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《2002版美国肠外与肠内营养在成人和儿童患者中的应用指南》重要信息提炼

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内容大纲

❶ 指南简介

❷ 指南内容框架

❸ 指南中对左卡尼汀的推荐-摘录及翻译

指南简介

JOURNAL OF PARENTERAL AND ENTERAL NUTRITION

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Guidelines for the Use of Parenteral and Enteral Nutrition in Adult and Pediatric Patients

中文全称：肠外与肠内营养在成人和儿童患者中的应用指南

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指南内容框架

第1部分：引言
第2部分：营养护理过程
第3部分：营养评估-成人
第4部分：营养评估-儿童
第5部分：特殊营养支持的管理
第6部分：正常的需求-成人
第7部分：正常的需求-儿童
第8部分：营养支持实施的过程
第9部分：药物-营养相互作用
第10部分：生命周期和代谢状况
第11部分：针对特定疾病的指南-成人
第12部分：特殊营养支持的管理—专门针对儿科的问题
第13部分：针对特定疾病的指南-儿童

各部分主要由**知识背景、循证医学证据和实践指南**等内容构成，其中，*红色字体标示的部分含有描述或推荐可益能（左卡尼汀）的内容*。

指南中对左卡尼汀的推荐-摘录及翻译

Section VII: Normal Requirements-Pediatrics 第7部分：正常的需求-儿童

MICRONUTRIENT REQUIREMENTS 微量营养素的需求（见该指南第31页）

原文摘录

Carnitine, synthesized from methionine and protein bound lysine, is the requisite transporter of long chain fatty acids across the inner mitochondrial membrane for β-oxidation. Carnitine can be obtained through the diet, exogenous supplementation, and endogenous biosynthesis. Now a standard addition to many enteral formulas, carnitine may become conditionally essential in the neonate on long term PN secondary to limited biosynthetic capability and immature conservation mechanisms.[3] Carnitine supplementation should be provided once a deficiency has been confirmed.

中文翻译

左卡尼汀，由蛋氨酸和赖氨酸合成，是长链脂肪酸通过线粒体内膜参与β氧化的必需转运因子。体内左卡尼汀可通过饮食，外源性补充和内源性合成三种方式获得。**现在已是许多肠内营养配方的标准添加剂**，由于生物合成能力受限和不成熟的保护机制，**在长期肠外营养的新生儿中左卡尼汀成为条件性的必需营养素。一旦确诊卡尼汀缺乏应立即提供左卡尼汀补充。**

Section XII: Administration of Specialized Nutrition Support-Issues Unique to Pediatrics 第12部分：特殊营养支持的管理—专门针对儿科的问题

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COMPLICATIONS UNIQUE TO NEONATES:HYPERTRIGLYCERIDEMIA

针对新生儿并发症：**高甘油三酯血症**（见该指南第106页）

原文摘录

Premature infants have low carnitine reserves. Low carnitine levels can also be seen in full-term neonates receiving long-term PN.[20,21] Carnitine supplementation may increase lipid clearance,[22,23] but low plasma carnitine concentrations do not necessarily correlate with elevated serum triglyceride concentrations.[24] It has been shown that fatty acid oxidation and improved triglyceride levels are seen in premature infants receiving supplemental intravenous carnitine who are also receiving lipid emulsions.[25] Although studies are somewhat inconclusive, the administration of L-carnitine (at a dose of 10 mg/kg per day) seems to enhance fatty acid oxidation, especially in carnitine-deficient infants. L-carnitine may be useful in infants with hypertriglyceridemia when other etiologies have been ruled out. Guidelines on monitoring for hypertriglyceridemia are predominately empiric.

中文翻译

早产儿体内左卡尼汀储备量少。长期接受PN治疗的足月新生儿也同样存在体内卡尼汀水平不足[20,21]。补充卡尼汀可以增加脂质清除[22,23]，但是低血浆卡尼汀浓度并不一定与血清甘油三酯浓度升高有关[24]。已被证实，**在补充脂肪乳的早产儿中，同时静脉补充左卡尼汀可以改善脂肪酸氧化和甘油三酯水平**[25]。虽然研究有一定的争议，但**补充左卡尼汀（按每天10 mg/kg的剂量）**似乎确实能够提高脂肪酸氧化，**尤其是对那些存在卡尼汀缺乏的患儿**。对于高甘油三酯患儿而言，当排除其他病因时，左卡尼汀补充可能是有用的。对高甘油三酯血症的监测的临床指引是主要的治疗经验。

Section XII: Administration of Specialized Nutrition Support-Issues Unique to Pediatrics 第12部分：特殊营养支持的管理—专门针对儿科的问题

Practice Guidelines Complications Unique to Neonates: Hyperglycemia

针对新生儿并发症的临床指南：**高甘油三酯血症**（见该指南第106页）

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原文摘录

1. Lipid emulsion infusions in infants should begin at 0.5 to 1 g/kg per day and advance at rate of 0.5 g/kg per day to a maximum of 3 g/kg per day. (A)
2. Lipid emulsion infusion rates should be reduced in premature or septic infants and serum triglyceride concentrations should be monitored. (B)
3. If serum triglyceride concentrations exceed 200 mg/dL in the neonate, lipid emulsion infusion should be suspended and then restarted at a rate of 0.5 to 1 g/kg per day. (B)
4. Intravenous heparin, at a dose of 1 unit/mL of PN fluids, should be given to enhance the clearance of lipid emulsions. (B)
5. A trial of carnitine supplementation should be given to premature infants with unexplained hypertriglyceridemia. (B)
6. Infants should receive 20% lipid emulsion to improve clearance of triglycerides and phospholipids. (B)

中文翻译

1. 婴幼儿应按照每天0.5到1g/kg的起始剂量开始脂肪乳输注治疗，并以每天0.5g/kg的速度递增，最大剂量为每天3g/kg。（A）
2. 在早产儿和脓毒症患儿中脂肪乳的输注速率应降低，应监测血清甘油三酯浓度。（B）
3. 如果新生儿血清甘油三酯浓度超200mg/dL应暂停脂肪乳滴注，之后以每天0.5到1g/kg的剂量重新开始治疗。（B）
4. 在PN营养液中以1 unit/mL的剂量加入肝素静脉注射，应该可以提高脂肪乳的清除率。(B)
5. **出现不明原因的高甘油三酯血症的早产儿应补充左卡尼汀治疗。（B）**
6. 对于接受20%脂肪乳补充的婴儿，应该提高甘油三酯和磷脂的清除率。（B）

Section XII: Administration of Specialized Nutrition Support-Issues Unique to Pediatrics 第12部分：特殊营养支持的管理—专门针对儿科的问题

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COMPLICATIONS UNIQUE TO NEONATES:HEPATOBILIARY

新生儿并发症：**肝胆科**（见该指南第107页）

原文摘录

Background

PN-associated cholestasis (PNAC) is the most common and life-threatening long-term complication of PN in children.[1–3] About 30% to 60% of children develop PN-associated hepatic dysfunction during long-term PN.[4] The mean time to onset of PNAC was reported to

tecting against PNAC are inconclusive. Carnitine is not a routine constituent of PN formulas and carnitine deficiency in PN patients has been suggested as a predisposing factor to liver dysfunction. Two case reports of adults with hepatocyte fatty infiltration showed improvement in LFTs and normalized bilirubin concentrations with carnitine supplementation.[48,49] Another report in four adults showed no change in liver morphology with carnitine,[50] and declines in carnitine levels probably have little adverse effect.[51] Cyclic infu-

中文翻译

背景：PN相关性胆汁淤积（**PNAC）是最常见的且危及生命**的小儿PN患者**慢性并发症**[1-3]。大约**30%-60%**的儿童在长期PN期间会发生PN相关的肝功能异常[4]。

左卡尼汀不是PN配方中的一种常规组成成分，PN患者**左卡尼汀的缺乏已被证实为肝功能紊乱的易感因素**。2例关于肝细胞脂肪浸润的成人患者的报道显示，**左卡尼汀补充治疗可以改善肝功能，使胆红素浓度恢复正常**[48,49]。另一例4名成人患者的病例报道显示左卡尼汀对肝脏形态无影响[50]，卡尼汀浓度的下降可能有很小的不利影响[51]。

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INBORN ERRORS OF METABOLISM 先天代谢异常（见该指南第129页）

原文摘录

minimizing any metabolic complications. Examples of nutritional therapy for these disorders include the following:

1. Restricting substrates in patients with blocked metabolic pathways to prevent accumulation of toxic precursors such as phenylalanine (phenylketonuria), galactose (galatosomia), and leucine (maple syrup urine disease);
2. Facilitating alternative metabolic pathways to decrease accumulated toxic precursors in blocked reaction sequences (eg, glycine therapy in patients with isovaleric acidemia);
3. Supplying products of blocked primary pathways [eg, arginine in patients with urea cycle disorders, tyrosine in phenylketonuria (PKU), and glucose in glycogen storage disease type I];
4. Supplementing conditionally essential nutrients (eg, carnitine in patients with organic acidemias);

中文翻译

先天代谢障碍儿童营养治疗的目标包括优化生长和发育，尽可能减少代谢并发症。针对这些障碍的营养治疗实例如下：

1、限制代谢途径受阻的底物的摄取，以防止毒性前体的堆积；

2、促进其他的代谢途径以减少受阻反应序列中堆积的毒性前体物质；

3、补充受阻的主要代谢途径的反应产物；

4、有条件地补充必需营养素（如有机酸尿症患者应补充左卡尼汀）；

5、。。。。。

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HOME SPECIALIZED NUTRITION SUPPORT 家庭专业化营养支持（见该指南第137页）

原文摘录

Iron studies, platelet, folate/vitamin B-12, and carnitine levels should be obtained as indicated. Trace element studies every 2 to 6 months and fat-soluble vitamin assessment every 6 to 12 months are appropriate.

中文翻译

铁的检测，血小板，叶酸/维生素B12以及左卡尼汀水平应该达到参考值。

每2-6个月进行一次微量元素检测，每6-12个月进行一次水溶性维生素的评估是合理的。

参考文献

3. Borum PR: Carnitine in neonatal nutrition. J Child Neurol 10(Suppl 2):2S25-2S31, 1995

20. Schmidt-Sommerfeld E, Penn D, Wolf H: Carnitine deficiency in premature infants receiving total parenteral nutrition: Effect of L-carnitine supplementation. J Pediatr 931-935, 1983

21. Tibboel D, Delemarre FMC, Przyrembel H, et al: Carnitine deficiency in surgical neonates receiving total parenteral nutrition. J Pediatr Surg 25:418-421, 1990

22. Larson LE, Olegard R, Ljung ML, et al: Parenteral nutrition in preterm neonates with and without carnitine supplementation. Acta Anaesthesiol Scand 34:501-505, 1990

23. Smith RB, Sachan DS, Plattsmier J, et al: Plasma carnitine alterations in premature infants receiving various nutritional regimens. JPEN 12:37-42, 1988

24. Helms RA, Mauer EC, Hay WW Jr et al. Effect of intravenous L-carnitine on growth parameters and fat metabolism during parenteral nutrition in neonates. JPEN 14:448-453, 1990

25. Bonner CM, DeBrie KL, Hug G, et al: Effects of parenteral L-carnitine supplementation on fat metabolism and nutrition in premature infants. J Pediatr 126:287-292, 1995

48. Palombo JD, Schnure F, Bistrian BR, et al: Improvement of liver function tests by administration of L-carnitine to a carnitine deficient patient receiving home parenteral nutrition: A case report. JPEN 11:88-92, 1987

49. Worthley LI, Fishlock RC, Snoswell AM: Carnitine deficiency with hyperbilirubinemia, generalized skeletal muscle weakness and reactive hypoglycemia in a patient on long-term total parenteral nutrition: treatment with intravenous L-carnitine. JPEN 7:176-180, 1983

50. Bowyer BA, Miles JM, Haymond MW, et al: L-carnitine therapy in home parenteral nutrition patients with abnormal liver tests and low plasma carnitine concentrations. Gastroenterology 94:434-438, 1988

51. Moukarzel AA, Dahlstrom KA, Buchman AL, et al: Carnitine status of children receiving long-term total parenteral nutrition: a longitudinal prospective study. J Pediatr 120:759-762, 1992

备注：参照指南原文中的参考文献编号，未重新排序

谢 谢！