Report of audit visit to the

Belfast Comprehensive Care Haemophilia Centre

1 - 2 June 2000

Audit visit carried out under the auspices of the
UK Haemophilia Centre Directors' Organisation by:

Dr M Makris - Director
Sheffield Comprehensive Care Haemophilia Centre

Mr K Lawrence - Haemophilia Nurse Specialist
Cambridge Comprehensive Care Haemophilia Centre

1.0 Audit Format.

The format of the review was according to the proforma supplied by the UK Haemophilia Centre Directors Organisation. It consisted of a two day site visit and review of questionnaires sent randomly to patients or parents who use the services of the Centre. On the first day of the audit, the services of the Paediatric Centre at the Royal Belfast Hospital for Sick Children were reviewed, whilst on the second day the adult services, which are currently based at the Royal Victoria Hospital, Belfast were audited.

2.0 Background.

The Belfast Comprehensive Care Haemophilia Centre is responsible for the diagnosis and management of all patients with hereditary haemostatic disorders in Northern Ireland, a province with a population of 1.5million, of which approximately one third live in Belfast. The Centre is based in Belfast, at the Royal Hospitals site. Children under the age of 16 are seen at the Royal Belfast Hospital for Sick Children and this service is supervised by Dr S Dempsey. The adult services are currently based at the Royal Victoria Hospital and, until May 1999, were supervised by Dr E Mayne. Since Dr Mayne's retirement there has been a vacancy for the replacement post, and the adult service has been supervised by Dr F Jones, a Consultant Haematologist at the Royal Victoria Hospital. Clinical haematology, including the adult haemophilia service, is due to relocate at the City Hospital site (about 1½ miles from the Royal Hospitals site) in Spring 2001. The plans of exactly where the haemophilia centre will be, how much space will be available or the space provision for the coagulation laboratory have not yet been finalised.

3.0 PAEDIATRIC SERVICES: ROYAL BELFAST HOSPITAL FOR SICK CHILDREN.

All children with haemostatic disorders are seen at the Children's Hospital. They get transferred to the adult service usually shortly after their 16th birthday.

3.1 Patients registered.

Currently the paediatric unit looks after 75 patients with congenital bleeding disorders, and the distribution is:

Diagnosis	Total patients	Severe patients
Haemophilia A	47	24
Haemophilia A carrier	1	0
Haemophilia B	4	4
Factor X deficiency	1	0
Von Willebrand's disease	15	0
Platelet abnormalities	7	0

3.2 Haemophilia medical staff.

The Director of the paediatric service is Dr S Dempsey. He is supported by Dr Cairns, a Clinical Medical Officer, Dr McCarthey, a Consultant Paediatric Oncologist, as well as a full time Paediatric Registrar and a Paediatric SHO. At the time of the visit there was no Haematology SpR rotating through the unit.

3.3 Haemophilia nursing staff.

There is no dedicated Haemophilia Nurse Specialist. The Ward Sister provides in-patient support for the haemophiliacs. The in-patient haematology ward is staffed by 14 nurses, none of which are specifically dedicated to haemophilia care. 3 of these nurses, however, are registered on the 'Essentials of Haemophilia Nursing' course which is due to run at Canterbury in Autumn 2000.

3.4 Physiotherapy services.

There is no dedicated haemophilia physiotherapy service. There is, however, a well-appointed physiotherapy department in close proximity to the in-patient haematology unit which includes a hydrotherapy pool. Physiotherapy services are provided to haemophiliacs when specifically needed and requested from the hospital pool of physiotherapists. There is no regular review of haemophiliacs by physiotherapy specialists.

3.5 Dental care.

There is an excellent dental service provided by Mr Martin Kinirons. There are two paediatric-sized dental chairs in a specialised room, in very close proximity to the area where haemophiliacs have their out-patient reviews. As a result of this, all haemophilic children have a dental review every 3-4 months.

3.6 HIV / Hepatitis / orthopaedic input.

None of the children currently attending the Children's Hospital are HIV, or hepatitis B or C, positive. When required, orthopaedic support is provided by Mr Harry Cowie, a Paediatric Orthopaedic Surgeon.

3.7 Social work support.

This is currently provided by a Malcolm Sargent Social Worker who is available for one session per week. This is funded by a charitable trust.

3.8 In-patient facilities.

There is no dedicated Haemophilia Centre, but the in-patient haematology ward is a hub of activity and haemophilic children can be reviewed there at any time. The haematology unit is near the main entrance to the hospital, and this is well signposted and can be accessed via the Allen Ward. There is currently a considerable amount of building work taking place on the hospital site and parking may be a problem, although patients can be dropped near the entrance. There is a hospital bus system travelling from the city centre to the hospital. Disabled parking spaces are available. There is a direct phone line to the ward and out-of-hours telephone calls are taken by the in-patient haematology unit.

The in-patient unit is eight-bedded. There are two refrigerators containing factor VIII and IX concentrates respectively.

3.9 Out-patient facilities.

Routine out-patient clinics are held in the out-patient department, which is in close proximity to the in-patient facility. There are haemophilia clinics on the first and third Friday of every month.

3.10 Emergency treatment.

Patients are able to obtain emergency treatment by telephoning the haematology ward and attending. Out-of-hours the patients will be seen by the Senior House Officer on call. There are excellent printed guidelines about what to do when a patient attends. Doctors are advised to seek senior help for every new bleed, and they are given advice on what treatment to give and where to find the concentrate, as well as how to record it.

3.11 Medical records.

The patients' medical notes are housed within the Haematology Department. The quality of the notes is excellent, both in content as well as in presentation. The patients' factor deficiency is written at the front of each cover. The laboratory results are colour-coded and are very easy to find. There was plenty of evidence of letters to GPs and other services. Family pedigrees, however, were not evident in any of the notes reviewed.

3.12 Clotting factor usage and administration.

All patients with haemophilia are on recombinant factor VIII or IX concentrates. The product usage is compatible with the recommendations from the UKHCDO guidelines. The Centre uses approximately 2.5 million units of factor annually.

Three of the patients have portacaths in situ. Twenty-two of the patients are on prophylaxis with FVIII/IX concentrate, the vast majority of which is given at home by the parents. There are, however, Four patients who receive their prophylactic treatment from the ward.

3.13 Laboratory support.

All the laboratory support is provided by the adult service which is based at the Royal Victoria Hospital. At present the paediatric service receives very good laboratory support and this is undoubtedly helped by the close proximity of the two hospitals. There is a potential risk to this service when the adult haematology and haemophilia services are re-sited at the City Hospital site.

3.14 Molecular genetic support.

Other than explaining the genetics of haemophilia, all other testing is provided by the adult site.

3.15 Patient satisfaction questionnaire.

Prior to the audit visit, an anonymised list of all the severe and moderate patients attending was forwarded to Dr Makris. Ten patients were selected at random, and the questionnaires were sent to their parents. Of these ten, four (40%) were returned to the auditors.

Although only four questionnaires were returned, there was universal satisfaction and praise for the service. Each questionnaire consisted of ten questions, and of the total of forty replies there was not a single negative comment.

4.0 Recommendations for the Royal Belfast Hospital for Sick Children.

Although the overall standard of paediatric haemophilia care has been maintained since previous audits, it is disappointing that some of the previous recommendations (see section 7.0) have not been acted upon. The Centre must be unique in the UK in not having a dedicated Haemophilia Nurse Specialist. At present, a lot of the workload that would be carried out by a Haemophilia Nurse Specialist ends up being provided by the Consultant Haematologist and the Clinical Medical Officer. In the view of the auditors, this is unacceptable because these two persons already have very heavy workloads and are unable to provide many of the services provided by paediatric haemophilia nurses such as school visits, home visits, primary care visits, etc. The auditors would therefore recommend:

a) By far the most important, urgent need for the Centre is the appointment of a dedicated Haemophilia Nurse Specialist.

b) As recommended at the last audit, physiotherapy should be integrated into the children's clinic.

c) A specific physiotherapist should be named for provision of haemophilia care.

d) Increased dedicated social work support should be provided.

e) Although current laboratory support is adequate, there should be specific points made in the contract, following the transfer of the laboratories to the City Hospital, for provision of all the tests required by the Haemophilia Centre. As well as the usual routine tests, there should be 24-hour support for factor VIII and IX estimation, as well as for inhibitor detection.

f) There should be increased liaison between the paediatric and adult services to provide more cohesive haemophilia care, especially since Belfast is a single Comprehensive Care Centre.

5.0 ADULT SERVICES: ROYAL VICTORIA HOSPITAL.

5.1 Patients registered.

Diagnosis	Total patients	Severe patients
Haemophilia A	106	40
Haemophilia A carrier	2	0
Haemophilia B	7	4
Haemphilia B carrier	1	0
Von Willebrand's disease	85	1
Platelet abnormalities	30	?

5.2 Haemophilia medical staff.

The Centre Director, Dr E Mayne, retired in May 1999 and has not been replaced. The post was previously advertised, but only one candidate - who was not considered suitable - attended for interview. Following this, senior staff at the hospital asked around the UK and tried to see if anybody in training would be suitable. As a result of this there was a deliberate delay in advertising the post. The post has now been re-advertised and interviews will be held at the end of June 2000. Over the last 12 months, continuity of haemophilia care has been provided by Dr Orla McNulty with support from the two haematology consultants, Dr F Jones and Dr M F McMullin. The other medical staff providing help are the Haematology SpRs and SHOs.

5.3 Haemophilia nursing staff.

In February 2000, Sister Collette McAffee was appointed as the first Haemophilia Nurse Specialist. She works full-time and has the support of the other haematology nurses working in the haematology day unit.

5.4 Physiotherapy services.

There is a haemophilia specialist physiotherapist working on two days a week. She primarily provides out-patient physiotherapy and is always available for the out-patient haemophilia clinics.

5.5 Dental care.

The dental care provided for adult haemophiliacs was severely criticised in the previous audit. The reason for this was felt to be the care provided by a specific dental surgeon, who has now retired. Unfortunately no replacement appointment has been made. There is no dedicated dental service for adult haemophiliacs in Northern Ireland at present. This is disappointing, considering the fact that the University Dental School is situated on the site of the Royal Hospitals.

5.6 HIV / hepatitis / orthopaedic input.

There are currently only three HIV positive haemophiliacs, and these are managed in conjunction with the HIV specialists in the hospital. It was noted that the HIV viral load results are kept separately from the main medical notes of the patients for 'confidentiality' reasons. The auditors believe that improved patient care will result by including all the HIV viral loads in the patients' notes.

Most adult haemophiliacs are infected with the hepatitis C virus. There is, at present, only very limited input from the hepatologists. The UKHCDO guidelines on the management of liver disease in haemophiliacs recommend that patients are managed in conjunction with a hepatologist, and there should certainly be very much more hepatology input in the management of these patients. The combination of interferon/ ribavirin treatment is not available in the Centre for financial reasons, in common with the rest of the UK. A number of patients have been previously treated with the combination as part of a UK trial.

Orthopaedic surgery is carried out by Mr McClelland at the Royal Victoria Hospital. Mr McClelland has considerable experience in haemophilic orthopaedic surgery. Post-surgery patients are initially on the Orthopaedic Ward for 48 hours and are then transferred to the Haematology Ward for the rest of their care. There are infrequent combined haemophilia/orthopaedic clinics. Because there will be no transfer of orthopaedic services after transfer of haematology to the City Hospital, the question of where orthopaedic surgery is performed in future should be discussed before the move takes place. Overall, so far, orthopaedic input has been adequate, with the exception that since Dr Mayne's retirement no major orthopaedic surgery has taken place.

5.7 **Social work support.**
This is provided by Finulla Ford, a Social Worker working 18 hours per week.

5.8 **In-patient facilities.**
Haemophilia in-patients are currently managed on the Haematology Ward. In common with other Centres, haemophiliacs are rarely admitted these days and there are no problems getting them onto the ward. When they start, the nursing staff on the ward get an induction, which includes some haemophilia training, and there are very good medical and nursing protocols available on the ward for the management of haemophiliacs.

5.9 **Out-patient facilities.**
The Haemophilia Centre is currently housed in a portacabin and shares the facilities with the Haematology Day Ward. The Centre transferred to this purpose-build accommodation about a year ago. The term 'portacabin', however, fails to convey the standard of this accommodation, which is good. There are three private single-bedded rooms as well as an open ward area for the management of patients. There are separate reception and secretarial rooms and all the haemophilia notes are kept within the Centre. There are toilets, including disabled ones, on site. Patients can be dropped just outside the door and there is a small number of parking spaces. Despite the temporary nature of the accommodation, the facilities are adequate, and probably superior to many provided by other Comprehensive Care Centres in the UK.

5.10 **Emergency treatment.**
Patients are able to turn up at the Haemophilia Centre for treatment during the day on weekdays. Out of hours, they telephone the Haematology Ward and attend to receive treatment. There are adequate protocols in place and patients are discussed with, or seen by, a Haematology Registrar before treatment is given.

5.11 Medical records.

The quality of medical records is good, filing was neat and dividers were present. There were family trees in all the sets of notes that were picked up at random. The auditors were impressed by one set of notes that was the 21st set for a single patient. The medical records are kept in the Haemophilia Centre and are easily accessible both during working hours as well as out-of-hours if a patient attends the ward.

5.12 Clotting factor usage and administration.

Most patients are managed with high-purity plasma-derived products. All product usage complies with the UKHCDO guidelines. An increasing number of patients are started on recombinant products. There are no problems with supplies.

When patients require products for home use they telephone the Haemophilia Centre and order a supply. For patients who live near the Centre, the factor is prepared by the Blood Bank and delivered to the Haemophilia Centre from where patients collect it. For patients living further away, the factor is delivered to a nearby hospital. The system appears to work very well.

5.13 Laboratory support.

The dedicated specialist haemostasis section is housed in quite a small space for the range of tests offered. All coagulation factors, as well as inhibitor quantitation, are available. Bleeding times are performed by nursing staff. Von Willebrand factor activity and antigen levels are performed locally, but the von Willebrand factor multimers are sent to the Royal Free Hospital in London. Platelet aggregometry can be performed locally, but there are no tests available for 5HT uptake and release or for platelet nucleotides. The laboratory provides all the haemophilia support for the paediatric service as well. Factor VIII and IX levels, as well as inhibitors, can be performed on-call 24-hours a day.

5.14 Molecular genetic support.

There is a molecular genetics laboratory with a service provided by a single scientist with ten years post-doc experience. He performs the routine factor V Leiden, prothrombin 20210A and MTHFR screening, which take about two thirds of his time. In the remainder he performs some molecular genetics to identify carriers of haemophilia. A significant part of this Post-doc's time is, in the eyes of the auditors, inappropriately used. The auditors would recommend that the indications for thrombophilic screening are examined since the rate of positive results is very low. The thrombophilic tests could be considered 'routine' and could be performed by a junior MLSO under the supervision of the Molecular Biologist. The current haemophilia/genetic service could be significantly improved, and this should easily be within the capabilities of the current employee, provided he was given adequate time and support.

5.15 Patient satisfaction questionnaire.

An anonymised list of all the severe and moderate haemophilia patients attending the adult site was sent to the auditors before the audit visit. Fifteen patients were selected at random to receive questionnaires. Of these eleven (73%) returned their questionnaires to Dr Makris.

Of the eleven returned questionnaires, six felt all aspects of care were excellent and were very pleased with the service with no adverse comments or suggestions for any changes. Four other patients felt the overall service was very good but had some comments. Two mentioned that they did not like the portacabin and would prefer the Haemophilia Centre to stay at the Royal Victoria Hospital, respectively. Two others mentioned that sometimes the clinic had to be reminded to send the patients' appointment and a further patient felt that sometimes chronic joint problems were allowed to linger on (but felt that the problem was outside the Haemophilia Unit).

There was only a single patient questionnaire that was critical in a number of parts. This patient felt that orthopaedic follow-up was irregular, there was no input from hepatology, he was irregularly seen by a consultant, and that dental support was poor due to the lack of replacement of the dental surgeon. The questionnaire asked for 'what improvements in the care of haemophilia would you like to see at your centre?' and the patient's suggestions in terms of orthopaedic, hepatology, Centre Director and dental support are in line with the recommendations of the auditors that are given at the end of this report. This patient felt that "the lack of adequate bridging arrangements between the retirement of Centre Director and appointment of a replacement has been generally detrimental to good haemophilia care".

6.0 Recommendations for the adult services at the Royal Victoria Hospital.

There have clearly been some difficulties over the last year due to the failure to find a successor for Dr Elizabeth Mayne's post. There will be interviews in the very near future, and hopefully a successful candidate will be appointed. The auditors would recommend:

a) Haemophilia Centre Director.

A new Haemophilia Centre Director for the adult site should be appointed as soon as possible. In the event of not appointing a Director, a Locum appointment should be made, if possible, to help with the running of the service. Although over the last year major elective and orthopaedic surgery has not been performed due to the lack of a Haemophilia Centre Director, this cannot be allowed to continue. Two possible options are to transfer patients for major surgery to another Comprehensive Care Haemophilia Centre, or alternatively perform the surgery locally following discussions with the Director of another Comprehensive Care Centre. The auditors feel that it is reasonable and acceptable to perform major surgery in Belfast even in the absence of a Haemophilia Centre Director, provided that the patients do not have inhibitors, that Dr McNulty and Sister McAffee are both available, that one of the local Haematology Consultants is available and also provided there is a clear management plan for the patient and this has been discussed with another Comprensive Care Centre Director.

b) The new haemophilia centre.

The auditors did not see the plans for the new haemophilia centre. It is important that adequate space and facilities are provided in a dedicated space for the haemophilia centre. There should be adequate parking for patients with mobility problems due to haemophilic arthropathy, as well as adequate access for these patients within the new haemophilia centre itself.

c) **New laboratory services.**
With the relocation of the Haemophilia Centre at the City Hospital, it is important that all the currently available routine coagulation tests should be available at the City site. Furthermore it is imperative that factor VIII and IX assays, as well as inhibitor detection, should be offered on a 24-hour basis at the new Centre.

d) **Hepatology input.**
There is currently inadequate input from hepatology for the management of HCV positive haemophiliacs. Since most adults with haemophilia are HCV positive, it is important that their management is guided by a hepatologist and **all** patients should be reviewed or discussed with that person. One way of achieving this is to have combined hepatology/haemophilia clinics.

e) **Genetic services.**
Currently a single person provides these services. The auditors recommend that a laboratory technician should be identified to work with the scientist. The indications for factor V Leiden, prothrombin 20210A and MTHFR testing should be reviewed.

f) **Dental services.**
A dedicated dental surgeon should be identified, who can take responsibility for the provision of dental support for haemophiliacs at the Belfast Centre

7.0 Closure of the loop: action based on previous audits.

There have been two previous full audits, in 1994 the Centre was audited by Dr Gordon Lowe and in 1996 it was audited by Dr Ian Hann. A number of specific recommendations have been previously made:

a)	Improvement of adult patient notes with proper dividers	Achieved
b)	Place family trees in all case notes	Achieved for the adult site but not for the children's site.
c)	Urgently investigate and improve adult dental care	There was a major problem relating to a specific dental surgeon, who has now retired. No replacement has been appointed, therefore action point not achieved.
d)	Continue IT improvements to allow free data flow.	A new computer system has recently been installed.
e)	Improved parking for the future.	The relocation of the Haemophilia Centre on the adult site has resulted in improvements for this.
f)	Integrate physiotherapy into children's clinics.	Not achieved.
g)	Consider more community care especially for children, eg school visits/ home deliveries/primary care visits.	Not achieved.

GRO-C

M Makris
Senior Lecturer in Haematology

GRO-C

K Lawrence
Specialist Haemophilia Nurse

1 July 2000