“Recently, nutrition has taken its place along with surgical techniques and drugs as therapy for diseases.” (Ekvall SW, et al. 2005)

Nutrition intervention strategies for children with chronic diseases and developmental disorders must have both short- and long-term goals.
• **Short-term goals** involve support of growth and development while avoiding nutritional deficiencies in both energy and specific nutrients.

• **Long-term goals** must take into account the avoidance of nutritional risk factors for chronic adult diseases such as hypertension, cancer, and coronary artery disease.

• Each child will need an **individualized program**, depending on the severity of the disorder and his or her personal nature.

• A trained dietician should be consulted in order to plan such an individualized diet for the child.
The aim of this lecture is to point out some of the needs and challenges regarding the maintenance of nutrition in a number of conditions in children.

**Conditions**

<table>
<thead>
<tr>
<th>Kidney disease</th>
<th>Cystic fibrosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diabetes Mellitus</td>
<td>Gut disease</td>
</tr>
<tr>
<td>Cancer</td>
<td>Heart disease</td>
</tr>
<tr>
<td>Liver disease</td>
<td></td>
</tr>
</tbody>
</table>
Kidney Disease

Chronic renal disease (CRD)
in children

Chronic renal disease involves a progressive decline in the number of functioning nephrons that may ultimately result in kidney failure.
Functions of the kidney

Excretory organ that filters, reabsorbs and excretes:
Water
Electrolytes
Glucose
Amino acids
Protein by-products
Toxic substances

Non-urinary functions of the kidney

Glomerular filtration rate (GFR) as measure of kidney function

GFR is most accurately estimated using formulas specifically developed for children. 
Counahan-Baratt formula \[ \text{GFR (mL/min/1.73m}^2\) = 0.43 \times \text{Length (cm)/serum creatinine (mg/mL)} \]

Biochemical abnormalities in kidney damage

• Proteinuria → hypo-albuminaemia and oedema
  
  • Elevated serum creatinine and blood urea nitrogen (BUN) → uraemic symptoms of nausea, vomiting, poor appetite and general malaise → adversely affect dietary intake
• ± 50% of children 5-17 years on dialysis exhibit metabolic acidosis

• Serum bicarbonate levels < 22 mmol/L should be corrected with oral administration of alkali therapy and/or a sodium bicarbonate additive in the hemodialysis dialysate solution.

• As creatinine clearance decrease below 80 mL/min plasma calcitriol starts to decrease and plasma PTH starts to increase (secondary parathyroidism).
Challenges

• The goals in feeding a child with renal disease are to promote normal growth and protect his/her health as much as possible by slowing down the deterioration of renal function.

• Children are in an active anabolic state, with dietary protein being used both for production of new and maintenance of existing body protein.

• Children with CKD typically have shorter stature and lower BMI than non-CKD children

• Growth and nutrition status are negatively correlated with abnormal acid-base status in children.

• Growth hormone secretion is interrupted, changed, and/or suppressed in multiple metabolic pathways in CKD.

• Glucocorticoid treatment is the main inhibiting factor for longitudinal growth after kidney transplantation.
Dietary management

- The major challenge in children is maintaining sufficient oral intake to promote normal growth.

- Nocturnal nutritional supplementation by an appropriate individualised chronic feeding tube route could also be used.

- Although a sufficient energy intake is recommended for children with CRF, excessive kJ intake may induce hyperlipidemia, hyperinsulism and arteriosclerosis in the long run.

---

**Dietary management of chronic renal failure**

**Table 2**

<table>
<thead>
<tr>
<th>Nutrition and diet</th>
<th>Water and electrolyte management</th>
</tr>
</thead>
<tbody>
<tr>
<td>Energy intake</td>
<td>Balance intake and output</td>
</tr>
<tr>
<td>Protein intake</td>
<td>Restrict in the presence of severe hypertension</td>
</tr>
<tr>
<td>Vitamins/tonics</td>
<td>In face of hyperkalaemia, restrict intake, Kayedalate®</td>
</tr>
<tr>
<td></td>
<td>1 g/kg/dose, oral bicarbonate</td>
</tr>
<tr>
<td>High energy (carbohydrates, fats)</td>
<td>Balance intake and output</td>
</tr>
<tr>
<td>Moderate protein restriction</td>
<td>Restrict in the presence of severe hypertension</td>
</tr>
<tr>
<td>depending on blood urea</td>
<td>In face of hyperkalaemia, restrict intake, Kayedalate®</td>
</tr>
<tr>
<td>Use high biological value proteins 2.5 g/kg/d</td>
<td>1 g/kg/dose, oral bicarbonate</td>
</tr>
<tr>
<td>Water soluble vitamins routinely</td>
<td>High energy (carbohydrates, fats)</td>
</tr>
<tr>
<td>Fat-soluble vitamins not necessary; iron and zinc for maintenance</td>
<td>Moderate protein restriction depending on blood urea</td>
</tr>
</tbody>
</table>

Coovadia HM, Wittenberg DF. Paediatrics & child health. 5th ed. p547
Protein and energy intake

• Aim at high energy intake with a protein-restricted diet (of high biological value) if the blood urea is raised. Sweets and high-energy formulas can be introduced.

• Children receiving peritoneal dialysis absorb significant amounts of calories from the dialysate glucose-based solution which should be taken into account.

• Protein intake should be higher for peritoneal dialysis patients due to constant loss of amino acids through the peritoneal membrane carrier.

Recommended dietary protein and energy for children on initiation of maintenance dialysis

Table 3

<table>
<thead>
<tr>
<th>Age (y)</th>
<th>Recommended Dietary Allowances for Protein (g/kg/day)</th>
<th>Protein Intake for Hemodialysis (g/kg/day)</th>
<th>Protein Intake for Peritoneal Dialysis (g/kg/day)</th>
<th>Energy Intake (kcal/day)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infants 6-6.5</td>
<td>2.2</td>
<td>2.6</td>
<td>2.0-3.0</td>
<td>108</td>
</tr>
<tr>
<td>Infants 6.5-10</td>
<td>1.6</td>
<td>2.0</td>
<td>2.3-2.4</td>
<td>98</td>
</tr>
<tr>
<td>Children 4-6</td>
<td>1.2</td>
<td>1.6</td>
<td>1.8-2.0</td>
<td>102</td>
</tr>
<tr>
<td>Children 7-10</td>
<td>1.2</td>
<td>1.4</td>
<td>1.7-1.8</td>
<td>70</td>
</tr>
<tr>
<td>Males 11-14</td>
<td>1.0</td>
<td>1.4</td>
<td>1.7-1.8</td>
<td>55-59</td>
</tr>
<tr>
<td>Males 15-18</td>
<td>0.9</td>
<td>1.3</td>
<td>1.4-1.5</td>
<td>45-49</td>
</tr>
<tr>
<td>Females 14-17</td>
<td>0.8</td>
<td>1.2</td>
<td>1.7-1.8</td>
<td>40-49</td>
</tr>
<tr>
<td>Females 18-21</td>
<td>0.8</td>
<td>1.2</td>
<td>1.4-1.5</td>
<td>40</td>
</tr>
</tbody>
</table>

Modified from National Kidney Foundation Kidney Disease Outcomes Quality Initiative Clinical Practice Guidelines for Nutrition in Chronic Renal Failure: Pediatric Guidelines. 1

Vitamins and Minerals

- Normal intakes for pyridoxine, vitamin B\textsubscript{12}, folic acid, zinc, vitamins A, C, E, and K are recommended.

- Calcium supplementation, limited phosphate intake and low dose calcitriol supplementation should be considered to prevent the onset of secondary hyperparathyroidism.

- Anaemia is relatively common in stages 1-4 of renal failure. Recombinant EPO is often required.

- Oral iron supplementation needs to be individually assessed and is often given intravenously in conjunction with dialysis therapy.

Diabetes Mellitus
Diabetes Mellitus

Is a group of metabolic diseases characterized by hyperglycaemia resulting from defects in insulin secretion, insulin action, or both.

Apart from hyperglycaemia, lack of diabetes control could be associated with a variety of serious complications, including retinopathy, nephropathy, neuropathy, and cardiovascular disease.

Table 4

<table>
<thead>
<tr>
<th></th>
<th>Type 1</th>
<th>Type 2</th>
</tr>
</thead>
<tbody>
<tr>
<td>Prevalence in diabetic population</td>
<td>≥10% of cases; ≤30 years; autoimmune diseases, viral infections, inherited factors</td>
<td>≥90-95% of cases; ≥49 years; obesity, aging, inherited factors</td>
</tr>
<tr>
<td>Age of onset</td>
<td>Destruction of pancreatic beta cells; insulin deficiency</td>
<td>Insulin resistance; insulin deficiency (relative to needs)</td>
</tr>
<tr>
<td>Associated conditions</td>
<td>Little or none</td>
<td>Varies: may be normal, increased, or decreased</td>
</tr>
<tr>
<td>Major defect</td>
<td>Always</td>
<td>Sometimes</td>
</tr>
<tr>
<td>Insulin secretion</td>
<td>Juvenile-onset diabetes; insulin-dependent diabetes mellitus (IDDM)</td>
<td>Non-insulin-dependent diabetes mellitus (NIDDM)</td>
</tr>
<tr>
<td>Requirement for insulin therapy</td>
<td>Order names</td>
<td></td>
</tr>
</tbody>
</table>

Whitney E, Rolfes SR. Understanding nutrition. 11th ed. p638
Challenges

Nutrition therapy for type 1 diabetes (T1DM) should focus on

- maintaining optimal nutrition status
- controlling blood glucose
- achieving a desirable blood lipid profile
- controlling blood pressure
- preventing and treating complications of diabetes
Dietary recommendations for Diabetes Mellitus

- The diet should provide a fairly consistent carbohydrate intake from day to day and at each meal and snack to minimize fluctuations in blood glucose.

- Carbohydrate counting (interchanging foods of equal carbohydrate content) is advised as a means of matching injected insulin with carbohydrate intake and improving glycaemic control in T1DM.

- This approach is more flexible than the traditional method of prescribing ‘carbohydrate portions’ since it permits altering insulin dose on the basis of the quantity of carbohydrate consumed.

Balancing injected insulin with carbohydrate-containing food


† Injection of insulin mixtures containing soluble (=) and longer lasting insulin (III)
Physical Activity

- Children and adolescents with T1DM should take part in a minimum of 30-60 minutes of moderate physical activity daily.

- To avoid hypoglycaemia, blood glucose should be monitored before and after activity to determine when changes in insulin or food intake are needed.

- Carbohydrate-rich foods should be readily available during and after activity.

Key aspects of the current recommendations for diabetic nutrition and lifestyle

Table 5
Dietary energy and body weight
Achieve and/or maintain BMI of 18.5–25
Diet and exercise important

Dietary fat
Saturated plus trans-unsaturated fatty acids: < 10% total energy,
< 8% if low-density lipoprotein raised
Polyunsaturated fatty acids: < 10% total energy
Monounsaturated fatty acids: 10–20% total energy
Total fat: < 35% total energy (if overweight < 30%)
Oily fish, soybean and rapeseed oil, nuts and green leafy vegetables to provide ω-3 fatty acids
Cholesterol: < 300 mg/day
Carbohydrate

Total carbohydrate: 45–60% total energy, influenced by metabolic characteristics
Vegetables, fruits, legumes and cereal-derived foods preferred

Dietary fiber and glycemic index

Naturally occurring foods rich in dietary fibre are encouraged
Ideally dietary fibre intake should be more than 40 g/day (or 20 g/1000 kcal/day), half soluble (lesser amounts also beneficial)
Five servings/day of fibre-rich vegetables and fruit and four or more servings of legumes/week help to provide minimum requirements
Cereal-based foods should be wholegrain and high in fibre
Carbohydrate-rich low-glycaemic-index foods are suitable choices, provided other attributes are appropriate

Protein and renal disease

Total protein intake at lower end of normal range (0.8 g/kg/day) for type 1 patients with established nephropathy
For all others, protein should provide 10–20% total energy

Vitamins, antioxidants, minerals and trace elements

Increase foods rich in tocopherols, carotenoids, vitamin C and flavonoids, trace elements and other vitamins
Fruits, vegetables, wholegrains rather than supplements recommended
Restrict sodium to less than 6 g/day

Families

Most recommendations suitable for whole family

Cancer

- Cancer, also known as a malignant neoplasm, is a broad group of various diseases, all involving unregulated cell growth.

- In cancer, cells divide and grow uncontrollably, forming malignant tumours, and invade nearby parts of the body. The cancer may also spread to more distant parts of the body through the lymphatic system or bloodstream.

- Treatment of cancer depends on the type of tumour, the site of the primary tumour, and the extent of spread. Surgery, chemotherapy, or radiation could be applied.
The most common paediatric cancers are acute leukemias and brain tumours.

**Goals for nutritional support:**

- Reverse nutritional deficits
- Prevent further deficits
- Allow for normal growth and development
- Minimize morbidity
- Maximise the quality of life
Advantages of good nutrition

Children who get good nutrition while they are being treated for cancer:

- Tolerate treatment and treatment side effect better
- Are able to stay on schedule for treatment
- Heal and recover faster
- Have less risk of infection during treatment
- Are better able to keep up normal growth and development
- Feel better and have a better quality of life

Challenges

- Children being treated for cancer need extra kJ for energy, growth, to fight infection, rebuild damaged tissues, etc. Compared to healthy children, 20-50% more kJ may be required.

- Cancer and its treatments may affect a child’s appetite, tolerance to foods, and their ability to use nutrients. Common treatment side effects, like nausea and vomiting and mouth sores can make it hard to eat well.

- Children with cancer often present with protein-energy malnutrition (PEM) which needs to be corrected.

Useful website with practical advise:
Methods for nutritional support

Oral feeding/supplements

- If at all possible, the child should get the required nutrients from eating and drinking nutrient-rich foods and fluids that are part of a healthy, well-balanced diet.

- After a child has had surgery, chemotherapy, or radiation treatments, he may need extra protein to heal tissues and to help prevent infection.

- Extra nutrients should be added in the form of high-energy, high-protein meals supplemented with snacks, and homemade drinks and shakes.

Enteral feeds

- If a child cannot eat or drink or it gets too hard for the child to maintain or gain weight by normal intake, or if his kJ and nutrients needs have greatly increased, a feeding tube may be indicated.

- Most of the time, these liquid feedings can supply the child with all of the calories, protein, vitamins, and minerals needed.

- Children who have feeding tubes usually can still eat by mouth. The child may be tube fed at night while sleeping to allow him to eat during the day.
**Parenteral nutrition (PN)**

- Sometimes tube feedings are not able to give all the fluids and nutrients a child needs. In these cases, nutrient solutions can be given right into a vein. PN solutions can usually supply all of a child’s nutritional needs.

- PN is most often used when the stomach and intestines are not working properly. For example, in children who have:
  - Had surgery involving the digestive system
  - Complete blockage of the intestines
  - Uncontrolled nausea, vomiting, or diarrhea
  - An infection that requires the digestive system to rest so that it can heal

---

**When the child is taking steroids**

- Children with cancer often take steroids, such as prednisone or dexamethasone, as part of their treatment.

- Children taking steroids usually feel hungry all the time and may gain weight.

- Steroids also tend to make people retain fluid.

- Some dietary changes to help prevent fluid retention and limit weight gain may be needed. Salt intake should be limited. The appetite changes and fluid retention caused by steroids are short-term and will go away when treatment ends.
Liver Disease

Functions of the liver

Fig 7

Silverthorn DG. Human Physiology. Pearson, San Francisco. 5th ed. 2010. p 698
Liver disease

- The liver is susceptible to a wide variety of diseases in childhood.

- Jaundice and hepatomegaly are most commonly due to a viral infection of the liver.

- Pediatric chronic liver disease could eventually lead to liver insufficiency, liver cirrhosis, and may be associated with profound cholestasis.

Cholestasis may be associated with malabsorption, malnutrition, and growth retardation.

Fig 8
**Nutritional defects due to cholestasis**

- Metabolic alkalosis is regularly observed in cholestatic children.
- Urea synthesis is a highly bicarbonate consuming system.
- Impairment of urea synthesis may lead to excessive bicarbonate levels and alkalosis.
- Zinc deficiency is common in cholestasis. Zinc is a common cofactor of various enzymatic systems, and deficiency may affect liver metabolism such as for example ammonium metabolism and the urea cycle.

- Cholestasis could interfere with fat absorption due to low levels of bile acids being secreted into the duodenum.
- Medium chain triglycerides could be added to the diet as they are directly absorbed by the intestinal mucosa, without requiring emulsification by bile acids.
- Due to a derangement in the digestion and absorption of fats, levels of the fat-soluble vitamins A,D,E,K could also be affected.
- Calcium deficiency and bone demineralization are common in these patients, due to calcium malabsorption.
- Calcium absorption will be improved by vitamin D supplementation in deficient patients.
### Vitamin and mineral support for cholestatic infants

<table>
<thead>
<tr>
<th>Table 6</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Vitamin A</td>
<td>25,000 to 50,000 IU, IM, every other month</td>
</tr>
<tr>
<td>Vitamin D</td>
<td>30,000 to 60,000 IU, IM, every other month</td>
</tr>
<tr>
<td>Vitamin E</td>
<td>10 mg/kg, intra-muscular, every two weeks</td>
</tr>
<tr>
<td>Vitamin K</td>
<td>1 mg/kg, maximum 10 mg, intra-muscular, once weekly to 2 weeks</td>
</tr>
<tr>
<td>Calcium</td>
<td>50 mg/kg/day, orally</td>
</tr>
<tr>
<td>Zinc</td>
<td>1 mg/kg/day, zinc sulfate, orally</td>
</tr>
</tbody>
</table>


### Cystic fibrosis
Cystic fibrosis

- Cystic fibrosis (CF) is a hereditary disease affecting cells of the exocrine glands.
- Affected individuals lack a protein, cystic fibrosis transmembrane regulator (CFTR), that enables the transport of Cl⁻ across cell membranes.
- The disease is characterised by impaired epithelial transport of electrolytes and fluid, producing thick, viscous secretions in many organs.

Organs affected by cystic fibrosis

The digestive system in cystic fibrosis

- In cystic fibrosis, mucus can block pancreatic ducts preventing enzymes from reaching the intestines.
- As a result, the intestines can't fully absorb fats and proteins. This can cause ongoing diarrhea or steatorrhea.
- Intestinal blockages also may occur, especially in newborns.
- Too much gas or severe constipation in the intestines may cause stomach pain, discomfort and lack of appetite.
- A hallmark of CF in children is poor weight gain and growth.

Nutritional management

- The overall aim of nutritional therapy is to ensure that patients maintain normal linear growth and weight gain.
- A well-balanced diet rich in calories, fat, and protein is recommended. The diet should provide a calorie intake of 120-140% of the Recommended Daily Allowance for calories.
- Supplemental nocturnal tube feeding should be considered.
- Oral pancreatic enzymes to help digest fats and proteins and absorb more vitamins should be prescribed.
Recommendations for vitamin supplementation

Table 7

<table>
<thead>
<tr>
<th>Age</th>
<th>Vitamin A (IU)</th>
<th>Vitamin E (IU)</th>
<th>Vitamin D (IU)</th>
<th>Vitamin K (mg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0–12 months</td>
<td>1500</td>
<td>40–50</td>
<td>400</td>
<td></td>
</tr>
<tr>
<td>1–3 years</td>
<td>5000</td>
<td>80–150</td>
<td>400–800</td>
<td>0.3–0.5</td>
</tr>
<tr>
<td>4–8 years</td>
<td>5000–10,000</td>
<td>100–200</td>
<td>400–800</td>
<td></td>
</tr>
<tr>
<td>&gt;8 years</td>
<td>10,000</td>
<td>200–400</td>
<td>400–800</td>
<td></td>
</tr>
</tbody>
</table>

In addition to a standard age-appropriate dose of non-fat-soluble multivitamins, the following should be given:

Bone health in cystic fibrosis

- Malabsorption, reduced weight-bearing exercise and corticosteroid use can result in premature osteopenia and osteoporosis.

- Prevention and treatment of osteopenia in children with CF should focus on
  - sufficient vitamin D and K intake to normalize blood levels
  - calcium intake of 1300 mg/day for children ages 9-18
  - increasing weight-bearing activity

Gastrointestinal tract diseases

• For a child to grow and develop normally, food must be ingested, digested, and absorbed.

• The gastrointestinal (GI) tract is crucial in this process.

• Inflammatory bowel disease such as Crohn’s disease and ulcerative colitis could interfere with digestion.
Inflammatory bowel disease

Fig 10

- Chronic disease could cause partial obstruction of the intestine, leading to pain, diarrhoea, and malabsorption.
- Children are prone to develop electrolyte, mineral and vitamin deficiencies due to inadequate intake, malabsorption or excessive losses.
- Children should be assessed for nutrient deficiencies.
Dietary treatment

- Patients are at risk for iron deficiency due to chronic bleeding. Iron and folic acid supplementation could be required.

- Fish oil could help in the treatment of ulcerative colitis and Crohn’s disease as the increased ω-3 fatty acids decrease the production of pro-inflammatory cytokines.

- Lactose should not be restricted unless there is evidence of intolerance. If restricted, adequate calcium and vitamin D should be provided.

- Every attempt should be made to maximize the nutritional intake.

- If necessary, supplementation should be considered.

- The most effective way is by means of nocturnal (10-12 hours) nasogastric feedings by which formula is infused at a constant rate using a pump.
Heart Disease

A congenital heart defect (CHD) is a defect in the structure of the heart and great vessels which is present at birth. Many types of heart defects exist, most of which either obstruct blood flow in the heart or vessels near it, or cause blood to flow through the heart in an abnormal pattern.

Congestive heart failure (CHF) is generally defined as the inability of the heart to supply sufficient blood flow to meet the needs of the body.
Heart disease

Congestive heart failure may restrict growth through a variety of mechanisms:

• Nutritional intake may be reduced by tachypnea, dyspnea, and increased fatigability.

• Reduced systemic perfusion, particularly to the GIT may limit gastric emptying, intestinal mobility and nutrient uptake.

Nutritional management

• Higher calorie and moderate protein supplementation, compared to normal age-matched children, is needed for postoperative wound healing and long-term catch-up growth (20-30%> than normal).

• Partial or total parenteral nutrition should be used in acute settings where patients are not able to receive sufficient enteral nutrition.

• Breast-feeding should be encouraged whenever it is medically feasible. Breast-feeding provides highly absorbable nutrients and abundant amounts of immunoglobulin.
• Supplemental higher-calorie formulas may be used in combination with breast-feeding to achieve desired growth.

• Fluid status is one of the major factors affecting the medical and nutritional condition of CHD patients. Insensible fluid loss can be as high as 10-15% above normal due to tachypnea or fever.

• Conversely, fluids may be restricted in patients with CHF to as little as 60-80 ml/kg and diuretics administered to prevent fluid overload.

• Strict fluid limits make adequate nutritional support extremely challenging.

---

**Lipid disorders in children**

• Cardiovascular disease (CVD) is a leading cause of morbidity and premature mortality in women and men.

• Epidemiological studies have established a link between diet, serum cholesterol levels and subsequent CVD risk.

• The process of atherosclerosis evolves over decades, and begins as early as childhood.
American Heart Association 2006
Diet and Lifestyle Goals for Cardiovascular Disease Risk Reduction

- Consume an overall healthy diet
- Aim for a healthy body weight
- Aim for recommended levels of low-density lipoprotein cholesterol, high-density lipoprotein cholesterol, and triglycerides
- Aim for a normal blood pressure
- Aim for a normal blood glucose level
- Be physically active

Cholesterol modification in children

- Dietary intervention is the primary approach for children with elevated cholesterol.

- Nutritional adequacy should be achieved by eating a wide variety of foods with sufficient calories to support normal growth and development and to reach and maintain desirable body weight.

- A nutrient intake pattern in which total fat constitutes no more than 30% of total calories, with <10% of total calories from saturated fat and dietary cholesterol limited to <300 mg/day is recommended.
• The intake of foods high in n-3 fatty acids such as salmon, nuts, canola, etc. should be increased. At least 3 fish servings per week are recommended.

• The American Academy of Pediatrics has cautioned against the introduction of an extremely low-fat diet (<20% of calories) in children. Such diets may result in deficiencies in essential fatty acids, fat-soluble vitamins and other nutrients.

• See table 5

http://circ.ahajournals.org/content/119/8/1161