CONGENITAL HEART DEFECTS: NUTRITION CONSIDERATIONS FOR INFANTS

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DISCLOSURES

Amy R. Mahar created the content of this learning module and she is an employee of Nutricia North America

There are no additional disclosures or conflicts of interest to report

Nutricia North America supports the use of human milk wherever possible

SELF- STUDY LEARNING OBJECTIVES

1

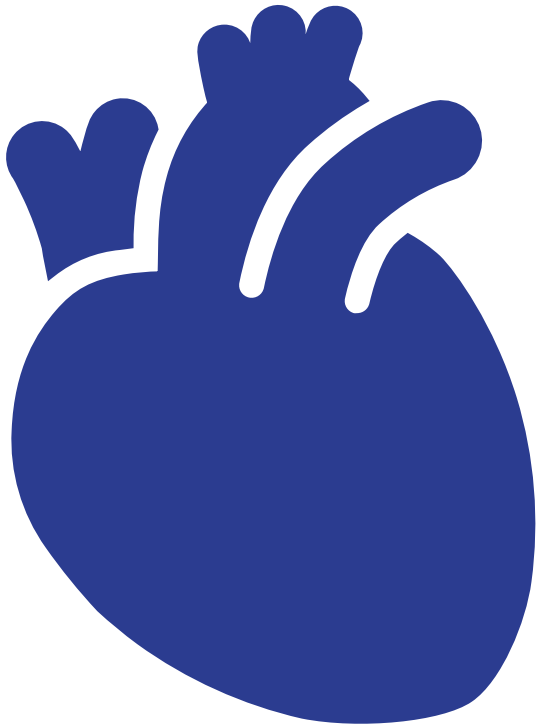
Define types of congenital heart disease (CHD)

2

Understand nutrition considerations associated with infants with CHD in the neonatal period

3

Determine nutrition interventions and care plans for infants with CHD using a novel energy- and nutrient-dense infant formula

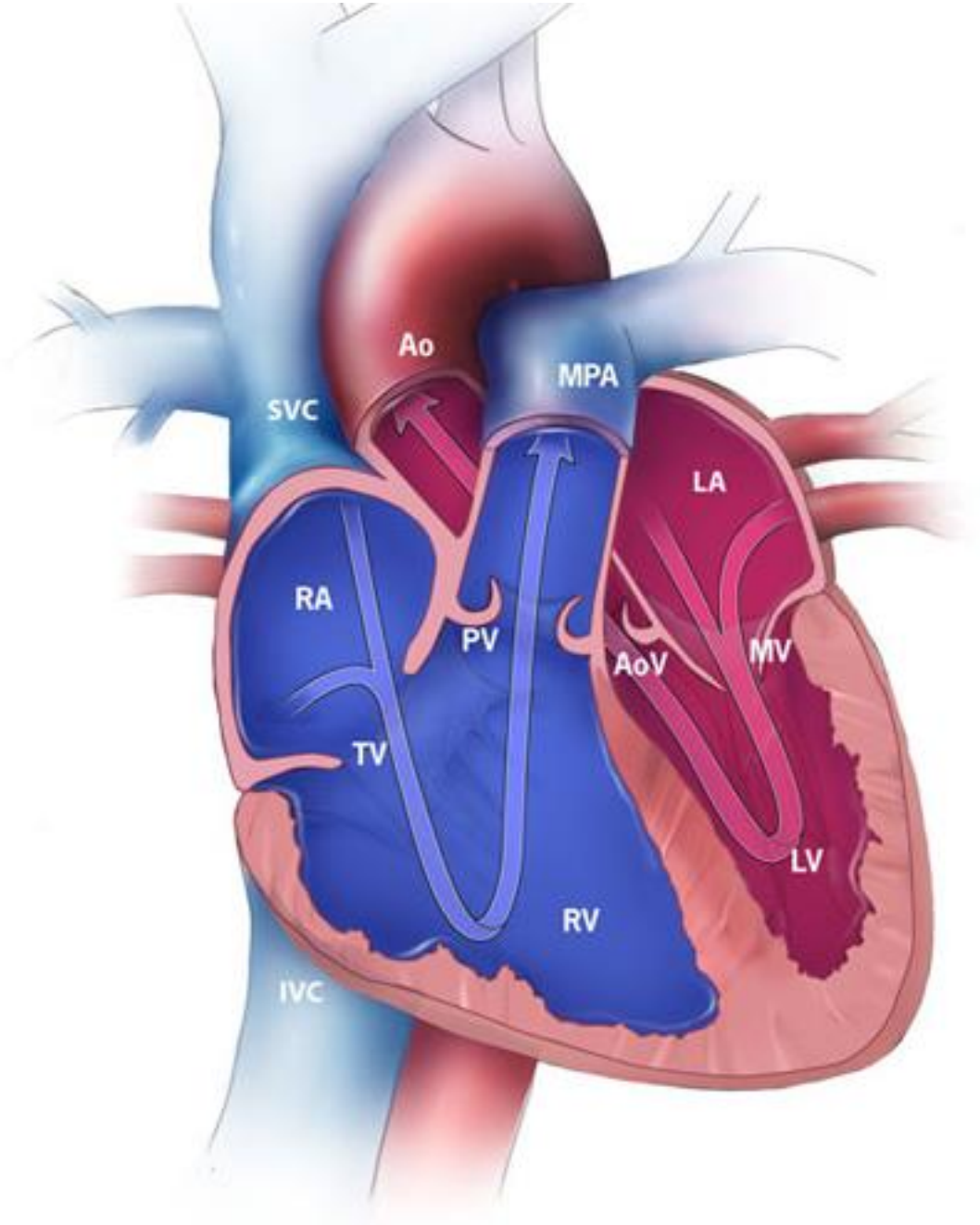


NORMAL ANATOMY OF THE HEART

NORMAL HEART

Function

- Oxygen depleted blood sent to heart→ lungs → heart →
- Oxygen rich blood sent to the body



RA. Right Atrium
RV. Right Ventricle
LA. Left Atrium
LV. Left Ventricle

SVC. Superior Vena Cava
IVC. Inferior Vena Cava
MPA. Main Pulmonary Artery
Ao. Aorta

TV. Tricuspid Valve
MV. Mitral Valve
PV. Pulmonary Valve
AoV. Aortic Valve

Photo credit: Centers for Disease

References: Centers for Disease Control and Prevention, National Center on Birth Defects and Developmental Disabilities
Puri et al Pediatrics in Review Oct 2017

NORMAL HEART
PATENT DUCTUS ARTERIOSUS (PDA)

What is it?

- Normal anatomy present in all infants
- Functionally closes within 24 hours after birth
- Patent (open) longer in preterm infants

Clinical Presentation and Treatment

- In an older infant, if unclosed, MD may hear a murmur
- Indomethacin/ibuprofen
- Surgery

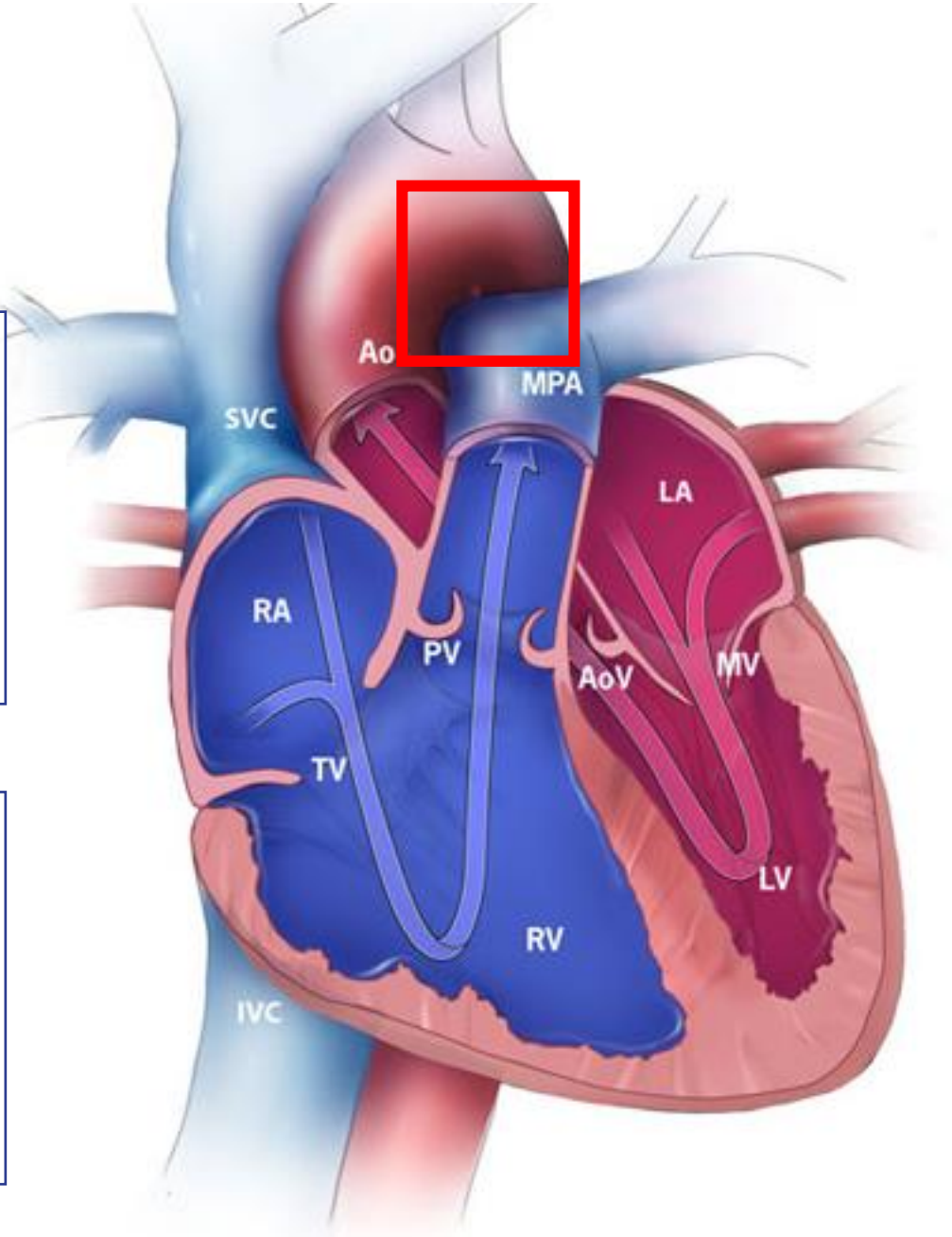


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Photo credit: Centers for Disease Control and Prevention, National Center on Birth Defects and Developmental Disabilities;
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Puri et al Pediatrics in Review Oct 2017

ACYANOTIC CONGENITAL HEART DISEASE

CYANOTIC CONGENITAL HEART DISEASE

WHAT'S THE DIFFERENCE?

TYPES OF CONGENITAL HEART DISEASE: EXAMPLES OF ACYANOTIC VS CYANOTIC DEFECTS

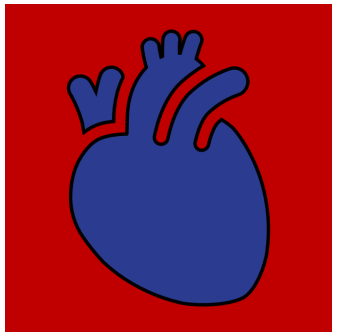
Acyanotic Heart Disease

Atrial septal defect

Ventricular septal defect

Atrioventricular septal defect (AVSD)

Coarctation of the aorta

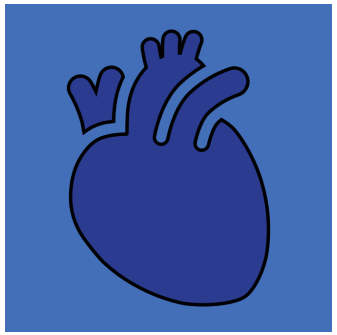


Cyanotic Heart Disease

Tetralogy of Fallot

Transposition of the great arteries

Hypoplastic left heart syndrome



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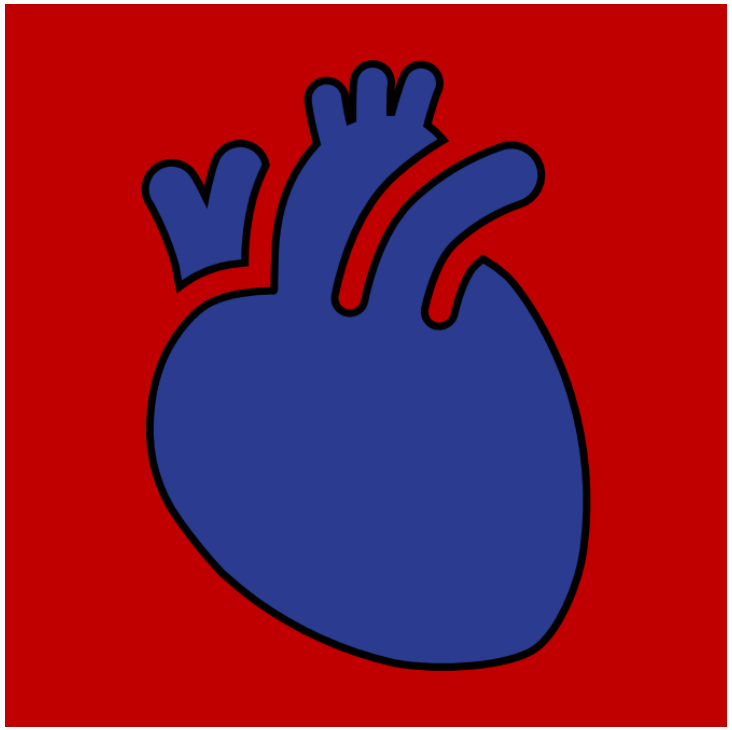
ACYANOTIC CONGENITAL HEART DISEASE



ACYANOTIC HEART DISEASE: ATRIAL SEPTAL DEFECT

What is it?

- A hole that divides the atria of the heart (upper chambers)

Prevalence

- 1 in 1,859 infants annually (CDC)
- 7-10% of CHD (Puri 2017)

Clinical Presentation

- Many infants do not have signs/symptoms at birth
- MD may hear murmur ~4-6 mos of age
- May tire with feeds due to extra work on the heart
- Sometimes not diagnosed until adulthood

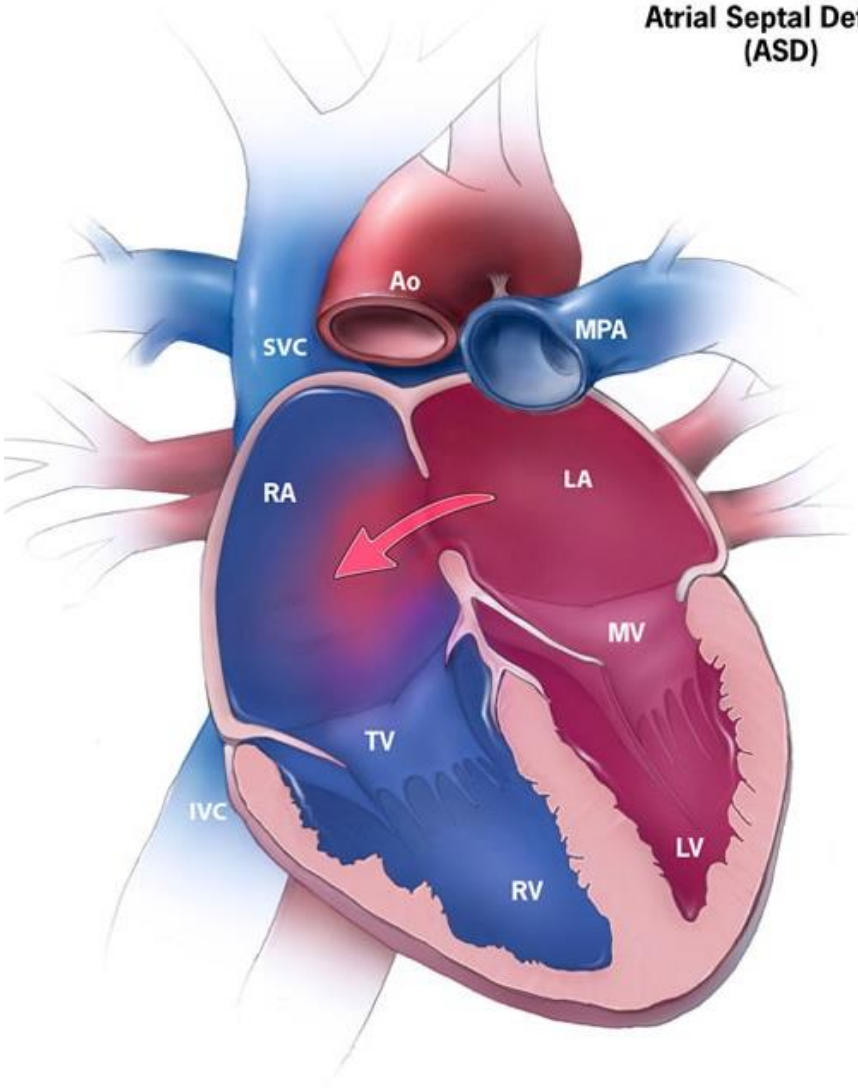


Photo credit: Centers for Disease Control and Prevention, National Center on Birth Defects and Developmental Disabilities;
References: Centers for Disease Control and Prevention, National Center on Birth Defects and Developmental Disabilities
Puri et al Pediatrics in Review Oct 2017
MD: medical doctor; physician
CDC: Centers for Disease Control

ACYANOTIC HEART DISEASE: VENTRICULAR SEPTAL DEFECT

What is it?

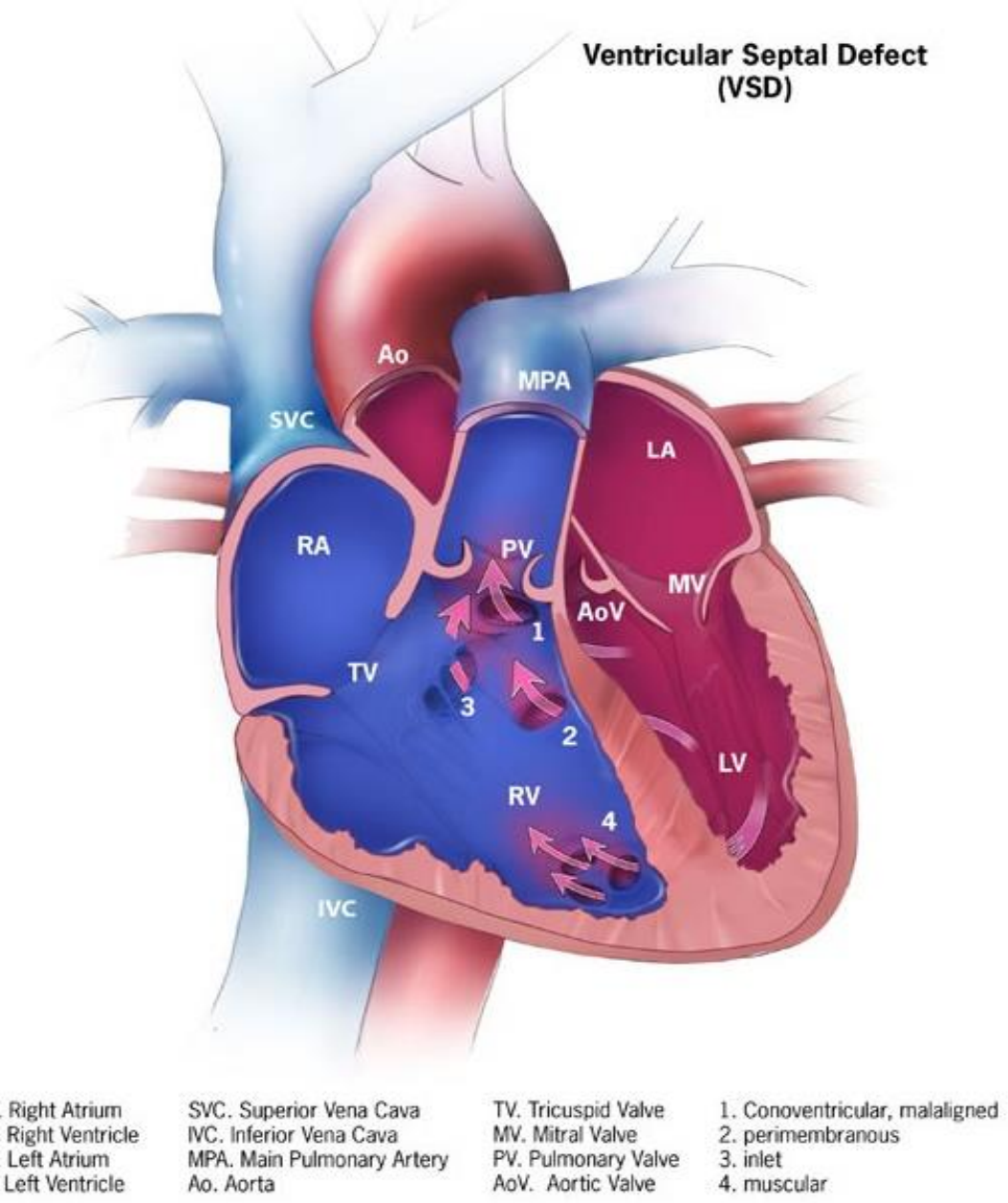
- Holes between the chambers of the R and L sides of the heart
- Can be in various locations

Prevalence

- 1 in every 240 births (CDC)
- 50-60% of CHD (Puri et al)

Clinical Presentation and treatment

- Size of VSD impacts symptoms
- MD may notice murmur
- Surgical repair
- Infants may tire with feeding, at risk of faltering growth
- May need up to 150 kcal/kg/d
- Monitor intake, weight gain



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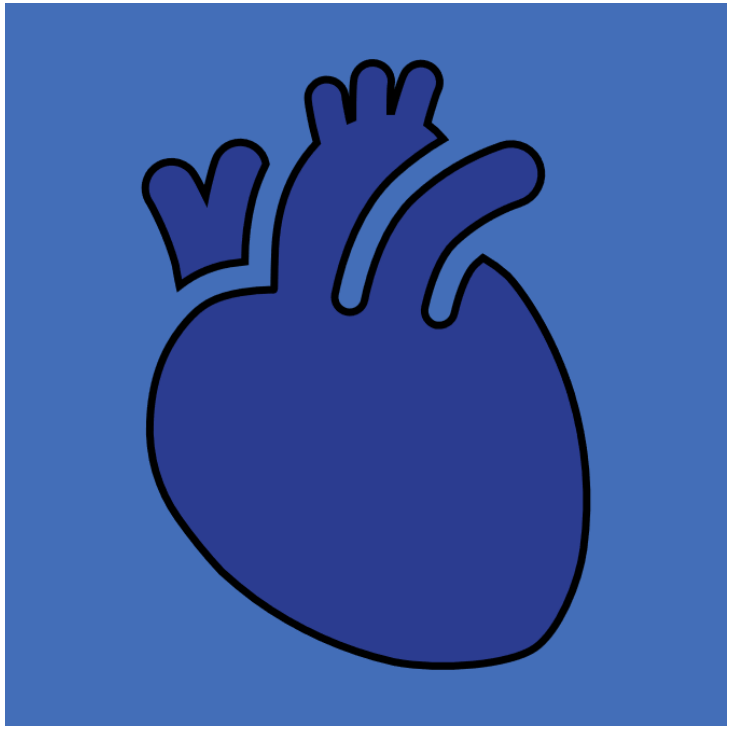
CYANOTIC CONGENITAL HEART DISEASE



CYANOTIC HEART DISEASE: TETRALOGY OF FALLOT

What is it?

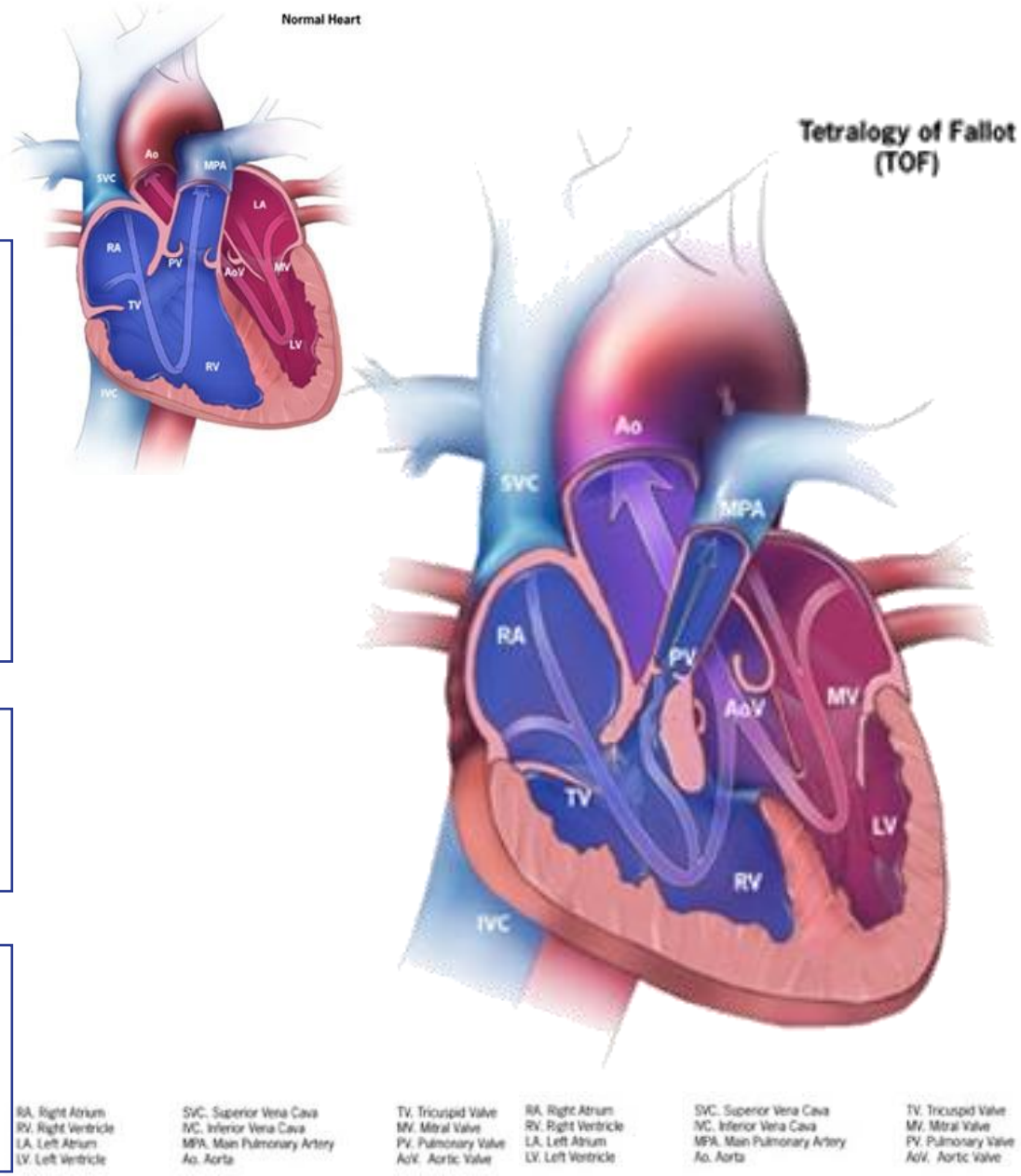
- Critical congenital heart defect
- Comprised of 4 defects:
  - VSD
  - A narrowing of the pulmonary valve and main pulmonary artery
  - Enlarged aortic valve
  - Ventricular hypertrophy

Prevalence

- About 1 in 2518 births
- Most common cyanotic CHD, about 5% of all CHD

Clinical Presentation

- MD might diagnose prenatally, or after birth
- Higher risk of endocarditis, arrhythmia
- Delayed growth and development



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Puri et al Pediatrics in Review Oct 2017

CYANOTIC HEART DISEASE: HYPOPLASTIC LEFT HEART SYNDROME

What is it?

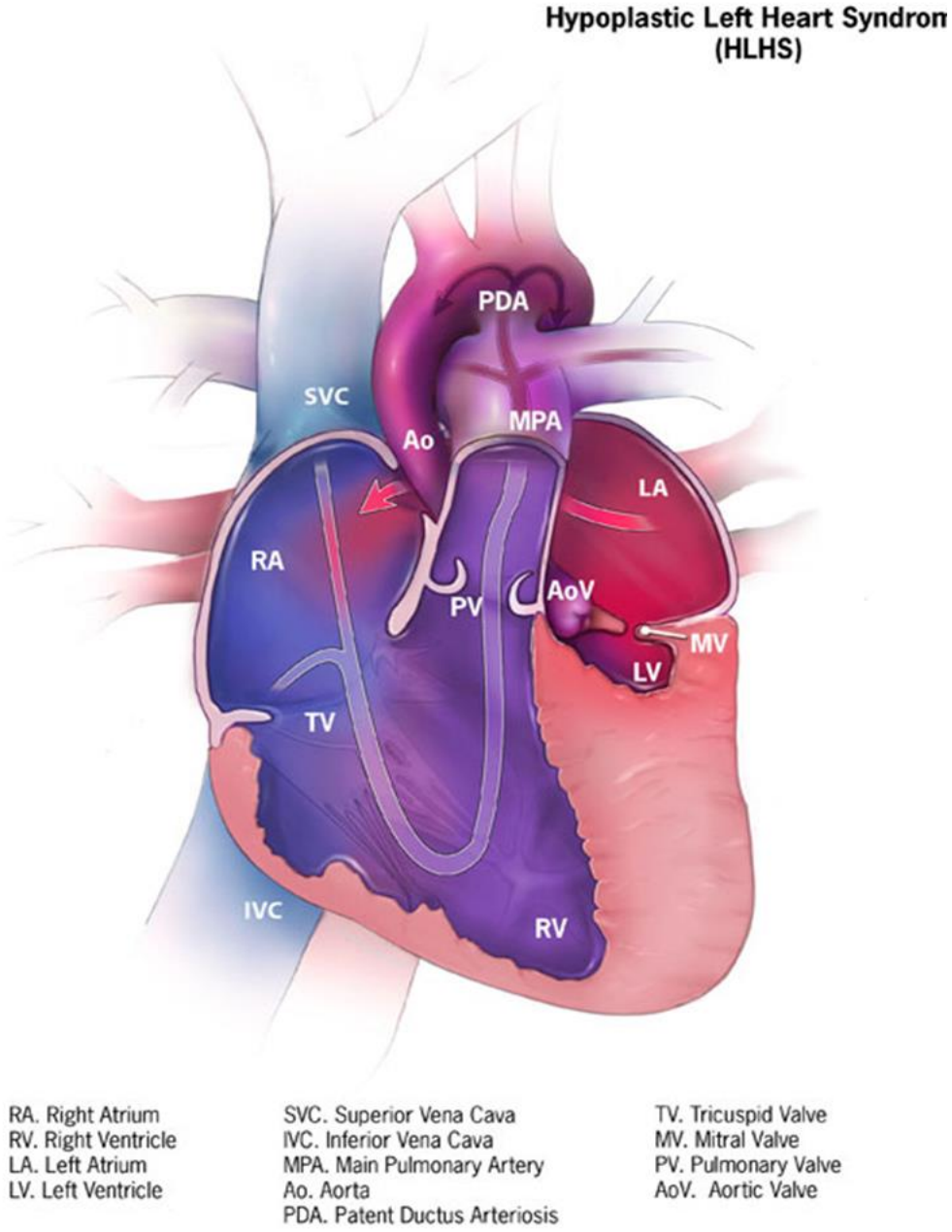
- Critical congenital heart disease
- Effects several structures on the L side of the heart, which do not fully form (e.g., left ventricle, mitral valves, aortic valves aren't formed properly)
- Affects normal blood flow through the heart

Prevalence

- 1 in 3,841 babies

Clinical Presentation

- PDA dependent (also known as ductal-dependent lesions)
- MD can hear murmur
- Shock, tachypnea, respiratory distress
- Multiple surgeries
- Tire with feeds



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NUTRITION CONSIDERATIONS FOR INFANTS WITH CHD

WHY IS NUTRITION SO IMPORTANT IN INFANTS WITH CHD?

Growth failure in CHD

- 15-41% develop malnutrition in the first few months of life[1]
- More common among neonates with cyanotic CHD
  - worsened by presence of pulmonary hypertension[1,2]
- Better growth among infants with CHD with standardized feeding protocols[3,4]

Lower HAZ and WAZ in neonates with CHD are associated with[5]:

- Increased mortality
- Increased infection
- Longer hospitalizations
- Adverse surgical outcomes (increased mortality, infection and length of stay)

HAZ = height-for-age z-score; WAZ = weight-for-age z-score.

1. Martini et al. Nutrients. 2021. 2. Blasquez et al. Eur J Clin Nutr. 2016. 3. Anderson et al. J Pediatr. 2021. 4. Lisanti et al. J Pediatr. 2021. 5. Ross et al. Am Heart J. 2020.

HOW DO THESE INFANTS WITH CHD DEVELOP GROWTH FAILURE?

Inadequate Intake[1,2,3]

- Gastroesophageal Reflux (GER)
- Tachypnea
- Poor feeding coordination
- Early satiety
- Endotracheal tube trauma/vocal cord injury
- Fluid restriction

Increased Energy Needs[1,2,3]

- Cardiopulmonary bypass (CPB) activates an inflammatory cascade [4]
- Energy requirements may increase by 30-50% in critically ill children [5]
- Increased work of breathing
- Resting energy expenditure may increase postoperatively [6,7,8,9]

Malabsorption[3,10]

- Altered systemic perfusion
- Intestinal edema
- Poor gut perfusion
- Splanchnic ischemia[11]

1) Shine AM et al, J Pediatr. 2021 2) Ross et al., Am Heart J 2020 3) Karpen HE Clin Perinatol. 2016 4) Floh AA Pediatr Crit Care Med. 2015 5) Skillman HE Curr Opin Crit Care. 2012 6) Nydegger, A Eur J Clin Nutr 63, 392–397 (2009) 7) De Wit B, . Pediatr. Crit. Care Med. 2010;11 (4): 496-501. 8) Mehta NM, Enteral Nutr. 2012;Ↄ6(6):685-692 9). Li Pediatr Crit Care Med. 2008;9(1):55-61 10)Martini et al. Nutrients. 2021 11) Kolkman et al World J Gastroentrol. 2008.

CALORIE AND PROTEIN NEEDS: NEONATES WITH CHD

ASPEN Guidelines for the Provision and Assessment of Nutrition Support Therapy in the Pediatric Critically Ill Patient

Recommended energy requirement for critically ill children:

"…We suggest that measured energy expenditure by IC be used to determine energy requirements and guide prescription of the daily energy goal."

Minimum recommended protein requirement for critically ill children:

"…we recommend a minimum protein intake of 1.5 g/kg/d. Protein intake higher than this threshold has been shown to prevent cumulative negative protein balance in RCTs."

IC = indirect calorimetry; RCT = randomized controlled trials

Mehta et al., 2017 Pediatr Crit Care 18(7)675-715

CALORIE AND PROTEIN NEEDS: NEONATES WITH CHD

ASPEN Guidelines for the Provision and Assessment of Nutrition Support Therapy in the Pediatric Critically Ill Patient

In the absence of indirect calorimetry ASPEN recommends the following equations without the addition of stress factors:

- The Schofield equation or
- The World Health Organization Food and Agriculture Organization's equation

IC = indirect calorimetry; RCT = randomized controlled trials

Mehta et al., 2017 Pediatr Crit Care 18(7)675-715
WHO/FAO equation: https://www.fao.org/3/y5686e/y5686e00.htm

CALORIE AND PROTEIN NEEDS: NEONATES WITH CHD

For step-down/acute care of infants with CHD

While nutrient recommendations vary depending on the infant's age, clinical status, other medical diagnoses, and care plan goals, Non-critically ill infants may require:

- **Calories:** 120-150 kcal/kg/d, sometimes up to 200 kcal/kg/d for catch up growth
- **Protein:** 3-3.5 g/kg/d
- **Fluid**: May be restricted per medical management, up to maintenance calculated by Holiday-Segar Methods

Roman. Nourishing Little Hearts: Nutritional Implications for Congenital Heart Disease 2011
Luca et al., Optimal Nutrition Parameters for Neonates and Infants with Congenital Heart Disease 2022

WHY IS NECROTIZING ENTEROCOLITIS (NEC) A FEEDING CONCERN?

CHD is a Risk Factor for NEC

- 10-100 times more common among infants with CHD
- Different pathophysiology among CHD vs preterm neonates
- Reduced intestinal perfusion and oxygen delivery
- Higher risk in ductal-dependent anomalies

McElhinney et al., Pediatrics 2000 Nov;106(5):1080-7
Scahill et.al., World J Pediatr Congenit Heart Surg. 2017 Jan;8(1):62-68
Giannone et al., Life Sciences 2008; 82(7-8):341-7
NEC: Necrotizing enterocolitis

EVIDENCE OF NEC AND FEEDING: NO INCREASED RISK

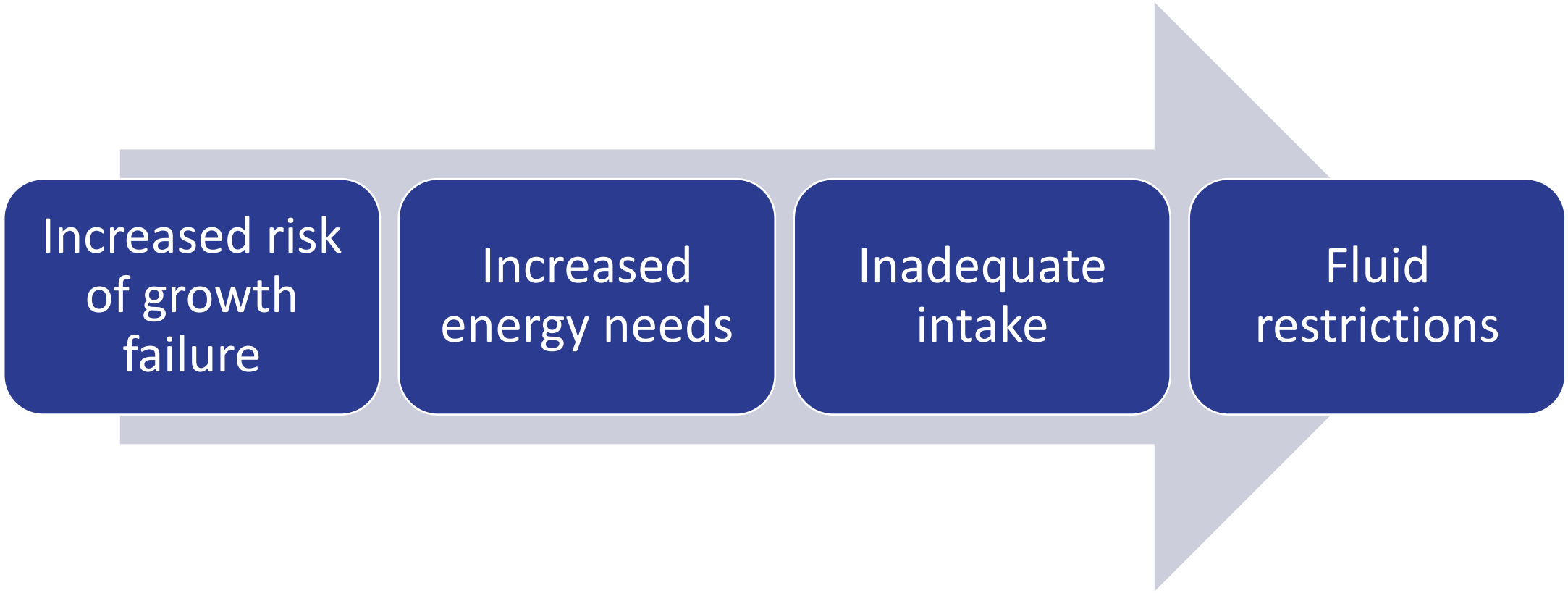
Scahill, et al., 2017 *World J Pediatr Congenit Heart* Surg 8(1):62-68		
n=130		
Infants ≤31 days of life requiring neonatal cardiac surgery		
61% with single ventricle physiology	55% PDA-dependent	61% (n=79) received pre-op feeds
No associations with preoperative feeding and NEC prevalence (n=130) Prematurity was only variable associated with NEC (P=0.03)		

Kataria-Hale et al., 2019 Hosp Pediatr 9(12):998-1006
Systematic review and meta-analysis evaluating pre-op feedings and ductal dependent heart disease
five retrospective cohort studies were included (high risk of bias)
No significant difference in NEC when comparing infants who were fed vs not fed Authors concluded "insufficient evidence to suggest pre-op feeding adversely influence rate of NEC, LOS or feeding intolerance"

Breast milk is associated with reduced NEC rates for infants with CHD- for further reading:

Cognata A, et al. "Human Milk Use in the Preoperative Period Is Associated with a Lower Risk for Necrotizing Enterocolitis in Neonates with Complex Congenital Heart Disease". J Pediatr. 2019;215:11-16.e2

NUTRITION CONSIDERATIONS FOR CHD INFANTS IN CASES OF:



WHAT IS AN ENDF?

- ENDF = energy- and nutrient-dense formula
- 30 kcal/ounce term infant formula
- 2.6 g protein/100 kcals
- Lower osmolality (<400mOsm/L)
- Well tolerated and supports growth
- Ready to feed and sterile
- Nutritionally complete
- Can be used to supplement breastmilk

Nutrition Management of Term Infants With Growth Failure
ASPEN 2022

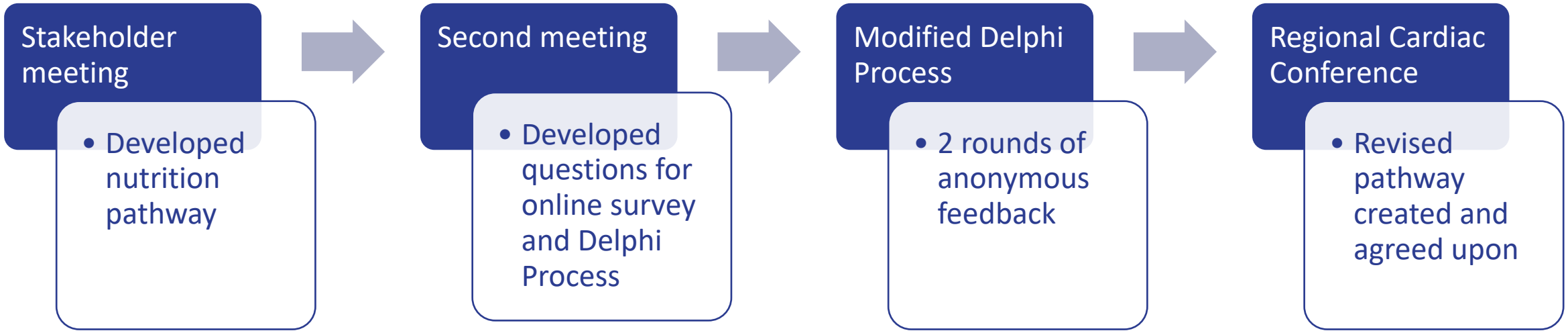
CARE PLANS FOR THE INFANT WITH CHD USING ENERGY- AND NUTRIENT-DENSE INFANT FORMULA (ENDF)

Improving growth of infants with congenital heart disease using a consensus-based nutritional pathway

Luise V. Marino, et al., . Clin Nutr. 2020;39(8):2455-2462

Improving growth of infants with congenital heart disease using a consensus-based nutritional pathway

Marino LV et al.,



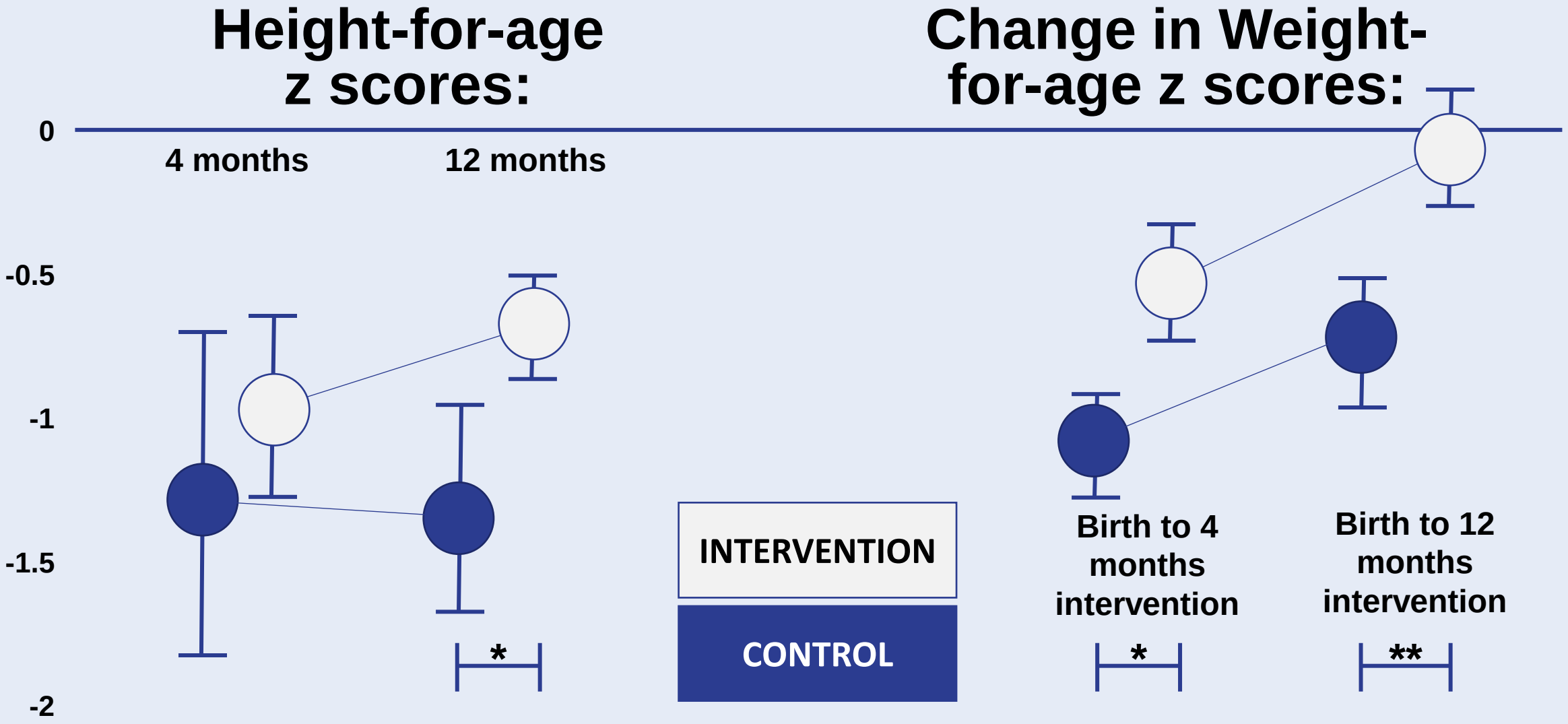
Improving growth of infants with congenital heart disease using a consensus-based nutritional pathway

Marino LV et al.,

Clinical Scenarios	Nutrition Care Plan	Energy Needs		Protein Needs		Fluids	Enteral nutrition sources
			kcal/kg/d	%EN	g/kg/d		
• Growing well • Can meet nutrient needs PO	**A**	Normal	90-100	No special considerations		Normal, e.g. 150 ml/kg/d	Breast milk or standard infant formula on demand
• Not growing well • CHD lesion + higher nutrition risk, feeding well • Finishes >75% of feeds PO • Fluid intake <120 mL/kg/d	**B**	+ ~10%	100-110	9-12	~2.5		Breast milk or standard infant formula + 30-80% of nutrition from ENDF
• Not growing • CHD lesion + higher nutrition risk • Needs NG/NJ tube • Fluid intake <100 mL/kg/d	**C**	+ ~10-20%	120-150	10-15	<4	May be restricted Check renal function	Breast milk or standard infant formula + 50-100% of nutrition from ENDF or as ON or NG feeds

CHD = congenital heart disease; %EN = Percent total energy; ENDF = energy- and nutrient-dense formula; NG = nasogastric; NJ = nasojejunal; ON = overnight; PO = by mouth

CONSENSUS-BASED NUTRITIONAL PATHWAY IMPROVED GROWTH OUTCOMES



Marino et al, Clin Nutr, 2020.

Effects and tolerance of protein and energy-enriched formula in infants following congenital heart surgery: a randomized controlled trial.

Cui Y, et al. JPEN J Parenteral Enteral Nutr. 2018;42:196-204.

Cui et al., ENDF Use Among Infants Post-Op for CHD Surgical Repair

NUTRICIA
LIFE-TRANSFORMING NUTRITION

Design

- Randomized, double-blind controlled trial
- 5-day intervention
- Fed continuously via NG tube
  - Start 12-24 hours post-op at 1 mL/kg/h (24 mL/kg/d), advance 1 mL/kg/h Q6H as tolerated
- Study formulas

Intervention (n = 26)	Control (n = 24)
ENDF • 1 kcal/mL • 2.6 g protein/ 100 kcal (10.4% PE)	SIF • 0.67 kcal/mL • 2.0 g protein/100 kcal (8% PE)

Study Population

- Term infants, 4 weeks -12 months old, post-op for CHD repair (biventricular repairs only)

Outcomes

1 – Nutrition status

- Macronutrient intake
- Daily 24-hr urinary urea nitrogen
- Biochemical

2 – Tolerance

- Emesis + stools
- GRV Q4H
- GI bleeding
- Gastric motor drugs (anti-emetic or anti-diarrheal agents)

3 – Outcomes

- Infections
- Length of stay

NG = naso-gastric; SIF = standard infant formula; ENDF = energy- and nutrient-dense formula; CHD = congenital heart defect. PE= protein energy 1. Cui Y et al. JPEN J Parenteral and Enteral Nutrition. 2018.

ENDF supported meeting nutrition goals sooner than standard infant formula with comparable tolerance

NUTRICIA
LIFE-TRANSFORMING NUTRITION

Results

1 – Nutritional Status

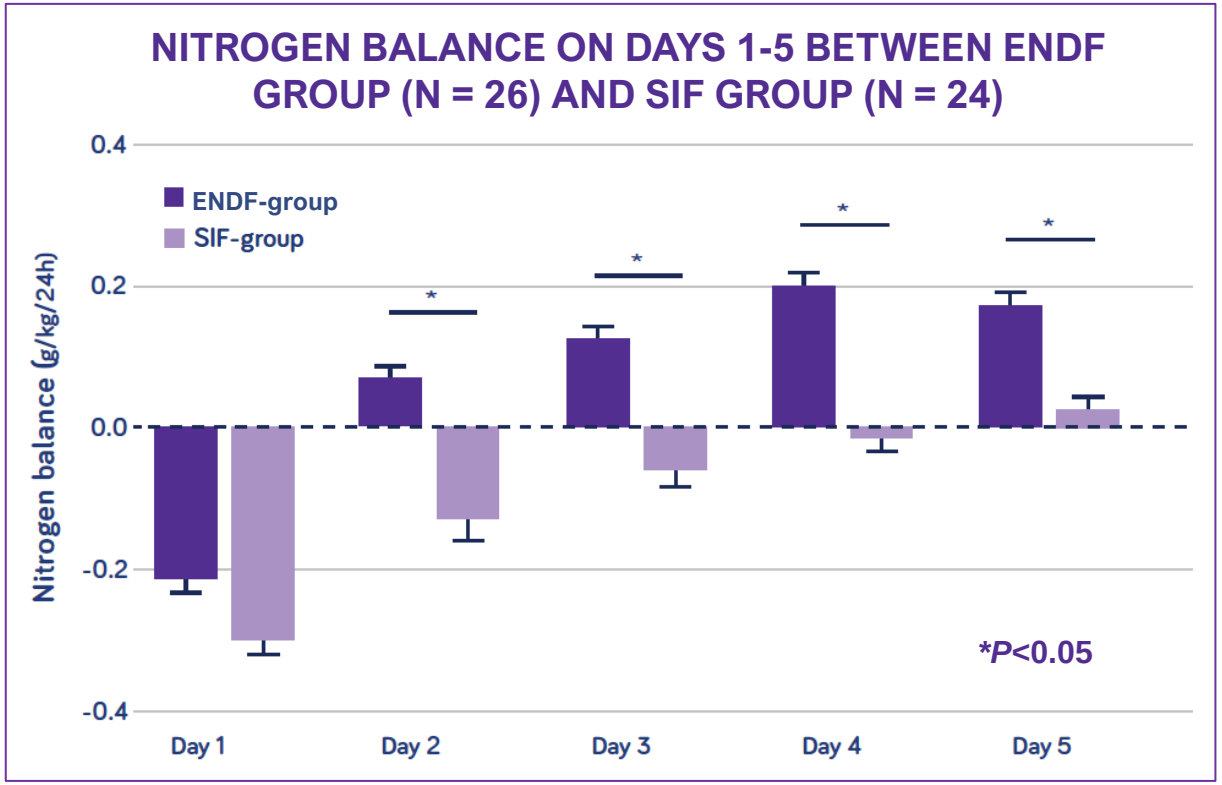
- ENDF group met adequate intake for energy and macronutrients before day 2
- SIF group only achieved adequate intake for CHO on days 2- 5

2 – Tolerance

- Increased stool frequency in the ENDF group on days 3 and 4 in the ENDF group
- Stool frequency and volume did not differ between groups overall

3 – Clinical outcomes

- No significant differences in infection, poor wound healing, prolonged ICU stay or prolonged ventilation time



CHO = carbohydrate; SIF = standard infant formula; ENDF = energy- and nutrient-dense formula. 1. Cui Y et al. JPEN J Parenteral and Enteral Nutrition. 2018.

Cui, et al. 2018.- Author conclusions

Nutritional Status

1) Achieve positive nitrogen balance in as little as 2 days
2) Promote anabolism compared to SIF

Tolerance

1) Equivalent tolerance to SIF
2) Well-tolerated in critically ill infants, including infants post-op for CHD repair

Safety

Safe in infants with growth failure due to CHD & other causes of growth failure

IN SUMMARY

CHD severity varies based on defect

Growth failure is common among infants with CHD

Providing adequate nutrition is challenging due to:

- Infants tire easily
- Feeding interruptions
- Potential malabsorption
- Increased energy needs

Infants with CHD have better growth if a feeding protocol is in place

Incorporating ENDF into care plans may improve growth among infants

ENDF is safe, well tolerated and supports catch up growth among infants with CHD

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THANK YOU

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