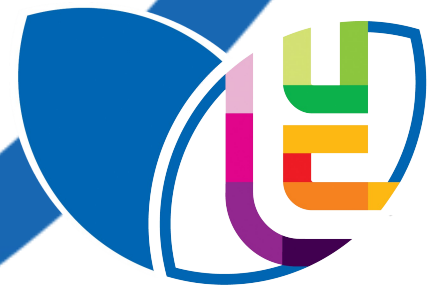


**La dyskinésie ciliaire primitive, une pathologie pour le  
kinésithérapeute :**

**Quelle prise en charge pour le kinésithérapeute ?**

Thibault COPPENS,

*Kinésithérapeute, Service de Médecine physique Pneumologie*

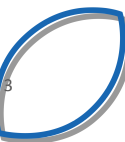


Cliniques universitaires  
**SAINT-LUC**  
UCL BRUXELLES

- Dyskinésie ciliaire (DCP) = mucoviscidose ?
- Introduction prise en charge
- Prise en charge
- Conclusion et Take home message

## Dyskinésie ciliaire = mucoviscidose ?

- Patients sous diagnostiqués => orientés vers centres mucos
- Patients traités par les mêmes équipes
- Nombre nettement plus restreint (sous-diagnostiqués ?)
- Même type d'atteinte mais...



## Mucoviscidose

Inflammation dès la naissance

Dégradation progressive

3,6% de déclin de VEMS par an moyen

« dysfonction » du mucus

## Dyskinésie ciliaire primitive

Pas d'inflammation à la naissance

Si traité, stabilisation

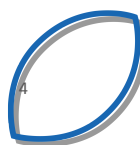
± 0,8% de déclin de VEMS par an

« dysfonction » des cils

Table 1—Comparison of Patient Demographics and Sputum Properties\*

| Variables                             | CF            | PCD           | p Value  |
|---------------------------------------|---------------|---------------|----------|
| Age, yr                               | 10.8 (2.6)    | 9.5 (3.0)     | 0.10     |
| FEV <sub>1</sub> , % predicted        | 62 (23)       | 65 (8)        | 0.60     |
| FVC, % predicted                      | 80 (24)       | 79 (15)       | 0.84     |
| DNA, µg/mL                            | 1.16 (1.2)    | 0.44 (0.4)    | 0.11     |
| IL-8, ng/mL                           | 163 (130)     | 606 (387)     | < 0.0001 |
| G' (elasticity), dyne/cm <sup>2</sup> | 905 (769)     | 1,651 (3,009) | 0.15     |
| G'' (viscosity), dyne/cm <sup>2</sup> | 1,190 (1,621) | 1,949 (2,730) | 0.21     |
| Cohesivity, mm                        | 15.8 (8.8)    | 11.7 (4.1)    | 0.10     |
| Wettability degrees                   | 43.6 (12.0)   | 47.3 (11.5)   | 0.23     |
| Interfacial tension dyne/cm           | 79.1 (17.0)   | 82.9 (15.2)   | 0.45     |
| Percentage of solids                  | 7.0 (2.7)     | 5.7 (1.7)     | 0.07     |
| Tangent δ (dimensionless)             | 8.44 (39)     | 2.48 (4.2)    | 0.57     |
| MCTR, normalized                      | 0.74 (0.27)   | 0.79 (0.14)   | 0.46     |
| CTR, cm                               | 24.6 (8.7)    | 25 (5.7)      | 0.69     |

\*Data are presented as mean (SD). MCTR = mucociliary transportability of sputum.



## Qualité de vie et DCP

- 78/91 (96%) réponses
- 34 hommes et 44 femmes
- Age entre 1,7 et 48,5 ans ( $21,4 \pm 12,9$  ans)
- Questionnaires utilisés : SGRQ, SF-36, questionnaire spécifique DCP



Source: M. Pifferi, Eur Respir J 2010; 35: 787–794



## Qualité de vie et DCP

### Résultats :

**TABLE 1** Multiple linear regression analysis on dependent and independent variables

| Dependent variable | Independent variables |         |                      |         |                          |         |
|--------------------|-----------------------|---------|----------------------|---------|--------------------------|---------|
|                    | Age yrs               |         | Age at diagnosis yrs |         | Time since diagnosis yrs |         |
|                    | $\beta^{\#}$          | p-value | $\beta$              | p-value | $\beta$                  | p-value |
| SGRQ symptoms      | NS                    | NS      | 0.243                | 0.040   | 0.242                    | 0.041   |
| SGRQ activity      | NS                    | NS      | 0.404                | 0.001   | NS                       | NS      |
| SGRQ impacts       | NS                    | NS      | 0.452                | <0.0001 | 0.464                    | <0.0001 |
| SF-36 PCS          | NS                    | NS      | NS                   | NS      | NS                       | NS      |
| SF-36 MCS          | NS                    | NS      | -0.388               | 0.001   | -0.351                   | 0.002   |

SGRQ: St George's Respiratory Questionnaire; SF-36: Short Form-36; PCS: physical component summary; MCS: mental component summary; NS: not significant.  $\beta^{\#}$ : regression coefficient.

## Qualité de vie et DCP

### Résultats :

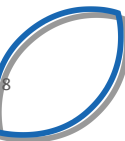
**TABLE 3** Treatments used by primary ciliary dyskinesia patients

|                              | Usage      |             |            | Correlation of usage |                      |                          |
|------------------------------|------------|-------------|------------|----------------------|----------------------|--------------------------|
|                              | Never used | In the past | At present | Age yrs              | Age at diagnosis yrs | Time since diagnosis yrs |
| Daily physiotherapy          | 10 (12.82) | 10 (12.82)  | 58 (74.36) | -0.125 (NS)          | 0.097 (NS)           | -0.230 (0.04)            |
| Regular antibiotics          | 9 (11.54)  | 13 (16.67)  | 56 (71.79) | 0.147 (NS)           | 0.186 (NS)           | -0.003 (NS)              |
| Regular bronchodilators      | 10 (12.82) | 14 (17.95)  | 54 (69.23) | -0.069 (NS)          | 0.107 (NS)           | -0.176 (NS)              |
| Intermittent bronchodilators | 20 (25.64) | 11 (14.10)  | 47 (60.26) | -0.006 (NS)          | -0.043 (NS)          | 0.032 (NS)               |
| Mucolytics <sup>#</sup>      | 17 (21.79) | 29 (37.18)  | 32 (41.03) | 0.258 (0.02)         | 0.244 (0.03)         | 0.069 (NS)               |

Data are presented as n (%), for usage and r-value (p-value) for correlation of usage. NS: not significant. <sup>#</sup>: N-acetylcysteine.

## Suivi spirométrique...

- 24 patients suivis minimum 2 ans
- Suivi : FVC, FEV<sub>1</sub> (3 mois), prélèvement bactériologique (1 mois)
- Traitement : antibiotique ciblé, kinésithérapie respiratoire (minimum 2x10 min PEP à 10 cmH<sub>2</sub>O)



## Suivi spirométrique...

### Résultats :

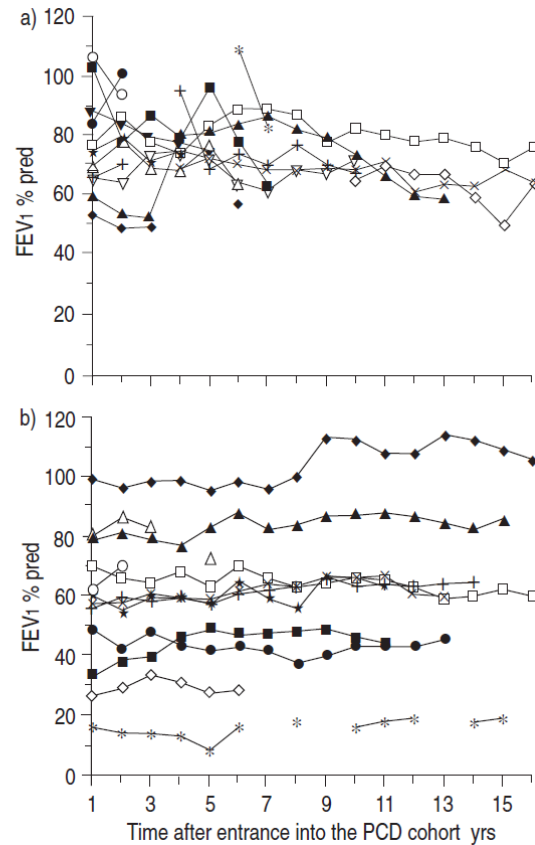
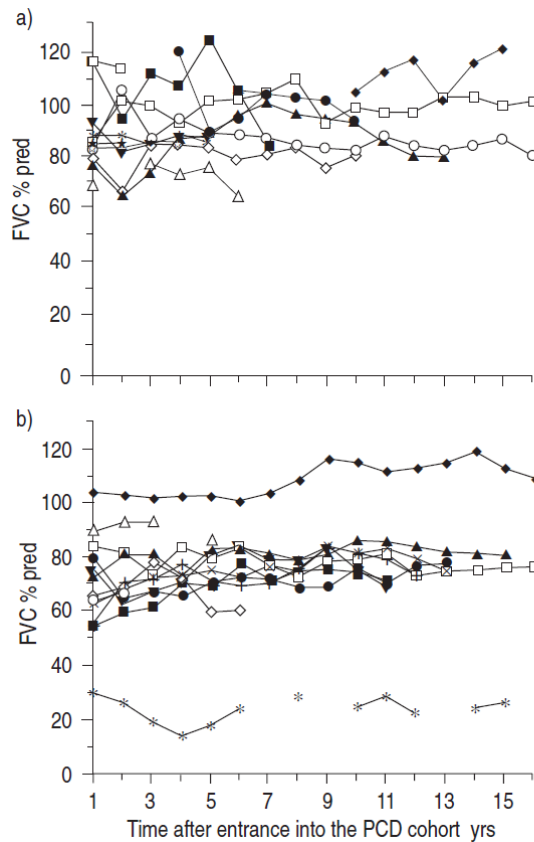
Table 1. – Patient characteristics and bacterial growth in sputum samples from lower airways obtained from the primary ciliary dyskinesia (PCD) cohort

| Patient No.                  | Age at entrance to cohort | Age by end of 1994 | Presence of bronchiectasis† | FVC<br>% pred |          | FEV1<br>% pred |          | Cilia function | Hi | P  | Sa | Pa |
|------------------------------|---------------------------|--------------------|-----------------------------|---------------|----------|----------------|----------|----------------|----|----|----|----|
|                              |                           |                    |                             | 1st year      | End 1994 | 1st year       | End 1994 |                |    |    |    |    |
| yrs                          |                           |                    |                             |               |          |                |          |                |    |    |    |    |
| yrs                          |                           |                    |                             |               |          |                |          |                |    |    |    |    |
| % of positive cultures       |                           |                    |                             |               |          |                |          |                |    |    |    |    |
| ≤18 yrs of age at enrollment |                           |                    |                             |               |          |                |          |                |    |    |    |    |
| 1                            | 2                         | 11                 | -                           | 69            | 74       | 70             | 64       | A              | 95 | 40 | 0  | 0  |
| 2                            | 4                         | 13                 | -                           | 87            | 85       | 88             | 74       | A              | 83 | 27 | 0  | 0  |
| 3                            | 6                         | 15                 | -                           | 116           | 84       | 103            | 63       | A              | 71 | 29 | 50 | 0  |
| 4                            | 6*                        | 13                 | -                           | 83            | 104      | 84             | 83       | A              | 50 | 54 | 46 | 7  |
| 5                            | 7                         | 9                  | -                           | 117           | 114      | 107            | 93       | A              | 44 | 29 | 0  | 0  |
| 6                            | 8*                        | 17                 | -                           | 85            | 93       | 65             | 67       | A              | 92 | 23 | 8  | 0  |
| 7                            | 11                        | 20                 | +                           | 80            | 79       | 65             | 71       | I              | 45 | 24 | 12 | 10 |
| 8                            | 11                        | 26                 | -                           | 93            | 120      | 53             | 64       | A              | 52 | 16 | 55 | 0  |
| 9                            | 12                        | 16                 | -                           | 83            | 85       | 74             | 74       | A              | 73 | 18 | 32 | 0  |
| 10                           | 13                        | 25                 | +                           | 77            | 79       | 60             | 59       | A              | 39 | 23 | 23 | 13 |
| 11                           | 17                        | 32                 | -                           | 85            | 100      | 76             | 76       | A              | 87 | 24 | 17 | 2  |
| 12                           | 18                        | 34                 | -                           | 87            | 79       | 59             | 64       | A              | 73 | 3  | 35 | 1  |
| Median                       | 9.5                       | 16.5               |                             | 85            | 85       | 72             | 69       |                | 72 | 24 | 20 | 0  |
| >18 yrs of age at enrollment |                           |                    |                             |               |          |                |          |                |    |    |    |    |
| 13                           | 22                        | 26                 | +                           | 90            | 87       | 81             | 73       | A              | 50 | 7  | 43 | 0  |
| 14                           | 24                        | 36                 | -                           | 80            | 76       | 49             | 60       | I              | 20 | 20 | 16 | 27 |
| 15                           | 26                        | 41                 | -                           | 104           | 110      | 99             | 106      | H              | 38 | 6  | 43 | 20 |
| 16                           | 28                        | 38                 | -                           | 76            | 72       | 61             | 64       | A              | 40 | 8  | 1  | 36 |
| 17                           | 29                        | 43                 | +                           | 74            | 82       | 79             | 86       | A              | 71 | 20 | 5  | 0  |
| 18                           | 30                        | 45                 | +                           | 84            | 77       | 70             | 60       | I              | 83 | 16 | 3  | 2  |
| 19                           | 30                        | 42                 | +                           | 63            | 78       | 57             | 45       | A              | 63 | 19 | 18 | 7  |
| 20                           | 36                        | 46                 | -                           | 55            | 69       | 33             | 44       | A              | 75 | 28 | 14 | 0  |
| 21                           | 36                        | 41                 | -                           | 66            | 61       | 26             | 28       | A              | 84 | 34 | 13 | 0  |
| 22                           | 33                        | 48                 | +                           | 30            | 27       | 16             | 19       | A              | 1  | 1  | 8  | 97 |
| 23                           | 43                        | 56                 | +                           | 55            | 79       | 55             | 64       | H              | 83 | 19 | 0  | 2  |
| 24                           | 53                        | 55                 | -                           | 64            | 66       | 52             | 70       | I              | 0  | 0  | 0  | 0  |
| Median                       | 30                        | 44                 |                             | 70            | 76.5     | 59             | 62       |                | 57 | 18 | 11 | 2  |

\*: pair of siblings with primary ciliary dyskinesia. †: detected by radiograph. FVC: forced vital capacity; FEV<sub>1</sub>: forced expiratory volume in one second; % pred: percentage of predicted values; +: present; -: absent; A: asynchronous; I: immotile; H: hyperfrequent. Hi: *Hemophilus influenzae*; P: *Pneumococcus*; Sa: *Staphylococcus aureus*; Pa: *Pseudomonas aeruginosa*.

## Suivi spirométrique...

### Résultats



## Suivi spirométrique...

### Résultats :

- Importance du suivi régulier des patients
- Importance d'une prise en charge précoce
- Importance d'avoir des centres de référence

## Prise en charge

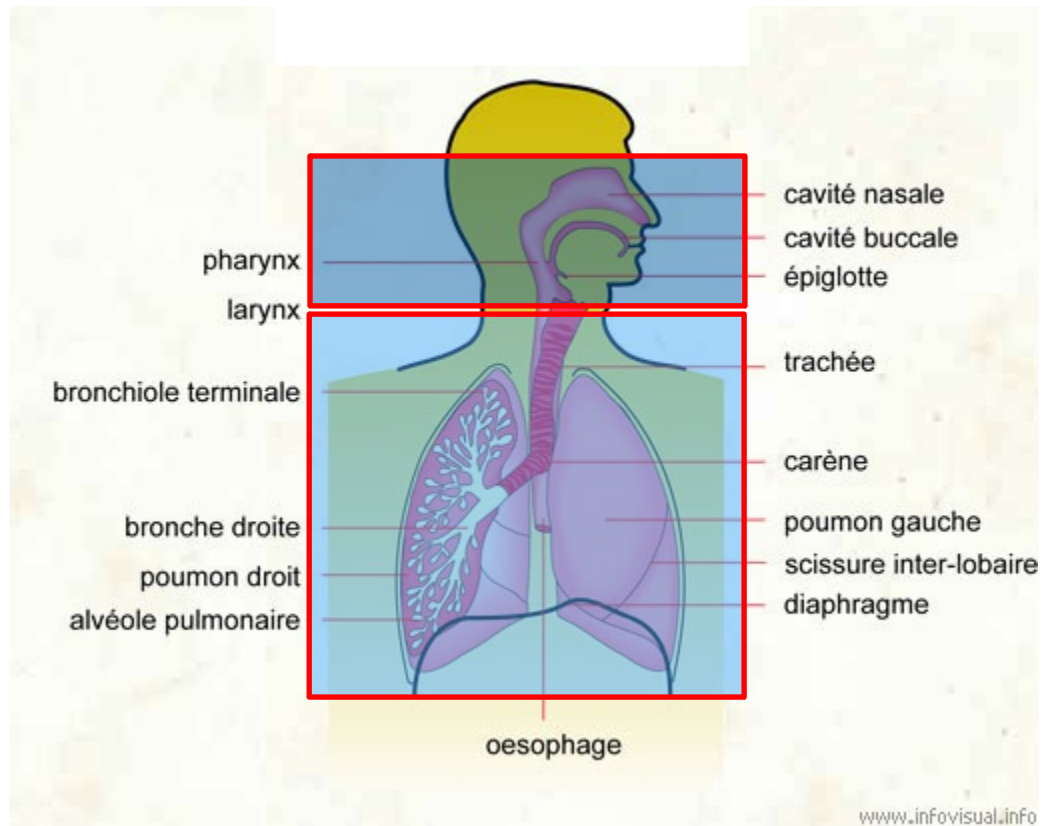
- Non basée sur l'Evidence Based Medecine
- Basée sur la prise en charge des patients mucos
- La plus précoce possible



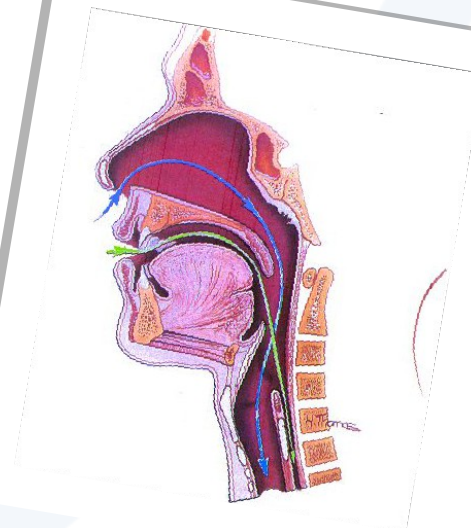
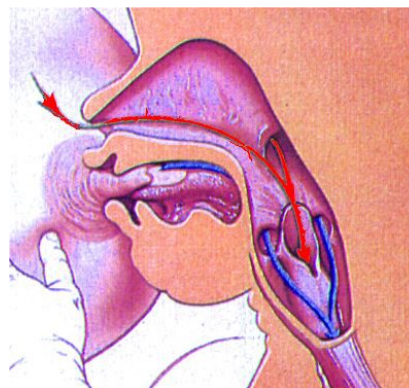
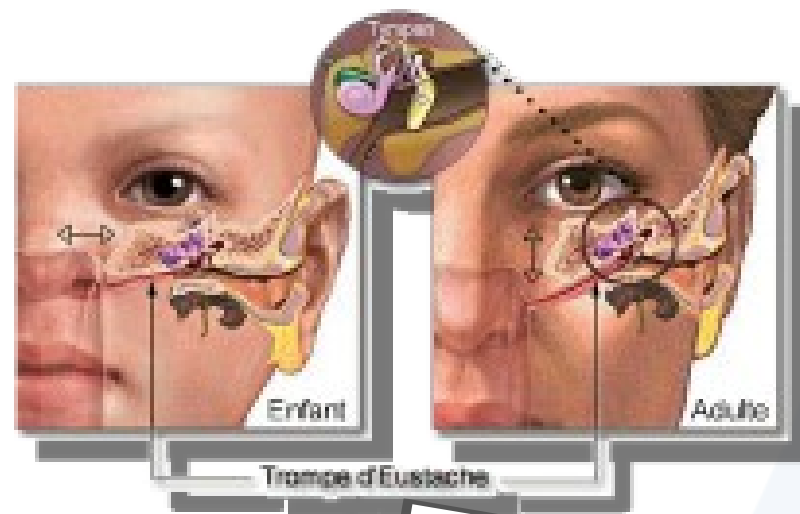
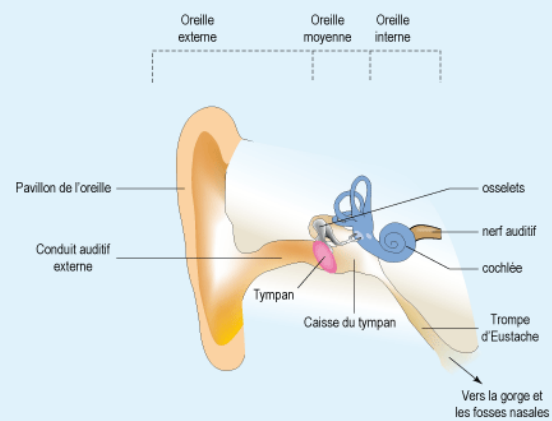
Source: B. Fauroux, Paediatric Respiratory Reviews 10 (2009) 55–57

M. Pifferi, Eur Respir J 2010; 35: 787–794

## Quelle kinésithérapie ?



## Voies aériennes supérieures (VAS)



## Voies aériennes supérieures (VAS)



Coupe de la région nasale de face

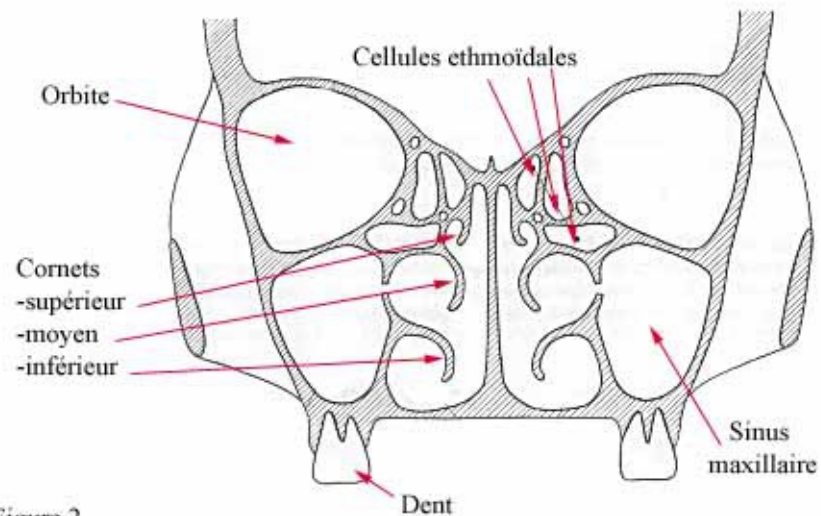
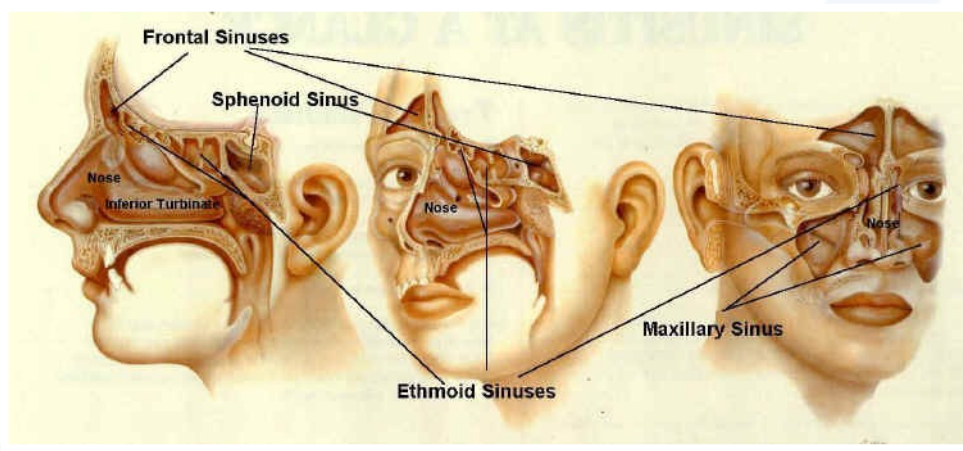


Figure 2



## Voies aériennes supérieures (VAS)

Coupe verticale montrant la paroi externe de la fosse nasale droite

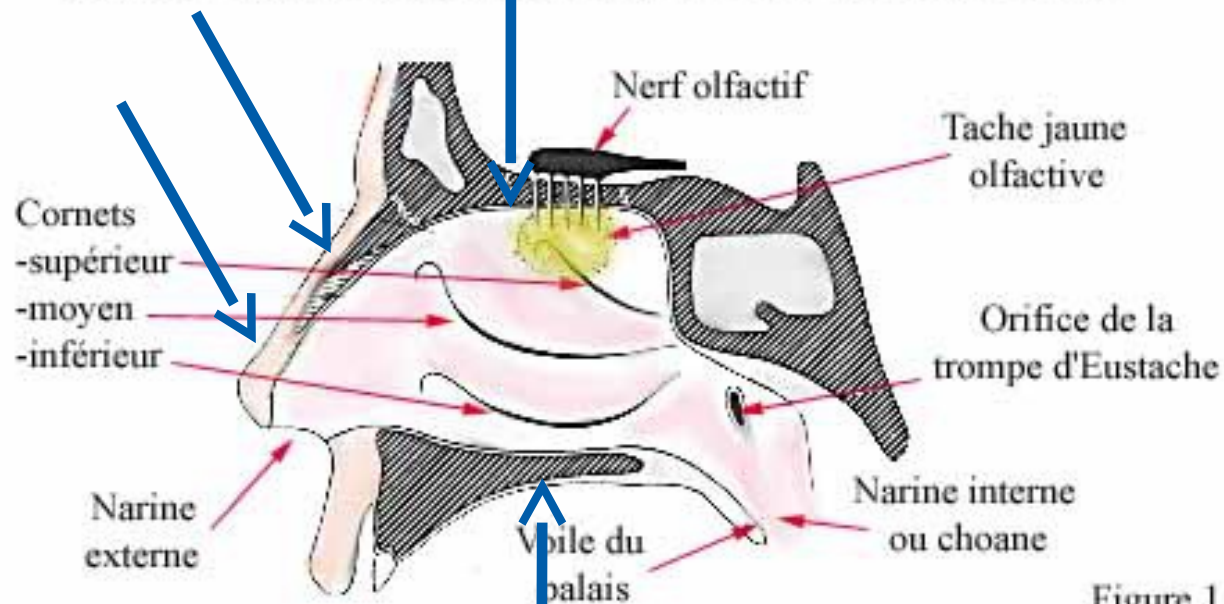
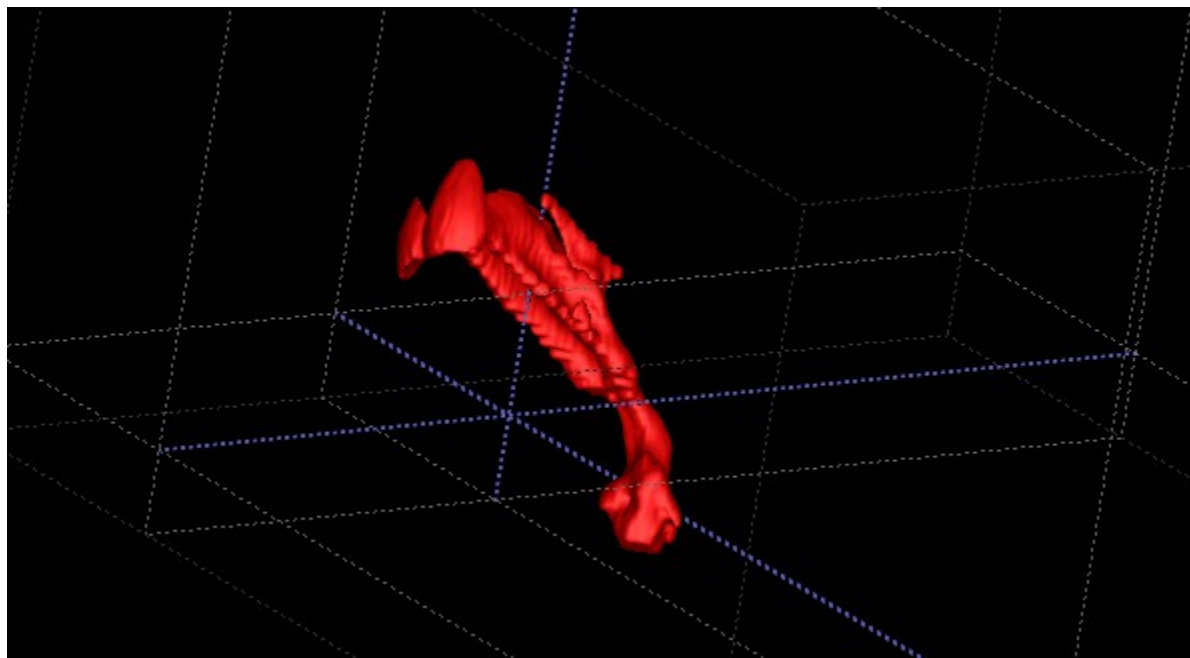


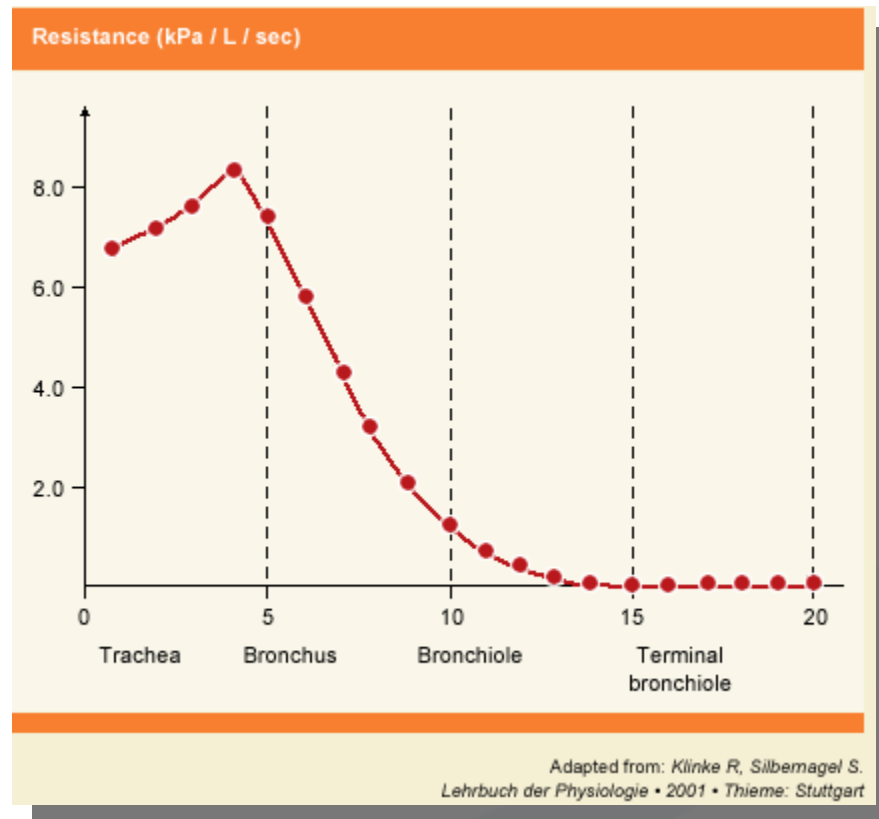
Figure 1



## Voies aériennes supérieures (VAS)

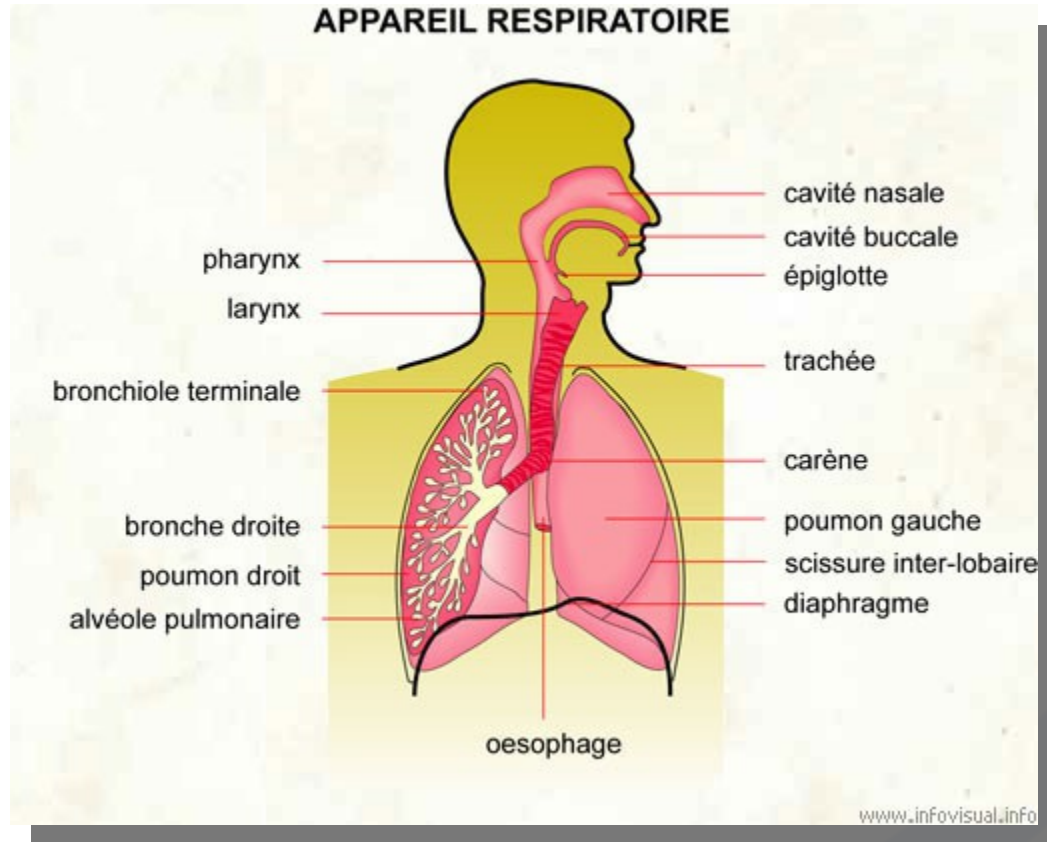


## Voies aériennes supérieures (VAS)



Adulte

Pédiatrique



1/4

3/4

1/3

2/3



## Voies aériennes supérieures (VAS)

- 390 enfants (6-10 ans)
- « Common cold or flu »
- Traitement « classique » (+ serum physiologique)
- Evaluation en phase aigue et ensuite prévention



Source: Slapak I., Arch Otolaryngol Head Neck Surg. 2008;134(1):67-74

# Efficacy of Isotonic Nasal Wash (Seawater) in the Treatment and Prevention of Rhinitis in Children

Ivo Šlapak, MD; Jana Skoupá, MD; Petr Strnad, MD; Pavel Horník, MD

## Voies aériennes supérieures (VAS)

**Table 3. Efficacy Parameters at Visit 2 (Acute Phase)<sup>a</sup>**

| Characteristic                     | Control<br>(n=101) | Isotonic Saline Wash              |                                   |                                     |                                  |
|------------------------------------|--------------------|-----------------------------------|-----------------------------------|-------------------------------------|----------------------------------|
|                                    |                    | Medium Jet<br>(n=99) <sup>b</sup> | Fine Spray<br>(n=95) <sup>b</sup> | Dual Formula<br>(n=95) <sup>b</sup> | All Wash<br>(n=289) <sup>c</sup> |
| Rhinologic score <sup>d</sup>      |                    |                                   |                                   |                                     |                                  |
| Sore throat                        | 1.23 [0.47]        | 1.08 [0.27]                       | 1.07 [0.26]                       | 1.11 [0.43]                         | 1.09 [0.33] <sup>e</sup>         |
| Dry cough                          | 1.14 [0.38]        | 1.09 [0.35]                       | 1.14 [0.43]                       | 1.12 [0.35]                         | 1.11 [0.38]                      |
| Productive cough                   | 1.38 [0.63]        | 1.21 [0.54]                       | 1.22 [0.49]                       | 1.25 [0.62]                         | 1.23 [0.55]                      |
| Nasal secretion                    | 2.10 [0.74]        | 1.76 [0.67] <sup>e</sup>          | 1.86 [0.79]                       | 1.74 [0.67] <sup>e</sup>            | 1.79 [0.71] <sup>e</sup>         |
| Sneezing                           | 1.06 [0.24]        | 1.01 [0.10]                       | 1.06 [0.29]                       | 1.06 [0.25]                         | 1.04 [0.22]                      |
| Itching                            | 1.06 [0.24]        | 1.01 [0.10]                       | 1.03 [0.23]                       | 1.04 [0.20]                         | 1.03 [0.18]                      |
| Loss of smell/taste                | 1.09 [0.35]        | 1.01 [1.01]                       | 1.00 [0.00]                       | 1.00 [0.00]                         | 1.00 [0.06]                      |
| Nasal secretion type <sup>f</sup>  | 2.06 [0.69]        | 1.74 [0.65] <sup>e</sup>          | 1.74 [0.62] <sup>e</sup>          | 1.69 [0.65] <sup>e</sup>            | 1.72 [0.64] <sup>e</sup>         |
| Nasal breathing score <sup>g</sup> | 1.58 [0.68]        | 1.27 [0.51] <sup>e</sup>          | 1.27 [0.56] <sup>e</sup>          | 1.20 [0.50] <sup>e</sup>            | 1.25 [0.52] <sup>e</sup>         |
| Medication                         |                    |                                   |                                   |                                     |                                  |
| Antipyretics                       | 13 (12.9)          | 5 (5.1)                           | 7 (7.4)                           | 10 (10.5)                           | 22 (7.6)                         |
| Nasal decongestants                | 36 (35.6)          | 15 (15.2) <sup>e</sup>            | 13 (13.7) <sup>e</sup>            | 18 (18.9) <sup>e</sup>              | 46 (15.9) <sup>e</sup>           |
| Mucolytics                         | 32 (31.7)          | 15 (15.2) <sup>e</sup>            | 14 (14.7) <sup>e</sup>            | 21 (22.1)                           | 50 (17.3) <sup>e</sup>           |
| Systemic antibiotics               | 9 (8.9)            | 7 (7.1)                           | 6 (6.3)                           | 3 (3.2)                             | 16 (5.5)                         |
| Health status score <sup>h</sup>   |                    |                                   |                                   |                                     |                                  |
| Entry during cold                  | 2.60 [1.02]        | 1.88 [0.88] <sup>e</sup>          | 1.93 [0.79] <sup>e</sup>          | 1.80 [0.84] <sup>e</sup>            | 1.87 [0.84] <sup>e</sup>         |
| Entry during flu                   | 2.00 [0.91]        | 1.63 [0.75]                       | 1.58 [0.79]                       | 1.57 [0.68]                         | 1.59 [0.74] <sup>e</sup>         |

<sup>a</sup>All data are reported as mean [SD] score or number (percentage) of patients.

<sup>b</sup>Multiple comparisons of treatments vs control if Kruskal-Wallis or logistic regression results were significant.

<sup>c</sup>Mann-Whitney or  $\chi^2$  tests.

<sup>d</sup>Rhinologic symptom scores: 1, no symptoms; 2, mild; 3, moderate; and 4, severe.

<sup>e</sup> $P < .05$ .

<sup>f</sup>Secretion types: 1, absent; 2, serosal; 3, seropurulent; and 4, purulent.

<sup>g</sup>Breathing scores: 1, without any difficulty; 2, minor difficulties; 3, difficult; and 4, impossible.

<sup>h</sup>Health status scores: 1, cured; 2, significant improvement; 3, partial improvement; and 4, no change.

# Efficacy of Isotonic Nasal Wash (Seawater) in the Treatment and Prevention of Rhinitis in Children

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## Voies aériennes supérieures (VAS)

Table 4. Efficacy Parameters at Visit 3 (Preventive Phase)<sup>a</sup>

### Isotonic Saline Wash

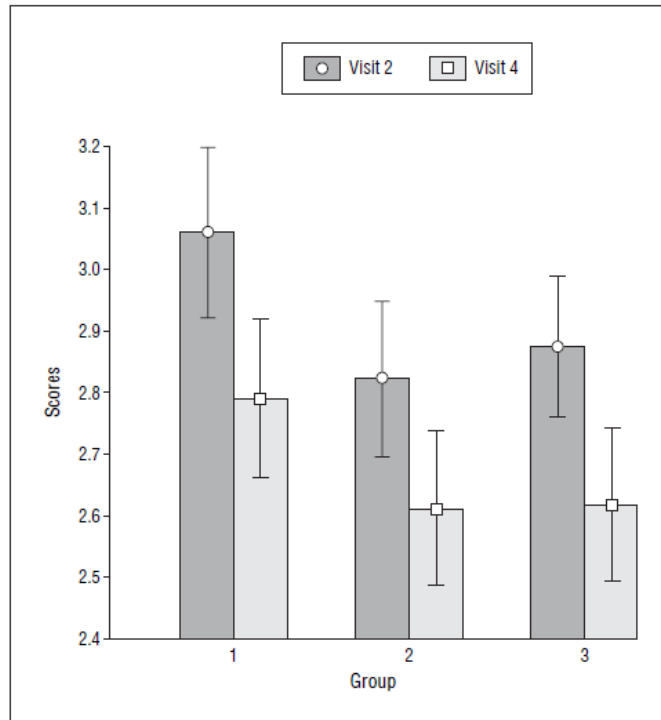
Table 5. Efficacy Parameters at Visit 4 (Preventive Phase)<sup>a</sup>

| Characteristic                     | Isotonic Saline Wash |                                |                                |                                  |                               |
|------------------------------------|----------------------|--------------------------------|--------------------------------|----------------------------------|-------------------------------|
|                                    | Control (n=101)      | Medium Jet (n=99) <sup>b</sup> | Fine Spray (n=95) <sup>b</sup> | Dual Formula (n=94) <sup>b</sup> | All Wash (n=288) <sup>c</sup> |
| Rhinologic score <sup>d</sup>      |                      |                                |                                |                                  |                               |
| Sore throat                        | 1.12 [0.41]          | 1.06 [0.24]                    | 1.04 [0.25]                    | 1.03 [0.23]                      | 1.05 [0.24]                   |
| Dry cough                          | 1.04 [0.24]          | 1.02 [0.14]                    | 1.04 [0.20]                    | 1.03 [0.23]                      | 1.03 [0.19]                   |
| Productive cough                   | 1.13 [0.46]          | 1.02 [0.20]                    | 1.03 [0.23]                    | 1.02 [0.15]                      | 1.02 [0.19]                   |
| Nasal secretion                    | 1.55 [0.78]          | 1.20 [0.55] <sup>e</sup>       | 1.24 [0.50]                    | 1.23 [0.50]                      | 1.23 [0.52] <sup>e</sup>      |
| Sneezing                           | 1.10 [0.30]          | 1.04 [0.20]                    | 1.05 [0.22]                    | 1.10 [0.36]                      | 1.06 [0.27]                   |
| Itching                            | 1.11 [0.31]          | 1.04 [0.24]                    | 1.05 [0.27]                    | 1.04 [0.25]                      | 1.05 [0.25]                   |
| Loss of smell/taste                | 1.11 [0.37]          | 1.00 [0.10]                    | 1.02 [0.14]                    | 1.01 [0.10]                      | 1.01 [0.10]                   |
| Nasal secretion type <sup>f</sup>  | 1.53 [0.81]          | 1.16 [0.51] <sup>e</sup>       | 1.23 [0.52]                    | 1.23 [0.54]                      | 1.21 [0.52] <sup>e</sup>      |
| Nasal breathing score <sup>g</sup> | 1.39 [0.60]          | 1.10 [0.36] <sup>e</sup>       | 1.12 [0.32]                    | 1.17 [0.43]                      | 1.13 [0.37] <sup>e</sup>      |
| Medication                         |                      |                                |                                |                                  |                               |
| Antipyretics                       | 20 (19.8)            | 8 (8.1) <sup>e</sup>           | 5 (5.3) <sup>e</sup>           | 6 (6.4) <sup>e</sup>             | 19 (6.6) <sup>e</sup>         |
| Nasal decongestants                | 43 (42.6)            | 4 (4.0) <sup>e</sup>           | 7 (7.4) <sup>e</sup>           | 0 (0.0) <sup>e</sup>             | 11 (3.8) <sup>e</sup>         |
| Mucolytics                         | 24 (23.8)            | 4 (4.0) <sup>e</sup>           | 4 (4.2) <sup>e</sup>           | 6 (6.4) <sup>e</sup>             | 14 (4.9) <sup>e</sup>         |
| Systemic antibiotics               | 9 (8.9)              | 6 (6.1)                        | 3 (3.2)                        | 3 (3.2)                          | 12 (4.2)                      |
| Illness and complications          |                      |                                |                                |                                  |                               |
| Reported illness                   | 53 (52.5)            | 22 (22.2) <sup>e</sup>         | 21 (22.1) <sup>e</sup>         | 21 (22.3) <sup>e</sup>           | 64 (22.2) <sup>e</sup>        |
| Reported school absence            | 25 (24.8)            | 7 (7.1) <sup>e</sup>           | 8 (8.4) <sup>e</sup>           | 10 (10.6) <sup>e</sup>           | 25 (8.7) <sup>e</sup>         |
| Complications                      | 14 (13.9)            | 6 (6.1)                        | 3 (3.2) <sup>e</sup>           | 3 (3.2) <sup>e</sup>             | 12 (4.2) <sup>e</sup>         |

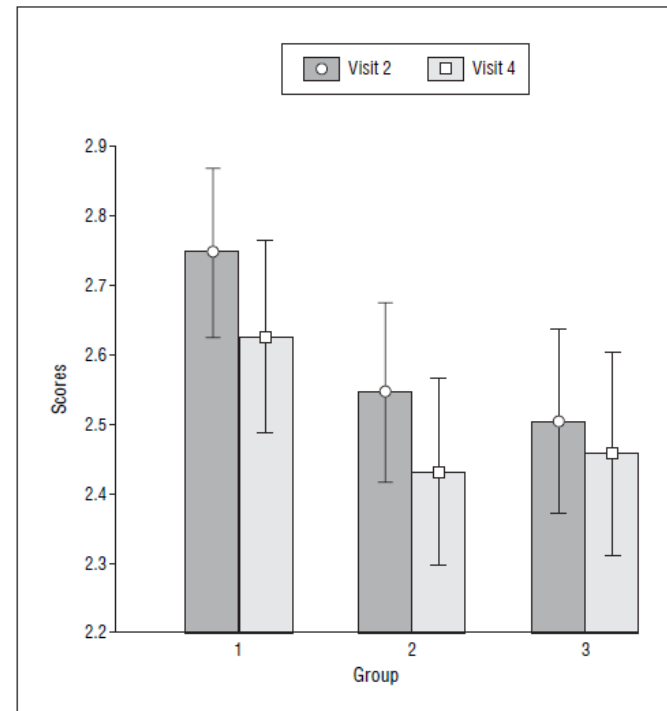
# Efficacy of Isotonic Nasal Wash (Seawater) in the Treatment and Prevention of Rhinitis in Children

Ivo Šlapak, MD; Jana Skoupá, MD; Petr Strnad, MD; Pavel Horník, MD

## Voies aériennes supérieures (VAS)



**Figure 1.** Sensations during application. Patients rated their sensations during isotonic saline wash application using a qualitative scale transformed into a quantitative range for evaluation (1, very pleasant; 2, pleasant; 3, no complaints; 4, unpleasant; and 5, very unpleasant).

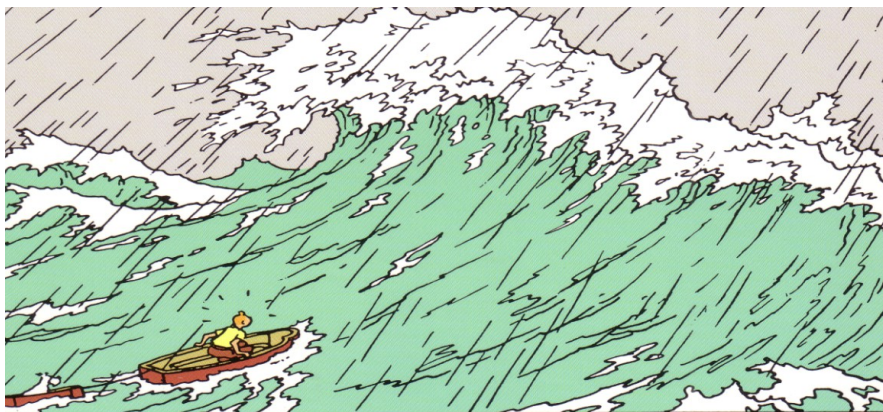


**Figure 2.** Sensations after application. Patients rated their sensations 5 minutes after isotonic saline wash application using a qualitative scale transformed into a quantitative range for evaluation (1, very pleasant; 2, pleasant; 3, no complaints; 4, unpleasant; and 5, very unpleasant).

Quelle technique ?

Poire

Lota



## Voies aériennes inférieures (VAI)

Techniques de désencombrement « classiques » :

Manuelles :

- Drainage autogène
- Eltgol
- AFE
- ...

Instrumentales :

- PEP
- Flutter

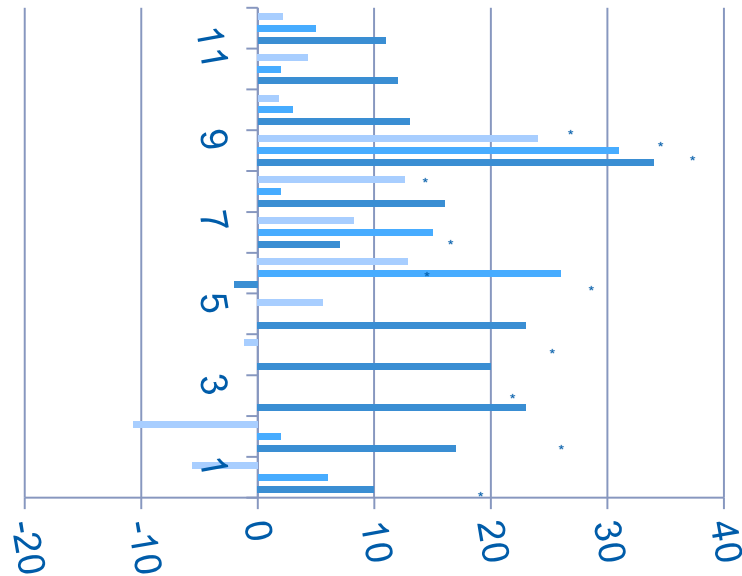


## Exercice physique...

Changes in peak expiratory flow rate (PEFR) and forced expiratory volume expired in 1 seconde (FEV<sub>1</sub>) in response to exercise and to  $\beta_2$ -agonist salbutamol

- % Rise in PEFR with exercise
- % Fall in PEFR with exercise
- % Change in FEV<sub>1</sub> with  $\beta$ -agonist

\* : significant clinical response



Exercice !!!

## Take home message

- Prise en charge dès le diagnostique
- Techniques de kinésithérapie respiratoire :
  - Voies aériennes supérieures
  - Voies aériennes inférieures
- Stimuler l'exercice physique
- Vigilance pendant la période de l'adolescence



A scenic landscape photograph of a mountain range at dawn or dusk. The sun is low on the horizon, creating a warm, golden glow that fills the sky and the valleys. The mountains are layered, with the foreground peaks in sharp silhouette and the distant ranges fading into a thick mist or haze. The trees on the slopes are dark against the lighter background, and the overall atmosphere is serene and ethereal.

Merci pour votre attention !