ERNDIM
Urine mucopolysaccharides
ANNUAL REPORT 2018

Scheme Organiser	Scientific Advisor	Website for reporting results	Administration office:
Dr. C. Weykamp Streekziekenhuis Koningin Beatrix Beatrixpark 1 7101 BN Winterswijk Netherlands e-mail: c.w.weykamp@skbwinterswijk.nl	Dr. G.J.G. Ruijter Erasmus Medical Center Dep. Clinical Genetics Ee2422 P.O. Box 2040 3000 CA Rotterdam e-mail: g.ruijter@erasmusmc.nl	Dr. Xavier Albe CSCQ Swiss Center for Quality Control 2 chemin du Petit-Bel-Air CH-1225 Chêne-Bourg Switzerland e-mail : Xavier.Albe@hcuge.ch	ERNDIM Adminsitration Office Manchester Centre for Genomic Medicine 6th Floor, St Mary's Hospital, Oxford Road, Manchester M13 9WL, United Kingdom. e-mail: admin@erndim.org

January 3, 2019

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 Erasmus Medical Centre (Rotterdam, NL) in conjunction with SKML, 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. Participants

In 2018 100 laboratories from many different countries have initially registered for the Urine MPS scheme (Table 1). The number of participants is relatively stable over the years (2015: 105, 2016: 99, 2017: 102 participants). Two laboratories have withdrawn from scheme participation during the 2018 scheme year, thus the final number of participants is 98. Two laboratories were educational participants in 2018 (6 in 2017). 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.

Table 1. Number of participants in 2018 per country.

Country	No. Participants	Country	No. Participants
ARGENTINA	2	MALAYSIA	2
AUSTRALIA	5	NETHERLANDS	4
AUSTRIA	1	NEW ZEALAND	2
Bahrain	1	NORWAY	1
BELGIUM	4	PAKISTAN	1
BRAZIL	1	POLAND	1
CANADA	5	PORTUGAL	2
COLOMBIA	1	REPUBLIC OF SINGAPORE	1
CROATIA	1	SERBIA	1
CYPRUS	1	SLOVAKIA	1
CZECH REPUBLIC	1	SOUTH AFRICA	2
DENMARK	1	SPAIN	4
ESTONIA	1	SWEDEN	1
FRANCE	7	SWITZERLAND	2
GERMANY	7	TAIWAN	1
GREECE	1	TURKEY	4
HONG KONG	2	UK	15
INDIA	1	URUGUAY	1
ITALY	4	USA	6
LATVIA	1		

3. Design of the scheme and logistics

As usual, the samples used in 2018 were authentic human urine samples, 5 from MPS patients and 1 from a healthy individual (Table 2). Samples were selected by the Scientific Advisor and tested for suitability in the Scientific Advisor's laboratory (Erasmus Medical Centre, Rotterdam, Netherlands). Bulk sample volumes were 350-600 mL. Samples were prepared by lyophilisation of 2.9-5 mL aliquots. Preparation and dispatch of the samples was done by the Scheme organiser (MCA Laboratory, Winterswijk, Netherlands). Integrity of the samples was checked in the Scientific Advisor's laboratory before shipment to participants.

To be able to continue this scheme we need a steady supply of new patient samples. Several laboratories have donated samples to the Urine MPS scheme in the past, for which they are gratefully acknowledged. If you have one or more samples available and are willing to donate these to the scheme, please contact us at g.ruijter@erasmusmc.nl.

Table 2. Samples included in the 2018 ERNDIM Urine MPS scheme. One sample was donated by dr Songaliene, Vilnius, Latvia and another sample by dr Filocamo, Genova, Italy. The other four samples were from the sample repository at Erasmus MC, Rotterdam, The Netherlands.

Survey, reporting deadline	Sample no.	Sample type
2018-1, April 30, 2018	MPS2018.01	MPS I Scheie (f, 38 y)
	MPS2018.02	Normal control (m, 4 y)
	MPS2018.03	MPS II (m, 5 y)
2018-2, October 1, 2018	MPS2018.04	MPS II (m, 42 y)
	MPS2018.05	MPS VII (f, 19 y)
	MPS2018.06	MPS III A (f, 32 y)

The scheme format was kept identical to those of previous years. Samples were shipped by regular mail in February along with other ERNDIM samples. Details regarding stability of (reconstituted) samples are provided in the sample package. Participants were asked to reconstitute each sample in 5 mL deionised water, to determine creatinine concentration (mmol/L) and GAG concentration (mg/mmol creatinine), to qualify the GAG level according to age-matched reference values (i.e normal or increased), to analyse GAG sub fractions and qualify (i.e. normal or increased CS, HS, DS and KS) and to give the most likely diagnosis.

Please see item 4 (scoring of results) for a note on the use of check boxes and the comments box for reporting results

Participants submitted results to the CSCQ website https://cscq.hcuge.ch/cscq/ERNDIM/Initial/Initial.php. The due dates for submitting results in 2018 were April 30th and October 1st.
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, 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, i.e. including scores, for each individual participant.

4. Scoring 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. Scores are allocated to different elements of the results reported (Table 3).
Qualitative results and diagnostic proficiency of the 2018 samples were scored using the criteria given in Table 4 and 5. 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 of the scheme is made in the Scientific Advisory Board (SAB) during the autumn meeting (November 29-30, 2018 for the 2018 scheme). Sample 2018.05, obtained from an MPS VII patient, appeared to be particularly challenging. Based on initial marking, overall proficiency of this sample was 48%. The Scientific Advisory board has decided to class sample 2018.05 as educational. For that reason, sample 2018.05 will not be included in the final scores of the 2018

surveys. As a result, satisfactory performance requires at least 12 points out of the maximum 20 in the 2018 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.

Table 3. 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 (Table 4)	1
	Incorrect: minimally required results not reported	0
Diagnostic proficiency	Correct according to criteria set for the sample (Table 5)	2
	Partially correct	1
	Unsatisfactory or misleading	0
	Maximum total score	4

Table 4. Criteria used for scoring qualitative GAG results (electrophoresis, TLC, LC-MS/MS) of 2018 samples. Sample 2018.05 has been classed as an educational sample and interim scores have been retracted.

Sample	To obtain 1 point the report should state (minimally)
MPS2018.01	Increased DS
MPS2018.02	Normal results for all GAG types, or increased CS only
MPS2018.03	Increased DS
MPS2018.04	Increased DS
MPS2018.05	Educational sample; not scored
MPS2018.06	Increased HS

Starting with the 2014 schemes the concept of 'critical error' is introduced to the assessment of the qualitative schemes. 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 is 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 November 29-30, 2018. Samples MPS2018.01, MPS2018.03 and MPS2018.04 were eligible for critical error. Amongst the reports of regular participants, 2 critical errors were identified in 2018 (both in sample 2018.01). Details are given under item 7 'Results of individual samples and evaluation of reporting'.

Table 5. Criteria for scoring of diagnostic proficiency of 2018 samples. Sample 2018.05 has been classed as an educational sample and interim scores have been retracted.

Sample	Diagnoses (or combinations of possible diagnoses) scored as correct - 2 points	Combinations of possible diagnoses scored as partially correct - 1 point	Not correct - 0 points
MPS2018.01	MPS I (VII) MPS I, II (VII) MPS I, VI (VII) MPS I, II, VI (VII)	MPS VI (VII) MPS II, VI (VII)	Normal Any other (combination of) MPS No diagnosis
MPS2018.02	Normal	-	Any (combination of) MPS No diagnosis
MPS2018.03	MPS II (VII) MPS I, II (VII)	MPS I, II, VI (VII)	Normal Any other (combination of) MPS No diagnosis
MPS2018.04	MPS II (VII) MPS I, II (VII)	MPS I, II, VI (VII)	Normal Any other (combination of) MPS No diagnosis
MPS2018.05	-	-	-
MPS2018.06	MPS III	MPS III, normal	Normal Any other (combination of) MPS No diagnosis

Please see item 4 (scoring of results) for a note on the use of check boxes and the comments box for reporting results.

5. Communication of results

Interim reports with diagnoses, summaries of the results submitted and interim scores were made available June 21, 2018 (survey 2018-1) and November 7, 2018 (survey 2018-2). Sample 2018.05 has been classed as an educational sample and interim scores have been retracted. Scores of the other 5 samples have not been adjusted; scores provided in the interim reports are final scores.
The annual report summarises scheme organisation and results.

ERNDIM provides a single certificate for all its schemes with details of participation and performance.

Seven Performance Support letters will be sent for the 2018 surveys. One of these 7 participants has also received a performance support letter in 2016. Unsatisfactory performance (either due to overall score or due to critical error) within an EQA scheme for at least 2 out of 3 years that the participant has subscribed for will result in a notification letter of unsatisfactory performance to the quality manager or head of department.
For the 2017 scheme also 7 Performance Support letters were sent.

6. Proficiency of the 2018 surveys

In 2018 a total of 96 reports were received for survey 1 (samples MPS2018.01 to MPS2018.03) and 95 reports for survey 2 (samples MPS2018.04 to MPS2018.06). 94 labs submitted results for both surveys, including 2 educational participants. Three participants did not submit any report, while 3 other participants submitted one of the two reports. In 2017 the number of reports was 98-99 per sample. From the 96 ordinary (non-educational) participants 85 (89%) achieved satisfactory performance (2 reports submitted, score ≥12, no critical error). Eleven participants did not accomplish satisfactory performance, including 4 due to incomplete submission of results (i.e. no report or 1 survey report submitted instead of 2 reports).
Overall proficiency in 2018 was 85.7%. Proficiency of each sample is depicted in Table 6.

Table 6. Proficiencies of the 2018 surveys.

Sample ID	Sample type	Proficiency (%)		
		Analytical	Interpretation	Total
MPS2018.01	MPS I Scheie (f, 38 y)	91	78	84
MPS2018.02	Normal control (m, 4 y)	94	92	93
MPS2018.03	MPS II (m, 5 y)	95	78	86
MPS2018.04	MPS II (m, 42 y)	94	75	84
MPS2018.05	MPS VII (f, 19 y)	Educational sample, not scored		
MPS2018.06	MPS III A (f, 32 y)	85	77	81

7. Results of individual samples and evaluation of reporting

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 GAG analysis. The number of participants using other GAG screening methods is smaller.

On average, 8% of the laboratories did not report a diagnosis (range 4-15% for samples 2018.01 to 2018.06). This was partly due to the fact that some laboratories did not perform qualitative analysis of GAG, but also inconclusive test results, e.g. for the MPS III and MPS VII samples, affected the number of diagnoses.

The number of participants reporting the use of LC-MS/MS methods to analyse GAG-derived di- or oligosaccharides is slowly increasing. In 2018, six labs have reported the use of methanolysis to hydrolyse GAGs and 5 have used enzymatic hydrolysis. Amongst these, 2 participants reported the use of both methanolysis and enzymatic hydrolysis. One lab determines sugars and oligosaccharides without hydrolysis. So altogether 10 labs report using LC-MS/MS. While overall proficiency of the 2018 surveys (not including sample 2018.05) was 86%, the 10 participants using LC-MS/MS methods on average scored 96%.

Sample MPS2018.01

Sample type. MPS I, attenuated phenotype, female aged 38 y. This patient was receiving ERT when the urine sample was collected. Since the GAG concentration was still clearly abnormal, treatment did not prohibit use of the sample in the scheme.

Analytical proficiency. In this sample 97% (88/91) of the participants reported increased total GAG and 92% (80/87) increased DS. Only 49% (42/85) of the participants reported increased HS in this sample. This once again illustrates that HS is generally less elevated in MPS I urine samples compared to MPS II samples (e.g. 2018.03 and 2018.04).

Interpretative proficiency. In total 64 participants reported a combination of MPS I, II, VI and VII (including MPS I), which was marked 2. Combinations of MPS II, VI and VII (not including MPS I; n=18) were marked 1. Two participants concluded that this was a normal sample. The relatively low percentage of correct diagnoses reported for sample 2018.01 is because many laboratories (15) diagnosed this sample as MPS VI. As described above, HS was detected by only half of the participants in this sample. In previous years other MPS I samples (e.g. 2016.03 gave similar results and shows the difficulty to distinguish MPS I from MPS VI on the basis of urine mucopolysaccharide analysis with traditional methods for GAG subtype analysis (electrophoresis, TLC).

Overall proficiency (based on points): 84%

Critical error. Reporting ‘normal’ as the most likely diagnosis was considered a critical error in this sample (n=2).

2018.01	GAG screening	CS	DS	HS	KS
	(number)				
Normal	3	62	2	25	21
Increased	88	4	80	42	0
Not detected	-	16	5	18	57
N	91	82	87	85	78

2018.01

Diagnosis	**n (total = 92)**
MPS VI	15 (16 %)
MPS I	13 (14 %)
MPS I/MPS II/MPS VII	12 (13 %)
MPS I/MPS II/MPS VI/MPS VII	11 (12 %)
MPS I/MPS II	9 (10 %)
MPS I/MPS VI	8 (9 %)
MPS I/MPS II/MPS VI	8 (9 %)
No Diagnosis	3 (3 %)
Normal	2 (2 %)
MPS I/MPS VII	2 (2 %)
MPS III	2 (2 %)
MPS I/MPS III	1 (1 %)
MPS I/MPS IV/MPS VI	1 (1 %)
MPS I/MPS VI/MPS VII	1 (1 %)
MPS I/MPS II/MPS III/MPS/VI/MPS VII	1 (1 %)
MPS VI/Normal	1 (1 %)
MPS VI/MPS VII	1 (1 %)
MPS VII	1 (1 %)

Sample MPS2018.02

Sample type. Normal control, 4-year old male.

Analytical proficiency. 95% (86/91) of the participants reported a normal result in the quantitative GAG test. Most participants reported normal test results of GAG electrophoresis/TLC. Three labs reported elevated DS and one increased HS. None of the participants reported increased KS.

Interpretative proficiency. As ususal for normal samples, most participants (91%) correctly concluded that this was not an MPS sample. Three participants concluded a mucopolysaccharidosis in this sample.

Overall proficiency (based on points): 93%

Critical error. This sample was not considered eligible for critical error.

2018.02	GAG screening	CS	DS	HS	KS
	(number)				
Normal	86	83	29	43	23
Increased	5	1	3	1	0
Not detected	-	3	58	46	59
N	91	87	90	90	82

2018.02

Diagnosis	**n (total = 94)**
Normal	84 (89 %)
No Diagnosis	4 (4 %)
MPS I/MPS II/MPS VII	1 (1 %)
MPS I/MPS II/MPS VI/MPS VII	1 (1 %)

MPS IV/Normal	1 (1 %)
MPS IV/MPS VI/Normal	1 (1 %)
MPS VI	1 (1 %)
MPS VI/Normal	1 (1 %)

Sample MPS2018.03

Sample type. MPS II patient, aged 5 y, not receiving ERT treatment.

Analytical proficiency. All but one of the 91 participants that submitted results of GAG screening in this sample reported an elevated concentration (99%). Also, most labs reported abnormal test results of GAG electrophoresis or TLC. 98% (87/89) reported elevated DS, while 93% (83/89) found elevated HS.

Interpretative proficiency. The diagnosis MPS II with or without MPS I or VII was reported as the most likely diagnosis by 64 participants (70%), while another 15 (16%) concluded MPS I, II or VI (or VII). In total, 86% mentioned MPS II among the correct possible differential diagnoses. This sample apparently was a clear MPS case, since none of the participants reported 'normal' as the most likely diagnosis.

Overall proficiency (based on points) 86%.

This sample was also circulated in 2013 (sample ID: MPS23). Overall proficiency was also 86% in 2013.

Critical error. Reporting 'normal' as the most likely diagnosis was considered a critical error in this sample (n=0).

2018.03	**GAG screening**	**CS**	**DS**	**HS**	**KS**
	(number)				
Normal	1	62	0	4	19
Increased	90	16	87	83	2
Not detected	-	4	2	2	57
N	91	82	89	89	78

2018.03 **Diagnosis**	**n (total = 92)**
MPS I/MPS II	33 (36 %)
MPS I/MPS II/MPS VII	27 (29 %)
MPS I/MPS II/MPS VI/MPS VII	10 (11 %)
MPS I/MPS II/MPS VI	5 (5 %)
MPS II	4 (4 %)
MPS VI	4 (4 %)
MPS I	2 (2 %)
MPS III	2 (2 %)
MPS I/MPS VI	1 (1 %)
MPS I/MPS II/MPS III	1 (1 %)
MPS I/MPS II/MPS IV/MPS/VI/MPS VII	1 (1 %)
MPS II/MPS III	1 (1 %)
MPS VII	1 (1 %)

Sample MPS2018.04

Sample type. A sample from an adult MPS II patient (42 y) not receiving ERT treatment.

Analytical proficiency. All 88 participants that submitted results of GAG screening in this sample reported elevated GAG concentration. Also, most labs reported abnormal test results of GAG

electrophoresis or TLC. 99% (84/85) reported elevated DS, while 77% (65/84) found elevated HS. The HS concentration in this sample apparently was lower compared to sample 2018.03 for which 93% of the respondents reported elevated HS.

Interpretative proficiency. MPS II in combination with MPS I or VII was reported as the most likely diagnosis by 67% of the participants, while another 25% concluded MPS I, II or VI (or VII). In total, 92% mentioned MPS II among the possible diagnoses. More participants included MPS VI in the differential diagnosis in sample 2018.04 compared to 2018.03 (25 vs. 20), which may be explained by the less apparent HS storage in 2018.04 compared to sample 2018.03 (obtained from a severely affected MPS II patient).

Overall proficiency (based on points) 84%.

This sample was also circulated in 2014 (sample ID: MPS32). Overall proficiency was 82% in 2014. The proficiencies of the 2 MPS II samples (2018.03 and 2018.04) included in the 2018 surveys were very similar: 86 and 84% respectively.

Critical error. Reporting 'normal' as the most likely diagnosis was considered a critical error in this sample (n=0).

2018.04	**GAG screening**	**CS**	**DS**	**HS**	**KS**
	(number)				
Normal	0	63	1	14	23
Increased	88	5	84	65	0
Not detected	-	10	0	5	50
N	88	78	85	84	73

2018.04

Diagnosis	**n (total = 87)**
MPS I/MPS II	28 (32 %)
MPS I/MPS II/MPS VII	25 (29 %)
MPS I/MPS II/MPS VI	13 (15 %)
MPS I/MPS II/MPS VI/MPS VII	9 (10 %)
MPS II	5 (6 %)
MPS VI	2 (2 %)
MPS I/MPS VI	1 (1 %)
MPS I/MPS II/MPS III/MPS VI	1 (1 %)
MPS I/MPS II/MPS III/MPS VII	1 (1 %)
MPS III	1 (1 %)
MPS VII	1 (1 %)

Sample MPS2018.05

Sample type. 19-year old female MPS VII patient. This sample appeared to be particularly challenging. Based on initial markings, overall proficiency was 48%. Because of the low proficiency, the Scientific Advisory board has decided to class sample 2018.05 as educational. For that reason, sample 2018.05 will not be included in the final scores of the 2018 surveys.

Analytical proficiency. 81% (72/89) of the participants reported an abnormal result in the GAG screening test. The results of GAG subtype analysis were variable with 43% of the participants reporting elevated DS, 35% increased HS, 30% increased CS and 9% increased KS. Since DS, CS and HS all contain glucuronic acid residues; elevation of these 3 GAG species could be expected in an MPS VII urine sample. The results reported of 2018.05 do confirm this, but the elevations were not very clear in this particular sample.

Interpretative proficiency. Many different differential diagnoses were reported (see below). Interestingly, MPS VII as a single possible diagnosis was reported by 11 participants. On the other

hand, 24 participants concluded that this was a normal sample, i.e. not an MPS, and 7 participants reported ‘no diagnosis’.

Overall proficiency (based on points): sample not scored.

Critical error. This sample was not considered eligible for critical error.

2018.05	GAG screening	CS	DS	HS	KS
	(number)				
Normal	17	52	14	34	21
Increased	72	24	36	30	7
Not detected	-	4	34	21	48
N	89	80	84	85	76

2018.05

Diagnosis	**n (total = 88)**
Normal	24 (27 %)
MPS VII	11 (13 %)
MPS IV	7 (8 %)
No diagnosis	7 (7%)
MPS III	6 (7 %)
MPS VI	5 (6 %)
MPS I/MPS II	4 (5 %)
MPS IV/Normal	4 (5 %)
MPS I/MPS VII	3 (3 %)
MPS I/MPS II/MPS VII	3 (3 %)
MPS I/MPS VI	2 (2 %)
MPS I	1 (1 %)
MPS I/MPS II/Normal	1 (1 %)
MPS I/MPS II/MPS VI	1 (1 %)
MPS I/MPS VI/MPS VII	1 (1 %)
MPS I/MPS II/MPS VI/MPS VII	1 (1 %)
MPS I/MPS III/MPS IV/MPS VI	1 (1 %)
MPS I/MPS II/MPS III/MPS VII	1 (1 %)
MPS I/MPS II/MPS III/MPS/VI/MPS VII	1 (1 %)
MPS III/MPS IV/No Diagnosis	1 (1 %)
MPS IV/No Diagnosis	1 (1 %)
MPS VI/Normal	1 (1 %)
MPS VII/No Diagnosis	1 (1 %)

Sample MPS2018.06

Sample type. MPS III A, female aged 32 y

Analytical results. The GAG concentration in this sample was not very high, but clearly abnormal for an adult; 90% of the participants (80/89) reported an abnormal GAG screening result. Many labs (88%) reported elevated HS.

Interpretation. MPS III was reported as the most likely diagnosis by 80% of the participants (71/89). In total 11 participants (13%) concluded normal or ‘no diagnosis’.

Overall proficiency (based on points) 81%.

This sample was also circulated in 2013 (sample ID: MPS22). Overall proficiency was 74% in 2013.

Critical error. This sample was not considered eligible for critical error.

2018.06	GAG screening	CS	DS	HS	KS
	(number)				
Normal	9	58	29	3	24
Increased	80	2	3	75	0
Not detected	-	17	50	7	50
N	89	77	82	85	74

2018.06

Diagnosis	**n (total = 89)**
MPS III	71 (80 %)
No Diagnosis	5 (6 %)
Normal	5 (6 %)
No Diagnosis/Normal	1 (1 %)
MPS I	1 (1 %)
MPS I/MPS II/MPS VII	1 (1 %)
MPS I/MPS II/MPS III/MPS VII	1 (1 %)
MPS I/MPS II/MPS VI/MPS VII	1 (1 %)
MPS I/MPS III/MPS IV/MPS VI	1 (1 %)
MPS III/No Diagnosis	1 (1 %)
MPS III/MPS IV/No Diagnosis	1 (1 %)

8. Preview of the scheme in 2019

The format of the MPS 2019 scheme will be similar to that of previous years.

Website reporting will be maintained in the Urine MPS scheme in 2019. The URL is https://cscq.hcuge.ch/cscq/ERNDIM/Initial/Initial.php, choose 'Urine Mucopolysaccharides'. Currently software is developed to produce annual reports. This will most probably be ready for use in the 2019 surveys. As for the interim reports, annual reports produced by the software will be customized for each participant.

Tentative planning:

Shipment of samples by SKML (all 6 samples in one box):	February 2019
Analysis start survey 1 (website open):	April 1, 2019
Deadline for reporting results of survey 1:	April 29, 2019
Interim report survey 1 available:	June 2019
Analysis start survey 2 (website open):	September 2, 2019
Deadline for reporting results of survey 2:	September 30, 2019
Interim report survey 2 available:	November 2019
Annual report 2019	December 2019

Rotterdam
January 3, 2019

Dr George Ruijter
Scientific Advisor

Note: *This annual report is intended for the participants of the Urine MPS scheme. The contents of this report or data derived from the use or analysis of ERNDIM EQA materials must not be used in written publications or oral presentations unless the explicit prior consent of ERNDIM has been granted*