Lab Dept:	**Chemistry**
Test Name:	**PIPECOLIC ACID, SERUM**

General Information

Lab Order Codes:	PIPE
Synonyms:	N/A
CPT Codes:	82542 – Column chromatography, includes mass spectrometry, if performed, non-drug analytes, not elsewhere specified, qualitative or quantitative, each specimen
Test Includes:	Pipecolic Acid reported in nmol/mL.

Logistics

Test Indications:	Differential diagnosis between disorders of peroxisomal biogenesis and disorders with loss of a single peroxisomal function.
Lab Testing Sections:	Chemistry – Sendouts
Referred to:	Mayo Clinic Laboratories (MML Test: PIPA)
Phone Numbers:	MIN Lab: 612-813-6280 STP Lab: 651-220-6550
Test Availability:	Daily, 24 hours
Turnaround Time:	2 - 9 days
Special Instructions:	Fasting is recommended. See Patient Preparation

Specimen

Specimen Type:	Blood
Container:	SST (Marble, gold or red top tube) Acceptable: Red top no gel
Draw Volume:	4.5 mL (Minimum: 3 mL) blood
Processed Volume:	1.5 mL (Minimum: 1 mL) serum

Collection: Routine venipuncture

Special Processing: Lab Staff: Centrifuge specimen, remove serum aliquot into a screw-capped round bottom plastic vial. Store and ship at frozen temperatures. Forward promptly.

Specimen stable frozen (preferred) for 94 days, refrigerated for 14 days.

Patient Preparation: Patient should fast 12 hours or more, infants and small children just before next feeding.

Sample Rejection: Warm; cold refreeze; mislabeled or unlabeled specimens

Interpretive

Reference Range:

Age	Range (nmol/mL)
<6 months:	<or =6.0 nmol/mL
6 months to <1 year:	<or =5.9 nmol/mL
1 – 17 years:	<or =4.3 nmol/mL
>or=18 years:	<or =7.4 nmol/mL

Interpretation: Elevated pipecolic acid levels are seen in disorders of peroxismal biogenesis; normal levels are seen in disorders with loss of a single peroxisomal function.

Abnormal levels of pipecolic acid should be interpreted together with the results of other biochemical markers of peroxisomal disorders, such as plasma C(22)-C(26) very long chain fatty acids, phytanic acid, pristanic acid, RBC plamalogens, and bile acid intermediates.

Critical Values: N/A

Limitations: Newborn with disorders of peroxisomal biogenesis often have normal levels of pipecolic acid which increase with age.

Abnormal results may reflect either prematurity or nongenetic liver and/or renal disease.

Vigabatrin interferes with pipecolic acid determination.

Methylmalonic acid interferes with pipecolic acid determination.

Methodology: Gas Chromatography-Mass Spectrometry (GC-MS).

References: Mayo Clinic Laboratories (December 2023)

Updates:	5/8/2012: Reference range updates. 1/26/2016: CPT update 12/1/2023: Updated TAT, added stability, added interfering drugs.