Laboratory number
ERNDIM number ERN***

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8th June 2006

ERNDIM QAP
for qualitative urinary organic acid analysis

Annual Report 2005 (Sheffield)

Participation

Active participants (reporting on at least one sample in the year) are shown in Table 1. No further laboratories were transferred to the Heidelberg scheme for 2005. The two schemes are run separately, usually circulating different samples, but try to keep the same general philosophy and format. To assist this, the two organising laboratories each participate in the other's scheme.

Table 1: Geographical distribution of participants

	2005	***2004***	***2003***	***2002***	***2001***	***2000***
United Kingdom	21	21	21	22	21	21
France	12	13	13	11	11	11
Italy	0	0	0	0	1	9
The Netherlands	0	0	10	9	8	8
Belgium	6	6	6	6	6	7
Germany	1†	1†	1†	1†	1†	9
Australia	6	6	6	6	6	6
Spain	5	5	5	5	5	5
USA	1	0	0	0	5	5
Austria	0	0	0	0	0	3
Canada	0	0	0	0	3	3
Czech Republic	0	0	0	0	0	2
Denmark	0	0	0	0	2	2
Republic of China	4	4	4	3	3	2
Israel	2	2	2	1	1	1
Portugal	2	2	2	1	1	1
Sweden	0	0	0	0	2	2
Switzerland	0	0	0	0	0	2
Other countries	9*	7	7	6	11	14
TOTAL	**69**	**67**	**77**	**71**	**87**	**113**

† Heidelberg laboratory; * One participant each from Argentina, Brazil, Finland, Republic of Ireland, Lebanon, Malaysia, New Zealand, South Korea and Taiwan

Samples and results

Three sets of three samples (total 9; sample numbers 133 - 141) were distributed in 2005. Sixty-two laboratories returned results for all three circulations.

Table 2: Receipt of results into the executive centre within the specified time period (approximately 6 weeks from dispatch) :

Number of returns in 2005	*Number of participants*				
	0 Late	*1 Late*	*2 Late*	*3 Late*	*Total*
1	1	1	-	-	2
2	2	2	1	-	5
3	47	9	3	0	62

Instrumentation

Currently only four active participants are relying on gas-chromatography alone, the remainder performing their analyses wholly or in part by GC-MS.

Scoring of results

Summary results for the individual returns were dispatched earlier. To enable data reduction and analysis of long-term performance the results were scored as shown below:

2	satisfactory
1	helpful but incomplete
0	unhelpful
-1	slightly misleading
-2	misleading.

A score of zero was given for failing to return an individual result.

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

Sample	**Scores**				
	-2	**-1**	**0**	**1**	**2**
#133 Normal pattern	2	-	-	1	64
#134 Maple syrup urine disease (intermittent/mild in an adult)	5	1	5	11	45
#135 Propionic acidaemia	-	-	1	1	66
#136 MCAD deficiency, non-crisis in an adult	9	4	-	2	51
#137 Tyrosinaemia type 1	1	-	1	9	55
#138 Neonate, no abnormality	3	2	4	2	55
#139 4-Hydroxybutyrate ingestion in a teenager	2	5	9	3	46
#140 Normal pattern with ? slightly increased lactate	4	2	5	6	48
#141 Succinic semialdehyde dehydrogenase deficiency	10	1	3	1	50

Table 4: Cumulative scores for 2004 and the five preceding years (current Sheffield participants only)

Year	2005			2004	2003	2002	2001	2000
Lab ID no	Number of returns	Late returns	Total score	Total score	Total score	Total score	Total score	Total score
3	3	0	17	17	16	12	13	10
4	3	0	16	12	14	17	12	15
5	3	0	11	15	12	15	17	18
6	3	0	9	18	13	18	17	14
7	3	0	10	14	16	18	18	14
9	3	1	17	18	9	18	18	18
10	3	0	18	17	16	14	15	15
11	3	1	17	18	12	18	18	18
12	3	0	18	12	16	14	18	18
13	3	0	16	17	12	12	17	18
14	3	2	17	12	10	13	17	8
15	3	0	18	16	16	11	17	17
17	3	0	15	13	13	14	11	12
18	3	0	18	11	16	18	17	14
19	3	0	14	18	16	18	15	13
21	2	1	12	14	16	12	12	16
24	3	0	17	18	12	18	17	18
25	3	0	18	17	14	16	17	18
26	3	0	16	18	16	18	17	18
27	2	2	-3	9	1	4	-1	11
28	3	0	5	7	4	14	15	14
29	3	0	18	17	16	14	15	18
31	3	0	17	18	16	18	17	17
32	3	1	18	11	16	18	12	18
35	3	0	14	17	16	18	17	18
38	3	0	18	18	16	18	18	18
42	3	0	14	14	16	18	18	18
43	3	1	11	16	11	17	18	16
44	3	0	14	17	15	18	15	14
48	2	0	12	11	8	16	10	14
49	3	2	15	12	11	15	18	14
51	3	0	18	17	12	18	18	17
52	3	0	16	16	13	10	18	18
65	3	0	10	18	16	16	14	18
66	3	0	18	18	14	14	17	18
76	3	0	6	13	13	18	16	18
79	3	0	17	13	14	17	11	13
83	3	0	14	18	16	15	17	18
85	3	2	11	14	12	16	17	18
86	3	1	17	16	12	11	17	14
88	3	0	13	14	5	8	10	18
90	1	0	6	17	15	11	11	17
92	3	0	14	17	16	17	17	12

Year	2005			2004	2003	2002	2001	2000
Lab ID no	Number of returns	Late returns	Total score	Total score	Total score	Total score	Total score	Total score
93	3	0	17	18	16	18	17	14
94	3	0	15	17	6	14	13	11
96	3	2	13	15	10	12	17	6
98	3	1	16	16	16	17	18	16
101	3	0	17	17	16	16	18	18
102	3	0	18	17	13	17	16	18
104	3	0	10	14	12	16	17	14
106	3	0	18	18	10			
108	3	0	13	14	12	16	8	10
111	3	0	18	18	9	18	17	18
113	3	1	2	9	0	10	12	7
114	3	1	8	13	7	6	17	14
119	3	0	18	17	12	18	17	6
120	2	0	11	12	8	16	10	
121	3	0	16	16	12	11	12	
126	3	2	13	11	15			
127	1	1	0	7				
128	2	1	5	12	4			
130	3	0	15	18	16			
131	3	0	7	13	9			
132	3	1	11	8	8			
133	3	2	9	15	5			
134	3	0	7	17	9			
136	3	0	4					
137	3	0	16					
138	3	0	4					
Maximum score	3	3	18	18	16	18	18	18

Table 5: Ranking of scores

In 2005 Fourteen laboratories scored 18, ten scored 17 and were ranked 15th equal; seven scored sixteen and were ranked 25th equal, etc. Rankings for 2004 are shown for comparison.

Score	18	17	16	15	14	13	12	11	10	9	8	<8
2005	1	15	25	32	36	42	46	48	53	55	57	58
2004	1	16	31	37	40	47	52	58	62	63	65	66

Commentary

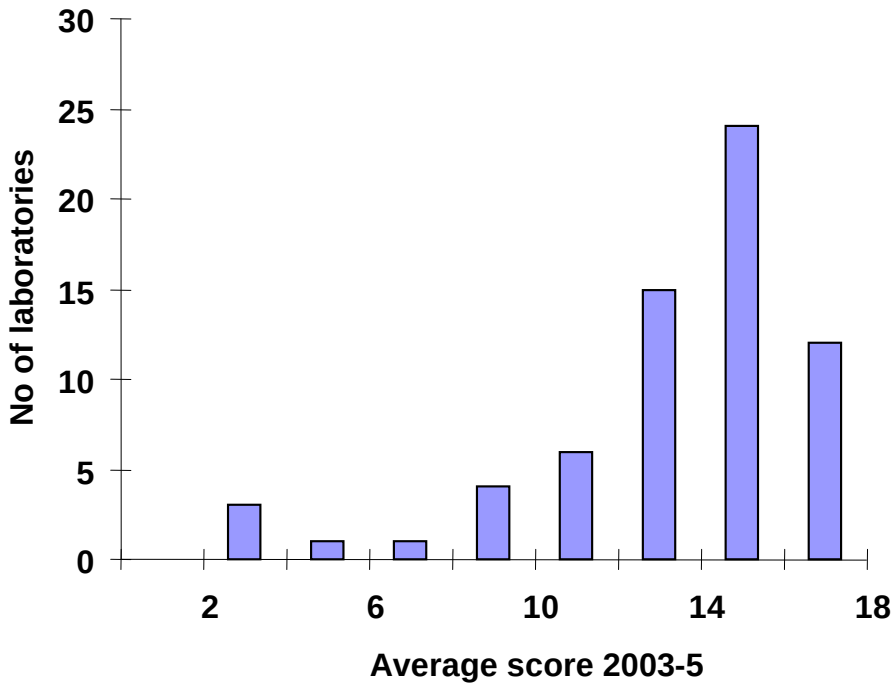
Most participating laboratories are proficient at producing high quality chromatograms and at identifying the metabolites present and no major surprises emerged from this year's distributions. The main emphasis is now on placing laboratory investigations in their broader context. As described in last year's Annual Report, we introduced the structured report form to encourage participants to address the main questions implicit in the referring physician's request:

- what are the major analytical findings?
- what is the most likely diagnosis?
    - how certain is it?
    - what, if any, are the possible alternatives?
- what further investigations are required to confirm or clarify the diagnosis?

This experiment has largely been successful but with two of this year's samples some participants scored badly through failure to bear clinical context in mind. With sample # 139 this resulted in misinterpretation of the analytical findings. In the case of the moderately prominent lactate peak from sample #140 suggestions for further investigation should have taken into consideration both the burden on the patient and the cost, the obvious next step being measurement(s) of plasma lactate rather than glucose loading test, muscle biopsy, mutation analysis ….

Unsatisfactory performance

For many years we have been attempting to quantify participant's performance by allocating an overall score (from +2 to -2) for each sample. Scoring a qualitative test is more difficult than for a quantitative assay. Inevitably there will be some degree of arbitrariness, particularly with the more difficult samples. For this reason, and because our participants work in diverse clinical and institutional settings, we have avoided setting a firm boundary to define satisfactory performance. However, a laboratory's long-term average score must give some indication of its effectiveness in routine diagnosis and we are disturbed that a few participants have consistently low scores.



Your laboratory's average score for 2003-2005 was -----

We urge participants with low average scores, perhaps 12 or less, to review their staffing and procedures to ensure that they are providing as good a service as circumstances permit. For those with limited resources it may be helpful to form a working relationship with a larger centre. A poster analysing factors associated with suboptimal performance is being prepared, jointly with the organisers of the Heidelberg scheme, for this September's ICIEM in Japan. A copy will be included with a future report.

We hope that you continue to find the scheme useful.

Yours sincerely

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Scheme organisers