

## **How Elderly People with Bleeding Disorders Manage Their Condition**

Chair: June Ward, Haemophilia Specialist Nurse, Ninewells Hospital, Dundee, United Kingdom

### *Impact of Hemophilia on Everyday Life of Elderly Patients*

Sylvia von Mackensen, Institute of Medical Psychology, University Hospital Hamburg, Germany

Since treatment advances continue to improve life expectancy for people with hemophilia, different approaches, special care, and dedication are needed to meet the needs of the growing number of elderly patients, said Dr. Sylvia von Mackensen.

She described findings from a recent multicentre study of the elderly population in Italy and its two main objectives:

- To describe health status, cognitive functioning, and health-related quality of life (HRQoL) of elderly people with hemophilia
- To evaluate the influence of co-morbidities on the health status and HRQoL in elderly people with hemophilia

To address the needs of the growing elderly population, researchers must assess what contributes to health-related quality of life in the elderly and how elderly individuals manage with chronic disease and other health problems, Dr. von Mackensen said. The Italian study assessed both clinical and geriatric dimensions:

- Clinical assessment: demographic characteristics, family history, hemophilia treatment, hemorrhagic history, viral infections, concomitant diseases, cardiovascular disease, surgery, orthopedic score, and subjective health complaint list
- Geriatric assessment: physical functioning, autonomy in daily life, gait and balance, cognitive functioning, and mental health and depression

The study used a combination of generic HRQoL instruments, an instrument specifically for the elderly, and a hemophilia-specific questionnaire for the elderly developed by the study's investigators. The elderly hemophilia-specific quality of life instrument was validated through this study.

Of the 46 elderly patients with hemophilia registered in Italy, 39 participated in the study and were matched with male controls without bleeding disorders. Clinical data on the patients with hemophilia include:

- Mean age of 69.5 years
- 85% with hemophilia A
- 21% on prophylaxis (79% on on-demand therapy)
- 13% with inhibitors
- 87% with chronic hepatitis C
- 13% with HIV
- 37% reporting chronic pain

The impacts of hemophilia on the health status and physical functioning of elderly patients are considerable, Dr. von Mackensen noted. Medical conditions such as hypertension, viral infections, musculoskeletal problems, and increased mobility challenges, along with impairment affecting dressing, bathing, use of transportation, gait, and balance, often diminish quality of life as patients age. These changes are frequently accompanied by emotional changes and depression. The study found significant differences in orthopedic health and autonomy in daily life between the patients with hemophilia and the controls, but no significant difference in cognitive functioning and mental state.

Sociodemographic factors also affect elderly people with hemophilia, Dr. von Mackensen said. One of the findings of the Italian study showed the impact of hemophilia on the everyday lives of elderly patients: 64% of patients are married, compared with 91% in the control group. "This can result in a worse situation because they need assistance but do not have a wife or children to support them. It is really a big problem because they gradually lose autonomy in daily activities," she said. The hemophilia-specific questionnaire found that 48% of those with hemophilia are afraid of losing their autonomy and 44% fear that their condition is worsening.

#### *Physiotherapy and the Hemophilia "Old Boys"*

Ethelwyn Remmers, Pretoria Academic Hospital, South Africa

Ethelwyn Remmers described physiotherapy as an adjunct therapy for the management of decreased joint and muscle mobility and pain associated with hemophilic arthropathy. Most older individuals with hemophilia are affected by chronic hemophilic arthropathy as the result of target joint development. The main characteristics of chronic arthropathy are pain, reduced movement, deformity, and muscle wasting. Those with hemophilia also seem to have an earlier onset of osteoarthritis.

Evidence points to the efficacy of the treatment of an arthritic patient with different physiotherapy modalities. These modalities may vary according to the required effect and are also used for the treatment of osteoarthritic patients with hemophilia. Different physiotherapy modalities are used to treat the osteoarthritic patient with hemophilia, including:

- Heat and cold treatments: relieves pain and improves circulation using infrared hot packs or lamps, or ice packs and cold packs
- Hydrotherapy: using the buoyancy and resistance of water for exercises (preferably, using a heated pool, as the heat also helps relieve pain)
- Electrotherapy modalities:
  - Ultrasound: relieves pain and may improve circulation
  - Interferential electrotherapy: reduces muscle spasms and relieves pain
- Exercises for strengthening and stretching muscles, to improve suppleness, balance, proprioception, joint mobilization, and relaxation

These modalities are often used in combination.

Remmers presented a case study of Justice Mabo, a 49-year-old man in South Africa with severe hemophilia. Mabo has been visiting the Pretoria Hemophilia Clinic regularly since it opened 11 years ago. Over the years, he has had two serious elbows bleeds, several serious chest infections, and gastric and kidney problems. In 2005, orthopedic surgeons assessed Mabo's swollen, stiff, and painful knees. He was placed on the waiting list for knee replacements and is still waiting his turn for surgery. In 2007, he suffered a serious iliopsoas bleed. Later that year, he had bleeds in his right shoulder, elbow, wrist, and left knee. He subsequently sustained a fall that resulted in a patella fracture and compartment syndrome in his lower leg. At the end of the year, he had another elbow bleed. With every admission to hospital, Mabo received physiotherapy treatment to help reduce pain, maintain muscle function, and improve the range of motion of the affected muscles and joints.

In January 2008, Mabo began a three-month physiotherapy program involving one-hour sessions twice a week. The first two weeks consisted of simple exercises; then treatment progressed to mobilization and strengthening exercises. After a month, the focus turned to balancing and proprioception. Mabo remained committed to the intensive physiotherapy program throughout the three months, which substantially improved his mobility while reducing pain, and he continues to benefit from daily exercise.

People with hemophilia need to be encouraged to do exercises more regularly, Remmers concluded, and physiotherapists have a definite role to play in quality of life of older patients.

#### *Multidisciplinary Approach to the Elderly Hemophilia Patient*

Els Haan, Van Creveldkliniek, Utrecht, the Netherlands

Els Haan described the multidisciplinary approach adopted at the Van Creveldkliniek for caring for elderly hemophilia patients. The clinic was established in 1964 as the first comprehensive hemophilia care centre in the world. Before the clinic was founded, Dutch patients were unlikely to survive childhood because of the absence of treatment. But today, the availability of clotting factor concentrates, prophylactic home treatment, and antiviral treatment, as well as surgical precautions, has helped raise life expectancy to 74 years for patients with viral infections, and 66 years for those with HIV and/or HCV.

In the 44 years since it first opened, the clinic's staff has grown to reflect the advances in treatment of the past four decades:

- Pediatric specialist (2)
- Orthopedic surgeon
- Rehabilitation specialist
- Dentist
- Nurse (4)
- Physiotherapist (2)
- Psychologist
- Hematologist
- Pediatric hematologist
- Hemophilia specialist (2)
- Orthopedic surgeon
- Rehabilitation specialist
- Dentist
- Hemophilia nurse (3)
- Research nurse
- Physiotherapist
- Genetic counsellor
- Social worker
- Data manager
- Secretarial staff (2)

2008

Today, the clinic's multidisciplinary staff also manages the needs of elderly patients. The hemophilia doctor prescribes treatment, while nurses observe outpatients, evaluate their coping abilities with daily activities, and provide education and treatment to inpatients. Social workers help patients cope with the possible psychological consequences of ageing, employment issues, social concerns and family matters, and adaptations at home. The orthopedic surgeon works closely with the hemophilia specialist on musculoskeletal surgery. And finally, physiotherapists and rehabilitation specialists assess joint mobility and muscle function, and prescribe exercise programs. Haan said Dutch researchers have noticed that it takes elderly patients more time to decide to undergo surgery and that they are often wary of being in hospital because of childhood memories of long hospital stays.

Treatment side effects in the Dutch population have included the following:

- Hepatitis B virus (HBV): the majority of affected patients spontaneously cleared the virus, but 5% remain carriers. The HBV test was introduced in the Netherlands in 1970, and HBV vaccination has been available since 1985.
- HIV: 16% of patients are infected. HIV testing has been available in the Netherlands since 1985. Nowadays, 50% are still alive on highly active antiretroviral therapy (HAART).
- Hepatitis C virus (HCV): 98% of patients are infected, with 10-12% clearing the virus spontaneously. Of the chronically infected patients, 20% will develop end-stage liver disease. The HCV test has been available in the Netherlands since 1985.

In 2008, the life expectancy of individuals with hemophilia in the Netherlands is similar to the majority of the Dutch population, owing to treatment with clotting factor concentrates, prophylaxis, self-infusion, precautions for surgical procedures, pregnancy, and childbirth, antiviral treatment, and educational projects at school and work, Haan said.