|  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- |
| Frequency | Center 1 | Center 2 | Center 3 | Center 4 | Center 5 | Center 6 |
| Less than every 3 months | 8.7% | 0.0% | 0.0% | 1.9% | 0.0% | 0.0% |
| Every 3 to 5 months | 26.1% | 0.0% | 26.9% | 7.8% | 11.9% | 10.0%  |
| Every 6 to 11 months | 52.2% | 21.4% | 21.2% | 36.9% | 23.7% | 30.0% |
| Less than once a year | 13.0% | 78.6% | 51.9% | 53.4% | 64.4% | 60.0% |
| Total | 100.0% | 100.0% | 100.0% | 100.0% | 100.0% | 100.0% |

Table A: frequency of sweat tests performed in CRMS/CFSPID subjects by centre

|  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- |
|  | nr | Group A° | nr | Group B°° | P |
|   |  |  |  |  |  |
| Median IRT2 (ng/ml)  | 53 | 45.6 | 46 | 113 | 0.015\* |
| IQR§ 25-75 |  | 37.8-50.8 |  | 23-55 |  |
| Median Sweat chloride (mmol/l)  | 139 | 19 | 186 | 33 | <0.010\* |
| IQR 25-75 |  | 12-26 |  | 37-45 |  |
|  |  |  |  |  |  |
|  ***Respiratory Infection*** |  |  |  |  |  |
|  *Isolation of PA* | 18/117 | 15.4% | 29/111 | 26.1% | 0.045\*\* |
|  *Isolation of MSSA* |  |  |  |  | <0.001\*\* |
| *At least one Isolation*  | 57/117 | 48.7% | 27/111 | 9.4% |  |
| *Chronic*  | 11/117 | 9.4% | 2/111 | 1.8% |  |
| *Isolation of HI* | 44/117 | 37.6% | 23/110 | 20.9% | 0.022\*\* |
|  |  |  |  |  |  |
|  ***Diagnostic tools performed*** |  |  |  |  |  |
|  *Oropharyngeal swab* | 111/137 | 81.0% | 107/181 | 59.1% | <0.001 \*\* |

Table B: Main differences in clinical characteristics between CRMS/CFSPID subjects with sweat test in normal or intermediate range

§IQR= Inter Quartile Range 25-75; \*Test T; \*\*chi square test

° CRMS/CFSPID subjects having sweat chloride <30 mmol/L and 2 CFTR variants, at least one of which having unclear phenotypic consequences;

°° CRMS/CFSPID subjects having sweat chloride 30–59 mmol/L and 1 or 0 CF-causing pathogenetic variant.

Abbreviations: MSSA: methicillin sensible staphylococcus aureus; HI: haemophilus influentiae; PA: pseudomonas aeruginosa.

|  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- |
|  | Center 1 | Center 2 | Center 3 | Center 4 | Center 5 | Center 6 |
| ***Diagnostic tools performed*** |  |  |  |  |  |  |
| Chest X-rays | 34.5% | 43.8% | 8.2% | 66.9% | 63.5% | 100.0% |
| Lung CT scan | 17.2% | 0.0% | 0.0% | 0.7% | 5.4% | 0.0% |
| Throat swab or nasopharyngeal aspiration | 79.3% | 43.8% | 16.4% | 71.3% | 94.6% | 100.0% |
| ***Therapies*** |  |  |  |  |  |  |
| Salt supplementation | 89.7% | 56.3% | 11.5% | 34.6% | 10.8% | 54.5% |
| Respiratory physiotherapy | 24.1% | 31.3% | 1.6% | 44.1% | 20.3% | 54.5% |

Table C: Percentage of CRMS/CFSPID subjects who underwent at least once each type of diagnostic intervention or treatment. Differences among centers.