# Chapter 6
# Histidine and branch chain amino acid

## Histidine

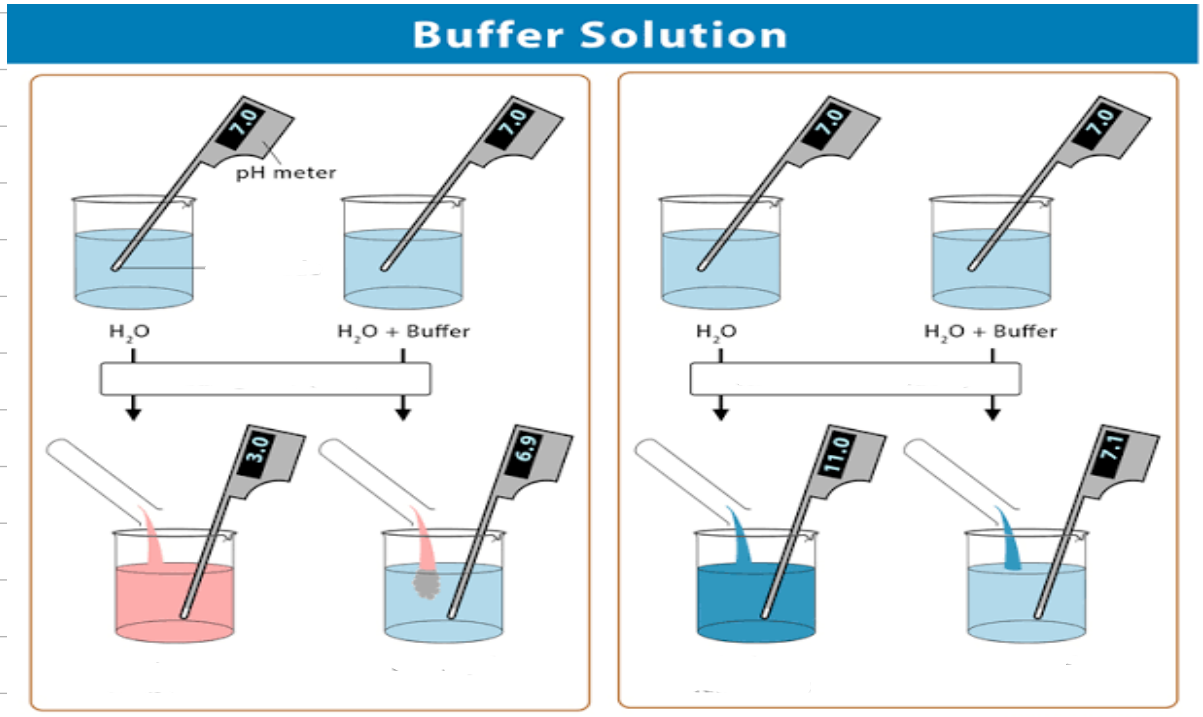
Essential / Non-essential

Aromatic / Non aromatic

Water soluble / water insoluble

Histidine structure Contains …… ring.

## Concept of buffer solution




Histindine has …… buffer capacity among all AAs.

## Function of histidine

Histidine ⟶

PL P - pyridoxal phoshphate

☆ **Pharma connection -**

| | Location | Function | Histamine rec. blocker |
|---|---|---|---|
| H1 receptor | | | |
| H2 receptor | | | |
| H3 receptor | | | |

**Histdine metabolism**

Histdine → [ ] → [ ]

↓

[ ]

**Clinical co-relation**

Vit. B9 (……) deficiency leads to. -

-



30 year old female comes to OPD . giving history of NTD in last child 4years back ,Now she is again planning for pregnancy. She want to make sure she is having adeqaute vit B9 level , what to be done next ?

Histidine

↓

FIGLU

Vit …

↓

Glutamic acid

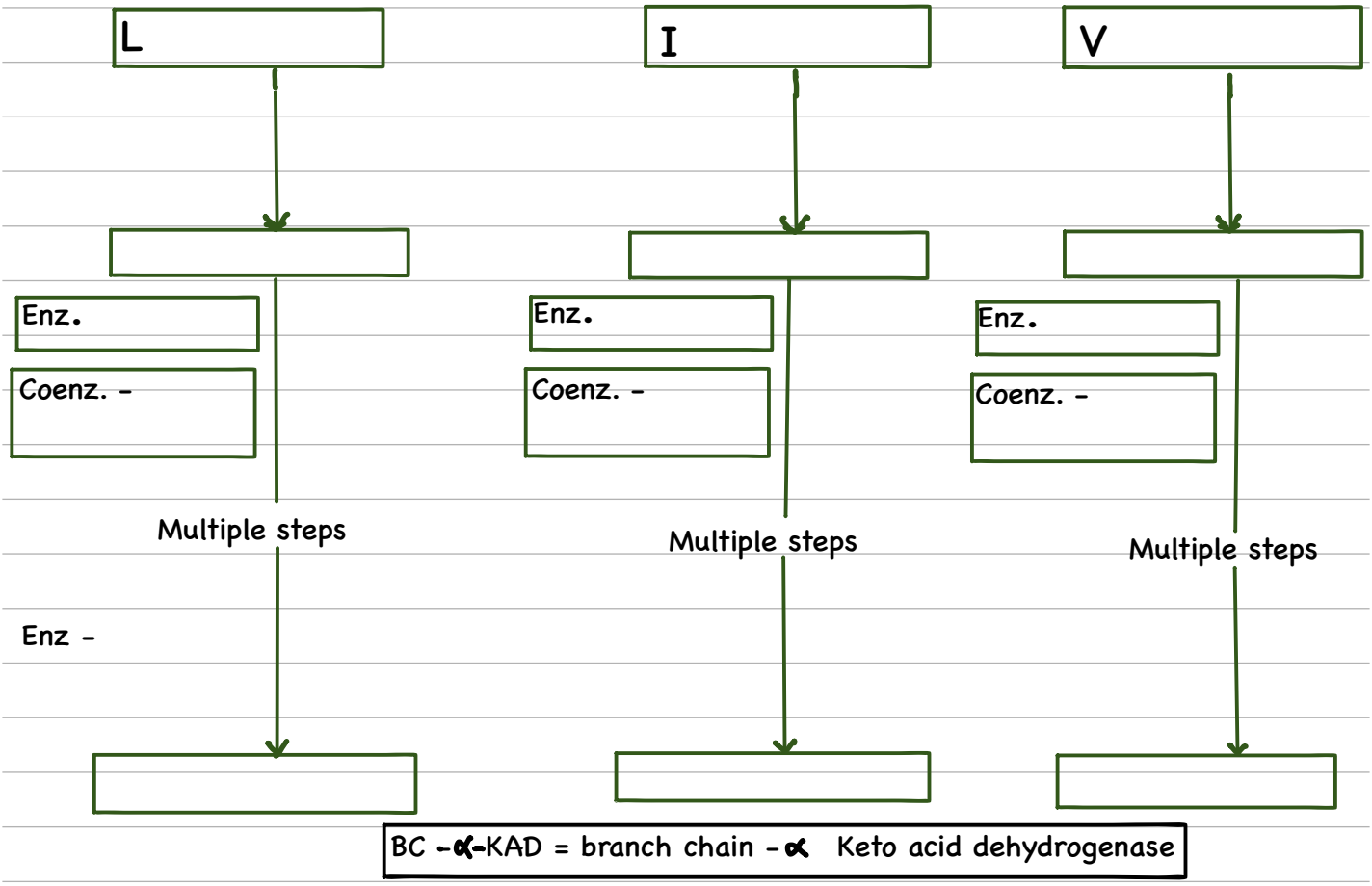
Ans - give a i/v injection of …… AA , if there is deficiency of vit B9 , ..... will increase is the body ,this is called as histidine loading test or histidine challange test.

## Branch chain AA

Eg.

Most ketogenic AA -

## Metabolism of branch chain AA




## Clinical co-relation

### MSUD

Deficiency of branch chain. α- Keto-acid dehydrogenase leads to ……, in this urine smells like ……the main reaction defective in MSUD is……

Inv in MUSD.-

Fecl3 can also be done in …… …… ……

## Isovaleric acidemia

Is a defect of …… metabolism.

is due to deficiency of

Urine smells like

## Summary

☆ Maximum buffer capacity among all AA -

☆ Cetrizine is …… receptor blocker

☆ Ranitdine is …… receptor blocker

☆ Histidine loading test is done to detect deficiency of vit. ……

& if there is deficiency - ……will be increased.

☆ Match the following

| | |
|---|---|
| A) mousy smell | 1) alkaptonuria |
| B) swimming pool like smell | 2) MSUD |
| C) burnt sugar smell | 3) isovaleric acidemia |
| D) sweaty feet smell | 4) tyrosinemia |
| E) darkening of urine on standing | 5) PKU |

A……b……c……d……e……