**Table S1.** **Electron microscopy, NO measurements and situs status in the PCD patients studied**

|  |  |  |  |
| --- | --- | --- | --- |
| Patient # | EM | NO (ppb) | *Situs inversus* |
| 1  | ODA/IDA | 67.1/120.0/90.1/133/177 | – |
| 2  | ND | 188/481/137 | – |
| 3  | ODA/IDA | 25.5/33 | – |
| 4\*  | ODA/IDA | 75.6/59.9/116 | – |
| 5  | ODA/IDA | 626/669/687 | + |
| 6  | IDA | ND | + |

\*Two *DNAH5*  mutations; EM electron microscopy; ODA/IDA: lack of both outer and inner dynein arms; IDA: isolated lack of inner dynein arms; NO: nitric oxide measurements; ppb: parts per billion; ND: not determined.