

CHI and Estonia.

Early Days.

CHI undertook a preliminary visit to Vilnius, Lithuania in December 2005 and a fuller visit in March 2006. Thereafter a steady programme of collaboration with Lithuania has continued. It led to a visit to UK in February 2008 by Professor Sigitas Dumcius and three colleagues, primarily to the RBH, London. This represents the first strand of CHI liaison with the Baltic States of Lithuania, Latvia and Estonia.

In March 2007, I found a message on the European CF Society website from a Dr Vija Svabe in Riga, Latvia, pleading for help with her CF patients and began an E Mail correspondence with her. After establishing some basic facts with her on the CF situation in Latvia, CHI sent Dr Chris Rolles, Medical Adviser to the charity, and Dr Gary Connett, Consultant in paediatric respiratory medicine, Southampton General Hospital to Riga in late February 2008. This visit much consolidated our understanding of Dr Svabe's needs and gave rise to several ideas for collaboration – or, more exactly, assistance to Latvia. This represented a second “Baltic” strand.

Continuing our association with Solvay Pharmaceuticals, Chris Rolles and I went to their Hannover HQ in May 2008 for the day and to talk with Dr Ulrike Tanneberger and her manager. We wished to enlist Solvay's support for the projects in Lithuania and Latvia – and we floated the idea of a combined Baltic States approach. Solvay offered rather lukewarm support to this idea. The con was the awareness that the Baltic states are each proud of their independence. The pro was the idea of a bigger market commercially, not least for Creon or for clinical trials of any CF related drugs. We realised, however, that we knew little about CF in Estonia. Solvay offered to do some research through their Estonian manager in Tallinn.

On return to UK, via the internet, I found a PhD thesis by Dr Tiina Kahre, University of Tartu, Estonia on CF in Estonia, dated 2004. This provided reliable data on the number of CF patients, incidence of CF in Estonia, the CF gene mutations encountered etc as well as Dr Kahre's contact details. Her thesis is at <http://dspace.utlib.ee/dspace/bitstream/10062/577/5/KahrePhD.pdf>

It emerged that Dr Kahre, a geneticist, is the President of the Estonian CF Association and that there are two centres for the CF service in Estonia – one in the capital, Tallinn, and the other in Tartu. CHI now needed to identify a UK CF specialist, who would be willing to undertake an initial exploratory visit to Estonia and hopefully become a mentor to Estonian CF professionals. Thanks to Chris Rolles, such a person was identified at the University of Cardiff, School of Medicine – Dr Iolo Doull.

Pre-visit to Estonia planning.

Dr Urve Putnik, Tallinn Children's Hospital, and Dr Maire Vasar, Tartu University Children's Hospital with Dr Kahre completed the CHI CF questionnaire – providing good information about their patients, their experience, availability of drugs and their needs. The chief point of contact was and is Dr Tiina Kahre. Once Dr Iolo Doull had found a 'slot' in his diary (21st –25th January 2009) for the visit, planning proceeded swiftly. Flights were booked on-line with KLM, such that Iolo, starting from Cardiff,

and I, starting from Heathrow, met in Amsterdam for the last leg of the flight to Tallinn. A hotel in Tallinn was also booked on-line for the 4 nights in-country. The programme for the visit is at Appendix 1. We needed to spend some time in both CF centres, Tallinn and Tartu – and a meeting with the families – members of the Estonian CF Association – was sensibly convened in Tallinn. This meant a long first day with travel to Tartu and back to Tallinn but meant we stayed in only one hotel.

Report of the visit.

22nd January

The rail system is being modernised and we were able to catch a new non-stop service from Tallinn to Tartu (a journey of 185km) at 0746. On arrival at Tartu we were met by Dr Kahre and driven to the University Hospital Children's wing.

We met Dr Maire Vasar, lead CF doctor in Tartu and several of her colleagues, including Dr Kaja Julge, allergy specialist and Karin Tammik, physiotherapist, as well as Drs Putnik and Karin Puks from Tallinn. After introductions – all in English and without the need for an interpreter – Dr Doull asked questions to establish the local CF situation.

Some basic facts about the CF service in Estonia.

With Dr Urve Putnik leading, there are 12 registered patients in Tallinn, of whom the oldest is 17. Dr Putnik has had responsibility for CF patients for 5 years. In Tartu (Dr Vasar), there are 20. She has had responsibility for CF for 4 years. Incidence of CF is much lower than in UK with about 1:7500 births, ie about two new cases per year. There is no new-born screening in Estonia. Average age of diagnosis is around 2 years and reducing. Apart from the two CF centres, there are 10 other district hospitals, where children might go for treatment or advice. On average the CF Drs in Tallinn and Tartu meet once a year but may discuss difficult cases by telephone. The CF team in Tartu has the services of a physio (Karin) but she has limited knowledge of CF specific physio (– hence the request for a videocassette/ DVD of physio for CF children – obtained from the UK Cf Trust and handed over), whilst the Tallinn “CF” physio is away on maternity leave until the Autumn. Both teams have psychological/ psychiatric/ psychotherapeutic support but no dietary expertise. DNase is available free in Estonia.

Regarding medication and enzymes (the rules and regulations were confusing), all medicines for in-patients are free. Generally, only medicines registered in Estonia may be prescribed but there are exceptions involving applications to the “Sick fund”. There are constraints on the availability of antibiotics – and their use for prophylactic purposes seemed limited by UK standards. Dobromycin is registered but gentomycin is not. Vitamin supplements are not funded or used and patients/families buy their own high calories drinks. Creon is the main enzyme in use (the other is pangrol). The doctors thought this was supplied free but parents later indicated that they do have to pay – between EEK 100 and 200 per month (£6-12 per month). Where medicines are prescribed for CF children as out-patients, a family pays EEK 10 per prescription for a child under 4 and 10% of the cost for a child over 4.

CF in Wales.

Dr Doull then described his work in Wales under the title “CF in the 21st Century”. This covered experience with 250 CF children both in Cardiff and further afield, such as in Aberystwyth, life expectancy, treatment regimes etc etc. There was a useful discussion of Sweat test interpretation and a recommendation that the minimum weight of sweat for a reliable result is 75mg and should be more than 100mg. Thanks to new-born screening in Wales, 97% of CF cases are detected at ‘birth’. Graphs illustrated the serious impact on a child’s life expectancy if it has *Pseudomonas aeruginosa* and was malnourished. His advice included the use of hypertonic saline as one way to reduce intake of antibiotics. A good routine, morning and evening, is to give Salbutamol, 4ml of 7% hypertonic saline solution, followed by physio.

For a boy with Meconium ileus and CF liver disease in Wales, ‘urso’ (ursodeoxycholic acid) is prescribed from birth and he remains on it for life. In Estonia, if liver function reverts to normal, ‘urso’ is discontinued.

His presentation is available on request from CHI’s Coordinator and a copy is held by our Estonian colleagues.

The afternoon in Tartu.

We paid a courtesy call on the Head of the Children’s Unit, Professor Valle Tillmann. He has spent time in both Manchester and Sheffield hospitals.

From 1400, Dr Doull gave his presentation “CF in the 21st Century” to a much wider audience of the Hospital’s doctors (incidentally, almost all female, about 60 in all) and answered questions.

A joint consultation then took place of a young man with CF and aged 24.

We adjourned then to an “Irish” pub in Tartu and had an evening meal together – hosted jointly by Iolo and CHI..

On a wintry night and with the road in a slushy, wet state, Dr Urve Putnik drove us the 185 km back to Tallinn. We reached the hotel around 2115.

23rd January.

We arrived at the Tallinn Children’s Hospital about 0800. We met the local Solvay manager, Dr Terje Nirgi; we had a short chat and she stayed for the first part of the morning.

From 0815 to 0900, Dr Doull gave his presentation “CF in the Twenty First Century” to most of the Doctors at the hospital (an audience of about 60), answered some questions and I gave a short description of CHI.

The planned meeting with the Head of the Hospital did not take place.

We were given a tour of the hospital. It was modernised three years ago. It is very impressive – fresh décor, bright, very clean, quiet, modern medical equipment and extensive modern IT facilities. We noted a cheerful atmosphere and a good morale amongst the staff.

Dr Urve Putnik introduced 3 short presentations on aspects of some of her CF cases. The first was by a psychotherapist and concerned a girl aged 10. The second presentation was by a geneticist, Kairit Joost, on strategies for molecular diagnostics – and how to offer advice on the results of genetic analysis to parents. If a pre-natal test indicates a CF positive child, the general Estonian ethos is to terminate the pregnancy but a few families have ruled out termination on religious grounds.

Sweat testing was discussed. In 2008, Tallinn had undertaken 37 sweat tests and Tartu – 100. Dr Doull mentioned the existence on-line of comprehensive guidance on sweat testing <http://www.acb.org.uk/docs/sweat.pdf>

The third presentation was by the physiotherapy instructor and concerned a problematical case of a boy of 12.

A session followed in which many CF cases in Estonia were reviewed together. This led to a good exchange of experience and practice and these points are recorded: -

- Both Dr Doull and Dr Putnik carry out annual reviews – Iolo agreed to send an example of his annual report to Urve and a copy of his guidelines for annual review.
- Dr Doull recommended exclusion of the IDG test (EKG in Estonian).
- Dr Doull passed copies of two documents – on exercise and on hypertonic saline solution to Dr Putnik.
- Advocacy of treating phenotype rather than genotype.
- Dr Doull recommends wide useage of azithromycin. Many of his children are prescribed a combination of azithromycin and flucloxacilin.
- A joint agreement that “Guidelines for CF Care in Estonia” should be drawn up. Dr Doull recommended that these should be aspirational but realistic.
- Is it a role for CHI to intervene with Estonian health policy makers/ resource allocators regarding the availability and cost of medication for CF children/ adults?

Dr Tiina Kahre gave a presentation both about her 2004 thesis on CF in Estonia, updating some of the data to 2008 and about the Estonian CF Foundation. She presented copies of her thesis to Dr Doull and to me. The median age of those with CF has now reached 15 ½ years and, whilst there were no patients with CF over the age of 18 in 2003, there are now 9.

The Estonian CF Association has been in existence for about 15 years. It has enjoyed good relations with a similar body in Denmark. Families pay a membership fee. An important annual event is a summer camp. This has rules to prevent cross infection and camp activities include positive thinking, and musical and laughing therapy! There is an ECFA (rather dormant-looking) website at www.etfy.ee - apparently sponsored by Roche.

In the evening, Dr Doull and I accompanied Dr Tiina Kahre to a Burns Dinner (250th anniversary of Robbie Burns' birth) at the Black Heads House in Pikk street, Tallinn. This was a genuine Scottish celebration, organised by the British Estonian Chamber of Commerce (BECC) with pipers and haggis but it was also a fundraising dinner with the Estonian CF association as one of the three main beneficiaries. It turned out that the Deputy Chairman of BECC (Phil Marsdale) has a 10 year old niece living in Kent with CF – and it was his idea to support the Estonian CF Association.

24th January

At 1100 at the Tallinn Children's Hospital, we attended a meeting of the Estonian CF Association – which included most of the key CF professionals from Tallinn and Tartu. We noted with gratitude the way in which Drs in Tartu had come to Tallinn to meet us and Drs from Tallinn had travelled similarly to Tartu. Altogether we numbered about 30, with a sprinkling of CF children and some 16 CF family members.

I began proceedings with a presentation about CHI (available from the CHI Coordinator) and explaining in some detail how our projects had developed in Lithuania and Latvia – and what might be ongoing work in these countries in order to stimulate thinking about future collaboration with Estonia and to test feelings about an integrated Baltic States programme. I sensed no particular enthusiasm for the latter.

Dr Iolo Doull gave his presentation – by now familiar to the CF Drs but new to the families – and answered questions.

After the families had left, we convened to discuss outcomes of the visit and to discuss “where now?” Iolo and I thanked our hosts for their hospitality. Much of what we had seen was excellent. These were our conclusions – and actions already agreed are repeated for the sake of convenience - and form the basis for ongoing collaboration: -

Conclusions

1. “Guidelines for the care of children with CF” should be drawn up locally.
2. The CF teams in Tallinn and Tartu might benefit from more regular meeting, particularly to review the more problematical patients.
3. The same “problematical” patients can be referred to Dr Doull in Cardiff, using current technology and at a time to be mutually agreed. (Photos were taken of all doctors involved so that telephone calls can be linked to a “face”!). This arrangement should be trialled at an early date.
4. Dr Doull will send a copy of his annual CF review report and guidelines for the review to Dr Putnik.
5. To strengthen the CF physiotherapy capability, CHI will try to send a UK physiotherapist to Estonia for 1-2 weeks. She would give a course to 4 Estonian physios – 2 from Tallinn and 2 from Tartu, including Karin Tammik – and give a lecture at Tartu University.
6. We considered that a visit to Cardiff of an Estonian team would be helpful, not least to see the UK's team approach to CF. Such a team should be equally drawn from Tallinn and Tartu. Some thought would be required to find a

suitable “dietician representative” – a candidate in Tartu has Estonian, Finnish and German as her languages. The Estonian side is welcome to propose such a visit.

7. More work would be required to justify new-born screening in Estonia.
8. We felt that there should be better availability of medication, for example vitamin supplements, some antibiotics and at lower cost to families. Further that the number of families involved is small and therefore the cost not great. But we didn’t decide if this is an action locally or for CHI involvement.

25th January

We flew back to UK.

8th February

Tony Wolstenholme