



ERNDIM DPT Center Eastern Europe

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Proficiency Testing Center Eastern Europe: Annual Report 2002

1. Introduction

In 2002 this DPTC has been running as a regular ERNDIM scheme.

2. Geographical distribution of the participants

Twenty laboratories from 10 countries of Eastern, Central and Southern Europe have participated in our DPT scheme in 2002.

Country	Number of participating laboratories
Austria	2
Bulgaria	1
Croatia	1
Cyprus	1
Czech Republic	2
Germany	4
Greece	2
Poland	1
Slovak Republic	3
Switzerland	3
TOTAL	20

3. Logistic of the scheme

- 2 surveys: 2002/1 – patients A, B and C
2002/2 – patients D, E and F
- Origin of samples – 5 urines from the patients with established inborn errors of metabolism contributed by participants; the samples were checked before distribution in Institute of Inherited Metabolic Diseases in Prague.
- A common sample F from Nijmegen distributed in all 4 DPT centres was included.
- Six heat-treated urines were shipped by express courier service at RT.

- Communication between the organizers and the participants occurred by e-mail, fax and regular mail.

4. Schedule of the scheme in 2002

Sample distribution	April 15
Survey 2002/1 – results submission	May 27
Survey 2002/1 – report	June 19
Survey 2002/2 – results submission	July 22
Survey 2002/2 – report	August 16
Annual meeting of the participants	September 3
Annual report 2002	November 15

5. The receipt of samples and results

Date of receipt of samples (samples sent on April 15, 2002)

Date (reported by participants)	Number of participants	Date (reported by courier service)	Number of participants
the same day	1	the same day	1
1 day	4	1 day	9
2 days	7	2 days	6
3 days	2	3 days	3
4 days	1	not reported	1
5 days	1	-	-
not reported	4	-	-

Date of results submission

submission	2002/1	2002/2
in time	17	18
2 days delay	2	1
3 days delay	-	1
4 days delay	1	-

6. Scoring of results

Three criteria (analytical performance, interpretative proficiency and recommendations) were scored, the **total score** is calculated as a sum of these three criteria. The maximum score that could have been achieved was 6 for one sample, 18 points per survey.

Two points for analytical performance were given if the crucial analyses were performed and typical results were found (e.g. glycine conjugates in MCAD deficiency or succinylacetone in tyrosinemia I), or if concentration of critical metabolite was abnormal according to local reference ranges (e.g. 3-hydroxyisovalerate in biotinidase deficiency).

Two points for interpretative performance were given if diagnosis was fully established in respect to the urinary metabolite pattern (e.g. diagnosis of fatty acid oxidation defect or specifically of MCADD in sample A).

Two points for recommendations were given if all therapeutic advices were reported (e.g. dietary treatment with phenylalanine and tyrosine restriction / NTBC treatment / monitoring the possibility of developing hepatoma and/or recommendation of liver transplant in tyrosinemia I or avoidance of drugs that can precipitate hemolytic crisis and/or administration of free radical scavengers vitamin C and E / acidosis correction in pyroglutamic aciduria).

The overview of scoring criteria is as follows:

A	Analytical performance	Correct results of the appropriate tests	2
		Partially correct or non-standard methods	1
		Unsatisfactory or misleading	0
I	Interpretative proficiency	Good (diagnosis was established)	2
		Helpful but incomplete	1
		Misleading/wrong diagnosis	0
R	Recommendations	Complete	2
		Helpful but incomplete	1
		Unsatisfactory or misleading	0

7. Score of participants for individual samples

Survey 2002/1

Sample A MCAD				Sample B MPS VII				Sample C Tyrosinemia I			
A	I	R	Total	A	I	R	Total	A	I	R	Total
0	0	1	1	0	0	1	1	2	2	2	6
0	0	0	0	2	1	1	4	1	2	1	4
2	2	2	6	2	1	1	4	2	2	2	6
1	2	2	5	2	2	2	6	2	2	2	6
2	2	2	6	2	2	2	6	2	2	2	6
0	0	0	0	2	1	1	4	1	2	2	5
2	2	2	6	2	2	1	5	2	2	2	6
2	2	1	5	1	2	1	4	2	2	0	4
2	2	2	6	1	2	2	5	2	2	2	6
2	2	2	6	2	0	1	3	2	2	2	6
0	2	2	4	1	2	2	5	2	2	2	6
2	2	2	6	1	2	2	5	2	2	2	6
2	2	1	5	1	2	1	4	1	2	2	5
0	2	2	4	1	2	2	5	1	2	2	5
0	2	1	3	1	2	2	5	1	2	2	5
2	2	2	6	0	0	1	1	2	2	2	6
2	2	1	5	1	1	1	3	2	2	1	5
2	2	1	5	2	2	1	5	2	2	1	5
2	2	1	5	2	2	1	5	2	2	1	5
2	2	1	5	0	2	2	4	1	2	2	5

Survey 2002/2

Sample D Pyroglutamic aciduria				Sample E Argininosuccinic aciduria				Sample F Biotinidase deficiency			
A	I	R	Total	A	I	R	Total	A	I	R	Total
2	2	2	6	0	0	0	0	2	2	2	6
0	0	1	1	2	2	2	6	0	0	1	1
2	2	2	6	2	2	2	6	0	0	1	1
2	2	2	6	2	2	2	6	2	2	2	6
2	2	2	6	2	2	2	6	2	2	1	5
2	2	2	6	0	0	0	0	2	2	2	6
2	2	2	6	2	2	2	6	2	2	2	6
2	2	1	5	0	0	0	0	2	0	1	3
2	2	2	6	2	2	2	6	2	2	2	6
2	2	2	6	1	0	1	2	0	0	1	1
2	2	2	6	2	2	2	6	2	2	2	6
2	2	2	6	2	2	2	6	2	2	2	6
2	2	2	6	0	0	1	1	0	0	0	0
2	2	2	6	2	2	2	6	2	2	2	6
2	2	2	6	2	2	2	6	2	2	2	6
2	2	2	6	2	2	2	6	2	2	2	6
2	2	2	6	2	2	1	5	2	2	2	6
2	2	2	6	2	2	2	6	0	0	0	0
2	2	0	4	0	0	0	0	2	1	0	3
2	2	2	6	2	2	2	6	2	0	1	3

8. Score summary in 2002

Sample	Diagnosis	Analytical [%]	Interpretative [%]	Recommendations [%]	Total [%]
A	<i>MCAD deficiency</i>	68	85	70	74
B	<i>MPS type VII</i>	65	75	70	69
C	<i>Tyrosinemia type I</i>	85	100	85	90
D	<i>Pyroglutamic aciduria</i>	95	95	90	93
E	<i>Argininosuccinic aciduria</i>	73	70	73	72
F	<i>Biotinidase deficiency</i>	75	63	70	69

9. Performance scores for individual participants

[% of maximum achievable]

Survey 2001/2				Survey 2002/1				Survey 2002/2				Sliding window (the last 3 surveys)				Total performance (all surveys)			
A	I	R	T	A	I	R	T	A	I	R	T	A	I	R	T	A	I	R	T
100	100	83	94	33	33	67	44	67	67	67	67	67	67	72	69	64	64	68	65
17	33	50	33	50	50	33	44	33	33	67	44	33	39	50	41	36	41	50	42
67	67	50	61	100	83	83	89	67	67	83	72	78	72	72	74	82	77	77	79
83	83	83	83	83	100	100	94	100	100	100	100	89	94	94	93	91	95	91	92
100	100	83	94	100	100	100	100	100	100	83	94	100	100	89	96	100	100	73	91
67	67	67	67	50	50	50	50	67	67	67	67	61	61	61	61	68	68	68	68
100	100	100	100	100	100	83	94	100	100	100	100	100	100	94	98	100	100	95	98
83	100	83	89	83	100	33	72	67	33	33	44	78	78	50	69	73	73	45	64
100	100	83	94	83	100	100	94	100	100	100	100	94	100	94	96	95	100	95	97
100	100	100	100	100	67	83	83	50	33	67	50	83	67	83	78	86	73	86	82
67	83	83	78	50	100	100	83	100	100	100	100	72	94	94	87	77	95	91	88
100	100	83	94	83	100	100	94	100	100	100	100	94	100	94	96	95	100	95	97
83	100	100	94	67	100	67	78	33	33	50	39	61	78	72	70	64	73	64	67
100	100	100	100	33	100	100	78	100	100	100	100	78	100	100	93	82	100	100	94
100	100	100	100	33	100	83	72	100	100	100	100	78	100	94	91	82	100	95	92
83	83	83	83	67	67	83	72	100	100	100	100	83	83	89	85	86	86	91	88
100	100	100	100	83	83	50	72	100	100	83	94	94	94	78	89	95	95	73	88
100	100	83	94	100	100	50	83	67	67	67	67	89	89	67	81	91	91	68	83
-	-	-	-	100	100	50	83	67	50	0	39	83	75	25	61	83	75	25	61
-	-	-	-	50	100	83	78	100	67	83	83	75	83	83	81	75	83	83	81

At the previous meeting in Prague in 2001 the participants agreed that the overall performance will be scored for the last 3 surveys and that 75% and more of the maximum achievable score will be considered good performance (poor performers according to this criteria are shown in red and blue numbers in the table above).

10. Annual meeting of the participants

The participants met at the 40th Annual Symposium of SSEIM, 3rd September 2002, 9.30-11.00, Dublin, Ireland, for details see the minutes of meeting (reported in October 2002)

11. Organization and scoring of DPT in 2003

The harmonisation of DPT schemes in all four DPT centres is in progress. Starting in 2003 six samples will be distributed yearly, heat-treated urines will be shipped at ambient temperature. Also normal samples may be included (real clinical samples with no known inborn error of metabolism). In all 4 centres the results should be reported within 3 weeks after receipt of samples. New evaluation and scoring system will be implemented in 2003 (in general, it is a modified version of the scoring system used in our DPTC). Three criteria will be evaluated: analytical, interpretative and recommendations for further investigations. Due to the large variability in reporting results in

various countries recommendations pertaining to treatment will not be evaluated in proficiency testing, however, they can still be reported and summarized by the scheme organizers. The new system will be evaluated and criteria for poor performers will be established within approximately 2 years.

The application forms for participation in QA Schemes were distributed from ERNDIM in October. If you did not receive it, please contact the Treasurer Dr. J R Bonham or the Assistant to Treasurer Mr. Malcolm Heron (mjheron@onetel.net.uk). The Executive Board has set the fee for participation in Proficiency testing scheme in 2003 to 250 Euro, the fee is payable to the Treasurer of ERNDIM.

12. Tentative schedule of DPT scheme in 2003

Samples distribution	February 18, 2003
Survey 2003/1 – results submission	March 11, 2003
Survey 2003/1 – report Survey on participation in ICIEM in Australia	April 15, 2003
Survey 2003/2 – results submission	June 3, 2003
Survey 2003/2 – report	July 15, 2003
Annual meeting of participants (Australia or Prague)	? September ?, 2003
Annual report 2003	November 15, 2003

Next meeting of the scheme participants

Two possible dates were suggested. The annual meeting of DPT scheme participants can take place in September 2003 in Brisbane, Australia (ICIEM) or at another date in Prague. The time and place of the next meeting will be specified during April 2003 according to a number of participants, who would be able to take part at ICIEM in Australia.

Prague, November 12, 2002

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