

PKU and MSUD Screening Methods

Phenylketonurea (PKU) and Maple Syrup Urine Disease (MSUD) are amino acid disorders – rare inherited conditions that prevent people from digesting certain amino acids in food proteins. Untreated, these serious conditions lead to mental and physical disability and even death. PKU and MSUD are part of newborn screening program and are detected and monitored through Amino Acids analysis of blood samples.

Pickering Laboratories offers full range of products for post-column Amino Acids analysis of physiological fluids. The same column and buffers can be used for running full amino acid profile as well as accelerated screening methods for specific medical conditions. This application note describes PKU and MSUD screening methods for Pickering 0354100T and 0354675T columns.

PKU MSUD Screening Method for Column 0354100T

Analytical Conditions

Column: High-efficiency Lithium cation-exchange column,

4x100 mm, Catalog Number 0354100T

Guard: Cation-exchange GARD™,

Catalog Number 1700-3102

Flow Rate: 0.4 mL/min

Mobile Phase: 1700-1125, Li265, Li375, RG003

| | Post-Col | lumn | Cond | itions |
|--|----------|------|------|--------|
|--|----------|------|------|--------|

Post-Column System: Onyx PCX or Pinnacle PCX

Reactor Volume: 0.5 mL Reactor Temperature: 130 $^{\circ}$ C

Reagent: Trione Ninhydrin reagent
Reagent Flow Rate: 0.3 mL/min

Detection: UV/VIS 570 nm for primary amino acids

440 nm for secondary amino acids

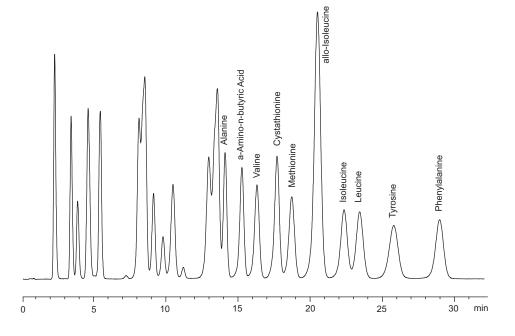
Run Time: 33 min

Equilibration Time: 10 min

Contact Pickering Laboratories for information about full amino acid profile method for this column.

| Column Oven Program | | | |
|---------------------|---------|--|--|
| Time | Temp °C | | |
| 0 | 50 | | |
| 20 | 61 | | |
| 30 | 61 | | |
| 30.1 | 50 | | |

| HPLC Program | | | | | |
|--------------|-------------|---------|---------|---------|--|
| Time | 1700-1125 % | Li365 % | Li375 % | RG003 % | |
| 0 | 70 | 30 | 0 | 0 | |
| 20 | 0 | 100 | 0 | 0 | |
| 30 | 0 | 0 | 100 | 0 | |
| 30.1 | 0 | 0 | 0 | 100 | |
| 33 | 0 | 0 | 0 | 100 | |
| 33.1 | 70 | 30 | 0 | 0 | |



PKU MSUD Screening Method for Column 0354675T

Analytical Conditions

Column: High-efficiency Lithium cation-exchange column,

4.6x75 mm, Catalog Number 0354675T

Guard: Cation-exchange GARD™,

Catalog Number 1700-3102

Flow Rate: 0.55 mL/min

Mobile Phase: 1700-1125, Li265, Li375, RG003

Post-Column Conditions

Post-Column System: Onyx PCX or Pinnacle PCX

Reactor Volume: 0.5 mL
Reactor Temperature: 130 °C
Reagent: Trione Ninhydrin reagent
Reagent Flow Rate: 0.5 mL/min

Detection: UV/VIS 570 nm for primary amino acids

440 nm for secondary amino acids

Run Time: 16 min

Equilibration Time: 10 min

Contact Pickering Laboratories for information about full amino acid profile method for this column.

| Column Oven Program | | | |
|---------------------|---------|--|--|
| Time | Temp °C | | |
| 0 | 50 | | |
| 14 | 61 | | |
| 15 | 50 | | |
| 30.1 | 50 | | |

| HPLC Program | | | | |
|--------------|-------------|---------|---------|---------|
| Time | 1700-1125 % | Li365 % | Li375 % | RG003 % |
| 0 | 60 | 40 | 0 | 0 |
| 9 | 0 | 100 | 0 | 0 |
| 14 | 0 | 30 | 70 | 0 |
| 14.1 | 0 | 0 | 0 | 100 |
| 16 | 0 | 0 | 0 | 100 |
| 16.1 | 60 | 40 | 0 | 0 |

