



# **From Practice - For Practice**

## **Mucoviscidosis**

**by Dr. Konrad Werthmann**



Mucoviscidosis (viscidus = sticky, viscous) is connatal cystic fibrosis, an autosomal-recessive inherited disorder with a defective 7<sup>th</sup> chromosome. It is not inherited by heterozygotic carriers.

A characteristic feature is the generalised dysfunction of the exocrine glands. The secretions have a non-physiological, viscous composition, resulting in both drainage difficulties and in complete obstruction of the excretory ducts of the glands in question. Considerable complications occur within the individual organ systems. In the respiratory tract, this finds expression as breathlessness, in the intestinal tract as maldigestion and malabsorption. As a consequence of the elevated electrolyte content

of the sweat, the patient loses fluids and electrolytes additionally.

The three most important symptom pictures to emerge in the course of such a disease are meconium-ileus in the newborn, pancreatic fibrosis with diarrhoeic, fatty stools, and increasing dystrophy with bronchiectasis and bronchitis which is resistant to treatment.

In the newborn, the symptoms are abdominal enlargement with stinking diarrhoea, prolonged infections of the airways with periods of dyspnoea, intransigent cough (not to be confused with pertussis!), the secretions being purulent, yellow or green.

Confirmation is via pilocarpine

iontophoresis (sweat test): a reading of more than 60 mmol chloride/litre is considered pathological.

Treatment: more fluid than usual, the therapeutic diet should always have enzymes added (e.g. Kreon<sup>®</sup> Granules for Children), mucolytic medication, exercise and physiotherapy, early treatment of the bronchial system (breathing exercises, fresh air); antibiotics, the choice of which varies according to the germ and is almost always necessary.

In the case of this inherited disease, treatment with isotherapy is very difficult. Because of their immediate action, antibiotics afford the patient, and especially his lungs, a rapid improvement in quality of life.

### **Infants and children up to the age of 12:**

- SANUVIS 2X drops, 1 drop once or twice daily in warm water; LEPTOSPERMUSAN drops, one drop once every 2 days; both for the whole duration of treatment.
- NOTAKEHL 5X drops, 1 drop per year of age twice daily for 1-2 weeks; then change to
- SANKOMBI 5X drops 1 drop per year of age twice daily, but always following the pattern 5-2-5-2 (Mon. - Fri. = 5 days SANKOMBI 5X drops, and Sat./Sun. = 2 days NOTAKEHL 5X drops).
- Depending on the microbial presence: SANUKEHL Staph 6X drops, or SANUKEHL Strep 6X drops, 1-4 drops 3 times a week, to be massaged in. (no. of drops depending on age and constitution.)

### **Children over 12 and adults:**

- SANUVIS 2X drops, 5 drops twice a day; LEPTOSPERMUSAN drops, 2-4 drops twice a day; LUFFA-SAN 4X drops, 5 drops once a day; all for the whole duration of treatment.
- FORTAKEHL 5X drops, 2-10 drops twice daily for 1-2 weeks; then change to
- SANKOMBI 5X drops, 2-5 drops twice daily, but always following the pattern 5-2-5-2 (Mon. - Fri. = 5 days SANKOMBI 5X drops, and Sat./Sun. = 2 days FORTAKEHL 5X drops).
- Depending on the microbial presence: SANUKEHL Staph 6X drops, or SANUKEHL Strep 6X drops, 5-10 drops 3 times a week, to be massaged in or taken orally.

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