

Nutrition in the Paediatric Liver Patient

Cape Town Metropole Paediatric Interest Group

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0. Paediatric Working Group Guidelines: Developers Summary

Scope and Purpose

The Guidelines for Liver 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 this condition.

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 liver 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 Liver 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

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Rigour of Development

A Pubmed search was completed using key words such as liver disease, biliary atresia, liver transplant, acute liver failure, chronic liver disease and children. Table 1 was used to define the type of articles desired. Thirty-eight articles were identified using the key words. The review include papers graded as being grades 1 and 2 levels of evidence.

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 Liver Disease Clinical 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 clinicians who are considered to be experts in their field. Comments received have been incorporated into the clinical guidelines.

This guideline will be reviewed in 2009 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.

1. Glossary

Term	Definition
AMA	Arm muscle area [requires MUAC & TSF to calculate]
DRV's	Dietary reference values [Appendix 1]
RDI	Recommended daily intake
EFA	Essential fatty acids
% EWA	Percentage estimated weight for age
% EHA	Percentage estimated height for age
% EWH	Percentage estimated weight for height
ESLD	End stage liver disease
HA	Height age <i>Is this height-for-age or height age?</i>
HE	Hepatic encephalopathy
IMCI	Integrated management of childhood illness
IMCI: Not Growing Well	<p><i>Severe Malnutrition:</i></p> <ul style="list-style-type: none"> ▪ Very low weight < 60% EWA. ▪ Visible signs of severe wasting ▪ Oedema on the feet <p><i>Not Growing Well:</i></p> <ul style="list-style-type: none"> ▪ Low weight < 3rd centile ▪ Poor weight gain - gaining weight but curve flattening or ▪ Mother reports weight loss. <p><i>Growing Well:</i></p> <ul style="list-style-type: none"> ▪ Not low weight and ▪ Good weight gain.
LCPUFA	Long chain polyunsaturated fatty acids
MCT	Medium chain triglycerides
NPE	Non-protein energy
MUAC	Mid upper arm circumference [6 months – 5 years of age] <ul style="list-style-type: none"> ▪ > 15cm normal ▪ >11.5cm - <14.5cm moderately malnourished ▪ <11.5 cm [<-3SD] severely malnourished
MAC	Mid arm area circumference
Schofield Equation	Predicting estimated energy requirements [Appendix 1]
NSP	Nutrition supplementation programme
Growth faltering (NSP Definition):	<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 disease, 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	<p>a) <i>Successful:</i></p> <ul style="list-style-type: none"> • <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 chart within the 6 months period on the scheme <p>b) <i>Unsuccessful:</i></p> <ul style="list-style-type: none"> • <i>Birth – 5 years:</i> 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 • <i>>5 - < 18 years:</i> who do not attain a normal growth curve according to the growth chart with in the 6

	<p>months period.</p> <p>c) <i>Defaulter:</i></p> <ul style="list-style-type: none"> ▪ <i>Birth – 5 years:</i> Failure to attend the clinic for a period of three consecutive months. • <i>> 5 - <18 years:</i> 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. <p><i>** Re-entry:</i></p> <ul style="list-style-type: none"> • <i>UNSUCCESSFUL</i> and <i>DEFAULT</i> cases <i>MAY NOT</i> be re-entered onto the programme. • <i>SUCCESSFUL</i> cases <i>MAY</i> be re-entered onto the programme according to entry criteria.
SD	<p>Standard Deviations used to determine moderate to severe malnutrition:</p> <ul style="list-style-type: none"> • 0 - <-1 Z scores Normally Nourished • >-2 – -3 Z scores Moderately Malnourished • >-3SD Severely Malnourished
TSF	Tricep Skinfold Thickness
WA	Weight age
WH	Weight for height
Waterlow Criteria (WHO)	<p>Used to determine malnutrition:</p> <p><i>Acute malnutrition: Weight-for-height/ length</i></p> <ul style="list-style-type: none"> • Normal WH >90%, • Mild 81% - 90%, • Moderate 70% - 80%, • Severe <70%. <p><i>Chronic malnutrition: Height-for-age</i></p> <ul style="list-style-type: none"> • Normal >95%, • Mild 90 –95%, • Mild – moderate 85% to 89% • Severe < 85%
Gomez Classification	<p><i>Acute malnutrition: 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%
GIT/ GI tract	Gastro-intestinal tract
CHO	Carbohydrate
IBM	Inborn error of metabolism
NPE	Non-protein energy
U&E	Urea, creatinine, sodium, potassium
ALT	Alanine aminotransferase
AST	Aspartate aminotransferase
ALP	Alkaline phosphatase
GGT	Gamma glutamyltransferase
INR	International normalisation ratio
FBC	Full blood count
Hb	Haemoglobin
WCC	White blood cell count
NG	Nasogastric
PEG	Percutaneous endoscopic gastrostomy
CHC	Community Health Centre
Mg++	Magnesium
PO ₄	Phosphate
K+	Potassium
Na+	Sodium
IV	Intravenous
IM	Intramuscular
RDA	Recommended daily allowance
DHA	Docosahexanoic acid
AA	Arachidonic acid
EBM	Expressed breastmilk
FBDG	Food based dietary guidelines

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

Summary Nutrition Recommendations: Acute Liver Failure

Acute Liver Failure	
Overview	<ul style="list-style-type: none"> In rapid presentation infants and children are usually moderately – well nourished. Management is based on maintaining nutrition status. If child survives liver function should return to near normal.
Energy	<ul style="list-style-type: none"> RDI [Appendix 1: Table 1] Do not usually require additional energy during an acute episode.
Protein	<ul style="list-style-type: none"> Not restricted
Fat	<ul style="list-style-type: none"> Not restricted
Vitamins	<ul style="list-style-type: none"> Vitamin K may be indicated if there is a deranged coagulopathy. However, the administration of other fat-soluble vitamins is not advised.

Acute Fulminant Liver failure

Overview

- Fulminant failure is difficult to define: as it occurs as an acute episodes arising from a previously undiagnosed chronic disease e.g. inborn error of metabolism or as a result of ingestion of a toxin or poison.
- Mitochondrial disorders of metabolism should be excluded.

Anthropometry

Complete on admission & weekly until discharge

<i>Determine:</i>	<i>Plot weight and length/ height using:</i>	<i>Calculate</i>
<ul style="list-style-type: none"> Length/ Height (m) Weight (kg) MUAC TSF Head Circumference < 3 years of age 	<ul style="list-style-type: none"> Appropriate growth charts. Weight may not be useful when there is gross ascites 	<ul style="list-style-type: none"> HA/ HFA WA WH % EWA % EWH % EHA

Entry Criteria for nutrition support during hospitalisation based on anthropometry

- Poor oral intake
- Weight loss or growth failure during hospitalisation
- Nutrition risk score > 6
- Growth curve flattens or drops over two consecutive months (NSP definition)
- < 5th or 3rd centile
- < 80% % EWH
- <-2 or -3 ZD
- Downward crossing of 2 or more centiles

Biochemistry

- Complete daily until acute episode has resolved.
- Once in recovery complete x 2 week until discharge
- [Follow Appendix 2: King's College Investigation protocol]

Monitor the following

- Urea, creatinine, sodium, potassium
- Calcium, magnesium and phosphorus
- Ammonia
- Glucose
- Albumin, ALT, GGT, Bilirubin [conjugated, unconjugated]
- INR & Fibrinogen
- Hb, platelets, WCC

Clinical		
Acute presentation associated with:		
<ul style="list-style-type: none"> Severe liver impairment associated with hepatocellular necrosis. Fulminant liver failure is usually associated with encephalopathy 		
Encephalopathy in acute liver failure		
Stage	Clinical	Signs
<ul style="list-style-type: none"> Stage 0 	<ul style="list-style-type: none"> Subclinical encephalopathy 	<ul style="list-style-type: none"> Abnormalities in psychometric testing
<ul style="list-style-type: none"> Stage 1 	<ul style="list-style-type: none"> Subtle behaviour changes Altered sleep patterns – switching night to day sleep Shortened attention spans 	<ul style="list-style-type: none"> Poor feeding Vomiting Incoordination Fine tremor Trivial lack of awareness Shortened attention span
<ul style="list-style-type: none"> Stage 2 	<ul style="list-style-type: none"> Personality changes Drowsiness/ lethargy Disorientation Agitation 	<ul style="list-style-type: none"> Asterixis Ataxia Hypertonia
<ul style="list-style-type: none"> Stage 3 	<ul style="list-style-type: none"> Temporal and spatial disorientation Delerium, hallucinations Mania Stupor, seizures 	<ul style="list-style-type: none"> Hyperreflexia Babinski's reflex Somnolence to semistupor Responsive to stimuli
<ul style="list-style-type: none"> Stage 4 	<ul style="list-style-type: none"> Comatose Semi- or unconscious 	<ul style="list-style-type: none"> Decorticate and decerebrate posturing Opisthotonus Ocular palsies Cardio respiratory arrest
Clinical Signs usually present during liver disease in children:		
<ul style="list-style-type: none"> Clubbing Jaundice Oedema Large abdomen Hepatomegaly 	<ul style="list-style-type: none"> Splenomegaly Ascites < Muscle bulk < Skinfold thickness Vasodilatation 	
Supportive Medical Management of Fulminant Liver failure		
<ul style="list-style-type: none"> Sedation: No sedation for procedures. Monitor: neuro obs 4 – 6 hrly, gastric pH [>5], blood glucose [$> 4\text{mmol/kg}$]; acid/ base electrolytes, PT, PTT Fluid balance: 75% maintenance – maintain circulating volume with colloid/ fresh frozen plasma Coagulation support: (fresh frozen plasma) Drugs: Vitamin K (2 – 10mg/d orally or IV) more is required if the INR is extremely deranged, sucralfate (2- 4g/day), lactulose (5 – 20ml/day), N-acetyl cysteine – IV (70mg/kg/6 hours), +/- broad spectrum antibiotics Enteral feed: enteral feed (aim for 1 – 2g/kg protein per day depending on age), parenteral nutrition may be occasionally required. 		
Dietary		
Overview of pathophysiology		
Potential problems requiring nutrition intervention occur when there is a disturbance of normal metabolic functions of the liver including:		
<ul style="list-style-type: none"> Glucose homeostasis Protein synthesis 		
Diet Therapy includes management of:		
<ul style="list-style-type: none"> Hypoglycaemia for poor glycogen storage Reduction in protein synthesis especially albumin exacerbating ascites. 		
At each follow up a thorough nutrition history should be completed		
<ul style="list-style-type: none"> Diet history Review through 24hour dietary recall quarterly or at each follow up review in conjunction with food frequency. 		
Fluid		
Patients may be fluid restricted to 60 – 75% of volume if cerebral oedema is a concern.		
Age (years)	ml/kg actual body weight	
Premature	180-200	
0-1	150	
1-3	100	
3-6	90	
7-10	70	
10-15	60	

Energy Expenditure & Requirements

Energy expenditure:

- In sedated, ventilated children energy expenditure is often significantly reduced.
- Caution should be taken not to overfeed in these children as they have an increased risk of metabolic and clinical complications

Infants:

- Ventilated: 90 – 100 kcal/kg
- Non ventilated: 100 – 120kcal/kg

Children:

- Ventilated: Schofield equation or WHO/FAO/UNU x 1.3 – 1.5 [Stress factor; No activity factor]
- Non ventilated: Schofield equation or WHO/FAO/UNU x 1.7 – 1.8 [Combined activity and Stress factor] [Appendix 1: Tables 2, 3 and 4]

Recommendations for energy density of feeds

Infants:

- No additional energy should be required breastmilk and or standard ready to use/ hang infant formula [0.67kcal/ml] may be given.
- If the patient is volume-restricted breastmilk may be supplemented with a carbohydrate and fat powder or a ready to use/hang energy dense infant feed [1kcal/ml] may be given.

Children:

- No additional energy should be required and a standard feed [1kcal/ml] should meet energy requirements in the volume prescribed.
- If a 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 boluses flushes of super soluble fat and carbohydrate powder should be given prior to a drink or feed (including breastmilk).
- Recommendations for fat, protein and carbohydrate per age group concentrations should not be exceeded. [See sections below]

Carbohydrate Recommendations

Glucose requirements

- > According to tolerance

Infants

- 8-9mg/kg/min [11.5g –12.9g/kg/day]
- Max 12.5mg/min/kg [18g/kg/day]

Toddlers

- 7mg/kg/min [10g/kg/day]

Adolescents

- 4mg/kg/min [5.7g/kg/day]

NB: Always increase glucose gradually and according to tolerance.

The following concentrations of CHO per 100ml will be tolerated if a glucose polymer is used:

- Infants under 6 months: 10-12% carbohydrate concentrations (i.e. 7g from formula, 3-5g of glucose polymer added)
- Infants 6months to 1 year: 12-15%
- Toddlers 1-2 years: 15-20%
- Older children > 2 years: 20-30%

Form of CHO

- CHO's in the form of glucose polymer should be used instead of disaccharides as they have a lower osmotic effect.
- It is recommended that glucose polymer be added in increments of 1g per kg per 100ml per day until the goal amount is reached in order to decrease the risk of diarrhoea

Protein Recommendations

Protein during the first 20 – 48 hours:

Infants:

- Initial protein per kg of 0.5 – 1g/kg
- Increase with 0.5g/kg increments according to ammonia levels and the infant's clinical condition until the goal amount of protein is met.
- Goal: 1.5g – 1.9g/kg

Children > 1 year of age:

- Increase with 0.5g/kg increments according to ammonia levels and the child's clinical condition until the goal amount of protein is met.
- Goal: 0.8 – 1.0g/kg

If baseline ammonia > 100 mmol/l or be lead by clinical signs

- Start with increments of 0.25 – 0.5g/kg protein.
- Increase to goal amount daily.

If ammonia < 100 mmol/l or be lead by clinical signs

- Start with increments of 0.5g/kg day
- Increase to goal amount daily.

If ammonia levels show a sudden upward trend or there is a sudden deterioration in patients clinical condition

- Decrease protein intake by 0.25 – 0.5g/kg/day
- Increase again slowly as tolerated and according to levels/

NB:

- Lactulose 5 – 20ml/day should be prescribed in all children with encephalopathy with the aim of producing 2 – 3 loose stools per day.
- In addition to this fleet enema may be used along with neomycin to purge the large bowel of faecalant matter.

Fat Recommendations

Birth - < 5 years of age

- 40% NPE

Children > 5 years of age

- 30 – 35% NPE

Adding fat to feeds:

- 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.
- Liquigen, Calogen, Duocal oe Energivit is recommended for supplementing fat.
- If the intake of long chain fats is restricted in order to prevent EFA deficiency: 2 ml per 100kcal/ day Walnut oil to prevent EFA.
- Increments of 1% per 24 hours or 0.5g/kg per 100ml per day up to goal amount.

NB: Fat is restricted only if an inborn error of metabolism (IBM) is suspected.

Vitamins Recommendations

- Coagulopathy will present when INR > 1.5
- Supplement Vitamin K: 2-10 mg/ day orally
- Do not supplement with other fat soluble vitamins
- Water soluble vitamins in the form of a multivitamin may be prescribed

Entry and Exit Criteria for Nutrition Support

Entry Criteria for nutrition support during hospitalisation: Daily Dietetic Review

- Poor oral intake
- Weight loss or growth failure during hospitalisation
- Nutrition risk score > 6

Entry Criteria for nutrition support on discharge home: Monthly Dietetic Review

Acute malnutrition: Weight/ Height

- < 80%

Acute Malnutrition: Weight for age

- < 76%

Chronic malnutrition: height for age

- < 89%

Moderate Malnutrition

- MUAC < 14.5 cm - >11.5cm in children < 5 years of age

Severe Malnutrition

- MUAC < 11.5cm in children < 5years of age.

Qualifies for entry into the Nutrition supplementation programme (NSP)

- Infants: 6 months – 1 year: when infants' growth curve flattens or drops over two consecutive visits.
- Children > 5 years ≤ 18 years: When child's growth curve flattens or drops over two consecutive months.

Referral NSP Scheme

- Access from local day hospital/ CHC

Private Medical Aid Patients

- Growth failure – flattening or down ward crossing of centiles.
- Downward crossing of 2 or more centiles over a period of 1 month or 2 consecutive visits.

Acute malnutrition: Weight/ Height

- < 80%

Acute Malnutrition: Weight for age

- < 76%

Chronic malnutrition: height for age

- < 89%

Moderate Malnutrition

- MUAC < 14.5 cm - >11.5cm in children < 5 years of age

Severe Malnutrition

- MUAC < 11.5cm in children < 5years of age.

Exit Criteria for nutrition support:

- **Monthly review for first 3 months, if growing well quarterly, followed by 6 monthly and annually.**

NSP

- 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.
- WH >90%
- HA >95%
- WA > 90%

Or Private Patients

- 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%
- WA > 90%

Summary: Establishing Nutrition Support in the Acute Liver patient

Goal: To ensure that each patient with chronic liver 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 to determine patient's nutritional status & risk:

- Height MAC TSF %EHA HC MUAC Weight %EWA
- %EWH AMA

Assess dietary intake: Interview caregiver

- Complete 24 hour diet recall
- Food frequency
- Is the intake appropriate according to the DRV's?

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

Yes
Yes
No

Monitor
3 month review

Good intake: Pre Discharge

- Educate caregiver about appropriate and affordable food intake.
- Inform caregiver of the food based dietary guidelines [FBDG]

Supportive Medical Management of Acute Liver failure with encephalopathy

- No sedation for procedures
- **Monitor:** neuro obs 4 – 6 hrly, gastric pH [>5], blood glucose [>4 mmol/kg]; acid/ base electrolytes, PT, PTT.
- **Fluid balance:** 75% maintenance – maintain circulating volume with colloid/ fresh frozen plasma.
- **Coagulation support:** (fresh frozen plasma)
- **Drugs:** Vitamin K (2 – 10mg/d IV or orally), sucralfate (2- 4g/day), lactulose (5 – 20ml/day), N-acetyl cysteine – IV (70mg/kg/6 hours), +/- broad spectrum antibiotics, neomycin.
- **Enteral feed:** enteral feed (aim for 1 – 2g/kg protein per day depending on age), parenteral nutrition may be occasionally required.

Poor intake: Pre Discharge

- Encourage caregiver.
- 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.

- Growth faltering
- WH $< 90\%$ expected
- HA $< 95\%$ expected
 1. Government: NSP Programme
 2. Private Medical aid motivation

Nutrition Support

Infants

- Breastmilk
- Infant cereal > 6 months age

Children

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

ACUTE PHASE

Provide sufficient glucose:

- Infants 8 – 10 mg/min/kg/gluc
- Children 7mg/min/kg/gluc
- Adolescents 4 mg/min/kg /gluc

Protein

- If baseline ammonia > 100 mmol/l.
- Start with 0.5g/kg increments during first 24 – 28 hours.
- Increase 0.5g/kg daily until goal amount is reached.
- If there is an increase in ammonia levels or $<$ in levels of consciousness $<$ protein by 0.5g/kg.
- All patients should receive lactulose therapy – 5 – 20ml per day.

Energy

- Aim to provide sufficient non protein energy in order to prevent endogenous catabolism of somatic and visceral tissues and maintain glucose homeostasis

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 > 15 cm in children < 5 years of age.
- WH $> 90\%$ expected
- HA $> 95\%$ expected

Recovery Phase

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

Infants

- 2 - 3g/kg protein
- 100 - 120kcal/kg

Children

- 1.5 - 2g/kg protein
- 1.2 – 1.5 x RDA OR
- Schofield equation or WHO x 1.7 – 1.9 [combined activity & stress factor]

3. Summary of recommendations for nutrition management of infants and children with chronic liver disease

3. Summary Nutrition Recommendations: Chronic Liver Failure	
Overview	
<ul style="list-style-type: none"> Biliary atresia is the most common cause of chronic liver failure with destruction of intra- and extrahepatic ducts leading to cholestasis, fibrosis and cirrhosis. Aims of medical management are to prevent progression of liver failure, prevent malnutrition and control or prevent complications. 	
Recommendations	Comments
Anthropometry	
<p>Measure At least monthly:</p> <ul style="list-style-type: none"> Head Circumference [< 3 years of age] SFTT MUAC MAC AMA Plot growth velocity biannually. Plot parental heights and determine mid height to be used as an index for target height. [See Anthro Guideline] Medically stage sexual maturation using Tanner system [when appropriate] [See Anthro Guideline] 	<p>Calculate At least monthly:</p> <ul style="list-style-type: none"> HA/HFA WA WH % EWA % EWH % EHA/HFA
<p>Classifications of malnutrition: Requiring Nutrition intervention:</p> <ul style="list-style-type: none"> Immediate nutrition support should be commenced in all patients classified as having moderate malnutrition. In those with mild malnutrition their status should be the following month, if there is no improvement with respect to weight or height/length, nutrition support should be commenced immediately. 	<p>All children who are classified as malnourished in hospital using height for age, weight for height or nutrition risk score.</p> <p>Height for age: Chronic malnutrition; stunting</p> <ul style="list-style-type: none"> Mild 90 – 95% Moderate 85 – 90% Severe <85% <p>Weight for Height: Acute Malnutrition; wasting</p> <ul style="list-style-type: none"> Normal 90 – 110% Mild 80 – 90% Moderate 70 – 80% Severe < 70% <p>Weight for age: acute malnutrition; wasting</p> <ul style="list-style-type: none"> Obese >120% Normal > 90% Mild malnutrition 76 – 90% Moderate malnutrition 61 – 75% Severe malnutrition < 60% <p>Nutrition Risk Screening Tool score:</p> <ul style="list-style-type: none"> 1 – 3 no current nutrition risk 4 – 5 some nutrition risk > 6 malnourished <p>All children should be reviewed monthly. If post hospital discharge malnutrition occurs e.g.</p> <ul style="list-style-type: none"> When child's growth curve flattens or drops over one consecutive month. Nutrition risk score of > 6. < 5th or 3rd centile < 80% EWH <-2 and - 3 ZD Downward crossing of 2 or more centiles

Biochemistry		
<p><i>Monitor the following</i></p> <ul style="list-style-type: none"> • Urea, creatinine, sodium, potassium • Magnesium, calcium and phosphate • Ammonia • Glucose • Albumin, ALT, AST, GGT • Bilirubin [conjugated, unconjugated] • INR & Fibrinogen • FBC: Hb, platelets, WCC • Triglycerides <p><i>Others</i></p> <ul style="list-style-type: none"> • Insulin Like Growth Factor –1 [IGF-1] • Pre albumin • C – reactive protein 		<ul style="list-style-type: none"> • If Ammonia > 100mmol/l restrict protein or reduced amount by 0.25g – 0.5g/kg. • Increase again as soon as possible according to ammonia levels. • If Mg, PO₄ or K low provide IV supplementation as per Refeeding syndrome guideline. • If INR > 1.5 and TPN required consider delaying commencement until INR < 1.5 • If triglycerides are high post transplant provide fat free parenteral nutrition – otherwise lipid-containing bag should be given as indicated.
Entry Criteria for nutrition support during hospitalisation based on biochemistry		
<ul style="list-style-type: none"> • Growth faltering, bilirubin > 70umol/l or overfeeding on normal formula 	<p>Formula</p> <ul style="list-style-type: none"> • Change to MCT containing milk < 1-year age and nutrition complete supplement > 1 year of age. <p>Breastmilk</p> <ul style="list-style-type: none"> • Alternate feeds between bottle and breast. • If poor weight gain continues maximise nutrient density with MCT containing feed. • Breastfeeding may be used as comfort feeding in those with persistent growth failure requiring more kcal. 	<p>Formula</p> <ul style="list-style-type: none"> • High MCT oil content 50 – 60% <p>Breastfeeding</p> <ul style="list-style-type: none"> • Top up feeds with MCT containing milk providing ½ requirements per day e.g. 75 – 90ml/kg.
<ul style="list-style-type: none"> • Cholestatis resolves, bilirubin < 30 umol/l 	<ul style="list-style-type: none"> • If bilirubin is <30umol/l and the child is thriving, MCT containing feeds may be changed to a normal feed. • If child is not thriving consider provide energy dense RTU feed in infants 1kcal/ml and 1.5kcal/ml in children. 	<ul style="list-style-type: none"> • Nasogastric feeds may be required in children especially if there is poor weight gain or linear growth is poor.
Clinical		
Clinical Signs usually present during liver disease in children:		
<ul style="list-style-type: none"> • Clubbing • Jaundice • Oedema • Large abdomen 	<ul style="list-style-type: none"> • Hepatomegaly • Splenomegaly • Ascites < Muscle bulk • < Skinfold thickness 	
<p>There are many causes of chronic liver failure most of which are likely to present early in life.</p> <p>Cirrhosis</p> <ul style="list-style-type: none"> • Cirrhosis represents ESLD occurring as a result of repetitive sequence of cell injury and repair. • Leads to cyclical necrosis and fibrogenesis leading to irreversible damage in addition to primary disease process. • Cirrhosis may be asymptomatic but can decompensate when damage to the liver causes blood flow to be impaired resulting in symptoms such as: • Portal hypertension • Ascites • Varices 		

Symptoms that require nutrition intervention include:

Jaundice

- Results in dark urine and pale stools.
- Occurs when there is an increase total bilirubin of which more than 20% is conjugated (normal = 5%).
- This may be significant hepatobiliary disease and cholestatic liver disease.
- May result in reduced bile flow from the liver into the gut.
- Decreased fat emulsification and digestion; malabsorption of fat, fat-soluble vitamins and some minerals.
- Steathorrea, growth failure and rickets are common.

Fat malabsorption

- Fat should not be restricted but given to tolerance.
- Infants require significant amounts of fat in their diet 40%.
- Of this it is important to provide a percentage of the total fat as MCT's e.g. 40 - 50%.

Hypoglycaemia

- Infants may require overnight continuous feeds with up to 6g/ 100ml CHO.
- In children a diet high in complex CHO should be given.

Failure to thrive

- A diet high in energy and protein.

Ascites & hepatomegaly

- Reduced abdominal capacity for food/ feeds - give smaller, frequent, energy dense meals/ snacks.
- In volume restriction may require lower sodium feed e.g. 1.2 – 1.5mmol/l.
- However, the use of diuretics is favoured.

Portal hypertension & malabsorption

- Cirrhosis can obstruct blood flow leading to portal hypertension with assoc. enteropathy and malabsorption secondary to pressure in the mesenteric venous system.
- Semi elemental and continuous feeds may help – if not TPN may be required.

Oesophageal varices

- Occasionally in ESLD a huge bleed will require TPN, otherwise 24 - 48 hours post sclero therapy clear fluids may be allowed followed by soft diet.
- Chronic encephalopathy – protein restrictions can < nutr status.
- Degree of encephalopathy should be determine level of restriction.
- Energy ratio should be > to < endogenous breakdown (sodium benzoate may allow for higher protein tolerance & or use of BCAA).
- May require restriction of protein to 1 – 2 g/kg infants and children 0.8 – 1.5g/kg.

Dietary

Potential problems requiring nutrition intervention occur when there is a disturbance of normal metabolic functions of the liver including:

- Glucose homeostasis
- Protein synthesis
- Bile Salt production
- Lipid metabolism
- Vitamin absorption and storage

Diet therapy includes management of:

- Hypoglycaemia for poor glycogen storage
- Fat malabsorption as a result of poor bile production or flow
- Reduced in protein synthesis especially albumin exacerbating ascites.
- Vitamin deficiencies due to malabsorption of fat soluble vitamins

At each follow up a thorough nutrition history should be captured.

Components of a nutrition history include

Diet history

- Review through 24-hour dietary recall quarterly or at each follow up, use in conjunction with food frequency.
- Many patients may eat < 65% of RDI.

- Weight Change
- Appetite
- Satiety Level
- Taste Changes/ aversions
- Nausea/ vomiting
- Bowel habits – constipation, diarrhoea
- Chewing/ swallowing ability
- Pain when eating

- Long-term disease(s) affecting absorption/use of nutrients
- Surgical resection or disease of GI Tract
- 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.

Fluids		
<ul style="list-style-type: none"> It is important to note that on presentation some children will be consuming up to 2 – 3 x normal fluid requirements e.g. 200 – 300ml/kg and may demand frequent breastfeeds, this may be a compensation mechanism for fat malabsorption. Once feeds containing a higher amount of MCT's are introduced this phenomenon should resolve. Avoid excessive sodium (<2mmol/kg/day) Ascites treat with: spironolactone, furosemide 		
Fluid Range		
Age (years)	ml/kg actual body weight	
Premature	180-200	
0-1	150	
1-3	100	
3-6	90	
7-10	70	
10-15	60	
Energy Requirements		
<ul style="list-style-type: none"> Provide sufficient energy for linear growth and development. Infants use breastmilk or infant formulas ranging from 0.67 - 0.74 – 1kcal/ml Infants Children > 1 year: Use age-appropriate feeds with standard or energy dense calorie concentrations, ranging from 1 – 1.5kcal/ml respectively <p><i>Infants:</i></p> <ul style="list-style-type: none"> 120 – 150kcal/kg <p><i>Children:</i></p> <ul style="list-style-type: none"> Schofield equation x 1.7 – 1.9 or WHO/FAO/UNU [Appendix 1:Table 2, 3 and 4] 		
Or		
<ul style="list-style-type: none"> 1.2 – 1.5 x DRV [Appendix 1: Table 1] 		
Dietary Recommendations		
Infants	Discussion	Recommendations
<p>Stage 1: Growth Faltering, bilirubin > 70mmol/l or over feeding on normal formula.</p>	<p>Formula</p> <ul style="list-style-type: none"> Change to MCT containing feed. 	<p>Formula</p> <p>High MCT 50 – 60% 150 – 200ml/kg/day</p>
<p>Stage 2: If poor growth persists</p>	<ul style="list-style-type: none"> For infants with persistent cholestasis energy supplementation with glucose polymer and MCT oil is required to achieve weight gain. Add glucose polymer in 1 – 2% daily increments as tolerated, in conjunction with MCT oil 1- 2% to achieve an energy density of 1kcal/ml. Do not exceed 50% of total energy from fat. 	<ul style="list-style-type: none"> Assess total daily feed intake to monitor protein intake. Consider use of RTU energy and protein dense feed.
<p>Stage 3: Cholestasis resolves < 30 mmol/l</p>	<ul style="list-style-type: none"> If bilirubin <30 mmol/l and the child is thriving, MCT feeds may be changed to normal feed. If infant is not thriving provide energy dense RTU feed [1kcal/ml] 	<ul style="list-style-type: none"> Standard formula or breastfeeds. Energy dense RTU feed 1kcal/ml
<p>Stage 4: If cholestasis does not resolve</p>	<ul style="list-style-type: none"> Continue with MCT formula, energy supplements and diet. Frequent monitoring, review of growth, feed intake and tolerance. 	<ul style="list-style-type: none"> Nasogastric feeds may be required especially if weight gain or linear growth is poor.
Older Children		
<ul style="list-style-type: none"> Older children usually tolerate a normal fat diet well and should be encouraged to have fat to tolerance. Some children will self regulate to avoid fatty foods. Children who are failing to thrive need to be on high calories and protein, which may require the use of supplements. Use of 1.5kcal/ml feeds should be encouraged. 		

Enteral Feeding
<ul style="list-style-type: none"> • Bilirubin < 30mmol/l • If patient cholestatic or has ascites. • Volume of feed given depends on growth failure – add daytime boluses in addition to overnight feeds if growth failure persists. • Patients with poor growth on high calorie MCT feeds should be followed up weekly via telephone, with monthly reviews. <p>Infants:</p> <ul style="list-style-type: none"> • 1kcal/ml <p>Children:</p> <ul style="list-style-type: none"> • 1 - 1.5kcal/ml RTU feed with fibre. <p>Overall:</p> <ul style="list-style-type: none"> • Lower sodium feed. • 50 –75% of energy requirements with the remainder made up from normal oral food intake.
Nutrition Support
<p>To increase nutrition content of food.</p> <ul style="list-style-type: none"> • Overnight NG feeds – MCT containing formula in those patients awaiting transplant. • PEG's relatively contraindicated
Growth failure
<p>CHO:</p> <ul style="list-style-type: none"> • 26 – 28% may be required in ESLD to maintain glucose homeostasis. <p>Fat:</p> <ul style="list-style-type: none"> • Not all ESLD will be cholestatic and LCT may be used. • For cholestatic disease proportions of 50:50 or 70:30 ratio is usually required. • Where there is low intake of LCT < 2 – 3% of total energy walnut oil will be required 2% energy in cholestasis and 1% energy in non cholestatic. • Sodium chloride: 1.5mmol/kg should be used where majority of nutrition is coming from feed and 1mmol/100ml when the feed is used to supplement food. • Potassium chloride: If the patient is on frusemide may have increased requirements for potassium. <p>Aim to provide</p> <ul style="list-style-type: none"> • Protein: 3 – 4 g/kg protein [unless protein restricted] • CHO: up to 18% • Fat: 0.5% fat increments (1% emulsion) up to 5 – 8 % fat per 100ml. • Sodium Chloride: 1mmol/100ml feed or 1.5mmol/kg unless otherwise indicated. • Potassium Chloride: 2mmols/100ml feed or 3mmols/kg unless otherwise indicated.
Energy Supplementation
<p>Infants:</p> <ul style="list-style-type: none"> ▪ Breastfeeding should be supported where possible. ▪ If supplementation is required a carbohydrate and fat [MCT/LCT] containing powder should be added to a small amount of expressed breastmilk [5 – 10ml] and given to the infant prior to a feed as prescribed by a dietitian. ▪ If additional calories are required in formula feeding a carbohydrate and fat [MCT/LCT] containing powder should be dissolved in a small amount of sterile water. <p>Ready to use/ hang feeds:</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 then boluses of super soluble carbohydrate and fat powder should be administered prior to a drink or feed. ▪ Recommendations regarding concentrations for fat, carbohydrate and protein per age category should not be exceeded. [See sections below]

Carbohydrate Recommendations

Glucose requirements:

Infants

- 8-9mg/kg/min [11.5g –12.9g/kg/day]
- Max 12.5mg/min/kg [18g/kg/day]

Toddlers

- 7mg/kg/min [10g/kg/day]

Adolescents

- 4mg/kg/min [5.7g/kg/day]

NB: Always increase glucose gradually and according to tolerance.

CHO Concentrations

The following concentrations of CHO per 100ml will be tolerated if a glucose polymer is used.

- Infants under 6 months: 10-12% carbohydrate concentrations (i.e. 7g from formula, 3-5g added)
- Infants 6months to 1 year: 12-15%
- Toddlers 1-2 years: 15-20%
- Older children > 2 years: 20-30%

Format of CHO

- CHO's in the form of glucose polymer should be used instead of disaccharides as they have a lower osmotic effect.
- It is recommended that glucose polymer be added in increments of 1g per kg per 100ml per day until the goal amount is reached in order to decrease the risk of diarrhoea.

Protein Recommendations

- Protein may appear in feeds in the form of amino acids, peptides or whole proteins.
- Protein supplementation is rarely required and addition of a protein powder and is not recommended in infants or children > 1 year of age due to the dangers of providing too much protein and sodium.
- Feeds with added protein should usually be accompanied with an increase in energy e.g. energy-enriched products, otherwise protein will be used for gluconeogenesis.

Infants:

- 3 – 4g/kg/day

Children:

- up to 2kg/day

Protein-energy ratio of a feed should be kept within a range of:

- 7.5% - 12% for infants [i.e. 7.5% - 12% of total energy requirements from protein]
- However, for most infants an intake of between 9 – 10% is a practical ratio to aim for.
- 5 – 15% in older children.
- At least 9% of energy from protein for “catch up growth”

Fat Recommendations

NB: Fat should not be restricted and a portion of fat should be in the form of MCT

Ensure DHA and AA supplementation

- Long chain fat emulsions in the form of olive/ canola oil are often favoured over MCT's as they have a lower osmotic effect and provide EFA's.
- Where additional EFA are required provide 2ml per 100kcal walnut oil.
- If fat malabsorption, aim for the upper limit of DHA:AA

EFA supplements:

Infants:

- DHA and AA supplement in the form of a LCPUFA enriched infant formula with the following ratio's:
 - Linoleic acid 10.2 – 12.5%
 - Alpha linolenic acid 1.2 – 1.5%
 - Arachidonic acid 0.3 – 0.4%
 - Docosahexanoic acid 0.04%

Children > 1 year of age:

- 4.5 – 10.8% Linoleic acid by means of a nutritionally complete feed or supplement drink.
- Linoleic: alpha linolenic ratio - 5:15
- Children have a higher tolerance of LCT than infants

Additional Fat Sources:

- In those children with fat malabsorption or awaiting transplant provide nutritionally complete drinks, which have MCT's as part of the lipid profile.
- MCT oil can also be supplemented – 0.3ml/ kg:
 - Give as a medicine; as a bolus prior to feeds or add to food once the food has been cooked (Do not use MCT oil to cook or fry with as it has a low temperature threshold).
 - Excessive amounts of MCT's may also be neuro toxic.
 - Avoid providing more than 40% of total fat as MCT.
 - It will not be tolerated at higher concentrations and may cause abdominal cramps and osmotic diarrhoea.
- Fat emulsions or those in the form of a super soluble powder with carbohydrates are preferred for increasing calories and should be added in small increments until goal amount is reached
- Adding fat to feeds should be done as a last resort. Try to rather add extra oil/ margarine to food.
- Excess fat supplementation can lead to nausea, vomiting and delayed gastric emptying. The following goal amounts should not be exceeded:
 - Infants: 1% or 0.5g fat per 100ml per day up to 5 –6% per day (5 – 6g fat per 100ml feed).
 - Children > 1 year: 7% total fat (7g fat per 100ml feed).

Fat Soluble Vitamins

Only oral preparations of fat-soluble vitamins are currently available in RSA. Use an aqueous preparation where possible.

- Supplement vitamins A, D, E and K in all children with cholestatic disease.
- Aqueous forms available as ADEK®

ADEK® Dosage:

- **Liquid**
 - 0 – 12 months: 1 ml per day
 - 1 – 3 years: 2 ml per day
- **Tablet**
 - 4 – 10 years: 1 tablet per day
 - > 10 years: 2 tablets per day

Vitamin A (aqueous)

- Plasma retinol/ RBP: 5, 000 – 10, 000 – 50, 000 IU

25-OH vitamin D or Alphacalcidol

- Plasma 25 – OHD: 2 – 4 ug/kg/day **or** 50 ng/kg/day

Vitamin E

- Plasma E/ total lipids: 25 IU kg/d **or** 50mg – 200mg/day
- As d-alpha tocopheryl polyethylene glycol-1000 succinate:
 - 2 – 10mg/d o

Vitamin K

- Check prothrombin time and for coagulopathy.

Introduction of Complimentary Foods

- Normal complimentary foods may be commenced at 4 – 6 months age.
- Dried baby cereal may be made with formula or breastmilk rather than water.
- No foods need be avoided.
- For infants with poor growth high energy weaning advice should be given.
- Parents should be encouraged to sustain oral intake no matter how little to develop oral feeding skills.

Addition of:

- Sugar/ glucose polymer
- Margarine, olive or canola oil, butter or MCT oil to foods.

End stage liver disease Recommendations		
<ul style="list-style-type: none"> • Patients may be fluid restricted to 60 – 80% of maintenance fluid • Reducing fluid impacts on calorie, protein, vitamin and mineral intake. 		
Ascites	<ul style="list-style-type: none"> • Fluid and salt restriction • Energy dense, low sodium feeds • Ice lollies useful way of making fluid last longer. 	<ul style="list-style-type: none"> • No added salt diet, 1 – 1.5mmol/kg Sodium. • Negotiate with team for more volume!
Steatorrhea	<ul style="list-style-type: none"> • MCT: LCT feed – additional MCT may be required. • Low fat diet is rarely required. 	<ul style="list-style-type: none"> • Skimmed milk with CHO: MCT powder may be useful.
Encephalopathy	<ul style="list-style-type: none"> • Unless acutely encephalopathic – protein restriction rarely required. • Enteral feeds will probably be required. 	<ul style="list-style-type: none"> • Temporary restriction of 0.5 – 1g/kg.
Hypoglycaemia NB: If child is NPO provide 10 – 20% dextrose, as 5% will not maintain blood glucose.	<ul style="list-style-type: none"> • Glucose homeostasis may be difficult. • Measure pre feed bloods. • Offer high CHO drinks and snacks. • Continuous feeds overnight. 	<ul style="list-style-type: none"> • Feed 3 – 4 hourly or more frequently 2 hourly. • Diet should be offered for as long as possible.
Liver Transplant Recommendations		
Anthro	<ul style="list-style-type: none"> • Nutrition review pre surgery • Commence NG feeds if required pre surgery. 	
Waiting period	<ul style="list-style-type: none"> • Reviewed monthly in OPD. • Infants may require more frequent review especially if on NG feeds. 	
Post Transplant	<ul style="list-style-type: none"> • Oral fluids usually started 72 hours post surgery via NGT. • If after 72 hours feeds cannot be commenced consider PN until 50% of total energy requirements can be met enterally. • Feeds usually semi-elemental, started whilst child is fluid restricted e.g. 80% maintenance. • Feeds increased as fluids are liberalised. • Re start age appropriate diet as soon as possible – favourite foods may be used to tempt children. • Some children have may have developed food aversions – speech therapist or psychologist intervention may be warranted. • Majority of post transplant patients experience some loose stools once oral diet is commenced but this settles within in a few days. • Ongoing diarrhoea/ vomiting merits investigation. • Patients are usually on high doses of steroids for 3 months post surgery, which helps to improve their appetite. • As steroids are stopped appetite may deteriorate and energy supplementation may be required. • Anthropometry is essential during this time as weight gain may be rapid and excessive. • Children in whom significant catch up growth is required may benefit from over night feeding until this has been achieved. 	<ul style="list-style-type: none"> • Clear fluids ½ dd introduced, as bowel sounds present or flatus/ stool passed. • Previously NG fed – start semi elemental feed e.g. < 1 year 0.67kcal/ml RTU, 1kcal/ml powder/RTU > 1 year.
Follow up		
<ul style="list-style-type: none"> • Patients with poor growth on high calorie MCT feeds should be followed up weekly via telephone. • Monthly reviews 		

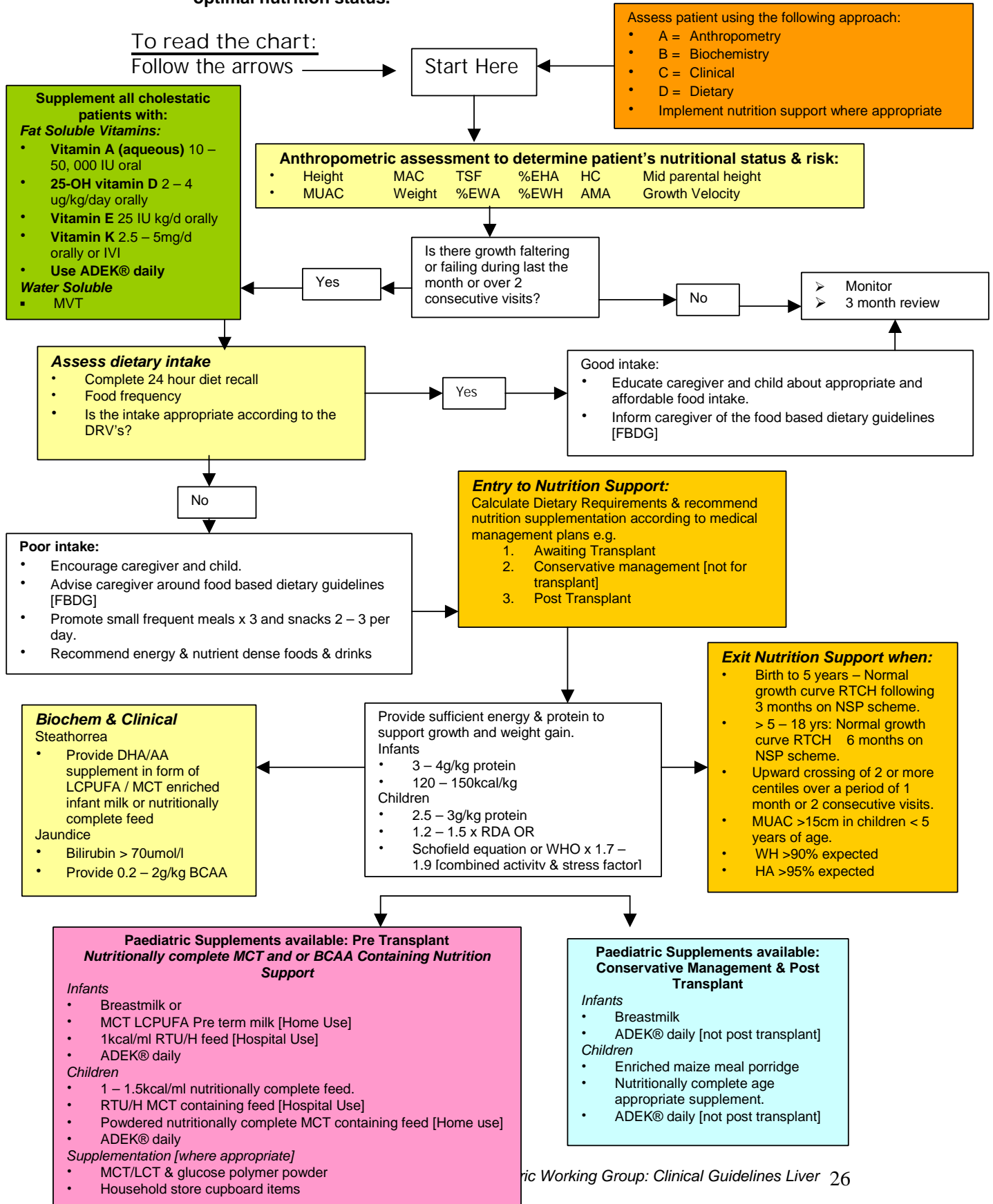
Product Choices for patients with Chronic Liver Disease
<p>Overall Recommendations</p> <ul style="list-style-type: none"> • In infants a max of 1.2kcal/ml is recommended. • In children a max of 1.5kcal/ml is recommended. <p>NB</p> <ul style="list-style-type: none"> • Feeds enriched with modulars > of 1.5kcal/ml in children and 1.2kcal/ml in infants may cause early satiety, nausea and vomiting. • Recommend utilising max 1kca/ml feeds in infants and 1.5kcal/ml children and liberalising volume of feed provided. • Encourage energy dense food providing advice to caregiver around enriching store cupboard options with an emphasis on favourite foods. • To enrich milk add 4 tablespoons (60g) full cream milk powder to 500ml ml milk/maas [605kcal & 30.7g protein > 1 year old.
Infants
<p>Home use</p> <p>Feed of Choice: Breastmilk</p> <ul style="list-style-type: none"> • If no weight gain top up with energy dense pre term formula MCT & LCPUFA containing 0.74kcal/ml or alternate feeds e.g. breast then formula. • If still no weight gain enrich with duocal/ EBM flushes before each feed. • Provide 75 – 90ml/kg from formula feeds. • ADEK® daily <p>Breast milk substitute: Pre Term infant feed</p> <ul style="list-style-type: none"> • MCT & LCPUFA containing 0.74kcal/ml • If no weight gain enrich with carbohydrate and MCT/LCT powder flushes • Ensure nutritional requirements are first being met from the feed e.g. 9 - 12% protein, 40% fat, 48 -51% CHO, before providing extra energy and that • All growth factors are within normal range e.g. sodium, potassium, haemoglobin etc. • ADEK® daily <p>Hospital Use</p> <p>Ready to Hang/ Use 1kcal/ml infant formula</p> <ul style="list-style-type: none"> • 1kcal/ml & 2.6g protein per 100ml • Do not add energy supplements to sterile milk. • If additional energy is required provide as bolus flushes prior to feeds. • ADEK® daily
Children
<p>Home Use</p> <ul style="list-style-type: none"> • Nutritionally complete 1kcal/ml MCT containing powdered feed. • Try with 1kcal/ml first • Enrich up to max 1.5kcal/ml with carbohydrate and MCT/LCT powder. • ADEK® daily <p>Hospital Use</p> <ul style="list-style-type: none"> • 1.5kcal/ml Ready to use/ hang paediatric feed. • Do not add additional energy or protein supplement to feed. • ADEK® daily
Additional Specialised Products
<p>Infants:</p> <ul style="list-style-type: none"> • Caprilon [SHS] <p>Children:</p> <ul style="list-style-type: none"> • Generaid [SHS] BCCA Enriched • Poor palatability <p>BCAA</p> <ul style="list-style-type: none"> • A range of 0.2 – 2g/kg has been provided, however, efficacy appears to occur at 1g/kg. • Universal Nutrition BCAA powder 10g Powder contains:2.75g Glutamine, 2.5g Leucine, 1.25g Isoleucine, 1.25g valine. • Efficacy of supplementation not fully elucidated but some studies have shown some beneficial effect on Fisher ratio.

Entry Criteria for Nutrition Support in Hospital: Daily Dietetic Review
<ul style="list-style-type: none"> • Poor oral intake • Weight loss or growth failure during hospitalisation • Over feeding on normal formula • Growth failure classified as a downward crossing 2 or more centiles over a period of 1 month or 2 consecutive visits. • Nutrition risk score > 6 <p><i>Moderate Malnutrition</i></p> <ul style="list-style-type: none"> • MUAC < 14.5 cm - >11.5cm in children < 5 years of age <p><i>Severe Malnutrition</i></p> <ul style="list-style-type: none"> • MUAC < 11.5cm in children < 5years of age. <p>Acute Malnutrition: Weight for age</p> <ul style="list-style-type: none"> • <80% <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%
Entry Criteria for Nutrition Support on discharge from Hospital: Monthly Dietetic Review
<p><i>Acute malnutrition: Weight/ Height</i></p> <ul style="list-style-type: none"> • < 80% <p>Acute Malnutrition: Weight for age</p> <ul style="list-style-type: none"> • < 76% <p><i>Chronic malnutrition: height for age</i></p> <ul style="list-style-type: none"> • < 89% <p><i>Moderate Malnutrition</i></p> <ul style="list-style-type: none"> • MUAC < 14.5 cm - >11.5cm in children < 5 years of age <p><i>Severe Malnutrition</i></p> <ul style="list-style-type: none"> • MUAC < 11.5cm in children < 5years of age
Additional Entry Criteria for Nutrition supplementation programme (NSP): Monthly Dietetic Review
<ul style="list-style-type: none"> • Infants: 6 months – 1 year: when infants' growth curve flattens or drops over two consecutive visits. • Children > 5 years ≤ 18 years: When child's growth curve flattens or drops over two consecutive months. • If nutrition supplementation must be continued for only 6 months patient can be re-entered with additional motivation from a dietitian. • It is possible to motivate for patients with chronic liver disease to remain on the NSP programme. • Home visits including assessment of food security and access should be completed with a referral to Social Welfare for available grants, food assistance where appropriate.
Entry Criteria for Nutrition Support in Private Medical Aid Patients: Monthly Dietetic Review
<ul style="list-style-type: none"> • Growth failure – flattening or down ward crossing of centiles. • Downward crossing of 2 or more centiles over a period of 1 month or 2 consecutive visits. <p><i>Acute malnutrition: Weight/ Height</i></p> <ul style="list-style-type: none"> • < 80% <p>Acute Malnutrition: Weight for age</p> <ul style="list-style-type: none"> • < 76% <p><i>Chronic malnutrition: height for age</i></p> <ul style="list-style-type: none"> • < 89% <p><i>Moderate Malnutrition</i></p> <ul style="list-style-type: none"> • MUAC < 14.5 cm - >11.5cm in children < 5 years of age <p><i>Severe Malnutrition</i></p> <ul style="list-style-type: none"> • MUAC < 11.5cm in children < 5years of age.

Recommended Product Choice for Nutrition Support
<p>Conservative Management: Not for transplant</p> <ul style="list-style-type: none"> • Aim maintain/ improve quality of life, providing sufficient energy for growth. • Enrich foods with household food sources. • Refer to NSP scheme for nutrition support, where appropriate. • Refer to medical aid for nutrition support, where appropriate. <p><i>Use products such as:</i></p> <ul style="list-style-type: none"> • Enriched maize meal porridges and or age appropriate nutritionally complete supplements. • Supplement fat Soluble vitamins • ADEK® daily
<p>Awaiting Transplant</p> <ul style="list-style-type: none"> • Aim maintain/ improve nutrition status, providing sufficient energy for growth. • Enrich foods with household food sources • Refer to NSP scheme for nutrition support, where appropriate. • Refer to medical aid for nutrition support, where appropriate. <p><i>Use products such as:</i></p> <ul style="list-style-type: none"> • Glucose or MCT/LCT/ glucose polymer duo product • MCT & LCPUFA containing nutrition complete supplement/ infant feed. • Supplement fat-soluble vitamins. • ADEK® daily <p><i>Additional Product Considerations:</i></p> <ul style="list-style-type: none"> • Branched chain amino acids 1g/kg in those patients with bilirubin >400mmol/l. • Source either age appropriate branched chain specialised amino acid feed e.g. Generaid or vitamin supplement powder [Health Food Shops].
<p>Post Transplant</p> <ul style="list-style-type: none"> • Improve nutrition status providing sufficient energy for catch up growth • Enrich foods with household food sources. • Refer to NSP scheme for nutrition support, where appropriate. • Refer to medical aid for nutrition support, where appropriate. <p><i>Use products such as:</i></p> <ul style="list-style-type: none"> • Use products such as Enriched maize meal porridges and nutritionally complete age appropriate supplements.
NSP Scheme Product Motivation for Chronic Liver failure
<ul style="list-style-type: none"> • Motivation required for MCT containing infant feeds [Pre term] and or nutritionally MCT containing complete drinks for those patients > 1 year of age awaiting transplant. • Access Pre Term and nutritionally complete paediatric feed from local day hospital/ CHC. • Additional motivation will be required by the referring dietitian.
Medical Aid Patients Product Motivation
<ul style="list-style-type: none"> • A motivation by a dietitian may be required for MCT containing infant feeds [Pre term] and or nutritionally MCT containing complete drinks for those patients > 1 year of age awaiting transplant. • If on Medical Aid scheme either access directly from company or from local pharmacy.
Exit Criteria for nutrition support: Monthly Dietetic Review for first 3 months, if growing well consider quarterly review.
<p>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. • WH >90% • HA >95% • WA > 90% <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% • WA > 90%

2.4 Summary: Establishing Nutrition Support in the Chronic Liver patient

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



3. Nutrition in Paediatric Liver Patient

3.1 Aim

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

Nutrition management usually aims to maintain glucose homeostasis, manage fat malabsorption including steatorrhoea, promote appropriate aminogenesis and prevent.¹

3.2 Objectives

The objectives of these guidelines are to:

- Identify appropriate feeding practices in the paediatric liver patient.
- Identify appropriate routes of feeding in the paediatric liver 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 e.g. enteral or parenteral feeding, where oral feeding has failed in the paediatric liver patient.

3.3 Statement Regarding Promotion, Protection and Support of Exclusive Breastfeeding in infants with liver 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.^{30, 31}

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.³²

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 liquid infant feed be used in all Neonatal and Paediatric Intensive Care /High Care Units, as well as 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 breastmilk 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”.³¹

4. Introduction

Liver disease in children is uncommon and can be split into two groups e.g. those with acute liver failure and those with chronic degenerative liver disease.

Acute liver failure may be as a result of a viral infection, ingestion of a toxin, poison and or as a result of an immune mediated attack. ^{1, 25, 26}

Causes of chronic liver failure are usually precipitated by inborn errors of metabolism such as glycogen storage disease or galactocaemia, biliary tract disease, infections and autoimmune disorders. ^{1, 26}

Inborn errors of metabolism will not be considered in this guideline. However, for completeness disease conditions such as tyrosinaemia, glycogen storage disease, fatty acid oxidation disorders, urea cycle defects, galactocaemia and fructosinaemia require specific dietary treatment often including restriction or exclusion of some dietary components. ¹

The majority of chronic liver disease cases in infancy arise from biliary atresia, whereas in the older child it may be as a result of a “silently” progressing chronic condition with acute decompensation as a result of an inborn error of metabolism or autoimmune liver dysfunction. ¹

Nutrition management is dependant on the aetiology of the disorder e.g. acute or chronic or metabolic. Liver dysfunction can result in profound metabolic and biochemical abnormalities such as disruption of glucose homeostasis, protein synthesis, bile salt production, lipid metabolism and storage of vitamins. ^{1,29}

There are a number of goals with respect to nutrition management:

- Promote normal growth and development.
- Maintain good nutrition status in order to prolong need for transplant.
- Provide most appropriate nutrition treatment to minimize symptoms and prevent nutrition deficiencies. ²⁹

A multidisciplinary liver team including a dietitian, physiotherapist, social worker, hepatologist, psychologist, should complete a global assessment of the medical, nutritional and quality of life status of individuals with liver disease. ²⁹

5. Anthropometry

5.1 Nutritional Assessment and History

Regular nutrition assessment is vital in attaining good nutrition status as it allows for early indication of malnutrition. However, the severity of malnutrition is not always correlated with individual anthropometrical, biochemical and clinical markers such as weight, liver function tests, vitamin and mineral status. ^{1,29} In a general hospital population, the use of a single marker such as weight would result in up to 80% of the population to be considered at risk of malnutrition, however a combination of markers would result in only 28%, a more realistic figure. ²⁹

Table 1 outlines nutrition assessments, which are considered useful or misleading in liver disease. ^{1 10, 14, 21}

Table 1: Evaluation of nutrition assessment parameters in childhood liver disease ^{1,10, 14, 21}

	Valuable	Misleading
Body weight		X
Height	X	
Upper extremity anthropometrics e.g. arms	X	
Lower extremity anthropometrics e.g. legs		X
Plasma protein		X
Nitrogen balance studies		X
Creatinine height index		X
Immune status: Lymphocyte count		X
Subjective assessment	X	
24 hour dietary recall	X	

5.1.1 Weight

Weight-for-age may be useful in the early stages of disease but becomes misleading and useless when hepatosplenomegaly, oedema and ascites are present. Weight-for-height provides an index of severity with respect to long-term malnutrition.¹ In addition to this, it may provide an indication of worsening liver disease related to ascites or an increase in water retention. Weight-for-height should be used to determine wasting.²⁹

5.1.2 Height

Growth velocity should be plotted at least 6 monthly using appropriate charts. An average of both parents' heights may be used to predict future stature. Tanner staging should be completed as the child approaches puberty as growth spurts would normally be expected. Malnutrition is not usually the cause of delayed puberty and other reasons should be investigated.²⁹

5.1.3 Body Mass Index

Body mass index (BMI) has limited value in chronic liver disease due to the confounding weight factor. However, it may be used to determine liver disease-related obesity, particularly post transplant.²⁹

5.1.4 Head Circumference

Head circumference should be plotted in all children up until the age of 3 years. Head circumference may be influenced by chronic malnutrition.²⁹

5.1.5 Skinfold Thickness

In chronic liver disease the upper body is less affected by oedema. Using four skin folds measurement of tricep, suprailiac, bicep and subscapular is not appropriate in this population group due to the confounding factors of fluid shifts.²⁹

Measurement of mid upper arm circumference (MUAC) and tricep skinfold thickness (TSF), can be used to determine arm muscle area (AMA). AMA is a sensitive indicator of changes in caloric intake and muscle mass. These two measurements are able to determine total body fat [*Figure 1*].^{1, 10,21, 24,29}

They are essential parameters in monitoring chronic liver disease and should be completed at least quarterly. Skinfolts cannot be taken in infants younger than 3 months of age.²⁹

Figure 1: Equations for arm muscle area using tricep skinfold thickness and mid upper arm circumference:

$$\text{AMA (cm}^2\text{)} = (\text{MAC (cm)} - [\text{TFSF} \times 4])^2 / (4 \times 3.14)$$

$$\text{AFA (cm}^2\text{)} = \text{upper arm area} - \text{AMA, where upper arm area} = \text{MAC}^2 \text{ (cm)} / (4 \times 3.14)$$

5.2 Other anthropometric measurements

Bioelectrical impedance (BIA) estimates body fluid volumes. Its use has been validated in adult and paediatric populations. The measurements are easy to obtain, however in chronic liver disease it is affected by ascites and peripheral oedema. Measurements obtained should be interpreted with caution.²⁹

Dual energy x-ray absorbitometry is used to determine bone mass, fat free mass and fat body mass. It is used in some centres as the gold standard for body composition and should be considered for use in children with chronic liver disease where available.²⁹

5.3 Mechanisms for Growth failure and Malnutrition

5.3.1 Acute Liver Disease

Acute liver failure as a result of viral hepatitis, may lead to vomiting, diarrhoea and acute weight loss, but malnutrition is unusual.^{1, 10} If the presentation is acute then the children are often well nourished, in these cases the aim of nutrition management is to maintain nutrition status until there is some improvement in symptoms.^{1, 26}

Life threatening complications of cerebral oedema may occur, however if the child survives, liver function should return too normal.^{1, 26}

5.3.2. Chronic Liver Disease

Malnutrition and or rapidly declining nutrition status increase the risk of morbidity and mortality especially in those children awaiting liver transplant.²³ More than 50% of patients presenting with obstructive liver disease have some related nutrition disorder in addition to being malnourished. The severity of malnutrition also appears to be associated with the duration and severity of jaundice experienced.¹

Obstructive jaundice leads to hypodipsia and hypophagia and over 60% of patients have an intake lower than their energy requirements despite recommendations to increase energy consumption.^{13,21,27,28}

Anorexia may be partly explained by an induction of an inflammatory response with a concomitant release of cytokines. Malnutrition is more frequent in patients with marked liver dysfunction and low anabolic capacity. Alkaline phosphatase (ALT) may be a useful prognostic factor for malnutrition in obstructive jaundice. In biliary atresia following biliary decompression e.g. post transplant, appetite is significantly improved.²¹

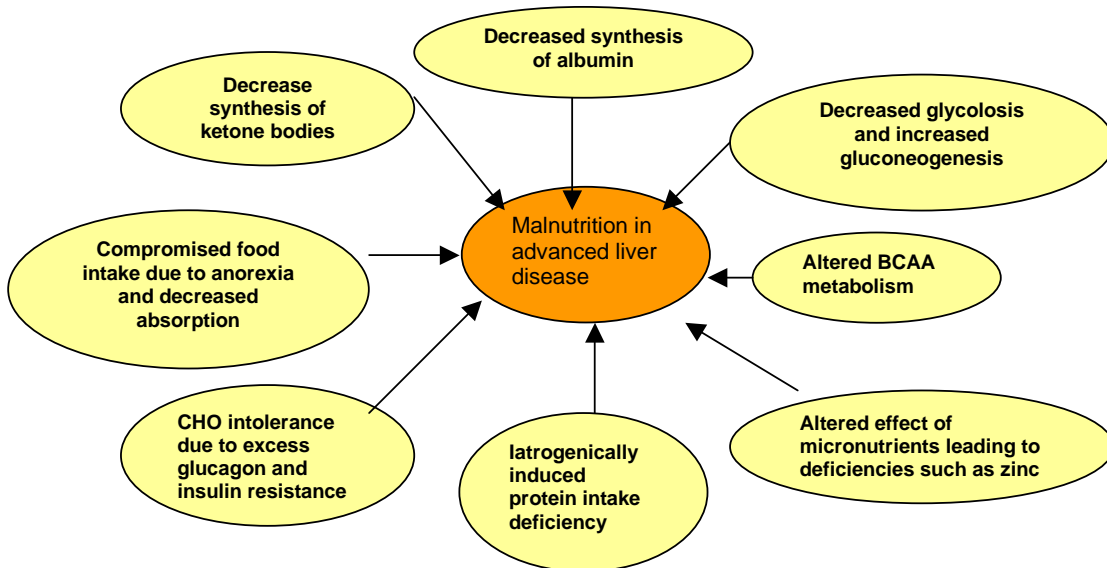
Preoperative nutritional status of children awaiting orthotopic liver transplant is often poor with many of them being chronically malnourished. Nutrition status significantly impacts on pre and postoperative survival in children following liver transplant with those children who were normally nourished experiencing longer survival rates than those who were malnourished.^{1, 10}

In chronic liver disease of infancy and childhood there are many causes for malnutrition as outlined in table 2 and *figure 2*.^{1, 10}

Table 2: Potential Causes of malnutrition in chronic liver disease

Potential Causes of Malnutrition in Childhood Liver Disease ^{1, 10}	
1. Well characterised in children	
A. Decreased intake	<ul style="list-style-type: none"> • Anorexia, nausea, vomiting • Mechanical difficulties caused by tense ascites • Hospitalisation related depression • Unpalatable diet
B. Impaired nutrient digestion and absorption	<ul style="list-style-type: none"> • Bile salt deficiency • Pancreatic insufficiency
C. Increased energy requirements	<ul style="list-style-type: none"> • Hypermetabolism • Stresses such as infection • Increase protein oxidation
2. Predicted from studies in adults	
A. Accelerated protein breakdown	
B. Inefficient protein synthesis	
C. Enteropathy from portal hypertension	
D. Malnutrition related to villous blunting	

Figure 2: Factors leading to malnutrition in advanced liver disease.



The presence of portal hypertension with cirrhosis may lead to enteropathy and malabsorption secondary to increased pressure on the mesenteric venous system. Some conditions such as Alagille Syndrome, Byler's disease and choledochal cysts may be accompanied by pancreatic insufficiency aggravating malabsorption. In addition to this, cholestasis results in malabsorption of dietary lipids and fat-soluble vitamins.^{1, 10, 26, 28}

Energy requirements may also be increased as a result of the disease pathophysiology e.g. biliary atresia in addition to the impact of increased stress on the liver itself from concomitant co-morbidities such as infections.^{1, 10, 26}

6. Biochemistry

6.1 Acute Liver failure

A diagnosis of acute liver failure may be made by performing standard liver function tests and coagulation. Results will show:

- Hyperbilirubinaemia
- Raised aminotransferase (>10, 000 IU/L)
- Raised plasma ammonia (>100IU/L)
- Coagulopathy (prothrombin time > 40 seconds)²⁶

All children present with a coagulopathy in addition to jaundice and or encephalopathy depending on the extent of damage, vitamin K 2 – 10mg/day orally or IV is recommended.²⁶

Serial biochemistry values are recommended as frequently as indicated:

- Urea, creatinine, sodium, potassium
- Calcium, magnesium and phosphorus
- Glucose
- Albumin, ALT, AST, GGT, Bilirubin [conjugated, unconjugated]
- INR & Fibrinogen
- Hb, platelets, WCC
- Ammonia, baseline

A poor prognosis is indicated by:

- Prothrombin time >60 seconds
- Decreasing transaminase levels
- Rising bilirubin > 300mmol/l
- Decreasing liver size
- Acid base – pH 7.3
- Hypoglycaemia < 4mmol/l with increasing dextrose requirements
- Hepatic coma grade 2 or 3.²⁶

6.2 Chronic Liver failure

The above-mentioned biochemical values should be measured regularly. Measurement of individual nutrients such as vitamins and minerals is important for identifying deficiencies. Assessment should also include observations of physical signs of vitamin deficiencies, dietary intake, degree of early satiety and any self imposed restrictions. Nutrition related issues such as nausea, vomiting, anorexia and or diarrhoea should be considered, as they may impact on biochemistry.^{1,29}

6.3 Albumin

The measurement of biochemical parameters such as albumin is not useful as synthesis may be decreased, it has a long ½ life and a large extravascular pool. A

low albumin may be a prognostic marker reflecting the severity of the liver disease. Nitrogen balance studies are similarly difficult to interpret.^{1, 10, 21, 29}

6.4 Pre Albumin

Although not routinely done, prealbumin is a more sensitive index of nutrition status as the half-life is 24 – 48 hours. It is however, influenced by liver disease, infection and inflammation. Prealbumin levels may be inversely correlated to C-reactive protein [CRP]. It is difficult to discern whether low prealbumin levels with a high CRP are as a result of poor nutrition or inflammation. Results should therefore be interpreted with caution.²⁹

6.5 Transferrin and Retinol Binding Protein

Both are sometimes used as quasi nutrition indices. Transferrin is manufactured in the liver and is therefore influenced by liver disease and retinol-binding protein is affected by vitamin A and zinc deficiency.²⁹

6.6 Insulin-like Growth Factor (IGF-1)

Growth failure occurs in up to 60% of children as a result of decreased levels of insulin-like growth factor (IGF-1). Following liver transplant growth failure appears to continue to occur in 15 – 20% of children. Treatment with IGF-1 and/ or growth hormone may precipitate linear growth.^{16, 18, 24}

6.7 Immune Competence

Humoral immunity remains intact unless there is severe malnutrition. Cell mediated immunity is affected early by compromised nutrition status. The most common is a delayed cutaneous hypersensitivity. This can be affected by uraemia, infections, cirrhosis, and hepatitis amongst others.²⁹

6.8 Serum Cholesterol

Patients with malnutrition commonly have decreased levels of cholesterol. Very low levels occur in patients with liver and renal disease and malabsorption syndromes.²⁹

6.9 Cirrhosis and Portal Hypertension

Cirrhosis with portal hypertension is the end point for all children with chronic liver disease. There are two forms, namely, compensated and decompensated. Decompensation occurs when there is a loss of hepatic synthetic function in addition to the presentation of complications such as malnutrition, bleeding oesophageal varices, and encephalopathy as well as hepatorenal failure.²⁶

Most diagnoses are confirmed by means of histology (extensive fibrosis and regenerative nodules) but some biochemical and clinical findings include:

- An echogenic liver with splenomegaly and varices on ultrasound,
- Oesophageal and gastric varices on endoscopy,
- Mild transaminitis (aspartate aminotransferases and alanine aminotransferase > 3 x normal)
- Increase serum alkaline phosphatase and gamma glutamyltransferase (twice normal)
- Low serum albumin

- Low serum calcium and phosphate secondary to rickets
- Anaemia
- Prothrombin time > 20 seconds
- INR >1.5²⁶

7.Clinical

7.1.1 Physical Examination

Fat loss in a child may be a subjective indicator of nutrition status. In children with chronic liver disease this is most likely to be seen on the upper extremities such as the arms, which will appear wasted and thin. Table 3 outlines the physical signs of nutrient deficiencies.²⁹

Table 3: Physical signs of nutrient deficiencies.²⁹

Physical Sign	Possible Nutrient Deficiency
Oedema	Protein
<i>Signs of bone disease:</i> Thickened costochondrial junction Bossing (protruding forehead) Delayed fontanel closure Bow legs (once able to walk)	Vitamin D
<i>Skin Lesions</i> Petechiae, subcutaneous haemorrhages Perifollicular keratosis Scaly rash Red scaly rash	Vitamin C, K Vitamin A Niacin, tryptophan Zinc
<i>Head and Neck</i> Conjunctival changes (dryness, white plaques) Ophthalmoplegia Chelosis Angular stomatitis Glossitis	Vitamin A Thiamine Vitamin B complex Vitamin B complex Vitamin B complex
<i>Cardio-respiratory system</i> Heart failure with dyspnoea, peripheral oedema, tachycardia Severe acidosis causing tachypnoea	Thiamine Thiamine
<i>Nervous system</i> Peripheral neuropathy Myopathy with ataxia and retinopathy Subacute demyelination of the spinal cord	Thiamine Vitamin E Vitamin B ₁₂

7.1.2 Medical History

A thorough medical history forms an integral part of the past nutrition history. Any gastrointestinal problems such as vomiting, diarrhoea, type of stools should be elicited. Questions around oral function are important with respect to the presence of dysphagia and the ability to consume a normal diet, which is age appropriate.

Medication prescribed may also cause anorexia and gastrointestinal disturbances in addition to altering absorptive capacity of vitamins (e.g. Lactulose, antibiotics, antacids, Cholestyramine). A six monthly review of all medication is recommended. Caregivers should be encouraged to take note of problems associated with particular medication.

7.1.3 Psychosocial History

A review of psychosocial factors is important to establish which other members of a health care team should be involved e.g. social worker, speech therapist etc. Behavioural problems relating to food are not uncommon and are important to resolve prior to the implementation of planned nutrition support. It is important to determine whether or not the family is able to fund the nutrition support planned. Sensitive questions around home facilities, knowledge and skills to provide a nutritious diet should be elicited.²⁹ Referral to the Health Facility Based Nutrition Support Scheme may be appropriate using referral forms in *Appendix 3*. A letter of motivation for the provision of an MCT containing infant and paediatric complete nutrition supplement will need to be provided by the referring dietitian.

7.1.4 Acute Liver Failure

The clinical presentation depends on the aetiology of the acute liver failure and whether or not the presentation is acute i.e. within days, or prolonged i.e. within weeks, as seen in metabolic liver disease.²⁶

The main pathological features of acute liver failure are severe hepatic necrosis, loss of hepatic architecture and absence of hepatic regeneration.²⁶

Fulminant liver failure often results in hepatic encephalopathy. Common clinical signs of encephalopathy are:²⁶

- Irritability
- Drowsiness
- Coma
- Hepatomegaly
- Many causes of chronic liver failure, likely to present early in life
- Vomiting
- Poor feeding
- Switching of day/night sleep pattern
- Aggressive behaviour
- Convulsions

Support management of fulminant hepatitis includes²⁶:

- No sedation or lower doses, except for medical procedures
- Consider frusemide infusion or mannitol to prevent cerebral oedema and monitor the response with neuro observations every 4 – 6 hours
- Blood glucose > 4mmol/l
- Maintenance of appropriate acid-base balance
- Prothrombin time (PT), partial thromboplastin time, INR – coagulopathy treated with vitamin K, severe coagulopathy (PT > 60 seconds) managed with frozen plasma
- Fluid balance 75% maintenance
- Maintain circulating volume with colloid or fresh frozen plasma
- Coagulation support
- Drugs: Vitamin K (2 – 10mg/day) IV or orally, omeprazole, Sucraflata (2 – 4g/day), Lactulose (5 – 20ml/day), Neomycin, N-acetylcysteine (70mg/kg/6 hour), broad-spectrum antibiotic, mycostatin.
- Nutrition: enteral feeding (1 – 2 g/kg protein /day) parenteral nutrition where indicated.

7.2 Chronic Liver Failure

Cirrhosis represents ESLD occurring as a result of repetitive sequence of cell injury and repair. It leads to cyclical necrosis and fibrogenesis causing irreversible damage in addition to the primary disease process. Cirrhosis may be asymptomatic but can cause decompensation when damage to the liver causes blood flow to be impaired which results in portal hypertension, ascites and varices. Table 4 outlines the complications of liver cirrhosis in children.²⁶

Table 4: The complications of cirrhosis in children²⁶

<ul style="list-style-type: none">• Malnutrition and growth failure• Coagulopathy• Portal hypertension, hypersplenism, variceal bleeding, ascites and encephalopathy• Hepatopulmonary syndrome	<ul style="list-style-type: none">• Hepatorenal syndrome• Bacterial infections, spontaneous bacterial peritonitis• Hepatocellular carcinoma
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7.3 Ascites and Hepatomegaly

Patients with gross ascites and/ or hepatomegaly often experience anorexia due to decreased stomach volume/ capacity. During this time nutrition management goals extend to smaller frequent energy dense meals and/ or snacks in addition to supplementary nasogastric feeding where appropriate.^{1,10,16,26}

Ascites is often associated with fluid and sodium restriction although diuretics are often used in place of fluid restriction in order to promote adequate nutrition intake. In some cases patient's will self restrict intake due to abdominal discomfort, nausea and early satiety, in which case energy dense feeds are required, providing 1– 1.5kcal/ml. However, caution must be used with the higher end of the energy range as problems may occur with nausea, vomiting and/ or early satiety.^{1,10,16,26}

Care should be exercised when teaching parents to make enriched feeds at home especially when they are divergent from the manufacturer's instructions and/ or when more than one powder/ liquid is used e.g. milk with additional carbohydrate powder and fat. It is advised that test feeds are made up in front of the dietitian on more than one occasion to ensure caregivers have interpreted individually tailored special recipes properly.¹

It is recommended that nutritious liquids replace those with little or no nutritive value such as fruit juices or cool drinks.¹

7.4 Portal Hypertension and malabsorption

Patients with cholestatic liver disease resulting in cirrhosis can present with portal hypertension obstructing blood flow to the liver. This is associated with malabsorption in addition to enteropathy due to increased pressure on the mesenteric venous system.^{1,10}

Nutrition management varies and consensus is lacking however, continuous semi elemental feeds may increase absorption but in severe cases total parenteral nutrition (TPN) will be required.^{1,10}

7.5 Oesophageal Varices

A soft textured diet is recommended 24-48 hours post sclerotherapy and TPN is only required in severe cases of ESLD with huge bleeding varices. With large varice

bleeds, a nasogastric tube can be inserted during the procedure in theatre under guidance. This will enable early enteral feeding while trying to establish oral intake again.¹

8. Dietary

8.1.1 Overview

Potential problems requiring nutrition intervention occur when there is a disturbance of normal metabolic functions of the liver including:

- Glucose homeostasis
- Protein synthesis
- Bile Salt production
- Lipid metabolism
- Vitamin absorption and storage²⁶

8.1.2 Diet Therapy includes management of:

- Hypoglycaemia due to poor glycogen storage
- Fat malabsorption as a result of poor bile production or flow
- Reduction in protein synthesis especially albumin exacerbating ascites
- Vitamin deficiencies due to malabsorption of fat soluble vitamins²⁶

At each follow up a thorough nutrition history should be completed collecting information around the following areas:

- Weight change
- Appetite
- Satiety level
- Taste changes/ aversions
- Nausea/ vomiting
- Bowel habits – constipation, diarrhoea
- Chewing/ swallowing ability
- Pain when eating
- Long-term disease(s) affecting absorption/use of nutrients
- Surgical resection or disease of GI tract
- 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 (food security at home)
- Use of any other or unprescribed, over the counter medications, vitamin and mineral supplements and herbal remedies.^{26,29}

Nutrition counselling regarding appropriate and healthy food choices should be provided in line with the paediatric food based dietary guidelines. Additional information should be provided to caregivers on how to enrich foods using common household items such as margarine, butter, oil, sugar, full cream milk powder and peanut butter. As children's diets vary it is important to gather a 3-day diet history, where possible.²⁹

In addition to this it is important to establish whether any traditional medicines, over the counter vitamin and mineral supplements and any other herbal remedies are being used.²⁹

8.2 Acute Liver Disease

There is generally no restriction on protein intake in this group except for those with fulminant liver failure and encephalopathy where protein restrictions will apply. However it is important not to over restrict, as this will promote endogenous ammoniogenesis.^{1, 19, 20}

In older children an undiagnosed inborn error of metabolism is unlikely except in the case of Wilson's disease so normal enteral feeds may be used allowing nutrition requirements to be met timeously.¹

8.3 Fulminant Liver Failure with encephalopathy

Infants may receive 1.0 – 1.5g/kg/day of protein and children or teenagers should get between 0.5 – 1.0g/kg/day of protein. Glucose infusion should provide 6 – 8 mg glucose/min/kg day in order to prevent hypoglycaemia and protein catabolism. Infusions should provide sufficient potassium in order to prevent hypokalaemia especially when there is concomitant use of a diuretic. Some centres use BCAA containing formula in HE, as it appears there may be some improvement in symptoms.¹

8.3.1 Step 1 – Emergency Regime

Aim: To meet glucose oxidation rate and prevent endogenous protein catabolism.

- Provide carbohydrate in the form of glucose for the first 20 – 48 hours.
- IV Dextrose with potassium if diuretic therapy is being prescribed.
- Enteral Feeding: Glucose polymer added to oral rehydration solution – lactose/fructose free.
- Additional sodium and potassium should be added to the feed as required.
- Feed continuously over 24 hours

Guidelines for Glucose requirements

- Glucose rates may be increase according to tolerance.
- Infants 8-9mg glucose/kg/min or (11.5 – 12.9g/kg)
Can tolerate up to 12.5mg glucose/kg/min or 18g/kg/day
- Toddlers 7mg glucose/kg/min or (10g/kg)
- Adolescents 4mg glucose/kg/min or (5.7g/kg)¹

8.3.2 Step 2 – Addition of protein

- Protein should be added to the feed within 20 – 48 hours post onset of encephalopathy.
- The feeds should contain a lactose free protein source e.g. soya-based formula until galactocaemia is ruled out and should be fat free until fatty acid oxidation defects are excluded.
- A child with an inborn error of any type should tolerate some protein containing all amino acids.

8.3.3 Protein requirements – WHO minimum requirements

- Levels of consciousness and ammonia should be checked daily before additional protein is given.

- It is recommended that the amount of protein provided be a minimum of 0.5 – 1.0g/kg, increased by 0.5g/kg daily until the goal amount is reached:
 - Infants: 1.5 – 1.9g/kg/dry weight/day
 - Children: 0.8 – 1.0g/kg/dry weight/day¹

8.3.4 Step 3 – Addition of Fat

- The addition of fat enables energy and nutrition requirements to be met as soon as possible, preventing the build up of toxic metabolites
- Increments of 1% of fat per 24 hours or 0.5g fat per 100ml per day are recommended in the form of Duocal, Liquigen, Calogeen or long chain fat [Canola or olive oil, not sunflower oil].
- In the cases of severe fat restriction and to prevent essential fatty acid deficiency (EFA) walnut oil is recommended at 2ml per 100kcal.
- Energy requirements can also be met by adding Energivit (SHS) which provides fat, carbohydrate and micronutrients.¹

8.4 Chronic Liver Disease

8.4.1 Energy

Although significant changes in energy metabolism are anticipated in liver disease there is very little research regarding this in chronic liver disease in children. Two studies considering extrahepatic biliary atresia noted resting energy expenditure (REE) was increased by 30%. However, REE was only found to be 48% of total energy expenditure (TEE). This was thought to be related to a hyperdynamic circulation in addition to deranged metabolism. It is recommended that REE values be used as a baseline with 50% additional calories being provided in order to accommodate the increase in TEE.^{10,29} If available indirect calorimetry measuring REE should be performed on children with chronic liver disease at each follow up visit.²⁹

Energy requirements are dependant on the age of the child, disease staging, nutritional status and degree of malabsorption. Infants are estimated to require between 120 - 150kcal/kg/day with 50% of the dietary fat being derived from MCT's. In older children 1.2 – 1.5 x DRV [*Appendix 1: Table 1*] for energy is recommended.^{1, 10, 16, 26} Alternatively, predictive equations may be used to calculate energy requirements. [*Appendix 1: Tables 2-4*]

The caloric density of the feed may be increased to 1kcal/ml in infants and up to 1.5kcal/ml in children. This may be achieved through a sterile ready to use/ hang energy dense feed or by the addition of modulars such as glucose polymer and LCT/ MCT powders.^{1, 10, 16, 26}

Tolerance of the feeds should be closely monitored. Breast-feeding should be encouraged and can be maintained as long as there is sufficient growth. Breast milk may be fortified with combined fat and carbohydrate powders. The addition of protein powders is not recommended.¹

In children who are unable to take sufficient calories by mouth during the day nocturnal enteral feeds should be considered via the naso gastric route. Percutaneous gastrostomies (PEGs) are relatively contraindicated in this population group however, some success has occurred in patients with Alagielles syndrome.³⁵

8.4.2 Protein

Typically, protein oxidation in healthy infants occurs at around 4 – 9%. Deranged protein metabolism has been demonstrated in children with biliary atresia and protein oxidation was around 17% despite adequate nutrient intake and a positive nitrogen balance.¹⁰

Abnormalities of albumin and muscle protein turnover have been demonstrated in cirrhotic liver patients. The possible reason for this is multifactorial including hormonal and metabolic abnormalities in addition to upregulation of proinflammatory cytokines.²²

Sufficient protein is required in order to prevent the endogenous breakdown of protein stores. For infants a goal of 3-4g/kg/day is recommended. In children over 1 year of age the recommendation for protein is 2.5 – 3g/kg. Protein oxidation in children occurs maximally at 4g/kg. Protein given in excess of this will be futilely oxidised increasing the risk of metabolic acidosis and hypertonic dehydration and exacerbating any hepatic/ renal impairment.^{1, 10, 16}

8.4.3 Branched Chain Amino Acids

The liver is the primary organ for metabolism of various amino acids with 40 – 60% metabolised in a single pass through the liver. Protein breakdown from the muscles, of the BCAAs in particular (valine, isoleucine and leucine), provides energy through gluconeogenesis. Abnormal amino acid profiles are seen in liver disease. Plasma concentrations of aromatic amino acids increase as levels of the BCAA decrease and this in turn is thought to correlate with the degree of encephalopathy.²⁻⁸

The use of BCAA in the management of HE remains controversial in both adults and paediatrics. Clinical studies in critically ill HE patients have shown the following benefits of BCAA given on their own or in the form of a BCAA enriched parenteral nutrition solution:²⁻⁸

1. Improved nitrogen retention and balance
2. Stimulation of protein synthesis
3. Improved visceral protein status
4. Improved immune function parameters
5. Normalisation of plasma amino acid profile

However none of these effects have changed the morbidity or mortality rates associated with injury and sepsis.⁸

If portal systemic encephalopathy develops, negative nitrogen balance is exacerbated by dietary protein restriction. BCAA appear to be primarily metabolised extrahepatically in skeletal muscle and used as a source of glucose in catabolism, thus having less impact on protein loads than other sources.^{2-8, 27}

As a result supplementation may allow a higher protein intake without deterioration in mental state. In addition to this, leucine stimulates muscle protein synthesis and inhibits protein breakdown. In patients given isonitrogenous parental nutrition, a 45%

BCAA rich formula was associated with increased pre-albumin, lower mortality rate, but no difference in length of hospital stay.²⁻⁸

The Fischer molar ratio is the difference between the serum concentration of BCAA and aromatic amino acids [Valine, Leucine, Isoleucine vs. Phenylalanine and Tyrosine]. It has been shown to correlate with intrinsic liver function test values that yield abnormal results when there is functional impairment of liver cells. The Fischer ratio also correlates with pre albumin and albumin levels.²⁴

A Fischer ratio of below 2.0 is common in children with persistent jaundice and is likely to be a predictor of growth disturbance. The Fischer molar ratio may prove to be a useful index of intrinsic liver function and nutritional status on which to base nutrition supplementation. In studies completed comparing the concentrations of BCAA with the Fischer ratio a correlation was found to occur between the supplementation of BCAA and an improvement in the Fischer ratio.²⁴

The amount of BCAA orally supplemented in studies ranges from 2.5% - 50% [total protein], which is equivalent to 0.2g – 2g/kg/day BCAA. The BCAA supplement is provided in addition to the normal dietary protein content.²⁻⁸ Most standard enteral feeds have a BCAA content between 16 - 53% of the protein content, however BCAA enriched formulations will contain a significantly higher percentage of BCAA when compared to standard supplements. The cost benefit ratio of these expensive products has not been fully elucidated.²⁴

In both adults and children with ESLD, significant improvements in weight gain, liver function and quality of life were found when a supplementation of 0.272g/kg/day of BCAA was given over a period of 1 year. The BCAA requirements of children with ESLD have been found to be significantly higher than age matched healthy children.²⁷

Although promising results have been shown with respect to improvement in nutrition status of children receiving a BCAA enriched product further studies are required to elucidate whether there is a dose related response and what the ideal amount of BCAA per kg may be.⁷

8.4.4 Fat

One of the liver's many functions is the transport and synthesis of lipids. In cholestatic liver disease the amount of bile salts in the GI tract is decreased whereas serum concentrations of bile salts become elevated leading to pruritis.^{10, 35, 36}

A deficiency of bile salts results in growth failure due to fat malabsorption, fat-soluble vitamin and some mineral deficiencies. Studies have shown that even when infants are supplemented with fat there is a decreased level of cellular DHA impacting on neurodevelopmental function.¹⁰ This may remain for up to one year post transplant in some patients and is thought to be as a result of a defect in the delta 5 desaturase pathway. Appropriate dietary approaches have yet to be elucidated.³⁷

Fat restrictions are not recommended in the management of chronic liver disease, especially in lieu of EFA deficiencies and should instead be given according to tolerance. However, children with cholestatic disease may have significant steatorrhoea and a reduced fat diet may be better tolerated e.g. Alagielles Syndrome.
1, 10, 16, 26

Medium chain triglycerides (MCT's) may be useful in the management of these children as they do not require micelle formation and are absorbed via the portal vein. The use of MCT containing products has been shown to decrease the incidence of steatorrhoea and promote growth. MCT's can be given in small doses throughout the day either mixed into food, sauces or beverages. However as they do not contain EFA other fat sources should also be provided.^{1, 10, 16, 26, 35, 36}

MCT's have a high osmolality (> 600mOsmo/kg) which may result in abdominal cramping, diarrhoea and nausea. In high amounts they can also be neurotoxic thus judicious use is advised. Where required a dose of 0.3g/kg is recommended. It is advisable not to cook with MCT oil as it has a low temperature threshold, however it may be added to food once cooked.²⁴

8.4.5 Long Chain Polyunsaturated Fats (LPUFA's) and ESLD

Paediatric patients with cholestatic liver disease often have PUFA deficiency as a result of nutrition and metabolic factors. Since PUFA deficiency may develop early in the disease dietary intervention is vital to promote growth and prevent neurodevelopmental delay in addition to concomitant deficiencies of fat soluble vitamins especially vitamin E.⁹

PUFA depletion may occur as a result of enhanced oxidation of fat body mass for energy. In addition to this, low levels of antioxidants such as vitamin E may result in lipid peroxidation contributing further to their depletion. Children with cholestasis are prone to free radical damage but the clinical significance of this remains unknown.⁹

Providing PUFA's in the form of linoleic acid (LA) and α -linoleic acid (ALA) may not be sufficient as a result of impaired conversion of EFA with cholestasis and associated hepatocyte injury. Increased levels of mead and palmetic acids are shown in this patient population, which can affect immune function, platelet aggregation, clotting profile and inflammatory markers. LA and ALA were deficient compared to docosahexanoic acid (DHA) and arachidonic acid (AA) but appeared to improve when supplemented with DHA and AA.⁹

8.4.6 Fat Soluble Vitamins

Fat soluble vitamin deficiencies are well described in cholestatic liver disease as a result of fat malabsorption due to the absence of bile in the gastrointestinal tract. Aggressive supplementation of all fat-soluble vitamins is recommended in addition to regular monitoring for signs of deficiencies [Table 6].^{1, 10, 16} Supplements may be given orally in doses of 3 – 5 times the normal requirements if cholestasis is incomplete. However, in complete cholestasis, supplements should be given intravenously, where possible, once a month.³⁵

Table 6: Recommendations for vitamin supplementation in cholestatic liver disease

Fat Soluble Vitamins and the measuring units	Amounts
Vitamin A (aqueous)	• 10 – 50, 000 IU
• Levels may be tested via plasma retinol/ RBP	
25-OH vitamin D	• 2 – 4 ug/kg/day
• Levels may be tested via plasma 25 – OHD	
Vitamin E	• 25 IU kg/d
• Levels may be tested via plasma E/ total lipids	• As d-alpha tocopheryl polyethylene glycol-1000 succinate
Vitamin K	• 2.5 – 10mg/d
• Levels may be tested via prothrombin time.	
Water Soluble Vitamins	Amounts
• Consider multivitamin	• 5 – 10ml per day

In place of supplementing each vitamin, mineral individually a fat-soluble vitamin liquid preparation can be given. Cernevit [Baxter] is an intravenous preparation and ADEK® [Pharmaplan] is an oral preparation. Both are available locally and contain high levels of fat-soluble vitamins A, D, E and K. The vitamins are present in their water miscible forms, which is thought to improve their absorption. ADEK® also contains vitamins C, B complex, folic acid and zinc [Table 7 & 8]. Tablets should be thoroughly chewed or crushed prior to swallowing.

Table 7: ADEK® Vitamin Composition and age requirements

	% Daily values for children			% Daily values for adults		
	Amount per ml	0-6 months % DRI	7 – 12 months % DIR	1 – 3 years % DRI	Per table	% Based on 2,000kcal diet
Calories	5					
Total CHO	1g					
Vitamin A (as palmitate and 50% as beta carotene)	3170 IU	250	250	240	9000 IU	180
Vitamin C (as ascorbic acid)	45mg	150	130	115	60 mg	100
Vitamin D (as cholecalciferol)	400 IU	75	100	100	400 IU	100
Vitamin E (as d -alpha tocopheryl)	40 IU	900	670	450	150 IU	500
Vitamin K (as phtonadione)	100 mg	*	*	*	150 ug	190
Thiamine	0.5 mg	165	125	70	1.2 mg	80
Riboflavin	0.6 mg	150	120	75	1.3 mg	80
Niacin (as niacinamide)	6 mg	120	100	65	10 mg	50
Folic acid	-	-	-	-	0.2 mg	50
Vitamin B6 (as pyridoxine)	0.6 mg	200	100	60	1.5 mg	80
Vitamin B12 (as cyanocobalamine)	4 mg	1300	800	570	12 ug	200
Biotin	15 mg	150	100	75	50 ug	15
Pantothenic acid (as d-pantothenol)	3 mg	150	100	100	10 mg	100
Zinc (as sulphate)	3 mg	100	100	50	7.5 mg	50

* No DRI established

Table 8: Recommended dosage of ADEK® vitamin preparation

Liquid		Tablet	
0 – 12 months	1 ml per day	4 – 10 years	1 tablet per day
1 – 3 years	2 ml per day	> 10 years of age	2 tablets per day

8.4.7 Vitamin A

Up to 20 – 50% of children with chronic cholestatic liver disease have low retinol concentrations, which correlate with the duration and intensity of the liver disease. A retinol range of > 20ug/dl is considered acceptable. Some centres advocate the use of up to 25, 000 – 50, 000 IU of vitamin A esters although the safety and efficacy of this remains unclear.^{1,10}

8.4.8 Vitamin D

Vitamin D deficiency is associated with rickets and is commonly seen in patients with biliary atresia. Even large doses of vitamin D₂ may be malabsorbed in cholestatic liver disease. Absorption of 25 - hydroxycholecalciferol (25-OHD) is better and the provision of 2 – 4 ug/kg/d is recommended.^{1, 10}

8.4.9 Vitamin E

Vitamin E deficiency presents with peripheral neuropathy, ataxia, ophthalmoplegia, staggering gait and muscle weakness, which is reversible if treated before the age of 3 years. Biochemical vitamin E deficiency is treated with < 0.8mg total tocopherol/g total lipid in older children and adults and with <0.6mg tocopherol/g total lipid in children under the age of 12 years.^{1, 10}

8.4.10 Vitamin K

Assessed indirectly through prothrombin time and INR, which is dependant on vitamin K clotting factors. A dose of 2.5 – 5mg given orally, two to seven times per week is recommended. Alternatively, between 2 – 5 mg is administered monthly via a slow intravenous infusion or by the intramuscular route, in those children with severe cholestasis.^{1, 10}

8.4.11 Monitoring

Careful monitoring is required of all patients receiving fat soluble vitamin supplementation to avoid either deficiency or toxicity.¹⁰

8.4.12 Water Soluble Vitamins

The incidence of deficiencies of water-soluble vitamins is considered to be relatively rare and may only occur in those patients with profound enteropathies.^{1, 10}

8.5 Complications of Chronic Liver Disease

Complications of chronic liver disease, which require nutrition intervention, are outlined in the following paragraphs.

8.5.1 Jaundice

Jaundice indicator of significant hepatobiliary disease and cholestatic liver disease. High conjugated bilirubin may result in reduced bile flow from the liver into the gut.^{1, 26, 28, 29}

The nutrition implications are that fat emulsification and digestion do not occur normally. This results in malabsorption of fat, fat soluble vitamins and some minerals. Steatorrhea, growth failure and rickets are common in children with chronic liver failure.^{1, 26, 28, 29}

8.5.2 Portal hypertension & malabsorption

Cirrhosis can obstruct blood flow leading to portal hypertension with associated enteropathy and malabsorption secondary to increased pressure in the mesenteric venous system. Nutrition management is difficult in these cases and sometimes a semi elemental and or continuous enteral feeds may help. If malabsorption continues TPN may be required.^{1, 26, 28, 29}

8.5.3 Fat Malabsorption

Where there is fat malabsorption, fat should not be restricted but given to tolerance. Infants require a significant amount of fat for growth and development. Up to 50% of total fat content may be provided as MCT's.^{1, 26, 28, 29}

8.5.4 Hypoglycaemia

Infants may require overnight continuous feeds with up to 6g/ 100ml carbohydrates. In children who can eat, complex, slow release carbohydrates should be given.²⁶

8.5.5 Ascites and hepatomegaly

With ascites and hepatomegaly there is reduced abdominal capacity for food/ feeds. Smaller, frequent, energy dense meals/ snacks are recommended. Where there is volume restriction, a lower sodium feed e.g. 1.2 – 1.5mmol/l may be required, however use of diuretics are favoured.^{1, 26, 28, 29}

8.5.6 Oesophageal varices

Occasionally in ESLD a huge bleed from oesophageal varices will require TPN. If this is not required clear fluids followed by a soft diet may be provided 16 – 24 hours post sclero therapy. With large varice bleeds, a nasogastric tube can be inserted during the procedure in theatre under guidance. This will enable early enteral feeding while trying to establish oral intake again.^{1, 26, 28, 29}

8.5.7 Chronic encephalopathy

As discussed earlier protein restrictions can significantly decrease nutrition status, however a protein restriction to 1 – 2 g/kg may be required. The degree of encephalopathy should determine the level of restriction. In order to ameliorate gluconeogenesis, the energy ratio should be increased to prevent endogenous muscle breakdown. Sodium benzoate or the use of BCAA may allow for higher protein tolerance.^{1, 26, 28, 29}

8.6 Dietary Interventions

All children with growth failure should receive nutrition supplementation in order to attain a good nutrition status and linear growth.^{1, 26, 28, 29}

Nutrition support in children awaiting liver transplant may take the form of MCT enriched, age appropriate nutritionally complete products e.g. Pre Term formula for infants. The enrichment of food should be encouraged using household store items such as margarine, butter, oil, sugar, peanut butter, full cream milk powder etc.

In those children not awaiting transplant a more conservative approach may be adopted, however, it is important to maintain nutrition status in these children in order to provide a good quality of life. Nutrition support should be provided in the form of nutritionally complete maize or cow's milk based drinks.

All growth failing children should be referred to the NSP scheme for long term, ongoing nutrition support. Anthropometry should be reviewed regularly and entry and exit criteria adhered to. Private medical aid members should seek nutrition support through their medical aid with supporting motivation from the medical team including a dietitian.

8.6.1 Nutrition Supplementation

The following recommendations should be applied for choice or feed and or supplementation for children with chronic liver disease [Table 9]. If modulars are to be used it is important to calculate the amount of additional protein, fat, carbohydrate and sodium per 100ml and per kg actual body weight to ensure recommendations are not being exceeded.

A 1kcal/ml RTU infant or 1.5kcal/ml RTU paediatric feed should be used, when an energy dense feed is required. Modulars should not be added to RTU feeds and should be given as a bolus flush with sterile or boiled water [infants] prior to a feed. Modulars should be used with caution as they increase the osmolality of feeds and may result in vomiting, nausea and diarrhoea if used incorrectly.

If a recipe for an enriched powdered feed with modulars is to be used at home it is imperative this be provided as a written recipe. Verbal instructions, including telephonic recipes should not be given due to the risk of annotation errors. In addition the caregivers should be requested to make the recipe up a number of times, on separate occasions, in front of the dietitian prior to being discharged.

Table 9: Summary of macronutrient supplementation guidelines

	Infants	Children
Energy Concentration	<ul style="list-style-type: none"> ▪ 0.67 – 0.74 – 1kcal/ml ▪ Standard – Pre Term – Energy enriched [RTU] 	<ul style="list-style-type: none"> ▪ 1 – 1.5kcal/ml • 2kcal/ml not recommended due to early satiety, nausea, vomiting.
Protein: <ul style="list-style-type: none"> ▪ Protein supplementation is rarely required without an accompanying > in energy. 	<ul style="list-style-type: none"> ▪ Protein supplementation in the form of a protein powder is not recommended in children < 1 years of age. ▪ 7.5% - 12% energy from protein ▪ For “catch up growth” at least 9% of energy from protein will need to be provided. 	<ul style="list-style-type: none"> ▪ Protein may be added to feeds in the form of amino acids, whole protein or peptides. ▪ 5 – 15% in older children. ▪ For “catch up growth” at least 9% of energy from protein will need to be provided.
Carbohydrate: <ul style="list-style-type: none"> ▪ Glucose polymers are preferred lower osmotic effect add in small increments e.g. 1% per 24 hours. 	<ul style="list-style-type: none"> ▪ 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 	<ul style="list-style-type: none"> ▪ 15-20% in toddlers aged 1-2 years ▪ 20-30% in older children
Fat: <ul style="list-style-type: none"> ▪ Fat emulsions are preferred and should be added in small increments until goal amount is reached e.g. 1% or 0.5g fat per 100ml per 24 hours. 	<ul style="list-style-type: none"> ▪ Infant’s will tolerated a total fat concentration of 5 – 6 % [e.g. 5 – 6g per 100ml of feed]. ▪ 40% fat total energy of which: ▪ 40 – 50% MCT’s ▪ LCFPUFA’s 	<ul style="list-style-type: none"> ▪ Children > 1yr will tolerate a fat concentration of 7% - concentrations above this may cause nausea/ vomiting. ▪ 30 – 35% total energy ▪ LCPUFA’s better tolerated.
Osmolality [osmo]:	<ul style="list-style-type: none"> ▪ Standard dilution infant feeds [incl. Pre term] osmo/l141 – 169mosmo/l. ▪ 1kcal/ml RTU 283 osmo/l [prescribed under medical supervision] ▪ osmolality of > 277mosml [FDA] is not recommended to children < age of 1 years. 	

8.7 Enteral Feeding and TPN

Fine bore polyurethane tubes (Fr size 6 – 8), are recommended for use in ESLD especially where there are varices. Continuous, bolus or nocturnal feeds may be provided via an enteral feeding pump. Continuous enteral feeds are usually only warranted in severe cases of malabsorption or where there is hypoglycaemia. ^{1, 10, 24}

Lipid containing TPN may be provided in patients with chronic and acute liver disease where appropriate, without cholestasis.^{1, 10} The best form of lipid is one which contains a mixture of MCT and LCT fat emulsions in a 50:50 split, which are currently available in South Africa in the form of a structolipid.²⁴

8.8 Gastrostomy

The placement of a gastrostomy or PEG is contraindicated in liver disease, especially if there is ascites due to increased risk of peritonitis as well as problems arising from adhesions to the abdominal cavity. However, they have been placed in exceptional circumstances e.g. chronic cholestasis, Alagilles syndrome, post transplant with failure to thrive.^{1, 12, 13, 10}

8.9 Nutrition Support Post Orthotopic Liver Transplant

Liver transplant is considered in children with chronic liver failure and end stage liver disease. The indications for transplant include acute liver disease with fulminant liver failure, chronic liver failure, inherited metabolic disease and unresectable liver tumours.³³

Useful indicators as to when a liver transplant becomes necessary include:

- Persistently raised total bilirubin >150 umol/l.
- Prolonged prothrombin ratio/ INR > 1.4
- Fall in serum albumin <35g/l
- Decrease in psychosocial development.
- Failure to thrive despite aggressive nutrition intervention.
- Chronic encephalopathy
- Refractory ascites
- Recurrent variceal bleeding despite optimum management
- Intractable pruritis
- Severe metabolic bone disease
- Diminishing quality of life²⁶

The majority of transplants occur in children with biliary atresia. Survival rates are more than 90% at one year and 80% at 10 – 15 years. Aggressive nutrition support pre and post transplant contributes to the increased survival.^{11, 26, 33}

Due to the nature of liver transplant in children a Roux loop is often fashioned from a loop of bowel through which the bile will drain from the transplanted liver. Enteral feeding is often delayed for 3 – 5 days post surgery.^{1, 11, 10, 16, 33}

Parenteral nutrition (PN) may be considered in children post transplant if the initiation of enteral feeds is delayed beyond 5 days. If enteral feeds have not been commenced 72 hours post surgery PN should be provided. Lipid free PN should only be considered if a patient has elevated triglyceride levels, indicating poor fat utilisation.^{34, 38}

A good enteral nutrition intake usually occurs by day 5-post transplant. Semi elemental feeds are usually commenced in the immediate post operative phase with a graded progression to polymeric, MCT containing feeds if there is no evidence of diarrhoea often associated in the early post transplant period.^{1, 10, 16, 34}

Catch up growth post transplant is most effective in children under the age of 2 years and occurs less so in extremely nutritionally compromised older children.^{11,17} If there is continued growth failure following transplant, then IGF-1 levels should be measured with the possibility of providing growth hormone to promote catch up linear growth.¹⁰

Post transplant, and once catch-up growth has been achieved where necessary, care should be taken to ensuring age appropriate healthy dietary guidelines. Steroid and immune suppressing medication, physical inactivity and unhealthy eating habits can all contribute to an increased risk in these children of developing obesity, insulin resistance and steroid-induced diabetes.

9. Summary

Acute liver failure rarely results in nutrition compromise. In contrast, one of the hallmarks of chronic liver failure is growth failure with 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 chronic liver failure.

All children with growth failure should be provided with nutrition support to attain optimal nutrition status and support linear growth. They should be referred to the Nutrition Supplementation Programme (NSP) and or motivations written to medical aid schemes to provide appropriate supplements on a regular basis. The enrichment of food should be encouraged with small frequent meals and snacks.

Children awaiting liver transplant should receive nutrition support, which contains MCT oil. The addition of modulars may play a role in attaining required energy although caution should be exercised in prescribing additives as they can cause nausea, early satiety, diarrhoea and vomiting.

10. 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

11. Appendix 2: ²⁵

King's College Hospital, Special Investigations protocol for acute liver failure

Biochemical Tests

- Liver function Tests (Total & direct bilirubin, AST, ALT, GGT, ALP, albumin)
- Blood sugar
- Serum calcium, phosphorus, magnesium
- Uric Acid
- Cholesterol
- Triglyceride
- Amalyse
- Alpha-1 antitrypsin phenotype
- Galactose –1 phosphate uridyl transferase (in infants and neonates)
- Serum copper & caeruloplasmin (in children 3 years old)
- Serum aminoacids
- Blood gas analysis

Haematological Tests

- Full blood count
- Reticulocyte count
- Prothrombin time or INR
- Blood for grouping & cross matching
- Direct coombs test
- Bone marrow examination (in seronegative hepatitis or haematological malignancy or HLH is suspected)

Ultrasound

- US scan of abdomen especially liver, portal and hepatic vein, inferior vena cava, biliary system and spleen
- Microbiological Tests
- Bacterial cultures: blood, urine, stool, throat swab, sputum, skin lesion if present, ascetic fluid if present.
- Viral culture of urine and skin lesions if present

Serological tests

- Viral hepatitis: anti HAV IgM antibody, HbsAg, HB core antigen, Hepatitis D antigen and antibody, anti hepatitis C antibody, anti hepatitis E antibody.
- CMV
- EBV
- HIV
- Measles
- Varicella
- Herpes simplex virus
- Adenovirus
- Echovirus

Immunological tests

- Immunoglobulins (IgG, IgA, IgM)
- Tissue antibodies (anti SMA, GPC, anti SLA, LKM and anti nuclear antibody)
- Complement C3 & C4
- Ascitic fluid or cerebrospinal fluid cytopsin for evidence of hemophagocytosis
- Urine
- Toxicology
- Chemical analysis, osmolality and electrolytes
- Succinyl acetone
- 24 hour urinary copper pre penicillamine and post penicillamine (2 doses of 500mg 12 hours apart)

Tissue studies

- Buccal mucosal biopsy
- Skin fibroblast culture

APPENDIX 3

Nutrition Risk Score

Patients Name ----- Ward -----
 Hospital Name ----- Date -----
 Date of birth ----- Height/ Length -----

Please circle relevant score. Only select one score from each section. Select the highest score that applies.

COMPLETE ON ADMISSION AND WEEKLY IF PATIENTS CONDITION HAS CHANGED

1

Paediatrics (0-17 years) score

Adults (18 years) score

Present Weight

Expected weight for length	0
90-99% of expected weight for length	2
80-89% of expected weight for length	4
<79% of expected weight for length	6

Weight loss in last 3 months (unintentional)

No weight loss	1
0-3kg weight loss	2
>3-6kg weight loss	3
>6kg weight loss	3

2

Omit question 2
For paediatrics

BMI (Body Mass Index)

20 or more	0
18 or 19	1
15-17	2
Less than 15	3

3

Appetite

- Good appetite, manages most of 3 meals/ day (or equivalent) 0
- Poor appetite, poor intake – leaving > half of meals provided (or equivalent) 2
- Appetite nil or virtually nil, unable to eat. NMB (No food for > 4 meals) 3

4

Ability to eat/ retain food		
•	No difficulties in eating, able to eat independently No diarrhoea or vomiting	0
•	Problems handling food e.g. needs special cutlery 1 Vomiting/ frequent regurgitation (or possetting)/ mild diarrhoea	
•	Difficulty swallowing, requiring modified consistency. Problems with dentures, affecting food intake. Problems with chewing affecting food intake. Slow to feed. Moderate vomiting and/or diarrhoea (1-2/day children) Needs help with feeding (e.g. physically handicap)	2
•	Unable to take food orally. Unable to swallow (complete dysphagia) Severe vomiting and/or diarrhoea (>2/ day for children). Malabsorption	3

5

Stress Factor		
•	No stress factor (includes admission for investigations only)	0
•	Mild Minor surgery. Minor Infection	1
•	Moderate Chronic disease. Major surgery/ infarctions Fractures. Pressure sore/ ulcers. CVA Inflammatory bowel disease. Other gastrointestinal disease	2
•	Severe Multiple injuries. Multiple fractures/burns Multiple deep pressure sores/ ulcers Severe sepsis. Carcinoma/ malignant disease	3

Total

Nutrition Risk Score Results

Score	Action
0 – 3 Low Risk	No action necessary Check Weight weekly
4 – 5 Needs Monitoring	Check weight weekly Encourage eating & drinking Replace missed meals with Supplements. (Check with Dietitian if on special diet) Repeat scores after 1 week refer to dietitian if no improvement
6 – 15 High Risk	Refer to dietitian as soon possible

ALSO REFER TO DIETITIAN IF:

- The patients needs a special diet not available on the normal menu
- The patient needs advice about a special diet

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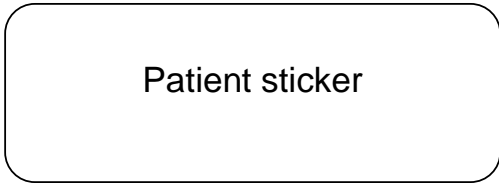
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13. Appendix 3: NSP Referral Form

REFERRAL TO THE NUTRITION SUPPLEMENTATION PROGRAM OF THE INTEGRATED NUTRITION PROGRAMME, WESTERN CAPE

COMPLETE IN QUADRUPPLICATE:

- Copy 1: (white) give copy to patient's mother or caregiver at the clinic
- Copy 2: (green) send copy to referred health facility
- Copy 3: (pink) place the copy in the patient's medical folder
- Copy 4: (blue) send to regional INP manager/dietician in sub-district



Patient's name:

Address:

..... Tel no:

Referred to:

Age: years Diagnosis:

Current weight: kg Length: cm BMI:

This patient meets the **inclusion criteria** to the Nutrition Supplementation Program of the HFBNP (*Health Facility Based Nutrition Program) as marked by ☒. It would be appreciated if you could supply the patient or infant with the necessary supplementation.

Thank you.

Date: / /
.....

Referring dietician's/doctor's/nursing officer's name:

Referring health facility: Tel:

Dispensing person's signature:

Comply with the following criteria:

1. Entry criteria for nutritionally at risk pregnant women:

Teenage pregnancy (∴ patient under 18 years of age)	
A history of low birth-weight, preterm (< 37 weeks gestation), Small for gestational age and underweight for gestational age	
Short birth intervals of less than 12 months	
Insufficient growth according to curve on symphysis-fundus graph on antenatal chart	
Mid-Upper-Arm-Circumference (MUAC) < 23 cm	

2. Entry criteria for underweight lactating women:

Her young infant (0 – 6 months) has growth faltering ⁱ for 2 consecutive VISITS on the RTHC ⁱⁱ	
Her older infant (6 to 12 months) has growth faltering for 2 consecutive MONTHS on the RTHCard	

3. Entry criteria for infants and children:

Infant 0 – 6 months and growth faltering for 2 consecutive VISITS on the RTHC and not on PMTCT ⁱⁱⁱ -program (Mother unable to breastfeed)	
Infant 6 – 12 months and growth faltering for 2 consecutive MONTHS on the RTHC and not PMTCT-program	
Infant 12 – 60 months (1 – 5 years) and growth faltering for 2 consecutive MONTHS on the RTHC	
Children 6 to 14 years who are chronically ill and growth faltering for 2 consecutive MONTHS on the RTHC	

4. Entry criteria for adults (> 14 years):

The patient is underweight (∴ BMI ≤ 18,5)	
The patient unintentionally lost more than 10% of his/her weight during the past 6 months	
14- 18 years who are chronically ill and growth faltering for 2 consecutive MONTHS on the Growth Cards	

ⁱ **Growth faltering** is when an infant's growth curve flattens or dips over two consecutive visits (aged < 6 months) or months (aged > 6 months) on his/her Road-to-Health card

ⁱⁱ **RTHC** – Road-to-Health Card

ⁱⁱⁱ **PMTCT** or Prevention of Mother-to-Child-Transmission p

ADDENDUM 4