How to prepare for stage I?

Thomas Krasemann

Fetal cardiologist

- Council parents openly
- Give honest numbers- only 50% of operated children reach adulthood
- Speak not only about survival – quality of life is important
- Take the beliefs of ouders into account

It’s not just the heart

- The study cohort included 29 neonates with hypoplastic left heart syndrome and 13 neonates with transposition of the great arteries at a mean gestational age of 38.9 ± 1.1 weeks. Mean head circumference was 1 standard deviation below normal. The mean total maturation score for the cohort was 10.15 ± 0.94, significantly lower than reported normative data in infants without congenital heart defects, corresponding to a delay of 1 month in structural brain development.

Multidisciplinary meeting

- Cardiac anatomy, function, details
- Extracardiac issues
- Medications
- Ventilation details
- ECG
- Echocardiography
- Chest X-ray
- Cross sectional imaging as approriate
- Consider specific risks (TI, pulmonary venous return, position of the heart, extracardiac), specify strategy

Pre-operative condition

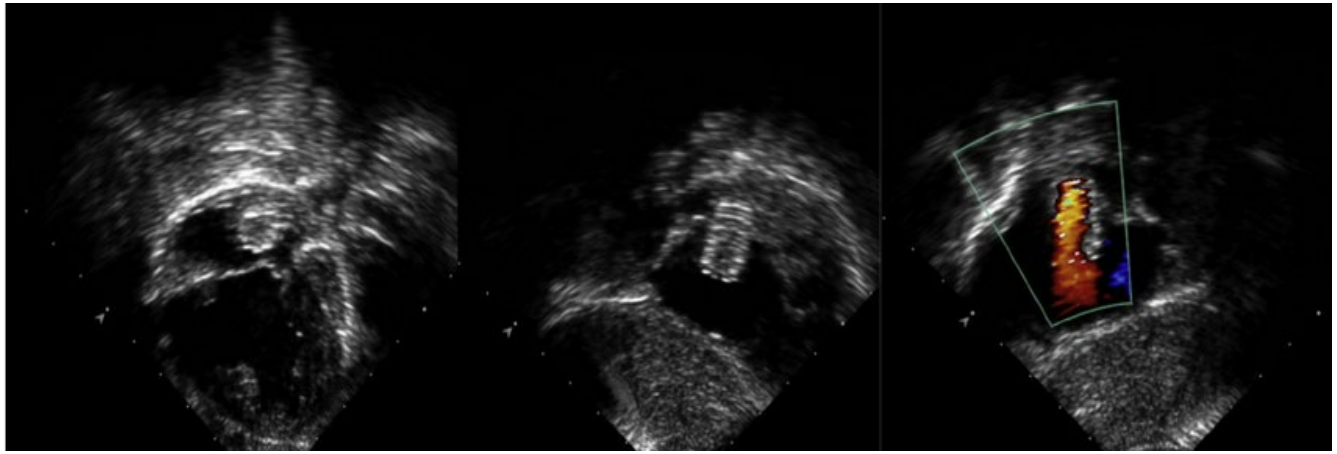
322 neonates, 50% BT, 50% Sano

		Attrition S1P and S2P						Attrition S2P and S3P	Interstage deaths (*n* = 32)	*P*-value
Birth weight (g)	3.2 (2.9–3.5)	3.2 (2.9–3.5)	3.2 (2.9–3.7)	0.40	**3.3 (2.9–3.5)**	**3.0 (2.6–3.4)**	**0.008**			
									22 (68.8)	0.89
									38.9 ± 1.6	0.61
Preterm	38 (11.8)	33 (16.3)	5 (13.2)	0.96	28 (10.7)	5 (21.7)	0.17	22 (10.2)	3 (9.4)	0.79
					3.5)	**3.0 (2.6–3.4)**	**0.008**	3.3 (2.9–3.5)	3.4 (2.9–3.7)	0.96
Low birth weight (<2500 g)	18 (5.6)	**12 (4.2)**	**6 (15.8)**	**0.002**						
						2 (8.7)	0.28	7 (3.2)	2 (6.3)	0.43
						2 (8.7)	0.41	11 (5.1)	1 (3.1)	0.60
Extracardiac anomalies	42 (13.0)	35 (12.3)	7 (18.4)	0.26	32 (12.3)	3 (13.0)	0.96	**24 (11.1)**	**8 (25.0)**	**0.036**
EFE	25 (7.8)	23 (8.1)	2 (5.3)	0.57	22 (8.4)	1 (4.3)	0.47	16 (7.4)	3 (9.4)	0.73
HLHS	271 (84.2)	237 (83.5)	34 (89.5)	0.34	215 (82.4)	22 (95.7)	0.100	177 (81.9)	27 (84.4)	0.74
AA/MA	69 (21.4)	56 (19.7)	13 (34.2)	0.068	49 (18.8)	7 (30.4)	0.34	43 (19.9)	5 (15.6)	0.58
AA/MS	67 (20.8)	60 (21.1)	7 (18.4)	0.55	55 (21.1)	5 (21.7)	0.77	45 (20.8)	4 (12.5)	0.27
AS/MA	29 (9.0)	25 (8.8)	4 (10.5)	0.83	24 (9.2)	1 (4.3)	0.34	21 (9.7)	2 (6.3)	0.54
Extracardiac anomalies	42 (13.0)	35 (12.3)	7 (18.4)	0.26	32 (12.3)	3 (13.0)	0.96	**24 (11.1)**	**8 (25.0)**	**0.036**
Other	4 (1.2)	4 (1.4)	0 (0)	0.46	3 (1.1)	1 (4.3)	0.21	2 (0.9)	0 (0)	0.59
TGA	29 (9.0)	28 (10.0)	1 (2.6)	0.16	28 (10.7)	0 (0)	0.095	27 (12.5)	1 (3.1)	0.11
DORV	18 (5.6)	14 (4.9)	4 (10.5)	0.14	12 (4.6)	2 (8.7)	0.40	10 (4.6)	1 (3.1)	0.69
CoA	84 (26.1)	77 (27.1)	7 (18.4)	0.30	70 (26.8)	7 (30.4)	0.74	58 (26.9)	10 (31.3)	0.65
Dextrocardia	4 (1.2)	3 (1.1)	1 (2.6)	0.39	3 (1.1)	0 (0)	0.60	2 (0.9)	1 (3.1)	0.30
Heterotaxy	1 (0.3)	1 (0.4)	0 (0)	0.72	1 (0.4)	0 (0)	0.76	1 (0.5)	0 (0)	0.70
Azygos continuation	1 (0.3)	1 (0.4)	0 (0)	0.72	1 (0.4)	0 (0)	0.76	1 (0.5)	0 (0)	0.70
Bilateral SVC	2 (0.6)	2 (0.7)	0 (0)	0.61	2 (0.8)	0 (0)	0.67	2 (0.9)	0 (0)	0.58
TAPVC	9 (2.8)	7 (2.5)	2 (5.3)	0.30	6 (2.3)	1 (4.3)	0.55	6 (2.8)	0 (0)	0.34
PLSVC	10 (3.1)	18 (6.3)	2 (5.3)	0.84	16 (6.1)	2 (8.7)	0.64	15 (6.9)	1 (3.1)	0.40
AA	138 (42.9)	118 (41.5)	20 (52.6)	0.21	105 (40.2)	13 (56.5)	0.14	89 (41.2)	9 (28.1)	0.14
AS	[illegible]	[illegible]	[illegible]	[illegible]	[illegible]	10 (43.5)	0.21	20 (9.3)	22 (68.8)	0.19
						9 (39.1)	0.16	56 (25.9)	7 (21.9)	0.61
MA	91 (28.3)	**74 (26.1)**	**17 (44.7)**	**0.017**		10 (47.6)	0.76	78 (36.1)	15 (46.9)	0.22
						0 (0)	0.11	20 (9.3)	3 (9.4)	0.93
AVVR	19 (5.9)	15 (5.3)	4 (10.5)	0.19	15 (5.7)	0 (0)	0.23	**10 (4.6)**	**4 (12.5)**	**0.025**
						11 (47.8)	0.091	63 (29.2)	5 (15.6)	0.15
Restrictive atrial septum	104 (32.3)	**84 (29.6)**	**20 (52.6)**	**0.003**		13.4 ± 1.6	0.38	**14.0 ± 2.3**	**15.5 ± 3.5**	**0.026**
						3.3 ± 1.9	0.12	4.1 ± 2.2	3.7 ± 1.7	0.45

Ono M, Kido T, Wallner M, Burri M, Lemmer J, Ewert P, Strbad M, Cleuziou J, Hager A, Hörer J. Preoperative risk factors influencing inter-stage mortality after the Norwood procedure. Interact Cardiovasc Thorac Surg. 2021 Jul 26;33(2):218-226

Interventionalist

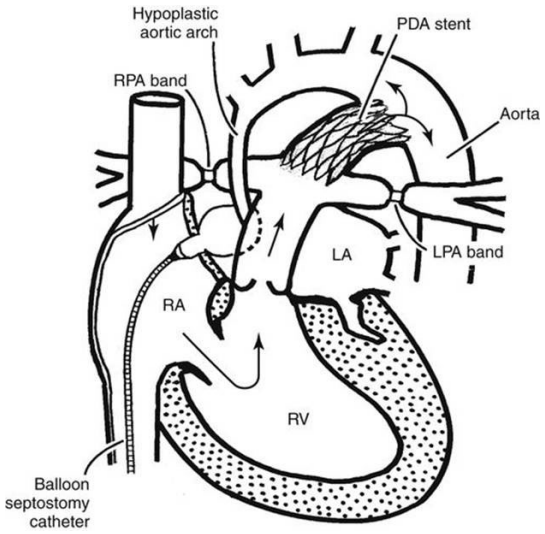
- Is the interatrial communication adequate?
- Is there need for a septostomy/atrial stenting?
- Which patients need an immediate postnatal intervention?



Hermuzi A, McBrien A, De Rita F, McCheyne A, Griselli M, O'Sullivan JJ, Hasan A, Crossland DS. Hybrid transatrial stent insertion for left atrial decompression in hypoplastic left heart syndrome with intact atrial septum. Catheter Cardiovasc Interv. 2016 Jan 1;87(1):109-16

Keep the baby alive!

- Prostaglandin 5-10mcg/kg/min – if concomittant CoA higher doses as necessary
- Monitor: BP on all extremities, pre- and postductal SaO2, fluid intake (restriction usually not necessary if patient is stable), brain oxigenation (NIRS), HR, lactate (?)
- Intubation/ventilation only if necessary, sedation, analgesia
- If high prostaglandin doses needed or side effects: ductal stenting?
- Balancing the circulations



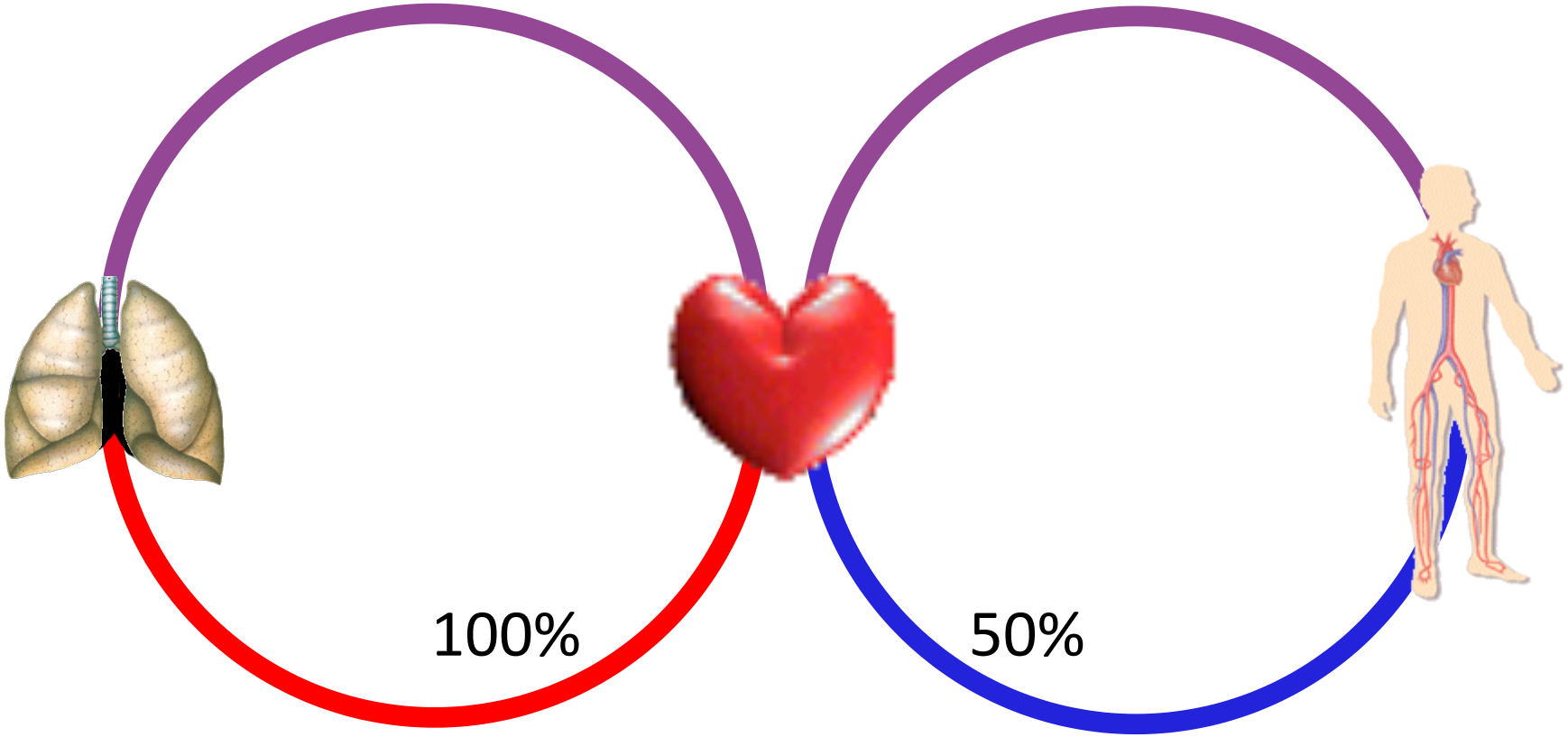
Alphonso N, Angelini A, Barron DJ, Bellsham-Revell H, Blom NA, Brown K, Davis D, Duncan D, Fedrigo M, Galletti L, Hehir D, Herberg U, Jacobs JP, Januszewska K, Karl TR; (Chaiman HLHS Guidelines Task Force); Malec E, Maruszewski B, Montgomerie J, Pizzaro C, Schranz D, Shillingford AJ, Simpson JM. Guidelines for the management of neonates and infants with hypoplastic left heart syndrome: The European Association for Cardio-Thoracic Surgery (EACTS) and the Association for European Paediatric and Congenital Cardiology (AEPC) Hypoplastic Left Heart Syndrome Guidelines Task Force. Eur J Cardiothorac Surg. 2020 Sep 1;58(3):416-499

Balancing the circulations

- Right ventricle pressure loaded and volume loaded
- Both pulmonary and systemic circulation supplied by RV
- Systemic circulation duct dependent - prostaglandin
- Postnatal transition – decreasing pulmonary vascular resistance
- Prostin – decreases pulmonary vascular resistance

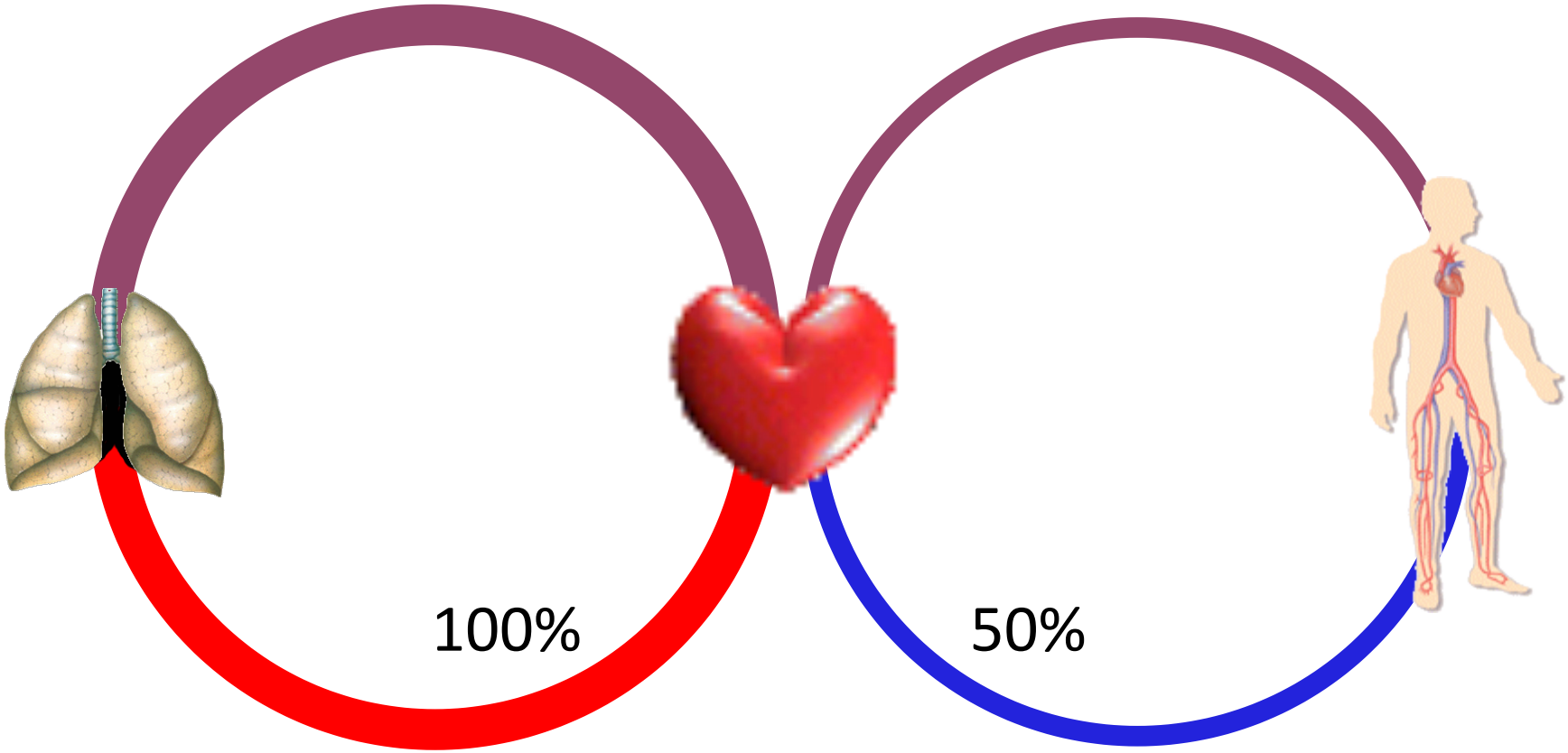
Balancing the circulations II

Qp:Qs 1:1



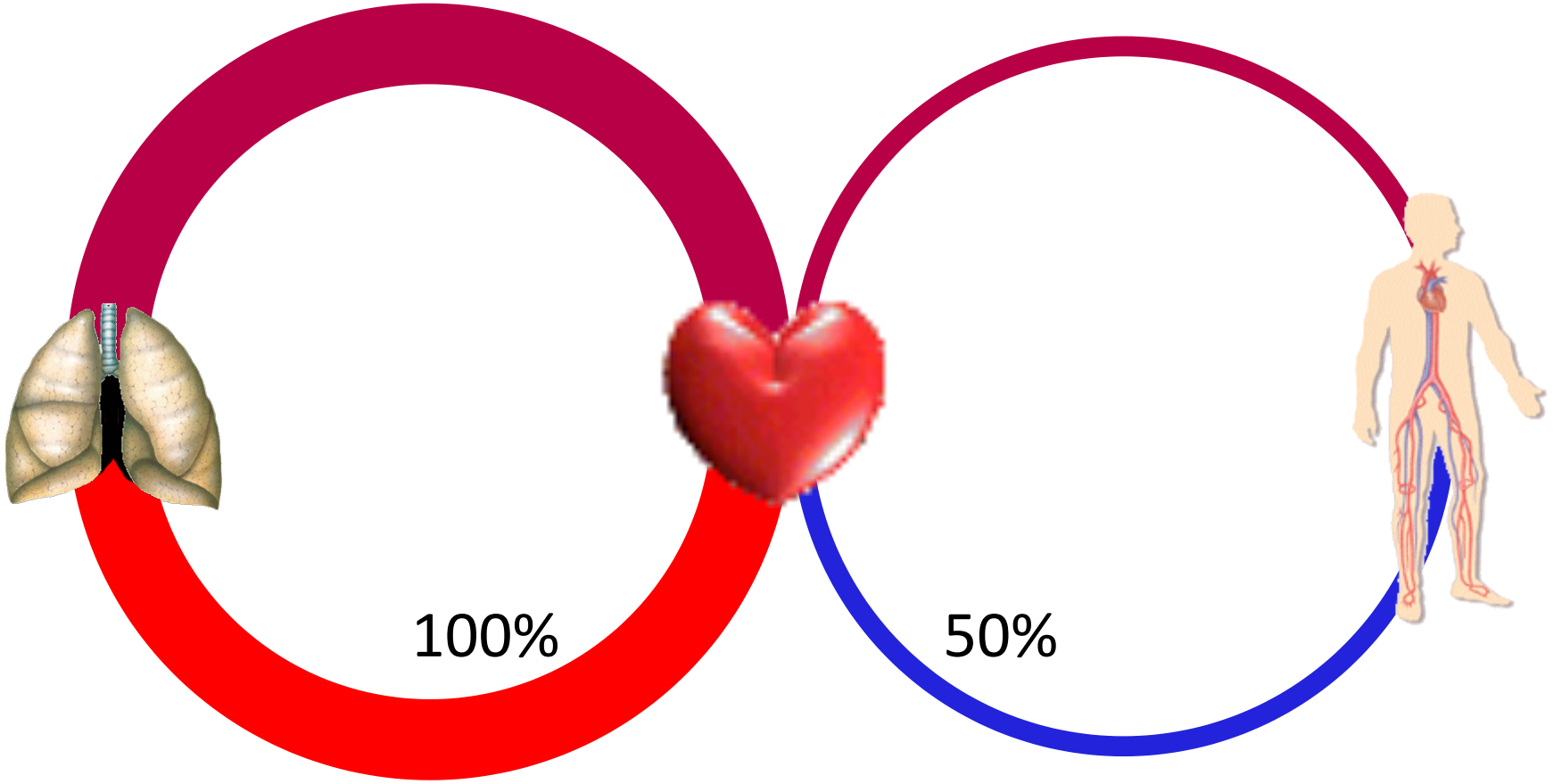
Balancing the circulations III

Qp:Qs 2:1



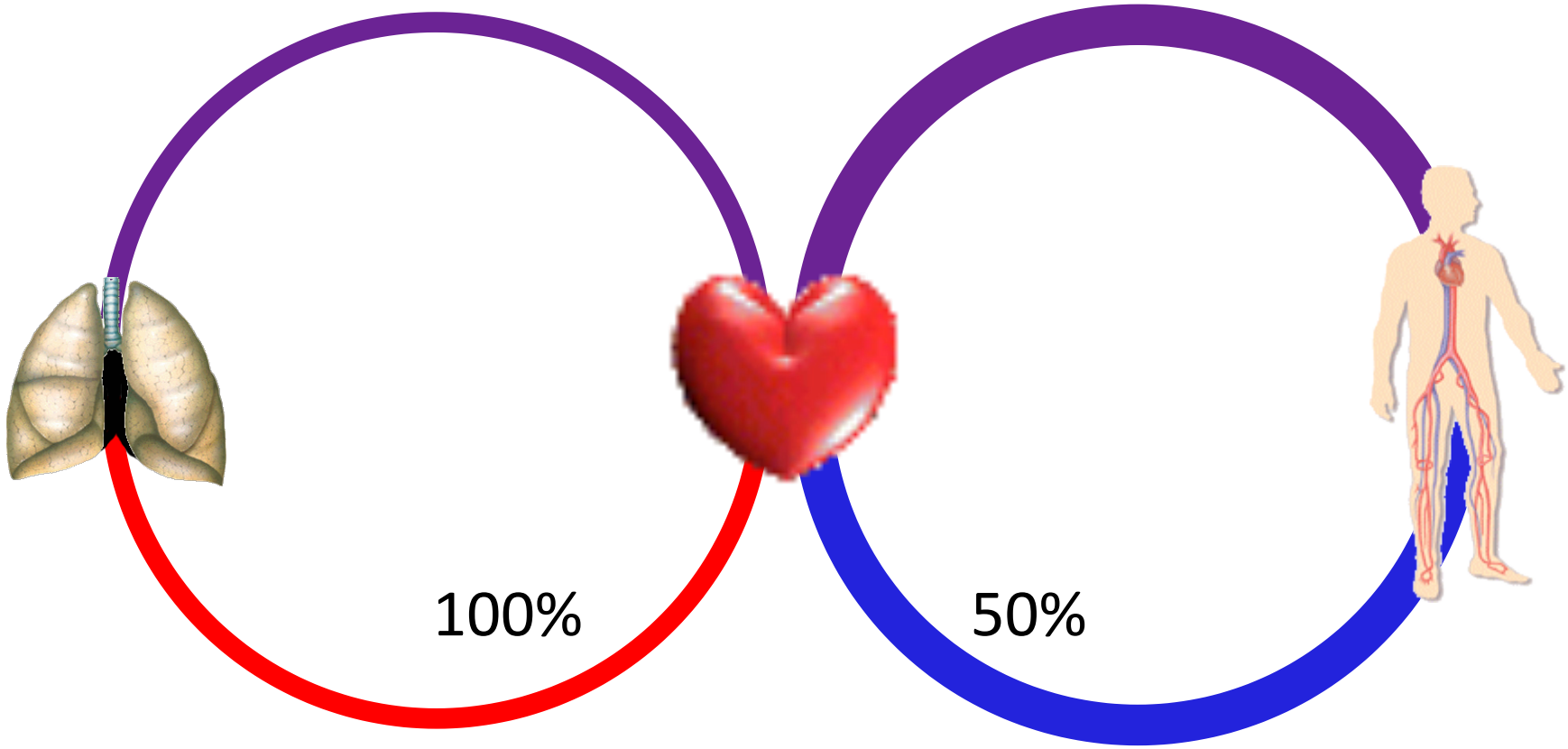
Balancing the circulations IV

Qp:Qs 4:1



Balancing the circulations V

Qp:Qs 1:2

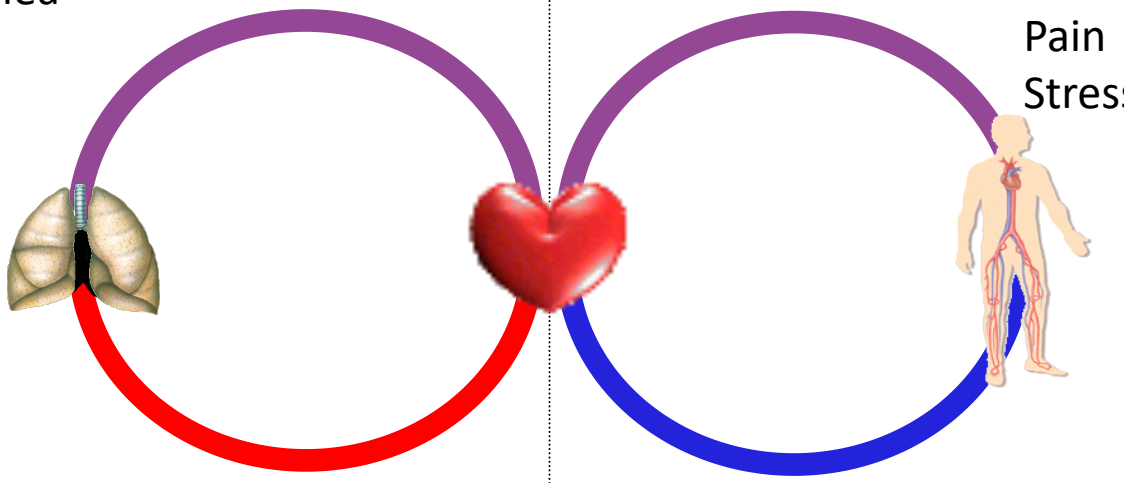


Balancing the circulations VI

Increase resistance

Acidosis
Hypercapnea
fiO2↓
PEEP↑

Adrenaline
Noradrenaline
Pain
Stress



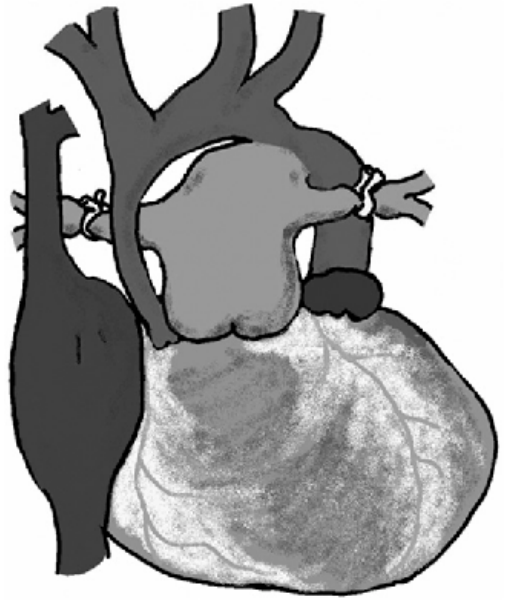
NO
Prostaglandin

Decrease resistance

Na-Nitroprusside
Phosphodiesterase-Inhibitors
Penoxybenzamine
Temperature↑
Beta blokkers

Balancing the circulations III

- Influenscing systemic vascular resistance:
  - Sodium-Nitroprussid?
  - Betablokkers?
  - Milrinone?
- Influencing pulmonary vascular resistance:
  - Hypoxic mix?
  - Metabolic? pH?
  - Mechanical? – bilateral bandings until child is stable/brains matured?



Hansen JH, Schlangen J, Voges I, Jung O, Wegmann A, Scheewe J, Kramer HH. Impact of afterload reduction strategies on regional tissue oxygenation after the Norwood procedure for hypoplastic left heart syndrome. Eur J Cardiothorac Surg. 2014 Feb;45(2):e13-9
Schranz D, Krasemann T. A word on netting of angiotensin-converting enzyme inhibitor therapy in hypoplastic left heart syndrome following stage-I. Cardiol Young. 2021 Aug;31(8):1323-1326.

Intensivist

- Try to understand the pathophysiology
- Listen to the Paediatric Cardiologist
- Do not miss any alarm symptoms- know them!
- Prepare the postoperative admission – NO?
- Make sure the pre- and postoperative handover to/from the anesthetist contains all necessary information
- Deliver the baby to theatres in optimal state

Optimal state?

- Well perfused
- Organ systems in perfect condition (except heart, maybe)
- Medication? Vasopressors? Effect on oxigenation-NIRS?

CONCLUSION

Go to:

Multivariate analyses demonstrate dopamine is independently associated with improvement in markers of systemic oxygen delivery 6 h after initiation. Dobutamine and milrinone were not independently associated with improvements in systemic oxygen delivery in this patient population.

Hendon E, Kane J, Golem GM, McBroom A, Flores S, Wong J, Villarreal EG, Loomba RS. Acute effects of vasoactive medications in patients with parallel circulation awaiting hybrid or Norwood procedure. Ann Pediatr Cardiol. 2022 Jan-Feb;15(1):34-40

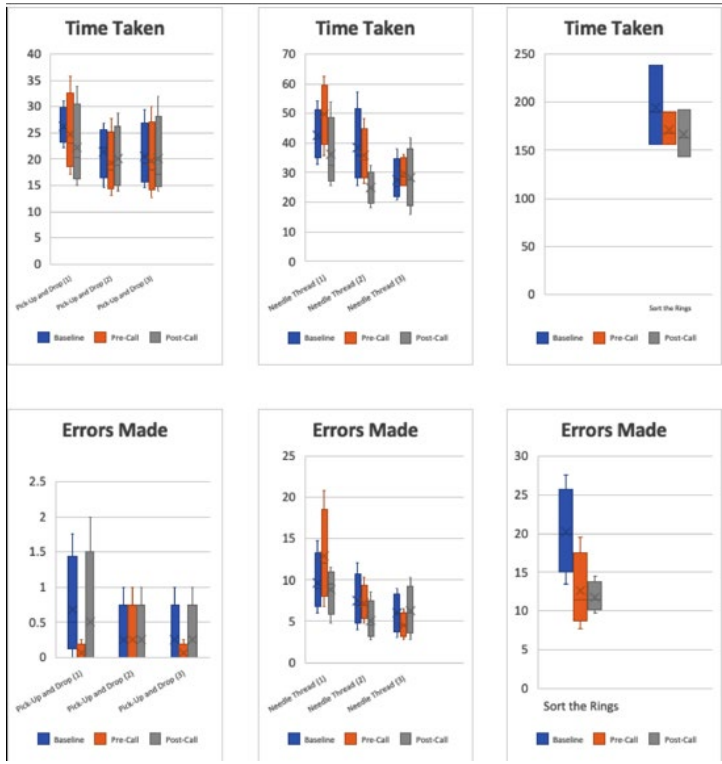
NIRS – preoperative brain oxigenation

- 43 patients
- Cerbral tissue oxigenation measured (NIRS) pre- and postoperative
- Full-scale, verbal, and performance IQ scores were evaluated with the Wechsler Preschool and Primary Scale of Intelligence at a median of 4.5 years (range, 3.5-6.8 years). Cognitive functions were assessed with the German Kognitiver Entwicklungstest für das Kindergartenalter (KET-KID)
- Preoperative low NIRS predictive for low values

Hansen JH, Rotermann I, Logoteta J, Jung O, Dütschke P, Scheewe J, Kramer HH. Neurodevelopmental outcome in hypoplastic left heart syndrome: Impact of perioperative cerebral tissue oxygenation of the Norwood procedure. J Thorac Cardiovasc Surg. 2016 May;151(5):1358-66

Surgeons

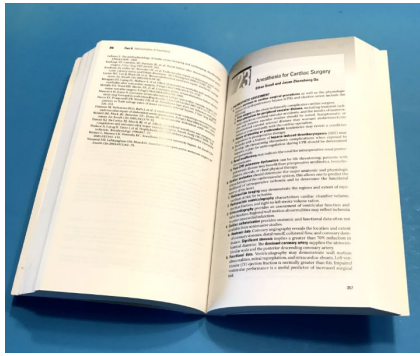
- Homograft ordered and available?
- Know the anatomy, as many details as possible: Was any 3D imaging available? Any unknowns you want to know in advance? Who knows them/can bring it into view?
- Look at the TOE imaging as if you don’t know the case- you might see something you haven’t before
- Go to bed early the day before
- Don’t drink too much coffee before starting the operation



Whelehan DF, Alexander M, Connelly TM, McEvoy C, Ridgway PF. Sleepy Surgeons: A Multi-Method Assessment of Sleep Deprivation and Performance in Surgery. J Surg Res. 2021 Dec;268:145-157

Anesthetist

- Sleep well the night before
- You may have some coffee before
- Understand preoperative circulation, thus: Check all physiological issues before: BP arm/leg, SaO2 (measured where)
- Consider different anesthetic agents, choose wisely
- This will not be a short operation. What do you want to read?



Zajonz T, Cupka P, Koerner C, Mann V, Menges T, Akintuerk H, Valeske K, Thul J, Schranz D, Mueller M. Anesthesia for bilateral pulmonary banding as part of hybrid stage I approach palliating neonates with hypoplastic left heart syndrome. Paediatr Anaesth. 2020 Jun;30(6):691-697

Alphonso N, Angelini A, Barron DJ, Bellsham-Revell H, Blom NA, Brown K, Davis D, Duncan D, Fedrigo M, Galletti L, Hehir D, Herberg U, Jacobs JP, Januszewska K, Karl TR; (Chaiman HLHS Guidelines Task Force); Malec E, Maruszewski B, Montgomerie J, Pizzaro C, Schranz D, Shillingford AJ, Simpson JM. Guidelines for the management of neonates and infants with hypoplastic left heart syndrome: The European Association for Cardio-Thoracic Surgery (EACTS) and the Association for European Paediatric and Congenital Cardiology (AEPC) Hypoplastic Left Heart Syndrome Guidelines Task Force. Eur J Cardiothorac Surg. 2020 Sep 1;58(3):416-499

Paediatric Cardiologist

- Know your place
- Don’t stand in the way
- Know the answer to all question
- Give advice as needed
- Think like a red blood cell- where will I go if…
- Are there extracardiac issues? Make them known.
- Brains of HLHS babies ca. 4 weeks less mature
- Prepare for potential exit-angiography

Timing of operation

- Too early?
- Too late?
- Which parameters determine the perfect timing?

Aims of the stage I operation

- Survival
- Independency from prostaglandin-infusion
- Balanced circulations
- Decision: uni- or biventricular?
- Quality of life on the long run

- Feasability?

Akintürk H, Yörüker U, Müller M, Schranz D. Hypoplastic Left Ventricle: Left Ventricular Recruitment With Hybrid Approach. World J Pediatr Congenit Heart Surg. 2022 Sep;13(5):637-644
Edwin F, Edwin AK, Palacios-Macedo A, Mamorare H, Yao N Management of Hypoplastic Left Heart Syndrome in Low-Resource Settings and the Ethics of Decision-Making. A. World J Pediatr Congenit Heart Surg. 2022 Sep;13(5):609-614