



Universitätskinderklinik und Poliklinik

Kinderheilkunde I

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

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ERNDIM QA Scheme for qualitative urinary organic acid analysis

Annual Report 2003

Participation

Because of the increasing number of participants the scheme has been split between Sheffield and Heidelberg, starting from the beginning of 2001. In 2001 26 participants, in 2002 forty-seven participants and in 2003 forty-nine participants received their samples from Heidelberg. The geographical distributions of the active participants in 2003 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.

Country	Number of laboratories	Country	Number of laboratories
Austria	2	Poland	1
Canada	4	Saudi Arabia	1
Croatia	1	Slovakia	2
Czech Republic	2	Slovenia	1
Denmark	1	Sweden	2
Finland	1	Switzerland	2
Germany	9	Turkey	1
Italy	10	United Kingdom	1
Norway	1	USA	7

Table 1: Geographical distribution of participants

Samples and results

Three sets of three samples (total 9; sample number 115-123) were distributed to 49 laboratories in 2003. 44 laboratories returned results to the first and second circulation and 42 returned results to the third circulation.

Circulation	Number of returns
1. circulation	44
2. circulation	44
3. circulation	42

Table 2: Receipt of results

A few laboratories returned their results extremely late, sometimes after the circulation report. We have included such late results wherever possible, but this is not possible once the returns have been analysed and the report written. **In future, any results returned late may be disregarded.**

Shipment of the samples

Last year we sent out the samples for all three circulations together. This is only for organisation reasons and keep the costs for participating in this scheme as low as possible. Some laboratories reported all three circulations together. **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 115 Maple sirup urine disease (MSUD)	0	4	3	37
Sample 116 Glutaric aciduria type I (manufactured sample)	24	2	0	18
Sample 117 Normal pattern (same sample as sample 122)	0	2	2	40
Sample 118 Canavan disease	3	0	0	41
Sample 119 Normal pattern (same sample as sample 121)	0	0	0	44
Sample 120 3-Methylcrotonyl-CoA carboxylase deficiency	1	0	0	43
Sample 121 Normal pattern (same sample as sample 119)	0	1	1	40
Sample 122 Normal pattern (same sample as sample 117)	0	0	8	34
Sample 123 Phyenyketonuria (PKU)	0	0	0	42

Scoring scheme

Individual returns for each sample were scored on the scale

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

Most active laboratories diagnosed correctly MSUD (84%), Canavan disease (93%), 3-Methylcrotonyl-CoA carboxylase deficiency (98%) and PKU (100%). The greatest challenge was presented by sample 116. The idea was to test whether routine organic acid analysis is likely to detect low amounts of 3-hydroxyglutarate. To manufacture the sample was necessary because we have no access to genuine samples from any low-excretor GA1 patients. Manufactured samples are also in future the exception but in general we run out of good samples to distribute!

This year, we sent out the same control samples twice but with different description about the clinical picture. Sample 117 was identically with sample 122 and sample 119 was identically with sample 121. By most laboratories same results were obtained for these samples in both cases, few laboratories came to different conclusions. For sample 117/122, 21% of the laboratories came to (slightly) different interpretations of the chromatograms. Some of the laboratories interpreted only one of the identical samples as normal and the other sample as coming from a patient with propionic acidaemia, β -oxidation defects, mitochondrial defects or glutaric aciduria. This is probably due to the different clinical descriptions or the expectation of the participants to have always only one control in each circulation (we sent out 2 controls in the third circulation).

Comments on performance

The participants cumulative scores are shown in diagram 1 and in table 4. Cumulative scores are the scores for the whole year 2003. Thirty-one participants scored more than 12 this year. This means that over 82% of the laboratories, which participated in all 3 circulations, achieved a score over 12 which we regard as a very good result!

The poor performance of some laboratories scoring less than 10 this year is due to missing returns. Six laboratories only returned 2 distributions; four laboratories only returned 1 distribution for the whole year and one laboratory did not send any results.

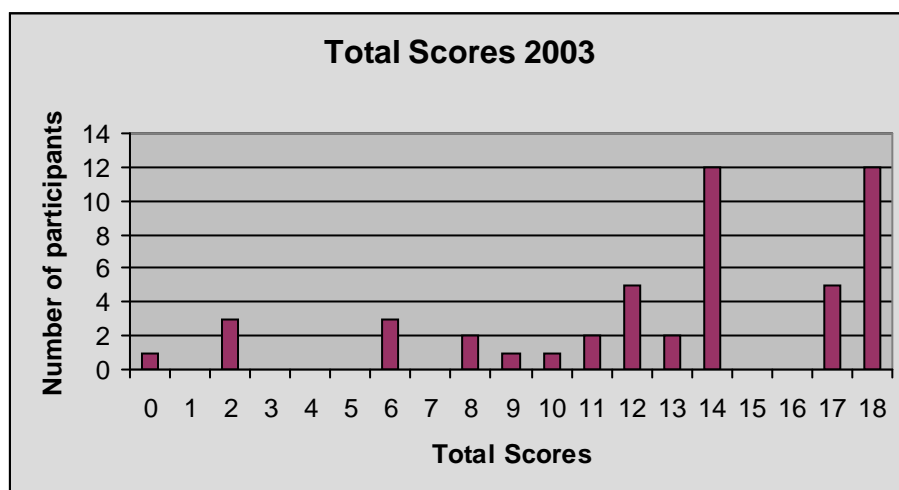


Diagram 1: Total scores 2003

Table 4: total scores 2001

Cumulative scores	Numbers of laboratories
18	12
17	5
16	/
15	/
14	12
13	2
12	5
11	2
10	1
9	1
8	2
7	/
6	3
5	/
4	/
3	/
2	3
1	/
0	1

Your total score 2003

Your total score for 2003 was:

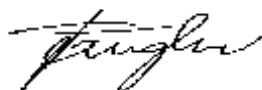
General comments

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 200 ml urine of any interesting patients you may have. We will cover the costs.**

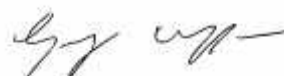
Yours sincerely,



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