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To

Stoffwechselzentrum Heidelberg Stoffwechsellabor

Kinderheilkunde I

(Schwerpunkt: Allgemeine Pädiatrie, Stoffwechsel, Gastroenterologie u. Nephrologie)

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Universitätsklinik für Kinder- und Jugendmedizin

ERNDIM QA Scheme for qualitative urinary organic acid analysis

Annual Report 2010

Participation

The geographical distributions of the active participants of the quality assurance scheme organized and distributed through the centre of Heidelberg in 2010 are shown in Table 1. Sheffield and Heidelberg participate in each other's scheme and the two centers work closely together under the auspices of the ERNDIM Scientific Advisory Committee.

Table 1: Geographical distribution of participants						
Country	Number of		Country	Number of		
	laboratories			laboratories		
Austria	3		Lithuania	1		
Belgium	1		Norway	1		
Canada	6		Philippines	1		
Croatia	1		Poland	1		
Cypres	1		Saudi Arabia	1		
Czech Republic	2		Slovakia	1		
Denmark	1		Slovenia	1		
Estonia	1		Spain	3		
France	2		Sweden	2		
Germany	13		Switzerland	3		
Greece	1		The Netherlands	10		
Hungary	1		Ukraine	1		
India	1		United Kingdom	1		
Italy	11		USA	12		
Latvia	1		Vatican City	1		
Luxembourg	1					

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Samples and results

Three sets of three samples (total 9; sample number 178 --186) were distributed to 87 laboratories.

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

Table 2: Receipt of results					
Circulation	ation Number of returns Late returns				
1. circulation	79	1			
2. circulation	83	3			
3. circulation	84	3			

Shipment of the samples

As the years before we sent out the samples for all three circulations together. This is only for organizational reasons especially 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 178	Methylmalonic aciduria				79		
Sample 179	Normal pattern				79		
Sample 180	Glutaric aciduria type I				79		
Sample 181	Normal pattern	3			80		
Sample 182	Normal pattern	3	1		79		
Sample 183	Succinic semialdehyde dehydrogenase deficiency (SSADHD)	1	1		81		
Sample 184	Hawkinsinuria	26	4	2	52		
Sample 185	Mevalonic aciduria	1			83		
Sample 186	Maple syrup urine disease (MSUD)	4			80		



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Scoring scheme

Individual returns for each sample were scored on the scale

- 2 Correct/satisfactory
- 1 helpful but incomplete
- o 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 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

The overall performance was 100% for methylmalonic aciduria and glutaric aciduria type I, and 99% and 95% for mevalonic aciduria and maple syrup urine disease (MSUD) respectively.

In contrast, analytical performance and interpretative proficiency varied widely for **hawkinsinuria**. Whereas 4-hydroxycyclohexylacetic acid was identified by 75% of the laboratories, the correct diagnosis was given by 62% of the participants.

The three circulated **normal organic acid profiles** were clearly identified by nearly all participants. False positive results are at 2.5%

The participants' cumulative scores are shown in table 4. Cumulative scores are the scores for the whole year. This year forty-six participants (55%) got full marks!



Table 4: cumulative total scores 2010 - 2005

	Number of laboratories					
Cumulative scores	2010	2009	2008	2007	2006	2005
18	46	27	21	55	16	25
17	2	3	26	4	10	4
16	4	2	5	-	10	12
15			1	-	4	1
14	19	31	11	2	12	6
13		1	2	-	2	2
12	3	6	6	7	5	6
11			3	2	-	1
10	3	1	1	1	4	3
9	2	1	-	-	1	1
8	3	4	1	1	-	-
7			1	-	-	1
6		2	1	3	3	-
5			-	-	-	-
4	3	1	-	-	-	-
3			-	-	-	-
2			-	-	-	2
1			-	-	-	-
O	2	6	4	3	4	3

Your total score 2010

Your total score for 2010 was:

Your number of returns in 2010 was:





General comments

We would 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 route (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.

In 2010 samples for the QOA scheme were kindly contributed by

• Dr. Jeannette Klein, Berlin, Germany

Yours sincerely,

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