Clinical Correlation

Usman Sumo Friend Tambunan
Arli Aditya Parikesit
Geraldine Marcella L

Bioinformatics Group
Department of Chemistry
Faculty of Mathematics and Science
University of Indonesia

- Diabetes Mellitus
- Galactosemia
- Protein Malnutrition
- Reye's Syndrome
- Obesity

Protein Malnutrition

Definition

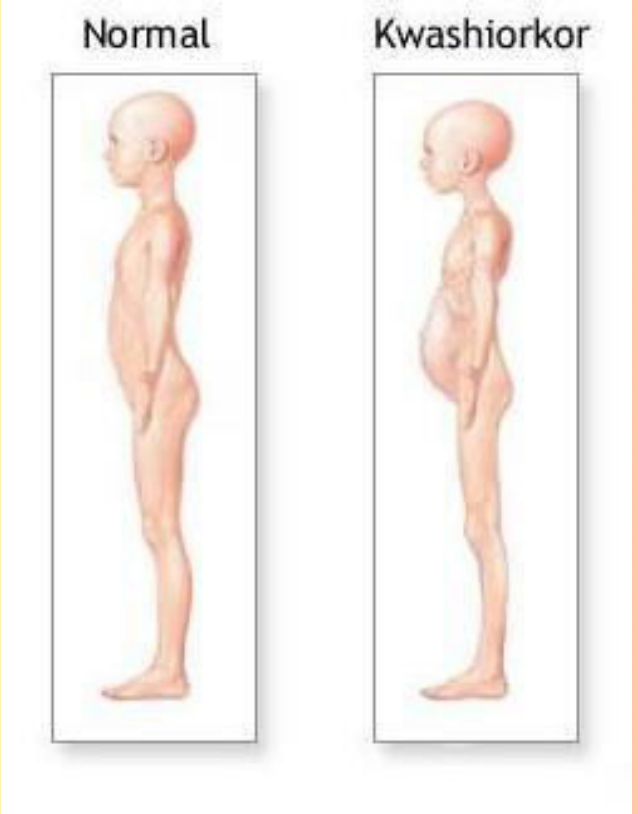
- Malnutrition is the result of extended inadequate intake or severe illness on body composition and function and affect all system of the body.

Two types of malnutrition:

- Protein malnutrition, refers to kwashiorkor.
- Protein Calorie (Energy) Malnutrition, refers to marasmus

KWASHIORKOR

- Usually occur in children aged 1-4 years although can be occurred in older children or adults.
- Symptoms: swollen abdomen (known as pot belly), edema, failure to gain weight and failure of linear growth, hair changes, depigmented skin.
- Long term impact is on the physical and mental development. In severe cases may lead to the death.



Protein Correlation

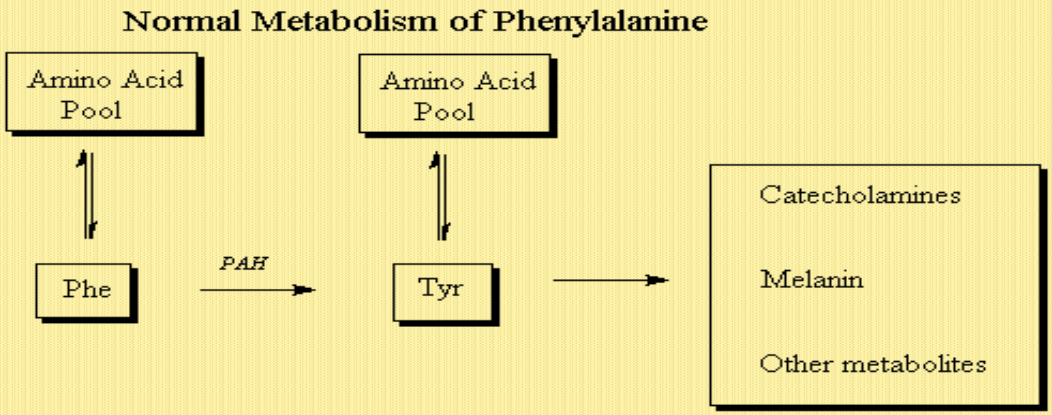
- Protein is composed from amino acids.
- Amino acid can be classified into two types, which are: essential amino acid and nonessential amino acid.
- Essential amino acid cannot be produced in human body, it supplied from food we eat.
- Kwashiorkor can be occurred when there is no food supply to body or the food doesn't provide dietary protein needed (essential amino acid)

Nutritionally Essential	Nutritionally Nonessential
Arginine[1]	Alanine
Histidine	Asparagine
Isoleucine	Aspartate
Leucine	Cysteine
Lysine	Glutamate
Methionine	Glutamine
Phenylalanine	Glycine
Threonine	Hydroxyproline[2]
Tryptophan	Hydroxylysine[2]
Valine	Proline
	Serine
	Tyrosine

[1]"Nutritionally semiessential." Synthesized at rates inadequate to support growth of children.
[2]Not necessary for protein synthesis but formed during post-translational processing of collagen.

- Essential amino acids are very important because they are precursor for many others metabolism pathways.
- For example: Phenylalanine
- In humans, melanin is the primary determinant of human skin color and also found in hair

Protein source	Limiting amino acid
Wheat	Lysine
Rice	lysine
Legumes	tryptophan
Maize	lysine and tryptophan
Beef	phenylalanine (or tyrosine)

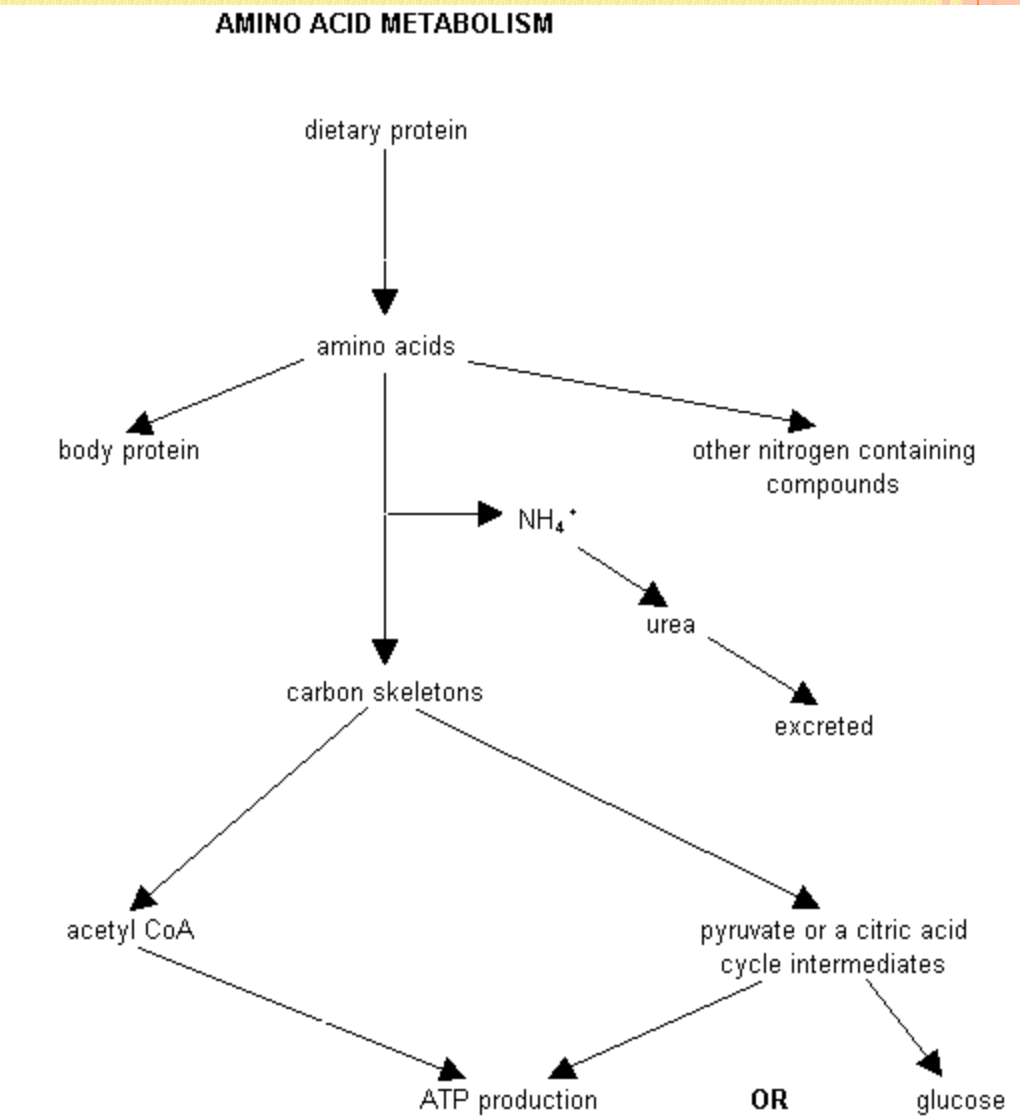


Possible Causes of the Development of Kwashiorkor

- The protein deficiency, in combination with energy and micronutrient deficiency, is not the key factor of the development of kwashiorkor.
- One important factor in development of kwashiorkor are aflatoxin poisoning.
- Aflatoxin can cause mycotoxin. Aflatoxins are naturally produced by aspergillus, a fungus.
- After aflatoxin enter the body, it will be metabolized in liver with enzyme cyttochrome p450 help produce reactive intermediate aflatoxin M1, the final product is epoxide.
- This epoxide will damage liver DNA.

The correlation between damage liver DNA and development of kwashiorkor

- Protein catabolism involves urea cycle that take place in liver. The product is urea or uric acid and will be removed from body.
- Because of the liver is damage and cannot do the urea cycle, it cannot converted amonia compound to urea and cause the accumulation of amonia, resulting the failure damage.
- Liver is the place to produced many serum proteins. With the damage of liver, symptoms of kwashiorkor can be clearly explained.



Treatment of Kwashiorkor

- Treatment of kwashiorkor begins with rehydration. Subsequent increase in food intake must proceed slowly, beginning with carbohydrates followed by protein supplementation.

Reye's Syndrome

- Reye's syndrome is a potentially fatal disease that affect all organs of body, especially liver and brain.
- In almost case, reye's syndrome is associated with viral infection. Such as: influenza, cold, or chicken pox.
- The mechanism of how Reye's syndrome occurs is still unknown.

Symptoms and Sign

Reye's syndrome progress through five stages, which are:

Stage I

- heavy vomiting
- Confusion
- Nightmares

Stage II

- Fatty liver (found by biopsy)
- Hyperventilation
- Hyperactive reflexes

Stage III

- Possible coma
- Possible cerebral edema
- Rarely, respiratory arrest

Stage IV

- Deepening coma
- Large pupils with minimal response to light
- Minimal but still present hepatic dysfunction

Stage V

- Very rapid onset following stage IV
- Deep coma
- Seizures
- Respiratory failure
- Flaccidity
- Extremely high blood ammonia (above 300mg per 100mL of blood)
- Death

Causes

- Reye's syndrome causes fatty accumulation in the organs of the body, especially in liver.
- In the brain, it causes accumulation of fluid (edema), and rising the pressure.
- This pressure squeezes blood vessels, preventing blood from entering the brain.
- Untreated, this pressure increase leads to brain damage and death.

Test

The following tests may be used to diagnose Reye syndrome:

- Blood chemistry tests show low blood sugar (glucose) levels
- Liver function tests show higher than normal levels of liver enzymes
- Serum ammonia test may be higher than normal
- Liver biopsy
- Spinal tap
- Head CT or head MRI scan help rule out other causes of mental status changes

Conclusion

- Kwashiorkor is one of the more severe forms of protein malnutrition and is caused by inadequate protein intake.
- The development of kwashiorkor can lead to damage liver DNA.
- Reye's syndrome is a fatal disease that can damage organs of body, especially liver and brain
- The mechanism of reye's syndrome remains unknown.

References

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