

DEMOGRAPHICS**1.1a Number of patients registered in year 2019**

Year of follow-up year	Frequency
2019	1021

DEMOGRAPHICS**1.1b Number of patients registered in year 2019 by diagnosis and status**

Table of diagnosis by status					
diagnosis(Diagnosis confirmed) Frequency	status(Status of patient)				
	Deceased	Alive	Not seen during the year of follow-up	Lost to follow-up	Total
No	0	0	0	1	1
Yes	4	971	24	20	1019
Diagnosis to be confirmed	0	1	0	0	1
Total	4	972	24	21	1021

DEMOGRAPHICS**1.1c Number of patients registered, not lost to follow-up, in year 2019**

Year of follow-up

year	Frequency
2019	999

DEMOGRAPHICS

1.1d Number of patients with diagnosis confirmed seen in year 2019

Year of follow-up	year	Frequency
2019		975

DEMOGRAPHICS

1.2 Status of patients

Status of patient			
	status	Frequency	Percent
	Deceased	4	0.40
	Alive	971	97.20
Not seen during the year of follow-up		24	2.40

DEMOGRAPHICS

1.3 Age at follow-up distribution. Patients alive, also not seen, on 31/12/2019

age_fupl	Frequency	Percent
0	14	1.41

1	20	2.01
2	22	2.21
3	22	2.21
4	22	2.21
5	33	3.32
6	20	2.01
7	29	2.91
8	32	3.22
9	34	3.42
10	31	3.12
11	17	1.71
12	25	2.51
13	24	2.41
14	21	2.11
15	23	2.31
16	21	2.11
17	18	1.81
18	23	2.31
19	24	2.41
20	25	2.51
21	23	2.31
22	22	2.21
23	31	3.12
24	26	2.61
25	26	2.61
26	24	2.41
27	26	2.61

28	17	1.71
29	18	1.81
30	16	1.61
31	22	2.21
32	10	1.01
33	13	1.31
34	9	0.90
35	14	1.41
36	12	1.21
37	22	2.21
38	10	1.01
39	14	1.41
40	12	1.21
41	7	0.70
42	14	1.41
43	10	1.01
44	7	0.70
45	6	0.60
46	5	0.50
47	8	0.80
48	4	0.40
49	7	0.70
50	6	0.60
51	10	1.01
52	3	0.30
53	5	0.50
54	7	0.70

55	3	0.30
56	2	0.20
58	2	0.20
59	4	0.40
60	2	0.20
63	1	0.10
64	3	0.30
65	1	0.10
66	1	0.10
67	1	0.10
70	2	0.20
71	2	0.20
72	2	0.20
73	1	0.10
79	1	0.10
85	1	0.10
Frequency Missing = 4		

DEMOGRAPHICS

1.4 Age at follow-up distribution by sex. Patients alive, also not seen, on 31/12/2019

Table of age_fupl by Gender			
age_fupl	Gender(Gender)		
Frequency			
Col Pct	M	F	Total

0	8 1.48	6 1.32	14
1	12 2.22	8 1.76	20
2	13 2.40	9 1.98	22
3	8 1.48	14 3.08	22
4	12 2.22	10 2.20	22
5	17 3.14	16 3.52	33
6	11 2.03	9 1.98	20
7	19 3.51	10 2.20	29
8	16 2.96	16 3.52	32
9	21 3.88	13 2.86	34
10	17 3.14	14 3.08	31
11	8 1.48	9 1.98	17
12	10 1.85	15 3.30	25
13	13 2.40	11 2.42	24
14	11 2.03	10 2.20	21
15	17 3.14	6 1.32	23
16	4 0.74	17 3.74	21
17	12 2.22	6 1.32	18

18	8 1.48	15 3.30	23
19	15 2.77	9 1.98	24
20	14 2.59	11 2.42	25
21	11 2.03	12 2.64	23
22	9 1.66	13 2.86	22
23	16 2.96	15 3.30	31
24	13 2.40	13 2.86	26
25	17 3.14	9 1.98	26
26	14 2.59	10 2.20	24
27	12 2.22	14 3.08	26
28	10 1.85	7 1.54	17
29	6 1.11	12 2.64	18
30	9 1.66	7 1.54	16
31	9 1.66	13 2.86	22
32	5 0.92	5 1.10	10
33	10 1.85	3 0.66	13
34	6 1.11	3 0.66	9
35	8 1.48	6 1.32	14

36	9 1.66	3 0.66	12
37	15 2.77	7 1.54	22
38	5 0.92	5 1.10	10
39	7 1.29	7 1.54	14
40	7 1.29	5 1.10	12
41	3 0.55	4 0.88	7
42	8 1.48	6 1.32	14
43	6 1.11	4 0.88	10
44	3 0.55	4 0.88	7
45	6 1.11	0 0.00	6
46	3 0.55	2 0.44	5
47	5 0.92	3 0.66	8
48	2 0.37	2 0.44	4
49	4 0.74	3 0.66	7
50	6 1.11	0 0.00	6
51	8 1.48	2 0.44	10
52	1 0.18	2 0.44	3
53	3 0.55	2 0.44	5

54	3 0.55	4 0.88	7
55	2 0.37	1 0.22	3
56	2 0.37	0 0.00	2
58	1 0.18	1 0.22	2
59	2 0.37	2 0.44	4
60	2 0.37	0 0.00	2
63	1 0.18	0 0.00	1
64	0 0.00	3 0.66	3
65	1 0.18	0 0.00	1
66	0 0.00	1 0.22	1
67	1 0.18	0 0.00	1
70	1 0.18	1 0.22	2
71	2 0.37	0 0.00	2
72	1 0.18	1 0.22	2
73	0 0.00	1 0.22	1
79	0 0.00	1 0.22	1
85	0 0.00	1 0.22	1
Total	541	454	995

Frequency Missing = 4

DEMOGRAPHICS

1.5 Proportion of children (<18 years) and adults (>=18 years). Patients alive, also not seen, on 31/12/2019

age_fupl	Frequency	Percent
<18	428	43.02
>=18	567	56.98
Frequency Missing = 4		

DEMOGRAPHICS

1.6 Age at follow-up: descriptive statistics. Patients alive, also not seen, on 31/12/2019

Analysis Variable : age_fup						
N	Mean	Minimum	25th Pctl	Median	75th Pctl	Maximum
995	22.8	0.0	10.0	20.9	31.9	85.3

DEMOGRAPHICS

1.7 Sex distribution. Patients alive, also not seen, on 31/12/2019

Gender		
Gender	Frequency	Percent
M	541	54.37
F	454	45.63

DEMOGRAPHICS

1.8 Sex distribution. Patients alive, also not seen, on 31/12/2019 and aged 18 years or more

Gender		
Gender	Frequency	Percent
M	312	55.03
F	255	44.97

DIAGNOSIS

2.1 Age at diagnosis: descriptive statistics. All patients seen in 2019

Analysis Variable : age_dia0							
N							
N	Miss	Mean	Minimum	25th Pctl	Median	75th Pctl	Maximum
864	111	4.00	0.00	0.10	0.40	2.50	81.90

DIAGNOSIS

2.2 Proportion of patients diagnosed at age 1 month or younger and patients diagnosed when older than 1 month. All patients seen in 2019

age_dia0	Frequency	Percent
<=1 month	167	19.33
>1 month	697	80.67
Frequency Missing = 111		

DIAGNOSIS**2.3 Proportion of patients diagnosed when younger than 1 year, when older than 18 years and in between. All patients seen in 2019**

age_dia0	Frequency	Percent
<1	533	61.69
1-18	279	32.29
>18	52	6.02
Frequency Missing = 111		

DIAGNOSIS**2.4 Proportion of patients who underwent neonatal screening. All patients seen in 2019**

Neonatal screening			
neo_scee	Frequency	Percent	
Missing/Unknown	11	1.13	
Not done	744	76.31	
Performed	220	22.56	

DIAGNOSIS**2.5 Proportion of patients who underwent neonatal screening. Patients 5 years old or younger seen in 2019**

Neonatal screening			
neo_scee	Frequency	Percent	

Missing/Unknown	2	1.52
Not done	2	1.52
Performed	128	96.97

DIAGNOSIS

2.6 Patients with meconium ileus by age. All patients seen in 2019

age1 Frequency Row Pct	Table of age1 by ileus					Total
	ileus(Meconium ileus)					
	Missing/Unknown	No	Yes, operated	Yes, not operated	Yes, I don't know if operated	
<=10	16 5.80	222 80.43	30 10.87	6 2.17	2 0.72	276
>10	58 8.30	553 79.11	73 10.44	7 1.00	8 1.14	699
Total	74	775	103	13	10	975

DIAGNOSIS

2.7 Proportion of patients who underwent nasal potential difference measurement. All patients seen in 2019

Nasal Potential Difference (NPD)		
NPD	Frequency	Percent
Missing/Unknown	2	0.21
Not done	973	99.79

DIAGNOSIS

2.9 Proportion of patients who underwent intestinal current measurement. All patients seen in 2019

Intestinal Current Measurement (ICM)		
ICM	Frequency	Percent
Missing/Unknown	2	0.21
Not done	973	99.79

GENETICS

3.1 Proportion of patients with DNA analysis

mut_test	Frequency	Percent
done	971	99.59
not done/missing	4	0.41

GENETICS

3.2 Proportion of alleles which underwent DNA analysis

DNA_test	Frequency	Percent
done	1942	99.59
not done/missing	8	0.41

GENETICS

3.3 Number of patients for whom both mutations were identified and number of patients for whom at least one mutation was unknown

mut_ident	Frequency	Percent
both identified	928	95.57
at least one unknown	43	4.43
Frequency Missing = 4		

GENETICS

3.4 Proportion of identified mutations

mut_cat	Frequency	Percent
identified	1881	96.86
unknown	61	3.14
Frequency Missing = 8		

GENETICS

3.5 Prevalence of F508del homozygous and heterozygous patients. All patients seen in 2019

df	Frequency	Percent
F508del heterozygote	376	38.72
F508del homozygote	436	44.90
Not F508del	159	16.37
Frequency Missing = 4		

GENETICS

3.6 Allelic frequencies of mutations

mut	Frequency	Percent
F508del	1248	64.26
3905insT	64	3.30
unknown	61	3.14
1717-1G->A	60	3.09
G542X	48	2.47
R553X	44	2.27
R347H	27	1.39
R347P	24	1.24
N1303K	20	1.03
W1282X	20	1.03
2347delG	12	0.62
2789+5G->A	12	0.62
Q525X	11	0.57
R1162X	11	0.57
621+1G->T	9	0.46
R117H	9	0.46
3849+10kbC->T	7	0.36
Q39X	7	0.36
2183AA->G	6	0.31
420del9	6	0.31
D1152H	6	0.31
G551D	6	0.31
R1066C	6	0.31

1677delTA	5	0.26
405+1G->A	5	0.26
A455E	5	0.26
CFTRdele14b-17b	5	0.26
E585X	5	0.26
R1158X	5	0.26
1525-1G->A	4	0.21
3659delC	4	0.21
4016insT	4	0.21
5T	4	0.21
L1040P	4	0.21
P5L	4	0.21
S945L	4	0.21
Y1092X	4	0.21
2176insC	3	0.15
5T;TG12	3	0.15
711+5G->A	3	0.15
R1066H	3	0.15
R334W	3	0.15
S1235R	3	0.15
S549R	3	0.15
2184insA	2	0.10
2708del13	2	0.10
3120+1G->A	2	0.10
3271+1G->A	2	0.10
3272-26A->G	2	0.10
3272-4A->G	2	0.10

3359delCT	2	0.10
3732delA	2	0.10
4382delA	2	0.10
5T;TG11	2	0.10
712-1G->T	2	0.10
A561E	2	0.10
C524X	2	0.10
CFTRdele17a,17b	2	0.10
D110H	2	0.10
G1244E	2	0.10
G239R	2	0.10
G85E	2	0.10
I507del	2	0.10
K710X	2	0.10
L206W	2	0.10
L333H	2	0.10
L997F	2	0.10
M1101K	2	0.10
R352Q	2	0.10
R933G	2	0.10
T338I	2	0.10
W1089X	2	0.10
1001+2T9A	1	0.05
1001+4A->C;993delCTTAA	1	0.05
1199delG	1	0.05
17171G2A	1	0.05
1717A	1	0.05

1811+1634A->G	1	0.05
1898+1G->A	1	0.05
2143delT	1	0.05
2183delAA	1	0.05
3095insT	1	0.05
3132delTG	1	0.05
3359delCTCTG	1	0.05
3849+4A->G	1	0.05
4005+1G->A	1	0.05
4022insT	1	0.05
4222delG	1	0.05
5G>A	1	0.05
612T/A	1	0.05
675del4	1	0.05
875+1G->C	1	0.05
A613T	1	0.05
C2175dupA	1	0.05
C276X	1	0.05
C276X;R1438W	1	0.05
CFTRdele2	1	0.05
CFTRdele2,3	1	0.05
CFTRdele22,23	1	0.05
Delta N 1303	1	0.05
E1371X	1	0.05
E528K	1	0.05
E588X	1	0.05
E822X	1	0.05

E831X	1	0.05
E92K	1	0.05
F587S	1	0.05
G-P621	1	0.05
G126D	1	0.05
G178R	1	0.05
G194V	1	0.05
G486E	1	0.05
G576A	1	0.05
I105N	1	0.05
I148N	1	0.05
IVS8 5T-(TG) 12	1	0.05
K464N;p.Arg747Trp	1	0.05
K688X	1	0.05
K68X	1	0.05
L1065R	1	0.05
L1077P	1	0.05
L227R	1	0.05
L467F	1	0.05
L863R	1	0.05
M1V	1	0.05
R1162L	1	0.05
R3147H	1	0.05
R31C	1	0.05
R3342W	1	0.05
R668C	1	0.05
R709X	1	0.05

R75X	1	0.05
R785X	1	0.05
R851L	1	0.05
S1455X	1	0.05
S341P	1	0.05
S466X	1	0.05
S549N	1	0.05
T501I	1	0.05
TG11_T	1	0.05
V201M;D1270N	1	0.05
V456A	1	0.05
V97_P99del (LRG_663t1; c.288-296del]	1	0.05
V97_P99del(c.288_296del)	1	0.05
Y563C	1	0.05
c.1521C>G	1	0.05
c.2225delA	1	0.05
c.3180delA	1	0.05
c.4137-116G>T	1	0.05
c.723_743+1del	1	0.05
c.946delT	1	0.05
Frequency Missing = 8		

LUNG FUNCTION (FEV1_pp percent of predicted computed with GLI equations)

4.0 All patients 6 years old or more: transplants, gender, height and fev1 information to check the numbers of patients

lung_t	liver_t	Gender	hgt	age_fevl	fev1	FEV1_pp	Frequency	Percent
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No	No	M	missing	missing	missing	missing	4	0.48
No	No	M	missing	value	missing	missing	1	0.12
No	No	M	value	value	missing	missing	5	0.60
No	No	M	value	value	value	value	405	48.33
No	No	F	missing	missing	missing	missing	3	0.36
No	No	F	value	value	missing	missing	4	0.48
No	No	F	value	value	value	value	349	41.65
No	Yes	M	value	value	value	value	4	0.48
Yes	No	M	missing	missing	missing	missing	2	0.24
Yes	No	M	value	value	value	value	30	3.58
Yes	No	F	missing	missing	missing	missing	2	0.24
Yes	No	F	value	value	missing	missing	1	0.12
Yes	No	F	value	value	value	value	24	2.86
Yes	Yes	M	value	value	value	value	3	0.36
Yes	Yes	F	value	value	value	value	1	0.12

LUNG FUNCTION (FEV1_pp percent of predicted computed with GLI equations)

4.1 FEV1% of predicted: descriptive statistics. All patients seen in 2019 who have never had a transplant

Analysis Variable : FEV1_pp							
N							
N	Miss	Mean	Minimum	25th Pctl	50th Pctl	75th Pctl	Maximum
754	10	78.0	18.8	58.8	81.8	96.7	138.6

LUNG FUNCTION (FEV1_pp percent of predicted computed with GLI equations)

4.2 FEV1% of predicted: descriptive statistics. All patients seen in 2019 aged 6-17 years who have never had a transplant

Analysis Variable : FEV1_pp							
N							
N	Miss	Mean	Minimum	25th Pctl	Median	75th Pctl	Maximum
287	9	93.7	25.4	84.9	95.3	104.9	138.6

LUNG FUNCTION (FEV1_pp percent of predicted computed with GLI equations)

4.3 FEV1% of predicted: descriptive statistics. All patients seen in 2019 aged 18 years or older who have never had a transplant

Analysis Variable : FEV1_pp							
N							
N	Miss	Mean	Minimum	25th Pctl	Median	75th Pctl	Maximum
467	1	68.4	18.8	49.8	68.3	86.1	122.7

LUNG FUNCTION (FEV1_pp percent of predicted computed with GLI equations)

4.4 FEV1% of predicted: descriptive statistics by age group. All patients seen in 2019 aged 6 years or older have never had a transplant

Analysis Variable : FEV1_pp										
age_fevl	N		N		Mean	Minimum	25th Pctl	Median	75th Pctl	Maximum
	Obs	N	Miss							
6-9	122	117	5		101.4	62.8	93.0	100.8	109.6	138.6
10-14	115	111	4		90.6	33.0	82.6	92.9	101.3	114.5
15-19	108	108	0		81.8	24.8	70.8	84.1	95.3	124.1
20-24	107	107	0		73.4	27.9	53.2	72.5	91.4	122.7
25-29	96	95	1		68.6	22.8	52.7	68.7	84.0	118.6
30-34	60	60	0		62.1	24.5	44.3	57.5	77.8	110.3

35-39	55	55	0	59.2	18.8	43.3	56.1	77.9	116.4
40-44	35	35	0	64.6	21.5	44.6	64.0	82.1	108.4
45+	66	66	0	67.4	26.2	49.6	67.0	83.1	114.9

LUNG FUNCTION (FEV1_pp percent of predicted computed with GLI equations)

4.5 FEV1% of predicted according to severity group and age group. All patients seen in 2019 who have never had a transplant

age_fevl	FEV1_pp			Total
	<40	40-80	>80	
6-17	2 0.70	48 16.72	237 82.58	287
18-29	22 8.76	135 53.78	94 37.45	251
30+	28 12.96	131 60.65	57 26.39	216
Total	52	314	388	754

Frequency Missing = 10

LUNG FUNCTION (FVC_pp percent of predicted computed with GLI equations)

4.6 All patients 6 years old or more: transplants, gender, height and fvc information to check the numbers of patients

lung_t	liver_t	Gender	hgt	fvc	FVC_pp	Frequency	Percent
No	No	M	missing	missing	missing	5	0.60
No	No	M	value	missing	missing	4	0.48
No	No	M	value	value	value	406	48.45

No	No	F	missing	missing	missing	3	0.36
No	No	F	value	missing	missing	5	0.60
No	No	F	value	value	value	348	41.53
No	Yes	M	value	value	value	4	0.48
Yes	No	M	missing	missing	missing	2	0.24
Yes	No	M	value	value	value	30	3.58
Yes	No	F	missing	missing	missing	2	0.24
Yes	No	F	value	value	value	25	2.98
Yes	Yes	M	value	value	value	3	0.36
Yes	Yes	F	value	value	value	1	0.12

LUNG FUNCTION (FVC_pp percent of predicted computed with GLI equations)

4.7 FVC% of predicted: descriptive statistics. All patients seen in 2019 who have never had a transplant

Analysis Variable : FVC_pp							
N							
N	Miss	Mean	Minimum	25th Pctl	50th Pctl	75th Pctl	Maximum
754	10	88.5	25.9	75.4	91.3	102.1	139.7

LUNG FUNCTION (FVC_pp percent of predicted computed with GLI equations)

4.8 FVC% of predicted: descriptive statistics. All patients seen in 2019 aged 6-17 years who have never had a transplant

Analysis Variable : FVC_pp							
N							
N	Miss	Mean	Minimum	25th Pctl	Median	75th Pctl	Maximum
286	10	98.1	55.4	89.6	98.9	107.3	139.7

LUNG FUNCTION (FVC_pp percent of predicted computed with GLI equations)

4.9 FVC% of predicted: descriptive statistics. All patients seen in 2019 aged 18 years or older who have never had a transplant

Analysis Variable : FVC_pp							
N	Miss	Mean	Minimum	25th Pctl	Median	75th Pctl	Maximum
468	0	82.6	25.9	69.2	82.4	97.5	130.5

LUNG FUNCTION (FVC_pp percent of predicted computed with GLI equations)

4.10 FVC% of predicted: descriptive statistics by age group. All patients seen in 2019 aged 6 years or older have never had a transplant

Analysis Variable : FVC_pp									
age_fevl	N Obs	N	N Miss	Mean	Minimum	25th Pctl	Median	75th Pctl	Maximum
6-9	122	116	6	103.0	77.3	94.1	102.5	112.3	139.7
10-14	115	111	4	94.7	55.4	86.6	96.7	104.9	121.2
15-19	108	108	0	93.3	38.7	83.7	95.6	103.9	124.0
20-24	107	107	0	85.9	41.4	72.9	88.0	98.9	128.7
25-29	96	96	0	83.2	25.9	71.4	83.1	100.0	125.6
30-34	60	60	0	78.9	33.4	66.7	76.6	92.5	123.1
35-39	55	55	0	75.7	43.0	61.1	72.0	91.0	130.5
40-44	35	35	0	81.7	33.6	64.3	81.6	97.4	115.1
45+	66	66	0	79.2	39.1	66.6	78.6	92.2	112.1

LUNG FUNCTION (FVC_pp percent of predicted computed with GLI equations)

4.11 FVC% of predicted according to severity group and age group. All patients seen in 2019 who have never had a transplant

Table of age_fevl by FVC_pp				
age_fevl	FVC_pp			
Frequency	<40	40-80	>80	Total
Row Pct				
6-17	0 0.00	30 10.49	256 89.51	286
18-29	3 1.19	93 36.90	156 61.90	252
30+	5 2.31	117 54.17	94 43.52	216
Total	8	240	506	754
Frequency Missing = 10				

LUNG FUNCTION (LCI, lung clearance index)

4.12 LCI: percentage of patients with a value for LCI measurement by age group (age at follow-up). All patients seen in 2019 who have never had a transplant

Table of age1 by lci			
age1	lci(Value of lowest LCI 2.5%%)		
Frequency	missing	value	Total
Row Pct			
0-5	103 78.03	29 21.97	132
6-17	68 23.61	220 76.39	288
18-29	251 95.80	11 4.20	262

30+	223	3	226
	98.67	1.33	
Total	645	263	908

LUNG FUNCTION (LCI, lung clearance index)

4.13 LCI: percentage of patients with a date for LCI measurement. All patients seen in 2019 who have never had a transplant

age_lcil	Frequency	Percent
.	1	0.38
0-5	35	13.31
6-17	216	82.13
18-29	8	3.04
30+	3	1.14

LUNG FUNCTION (LCI, lung clearance index)

4.14 LCI: descriptive statistics by age group (age at LCI measurement). All patients seen in 2019 who have never had a transplant

Analysis Variable : Ici Value of lowest LCI 2.5%%									
age_lcil	N	N	N	Mean	Minimum	25th Pctl	Median	75th Pctl	Maximum
	Obs	N	Miss						
0-5	35	35	0	7.6	6.3	6.9	7.4	8.2	9.6
6-17	216	216	0	9.4	6.1	7.4	8.5	10.5	25.0
18-29	8	8	0	10.0	7.2	7.9	9.3	11.2	16.3
30+	3	3	0	9.5	6.9	6.9	7.0	14.8	14.8

LUNG FUNCTION (LCI, lung clearance index)

4.15 LCI according to severity group and age group (age at LCI measurement). All patients seen in 2019 who have never had a transplant

LUNG FUNCTION (LCI, lung clearance index)

4.16 Distribution of kind of device used for lowest LCI 2.5%. All patients seen in 2019 who have never had a transplant

Device used for lowest LCI 2.5%			
	Ici_dev	Frequency	Percent
	Missing/Unknown	6	2.28
	Ecomedics Exhalyzer D N2-Washout	241	91.63
	Ecomedics Exhalyzer SF6 (Tracergas 4 %)	2	0.76
	Other, not listed	14	5.32

MICROBIOLOGY

5.1a Prevalence of chronic Pseudomonas aeruginosa infection in all patients seen in 2019 who have never had a transplant

Chronic Pseudomonas aeruginosa			
	pseudo	Frequency	Percent
	Missing/Unknown	14	1.54
	No	606	66.74

Yes	288	31.72
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MICROBIOLOGY

5.1b Prevalence of chronic Pseudomonas aeruginosa infection in all patients seen in 2019 who have never had a transplant by age group

Table of age1 by pseudo					
age1		pseudo(Chronic Pseudomonas aeruginosa)			
Frequency					
Row	Pct	Missing/Unknown	No	Yes	Total
<18		7 1.67	365 86.90	48 11.43	420
>=18		7 1.43	241 49.39	240 49.18	488
Total		14	606	288	908

MICROBIOLOGY

5.2a Prevalence of chronic Burkholderia species infection in all patients seen in 2019 who have never had a transplant

Chronic Burkholderia cepacia complex			
burkho	Frequency	Percent	
Missing/Unknown	19	2.09	
No	867	95.48	
Yes	22	2.42	

MICROBIOLOGY

5.2b Prevalence of chronic Burkholderia species infection in all patients seen in 2019 who have never had a transplant by age group

Table of age by burkho					
age	burkho(Chronic Burkholderia cepacia complex)				
Frequency Row Pct	Missing/Unknown	No	Yes	Total	
<18	8 1.90	410 97.62	2 0.48	420	
>=18	11 2.25	457 93.65	20 4.10	488	
Total	19	867	22	908	

MICROBIOLOGY

5.3a Prevalence of chronic Staphylococcus aureus infection in all patients seen in 2019 who have never had a transplant

Chronic Staphylococcus aureus			
staph	Frequency	Percent	
Missing/Unknown	19	2.09	
No	355	39.10	
Yes	534	58.81	

MICROBIOLOGY

5.3b Prevalence of chronic Staphylococcus aureus infection in all patients seen in 2019 who have never had a transplant by age group

Table of age by staph	
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agel	staph(Chronic Staphylococcus aureus)			
	Missing/Unknown	No	Yes	Total
<18	9 2.14	175 41.67	236 56.19	420
>=18	10 2.05	180 36.89	298 61.07	488
Total	19	355	534	908

MICROBIOLOGY

5.4a Prevalence of non tuberculous mycobacteria infection in all patients seen in 2019 who have never had a transplant

Nontuberculous mycobacteria			
myco	Frequency	Percent	
Missing/Unknown	31	3.41	
No	830	91.41	
Yes	47	5.18	

MICROBIOLOGY

5.4b Prevalence of non tuberculous mycobacteria infection in all patients seen in 2019 who have never had a transplant by age group

Table of agel by myco				
agel	myco(Nontuberculous mycobacteria)			
	Missing/Unknown	No	Yes	Total
<18	7 1.67	407 96.90	6 1.43	420

>=18	24	423	41	488
	4.92	86.68	8.40	
Total	31	830	47	908

MICROBIOLOGY

5.5a Prevalence of Stenotrophomonas maltophilia infection in all patients seen in 2019 who have never had a transplant

Stenotrophomonas maltophilia			
malto	Frequency	Percent	
Missing/Unknown	14	1.54	
No	785	86.45	
Yes	109	12.00	

MICROBIOLOGY

5.5b Prevalence of Stenotrophomonas maltophilia infection in all patients seen in 2019 who have never had a transplant by age group

Table of age by malto					
age	malto(Stenotrophomonas maltophilia)				
Frequency	Missing/Unknown	No	Yes	Total	
Row Pct					
<18	5	377	38	420	
	1.19	89.76	9.05		
>=18	9	408	71	488	
	1.84	83.61	14.55		
Total	14	785	109	908	

MICROBIOLOGY

5.6a Prevalence of Haemophilus influenzae infection in all patients seen in 2019 who have never had a transplant

Haemophilus influenzae this year			
HaemInf	Frequency	Percent	
Missing/Unknown	17	1.87	
No	663	73.02	
Yes	228	25.11	

MICROBIOLOGY

5.6b Prevalence of Haemophilus influenzae infection in all patients seen in 2019 who have never had a transplant by age group

Table of age1 by HaemInf					
age1		HaemInf(Haemophilus influenzae this year)			
Frequency					
Row Pct	Missing/Unknown	No	Yes	Total	
<18	6 1.43	271 64.52	143 34.05	420	
>=18	11 2.25	392 80.33	85 17.42	488	
Total	17	663	228	908	

MICROBIOLOGY

5.7a Prevalence of Achromobacter species infection in all patients seen in 2019 who have never had a transplant

Achromobacter species this year

achro	Frequency	Percent
Missing/Unknown	17	1.87
No	849	93.50
Yes	42	4.63

MICROBIOLOGY

5.7b Prevalence of Achromobacter species infection in all patients seen in 2019 who have never had a transplant by age group

Table of agel by achro					
achro(Achromobacter species this year)					
agel	Frequency	Missing/Unknown	No	Yes	Total
Row Pct					
<18	6	1.43	404	10	420
			96.19	2.38	
>=18	11	2.25	445	32	488
			91.19	6.56	
Total	17		849	42	908

MICROBIOLOGY

5.8a Prevalence of Methicillin-resistant Staphylococcus Aureus infection in all patients seen in 2019 who have never had a transplant

Methicillin-resistant Staphylococcus Aureus this year			
MRSA	Frequency	Percent	
Missing/Unknown	37	4.07	
No	856	94.27	

Yes	15	1.65
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MICROBIOLOGY

5.8b Prevalence of Methicillin-resistant Staphylococcus Aureus infection in all patients seen in 2019 who have never had a transplant by age group

Table of age1 by MRSA				
age1	MRSA(Methicillin-resistant Staphylococcus Aureus this year)			
Frequency	Missing/Unknown	No	Yes	Total
Row Pct				
<18	7 1.67	405 96.43	8 1.90	420
>=18	30 6.15	451 92.42	7 1.43	488
Total	37	856	15	908

EXACERBATIONS

5.9a Total days on intravenous antibiotics (for CF-related reasons), at home + in hospital. All patients seen in 2019 who have never had a transplant.

Table of age1 by ivDaysTotal	
age1	ivDaysTotal(Total days on intravenous antibiotics (for CF-related reasons), at home + in hospital, this year)

Frequency Row Pct	Missing	No days	At least one day	Total
0-4	1 1.01	90 90.91	8 8.08	99
5-9	0 0.00	137 93.84	9 6.16	146
10-14	1 0.85	98 83.76	18 15.38	117
15-19	1 0.95	65 61.90	39 37.14	105
20-24	0 0.00	66 57.39	49 42.61	115
25-29	1 1.00	65 65.00	34 34.00	100
30-34	0 0.00	30 50.00	30 50.00	60
35-39	0 0.00	31 53.45	27 46.55	58
40-44	1 2.63	24 63.16	13 34.21	38
45+	0 0.00	46 65.71	24 34.29	70
Total	5	652	251	908

EXACERBATIONS

5.9b Total days on intravenous antibiotics (for CF-related reasons), at home + in hospital: descriptive statistics. All patients seen in 2019 who have never had a transplant.

Analysis Variable : ivDaysTotal Total days on intravenous antibiotics (for CF-related reasons), at home + in hospital, this year

age	N Obs	N Miss	Mean	Minimum	25th Pctl	Median	75th Pctl	Maximum
0-4	99	98	1	0.8	0.0	0.0	0.0	15.0
5-9	146	146	0	2.8	0.0	0.0	0.0	195.0
10-14	117	116	1	2.8	0.0	0.0	0.0	32.0
15-19	105	104	1	7.8	0.0	0.0	14.0	79.0
20-24	115	115	0	12.4	0.0	0.0	14.0	172.0
25-29	100	99	1	9.0	0.0	0.0	15.0	80.0
30-34	60	60	0	11.4	0.0	2.0	15.0	100.0
35-39	58	58	0	8.7	0.0	0.0	14.0	45.0
40-44	38	37	1	14.2	0.0	0.0	14.0	212.0
45+	70	70	0	7.3	0.0	0.0	14.0	87.0

EXACERBATIONS

5.10a Total days on intravenous antibiotics (for CF-related reasons), in hospital only. All patients seen in 2019 who have never had a transplant.

Table of age by ivDaysHosp				
age	ivDaysHosp(Total days on intravenous antibiotics (for CF-related reasons), in hospital only, this year)			
Frequency Row Pct	Missing	No days	At least one day	Total
0-4	1 1.01	90 90.91	8 8.08	99

5-9	0	138	8	146
	0.00	94.52	5.48	
10-14	1	100	16	117
	0.85	85.47	13.68	
15-19	1	69	35	105
	0.95	65.71	33.33	
20-24	0	77	38	115
	0.00	66.96	33.04	
25-29	1	76	23	100
	1.00	76.00	23.00	
30-34	0	38	22	60
	0.00	63.33	36.67	
35-39	1	44	13	58
	1.72	75.86	22.41	
40-44	1	29	8	38
	2.63	76.32	21.05	
45+	1	54	15	70
	1.43	77.14	21.43	
Total	7	715	186	908

EXACERBATIONS

5.10b Total days on intravenous antibiotics (for CF-related reasons), in hospital only: descriptive statistics. All patients seen in 2019 who have never had a transplant.

Analysis Variable : ivDaysHosp Total days on intravenous antibiotics (for CF-related reasons), in hospital only, this year									
age	N		N						
	Obs	N	Miss	Mean	Minimum	25th Pctl	Median	75th Pctl	Maximum
0-4	99	98	1	0.7	0.0	0.0	0.0	0.0	15.0
5-9	146	146	0	1.9	0.0	0.0	0.0	0.0	162.0
10-14	117	116	1	2.1	0.0	0.0	0.0	0.0	32.0
15-19	105	104	1	6.1	0.0	0.0	0.0	9.0	79.0

20-24	115	115	0	5.6	0.0	0.0	0.0	10.0	91.0
25-29	100	99	1	4.1	0.0	0.0	0.0	0.0	44.0
30-34	60	60	0	7.6	0.0	0.0	0.0	11.5	100.0
35-39	58	57	1	2.8	0.0	0.0	0.0	0.0	33.0
40-44	38	37	1	3.1	0.0	0.0	0.0	0.0	28.0
45+	70	69	1	4.8	0.0	0.0	0.0	0.0	87.0

EXACERBATIONS

5.11a Total days in hospital this year, for any reason (routine check-up days not included). All patients seen in 2019 who have never had a transplant.

Table of age1 by HospDaysTotal					
age1		HospDaysTotal(Total days in hospital this year, for any reason, this year. Routine check-up days should not be included.)			
Frequency	Row Pct	Missing	No days	At least one day	Total
0-4		1	82	16	99
		1.01	82.83	16.16	
5-9		0	135	11	146
		0.00	92.47	7.53	
10-14		1	95	21	117
		0.85	81.20	17.95	
15-19		1	65	39	105
		0.95	61.90	37.14	
20-24		0	70	45	115
		0.00	60.87	39.13	

25-29	0	72	28	100
	0.00	72.00	28.00	
30-34	0	37	23	60
	0.00	61.67	38.33	
35-39	1	42	15	58
	1.72	72.41	25.86	
40-44	0	28	10	38
	0.00	73.68	26.32	
45+	1	48	21	70
	1.43	68.57	30.00	
Total	5	674	229	908

EXACERBATIONS

5.11b Total days in hospital this year, for any reason (routine check-up days not included): descriptive statistics. All patients seen in 2019 who have never had a transplant.

Analysis Variable : HospDaysTotal Total days in hospital this year, for any reason, this year. Routine check-up days should not be included.										
age	N	N	N	Mean	Minimum	25th Pctl	Median	75th Pctl	Maximum	
	Obs	Miss	Miss							
0-4	99	98	1	1.6	0.0	0.0	0.0	0.0	21.0	
5-9	146	146	0	2.1	0.0	0.0	0.0	0.0	162.0	
10-14	117	116	1	2.6	0.0	0.0	0.0	0.0	32.0	
15-19	105	104	1	6.9	0.0	0.0	0.0	10.5	86.0	
20-24	115	115	0	6.5	0.0	0.0	0.0	10.0	101.0	
25-29	100	100	0	6.7	0.0	0.0	0.0	5.5	122.0	
30-34	60	60	0	7.9	0.0	0.0	0.0	11.5	100.0	
35-39	58	57	1	3.2	0.0	0.0	0.0	4.0	33.0	
40-44	38	38	0	4.0	0.0	0.0	0.0	2.0	40.0	
45+	70	69	1	6.3	0.0	0.0	0.0	3.0	95.0	

NUTRITION

6.1 Use of pancreatic enzymes for all patients seen in 2019 who have never had a transplant

Use of pancreatic enzymes this year	Frequency	Percent
Missing/Unknown	1	0.11
No	124	13.66
Yes	783	86.23

NUTRITION

6.2 Z-scores for height: descriptive statistics. All patients seen in 2019 aged 17 years or younger who have never had a transplant

Analysis Variable : hgt_z							
	N	Mean	Minimum	25th Pctl	Median	75th Pctl	Maximum
	429	-0.2	-2.9	-0.8	-0.1	0.4	3.3

NUTRITION

6.3 Z-scores for height: descriptive statistics. All patients seen in 2019 aged 18 years or older who have never had a transplant

Analysis Variable : hgt_z							
	N	Mean	Minimum	25th Pctl	Median	75th Pctl	Maximum
	468	-0.2	-3.7	-0.8	-0.2	0.4	2.7

NUTRITION**6.4 Z-scores for height by age group. All patients seen in 2019 who have never had a transplant**

Analysis Variable : hgt_z									
age_fevl	N		N	Mean	Minimum	25th Pctl	Median	75th Pctl	Maximum
	Obs	N	Miss						
0-4	116	113	3	-0.1	-2.3	-0.6	0.0	0.4	3.1
5-9	143	142	1	-0.1	-2.8	-0.8	-0.2	0.5	3.3
10-14	115	115	0	-0.2	-2.6	-0.9	-0.1	0.3	1.9
15-19	108	108	0	-0.4	-2.9	-1.1	-0.4	0.3	2.9
20-24	107	107	0	-0.3	-3.4	-1.1	-0.4	0.3	2.4
25-29	96	96	0	-0.3	-2.6	-1.0	-0.4	0.3	2.4
30-34	60	60	0	-0.2	-3.7	-0.7	-0.3	0.6	2.7
35-39	55	55	0	-0.0	-2.3	-0.7	-0.2	0.7	1.9
40-44	35	35	0	-0.0	-2.4	-0.7	0.0	0.4	2.7
45+	66	66	0	-0.1	-2.2	-0.5	-0.0	0.4	2.0

NUTRITION**6.5 Z-scores for weight: descriptive statistics. All patients seen in 2019 aged 17 years or younger who have never had a transplant**

Analysis Variable : wgt_z								
	N		Mean	Minimum	25th Pctl	Median	75th Pctl	Maximum
	N	Miss						
433	0		-0.3	-4.1	-0.9	-0.2	0.3	4.1

NUTRITION

6.6 Z-scores for weight: descriptive statistics. All patients seen in 2019 aged 18 years or older who have never had a transplant

Analysis Variable : wgt_z							
N	Miss	Mean	Minimum	25th Pctl	Median	75th Pctl	Maximum
467	1	-0.4	-4.9	-1.0	-0.3	0.2	2.1

NUTRITION

6.7 Z-scores for weight by age group. All patients seen in 2019 who have never had a transplant

Analysis Variable : wgt_z									
age_fevl	N Obs	N	Miss	Mean	Minimum	25th Pctl	Median	75th Pctl	Maximum
0-4	116	116	0	-0.2	-2.3	-0.9	-0.1	0.4	4.1
5-9	143	143	0	-0.2	-4.1	-0.7	-0.2	0.4	2.9
10-14	115	115	0	-0.4	-2.7	-1.0	-0.3	0.2	1.7
15-19	108	108	0	-0.6	-4.0	-1.0	-0.4	0.1	2.3
20-24	107	107	0	-0.7	-4.9	-1.3	-0.6	0.1	1.6
25-29	96	96	0	-0.4	-2.7	-1.1	-0.4	0.3	1.5
30-34	60	59	1	-0.3	-4.4	-1.0	-0.1	0.4	1.9
35-39	55	55	0	-0.3	-2.5	-1.0	-0.2	0.3	1.8
40-44	35	35	0	-0.4	-1.9	-1.3	-0.2	0.4	1.7
45+	66	66	0	-0.1	-4.6	-0.6	-0.0	0.7	2.1

NUTRITION

6.8 Z-scores for BMI: descriptive statistics. All patients seen in 2019 aged 2-17 years who have never had a transplant

Analysis Variable : bmi_z

N		Mean	Minimum	25th Pctl	Median	75th Pctl	Maximum
N	Miss						
396	2	-0.2	-3.1	-0.7	-0.2	0.4	3.9

NUTRITION

6.9 BMI: descriptive statistics. All patients seen in 2019 aged 18 years or older who have never had a transplant

Analysis Variable : bmiECFSPR

N		Mean	Minimum	25th Pctl	Median	75th Pctl	Maximum
N	Miss						
467	1	21.7	13.8	19.4	21.3	23.4	36.6

NUTRITION

6.9a BMI: descriptive statistics. All patients seen in 2019 aged 18 years or older (males) who have never had a transplant

Analysis Variable : bmiECFSPR

N		Mean	Minimum	25th Pctl	Median	75th Pctl	Maximum
N	Miss						
254	0	22.1	13.8	20.0	21.9	24.0	31.4

NUTRITION

6.9b BMI: descriptive statistics. All patients seen in 2019 aged 18 years or older (females) who have never had a transplant

Analysis Variable : bmiECFSPR

N							
N	Miss	Mean	Minimum	25th Pctl	Median	75th Pctl	Maximum
213	1	21.2	15.1	19.1	20.5	22.4	36.6

NUTRITION

6.10 Z-scores for BMI by age group. All patients seen in 2019 aged 2-17 years who have never had a transplant

Analysis Variable : bmi_z										
age_fevl	N		N		Mean	Minimum	25th Pctl	Median	75th Pctl	Maximum
	Obs	N	Miss							
2-7	160	159	1		-0.0	-2.7	-0.6	-0.1	0.6	3.9
8-11	109	108	1		-0.4	-3.1	-0.9	-0.4	0.2	1.2
12-17	129	129	0		-0.3	-2.5	-0.7	-0.3	0.3	2.2

NUTRITION

6.11 BMI by age group. All patients seen in 2019 aged 18 years or older who have never had a transplant

Analysis Variable : bmiECFSPR										
age_fevl	N		N		Mean	Minimum	25th Pctl	Median	75th Pctl	Maximum
	Obs	N	Miss							
18-21	86	86	0		20.5	13.8	18.9	20.2	22.0	30.1
22-27	138	138	0		21.6	17.6	19.6	21.7	23.5	27.4
28-31	57	56	1		21.7	17.7	19.4	21.2	23.1	30.8
32-37	65	65	0		21.9	17.0	19.7	22.1	23.7	28.1
38+	122	122	0		22.5	15.1	19.7	21.5	24.4	36.6

NUTRITION

6.12a Proportion of adult patients with BMI<=20 by age and gender. All patients seen in 2019 aged 18 years or older who have never had a transplant

		bmi20				All N
		>20 kilogram/(square metre)		<=20 kilogram/(square metre)		
		N	PctN	N	PctN	
Gender	age_fevl					
M	18-21	24	54.55	20	45.45	44
	22-27	59	76.62	18	23.38	77
	28-31	19	73.08	7	26.92	26
	32-37	35	83.33	7	16.67	42
	38+	52	80.00	13	20.00	65
F	18-21	21	50.00	21	50.00	42
	22-27	36	59.02	25	40.98	61
	28-31	19	63.33	11	36.67	30
	32-37	13	56.52	10	43.48	23
	38+	34	59.65	23	40.35	57

NUTRITION

6.12b Proportion of adult patients with BMI<=18.5 by age and gender. All patients seen in 2019 aged 18 years or older who have never had a transplant

		bmi18_5	All

		>18.5 kilogram/(square metre)		<=18.5 kilogram/(square metre)		
		N	PctN	N	PctN	N
Gender	age_fevl					
M	18-21	37	84.09	7	15.91	44
	22-27	69	89.61	8	10.39	77
	28-31	25	96.15	1	3.85	26
	32-37	40	95.24	2	4.76	42
	38+	61	93.85	4	6.15	65
F	18-21	31	73.81	11	26.19	42
	22-27	51	83.61	10	16.39	61
	28-31	25	83.33	5	16.67	30
	32-37	20	86.96	3	13.04	23
	38+	52	91.23	5	8.77	57

NUTRITION

6.12c Proportion of children with ZBMI<-2 by age and gender. All patients seen in 2019 aged 2-17 years who have never had a transplant

		bmizclass			
		>=-2 zscore		<-2 zscore	
		N	PctN	N	PctN
Gender	age_bmi_class				
M	2-7	86	100.00	.	.
	8-11	56	96.55	2	3.45

F	12-17	63	98.44	1	1.56
	2-7	68	93.15	5	6.85
	8-11	48	96.00	2	4.00
	12-17	62	95.38	3	4.62

COMPLICATIONS AND THERAPY

7.1 Prevalence of allergic broncho-pulmonary aspergillosis in patients seen in 2019 who have never had a transplant

ABPA this year		
ABPA	Frequency	Percent
Missing/Unknown	6	0.66
No ABPA this year	869	95.70
Yes, current ABPA	33	3.63

COMPLICATIONS AND THERAPY

7.2 Prevalence of CF related diabetes in patients seen in 2019 by age group who have never had a transplant

age1	Table of age1 by Diabetes						
	Diabetes(CF related diabetes and type of treatment this year)						
Frequency Row Pct	Missing/Unknown	No diabetes	Yes, treated with daily insulin	Yes, treated with oral hypoglycaemic agents	Yes, dietary advice only	Yes, therapy unknown	Total
<18	5 1.19	395 94.05	9 2.14	0 0.00	10 2.38	1 0.24	420

>=18	0	341	122	4	20	1	488
	0.00	69.88	25.00	0.82	4.10	0.20	
Total	5	736	131	4	30	2	908

COMPLICATIONS AND THERAPY

7.3 Prevalence of pneumothorax in patients seen in 2019 who have never had a transplant

Pneumothorax requiring chest drain this year			
	pneumo	Frequency	Percent
Missing/Unknown		6	0.66
No		900	99.12
Yes, chest drain		2	0.22

COMPLICATIONS AND THERAPY

7.4 Prevalence of hemoptysis in patients seen in 2019 who have never had a transplant

Hemoptysis major over 250 ml this year			
	hemo	Frequency	Percent
Missing/Unknown		5	0.55
No		887	97.69
Yes		16	1.76

COMPLICATIONS AND THERAPY

7.5a Prevalence of malignancy in patients seen in 2019

Occurrence of malignancy this year		
malignancy	Frequency	Percent
Missing/Unknown	5	0.51
No	966	99.08
Colorectal cancer	1	0.10
Other	3	0.31

COMPLICATIONS AND THERAPY

7.5b Prevalence of malignancy in patients seen in 2019 who have never had a transplant

Occurrence of malignancy this year		
malignancy	Frequency	Percent
Missing/Unknown	5	0.55
No	900	99.12
Colorectal cancer	1	0.11
Other	2	0.22

COMPLICATIONS AND THERAPY

7.6 Prevalence of liver disease in patients seen in 2019 who have never had a transplant

Liver disease this year			
	liver	Frequency	Percent
	Missing/Unknown	12	1.32
	No	665	73.24

Cirrhosis with hypertension/hypersplenism	29	3.19
Cirrhosis without hypertension/hypersplenism	9	0.99
Cirrhosis, hypertension unknown	2	0.22
Liver disease without cirrhosis	189	20.81
Variceal bleeding	2	0.22

COMPLICATIONS AND THERAPY

7.7 Prevalence of distal intestinal obstruction syndrome in patients seen in 2019 who have never had a transplant

Distal intestinal obstruction syndrome this year			
	DIOS	Frequency	Percent
Missing/Unknown		5	0.55
No		880	96.92
Yes		23	2.53

COMPLICATIONS AND THERAPY

7.8 Prevalence of salt loss syndrome in patients seen in 2019 who have never had a transplant

Salt loss syndrome this year			
	SaltLoss	Frequency	Percent
Missing/Unknown		8	0.88
No		895	98.57
Yes		5	0.55

COMPLICATIONS AND THERAPY**7.9 Use of ursodeoxycholic acid in patients seen in 2019 who have never had a transplant**

Use of ursodeoxycholic acid this year			
	urso	Frequency	Percent
Missing/Unknown		5	0.55
	No	693	76.32
	Yes	210	23.13

COMPLICATIONS AND THERAPY**7.10 Continuous use of inhaled hypertonic saline in patients seen in 2019 who have never had a transplant**

Inhaled hypertonic NaCl this year			
	NaCl	Frequency	Percent
Missing/Unknown		5	0.55
	No	200	22.03
	Yes	703	77.42

COMPLICATIONS AND THERAPY**7.11 Continuous use of rhDNase in patients seen in 2019 who have never had a transplant**

Use of rhDNase this year			
	rhDNase	Frequency	Percent
Missing/Unknown		3	0.33
	No	466	51.32

Yes	439	48.35
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COMPLICATIONS AND THERAPY

7.12 Continuous use of bronchodilators in patients seen in 2019 who have never had a transplant

Inhaled bronchodilators this year			
	broncho	Frequency	Percent
Missing/Unknown		2	0.22
	No	174	19.16
	Yes	732	80.62

COMPLICATIONS AND THERAPY

7.13 Continuous use of inhaled antibiotics in patients seen in 2019 who have never had a transplant

Inhaled antibiotic this year			
	Antibiotic	Frequency	Percent
Missing/Unknown		1	0.11
	No	567	62.44
	Yes	340	37.44

COMPLICATIONS AND THERAPY

7.14 Use of oxygen therapy in patients seen in 2019 who have never had a transplant

In oxygen therapy this year

Oxygen	Frequency	Percent
Missing/Unknown	5	0.55
No	865	95.26
Yes	38	4.19

COMPLICATIONS AND THERAPY

7.15 Continuous use of macrolides in patients seen in 2019 who have never had a transplant

Use of macrolide this year		
azitro	Frequency	Percent
Missing/Unknown	3	0.33
No	672	74.01
Yes	233	25.66

COMPLICATIONS AND THERAPY

7.16 Continuous use of inhaled mannitol in patients seen in 2019 who have never had a transplant

Continuous inhaled mannitol this year		
Mannitol	Frequency	Percent
Missing/Unknown	6	0.66
No	901	99.23
Yes	1	0.11

COMPLICATIONS AND THERAPY

7.17 Continuous use of NIPPV in patients seen in 2019 who have never had a transplant

Continuous NIPPV this year			
	NIPPV	Frequency	Percent
	Missing/Unknown	3	0.33
	No	899	99.01
Yes - CPAP (Continuous Positive Airways Pressure)		3	0.33
Yes - BiPAP (Bilevel Positive Airways Pressure)		3	0.33

COMPLICATIONS AND THERAPY

7.18 Continuous use of inhaled steroids in patients seen in 2019 who have never had a transplant

Continuous inhaled steroids this year			
	steroid_inhaled	Frequency	Percent
	Missing/Unknown	3	0.33
	No	606	66.74
	Yes	299	32.93

COMPLICATIONS AND THERAPY

7.19 Continuous use of oral steroids in patients seen in 2019 who have never had a transplant

Continuous use of oral steroids this year			
	steroid_oral	Frequency	Percent
	Missing/Unknown	2	0.22
	No	874	96.26

Yes	32	3.52
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COMPLICATIONS AND THERAPY

7.20 Continuous use of proton pump inhibitors in patients seen in 2019 who have never had a transplant

Continuous use of proton pump inhibitors this year		
PPI	Frequency	Percent
Missing/Unknown	8	0.88
No	714	78.63
Yes	186	20.48

COMPLICATIONS AND THERAPY

7.21 Use of Ivacaftor in patients seen in 2019 who had never had a transplant (eligible or not for Ivacaftor)

	Eligibility for Ivacaftor			
	No		Yes	
	Use of Ivacaftor this year		Use of Ivacaftor this year	
	No	Yes	No	Yes
	N	N	N	N
Age at follow-up				
0-1	34	.	.	.
2-5	97	.	.	1

6-11	159	.	.	2
12-17	120	2	.	5
>=18	469	9	6	4
Total	879	11	6	12

COMPLICATIONS AND THERAPY

7.22 Use of Lumacaftor/Ivacaftor in patients seen in 2019 who had never had a transplant (eligible or not for Lumacaftor/Ivacaftor)

COMPLICATIONS AND THERAPY

7.23 Use of Tezacaftor/Ivacaftor in patients seen in 2019 who had never had a transplant (eligible or not for Tezacaftor/Ivacaftor)

	Eligibility for Tezacaftor/Ivacaftor	
	No	
	Use of Tezacaftor and Ivacaftor this year	
	No	Yes

	N	N
Age at follow-up		
0-1	34	.
2-5	98	.
6-11	156	5
12-17	121	6
>=18	452	36
Total	861	47

COMPLICATIONS AND THERAPY

7.24 Use of Elexacaftor/Tezacaftor/Ivacaftor in patients seen in 2019 who had never had a transplant

	Use of Elexacaftor, Tezacaftor and Ivacaftor this year	
	No	Yes
	N	N
Age at follow-up		
0-1	34	.
2-5	98	.
6-11	161	.
12-17	126	1
>=18	470	18
Total	889	19

TRANSPLANTATION**8.1 Number of patients living with/without lung transplant. Patients alive on 31/12/2019**

Lung transplant		
lung_t	Frequency	Percent
No	911	93.82
Yes	60	6.18

TRANSPLANTATION**8.2 Number of patients living in 2019 with transplanted lung(s), by age and gender**

Table of age_fupl by Gender			
age_fupl	Gender(Gender)		
Frequency	M	F	Total
15-19	1	1	2
20-24	1	6	7
25-29	3	6	9
30-34	4	2	6
35-39	9	3	12
40-44	5	4	9
45+	11	4	15
Total	34	26	60

TRANSPLANTATION

8.3 Number of lung transplants in 2019 by age (only alive patients)

Table of age_fupl by lung_t_yy		
age_fupl	lung_t_yy(Year of the latest lung transplant)	
Frequency	2019	Total
20-24	1	1
25-29	1	1
45+	1	1
Total	3	3

TRANSPLANTATION

8.4 Number of patients living with/without liver transplant. Patients alive on 31/12/2019

Liver transplant		
liver_t	Frequency	Percent
No	963	99.18
Yes	8	0.82

TRANSPLANTATION

8.5 Number of patients living in 2019 with transplanted liver, by age and gender

Table of age_fupl by Gender			
age_fupl	Gender(Gender)		
Frequency	M	F	Total
15-19	1	0	1

20-24	2	0	2
30-34	3	0	3
40-44	0	1	1
45+	1	0	1
Total	7	1	8

TRANSPLANTATION

8.7 Number of patients living with/without kidney transplant. Patients alive on 31/12/2019

Kidney transplant		
kidney_t	Frequency	Percent
No	967	99.59
Yes	4	0.41

TRANSPLANTATION

8.8 Number of patients living in 2019 with transplanted kidney, by age and gender

Table of age_fupl by Gender			
age_fupl	Gender(Gender)		
Frequency	M	F	Total
35-39	0	1	1
40-44	1	1	2
45+	1	0	1
Total	2	2	4

TRANSPLANTATION

8.10 Number of patients living with/without transplanted organ different from lung, liver and kidney. Patients alive on 31/12/2019

Other transplant		
other_t	Frequency	Percent
No	970	99.90
Yes	1	0.10

TRANSPLANTATION

8.11 Number of patients living in 2019 with transplanted organ different from lung, liver and kidney, by age and gender

Table of age_fupl by Gender		
age_fupl	Gender(Gender)	
Frequency	F	Total
40-44	1	1
Total	1	1

MORTALITY

9.1 Number of deaths in 2019

Status of patient		
status	Frequency	Percent
Deceased	4	0.41
Alive	971	99.59

MORTALITY

9.2 Number of deaths in 2019, males

Status of patient		
status	Frequency	Percent
Deceased	2	0.38
Alive	524	99.62

MORTALITY

9.3 Number of deaths in 2019, females

Status of patient		
status	Frequency	Percent
Deceased	2	0.45
Alive	447	99.55

MORTALITY

9.4 Cause of death distribution of deaths in 2019

Cause of death		
death_cause	Frequency	Percent
Respiratory	2	50.00
Non-CF related	1	25.00
Other CF related	1	25.00

MORTALITY

9.5 Age at death distribution of patients deceased in 2019

age_dthl	Frequency	Percent
21-30	3	75.00
31-40	1	25.00

MORTALITY

9.6 Age at death distribution of patients deceased in 2019, males

age_dthl	Frequency	Percent
21-30	2	100.00

MORTALITY

9.7 Age at death distribution of patients deceased in 2019, females

age_dthl	Frequency	Percent
21-30	1	50.00
31-40	1	50.00