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Carnitine Deficiency: A Missed Diagnosis

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Carnitine Deficiency: A Missed Diagnosis

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BY DIANE W. SHANNON, MD, MPH, AND GARY S. WOLFE, RN, CNA, CCM

Carnitine deficiency in pediatric patients is an underdiagnosed disorder that, left untreated, may have serious clinical consequences and substantially impair quality of life. The article describes signs and symptoms, diagnosis, treatment, and consequences of a missed diagnosis, and discusses case management issues in carnitine deficiency, including suggestions for early case management intervention.

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Monday 8:30 a.m. You check in for your messages and discover that one of your pediatric patients has just been readmitted. Jennifer has spent most of her 12 months of life in the hospital with recurrent bouts of unexplained hypoglycemia. She has also had frequent upper respiratory and gastrointestinal infections. Every few weeks she is back in the ER again. Her parents are understandably panicked. Her pediatrician has no definitive answers. And your organization is concerned about the high cost of repeated admissions. What is going on with Jennifer?

It may be that Jennifer is suffering from a disorder that, when considered, can be noninvasively and inexpensively diagnosed and treated. Carnitine deficiency is an enigmatic disorder, presenting in a myriad of ways and often masking as nondescript symptoms, such as recurrent infections, hypoglycemia, failure to thrive, or cardiomyopathy of unknown etiology. In addition, carnitine deficiency often signals an underlying metabolic disease, iatrogenic factor, or acquired medical condition that needs to be addressed. Although there is a paucity of definitive statistics on the prevalence of carnitine deficiency, estimates suggest that it may be quite common in certain populations.

Dr. Susan Winter, Medical Director of Medical Genetics/Metabolism at Valley Children's Hospital in Fresno, CA, and an expert on carnitine deficiency, found a disturbingly high rate of carnitine deficiency in a 1987 study.[1] Dr. Winter and her colleagues screened 165 children deemed to be at risk for carnitine deficiency, because they displayed symptoms such as failure to thrive, cardiomyopathy, nonketotic hypoglycemia, or a disorder known to predispose to carnitine deficiency, such as a renal disorder. A surprising 32% of the children tested had carnitine deficiency.

Could your organization be caring for patients with scenarios similar to Jennifer's? Finding out may be crucial to your patients' health and the health plan's financial bottom line. Recurrent hospitalizations, repeated ER visits, and frequent relapses of nondescript symptoms add up to decreased health and quality of life for your patients and increased costs to the health plan. In addition, family members frustrated by their loved one's repeated relapses without definitive diagnoses do not make for satisfied health plan members. For these reasons and more, carnitine deficiency is a diagnosis you don't want to miss.

Carnitine is a naturally occurring amino acid derivative, produced endogenously in the liver and kidneys[2] and obtained in the diet from red meat and dairy products. Carnitine blood levels increase during the first year of life and then remains steady through-

out childhood and adulthood.[3] The normal serum level for infants is 25 mol/liter,[4] while the normal level for adults ranges from 30 to 89 mol/liter.[5] Carnitine serves two crucial functions in the body. It transports long-chain fatty acidsone of the body's key forms of fuel into the mitochondria, where the molecules undergo fatty acid oxidation, releasing energy in the form of ATP. This energy-producing process is especially important in the heart, liver, and skeletal muscles—organs that rely heavily on fat degradation for energy.[6] Carnitine also acts as a waste remover, transporting fatty acid esters (a waste product of fatty acid oxidation) out of the mitochondria to be excreted from the body.[6] If either of carnitine's vital functions is disrupted, the ability to produce energy by metabolizing fatty acids and to avoid buildup of toxic wastes may be impaired.

Carnitine Deficiency

Carnitine deficiency is defined as a level of carnitine in the plasma or tissues that is insufficient for normal function.[6] While carnitine levels can be measured in plasma, tissue, or urine, plasma values are generally used in clinical practice. However, while plasma levels may be easier to obtain, they do not always register carnitine deficiency in the tissues, where the majority of carnitine is stored.

Carnitine requirements vary by age, diet, and metabolic conditions such as fasting and stress.[6] Under normal conditions, adequate tissue levels of carnitine are maintained through biosynthesis, dietary intake, and renal reabsorption. Carnitine balance can be disrupted by decreased synthesis, decreased intake, malabsorption, increased loss, or decreased mitochondrial membrane transport of carnitine.[7] Increased renal excretion of carnitine due to renal disorders or hemodialysis may cause an excess loss of carnitine. Infants are especially susceptible to carnitine deficiency, because they cannot readily synthesize the compound until six months of age and because their diets may not contain carnitine (Table 1).

Primary and Secondary Carnitine Deficiency

Carnitine deficiency has only been recognized as a disorder relatively recently. In 1973, Engel and Angelini described a case of carnitine deficiency localized to the muscle.[8] Two years later, Karpati and colleagues first described a case of systemic carnitine deficiency.[9] Carnitine deficiency has been divided into primary and secondary types, the former showing a profound reduction in carnitine levels in affected tissues unassociated with another underlying abnormality. Primary carnitine deficiency is felt to be fairly rare and generally presents in one of two syndromes: myopathic, in which the serum levels of carnitine are normal but the muscle levels are low; and systemic, in which the serum, muscle, and often cardiac levels are very low.[10]

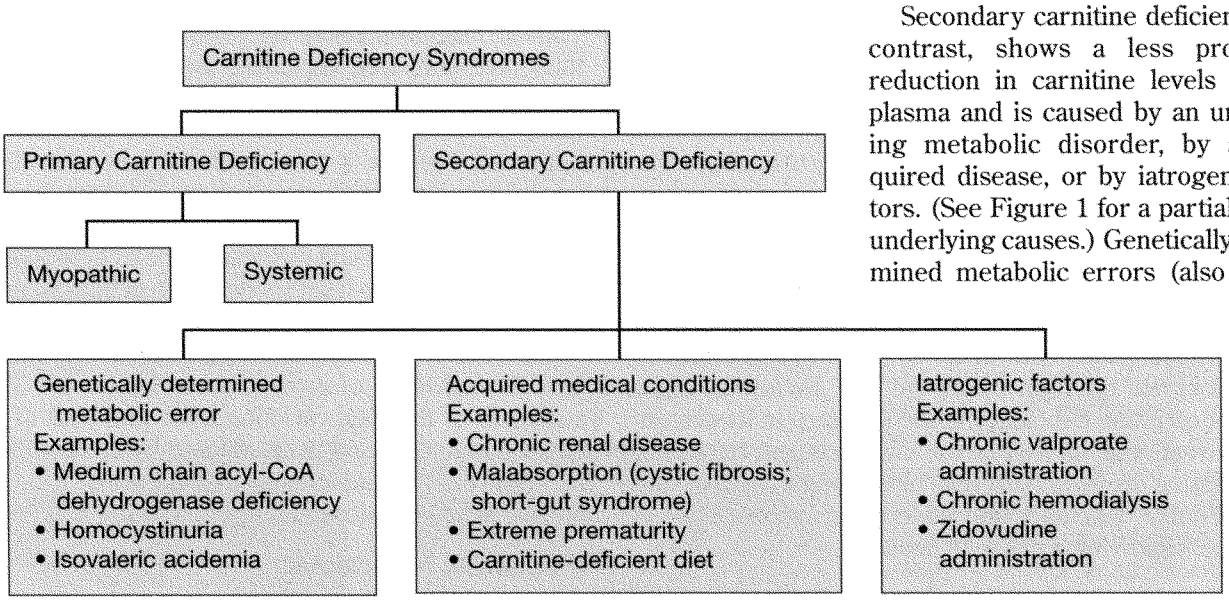
Secondary carnitine deficiency, by contrast, shows a less profound reduction in carnitine levels in the plasma and is caused by an underlying metabolic disorder, by an acquired disease, or by iatrogenic factors. (See Figure 1 for a partial list of underlying causes.) Genetically determined metabolic errors (also called

TABLE 1. CARNITINE AND PEDIATRIC FOOD SOURCES

Pediatric Food Source	Contains Carnitine
Breast milk	Yes
Colostrum (at term)	Yes
Cow-milk-based products	Yes
Semi-elemental formulas based on hydrolyzed cow milk	No
Soy-based products	No, unless added
TPN	No, unless added

TPN=Total Parenteral Nutrition
Sources: Rubaltelli FF, Orzali A, Rinaldo P, Donzelli F, Carnielli V. Carnitine and the premature. *Biol Neonate.* 1987;52(S1):65-77.
Borum PR, et al. Carnitine content of liquid formulas and special diets. *Amer J Clin Nutr.* 1979;32:2272-2276.

Fig 1. Carnitine deficiency Syndromes



Adapted with permission from DeVivo DC, Tein I. Primary and secondary disorders of carnitine metabolism. *International Pediatrics.* 1990;5(2):134-141. Information also included from: Pons R, DeVivo DC. Primary and secondary carnitine deficiency syndrome. *Journal of Child Neurology.* 1995; 10(25):2S8-2S24.

inborn errors of metabolism) commonly cause a disruption in fatty acid metabolism[10] and are often the underlying cause of carnitine deficiency.

Signs and Symptoms

Carnitine deficiency can mimic other disorders, prompting diagnostic and treatment procedures that may make it difficult to diagnose. In addition, secondary carnitine deficiency is, by definition, found in conjunction with other abnormalities. The diagnosis is also complicated by the fact that often carnitine deficiency cannot be diagnosed until a stressor event, such as fever or fasting, triggers a metabolic crisis. With such an indistinct and often masked presentation, the diagnosis of carnitine deficiency may be easily missed.

There are, however, a series of signs that can alert you to the possibility of carnitine deficiency. Being aware of these warning signs may result in a prompt and accurate diagnosis, which prevents continued ill-health and costly relapses. These warning signs include failure to thrive (FTT), recurrent infections, muscle weakness (hypotonia), lethargy, cardiomyopathy of unknown etiology, and liver dysfunction of unknown etiology (Table 2). The younger the patient and the more warning signs present, the greater the likelihood of carnitine deficiency.[11]

In addition to these warning signs, there are other key signs and predisposing factors to keep in mind. The previously described 1987 Winter study[1] used the following list to determine the need to test for carnitine deficiency:

- cardiomyopathy
- encephalopathy
- nonketotic hypoglycemia
- hypotonia
- failure to thrive
- recurrent infections
- organic aciduria
- renal disease
- gastrointestinal disease
- carnitine-deficient diet
- therapy with valproic acid

Many of these factors are common in hospitalized pediatric patients, and some patients may show several risk factors or symptoms concurrently; thus, a low threshold for suspicion of carnitine deficiency may be key to its diagnosis.

After Maria Shaffer, RN, CCM, a case manager in central Pennsylvania read about carnitine deficiency, she was certain she had seen cases of undiagnosed carnitine deficiency in her caseload. "In the past 6 months, I have seen 5 or 6 pediatric patients with FTT and the diagnosis of nonspecific developmental delay. . . .I read [about carnitine deficiency] and the light bulb went off." Becoming more aware of carnitine deficiency is the first step in its diagnosis and treatment, as well as the diagnosis and treatment of underlying disorders that may be causing it. Addressing both carnitine deficiency and underlying abnormalities is crucial to improving a patient's health.

Diagnosis

While the diagnosis of carnitine deficiency may be straightforward in the research setting, there are several caveats to its diagnosis in the clinical arena. First, a tissue sample is needed to document a cell level insufficiency of carnitine. However, tissue samples may not be available in the clinical setting due to time constraints or other factors. Thus, practitioners most commonly diagnose carnitine deficiency based on serum samples. Second, in the clinical setting, laboratory results may not be available in a timely fashion. Most samples for carnitine level determination must be sent to a reference lab, and results may not be available for a few days. In crisis situations and sometimes in nonacute ones, clinicians often elect to treat empirically before receiving lab results. Third, in order to accurately diagnose metabolic defects during a triggered crisis, serum samples must be drawn prior to the administration of glucose, since treatment may obscure metabolic abnormalities. If an emergency situation precludes obtaining lab values before glucose administration, serum samples may be drawn immediately after therapy.[12]

TABLE 2. WARNING SIGNS OF CARNITINE DEFICIENCY

FTT (Failure to Thrive)
Recurrent infections
Muscle weakness (Hypotonia)
Lethargy
Cardiomyopathy
Cardiomyology of unknown etiology
Liver dysfunction of unknown etiology

Source: In conversations with SC Winter (February 10, 1997, and March 20, 1997).

Despite these challenges, it is possible to make a relatively prompt and reliable diagnosis of carnitine deficiency. Clinical diagnosis relies on several serum tests: plasma total carnitine, free carnitine, acyl-carnitine (the bound form of carnitine), and the acyl-carnitine/free carnitine ratio. Obtaining the acyl-carnitine/free carnitine ratio is important because it reflects abnormalities in the tissue level (mitochondrial) metabolism.[13] Some laboratories include this calculation. To calculate it yourself, take the acyl-carnitine value and divide it by the free carnitine value. A low free carnitine level (generally less than 20 mol/liter) or a high acyl-carnitine/ free carnitine ratio (greater than 0.4) should prompt consideration of carnitine therapy and further evaluation of organic acid disorders.[14] The measurement of the total carnitine level (the sum of the free and the bound forms) is considered by some clinicians to be relatively unreliable, since, on occasion, it can appear normal despite clinically relevant carnitine deficiency.[1]

Diagnostic Algorithms

Figure 2 presents an algorithm adapted from the work of Dr. Susan Winter that outlines the steps for diagnosing carnitine deficiency. Algorithms, such as this one, could be

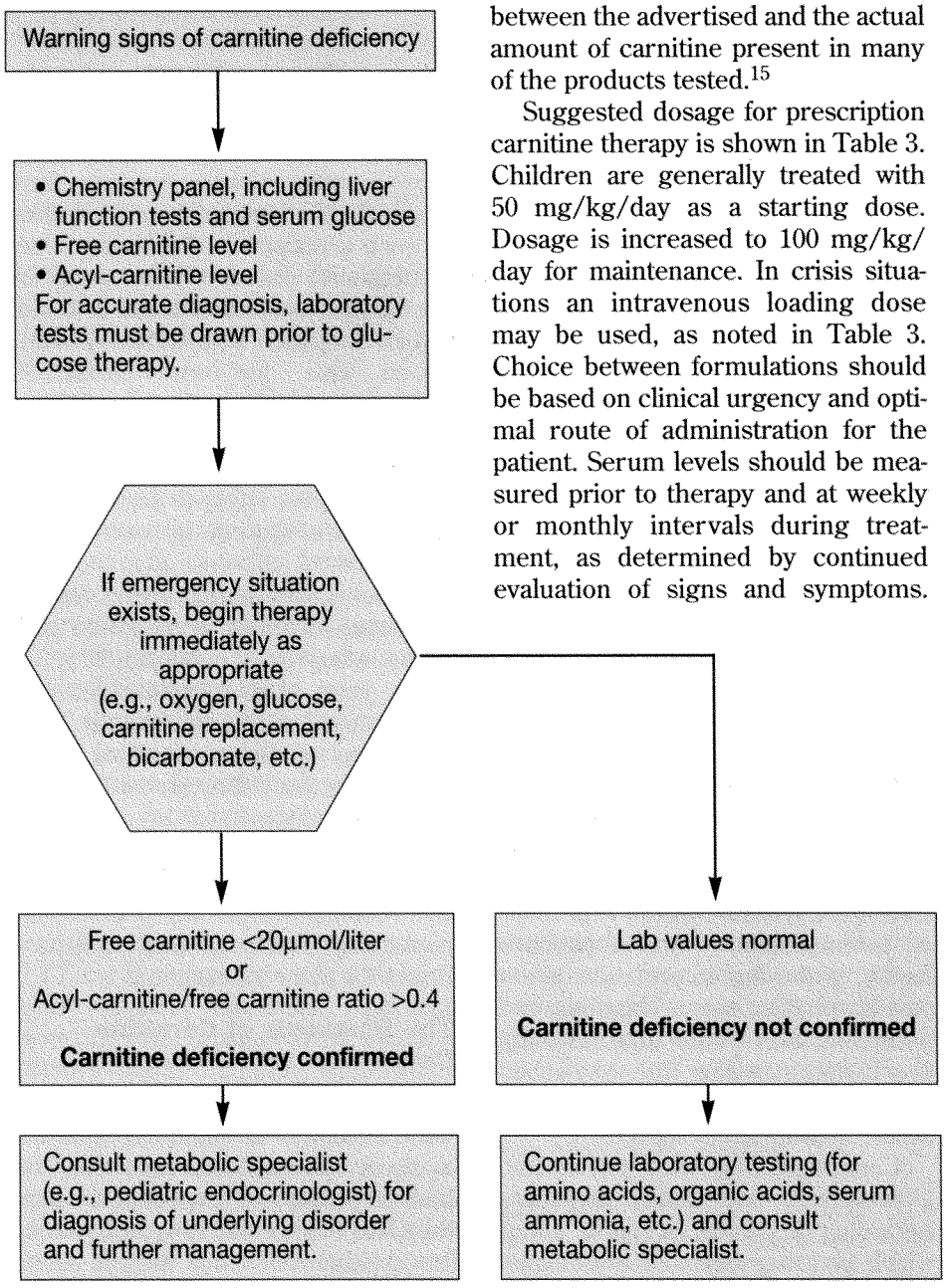
used to create care pathways for children presenting with symptoms suggestive of metabolic abnormalities such as carnitine deficiency.

Treatment

Levocarnitine, or L-carnitine, the naturally occurring racemic form of carnitine, is available for therapeutic intervention. Prescription therapy is available only under the brand name Carnitor as tablets, oral solution, and ampoules for intravenous injection. While non-prescription carnitine products are also available in tablet, capsule, and syrup forms from health food stores, they are marketed for athletic rather than therapeutic use. In addition, health food products are not subject to the same regulatory requirements as prescription products. In fact, a 1993 Duke University study indicated a lack of quality control in the non-prescription carnitine products, finding inconsistent disintegration rates and/or variance between the advertised and the actual amount of carnitine present in many of the products tested.[15]

Suggested dosage for prescription carnitine therapy is shown in Table 3. Children are generally treated with 50 mg/kg/day as a starting dose. Dosage is increased to 100 mg/kg/day for maintenance. In crisis situations an intravenous loading dose may be used, as noted in Table 3. Choice between formulations should be based on clinical urgency and optimal route of administration for the patient. Serum levels should be measured prior to therapy and at weekly or monthly intervals during treatment, as determined by continued evaluation of signs and symptoms.

Fig 2. Diagnostic algorithm for carnitine deficiency



Source: In a conversation with Dr. Susan Winter, February 10, 1997; and Winter, SC. Diagnosing metabolic disorders: one step at a time. *Contemporary Pediatrics*. October 1993:35-63.

Side effects to treatment are rare, but diarrhea, nausea and vomiting, and body odor (at high oral doses) have been reported. There are no contraindications or warnings for the drug's use, and toxicity from overdosage has never been reported.[16]

The cost of prescription carnitine therapy is modest compared with the other costs incurred with many chronic illnesses. Oral maintenance therapy costs approximately $2 per gram (based on AWP price), while intravenous therapy costs approximately $36 per gram (AWP price). The cost of non-prescription carnitine products is approximately $2 per gram at retail prices.

Consequences of a Missed Diagnosis

In an ideal world, diagnosis of illness would be precise, complete, and immediate. In the real world, diagnosis is often an imperfect art. What is the bottom line with regard to carnitine deficiency? What are the consequences of a missed diagnosis of carnitine deficiency? Obviously, we cannot know for certain the effects on patients who, by definition, are not classified as having the disorder. However, evaluating the experiences of patients who are diagnosed provides us with some useful insights.

First, there are clinical consequences to a missed diagnosis. These include continued symptoms, such as failure to thrive, cardiac dysfunction, recurrent infections, and even death. A 1995 study found a correlation between defects in fatty acid oxidation and sudden infant death syndrome (SIDS),[17] a tragic outcome to undiagnosed carnitine deficiency. A less tragic but debilitating effect is continued developmental delay.

Second to consider are the effects of a missed diagnosis on quality of life. While direct studies of quality of life have not been performed, one can infer that the development of a child who experiences repeated hospitalizations or infections, or other effects of untreated carnitine deficiency or its causative disorder, is adversely affected. Likewise, families who sense that their child's underlying disorder has not been properly diagnosed or treat-

TABLE 3. RECOMMENDED DOSAGE OF PRESCRIPTION CARNITINE THERAPY

Dosage Form	Recommended Dosage		Comments
	Pediatric	Adult	
Tablets	50 to 100 mg/kg/day in divided doses; starting dose is 50 mg/kg/day	990 mg two or three times per day	Pediatric maximum is 3 g/day
Oral Solution	50 to 100 mg/kg/day (0.5 to 1.0 mL/kg/day) in divided doses; starting dose is 50 mg/kg/day	1 to 3 g/day (10 to 30 mL/day) for a 50 kg person; starting dose is 1 g/day	Pediatric maximum is 3g/day (30mL/day)
Intravenous Solution	50 mg/kg as a slow injection or by infusion; recommended subsequent dosage is 50 mg/kg/day	same as pediatric	for crisis situations a loading dose is often administered, with an equivalent dose administered over the next 24 hours at q3h or q4h intervals

Carnitor® Package Insert, Sigma-Tau Pharmaceuticals, Inc., Gaithersburg, MD.

ed will suffer. Other effects on a patient's family may include missed days at work, child care issues, and a significant emotional toll. It is easy to imagine the impact that the ongoing crises and relapses associated with undiagnosed carnitine deficiency could have on families.

Lastly, untreated carnitine deficiency can increase health care costs. In a retrospective analysis of 20 pediatric patients, Dr. Susan Winter compared the costs associated with patients who had carnitine deficiency due to metabolic disease that was diagnosed and treated with prescription therapy versus patients with carnitine deficiency due to metabolic disease that was not diagnosed and, consequently, not appropriately treated (Table 4).

In this analysis, the cost savings was estimated to be $8,774.00 per year. Note that $600 per day for hospitalization fees is a conservative estimate; cost savings will be even greater with higher hospitalization costs. Other anecdotal evidence suggests that in some circumstances the savings may be even more substantial. Five years ago, Dr. Winter treated a child with cardiomyopathy of unknown etiology who was awaiting cardiac transplantation. After empirical treatment with carnitine therapy, the child's cardiac function improved and she was able to avoid cardiac transplantation. Later testing revealed that the child had an underlying metabolic disorder.[18] While this case may not be typical, the savings in terms of both quality of life and cost are obviously immense.

Case Management Issues

What about the case management issues surrounding the diagnosis of a suspected case of carnitine deficiency? The most salient issues surround communication of your suspicion to your patient's family members and health care providers. In such a situation, Juanita Lopez, RN, a PhD candidate in holistic nursing and case manager for an HMO in New York City, would rely on her already established relationships. She might distribute an educational article to staff nurses or speak to the primary care physician directly. Maria Shaffer, RN, CCM, from Pennsylvania, would rely more on communication with the patient's family, gathering information about past hospitalizations, evaluations, and treatment. She would then help the providers, including the physician, and the family to understand and access appropriate resources.

Communicating with physicians, pharmacists, and family members regarding a clinical scenario about which you may be more knowledgeable than they is a delicate situation. As case managers in a focus group convened by Sigma-Tau Pharmaceuticals noted, tactful communication in such situations is a necessity. One participant stated, "The word is encourage. I will encourage. I would say [to the patient's physician], 'Listen, check this for me please.'"[19] Determine which strategies will successfully help your patient receive an accurate diagnosis and correct treatment. Use your ability to negotiate the health care system to assist your patients.

Other case management issues surround obtaining prescription carnitine when it is non-formulary, or when your plan doesn't cover the medication. Options include approaching your plan's medical director or pharmacy director with direct requests, substantiated by cost information and reprints of articles on carnitine deficiency; researching alternative funding sources for patients; or contacting pharmaceutical manufacturers for more information.

The Diagnosis of Carnitine Deficiency: What Case Managers Can Do

Case managers are uniquely situated to discern undiagnosed and recurrent health problems, especially when they lead to repeated hospitalizations, as carnitine deficiency may. A chart review for illnesses in siblings, prior illnesses, diagnostic tests, treatments, and hospitalizations, in conjunction

TABLE 4. COST COMPARISON BETWEEN TREATED AND UNTREATED CARNITINE DEFICIENCY

	Undiagnosed Carnitine Deficiency	Diagnosed and Treated
Hospitalization (average number of days per month)	2.99/month	1.67/month
Total hospital days (average, per year)	35.88	20.04
Average cost of hospitalization (conservative estimate, non-ICU costs, 1996 dollars)	$600.00/day	$600.00/day
Total cost of hospitalization	$21,528.00	$12,024.00
Total carnitine therapy (Carnitor® tablets/oral solution)	0	365 g (100 mg/kg/day for 10 kg child for 1 year)
Cost of carnitine therapy	0	$730.00 ($2.00/g, AWP price, 1996 dollars
Total cost	$21,528.00	$12,754.00

Adapted from an unpublished 1996 study by Dr. Susan Winter.

with an awareness of the warning signs of this disorder can provide you with powerful clues about potential carnitine deficiency.

Case managers are also well situated to provide information on carnitine deficiency to family members, health care providers, plan administrators, and professionals acting in a care management capacity. Informal networking, in-services, and case-specific discussions are excellent opportunities to educate others about this underdiagnosed malady.

In a more direct capacity, case managers shape the quality and completeness of care by influencing the development, use, and enforcement of care pathways. Consider the ways in which algorithms such as the one detailed in Figure 2 could be incorporated into existing or future care pathways for FTT, cardiomyopathy, and other high-cost pediatric diagnoses. Approach your medical management administrators about updating appropriate care pathways. *After noting Jennifer's recent presentation and frequent hospitalizations, you place a call to her pediatrician, inquiring about the possibility of carnitine deficiency and letting her know that the health plan would cover serum testing for carnitine deficiency. Testing reveals carnitine deficiency, which responds well to carnitine therapy, initially administered intravenously and later orally. Two months later you place a follow-up call to Jennifer's mother, who tells you that Jennifer is being treated for an underlying metabolic disorder but has had no metabolic crises requiring hospitalization since beginning carnitine and other appropriate therapy.*

Conclusion

Carnitine deficiency is an underdiagnosed disorder that may signal the presence of another abnormality. With attention to warning signs and with appropriate testing and therapy, carnitine deficiency can be cost-effectively treated. Case managers are uniquely positioned to recognize carnitine deficiency, prompt its diagnosis and treatment, and effect investigation of other underlying disorders. An awareness of carnitine deficiency can help you improve your patients' health and quality of life, as well as your organization's financial bottom line. TJCM

For More Information:

Journal of Child Neurology, supplement devoted to L-Carnitine. November 1995; Volume 10, Supplement 2. Reprint contact: The Sheridan Press, Hanover, PA (717) 632-3535.

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Sigma-Tau Pharmaceuticals, Inc. Medical Information: toll-free information number (800) 447-0169. Informational video available.

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CEU APPLICATION

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Carnitine Deficiency: A Missed Diagnosis

This educational manuscript has been approved for one (1) hour of CCM education credit by The Commission for Case Manager Certification.

To obtain your education credit, please submit a copy of this form:

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Questions:

_____ 1. In the 1987 study by Dr. Susan Winter, what percentage of children deemed to be at risk for carnitine deficiency were found to have the disorder when tested?
A. 5%
B. 12%
C. 20%
D. 32%

_____ 2. Which of the following is true? (More than one may be correct.)
A. Carnitine is obtained in the diet from red meat and dairy products.
B. Carnitine is absent from soy-based products, unless it has been added.
C. Renal disorder and hemodialysis never cause excess carnitine losses.
D. Infants can readily synthesize adequate amounts of carnitine at birth.

_____ 3. Which of the following is known to cause secondary carnitine deficiency? (More than one may be correct.)
A. blood transfusions
B. zidovudine administration
C. chronic valproic acid administration
D. chronic hemodialysis

_____ 4. Which of the following is *not* considered a warning sign of carnitine deficiency? (More than one may be correct.)
A. headaches
B. failure to thrive
C. lethargy
D. muscle weakness

_____ 5. Which of the following is a potential consequence of a missed diagnosis of carnitine deficiency? (More than one may be correct.)
A. increased health care costs
B. emotional toll on families
C. developmental delay
D. all of the above

_____ 6. Which of the following are actions mentioned in the article that case managers may take in a suspected case of carnitine deficiency? (More than one may be correct.)
A. distribute educational materials to staff nurses
B. order serum lab tests for carnitine deficiency
C. review history of hospitalizations, evaluations, and treatments
D. help providers and family members to understand and access appropriate resources

Name:_____ CCM ID# _____ CRC#_____ CDMS #_____

Address: _____

City: _____ State: _____ Zip: _____

Signature: _____

An individual application and $10 payment must accompany each request. Applicants who do not score 80% or above may re-apply with another application and additional $10 payment. Documentation of credit and an approval number will be mailed from The Commission for Case Manager Certification.