Disease-related malnutrition in congenital heart disease: what is the risk and impact on patients' outcomes?

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Abstract

The most frequent congenital defect in newborns is congenital heart disease (CHD). In children with CHD, malnutrition often occurs, posing negative impacts on their health. Malnutrition is associated with increasing morbidity and mortality rate in children with CHD. To decrease unfavorable outcomes, nutritional screening and management are indicated before and after surgery. This review focuses on delivering information on prevalent CHDs in children, as well as nutritional management before and after heart surgery.

Keywords: congenital heart disease, malnutrition, surgery

Introduction

Congenital heart disease (CHD) refers to all heart problems occurring before delivery. It comprises flaws in the heart architecture that obstruct venous outflow, cardiac section septation and sequence, and normal valve apparatus function.\(^1\) It is the most typical congenital birth defect, accounting for almost one-third of all congenital defects.\(^2\) Congenital cardiac disease affects 6 to 8 out of every 1000 newborns.\(^3\) Due to various types of cardiac abnormalities, CHD frequently presents with a variety of signs and symptoms. Excessive perspiration, intense exhaustion, poor feeding, shortness of breath, fatigue, rapid heartbeat, chest pain, cyanosis (blue tinge to the skin), and clubbing fingernails are some of the symptoms of this illness. Clinical examination, chest x-ray, echocardiography, and electrocardiogram (ECG) are among the various diagnostic workup for this disease. If the transthoracic windows are not optimum, a transesophageal echo may be needed. Children with pulmonary hypertension and probable

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pulmonary vascular disease often need to undergo diagnostic catheterization.\textsuperscript{2} Malnutrition often occurs among children with CHD due to many factors. Malnourished patient is more likely to acquire infection and have poor wound healing due to fewer nutritional substrates available to respond to the enhanced catabolic consequences of injury from cardiac surgeries.\textsuperscript{4}

The review aims to provide information on common CHD in children as well as nutritional management before and after heart surgery.

**Classification of CHD**

1. **Acyanotic heart disease**

   1.1. **Atrial septal defect**

   The second most prevalent congenital heart disease is atrial septal defect (ASD), comprising around 7-10\% of all CHDs.\textsuperscript{3,5} When the communication between the right and left atria is not closed, an atrial septal defect occurs. Because the left atrium level is higher than the right one, ASD causes a left to right shunt.\textsuperscript{6} If the septum is damaged or absent, oxygen-rich blood from the left side of the heart can flow directly to the oxygen-poor blood in the right one (left-to-right shunt), or vice versa.\textsuperscript{9} Ostium secundum, sinus venosus, ostium primum, patent foramen ovale and coronary sinus defects, are the five major types of ASD. A quiet systolic crescendo-decrescendo outflow tract murmur, as a result of a rigid split in the S2 heart sound, may be heard on physical examination due to increased flow over the pulmonary valve.\textsuperscript{10}

   Small ASDs are likely to shut on their own and do not require closure. On echocardiogram, right ventricle volume overload (dilatation of the right side of the heart, with flat-to-paradoxical interventricular septal flow) and a pulmonary-to-systemic flow ratio of more than 1.5:1.\textsuperscript{10} are both indications for ASD closure. Elective surgical or catheter-based closure is suggested to be delayed until the patients are up to 3-5 years of age. Closure is not performed on infants unless they are exhibiting symptoms.\textsuperscript{8}

   **1.2. Ventricular septal defect**

   The most common CHD is ventricular septal defect (VSD), which affects nearly half of all children with CHD. The interventricular septum, splitting the heart's right and left ventricles, will be defectively formed, and VSD develops as a result.\textsuperscript{5,12} A left-to-right shunt occurs after an uncomplicated VSD causes oxygenated blood to flow from the high pressure in the left ventricle to the low pressure in the right ventricle, and back into the lungs. As a result, the patient is acyanotic.\textsuperscript{5,13} The characteristic pansystolic murmur, best-heard at the left mid-to-lower sternal border, could go unnoticed until the shunt becomes maximum at a few weeks of age.\textsuperscript{3,5} In general, the smaller the defect, the more intense the murmur.\textsuperscript{3,5}

   Anti-congestive medications, such as furosemide, chlorothiazide, and/or spironolactone, are employed in the management of VSD with congestive heart failure symptoms.\textsuperscript{5} The reduction of afterload is required to promote direct systemic flow from the left ventricle, reducing the sum of left-to-right shunting through the defect. Angiotensin-converting enzyme inhibitors (ACEIs) are employed to reduce afterload.\textsuperscript{5} Milrinone can also be given intravenously to produce inotropy and afterload reduction, but this is normally reserved for patients who are about to undergo surgery.\textsuperscript{13} Patients with failure to thrive might require caloric intake of up to 125 until 150 kcal/kg per day from fortified formula.\textsuperscript{5}

   The size and type (location) of VSD are the most important factors in determining whether or not surgery is necessary. Small VSDs do not need to be closed. Anti-congestive treatments should be used first for moderate VSD with congestive heart failure. A large VSD should be closed if the right ventricular and pulmonary artery pressures are equivalent to the left ventricle and aortic systolic pressures. VSD closure should be done before the child turn 6 to 12 months (should not exceed 18 months of age).\textsuperscript{5}

   **1.3. Patent Ductus Arteriosus**

   Ductus arteriosus that failed to close in 72 hours post-delivery is known as patent ductus arteriosus (PDA).\textsuperscript{15} All newborns have an arterial duct, which
is a typical connection between the aorta and the pulmonary artery that exists since their early development as a fetus.\textsuperscript{5} Greater oxygen tension and increased circulating PGE\textsubscript{2} and prostacyclin (PGI\textsubscript{2}) in full-term neonates cause it to shut completely within 24–72 hours.\textsuperscript{5} In some patients, particularly preterm infants, the duct could stay open for a longer period of time.\textsuperscript{5,17,18} Clinical symptoms become less reliable than echocardiographic outcomes in most cases. The gold standard for detecting the existence of a patent ductus arteriosus to date is echocardiography. Bounding pulses, tachypnoea, tachycardia, a broad pulse pressure, and a systolic murmur are all common symptoms of PDA.\textsuperscript{15} Wide pulse pressures, a distinctive coarse systolic murmur at the left sternal boundary, cardiac hypertrophy (as a symptom of systemic hypoperfusion compensation), and hypotension are the most typical symptoms of PDA.\textsuperscript{5,17,19}

PDA has been treated with ibuprofen, indomethacin, and acetaminophen.\textsuperscript{17,18} If they are hemodynamically significant, tiny and small PDAs should be sealed.\textsuperscript{5,18,19} To address the PDA, surgical ligation, video assisted thoracoscopic surgical (VATS) interruption, and trans-catheter occlusion of PDAs are currently viable alternatives.\textsuperscript{8,19} PDA closure can be done at any moment, primarily if heart failure or pulmonary compromise are present. However, due to the decreased risk profile at 6-12 months, kids who are asymptomatic should wait until they reached this particular age before they can undergo closure.\textsuperscript{10}

2. Acyanotic heart disease

2.1. Tetralogy of Fallot

The most frequent cyanotic CHD is Tetralogy of Fallot, which affects 0.34 out of every 1,000 live infants.\textsuperscript{20} A ventricular septal defect (VSD), overriding aorta, right ventricular (RV) outflow tract obstruction (RVOTO) which is often dynamic, and right ventricular hypertrophy (RVH) are the four cardinal characteristics of Tetralogy of Fallot.\textsuperscript{21} A right-to-left shunt is possible with a VSD. Blood flow to the pulmonary circuit is restricted by the RVOTO. The presence of an overriding aorta permits more deoxygenated and oxygenated blood to enter the circulatory system. The additional workload required to circulate blood to the right ventricular blockage and the aorta obstructing the flow of blood causes right ventricle hypertrophy.\textsuperscript{13,21}

A PDA may be needed by patients with Tetralogy of Fallot with severe RVOTO to provide adequate pulmonary blood flow; for instance a duct-dependent circulation necessitates prostaglandin infusion (either alprostadil [prostin E1] or dinoprostone [prostin E2]) until more temporizing palliative procedures or definitive surgical procedures can be performed.\textsuperscript{18,19} If spells occur frequently in infants with dynamic RVOTO, beta-blockade (e.g. propranolol) may be required.\textsuperscript{18} Patients with Tetralogy of Fallot and a large ventricular septal defect who are acyanotic will possibly develop a massive left-to-right shunt, which may necessitate the use of diuretics and may lead to heart attacks.\textsuperscript{18}

Surgery is recommended for children whose saturation levels are below 80% or who have hypercyanotic spells.\textsuperscript{5} When there is a significant stenosis or pulmonary valve atresia, surgical therapy may include palliative operations such as a modified Blalock-Taussig-Thomas (mBTT) shunt to assist in the provision of continuous pulmonary blood flow.\textsuperscript{5} While waiting for a full repair as the infants grow, catheterization may help increase blood flow in the lungs.\textsuperscript{5,21} Complete cardiac repair procedure is usually performed before the child reached the age of six months. The procedure includes closing the ventricular septal defect and septating the aorta back to the left ventricle, as well as the surgically removing the RVOT muscle bundles and reducing the degree of RVOT valvular stenosis. This results in normal saturations.\textsuperscript{5,18}

Nutrition status of congenital heart disease patients

Malnutrition is described as a discrepancy between dietary needs as well as consumption, causing cumulative calorie, protein, or micronutrient shortages which can have a deleterious impact on growth, development, and other outcomes.\textsuperscript{5,21} The WHO advises that malnutrition be defined and classified using established concepts and categories.
depending on the anthropometric indicators' computed Z scores. Moderate malnutrition is defined as a Z score of \(-2 \leq Z < -3\) SD for low WAZ (underweight), low WHZ (wasting) and, low HAZ (stunting), and severe malnutrition is described as a Z score of \(Z \leq -3\) SD.\(^{23}\) Malnutrition or nutritional deficiencies generally occurred in children with CHD due to a range of factors, such as lesser energy diet owing to the lack of food and poor nutrient uptakes from the digestive system, extra energy demands due to their cardiac and respiratory conditions as well as due to poor neurohumoral functions in CHD, or both.\(^{21-23}\) Malnutrition in children with CHD can also be caused by chromosomal anomalies/genetic disorders, high blood pressure in the blood vessels that supply the lungs, as well as the age at presentation.\(^{20-22}\)

Malnutrition is associated with adverse health outcomes in children with CHD, as indicated by regular admissions, poor surgical outcomes, permanent visceral development impairment, and higher death.\(^{21}\) Children with cyanotic cardiovascular disease and pulmonary hypertension tend to have stunting, whereas those with acyanotic cardiovascular disease are likely to be wasted.\(^{21,22,24}\)

**Pre-operative phase**

Corrective procedures for CHD are delayed in underdeveloped nations due to resource constraints, resulting in a never-ending phase of heart problems and respiratory disease. Malnutrition may be associated with other risk factors in developing countries compared with developing countries. Patients with CHD have a significant prevalence of nutritional deficiencies just prior to surgery.\(^{25,27}\)

The prevalence of malnutrition was observed to be 84.0 % in patients with CHD in Egypt, according to Hassan et al.\(^{25}\) Vaidyanathan et al.\(^{29}\) found that 59.0 % of infants with CHD were malnourished prior to surgery in South India. Even before surgery, 23.3 % were underweight, 23.3 % were stunted, and 14.3 % were wasted, according to Zhang et al.\(^{26}\)

Children with CHD who also had malnutrition have a higher risk of poor outcomes.\(^{21,23,30,31}\) Stunted children had a higher risk for longer length-of-stay, longer period of mechanical ventilation, and required over three inotropes post-operatively, whereas children who were underweight were at a greater danger of 30-day mortality and respiratory failure.\(^{26,30,31}\) A complete preoperative evaluation is recommended to provide crucial baseline data for evaluating progress after surgical correction. Accurate measurements of weight, height, and head circumference is typically needed.\(^{20}\)

Strategies to increase nutritional balance in children with CHD include: (1) cautious reintroduction of high-calorie foods; (2) avoidance of excessive fluid loads; (3) salt restriction; and (4) electrolyte monitoring.\(^{33}\)

Significant daily calorie multivitamin supplements in children with CHD contribute to enhanced growth and surgery outcomes, and interventions such as the introduction of calorie-rich foods, infant formula, protein additive or vitamins and minerals. In some cases, changing the technique of feeding, such as bolus or continuous enteral feeding via nasogastric, small bowel, or gastrostomy feeding, could also be used.\(^{26,30}\)

The standard 75 to 120 kcal/kg/day requirements is needed to maintain caloric intake. Protein must account for 8-10% of caloric intake, with carbohydrate accounting for 35-65% and fat accounting for 35-50% (4% being essential fatty acids). Potassium balance is essential and should be provided due to anabolism, particularly if diuretics are used often. The daily dose is 2-3 mEq/kg, but in cases of acute depletion, it can be as high as 4-5 mEq/kg/day. Even if salt is reduced to prevent heart problems, at least 2-3 mEq/kg daily sodium intake is required. In children with CHD, urinary osmolarity should not surpass 400 mOsm/L.\(^{33}\)

**Post-operative phase**

In the early days after surgery, patients require more energy, thus enteral feeds should be started after it is feasible, and parenteral nutrition should only be employed if definitely necessary.\(^{26,39}\) A combination of genetic factors, multiplied metabolic needs, inadequate nutrient uptake due to ineffective gut function to obtain energy, postsurgical fluid restriction to avoid volume overload, oropharyngeal dysfunction, and interruptions of enteral feeding for procedures can make achieving adequate nutritional intake difficult.\(^{27,32,33}\)
Table 1. Summary of the correlation between pre-operative nutritional status and post-operative outcomes

<table>
<thead>
<tr>
<th>References</th>
<th>Sample size (n)</th>
<th>Age-range</th>
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<tr>
<td>Anderson et al.34</td>
<td>100</td>
<td>2-10 months</td>
<td>WAZ</td>
<td>Hospital Length of Stay</td>
<td>Lower WAZ suggested a longer hospital Length of Stay</td>
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<td>Duration of mechanical ventilation</td>
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<td>Chest tube duration</td>
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<td>Anderson et al.35</td>
<td>55</td>
<td>18-72 months</td>
<td>WAZ</td>
<td>Hospital Length of Stay</td>
<td>WAZ&lt;-2 predicted a higher probability of severe postoperative infections, and severe postoperative infections predicted a longer stay in the hospital</td>
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<td>Duration of mechanical ventilation</td>
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<td>Post-operative infections</td>
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<td>Chest tube duration</td>
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<td>Wallace et al.36</td>
<td>2,747</td>
<td>&lt;6 years</td>
<td>Age, Weight, WAZ</td>
<td>In-hospital mortality</td>
<td>WAZ&lt;-2 predicted higher in-hospital mortality, Fontan failure, and a longer hospital Length-of-Stay</td>
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<td>Fontan failure*</td>
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<td>Hospital Length-of-Stay</td>
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<td>Complications</td>
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<td>Low WAZ predicted a longer combined mechanical ventilation and non-invasive ventilation duration and greater mortality at one year.</td>
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<td>Mortality at 1 year</td>
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<td>Duration of Mechanical/Non-invasive ventilation</td>
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<td>ICU Length-of-Stay</td>
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<td>Maximum lactate</td>
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<td>Marwali et al.37</td>
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<td>5-36 months</td>
<td>WAZ</td>
<td>ICU Length of Stay</td>
<td>Lower WAZ was linked to a longer ICU Length of Stay and mechanical ventilation length.</td>
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<td>Duration of mechanical ventilation</td>
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The most common concern with early feed initiation is the risk of poor cardiac output due to gut hypoperfusion, which can develop to necrotizing enterocolitis in infants with duct-dependent circulation. Feeds are generally triggered at 1 ml/kg/hour in newborns and young infants within 12–24 hours following surgery and increased at the same rate every 4 to 6 hours to attain the goal volume. The maintenance fluid rate determines feed volume in the early post-operative phase. To attain a negative fluid balance, fluid intake is generally restricted.

The resting energy consumption in the first 3–5 days is expected to be 55–75 kcal/kg/day based on research in CHD patients. This helps guide initial feeding, which can be increased to 120–150 kcal/kg/day when transitioning to a lower level of care to facilitate catch-up growth. Breast milk is the best source of nourishment for neonates and babies, especially in low- and middle-income countries (LMIC).
In addition to breastmilk, new research suggests that protein- and energy-enriched infant formula (PE formulas) may aid in achieving nutrition goals and promoting anabolism in infants following cardiac surgery.\textsuperscript{26,41} Scheeffe et al.\textsuperscript{42} found that energy-enhanced formula is well tolerated after heart surgery in CHD patients and could help with short-term nutritional outcomes, minimizing hospital length-of-stay and the use of antibiotics. In newborns, PE milk is well-tolerated by infants with congenital heart surgery and is helpful in attaining higher nutritional intake even in the first days after surgery, according to Cui et al.\textsuperscript{43}

A study conducted in developed countries found that two years following surgery, catch-up growth is essentially complete.\textsuperscript{27} However, another study found that at the third year after surgery, numerous children remain malnourished (1.9 % wasting, 2.7 % stunting, and 3.2 % underweight).\textsuperscript{26}

**Conclusion**

One of the most common human developmental anomaly is congenital heart disease, which in children is frequently associated with malnutrition and failure-to-thrive. Growth failure has been linked to higher morbidity and mortality in children with CHD. To avoid unfavorable outcomes, nutritional management is required both before and after surgery. Human breast milk is the best nourishment for children with CHD. Protein- and energy-enriched formulas, in addition to human breast milk, can help children with CHD who are undergoing surgery have a better outcome.

**Conflict of Interest**

Authors declared no conflict of interest regarding this article.

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