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March 2007

**ERNDIM QAP
for qualitative urinary organic acid analysis**

Annual Report 2006 (Sheffield)

Participation

Active participants (reporting on at least one sample in the year) are shown in Table 1. Three laboratories left the scheme and six joined. 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 of the samples will be distributed in both schemes.

Table 1: Geographical distribution of participants

	2006	2005	2004	2003	2002
Argentina	1	1	1	1	1
Australia	6	6	6	6	6
Belgium	4	6	6	6	6
Brazil	1	1	1	1	1
Canada	1	0	0	0	0
Democratic Republic of China	1	1	1	1	1
Finland	1	1	1	1	0
France	11	12	13	13	11
Germany†	1	1	1	1	1
Israel	2	2	2	2	1
Japan	1	0	0	0	0
Lebanon	1	1	1	1	1
Malaysia	2	1	1	1	0
The Netherlands	0	0	0	10	9
New Zealand	2	1	0	0	0
People's Republic of China	4	4	4	4	3
Portugal	2	2	2	2	1
Republic of Korea	1	1	0	0	0
Republic of Ireland	1	1	1	1	1
Spain	5	5	5	5	5
United Kingdom	21	21	21	21	22
USA	2	1	0	0	0
Venezuela	1	0	0	0	0
TOTAL	72	69	67	77	70

† Heidelberg laboratory

Samples and results

Three sets of three samples (total 9; sample numbers 142 - 150) were distributed in 2006. Sixty-three laboratories returned results for all three circulations, nine for only two.

Instrumentation

Currently only three 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. Two points were deducted for transposed sample numbers.

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

Sample	Scores				
	-2	-1	0	1	2
#142 2-month-old boy, poor feeding, metabolic acidosis. Massive excretion of methylmalonate.	-	-	-	14	54
#143 18-month-old girl, vomiting, acidosis and hypoglycaemia following viral illness. Beta-ketothiolase deficiency.	1	1	1	4	61
#144 3-year-old boy, developmental delay, ? regression. No abnormality.	-	-	-	-	68
#145 4-year-old boy, vomiting, seizures and acidosis during the first week of life. Now on anti-epileptic drugs. 3-Hydroxy-3-methylglutaryl-CoA lyase deficiency.	-	-	7	2	59
#146 6-year-old girl, microcephaly and severe developmental delay. Fumarate hydratase deficiency.	7	3	1	2	55
#147 8-year-old boy. Younger sibling recently diagnosed with medium-chain acyl-CoA dehydrogenase deficiency. No excess of hexanoylglycine or suberylglycine or other abnormality.	-	2	-	1	65
#148 1-month-old boy, severe metabolic acidosis since 3 days of age. 5-Oxoprolinuria (glutathione synthase deficiency)	-	-	1	3	67
#149 4-year-old boy, developmental regression, skin rash. No abnormality.	-	1	1	3	66
#150 3-year-old girl, episodic vomiting, lethargy, hypotonia. Increased orotic acid and uracil. On benzoate therapy.	1	1	4	4	61

Table 3: Cumulative scores for 2006 and the two preceding years (current Sheffield participants only)

Laboratory Number	2006			2005		2004		2004-6	
	No of returns	Late returns	Total score	No of returns	Total score	No of returns	Total score	No of returns	Average Score*
3	3	0	18	3	17	3	17	9	5.78
4	3	0	17	3	16	2	12	8	5.63
5	3	0	18	3	11	3	15	9	4.89
6	3	0	18	3	9	3	18	9	5.00
7	3	0	17	3	10	3	14	9	4.56
9	3	0	17	3	17	3	18	9	5.78
10	3	0	15	3	18	3	17	9	5.56
11	3	0	17	3	17	3	18	9	5.78
12	3	0	18	3	18	2	12	8	6.00
13	3	0	18	3	16	3	17	9	5.67
14	3	2	18	3	17	2	12	8	5.88
15	3	0	14	3	18	3	16	9	5.33
17	3	0	12	3	15	3	13	9	4.44
18	3	1	16	3	18	3	11	9	5.00
19	3	0	18	3	14	3	18	9	5.56
21	3	1	18	2	12	3	14	8	5.50
24	3	0	16	3	17	3	18	9	5.67
25	2	0	11	3	18	3	17	8	5.75
26	3	1	14	3	16	3	18	9	5.33
27	3	0	10	2	-3	3	9	8	2.00
28	3	0	8	3	5	3	7	9	2.22
29	3	0	18	3	18	3	17	9	5.89
31	2	0	12	3	17	3	18	8	5.88
32	3	0	18	3	18	3	11	9	5.22
35	3	0	18	3	14	3	17	9	5.44
38	3	0	18	3	18	3	18	9	6.00
42	3	1	18	3	14	3	14	9	5.11
44	3	0	15	3	14	3	17	9	5.11
48	3	0	18	2	12	2	11	7	5.86
51	2	0	18	3	18	3	17	8	5.89
52	3	0	15	3	16	3	16	9	5.22
65	3	0	18	3	10	3	18	9	5.11
66	3	0	17	3	18	3	18	9	5.89
69	0	0	0	0	0	2	10	2	5.00
76	0	0	0	3	6	3	13	6	3.17
79	3	1	18	3	17	3	13	9	5.33
83	3	0	17	3	14	3	18	9	5.44
85	3	2	18	3	11	3	14	9	4.78
86	2	0	7	3	17	3	16	8	5.00
88	3	1	18	3	13	3	14	9	5.00
90	2	0	12	1	6	3	17	6	5.83
92	3	0	15	3	14	3	17	9	5.11
93	3	2	17	3	17	3	18	9	5.78
94	3	0	17	3	15	3	17	9	5.44
96	2	1	11	3	13	3	15	8	4.88

Laboratory Number	2006			2005		2004		2004-6	
	No of returns	Late returns	Total score	No of returns	Total score	No of returns	Total score	No of returns	Average Score
98	3	0	17	3	16	3	16	9	5.44
101	3	0	18	3	17	3	17	9	5.78
102	3	1	18	3	18	3	17	9	5.89
104	3	0	12	3	10	3	14	9	4.00
106	3	0	18	3	18	3	18	9	6.00
108	3	0	18	3	13	3	14	9	5.00
111	3	0	14	3	18	3	18	9	5.56
113	3	2	13	3	2	3	9	9	2.67
114	3	1	16	3	8	3	13	9	4.11
119	3	0	18	3	18	3	17	9	5.89
120	3	1	16	2	11	3	12	8	4.88
121	3	0	18	3	16	3	16	9	5.56
126	3	0	15	3	13	2	11	8	4.88
127	2	0	6	1	0	2	7	5	2.60
128	3	1	15	2	5	3	12	8	4.00
130	2	0	12	3	15	3	18	8	5.63
131	3	1	18	3	7	3	13	9	4.22
132	3	0	18	3	11	3	8	9	4.11
133	3	3	15	3	9	3	15	9	4.33
134	3	0	18	3	7	3	17	9	4.67
135	2	0	8	0	0				
136	3	1	16	3	4				
137	3	1	18	3	16				
138	3	0	13	3	4				
139	3	0	15						
140	3	1	14						
141	3	0	15						
142	3	0	16						
143	2	0	10						

*The average score is **per return received**. Every return over this period had a maximum score of 6.

Commentary

None of this year's samples presented any great difficulties. Sample # 146, from a patient with fumarate hydratase deficiency generated the lowest scores with several participants missing the diagnosis entirely due mainly to the relatively non-specific nature of the abnormalities. Paradoxically several participants lost points with the "easiest" sample, #142 which showed a gross excretion of methylmalonic acid, mainly because of inadequate advice on further investigation.

As 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. We are intending to make greater use of E-mail to circumvent such problems:

a) We will send out a group E-mail to advise when samples have been dispatched. This year we will be sending the entire set of nine samples as a single consignment, to be analysed and reported in three sets as at present.

b) We will send out an E-mail reminder to participants whose reports are outstanding after the closing dates.

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 2007 your E-mail address appears below:

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

Another small change for 2007 is that we shall be using your laboratory's ERNDIM membership number as identifier rather than the "laboratory number" series which preceded the ERNDIM scheme.

We hope that you continue to find the scheme useful.

Yours sincerely

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