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ERNDIM QAP
for qualitative urinary organic acid analysis

Annual Report 2007 (Sheffield)

Participation

Active participants (reporting on at least one sample in the year) are shown in Table 1. The number of participants continues to grow. The Sheffield and Heidelberg qualitative urinary organic acid schemes are run separately but try to keep the same general philosophy and format. To assist this, the two organising laboratories each participate in the other's scheme and in 2007 one sample was distributed in both schemes (Table 2).

Table 1: Geographical distribution of participants

	2007	*2006*	*2005*	*2004*	*2003*
Argentina	2	1	1	1	1
Australia	6	6	6	6	6
Belgium	5	4	6	6	6
Brazil	1	1	1	1	1
Canada	1	1	0	0	0
Democratic Republic of China	1	1	1	1	1
Finland	1	1	1	1	1
France	13	11	12	13	13
Germany†	1	1	1	1	1
Israel	2	2	2	2	2
Japan	1	1	0	0	0
Lebanon	1	1	1	1	1
Malaysia	2	2	1	1	1
The Netherlands	0	0	0	0	10
New Zealand	2	2	1	0	0
People's Republic of China	6	4	4	4	4
Portugal	2	2	2	2	2
Republic of Korea	1	1	1	0	0
Republic of Ireland	1	1	1	1	1
Spain	5	5	5	5	5
United Kingdom	20	21	21	21	21
USA	4	2	1	0	0
Venezuela	1	1	0	0	0
TOTAL	**79**	**72**	**69**	**67**	**77**

† Heidelberg laboratory

Samples and results

Three sets of three samples (total 9; sample numbers 151-159) were distributed in 2007. As an experiment to save on distribution costs all nine samples were dispatched together. This has been the practice of the Heidelberg scheme or some years. Seventy-one laboratories returned results for all three circulations (90%, 87.5% in 2006), seven for only two, and one laboratory made only a single return.

Instrumentation

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

Scoring of results

To enable data reduction the results were scored as shown below:

Satisfactory	2	Helpful but incomplete	1
Unhelpful	0	Slightly misleading	-1
Misleading	-2	Failing to return an individual result	0

Two points are deducted for transposed sample numbers.

Table 2: Distribution of scores for individual samples (laboratories making returns)

Sample	Scores				
	-2	**-1**	**0**	**1**	**2**
#151 Teenage girl with muscle weakness: 3-Methylcrotonyl-CoA carboxylase deficiency	1	1	-	1	72
#152 Eight-year-old girl with enlarged liver: No abnormality detected.	-	-	-	2	73
#153 Six-year-old boy, admitted with pancreatitis: Isovaleric acidaemia	-	-	1	-	74
#154 Seven-year-old boy, hyperkeratosis and blisters on feet and hands: Tyrosinaemia type 2	3	2	2	3	68
#155 Seven-year old boy with chronic liver disease: Medium-chain acyl-CoA dehydrogenase deficiency	*	*	*	*	*
#156 Fifteen-year-old boy, speech problems, intermittent dyskinesia, severe gait ataxia: Glutaric aciduria type 1†	-	1	-	1	76
#157 Eighteen-month-old girl with ? Reye syndrome: Medium-chain acyl-CoA dehydrogenase deficiency	-	2	3	-	70
#158 Three-year-old boy, found at home acidotic and comatose: Ethylene glycol ingestion	1	-	12	1	61
#159 Five-year-old boy, developmental regression: No abnormality	2	-	2	1	70

* This sample was unstable on prolonged storage and no individual scores were allocated

† Common sample with the Heidelberg scheme: all 70 participants scored 2

Table 3: Cumulative scores for 2005 - 2007 (current Sheffield participants only)

	2007			2006		2005		2005-7
Laboratory Number	No of returns	Late returns	Total score	No of returns	Total score	No of returns	Total score	Average score*
3	3	0	16	3	18	3	17	1.96
4	3	0	16	3	17	3	16	1.88
5	3	0	10	3	18	3	11	1.50
6	3	0	16	3	18	3	9	1.65
7	3	0	16	3	17	3	10	1.65
9	3	0	16	3	17	3	17	1.92
10	3	1	16	3	15	3	18	1.88
11	3	0	16	3	17	3	17	1.92
12	3	0	16	3	18	3	18	2.00
13	3	0	16	3	18	3	16	1.92
14	3	0	16	3	18	3	17	1.96
15	3	0	16	3	14	3	18	1.85
17	3	0	15	3	10	3	15	1.54
18	3	0	16	3	16	3	18	1.92
19	2	0	10	3	18	3	14	1.83
21	3	0	16	3	18	2	12	2.00
24	3	0	16	3	16	3	17	1.88
25	3	0	16	2	11	3	18	1.96
26	3	0	15	3	14	3	16	1.73
27	3	0	16	3	10	2	-3	1.00
28	3	1	13	3	8	3	5	1.00
29	3	0	14	3	18	3	18	1.92
31	3	0	14	2	12	3	17	1.87
32	3	0	16	3	18	3	18	2.00
35	3	0	16	3	18	3	14	1.85
38	3	0	16	3	18	3	18	2.00
42	2	0	16	3	18	3	14	2.09
44	3	0	15	3	15	3	14	1.69
48	3	0	16	3	18	2	12	2.00
49	3	0	16			3	15	1.82
51	3	0	16	3	18	3	18	2.00
52	3	1	15	3	15	3	16	1.77
65	3	1	16	3	18	3	10	1.69
66	3	0	16	3	17	3	18	1.96
83	3	1	16	3	17	3	14	1.81
85	3	0	16	3	18	3	11	1.73
86	3	0	16	2	7	3	17	1.74
88	3	0	16	2	12	3	13	1.78
90	1	1	6	2	12	1	6	2.00
92	3	0	11	3	15	3	14	1.54
93	3	0	16	3	17	3	17	1.92
94	3	0	16	3	17	3	15	1.85
96	3	0	16	2	11	3	13	1.74

	2007			2006		2005		2005-7
Laboratory Number	No of returns	Late returns	Total score	No of returns	Total score	No of returns	Total score	Average score*
98	3	2	16	3	17	3	16	1.88
101	3	0	16	3	18	3	17	1.96
102	3	0	13	3	18	3	18	1.88
104	2	1	10	3	12	3	10	1.39
106	3	0	16	3	18	3	18	2.00
108	3	1	14	3	18	3	13	1.73
111	3	0	16	3	14	3	18	1.85
113	3	0	10	3	13	3	2	0.96
114	3	0	10	3	16	3	8	1.31
119	3	0	16	3	18	3	18	2.00
120	3	0	10	3	16	2	11	1.61
121	3	0	12	3	18	3	16	1.77
126	3	1	11	3	15	3	13	1.50
128	2	0	5	3	15	2	5	1.25
130	3	0	16	2	12	3	15	1.87
132	3	0	16	3	18	3	11	1.73
133	3	2	14	3	15	3	9	1.46
134	3	0	16	3	18	3	7	1.58
135	3	1	14	2	8	0	0	1.57
136	3	0	10	3	16	3	4	1.15
137	3	0	16	3	18	3	16	1.92
138	3	0	15	3	13	3	4	1.23
139	3	0	14	3	15			
140	3	0	14	3	14			
141	3	1	12	3	15			
142	3	0	16	3	16			
143	3	0	11	2	10			
144	3	0	14					
146	2	0	8					
147	3	0	16					
148	2	0	10					
149	3	0	16					
150	3	0	12					
151	3	3	16					
152	3	1	5					
153	2	1	10					

*The average score is **per sample reported**. The maximum score for 2007 was 16. For 2005 and 2006 the maximum scores were 18.

Your Laboratory Number in the above Table is ***

Commentary

None of this year's samples presented any great difficulties though the relatively poor performance on samples #154 and #158 highlighted the importance of interpreting results in their clinical context. Some laboratories still have difficulty in detecting small but diagnostically significantly increases in hexanoyl- or suberylglycine excretion.

In previous years some participants have experienced problems with mail, their samples or the subsequent reports having gone astray. If anything, FAX has proved to be less reliable than conventional mail and we recommend using both as a precaution.

This year we sent the entire set of nine urine samples as a single consignment, to be analysed and reported in three sets, with a group E-mail to advise that they had been dispatched. We also sent out E-mail reminders to participants whose reports are outstanding after the closing dates. This revealed that a small number of returns had indeed gone missing in the mail and that a slightly larger number of laboratories had overlooked the closing date or lost their response forms – a disadvantage of sending all the samples out together.

In order to do this we need up-to-date E-mail addresses for all participants (not their hospital finance departments please). If you are registered as a Sheffield participant for 2008 your E-mail address appears below:

###########@#####.###

If this is incorrect please let us know on rodney.pollitt@sch.nhs.uk .

The ERNDIM organisation is moving towards providing a single "Certificate" to cover participation and performance in all its schemes. This requires us to define criteria for both "Participation" and "Satisfactory Performance". We have defined Participation as requiring at least two returns during the year. Satisfactory Performance is more difficult to define. In theory, any missed diagnosis is unsatisfactory but we are aware that our participants are working in a variety of contexts and that the statistical significance of a single year's results is limited. Thus we have adopted the rather arbitrary criterion that a score of 11 or more based on three returns (maximum possible score 18), or of 7 or more where only two returns have been received (maximum possible score 12), is satisfactory. We repeat the advice given in 2005 that participants with low scores should 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.

We have some "interesting" samples lined up for 2008 and hope that you will continue to find the scheme useful.

Yours sincerely

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Scheme organisers