Learning How Patients with Heritable Skin Diseases are Cared For in the UK: Centralization Concentrates Expertise

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A group of four dermatologists who treat heritable skin diseases in Finland visited two dermatology centres in London, UK. The visitors witnessed the effectiveness of centralization, which has created expertise, the power of multidisciplinary teamwork and the presence of research as leading factors in diagnosis.

Patients with rare genodermatoses, specifically epidermolysis bullosa (EB), are treated in two main locations in London. The children attend Great Ormond Street Hospital for Children National Health Service (NHS) Trust (GOSH), and adults are treated at St Thomas' Hospital at Westminster Bridge Road. GOSH is the oldest children's hospital in London; it opened in 1852. Together with University College London (UCL) Institute of Child Health, this NHS trust is the largest centre for research into childhood illness outside the USA. St John's Institute of Dermatology opened in 1863, and is nowadays integrated into Guy's and St Thomas' NHS Foundation Trust and King's and St Thomas' School of Medicine (GKT) of King's College London. St John's Institute of Dermatology is one of the world's leading research centres for skin diseases.

The Finnish Dermatological Society has a section for heritable skin diseases, comprising the doctors responsible for this patient group in the university hospitals. This group of doctors perceived a need for collaboration and deepening of their practical experience, especially with regard to EB. Therefore, Dr Kaisa Tasanen from Oulu organized a visit to GOSH and St Thomas' Hospitals for a group of four Finnish doctors: herself, Maja-Leena Tuomi from Tampere, Hannele Heikkilä (Fig. 1) from Helsinki, and Sirkku Peltonen from Turku. The group had the opportunity to observe the work of Dr Jemima Mellerio (Fig. 2) and her colleagues for 3 days in the two hospitals. Without the kind help of Dr Mellerio this visit would not have been possible. In order to obtain visitor status for GOSH a multi-page form, including signatures from the home hospital, had to be completed weeks before the trip (in addition to a good deal of e-mail correspondence), and the visitors had to attend the administration and security offices and to wear official visitor identity badges. In addition, visitors usually have to pay a fee of £150. The paperwork needed for St Thomas' Hospital was much less extensive.

The reward for completing this paperwork was the opportunity for the visitors to observe patients with rare diseases that none

Fig. 1. Visitors from Finland (*left to right*): Hannele Heikkilä, Marja-Leena Tuomi and Kaisa Tasanen-Määttä on their way to St Thomas' Hospital (the building on the right), located near the River Thames opposite the Houses of Parliament.

Fig. 2. Dr Jemima Mellerio (*left*) with secretary Sonia Ama. Jemima Mellerio is a dermatologist with special expertise in epidermolysis bullosa. Sonia Ama plays a key role in organizing and coordinating the visits of children with epidermolysis bullosa to different specialists and services at GOSH.

of them had seen before, and to observe an expert team working together for patients with EB. In the UK the diagnostics and responsibility for treatment of patients with EB is centred in two cities: London and Birmingham. Although these areas are densely populated, patients may need to travel from some distance to the centres. The London clinics are responsible for more than 300 children and approximately 250 adults with EB. Approximately 50 of the patients have severe EB, and approximately 16 babies are born each year with severe EB. This number of patients has created a lot of expertise on treatments and everyday living with EB. Centralization has also enabled research to be carried out, which has led to the discovery of new disease entities, such as desmosomal genodermatoses. This strong research tradition surrounds the clinical work, and obtaining a molecular diagnosis for each patient is a permanent goal. Many diagnoses can, in fact, be obtained from the laboratory located at St John's Institute of Dermatology, led by Professor John McGrath (Fig. 3).

Fig. 3. Professor John McGrath running his clinic for heritable skin diseases.

The EB team includes at least a dermatologist, a paediatrician, EB nurses, dieticians, a social worker, physiotherapists and a psychologist. It has become evident that patients with severe EB need a holistic approach in order to thrive. Consultations about surgery are often needed, for example for oesophageal strictures and skin cancers. Trained EB nurses also visit local hospitals as soon as the centre is informed of the birth of a baby with blistering skin disease. The nurses carry an "EB pack", which includes, for example, a small mattress to help with moving and carrying the baby without causing stress to the skin, a special feeder (Haberman Feeder™, Medela Special Needs Feeder, Medela AG Baar, Switzerland), which allows the milk to flow without strong sucking, and various bandages. The EB nurses also take skin biopsies for diagnostics. They make home visits to children with EB and help solve difficulties with practical everyday issues, such as skin treatments, bathing, feeding, housing and schooling. The EB team has also produced a range of practical information leaflets on various topics, such as infant nutrition, eye care, physiotherapy, school, etc.

Diagnosing and treating rare heritable skin diseases is always challenging, even more so if one only sees patients with these diseases once or twice during one's career. Most of the Nordic countries have large areas with small populations, and thus are not ideal for building special expertise with rare diseases. Centralizing the treatment of rare diseases, the formation of teams with different disciplines, and networking of those teams should be priorities, and should be promoted by the university hospitals to guarantee high-quality healthcare for patients.

The visitors are grateful to Dr Jemima Mellerio and Professor John McGrath for providing the opportunity to visit their clinics, and to all the other doctors and personnel whose work they observed.