

ERNDIM QA Scheme for qualitative urinary organic acid analysis

Annual Report 2007

Participation

The geographical distributions of the active participants in 2007 are shown in Table 1. Sheffield and Heidelberg participate in each other's scheme and the two centres work closely together under the auspices of the ERNDIM Scientific Advisory Committee.

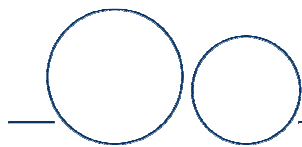
Table 1: Geographical distribution of participants

Country	Number of laboratories	Country	Number of laboratories
Austria	3	Norway	1
Belgium	1	Philippines	1
Canada	6	Poland	1
Croatia	1	Saudi Arabia	3
Cyprus	1	Slovakia	1
Czech Republic	2	Slovenia	1
Denmark	1	Spain	2
Estonia	1	Sweden	2
France	2	Switzerland	3
Germany	11	The Netherlands	9
Hungary	1	United Kingdom	1
Italy	9	USA	12
Latvia	1	Vatican City	1

Im Neuenheimer Feld 153
69120 Heidelberg
Stoffwechsellabor:
Fon +49 (0)6221 56 8276
8423
Fax +49 (0)6221 56 5565
Stoffwechselklinik und -ambulanz:
Fon +49 (0)6221 56 2319 (Anmeldung)
2311 (Information)

Neugeborenencreening:
Fon +49 (0)6221 56 8278

stoffwechsellabor@uni-hd.de
www.stoffwechsel.uni-hd.de



Samples and results

Three sets of three samples (total 9; sample number 151 --159) were distributed to 78 laboratories.

Three participants did not answer to any of the three circulations. Nine laboratories returned results for two circulations, three for only one.

Circulation	Number of returns	Late returns
1. circulation	70	4
2. circulation	70	6
3. circulation	67	2

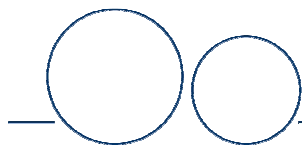
Shipment of the samples

As the years before we sent out the samples for all three circulations together. This is only for organisation reasons and to keep the costs for participating in this scheme as low as possible.

Please remember, the idea of the scheme is to measure the samples evenly spread over the year and report the results near the closing date!

Table 3: Distribution of scores for individual samples (laboratories making returns)

		-2	0	1	2
Sample 151	fumaric aciduria (fumarase deficiency)	1	-	-	69
Sample 152	Normal pattern	1	-	1	68
Sample 153	Normal pattern	2	-	-	68
Sample 154	Normal pattern	-	-	-	70
Sample 155	glutaric aciduria type I (GA I)	-	-	-	70
Sample 156	methylmalonic aciduria	1	-	-	69
Sample 157	propionic aciduria	-	-	-	67
Sample 158	Normal pattern	-	-	-	67
Sample 159	glutaric aciduria type II (GA II)	-	-	5	62



Scoring scheme

Individual returns for each sample were scored on the scale

2	Correct/satisfactory
1	helpful but incomplete
0	unhelpful
-2	misleading

The ERNDIM organisation is moving towards providing a single “Certificate” to cover participation and performance in all its schemes. The scheme organizers of the “Qualitative Organic Acid Scheme” in Sheffield and Heidelberg agreed on criteria to define “Participation” and “Satisfactory Performance”.

We are aware that these criteria are rather arbitrary but we are convinced that they will represent all the different contexts in which the participants are working.

So “Participation” will be defined as requiring at least two returns during a year and “Satisfactory Performance” as obtaining a score of 11 or more based on three returns (out of maximum 18). When two returns have been received a score of 7 or more (in this case possible maximum score 12) is satisfactory.

Comments on performance

In 2007 nearly all participants correctly diagnosed the circulated samples. Diagnosis of classical organoacidopathies like **propionic aciduria** or **methylmalonic aciduria** as well as the diagnosis of disorders of the tricarboxylic acid cycle (**fumaric aciduria**) might be less challenging.

In case of **glutaric aciduria type II** identification of noticeable changes in organic acid profiles and assigning these analytical findings to disorders of fatty acid oxidations were comparable for all laboratories.

For the first time a common sample was distributed by Sheffield and Heidelberg in 2007. For this sample the diagnosis of **glutaric aciduria type I** was correctly found by all active laboratories (70). In the Sheffield scheme seventy-six participants scored 2, one scored 1 and one obtained a score of -1.

The participants cumulative scores are shown in table 4. Cumulative scores are the scores for the whole year. This year fifty-five participants (71%) got full marks!

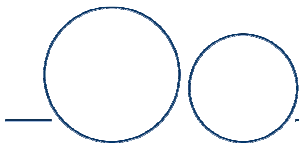


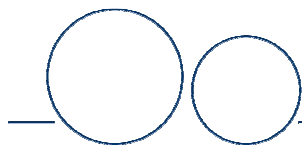
Table 4: cumulative total scores 2007 - 2005

Cumulative scores	Number of laboratories		
	2007	2006	2005
18	55	16	25
17	4	10	4
16	-	10	12
15	-	4	1
14	2	12	6
13	-	2	2
12	7	5	6
11	2	-	1
10	1	4	3
9	-	1	1
8	1	-	-
7	-	-	1
6	3	3	-
5	-	-	-
4	-	-	-
3	-	-	-
2	-	-	2
1	-	-	-
0	3	4	3

Your total score 2007

Your total score for 2007 was:

Your number of returns in 2007 was:



General comments

We would just like to point out here that we are not able to accept returns sent in after the report for the corresponding circulation has been mailed because this would not be compatible with the overall intention of the scheme. We are conscious of the fact that posted results could get lost on a variety of ways. Therefore it would be a good advice to send in results on more than one routes (e.g. FAX and email, regular mail and FAX or email).

Special thank for the laboratories that supported us last year with samples. This is critical for the success of the program and will keep the scheme interesting. **It is most appreciated that you will continue to support us with urine from patients. Please send us at least 250 ml urine of any interesting patients you may have. We will cover the costs.**

Yours sincerely,

Dr. C. D. Langhans

*Laboratory of Metabolic
Diseases*

Dr. V. Peters

*Laboratory of Metabolic
Diseases*

Prof. Dr. G. F. Hoffmann

*Director
Department of General
Paediatrics*