**Supplementary Data**

**Table S1.** Diagnostic parameters of PCD patients.

|  |  |  |  |
| --- | --- | --- | --- |
| **Diagnostic parameters** | **n** | **CF** |  |
| Situs | 24 | Situs inversusSitus solitus | 1212 |
| HVMA | 21 | ImmotileDyskinetic | 165 |
| TEM | 15 | IDA + ODA defectODA defectIDA defectMicrotubule disorganizationIDA defect + microtubule disorganizationnormal | 811212 |
| IF | 11 | DNAH5DNAH5 + DNALI2DNAH5 + DNALI1RSPH9 | 8111 |
| Genetics | 16 | DNAH5ARMC4DNAAF3DNAI2DYX1C1SPAG1DNAI1CCDC39CCDC40RSPH4A | 6211111111 |

Abbreviations. n = number of subjects, HVMA = high-frequency video-microscopy analysis, TEM = transmission electron microscopy, IF = immunofluorescence

**Table S2.** Data of patients with cystic fibrosis on treatment with CFTR modulators.

|  |  |
| --- | --- |
| **Numbers of subjects** | **25** |
| CFTR modulator* Ivacaftor
* Lumacaftor/Ivacaftor
* Tezacaftor/Ivacaftor
* Tezacaftor/Ivacaftor/Elexacaftor
 | 9 (36%)2610 |

**Table S3.** Pulmonary bacterial colonization in patients with primary ciliary dyskinesia (PCD) and cystic fibrosis (CF).

|  |  |  |
| --- | --- | --- |
|  | **PCD** | **CF** |
| Number of subjects  | 23 | 25 |
| Pathological bacterial colonization* Aspergillus fumigatus
* Burkholderia cenocepacia
* Candida albicans
* Candida tropicalis
* Enterobacter cloacae
* Escherichia coli
* Haemophilus influenzae
* Haemophilus parainfluenzae
* Moraxella catarrhalis
* Methicillin-resistant Staphylococcus aureus (MRSA)
* Mycobacterium abscessus
* Neisseria flavescens
* Neisseria meningitidis
* Pseudomonas aeruginosa
* Proteus mirabilis
* Rothia mucilaginosa
* Staphylococcus aureus
* Serratia marcescens
* Streptococcus pneumoniae
 | 10 (43.48%)0010005110021101512 | 20 (80%)11011200011007101200 |

**Table S4.** Contents in plasma (µmol/L) and urine (µM or µM/mM creatinine) of metabolites of the L-Arg/NO pathway in patients with PCD or CF and in healthy controls (HC). Data are presented as median [25-75th interquartile range].

|  |  |  |  |  |
| --- | --- | --- | --- | --- |
| **Plasma** | **PCD** | **CF** | **HC** | ***p* value** |
| Number of subjects | 22 | 24 | 14 |  |
| L-Arg | 77.14[67.70 – 89.48] | 87.56[72.65 – 116.86] | 83.45[71.96 – 86.66] | 0.110 |
| h-Arg | 1.25[0.93 – 1.70] | 1.40[0.93 – 1.79] | 1.43[1.10 – 1.67] | 0.606 |
| ADMA | 0.51[0.36 – 0.55] | 0.53[0.46 – 0.59] | 0.41[0.36 – 0.52] | 0.886 |
| **Nitrite** | 23.90[22.19 – 24.92] | 22.22[21.45 – 23.62] | 21.02[19.65 – 22.64] | \* 0.163**# < 0.001****+ 0.038** |
| **Nitrate** | 63.26[56.26 – 67.75] | 81.61[73.88 – 100.86] | 86.96[70.93 – 110.41] | **\* < 0.001****# 0.002**+ 0.999 |
| **Orn/Cit** | 45.25[38.36 – 50.88] | 58.03[44.38 – 64.53] | 42.17[38.87 – 48.45] | \* 0.087# 1.00**+ 0.046** |
| L-Arg/ADMA | 166.72[144.10 – 199.50] | 176.34[136.98 – 222.85] | 172.15[158.26 – 234.17] | 0.531 |
| **Urine** |  |  |  |  |
| Number of subjects | 24 | 25 | 14 |  |
| L-Arg | 33.17[18.88 – 43.22] | 35.89[25.68 – 61.10] | 32.27[10.55 – 48.17] | 0.145 |
| hArg | 2.82[1.12 – 6.58] | 2.10[1.61 – 3.70] | 1.99[1.10 – 2.72] | 0.734 |
| ADMA/Crea | 4.08[3.21 – 5.26] | 4.29[3.41 – 4.99] | 3.22[2.57 – 3.65] | 0.077 |
| Nitrite/Crea | 0.20[0.12 – 0.45] | 0.22[0.15 – 0.34] | 0.23[0.15 – 0.66] | 0.320 |
| **Nitrate/Crea** | 54.84[44.57 – 82.64] | 93.80[75.54 – 133.66] | 74.58[57.04 – 103.02] | **\* 0.006**# 1.000+ 0.185 |
| Orn/Cit | 42.63[25.26 – 65.70] | 58.86[34.26 – 73.9] | 49.77[15.09 – 88.32] | 0.170 |
| UnoR | 311.70[175.81 – 403.66] | 530.48[268.08 – 655.81] | 204.78[168.53 – 633.59] | 0.070 |

Statistics: Bold indicates statistical significance; \* = Significance PCD vs. CF; # = Significance PCD vs. HC; + = Significance CF vs. HC

Abbreviations. L-Arg, L-Arginine; hArg, Homoarginine, ADMA, asymmetric dimethylarginine; Crea = creatinine; Orn/Cit, Ornithine/Citrulline ratio; UnoR = Urinary nitrate/nitrite ratio

**Figure S5.** Correlation between L-Arginine (µM/mg sputum, y-axis) and Ornithine/Citrulline ratio (µM/mg sputum, x-axis) in sputum of patients with primary ciliary dyskinesia (PCD), cystic fibrosis (CF) and healthy control (HC). R = Pearson correlation coefficient.