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Qualitative Organic Acids
Centre: Germany

Final Report 2025

prepared by
Dr. J. Janda

Note: This annual report is intended for participants of the ERNDIM Qualitative Organic Acids in urine scheme. The contents should not be used for any publication without permission of the Scientific Advisor.

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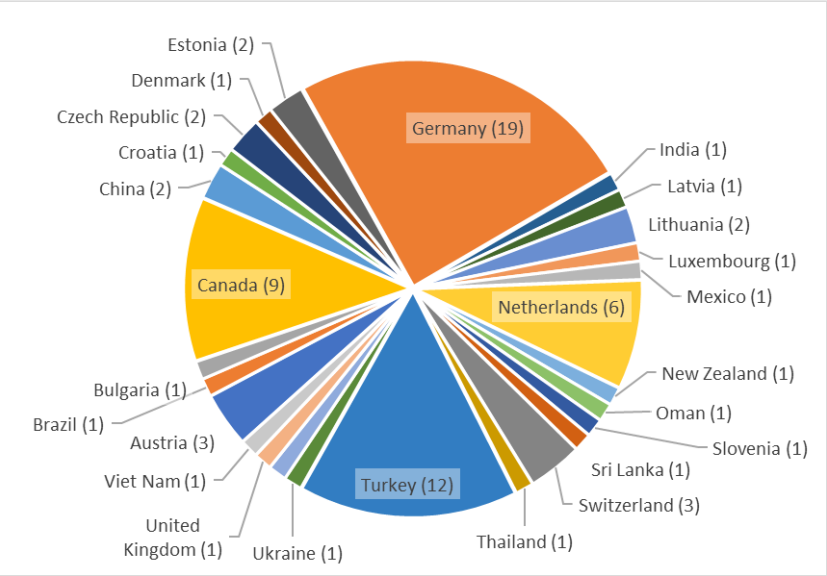
1. Introduction

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 is organised by Dr Joachim Janda (Metabolic Center Heidelberg) in conjunction with Centre Suisse de Contrôle de Qualité (CSCQ, the Swiss organisation for quality assurance in medical laboratories) 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, seventy-seven laboratories from various countries participated in the QLOU Heidelberg scheme. There was no educational participant in 2025 (as in 2024). Educational participants take part in all aspects of the scheme and receive interim reports with scores. However, performance is not indicated on the ERNDIM certificate for them.
Participants and new applicants are distributed between the Barcelona, Heidelberg and Sheffield qualitative urinary organic acid schemes which are run separately. The three organising laboratories each participate in the other's scheme by rotation.

[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
Undefined country	1
Austria	3
Brazil	1
Bulgaria	1
Canada	9
China	2
Croatia	1
Czech Republic	2
Denmark	1
Estonia	2
Germany	19
India	1
Latvia	1
Lithuania	2

Country	Number of participants
Luxembourg	1
Mexico	1
Netherlands	6
New Zealand	1
Oman	1
Slovenia	1
Sri Lanka	1
Switzerland	3
Thailand	1
Turkey	12
Ukraine	1
United Kingdom	1
Viet Nam	1

3. Design and logistics of the scheme including sample information

As usual, the samples used in 2025 were authentic human urine samples, five from affected patients and one from a healthy individual. All samples selected by the Scientific Advisor have been heat-treated and were tested for suitability in the Scientific Advisor's laboratory.
CSCQ dispatched the QLOU EQA samples to the scheme participants and provides a website for online submission of results and access to scheme reports. Existing QLOU, ACDB, DPT and Urine MPS scheme participants can log on to the CSCQ results submission website at:
https://cscq.hcuge.ch/cscq/ERNDIM/Initial/Initial.php

Participants are also encouraged to make use of the option to upload labelled copies of scans and/or chromatograms on the CSCQ website together with their analytical and interpretative results.

4. Schedule of the scheme

Time schedule in the 2025 ERNDIM QLOU Heidelberg scheme.

	1st Submission Round	2nd Submission Round
Sample IDs	QLOU-DH-2025-**A** QLOU-DH-2025-**B** QLOU-DH-2025-**C**	QLOU-DH-2025-**D** QLOU-DH-2025-**E** QLOU-DH-2025-**F**
Shipment of samples	07 February 2025	
Start of analysis (clinical data available)	06 May 2025	18 August 2025
Reminder for result submission	20 May 2025	01 September 2025
Results submission deadline	27 May 2025	08 September 2025
Interim reports available on CSCQ website	04 June 2025	16 September 2025

Samples included in the 2025 ERNDIM QLOU Heidelberg scheme.

Survey	Sample	Diagnosis
25-05-OUH	QLOU-DH-2025-A	Propionyl-CoA carboxylase deficiency
	QLOU-DH-2025-B*	3-Methylcrotonyl-CoA carboxylase (3-MCC) deficiency
	QLOU-DH-2025-C	Mevalonate kinase deficiency
25-08-OUH	QLOU-DH-2025-D	Methylmalonic aciduria mut(0) type
	QLOU-DH-2025-E	3-Hydroxy-3-methylglutaryl-CoA lyase deficiency
	QLOU-DH-2025-F	Normal control

**common sample for all centres in 2025*

The scheme format was kept identical to those of previous years. All samples selected by the Scientific Advisor have been heat-treated and were tested for suitability in the Scientific Advisor's laboratory before and after shipping process. Samples were sent by DHL; FedEx or the Swiss Post at room temperature. The samples are stable for the duration of the scheme's submission calendar when stored under defined conditions. Interim reports were generated by the evaluation program developed by CSCQ.

Origin of patients: All urine samples have been provided by the scheme organizers or specified participants.

Patient A:	Propionic aciduria, PA	Metabolic Center Heidelberg
Patient B:	3-MCC deficiency*	
Patient C:	Mevalonic aciduria	
Patient D:	MMA mut0	
Patient E:	HMG CLD	
Patient F:	Normal control	

**common sample for all centres in 2025*

5. Results

Numbers of result submissions in the 2025 ERNDIM QLOU Heidelberg scheme

	Survey 1	Survey 2
Receipt of results	77	77
No answer	4	7

6. Web site reporting

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

- **Selection of tests**
  Do not select a test if you will not perform it, otherwise the evaluation program includes it in the report.
- **Results**
  - Give quantitative data as much as possible.
  - Enter the key metabolites with the evaluation **in the tables** even if you do not give quantitative data.
  - If the profile is normal: enter “Normal profile” in “Key metabolites”.
  - **Do not enter results in the “comments” window**, otherwise your results will not be included in the evaluation program.
- **Diagnosis**
  Do not enter the diagnosis in the “comments” window, otherwise it will not be included in the evaluation program.
- **Recommendations (= advice for further investigation).**
  - Scored together with the interpretative score.
  - Advice for treatment will not be scored.
  - Do not give advice for further investigation in “Comments on diagnosis”: it will not be included in the evaluation program.

7. Scoring and evaluation of results

A scoring system was developed in 2012 and approved by the ERNDIM Scientific Advisory Board. Similar to other qualitative (proficiency testing) ERNDIM schemes, the maximum score for a sample is 4 points. 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)

Qualitative results and diagnostic proficiency of the samples were scored using the criteria given below. These criteria have been set by the Scientific Advisor, approved by the ERNDIM Scientific Advisory Board (SAB). A second evaluation of this year's results was carried out by Judit Garcia Villoria, Barcelona, scientific advisor of the QLOU Barcelona scheme. The final decision on the scoring in the scheme was made by the SAB at its autumn meeting (27 November 2025).

General criteria used to score results

Item	Description of scoring criteria	Score
Quantitative results	Correct classification of quantitative results (i.e., normal or increased) according to reference values	1
	Incorrect classification of quantitative results	0
Qualitative results	Correct results according to criteria set for the sample	1
	Incorrect: minimally required results not reported	0
Diagnostic proficiency	Correct according to criteria set for the sample	2
	Partially correct	1
	Unsatisfactory or misleading	0
	Maximum total score	4

ERNDIM applies the concept of ‘critical error’ in the scoring of results. In principle this is a category of error that would be unacceptable to the majority of labs and would have a serious adverse effect on patient management. Labs failing to make a correct diagnosis of a sample considered eligible for this category will be deemed not to have reached a satisfactory performance even if their total points for the year are sufficient according to the requirement set by the SAB. The classification of samples to be judged for critical error was undertaken at the SAB meeting held on 17 November 2025.

Score for satisfactory performance
At least 17 points out of a maximum of 24 (71%) are required for satisfactory performance.

The ERNDIM Annual Certificate covers all ERNDIM schemes in which a laboratory has participated during the scheme year. For the ‘Qualitative Organic Acids in Urine’ scheme, “participation” is defined as requiring full returns for both surveys during the year. Failure to meet this requirement will result in the certificate of participation showing ‘non-submitter’ or ‘partial submitter’ rather than ‘satisfactory’ or ‘unsatisfactory’.

8. Results of samples and evaluation of reporting

8.1. Patient A

Propionyl-CoA carboxylase deficiency

Patient details provided to participants

21-year-old woman admitted with metabolic acidosis and acute encephalopathy.

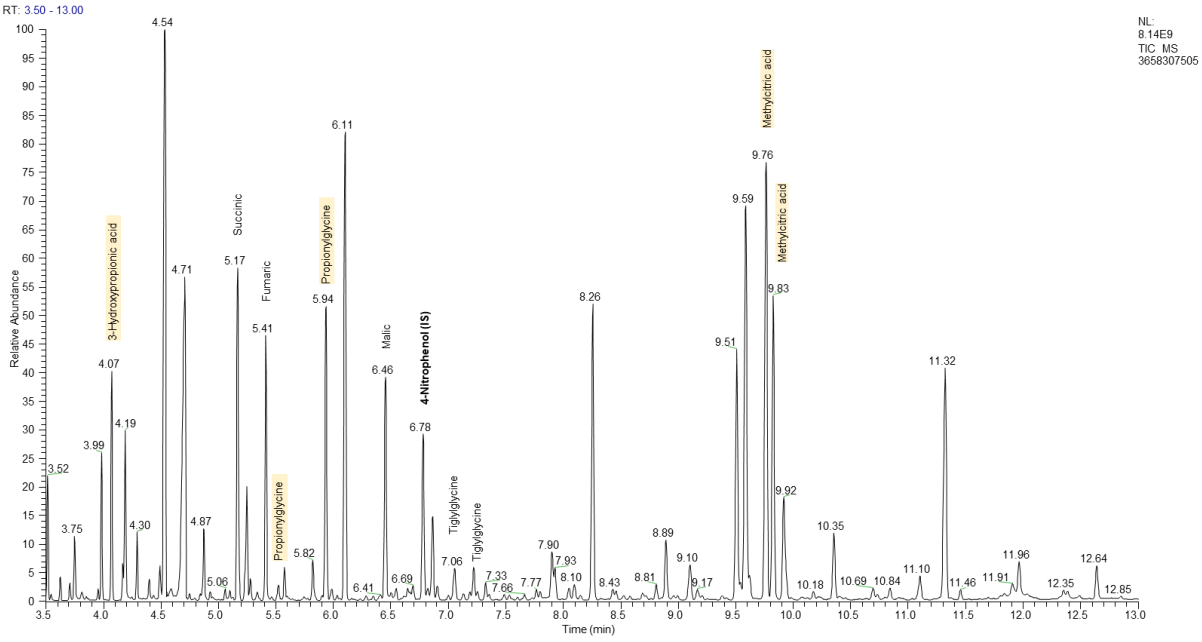
Biochemical diagnosis of propionic aciduria (PA) after metabolic decompensation at 7 days of age. Diagnosis was confirmed subsequently by identification of biallelic variants in *PCCB*. Frequent decompensations with hyperammonemia and metabolic acidosis. Today, she has hypertrophic cardiomyopathy and a chronic kidney disease.

In this organic acid profile, the relevant key metabolites for PA, methylcitric acid (MCA), 3-hydroxypropionic acid (3OHPA) and propionylglycine (ProGly) are markedly elevated along with tiglylglycine and several Krebs cycle metabolites representing secondary mitochondrial impairment.

Analytical performance

The analytes most frequently reported as elevated or grossly elevated were the key metabolites MCA (91% of participants), ProGly (90%) and 3OHPA (81%), followed by tiglylglycine (61%) and Krebs cycle metabolites, i. e., fumaric (45%), succinic (36%), and malic acids (31%). The distribution of these substances reflects very well our results after analysis of the validation sample.
Other metabolites reported less frequently were glutaric (17%) and lactic (16%) acids, isobutyryl (13%) and 3-methylcrotonyl glycine (12%). Four participants mentioned a normal level of methylmalonic acid, one participant rated it elevated.

Evaluation criteria:
One point is given if one of the key metabolites (MCA, 3OHPA or ProGly) is reported at least as elevated. Reporting more than one of them as elevated or grossly elevated yields two points.



Section of an example chromatogram for the validation of sample A analysed in the Heidelberg lab. 4-Nitrophenol is internal standard, key metabolites are highlighted. Note that due to the extent of TMS derivatisation, more than one peak can be observed for an analyte (MCA, ProGly).

Diagnosis / Interpretative proficiency

All participants mentioned PA in their interpretations, 69 as the principal diagnosis and four as alternative. Three labs opted for a type of multiple carboxylase deficiency as the most likely diagnosis, one lab opted for methylmalonic aciduria.

Evaluation criteria: Two points are given for reporting PA as the principal or alternative diagnosis including a recommendation for a test suitable for its confirmation. One point is given for PA as alternative without mentioning an appropriate confirmation method.

Recommendations
In their recommendations for further testing, the participants focused on confirmation by genetic testing (n=62), acylcarnitines in plasma (n=31) or DBS (n=18) and by enzymatic activity testing (n=20). 27 labs recommended to analyse plasma amino acids and 16 responses contained therapeutic advice.

Overall impression
The organic acids profile for this sample was pretty clear, and all participants performed their analyses and diagnostic conclusions very skilfully, resulting in 99% proficiency in analysis and interpretation.

8.2. Patient B

3-Methylcrotonyl-CoA carboxylase (3-MCC) deficiency

This sample was distributed to all participants across the three QLOU centres.

Patient details provided to participants

Man aged 37 years, diagnosed in childhood, now well.

The man was actually diagnosed after diagnosis of 3-MCC deficiency in his newborn nephew, who was identified in neonatal screening in 2004. No other information.

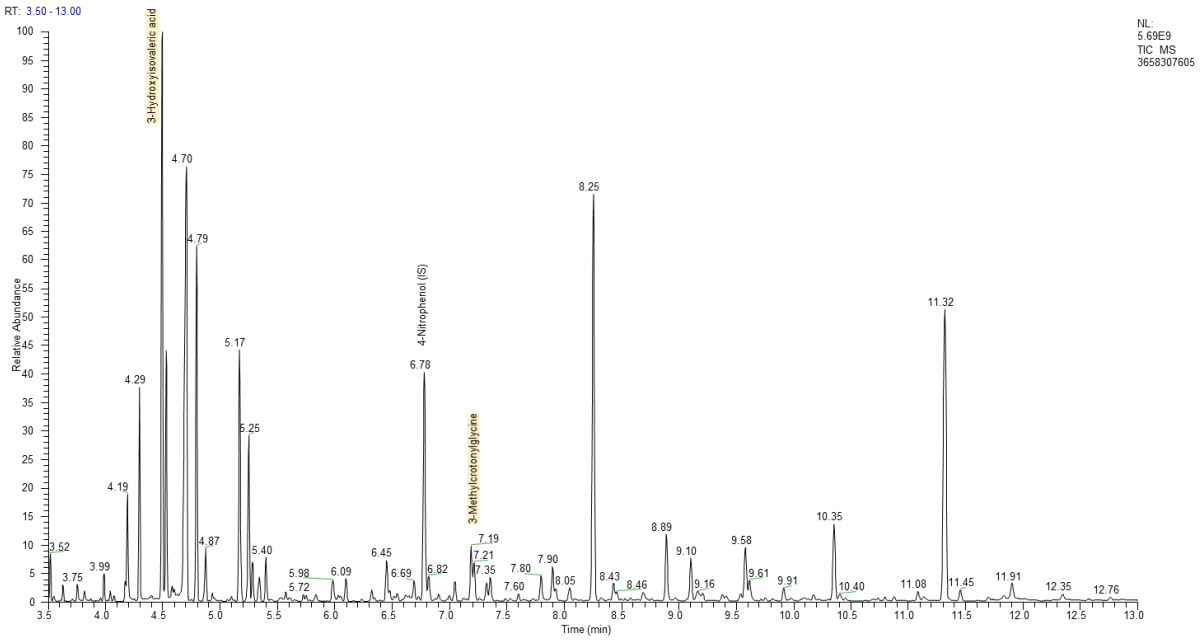
Organic acids analysis by GC-MS reveals massively increased 3-hydroxyisovaleric acid (3-HIVA) and 3-methylgrotonylglycine (3-MCG).

Analytical performance

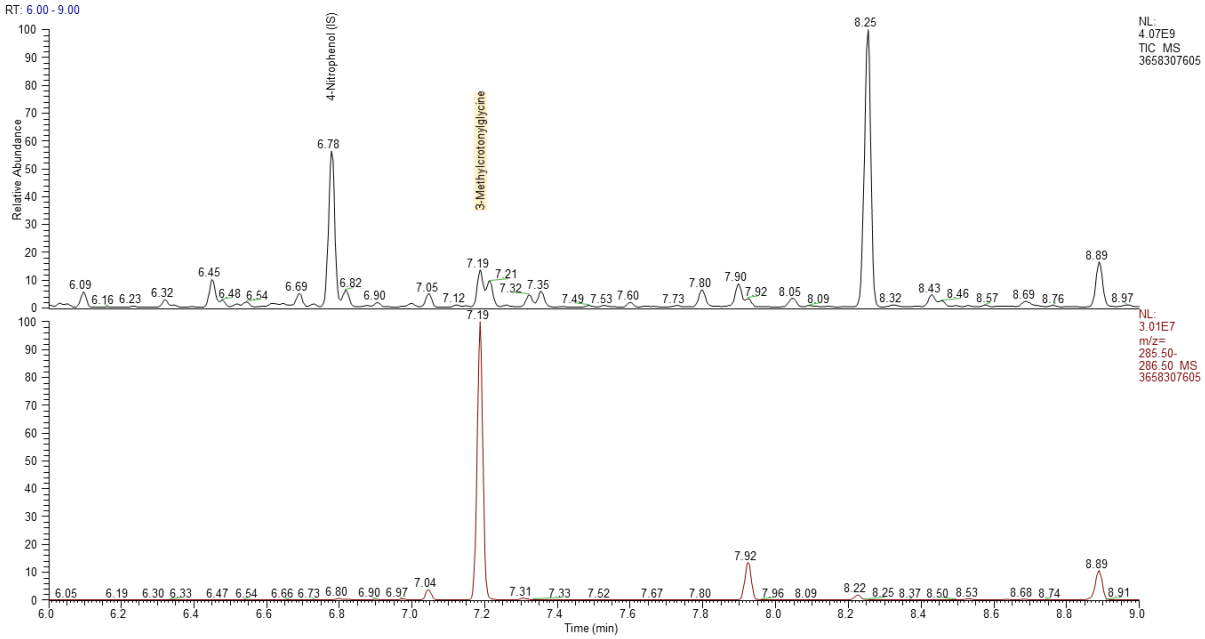
While 95% of the participants submitting results reported 3-HIVA, somewhat less (81%) reported 3-MCG as in elevated or grossly elevated concentrations. The latter was reported as normal by two and as decreased by one laboratory. Other frequently mentioned metabolites were benzoic (32%), succinic (23%) and lactic (11%) acids.

Analytes useful for differential diagnosis, such as isovalerylglycine or 3-methylglutaconic acid, have been highlighted by several laboratories and were mostly classified as normal.

Evaluation criteria: One point each is given for mentioning the two key metabolites, 3-HIVA and/or 3-MCG, in at least elevated concentration.



Chromatogram for the validation of sample B analysed in the Heidelberg lab. 4-Nitrophenol is internal standard, key metabolites are highlighted.



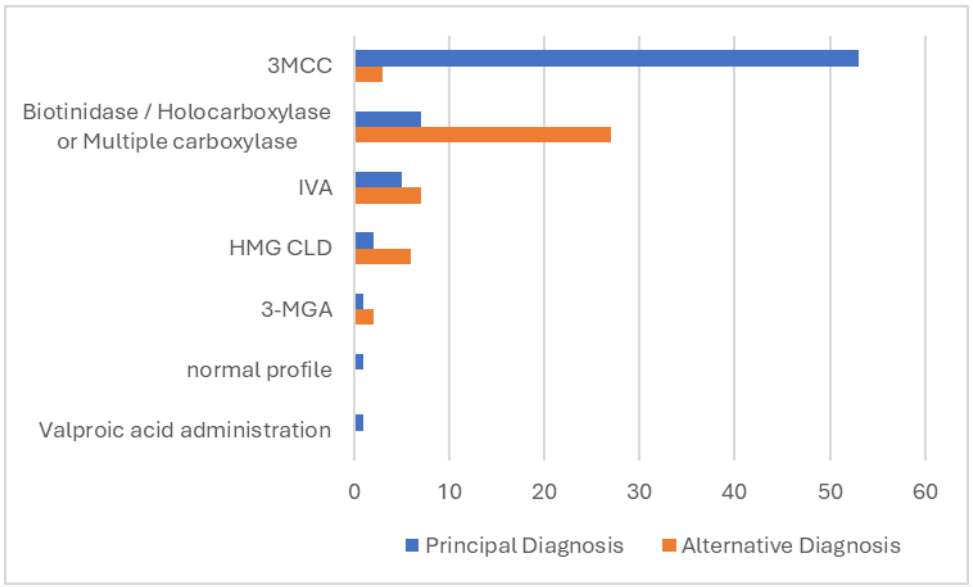
Chromatogram section of the TIC (top) and extracted ion trace (bottom) for 3-methylcrotonylglycine in sample B.

Diagnosis / Interpretative proficiency

53 laboratories (73%) stated 3MCC as their principal diagnosis and three (4%) as alternative. Types of Multiple Carboxylase deficiency were also reported frequently (principal: n=7, alternative: n=27) as interpretation. Five laboratories opted for isovaleric aciduria and two others for HMG CLD as their principal diagnoses. One lab each stated 3-methylglutaconic aciduria, valproate administration and a normal profile.

Evaluation criteria: Two points are awarded for reporting 3-MCC as the principal diagnosis or for mentioning it as alternative, if an appropriate method for confirmation is recommended. Reporting biotinidase, holocarboxylase synthetase or multiple carboxylase deficiency yields one point.

At the SAB meeting it was decided to evaluate this sample without assessment of critical errors.



Principal and alternative diagnoses reported by the participants for sample B. Please note that for alternatives, several entries can be made.

Recommendations

To support or confirm their results, most participants recommended to perform genetic (n=58) or enzymatic (n=28) analyses, measure acylcarnitines in plasma or DBS (n=44), plasma amino acids (n=9), investigate the carnitine status (n=7) or repeat the urinary organic acid analysis (n=7).

Overall impression
For some labs it was not easy to identify the elevated 3-MCG, which was, however, important to find the correct diagnosis. The analytical proficiency resulted in 88%, and the interpretive proficiency 82%.

8.3. Patient C

Mevalonate kinase deficiency

Patient details provided to participants

Boy of 12 years with mild cognitive impairment and ocular problems.

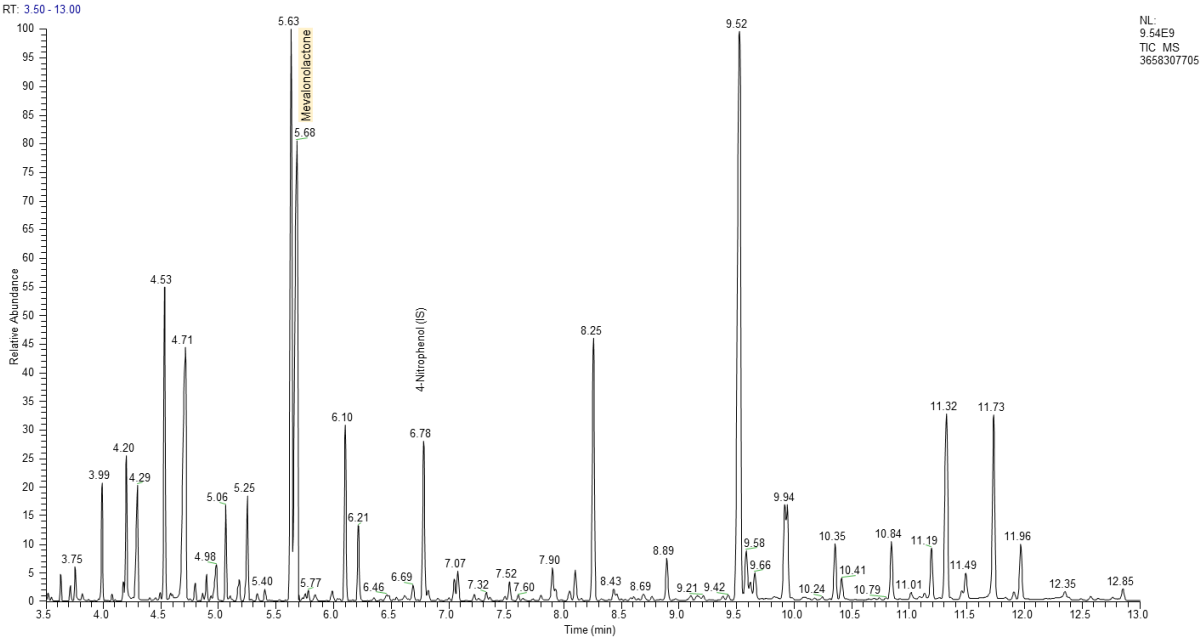
The sample was provided by a patient with developmental delay (predominantly speech delay) and hyperopia, therefore metabolic investigations were initialized. He never had any febrile episodes. In OA, massively elevated mevalonolactone led to the suspicion of mevalonic aciduria, which was confirmed by detection of biallelic pathogenic variants in *MVK*. Ophthalmologic examination showed bilateral papilledema and chorioretinitic pigment changes in the periphery. Later on, he developed cerebellar atrophy and an atactic movement disorder.

The organic acids profile of this sample exhibits a strongly elevated concentration of mevalonolactone.

Analytical performance

The vast majority of participants reported the concentrations of mevalonic acid (69%) and/or mevalonolactone (81%) as markedly increased. Other metabolites were only reported by few participants and included hippuric acid (n=6), 3-hydroxyhippuric acid (n=4) or tiglyl glycine (n=1).

Evaluation criteria: Full score is awarded for reporting mevalonic acid and/or its lactone at least as in elevated concentration.



Chromatogram for the validation of sample C analysed in the Heidelberg lab. 4-Nitrophenol is internal standard, mevalonolactone is highlighted.

Diagnosis / Interpretative proficiency

Mevalonic aciduria was reported as principal diagnosis by 69 labs (95%). Two labs chose it as an alternative. Hyper IgD Syndrome (HIDS) was given as principal diagnosis by four labs and as alternative by 36. Other alternatives mentioned less frequently were phosphomevalonate kinase deficiency (n=2) and mevalonate pyrophosphate decarboxylase deficiency.

One participant stated a normal organic acids profile, which was considered a critical error at the final SAB meeting.

Evaluation criteria: Full score is given if MKD is stated as diagnosis or if HIDS is mentioned as alternative, both if also an appropriate method for confirmation or differentiation is recommended.

Recommendations

Frequent recommendations reported by the participants were genetic tests (n=68), measurement of enzymatic activity (n=34), determination of IgD (n=30), creatine kinase activity (n=13), plasma

cholesterol (n=10), transaminases (n=10), prostanoids and leukotrienes (n=11) and exact quantitation of mevalonic acid (n=6).

Overall impression
The participants performed excellent yielding 99% analytical and interpretive proficiency.

8.4. Patient D

Methylmalonic aciduria mut(0) type

Patient details provided to participants

8-year-old girl on treatment after several episodes of metabolic decompensation.

The sample was taken from an 11-year-old girl who was diagnosed with methylmalonic aciduria after a metabolic decompensation with metabolic stroke at the age of nearly 3 years resulting in dystonic hemiparesis. Molecular analysis showed compound heterozygosity for two pathogenic variants in *MMUT*. At the age of 11 years, she has developed chronic kidney disease (CKD III).

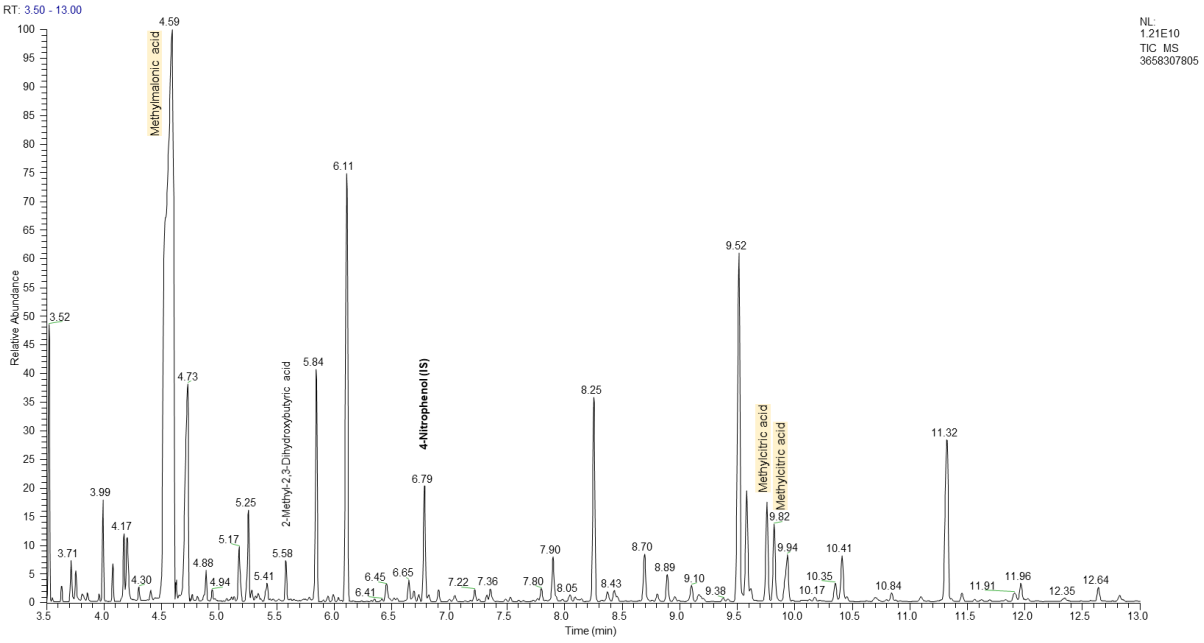
The urinary organic acids analysis reveals a profile typical for MMA with massively elevated methylmalonic acid a clear elevation of 2-methylcitric acid (MCA). Other representative metabolites such as 3-OH-propionic acid or propionylglycine are only slightly elevated. Concentrations of the secondary metabolites lactic and 2,3-dihydroxy-2-methylbutyric acids are also notably elevated.

Analytical performance

Almost all participants (n=69, 99%) described the level of MMA as grossly elevated in their reports. The MCA concentration was stated as grossly elevated by 25 (36%) and as elevated by 39 (56%) participants. One participant described it as normal. 34 participants reported 3OH-PA concentrations (elevated: n=32, normal: n=2) for this sample.

In addition, 43% of participants mentioned elevated or grossly elevated lactate concentrations. Other metabolites were mentioned only occasionally, with propionylglycine being the most frequently named (grossly elevated: n = 3, elevated: n = 8).

Evaluation criteria: One point is awarded for reporting methylmalonic acid at least as elevated. A second point is given for reporting elevated MCA and/or elevated 3OH-PA.



Chromatogram for the validation of sample D analysed in the Heidelberg lab. 4-Nitrophenol is internal standard, Key metabolites are highlighted.

Diagnosis / Interpretative proficiency

Sixty-nine participants reported MMA as their principal diagnosis, mostly specifying certain types of MMA, i. e., mut (0) or cobalamin dependent variants. MMA cbl types were also the most frequently mentioned alternative diagnoses (n= 46). Other diagnoses reported by the participants were combined malonic and methylmalonic aciduria, vitamin B_{12} deficiency and methylmalonyl-CoA epimerase deficiency.

One participant, who found elevated levels of 3-OH- and 4-OH-phenylacetic acids and 5-oxoproline decided for phenylketonuria as principal diagnosis. This was assessed as a critical error and confirmed by the SAB in their final meeting.

Evaluation criteria: Two points are given for reporting MMA as the principal diagnosis or one point is given if it is given as alternative diagnosis. Another point is awarded for recommending a test suitable to find/confirm the correct diagnosis.

Recommendations

In their recommendations for further testing, the participants focused on test to supporting, differentiate and/or confirming their diagnoses, such as acylcarnitines in blood or plasma (n=43), determination of methylmalonic acid in plasma (n=26), investigating the vitamin B_{12} status (n=26) or amino acids in plasma (n=34), often specially referring to homocysteine (n=47). Fifty-nine labs recommended genetic analyses and 14 labs decided for enzymatic tests.

Overall impression

The participants handled this MMA test very competently, resulting in 96% analytical and 97% interpretive proficiency.

8.5. Patient E

3-Hydroxy-3-methylglutaryl-CoA lyase deficiency

Patient details provided to participants

23-year-old man with sinus bradycardia and episodes of hypoglycaemia.

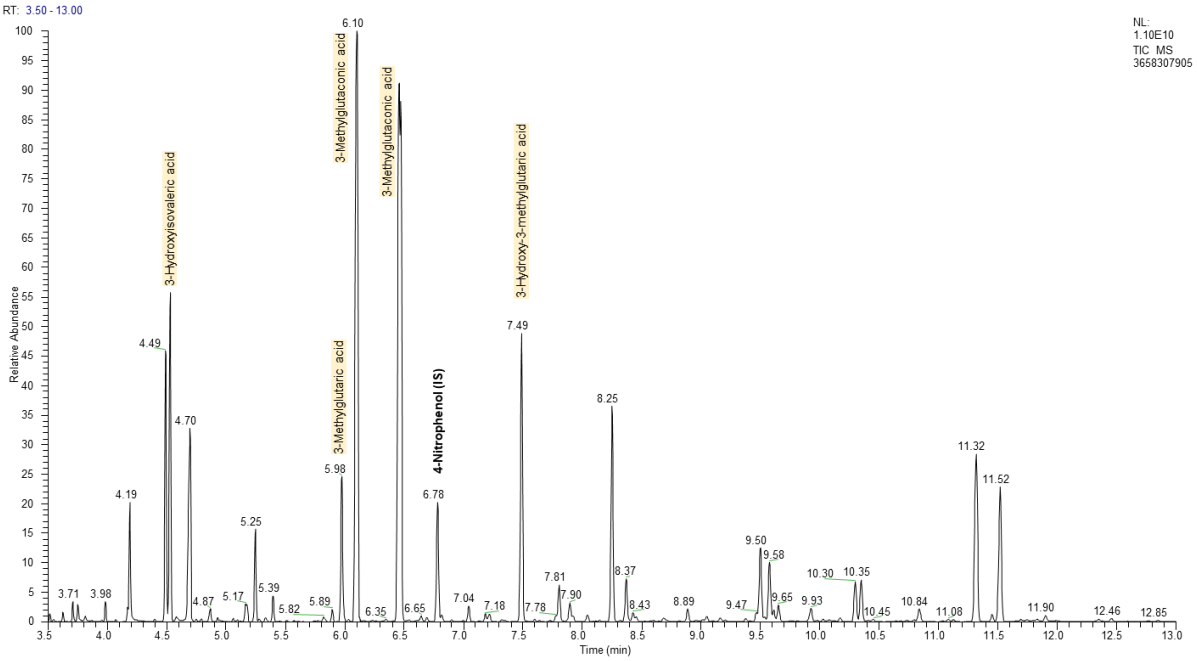
The sample was from a 32-year-old patient with HMG CLD. Metabolic decompensation with hypoketotic hypoglycemia, metabolic acidosis and hepatopathy occurred at age 5 months. The diagnosis was suspected by abnormal organic acids and confirmed by enzyme analysis in fibroblasts. Carnitine treatment and protein and fat reduced diet was started. In the further course he experienced several metabolic decompensations in catabolic situations. He has slight hepatomegaly; his cognitive abilities are in the normal range.

The four signature metabolites indicative for HMG CLD, i. e., 3-hydroxy-3-methylglutaric, 3-methylglutaric, 3-methylglutaconic, 3-hydroxyisovaleric acids are all markedly elevated in this profile of organic acids.

Analytical performance

Of the 70 participants who submitted results for this sample, 96% reported on 3-methylglutaconic acid (grossly elevated: n=57, elevated: n=10), 94% reported on 3-hydroxy-3-methylglutaric acid (grossly elevated: n=43, elevated: n=23), 93% reported on 3-methylglutaric acid (grossly elevated: n=22, elevated: n=43), and 81% reported on 3-hydroxyisovaleric acid (grossly elevated: n=20, elevated: n=37). Thirty-one labs also reported on 3-methylcrotonylglycine (grossly elevated: n=2, elevated: n=29).

Evaluation criteria: If more than two of the key metabolites (3-hydroxy-3-methylglutaric, 3-methylglutaric, 3-methylglutaconic, 3-hydroxyisovaleric acid) are reported at least as elevated two points are awarded. One point is given if one or two of them are reported.



Chromatogram for the validation of sample E analysed in the Heidelberg lab. 4-Nitrophenol is internal standard, Key metabolites are highlighted.

Diagnosis / Interpretative proficiency

HMG CLD was the most frequently mentioned principal diagnosis (n=62, 89%), and also given as alternative diagnosis (n=3). The second most frequently reported diagnosis was 3-methylglutaconic aciduria type 1 (principal: n=6, alternative: n=13), followed by other types of 3MGA.

One participant reported methylmalonic aciduria for this sample, and another participant stated 3-MGA1 without alternative or recommendations. Both results were considered critical errors and confirmed at the SAB meeting.

Evaluation criteria: Two points are given, if HMG CLD is reported as principal diagnosis or one point as alternative diagnosis. A useful recommendation for identifying the correct diagnosis is worth one point.

Recommendations
The most frequently recommended further tests to perform were genetic analyses (n=65), acylcarnitines in plasma or blood (n=48), determination of enzymatic activity (n=22) and plasma amino acids (n=12). Sixteen participants suggested basic tests, such as blood count or blood gas analysis, and 17 labs gave recommendations for patient treatment.

Overall impression
For most participants, it was not challenging to identify this unequivocal profile of organic acids and to draw the right conclusion. The analytical proficiency resulted in 96%, and the interpretive proficiency in 93%.

8.6. Patient F

Normal control

Patient details provided to participants

22-year-old man with muscle weakness.

This normal control sample was from a teenager without any known metabolic disorder.

Analytical performance

Roughly half (n=34) of the laboratories that submitted results reported a normal organic acid profile. The other half of the participants mentioned slightly elevated concentrations of metabolites, with lactate (n=14), glycerate (n=7), oxalic acid (n=6) and hippuric acid (n=9) being the most frequently mentioned, as well as some more general comments on Krebs cycle metabolites (n=4) and/or dicarboxylic acids (n=7).

Evaluation criteria: Full points are awarded for reporting a normal profile / considering absence of a metabolic disorder.

Diagnosis / Interpretative proficiency

In addition to those labs who directly reported a normal organic acids profile, 23 participants interpreted their analytical findings as non-specific elevations of metabolites, i. e., reported them as not indicative for a metabolic disease. Frequently mentioned diagnoses for this sample were mitochondrial disorders, hyperoxaluria, 5-oxoprolinuria, propionic aciduria or glutaric aciduria type I. Two labs that reported elevated vanillactic acid concluded aromatic amino acid decarboxylase deficiency.

Evaluation criteria: Full points are awarded for reporting a normal profile / considering absence of a metabolic disorder.

Recommendations

The most frequently recommended further tests were acylcarnitines in blood or plasma (n=23), plasma or urinary amino acids (n=16), basic laboratory tests (n=10), including creatine kinase analysis (n=7) or a more general recommendation for a broad metabolic workup (n=12).

Overall impression

Fifty-seven participants reported a normal profile or considered their analytical findings to be non-specific, resulting in a combined analytical and interpretive proficiency of 81%.

9. 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.

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.

9.1. Round 1

Lab n°	Patient A Propionic aciduria, PA			Patient B 3-MCC deficiency			Patient C Mevalonic aciduria			
	A	I	Total	A	I	Total	A	I	Total	Total
1	2	2	4	2	2	4	2	2	4	12
2	2	2	4	2	2	4	2	2	4	12
3	2	2	4	2	0	2	2	2	4	10
4	2	2	4	2	2	4	2	2	4	12
5	2	2	4	2	2	4	2	2	4	12
6	2	2	4	2	2	4	2	2	4	12
7	2	2	4	2	2	4	2	2	4	12
8	2	2	4	2	2	4	2	2	4	12
9	2	2	4	2	2	4	2	2	4	12
10	2	2	4	2	2	4	2	2	4	12
11	2	2	4	2	2	4	2	2	4	12
12	2	2	4	2	2	4	2	2	4	12
13	2	2	4	2	2	4	2	2	4	12
14	2	2	4	1	0	1	2	2	4	9
15	2	2	4	2	0	2	2	2	4	10
16	2	2	4	2	2	4	2	2	4	12
17	2	2	4	2	2	4	2	2	4	12
18	2	2	4	2	2	4	2	2	4	12
19	2	2	4	1	1	2	2	2	4	10
20	2	2	4	2	2	4	2	2	4	12
21	2	2	4	2	2	4	2	2	4	12
22	2	2	4	2	2	4	2	2	4	12
23	2	2	4	1	1	2	2	2	4	10
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	2	2	4	1	0	1	2	2	4	9

Lab n°	Patient A Propionic aciduria, PA			Patient B 3-MCC deficiency			Patient C Mevalonic aciduria			
	A	I	Total	A	I	Total	A	I	Total	Total
28	2	2	4	1	2	3	2	2	4	11
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	2	2	4	2	2	4	2	2	4	12
33	2	2	4	2	2	4	2	2	4	12
34	2	2	4	1	1	2	2	2	4	10
35	2	2	4	2	2	4	0	0	0	8
36	2	2	4	2	2	4	2	2	4	12
37	2	2	4	1	2	3	2	2	4	11
38	2	2	4	1	0	1	2	2	4	9
39	2	2	4	2	1	3	2	2	4	11
40	2	2	4	2	2	4	2	2	4	12
41	2	2	4	2	2	4	2	2	4	12
42	2	2	4	2	2	4	2	2	4	12
43	2	2	4	2	2	4	2	2	4	12
44	2	2	4	2	2	4	2	2	4	12
45	2	2	4	2	1	3	2	2	4	11
46	2	2	4	2	2	4	2	2	4	12
47	2	2	4	1	0	1	2	2	4	9
48	2	2	4	2	2	4	2	2	4	12
49	2	2	4	2	1	3	2	2	4	11
50	0	0	0	0	0	0	0	0	0	0
51	2	2	4	0	0	0	2	2	4	8
52	2	2	4	2	2	4	2	2	4	12
53	2	2	4	2	2	4	2	2	4	12
54	2	2	4	1	1	2	2	2	4	10
55	2	2	4	2	2	4	2	2	4	12
56	2	2	4	2	2	4	2	2	4	12
57	2	2	4	2	2	4	2	2	4	12
58	0	0	0	0	0	0	0	0	0	0
59	2	1	3	2	2	4	2	2	4	11
60	2	2	4	2	2	4	2	2	4	12
61	2	2	4	2	2	4	2	2	4	12
62	0	2	2	2	2	4	2	2	4	10

Lab n°	Patient A Propionic aciduria, PA			Patient B 3-MCC deficiency			Patient C Mevalonic aciduria			
	A	I	Total	A	I	Total	A	I	Total	Total
63	1	2	3	1	0	1	2	2	4	8
64	2	2	4	1	1	2	2	2	4	10
65	0	0	0	0	0	0	0	0	0	0
66	0	0	0	0	0	0	0	0	0	0
67	2	2	4	2	2	4	2	2	4	12
68	2	2	4	2	2	4	2	2	4	12
69	2	2	4	1	2	3	2	2	4	11
70	2	2	4	2	2	4	2	2	4	12
71	2	2	4	2	2	4	2	2	4	12
72	2	2	4	2	2	4	2	2	4	12
73	2	2	4	2	2	4	2	2	4	12
74	2	2	4	0	0	0	2	2	4	8
75	2	2	4	2	2	4	2	2	4	12
76	2	2	4	2	2	4	2	2	4	12
77	2	2	4	2	2	4	2	2	4	12

9.2. Round 2

Lab n°	Patient D MMA mut0			Patient E HMG CLD			Patient F Normal control			
	A	I	Total	A	I	Total	A	I	Total	Total
1	2	2	4	2	2	4	2	2	4	12
2	0	0	0	0	0	0	0	0	0	0
3	2	2	4	2	2	4	2	2	4	12
4	2	2	4	2	2	4	2	2	4	12
5	2	2	4	2	2	4	2	2	4	12
6	2	2	4	2	2	4	2	2	4	12
7	2	2	4	2	2	4	2	2	4	12
8	2	2	4	2	2	4	2	2	4	12
9	2	2	4	2	2	4	2	2	4	12
10	2	2	4	2	2	4	2	2	4	12
11	2	2	4	2	2	4	2	2	4	12
12	2	2	4	2	2	4	2	2	4	12
13	2	2	4	2	2	4	0	0	0	8
14	2	2	4	2	2	4	0	0	0	8
15	2	2	4	2	2	4	2	2	4	12
16	2	1	3	2	2	4	0	0	0	7

Lab n°	Patient D MMA mut0			Patient E HMG CLD			Patient F Normal control			
	A	I	Total	A	I	Total	A	I	Total	Total
17	2	2	4	2	2	4	2	2	4	12
18	2	2	4	2	2	4	2	2	4	12
19	2	2	4	2	2	4	2	2	4	12
20	2	2	4	2	2	4	2	2	4	12
21	1	2	3	1	0	1	0	0	0	4
22	2	2	4	2	2	4	2	2	4	12
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	0	0	0	8
27	2	2	4	2	2	4	2	2	4	12
28	2	2	4	2	2	4	2	2	4	12
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	2	2	4	2	2	4	2	2	4	12
33	2	2	4	2	2	4	2	2	4	12
34	0	0	0	0	0	0	0	0	0	0
35	2	2	4	2	2	4	2	2	4	12
36	2	2	4	2	2	4	2	2	4	12
37	2	2	4	2	2	4	2	2	4	12
38	2	2	4	2	2	4	2	2	4	12
39	2	2	4	0	0	0	2	2	4	8
40	2	2	4	2	2	4	2	2	4	12
41	2	2	4	2	2	4	2	2	4	12
42	2	2	4	2	2	4	2	2	4	12
43	0	0	0	0	0	0	0	0	0	0
44	1	2	3	2	2	4	2	2	4	11
45	0	0	0	0	0	0	2	2	4	4
46	2	2	4	2	2	4	2	2	4	12
47	2	2	4	2	2	4	0	0	0	8
48	2	2	4	2	0	2	2	2	4	10
49	2	2	4	2	2	4	0	0	0	8
50	0	0	0	0	0	0	0	0	0	0
51	2	2	4	1	2	3	0	0	0	7

Lab n°	Patient D MMA mut0			Patient E HMG CLD			Patient F Normal control			
	A	I	Total	A	I	Total	A	I	Total	Total
52	2	2	4	2	2	4	2	2	4	12
53	2	2	4	2	2	4	2	2	4	12
54	2	2	4	2	2	4	2	2	4	12
55	2	2	4	2	2	4	2	2	4	12
56	2	2	4	2	2	4	2	2	4	12
57	2	2	4	2	2	4	2	2	4	12
58	0	0	0	0	0	0	0	0	0	0
59	2	2	4	2	2	4	2	2	4	12
60	2	2	4	2	2	4	2	2	4	12
61	2	2	4	2	2	4	2	2	4	12
62	2	2	4	2	2	4	2	2	4	12
63	2	2	4	2	2	4	2	2	4	12
64	2	2	4	2	2	4	0	0	0	8
65	0	0	0	0	0	0	0	0	0	0
66	2	2	4	2	2	4	2	2	4	12
67	2	2	4	2	2	4	2	2	4	12
68	2	2	4	2	2	4	2	2	4	12
69	2	2	4	2	2	4	0	0	0	8
70	2	2	4	2	2	4	2	2	4	12
71	2	2	4	2	2	4	0	0	0	8
72	2	1	3	2	2	4	2	2	4	11
73	2	2	4	2	2	4	2	2	4	12
74	1	2	3	2	2	4	0	0	0	7
75	2	2	4	2	2	4	0	0	0	8
76	2	2	4	2	0	2	2	2	4	10
77	0	0	0	0	0	0	0	0	0	0

9.3. Total scores

Lab n°	A	B	C	D	E	F	Cumulative score	Cumulative score in %	Critical error
1	4	4	4	4	4	4	24	100	
2	4	4	4	0	0	0	12	50	
3	4	2	4	4	4	4	22	92	
4	4	4	4	4	4	4	24	100	
5	4	4	4	4	4	4	24	100	

Lab n°	A	B	C	D	E	F	Cumulative score	Cumulative score in %	Critical error
6	4	4	4	4	4	4	24	100	
7	4	4	4	4	4	4	24	100	
8	4	4	4	4	4	4	24	100	
9	4	4	4	4	4	4	24	100	
10	4	4	4	4	4	4	24	100	
11	4	4	4	4	4	4	24	100	
12	4	4	4	4	4	4	24	100	
13	4	4	4	4	4	0	20	83	
14	4	1	4	4	4	0	17	71	
15	4	2	4	4	4	4	22	92	
16	4	4	4	3	4	0	19	79	
17	4	4	4	4	4	4	24	100	
18	4	4	4	4	4	4	24	100	
19	4	2	4	4	4	4	22	92	
20	4	4	4	4	4	4	24	100	
21	4	4	4	3	1	0	16	67	
22	4	4	4	4	4	4	24	100	
23	4	2	4	4	4	4	22	92	
24	4	4	4	4	4	4	24	100	
25	4	4	4	4	4	4	24	100	
26	4	4	4	4	4	0	20	83	
27	4	1	4	4	4	4	21	88	
28	4	3	4	4	4	4	23	96	
29	4	4	4	4	4	4	24	100	
30	4	4	4	4	4	4	24	100	
31	4	4	4	4	4	4	24	100	
32	4	4	4	4	4	4	24	100	
33	4	4	4	4	4	4	24	100	
34	4	2	4	0	0	0	10	42	
35	4	4	0	4	4	4	20	83	CE
36	4	4	4	4	4	4	24	100	
37	4	3	4	4	4	4	23	96	
38	4	1	4	4	4	4	21	88	
39	4	3	4	4	0	4	19	79	CE
40	4	4	4	4	4	4	24	100	

Lab n°	A	B	C	D	E	F	Cumulative score	Cumulative score in %	Critical error
41	4	4	4	4	4	4	24	100	
42	4	4	4	4	4	4	24	100	
43	4	4	4	0	0	0	12	50	
44	4	4	4	3	4	4	23	96	
45	4	3	4	0	0	4	15	62	CE
46	4	4	4	4	4	4	24	100	
47	4	1	4	4	4	0	17	71	
48	4	4	4	4	2	4	22	92	
49	4	3	4	4	4	0	19	79	
50	0	0	0	0	0	0	0	0	
51	4	0	4	4	3	0	15	62	
52	4	4	4	4	4	4	24	100	
53	4	4	4	4	4	4	24	100	
54	4	2	4	4	4	4	22	92	
55	4	4	4	4	4	4	24	100	
56	4	4	4	4	4	4	24	100	
57	4	4	4	4	4	4	24	100	
58	0	0	0	0	0	0	0	0	
59	3	4	4	4	4	4	23	96	
60	4	4	4	4	4	4	24	100	
61	4	4	4	4	4	4	24	100	
62	2	4	4	4	4	4	22	92	
63	3	1	4	4	4	4	20	83	
64	4	2	4	4	4	0	18	75	
65	0	0	0	0	0	0	0	0	
66	0	0	0	4	4	4	12	50	
67	4	4	4	4	4	4	24	100	
68	4	4	4	4	4	4	24	100	
69	4	3	4	4	4	0	19	79	
70	4	4	4	4	4	4	24	100	
71	4	4	4	4	4	0	20	83	
72	4	4	4	3	4	4	23	96	
73	4	4	4	4	4	4	24	100	
74	4	0	4	3	4	0	15	62	
75	4	4	4	4	4	0	20	83	

Lab n°	A	B	C	D	E	F	Cumulative score	Cumulative score in %	Critical error
76	4	4	4	4	2	4	22	92	
77	4	4	4	0	0	0	12	50	

9.4. Performance

	Number of labs	% total labs
Satisfactory performers (≥ 71 % of adequate responses)	63	82
Unsatisfactory performers (< 71 % adequate responses and/or critical error)	6	8
Partial and non-submitters	8	10

9.5. Overall Proficiency

Sample	Diagnosis	Analytical (%)	Interpretation (%)	Total (%)
QLOU-DH-2025-A	Propionic aciduria, PA	98	99	99
QLOU-DH-2025-B	3-MCC deficiency	88	82	85
QLOU-DH-2025-C	Mevalonic aciduria	99	99	99
QLOU-DH-2025-D	MMA mut0	96	97	97
QLOU-DH-2025-E	HMG CLD	96	93	94
QLOU-DH-2025-F	Normal control	81	81	81

10. Tentative 2026 schedule

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	7th September 2026
Survey 2026/2 - Reports	September/October 2026
Annual Report 2026	January-March 2027

11. 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. For the QLOU scheme this certificate will indicate if results were submitted and whether satisfactory performance was achieved in the scheme.

12. Questions, Suggestions and Complaints

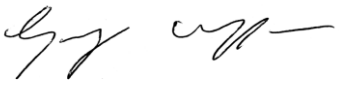
If you have any questions, comments or suggestions please address to the Scientific Advisor of the scheme, Dr. Joachim Janda and/or to the ERNDIM Administration Office (admin@erndim.org).

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/

To be able to continue this scheme, we need a steady supply of new patient samples. Several laboratories have donated urine samples to the QLOU scheme in the past, for which they are gratefully acknowledged. If you are able to collect one or more samples and are willing to donate these to the scheme, please contact us. Laboratories which donate samples that are used in the scheme are eligible for a 20% discount on their participation in the QLOU scheme in the following year.

Date of report, 2026-01-20
Name and signature of Scientific Advisor

Dr. J. Janda
Scientific Advisor
Laboratory of Metabolic Diseases



Prof. Dr. G. F. Hoffmann
Director
Department of General Paediatrics

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

Version Number	Published	Amendments
1	20 January 2026	2025 annual report published

END