



# WHAT IS CITRIN DEFICIENCY

## AND HOW WE CAN HELP

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### Citrin deficiency is...

Citrin deficiency is a hereditary condition which has some difficulty in metabolism in liver including energy production from carbohydrate and detoxication of alcohol.

It is usually manageable by diet and no activity restriction is necessary.

### Presentations

Typically citrin deficiency patients do not eat much food containing high carbohydrate (rice, bread, sweets, etc.) but instead take a lot of food containing fat and protein (milk, cheese, meat, fried food, etc.) to compensate the energy supply shortage.

Some patients may also experience failure to thrive, fatigue, abdominal pain, dizziness, and convulsion, depends on the physical and nutritional condition.

It may sometimes take patients a longer time to fully recover from a general cold if they cannot eat enough during sickness.

Dietary imbalance especially from eating too much carbohydrates/sweets can lead to sickness.

### How patients cope

The best thing the patients can do to cope with the condition is the diet management.

There is a natural reaction of patients to meet their needs by selecting and avoiding certain foods. This is a key for them to stay energetic and healthy.

There is also a need to eat frequently to have the continuous flow of energy intake.

### Tips to support patients

- ★ Understand that their food preference is a body reaction for their well-being and not something to be corrected.
- ★ Help them eat what they wish to eat in the amount they wish.
- ★ Make snacking time available at school.
- ★ Inform doctors that the patient has citrin deficiency in the case of an emergency.
- ★ No alcohol.

Please read the back of this leaflet for details.  
Thank you very much.

# Support you can provide

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## 1. School meals

### Serve more meat and dairy products, and less rice/bread/pasta

In schools where lunch is provided, please be informed that the portion of each dish may need to be adjusted to fit the needs of patients. Typically, having more meat and dairy products (fat/protein) and consuming less rice/bread/pasta (carbohydrate) will be the right balance for them.

Please allow the patients to decide on what and how much they want to eat so their dietary needs on the day can be naturally fulfilled.

It is different from being selfish or picky.

It is also different from allergy.

Too much carbohydrates and sweets tend to make the patients feel sick, but they may favor consuming small amounts of carbs and sweets if it is necessary.

It may be more preferable if understanding from their classmates can be obtained in advance.

### Some seasonings may not be suitable

Some seasonings may taste sweet to the patients which may cause them to avoid consuming the dish.

If there is no alternative dish they can choose from, it may be necessary for them to bring lunch from home.

## 2. Supplementary food and drinks

### Frequent energy intake throughout the day is necessary

Energy supplementation between meals is suggested by doctors to respond to the metabolic conditions. Parents may ask the school to keep supplementary food/drinks in a cool place if it is necessary.

## 3. Snacks

### Typical children snacks may not be their favorite

Children with citrin deficiency do not enjoy sweets, candies, or juice as much as other kids. But especially younger children may take more than what their body can take in a social environment. Please pay a kind attention so they do not eat too much sugar/carbs.

### List of typical likes/dislikes:

(It may depend on what was consumed in previous meals or their condition.)

Likes: whole milk, cheese, yogurt, sausage, bacon, beef jerky, salmon, tuna, chicken, tofu, egg, nuts, butter

Dislikes: sticky rice, bread, buns, jam, sweet beans, honey, candy

## 4. When patients get sick

### Inform the doctors

When patients say they feel sick, please allow them to take what they wish to eat/drink regardless of the usual medical protocol.

In case they need medical attention, please inform doctors that the patient has citrin deficiency. The use of GLYCEOL® is prohibited and high calorie infusion with glucose is not recommended as they may further exacerbate the condition of the patients. There is no issue with general infusion of glucose to treat hypoglycemia, diarrhea, or vomiting. But it is advisable to let patients consume the food they prefer after their appetite comes back for faster recovery.

## 5. No alcohol

Alcohol consumption may put the patients life at risk. No alcohol, please.

Fully evaporated cooking wine may not be a problem but it may be safer to avoid it.