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。

Premature infants have low carnitine reserves. Low

carnitine levels can also be seen in full-term neonates

receiving long-term PN.2o,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.

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。

Practice Guidelines Complications Unique to Neonates: .Hypertriglyceridemia,

专门针对新生儿的并发症：高甘油三酯血症的临床实践指南

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)

**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过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）**

**COMPLICATIONS UNIQUE TO NEONATES:HEPATOBILIARY**

**新生儿并发症：肝胆管的**

**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.’**

**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。**

**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**

**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]。**

**Practice Guidelines Extracorporeal Membrane# Oxygenation 实践指南：体外膜氧化**

**Homocysteine, carnitine, and glutamine are three**

**nutrients currently under study.**

**同型半胱氨酸，左卡尼汀，谷氨酰胺是三种目前正在研究的营养物质。**

**There are insufficient data to support**

**the routine use of L-carnitine in chronic dialysis**

**patients; however, it may help treat erythropoeitinresistant**

**anemia.8**

**没有充分的证据支持长期透析患者使用左卡尼汀，然而，左卡尼汀确实可以治疗EPO抵抗性贫血[8]。**

**INBORN ERRORS OF METABOLISM 先天代谢异常**

**Goals of nutrition support in children with inborn errors of metabolism**

**include optimizing growth and development and**

**minimizing any metabolic complications. Examples of**

**nutritional therapy for these disorders include the following:**

**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、 。。。。。**

**HOME SPECIALIZED NUTRITION SUPPORT 家庭专业化营养支持**

**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.**

**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。**