Редакционное приветствие

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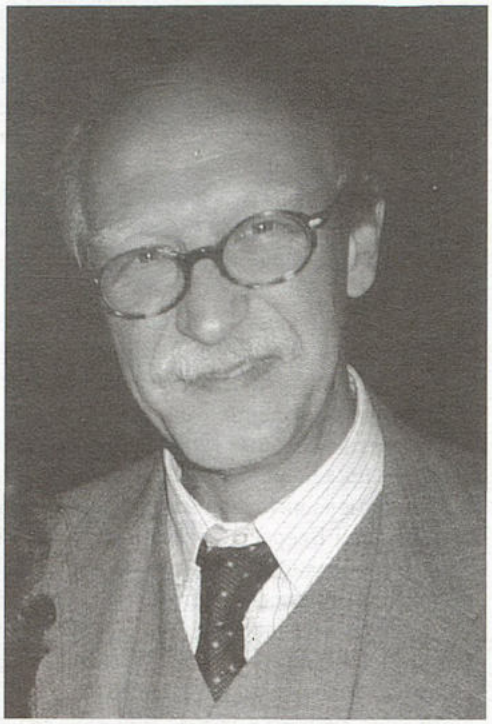

Ladies and Gentlemen,

I think that I don't exaggerate too much when I say that we live in a time of rapid changes. Not only rapid changes but also fundamental changes. Many of these changes (but not necessarily all) are positive and CF is a very illustrative one.

Known for at least 6000 years, CF dreamt the silent dream of unknown adulthood. Patients died soon after birth only recognized by their helpless patients to have salty skins. According to old German folklore a child who tastes salty on being kissed has a poor prognosis.

The 20-th century brought the breakthrough. Described between 1930 and 1940 as a new clinical entity, CF was recognized more and more in Europe and America. Using the sweat test, diagnosis was more and more facilitated, the autosomal recessive inheritance of CF was established in the 1960-s, the localization of the gene to chromosome 7 in 1985, and the gene was cloned four years later.

The genetic revolution resolved the biochemical puzzle in 1989 which had led to many intuitive but false hypotheses, brilliant but completely wrong scientific papers, great hopes but bitter delusions! Thus, CF was finally determined as a chlorid channel mutation defect. This fundamental finding for CF stands like the famous statue of Peter the Great in this city as a solitair. Certainly, it will take some time before we have digested this information and adopted it to reach new horizons for CF treatment.

The application of modern genetic tools has allowed to determine the prevalence of CF in Russia more precisely than before. Unlike in North America or Western Europe where we face a prevalence of 1 CF child in about 2500 births, Russian scientists have determined a prevalence of 1 CF child in about 12 000 life births. Future genetic studies will lead to a clearer picture of the distribution of mutations in the CF gene. The identification of unknown mutations which account for about 25% of CF cases in Russia will also be very valuable for prenatal diagnosis of CF. But first of all we have to diagnose CF. The classical sweat test is available in 50% of 89 geographical regions of Russia. As with many biochemical tests, reference laboratories are needed to ensure high quality standards.

Besides the identification of the CF gene, the year 1989 also saw the opening of the first two officially designated CF centres in Russia, one in Moscow and the second one here in St.Petersburg. Many other centers followed and with its 150 Million inhabitants, Russia has now 26 CF centers. The treatment of this complicated disease has clear advantages for the CF patient in specialized centers, where expert teams and expensive equippment can provide better care than in smaller private practises. Let us hope that such teams including besides the clinician trained physiotherapists, dieticians, specialist nurses, social workers and other experts, will be established in these centers in Russia in the future. Clearly, the physician himself cannot fullfill all these tasks alone in 24 hours! Let us also hope, that the recent political changes which led to the break of the former USSR will not impair the dialogue between CF centers in Russia, the Ukraine, Byelorussia or the Baltic Republics, to name only a few.

We have to admit that the identification of the CF gene had no direct impact on the treatment and the prognosis of CF patients. Rather the fundamental change in nutrition based on pancreatic enzyme supplementation together with the antibiotic revolution in the last three decades has increased life expectancies for CF patients significantly and enabled CF patients to reach adulthood where this treatment was applied. Many efforts had been made in Russia in this direction, and I only want to mention the close cooperation of the Moscow CF center with the CF center of Southampton in England and the Solvay company who have generously supported these efforts. This has already led to a 50% survival rate of CF patients in Moscow and St.Petersburg of 23.3 years.

If antibiotics and pancreatic enzyme supplementation would be available for all CF patients in Russia, a similar life expectancy will result also in this country.

All drugs have their price. The cost of appropriate treatment per patient for one year in Russia is approximately 7000 USD. And this figure may increase with the use of other, more expensive antibiotics to approximately 17 000 USD. It is good news that in Moscow CF patients can receive their drugs free of cost since 1999. However, such help is not offered in every part of the country and strong efforts have to be taken to improve the situation where it needs improvement. But, besides the financial aspect, particularly with antibiotics we have to pay another price. Bacteria get resistant to every antibiotic and sooner or later we have to change our antibiotic strategy.

Education of the various aspects of CF to the young clinicians in other parts of the country is mandatory. National and international symposia or congresses devoted to CF are valuable strategies to distribute and increase the knowledge about CF in distant parts of the country. The 5-th CF Symposium which starts today together with the 10-th National Congress of lung diseases here in St.Petersburg is an optimal occasion to learn from the many experts who have come here all about CF, to make new friends and to plan future scientific cooperations. Research is needed to investigate host-parasite interactions and many other aspects of the multifactorial CF pathogenesis. We have to move, to change, to modify, to create new concepts. Research is in the air and we all sense it with our hearts. Let's start together into this new era!

Dr. Chris Rolles (UK),
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Professor Kapranov and his team are to be congratulated for the wonderful development of the cystic fibrosis service in Moscow over the last 10 years.

When Professor Kapranov started the service he encountered a great many obstacles both political and financial. He had a very loyal team who stayed with him and supported him during those difficult times. At first he needed to find a base in which the cystic fibrosis service could run. Eventually his persistence paid off and he was provided with accommodation for both the staff and the patients at the Republican Children's Hospital. In spite of having very few links with any other centre of expertise the clinic developed and patients from the whole of the country were seen, diagnosed and cared for as well as those from Moscow.

In 1993 a British based charity (International Integrated Health Association IIHA) made contact and was able to initiate a very productive collaborative programme which still continues.

Initially the collaborative programme was based on a "twinning" with the cystic fibrosis centre in Southampton which was of comparative size in terms of patients. It was immediately recognised that cystic fibrosis care in the UK could not be absolutely paralleled in Russia because of the sheer resource implications. For example in the UK at that time the average patient with cystic fibrosis cost the National Health Services about 15 000 USD per year. That type of resource would not be available in Russia and would not even be appropriate in the Russian setting. With the help of a pharmaceutical company (Solvay) a project was set up to see whether a much smaller investment in Moscow could produce results that might at least be comparable. The end point in a 4 year project was to prove that perhaps 90% of the benefit seen in the UK could be obtained in Russia with less than 10% of the resources. This was not only extremely good news in the Russian setting but was also very important information in the UK where it was recognised that a lot of the additional expenditure was probably unnecessary. The results of that collaborative study have now been published. The essence of the modified clinical approach was based on the knowledge that the Moscow team had good training and expertise and that the British team had practical experience in the running of a clinical service.