

Nutrition in Congenital Heart Disease

Cape Town Metropole Paediatric Interest Group

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Nutrition in the Paediatric Cardiac Patient

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1. Glossary

Term	Definition
% EHA	Percentage estimated height for age
% EHW	Percentage estimated height for weight
% EWA	Percentage estimated weight for age
ABW	Actual body weight
AMA	Arm muscle area [requires MUAC & TSF to calculate]
CHD	Congenital Heart Disease
CHO	Carbohydrate
DRV's	Dietary Reference Values [Appendix 1]
FBC	Full Blood Count
FBDG	Food Based Dietary Guidelines
FTT	Failure to Thrive
HA	Height age
Hb	Haemoglobin
IMCI	Integrated management of childhood illness
IMCI: Not Growing Well	<ul style="list-style-type: none"> • <i>Severe Malnutrition:</i> • Very low weight < 60% EWA. • Visible signs of severe wasting • Oedema on the feet • <i>Not Growing Well:</i> • Low weight < 3rd centile; g • Poor weight gain - gaining weight but curve flattening or • Mother reports weight loss. • <i>Growing Well:</i> • Not low weight and • Good weight gain.
LCPUFA	• Long chain polyunsaturated fatty acids
LCT	• Long Chain Triglyceride
MAC	• Mid arm area circumference
MCT	• Medium Chain Triglyceride
MUAC	<ul style="list-style-type: none"> • Mid upper arm circumference [6months – 5 years of age] • > 15cm normal • >11.5cm - <14.5cm moderately malnourished • <11.5 cm [<-3SD] severely malnourished
NOBI	• Non Occlusive Bowel Injury
NSP	• Nutrition supplementation programme (NSP)
NSP Definition: Growth faltering	<ul style="list-style-type: none"> • Birth – 5 years: when an infant or child's growth curve flattens or drops over two consecutive visits on his/her RTHC. • >5 - < 18 yrs: when a child's growth curve flattens or drops over two consecutive months on his/her weight-for-age growth chart.
NSP: Entry Criteria	<ul style="list-style-type: none"> • Supplementation must be continued for only 6 months if entered onto the Nutrition Supplementation Programme. • <i>Infants: 0 – 12months</i> growth curve flattens or drops over two consecutive visits on his/her RTHC and the mother is unable to breastfeed because of the following reasons: <ul style="list-style-type: none"> • Serious systemic, on long-term medication or treatment e.g. chemotherapy, hypothyroidism; is addicted to alcohol or drugs (condition must be formally documented/assessed); is mentally disabled and poses a threat to the baby; the infant is in foster care. • <i>Children > 5 years ≤ 18 years:</i> When child's growth curve flattens or drops over two consecutive months.
NSP: Exit Criteria	<ul style="list-style-type: none"> • <i>Successful:</i> • <i>Birth – 5 years:</i> gained sufficient weight to attain a growth curve in relation to his/her normal growth curve and maintains the curve for three consecutive months. • <i>>5< 18 years:</i> gained sufficient weight to attain normal growth curve according to the growth

	<p>chart within the 6 months period on the scheme</p> <ul style="list-style-type: none"> • <i>Unsuccessful:</i> • Birth – 5 years: Failure to attain growth curve in relation to his/her normal growth curve over a period of 6 months and if no underlying disease/condition is present e.g. Foetal Alcohol Syndrome • >5< 18 years: who do not attain a normal growth curve according to the growth chart with in the 6 months period. • <i>Defaulter:</i> • Birth – 5 years: Failure to attend the clinic for a period of three consecutive months. • > 5 - <18 years: Failure to attend the clinic for a period of three consecutive months within the 6 months period. • Client has a history of irregular clinic attendance (less than three visits in a 6 month period) with in the 6 months period. • ** Re-entry: • UNSUCCESSFUL and DEFAULT cases MAY NOT be re-entered onto the programme. • SUCCESSFUL cases MAY be re-entered onto the programme according to entry criteria.
RDA	• Recommended Daily Allowances
REE	• Resting Energy Expenditure
RTHC	Road to health card (Clinic Card)
RTU/RTU	Ready to use/ Ready to hang
Schofield Equation	• Predicting estimated energy requirements [Appendix 1]
SD	<ul style="list-style-type: none"> • Standard Deviations used to determine moderate to severe malnutrition: • 0 - <-1 SD Normally Nourished • >-2 – -3 SD Moderately Malnourished • >-3SD Malnourished
TSF	• Tricep Skinfold Thickness
TTO	To Take Out
WA	• Weight age
Waterlow Criteria (WHO)	<ul style="list-style-type: none"> • Used to determine malnutrition: • <i>Acute malnutrition: Weight/ Height</i> • Normal WH >90%, • Mild 81% - 90%, • Moderate 70% - 80%, • Severe <70%. • <i>Chronic malnutrition: height for age</i> • Normal >95%, • Mild 90 –95%, • Mild – moderate 85% to 89% • Severe < 85%.
WCC	White Cell Count
WH	Weight for height

2. Summary of recommendations for nutrition management of infants and children with congenital heart disease

Summary Recommendations: Congenital Heart Disease ^{1, 2, 12, 25}

2.1 Anthropometry		
<ul style="list-style-type: none"> Complete on admission & weekly until discharge. 	<ul style="list-style-type: none"> Height Weight MUAC Head Circumference < 3 years of age <p><i>Calculate</i></p> <ul style="list-style-type: none"> % EWA % EWH % EHA HA WA WH <p>Classify degree of malnutrition</p> <p><i>Waterlow: Acute malnutrition: Weight for Height (wasting)</i></p> <ul style="list-style-type: none"> Normal WH >90% Mild 81% - 90% Moderate 70% - 80% Severe <70% <p><i>Waterlow: Chronic malnutrition: height for age (stunting)</i></p> <ul style="list-style-type: none"> Normal >95% Mild 90 –95% Mild – moderate 85% to 89% Severe < 85% <p><i>Gomez: Acute wasting: Weight for age</i></p> <ul style="list-style-type: none"> Obese >120% Normal > 90% Mild malnutrition 76 – 90% Moderate malnutrition 61 – 75% Severe malnutrition < 60% 	<ul style="list-style-type: none"> Plot weight & height on appropriate growth charts. (CDC or WHO or disease specific e.g. Downs Syndrome) Expected weight gain for an infant (< 6 months) with CHD is 10 – 20g per day and for infants (6 – 12 months) 120 – 210g/week
2.2 Biochemistry		
<ul style="list-style-type: none"> Complete daily post operatively whilst in ICU Once in recovery complete x 2 week until discharge 	<p><i>Monitor the following</i></p> <ul style="list-style-type: none"> U & E: Urea, creatinine, sodium, potassium Calcium, magnesium and phosphorus Glucose FBC: Hb, platelets, WCC 	
2.3 Clinical		
<ul style="list-style-type: none"> Mechanisms of malnutrition effected by: Type of cardiac lesion Increased metabolic rate Inadequate caloric intake Weight at time of operation Prenatal factors Medication Cardiac cachexia Urinary sodium losses (especially on Frusemide-Lasix) Low Hb Microcytic anaemia Macrocytic anaemia Low Zinc or Selenium 	<p>Comments:</p> <ul style="list-style-type: none"> High urinary sodium losses can result in failure thrive. Establish sodium balance over a 24 hour period Check Iron status Check folate and Vit B12 levels Infants with cyanotic CHD may have a normal Hb but iron deficient. Check iron status using ferritin, red cell indices and total iron binding capacity. 	<ul style="list-style-type: none"> Common clinical signs are: Fatigue on feeding Early satiety Anorexia Failure to thrive Frequent Infections

2.4 Dietary

At each follow up a thorough nutrition history should be completed

Components of a nutrition history include			Components of a nutrition history include (cont.)		
<ul style="list-style-type: none"> • Weight Change • Appetite • Satiety Level • Taste Changes/ aversions • Nausea/ vomiting • Bowel habits – constipation, diarrhoea • Chewing/ swallowing ability • Shortness of breath on feeding 			<ul style="list-style-type: none"> • Long-term disease(s) affecting absorption/use of nutrients • Dietary history – 24 hour recall/ food frequency • Use of vitamin/ mineral or nutritional supplements • Medications • Level of activity/ exercise • Ability to secure and prepare food • Over the counter medications, vitamins and herbal remedies. 		
Diet history			Method of feed administration (employ a stepwise downward approach)		
<ul style="list-style-type: none"> • Review through 24 hour dietary recall quarterly or at each follow up review. • Use in conjunction with food frequency. • Many patients may eat < 65% of RDI. 			<ul style="list-style-type: none"> • Offer smaller volumes and more frequent feeds orally • Give any unfinished feeds via naso-gastric tube if required • Give small frequent bolus feeds via naso-gastric tube • Top up small frequent daytime feeds with continuous overnight feeds via an enteral feeding pump • Give feeds continuously over 24 hours via an enteral feeding pump. <p>NOBI (Non occlusive Bowel Injury) <i>Initiation of enteral feeds in patients with circulatory compromise (sepsis, cardiogenic shock, haemodynamic instability) may lead to deleterious changes on the structure and function of the gut. It is therefore imperative to monitor for any signs of feeding intolerance.</i></p>		
Fluid	<p>Fluid Ranges</p> <p>Age (years)</p> <p>Premature</p> <p>0-1</p> <p>1-3</p> <p>3-6</p> <p>7-10</p> <p>10-15</p> <p>Weight</p> <p>Premature < 2kg</p> <p>Neonates and infants 2 – 10kg</p> <ul style="list-style-type: none"> • 0 – 6 months • 6 – 12 months <p>Infants and children 10 -20kg</p> <p>Children > 20kg</p> <p>DO NOT ALTER MEDICALLY INDICATED FLUID RESTRICTION</p> <p>(Fluid requirements include fluid from feed, medication, IV fluids and oral sips)</p>	<p>ml/kg actual weight</p> <p>180-200</p> <p>150</p> <p>100</p> <p>90</p> <p>70</p> <p>60</p> <p>Fluid Volume per 24 hours</p> <p>150ml/kg</p> <p>150ml/kg</p> <p>120ml/kg</p> <p>1000ml + 50ml/kg over 10kg</p> <p>1500ml + 20ml/kg over 20kg</p>			
Energy	<p>Infants:</p> <p><u>Pre-operative or post shunt</u></p> <ul style="list-style-type: none"> • Ventilated: 90 – 100 kcal/kg • Non ventilated: 120 – 150kcal/kg (maximum 170 kcal/kg) <p><u>Post definitive operation (Cardiac Repair)</u></p> <ul style="list-style-type: none"> • Ventilated: 90 – 100 kcal/kg • Not ventilated: Schofield using abw + activity 1.2 + stress 1.5 – 1.6 <ul style="list-style-type: none"> • In sedated, ventilated children's energy expenditure if often significantly reduced. • Aim to feed at 150kcal/kg and only increase to 170kcal/kg if other factors effecting growth have been excluded 	<p>Children:</p> <p><u>Pre-operative or post shunt</u></p> <ul style="list-style-type: none"> • Ventilated: Schofield equation or WHO/FAO/UNU x 1.3 – 1.5 [No activity factor] • Non ventilated: 120 – 150% or DRV's <p><u>Post definitive operation (Cardiac Repair)</u></p> <ul style="list-style-type: none"> • Ventilated: Schofield equation or WHO/FAO/UNU x 1.3 – 1.5 [No activity factor] • Non ventilated: Schofield using abw + activity 1.2 + stress 1.5 – 1.6 			

Energy Supplementation	<p><i>Infants:</i></p> <ul style="list-style-type: none"> No additional energy may be required in the pre-repair infant and breast milk and or standard ready to use/ hang infant formula [0.67kcal/ml] should be given. If the patient is volume-restricted breast milk may be supplemented with a human milk fortifier or carbohydrate/fat powder and or a ready to use/hang energy dense infant feed [1kcal/ml] should be given. <p><i>Children:</i></p> <ul style="list-style-type: none"> No additional energy may be required and a standard feed [1kcal/ml] should meet requirements in the volume prescribed. If the patient is volume restricted an energy dense [1.5 kcal/ml] ready to use/hang feed should be given. <p>NB:</p> <ul style="list-style-type: none"> No powders or liquids e.g. oil should be added to a sterile ready to use feed. If additional energy is required in non-ventilated children then flushes of energy boluses should be provided prior to a drink or feed including breastmilk. Recommendations for fat, protein and carbohydrate concentrations should not be exceeded. [See sections below] 	
Carbohydrate	<p><i>Glucose requirements</i></p> <ul style="list-style-type: none"> > According to tolerance <p><i>Infants</i></p> <ul style="list-style-type: none"> 8-9mg/kg/min [11.5g –12.9g/kg/day] Max 12.5mg/min/kg [18g/kg/day] <p><i>Toddlers</i></p> <ul style="list-style-type: none"> 7mg/kg/min [10g/kg/day] <p><i>Adolescents</i></p> <ul style="list-style-type: none"> 4mg/kg/min [5.7g/kg/day] 	<ul style="list-style-type: none"> The following concentrations of CHO per 100ml will be tolerated if a CHO/fat powder is used. <p>10-12% carbohydrate concentrations in infants under 6 months (i.e. 7g from formula, 3-5g added)</p> <ul style="list-style-type: none"> 12-15% in infants aged 6months to 1 year 15-20% in toddlers aged 1-2 years 20-30% in older children
Protein	<p><i>Infants:</i></p> <p><u>Pre-operative or post shunt</u></p> <ul style="list-style-type: none"> 9 – 11% of total energy Up to 4 g/kg abw <p><u>Post definitive operation (Cardiac Repair)</u></p> <ul style="list-style-type: none"> 9 – 11% of total energy DRV's <p><i>Children</i></p> <p><u>Pre-operative, post shunt and post definitive operation (Cardiac Repair)</u></p> <ul style="list-style-type: none"> Up to 2 g/kg abw 	
Fat	<p><i>Birth - < 5 years of age</i></p> <ul style="list-style-type: none"> 40% NPE <p><i>Children > 5 years of age</i></p> <ul style="list-style-type: none"> 30 – 35% NPE 	<p><i>Adding Fat to formula/enteral feeds:</i></p> <ul style="list-style-type: none"> Should be done as a last resort – rather add extra oil/ margarine to food. Infants will tolerate a total fat concentration of 5 – 6 % [e.g. 5 – 6g per 100ml of feed]. Children > 1yr will tolerate a fat concentration of 7% - concentrations above this may cause nausea/ vomiting. It is recommended that a soluble carbohydrate/fat powder be used in bolus form over modulars such as oil and/or a glucose polymer
Micronutrients	<ul style="list-style-type: none"> A daily multi-vitamin should be given e.g. Abidec® or Vidaylin® Elemental Zn 1 – 3 mg/kg abw Selenium 2mg/kg abw with a max of 30mg/day Iron: 2mg/kg/day for prophylaxis 6mg/kg/day if Fe deficient 	

Discharge Planning	<ul style="list-style-type: none"> Educate the parent/caregiver on the supplementation of feeds (this can be started prior to discharge). The caregiver is asked to make up the child's supplemented feeds at ward level as a way of educating him/her under supervision. Provide caregiver with a date for dietetic follow up or refer to a private practising dietitian. Ask the doctor to write up a prescription TTO (To Take Out) for multivitamins 	<p>Government: Supply sufficient micronutrient supplements until follow up appointment Private: 7 day TTO on hospital discharge and provide a second script for at least 1 month supply. Continue multi-vitamins until catch up growth has been achieved.</p>
Post Discharge	<ul style="list-style-type: none"> Frequency of follow up 	<ul style="list-style-type: none"> Poor growth: monthly If failing to thrive, follow guidelines below Thriving: Quarterly
2.5 Entry and Exit Criteria for Nutrition Support		
<p>Entry Criteria Nutrition Support NSP</p> <ul style="list-style-type: none"> Supplementation must be continued for only 6 months if entered onto the Nutrition Supplementation Programme. Children > 5 years ≤ 18 years: When child's growth curve flattens or drops over two consecutive months. <p>Or Private Patients</p> <ul style="list-style-type: none"> Growth failure Downward crossing 2 or more centiles over a period of 1 month or 2 consecutive visits. MUAC < 12.5cm in children < 5years of age <p><i>Acute malnutrition: Weight/ Height</i></p> <ul style="list-style-type: none"> < 80% <p><i>Chronic malnutrition: height for age</i></p> <ul style="list-style-type: none"> < 89% 	<p>Exit Criteria for nutrition support NSP</p> <ul style="list-style-type: none"> Birth – 5 years: gained sufficient weight to attain a growth curve in relation to his/her normal growth curve and maintains the curve for three consecutive months. > 5yrs – 18 years who attain normal growth curve according to the growth chart within the 6 months period on the NSP scheme. <p>Or Private Patients</p> <ul style="list-style-type: none"> Upward crossing of 2 or more centiles over a period of 1 month or 2 consecutive visits. MUAC >15cm in children < 5 years of age. WH >90%, HA >95%, 	
<p>Referral</p> <ul style="list-style-type: none"> NSP Scheme 	<p>Additional Requirements</p> <ul style="list-style-type: none"> Nutritionally complete age appropriate supplement. Access from local day hospital/ CHC 	

2.6 Complications

2.6.1 Chylothorax

Chylothorax is an uncommon post operative complication resulting in leakage of lymphatic fluid into the pleural space due to surgical disruption of the thoracic duct or increased venous pressure of one of its main tributaries resulting in increased pressure within the intrathoracic lymph system

Diagnosis

The following needs to be present in the pleural fluid:

- 1) Triglycerides > 1.1 mmol/L
- 2) Chylomicrons positive
- 3) Chylomicrons negative with a lymphatic fraction > 80%

Treatment

1) Dietary

- Bowel rest with total parenteral nutrition
- Fat free diet or high LCT diet: this type of diet should be followed in a clinical environment only due to the high risk of developing essential fatty acid (EFA) deficiency and should not be followed for more than 2 weeks.
- High MCT enteral nutrition: Use monogen

2) Surgical

3) Octreotide

Refer to Appendix 2 for the recommended treatment algorithm

2.6.2 Chylous Ascites

Chylous ascites is an accumulation of chyle in the peritoneal cavity due to obstruction or rupture of the peritoneal or reperiitoneal lymphatic glands, increased venous pressure or congestive cardiac failure.

Diagnosis (Paracentesis)

The following needs to be present in the fluid

- 1) Triglycerides 200mg/dl
- 2) Predominance of lymphocytes > 75%

Treatment

Bowel rest with total parenteral nutrition

2.6.3 Acute Myocarditis

Treatment

CCME:

- L-carnitine 5 – 15 mg/kg (max 1g) 6H(IV) or orally 25 mg/kg 6 – 12H (max 3g/day)
- Co-enzyme Q10 1 – 4 mg/kg daily oral
- Magnesium Sulphate IV 2 mmol/ml (max 10ml) 12H slow IV
- Vitamin E 50 – 1000IU < 3yr or 200 – 400IU >3yr

The CHD patient

Goal: To ensure that each patient with congenital heart disease attains/ maintains an optimal nutrition status.

To read the chart:
Follow the arrows

Assess patient using the following approach:

- A = Anthropometry
- B = Biochemistry
- C = Clinical
- D = Dietary
- Implement nutrition support where appropriate

Start Here

Anthropometric assessment determine patient's nutritional status & risk:

- Height MAC TSF %EHA HC Mid parental height
- MUAC Weight %EWA %EWH AMA Growth Velocity

Is there growth faltering or failing during last month or over 2 consecutive visits?

Yes

No

Monitor
3 month review

Assess dietary intake

- Complete 24 hour diet recall
- Food frequency
- Analyse where possible
- Is the intake appropriate according to the

Yes

Good intake:

- Encourage caregiver and child around good food intake.
- Advise caregiver around food based dietary guidelines [FBDG]

No

Poor intake:

- Encourage caregiver and child.
- Advise caregiver around food based dietary guidelines [FBDG]
- Promote small frequent meals x 3 and snacks 2 – 3 per day.
- Recommend energy & nutrient dense foods & drinks

Entry to Nutrition Support:
Calculate Dietary Requirements & recommend nutrition supplementation according to treatment modality (pre operative/post shunt or post definitive operation – cardiac repair)

Exit Nutrition Support when:

- Birth to 5 years – Normal growth curve RTCH following 3 months on NSP scheme.
- > 5 – 18 yrs: Normal growth curve RTCH 6 months on NSP scheme.
- Upward crossing of 2 or more centiles over a period of 1 month or 2 consecutive visits.
- MUAC >15cm in children < 5 years of age.
- WH >90%,
- HA >95%,

Biochemistry & Clinical Monitor the following:

- Urinary sodium losses
- Hb
- Infection

Provide sufficient energy & protein to support growth and weight gain.

Infants

- 3 – 4g/kg protein
- 120 – 150kcal/kg (max 170kcal/ml)

Children

- 2.g/kg protein
- 1.2 – 1.5 x RDA OR
- Schofield equation or WHO x 1.5 – 1.6 (combined activity & stress factor)

Paediatric Supplements available: Hospitalised

Infants

- Breast milk or
- 0.67kcal/ml RTU/H feed
- 1kcal/ml RTU/H feed (fluid restricted)

Children

- 1kcal/ml RTU/H nutritionally complete feed. (Standard)
- 1.5kcal/ml RTU/H nutritionally complete feed. (Fluid Restricted)

For additional energy required for both infants and children consider a super soluble CHO/fat powder.

Paediatric Supplements available: Discharged

Infants

- Breast milk with a breast milk fortifier if required

Children

- Enriched maize meal porridge
- Nutritionally complete age appropriate supplement.

Supplementation of complementary foods: Discharged

Infants

- Soluble CHO powder
- Long chain fat/oil

Children

- Soluble CHO powder
- Long chain fat/oil

Maximum duration 10 – 14 days

Requirement of LCT: 1g LCT per year of life up to a maximum of 4 – 5g LCT per day

Suitable foods for use in a low LCT diet

Food	Average Portion Size (g)	LCT per portion (g)
Breakfast Cereals		
Cornflakes	25	0.2
Frosties	20	0.1
Special K	20	0.2
Cocopops	20	0.2
Rice Crispiers	25	0.2
Weetbix	35 (1)	0.9
Bread		
White, large thin slice	35	0.4
Matzos	20	0.2
Crumpets Toasted	40(1)	0.4
Dairy Foods		
Reduced fat cottage cheese	50	0.7
Condensed milk, skimmed sweetened	50	0.5
Fish		
White hake fillet	100	0.7
Fish fingers	25 (1)	1.8
Tuna	100	0.5
Meat and Poultry		
Roast Turkey, light meat	70	1.0
Roast chicken, light meat	25	1.0
Roast lamb, lean	25	2.0
Roast beef, lean topside	45	2.0
Silverside, lean	40	2.0
Legumes, Pasta, Rice		
Baked beans in tomato sauce	200	1.0
Tinned spaghetti in tomato sauce	125	0.5
White rice, boiled	150	0.4
White pasta, boiled	130	1.0

Fortify skimmed milk with a glucose polymer.

Free Foods for a Minimal LCT Diet

All fruit, fresh, frozen or tinned (except olives and avocado)
 All vegetables fresh, tinned or frozen
 Sugar, honey, golden syrup, treacle, jam and marmalade
 Jelly and jellied sweets such as Jelly Tots, Jelly Babies, wine gums, lollie pops or fruit pastilles
 Boiled sweets, mints (not butter mints)
 Fruit sorbets (milk free), water ices and ice lollies
 Meringue, egg white
 Spices and essences
 Salt, pepper, vinegar, herbs, tomato sauce, most chutneys, Marmite, Oxo and Bovril
 Fruit juices, fruit squashes, bottle fruit sauces
 Fizzy drinks, lemonade, cola

3.1 Aim

The aim of these guidelines is to outline appropriate nutrition care practices in the dietary management of children with congenital heart disease.

Nutrition management usually aims to maintain and support optimal nutrition status.

3.2 Objectives

The objectives of these guidelines are to:

- Identify appropriate feeding practices in the paediatric cardiac patient.
- Identify appropriate routes of feeding in the paediatric cardiac patient.
- Identify appropriate nutrition care plan strategies for the maintenance and support of optimal nutrition status.
- Promote early and appropriate nutrition intervention with non-volitional nutrition support where oral feeding has failed in the paediatric cardiac patient.

3.3 Statement Regarding Promotion, Protection and Support of Exclusive Breastfeeding in infants with congenital heart disease

3.3.1 Breastfeeding

Exclusive breastfeeding should be encouraged for the first 6 months of life and up to the age of 2 years following the introduction of a variety of safe and healthy complimentary foods.

However, in some instances breastfeeding may not be possible and breast milk substitutes are required. Breast milk substitutes should be prepared to standards recommended by Codex Alimentarius.

3.3.2 Liquid Infant Formula

Liquid infant formula is sterile and does not contain any pathogenic organisms and as such does not present any potential source of infection. Wherever possible, in a hospital setting sterile liquid formula should be used. It is recommended that sterile infant feed liquids are used in all Neonatal and Paediatric Intensive Care /High Care Units, in addition in all infants considered to be immunocompromised and or where the safe preparation of powdered infant feeds may not be guaranteed.

3.3.3 Infant Feeds

Breast-feeding should be supported at all times. However there may be instances where specialized infant feeds (IF) are required and or breast milk is topped up with a breast milk substitute. However this should be provided under the guidance that a replacement feed should only be given if it is “acceptable, feasible, affordable and sustainable”.

4. Introduction

Congenital heart defects are classified into two broad categories: acyanotic and cyanotic lesions. The most common acyanotic lesions are ventricular septal defect (VSD), atrial septal defect (ASD), atrioventricular canal, pulmonary stenosis, patent ductus arteriosus (PDA), aortic stenosis and coarctation of the aorta. Congestive heart failure (CHF) is the primary concern in infants with acyanotic lesions. The most common cyanotic lesions include tetralogy of Fallot (TOF) and transposition of the great arteries.⁵

The reported incidence of congenital heart disease (CHD) is eight cases per 1000 live births. Up to 30% of infants with CHD have features of various genetic syndromes.

4.1 Features of Common Congenital Heart Defects⁵

4.1.1 Acyanotic Lesions

Ventricular Septal Defect (VSD)	Spontaneous closure in 30 –40% of cases in the first 6 months. The most common 15 –20%. Surgical repair recommended if infant exhibits failure to thrive, pulmonary hypertension
Atrial Septal Defect (ASD)	Often asymptomatic and 87% of secundum types close by age 4. Primary and sinus types require surgery
Atrioventricular canal	Presentation similar to that of the VSD.
Pulmonary Stenosis	May be asymptomatic or result in severe CHF.
Patent Ductus Arteriosus (PDA)	In premature infants, spontaneous closure or indomethacin-induced closure may occur. In term infant spontaneous closure is less likely. Recurrent pneumonia may occur. Surgical ligation is usually required.
Aortic Stenosis	May be asymptomatic. Valve replacement and anti-coagulation may be required.
Coarctation of the aorta	98% of cases occur at origin of left subclavian artery. Blood pressure is higher in arms than in legs. Surgical repair usually required between 2 and 4 years of age.

4.1.2 Cyanotic Lesions

Tetralogy of Fallot (TOF)	Most common CHD beyond infancy. Intermittent episodes of hyperapnoea, irritability, cyanosis. Surgical repair required with suitable anatomy.
Transposition of the great arteries	Incidence of about 5% in children with CHD. Late complications include pulmonary stenosis, mitral regurgitation, aortic stenosis, coronary artery obstruction, ventricular dysfunction and arrhythmias.

5. Anthropometry ^{2,6}

It has been reported that 52% of children with CHD were below the 16th percentile for height, 55% were below the 16th percentile for weight and 27% were below the 3rd percentile for both. Weight was more affected than height and boys tended to be more malnourished than girls. Stunting was more frequent in children below 2 years of age at 49%. Children with CHD tended to fall behind in weight-for-height, especially between 6 and 12 months of age.² Other studies found that acute and chronic malnutrition occurred in 33% and 64% respectively of hospitalized infants and children with CHD.¹

5.1 Assessment

- 1) Record weight, height and head circumference (below 3 years of age)
- 2) Plot on relevant growth chart. (Down's Syndrome charts and Gestational age charts should be used where appropriate)
- 3) If failing to thrive:
 - i) Calculate weight at 100% expected weight for child's current length/height
 - ii) Calculate ideal body weight (IBW) using the following equation:

$$\text{IBW} = (\text{Expected weight-for-height} + \text{expected weight-for-age} + \text{expected weight-for-head circumference} - \text{if the child is less than 3 years of age}) / \text{Present body weight}$$

The calculated ideal body weight will give you an indication of your aim, but always use current body weight when calculating requirements. To provide sufficient calories for catch up growth 1.2 – 1.5 DRV may be used in children >1 year of age

Refer to the clinical guideline on anthropometry.

5.2 Expected weight gain in children

Normal birth weight in the RSA is 3 kg for both sexes. There is some weight loss during the first 5-7 days, but this is normally regained by day 10-14. Expected weight gain should be as follows.

Table 1: Expected weight gain in healthy children ¹

Age	g/week	g/day	g/month
0-3 months	200	28.6	
3-6 months	150	21	
6-9 months	100	14	
9-12 months	50-75	7-11	
12-18 months	56	8	
18-24 months	42	6	
2-7 years			38
7-9 years			56-62
9-11 yrs			67-77
11-13yrs			85-110

Take note that normal weight gain would not normally be anticipated in a cardiac child and an average gain of 10 –20g / day should be aimed for in children < 6 months, 120 – 210g per week 6 – 12 months.

6. Biochemistry

Monitor biochemistry values and in particular consider urea, creatinine and sodium. If a child is failing to gain weight despite appropriate nutritional support, the following biochemical growth factors may be contributing and should be evaluated.

- High sodium losses in urine, normal or low serum sodium – a 24-hour urine sodium balance should be done to establish sodium balance. Sodium losses may be expected in the urine if the infant/child is on Lasix. Children under 1 year normally require 3 mmol/kg sodium to support growth. Sodium may be supplemented orally via hypertonic saline (1ml = 3mmol) which may be administered via NGT. If given orally it may cause nausea due to the saltiness. Additional salt should not be added to feeds.
- *Low potassium:* As a result of diuresis. Consider a K sparing diuretic.
- *Low HB:* (microcytic anaemia – check iron status and macrocytic anaemia – check folate and Vitamin B12 levels). Infants with cyanotic CHD may have a normal Hb but iron deficient. Check iron status using ferritin, red cell indices and total iron binding capacity.¹²
- *Infection:* An increase of 1 degree Celsius above normal temperature can result in a 10% increase in energy requirements. Monitor CRP.
- *Thyroid function:* deranged thyroid function may cause growth retardation
- *Acidosis:* may result in weight loss
- Serum zinc and selenium¹

Normal blood values for infants and children

	Unit	Age	Paediatric normal values
Potassium	mmol/l	<1mth	3.0-6.6
		>1mth	3.0-5.6
Sodium	mmol/l		132-142
Urea	mmol/l		2.5-6.5
Creatinine	mmol/l		
Haemoglobin	g/dl	1wk	11.0-25.2
		1month	12.0-21.8
		6month	10.0-15.0
		1-2yr	10.5-13.7
		4-5yr	11.1-14.7
		8-13yr	10.3-15.5
CRP	mg/l		0 - 10

7. Clinical

7.1 Pathogenesis of malnutrition in infants with CHD

Many children with CHD have increased metabolic rate. There is increased brain metabolism, cell number and over activity of the sympathetic nervous system. There is also an increased metabolic demand by specific tissues such as cardiac and respiratory muscle and those tissues, which are haematopoietic.^{1,2}

There is often an increase in respiratory rate and hypertrophied cardiac muscle which may use up to 20-30% of the total oxygen consumption compared to the normal 10%. Infections are common which may also impact on growth.^{1,2}

There are a number of factors associated with poor growth in infants with CHD of which nutrition is one.^{1,2}

Inadequate calorie intake may be as a result of:

1. Fatigue on feeding leading to low intake
2. Fluid restriction

3. Poor absorption
4. Increased metabolic expenditure
5. Early satiety
6. Anorexia
7. Frequent infections
8. Frequent use of antibiotics affecting gut flora. ¹

7.2 Mechanisms of Malnutrition in Children with CHD

7.2.1 Type of Cardiac Lesion

Cyanotic lesions usually result in reduced height and weight, whereas acyanotic lesions tend to affect weight rather than height. Children with left to right shunts tend to weigh less than cyanotic children and this may be due to the greater occurrence of pulmonary hypertension. In pulmonary stenosis and coarctation of the aorta, linear growth is usually more impaired than weight. Hypoxia and breathlessness is commonly seen in children with CHD and while the duration of the hypoxia may affect growth, the severity does not appear to affect tissue metabolism profoundly. ²

7.2.2 Increased Metabolic Rate

Increased metabolic rate is common especially if Congestive Heart Failure (CHF) is present. Energy expenditure usually comprises of five elements:

- a. Resting metabolic rate
- b. Physical activity
- c. Thermic effect of food digestion/metabolism
- d. Energy cost of new tissue synthesis
- e. Energy losses.

In children with CHD the most important difference is the higher energy cost for maintenance due to an increased metabolic rate, leaving little energy to be directed toward growth.

Immediate post-operative oxygen consumption and resting energy expenditure (REE) appears to be close to the predicted values in healthy children. Oxygen consumption in an infant with CHD and failure to thrive (FTT) is increased in comparison to infants with CHD without FTT. The same can be observed in severely malnourished children with CHD when compared to children with CHD and normal growth. Malnourished children have an abnormal body composition, which is higher in lean body mass and lower in fat tissue as manifested by reduced skinfold thickness measurements. The metabolic rate is affected by the fat content of the body and once the fat is depleted, lean body mass is more metabolically active and consumes relatively more oxygen resulting in an increased basal metabolic rate.

The following mechanisms explain this increased metabolic demand:

- a) Increased brain metabolism in undernourished children, with a twofold increase in energy expenditure
- b) Increased metabolism related to increased cell number rather than cell mass
- c) Over activity of the sympathetic nervous system
- d) Increased demand of certain tissues e.g. hematopoietic tissue, cardiac and respiratory muscle.

There is often an increased respiratory rate and hypertrophied cardiac muscle which may use up to 20-30% of the total oxygen consumption of the body compared to the normal 10%. Infections are common which also impact on growth. ^{1,2}

Infants with FTT due to ventricular septal defects (VSD) have been shown to have a 40% elevation in total energy expenditure (TEE). REE was found to be the same between the control and VSD group. The difference between REE and TEE was 2.5 times greater in the VSD group than the control indicating that their energy expenditure during activity is much greater. In patients with a VSD there is a significant mixing of venous and arterial blood reducing arterial oxygen saturation. Whilst at rest, this is not posing a problem and REE does not increase. However when active, oxygen delivery to tissue is resulting in anaerobic metabolism. This is ineffective and causes an increase in energy expenditure increasing TEE. Children with CHD in which there is CHF present, often present with an increased REE. This is the heart muscle having to work much harder in order to pump an adequate amount of blood against a greater opposing force. In contrast to VSD, this type of lesion leads to a more inefficient use of energy at all times including rest leading to an elevation in REE.^{2,6}

Frequent respiratory infections also lead to growth impairment. Systemic and respiratory illnesses increase body temperature and metabolic rate. Metabolic rate may increase up to 10% for each degree Celsius above normal 37.5 degrees Celsius.

7.2.3 Inadequate Caloric Intake

Inadequate caloric intake has been shown to be the most important cause of growth disturbances in children with CHD. Evidence indicates that caloric intake of CHD patients was 76% that of normal age matched controls. Inadequate caloric intake may occur as a result, secondary to the body's inability to utilize nutrients for growth as a result of anoxia, acidosis, malabsorption and the relative increase in nutrient requirements.

Hypoxia leads to both dyspnoea and tachypnoea during feeding, causing the child to tire easily reducing the quantity of food consumed. A child with CHD will typically eat in the following manner: a few minutes of sucking or chewing and swallowing, followed by a decrease in appetite, increased respiratory rate and sweating. The meal is often not finished and the ingested part may be vomited. Anorexia and early satiety may also be related to the side effects of drugs such as diuretics. A compressive hepatomegaly secondary to CHF in addition may reduce the gastric volume and increasing the potential for gastroesophageal reflux and aspiration.

CHF is also responsible for oedema and hypoxia of the gut with subsequent dysmotility and malabsorption. It has been speculated that gastrointestinal maturation and function are delayed due to chronic hypoxia. Protein losing enteropathy and steatorrhoea were two of the most common abnormalities. Ideopathic diarrhea is common and may be related to congestion of the mesenteric, splachnic and portal systems.

Disorders of carbohydrate metabolism are commonly seen in children with CHD. These children are found to have lower fasting glucose levels and elevated insulin secretion rates. This could be related to higher levels of circulating catecholamines or a switch from fatty acid -oxidation to glycolytic metabolism, which is inefficient and uses more available glucose. This raises the possibility that CHD children are chronically hypoglycaemic which could contribute to the fatigue they experience whilst feeding.^{2,6}

7.2.4 Weight at the time of operation

Children below 4.5kg have a high risk of intra-operative mortality. Children with PDA experience a marked improvement in both weight and height after surgical correction. Normal birth weight children with large VSD's and CHF show significant signs of improvement in weight, height and head circumference after early surgical closure occurring within 6 – 12 months after surgery. Different results were found for low birth weight children with CHD. For these children, head circumference, length and weight had limited postoperative increase and they never reached the norm.²

7.2.5 Prenatal factors

Traditional prenatal factors effecting growth play a more significant role once the cardiac defect is corrected. Among these factors, parental height, genetic factors, intrauterine factors and birth weight have been identified as the most important.

7.2.6 Medication

7.2.6.1 Inotropes

Vasoactive drugs are used to support cardiac output and blood pressure in patients with cardiac failure.¹³⁻¹⁶ Dopamine is used with the aim of decreasing afterload resistance against which the heart contracts. Dopamine is still commonly used in low doses (1 – 3ug/kg/min) to improve renal perfusion and urinary output although there is little evidence to suggest there is a subsequent improvement in renal function.¹⁴

Splanchnic perfusion may be compromised following cardiac surgery due to poor cardiac output. As a result an increase in metabolic demands to the splanchnic bed may be considered harmful if there is not committant increase in blood flow to the region causing gastric mucosal acidosis, even in the presence of dopamine infusion.^{14,15}

Dopamine results in local redistribution of blood flow in the splanchnic region however there is controversy surrounding whether or not blood flow and oxygen consumption in the splanchnic organs is always increased, remains unchanged or sometimes decreases. Some studies have found that in at least some of the subject's oxygen perfusion to the splanchnic bed is compromised.^{14,15} This may be more commonly seen in those patients who are receiving high doses of dopamine e.g. > 8 ug/kg/min/day.

However, in other clinical trials a dopamine infusion of 6 ug/kg/min does not effect hepatosplanchnic metabolism. In children following cardiac surgery dobutamine at 5 ug/kg/min resulted in improved splanchnic circulation and resulted in the reversal of intramucosal acidosis.¹⁴

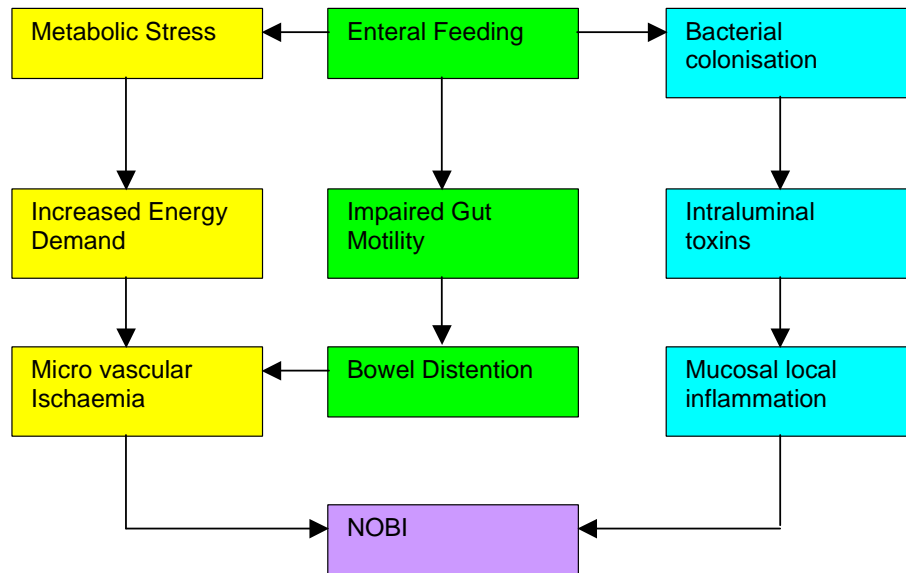
A recent study indicated, following the introduction of enteral feeds postoperative cardiac patient receiving inotropic support, indicated that cardiac index increased in conjunction with splachnic blood flow. This suggests that enteral feeding has a trophic effect on the gut. There were no signs of feeding intolerance with biochemical and metabolic indices indication that nutrients were utilized.^{17, 18}

Non-occlusive bowel injury (NOBI)

This is an uncommon but potentially fatal complication, which is related to the administration of enteral feeding in the acutely stressed patient. Criteria used to

diagnose non-occlusive bowel injury (NOBI) are patent mesenteric vascular bed in conjunction with the absence of bowel obstruction.

Potential mechanisms for NOBI ¹⁹



Administration of intraluminal nutrients may increase energy demands negatively impacting on enterocytes. The reasons for this are multifactorial, which include an imbalance in energy demand and supply, blood being shunted away from the mucosa and a defect in the mitochondrial function of the enterocyte. ¹⁹

Recent studies have shown that early enteral feeding is not harmful and may even have a trophic effect on the gut; the initiation of enteral feeding in patients with circulatory compromise who are at risk of splanchnic hypoperfusion (sepsis, cardiogenic shock, haemodynamic instability) may lead to deleterious changes in the structure and function of the gut. It is therefore imperative to monitor for ant signs of feeding intolerance and take early corrective measures. ¹⁹

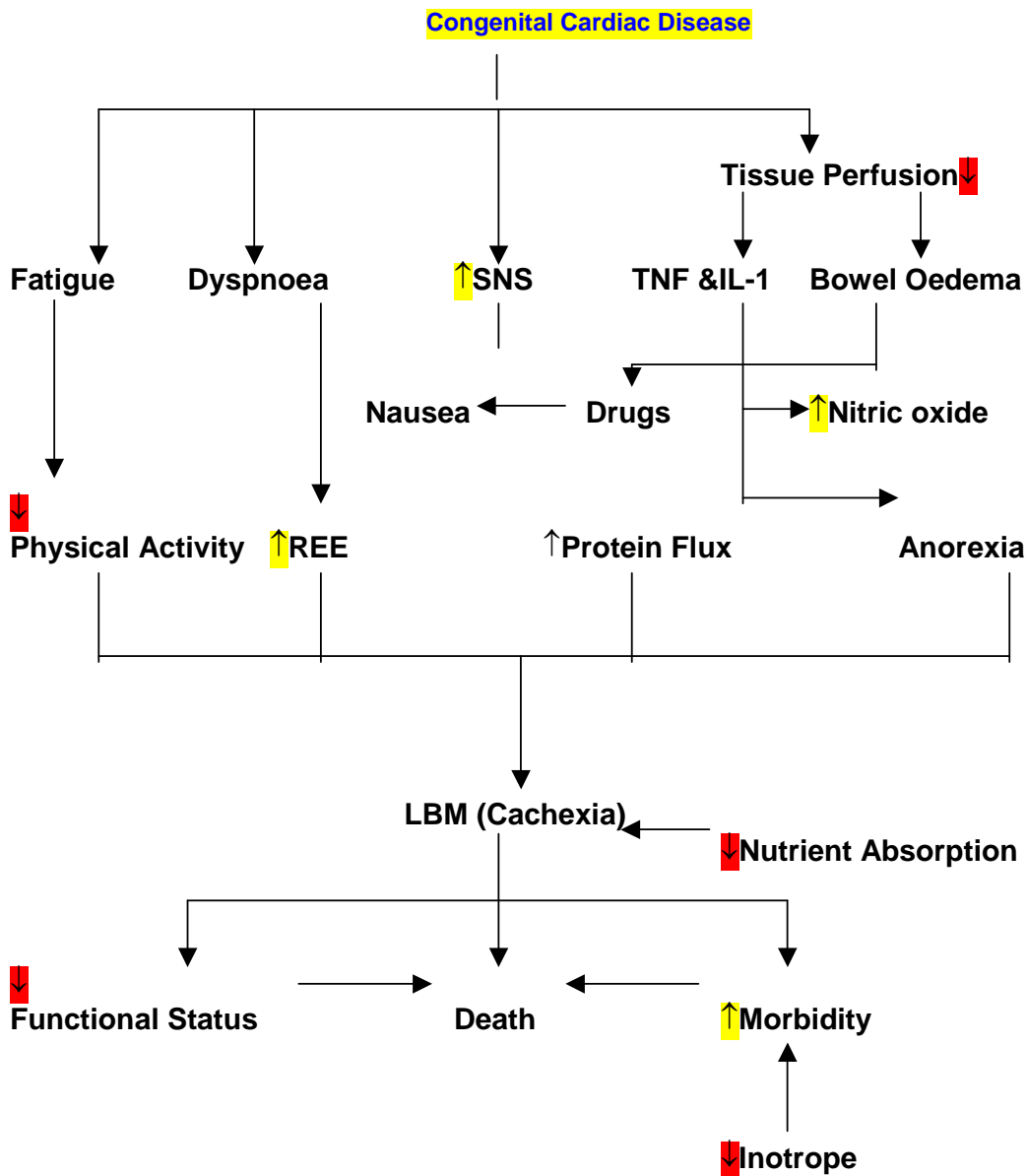
Nutrients have different effects on splanchnic flow during normal physiological conditions. Nutrients, which are found to be most capable of inducing mucosal hyperemia, are glucose and other carbohydrates, which is followed by proteins and peptides with amino acids being the least capable nutrient inducers, although glutamine, aspartate and glycine are the exceptions promoting vasodilation. ²⁰

7.2.6.2 Diuretics

Infants with CHD are often on diuretic therapy. Commonly used diuretics cause increased urinary sodium, potassium, chloride and calcium losses. Whereas baseline sodium requirement for a healthy preterm infant is 3 – 4mEq/kg per day, diuretic usage may increase this need to as high as 12mEq/kg/day. Similarly, the potassium requirement, which normally averages 2 – 4mEq/kg/day, will rise to 7 – 10mEq/kg/day under diuretic pressure. Because the primary deficit is chloride, sodium and potassium, it must be repleted as sodium chloride or potassium chloride. Infants who have persistent hyponatraemia exhibit poor growth. Infants receiving diuretics and electrolyte supplements may demonstrate wide swings in serum potassium levels. Urinary calcium loss with diuretics exacerbates an already tenuous balance and increases the risk of osteopaenia and nephrocalcinosis. ¹²

7.2.7 Cardiac Cachexia

Cachexia is a syndrome of tissue wasting, which was first described in chronic heart failure (CHF) by Hippocrates 2300 years ago, and is associated with significant morbidity and mortality. Many different mechanisms have been proposed for the pathogenesis of wasting in cardiac cachexia. It has been suggested that malnutrition, malabsorption, metabolic dysfunction, anabolic/catabolic imbalance and the loss of nutrients via the urinary and digestive tracts are important for the development of wasting, but the mechanisms of the transition from heart failure to cardiac cachexia are not known.^{3,4} Cachexia is defined as “accelerated loss of skeletal muscle in the context of a chronic inflammatory response. The development of cachexia in CHD is a dynamic process of non-intentional weight loss measured in a non-oedematous state.



Key: Congestive Heart Failure (CHF), Sympathetic nervous system (SNS), Tumour necrosis factor (TNF), Interleukin – 1 (IL 1), Resting energy expenditure (REE), Lean body mass (LBM), nitric oxide (NO)

8. Dietary

Adequate nutrition is important for infants with CHD. Many infants with CHD are able to breastfeed and gain adequate weight and in addition enjoy the other benefits of breastfeeding. In infants who are unable to gain sufficient weight through breastfeeding alone, breast milk can be supplemented with a super soluble carbohydrate and super soluble fat supplement to increase the feed concentration to fulfill requirements.⁶

A child with CHF will typically eat in the following manner

1. Sucking and swallowing for a few minutes.
2. Early satiety
3. Increase in respiratory rate and sweating.
4. The meal is often not finished and ingested portion will be vomited up.
5. Compressive hepatomegaly secondary to CHF may reduce the gastric volume and increase the risk of gastro-oesophageal reflux/ aspiration^{1,2}

8.1 General guidelines

1. Provide sufficient calorie and protein to facilitate weight gain
2. Avoidance of large fluid loads if fluid restricted
3. Monitor sodium requirement and intake
4. Electrolyte monitoring.

Dietary	
At each follow up a thorough nutrition history should be completed	
Components of a nutrition history include <ul style="list-style-type: none"> • Weight Change • Appetite • Satiety Level • Taste Changes/ aversions • Nausea/ vomiting • Bowel habits – constipation, diarrhoea • Chewing/ swallowing ability • Shortness of breath on feeding 	Components of a nutrition history include (cont) <ul style="list-style-type: none"> • Long-term disease(s) affecting absorption/use of nutrients • Dietary history – 24 hour recall/ food frequency • Use of vitamin/ mineral or nutritional supplements • Medications • Level of activity/ exercise • Ability to secure and prepare food • Over the counter medications, vitamins and herbal remedies.
Diet history <ul style="list-style-type: none"> • Review through 24-32hour dietary recall quarterly or at each follow up review. • Use in conjunction with food frequency. • Many patients may eat < 65% of RDI. 	Method of feed administration (employ a stepwise downward approach) <ul style="list-style-type: none"> • Offer smaller volumes and more frequent feeds orally • Give any unfinished feeds via naso-gastric tube if required • Give small frequent bolus feeds via naso-gastric tube • Top up small frequent daytime feeds with continuous overnight feeds via an enteral feeding pump • Give feeds continuously over 24 hours via an enteral feeding pump.

Fluid	<p align="center">Fluid Ranges</p> <p align="center">Age (years)</p> <p align="center">Premature 0-1 1-3 3-6 7-10 10-15</p> <p align="center">Weight</p> <p align="center">Premature < 2kg Neonates and infants 2 – 10kg</p> <ul style="list-style-type: none"> • 0 – 6 months • 6 – 12 months <p align="center">Infants and children 10 -20kg Children > 20kg</p> <p align="center">DO NOT ALTER MEDICALLY INDICATED FLUID RESTRICTION</p>	<p align="center">ml/kg actual weight</p> <p align="center">180-200 150 100 90 70 60</p> <p align="center">Fluid Volume per 24 hours</p> <p align="center">150ml/kg</p> <p align="center">150ml/kg 120ml/kg 1000ml + 50ml/kg over 10kg 1500ml + 20ml/kg over 20kg</p>
Energy	<p><u>Infants:</u></p> <p><u>Pre-operative or post shunt</u></p> <ul style="list-style-type: none"> • Ventilated: 90 – 100 kcal/kg • Non ventilated: 120 – 150kcal/kg (maximum 170 kcal/kg) <p><u>Post definitive operation (Cardiac Repair)</u></p> <ul style="list-style-type: none"> • Ventilated: 90 – 100 kcal/kg • Not ventilated: Schofield using abw + activity 1.2 + stress 1.5 – 1.6 <ul style="list-style-type: none"> • In sedated, ventilated children energy expenditure if often significantly reduced. • Aim to feed at 150kcal/kg and only increase to 170kcal/kg if other factors effecting growth have been excluded. 	<p><u>Children:</u></p> <p><u>Pre-operative or post shunt</u></p> <ul style="list-style-type: none"> • Ventilated: Schofield equation or WHO/FAO/UNU x 1.3 – 1.5 [No activity factor] • Non ventilated: 120 – 150% or DRV's <p><u>Post definitive operation (Cardiac Repair)</u></p> <ul style="list-style-type: none"> • Ventilated: Schofield equation or WHO/FAO/UNU x 1.3 – 1.5 [No activity factor] • Non ventilated: Schofield using abw + activity 1.2 + stress 1.5 – 1.6
Energy Supplementation	<ul style="list-style-type: none"> • <u>Infants:</u> • No additional energy should be required and breast milk and or standard ready to use/ hang infant formula [0.67kcal/ml] should be given. • If the patient is volume-restricted requirements breast milk may be supplemented with a human milk fortifier or carbohydrate/fat powder and or a ready to use/hang energy dense infant feed [1kcal/ml] should be given. • <u>Children:</u> • No additional energy should be required and a standard feed [1kcal/ml] should meet requirements in the volume prescribed. • If the patient is volume restricted an energy dense [1.5 kcal/ml] ready to use/hang feed should be given. • NB: • No powders or liquids e.g. oil should be added to a sterile ready to use feed. • If additional energy is required in non-ventilated children then flushes of energy boluses should be provided prior to a drink or feed including breast milk. • Recommendations for fat, protein and carbohydrate concentrations should not be exceeded. [See sections below] 	
Carbohydrate	<p><u>Glucose requirements</u></p> <ul style="list-style-type: none"> • > According to tolerance <p><u>Infants</u></p> <ul style="list-style-type: none"> • 8-9mg/kg/min [11.5g –12.9g/kg/day] • Max 12.5mg/min/kg [18g/kg/day] <p><u>Toddlers</u></p> <ul style="list-style-type: none"> • 7mg/kg/min [10g/kg/day] <p><u>Adolescents</u></p> <ul style="list-style-type: none"> • 4mg/kg/min [5.7g/kg/day] 	<ul style="list-style-type: none"> • The following concentrations of CHO per 100ml will be tolerated if a CHO/fat powder is used. • 10-12% carbohydrate concentrations in infants under 6 months (i.e. 7g from formula, 3-5g added) • 12-15% in infants aged 6months to 1 year • 15-20% in toddlers aged 1-2 years • 20-30% in older children

Protein	<p><i>Infants:</i> <u>Pre-operative or post shunt</u></p> <ul style="list-style-type: none"> • 9 – 11% of total energy • Up to 4 g/kg abw <p><u>Post definitive operation (cardiac Repair)</u></p> <ul style="list-style-type: none"> • 9 – 11% of total energy • DRV's <p><i>Children</i> Pre-operative, post shunt and post definitive operation (Cardiac Repair)</p> <ul style="list-style-type: none"> • Up to 2 g/kg abw 	<ul style="list-style-type: none"> • unless renal function is impaired
Fat	<p><i>Birth - < 5 years of age</i></p> <ul style="list-style-type: none"> • 40% NPE <p><i>Children > 5 years of age</i></p> <ul style="list-style-type: none"> • 30 – 35% NPE 	<ul style="list-style-type: none"> • <i>Adding Fat to feeds:</i> • Should be done as a last resort – rather add extra oil/ margarine to food. • Infants will tolerate a total fat concentration of 5 – 6 % [e.g. 5 – 6g per 100ml of feed]. • Children > 1yr will tolerate a fat concentration of 7% - concentrations above this may cause nausea/ vomiting. • It is recommended that a soluble carbohydrate/fat powder be used in bolus form over modulars such as oil and/or a glucose polymer
Micronutrients	<ul style="list-style-type: none"> • A daily multi-vitamin should be given e.g. Abidec® or Vidaylin® • Elemental Zn 1 – 3 mg/kg abw • Iron: 2mg/kg/day for prophylaxis 6mg/kg/day if Fe deficient 	<ul style="list-style-type: none"> • The multivitamin should contain folate, niacin, thiamine and B12 and vitamin E. • Do not supplement for longer than 2 weeks. • Do not supplement during the first 10 days post-operatively as it increases the risk of redox.

To recover from a downward spiral of decreased energy intake and increased energy requirements, catch-up growth is the goal for children with CHD and FTT.

8.2 Nutrition Strategies for Children with CHD and FTT²

1. Slow re-introduction of foods with high caloric content and of high nutritional value
 - 1kcal/mL (Infants) 1.5kcal/ml (Children)

Increase caloric intake by

 - Concentrating the feed
 - Increasing carbohydrate and fat content (Super soluble CHO/fat supplement)
2. Avoid large loads of non-nutritive fluids e.g. rooibos tea
3. Sodium restriction **only** where necessary (2.2 – 3mEq/day)
4. Electrolyte monitoring and supplementation where required – refer to biochemistry section

8.3 Type of Feed

Reports indicate that when dietary intervention is provided, including nutritional analysis and counseling. There is an increased mean intake from 90% to 104% of the RDA for calories and increased weight from 83.1% to 88.3% of ideal body weight. Parental education from a dietitian is important to optimize feeding.⁶

8.4 Method of Feed Administration

Oral intake in children with CHD is often impaired as they frequently tire during feedings and vomit a substantial amount of their intake. Continuous enteral feeding can result in a 32% mean increase in caloric intake.⁷

There are four proposed mechanisms for improved weight gain on continuous enteral feeds:

1. Improved caloric and nitrogen intake
2. Continuous infusion of feeds prevented gastric distention and vomiting
3. Improved absorption due to continuous saturation of the small intestinal absorptive sites
4. Reduced metabolic demands.

8.5 Iron supplementation

Infants who have cyanotic congenital heart disease have high iron requirements due to their expanded red cell mass. Persistent cyanosis increases the production of endogenous erythropoietin, resulting in secondary polycythemia, to improve oxygen delivery. The synthesis of each additional gram of hemoglobin requires an additional 3.4 mg of elemental iron. It is important to screen infants who have cyanotic CHD for iron deficiency using ferritin, red cell indices and total iron binding saturation as hemoglobin may be within normal range even though the infant is iron deficient. Minimum dose recommended is 2mg/kg actual body weight of iron.¹²

Post-operative iron supplementation has been shown to increase transferrin saturations and results in smaller decreases in ferritin level and results in a lower incidence of depleted iron stores.²⁴

Iron supplementation should not be given for the first 10 days post-operatively due to an increased risk of redox.

9. Complications

9.1 Chylothorax

Chylous pleural effusion, or chylothorax, is an uncommon early postoperative complication. The postoperative leakage of lymphatic fluid into the pleural space may result from the surgical disruption of the thoracic duct or one of its main tributaries resulting in increased pressure within the intrathoracic lymph system. The incidence of chylothorax is 2.5 – 4.7%.⁹

Chyle is an opaque milky white odourless fluid consisting of protein, fat, lymphocytes and electrolytes absorbed through the gut into the lymph channels of the gastrointestinal tract. The lymph channels drain into the cisterna chyli and then travel up the thoracic duct to re-enter the vascular system near the junction of the subclavian and left internal jugular veins. It is mainly long chain fatty acids that are absorbed from the intestines via lacteals and enter the circulation at the thoracic duct.¹⁰

Because chyle fluid originates in the gut and chyle flow increases dramatically after enteral intake, management of a patient with chylothorax has focused on altering

dietary intake. If this is not successful, other approaches include surgical management via thoracic duct ligation, pleurodesis or pleuroperitoneal shunt, more recently; Octreotide infusion has also been used.¹⁰

Buttiket and coworkers have defined chylothorax with the following parameters:

- a. Triglycerides > 1.1 mmol/L
- b. Chylomicrons (+)
- c. Chylomicrons (-) plus lymphocyte fraction > 80%
Present in the pleural fluid

When diagnosing chylothorax, cogniscance should be taken of previously poor enteral nutrition as this will decrease chylomicron and triglyceride levels in the pleural fluid. As a longer time to diagnosis is correlated with increased drainage duration, it may be useful to re-test pleural fluid after an enteral fat challenge.⁹

Dietary Management

Dietary management includes complete gut rest with parenteral nutrition, relatively fat free enteral nutrition, or very low long chain triglyceride (LCT), high medium chain chain triglyceride (MCT) enteral feeding. Medium chain fatty acids (6 – 12 carbon lengths) are absorbed directly into the portal system and do not enter the lymphatic system.

Adequate calories, fluid, protein and electrolytes must be provided regardless of feeding method. It is also important to provide enough essential fatty acids (linoleic and linolenic acid) to prevent essential fatty acid (EFA) deficiency. The general guide is to not give more than 1g of LCT per year of life up to a maximum of 4 to 5g LCT per day. (1) The American Academy of Paediatrics (AAP) recommends that at least 3% of daily calories come from EFA's. Others report adequate EFA if linoleic acid supplies 1 – 2% of total calories and linolenic acid supplies 0.54% of total calories.¹⁰

- 1) *Total parenteral nutrition*: Since the fat in TPN is delivered directly into the blood stream, it never enters the lymphatic system and therefore has no effect on the thoracic duct.
- 2) *Low fat nutrition*: EFA deficiency has been found in infants and children receiving a fat free diet for greater than three weeks. Symptoms of deficiency are scaly skin, delayed wound healing, poor growth, diarrhoea, platelet dysfunction and hair loss. (10) It is important to monitor infants and children on the low fat diet to prevent EFA deficiency by providing EFA in the form of walnut oil 2 – 3% of total calories per day.
- 3) *High MCT enteral nutrition*: Monogen is a nutritionally complete, low fat, whole whey protein, powdered feed containing 93% of fat as MCT and 7% of fat as LCT provided by walnut oil. It was designed for infants and children with lipid and lymphatic disorders. Monogen also has a lower osmolality than most elemental, fat free formulas, which improves gastrointestinal tolerance.¹¹

Surgical Management

Surgical management of chylothorax remains controversial and quite variable. Some centers ligate the thoracic duct after a certain number of weeks of drainage, while

others use volume of drainage and rate of decline in drainage as determining factors to plan surgery. Some hospitals use pleurodesis, the stripping of the pleura off the surface of the lung, to control fluid drainage. Others use irritant agents (such as talc) to sclerose the pleura to the lung. Also reported is the placement of a pleuroperitoneal shunt to shift the fluid from the thoracic cavity to the abdominal cavity where it can be reabsorbed. This approach prevents the problem of electrolyte and protein loss. This technique could be successful or at least minimally helpful depending on the volume of fluid shunted but could create an ascites like picture in the patient.¹⁰

Octreotide

Octreotide is known to decrease splanchnic, hepatic and portal blood flow, thereby decreasing the volume of lymph produced and, ultimately, thoracic duct flow. It inhibits the absorption of triglycerides and decreases acetylcholine release in the gut. Acetylcholine is known to increase lymph flow therefore reduced acetylcholine would result in reduced lymph flow. A case report suggests 1 – 4mcg/kg/hr infusion when used in conjunction with dietary management. Anecdotal reports of side effects include vomiting and other gastrointestinal issues related to decreased blood flow.¹⁰ Chan et al. postulate that Octreotide may be more beneficial for use in mild to moderate chylothoraces.⁹

Recommended infusion rate: Octreotide infusion 1 – 4 mcg/kg/hr when used in conjunction with dietary intervention.¹⁰

Refer to treatment Appendix 2. 3

9.2 Chylous Ascites

Chylous ascites, an uncommon disease usually caused by obstruction or rupture of the peritoneal or retroperitoneal lymphatic glands, is defined as the accumulation of chyle in the peritoneal cavity.

Many pathological conditions can result in this disease, including congenital defects of the lymphatic system, non specific bacterial, parasitic and tuberculous peritoneal infection, blunt abdominal trauma and surgical injury.²¹

The diagnosis is confirmed with paracentesis The fluid has:

Elevated triglycerides 200mg/dl

Predominance of lymphocytes > 75%^{22, 23}

Recommended treatment is conservative management with bowel rest, total parenteral nutrition (TPN) in combination with Somatostatin therapy. Studies have shown that fasting together with TPN can decrease the lymph flow in the thoracic duct dramatically from 220ml/ (kg.h) to 1 ml/ (kg.h).²¹

Somatostatin has been shown to decrease intestinal absorption of fats, lower triglyceride concentrations in the thoracic duct and attenuate lymph flow in the major lymphatic channels. In addition it also decreases gastric, pancreatic and intestinal secretions, inhibits motor activity of the intestine, slows the process of intestinal absorption and decreases splanchnic blood flow, which may further contribute to decreased lymph production. It has also been speculated that Somatostatin inhibits lymph fluid excretion through specific receptors found in normal lymphatic vessel of the intestinal wall.²¹

No specific recommendations regarding the optimum dose of Somatostatin have been identified in the evidence.

9.3 Acute Myocarditis

CCME stands for L-Carnitine, Coenzyme Q10, Magnesium and Vitamin E. Carnitine, coenzyme Q10, magnesium and vitamin E interact in the mitochondrial generation of energy.²⁶

Animal and human trials have shown specific nutrient deficiencies in the failing myocardium. There is a reduction in L-carnitine, coenzyme Q10 and creatinine, which are important cofactors in energy metabolism. Deficiencies in carnitine and taurine have been associated with dilated cardiomyopathy. Antioxidant endogenous defences, including vitamin E and selenium, have also been shown to be reduced. Myocardial oxidative stress is increased during ischaemia-reperfusion.¹⁷

L-carnitine increases ATP generation via its effect on beta oxidation, as well as its role in the removal of acetyl units from the mitochondria, which is important as accumulation of acetyl units is known to inhibit various parts of the respiratory process. It also acts as a vasodilator and has an increased ability to sustain cardiac contraction.²⁶

Coenzyme Q10 is a rate-limiting carrier for the electron flow in the mitochondrial respiratory change, and is also an endogenous antioxidant protecting membranes from oxidation. A reduction of up to 50% in myocardial coenzyme Q10 has been documented in heart failure.¹⁷

Magnesium is considered to be a “natural” calcium channel blocker and it can also mitigate the cardio toxic effects of catecholamines.²⁶

Vitamin E plays a role in the inhibition of platelet aggregation and is able to block redox cycling of catecholamines resulting in diminution in abnormal sympathetic stimulation of the heart.²⁶

Folic acid supplementation has been shown to be essential in the prevention of coronary heart disease, but this type of supplementation is not an issue in acute illness.¹⁷

Recommended CCME:

- L-carnitine 5 – 15 mg/kg (max 1g) 6H(IV) or orally 25 mg/kg 6 – 12H (max 3g/day)
- Co-enzyme Q10 1 – 4 mg/kg daily oral
- Magnesium Sulphate IV 2 mmol/ml (max 10ml) 12H slow IV
- Vitamin E 50 – 1000IU < 3yr or 200 – 400IU >3yr

10. Summary

Congenital heart disease results in growth failure and malnutrition. Aggressive nutrition support is required to promote good nutrition status. Entry and exit criteria for nutrition support have been delineated in the summary tables and should be used as a guide for the appropriate nutrition management of children with congenital heart disease.

All children with growth failure should be provided with nutrition support to attain optimal nutrition status and support linear growth. Children awaiting surgical intervention should receive nutrition support.

All children with growth failure should be referred to the Nutrition Supplementation Programme (NSP) and/or motivations written to medical aids to provide appropriate supplements on a regular basis. The enrichment of food should be encouraged with small frequent meals and snacks.

11. Appendix 1: Energy Calculations

Table 1: Selected Dietary Reference Values (DRV's) for Infants and Children requiring Oral/Enteral Nutrition ¹

Age	Weight (kg)	KJ/kg/day	Kcal/kg/day	Protein g/kg/day
Males				
0 – 3months	5.1	420 – 480	100 – 115	2.1
4 – 6	7.2	400	95	1.6
7 – 9	8.9	400	95	1.5
10 –12	9.6	400	95	1.5
1 – 3 years	12.9	400	95	1.1
4 – 6	19.0	380	90	1.1
7 – 10		8240/day	1970/day	28.3g/day
11 – 14		9270/day	2220/day	42.1g/day
15 – 18		11510/day	2755/day	55.2g/day
Females				
0 – 3 months	4.8	420 – 480	100 – 115	2.1
4 – 6	6.8	400	95	1.6
7 – 9	8.1	400	95	1.5
10 –12	9.1	400	95	1.5
1 – 3 years	12.3	400	95	1.1
4 – 6	17.2	380	90	1.1
7 – 10		7280/day	1740/day	28.3g/day
11 – 14		7920/day	1845/day	42.1g/day
15 - 18		8830/day	2110/day	45.4g/day

Table 2: Schofield Equation for Calculating Resting Metabolic Rate (RMR) – Kcal/day ^{1,29}

Age (yr)	Male	Female
< 3	$0.167(W) + 1517.4(H) - 617.6$	$16.252(W) + 1023.2(H) - 413.5$
3 – 10	$19.59(W) + 130.3(H) + 414.9$	$16.696(W) + 161.8(H) + 371.2$
10 –18	$16.25(W) + 317.2(H) + 515.5$	$8.365(W) + 465(H) + 200.0$
> 18	$15.057(W) + 10.04(H) + 705.8$	$13.623(W) + 283(H) + 98.2$

Table 3: FAO/WHO/UNU kcal/day ^{1,29}

Age (yr)	Male	Female
3 – 10	$22.7 (W) + 495$	$22.5 (W) + 499$
10 - 18	$17.5 (W) + 651$	$12.2 (W) + 746$

PHYSICAL ACTIVITY FACTORS ¹	
ACTIVITY	ACTIVITY FACTOR (AF)
• Sleeping (ICU, Sedation and muscle relaxation)	1.0
Hospitalized	
• Non Ambulant	1.2
• Ambulant	1.3
At Home	
• Relatively inactive	1.4
• Very active	1.9
STRESS FACTORS	
DISEASE	STRESS FACTOR
Trauma	
• Little (long bone fracture)	1.2
• Central Nervous System	1.3
• Moderate to severe (multiple)	1.5
Sepsis	
• Moderate	1.3
• Severe	1.6

12. Metropole Paediatric Working Group:

1. Principal Author: Sonja Stevens
2. Working Group members: Nadia Bowley [Netcare], Gina Stear [PPD], Laurentia van Wyk [TBH]; Elisna van Wyk [TBH]; Luise Marino [RXH]; Nazneen Osmany [TBH]
3. Ad Hoc Reviewer Claudia Schubl [TBH]; Shihaam Cader [RXH], Melanie Davids [RXH], Bernadette Saayman [RXH]; Tamlyn Lippiat Moll [RXH]; Carmen van Zyl [RXH]
4. Clinical Reviewer: Dr S Vosloo
Dr H Pribut
Dr. J Lawrenson
Dr. S Shipton

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14. Paediatric Working Group Guidelines: Developers Summary

Scope and Purpose

The Guidelines for Congenital Heart Disease have been developed by the Western Cape Paediatric Nutrition Working Group in response to the need for evidence-based guidelines with respect to the nutrition management of Congenital Heart Disease.

The aim of this Guideline is to provide an evidence based nutrition management resource tool, which may be used by health professionals involved in the prescription and supply of nutrition support to infants or children with Congenital Heart Disease.

This Guideline uses an “A, B, C, D” approach e.g. Anthropometry, Biochemistry, Clinical and Dietary, to provide a step by step reference as to how to approach nutrition support.

These guidelines outline nutrition support in children with Congenital Heart Disease from the ages of 0 – 18 years of age. They are not meant to be prescriptive and there may be individual case variations.

Stakeholder Involvement

Members of the Paediatric Working Group are outlined in table 1:

Table 1: Paediatric Working Group Members and Reviewers

	Principal Author	Affiliations
Full Time Members		
Sonja Stevens	◆ Cardiac Disease	Dietitian, Netcare Christian Barnard Hospital
Lourentia van Wyk	◆ Diabetes [May 2007]	Department of Dietetics, Tygerberg Hospital
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Elisna van Wyk	◆ Pre Term Infants	Department of Dietetics, Tygerberg Hospital
Luise Marino	◆ Refeeding Syndrome ◆ Total Parenteral Nutrition ◆ Liver Disease ◆ GORD	Department of Dietetics, Red Cross War Memorial Children's Hospital
Gina Stear		Dietitian, Private Practice
Nadia Bowley		Dietitian, Netcare Regional Office N1 City
Ad Hoc Members		
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Clinical Reviewers		
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Dr. L Cooke	◆ Liver Disease ◆ GORD	Ambulatory Medicine, Tygerberg Hospital
Dr. E Nel	◆ Liver Disease ◆ GORD	Department of Gastroenterology, Tygerberg Hospital
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Dr. J Lawrenson	◆ Cardiac Disease	Department of Cardiology, Red Cross War Memorial Children's Hospital
Dr. S Vosloo	◆ Cardiac Disease	Private Cardio Thoracic Surgeon, Netcare:

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Prof. Hartley	♦ Oncology	Department of Oncology, Red Cross War Memorial Children's Hospital
Prof H Rode	♦ Short Bowel Syndrome	Department of General Surgery, Red Cross War Memorial Children's Hospital
Dr. Kaposky	♦ Short Bowel Syndrome	Department of General Surgery, Red Cross War Memorial Children's Hospital

Rigour of Development

A Pubmed search was completed using key words such as “nutritional management, Congenital Heart Disease, Chylothorax, Chylous Ascites”. Table 1 was used to define the type of articles desired. 52 articles were identified using the key words. The search was narrowed to include papers graded as being 1+++ to 2+ levels of evidence. [If this was not available change to which ones were included and rationale – e.g. consensus papers etc.]

Grading of levels of evidence (LOE) according to the Scottish Intercollegiate Guideline Network (SIGN) 2000

Grading	Level of evidence
1+++	High quality meta analyses, systematic reviews of RCT's or RCT's with very low risk of bias
1+	Well conducted meta analyses, systematic review of RCT's or RCT's with low risk of bias
1-	Meta analyses, systematic reviews of RCT's or RCT's with a high risk of bias
2++	High quality systematic reviews of case controlled or cohort studies
2+	Well conducted case control or cohort studies with a low risk of confounding, bias, or chance and a moderate probability that the relationship is causal
2-	Case control or cohort studies with a high risk of confounding, bias or chance and a significant risk that the relationship is not causal
3	Non-analytical studies e.g. case reports, case series. Evidence from non analytical studies e.g. case reports, case series
4	Evidence from expert opinion

The principle author was responsible for compiling the Congenital Heart Disease guideline, which was circulated amongst members of the working group in addition some of the ad hoc members.

All guidelines went through a process of first to third drafts. The recommendations within the guidelines were drafted following a review of the literature and discussions within the group.

All benefits and potential harm of the nutrition recommendations within the guidelines have been discussed and reviewed by the panel at length. The recommendations provided within the text and summary tables are referenced and evidence based.

This guideline has been reviewed by Dr Vosloo, Dr Pribut, Dr Lawrensen and Dr Shipton are considered to be experts in their field. Comments received have been incorporated into the clinical guidelines.

This guideline will be reviewed in 2008 and updated accordingly.

Clarity and Presentation

The format of this clinical guideline aims to direct the health professional through a logical Nutrition Care Plan approach using A, B, C, D e.g. Anthropometry, Biochemistry, Clinical and Dietary using a series of summary tables, which can be used as a quick reference abridged version for the key recommendations. In addition to these tables the full text may be consulted as required.

A variety of management options have been present targeting clients within the Public and Private Health Care sector. The guideline provides a stratified management approach and identifies current nutrition support systems through which they could be implemented.

Applicability

The working group did not perceive any potential barriers as all nutrition support strategies are currently available within Public and Private Health Care centres and are available on national tenders. All cost implications have been considered and the most cost effective nutrition management strategies have been recommended.

Within the Nutrition Care Plan Summary Tables appropriate review processes have been identified. In addition all tools are presented with an audit process.

Editorial Independence

The principal author, working group and or reviewers did not receive any funding to complete these guidelines and no conflicts of interest are recorded by the team.