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Qualitative Organic Acids
Centre: United Kingdom

Final Report 2025

prepared by
Mrs C Scott and Miss S Colyer

Note: This annual report is intended for participants of the ERNDIM QLOU Sheffield scheme. The contents should not be used for any publication without permission of the Scientific Advisor.

The fact that your laboratory participates in ERNDIM schemes is not confidential, however, the raw data and performance scores are confidential and will only be shared within ERNDIM for the purpose of evaluating your laboratories performance, unless ERNDIM is required to disclose performance data by a relevant government agency. For details, please see the ERNDIM Privacy Policy on www.erndim.org

1. Introduction

The ERNDIM Qualitative Organic Acids in urine scheme offers urine samples obtained from patients with confirmed diagnoses to enable laboratories to gain or maintain experience to identify organic acid disorders. The scheme has been designed and planned by Camilla Scott as Scientific Advisor, Sharon Colyer as Deputy Scientific Advisor, and coordinated by Alessandro Salemma as scheme organiser (sub-contractor on behalf of CSCQ), both appointed by and according to procedures laid down the ERNDIM Board.
As in previous years, samples were sent out to cover the spectrum of what is typically observed in the metabolic laboratory. A mix of clearly diagnostic profiles and some more challenging profiles were provided. As in previous years, normal profiles were also sent out. The requirement to interpret a normal profile, as such, is as important as correctly identifying abnormal profiles. Correctly identifying a profile as normal can avoid unnecessary further investigation and distress to the patient and family.

2. Geographical distribution of participants

In 2025, 75 labs participated to the Qualitative Organic Acid Scheme, Sheffield.

For the first survey, 73/75 and second survey 73/75 laboratories submitted results.

[1] If this report is not Version 1 for this scheme year, go to APPENDIX 1 for details of the changes made since the last version of this document.

Country	Number of participants
Australia	6
Azerbaijan	1
Belgium	8
Finland	1
Hungary	1
Ireland	1
Israel	3
Japan	6
Malaysia	3
New Zealand	1
Norway	1
Pakistan	1
Poland	4
Slovakia	1
South Africa	2
Spain	1
Sweden	2
United Kingdom	17
United States	15

3. Design and logistics of the scheme including sample information.

CSCQ dispatches QLOU EQA samples to the scheme participants and provides a website for on-line submission of results and access to scheme reports. Existing QLOU scheme participants can log on to the CSCQ results submission website at:
https://cscq.hcuge.ch/cscq/ERNDIM/Initial/Initial.php

2 surveys	Round 1: patients A, B and C
	Round 2: patients D, E and F

Origin of patient samples: all urine samples have been provided by the scheme organizers.

The samples used in 2025 were human urine samples. Four patient sample donations, and one control sample donation were collected through Sheffield Children's NHS Foundation Trust, Sheffield UK. A sample was also selected and donated by the Scientific Advisor for QLOU-Heidelberg, Joachim Janda through the Centre for Metabolic Diseases Heidelberg, Heidelberg, Germany.

The samples have been heat-treated. They were pre-analysed in the Scientific Advisors institute after 3 days incubation at ambient temperature (to mimic possible changes that might arise during transport). In all six samples the typical metabolic profiles were preserved after this process. Samples were shipped by DHL, FedEx or the Swiss Post at room temperature. Details regarding stability of (reconstituted) samples are provided in the sample package. The samples are stable for the duration of the scheme's submission calendar when stored under defined conditions.

4. Tests

Analysis of qualitative organic acids.

5. Schedule of the scheme

- Feb 5, 2025: shipment of samples by CSCQ
- May 6, 2025: analysis start and website submission 1st round (A-C)
- May 27, 2025: results submission deadline
- August 18, 2025: analysis start and website submission 2nd round (D-F)
- Sep 8, 2025: deadline for result submission (Survey 2)
- July 15, 2025: report of Survey 1 by e-mail
- November 6, 2025: report of Survey 2 by e-mail
- January 31, 2026: annual report with scoring.

6. Results

	Survey 1	Survey 2
Receipt of results	71	72
No answer	4	3

7. Web site reporting

The website reporting system is compulsory for all centres. Please carefully read the following advice:

- Results
  - Enter the key metabolites with the evaluation **in the tables.**
  - If the profile is normal: enter “Normal profile” in “Key metabolites”.
  - **Don’t enter results in the “comments” window**, **otherwise your results may not be included in the evaluation program.**
- Recommendations = **advice for further investigation**.
  - Scored together with the interpretative score.
  - Advice for treatment are not scored but may be used in the overall assessment.
  - **Don’t give advice for further investigation in “Comments on diagnosis”**: it may not be included in the evaluation program.
  - Enter the key metabolites with the evaluation **in the tables** even if you don’t give quantitative data.
  - If the profile is normal: enter “Normal profile” in “Key metabolites”.
  - **Don’t enter results in the “comments” window**, **otherwise your results may not be included in the evaluation program.**

8. Scoring and evaluation of results

The scoring system has been established by the International Scientific Advisory Board of ERNDIM. Two criteria are evaluated: 1) analytical performance, 2) interpretative proficiency also considering recommendations for further investigations.

The total score is calculated as a sum of these two criteria. The maximum to be achieved is 4 points per sample. The scores were calculated only for laboratories submitting results. For further information, please refer to the Framework for Assessment and Education for Qualitative Schemes on our website (https://eqa.erndim.org/information/view/14)

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

Scoring and certificate of participation: scoring is carried by a second assessor who changes every year as well as by the scientific advisor. The results of QLOU US have also been second scored by Joachim

Janda, QLOU SA, Heidelberg. At the SAB meeting in November 2025 the definitive scores were finalised. The concept of critical error was introduced in 2014. A critical error is defined as an error resulting from seriously misleading analytical findings and /or interpretations with serious clinical consequences for the patient. Thus, labs failing to make a correct diagnosis of a sample considered as eligible for this category will be deemed not to have reached a satisfactory performance even if their total points for the year exceed the limit set at the SAB. For 2025, the SAB decided that critical error would be awarded for sample D (Fumarase Deficiency) and for sample E (SSADH) if the diagnosis was missed.

A certificate of participation will be issued for participation, and it will be additionally notified whether the participant has received a performance support letter. This performance support letter is sent out if the performance is evaluated as unsatisfactory. Four performance support letters will be sent by the Scheme Advisor for 2025. Four critical error letters will be sent out for this scheme for 2025. Any partial submitters will receive a letter from the ERNDIM Executive Administrator, Dr Sara Gardner.

8.1. Score for satisfactory performance

At least 14 points from the maximum of 20 (70%). Sample F was deemed educational, and scores will not be counted towards performance for this sample.

9. Results of samples and evaluation of reporting

9.1. QLOU US 2025 Sample A Hereditary Tyrosinaemia Type 1

Patient details provided to participants.
Unwell, liver transplant when aged 5 years old. 40year old male at presentation.

Patient details
This sample was donated by a patient diagnosed with Tyrosinaemia Type 1 (Fumarylacetoacetase (FAH gene)) who had undergone therapeutic liver transplantation in childhood. He had been lost to follow up and presented to the local hospital acutely unwell. He presented at aged 40 years and is male.

Analytical performance

	Number of labs:
Succinyl acetone	40
No abnormality	31

Diagnosis / Interpretative proficiency

	Number of labs:
Tyrosinaemia Type 1	41
Normal	30

Recommendations
Molecular analysis *FAH* gene, liver function tests, clotting screen. Plasma and urine amino acids.

Scoring
Analytical:

Succinyl acetonte	2 points

Clinical:

Tyrosinaemia Type one (with succinyl acetone)	2 points

Overall impression

This was a difficult sample due to the low excretion of the succinyl acetone however as the detection of this metabolite was key to the diagnosis in this patient it was decided at the recent Scientific Advisory Board that this sample should be scored for performance. This sample however would not be considered for critical error.

9.2. QLOU US 2025 Sample B. Isolated 3-Methylcrotonyl-CoA carboxylase deficiency (3MCC)

Patient details provided to participants.
Diagnosed in childhood. Adult – 37-year-old now well.

Patient details.

This sample was donated from a 58-year-old man with isolated 3-Methylcrotonyl-CoA carboxylase deficiency. He was diagnosed following family studies and had no health concerns.

Analytical performance

	Number of labs:
3-methylcrotonyl glycine	65
3-hydroxy isovaleric acid	70

Diagnosis / Interpretative proficiency

	Number of labs:
3-Methylcrotonyl-CoA carboxylase deficiency	66
Biotinidase/Holocarboxylase Deficiency	4

Recommendations
Recommendations ranged from follow up acylcarnitine studies, biotinidase activity and specific mutational analysis of the various genes including MCCC1 and MCCC2 genes

Scoring
Analytical:

3-methylcrotonyl glycine	1 point
3-hydroxy isovaleric acid	1 point

Clinical:

3MCC	2 points
Biotinidase/Holocarboxylase	1 point

Overall impression
This was a good scoring survey with most participants picking up the abnormal metabolites. Points however were lost if participants did not consider isolated 3-Methylcrotonyl-CoA carboxylase deficiency as a diagnosis. Because of the relatively benign nature of this condition, it was decided at the Scientific Advisory Board that this sample would not be considered for critical error.

9.3. QLOU US 2025 Sample C. Secondary pyroglutamic aciduria

Patient details provided to participants.
Unexplained metabolic acidosis. Sample taken whilst on the Intensive Care Unit. Male 51 years old.

Patient details
This sample was donated from a critically ill patient presenting with an unexplained metabolic acidosis on a background of well managed previously diagnosed Classical Homocystinuria

Analytical performance

	Number of labs:
Pyroglutamic acid	70

Diagnosis / Interpretative proficiency
Clinical:

	Number of labs:
Acquired Pyroglutamic aciduria	41
Glutathione synthase deficiency	17
5-Oxoprolinase deficiency	5

Recommendations
Recommendations included medication and dietary review. Molecular analysis of the GSS and OPLAH genes if acquired causes have been excluded.

Scoring
Analytical:

Increased pyroglutamate	2 points

Clinical:

Pyroglutamic aciduria	2 points

Overall impression

Most participants did suggest acquired pyroglutamic aciduria secondary to depletion of glutathione stores as a cause of the increased pyroglutamic acid in this patient. It is important to consider acquired pyroglutamic aciduria, particularly in the acutely unwell patient, as treatment requires prompt removal of glutathione-depleting medication and further extensive testing may not be required. Marks were given if secondary causes were included in the report. It was decided at the Scientific Advisory Board that this sample would not be considered for critical error due to the secondary origin of the pyroglutamic acid.

9.4. QLOU US 2025 Sample D. Fumarate Hydratase Deficiency

Patient details provided to participants.
Congenital lactic acidosis, male 4 weeks old.

Patient details
This is a historical sample donated from a patient with a confirmed diagnosis of Fumarate Hydratase deficiency.

Analytical performance

	Number of labs:
Fumaric acid	71

Diagnosis / Interpretative proficiency

	Number of labs:
Fumarate Hydratase deficiency	59
Mitochondrial respiratory chain	10
Normal	2

Recommendations
Molecular analysis of the Fumarate Hydratase gene. Enzymatic studies.

Scoring
Analytical:

Fumarate	2 points

Clinical:

Fumarate Hydratase deficiency (most likely or alternative diagnosis)	2 points

Overall impression
Most participants identified the increased fumaric acid. Because the profile also contained a small amount of malate (origin unknown) diagnostic interpretation also centred around mitochondrial disease as well as Fumarate Hydratase Deficiency. Participants were not awarded critical error if mitochondrial disease was given in the diagnosis. Critical error was awarded in the cases of no abnormality reported.

9.5. QLOU US 2025 Sample E. Succinic Semialdehyde Dehydrogenase Deficiency (SSADH).

9.6.

Patient details provided to participants.
Developmental delay and aggressive behaviour, female 3 years old.

Patient details
This historical sample was donated from a patient diagnosed with Succinic Semialdehyde Dehydrogenase Deficiency (SSADH).

Analytical performance

	Number of labs:
4-hydroxybuyrate	70
5-oxoproline (in isolation)	2
Normal	1

Diagnosis / Interpretative proficiency

	Number of labs:
SSADH	69
5-oxoprolinuria	2
Normal	1

Recommendations
Molecular analysis of the *ALDH5A1* gene.

Analytical:

4-hydroxybutyrate	2 points

Interpretative:

SSADH	2 points

Overall impression
This was a high scoring survey with most participants identifying the correct metabolites and reaching the correct diagnosis. It was agreed at the Scientific Advisory Board that participants that did not reach the correct diagnosis would be awarded Critical Error.

9.6 QLOU US 2025 Sample F. Glycerol contamination from Slush consumption in a child without an underlying metabolic defect.

Patient details provided to participants.
Very lethargic post a visit to a play centre. Male 3 years old.

Patient details
This sample was donated by a healthy child who had consumed a slush style drink earlier in the day.

Analytical performance

	Number of labs:
Increased glycerol	40

Diagnosis / Interpretative proficiency

Secondary cause of glycerol	27
Glycerol kinase deficiency	10
Fructose 1-6 bisphosphatase deficiency	6
Glycerol encephalopathy	13

Recommendations
A repeat fresh sample. Discuss with the clinical team.

Scoring
This sample was not scored.

Overall impression

This was a difficult sample due to the patient having no known metabolic disease with a moderate peak of glycerol present on the organic acid profile. It was approved as educational at the SAB in November 2025. Subsequently scores will not be assigned, and this sample will be removed from performance criteria. For education purposes the scientific advisors draw participants attention to the possibility of glycerol intoxication syndrome which is an emerging condition often presenting secondary to consumption of slush style drinks.

10. Scores of participants

All data transfer, the submission of data as well as the request and viewing of reports proceed via the QLOU-CSCQ results website. The results of your laboratory are confidential and only accessible to you (with your username and password). The anonymous scores of all laboratories are accessible to all participants in the table below.

If your laboratory is assigned poor performance and you wish to appeal against this classification, please email the ERNDIM Administration Office (admin@erndim.org), with full details of the reason for your appeal, within one month receiving your Performance Support Letter. Details of how to appeal poor performance are included in the Performance Support Letter sent to poor performing laboratories.

Detailed scores – Round 1

Lab n°	Patient A Tyrosinaemia type 1 (treated).			Patient B 3MCC			Patient C Acquired pyroglutamic aciduria secondary to glutathione depletion.			
	A	I	Total	A	I	Total	A	I	Total	Total
1	2	2	4	1	0	1	2	2	4	9
2	2	2	4	1	1	2	2	2	4	10
3	2	2	4	2	2	4	2	2	4	12
4	2	2	4	2	2	4	2	2	4	12
5	0	0	0	2	2	4	2	0	2	6
6	2	2	4	2	2	4	2	2	4	12
7	2	2	4	2	2	4	2	2	4	12
8	0	0	0	2	2	4	2	2	4	8
9	2	2	4	1	2	3	2	1	3	10
10	0	0	0	0	0	0	0	0	0	0
11	2	2	4	2	2	4	2	2	4	12
12	2	2	4	2	2	4	2	2	4	12
13	0	0	0	2	2	4	2	2	4	8
14	2	2	4	2	2	4	2	2	4	12
15	0	0	0	2	2	4	2	2	4	8
16	0	0	0	2	2	4	2	2	4	8
17	0	0	0	2	2	4	2	2	4	8
18	2	2	4	2	2	4	2	2	4	12
19	2	2	4	2	2	4	2	2	4	12
20	0	0	0	2	2	4	2	2	4	8
21	2	2	4	2	1	3	2	2	4	11
22	2	2	4	1	1	2	2	2	4	10

Lab n°	Patient A Tyrosinaemia type 1 (treated).			Patient B 3MCC			Patient C Acquired pyroglutamic aciduria secondary to glutathione depletion.			
	A	I	Total	A	I	Total	A	I	Total	Total
23	2	2	4	2	2	4	2	2	4	12
24	2	2	4	2	2	4	2	2	4	12
25	2	2	4	2	2	4	2	2	4	12
26	2	2	4	2	2	4	2	2	4	12
27	0	0	0	1	2	3	2	2	4	7
28	0	0	0	2	2	4	2	2	4	8
29	2	2	4	2	2	4	2	2	4	12
30	2	2	4	2	2	4	2	2	4	12
31	2	2	4	2	2	4	2	2	4	12
32	0	0	0	2	2	4	2	2	4	8
33	2	2	4	2	2	4	2	2	4	12
34	0	0	0	2	2	4	2	2	4	8
35	2	2	4	2	2	4	2	2	4	12
36	0	0	0	2	2	4	2	2	4	8
37	2	2	4	2	2	4	2	2	4	12
38	0	0	0	2	2	4	2	1	3	7
39	0	0	0	2	2	4	2	2	4	8
40	2	2	4	2	2	4	2	2	4	12
41	0	0	0	2	2	4	2	2	4	8
42	0	1	1	0	0	0	2	2	4	5
43	0	0	0	2	2	4	2	2	4	8
44	2	2	4	2	2	4	2	2	4	12
45	2	2	4	2	2	4	2	2	4	12
46	0	0	0	2	2	4	2	1	3	7
47	2	2	4	2	2	4	2	2	4	12
48	2	2	4	2	2	4	2	2	4	12
49	2	2	4	2	2	4	2	2	4	12
50	2	2	4	2	2	4	2	0	2	10
51	2	2	4	2	2	4	2	2	4	12
52	0	0	0	2	2	4	2	2	4	8
53	0	0	0	2	2	4	2	2	4	8

Lab n°	Patient A Tyrosinaemia type 1 (treated).			Patient B 3MCC			Patient C Acquired pyroglutamic aciduria secondary to glutathione depletion.			
	A	I	Total	A	I	Total	A	I	Total	Total
54	2	2	4	2	2	4	2	2	4	12
55	2	2	4	2	2	4	2	2	4	12
56	0	0	0	2	2	4	2	2	4	8
57	0	0	0	2	2	4	2	2	4	8
58	2	2	4	2	2	4	2	2	4	12
59	0	0	0	0	0	0	0	0	0	0
60	0	0	0	2	2	4	2	0	2	6
61	0	0	0	1	2	3	2	1	3	6
62	0	0	0	2	2	4	2	0	2	6
63	0	0	0	2	2	4	2	2	4	8
64	0	0	0	2	2	4	2	2	4	8
65	2	2	4	2	2	4	2	2	4	12
66	0	0	0	2	2	4	2	2	4	8
67	2	2	4	2	2	4	2	2	4	12
68	2	2	4	2	2	4	2	2	4	12
69	0	0	0	1	2	3	2	2	4	7
70	0	0	0	2	2	4	2	2	4	8
71	2	2	4	2	2	4	2	0	2	10
72	2	2	4	2	2	4	2	2	4	12
73	0	0	0	0	0	0	0	0	0	0
74	0	0	0	2	2	4	0	0	0	4
75	0	0	0	0	0	0	0	0	0	0

Detailed scores – Round 2

Lab n°	Patient D Fumarate Hydratase (FH) deficiency.			Patient E SSADH			Patient F A profile with a small peak of glycerol which may be exogenous but requires further follow up samples.			
	A	I	Total	A	I	Total	A	I	Total	Total
1	2	2	4	2	2	4	--	--	--	8
2	2	2	4	2	2	4	--	--	--	8
3	2	2	4	2	2	4	--	--	--	8
4	2	2	4	2	2	4	--	--	--	8
5	2	2	4	2	2	4	--	--	--	8
6	2	2	4	2	2	4	--	--	--	8
7	2	2	4	2	2	4	--	--	--	8
8	2	2	4	2	2	4	--	--	--	8
9	2	2	4	2	2	4	--	--	--	8
10	2	2	4	2	2	4	--	--	--	8
11	2	0	2	2	2	4	--	--	--	6
12	2	2	4	2	2	4	--	--	--	8
13	2	2	4	2	2	4	--	--	--	8
14	2	2	4	2	2	4	--	--	--	8
15	2	2	4	2	2	4	--	--	--	8
16	2	0	2	2	2	4	--	--	--	6
17	2	2	4	2	2	4	--	--	--	8
18	2	2	4	2	2	4	--	--	--	8
19	2	2	4	2	2	4	--	--	--	8
20	2	2	4	2	2	4	--	--	--	8
21	2	2	4	2	2	4	--	--	--	8
22	2	2	4	2	2	4	--	--	--	8
23	2	2	4	2	2	4	--	--	--	8
24	2	2	4	2	2	4	--	--	--	8
25	2	2	4	2	2	4	--	--	--	8
26	2	0	2	2	2	4	--	--	--	6
27	2	2	4	2	2	4	--	--	--	8
28	2	2	4	2	2	4	--	--	--	8
29	2	2	4	2	2	4	--	--	--	8

Lab n°	Patient D Fumarate Hydratase (FH) deficiency.			Patient E SSADH			Patient F A profile with a small peak of glycerol which may be exogenous but requires further follow up samples.			
	A	I	Total	A	I	Total	A	I	Total	Total
30	2	2	4	2	2	4	--	--	--	8
31	2	2	4	2	2	4	--	--	--	8
32	2	2	4	2	2	4	--	--	--	8
33	2	2	4	2	2	4	--	--	--	8
34	2	2	4	2	2	4	--	--	--	8
35	2	2	4	2	2	4	--	--	--	8
36	2	0	2	2	2	4	--	--	--	6
37	2	2	4	2	2	4	--	--	--	8
38	0	0	0	2	2	4	--	--	--	4
39	2	2	4	2	2	4	--	--	--	8
40	2	2	4	2	2	4	--	--	--	8
41	2	0	2	2	2	4	--	--	--	6
42	2	2	4	2	2	4	--	--	--	8
43	2	0	2	2	2	4	--	--	--	6
44	2	2	4	2	2	4	--	--	--	8
45	2	2	4	2	2	4	--	--	--	8
46	2	2	4	2	2	4	--	--	--	8
47	2	2	4	2	2	4	--	--	--	8
48	2	2	4	2	2	4	--	--	--	8
49	2	2	4	2	2	4	--	--	--	8
50	2	2	4	2	2	4	--	--	--	8
51	2	2	4	2	2	4	--	--	--	8
52	2	2	4	2	2	4	--	--	--	8
53	2	2	4	2	2	4	--	--	--	8
54	2	2	4	2	2	4	--	--	--	8
55	2	0	2	2	2	4	--	--	--	6
56	2	2	4	2	2	4	--	--	--	8
57	2	2	4	2	2	4	--	--	--	8
58	2	2	4	2	2	4	--	--	--	8
59	0	0	0	0	0	0	--	--	--	0
60	2	2	4	0	0	0	--	--	--	4

Lab n°	Patient D Fumarate Hydratase (FH) deficiency.			Patient E SSADH			Patient F A profile with a small peak of glycerol which may be exogenous but requires further follow up samples.			
	A	I	Total	A	I	Total	A	I	Total	Total
61	2	2	4	2	2	4	--	--	--	8
62	2	2	4	2	2	4	--	--	--	8
63	2	0	2	2	2	4	--	--	--	6
64	2	2	4	2	2	4	--	--	--	8
65	0	0	0	0	0	0	--	--	--	0
66	2	2	4	2	2	4	--	--	--	8
67	2	2	4	0	0	0	--	--	--	4
68	2	2	4	2	2	4	--	--	--	8
69	2	2	4	2	2	4	--	--	--	8
70	2	2	4	2	2	4	--	--	--	8
71	2	2	4	2	2	4	--	--	--	8
72	2	2	4	2	2	4	--	--	--	8
73	0	0	0	0	0	0	--	--	--	0
74	2	0	2	0	0	0	--	--	--	2
75	2	2	4	2	2	4	--	--	--	8

Total scores

Lab n°	A	B	C	D	E	F	Cumulative score	Cumulative score (%)
1	4	1	4	4	4	--	17	85
2	4	2	4	4	4	--	18	90
3	4	4	4	4	4	--	20	100
4	4	4	4	4	4	--	20	100
5	0	4	2	4	4	--	14	70
6	4	4	4	4	4	--	20	100
7	4	4	4	4	4	--	20	100
8	0	4	4	4	4	--	16	80
9	4	3	3	4	4	--	18	90
10	0	0	0	4	4	--	8	40
11	4	4	4	2	4	--	18	90
12	4	4	4	4	4	--	20	100
13	0	4	4	4	4	--	16	80
14	4	4	4	4	4	--	20	100
15	0	4	4	4	4	--	16	80
16	0	4	4	2	4	--	14	70
17	0	4	4	4	4	--	16	80
18	4	4	4	4	4	--	20	100
19	4	4	4	4	4	--	20	100
20	0	4	4	4	4	--	16	80
21	4	3	4	4	4	--	19	95
22	4	2	4	4	4	--	18	90
23	4	4	4	4	4	--	20	100
24	4	4	4	4	4	--	20	100
25	4	4	4	4	4	--	20	100
26	4	4	4	2	4	--	18	90
27	0	3	4	4	4	--	15	75
28	0	4	4	4	4	--	16	80
29	4	4	4	4	4	--	20	100
30	4	4	4	4	4	--	20	100
31	4	4	4	4	4	--	20	100
32	0	4	4	4	4	--	16	80
33	4	4	4	4	4	--	20	100

Lab n°	A	B	C	D	E	F	Cumulative score	Cumulative score (%)
34	0	4	4	4	4	--	16	80
35	4	4	4	4	4	--	20	100
36	0	4	4	2	4	--	14	70
37	4	4	4	4	4	--	20	100
38	0	4	3	0	4	--	11	55
39	0	4	4	4	4	--	16	80
40	4	4	4	4	4	--	20	100
41	0	4	4	2	4	--	14	70
42	1	0	4	4	4	--	13	65
43	0	4	4	2	4	--	14	70
44	4	4	4	4	4	--	20	100
45	4	4	4	4	4	--	20	100
46	0	4	3	4	4	--	15	75
47	4	4	4	4	4	--	20	100
48	4	4	4	4	4	--	20	100
49	4	4	4	4	4	--	20	100
50	4	4	2	4	4	--	18	90
51	4	4	4	4	4	--	20	100
52	0	4	4	4	4	--	16	80
53	0	4	4	4	4	--	16	80
54	4	4	4	4	4	--	20	100
55	4	4	4	2	4	--	18	90
56	0	4	4	4	4	--	16	80
57	0	4	4	4	4	--	16	80
58	4	4	4	4	4	--	20	100
59	0	0	0	0	0	--	0	0
60	0	4	2	4	0	--	10	50
61	0	3	3	4	4	--	14	70
62	0	4	2	4	4	--	14	70
63	0	4	4	2	4	--	14	70
64	0	4	4	4	4	--	16	80
65	4	4	4	0	0	--	12	60
66	0	4	4	4	4	--	16	80
67	4	4	4	4	0	--	16	80

Lab n°	A	B	C	D	E	F	Cumulative score	Cumulative score (%)
68	4	4	4	4	4	--	20	100
69	0	3	4	4	4	--	15	75
70	0	4	4	4	4	--	16	80
71	4	4	2	4	4	--	18	90
72	4	4	4	4	4	--	20	100
73	0	0	0	0	0	--	0	0
74	0	4	0	2	0	--	6	30
75	0	0	0	4	4	--	8	40

Performance

	Number of labs	% total labs
Satisfactory performers (≥ 70 % of adequate responses)	67	89
Unsatisfactory performers (< 70 % adequate responses and/or critical error)	5	6
Partial and non-submitters	3	4

Overall Proficiency

Sample	Diagnosis	Proficiency Total (%)
QLOU US A	Hereditary Tyrosinaemia Type 1	**56**
QLOU US B	Isolated 3-Methylcrotonyl-CoA carboxylase deficiency (3MCC)	**95**
QLOU US C	Secondary pyroglutamic aciduria	**95**
QLOU US D	Fumarate Hydratase Deficiency	**92**
QLOU US E	Succinic Semialdehyde Dehydrogenase Deficiency (SSADH).	**94**
QLOU US F	Glycerol contamination in a child without an underlying metabolic defect	**n/a**

11. Information from the Executive Board and the Scientific Advisory Board

- **Urine samples**: we remind you that every year, we make an urgent request to participants to donate to the scheme organizer at least 200 ml of urine from a patient affected with an established inborn error of metabolism or “normal” urine, together with a short clinical report. Each urine sample must be collected from a single patient (don’t send urine spiked with pathological compounds). Please don’t send a pool of urines, except if urine has been collected on a short period of time from the same patient.

  As soon as possible after collection, the urine sample must be heated at 50 °C for 20 minutes. Make sure that this temperature is achieved in the entire urine sample, not only in the water bath. Then aliquot the sample in 10 ml plastic tubes (minimum 20 tubes), add stoppers and freeze. Send the aliquots frozen by rapid mail or express transport to:

  Mrs C Scott and Miss S Colyer NHS
  Department of Clinical Chemistry and Newborn Screening
  The Children’s Hospital
  Sheffield
  S10 2TH
  United Kingdom

  Please send us an e-mail on the day you send the samples.

12. Tentative schedule and 2026

Sample distribution	4th February 2026
Start of analysis of Survey 2026/1 Website open	5th May 2026
Survey 2026/1 - Results submission	26th May 2026
Survey 2026/1 - Reports	June 2026
Start of analysis of Survey 2026/2 Website open	17th August 2026
Survey 2026/2 – Results submission	31st August 2026
Survey 2026/2 - Reports	October 2026
Annual Report 2026	January-March 2027

13. ERNDIM certificate of participation

A combined certificate of participation covering all EQA schemes will be provided to all participants who take part in any ERNDIM scheme.

14. Questions, Comments and Suggestions

If you have any questions, comments or suggestions please address to the Scientific Advisor of the scheme, Mrs C Scott and/or to the ERNDIM Administration Office (admin@erndim.org).

Most complaints received by ERNDIM consist of minor misunderstandings or problems with samples, which can usually be resolved via direct contact with the ERNDIM administrative staff. If you wish to file a formal complaint, please email your complaint with details of your issue to admin@erndim.org or contact us through our website at https://www.erndim.org/contact-us/

Date of report, 2026-01-31
Name and signature of Scientific Advisor

Mrs C Scott and Miss S Colyer
NHS
Department of Clinical Chemistry and Newborn Screening
The Children's Hospital Sheffield
S10 2TH
United Kingdom

APPENDIX 1. Change log (changes since the last version)

Version Number	Published	Amendments
1	3rd February 2026	2025 annual report published

END