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SUMMER SCHOOL: PEDIATRIC HEPATOLOGY

Complications of chronic liver disease

Alexandra Tsouka, Valérie A. McLin*

Department of Pediatrics, University Hospitals Geneva, 5, rue Willy-Donze, 1205 Geneva, Switzerland

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Summary Children with chronic liver disease (CLD) need a head to toe approach and an early suspicion of multi organ involvement. Nutritional assessment and management is the cornerstone of management. Consider immune dysfunction in everyday treatment decisions. Consider early heart-lung-brain involvement in transplant evaluation.

Take home messages

1. Children with CLD need a head to toe approach and an early suspicion of multi organ involvement.
2. Nutritional assessment and management is the cornerstone of management.
3. Consider immune dysfunction in everyday treatment decisions.
4. Consider early heart-lung-brain involvement in transplant evaluation

Introduction

Managing a child with chronic liver disease (CLD) involves much more than dealing organ specific deficiencies (Fig. 1). This is particularly true in children who warrant meticulous care owing to their ongoing development.

The aim of this review is to focus on nutrition, bone metabolism, renal function, immunity, central nervous system disturbances and cardiopulmonary complications of CLD. Portal hypertension (PH) is the culprit of many symptoms related to CLD and is therefore also briefly discussed. The focus of the next pages is not clinical management, for which we refer the reader to several excellent and current reviews. Rather, we aim to focus on the often unclear pathophysiology of CLD.

Nutrition

Nutritional status is considered one of the most important prognostic factors in CLD. Malnutrition affects pre- and post-transplant survival, and nutritional assessment should be a priority in children with liver disease. It should be assumed that all children with CLD present with substrate and nutrient deficiencies for two reasons: impaired absorption or intake, and impaired hepatic homeostasis. Basic energy requirements are increased as detailed below and disordered substrate use may further complicate nutritional demands [1].

There are several factors leading to malnutrition in CLD, and these are outlined in Table 1.

There are two categories of patients with CLD: those with anicteric cirrhosis and PH, and those with cholestatic

* Corresponding author. Tel.: +4122 372 4572; fax: +4122 372 5489.
E-mail address: valerie.mclin@hcuge.ch (V.A. McLin).

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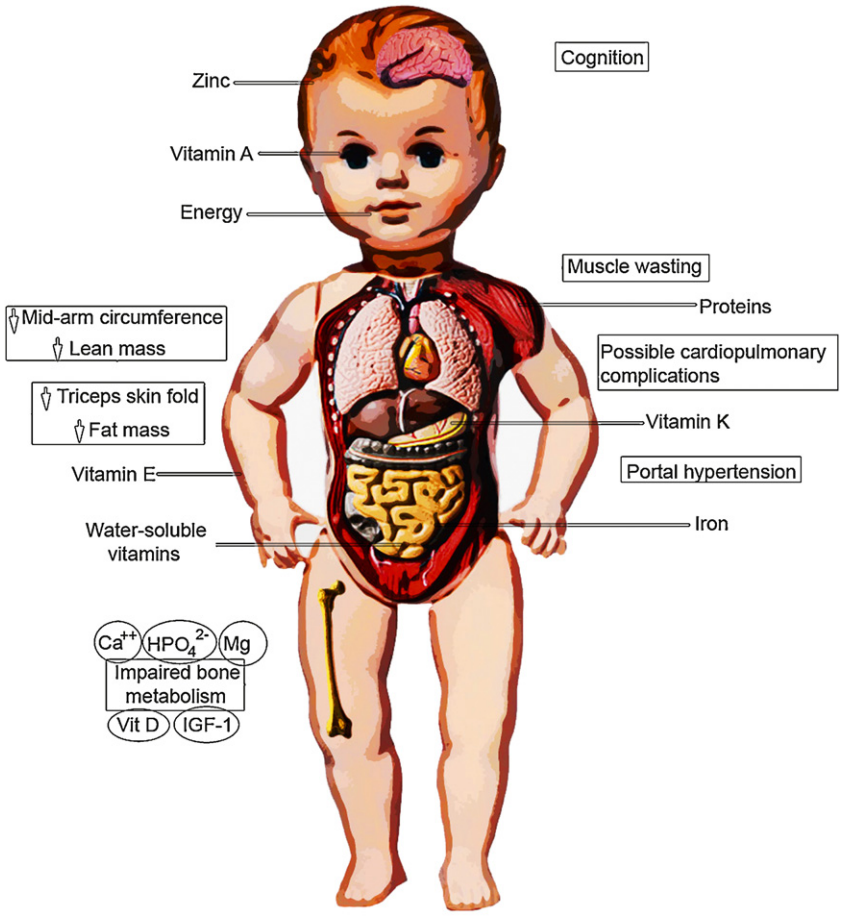

Figure 1 Head to toe assessment of the child with chronic liver disease.

cirrhosis. The latter group warrants particular attention as their poor nutritional status is further exacerbated by defective bile acid circulation.

Energy

Various factors contribute to increased energy requirements, as described in Table 2.

Proteins

Several factors lead to protein deficiency in CLD (Table 1). Studies in adults suggest that an overnight fast in cirrhotic patients is the equivalent of a 3-day fast in healthy individuals. In other words, the switch to amino-acid-derived gluconeogenesis occurs earlier in the cirrhotic patient.

Protein restriction in order to avoid ammonia overload is not recommended for children. The incidence or severity of encephalopathy appears to be independent of protein load in children who have high protein needs for growth.

Reduced Abnormal Aromatic Amino Acid (AAA) oxidation by the liver and increased Branched-Chain Amino Acid (BCAA) metabolism in extra hepatic tissues (AAA) to (BCAA) ratio in CLD results in an altered AAA: BCAA ratio in CLD. Formulas enriched in BCAA reduce the catabolic state, keep skeletal muscle intact and reduce ammonia production. Nocturnal BCAA administration may stimulate hepatic albumin synthesis. Moreover, as BCAA competes with tryptophan for the same amino acid transporter in the blood-brain barrier, supplementation of BCAA could also be beneficial in preventing hepatic encephalopathy (HE) [1].

Carbohydrates

Reduced glycogen storage and gluconeogenesis capacity exposes infants and young children to a higher risk of fasting hypoglycemia. Older children rely on fat and protein as alternative energy sources as previously mentioned [1].

Fats

As previously mentioned, children with cholestasis are exceptionally vulnerable to protein/energy deficiencies. Cholestatic liver disease leads to a decreased bile salt pool. In children with biliary atresia and blind small bowel loops following portoenterostomies, the pool is further decreased by bile salt deconjugation from bacterial overgrowth. This deficit in enteral bile salts results in fat malabsorption, impairing overall energy balance in children with CLD, as children largely rely on fat to cover their total energy needs.

In advanced liver disease, insulin resistance leads to diminished peripheral delivery of glucose and impaired