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Use of propofol for short duration procedures in children with long chain 3-hydroxyacyl-CoA dehydrogenase (LCHAD) or trifunctional protein (TFP) deficiencies

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Abstract

The medication propofol, commonly used for anesthesia, has been avoided in patients with mitochondrial fatty acid oxidation disorders (FAODs) due to concerns that it contains long-chain fatty acids (LCFAs), and because of reports of severe side effects in some critically ill patients receiving high-dose propofol infusions that mimic some of the symptoms regularly found in FAOD patients. In this secondary analysis, we examined the outcomes of 8 children with long-chain 3-hydroxyacyl CoA dehydrogenase (LCHAD) deficiency or trifunctional protein (TFP) deficiency who were repeatedly sedated for an electroretinogram (ERG) as part of a longitudinal study of the progression of chorioretinopathy commonly found in this population. A total of 39 sedated ERG procedures were completed using propofol for sedation. The propofol dosing, estimated total energy needs of the subject, and inpatient dietary intake recording was complete in 32 of these procedures. The LCFAs in the propofol provided approximately 1.0% of the average total daily energy needs. The sedation with propofol resulted in no adverse side effects and was safely used in this short duration procedure.

Keywords

Long-chain 3-hydroxyacyl-CoA dehydrogenase deficiency; Trifunctional protein deficiency; Propofol; anesthesia; fatty acid oxidation disorders

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Introduction

Management of routine medical procedures in patients with mitochondrial fatty acid oxidation disorders (FAODs) is complicated by the inherent metabolic defect. Patients with a FAOD have a decreased ability to use fatty acids to generate energy that decreases their ability to tolerate fasting. Symptoms observed among patients with long-chain FAODs include hypoketotic hypoglycemia due to fasting or the stress of illness during infancy, cardiomyopathy, hepatic steatosis, rhabdomyolysis, and sudden death [1]. Treatment for long-chain FAODs includes fasting avoidance and consumption of a diet that limits long-chain fatty acid (LCFA) intake to 10% of total energy intake while providing 10-20% of energy as medium chain triglycerides [2]. Current practice for routine medical procedures that require general anesthesia include prolonged fasting prior to the procedure, which increases the risk for hypoketotic hypoglycemia among patients with long-chain FAODs.

Providing safe general anesthesia for people with FAODs has been a concern because propofol, a popular anesthetic agent, is prepared with long-chain fatty acids for intravenous administration. Propofol, approved for intravenous administration in the United States since 1989, is used to induce and maintain sedation [3, 4]. It has achieved widespread use because an effective level of sedation is achieved rapidly, and patients are easily aroused and recover quickly after the infusion stops. Rapid recovery from procedures that require sedation and resumption of oral intake could be desirable in patients with FAODs, in which the ability to fast is limited [5]. However, propofol is formulated as an oil and water emulsion which contains soybean oil (100 mg/mL), composed of fatty acids such as alpha-linoleic acid, linoleic acid, oleic acid, stearic acid and palmitic acid, all long-chain fatty acids of 16 to 18 carbons in length [3, 6]. The safety of providing long-chain fatty acids, even in the small doses usually needed for short-term sedation has been questioned in people with FAODs [7-9].

Concern about the use of propofol in all children (not just those with fatty acid oxidation disorders) was generated when metabolic acidosis and myocardial failure were reported in five critically ill children following prolonged propofol infusion [10]. A new syndrome, propofol infusion syndrome (PRIS), was defined by a published review that reported the cases of eighteen children who had suffered serious adverse events while receiving high-dose, sustained infusions of propofol [11]. The syndrome was defined as the sudden onset of treatment resistant bradycardia progressing to asystole with at least one of the following: the presence of lipemic plasma, enlarged liver, metabolic acidosis, or muscle involvement with evidence of rhabdomyolysis or myoglobinuria. None of the 18 children involved were known to have a fatty acid oxidation disorder and only five of the children (28%) had muscle involvement [11]. AstraZeneca Pharmaceuticals conducted an unpublished randomized controlled trial in 327 pediatric patients receiving propofol for sedation and found a non-significant dose dependent increase in 28-day mortality in the propofol group (8% mortality for 1% propofol and 11% mortality for 2% propofol) compared to standard sedation (4% mortality) [12]. This trial was stopped early; and even though propofol was already only approved for use in general anesthesia in the pediatric population, the US Food and Drug Administration (FDA) issued a product labeling change in 2001 recommending against off-label use of propofol for sedation in pediatric patients in intensive care units [4].

Concern about infusing long-chain fatty acids, and the similarity of reported side effects of high-dose, sustained infusions of propofol to symptoms of FAODs, appears to have led to recommendations to avoid propofol use in all patients with FAODs. To date there are no published reports specifically citing the use of propofol in patients with FAODs. In this study, propofol was used safely in eight children with long-chain 3-hydroxyacyl CoA dehydrogenase (LCHAD) or trifunctional protein (TFP) deficiency when they received short-term sedation for a total of 39 ophthalmological electroretinogram (ERG) procedures. ERGs were conducted in these patients to investigate the progression of the unique pigmentary retinopathy associated with LCHAD deficiency [13, 14].

Methods

A total of 14 children ranging in age from 9 months to 12 years were enrolled in this study, which was originally designed to determine if dietary therapy with the addition of docosahexaenoic acid (DHA) supplementation could prevent the progression of chorioretinopathy which occurs in children with LCHAD or TFP deficiency. The results of this research and a detailed report of the methods used have already been published [14]. The methods pertaining to this examination of sedation tolerance are discussed below.

The children in this study were enrolled at Oregon Health & Science University, Portland, OR) and diagnosis of LCHAD or TFP deficiency was confirmed biochemically or molecularly. Study visits occurred twice in the first year and then annually for up to 5 years for each volunteer and involved a 3-day inpatient stay in the Oregon Clinical and Translational Research Institute (OCTRI). All of the subjects were counseled to follow a diet low in long-chain fat (10% of calories) and most were also supplemented with medium chain triglycerides (MCT) and carnitine in addition to the study supplement, DHA. Prior to admission, each subject's usual dietary intake was recorded for 3 days by their legally authorized representative (LAR) and was analyzed for nutrient content using commercially available software (Food Processor, ESHA Research, Salem, OR, 1996). Each subject's actual dietary intake was also recorded and analyzed with the Food Processor software during all inpatient study visits. Energy average of the home 3-day food record was used to estimate usual intake, because children often have specific food preferences which are not satisfied in an inpatient setting and result in altered dietary intake. The OHSU Institutional Review Board approved the protocol and each subject's LAR gave informed consent. Children older than 7 years also gave assent to participate. The study was conducted from 1996 to 2004 and was not registered on clinical trials.gov. Evaluations at the baseline and annual visits included physical, neurological, and ophthalmologic examinations. Fasting plasma fatty acid analysis, a metabolic panel, liver function tests, and plasma acylcarnitines were also measured.

The ophthalmologic examinations included a clinical ERG and a determination of visual acuity with a swept-spatial-frequency visual evoked cortical potential (sweep VEP). Eight of the children who underwent these procedures were sedated using propofol injectable emulsion 1% (Diprivan®; AstraZeneca Pharmaceuticals, Wilmington DE), each one completing 3-6 procedures. Sedation was performed by a Pediatric Sedation Team from Doernbecher Children's Hospital in Portland, Oregon. As described in Table 1, all subjects

were instructed to consume a light meal 6 hours before sedation, to drink 4-6 oz of clear liquids such as apple or grape juice 4 hours prior to sedation, and then take nothing by mouth (NPO) until after the ERG. Intravenous administration of 10% dextrose with half normal saline (D₁₀ 1/2NS) was initiated approximately 45 minutes prior to the start of sedation as a precaution to reduce the risk of hypoglycemia during the procedure. The duration of sedation was less than an hour for these procedures, with most of them completed within 40 minutes. Sedation was discontinued rapidly after the procedure to allow the child to wake up and resume oral intake as quickly as possible. The children were monitored closely over the 24-hours following the procedure for any adverse events, but no laboratory testing was performed immediately after the procedure.

Results

Intake of Long-Chain Fatty Acids

Eight children enrolled in the study received propofol during the ERG procedure at 39 different study visits. Complete data on propofol dosing, the 3-day food diary results reflecting usual home intake, and actual dietary intake from inpatient diet records was available for 32 of the visits, which are summarized in Table 2. The amount of LCFAs found in each propofol dose was calculated from the actual dose provided during the procedure (10 mg propofol = 1 mL). Each milliliter of the propofol emulsion provided 100 mg of soybean oil [3]. The average dose of propofol given for the ERG sedation provided 1.83 grams of LCFA and the largest dose given was 3.10 grams. The recommended daily intake of LCFAs was 10% of calorie intake or less, and this amount was estimated for each subject based on the calorie intake averaged from 3 days of home diet diary records. The total amount of dietary fat to maintain 10% of total energy from LCFAs ranged from 6.75 to 29.50 grams per day due to the wide range of calorie needs in this population, which included children from infancy to adolescence. The actual range of daily dietary intake of LCFAs was greater (2.77 to 43.83 grams) as calculated during inpatient visits using food weighed before and after consumption. On average, the amount of LCFAs provided in the propofol emulsion provided just over 10% of the recommended allowance of daily LCFA intake, or just 1.0% of total energy needs. A review of anesthesia records from the procedures reveal that comments were made in 19 of the 39 procedures, recording the time from the discontinuation of propofol until the patient was awake and taking liquid calories. This time ranged from 13 to 30 minutes.

Adverse Event Reporting

No adverse events were observed in any of the subjects in response to the sedation used at the study visits. Because there were no symptoms of muscle pain, lethargy, or discolored urine during monitoring at the OCTRI in the 24-hours after the procedures, no laboratory measurements were completed. Between annual visits for the study, 7 of the 14 subjects were hospitalized at least once for rhabdomyolysis, elevated CPK levels, and dehydration, which was associated with illness and/or exercise. All received standard treatments of IV fluids and dextrose during those hospitalizations. One subject experienced a cardiac arrest and was resuscitated which resulted in physical and mental delays related to hypoxia.

Another subject was hospitalized for a tonsillectomy and another with a broken femur, as previously reported [14].

Discussion

These results show that propofol can be used safely in children with LCHAD or TFP deficiency for procedures of short duration that induce low post-procedural physical stress. These children were not physically ill at the time of the sedation and it is unknown if similar results would be seen if children with FAODs were sedated during episodes of metabolic decompensation, such as during rhabdomyolysis. Pre-sedation fasting guidelines were modified and children were also supplemented with intravenous dextrose during the procedure to decrease the risk of hypoglycemia (Table 1). The rapid recovery seen after the procedure further reduced the risk of hypoglycemia and other symptoms of LCHAD or TFP deficiency by allowing for quick resumption of oral intake. Propofol should be considered for use in similar procedures among patients with other FAODs such as carnitine palmitoyltransferase II (CPT2) deficiency, very-long-chain acyl-coenzyme A dehydrogenase (VLCAD) deficiency, medium-chain acyl-CoA (MCAD) deficiency or short-chain acyl-CoA dehydrogenase (SCAD) deficiency. Although this study was not conducted in children with those disorders, we would expect that children with other long-chain FAODs (CPT2 or VLCAD deficiency) would respond in a similar way to the LCHAD and TFP deficient subjects we studied. It is generally accepted that people with MCAD or SCAD deficiency tolerate more dietary fat and therefore dietary fat is not routinely restricted for these FAO disorders. The minimal amount of LCFA in propofol for routine sedation should be of less concern in these disorders. However, caution should still be exercised in the use of propofol in any of these disorders if patients are under metabolic stress. Although this study was done in children, adults with FAODs should theoretically also be able to tolerate short-term sedation with propofol, although no cases have been reported to date.

The recommendation to avoid general anesthesia for patients with an FAOD, was first mentioned by Wieser et al. in a review of CPT2 deficiency in 1993, but no reference was cited [15]. A case report was published in 2002 regarding perioperative management of a child with -VLCAD deficiency [7]. In this report, alternative anesthesia was provided to a 9-year-old child who underwent an elective minor surgical procedure (circumcision) because the authors hypothesized that propofol infusion syndrome presents with symptoms similar to those found in patients with VLCAD deficiency; bradydysrhythmias, rhabdomyolysis and metabolic acidosis. In addition to the concern about the similarity of symptoms, the authors remarked that free fatty acid levels can be elevated during minor surgical procedures (citing a study by Kleemann et al. conducted in healthy subjects) which they worried could cause a build-up of long-chain acylcarnitines in their VLCAD patient [7, 16]. Propofol was avoided in the sedation of another VLCAD child for placement of a gastrostomy tube with the authors citing similar concerns about the risk of overload with fatty acids [8]. Two other case reports have been published in children with long-chain FAODs, one in a child with LCHAD deficiency and the other in a child with TFP deficiency, in which alternate sedation was used for short-term sedation due to concerns about the use of propofol [9, 17]. Alternative anesthesia was also given to a child with SCAD deficiency for elective surgery [18]. The reason discussed for avoidance of propofol was not hypoglycemia, which is not a

usual characteristic of SCAD, but the potential for the build-up of toxic intermediates during surgery, particularly butyryl-CoA which could potentially lead to acidosis. MCAD deficiency has also been mentioned in two review articles on PRIS as a possible risk factor due to the similarity of symptoms during acute episodes of MCAD metabolic decompensation, but no case reports were cited [19, 20].

The similarity in the symptoms of PRIS and the symptoms of exacerbation of FAODs appears to be a major cause of concern and the primary reason for recommendations to avoid propofol for anesthesia in people with these rare genetic disorders. The exact cause of PRIS is not completely known but disruptions of energy metabolism have been suggested. Propofol is thought to inhibit intracellular energy production by inhibiting transportation of LCFAs in to the mitochondrion (caused by inhibition of carnitine palmitoyltransferase I) [4]. In addition, propofol has been shown to directly impair the flow of electrons down the mitochondrial respiratory electron transport chain [21], potentially through secondary inhibition of respiratory chain Complex II [22]. This is thought to disrupt adenosine triphosphate (ATP) production resulting in cellular energy deficiency and a build-up of toxic fatty acid intermediates during PRIS. Intercurrent critical illness is considered to be an essential prerequisite for the occurrence of PRIS; and receiving high dose sustained propofol infusion, along with steroids or catecholamines can be triggering factors [23]. The clinical manifestations of PRIS reported in the order of frequency from the FDAs MEDWATCH system include cardiac (43.7%), hypotension (34.0%), rhabdomyolysis (27.0 %), hepatic (24.1%), renal (23.1%), metabolic acidosis (20.1), hypoxia (17.5%), hyperthermia (11.6%), and dyslipidemia (5.3%) [24]. We are not aware of any cases of PRIS in a patient diagnosed with a FAOD.

PRIS mimics some of the symptoms found during exacerbations of FAODs. People with FAODs have deficient production of energy-yielding substrates such as acetyl CoA and ketone bodies which causes accumulation of free fatty acids and potentially toxic acyl-CoA intermediates [1]. When physiologically stressed by illness, infection, surgery or excessive exercise, people with LCHAD cannot effectively transport the long-chain fatty acids inside the mitochondria for metabolism and energy generation and this can result in a spectrum of symptoms similar to PRIS; including cardiomyopathy, rhabdomyolysis, and hepatic steatosis [1, 4]. These similarities in symptoms and the proposed mechanism of action of propofol in causing PRIS should result in caution with prescribing propofol to anyone with an FAOD who is ill or in a catabolic state; however, this does not rule out propofol for use in short term sedation in clinically stable healthy people with a FAOD.

It has been suggested that it may be possible to prevent PRIS in critically ill patients by maintaining an adequate carbohydrate supply and by limiting the lipid load by keeping the propofol infusion running at 4 mg/kg/hour or lower [25]. Providing intravenous dextrose during medical procedures and providing the minimum amount of propofol needed for the procedure may reduce the risk of negative effects in patients with LCHAD or TFP deficiency.

Conclusions

The successful sedation of these children shows that propofol can be used safely for short-duration procedures in patients with LCHAD or TFP deficiency. The amount of LCFAs provided by the propofol in these procedures was minimal (approximately 1.0% of daily energy needs) and no adverse effects were noted. However, no other clinical studies or case reports are available in which propofol has been used in this patient population and more research is needed to confirm the safety of this drug for sedation in clinical practice. Future research might include laboratory testing to measure acylcarnitines, free fatty acids, and CPK levels in addition to assessment of muscle pain after the procedure to better monitor tolerance. Caution should still be exercised before administering propofol in patients with FAODs requiring longer duration sedation or if a patient is experiencing a critical illness or metabolic decompensation related to a FAOD.

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Highlights

- The medication propofol, commonly used for anesthesia, has been avoided in patients with mitochondrial fatty acid oxidation disorders (FAODs) due to concerns that it contains long-chain fatty acids (LCFAs).
- The outcomes of 8 children with long-chain 3-hydroxyacyl CoA dehydrogenase (LCHAD) deficiency or trifunctional protein (TFP) deficiency who were repeatedly sedated for an electroretinogram (ERG) with propofol were examined.
- The LCFAs in the propofol provided only 1.0% of the average total daily energy needs.
- The sedation with propofol resulted in no adverse side effects and was safely used in this short duration procedure.

Table 1
Fluid Management Protocol for Short-Duration Sedation with Propofol

Sedation Timeline	Procedure
6 hours prior to sedation	Consumption of a light low-fat meal
4 hours prior to sedation	4-6 oz clear liquids; then nothing by mouth (NPO)
45 minutes prior to sedation	Begin intravenous (IV) infusion of 10% dextrose with ½ normal saline (D ₁₀ 1/2NS) at 1.5 times maintenance rate
During the procedure	Continue the IV infusion
15-30 minutes after end of sedation	Once awake, resume clear liquids orally and discontinue the IV

Table 2
Propofol Dosing Compared to Actual and Recommended Fat Intake

Subject	Age (yr)	3-Day Usual Energy Intake Average (kcal)	Predicted 10% Kcal as LCFA from Usual Intake (gm)	Actual LCFA Intake, Visit Average (gm)	Propofol Dose LCFA Intake (gm)	Propofol dose/Predicted LCFA Intake (%)
1	3	1143	12.70	10.77	1.66	13.07
1	4	1506	16.73	23.10	1.71	10.22
1	5	1263	14.03	16.77	1.75	12.47
1	6	1905	21.17	20.75	1.32	6.24
2	4	1593	17.70	14.20	1.19	6.74
2	5	1845	20.50	9.58	1.67	8.15
2	6	2512	27.92	28.31	1.62	5.80
2	7	2191	24.34	24.54	1.25	5.13
3	10	2046	22.73	32.24	2.90	12.76
3	10	2359	26.21	17.20	2.22	8.47
3	11	2655	29.50	28.83	3.00	10.17
3	12	2156	23.96	28.36	2.70	11.27
4	4	1357	15.08	13.21	3.05	20.23
4	5	1407	15.63	15.60	3.10	19.83
4	6	1615	17.94	28.00	3.07	17.11
4	8	1653	18.37	18.70	1.78	9.69
5	9m	608	6.75	2.77	1.10	16.29
5	1	991	11.01	11.40	1.07	9.72
5	2	1331	14.79	24.79	2.00	13.53
5	3	1375	15.28	33.91	2.20	14.40
6	3	1686	18.73	25.04	1.65	8.81
6	4	1646	18.29	23.00	1.18	6.45
6	5	1842	20.47	24.38	1.45	7.08
6	6	1951	21.68	42.69	1.50	6.92
6	7	2212	24.57	30.44	1.90	7.73
7	2	1510	16.77	20.21	1.70	10.14

Subject	Age (yr)	3-Day Usual Energy Intake Average (kcal)	Predicted 10% Kcal as LCFA from Usual Intake (gm)	Actual LCFA Intake, Visit Average (gm)	Propofol Dose LCFA Intake (gm)	Propofol dose/Predicted LCFA Intake (%)
7	3	1705	18.95	28.61	2.00	10.56
7	5	2082	23.14	43.83	1.21	5.23
7	6	2100	23.33	27.90	1.43	6.13
8	1	797	8.85	10.50	1.20	13.56
8	2	863	9.58	5.66	1.85	19.30
8	3	1197	13.30	41.59	1.20	9.02
Average:		1659	18.44	22.72	1.83	10.69