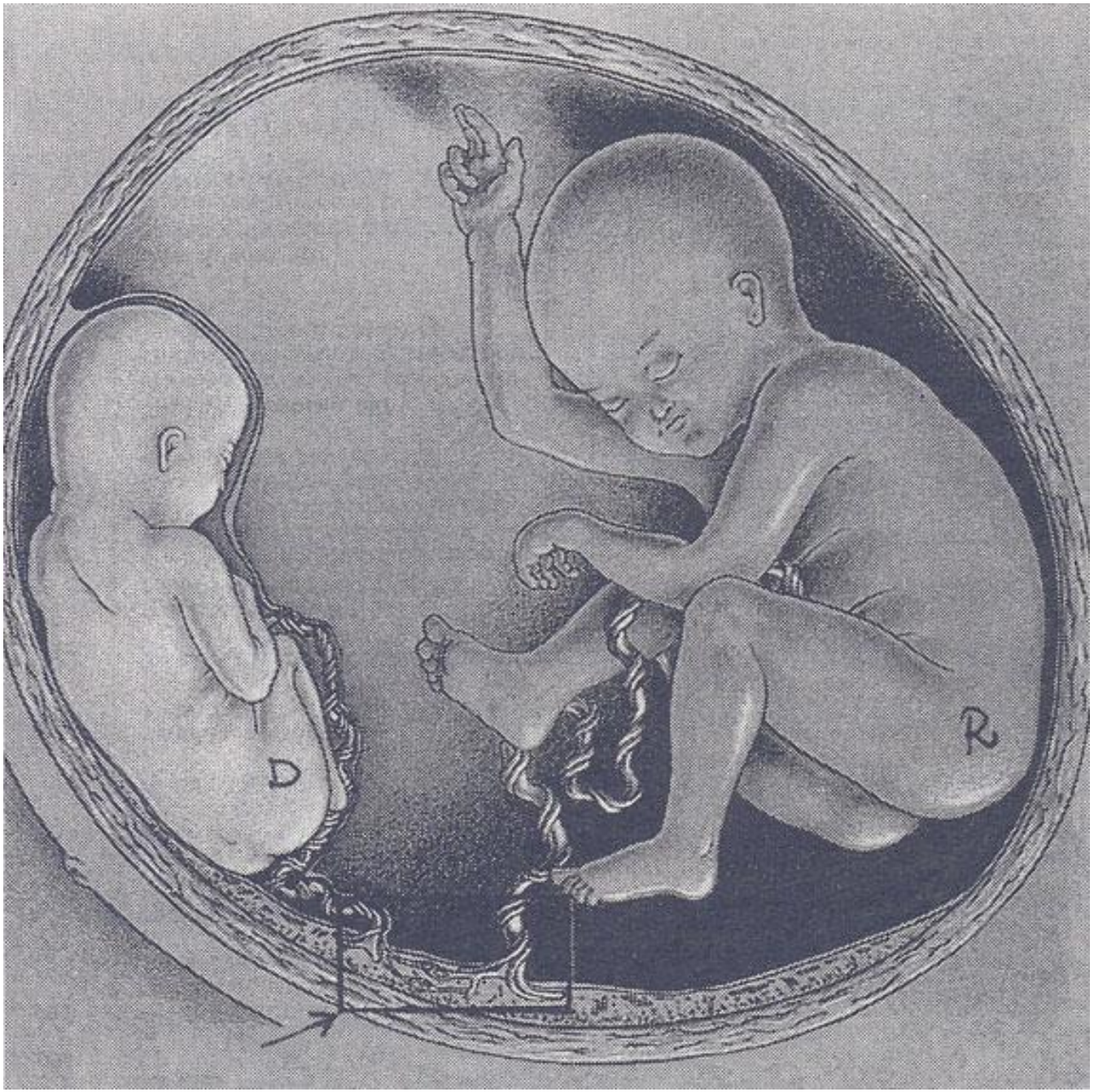
Cardiac overload (r)

Polyhydramnios

Hormonal output (r)

Oligohydramnios (d)

Twin Poly Oligohydramnios Sequence = TPOS


F
D
R

TOPS

Gestational age	DVP recipient	DVP donor
<20 weeks	≥ 8 cm	< 2 cm
≥ 20 weeks	≥ 10	< 2 cm

+

bladder filling in donor	no bladder filling in donor	abnormal Doppler findings: abnormal umbilical artery Doppler donor abnormal venous Doppler recipient	hydrops	IUFD
Stage I	Stage II	Stage III	Stage IV	Stage V

Table 2
Cincinnati staging system

Stage	Donor	Recipient	Recipient Cardiomyopathy
I	Oligohydramnios (deepest vertical pocket <2 cm)	Polyhydramnios (deepest vertical pocket >8 cm)	No
II	Bladder not visible	Bladder visible	No
III	Abnormal Doppler	Abnormal Doppler	None
IIIA			Mild
IIIB			Moderate
IIIC			Severe
IV	Hydrops	Hydrops	
V	Death	Death	
Variables			
Cardiomyopathy	Mild	Moderate	Severe
AV regurgitation	Mild	Moderate	Severe
RV/LV thickness[a]	>2 + Z-score	>3 + Z-score	>4 + Z-score
MPI[a,b]	>2 + Z-score	>3 + Z-score	Severe biventricular dysfunction

Crombleholme et al, 2009

So what ?

Table 5
Short-term neonatal outcome inTTTS

Author/Year	TTTS Pregnancy (n)	Gestational Age at Delivery (wk)	Delivery Mode: Spontaneous Vaginal Delivery (%)	Delivery Mode: Cesarean Section (%)	Preterm Labor <33 wk (%)	Acute Renal Failure (%)	Necrotizing Enterocolitis (%)	Respiratory Distress Syndrome (%)	Intraventricular Hemorrhage Grade 3–4 (%)
Dickson/2000[103]	112	29	NA	NA	NA	7	3	27	16
Duncomb/ 2003[113]	69	29.4	NA	NA	NA	4.8	2.9	62	5.8
Lepoir/2005[104]	85	32.6	34	66	NA	NA	3	34	14
Lufti/2005[106]	48	NA	27	73	27	5.2	NA	27	13
Acosta/2007[107]	101	33	NA	NA	44	NA	4.3	43	4
Lenclen/2007[108]	79	29	NA	NA	NA	NA	NA	NA	NA

Crombleholme et al, 2009

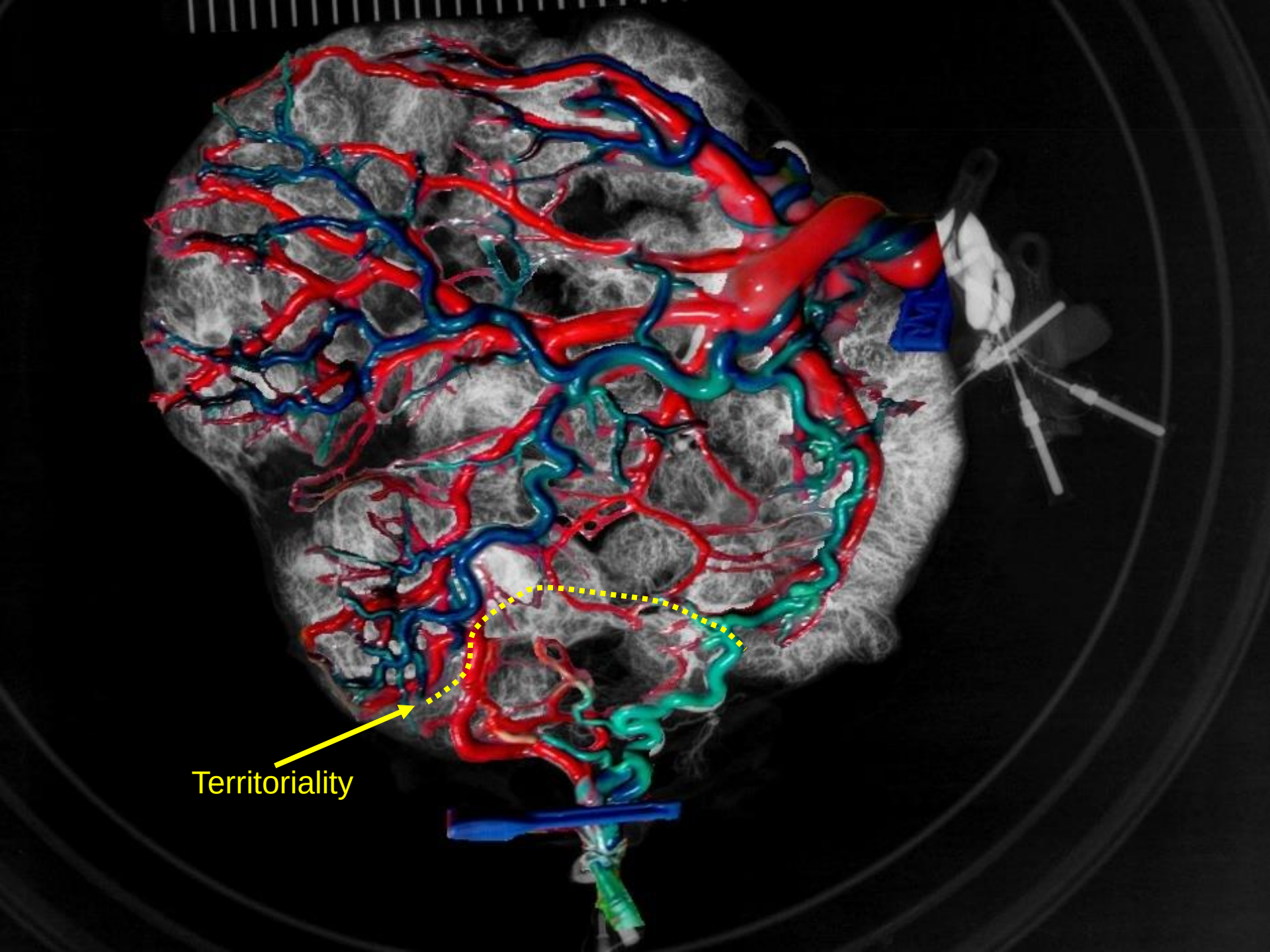
Long term cardiac follow up of severe twin to twin transfusion syndrome after intrauterine laser coagulation

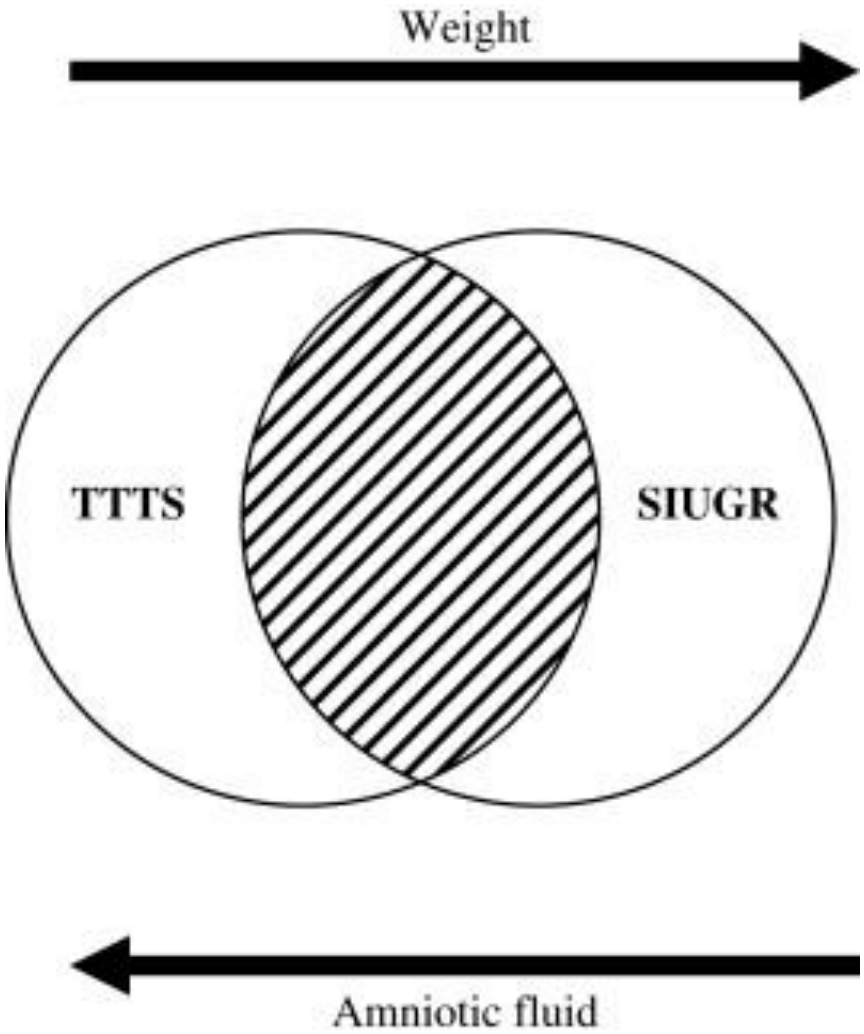
U Herberg, W Gross, P Bartmann, C S Banek, K Hecher, J Breuer

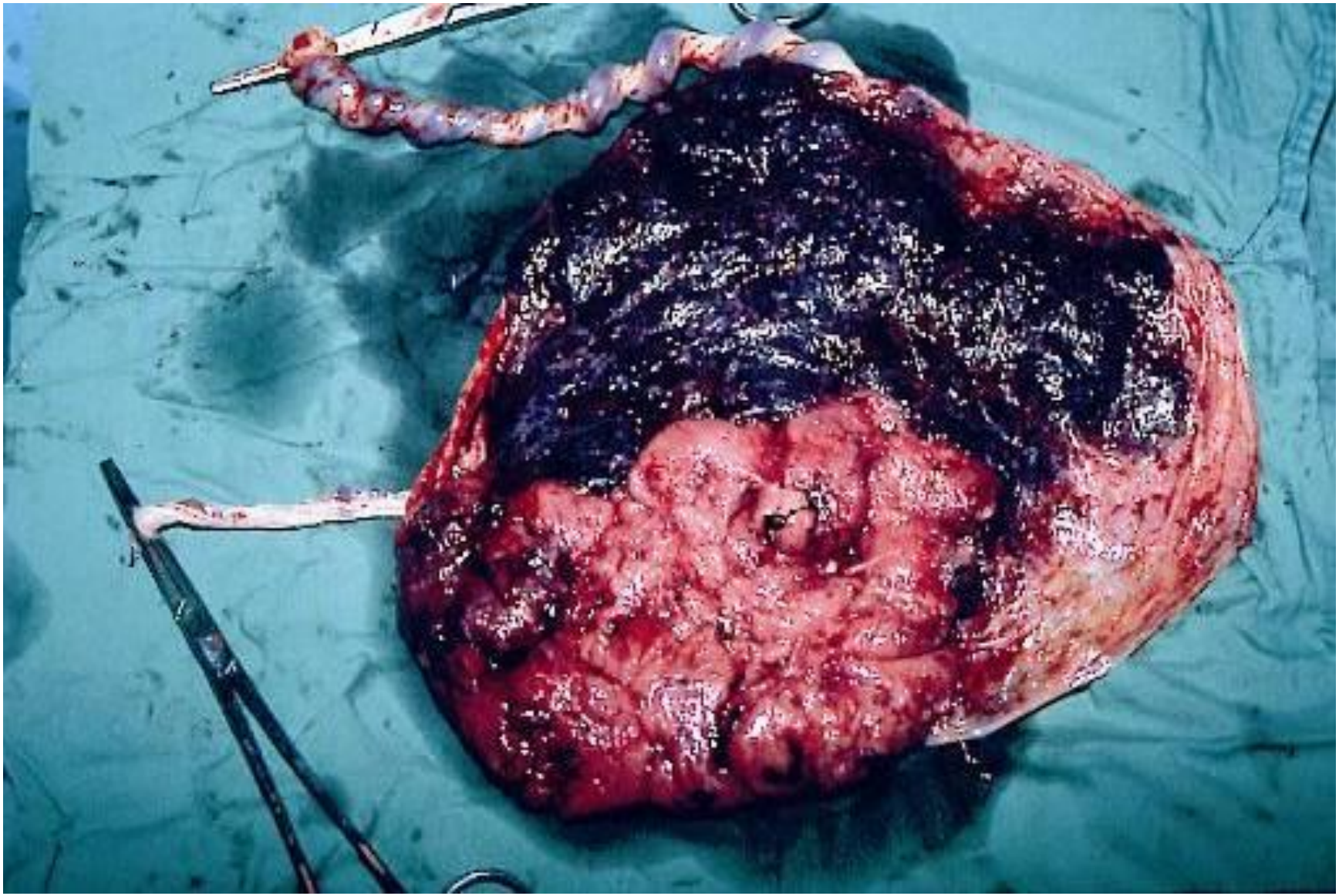
Table 1 Prevalence of congenital heart disease in survivors after twin to twin transfusion syndrome in the study cohort compared with the prevalence at birth reported by the Baltimore-Washington infant study (WIS)[28] and Wren *et al*[29]

Postnatal examination	All (n = 89)	Recipients (n = 51)	Donors (n = 38)	Prevalence per 100 live births	
				WIS[28]	Wren[29]
Normal	78 (87.6%)	44 (86.3%)	34 (89.5%)		
Acquired pulmonary HT	1	0	1		
Congenital heart disease	10 (11.2%)	7 (13.7%)	3 (7.89%)	0.49	0.559
95% CI	5.52 to 19.7	5.70 to 26.2	1.66 to 9.25		
Pulmonary stenosis	4 (4.49%)	4 (7.84%)	0 (0)	0.038	0.033
95% CI	1.24 to 11.1	2.18 to 18.9	0 to 9.25		
ASD (all sizes)	5 (5.62%)	2 (3.92%)	3 (7.89%)		
95% CI	1.86 to 12.6	0.48 to 13.5	0.48 to 13.5		
Medium sized ASD	2 (2.25%)	1 (1.96%)	1 (2.63%)	0.032	0.023
95% CI	0.27 to 7.88	0.05 to 10.4	0.07 to 13.8		
Ventricular septal defect	1 (1.12%)	1 (1.96%)	0 (0)	0.146	0.188
95% CI	0.03 to 6.10	0.05 to 10.4	0.0 to 9.25		

ASD, atrial septal defect; CI, confidence interval; HT, hypertension.


Territoriality


Weight
TTTS
SIUGR
Amniotic fluid



IUGR in twins

- Relative growth restriction
- Absolute growth restriction

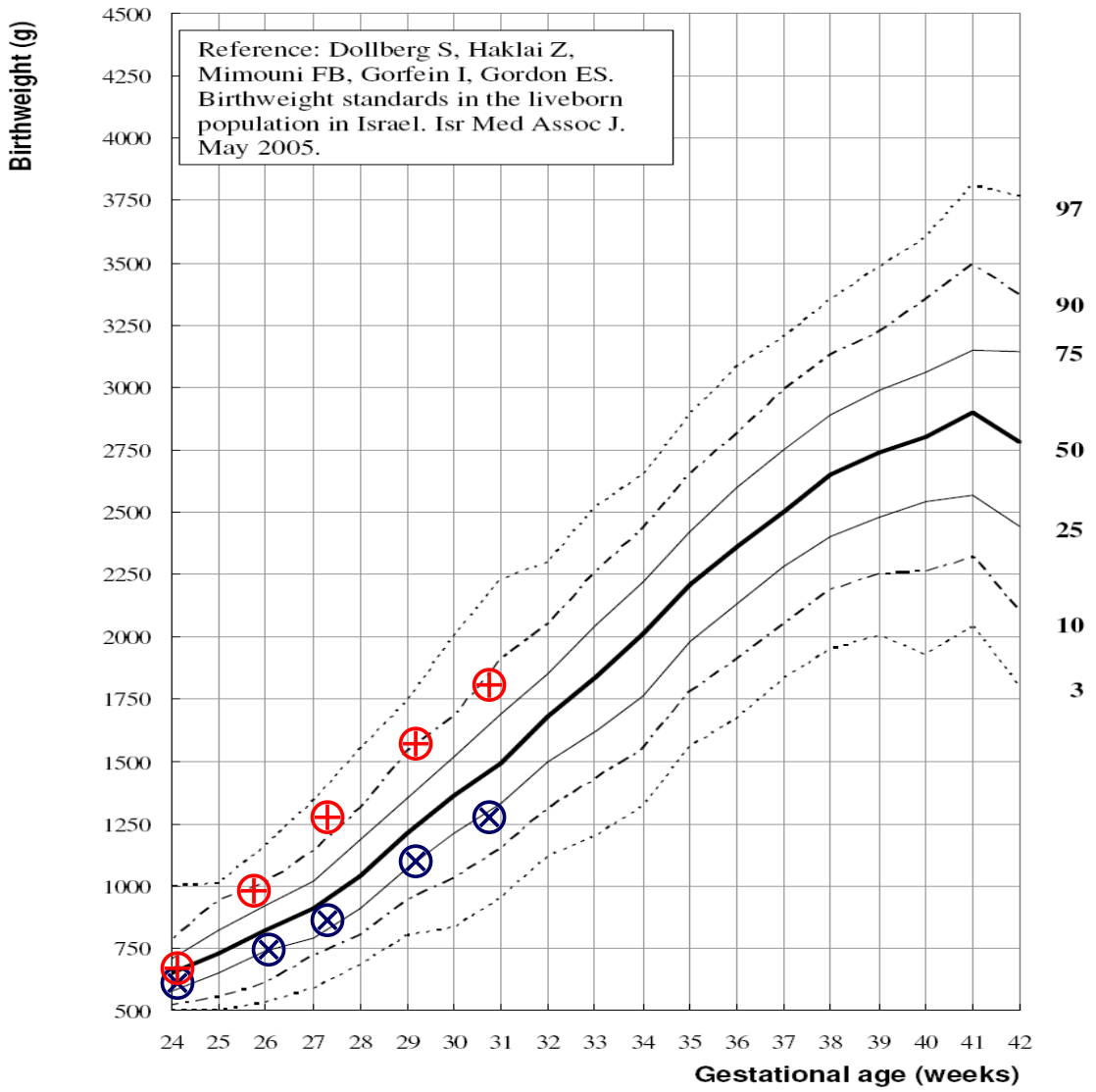
sIUGR in twins

- Relative growth restriction

+

- Absolute growth restriction

Classification of Israeli multiple gestation neonates (both sexes) by intrauterine growth and gestational age (3-97th percentiles)

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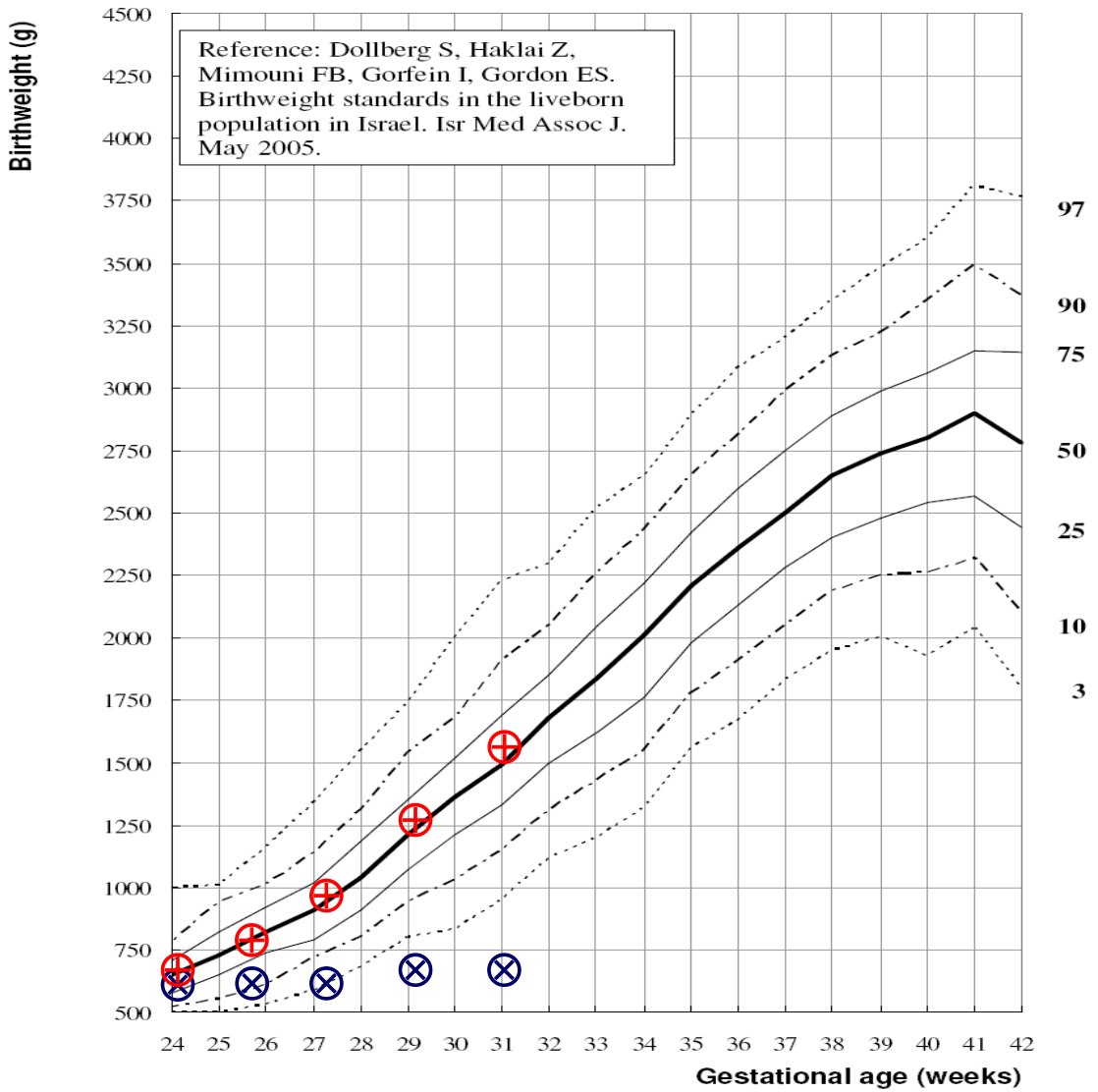
Kanpai-


Kanpai-

Classification of Israeli multiple gestation neonates (both sexes) by intrauterine growth and gestational age (3-97th percentiles)



1540 g 450 g

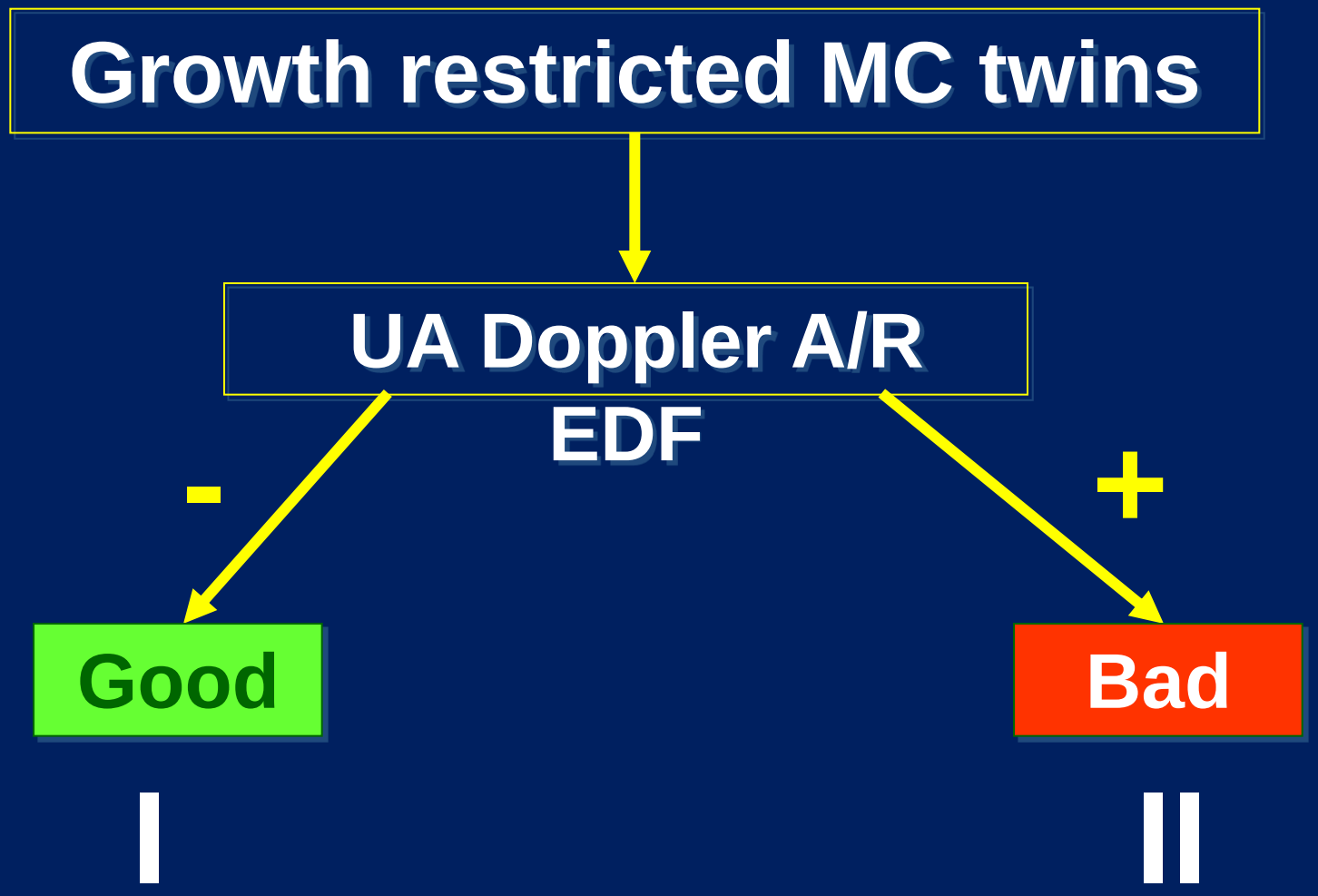
Growth restricted MC twins

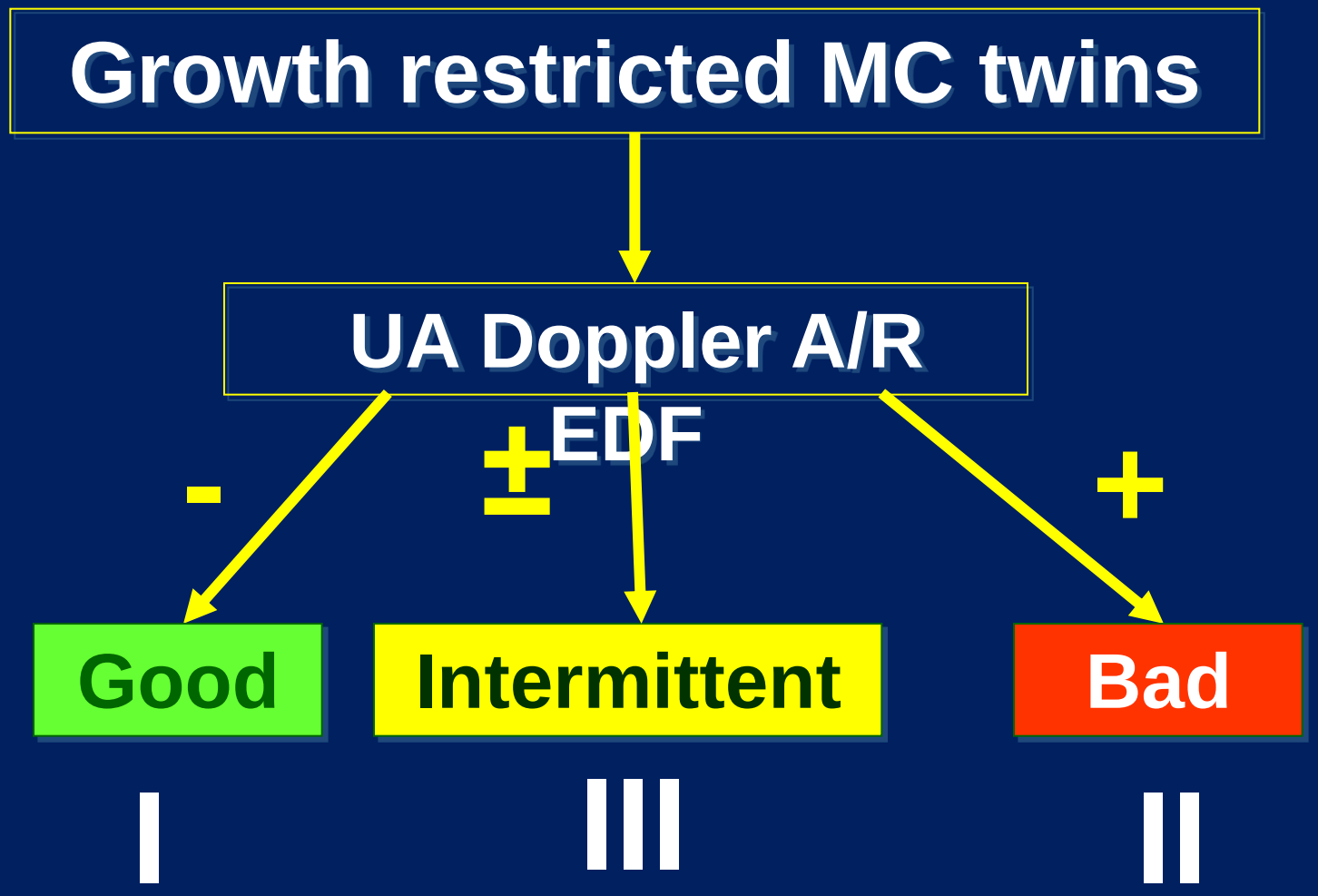
UA Doppler A/R
EDF

-

Good

I


Growth restricted MC twins
UA Doppler A/R
EDF
-
+
Good
Bad
I
II


Growth restricted MC twins
UA Doppler A/R
EDF
-
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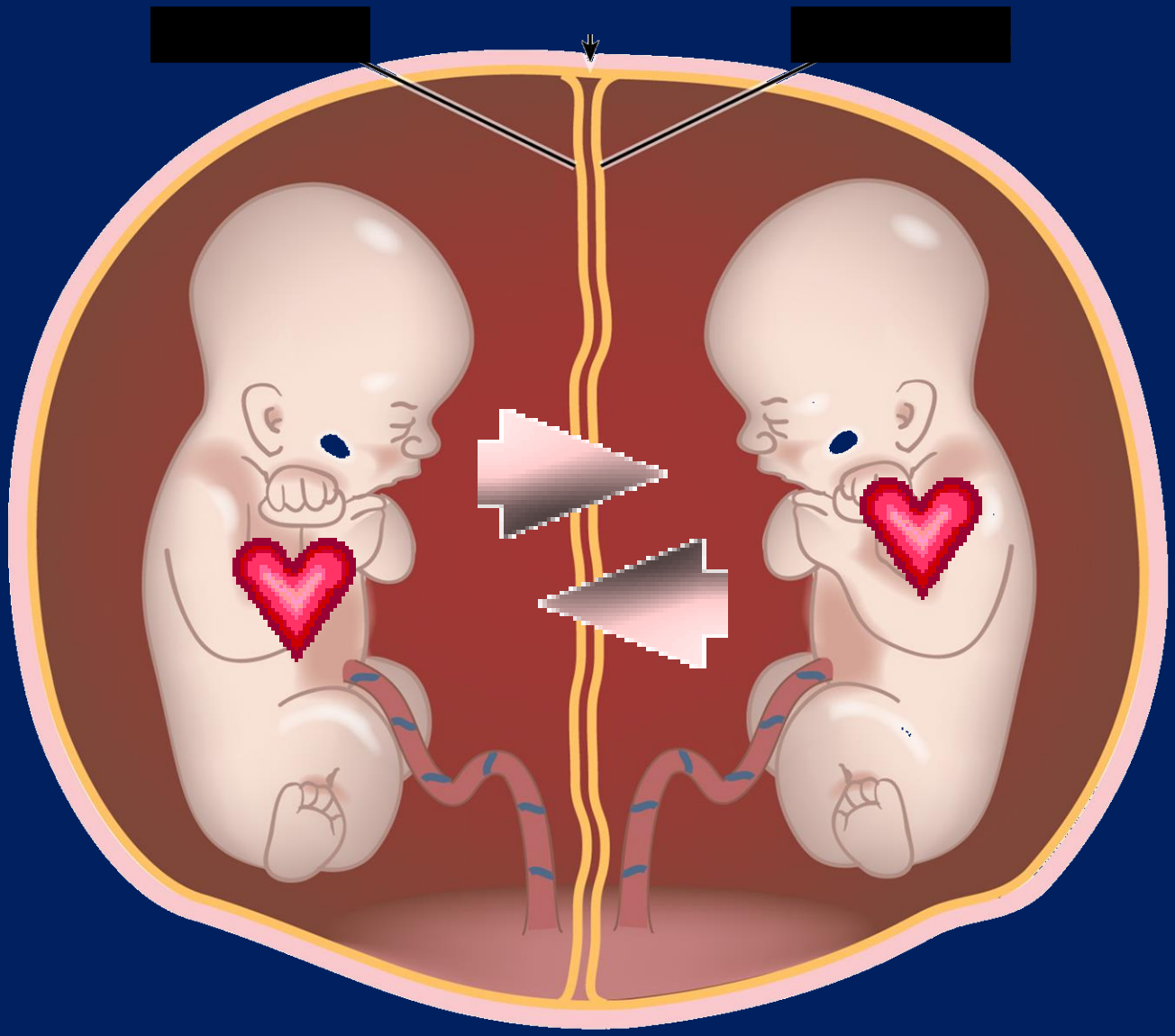
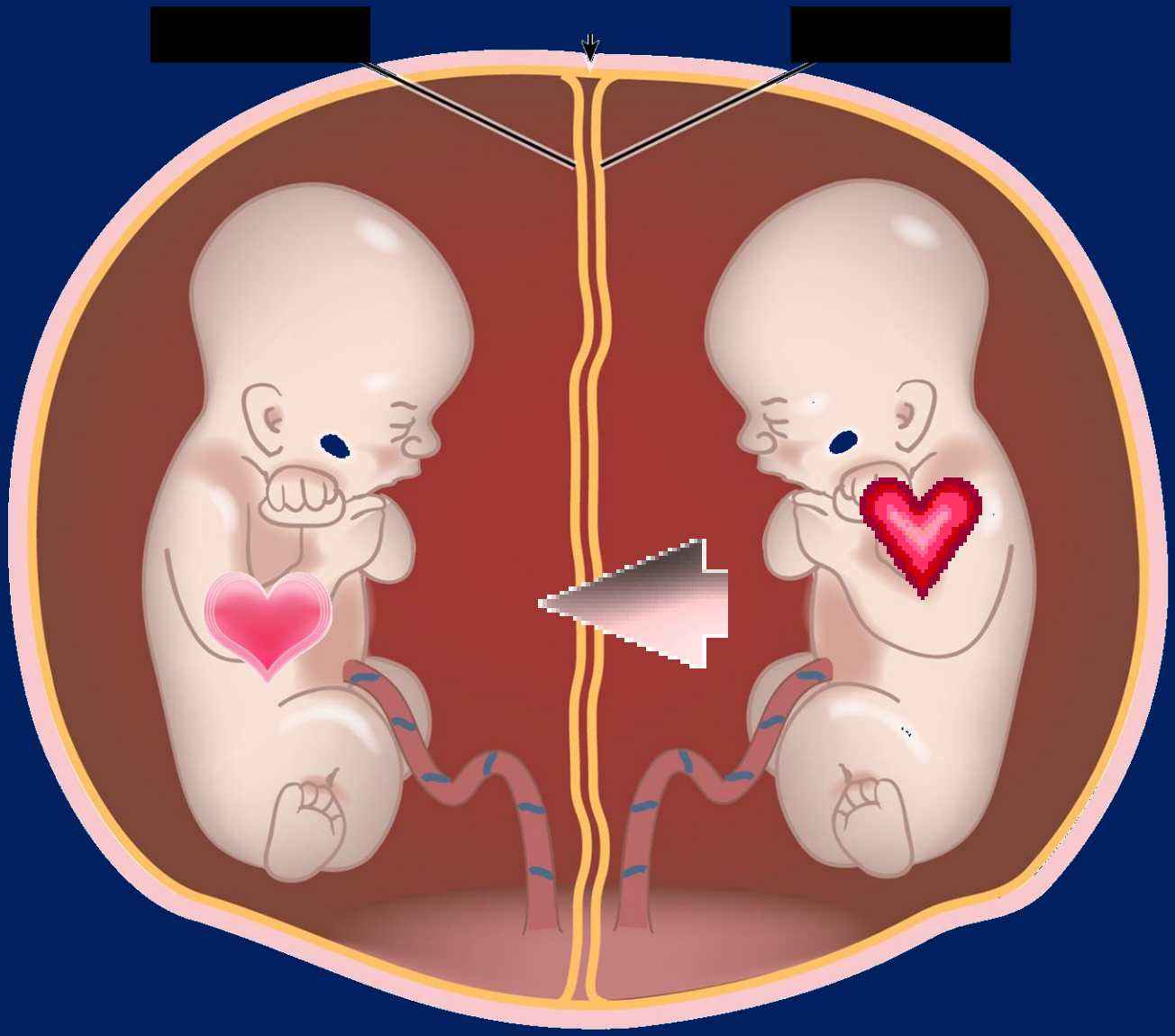
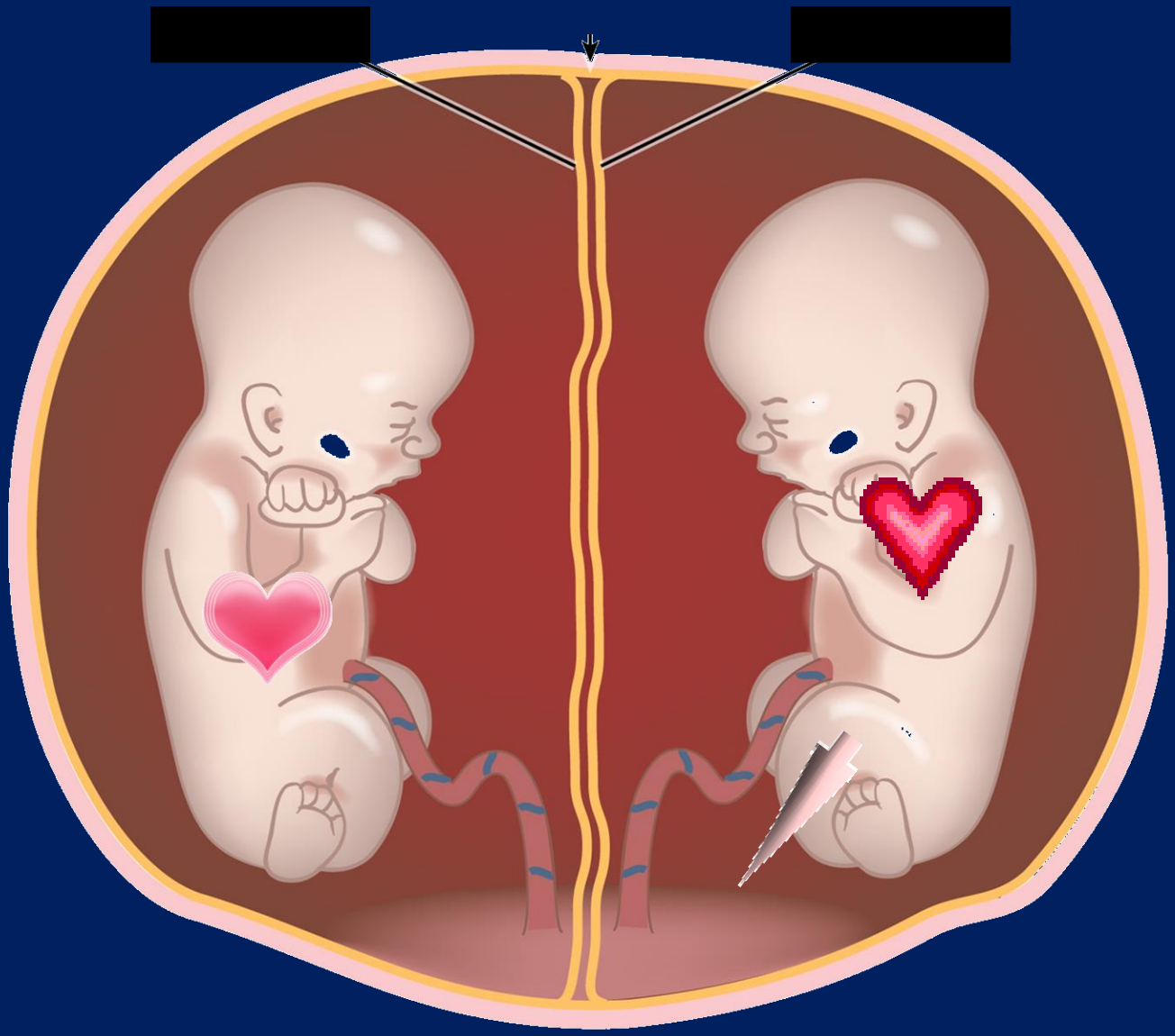
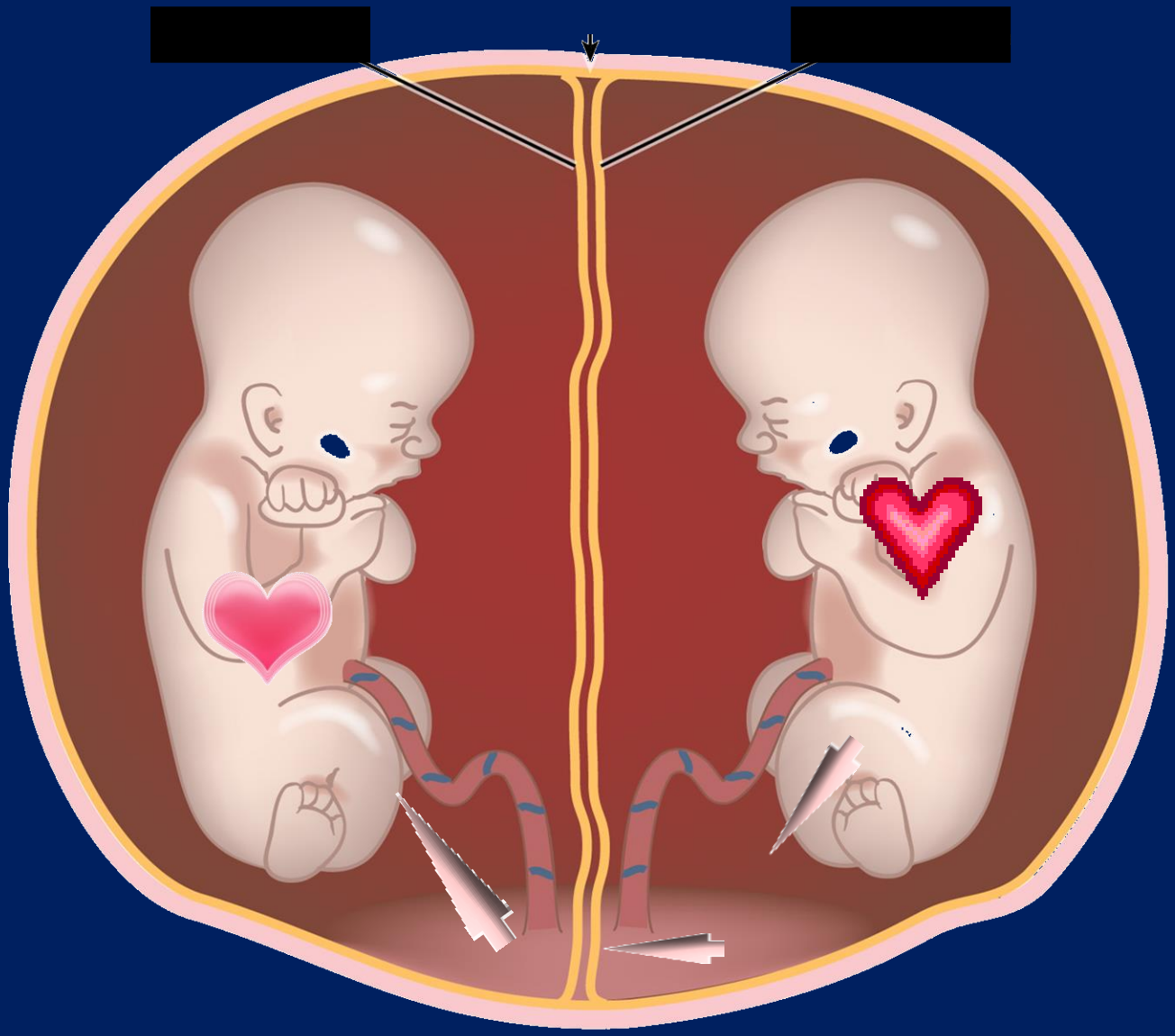
+
Good
Intermittent
Bad
I
III
II

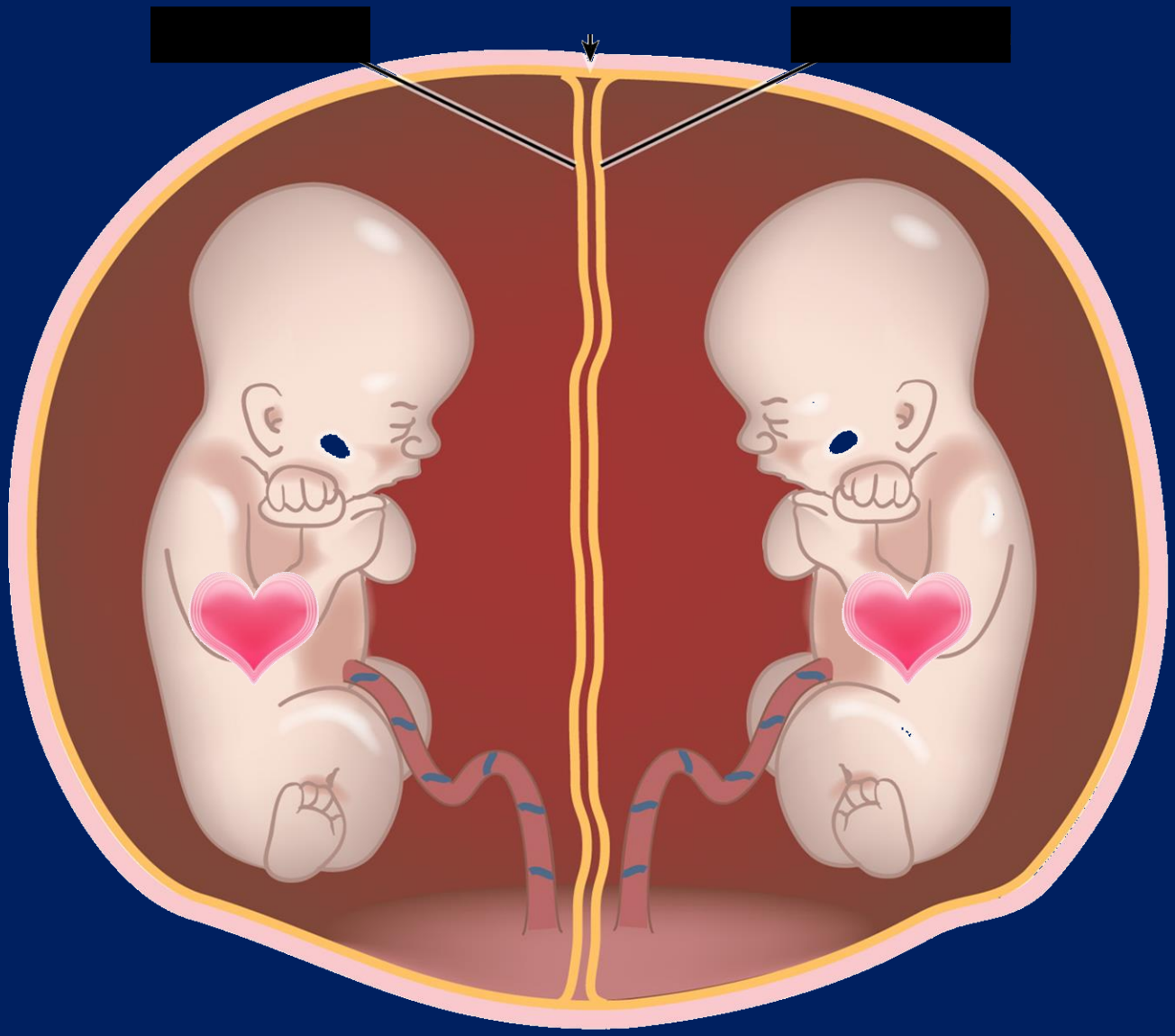
So what ?



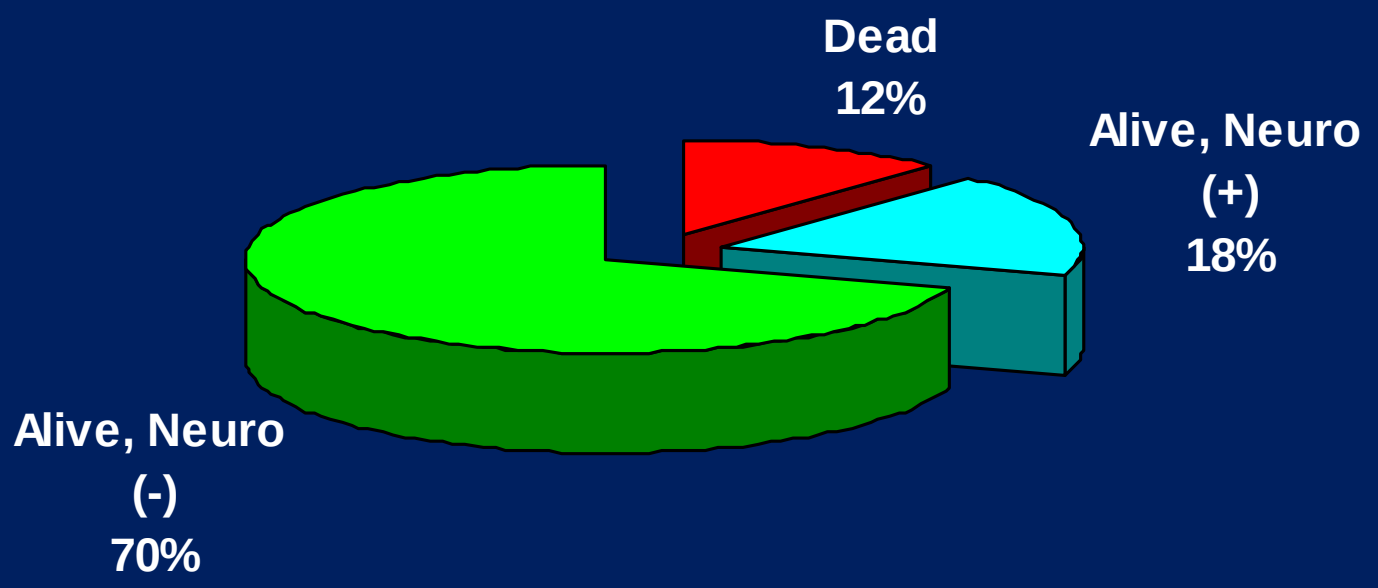








Prognosis for the co-twin following single-twin death in MC twins



Ong et al, BJOG 2006

In MC twins

Risk of sIUGR:

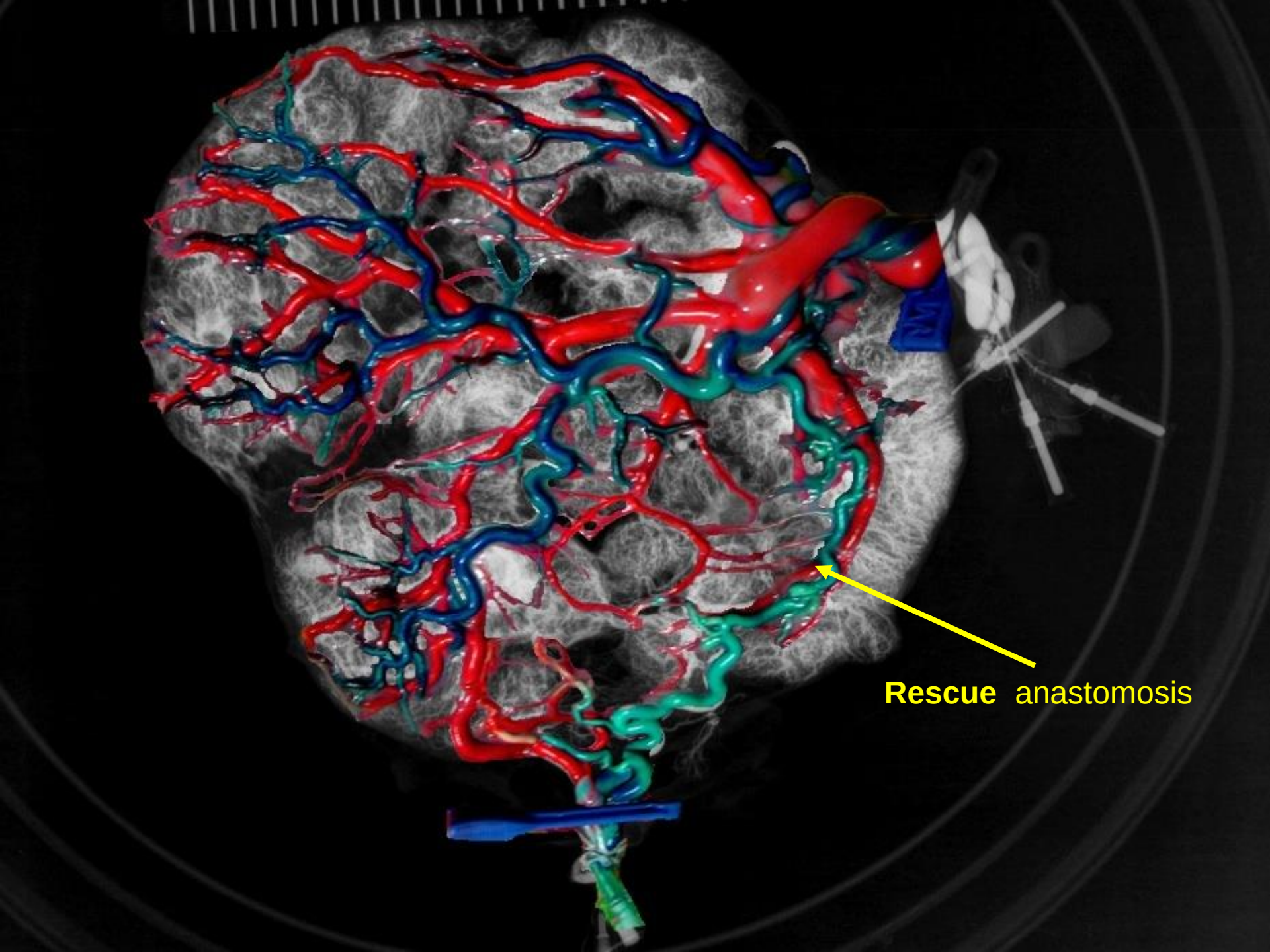
sIUFD

Ultrasound predictors of mortality in monochorionic twins with selective intrauterine growth restriction

Predictor	*Survival* (n *(%))*	*Death* (n *(%))*	*OR (95% CI)*	P
UA Doppler				
Type I	30 (97)	1 (3)	—	—
Type II	28 (51)	27 (49)	28.9 (3.7–227.3)	< 0.001
Type III	12 (80)	3 (20)	7.5 (0.7–79.4)	—

ISHII et al

Ultrasound Obstet Gynecol 2011; 37: 22–26


Rescue anastomosis

Fetal Diagnosis and Therapy

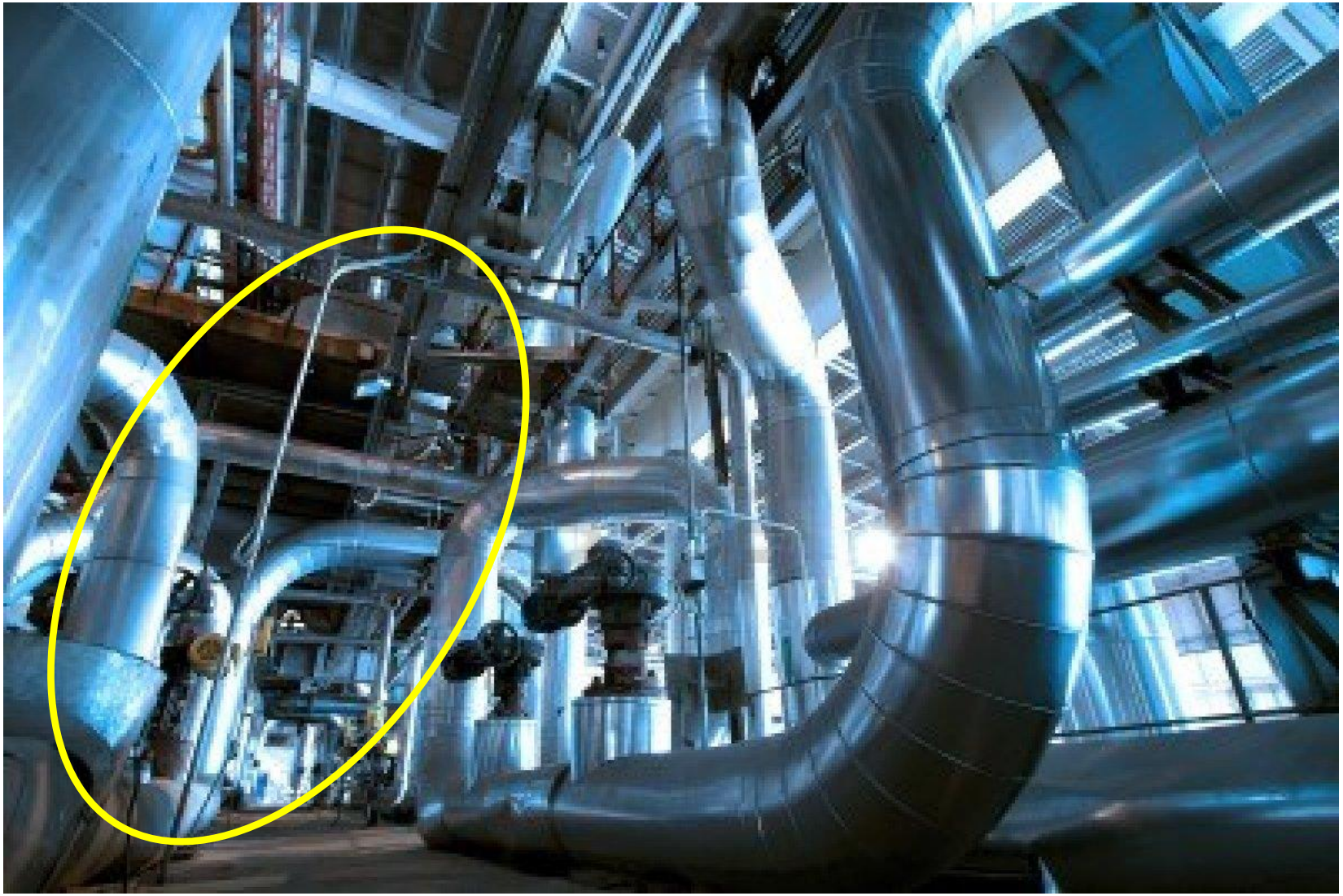
Fetal Diagn Ther 2010;27:181–190
DOI: 10.1159/000304512

Twin Anemia-Polycythemia Sequence: Diagnostic Criteria, Classification, Perinatal Management and Outcome

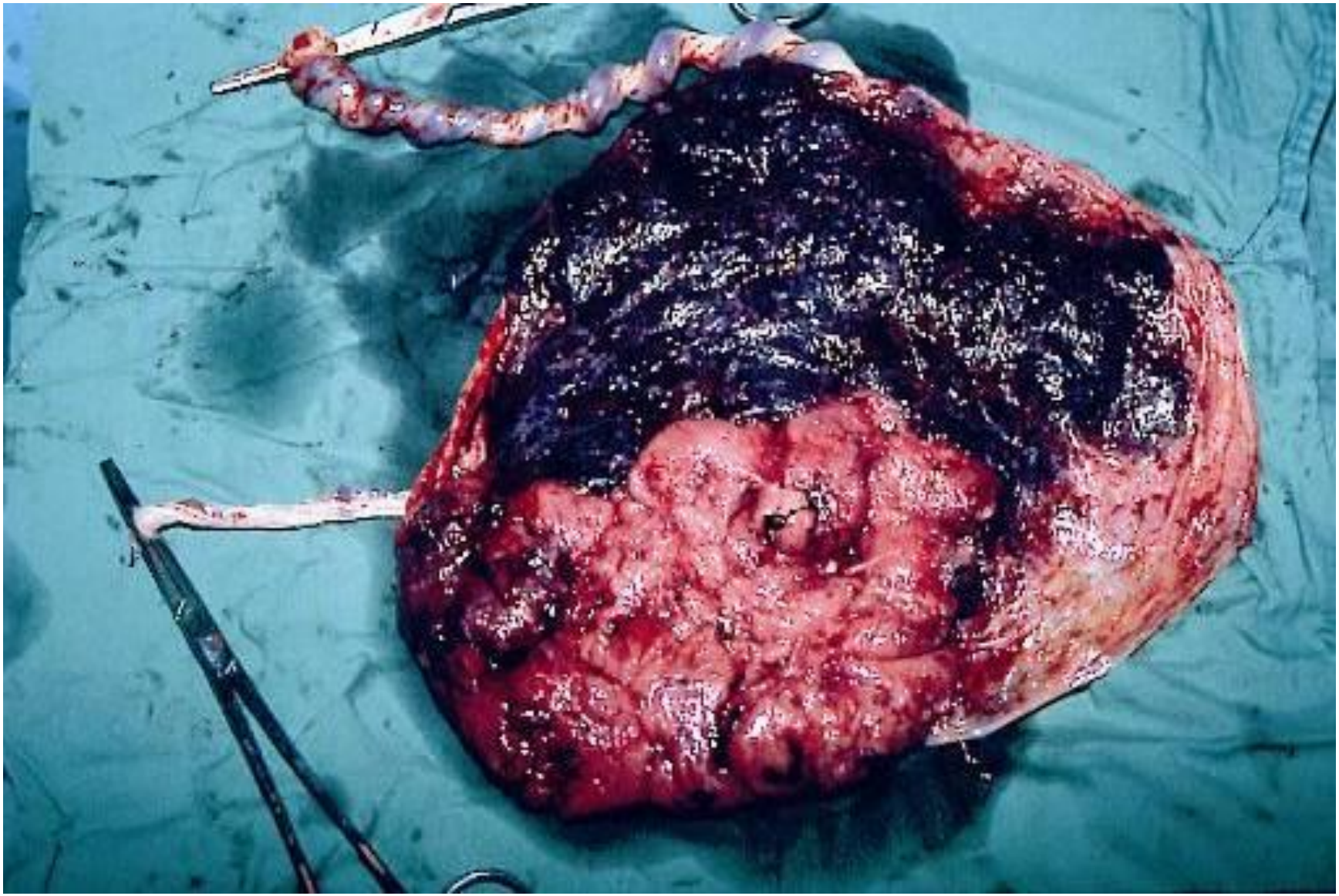
F. Slaghekke[a] W.J. Kist[a] D. Oepkes[a] S.A. Pasman[a] J.M. Middeldorp[a]
F.J. Klumper[a] F.J. Walther[b] F.P.H.A. Vandenbussche[a] E. Lopriore[b]

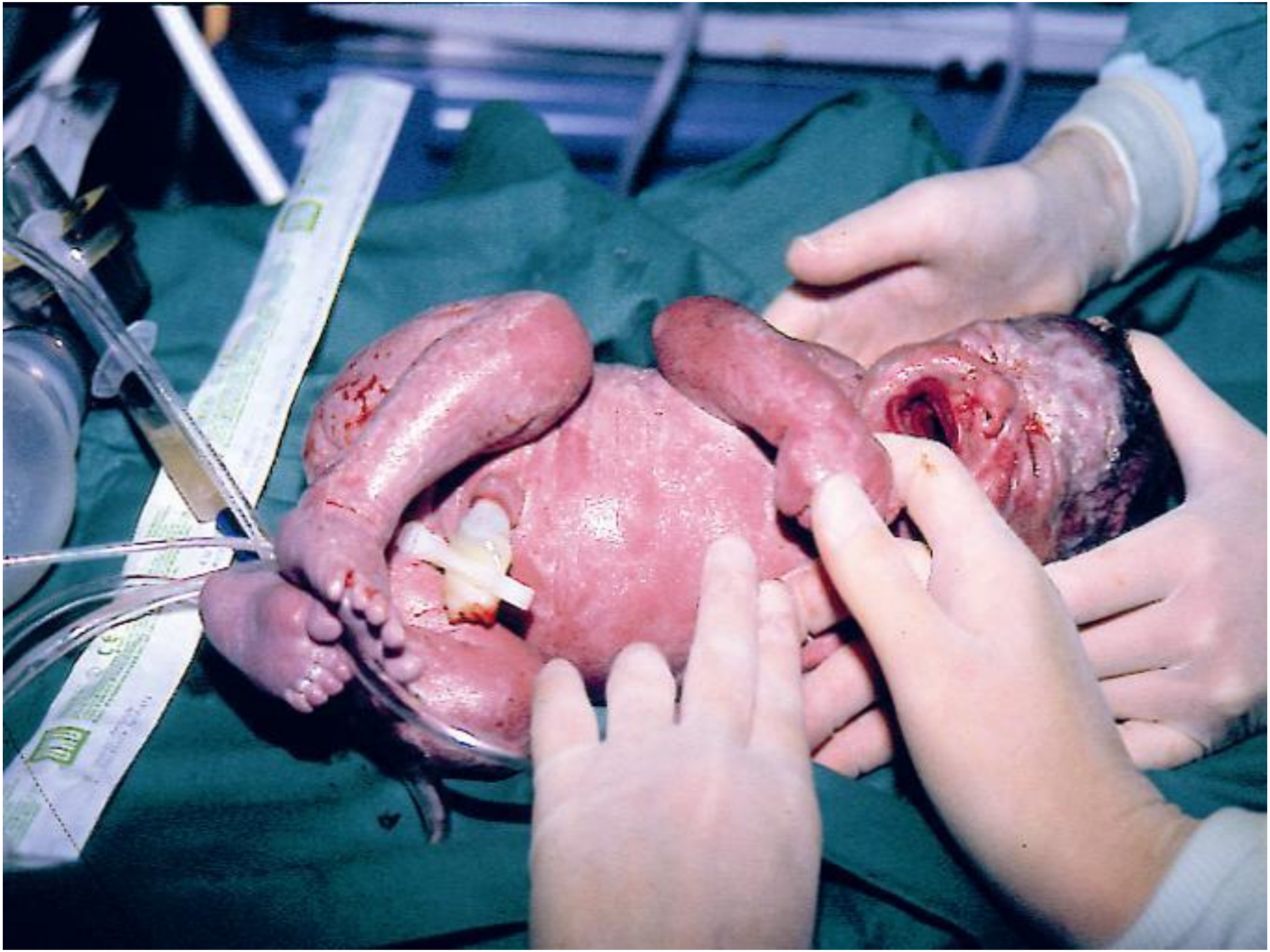
Twin Anemia Polycythemia Sequence
Large Hb difference without TTTS

- Spontaneous in 5%
- Following incomplete laser Tx for TTTS




KAPLAN OB/GYN Ultrasound Uni
06-09-2001
stetric
13.8cm
33Hz
12:17:55 PM
2+3 Trim.
6.00 - 3.00
Pwr -3
Gn 2
150/C 6
Map 7
Per 3
EE 1
Freeze





Prenatal Dx of TAPS

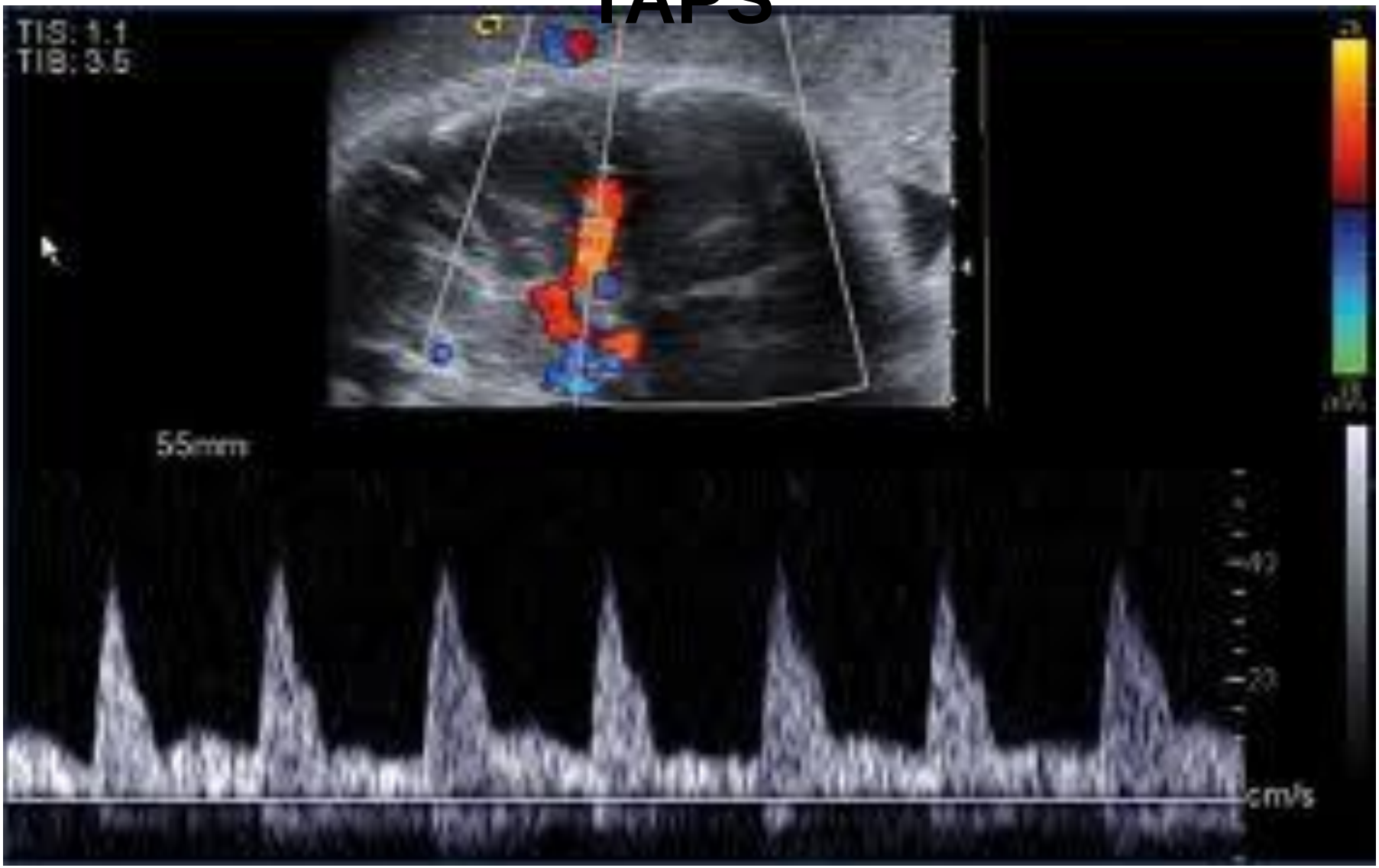


Table 1. Antenatal and postnatal diagnostic criteria for TAPS

Antenatal criteria	Postnatal criteria
MCA-PSV >1.5 MoM in the donor *and* MCA-PSV <1.0 MoM in the recipient	Intertwin Hb difference >8.0 g/dl *and* at least one of the following: – Reticulocyte count ratio >1.7 – Placenta with only small (diameter <1 mm) vascular anastomoses

Table 2. Antenatal TAPS classification

Antenatal stage	Findings at Doppler ultrasound examination

Table 3. Postnatal TAPS classification

Postnatal stage	Intertwin Hb difference, g/dl

So what ?

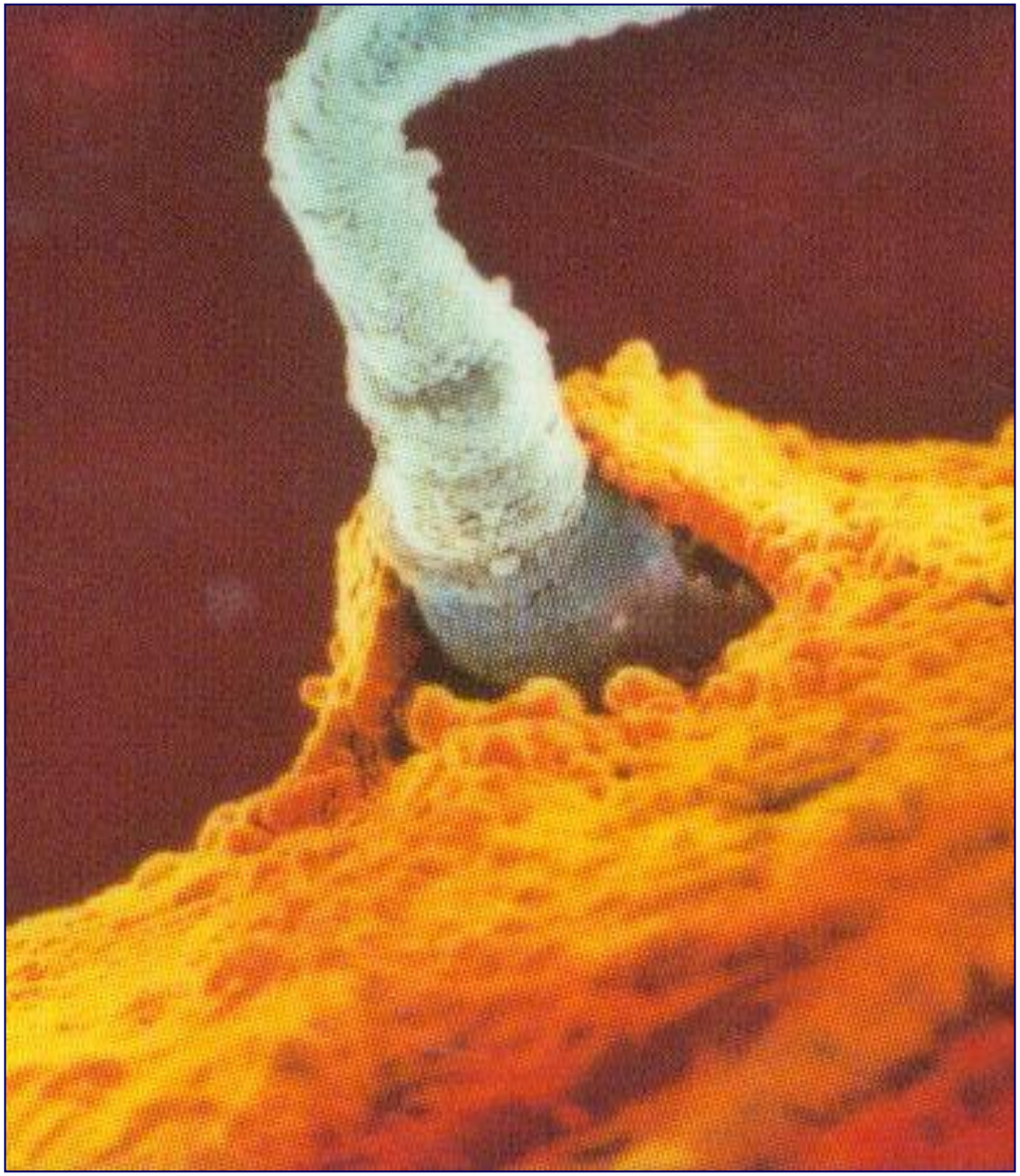
Table 4. Perinatal management and outcome in 18 antenatal TAPS cases detected at our center

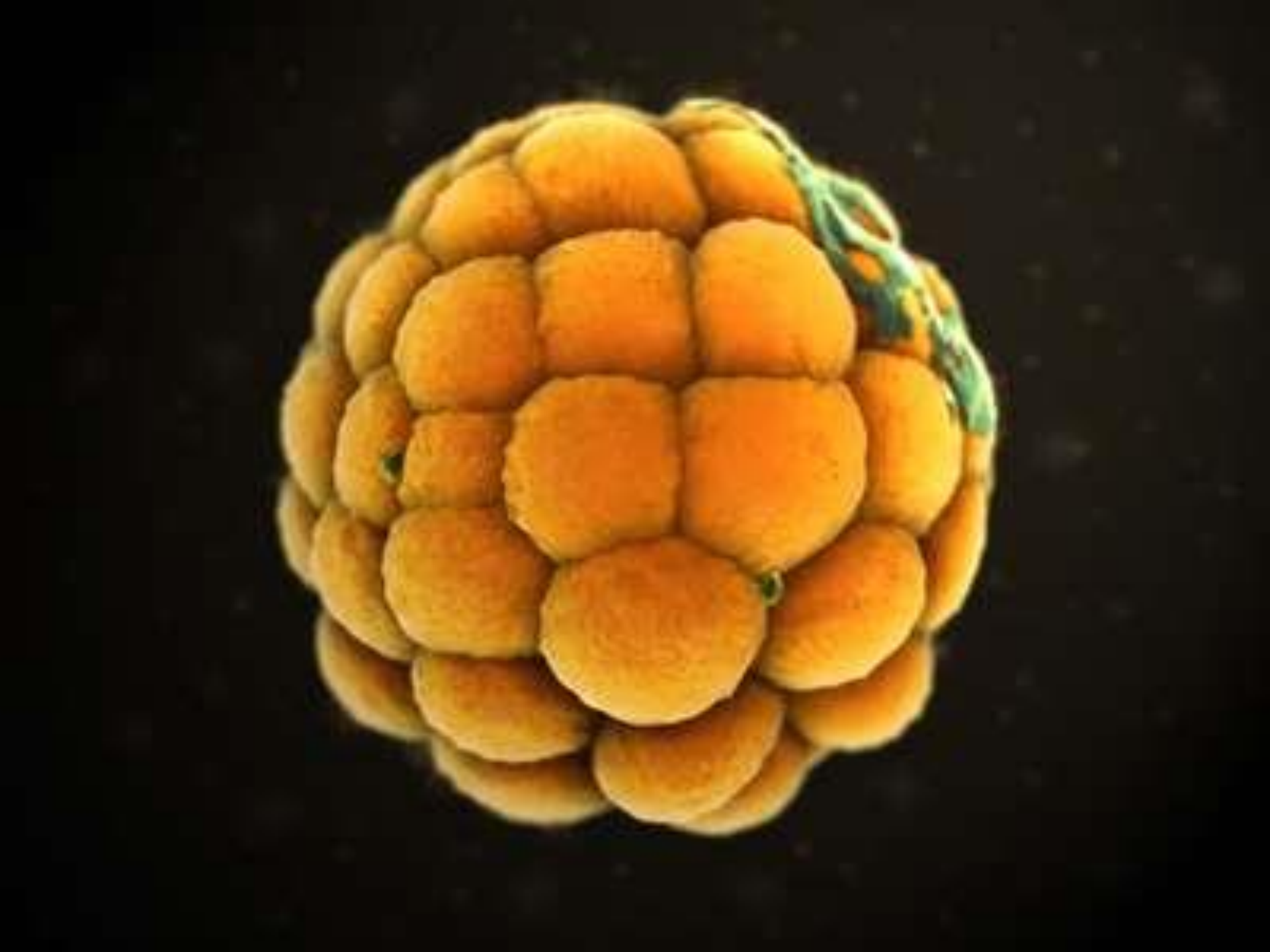
	Expectant management	IUT	IUT + laser	Laser	Selective feticide	TOP
Pregnancies, n	10	4[a]	1	1	1	1

Severe anemia
Hyper-viscosity

↓

Brain (other organs) damage





MZ ≡ embryonic accident

Pathogen

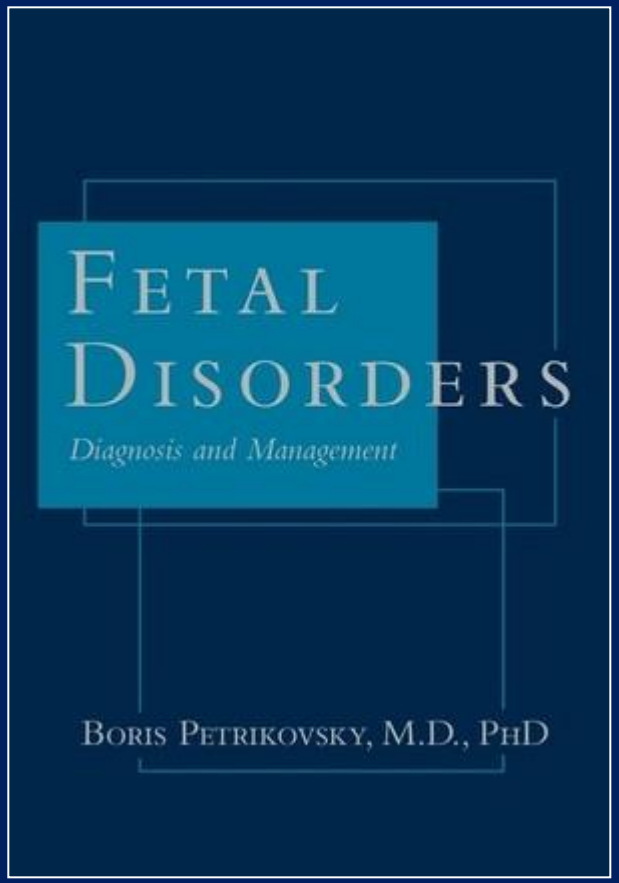
Zygotic splitting

Pathogen

Zygotic splitting

Malformations

Classification of malformations among multiples



Isaac Blickstein, 1999

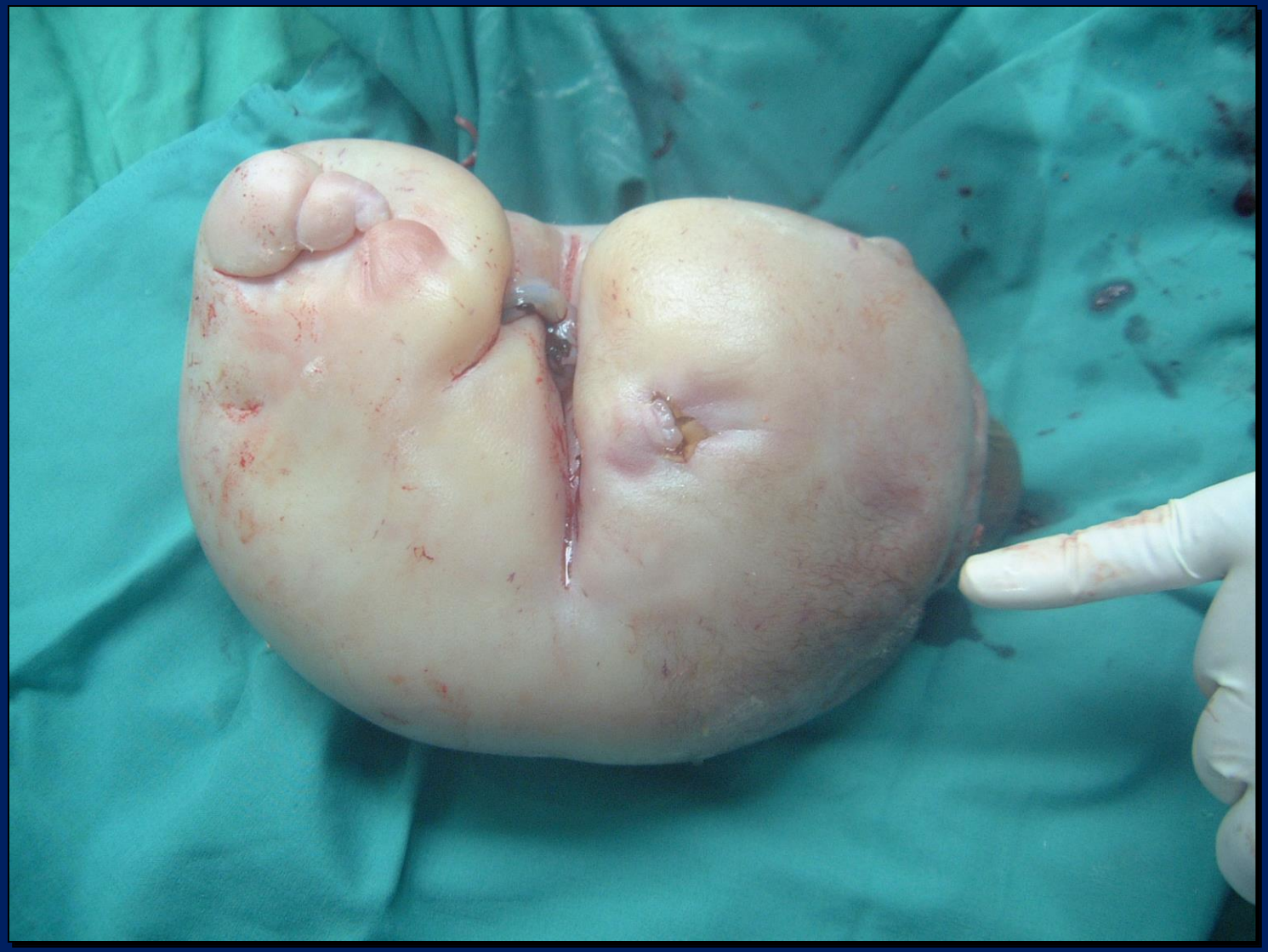
Classification of malformations among multiples

Category
1. Malformations more common in twins than in singletons

Classification of malformations among multiples

Category
2. Malformations unique to MZs


p 3
dB/C 2
slst Low
Opt:HRes
Rate:Max
-5
-10
-15
ACARDIAC
TWIN
TWIN



Classification of malformations among multiples

Category	
4. Deformation owing to intrauterine crowding	

Classification of malformations among multiples

Category
3. Placental malformations

Discordant

Table 1. Composite Definition of the Twin-Twin Transfusion Syndrome*

Criteria	Suggested cutoff values
I. Minor	
1) Sonographic	Inter-twin abdominal circumference difference >18 mm, poly/oligohydramnios, signs of monozygosity
2) Doppler velocimetry (umbilical arteries)	Inter-twin difference in S/D ratios >0.4
II. Major	
1) Transplacental shunt	
2) Birth weight	Inter-twin difference ≥15% (heavier twin = 100%)
3) Hemoglobin†	Inter-twin difference ≥5 g/dl

S/D = systolic/diastolic.
* Two major criteria or one minor and one major criterion are needed.
† Criteria may be found by antenatal funipuncture or postnatally.

the twin-twin transfusion syndrome. Wittmann et al

Size — sIUGR

AFI — TTTS, TOPS

Dopplers — TTTS, sIUGR, TAPS

Size — sIUGR

Hb — TAPS

Thank you