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Nutrition in congenital heart disease: consensus document

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Delphi technique

Abstract
Introduction: The prevalence of malnutrition among infants with congenital heart disease (CHD) is high. Early nutritional assessment and intervention contribute significantly to its treatment and improve outcomes. Our objective was to develop a consensus document for the nutritional assessment and management of infants with CHD.
Material and methods: We employed a modified Delphi technique. Based on the literature and clinical experience, a scientific committee prepared a list of statements that addressed the referral to paediatric nutrition units (PNUs), assessment, and nutritional management of infants with CHD. Specialists in paediatric cardiology and paediatric gastroenterology and nutrition evaluated the questionnaire in 2 rounds.

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Results: Thirty-two specialists participated. After two evaluation rounds, a consensus was reached for 150 out of 185 items (81%). Cardiac pathologies associated with a low and high nutritional risk and associated cardiac or extracardiac factors that carry a high nutritional risk were identified. The committee developed recommendations for assessment and follow-up by nutrition units and for the calculation of nutritional requirements, the type of nutrition and the route of administration. Particular attention was devoted to the need for intensive nutrition therapy in the preoperative period, the follow-up by the PNU during the postoperative period of patients who required preoperative nutritional care, and reassessment by the cardiologist in the case nutrition goals are not achieved.
Conclusions: These recommendations can be helpful for the early detection and referral of vulnerable patients, their evaluation and nutritional management and improving the prognosis of their CHD.

PALABRAS CLAVE
Cardiopatías congénitas;
Evaluación nutricional;
Estado nutricional;
Terapia nutricional;
Apoyo nutricional;
Trastornos nutricionales;
Técnica delfos

Asociación Española de Pediatría

Resumen
Introducción: La tasa de desnutrición entre los lactantes con cardiopatías congénitas (CC) es elevada. Una evaluación e intervención nutricional tempranas ayudan a su tratamiento y mejoran el pronóstico. El objetivo fue elaborar un documento de consenso para la evaluación y el tratamiento nutricional del lactante con CC.
Material y Métodos: Se utilizó una técnica Delphi modificada. En base a la literatura y a su experiencia clínica, un comité científico elaboró un listado de afirmaciones que abordaban la derivación a Unidades de Nutrición Pediátrica (UNP), la evaluación y el manejo nutricional de los lactantes con CC. Especialistas en cardiología pediátrica, y gastroenterología y nutrición pediátrica evaluaron el cuestionario en 2 rondas.
Resultados: Participaron 32 especialistas. Tras dos rondas de evaluación, se consensuaron 150 de 185 ítems (81%). Se determinaron patologías cardiacas de bajo y alto riesgo nutricional y factores asociados cardíacos o extracardíacos que confieren riesgo nutricional alto. Se elaboraron recomendaciones para la evaluación y seguimiento en unidades de nutrición y sobre el cálculo de los requerimientos nutricionales, el tipo de nutrición y la vía de administración. Se enfatiza la necesidad de un tratamiento nutricional intensivo en el preoperatorio, del seguimiento por la UNP en el postoperatorio cuando se haya necesitado intervención preoperatoria, y de la reevaluación por el cardiólogo cuando no se alcancen los objetivos nutricionales.
Conclusiones: Estas recomendaciones pueden ser de ayuda para la detección precoz y derivación temprana de población vulnerable, su evaluación y tratamiento nutricional y para mejorar el pronóstico de su CC.

Introduction

Congenital heart diseases (CHDs) constitute the most frequent group of congenital malformations, with an incidence of 8–12 cases per 1000 live births.[1] Approximately one third of affected children have haemodynamically significant CHD and are likely to require intervention (open surgery, catheterization or pharmacological treatment).[2] Most infants with CHD have normal weights at birth but develop nutritional and growth deficiencies in the first months of life, depending on the type of CHD.[3] Infants with mild CHD usually have normal growth and development,[4] but infants with moderate or severe CHD are at risk of nutritional problems that may affect development and growth and associated with an increased morbidity and mortality.[5] The prevalence of malnutrition in children with CHD ranges from 15% to 64%.[6]

The cause of these nutritional abnormalities is multifactorial, including both cardiac and extracardiac factors involving important aspects such as metabolic demand, energy expenditure, intake or intestinal absorption.[3] Early identification and prompt and appropriate intervention with frequent assessments are key to reduce the morbidity and mortality associated with malnutrition.[7] However, current nutritional assessment and management practices in infants with CHD are heterogeneous and vary between care settings and hospitals.[7,8] Guidelines for the assessment and nutrition of critically ill children have been proposed by scientific

societies in the fields of nutrition, paediatric cardiology or intensive care, but their recommendations are not homogeneous and do not focus on infants with CHD.[5,8–14]

The aim of this project was to establish consensus recommendations for the referral to the paediatric nutrition unit (PNU), the assessment and estimation of nutritional needs and nutrition therapy of infants with CHD in a multidisciplinary approach.

Material and methods

We used a modified Delphi method following the RAND/UCLA recommendations.[15,16] The first step was to gather a scientific committee comprised of 10 members of the Spanish societies of gastroenterology, hepatology and nutrition (Sociedad Española de Gastroenterología, Hepatología y Nutrición Pediátrica, SEGHNP) and paediatric cardiology and congenital heart diseases (Sociedad Española de Cardiología Pediátrica y Cardiopatías Congénitas, SECPCC). Following a literature search, the committee drafted statements or proposals regarding controversial aspects of referral, assessment, need estimation and nutrition therapy of infants with CHD.

Thirty-two panellists were invited to participate in the study. They were specialists in paediatric nutrition and paediatric cardiology with years of experience in the field of CHD. All participated in the 2 Delphi rounds.

Literature search

We conducted a literature search in the PubMed database using the following terms: *heart defects*, *congenital*; *nutritional status*; *nutrition therapy*; *nutritional support*; *nutrition assessment*; *enteral nutrition*; *parenteral nutrition*; *nutrition disorders*; *malnutrition*. We made a qualitative assessment of the literature, selecting articles in Spanish and English dealing with the management of CHD in children with particular emphasis on clinical practice guidelines (CPGs) and recent reviews. We also performed searches in the websites of the following scientific societies to find CPGs: Asociación Española de Pediatría (AEP), SECPCC, Sociedad Española de Nutrición Clínica y Metabolismo (SENPE), SEGHNP, European Society for Clinical Nutrition and Metabolism (ESPEN), American Society for Parenteral and Enteral Nutrition (ASPEN), European Society of Paediatric and Neonatal Intensive Care (ESPNIC), Pediatric Cardiac Intensive Care Society (PCICS) and National Pediatric Cardiology Quality Improvement Collaborative (NPC-QIC).

Panellist selection

In a second phase, a panel of experts on paediatric cardiology and paediatric gastroenterology and nutrition was selected for the task of evaluating proposed statements. The panellists were selected by the scientific societies they were affiliated with on the basis of their expertise and knowledge of or involvement in CHD.

Evaluation of proposed statements and consensus criteria

The questionnaire was sent to the panellists to be completed online in two rounds. Between rounds, it was possible to edit confusing statements. Panellists rated statements by means of a 9-point Likert scale (1: completely disagree; 9: completely agree). Ratings were grouped into 3 categories (1–3: disagree; 4—6: neither agree nor disagree; 7–9: agree).

To consider that consensus had been reached in agreeing or disagreeing with an item, the median of the ratings by the panellists had to be within the 7–9 points or the 1–3 points range, respectively. In addition, the number of panellists that voted outside the 1–3 or 7–9 ranges had to be less than 1/3 of the total and the interquartile range had to be less than 4. Items for which a consensus was not reached in the first round were subjected to a second evaluation round, the results of which were analysed with the same method used in the first round. Panellists were informed of the results of the first round before participating in the second round. The results are summarised in tables available in the supplemental material (Appendix B, Tables S1–S4).

Results

In the first round, the panel reached a consensus in agreeing with 143 items, and in the second round, it reached a consensus in agreeing with an additional 7 items. After the two Delphi rounds, a consensus was reached on 150 of the 185 proposed items (81%), in agreement in every case (Appendix B, Tables S1–S4).

General aspects of nutrition in children with CHD

Panellists considered that patients with CHD were a group with particular nutritional risk requiring specific assessment, and whose nutritional needs depend on the type of cardiac lesions and clinical significance. They also agreed that children with CHD require periodic assessments of their nutritional status in order to detect delays in growth early and allow intervention, and that recommendations for the nutritional management of these children need to be established to improve nutritional status before surgery (Appendix B, Table S1).

Identification and referral of vulnerable individuals

In this set of proposals, panellists reached a consensus on the CHDs and associated factors associated with a low or high nutritional risk at diagnosis (Tables 1–2) and the criteria for referral to nutrition services (Table 3). The panel considered that patent ductus arteriosus (in the case of early surgical repair), atrial septal defect and pulmonary stenosis carry a low nutritional risk. The diseases considered to carry a high nutritional risk were complex CHDs and moderate or severe cardiac shunts, including aortopulmonary window.

The following cardiac factors were considered to be associated with a high nutritional risk: pressure and/or volume

Table 1 Congenital heart diseases associated with low or high nutritional risk at diagnosis.

Low nutritional risk
Patent ductus arteriosus (early surgery)
Atrial septal defect
Pulmonary stenosis
No consensus: cor triatriatum and total anomalous pulmonary venous return
High nutritional risk
Pulmonary atresia
Tetralogy of Fallot
Atrial septal defect (severe lesion)
Transposition of the great vessels
Ventricular septal defect (moderate or severe)
Atrioventricular septal defect
Hypoplastic left heart syndrome
Truncus arteriosus
Aortopulmonary window
Patent ductus arteriosus (large or with late surgery)
Tricuspid atresia
Ebstein anomaly
Double outlet right ventricle
No consensus: coarctation of the aorta and partial anomalous pulmonary venous return

Table 2 Factors associated with congenital heart diseases that carry a high nutritional risk.

Associated cardiac factors
Right or left heart pressure and/or volume overload
Myocardial dysfunction
Congestive heart failure
Chronic hypoxaemia
Pulmonary hypertension
Increased pulmonary blood flow
Associated extracardiac factors
Malabsorption
Specific nutrient deficiency
Congenital heart disease as part of an identifiable syndrome
Certain chromosomal anomalies (e.g., trisomy 21, 18 or 13)
Associated extracardiac anomalies (e.g., intestinal atresia, congenital chylothorax...)
Intrauterine growth restriction and preterm birth
Recurrent respiratory infection
Adverse psychosocial conditions
Gastro-oesophageal reflux disease
Other associated clinical conditions
Inadequate energy intake
Increased energy expenditure
Inadequate weight or length gain (e.g., weight gain >10 g/kg/day)
Vomits most of the food
Inadequate intake amount
Fatigue during feedings
Requires nutritional support (supplementation, nasogastric tube feeding, etc.)

overload, myocardial dysfunction, congestive heart failure, chronic hypoxaemia, pulmonary hypertension and pulmonary overflow. Among the extracardiac factors and other clinical conditions that could be considered to be associated with a high nutritional risk, the panel reached a consensus for intestinal malabsorption, intrauterine growth restriction, prematurity, inadequate energy intake or increased energy expenditure, among others.

As regards the criteria for referral to the PNU, the panel agreed that referral is required in infants with CHD considered to carry a high nutritional risk (Table 1), with any risk factor (cardiac or extracardiac) or clinical condition associated with high nutritional risk (Table 2), or with CHD and high surgical risk established by means of a validated scale. In patients with nutritional risk, referral need not be contingent on the development of malnutrition or weight loss.

Table 3 Recommendations for the referral of infants with CHD to nutrition services.

Who should be referred
Infants with CHD considered to carry high nutritional risk
Infants with CHD and nutritional risk associated with a cardiac factor (myocardial dysfunction, chronic hypoxaemia etc.)
Infants with CHD and nutritional risk associated with an extracardiac factor (chromosomal abnormalities, gastro-oesophageal reflux etc.)
Infants with CHD if they have any clinical condition associated with high nutritional risk (weight loss, vomiting etc.)
Infants with CHD and high surgical risk based on a validated scale (e.g., Aristotle basic score, RACHS-1, STS-EACTS mortality score...)
No consensus: any infant with CHD
When to refer
When the patient is at nutritional risk, without waiting for weight loss or malnutrition to develop.
How to refer
Additional information to convey at the time of referral:
• Myocardial dysfunction
• Presence or absence of pulmonary hypertension
• Presence or absence of heart failure
• Planned surgeries
• Surgical risk
• Cardiological prognosis
• Cardiological treatments

CHD, congenital heart disease.

Nutritional assessment and follow-up

This section was devoted to establishing consensus on the information that must be documented in the health record (such as changes in the infant weight and height gain charts, presence of fatigue or increasing cyanosis during feedings, symptoms associated with intake, etc.) and the nutritional assessment of infants with CHD managed in a paediatric nutrition clinic, among which anthropometric measurements and assessment of warning signs of cardiac failure and undernutrition are essential. The panel agreed on the diagnostic tests that needed to be performed, such as kidney and liver function tests, iron panels, and measurements of electrolyte, albumin, prealbumin and thyroid hormone levels, among others (Table 4). The panel agreed on the application of criteria based on percentiles or z scores of indices including weight-for-height, weight-for-age and height-for age to establish the nutritional status of the patients.

As regards the estimation of nutrient requirements (Table 5), panellists agreed that the nutritional recommendations for healthy infants could be applied making adjustments based on the progress of the patient, for instance, with increases of 50%–100% in the case of chronic undernutrition or of 25%–50% if major surgery is anticipated. Energy and protein requirements should also be adjusted based on the level of nutritional risk. An intake of 90–100 kcal/kg and 1.5 g protein/kg could serve as reference in patients at low nutritional risk, an intake of 110–120 kcal/kg and 2.5 g protein/kg in patients at moderate nutritional risk, and an intake of 20–150 kcal/kg and up to 4 g protein/kg in patients at high nutritional risk. The recommended energy intake in infants with haemodynamically significant CHD and malnutrition could be as high as 3 times the basal metabolic rate.

On the other hand, the recommended follow-up in infants with CHD (Table 5) could be weekly at first, followed by follow-up every 15 days and then monthly. In the case of high nutritional risk, the recommended frequency of follow-up is of at least once a week. Lastly, the panel reached a consensus on the clinical criteria for referral to speech therapy (Table 6).

Nutritional support

The panel agreed by consensus on recommendations regarding nutritional support and the route of administration (Table 7). Efforts should be made to maintain breastfeeding. When not possible, conventional infant or follow-up formula should be used. If the child has malabsorption, the use of semi-elemental formula can be contemplated. To increase the caloric density of the diet, a modular lipid and/or carbohydrate supplement can be added to the formula, complementary feeding may be started earlier than usual (never before 4 months) or a hypercaloric (1 kcal/mL) polymeric enteral nutrition formula for infants may be used. Generally, the total oral fluid intake should not exceed 165 mL/kg/day and the sodium intake should not exceed 2.2–3 mEq/kg/day.

As for the route of administration, the oral route is recommended as the route of choice, restricting the use of nasogastric tube feeding to cases in which oral delivery worsens haemodynamic status or causes fatigue, respiratory distress or hypoxaemia, or in patients with significantly increased nutritional requirements (nutritional rehabilitation) or a high energy expenditure.

The panel also agreed on recommendations for pre- and postoperative nutritional support (Table 8). When a surgical intervention is planned in a malnourished infant with CHD, the patient should receive preoperative intensive nutritional

Table 4 Recommendations on the information to document and tests to order in the assessment of infants with CHD in the department of nutrition.

Health record	Nutritional assessment*	Diagnostic tests
Pregnancy data	Weight	Oxygen saturation (pulse oximetry)
Anthropometry at birth	Length or height	Complete blood count
Socioeconomic factors	Weight-for-height percentile	Kidney function
Weight and height gain charts	Head circumference	Liver function
Growth velocity	Arm circumference	Electrolytes (sodium, potassium)
Concomitant medication	Body mass index	Ionised calcium
Detailed dietary questionnaire	Percentage of expected	Iron metabolism
Appetite	weight-for-height (Waterlow	Total protein
Strength and duration of suck	classification)	Albumin
Fatigue and/or increasing cyanosis	Heart failure signs	Prealbumin
during feeding	Undernutrition warning signs	Thyroid hormone levels
Physical activity and quality of rest	Some form of assessment of body	**Other tests for which consensus was**
Associated symptoms (e.g. vomiting,	composition	**not reached**: lipid panel,
diarrhoea, recurrent infection, etc.)	**Other measurements for which**	immunoglobulins, IGF-1, retinol-binding
	consensus was not reached:	protein, fibronectin, water-soluble
	subscapular skinfold, chest	vitamins (folic acid and vitamin B12),
	circumference, waist circumference,	fat-soluble vitamins (vitamins A, D, E
	triceps skinfold, McLaren index,	and K), micronutrients (zinc, copper,
	Quetelet indices	magnesium, etc.), α_1-antitripsin in stool,
		faecal elastase, quantitative faecal fat
		test, immune cell function tests

CHD, congenital heart disease.
* The panel recommends the use of percentile or z score-based criteria and indices including the weight-for-height, weight-for-age and height-for-age for the classification of nutritional status.

Table 5 Recommendations regarding the estimation of the nutritional requirements and the follow-up of infants with CHD.

- To estimate nutritional requirements, it is possible to apply the recommendations for normal infant nutrition (eg, predictive requirement equations adjusted for age):
- The estimation should be adjusted based on the progress of the patient.
- Increasing by 50%–100% in the case of chronic malnutrition.
- Increasing by 25%–50% in the case major surgery is anticipated.
- Energy and protein requirements should be adjusted based on the nutritional risk:
- Patients at LOW nutritional risk: an energy intake of 90—100 kcal/kg and protein intake of 1.5 g/kg can be used as reference.
- Patients at MODERATE nutritional risk: an energy intake of 10—120 kcal/kg and a protein intake of 2.5 g/kg can be used as reference.
- Patients at HIGH nutritional risk: an energy intake of 120—150 kcal/kg and a protein intake of up to 4 g/kg can be used as reference.
- The recommended daily energy intake in infants with haemodynamically significant CHD and malnutrition can be up to 3 times the basal metabolic rate, that is, near 175–180 kcal/kg to achieve catchup growth and maintain adequate growth thereafter.
- *Recommendation without consensus*: The protein intake must account for 6%–8% of total calories.

Follow-up:

- Initially, the follow-up of infants with CHD may be weekly, followed by every two weeks and monthly thereafter.

It is recommended that infants at high nutritional risk be followed up by the nutrition team weekly (or more frequently if considered necessary).

support for a minimum of 10 days. After the intervention, children with a history of undernutrition before surgery or who required nutritional support before or after surgery should continue to be followed up by a nutritionist.

In cases in which, despite the surgical intervention, the established nutritional targets are not met, the child should be re-evaluated by a paediatric cardiologist.

Discussion

This consensus document offers guidelines for the referral to nutrition services, nutritional evaluation, estimation of nutritional requirements and nutritional therapy of infants with CHD. It may be a useful resource for clinicians who manage this type of patient and may have a positive impact

Table 6 Recommendations regarding the need of assessment by a speech therapist.

Infants with CHD require assessment by a speech therapist if:
- Breathing sounds noisy or wet during or after feeding.
- There are episodes of coughing, gagging or asphyxia during or after feeding.
- Fluid leaks through the mouth or fluid or food is retained in the mouth.
- Respiratory rate or oxygen saturation change during feeding.
- There is a change in colour during or after feeding.
- Oral feeding skills regress or oral motor disorders develop.
- There are difficulties transitioning from enteral to oral feeding.
- *Recommendation without consensus*: if the child exhibits signs of discomfort or unease during or after feeding.

by improving the treatment and outcomes of infants with CHD.

Concerning the identification of patients at risk, the classification of patients into high or low nutritional risk based on the type of heart disease (Table 1) may be a useful first step, especially for the paediatricians initially in charge of the infant. However, the actual nutritional risk in each case of heart disease depends on a host of associated factors.[11,17,18] Therefore, other cardiac and extracardiac factors must be taken into account, in addition to other associated clinical conditions (Table 2).[18,19] Generally speaking, cyanotic heart disease or CHD with pulmonary hypertension are associated with greater growth delays, whereas acyanotic heart disease is associated with greater wasting.[2] The development and progression of undernutrition in these patients is largely dependent on the haemodynamic impact of the cardiac lesions, the development of heart failure, delays in surgical repair, prolonged intubation and feeding intolerance, and pulmonary hypertension is the factor most strongly associated with preoperative undernutrition.[2] In this consensus process, an agreement was not reached for the classification of specific CHDs as carrying a high or low nutritional risk (Table 2). It must be taken into account that the nutritional risk associated with cor triatriatum may vary based on the degree of stenosis, the presence of additional anomalies and especially the presence of pulmonary hypertension.[20,21] In the case of coarctation of the aorta, nutritional risk depends on the severity and location of the coarctation, and whether the coarctation is repaired early. In the case of partial anomalous pulmonary venous return, there are certain haemodynamically significant conditions, such as scimitar syndrome or sinus venosus atrial septal defect, which would have an impact on the degree of pulmonary hypertension and therefore nutritional risk.[22,23] Thus, in addition to the specific CHD, the assessment of nutritional risk must take into account associated factors that may affect this risk.[11,17,18]

When it comes to the referral to the PNU (Table 3), we consider that the key message is that patients should be referred early, that is, when nutritional risk is identified, without needing to wait for the patient to lose weight or become malnourished.[24]

Section III proposed statements regarding the assessment to be performed at the PNU. These statements were based on the guidelines of the AEP[18,25] and the SECPCC.[5] With the items assessed in the consensus process, an instrument was developed that may prove useful in clinical practice (Table 4). The recommendations for the initial workup depend largely on the particular clinical picture and on the suspicion of any specific disease. For instance, measurement of immunoglobulin levels may be useful in patients who have undergone a Fontan procedure in whom protein-losing enteropathy is suspected.[26] Various laboratory measurements were not considered indispensable in all infants with CHD. However, they are shown in the table because these tests may be considered in some cases.

The panel agreed that some form of assessment of body composition should be included in the evaluation of infants with infant with CHD. In addition to anthropometric measurements, there are other direct methods, such as densitometry, bioimpedance, imaging techniques, isotopic methods, etc.[5] Their use will depend on the resources of each facility and the clinical features of the patient.

When it came to the classification of nutritional status, the consensus was that criteria based on percentiles or z scores and the use of indices including the weight-for-height, weight-for-age and height-for-age should be applied, in agreement with the recommendations of different guidelines.[10] On the other hand, a consensus could not be reached regarding the use of national growth charts or nutritional values in healthy infants in Spain as reference in the assessment of infants with CHD (Appendix B, Table S3, item 112). In their comments, numerous panellists recommended the use of the growth standards of the World Health Organization (WHO), as recommended in the guidelines of the American Society for Parenteral and Enteral Nutrition (ASPEN) or the European Society of Pediatric and Neonatal Intensive Care (ESPNIC) for critically ill children.[9,13,27] The WHO has proposed international growth standards for weight, length/height, head circumference, arm circumference and triceps and subscapular skinfolds and calculation of the weight-for-height and the body mass index (BMI). These standards include data for breastfed children aged 0–5 years from different countries across the world. The data are presented in the form of tables or charts of percentiles or z scores.[18,28] However, it is important to keep in mind that when there is an associated genetic disorder (Down syndrome, Noonan syndrome, etc.) the pattern of growth is not consistent with the growth charts and standards published to date. In these cases it is possible to use specialised growth charts, although being aware that these charts are based on descriptive studies and not reference standards.[18]

For the estimation of nutritional requirements, panellists reached a consensus on recommendations regarding energy

Table 7 General nutritional support guidelines for infants with CHD.

Type of nutrition:	Route of administration:
• Efforts should be made to maintain breastfeeding through direct sucking (feeding at breast or with bottle) or through a NGT. • If breastfeeding is not possible, use conventional infant formula (up to age 4—5 months) or follow-up formula (age > months receiving complementary foods). • In infants with malabsorption, the use of semi-elemental formula based on extensively hydrolysed protein, carbohydrates in the form of glucose polymers and lipids with a certain percentage of medium-chain triglycerides may be indicated. **Energy intake:** • There may be recommended maximum and minimum energy intake thresholds. • It may be necessary to increase the caloric density of the delivered nutrition if the infant cannot tolerate intake of large volumes. • The following may be indicated to increase energy intake: - Addition of a modular lipid and/or carbohydrate supplement (always ensuring an adequate macronutrient distribution in the total energy intake). - Introduction of complementary foods (usually gluten-free cereal) earlier than usual, but never before age 4 months. - Delivery of a hypercaloric (1 kcal/mL) polymeric enteral nutrition formula for infants. - *No consensus*: increase the concentration of the formula (under medical supervision and warning the family that this is an exceptional measure). • When the calorie density of the formula is increased to ensure the necessary energy intake without increasing volume, it must be taken into account that insensible fluid loss may increase by 10%–15% or more. • *No consensus*: if the volume required to deliver the necessary amount of protein and energy is poorly tolerated, it may be preferable to increase the dose of diuretics than to decrease volume. **Fluid and electrolyte dosage:** • There may be recommended maximum and minimum fluid intake thresholds. • In general, the total oral fluid intake should not exceed 165 mL/kg/day. • If the volume required to deliver an adequate amount of protein and calories is poorly tolerated, it may be preferable to intensify diuretic therapy than to reduce the delivered volume. • In general, the total sodium intake should be limited to 2.2—3 mEq/kg/day. • *No consensus*: recommendation to perform routine urine osmolality tests to maintain it in the 300—400 mOsm/L range. • *No consensus*: there may be an advisable maximum for daily/weekly weight gain in infants with CHD. • *No consensus*: recommendation to monitor of serum vitamin D levels and deliver supplementation to ensure 25-hydroxyvitamin D levels $\geq$ 80 nmol/L.	• The oral route is the route of choice for delivery of nutrition in infants with CHD. • As long as the patient remains haemodynamically stable, exclusive oral feeding should be maintained. • If exclusive oral feeding is not possible, maintenance of oral feeding during the day combined with night-time nasogastric tube (NGT) feeding to complete the nutritional requirements is recommended. • NGT feeding is indicated: - When oral feeding results in worsening haemodynamic status or causes fatigue, respiratory distress or hypoxaemia. - In patients with increased nutritional requirements for nutritional rehabilitation. - In patients with a high energy expenditure. • The initiation and adaptation to NGT feeding should take place in the hospital, as NGT feeding requires learning and adaptation by the parents and the child. • The approach to NGT feeding that mimics normal physiology most closely is bolus delivery over 4–8 feedings, depending on the age of the patient. • When the patient does not tolerate feeding through a nasogastric or gastrostomy tube, contemplate placement of a postpyloric feeding tube. • If the patient cannot tolerate bolus feeding, consider continuous enteral feeding by gravity tube feeding or with a feeding pump. • Parenteral nutrition is only indicated when it is impossible to use the gastrointestinal tract, for supplementation of enteral nutrition or if it is not possible to achieve the necessary energy intake with enteral nutrition. • *No consensus*: percutaneous endoscopic gastrostomy is the route of choice if a duration of enteral nutrition greater than 8 weeks is anticipated. **Other recommendations** • Treatment of chylothorax requires the replacement of conventional infant or follow-up formula by formula rich in medium-chain triglycerides that are directly absorbed into the portal venous system. • There is not sufficient evidence to support the recommendation of routine administration of probiotics to children with CHD. • Given the strong associations with overall benefit in the paediatric population, the dosage of omega-3 fatty acids should be the one currently recommended in the healthy population. • Patients who continue to have an inadequate nutritional status despite maximum optimization of nutrition should have their cardiovascular health re-evaluated by a paediatric cardiologist. • Educate and support parents and caregivers of children with CHD in the care and management of the different nutritional therapies in these patients.

CHD, congenital heart disease; NGT, nasogastric tube.

Table 8 Recommendations for pre- and postoperative nutritional support of infants with CHD.

Recommendations for the preoperative period:
• When a malnourished infant with CHD is scheduled to undergo surgery, preoperative intensive nutrition therapy should be delivered for a minimum of 10 days.
• Prostaglandin infusion is not a contraindication for trophic feeding.
• *No consensus:* in infants with CHD who require preoperative intensive nutrition therapy, the latter should be delivered via continuous enteral feeding 24 h a day.
Recommendations for the postoperative period:
• Nutritional status should be monitored after surgical intervention in all infants with CHD.
• After surgery, children with a history of undernutrition before surgery or who required nutrition support before or after the intervention should continue to be followed up by a nutrition specialist.
• Enteral feeding should be initiated as soon as it is considered safe based on the condition the patient.
• Indirect calorimetry is the gold standard for the calculation of calorie requirements.
• Energy requirements can be calculated with equations used to estimate energy requirements in critically ill children.
• In the immediate postoperative period in patients with CHD, clinicians should remain alert for red flags that indicate the need to initiate nutrition therapy:
- Failure to gain 20 g of weight in 3 days.
- Intake < 100 mL/kg/day.
- Recurrent vomiting.
- Change in stool frequency and consistency.
- *No consensus*: weight loss > 30 g in one day.
• Postoperative nutritional support in infants with CHD may differ from preoperative nutritional support based on postoperative outcomes.
• All infants with CHD who develop postoperative complications that may affect nutritional status (protein-losing enteropathy, chylothorax, malabsorption) should receive specific nutrition therapy.
- The patient must be re-evaluated by a paediatric cardiologist if the maximum energy and volume intake has been reached.

CHD, congenital heart disease.

and protein requirements (Table 5) consistent with those of other guidelines and consensus documents.[5,19] A consensus was not reached for an item proposing that the protein intake should amount to 6%–8% of the total calorie intake. The literature suggests an optimal protein-to-energy ratio of 9%–12%.[19,25] For critically ill children, the ASPEN guidelines propose a protein intake of 2–3 g/kg/day and the ESPNIC guidelines a minimum protein intake of 1.5 g/kg/day.[9,13,27]

The section devoted to evaluation included items on the need for assessment by a speech therapist (Table 6). These recommendations are based on previous consensus documents.[19] All proposed recommendations were agreed on, except for the one that contemplated referral to the speech therapist if the infant exhibited signs of discomfort or unease during or after feeding. Some panellists considered that these symptoms were not specific enough and that other possible causes, such as gastro-oesophageal reflux, should be ruled out before referring the patient.[29]

Lastly, the panel agreed on general recommendations for the delivery of nutritional support, its route of administration (Table 7) and nutrition in the perioperative period (Table 8) in line with previous recommendations.[5,25]

With regard to surgery, we ought to highlight the item that contemplates that when surgical intervention is planned in infants with CHD and malnutrition, the infant must receive intensive nutrition therapy for a minimum of 10 days before surgery (Appendix B, Table S4.3). Nearly one third of infants with CHD require some form of surgical intervention, usually in the first year of life and increasingly frequently in the neonatal period.[7] Early repair decreases the probability of undernutrition, but up to 50% of children may have protein-energy undernutrition at the time of the intervention.[5] In addition, a poor nutritional status in the preoperative period may be associated with unfavourable postoperative outcomes, increasing the risk of nosocomial infection and poor wound healing.[7,30] Consequently, early diagnosis and adequate pre-and postoperative intervention are essential in this context.[7]

This work has the limitations intrinsic to the Delphi method, including the impossibility of discussing recommendations in depth or the possibility of bias in the selection of panellists. However, the scientific committee took into account the comments made by the panellists in the writing of the Discussion section and the selection of participants was very careful and included only physicians with demonstrated expertise on the subject.

In summary, infants with CHD, especially those with haemodynamically significant disease, may be at risk of undernutrition, which is associated with an increased morbidity and mortality. Thorough assessment and appropriate nutritional support of these children is crucial to improve treatment, long-term outcomes and quality of life. The committee developed consensus-based recommendations on the nutritional management of infants with CHD with the participation of physicians in all the specialities involved in their management. These recommendations could facilitate early detection and referral of patients at risk and guide the assessment, rapid estimation of nutritional requirements

and appropriate nutrition and feeding of children with CHD in PNUs.

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Conflicts of interest

The authors have no conflicts of interest to declare.

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Appendix A. Supplementary data

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