

Amino Acid Pot Pourri

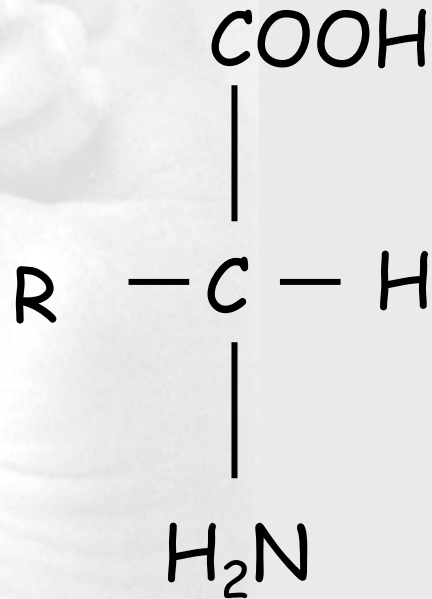
Dr Mick Henderson

*Department of Clinical Biochemistry
and Immunology
St James's University Hospital, Leeds*

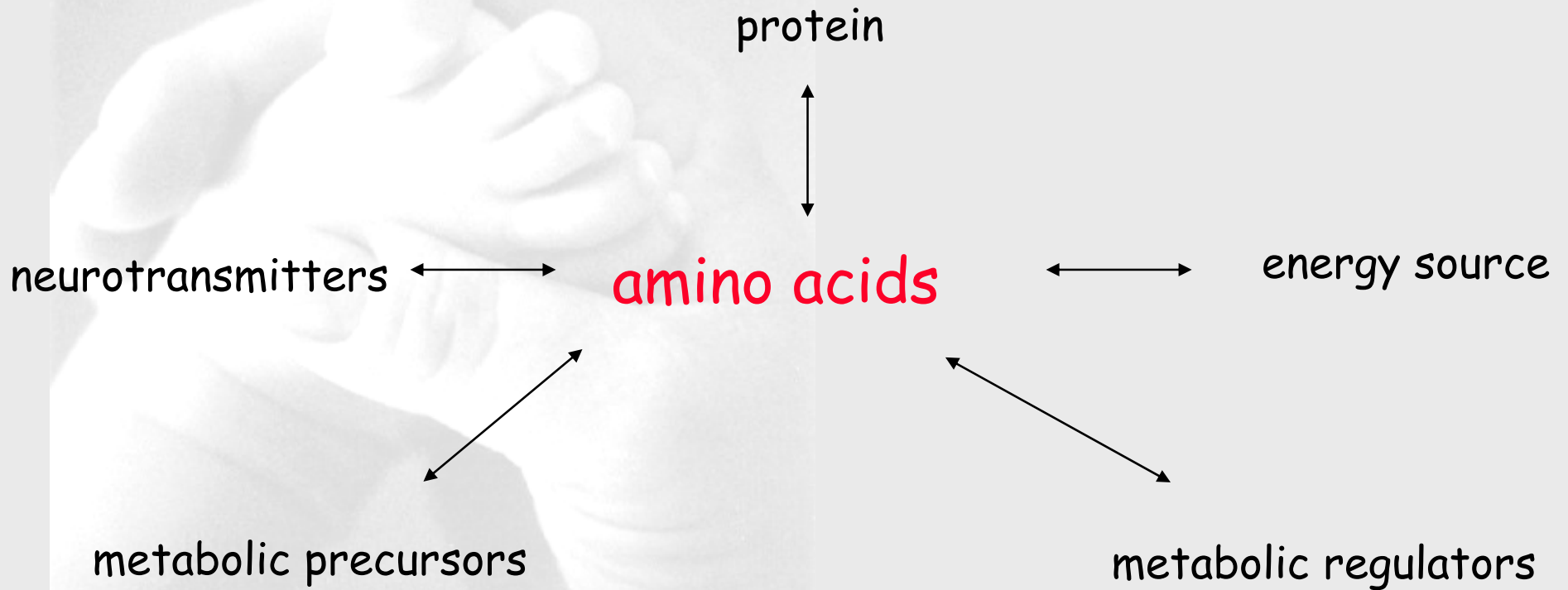


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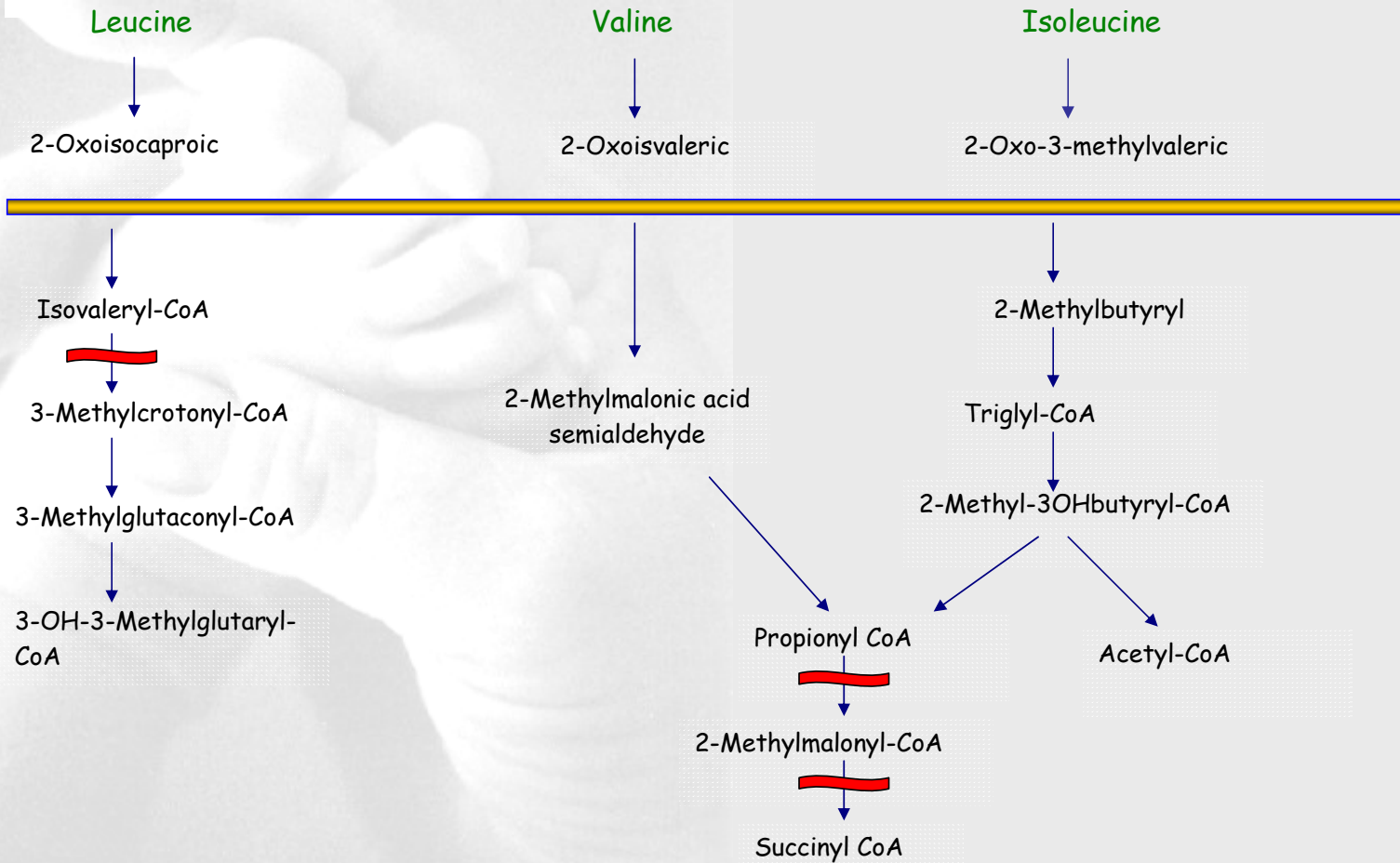
Amino acids



What use are amino acids?



Branched chain amino acid catabolism



Essential amino acids

- Histidine, isoleucine, leucine, lysine, methionine, phenylalanine, threonine, tryptophan, valine

conditionally essential for neonates

- Cysteine, tyrosine, taurine, arginine and glycine



Difficult to emulate ex utero

Nutritional monitoring

Plasma profiles of limited value

Affected by;

- quantity and quality of protein ingested or TPN
- time of samples relative to feeds
- energy supply
- growth rate
- relative maturation of enzymes and organs e.g. liver
- action of insulin, distribution between ICF and ECF

Most meaningful data has come from stable isotope studies

- Flux thro transsulfuration pathway inadequate for cys requirement

Other factors

- Protein sparing effect of
 - adequate calorific intake
 - appropriate balance of aa in neonatal feeds

tyrosine and cysteine missing from most PN preps
- Preference of neonatal brain for ketones
- Ketones as source of amino acids
 - particularly within the developing brain

AA Morris, JIMD, 2005, 28; 109-121

Microsoft Internet Explorer window: **Nutrient Requirements For Preterm Infant Formulas -- Klein 132 (6): 1395 -- Journal of Nutritio**

Address: http://www.nutrition.org/cgi/content/abstract/132/6/1395?ijkey=e3c7c6ffbf409d139fe1fe82a8bae209ae7d1ab&keytype2=tf_ipsecsha

nutrition.org Diets Inc. Experimental Diets For Laboratory Animals

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Supplement: Nutrient Requirements For Preterm Infant Formulas

Nutrient Requirements For Preterm Infant Formulas^{1,2,3}

Catherine J. Klein, Editor⁴

Life Sciences Research Office, 9650 Rockville Pike, Bethesda, Maryland 20814

Achieving appropriate growth and nutrient accretion of preterm and low birth weight (LBW) infants is often difficult during hospitalization because of metabolic and gastrointestinal immaturity and other complicating medical conditions. Advances in the care of preterm-LBW infants, including improved nutrition, have reduced mortality rates for these infants from 9.6 to 6.2% from 1983 to 1997. The Food and Drug Administration (FDA) has responsibility for ensuring the safety and nutritional quality of infant formulas based on current scientific knowledge. Consequently, under FDA contract, an ad hoc Expert Panel was convened by the Life Sciences Research Office of the American Society for Nutritional Sciences to make recommendations for the nutrient content of formulas for preterm-LBW infants based on current scientific knowledge and expert opinion. Recommendations were developed from different criteria than that used for recommendations for term infant formula. To ensure nutrient adequacy, the Panel considered intrauterine accretion rate, organ development, factorial estimates of requirements, nutrient interactions and supplemental feeding studies. Consideration was also given to long-term developmental outcome. Some recommendations were based on current use in domestic preterm formula. Included were recommendations for nutrients not required in formula for term infants such as lactose and arginine. Recommendations, examples, and sample calculations were based on a 1000 g preterm infant consuming 120 kcal/kg and 150 mL/d of an 810 kcal/L formula. A summary of recommendations for energy and 45 nutrient components of enteral formulas for preterm-LBW infants are presented. Recommendations for five

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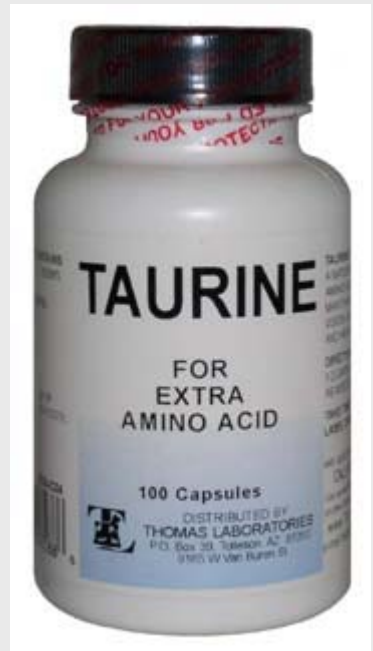
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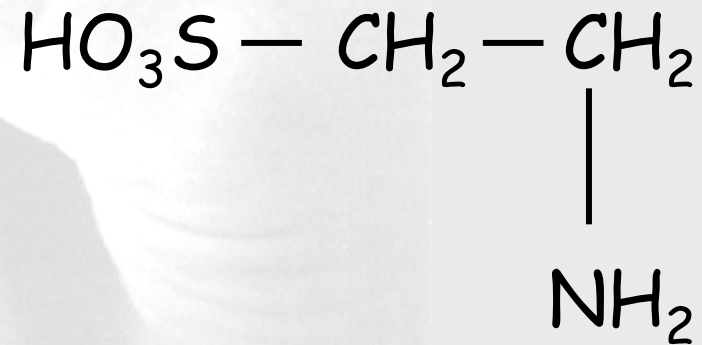
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Taurine



Taurine

an honorary amino acid!



Cats

Essential for



And babies



Taurine

Vital for:

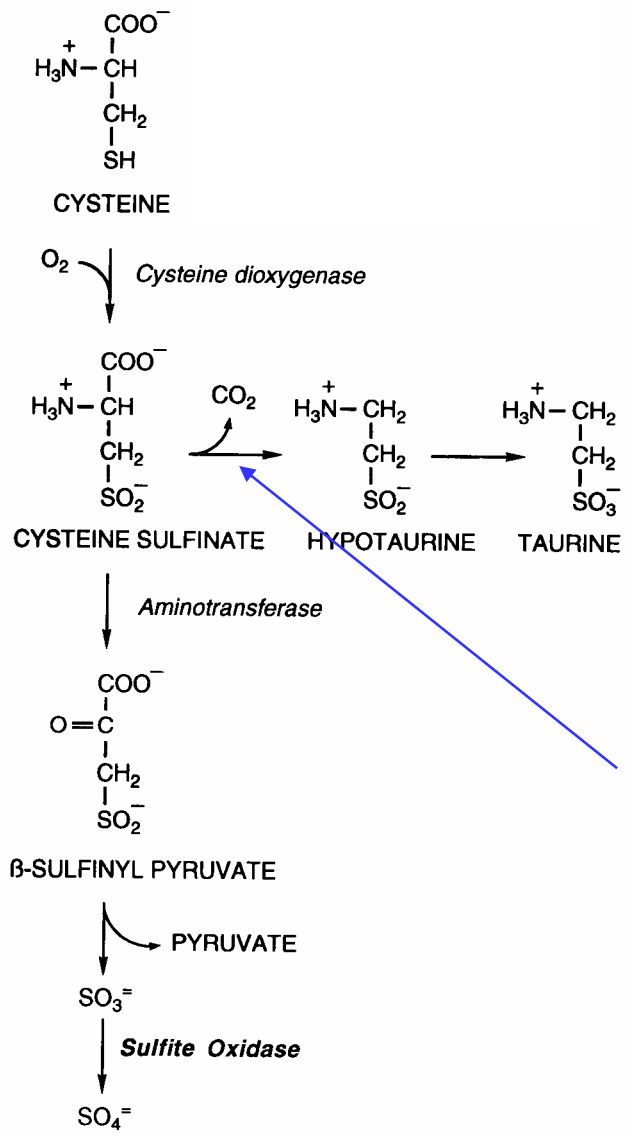
- Bile salt formation
- Vision
- Growth

Made from methionine and
cysteine

- Neonates have low CSAD activity
- Breast milk is rich in taurine

Taurine designated 'essential nutrient' by the FDA Expert Panel, 2002

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Cysteine sulfinic acid
decarboxylase

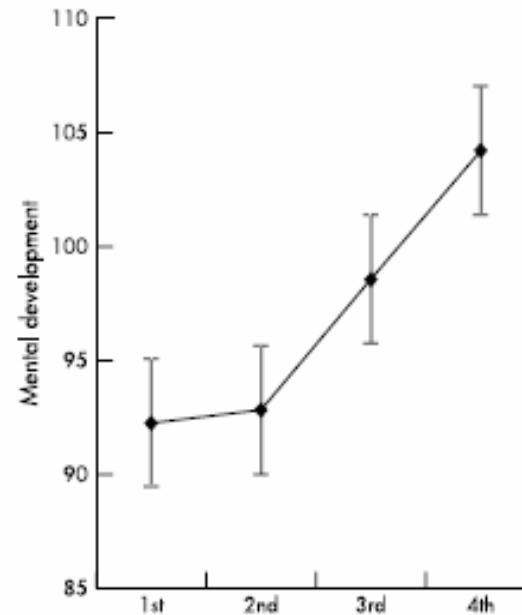
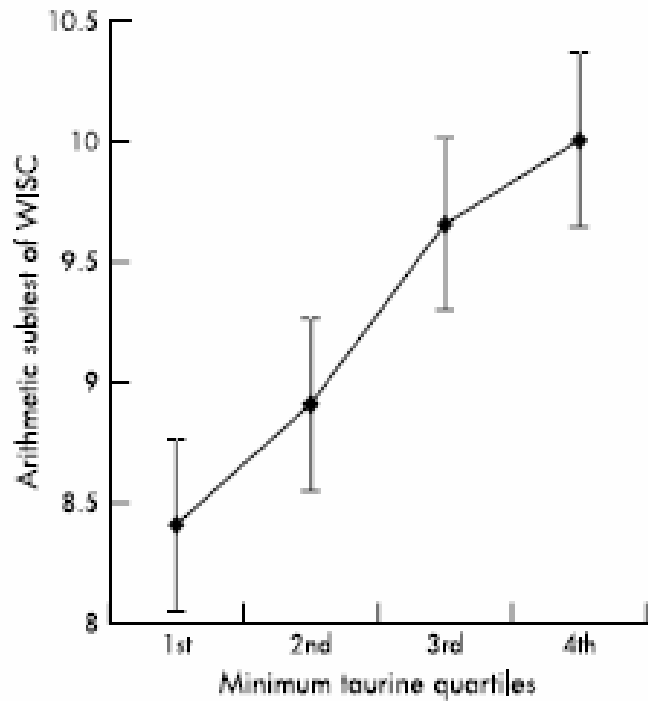
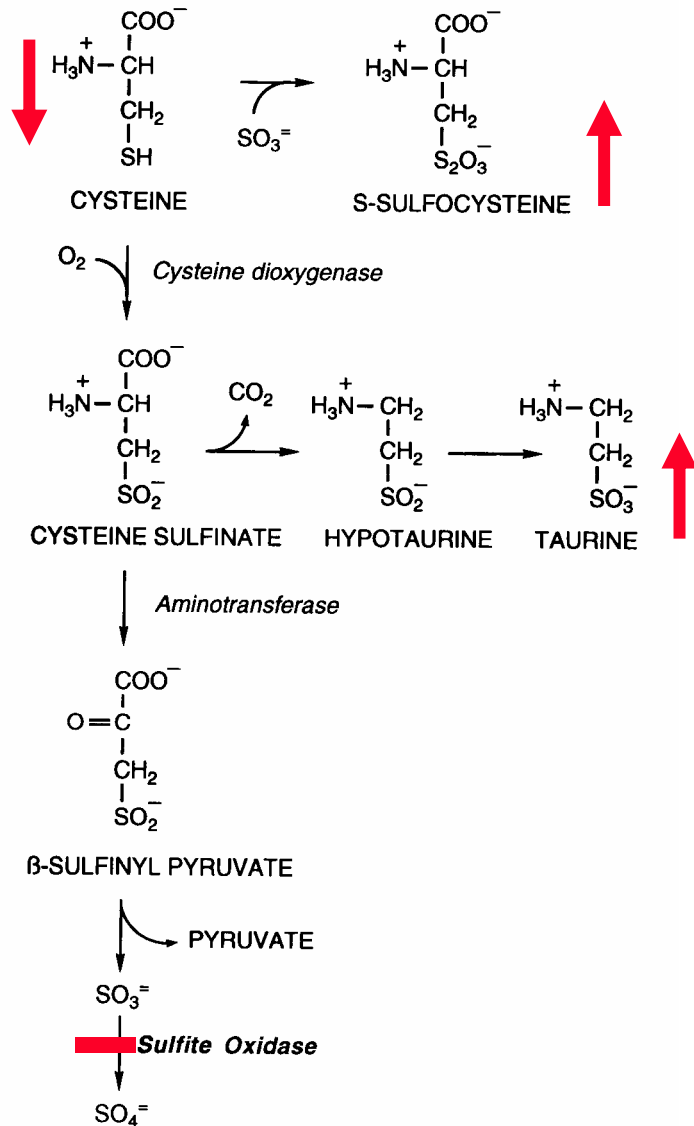


Figure 1 Bayley mental development index at 18 months, arithmetic subtest of WISC-R at 7 years, and minimum neonatal plasma taurine concentration ($\mu\text{mol/l}$). Taurine, 1st quartile, 20–43; 2nd quartile, 44–55; 3rd quartile, 56–67; 4th quartile, 68–180. Mental development index, mean (SE) 97 (2). Arithmetic score, mean (SE) 9 (0.3).

Wharton BA et al Arch Dis Child Fetal Neonatal Ed 2004;89:F497-F498.



Sulfite Oxidase Deficiency

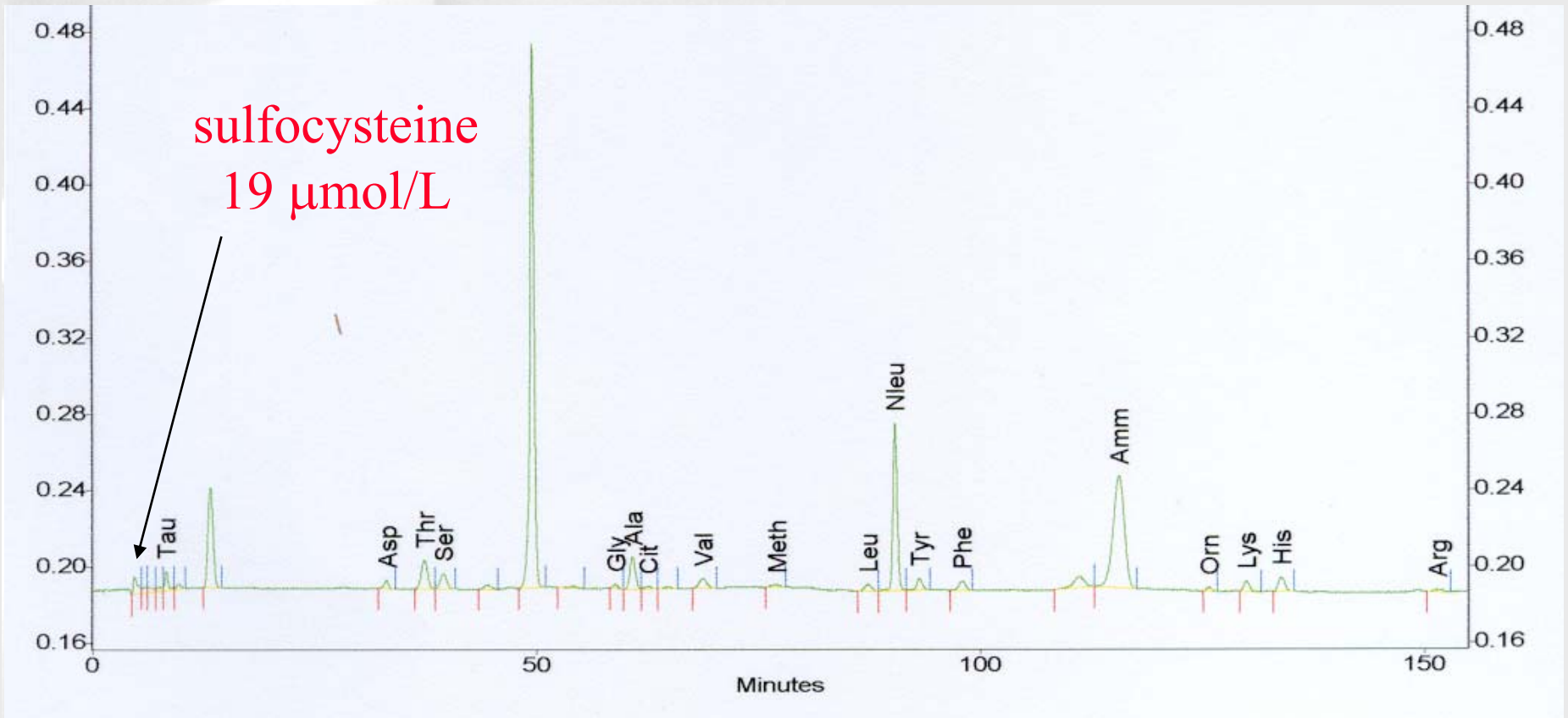
Johnson and Wadman,
7th Edition, Scriver, MMBID

Female baby

- unrelated parents
- term baby, no recorded neonatal problems
- severe persistent fitting from day 2
- died at 3 weeks

- early apnoea, lactate 8 mmol/L
- no evidence of hyperammonaemia, hypoglycaemia
- urine organic acids & blood acyl carnitines: NAD
- amino acids.....

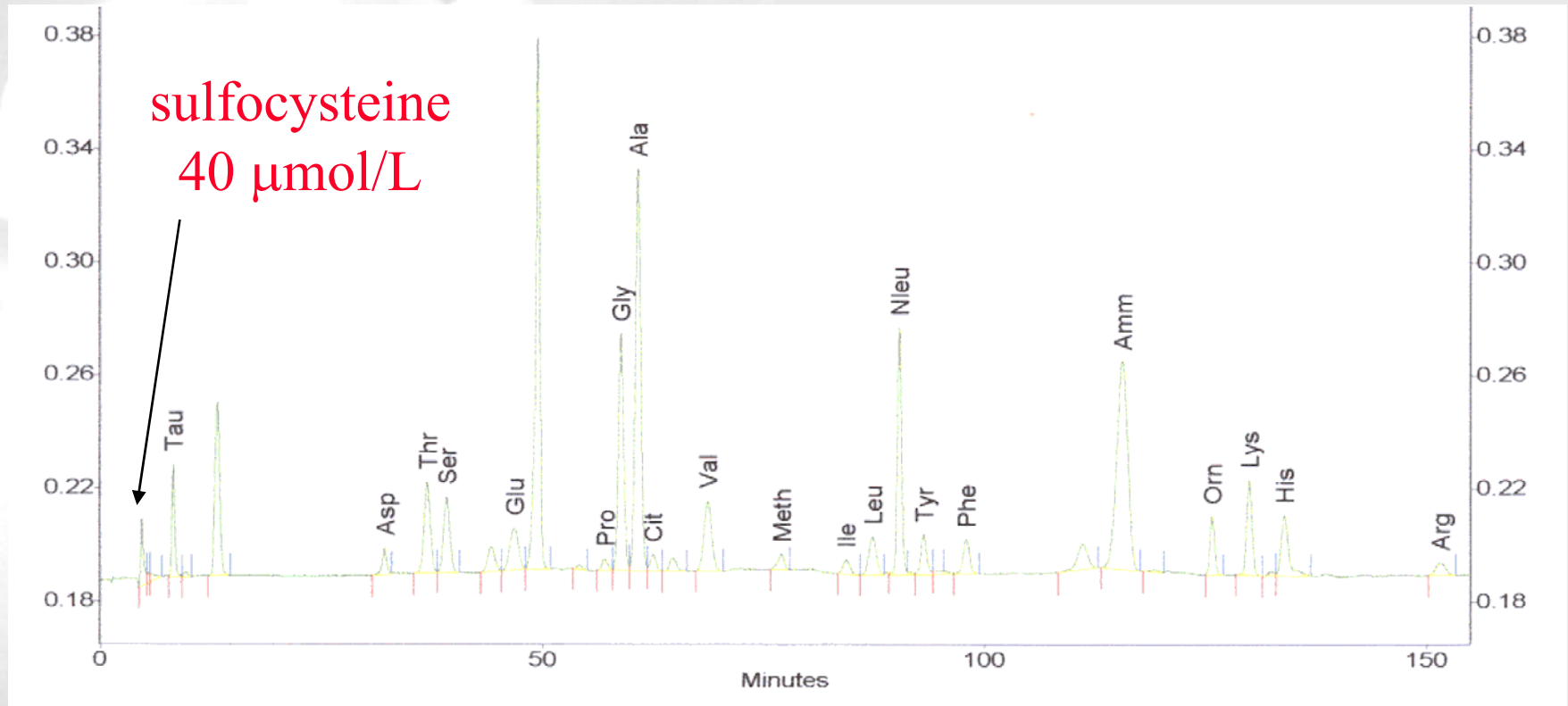
CSF amino acids



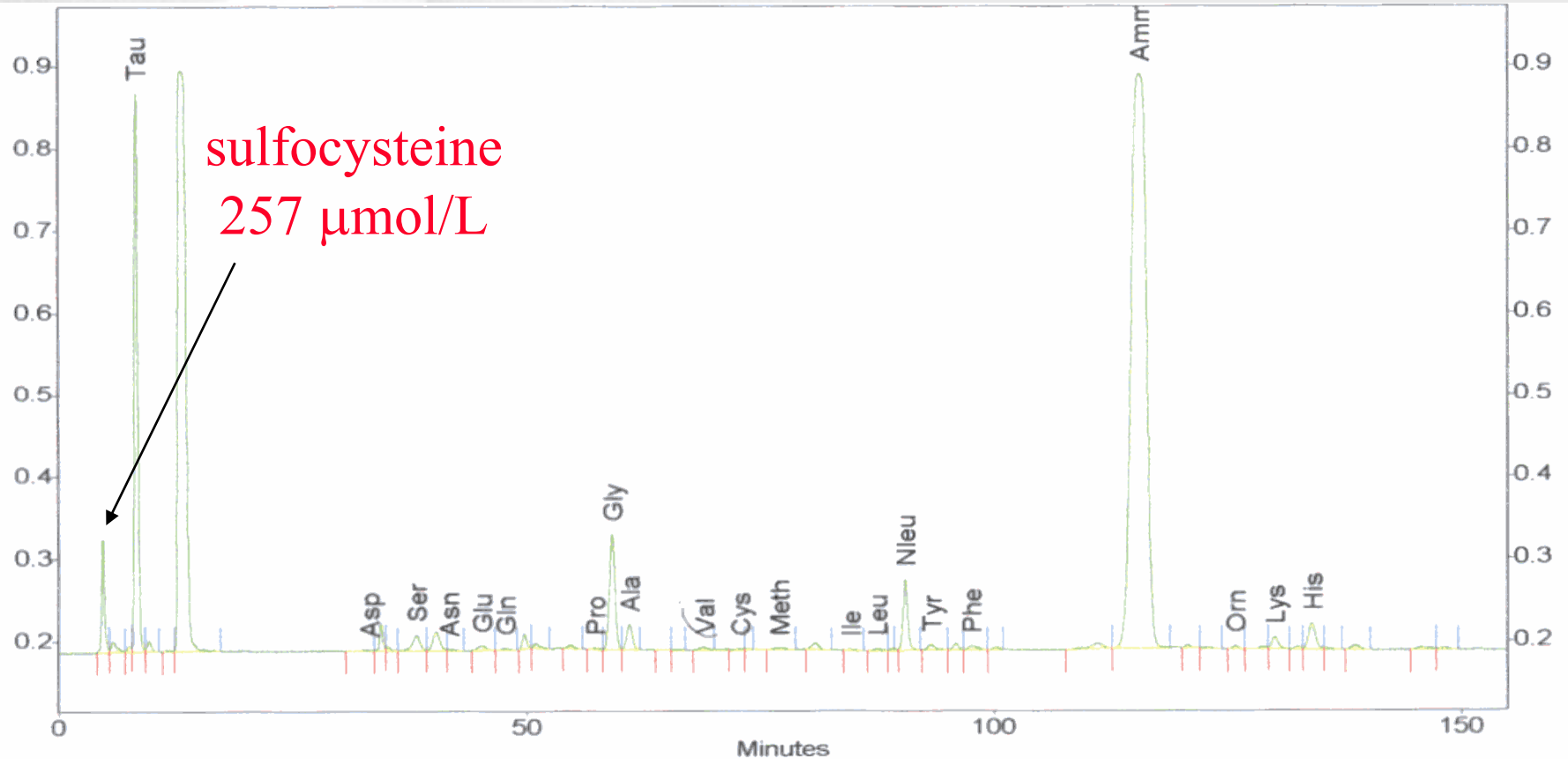
Results summary

Date	Urine						Plasma			
	sulfocys	taurine	cystine	glycine	sulphite		sulfocys	taurine	cystine	glycine
<i>ref value</i>	<i>ND</i>	<i><1051</i>	<i><37</i>	<i><938</i>	<i>neg</i>		<i>ND</i>	<i>92-392</i>	<i>21-73</i>	<i>220-527</i>
6.8.00	139	448	3	504	neg		40	76	ND	244
14.8.00							55	298	ND	449
15.8.00							46	308	ND	412
17.8.00	356	1067	19	2070	pos		44	319	ND	438
22.8.00							60	112	ND	288
24.8.00	304	2087	6	557	neg		40	148	ND	256
25.8.00	367	2404	11	591	neg					

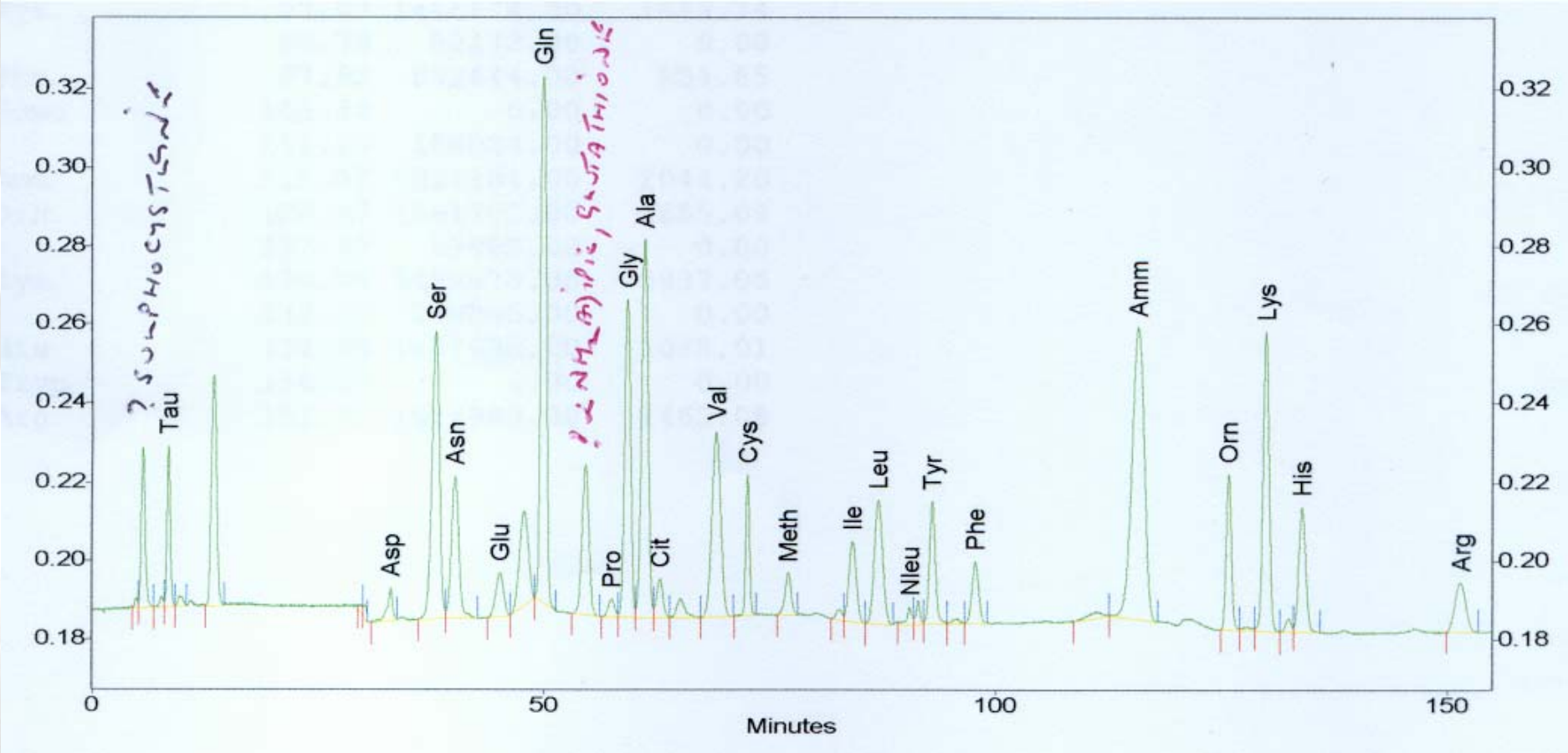
Plasma amino acids



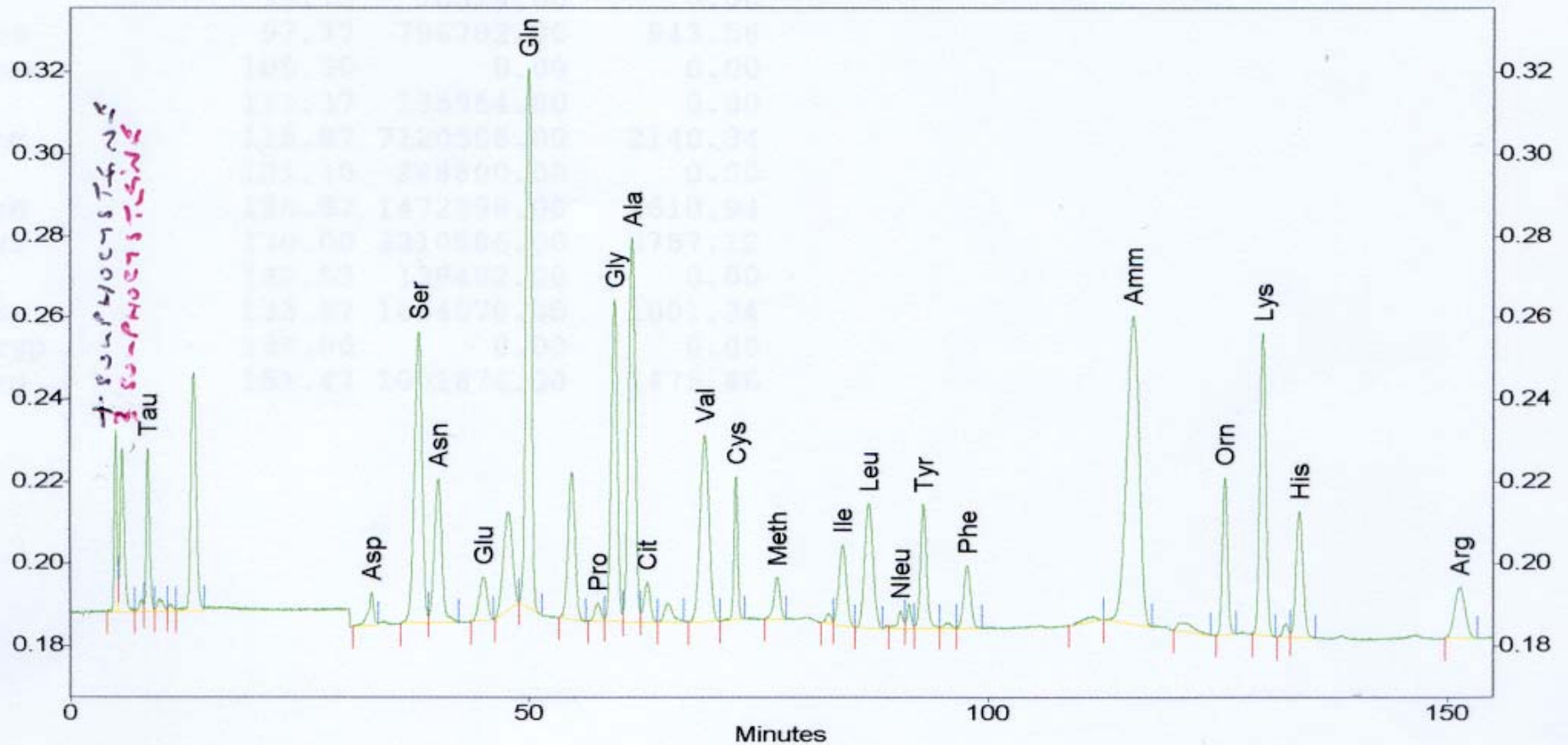
Urine amino acids



Plasma amino acids, referred sample



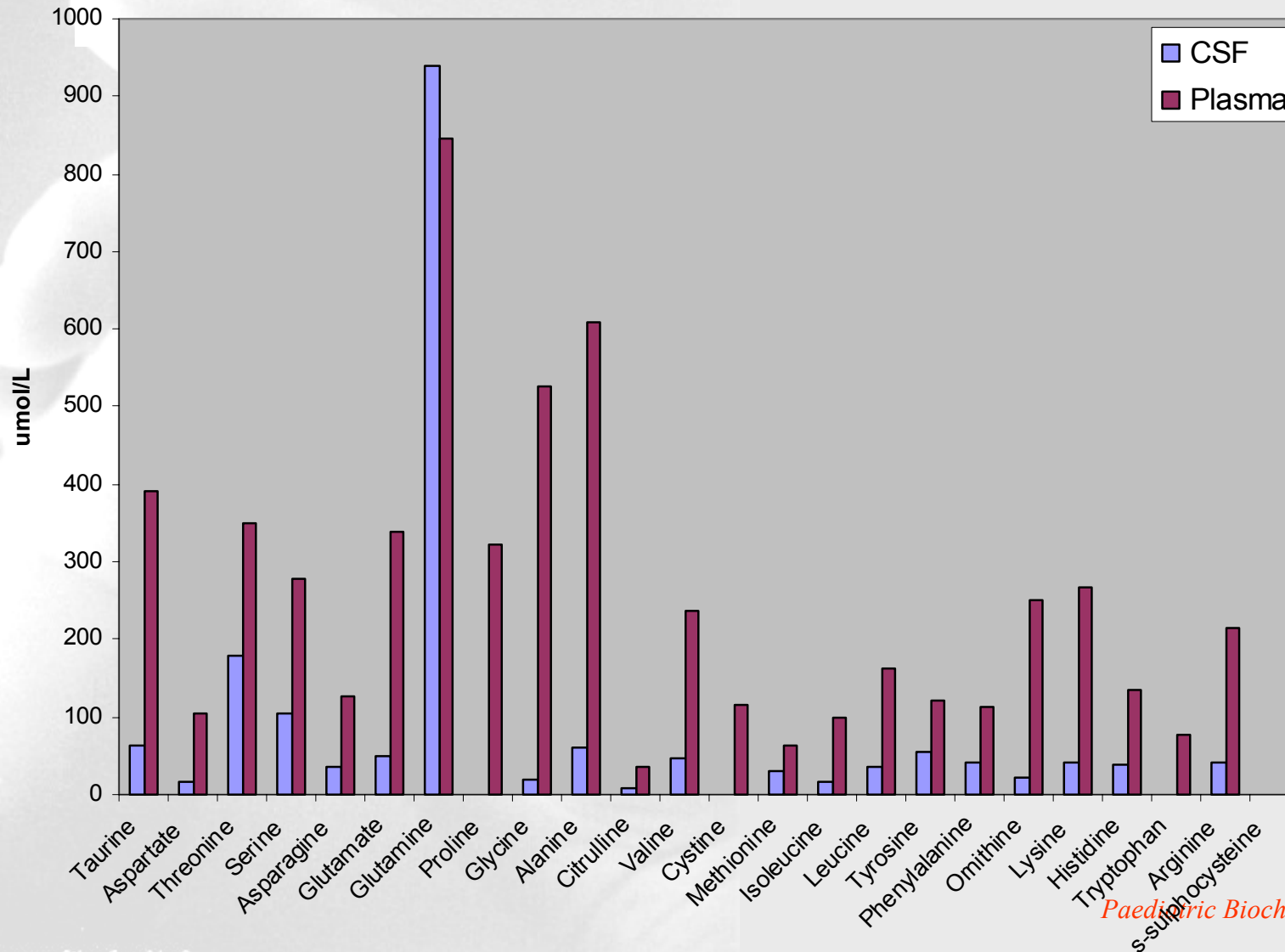
Plasma amino acids, referred sample spiked with sulphocysteine



570:440 ratios, scys 4.3, unknown 10.0

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Amino acid concentrations; 95th Centile upper limits



Glycine and serine

- Glycine encephalopathy (NKH)
 - ↑ gly CSF, plasma, urine
 - Intractable seizures
- 3 phosphoserine dehydrogenase deficiency
 - ↓ ser CSF
 - Intractable seizures, microcephaly

Break through in ante natal Rx serine disorder

- Successful prenatal treatment of 3PGD def, De Koning et al Lancet Dec 2004 (364 p2221)
- 2 previous sibs
 - Serine suppl diet controlled seizures
 - Severe microcephaly
- Maternal serine supplementation *from 26 w*

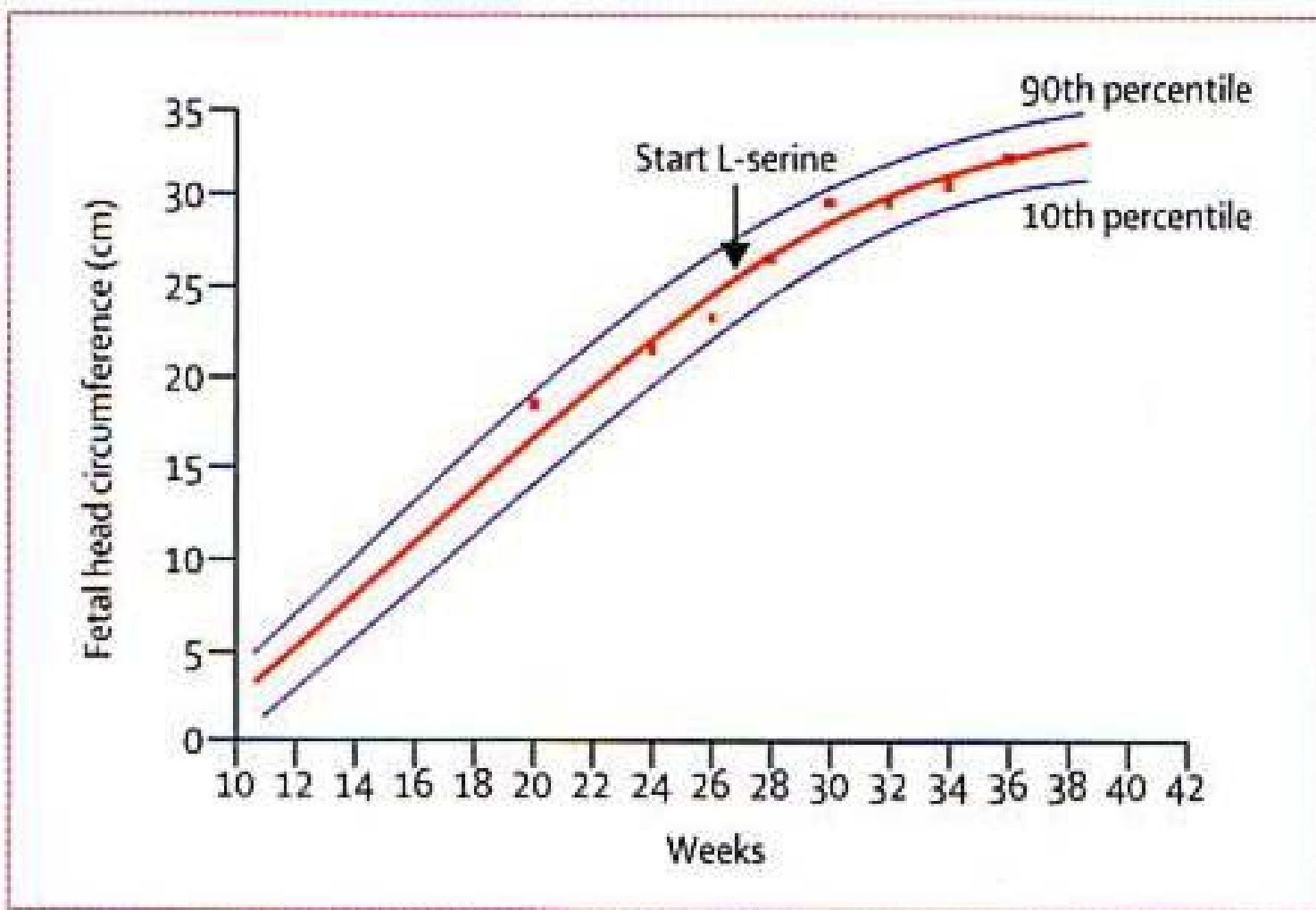


Figure: Fetal head circumference before and during maternal L-serine treatment

De Koning et al, Lancet 2004, 364; 2221

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Prenatal serine therapy: outcome

- Healthy baby girl born at term
- Wt and ht on 40th centile, head 30th
- Cord plasma ser 52 $\mu\text{mol/L}$ (124 - 180)
- Serine supplements given from birth
- Child 4 yrs at time of report, growth neurological status and psychmotor dev normal

Amino acids

Interesting little molecules

Vital to health

In the right balance

Which is different for neonates

And may be compounded by IEMs

Encouraging progress in therapy of 3PGD

We are still learning!

www.metbio.net

