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Urine Mucopolysaccharides Centre: The Netherlands Final Report 2021

prepared by
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1. Introduction

The ERNDIM Urine Mucopolysaccharide scheme offers (1) urine samples obtained from confirmed MPS patients to enable laboratories to gain or maintain experience to identify MPS patients and (2) proficiency testing for laboratories providing urine screening of mucopolysaccharidoses. The scheme is organised by UMC Utrecht (Utrecht, The Netherlands) in conjunction with MCA, the Dutch organisation for quality assurance in medical laboratories (MCA laboratory, Winterswijk, NL) and CSCQ, the Swiss organisation for quality assurance in medical laboratories.

2. Geographical distribution of participants

In 2021 87 laboratories from many different countries have registered for the Urine MPS scheme. The number of participants is relatively stable over the years (2018: 100, 2019: 96, 2020: 97 participants), but decreased in 2021. One laboratory was an educational participant. Educational participants take part in all aspects of the scheme and receive interim reports with scores, but performance is not indicated on the ERNDIM certificate of performance.

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	Country	Number of participants
Undefined country	4	Malaysia	2
Argentina	2	Mexico	1
Australia	4	Netherlands	3
Austria	1	New Zealand	2
Belgium	4	Norway	1
Brazil	1	Poland	1
Canada	4	Portugal	2
Colombia	1	Serbia	1
Croatia	1	Singapore	1
Cyprus	1	South Africa	2
Czechia	1	Spain	4
Denmark	1	Sweden	1
Estonia	1	Switzerland	2
France	5	Taiwan	1
Germany	7	Turkey	2
Hong Kong	1	United Kingdom	13
Italia	4	United States of America	4
Latvia	1	Uruguay	1

3. Design and logistics of the scheme including sample information

The scheme has been designed and planned by dr. Berthil Prinsen as Scientific Advisor and coordinated by CSCQ (sub-contractor on behalf of ERNDIM) and Dr. Cas Weykamp and Dr. Eline van der Hagen (MCA, sub-contractors on behalf of ERNDIM) as scheme organisers, all appointed by and according to procedures laid down the ERNDIM Board.

MCA prepares lyophilised sample aliquots and dispatches UMPS EQA samples to the scheme participants by courier. CSCQ provides a website for on-line submission of results and access to scheme reports. Existing Urine MPS 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: samples UMPS-NL-2021-A, B and C
	Round 2: samples UMPS-NL-2021-D, E, and F

As usual, the samples used in 2021 were authentic human urine samples, five from MPS patients and one from a non-MPS individual. Two samples were from the sample repository at UMC Utrecht, Utrecht, the Netherlands and four samples were from the sample repository at Erasmus MC, Rotterdam, The Netherlands. Samples were selected by the Scientific Advisor and tested for suitability in the Scientific Advisor's laboratory (UMC Utrecht, Utrecht, the Netherlands). Integrity of the samples was checked after preparation of the lyophilized aliquots in the Scientific Advisor's laboratory before shipment to participants. Details regarding stability of (reconstituted) samples are provided in the sample package.

UMPS-NL-2021-A	female, 3 years	MPS I
UMPS-NL-2021-B	female, 9 years	MPS VI
UMPS-NL-2021-C	male, 5 years	MPS IVA
UMPS-NL-2021-D	male, 42 years	MPS II
UMPS-NL-2021-E	male, 9 years	Control
UMPS-NL-2021-F	male, 32 years	MPS III

4. Tests

Test required for participation in the Urine MPS scheme are creatinine concentration and GAG analysis (quantitative total GAG and GAG sub fractions, either qualitative by electrophoresis/TLC or quantitative by LC-MS). Participants are asked to interpret the GAG level according to age-matched reference values (i.e normal or increased), interpret GAG subfractions (i.e. normal or increased CS, HS, DS and KS) and to give the most likely diagnosis.

5. Schedule of the scheme

- February, 2021: sample dispatch
- March 1, 2021: analysis start (survey 1)
- March 29, 2021: website available for result submission (survey 1)
- April 26, 2021: deadline for result submission (survey 1)
- May 31, 2021: interim report of survey 1 available for download
- August 1, 2021: analysis start (survey 2)
- August 30, 2021: website available for result submission (survey 2)
- September 27, 2021: deadline for result submission (survey 2)
- October 11, 2021: interim report of survey 2 available for download
- March, 2022: annual report with final scoring, confirmed by the SAB, available for download

6. Results submitted

83 out of the 87 labs that were registered returned results for both surveys. Due to COVID-19, one lab submitted results for survey 1 two weeks later.

	Survey 1	Survey 2
Receipt of results	83	82
No report	4	5

7. Website reporting

Website reporting system is compulsory for all participants. Please note, the website includes a section to specify methods. Method specification is required for correct evaluation of the quantitative results (method specific statistics for DMB, harmine, Alcian Blue, CPC and LC-MS/MS test results). Unfortunately, not all participants have specified their methods.

In 2017 an evaluation program made by dr Albe from CSCQ was used for the first time to evaluate and score results submitted by participants. The use of this software enabled production of customised interim reports and the annual report, i.e. including scores, for each individual participant.

8. Scoring and evaluation of results

Information regarding procedures for establishment of assigned values, statistical analysis, interpretation of statistical analysis can be found in generic documents on the ERNDIM website.

The scoring system has been established by the Scientific Advisory Board of ERNDIM. Scores are allocated to different elements of the results reported. Two aspects are evaluated: 1) analytical performance, 2) interpretative proficiency. The total score is calculated as a sum of these two aspects. Similar to other qualitative (proficiency testing) ERNDIM schemes, the maximum score for a sample is 4 points. The scores were calculated only for laboratories submitting results.

A	Analytical performance	Correct results of the appropriate tests	2
		Partially correct or missing results	1
		Unsatisfactory or misleading	0
I	Interpretative proficiency	Correct (differential) diagnosis was established	2
		Helpful, but (partially) incorrect	1
		Misleading or wrong diagnosis	0

The specific criteria applied to score the results of the samples included in the 2021 scheme are given under item 9. These criteria have been set by the Scientific Advisor, approved by the Scientific Advisory Board, and have been devised on the basis of (1) for each sample: the type of MPS, (2) current possibilities of routine MPS testing, and (3) actual achievable results for a particular sample. The final decision about scoring was made in the Scientific Advisory Board (SAB) during the autumn meeting (November 25-26, 2021 for the 2021 scheme).

A note on scoring of diagnostic proficiency and the use of check boxes and the comment box:
To indicate the most likely diagnosis check boxes must be used to facilitate evaluation of results. The use of the 'comments' box in the website form is recommended to explain your interpretation of results. Comments will be taken into account to score interpretation.

For example, we have noted in previous surveys that it may be hard to distinguish MPS I and VI. In the case of increased DS with normal or undetectable HS, checking just the MPS VI box may result in lower than maximum marks if this actually was a MPS I sample. In this case we advise to check the MPS VI box and explain in the comments box that MPS I (and perhaps II) cannot be excluded on the basis of the results. Or alternatively the boxes for MPS I, II and VI could be checked with a comment entered explaining that MPS VI is more likely.

It is important to realize, when no diagnosis is selected a comment or recommendation is mandatory that needs to explain why the diagnosis 'no diagnosis' is selected. This information is essential for correct scoring of your samples.

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 2021, the SAB decided that samples UMPS-NL-2021-B, UMPS-NL-2021-D and UMPS-NL-2021-F were eligible for critical error (details provided under item 9).

Score required for satisfactory performance: at least 17 points from the maximum of 24 (70%).

From the 87 regular (non-educational) participants 75 participants (86%) submitted results for two rounds and achieved satisfactory performance (2 reports submitted, score ≥ 17 , no critical error).

Twelve participants did not accomplish satisfactory performance, including four due to incomplete submission of results (e.g. no results submitted or 1 survey submitted instead of two reports submitted).

A certificate of participation, including a statement on performance (satisfactory yes/no) will be issued for participation. In addition, performance support letters will be sent out if the performance is evaluated as unsatisfactory. Seven performance support letters were sent by the Scheme Advisor for 2021. Any partial submitters or non-submitters will receive a letter from the ERNDIM office.

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. Results of the samples and evaluation of reporting

9.1. Creatinine and total GAG results of all samples

Quantitative results of creatinine and total GAG were summarised in the two interim reports. Quantitative GAG results were evaluated separately for most methods (DMB, Alcian Blue, Harmine/carbazole, CPC/turbidity). Most participants use DMB (approx. 70 %) for quantitative total GAG analysis. The number of participants using other GAG screening methods is smaller.

Parameter/Method	UMPS-NL-2021-A	UMPS-NL-2021-B	UMPS-NL-2021-C	UMPS-NL-2021-D	UMPS-NL-2021-E	UMPS-NL-2021-F
Creatinine (mmol/L)						
Average	2,35	2,17	1,62	3,56	5,49	5,79
SD	0,19	0,21	0,18	0,17	0,30	0,28
Median	2,37	2,19	1,60	3,57	5,50	5,80
N	80	81	81	76	79	79

Parameter/Method	UMPS-NL-2021-A	UMPS-NL-2021-B	UMPS-NL-2021-C	UMPS-NL-2021-D	UMPS-NL-2021-E	UMPS-NL-2021-F
GAG quantitative (mg/mmol creat) DMB colorimetric method						
Average	58,3	27,7	27,0	24,3	6,2	11,4
SD	24,0	8,6	10,0	7,2	2,3	4,1
Median	59,6	27,1	25,1	23,4	5,9	10,8
N	56	55	56	54	54	55
GAG quantitative (mg/mmol creat) Alcian blue colorimetric tests						
Average	52,2	28,3	23,5	25,7	5,5	10,6
SD	20,8	2,1	1,2	1,6	2,2	1,4
Median	58,0	28,1	23,5	25,3	5,5	10,5
N	4	4	4	4	4	4
GAG quantitative (mg/mmol creat) CPC turbidity method						
Average	75,0	37,0	26,5	41,7	7,9	17,0
SD	39,6	25,5	3,5	15,2	2,6	5,6
Median	75,0	37,0	26,5	41,7	7,9	17,0
N	2	2	2	2	2	2
GAG quantitative (mg/mmol creat) Uronic acids - carbazole/harmine method						
Average	10,8	4,6	4,3	10,3	1,9	4,1
SD	7,3	1,8	1,2	10,0	1,5	3,4
Median	10,8	4,6	4,3	7,0	1,3	3,4
N	2	2	2	3	3	3
GAG quantitative (mg/mmol creat) LC-MS/MS GAG fragments (Saville method)						
Average	81,1	32,7	44,7	--	--	--
SD	0,0	0,0	0,0	--	--	--
Median	81,1	32,7	44,7	--	--	--
N	1	1	1	--	--	--

9.2. Your results

Parameter/Method	UMPS-NL-2021-A	UMPS-NL-2021-B	UMPS-NL-2021-C	UMPS-NL-2021-D	UMPS-NL-2021-E	UMPS-NL-2021-F
Creatinine (mmol/L)						
GAG quantitative (mg/mmol creat) DMB colorimetric method						

The number of non-submitters and registered participants displayed in the tables from section 9.3 – 9.8 are not correct (see section 8 for the correct numbers).

9.3. Sample UMPS-NL-2021-A

Patient details

This sample was from a 3 years old female with MPS-I. Diagnosis was confirmed by enzyme testing and DNA sequencing of the IDUA gene.

Analytical performance

98% of the participants reported an abnormal GAG-screening test (e.g. DMB-test). One participant reported that the GAG-screening test was normal. All participants reported elevated dermatan sulphate (DS) and 49/79 (62%) participants reported an increased heparan sulphate (HS) as well. The analytical performance of this sample was 99%.

Diagnosis / Interpretative proficiency

In total 78 participants (96%) reported a differential diagnosis including MPS I in various combinations with MPS II, VI and VII. Fourteen participants (17%) concluded that this sample was of a patient with MPS I. MPS-II was reported by 1 subject. This is unlikely since MPS-II is an X-linked disorder and this sample was obtained from a female patient (see clinical information). Four participants, that reported both an abnormal GAG-screening test and abnormal GAG-subtype analysis, lost points with the interpretation. The diagnostic performance of this sample was 94% and total performance was 96%.

Diagnosis	N	%
MPS I/MPS II/MPS VI/MPS VII	16	19,8
MPS I	14	17,3
MPS I/MPS II/MPS VII	11	13,6
MPS I/MPS II	9	11,1
MPS I/MPS II/MPS VI	9	11,1
MPS I/MPS VI/MPS VII	7	8,6
MPS I/MPS VI	6	7,4
MPS I/MPS VII	6	7,4
MPS VII	1	1,2
MPS II	1	1,2
MPS III	1	1,2
N results	81	100
N non-submitters	17	
N registered	98	

Scoring

- Analytical results: Elevated total GAG: 1 mark, elevated DS: 1 mark.
- Interpretation: MPS I mentioned in the differential diagnosis (based on elevated DS): 2 marks. Combinations of MPS II, VI or VII based on elevated DS: 1 mark.
- Critical error: The sample was not considered eligible for critical error.

9.4. Sample UMPS-NL-2021-B

Patient details

This sample was from a 9 years old female with MPS VI. Diagnosis was confirmed by enzyme testing.

Analytical performance

97% of the participants reported an elevated GAGs, while two participants reported that the GAG-screening test was normal. Elevated DS was reported by 79/81 participants (98%). The analytical performance for this sample was 97%.

Diagnosis / Interpretative proficiency

The majority of the labs reported MPS VI or MPS VI in different combinations as the most likely diagnosis (71/81, 88%). All combinations with MPS VI included were scored 2 marks. Eight participants did not report the correct diagnosis and lost points with interpretation. Two participants reported normal/no diagnosis (critical error, CE). The diagnostic performance of this sample was 90% and total performance was 94 %.

Diagnosis	N	%
MPS VI	29	35,8
MPS I/MPS II/MPS VI/MPS VII	17	21,0
MPS I/MPS II/MPS VI	8	9,9
MPS I/MPS VI/MPS VII	7	8,6
MPS I/MPS VI	5	6,2

Diagnosis	N	%
MPS VI/MPS VII	4	4,9
MPS I	3	3,7
MPS I/MPS VII	2	2,5
MPS I/MPS II/MPS VII	2	2,5
No Diagnosis	1	1,2
MPS IV/MPS VI	1	1,2
Normal	1	1,2
MPS I/MPS II	1	1,2
N results	81	100
N non-submitters	17	
N registered	98	

Scoring

- Analytical results: Elevated total GAG: 1 mark, elevated DS: 1 mark.
- Interpretation: MPS VI mentioned in the differential diagnosis (based on elevated DS): 2 marks. Combinations of MPS I, II or VII based on elevated DS: 1 mark.
- Critical error: normal profile/no diagnosis was considered as a critical error (CE) (n=2).

9.5. Sample UMPS-NL-2021-C

Patient details

This sample was obtained from a male subject of 5 years old with MPS IVA (severe type). Diagnosis was confirmed by enzyme testing.

Analytical performance

Abnormal GAG-screening was reported by 72 participants (72/78, 92%) and 6 participants (8%) reported a normal GAG-screening result for this sample. Seventy-two participants submitted a result for KS and 82% (59 participants) reported an elevated amount of this GAG-species. N-acetyl-galactosamine 6-sulphatase (GALNS) deficiency may lead to storage of chondroitine-6-sulphate as well and indeed an elevated amount of CS was reported by 22/76 participants (29%). Remarkably 5/76 participants (7%) and 4/76 participants (5%) reported an elevated DS or HS. The analytical performance of this sample was 87%.

Diagnosis / Interpretative proficiency

MPS IV was reported as the most likely diagnosis by 66/80 participants (83%). One participant reported MPS IV in combination with MPS VII/normal diagnosis. Seven participants (87%) concluded that this sample was a normal sample/no diagnosis. Overall proficiency of this sample was 84%, which is slightly higher in comparison with the analysis results of the urine sample of a patient with MPS IV (2020.05). The 2020.05 sample and this sample were from patients with a severe phenotype. The diagnostic proficiency was 81% and the total proficiency was 84%.

Diagnosis	N	%
MPS IV	66	82,5
Normal	6	7,5
MPS VI	2	2,5
MPS VII	2	2,5
No Diagnosis	1	1,3
MPS IV/MPS VII/Normal	1	1,3
MPS I/MPS II/MPS VI	1	1,3
MPS III	1	1,3
N results	80	100

Diagnosis	N	%
N non-submitters	17	
N registered	97	

Scoring

- Analytical results: Elevated total GAG: 1 mark, elevated KS or CS: 1 mark.
- Interpretation: MPS IV mentioned in the differential diagnosis (based on elevated KS or CS): 2 marks.
- Critical error: The sample was not considered eligible for critical error.

9.6. Sample UMPS-NL-2021-D

Patient details

This sample was obtained from an adult patient with MPS II not receiving ERT treatment.

Analytical performance

From the 76 participants that submitted results of total GAG-screening for this sample, 75 participants (99%) reported elevated GAG-concentrations. Most participants also reported abnormal results for GAG-subtyping. DS was reported elevated by 78/79 participants (99%) and HS was found to be elevated by 53/79 participants (67%). These results are similar to the samples circulated in the 2020-scheme (2020.03 and 2020.06). For diagnosis of MPS I/II DS is more aberrant than HS. Analytical performance was 98%. Reporting a normal DS excretion was considered as a critical error (CE) for this sample.

Diagnosis / Interpretative proficiency

The majority of the participants reported a combination of MPS I/II as the most likely diagnosis. In total 77/81 (95%) participants mentioned MPS II among the correct possible diagnoses. Diagnostic performance was 95% and total performance was 97%.

Diagnosis	N	%
MPS I/MPS II	27	33,3
MPS I/MPS II/MPS VI/MPS VII	19	23,5
MPS I/MPS II/MPS VII	15	18,5
MPS I/MPS II/MPS VI	10	12,3
MPS II	5	6,2
MPS VII	1	1,2
MPS I/MPS II/To be entered	1	1,2
MPS I/MPS VI/MPS VII	1	1,2
MPS I	1	1,2
MPS VI	1	1,2
N results	81	100
N non-submitters	6	
N registered	87	

Scoring

- Analytical results: Elevated (total) GAG and elevated DS were each scored 1 mark.
- Interpretation: MPS II with MPS I, VI or VIII in various combinations were scored 2 marks.
- Critical error: Reporting a normal DS excretion was considered as a critical error (CE) for this sample (n=1).

9.7. Sample UMPS-NL-2021-E

Patient details

This urine sample was obtained from a healthy boy of 9 years old.

Analytical performance

Most of the participants (73/76, 96%) reported a normal quantitative GAG screening test. However, 2 participants reported an abnormal quantitative GAG but concluded that this sample was normal after GAG-subtyping. Most participants indeed reported normal GAG-subtyping results by GAG-electrophoresis, TLC or mass-spectrometry. One lab reported elevated HS in combination with elevated CS and one lab reported elevated KS by GAG-subtyping. The analytical performance of this sample was 98%.

Diagnosis / Interpretative proficiency

As is usual for normal samples, most participants (81/82, 99%) correctly conclude that this was not a sample of a patient with a mucopolysaccharidosis. One laboratory concluded that this sample was of a patient with MPS IV. The diagnostic performance was 99% and the overall proficiency of this sample was 98%.

Diagnosis	N	%
Normal	76	92,7
No Diagnosis/Normal	2	2,4
No Diagnosis/To be entered	1	1,2
MPS IV	1	1,2
No Diagnosis	1	1,2
MPS IV/Normal	1	1,2
N results	82	100
N non-submitters	5	
N registered	87	

Scoring

- Analytical results: Normal quantitative GAGs and normal GAG-subtyping were each scored 1 mark.
- Interpretation: A normal profile and other combinations with normal profile/no diagnosis were scored 2 marks.
- Critical error: The sample was not considered eligible for critical error.

9.8. Sample UMPS-NL-2021-F

Patient details

This sample was obtained from an adult patient with MPS-IIIa.

Analytical performance

In this sample 95% of the participants (72/76) reported an increased total quantitative GAGs. Four participant reported a normal quantitative GAGs (5%). The majority of the participants (72/80, 90%) reported elevated HS. The analytical performance of this sample was 91%.

Diagnosis / Interpretative proficiency

In total 72/81 participants reported that this urine specimen was of a patient with MPS III, while 9 participants did not report the correct diagnosis. For this sample reporting a normal profile was considered to be a critical error. The diagnostic performance of this sample was 88% and the overall performance was 90%.

Diagnosis	N	%
MPS III	69	85,2
No Diagnosis	3	3,7
Normal	2	2,5
MPS III/No Diagnosis	1	1,2
MPS IV/MPS VII	1	1,2
MPS I/MPS II	1	1,2

Diagnosis	N	%
MPS I/MPS II/MPS VI	1	1,2
MPS III/Normal	1	1,2
MPS III/To be entered	1	1,2
MPS I/MPS II/MPS VII	1	1,2
N results	81	100
N non-submitters	6	
N registered	87	

Scoring

- Analytical results: Elevated (total) GAG and elevated HS were each scored 1 mark.
- Interpretation: MPS III was scored 2 marks.
- Critical error: Reporting a normal profile as the most likely diagnosis was considered as a critical error (CE) in this sample (n=2).

10. Scores of participants

All data transfer, i.e. the submission of data as well as viewing and downloading of reports proceed via the 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 and only in your version is your laboratory highlighted in the leftmost column.

Detailed scores – Round 1

Lab n°	Sample 1			Sample 2			Sample 3			Total
	MPS-I.			MPS-VI.			MPS-IV(A).			
	A	I	Total	A	I	Total	A	I	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	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	2	2	4	12
14	2	2	4	2	2	4	2	2	4	12
15	2	2	4	2	2	4	2	2	4	12
16	2	2	4	2	2	4	2	2	4	12

Lab n°	Sample 1 MPS-I.			Sample 2 MPS-VI.			Sample 3 MPS-IV(A).			Total
	A	I	Total	A	I	Total	A	I	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	0	2	2	2	4	2	2	4	10
21	--	--	--	--	--	--	--	--	--	0
22	2	1	3	2	2	4	2	2	4	11
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	1	0	1	9
26	2	2	4	2	1	3	2	0	2	9
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	0	0	0	8
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	2	2	4	2	2	4	12
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	1	2	3	2	2	4	11
38	2	2	4	2	2	4	2	2	4	12
39	2	2	4	2	2	4	2	2	4	12
40	2	2	4	2	2	4	2	2	4	12
41	2	2	4	2	2	4	0	0	0	8
42	2	1	3	2	1	3	2	2	4	10
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	2	4	2	2	4	12
46	2	2	4	2	2	4	2	2	4	12
47	2	0	2	2	0	2	1	0	1	5
48	2	2	4	2	2	4	2	2	4	12
49	2	2	4	2	2	4	2	2	4	12

Lab n°	Sample 1 MPS-I.			Sample 2 MPS-VI.			Sample 3 MPS-IV(A).			Total
	A	I	Total	A	I	Total	A	I	Total	
50	2	2	4	2	2	4	1	0	1	9
51	2	2	4	2	1	3	2	2	4	11
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	1	0	1	9
56	2	2	4	2	2	4	1	2	3	11
57	2	2	4	2	2	4	2	2	4	12
58	2	2	4	2	2	4	2	2	4	12
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	1	3	2	2	4	11
62	2	2	4	2	2	4	0	0	0	8
63	2	2	4	1	0	1	1	0	1	6
64	2	2	4	0	0	0	1	2	3	7
65	2	2	4	2	2	4	2	2	4	12
66	2	2	4	2	1	3	2	2	4	11
67	2	2	4	2	2	4	2	2	4	12
68	2	2	4	2	2	4	2	0	2	10
69	2	2	4	2	2	4	1	0	1	9
70	--	--	--	--	--	--	--	--	--	0
71	2	0	2	2	2	4	0	0	0	6
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	2	2	4	2	2	4	12
75	2	2	4	2	1	3	2	2	4	11
76	2	2	4	2	2	4	2	2	4	12
77	1	2	3	2	2	4	2	2	4	11
78	2	2	4	2	2	4	2	2	4	12
79	2	2	4	2	2	4	2	2	4	12
80	2	2	4	2	1	3	2	2	4	11
81	2	2	4	2	2	4	2	0	2	10
82	2	2	4	2	2	4	0	0	0	8

Lab n°	Sample 1 MPS-I.			Sample 2 MPS-VI.			Sample 3 MPS-IV(A).			Total
	A	I	Total	A	I	Total	A	I	Total	
83	2	2	4	2	2	4	1	2	3	11
84	--	--	--	--	--	--	--	--	--	0
85	1	0	1	1	0	1	1	0	1	3
86	2	2	4	2	1	3	0	0	0	7
87	--	--	--	--	--	--	--	--	--	0
88	--	--	--	--	--	--	--	--	--	0

Detailed scores – Round 2

Lab n°	Sample 4			Sample 5			Sample 6			Total
	MPS-II.			Normal profile.			MPS-III(A).			
	A	I	Total	A	I	Total	A	I	Total	
1	2	2	4	2	2	4	1	0	1	9
2	2	2	4	2	2	4	2	2	4	12
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	1	0	1	2	2	4	9
13	2	2	4	2	2	4	2	2	4	12
14	2	2	4	2	2	4	2	2	4	12
15	2	2	4	2	2	4	2	2	4	12
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	2	2	4	2	2	4	12
20	2	2	4	2	2	4	2	2	4	12
21	--	--	--	--	--	--	--	--	--	0
22	2	2	4	1	2	3	2	2	4	11
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	2	2	4	2	2	4	2	2	4	12
28	2	2	4	2	1	4	2	0	2	10
29	2	2	4	2	2	4	2	2	4	12
30	2	2	4	2	2	4	2	2	4	12

Lab n°	Sample 4			Sample 5			Sample 6			Total
	MPS-II.			Normal profile.			MPS-III(A).			
	A	I	Total	A	I	Total	A	I	Total	
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	2	2	4	2	2	4	12
35	2	2	4	2	2	4	2	2	4	12
36	2	2	4	2	2	4	1	2	3	11
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	2	2	4	2	2	4	12
40	2	2	4	2	2	4	2	2	4	12
41	2	2	4	2	2	4	0	0	0	8
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	2	4	2	2	4	12
46	2	2	4	2	2	4	2	2	4	12
47	2	2	4	2	2	4	1	0	1	9
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	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	1	2	3	2	2	4	11
56	2	2	4	2	2	4	2	2	4	12
57	1	0	1	2	2	4	1	2	3	8
58	2	2	4	2	2	4	2	2	4	12
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	1	0	1	9
63	--	--	--	--	--	--	--	--	--	0

Lab n°	Sample 4			Sample 5			Sample 6			Total
	MPS-II.			Normal profile.			MPS-III(A).			
	A	I	Total	A	I	Total	A	I	Total	Total
64	1	2	3	2	2	4	2	2	4	11
65	2	2	4	2	2	4	2	2	4	12
66	2	2	4	2	2	4	1	2	3	11
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	2	2	4	12
70	--	--	--	--	--	--	--	--	--	0
71	2	2	4	2	2	4	2	2	4	12
72	2	1	3	2	2	4	2	2	4	11
73	2	2	4	2	2	4	2	2	4	12
74	2	2	4	2	2	4	2	2	4	12
75	2	2	4	2	2	4	2	2	4	12
76	2	2	4	2	2	4	1	2	3	11
77	2	2	4	2	2	4	2	2	4	12
78	2	2	4	2	2	4	1	0	1	9
79	2	1	3	2	2	4	2	2	4	11
80	2	2	4	2	2	4	2	2	4	12
81	2	2	4	2	2	4	1	0	1	9
82	2	0	2	2	2	4	2	2	4	10
83	2	2	4	2	2	4	2	2	4	12
84	--	--	--	--	--	--	--	--	--	0
85	1	0	1	2	2	4	1	0	1	6
86	2	2	4	1	2	3	0	0	0	7
87	--	--	--	--	--	--	--	--	--	0
88	--	--	--	--	--	--	--	--	--	0

Total scores

Lab n°	1	2	3	4	5	6	Cumulative score	Cumulative score (%)	Critical error
1	4	4	4	4	4	1	21	88	
2	4	4	4	4	4	4	24	100	
3	4	4	4	4	4	4	24	100	
4	4	4	4	4	4	4	24	100	
5	4	4	4	4	4	4	24	100	
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	1	4	21	88	
13	4	4	4	4	4	4	24	100	
14	4	4	4	4	4	4	24	100	
15	4	4	4	4	4	4	24	100	
16	4	4	4	4	4	4	24	100	
17	4	4	4	4	4	4	24	100	
18	4	4	4	4	4	4	24	100	
19	4	4	4	4	4	4	24	100	
20	2	4	4	4	4	4	22	92	
21	--	--	--	--	--	--	0	0	
22	3	4	4	4	3	4	22	92	
23	4	4	4	4	4	4	24	100	
24	4	4	4	4	4	4	24	100	
25	4	4	1	4	4	4	21	88	
26	4	3	2	4	4	4	21	88	
27	4	4	4	4	4	4	24	100	
28	4	4	4	4	4	2	22	92	
29	4	4	4	4	4	4	24	100	
30	4	4	0	4	4	4	20	83	
31	4	4	4	4	4	4	24	100	
32	4	4	4	4	4	4	24	100	

Lab n°	1	2	3	4	5	6	Cumulative score	Cumulative score (%)	Critical error
33	4	4	4	4	4	4	24	100	
34	4	4	4	4	4	4	24	100	
35	4	4	4	4	4	4	24	100	
36	4	4	4	4	4	3	23	96	
37	4	3	4	4	4	4	23	96	
38	4	4	4	4	4	4	24	100	
39	4	4	4	4	4	4	24	100	
40	4	4	4	4	4	4	24	100	
41	4	4	0	4	4	0	16	67	CE
42	3	3	4	4	4	4	22	92	
43	4	4	4	4	4	4	24	100	
44	4	4	4	4	4	4	24	100	
45	4	4	4	4	4	4	24	100	
46	4	4	4	4	4	4	24	100	
47	2	2	1	4	4	1	14	58	
48	4	4	4	4	4	4	24	100	
49	4	4	4	4	4	4	24	100	
50	4	4	1	4	4	2	19	79	
51	4	3	4	4	4	4	23	96	
52	4	4	4	4	4	4	24	100	
53	4	4	4	4	4	4	24	100	
54	4	4	4	4	4	4	24	100	
55	4	4	1	4	3	4	20	83	
56	4	4	3	4	4	4	23	96	
57	4	4	4	1	4	3	20	83	CE
58	4	4	4	4	4	4	24	100	
59	4	4	4	4	4	4	24	100	
60	4	4	4	4	4	4	24	100	
61	4	3	4	4	4	4	23	96	
62	4	4	0	4	4	1	17	71	
63	4	1	1	--	--	--	6	25	CE
64	4	0	3	3	4	4	18	75	CE
65	4	4	4	4	4	4	24	100	
66	4	3	4	4	4	3	22	92	

Lab n°	1	2	3	4	5	6	Cumulative score	Cumulative score (%)	Critical error
67	4	4	4	4	4	4	24	100	
68	4	4	2	4	4	4	22	92	
69	4	4	1	4	4	4	21	88	
70	--	--	--	--	--	--	0	0	
71	2	4	0	4	4	4	18	75	
72	4	4	4	3	4	4	23	96	
73	4	4	4	4	4	4	24	100	
74	4	4	4	4	4	4	24	100	
75	4	3	4	4	4	4	23	96	
76	4	4	4	4	4	3	23	96	
77	3	4	4	4	4	4	23	96	
78	4	4	4	4	4	1	21	88	
79	4	4	4	3	4	4	23	96	
80	4	3	4	4	4	4	23	96	
81	4	4	2	4	4	1	19	79	
82	4	4	0	2	4	4	18	75	
83	4	4	3	4	4	4	23	96	
84	--	--	--	--	--	--	0	0	
85	1	1	1	1	4	1	9	38	
86	4	3	0	4	3	0	14	58	CE
87	--	--	--	--	--	--	0	0	
88	--	--	--	--	--	--	0	0	

Performance

	Number of labs	% total labs
Satisfactory performers (≥ 70 % of adequate responses)	75/82	91,4%
Unsatisfactory performers (< 70 % adequate responses and/or critical error)	7/82	8.6%
Partial and non-submitters	1 and 3	1,1% and 3,4%

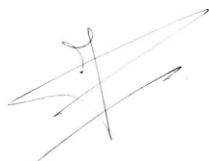
Overall Proficiency

Sample	Diagnosis	Analytical (%)	Interpretation (%)	Total (%)
UMPS-NL-2021-A	MPS-I.	99	94	96
UMPS-NL-2021-B	MPS-VI.	97	90	94
UMPS-NL-2021-C	MPS-IV(A).	87	81	84
UMPS-NL-2021-D	MPS-II.	98	95	97
UMPS-NL-2021-E	Normal profile.	98	99	98
UMPS-NL-2021-F	MPS-III(A).	91	88	90

11. Tentative schedule for 2022

Sample distribution	8 February
Start of analysis of Survey 2022-1. Website open	21 March
Survey 2022-1 - Results submission	19 April
Survey 2022-1 - Reports	16 May
Start of analysis of Survey 2022-2	25 July
Survey 2022-2 – Results submission	12 September
Survey 2022-2 - Reports	October
Annual Report 2022	December/January

Date of report, 2022-03-03



Dr. H.C.M.T. Prinsen

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

Version Number	Published	Amendments
1	26 April 2022	2021 annual report published

END