Clinical Perspective: Nutrition Interventions for Youth with Dyslipidemia

Overview: Dyslipidemia, or high cholesterol, means you have too many lipids (fat) in your blood. Nutrition interventions in youth with dyslipidemia provide short- and long-term benefits without adverse effects on growth or maturation. Counseling by a registered dietitian nutritionist (RDN) is strongly recommended to help youth and their families successfully alter dietary intake to meet nutritional needs, provide ongoing support, and encourage long-term adherence to healthy nutrition and exercise patterns.

Highlights
- Atherosclerosis, the origin of cardiovascular disease (CVD), begins at a very early age and is significantly influenced by lifestyle behaviors.
- Adopted at an early age and maintained over a lifetime, nutrition interventions and other heart-healthy lifestyle behaviors can delay the onset or prevent the occurrence of CVD-related events.
- Even in youth who require lipid lowering medication, lifestyle interventions provide additional benefits and enhance lipid-lowering.
- There are limited data supporting the use of dietary supplements in youth with dyslipidemia.
- Changes in lifestyle habits are best achieved with a multidisciplinary, family-based approach.

“Heart-Healthy Lifestyle”
Beginning at an early age and sustained throughout life, a heart-healthy lifestyle may reduce risk for cardiovascular disease in youth. The goal of this National Lipid Association Clinical Perspective is to provide guidance for healthcare professionals caring for youth with disorders of lipid and lipoprotein metabolism, including nutritional guidance that complements the use of lipid lowering medications.

Therapeutic Objective
A family-centered approach and the support of a multi-disciplinary healthcare team, which includes a RDN to provide nutrition counseling, provides the best opportunity for primary prevention and improved outcomes.

Components of a Heart-Healthy Lifestyle
- Decrease saturated fatty acid intake and replace with unsaturated fatty acids
- Avoid trans fatty acids
- Reduce dietary cholesterol
- Increase dietary soluble fiber
- Incorporate omega-3 fatty acids into the diet
- Choose complex carbohydrates
- 60 minutes moderate-to-vigorous physical activity

Dietary Supplements
A limited number of dietary supplements are included in national guidelines for youth and discussed in this paper. With all patients, it is important to inquire about use of dietary supplements, including herbal products, and provide appropriate guidance. Parents and caregivers should be informed that some dietary supplements may have harmful side effects, particularly if started at a young age and taken for a prolonged period-of-time. Dietary supplements should also be discussed with the medical care team, which ideally includes a pharmacist, to ensure that no drug-drug interactions exist for dietary supplements and any prescribed medications used by patients.

Psychosocial Aspects of Nutrition Counseling in Youth
There are many psychosocial aspects to nutrition counseling to be considered, including individualized approaches for youth with underweight, overweight, or disordered eating patterns. The family’s ability to understand and implement recommendations for lifestyle behavioral changes are affected by modifiable and non-modifiable individual, family, community, and healthcare system-level factors. It is important to be aware of the patient’s psychosocial state when counseling families regarding the need for interventions.
Table 2: Summary of nutrition interventions for youth by disorder

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<th>Lipid Disorder</th>
<th>Nutrition Intervention</th>
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| Familial Hypercholesterolemia (FH) | • FH is a common genetic disorder characterized by moderate-to-severe elevations of LDL-C that increase the likelihood of premature CHD.  
  • While LLM is generally required to reduce LDL-C and non-HDL-C, nutrition interventions to reduce SFA to <7% daily caloric intake, avoidance of TFA, <200 mg/day of dietary cholesterol, and increased dietary soluble fiber provides additional benefits. Supplements, such as soluble fiber or plant sterols, may also be beneficial.  
  • All diet modifications should exist in the context of an age-appropriate dietary pattern, with adequate calorie and macro- and micronutrient intake to promote normal growth and development. |
| Familial Combined Hyperlipidemia (FCHL) | • FCHL is a common metabolic disorder characterized by: (a) elevated levels of Apo B that may present as either mixed hyperlipidemia, isolated hypercholesterolemia, HTG, or as normal serum lipid levels with an elevated level of Apo B; (b) intra-individual and intrafamilial variability of the lipid phenotype; a (c) increased risk of premature CHD; and (d) a polygenic inheritance.  
  • Nutrition interventions are similar to those for FH, with additional recommendations for TG lowering, such as reduction in simple carbohydrates foods and sugar sweetened beverages, as needed. |
| Elevated Lipoprotein(a) [Lp(a)] | • Serum Lp(a) reaches adult levels by school age and remains relatively constant into adulthood.  
  • Elevated Lp(a) is recognized as a causal, independent risk factor for premature CVD.  
  • While nutrition interventions do not significantly lower Lp(a), a lifelong heart-healthy lifestyle is helpful in minimizing additional CVD risk factors. |
| Sitosterolemia | • Hyperabsorption and decreased biliary excretion of cholesterol and non-cholesterol sterols leads to accumulation of serum sterols, such as campesterol and sitosterol.  
  • Effective nutrition intervention includes:  
    • Dietary restriction of cholesterol and plant-based non-cholesterol sterols.  
    • Limiting intake or avoidance of shellfish (e.g., clams, scallops, oysters) and plant foods that are high in fat (e.g., vegetable oils, olives, margarine, nuts, seeds, avocados, and chocolate).  
    • Fruits, vegetables, and refined cereal products (not whole grain) may be used.  
    • Margarines/spreads and other sterol- or stanol-fortified products are contraindicated. |
| Cerebrotendinous Xanthomatosis (CTX) | •CTX, characterized by impaired bile acid synthesis, leads to accumulation of cholestanol and cholesterol in many tissues, including the brain.  
  • The treatment of choice for CTX is oral CDCA therapy, although it is currently not approved by the FDA for this indication.  
  • A dietary pattern low in cholestanol (egg yolks, meat, fish/shell fish and poultry, and high fat dairy), especially when implemented at a young age, may also be helpful. |
| Lysosomal Acid Lipase Deficiency (LAL-D) | • LAL-D is a rare autosomal recessive disease, the manifestations of which include a clinical continuum from infancy through adulthood.  
  • The infantile form generally presents with severe failure to thrive, may require a low-fat, amino acid-based formula and, in the absence of timely enzyme replacement, is most often fatal.  
  • Patients with childhood/adult-onset LAL-D may benefit from a dietary pattern with <25-30% daily caloric intake from fat and <200 mg dietary cholesterol daily. Fat-soluble vitamin supplementation may also be helpful in those who have malabsorption and malnutrition.  
  • Enzyme replacement therapy with sebelipase alfa is recommended for the treatment of LAL-D.  
  • Nutrition intervention is an important supportive measure to medical intervention and not a primary therapy to promote changes in lipid levels. |
### Hypobetalipoproteinemias

**Abetalipoproteinemia (ABL)** - Homozygous ABL
- A rare, inherited, autosomal-recessive disorder resulting from a microsomal triglyceride transfer protein deficiency characterized by the absence, or near absence, of LDL-C.
- Disruption of cellular fat transport causes symptoms of fat malabsorption (steatorrhea, diarrhea) and failure to thrive, which often present in infancy or early childhood.
- Dietary fat, cholesterol, and fat-soluble vitamins, such as A, E, D, and K, are poorly absorbed, leading to deficiencies.
- A low-fat diet (20-30% daily caloric intake), adequate intake of EFAs (2-4% daily caloric intake) with supplementation as needed, and vitamin supplementation, are critical in nutritional management. These interventions are most effective when started at a young age.
- Prognosis is variable, but early diagnosis and strict adherence to treatment can improve neurological function and halt disease progression.

**Heterozygous ABL**

**Patients with heterozygous ABL usually have normal lipids**

**Hypobetalipoproteinemia (HBL)** - Homozygous HBL
- A rare, inherited, autosomal co-dominant disorder resulting from mutations in both alleles of the APOB, characterized by the absence, or near absence, of LDL-C.
- Disruption of cellular fat transport causes symptoms of fat malabsorption (steatorrhea, diarrhea) and failure to thrive, which often present in infancy or early childhood.
- Dietary fat, cholesterol, and fat-soluble vitamins such as A, E, D, and K are poorly absorbed, leading to dietary deficiency.
- A low-fat diet (20-30% daily caloric intake), adequate intake of EFAs (2-4% daily caloric intake) with supplementation as needed, and vitamin supplementation, are critical in management. These interventions are most effective when started at a young age.
- Prognosis is variable, but early diagnosis and strict adherence to treatment can improve neurological function and halt disease progression.

**Heterozygous HBL**

**Patients with heterozygous HBL typically have half-normal levels of Apo B-containing lipoproteins. Some may be at-risk of steatohepatitis.**

### Cholesterol Disorders

**Familial Chylomicronemia Syndrome (FCS) and Multifactorial Chylomicronemia Syndrome (MCS)**
- Individuals with FCS have impaired or absent LPL activity caused by a monogenic variant; MCS, which is 50-100 times more common, occurs in individuals with co-existence of genetic and secondary causes.
- Both FCS and MCS lead to severe elevations in TG (>1000 mg/dL).
- The mainstay of treatment is a specialized dietary pattern:
  - Very-low-fat (<15-20 g per day (<10%-15% of total daily caloric intake) while meeting EFA needs (2-4% daily caloric intake).
  - MCT oil to increase overall caloric intake and balance macronutrients in the dietary pattern, as needed.
  - Emphasis on complex carbohydrate foods (e.g., oatmeal, brown rice, quinoa, beans) while limiting simple and refined carbohydrate foods.
  - Avoidance of alcohol.
  - Fat-soluble vitamin and mineral supplementation, as needed.

### Triglyceride Disorders

**Familial Hypertriglyceridemia (FHTG)**
- FHTG may be present in youth, typically in those with overweight or obesity and/or insulin resistance.
- A low-fat diet (<30% calories from fat), limited intake of foods and beverages with added sugars, and the addition of complex carbohydrate foods and dietary sources of O3FAs is helpful in lowering TGs.
- Promotion of a healthy weight is especially helpful in youth with overweight or obesity and/or insulin resistance.

**Acquired Hypertriglyceridemia (HTG)**
- Acquired HTG is closely associated with adverse lifestyle behaviors.
- Nutrition intervention with lifestyle behaviors designed to achieve and maintain healthy body weight are the primary treatments for acquired HTG.
- For youth with mild- to moderate-HTG, emphasis on vegetables, fruits, and lean protein intake with reduced intake of refined carbohydrate foods and foods/beverages with added sugar can be effective.
- For youth with severe HTG (fasting TGs >1000mg/dL), fat intake should be decreased to <10-15% of total daily caloric intake (<15-20 g per day) with avoidance of simple carbohydrates, such as foods and beverages with added sugars and 100% fruit juice.

**Abbreviations:**
- FH=familial hypercholesterolemia, LDL-C=low density lipoprotein cholesterol, CHD=coronary heart disease, LLM=lipid-lowering medication, non-HDL-C= non high density lipoprotein cholesterol, SFA=saturated fatty acids, TFA=trans fatty acids, FCHL=familial combined hyperlipidemia, Apo B=apolipoprotein B, HTG=hypertriglyceridemia, CVD=cardiovascular disease, TG=triglycerides; Lp(a)=lipoprotein(a), CTX=cerebrotendinous xanthomatosis, CDCA=chenodeoxycholic acid, FDA=Food and Drug Administration, LAL-D=lysosomal acid lipase deficiency, ABL=abetalipoproteinemia, EFA=essential fatty acids, HBL=hypobetalipoproteinemia, FCS=familial chylomicronemia syndrome, LPL=lipoprotein lipase, MCS=multifactorial chylomicronemia syndrome, MCT=medium chain triglyceride, HTG=familial hypertriglyceridemia, O3FAs=omega-3 fatty acids

Read the National Lipid Association’s Clinical Perspective in the *Journal of Clinical Lipidology*.  
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